Gayle Fischer &
Jennifer Bradford

THIRD EDITION

The VULVA

A Practical Handbook for Clinicians

CAMBRIDGE Medicine

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Contents

Glossary

Acanthosis nigricans – a velvety eruption, sometimes with wart-like growths, accompanied by hyperpigmentation in the skin of the armpits, neck, anogenital area and groin

Alopecia areata – an autoimmune condition that causes hair loss with round bald patches that can evolve to complete baldness

Amoebiasis – a tropical infection with *Entamoeba histolytica*, most commonly causing gastroenteritis

Angiokeratoma – harmless raised, purple lesions composed of blood vessels with a hyperkeratotic surface, often found on the labia majora

Apareunia – inability to perform coitus because of a physical or psychological sexual dysfunction

Aphthae – small, shallow, painful ulcers that usually affect the oral mucosa but less commonly affect the vulva

Atopy – a common genetic condition characterised by asthma, hay fever and dermatitis, as well as, in some patients, exaggerated IgE responses

Autoimmune thyroiditis – an inflammatory disease of the thyroid associated with high levels of thyroid autoantibodies

Bleb – a blister filled with fluid (*see also* vesicle)

Campbell de Morgan spots – red papules on the skin containing a proliferation of blood vessels, very commonly found in middle-aged people

Candidiasis – fungal infection caused by *Candida* species, most often *C. albicans*

Cellulitis – a bacterial skin infection characterised by spreading painful erythema, most often due to group A *Streptococcus*

Chancroid – a sexually transmissible tropical infection caused by *Haemophilus ducreyi* and characterised by genital ulcers

Comedones – also known as blackheads: papules with a dark centre caused by a build-up of sebaceous material in hair follicles

Crohn's disease – an inflammatory disease that may affect any part of the gastrointestinal tract from mouth to anus, which causes a wide variety of symptoms

Cytokines – immunoregulatory chemicals

Dermoscopy – a technique for examining skin lesions using a hand-held magnifying device

Desquamation – shedding of the outer layers of the skin

Desquamative inflammatory vulvovaginitis – an uncommon chronic non-infective vulvovaginitis characterised by an introital and vaginal rash, soreness, dyspareunia and discharge

Dowling–Degos disease – a rare disease that causes reticulated hyperpigmentation of the vulval and axillary skin

Dyspareunia – painful sexual intercourse

Erythema – redness of the skin

Folliculitis – inflammation or infection of one or more hair follicles, characterised by a pustular eruption

Fomite – any inanimate object or substance capable of carrying organisms

Fourchette – a small fold of membrane connecting the labia minora in the posterior part of the vulva

Fox–Fordyce disease – a rare skin disorder characterised by the development of itchy bumps around the hair follicles of the underarm area, pubic region and/or around the nipples

Hamartoma – a neoplasm resulting from overgrowth of normal tissue

Hidradenitis suppurativa – a severe, chronic recurrent condition of the apocrine sweat glands, characterised by nodules, pustules and sinuses

Hyperalgesia – pain or discomfort from light touch, causing intolerance of tight clothes

Inguinal – pertaining to the groin

Intertriginous – where two skin areas touch or rub together

Koebner phenomenon – refers to skin lesions appearing in areas of chronic trauma

Leishmaniasis – a skin disease characterised by ulcers and nodules, caused by protozoan parasites of the genus *Leishmania* transmitted by sandfly bite

Lichenification – thickening of the surface of the skin, caused by scratching

Lymphogranuloma venereum – a sexually transmissible disease caused by *Chlamydia trachomatis*, causing genital abscesses and ulcers

Maceration – softening and whitening of skin due to chronic wetness

Macule – a change in skin colour without elevation or depression

Marsupialisation – cutting off the top of a cyst and suturing the cyst edges of the skin

Molluscum contagiosum – a viral skin disease characterised by small umbilicated papules

Morphoea – a condition in which there are areas of skin fibrosis similar to a scar

Mucosal petechiae – red–purple lesions of the skin or mucosa due to extravasation of blood from capillaries

Naevi – birthmarks or coloured skin markings

Neurofibromatosis – a genetic disease in which patients develop multiple soft tumours (neurofibromas) under the skin and throughout the nervous system associated with pigmented skin lesions

Non-sexual acute genital ulceration (NSAGU) – aphthous ulceration of the vulva (*see also* aphthae)

Papilloma – a benign pedunculated tumour

Papillomatous –similar to a wart, raised and rough

Papules – firm raised lesions on the skin

Pedunculated – growing or attaching to a peduncle or stalk

Pernicious anaemia – anaemia due to vitamin B12 deficiency

Pruritus – itch

Punctum – the opening of a sebaceous cyst

Rugose – wrinkled or ridged

Sebaceous adenitis – recurrent inflammation of the sebaceous glands of the labia minora

Sebaceous hyperplasia – a common harmless enlargement of the skin oil glands, which features skin-coloured to yellow-white elevations of the skin

Seborrhoeic keratoses – harmless skin lesions occurring in adulthood, also known as 'age warts'

Spongiosis – a histopathological term meaning intercellular oedema between keratinocytes, or cells found in the epidermis

Stenosed – narrowed

Striae – stretchmarks

Syringomas – harmless sweat duct tumours

Telangiectasia – small, superficial, dilated blood vessels

Tuberous sclerosus – rare, multisystem genetic disease characterised by skin lesions and internal tumours

Umbilicated – marked by depressed spots resembling the umbilicus

Vesicle – a fluid-filled sac within the epidermis

Violaceous – of a violet colour

Vitiligo – an autoimmune disease characterised by loss of skin pigment

Vulvovaginitis – inflammation of the vagina and vulva

Chapter 1

The Basics

Contents

Patients with vulval problems have often spent many years in fruitless pursuit of a diagnosis and effective treatment. The reasons for this are varied.

- Most vulval conditions are chronic dermatological diseases that cannot be cured. They must be managed. Such conditions on the vulva often look and behave differently from the same conditions on other parts of the skin. Management strategies that are effective for non-genital skin must often be modified in order to be effective on the vulva.
- The vulva and vagina are in the centre of the lower pelvis and are closely related to other pelvic organs, bound to them by the myo-fascial structure of the pelvic floor (see Figures 1.1, 1.2 and 1.3). Referred vulval pain from other pelvic viscera and from the lumbosacral spine and hip joints is an important concept in understanding vulvo-vaginal disorders.
- Dermatological disease of the vulva has far more importance for the patient than the same disease on less emotionally significant areas of the body. Patients frequently present, not with symptoms of the vulval disease, but of its sexual or relationship consequences. It is therefore no wonder that in the past, women with vulval disorders have been unfairly told 'it's all in your head'.
- Vulval disease comes with a significant emotional overlay. Embarrassment commonly prevents patients from seeking help. Doctors are only human, and embarrassment can affect them too: patients often tell us that they were not examined. History taking is difficult because of the intimate nature of a woman's symptoms. A detailed sexual and environmental history is essential, and eliciting such histories takes patience and empathy. Patients may either avoid saying what is really on their mind or, alternatively, pour out huge amounts of disorganised, emotionally charged information. It is important to help them to organise their thoughts. Start at the beginning and get them to

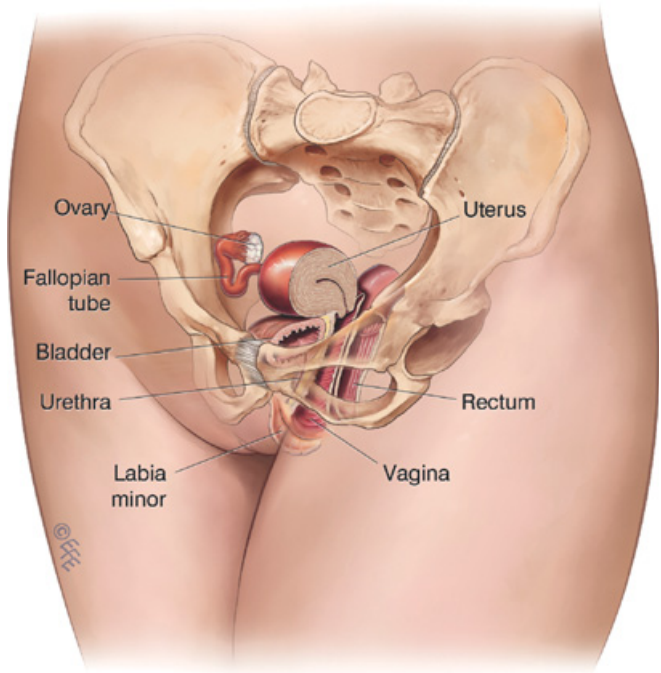

Figure 1.1 The female reproductive tract. With permission from Dr Levent Efe, CMI

think back on how their complaint evolved and how they came to be in your consulting room.

- An emerging issue in the management of vulval disease is the transgender patient. Male-to-female patients with a neovagina and female-to-male patients who have retained a vagina that has been changed by exposure to androgen form a unique group with their own special needs.

Understanding Vulval Conditions

The management of patients with vulval disease fundamentally requires an understanding of dermatological diagnosis and therapy, especially those skin diseases with a predilection for this part of the body.

However, dermatological knowledge is not enough. An understanding of gynaecology, gastroenterology, urology, vaginal bacteriology, spinal function and dysfunction, and finally sexual medicine are all essential for optimal management.

Another challenge is the group of patients who appear to defy diagnosis. We believe that this group is very small indeed, and that it is possible to classify virtually all patients. However, achieving this relies on very thorough history taking and on combining many different medical disciplines. The more difficult vulval cases are always multifactorial. The term 'vulvodynia' (vulval pain of unknown origin) is not used in this book, as we believe that a rational diagnosis can eventually be found for almost all vulval symptoms.

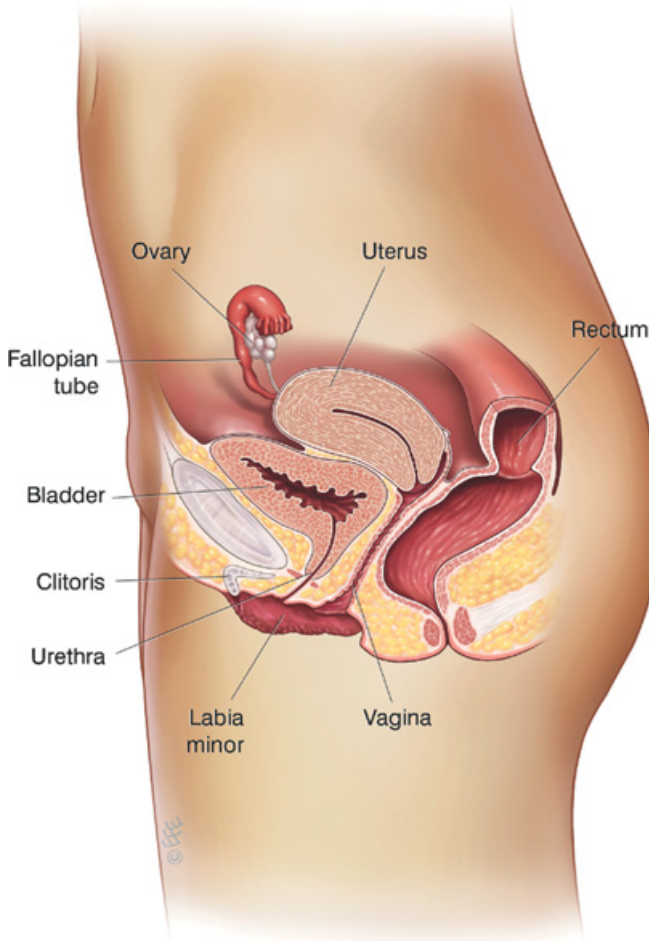

Figure 1.2 The female reproductive tract: side view. With permission from Dr Levent Efe, CMI

The purpose of this handbook is to introduce these concepts and to provide practical management recommendations. These are based on our 60 years' collective experience of helping patients with vulval disease, as well as published research by ourselves and others. We hope that this book will give clinicians the tools to approach vulval patients with confidence, and that this will in turn improve the lives of many women.

Anatomy

The Vulva

The vulva is not only part of the skin but also the entrance to the genital tract. It is essential to understand that vulval skin extends all the way to the hymen. This means that rashes within the vaginal introitus are still classified as vulval, and the patient must be instructed to apply any treatment far enough inside to adequately control these rashes.

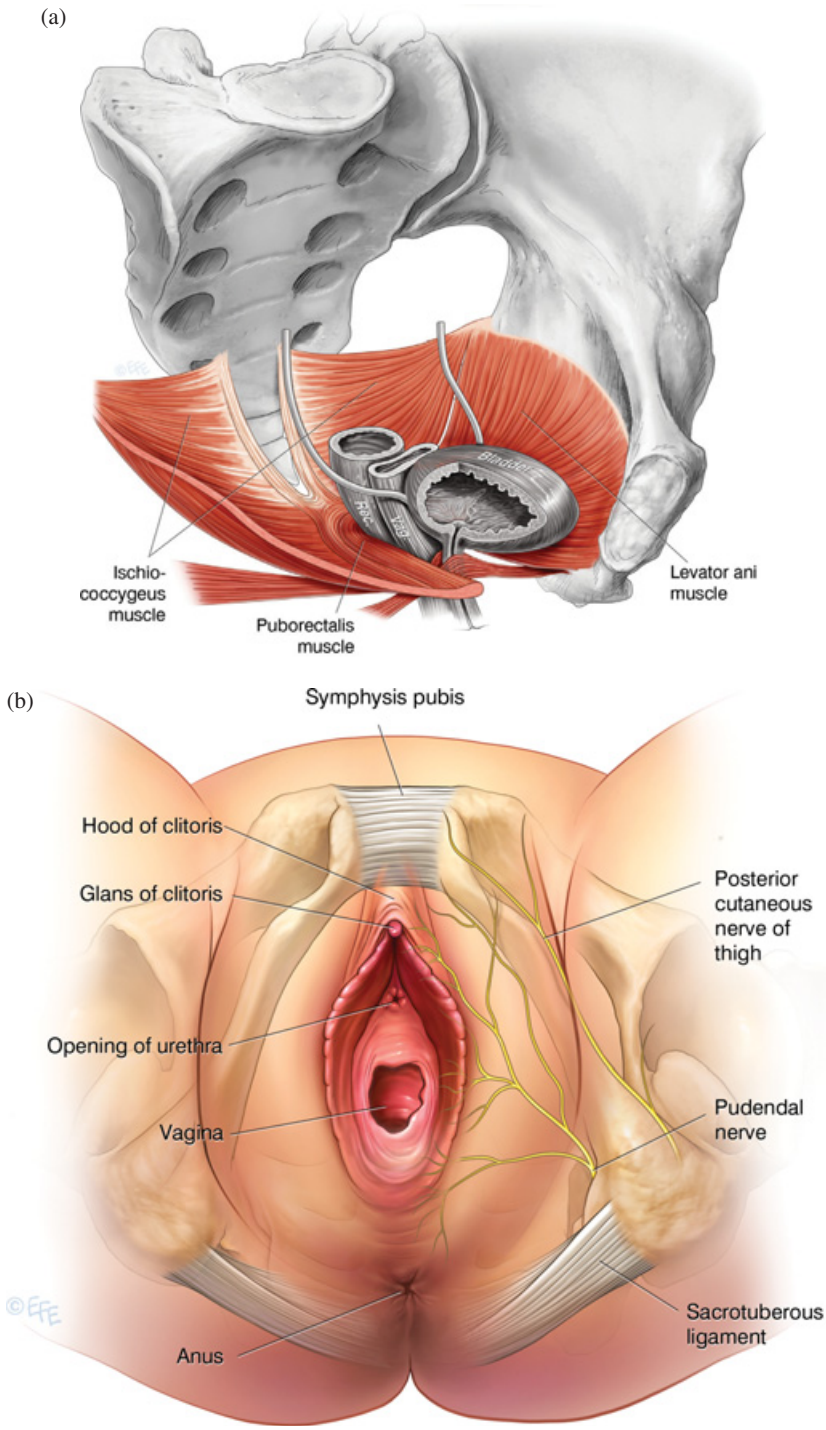

Figure 1.3 (a) The pelvic floor. (b) View of the vulval area. With permission from Dr Levent Efe, CMI

The vulva is exposed to many potential irritants, which can result in dermatological symptoms. These include:

- menstrual fluid
- urine
- faeces
- sweat
- vaginal discharge, both normal and abnormal
- semen
- tight clothes
- lubricants
- perfumed products including toilet paper, wet wipes and feminine hygiene products
- medications, both vaginal and oral
- pads and panty liners
- hair removal practices.

The Vagina

The vagina is the conduit between the uterus and the vulva. Its mucosa is prone to similar diseases as in the mouth. Located in the middle of the lower pelvis (see Figure 1.1), its anatomical relations include:

- bladder and urethra
- cervix
- rectum and anus
- utero-vesical and recto-vaginal peritoneal pouches
- sacrum and coccyx.

The Pelvic Floor

The pelvic floor is a complex myo-fascial structure that encompasses the entire pelvis. It acts as a conduit for pain referral throughout the pelvis. It is closely related to the muscles of the lower back and hip, and may be affected by lumbo-sacral or hip dysfunction.

One way of understanding functional pelvic floor anatomy is to think of it as having upper and lower parts. The upper part supports the bladder neck, cervix and upper rectum. The lower part supports the urethra, vagina, lower rectum and anus. When considering the lower pelvic floor, it is helpful to think of the attached structures arranged posterior to anterior: lumbo-sacral spine and hip joints, rectum and anus, vulva and vagina, and urethra and bladder. This is because referral of pain and dysfunction in the lower pelvis tends to be from posterior to anterior (see Chapter 8 on pain).

The Innervation

The innervation of the lower vagina, vulva and anus is from sacral nerve roots S2, 3 and 4 via the pudendal nerve. The anterior vulva is supplied by the genital branch of the genito-femoral nerve (L1 and L2) and the ilio-inguinal nerve (L1). Thus lumbo-sacral, coccygeal and even lower thoracic spinal disorders may produce referred vulval pain (see Chapter 8 for a discussion on pain and dysfunction referral in the pelvis).

Clinical Presentation

The majority of patients presenting with chronic vulval symptoms have a skin disease such as eczema, psoriasis or lichen sclerosus, or are suffering from chronic vulvo-vaginal candidiasis. Many have a personal or family history of the vulval skin condition or have evidence of it elsewhere on their skin, so a general skin examination is very helpful. Patients with eczema are usually atopic. This historical information can provide very helpful clues to a possible diagnosis.

When a patient presents with a vulval complaint, she usually complains of one or more of the following symptoms. Patients sometimes have trouble communicating their thoughts. It can be helpful to run through this list with them in order to better delineate their real story.

- itch
- pre-menstrual or post-menstrual exacerbation of symptoms
- irritation
- soreness
- pain
- dyspareunia
- burning
- stinging
- stabbing
- crawling sensations (formication)
- awareness of the vulva
- dysuria.

The duration of symptoms, any precipitating and exacerbating factors, and previous treatments should be recorded. Bladder, menstrual and bowel function also needs to be recorded, as vulvo-vaginal disease is frequently associated with dysfunction in these systems. Spinal and hip joint disease and dysfunction in voiding may play a significant role in vulval symptoms and should be recorded.

What Do the Symptoms Mean?

Itch and irritation are usually due to a non-eroded inflammatory skin condition. **Soreness and pain** are often due to erosions or fissures, either caused by a skin condition or secondary to excoriations produced by scratching. They can less often be due to neuro-muscular disorders. Burning, stinging, stabbing, formication and vulval awareness are usually due to a neuro-muscular dysfunction.

Dyspareunia

Dyspareunia means pain during sexual intercourse. We find it helpful to categorise dyspareunia into abdominal and vulvo-vaginal types.

Abdominal dyspareunia is sexual pain experienced in the lower abdomen, as it relates to the upper pelvic floor. It usually relates to disease or dysfunction at or above the level of the cervix, for example endometriosis.

Vulvo-vaginal dyspareunia is experienced at the vaginal entrance, or further up into the vagina proper. It is caused by disease or dysfunction at the level of the lower pelvic floor. This pain is usually caused by a vulval and/or vaginal skin condition. It can also be caused by disease or dysfunction of the bowel, anus, the lumbo-sacral spine and hip joints.

Important points to elicit in a history of dyspareunia are:

- Is the onset of pain:
  - with foreplay (or masturbation)?
  - during vaginal intercourse?
  - after intercourse is concluded?
  - gradual or sudden?
- Is the duration of pain
  - at intromission, and then improves?
  - at intromission and then relieves rapidly after withdrawal?
  - throughout intercourse then continues for a variable time after intercourse has ceased?
- Where is the site of entry (or vaginal) dyspareunia (with a mirror if necessary)?
- What is the nature of the pain: tearing, splitting, dull or sharp?
- What relieves the pain?
- Is the pain severe enough to result in apareunia?
- Is the same pain also experienced in a non-sexual context, particularly tampon insertion or with pressure on the vulva (especially with tight clothes)?

History Taking

The Dermatological History

The following factors in a patient's dermatological history may be relevant to the vulva:

- atopic disease (e.g., eczema, hay fever or asthma)
- psoriasis
- autoimmune conditions (e.g., systemic lupus erythematosus, Sjogren's syndrome, autoimmune thyroiditis or pernicious anaemia)
- allergic reactions to drugs or topical therapy
- lichen planus, particularly oral.

The Gynaecological History

Menstrual disturbance often results in more frequent use of menstrual protection, leading to more contact irritation.

Oestrogen status is important. It is low in post-menopausal and lactating women, and of course in pre-pubertal girls. In general, vaginal candidiasis does not occur in a low-oestrogen environment, and so a post-menopausal woman who does not use systemic or vaginal oestrogen should be assumed not to have candidiasis, unless proven otherwise.

Gynaecological surgery including laser surgery, even of a very minor nature, may cause or worsen vulval disorders.

Patients often assume that their symptoms are due to sexually transmissible infections (STIs). It is important to assess this possibility, but investigation is frequently negative.

Herpes simplex infection of the vulva is an acute event that is rarely by itself responsible for chronic vulval symptomatology. However, it can be the precipitating factor for chronic vulval dermatitis, entry dyspareunia, neuropathic pain or, very occasionally, anxiety or obsessive compulsive behaviour.

The Urological History

Urinary incontinence has a strong association with vulval disorders. This is partly due to simple vulval maceration caused by contact with urine and pads, but also because vulval skin disorders often cause or worsen bladder dysfunction via the pelvic floor.

Vulvo-vaginal disorders often result in bladder dysfunction disorders, either infective or non-infective. Many patients, however, will present with the secondary bladder symptoms only, and it will become apparent only after careful history taking that the real culprit is in the vagina or vulva.

Urological surgery, even diagnostic cystoscopy, may cause vulval symptoms.

Very occasionally, symptoms originating from the bladder may be experienced in the vulva and vagina without obvious bladder symptomatology. This is especially true of urethral disorders.

The Gastroenterological History

Bowel disturbances may cause or worsen vulvo-vaginal disorders. Diarrhoea often results in peri-anal and vulval irritant contact dermatitis. Constipation tends to 'wind up' the posterior pelvic floor, and may lead to vaginal dyspareunia of neuro-muscular origin.

Haemorrhoids make anal cleansing after defecation more difficult, and often causes dermatitis due to excessive cleaning.

Faecal incontinence must always be asked about in women who have had vaginal deliveries. It is surprising how frequently this occurs.

The presence of diseases that produce problems with absorption, most commonly coeliac disease, may result in reduced effectiveness of medications.

Crohn's disease may rarely directly affect the vulva and may do so in the absence of active gastro-intestinal disease.

The Musculoskeletal History

It is essential to enquire about the following:

- back injuries (motor vehicle accidents, falls onto the coccyx, heavy lifting, falls causing back injury)
- sciatica
- hip joint pain, arthritis and injury
- lumbo-sacral osteoarthritis with/without disc protrusion
- spinal surgery
- exercise routines
- weight gain.

The Environmental History, 'Secret Women's Business'

It is very likely that your patient has her own personal hygiene beliefs, practices and rituals. These are often cherished and difficult to change. You need to find these out – and you won't unless you ask. Ask specifically about possible irritants and allergens including:

- washing routines: frequency, use of soap, bubble baths and perfumed oils
- sanitary pads, incontinence pads, liners and tampons
- lubricants
- condoms
- shaving and waxing
- douching
- underwear, G-strings
- over-the-counter and home remedies
- exercise routines, including clothing worn
- sports, particularly cycling and horse riding
- swimming, saunas and spa baths.

The Psychological History

Although this need not be exhaustive, it is important to determine whether:

- the patient is still able to enjoy intercourse
- her partner is sympathetic
- her problem has ended any previous sexual relationships
- she is suffering from depression, shame or anxiety independent of, or related to, her problem
- she has had any traumatic sexual experiences, either recently or as a child
- if the patient is a child, consider possible sexual abuse and how this could affect the family
- she has beliefs about her condition that are related to misleading information, often from the Internet
- she is angry with the medical profession regarding previous treatment failures.

Patient Beliefs

It is important to find out what patients believe is responsible for their symptoms and also their attitude to your possible treatments. Examples of beliefs that may impact on your therapeutic strategies include:

- the assumption that symptoms are due to thrush (although this involves 20% at most)
- fear that treatment with oestrogen will predispose to breast cancer
- fear that the use of tampons will result in toxic shock
- a belief that symptoms are due to genital herpes, even when there is no objective evidence
- fear of the use of any form of corticosteroid (it will 'thin the skin')
- fear that their skin condition is transmissible

- fear of cancer
- a belief that their condition is the result of a sexual encounter
- a belief that their condition was transmitted from contact with a fomite (e.g., a toilet seat).

Summary of History Making

- symptoms
- cycling of symptoms
- duration of problem
- previous treatment and whether it has helped, even briefly
- personal habits
- dermatological personal and family history
- atopic disease
- dyspareunia
- effect on sexual relationships
- gynaecological history
- gastro-intestinal history
- urological history
- general medical history
- psychological history
- medications, including over the counter
- allergies
- secret women's business.

Examination

The vulva and vagina display a high level of anatomical and colour variation. Some of this is congenital, some age-related and much due to vaginal childbirth. Female genital mutilation will present from time to time, and there is also the increasingly common phenomenon of cosmetic reduction labiaplasty. The clinician's ability to define what is 'normal' on examination will therefore be determined by their clinical experience in women's medicine.

Although most vulval problems are fundamentally dermatological, the typical appearance of most skin diseases is very different when they occur on genital skin, and may be quite subtle. A good light and adequate access to the genital skin by careful patient positioning is essential. A couch with foot rests is ideal, however many patients are humiliated and stressed by having their feet placed in stirrups, and this is not essential for an adequate vulval examination.

Inspect the groins and pubic area first, then the external labia majora, the inter-labial sulcae and then the vaginal introitus. The clitoral hood should be gently retracted to inspect the glans clitoris. Include the peri-anal area and natal cleft with the patient lying on her side. It may then be helpful to perform a general skin and oral examination to look for clues that will help with diagnosis, for example, of possible psoriasis or lichen planus. Include the buccal mucosa as this may give a clue to lichen planus, which is not always symptomatic.

It is very helpful to use a hand mirror to allow the patient to demonstrate the area of her concern, and so that the clinician can show the patient the areas that actually require application of any topical treatments. Many women have never inspected their own vulva and it is important to familiarise them with their own anatomy and the names of the components.

Common Physiological Variations

Common physiological variations of the vulva include:

- pigmentation: Non-Caucasians commonly demonstrate hyper-pigmentation of the labia minora. However, patients of all races may develop vulval hyper-pigmentation, which becomes more apparent with advancing age
- size of labia minora: pre-pubertal girls have very small labia minora, however, virtually all normal adult pre-menopausal women possess labia minora. After menopause, these may reduce significantly in size but absence of labia minora, particularly if it is asymmetrical, should be cause for concern regarding possible scarring vulval conditions such as lichen sclerosus and lichen planus
- asymmetry of the labia minora
- size of the vaginal opening
- degree of rugosity of the mucosal surface of labia minora and vaginal mucosa
- vulval papillomatosis (tiny projections from the inner surface of the labia minora, a normal variant)
- prominence of sebaceous glands (Fordyce spots)
- erythema: in some patients, the sulcus between the minora and majora is persistently red in the absence of any pathology
- length and density of pubic hair
- clitoral size
- prominence of gland openings (these may form obvious pits on the mucosal surface)
- amount of normal discharge

apparent webbing at the base of the fourchette.

Finding Abnormalities during the Physical Examination

Vulval rashes can be subtle, and initial examination may suggest a normal vulva. Look carefully for:

- increased erythema of labia minora, and the sulcus between minora and majora
- wet desquamation (scale), especially in the inter-labial sulcus
- fissuring (skin splits): you may have to gently stretch the skin to find these
- textural change: lichenification of labia majora and peri-anal skin, 'cigarette paper wrinkling'
- evidence of erosions or ulcers
- mucosal petechiae
- colour change: light or dark
- atrophy of mucosa
- presence of any unusual lesions.

Do not be surprised if there are no abnormalities. Many patients with significant vulval symptoms are normal on examination. This finding means that either they have a skin disease that is episodic, or that their symptoms are not caused by a dermatological problem.

The Speculum Examination

Many vulval skin diseases involve the vulva only, and there is no need for a speculum examination. The diseases that do extend into the vagina are often so uncomfortable that speculum examination is very difficult and may need to be postponed. While there is a place for examination under anaesthesia, initial treatment of the vulval problem may make speculum examination in the consulting room possible at a subsequent visit.

If when you examine the vulva the introitus is involved and there appears to be extension into the vagina, attempt a speculum exam using a small, straight-bladed instrument. Remember, you are not taking a Pap test, and therefore a complete view of the cervix is not essential. Your aim is to visualise enough of the vaginal wall to determine the degree of intravaginal inflammation.

Look for:

- erythema, and whether it is confluent or patchy; refer to the oral mucosa as a guide to what is normal
- erosions
- petechiae
- degree, colour and type of discharge.

In the rare case of intra-vaginal adhesions, a one-finger digital examination will usually give more information than a speculum examination.

Investigations

The most commonly performed investigations in patients with vulval disease are:

- low vaginal swab for *Candida* (or rarely bacterial) infection
- viral swab for polymerase chain reaction testingfor herpes simplex if suspected. This must be taken from an ulcer or erosion and the lesion should be firmly swabbed. This will be uncomfortable. Warn the patient of this prior to performing the test.
- vulval swab for *Candida* or bacterial infection
- scraping of vulval skin for dermatophyte (tinea) infection: this is done with a scalpel blade into a specimen jar (check with your pathologist)
- skin biopsy.

All patients should have a low vaginal swab taken for culture. A high vaginal swab taken during a speculum examination is unnecessary, and may not reflect the environment of the lower vagina. Furthermore, passing a speculum past inflamed vulval skin is often exquisitely painful.

A vulval swab or scraping is performed if infection of the hair-bearing skin is suspected clinically.

A skin biopsy is performed if there is a visible rash or lesion that is diagnosable by biopsy, particularly for suspected malignancy. It should never be performed on clinically normal skin. Specific situations that always require biopsy are:

- a white lesion
- a unilateral lesion

- suspicion of malignancy: persistent ulcer, nodule, fissure, pigmented lesion, white hyperkeratotic lesion
- a lesion that has not adequately responded to initial treatment.

Other tests that may be relevant include:

- mid-stream urine for microscopy and culture if urological symptoms are present
- patch testing if a contact allergy is suspected
- serum follicle-stimulating hormone (FSH) may in some cases be required to assist in confirming menopause
- tests for systemic autoimmune disease may be relevant in patients with conditions such as lichen sclerosus.

Colposcopy is usually unnecessary in the diagnosis of vulvo-vaginal disorders, except when vulval intraepithelial neoplasia is suspected. The use of acetic acid on the vulva is unnecessary in benign skin diseases and usually produces significant stinging that patients find traumatic.

Patient Categorisation

At the end of your history and examination, you will find that patients with vulval disease fall into the following broad categories:

- patients with a rash and non-cycling symptoms
- patients with a rash and cyclical symptoms
- patients with a rash or lesion and no symptoms
- patients with no rash but symptoms.

This will be discussed more later in the book.

Summing Up: Common Things Occur Commonly

The majority of patients presenting with chronic vulval symptoms have a vulval presentation of a common skin disease such as eczema, psoriasis or lichen Sclerosus or are suffering from chronic vulvo-vaginal candidiasis.

Many will have a personal or family history of the same condition elsewhere on the skin. Many patients with eczema are atopic. This historical information can provide very helpful clues to a possible diagnosis.

Chapter 2

Using Topical Steroids on the Vulva

Topical steroids on the vulva are very safe if used properly and supervised regularly. We provide a guide to the safe and effective use of these essential drugs.

Topical corticosteroids (TCSs) are a dermatological therapeutic mainstay. They are appropriate in the treatment of most inflammatory dermatoses everywhere on the skin. This includes the vulva.

Used in different ways in different situations, and if used appropriately and correctly, TCSs are very safe. For instance, in lichen sclerosus and lichen planus, they are used continuously, but in psoriasis and dermatitis, they are used intermittently, first for initial treatment and then for flare ups. Details of how to treat these conditions are found in the appropriate chapters.

These time-tested medications, which have been available for over sixty years, have been much maligned in the last fifteen. So-called corticosteroid phobia has resulted in doctors and patients being too scared to use them adequately; this then denies patients the right sort of treatment that will greatly improve their quality of life.

In general, the following principles will keep your patients safe:

- Use ointment bases in preference to creams. Ointments adhere better to genital skin, and creams contain preservatives that may sting.
- Make sure your patient has a corticosteroid-responsive dermatosis.
- Know your corticosteroid potencies (see Table 2.1).
- Titrate the dose of your treatment to the severity of the condition that you are treating.
- Start strong and work down.
- Clobetasol propionate is super-potent and therefore it is much easier to get into trouble with than weaker preparations. Do not ever leave a patient on a potent TCS without regular monitoring. It is mentioned in many publications on vulval disease, but is only one of many TCSs available, and in many situations, it is not the best. In Australia, it is only available through compounding chemists; however, in most other countries, it is a commercial preparation.
- Use clinical response rather than guidelines for length of treatment. One of the biggest mistakes made by clinicians is under-treatment. Your patient's skin should be back to subjective and objective normality before you reduce.
- Educate your patient that it is OK to use TCSs long term. Most vulval dermatoses need long-term treatment. Peri-anal skin is less tolerant of TCSs than the vulva. In general, you should use weaker preparations there.

Table 2.1 Topical corticosteroids
This table gives you a selection of TCSs that we have found useful and affordable.
Not all of them are available in every country. Remember that the percentage on the tube does not necessarily indicate that the product is more or less potent.
If you can access methylprednisolone aceponate and or aclometasone dipropionate 0.05%, we find these particularly well tolerated and unlikely to produce any side effects, even with very long-term use.

Potency class	Generic name	Comments
Mild	Hydrocortisone 1%	Useful in dermatitis, psoriasis and maintenance treatment of lichen sclerosus
Moderate	Triamcinolone acetonide 0.02% Clobetasone butyrate 0.05% Methylprednisolone aceponate 0.01% Desonide 0.05% Betamethasone valerate 0.02%	Useful in dermatitis, psoriasis and maintenance treatment of lichen sclerosus
Potent	Mometasone furoate 0.01% Betamethasone dipropionate 0.05% Betamethasone valerate 0.05–0.1%	Useful for initial treatment of all inflammatory dermatoses but in general not for very long-term use outside of lichen planus
Super potent	Clobetasol propionate 0.05% Halobetasol propionate 0.05%	Take care! Only for severe lichen sclerosus, lichen planus and very lichenified dermatitis. Monitor carefully Same precautions as for clobetasol propionate

- Avoid combination products containing antibiotics or anti-fungals. Long-term use can result in sensitisation. Genuine allergies to topical steroids are rare.
- In dermatitis and psoriasis, it is often possible to substitute moisturisers for topical steroids once the condition is controlled.
- However, we do not recommend substituting calcineurin inhibitors such as pimecrolimus and tacrolimus for TCSs, or use them just because they are not ‘steroids’. They are expensive, they sting and the risks of long-term use are unknown. This is also true for the PD4 inhibitor crisaborole.
- The vulva is less than 1% of the total body surface. The use of TCSs here will not result in systemic absorption.
- The commonest corticosteroid side effect is redness. This always gets better with a reduction in dose.
- As long as you are following these rules, atrophy is most unlikely. Atrophy will be evidenced by tears during intercourse or striae. Both are usually seen only in unsupervised patients using potent preparations for long periods of time.
- Occasionally you may produce telangiectasia, and in small girls, even early pubic hair. Does this really matter when the patient is getting better?
- Do not be scared of TCSs. Embrace them!

Chapter

3

Red Vulval Rashes

Contents

The most common presentation of a vulval skin problem is an itchy red rash. This group includes inflammatory dermatoses, infections, hypersensitivity reactions and one malignancy.

Dermatitis, psoriasis and chronic vulvo-vaginal candidiasis are all very common causes of red, itchy rashes with variable degree of scaling. Corticosteroid-induced dermatitis occurs when moderate to potent topical corticosteroid is used for long periods of time. Tinea is uncommon and extra-mammary Paget's disease and oestrogen-hypersensitivity vulvitis are rare.

On first sight, all of these conditions look much the same. A combination of history taking, investigation and response to therapy will ultimately enable a diagnosis and effective treatment.

Dermatitis

Dermatitis is common and usually dominated by itch. There are two forms: endogenous and exogenous.

Endogenous dermatitis (or eczema) includes inflammatory dermatoses that are intrinsic to the patient. The majority of these patients are atopic, and their condition is a localised form of atopic dermatitis. History taking often reveals asthma, hay fever or dermatitis on other parts of the skin, or a strong family history of atopy.

Exogenous dermatitis describes inflammatory skin disease caused by exogenous agents: allergens and irritants.

All forms of dermatitis have in common a histological appearance known as 'spongiosis'. Spongiosis is oedema of the epidermis, 'Spongiotic dermatitis' is therefore a term that is

commonly used by pathologists when describing dermatitis. It is not however a clinical diagnosis.

The forms of endogenous dermatitis that involve the vulva are:

- atopic dermatitis
- seborrhoeic dermatitis.

The forms of exogenous dermatitis that involve the vulva are:

- irritant contact dermatitis
- allergic contact dermatitis.

The following conditions, which will be discussed later, resemble chronic dermatitis but have unique characteristics:

- chronic vulvo-vaginal candidiasis
- oestrogen-hypersensitivity vulvitis
- corticosteroid dermatitis (peri-orificial dermatitis).

Presentation

Patients with dermatitis are almost always itchy, but if the mucosal surface is involved, they will also experience burning, and excoriations from scratching and fissures that are commonly found in all vulval dermatoses will produce pain and dyspareunia.

The symptoms are normally worse at night and interfere with sleep. Clothes and activities that increase heat, sweat and friction often exacerbate dermatitis.

Examination

Examination shows poorly defined erythema of the labia and perineum that may extend onto the mons pubis and the inner thighs. In long-standing cases, lichenification is common, resulting in rugosity of the labia majora, and peri-anal skin. The vestibule is erythematous, and there may be white plaques present on the mucosal surface.

Close examination often reveals fissuring, particularly around the introitus or in the inter-labial clefts. The vagina is not involved, but patients with dermatitis may report an offensive discharge. This is actually desquamation, which remits when the dermatitis is controlled. Dermatitis is worsened by super-infection, and if this is identified on vulval and vaginal swabs, it should be actively treated.

Atopic Dermatitis

Most patients have a current or past history of atopic dermatitis on other parts of their skin or of asthma or hay fever. Atopic dermatitis of the vulva may occur at any age. It usually responds readily to simple management. Very occasionally, patients with severe lichenification may be treatment resistant (see Figure 3.1).

The distribution of the rash includes the labia majora and minora, perineum and sometimes the peri-anal skin. The vagina is not involved.

Atopic patients are typically intolerant of irritants. They are unable to use soap or wear wool, occlusive clothing without discomfort.

Post-menopausal women may experience vulval atopic dermatitis for the first time in connection with oestrogen deficiency. They suffer not only from vaginal dryness but also

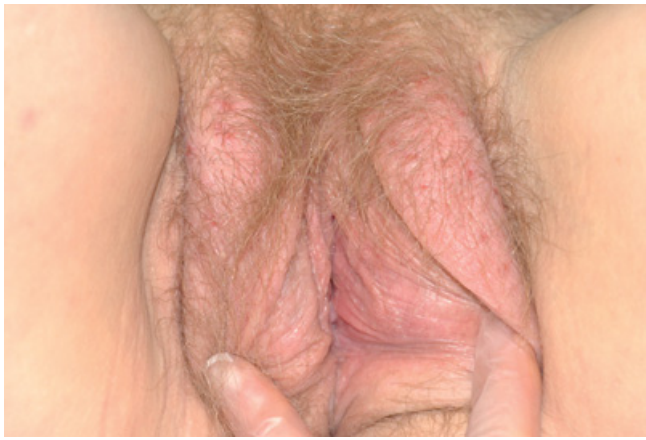

Figure 3.1 Atopic dermatitis. Note excoriations due to scratching

from itching of the labia. In this group, urinary incontinence and therefore pad wearing will exacerbate the problem.

Seborrhoeic Dermatitis

Seborrhoeic dermatitis is very common as a cause of dandruff, but it is relatively uncommon as a cause of vulvitis. Such patients are not atopic but do have chronic problems with an itchy, scaly scalp and sometimes rashes on the central chest and in the axilla.

It can be differentiated from atopic dermatitis by the involvement of other typical sites such as the scalp and axilla, as well as a negative history of atopy.

In vulval seborrhoeic dermatitis, the extent of the rash is often more extensive and less itchy compared to atopic dermatitis extending into the inguinal folds, inner thighs and mons pubis. There is a characteristic greasy, scaly surface. The vagina is not involved. It is a very chronic condition that tends to be exacerbated at times of stress.

The cause of seborrhoeic dermatitis is unknown but, in some patients, it is thought that the presence of Malassezia yeast on the skin is a trigger. For this reason, some patients benefit from the use of anti-fungal creams in addition to topical corticosteroids.

Irritant Contact Dermatitis

Case Study

A 32-year-old woman requests your help for low libido of six months' duration. Questioning reveals dry vaginal dyspareunia for about nine months. She is in a long-term relationship. Ten months ago, she had started an intensive exercise programme to lose weight in response to her partner's negative comments.

Examination reveals a well-demarcated erythematous rash involving the hair-bearing vulval skin and inter-labial sulcae. A vaginal culture is negative.

