vulval disorders

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Non-neoplastic disorders of the vulva

For many years a confusing variety of terms have been applied to vulval conditions,

The International Society for Gynaecological Pathologists and the International Society for the Study of Vulvar Disease have recently urged that the term chronic epithelial dystrophy should be replaced by the term

non-neoplastic epithelial disorders of skin and mucosa.

Non-neoplastic disorders;

1. Squamous cell hyperplasia (formerly hyperplastic dystrophy).

2. Lichen sclerosus

3. Other dermatoses

a. Inflammatory : e.g.; Contact Dermatitis

Intertrigo, Eczema, Psoriasis

b. Bullous lesions; e.g. STEVENS-JOHNSON SYNDROME)

c. Ulcerative lesion e.g. primary syphilis , aphthous ulcers, herpes genitals,

LICHEN SIMPLEX CHRONICUS;

(previously known as neurodermatitis)

Chronic trauma secondary to rubbing or scratching elicits lichenification, a protective response of the involved skin.Most lesions of squamous cell hyperplasia are

symmetrical bilaterally and may extend beyond the labia majora.

Behavioral risk factors for developing squamous cell hyperplasia include persistent rubbing, chronic scrubbing, and development of a chronic itch-scratch cycle . There are numerous triggering factors, including chemical substances contained within hygiene products and topical medicines .

Physical examination of affected areas reveals excoriations in a background of erythematous skin. Correlation of these physical findings with historical information is typically sufficient to reach the diagnosis. Treatment involves halting the itch-scratch cycle.

Topical steroid ointments help to reduce inflammation, whereas lubricants help to restore the skin's barrier function.

If symptoms fail to resolve within 1 to 3 weeks, biopsy is indicated to exclude other pathology.

Lichen sclerosus;

This is the commonest condition found in elderly

women complaining of vulval itch but may also be

seen in children and, less commonly, in younger

women. The cause is not known, but the condition is

associated with autoimmune disorders.

Although it most commonly affects the vulva and Peri -anal skin, lesions do appear elsewhere. The lesion is white and the skin looks thin, with a crinkled surface. The contours of the vulva slowly disappear And labial adhesions form. If the patient has been rubbing the area, the skin will become thickened (lichenified).

Genital mucosal involvement does not occur in LS; the vagina and cervix are always spared. However, some mucosal involvement at the edge of mucocutaneous junctions may lead to introital narrowing. Involvement of labial, perineal and perianal areas along with introital narrowing is referred to as *keyhole* or *hourglass* or *figure of 8*.

There is much uncertainty about the risk to patients with lichen sclerosus of developing vulval cancer.

Vulval intraepithelial neoplasia (VIN) and lichen sclerosus can coexist in the same patient, and many patients with invasive carcinoma also have lichen sclerosus in the surrounding skin. About 4 % of women with lichen sclerosus develop invasive cancer.

Although there is a 9 % prevalence of thyroid disease, pernicious anaemia or diabetes in these patients, screening for these conditions may not be of value, as in most cases the diagnosis has already been made by the time the patient present!; with her vulval complaint.

If the patient is asymptomatic, . no treatment is required. Mild itching may be helped by aqueous cream or 1 % hydrocortisone ointment applied three times daily. Some have suggested the long-term use of potent steroid creams such as clobetasol propionate. Some women benefit from 2% testosterone ointment, which should be applied twice or three times per day for 6 weeks; thereafter the frequency can be reduced to once or twice a week.

Benign tumours of the vulva;

Cystic tumours;

The commonest of these are the cysts that arise from the duct of Bartholin's gland, which lies in the subcutaneous tissue below the lower third of the labium majora.

When the duct becomes blocked, a tense retention cyst forms. The patient usually presents only after infection has supervened and a painful abscess has formed. Incision and marsupialization of the abscess and antibiotic therapy give excellent results. The pus from the abscess should be sent for culture in media suitable for the detection of gonococcal infection. In women aged over 40 years, a biopsy of the cyst wall should be sent for histological examination to exclude carcinoma. A large variety of other cystic lesions may be found on the vagina. In most cases, surgical excision and histological examination will be required to determine their nature.

