Skin diseases affecting the vulva

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Abstract

The vulva stretches between the mons pubis and the anus and is bounded by the genitocrural folds. The epithelium includes the keratinised, hair-bearing squamous area of the labia majora and mons pubis and the squamous mucosa of the vaginal introitus. The anatomy produces relative occlusion of the area and contributes to high humidity and levels of surface organisms, whist hormonal variation also influences skin and mucosal function. Skin disorders that can affect any part of the skin can appear slightly different in the vulval area and there are a number of disorders that occur more frequently at anogenital sites than elsewhere on the body.

Keywords blister; infection; inflammation; pruritus vulvae; skin; ulcer; vulva; vulvar diseases; vulvar neoplasms

Skin diseases affecting the vulva can cause significant morbidity and impact negatively on quality of life. Patients are often embarrassed by their symptoms, and may delay seeking a medical opinion.

In addition to a thorough history and examination, further investigations including vulval swabs, skin biopsy or allergy patch testing may be indicated to help reach the diagnosis (Table 1).

Infections

Given the relative warmth and moisture of the vulva, infections in this area are common. It is important to remember that superadded infection can complicate and exacerbate a pre-existing inflammatory dermatosis, making treatment of the underlying condition more difficult.

Yeast & fungal infections

Vulvovaginal candidiasis: is characterised by itch and usually a thick, creamy discharge. A vaginal swab should be taken to confirm the diagnosis before initiating treatment with topical imidazoles (in a pessary or intravaginal cream formulation), or orally. Chronic or incompletely treated candidiasis can present with persistent vulval erythema and fissuring in the interlabial sulci or perineum. Recurrent episodes may respond best to a 7–10 day course of oral fluconazole. Episodes may be precipitated by antibiotic use or associated with the menstrual cycle. Recurrence is common, particularly as Candida is a vulval commensal. Poorly controlled diabetes, pregnancy and oral antibiotics all increase levels of Candida in the vulva, predisposing to further infection. In difficult to treat cases, prophylactic fluconazole (150 mg weekly or monthly) may be a useful adjunct.

Tinea cruris (groin ringworm): often occurs in conjunction with tinea pedis (Athlete's foot). In the groin, buttocks and upper thighs, fungal infection causes well-demarcated erythematous plaques with fine scaling, particularly at the leading edge. In hair bearing areas, there may be deeper fungal involvement with pustules or nodules. Skin scrapings of scaly areas for mycology are useful to identify the causative fungus, generally *Trichophyton* species. If there is intercurrent tinea pedis, oral antifungal treatment with terbinafine or pulsed itraconazole to treat the nail infection is necessary to prevent recurrence of the groin rash.

Bacterial infections

Erythrasma: is caused by *Corynebacteria*, and may be confused with tinea cruris. It presents with a faint, brownish discolouration of the groin, with fine peeling. Examination under a Wood's light (low wavelength ultraviolet light) demonstrates a characteristic coral-pink fluorescence of affected skin. Erythrasma is more common in the immunosuppressed or diabetic population. Treatment is with a topical antibiotic, such as fusidic acid.

Folliculitis: is visible as small papules and pustules associated with hair follicles. A swab taken from pus within a pustule often yields *Staphylococcus aureus*, but other aerobes and anaerobes can also cause this infection. Diabetes, poor hygiene, immunodeficiency and obesity are all aggravating factors. Damage to the skin through shaving presents with a similar picture, but the infection tends to be superficial, without deeper involvement of the hair follicles, and is termed pseudofolliculitis. Folliculitis usually responds to topical antiseptics and systemic antibiotics, although recurrence is common unless the course of treatment is prolonged. General measures to reduce surface bacteria by washing with antiseptic preparations plus increased ventilation to the area (weight loss if the patient is obese, reduced sitting and loose-fitting, natural fibre clothing) will all help to minimise episodes of recurrence. In recurrent/difficult to manage cases, it is worth taking a nasal swab to identify chronic Staphylococcal carriers. Appropriate eradication treatment may help in these cases.

Occasionally infection from an infected follicle can burst into the adjacent subcutaneous tissue forming a larger inflammatory mass with a collection of pus recognisable as a boil or furuncle.

Vulval cellulitis: presents with aching pain, tenderness and a beefy-red swelling of the labia. There may be associated increase in vaginal discharge and general symptoms of malaise and fever. Culture from a vulval swab may confirm the organism (usually *Streptococcus pyogenes*) but treatment should be started immediately to reduce the risk of secondary lymphatic damage and the possibility of lymphoedema with recurrent episodes. Penicillin, flucloxacillin or erythromycin are appropriate choices for oral antibiotics.

