When a vulval rash causes symptoms, it is often assumed to be acute candidiasis. However, there are many other vulval dermatological conditions. Dermatitis, chronic vulvovaginal candidiasis, lichen sclerosus and psoriasis are other common causes.

Vulval disease

Part 2: patients with a symptomatic rash

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In Part 2 of this three-part article about vulval disease, we describe those conditions that present with a rash and other symptoms. Last issue, Part 1 presented the principles of a dermatogynaecological approach to the evaluation, investigations and management of vulval conditions. Next issue, Part 3 will describe those conditions that present with either symptoms and no rash, or a rash or lesion with no symptoms.

The patient with a rash and noncyclical symptoms

The many dermatological conditions that can lead to a vulval rash with noncyclical symptoms can usually be differentiated from each other by physical examination plus or minus biopsy. They can be subdivided further into those where:

- the rash does not involve the vagina
- the rash does involve the vagina
- the rash includes blister and erosions.

Rash, noncyclical symptoms and vagina uninvolved

Dermatitis

Dermatitis is the most common cause of chronic vulval symptoms, and accounts for about a third of all vulval presentations.

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Types

Atopic dermatitis. About 80% of patients with vulval dermatitis are atopic and their condition is a localised form of atopic dermatitis. History taking will reveal asthma, hay fever or dermatitis of other parts of the skin.

Irritant contact dermatitis. Of the remaining 20% of patients with vulval dermatitis, most will be suffering from irritant contact dermatitis as a result of overexposure to soap, bubble bath, occlusive clothing, or daily use of panty liners or other irritants. Patients with a normal but heavy vaginal discharge or patients who dislike discharge getting onto their clothes may wear panty liners every day. Some women are too fearful of the toxic shock syndrome to wear tampons.

Incontinence is a very important cause of persistent irritant dermatitis, particularly in postmenopausal women. The skin is macerated because of constant exposure to urine, and to pads and panty liners which tend to have a plastic backing. Patients rarely mention their problem because of embarrassment so it needs to be enquired about.

Allergic contact dermatitis. True allergic contact dermatitis to a substance such as perfume in toilet paper, latex in condoms or an over-the-counter or applied medication is an uncommon event but one to be kept in mind in recalcitrant, severe cases of dermatitis, particularly when the rash is eroded, blistering or ulcerated (Figure 1). Rarely, a patient may be allergic to seminal fluid.

Corticosteroid-induced dermatitis. Prolonged use of fluorinated topical steroids on the vulva can produce a dermatitis that flares every time an attempt is made to cease the topical steroid. This is a situation analogous to the periorificial dermatitis seen when similar creams are used on the face.

Seborrhoeic dermatitis. Seborrhoeic dermatitis, often with involvement of other typical sites such as the scalp and axillae, may involve the vulva (Figure 2).

Symptoms and signs

Patients with dermatitis are almost invariably itchy. If the mucosal surface is involved, they will also experience burning. Excoriations and fissures produce pain and dyspareunia (Figure 3).

Examination shows poorly defined erythema and scaliness of the labia, which may extend onto the pubis and the thighs. However, the signs may be very subtle and minimal (Figure 4). In longstanding cases, lichenification is common resulting in rugosity of the labia majora and perianal 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.

The vagina is not involved, and there is no discharge unless superinfection is present, most often due to *Staphylococcus aureus*, *Streptococcus pyogenes* or *Candida albicans*.

Management

The principles of management of all vulval disorders, including environmental modification and management of incontinence, were discussed in Part 1 of this article. Specifically, most dermatological conditions, including dermatitis as well as psoriasis and lichen sclerosus, are corticosteroid responsive. Therapy should begin with a potent topical corticosteroid (we favour

IN SUMMARY

- **Vulval conditions with a symptomatic rash can be divided into those with cyclical symptoms and those with noncyclical symptoms.**
- **Rash with noncyclical symptoms can be further subdivided into rash which does or does not involve the vagina and rash with blistering and erosions. Physical examination, with or without biopsy, will usually differentiate the causes.**
- **Dermatitis is the most common cause of chronic vulval symptoms, and accounts for about a third of all vulval presentations.**
- **Most dermatological conditions, such as dermatitis, psoriasis and lichen sclerosus, are corticosteroid responsive.**
- **Most women with chronic vulvovaginal candidiasis are otherwise normal and their susceptibility to the infection is as yet unexplained. Chronic candidiasis accounts for about 15% of patients with longstanding vulval symptoms.**
- **It is essential to consider options other than ‘thrush’ when a patient presents with vulval symptoms. There are many other dermatological conditions of the vulva, and specific management is dependent on the underlying condition.**

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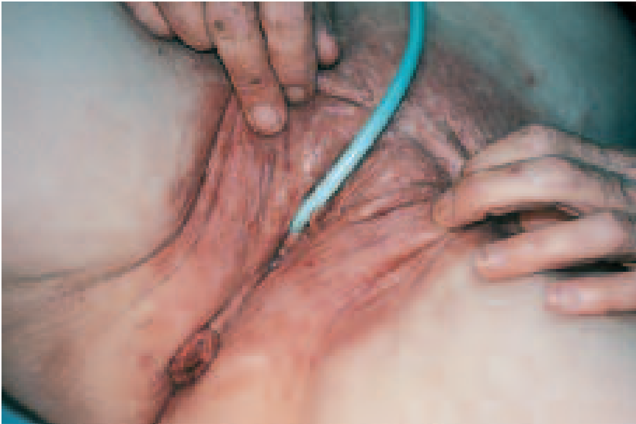

Figure 1. Acute contact dermatitis, showing spreading erythema, swelling and erosions. This is the result of an applied topical antibiotic.

