EQA Circulation 43 Educational Cases

E1-E2

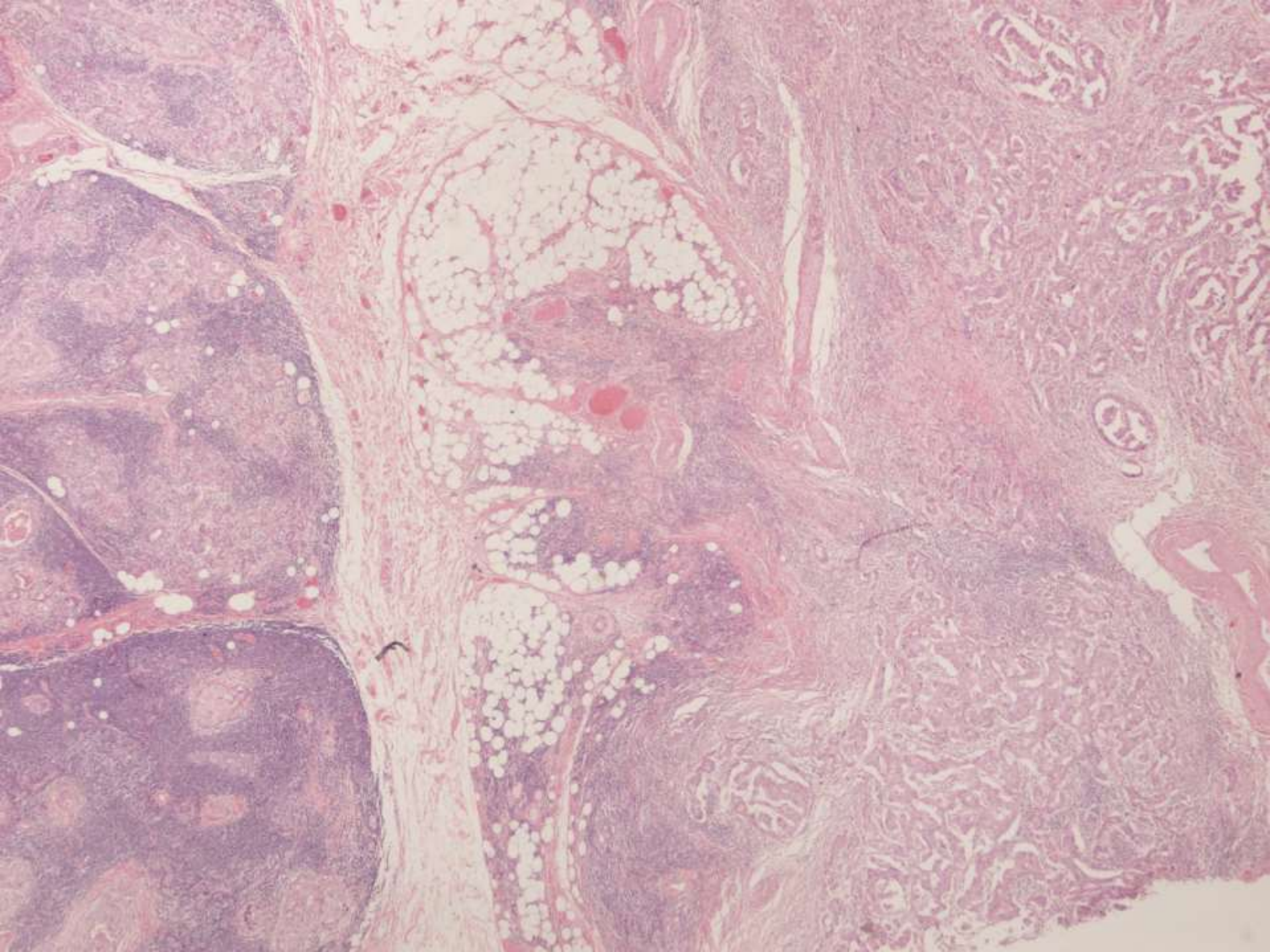
Monica Agarwal
Monklands Hospital

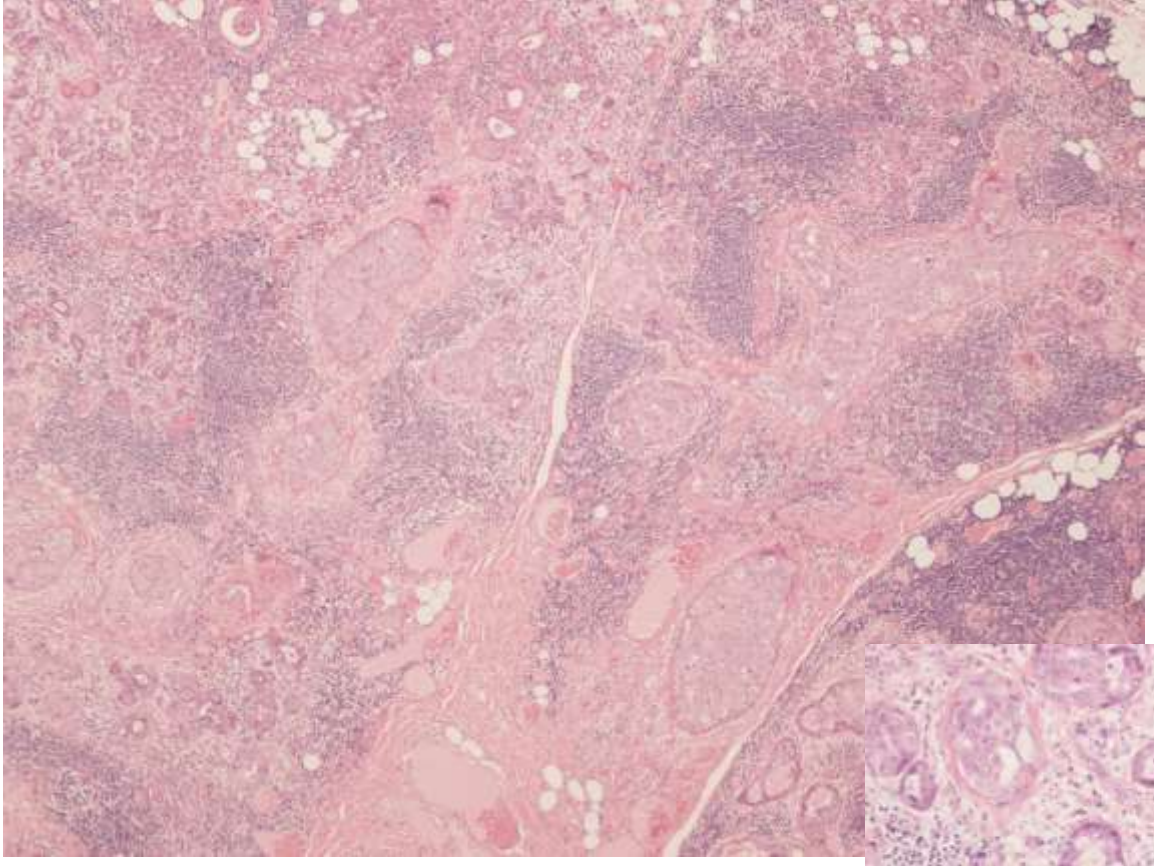
E1

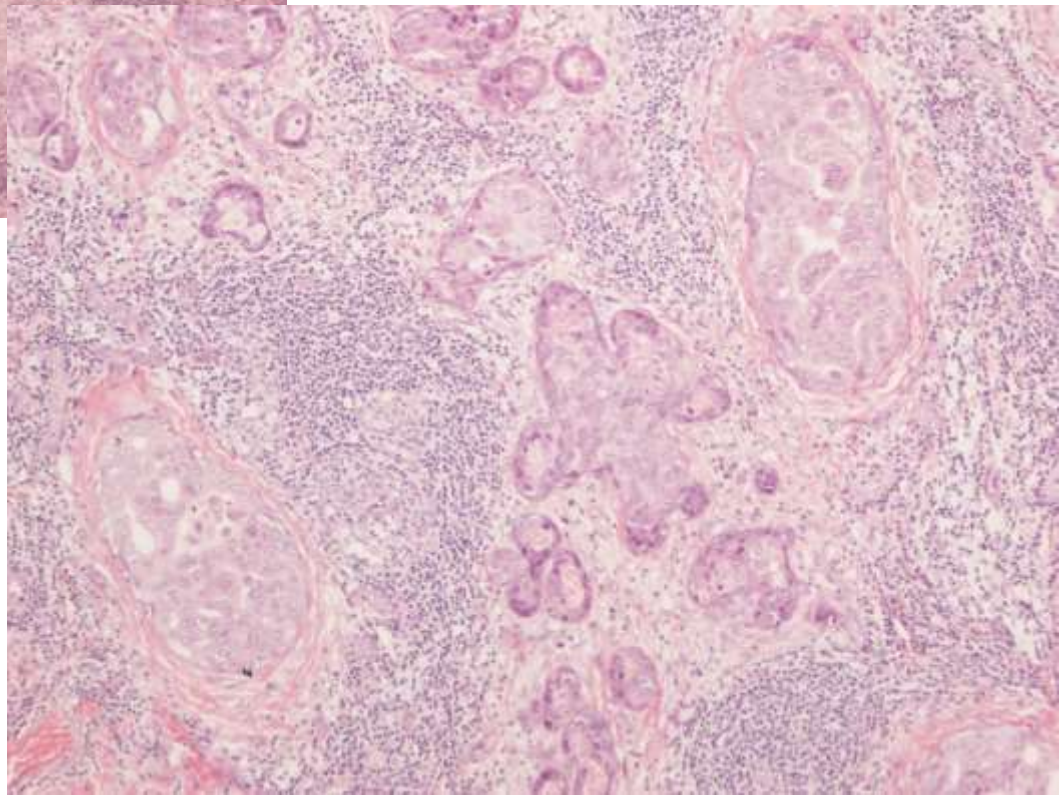
- 38 yrs male
- Submandibular gland tumour

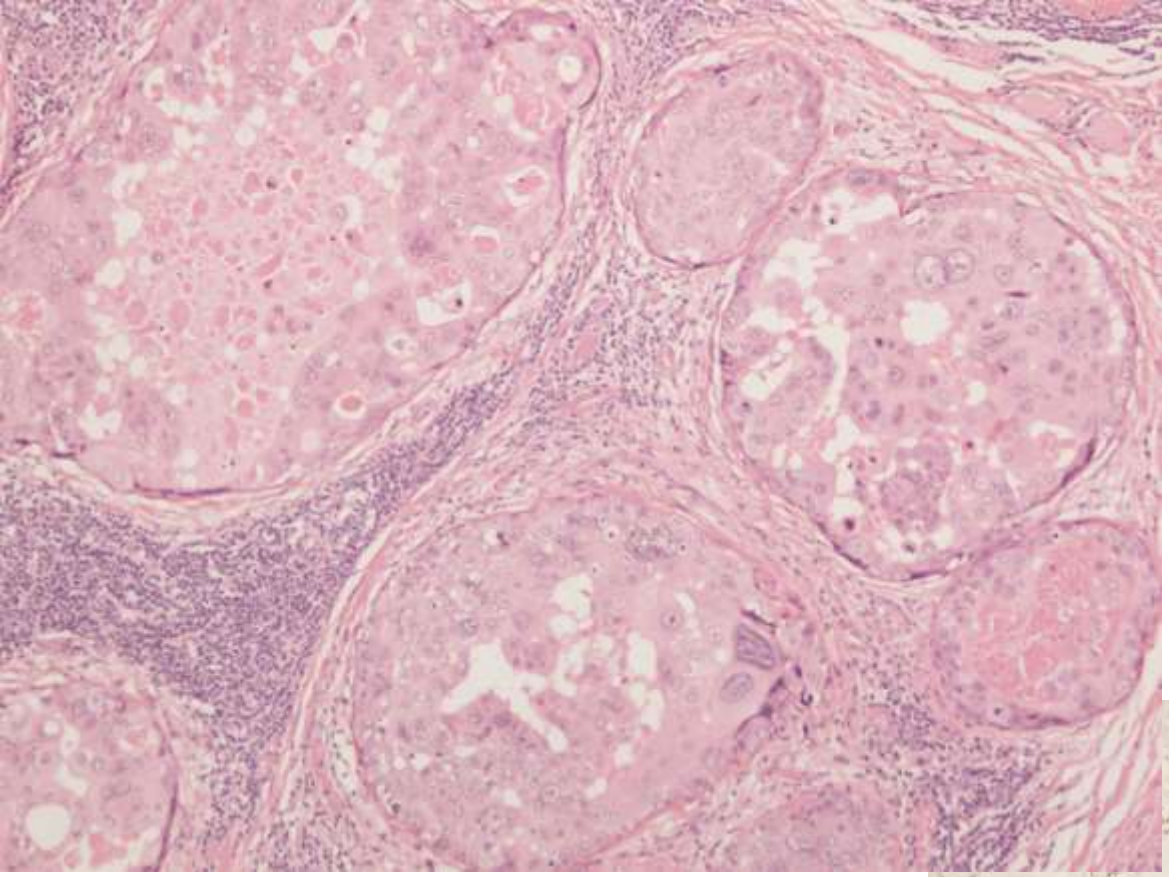
E1

- Formal excision following diagnosis of poorly differentiated carcinoma on core biopsy
- 20 mm tumour
- Grey/white cut surface







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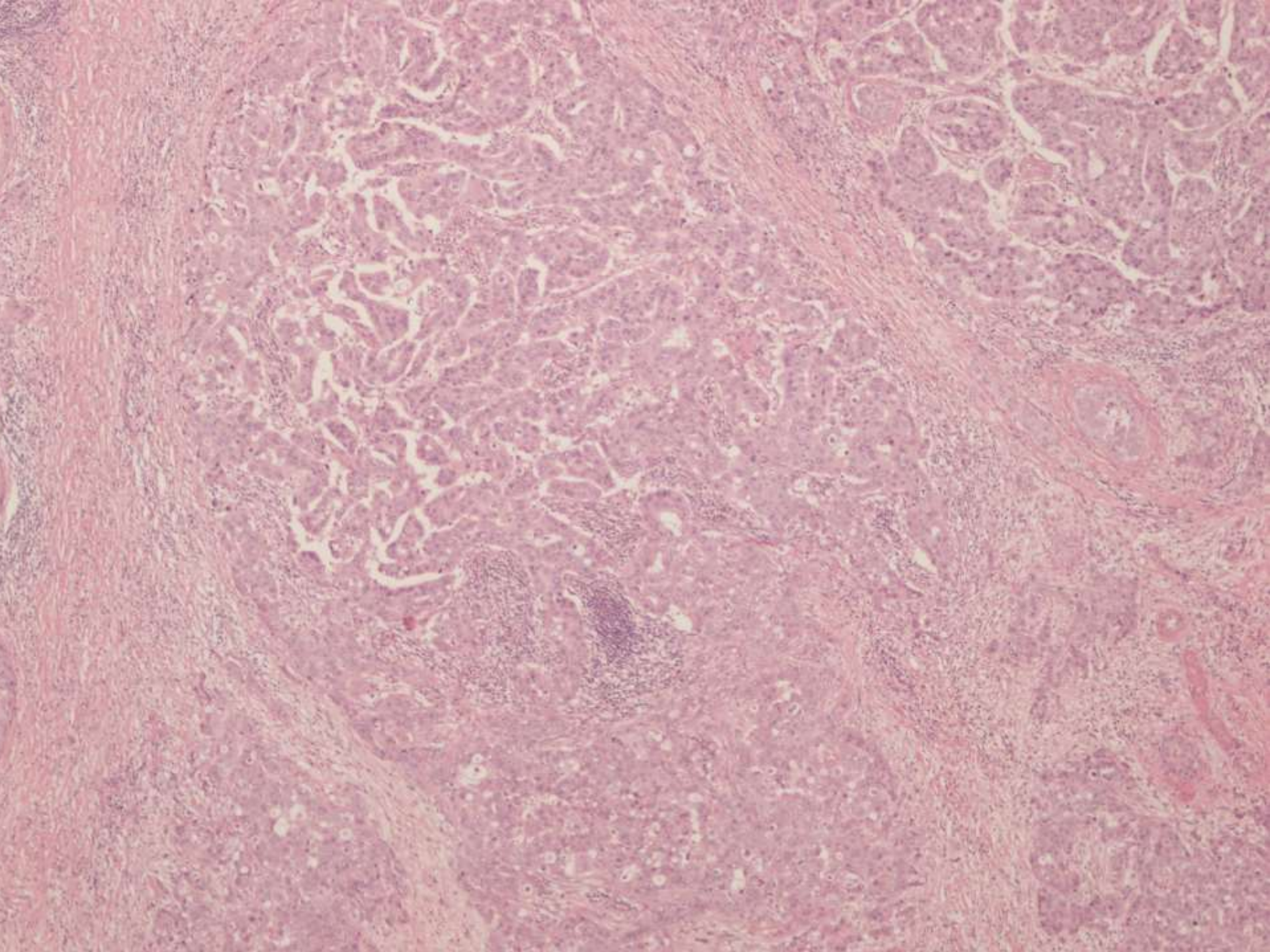

