Skin diseases affecting the vulva

Moumita Chattopadhyay
Jane Sterling

Abstract
The vulva stretches between the mons pubis and the anus and is bounded by the genitocrural folds. The epithelium includes the keratinized, 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

Patients with vulval disease are often embarrassed by their problem and may delay seeking help or self-treat. Careful elucidation of the symptoms and signs of vulval disease can be sufficient to make the diagnosis (Table 1), but investigations such as vaginal swabs, allergy patch testing and skin biopsy are often needed to confirm suspicions or distinguish between diseases with similar presentations.

A 'shortlist' of the most common vulval conditions would include vulvovaginal candidiasis, dermatitis and lichen sclerosus, but there are many other less common causes of vulval symptoms which must not be forgotten. It is always worthwhile considering infection as a cause of or an exacerbating factor in vulval skin disease.

Infections

Candida

A sensation of itch, both in the vagina and in the vulval skin, combined with an increase in creamy vaginal discharge are the hallmarks of vulvovaginal candidiasis. Treatment should be preceded by vaginal swab to confirm the presence of the yeast. Chronic or incompletely treated candidiasis may present with persistent vulval erythema and fissuring in the interlabial sulci or perineum. Again, culture will help to confirm the diagnosis.

Moumita Chattopadhyay *MB BS DDVL MRCP is a Specialist Registrar at the Department of Dermatology, Addenbrooke's Hospital, Cambridge, UK. Conflicts of interest: none declared.*

Jane Sterling *MB BChir MA FRCP PhD FHEA is a Consultant at the Department of Dermatology, Addenbrooke's Hospital, Cambridge, UK. Conflicts of interest: none declared.*

Acute or chronic candidiasis should respond to imidazoles, either intravaginally as pessary or cream, or orally. Persistent symptoms can develop if only external treatment is used. Recurrent episodes may respond best to a 7–10 day course of oral fluconazole.

Tinea cruris

Ringworm of the groin is another readily diagnosable and easily-treated cause of vulval irritation. A pre-existing problem with athlete's foot (tinea pedis) may lead to the suspicion of involvement of the groin and vulval skin. In the groin, buttocks and upper thighs, fungal infection causes mild erythema with a well-defined edge with fine scaling, but in hair-bearing areas, there may be deeper involvement of the follicles with pustules or nodules. Skin scrapings to capture fine scales for fungal culture can reveal the causative agent, most commonly *Trichophyton rubrum*. If extensive, systemic therapy with terbinafine is most effective.

Easily confused with tinea is the condition **Erythrasma**, caused by corynebacteria. The infection is more common in immunosuppression or diabetes. A faint brownish discolouration of the groin with fine peeling should suggest the diagnosis and, if available, examination with Wood's light (low wave length ultraviolet) will show coral-pink fluorescence of affected skin.

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
		Behçet's disease

Table 1

Treatment for 2 weeks with topical fusidic acid or an imidazole cream is usually effective.

Folliculitis/boils

Folliculitis is visible as small papules and pustules sited at hair follicles. Careful swabbing, if possible from the pus of an intact pustule, is likely to show infection with *Staphylococcus aureus*, but other aerobes and anaerobes can occasionally cause this common infection. Damage to the skin through shaving presents with a similar picture, but the infection may be more superficial without deep involvement of the follicles and so is termed pseudofolliculitis.

The infection usually responds to a combination of topical antiseptic measures and systemic antibiotics. Although the bacteria are usually sensitive to flucloxacillin and erythromycin as well as other anti-staphylococcal drugs, recurrence is common unless the course of treatment is prolonged. General measures to reduce surface bacteria by washing and antiseptic applications 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 minimize infection recurrence.

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

Hidradenitis suppuritiva

Recurrent boils in the groin plus other flexural sites such as axillae and sub-mammary areas together with comedones and bridged scarring suggests the condition hidradenitis suppuritiva, which may be familial.

Cellulitis

Streptococcal infection of the vulva 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 may confirm the organism but treatment should be started immediately to reduce the risk of secondary lymphatic damage and hence increased risk of repeated episodes and development of lymphoedema. Penicillin, flucloxacillin or erythromycin are appropriate choices for oral antibiotics.

Crohn's disease

Vulval Crohn's disease has many features similar to those of chronic cellulitis or hidradenitis suppuritiva. It may develop before any intestinal disease or together with bowel Crohn's. Pustules, ulcers, fissures, sinuses, fistulae, papules, nodules or skin tags with inflammation, swelling and induration can all feature as part of this granulomatous inflammation. If suspected, a search for bowel abnormality is essential.

Herpes simplex

Genital herpes is not uncommon and is usually diagnosed in general practice or in genitourinary medical clinics. In primary genital herpes, the onset is acute and the pain is severe. There may 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. Aciclovir by mouth will help to shorten the duration of the acute eruption provided it is taken early in the course of the infection. If episodes are frequent, regular prophylactic aciclovir (200 mg twice daily for 6 months) can help to reduce the number of reactivation episodes.

Molluscum contagiosum

Infection with this molluscipox virus produces small pearly papules with a central dip 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.

Warts

Anogenital warts in adults are usually sexually transmitted and are caused by the low-risk human papillomaviruses (HPVs), usually type 6 or 11. The diagnosis is suggested by small papules or cauliflower-shaped growths at any site in the vulva or vagina, but the most common site is at the posterior fourchette.

The differential diagnosis includes molluscum contagiosum, epidermoid cysts, lymphangiomata, angiokeratomas, seborrhoeic keratoses and vulval intraepithelial neoplasia. If the diagnosis is in doubt, histology and possibly also molecular testing for HPVs should be obtained.

Treatment of vulval warts will depend on the number, size and position of the warts. In immunosuppressed individuals, standard therapies may not lead to clearance, as an immune response against the virus is essential to clear the infection. Topical antiproliferative or destructive treatments are used most commonly and include podophyllotoxin, trichloroacetic acid and liquid nitrogen cryotherapy. The immune response modifier, imiquimod, is also effective and can reduce the risk of recurrence. Warts that are keratinized may not respond so well as the treatment may not penetrate and in this case, surgery with snip or curettage may be the treatment of choice.

Vulval warts can be associated with acquisition of the other HPV types that are associated with cervical disease, so cervical smear testing should not be forgotten.

Disorders associated with malignancy

Vulval intraepithelial neoplasia (VIN)

Vulval intraepithelial neoplasia, like intraepithelial neoplasia of the cervix, vagina, penis and perianal skin, is closely associated with high-risk human papillomavirus infection, most commonly HPV type 16. The presentation of VIN is varied. There may be no symptoms but a change in surface texture or a change in appearance may be noted either by the patient or at examination, for instance at cervical screening. When present, symptoms include irritation, mild discomfort, pain or ulceration. Visibly VIN can appear as an area of glazed erythema, as an area of slightly thickened, macerated skin, as an area of brown pigmentation or a combination of any of these (Figure 1). VIN is more common in patients with a history of cervical intraepithelial neoplasia.

If VIN is suspected, tissue diagnosis is essential. There are two main histological types of VIN — well differentiated VIN and basaloid VIN. The latter is more common, especially in the premenopausal woman.

The main cause for concern is the risk of progression to invasive squamous cell carcinoma (SCC), which is of the order of about 5% over lifetime. Unifocal disease may be best treated surgically and as long as the resection margins are clear, recurrence is unlikely. If margins are involved, recurrence is likely.