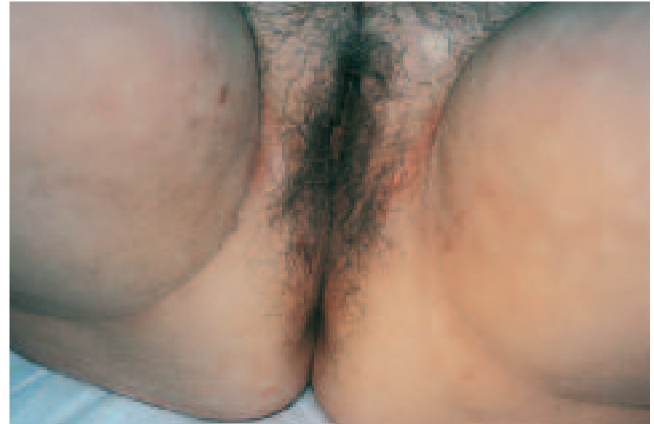

Figure 2. Seborrhoeic dermatitis, showing typical involvement of the inguinal folds.

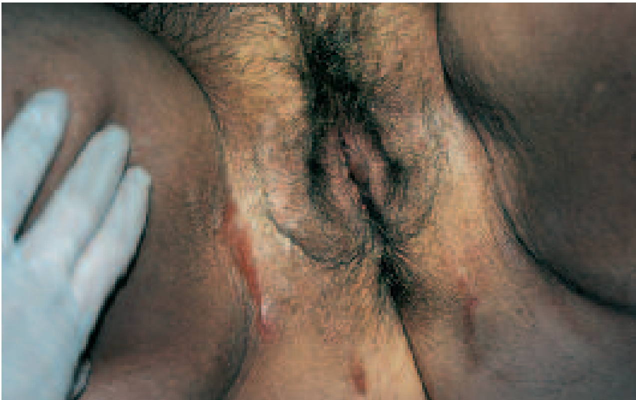

Figure 3. Dermatitis. The skin is erythematous, thickened and fissured.

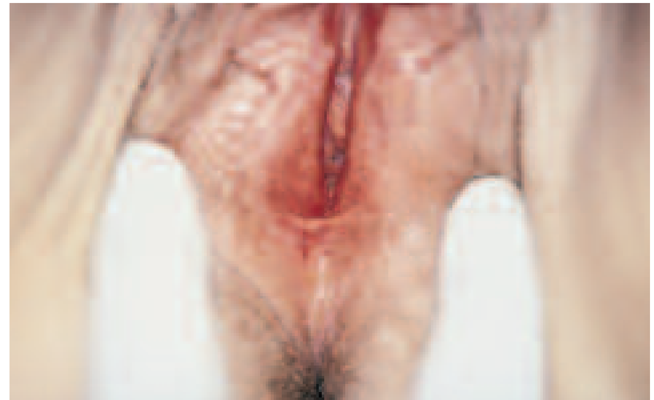

Figure 4. Some subtle signs of dermatitis: a split at 6 o'clock on the introitus associated with erythema and splitting of the mucosal surface.

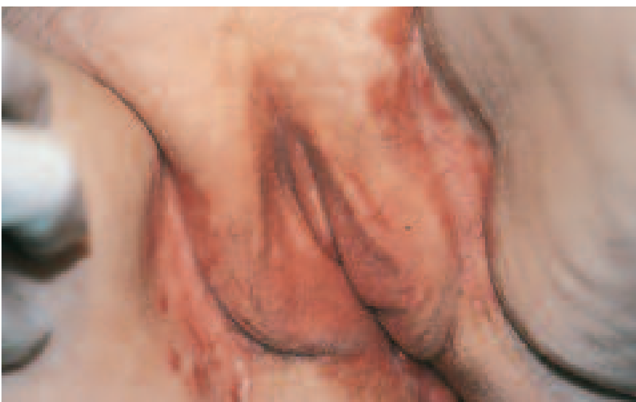

Figure 5. Psoriasis. Well demarcated red plaque without the usual white scale associated with psoriasis elsewhere.

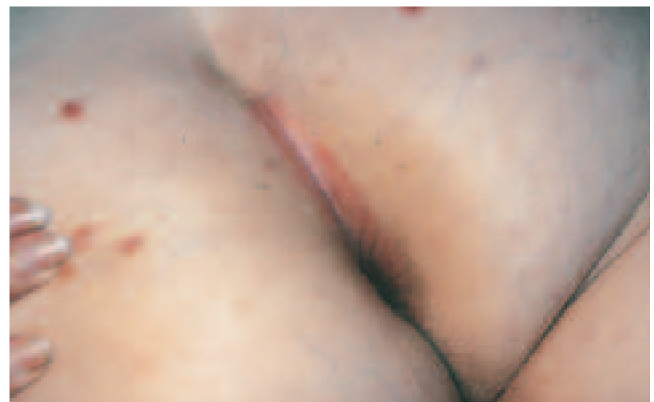

Figure 6. Natal cleft involvement is a clue to psoriasis.

