CASE OF THE MONTH APRIL 2019

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CASE PRESENTATION

- 75 Year old male
- Presented with hemoptysis
- Suspected case of carcinoma lung

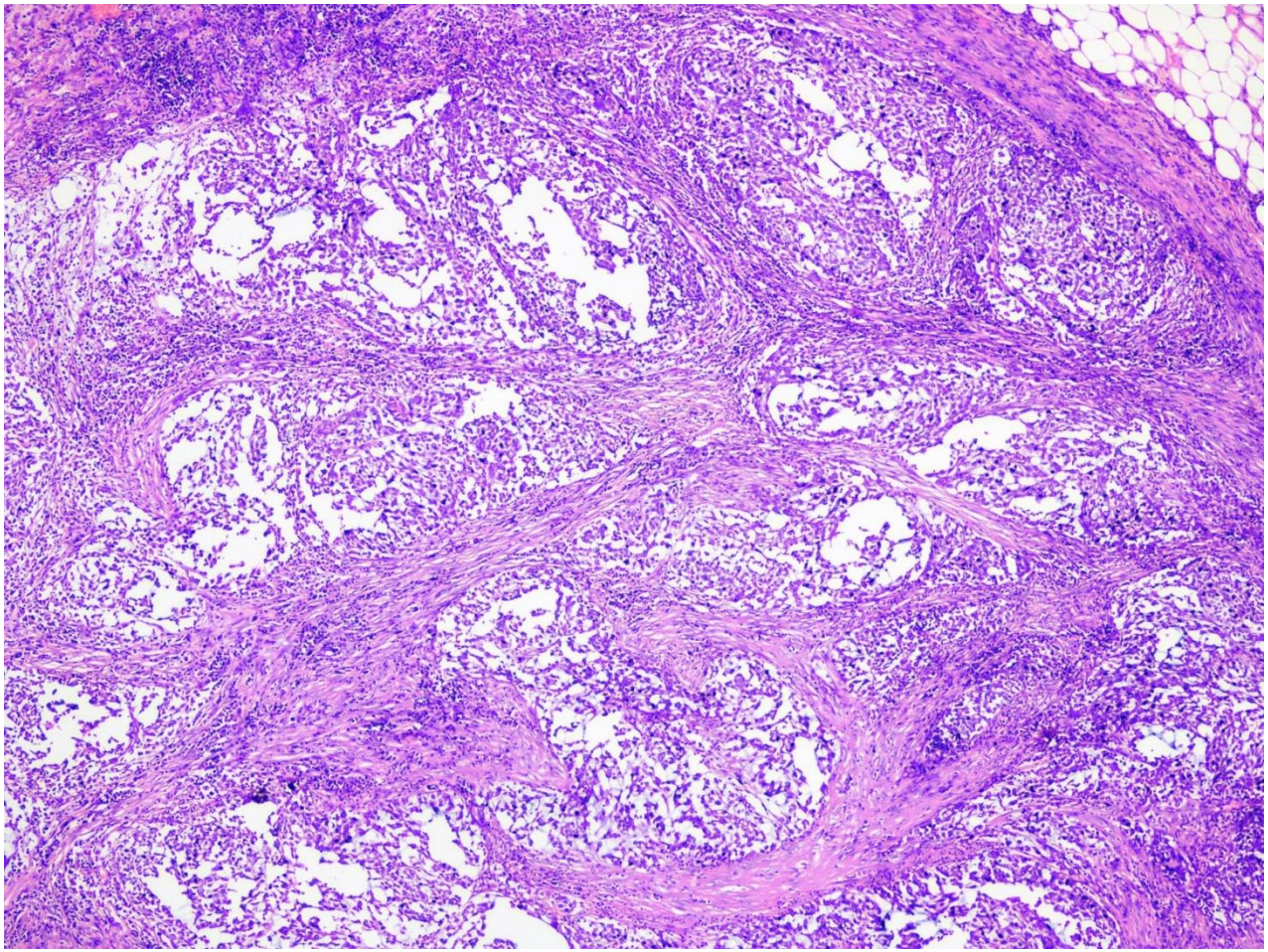
PET-CT

- Mass lesion measuring 9.1 x 7.3 x 8.0 cm
- Infiltrating the diaphragm
- Involve the lower lobe of lung
- Abutting the spleen and left adrenal
- Paratracheal and bilateral hilar lymphnodes

