Vulval Diseases

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Vulval Diseases

- Introduction
- Benign vulval lesions
- Pre-invasive disease of the vulva
- Malignant vulval lesions
Introduction
The vulva

- Highly vascular
- Anatomy and embryological development help determine Dx & Rx
- Specific gynae, medical & dermatological disorders
- Rx: Requires knowledge in gynae, derma. and infectious diseases
- Care and compassion during consultation
Surface anatomy

- Mons pubis
- Glans of clitoris
- Openings of paraurethral (Skene) ducts
- Labium minus
- Labium majus
- Bartholin glands
- Anus
- Prepuce of clitoris
- Urethral opening
- Vestibule of vagina
- Vaginal opening
- Hymenal caruncle
- Hart’s line
Blood supply

The arterial supply:
- **Internal pudendal**: From Internal iliac arteries
- **External pudendal**: From the Femoral arteries

The venous drainage
- Via the labial veins to the internal pudendal veins
- These large plexuses become significantly enlarged in pregnancy
Nerve supply

Innervation of the vulva is via:

- Pudendal nerve (S2–4)
- Branches of the Ilioinguinal nerve (L1)
- Genital branch of the Genitofemoral nerve (L1–h2)
- Perineal branch of the Lateral Femoral cutaneous nerve of the thigh (L2–4)
Lymphatic drainage

- A meshwork of lymphatic vessels
- Lymph vessels pass laterally to lymph nodes in the groin, at the base of the femoral triangle
- Then drain to the deep nodes in the pelvis
- Then to the para-aortic nodes

Perineum and vulva has bilateral lymph drainage
Clinical approach

Specialised vulval clinic
Expertise in Derma, Gynae, infectious diseases

Approach
- History
- Clinical examination
- Investigation
  - Microbiological swabs
  - Vulval biopsy if necessary
History

- Duration, appearance and change, Previous Rx
- Symptoms: Itching, burning, pain
- Affected family members
- Lesions elsewhere on the skin or oral/vaginal mucosa
- Is anything new or different: self care products, new sexual partner….
- Factors that improve or exacerbate the condition
- Vaginal discharge or history of STI
Clinical examination

Characteristics of the vulvar lesion should be evaluated:

• Morphology (macule, papule, patch, nodule, ulcer)
• Size and shape
• Number and distribution
• Color
• Consistency
• Secondary changes (excoriation, lichenification, scale, fissure, erosion, bleeding)
• Non-vulvar skin: vaginal, cx, ocular, oral, nasal, anal
• Regional lymph nodes
Investigations

• Dx  often made on basis of hx & exam
• Biopsy may be needed to confirm Dx.
  o  Difficultly in establishing a Dx
  o  Pigmented lesions
  o  Persistently eroded areas
  o  Indurated areas
  o  No response to Rx following initial diagnosis
• Culture swabs
Benign lesions of the vulva
Bartholin’s gland cyst
Bartholin gland

- The greater vestibular glands
- A pair of 0.5 cm glands located at 4 o'clock and 8 o'clock positions of the introitus
- A mucus-secreting gland
- Plays a role in vaginal lubrication
- Generally nonpalpable when not obstructed
Bartholin’s gland cyst

• A benign swelling of the Bartholin’s gland
• Usually unilateral, asymptomatic, and may be incidental finding
• Predominantly in women of child-bearing age
  o Often found after onset of puberty
  o Decrease in incidence after menopause
• Obstruction may occur after trauma to the area, episiotomy, or childbirth or without an known cause
• Cysts and abscesses account for 2% of all annual gynae consultations
Bartholin’s gland cyst

Presentation

- **Cyst:** Painless
- **Abscess:** Painful

Treatment:

- **Asymptomatic:** ? No intervention
- **Abscess:** Marsabialisation

Risk of recurrence
Lichen sclerosus

- Presents as well-demarcated white, finely wrinkled, and atrophic patches
- Normal genital landmarks may be obliterated with chronic inflammation and adhesions
- Consider biopsy if suspicious lesion
- The risk of malignancy 4 - 5 %

Rx:
- Medical: potent topical corticosteroid ointment (clobetasol 0.05% ointment)
- Follow up closely; risk of ca
Lichen sclerosus
Pre-invasive disease of the Vulva
Vulval intraepithelial neoplasia (VIN)
VIN

• Benign changes that can occur in the vulval skin
• Can resolve without treatment
• May progress to ca
Clinical manifestations of VIN

