Vulval Disease

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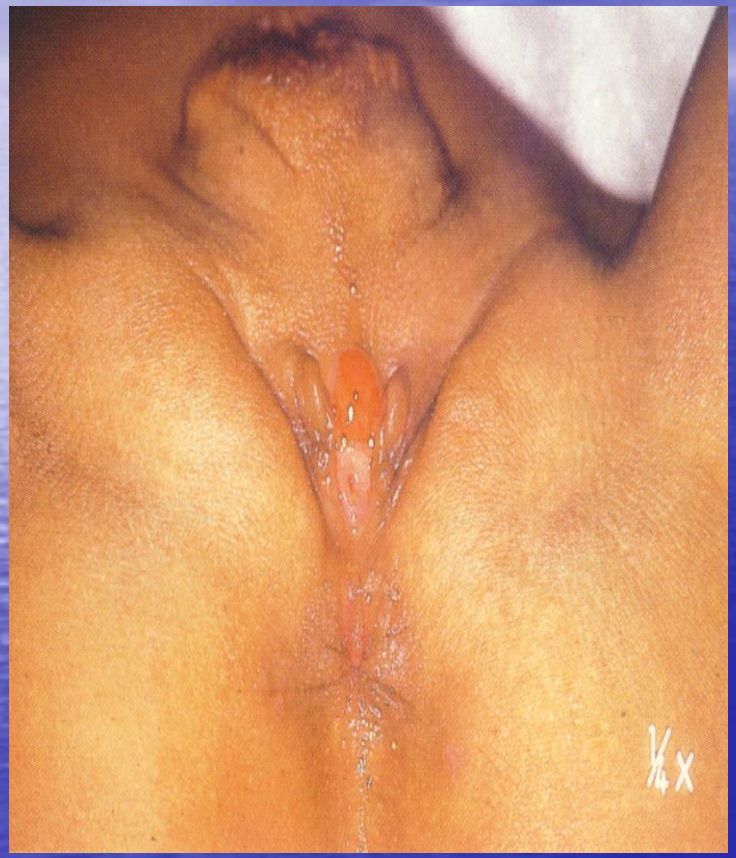
Vulval disorders

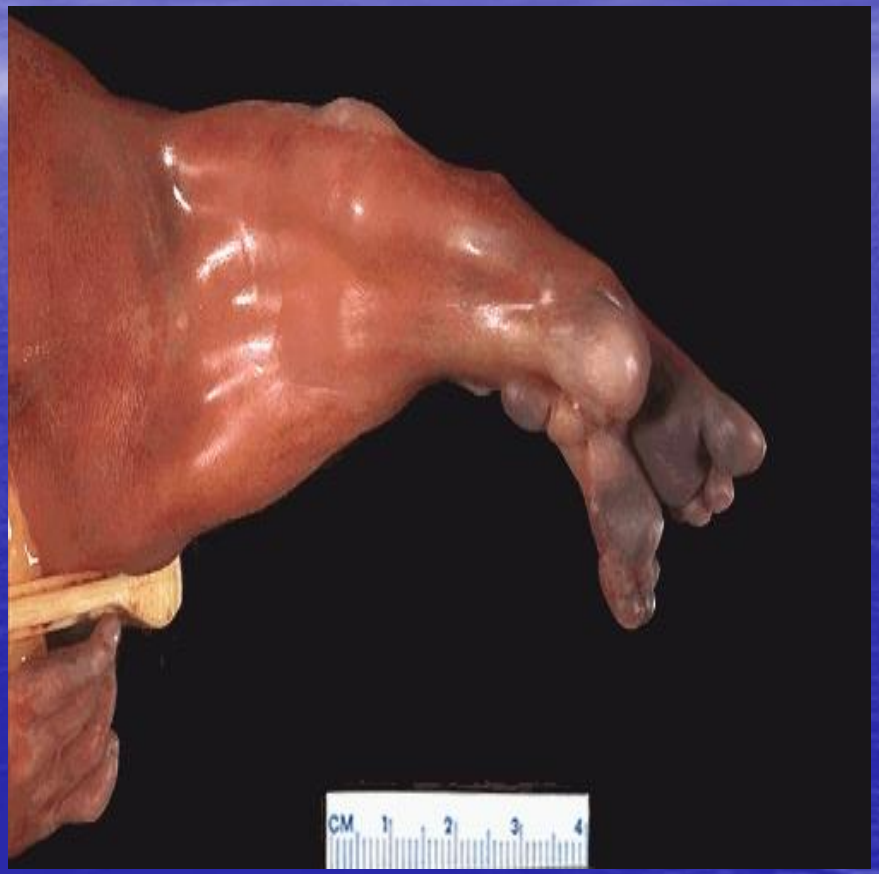
- The skin of the vulva is like that of other areas of the body with keratinized stratified squamous epithelium
- The monis pubis and labia majora contain fatty tissue, sebaceous glands, apocrine glands, eccrine sweat glands and blood vessels that can form varicosities
- The labia minora are rich with sebaceous glands but have few sweat glands and no hair follicles
- The epithelium of the vestibule is neither pigmented nor keratinized and it contains eccrine glands

Developmental anomalies

- Aplasia
- May present with or without developed hind limbs
- *Severe agenesis results in obliteration of perinum with absence of genital, urinary or anal orifices and fusion of lower limbs into one extremity (sirenomelia) incompatible with life.