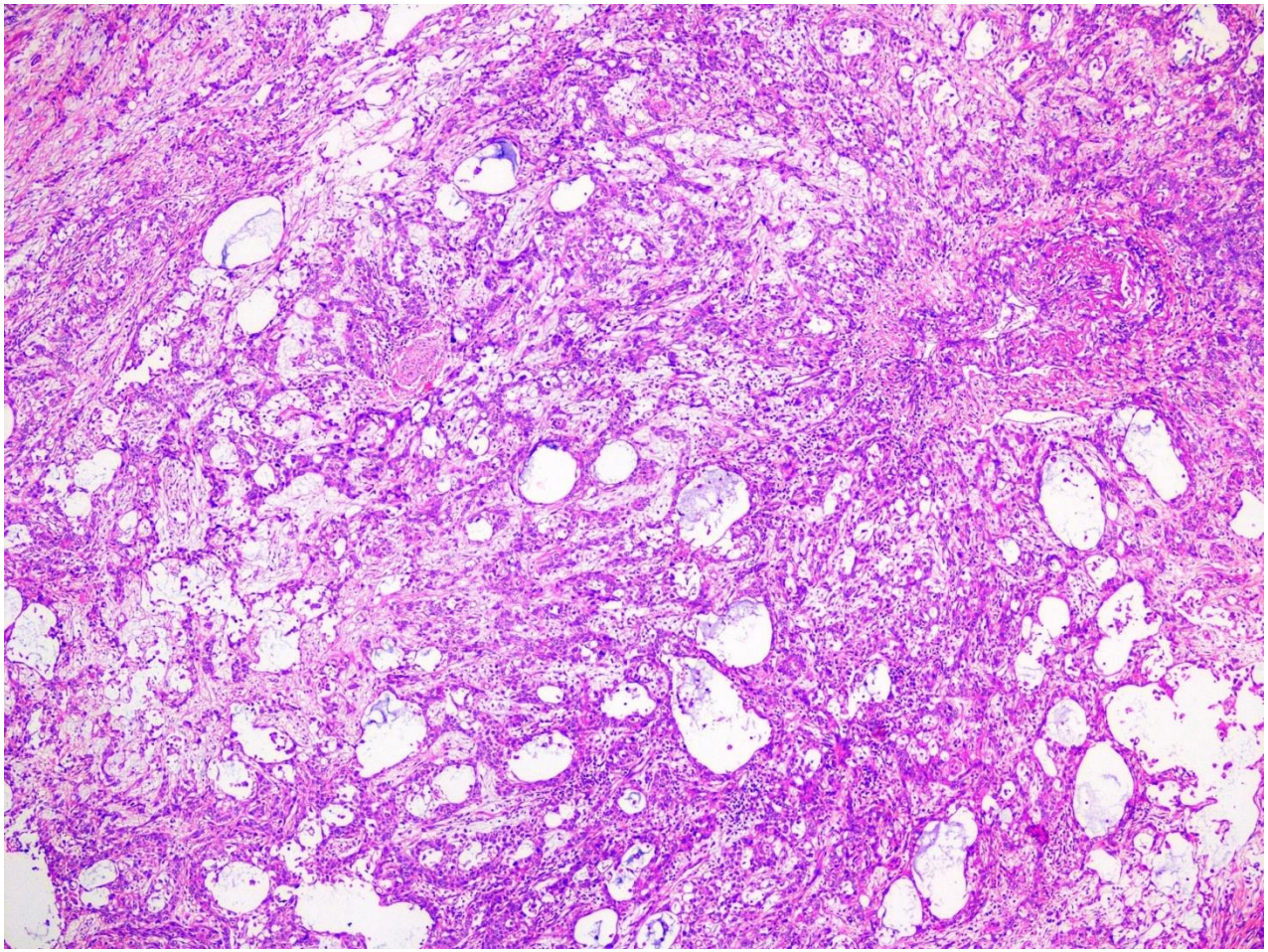
Procedure

- Wedge resection of left lower lobe lung

