In this article the author looks at a number of skin complaints that may affect the vulva and which are not caused by sexually transmitted diseases.

Dr Maggie Kirkup
Consultant, Western Area Health Trust

This article aims to discuss some of the most common skin disorders which affect the vulva and surrounding skin in adults. Sexually transmitted diseases and issues around female genital mutilation are not included in this article.

The vulva consists of the mons pubis, labia majora and minora, clitoris and vaginal vestibule. The surface of most of the vulva is skin, the vestibule mucosal. The vulva contains sebaceous and sweat glands as well as specific vestibular glands known as Bartholin’s glands. Enlargement of the sebaceous glands is of no clinical significance but can cause anxiety.

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There are intrinsic changes in the anatomy and physiology of the female genital skin which occur with advancing age. Atrophy of the skin and mucosa of the epithelium, reduced collagen and water content plus loss of subcutaneous fat are at least in part due to the fall in oestrogen levels at the menopause. These alterations together with the reduced vaginal secretions make the older vulva liable to shear forces and irritation. Irritants include dampness, urine, soap and other washing products, toiletries and friction from clothing. Incontinence will therefore increase difficulties because of the exposure to irritants. Elderly patients are also susceptible to contact irritation leading to dermatitis in the vulvar area as ammonia in urine elevates local pH, which alters barrier function, further compromising skin integrity and increasing risk of infection. The proximity to the vagina and rectum increase exposure to the microbes prevalent in these areas. Diseases of the female genitalia are likely to have a deleterious effect on sexual function. There may be a perception that older females do not indulge in sexual activity. However, studies of sexual activity in older adults in the USA in 2007 and 2010 reported frequent activity among older women (around 50% and 25% of those aged 65-74 and 75-95 years respectively) There was also agreement that women still engage in, or wish to engage in, sexual activity.

Many factors contribute to sexual dysfunction, among these being skin disease. The significant impact of lichen sclerosis et atrophicus (LSA) on sexual dysfunction has been recently quantified.

Delayed diagnosis of vulvar problems is common as patients may be reluctant to complain to doctors who may in turn not enquire about genital symptoms. Multidisciplinary vulval clinics involving dermatologists, gynaecologists and GU physicians are extremely important in improving patient care but are not available in every area and tend to be over-subscribed. Hence there is a need for familiarity with the common problems which can be successfully managed in primary care.

Inflammatory dermatoses

Common inflammatory dermatoses such as the eczemas and psoriasis can manifest on the vulva where the features may differ from those on any other area of skin. Seborrhoeic eczema affects skin creases such as the inguinal and genitocrural folds (the body creases around the genitals) but can also affect the skin of the vulva. Allergic contact dermatitis is rare in the female genital area but irritant contact dermatitis is very common for reasons mentioned above. Although psoriasis is most frequently found on extensor aspects of limbs and on the scalp, flexural involvement is not rare. The usual scaling of psoriasis is absent in the skin creases, the appearance being of a “beefy” red, well demarcated area. Topical applications need to be appropriate for the area, for example, avoiding potent topical steroids to reduce the risk of absorption and localised skin thinning.

Lichen simplex

Lichen simplex is the name given to a condition caused by repeated scratching or rubbing of the skin in the absence of a visible dermatosis. The appearance is likened to the bark of a tree. On the vulva this presents as a localised area, white or grey in colour, usually on
Lichen sclerosis of the vulva in a 49 year old female patient. Also known as lichen sclerosus et atrophicus, this inflammatory skin disorder that affects the vulva, penis and perianal area, leads to loss of pigment and atrophy (thinning) of the skin.

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The labia majora or mons pubis. Management plan includes taking steps to interrupt the “itch-scratch cycle” by application of potent topical steroid, use of soap substitutes and avoiding irritants. Treatment failure may be due to an unrecognised topical allergen and patch tests may reveal this.

Lichen sclerosis
Lichen sclerosis et atrophicus can occur at any age but the incidence is highest in pre-pubertal girls and menopausal women. It is one of the commonest skin disorders of the older vulva. The prognosis is unpredictable. It may follow a chronic or relapsing and remitting course and can resolve. The most frequent symptom is itch. However, pain, superficial bleeding, fissuring and narrowing of the orifices due to scarring can also occur. Difficulties with sexual penetration and urination can follow. The genital skin appears white and indurated with telangiectases and ecchymoses. Perianal involvement is frequent giving a “figure of eight” appearance. Around 10% of women with ano-genital disease also have extragenital involvement which can be on any body site, most commonly on the trunk. The cause is unknown but evidence suggests that it is a genetically determined autoimmune condition.