Symptoms
- Vuval pruritus, pain, burning, dyspareunia
- 50% of cases are asymptomatic
- Represent “a field change”, therefore, cervix and perianal area must be examined to exclude CIN and anal neoplasia
- 24% of women with VIN have CIN
- Progression to cancer: 2–14% of patients

Examination
- Lesions (may be raised, erythematous, leukoplakic, keratotic, ulcerated or pigmented)

Vulvoscopy
- Acetowhite change
Classification of VIN

Differentiated VIN
- Unifocal
- High risk of developing squamous cell carcinoma (SCC)
- Postmenopausal women
- Associated with lichen sclerosis
- Have a non-viral etiology
- Not classically associated with CIN

Usual type
- Multifocal
- Low risk of developing SCC
- Premenopausal women
- Associated with HPV, smoking and immunodeficiency
- May have similar pathophysiology to CIN
VIN; Investigation

Investigation
- Vulvoscopy
- Biopsy
VIN: Treatment

Options include

• Wide local excision
• Skinning vulvectomy
• Laser ablation
• Topical treatment (e.g. imiquimod)
Vulval cancer
Vulvar cancer

- Fourth most common gynecologic ca
- 5% of ca of female genital tract
- Most frequently in postmenopausal women
- Mean age at diagnosis: 65 yrs
- Early detection and Rx of VIN may prevent ca

Risk factors

- Cigarette smoking
- Vulval dystrophy (eg, lichen sclerosus)
- Vulval or cervical intraepithelial neoplasia
- HPV infection
- Immunodeficiency syndromes
- Prior history of cervical cancer
- Northern Europeans
Vulval carcinogenesis

Related to HPV infection

- HPV responsible for 60% of vulvar ca
- HPV 16 + 33 in 55% of cases

Related to chronic inflammatory (vulvar dystrophy) or autoimmune processes
Clinical presentation

Common symptoms

- Pruritus (most common; 38–71%)
- Bleeding
- Swelling
- Ulceration
- Pain or burning
- Discharge
- Asymptomatic (up to 5% detected histologically in association with VIN or ca of cervix or anus)
Pathology

- Squamous carcinomas: 90%
- Malignant melanoma: 5%
- Basal cell carcinoma: 2%
- Bartholin's gland ca: <1%
- Adenocarcinoma: <1%
- Verrucous carcinoma: <1%
- Sarcomas: 1%
Clinical presentation

• Unifocal vulvar plaque, ulcer, or mass (fleshy, nodular, or warty)
• Typical appearance: raised ulcer with rolled edges
• Most common site: labia majora
• Less frequent: Labia minora, perineum, clitoris, mons
• Multifocal in 5%; therefore, all vulval and perianal skin surfaces, the cervix and vagina, should be examined
• A synchronous second ca, most commonly cervical: in up to 22% of cases
Histologic types

Squamous cell carcinoma

Over 90 % of vulval ca

Two subtypes

The keratinizing type (differentiated, or simplex)
- More common
- Older women
- Not related to HPV infection
- Associated with dystrophies (lichen sclerosus)

The classic type (warty)
- Predominantly associated with HPV 16, 18, 33
- Younger women
- Present with early stage disease
Histologic types

Melanoma

- Second most common
- 5%
- Postmenopausal (median age: 68 years)
- Usually pigmented lesion, but amelanotic lesions occur
- Most common sites: Clitoris, labia minora
Histologic types

Verrucous carcinoma

• Variant of squamous cell ca
• <1% of ca vulva
• Cauliflower-like in appearance
• Differentiated from squamous cell ca with a verrucous configuration by biopsy of the lesion base
• Grows slowly
• Rarely metastasizes to lymph nodes
• May be locally destructive
Histologic types

Basal cell carcinoma

- 2% of vulval ca

Sarcoma

- Soft tissue sarcomas (including leiomyosarcomas, rhabdo., lipo., angio., neurofibrosarcomas, ...)
- 1 - 2 % of vulval ca
- Generally poor prognosis
Histologic types
Extramammary Paget’s disease

- An intraepithelial adenocarcinoma
- < 1% of vulval ca
- Age: 60s and 70s and Caucasian
- Most common symptom: pruritus
- Lesion: eczematoid appearance; well-demarcated, slightly raised edges and a red background,
- Usually multifocal
- Sites: vulva, mons, perineum/perianal area, inner thigh
- Diagnosis: often delayed
- Biopsy should be performed if dermatitis does not respond to Rx
Mode of spread

Direct extension
• To adjacent structures (vagina, urethra, clitoris, anus)