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Presentation of vulval disease

Symptom	Possible diagnosis	
	Acute presentation	Chronic continuous/intermittent
Itch	Candida	Lichen simplex
	Contact dermatitis	Lichen sclerosus
		Lichen planus
		Tinea cruris (ringworm)
		Partially controlled contact dermatitis
		Incompletely treated candida
Discomfort/ Pain	Irritant dermatitis	Psoriasis
	Folliculitis	Seborrhoeic dermatitis
	Cellulitis	Intertrigo
	Boils	Erythrasma
		Lichen sclerosus
		Lichen planus
		Plasma cell vulvitis
		Vulvodynia/Vestibulodynia
Swelling of vulva	Cellulitis	Urticaria
		Hidradenitis suppuritiva
		Crohn's disease
		Lymphoedema (primary or secondary)
Lump(s)	Warts	Cysts
	Molluscum contagiosum	Hidradenitis suppuritiva
		Malignancy
Blisters	Herpes simplex	Pemphigus (vulgaris or foliaceous)
Ulcers	Aphthous ulcers	Nicorandil-induced ulceration
	Aphthous ulcers	Behçet's disease

Table 1

Viral infections

Genital herpes simplex virus: (HSV) infection is not uncommon and is generally diagnosed in primary care or genitourinary clinics. Infection is usually with HSV2, but rates of genital HSV1 infection are increasing. Primary genital HSV in a previously seronegative individual causes symptoms within 7 days of infection. There may be a prodromal phase with localised pain prior to the development of papules and vesicles which ulcerate and are extremely painful. There can be associated general malaise. Reactivation episodes are less severe and more commonly present with pain plus a small group of erosions and possibly short-lived blisters amongst them. Oral aciclovir will help to shorten the duration of the acute eruption provided it is taken early in the course of the infection (within 5 days of symptom onset if new lesions are forming). If episodes are frequent, regular prophylactic acyclovir (200 mg twice daily for 6 months) can help to reduce the number of reactivation episodes.

Molluscum contagiosum: produces small pearly papules with a central shallow pit or umbilication. In children, infection is common on the trunk and limbs, but in young adults, genital infection is more common. Clearance is usually spontaneous, but can be hastened with cryotherapy.

Anogenital warts: in adults are generally sexually transmitted, and are usually caused by low-risk human papillomaviruses (HPV) 6 or 11. They present as small papules or cauliflower-shaped growths, and are most commonly found at the posterior fourchette, but can occur at any site within the vulva or vagina.

Treatment of vulval warts depends on the number, size and location. Topical antiproliferative treatments (podophyllotoxin, trichloroacetic acid) or destructive techniques (cryotherapy) are most commonly used. Immunotherapy with topical imiquimod is also effective, and may reduce recurrence rates. Treatment is more difficult in the immunosuppressed, as an immune response against the virus is necessary to clear the infection. Keratinised warts may not respond as effectively to treatment due to poor penetration, and a surgical approach with curettage or shave excision may be more successful.

It is important to undertake a cervical cytology sample in these patients to exclude HPV-associated cervical disease.

Disorders associated with malignancy

Vulval intraepithelial neoplasia (VIN): is commonly associated with HPV 16, and has a varied clinical presentation. Symptoms include irritation, mild discomfort, pain or ulceration. It can however be asymptomatic, and a change in texture and/or appearance is noted. On examination, VIN presents as an area of glazed erythema, thickened, macerated skin, an area of brown pigmentation or a combination of these features (Figure 1). VIN patients are more likely to develop cervical intraepithelial neoplasia (CIN) and anal intraepithelial neoplasia (AIN), and should be examined to exclude this.

There are two main histological subtypes of VIN — well differentiated and basaloid. The latter is more common, particularly in pre-menopausal women. Untreated VIN may progress to invasive squamous cell carcinoma (SCC) in approximately 5% of cases. Unifocal disease may be best treated surgically, and recurrence is unlikely if clear margins are obtained. Multifocal or recurrent disease poses a greater problem as extensive vulval surgery may be mutilating. Alternative treatments include laser and topical immunotherapy with imiquimod or cidofovir. Both topical treatments have been reported to clear VIN, but treatment produces an intense inflammatory reaction with discomfort and occasionally ulceration which limit their efficacy.

Lichen sclerosus (LS): most frequently presents with itch, but is often initially misdiagnosed as Candida. Soreness and dyspareunia are also common complaints, but occasionally LS can be asymptomatic. Examination reveals white, shiny patches classically in a 'figure of eight' distribution affecting the vulva, perineum and perianal skin (Figure 2). The clitoral hood, clitoris, labia minora and interlabial sulci are also commonly affected. Other skin changes include bruising (ecchymoses), telangiectasia, thickening, hyperkeratosis, fissuring and ulceration. The vaginal mucosa is not involved but loss of elasticity and sclerotic change at the vaginal introitus can make intercourse painful. Patients with long-standing LS may develop architectural change due to scarring secondary to chronic inflammation damaging to the dermis and dermo-epidermal junction. This may be evident as burying of the clitoris, resorption of the labia minora, anterior