Histological changes include chronic inflammation and thinning of epidermis. However some cases show epidermal thickening with squamous hyperplasia. The risk of development of squamous cell carcinoma (SCC) of the vulval skin (approximately 4%) has long been recognised. Topical treatment with superpotent topical steroids remains the mainstay of treatment. The optimal regimen is uncertain. A typical course of treatment would be clobetasol propionate once nightly for four weeks, then alternate nights for four weeks, and twice weekly for a further month. Most patients require 30-60g annually allowing for occasional “as required” ongoing treatment. For resistant cases and to reduce the need for steroids, short-term use of topical calcineurin inhibitors (tacrolimus and pimecrolimus) can be introduced but they are immune suppressive and it is uncertain if there is an increased risk of neoplastic change.

A significant improvement in quality of life with topical steroid treatment has been demonstrated. Biopsy is not always practical and may be unnecessary if the clinical features are typical but should be done if there is poor response to treatment or development of raised lesions which might indicate neoplastic change.

Lichen planus
The main differential diagnosis from LSA is lichen planus (LP). There can be clinical and histological overlap and the two conditions are considered by some to be part of a spectrum. LP most commonly presents as a generalised skin eruption and 20% of patients will have genital lesions. The clinical appearance is violaceous or erythematous papules and plaques and erosions which may or may not have the typical white lines known as Wickham’s striae or a white lacy border. Involvement of oral and genital surfaces may occur without other cutaneous sites being affected. The symptoms and signs can often mimic LSA. A distinctive variant involves vaginal and gingival mucosa where painful erosive lesions predominate.

First-line management is with potent topical steroids, using a regime such as that used in lichen sclerosis, applied by steroid foam or suppositories if there is vaginal involvement. Topical retinoids and calcineurin inhibitors have been used. There are case reports of use of oral steroids, oral retinoids, methotrexate and other
systemic immune suppressants.

**Zoon’s vulvitis**

Presenting as red-brown patch of skin with a rather glazed appearance, this may be asymptomatic. The characteristic finding on biopsy is dermal infiltration with plasma cells. Zoon’s is considered to be a reactive process consistent with an irritant dermatosis or another chronic inflammatory condition. It tends to be chronic but benign, although premalignant changes can occasional look similar. Treatment is of the underlying condition and a protective barrier ointment such as yellow soft paraffin may help reduce irritation.

**Vulvodynia**

Vulvodynia is a chronic pain condition associated with local hypersensitivity of the vulva which can be provoked (e.g. by tampons or intercourse) or unprovoked or both. The majority of sufferers remain undiagnosed and inadequately treated. Prevalence in a population-based study was around 8%, the rate decreasing after the age of 70, with evidence that this was due to reduced sexual activity.

Vulvodynia is a chronic but benign, although premalignant changes can occur in the cervix, vagina and perianal areas so comprehensive examination should be carried out. Treatment options depend on the individual case.

**Neoplasia**

As the vulva is sun-protected it might be assumed that it is not susceptible to skin malignancies. However, UV exposure is only one factor in oncogenesis and importantly skin tumours do develop on genital skin. Some strains of the human papilloma virus (HPV) are known to have oncogenic potential.

Basal cell carcinomas (BCC) have the appearance of an ulcer or nodule, which can be skin coloured or translucent. They are rare on the genital skin, representing no more than 5% of vulval neoplasia and less than 1% of all BCCs. Recurrence after complete excision is very unlikely.

Malignant melanomas are usually present on sun exposed sites but can appear on genital skin. Melanoma is the second most common vulvar malignancy. They tend to have an adverse prognosis, attributed to late presentation. The majority of melanomas, at least initially are macular (flat) and can only be detected by inspecting the area. Benign pigmentation of the vulva is common, occurring in around 10% white women. It can be difficult to be certain of the nature of pigmented macules and biopsy may be necessary for accurate diagnosis.

Vulval Intraepithelial Neoplasia (VIN), previously known as Bowenoid papulosis or carcinoma in situ, represents cellular atypia and is graded depending on the percentage of epithelium involved. Malignant potential is said to exist if over two thirds of the epithelium is involved. Management of both can be difficult and these patients need long consultation times and gentle sensitive examination technique. Topical therapy with soap substitutes and regular application of 5% lidocaine ointment is first line treatment. Addition of systemic medication as used for other chronic pain syndromes may be needed e.g. pregabalin or a tricyclic antidepressant. Some individuals may benefit from contact with patient support groups such as Vulval Pain Society (see box).