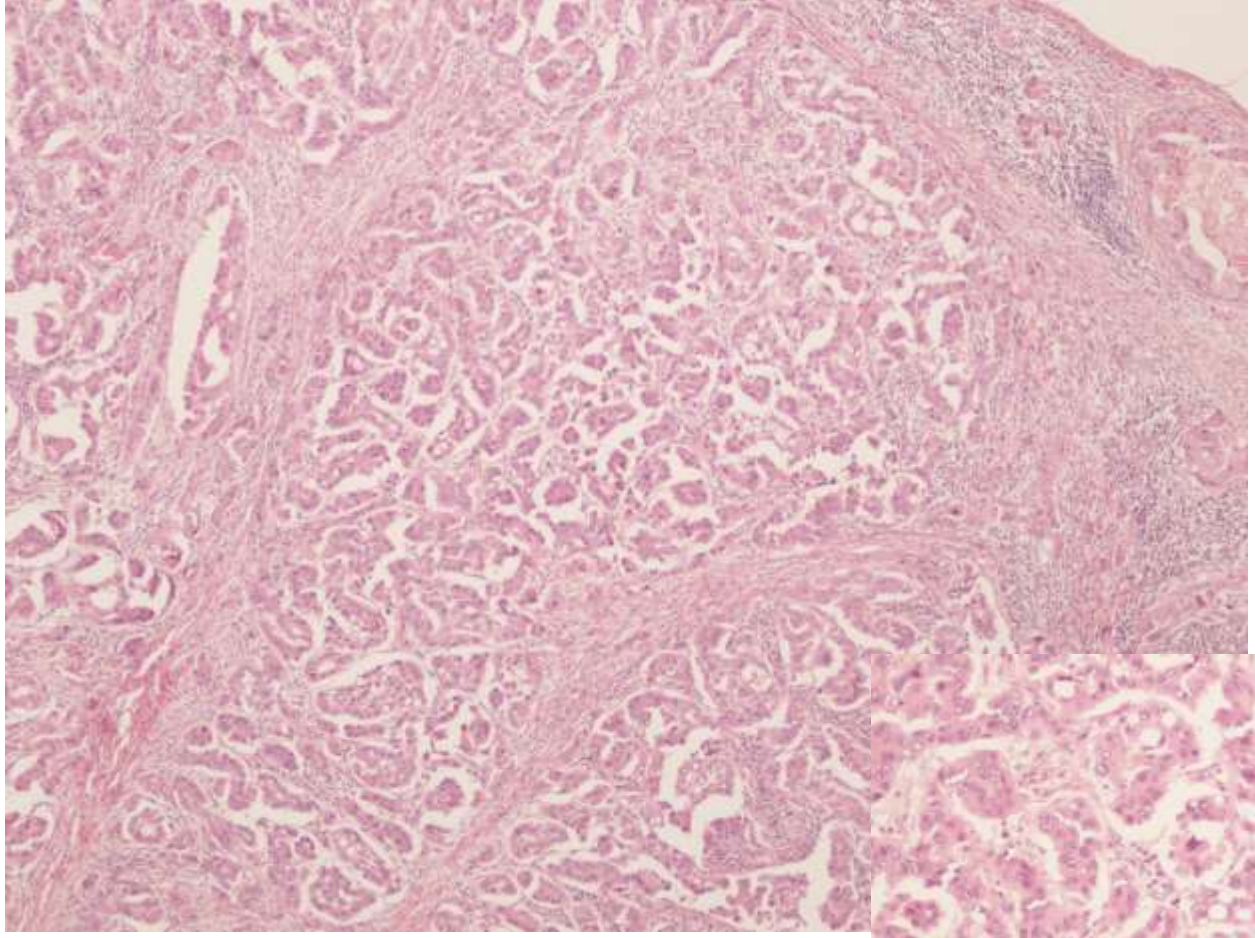

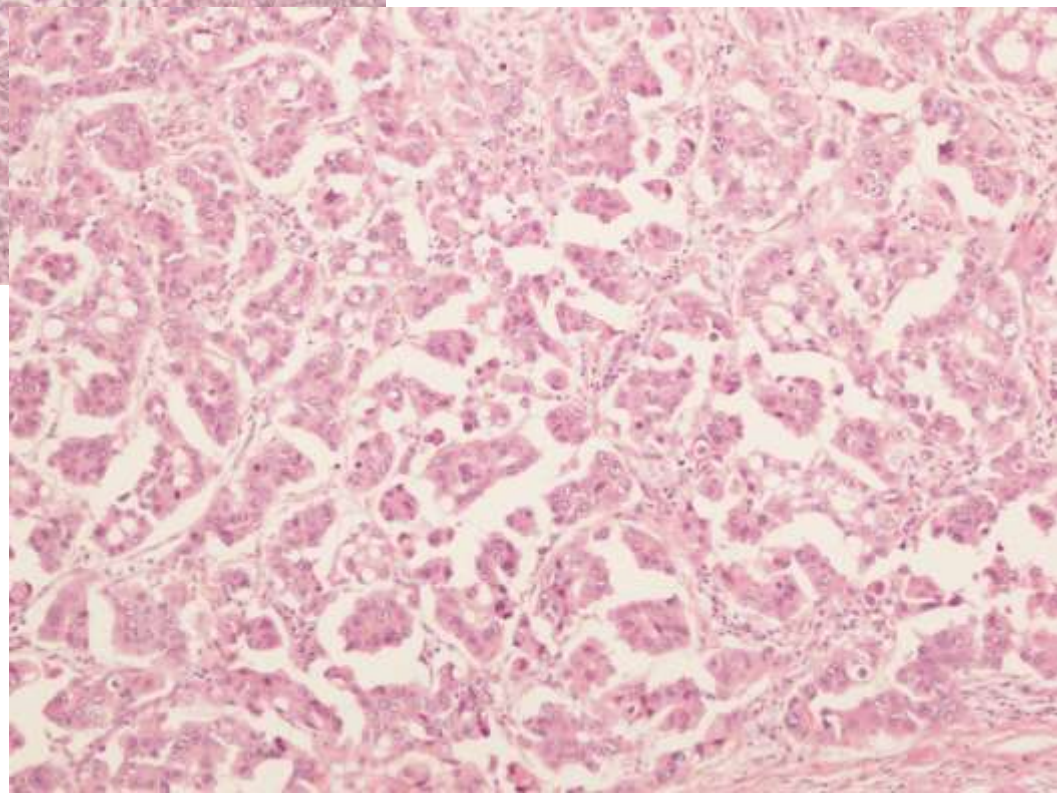

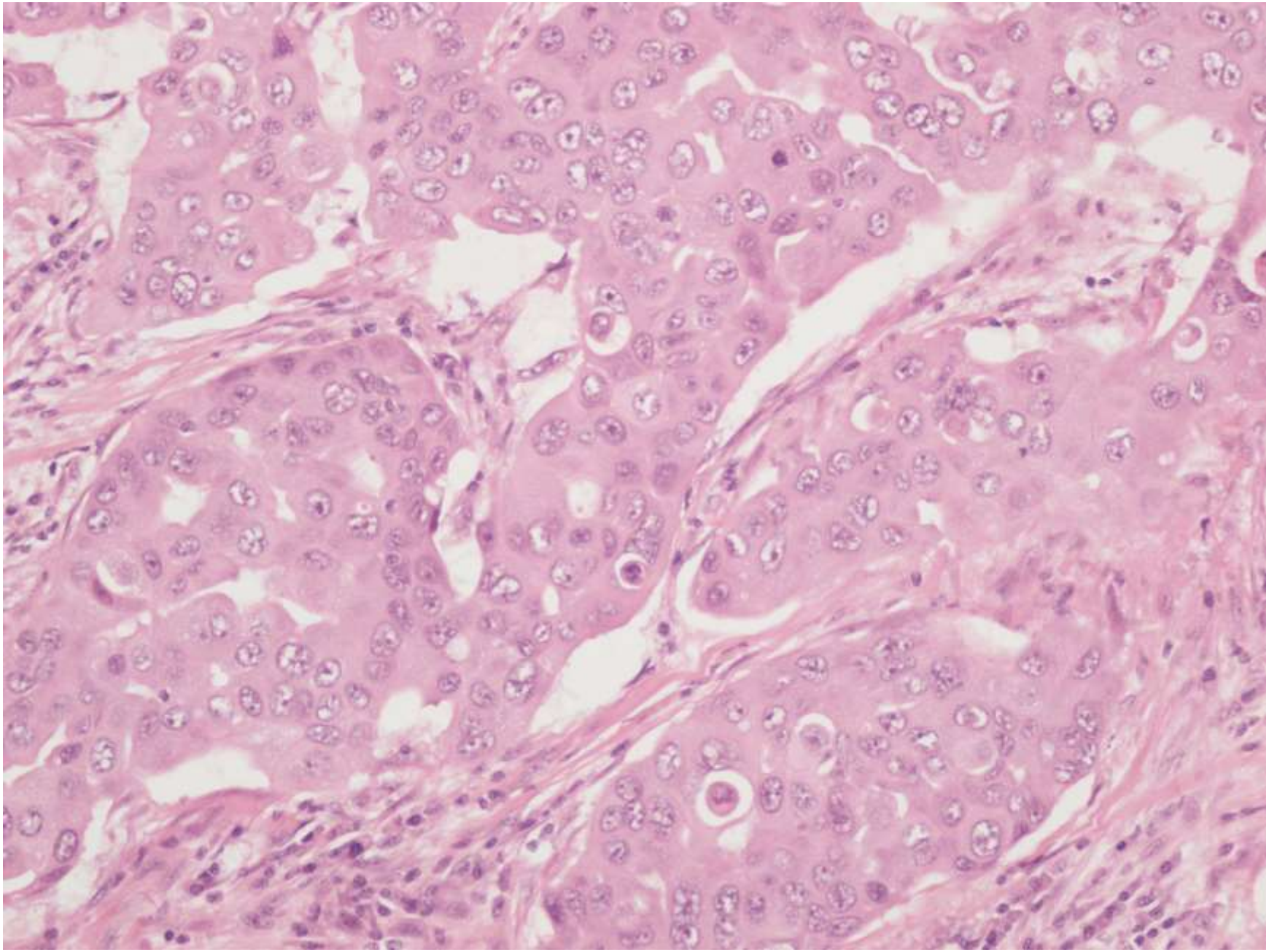

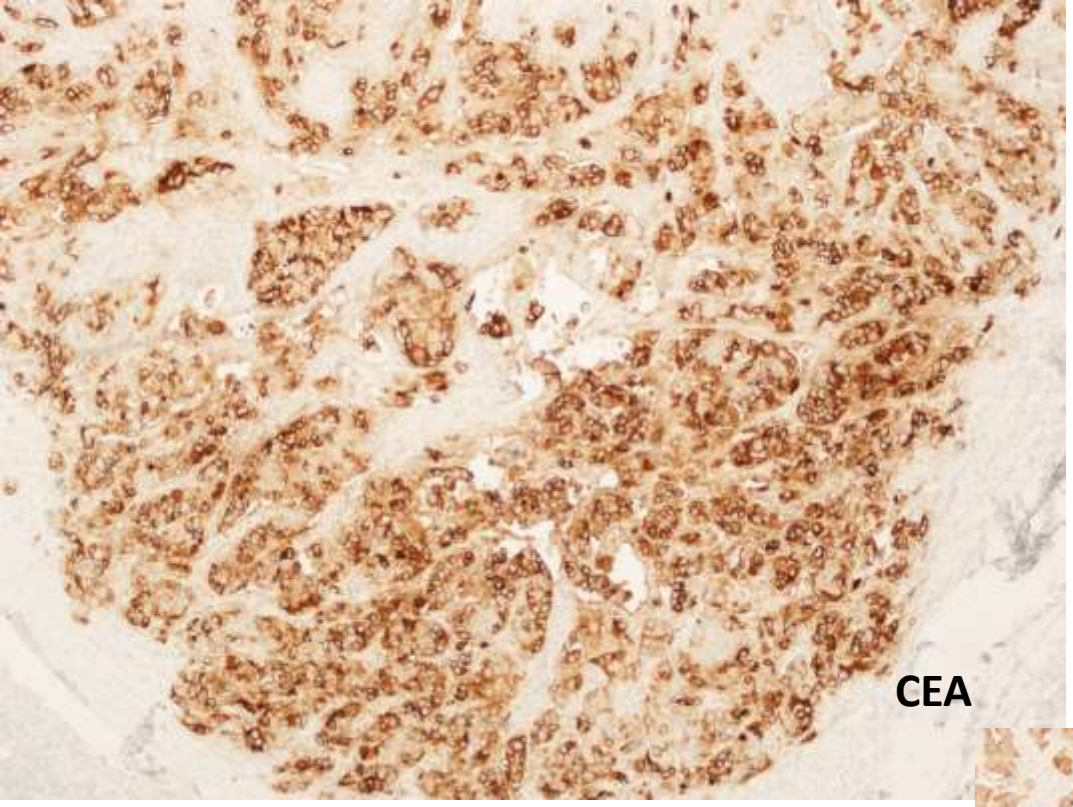
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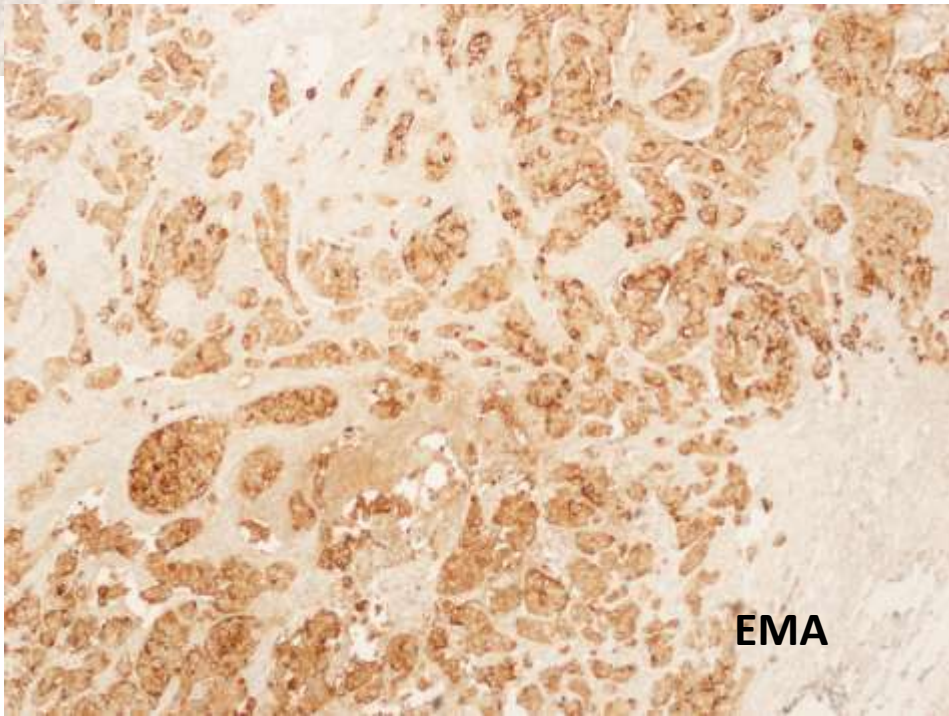

EMA


S100


ER

Responses

Diagnoses and D/D

- Salivary duct ca – 74
- Mucoepidermoid ca – 10
- Oncocytic ca – 6
- Squamous cell ca with cancerisation of salivary ducts – 2
- Malignant Warthin's tumour – 1
- Epithelial myoepithelial ca – 1
- Micropapillary ca with squamous differentiation ?thyroid metastasis to lymph node – 1
- Mammary analogue secretary ca – 2
- Lymphoepithelial ca – 5
- Papillary adenoca - 1
- Necrotizing sialometaplasia with marked atypia - 1

Diagnosis

Salivary duct carcinoma

Salivary duct carcinoma

- Uncommon salivary gland malignant tumour (about 9%)
- Frequently seen in elderly population
- Commonly in 6^{th} and 7^{th} decades
- More common in males (M:F 3-6:1)
- Majority in parotid gland, some occur in submandibular gland and rarely in minor salivary gland
- Rarely reported in longstanding chronic obstructive sialadenitis

Salivary duct carcinoma

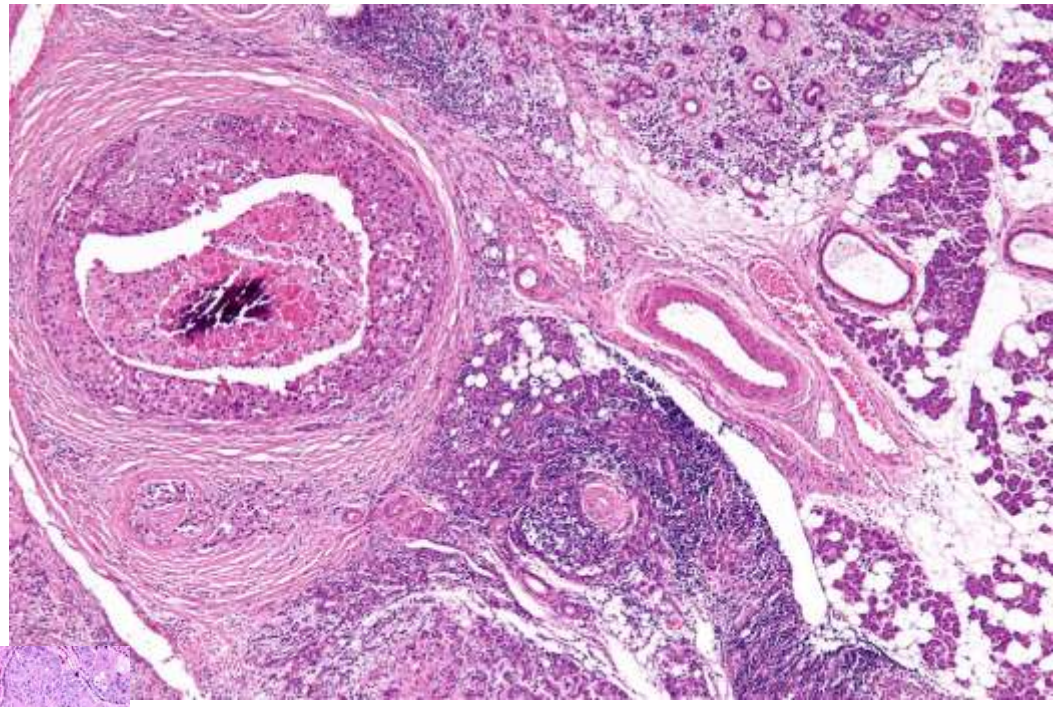
- One of the most aggressive salivary gland malignant tumour
- Local recurrence 33%; distant metastasis 46%
- Metastasis – lymph nodes, distant
- Frequent sites of distant metastasis – lung, bone, brain, liver, skin
- 65% patients die of disease usually within 4 years of diagnosis

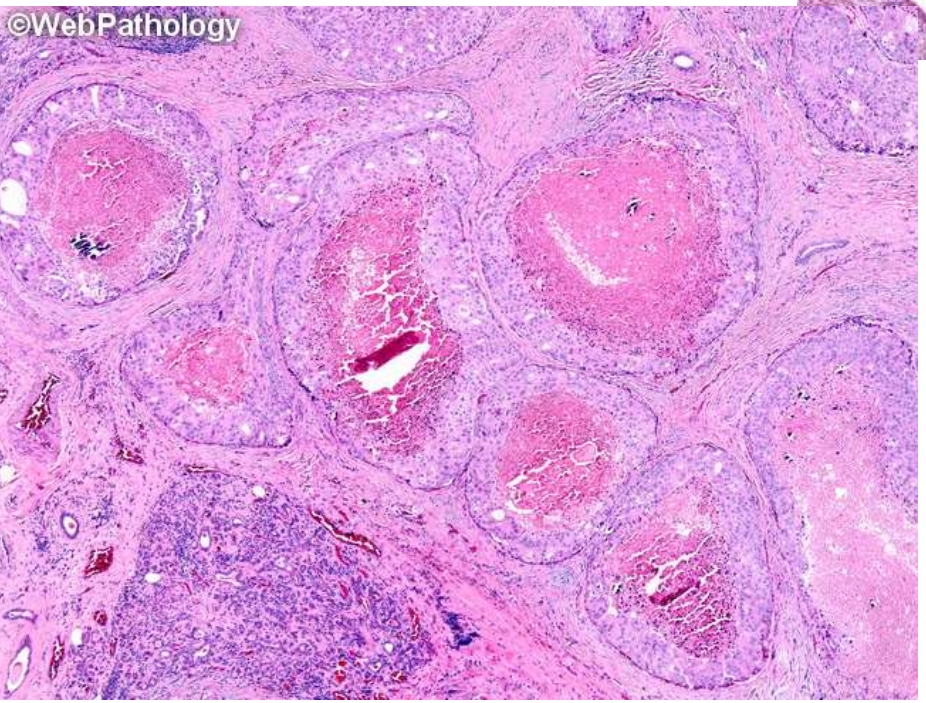
Salivary duct carcinoma

- Usually poorly circumscribed, tan coloured and usually solid
- Morphology resembles ductal carcinoma of breast
- Intraduct like and invasive components

Intraduct like component

Cribriform, papillary, solid with frequent comedo necrosis

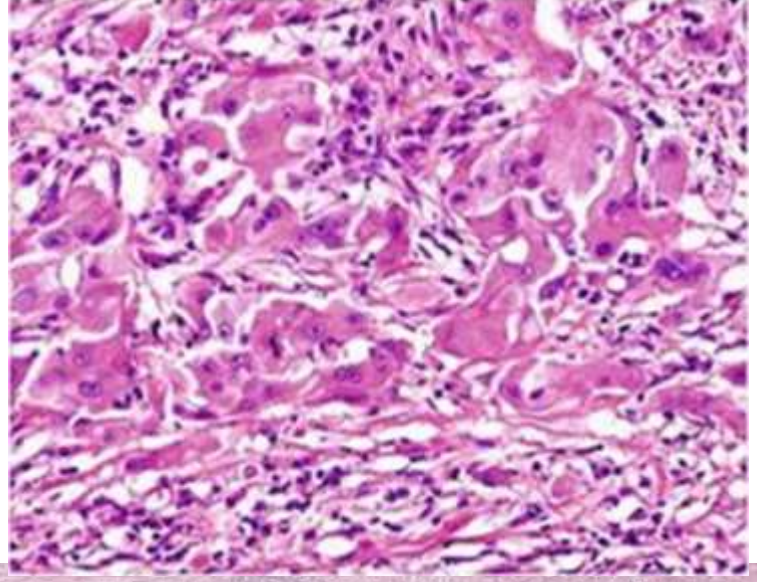


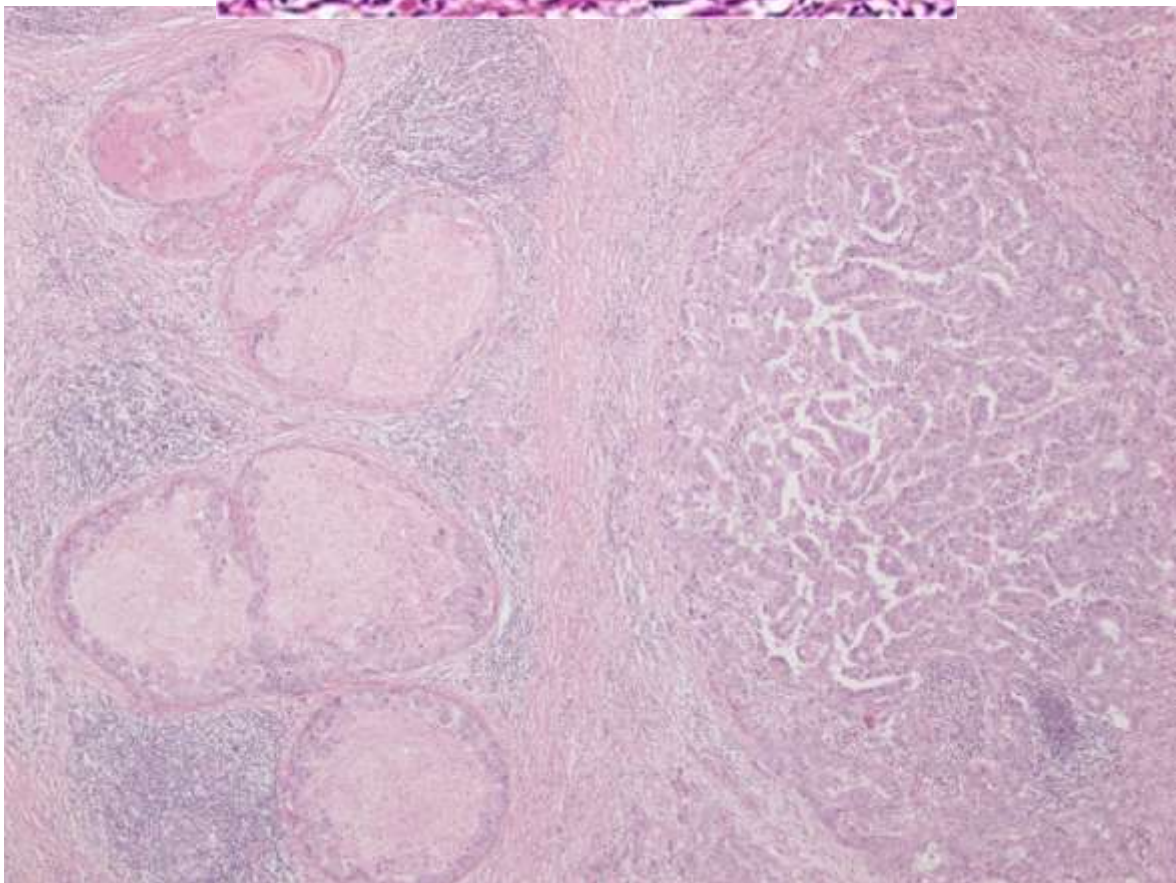


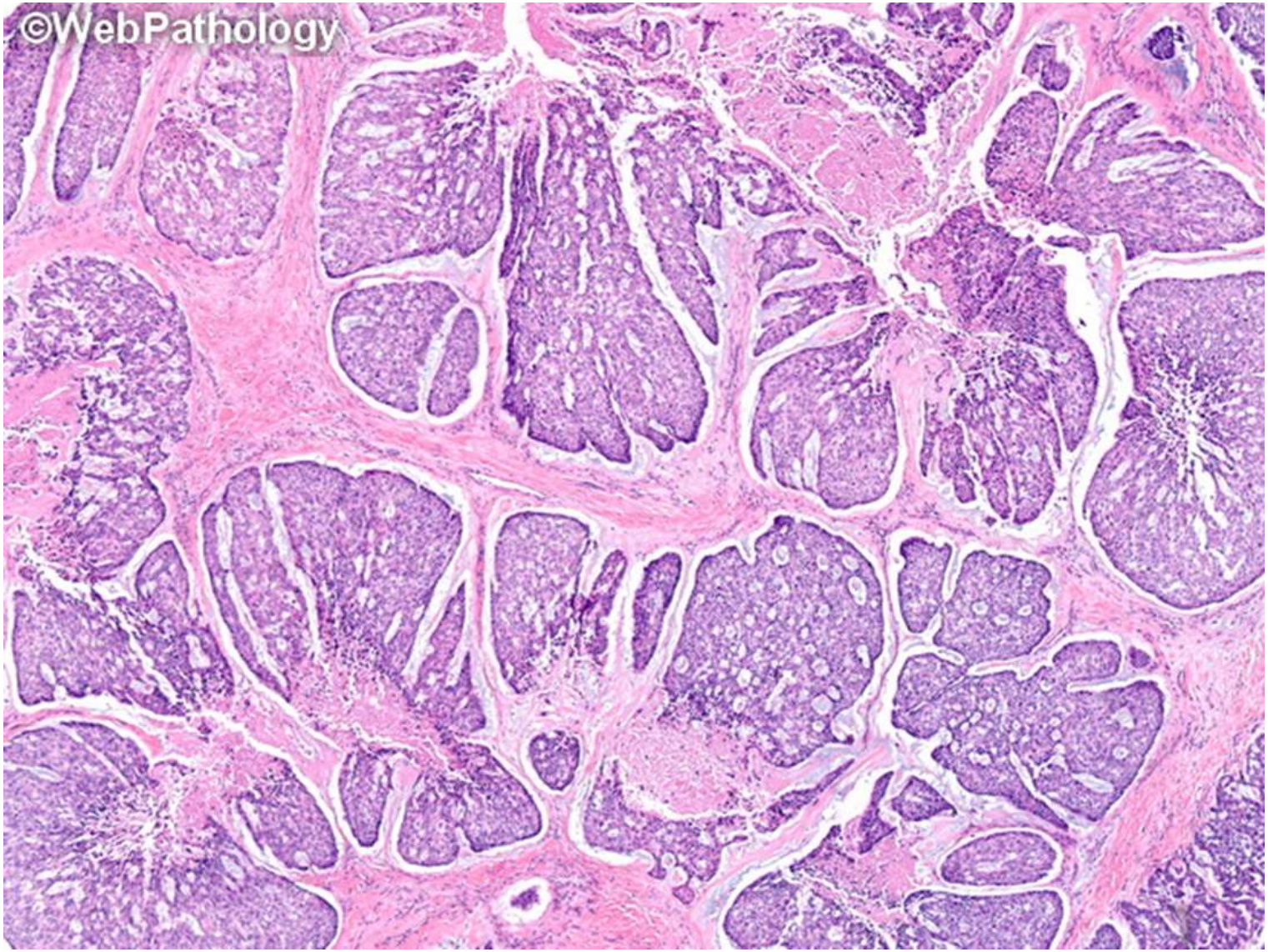
Infiltrative component

- Cribriform, solid, cords, glands
- Apocrine appearance – abundant pink cytoplasm, pleomorphic epithelioid cells, coarse chromatin and prominent nucleoli
- Squamous differentiation can be seen
- Stroma is fibrous/desmoplastic