This woman's fundamental problem is an irritant contact dermatitis that has produced entry dyspareunia and low libido. She is advised to exercise in loose clothing rather than stretch leggings, and use 1% hydrocortisone ointment twice daily to the affected skin for one month.

At follow-up at one month, the dyspareunia is largely improved, but the low libido remains. The woman reports resentment about her partner's continued criticism of her physical appearance. She and her partner are referred for relationship counselling.

Vulval dermatitis due to chronic exogenous irritants presents with persistent low-grade erythema and scale in the area in contact with the irritant. In most cases, only the labia majora and perineum are involved.

Atopic patients are more prone to dermatitis from irritants.

When irritant dermatitis is not associated with atopy, there is often a severe compromise of the cutaneous barrier. This occurs in patients incontinent of urine and/or faeces, who constantly wear macerated incontinence garments or who are confined to a wheelchair. Such patients are often in nursing homes. They present a very difficult challenge.

Women who engage in intense sporting activities, particularly bicycle riding, may develop a chronic vulvitis from friction, occlusive nylon sports clothes and sweating. In the case of serious cyclists, vulval oedema may become a disabling problem.

Some feminine hygiene products may also cause irritant contact dermatitis and this includes the use of shower gels, perfumed wipes, sprays, douches and essential oils. Many women feel the need to clean the genital area more thoroughly than other parts of the skin, and unintentionally produce dermatitis in their attempts to feel 'fresh'. The daily use of pantiliners also falls into this category.

Tight, occlusive nylon clothes, G-string underwear, pantyhose and control garments can also cause irritation by producing constant friction.

Waxing and other hair removal products may cause irritation. Waxing often produces a folliculitis.

The constant use of imidazole anti-fungal creams on the vulva frequently causes an irritant reaction.

When skin is inflamed, possible irritants will often aggravate the condition and things that are normally tolerated may begin to cause a problem. Friction from intercourse may become intensely uncomfortable and contact with semen and saliva may cause burning and stinging.

Allergic Contact Dermatitis

Allergic contact dermatitis results from contact with a true allergen: a substance to which a patient mounts a Type IV allergic response, resulting in severe, vesicular, eroded dermatitis (see Figure 3.2). Fortunately, it is a rare event.

It can present either as a sudden onset acute dermatitis or a chronic non-responsive condition that appears frustratingly resistant to treatment. Because of the severity of the allergic reaction, pain and soreness is often more of a feature than itch. It often extends to the skin outside of the area of direct contact and may be found in other places such as the fingers or eyelids as a result of accidentally transferring the allergen.

The most common cause of allergic contact dermatitis of the vulva is medications, both prescribed and over the counter. These include azole anti-fungal creams, oestrogen creams, antibiotics, products containing tea tree oil, topical anaesthetics, particularly benzocaine, other vaginal creams and topical corticosteroids. The latter are particularly difficult to identify as the cortisone both causes an allergic reaction and suppresses it simultaneously. Compounded products containing amitriptyline may also cause allergic contact dermatitis.

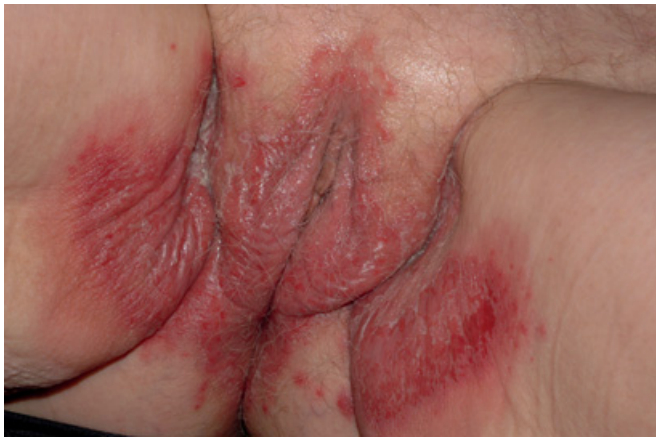

Figure 3.2 Allergic contact dermatitis

When moisturisers, lubricants, sanitary pads and toilet paper cause an allergic contact dermatitis, the culprit is usually the perfumes and preservatives in these products.

The use of wet wipes containing the preservative methylisothiazolinone has become a worldwide phenomenon in causing contact dermatitis of the genital region in adults and children.

Latex in condoms may cause allergy. Polyurethane condoms are non-allergenic and may be substituted.

When vulval dermatitis is of sudden onset, is severe and related to the use of a new product, suspect allergic contact dermatitis. It is also a diagnostic possibility in recalcitrant or severe cases of dermatitis. The allergen is identified by patch testing, a painstaking and sub-specialised process that requires referral to a dermatologist.

In rare cases, seminal fluid may cause true contact allergic reaction. Patients who suffer from this experience severe irritation when their partner ejaculates. This is followed by persistent vulval and vaginal dermatitis. Many of them discover for themselves that condoms protect them and this history is a clue to the nature of the problem. A confounder, however, is that seminal fluid can be irritating to an already inflamed vagina without true allergy. If a patient gives this history, referral to a dermatologist is recommended.

Corticosteroid Dermatitis

Corticosteroid dermatitis, also known as peri-orificial dermatitis, is not a true dermatitis, but a side effect of the use of potent corticosteroid on the vulva. It usually occurs when patients use potent, fluorinated corticosteroids continually for long periods of time and is very unlikely with weak corticosteroids, even when these are used chronically. Patients complain that although their condition is not well controlled with their medication, any attempt to withdraw it results in even more severe symptoms.

Patients commonly complain of a constant burning sensation. The labia majora and minora become erythematous or even violaceous (see Figure 3.3). Telangiectasia may appear and skin fragility may result in fissuring and tearing during intercourse. Pustules and papules can occur but, scaling is not present.

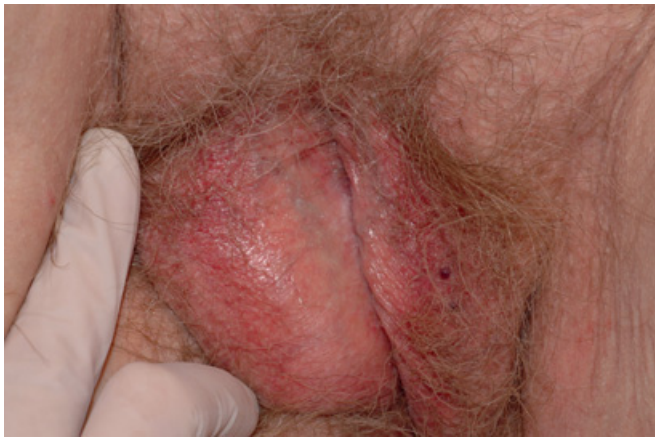

Figure 3.3 Corticosteroid dermatitis

It is important to recognise this symptom as a side effect of corticosteroid. If it is not differentiated from ordinary dermatitis, there is a risk that even stronger preparations will be used, which will exacerbate the condition.

Corticosteroid dermatitis is a completely reversible condition, although the withdrawal process may be difficult. Gradual reduction in corticosteroid potency over six weeks is usually effective but the addition of oral doxycycline at doses of 50–100 mg/day will improve any symptomatic exacerbations during the process.

Management

By the time that a patient presents with vulval dermatitis, she has often developed one or more secondary problems. The most common of these is dyspareunia, the management of which is discussed in Chapter 6. The first step in management is the control of the dermatitis before dealing with secondary issues.

The principles of treating vulval dermatitis include:

- environmental modification, which includes eradication of irritant substances
- diagnosis and treatment of super-infection
- identification of possible allergens
- topical corticosteroids
- management of secondary dyspareunia.

Environmental Modification

Topical corticosteroids alone will not substantially improve any form of genital dermatitis. This is because genital skin is exposed to many contact irritants and possibly to allergens. Even in endogenous dermatitis, these local factors exacerbate the clinical problem.

Treatment must start with identifying and managing the causes of an exogenous dermatitis, and the exacerbating factors in an endogenous dermatitis.

Most patients have more than one causative or exacerbating factor. The patient needs to understand that significant improvement will not be possible until these factors are controlled.

If you suspect that one of the patient's topical medications or cosmetic products is responsible, stop that product.

Common irritants include:

- soap
- bubble bath
- feminine hygiene products
- sanitary pads and liners
- prolonged use of anti-fungal creams
- lubricants
- wet wipes
- douches
- waxing and shaving products
- nylon underwear
- pantyhose
- G-strings
- tight clothes
- gym clothes
- friction from bicycle seats.

Patients should be advised to:

- use a soap substitute
- wear cotton underwear
- use tampons, menstrual underwear or menstrual cups, rather than pads
- substitute a wet cloth for wet wipes (this can be kept in a snap-lock bag)
- avoid routine use of panty liners
- discard G-strings
- stop using anti-fungal creams
- discard perfumed products
- wear loose clothes
- avoid nylon bike pants and pantyhose
- avoid activities that cause heavy sweating and/or friction until improved.

Dermatitis always involves an element of dryness, so a bland emollient is a necessary part of management and prevention. In general, this should be non-perfumed, non-irritating and of a consistency that is comfortable and acceptable for the patient. We recommend daily use as often as possible. On inflamed skin, many products cause stinging. This is less likely to happen with ointments than creams.

Diagnosis and Treatment of Super-infection

All patients should have a low vaginal swab and a vulval swab. Occasionally, bacterial super-infection can occur in vulval dermatitis. This may present as a cellulitis, producing oedema; however, weeping and crusting may also occur. The organisms that may cause super-infection are:

Candida albicans (vagina)

Staphylococcus aureus (vulva)

Streptococcus pyogenes (vulva).

A finding of *C. albicans* may indicate super-infection in dermatitis, or possibly the independent diagnosis of chronic vulvo-vaginal candidiasis. If the history is typical of chronic candidiasis, the patient will need prolonged anti-fungal treatment. A rapid response to a single oral dose of fluconazole 150 mg is the most pragmatic way to determine whether *C. albicans* is partially responsible for the patient's symptoms. We recommend oral azoles because, in patients with dermatitis, topical anti-fungals run the risk of causing irritation.

Bacterial infection is treated with appropriate oral antibiotics, according to antibiotic sensitivities.

Group B Streptococcus vaginal isolates are usually irrelevant in these situations, and can be ignored.

Identification of Possible Allergens

If allergic contact dermatitis is suspected, the initial examination should include taking a careful history. While in the office, patients often do not remember every substance that has been applied to their skin. Ask them to go home and write down everything that they can think of that is applied to the vulva.

You will note that a number of products are on both the irritant and allergen list. It is common for many substances to irritate but rarer for them to cause a true allergy.

Latex allergy may be diagnosed with specific IgE serology.

Use of Topical Corticosteroids

When using topical corticosteroid on the vulva, ointments are preferable to creams. Creams contain preservatives that may cause stinging, irritation or even frank allergy. Ointments tend to adhere better to the moist skin of the inner vulva.

In general, a weak topical corticosteroid (1% hydrocortisone) is effective and appropriate in treating all forms of genital dermatitis, provided environmental modification is in place. An application twice daily for up to one month is often all that is necessary to control the inflammation. It may then be used as needed for recurrences. The advantage of this weak cortisone is that it can be used for long periods of time without adverse effects. This is essential as many forms of dermatitis are chronic conditions that require long-term control.

If lichenification is present, a more potent topical corticosteroid ointment may be safely used once daily until this has resolved. This may take as long as a month. As soon as the patient has recovered, the topical corticosteroid should be reduced gradually, first to medium potency for a month and then to 1% hydrocortisone.

Intralesional intradermal triamcinolone may sometimes be needed for patients with recalcitrant areas of lichen simplex chronicus.

These warnings about potent topical corticosteroids are not intended to discourage their use. Even with long-term treatment, the weak preparations are safe and appropriate on the vulva. 'Corticosteroid phobia' is common in the community and patients frequently receive warnings from health professionals about their potential to 'thin the skin'. Such warnings only serve to discourage treatment adherence and deny patients with a genuine problem the treatment that they need. In practice, adverse effects of cortisone are seen only with

prolonged inappropriate use, and even so, are reversible. As a doctor, it is important to be very positive about the use of these products.

Alternatives to Topical Corticosteroids

There are very few alternatives to the use of topical corticosteroids when treating dermatitis, particularly where it is severe.

The topical calcineurin inhibitors pimecrolimus 1% and tacrolimus 0.03% and 0.1% and the PD4 inhibitor crisaborole 2% may be used to treat vulval dermatitis, however, these are significantly more expensive than topical corticosteroids and frequently cause stinging.

It is usually necessary to gain control of the dermatitis first with a topical corticosteroid before initiating them. Because of corticosteroid phobia, long-term maintenance with other anti-inflammatory products may have better acceptance by some patients.

Management of Dyspareunia

All forms of vulval dermatitis have a tendency to cause splitting, and scratching may cause excoriation. As a result, intercourse may cause discomfort.

In many cases, simple treatment of the underlying condition is all that is required for dyspareunia to resolve. However, in any painful vulval condition, secondary pelvic floor muscle spasm may continue after adequate control of the dermatitis.

If dyspareunia does not resolve after symptoms have been controlled and the vulva has returned to normal objectively, pelvic floor muscle dysfunction should be suspected and treated (see Chapter 6).

Psoriasis

Psoriasis is a common vulval condition, but less so than dermatitis, with which it is most often confused.

History

History often provides clues that differentiate psoriasis and dermatitis. Psoriasis is often episodic in nature, with episodes not always clearly related to any triggers. Psoriasis is classically exacerbated by situational stress or physical illness, particularly streptococcal throat infections. It may also be precipitated by vaginal candidiasis, or vaginal surgery. In some cases, psoriasis flares pre-menstrually.

A common history is of a very longstanding itchy vulvitis, present for years and treated intermittently by potent topical corticosteroid with some response but without lasting improvement.

Vulval psoriasis is often itchy. It may become sore if eroded by scratching. It may flare pre-menstrually but this is not invariable.

Examination

Psoriasis of the vulva is easier to diagnose if present elsewhere on the skin, but it can occur solely on genital skin. On the pubic area, it is usually typical with well-defined scaly red plaques, but on the vulva itself, it lacks the scale and sometimes the sharp edge of typical psoriasis. When scale occurs in vulval psoriasis, it is usually macerated and found in the sulcus between the labia minora and majora.

The lesions are usually more erythematous and well defined than dermatitis, and are usually bilaterally symmetrical (Figure 3.4), although unilateral lesions are possible. The peri-anal area and natal cleft may be involved (Figure 3.5). Psoriasis does not involve the vagina, but the labia are usually involved and the rash can extend inwards as far as the vestibule and externally into the inguinal folds and inner thighs (Figure 3.6). Natal cleft involvement is a useful sign, as it is not seen in dermatitis. Lichenification of the peri-anal skin and labia majora may be severe.

A search for subtle signs of psoriasis such as nail pitting, scalp scaling and thickening and scaling of the dorsal surface of the elbows and knees may provide helpful diagnostic clues, as does a family history of psoriasis.

There is no specific diagnostic test for psoriasis. Although a classic histopathology is described, this is often lacking on the vulva and biopsies are often reported as 'non-specific

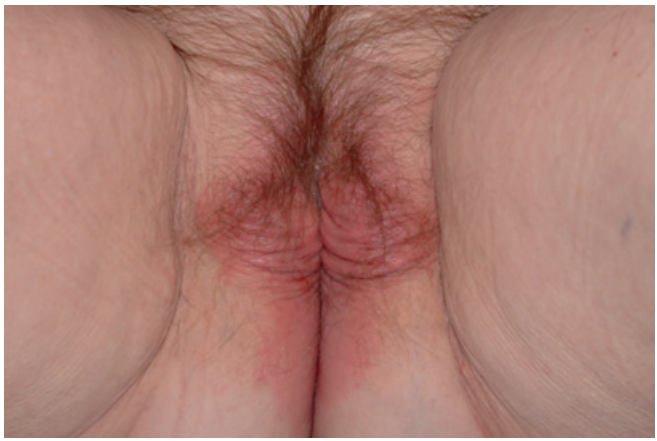

Figure 3.4 Psoriasis. Note the well-demarcated border and symmetry

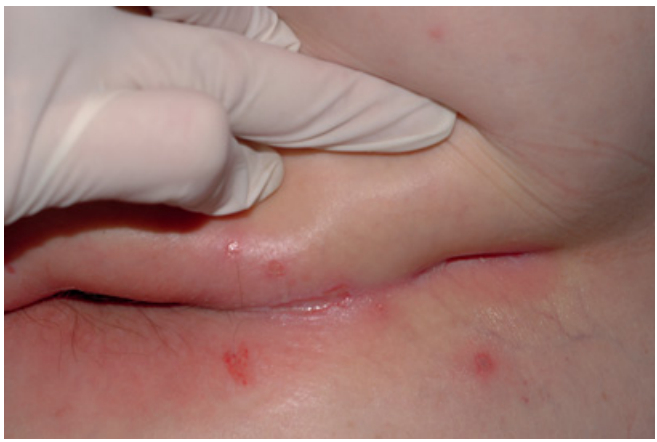

Figure 3.5 Psoriasis in the natal cleft with fissuring

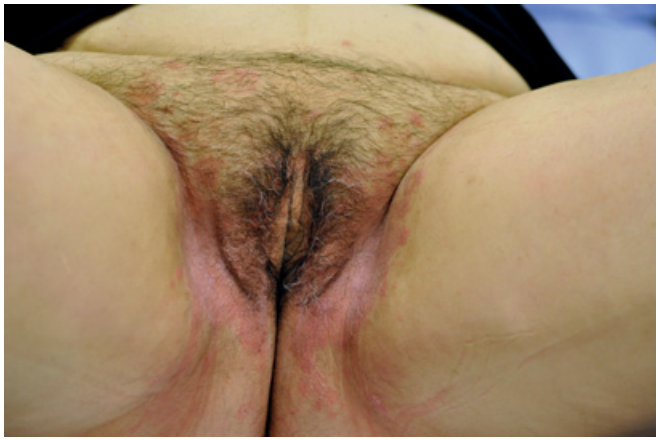

Figure 3.6 Psoriasis, extending out to groins. Note the typical silvery appearance

inflammation' or 'spongiotic dermatitis'. Therefore, diagnosis relies on the clinician's judgement and is supported by family history and/or signs of psoriasis elsewhere on the skin.

Management

Psoriasis usually improves initially with topical corticosteroids, but these can lose effectiveness after some weeks. Treatment with non-corticosteroid topical therapy specific for psoriasis is necessary to continue improvement and maintain control, and the patient should be given a management plan for the inevitable flare-ups.

A diagnosis of psoriasis can be distressing for many patients. The condition is common enough for many people to have heard of it and also to realise that it is not curable, and in a few cases very severe. It is important to put this into perspective. Psoriasis is common, in most cases not severe, and on the vulva, nearly always able to be controlled with topical therapy. If this is not possible, there are many systemic treatments that are effective.

The same principles of environmental modification and infection control that are used for patients with dermatitis should be put into place.

Weak topical corticosteroids are usually ineffective for initial treatment of psoriasis. A topical medium-potency cortisone ointment is used at night until significant symptomatic and objective improvement is achieved. The potent topical corticosteroid betamethasone dipropionate 0.05% is available combined with calcipotriol (vitamin D) as an ointment or foam. This is a specific treatment for psoriasis and can also be effective on the vulva. This often takes at least four weeks. Psoriasis can, like dermatitis, be complicated by lichenification. In this situation, a potent topical corticosteroid is used to initiate treatment, as described earlier.

As soon as the patient is comfortable and eroded or split areas have healed, introduce a non-corticosteroid preparation:

- 2% LPC (liquor picis carbonis – a tar product) in aqueous cream or white soft paraffin. This should be tested on the cubital fossa for a few days before applying to the vulva to ensure that it does not cause a severe irritant or allergic reaction.
- calcipotriol ointment.

If neither of these preparations are tolerated, a greasy emollient should be used. Bepanthen® ointment, Amolin® cream or zinc and castor oil give many patients relief.

There are a number of other maintenance formulations that can be helpful. Topical calcineurin inhibitors such as pimecrolimus 1% or tacrolimus 0.1% or 0.03% may be tried but may cause unacceptable stinging, as in dermatitis. It is important to remember that many psoriasis patients have unpredictable reactions to various preparations. Prolonged burning and stinging after using one treatment mean that it should be discontinued, and another tried.

It usually takes several weeks for optimal efficacy to be achieved. Once adequate improvement is achieved, the steroid ointment is used intermittently for flares and the LPC continued daily as a preventer. Some patients benefit from using both daily. Long-term management is required on the vulva, and it is important to make sure patients realise this.

If there is co-existent candidiasis, prolonged anti-fungal treatment is essential until the psoriasis is well controlled. This is because many cases of psoriasis are driven by chronic candidiasis.

Psoriasis can be an unpredictable and frustrating disease. It is a matter of trial and error to find the ideal maintenance and flare-up regimen. Genital psoriasis not able to be controlled by topical therapy should be referred to a dermatologist.

Tinea

Although common elsewhere, tinea is an uncommon cause of vulval disease. Tinea is a dermatophyte fungus that also causes tinea pedis.

Presentation

Patients present with an itchy, scaly bilateral or unilateral rash involving the labia majora, which may extend to both inguinal folds and lower abdomen (Figure 3.7). The edge is usually better demarcated than in dermatitis and it is often less symmetrical. It may be difficult to differentiate clinically from psoriasis or dermatitis, but is worsened by topical corticosteroid treatment, even if there is temporary improvement at first.

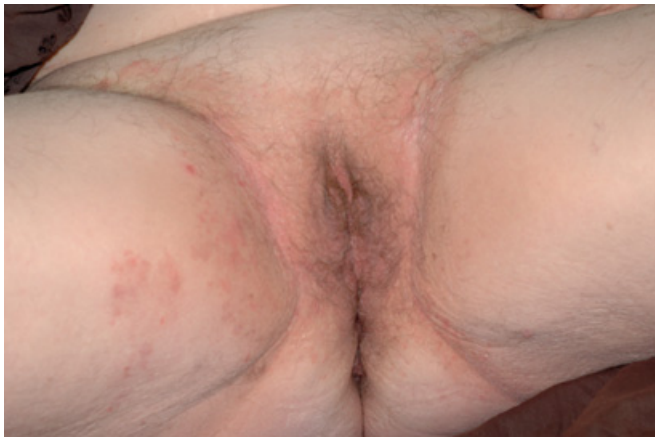

Figure 3.7 Tinea. Note the asymmetry and extension onto the lower abdomen

Rarely, vulval tinea may present with pustules and nodules and may closely simulate boils or hidradenitis suppurativa.

Investigation

A fungal skin scraping or biopsy is essential to make a diagnosis.

Management

Most cases of vulval tinea require oral anti-fungal treatment for two reasons:

- the hair follicles are involved
- there is almost invariably foot involvement with the same fungus and unless this is adequately treated, the patient runs the risk of re-inoculating herself.

The following anti-fungal medications may be used for tinea:

terbinafine 250 mg/day

fluconazole 50 mg/day

itraconazole 100 mg/day.

Each needs to be given until there is complete clearance of the rash on the vulva and the feet (if present), and a repeat scraping is negative. The minimum duration of treatment is typically four to six weeks; however, longer treatment periods may be required.

For patients who are unwilling to take oral medication, topical terbinafine will control symptoms. It should be used daily for several weeks. Relapse is common when it is stopped.

Chronic Vulvo-vaginal Candidiasis

Case Study

A 36-year-old woman is referred with a two-year history of urinary frequency, dysuria and minor stress urinary incontinence. She has previously been investigated by a urologist, but urine culture, urodynamic studies and cystoscopy were all normal.

Questioning reveals a three-year history of chronic sinusitis, managed with multiple courses or oral antibiotics, leading to monthly episodes of (confirmed) vulvo-vaginal candidiasis, usually occurring in the pre-menstrual phase. Recently, her candidiasis symptoms occur daily, with pre-menstrual exacerbations. She admits to episodes of acute candidiasis in her twenties.

Examination reveals a poorly demarcated, swollen erythematous rash involving the labia minora and vaginal vestibule.

This woman has chronic vulvo-vaginal candidiasis that has produced bladder dysfunction. She has the typical history of a susceptibility to candidiasis, with a trigger (antibiotics) that has caused the chronic form of this disease.

Oral fluconazole at a daily dose of 50 mg is commenced, and 1% hydrocortisone ointment is used twice daily for any vulval discomfort. On review at three months, the bladder is now functioning normally, and the examination is normal. The dose of fluconazole is then slowly reduced to a maintenance dose of twice weekly. This woman needs appropriate advice to better manage her chronic sinusitis.

Approximately 20% of women carry *Candida* yeast (the majority *C. albicans*, and about 5% *C. glabrata*) in the vagina, but only a small number suffer from recurrent or chronic candidiasis.

Vulvo-vaginal candidiasis is oestrogen-dependent and in healthy women. The condition starts from menarche onwards. It ceases after menopause unless oestrogen-replacement therapy (ERT) is used.

Acute and recurrent candidiasis are well understood, easily recognised and treated. Recurrent vulvo-vaginal candidiasis is defined as four or more attacks of microbiologically proven vulvo-vaginal candidiasis per year, with the patient being asymptomatic in between. However, patients with the chronic form of vulvo-vaginal candidiasis are a much less well recognised and managed group.

We define chronic candidiasis as patients who are constantly symptomatic. It is a real diagnosis, but unfortunately not recognised in much of the literature on the subject.

Presentation

Chronic vulvo-vaginal candidiasis usually presents with recurrent or constant itch, but dyspareunia, soreness, burning, swelling, fissuring and pain are common. It is characteristic but not invariable for these symptoms to cycle, being most severe in the pre-menstrual week, and suddenly improving on the first day of menstruation.

Discharge is common but may be absent, and the typical cheesy exudate of acute candidiasis is not seen. It is typical for courses of oral antibiotics to exacerbate or precipitate symptoms.

The patient's partner may sometimes experience post-coital itching and penile rash, which is also characteristic. Patients often give a history of response to topical and oral anti-fungals, however, recurrence of symptoms is the rule when they are ceased. There may also be a history of worsening symptoms with the use of potent topical steroids. As most patients do benefit to some extent from anti-fungal medications, they use them frequently and as a result vaginal cultures may return a false negative. Therefore, a negative swab does not rule out this condition, particularly if other aspects of the history are present.

Many patients can recall recurrent attacks of acute candidiasis from adolescence onwards. They state that initially such attacks responded to topical anti-fungal medication but, with time, resistance to topical therapy occurred, attacks became more frequent and eventually symptoms became constant.

The rash in this condition may vary from no apparent abnormality to severe erythema, involving the whole vulva and peri-anal area and vagina, with varying degrees of oedema and fissuring on the perineum and inter-labial sulcus (Figures 3.8 and 3.9). The most typical presentation, however, is a vulvo-vaginitis with erythema of the labia minora extending into the sulcus between the minora and majora.

Chronic vulvo-vaginal candidiasis can be a very difficult diagnosis. If your patient has:

- a persistent non-erosive vulvo-vaginitis that cycles, being most severe in the pre-menstrual week
- a vulvo-vaginitis that has previously responded (even if briefly) to topical or oral anti-fungals
- had positive swabs for *Candida*, even if this has been inconsistent and not positive on THIS occasion
- a history of exacerbation as a result of taking antibiotics.

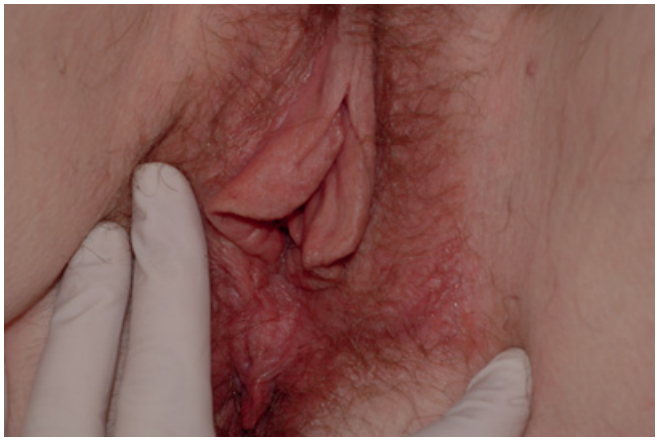

Figure 3.8 Chronic candidiasis. Note the oedematous rash extending out to the hair-bearing surface of the labia majora. The external border is poorly demarcated

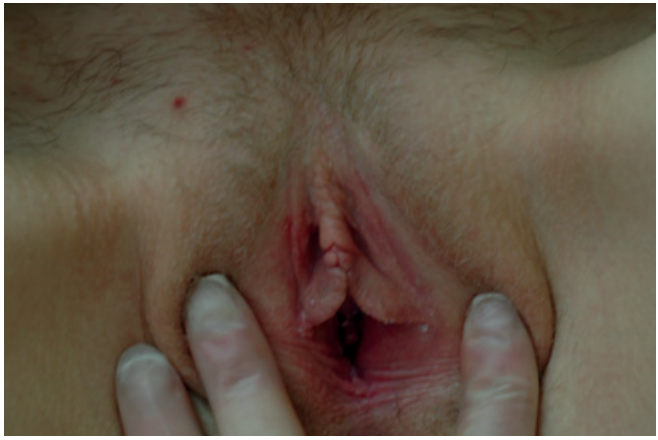

Figure 3.9 Chronic candidiasis. Note the associated rash with fissuring

Then a trial of prolonged anti-fungal therapy is indicated. Such a trial often becomes the ultimate diagnostic test. It is most easily done using oral medications as pessaries can be irritating and messy. However, if the isolate is *C. glabrata*, there is no practical choice.

Investigation

When taking the swab, the yield is best from a low, rather than high, vaginal swab. If the typical history is present, any degree of culture-positivity is significant. Swabs are unreliable as so many patients self-medicate with over-the-counter anti-fungals. Biopsy is usually non-specific, and can be misleading as it rarely shows hyphae. The diagnosis is therefore made on history.

Although a number of factors are associated with an increased tendency to candidiasis (ERT, diabetes, local or systemic corticosteroid treatment, antibiotic therapy and immunosuppression), the majority of women with chronic vulvo-vaginal candidiasis are otherwise normal. Patients often relate that their female siblings and mother also had a susceptibility to candidiasis. This condition may therefore have a genetic basis. It is exceptional to see it in Asian women.

The first symptoms are often seen around the time a patient first becomes sexually active. The reason for this is unknown. Attempts to relate vulvo-vaginal candidiasis to various personal and hygiene habits have been inconsistent.

The pathogenesis of chronic vulvo-vaginal candidiasis is still unknown but it behaves as though the commensal organism *Candida* is an antigen that causes a maladaptive inflammatory response rather than an infection. If chronic vulvo-vaginal candidiasis is conceptualised in this way, the condition is much more easily understood. The exact mechanism is, however, still unknown.

The important role of oestrogen in candidiasis may also explain why patients with a severe problem in the vagina rarely complain of oral symptoms. The vulvo-vaginal epithelium contains different oestrogen receptors to those in the oral mucosa, and although the presence of oestrogen is essential to this condition, its exact role has not been elucidated. It is known that *Candida* possess an oestrogen receptor but the pathogenesis of how this relates to symptoms is unknown as is the role of the vaginal microbiome. Research into this during the next few years may explain some of the mysteries of why a commensal organism that is tolerated by most women causes severe symptoms in some.

Almost 95% of cases are caused by *C. albicans* but other yeasts may be responsible, most often *C. glabrata*. These fungi may be reported as 'non-pathogenic' by some laboratories, and a request for further characterisation should be made if necessary.

Management

If the history is typical, a trial of anti-fungal treatment should be given. This must be with a daily oral anti-fungal for at least one month. There are two appropriate oral anti-fungals available: fluconazole and itraconazole. Fluconazole is used at a dose of 50–100 mg/day and itraconazole at a dose of 100 mg/day. Ketoconazole is not used because of risks of hepatic damage over the long treatment periods required.

Both itraconazole and fluconazole have a good safety record and are not usually associated with abnormal liver function tests or frequent side effects. Routine monitoring of liver function tests is usually normal.

Side effects occur rarely and include nausea, diarrhoea, allergic reactions, pustular facial rash, sensory neuropathy, peripheral oedema and, with fluconazole, reversible hair loss. It is very unusual for resistance to be encountered, although it has been described.

Both anti-fungals may potentially interact with many other drugs, including fexofenadine hydrochloride, an antihistamine available over the counter, and most statins. The interaction with statins is particularly serious and may result in rhabdomyolysis.

Treatment must be prolonged, and the length of time will vary from patient to patient. A useful rule of thumb is to continue daily treatment until the patient is totally asymptomatic. Physical examination is unreliable and persistent erythema of the inter-labial sulcus is common, even when patients have recovered symptomatically. Typically, this takes three

months but some patients take much longer to recover, some as long six to twelve months. At this point, the dose is gradually reduced to the lowest level that will control symptoms over the next six months. Again, this varies from patient to patient and a few are not able to reduce their medication at all without recurrence of symptoms. We find that maintenance doses vary from 100 mg/day in some patients to the occasional 50 mg dose for return of symptoms. The majority do well on 50 mg of fluconazole twice a week. The half-lives of these drugs are such that maintenance intermittent dosing is often effective, although we have found this to be less so during the initial induction phase.

Published reports of the use of oral azole anti-fungals often recommend 150 mg/week. Our personal experience has been that this is not as effective as daily dosing in the induction phase. There have, however, been no side-to-side trials to establish an evidence-based treatment regimen.

Once patients are on a low maintenance dose, they need to be made aware that a course of antibiotics may result in a relapse and that they should take their medication daily for a week during and after any such course. Any flares of their condition are again treated with an increased dose until symptoms have settled.

Some patients do appear to require higher rather than lower doses over time and this may be due to a slow increase in the minimum inhibitory concentration (MIC) of the drug, or individual variation in metabolism and absorption. If this happens, ask about other medications that your patient is taking (especially herbal preparations) as they may interfere with absorption. Patients with any disease that causes malabsorption may not absorb these medications well.

Once a remission of at least six months has been achieved, patients may attempt to stop their treatment; however, many relapse and should be given permission to restart immediately and to titrate their dose to a level that keeps them symptom free. The situation is no different from the use of long-term anti-viral medication in the management of recurrent genital herpes.

In patients where an atypical *Candida* such as *C. glabrata* is isolated, there are two treatment options: boric acid vaginal suppositories (dose 600 mg) or the oral azole voriconazole. The latter has significant toxicities not shared by fluconazole and itraconazole. The same principles of management apply with daily treatment until symptomatic remission is achieved and then regular long-term maintenance therapy.

It is important to reduce environmental or pharmaceutical influences that may make treatment of candidiasis less effective and increase the recurrence rate. Reducing heat, sweat and friction, improving any medical condition that calls for long-term use of antibiotics (asthma, acne, recurrent urinary tract infection and recurrent tonsillitis) and managing bowel disorders so that faecal soiling is less of a problem are just some of the wider medical issues that should be considered.

The role of oestrogen in this condition means that a symptomatic vulvo-vaginal problem in pre-menarchal girls and healthy post-menopausal women who are not on ERT is very unlikely to be candidiasis. During pregnancy, symptoms frequently and counter-intuitively improve in most suffers. Although the use of high oestrogen oral contraceptive pills may be associated with candidiasis, cessation of the pill does not usually improve it. Low oestrogen oral contraceptive pills are rarely implicated.

At menopause this condition remits, but it may take up to a year after the last menstrual period for all symptoms to cease. In non-diabetic healthy post-menopausal women, vulvo-vaginal candidiasis is usually associated with systemic or vaginal oestrogen. Chronic

candidiasis in this group is usually controllable within one month of oral azole treatment if all oestrogen is stopped during this time.

In patients who are unable to cease oestrogen supplements, a weekly dose of oral anti-fungal medication usually keeps symptoms under control once symptoms are controlled.

It must be stressed that the finding of atypical *Candida* does not always imply clinical disease in the vagina. In an asymptomatic patient, it can be regarded as a carrier state.

Oestrogen-Replacement Therapy (ERT)-Associated Vulvo-vaginal Candidiasis

It is important to remember that although healthy post-menopausal women do not suffer from vaginal candidiasis, the situation can change significantly if they begin ERT. However, there may be a lag of up to five years of treatment before this occurs.

Patients on ERT may develop candidiasis for the first time but more often they are patients who have had relatively frequent attacks of acute candidiasis pre-menopause.

Patients who have ERT-related candidiasis require cessation of ERT and treatment with oral anti-fungal agents until symptoms have resolved (usually four to six weeks of daily treatment).

Once a remission of signs and symptoms has been achieved, permanent cessation of ERT is the ideal advice. If ERT is essential, it should be at the lowest possible dose. Maintenance doses of anti-fungals are often required to prevent relapse. Our usual recommendation is fluconazole 50 mg twice a week.

Diabetes and Vulvo-vaginal Candidiasis

Diabetes is known to be a risk factor for vulvo-vaginal candidiasis. Candidiasis may occur in post-menopausal women with diabetes who are not on oestrogen replacement.

The hypoglycaemic agents of the group of SBLT2 inhibitors are associated with an increased risk of candidiasis in type 2 diabetes, via an increase in glycosuria. Although this can be managed with anti-fungal agents, there are times when the drug has to be ceased.

Oestrogen-Hypersensitivity Vulvitis

Oestrogen-hypersensitivity vulvitis is a very rare condition that produces a strikingly cyclical vulvitis, mimicking chronic candidiasis. Patients suffer from vulvo-vaginitis throughout the menstrual cycle, with a pre-menstrual exacerbation.

Both endogenous oestrogen and progesterone are recognised as rare causes of cyclical rashes on extra-genital skin. Some women who have cyclical vulval symptoms suffer from this sort of hypersensitivity. The vulval skin and vagina contain oestrogen but not progesterone receptors, and for this reason, the culprit in the vagina is usually oestrogen.

History Taking

Patients often give a history that is very similar to chronic vulvo-vaginal candidiasis; however, they have had consistently negative swabs, and no response to anti-fungal therapy. They tend to be refractory to topical steroid treatment.

Investigation

There is no commercially available diagnostic test and diagnosis relies on good history taking and a trial of oral anti-fungal therapy to rule out chronic candidiasis. In a research setting, patients do have positive intradermal tests to oestrogen.

Management

Many patients with this condition are intolerant of the combined oral contraceptive pill. Menstrual suppression with progestogens such as cyproterone acetate has been our most successful therapy to date. Successful treatment with tamoxifen has been described, however, it does have potential side effects. Raloxifene may prove to be a useful agent.

If this rare condition is suspected, the patient should be referred to a specialist with an interest in vulval disease. The use of hormones to suppress the menstrual cycle is best supervised by a gynaecologist.

Extra-mammary Paget's Disease

Extra-mammary Paget's disease is a very rare condition, but an important one, as it is easily mistaken for dermatitis or psoriasis.

Presentation

The presentation is with an eczematous erythematous bilateral or unilateral eruption, which can occur on the vulva, peri-anal area or both. The erythematous base may have a white, flaky surface that has been likened to icing sugar. With time, the area becomes raw and weeps. The eruption is usually itchy and sometimes sore. There is no response to topical corticosteroid treatment. The typical patient is post-menopausal.

Investigation

Biopsy is essential for diagnosis. The literature states that up to 20% of patients have an underlying adenocarcinoma, and another 30% have an adenocarcinoma at another location.

Management

Patients with this condition should be referred to a gynaecological oncologist.

Common Pitfalls in Therapy of Red Scaly Rashes on the Vulva

Prolonged Use of Vaginal Azoles

Intra-vaginal azoles are effective treatments for acute candidiasis, but are usually not convenient or effective for longer-term use in chronic candidiasis. Prolonged or recurrent use may cause irritancy. If one course of a standard vaginal anti-fungal does not bring lasting symptomatic relief, further courses of similar drugs should not be used, and the original diagnosis reviewed. In confirmed cases of chronic candidiasis where long-term vaginal therapy is the only option, we recommend vaginal nystatin, 100,000 U/5 g dose.

Vulval Application of Topical Anti-fungals

Candidiasis is usually an infection of the vaginal mucosa only, and the external vulval symptoms are caused by hypersensitivity to this infection. External symptoms therefore should be controlled with a low-potency steroid ointment. Applying topical anti-fungals to vulval skin is not only ineffective but may worsen this irritant dermatitis.

Vaginal Cultures

It is essential to understand that a vaginal culture cannot reliably exclude candidiasis. The use of a vaginal speculum to collect the swab is unnecessary. Patients can do their own vaginal swab when they are symptomatic. This eliminates the problem of urgent appointments simply for specimen collection. Nevertheless, the history is more important than the swab in making the diagnosis of chronic vulvo-vaginal candidiasis. A history of positive swabs in the past is important.

The Use of Topical Oestrogen

The use of topical oestrogen for 'dryness' of the vulva is useless in pre-menopausal women with regular periods. These women are already very adequately oestrogenised, and their sensation of dryness usually relates to some form of dermatitis, as the latter usually produces a dry, flaky skin surface.

Prolonged Use of Potent Topical Corticosteroids

There are very few situations where long-term use of potent topical corticosteroids is required to control symptoms of any vulval dermatosis other than lichen sclerosus and lichen planus (see Chapter 4).

Although they are appropriate and useful to bring initial symptoms under control, if you find that your patient is unable to reduce to a weaker preparation, it is time to review the diagnosis, question compliance to environmental modification, rule out infection, allergy or consider corticosteroid dermatitis. Any unilateral or very recalcitrant rash should be biopsied to rule out extra-mammary Paget's disease.

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Chapter

4

Things That Look White[1]

Contents

White- or pale-appearing patches on the vulva are uncommon. Most white vulval lesions are lichen sclerosus. However, vulval intra-epithelial neoplasia (VIN) may also appear white, and dermatitis complicated by lichenification or lichen simplex chronicus may also appear white.

In the past, white patches on the vulva were called 'leukoplakia'. This term rarely appears anymore and should be regarded as out of date. It should be abandoned in favour of specific diagnostic terms. Because of the potential diagnostic confusion, white lesions on the vulva in adults should be biopsied if possible.

Lichen Sclerosus

Case Study

A 62-year-old woman presents with a four-year history of dry vaginal dyspareunia and vulval itch, for which vaginal oestrogen treatment has been ineffective. Examination reveals a thick, white rash around both the vulval and peri-anal skin. A biopsy confirms the clinical suspicion of lichen sclerosus. Betamethasone dipropionate 0.05% in optimised vehicle is applied every night, and after six weeks, the itch is eliminated and the dyspareunia is improved. Examination reveals complete visual suppression of the lichen sclerosus. The corticosteroid regimen is then reduced to methylprednisolone aceponate ointment five nights/week, with betamethasone dipropionate ointment two nights/week.