- *With presence of lower limbs absent genitalia is noted by smooth perineum without orifices. Usually associated with extensive internal developmental defects





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- Fusion

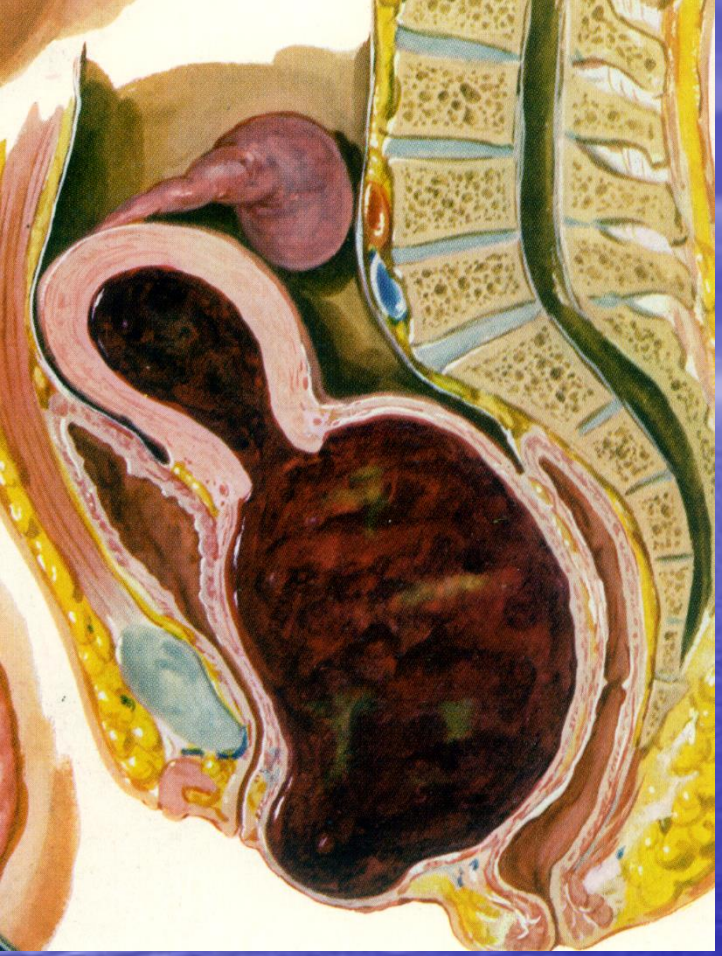
- Agglutination of the labia present from dense adhesions that hold the labia minora and majora together in the midline. This probably result from postnatal inflammation which is unnoticed

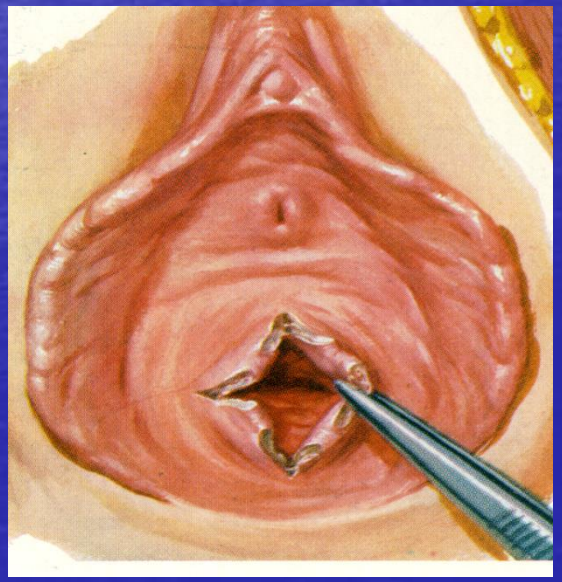
- Occasionally, fusion is complete as to stimulate the median raphe of male perineum and result in disposal of urinary stream. This is treated by separation of the adhesions and application of surface ointment to prevent recurrence