Vascular invasion, perineural invasion commonly seen



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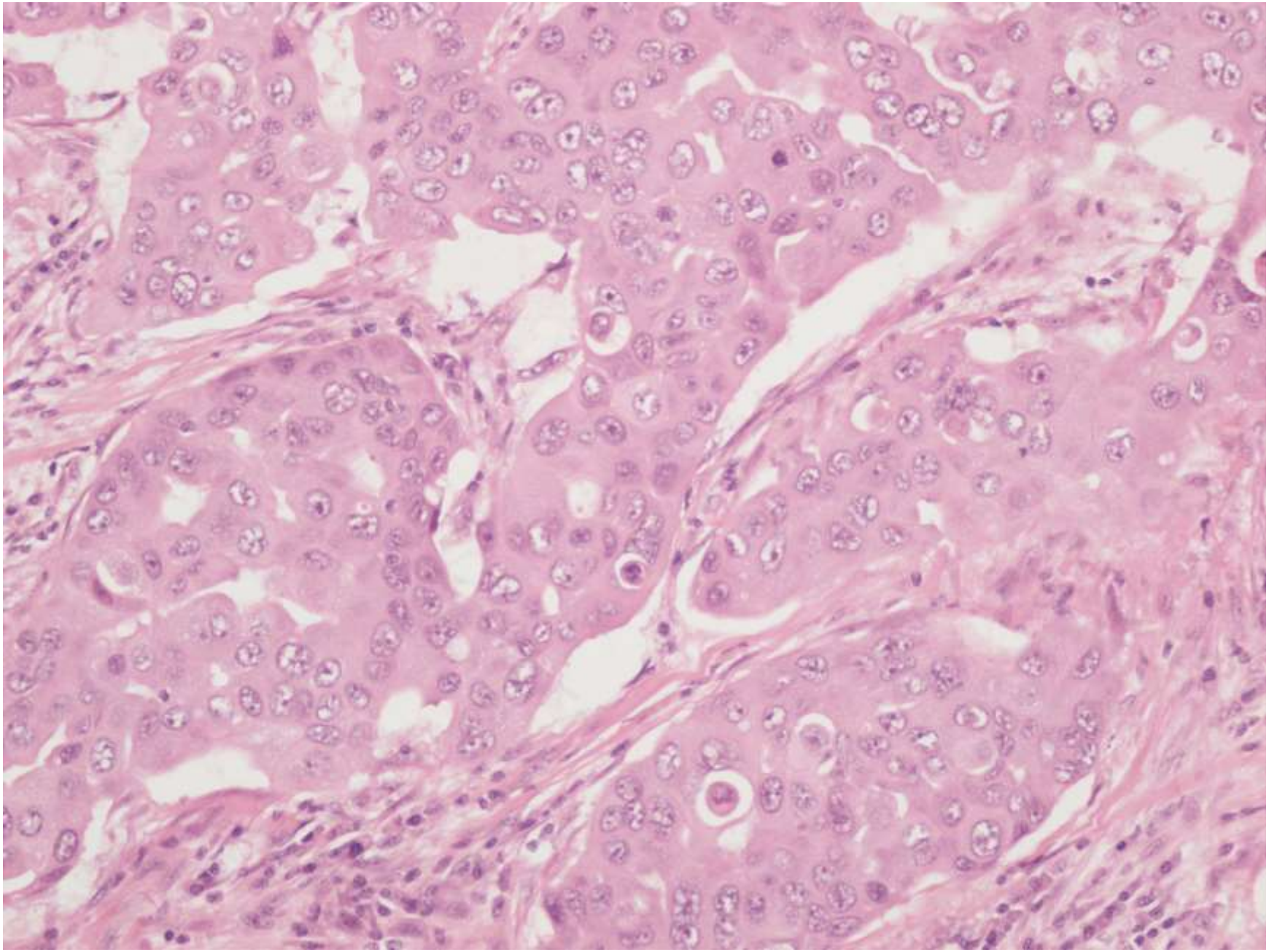
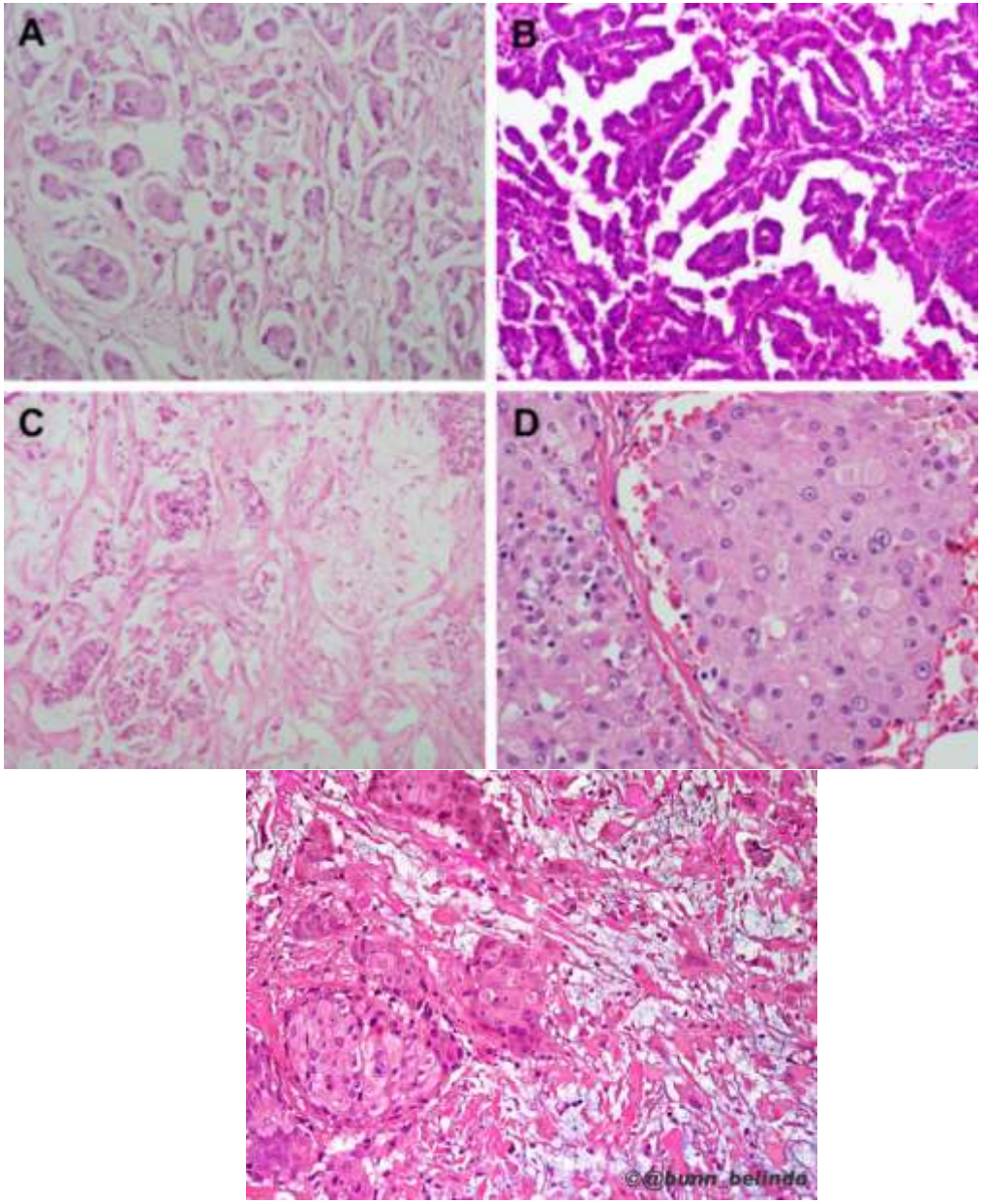


Variants:

Micropapillary, papillary, mucin rich, spindle cells



Salivary duct carcinoma

ICC

Positivity-

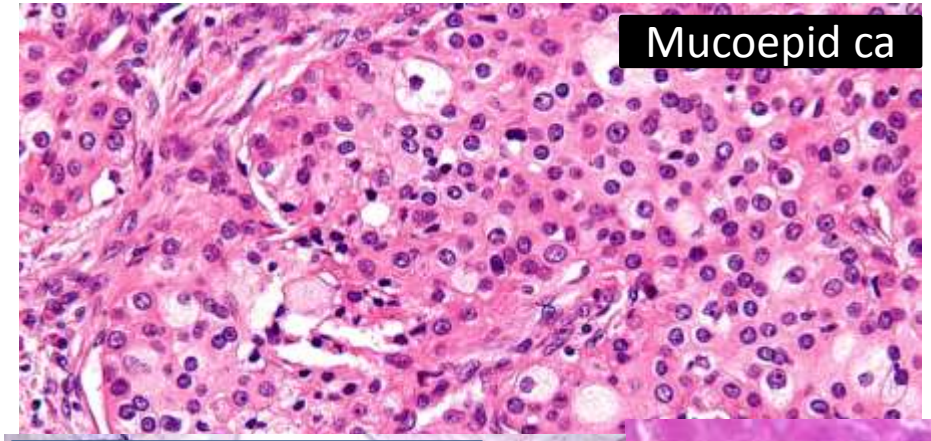
- Low and high molecular wt cytokeratins
- CEA, EMA
- Androgen receptors – strong nuclear
- GCDFP-15
- Her2 – commonly positive
- PSA, PAP - variable

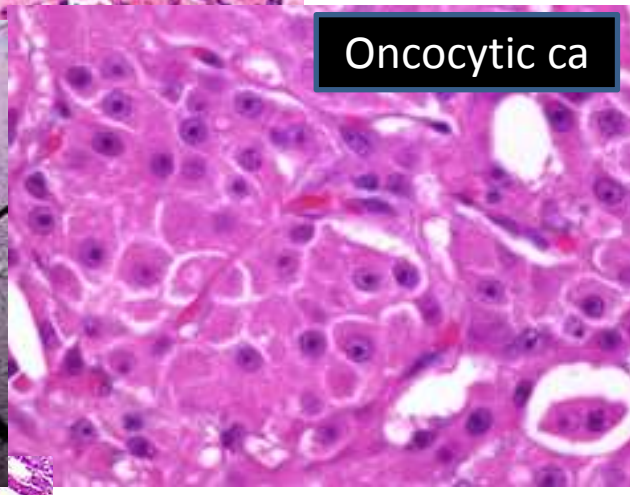
Negative-

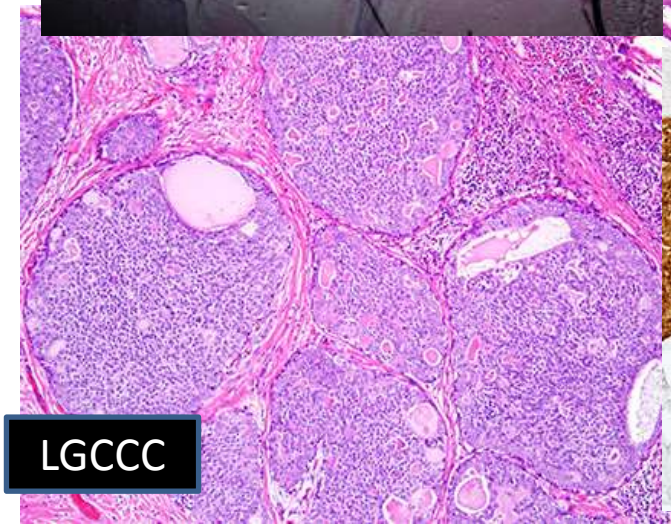
- S100
- Myoepithelial markers
- ER, PR

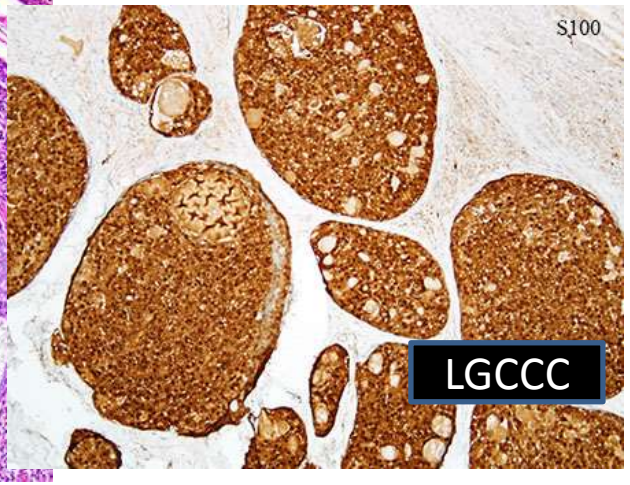
D/D

- Metastatic breast ca
- High grade mucoepidermoid carcinoma
- Oncocytic carcinoma
- Cystadenocarcinoma
- Intraductal carcinoma/Low grade cribriform cystadencarcinoma (LGCCC)







