Vulval Diseases

Dr Khaldoun Khamaisheh MRCP FRCOG
Consultant Obstetrician & Gynaecologist
Vulval Diseases

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

- Highly vascular
- Specific gynae, medical & dermatological disorders
- Rx: Requires knowledge in gynae, derma. and infectious diseases

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
Surface anatomy

- Mons pubis
- Glans of clitoris
- Openings of paraurethral (Skene) ducts
- Labium minus
- Labium majus
- Bartholin glands
- Anus
- Prepucce of clitoris
- Urethral opening
- Vestibule of vagina
- Vaginal opening
- Hymenal caruncle
- Hart’s line
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, L2)
- 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 may include
  o Microbiological swabs
  o Vulval biopsy
  o Vulvoscopy
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 vulval lesion should be evaluated:

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

• Biopsy
  o Difficultly in establishing a Dx
  o Pigmented lesions
  o Erosions
  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 and 8 o'clock positions of the introitus
• A mucus-secreting gland
• Plays a role in vaginal lubrication
• Generally non palpable when not obstructed
Bartholin’s gland cyst

• A benign: 2% of all annual gynae consultations
• Usually unilateral, asymptomatic
• May be an incidental finding
• Predominantly in women of child-bearing age
  o Decrease in incidence after menopause
• Obstruction may occur after trauma such as episiotomy, childbirth or without a known cause
• Consider biopsy after age of 40 (risk of ca)
Bartholin’s gland cyst

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

**Treatment:**
- Asymptomatic: ? No intervention
- Abscess: Marsupialisation
- Recurrent Cyst: Consider excision
  - Risk of cancer
Lichen sclerosus

- Presents as well-demarcated white, atrophic patches
- Consider biopsy if suspicious lesion
- The risk of malignancy: 4 - 5%

**Treatment**

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

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

Vulvoscopy
- Acetowhite change
Classification of VIN

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

**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
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 ca
- 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)
- VIN and CIN (HPV in 60%: type 16 & 33)
- Immunodeficiency syndromes
- Prior history of cervical cancer
Clinical presentation

• Pruritus (most common; 38–71%)
• Bleeding
• Swelling
• Ulceration
• Pain or burning
• Discharge
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
Pathology

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

The keratinizing type (differentiated)

- More common
  - Older women
  - Not related to HPV infection
  - Associated with dystrophy (lichen sclerosus)

The classic type (warty)

- Predominantly associated with HPV 16, 18, 33
- Younger women
- Present with early stage disease
Histologic types

Melanoma

• 5%
• Postmenopausal (median age: 68 years)
• Usually pigmented lesion, but amelanotic lesions occur
• Most common sites: Clitoris, labia minora
Mode of spread

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

Lymphatics
- To regional lymph nodes
- First to groin (inguinal-femoral lymph nodes)

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

Clinical evaluation
• Guides 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
  • Considered the most important factor re prognosis:
  • Involves
    o Tumor size
    o Depth of invasion
    o Lymph node involvement
    o Presence of distant metastases

• Inguino-femoral LN status is the most important predictor of overall prognosis
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
Staging

- **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, 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

Complications
- Wound infection and breakdown (20 - 40 %)
- Lower extremity lymphedema (30 - 70 %)
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.
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)**

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<th>FIGO stage</th>
<th>Number of patients</th>
<th>Overall survival, percent</th>
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<td></td>
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<tr>
<td>I</td>
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Thank you