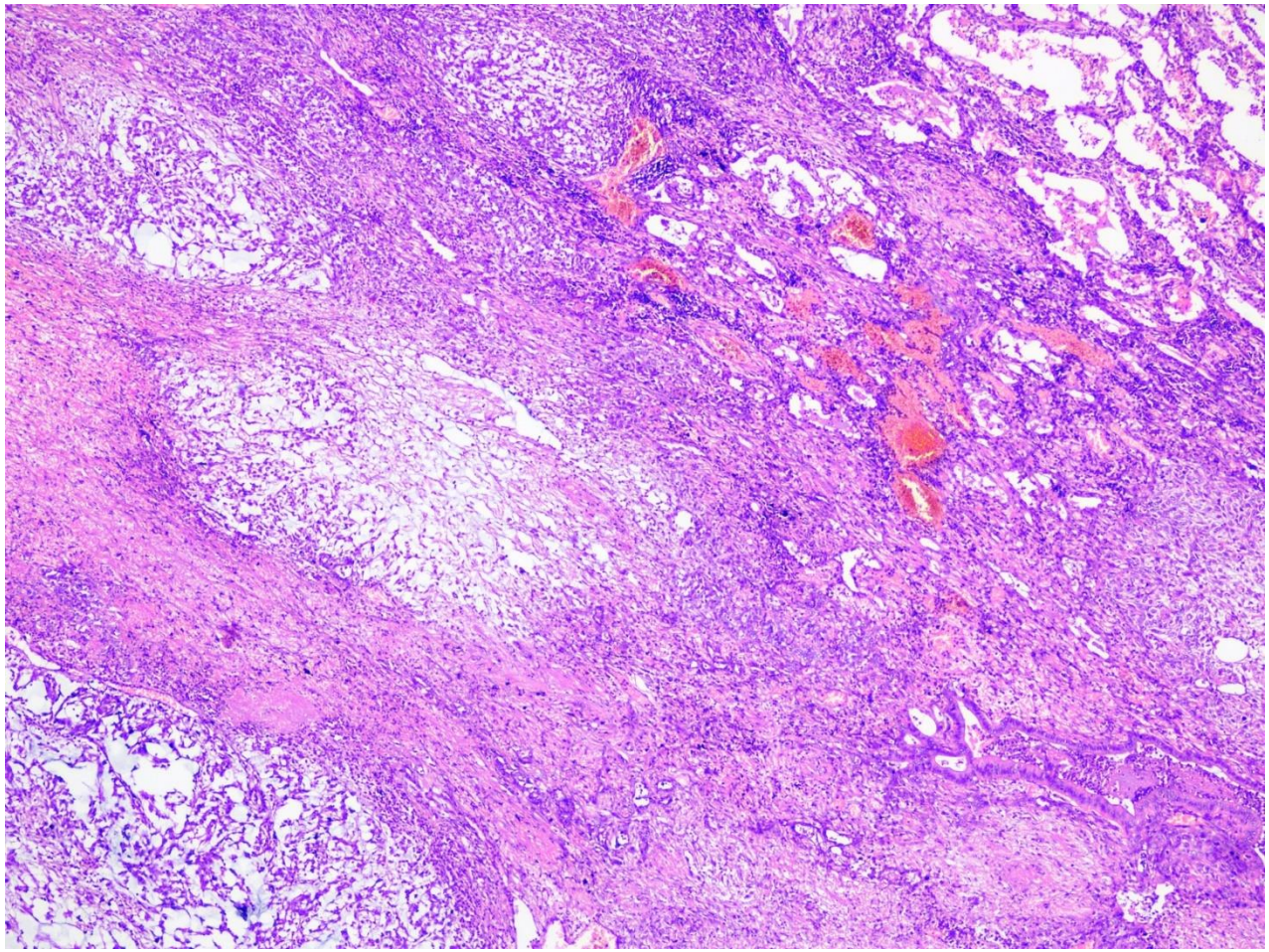
Histopathology



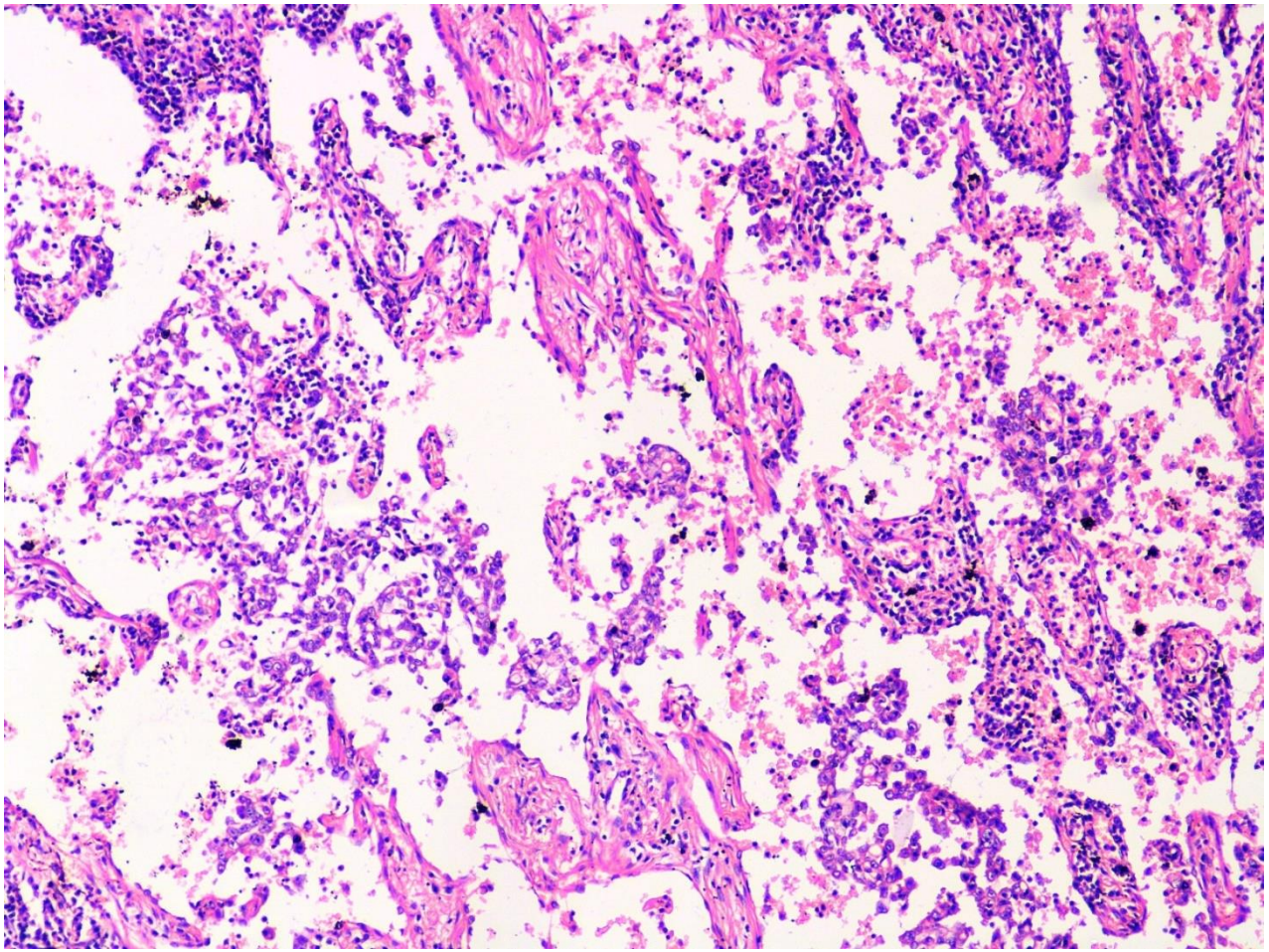
Tumour present predominantly in nodular and reticular growth pattern