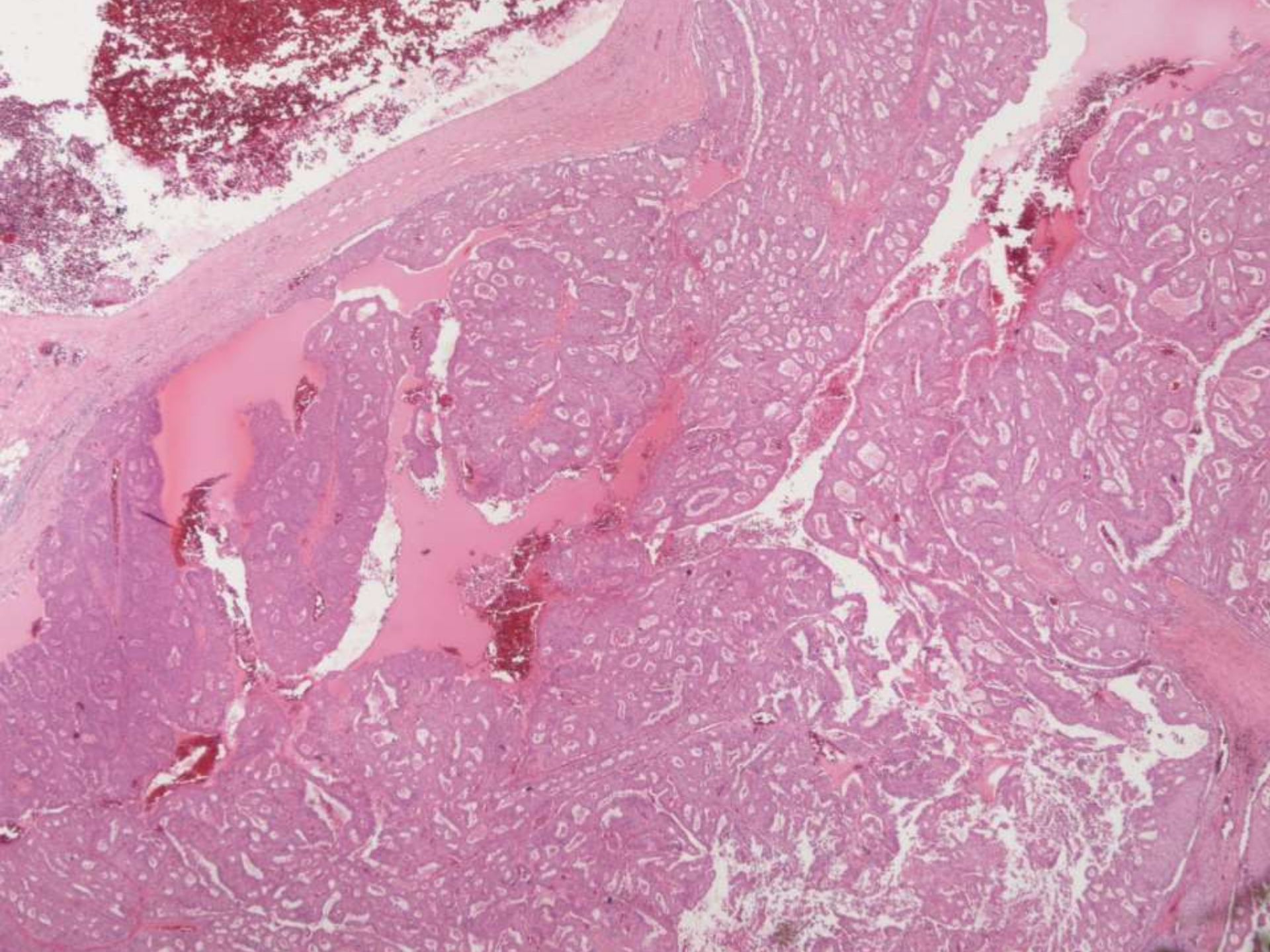


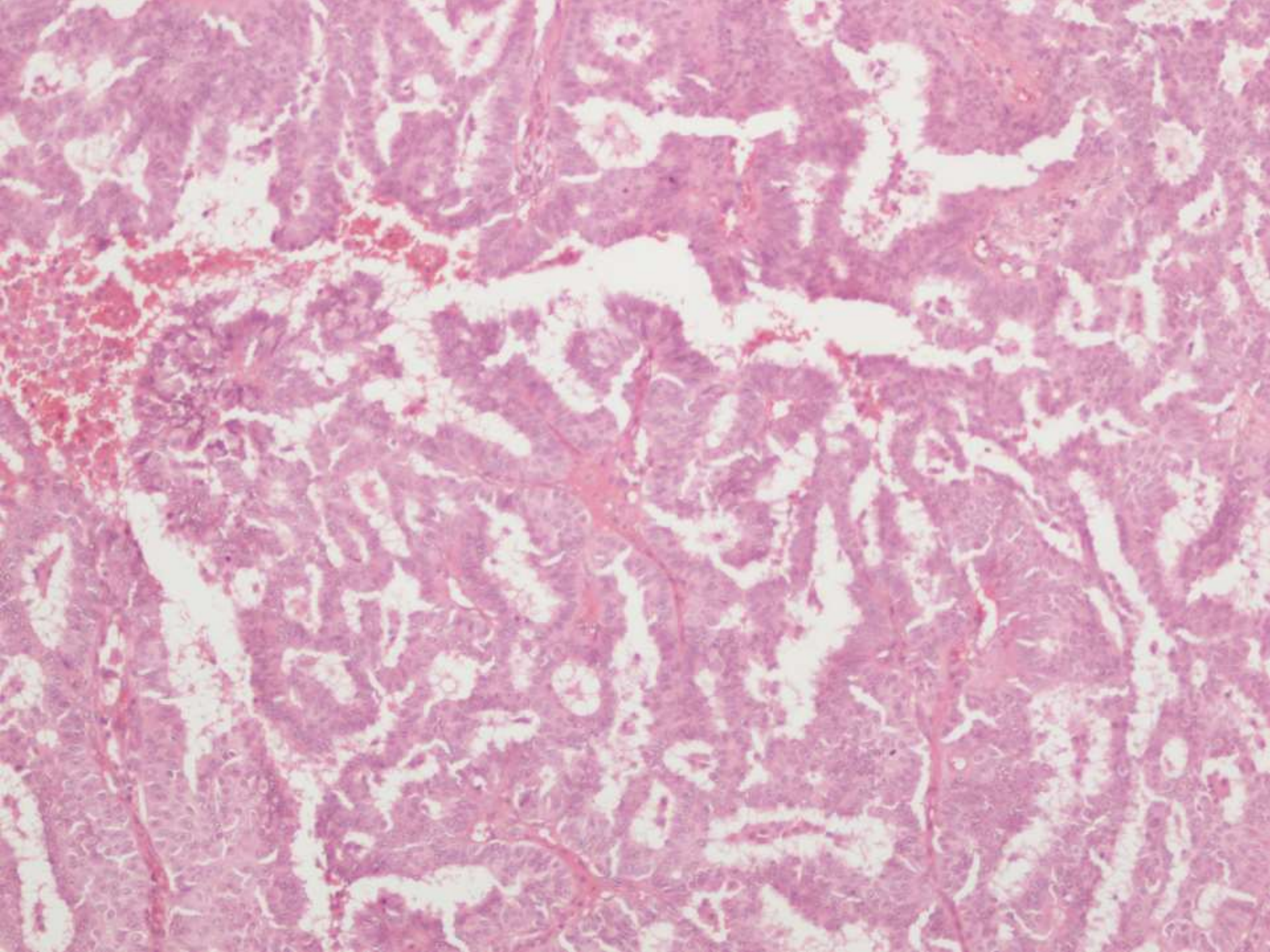
E2

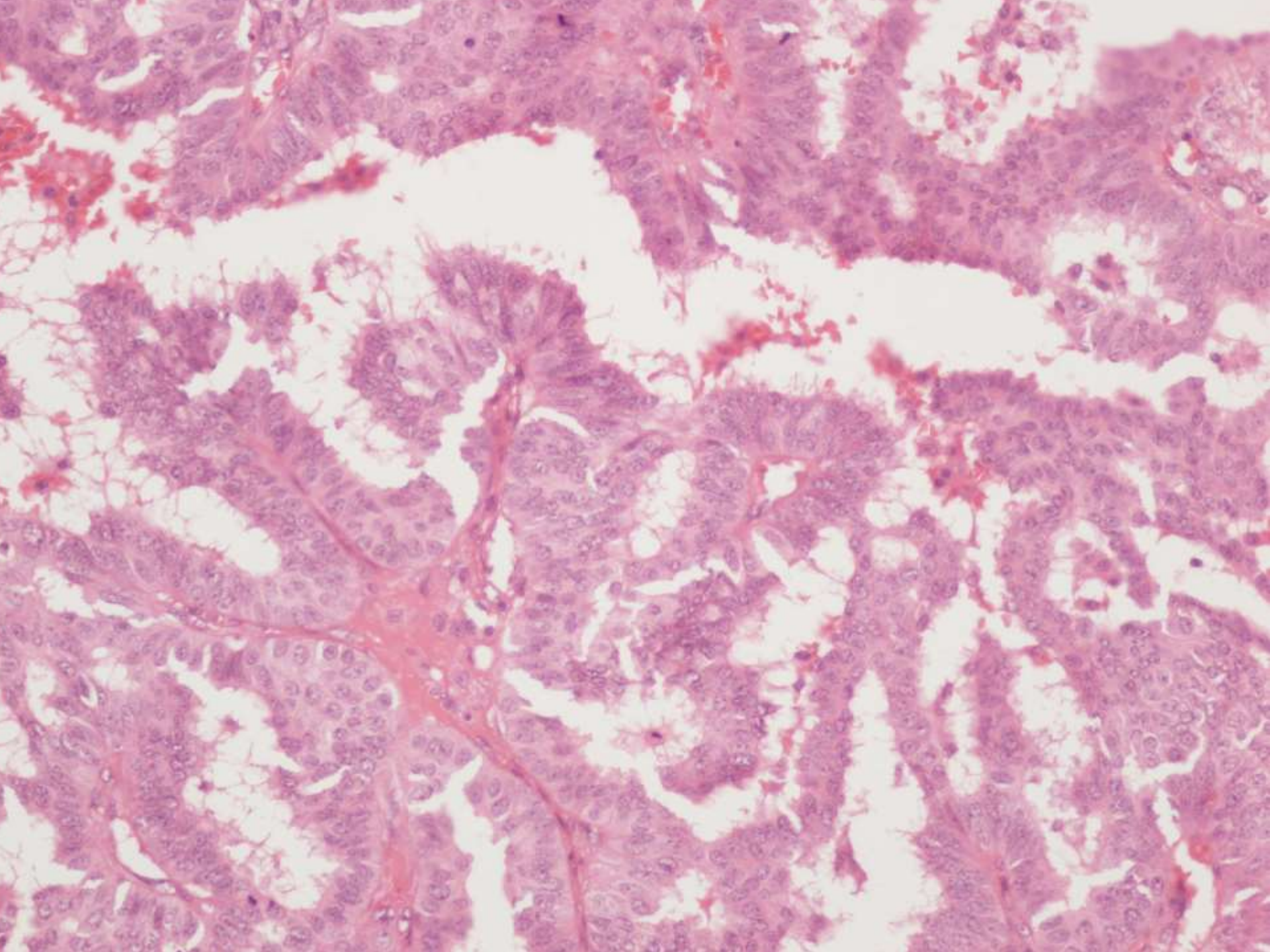
- 68 year old female
- WLE right breast

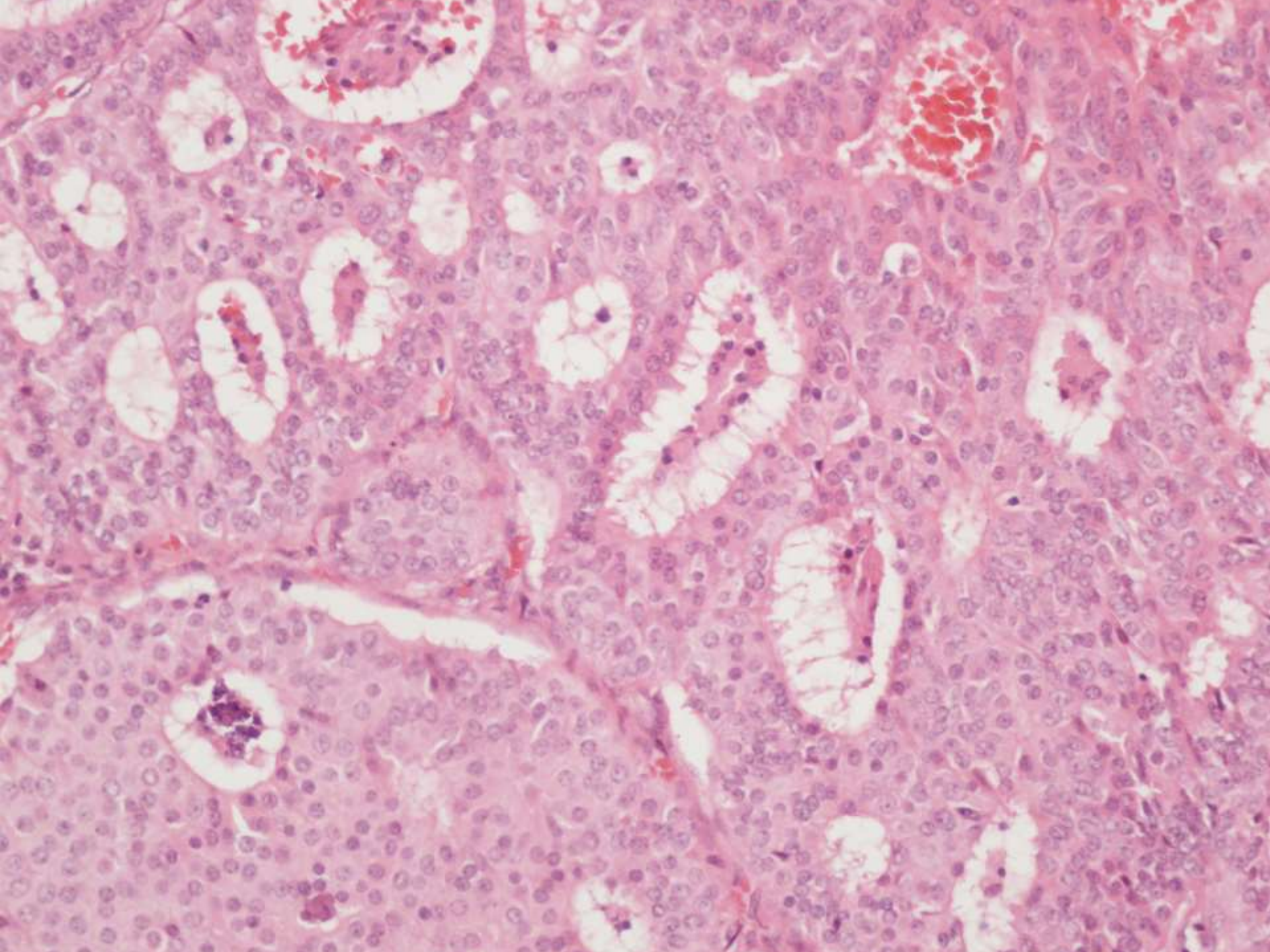
E2

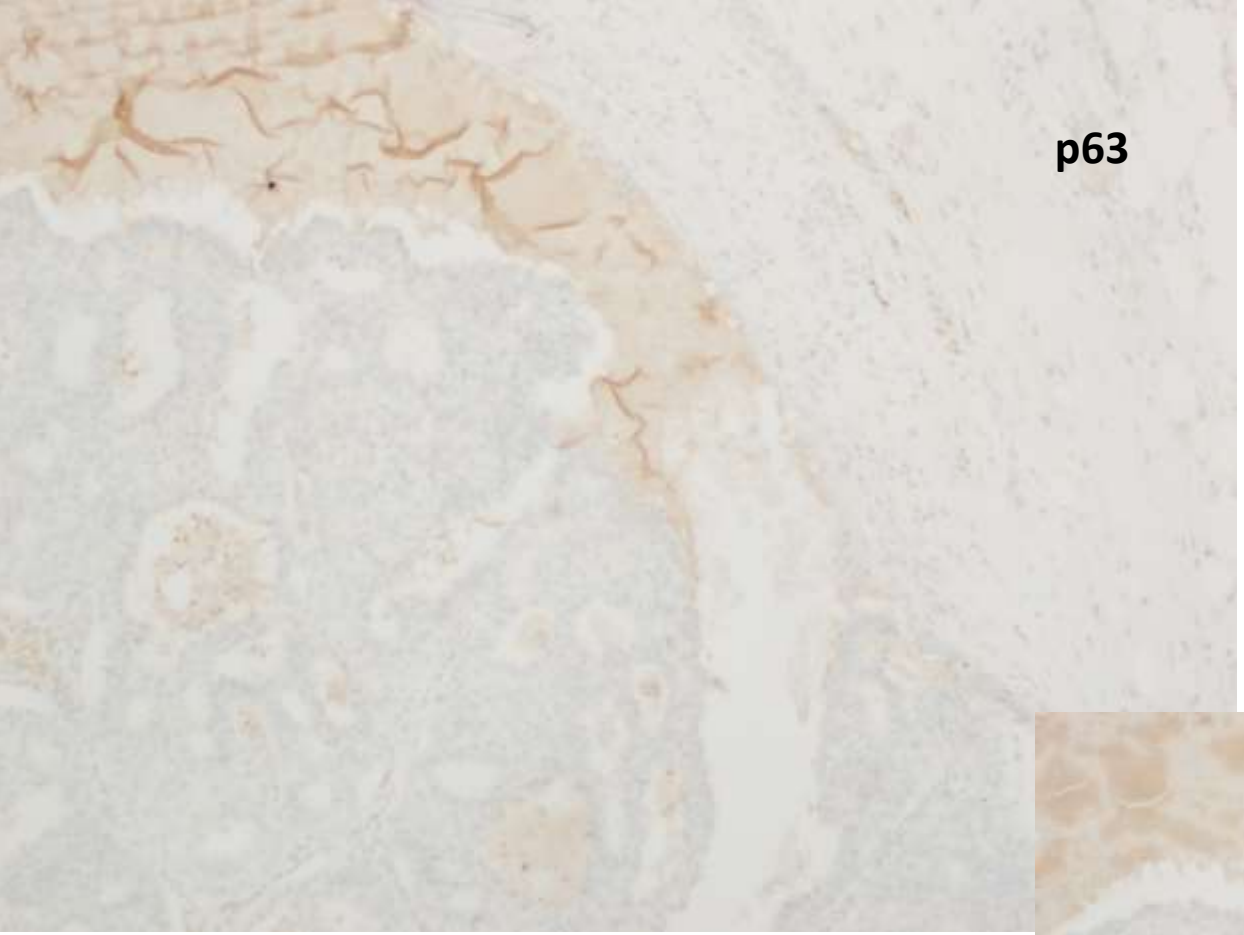
- Papillary lesion seen on core biopsy
- WLE showed a 22 mm nodular haemorrhagic lesion







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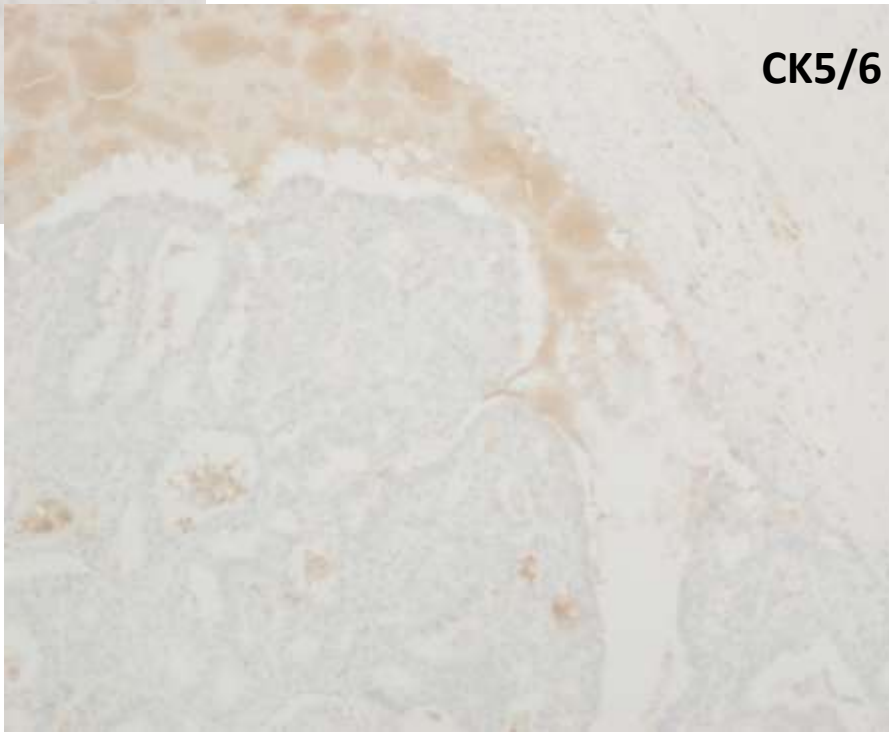
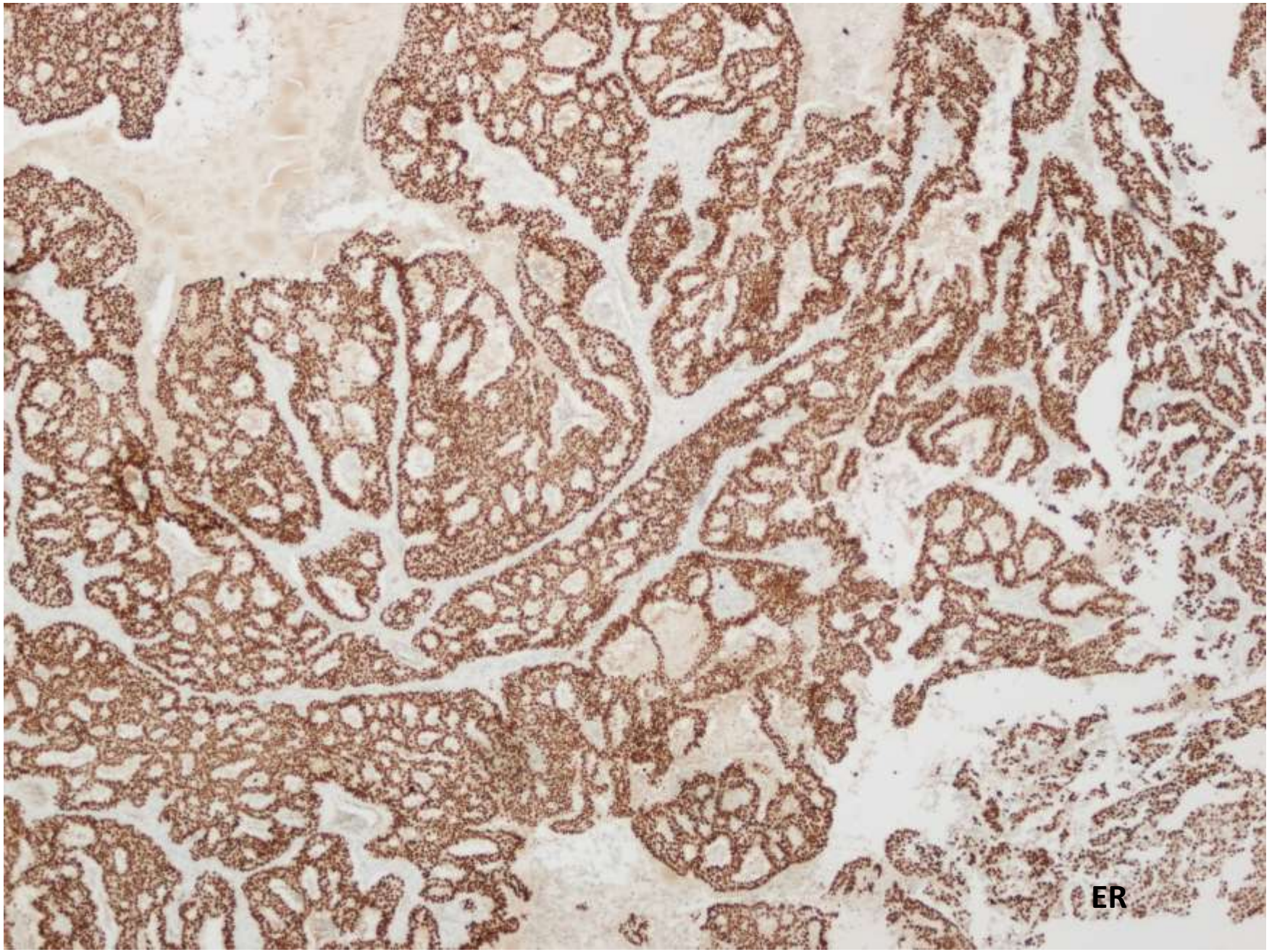

CK5/6


SMM


ER

Responses

- Encysted papillary ca – 65
- Solid papillary ca – 10
- Papillary ca – 7
- Apocrine ca – 1
- Cribriform ca – 12
- Breast ca with neuroendocrine features – 1
- Adenoca/ca – 2
- Invasive ductal ca – 1
- Intraductal papilloma/papilloma with atypical features -2
- DCIS with microinvasion – 1
- No response - 2

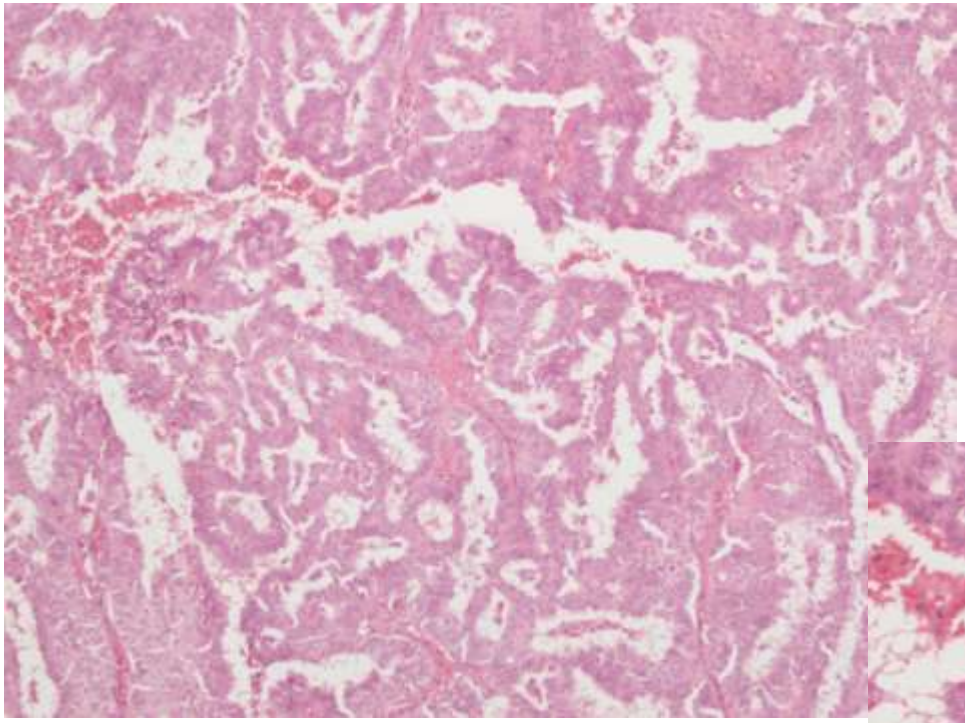
Diagnosis

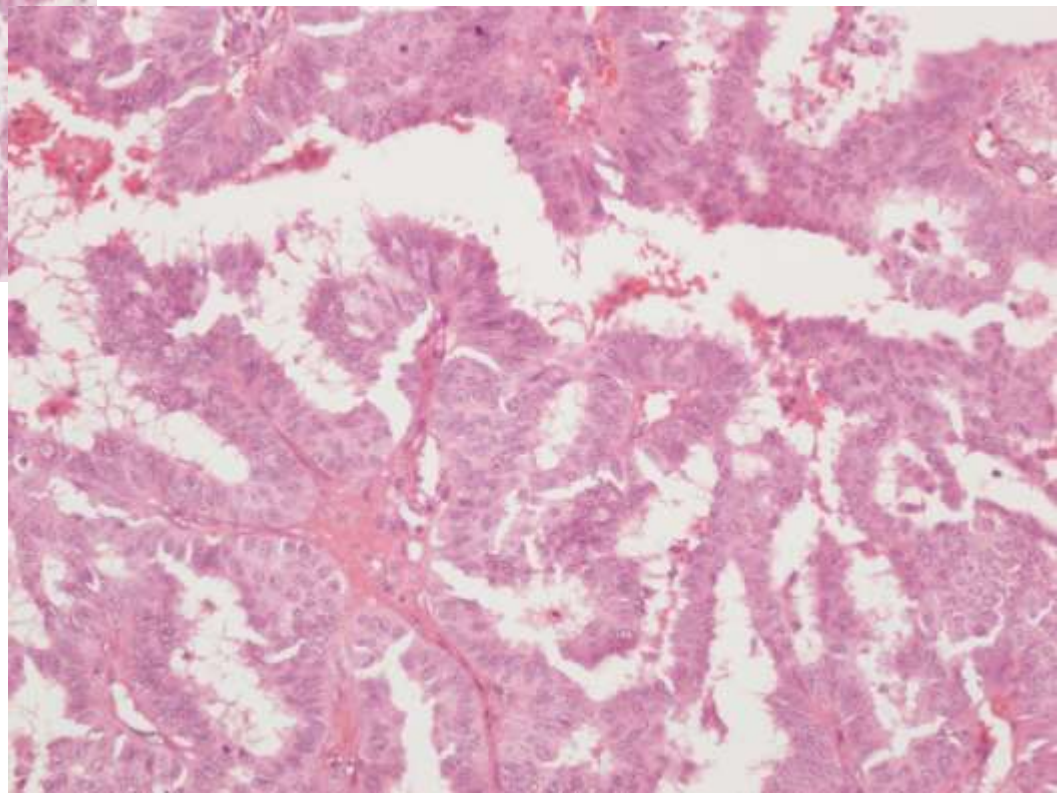
- Encysted papillary carcinoma

Papillary tumours of breast WHO 4th edition

- Benign
  - Intraductal papilloma
- Malignant
  - In-situ
    - Intraductal papilloma with DCIS
    - Intraductal papillary carcinoma
    - Encapsulated papillary carcinoma
    - Solid papillary carcinoma
  - Invasive
    - Invasive papillary carcinoma
    - Micropapillary carcinoma