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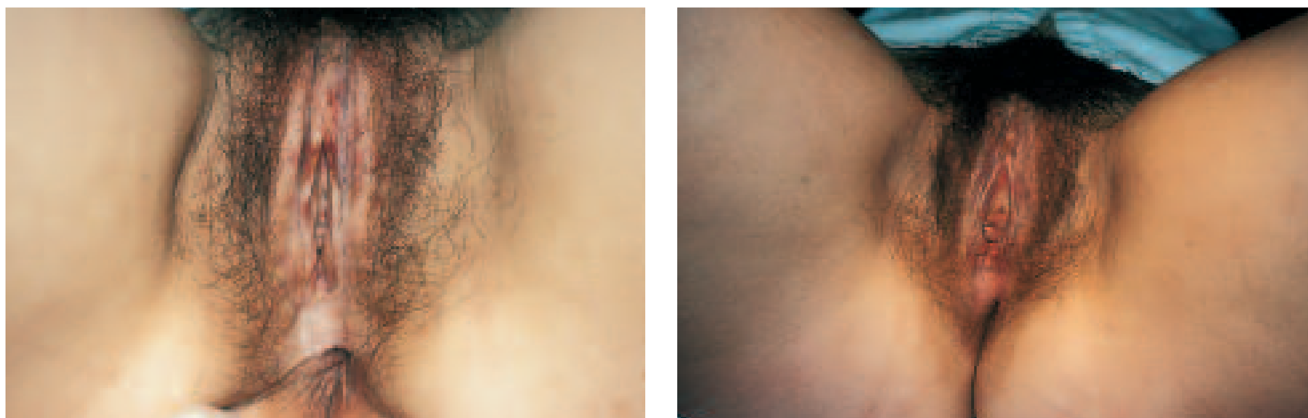

Figures 7a and b. Lichen sclerosus in a young woman. a (left). Before treatment. b (right). After treatment.

methylprednisolone aceponate as it is potent but appears to have a relatively low potential to cause stinging or atrophy) until symptoms have resolved and the skin has normalised. Then, a reduction to a weaker preparation such as 1% hydrocortisone should be attempted and continued for three months. Any recrudescence after this should be managed by a repeat of this complete cycle.

Psoriasis

Psoriasis is also a common vulval condition but less so than dermatitis, with which it is most often confused. Psoriasis accounts for 5 to 10% of patients with vulval rashes.

Symptoms and signs

Psoriasis of the vulva is easy to diagnose if present elsewhere on the skin, but it can be localised only to the vulva. 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 (Figure 5).

The lesions are usually more erythematous and well defined than dermatitis, and are usually bilaterally symmetrical. Psoriasis does not involve the vagina but the labia are usually involved and the rash can extend as far as the vestibule. The perianal area and natal cleft may be involved and natal cleft involvement is a useful sign, as it is not seen in other conditions (Figure 6).

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

Management

Treatment of psoriasis tends to be more difficult than treatment of dermatitis; it often needs to be more aggressive and more prolonged. Initial treatment with a potent topical steroid is indicated but it is rarely possible to maintain patients on a weak topical corticosteroid, and addition of a weak coal tar cream such as 3% liquor picis carbonis (LPC) in aqueous cream is necessary.

It is well known that psoriasis may be precipitated by streptococcal infections. We find that patients with vulval psoriasis often have a positive vaginal swab for streptococci; this requires treatment.

Lichen sclerosus

Lichen sclerosus is an uncommon skin disease that has a predilection for the genital skin and is ten times more common in women than men. This makes it relatively common in vulval practice, responsible for about 10% of cases.

Lichen sclerosus is an important condition to diagnose correctly for two reasons. Firstly, if not treated aggressively, it may significantly deform the vulva. Secondly, it is associated with squamous cell carcinoma of the vulva, with the lifetime risk said to be 2 to 6%. Thus patients will require lifelong surveillance and a biopsy to confirm the diagnosis should be performed wherever possible.

Symptoms and signs

Lichen sclerosus may be asymptomatic, discovered by chance by the patient or by the GP when a Pap smear is performed; however, it is usually intensely itchy, and sometimes painful as a result of excoriation or fissuring.

The distribution of lichen sclerosus is variable. It may involve any part of the vulva and perianal area. Rarely, lesions may be present on other areas of the skin.

The appearance of a well defined white plaque with an atrophic wrinkled surface and areas of purpura and erosion is typical (Figure 7a). Areas of hyperpigmentation and thickening may appear with time, and occasionally blistering is present. In longstanding cases, atrophy of the vulva occurs, with fusion of the labia and disappearance of the labia minora and clitoris typical (Figure 8). Although the vagina is not involved, the vestibule is, and stenosis of the introitus may occur.