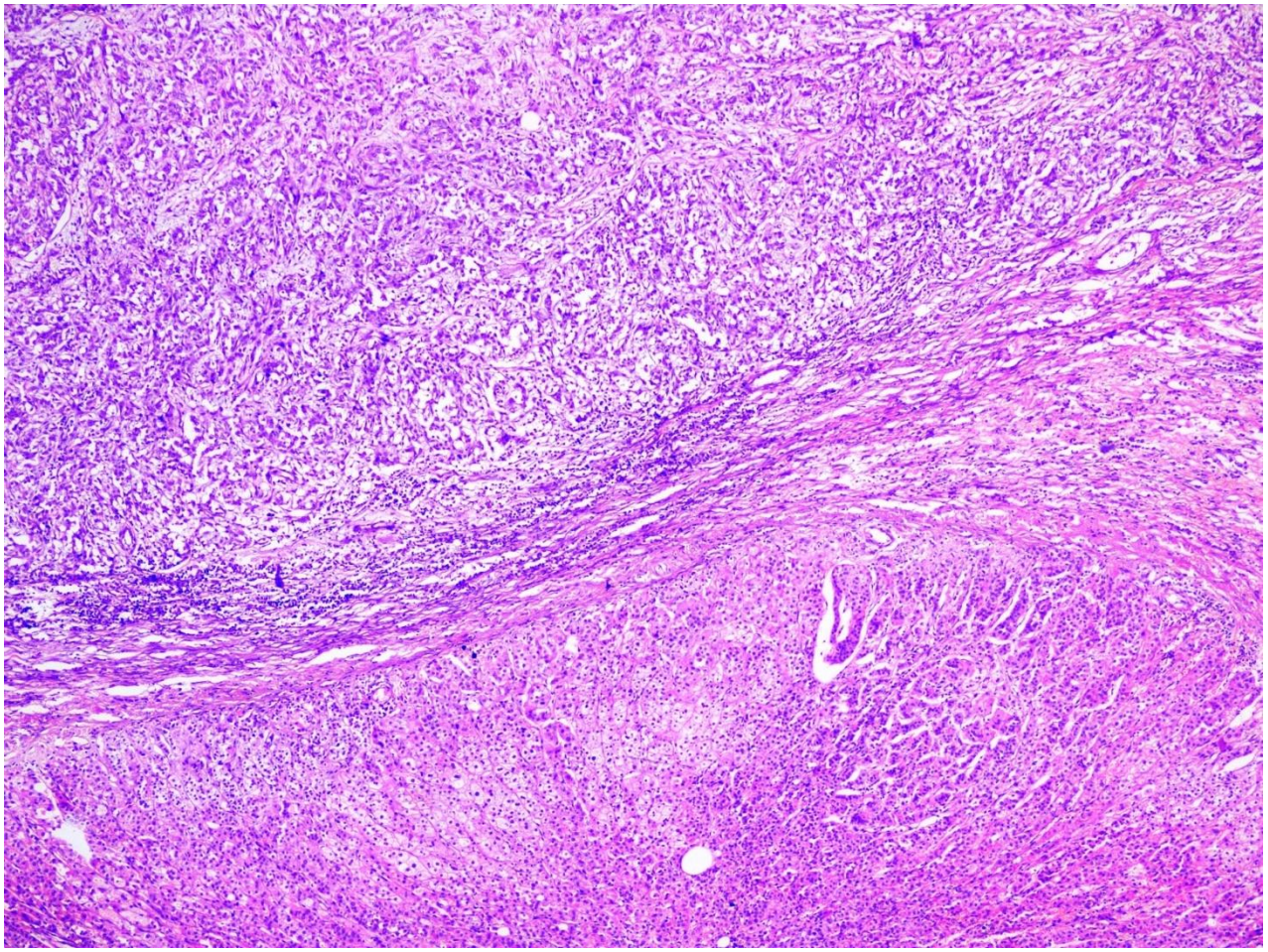
Reticular pattern of growth with loosely textured myxoid background