Lymphatics
• To regional lymph nodes
• May occur early in the course of disease
• First to groin (inguinal-femoral) lymph nodes
• Ipsilateral disease generally spread to ipsilateral nodes

Hematogenous
• Typically occurs late in the course of the disease
• Rare in patients without lymph node involvement
Evaluation prior to Rx

Clinical evaluation
• Guide surgical and medical approach (eg, choice of incision, use of neoadjuvant chemo-radiation)

Pelvic and general physical examination
• Measure diameter of primary tumor
• Palpate inguinal, axillary, supraclavicular lymph nodes

Cervical cytology and colposcopy of the cervix, vagina, vulva
• Squamous intraepithelial lesions are multifocal

Abdominal/pelvic computed tomography
• Detect lymphadenopathy and metastases
• Additional imaging studies performed as appropriate
Staging of vulval ca

• Staging is surgical / pathological
• Considers most important factors related to prognosis:
  o Tumor size
  o Depth of invasion
  o Lymph node involvement
  o Presence of distant metastases
• Essential because inguino-femoral LN status is the most important predictor of overall prognosis
• Staging and surgical Rx performed as a single procedure
Stages of ca vulva

**IA**  Tumor confined to the vulva or perineum, ≤ 2cm in size with stromal invasion ≤ 1mm, negative nodes

**IB**  Tumor confined to the vulva or perineum, > 2cm in size or with stromal invasion > 1mm, negative nodes

**II**  Tumor of any size with adjacent spread (1/3 lower urethra, 1/3 lower vagina, anus), negative nodes

**IIIA**  Tumor of any size with positive inguino-femoral lymph nodes
(i) 1 lymph node metastasis greater than or equal to 5 mm
(ii) 1-2 lymph node metastasis(es) of less than 5 mm

**IIIB**  (i) 2 or more lymph nodes metastases greater than or equal to 5 mm
(ii) 3 or more lymph nodes metastases less than 5 mm

**IIIC**  Positive node(s) with extracapsular spread

**IVA**  (i) Tumor invades other regional structures (2/3 upper urethra, 2/3 upper vagina), bladder mucosa, rectal mucosa, or fixed to pelvic bone
(ii) Fixed or ulcerated inguino-femoral lymph nodes

**IVB**  Any distant metastasis including pelvic lymph nodes
Surgical Management

Radical vulvectomy (RV)
- Historically, vulvar cancer staged and Rx with radical vulvectomy and inguinofemoral lymph node dissection
- Associated with high rates of survival as well as morbidity

Wide local excision
- An alternative surgical approach
- Limited excision with same depth of dissection as RV
- Most commonly performed

For all surgical Rx:
- Tumor-free margin of at least a 1 cm decreases risk of local recurrence
Examples of incisions used for radical vulvectomy

(A) Radical vulvectomy via butterfly incision. (B) Modified radical vulvectomy: triple incision technique; a skin bridge is left between the radical vulvectomy and the groin incisions. (C) Modified radical vulvectomy: anterior horseshoe incision.
Inguinofemoral lymphadenectomy

- Superficial inguinal and deep femoral LN
- For all stages of disease except stage IA
- Pelvic LN (obturator and iliac nodes) not required for staging or Rx
- High morbidity
  - Wound infection and breakdown (20 - 40 %)
  - Lower extremity lymphedema (30 - 70 %)
Chemoradiation

Indications for primary radiotherapy

• Locally advanced (stage III to IVA)
• Inoperable disease

Postoperative RT

• Advanced stage surgically treated disease with high-risk of local recurrence
• Adjuvant pelvic RT including the groin for patients with two or more positive inguinal LN
Presence of +ve inguinofemoral nodes is the most important prognostic factor for survival in patients with vulval ca

## Prognosis (FIGO)

<table>
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<th>FIGO stage</th>
<th>Number of patients</th>
<th>Overall survival, percent</th>
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<tr>
<td></td>
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<td>1 year</td>
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<tr>
<td>I</td>
<td>286</td>
<td>96.4</td>
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<tr>
<td>II</td>
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<td>III</td>
<td>216</td>
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<td>IV</td>
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Post Rx follow up

Clinical evaluation
• Symptoms
• Physical examination of vulva, skin bridge, and inguinal LN

Intervals of follow up
Low-risk disease
• Early stage, treated with surgery alone, no adjuvant therapy
• For the first two years, every six months; and then annually

For high-risk disease
• Advanced stage, treated with primary chemotherapy/radiation therapy or surgery plus adjuvant therapy
• For the first two years, every three months
• For year 3 through 5, every six months, and then annually