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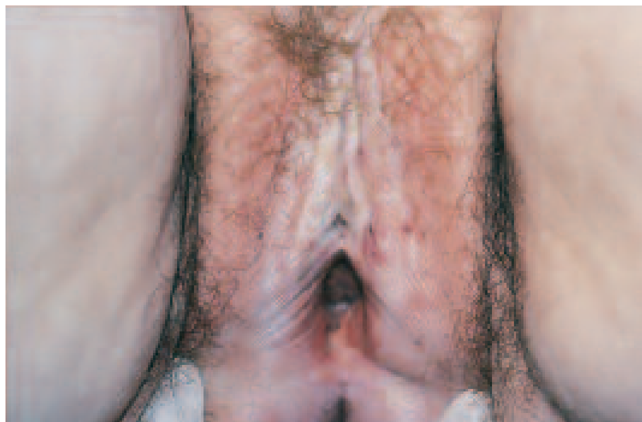

Figure 8. Lichen sclerosus. White plaque with finely wrinkled surface and telangiectasia. The labia minora have resorbed and the clitoris is buried by adhesions.

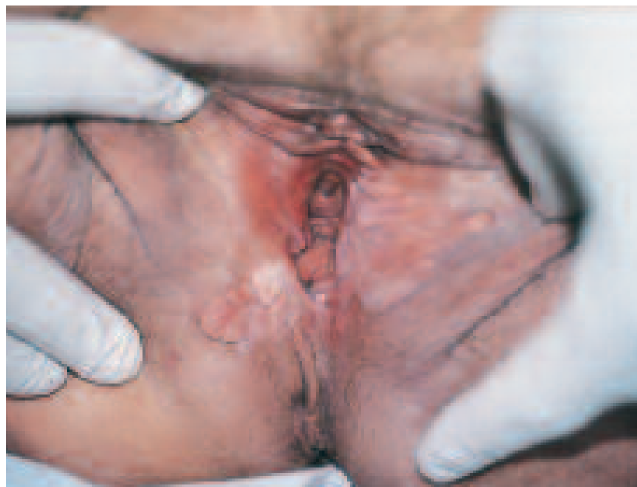

Figure 9. VIN III. Multiple indurated plaques.

Management

Lichen sclerosus should initially be treated with a very potent topical steroid such as Diprosone OV, initially twice daily for a month, then daily for two months, before reducing to weaker preparations.

Response is usually rapid and although the vulva may not completely return to normal appearance, it is usually possible to completely control symptoms and prevent further stenosis and loss of vulval architecture (Figure 7b).

Maintenance therapy with a moderate strength topical steroid is needed as is six-monthly surveillance for squamous cell carcinoma. We usually suggest sharing care with the patient's GP for this lifelong condition but recommend referral if it is suspected.

VIN III (vulval intraepithelial neoplasia)

The histopathology of VIN III is of squamous cell carcinoma *in situ*.

Symptoms and signs

In about half the patients with VIN III, symptoms are not present at diagnosis, but if they are, pruritus is the commonest complaint.

The appearance of VIN III is highly variable. It may present as thickened white or red mucosal patches, hyperpigmented plaques, warty lesions or as persistent erosions or ulcers (Figure 9). The distribution may be single or multifocal, and is usually asymmetrical.

Management

Any lesion suspected of being VIN III should be biopsied immediately. We usually refer these patients to a gynaecological oncologist. Like carcinoma of the cervix, there may have been preceding infection with human papilloma virus and the patient should have a Pap smear.

Tinea

This very common fungal infection is an uncommon cause of vulval disease.

Symptoms and signs

Tinea presents with an itchy, scaly, bilaterally symmetrical rash involving the labia majora and may extend to both inguinal folds and lower abdomen (Figure 10). The edge is usually better demarcated than in dermatitis.

Management

Tinea may be difficult to differentiate clinically from psoriasis or dermatitis and a fungal scraping or biopsy is essential to make this diagnosis.

Treatment usually requires the use of oral antifungal medications, such as griseofulvin (Fulcin, Griseostatin, Grisovin), over a three month period. Tinea is only worsened by topical steroid treatment, even if there is temporary improvement at first.

Extramammary Paget's disease

Extramammary Paget's disease is a very rare condition, but an important one as it is easily mistaken for dermatitis. The typical patient is postmenopausal.

Symptoms and signs

The presentation is with an eczematous, very erythematous bilaterally symmetrical eruption (Figure 11). The eruption is usually itchy. With time, the area becomes raw and weeps. There is no response to topical steroid treatment.

Management

Biopsy is essential for diagnosis. Up to 20% of patients with extramammary Paget's disease of the vulva have an underlying adenocarcinoma. Another 30% have an adenocarcinoma at another location.

Patients with extramammary Paget's disease should be referred to a gynaecological oncologist.

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Rash, noncyclical symptoms and vagina involved

Lichen planus

Lichen planus tends to be overemphasised in most articles on vulval disease; in practice it is quite rare. It is predominantly a mucosal condition which may involve the entire vagina and cervix; however, the labia minora and clitoral area are often involved. Lichen planus should be suspected in any patient with persistent vulvovaginal erosion, particularly if a similar pattern is seen in the mouth.

Symptoms and signs

Patients present with erosions and ulcers which are very painful (Figure 12). There is usually a heavy purulent discharge and superficial bleeding. A white reticulate pattern may be seen on the mucosa adjacent to erosions. It is not uncommon for the oral mucosa to also be involved. Lichen planus may result in severe scarring and stenosis of the vagina.

Management

Lichen planus is diagnosed by vulval biopsy.

The condition is very chronic and difficult to treat, often requiring the use of oral prednisone for long periods in addition to strong topical steroids and topical cyclosporin. Referral is recommended.

