Vulvar Cancer?

Vulvar Cancer

- 4th most common GYN cancer
  - 5% of malignancies of GYN type.
  - 4850 new cases annually
  - 1030 deaths

Risk Factors

- Cigarette smoking
- Vulvar dystrophy (Lichen Sclerosis)
- VIN / CIN
- HPV (40% of invasive lesions)
  - HPV 16 (85% of cases)
- Immunodeficiency syndromes
- Prior history of cervical cancer
### Etiology By Age Group

- **Younger patients**
  - Related to HPV infection and smoking
  - Commonly associated with Basaloid or warty VIN
- **Elderly (>65 y/o)**
  - High incidence of dystrophic lesions
  - More common with lichen sclerosis present

### Symptoms

- Pruritus -- #1
- Pain
- Vaginal Bleeding / Discharge
- Dysuria
- Unusual or foul odor
- Vulvar lump, plaque, or ulcer
- Groin mass

### Diagnosis

- Visual inspection
  - During routine annual exam or based on patient complaints
- Vulvoscopy
  - If no abnormal lesion seen with the naked eye but suspicion of underlying lesion
- Complete Exam
  - Examine entire Vagina and Cervix when suspected vulvar carcinoma because neoplasia most often is multi-focal
### Typical Lesions

- Raised
- Fleshy
- Ulcerated
- Leukoplakic
- Warty
- Grey – white area
- Nodular
- Most are single / solitary lesions

### Vulvoscopy

- Apply 5% acetic acid to vulva and look for acetowhite lesions or vascular changes
- Any area of concern should be biopsied
- If multiple areas → multiple biopsies

### Biopsy

- Keyes Biopsy
  - Important to get underlying dermis so pathologist can evaluate presence and depth of stromal invasion
  - Biopsy most abnormal area of vulvar lesion
Keyes Biopsy

Histology

- Squamous Cell
- Melanoma
- Basal Cell
- Sarcoma
- Extramammary Paget’s Disease
- Bartholin gland adenocarcinoma

Squamous Cell Carcinoma

- >90% of vulvar carcinomas
- Keratinizing or simplex type
  - Seen in older patients and associated with lichen sclerosus
- Warty or Bowenoid type
  - Younger women and associated with HPV 16, 18, and 33
- Verrucous
  - Variant of squamous cell
  - Cauliflower-like appearance
Melanoma

- Second most common type of vulvar carcinoma
- 5-10% of cases
- Common in postmenopausal women
  - Caucasian, non-hispanic
  - ~68 y/o
- Presents as a pigmented lesion
- Commonly seen in labia minora or clitoris
- Asymptomatic
- Treatment of choice radical vulvectomy with inguinal LN if >1 mm deep vulvar lesion

Melanoma Staging

- Uses depth of lesion and nodes for staging
- Commonly use Clarks staging
- New staging by AJCC that uses clarks with depth of lesion

Basal Cell

- 2% of vulvar carcinomas
- Present as
  - Ulcer "rodent" type
    - Rolled edges and central ulceration
    - Pigmented or pearly and gray
  - Pruritus and bleeding can be seen
  - Most commonly asymptomatic
Sarcoma

- 1-2% of vulvar carcinoma
- Leiomyosarcoma, liposarcoma, angiosarcoma, rhabdomyosarcoma, neurofibrosarcoma, epithelioid sarcoma

Extramammary Paget’s Disease

- Intraepithelial adenocarcinoma
- <1% of vulvar carcinomas
- Presents as
  - Pruritus (70%)
  - Eczematoid appearance
  - Slightly raised edges, red background, dotted with small pale islands
  - Usually multi-focal (examine entire vulva, perianal area, inner thighs)

Bartholin Gland Carcinoma

- Usually Solid and infiltrating tumors
- Biopsy any Bartholin gland in patients >40 years old to exclude carcinoma commonly adenocarcinoma
  - Even in cystic or abscessed glands
- Surgery – radical vulvectomy withinguinal lymphadenectomy
Route of Spread

- Direct
  - Adjacent structures – Vagina, urethra, anus
- Lymphatic
  - To regional lymph nodes (Inguinal first)
    - Superficial groin then femoral lymph node spread
- Hematogenous
  - To distant sites: Lung, liver, bone
  - Usually see lymph node involvement first

Staging

- Vulvar carcinoma is a surgically staged disease
- Simple vulvectomy vs. Radical vulvectomy with or without groin lymphadenectomy