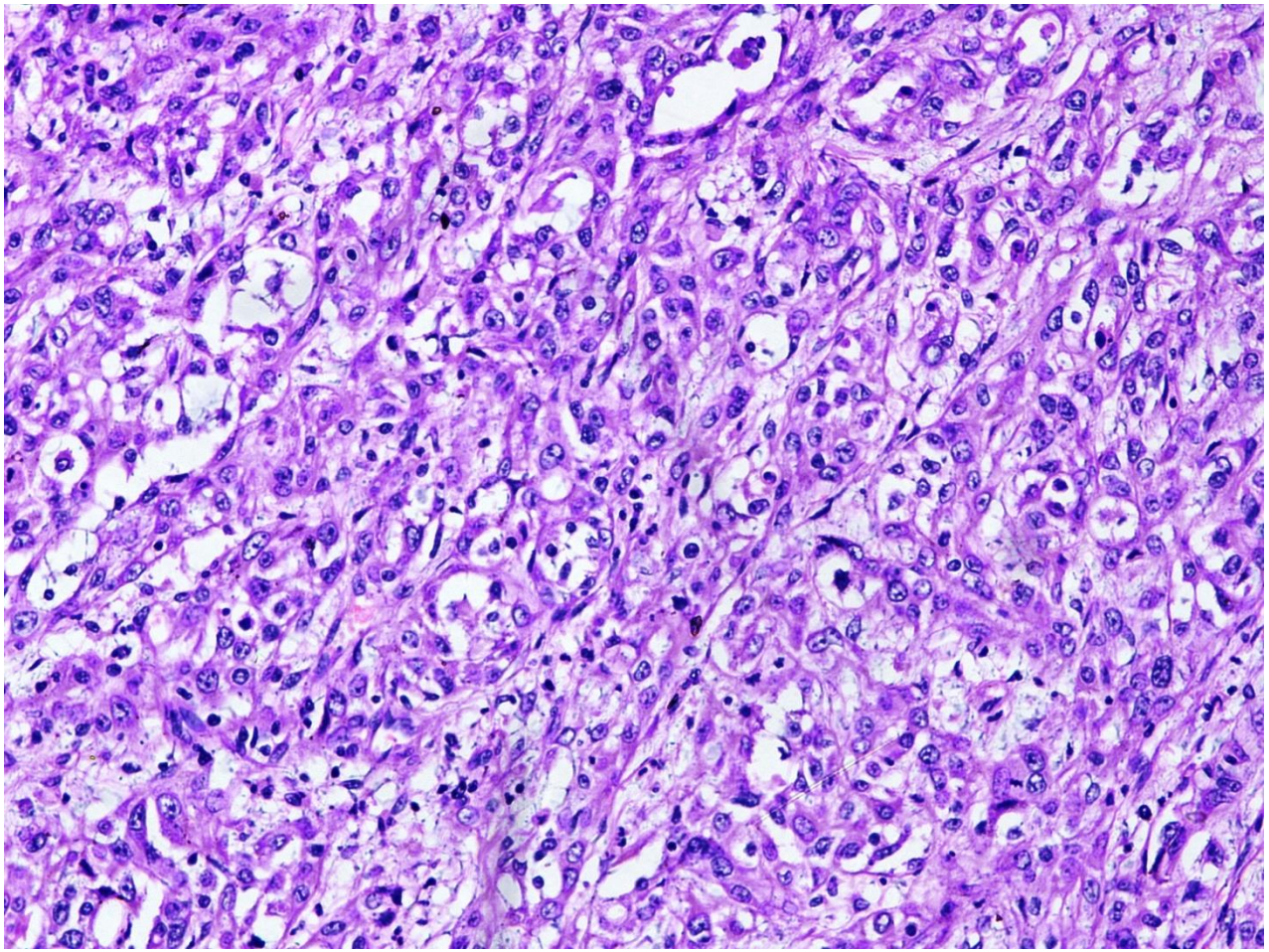
Lung infiltration and areas of necrosis