In-situ papillary lesions

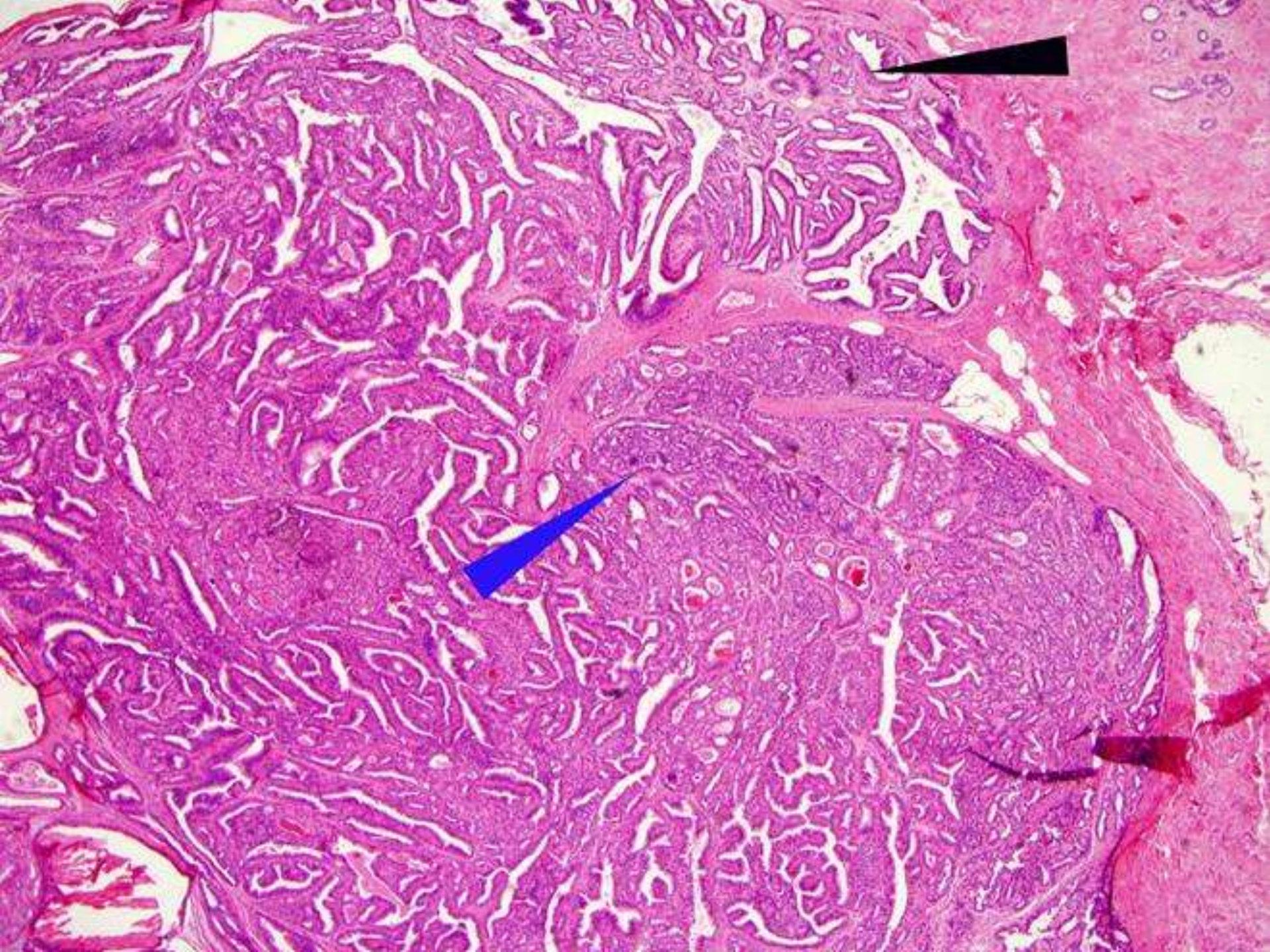


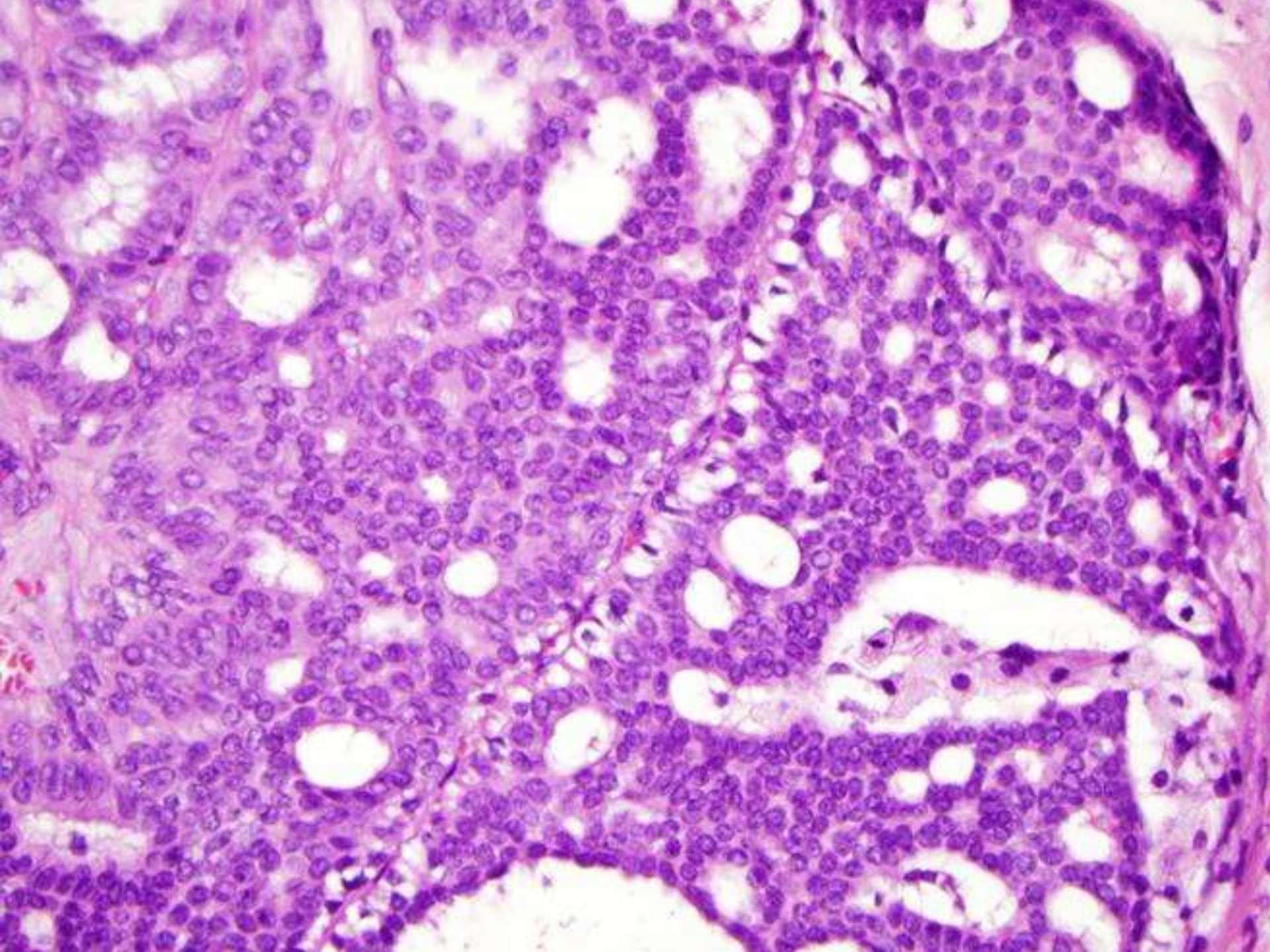


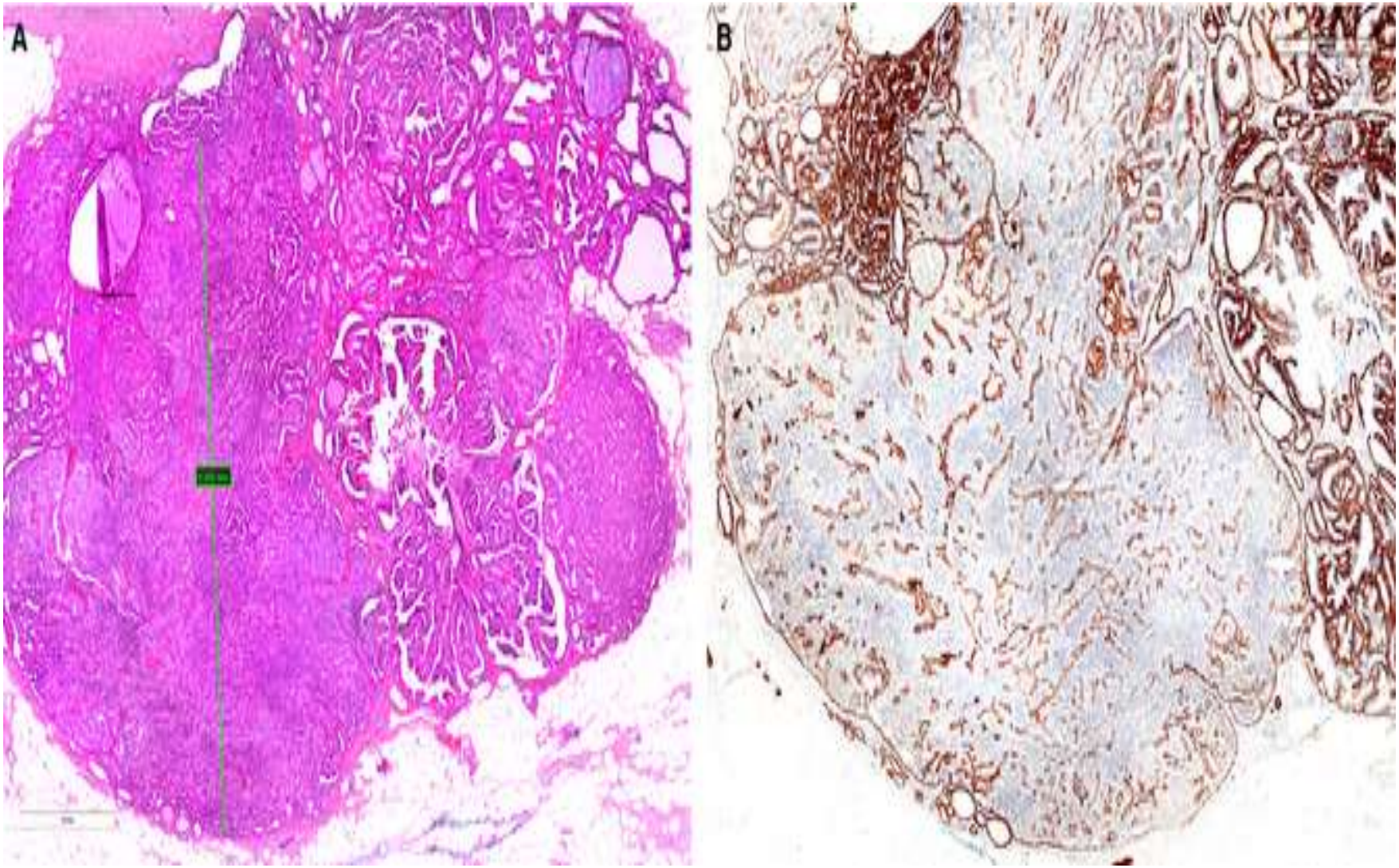
In-situ papillary lesions

Intraductal papillomas with DCIS

- Use of atypical papilloma is discouraged
- Low grade changes <3 mm ADH
- Low grade changes >3 mm DCIS
- Increased risk of subsequent invasive breast cancer
- Intermediate/High grade changes - DCIS



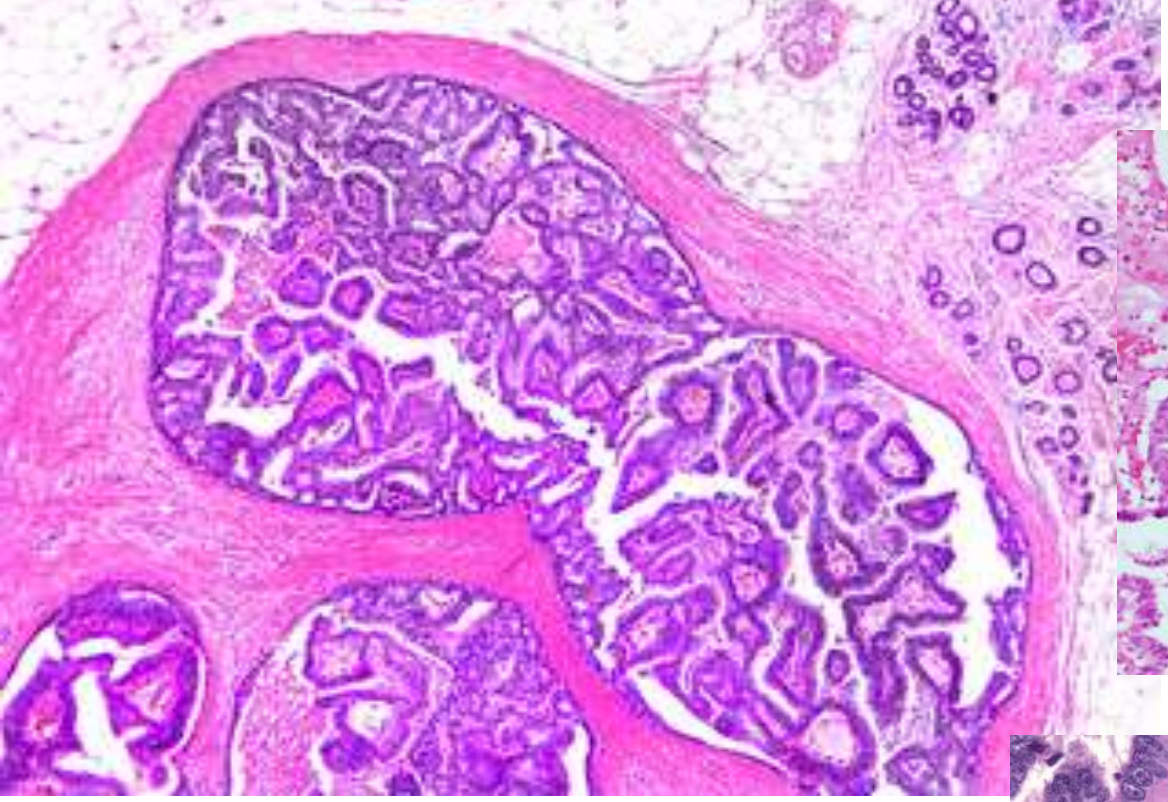


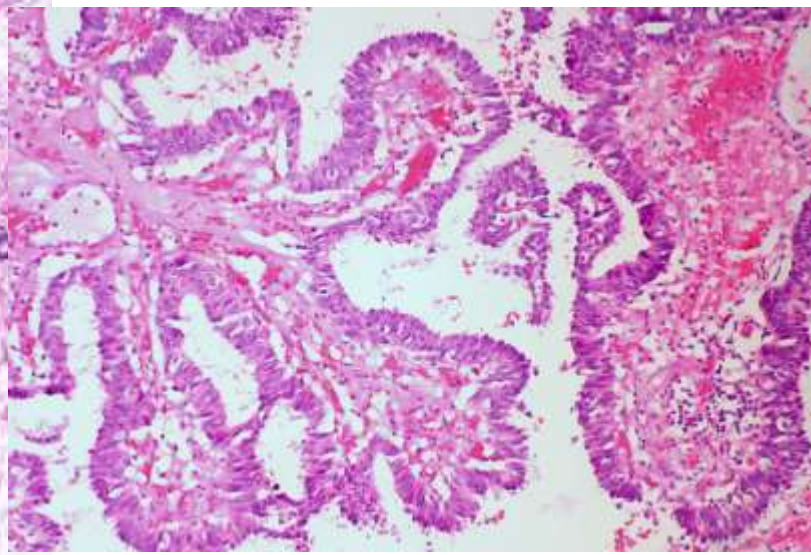

A
B

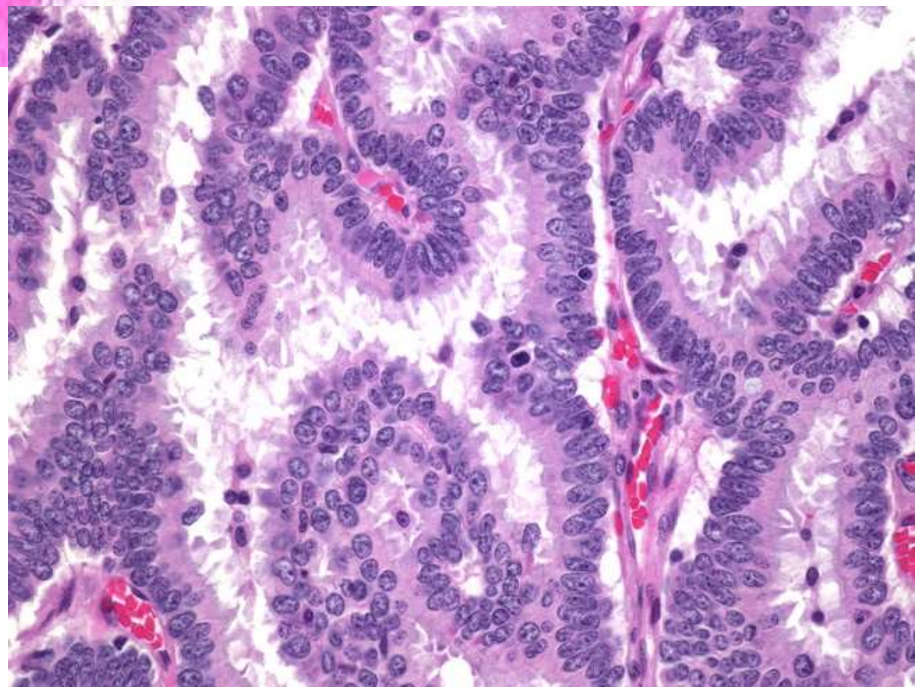
In-situ papillary lesions

Intraductal papillary carcinoma/ Papillary DCIS

- Intraductal papillary lesion with thin fibrovascular cores
- Columnar cells with nuclei aligned perpendicular to the stromal cores
- Usually lack myoepithelial cells within the lesion (although can sometimes be demonstrated)
- Myoepithelial cells are demonstrated around the lesion



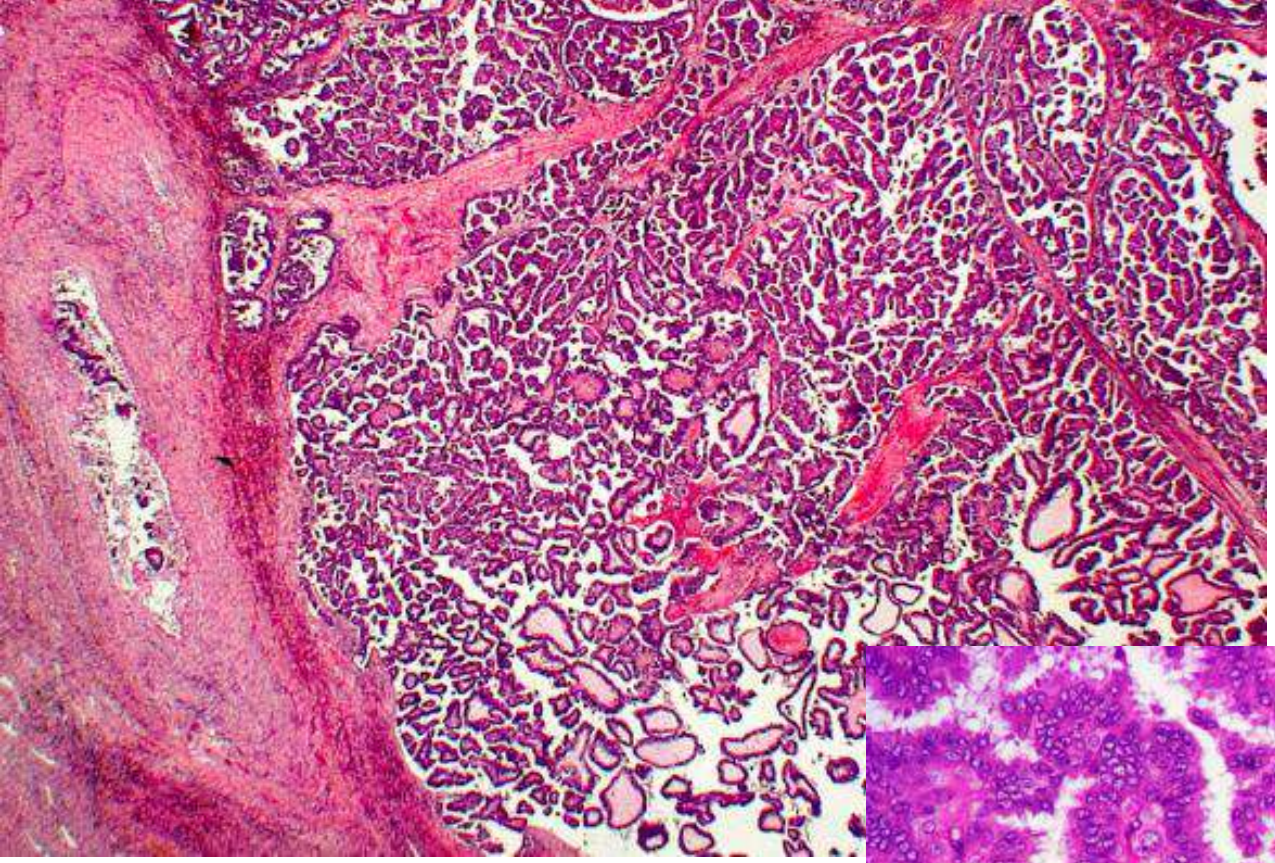


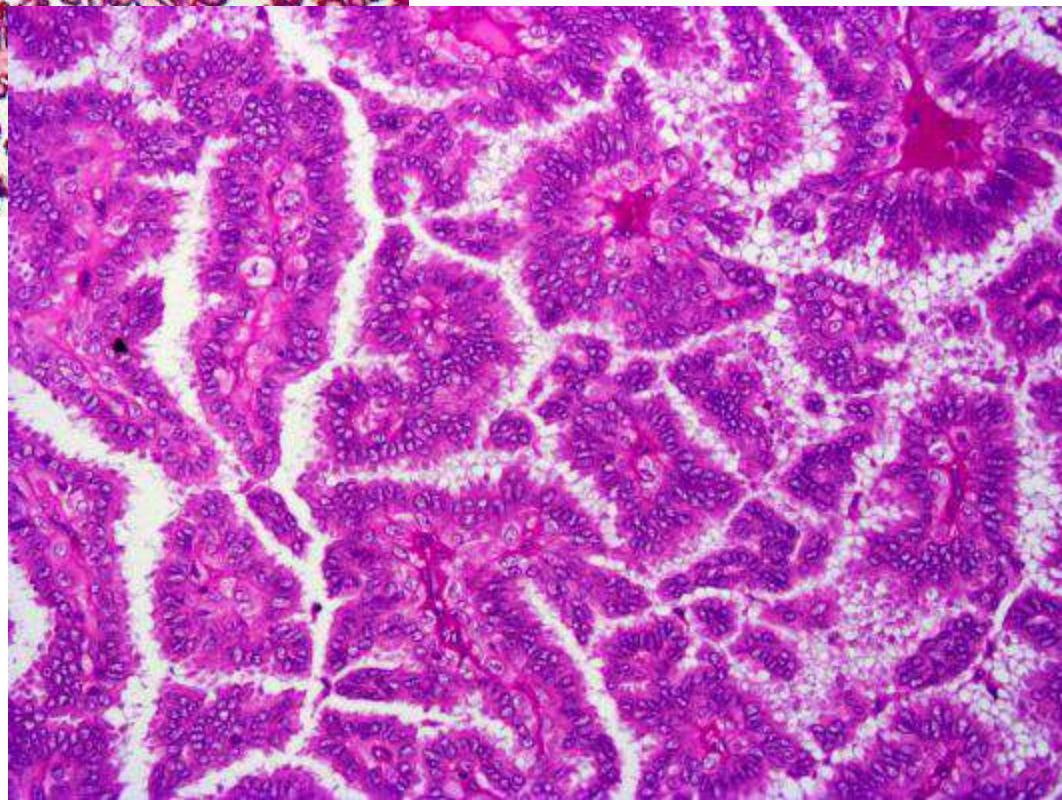


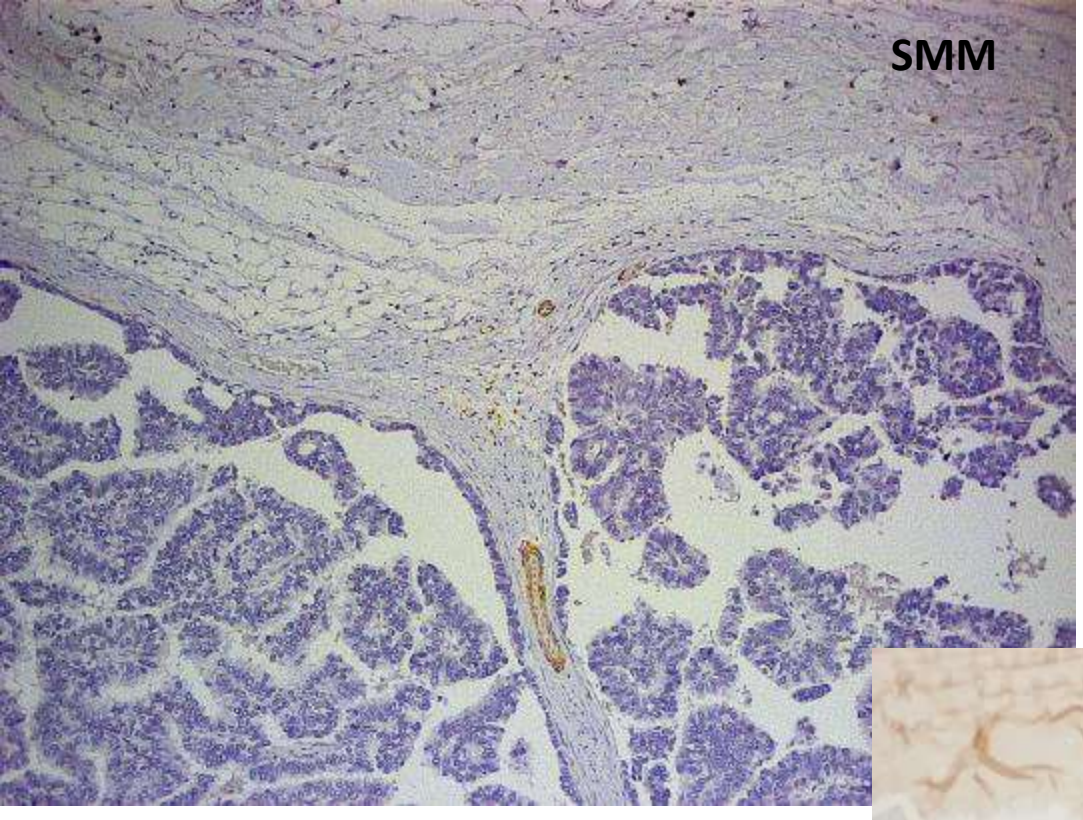
In-situ papillary lesions

Encysted/Encapsulated papillary carcinoma

- Circumscribed papillary lesion surrounded by a thick fibrous capsule
- Complete lack of myoepithelial cells



