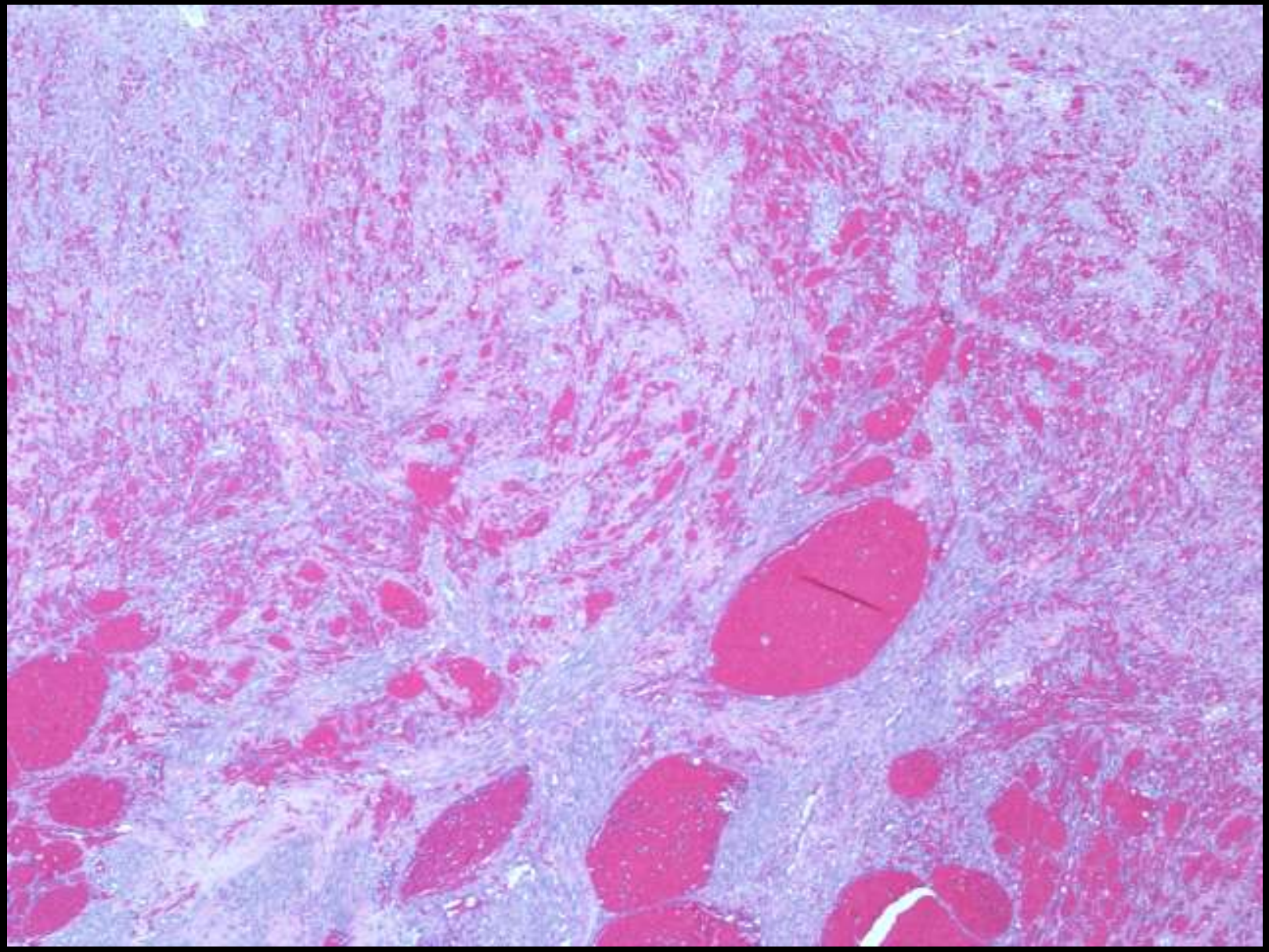
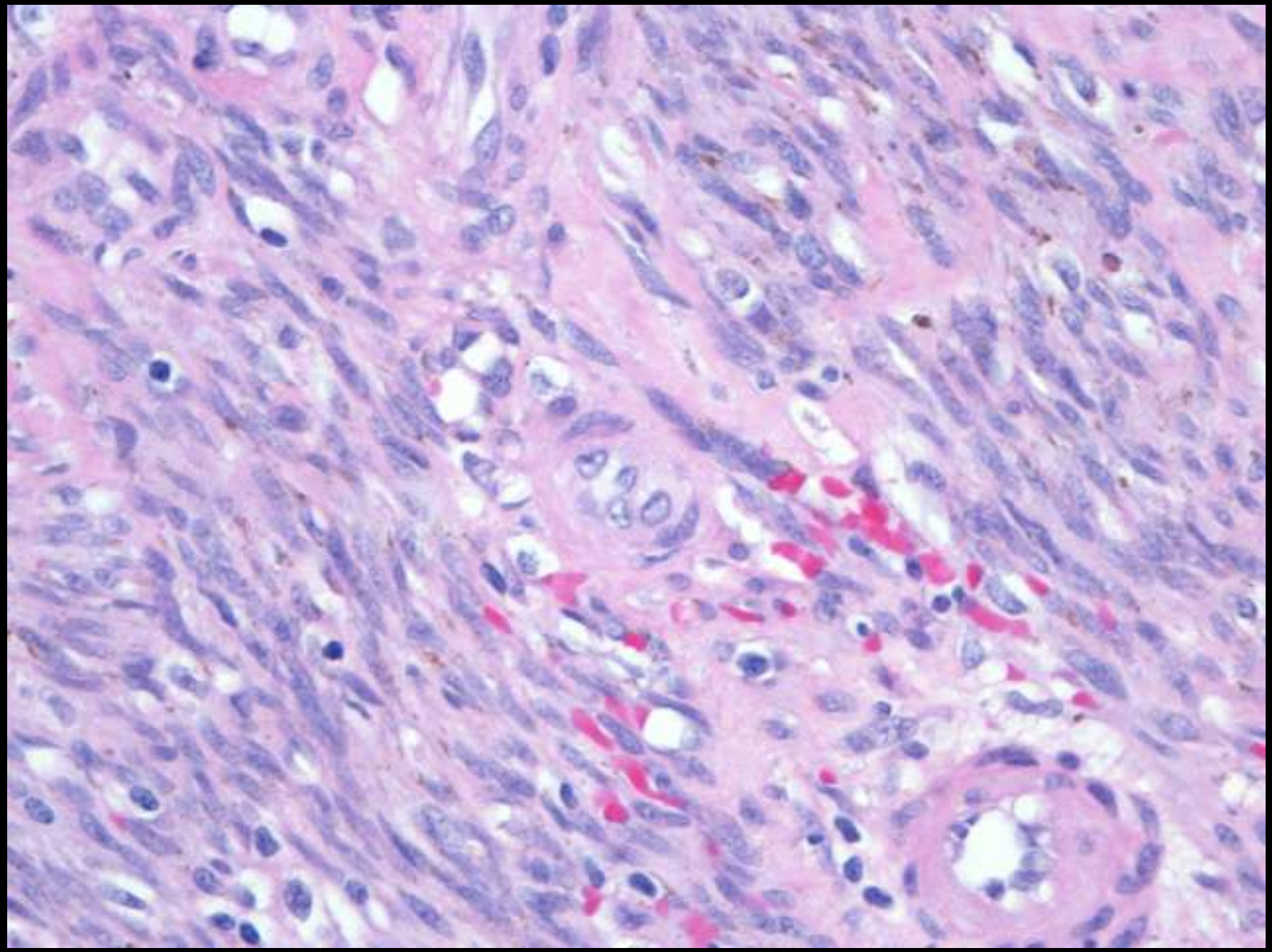
CASE 1

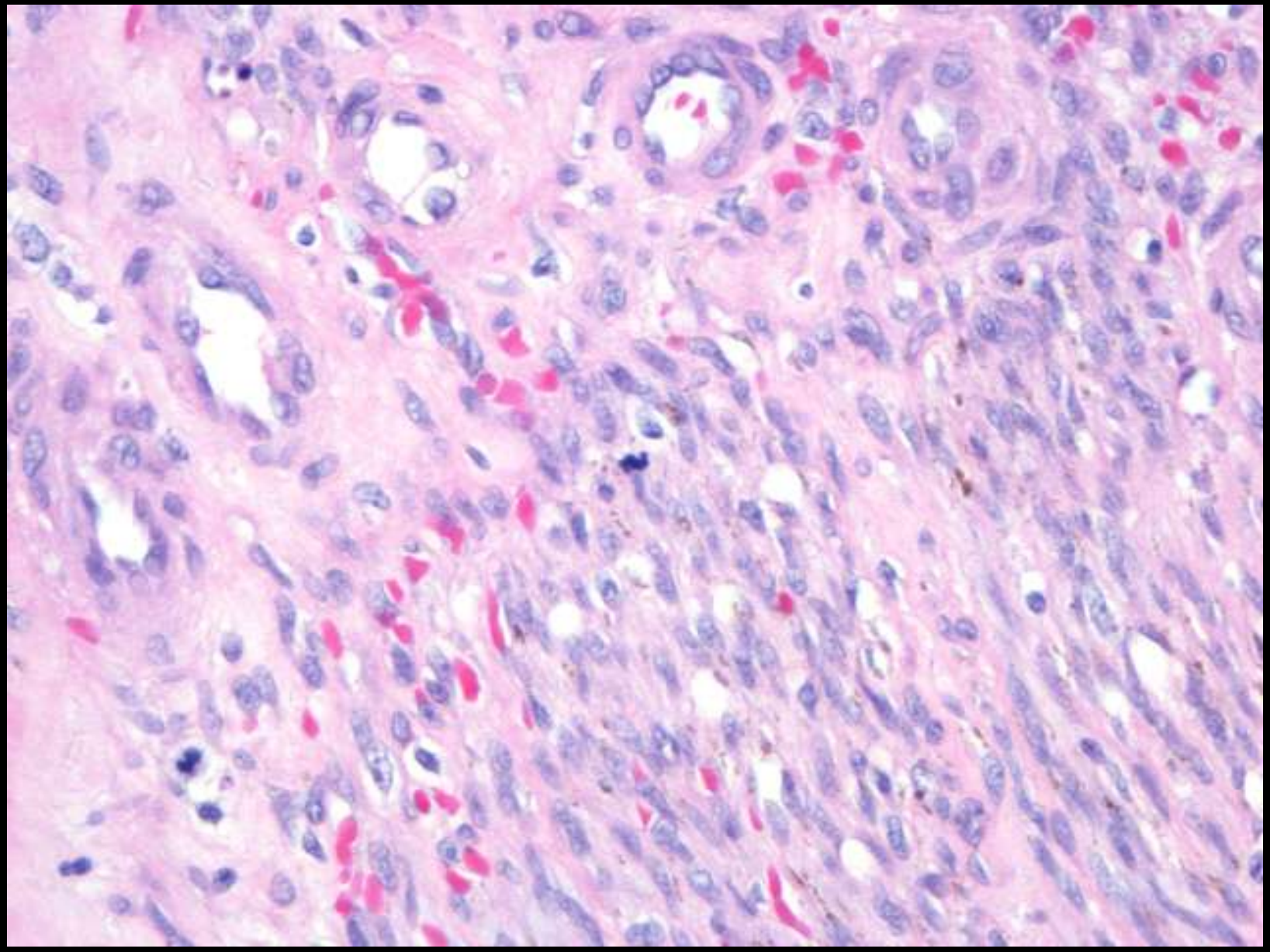
55 year old male with a PET avid nodule in the left adrenal gland

Case 1

- Adrenal gland parenchyma partly replaced by a spindle cell tumour with mild nuclear pleomorphism
- Atypical mitoses present
- Spindle cells surround slit-like vascular spaces
- Prominent red blood cell extravasation and focal haemosiderin deposition



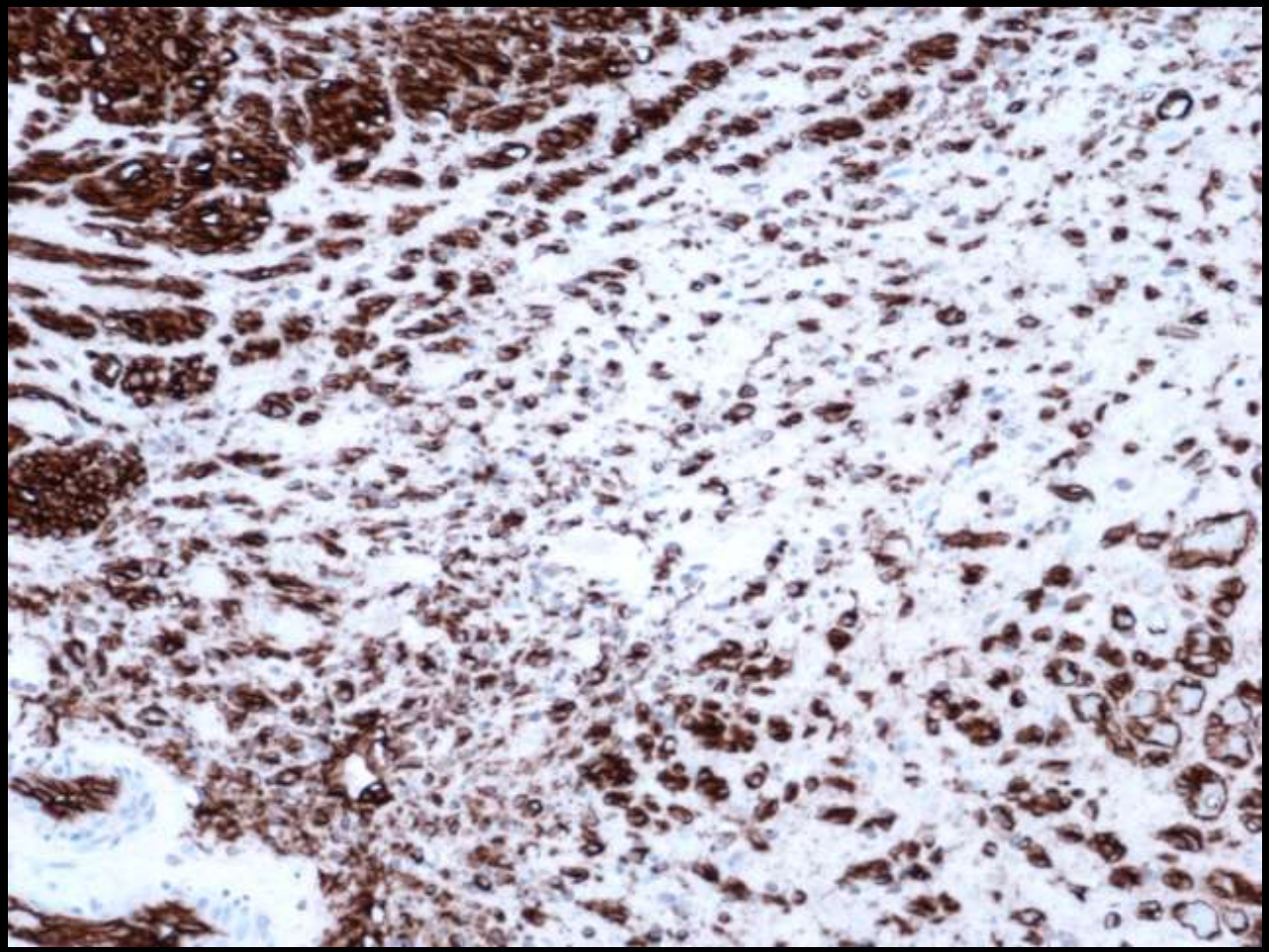




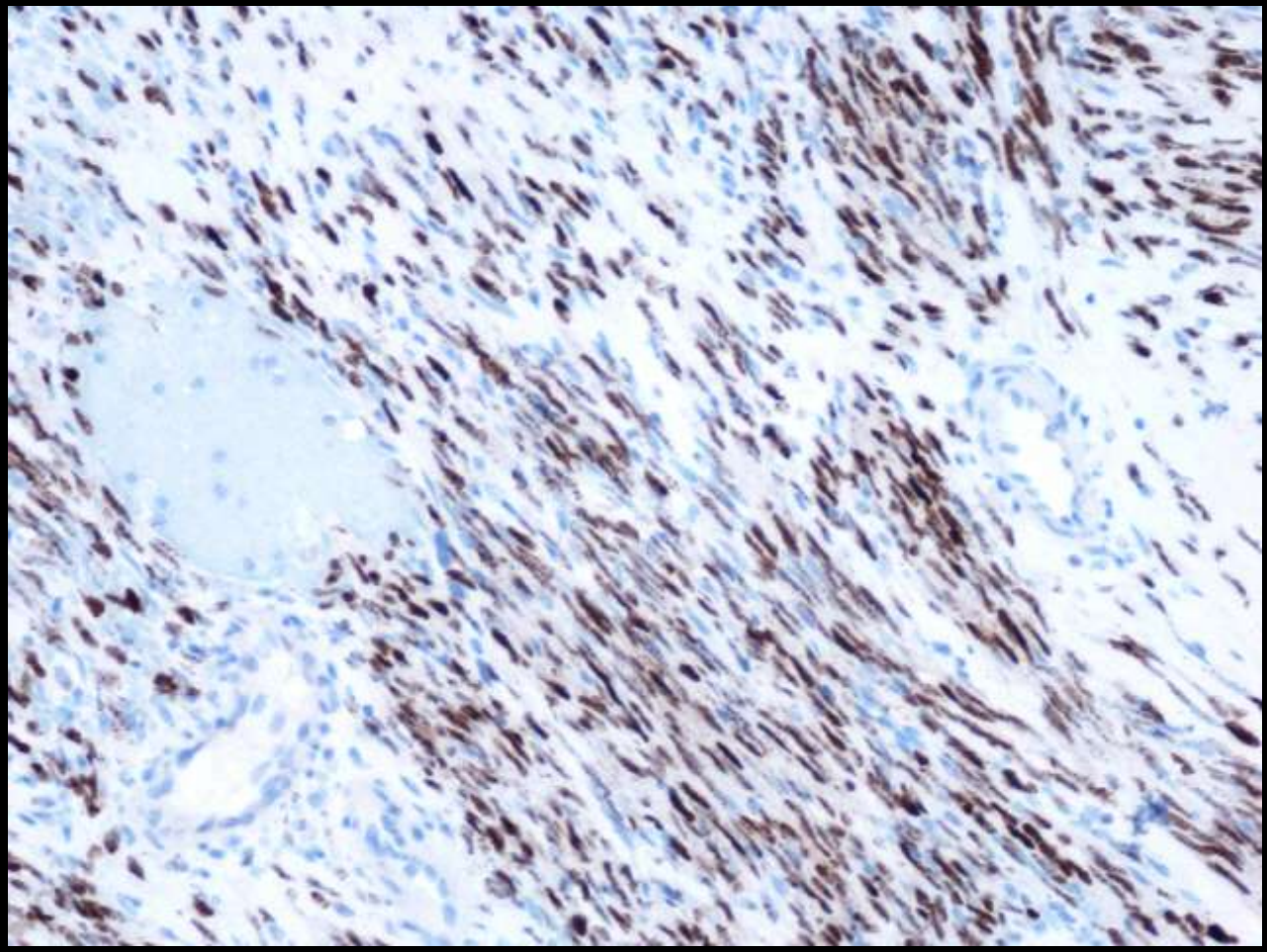
Answers

- Kaposi’s sarcoma (most popular answer)
- Composite phaeochromocytoma
- Angiosarcoma
- Schwannoma
- Spindle cell haemangioma
- Angioleiomyoma
- Ganglioneuroma
- Adrenal cortical adenoma
- PEComa

CD 34



HHV 8



Case 1

- Kaposi's sarcoma
- Patient known to be HIV positive with cutaneous and lung involvement
- Good response with highly active retroviral therapy
- Lung lesions currently static on imaging and no recurrence in adrenal bed
- Has had radiotherapy for skin lesions
- Involvement of the adrenal glands by Kaposi's sarcoma is usually a manifestation of a more widely disseminated tumour in patients with acquired immunodeficiency syndrome

Case 1

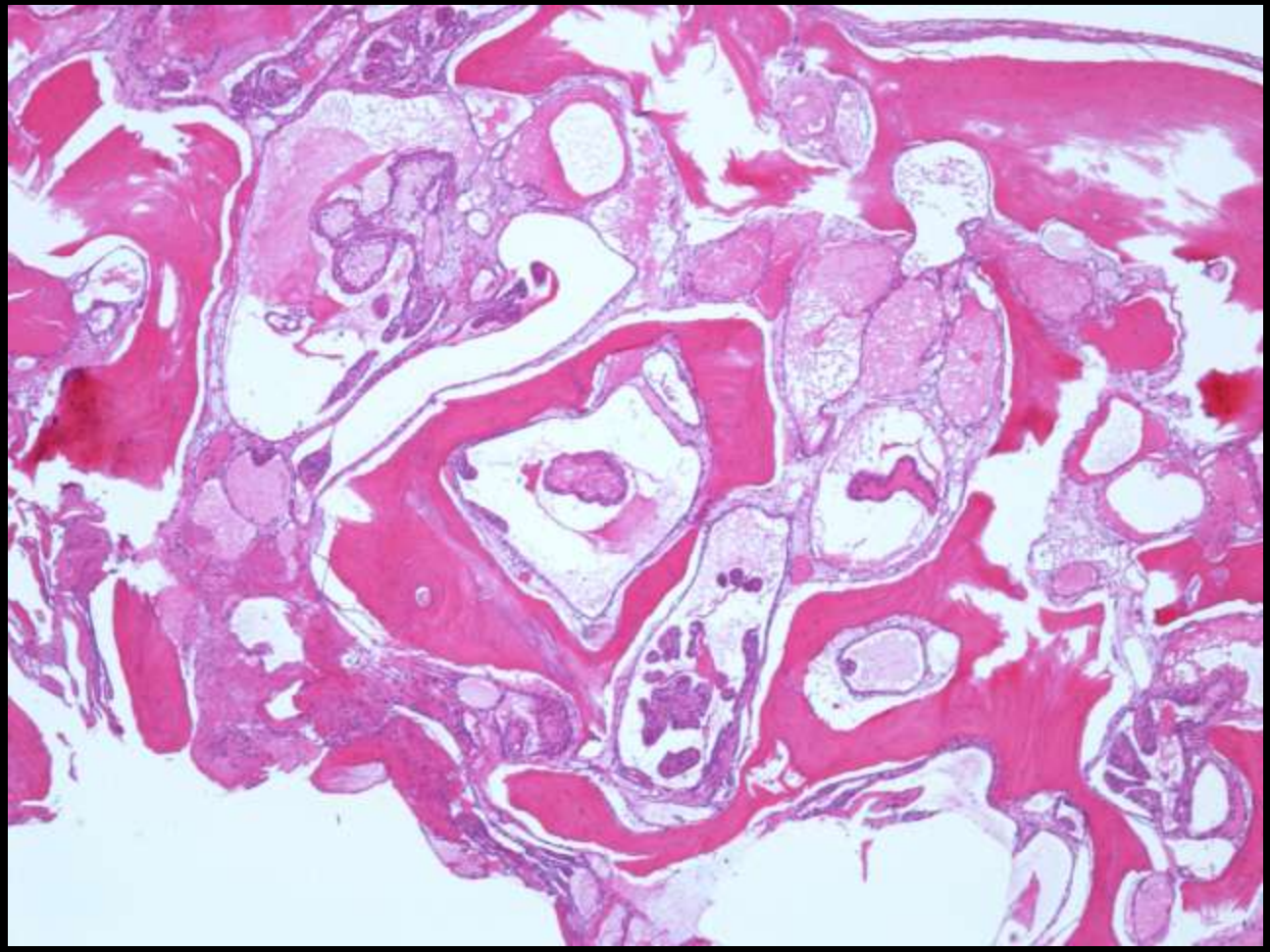
- There are very few detailed reports of adrenal KS published
- Adrenal KS has been documented in post mortem studies in 19% of examined patients with classic (sporadic) KS, 18% with African (endemic) KS and 17% with AIDS-related (epidemic) KS.
- The adrenal cortex appears to be involved far more than the medulla

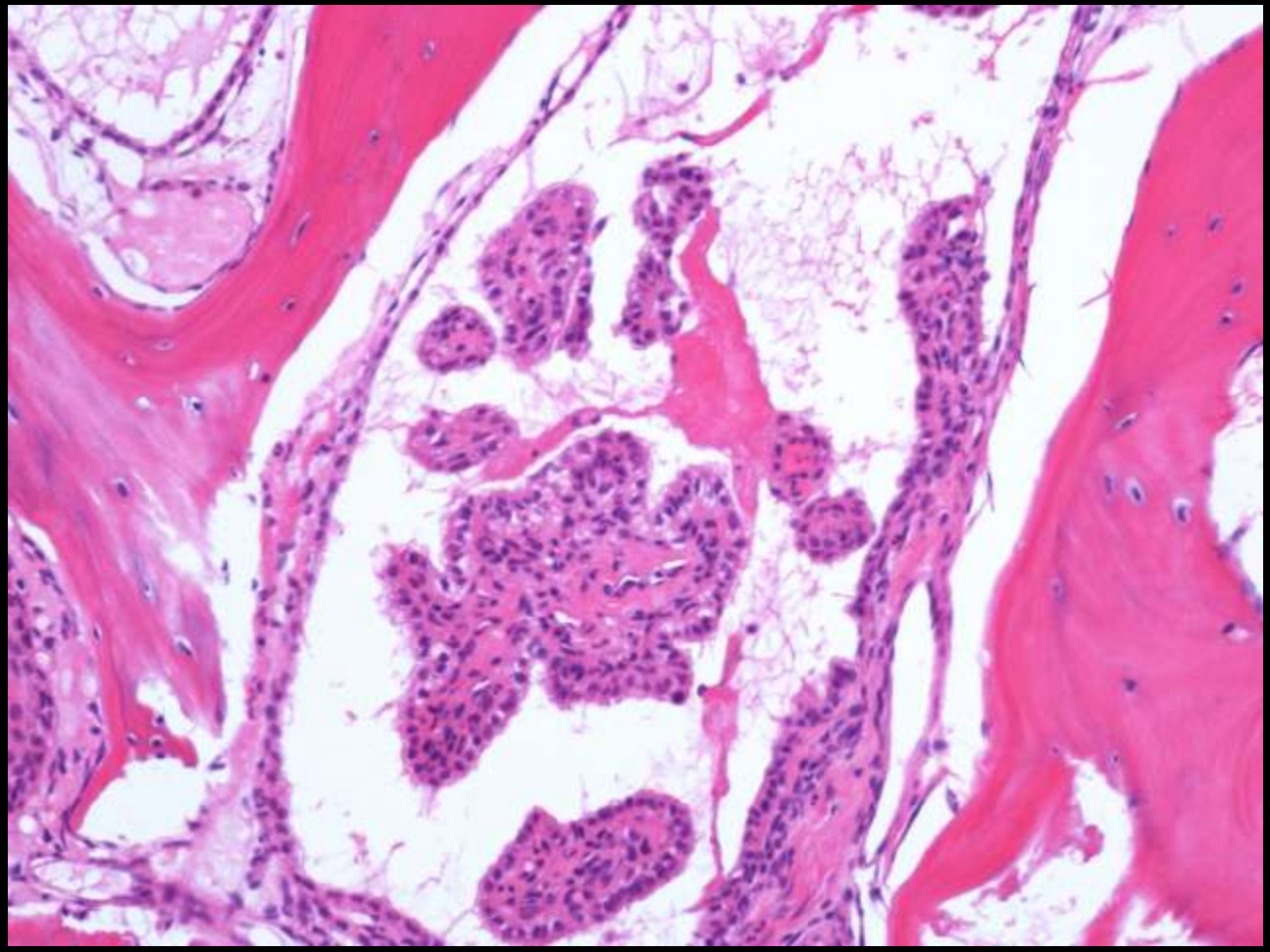
CASE 2

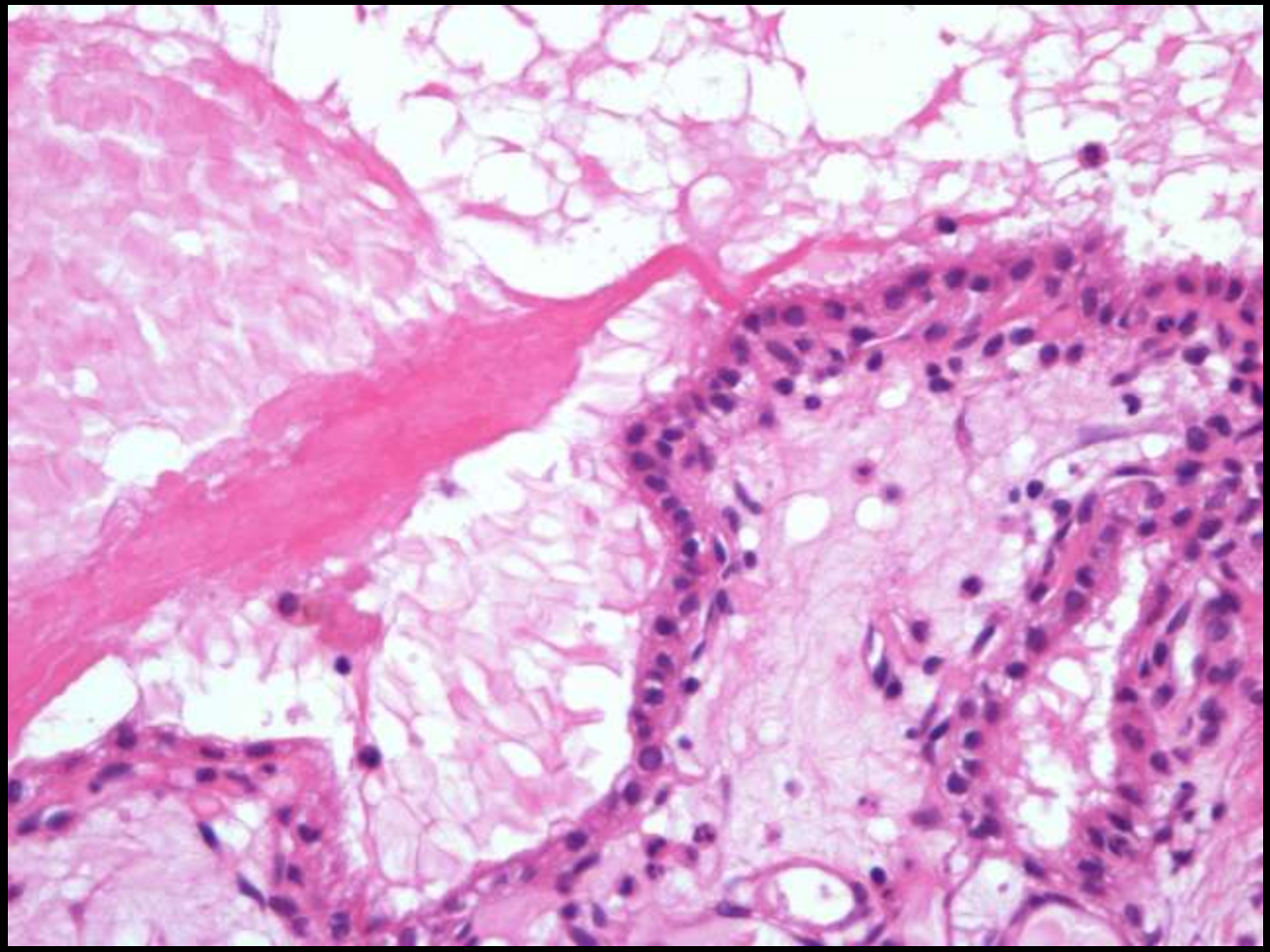
62 year old female- hearing loss and tinnitus. CT showed destructive lesion involving skull base

Case 2

- Architecture is papillary and cystic
- Papillae lined by a single layer of epithelial cells with bland nuclei
- Small glands and follicular structures containing eosinophilic secretions resembling thyroid tissue may be seen
- Mucin stains negative
- Mitoses and necrosis not seen







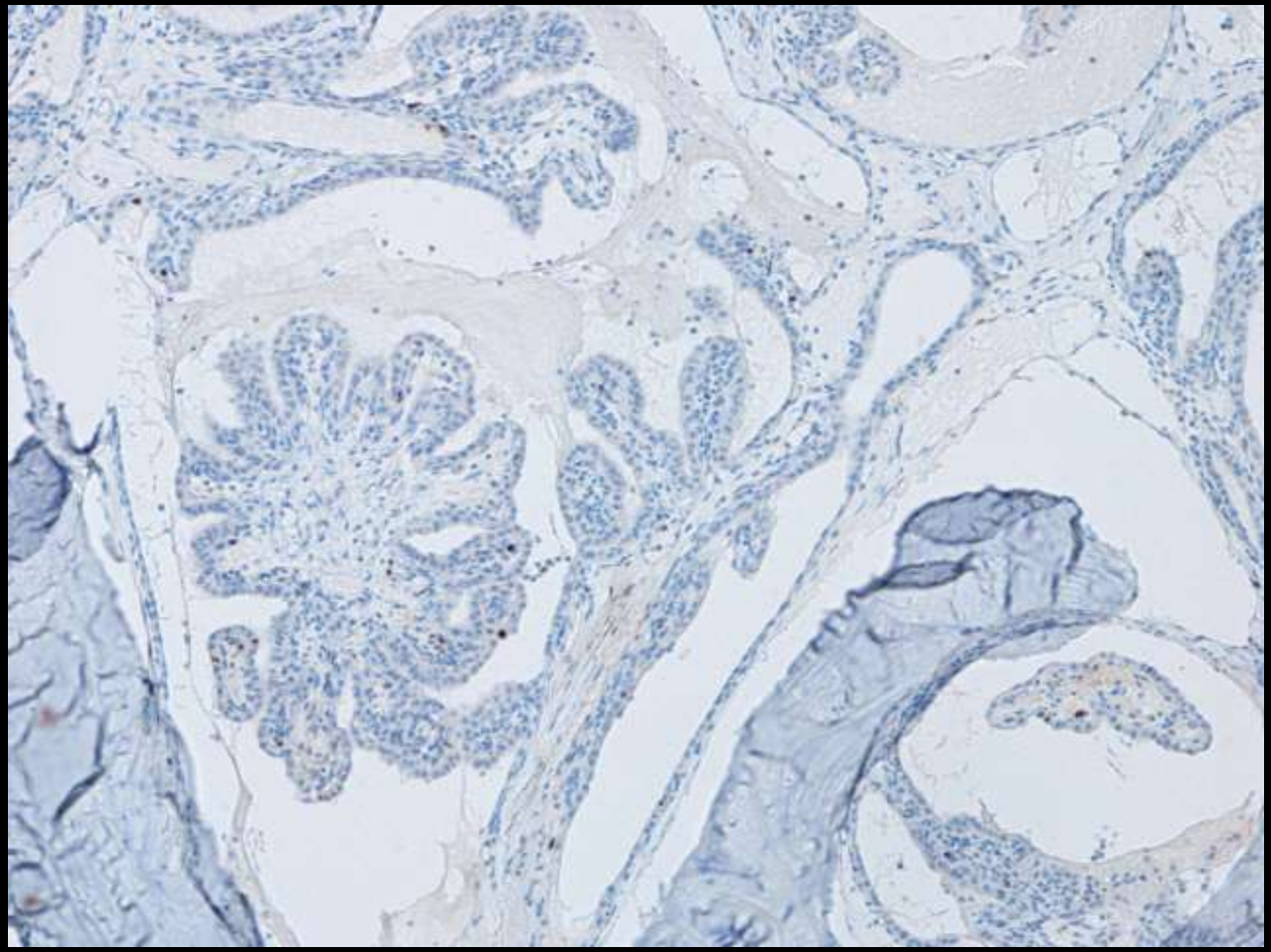
Answers

- Endolymphatic sac tumour
- Metastatic tumour- suggestions included thyroid, prostate or renal primaries
- Papillary ependymoma
- PLGA
- Papillary meningioma
- Papillary variant of craniopharyngioma
- Choroid plexus tumour
- Sinonasal adenocarcinoma

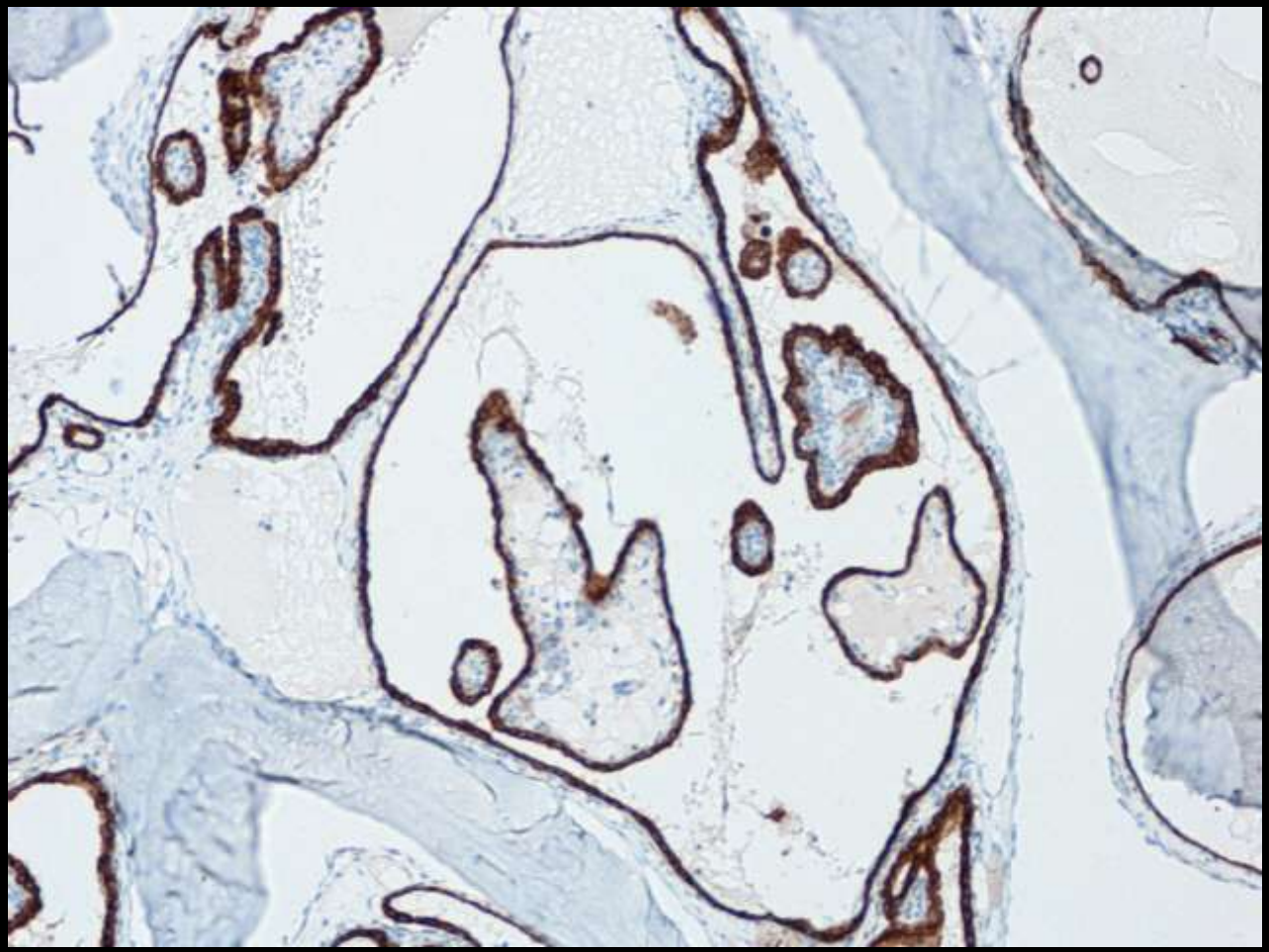
ICC

- CK 7 +, CK20-
- TTF1 -, thyroglobulin-
- CD10-, PAX 8 –
- PSA –
- Ki67 very low

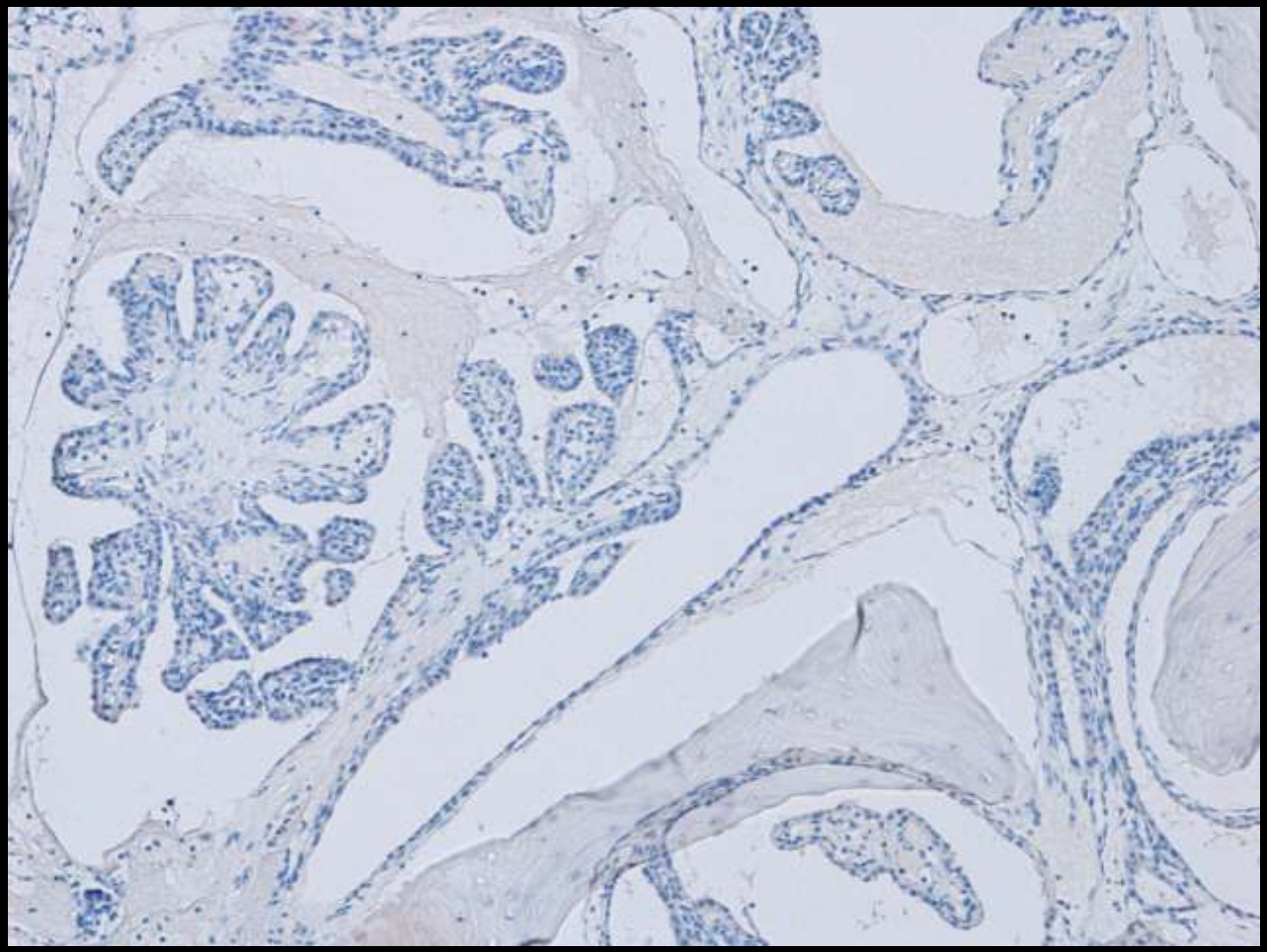
KI67



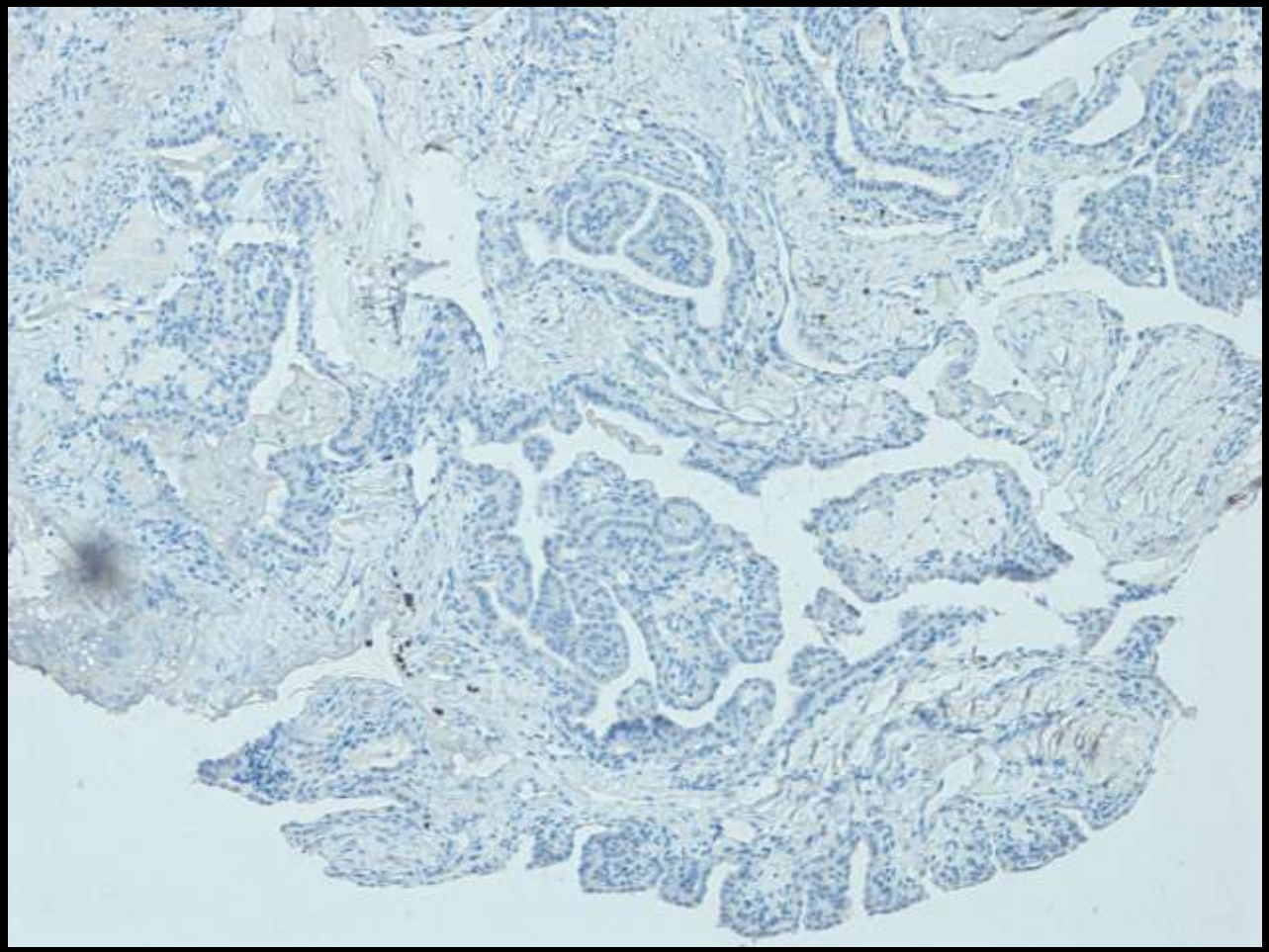
CK7



RCC



TTF1

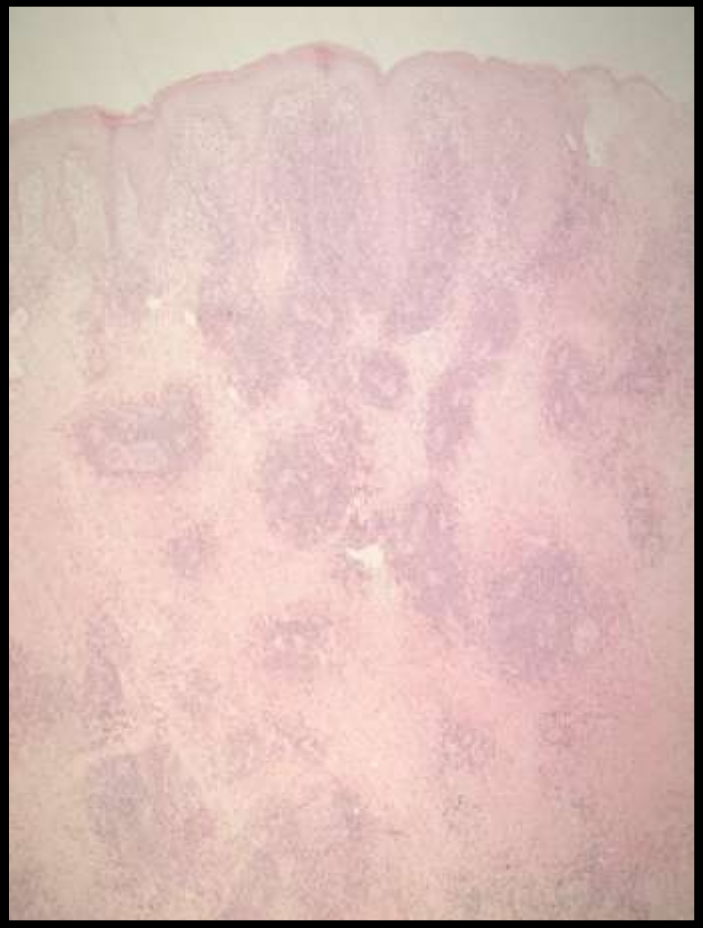


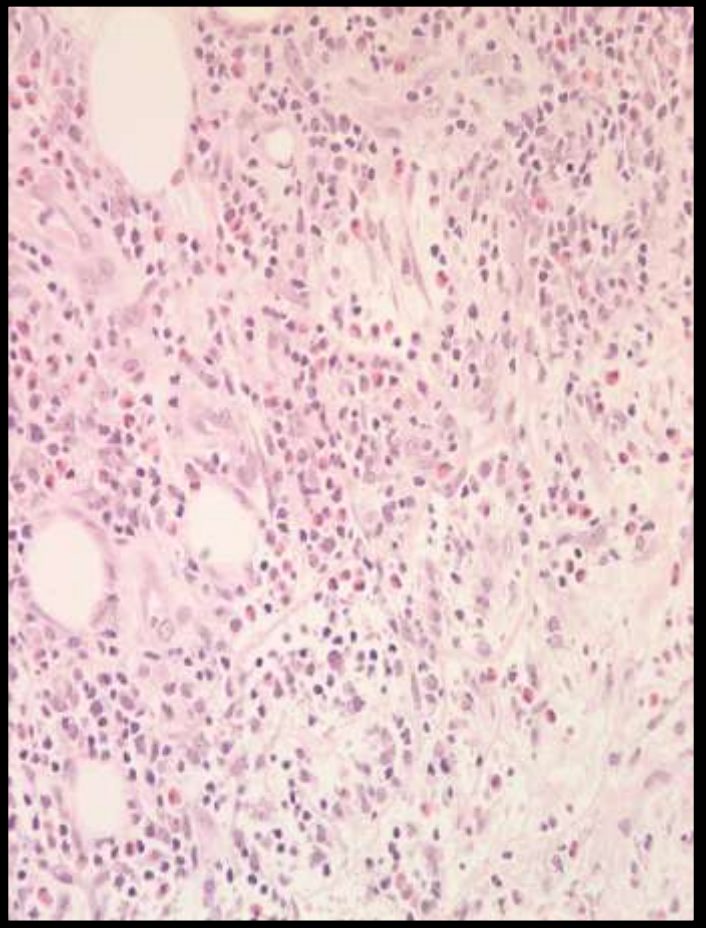
Answer

- Clinically, radiologically and histologically in keeping with **endolymphatic sac tumour** (low grade papillary adenocarcinoma of endolymphatic sac origin)
- 1/3 of cases associated with VHL syndrome
- Treated by complete surgical excision +/- radiotherapy
- Distant mets rare but have been described

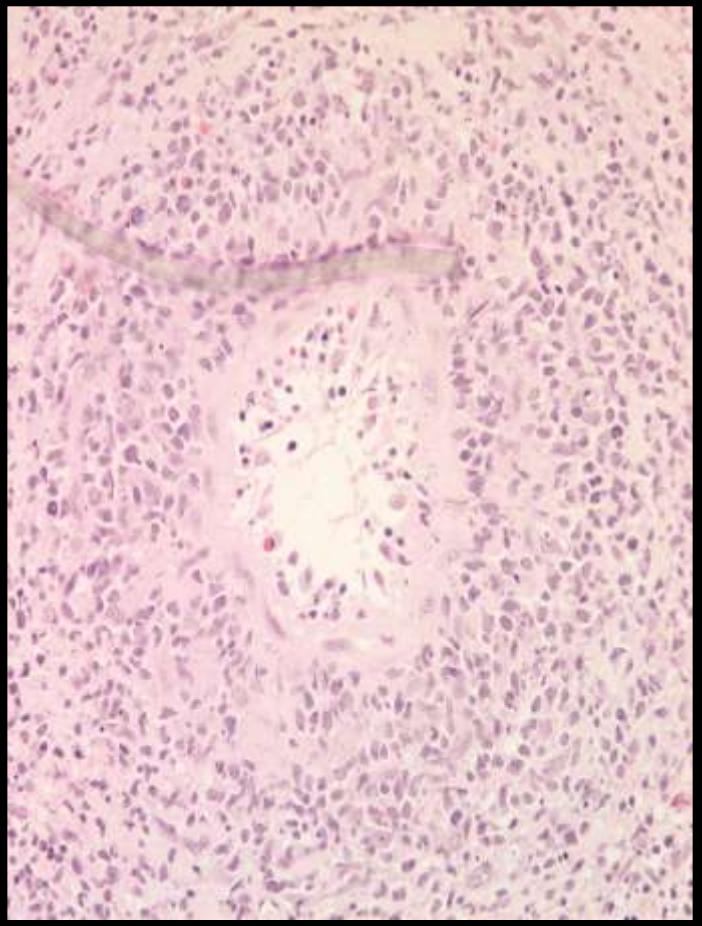
CASE E3

Case E3 Histology

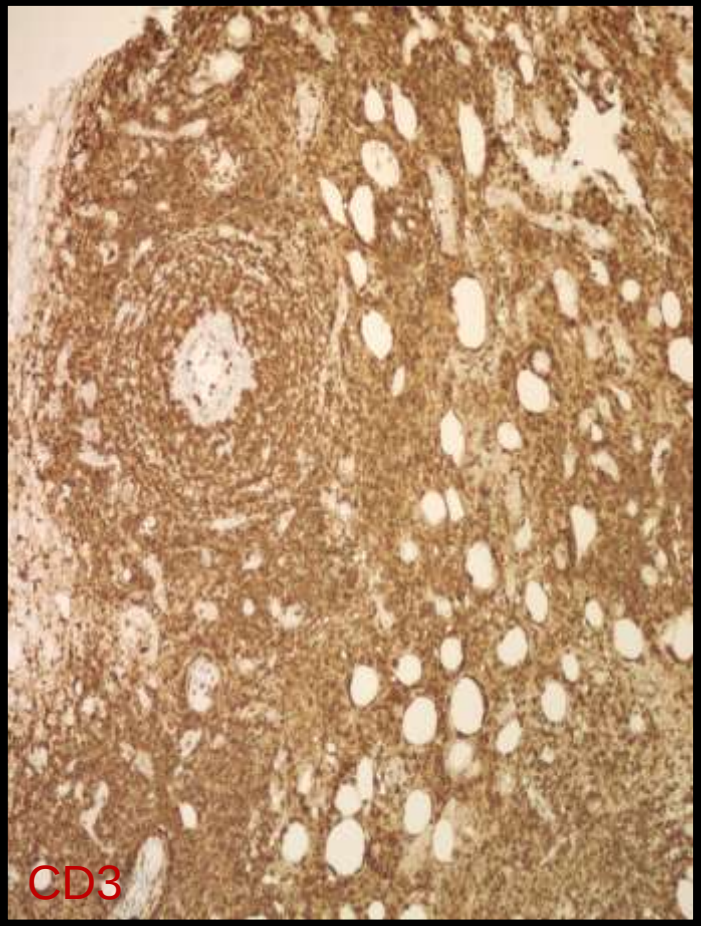


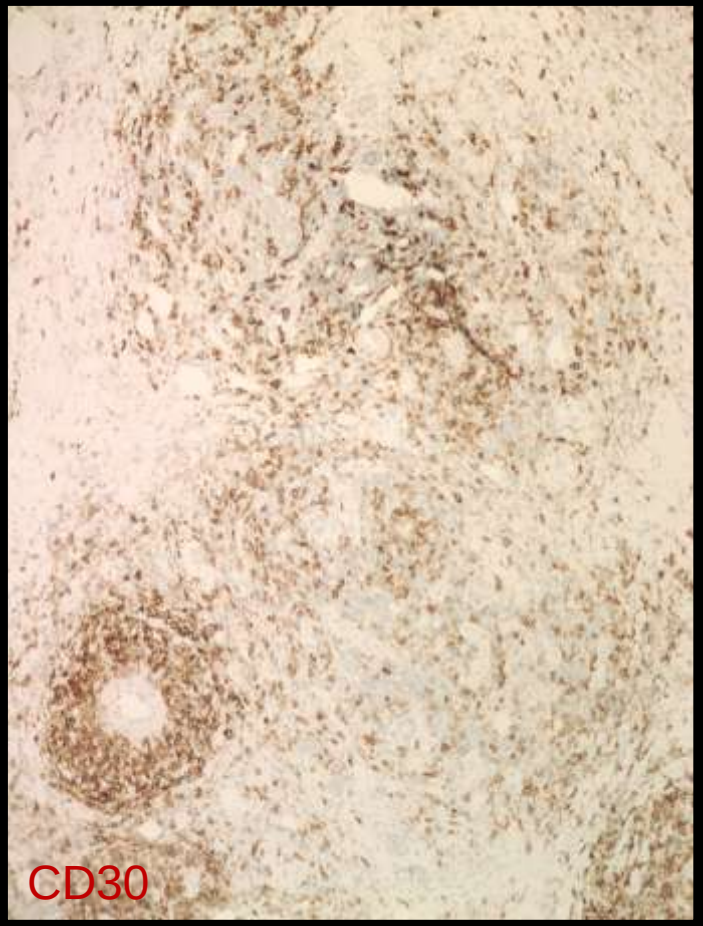


Case E3 Histology



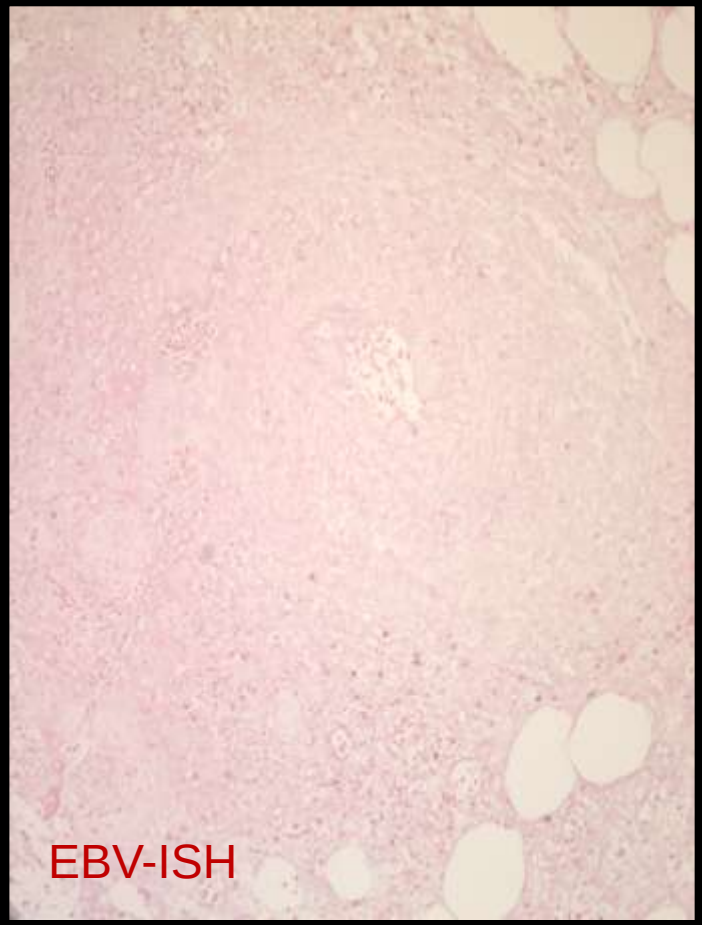
Case E3 IHC





Immunophenotype

- T cell dominant infiltrate
  - CD2+, CD3+, CD5+
  - CD7 down-regulated
  - CD30+
  - CD4+ > CD8+
  - Cytotoxic markers = CD8+
  - CD10 -
  - Alk-1-
  - EBV ISH –
- Some background B



Principal Diagnoses

- Necrotising eosinophilic vasculitis / Churg Strauss 21
- Angiolymphoid hyperplasia with eosinophilia 18
- Lymphomatoid papulosis 13
  - 1 specified Type E
  - 1 further response suggested CD30+ LPD 1
- Angocentric T-NHL / AITL 4

Other Suggested Diagnoses

- Weil's Disease

4

- Insect bite

3

- LCH

5

- Erythema nodosum

1

- Erythema elevatum diutinum

1

LYMPHOMATOID PAPULOSIS, TYPE E

Lymphomatoid papulosis

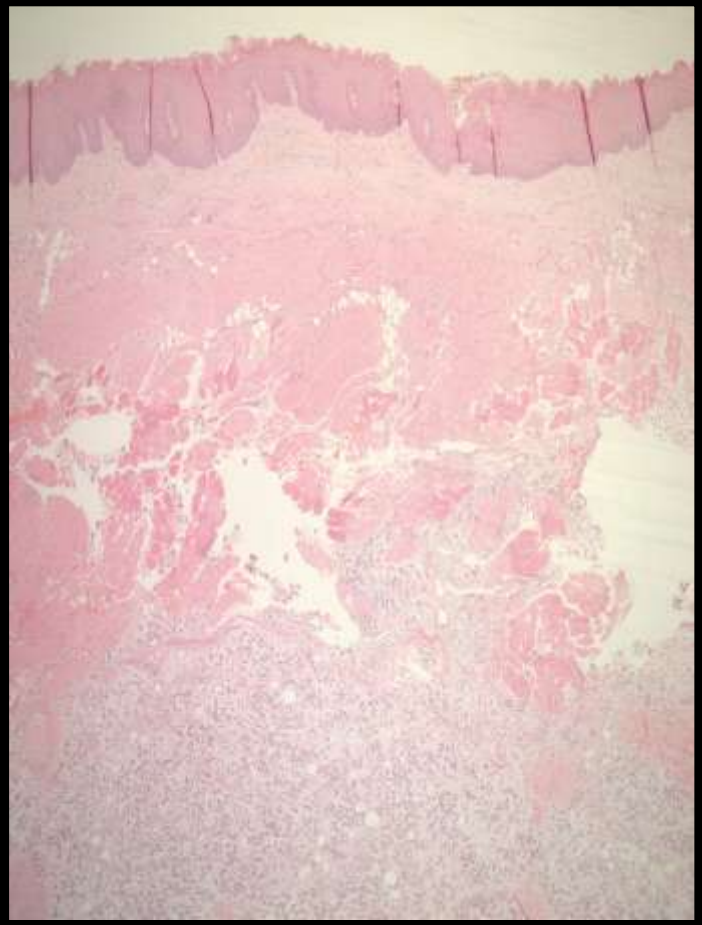
- Primary cutaneous CD30+ disorder
- Clinically characterised by relapsing course of spontaneously resolving papulo-nodular lesions that may ulcerate
- A spectrum of histological features
- Classification into variant forms A to F
- Type A is most common
- Other forms rarer – cases with mixed features may be encountered
- To a greater or lesser extent DDx from T NHL may be difficult

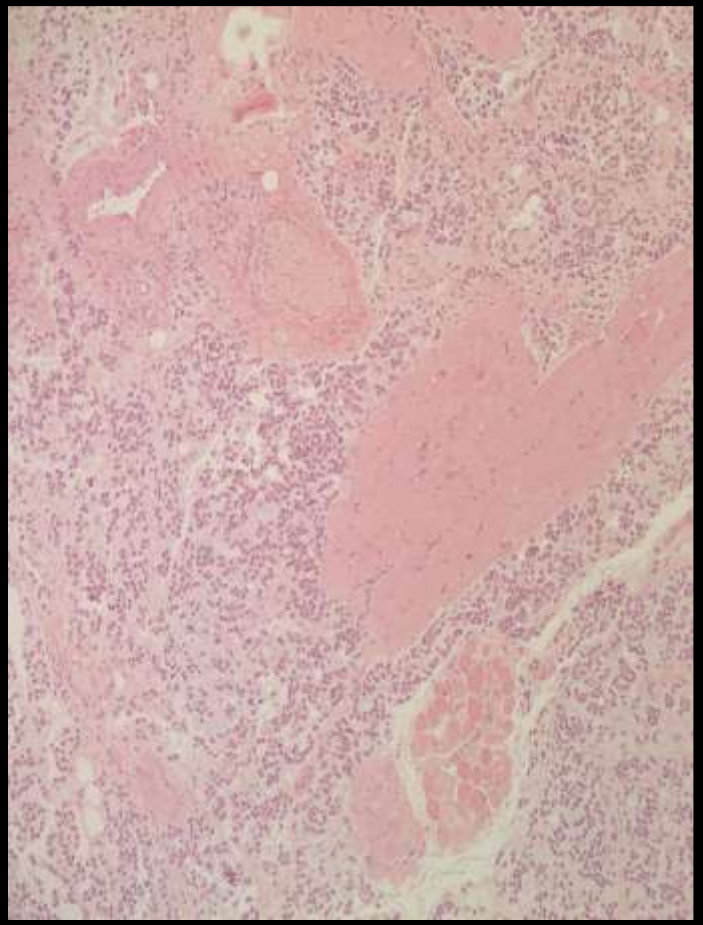
Angio-invasive LyP (Type E)

- Described by Kempf et al (Am J Surg Path, 2013: 37(1); 1-13)
- Lesions grow rapidly and ulcerate.
- Angio-invasive features that with cytological atypia can simulate aggressive T cell lymphomas.
- Atypical cells are of T cell lineage - CD30+ and often CD8+
- Spontaneous regression – complete remission in 9/16 cases.

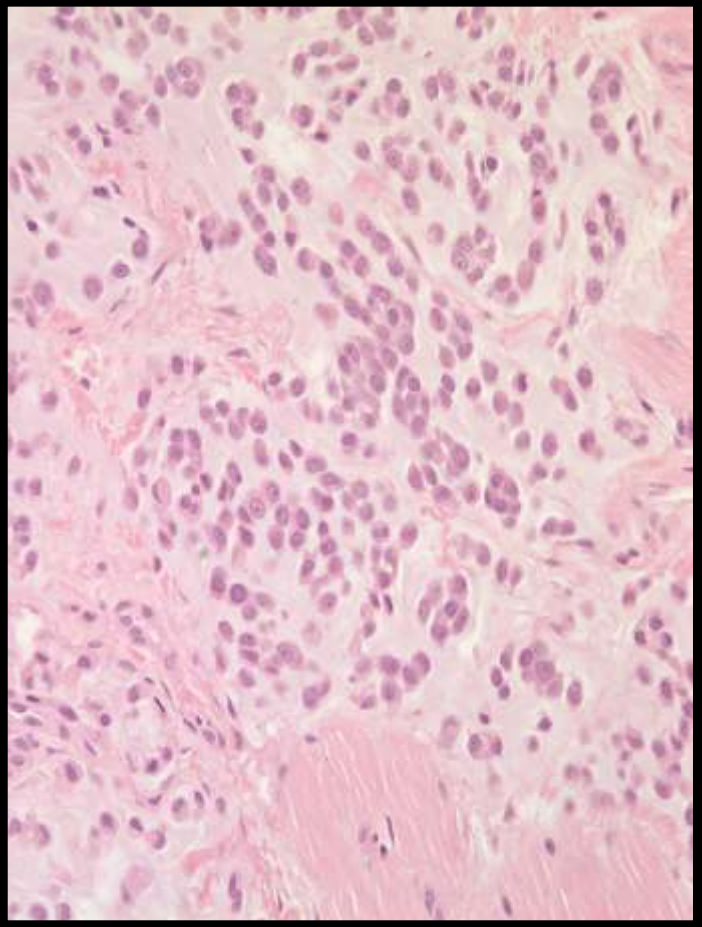
CASE E4

Case E4 Histology

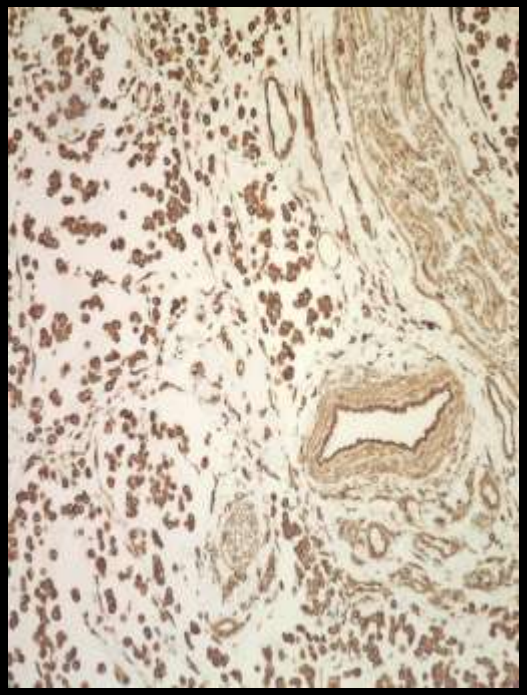




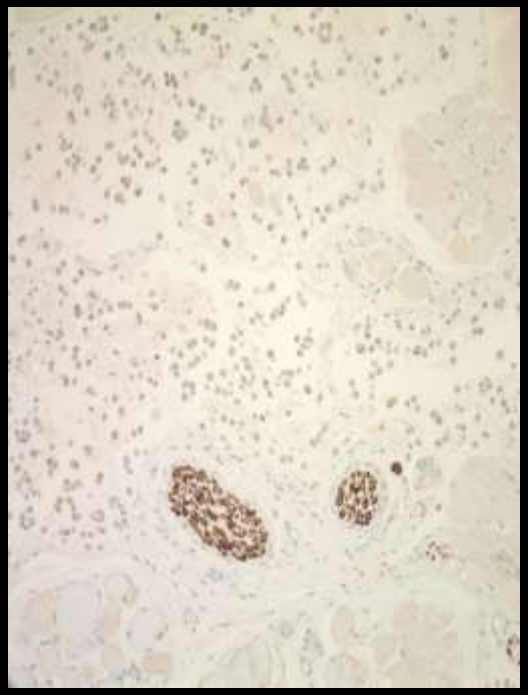
Case E4 Histology



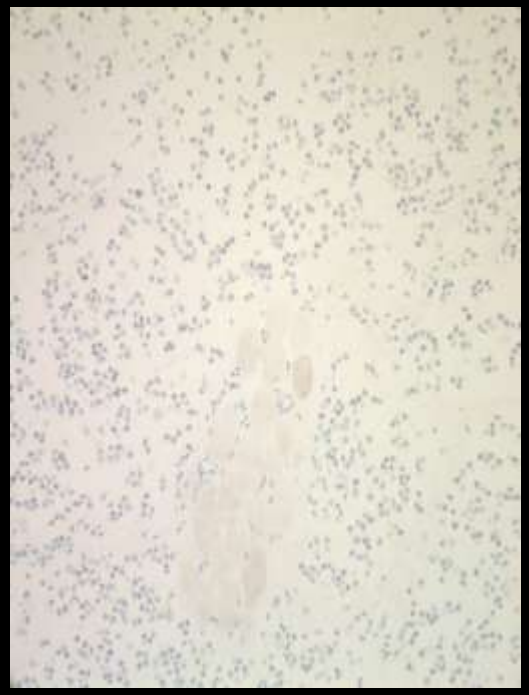
Case E4 Histology



Vimentin

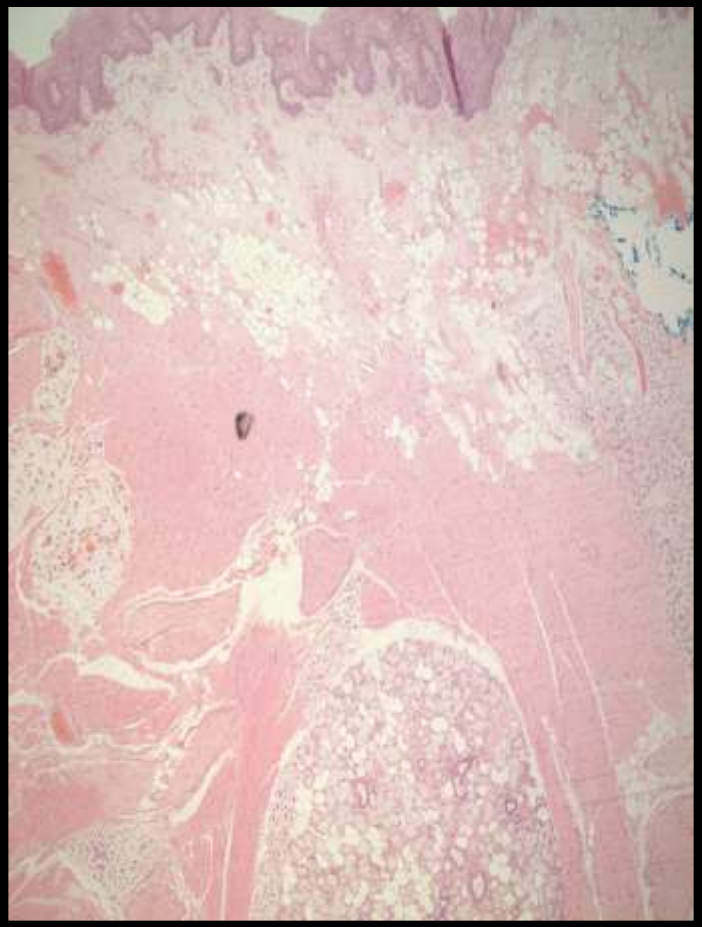


S100



CK

Resection specimen



Principal Diagnoses

- **Myxoid chondrosarcoma 19**
  - **Myxoid sarcoma 3**
  - **Sarcoma 4**
- **Metastatic mucinous adenocarcinoma 14**
- **Ectomesenchymal chondromyxoid tumour 10**

Other Suggested Diagnoses

- Myeloid sarcoma
  3
- Carcinoma ex PSA
  1
- Myoepithelial carcinoma
  6
- Mucoepidemoid carcinoma
  1
- Clear cell carcinoma
  1
- Malignant lesion
  2
- PSA
  5
- Monomorphic adenoma / myoepithelioma
  3

METASTATIC MYXOID CHONDROSARCOMA

Metastatic myxoid chondrosarcoma

- Differential diagnosis of myxoid chondrosarcoma
- In this case
  - Pulmonary lesions on radiology – new and rapid development
  - Comparison with the original resection specimen
    - Specialist review
  - (Genetic abnormalities)
- Ectomesenchymal chondromyxoid tumour