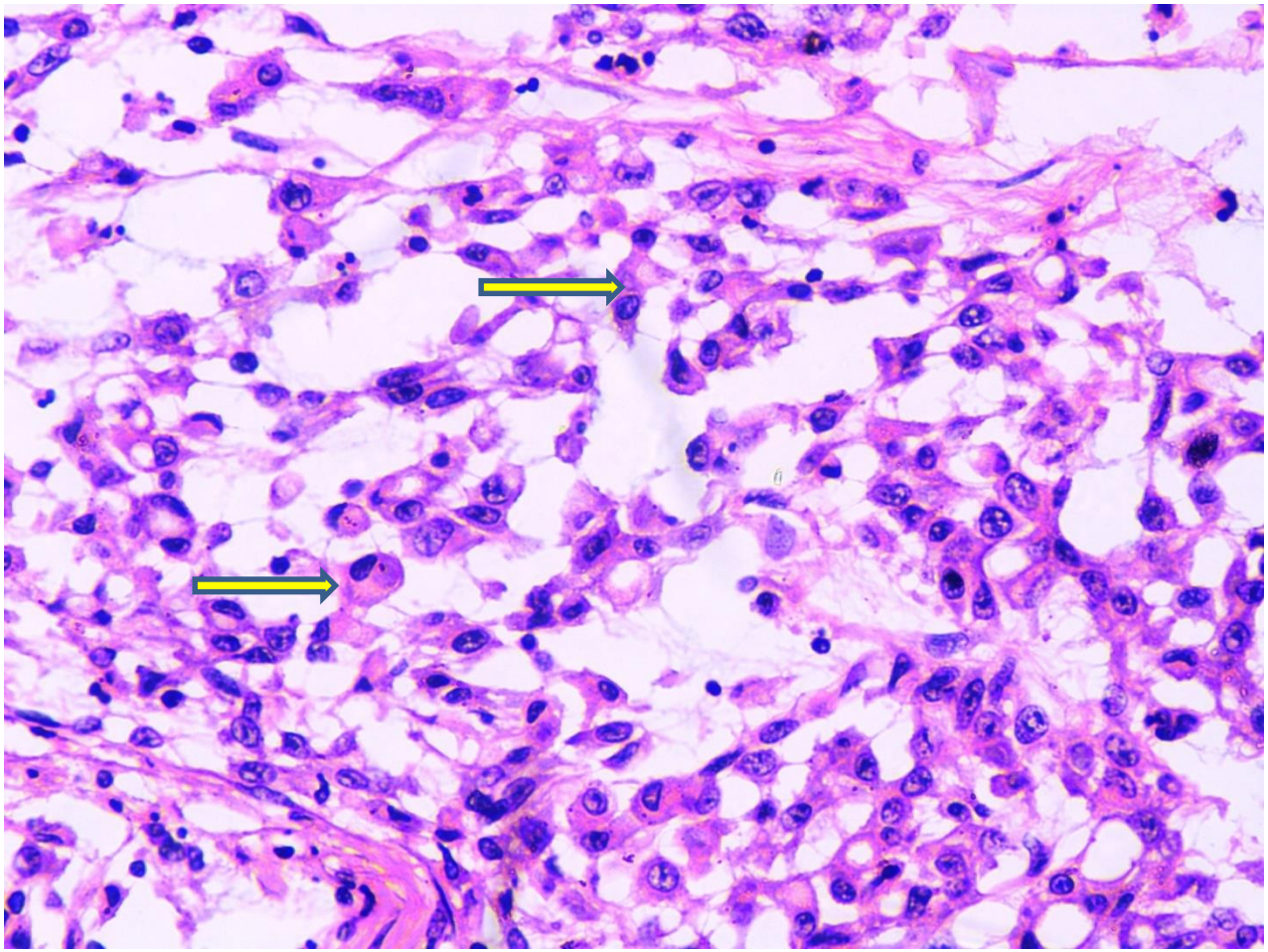
Tumour showing STAS (spread through air spaces)