[1] We would like to thank Dr Greg Gard, FRANZCOG, CGO, for his help in preparing this chapter.

On review at three months, the dyspareunia is minimal but feels 'dry'. Examination reveals that the lichen sclerosus is still completely visually suppressed, but that there is menopausal atrophy. Vaginal oestrogen is added, which eventually eliminates the residual dyspareunia.

Lichen sclerosus is a skin disease that has a predilection for the genital skin and has a female-to-male ratio of 10:1. It is relatively prevalent after the age of 50, being found in 3% of females. This makes it relatively common in vulval practice, responsible for about 10% of cases.

Although lichen sclerosus may occur on any part of the skin, it is almost always a genital condition. While we will be referring to 'vulval lichen sclerosus' in this chapter, it is essential to remember that perineal and peri-anal involvement is common as well.

While lichen sclerosus occurs in all age groups, it is most common in peri-menopausal and post-menopausal women and the mean age of onset is around 55. It can occur in children and in babies. Until recently, it was believed that pre-pubertal lichen sclerosus resolved at puberty but recent evidence contradicts this.

Lichen sclerosus is an important condition to diagnose correctly for two reasons. First, if not treated aggressively, it may significantly scar, shrink and deform the vulva and cause stenosis of the introitus. Second, lichen sclerosus carries with it a 2–6% lifetime risk of squamous cell malignancy, both intra-epithelial (VIN) and invasive (squamous cell carcinoma (SCC)). Both of these complications can be either prevented or greatly reduced with adequate treatment. Therefore, these patients require lifelong observation.

Epidemiology

Vulval lichen sclerosus has a reported prevalence of between 1 in 300 to 1 in 1,000 women and 1 in 900 girls. These figures may well be an underestimate because many cases go unrecognised.

Lichen sclerosus occurs predominantly in peri- and post-menopausal women, although about one-third may occur in women under 50 years. Paediatric lichen sclerosus accounts for 5–15% of all cases.

Extra-genital lesions can be found on any part of the skin but are most common on the neck, buttocks, inner thigh, shoulders and wrists. Extra-genital lichen sclerosus can also occur as multiple small 'confetti' lesions.

In children, lichen sclerosus almost always affects the genital area, with only approximately 6% of these patients having extra-genital involvement.

Aetiology

The true aetiology of lichen sclerosus remains unknown; however, there is a well-documented association with autoimmune disease, particularly Hashimoto's thyroiditis and vitiligo.

There seems to be an association between lichen sclerosus and the presence of serum auto-antibodies such as anti-nuclear factor and thyroid auto-antibodies, but this association is not aetiological. It is still uncertain if lichen sclerosus is an autoimmune disease.

Should We Test for Associated Diseases?

When patients are diagnosed with lichen sclerosus and start to investigate it on the Internet, they invariably discover frightening accounts of associated diseases. They worry that their immune systems are compromised. This has to be put into perspective.

A recent well-conducted study found that relative to aged-matched controls, lichen sclerosus patients do suffer from clinical autoimmune disease more often – about 30% as opposed to 10% of the whole population. About 30% also had a positive family history. However, the same study found that there was no significant difference between the rate of asymptomatic auto-antibody detection between patients with lichen sclerosus and controls.

The majority of patients we encounter with lichen sclerosus are otherwise well with no personal or family history of autoimmune disease. Thyroid auto-antibodies may also be present, and if this is the case, further investigation is warranted.

The question remains as to whether it is best practice to investigate all patients with lichen sclerosus for evidence of autoimmune tendencies. Most patients who have associated autoimmune disease are already diagnosed when we encounter them but, occasionally, we do identify patients with thyroid auto-antibodies who were unaware that there was a problem.

We recommend only TSH and thyroid auto-antibodies. Other non-directed testing is likely to have a very low yield.

Genetic Factors

Lichen sclerosus has been reported to run in families and there have been attempts to find a genetic association. Although no association to the autoimmune-related HLA antigens (HLA A1, B8 and DR3) has been reported, the HLA class II antigen HLA DQ7 has the strongest association with lichen sclerosus.

While these documented HLA associations are of interest, unfortunately, there is a lack of significant data to conclusively comment on the strength of these associations.

Presentation

The most common presenting symptom is itch, often of a severe, life and sleep-disrupting nature. There is sometimes pain as a result of excoriation or fissuring. Distressing clitoral hyperaesthesia may occur, and dyspareunia is very common. This has a marked effect on quality of life. However, occasionally, lichen sclerosus can be completely asymptomatic, discovered by chance by the patient or by the general practitioner during a Pap test.

The appearance of a well-defined white sclerotic plaque with an atrophic wrinkled surface and areas of purpura and erosion is typical. However, there are many variations (Figure 4.1). These include:

- multiple white papules or macules
- hyperkeratotic lesions
- plaques limited to small areas such as the tips of the labia minora or the clitoris or clitoral hood
- oedema on a background of pallor
- telangiectasia, purpura, haemorrhagic blistering on a background of pallor
- fissures, traumatic ulcers

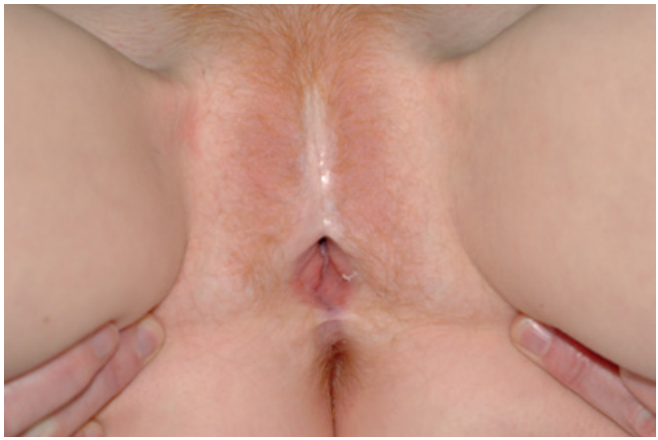

Figure 4.1 Lichen sclerosus with severe anterior labial fusion

- erosions
- lichen sclerosus associated with vulval psoriasis, which appears erythematous
- brown hyper-pigmentation similar to melanosis vulvae that can supervene.

The distribution of lichen sclerosus is also very variable. The classic textbook description is of a figure of eight encircling the vulva, perineum and peri-anal skin. However, it can affect only one area of the genital skin. Lichen sclerosus does not involve the vagina proper (i.e., within the hymen).

Lichen sclerosus obeys what is called the Koebner phenomenon, which means it localises into areas of friction and trauma. This possibly explains why it is usually most recalcitrant on the perineum and the inner surfaces of the labia minora.

A key point in recognising lichen sclerosus, particularly in the late stage, is that the vulval shape is not normal. If left untreated, the labia minora eventually become reabsorbed and the clitoris becomes entrapped under clitoral hood scarring, revealing an overall atrophic, shiny, white vulva missing normal anatomy. It is very typical for the labia minora to fuse, most commonly posteriorly, but also anteriorly (see Figure 4.2). The fusion line is brittle and easily tears during intercourse. Perineal fissuring and tearing are also common. Eventually, the vaginal opening (introitus) may become significantly stenosed, with pooling of urine within the vagina, simulating urinary incontinence.

With end-stage disease, epithelial change may be hard to find, and all that is left is gross distortion of the vulva.

Clinical Presentation in Children

A recent study of 70 children with lichen sclerosus showed the mean age of development of symptoms was 5.0 years (range 1–12 years) and the mean age at diagnosis was 6.7 years (range 3–14 years). Another study of 46 children found the mean age of diagnosis to be 7.8 years with a delay in diagnosis of 1.6 years. Both studies indicate that many children suffer for long periods of time before being diagnosed and treated.

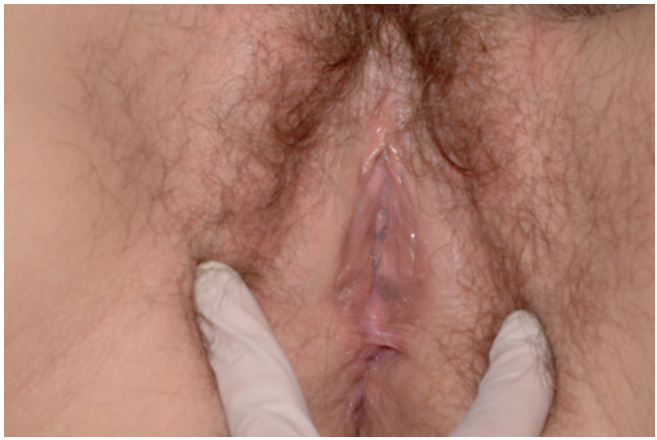

Figure 4.2 Lichen sclerosus with minimal lichenification

In both studies, the most common presenting symptoms were itching and soreness. However, other symptoms and signs at presentation are purpura, dysuria, constipation, genital erosions and extra-genital lesions.

Less than 10% of the children studied were asymptomatic and were discovered after biopsy for another reason.

Dysuria and pain with defecation leading to constipation are presentations quite different to adults who normally present with itch and dyspareunia. It is not uncommon for children with lichen sclerosus to be referred to urologists and gastroenterologists. If purpura is present, children with lichen sclerosus may be referred to child protection units.

The appearance in children is the same as in adults and atrophy, fusion of the labia as well as loss of vulval architecture also occurs.

Investigation

Although vulval lichen sclerosus generally has a characteristic clinical appearance and can be diagnosed clinically by an experienced health provider, a skin biopsy from the affected site provides diagnostic confirmation and exclusion of alternate diagnoses. False negatives are rare. A positive biopsy is helpful in counselling the patient about the important long-term consequences and the need for follow-up, and we recommend it for all patients. If biopsy is not possible or refused by the patient, a photographic record should be kept. It is also useful if the patient changes location or medical practitioners. Treated disease may appear normal and it is important that there is a clear, histopathological record of the diagnosis prior to treatment.

It should be noted that prior treatment with a topical corticosteroid may render the histological appearances non-specific. However, a biopsy from a white area is usually diagnostic.

In children, a clinical diagnosis is almost always sufficient because of the difficulties of a biopsy, limited differential diagnoses and also because neoplastic transformation has never been reported to occur in children with lichen sclerosus.

The histology is distinctive and uniform across ages and genders. The epidermis is atrophic with hydropic degeneration of basal cells and a homogenous pale zone in the upper dermis. There may also be a lichenoid infiltrate of mainly mononuclear cells in the dermis.

Differential Diagnosis

The differential diagnosis in adults is lichenification of any sort: dermatitis, extra-mammary Paget's disease, genital warts, non-pigmented seborrhoeic keratosis and VIN.

In children, vulval lichen sclerosus has the same characteristic clinical appearance seen in adults. Lichenified atopic dermatitis can simulate it in adults, but is much less likely to do so in children. Psoriasis may co-exist, causing diagnostic confusion because of superimposed erythema. Vitiligo lacks the epithelial changes seen in lichen sclerosus, presenting with sharply marginated white macules that fluoresce under ultra-violet light.

Lichen Sclerosus and Associated Malignancy

Before it was realised that lichen sclerosus could be adequately treated, about 60% of all vulval SCCs had histological evidence of adjacent lichen sclerosus, and it was well-established that adult women with vulval lichen sclerosus had a 2–6% lifetime risk of developing cancer. Any degree of lichen sclerosus, even mild disease, carries this non-negligible risk.

The appearance of a vulval SCC can include nodules, persistent fissures, hyperkeratotic plaques, non-healing ulcers and fungating tumours. Any change in an area of lichen sclerosus that does not promptly resolve with topical treatment must be biopsied.

There have been no reports of vulval malignancy associated with lichen sclerosus during childhood, but SCC of the vulva has been reported prior to the age of 40 in patients with childhood-onset lichen sclerosus.

The association of lichen sclerosus with genital malignancy has very important implications for management. Patients must be aware of the risk, be educated about what to look for and be regularly treated and followed up.

It should be noted that extra-genital lichen sclerosus is not associated with malignancy.

Malignancy after Treatment

Our prospective study of lichen sclerosus, published in 2015, compared 507 adult female patients who adhered to treatment and those who did not. It demonstrated that topical corticosteroid treatment that kept the skin objectively normal, also resulted in minimal scarring and a greatly reduced the risk of cancer. This confirms the findings of our previous retrospective study and the observations of other authors that adequate control of lichen sclerosus minimises the risk of subsequent malignancy. During the subsequent six years since publication, none of the patients in the cohort who have remained under observation using regular topical corticosteroid treatment have developed a cancer. This study also showed that regular treatment greatly reduces the likelihood of scarring or progression of existing scarring at the time of first presentation. This has also been born out in follow-up of the cohort.

We therefore believe that the interests of patients with lichen sclerosus are best served by making every attempt to complete disease suppression and careful surveillance.

Malignant melanoma accounts for 2% of all vulval cancers, and in the paediatric setting is considered a rare association with lichen sclerosus. It is important to bear in mind that

melanocytic proliferations associated with the condition have been documented. The significance of this is not known.

Prevention of Malignancy

Our research has demonstrated that regular suppressive treatment greatly reduces the risk of malignancy in patients with lichen sclerosus. A more recent study, published in 2019, showed that recurrence of malignancy in patients with lichen sclerosus who had had a cancer may also be reduced by regular treatment. Patients who have had a malignancy should remain under surveillance and it is of great importance that they continue their ongoing suppressive treatment.

Management

An Australian consensus statement for diagnosis and management of vulval lichen sclerosus has recently been published. Lichen sclerosus in adults is a lifelong disease that is unlikely to remit. Most patients are unable to stop treatment without eventual relapse, although this may take many months. It is important when counselling them to make sure that they understand that treatment should be assumed to be for life. This news is very difficult for many patients to assimilate and it is important to impress it upon them at every visit until you are sure that they understand. A comparison to diabetes, which most patients understand is not curable, is useful as an analogy.

In the very unusual instances where patients have apparently remitted, they need to be kept under long-term observation as lichen sclerosus can re-activate after years of dormancy.

Topical Therapy

There are two phases of treatment for lichen sclerosus:

1. induction of remission
2. maintenance treatment.

It is now accepted that potent topical corticosteroid is the gold standard for obtaining remission in vulval lichen sclerosus. The first report of this treatment was published in 1991 using clobetasol propionate 0.05%, an 'ultra-potent' topical corticosteroid. Since that first courageous study, lichen sclerosus has become one of the easiest vulval conditions to manage, and many further studies with potent and super-potent topical corticosteroid have confirmed this as a safe and highly effective treatment.

Lichen sclerosus is in fact so responsive to topical corticosteroid that failure to improve should be reason to suspect either that the diagnosis is wrong, the patient is not using the treatment or there are other factors confounding it, such as allergy or super-infection.

Most of the subsequent studies have also used clobetasol propionate, with more recent studies comparing its efficacy with that of mometasone furoate 0.1%. Our experience has been that, in almost all cases, less potent products will produce results that are just as good. The clinician should first decide if the lesions are more or less hyperkeratotic, and match the potency of the topical steroid to the severity of the skin disease.

The main focus of treatment should not be on the product used but the end result: attaining and maintaining normal skin. There is no single way to do this and clinicians can make their own judgement relative to the severity of the patient's disease and their preference for daily or intermittent treatment. It has, however, been our observation that the

regimens that work best are used at least three to four times a week. Feedback from our patients is that very intermittent regimens are easily forgotten.

Induction of Remission

The judgement of topical corticosteroid potency is made based on the degree of hyperkeratosis (thickening) of the vulval skin. Although scarring may contribute to severity, it cannot be changed by treatment.

We recommend for:

- severely hyperkeratotic disease: super-potent corticosteroid (e.g., clobetasol propionate 0.05% ointment) twice daily until itching has ceased (usually one to two weeks) then daily until review at six weeks
- hyperkeratotic disease: potent corticosteroid (e.g., betamethasone dipropionate 0.05%, mometasone furoate 0.1% twice daily until itching has ceased then daily until review at six weeks
- mild disease with only pallor and very little hyperkeratosis: moderate corticosteroid (e.g., triamcinolone acetonide 0.02%, methylprednisolone aceponate 0.1%, aclometasone dipropionate 0.05%) daily until review at six weeks.

The six-week review is to check for side effects, response to treatment and for emotional support, as the diagnosis of lichen sclerosus is difficult for most women. At this point, they are usually feeling much better and many assume they are cured. It is important to emphasise that treatment must now be maintained long-term and to explain that the reason for this is to prevent cancer and scarring.

The initial potency of topical corticosteroid is continued until the skin texture and colour has returned to normal. It should be noted that there may be residual hyper- or hypopigmentation. However, the clinical appearance of the surface of the skin usually improves markedly.

Patients are reviewed again three months later, and then every six months for the first two years. The potency of the topical corticosteroid is slowly titrated down to a moderate to mild potency for maintenance therapy.

The aim of treatment is the disappearance of abnormal signs as well as the resolution of symptoms. Symptom resolution occurs quickly but resolution of abnormal signs takes longer. Patients must therefore continue their regular treatment even after symptom resolution. It is important to be guided by objective clinical response. We have found that compliance is best when patients incorporate treatment into their daily routines, and that patients whose compliance is dubious benefit from regular six monthly reviews. The average time to return to normal skin once treatment is commenced is from four to six months of continuous treatment.

Maintenance Treatment

Regimens for maintenance treatment of lichen sclerosus have been much less well researched and as a result less defined than those for initial disease control. Although many reviews and published articles state that the condition does not spontaneously resolve and has to be controlled, there is no consensus on what this long-term control involves.

Prior to the publication of our research, the weakness of most published studies related to length of follow-up. For almost all publications, the longest period of observation documented is three years.

There are two exceptions: a descriptive cohort study from the UK, with a mean length of follow up of 66 months, and a long-term study from France, which was conducted prospectively over ten years. This latter study of 83 women is the best evidence we have to confirm what most experienced practitioners know: that although topical corticosteroid easily induces remission, it does not cure lichen sclerosus. This study reported an 84% recurrence rate if treatment was ceased.

Both of these studies suggested that treatment might also change the course of the disease, reducing the risk of cancer and scarring, which we later confirmed.

The maintenance regimen used in the French and UK studies was intermittent clobetasol propionate once to three times a week, and this is what most other published papers have stated ever since. However, there is no single way to treat lichen sclerosus long-term, because differing degrees of severity require different regimes. What is important is maintenance of normal skin texture and colour. Treatment should be titrated accordingly.

The main potential problems with long-term use of potent topical corticosteroids on the genital area are atrophy, evidenced by fragility and striae, peri-orificial dermatitis and super-infection with *Candida albicans*. Interestingly, the French and UK studies and another from the UK with a three-year follow-up period recorded that such side effects were rare when treating lichen sclerosus. This has been our experience as well. In our own studies, side effects were minimal and reversible and were confined to skin fragility and erythema. The argument that long-term topical corticosteroid will produce atrophy is therefore not valid in lichen sclerosus.

We recommend that treatment is re-evaluated every six months in order to determine the lowest maintenance regimen that will ensure continuing remission. Topical corticosteroid treatment is constantly titrated to the degree of hyperkeratosis. If this relapses, the strength of treatment increases. If atrophy or corticosteroid dermatitis, evidenced by irritability and redness, occurs, potency is reduced. We have found this method of managing lichen sclerosus patients long-term to be successful, safe, inexpensive and outstandingly effective. As stated, none of our compliant patients have developed a cancer, and over 95% have had no further disease progression or scarring. Over 90% have complete and sustained symptom control, and of those who are sexually active, over 90% no longer experience dyspareunia. Our most recent study demonstrates that good control results in a marked improvement in quality of life.

We strongly discourage regimens that are used on an 'as needed' basis to control symptoms only. Symptom control in lichen sclerosus is not difficult to achieve, but objective disease suppression should be the target outcome, or the patient is still at risk of complications. It is a common theme amongst patients who have succumbed to cancer or disease progression as a result of poor compliance with treatment that they had remained asymptomatic.

Our recommended long-term follow-up regimen is:

- that patients are reviewed every six months until they have been in a stable remission for two years, then yearly with the proviso that they have an examination by their general practitioner half way through that year and come back earlier if they have any concerns.
- if evidence of relapse occurs on treatment, more potent corticosteroid is used until this settles.

- if there is evidence of corticosteroid excess, less is used. Corticosteroid excess usually evidences itself with vulval redness and burning or fragility. This reverses quickly once treatment is adjusted.
- patients should be encouraged not to stop treatment once they are in remission, but to continue with the lowest dose of corticosteroid possible to maintain complete objective normality. The psychological impact of a recurrence on a patient who is finally in remission after years of suffering can be devastating. Furthermore, patients who do not comply with treatment have a 50% risk of scarring and a 5% risk of development of malignancy. Each review is an opportunity to remind your patients of the importance and safety of maintenance treatment.

The main outcome measures of treatment are:

- symptom control: no itch or soreness is expected
- ability to have intercourse: in post-menopausal women, this may also require topical oestrogen to reduce any associated menopausal atrophy
- prevention of scarring, fusion and loss of clitoral substance (reduction in labia minora after menopause is common, not problematic but not always prevented by treatment)
- prevention of malignancy
- lack of side effects.

Side Effects of Treatment

Side effects are remarkably few. We rarely see corticosteroid-induced atrophy.

We have encountered:

- candidiasis: this is easily controlled with antifungal therapy
- erythema: this responds rapidly to a reduction in corticosteroid strength
- stinging from topical therapy: this usually settles as fissures and erosions heal. It is virtually always possible to find a well-tolerated topical corticosteroid.

Some patients may have recalcitrant thickened areas that appear non-responsive even to super-potent corticosteroid. These should always be biopsied to rule out malignancy. Such lesions may respond to intra-lesional corticosteroid if they are causing distress.

In some patients with very hyperkeratotic disease, ablative laser treatment can be a useful adjunct to treatment. It is not a substitute for topical therapy, however, and this has to be continued after laser treatment to reduce hyperkeratosis.

The most important principle is to maintain observation. This condition is pre-malignant, potentially unpredictable and liable to recur if patients become complacent about its management. It has been argued by some authors that this type of approach is a burden on the health system. However, we argue that lichen sclerosus is not a common disease and that the cost of even one patient with a vulval cancer should be compared to the cost of follow-up of many to ensure that cancer does not occur.

Other Topical Therapies

Topical immunosuppressive agents, such as tacrolimus and pimecrolimus, have been described as potentially playing a role in the treatment of lichen sclerosus in children and adults.

There has been one phase II trial to assess the safety and efficacy of tacrolimus ointment 0.1% for the treatment of lichen sclerosus, and the results were released in 2006. Clearance

of active lichen sclerosus was reached by 43% of patients at 24 weeks of treatment and partial resolution was reached in a further 34% of patients. Maximal effects of therapy occurred between weeks 10 and 24 of treatment. The authors, who recommend topical immunosuppressives, state that they are less likely to cause atrophy. However, we have rarely experienced atrophy in our corticosteroid-treated patients, and when it occurs, it invariably improves with a lower dose.

While there were no adverse events during the 18 months of follow-up, the theoretical disadvantage of topical immunosuppressive agents is an increased risk of malignant transformation due to local immunosuppression. This is arguably an important consideration given the well-described association of vulval lichen sclerosus and malignancy. Squamous cell carcinoma has been reported in adults with lichen sclerosus in association with pimecrolimus and tacrolimus treatment.

There is insufficient data to recommend topical immunosuppressive agents to treat lichen sclerosus, and no justification when topical corticosteroid is so effective and safe. Topical immunosuppressive agents have no advantage over topical corticosteroids whatsoever. They are more expensive, very likely to sting and burn and their long-term safety is not established.

Historically, topical testosterone has been used to treat vulval lichen sclerosus. However, there is no longer any role for it, as it is ineffective and may produce androgenisation in girls.

Similarly, topical oestrogen is of no value, other than to reduce hypo-oestrogenic atrophy in sexually active post-menopausal women.

Management in Children and Adolescents

The situation in children is less well documented than in adults. In pre-pubertal children with lichen sclerosus, there is anecdotal evidence that prompt diagnosis and treatment may induce a remission. Children with lichen sclerosus rarely have severely hyperkeratotic disease and therefore the recommendation is to commence treatment with a potent corticosteroid and to manage in the same way as managing an adult.

Historically, it was thought that childhood vulval lichen sclerosus improved or remitted at puberty. This is not correct. Only two studies have examined lichen sclerosus in adolescents who developed it as children and both have cast doubt on this assumption. A study of twelve adolescents with a follow-up duration of up to ten years supports the conclusion that remission is very unlikely, and also suggested that treatment could prevent sequelae. Once children with lichen sclerosus reach puberty, they usually require long-term management just like an adult. It is therefore essential that parents and patients understand that it is unlikely to resolve at puberty, and that it requires long-term follow-up exactly as in adults.

Our group has recently completed a retrospective study of 46 children with lichen sclerosus, again comparing compliant patients with non-compliant ones. We have shown that when normal skin is attained and maintained, progression of the disease ceases and scarring and atrophy do not occur. Scarring that is present prior to treatment, however, does not reverse. The commonest is loss of labia minora and clitoral phimosis. The latter may be reversed by treatment, but once labia minora have resorbed, they will not re-grow. More recently, our research has shown that the risk of scarring in children who are not compliant with treatment is greatly increased. This appears particularly important around puberty when the genital area is developing adult proportions.

Follow-up of teenagers is difficult because of their embarrassment about examination. In order to avoid this embarrassment, many will assure their parents and doctor that they are asymptomatic, and as a result, may be lost to follow-up. A trusting relationship with their doctor prior to puberty is the best way to prevent this.

Lichen sclerosus may have a profound effect on children and adolescents. It is common for them to develop unvoiced fears around sexuality and reproduction. Many of them feel isolated and different to their peers. Fear of using tampons is common and related to difficulties related to their own genital area.

Around puberty when children should be developing autonomy, many start to refuse treatment and wish to self-manage. It is around this important stage that control can be lost, resulting in failure to develop normally. This is an area of social science that has never been researched. More work is required in determining how best to help these young adults.

Lichen Sclerosus and Sexual Abuse in Children

Sexual abuse concerns often arise when children with lichen sclerosus are examined because of the associated erosions, fissures, purpuric lesions, bleeding, and scarring. There have been numerous reports of patients with the classic presentation of lichen sclerosus undergoing extensive, inappropriate evaluation for sexual abuse. Although the awareness of sexual abuse amongst health workers has often resulted in more timely referrals by paediatricians for the proper diagnosis of lichen sclerosus, the added emotional trauma to the family is completely unnecessary.

Sexual abuse is common and many retrospective studies suggest that approximately 20–25% of all females have been abused as children. However, children who have been sexually abused rarely have clinical signs when examined. A diagnosis of lichen sclerosus neither rules out or proves sexual abuse.

Surgical Therapy

Historically, vulvectomies have been performed in adults for lichen sclerosus, but the disease recurs. This is no longer considered an acceptable method of treatment and is completely contraindicated. Surgery is rarely appropriate therapy in the paediatric population unless significant fusion of the labia has occurred.

Various surgical procedures have been used to treat labial and peri-clitoral adhesions. Simple division of adhesions gives a very satisfactory result, provided that potent topical steroids are used daily post-operatively until healing is well underway. It is sometimes necessary to apply the post-operative steroid on a dilator. We do not advocate perineoplasty for women with dyspareunia caused by posterior introital fusion. Again, simple division of adhesions works well.

Can Patients with Lichen Sclerosus Resume Normal Sexual Activity?

In most cases, the answer is yes. Physically, particularly in younger women and those in whom the disease was treated before it became too advanced, there is usually no reason why they should not be able to resume a normal sex life.

Women who have had a long history of painful sex have usually developed significant pelvic floor spasm and may need physiotherapy to overcome. Others admit to having developed a distaste for intercourse while they were as yet undiagnosed and may need psychological help. There are of course many older women who

decline any help because their lack of interest in sex has been legitimised by their disease. Those who want to become sexually active again usually do.

Other White Lesions on the Vulva

White lesions on the vulva other than lichen sclerosus are unusual and uncommon; however, there are a few that are important differential diagnoses.

Case Study

A 25-year-old woman is referred with intermittent vulval itch since childhood, becoming worse since her menarche. She has used anti-fungals all her life, which have largely been ineffective. Vaginal cultures for *Candida* have always been negative. The referring doctor has made a clinical diagnosis of lichen sclerosus, which has terrified her. She has chronic scalp itch, and a family history of psoriasis.

Examination reveals a thick white patch of skin on the perineum, extending onto the labia majora. A punch biopsy of this rash excludes lichen sclerosus. A vaginal culture is again negative.

This woman has vulval psoriasis. She is commenced on a moderate-potency corticosteroid and 2% LPC ointment, both daily for six weeks, and then LPC ointment alone, twice daily. At review after three months, she reports a large improvement in her itch, but with exacerbations during periods. She is advised to use either menstrual underwear or a menstrual cup instead of tampons and pads.

Lichenified Dermatitis (Also Known as 'Lichen Simplex Chronicus')

Any skin disease that is chronic and itchy can evolve into a state called 'lichenification'. It is thought to be the way that skin reacts to long periods of scratching. In the past, lichen simplex chronicus was thought to be psychological in origin, but this is not generally true.

Underlying conditions, in addition to lichen sclerosus, that may become lichenified on the vulva include dermatitis, psoriasis and lichen planus (see Chapters 2 and 4). As a result, these predominantly red dermatoses contain patchy areas that are white and have abnormal thickened texture.

Presentation

The clinical appearance shows obvious areas of thickening, rugosity, excoriation and scale (Figure 4.3). On the rest of the skin, lichenification usually looks red or brown, but on the vulva, it often looks white.

Lichenification does not display the same subtle atrophic, wrinkled surface of lichen sclerosus, and is never bullous or haemorrhagic. It is most often seen on the perineum and peri-anal area but sometimes is more widespread, or conversely, localised and asymmetrical. It is invariably very itchy. Patients are usually atopic and have dermatitis elsewhere.

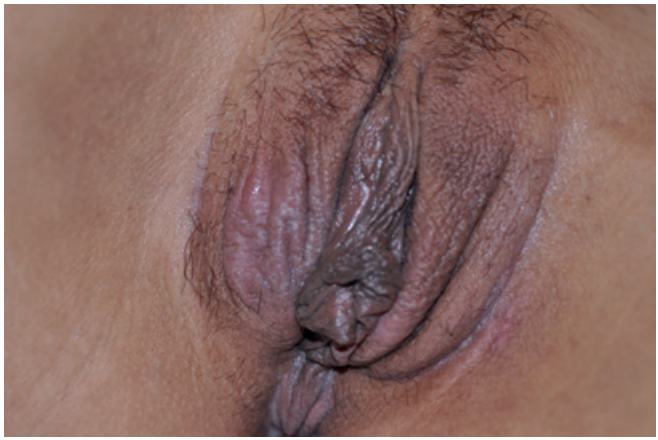

Figure 4.3 Lichenified dermatitis

Investigation

In most cases, the only way to differentiate lichenification from lichen sclerosus is with a skin biopsy. It is important to differentiate between the two because of long-term prognosis. But remember: if your clinical impression is lichen sclerosus, particularly if there is scarring or loss of substance, then you are probably correct no matter what the biopsy shows.

The histology of lichenified dermatitis usually shows evidence of spongiosis and acanthosis (thickening of the epidermis). If your report says 'non-specific', then you may just be dealing with treated lichen sclerosus.

When you read literature on vulval disease, you may encounter the term 'squamous hyperplasia' mentioned in articles on vulval cancer. This is a histopathological term that correlates clinically with lichenification. It may be confusing, however, because it suggests something malignant when what it is describing is completely benign. Squamous hyperplasia is not an indication for surgery.

Human Papillomavirus Infection

Human papillomavirus (HPV) infection usually presents on genital skin with discrete lesions that are unmistakably genital warts. There is, however, a variant that comes into the differential diagnosis of white lesions that presents with a hyperkeratotic plaque very often on the perineum.

This can be indistinguishable from lichenified dermatitis, lichen sclerosus and malignancy. Such white plaques should be biopsied and obviously the treatment for these various entities is very different.

Pigment Change: Vitiligo and Post-inflammatory Hypo-pigmentation Vitiligo

Vitiligo is a relatively common autoimmune disease, which results in patchy, very well-defined areas of complete loss of pigment on the skin, resulting in striking white decolouration (Figure 4.4). The surface of the skin retains its normal texture and this is the important clinical feature that differentiates it from lichen sclerosus, with which it may, confusingly, co-exist. Vitiligo is asymptomatic and is predominantly a cosmetic issue.

Figure 4.4 Vitiligo

Presentation

The most common sites for vitiligo are the face, hands, arms and legs; however, it can occur on any site. It usually occurs as multiple lesions but may be localised to one area. This is known as 'segmental vitiligo'. It may also be localised to the vulva.

Vitiligo is harmless. It may occur in association with other autoimmune diseases, particularly thyroiditis. If there is a family history of autoimmune disease or the patient is unwell, further investigation for other autoimmune diseases, particularly thyroid disease, should be carried out. However, most patients are well. Vitiligo on sun-exposed areas presents a risk for severe sun burn because the skin has lost melanin, its natural sun protection. Obviously, this is not an issue on the genital area.

Vitiligo can co-exist with lichen sclerosus. When both lichen sclerosus and vitiligo are found together on the vulva, the clinical presentation may be very confusing.

Investigation

The key to differentiating vulval vitiligo from vulval lichen sclerosus is that there is no textural change. Ultra-violet light is an easy way to confirm the diagnosis and an inexpensive hand-held device can be purchased online. Vitiligo is brightly fluorescent under ultra-violet light. You need a darkened room to do this. If there is doubt, a biopsy will diagnose vitiligo.

In vitiligo, all melanocytes have disappeared and the inflammatory signs of lichen sclerosus are not present. Therefore, even if lichen sclerosus has been treated and the classic inflammatory signs have resolved, it will still be histologically different to lichen sclerosus.

Management

Vitiligo is difficult to treat. There are many treatments including potent topical corticosteroids, tacrolimus, calcipotriol and topical psoralens (these are medications that must be exposed to sunlight to become activated and therefore not practical on the vulva). All treatments are slow to respond, requiring many months to become effective.

When vitiligo occurs on the genital area, it is doubtful that it requires treatment and patients are usually happy to be told that their condition is harmless. The treatments available are likely to be irritating and prolonged use of potent topical corticosteroid on vulval skin unaffected by lichen sclerosus will result in atrophy and peri-orificial dermatitis.

When vitiligo and lichen sclerosus occur together on the vulva, corticosteroid treatment of the lichen sclerosus can result in re-pigmentation of the vitiligo. However, in the absence of lichen sclerosus, potent topical corticosteroids should not be used on vulval vitiligo.

Post-inflammatory Hypo-pigmentation

Any inflammatory dermatosis can result in loss of pigment from the skin. This is most often seen in non-Caucasians and Caucasians with olive skin. It is commonly seen on the perineum after obstetric surgical repairs.

Presentation

When post-inflammatory pigmentation occurs, the edges are not well defined, and there is often some textural change present because of the underlying dermatosis that caused it.

Unlike vitiligo, there is not a complete loss of melanocytes and therefore the appearance is of colour attenuation. This means it looks paler than surrounding skin, rather than white. The dermatoses that occur on the vulva that result in loss of pigment include psoriasis, any form of dermatitis and lichen sclerosus.

When this type of colour loss occurs after lichen sclerosus has been treated, it can cause confusion about when to reduce treatment. When treating lichen sclerosus, the key is to observe very carefully for loss of textural change (thickening, atrophy, wrinkling). In some patients, it may take many months for normal colour to return and in some it never returns to normal.

History

The patient gives a history of a previous dermatosis in the same area, including symptoms of itch.

Management

Post-inflammatory hypo-pigmentation is harmless and does not specifically require treatment. It usually resolves spontaneously once the underlying dermatosis is treated.

Squamous VIN

Vulval neoplasia is rare. VIN is the most common form of vulval neoplasia and is primarily seen in patients with lichen sclerosus, or in conjunction with oncogenic HPV infection, most often with genotypes 16 and 18, which are associated with cancer of the cervix.

Other skin cancers such as basal cell carcinoma, extra-mammary Paget's disease and melanoma can also occur on the vulva, but these are even more uncommon than VIN and are unlikely to appear white as they are rarely hyperkeratotic. The exception is extra-mammary Paget's disease (which can be viewed in any case as a non-squamous subset of VIN), which classically is red but may have a white element that looks like 'icing sugar'.

Despite its name, VIN is histopathologically no different from what we call SCC in situ (or more commonly, Bowen's disease) on non-genital sun-exposed skin. It has not breached the basement membrane, which in turn means invasive disease has not yet occurred.

On non-genital skin, it is well accepted that in situ disease is the precursor of invasive disease. It is not known whether squamous VIN is the precursor of invasive vulval SCC and gynaecological oncologists therefore do not regard VIN as a true malignancy.

Dermatologists might disagree with this approach as they know that Bowen's disease rarely, but definitely, progresses to invasive SCC on sun-exposed skin, and they see the genital area as part of the skin.

Presentation

Vulval intra-epithelial neoplasia may look quite different from Bowen's disease. The latter usually presents as red patches, which look very much like eczema. The appearance of VIN is highly variable. Bowen's disease on sun-exposed skin has an excellent prognosis and is usually easily treated with a variety of minimally invasive procedures. Vulval intra-epithelial neoplasia has a more guarded prognosis and a tendency to recur after the sort of treatment that would be more than adequate for Bowen's disease. This possibly relates to the different carcinogens involved. In sun-exposed skin, this is ultra-violet light, whereas on the genital area, it is either oncogenic HPV or lichen sclerosus.

Vulval intra-epithelial neoplasia may present as thickened white or red mucosal patches, hyperpigmented plaques, warty lesions or persistent erosions or ulcers (see Figure 4.5). The distribution may be single or multifocal and is usually asymmetrical. Any part of the vulva can be affected, as well as the peri-anal skin and anus. Unless frank nodules are present, it is often very difficult to tell VIN from invasive SCC clinically. Only a biopsy will make this differentiation.

Unlike Bowen's disease on extra-genital skin, which is usually asymptomatic, VIN and vulval SCC can be itchy, particularly when seen on a background of lichen sclerosus.

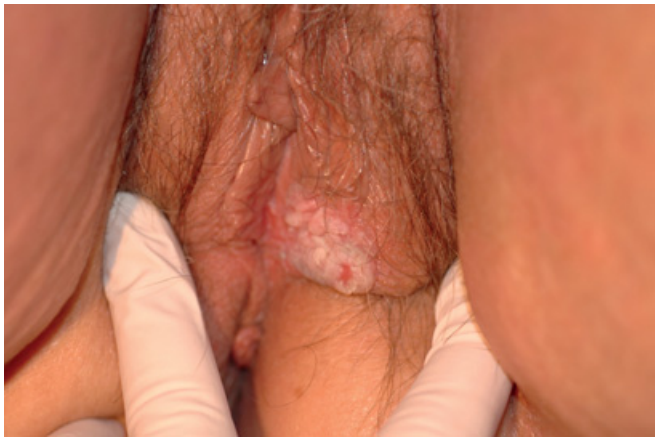

Figure 4.5 Vulval intra-epithelial neoplasia

About half of the patients are asymptomatic at diagnosis, but most symptomatic patients report pruritus. If ulceration occurs, VIN is painful and may bleed. Extra-mammary Paget's disease is characteristically itchy and is often mistaken for dermatitis.

VIN Terminology

The understanding of VIN has been confused by the previous application of cervical grading systems (cervical intra-epithelial neoplasia) to the vulva. In the past, a three-grade histopathological staging system of VIN, I, II and III, was proposed based on cervical intra-epithelial neoplasia.

This system was not helpful in describing the actual biology of VIN and was abandoned in favour of a new classification that was published in 2005.

This latest classification divides VIN into two types:

- 'usual' VIN (uVIN): this type is HPV related and is further divided into two sub-categories: warty and basaloid
- 'differentiated' VIN (dVIN): this type is most often found in association with lichen sclerosus and lichen planus.

Warty VIN is usually HPV related, occurs in younger patients and has a multifocal, warty appearance. It has the lowest potential for invasive carcinoma. Basaloid VIN is more likely to be a single, well-defined lesion and occurs in older patients.

Differentiated VIN occurs in older patients, usually with lichen sclerosus, and is the most dangerous in terms of the potential to invade and metastasise. This is confusing because, when talking about cancer, most of us think of differentiated as being less, rather than more dangerous.

There are a number of names that have been previously given to genital lesions with the same histopathological appearance. They include Bowenoid papulosis and erythroplasia of Queyrat. These are clinical descriptors. Bowenoid papulosis suggests multiple brown, warty papules, and erythroplasia of Queyrat suggests red, shiny patches.

You may also read about the term SIL, which stands for squamous intra-epithelial lesion. This plethora of terminology tells us something about how confused this subject is in the medical literature.

The take-home message is that although controversy exists about prognosis, all types of VIN have the potential to progress to invasive cancer, and older age of the patient and the presence of lichen sclerosus are the factors that make this more likely. From a clinician's perspective, the one term, VIN, is probably all we need or most lesions except melanoma and extra-mammary Paget's disease.

Epidemiology of VIN

Vulval cancer occurs in two settings:

1. younger women with genital oncogenic HPV infection, most commonly types 16 and 18. Smoking or immunosuppression are frequently associated. It has been shown that HPV immunisation reduced the incidence of carcinoma of the cervix. This may also extend to HPV-associated vulval carcinoma.
2. older women with hyperkeratotic lichen sclerosus and less commonly lichen planus. These women are not smokers nor do they have HPV-related disease. They are typically post-menopausal.

Vulval intra-epithelial neoplasia has been very rarely described before the third decade. Although the two types often occur in different age groups, both can occur at any age.

Any white lesion that is not rapidly responsive to therapy with potent topical corticosteroid should be biopsied as soon as possible, particularly in any of the settings described earlier.

Multi-focal squamous malignancy involving the cervix, vagina, vulva and anus is not rare, and a careful visual inspection of the entire lower genital tract, including a Pap test, is mandatory.

Investigation

Any lesion suspected of being VIN, including white patches, persistent ulcers, warty lesions, red shiny patches, bleeding lesions, pigmented plaques and papules, should be biopsied immediately.

When taking the biopsy, choose a thick warty area that has not responded to treatment. Biopsies from erosions are often non-specific. If the result of a small punch biopsy gives a non-specific result, refer to a gynaecologist for an excisional biopsy.

Especially in younger women, a Pap test should be performed. Any patient with lichen sclerosus should have 6–12-monthly checks for evidence of super-imposed VIN, and any non-responsive lesion should be immediately biopsied.

