DISEASES AND SWELLINGS OF THE VULVA

ANATOMY OF THE VULVA



Mons veneris

Prepuce of clitoris Frenulum of clitoris

Vestibule

Labium minus Hymen Labium majus

Fossa navicularis

Fourchette

Perineum

Vestibular bulb

Greater vestibular gland and duct (Bartholin's) Deep transverse

perineal muscle.

M. Ischiocovernosus M. Bulbocovernosus

Urogenital diaphragm (triangular ligament)

Superficial transverse perineal muscle Perineal body

M. Levator ani

M Sphincter oni externus Gluteal muscles

Most women will present to the gynaecologic clinic, once or more in their lives, complaining from different **diseases and conditions affecting the vulva and the vagina**.

Among these conditions; *infections, benign swellings, and premalignant conditions* are the commonest.

Benign neoplasms are relatively uncommon while malignant tumours of the vulva and vagina are among the rarest in the female genital tract.

DISEASES AND SWELLINGS OF THE VULVA

Pruritus vulvae, pain, discharge, and swelling are common manifestations of vulvar diseases.

Diagnosis is often delayed because of a tendency for most physicians to treat the symptoms without proper clinical examination and histologic evaluation.

It is important to establish a specific diagnosis before initiating any therapy.

Medical history:

General medical conditions that may have vulvar manifestations; such as diabetes mellitus, Crohn's disease, atopic sensitivity, psoriasis or other skin diseases.

Urinary incontinence or *chronic diarrhoea* may result in secondary vulvar reactions.

The use of soaps, perfumes, deodorant, and nylon or tight-fitting clothing especially in a patient with an atopic history.

Previous therapeutic measures and the patient's response to such medication

Physical examination:

A) Careful inspection of the vulva: under good light, with a simple hand-held magnifying lens if necessary, is of utmost importance as many lesions are small and can be easily missed.

B) Vaginal examination; both digital and speculum are necessary because diseases of the vagina, cervix, and the vulva can be interrelated.

C) Inguinal lymph nodes: The draining lymph nodes of the vulva (superficial inguinal L.N.) should be examined on both sides.

N.B.; the clinician should always examine the patient for a systemic disease.

Diagnosis:

Definitive diagnosis of lower genital tract lesions requires *biopsy*, which is best performed by a *Keye's cutaneous biopsy punch* under local anaesthesia.

Colposcopy with 5% acetic acid or toluidine blue may aid in localizing the best biopsy site.

PRURITUS VULVAE

Pruritus means a sensation of itching, usually arousing the desire to scratch. Pruritus vulvae properly refers to vulval irritation for which *no lesion is defined*, however it is commonly used in practice to describe this upsetting symptom regardless of whether or not a cause is found

. Pruritus should be distinguished from burning sensation commonly described by many women.

A) Pruritus associated with vaginal discharge (80%):

Trichomonas vaginalis and Candida albicans are the commonest vaginal infestations associated with pruritus. They account for at least 80 per cent of all cases.
B) Pruritus without vaginal discharge (20%):
Generalized pruritus: as in cases of Jaundice, diabetes mellitus and Uraemia.

Allergy and drug sensitivity: Skin sensitivity to various chemical constituents of toilet preparations such as soaps, bath salts and antiseptics may explain some cases. Rarely idiosyncrasy to chemical or rubber contraceptives is present.

Skin diseases not specific to the vulva: as scabies and seborrheic dermatitis.

Chronic epithelial dystrophies: as lichen sclerosis.

Diseases of the anus and rectum: oxyuris worms may migrate forward to cause vulval itching.Rectal Incontinence may also cause intense pruritus.

Urinary tract disorders: Glycosuria is almost the only condition associated with pruritus. Carcinoma of the vulva

Psychogenic (neurodermatitis) Idiopathic pruritus.