Desquamative vaginitis

Desquamative vaginitis is a recently described condition. We suspect that it may be what was described in the past as ‘plasma cell vulvitis’ as it responds to the same treatment.

Symptoms and signs

Patients present with pain and dyspareunia and a rash involving the vagina and introitus but not extending onto skin. The appearance of the rash is of petechiae and shiny erythematous patches that look like but are not erosions (Figure 13). There is usually a heavy, creamy, nonoffensive discharge and accumulations of macerated squames in the interlabial clefts.

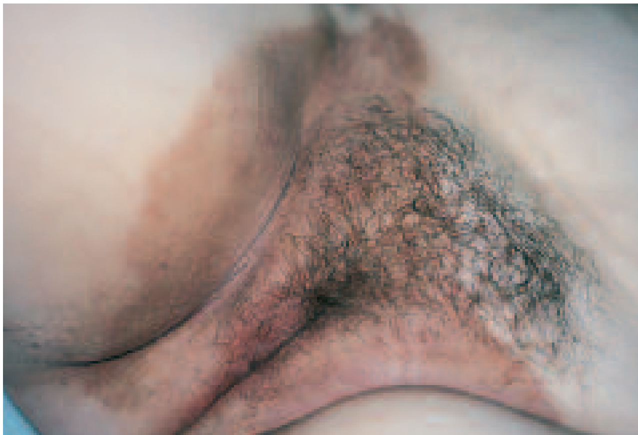

Figure 10. Tinea. The rash has a well demarcated edge and frequently extends onto the thighs and pubis.

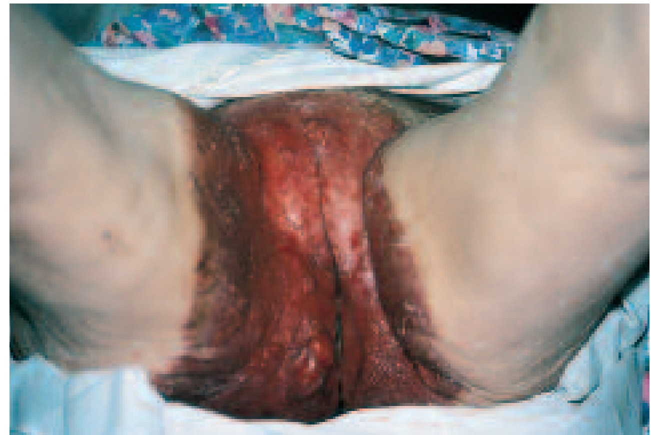

Figure 11. Advanced extramammary Paget’s disease.

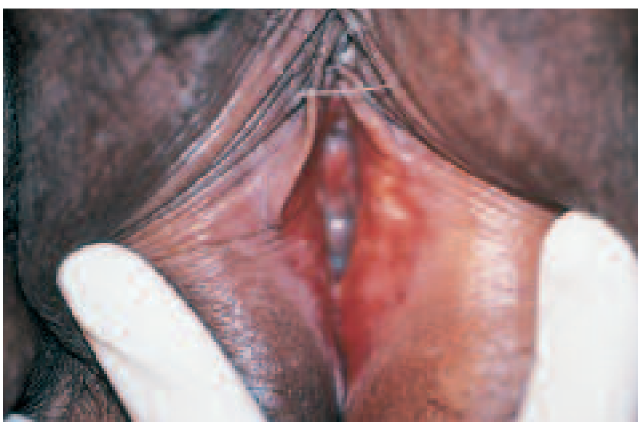

Figure 12. Lichen planus. The mucosal surface is completely eroded. There is a heavy purulent discharge and easy bleeding.

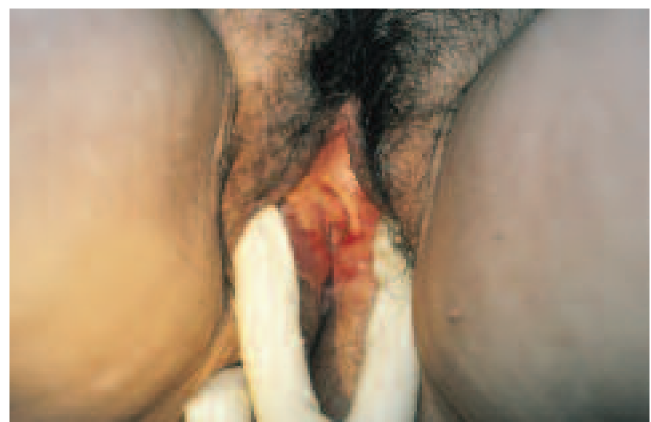

Figure 13. Desquamative vaginitis. The mucosal surface is erythematous, studded with petechiae. There is a heavy discharge.

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Management

Biopsy of desquamative vaginitis will often be reported as nonspecific, but it shows a heavy, mixed inflammatory infiltrate with plasma cells that is much more florid than would be expected in dermatitis.

In this condition, vaginal swabs do not show pathogens, but vaginal smears sent for Gram stain commonly show a predominance of Gram positive cocci rather than the usual lactobacilli of the normal vagina. The condition appears to be an inflammatory response to these organisms as it settles quickly with topical antibiotic and corticosteroid therapy.