Management

It is beyond the scope of this book to talk in detail about the management of vulval cancer. A number of modalities have been described including:

excisional surgery

laser surgery

photodynamic therapy

topical immunomodulators such as imiquimod.

Treatment is difficult because of multifocality and the surgical problems associated with operating on the genital area. Most studies report high recurrence rates no matter what modality is used and whether or not surgical margins have been reported as clear of tumour.

Evidence that removal of VIN prevents later SCC is lacking. Despite this, excisional surgery is currently the gold standard. Some experts have recommended a conservative approach, particularly with multifocal, HPV-related VIN, with only symptomatic treatment.

The main risk factors for invasive cancer are:

- age
- raised, solitary lesion
- immunosuppression
- previous radiotherapy to the genital tract.

These patients should always be referred to a gynaecological oncologist for further management. Even if the patient opts for medical therapy, she should first have the opportunity to discuss all options with a clinician experienced in this area.

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Chapter 5

Things That Ulcerate, Blister and Erode

Contents

Diseases of the vulva that are primarily erosive or ulcerative are uncommon. Notwithstanding, fissures or excoriations can occasionally complicate almost any dermatological disease of the vulva.

Common conditions such as dermatitis and psoriasis may become eroded by scratching, and allergic contact dermatitis often causes such severe oedema that blistering occurs.

Certain conditions, which are not usually ulcerative or bullous, may have rare variants that are, for example, the bullous variant of lichen sclerosus. Vulval cancer may ulcerate when advanced.

This chapter focuses on conditions where ulceration or erosion are a characteristic part of the disease. It is important to understand the difference between ulceration and erosion: ulceration means full-thickness loss of the epithelium, whereas erosion means partial-thickness epithelial loss.

Lichen Planus

Lichen planus is a rare disease. It is very difficult to recognise in general practice and patients are usually referred to a dermatologist before a diagnosis is made. It is almost always a disease of adults, usually after middle-age.

Mucosal lichen planus is the commonest type on genital skin. This is the type that involves the oral mucosa, and (rarely) the oesophagus and anal canal. Unlike lichen sclerosus, which never involves the vagina proper, lichen planus often does (see Figure 5.1).

The common type of cutaneous lichen planus presenting with itchy violaceous papules with white streaks (Wickham's striae) occurs on the vulva but is very uncommon compared to the mucosal type.

Presentation

The typical patient with vulval lichen planus is in her 50's and beyond; however, it can occur in women in the second or third decade of life. In younger patients, it is a devastating diagnosis. It is exceptionally rare in children.

The typical presentation is of pain, dyspareunia and heavy non-offensive discharge. Itch is less common. Dyspareunia is usually severe, and patients have often become completely apareunic. If the anal canal is involved, pain with defaecation is characteristic.

When oral disease is also present, patients notice oral soreness and sensitivity as well as tenderness, raw areas and gingivitis.

Aetiology

The pathogenesis of lichen planus is T-cell-related but the exact nature of the disease remains unknown. Rarely, it is related to hepatitis B and C and has been reported to be precipitated by hepatitis B vaccination.

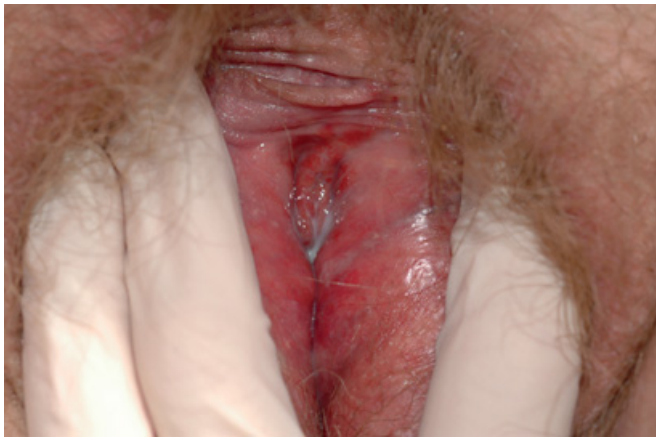

Figure 5.1 Vulvo-vaginal lichen planus. Note the subtle erosions and heavy vaginal discharge

Oral lichen planus has been associated with amalgam fillings. However, most cases are idiopathic.

Examination

Physical examination is often confusingly non-specific, ranging from non-specific erythema to frank ulceration. The hallmark of lichen planus is erosion of the mucosal surface of the introitus, often extending into the vagina (see Figure 5.1). This presents as glazed erythema but if one looks closely, the loss of epithelium is evident (see Figure 5.2). Interspersed with this bright erythema you may notice grey patches. This is a classic clinical finding that is very helpful but unfortunately not invariable.

The rash is usually confined to the labia minora, the sulcus between the majora and minora, introitus, perineum and vagina. However, extension to the peri-anal area and the anus may occur. As a result, the anal mucosa may also appear brightly erythematous and may also scar and stenose.

If lichen planus is not diagnosed and treated early, scarring will usually occur. On the vulva, this may involve loss of the labia minora, labial fusion (either anteriorly or posteriorly) and complete obliteration of the clitoris by clitoral hood adhesions. Clitoral scarring may produce recurrent clitoral abscesses. In the vagina, scarring may result in occlusion or stenosis, making speculum examination impossible.

Lichen planus is highly treatment-resistant and patients do not respond to weak topical corticosteroids.

Investigation

The classic histological appearance of lichen planus (a predominantly lymphocytic infiltrate at the dermo-epidermal junction) is often not seen in genital biopsies. Therefore, you may need to make a clinical diagnosis.

Vulval biopsy should never be taken from an eroded area. Take the sample from adjacent skin, or a grey area.

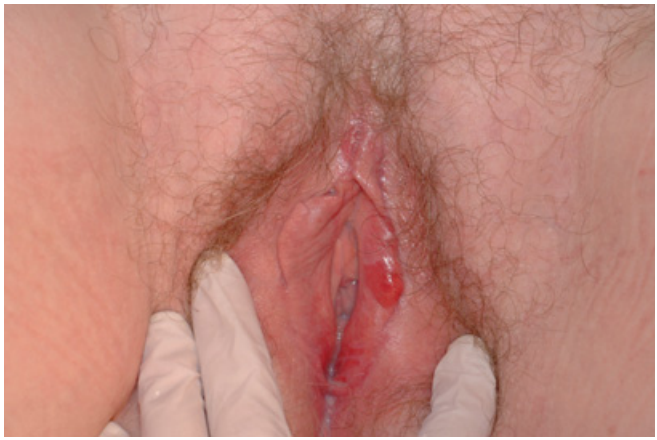

Figure 5.2 Vulval lichen planus. Note the more typical glazed erosions

Management

Initial management in all cases is with topical and/or oral corticosteroids, but these must be potent or super-potent. Even over many years, this is effective and without side effects. Milder cases, usually in elderly women, may be able to be controlled by daily potent or even moderate steroids.

More severe cases often require much more aggressive treatment including oral prednisone. In these cases, a steroid-sparing agent should be introduced as soon as possible to minimise side effects.

Steroid-sparing agents that are available include:

- topical tacrolimus
- oral retinoids
- oral weekly methotrexate
- oral azathioprine
- oral mycophenolate.

The details of how to use these agents are beyond the scope of this text. If you suspect that your patient has lichen planus, referral to a specialist is highly recommended. We do not recommend treatment of vulvo-vaginal lichen planus in general practice.

Surgical Treatment

Scarring and stenosis often require correction but this must only be attempted once medical treatment has stabilised the disease. Furthermore, surgical division of adhesions will inevitably flare the lichen planus, and medical management must be in place pre-operatively to prevent this.

Follow-up

Lichen planus is usually a long-term condition that must be controlled. Our aim is to induce remission with oral and/or topical therapy and tailor maintenance therapy to suit the individual, minimising this as much as possible.

In our experience, the majority of patients can achieve good control, but it may take up to a year or even longer to achieve this. Not all patients are able to achieve painless intercourse and a high degree of motivation is required.

Lichen planus is often a devastating and life-changing event for patients, necessitating a huge adjustment in lifestyle, including daily medication, some of which involves significant risk and loss of sexual enjoyment and activity. The impact on the patient should never be underestimated.

Differential Diagnosis

Both lichen planus and lichen sclerosus are scarring conditions where loss of vulval architecture is typical. There are, however, important differences (Table 5.1).

Other differential diagnoses are:

- graft versus host disease
- any autoimmune bullous disease, but particularly mucosal pemphigoid and bullous pemphigus
- desquamative inflammatory vulvo-vaginitis (DIV)
- fixed drug eruption of the vulva.

Table 5.1 Differentiating lichen sclerosus and lichen planus

	Lichen planus	Lichen sclerosus
Site	Vulva and/or vagina	Vulva only, never involves vagina
Appearance	Red, erythematous with patches of grey	White. Redness only seen with super-imposed psoriasis, candidiasis or corticosteroid
Erosions	Yes, primarily erosion of the mucosal surface of the introitus	Not primarily, however, may split or ulcerate as a complication of the disease
Childhood	Extremely rare	Occurs in children
Association with vulval cancer	Probable association	Association well documented
Treatment	Highly treatment-resistant	Responds readily to treatment

Graft versus Host Disease

This is the condition that most closely mimics vulval lichen planus. Patients with known chronic graft versus host disease not uncommonly experience mucosal involvement indistinguishable from lichen planus. Treatment is very similar.

Graft versus host disease occurs in 20–30% of stem cell transplant recipients. It often occurs in the mouth, but tends to be overlooked in the vagina because symptoms may be minimal or absent until intercourse is attempted. Vulval and vaginal scarring may make intercourse impossible. It is therefore essential that all female stem cell transplant recipients are examined at three to six months post-transplant, even if asymptomatic.

Only appropriately experienced specialists, in close conjunction with the treating haematologist, should manage these patients.

Non-infective Chronic Vulvo-vaginitis (DIV)

Desquamative inflammatory vulvo-vaginitis is an uncommon non-infective, non-erosive chronic vulvo-vaginitis. The precise aetiology is unknown, but there appears to be a loss of vaginal homeostasis.

It is not actually an erosive condition, but because it is very often confused with lichen planus, which may be erosive, it will be discussed here.

We believe that DIV is probably the same entity as Zoon's vulvitis and plasma cell vulvitis. All of these names create confusion and it would probably best for international consensus to provide an overarching term.

Like lichen planus, it involves the vagina and mucosal surface of the labia minora. However, unlike lichen planus, it does not extend onto the external hair-bearing vulval skin, does not involve non-genital skin or the oral mucosa, and it does not produce scarring.

Professor Sobel established diagnostic criteria based on a case series of 51 patients. These were:

- absence of infection, synechiae or stenosis
- purulent exudate
- increased parabasal cells on wet film
- elevated vaginal pH

- gram stain showing relative loss of gram-positive bacilli and the presence of gram-positive cocci with the presence of polymorphonuclear leukocytes.

Desquamative inflammatory vulvo-vaginitis is not usually a disease of young women and has never been described in a child. The typical patient is in the fourth decade and beyond.

Presentation

The symptoms are usually a combination of soreness, dyspareunia and discharge (occasionally bloody). Itch is not a prominent feature and scarring is never encountered.

The appearance of the vulvo-vaginitis varies from non-specific confluent dull or glazed erythema, to patchy erythema to petechiae and petechial patches (see Figure 5.3). A non-offensive, greenish discharge is often but not always present. Although on first inspection the rash may look erosive, there is no true epithelial breach.

Investigation

Microscopy may reveal polymorphs and loss of lactobacilli, both of which are non-specific. Vaginal cultures do not show any recognised pathogen associated with vaginitis. Group B *Streptococcus* is a frequent isolate, but in this context, is not pathogenic. All of these findings are by no means invariable.

In general, we do not biopsy these patients because it is so often non-specific. Histology shows either a non-specific inflammatory response or an interface dermatitis with a heavy mixed inflammatory infiltrate. This pattern is different from that seen in lichen planus.

In our published cases series, 56% had historical triggers, most frequently chronic diarrhea or antibiotic treatment. A total of 54% had no significant abnormality on microbiological testing. Other historical triggers included hormone-replacement therapy (HRT) and recent gynaecological surgery. Other publications cite changes on examination of vaginal wet mounts. Unfortunately, most clinicians possess neither a microscope in their office nor the skills to interpret these.

Figure 5.3 Desquamative inflammatory vaginitis with typical petecchial rash in vaginal introitus

Our diagnostic criteria are as follows:

- history: non-cyclical pain, dyspareunia, itch and discharge
- clinical findings: non-erosive, non-scarring patchy or confluent erythema or petechiae
- exclusion of other causes of chronic vaginitis
- drug and irritant causes excluded by trial of elimination
- microbiology: no pathogens isolated on culture
- treatment response: prompt improvement with intravaginal antibiotic.

Management

Desquamative inflammatory vulvo-vaginitis usually promptly responds to intravaginal antibiotics including clindamycin, mupirocin and metronidazole. Oral antibiotics are ineffective.

We most commonly use 2% clindamycin vaginal cream. Most patients tolerate it; however, in some cases, allergy or severe irritation occurs. In these patients, 0.75% metronidazole topical preparations may be substituted. Mupirocin 2% is a third option.

Our regimen is:

- 5 g of clindamycin 2% cream inserted intravaginally at night daily for four weeks
- 1% hydrocortisone ointment applied externally twice daily.

Almost all patients are asymptomatic and normal to examination by two to four weeks of treatment.

Any historical triggers should be modified where possible. If HRT is implicated, this is ceased at the beginning of the four weeks of treatment. After treatment is completed, HRT may be restarted, but at the risk of DIV recurrence.

Desquamative inflammatory vulvo-vaginitis often requires retreatment or maintenance therapy. Retreatment invariably results in rapid response. However, there is a small group of women who had initially presented with a clinical picture of DIV, who subsequently declare themselves as having lichen planus. We conclude from this that early lichen planus may resemble DIV. This may have resulted in confusion in the literature, since neither condition is reliably diagnosable by biopsy.

Aphthous Ulceration and Non-sexually Acquired Genital Ulceration

Aphthous ulceration of the oral mucosa is common, easily recognised and self-limiting. What is not so well known is that aphthae can involve the vulva, where they may cause a chronic, painful condition. These cases are now known as non-sexually acquired genital ulceration (NSAGU). Aphthae are found in all age groups and, although less common in children than adults, do occur before puberty. Patients with genital aphthae frequently have a history of oral aphthae.

Aphthous ulcers are classified as minor or major. Minor aphthous ulcers are small (2–4 mm), superficial and heal quickly, within 7–10 days. Major lesions are large (approx. 10 mm), deep, severely painful and can be very slow to heal, sometimes taking many months.

The aetiology of NSAGU is unknown, but often familial. It is by definition a disease without underlying sinister implications. Aphthous-like genital ulcers are also found in Behçet's disease and Crohn's disease, and very rare auto-inflammatory diseases such as PFAPA syndrome (periodic fever, aphthous stomatitis, adenitis and pharyngitis). However, these diseases have other symptoms that differentiate them from NSAGU.

A sudden onset of severe aphthous ulceration in association with a febrile prodrome has been described in adolescent girls. It has various names including 'Sutton's Ulcer', Lipschutz ulcer and 'Ulcus Vulvae Acutum'. These ulcers are very painful, alarmingly large and sometimes associated with severe oedema of the labia minora. This very acute form may possibly be a reaction to a viral illness, such as Epstein–Barr virus. It is therefore assumed to be unlikely to recur. This differs from NSAGU.

Presentation

The onset is sudden and associated with significant pain. In some cases, pain is so severe that the patient is unable to walk or urinate, and thus often presents to an emergency department, where they are frequently subjected to investigations for sexually transmitted infections. In virginal adolescents in particular, this causes enormous anger from their distraught families.

Typical aphthous ulcers are 'punched out' with a yellow, sloughy base and a red rim (Figure 5.4). They may be located on any part of the mucosa of the introitus and may be single or multiple. Even when large, they usually heal without scarring.

Recurrent lesions tend to be smaller and less severe and the patterns of recurrence may vary considerably. Some patients have cyclical recurrences, while others may have constant ulcers for months, with prolonged remissions in between.

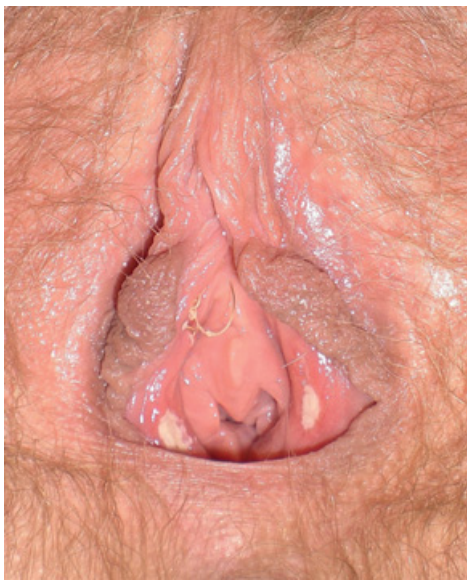

Figure 5.4 Aphthous ulcers showing typical sloughy base

Investigation

The most important part of management is initial, accurate diagnosis. Since there is almost no other differential diagnosis except for herpes simplex virus (HSV), the diagnosis can be made with confidence after HSV polymerase chain reaction (PCR) testing. This will prevent unnecessary additional emotional upset.

Textbooks usually advocate investigating patients for vitamin deficiencies; however, we have never found this to be helpful. Where an adolescent presents with aphthosis associated with fever, Epstein–Barr virus titres are interesting and may explain aetiology, but will not help management.

Management

In the acute stage, the main objective is to relieve pain and facilitate healing.

For minor aphthosis we usually recommend:

- topical very potent corticosteroid every two hours as soon as patients feel the typical pain of an impending attack.

For major aphthosis:

- oral prednisone 0.5 mg/kg/day as a single morning dose daily until the pain has resolved and the ulcer has healed. The prednisone is then withdrawn over the next four weeks
- adequate analgesia
- urination is easier in a warm bath.

For prevention of recurrent attacks:

- doxycycline 50–100 mg/day is often very effective
- nicotine patches may also be useful.

Other treatments that have been advocated include:

- thalidomide
- colchicine
- dapsone.

The use of colchicine, thalidomide or dapsone in general practice should be used only by experts.

Any patient with major or recurrent aphthosis should be reviewed by a specialist.

Crohn's Disease of the Vulva

Crohn's disease may, rarely, involve areas outside the gastrointestinal tract. Many patients with Crohn's vulvitis have a previous diagnosis of Crohn's colitis. Sometimes, however, the vulval involvement may predate the onset of symptomatic colitis by months or even years.

In a patient with known Crohn's colitis who presents with vulvitis, extra-gastrointestinal tract Crohn's must be considered. Furthermore, the gastrointestinal tract disease can be under good control when the vulval disease is not.

Presentation

The hallmarks of Crohn's disease of the vulva are:

- aphthous-like ulceration

- swelling of the labia majora, which may be unilateral or bilateral (see Figure 5.5)
- fissuring
- dyspareunia
- knife-like cuts
- sinuses
- peri-anal erythematous plaques
- peri-anal tags.

Investigation

The diagnosis is made on biopsy, which shows the typical granulomas found in the gastrointestinal tract. If the patient is not known to have Crohn's already, she should be referred to a gastroenterologist for assessment.

Management

Treatment of vulval Crohn's is very challenging. The condition will respond rapidly to oral prednisone, but usually a steroid-sparing agent will be needed before attempting to withdraw the steroid. Referral to a specialist is recommended.

Behçet's Disease

Behçet's disease is a multisystem disease found in certain racial groups, particularly in the Middle East, Asia and Japan. It is rare in women outside of these groups.

Presentation

It is diagnosed according to criteria, which include oral and genital aphthous ulcers, uveitis or retinal vasculitis, and a variety of inflammatory skin lesions. In addition to these criteria, many other symptoms are reported: meningo-encephalitis, synovitis, myocarditis and glomerulonephritis. The pathology of this condition is a vasculitis.

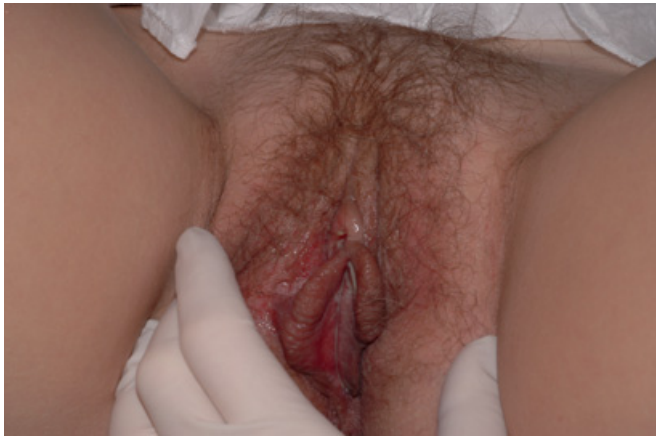

Figure 5.5 Vulval Crohn's disease. This early example shows only bilateral labial oedema

Investigation

Although the genital lesions of Behçet's may be indistinguishable from minor or major aphthosis, in order to make a diagnosis of Behçet's, the patient must have other signs and symptoms, inflammatory bowel disease must be ruled out and a biopsy that shows vasculitis is desirable.

The majority of patients with genital aphthosis do not have Behçet's disease and this need not be entertained unless there are other manifestations.

Management

Immunosuppressant medicine is used to manage the symptoms and prevent complications. Referral to a rheumatologist is recommended.

Autoimmune Bullous Disease of the Vulva

There are two autoimmune bullous diseases that may involve the vulva: bullous pemphigoid, both non-cicatricial and cicatricial, and pemphigus.

Both conditions are very rare. In most cases of pemphigus, it will be obvious that the disease is present elsewhere on the skin and oral mucosa. Cicatricial pemphigoid is often primarily an oral and/or ocular disease, which may also involve the vulva. However, bullous pemphigoid may occur only on the vulva.

Presentation

The hallmark of all these conditions is painful erosions and blisters. It is difficult to differentiate them from each other and from erosive lichen planus without a skin or mucosal biopsy.

Investigation

These conditions usually come into the differential diagnosis of erosive lichen planus, all presenting with vulval erosions. Tissue should always be submitted for histology and also immunofluorescence, which is the diagnostic test.

Management

Treatment is frequently challenging, involving prednisone and immunosuppressive medications. Patients should be referred to a dermatologist.

Cicatricial Pemphigoid

This very rare condition presents with recurrent, painful superficial erosions, which are very slow to heal and do so with scarring. The erosions are usually discrete and few in number. It is a disease of older patients, usually in their mid-60s.

This disease may involve the mouth and the conjunctiva where a similar cicatrising process occurs. In the mouth, it presents with oral erosions of the gingiva, buccal mucosa or palate. It may extend into the oesophagus.

In the genital area, ulceration of the vulva, peri-anal area and rectal mucosa may occur.

Cicatricial pemphigoid is very difficult to treat, and patients should be referred to a dermatologist.

Vulval Bullous Pemphigoid

Bullous pemphigoid is the most common of a group of rare autoimmune bullous dermatoses. It usually occurs in elderly patients, but is well reported in children, in whom it may only occur on the vulva. It does not scar and is usually a self-limiting condition lasting approximately two years.

In adults, vulval involvement is often part of generalised disease and can easily be inferred from this. Localised bullous pemphigoid does occur and a specific subtype occurring only on the vulva is well recognised in both adults and children.

Presentation

The presentation is with blisters and erosions, which involve not only the mucosal surface but also the labia and surrounding skin. The blisters are itchy and once they erode, painful.

Investigation

The clinical diagnosis is confirmed by biopsy and immunofluorescence.

Management

Bullous pemphigoid responds readily to oral and potent topical corticosteroids. Patients should be referred to a dermatologist.

Pemphigus

This very rare disease usually involves skin and oral mucosa with superficial, painful erosions.

Presentation

The initial presentation is often with oral disease; however, genital blistering often follows.

Investigation

Diagnosis requires a skin biopsy submitted for immunofluorescence.

Management

The disease is highly treatment resistant, requiring high-dose oral prednisone and steroid-sparing agents. It would be very unusual to find pemphigus only involving the genital area. Patients should be referred to a dermatologist.

Hailey–Hailey Disease (Benign Familial Pemphigus)

This is a rare dominantly inherited disorder. It is an inherited tendency to fragile skin in certain areas of the body. It frequently involves skin folds and this includes the genital area.

The name benign pemphigus is a misnomer, as this condition is not an immunobullous disease at all. The majority of patients have a positive family history but this is not invariable.

Presentation

The appearance on the genital area is of a non-specific erythematous and eroded eruption involving the labia majora. Super-infection with bacteria and HSV is a recurrent problem in many patients and overheating is an exacerbating factor as it increases epidermal fragility.

Investigation

Hailey–Hailey disease has very characteristic histology, which is diagnostic. The epidermal cells appear disjointed and this is often described as a 'dilapidated brick wall' by histopathologists.

Management

Because there is no cure for this genetic condition, treatment involves palliation with anti-infective agents and topical corticosteroids to reduce inflammation. Keeping cool in summer is important. Some patients improve with systemic retinoids. Patients should be referred to a dermatologist.

Vulval Fixed Drug Eruption

A fixed drug eruption is a rare adverse event related to many oral medications.

On non-genital skin, a fixed drug eruption is a harmless event, which is more of a nuisance. However, on the vulva, it may be much more significant and is almost always very difficult to diagnose. The diagnosis may go unrecognised for many years, causing unnecessary suffering. An *acute* vulval fixed drug eruption is the result of taking drugs that are used intermittently. A *chronic* vulval fixed drug eruption is the result of daily, long-term medications.

Aetiology

Although many drugs may be implicated in vulval fixed drug eruption, the drug classes that are strongly associated are statins, non-steroidal anti-inflammatory drugs, COX-2 inhibitors, pseudoephedrine and paracetamol. This means that the offending drug could be an over-the-counter medicine, which the patient may forget to disclose.

Patients who occasionally take analgesics or nasal decongestants often make the connection between their medication and vulval reaction, but those taking a daily medication frequently do not. Patients who are allergic to ibuprofen and take it for dysmenorrhea often attribute their symptoms to their pads or tampons.

When a patient is on a daily medication, a vulval fixed drug reaction becomes a continuous and chronic. All therapeutic attempts fail to make an impact and the only treatment that will work is cessation of the drug. Paracetamol is a significant cause in elderly patients who rely on it as an analgesic for arthritis. The clue to this diagnosis is the peculiar association of recalcitrant external vulvitis with an erosive mucositis.

Presentation

In a typical *acute non-genital* fixed drug eruption, the rash occurs minutes to hours after ingesting the offending drug. It is usually an erythematous plaque that may or may not blister. The eruption always occurs on exactly the same place every time, lasts about two weeks and usually leaves post-inflammatory hyper-pigmentation.

A chronic fixed drug eruption on the vulva presents differently. There is usually an itchy, bilateral, erythematous and eczematous rash involving the external vulva, peri-anal area and inner thighs. On vulval mucosa it usually causes erosions and therefore pain. It does not cause post-inflammatory hyper-pigmentation, unlike fixed drug eruption on non-genital skin. This non-specific appearance makes vulval fixed drug eruption very easy to miss. In a chronic vulval fixed drug eruption, the link between a daily medication and a chronic vulvitis is usually completely unsuspected.

Investigation

A fixed drug reaction of the skin is a particularly difficult diagnosis for non-dermatologists, as it is rare and slightly bizarre. When it occurs on the vulva, it is even more difficult to diagnose. The histology is non-specific.

The clinical diagnosis is confirmed when rapid improvement occurs within two weeks of stopping the offending drug. Re-challenge is the ultimate diagnostic test.

Management

A drug history including over-the-counter medications, even vitamins, is essential. If vulval fixed drug reaction is suspected, all non-essential drugs should be ceased. It takes only two weeks for the patient to notice improvement in their symptoms.

It should be remembered that some patients react to more than one drug, and that changing the offending drug to a related one may cause the same reaction (i.e., statins may cross react).

If you are unsure which drug has been implicated, a rechallenge will quickly reproduce the rash. With forgetful, elderly patients on paracetamol, this happens all too often. We usually recommend that they wear a drug alert bracelet.

Other Ulcerative Vulval Drug Eruptions

Unless a drug causes a fixed drug reaction, it is unusual to see specific ulceration of the vulva from a medication. Some chemotherapeutic agents, such as methotrexate and the anti-viral agent foscarnet are specifically associated with genital ulceration.

The new immunotherapy drugs for some cancers may cause genital mucosal erosions. The anti-CD20 monoclonal antibody rituximab, used for the treatment of B-cell non-Hodgkin's lymphoma, has been associated with severe genital ulceration, simulating pyoderma gangrenosum.

Erythema Multiforme (Stevens–Johnson Syndrome) and Toxic Epidermal Necrolysis

These two conditions, which are frequently mentioned together, are in fact quite different. Erythema multiforme is a benign condition of skin and mucosal surfaces, which, although it can cause significant morbidity, is not life threatening. It is usually

precipitated by infections with either HSV or *Mycoplasma pneumonia*, although drug eruptions may occasionally be involved. Very occasionally, it may occur only on the mucosa of the mouth and vagina. It mostly affects young adults.

Toxic epidermal necrolysis, by contrast, is a severe, life-threatening drug eruption where skin erosion is associated with significant multisystem disease. It is invariably a drug eruption, classically from anti-epileptic and sulpha drugs. Mucosal surfaces are always involved, but external skin is often affected as well. Genital involvement is often overlooked because the patient is so ill.

Presentation of Erythema Multiforme

Erythema multiforme presents with a sudden onset of skin lesions, which are classically described as 'target lesions' as they have a round to ovoid appearance with central clearing or blistering. When mucosal surfaces are involved, the presentation is with painful erosions.

The precipitating event is usually either an attack of HSV, which may have been subclinical, or a *Mycoplasma* infection, which may have only evidenced itself with a cough or apparent upper respiratory tract infection. When erythema multiforme is caused by HSV, it may be recurrent, occurring after every cold sore.

The diagnosis is usually made clinically, although there is a characteristic biopsy appearance. When vulval erosions are part of a larger clinical picture, a vulval biopsy is not necessary.

Recurrent erythema multiforme of the genital mucosa is usually accompanied by erosive disease in the mouth, or even the eyes. The presentation is usually with intermittent and recurrent attacks of simultaneous oral and genital ulceration lasting about two to three weeks. In between attacks, the patient is normal. This is very different from lichen planus, which is chronic but which it can mimic during an acute attack.

This confusing presentation requires a biopsy to make the diagnosis. Histology is characteristic.

Management

The treatment of these conditions still remains an area of controversy, but most experts agree that corticosteroids are of value only if started promptly at the onset of symptoms. They are of little value in established disease.

Toxic epidermal necrolysis invariably requires admission to hospital, usually to an intensive care unit.

Diagnosis and treatment of underlying infection that may have precipitated erythema multiforme is essential but treatment of the rash itself is supportive only. Swabs for HSV and *Mycoplasma* titres should be done and a macrolide antibiotic as well as an anti-viral agent should be commenced. Any drugs that may have been responsible should be ceased.

In both of these conditions, vaginal erosion may be severe and may be followed by adhesions, which may occlude the vagina. During the acute attack, intravaginal corticosteroid pessaries should be inserted daily, and as the vagina heals, topical steroids should be used with dilators daily to prevent adhesions.

Herpes Simplex

Herpes simplex virus (HSV types 1 and 2) is the most common infective condition that causes vulval erosions and ulcerations. HSV 2 used to be responsible for most cases of genital herpes, but this is changing because of the tendency for young people to engage in oral sex.

Unlike lip cold sores, which are usually acquired innocently during childhood, HSV of the vulva is a sexually transmissible infection. It is therefore rarely seen in children. When it does occur in children, however, herpes may not always be sexually acquired, particularly where the child has an underlying skin disorder such as eczema. The occurrence of genital herpes in a child with no predisposing factors does, however, raise the question of child sexual abuse.

Herpes may complicate vulval skin conditions such as lichen planus and lichen sclerosus, particularly when under treatment with potent topical corticosteroid or during episodes of immunosuppression.

Presentation

The first, or primary, attack is always the most severe. Painful blisters are found on any part of the genital and peri-anal skin and mucosa.

They range in size from about 3–10 mm and rapidly erode leaving ulcers (see Figure 5.6). Lesions may extend to the inner thighs and buttocks and may be unilateral. Lesions may be numerous or few in number. Lymphadenopathy is present and patients may be systemically unwell. Untreated, the attack usually lasts two weeks.

Many patients only ever experience one attack of genital herpes, but it may recur at any stage of life with greatly varying frequency. In otherwise healthy patients, the cause of this variation is not known. Patients with HIV disease may experience intractable HSV genital ulceration.

Investigation

Diagnosis of HSV is made by viral PCR, sampled by swabbing a new lesion. This technique is very sensitive and differentiates HSV 1 and 2. Biopsy of a blister will also diagnose HSV but this is usually not necessary.

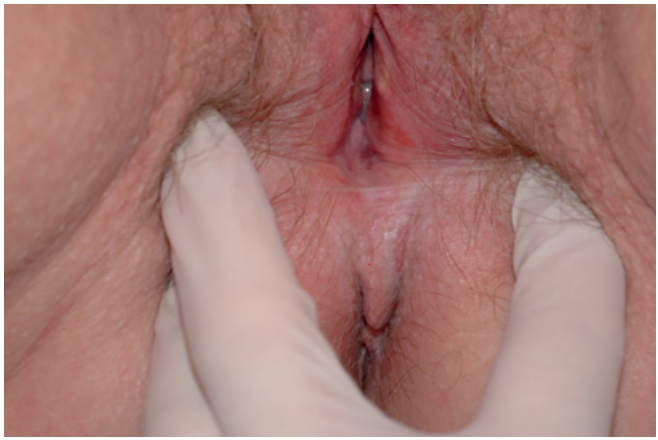

Figure 5.6 A posterior introital fissure in seborrhoeic dermatitis

Herpes serology is not usually helpful in the diagnosis of genital herpes. Positive IgG titres indicate past exposure only and therefore do not help to determine whether the current lesion is herpes or not. Rising herpes IgM titres are of value if started soon enough, but are not routinely required.

Management

Genital herpes is treated with anti-viral agents: acyclovir, valacyclovir and amciclovir. The two latter agents have virtually superseded acyclovir because of ease of dosing; however, all are similar in terms of efficacy. The symptoms and duration of acute attacks are reduced by these medications.

For recurrent herpes, all anti-viral agents may be used intermittently or continuously depending on the frequency of attacks. These medications are well tolerated and, long term, have an excellent safety record.

A diagnosis of genital herpes is emotionally devastating for most women, not only because of the knowledge that it is a sexually transmissible infection but because of the long-term, uncertain prognosis.

Although it is not unreasonable to commence anti-viral treatment on suspicion of genital herpes, it is equally important to confirm or deny the diagnosis. There are many erosive conditions of the vulva and not every blistering condition is genital herpes. We have seen many patients suffer for years because of a belief that they have genital herpes based on positive serology alone or simply on clinical impression that has never been confirmed. In particular, patients with recurrent aphthosis are often told that they have genital herpes and we have seen patients with this condition not only treated for long periods of time with anti-virals without improvement, but also under the false impression that they have contracted an sexually transmissible infection.

Other Infections that May Cause Ulceration

In Western countries, persistent vulval ulcers are much more likely to be non-infective than infective. However, the following infections may cause vulval ulceration:

- syphilis
- chancroid
- lymphogranuloma venereum
- granuloma inguinale
- leishmaniasis
- amoebiasis.

With the exception of syphilis, all of these conditions are tropical diseases and the patient will have a history of living in or travel to an endemic area. Chancroid, lymphogranuloma venereum and granuloma inguinale are usually sexually transmitted. A detailed description is beyond the scope of this book.

Any persistent vulval ulcer must be biopsied in the first instance and further investigations, including culture, PCR and serology, frequently depend on the result of that biopsy. When sending the specimen for histopathology, alert the histopathologist to the possibility of chronic infection. Referral to a sexual health or infectious diseases specialist is recommended.

Traumatic Fissures

Minor tears and splits are common on the vulva, particularly at six o'clock on the introitus, and usually arise from friction during intercourse and sporting activities, particularly bicycle riding. They heal rapidly in most cases.

Traumatic fissures that are recurrent, frequent and slow to heal may occur in three situations:

- where there is an underlying uncontrolled skin condition (see Figure 5.7)
- where there is oestrogen deficiency (menopause, lactation)
- when it is idiopathic.

It is the idiopathic group that presents the most difficult management problem. This group of patients presents with recurrent, slow-to-heal splits on the mucosal surface of the labia minora following intercourse. The splits are often on the same area each time that they occur. These patients have no underlying skin problem and are not oestrogen deficient. As a result, topical oestrogen does not help them. A careful history should be taken, seeking any activities that might cause excessive vulval friction. It is possible for very frequent sexual intercourse to cause vulval or vaginal skin damage, even in the absence of skin disease. Sometimes we need to advise women that they can no longer sustain very frequent intercourse.

Our approach to these patients is to trial topical testosterone, prescribed as 2% testosterone in white soft paraffin. It is initially applied only to the area that splits every day for two weeks, then twice a week. If this strategy is unsuccessful, excision of the area that splits recurrently is the next step and frequently ends the problem.

Conclusion: Approach to a Patient with Vulval Ulceration or Erosions

History must include a sexual and travel history, search for possible triggers or any oral or gastrointestinal tract disease.

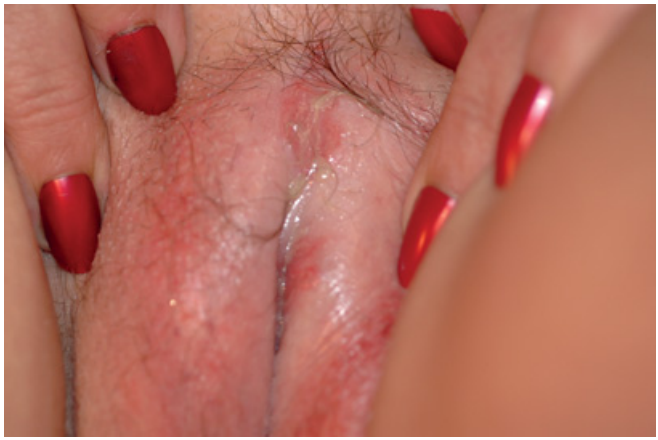

Figure 5.7 Acute genital herpes

A very careful examination must be made, and if possible, include a speculum examination of the entire vagina, although this may be very difficult because of pain. Look carefully to distinguish between an ulcer, erosion and a fissure. Note whether the rash extends onto external vulval skin, and if it extends further into the vagina than the introitus. A low vaginal swab should exclude infections.

It is important to remember what is common and what is very rare. This will make it easier to arrive at a working diagnosis.

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Chapter 6

Persistent Vaginitis

Contents

This chapter discusses persistent vaginitis that is not infective in aetiology. This is not only distressing for patients, but diagnostically challenging because the available tests are often unhelpful.

What Is Persistent Vaginitis?

Vaginitis is inflammation of the vaginal epithelium evidenced by erythema, discharge or other changes such as erosions. Infection is the most common cause but vaginitis can be non-infective, and there is usually a defined aetiology. Persistent vaginitis is when this symptomatic inflammation either recurs after, or is resistant to, initial treatment. It may occur in association with a vulval or perianal dermatosis.

Persistent vaginitis is uncommon and often perplexing. For the patient, it can be the cause of enormous misery, anxiety, sexual guilt and even relationship breakdown. Historically, it has three patterns: recurring attacks, chronic unremitting symptoms or symptoms that are chronic but exacerbate at certain times of the menstrual cycle. A previous systematic review has demonstrated that, taken individually, symptoms, signs and tests are poor predictors of the cause of vaginitis.

Does the Complaint of Persistent Vaginitis Indicate Pathology?

Our own differential diagnosis of persistent vulvo-vaginitis is not long (Table 6.1). The list is based on our published observations and is listed in the order of the prevalence that we think may occur in general practice.

Some of the less common conditions that may be unfamiliar to general clinicians include:

- chronic vulvo-vaginal candidiasis
- recurrent bacterial vaginosis.
- type 1 hypersensitivity responses
- intravaginal foreign body such as a retained tampon. This normally presents with a heavy discharge and or recurrent infection

Table 6.1 Differential diagnosis of chronic vulvo-vaginitis

Common
• Recurrent vulvo-vaginal candidiasis: defined as four attacks a year; however, some patients' candidiasis is more indicative of a chronic continuous process
• Recurrent bacterial vaginosis
• Contact dermatitis (allergic and irritant contact): caused by intra-vaginal pessaries and creams (usually obvious on history)
Uncommon
• Desquamative inflammatory vaginitis: an uncommon, non-infective, painful vaginitis of unknown cause characterised by shiny erythematous patches and/or petechiae
• Intra-vaginal foreign body (e.g., retained tampon): a cause of persistent vaginitis with a heavy discharge
• Chronic fixed drug eruption: an erosive vulvo-vaginitis most often associated with non-steroidal anti-inflammatory drugs and statins
• Type 1 hypersensitivity reactions: itch, burning, swelling and even anaphylaxis can result from exposure to latex condoms and seminal fluid
Rare
• Mucosal lichen planus: a skin disease that often involves the oral as well as the vaginal mucosa with very painful erosions that may eventually lead to scarring
• Oestrogen hypersensitivity vulvovaginitis: a cyclical vulvitis with a presentation closely similar to that of recurrent candidiasis but not causally associated with candida
Very rare
• Crohn's disease: a cause of vulvovaginitis
• Immunobullous disease: erosive vaginal involvement without generalised skin disease may occur, particularly in cicatricial pemphigoid
• Graft versus host disease: a vulvo-vaginitis indistinguishable from lichen planus

- chronic fixed drug eruption
- desquamative inflammatory vaginitis
- lichen planus
- oestrogen hypersensitivity vulvitis
- Crohn's disease is a rare manifestation of vulvo-vaginitis
- graft versus host disease can present exactly like vaginal lichen planus
- immunobullous diseases particularly mucosal pemphigoid. This is very rare.

Even though chronic vulvo-vaginal candidiasis and bacterial vaginosis are related to specific micro-organisms, they are not infective in nature but an immunological reaction to an organism.

With the exceptions of lichen planus, immunobullous disease and Crohn's disease, which have defined histopathology, none are diagnosable by biopsy. Since these diseases are very different in aetiology, an accurate diagnosis is therefore essential for rational management.

History

A detailed and specific history is essential.

Define the symptoms:

- itch, soreness, burning
- discharge, swelling

- superficial dyspareunia, skin splitting
- sudden or insidious onset
- duration
- whether continuous or recurrent
- whether there is a relationship to the menstrual cycle.