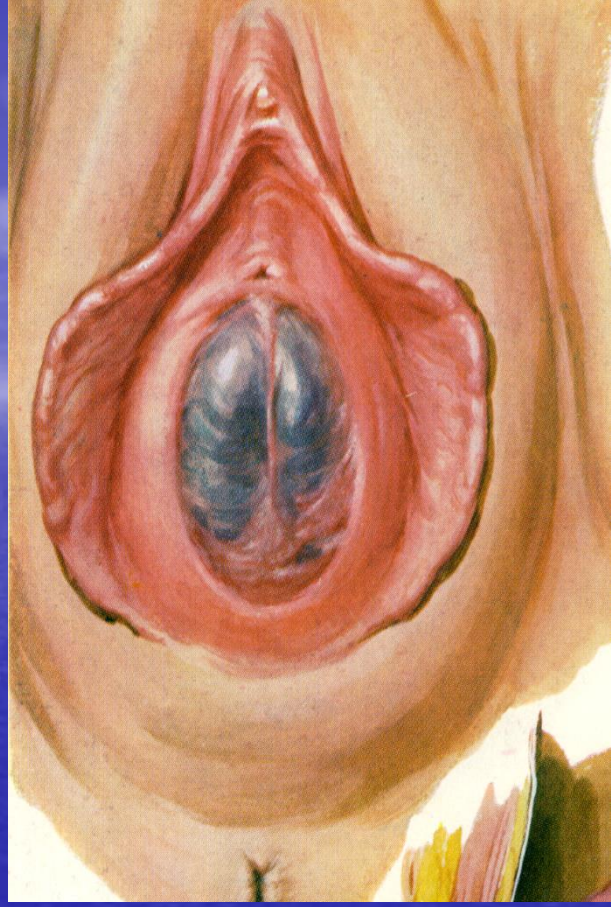
- **Imperforate hymen**

- The vaginal plate develops near the junction of the lower part of the vagina and the vestibule. Canalization of this plate is generally completed by the 6th. Month of fetal life. Failure of the final process of canalization occurs and results in imperforate hymen which obstruct the menstrual outflow

- Presentation:
- Usually present with primary amenorrhoea with monthly pain
- Sometimes present with retention of urine due to collection of blood in vagina (hematocolpos), uterus (hematometra) and tubes (hematosalpinx)
- Treated by cruciate incision or excision of hymen







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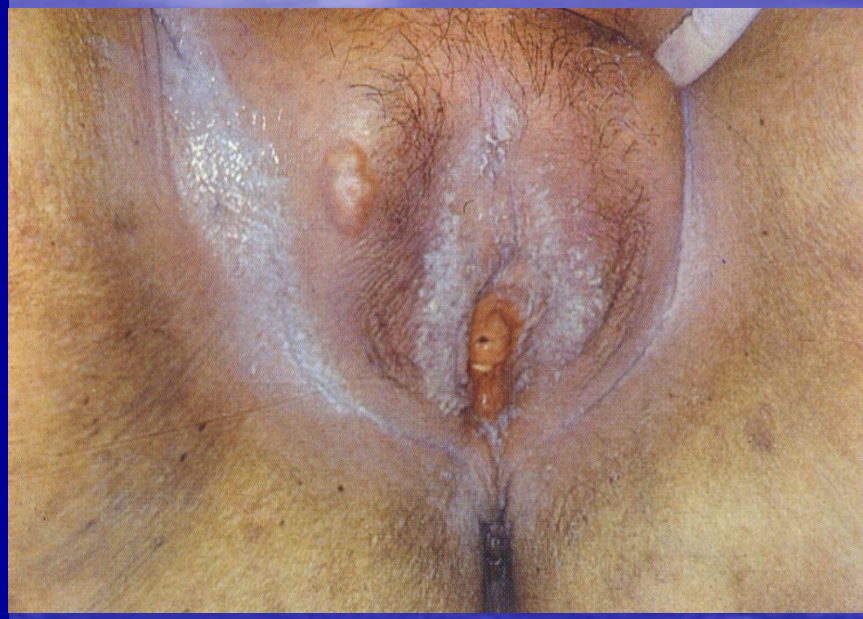
None-neoplastic Epithelial Disorders

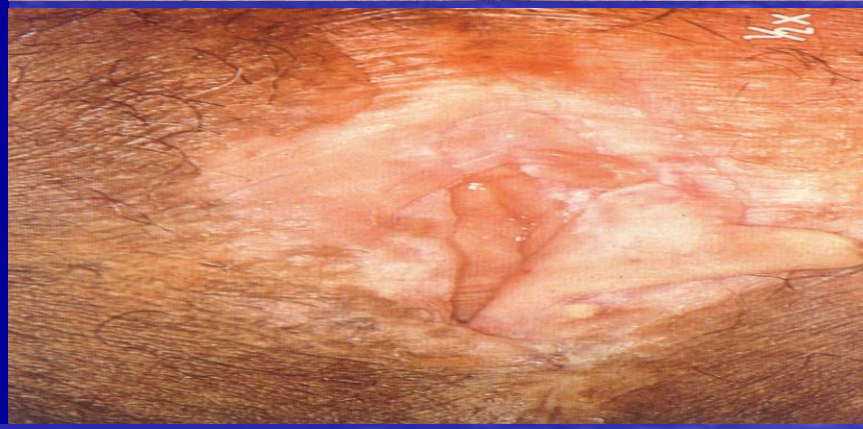
- This was formerly referred to as vulvar dystrophies, and renamed under a classification scheme adopted in 1987 by the International Society for the Study of Vulvar Disease(1987):
- 1-Lichen sclerosus 2-Squamous cell hyperplasia
- 3-Other Dermatosis:
- i-Primary irritant dermatitis
- ii-Allergic dermatitis iii-seborrheic dermatitis
- iv-Psoriasis v-Lichen planus
- vi-Hidradenitis suppurative vii-Behcet's syndrome