EPITHELIAL DISORDERS OF THE VULVA

A variety of terms have been applied to disorders of the vulvar epithelium that produce a number of non-specific gross changes

The term *vulvar dystrophy* has been replaced by the classification proposed by the

international society for the study of vulvar disease (ISSVD)

Non-neoplastic epithelial disorders	Vulvar intraepithelial neoplasia (VIN)
1.Lichensclerosis(hypoplastic dystrophy)2.Squamoushyperplasia(hyerplastic dystrophy)3.Other dermatosis (scabies,Psoriasis,lichenplanus,condyloma accuminata).	 Squamous VIN: type I, II, or III Non Squamous VIN: *Paget's disease *Melanoma in situ

ISSVD Classification of Vulvar epithelial disorders (1989)

NON-NEOPLASTIC EPITHELIAL DISORDERS OF THE VULVA

Lichen sclerosis

Squamous cell hyperplasia

Other dermatosis (scabes, Psoriasis, lichen planus, Condyloma accuminata

Lichen sclerosus et atrophicus:

Incidence: This is the commonest condition found in the *elderly women* complaining of vulval itching but may also be seen in children and younger women.

Aetiology: The cause is not known but it is associated with *autoimmune* disorders.

Clinically: In early lesions the skin may be reddish or of normal colour, later on it looks *thin* with a crinkled *surface* and <u>white shiny plaques</u>. The *contour* of the vulva slowly disappears and labial fusion may form.

Microscopically: there is hyperkeratosis, flattening of the rete pegs, and a zone of a homogenized pink staining collagenous appearing tissue beneath the epithelium. Lichen sclerosus is often associated with foci of both hyperplastic epithelium and atrophic changes.

Premalignant potential: It is uncertain if lichen sclerosus leads to vulvar cancer. VIN and lichen sclerosus can coexist in the same patient. Approximately 4% of women with lichen sclerosus develop invasive cancer.

Treatment: Topical application of testosterone ointment offers the best results.

Squamous cell hyperplasia:

The term is applied to lesions of epithelial *thickening* and *hyperkeratosis* with no specific cause.

The lesion may have a dusky red appearance when the degree of hyperkeratosis is slight.

At other times well-defined white plaques may be seen.

Lichenification is seen frequently,

while fissures and excoriation, as a result of chronic scratching, may be present.

Hyperplastic lesions are best treated by local application of cortisone.

VULVAR ULCERS

Most vulvar ulcers are benign however any persistent ulcer should be biopsied to exclude malignancy. Vulvar ulcers may be:

Traumatic e.g. infected tear or episiotomy

Syphilitic ulcer (painless)

Tuberculous ulcer (painful)

Herpetic ulcer (multiple and painful)

Carcinoma ulcer (everted edges, indurated base and floor with necrotic tissues)

VULVAR SWELLINGS

A swelling or a mass of the vulva should be differentiated from a mass protruding from the vulva, as cystocoele, rectocele, uterine prolapse, large fibroid polyp, inversion of the uterus, and Gartner cyst.



Vulvar swellings may be non neoplastic, or neoplastic (benign or malignant).

Non neoplastic swellings	Neoplastic swellings
l.Congenital	l.Benign tumours
2.Pretension cysts	2.Malignant tumours
*Bartholin duct cysts	
*Sebaceous cyst	
*Epidermal inclusion cyst	
*Hydrocoele of the canal of	
Nuck	
1.Endometrioma	
2. Traumatic lesions	
3. Circulatory disorders	
4.Inflammatory conditions	

NON-NEOPLASTIC SWELLINGS OF THE VULVA

1. Congenital: Hypertrophy of the clitoris

2. Retention cysts: These are the result of blockage of ducts of glands:

A. Bartholin's duct cyst:

The *commonest* vulvar swelling. In most cases is a cyst of the duct not of the glands.

The cyst contains *mucoid fluid* and is lined by transitional epithelium. *Clinically* it appears as a *cystic* swelling of variable sizes in the *posterior* part of the labium majus. Secondary *infection* leads to *Bartholin abscess*.

Treatment is by **marsupialization** to create a new opening between the duct wall and the skin. This line of treatment is preferable to excision as it is (a) easier with less bleeding (b) preserves the function of the gland (c) short convalescence.

N.B.: The fistulous tract created can best be preserved by placing a Word catheter in the cyst, inflating its bag with sterile water and leaving it in place for few weeks until it falls out. (Word catheter is a 10 Foley catheter, 2.5 cm long with a single barrel).





Bartholin cyst

Bartholin abscess



Bartholin duct cyst



Marsupiaalization of the cyst

B. Sebaceous cyst:

Sebaceous cysts usually present on the hairy region of the *labium majus*, as multiple small cysts that contain whitish cheesy sebaceous material.

The cyst may become infected and cause pain.

If painful they can be surgically removed.



C. Epidermal inclusion cyst:

Traumatic inclusion cysts: may occur from viable stratified squamous epithelium buried beneath either skin or mucosa, which will proliferate and desquamate forming an inclusion cyst. They may be seen at the site of circumcision (clitoridal cyst).

Epithelial inclusion cysts: The majority of epithelial cysts of the vulva are unrelated to traumatic implantation of skin fragments. They may have origin from embryonic tissue destined to become epithelium which remains in the dermis, or from pilosebaceous ducts that become occluded.

Treatment: Epidermal inclusion cysts are usually small and asymptomatic. If but if they are annoying the patient or become infected they



D) Hydrocele of the canal of Nuck:

Origin: During development the round ligament, which is inserted in the labia majora, is normally accompanied by a peritoneal pouch called the canal of Nuck which will be normally obliterated later on. Rarely if not obliterated, a cystic collection of serous fluid will accumulate in this pouch forming a Hydrocele of the canal of Nuck.

Clinically: it forms an elongated translucent swelling in the upper part of the labium majus and may extend to the inguinal canal. Sometimes it can be emptied by pressure on lying down if it communicates with the peritoneal cavity and it may be associated with inguinal hernia.

Treatment: Excision of the sac and dealing with any inguinal hernia.



3. Endometrioma:

Origin: Many suggestions have been made including; a congenital lesion, metaplastic change, embolic origin from the uterus, true neoplastic growth

Clinically: It is small bluish cyst containing altered blood and lined by endometrial epithelium that increases in size and becomes tender during menstruation.

Treatment. If painful or annoying it may be surgically excised.

4. Traumatic lesions:

Haematoma of the vulva may occur after labour. It spreads widely because of the loose tissue structure. Treatment is by incision, evacuation and drainage.

5. Swelling due to circulatory disorders:

Varicose veins: Usually occur during pregnancy as tortuous dark blue soft structures in one or both labia, which become prominent on standing and empty on lying down. *Treatment* is by excision or injection of sclerosing agents.



Oedema: It can be the result of a wide variety of systemic and local causes such as nephritic syndrome heart failure and allergic reactions

Oedema is usually diffuse and characterized by pallor of the skin and pitting on pressure with the finger.



6. Inflammatory conditions: Inflammatory vulvitis results in a diffuse swelling that surrounds the area of the inflammation due to congestion and oedema.

Some specific inflammatory conditions such as Bartholinitis, molluscum contagiosum, lymphogranuloma venereum, and condyloma accuminata give rise to localised lesions.





BENIGN NEOPLASMS OF THE VULVA

A) Cystic tumours Benign cystic tumours of the vulva are very rare

B. Solid tumours:

Lipoma: is found rarely. It arises from the subcutaneous tissues of the vulva and usually becomes pedunculated and dependent with growth. Treatment is by excision.

Fibroma: is also uncommon but present as a pedunculated tumour like a lipoma but is firmer. It arises from the fibrous tissue of the round ligament and the vulvar connective tissue. Treatment is by excision. These tumours on occasion become sarcomatous.

Squamous cell papilloma is usually a single small lesion, formed of papillae of stratified squamous epithelium, covering a core of vascular connective tissue the clinician has to differentiate it from: condylomata accuminata caused by HPV virus which are usually multiple, fibroepithelial polyps which are small, sessile, or pedunculated pieces of redundant skin, and bilharzial papillomata.



*Hidradenoma: It is an unusual lesion that originates

- * from the apocrine sweat glands.
- *It appears as a small nodule on the labium or the
- * interlabial sulcus.
- * Diagnosis and treatment is by excisional biopsy.
- * The hidradenoma is not malignant, although on histologic
- * examination the papillary nature of the tumour may be
- * mistaken for an adenocarcinoma.



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*Nevus: Pigmented nevi occur on the vulva as they do else where, but junctional activity, which carries a risk for subsequent malignant transformation is more common in this location.

* Excisional biopsy should be performed on all pigmented lesions on the vulva so that tissue can be sent for histologic evaluation.

* These lesions should not be treated by cryosurgery or laser therapy as histologic examination is essential.

Caruncle: A small tumour arising from the posterior part of the lower end of the urethra. It is composed of a very vascular stroma, almost like a haemangioma, usually infected and covered with squamous or transitional epithelium.

The caruncles are red in colour because of their vascularity and are extremely sensitive.

The patient is usually an elderly women complaining of dysuria and bleeding.

Treatment is by excision and histological examination although malignant change is rare. The base of the tumour on the urethral mucosa should be cauterised.

N.B Granulomatous caruncle: It is a chronic infection of the periurethraltissue. It is called a caruncle but it is **not neoplastic** and is often symptomless.

Granulomatous caruncle is often seen, while a true caruncle is uncommon.

Treatment if needed is by cautery and there is a tendency to recurrence.

Infection in this area must involve the paraurethral gland network and complete cure is difficult. A search should be made for a vaginal or bladder source of the infection. Other tumours include haemangioma, neurorofibroma.

PREMALIGNANT LESIONS OF THE VULVA

VULVAR INTRAEPITHELIAL NEOPLASIA (VIN)

VIN are classified after ISSVD (1998) into squamous and non squamous VIN.

Squamous VIN includes three degrees namely; VIN I, II, and III. They have been formerly knownas hyperplastic dystrophies with atypia.

Non squamous VIN includes Paget's disease and Melanoma in situ.

A. Squamous VIN

These include three degree namely ; VIN I, II, and III. They have been formerly known as hyperplastic dystrophies with atypia. The histological features and terminology of VIN and its grades I, II, & III, are analogous to those of cervical intraepithelial neoplasia "CIN" (see cancer cervix) although they seem to have a less malignant potential. It commonly occurs in the younger age where almost half of the cases are younger than 41 years.

Symptoms: 1/3 of cases are *asymptomatic*, however VIN often presents as *pruritus vulvae*.

N.E. inspection: Lesions are often raised above the surroundings skin with a rough surface.

Colour is variable: White due to hyperkeratinization, red due to thinning of epithelium, or dark brown due to melanin deposition in the epithelial cells.

Multicentric lesion: VIN lesions are often multifocal, that is why wide excision is mandatory.

VIN III or carcinoma in situ





Non squamous VIN

Non squamous VIN includes 1- Paget's disease (adenocarcinoma in situ) 2-Melanoma (is very rare.)

Paget's disease: this uncommon condition is similar to that found in the breast.

Clinically: Pruritus is the presenting complaint It often presents as a red crusted plaque with sharp edges.

Diagnosis must be made by biopsy. In almost 1/3 of cases there is an associated adenocarcinoma in the apocrine gland, and in 20% concomitant cervical cancer may be present.

Treatment is by very wide local excision usually including total vulvectomy, because unlike VIN III the histologic extent of Paget's disease is frequently beyond the visible lesion.

INVASIVE CANCER OF THE VULVA Incidence and Aetiology

Malignant tumours of the vulva are uncommon however during recent years it appears that this incidence has been increasing, reaching up to 8% of malignancies of the female genital tract.

This increase is both attributed to the continued rise in average age of female population, and the larger prevalence of VIN in the younger population.

No race is spared and neither parity nor gravidity is involved

There are two different aetiologic types of vulvar cancer.

One type is seen mainly in *younger patients*; is related to HPV infection and smoking, and is commonly associated with vulvar intraepithelial neoplasia (VIN). Carcinoma in situ of the vulva (VIN-III) appears to carry a significant risk of progression to invasive cancer if left untreated.

The more common type is seen mainly in the *elderly women*, is unrelated to smoking or HPV infection and concurrent VIN is uncommon.

Most tumours are *squamous cell carcinomas* (92%), with melanomas, adenocarcinomas, basal cell carcinomas, and sarcomas occurring much less frequently.

Squamous cell carcinoma Squamous cell carcinoma of the vulva occurs mainly in postmenopausal women , and the mean age at diagnosis is 65 years.