Historical triggers can be critical to the diagnosis, especially for contact dermatitis, type 1 hypersensitivity reactions, desquamative inflammatory vaginitis and fixed drug eruptions. Antibiotics use, especially with vaginal surgery, may trigger candidiasis and desquamative inflammatory vaginitis. Events that exacerbate symptoms are also useful, for example, the tendency of candidiasis to exacerbate in the pre-menstrual phase of the menstrual cycle.

Ask specifically about:

- medications including over-the-counter medications and whether the vaginitis occurred before they were commenced
- latex condoms
- relationship to contact with semen
- topically applied substances, lubricants, pessaries and devices
- presence of oral lesions that might indicate lichen planus
- previous results of swabs
- previous response to treatment.

Examination

Even though we are discussing vaginitis, there are several diagnoses that may either extend onto the external genital skin (e.g., candidiasis, fixed drug eruptions, lichen planus) or precipitate a reactive external dermatosis such as psoriasis.

External Examination

- Inspect the labia minora and majora for erythema, oedema and scale, and note whether there is an accentuation of erythema in the inter-labial sulcus, which may indicate chronic candidiasis. Loss of the labia minora, and introital fusion, may be associated with lichen planus.
- Look for fissures on the introitus, perineum or peri-anal skin, often seen in chronic candidiasis.
- Inspect the mucosal surface of the introitus: look for swollen, confluent erythema, or the petechial lesions typical of desquamative inflammatory vulvo-vaginitis.
- On the mucosal surface, look for erosions that might indicate a fixed drug eruption or erosive lichen planus.

Speculum Examination (If Possible)

- Note if the inflammation is confluent or patchy: patchy inflammation (especially with petechiae) is more typical of desquamative inflammatory vaginitis or lichen planus. Be aware that a vaginitis may only occur in the upper vagina.
- Note the type of discharge: chronic candidiasis may not produce the ‘cheesy’ discharge so typical of its acute counterpart. A green discharge may indicate desquamative inflammatory vaginitis.

- Look carefully for erosions, ulcers, adhesions or scarring.
- Make sure that there are no foreign bodies, either intravaginal or under the clitoral hood.

Investigations and Further Management

Appropriate tests should always be performed to exclude infective aetiologies, especially if this has not been performed recently. However, negative results should not preclude a trial of anti-infective medication if the history and clinical examination are consistent with any of these.

Biopsy from inflamed vaginal mucosa often triggers brisk bleeding, so this should be attempted only by clinicians with the necessary experience. There are no other reliable and valid tests for recurrent or chronic vaginitis and trials of therapy are often the only option.

Having excluded infections and bacterial vaginosis, it is usually possible to make a short list of likely diagnoses. It may be necessary to rule out chronic vulvo-vaginal candidiasis by a trial of oral azole therapy (see Chapter 3).

Contact dermatitis, which is a type IV hypersensitivity response, resolves after cessation of the offending item and can be confirmed by patch testing. If the patient has an erosive picture and is taking a drug that has been implicated in vulval fixed drug eruption, the next step is to stop the drug(s). A rapid improvement will occur within two weeks. Rechallenge will confirm the diagnosis in one or two days.

A non-erosive vaginitis extending only to the labia minora, particularly if patchy or petechial, may represent desquamative inflammatory vaginitis. This is a clinical diagnosis confirmed by exclusion of other causes, and by an adequate response to therapy. A two- to six-week trial of intravaginal clindamycin 2% cream with 1% hydrocortisone used externally will give an adequate clinical response.

If an erosive picture has not responded to stopping an offending drug (or if there are no suspect drugs), lichen planus or immunobullous disease should be suspected, and referral to a specialist arranged.

A vulval biopsy from near (not on) the edge of an erosion is indicated to rule out lichen planus. Note that this is highly specific but not sensitive. A negative biopsy in the presence of a strong suspicion of lichen planus warrants a trial of therapy with a potent topical corticosteroid or a short course of oral prednisone at a dose of 0.25 mg/kg/day. A good response is usually seen within four to six weeks.

The Patient with a Persistent Non-offensive Vaginal Discharge but No Other Symptoms

This clinical presentation can be one of the most difficult management issues in vulvology. The patient is often very young, and is distressed by what she perceives as an 'abnormal' discharge. She may less commonly describe an offensive vaginal odour, of which she only is aware, or which results from constantly wearing liners. Questioning reveals that the discharge has a physiological cyclical variation, is inoffensive and or normal consistency. When examination and relevant tests are normal, it can be very difficult to offer adequate reassurance.

It requires great clinical experience to be able to stop further tests and speculative treatments and encourage acceptance that the discharge is in fact physiological.

We recommend specialist referral for any perplexing vaginitis case: it is imperative that these women receive timely and effective help in order to prevent escalation of their distress. The one proviso is to make sure that the skin of the vulva is also examined. Surface desquamation, which is commonly seen in psoriasis, can be mistaken for vaginal discharge.

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Chapter 7

Lumps: Benign and Malignant[1]

Contents

The vulva is part of the skin, therefore, many common lesions found on the skin are also found on the vulva. Some of these lesions can be found anywhere; others are very specific to female genital skin.

This is also true of malignancy: most malignant lesions of the vulva are skin cancers. However, when skin cancer occurs on the vulva, it may have a more serious prognosis than equivalent lesions found on the rest of the skin. Extra-mammary Paget's disease is a specific vulval condition.

Normal Variants

- Melanocytic naevi
- epidermal naevi
- vulval papillomatosis
- prominent sebaceous glands
- melanosis vulvae

[1] We would like to thank Dr Greg Gard MB, BS, FRANZCOG, CGO, of Royal North Shore Hospital in Sydney, for his help in writing this chapter.

- hyper-pigmentation
- asymmetrical labia
- hymenal remnants.

Benign Lesions

- Warts
- molluscum contagiosum
- angiokeratomas
- syringomas
- Fox–Fordyce disease
- hidradenitis suppurativa
- seborrhoeic keratoses
- sebaceous cysts
- fibrous polyps
- hidradenoma papilliferum.

Malignant Lesions

- Vulval intraepithelial neoplasia (VIN; squamous cell carcinoma in situ)
- melanoma
- extra-mammary Paget's disease
- basal cell carcinoma

Normal Variants

The vulva is one of the most anatomically varied parts of the female body. This is due, not just to inherent differences, but also to obstetric trauma, obesity, pelvic organ prolapse and sexual intercourse.

The hymenal remnants can be very variable in size and appearance, and frequently cause concern. Patients (and sometimes doctors) may mistake them for abnormal lesions.

Vulval architecture is virtually always asymmetrical. As with any other part of the body, some normal vulvas display quite marked asymmetry.

We see more and more young women presenting with concerns about whether their vulvas are 'normal' or not. Usually this relates to the size or asymmetry of the labia minora. In almost every instance, the findings are well within normal limits, but the current media focus on vulval cosmetic surgery, particularly labiaplasty, has fueled many women's anxieties. Rarely, a very large unilateral hypertrophy may require surgical excision.

Warts and Other Lesions

External Genital Warts

Human papillomavirus (HPV) is the most common sexually transmissible infection (STI) and is known to cause ano-genital warts and also ano-genital malignancy. There are over 200 HPV genotypes, but most genital warts are caused by the non-oncogenic types 6 and 11. The risk of a wart being potentially neoplastic is therefore small but not zero.

The prevalence of genital warts in the adult population is about 1–2%, but many more carry HPV DNA on the external genitalia and lower genital tract. Figures range from 10 to 80% and are highly age dependent. As a result of this, people with no obvious lesions can still harbor latent virus in their ano-genital epithelium and transmit it to others.

In immune-competent patients, cell-mediated immunity controls latent infection and is responsible for regression of lesions; however, in immune-compromised patients, HPV-related lesions can be persistent and are more likely to be infected with high-risk HPV infections, particularly HPV 16, which may progress to neoplasia. This includes organ transplant patients and patients with HIV/AIDS.

How Are Genital Warts Acquired?

In adults and teenagers, genital warts are usually acquired by skin-to-skin contact during intercourse. The virus is highly transmissible, with up to 85% of sexual partners of patients with warts subsequently developing lesions within six weeks to eight months. The incubation period is around three months. There may be a period of latency where HPV DNA is present but sub-clinical. Re-activation with clinical warts may occur many years after initial infection. The use of condoms does not completely protect against the transmission of genital warts. A diagnosis of benign genital warts does not exclude the possibility of potentially oncogenic HPV carriage in other parts of the lower genital tract.

In children, however, a sexually transmissible aetiology is highly controversial, and published reports claim that there are many other ways for small children to acquire genital warts. This includes auto-inoculation, innocent transmission from other family members and vertical transmission at birth from an HPV-infected mother. There is very little logic in a medical belief system that immediately attributes genital warts in adults to sexual transmission but denies this in children.

When it comes to genital warts in children, there would seem to be a high degree of denial in the community about how they might have been acquired. The fact is, however, that many retrospective studies show that at least 1 in 5 women and 1 in 10 men can recall sexual abuse in childhood, a sobering statistic that should make us all think critically on the subject.

Despite the current media focus on child sexual abuse within institutions, child sexual abuse within the immediate family is a well-kept secret and disclosure is a rare event. With a medical literature that throws so much doubt on the source of genital warts in small children, the best way to manage a child with genital warts becomes a significant dilemma for which there is no straightforward answer.

Presentation

Typical genital warts are raised, papillomatous, often slightly pointed lesions with a rugose surface (see Figure 7.1). The warts are localised predominantly on the vulva, the vaginal introitus and peri-anal skin but can extend into the anal canal. It is not necessary to use a colposcope or acetic acid application to see genital warts. In general, they are asymptomatic; however, when traumatised, they may split or bleed, causing pain and anxiety (see Figure 7.2).

Warts vary greatly in shape and size. They may be tiny and multiple, cauliflower-shaped, dome-shaped skin-colored papules or flat-topped papules. The colour is usually skin-colored to pink to brown and the surface dull rather than shiny. Morphology does not

Figure 7.1 Genital warts

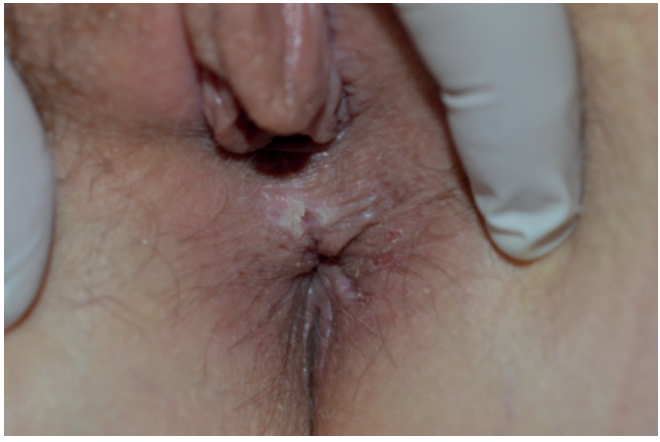

Figure 7.2 Perineal genital warts with central fissure

correlate with HPV type. Warts may be very large and numerous, particularly in the perianal area. HPV infection may also present as a fissured, painful perineal plaque that can simulate a malignancy.

HPV Vaccination

The most commonly used HPV vaccine targets HPV types 6, 11, 16 and 18. It covers the two most common genotypes of benign external genital warts and the two most common genotypes associated with lower genital tract cancer. New vaccines that immunise against more genotypes are now available.

Vaccination has demonstrated high-level protection against cervical dysplasia for at least 3.5 years after immunisation in adolescents aged 16–23 years and recent data demonstrate that it has also significantly reduced the incidence of genital warts. It promises also to prevent most cases of cervical carcinoma and VIN. It must be stressed, however, that this vaccine does not stop the need for routine cancer surveillance on the cervix.

Differential Diagnosis

Because HPV is a STI carrying a great degree of stigma for many patients, it is very important to be able to differentiate external genital warts from other similar lesions. If in doubt, a biopsy will usually provide the answer as warts have a typical histopathological appearance.

The main differential diagnoses are:

- seborrhoeic keratoses
- molluscum contagiosum
- VIN
- bowenoid papulosis, an unusual variant of external genital warts. In this condition, multiple domed or flat, hyper-pigmented papules are found on the vulva and perianal area. When biopsied, there is high-grade intraepithelial neoplasia, similar to VIN.

Management

The natural history of genital warts is to regress spontaneously within one to two years. However, in some patients, HPV can be very persistent.

Treatment of asymptomatic warts is often cosmetic, and they often recur after a single course of treatment. Further, there is no evidence that any treatment can change the natural history or reduce infectivity. We therefore point out to patients that observation is an appropriate treatment option.

A presentation with genital warts is an ideal opportunity to organise routine gynaecological screening and counselling on safe sex practices, especially for adolescents. A cervical screening test (for the presence of HPV, and/or cytology, depending on national guidelines) should always be performed at presentation. However, it should be pointed out that the cutaneous HPV infection that caused the warts might also cause a reversible low-grade Pap smear abnormality. Not all women with genital warts require a full STI screen. The clinician should use their judgement in this regard.

There is no definitive first-line treatment. All treatments aim to remove visible lesions, but patients often require a course of therapy rather than a single treatment. In general, warts located on moist surfaces and/or in intertriginous areas respond better to topical treatment than do warts on drier surfaces.

Treatments that Can Be Administered by the Patient

Many patients express a preference for applying treatment themselves. In this situation, make sure that they have been adequately counseled about how to properly and safely apply it, and warn about the potential for adverse local reactions. Make sure to warn patients not to apply the substances to normal skin. All treatments have a significant failure and recurrence rate and sometimes a combination of patient-applied treatment combined with in office treatment can be more effective than either alone.

1. Podophyllotoxin, also called podofilox and podophyllin, has been shown to be safe and effective. Patients may apply the solution with a cotton swab, or gel with a finger, to visible genital warts twice a day for three days, followed by four days of no therapy. After application, it is allowed to air dry and there is no need to wash it off. This cycle may be repeated for a total of four cycles. The total wart area that is treated should not exceed 10 cm^2, and a total volume of podophyllotoxin should not exceed 0.5 ml. The safety of podophyllotoxin during pregnancy has not been established.
2. Imiquimod 5 % cream (Aldara® cream) is a topically active immune enhancer that stimulates production of interferon and other cytokines. Patients should apply imiquimod cream with a finger at bedtime, three times a week for as long as 16 weeks. The treatment area should be washed with mild soap and water 6–10 hours after the application. This preparation always causes an inflammatory response on treated skin, and care should be taken to avoid excessive inflammation. It therefore may not be appropriate to use imiquimod on introital skin, especially in patients predisposed to atopic skin disease. Many patients will be clear of warts by 8–10 weeks, or even sooner. The safety of imiquimod during pregnancy has not been established.
3. Green tea sinecatechins (polyphenon E ointment) is approved for use in patients over 18 years of age for external genital and peri-anal warts. It has been shown to be safe and reasonably effective. It is applied three times daily, leaving a thin layer on the warts. It is not necessary to wash it off.
4. Both ingenol mebutate 0.015% gel and fluorouracil 5% cream are topical therapies used for actinic keratosis treatment. They have, anecdotally, been reported to be useful in eradicating genital warts. They should be prescribed only by a dermatologist.
5. Cidofovir is an anti-viral drug with activity against HPV. It is used topically by the patient as a 1% cream or gel, or intra-lesionally by the doctor. This use is off-label and it is expensive. There is a growing body of evidence that this treatment is useful for ano-genital warts in adults and children and it may be useful in future.

Treatments that Are Administered by a Doctor

1. Cryotherapy with liquid nitrogen. This is suitable only for small lesions. Keep the lesion frozen for 30 s, allow the wart to thaw and then repeat.
2. Surgical removal either by tangential scissors excision, tangential shave excision, curettage or electrosurgery. Surgical removal is especially helpful in the peri-anal area, and for large or numerous lesions. In children, a general anaesthetic is usually required.
3. CO_2 laser ablation, as an alternative to excision. The operator should use appropriate protection against aerosolised HPV.

External Genital Warts in Pregnancy

Genital warts can become much more severe in pregnant women but usually improve after the baby is born. Many experts advocate their removal at this time. HPV types 6 and 11 can cause laryngeal papillomatosis among infants and the route of transmission (transplacental, birth canal or postnatal) is not completely understood. It is not known whether Caesarean section might prevent transmission. Topical treatments are contra-indicated in pregnancy, except for cryotherapy.

Follow-up

Patients should be cautioned to watch for recurrences, which occur most frequently during the first three months.

Treatment of Sex Partners

Examination of sex partners is not necessary for management of genital warts because the role of reinfection is probably minimal. Therefore, treatment to reduce transmission is not necessary.

Psychological Issues

The emotional impact of genital warts is huge. Many patients say that they feel 'dirty' and this is usually why they seek treatment for a condition that is asymptomatic. The discovery of warts for a woman in a monogamous relationship may place great stress on the relationship because of the unknown incubation period. Couples may need counseling to come to terms with the diagnosis.

Molluscum Contagiosum

Molluscum contagiosum is a viral infection that is very common in children but not in adults. In adults, it is usually sexually acquired, but in children, is usually acquired from swimming pools and siblings with whom they share a bath. Auto-inoculation is also an important method of transmission. In children, genital mollusca are usually part of a more widespread infection of other parts of the skin.

There are four viral genotypes. MCV-1 is the type usually found in children, while MCV-2 is found in adults due to sexual transmission.

Presentation

Incubation is two weeks to several months. Lesions are seen as umbilicated flesh-coloured papules of 2–5 mm with a pearly appearance (see Figure 7.3). Giant mollusca can occur on the genital region, and occasionally they may become inflamed and superinfected. Very small lesions often lack typical umbilication.

Genital mollusca are usually asymptomatic, but itch and a secondary dermatitis may occur, particularly in atopic children. Mollusca can be severe in immunosuppressed patients, but in a healthy patient, severe mollusca is not necessarily a sign of any other disease.

Mollusca are self-limiting within six months to two years in immunocompetent patients, but may have a prolonged course in immunosuppressed patients. After they resolve, they are unlikely to recur.

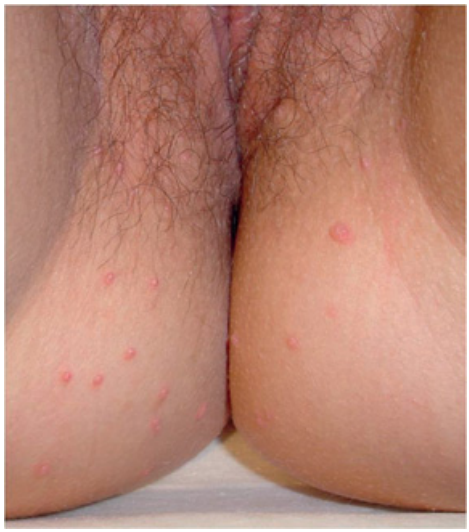

Figure 7.3 Molluscum contagiosum. Note the typical central umbilication

Investigation

Diagnosis is usually made clinically; however, in the genital area, it may occasionally be difficult to differentiate small mollusca without typical umbilication from genital warts. If there is any doubt, the diagnosis can be confirmed by microscopy performed on material from a lesion obtained by extruding the central core with pressure, a small curette or a simple shave biopsy. Histopathology is highly characteristic.

Management

Treatment is easy in adults, but more difficult in children. Avoidance of baths and swimming pools may reduce inoculation. If pruritus is a problem, topical corticosteroids are helpful, however, topical immunomodulators should be avoided as they may increase the severity of infection. In children, it may be best to allow them to resolve spontaneously, unless they are distressing. In adults, the best course of action is physical removal.

Mollusca lesions have a small round core containing viral particles. If this is removed or damaged, the lesions resolve quickly.

Methods that work for removing mollusca:

- scrape them off gently with a small skin curette
- squeeze the lesions to remove the core
- prick with a needle to flick out the core
- light freeze with a cryotherapy unit.

We do not recommend imiquimod because it is expensive, irritating and not completely reliable.

Cantharidin is a topical substance that causes blistering. It can be useful in mollusca in children, however, use on the genital area would be very irritating and better avoided.

Non-infective Lesions that May Be Confused with External Genital Warts

Seborrhoeic Keratoses

These very common, benign skin lesions are common in people over the age of 40. They can be found on any part of the skin, including the vulva. They do not occur in the vagina.

Presentation

Seborrhoeic keratoses have very varied shapes, sizes and colours. They may be flat, raised, pedunculated and often closely resemble warts (see Figure 7.4). Colours range from skin tone to black. They may thus be confused with melanoma. They become more numerous with age.

These lesions are usually not symptomatic but, if large, may rub on clothing and become irritating. If they are scratched or traumatised, they may suddenly enlarge and darken.

Investigation

The main significance of seborrhoeic keratoses on the vulva is their resemblance to genital warts and sometimes to vulval squamous cell carcinoma in situ (VIN). However, they are easily differentiated on biopsy as they have a typical histological appearance.

Dermoscopy is useful for seborrhoeic keratoses on non-genital skin, but is awkward to perform on the vulva.

Management

Treatment can be successfully achieved with:

- cryotherapy
- light curettage and cautery
- excision of large lesions.

Recurrences may occur, but are less common than with external genital warts.

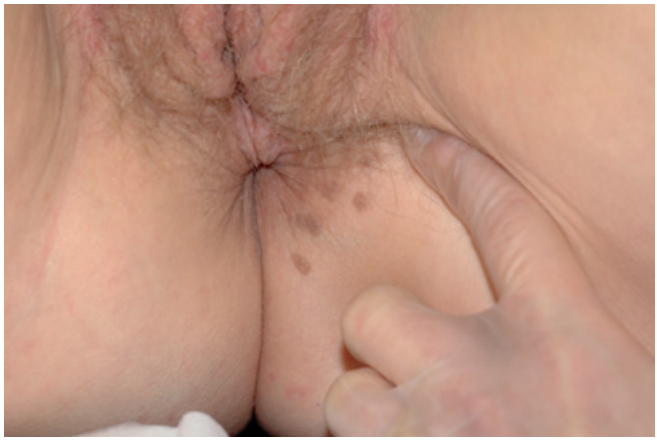

Figure 7.4 Seborrhoeic keratoses

Skin Tags (Fibromas)

These harmless lesions are common in the major flexures. They are most often found in the axillae, neck and the inguinal folds.

Skin tags usually form in middle age and can occur during pregnancy. The appearance of multiple skin tags in young people is unusual and should prompt a search for signs of tuberous sclerosus, neurofibromatosis or, if peri-anal, Crohn's disease.

Skin tags are small, soft, pedunculated lesions, flesh-coloured to brown in colour. They often accompany localised keratoses. The base of the peduncle is rarely wider than 1–2 mm.

Management

Patients may request removal because they rub on clothing. The easiest treatment is scissor amputation at the base of the peduncle. If this is done quickly, no local anaesthetic is required unless they are quite large. Bleeding can be stopped with pressure. Any destructive method will work: hyfrecation or cryotherapy is also effective.

Vulval Papillomatosis

These lesions are analogous to pearly penile papules found in males. They are normal anatomical variants but may often be confused with warts. Indeed, in the past, the medical literature has suggested that vulval papillomatosis is an HPV-related condition. This is now known to be incorrect.

Presentation

Multiple asymptomatic small papillae are seen on the mucosal surface of the labia minora in the vestibule. They are flesh-coloured with a smooth rounded surface unlike the dull, rough and often pointed shape of genital warts (see Figure 7.5).

The main differentiating factor from warts is the uniformity of the papillae and the symmetrical distribution.

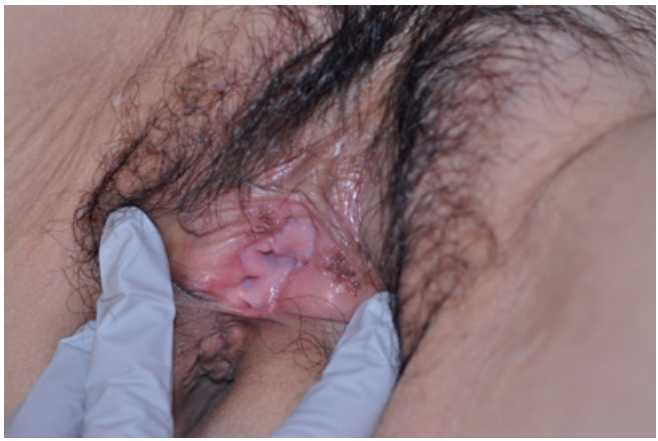

Figure 7.5 Vulval papillomatosis

Investigation

If there is any doubt, a small biopsy will differentiate them from genital warts.

Prominent Sebaceous Glands

The inner surface of the labia minora contains numerous sebaceous glands. This is normal and does not cause any symptoms.

Sebaceous glands are sometimes visible and palpable. If a patient examines herself and notices these glands, she may become alarmed and require reassurance. When this occurs, they are often described as hyper-plastic. Hyper-plastic sebaceous glands appear as small, discrete slightly yellowish flat papules (see Figure 7.6). This term is confusing because these lesions do not represent a disease state. To distinguish this from an abnormality, it is more appropriate to call them prominent sebaceous glands.

Fox–Fordyce Disease

This is a benign and very rare condition of unknown aetiology. The onset of this condition is usually in adolescence to young adulthood; however, there have been cases reported in children.

The pathology of this condition has been described as inflammation of the hair follicle, but histopathology is essentially non-specific. It may represent a disorder of apocrine glands because they occur only on genital, axillary and areolar skin. The glands are connected to the hair follicle in these areas and secrete an oily fluid, which is different to sweat.

Presentation

Multiple 2–4 mm itchy, dome-shaped papules occur bilaterally on the vulva and pubic area.

Management

Treatment is difficult. Many strategies and topical therapies have been suggested including topical retinoids, corticosteroids, antibiotics and surgical removal of the affected area.

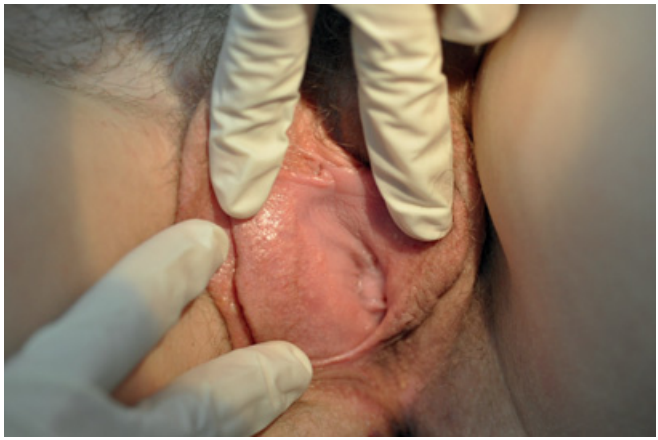

Figure 7.6 Sebaceous hyperplasia

A recent case report indicated that topical pimecrolimus was of benefit. We have also found topical tacrolimus to be useful.

Multiple Syringomas

Syringomas are benign adnexal tumours, consisting of eccrine sweat glands. They are usually multiple and are most commonly found under the eyes. There is a variant of multiple syringoma that is found on the vulva.

Presentation

Syringomas present with multiple small round papules, which are usually flesh-coloured but may also be brown or violaceous. The distribution is usually on the labia majora bilaterally.

The onset of these lesions is usually in adolescence or early adult life. Although on other body sites they are not symptomatic and present only a cosmetic problem, on the vulva, they may be intractably itchy.

Investigation

The histopathology is characteristic, demonstrating multiple clusters of eccrine glands in the dermis. This helps to differentiate them from Fox–Fordyce disease.

Management

Multiple itchy syringomas on the vulva can be a difficult clinical problem. Topical therapy with anti-inflammatory agents may be ineffective. Topical atropine has been reported to reduce itch. Definitive management is surgical removal.

Benign Vulval Neoplasia

Naevi and other benign neoplastic lesions may occur on any part of the skin, including the vulva. There are four categories:

1. melanocytic naevi and melanosis vulvae
2. vascular lesions of early and late onset
3. epidermal naevi
4. acquired benign tumours.

Melanocytic Naevi

Melanocytic naevi are quite common on the vulva and look no different from similar lesions on any other part of the skin.

Presentation

They present as small, slightly raised or pedunculated pink to brown papules, which may or may not grow hair. They generally appear from childhood until the mid-twenties.

Melanocytic naevi evolve slowly with time, gradually regressing with age.

On the vulva, melanocytic naevi rarely cause any morbidity unless they rub on clothes. Patients are often anxious about their potential to become malignant, but this risk is very small. Therefore, preventative removal is not necessary.

The main indication for removal is cosmetic or functional.

Examination

- Evaluation of a vulval melanocytic naevus is no different to a naevus elsewhere, but in practice, dermatoscopy of the vulva is difficult.

Photography and simple measurement is a good way to monitor melanocytic lesions. If such lesions have benign features, they can safely be observed. These features are:

- even colour
- regular border
- symmetry
- lack of itch or bleeding
- stability (remembering that over several years features may slowly change).

Melanosis Vulvae

Melanosis vulvae is strictly speaking not an acquired lesion, but acquired hyperpigmentation of the vulva. This condition is very common, but unusual before middle age.

The presentation is with multiple, discontinuous brown-to-black, well-defined macules on the labia minora and perineum (see Figure 7.7). These lesions are not raised. They are invariably asymptomatic and noted at Pap test or accidentally by the patient.

Melanosis vulvae often cause concern because it comes into the differential diagnosis of vulval melanoma. However, melanoma is usually a single lesion as opposed to melanosis vulvae, which is usually multiple.

Histopathology of melanosis vulvae simply reveals increased melanin in the basal layer of the epidermis. There is no evidence of malignancy.

Management

If there is any doubt, particularly with a solitary lesion and to allay a patient's fears, a small biopsy makes this diagnosis. Once this is established, reassurance is all that is required.

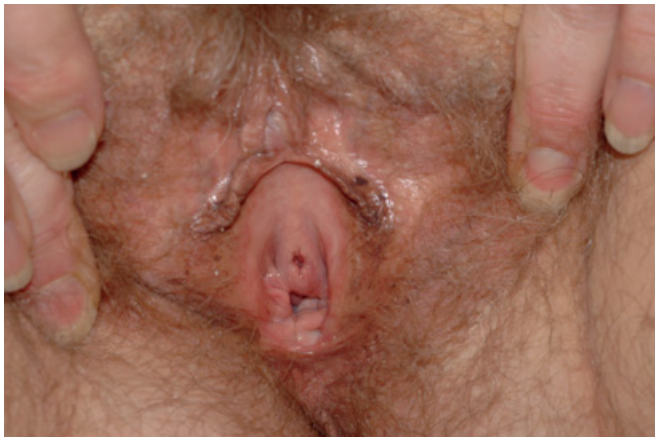

Figure 7.7 Benign vulval melanosis

Vulval Hyper-pigmentation

Vulval hyper-pigmentation is usually seen in dark skinned patients and is normal. It is similarly asymptomatic and is of earlier onset. The difference from melanosis vulvae is that it is confluent rather than patchy, involving usually the outer surfaces of the labia minora.

Drugs that cause hyper-pigmentation of the skin elsewhere may do the same on the vulva. The classic drug that does this is minocycline. A drug history should always be taken into account in any patient with acquired vulval hyper-pigmentation.

Acanthosis Nigricans

Acanthosis nigricans is an important condition to localised as it has associations with obesity, insulin resistance, type 2 diabetes, polycystic ovarian disease and internal malignancy.

Presentation

Acanthosis nigricans bilaterally affects the genital area and inner thighs, axillae and neck with a characteristic velvety hyperkeratotic thickening of the skin associated with hyper-pigmentation (see Figure 7.8). Vulval involvement alone is rare. It may be associated with multiple skin tags and has a characteristic histopathology showing localised keratosis-like thickening of the epidermis.

The difference between this condition and vulval hyper-pigmentation is the presence of hyperkeratosis evidenced by thickening of the skin surface.

Management

A finding of acanthosis nigricans mandates a glucose tolerance test and possible referral to an endocrinologist. In older patients where there is no obvious cause, a directed search for malignancy should be carried out.

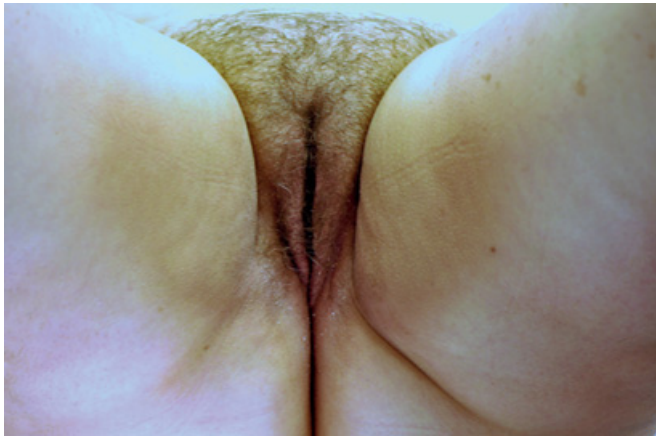

Figure 7.8 Acanthosis nigricans

Dowling Degos Disease

This is a very rare genetic condition but is included here because it comes into the differential diagnosis of acanthosis nigricans. It is inherited in an autosomal dominant fashion.

Presentation

It presents in adult life with multiple freckle-like lesions of the axillae and groins. The neck and other parts of the skin may also be involved. The pigmentation may also be reticulate or almost confluent.

It is not associated with endocrinopathies or insulin resistance. The characteristic histopathology will differentiate it from acanthosis nigricans.

Management

There is no effective treatment, nor is any required.

Vulval Angiokeratomas

Vulval angiokeratomas are vascular papules of the vulva. They are very common and are harmless. They are similar to Campbell de Morgan spots on the trunk.

Presentation

Patients present with multiple small red to purple papules on the labia majora (see Figure 7.9). The onset is from middle age and beyond, becoming more common with advancing age. When they are dark purple, there may be concern about melanoma.

Occasionally, these lesions may bleed if knocked or scratched, especially during a shower. Rarely, they may become much larger during pregnancy.

The histopathology is of a small haemangioma.

Angiokeratomas are usually easy to diagnose clinically, and like localised keratoses, there is a characteristic dermoscopic appearance.

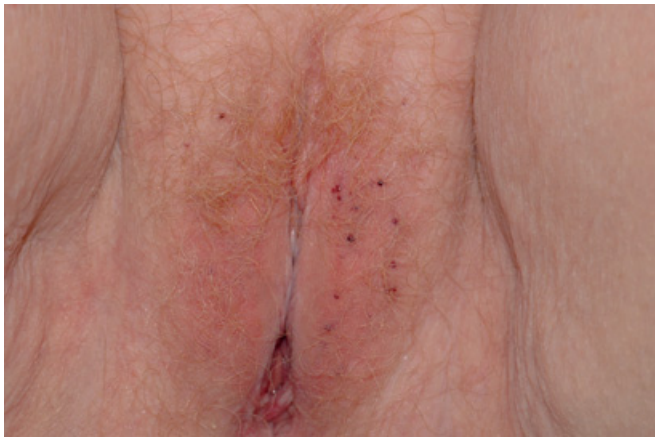

Figure 7.9 Angiokeratomas

Management

Reassurance is all that is required, but if any lesions are bleeding or causing discomfort, they can be easily ablated with simple diathermy or cryotherapy.

Vulval Lymphangiectases

These lesions are usually seen in patients who have had lymph node dissection and radiotherapy for a gynaecological malignancy and are the result of dermal fibrosis, which compresses the cutaneous lymphatics. They sometimes occur as the result of chronic bacterial cellulitis, or rarely Crohn's vulvitis. On occasion, they are idiopathic.

Presentation

These lesions present as clear blebs, which may from time to time leak clear lymphatic fluid. There is often also associated firm oedema of the surrounding area.

Patients with lymphatic stasis are at risk of secondary bacterial cellulitis, particularly with Group A *Streptococcus*.

Management

These lesions are incurable, but referral to a localised lymphoedema clinic is recommended for palliation. Patients should be warned to present immediately if they experience pain, redness, swelling or fever, which may represent cellulitis. Patients who have lymphangiectases as the result of chronic cellulitis will require long-term antibiotics.

Epidermal Naevi

In general, these lesions are uncommon and even less common on the vulva. They are hamartomas that usually appear in the first few years of life, extend for a few years, then are localised and persist through life.

There are many different types of epidermal naevi. On the vulva, the usual type is the keratinocytic or verrucous epidermal naevus.

Presentation

The clinical presentation is of a unilateral, flesh-coloured to brown keratotic papule that may be localised to the labia majora. They may be vaguely linear. Lesions may extend into the inguinal fold and onto the leg.

In the peri-anal area, epidermal naevi of the vulva may have a warty or pedunculated appearance. This will give rise to friction symptoms from clothes and wiping. The naevus may become intractably itchy and unresponsive to topical anti-inflammatory agents.

Investigation

Vulval epidermal naevi, particularly when not obviously part of a larger linear lesion, are frequently mistaken for lichenified eczema, or warts. In children, this may then lead to allegations of child sexual abuse. Biopsy will confirm the diagnosis.

Management

The most logical management is a combination of excision of symptomatic areas and reassurance for non-symptomatic ones. For symptomatic lesions, full thickness excision is a better option than shave excision or laser.

Many topical therapies have been advocated, including retinoids and calcipotriol, but our experience with them has been disappointing.

Cysts, Boils and Things that Simulate Them

Sebaceous Cysts

Case Study

A 54-year-old woman presents with two years of recurrently infected vulval cysts. She has a background history of itchy rashes under her breasts and under her pannus. She has had a large weight gain in the last five years, and wears incontinence pads daily for occasional urinary incontinence.

On examination, she is obese. On the hair-bearing vulval skin, there is a well-demarcated erythematous rash. There are multiple sebaceous cysts on the hair-bearing surface of the labia majora, but none that are infected. A vaginal culture is negative. There are similar rashes under the abdominal pannus and the breasts.

This woman has sebaceous cysts in the context of an irritant contact pad dermatitis. Both require active management to achieve the best outcome. She is advised to wear looser trousers in softer fabrics, and to dry her vulval skin after a shower with her hairdryer on the 'cool' setting. It is important that the use of pads be minimised, and alternatives such as incontinence underwear be used. A 1% hydrocortisone ointment is applied twice daily to the rash for six weeks, and thereafter a moisturiser is used daily. Any infected sebaceous cysts are managed with oral antibiotics if necessary, and incision and drainage.

Once the dermatitis is controlled, and there has been no infection for at least three months, the cysts can be excised. Weight loss will also help.

These benign lesions are quite common on the vulva and are often multiple, involving the labia majora bilaterally. They appear at any age from adolescence on but are unusual in children. They become slowly more numerous with time. The lesions tend to occur at points of friction, especially in obese women.

Sebaceous cysts of the vulva are usually merely a cosmetic problem, but occasionally they may become inflamed and even secondarily infected. In these situations, they may rupture, extruding their contents. Patients often make their situation worse by deliberately squeezing these lesions.

Presentation

Sebaceous cysts appear as dermal nodules with a yellowish colour. They are ovoid, mobile and well-defined (see Figure 7.10). The size varies from a few millimetres to 20 mm. These cysts have a characteristic histopathology showing a unilocular lesion with a dermally located keratin-filled cyst. These cysts have a punctum, from which the malodorous contents of the cyst can usually be extruded.

Management

Simple incision and drainage is adequate for most infected or sore cysts. Antibiotics are not usually necessary. If the distribution of cysts indicates a frictional aetiology, patients should modify their clothing, and must be warned never to squeeze these lesions.

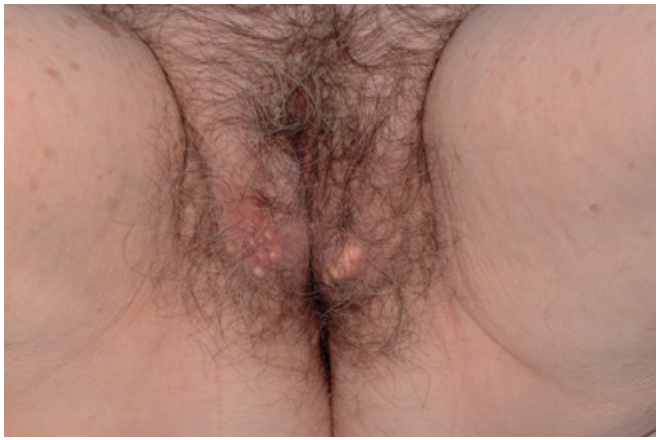

Figure 7.10 Sebaceous cysts

Our usual advice in uncomplicated cases is not to treat, but some patients are sufficiently concerned to seek surgery. The patient must understand that more cysts may form, and should be counseled about reducing heat, sweat and friction in this area.

The loose connective tissue of the vulva makes excision easier than on other parts of the skin. The cyst can usually be extracted through a small incision made over the lesion. Excision should be delayed for at least three months after a secondary infection in order to minimise post-operative infection.

Folliculitis and Boils

Folliculitis is a superficial *Staphylococcus aureus* infection of hair follicles. Boils are simply a deeper and more extensive form of folliculitis.

The vulva is prone to hair follicle infection because:

- it is a hair-bearing area
- it is a common site for chronic Staphylococcal carriage
- pubic and groin hair removal by waxing and shaving produces micro abrasions on the skin that are easily infected
- heat, sweat and friction further increases the chance of hair follicle disruption.

Presentation

Folliculitis presents with multiple superficial itchy pustules. Boils are larger, deeper and both tender and painful (see Figure 7.11). The two conditions may co-exist. Both tend to be very chronic because the underlying pathology is usually a Staphylococcal carrier state, which has to be addressed in order to obtain cure.

Investigation

Folliculitis should be proven with a simple bacterial swab, in order to prove the diagnosis and determine the sensitivity of the organism, and to exclude MRSA carriage.

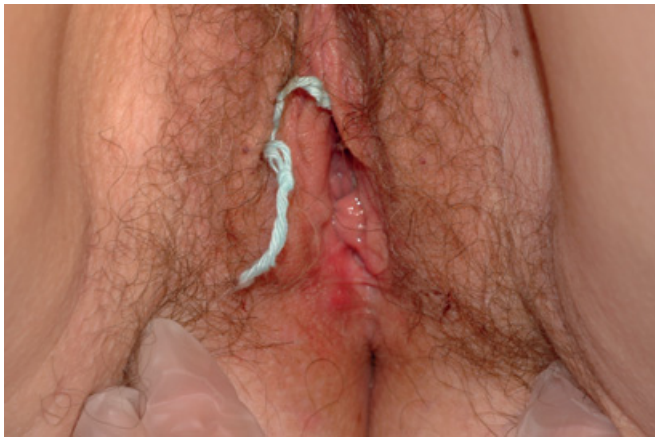

Figure 7.11 Boil

Management

Treatment involves the following:

- drain any boils
- treat the acute episode with appropriate oral antibiotics
- use an antiseptic wash in the shower daily
- hot wash all garments, sheets and towels for a month
- stop waxing and shaving until the episode is over
- ask about any symptoms in the patient's partner who may also require the same strategy. Even if the partner is asymptomatic, they should also wash with an antiseptic at the same time
- when the patient resumes hair removal, recommend clipping rather than shaving or waxing
- if the patient wants to be permanently free of genital hair, recommend laser hair removal.

