Diseases of the vulva & vagina

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Vulvar non-neoplastic epithelial disorders

- Lichen sclerosus
- Lichen simplex chronicus
Lichen sclerosus

• Thinning of the epidermis

• Disappearance of rete pegs

• Hydropic degeneration of the basal cells

• Dermal fibrosis

• Scant perivascular, mononuclear inflammatory cell infiltrate...mainly T cells
Lichen sclerosus, clinically:

• Smooth, white plaques... = leukoplakia
  or
• Papules...may extend and coalesce

• The labia may become atrophic and stiffened with vaginal orifice constriction

• In all age groups but most commonly affects postmenopausal women

• A small percentage of women (1% to 5%) with symptomatic lichen sclerosus develop squamous cell carcinoma of the vulva

• Increased frequency of autoimmune disorders
Lichen Simplex Chronicus

- Epithelial thickening (particularly of the stratum granulosum)
- Hyperkeratosis
- Increased mitotic activity is seen in the basal and suprabasal layers
- No epithelial atypia
- Leukocytic infiltration of the dermis is sometimes pronounced
Lichen Simplex Chronicus, cont’d

• Consequence of chronic irritation, often caused by pruritus

• Appears as an area of leukoplakia

• Lichen simplex chronicus often is present at the margins of established vulvar cancer
Vulvar neoplasms

• Benign...condyloma acuminata

• Malignant...squamous cell carcinoma
Condyloma acuminatum
Vulvar carcinoma

• 3% of female genital tract cancers

• Mostly >60 year old

• 90% are squamous cell carcinoma
  ...others: adenocarcinoma, basal cell carcinoma...etc.
Vulvar squamous cell carcinoma

• 2 forms:

1-Less common:

- In middle aged women
- Associated with HPV 16, 18...etc.
- Particularly in smokers, and immunodeficiency is also a risk factor
- Often preceded by precancerous lesions (VIN: vulvar intraepithelial neoplasia...I, II, & III)

...carcinoma occurs after many years and it is not inevitable
Vulvar squamous cell carcinoma, cont’d

2-The more common form:

- In older women
- Not associated with HPV 16, 18
- Preceded by years of lichen sclerosus or lichen simplex chronicus
- Not VIN I, II, or III but only a special lesion called: VIN simplex (subtle basal layer atypia & basal layer keratinization with good maturation)
- The invasive tumor is typically well-differentiated & keratinizing
Vulvar squamous cell carcinoma, morphology

**MORPHOLOGY**

VIN and early vulvar carcinomas manifest as areas of **leukoplakia** in the form of whitish patches of epithelial thickening. In about one fourth of the cases, the lesions are pigmented owing to the presence of melanin. Over time, these areas are transformed into overt **exophytic** or ulcerative **endophytic tumors**. HPV-positive tumors often are multifocal and warty and tend to be poorly differentiated **squamous cell carcinomas**, whereas HPV-negative tumors usually are unifocal and typically manifest as well-differentiated keratinizing squamous cell carcinomas.
Vulvar squamous cell carcinoma, clinical notes

• Both forms of vulvar carcinoma tend to remain confined to their site of origin for a few years but ultimately invade and spread, usually first to regional nodes

• The risk of metastasis correlates with the size of the tumor and the depth of invasion (stage)

• Women with tumors less than 2 cm in diameter have about a 90% 5-year survival rate after radical excision, whereas only 20% of those with advanced-stage lesions survive for 10 years
Extramammary Paget Disease

• Paget disease = intraepidermal proliferation of malignant epithelial cells that can occur in the skin of the vulva or nipple of the breast

• A majority of cases of vulvar (extramammary) Paget disease have no demonstrable underlying tumor...unlike that of the nipple

...Only occasionally, Paget disease in this location is accompanied by a subepithelial or submucosal tumor arising in an adnexal structure, typically sweat glands
Extramammary Paget Disease, clinically & histologically:

• Red, scaly, crusted plaque that may mimic the appearance of an inflammatory dermatitis

• Intraepidermal Paget disease may persist for years or even decades without evidence of invasion...invasion & metastasis is more if there is underlying skin adnexal malignancy and once metastasis occurs, the prognosis is poor

• Histologically: Large epithelioid cells with abundant pale, finely granular cytoplasm and occasional cytoplasmic vacuoles infiltrate the epidermis, singly and in groups...PAS differentiates it from melanoma (mucin)
Vaginal malignant neoplasms

• Squamous cell carcinoma
• Clear cell adenocarcinoma
• Sarcoma botryoides
Vaginal squamous cell carcinoma

• Extremely uncommon cancer

• Usually occurs in women older than 60 years of age

• In the setting of risk factors similar to those associated with carcinoma of the cervix

• Vaginal intraepithelial neoplasia is a precursor lesion that is nearly always associated with HPV infection

• Invasive squamous cell carcinoma of the vagina is associated with the presence of HPV DNA in more than half of the cases
Vaginal clear cell adenocarcinoma

• A very rare tumor

• Young women whose mothers took diethylstilbestrol during pregnancy...risk less than 1 per 1000, albeit about 40 times greater than in the unexposed population

• In about one third of exposed women, small glandular or microcystic inclusions appear in the vaginal mucosa
  ...these benign lesions are seen as red, granular-appearing foci that on histologic examination are lined by mucus-secreting or ciliated columnar cells... = vaginal adenosis
Vaginal Sarcoma Botryoides

= embryonal rhabdomyosarcoma

• A rare form of primary vaginal cancer that manifests as soft polypoid masses

• Usually is encountered in infants and children younger than 5 years of age

• Also may occur in other sites, such as the urinary bladder and bile ducts
Thank You