Lichen Sclerosus

- Common cause of white lesion of the vulva of unknown cause
- Can occur at any age but most common in the postmenopausal years
- At menarche symptoms and signs can improve spontaneously but it is not known whether there is long-term risk of recurrence

- Symptoms: Intractable pruritis is quite characteristic with less burning or pain, in early stage it may be asymptomatic
- Signs: lesion is crinkled appearance of skin (cigarette-paper) or parchment-like (shiny, delicate, pale, and wrinkled)
- Commonly extend around the anal area in a figure-of-eight or keyhole configuration
- There may be loss of the normal architecture with atrophy of the labia minora, constriction of the vaginal orifice (kraurosis)





Lichen sclerosus

- Histology: hyperkeratosis, epithelial thinning with flattening of the rete pegs, cytoplasmic vaculation of the basal layer of cells, homogenization of the subepithelial layer and inflammatory cell infiltration consisting of lymphocytes with few plasma cells

- Treatment
- Topical corticosteriods provide the best outcomes
- Use of systemic retinoids is currently being studied

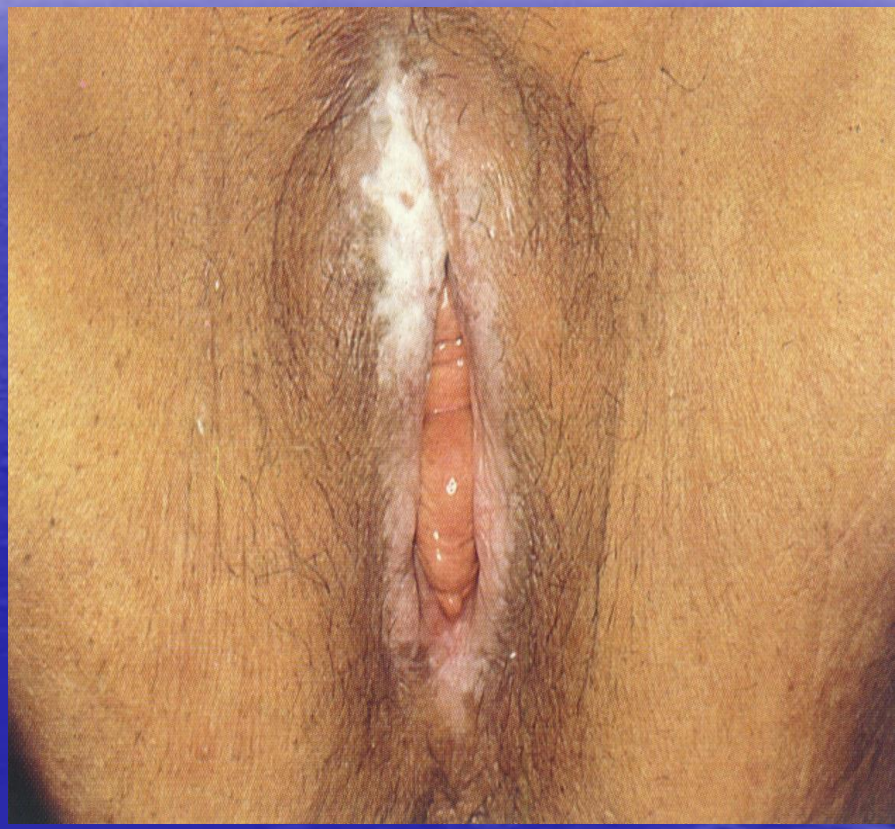
Squamous cell hyperplasia

- Generally result from repetitive surface trauma from irritants that cause scratching or rubbing---the notorious-itch-scratch cycle

- Clinical: a variety of stimuli cause pruritus and chronic irritation and inflammation result in excoriation, fissures and thickened, whitish hyperkeratotic patches, usually localized and well delinated but on occasion may be extensive and poorly defined

Squamous hyperplasia

- Histology: thickening of the keratin layer (hyperkeratosis) and lengthening and distortion (acanthosis) of the rete pegs and inflammatory cells in the dermis



Treatment

- General attention to proper hygiene
- If the skin is moist or macerated, aluminum acetate 5% solution applied 3-4 times daily for 30-60 minutes
- Systemic antihistamines or tricyclic antidepressants may help
- Treatment of choice is application of a potent corticosteroid cream ..betamethasone valerate 0.1 to 0.25%
- Surgical treatment better avoided because the recurrence rate is 40-50%

