

# Non-Squamous Lesions of the Vulva

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@saskmary



@saskpathbites





# Outline

## Adnexal

- Benign
  - Bartholin's gland
  - Mammary-like anogenital lesions
- Malignant
  - Extramammary Paget's

## Miscellaneous

- Verrucal xanthoma
- Granular Cell Tumour

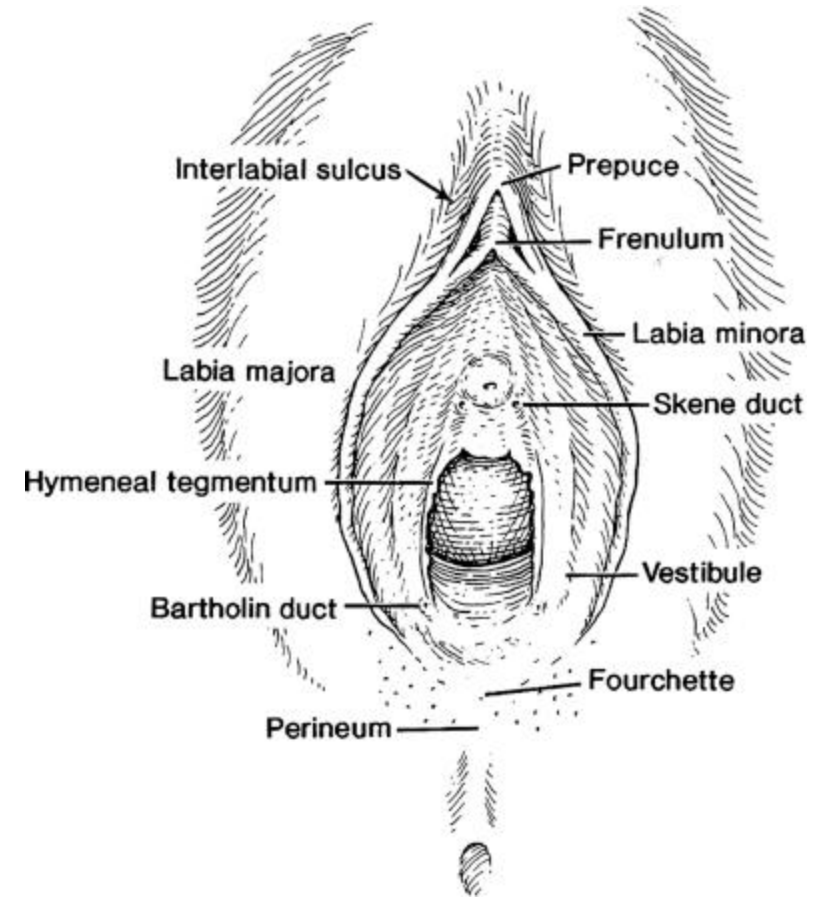
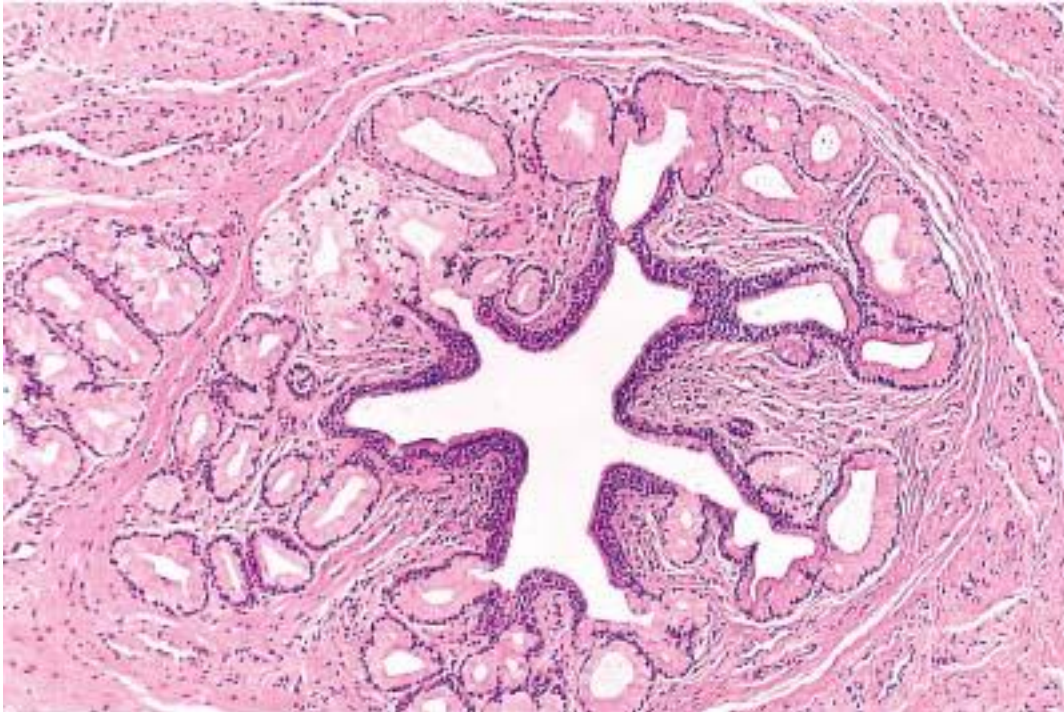
## Pigmented

- Non-melanocytic
  - Lichen planus pigmentosus inversus
  - Cavernous hemangioma
  - Pigmented apocrine hamartoma
- Melanocytic
  - Benign
  - Malignant



# Batholin's gland

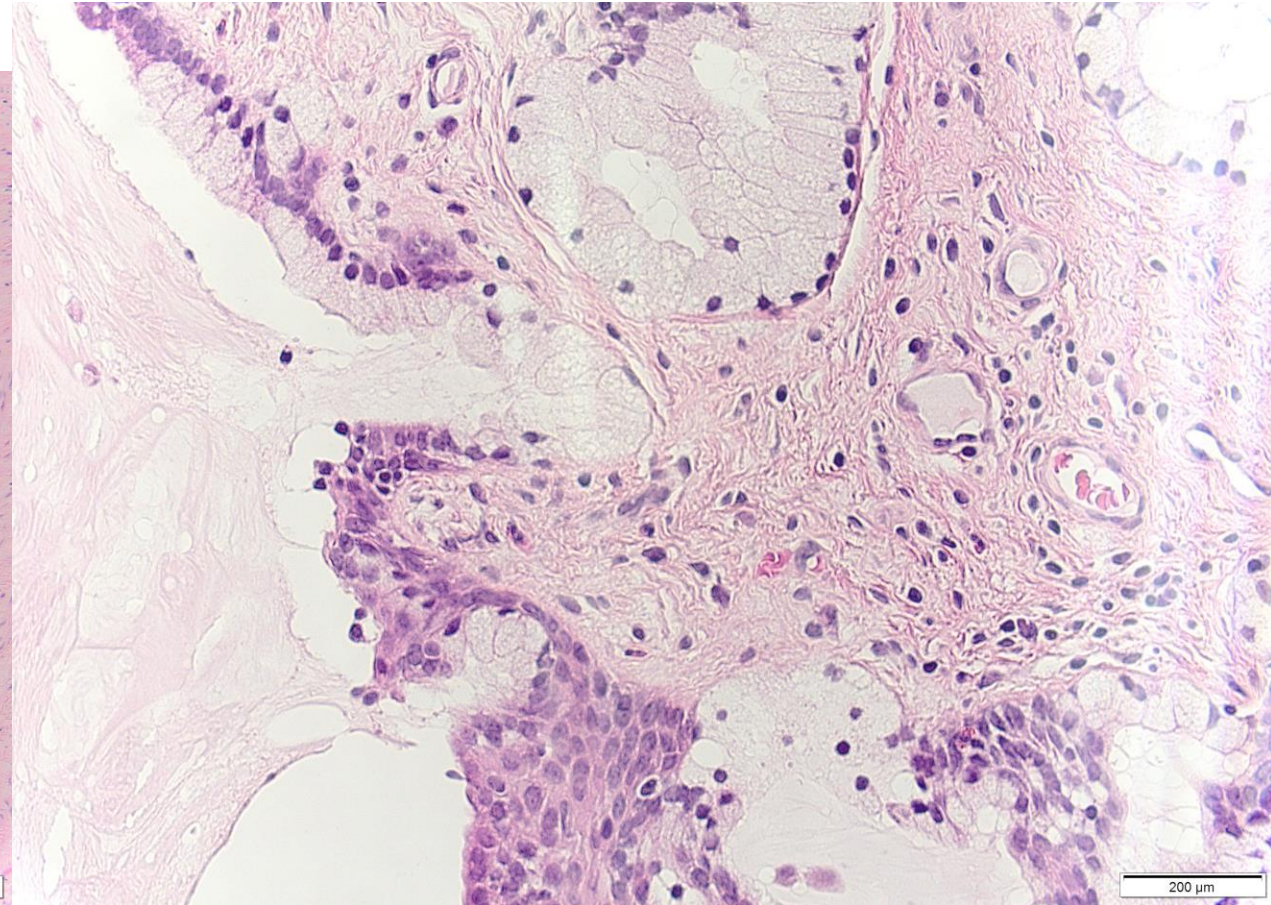
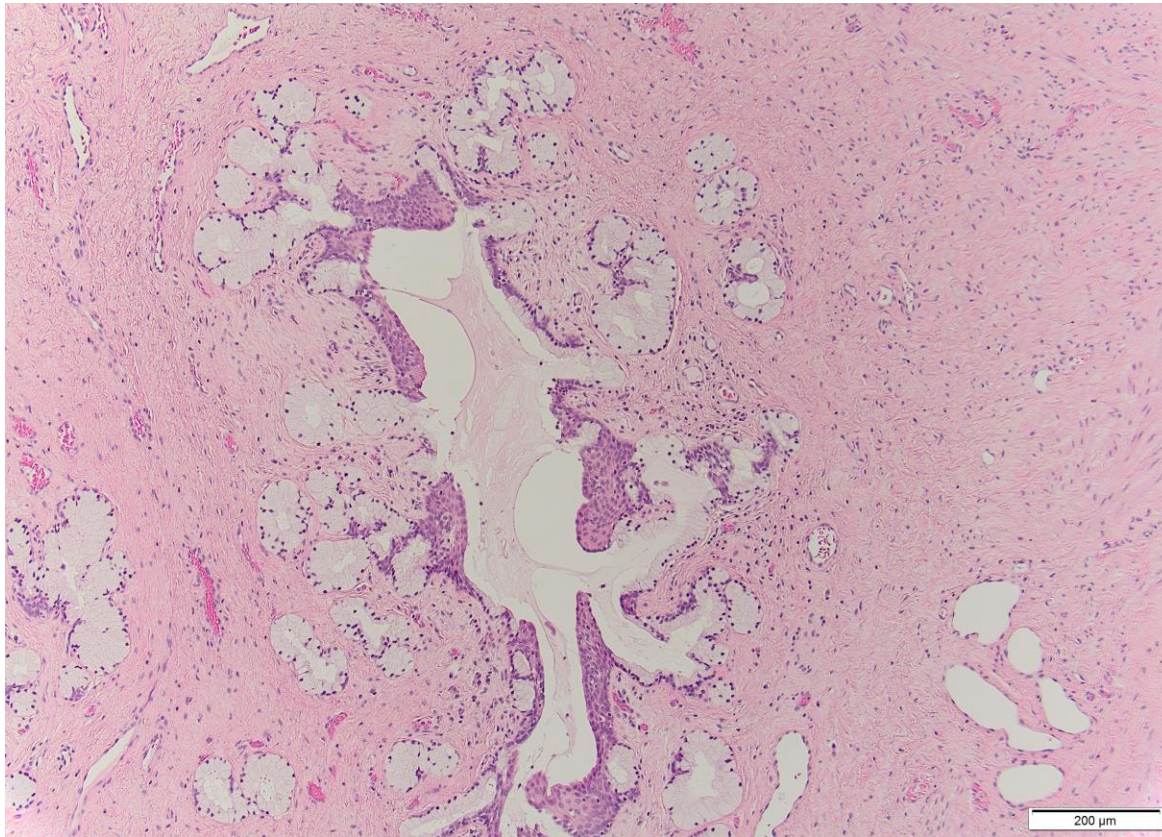
- Major vestibular gland
- Bilateral
- Acini are mucus-secreting columnar cells





# Bartholin's duct

- lined by squamous, transitional, mucinous, ciliated, or flattened nonspecific epithelium



# Bartholin's Duct Cyst



```
graph LR; A[Obstruction of orifice] --> B[secretions fills cyst]; B --> C[Secondary chronic and acute inflammation];
```

Obstruction  
of orifice

secretions  
fills cyst

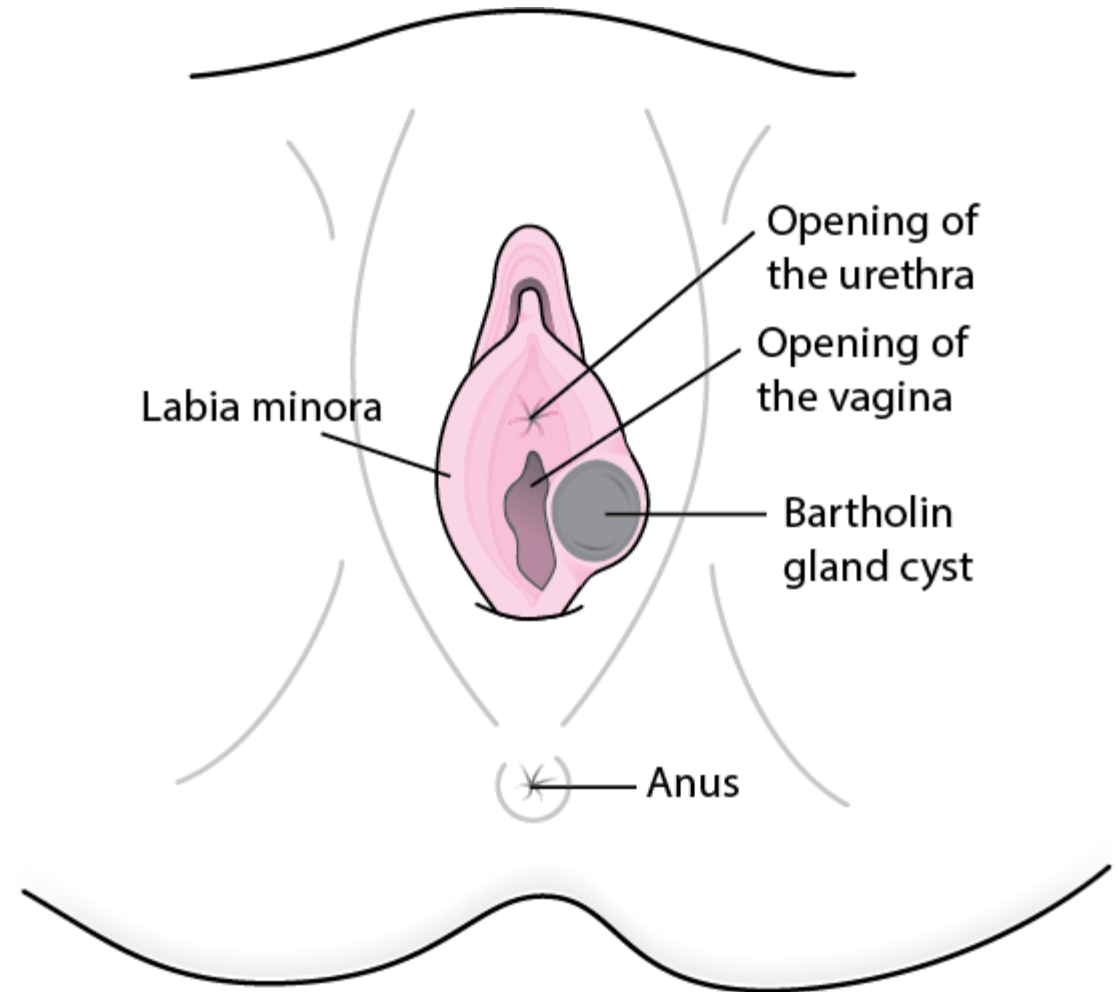
Secondary  
chronic and  
acute  
inflammation

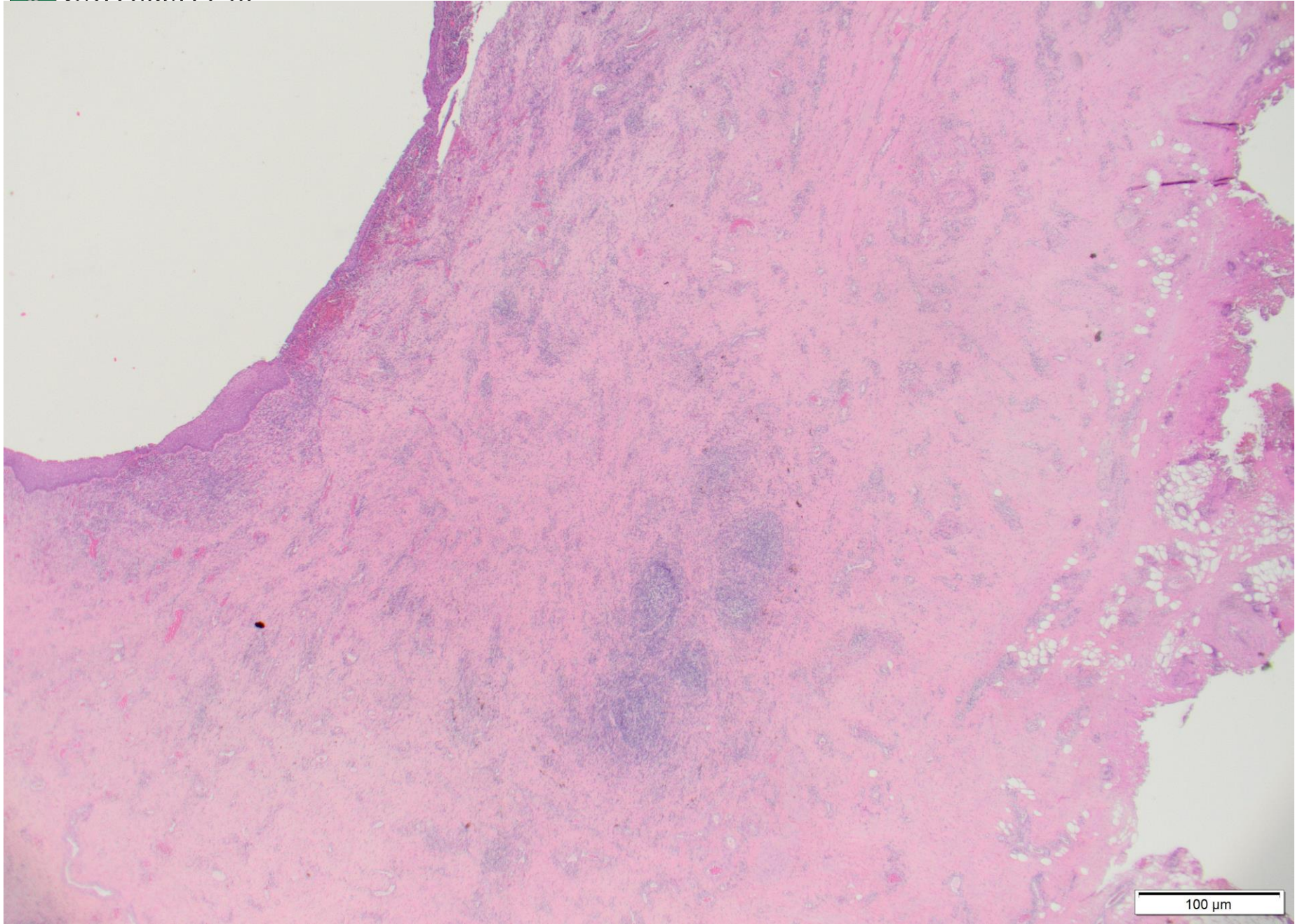


# Bartholin's gland cyst



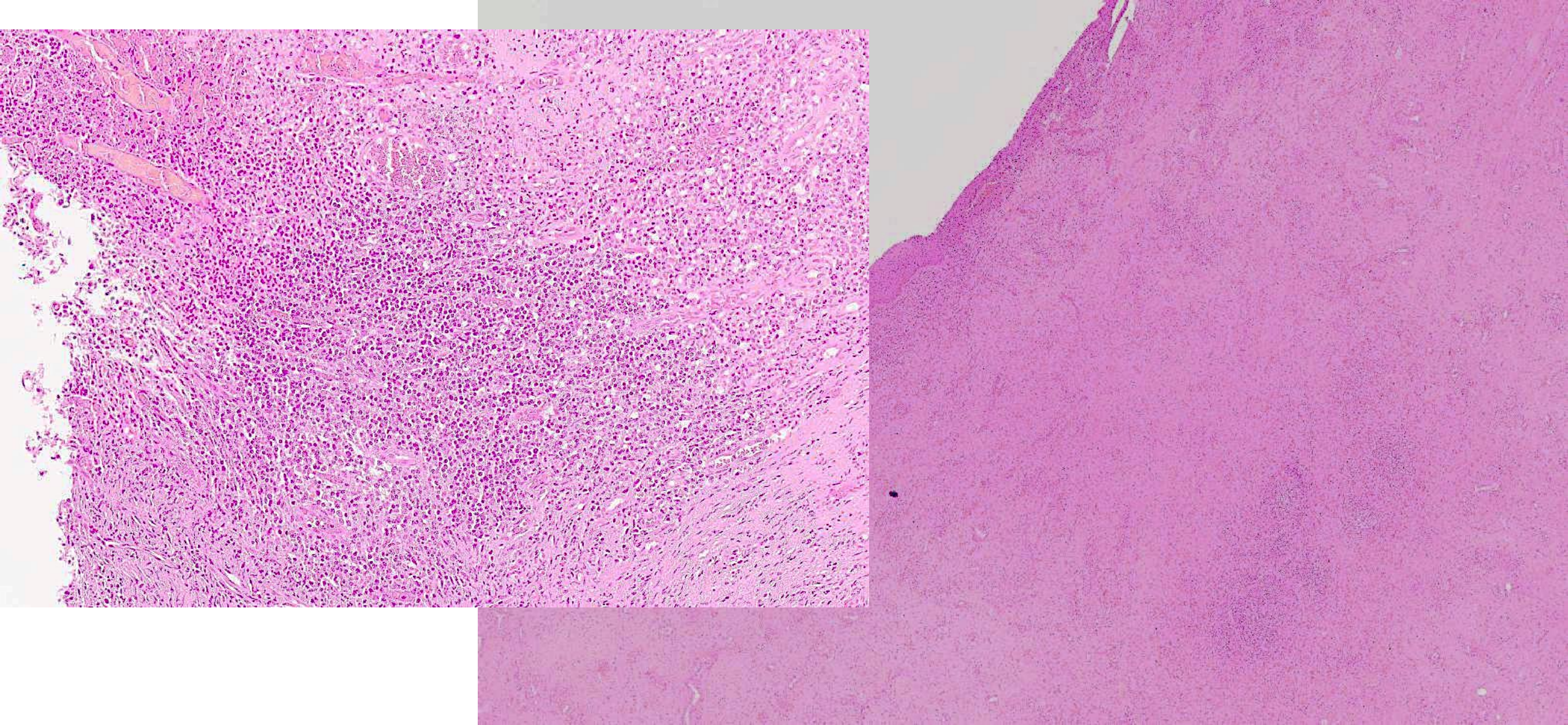
Lateral introitus



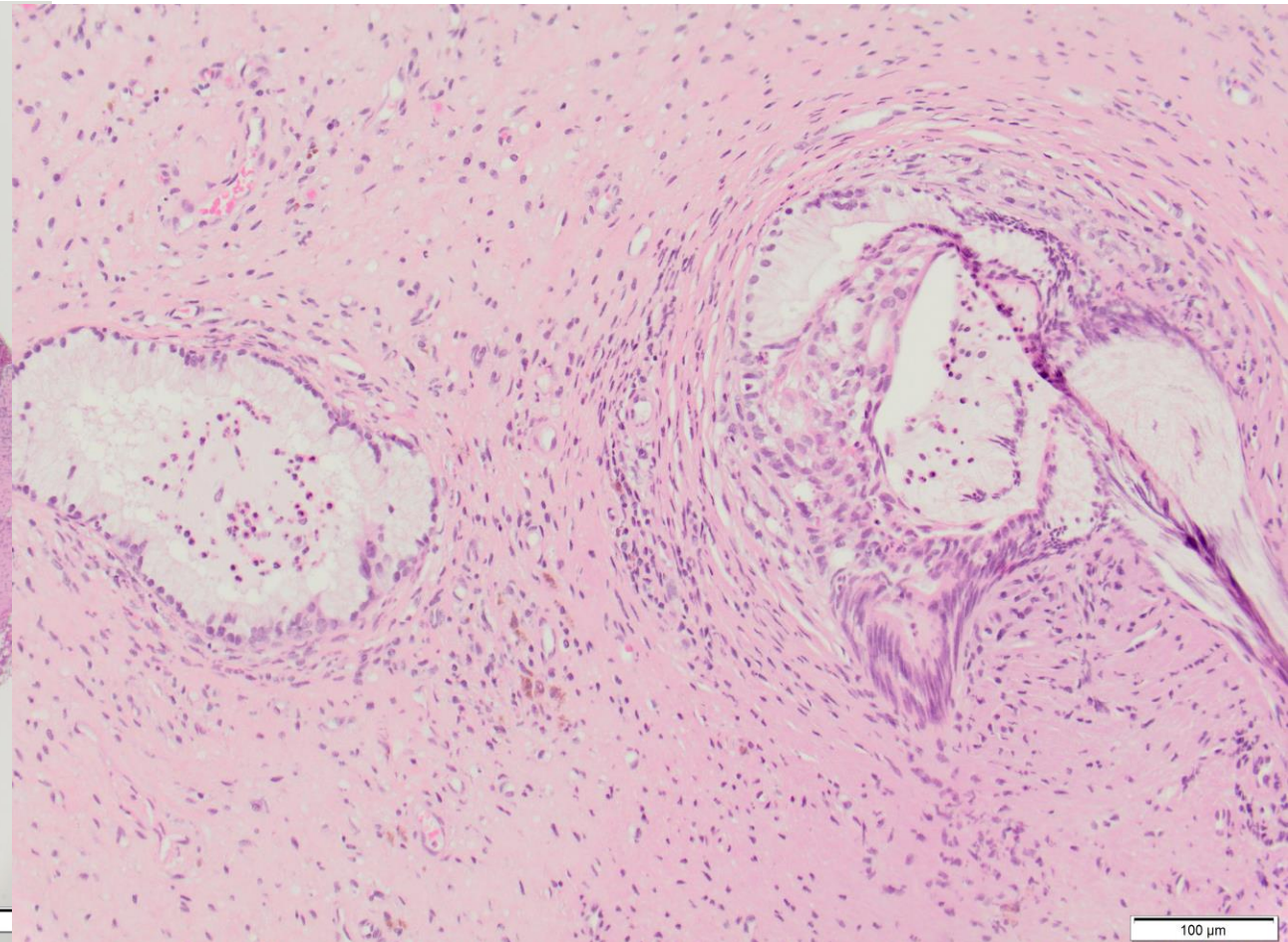
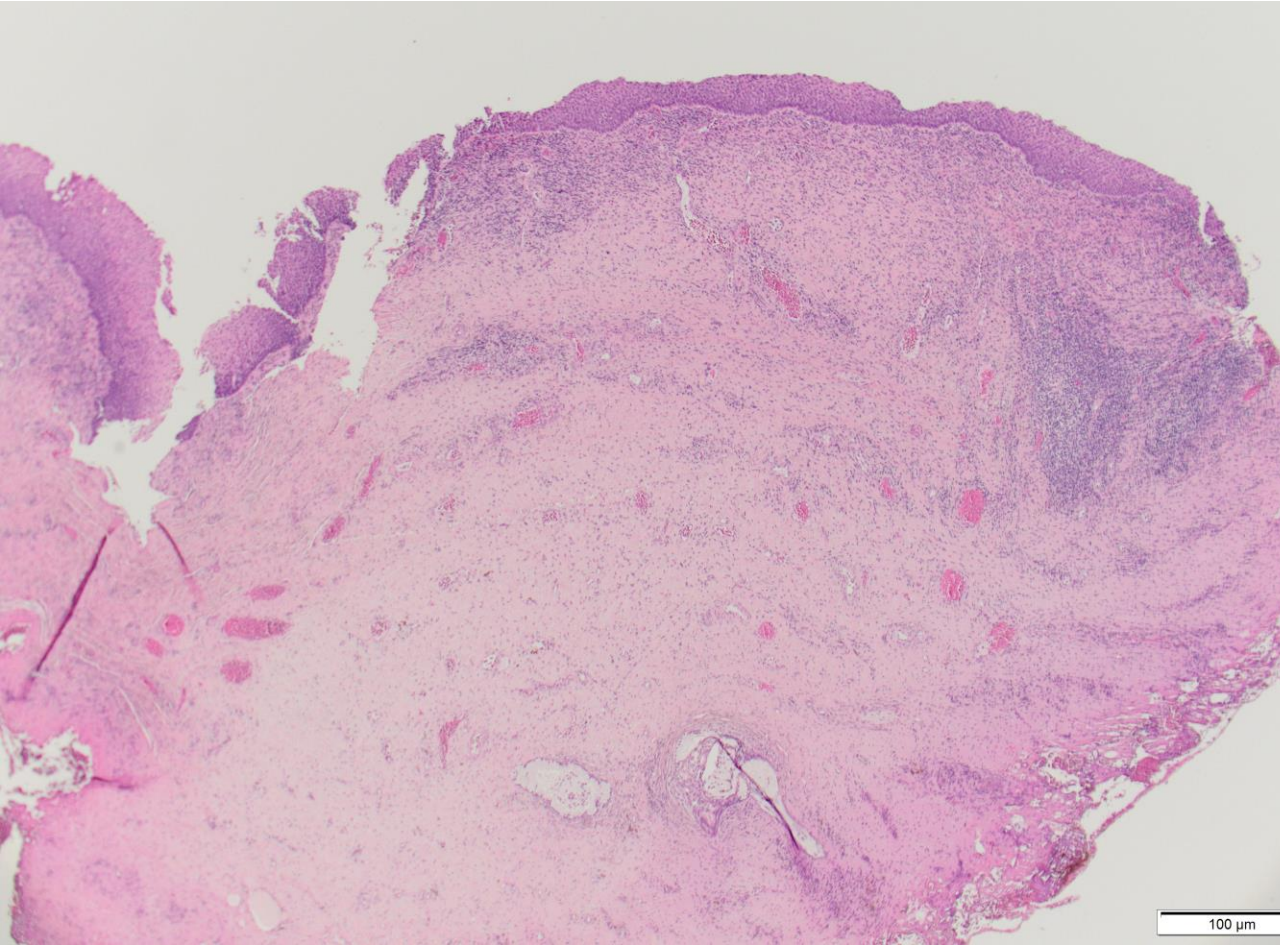




