Vulval Skin Conditions

The following might be useful:

https://www.dermnetnz.org/image-library/

**Inflammatory dermatoses**

* Eczema
* Psoriasis
* Seborrheic eczema
* Allergic contact dermatitis

**Lichen simplex**

Is 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

Presents as a localised area, white or grey in colour, usually on 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 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, often extreme.

However, pain, superficial bleeding, suturing 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 anogenital 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.

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.13 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. Patients would need referral for these treatments.

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.

**Management of Lichen Sclerosis**

* Uncomplicated vulval lichen sclerosus can be managed in primary care and diagnostic biopsies are only required if there is diagnostic doubt, or failure to respond to treatment
* However, it is preferable that no super potent topical steroid has been used for 4 weeks prior to biopsy

**Treatment**

* Appropriate vulval/genital skin care advice, including use of soap substitutes, emollients and avoidance of irritants.
* Use of lubricants for sex and or topical oestrogens if indicated, if sexually active.
* Super potent topical steroid (eg Dermovate ointment) applied at night, every night for the first month, alternate nights in the second month and in the third month and thereafter at a frequency which keeps their symptoms under control. (A 30g tube should last 3 - 4 months)

**Lichen planus**

https://www.dermnetnz.org/topics/genital-lichen-planus-images/

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. Patients would need referral for these treatments.

**Zoon’s vulvitis**

Presenting as red-brown patch of skin with a rather glazed appearance, this may be asymptomatic.

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

There are mainly two types of symptoms: vestibulitis and dysaesthetic vulvodynia.

Vestibulitis is also known as vestibulodynia, which is in fact a better term as there is no inflammation. The pain is usually localised to the vaginal vestibule. Dyspareunia is a common presenting symptom. Even light touch can cause severe pain. The cause is unknown but there may have been an episode of trauma preceding the onset.

By contrast, dysaesthetic vulvodynia is a more diffuse condition which may or may not be aggravated by touch.2This condition occurs most frequently in postmenopausal women who are not sexually active. The pain occurs spontaneously and there is an association with depression. It can follow an inflammatory dermatosis such as lichen planus (see above). Usually there is no visible abnormality. Examination needs to be carefully and sensitively carried out in both forms of vulval pain.

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.

**Neoplasia**

* **BCC**
* **Melanoma**
* **VIN**
* **SCC**
* **Extra-mammary Paget’s disease (EMPD)**

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.

Clinically, the lesions can be single or multifocal.

**They can resemble viral warts or consist of red or white plaques which can be smooth or velvety.**

The main symptom is itching.

Similar changes can occur in the cervix, vagina and perianal areas so comprehensive examination should be carried out.

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.30 Regular and long-term follow-up is advisable.

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.

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.

Extra-mammary Paget’s disease (EMPD) is 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. If patches of “eczema” are not resolving, please refer to genital dermatology. If you suspect cancer, please use the 2ww pathway.

There is usually itching or burning discomfort.

Onset is mainly after age 40.

Treatment is by excision which may need to be extensive.

Untreated, progression to invasive disease is likely, and the appearance of a raised, thickened or ulcerated area indicates this.

**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.32

**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 irritant 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.

**REFERENCE**

<https://www.bjfm.co.uk/skin-conditions-of-the-vulva>

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