The risk of progression to invasive disease is around 10%, although higher if there is immune compromise. Treatment options depend on the individual case and may include simple excision for a limited area. However, with extensive or multifocal disease, close follow-up and targeted excisions may be preferable. Topical 5-fluorouracil can be successful but not on hair-bearing areas. Imiquimod has been shown to be effective but is not currently licensed for use in this area. Regular and long-term follow-up is advisable.
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Squamous cell carcinoma (SCC) is the commonest vulvar malignancy and may develop from a chronic inflammatory condition such as LSA or LP. It can also develop from VIN with evidence of infection with oncogenic strains of HPV. These two types of SCC appear to be different entities both in terms of aetiology and clinical presentation, the non-HPV lesions tending to be keratinising. Surgical excision is the treatment of choice and five year survival is of the order of 75%, rising to 90% is there is no nodal involvement. It is of note that lymphatic drainage may be bilateral if the lesion is centrally located. Therefore staging investigations should examine lymph node basins on both sides. Radiotherapy can be a useful adjuvant treatment.

Paget’s disease of the nipple is usually associated with an underlying invasive tumour. However, the term extramammary Paget’s disease (EMPD) implies an intraepithelial adenocarcinoma. The vulva is the most common site for this condition to arise. Primary EMPD arises de novo while the secondary form is less common (25% of cases) representing skin involvement from a non-cutaneous tumour by extension or metastasis. Clinically, EMPD presents as a moist, red plaque which can mimic an inflammatory condition such as eczema or psoriasis. There is usually itching or burning discomfort. Onset is mainly after age 40. Treatment is by excision which may need to be extensive. Symptomatic relief and follow-up may be preferred by elderly patients or where there are significant co-morbidities. Topical 5FU, imiquimod and bleomycin have all been reported to be successful.

Benign genital ulceration

Recurrent acute ulceration of the vulva occurs in herpes simplex infection and this needs to be excluded in all cases. However, an important differential diagnosis is aphthous ulcers which may not necessarily be associated with oral lesions. Tending to occur at a young age they can be solitary and large, although more commonly look identical to the common oral ulcers. The cause is unknown and treatment includes analgesia, potassium permanganate soaks and topical steroids.

Where there is chronic ulceration, malignancy must be excluded. Having done so, the differential diagnosis includes infections (e.g. due to TB or other deep fungus) and many inflammatory skin conditions which cause blistering or erosions. Behçet’s syndrome is an uncommon multisystem, autoimmune condition whose symptoms include oral and genital ulceration.

Intertrigo

Intertrigo is not technically a condition of the vulva but involves the skin flexures. Inguinal and genital creases are common sites for this inflammation. The cause is often multifactorial with obesity, sweating, friction and incontinence contributing. There can be soreness and/or itching and secondary infection with bacteria or yeast following. Management should include modifying the skin environment where possible by avoid tight clothing and prolonged sitting, reducing friction by keeping opposed skin surfaces apart using appropriate dressings. Weight loss may help prevent recurrence. Topical steroids of low or medium potency, combined with antimicrobial agents can be used judiciously. There is a risk of developing chronic inflammation from irritants or sensitisation to topical preparations.

Other conditions

It is not possible in this article to cover all the known vulval conditions. While there may be a dermatosis, symptoms and signs may arise from conditions at adjacent sites e.g. Crohn’s disease, endometriosis and irritated or infective problems secondary to vaginal discharge for which the causes are many. In addition, as previously mentioned, sexually transmitted diseases and female genital mutilation are sources of concern but not dealt with here.

Summary

Skin conditions affecting the female genital skin are common and have a significant effect on quality of life. Although the risk is small, chronic inflammation in this area has potential to develop into neoplastic disease. Managing these difficult conditions may be optimised by multidisciplinary clinics.

References

For full references please visit www.bjfm.co.uk

FURTHER RESOURCES

Patient information sheets on many skin conditions including genital disorders are available from the British Association of Dermatology www.bad.org.uk/for-the-public/patient-information-leaflets Up-to-date contact details for the patient support groups are available on www.bad.org.uk/for-the-public/patient-support-groups

Patient support groups

Vulval Pain Society
Behçet’s Syndrome Society
Association for Lichen Sclerosus and Vulval Health
UK Lichen Planus
National Eczema Society
Psoriasis Association