Inflammatory Tinea of the Pubic Area

Although very unusual, tinea (a dermatophyte infection) may cause a dramatic eruption of sudden onset involving the vulva and pubic area.

Presentation

It appears indistinguishable from a severe attack of boils and may present with swelling, inflammatory nodules and draining sinuses.

The clue that you are not dealing with a Staphylococcal infection is lack of response to antibiotics and negative swabs. The patient may own guinea pigs or other pets from which the infection is acquired.

Investigation

The diagnosis is made with a skin biopsy and culture of skin scrapings.

Management

Treatment is with an oral anti-fungal drug such as griseofulvin, itraconazole, fluconazole or terbinafine in the same doses and duration used to treat tinea of the scalp or beard.

Hidradenitis Suppurativa

This is a relatively common, chronic and recurrent condition of the vulva and other skin sites, the severity of which ranges from a mild nuisance to a disabling, life-ruining condition.

Hidradenitis suppurativa is a genetic disease that may run in families. It is an androgen-sensitive condition that requires adult levels of hormone to express itself. It usually appears for the first time after puberty and rarely appears after menopause. Despite the association with androgen, women with hidradenitis suppurativa have normal serum androgen levels. The problem is the hair follicle, which is more sensitive than normal to androgenic stimulation.

It has been observed that cigarette smoking is a trigger in hidradenitis suppurativa and it has been postulated that nicotine may be causative in some patients. It is also more common in obese patients.

Aetiology

The aetiology is thought to be a chronic inflammation of apocrine sweat glands and it therefore is located on areas where these glands are present: vulva, peri-anal area, buttocks, axillae and under the breasts. The exact pathogenesis is unknown. The currently accepted theory is that apocrine glands are easily blocked in these patients, which then rupture, causing inflammation of surrounding tissue.

The disease is not an infection and swabs may grow a variety of non-pathogenic bacteria, the treatment of which is ineffective.

Presentation

Hidradenitis suppurativa presents with tender, painful nodules that drain pus, sinuses, pustules, abscesses and comedones. Lesions may be followed by significant scarring.

Patients often go without a diagnosis for many years, being thought to have a recurrent Staphylococcal infection even though swabs are consistently negative.

When hidradenitis suppurativa involves the vulva, the distribution is usually on the mons pubis, labia majora, inguinal folds, peri-anal area and buttocks (see Figure 7.12).

Although it is more common in women than men, it is less severe. It may exacerbate pre-menstrually.

It may be associated with severe acne, pilonidal sinuses and a condition known as dissecting cellulitis of the scalp.

A number of severity assessment tools have been proposed. However, severity does not always correlate well with response to treatment.

Investigation and Diagnosis

As long as Staphylococcal and fungal infection have been ruled out by culture, the diagnosis is a clinical one. The finding of vulval comedones is pathognomic: they occur in no other disease.

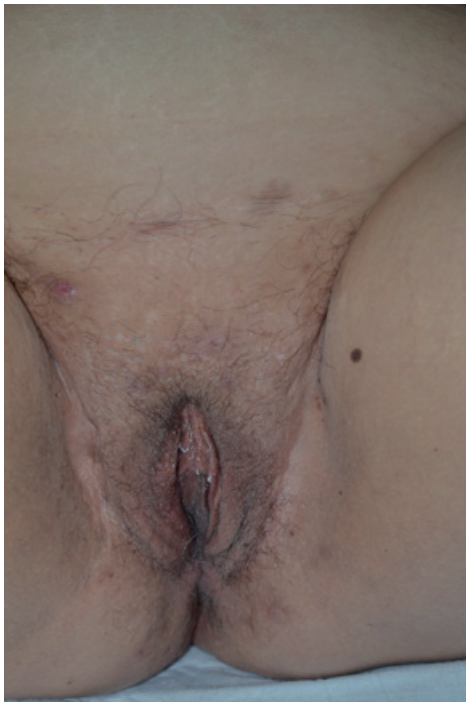

Figure 7.12 Hidradenitis suppurativa

A biopsy is generally not diagnostic.

Rare diseases that may come into the differential diagnosis, particularly with perianal disease, include atypical pyoderma gangrenosum, Crohn's disease and infections such as Mycobacterial folliculitis. However, none of these will be characterised by comedones.

Management

Many patients arrive with a long history of recurrent episodes of minor surgery to drain or excise lesions and multiple short courses of antibiotics.

Our experience has been that in most cases, treatment is straightforward. However, some patients are very challenging to treat. Unfortunately, evidence for treatment is largely anecdotal because there is little high-quality research.

Medical Therapy

In mild cases, topical clindamycin 2% or erythromycin 2% with regular antiseptic washes may reduce symptoms, but systemic therapy tends to be much more effective.

If the patient smokes, she should make every attempt to stop. Obesity will exacerbate the suffering in this condition, but weight-loss alone will not have a significant effect on the course of the disease.

Our usual first line of therapy is an oral anti-androgen, which has been shown to be more effective than antibiotics.

If the patient is on the combined oral contraceptive pill, we change this to one with an anti-androgenic progestogen such as cyproterone acetate or drosperinone.

If the combined oral contraceptive pill is not indicated, the most cost-effective and safe treatment in our hands is spironolactone. It is well tolerated and inexpensive, with high patient acceptance in the long term. If there is a risk of pregnancy, contraception must be advised, as this drug may feminise a male fetus. It may also result in menstrual irregularity in those not on the oral contraceptive pill and has been associated with breast lumps and tenderness. We start at 100 mg/day, but in resistant cases, the dose can be increased to 200 mg. Serum potassium levels should be regularly checked.

Some women, particularly when thin, may experience low blood pressure at 100 mg/day and it is safest in these patients to commence at 50 mg and slowly increase the dose. In practice, we find that these are relatively unusual adverse events are uncommon.

Other anti-androgens are:

- cyproterone acetate 5–25 mg/day
- finasteride 5 mg/day
- dutasteride 0.5 mg/week
- flutamide 250 mg/day.

Intralesional steroid injections using triamcinolone 10 mg/ml into early lesions may abort individual attacks.

Other Treatments

- Many authors advocate oral antibiotics; however, we find these relatively disappointing and a recent study showed that there was no difference to outcome when used with or without spironolactone.
- Oral and topical retinoids have been advocated in the medical literature but we have also found them disappointing.
- There are anecdotal reports of the use of immunosuppressants such as cyclosporin and methotrexate.
- There has been a case series of women treated with metformin.
- Recently, TNF-alpha inhibitors such as infliximab and adalimumab have shown promise in treating patients with resistant hidradenitis suppurativa. These 'biologic agents' are currently very expensive but our experience with them so far has been positive. Referral to a dermatologist is recommended.

Unfortunately, this condition does not always remit at menopause and treatment may need to be lifelong. Many patients are willing to accept this because of the severe morbidity imposed by untreated hidradenitis.

Surgery

Acute painful lesions may be incised and drained, but marsupialisation has a much better effect. This should be followed by topical application of clindamycin 2% or erythromycin 2%. This is palliation, however, and not cure.

Excisional surgery is curative and therefore the treatment of choice. However, this applies only to patients who have involvement in areas that can be widely excised without producing disfigurement. For patients with involvement of the labia, peri-anal skin or buttocks this may not be a practical option. CO_2 laser ablation has also been tried.

These options come with the risk of major post-operative implications for wound healing, scarring and sexual function, and should be reserved for very severe cases.

Sebaceous Adenitis

This recently described condition involves only the inner surface of the labia minora. It presents with recurrent tender and painful nodules, often flaring pre-menstrually. These nodules may drain purulent material. The onset of this condition is in adult life, the mean age being the mid-thirties. The sebaceous gland contains androgen receptors, which are thought to be causative.

Unlike hidradenitis suppurativa, this condition does not scar, nor is it associated with comedones.

Biopsy reveals sebaceous gland inflammation.

The differential diagnosis of such lesions includes bacterial infection. Once culture has ruled this out, the recurrent and multiple nature of these lesions differentiates it from both of these conditions. An infected Bartholin's cyst is easy to rule out because it arises in a different area.

Like hidradenitis suppurativa, this condition can be suppressed with topical macrolides, and oral anti-androgens, including the oral contraceptive pill with an anti-androgenic progestin. We have used spironolactone successfully in this condition.

Vulval Neoplasia

Because the vulva is part of the skin, it is not surprising that most neoplastic conditions affecting the vulva are occur on other parts of the skin.

These include:

- basal cell carcinoma
- squamous cell carcinoma
- VIN
- melanoma
- Paget's disease, which occurs on the vulva, where is it known as Extra-mammary Paget's disease.

Biopsy should immediately be performed for any new lesion that appears suspicious or presents with a change in characteristics over time. If there is genuine concern about malignancy, formal excision is advised.

Vulval Intraepithelial Neoplasia

Vulval intraepithelial neoplasia is the most common form of vulval neoplasia. Histopathologically, it is an in situ form of squamous cell carcinoma (vSCC), indistinguishable from what is known on sun-exposed skin as Bowen's disease.

Case Study

A 38-year-old woman is referred with a two-year history of vulval warts, which have been resistant to multiple topical therapies.

On examination, the lesions are in fact warty HSIL (high- grade squamous intraepithelial neoplasia). Biopsy confirms full-thickness HSIL with strong p16 positivity on

immunohistochemical testing. A cervical screening test is positive for HPV 18 with cervical cytology showing cervical intra-epithelial neoplasia (CIN) III.

The patient was referred to a gynaecologist for further management. This case illustrates the importance of biopsy when the initial diagnosis does not lead to effective treatment.

However, on the vulva, the most common aetiological factors are not sun but oncogenic genotypes of HPV, lichen sclerosus, and lichen planus.

There is still much controversy about the classification and prognosis for VIN. It used to be thought that VIN could be graded in the same way as CIN. This grading does not actually reflect the biological behaviour of VIN, and should be abandoned. Clinically significant VIN is full-thickness neoplasia with invasive potential.

Vulval intraepithelial neoplasia carries more risk than its equivalent on sun-exposed skin because if progression to invasive squamous cell carcinoma occurs, the risk for metastasis is greater on the vulva.

There are two types of VIN:

- uVIN (u = usual), which is HPV-associated and found in younger patients. It is more common in patients who smoke, who have genital or anal oncogenic HPV carriage or are immunosuppressed. The HPV genotypes most often associated with VIN are the oncogenic types 16 and 18, also found in CIN and squamous cell carcinoma of the cervix. uVIN has a lower invasive potential than dVIN, but a higher risk of recurrence after treatment. It is more likely to appear in multiple sites.
- dVIN (d = differentiated), which is lichen sclerosus- or lichen planus-associated and found in older patients. It is not HPV-associated, and has a higher risk of progression to invasive disease than uVIN.

Presentation

Vulval intraepithelial neoplasia presents as papules and plaques that may be any colour from white to brown. They may be solitary or multiple. Frequently, they have a warty, rough surface.

Other presentations include persistent raw or ulcerated areas. The lesions are frequently itchy but this is not invariable.

The difference between VIN and vSCC relates histologically to whether the lesion has invaded below the basement membrane of the epidermis. Once this has happened, the prognosis of the lesion becomes much more guarded and patients may require lymph node dissection in addition to wide local resection. Unlike squamous cell carcinoma of the skin, which rarely metastasizes, squamous cell carcinoma of the vulva may do so. If vulval cancer spreads beyond the groin lymph nodes, the prognosis is grave.

Management

Details of treating this condition are beyond the scope of this book. The first step is to make a diagnosis by taking a biopsy of the lesion.

Further treatment includes surgery, laser and in some cases topical therapy with agents such as imiquimod. Recurrence occurs frequently.

Referral to a gynaecological oncologist is highly recommended.

Extra-mammary Paget's Disease

This very uncommon condition is frequently a disease of the elderly. The risk of invasion is low. It is often confused with persistent dermatitis or psoriasis because of its appearance. The diagnostic clue therefore is its resistance to dermatitis and psoriasis treatment.

Presentation

Patients usually complain of itch, soreness and swelling or may present with the characteristic appearance but no symptoms.

There is often a plaque, which is usually red but may be pale, is either unilateral or bilateral and may involve the vulva and or peri-anal area. The edge of the plaque is usually better defined than for dermatitis, simulating psoriasis. The surface of the plaque may demonstrate a white scale that may look like 'icing sugar' (see Figure 7.13).

Investigation

The very characteristic histopathology makes the diagnosis easy on a biopsy.

The condition may be associated with underlying malignancy of the bowel and bladder and requires investigation. Immunohistochemical staining of the biopsy specimen may help to confirm the diagnosis and differentiate this risk.

Management

Treatment involves a combination of surgery and radiotherapy. Palliation with imiquimod may also be used and recent studies support this in preference to surgery in cases where

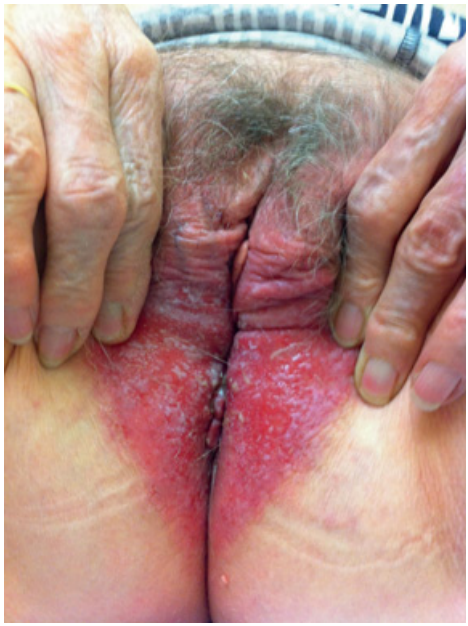

Figure 7.13 Extra-mammary Paget's disease

there is no invasive disease or underlying malignancy. Imiquimod therapy has to be prolonged and many patients find it difficult to tolerate.

We recommend referral to a gynaecological oncologist.

Melanoma of the Vulva

Melanoma of the vulva is extremely rare. The appearance is no different from elsewhere on the skin, presenting with a slowly expanding red to brown to black lesion that may itch and bleed.

The hallmarks of melanoma include variation in colour, irregular shape and constant evolution. Lesions are often not symptomatic until they ulcerate.

Any suspected vulval melanoma should be excised with urgency and the patient referred to a tertiary referral centre for further management.

Further Reading

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Chapter

8

Vulval Pain and Dyspareunia

Contents

Vulvodynia is a term that every doctor with an interest in vulval disease has heard of and read about. You will notice, however, that it is not the name of this chapter. This is because vulvodynia is by definition a collection of symptoms, not a disease entity in itself.

Vulvodynia is in fact a poorly defined concept that simply means vulval pain. When your patient presents with vulval pain, you need to sort her into a meaningful diagnostic group. The management of each sub-type is different. There is no single therapy that can be applied to all patients yet the existing literature on the subject can give the impression that there is.

Vulvodynia is a term developed by the International Society for the Study of Vulvar Disease (ISSVD) in 1983. Their current definition is 'vulvar discomfort, most often described as burning pain, occurring in the absence of relevant visible findings or a specific, clinically identifiable neurologic disorder'.

The ISSVD has also produced a system of classifying vulval pain, where patients are categorised into (A) those with an identifiable cause and (B) those that fit their case definition for vulvodynia (see Table 8.1).

- 'Provoked' means that pain is in response to friction, pressure, intercourse, insertion and so on.
- 'Unprovoked' means that the pain occurs spontaneously.

Despite the ISSVD's definition, doctors have diverse interpretations of the term, and the literature on the subject can be confusing. Some clearly see it as vulval pain, some as itch,

Table 8.1 ISSVD Terminology and Classification of Vulvar Pain (2003)[1]

A) Vulvar pain related to a specific disorder
   1) Infectious (e.g., candidiasis, herpes, etc.)
   2) Inflammatory (e.g., lichen planus, immunobullous disorders, etc.)
   3) Neoplastic (e.g., Paget's disease, squamous cell carcinoma, etc.)
   4) Neurologic (e.g., herpes neuralgia, spinal nerve compression, etc.)

B) Vulvodynia
   1) Generalised
      a) Provoked (sexual, nonsexual or both)
      b) Unprovoked
      c) Mixed (provoked and unprovoked)
   2) Localised (vestibulodynia, clitorodynia, hemivulvodynia, etc.)
      a) Provoked (sexual, nonsexual or both)
      b) Unprovoked
      c) Mixed (provoked and unprovoked)

others see it as vulval discomfort of any sort, and some see it as a disease in itself: often with which they have had difficulty managing! In other words, the label is often applied inappropriately to any treatment resistant vulval condition.

We prefer to avoid the term vulvodynia altogether. We refer to vulval pain because it is simple, straightforwards and does not carry with it the baggage of many years of medical confusion. If there is an observable lesion that is the cause for the pain, we call this 'lesional pain'. If there is no lesion, we call it 'non-lesional'.

We are of course at odds with many other authorities on the subject. We realise that there are websites and societies devoted to vulvodynia. However, our belief is that continuing to use this term, even though it is widely entrenched, will only prolong confusion on how to best manage the patient with vulval pain.

Pathophysiology of Vulval Pain

The mechanisms involved in vulval pain are still very poorly understood. There are many reasons why the vulva is a pain-prone part of the body. These include:

- The complex anatomical structure of the bony pelvis and lower spine, and its vulnerability to damage.
- The central position of the vulva and vagina within the pelvic floor myo-fascial complex, which facilitates pain referral from other pelvic viscera, specifically the uterus, bowel and bladder.
- The fact that this area is subject to a great deal of physical stress: urination, menstruation, sexual intercourse, childbirth, defaecation, and friction from clothes and pads.
- Personal hygiene habits that may inadvertently exacerbate the original problem.
- The high levels of anxiety and fear that are often attached to problems involving the genital area.

- The crucial importance of the genital area to a woman's sexual well-being and self-esteem.

Many of our patients with vulval or vaginal pain complain of multiple pelvic problems, and we think that they are often related to each other. Our experience is that disease or dysfunction in more posteriorly located organs tend to influence dysfunction in those more anteriorly located. Considering pelvic anatomy from a different point of view may assist.

Functionally, the lower pelvis may be understood to contain five components, arranged posterior to anterior:

- lumbo-sacral spine and hip joints
- sigmoid colon, rectum and anus
- vulva and vagina
- bladder and urethra
- the pubic symphysis where the two innominate bones join.

All of these elements are joined by the pelvic floor: a complex myo-fascial structure that provides support and sphincter function to the viscera, but also allows for referral of pain and dysfunction from one part to another. In general, referral of pain and dysfunction in the lower pelvis tends to be posterior to anterior. This means that the bladder and urethra, the most anterior parts of the pelvis, are especially susceptible to influence from disease or dysfunction in other parts. It is well known that colo-rectal disorders influence bladder dysfunction, but vulvo-vaginal disorders are also capable of the same influence. Since the lumbo-sacral spine forms the posterior portion of the pelvic girdle, it is also unsurprising that spinal disorders can also influence bladder function, not to mention the lower bowel and vulva and vagina. Further, the hip joints are related to the pelvic floor via the hip rotator muscles that attach onto the anterior aspect of the sacrum.

Nerve Supply to the Pelvis and Genital Area

Innervation to the vulva is provided by the pudendal nerve, which originates from S2 to S4 nerve roots and the ilio-inguinal and genito-femoral nerves, arising from L1 to L2 (Figure 8.1). The two latter nerves are predominantly sensory, but the pudendal nerve contains motor, sensory and sympathetic fibres, which supply the complex autonomic reflexes of the pelvic organs. The epithelium of the vagina proper (i.e., deep to the hymenal ring) is not normally sensitive to pain.

The pudendal nerve supplies both the anal and urinary sphincters whereas the muscles of the pelvic floor are mostly innervated via direct branches from the sacral plexus (S3–S5) with some input from the pudendal nerve, both voluntarily from higher centres in the brain and reflexly via the spinal cord.

It is known that there is a relationship between muscle function in the pelvis and pelvic pain, and studies have demonstrated that patients with pelvic pain have higher levels of resting muscle tone than other persons.

The convergence in the spinal cord of afferent impulses from viscera, skin and muscle can also lead to the phenomenon of referred pain. We are familiar with this when it comes to sciatica, but it is less well known that pain may also be referred to the vulva and distal vagina.

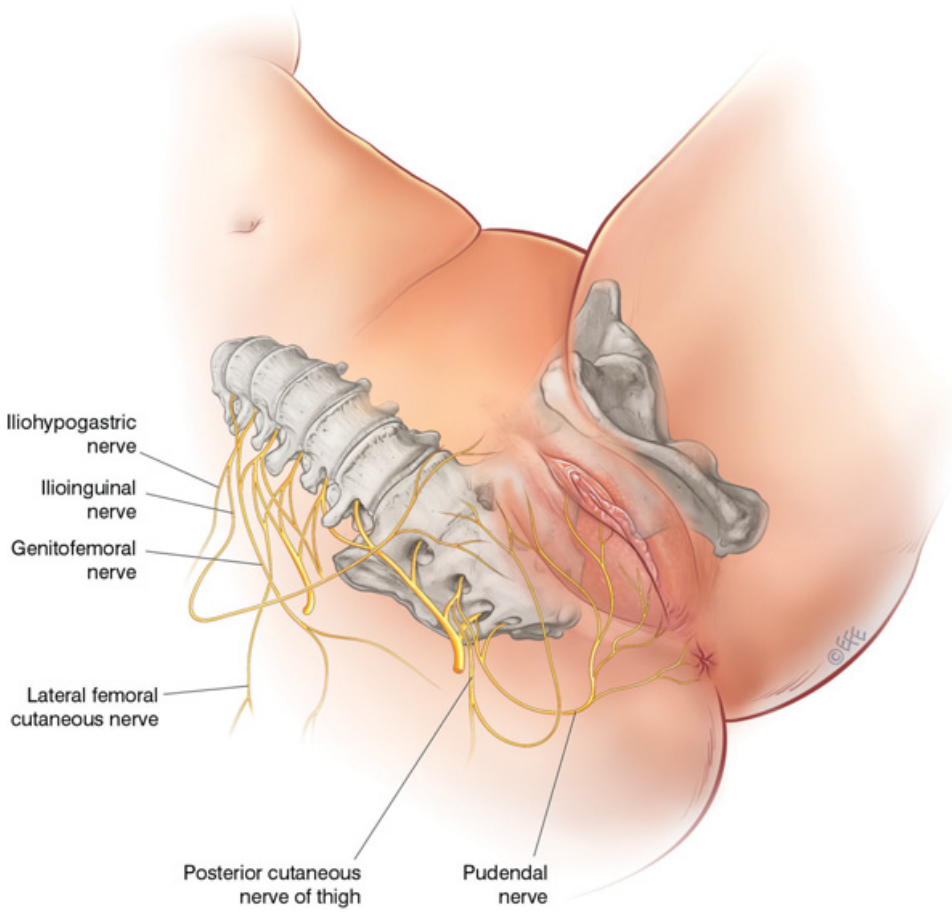

Figure 8.1 Nerve supply of the vulva. With permission from Dr Levent Efe, CMI

This convergence can also lead to alterations of sensation to nearby viscera, particularly the bladder. It is not uncommon for patients with vulval pain to complain of frequency, urgency, lower abdominal pain and burning on urination.

Autonomic dysfunction may lead to loss of control of the vascular system of the vulva. The result is a variable degree of erythema of the vulval skin and the epithelium of the introitus. This is often misinterpreted as a rash, and treated with anti-fungal agents or topical corticosteroids. The vasoconstricting action of these latter medications is followed by a reflex vasodilatation, which may increase erythema and produce discomfort.

Chronic Pain

Pain sensation is mediated by three types of afferent fibres: the large myelinated Type A-beta fibres that are responsible for touch; the smaller myelinated Type A nerve fibres; and the poorly myelinated or non-myelinated Type C fibres. These latter fibres are responsible for pain perception, and the pain mediated by Type C fibres in particular often has a burning quality. Type A fibres can, however, become involved in pain sensation, and when this

occurs, patients may develop allodynia. In this condition, stimuli that would normally elicit sensations of touch or pressure are perceived as pain. This explains why patients with vulval pain often find it difficult to cope with pressure from clothes.

When patients experience chronic pain, afferent sensory processes mediated by Type C peripheral nerve fibres via the dorsal horn spinal cord neurones appear to become sensitised, discharging more easily to lower levels of stimulation and at lower thresholds. They may even discharge spontaneously. This phenomenon has been termed 'wind-up'. There may be associated pathological changes in the dorsal horn connection and perhaps also in higher centres. It is helpful to understand this when there is no apparent noxious stimulus present. We also know that depressed and anxious patients have more problems with chronic pain, and that mental state is integral to pain experience.

It is important as a clinician to have a concept of how pain can occur in the absence of observable abnormality, particularly when it comes to explaining the diagnosis to the patient and indeed accepting it oneself.

Presentation

Broadly speaking, patients presenting with vulval pain tend to present either with something observable that explains their pain or with a normal looking vulva.

They can be divided into two main groups:

- pain that is directly attributable to an observable vulval or vaginal lesion or disease via cutaneous nociceptors: *lesional pain*
- pain that is experienced in the absence of any observable vulvo-vaginal pathology and where the physical examination is normal for the patient's age and ethnic group: *non-lesional pain.*

It must be remembered that the presenting symptoms may sometimes be due to *co-existing* lesional and non-lesional aetiologies.

Case Study

A 47-year-old woman is referred with a six-month history of vaginal dyspareunia and urinary frequency, which started after an acute episode of vulval itch, thought to be candidiasis. She has regular periods. Her general practitioner has performed vaginal and urine cultures, which are both normal. A gynaecologist has prescribed topical vaginal oestrogen and anti-fungals, both of which made the symptoms worse. A urologist has performed urodynamics and cystoscopy, which were unhelpful. A pelvic physiotherapist has tried 'trigger point therapy' on tight pelvic floor muscles without success.

This woman has had many years of proctalgia, which started during an acute exacerbation of her chronic L5/S1 spinal pain. The proctalgia is worse with defaecation and standing, but improved with lying down. A colo-rectal surgeon injected botulinum toxin into the anal muscles without improvement in the proctalgia.

On examination, there is an erythematous, silvery and scaly rash over the vulva and peri-anal skin, extending into the natal cleft. There is psoriatic nail pitting and psoriaform rashes behind both ears. Single-digit palpation of the vaginal introital and anal muscles shows fixed spasm.

The fundamental diagnosis is of genital psoriasis, complicated by pelvic floor muscular pain. The spinal dysfunction is also promoting the pelvic muscular pain.

Initial management involves controlling the psoriasis with a moderate-potency corticosteroid and then 2% LPC ointment. This reduced the urinary frequency, dyspareunia and proctalgia. Subsequently, physiotherapy with a practitioner with pelvis and spinal skill sets produced further improvement.

Lesional Pain

Pain due to an observable lesion or dermatosis is often caused by dermatological conditions that cause inflammation, ulceration, blisters, fissures and adhesions, and conditions that cause vaginitis.

- It is frequently provoked by physical stimuli such as friction during intercourse or when inserting or pulling out a tampon, rubbing, scratching or wearing tight clothes.
- It is often accompanied by the symptoms of the causative dermatosis.
- It is usually bilateral.
- It is often worse during the night, when there are fewer external stimuli.
- It resolves promptly when the underlying condition resolves.

Lesional pain is usually well localised and has the familiar qualities of pain induced by injury.

Application of creams, which contain irritating substances such as preservatives, may sting. Changing from a cream to ointment (which has no preservatives) usually solves this problem, and also helps to confirm that is lesional.

This sort of pain resolves promptly when the underlying condition resolves.

When patients describe lesional pain, they usually use the following words: cut, split, tearing, sandpaper and sore.

Non-lesional Pain

We believe that the most likely explanation for non-lesional pain is neuropathy and/or referred pelvic muscular dysfunction. The source of this neuro-muscular pain can include visceral pelvic problems such as prolapse, irritable bowel, particularly with chronic constipation, irritable bladder, uterine pain whether due to endometriosis or other pathology or neuromuscular problems such as spinal or hip disease or dysfunction. A patient may have pain caused by both. Sometimes a patient presents with pain, which seems to have started with an episode of vulval skin disease (especially genital herpes or chronic thrush), but which persists after the skin problem has been resolved. Even though there has been a physical historical trigger for the pain, it should still be treated in this group.

Conversely, if a skin disease is present, it should always be initially considered as the cause of the patient's symptoms, even if this eventually proves not to be the case. In other words, once the skin disease has been adequately treated, any ongoing pain is not caused by it. Teasing this out requires a trial of therapy.

Non-lesional pain usually has the qualities of neuropathic pain: it is poorly localised and usually has a burning quality. It is spontaneous but can also be worsened by the sort of physical stimuli that would not normally cause pain, such as pressure from tight clothes, sitting or the application of topical therapy. There is a sub-group of non-lesional pain that is well-localised. This tends to be more musculo-skeletal in origin, rather than purely neuropathic.

Any form of chronic painful vulvo-vaginal or pelvic condition can predispose a patient to non-lesional pelvic pain. This includes period pain, bowel pain, chronic irritable bladder and any painful vulvo-vaginal condition. The reason is that chronic pain of any sort not only causes painful muscle spasm but also changes the way the brain perceives pain so that the threshold for experiencing pain becomes lower with time. This is called neuro-plasticity.

Very occasionally, patients with psychiatric conditions can experience vulval pain as a symptom of their condition, and malingering patients may also sometimes complain of

vulval pain. Our experience is that vulval pain due *primarily* to psychiatric disease is no more common than in any other part of the body.

Non-lesional pain has the following characteristics:

- It typically is best on first arising, becomes worse throughout the day, and is improved with rest at night.
- It is often positional: worsened by prolonged sitting and tight trousers, and improved by standing.
- Hyperalgesia is characteristic of neuropathic pain and describes severe pain experienced from mild pain stimuli such as light touch. This can cause extra confusion because application of diverse topical therapies all seems to cause pain. This is inevitably attributed to the products themselves, but is actually hyper-sensitivity to touch.
- Poor localisation of pain is often a feature of neuropathic pain. When asked to localise this sort of pain, patients are often unable to do so, and are only able to indicate the general area that the pain is experienced.
- Well-localised and especially unilateral pain is usually referred from the spine (similar to sciatica) or hips. Patients will localise a portion of skin on the affected side, but examination will show normal skin, and no tenderness at this site.

When patients describe non-lesional vulvo-vaginal pain, they use the following words: burning, rawness, dryness (even when on examination there is no evidence of it), crawling, irritation, vulval awareness, itch or 'almost itch', stabbing, pulsing and stinging.

Which Skin Disease of the Vulva Is Likely to Be Painful?

Dermatological conditions that affect the vulva tend to present with itch. Pain may be present, but it is usually sharp, easily localised pain that is due to excoriation from scratching, or fissuring, which may occur in any dermatosis. Dyspareunia if present is usually due to friction involving raw areas.

Dermatological conditions that are predominantly painful rather than itchy are uncommon and include diseases that cause loss of epithelial integrity. This includes lichen planus, desquamative inflammatory vulvo-vaginitis, aphthous ulcers, erythema multiforme and fixed drug eruption. Bullous diseases such as bullous pemphigoid and cicatricial pemphigoid can cause painful erosions but are extremely rare. Crohn's disease can present with painful erosions associated with oedema.

Vulval varicose veins may cause a dull ache particularly after long periods of standing.

Infections such as genital herpes, vulval Staphylococcal cellulitis and hidradenitis suppurativa are characteristically painful.

Atrophic vulvo-vaginitis tends to present with dyspareunia and a sensation of dryness.

Vaginal Dyspareunia

It is important to differentiate vaginal (superficial/entry) dyspareunia (felt in the vagina) from abdominal (deep) dyspareunia (felt in the abdomen). The first is usually caused by problems in the lower pelvis, and the second by problems in the upper pelvis or abdomen. There is widespread confusion about what constitutes 'deep' dyspareunia, with many doctors assuming this means 'deep in the vagina'. Our experience is that the important distinction is between *abdominal* as opposed to *vaginal* dyspareunia, and that it does not

matter (in a diagnostic sense) how deep in the vagina it is felt. In the context of this book, we are discussing vaginal dyspareunia.

We find it most helpful to consider vaginal dyspareunia as a sub-set of vulval pain. Dyspareunia is usually primarily physical in origin, and therefore needs to be assessed in the context of the wider pelvis. Most often it is part of a syndrome of non-sexual vulval pain, and it is the history of this background pain that will help in making a diagnosis. Dyspareunia can however occur as the only presenting symptom. In other words, the patient has no symptoms except pain during intercourse and/or tampon insertion.

Like more generalised vulval pain, introital dyspareunia may also be lesional or non-lesional. Lesional dyspareunia clearly occurs because the vulva and vagina are raw and inflamed.

Non-lesional dyspareunia occurs in the absence of any observable disease that could explain it. We believe that it is most often due to neuromuscular dysfunction. This type of dyspareunia may also be triggered by an underlying lesional disease but may remain long after the disease resolves. Rarely, it may be a somatoform disorder, but we must stress that a somatoform disorder causing dyspareunia should be a diagnosis of exclusion.

When discussing dyspareunia, it is important to explain pubococcygeal muscular dysfunction. This is found in many patients who have experienced vulval conditions that have caused dyspareunia for any reason. It can also occur as a primary problem in response to emotional stress and anxiety.

In this situation, the pubococcygeus muscles (the muscles that attach anteriorly to the pubic bone and meet within the substance of the perineal body) go into spasm as soon as any pressure is applied to the introitus. The patient describes a sharp, tearing sensation on intromission and immediate relief as soon as intercourse ceases. Tampon insertion often causes similar symptoms. It is unusual for this pain to linger for long after intercourse ceases unless there is also a lesion that has been irritated during intercourse.

Pubococcygeus muscle spasm is detectable on per vaginal examination. It may be virtually impossible to insert an examining finger into the vagina because this spasm, and any attempt to do so is described as severe pain by the patient. However, a patient who is relaxed with you as a doctor but apprehensive about intercourse may appear deceptively normal.

In the Lesional Group (Nociceptive Pain)

When dyspareunia is due to an ulcer or fissure, patients usually complain of the sort of pain that we would all experience if we cut a finger. This is known as 'nociceptive pain', in other words mediated by nociceptors in skin. This pain is usually well-localised, and often accompanied by a small amount of bright post-coital bleeding because of a skin disease that is prone to fissuring, such as lichen sclerosus. In this situation, the pain may not be immediate but may develop during intercourse. It then typically lasts for several days, until the fissure has healed.

Dyspareunia due to local physical causes usually improves promptly when the underlying dermatological condition is healed. However, it must be recognised that by the time effective treatment for the dermatosis is initiated, it may have caused secondary pubococcygeus muscle dysfunction that continues to cause the typical superficial dyspareunia described earlier.

In the Non-lesional Group (Non-nociceptive Pain)

In these patients, there is often a background of poorly localised or unilateral vulval pain. Some patients with non-lesional vulval pain deny dyspareunia, but say that they no longer want to have sex due to low libido or fear of being hurt. Those who do experience dyspareunia are usually either experiencing hyperalgesia or the same secondary pubococcygeus muscle dysfunction that occurs in patients with nociceptive pain.

Dyspareunia due to Oestrogen Deficiency (Atrophic Vulvo-vaginitis)

Atrophic vaginitis is due to oestrogen deficiency and may present with dyspareunia alone, which patients often correctly identify as being associated with dryness.

These women are either lactating, post-menopausal or very thin with secondary amenorrhoea.

Some patients, particularly those with a tendency to dermatitis elsewhere or who are atopic, may also develop a mild dermatitis of the introitus and labia minora in association with oestrogen deficiency. They may then complain of itching or irritation.

Post-menopausal women who are prone to the vulvo-vaginal effects of post-menopausal oestrogen deficiency are however also the group who may suffer from neuropathic vulval pain. As a result, an observation of atrophy may not be the cause of their vulval pain.

Examination

Examination shows a pale mucosa, with little lubrication and loss of normal rugosity. When the labia minora are pressed together, they literally stick to each other. In younger lactating patients, however, there may be little to see other than a somewhat dry surface.

Management

Management should be initiated with topical treatment using oestrogen cream or pessaries, initially daily for two weeks and then twice weekly. Long-term use will be required if a beneficial response is achieved and it takes at least six weeks for an effect be achieved. Although the usual recommendation is for an initial loading dose of two weeks of daily treatment, some women find this difficult due to discharge or even acute candidiasis, which can be a side effect of the use of topical oestrogen and is dose dependent. However, they often tolerate twice weekly treatment from the outset. Some women require more frequent dosing than twice weekly.

If dermatitis is present, 1% hydrocortisone ointment used twice daily should be used concurrently, and the patient should use a soap substitute. Bland emollients and non-irritating lubricants during intercourse are helpful adjuncts to treatment. Symptomatic oestrogen deficiency should respond promptly to topical oestrogen cream or pessaries within six weeks. If there is no response, consider an alternate cause for the dyspareunia.

The Approach to the Patient with Constant Vulval Pain

Taking the History

Most patients with vulval pain will not volunteer the symptoms that help to make a diagnosis. They will (for example) complain about 'reduced libido' instead of dyspareunia; similarly, a patient is unlikely to volunteer that her vulval pain started (for example) during

an episode of acute generalised psoriasis. A careful and comprehensive history is therefore essential.

1. Duration of current episode of pain
2. History of similar pain previously
3. Historical triggers
    a. Exacerbating/relieving factors
    b. Associated symptoms (e.g., itch, vaginal discharge)
    c. History of skin disease either vulval or elsewhere
    d. History of menstrual pain
    e. History of bladder pain
    f. History of irritable bowel or chronic constipation
    g. History of hip and/or back pain or injury
    h. Leisure activities (e.g., cycling, horse riding, skating)
4. Pain descriptors
    a. sharp/dull/burn/sting/formication/stabbing
    b. associated itch, bleeding or abnormal vaginal discharge?
5. Continuous or episodic
    a. length of episodes
    b. triggers for episodes
6. Pain location
    a. on/within the labia majora
    b. central
    c. bilateral/unilateral
    d. anterior/posterior
    e. referral patterns
7. Previous treatments
    a. effective
    b. ineffective
8. Impact on quality of life/sex/relationships

Systems Review

Because this type of pain is often referred, a complete systems review should be undertaken. Of particular importance is disease, dysfunction or injury to the lumbo-sacral spine, lower intestinal tract and anus, and lower limbs. This is because pain referral in the lower pelvis is usually *posterior to anterior*.

1. Bladder: irritable bladder, recurrent infection, dysfunctional voiding
2. Menstrual cycle/period problems
3. Bowel problems: irritable bowel, constipation
4. Musculoskeletal review:

a. congenital problems
b. injury
c. orthopaedic/neurosurgical operations
d. sciatica/other neuralgic leg symptoms
e. orthotics ever worn
f. foot/knee problems

Examining the Patient

Remember the golden rule: first exclude dermatological or infective causes.

Most of the patients referred to us because of vulval pain have a dermatological or infective cause that has been previously overlooked. We even see patients referred from sex therapists, who have realised that the woman in front of them does not have a psychological or relational problem. The prevalence of these missed diagnoses means that most articles on 'vulvodynia' mistakenly include many women who do not have true non-lesional vulval pain. This confusion is, again, why we prefer not to use this term.

Conversely, there are patients with observable skin disease in whom the skin disease is not responsible for their symptoms. Nonetheless, a trial of treatment for the observed skin condition must be undertaken to ensure that it has been confidently excluded as the sole cause for the pain. In these patients, dermatological treatment will be at best only partially helpful.

Effective therapy is only possible after an accurate diagnosis has been made.

Having excluded dermatological or infective causes for vulval pain, the presentations can be divided into two groups:

- pain on insertion only
- constant or intermittent pain not just related to insertion (although this may or may not be present as well).

Examination

Patients with vulval pain are rarely so uncomfortable that they are unable to be examined, but they may experience severe tenderness around the vestibule and in the vagina. It is often not possible to perform a speculum examination.

The most common local physical finding that causes acute vulval pain is a fissure. Fissuring may occur in almost any vulval dermatosis. The typical location is at six o'clock on the introitus. Even the tiniest fissures at the introitus may cause severe dyspareunia so it is important to look closely, especially in vulval sulcae and to gently stretch the skin. Usually there is an accompanying vulval rash such as dermatitis, candidiasis, psoriasis or lichen sclerosus. However, occasionally patients develop persistent vulval fissuring in the absence of an obvious cause.

Surgical and obstetric scarring often make it difficult to distinguish between normal and abnormal appearances on genital skin. An assessment by an experienced gynaecologist may be necessary in these cases.

The most important questions to have in mind during the examination are the following.

Is There a Rash or Lesion?

- Look very closely for tiny fissures by gently stretching the skin.

- If there is a visible abnormality, is it able to account for the patient's symptoms?
- If there is some abnormality, is this simply a normal variant or consistent with the patient's age and ethnic group (e.g., small labia minora in an elderly woman or pigmentation in a dark-skinned woman)?

Where Is the Pain?

- Is the pain unilateral or bilateral?
- Does the pain radiate (e.g., to the groins)?
- Can the patient localise the pain?
- Is there tenderness out of proportion to the degree of pressure exerted (we find it helpful to lightly touch the leg and at the same time touch the vestibule and ask the patient to compare the sensation)?
- Is there any introital muscle spasm on digital vaginal examination? This is indicated by tightness, resistance and pain on gentle downward pressure at six o'clock in the posterior vaginal introitus. (When there is no spasm, the examining finger easily enters the vagina without any resistance.)

Does It Relate to Any Observable Abnormality or Not?

If you have been reading other literature on dyspareunia, you may have noticed reference to the use of a cotton-tipped swab ('Q-tip'). We do not use this instrument, as we have found that it may actually cause pain. Furthermore, its use cannot distinguish between tenderness of the skin, or underlying structures. We find that the educated finger is more helpful.

Coloscopic examination after the application of acetic acid will make *any* inflammatory dermatosis appear white, and is therefore diagnostically unhelpful, unless neoplasia is suspected. Its application usually provokes intense pain and we believe that it is contra-indicated in this setting.