# Bartholin abscess





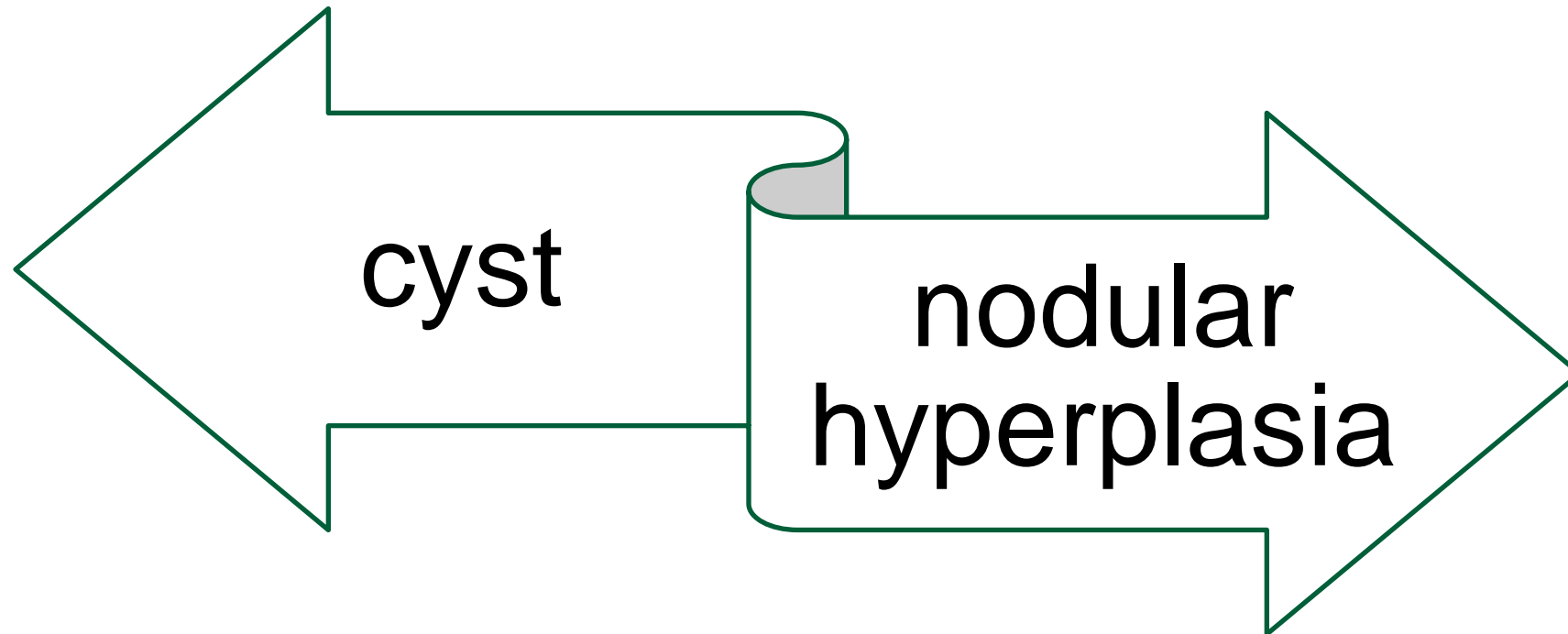




# Bartholin gland carcinoma

- involves the area of the Bartholin gland
  - ✓ histologically compatible with the origin from the Bartholin gland
  - ✓ areas of apparent transition from normal elements to neoplastic ones
  - ✓ no evidence of primary tumor elsewhere
- lined by squamous, transitional, mucinous, ciliated, or flattened nonspecific epithelium

# Bartholin's Gland enlargement



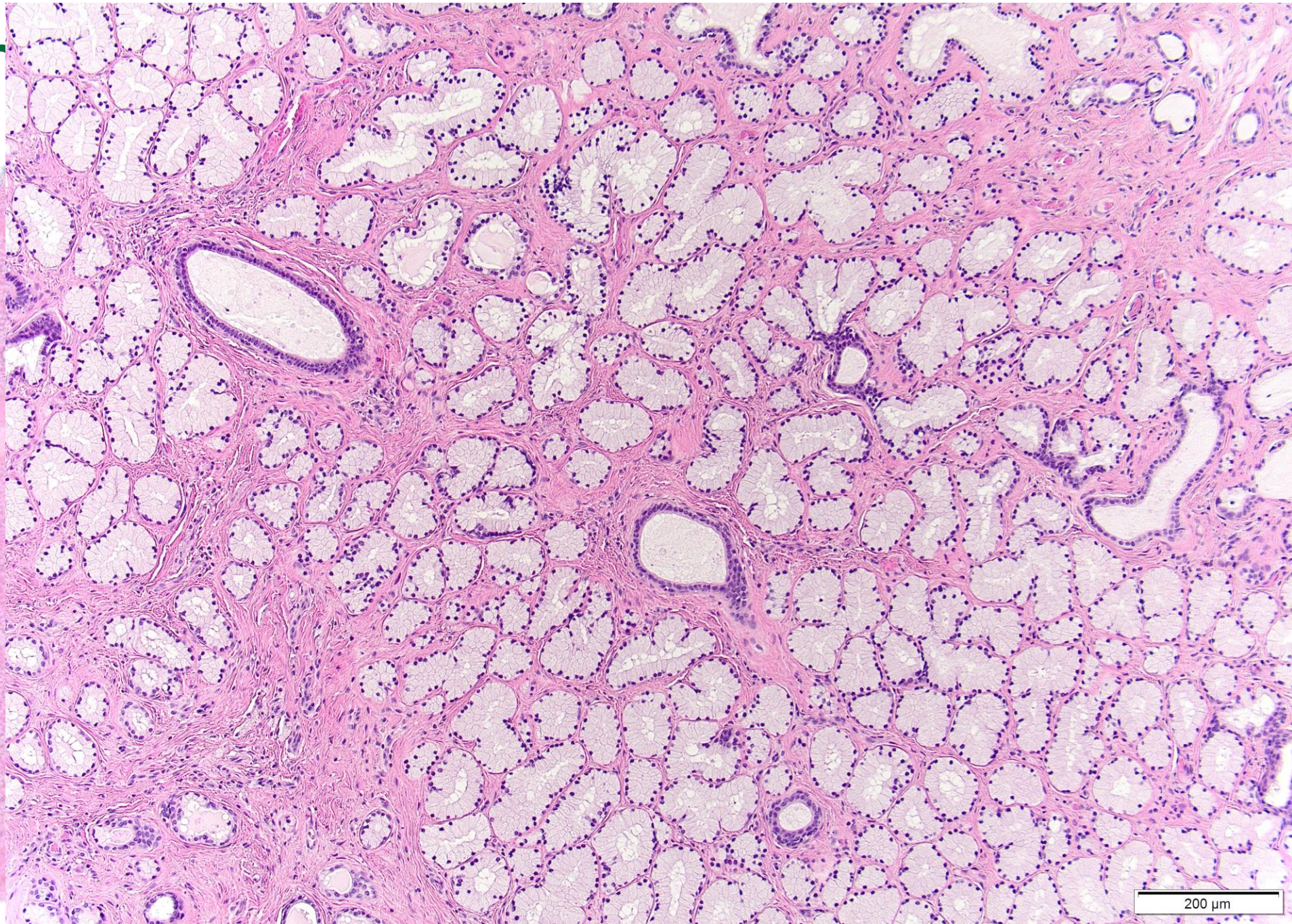


# Nodular Hyperplasia – Bartholin's gland

- Rare, solid, unencapsulated
- proliferation of mucus-secreting acini with preservation of the normal duct-to-acinar relationship
- Distinct from adenoma



# Bar

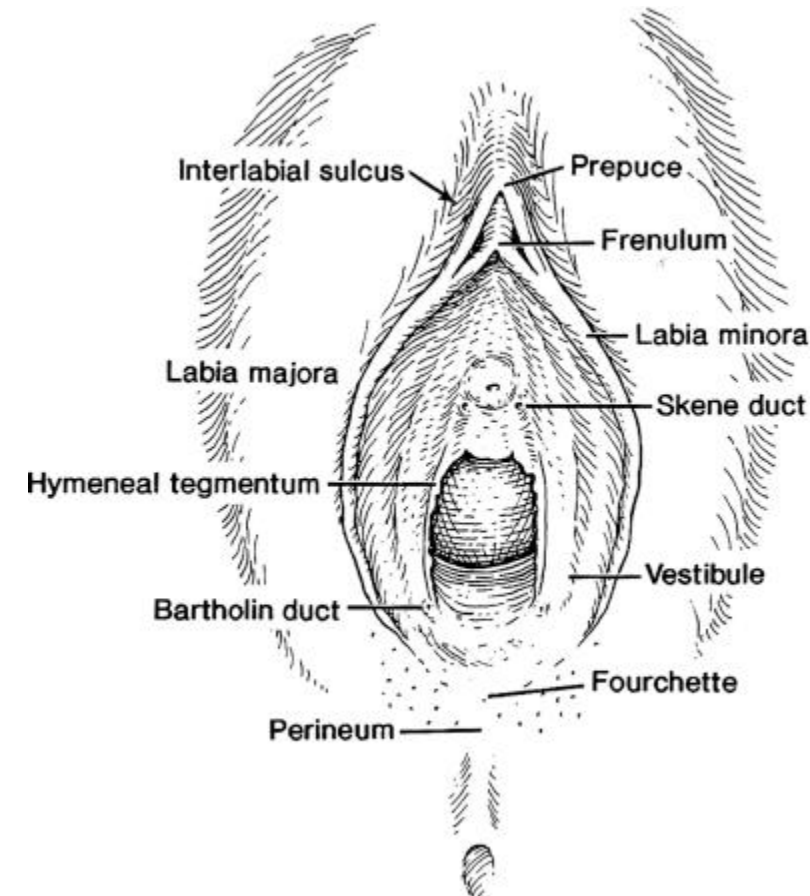


200  $\mu$ m



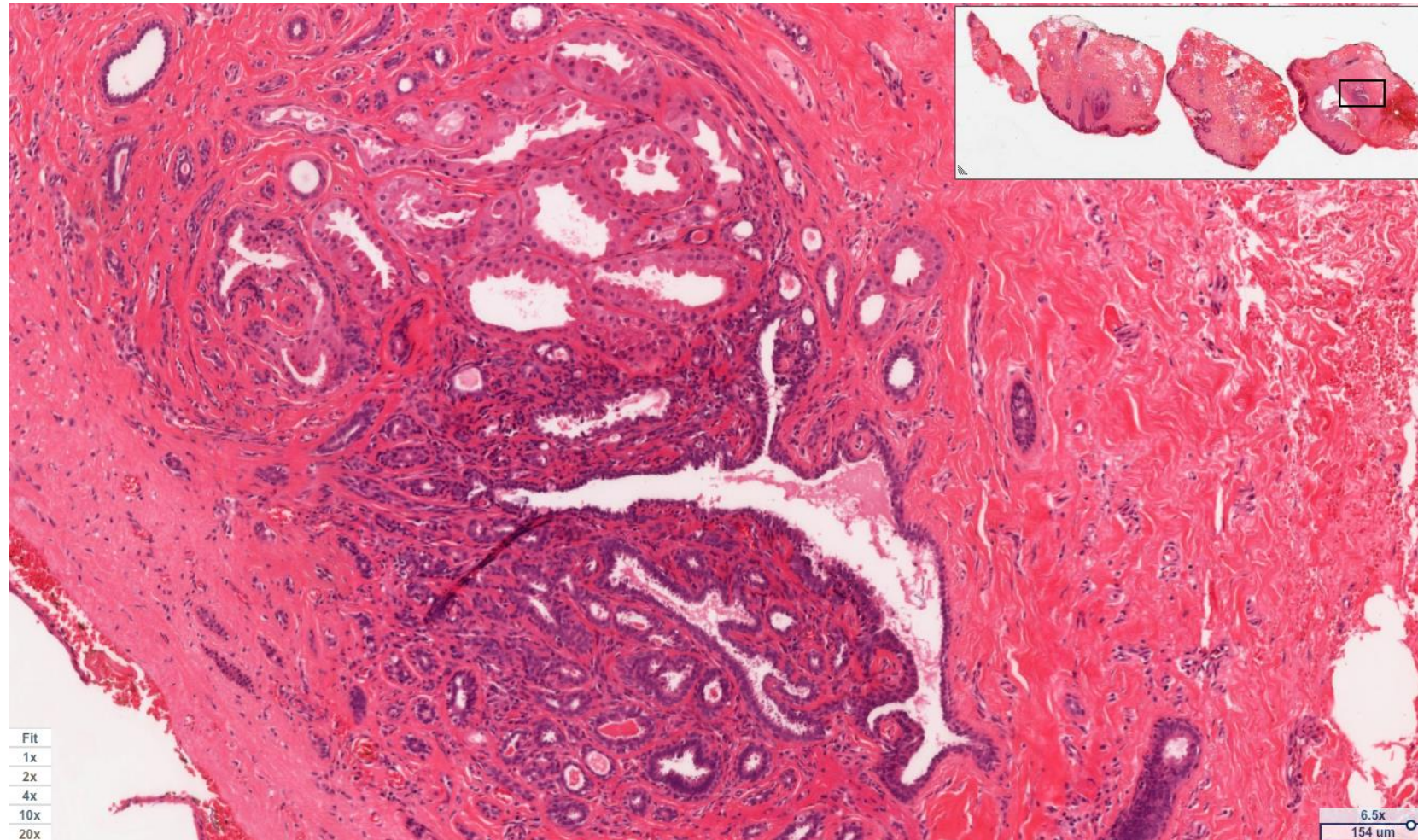
# Anogenital Mammary-like Tissue

- Why it's important to know about
  - Normal constituent of the anogenital region
- Where they occur
  - At the junction of the skin and cloaca-derived mucosa
  - Inter labial sulcus and inner side of labia majora
- What are they
  - Glandular lining with “snouts”
  - Myoepithelial lining



# Anogenital Mammary-Like Tissue

- Any breast pathology can happen in the vulva

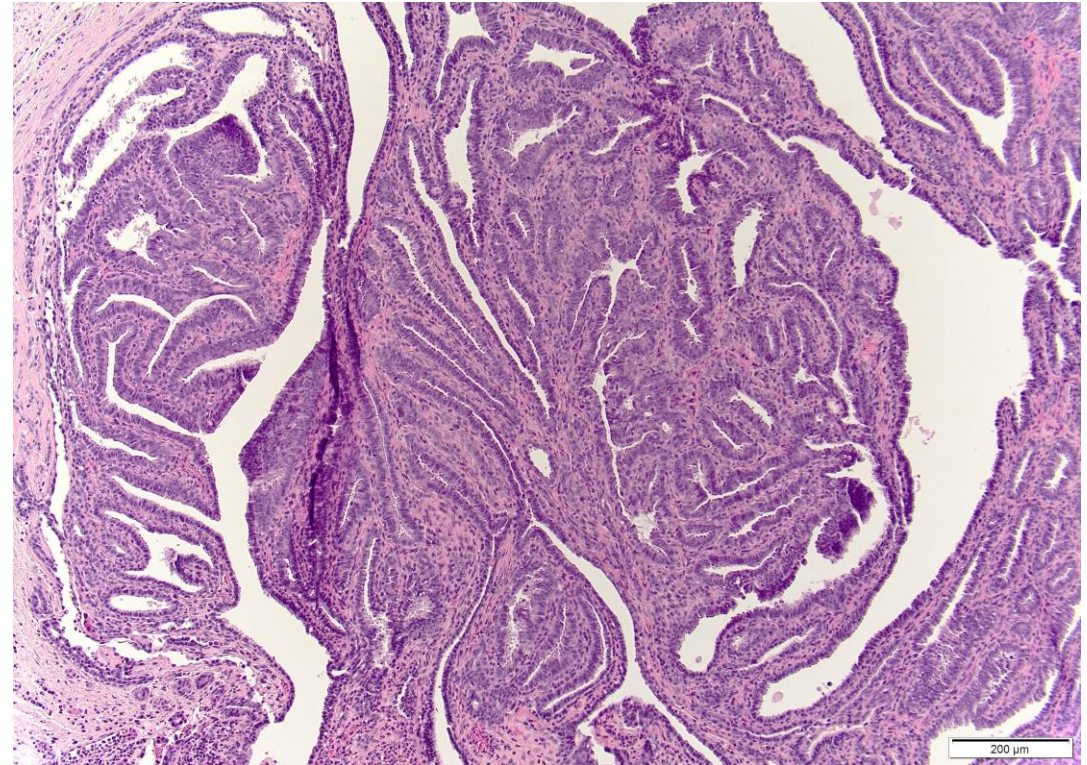
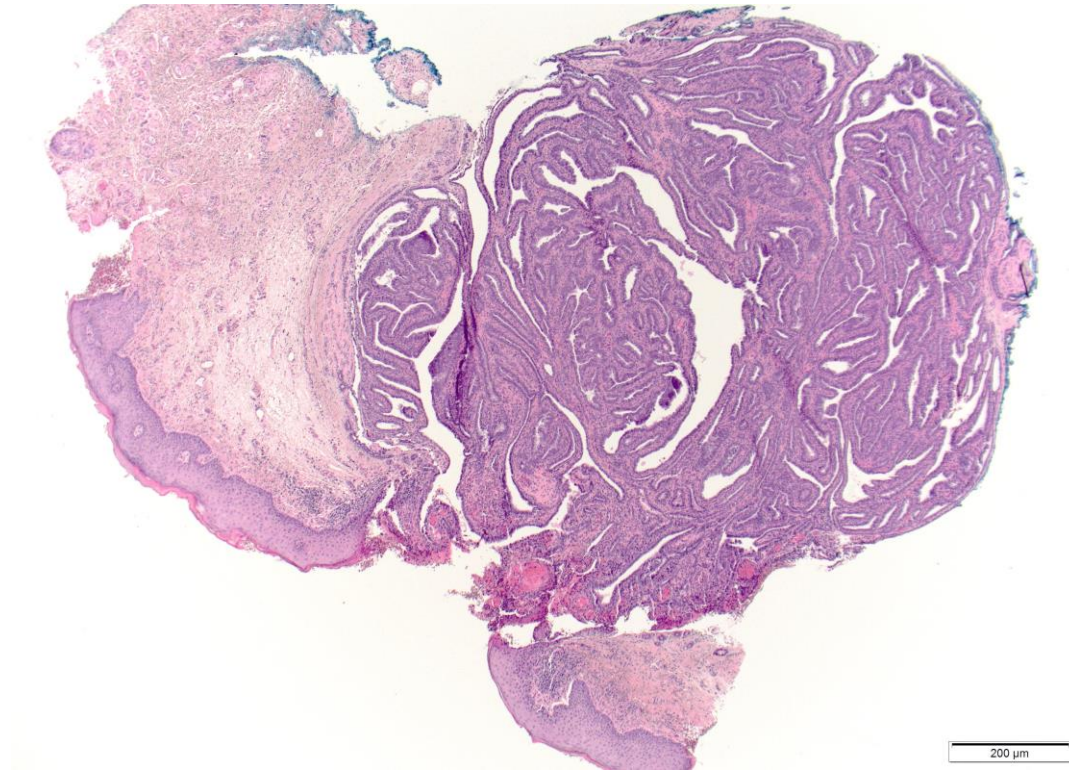




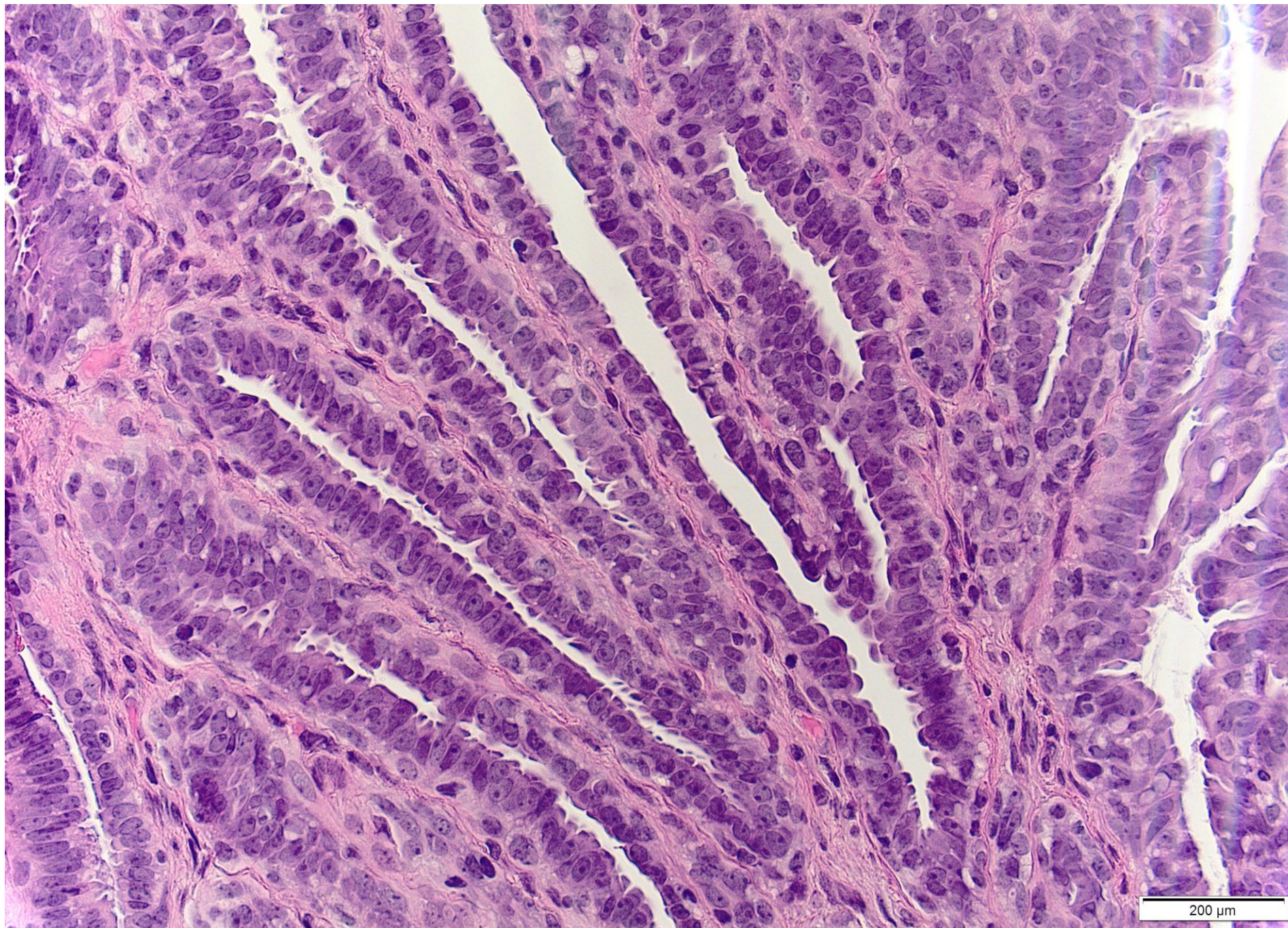
# Hidradenoma Papilliferum

- Most common anogenital mammary-like glandular adenoma
- Gross description:
  - Slow-growing, firm reddish nodule
- No clear correlate in the breast

