AMSER Rad-Path Case of the Month:

55 year old female with a vulvar mass

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Clinical History: Pt is a 55 year old female who presented with a palpable lump in her groin and a vulvar mass that had been present for 3 years. She noted discomfort with certain clothes and while sitting but denied pruritus, bleeding, abnormal discharge and weight loss. After initial treatment with mupirocin ointment failed, she had a lymph node biopsy and imaging done.

PE: 5cmx3cm firm mass in the left labia majora with no discoloration or ulceration of the overlying skin. There was also a 3cm palpable left inguinal lymph node.
CT Abdomen/Pelvis with Contrast (unlabeled)
Contrast enhanced coronal CT demonstrates inguinal lymphadenopathy (white arrow) and a partially visualized mass in the left labia majora (yellow arrow).

Contrast enhanced axial CT better visualizes the entire vulvar mass, which measured 2.7cm.
PET CT (unlabeled)
Corresponding PET CT shows hypermetabolic activity in the vulvar mass (yellow arrow) with a max SUV of 16.9) and bilateral inguinal lymph nodes (white arrows).
Ultrasound of Left Inguinal Lymph Node (unlabeled)
Ultrasound of Left Inguinal Lymph Node (labeled)

Enlarged, round lymph node with irregular borders and heterogeneous echogenicity.
DDX (Based on Imaging)

- Vulvar cancer (adenocarcinoma, squamous cell carcinoma, melanoma, extramammary Paget disease)
- Lipoma
- Leiomyoma
- Bartholin cyst
- Mucinous cyst
Gross Specimen

5cm firm mass with no ulceration of overlying skin
H&E stain with intact epidermis and dermis. There is a glandular mass in the subcutaneous layer (arrow).
Histology

H&E stain demonstrating tumor cells with irregular shaped nuclei and papillary architecture.

H&E stain of lymph node showing normal lymph tissue and tumor invasion (arrow).
Final Dx:

Vulvar Adenocarcinoma
Case Discussion

- Majority of primary vulvar malignancies are squamous cell carcinomas, with adenocarcinomas representing less than 10%
- The pathology results noted the adenocarcinoma arose from an anogential mammary-like gland
- Originally thought to represent remnants of the milk ridges, they are now favored to be a normal cell type in this region
- These glands differ from normal sweat glands in that they are hormone receptor (estrogen and progesterone) positive.
- Differences between breast mammary glands and mammary-like glands of vulva include:
  - Different acinar epithelium
  - Higher concentration of glands than if it were from rudimentary mammary tissue
  - Glands are organized in rows which may suggest cloacal origin vs breast tissue which has linear orientation
  - Mammary ridge is not believed to extend all the way to the vulva
Case Discussion

• Age at diagnosis is usually 50s-80s

• Often present with a unifocal, sometimes pruritic lesion on the labia majora
  • Lesion can also be on the labia minora, mons pubis or clitoris
  • Other symptoms can include bleeding, dysuria, and lymphadenopathy

• Diagnosis is confirmed by biopsy of lesion

• Treatment options for local disease include wide local excision and
  hemivulvectomy and radical vulvectomy with or without lymph node dissection for
  extensive disease
References:
