Scotland & Ireland EQA scheme of General Histopathology Educational cases Circulation 51

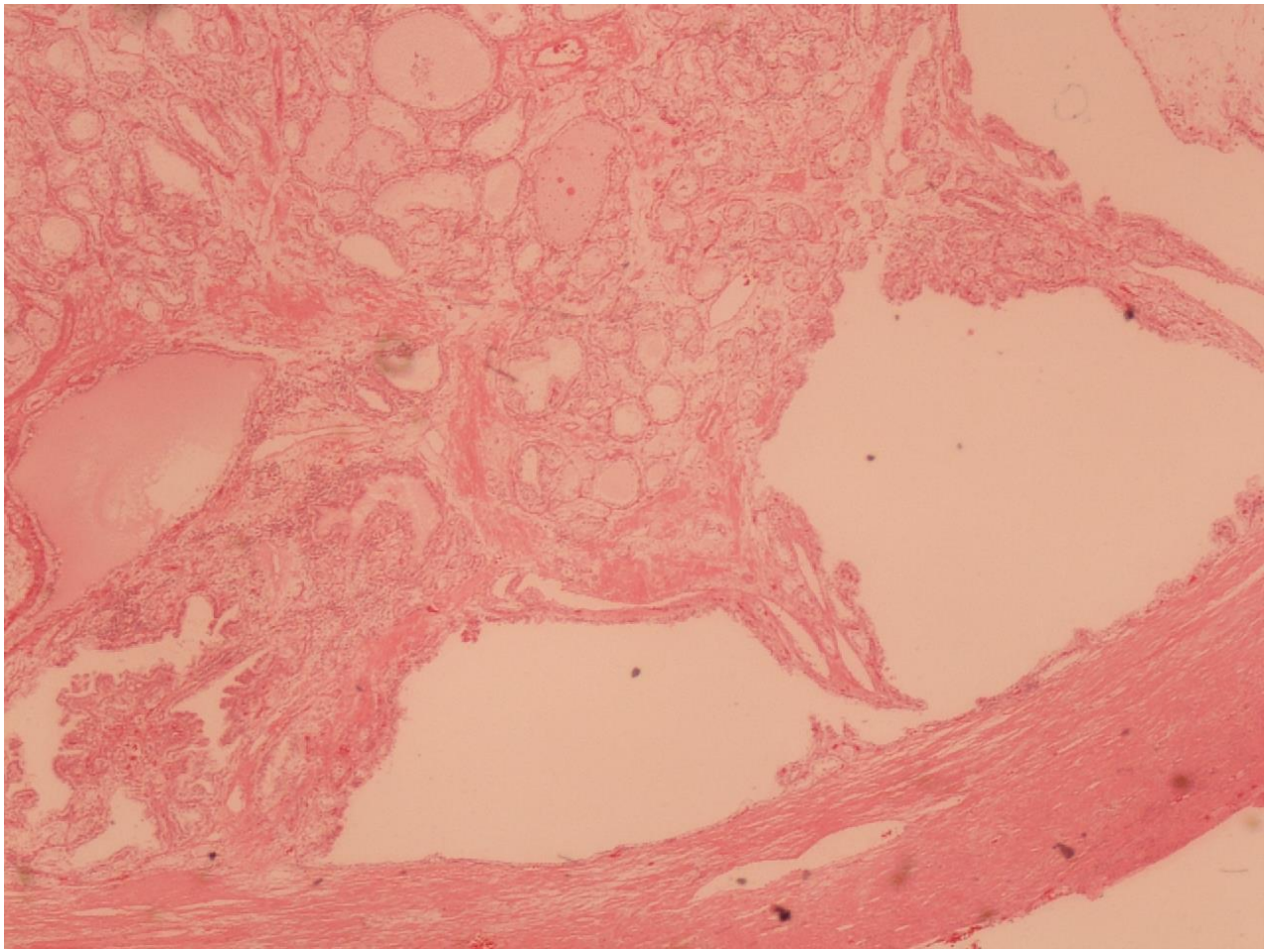
Dr Yvonne Woods

25/11/20

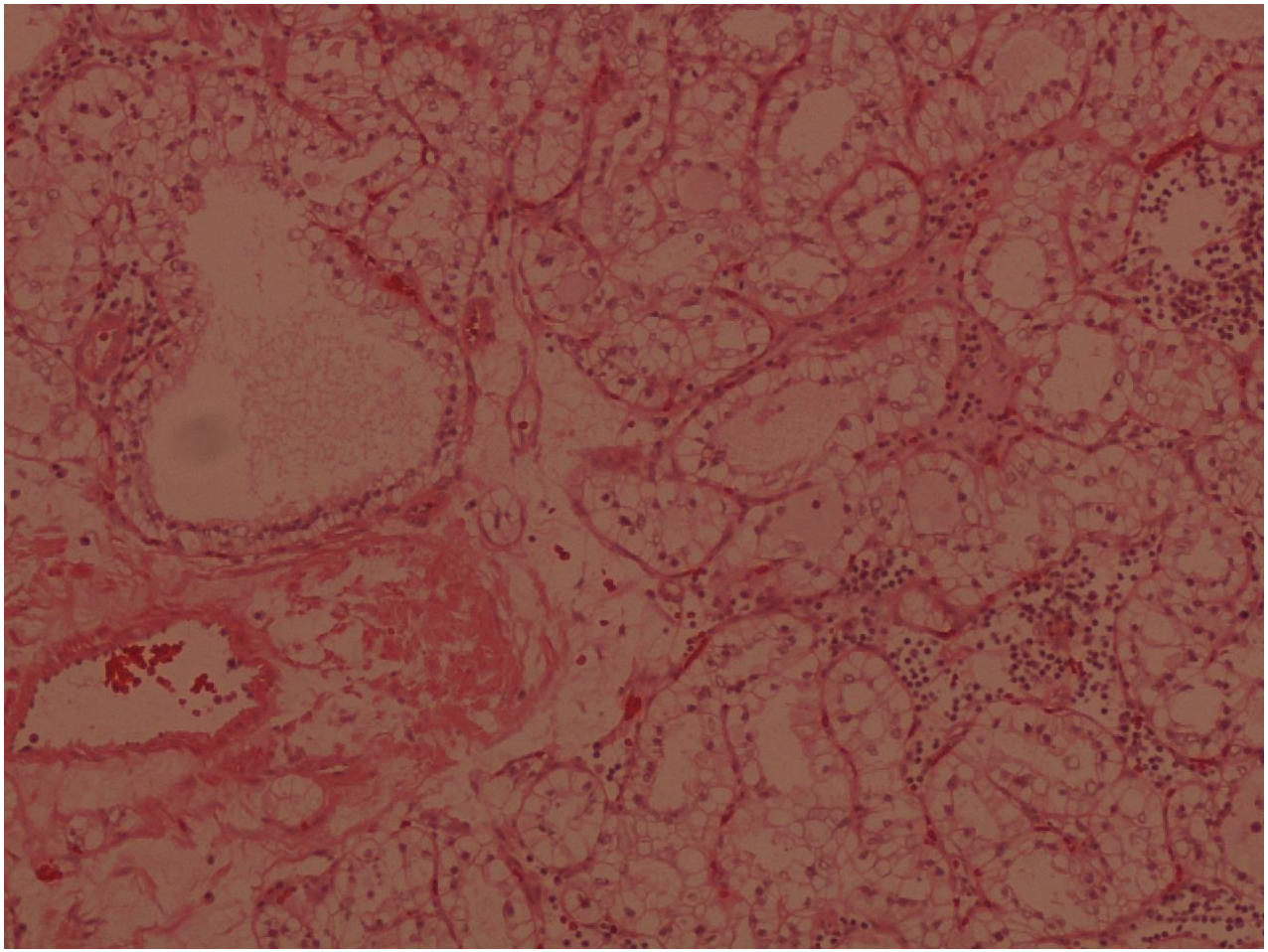
E1

- 68yr old male. Slowly enlarging right renal mass.

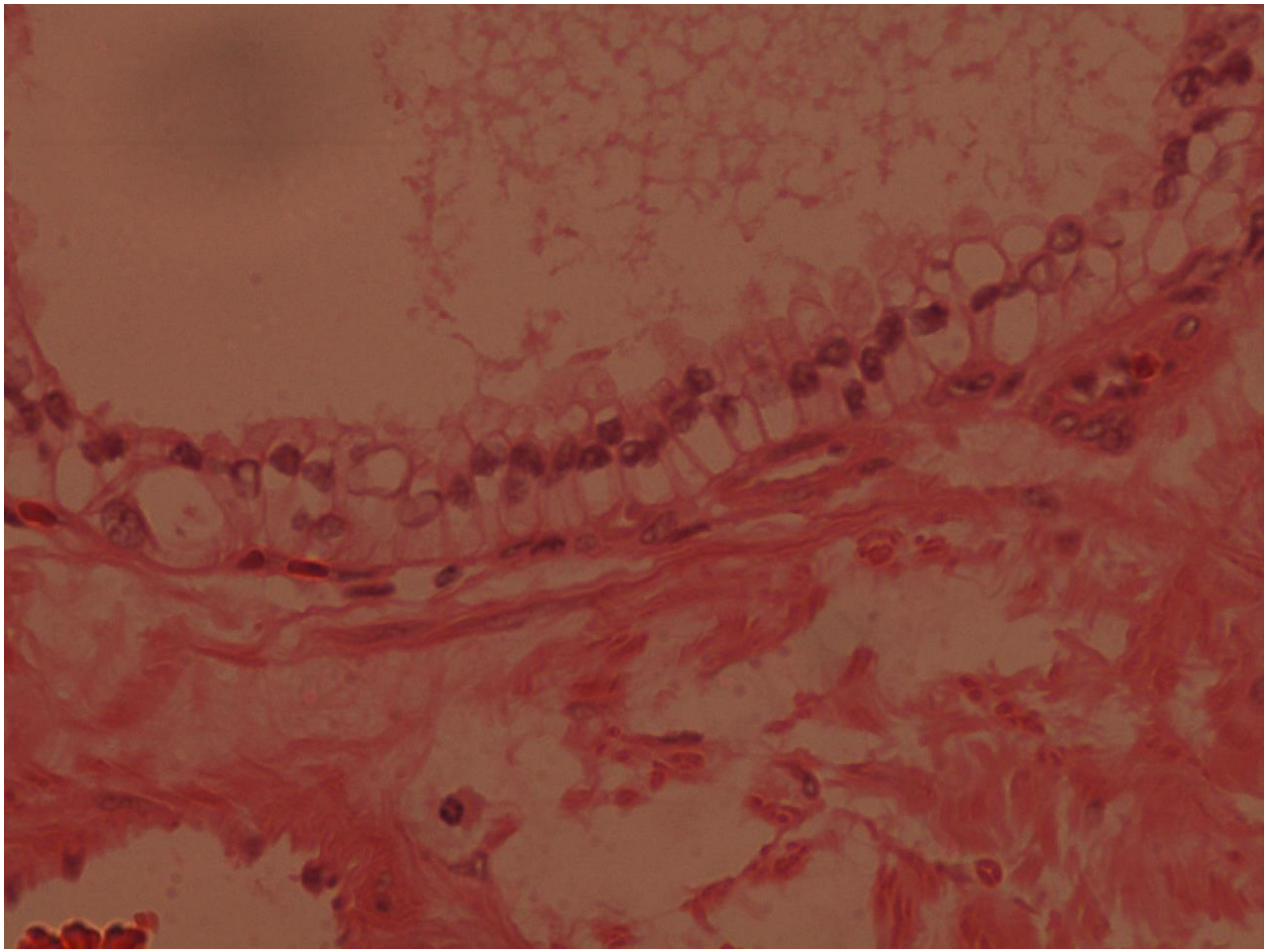
E1



E1



E1



E1 Responses (n=77)

