Vulvar Cancer

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Faculty/Presenter Disclosure

• **Faculty:** Alon Altman

• **Relationships with commercial interests:**
  – **Grants/Research Support:** Cancercare Manitoba foundation operational research grant.
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  – **Other:** Employee WRHA, Cancercare Manitoba and University of Manitoba; Site PI for ARRAY MEK Inhibitor study
Mitigating Potential Bias

No bias identified
Objectives

- Vulvar cancer basics
  - Epidemiology
  - Risk factors
  - Types
- How to diagnosis the disease?
- What is the appropriate work up?
- How is this disease treated?
- What complications can I expect to see in my office?
Basics

EPIDEMIOLOGY

- 3-5% of malignancies of the female tract
- 4\textsuperscript{th} most common gynecologic malignancy
- Disease of post-menopausal women
- Average age at diagnosis 65 years
  - 15\% of patients <40 years
Risk Factors:

- HPV (16, 18, 31, 33, 35) (20-60%)
- Granulomatous disease
- Diabetes, HTN
- Smoking
- Obesity
- SCC of Cx or CIN
- Age
- Multiple sexual partners
- Immuno-compromised /HIV
- Vulvar dystrophy
- VIN (associated with vulvar cancer)
- Radiation
## TYPES OF VULVAR ATYPIA

<table>
<thead>
<tr>
<th>Type</th>
<th>Details</th>
</tr>
</thead>
<tbody>
<tr>
<td>Squamous cell hyperplasia</td>
<td></td>
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<tr>
<td>Lichen sclerosus</td>
<td>4-5% risk of malignancy</td>
</tr>
<tr>
<td>VIN 1-3</td>
<td></td>
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<tr>
<td>Paget’s disease</td>
<td>10-12% with invasive component</td>
</tr>
<tr>
<td>Melanoma in situ</td>
<td></td>
</tr>
<tr>
<td>Aggressive Angiomyxoma</td>
<td></td>
</tr>
<tr>
<td>Lichen Planus</td>
<td></td>
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</table>
# TYPES OF VULVAR CANCER

<table>
<thead>
<tr>
<th>Type</th>
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<tbody>
<tr>
<td>Squamous cell carcinoma → 90% of cancers</td>
</tr>
<tr>
<td>Melanoma → 2(^{nd}) most common cancer</td>
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<tr>
<td>Basal cell carcinoma</td>
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<tr>
<td>Bartholin gland adenocarcinoma</td>
</tr>
<tr>
<td>Metastatic disease</td>
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<tr>
<td>Verrucous carcinoma</td>
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<tr>
<td>Sarcoma</td>
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</table>
Signs and Symptoms

- **Irritation or itching is most common**
- Dyspareunia or lump/mass
- Whitish change, thickened keratinized layer
- Thin skin with scarring or contractures
- Fissuring of skin
- May also be red or pigmented
- Discrete or multifocal
- Bleeding, discharge, dysuria
Diagnosis

• “If cancer’s the answer, and tumour’s the rumour, then tissue’s the issue”
  
  Dr. James Bentley

• i.e. Biopsy, Biopsy, Biopsy
Diagnosis:

- Need biopsy (Keyes punch biopsy or tischler)
- Monsel’s solution, AgNO₃, or suture for hemostasis

Colposcopy:

- May use 3% acetic acid to highlight affected areas
- No specific appearance is characteristic in vulvar lesions
Key points

The biopsy specimen must include some underlying dermis and connective tissue so that the pathologist can adequately evaluate the depth and nature of the stromal invasion.