Classifying Patients Who Have Non-lesional Vulval Pain

Pain Case: Simple Neuropathy

A 79-year-old woman presents with many years poorly defined vulval burning pain. This became worse following a left total hip replacement, and even worse after a fall onto her sacrum. The pain is worse with sitting, but relieved by lying down.

On examination, the genital skin and vagina mucosa are normal to inspection. She has hyperaesthesia to light touch on the vulval skin. This is a case of vulval neuropathic pain, and demonstrates how hip and spinal dysfunction can promote it. Amitriptyline is commenced at a dose of 5 mg at dinner time, and increased by 5 mg/day/week to minimise side effects. By eight weeks the pain is largely controlled at a dose of 20 mg. An attempt to reduce the dose after three months is unsuccessful.

Neuropathic Pain

This is the most common aetiology in this category. The typical patient is a middle-aged to elderly woman who complains of a constant poorly localised burning sensation in the vulva. The pain is least severe in the morning after rest in bed and builds up during the day,

especially with physical activity. Certain positions may be more uncomfortable than others, particularly sitting. The pain is not always associated with dyspareunia, but most patients develop a distaste for sexual intercourse because of it. This sensation is often not severe. It rarely wakes patients at night. However, it is constant and exhausting.

Some patients also complain of bladder symptoms such as urgency and frequency. When questioned, they often also have or have had low back pain or sciatica or have had a spinal disc protrusion. Occasionally we see women who complain of unwanted sexual clitoral feelings. Some of these cases are also due to neural dysfunction, related to spinal dysfunction.

When patients are examined, there is often no abnormality other than some degree of atrophy consistent with their age. However, atrophy alone does not cause burning pain. Erythema is often noted, and may relate to loss of sympathetic control of skin vasculature. However, many patients have been treated with strong topical steroids for months in an attempt to treat the pain. This also causes erythema.

When you ask your patient to indicate the painful area, she will often indicate a horseshoe shape involving the perineum, introitus and lower labia majora bilaterally. It may radiate to the groin or inner thighs. In some patients, the discomfort is unilateral, or even confined to one small spot usually on the mucosal surface of the labia minora. In some, the area affected is peri-clitoral.

We believe that patients with this problem have neuropathic pain. The cause is still not understood.

Management

Medical Therapy

Like other forms of neuropathic pain, it is possible to obtain relief using oral medication. The most effective ones include:

- Tricyclic anti-depressant medication 10–50 mg of amitriptyline or nortriptyline daily, two hours before retiring. We find amitriptyline to be more effective, but nortriptyline to be better tolerated.
- Doxepin 10–30 mg nocte appears to be useful in patients whose main complaint is itch.
- Pregabalin 75 mg nocte initially, increased to 75 mg twice daily. If tolerated and then increased gradually up to 300 mg twice daily, depending on the response.
- Gabapentin 100 mg nocte initially, increased to 100 mg twice daily, and then slowly increased up to 600 mg three times daily. Again, the eventual dose depends on the clinical response.

It is very important when commencing tricyclic anti-depressants to start with a very low dose of 5 mg nocte and slowly increase the dose, taking care not to go beyond a level that the patient can tolerate. It is important to explain that side effects usually pass. Although many texts advocate the use of high dose tricyclics, our experience has been that if the drug does not work at low doses (20 mg) it is unlikely to work at high doses, and the side effects rapidly become intolerable.

The side effects of tricyclics include drowsiness, disorientation, dry mouth, blurred vision, constipation, hypotension, urinary hesitancy and conduction defects causing palpitations. The drug should be taken in the early evening to minimise drowsiness the next morning. They should not be used in patients with narrow angle glaucoma, a history of cardiac arrythmia or those on other medications that may interact.

Many patients have great difficulty with the fact that these drugs are also used as anti-depressants. A statement we often hear is 'I don't want to be a pill popper'. It is important to explain that you are not using the drug as an anti-depressant but as a pain reliever, and to mention that it is also used for migraine and bowel pain. Even so, patients are often very apprehensive and any side effect will cause them to give up quickly. They need to be encouraged to persevere as it is usually the initial two weeks that are the most difficult.

Gabapentin and pregabalin may be combined with low-dose tricyclics.

If a medication is effective, most patients who respond will notice obvious, if small, improvements in their pain within four to six weeks. The dose should then be slowly increased until no more pain is felt. This can take up to one year, although some patients feel better very quickly. Once the pain has been relieved, it may be necessary for the patient to remain on their medication indefinitely, although an attempt is usually made to reduce them once the pain has been adequately controlled for some months.

At the time of writing, we do not believe that there is sufficient evidence for the use of compounded topical neuromodulating agents. Amitriptyline applied topically is a local anaesthetic but has much more potential for side effects than xylocaine. An integral part of the effect of neuromodulating drugs is on spinal neurological function, which topical agents cannot change. Further, topical agents may cause hyper-sensitivity reactions, and they are expensive.

Exercise

Rest is an important component of treating this condition. Once patients feel better, they often become much more active than before, and this can cause the pain to become worse again.

On the other hand, carefully supervised exercises that lower spinal function as well as weight loss can make a very big difference and eventually allow withdrawal of medication. We recommend this if the patient is well enough and young enough, particularly with the help of a physiotherapist.

We see young women, however, who exercise so much that their core and pelvic floor become too toned and tight. These individuals often require down-training of their muscles to improve vulval pain.

Complementary Medicine, Counselling and Explanation

Some patients appear to benefit from acupuncture and chiropractic and some find that counselling is helpful, particularly if the counsellor has expertise in chronic pain management.

In some patients, an explanation of what is happening is all they need. The pain is not severe and they would rather live with it than take medication.

Neuropathic vulval pain is much better recognised than it has been in the past and neuropathic pain in general has started to receive much more attention in the medical literature. It is important not to dismiss a patient with chronic vulval pain just because they appear normal. If they apparently have a skin condition and topical treatment is not effective within a reasonable time, consider the possibility of neuropathic pain as the real cause of their problem.

Referred Pain

The nerve supply to the vulva originates from L1 and L2 and S2–S4 nerve roots (see Figure 8.1), via the pudendal and genitofemoral nerves. Compression or injury to these nerves may result in pain referred to the vulva, similar to sciatica.

Presentation

There is much overlap between this problem and neuropathic pain, but in this situation, the pain is often well-localised, unilateral or much more pronounced on one side; may have a shooting, electric shock stabbing or cramping component; and may have been of sudden onset. The patient is often much younger, and may present from early in the third decade of life.

History

When questioned, these patients may also give a history of low back pain, sciatica and may have a history of a back injury from lifting, sport and exercise, a motor vehicle accident or falling onto the coccyx. Episodes of vulval pain can be associated with low back pain. The widespread fashion for weight-lifting and 'high-intensity training' can also be triggers for referred vulval pain.

This sort of pain typically also has a burning quality in exactly the same way as neuropathic pain.

Management

It is important to rule out gross spinal pathology, although this is very unusual in our patients. Management of any ano-rectal problems is also essential.

Management of this type of pain is ideally with a physiotherapist who is skilled in both pelvis and spine. Neuromodulating drugs may be necessary. General measures such as lifestyle modification and weight loss will help.

The Role of Physiotherapy in Non-lesional Vulvo-vaginal Pain

Physiotherapy is an essential element of treating many patients with non-lesional vulval pain. In these patients, re-training of the pelvic floor muscles will reduce resting muscle tone and in turn reduce dorsal horn sensitivity. The role of physiotherapy is either as an alternative to medication or to allow an eventual withdrawal of medication. It has high levels of patient acceptance and the psychological support of the physiotherapist should not be underestimated. We recommend this if the patient is well enough and young enough to embark on it.

The physiotherapy history is similar to the medical history. Physical examination, however, concentrates on the musculoskeletal components of the spine, pelvis and lower limbs. A gentle but thorough assessment may take more than one session due to the high levels of anxiety and fear in this patient group.

Resting tone, trigger points and pain scores of pelvic floor, hip and relevant spinal muscles are assessed, as well as the presence or absence of prolapse and any apparent fascial defects.

A more generalised musculoskeletal assessment is also performed including assessment of gait and posture, generalised muscle tone and breathing patterns and specific examination of the lumbar spine and pelvic girdle plus identification of any trigger points in the abdominal, hip, gluteal and back muscles.

Physiotherapy Management

Education is an important first step. A clear explanation of the possible causes of their pain and the nature of chronic pain is essential. Behaviour and lifestyle modification such as correct posture, good back care, good bladder and bowel habits, vulval hygiene, avoidance of aggravating factors, stress reduction and incorporation of appropriate general exercise and relaxation activities/techniques into daily life is important.

Physical techniques aim to increase awareness and proprioception, normalise tone, improve muscle discrimination and relaxation, desensitise and increase tissue elasticity and reduce the fear of vaginal penetration. Techniques may include myofascial massage and trigger point release. Use of vaginal dilators may be employed both during sessions and as part of the patient's home programme. Once normalisation of resting pelvic floor muscle tone had been achieved, further attention may need to be directed to strength, endurance, co-ordination and timing of these muscles.

Treatment of co-existing lumbar spine and sacroiliac joint dysfunction via joint mobilisation and muscle energy techniques is often necessary.

Somatoform Disorder

Until recently, vulval symptoms were considered to be mainly psychological in origin. In our practice, this is a very uncommon cause of chronic vulval pain.

Patients with psychogenic pain are a very difficult group to diagnose and, in many cases, impossible to treat. It takes experience to recognise this condition, and even so it is inevitably a diagnosis of exclusion.

Presentation

Typically, the complaint is of constant pain, often with a burning quality, that does not fit easily with physical pain patterns. There is a bizarreness to the descriptions, which may change with each consultation. The pain is often not confined to the vulva and may generalise to involve the whole body.

Patients with psychogenic pain rarely come to the consultation alone. They are brought in by relatives who are desperate for an end to the misery that the patient inflicts on other family members. Often, they have seen numerous doctors and are angry and frustrated. When they do come by themselves, they often remark that their family has insisted that they see you.

Examination

When one attempts to examine such patients, they may be quite uncooperative.

This type of patient experiences pain as a manifestation of an underlying psychiatric illness. They may have a conversion disorder or may be malingerers who derive secondary gain from their problem by manipulating those around them, particularly their relatives. Malingerers do not wish to recover, as the pain is a way of satisfying their needs. It is their relatives who want a cure.

For this reason, this is a very difficult situation to treat. They need the help of a psychiatrist, but this is one doctor that these patients usually flatly refuse to see. They medicalise their problem, and as soon as they encounter a doctor who refuses to legitimise their medical model, they seek help elsewhere.

We occasionally see patients with this sort of pain who are engaged in a court case to sue a person whom they feel is responsible for their pain. Some have appeared to recover after their legal action is resolved.

Management

Patients with psychogenic pain are a very difficult group to diagnose and, in many cases, impossible to treat. It takes experience to be able to recognise this condition and, even so, it is inevitably a diagnosis of exclusion.

Typically, the complaint is of constant pain, often with a burning quality, that does not fit easily with physiological pain patterns. There is a bizarreness to the descriptions, which may change with each consultation. The pain is often not confined to the vulva and may generalise to involve the whole body.

Patients with psychogenic pain rarely come to the consultation alone. They are brought in by relatives who are desperate for an end to the misery and expense that the patient inflicts on everyone in the family, who are inevitably controlled by the patient. Often, they have seen numerous doctors and are angry and frustrated. If a patient does come by herself, she often remarks that her family has insisted that she sees you.

When one attempts to examine such patients, they may be quite uncooperative.

This type of patient experiences pain as a manifestation of an underlying psychiatric illness. These patients derive secondary gain and do not wish to recover as the pain satisfies their needs. It is their relatives who want a solution. A psychiatrist is one doctor that these patients usually flatly refuse to see. They medicalise their problem, and as soon as they encounter a doctor who refuses to legitimise their medical model, they seek help elsewhere.

Our approach is to help the relatives cope with the patient. It is best for both if the secondary gain in truncated. Unfortunately, the relatives are often unwittingly part of the problem, and patience and persistence are needed to convince them of the real nature of the illness.

Sexual Abuse

Many studies have demonstrated that 20–25% of women can recall sexual abuse as a child or adolescent. It has been shown that sexual abuse is associated with a lifetime risk of psychiatric disease and somatoform symptoms, drug abuse, phobia, depression and panic disorder. Child sexual abuse has been correlated with chronic pelvic pain but the question of whether this is a major factor in subsequent dyspareunia is unknown.

Childhood sexual abuse is a broad term that includes any exposure to sexual acts and does not always imply penetration, particularly in young children. It is seen in all counties and social classes and the perpetrators are usually known to and trusted by the child.

You should of course include sexual abuse as a possible aetiology for vaginal dyspareunia. However, it is best mentioned further on in the history-taking, when you have the sense that you have built up a trusting relationship with your patient. Many patients do not wish to talk about or admit to it, and they may have forgotten about it, so any questioning about it is inherently problematic. Further, an answer in the affirmative cannot be assumed to explain the patient's problem.

With regard to non-lesional vulval pain where there appear to be clear non-sexual triggers, delving into childhood sexual abuse may simply alarm or upset your patient and not contribute to management. In our practice, we enquire about only if there are no other obvious causes.

The Depressed or Obsessive Patient with Vulval Disease

Depressed patients may also experience pain as a manifestation of their depression. This may present as psychogenic pain alone, but a more common scenario is a patient with an uncomfortable physical vulval condition who copes poorly due to a concurrent depressive illness.

As a result, a physical condition that would normally be mildly painful is experienced as excruciating and the most common term such patients use to describe themselves is 'in agony'. This is characteristic of the catastrophising that often happens in depression.

Patients with obsessive-compulsive disorder may also find vulval conditions extremely difficult to cope with. Washing rituals may further irritate their skin. They may become irrationally focused on infection, particularly genital herpes and they may experience great difficulty touching themselves to apply creams or insert tampons.

They may insist on wearing pads and liners at all times because they feel that this is hygienic, which further exacerbates their skin problem. As a result, they may find themselves in a vicious cycle. Not surprisingly, anxiety and depression commonly worsen.

Presentation

Unlike patients with a somatoform disorder who are commonly angry, these patients present as sad and anxious. Even when they lack insight into their condition, they are usually receptive to the suggestion of psychiatric help and anti-depressant medication.

Any patient with a long-standing vulval problem, particularly where pain is part of the symptomatology, may become depressed and the depression will exacerbate the pain. For many patients, the concept of long-term control of a condition that cannot be cured is very hard to accept.

Dyspareunia and Pain on Tampon Insertion without any Other Abnormalities

In a patient with dyspareunia, it is essential to rule out a physical cause. However, if vaginal bacteriology is normal, there is no oestrogen deficiency and physical examination (including careful examination for fissures) is normal, we believe that this is due to muscle spasm in response to any attempt at insertion into the vagina. The question then becomes: what has caused the muscular spasm?

Examination

It has been found that patients with this problem have a high resting muscle tone in the pelvic floor as measured on surface electromyography. They experience severe pain in response to stimuli that would ordinarily be experienced as touch, pressure or stretch.

This can usually be appreciated on digital examination. In a relaxed patient, it is possible to insert a finger into the vagina without encountering resistance. In the typical patient with muscle spasm, your finger will meet with firm resistance and the patient will experience pain from even light pressure.

Management

In this setting, oral tricyclic anti-depressants and other drugs used to treat neuropathic pain are in our experience relatively ineffective.

Many articles in the medical literature suggest that patients with entry dyspareunia require psychotherapy. This is not true in everyone and such a referral should be made on a case-by-case basis.

In patients with anxious personality, relationship problems or a history of sexual abuse, such referrals are an essential part of management. However, many just require very concrete advice on how to overcome muscle spasm and quickly overcome their problem. In our experience, psychotherapy alone without the pelvic floor advice is doomed to failure.

It is not known whether the pain causes the muscle spasm or vice versa, but expert pelvic physiotherapy can relieve this pain and often makes it possible for these patients to overcome their problem.

There are many pelvic floor physiotherapists who understand this type of dyspareunia and treat it effectively. These are our first line of management and one that patients welcome.

It is important to explain the mechanism and reassure very strongly and positively that this condition can be overcome with straightforward physiotherapy. We explain the nature of spinal reflexes such as the knee jerk response and find that patients can easily relate to this.

Patients who appear to have developed this as a result of a result of a skin condition should be warned at the outset that their dyspareunia may not resolve as quickly as their other symptoms.

There are patients in whom the prognosis has to be very guarded indeed. This group has primary dyspareunia, have never been able to insert anything into the vagina without severe pain and are angry, resentful, frustrated and have invariably seen numerous doctors without improvement. They have had the problem for many years, have failed physiotherapy entirely, and it seems that their condition serves a purpose in their lives, albeit a dysfunctional one.

Interestingly, they usually have partners who support them and come with them to one doctor's appointment after the other. In this difficult-to-manage group, the possibility of a somatisation disorder has to be considered. However, this is not easy to prove. It is possible that these patients may benefit most from a long, supportive therapeutic relationship with a doctor.

Surgical Treatment for Vulval Pain

There are three surgical approaches that have been advocated in the medical literature:

- Fenton's procedure, where the superficial muscles of the posterior introitus are incised to increase the dimensions of the introitus
- vestibulectomy, where the sensitive area of the introitus is excised, and the lining of the vagina is undermined and advanced to cover the area

- intra-muscular botulinum toxin.

Fenton's procedure is a very old operation, which is based on the notion that introital muscle tightness can be overcome surgically. We consider this notion to be incorrect, and never recommend this procedure.

The rationale for vestibulectomy is that the inside of the vagina is anaesthetic, and replacing sensitive introital skin with vaginal lining will eradicate the trigger point that sets off the muscle spasm. Advocates of this procedure claim a very high success rate where all other treatments have failed. The difficulty is that patients in these studies may have been characterised as having 'vulvodynia' without being further categorised and are therefore a heterogenous population. In our practice, we have never needed to resort to this procedure. We are of the opinion that vestibulectomy may not be necessary when an accurate diagnosis leads to effective (non-surgical) treatments.

Intralesional botulinun toxin, which is injected directly into the levator ani muscle, is used to temporarily paralyse the muscle that is in spasm, causing the pain. It does appear to be effective as an adjunct to physiotherapy, enabling patients who have a great deal of trouble releasing the pelvic floor, to progress with exercises. This therapy is still in an experimental stage. The effect lasts about three months, and the injections have to be repeated. Side effects include the possibility of urinary and faecal incontinence and the cost is not inconsiderable. At this stage, we still see it as something to be considered when more conservative therapy has failed.

Diagnosing Vulval Pain

Take a detailed history to determine:

the nature and timing of the pain

whether the pain is only associated with intercourse, or is present at other times.

Examine the patient to determine whether:

there is a lesion, fissure or rash

the vulva is essentially normal.

Investigations:

take a low vaginal swab in every case

biopsy may be required

treat any objective abnormality.

If the vulva appears normal and swabs are negative, the patient is likely to fall into one of the following groups:

introital hyperalgesia: pain only with intercourse

neuropathic pain: bilateral constant burning

referred pain: burning pain, more pronounced on one side, history of back injury

somatisation disorder: bizarre history and affect

See the diagnostic algorithm in Figure 8.2.

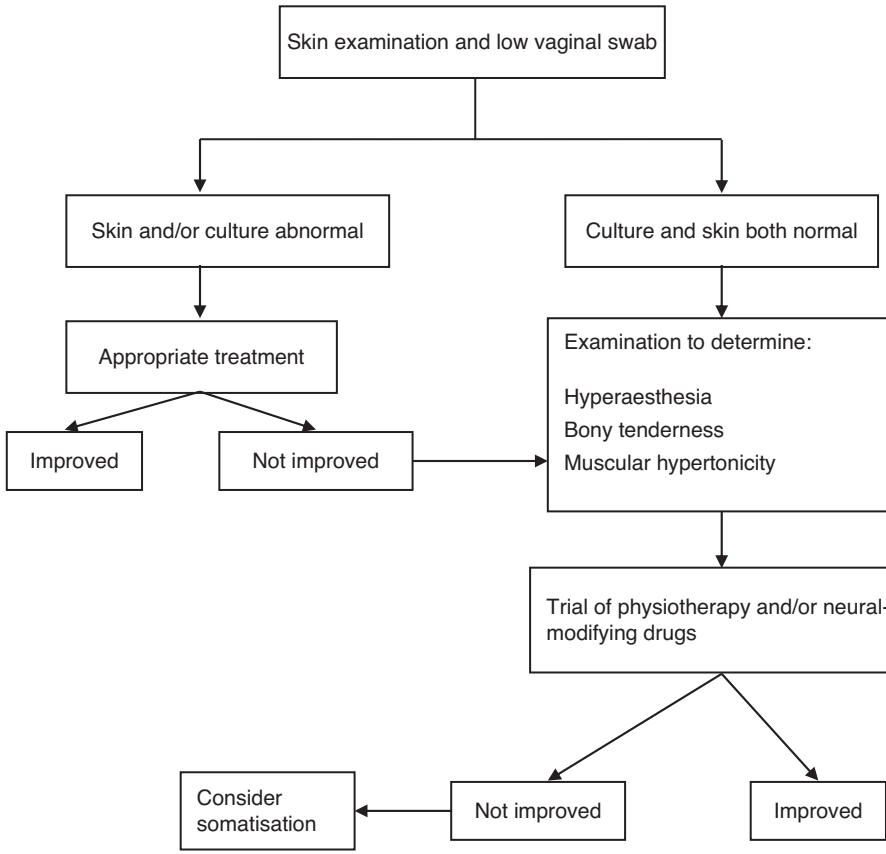

Figure 8.2 Diagnostic algorithm for vulval pain

Acknowledgement

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Chapter

9

Vulval Disease in Children

Contents

When a pre-pubertal girl presents with an itchy or sore vulval rash, she is usually assumed to have thrush or a urinary tract infection. Poor hygiene or sexual abuse may also be considered. In fact, none of these are likely to be true.

For a patient to experience symptoms from candidiasis, the vagina must be oestrogenised. It therefore does not occur in pre-pubertal girls. Urinary tract infections do not result in rashes unless prolonged incontinence is present (although contact of urine with inflamed skin may cause stinging), sexually abused children rarely have physical signs and over-zealous hygiene is more likely to produce a rash than lack of hygiene.

Vulval disease in children is less common than in adults. In both adults and children, dermatitis, psoriasis and lichen sclerosus (LS) are the most common dermatoses that cause a chronic vulval rash. Infective vaginitis is rare in children. Group A beta-haemolytic streptococcal vulvo-vaginitis is overwhelmingly the commonest infective vaginitis in children, but in adults it is seen only sporadically.

Birthmarks, particularly haemangiomas, of the vulva are an important issue in children, but not in adults. Fusion of the labia is a self-limiting condition seen in small children, but in adults is seen only in the setting of LS or lichen planus.

Sexual abuse is always an issue to be considered in any genital presentation in children, but in fact is rarely a cause of vulval disease.

Older texts often blame paediatric vulval disease on the theory that non-oestrogenised pre-pubertal vulva skin must be fragile and sensitive. In fact, there is no evidence to support this. The incidence of vulval skin disease in children is much lower than in adults, and this implies that their vulval skin is less prone to disease. The use of oestrogen creams as a speculative treatment in this setting therefore has no basis in fact. Indeed, oestrogen creams are often very irritating when applied to children's skin. Prolonged use of anti-fungal creams also has no basis in fact and can also be irritating.

The low-oestrogen environment of a child's vulva is physiological, not pathological. In fact, oestrogenisation in adult women is a liability that predisposes to vaginitis, particularly candidiasis.

Another common assertion is that vulval disease in children is due to poor hygiene and faecal contamination. This facile statement trivialises and stigmatises this problem. In fact, mothers of small girls, particularly those with a vulval problem, are usually highly conscientious about genital hygiene and are more likely to be doing too much washing, rather than too little.

Parents often arrive in your office in a state of defensive high anxiety. They are embarrassed, ashamed, worried that they have an abnormal child and frustrated with treatment that has been ineffective. They may have fears of child abuse and mortified by their child having been sent home from school because of scratching behaviour.

What Causes Itchy Rashes in Prepubertal Children?

The common causes of rashes of the vulva comprise the usual dermatoses of the prepubertal age group: dermatitis and psoriasis, and the rare condition LS.

Dermatitis

Most who suffer from vulval dermatitis, both adults and children, are atopic.

The vulval skin of babies is remarkably resistant to disease, even though they are in nappies. Babies who suffer from atopic dermatitis rarely have signs of it in the well-hydrated skin under the nappy, and the onset of vulval dermatitis is often delayed until the child is out of nappies.

The resistance of genital skin appears to be lost in the older child, where the ongoing wearing of nappies, for example, overnight in those with enuresis, starts to cause problems with irritant dermatitis similar to incontinent adults who wear absorbent garments.

The most common causes of irritant contact dermatitis in children are over-use of soap and bubble bath, using shampoo in the bath, swimming in chlorinated pools and wearing occlusive clothing, often ballet and athletic gear. Children who shower rather than bathe may miss washing the vulval area effectively.

Irritant contact dermatitis may occur as a result of contact with faeces, usually in the context of diarrhoea, or chronic constipation with soiling. Dysfunctional voiding may result in pooling of urine in the vestibule leading to maceration, which can also be irritating.

Irritation from over-use of topical medications and perfumed products is very common in adults but less so in children.

Presentation

Vulval dermatitis presents with itching and a fluctuating rash, often precipitated by contact with irritants. The child's scratching behaviour is often a source of embarrassment for her parents and causes unwelcome attention at school. It is common for children with vulval itching to wake in a distressed state at night with night terrors.

Examination

Examination is often fairly unremarkable, and parents may have trouble convincing a doctor that there is anything wrong. Close inspection will reveal some erythema, scale and slight rugosity of the labia majora, increased erythema and desquamation of the minora. The desquamation may stain the child's underwear and be misinterpreted as a vaginal discharge. If the rash is severe, it may extend to the inguinal areas and buttocks. It may become excoriated from scratching. Superinfection with *Staphylococcus aureus* may occur on the skin, but there is no true vaginitis and vaginal swabs and urine cultures are negative. However, in the setting of chronic wetness, the vestibule may appear inflamed and red.

Investigation

A skin swab should be performed if super-infection is suspected. Vaginal swabs are not necessary in pre-menarchal girls. A urine culture will rule out a urinary tract infection if urinary symptoms are present.

Management

Treatment of vulval dermatitis is easy if environmental modification can be put in place. The first step is to modify the environment. A bath is preferable to a shower, with no soap or bubble bath. Bland bath oil can be used. Shampoo should be rinsed out after the child gets out of the bath, or a soap substitute used instead of shampoo. If the child does shower, a soap substitute should be used and the parent needs to supervise parting and rinsing the labia.

If the child does any physical activity that involves wearing occlusive stretch clothing, this should be modified so that at least during practice sessions, loose cotton clothes are worn. Even nylon tights worn as part of a school uniform may have to be discarded, and you may need to give the parents a letter to take to the school. Some recreational activities that involve contact with a saddle can also be very irritating.

When it comes to clothing, loose cotton underwear is ideal, and underpants, particularly nylon ones, should be avoided in bed.

The chlorinated water in heated swimming pools can be a powerful irritant. Applying white soft paraffin or zinc cream before swimming is helpful. The bathing costume should be removed immediately after swimming, and the skin rinsed in the shower before going home.

Always actively ask about whether nappies (diapers) are being worn at night, and about urinary or faecal incontinence. Parents may be too embarrassed to mention them, but these issues must be addressed.

Ask about the use of over-the-counter topical preparations, as parents may not volunteer this information. The inappropriate use of anti-fungal creams, perfumed toilet paper and

wet wipes containing methyl isothiazolinone may all cause irritation, and should be ceased. Latex can also be a problem as some parents unknowingly sensitise their children when changing their diapers while wearing latex gloves.

Specific Treatment

Most cases of vulval dermatitis will respond to 1% hydrocortisone, as long as the environmental changes have also been made. Ointment is preferable to creams, which may cause stinging. If the dermatitis is severe, a stronger non-fluorinated topical corticosteroid may be used for a week or two.

Some topical corticosteroid preparations, particularly mometasone furoate, have a tendency to sting on the vulva and they should be avoided. Although adults can cope with this side effect, children rarely do. If a more potent corticosteroid has been used, it should be possible to reduce to 1% hydrocortisone once the rash has settled. If this is not possible, one should consider an alternative diagnosis.

Many parents are very apprehensive about using topical corticosteroids on their children, and even more so on the vulva where are they are concerned that the preparations will thin the skin. In practice, the treatment is very safe and it is wise to pre-empt any objections with strong reassurance and a warning that the pharmacist, the naturopath and well-meaning relatives may well recommend caution regarding their use.

In summary, we recommend the following:

- avoid soap and bubble bath
- shampoo hair in the shower not the bath
- shower after swimming lessons and do not go home in the wet swimming costume
- avoid perfumed wet wipes and toilet paper
- try to dispense with night nappies (diapers) or pull-ups as soon as possible
- pay attention to constipation with overflow and dysfunctional voiding
- avoid nylon ballet clothes and similar garments at other activities
- ask yourself whether sporting activities that irritate the vulva (cycling, horse riding, ballet) are worth it
- avoid nylon tights in winter
- wear cotton underwear
- do not wear underwear in bed at night
- never use anti-fungal creams
- use a simple non-perfumed moisturiser every day.

If skin swabs show infection, (usually with *S. aureus*) a course of appropriate antibiotics should be given. A finding of beta-haemolytic *Streptococcus* group A requires a 10-day course of penicillin, cephalexin or, for those allergic to the former, an antibiotic such as erythromycin, providing the organism is sensitive.

Case Study

An eight-year old girl has suffered from chronic vulval itching for the past year. At times it fluctuates, becoming more severe and waking her at night. She is quite distressed when she wakes and her mum has to put her in the bath to settle her down. At the time, her skin looks quite red but in between these attacks it appears normal.

She has already been investigated with swabs, which were negative, and urine culture, which was normal. Treatment with anti-fungal creams has not helped her, although there is some relief from 1% hydrocortisone.

She has a history of cradle cap and nappy rash as a baby, and her mum has psoriasis.

On examination, she has subtle but definite erythema and scale involving both labia majora, the perineum and perianal skin. You notice she also has scale and papules on the dorsum of her knees and elbows and a scaly scalp.

This girl has psoriasis. During exacerbations she needs a more potent topical corticosteroid than 1% hydrocortisone. Methylprednisolone aceponate 0.1% provided rapid relief. Once she is better, a low-potency tar ointment such as 2% LPC (liquor picis carbonis) in a bland base such as emulsifying ointment should be used preventatively on a daily basis. It is important to explain to her parents that this is a chronic but benign condition that may flare from time to time during intercurrent illnesses and at times of emotional and physical stress.

Psoriasis

Although the classic age of onset of psoriasis is early adulthood, psoriasis in children is not uncommon. If children with vulval disease are taken as a group, psoriasis is a very common cause of chronic vulval and perianal itchy rashes.

Presentation

In babies, psoriasis may present for the first time as a persistent nappy rash. The features at this age include a well-demarcated edge and involvement of the inguinal folds, but the typical scale of psoriasis can be lacking under the nappy. Macerated flakes of skin in the inter-labial sulcus may give the impression that there is a discharge. Although psoriatic nappy rash may respond to standard treatment with 1% hydrocortisone topically, it is not unusual to need a stronger topical corticosteroid.

In older children, the rash is more typical of psoriasis, with an itchy, red, well-demarcated, symmetrical plaque. Again, there is little scale, however, maceration with a build-up of white material in the inter-labial sulcus is typical. The vulva, perineum, peri-anal area and often natal cleft may all be involved. In older children, 1% hydrocortisone is also often ineffective.

If psoriasis is confined to the vulva, it is difficult to make a definite diagnosis unless there are other diagnostic clues present. A history of cradle cap or difficult nappy rashes as a baby, nail pitting, post-auricular or scalp rashes and a family history are all helpful. Signs can, however, be subtle and psoriasis in children is much more subtle than in adults and is often mistaken for eczema.

Management

Vulval psoriasis is more challenging to treat than vulval dermatitis. Although the environmental modification described is essential, more potent topical corticosteroids than 1% hydrocortisone are often required, as well as tar preparations. In this situation, referral to a dermatologist is recommended.

We usually commence a medium-potency corticosteroid ointment (preferably non-fluorinated) at night, continuing until the itch has been relieved. We then add a weak tar cream, 2% LPC in a thick emollient base to be used every day while reducing the strength of the topical corticosteroid back to 1% hydrocortisone.

The reduction in corticosteroid potency must be done gradually by initially alternating the weaker and stronger preparations, and slowly using less of the stronger one and more of the weaker one.

Most children can eventually be controlled on the weak tar cream alone. If this is not tolerated, usually because of stinging, we introduce calcipotriol 0.05% ointment for long-term control. If this is also problematic, a greasy emollient often suffices.

Case Study

A six-year old girl presents with a six-month history of vulval itch and pain. This has been accompanied by dysuria and constipation. The appearance of her vulva has resulted in a query of child abuse. You are asked whether there should be a child at risk report.

She has been seen by a paediatrician regarding the cause of the dysuria and constipation. A urine culture was negative and she is on a laxative regime, which has not been successful.

On examination, she has the typical white plaque of LS surrounding the vulva and perianal skin. It is complicated by purpuric blebs and fissuring, which are part of this condition, and she has a clitoral phimosis.

She is commenced on betamethasone dipropionate 0.05% in optimised vehicle daily. Within two weeks, her symptoms have resolved, however, it is another three months of daily treatment before her appearance normalises and the phimosis releases. She can then be treated with a less potent topical corticosteroid such as methylprednisolone aceponate 0.1% daily, gradually reducing the potency of the medication, just as one would do with an adult.

Follow-up should be every three months until her condition is stable, and follow-up is indefinite. It is particularly important that as she approaches puberty, treatment continues to prevent the possibility of vulval scarring.

Lichen Sclerosus

Lichen sclerosus may start at any age. It is relatively rare in children, however, if children with vulval disease are examined as a group, about 10% of them will have it.

Presentation

Lichen sclerosus may occur at any stage of childhood but the average age is from five to seven years Because it is unfamiliar, there is commonly a delay of diagnosis of at least one year. Girls with LS tend to present with more complex symptoms than adults. Itch is not always a prominent feature, and soreness, dysuria, bleeding and chronic constipation may occur. Night waking and night terrors are common. Not surprisingly, these children are therefore often investigated for bowel and urinary tract abnormalities and sometimes referred to child protection units.

The clinical appearance is of a well-demarcated white plaque with a wrinkled surface and scattered telangiectasia. Fissuring is common, causing bleeding from time to time. The typical distribution is of a figure of eight plaque surrounding the vagina and anus, but any pattern on the vulva, perineum or peri-anal area may be seen. The vagina is not involved and extra-genital involvement is very rare in children. Unless the labia majora are parted and a careful inspection made, it can be easy to miss this condition.

As in adults, LS in children can be complicated by loss of the labia minora and clitoris, which becomes buried under scar tissue. This is as common in children with LS as in adults with LS. The commonest distortion of vulval architecture is clitoral phimosis, easily missed unless it is sought. However, diminutive or missing labia minora are also seen often. With the delay in diagnosis, it is common for these changes to already be present on presentation.

Many children with LS are suspected of having been sexually abused. This relates to the unusual and unfamiliar appearance of the rash, particularly where purpura or bleeding is present. It is important for practitioners to realise that this is a skin condition. Its presence of course does not rule out abuse, but the disease itself is not a cause to suspect it.

Management

Lichen sclerosus in children is managed in exactly the same way as in adults, commencing with a super-potent topical corticosteroid and gradually reducing to the lowest strength that affects long-term control (see Chapter 3).

Parents often express fear of using such products on the vulva and are frequently cautioned by their pharmacist as well. A great deal of reassurance is required and a warning that the pharmacist may express reservations. We impress on parents the importance of effectively treating LS in their child to relieve symptoms and prevent loss of vulval structure. This is particularly important as girls approach puberty in order to provide the best chance of developing normally.

This diagnosis can be emotionally crippling to some families, who find the concept of their child having an incurable condition that will require life-long treatment of the genital area devastating. We encounter anger and denial at first in many but most eventually come to an acceptance of the need for treatment. Some parents are squeamish about applying treatment and negative parental attitudes are rapidly picked up by young patients. Treatment refusal follows and many parents need much emotional support to push through the temptation to allow children to apply or dictate how often they will use their medication.

There was once controversy about whether LS remits at puberty but in most cases it does not. Although symptoms may settle, silent progression with scarring may occur, and symptom activity may recur in adult life. Adolescent girls may be too embarrassed to allow anyone to examine their genital area. It is very important, for this reason, to build up a trusting relationship with a child with LS, and to start warning her from about the age of eight that when she is older, she will need ongoing check-ups and treatment.

We believe that when LS occurs in a child, it should be treated as aggressively as in adults. Although it is relatively easy to relieve symptoms with as needed topical corticosteroid application, the ideal outcome of management should be to preserve vulval architecture as well. Our most recent research shows that patients who do not keep the disease suppressed as they go through puberty are at least 25 times less likely to reach adulthood with normal vulval architecture than those who maintain objectively normal skin with continuous daily corticosteroid ointment. Inadequate treatment puts the patient at risk, not only physically but also psychologically, especially in adolescence.

The psychological impact of childhood LS should also not be under-estimated. About half our young patients are too scared to use tampons and many have unvoiced fears about

sexuality and reproduction. The unavoidable intrusion into their autonomy related to doctor's visits and application of ointment takes a toll. It is not surprising that treatment refusal commonly becomes an issue at adolescence at a time when it is so important to maintain good compliance.

In addition to this, there is a worrying association (about 2–6%) with squamous cell carcinoma of the vulva in untreated LS in adult life. This has been reported in relatively young women who have had LS since childhood. Therefore, this condition should be actively managed, and follow-up should ideally be life-long. Referral to a dermatologist is recommended for initial assessment.

Birthmarks of the Vulva

Birthmarks may occur on the vulva, as on any other part of the skin, but the importance of lesions in this location is that they may be mistaken for more sinister conditions.

For example, pigmented naevi on the vulva often raise queries of melanoma, where they might be ignored elsewhere, and epidermal naevi, which are rare and therefore not familiar to general practitioners, are often mistaken for warts or recalcitrant eczema.

Haemangiomas of Infancy

Haemangiomas of infancy are the most common birthmark and are found much more commonly in females than males. They can be located anywhere on the skin but may be located on the vulva and peri-anal area, where they have special features.

- They are prone to ulceration, which can be so extensive that it can be difficult to tell that there was ever a birthmark present.
- They may be mistaken for a sign of sexual abuse, particularly if ulcerated.
- If extensive, they may be associated with abnormalities of the bladder, bowel and lumbosacral spine.

The diagnostic feature of a typical haemangioma of infancy is the bright red colour, well-defined edge and the age of onset, which is in most cases is within the first four weeks of life. The lesions grow for a variable period, up to about four months of age and then regress.

Management

In many cases, no management other than reassurance is required and, like haemangiomas elsewhere, these lesions eventually resolve. It is rare for there to be significant sequelae unless the lesions are very large or ulcerated. Large or ulcerated vulval lesions can cause severe morbidity, and should be urgently referred to a dermatologist.

When ulceration occurs, it is not uncommon for a query of sexual abuse to be made. It is most important not to jump to such conclusions where there is ulceration of a haemangioma, and to seek the opinion of a dermatologist or paediatrician.

Oral propranolol is the treatment of choice for complex, ulcerated or disfiguring haemangiomas. Treatment of ulceration also involves the use of ulcer dressings. Topical beta-blockers have also been shown to be effective in small, flat haemangiomas but these lesions on the vulva are usually best left in the knowledge that they will resolve spontaneously.

All complex or ulcerated haemangiomas of infancy should be referred for urgent dermatological assessment.

Naevi

Pigmented naevi may occur on the vulva either as a congenital lesion or one that appears at any stage of childhood. The congenital lesions may be larger than late onset ones.

It is normal for children to acquire naevi at any age on any part of the skin, and the vulva and peri-anal area is no exception. However, melanoma in children is rare, and there have been very few reports of childhood vulval melanoma.

The same principles apply as on any part of the skin: if a lesion has benign features, such as symmetry, even colour, growth stability and benign features on dermoscopy, it can be safely observed. Pigmented naevi of the vulva do not have any more significant malignant potential than those elsewhere and, again, can be safely observed. It is worth noting, however, that benign *genital* naevi can have histological features that suggest malignancy. The site of the lesion should always be specified when an excision specimen is sent for histopathological examination.

Epidermal naevi are quite rare, and are not always present at birth. They commonly have a warty surface and may be arranged in whorls or streaks. Lesions that involve the vulva can be part of a larger one that extends to the leg and buttock.

They are sometimes very itchy and have a tendency to extend and become more raised with time and, if they become large, can interfere with function. Because of these features, epidermal naevi can be mistaken for warts, which in turn gives rise to queries of child abuse. If they are itchy, they can be mistaken for treatment-resistant eczema or nappy rash.

It is not uncommon for epidermal naevi of the genital region to cause enough trouble to require at least partial excision. For example, a warty peri-anal lesion is best removed. Sometimes recalcitrant itching is only relieved by removing the lesion.

However, if they are not causing problems, it is best just to reassure and leave them alone. They have no malignant potential.

Vulvo-vaginal Infections in Children

Acute Infective Vulvo-vaginitis

Group A beta-haemolytic *Streptococcus* can cause a low-grade, persistent peri-anal rash (streptococcal perianal dermatitis) and also acute vulvo-vaginitis. This is the most common cause of acute vulvo-vaginitis in pre-pubertal children. It is virtually never seen in adults, although the same organism can cause vulval cellulitis at any age.

Presentation

Streptococcal vulvo-vaginitis presents with sudden onset of an erythematous, swollen, painful vulva and vagina, with a thin mucoid discharge.

There may have been a preceding throat infection with the same organism, or preceding perianal dermatitis. Sometimes the infection can be low grade, similar to the peri-anal disease, presenting as a sub-acute vulvitis.

It is believed that the portal of entry is the throat and the organism reaches the vagina by haematogenous spread.

Occasionally, streptococcal genital infection may precipitate guttate, genital and or peri-anal psoriasis. This is a real trap for the unwary as it is usually interpreted as recurrent infection. A swab should always be taken if recurrence is suspected and, if it is negative, think of psoriasis.

Investigation

The infection is easily diagnosed by introital and peri-anal swabs It is not necessary to insert the swab right into the vagina, which children usually find traumatic, particularly when the area is tender.

Although a differential diagnosis of acute candidiasis would be reasonable in an adult, this is not the case in children. Pre-pubertal children do not suffer from vulvo-vaginal candidiasis. Recurrent streptococcal infections should raise the possibility of an intra-vaginal foreign body or chronic pharyngeal carriage.