We routinely use clindamycin 2% vaginal cream (Dalacin V Cream) and hydrocortisone 1% cream. Treatment has to be continued long term to maintain control.

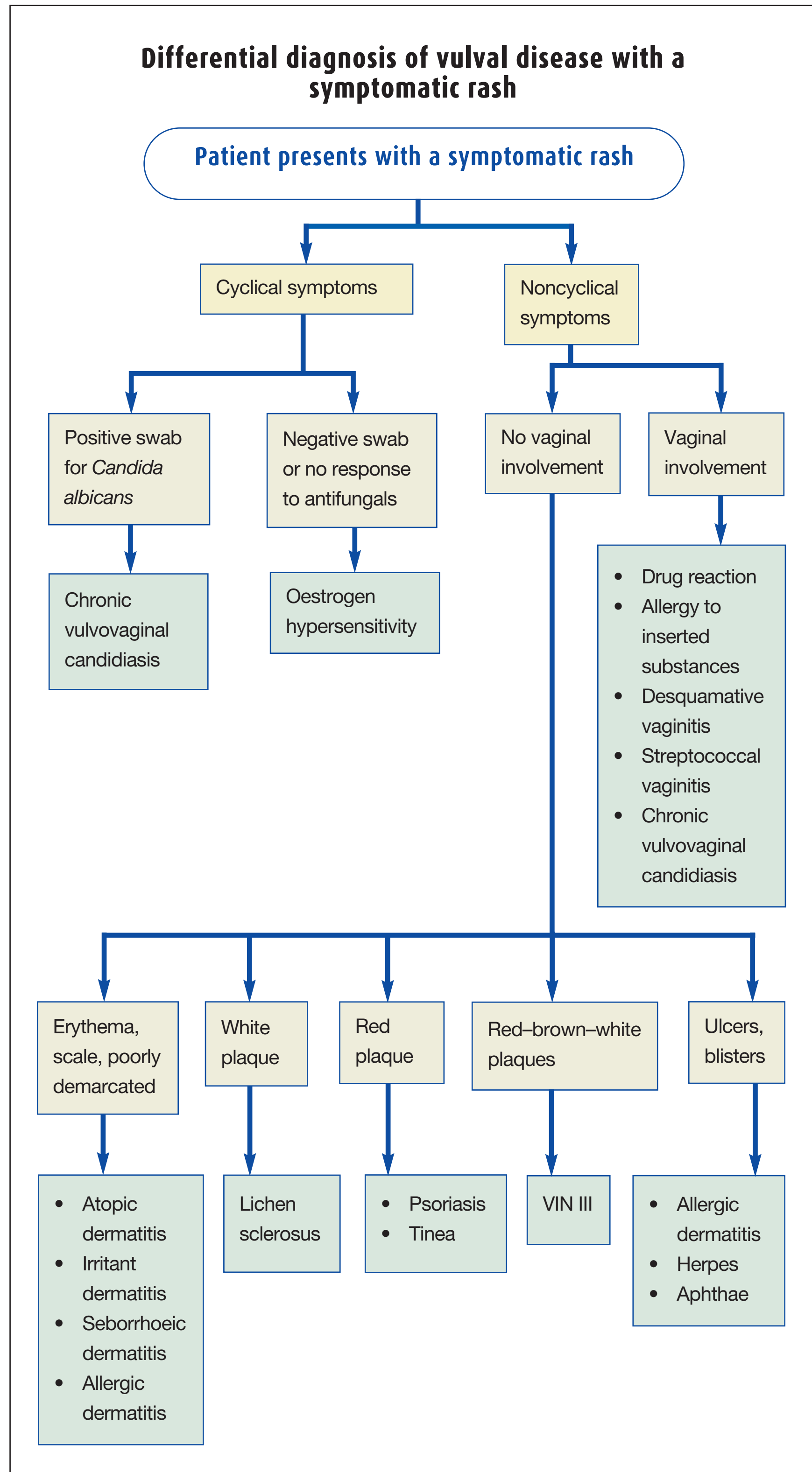
Streptococcal vaginitis with secondary vulvitis

Group B streptococci are often found on low vaginal swab. In many patients this is not clinically significant; however, in some women it causes a vaginitis with a heavy constant discharge. Most women with a heavy discharge wear pads and panty liners on a daily basis and this, together with the maceration of the discharge itself, produces a low grade vulvitis. In such cases, antibiotic treatment to eradicate the streptococcal colonisation is indicated, but usually has to be prolonged.

A finding of Group A beta-haemolytic streptococcus is always significant. In children, this causes a true acute primary vulvovaginitis and perianal dermatitis. In adults, it is usually found as superinfection on dermatitis or lichen sclerosus. A 10-day course of an appropriate antibiotic is indicated.

Hormone replacement therapy induced vulvitis

Although not widely recognised, it is possible for hormone replacement therapy



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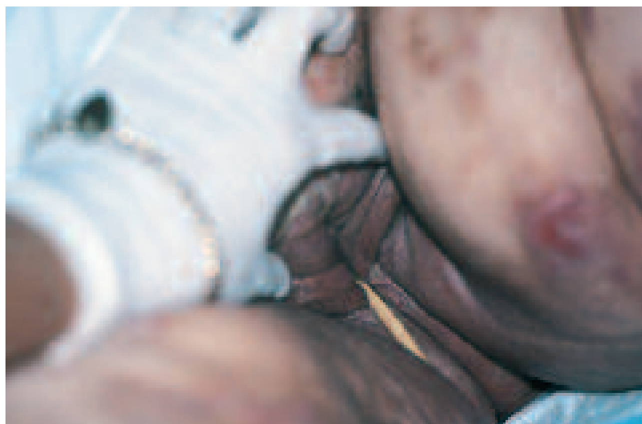

Figure 14. Drug reaction. This patient reacted to methotrexate prescribed for psoriasis.