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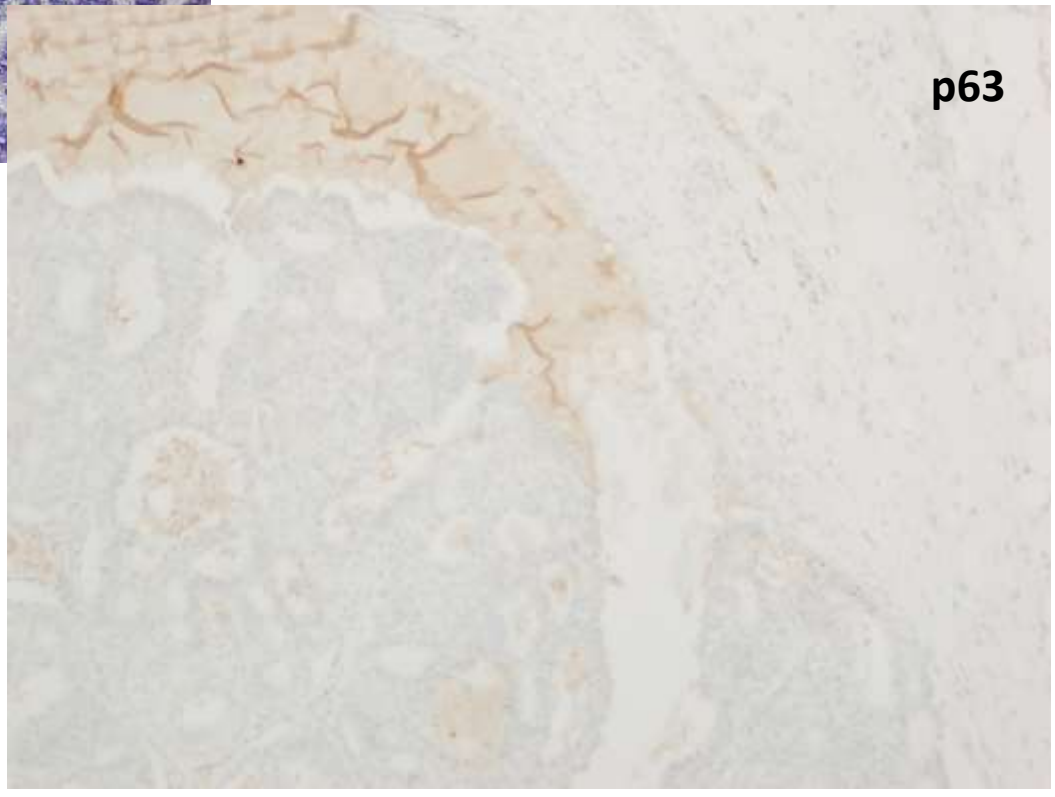
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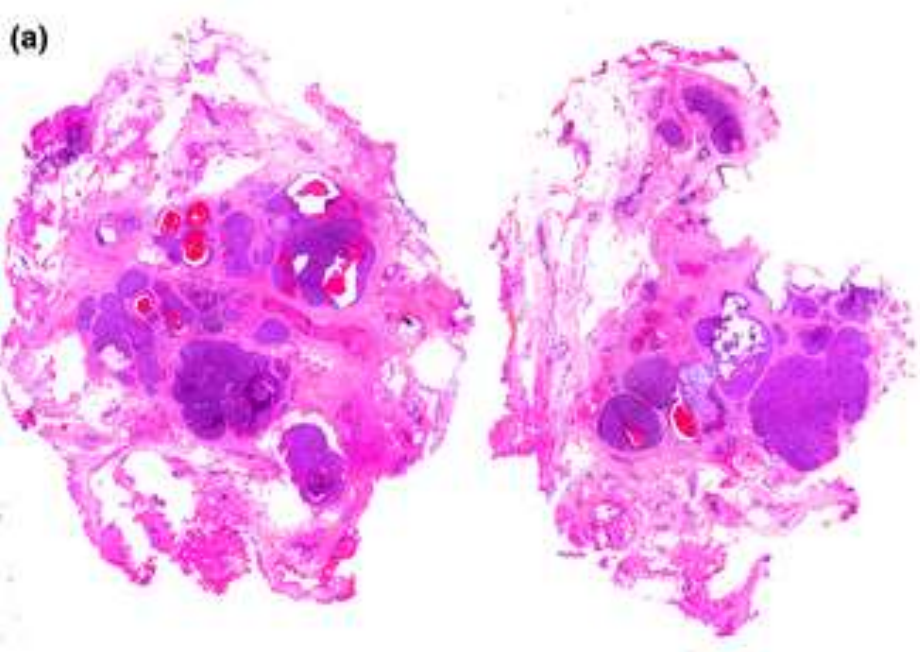
Encysted papillary carcinoma

- Ongoing debate regarding the true biological state
- Some of these lesions are probably low grade carcinomas growing with expansile edges
- However managed as in-situ lesions as behaviour is similar to DCIS

In-situ papillary lesions

Solid papillary carcinoma

- Single or multiple nodules
- Usually multiple expansile cellular nodules with smooth contours
- Solid papillary growth pattern
- Myoepithelial cells can be demonstrated at the periphery
- More commonly associated with invasive component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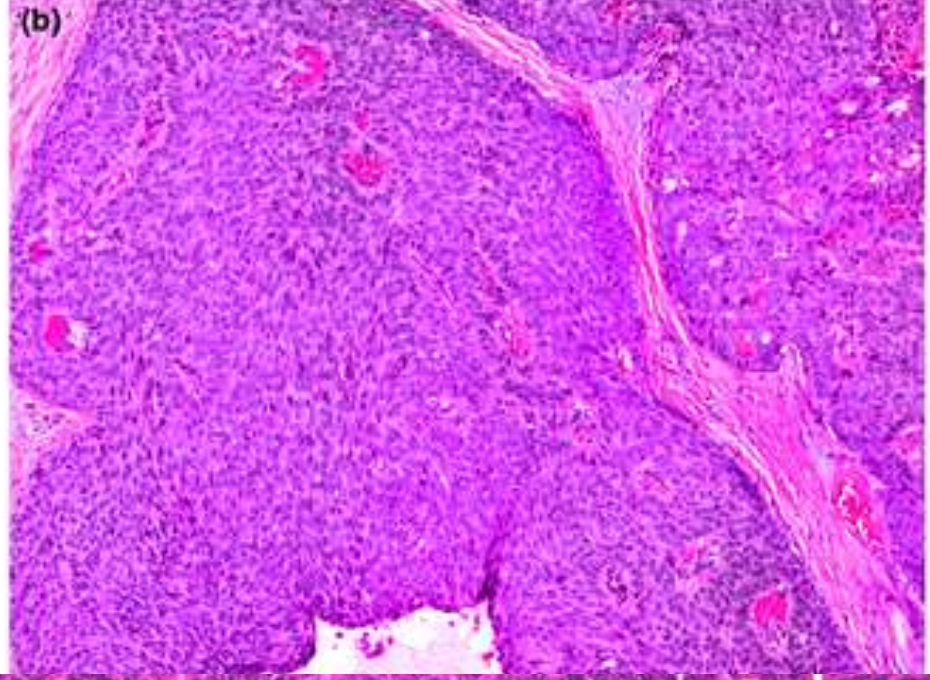
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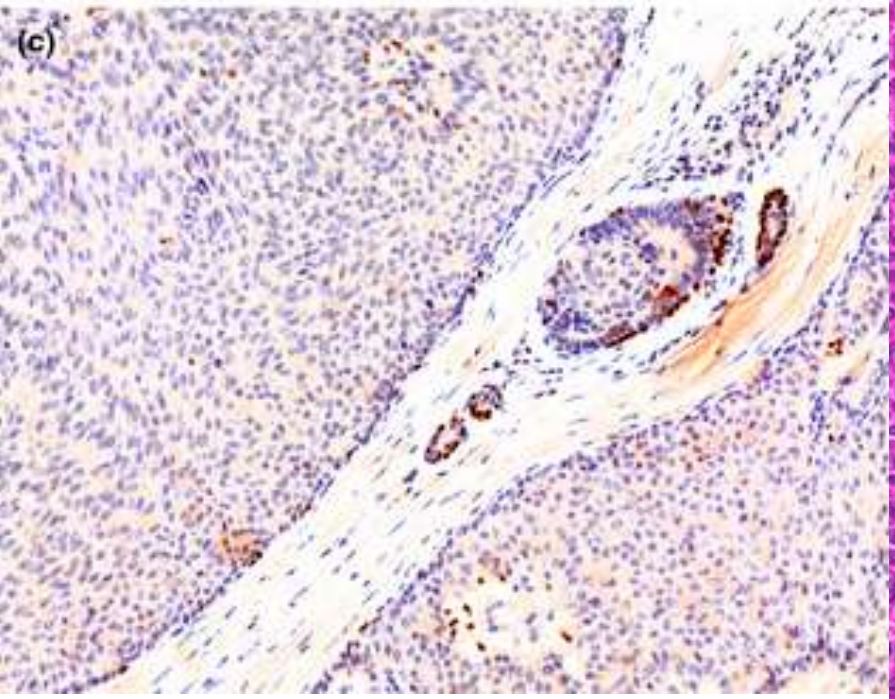
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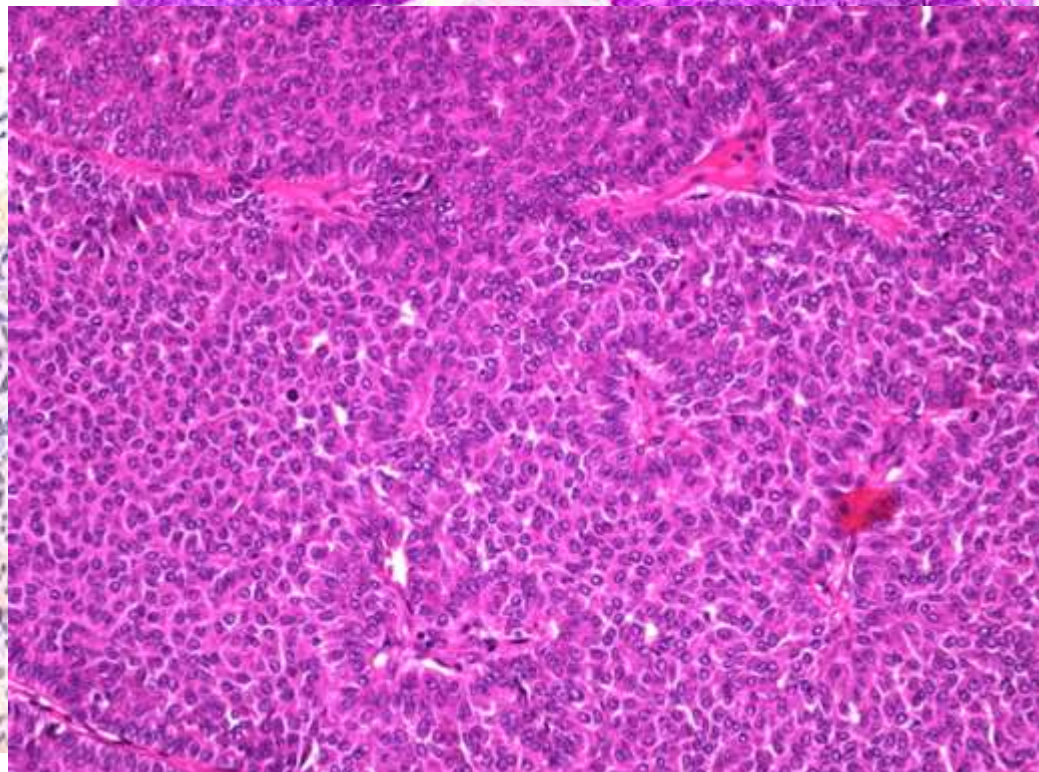


Solid papillary carcinoma

- In cases lacking mantle of myoepithelial cells – if the tumour islands are irregular with jagged edges and surrounded by desmoplastic stroma, consider diagnosis of invasive malignancy

In-situ papillary lesions

When diagnosis of papillary carcinoma is made, it is imperative to clarify in the report whether the lesion is in-situ or invasive

Reference

Review article

Papillary and neuroendocrine breast lesions: the WHO stance. Tan PH et al, Histopathology, 2015, 66, 761-770

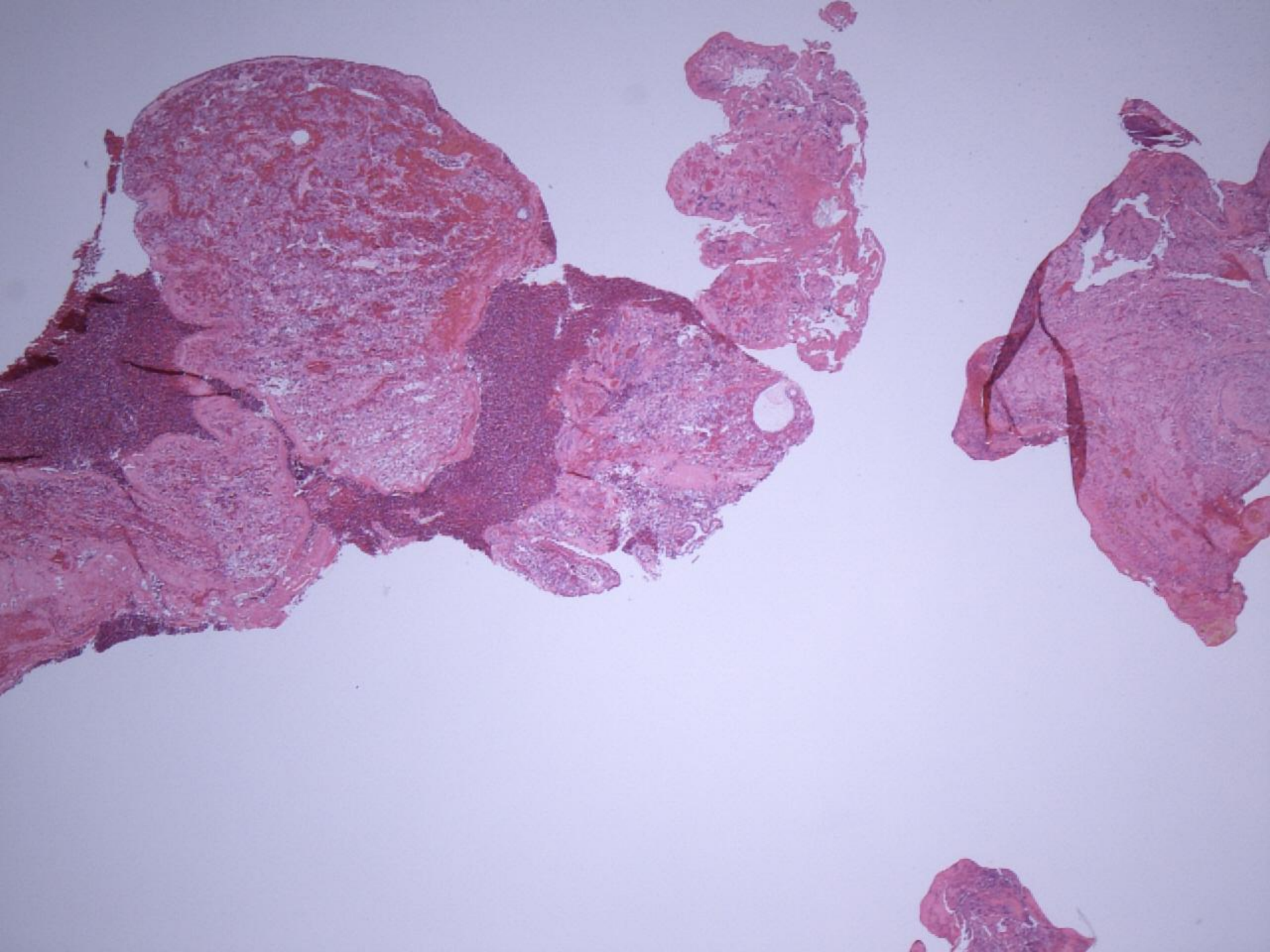
Thank you

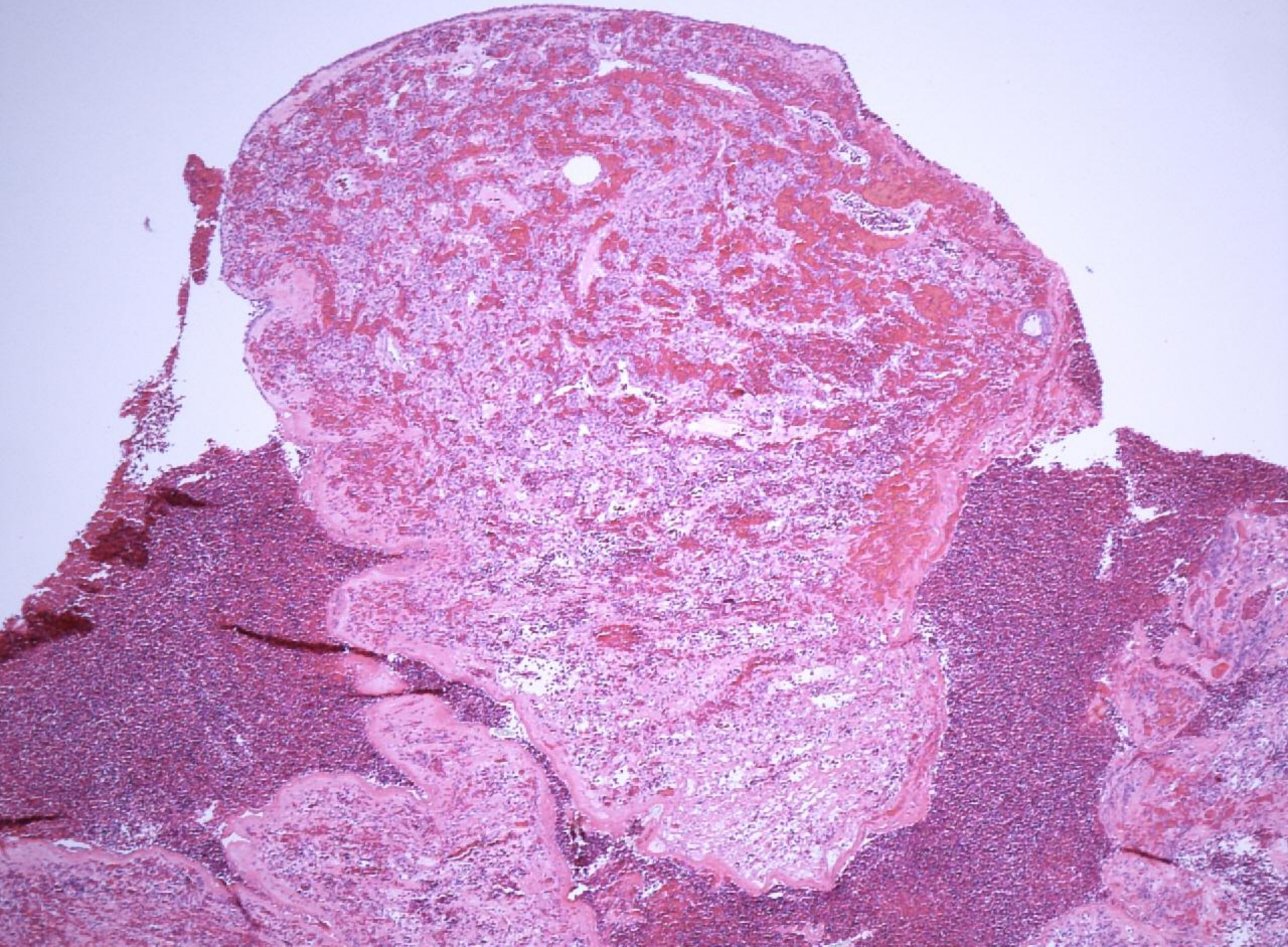
GENERAL EQA CIRCULATION 43 Educational Cases E3 & E4

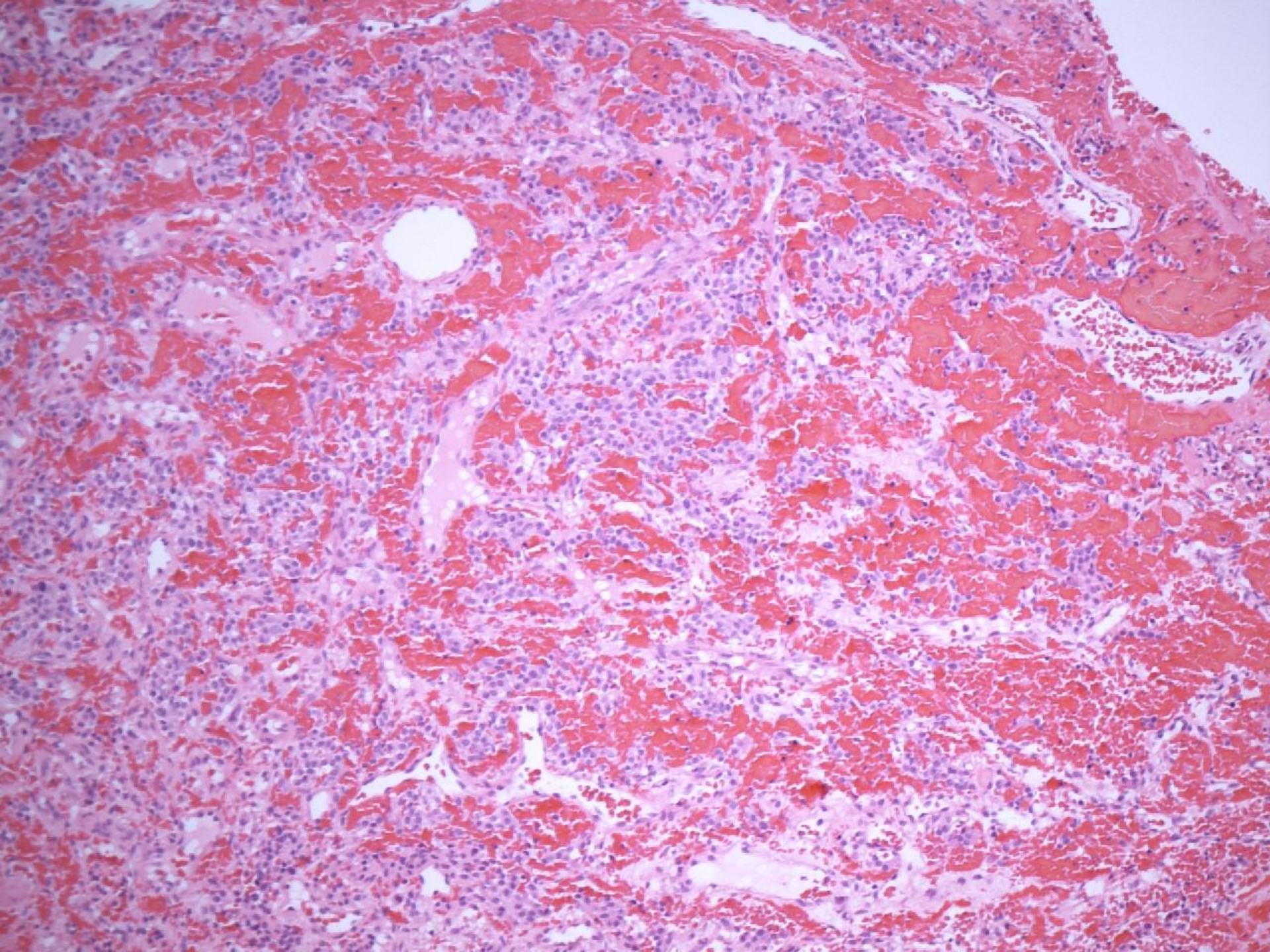
Dr John Robert Millar
Monklands General Hospital