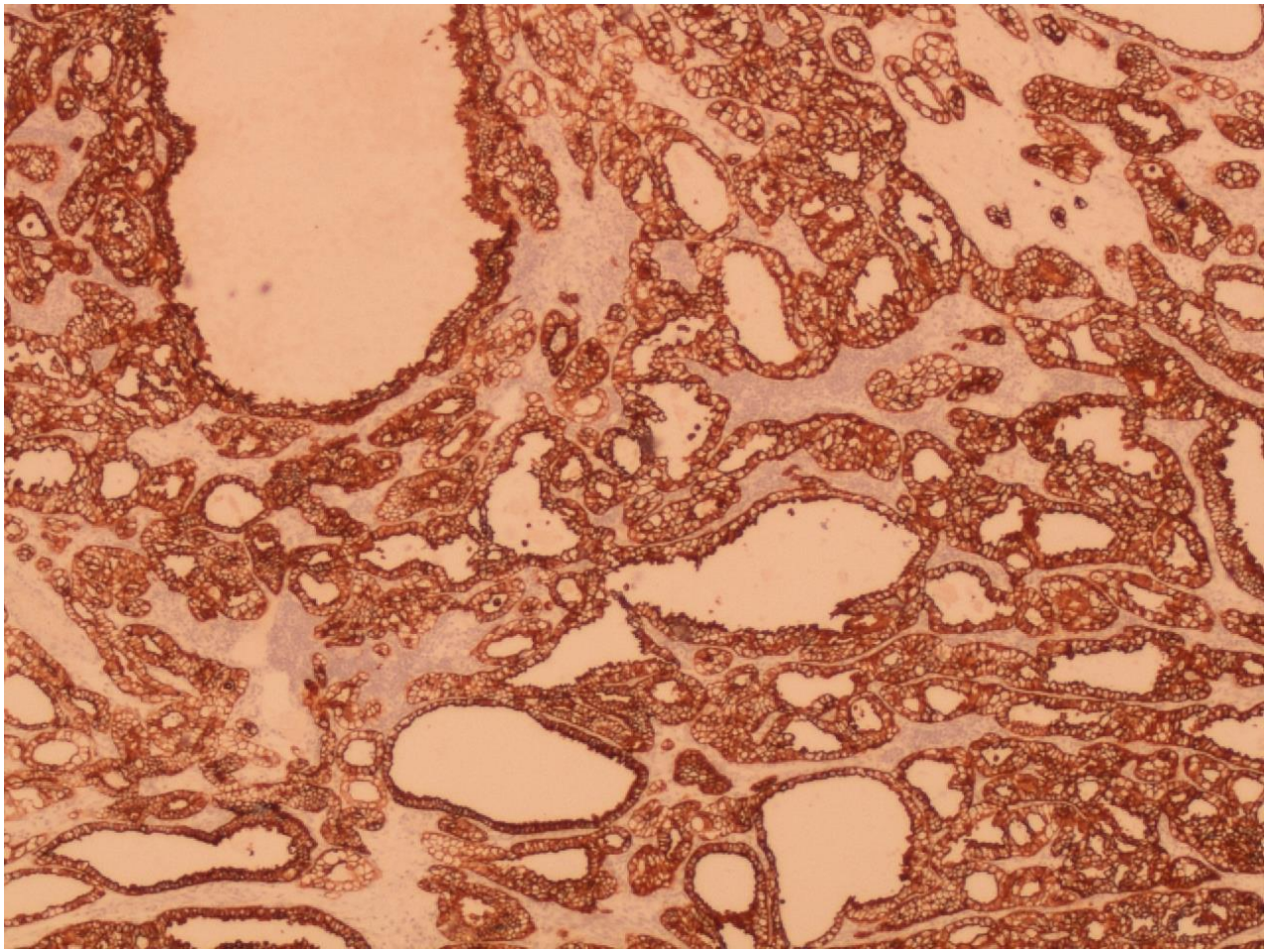
- Clear cell papillary renal cell carcinoma (42)
- Conventional clear cell RCC (13)
- MiT family translocation tumour (5)
- Papillary RCC (5)
- ?Clear cell tubopapillary (5)
- Collecting duct carcinoma (4)
- Metastatic thyroid cancer (1)

- Papillary adenoma (1)

E1 Diagnosis

- Clear cell papillary renal cell carcinoma
- 1-4% of resected renal tumours >95% pT1 at presentation
- M:F = 1:1, sporadic, ESRD and VHL
- Indolent, bland clear epithelial cells arranged in tubules & papillae with at least a predominantly linear nuclear alignment away from the basement membrane
- May have fibrous or smooth muscle stroma
- MOST ISUP grade1/2
- No necrosis, no LVI, no PNI
- Distinctive immunophenotype-diffuse CK7, cuplike carbonic anhydrase IX, -ve racemase
- No local recurrence or metastases documented