Prior to immunisation, *Haemophilus influenzae*, another respiratory pathogen, was also implicated in acute vulvo-vaginitis in children. This is now very rare.

Any case of acute vulvitis in a child should suggest streptococcal vulvo-vaginitis, not acute candidiasis.

Management

After swabs have been taken, the child should commence on either oral penicillin, or amoxycillin or cephalexin if they are allergic to penicillin. Other antibiotics are used based on sensitivities. The course must run for a full 10 days or recurrence may occur. Studies have shown that also using topical mupirocin twice daily reduces the risk of recurrence.

Staphylococcal Folliculitis and Impetigo

Staphylococcal folliculitis is common on the buttocks of children, particularly those with eczema and those who are still in night nappies. It may sometimes spread to the vulva or be found there primarily. Impetigo may also sometimes occur on the vulva and peri-anal area.

Presentation

The presentation is with pustules and crusted lesions, which are often more itchy and irritating rather than painful.

Investigation

The diagnosis is confirmed with a skin swab.

Management

Although impetigo usually responds quickly to a course of appropriate oral anti-staphylococcal antibiotics, folliculitis can be very persistent, and is often better treated by also using with topical agents such as mupirocin 2% ointment twice daily for a week. Staphylococcus carriage in fomites must simultaneously by treated by adding a quarter of a cup of household bleach to the bath water and hot washing clothes, sheets and towels with hot water.

Every attempt should be made to discard night nappies. If there is underlying eczema, this should be treated.

Pinworm

Although many children with pinworm infestation are asymptomatic, symptoms are that of peri-anal and vulval itching, particularly at night. An eczematous rash may occur.

Pinworm is very well known as a cause of genital itching in children, and many children with vulval disease will already have been treated with mebendazole by their parents, prior to seeing a doctor.

Molluscum Contagiosum

These viral lesions are very common in children. The virus is spread in water. Many children are infected at public swimming pools and then transmit the infection to younger siblings with whom they share a bath. As a result, it is not uncommon for mollusca to be found on the vulva, often as part of a more generalised eruption.

Sometimes vulval mollusca can be difficult to differentiate from genital warts, and close examination with a magnifier will be needed to see the typical central core. This distinction is important, as paediatric mollusca are not necessarily sexually transmitted, but paediatric genital warts should raise a strong suspicion of sexual abuse. There are four separate mollusca genotypes and studies have shown that the ones that cause genital lesions in adults are different to the ones that are usually found in children.

Management

In most cases, it is not necessary to treat vulval mollusca as spontaneous resolution invariably occurs within two years. Methods used to extract the viral core from the centre, which are tolerated on less sensitive areas, may be too painful on the vulva.

Genital Warts

Genital human papilloma virus lesions are very uncommon in children. Genital warts should raise the question of sexual abuse and many experts recommend that a consultation with a child protection unit should be arranged. This is invariably traumatic for the family and unfortunately it is often difficult even for experts to draw a firm conclusion. Unless either the child or a parent discloses that sexual abuse has occurred, no action can be taken unless sexually transmissible infection screening reveals an infection that is definitively sexually transmitted, for example, gonorrhoea.

There are many articles in the medical literature that state that there are many ways genital warts can be acquired in children other than sexually, but we do not see the logic in asserting this in children whilst assuming that they are invariably sexually acquired in adults. Unfortunately, this remains an area of controversy. There is probably truth in the contention that not all genital warts in children are sexually acquired but convincing research to support how common this is relative to sexual transmission is lacking.

Presentation

Genital warts typically have a filiform appearance and may involve the vulva, vaginal introitus and peri-anal area. These lesions are usually small, but can occasionally become large enough to interfere with toilet routines.

Management

Imiquimod and podophyllotoxin may be safely used in children and are preferable as first-line treatments to painful modalities such as cryotherapy or cautery, which require a general anaesthetic.

In children with very extensive peri-anal warts, surgical debulking may be required before this treatment is commenced.

Genital Herpes

Herpetic lesions of the vulval area are very uncommon in children. In infancy, herpes can be acquired at birth from a mother with genital herpes, or inoculated from an adult with a herpetic lesion on the mouth or finger.

Children with severe atopic eczema are also prone to unusual herpetic infections in unusual locations and this may involve the genital area.

Both herpes simplex virus 1 (HSV1; usually associated with cold sores) and herpes simplex virus 2 (HSV2; usually associated with genital herpes) can be isolated from genital lesions.

Presentation

The clinical findings are the same as in adults: painful vesicles that rapidly erode and ulcerate, associated with lymphadenopathy. There is sometimes some degree of oedema of the labia.

Investigation

The finding of a primary attack of genital herpes in an older child should be confirmed by polymerase chain reaction testing (PCR), and should raise the possibility of sexual abuse, particularly if HSV2 is isolated. The subject of whether a child with herpes of the genital region has been sexually abused is even more difficult than with genital warts because there are a number of innocent ways that a child might acquire genital herpes, particularly HSV1.

The differential diagnosis of herpes includes two very uncommon conditions: vulval aphthous ulcers and vulval bullous pemphigoid (see Chapter 4). Both present with painful ulcerating lesions, and it is not surprising that they are often mistaken for herpes. Herpes PCR is, however, negative, and sexual abuse should not be suspected on the grounds of these lesions.

Management

Genital herpes in children is treated in the same way as in adults, using oral anti-viral medication.

Fungal Infections

Tinea is a common cause of groin rashes in men, sometimes causes vulval rashes in women, but is rarely found on the vulva in children. When it does occur, it hardly ever has typical features, and is often the result of treatment with topical corticosteroids. It may be more common than one would think, as so many cases of vulval rashes are treated with imidazole creams, which fortuitously also treat tinea.

Presentation

In cases where no anti-fungal has been used, tinea of the vulva or under the nappy in a baby presents as a dermatitic rash that does not respond to topical corticosteroid treatment.

Investigation

The diagnosis requires a high index of suspicion but, once suspected, is easily confirmed by a skin scraping.

Candidiasis, on the other hand, does not occur in children out of nappies. In adults with chronic vulval symptoms, about 15% have candidiasis, but this oestrogen-dependent condition is not seen after infancy in children with normal immune systems.

This is an important point, as it is common for children with skin diseases such as dermatitis and psoriasis to be diagnosed with thrush and treated with anti-fungal creams, which may cause irritation, particularly if dermatitis is present.

Anatomical Abnormalities

Fusion of the Labia Minora

Fusion of the labia is sometimes seen in young children, usually of three years of age and under. It is not a congenital malformation and is acquired. The aetiology is unclear, but may be related to vulval dermatitis.

Presentation

Not all are symptomatic, but some experience soreness or itching. Urine can pool behind the fusion, causing irritating maceration as well as slow urination. Urinary tract infections are, however, rarely a complicating factor.

The labia minora or majora are agglutinated to a variable degree from the tip of the clitoris to the posterior fourchette. This may result in an abnormal-looking vulva with no apparent vaginal opening.

Investigation

Fusion of the labia is important in the differential diagnosis of ambiguous genitalia and imperforate hymen, and a specialist opinion should be sought if there is any doubt.

Management

This is the only condition where oestrogen cream is the treatment of choice in a pre-pubertal child. The cream need only be applied once a day and the fusion usually resolves over a two-

to six-week period. Once the fusion has separated, ongoing treatment with soap avoidance, topical lubricants and 1% hydrocortisone is recommended.

The fusion may reform and have to be retreated from time to time. This can be a problem as oestrogen creams are irritating in children and make co-operation difficult.

Very occasionally, a minor surgical procedure to separate the agglutinated labia may be required. The condition tends to resolve spontaneously in older children.

Pyramidal Perineal Protrusion

Although this has only recently been labelled as an entity in the medical literature, it is probably not rare. It is noticed in infancy as an asymptomatic soft protrusion of the median raphe in girls. The overlying skin is normal. This condition can be confused with genital warts.

The aetiology of this condition is unknown, there are no consistent associations with other conditions, it is not consistently the result of constipation and it resolves spontaneously.

No treatment is required.

Foreign Bodies

Although intra-vaginal foreign bodies are often mentioned as a cause of vulval disease in the medical literature, in fact, they are not a common event. The foreign material is usually a fragment of toilet paper or fluff. Small toys are less common.

The child presents with a persistent purulent discharge heavy enough to cause maceration of the vulval skin. Swabs show recurrent bacterial infection, which responds to courses of antibiotics but rapidly recurs.

The child will require examination under anaesthesia and saline lavage. Often there is very little to be seen on lavage, and it is likely that only small fragments can cause this clinical presentation.

Hair Tourniquet

Although very uncommon, the clitoris may be strangulated by a strand of hair that has become tightly wrapped around it. This causes swelling and acute pain.

As long as the condition is recognised for what it is and the hair cut off quickly, it will resolve without sequelae. There have been cases where delayed diagnosis has resulted in auto-amputation.

Labia Minora in Children and Adolescents: Size and Labial Asymmetry, What Is Normal?

Pre-pubertal children normally have very small labia minora, however, around puberty, they enlarge to the normal structures found in adults.

There is a very large variation in the size and bulk of labia minora. Rarely, they are congenitally absent but the vast majority of women have them and their absence during reproductive life is not normal.

In some girls, the labia grow asymmetrically, with one side growing first and the other catching up later. It is not uncommon for some minor degree of asymmetry to be present, but in some individuals, this is so marked that it becomes a source of embarrassment – particularly in adolescence.

Very large labia minora can also occur bilaterally and can become not only embarrassing but uncomfortable. As a result, patients may request surgical correction. This is called 'reduction labiaplasty'.

Labiaplasty is becoming more and more widely requested. It is very important to understand the motivation of the patient: there is a difference between trimming large labia that are producing genuine discomfort, and reducing normal labia for purely cosmetic reasons. Moreover, the complaint of large, uncomfortable labia may in fact be a presentation of a skin disease such as psoriasis. Rarely, Crohn's disease may present in children with vulval swelling.

It is not our place to pass judgement on those who perform this procedure, except to point out that it is not without risk, and patients should be adequately assessed, and fully informed, before proceeding. The stance of major societies such as the International Society for the Study of Vulvar Disease is that children requesting cosmetic genital procedures should be at least 18 years of age.

Psychological Aspects of Vulval Problems in Children

The Child with Symptoms but a Normal-Appearing Vulva

In adults with vulval pain (and less commonly itch), there is a small but significant group who present with symptoms but with no apparent abnormality. Although some are malingering or somaticising, there are many more who have a genuine complaint of neuropathy or referred pain.

When a child presents with symptoms but nothing to see, even after close examination when symptoms are maximal, it is unlikely that there is any physical cause. It is always worth checking for a scoliosis and obtaining a physiotherapy assessment if the child's complaints raise a suspicion of neuropathy. However, neuropathic pain in pre-pubertal girls is rare.

A common scenario is the asymptomatic child who presents because of a greenish discharge noticed as staining of the underwear. Swabs and urine culture are normal. This situation is a normal variant.

A somewhat less innocent situation is a child who constantly complains of vulval discomfort in the absence of findings and without any observable sign of being in pain. Children rapidly realise that complaining of genital pain, particularly at school or in public, attracts adult attention, and is a source of embarrassment for their parents.

They may even find that worried teachers rapidly send them home from school, and distraught parents are then summoned to answer sexual abuse allegations. In other words, havoc can be created, not only for the parents, but also for the unwary clinician!

Children who do this have no idea how much distress they are causing, but know that it is an effective attention-seeking device. The best way to deal with it is usually to withdraw the attention, but occasionally psychiatric help is needed.

Sometimes a child's complaint of vulval pain may be a smokescreen for her habit of masturbation. This can happen if her parents are shocked by this behaviour. It is then necessary to help the parents come to terms with the normality of the child's actions.

In most of these cases, non-intervention, reassurance and not giving in to attention-getting behaviour is the best treatment.

Vulval Disease and Sexual Abuse: What Should You Do?

Most parents of a child with a vulval condition of any sort will have considered the possibility of sexual abuse, even though they often do not tend to voice it, particularly at the first visit. It is reasonable for them to do so.

All forms of sexual abuse and paedophilia have rightly received enormous publicity in the lay press, however, there are never any details of what physical evidence there might be in an abused child, and this is therefore left up to the imagination.

Professionals who deal with children are also made very aware of child abuse as an issue because of legal requirements to reveal criminal records as a condition of employment. It is therefore common for carers and teachers to have these concerns about children who scratch the vulval area constantly or complain of vulval pain. Parents who suspect abuse in a child with a vulval condition may blame persons who care for the child in their absence.

Doctors in many countries are required by law to report any suspected case of sexual abuse, and when a general practitioner is faced with a vulval condition, they too may consider sexual abuse. However, this presents them with a very difficult problem. Reporting the patient may well ruin the doctor–patient relationship, and may result in an unnecessary, distressing invasion of privacy if they are proven to be incorrect. The fact remains that in Australia doctors are mandatory reporters.

The vulval conditions that should prompt doctors to consider sexual abuse are not the dermatoses such as psoriasis and LS but the infectious diseases that may be sexually transmissible.

The situation, however, is far from straightforwards. When it comes to genital warts, there are enough articles in the medical literature that state that these lesions in children are rarely sexually transmissible to be able to make this an area of ongoing controversy. It is also genuinely possible that genital herpes in a child can be the result of non-sexual transmission, particularly if there is an underlying condition such as eczema.

Even in expert hands, diagnosing sexual abuse is very difficult and impossible to prove without a disclosure from the child or a relative. Many cases remain unresolved even after investigation and interview in the child protection unit setting.

Numerous retrospective studies confirm that about 20% of adult women were in some way sexually abused as children. This does not necessarily mean sexual intercourse, however, all forms of unwelcome and inappropriate sexual contact in childhood can have devastating long-term psychological implications.

Perpetrators of sexual abuse are usually well known to the child, often relatives and close family friends who blackmail them into remaining silent. Child sexual abuse within the family is society's well-kept secret. It is almost never spoken of and the likelihood of obtaining a disclosure from any family member or the child is remote. Without disclosure, it is very difficult for any further action to be taken. Although institutionalised sexual abuse is now receiving much attention, studies show that child sexual abuse within the family unit and close acquaintances is common. This is very difficult to address. One can only hope that the experience of being questioned by a health professional will encourage the family to act of their own accord.

Most children who have been sexually abused do not have any physical signs, because trauma such as bruises resolve quickly, and abusive behaviour often does not involve attempts at penetration.

The presence of a rash such as eczema, psoriasis or LS should not raise queries of abuse. The child may well have been abused but the rash in itself is not evidence and there would have to be other reasons to suspect it.

In cases where the child has an infection that may have been acquired sexually such as genital warts or genital herpes, the issue of sexual abuse should be raised, and if there is no obvious explanation of non-sexual transmission, there are grounds to report.

Where there is a suspicion of sexual abuse, consultation with a child protection unit is the best first step. Child protection units are found in tertiary referral children's hospitals. We recommend that you call a child protection unit and speak to a paediatrician working there. This usually provides clarity about what to do next. If you have referred your patient and the family refuses to be assessed or does not attend their appointment, consult the child protection unit regarding your legal obligations.

When a child presents with a vulval rash, parents often have unvoiced concerns about sexual abuse, so it is worth discussing this. They will usually be greatly relieved that their child simply has a skin problem.

The medical literature contains many cases where skin conditions have been mistaken for sexual abuse, and this includes LS, ulcerated haemangiomas and rare skin conditions such as bullous pemphigoid, which may cause genital ulcers.

It is important to understand that lay people may attribute almost any vulval condition to sexual abuse. Although the presence of a skin condition does not rule it out, there would have to be other grounds to suspect it such as household composition, parental concerns, presence of sexually acquired infections and behavioural abnormalities in the child.

Psychological Management

There is often a much greater emotional overlay attached to any condition of the genital area than in other parts of the skin. As a result, the degree of distress experienced by the parents and sometimes the child may be out of proportion to the actual problem Therefore, much stronger reassurance is often required when skin disease affects the vulva than when it is found on other skin areas. Always enquire about fears of sexually transmissible disease and child abuse.

It is best to be matter of fact and help the parents to understand that the vulva is simply part of the skin. Inform them that children rapidly pick up their anxieties, and that an intelligent child may capitalise on this with attention-seeking or school-avoidance behaviour.

Parents are usually relieved and grateful for a diagnosis that they can believe and that allays any fears of the worst that they may have.

What to Say to an Adolescent with a Vulval Problem

There are two situations that you will face with adolescent girls: a patient who has had a chronic vulval condition, such as LS or psoriasis, since primary school and still has it at adolescence, and the adolescent presenting for the first time with a vulval condition.

The first scenario often raises issues around autonomy and non-compliance, which were not a problem in childhood. The girl who readily allowed her mum to assist with treatment and was unconcerned about a genital examination as a six-year old is often very different as a 12-year old.

In some ways, meeting an adolescent patient for the first time with a new presentation is much easier as the relationship you have with them is new and adult from the start and there does not exist a long history of unwanted interventions to cope with. The main things to pay attention to are:

- understanding of anatomy including correct anatomical names
- using tampons: this is particularly important for girls with vulval skin conditions where liners and pads will be irritating
- how to apply treatment properly: this means not just wiping topicals onto the labia majora but applying to all areas including retraction of the clitoral hood
- washing: similar to how to apply topicals
- sex and consent: although these are not directly related to what you are treating, it is an opportunity to listen to concerns, answer questions and set out a guide to your young patient's rights, particularly around pressure to engage in sexual activity that she does not feel OK about.

Adolescents with any sort of medical condition are likely to feel different, singled-out and isolated. They often have unvoiced fears around sexuality and reproduction and unless you ask what they are worried about, they will not tell you. However, when asked, it may surprise you to find that they are anxious for information and for permission to say no.

Our experience with these young people is that they do not want information to be sugar-coated, they would rather know the facts and the implications of not treating themselves. When this is discussed in a frank and open way, it is surprising how often co-operation follows. In other words, if you treat your adolescent patients as children, this is how they will behave. Treat them as adults, and most will rise to the occasion.

When to Refer

- The diagnosis is uncertain, or if there is apparent non-response to treatment. In practice, non-response is often a result of non-compliance, which is in turn the result of anxiety about the condition or fear of using topical corticosteroids.
- Extra reassurance may be required.
- In the case of proven infection that is recurrent. There may be an underlying abnormality causing the recurrence, such as a foreign body.
- The child with an ulcerating lesion. Most ulcers are not traumatic but pathological, with aphthous ulcers being the most common.
- You suspect the child has LS.
- The child has a labial fusion.
- There is a concern about sexual abuse.

Table 9.1 is a summary table of vulval disease in children.

Table 9.1 Causes of vulval disease in children

A red itchy rash	Eczema Psoriasis Tinea Folliculitis Allergic contact dermatitis Vaginal foreign body with persistent discharge
White rash	LS
Blisters, erosions or ulcers	Impetigo Herpes simplex Herpes zoster Varicella Aphthous ulcers Bullous pemphigoid Erythema multiforme
Acute vulvo-vaginitis	Group A – haemolytic streptococcal vulvo-vaginitis Fixed drug eruption Erythema multiforme
Lesions	Molluscum contagiosum Human papilloma virus Pyramidal perineal protrusion Birthmarks Haemangiomas
Normal-appearing vulva	Subacute vulvitis Pinworm Attention-getting behaviour

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Chapter

10 Myths and Pearls

Contents

Myths

. . . about Thrush

Most Itchy Vulval Conditions Are Due to Thrush

In chronic vulvitis, chronic thrush represents the minority (about 20%) of all cases. Many patients tell us that their doctor prescribed anti-fungal medication without either examining them or taking a vaginal swab.

Multiple courses of anti-fungals should not be given without confirmation of candidiasis on vaginal culture. Particularly for patients with chronic vulvovaginal candidiasis, the length of time that the patient needs to be withdrawn from such agents before vaginal culture is positive again can be many weeks. We therefore strongly recommend that patients always have a vaginal swab before any treatment is commenced.

The complication is that in many cases of genuine candidiasis, swabs are negative, mostly due to anti-fungal use. In this scenario, history taking is key.

Pre-pubertal Children Suffer from Vaginal Thrush

This is completely untrue. Healthy pre-pubertal children who are out of nappies do not suffer from thrush.

Thrush requires an oestrogenised environment and is therefore not seen before puberty. In Australia, the only infective vaginitis we see with any frequency is due to Group A *Streptococcus*.

Post-menopausal Women Can Suffer from Vaginal Thrush

Adequate oestrogen levels are necessary for a woman to acquire a vaginal candidiasis infection and therefore thrush is not seen in post-menopausal women. The only exceptions to this rule are:

- menopausal hormonal therapy, either systemic or vaginal
- diabetes, particularly in the setting of a patient taking SGLT2 inhibitors

- the over-use of vulval topical steroids
- immunosuppression.

Probiotics and the Anti-candida Diet Are the 'Natural' Answer for Thrush

These are very popular concepts, but have no clinical evidence for efficacy. The anti-candida diet is very difficult to comply with and probiotics are expensive. The patients we see have usually tried them without success.

Long-Term Oral Anti-fungal Drugs Will Harm Your Liver

The first oral anti-candidal medication, ketoconazole, had a risk of drug-induced hepatitis of about 10%. The newer oral anti-fungal medications recommended in this book, itraconazole and fluconazole, have a very minimal risk of liver damage. Our experience with the long-term use of these drugs is that they are of similar risk to the long-term use of anti-viral medication.

The risks of modern anti-fungals relate more their potential interactions with lipid-lowering drugs, some psycho-active drugs and some cardiac medications.

Chronic Thrush Can Be Managed by a Weekly Dose of Fluconazole

There have been publications that recommend this approach. It does work for some patients. Nevertheless, we see many failures from this regimen.

We believe that the best way to bring chronic thrush under control initially is with daily oral anti-fungal treatment. Intermittent regimens are appropriate once control has been achieved, but not before.

. . . about Oestrogen

If the Anti-fungal Has Not Worked, Try a Topical Oestrogen

There is only one condition in adults for whom topical oestrogen will have any benefit: oestrogen deficiency. This pertains only to post-menopausal, lactating and very under-weight women. Topical oestrogen will not assist women with adequate oestrogenisation.

In pre-menarchal children, the only condition that should be treated with topical oestrogen is fusion of the labia. It is completely inappropriate to use topical oestrogen in a child in any other situation.

Hormonal Therapy Does Not Cause Thrush or Allergies

This is also completely untrue. We see post-menopausal women who have intractable thrush until they cease their systemic or vaginal hormone-replacement therapy (HRT). In a post-menopausal woman on HRT who has thrush, it should be assumed that this is responsible.

Vaginal creams and pessaries not uncommonly cause an irritant vulvitis or vaginitis, which is reversible when the treatment is ceased.

Many allergic skin reactions to systemic HRT have been described, and we sometimes see women with similar vaginal allergic reactions to it.

. . . about Lichen Sclerosus

Lichen Sclerosus Can Be Managed Long-Term with a Twice-Weekly 'One Size Fits All' Topical Steroid

The opinion pieces that state this have been based on a small number of older, short-term studies. These have now been superseded by our prospective study of lichen sclerosus (LS), which confirms that there is no 'one size fits all' treatment for LS. We recommend that treatment be tailored to the individual patient for best results and high levels of long-term control.

Lichen Sclerosus Does Not Require Follow-Up

This myth has arisen because active LS may be asymptomatic for long periods of time. Most patients, if withdrawn from therapy, will eventually have a recurrence of their symptoms, and by the time that this happens, there will frequently be more irreversible damage.

Childhood Onset Lichen Sclerosus Resolves at Puberty and Therefore Does Not Need Treatment

This is not only untrue in most cases but risky. At least 50% of children with LS will experience scarring if it is not treated aggressively. True remission of LS at puberty is probably very unusual. Careful follow-up of LS is important in order to:

- monitor for cancer
- adjust treatment
- check for side effects of treatment
- make sure scarring is not interfering with the patient's life
- encourage ongoing compliance
- empower your patient to resist the many influences that tell her that long-term use of topical corticosteroids is dangerous.

If a patient's topical steroid requirements decrease so that they are using only 1% hydrocortisone, a trial of cessation of therapy is reasonable. Try to impress on your patient that, even if they may remain asymptomatic, the disease may re-activate.

If possible, patients should not be entirely discharged from care until they have been on no treatment and objectively disease-free for a year.

. . . about Topical Corticosteroids

Topical corticosteroid phobia is at epidemic proportions. Unfortunately, much of this fear derives from the incorrect attribution of systemic corticosteroid side effects to the topical agents.

The terms 'use sparingly' and 'not for internal use' on pharmacy labels frighten patients into reducing your treatment programmes. The Internet has also sensationalised the dangers of these medications and the popular push for all things 'natural' has demonised them as truly hazardous.

The fact is that there are sixty years of extensive research that demonstrates how safe and effective topical steroids are if used carefully.

The notion that skin will be somehow 'thinned' by topical corticosteroids is the most pervading fear. (Most patients when asked what they think this means are not really sure.) The super-potent corticosteroid clobetasol propionate may do this with prolonged use. This

medication has, unfortunately, become synonymous with treatment of vulval disease as it was the first topical corticosteroid reported to be effective for LS. As a result, most subsequent trials employed it, in our opinion, unnecessarily. Although it is useful for severe cases of LS and for lichen planus, it is rarely required to treat other vulval conditions, and weaker topical corticosteroids do not share its hazards.

On the vulva, 'thin skin' would mean visible veins, striae and fragility so that skin would tear during intercourse. In fact, we do not see this very often in our practices.

In general, in the genital area, the use of a weak topical corticosteroid such as 1% hydrocortisone is very safe, even long term. Stronger steroids may be used when needed, particularly in LS.

In reality, the main side effect that we see from topical corticosteroid is redness associated with a burning sensation. This is reversible with a reduction in potency. In some LS patients with high steroid requirements, candidiasis can supervene, however, this is surprisingly uncommon.

. . . That Psoriasis Can Be Cured

Psoriasis is a chronic endogenous dermatosis, for which there is most certainly no cure. It may unpredictably go into remission for long periods of time. However, there is always the risk that it will reactivate, particularly at times of emotional stress.

Always be very honest about the prognosis in patients with psoriasis: it is incurable, but can be controlled, although this can be more difficult for some patients than for others.

Many cases of psoriasis are mistakenly thought to be an allergic or irritant dermatitis, which potentially *can* be cured. If such a patient represents when you thought that they should have been cured, you may be dealing with psoriasis.

. . . That Vulval Disease Is Primarily Gynaecology

Vulval problems do not come under any one specialty. Gynaecology plays a role, but the dominant specialty involved is dermatology. However, pain management, neurology, gynaecological oncology, sexual health, physiotherapy and psychotherapy may all be important in certain patients.

Being a 'vulvologist' involves some knowledge of all of these areas, although no one person can have complete expertise. This is what makes vulval disease a truly multidisciplinary area.

The vulva is part of the skin with its own unique features. The outside of the vulva has much in common with the axilla because of the presence of hair and apocrine sweat glands, the lack of sun-exposure and because of its tendency to maceration.

The inner vulva has a great deal in common with the inside of the mouth. The lining of the vagina is keratinising mucosa with mucous glands and sebaceous glands.

Of the diseases that involve the vulva, the majority of those that have objective signs are skin diseases, and therefore come under the umbrella of dermatology. Diseases that involve the vagina such as lichen planus and autoimmune blistering diseases are also dermatological conditions, as are allergic reactions to suppositories, condoms and semen.

Gynaecological conditions that cause vulval disease are vaginitis due to infections, such as chronic thrush, and non-infective causes, such as desquamative inflammatory vulvovaginitis.

Gynaecology is of great importance in the field of vulval cancer, vulval conditions requiring surgery and where hormonal influences play a role in vulval disease. This includes

oestrogen-hypersensitivity vulvitis, atrophic vulvovaginitis and post-menopausal women with vulval conditions where there are issues related to HRT.

. . . That When All Else Has Failed, Take a Biopsy

Biopsies should be undertaken only if there is an objective abnormality.

There are many reasons for treatment failure. These include:

- non-compliance, often due to corticosteroid phobia
- the patient's own well-meaning hygiene habits
- sporting activities that irritate the skin
- undiagnosed infection
- allergy to a medication, condom, semen or over-the-counter preparations
- incorrect diagnosis, especially of musculoskeletal and neuropathic pain
- psychological problems.

None of these problems will be elucidated by a biopsy. If the vulva looks normal, the biopsy will be normal as well. Take another history and keep the reasons for possible treatment failure in mind.

. . . That Vulvodynia Is a Disease

Vulvodynia is a symptom, not a disease. The word literally means vulval pain, which the International Society for the Study of Vulvar Disease defines as rawness, irritation and burning *for which a demonstrable cause has not been found.*

Therefore, the term 'vulvodynia' should be used only when the clinician cannot determine a cause. We think the number of cases that satisfies this definition is very small indeed.

. . . That Women Who Complain of Chronic Vulval Symptoms Often Have a Psychiatric Disorder

When we were trainee specialists in the 1980s, this was a very common belief. Our subsequent experience, however, has been that vulval symptoms are no more due primarily to psychiatric disease than any other set of symptoms. In the last few years, as the field of vulval disease is becoming recognised and good-quality research is appearing, we have seen this attitude become much less prevalent.

There is no doubt that women with chronic vulval symptoms have high rates of anxiety, depression and sexual disorders but our opinion is that these are usually secondary to the physical vulval disease.

Pelvic floor muscle spasm is occasionally caused by psychological issues, but even in this situation, cure relies heavily on physiotherapy rather than psychotherapy. Our research demonstrates that most cases of dyspareunia are due to identifiable physical disorders. It is therefore of concern to us that non-medical sex therapists see primary presentations of female dyspareunia that have not been examined prior by a doctor.

Many women do feel a sense of relief that their vulval disease has legitimised their pre-existing reduction in sexual interest and some of them refuse to have sexual intercourse with their partners again, even when they have objectively recovered. This may be manipulative or even dishonest, but is a life choice, not madness.

. . . That There Is Such a Thing as an Ideal Vulva

The vulva is a bit like the face: everyone's is different. This is true of size, shape, colour and amount and texture of genital hair. When a patient, particularly an adolescent, comes to see you worried about the appearance of their labia, you have to be able to tell her confidently that everyone is different and there is no such thing as 'normal'. We, like many other health professionals, are not in favour of the rise of cosmetic labiaplasty. Any patient has the right to have cosmetic surgery, despite the expense and risks, but we do not believe that having smaller labia minora will enhance a woman's sexual pleasure. We are also disturbed by the way that these women have been convinced that they are 'abnormal'. An excellent website for your patients to have a look at is www.labialibrary.org.au.

Pearls

Environmental Modification Is Key, No Matter What Your Patient Suffers From

Every vulval condition will benefit from environmental modification. The minimum each patient should be doing includes:

- avoiding all contact with soap
- wearing cotton underwear
- using tampons, menstrual underwear or menstrual cups, rather than pads
- never wearing liners
- never wearing G-stings
- using a clipper or having laser treatment for hair removal instead of shaving or waxing
- wearing underwear and pants that are loose enough to be comfortable
- not wearing Lycra at the gym
- avoiding sporting activities that cause heavy sweating and over-heating
- using a non-irritating lubricant such as vegetable oil
- avoiding perfumed feminine hygiene products and wet wipes
- abstaining from sex if it hurts.

Realise that Anything Genital Is Highly Emotionally Charged

A patient with a genital rash will always attach a great deal more anxiety and significance to it than if the same skin disease occurred on another part of the skin. Patients may also have anxieties about sex that cause anxiety about any genital rash.

Always Ask about Sexual Functioning Even If Your Patient Does Not Mention It

Many patients are too embarrassed to talk about pain with intercourse. Even when this is their main complaint, they will often open their conversation with you in a very round-about way and it is not until you specifically ask if this is their main problem that they admit to it.

There is often a high degree of shame associated with not being able to function 'normally'. The popular press virtually never depicts sex as unpleasant and patients' expectations reflect this.

Ask about Faecal and Urinary Incontinence

Faecal staining of underwear is not uncommon in older women, but such women will rarely volunteer this important information because of embarrassment and shame. It is often caused by genuine faecal incontinence, but can be caused by inadequate cleaning due to obesity, arthritis or poor eyesight.

We see many older women whose vulval problems, which were previously thought intractable, become much easier to manage when their rectal and bladder problems improved. Even though faecal and urinary incontinence may not be curable in many cases, there are simple measures that will help: the application of petroleum jelly over the anus before each bowel motion, and using a hair dryer on the cool setting to properly dry genital skin.

Reassure Your Patient It Is Not Cancer or a Sexually Transmissible Infection

The two things that patients fear and are often too embarrassed to mention are sexually transmissible infections and cancer. Always ask if they are concerned about either or both of these issues.

Keep in mind also that repeated studies tell us that 20% of women were in some way sexually abused as children. They may have suppressed this or have kept it a secret all their lives. Any genital problem will make them wonder if there is a connection between what is going on now and what may have happened to them in the past. However, most genital diseases fall well outside this narrow range of frightening possibilities and are benign skin diseases.

Emphasising that their disease is not a sexually transmissible infection or a cancer is very reassuring. Keep in mind, however, that your patient may be so anxious that very little of what you say has been understood in the way that you hoped it would be. Information sheets that can be read later are very helpful.

Reassure Your Patient that She Cannot 'Give This' to Her Partner

Again, even when you think you have fully explained a benign skin disorder to your patient, you cannot assume that they will understand that it is not transmissible.

The question 'Can I give this to my partner?' after you have just spent ten minutes explaining psoriasis may seem ridiculous to you, but many lay people have trouble grasping the difference between something infective and something endogenous. Most patients assume that skin disorders are mostly infective and the vulva is no exception.

With a Child, Always Ask the Parents Whether They Are Worried about Sexual Abuse Even When They Have Not Mentioned It

When a child presents with a genital problem, the issue of sexual abuse is often on parents' minds, even if they do not immediately voice it. Again, ask specifically.

Although skin disease does not rule out sexual abuse, neither is it a reason alone to suspect it. This is a very different situation, however, to possible sexually transmissible infections such as genital warts or genital herpes. Here it is essential to consider the possibility of child abuse.

The medical literature on sexual abuse tells us that perpetrators are usually known and trusted by the child, so they are frequently family members. This is unbearably painful for the parents of the child and it is only natural that if a child goes to any form of childcare, parents will be directing their suspicions there, rather than at the immediate family. This sometimes has disastrous consequences for innocent child care workers.

With an Adolescent

If you treat an adolescent like an adult, chances are she will behave like one.

- Do not sugar-coat the facts: they want to hear it like it is.
- Ask if they want their parent to stay. They usually do.
- Make the young woman and parent aware that as of the age of 16, children have a right to make their own medical decisions. However, in the real world, they rarely do.
- Use correct anatomical language.
- Talk about tampons and sex.
- Find out what they might be scared of and defuse it.
- A lot of adolescents think their vagina is disgusting. The word 'gross' is often used. Tell them that the mouth has a lot more germs in it.
- Tell them that being pressured to do ANYTHING sexually that they do not like is not OK. This includes oral and anal sex.
- Tell them it is OK to wait for someone who they like very much to have their first sexual experience with.

Make Sure She Has Something to Read that Is Factual and Accurate

Most patients will go straight to their computer when they get home and look up their condition and its treatment on the Internet. Some do this more than others but there are very few who will not do some research of their own.

When it comes to vulval disease, the Internet is full of frightening, complex and sometimes inaccurate data. It is helpful to have information sheets and to direct patients to websites that are factual. The following websites contain good, factual information:

Care Down There: www.caredownthere.com.au

British Society for the Study of Vulval Disease: www.bssvd.org

International Society for the Study of Vulvar Disease: www.issvd.org

How to Do a Vulval Biopsy

We perform this minor procedure in the rooms and it is usually kinder to do it at the first consultation if possible. Sending patients home with an appointment to come back for the biopsy at a later date just gives them more time to worry about it.

Remember that in order to make a histopathological diagnosis of a skin condition, a piece of tissue about 3 mm in diameter to a depth of at least 3 mm is all that is required.

Check first for any underlying varices. To anaesthetise the skin, we inject about 0.1 ml of 1–2% Xylocaine with adrenalin, directly under the area to be biopsied, intra-dermally. If your patient is very apprehensive, topical anaesthetic cream may be applied before injecting. This should be left on for at least 20 minutes to be effective.

We do not use a prep on mucosa, as they are all likely to sting and irritate. On hair-bearing skin, any prep such as povidone-iodine or chlorhexidine may be used.

Before performing the biopsy, test with a sharp instrument to make sure that the area is numb.

There are two very easy and quick procedures:

1. Snip: suitable for lesions on the soft mucosal surface. Simply pinch up a small fold of skin with fine Adson forceps and then cut a small ellipse under the forceps.
2. Punch biopsy: suitable for lesions on hair-bearing skin or for a firm lesion on the mucosal surface, which is difficult to grasp with forceps. Use a round 3 mm punch, 'twiddle' while applying pressure to the lesion to a depth of at least 3 mm, and then withdraw the punch. The core of skin is then lifted up with forceps and snipped off with fine scissors.

It is not necessary to suture biopsies of 3 mm or smaller. Haemostasis is achieved with a haemostatic agent such as silver nitrate. We give the patient a panty liner to wear home in case of minor bleeding. No specific after care is required but the patient should not have intercourse for a week or until the wound has healed.

There are some situations where it is best to formally excise a lesion in its entirety. In this situation, traditional surgical techniques with scalpel excision and suture are used. It is usually not appropriate to attempt such a procedure in the office unless you are experienced in skin surgery.

Show Your Patient How and Where to Apply Her Ointment

Show her with a mirror exactly where the problem is so she knows where to apply her treatment. Most patients do not realise just how far 'inside' the vulval skin actually extends. Treatment failures will occur unless you show them how to do it.

What to Do if a Medication Stings

There are several reasons that medications may cause stinging:

- it is inherent to the medication itself
- it is caused by a preservative or a product found in the base
- the patient's skin is very inflamed and fissured
- the patient has hyperalgesia associated with a neuropathy
- the patient has used too much potent topical corticosteroid; her skin is red and has become very sensitive.

First, examine your patient for the presence of an underlying condition. If there is a severely inflamed dermatosis, and no topical therapy is tolerated, it may be necessary to use a systemic agent long enough to control this before changing back to a topical agent.

In many cases it will take a few days for the stinging to stop. If you suspect excess topical corticosteroid, reduce to a weaker preparation. If the examination is normal and you suspect hyperalgesia, stop all topical therapy.

If the patient is using a cream, cease this and change to an ointment. Certain topical therapies tend to sting even on intact skin, tacrolimus and pimecrolimus in particular. Some patients simply need an alternative product.

Tips to Optimise Compliance

Poor compliance tends to be related to:

- fear of topical corticosteroids
- cost of treatment
- the patient feels well and does not see the point of maintenance treatment
- the patient considers (perhaps falsely!) her condition to be low risk
- regimens that have to be applied multiple times per day
- treatment regimes that recommend intermittent treatment. This often gets forgotten
- treatment side effects
- psychological barriers such as depression, denial or forgetfulness
- embarrassment about having vaginal creams and pessaries where family members might find them
- fear of certain neuromodulators such as the tricyclic anti-depressants, which still carry the stigma of being used for major depression.

To optimise compliance:

- make sure that the patient understands the risks of non-compliance. In LS, this risk includes cancer. As a result, compliance is often excellent
- ask your patient what she is worried about and what gets in the way and try to find solutions together
- make sure that the patient has the knowledge to know what is being treated and why
- explain the concept of chronic disease and the difference between cure and control
- give very strong reassurance about topical corticosteroids
- explain that tricyclics are not being used as anti-depressants
- keep regimens simple, if possible, once daily
- minimise cost of treatment to your patient in any way you can, by recommending economical products and by making them aware that they can shop around for the lowest price
- make a follow-up visit and remind your patient to attend.

When to Refer to a Pelvic Floor Physiotherapist

The lower pelvic floor muscles lie directly under the skin of the vulva and vaginal mucosa. They frequently become hypertonic in response to vulvo-vaginal skin inflammation, leading to pain with stretch, and therefore to dyspareunia and pain with tampon insertion.

For most patients, this hypertonicity ('spasm') settles when the underlying cause improves. For others, it persists. Examination in this situation shows fixed spasm of the pubococcygeus muscle, with inability of the patient to either contract or relax the muscle. This persistent pelvic muscular spasm is often exacerbated by co-existing biomechanical problems, such as lumbo-sacral or hip joint dysfunction.

You should consider referral to a physiotherapist if:

- the patient's dyspareunia does not settle once the dermatosis is controlled
- there is evidence on examination of pubococcygeus spasm
- there is evidence of hyperalgesia.

When to Refer a Patient to a Counsellor

Relative to the number of patients we encounter with vulval disease, we refer only a few to counsellors. It is not essential to refer every patient with a vulval problem for psychosexual help.

Many patients with straightforward skin diseases of the vulva recover quickly and resume normal sex lives without any assistance other than appropriate medical treatment.

The reasons that we have referred in the past include:

- relationship problems
- sexual abuse
- depression
- high levels of anger related to previous adverse experiences with the medical profession
- obsessions with sexually transmissible infections
- patients with pelvic floor spasm who do not have the confidence to resume intercourse after treatment
- patients who are using their condition to manipulate others
- the very small group of patients with a genuinely psychosexual cause for their dyspareunia.

When to Refer to a Pain Management Specialist

Patients with neuropathic vulval pain are often easily helped with neuromodulating medications, exercise, weight loss and physiotherapy. However, there are those whose pain is intractable, has been present for many years and who have associated significant psychological problems. There is also a group where significant drug interactions with neuromodulators require a very expert prescriber.

Patients in this group need the expertise of a specialist in pain management. They frequently also need psychiatric help, although this suggestion tends to be less well received.

Should You Ever Tell a Patient You Have Nothing Further to Offer Her?

In the course of managing patients with vulval disease, we have often seen patients who claim that they have been told, sometimes on more than one occasion, that there is nothing that can be done to help them. This possibly relates more to the very limited information that is available to doctors on the subject of vulval disease than to a limit on therapeutic options.

We certainly cannot help everyone. In some cases, we lack the skills. In some, psychological issues get in the way of effective compliance, making it difficult to ever achieve a good result from therapy.

If we feel that a patient's problems are beyond our area of expertise, we should leave them with a diagnosis and a referral to another practitioner whom we feel may be able to

help. It is important to know when it is time for a patient to move on, but not with a sense that 'there is nothing that anyone can do to help you'.

There is something that will help most patients, even if it is just a validation that their problem is real and deserves our serious attention. Nothing in medicine has been more neglected than the field of vulval disease.

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