# Papillary fronds



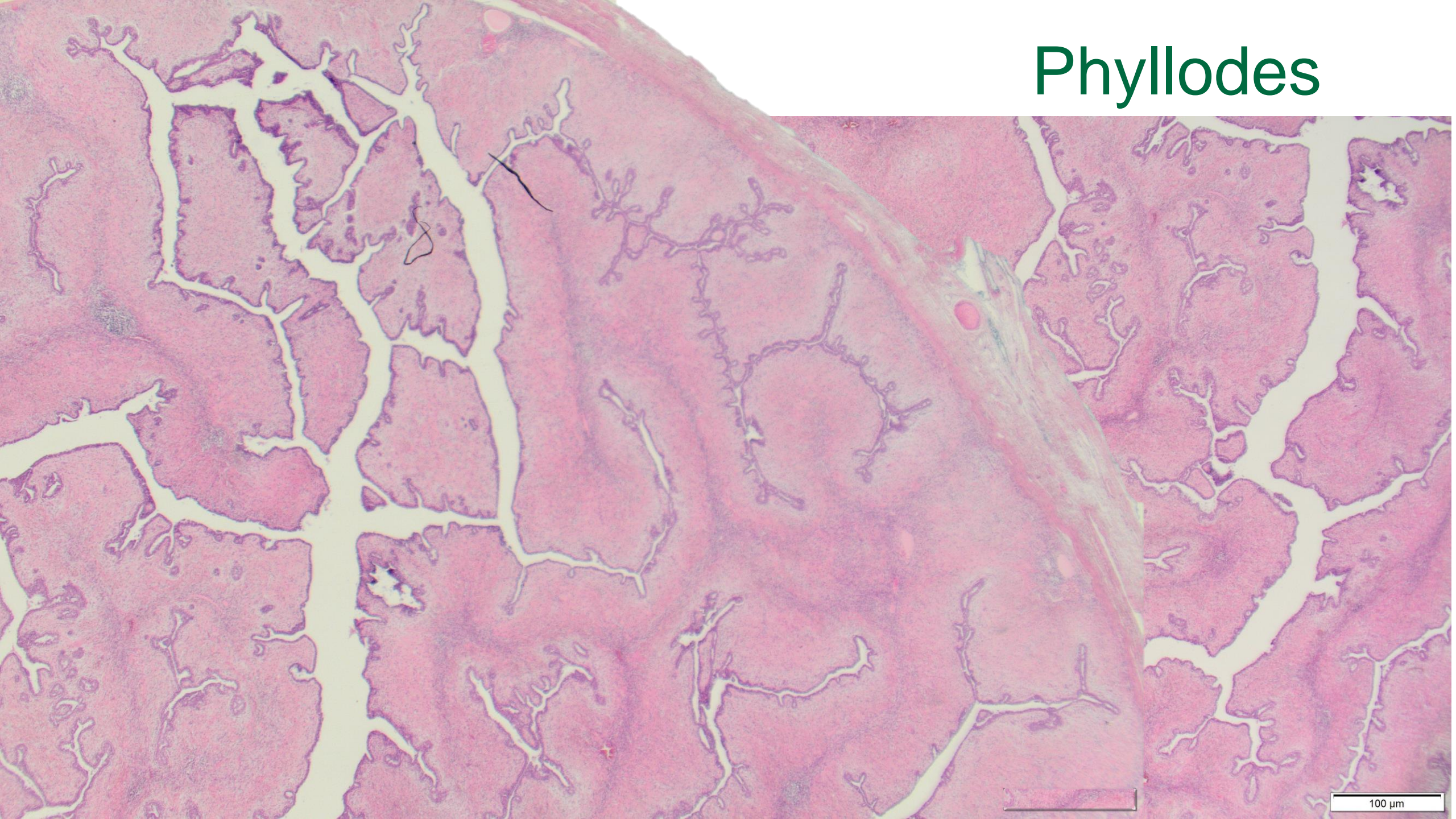




Apocrine-type cells  
Decapitation,  
secretion

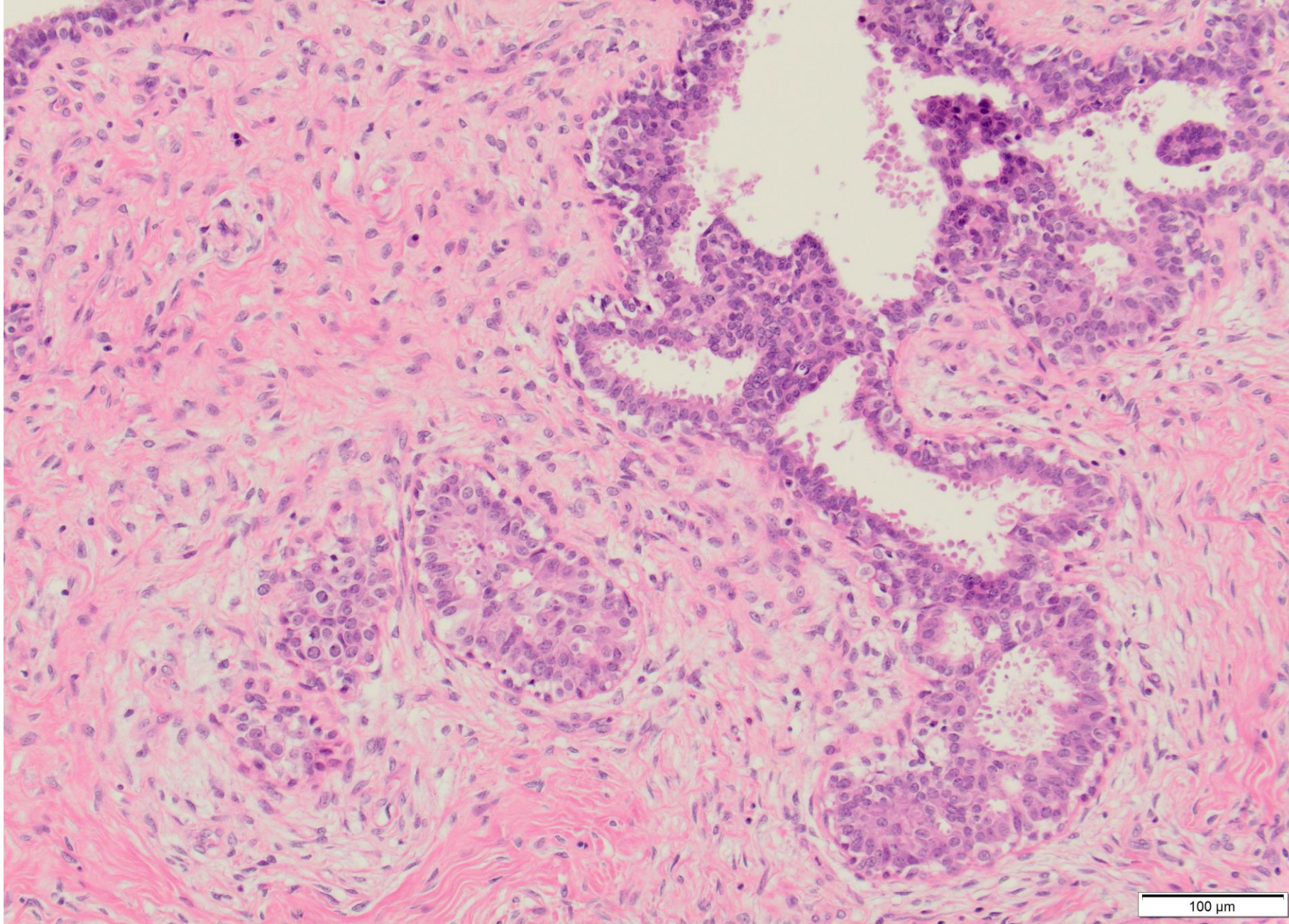


# Phyllodes



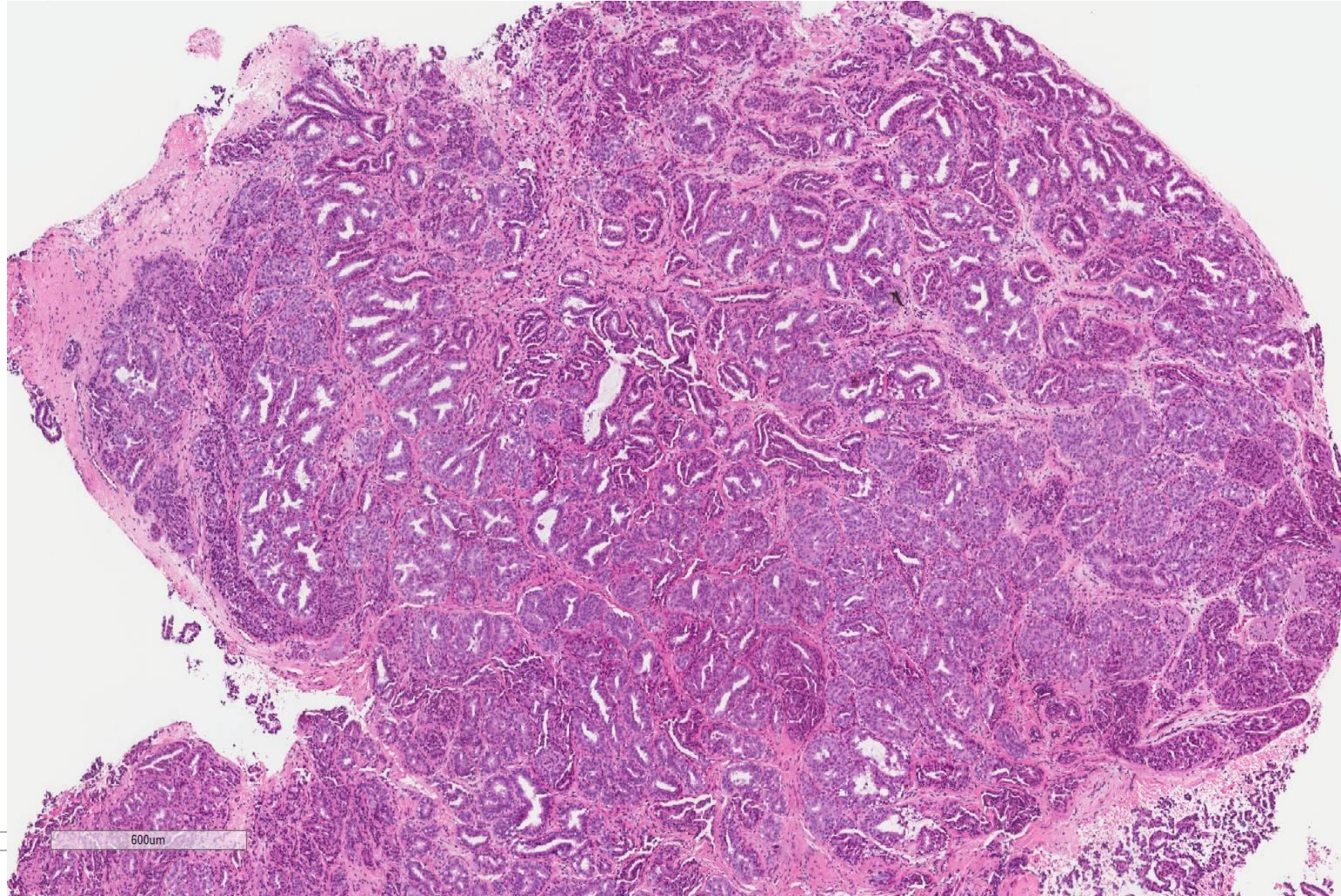
100  $\mu$ m



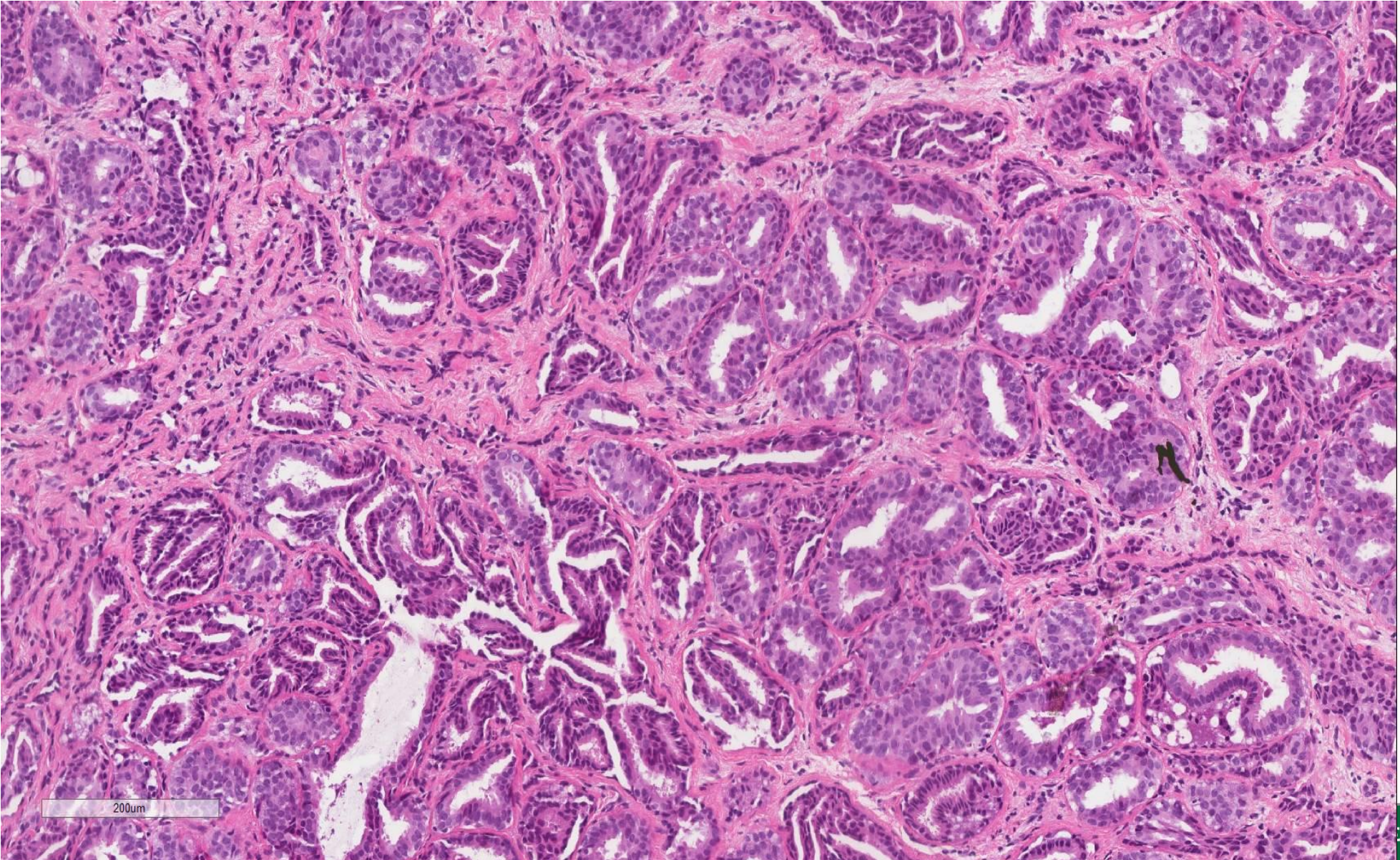




# 51 yo "lump" on labium majus

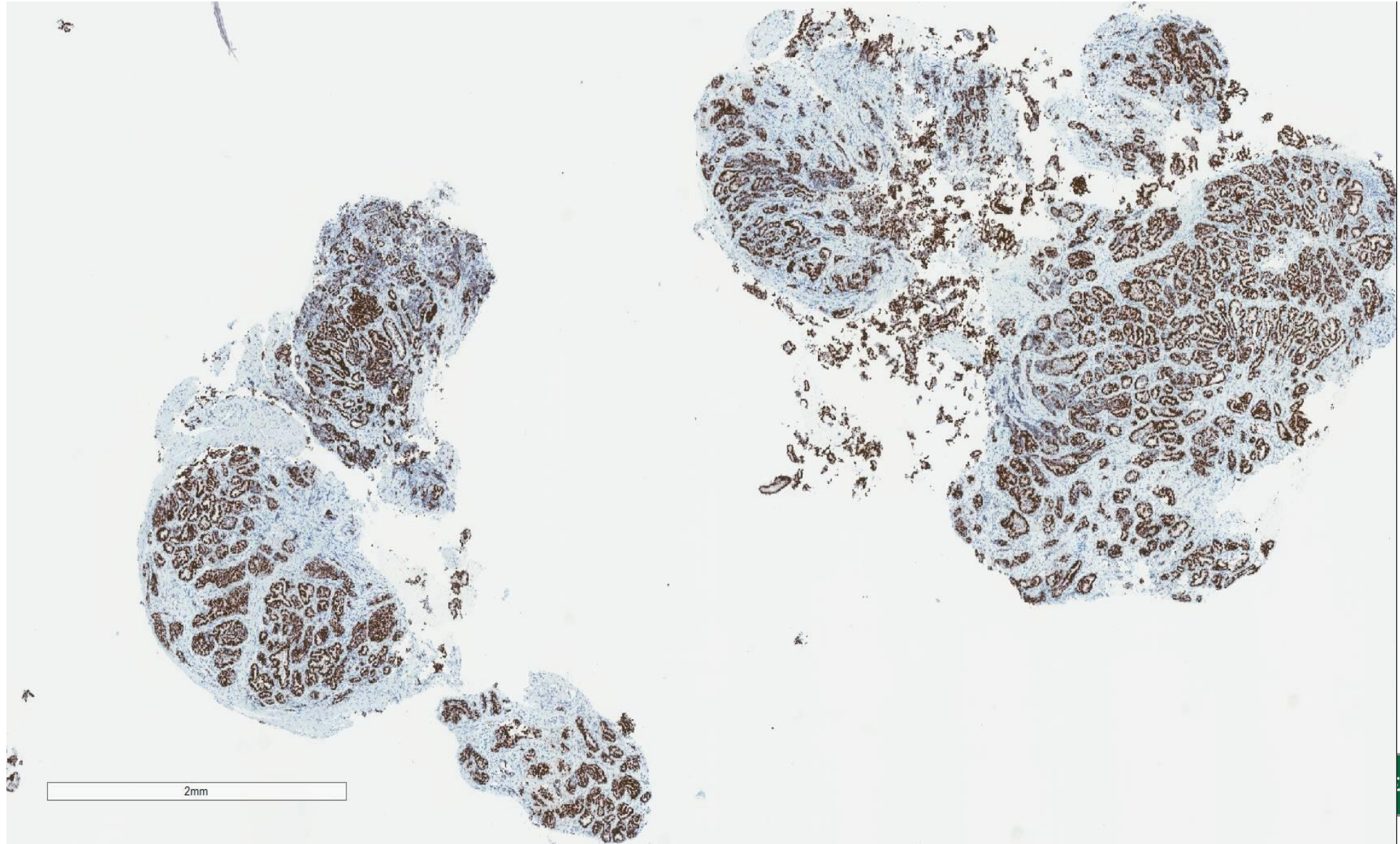






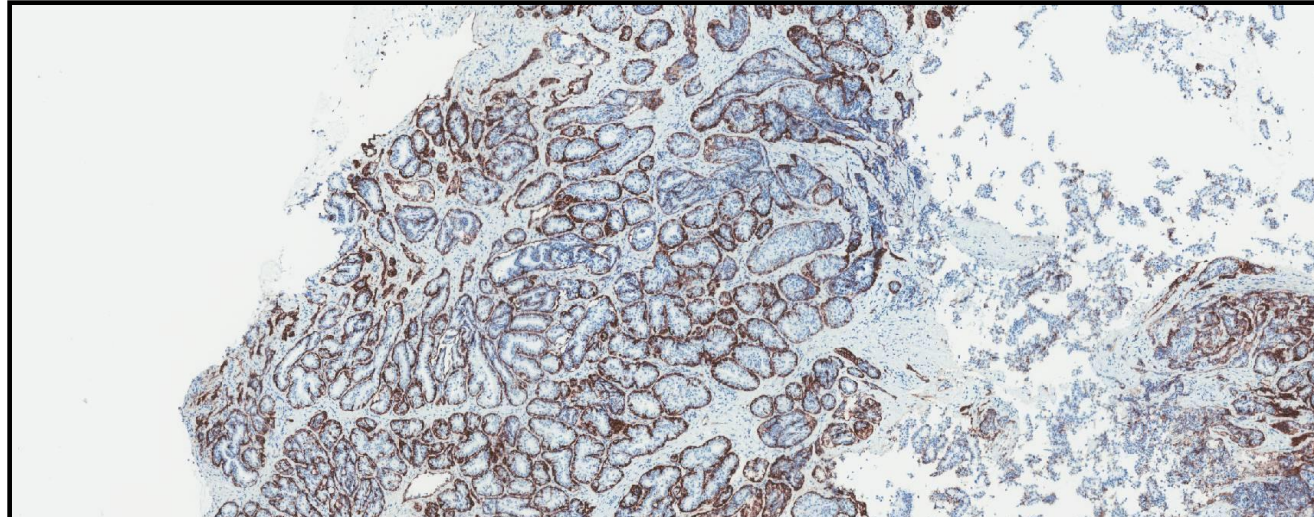


**GATA-3**



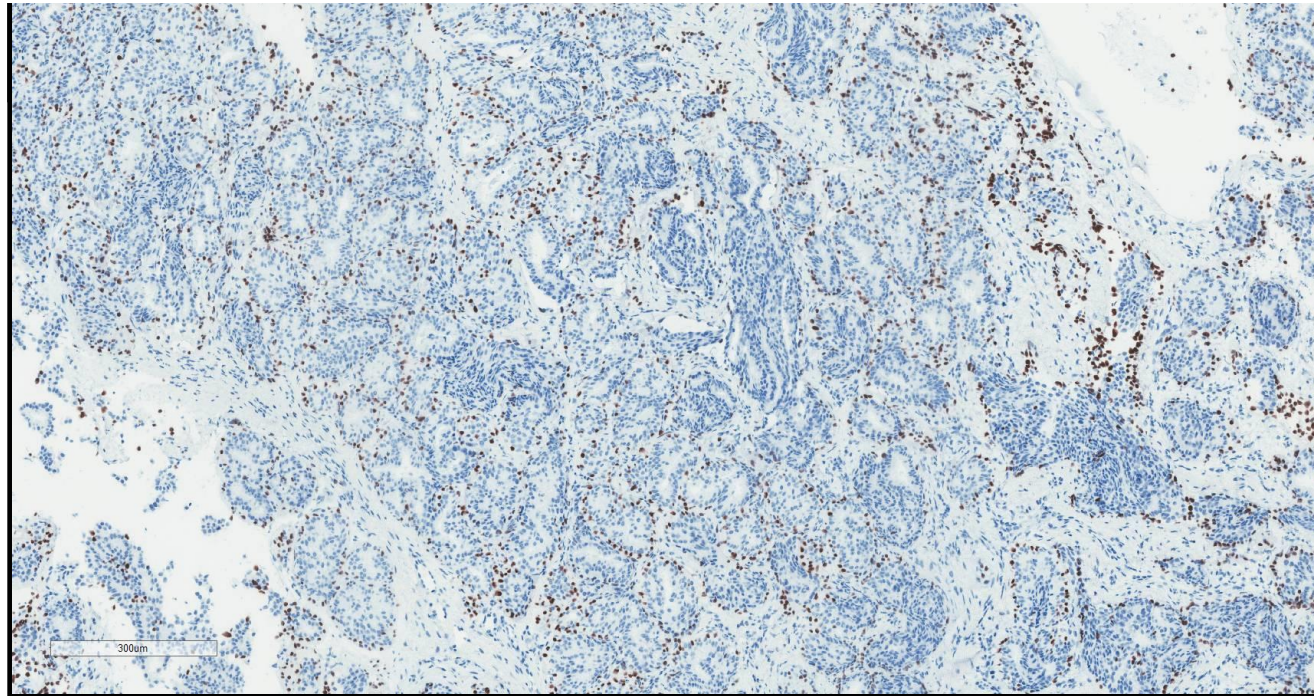


**CD10**



## Mammary-Like Anogenital Adenoma

**p63**





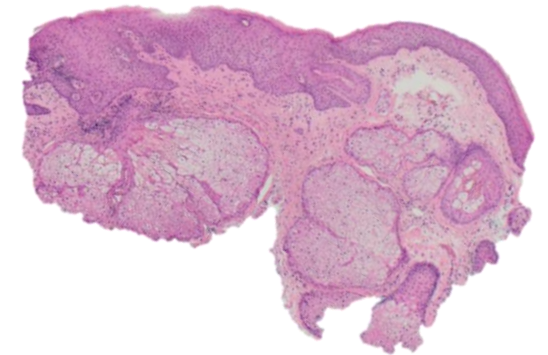
# Fordyce's spots/granules

Small (1-5 mm) slightly elevated yellowish or whitish papules or spots

- Vulva is one place (others are vermillion border or glans of penis)

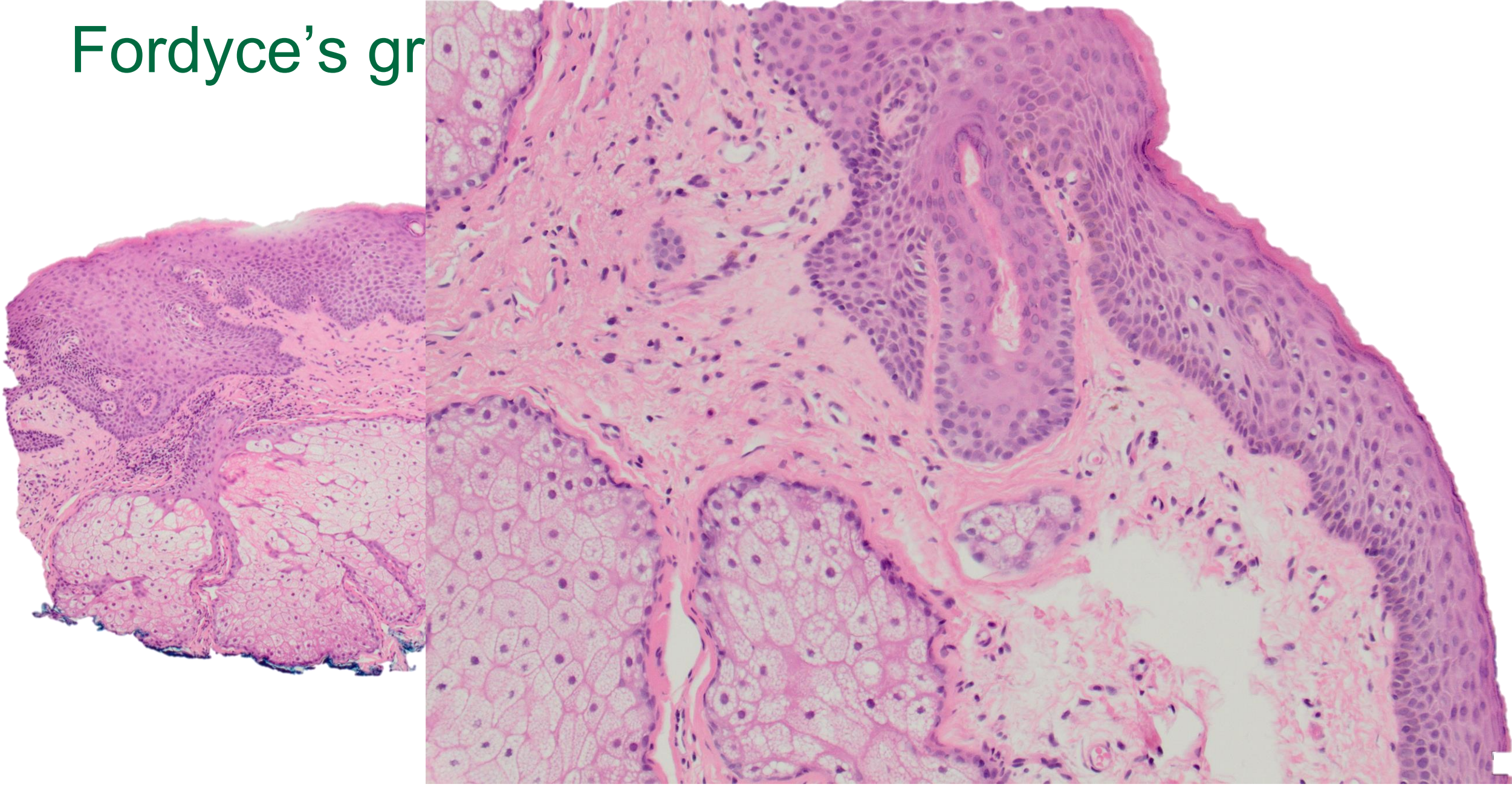
Definition is an ectopic sebaceous gland lacking a pilosebaceous unit

- Good clinical/pathologic correlation





# Fordyce's granules





# Miscellaneous

Verrucal Xanthoma

Granular Cell Tumour



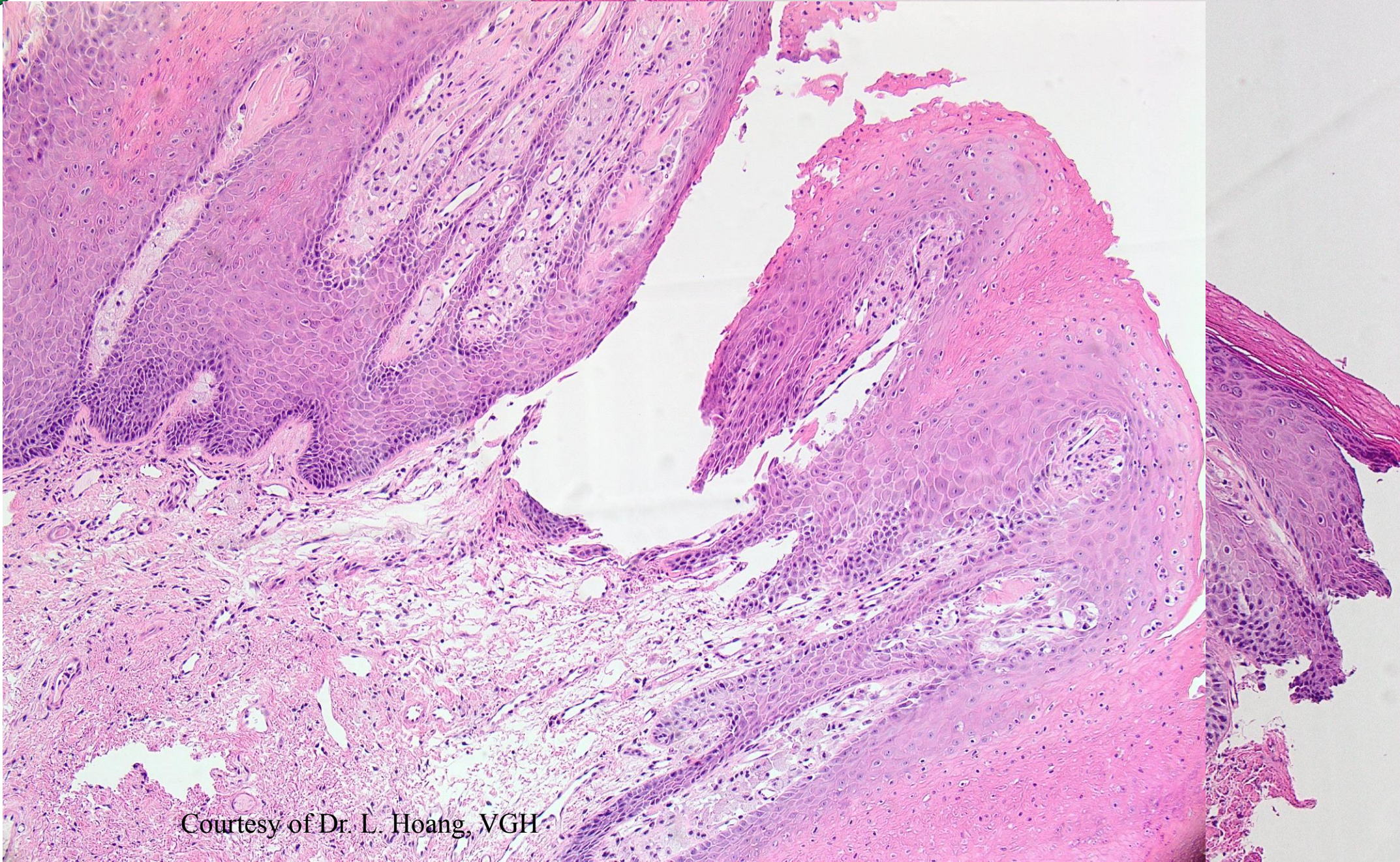
# Verrucal Xanthoma

Definition: rare, benign tumour of the oral or vulva

Clinical: asymptomatic yellowish-orange verrucous plaques

Can be clinically very worrisome





Courtesy of Dr. L. Hoang, VGH



# Verrucal Xanthoma

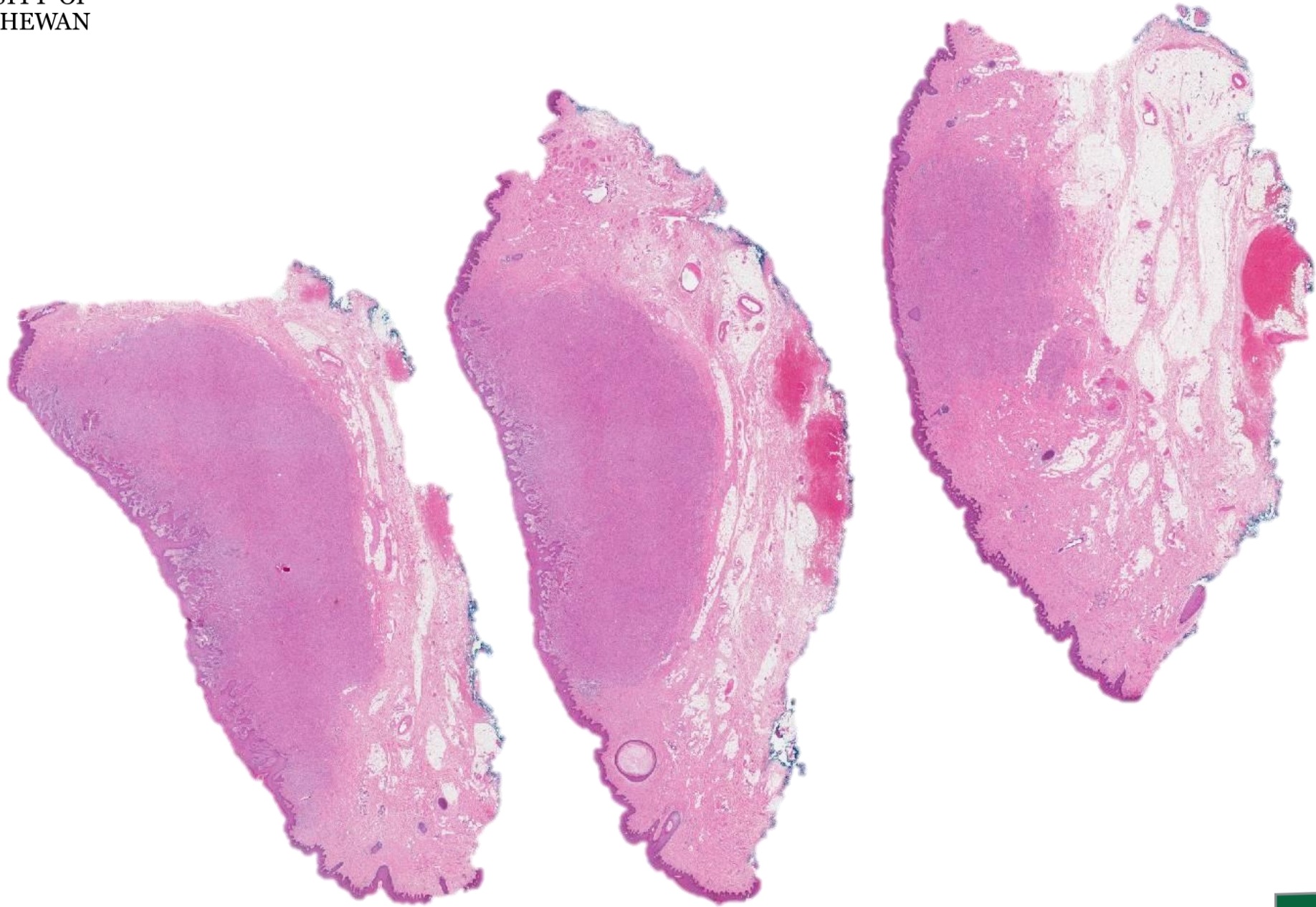
- Hyperkeratosis
- Acanthosis
- Elongation of rete ridges
- Granular layer absent
- Eosinophilic change
- Xanthomatous cells confined in the papillary dermis
  - Lipid-laden histocytes (foamy cells)
  - Tiny granules



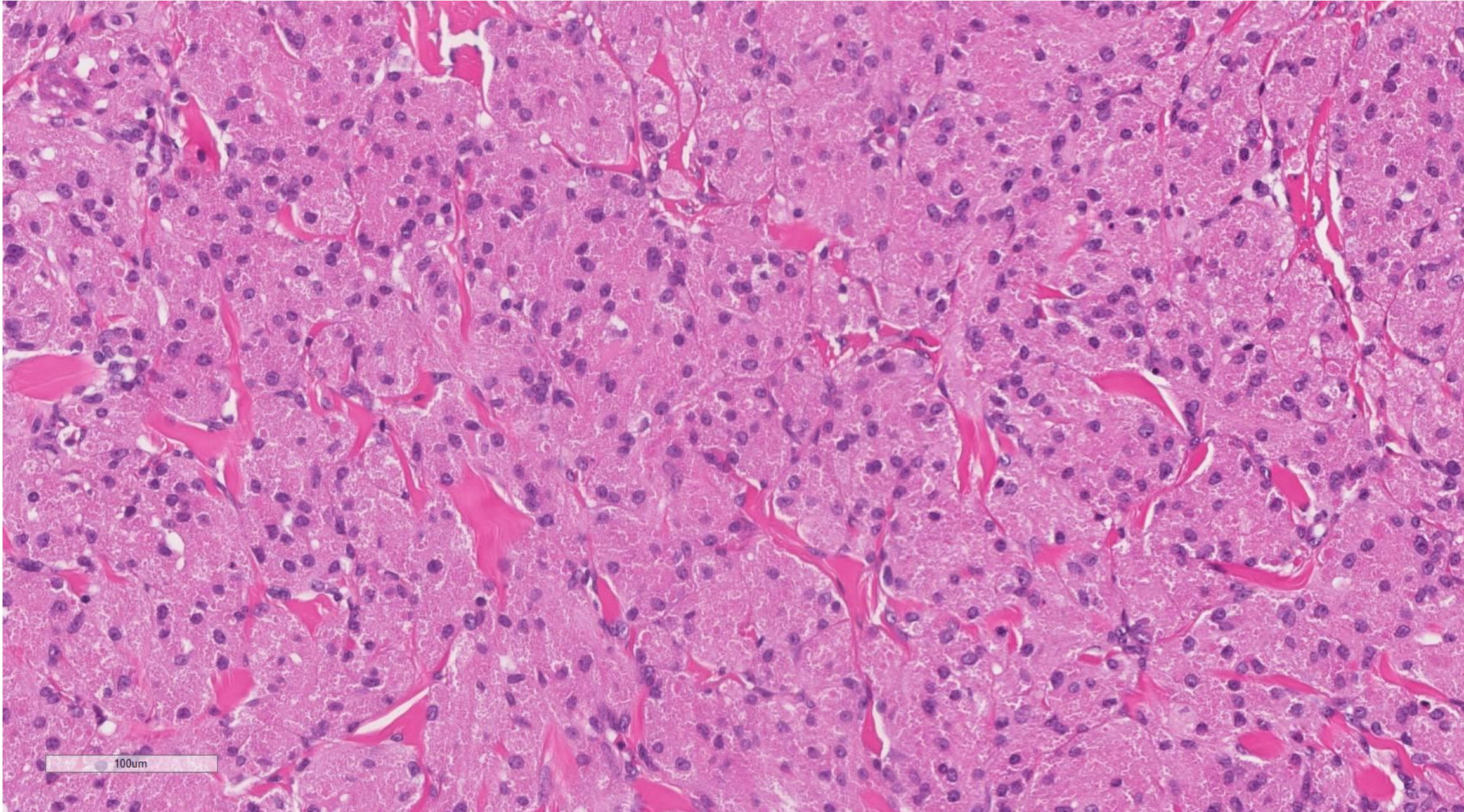
# 67 year old – treating with apple cider vinegar



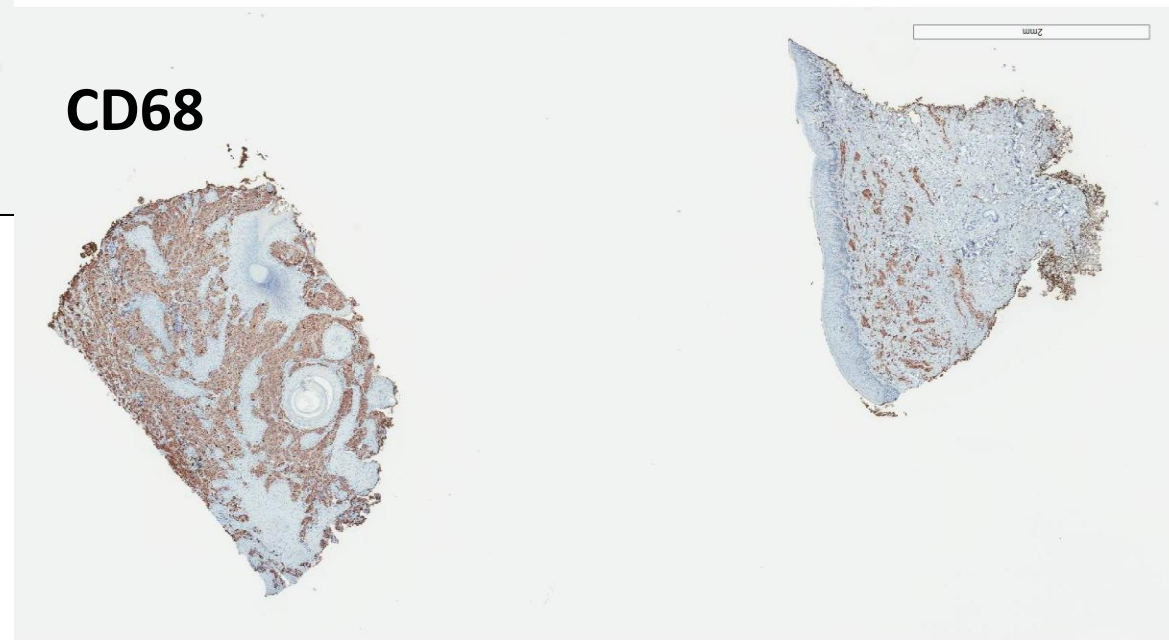
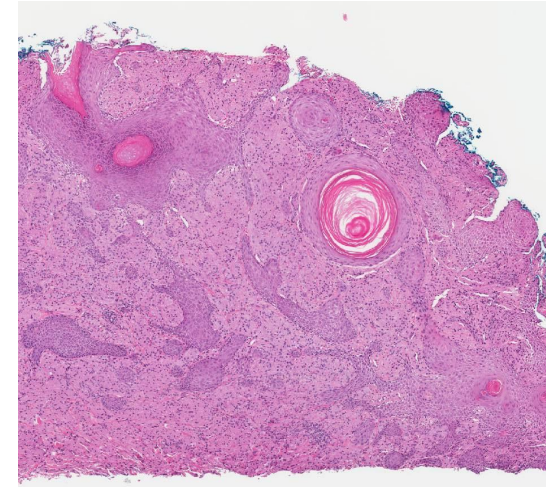
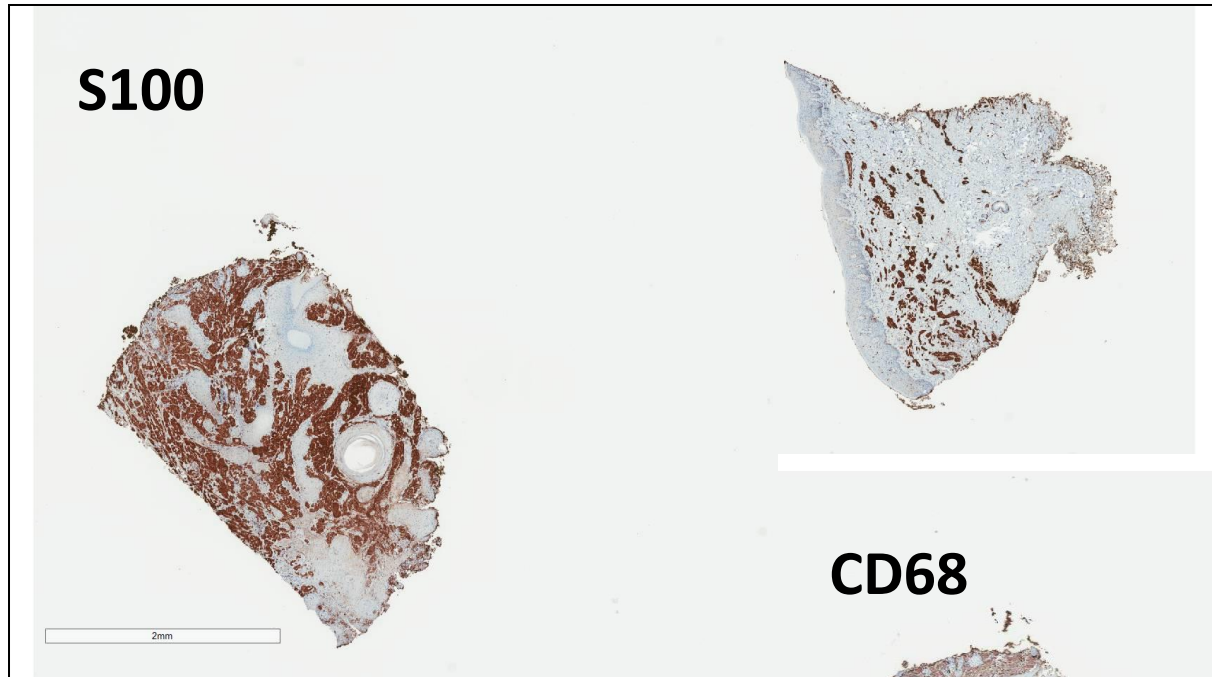














# Granular Cell Tumour

Definition: rare (usually benign) soft tissue neoplasm of neural origin

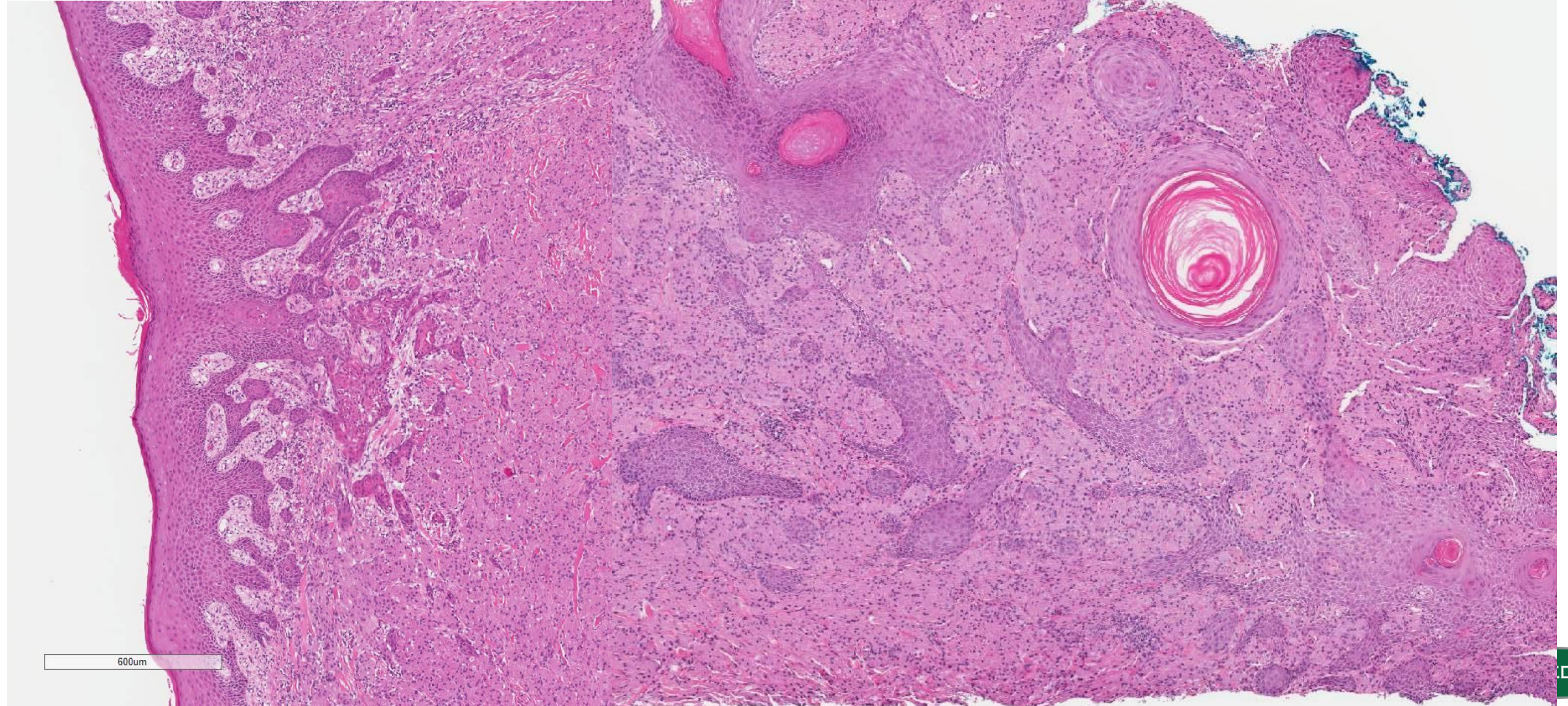
Clinical: females > males as an asymptomatic mass

Can occur anywhere in the body

- 15% situated in the vulva
- Highest incidence in the 4<sup>th</sup> and 5<sup>th</sup> decade



# Pseudoepitheliomatous Hyperplasia

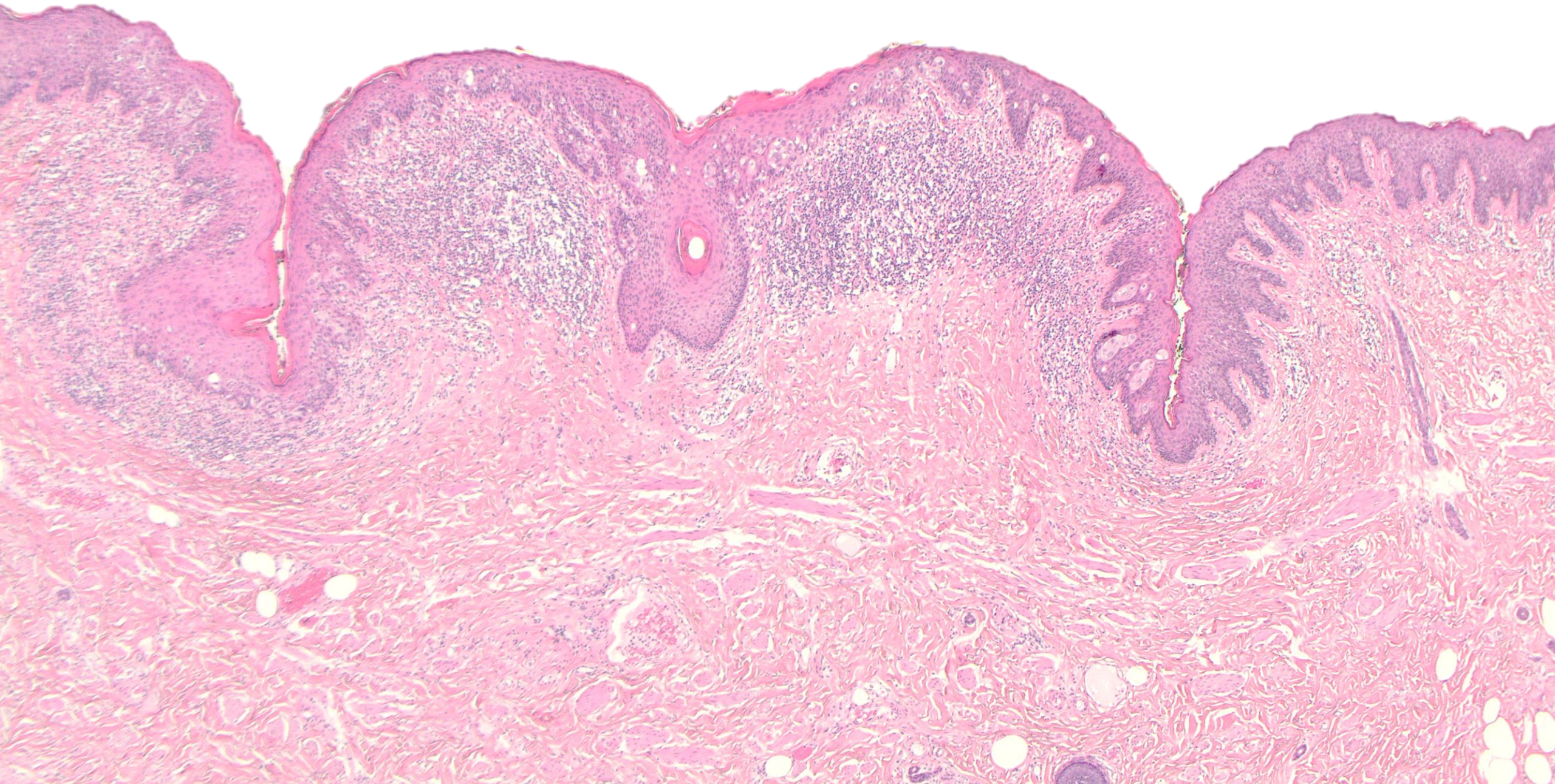




# Extra Mammary Paget's Disease

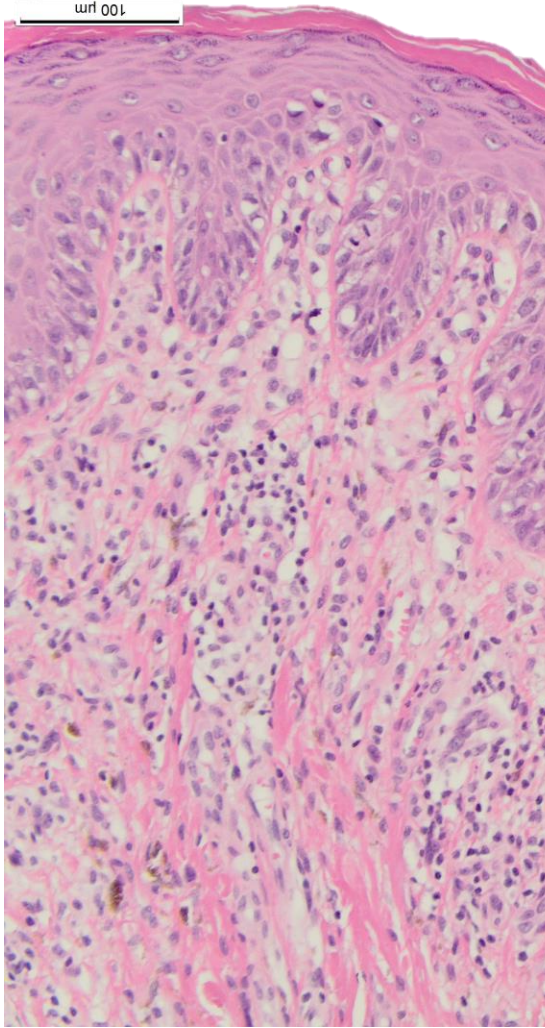
- Definition: intraepidermal adenocarcinoma in any region outside the breast
  - Anogenital being the most common
- Malignant in situ lesion of cutaneous sweat glands
- Underlying malignancy present in <25%
  - Colon
  - Bladder
  - Cervix



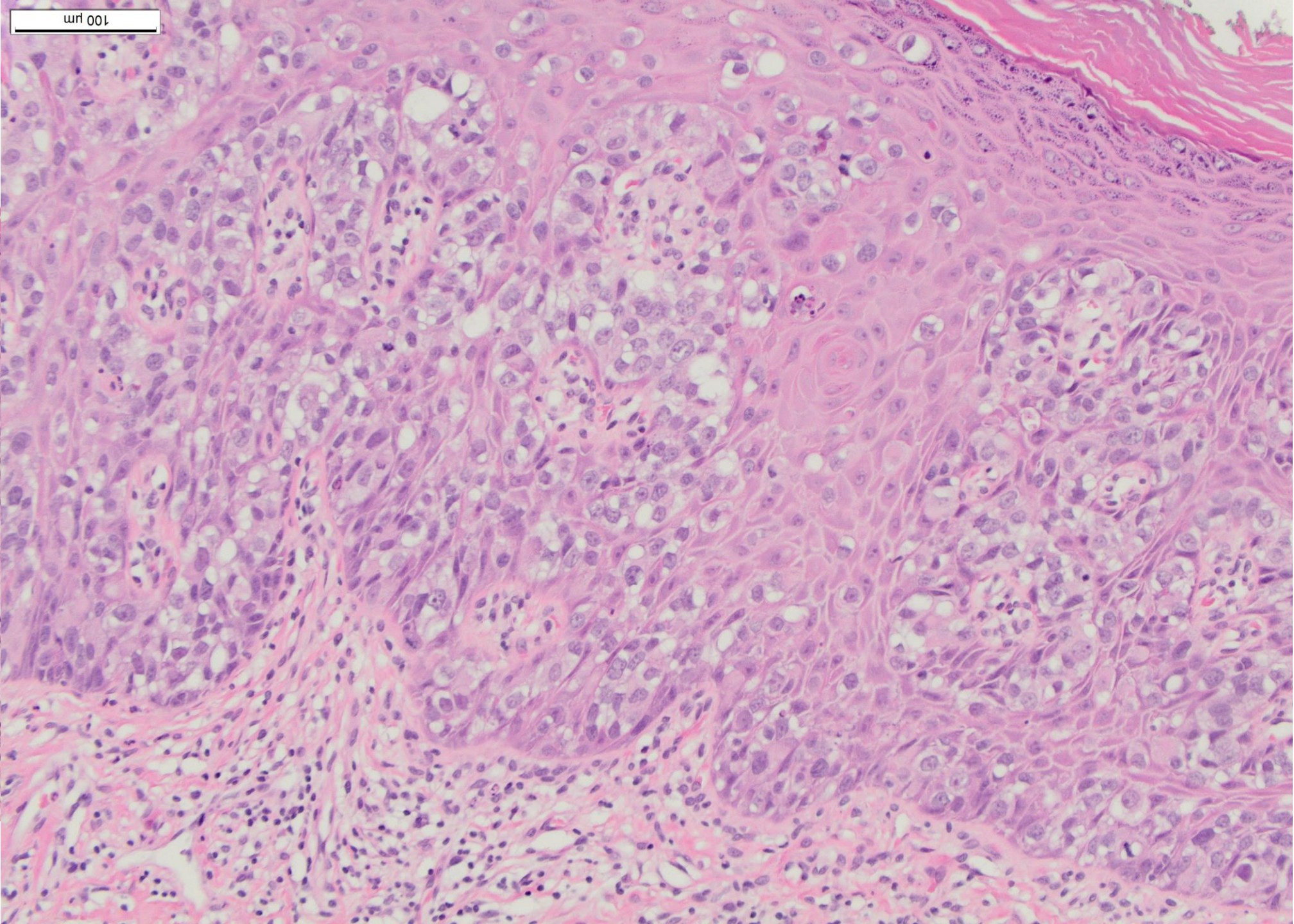




wrl 001



wrl 001



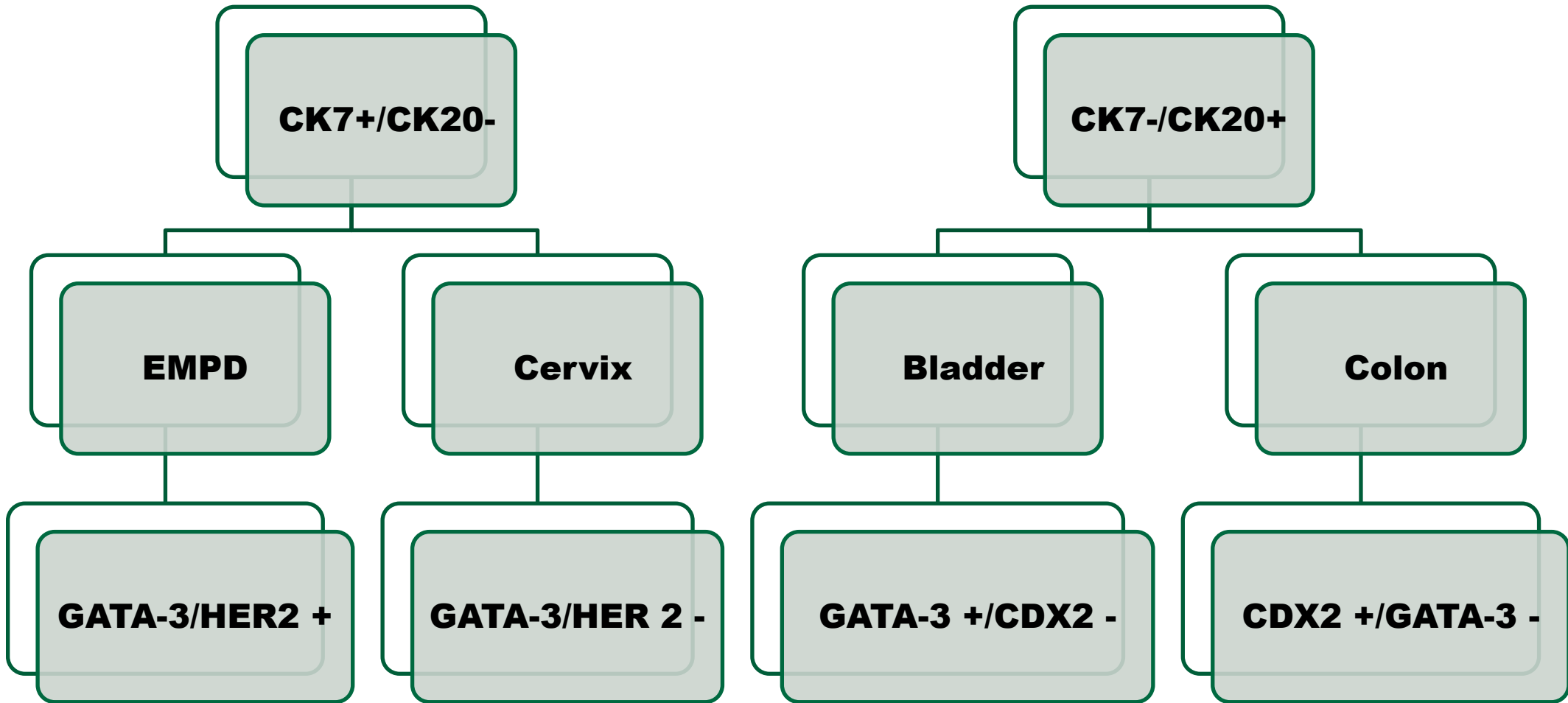


# Microscopy and differential

- Involving the epithelium of a related adnexal structure
- Isolated cells and nests at the basal and parabasal epithelial layers
  
- Toker Cell Hyperplasia
- Melanoma
- Underlying malignancy from colon, cervix, bladder

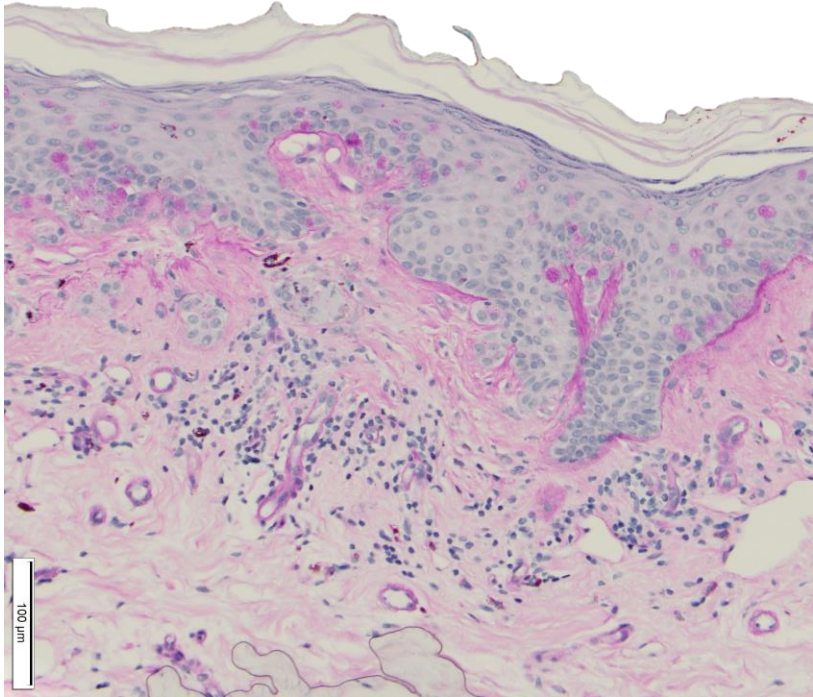


# Ancillary testing

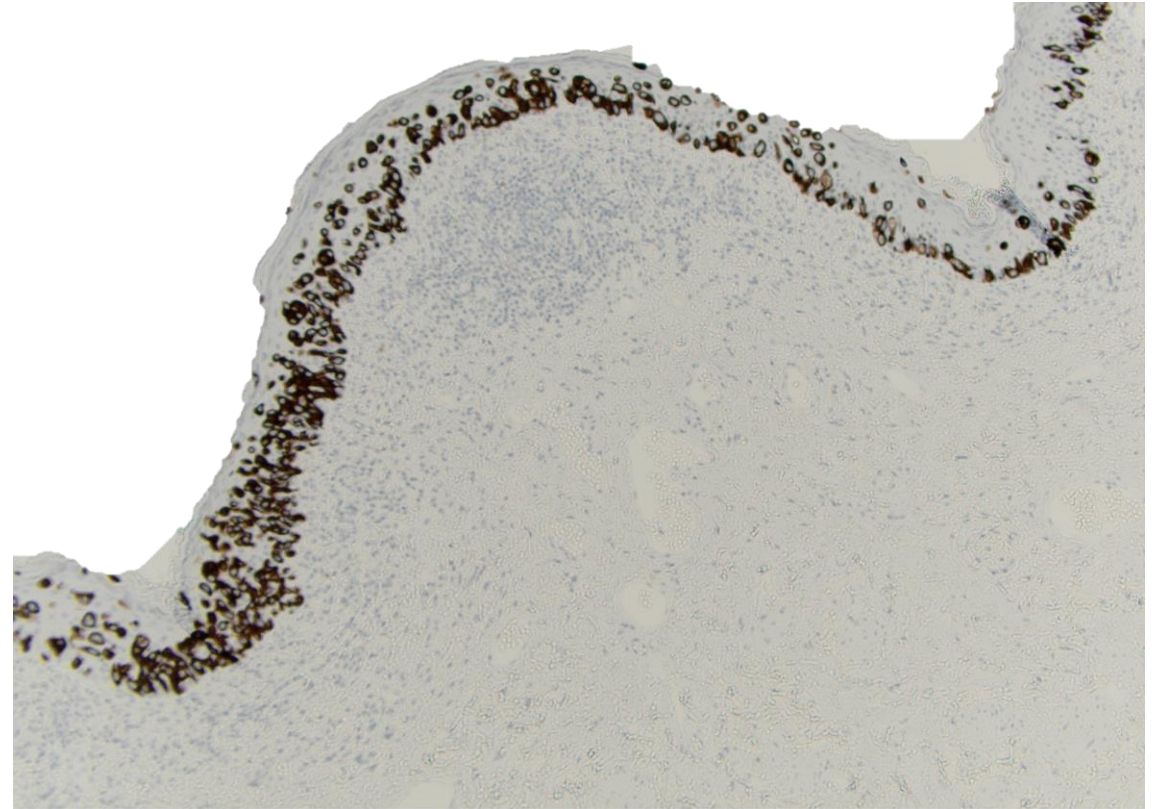




## PAS

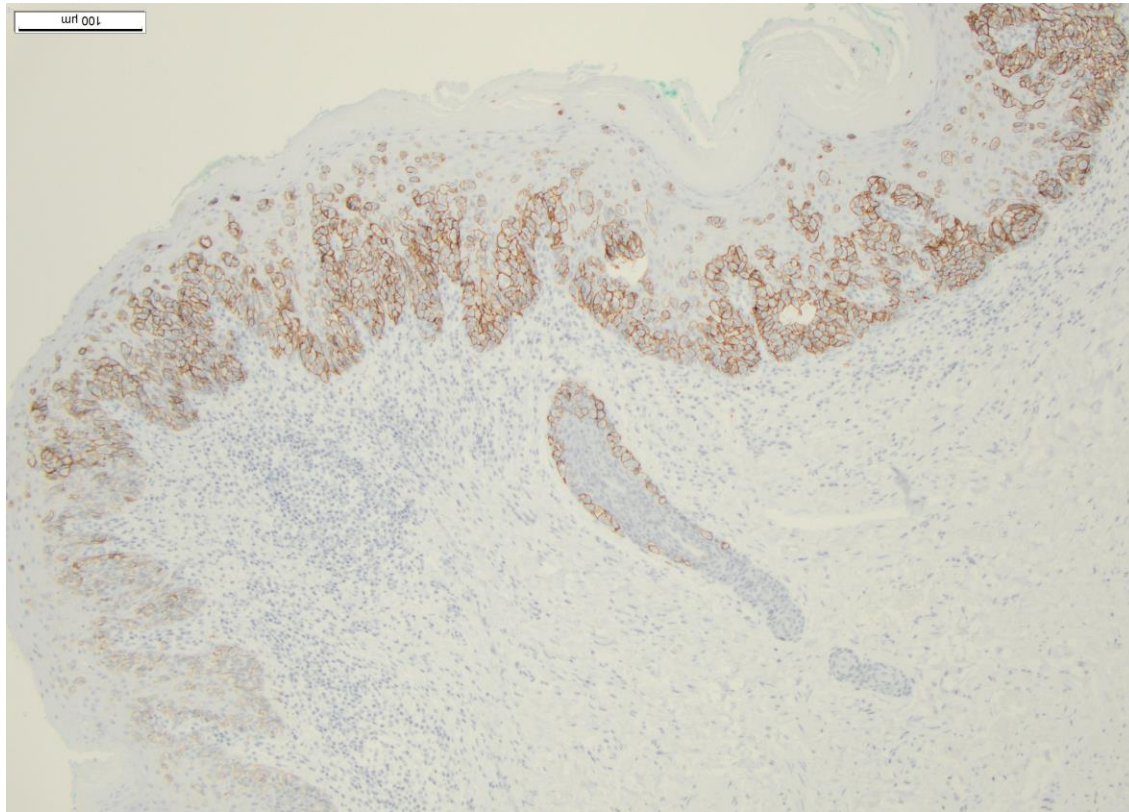


## CK7

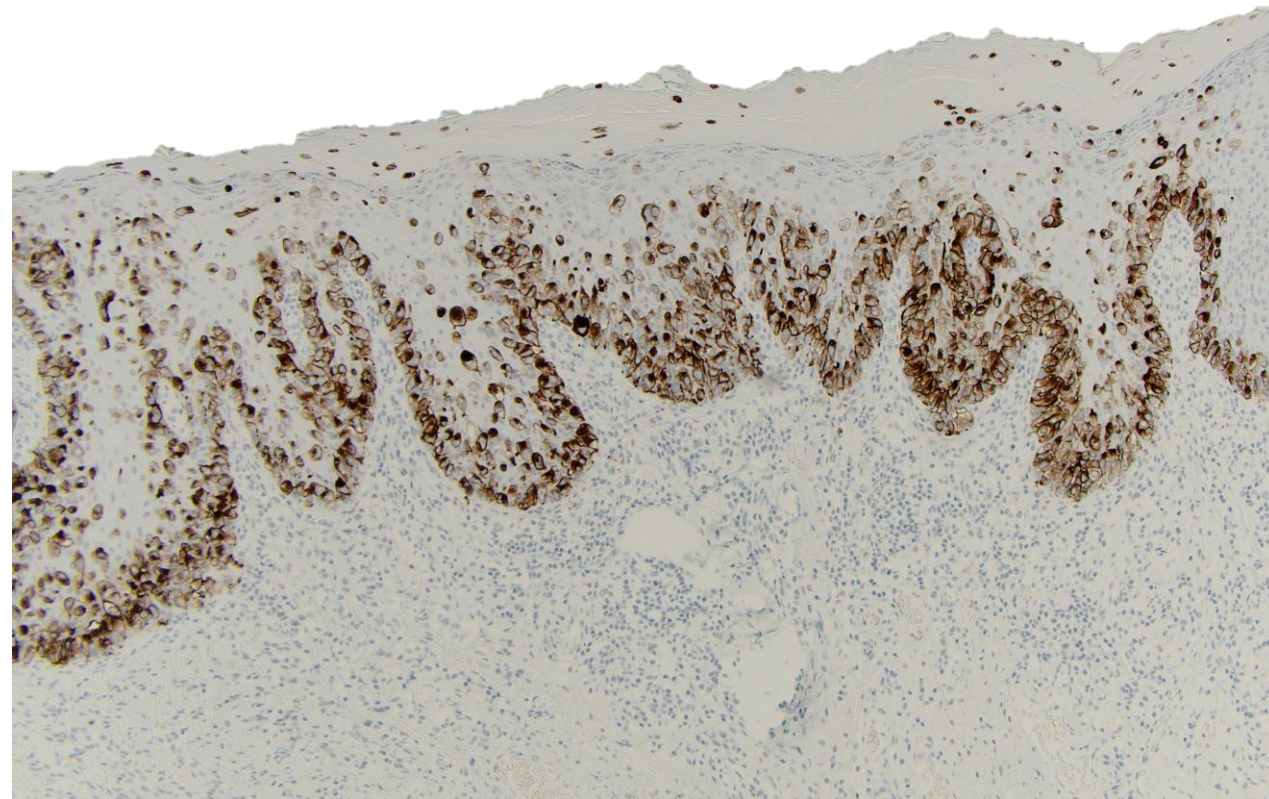




## Her2

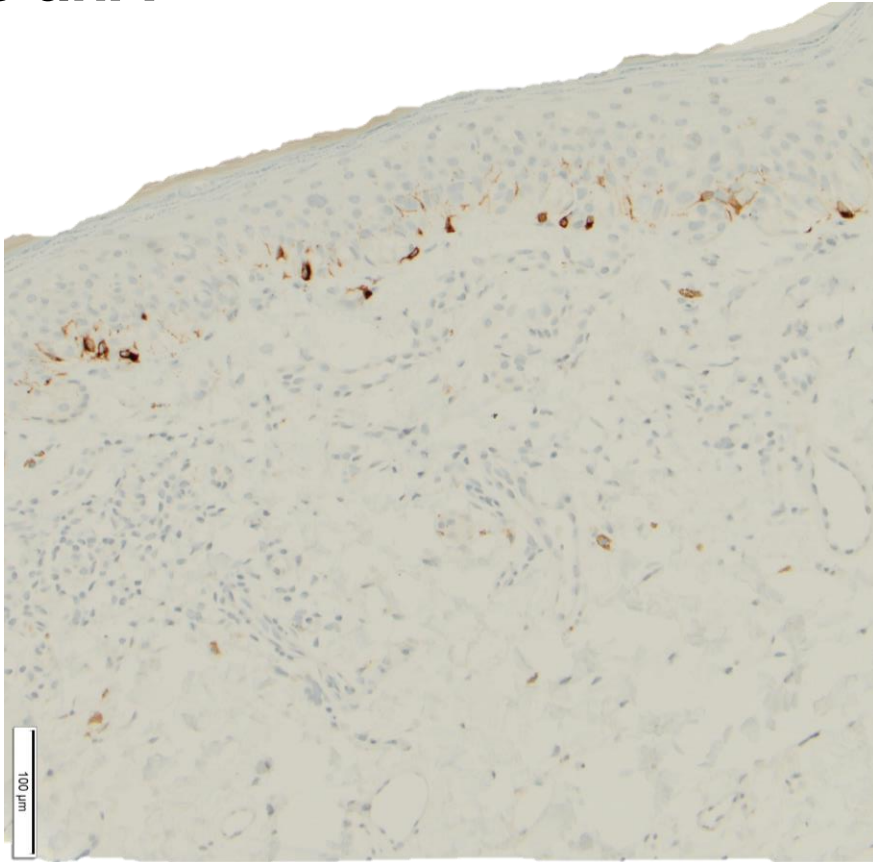


## mCEA

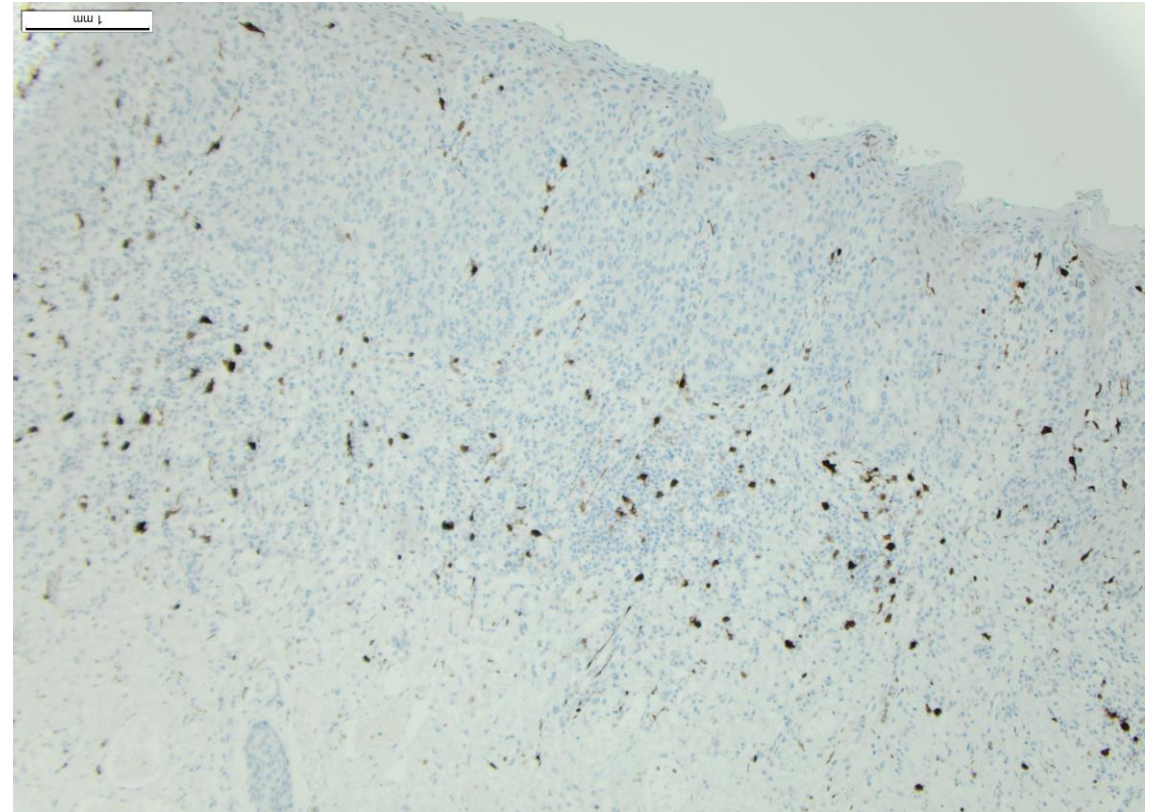




## MelanA



## S100





# Treatment

- Surgical Resection
  - Prognosis is good, but less so with even 1 mm invasion
  - Recurrences may occur, even with negative margin
  - Multi-focal field effect



# Pigmented

## Non-melanocytic

- Lichen Planus Pigmentosus Inversus (pigment incontinence)
- Cavernous hemangioma (blood)
- Pigmented apocrine hamartoma (melanin)

## Melanocytic

- Melanotic Macule
- Benign nevus of genital type
- melanoma

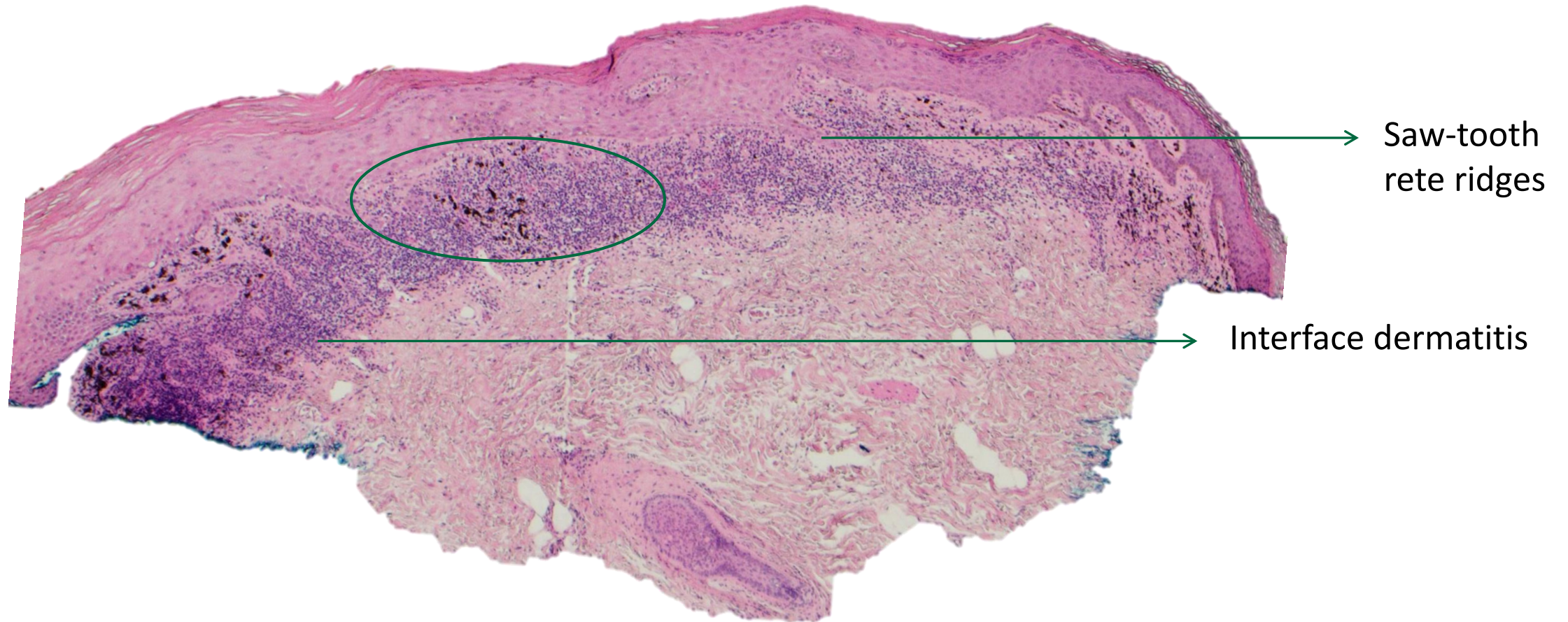


# 67 year old

- Itchy, grey/silver patches on the vulva?
- ?psoriasis

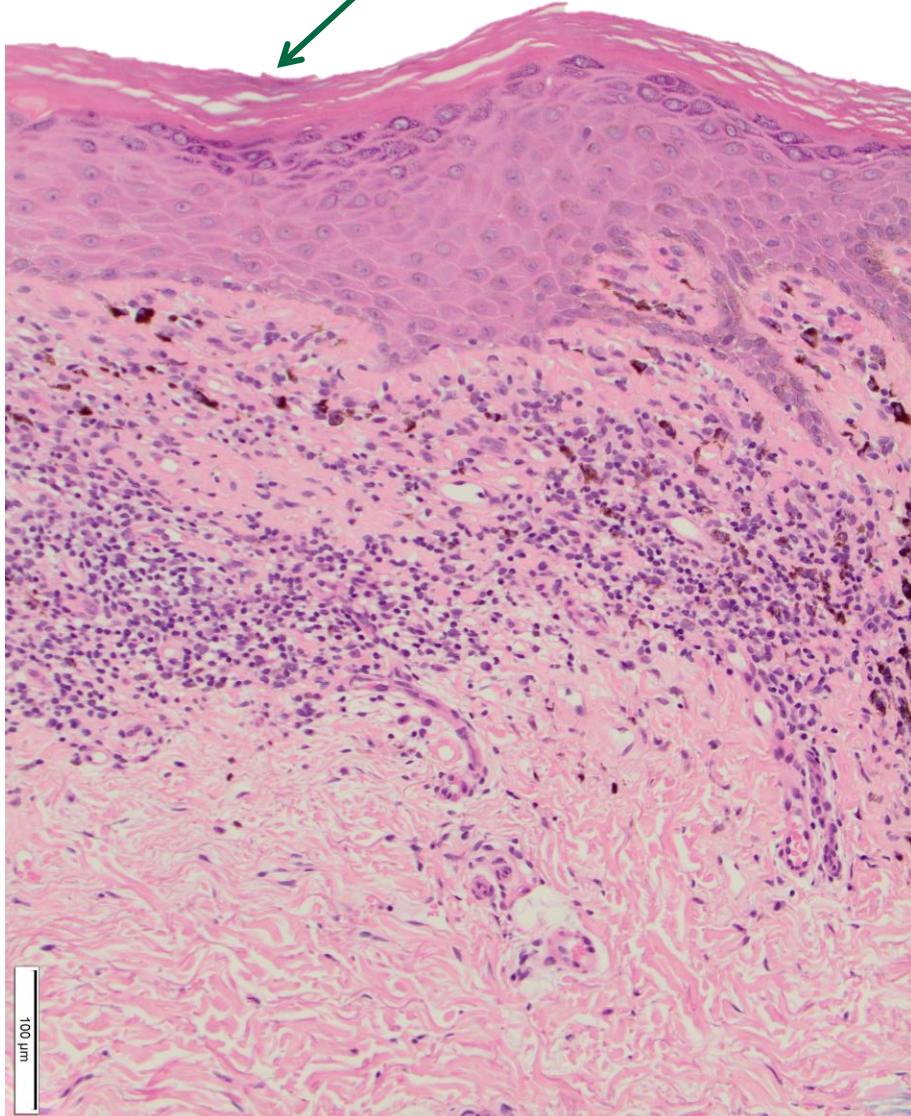


# Lichen Planus Pigmentosus Inversus



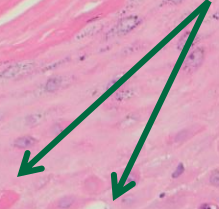


Wedge-shaped  
granulosis

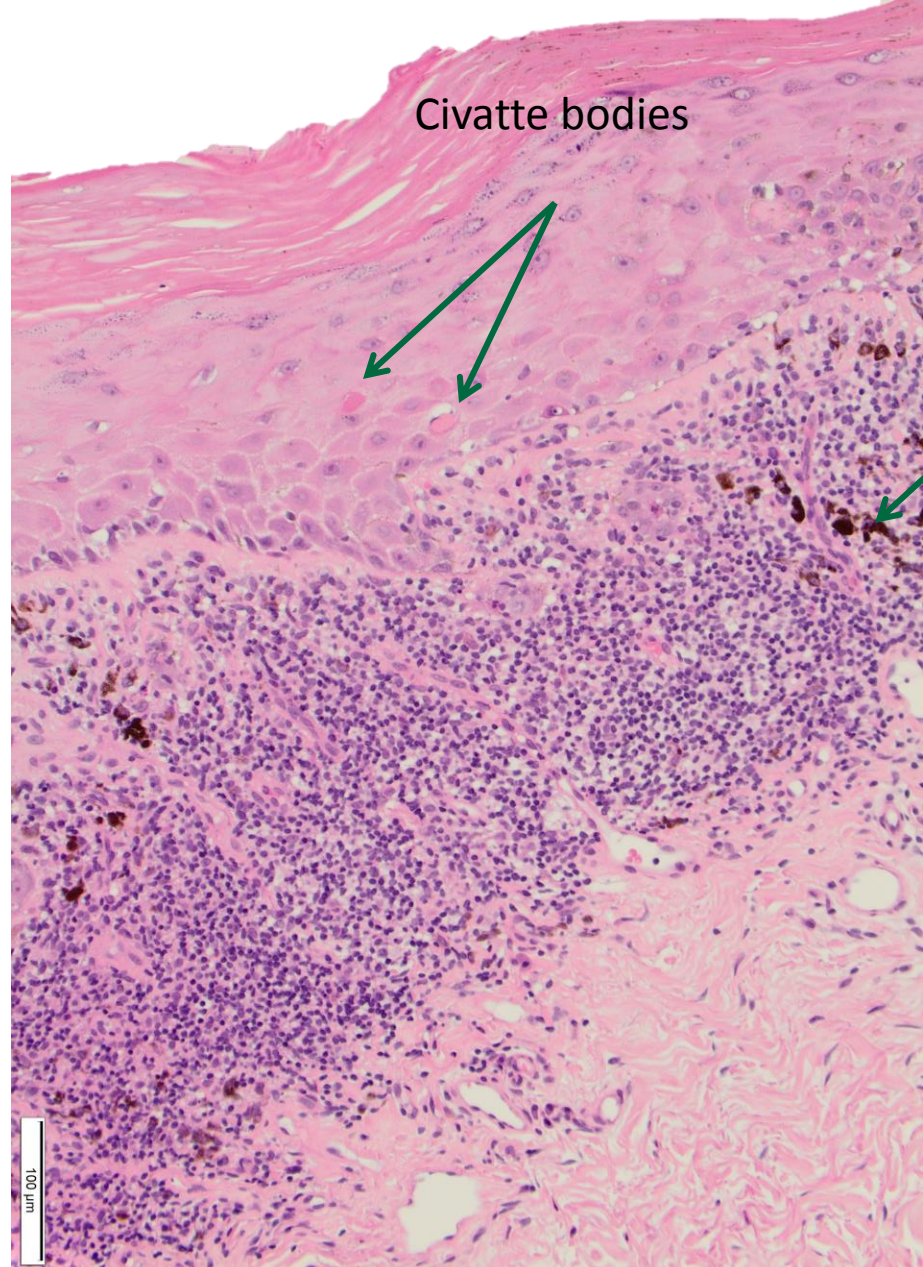
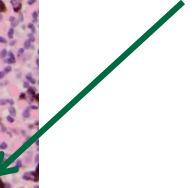


100µm

Civatte bodies



Dermal  
melanophages



100µm



# LPPI

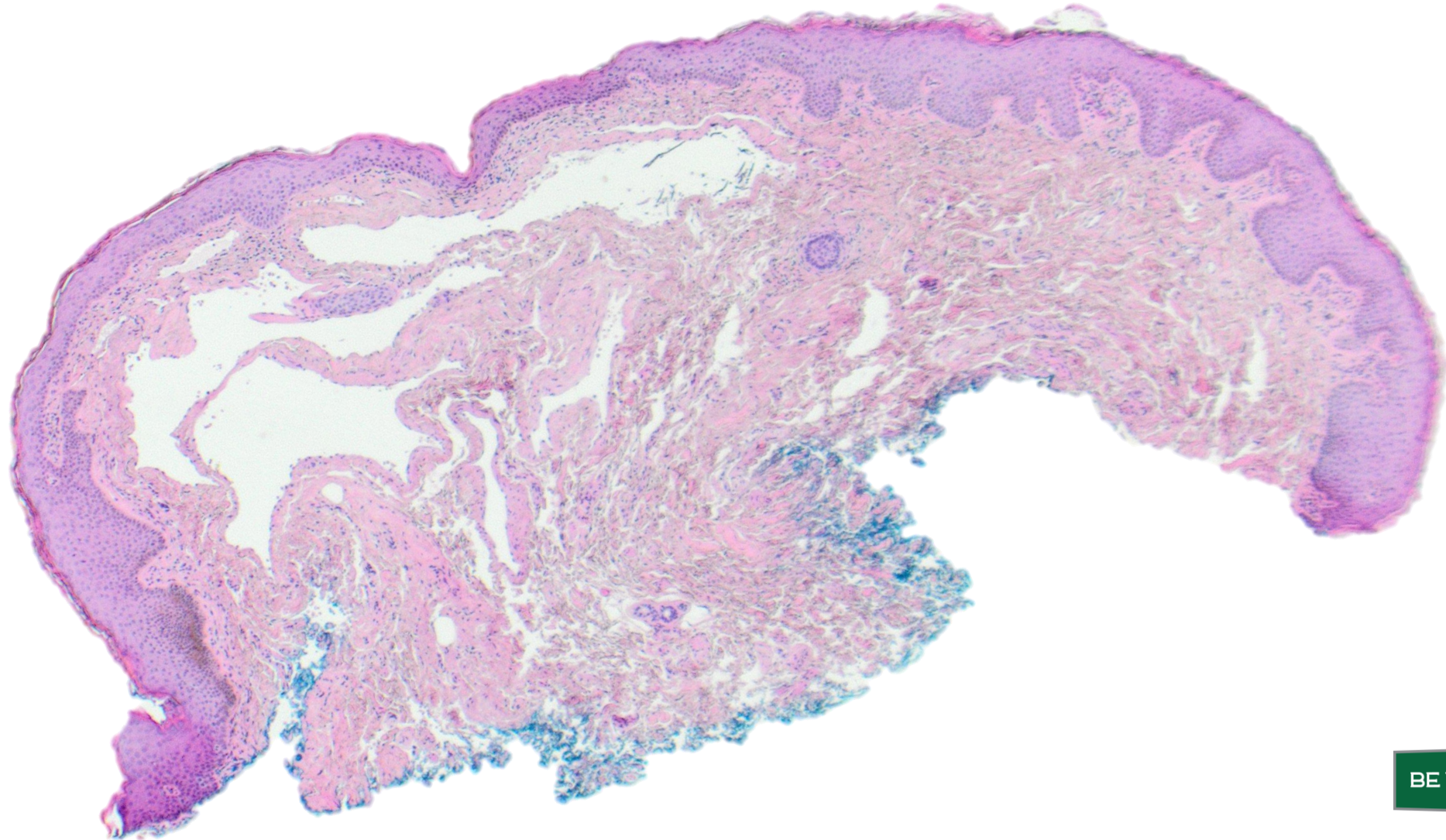
- rare variant of lichen planus pigmentosus
- typically affects sun protected areas
- rarely reported in the perineum/vulva
- Occasionally associated with lichen sclerosis
  
- Pigmented = dermal melanophages



# 77 year old

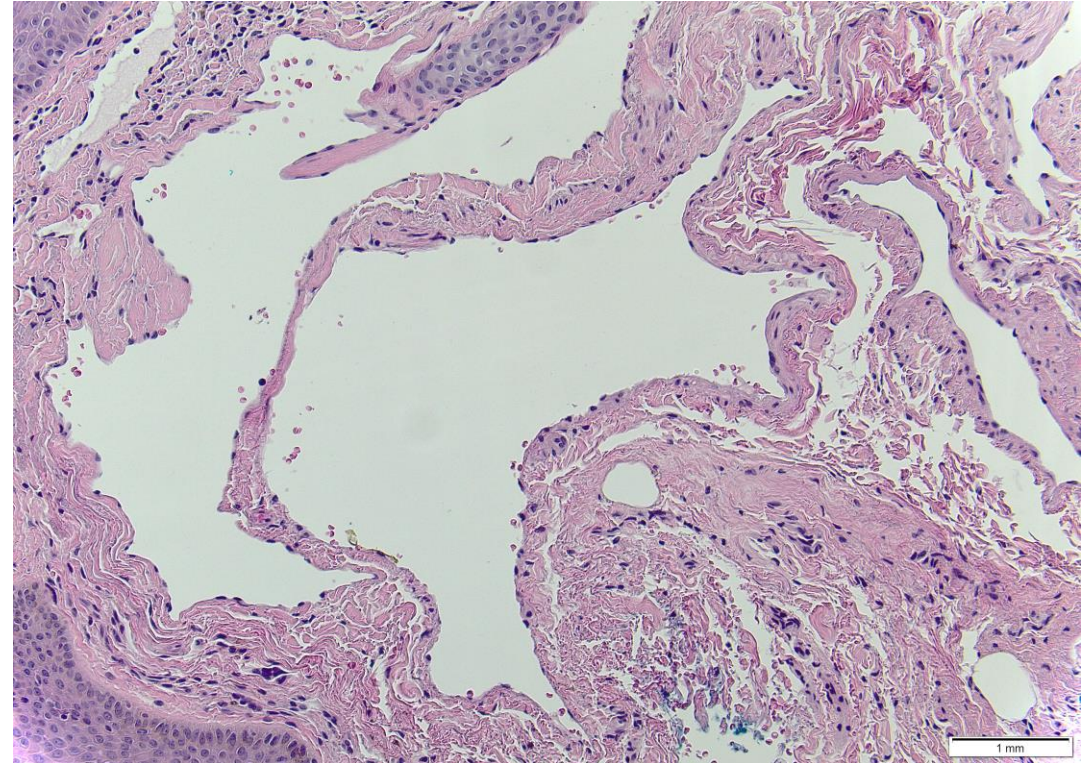
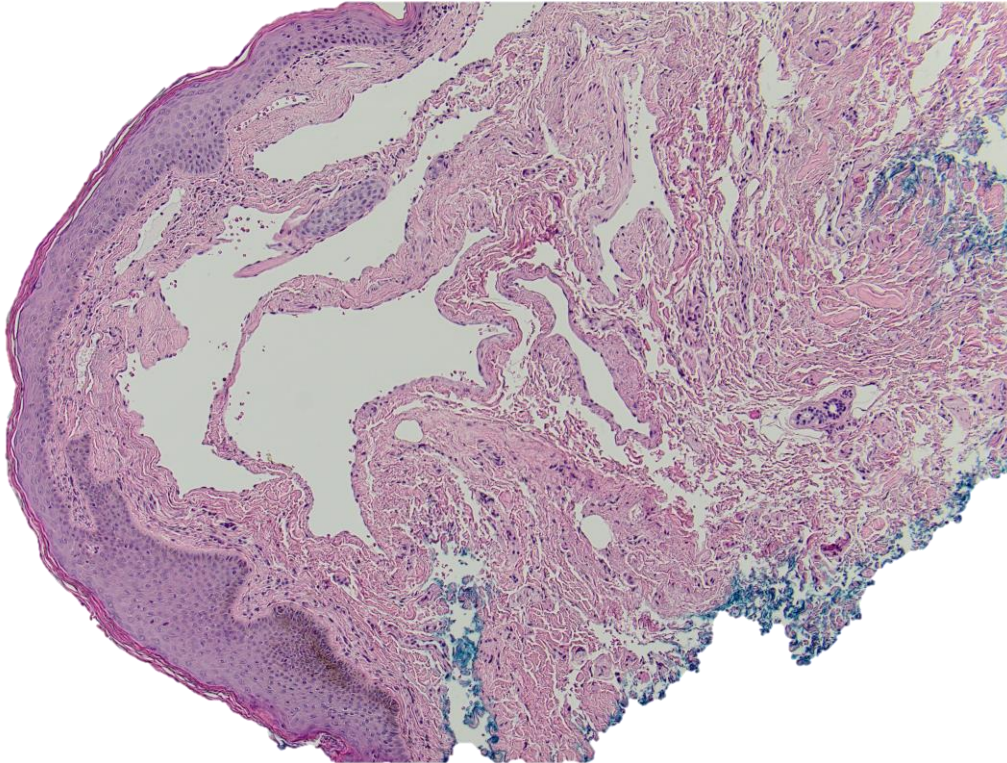
- Life-long lesion of dark spot on vulva







# Cavernous Hemangioma



Pigmented = blood

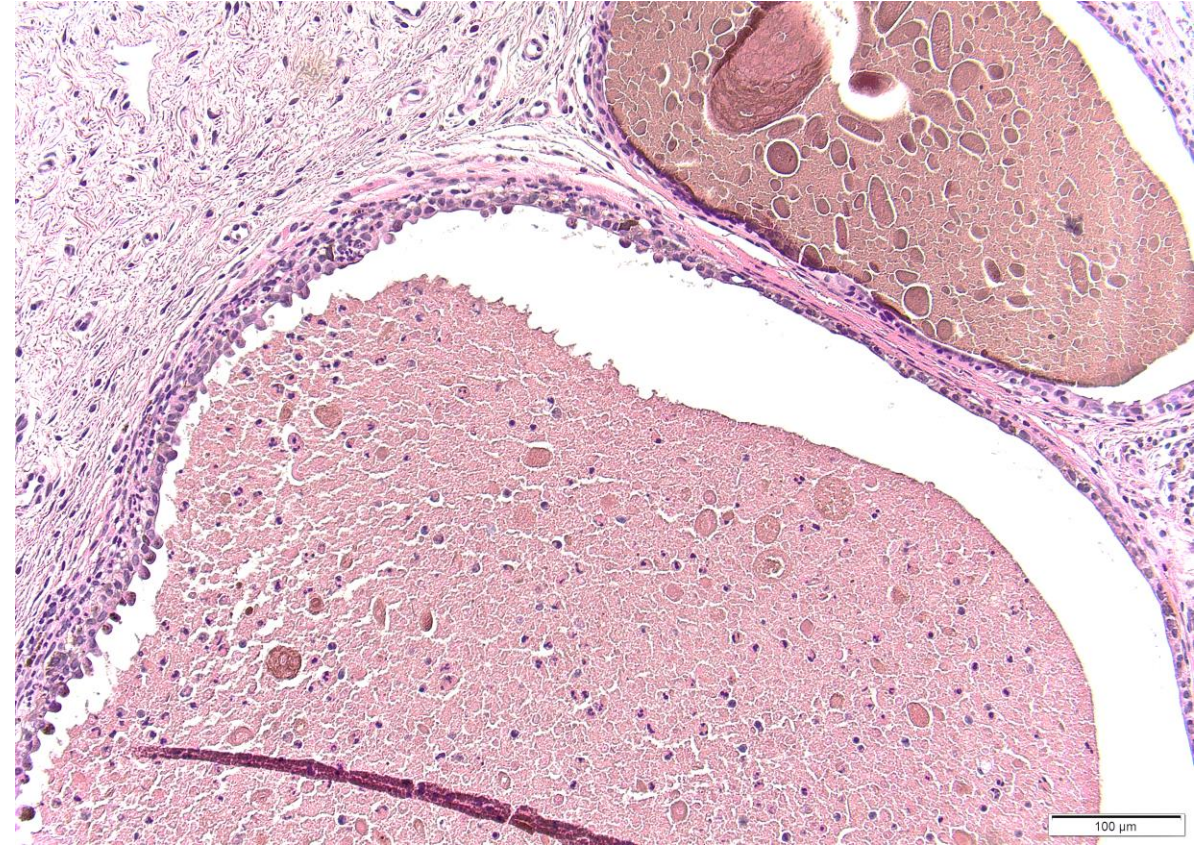
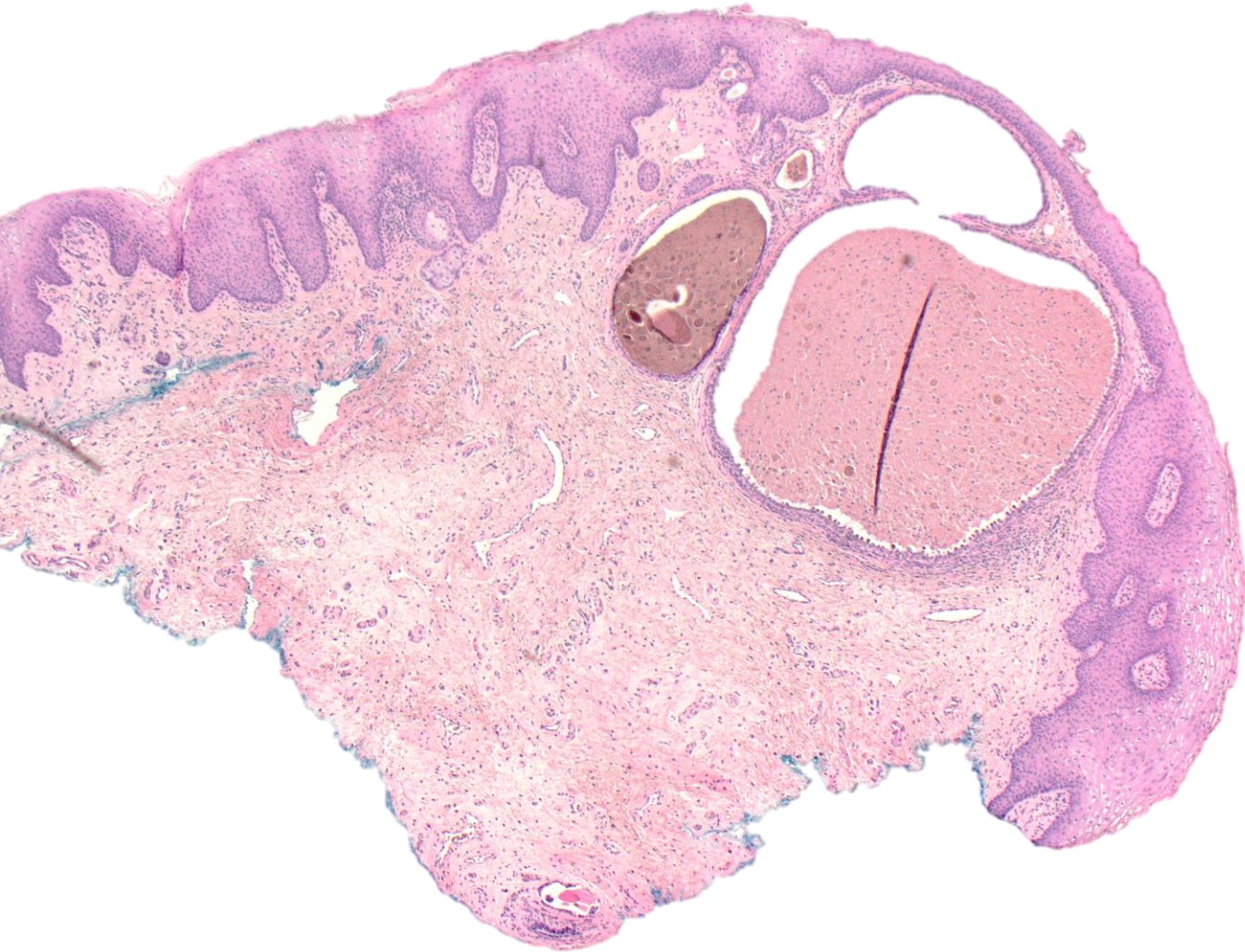


## 20 year old female

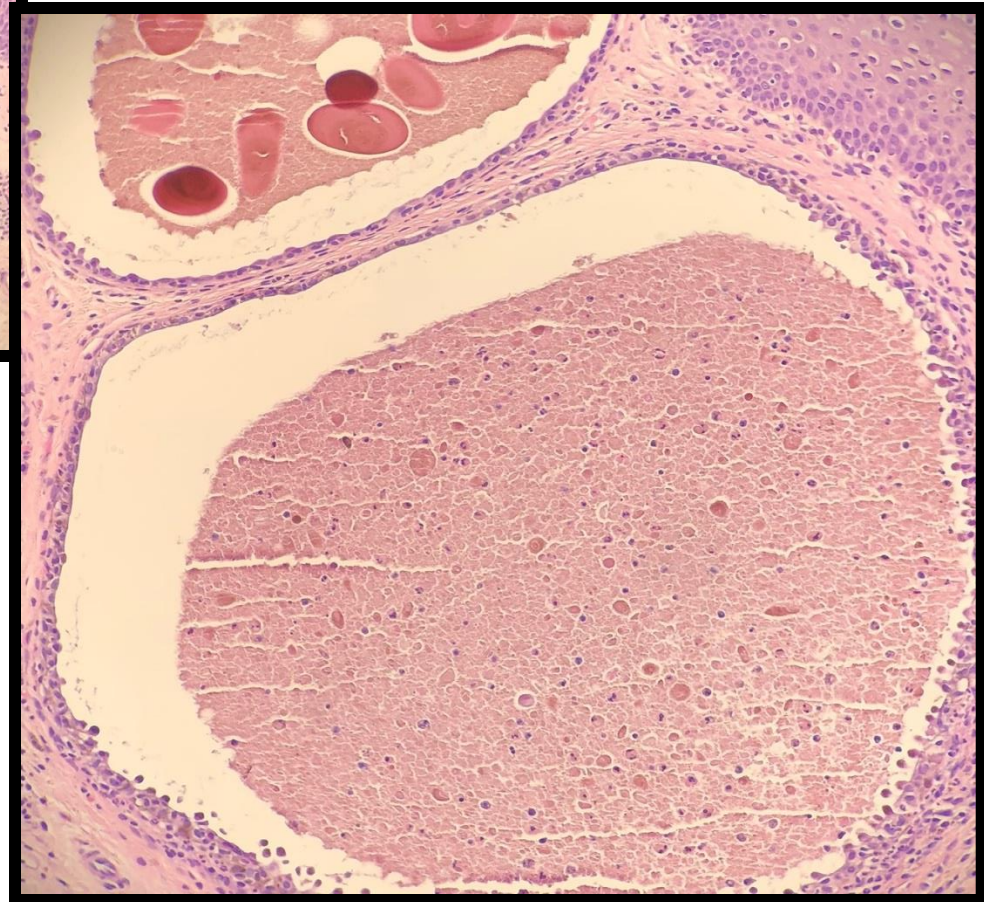
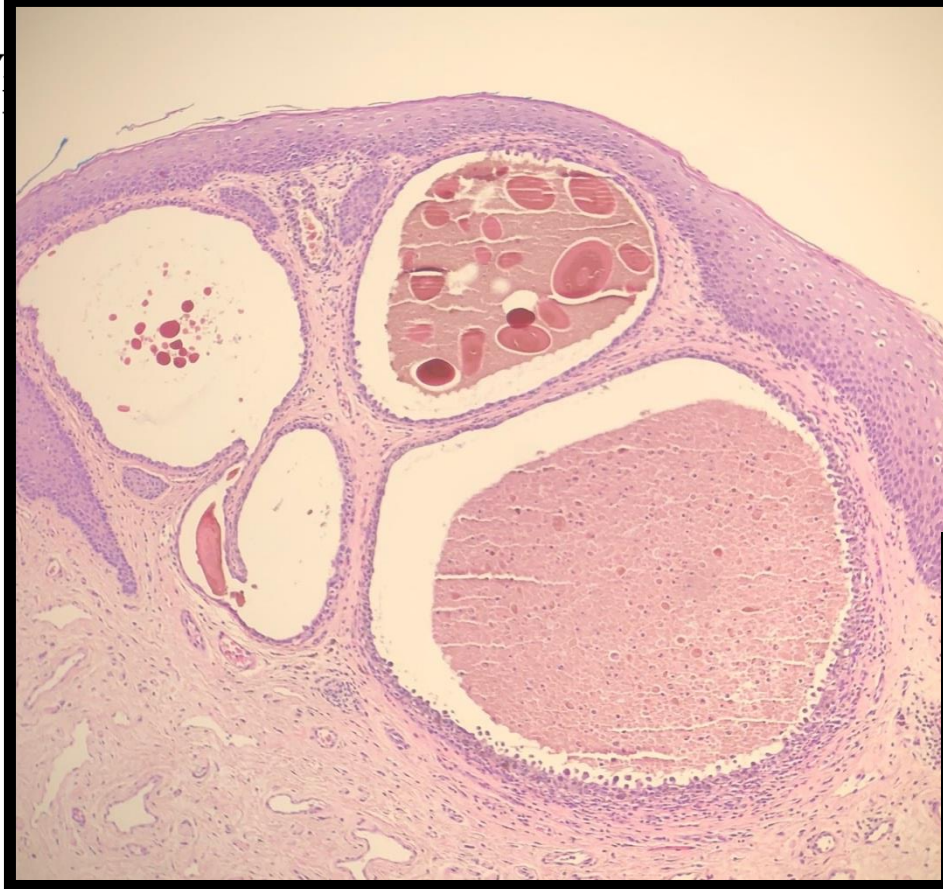
- Irregular-shaped small, dark mole on labium minus
- ? melanoma



# Pigmented Apocrine Hamartoma

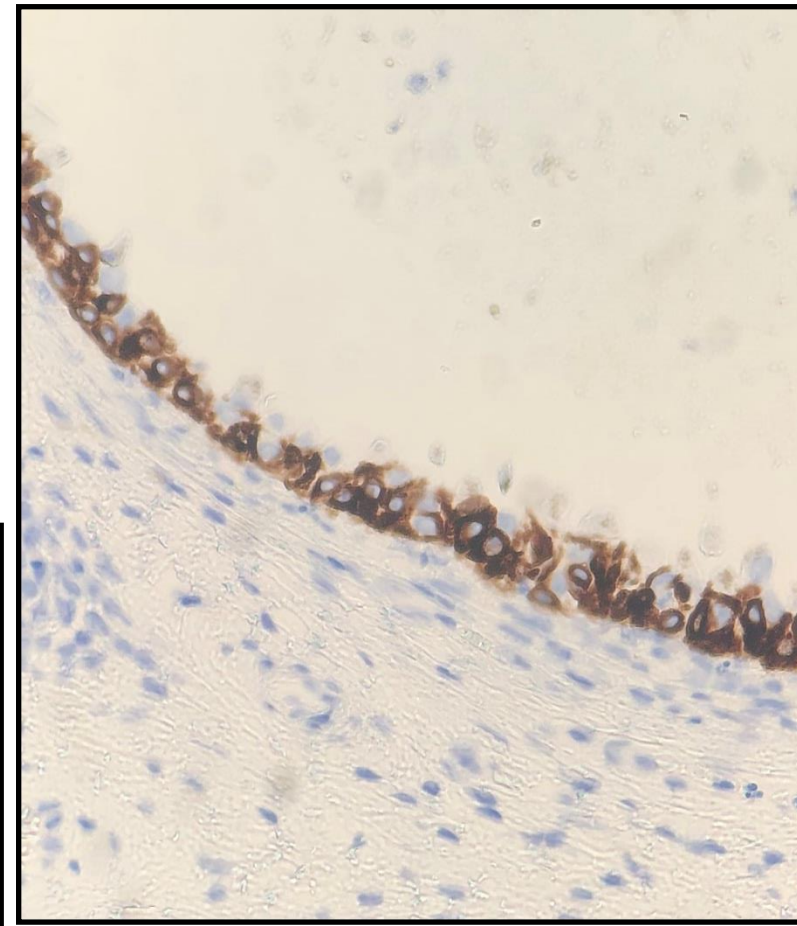
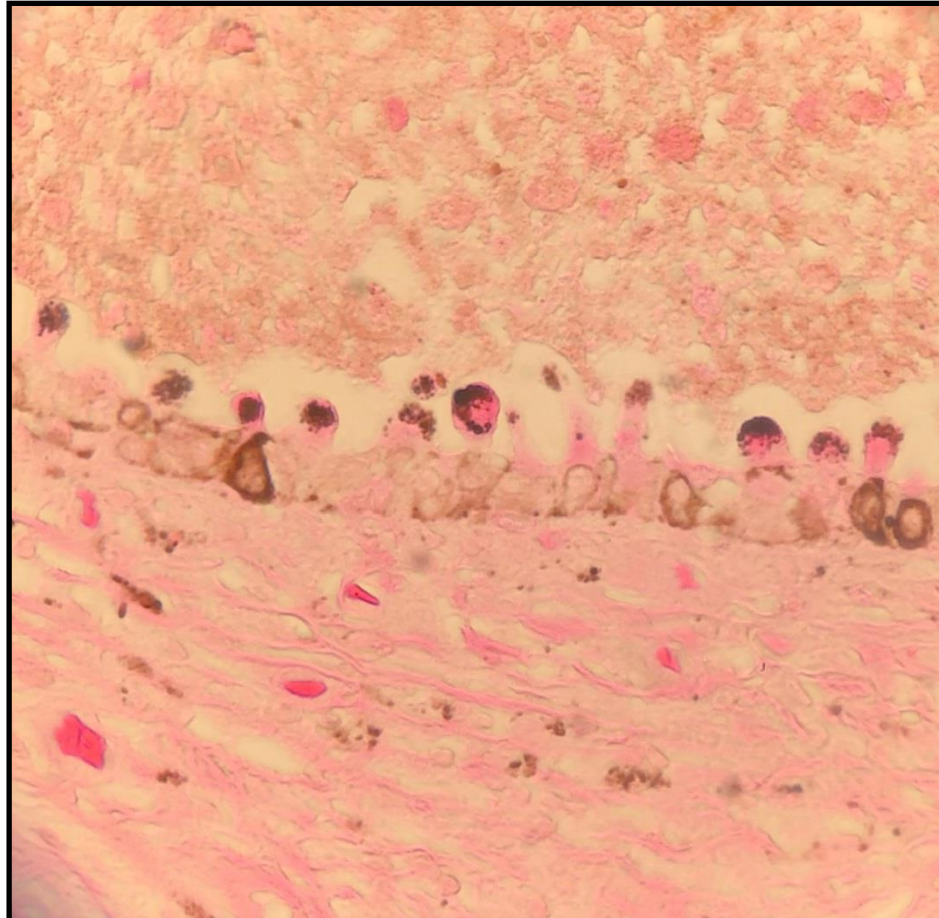
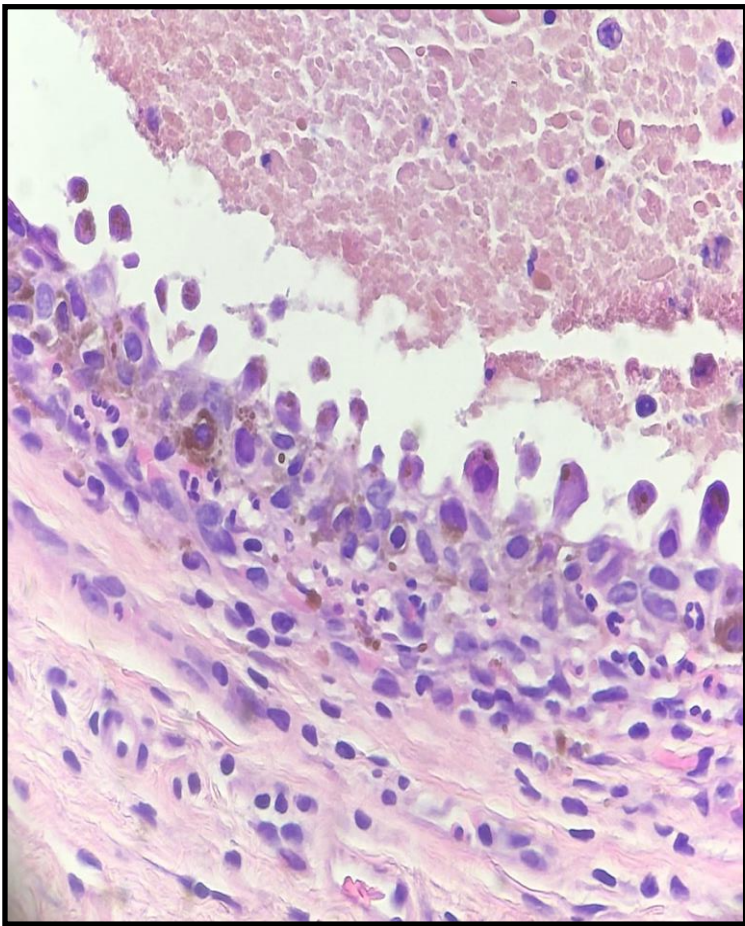






Courtesy of Dr. A. Osmond







## Case Report

# Pigmented Apocrine Hamartoma of the Vulva: A Report of Two Cases

Karl T. K. Chen, M.D.

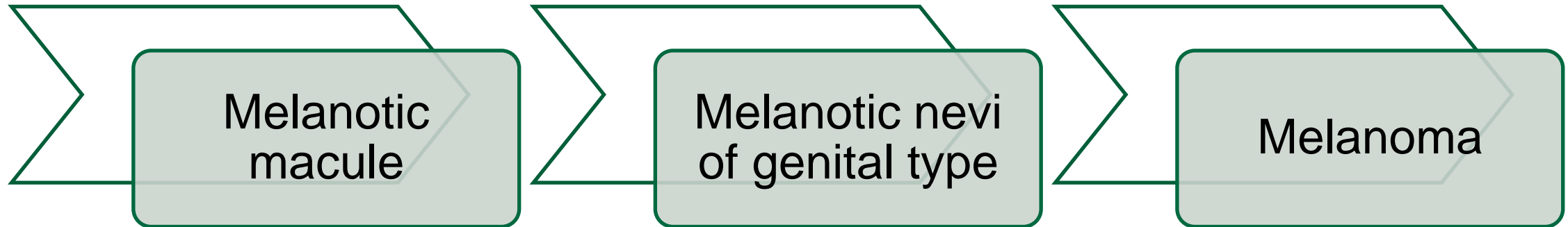
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**Summary:** Two cases of pigmented apocrine hamartoma of the vulva are reported; only one similar case involving the vulva was found in the literature. In each of the two cases, the lesion presented as a pigmented papule involving the labium minus of a young woman, and raised clinical concern for a malignant melanoma. On histological examination, the lesions were composed of tubules and cysts lined by melanin-containing apocrine cells and an outer layer of myoepithelial cells. The pigmentation appeared to be secondary to colonization of the apocrine cells by dendritic melanocytes. **Key Words:** Vulva—Apocrine tumor—Hamartoma—Melanin.

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# Melanocytic Lesions





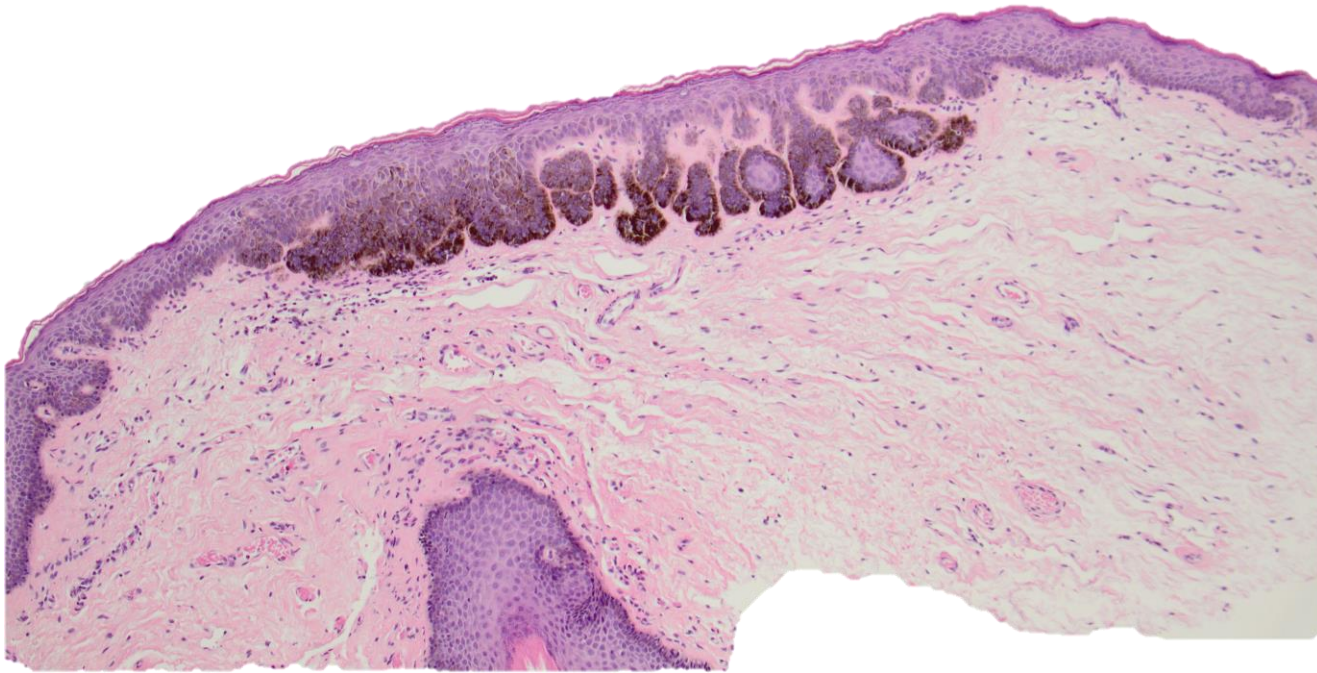
# Melanotic Macule

Definition: pigmented 'patch' on the mucosa

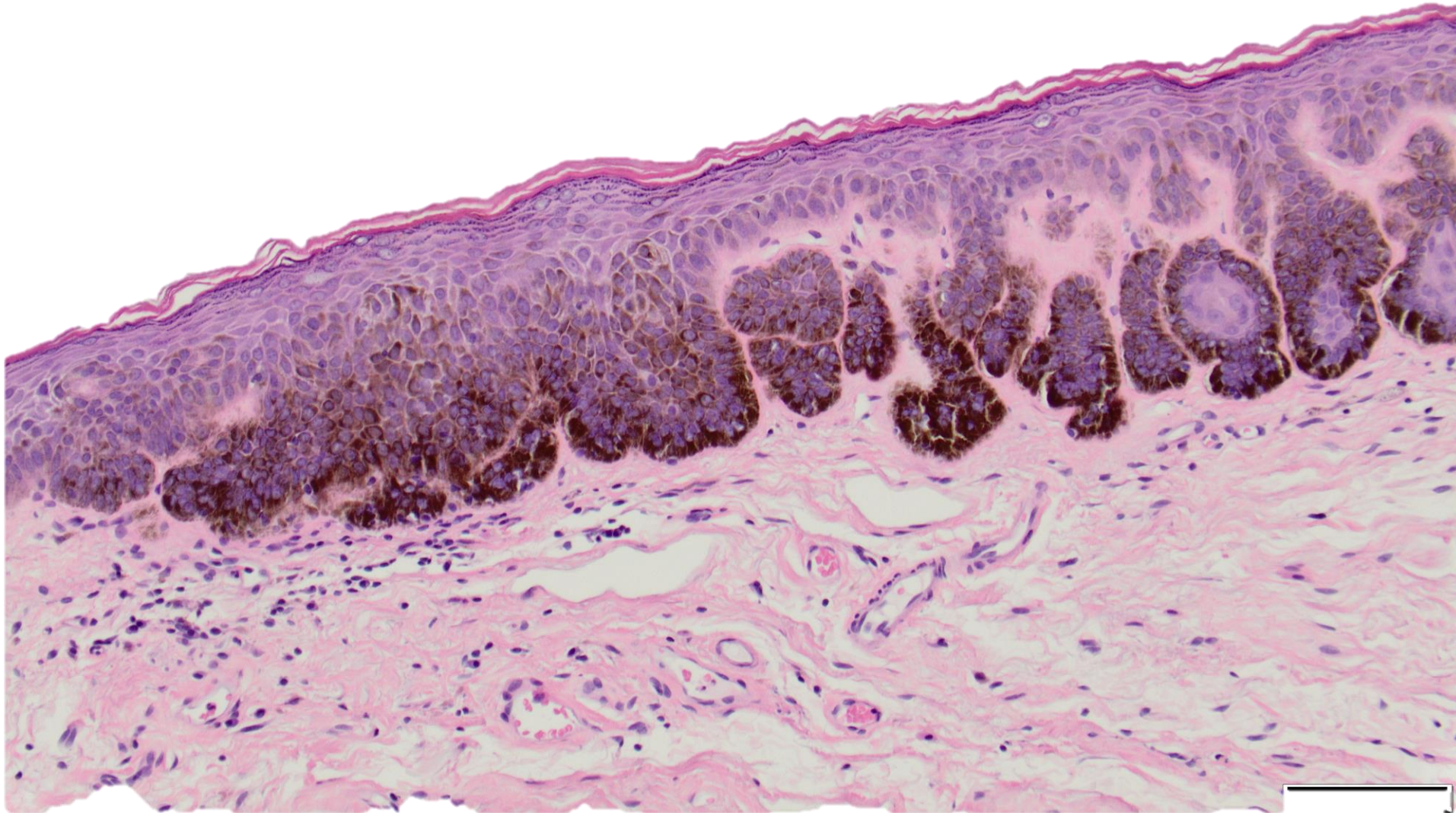
- Can be asymmetric, so cause for clinical concern



# 55 year old with pigmented lesion, previous HSIL (VIN3)



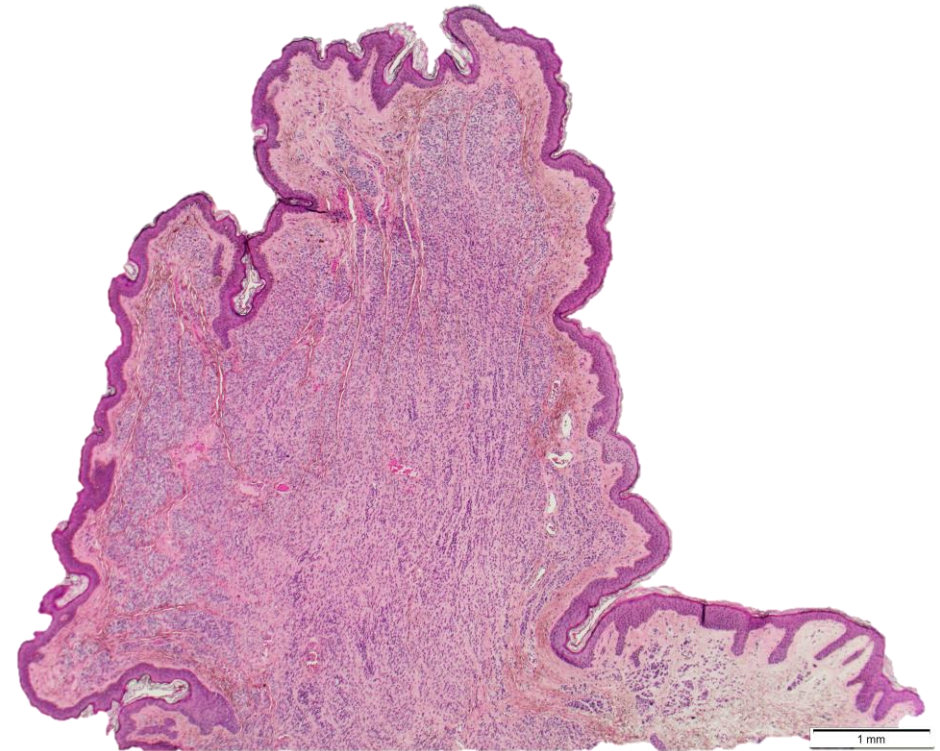
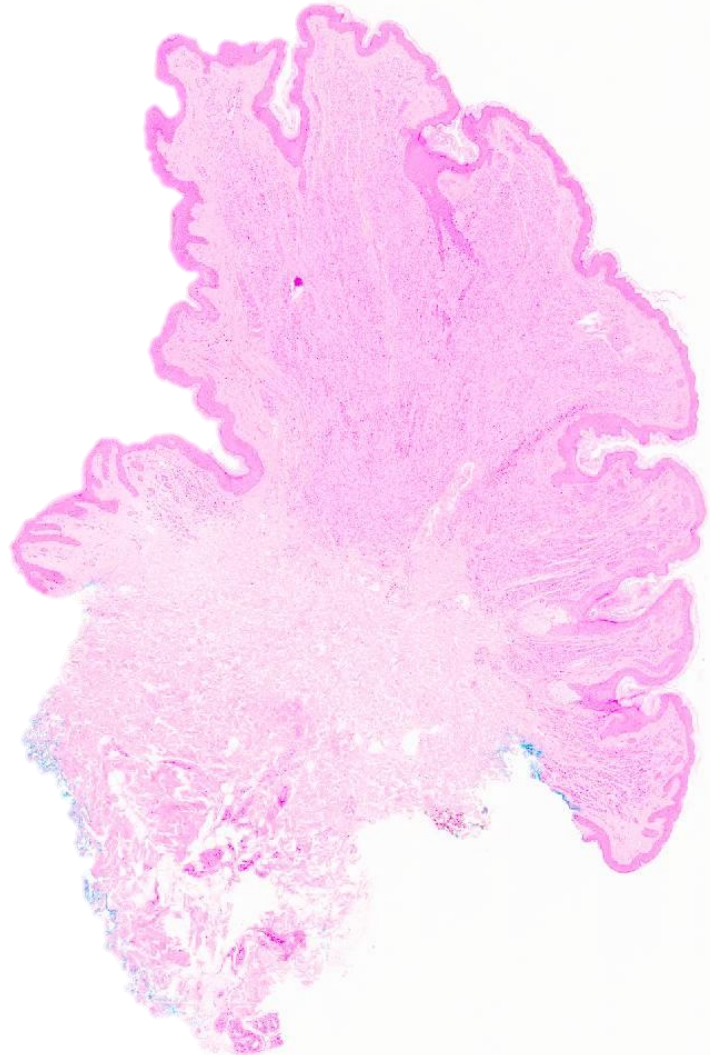
- ✓ Pigmentation
- ✓ Acanthosis
- ✓ No increase in melanocytes



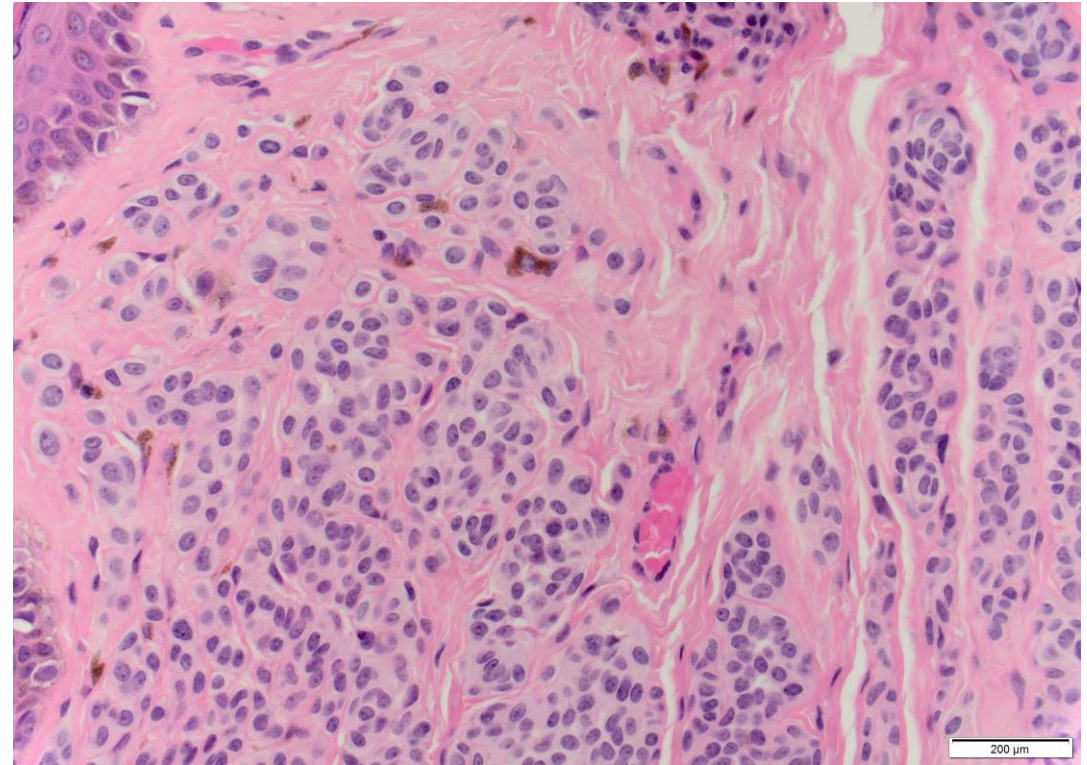
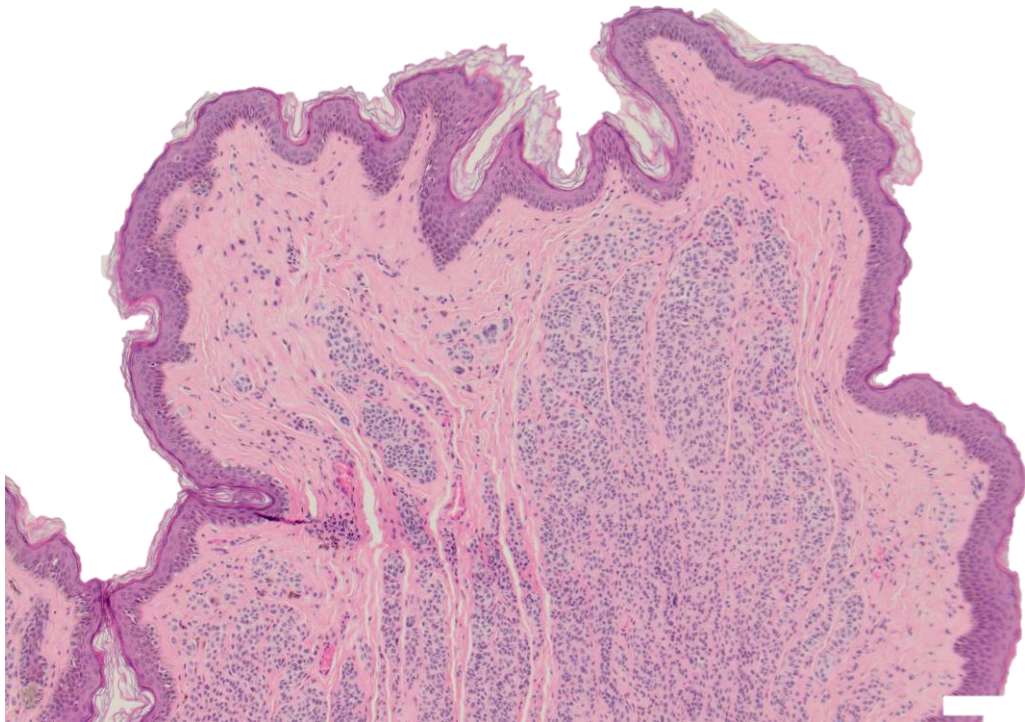


# Melanotic Nevi of Genital type

- Uncommon
- 2-3% of women
- Can sometimes have different features, which may be due to location and trauma/rubbing
- Important to recognize these special sites







# Vulvar Melanoma



## Definition:

- Malignant melanocytic neoplasm arising on the external genitalia
- Either hair-bearing skin or mucosa

## Incidence:

3% of melanomas

Cutaneous skin:vulva ratio of 71:1



# Patient demographics

- Older than patients with melanomas in sun-exposed areas
- Sixth-seventh decades
- Mostly Caucasian women
- Late presentation due to various reasons