FIGO Staging

- IA – Lesions ≤ 2 cm confined to vulva / perineum, Invasion ≤ 1mm, Neg nodes
- IB – Lesion >2 cm confined to vulva / perineum or invasion >1mm, Neg Nodes
- II – Any size with extension to adjacent structures (Lower 1/3 vagina or urethra, anus)
FIGO Staging

- IIIA – 1 LN met (≥5mm) or 1-2 LN met (<5mm)
- IIIB – ≥2 LN met (≥5mm) or ≥3 LN met (<5mm)
- IIIC – Positive LN with extracapsular spread

- IVA – Tumor invades upper urethra, vaginal mucosa, bladder mucosa, rectal mucosa or fixed to pelvic bone OR fixed or ulcerated inguino-femoral LN
- IVB – Any distant met (includes pelvic LN)

Surgery

- Wide local excision of lesion if < 1mm depth of invasion
- Radical local excision / Radical vulvectomy if > 1mm depth of invasion
- At least 1 cm margin of normal tissue (@ least 8 mm on final pathology)
- Lymph nodes only if >1mm depth of invasion
  - Ipsilateral LN in unilateral tumor
  - Bilateral LN if bilateral tumor or midline tumor or positive ipsilateral LN’s

Vulvar Cancer
**Post-operative**

![Image of post-operative incision]

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**Inguinal lymphadenectomy complications**

- **Early**
  - Lymphocyst
  - 40% of cases
  - Cellulitis, UTI, DVT, PE, MI, hemorrhage, osteitis pubis
- **Late**
  - Chronic leg edema
    - Can be seen up to 60%
  - Urinary stress incontinence / genital prolapse
  - Introital stenosis, femoral hernia
  - RV fistula
Sentinel Lymph node mapping

- Inject into the vulvar lesion TC 99 +/- isosulfan blue dye
- Dissect groin in usual fashion to find the sentinel node
- Can find with a gamma probe or visualization of the blue dye
- Frozen section of the node to determine if further nodes need to be removed
- If sentinel is negative then no further dissection needed
- False negative rate of this mapping is 5%

Sentinel lymph node

Positive LN.. Now What

- The determination of RT after surgery depends on how many lymph nodes are involved and the size
- If 1 micro met (≤ 5 mm) then observe
- If 3+ micro mets or 1 macro met (>5 mm) or extracapsular spread then groin and pelvic RT
- If 2 micro mets then observe or RT
**Advanced Vulvar cancer - unresectable**

- CT of abdomen, pelvis to look at pelvic and groin LN's
- If no suspicious nodes then proceed with bilateral inguinal lymphadenectomy and if the nodes are negative then use RT to the vulvar lesion and omit the groin RT
- If suspicious but resectable nodes
  - Remove all enlarged groin nodes, can remove pelvic nodes by extraperitoneal approach, give full pelvic and groin RT when healed
- If fixed and unresectable nodes then proceed with primary RT to groin and vulvar lesion

**Vulvar Cancer - Surgery**

- If primary tumor can be removed then proceed with radical vulvectomy with inguinal lymphadenectomy
- If tumor involves anus, rectum, RV septum, proximal urethra
  - Can opt for primary exenteration
  - Can proceed with Radiation therapy then surgery to follow if residual tumor is seen

**Who else receives RT**

- For groin nodes + that were prior explained
- Those patients with a positive vulvar margin
- Those patients with a close margin(<8mm)
### Recurrence
- Local recurrence at site distant (>2cm) from the primary tumor
  - Resect or RT
  - Good prognosis (66% 3 year survival)
- Local recurrence at primary tumor site
  - Poor prognosis (15.4% survival at 3 years)
- Distant recurrence
  - Consider Chemo

### Prognosis
- 5 year survival
- Stage I – 90%
- Stage II – 77%
- Stage III – 51%
- Stage IV – 18%

### Vulvar Cancer Pearls
- Younger patient often HPV related
- Older patient often related to VIN
- Squamous cell is most common
- Commonly present with a lump, mass, bleeding, itching, dysuria, discharge
- Diagnosis by keyes biopsy
- Spread by direct extension, lymphatic, and hematogenous spread
### Vulvar Cancer Surgery Pearls

- Stage IA – radical vulvectomy, no groin nodes needed
- Stage IB – IV = Radical vulvectomy, + groin LN
- If lesion single sided then can do unilateral LN
- If lesion is central or on both sides then need to do bilateral LN
- Need at least an 8 mm negative margin on final path