Follow-up

- Mixed diagnosis account for 20% of non-neoplastic epithelial disorders and vulvar neoplasia occurs more often in this setting
- The likelihood that superimposed carcinoma will develop from none-neoplastic epithelial lesions has been estimated to be 1-5% and is considered to be greater with lichen sclerosus or a lesion with atypia on initial biopsy
- Long-term surveillance of these lesions is recommended

Inflammatory Dermatoses

- Contact or primary irritant dermatitis
- Allergic dermatitis
- Sebaceous hyperplasia
- **Chronic Dermatosis**
- 1-Seborrheic dermatitis
- 2-Psoriasis
- 3-Lichen Planus
- 4-Hidradenitis suppurativa

Contact Dermatitis

- Primary irritant dermatitis is a common cause of vulvar irritation, burning and pruritus
- Common irritants include:
- *perfume *soap *sprays and deodorants
- *bath oils *colored toilet paper *urine *semen
- Also irritation arises from friction from tight clothing, trapping of moisture due to lack of ventilation (from wearing synthetic fabrics) or activities such as bicycling and horseback

Contact dermatitis

- Typical physical findings are diffuse reddening of the involved skin with areas of excoriation and ulceration
- Secondary infection may occur and candidal vulvitis can mimic this dermatosis
- Treatment
- *Identify and avoid irritants
- *symptomatic treatment by cool sitz bath
- *Aluminum acetate 5%(Burrow's solution)
- *Short term use of fluorinated corticosteroid ointment for symptomatic relief(avoid long-term use which can cause atrophy of skin)

Allergic Dermatitis

- Common in atopic individuals
- Known allergens include pollen, ingested foods or drugs, topical medications, semen, latex
- Physical examination reveals dryness, scaling, excoriations and at times ulceration

- **Diagnosis** made on basis of history. Vaginal smear may show eosinophils and serum eosinophilia may be present
- **Treatment**
- Identify the offending agent
- Use of oral antihistamine, short –term use of a topical corticosteroid ointment
- In rare cases immunotherapy
- If untreated may progress to sqaumous cell hyperplasia

Sebaceous Hyperplasia

- **Chronic inflammation and advancing age can lead to hypertrophy of the sebaceous glands of the labia majora**
- **Characterized by a cobblestone appearance and is localized to the inferior and medial aspect of the labia majora**
- **It is a benign condition not requiring treatment, but may be confused with other inflammatory processes**

Chronic Dermatoses

- **Seborrheic dermatitis**
- Occurs in areas of the skin where sebaceous glands are most active e.g face, scalp, body folds
- When vulva is involved, the labia majora and mons pubis primarily are affected
- Lesion appear as dry to greasy scales with ill-defined borders and usually pruritic
- Treatment: symptomatic relief by Burrow's solution and short –term use of steroid ointment
- Ketoconazole may help as Malassezia ovalis could play a role
- Apply medications immediately after bathing when keratin is softer to maximize dermal penetration

Psoriasis

- Hereditary disorder of the skin that affects 2% of population in the western world
- Lesion: silver-white present in sites with repetitive trauma e.g scalp, elbow, forearm, knee, hands, feet
- Typically involves the lateral aspects of the labia majora and the genitocrural areas and is characterized by erythematous patches without scales and it may be pruritis with areas of maceration
- Treatment:symptomatic with the use of calcipotriene ointment(Dovonex)..a topical vitamin D3 preparation
- In sever cases Mthotrexate can be used

Lichen Planus

- Can be acute or chronic and affect the skin or mucous membranes or both
- Cause is uncertain but evidence suggests that it is immunologically mediated
- Appearance ranges from delicate white reticulated papules to an erosive desquamting process. Diagnosis confirmed by biopsy
- Treatment: high-potency corticosteroid ointments or intralesion corticosteroid injections

Hidranitis suppurativa

- Chronic suppurative inflammatory disorder of the apocrine glands
- More common in black in second or third decades of life
- Lesion:deep painful subcutaneous nodules that may ulcerate and drain, leading to open sinuses and extensive scarring
- In vulva it primarily affects labia majora and intercrural folds, buut may affect clitoris,labia minora and mons pubis as well
- Surgery with wide excision of the involved areas sometimes is necessary for treatment of abscess and sinus tract formation
- Topical and systemic antibiotics, oral contraceptives, corticosteroids and leuprolide acetate gave limited success
- Because of similarity with acne vulgaris treatment with isotretinoin (Accutan) gave good results in selected cases