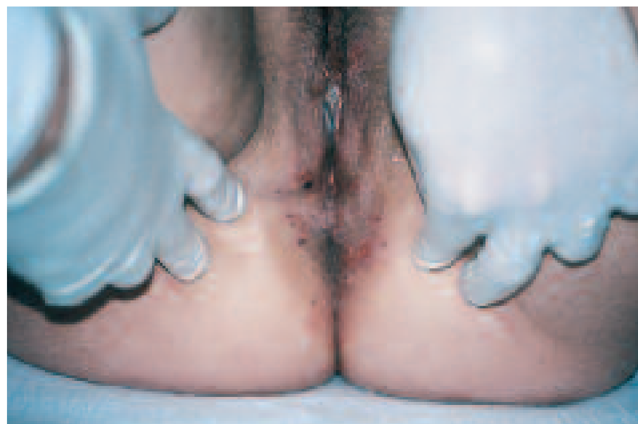

Figure 15. Genital herpes. Primary attack with multiple blisters.

to cause a nonspecific vulvovaginitis due to hypersensitivity to the oestrogen component. It is possible (although a commercial test is not available) to confirm this by intradermal testing with the oestrogen.

Patients will, if questioned, give a history of not having had the problem prior to commencing hormone replacement therapy. Often they will have queried a connection and been told that this is not possible. After ceasing hormone replacement therapy they will experience immediate improvement but full recovery usually takes about three months. Rechallenge will result in onset of symptoms within a month.

More commonly, however, an identical vulvovaginitis can occur when a so-called ‘standard’ dose of hormone replacement therapy is in fact too high for a particular woman. Treatment in this case involves withdrawing hormone replacement therapy (in addition to standard topical management) and later reintroducing hormone replacement therapy at a lower dose.

Postmenopausal women on hormone replacement therapy who have never previously suffered from vulvovaginal candidiasis may develop it for the first time. It will be persistent and treatment resistant if the hormone replacement therapy is not stopped. Once again, after adequate treatment it may be possible to restart hormone replacement therapy at a lower dose.

Other drug reactions

Very occasionally the vulva will be involved in a drug reaction, particularly when this is of the erythema multiforme or fixed drug eruption type. Drugs which can erode mucosal surfaces, particularly chemotherapeutic agents such as methotrexate, may cause a vaginitis (Figure 14).

Antibiotics, especially penicillins or cephalosporins, can cause a vulvovaginal reaction that may be confused with candidiasis.

Rash, noncyclical symptoms and blisters and erosions included

Conditions to consider include three discussed earlier – lichen planus, acute contact dermatitis and drug reactions – and three more – genital herpes, vulval aphthae and bullous disease, such as pemphigus.

Genital herpes

Genital herpes is a common condition but sometimes one that eludes diagnosis. Because of the implications of this condition – infectious, incurable, requires suppressive or recurrent therapy – every effort should be made to diagnose it accurately. The typical grouped painful blisters that erode rapidly are usually few in number and may occur anywhere on the vulva, perianal area or even the buttocks (Figure 15). A swab for immunofluorescence and viral culture should be taken from a new or very recent blister.

Vulval aphthae

These lesions are uncommon and may cause great diagnostic confusion with herpes simplex as they are also episodic, recurrent and painful. However, their appearance is exactly the same as the familiar oral aphthae, which these patients often suffer from. They are not shallow erosions, like herpes, but punched out ulcers with a yellow base and erythematous areola. Sometimes aphthae can be very large and alarming. If in doubt about the diagnosis, referral is recommended.

Bullous disease

Bullous disease, such as pemphigus, is a very rare condition of the vulva. When seen, it is generally accompanied by typical blisters on other parts of the skin. Biopsy is required for diagnosis, and referral is recommended.

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The patient with a rash and cyclical symptoms

Chronic vulvovaginal candidiasis

Approximately 20% of all women carry *Candida albicans* in the vagina, but less than 5% suffer from repeated or intractable clinical candidiasis. Although acute candidiasis is well understood, easily recognised and treated, patients with the chronic form of the infection are a much more difficult group.

A number of factors are associated with an increased tendency to acute and chronic candidiasis – pregnancy, hormone replacement therapy, uncontrolled diabetes, local or systemic corticosteroid treatment, antibiotic therapy and immunosuppression. However, most women with chronic vulvovaginal candidiasis are otherwise normal and their susceptibility to the infection is as yet unexplained. Chronic candidiasis accounts for about 15% of patients with longstanding vulval symptoms.

Symptoms and signs

The most common complaint is of recurrent or constant itch but dyspareunia, burning, swelling and pain are also experienced. It is characteristic but not invariable for these symptoms to cycle, being most severe in the premenstrual week and suddenly improving with the onset of menstruation. Discharge does not usually occur and the typical cheesy exudate of acute candidiasis is absent.