Solid tumours;

The commonest are condylomata acuminata presenting as small papules, which are sometimes sessile and often polypoid. They are due to infection with human papillomavirus, usually type 6 or 11. Such lesions may involve not only vulval skin but also the vagina and cervix

and may extend around the perianal area or out onto non genital skin. The transmission of the virus is usually by sexual contact. Vulval condylomata tend to increase in size in patients using oral contraceptives or during pregnancy.

Treatment;

The treatment of single or small numbers of condylomas

consists of the application of 25% trichloracetic

acid followed by 25% podophyllin, at weekly intervals.

This combination should be applied to the lesion and

the patient asked to bathe some 6-8 h later to remove

any excess. Prolonged application can lead to excessive

skin excoriation. Podophyllin should not be used during

pregnancy. Those condylomas that are resistant to

such treatment can be treated either with liquid nitrogen

application, cryosurgery, electrodiathermy or carbon

dioxide laser.

A great variety of other epithelial and non-epithelial tumours may be found. The commonest of these are squamous papillomata, skin tags, lipomas,Epidermal inclusion cyst this common cystic lesion arises from obstruction of a sebaceous gland and fibromas. Very

rarely, normal breast tissue may be found on the vulva, as may endometriosis

Vulval intraepithelial neoplasia (VIN)

Squamous VIN-

- VIN I Mild dysplasia
- VIN II Moderate dysplasia
- VIN III Severe dysplasia or carcinoma in situ

•Non-squamous VIN-

. (Paget's disease (adenocarcinoma in situ

Natural history of VIN;

Forty per cent of women with VIN are younger than 41 years. Although histologically very similar to CIN and often occurring in association with it.

Diagnosis and assessment of VIN

Intraepithelial disease of the vulva often presents as pruritus vulvae, but 20-45 % are asymptomatic and are frequently found after treatment of pre-invasive or invasive disease at other sites in the lower genital tract, particularly the cervix

These lesions are often raised above the surrounding skin and have a rough surface

The colour is variable: white, due to hyperkeratinization; red, due to thinness of the epithelium; or dark brown , due to increased melanin deposition in the epithelial cells .

They are very often multifocal.

However, the full extent of the abnormality is often not apparent until 5% acetic acid is applied After 2 minutes, VIN turns white and mosaic or punctuation may be visible. Al though these changes may be seen with the naked eye in a good light, it is much easier to use a hand lens or a colposcope.

Toluidine blue is also used as a nuclear stain, but areas of ulceration give false-positive results and hyperkeratinization gives false negatives

Adequate biopsies must be taken from abnormal areas to rule out invasive disease. These can usually be done under local anaesthesia in the outpatient clinic using a disposable 4 mm Stiefel biopsy punch.

Treatment of VIN

Provided invasion has been excluded as far as possible, topical steroids offer symptomatic relief for many women. A strong, fluorodinated steroid is usually required. This may be applied twice or thrice daily for not more than 6 months because of the thinning of the skin that may result. Frequent review is necessary initially

If the lesion is small, an excision biopsy may be both diagnostic and therapeutic. If the disease is multifocal or covers a wide area, a skin graft may improve the cosmetic result of a skinning vulvectomy. However, the donor site is often very painful and a satisfactory result can be obtained in most patients without grafting. An alternative approach used to be to vaporize the abnormal epithelium with the carbon-dioxide laser. In practice, laser vaporization has proved to be disappointing in the UK and is now seldom used.

Surgical excision is associated with crude recurrence rates of 15-43 per cent. Close observation and re-biopsy are essential to detect invasive disease among those who relapse. Repeated treatments are commonly required

Paget's disease;

This uncommon condition is similar to that found in the breast. Pruritus is the presenting complaint. It often presents as a red, crusted plaque with sharp edges. The diagnosis must be made by biopsy

In approximately one-third of patients there is an adenocarcinoma in the apocrine glands, and concomitant genital malignancies are found in 15-25 per cent. These are most commonly vulval or cervical, but transitional cell

carcinoma of the bladder (or kidney) and ovarian, endometrial, vaginal and urethral carcinomas have all been reported.

The treatment of Paget's disease is very wide local excision, usually including total vulvectomy because of the propensity of this condition to involve apparently normal skin.