Ulcerative Dermatoses

- Ulecerative lesions may be solitary or multiple and may be painful or non-tender
- *ulcers arising from vesicles are typical of **Herpes simplex (HVS)** infection
- Ulcers that arise from papules are characteristic of:
- 1-**syphilis** 2-**chancroid**
- 3- **Granuloma inguinal** 4-**Carcinoma**
- 5- **Lymphogranuloma venereum**
- 6- **Behcet's disease and crohn's disease**

Ulcers of vulva

- Solitary, non-tender ulcers a chracteristic of syphilis, lymphogranuloma venereum, and neoplasia
- Multiple painful ulcers occur in HSV, Behcet's disease and crohn's disease
- To deteromine the cause:
- 1-laboratory evaluation including serologic testing and culture is often necessary
- 2-solitary ulcer should be biopsed onece other entitis are excluded because 1/3 of vulvar neiolasm may ulcerate

- **Behcet's syndrome**
- Is a multisystem panvasculitis characterized by multiple painful genital and oral ulcers
- Other manifestifation include acneiform lesions or cutaneous nodules on the skin, thrombophelbitis and synovitis
- Diagnosis: based on history of recurrent oral ulcers in conjunction with genital ulcers, eye findings or skin lesions
- Local symptmatic treatment by corticosteroid ointments or intra-lesion injection with triamcinolone

Benign tumors of vulva

- Types of benign vulval tumors
- **1-Cystic**
- i-mucous cyst
- ii-cyst of canal of Nuck
- iii-Bartholin's cyst
- iv-skene's duct cyst
- V-epidermal inclusion cyst vi-frunculosis
- Anatomic: *varicosities *hernia

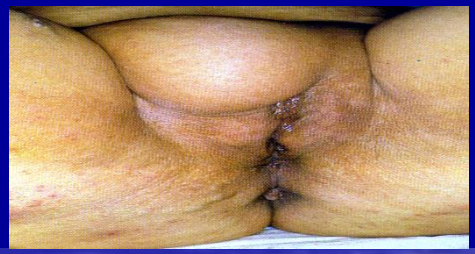
- **2-Solid tumors**
- i-Lentigo,nevus
- ii-seborrheic jeratosis
- iii-fibroepithelial polyp
- iv-papillomatosis
- v-fibroma, fibromyoma
- vi-lipoma
- vii-hidradenoma

Cystic tumors of vulva

- **Mucous cysts** (dysontogenetic) arise from minor vestibular glands or from mesonephric duct remenants and may be found at the interoitus and labia minora
- **Cyst of canal of Nuck** (processus vaginalis peritonei): filled with peritoneal fluid can give rise to a hydrocele located high in the labia majora, iguinal hernia present in 30% of cases

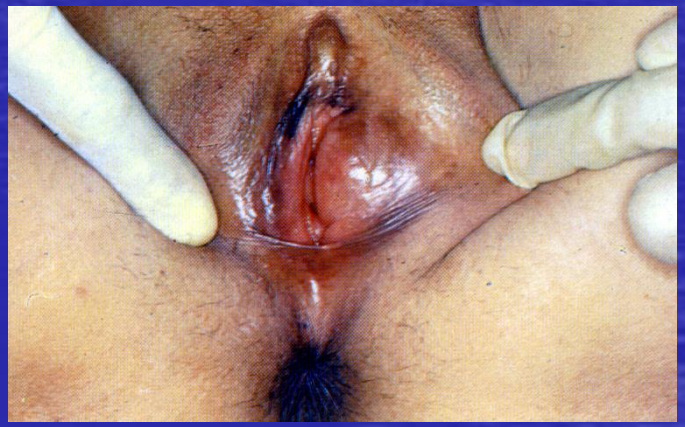
- Bilateral cyst of canal of Nuck with left inguinal hernia and cystocele

- Hydrocele of canal of Nuck



Cysts of vulva

- **Bartholin's cysts**
- Are the most common vuvar cystic growths result from ductal occlusion secondary to infection, trauma or neoplasm
- Symptoms: vulval swelling, dyspreunia, pain with walking
- Treatment:
- *asymptomatic cyst require no treatment
- *if painful, but not indurated, aspiration and antibiotic
- *Abscess treated by incision and drainage with creation of new ostia
- *LASER incision with cyst wall vaporization of the duct or marsuplization is indicated in cases of persistent or recurrent infection
- *Excision of the gland; in women over 40s to rule out adenocarcinoma



Cysts of Vulva

- **Skene duct abscess** occurs adjacent to urtheral meatus and if large enough may cause urinary obstruction
- Treated by incision and drainage
- **Epidermal inclusion sebaceous cyst**: common arise from obstruction of sebaceous gland, more common in vagina, usually asymptomatic unless infected and become painful, require incision and drainage
- **Folliculosis or Frunculosis:** result from obstruction of hair follicles or pilosebaceous unit, when infected folliculitis become papule or pastule and frunculosis become abscess which needs incision and drainage. These may be manifestation of hidradenitis supurative