Symptoms: Patients generally present with long standing pruritus, before a vulvar lump is detected.

Other common symptoms include vulvar pain, bleeding, and discharge.



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Signs on examination: The lesion may be raised, ulcerated, pigmented, or warty in appearances. Most lesions occur on the *labia majora*; the labia minora are the next most common sites.

Less commonly, the clitoris or perineum is involved . Approximately 5% of cases are multifocal

The groin lymph nodes should be evaluated carefully and a complete *pelvic examination* should be performed.

Colposcopic examination of the cervix and vagina should be considered to exclude the common association with other intraepithelial squamous neoplasia of the lower genital tract (CIN & VAIN).

Diagnosis: Definitive diagnosis requires *biopsy* from the lesion under local anaesthesia.



Methods of spread:

Direct extension to adjacent structures, such as the vagina, urethra, and anus.

Lymphatic embolization to regional lymph nodes (common).

Haematogenous spread to distant sites, including the lungs, liver, and bones (rare

Lymphatic pathways: In most cases, the initial lymphatic metastases are to the inguinal lymph nodes, located between Camper's fascia and the fascia lata. From these superficial nodes, spread occurs to the femoral nodes located along the femoral vessels

. Cloquet's node, which is situated beneath the inguinal ligament, is the most cephalad of the femoral node group.

From the inguinofemoral nodes spread occurs to the pelvic nodes, particularly the external iliac group.

The incidence of lymph node metastases in vulvar cancer is approximately 30%. It is related to the size of the lesion and the stage of the disease. Haematogenous spread usually occurs late in the disease and rarely occurs in the absence of lymphatic spread.



Clinical Staging

Staging was based on a clinical evaluation of the primary tumourand regional lymph nodes with a limited search for distant metastases.

International Federation of Gynaecology and Obstetrics staging of vulvar carcinoma 1989 (FIGO)

Stage 0	Carcinoma in situ, intraepithelial carcinoma
Stage I	Tumour confined to the vulva or perineum, or both (2 cm or less in greatest dimension); no nodal metastases
Stage II	Tumour confined to the vulva or perineum, or both (more than 2 cm in greatest dimension) no nodal metastases.
Stage III	Tumour of any size with adjacent spread to the urethra or vagina, or both, or the anus, or unilateral regional lymph node metastases or a combination
Stage IV a	Tumour invades any of the following; upper urethra, bladder mucosa, rectal mucosa, pelvic bone or bilateral regional node metastases, or a combination
Stage IV	Any distant metastases including pelvic lymph nodes

Management of Invasive cancer vulva:

Radical vulvectomy and en bloc groin dissection, with or without pelvic lymphadencetomy has long been considered the standard treatment for all operable patients, resulting in a corrected 5 year survival rate of nearly 90% for stages 1 and 2 disease.

Such extensive surgery had a very high morbidity affecting the quality of life after the procedure.





I de discussion Lesion is seen on right lower vulva.

During the past 20 years a number of significant advances have been made in the management of vulvar cancer that have markedly decreased the physical and psychological morbidity associated with the standard treatment. 1- Separate incisions are used for groin dissection; to decrease post operative wound breakdown



2-Postoperative pelvic and groin external radiation therapy; has become the standard treatment for patients with positive groin LNs, thus eliminating the need for routine pelvic lymphnectomy, unless metastases is documented in the inguinal node area. 3- Very early tumours in which the depth of penetration is less than 1 mm, groin dissection may be eliminated, where a wide and deep local excision (radical local excision) is as effective as radical vulvectomy in preventing local recurrence 4-In unilateral lesions on one of the labia majora, unilateral inguinofemoral lymphadencetomy is an acceptable approach.

5-For midline lesions invading more than1 mm bilateral groin dissection is necessary

6-In advanced vulvar cancer involving the proximal urethra, anus, or rectovaginal septum many centres have been using preoperative radiationor chemo radiation to shrink the primary tumour followed by more conservative surgical excision.

Prognosis

The 5-year survival rates ranges from nearly 90% for stage 1 to 15% for stage IV.

Patients with nodal involvement have a 5 year survival rate 50% whereas

those with no nodal involvement have a 5 year survival rate of about 90%.