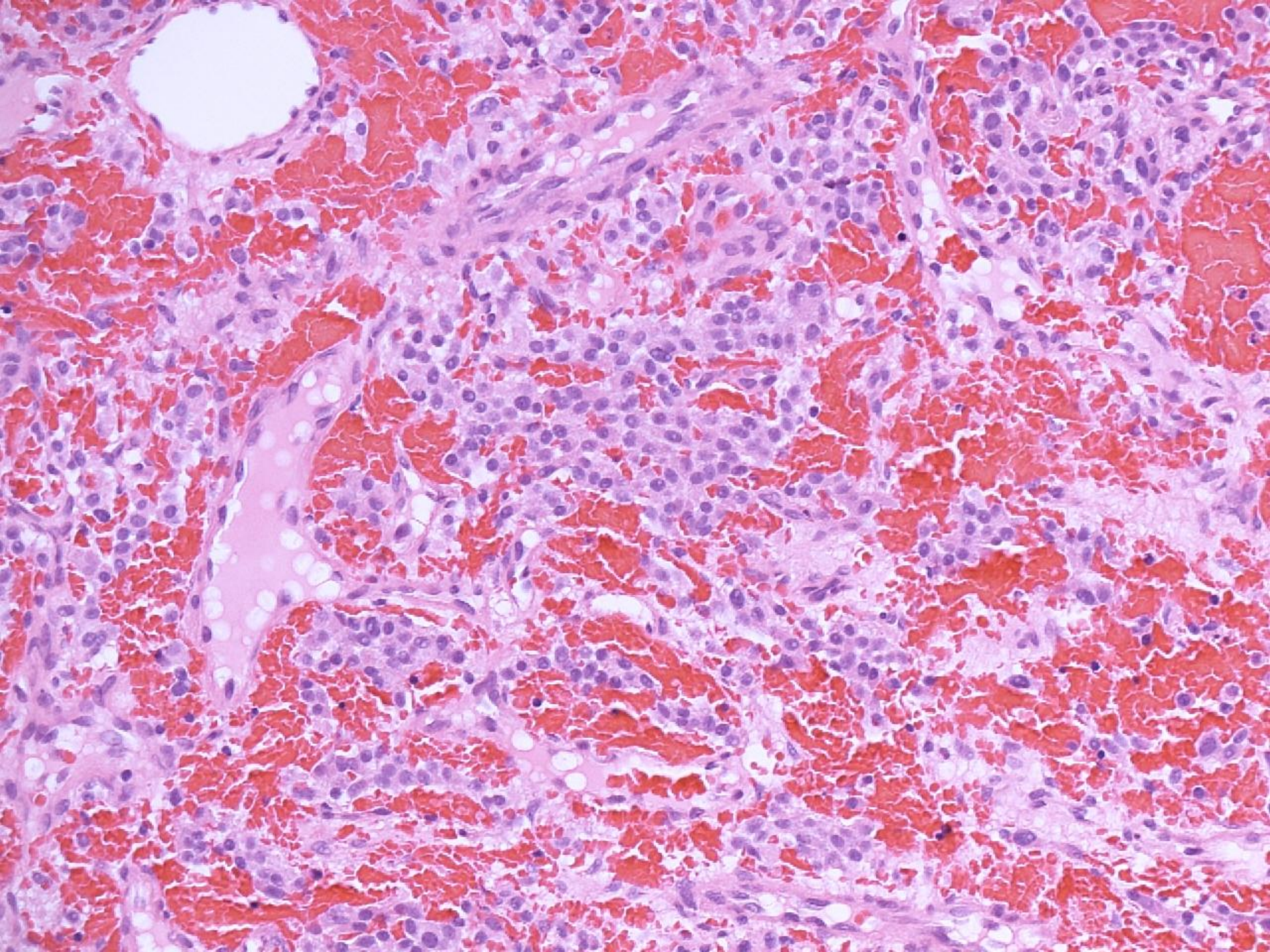
CASE E3

- F 35
- Hx of pulsatile tinnitus
- Bx red mass behind eardrum









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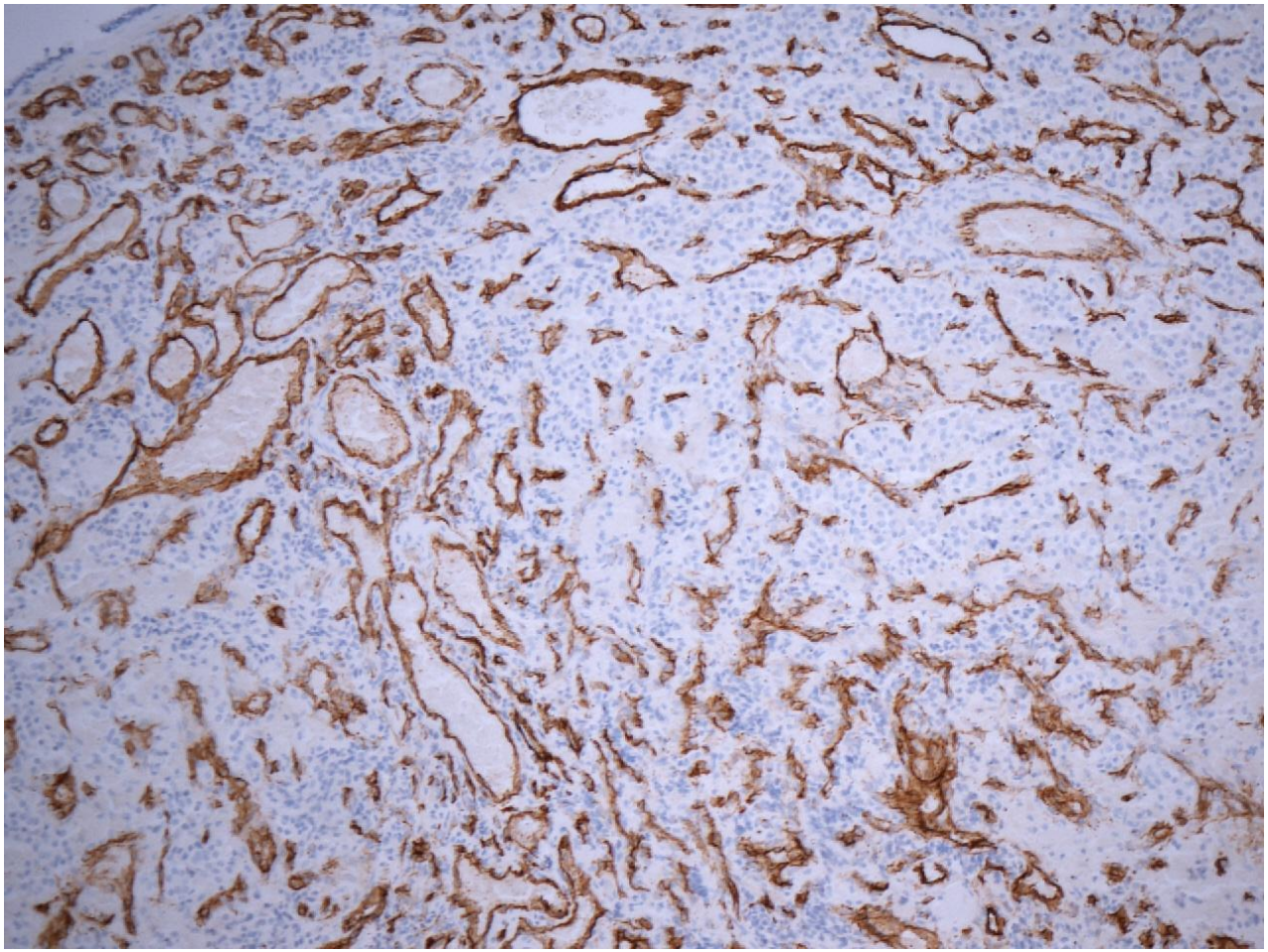
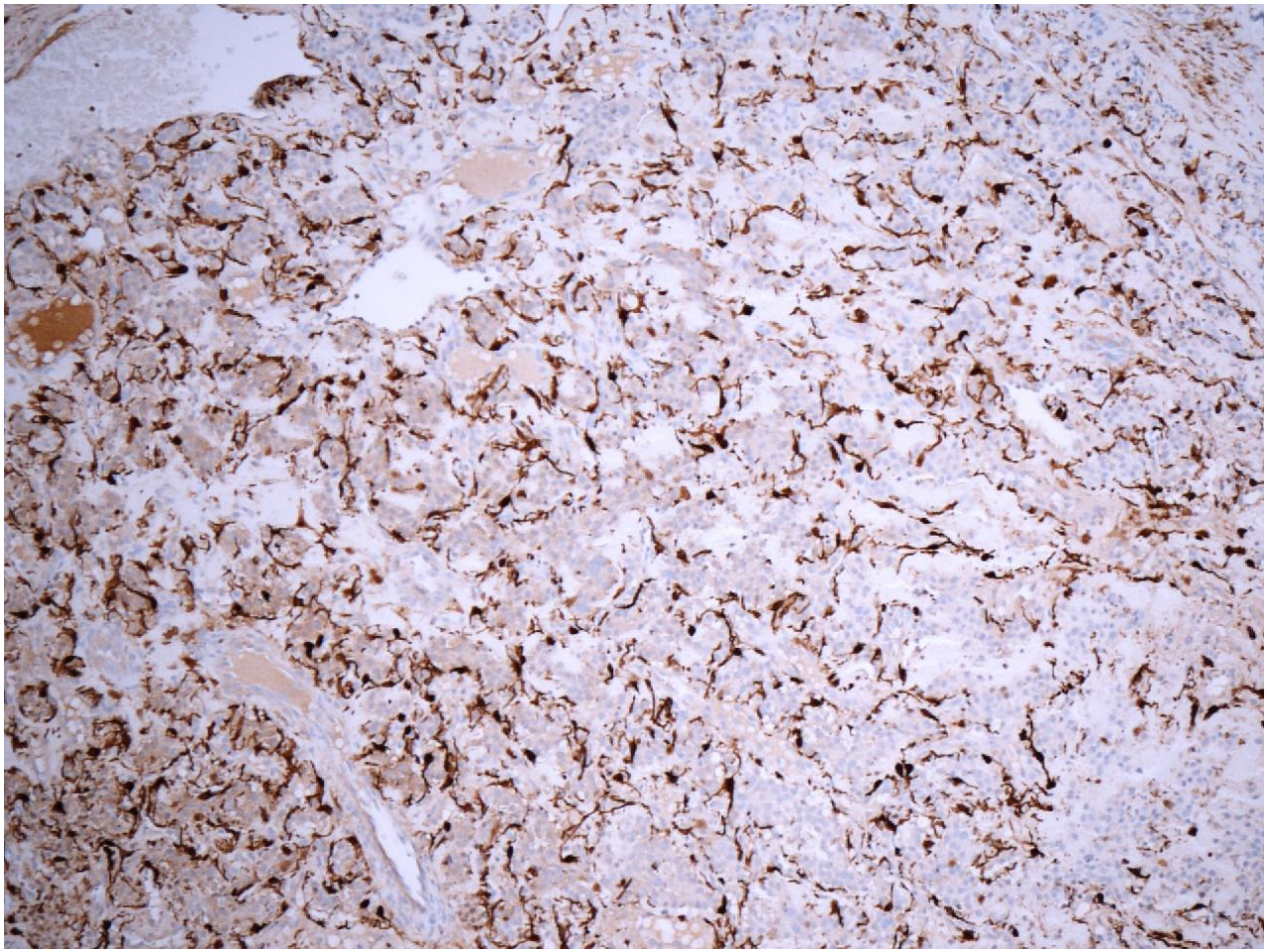
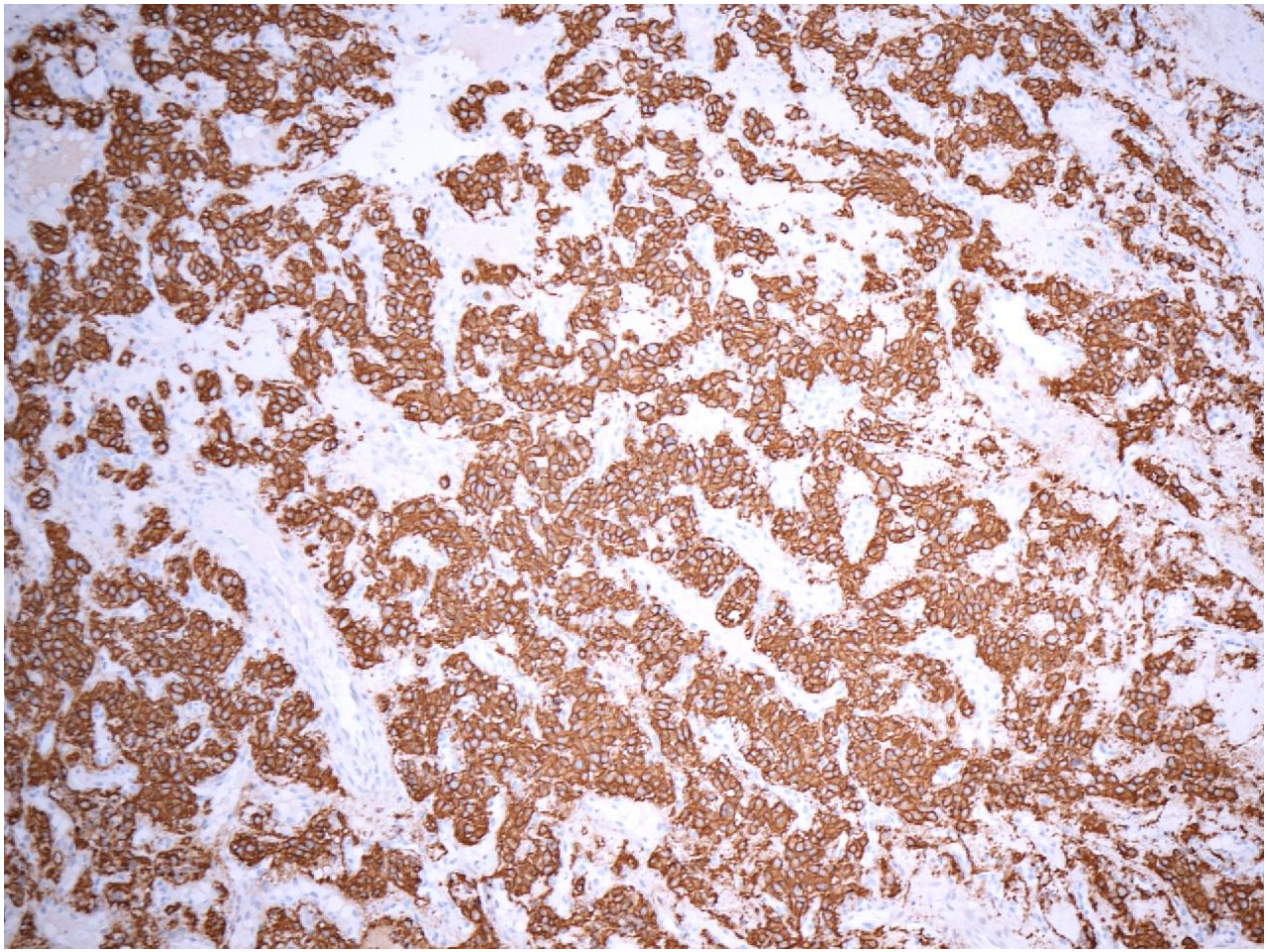


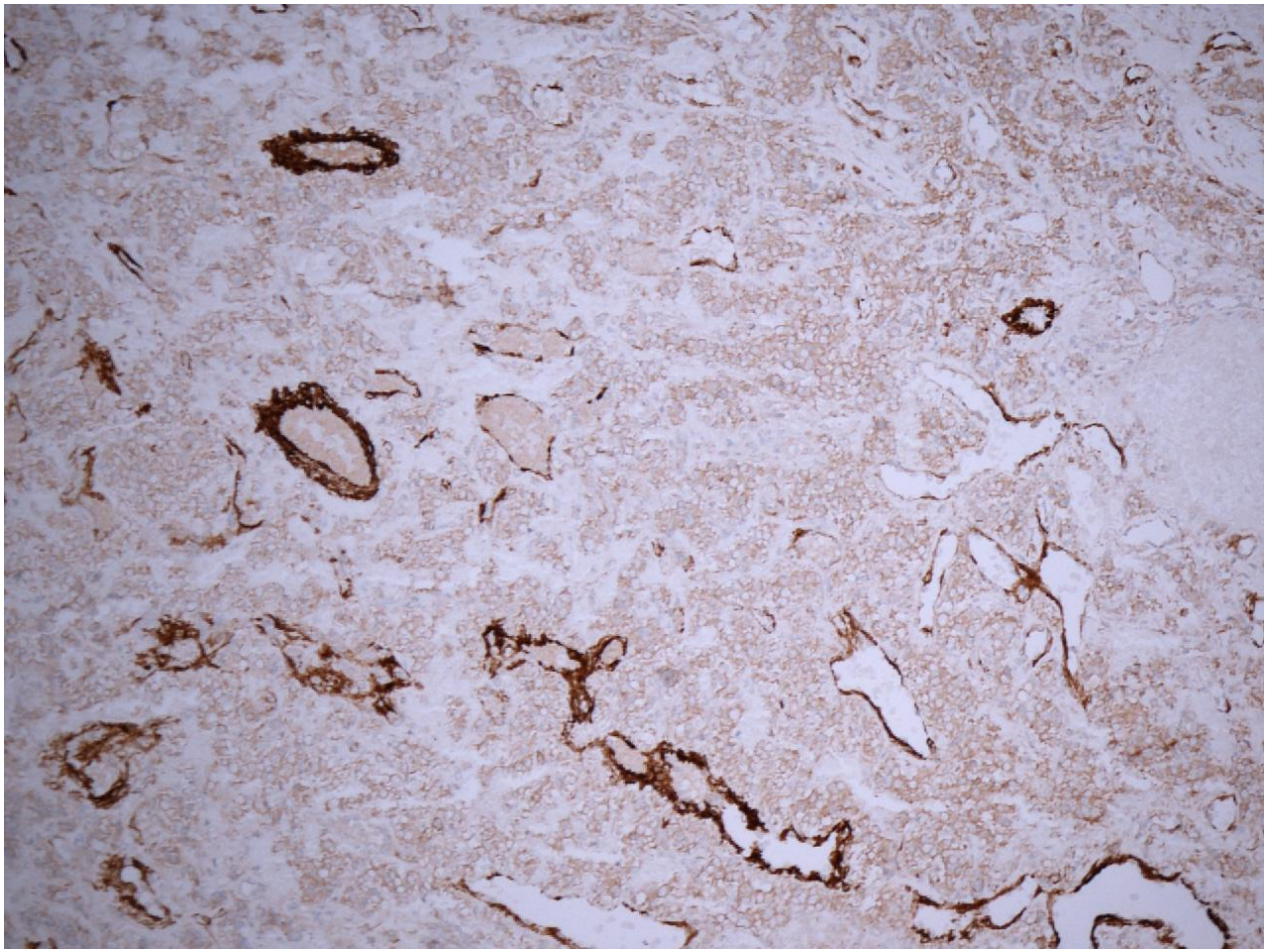
S100



CD56



SMA



ANSWERS

- Jugulotympanic paraganglioma, paraganglioma (57)
- Glomus tympanicum (2)
- Glomus tumour/glomangioma (27)
- Haemangioma (8)
- Carotid body paraganglioma (1)
- Glomus carotid body tumour (2)
- Adenoma (1)
- Pecoma (1)

TOTAL: 99

JUGOLOTYMPANIC PARAGANGLIOMA

- Also called glomus jugulare tumour or glomus tympanicum tumour
- Most common tumour of middle ear
- Usually women, ages 40-69 years
  - 85% arise in jugular bulb, causing mass in middle ear or external auditory canal
  - 12% arise from tympanic branch of glossopharyngeal nerve, causing middle ear mass;
  - 3% arise from posterior auricular branch of vagus nerve, causing external auditory canal mass
- Usually cause conductive hearing loss/tinnitus
- Tumours are fed by branches of nearby large arteries; may bleed profusely at biopsy
- Histology usually benign, but this does not predict behaviour

DIAGNOSTIC FEATURES

- Classic organoid (zellballen) or nesting pattern of paragangliomas with central round/oval chief cells containing abundant eosinophilic granular or vacuolated cytoplasm, uniform nuclei with dispersed chromatin

- Sustentacular cells (spindled, basophilic, difficult to see with H&E) are present at periphery of nests

- Prominent fibrovascular stroma separates nests

- No glandular or alveolar differentiation, although alveolar pattern like in middle ear adenoma has been described