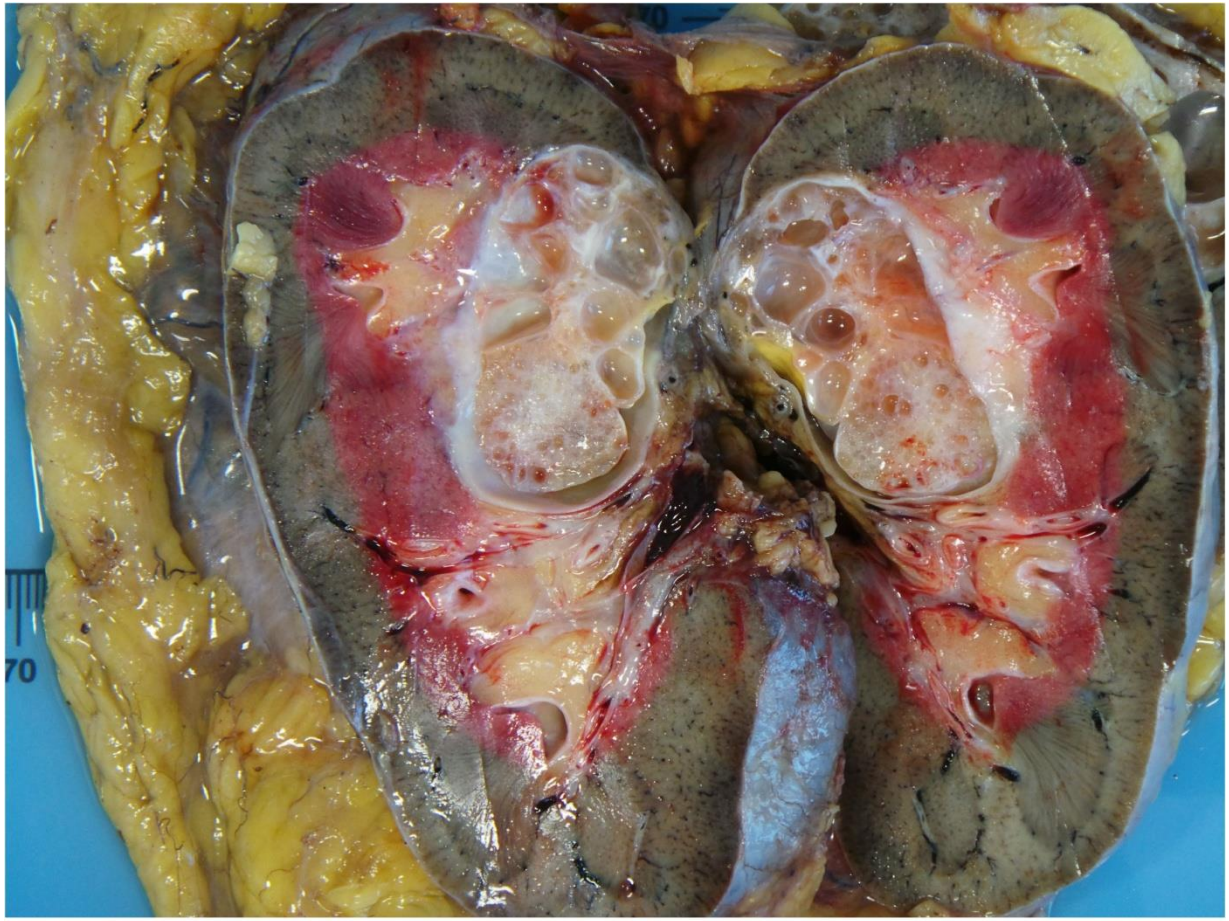
E1-CK7 IHC



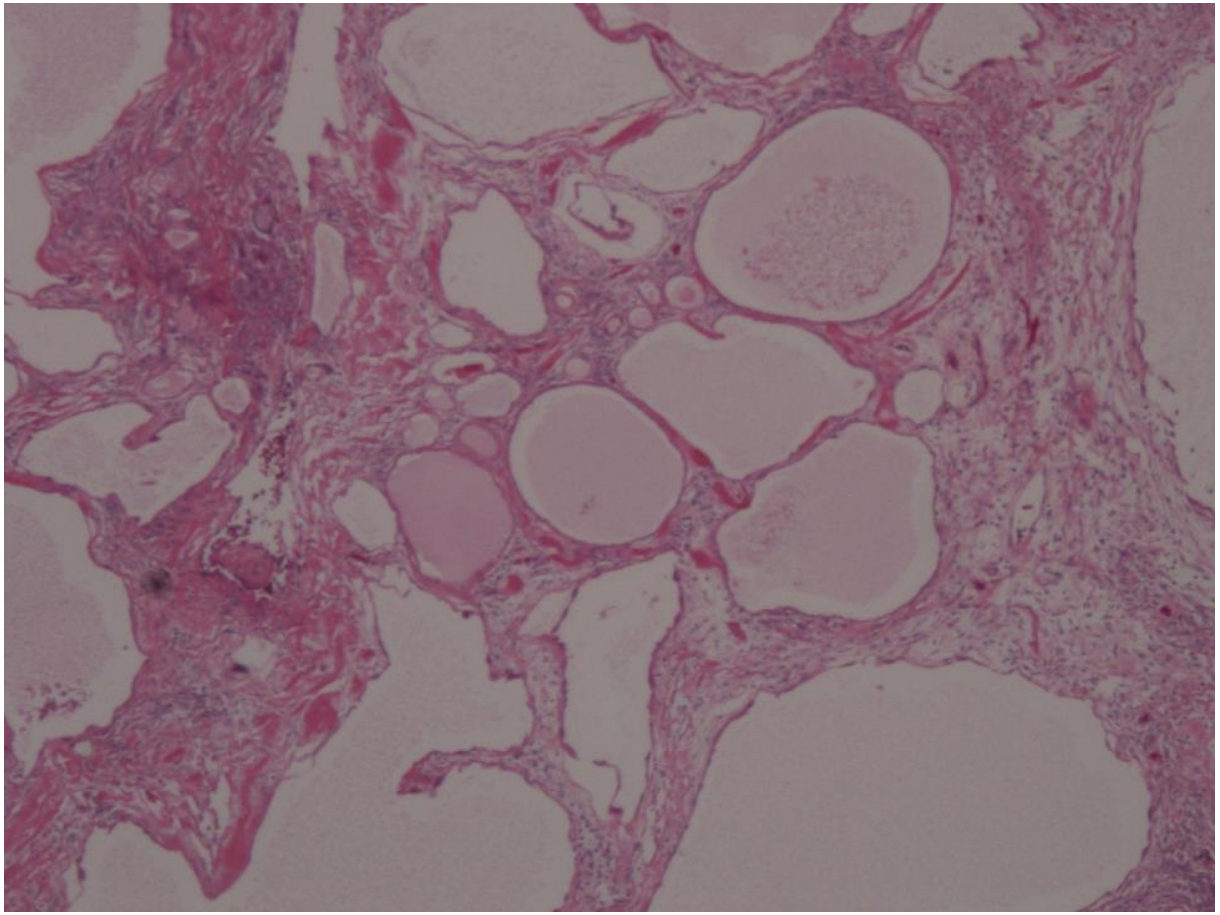
E2

- F 74yrs. Renal mass. Left nephrectomy

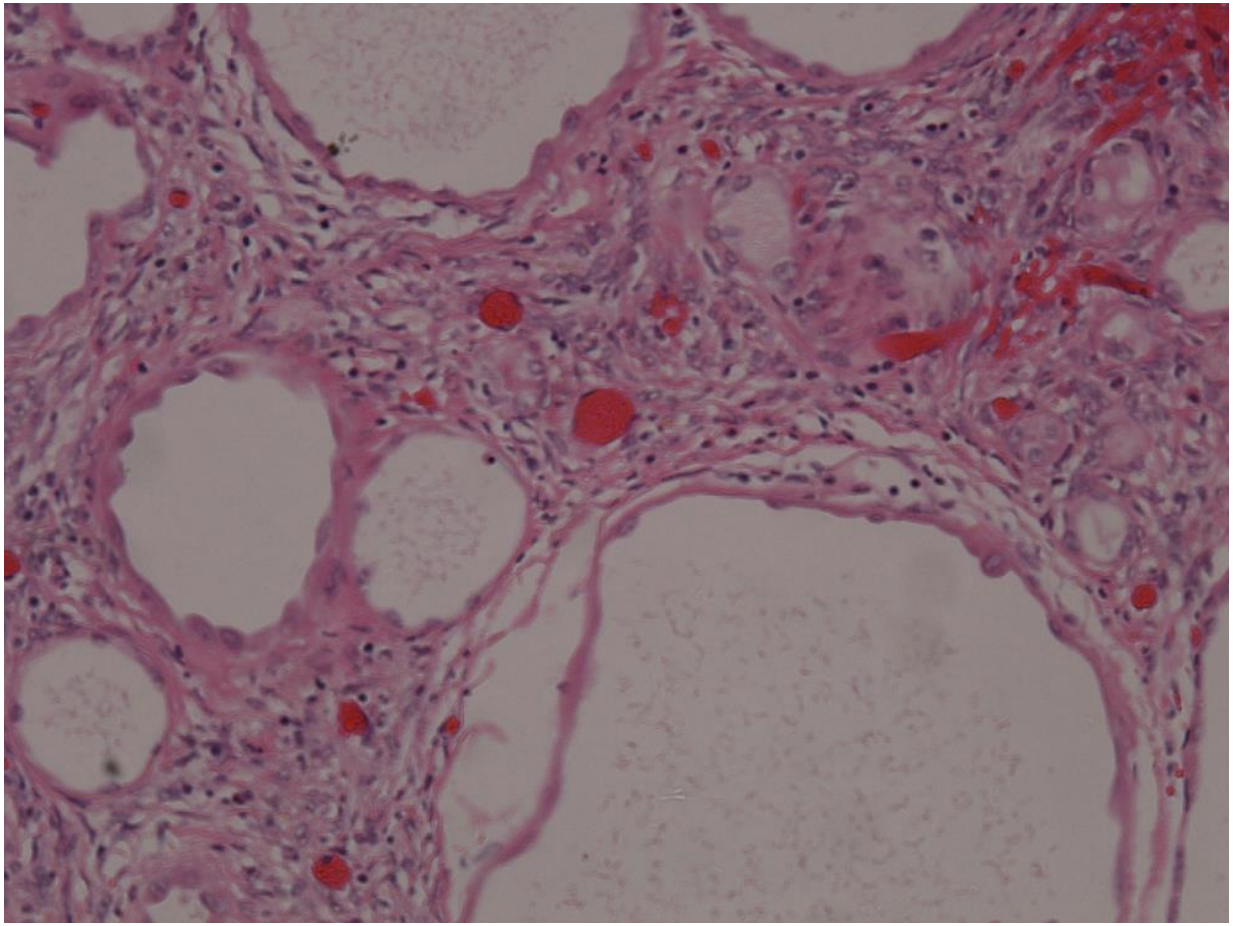
E2 Macro



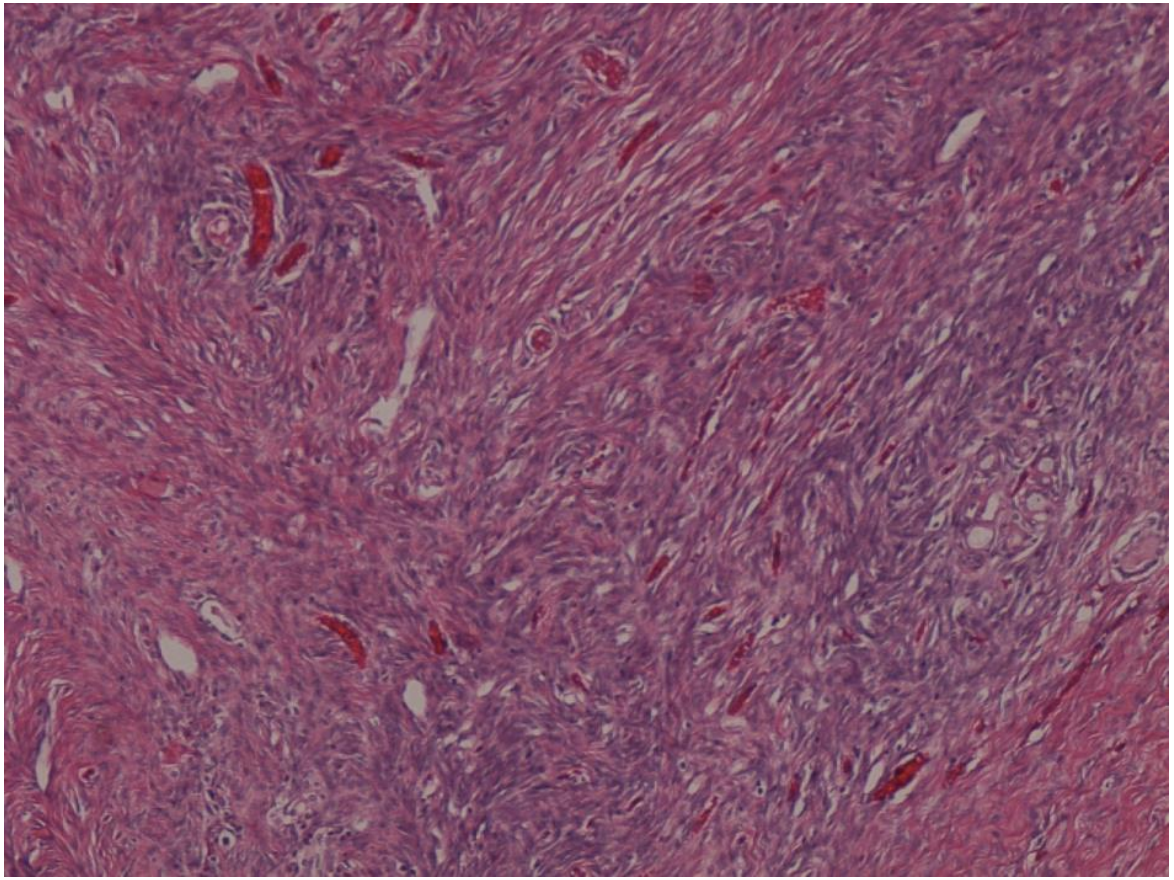
E2



E2



E2



E2 Responses (n=77)

- MEST/adult cystic nephroma (59)
- Multicystic nephroma (3)
- Multilocular cystic nephroma (1)

- Tubulocystic carcinoma (3)
- Thyroid like carcinoma (1)
- Multolocular cystic RCC (1)
- Collecting duct carcinoma/polycystic kidney (1)

- Polycystic kidney disease (3)
- Renal dysplasia (1)
- Medullary sponge kidney (1)
- Benign (2)

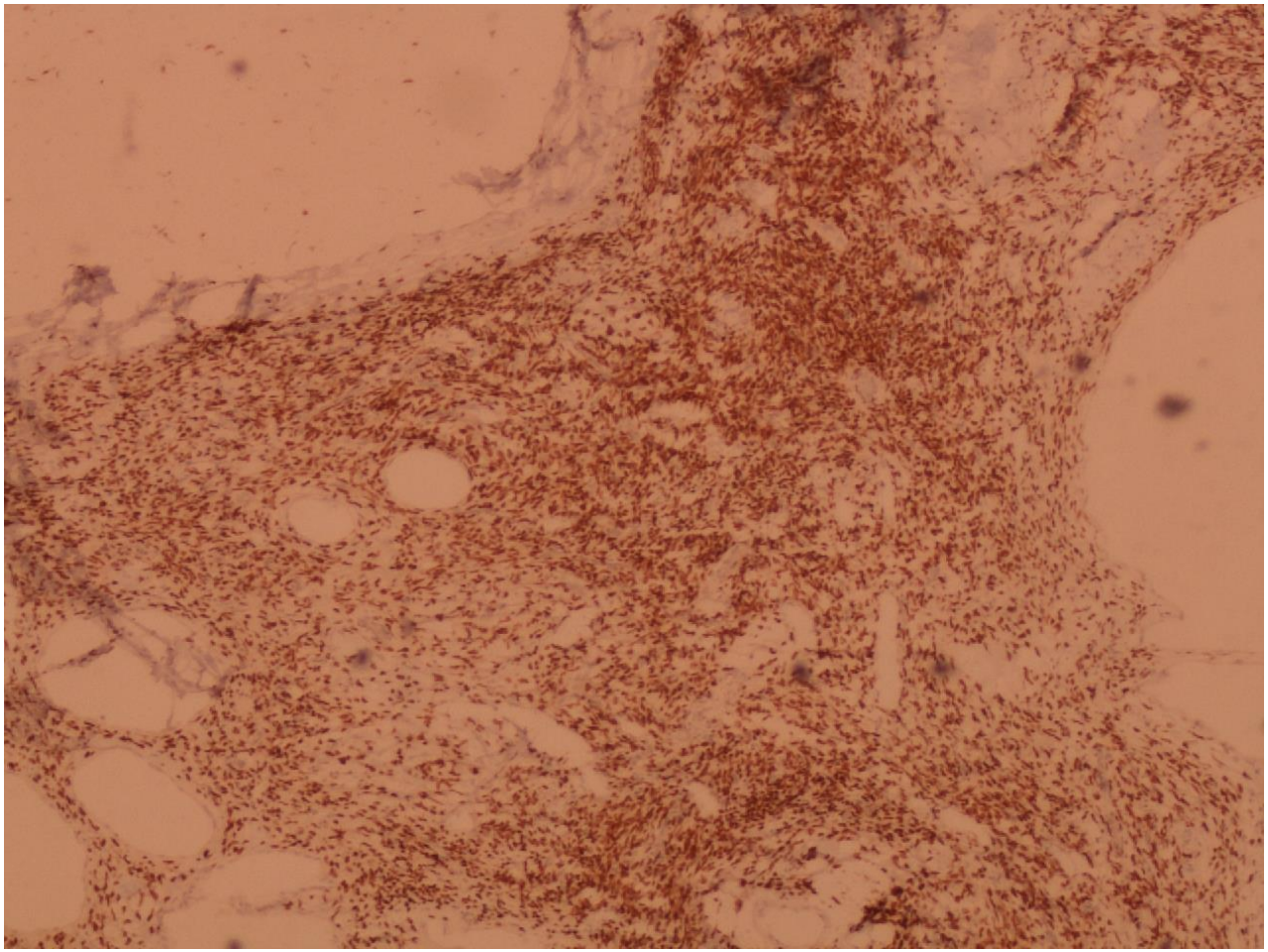
E2 Diagnosis

- Mixed epithelial and stromal tumour (MEST) family
- Ranges from predominantly cystic lesion (adult cystic nephroma) to tumours that are variably solid and contain biphasic stromal and epithelial components (MEST)
- Previously classified with paediatric cystic nephroma
- Rare-? Associated with hormonal imbalance
- Typically peri-menopausal women. 7:1 F:M ratio
- Always unilateral

E2 Diagnosis continued

- Well circumscribed, variably sized cysts and glands with intervening stroma
- Flat cuboidal, or hobnail cells (columnar rarely)
- Focally the epithelium has Mullerian features
- Stroma variably cellular
- Smooth muscle metaplasia common
- Ovarian type stroma often present-can be luteinised
- No significant atypia, necrosis or mitotic activity
- Stromal component typically positive for CD10, SMA, desmin, ER & PR
- Rarely aggressive-sarcomatoid

E2-ER IHC



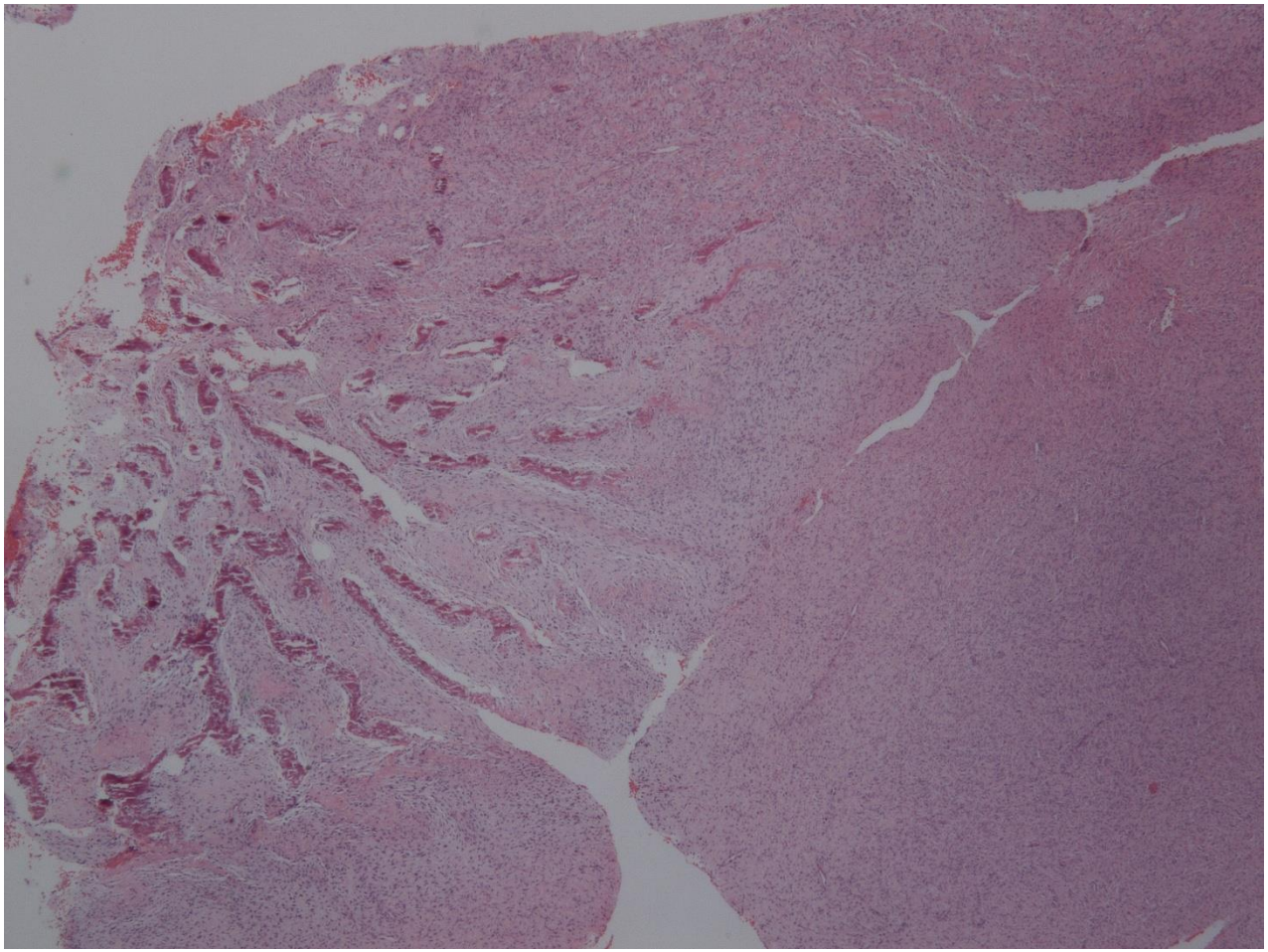
General EQA Circulation 51

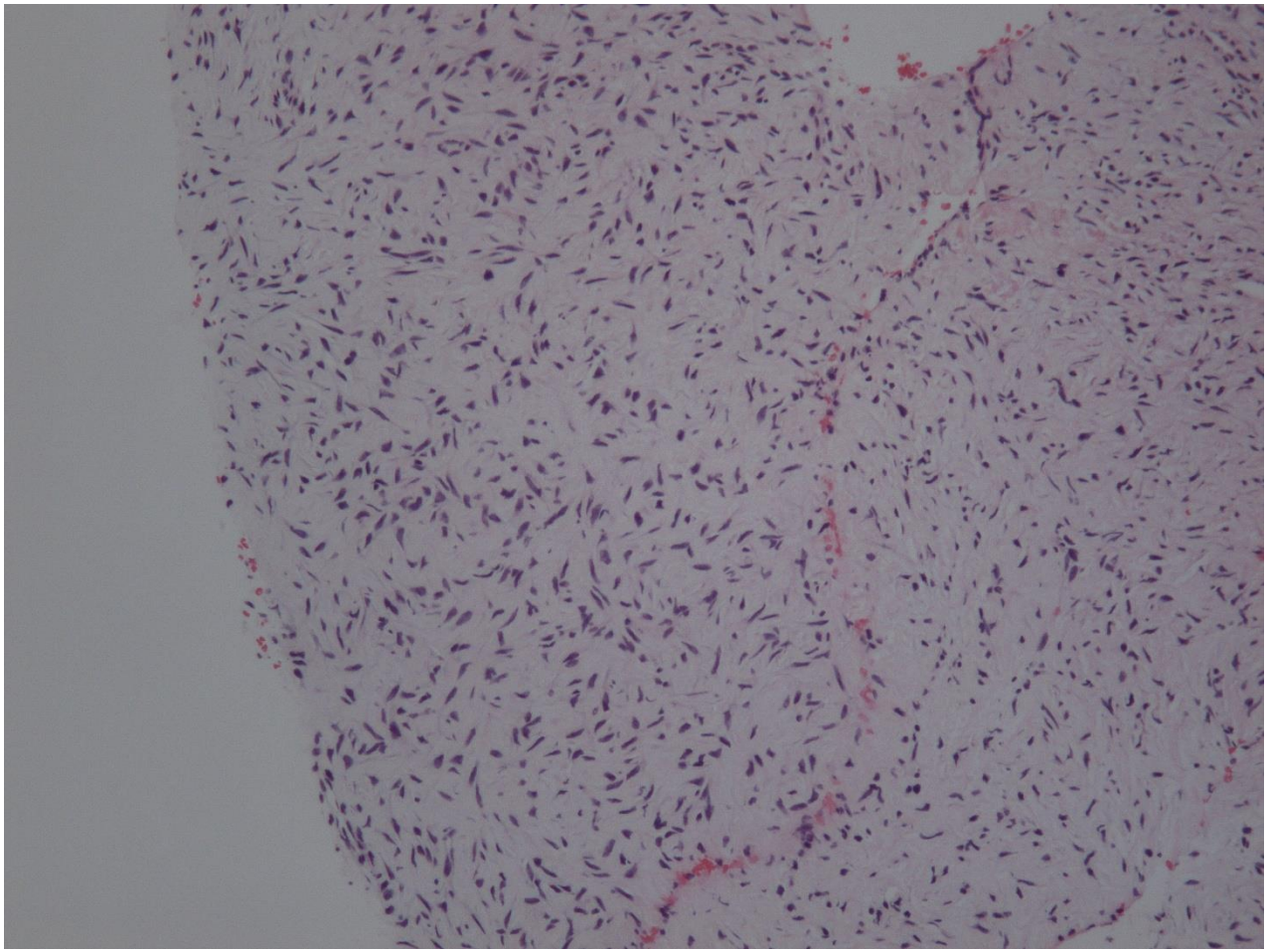
E3 and E4

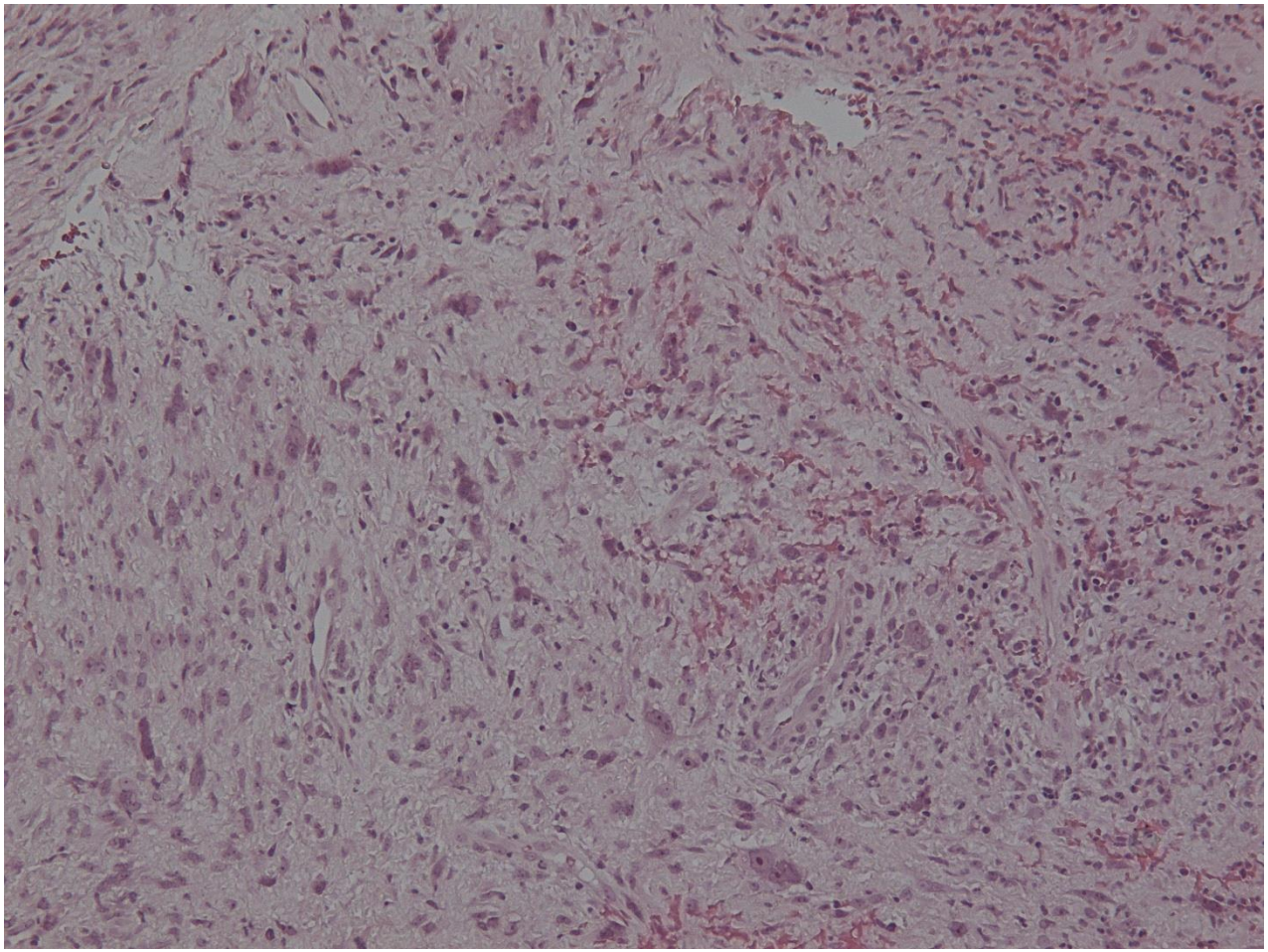
Dr Sarah Mukhtar

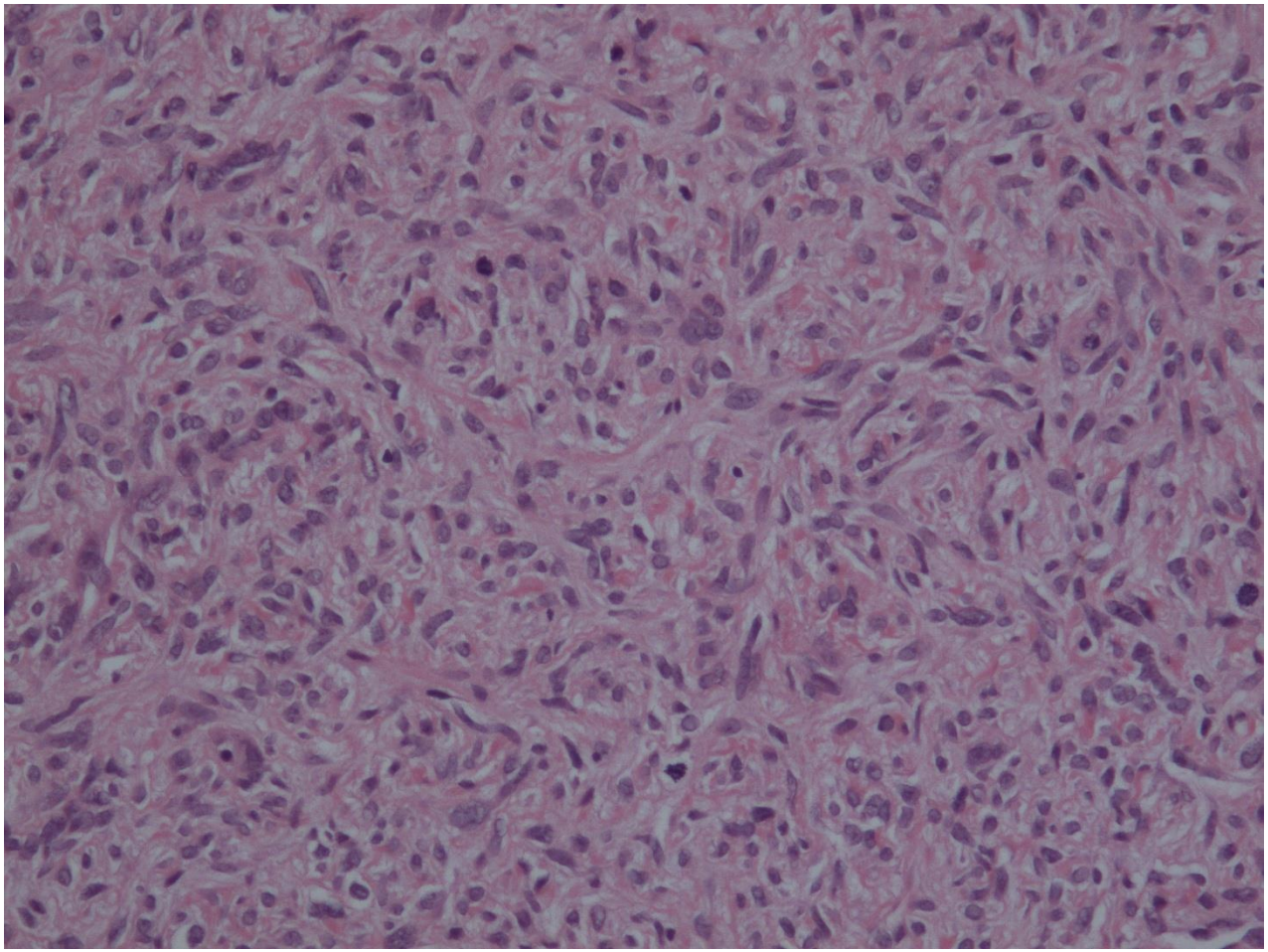
E3

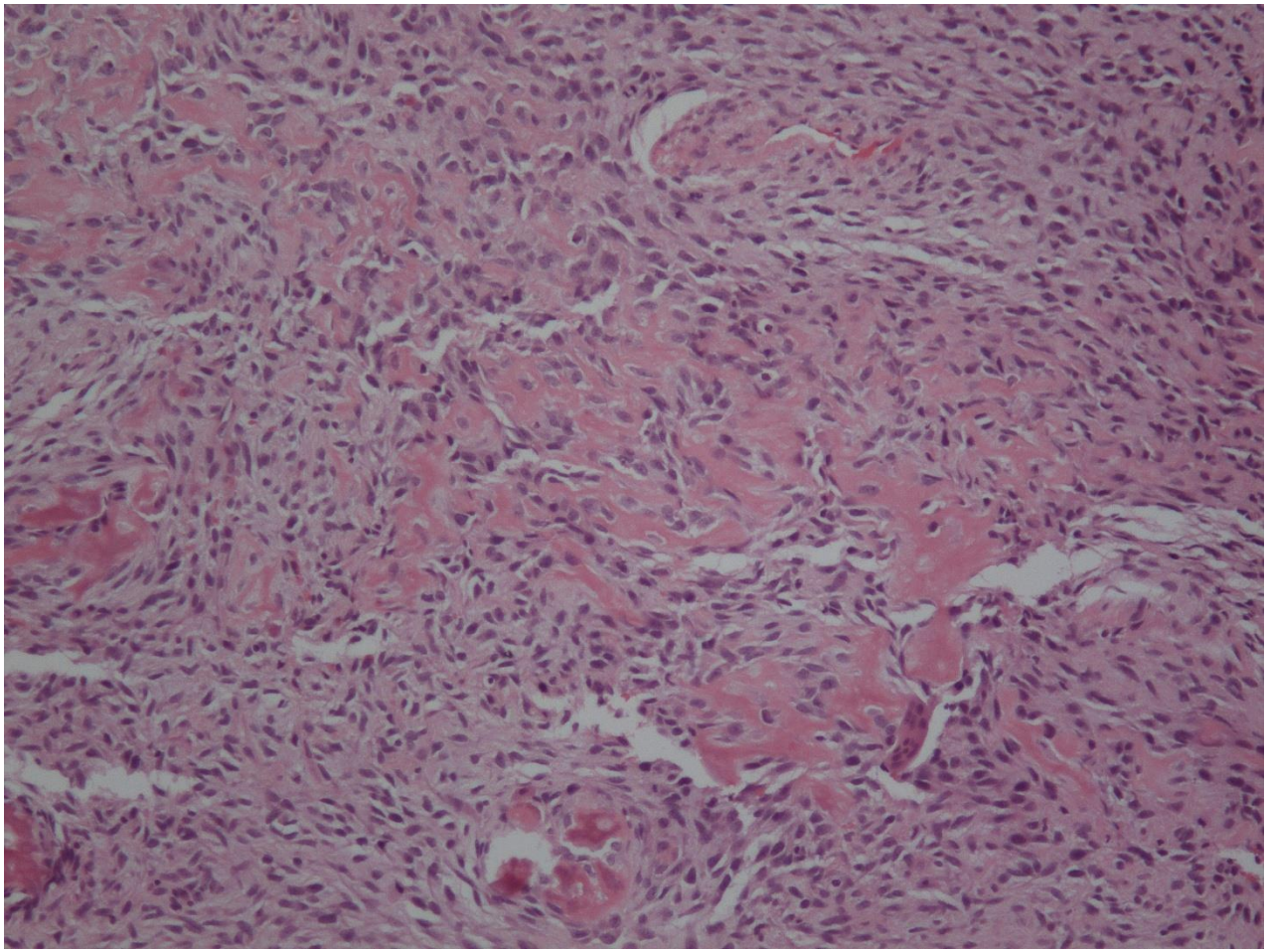
- 76 year old male. Left maxillary sinus. Previous SCC arising in inverted papilloma. ? recurrence.
- Multiple cream fragments of tissue measuring 3-19mm











Positive

- Vimentin
- S100

Negative

- AE1/3
- CAM5.2
- p63
- Desmin, actin, SMA
- CD34

Osteosarcoma (post radiation)

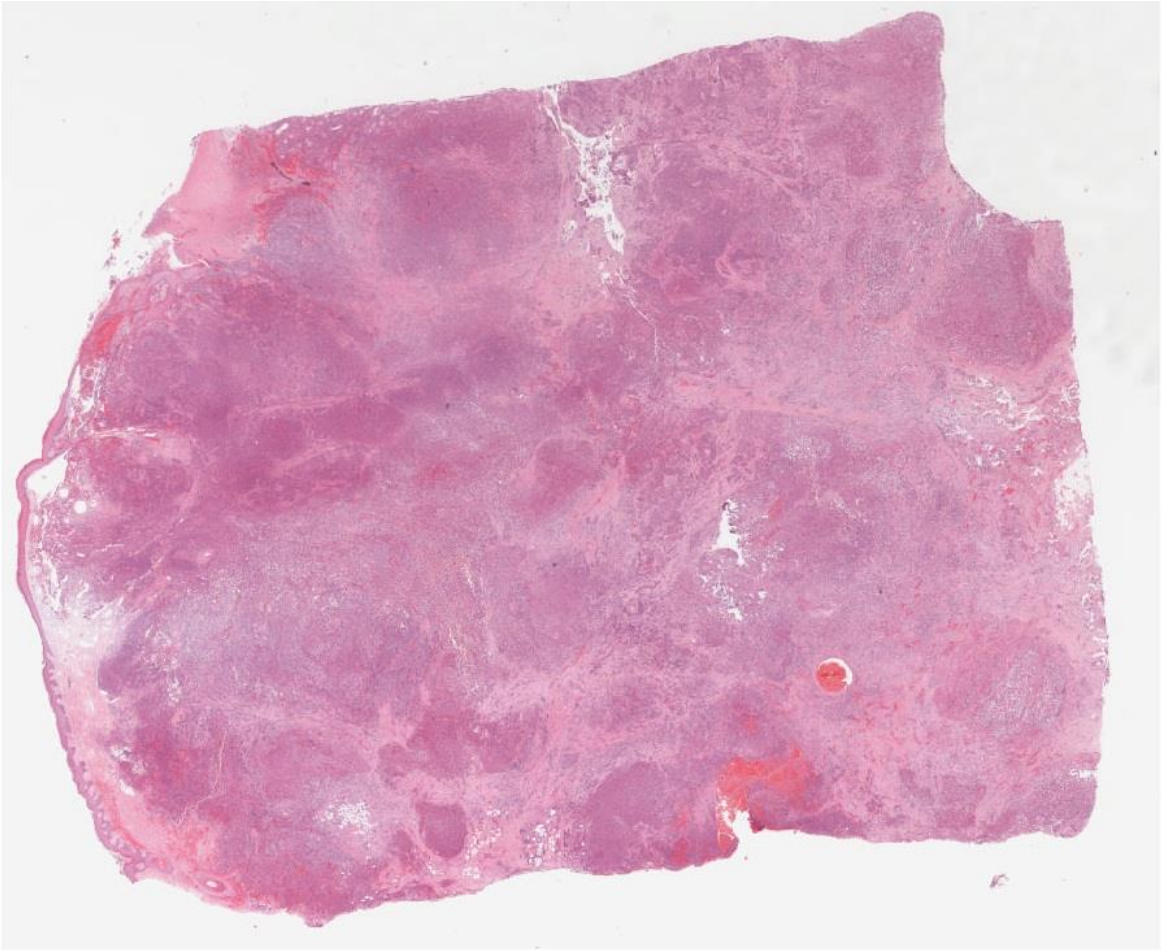
- 55 malignant (osteosarcoma, malignant spindle cell neoplasm, spindle cell carcinoma, carcinoma vs sarcoma, sarcoma, chondrosarcoma, carcinosarcoma, sclerosing rhabdomyosarcoma)
- 13 benign or malignant or not specified (differential, spindle cell tumour, fibrous dysplasia vs SCC, bone forming tumour)
- 10 benign (ossifying fibroma, solitary fibrous tumour, fibrous dysplasia, angiofibroma/solitary fibrous tumour)

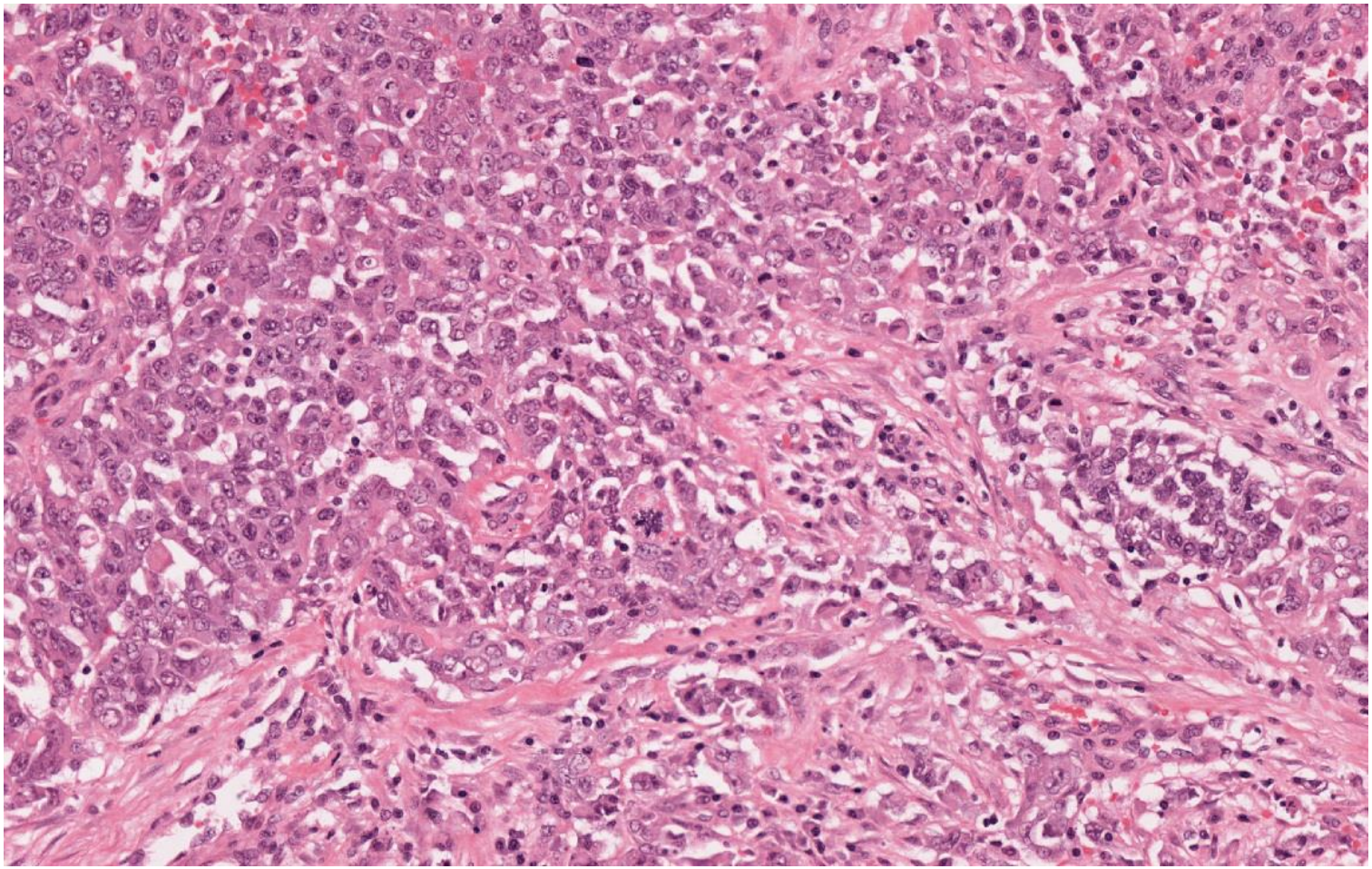
Osteosarcoma

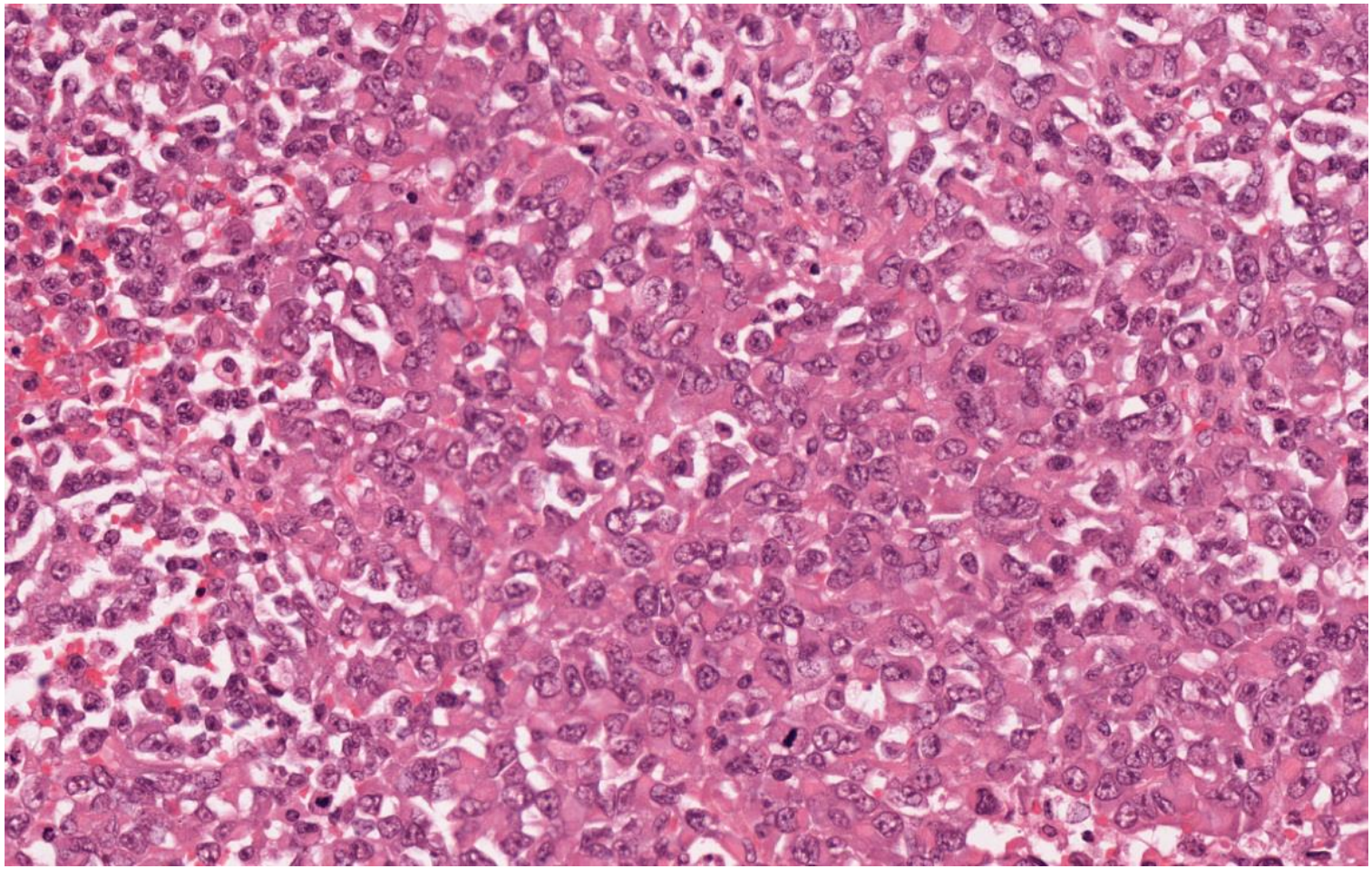
- 6% of all cases found in jaws, 10-20 years later than conventional osteosarcomas
- Most cases spontaneous, but can be post radiotherapy or associated with underlying Pagets
- Tend to be higher grade than conventional osteosarcomas, often have chondroblastic component
- Low grade dd ossifying fibroma or fibrous dysplasia
- Most common post radiotherapy sarcoma in jaws (50% cases)
- Treatment - surgery

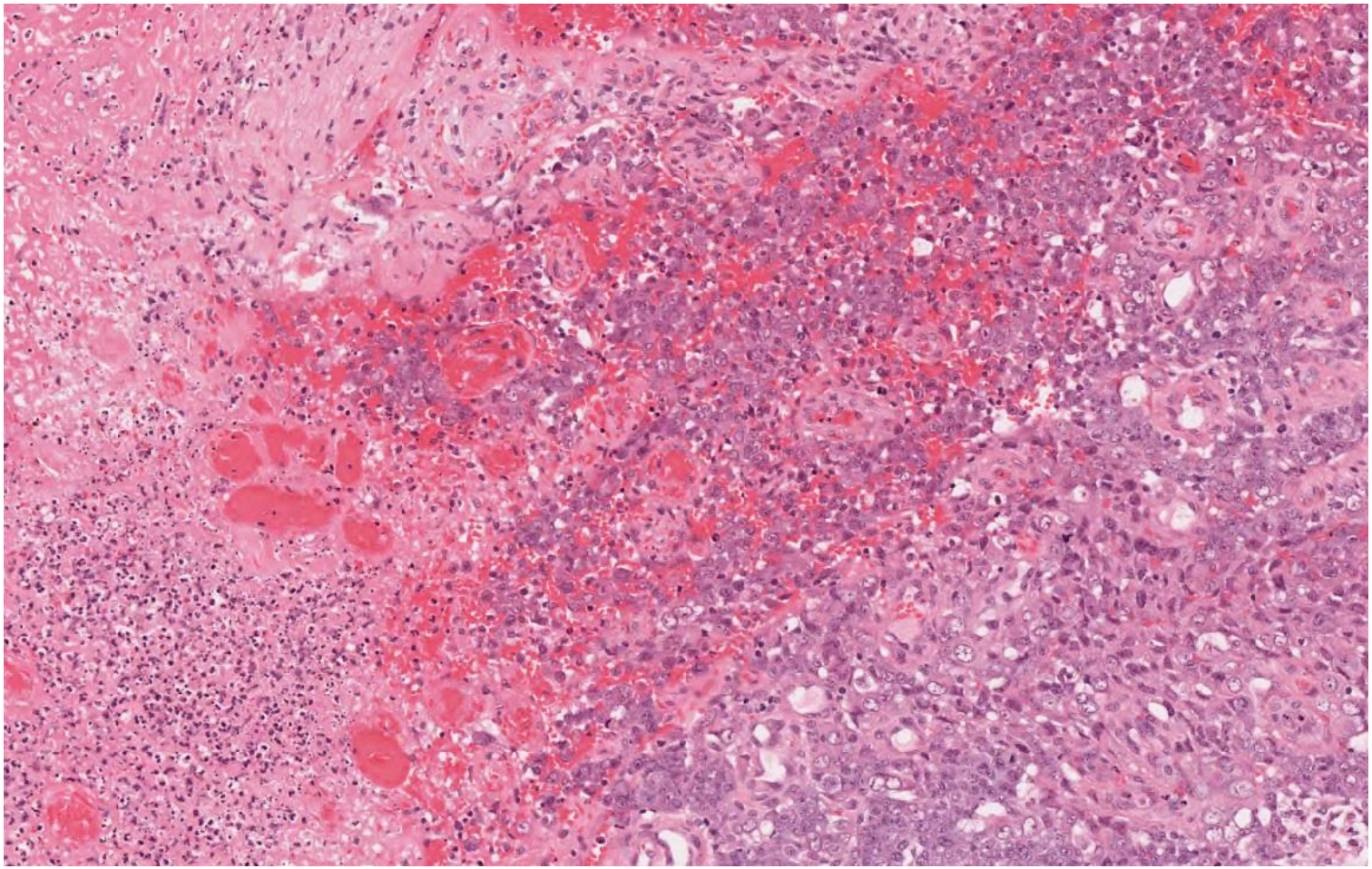
E4

- 44 year old female. Vulval lesion resection.
- Following bx for ‘vulval abscess’
- Hair bearing cutaneous tissue with an exophytic mass measuring 75x45x25mm and groin nodes







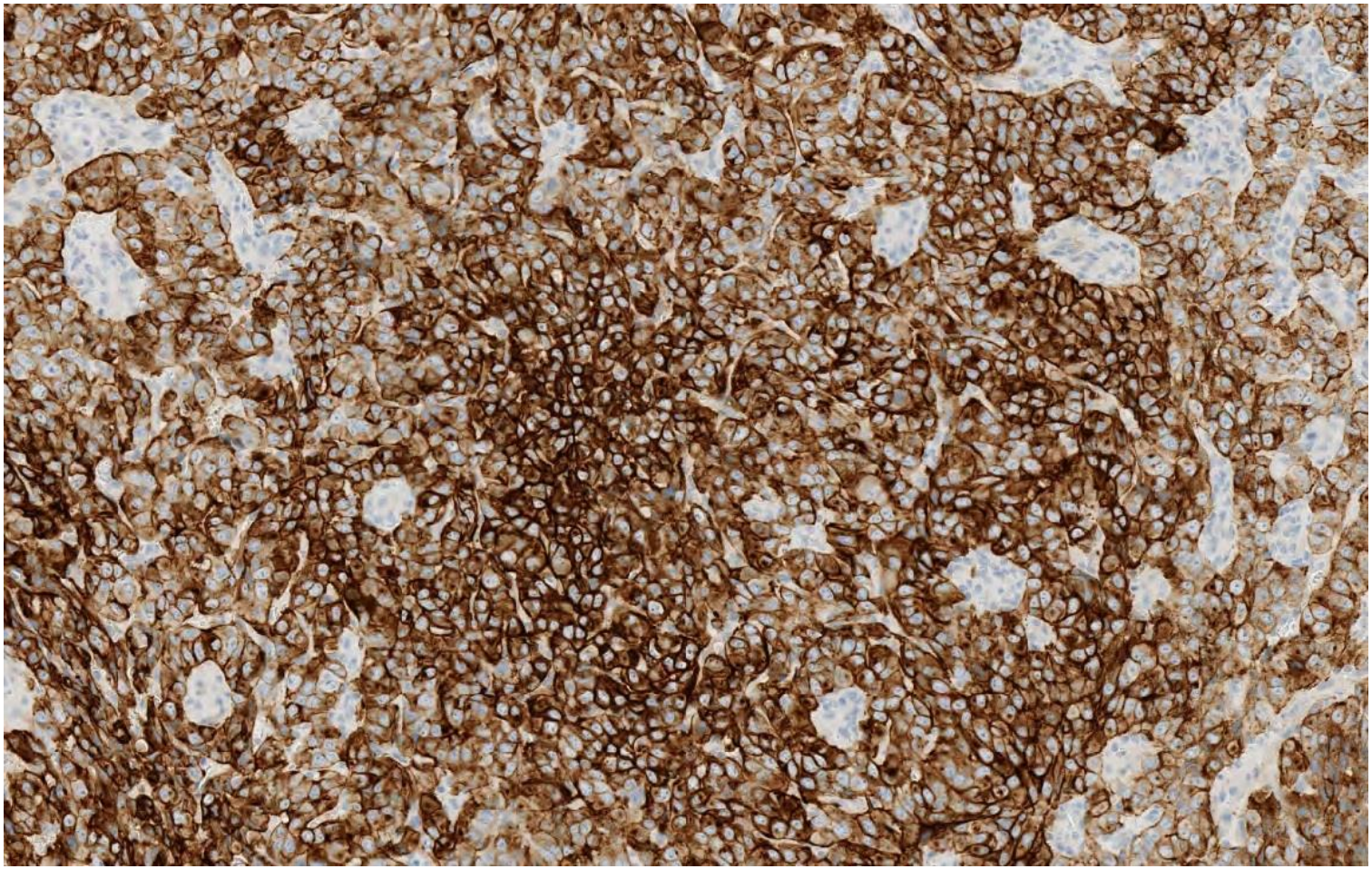


Positive

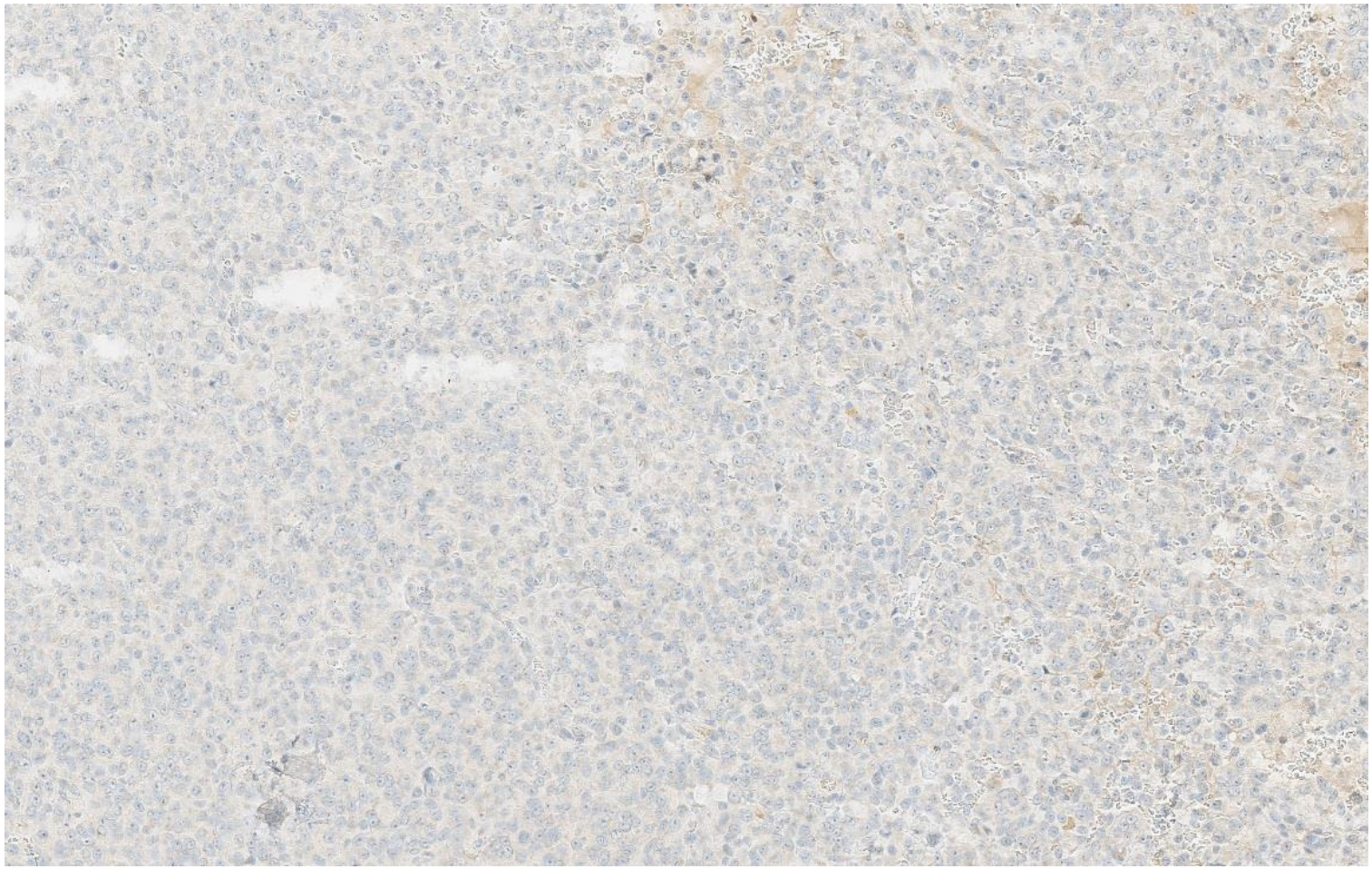
- Vimentin
- EMA
- CAM 5.2
- Actin
- CD56

- **Negative**
- CD34
- Chromogranin
- Synaptophysin
- Melanoma markers
- Desmin
- Caldesmin
- Myo D1
- FLI-1
- EGR
- INI-1

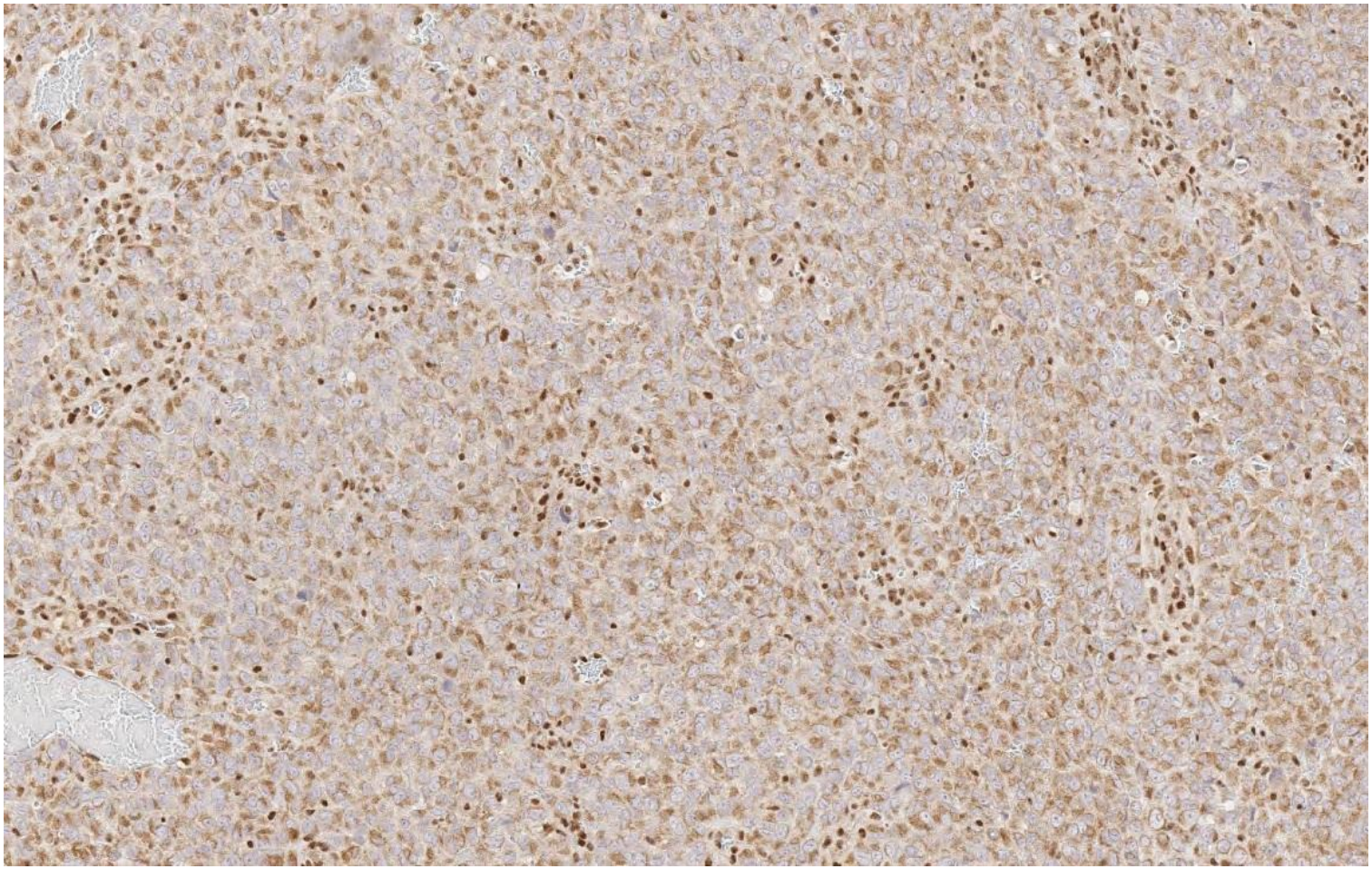
EMA



Myo D1



INI-1



Proximal epitheloid sarcoma

- EVERYONE thought malignant
- 35 gave broad differential +/- IHC or included sarcoma
- 12 specifically mentioned proximal epitheloid sarcoma
- 4 gave differential of two, but sarcoma not mentioned (p diff SCC vs carcinosarcoma, melanoma vs carcinoma
- 1 just sarcoma
- 28 gave one answer (angiosarcoma, melanoma, mixed Ca with Merkel, rhabdomyosarcoma, extra-renal rhabdoid tumour, malignant, metastasis, carcinosarcoma, rhabdoid carcinoma)

Proximal epitheloid sarcoma

- Epitheloid sarcoma 0.6-1%, with classic more common than proximal, slight male predominance, metastasis to nodes
- Proximal type occurs in older age group, predilection for vulva
- Histologically similar to epitheloid sarcoma of soft parts
- Dd melanoma or carcinoma – positive for HWMC, EMA, vimentin and CD34 (50%). Negative S100.
- Loss of SMARB1 (INI1)
- Different to extra-renal rhabdoid tumour
- Poor prognosis