Solid benign tumors of Vulva

1-Pigmented lesion occurs in 10-20%

- Lentigo is the most common pigmented lesion appear as small macule <5mm
- Vulvar navi are also common
- Seborrheic keratosis. Also common and is similar to that in other arts of body
- 2-Fibroepithelial polyp (acrochordon) is common in areas subject to irritation and is easily removed
- 3-fibroma: most common, develop along the line of insertion of round ligament into labia majora
- 4-Lipoma: second most common and removed if csmotically unacceptable or when become painful
- 5-Hidradenomas: usually 1-1.5cm in diameter occur in interlabial sulcus and always solitary and do not become malignant

Vulval Intraepithelial Neoplasia (VIN)

- Sqaumous epithelial lesion characterized by sidordered maturation and nuclear abnormalities such as loss of polarity, pleomorphism, large dark nuclei with increased mitotic figures which may be atypical, irregular and irregular membrane

VIN

- Grades
- VIN 1 abnormalities present in the lower third of the epithelium
- VIN2 abnormalities present in the lower 2/3 of the epithelium
- VIN3 abnormalities occupy the full thickness of the surface epithelium
- No invasion of the basement membrane

Risk Factors

- 1-Age: incidence increased with advanced age, although recently a trend in incidence was shown in young women below 35 years
- 2-HPV infection
- 3-Smoking
- 4-Low socioeconomic state
- 5-Young age at first birth
- 6-Chronic dermatoses …lichen sclorsus, sqamous hyperplasia
- 7-High coffee consumption and vitamin deficiency
- 8-DM, hypertension

- VIN comprises 2 entities:
- 1-one with risk factors similar to cancer cervix (HPV-related) called warty VIN
- 2-one with dissimilar risk factors (non-HPV-related) called keratinizing VIN
- The most common risk factors are *Age *HPV *smoking
- *Obesity is a risk factor because of difficulty in detection by patient and physician
- *Frequent pap smear reduce the risk because of frequent pelvic examination and early detection

Histologic Types of VIN

Based on morphological feature VIN3 lesions are subdivided into:

1-Warty lesion (condylomatous) lesion

2-Basaloid (usual type) lesion

3-Differentiated types (simplex type)

- **Warty type**: surface is spiked, large cells with nuclear pleomorphism, numerous mitosis, surface keratinocytes and koilocytosis
- **Basaloid type**: flat surface, immature parabasal cells, numerous mitosis, less koilocytes
- **Differentiated type**: presence of basal and parabasal cells, increased esinophilic cytoplasm, keratin pearl formation

Presentation Evaluation of VIN

- The most common clinical presentation is a patient who has been treated for CIN or VAIN
- *pruritus vulva lasting for more than 6 months or that not responding to treatment must be biopsed
- Condyloma should be biopsed to determine whether underlying neoplasm is also present
- Lesion may be white, velvety, erythematous, ulcerated or pigmented

- **Evaluation**
- Careful inspection
- Colposcopic examination with acetic acid (cells become white)
- Cervix should be evaluated by colposcope because the lesion is usually multifocal

Cancer Vulva

- Account for 3-4% of all gynecologic malignancies
- The average age at diagnosis is about 70 years, recently more cases are seen at the age of 40.
- Risk factors include chronic vulvar pruritus, HPV infection, lichen sclerosus, VIN, sqamous hyperplasia, history of sqamous cancer of cervix or vagina
- Pathology: 90% sqamous cell carcinoma, melanoma 5%

- **Spread**
- *Direct extension into adjacent structurs e.g urethera, bladder, vagina, perinum, anus, rectum
- *Lymphatic channels to inguinal LN (emboli) then to pelvic and para-aortic
- Hematogenous
- Classification based on TNM
- And stages are based on surgicopathological findings