It is typical for courses of oral antibiotics to exacerbate or precipitate symptoms. Patients often give a history of transient response to topical and oral antifungal agents.

The patient's partner may experience postcoital itching and penile rash, the latter characteristic but present in only about 15% of cases. The rash is not due to cross-infection, but to inflammation of the penile skin by irritative fungal discharge.

The patient's rash in chronic vulvovaginal candidiasis may vary from no apparent abnormality to acute erythema involving the whole vulva, perianal area and vagina, with varying degrees of oedema. Satellite lesions are classic but not always seen (Figure 16). Therefore, the clinical appearance is often not helpful is making the diagnosis, which rests on a typical history and positive swabs.

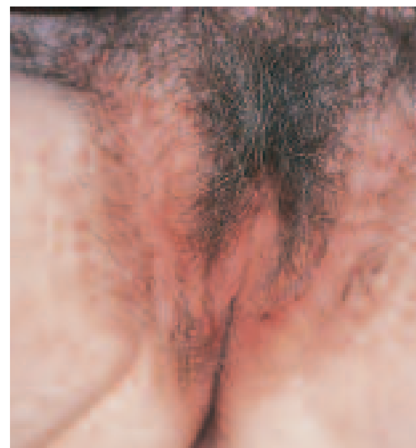

Figure 16. Chronic candidiasis showing classic satellite lesions.

Management

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 swab positivity is significant. False-negative swabs are rare, unless the patient has been on antifungal therapy within the last two weeks. Biopsy is often nonspecific and can be misleading as the fungus is not usually seen. Again, if the history is typical, a trial of antifungal treatment should be given, with the proviso that one must not persist with ineffective treatment in the face of repeatedly negative swabs.

Nearly all cases are caused by *Candida albicans* but other yeasts may be responsible, including *Candida glabrata*. These fungi may be reported as 'nonpathogenic' by some labs but a request for further characterisation should be made.

Chronic vulvovaginal candidiasis is a very chronic condition and in many cases long term suppressive treatment is needed. This can be achieved with either topical or oral therapy; however, most patients prefer to use and comply better with oral therapy – it is less messy and avoids the common experience of irritation from topical therapy.

We commence treatment with either itraconazole (Sporanox) 100 mg per day or fluconazole (Diflucan) 50 mg per day. Ketoconazole (Nizoral) 200 mg per day may be used, but its usefulness is limited by a higher incidence of hepatotoxicity, and regular liver function tests are required.

Therapy is continued on a daily basis until the patient is symptom free – this varies and may take up to six months. We then reduce to weekly dosing for a further six months. Many patients are unable to stop treatment without relapse.

Adjuvant treatment with 1% hydrocortisone is helpful to control itch and many patients report better response if they concurrently use a topical antifungal cream as well. Courses of oral antibiotics need cover with daily dosing again of the oral antifungal agent.

This treatment regimen is effective but expensive. Nevertheless, we have found that most patients are sufficiently impressed with the results to continue treatment. We have used nightly vaginal antifungal preparations for many months consecutively in the occasional patient who cannot afford the more effective oral treatments but who is motivated enough to comply.

Oestrogen hypersensitivity vulvitis

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

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continued

Symptoms and signs

Patients with oestrogen hypersensitivity vulvitis give a history that is very similar to chronic vulvovaginal candidiasis; however, they have had consistently negative swabs and have not responded to antifungal therapy.

They also tend to be refractory to topical steroid treatment.

Management

Oestrogen hypersensitivity vulvitis is not rare and we suspect that it explains many cases that were previously considered undiagnosable. Like hormone replacement therapy hypersensitivity it is possible to confirm the diagnosis by intradermal testing with oestrogen and its metabolites.

The difficulty lies in the treatment of the problem. Some patients with oestrogen hypersensitivity are improved by the oral contraceptive pill, which suppresses their own endogenous hormones. This improvement is not universal, however, and many patients are intolerant of the pill. Cycle suppression with progestogens such as cyproterone acetate (Androcur, Cyprone, Procur) has been our most successful therapy to date.

Long term cyclic suppression is not an attractive proposition for many women particularly if they are trying to become pregnant. However, patients are usually very relieved to find that their problem has an explanation and this alone has been sufficient for some of our patients.

Successful treatment with tamoxifen has been described; however, we have not used it because of its potential side effects.

Conclusion

The majority of patients with vulval symptoms have an observable rash. In most cases, this can be diagnosed clinically, but bacteriology and sometimes biopsy may be needed to determine the exact diagnosis. The importance of making an accurate diagnosis is clear when one considers the diversity of the conditions described above. It is essential to consider options other than ‘thrush’ when a patient presents with vulval symptoms. There are many other dermatological conditions of the vulva, and thrush is responsible for less than 20% of cases. Specific management is also diverse and dependent on the underlying condition. MT

Further reading

1. Fischer G, Ayer B, Frankum B, Spurrett B. Vulvitis attributed to hypersensitivity to estrogen: a report of 11 cases. J Reprod Med 2000. In press.
2. Sobel JD. Desquamative inflammatory vaginitis: a new subgroup of purulent vaginitis responsive to topical 2% clindamycin therapy. Am J Obstet Gynecol 1994; 171: 1215-1220.

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