IMMUNOHISTOCHEMISTRY

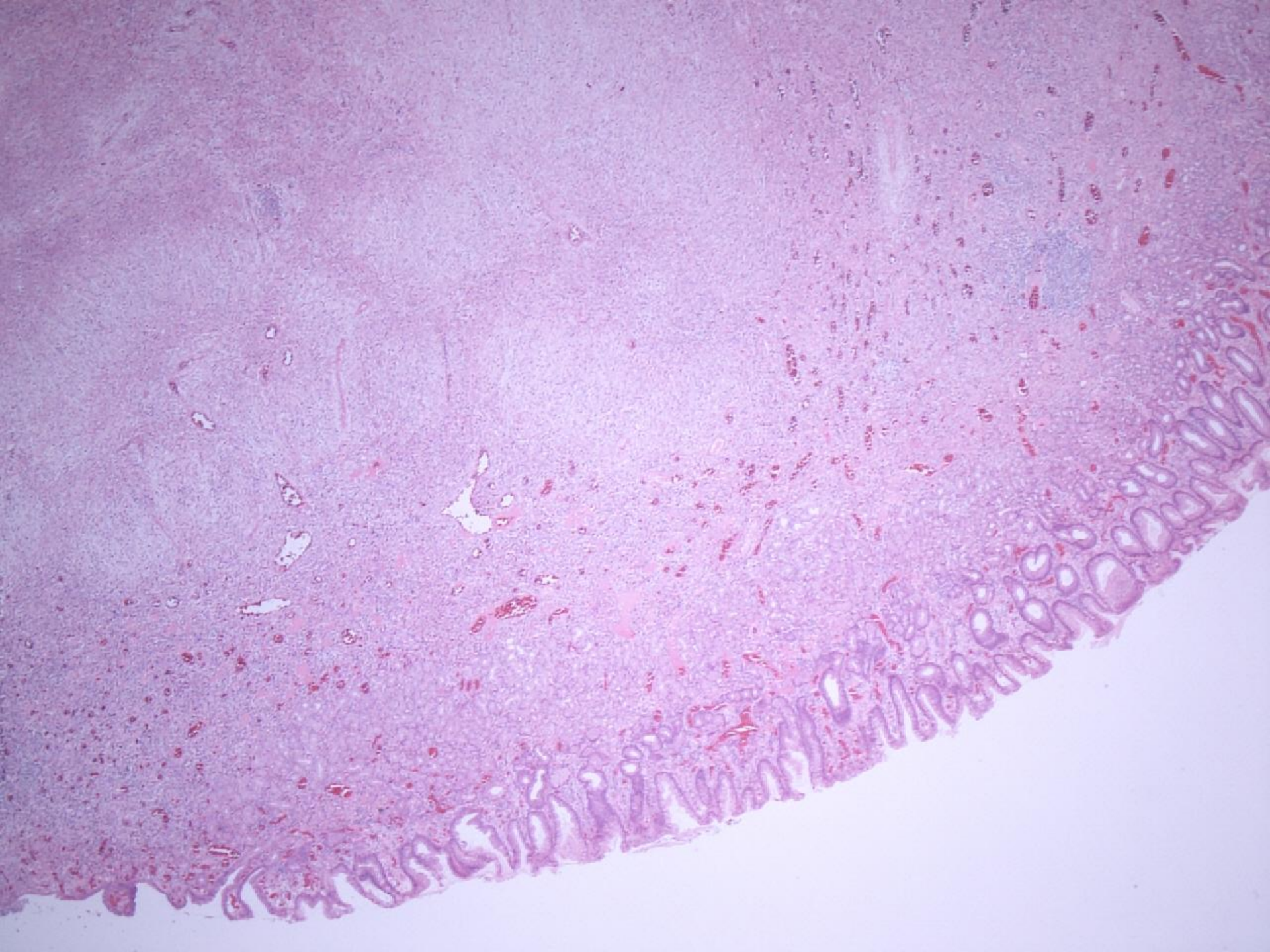
- Chromogranin and synaptophysin+ (chief cells), S100+ (sustentacular cells)
- Reticulin+ (stains stroma and delineates nesting pattern, particularly helpful with crushed specimens)
- Keratin, EMA, HMB45, desmin/other myogenic markers, PAS, mucicarmine -

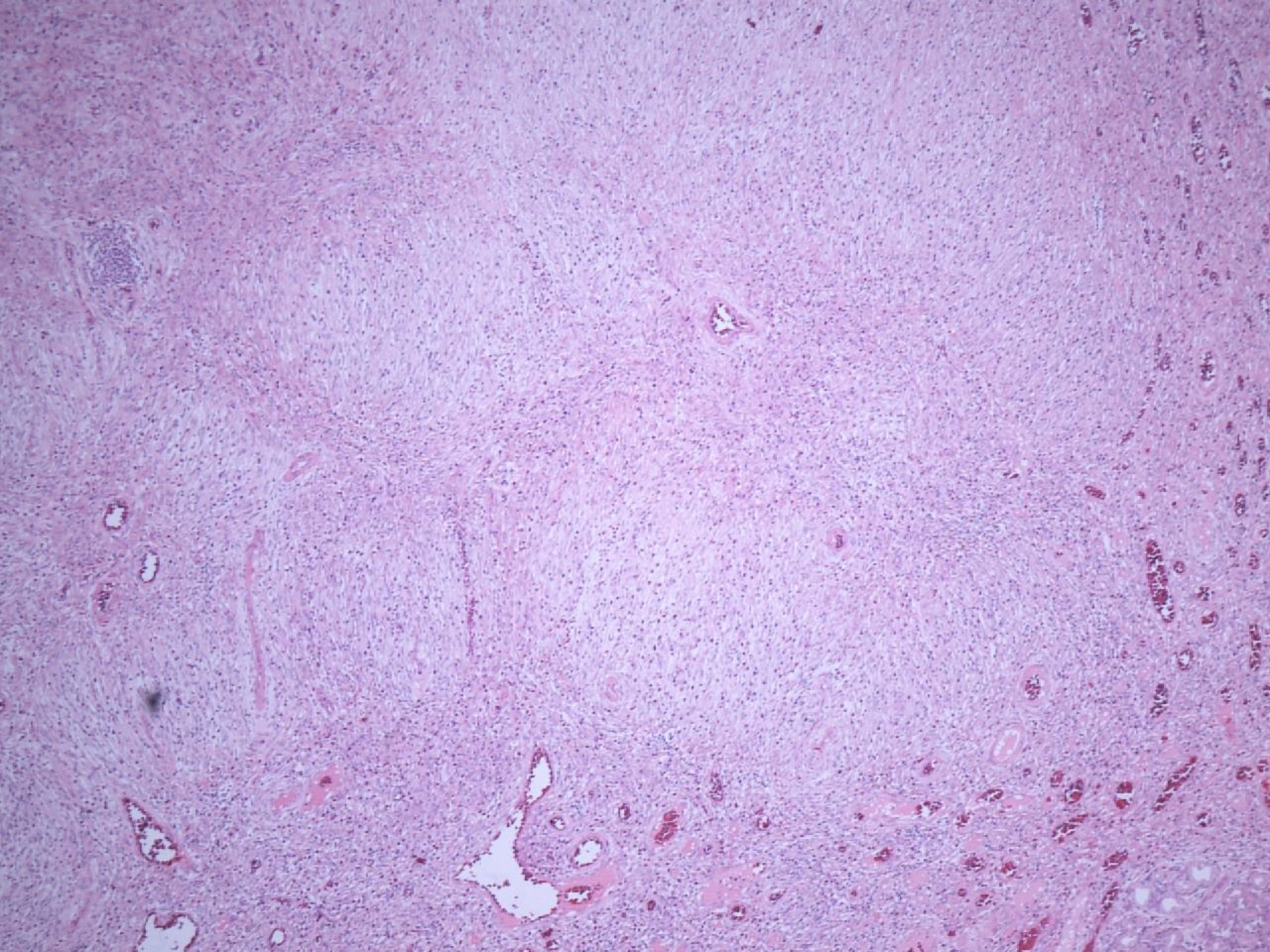
DIFFERENTIAL

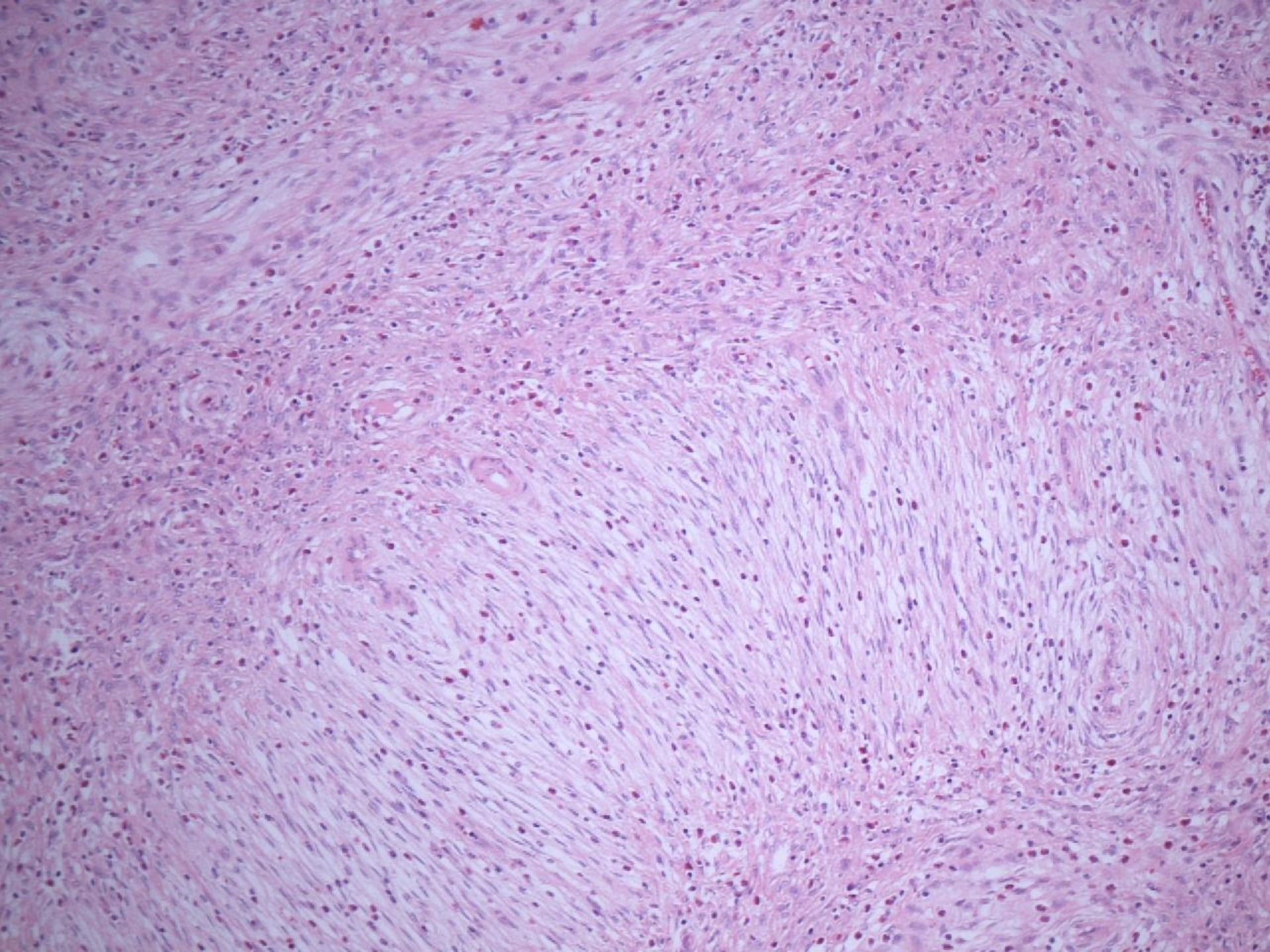
- Middle ear adenoma (glandular & NE differentiation, keratin/CK7/chromo+, intraluminal mucin+, non-vascular)

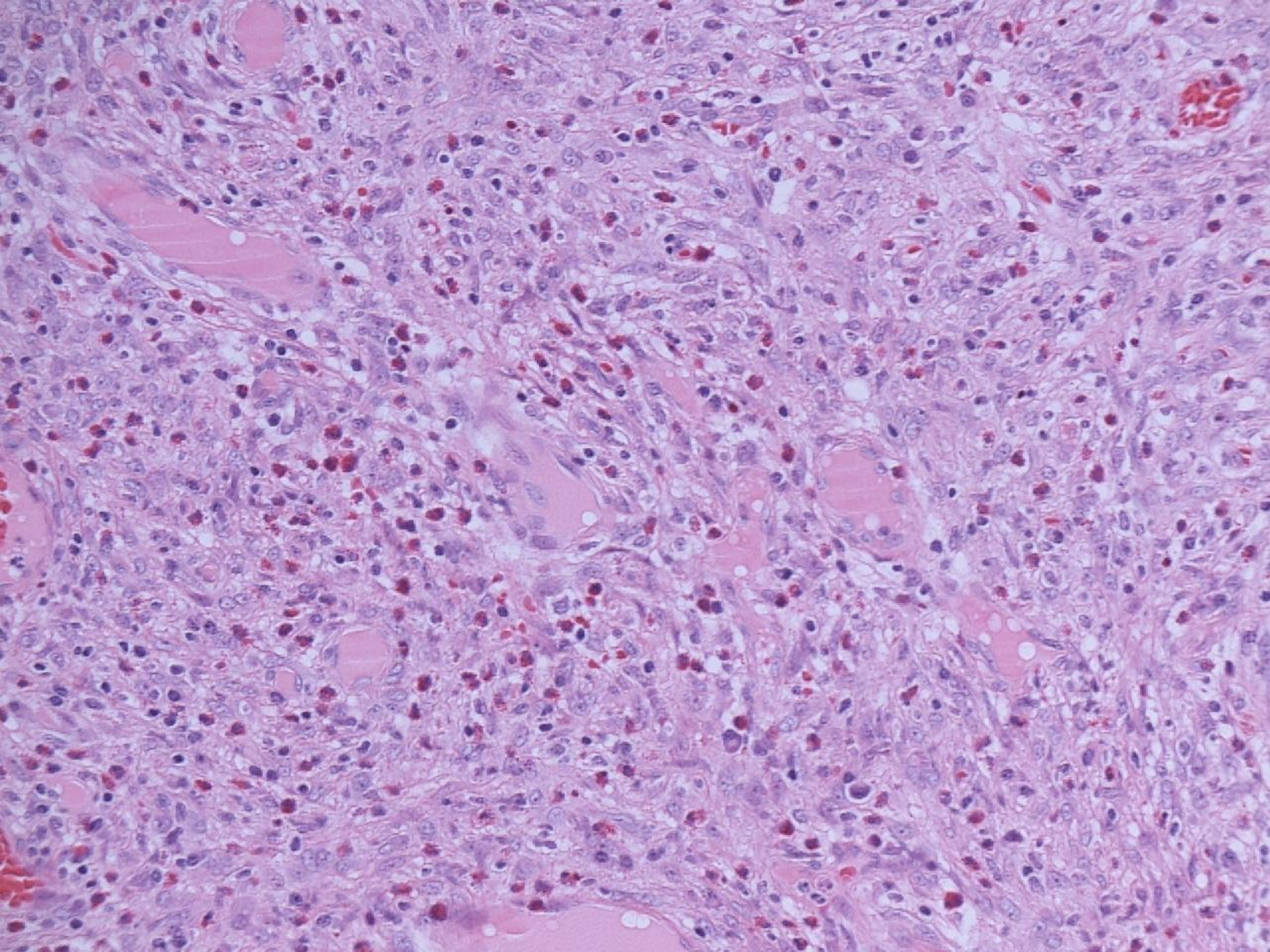
CASE E4

- F 79
- Large polyp prepyloric area at endoscopy

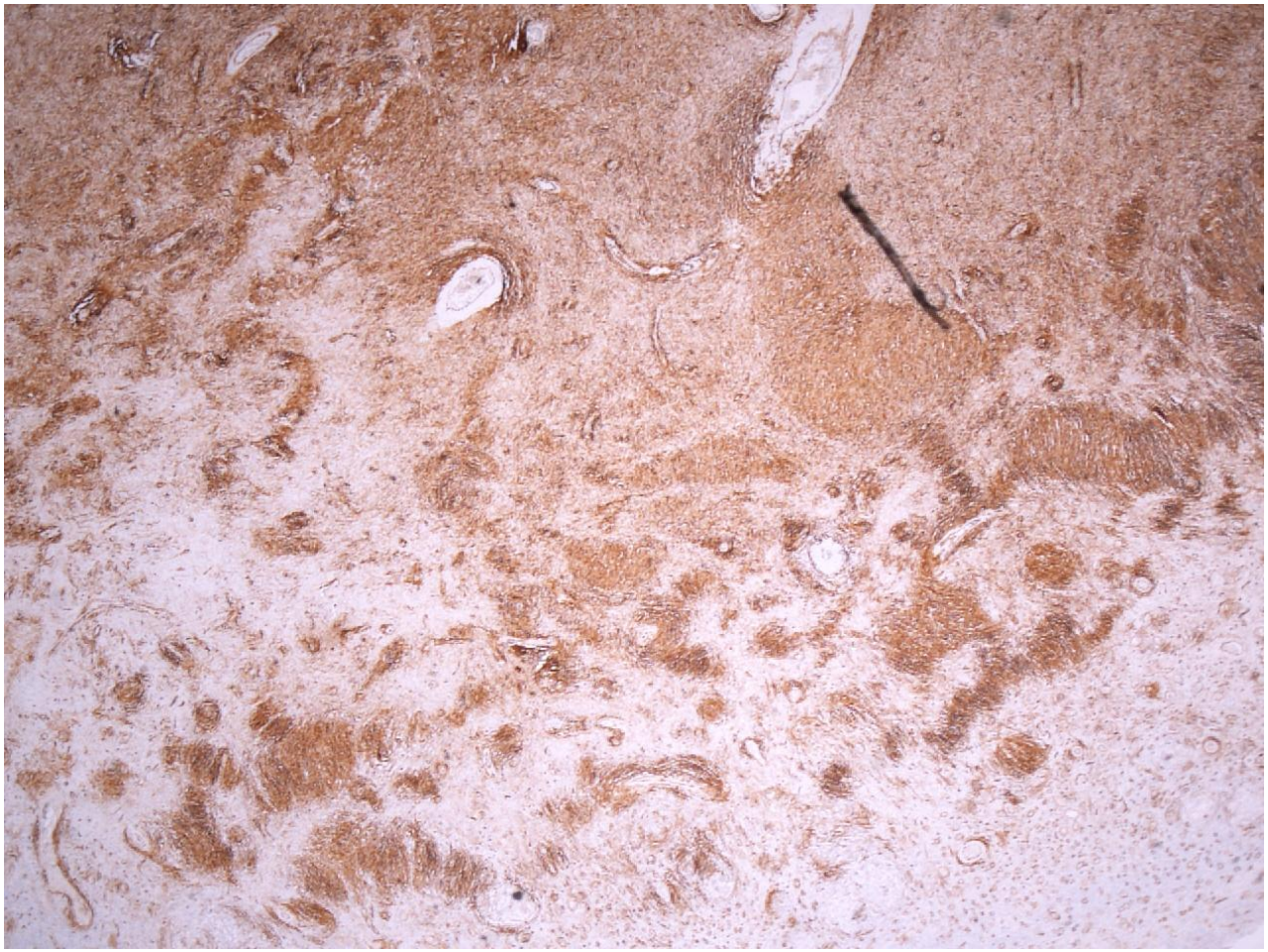




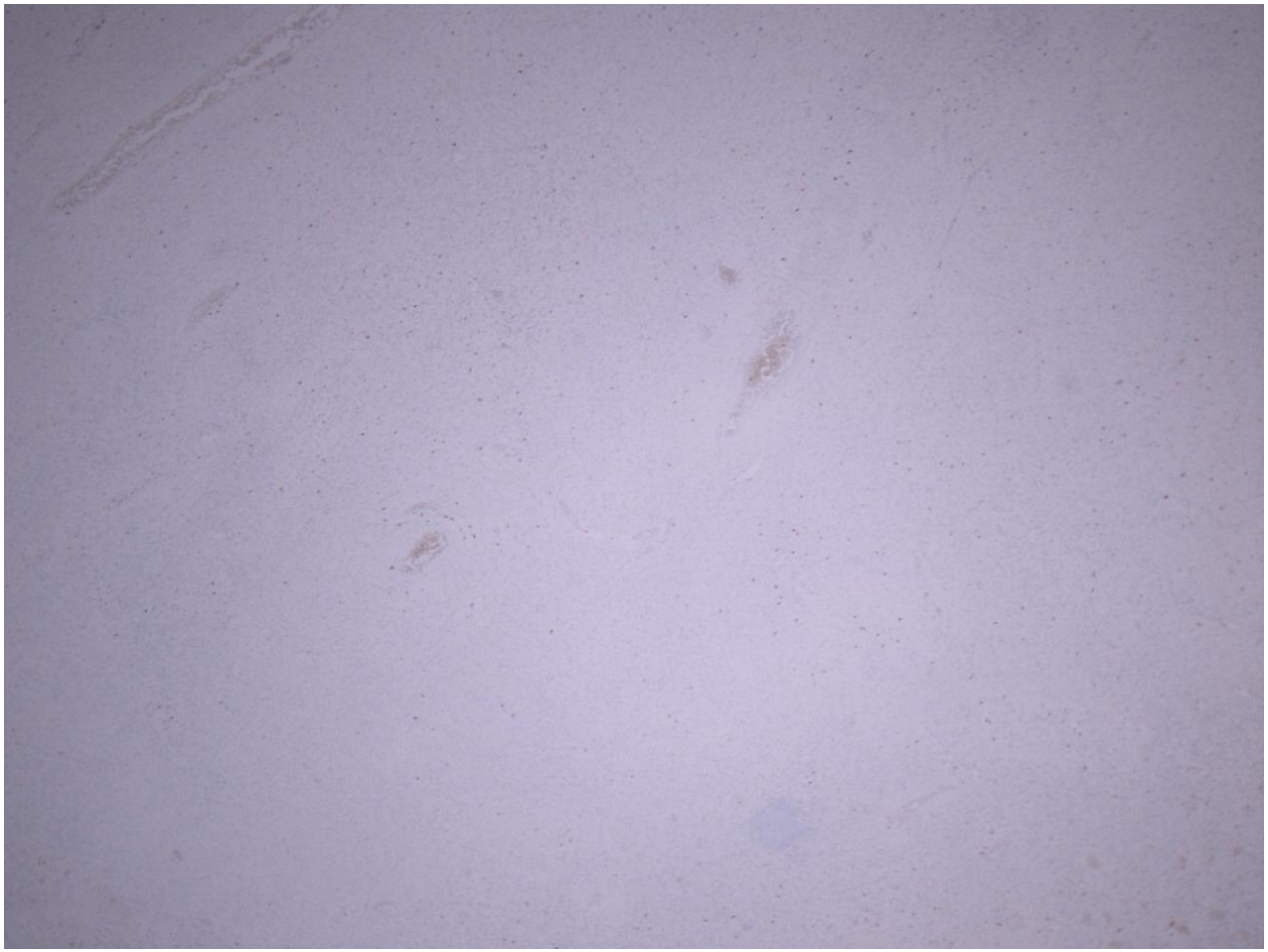




CD34



CD117



ANSWERS

- Inflammatory fibroid tumour/myofibroblastic (90)
- GIST (2)
- Inflammatory pseudopolyp/tumour (3)
- Eosinophilic granuloma (1)
- NF (1)
- Hamartoma (1)
- Schwannoma (1)

TOTAL: 99

INFLAMMATORY FIBROID TUMOUR/POLYP

- Gastrointestinal tract tumour characterised by spindle and stellate cells set in an inflammatory, myxoid stroma

- Most common in antrum, followed by small intestine

- 3rd to 8th decades of life (mean age 60)
  - May present with intussusception, obstruction, bleeding
  - Infrequently recurs
  - No metastases or local aggressive recurrences

- Most are semi-pedunculated polyps arising in the submucosa
  - Covered by mucosa or may be eroded
  - Occasional tumours may be restricted to the lamina propria and muscularis mucosae
  - Larger tumours may extend into muscularis propria
  - Most <5 cm, rarely up to 20 cm

DIAGNOSTIC FEATURES

- **Composed of bland, uniform spindled/stellate cells**
  - The lesional cells may be lost in the background and difficult to identify
  - Multinucleated giant cells in 1/3 of cases

- **Loose fibromyxoid background with regular vascular pattern**
  - Regular small to medium sized vessels throughout
  - May have granulation tissue appearance

- **Eosinophil rich mixed inflammatory infiltrate**
  - Also includes lymphocytes, plasma cells, macrophages, mast cells
  - Lymphoid aggregates may be seen

- **Frequent whorled, concentric "onion skin" pattern centred on blood vessels and glands**
  - 10% of cases may lack this pattern, but may be accentuated by CD34

DIFFERENTIAL

- **GIST** – CD117+, infrequent eosinophils, lacks regular vascular pattern
- **Solitary fibrous tumour –** arises in serosa, ropey collagen, inflammation infrequent
- **Schwannoma** – peripheral lymphoid cuff, lacks regular vascular pattern
- **Inflammatory myofibroblastic tumour –** children, plasma cells >eosinophils, desmin/keratin/ALK1+, CD34 -, lacks regular vascular pattern, nuclear pleomorphism
- **Leiomyoma** – desmin +, CD34-, infrequent eosinophils