It is preferable to leave the primary lesion in situ, to allow the treating surgeon to fashion adequate surgical margins.
Work up

- Colposcopy + directed biopsies
- Pap smear
- LN exam
- Pelvic/pelvirectal exam
Spread

• Direct
• Lymphatic
• Hematogenous
Important points to consider

- 18-22% of VIN III had invasive process
- Higher recurrence with positive margins
- Conversion of VIN III $\rightarrow$ invasive cancer = 3.4% (up to 5%)
- No treatment needed for asymptomatic HPV infections
So, how is it treated?
Treatments:

- VIN 3:
  - Wide local excision
  - Laser
  - Aldara/Imiquimod

- Paget’s
  - WLE/Radical local excision
  - Mammogram, colonoscopy
Treatments:

- Radical local excision
- +/- lymph nodes
- Radiation therapy with concurrent cisplatin
- Chemotherapy
# Staging

## Carcinoma of the vulva.

<table>
<thead>
<tr>
<th>Stage</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Tumor confined to the vulva</td>
</tr>
<tr>
<td>IA</td>
<td>Lesions $\leq 2$ cm in size, confined to the vulva or perineum and with stromal invasion $\leq 1.0$ mm*, no nodal metastasis</td>
</tr>
<tr>
<td>IB</td>
<td>Lesions $&gt;2$ cm in size or with stromal invasion $&gt;1.0$ mm*, confined to the vulva or perineum, with negative nodes</td>
</tr>
<tr>
<td>II</td>
<td>Tumor of any size with extension to adjacent perineal structures (1/3 lower urethra, 1/3 lower vagina, anus) with negative nodes</td>
</tr>
<tr>
<td>III</td>
<td>Tumor of any size with or without extension to adjacent perineal structures (1/3 lower urethra, 1/3 lower vagina, anus) with positive inguino-femoral lymph nodes</td>
</tr>
<tr>
<td>IIIA</td>
<td>(i) With 1 lymph node metastasis ($\geq 5$ mm), or (ii) 1–2 lymph node metastasis(es) (&lt;5 mm)</td>
</tr>
<tr>
<td>IIIB</td>
<td>(i) With 2 or more lymph node metastases ($\geq 5$ mm), or (ii) 3 or more lymph node metastases (&lt;5 mm)</td>
</tr>
<tr>
<td>IIIC</td>
<td>With positive nodes with extracapsular spread</td>
</tr>
<tr>
<td>IV</td>
<td>Tumor invades other regional (2/3 upper urethra, 2/3 upper vagina), or distant structures</td>
</tr>
<tr>
<td>IVA</td>
<td>Tumor invades any of the following: (i) upper urethral and/or vaginal mucosa, bladder mucosa, rectal mucosa, or fixed to pelvic bone, or (ii) fixed or ulcerated inguino-femoral lymph nodes</td>
</tr>
<tr>
<td>IVB</td>
<td>Any distant metastasis including pelvic lymph nodes</td>
</tr>
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*The depth of invasion is defined as the measurement of the tumor from the epithelial-stromal junction of the adjacent most superficial dermal papilla to the deepest point of invasion.*
Prognosis and survival

- LN status single most important prognostic factor

<table>
<thead>
<tr>
<th>LN status</th>
<th>Survival (5 yr)</th>
</tr>
</thead>
<tbody>
<tr>
<td>No LN mets</td>
<td>&gt;90%</td>
</tr>
<tr>
<td>LN mets</td>
<td>50%</td>
</tr>
<tr>
<td>1x microscopic LN</td>
<td>~90%</td>
</tr>
<tr>
<td>3+ LN</td>
<td>20-50%</td>
</tr>
<tr>
<td>+ve Pelvic LN</td>
<td>10%</td>
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</table>
Melanoma

- 4-5% of primary vulvar ca
- 2nd most common
- Asymptomatic

**Prognosis:**
- **Thickness**
- Number of regional lymph nodes
- **Ulceration**/bleeding at primary site
- Size
What am I going to see in my office?
Complications of surgery and radiation

- **Early**
  - Wound infection
  - Lymphocyst formation
  - Wound breakdown (incidence 44% with separate incisions, major in 14%)
Complications of Surgery and Radiation

- *Late*
  - Chronic lymphedema (30% patients)
  - Chronic cellulitis (10%)
  - Urinary incontinence (10%)
  - Dyspareunia
  - Depression, Altered body image and sexual dysfunction
Recurrence

- Occurs in 1/3 of patients

- Surgery (exenteration)
- Radiation
- Chemotherapy
Take home messages

- Visual assessment alone is often inadequate
- Biopsy anything unusual
- Refer to colposcopy for assessment