Treatment of VIN

1-Wide local excision

*cases with suspected malignancy

*cases above 40 years

-if only VIN present ,this cures 75%

2-Laser evaporation

*young patient *no suspecious of malignancy

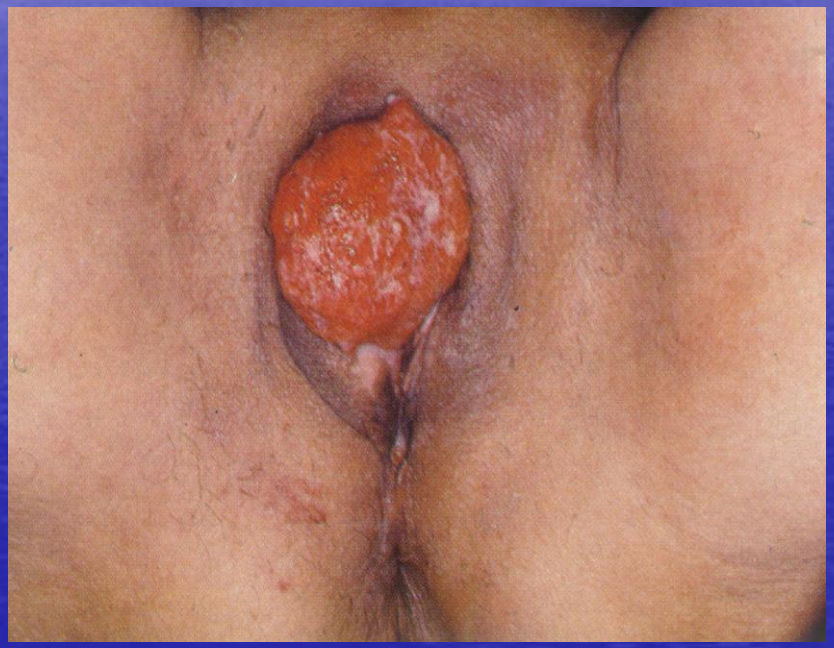
Recurrence in 10-20%

3-LEEp may be used but it leaves charred tissue

--New therapy: Retinyl acetate gel, aim at reversing the precancerous process

Clinical picture

- The most common complaint is a palpable vulvar lesion, the patient often has a long history of pruritus vulva
- About 20% of cases are asymptomatic and the lesion is identified during a routine pelvic examination
- Bleeding or watery discharge if the lesion become necrotic or ulcerated
- Diagnosis is often delayed because patients do not visit a physician because of embarrassment or fear



Diagnosis, prognosis

- Diagnosis
- Simple dermal punch biopsy using local anesthetic usually yield a definitive diagnosis
- Occasionaly wide local excision is necessary to differentiate a preinvasive from invasive carcinoma
- Subtle lesions may be delinated by staining the vulva with toludine blue or by using colposcpe
- DD:*veneral disease *basal cell carcinoma *VIN

  *Pget's disease *condyloma accuminata

- Prognosis is related to stage of the disease which is based on tumor size and location and regional lymph node status
- 5-year survival rates are >90% for stage I 80% for stage II, 50% for stage III, and 15% for stage IV
- Risk of lymph node spread is proportional to the tumor's and depth of invasion

Stages of vulvar cancer

- I:All lesions confined to vulva with a maximum diameter of <2cm and no suspicious groin nodes
- II:All lesions confined to the vulva with a diameter >2cm and no suspicious nodes
- III:Lesions extend beyond the vulva but without grossly positive groin nodes

-Lesions of any size confined to the vulva and having suspicious groin nodes

IV:Lesions with grossly positive groin nodes regardless of extent of primary Lesion involving mucosa of rectum, bladder, urthera or bone

-All cases with distant metastases or palpable deep pelvic metastases

Treatment

- Treatment of choice is radical excision of the local tumor and unilateral or bilateral inguinal lymphadenectomy
- *small lesions <2cm with invasion of <1mm, wide local excision can be used
- *Laterized lesion <2cm, unilateral hemivulvectomy and unilateral inguino-femoral lyphadenectomy
- *midline lesions requires bilateral node dissection
- *Lesion>2cm usually require a radical vulvectomy and bilateral inguino-femoral node dissection
- *For locally advanced tumor(III) bilateral node dissection followed by radiotherapy(+/-chemotherapy) followed by local radical excision

Pruritus Vulva

- Is the sensation of itching of the vulval area which usually occurs as an isolated phenomenon although may be associated with generalized itching
- The exact incidence and prevalence is not known, around 10% of women seen by Gynecologists experiience symptoms

Causes of Pruritus Vulva

1-Neoplastic conditions of vulva: sqaumous cell carcinoma, VIN, melanoma, bartholin's glsnd carcinoma

2-Non-neoplastic conditions:

- i-Lichen sclorosus ii-sqamous cell hyperplasia
- iii-Other dermatoses

3-Infections

- i-Yeast infection e.g candida vaginalis
- ii-Scabies
- iii-Pediculosis pubis
- iv-threadworm
- v-herpes simplex vi-vaginitis with discharge
- vii-urinary tract infection and vulvar vestibulitis

4-Other conditions

i-estrogen deficiency

Ii-GIT disease: prolonged contact of stool with vulva e.g fecal incontinence

Iii-Urinary incontinence

Iv-pregnancy through vulval engorgement

V-any cause of generalized itching : drug reactions and systemic disease.. lymphadenoma, jaundice, uraemia, anemia, psychological problems..etc

Treatment

- **Underlying cause should be treated**
- **Reassurance and tact to help reduce the possible embarrassment and stigma felt by the patient**
- **Non-drug treatment**
- ***Simple advice to avoid scratching, wear cotton underwear, clean affected area and avoid getting too hot**
- ***Avoidance of identified irritants: nylon underwear, tight fitting cloths, soaps, perfumes, over-zealous cleansing**

- Drug treatment
- **Topical steroid creams for primary idiopathic pruritus**
- **Systemic sedating antihistamines if there is difficulty in sleeping due to pruritus**