### Vulvar Cancer Treatment Pearls

- RT preop if lesion is on clitoris of younger patient and resection would not get optimal margin
- RT preop in patient with advanced disease who would otherwise need a pelvic exenteration
- Postop RT treatment
  - To prevent recurrence in patients with an involved or close margin (<8 mm)
  - In positive nodes
    - > 2 micromets, 1 macromet or extracapsular spread

### Vaginal Cancer
Vaginal cancer

- 1-2% of malignant tumors of female genital tract
- >50% of patients diagnosed in 70s, 80s, 90s y/o
- Squamous cell is most common path
- Usually a vaginal lesion is a met from somewhere else
  - Cervix 32%
  - Endometrium 18%
  - Colon and rectum 9%
  - Ovary 6%
  - Vulva 6%

Pathology

- Squamous cell
- Adenocarcinoma
- Melanoma
- Sarcoma
- Undifferentiated
- Small cell
- Lymphoma
- Carcinoid

Symptoms

- Painless vaginal bleeding or discharge
- Usually postmenopausal bleeding
- Can be postcoital bleeding
- Bladder pain and frequency
- Pelvic pain
- Rectal pain
Diagnosis

- PE
  - Most lesions are in the upper one-third of vagina
  - Apex or posterior wall
  - Biopsy of lesion for diagnosis
  - OR to biopsy or remove lesion if needed

FIGO staging

- Stage I – limited to vaginal wall
- Stage II – involved the subvaginal tissue but not extended to pelvic wall
- Stage III – Extension to the pelvic wall
- Stage IV A – Invades bladder or rectal mucosa and or spread beyond the true pelvis
- Stage IV B – spread to distant organs

Staging

- Clinically staged
  - Physical exam
  - Cystoscopy
  - Proctosigmoidoscopy
  - Chest x-ray
  - Skeletal x rays
Spread patterns

- Direct
  - To pelvic soft tissues and adjacent organs
- Lymphatic
  - Pelvic and then PA lymph nodes
  - Distal 1/3 vaginal lesions can met directly to groin LN
  - Posterior lesions can involve perirectal nodes
- Heme
  - Distant organs
  - Lungs, liver, bone

Treatment

- Surgery
  - Stage I with tumor in upper posterior vagina
    - Radical hysterectomy BSO and pelvic LN
    - Radical upper vaginectomy and pelvic LN
  - Stage IVA
    - Esp. if RV or VV fistula
    - Primary pelvic exenteration is suitable option
  - Central recurrence after RT
    - Pelvic Exenteration

Radiation

- All other patients that can't have surgery
Prognosis

- Stage 1 – 77%
- Stage II – 52%
- Stage III – 42%
- Stage IVA – 21%, Stage IVB – 12%
- Most recurrences are in the pelvis
- Survival after recurrence is 12%

Adenocarcinoma of vagina

- 10% of cases
- Younger patients
- Look for a met site first (colon, endometrium, ovary)
- Associated with patients who had DES exposure in utero
- Treatment is the same as squamous cell
- Better survival and prognosis of those associated with DES exposure vs. those who are not

Verrucous Carcinoma

- Rare
- Large, warty tumors
- Aggressive, but less tendency to met
- Wide surgical excision is treatment of choice
  - LN is not necessary
- RT can cause them to be more aggressive type of tumors
**Rhabdomyosarcoma**

- Malignant tumor
- Most common soft tissue tumor in children
- Rhabdomyosarcoma is most curable form of the disease
  - But only account for 4% of pediatric rhabdomyosarcomas
- Sarcoma botryoides
  - Embryonal Rhabdomyosarcoma
  - Highly malignant, grapelike tumor
  - In the vagina during infancy or childhood, cervix in reproductive years, corpus uteri in postmenopausal years
- Treatment
  - Chemotherapy has replaced radical surgery (except)

**Rhabdomyosarcoma Treatment**

- Chemotherapy
  - VAC (vincristine, actinomycin D, cyclophosphamide)
- RT or surgery if residual disease

**Vaginal Cancer Pearls**

- Squamous cell is most common path
- Diagnosis of exclusion after you have rule out other met source
- Most commonly present with painless vaginal bleeding and discharge
- Diagnosed by biopsy, but rule out met uterine, cervix, colon, rectum, ovary and vulva
- Spread through direct extension, lymphatic and hematogenous
Vaginal cancer Treatment Pearls

- Stage I disease
  - Can do radical hyst with upper vaginectomy and pelvic LN
- Stage II – IV
  - Radiation therapy ± weekly cisplatin chemo
  - If lower 1/3 of vagina involved then groin nodes should be dissected out or treated with RT
- Stage IV
  - If rectovaginal or vesicovaginal fistula then can do primary pelvic extenteration