# Anatomy affected



# Gross Description

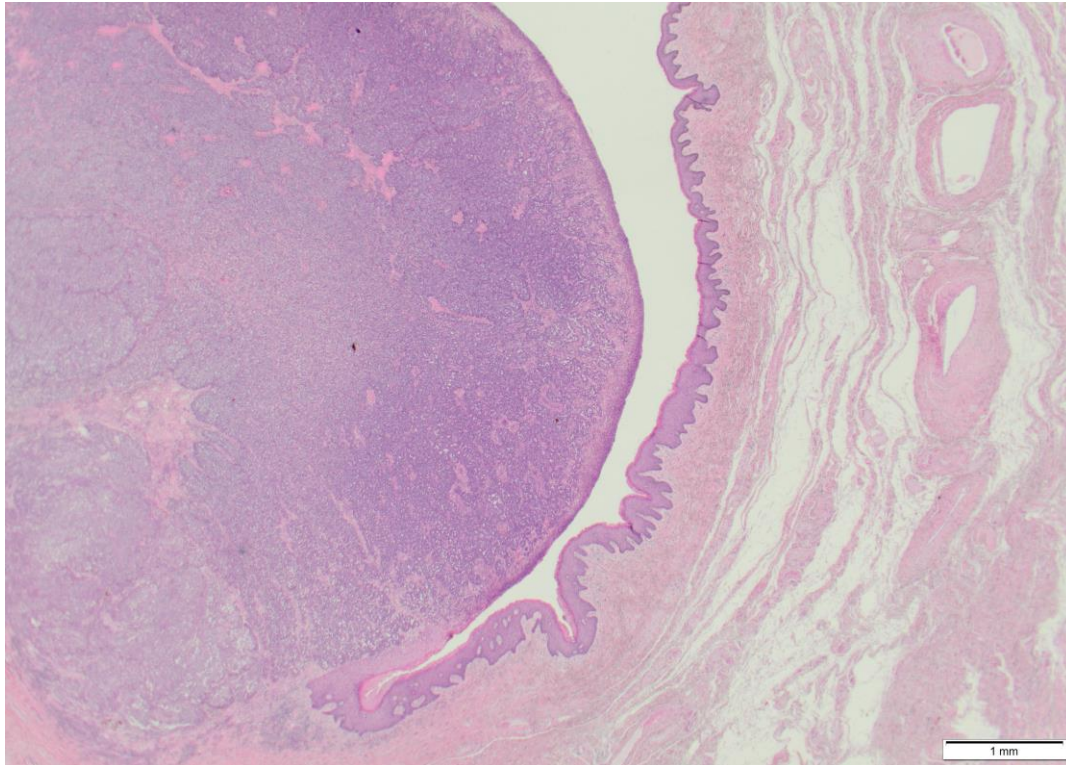




# Microscopic Findings

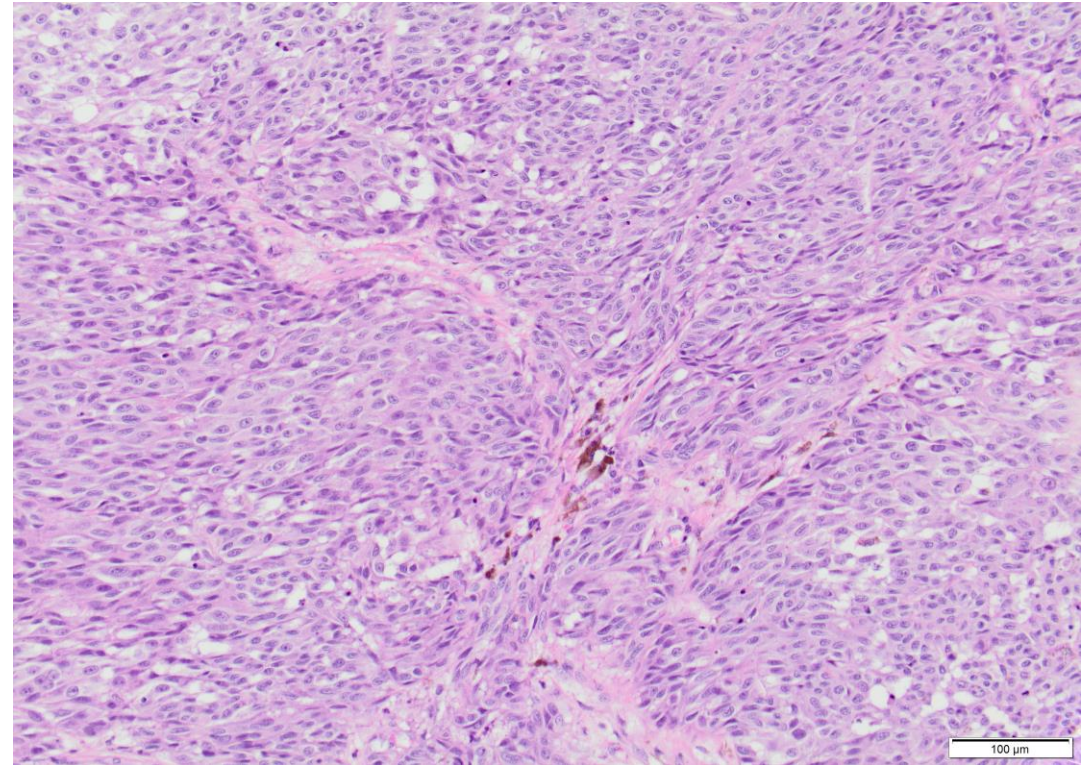
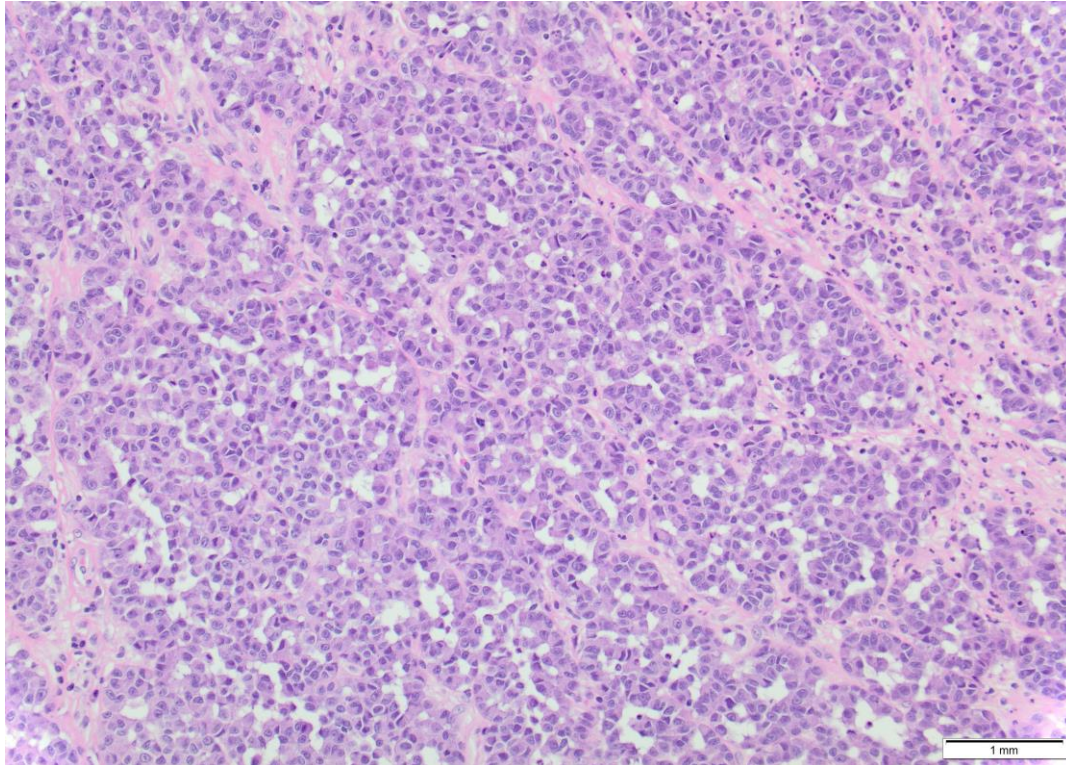
- Historically similar to acral lentiginous melanoma
  
- Mucosal > Superficial Spreading > Nodular
  
- Mucosal
  - Diffuse proliferation of large, epithelioid cells in nests along basal layer
  - Pagetoid spread common
  
- Nodular
  - Lacks radial growth phase
  - Ulceration common

# Melanoma





# Microscopy Patterns



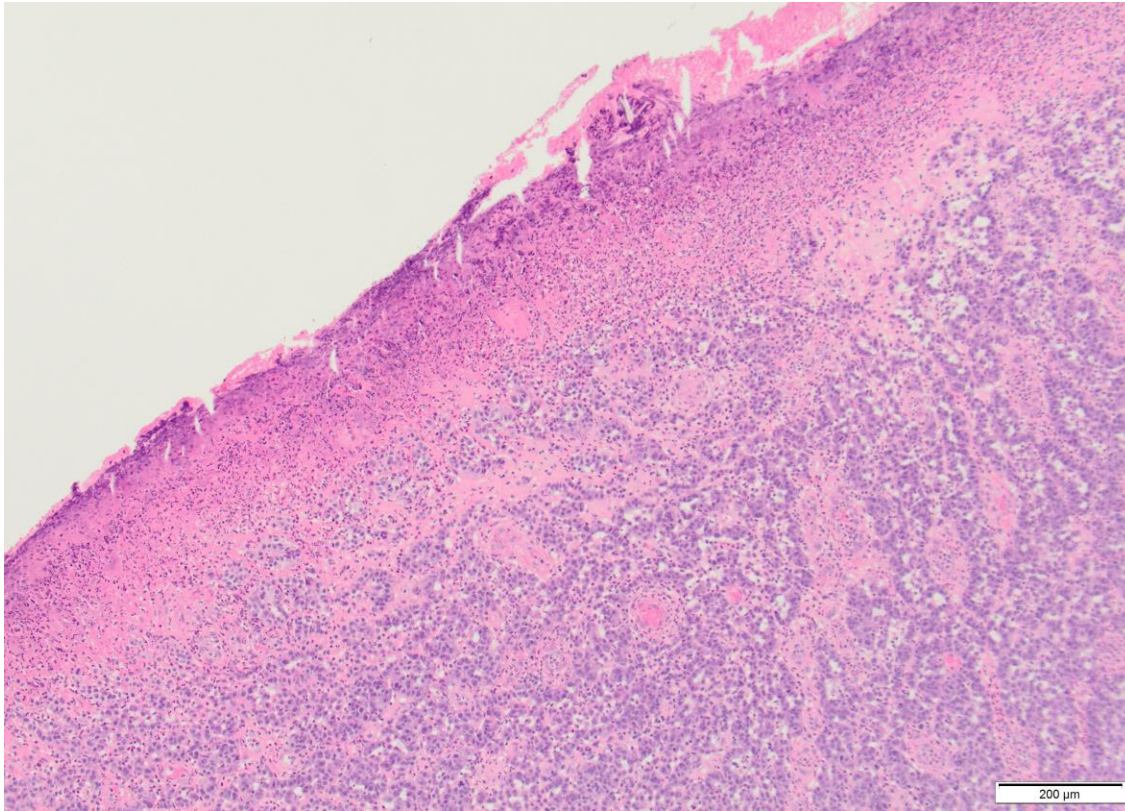
# Main Histologic Predictor in VMM

- Tumor thickness (Breslow)
  - Top of the granular cell layer
  - What if mucosa and no granular cell layer
  - Bottom of the ulceration base

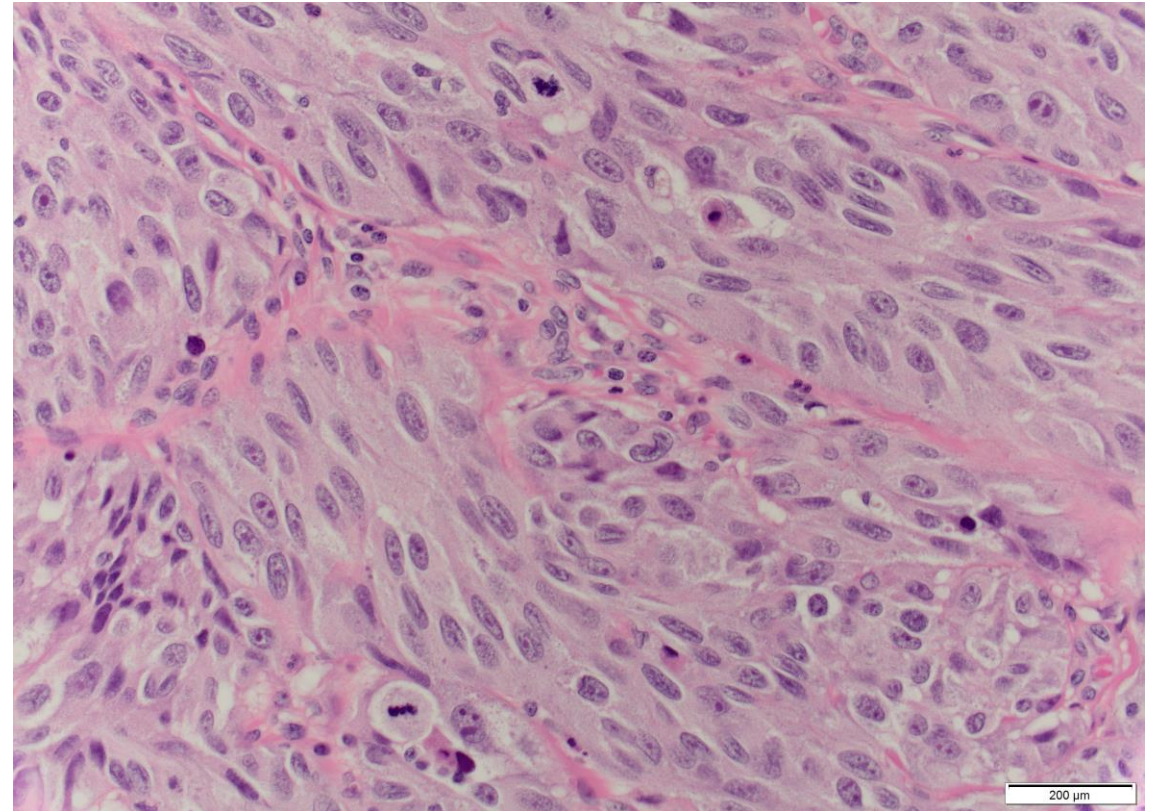


# Features reported in VMM similar to cutaneous

## Ulceration

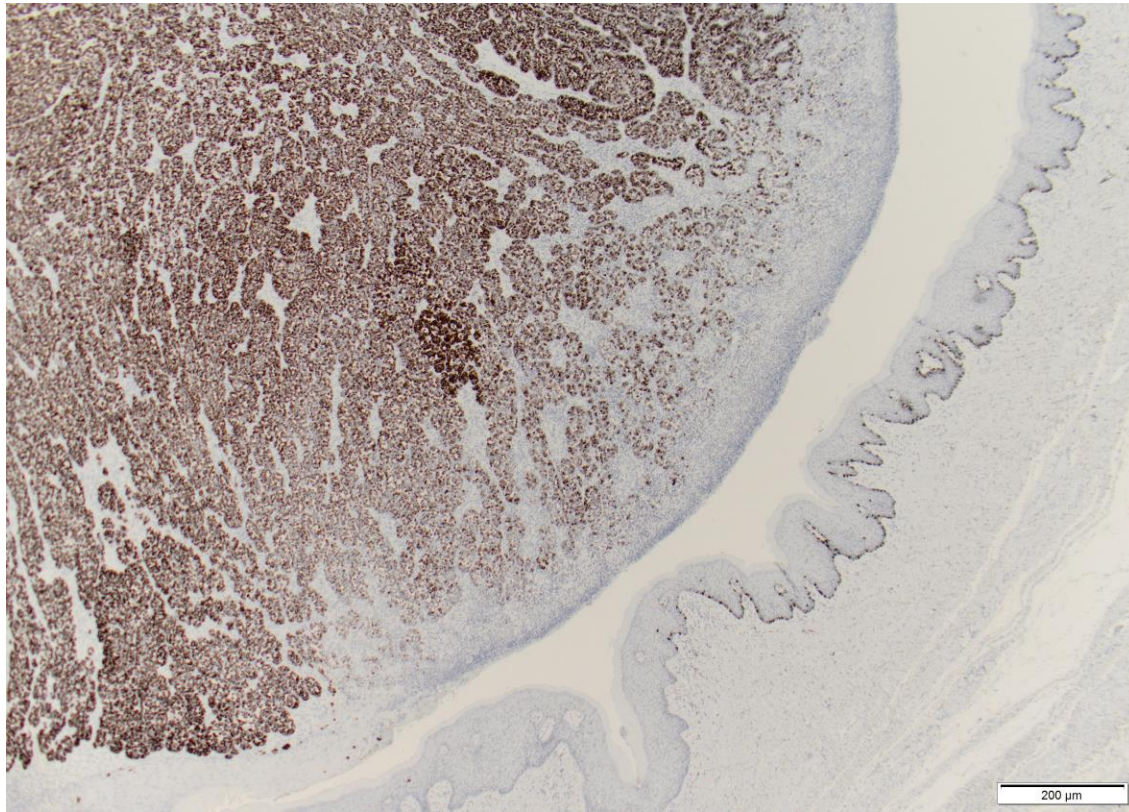


## Mitotic figures

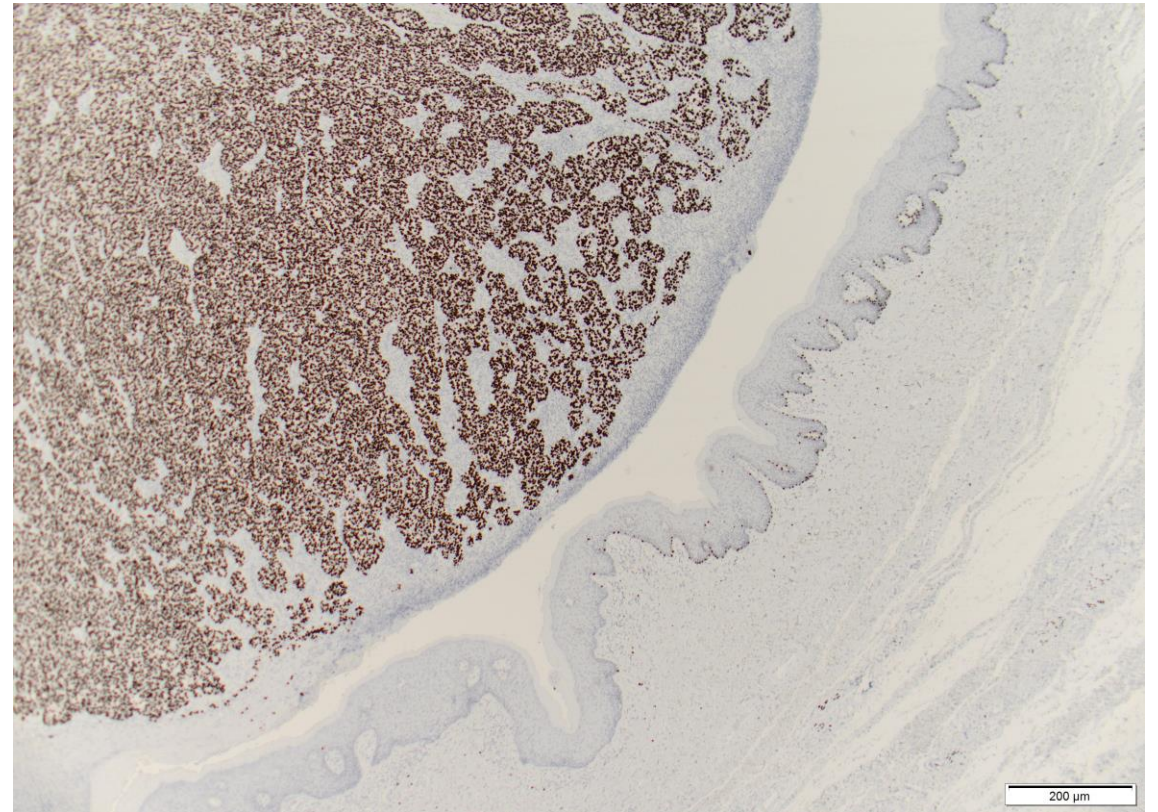




## Melan A



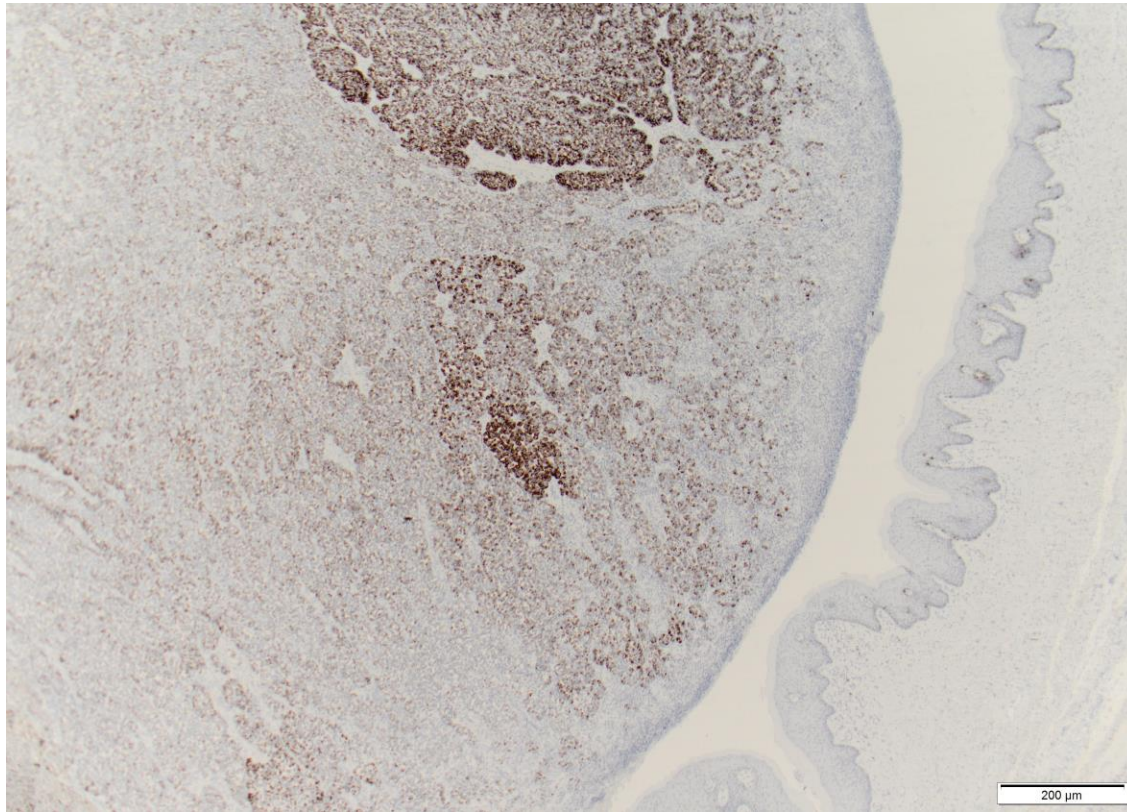
## SOX10



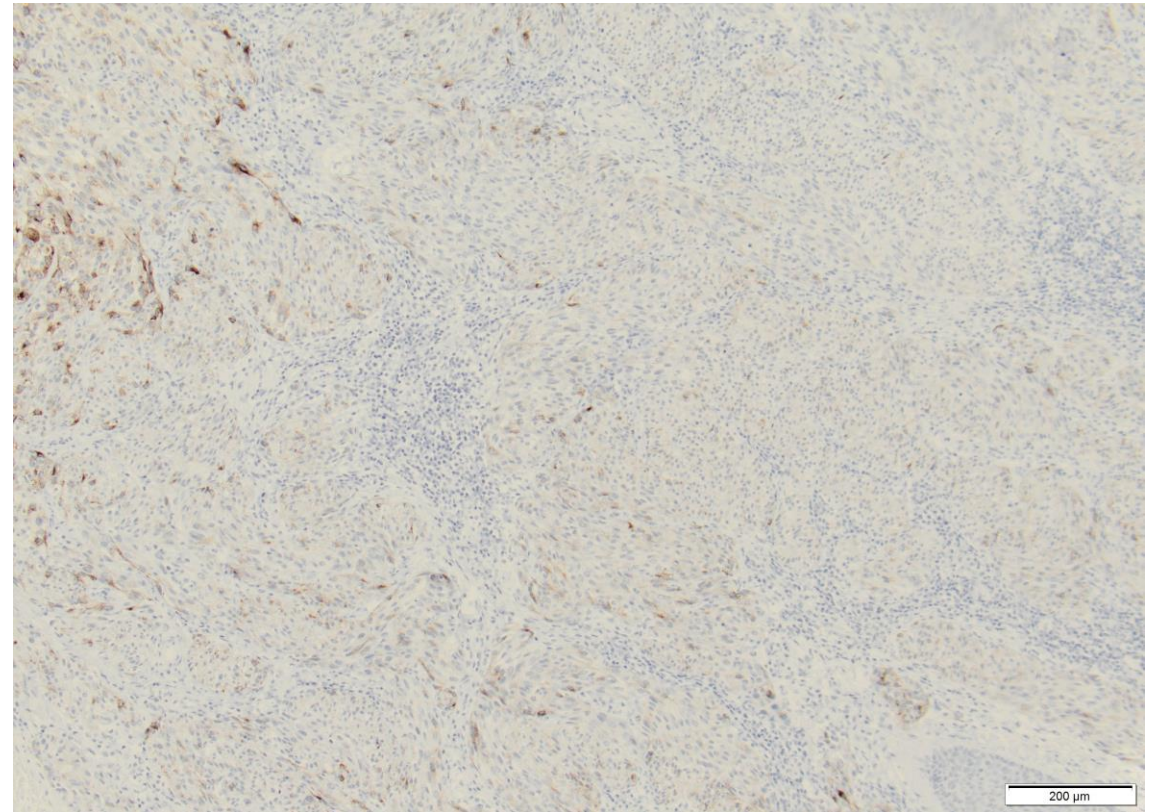


# HMB45

**Pattern 1**

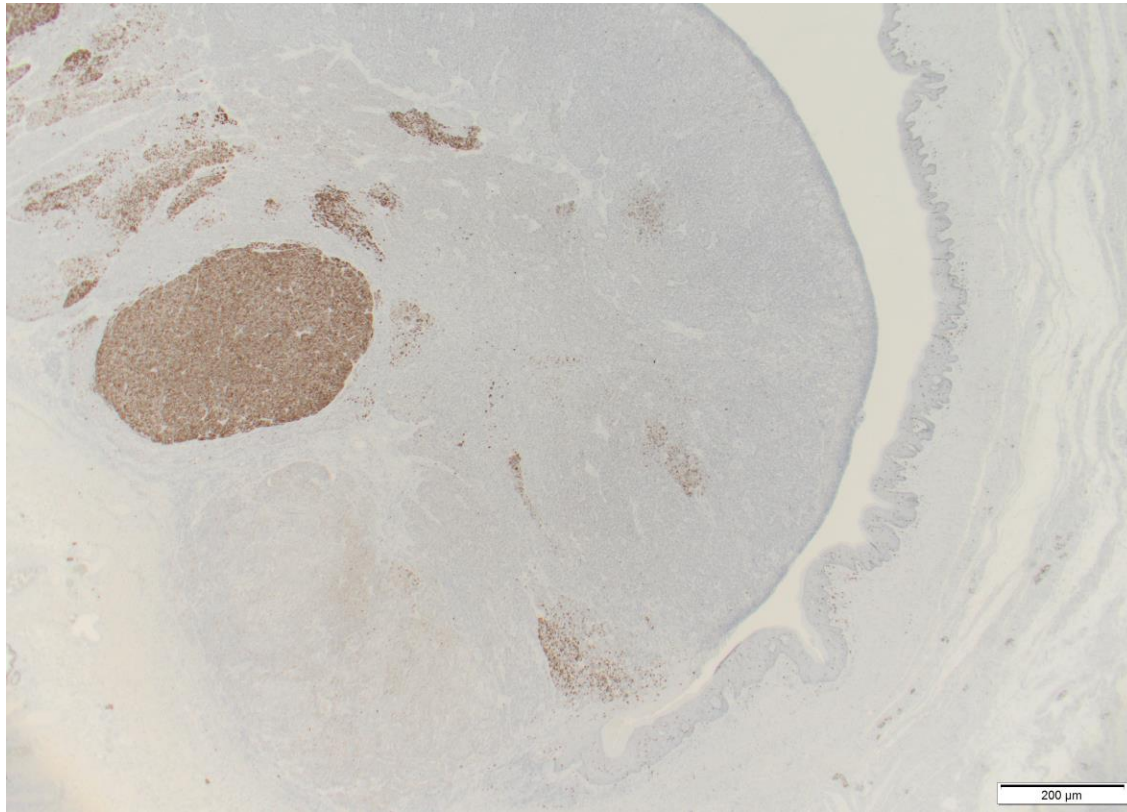


**Pattern 2**





## S100



## pancytokeratin





# Molecular Mutations

- KIT – NRAS – BRAF
- NRAS being uncommon in sun exposed skin

# Issues with VMM staging

- Location lends itself to more than one classification resulting in staging ambiguity
- No consensus on staging for the vulva
- Breslow's seems to have the most correlation with survival
  - Difficult if mucosal



# Predictors of Survival

- Breslow's thickness
- Increased Age
- Lymph node involvement
  - Sentinel lymph node?
- Ulceration

Things that aren't → histological type

# Thank you