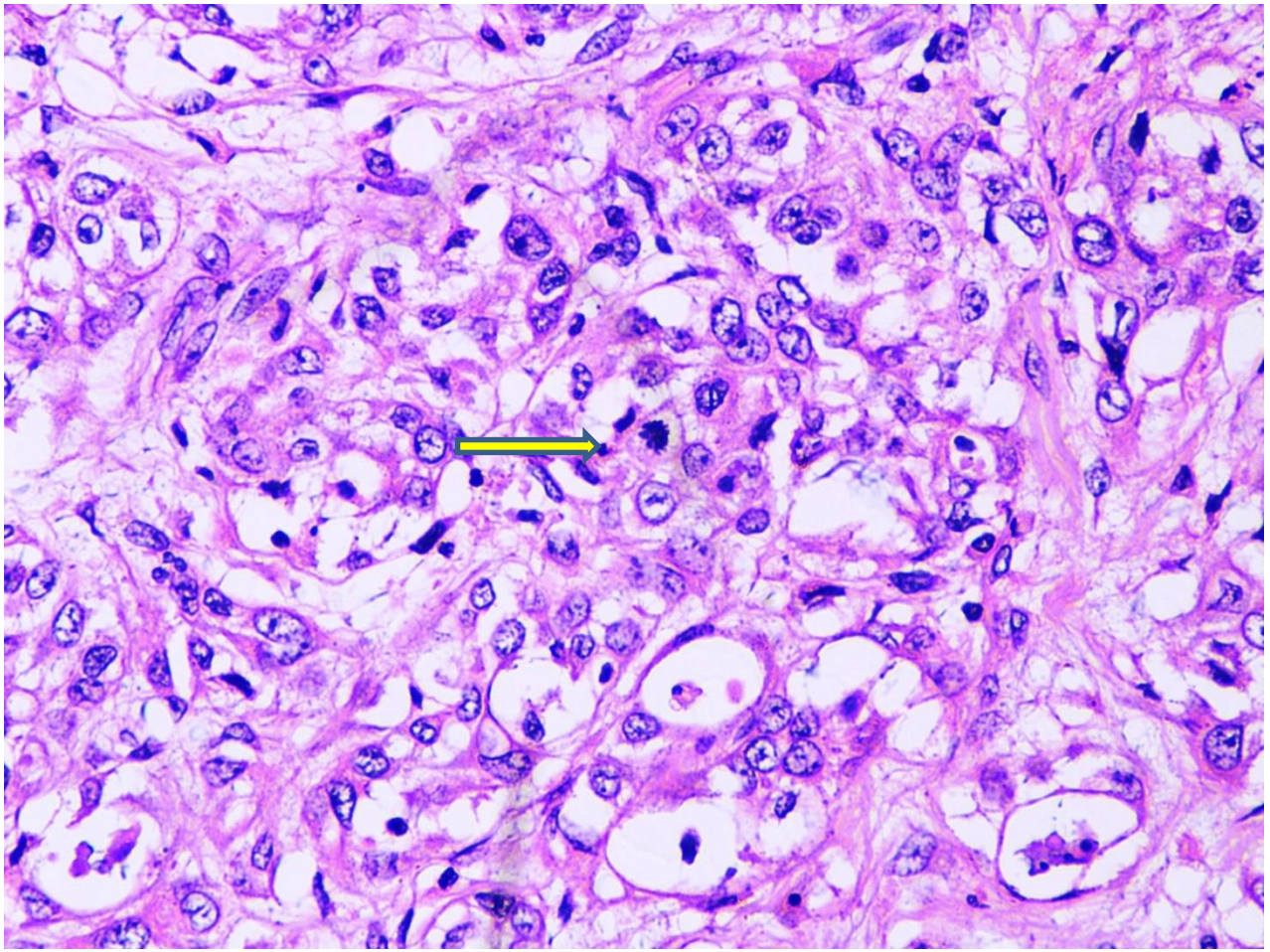
Abutting adrenal but not infiltrating



Cells are predominantly epithelioid, spindle with focal rhabdoid differentiation



Rhabdoid phenotype with abundant eosinophilic cytoplasm and cytoplasmic holes



Cells are having prominent nucleoli and brisk mitosis

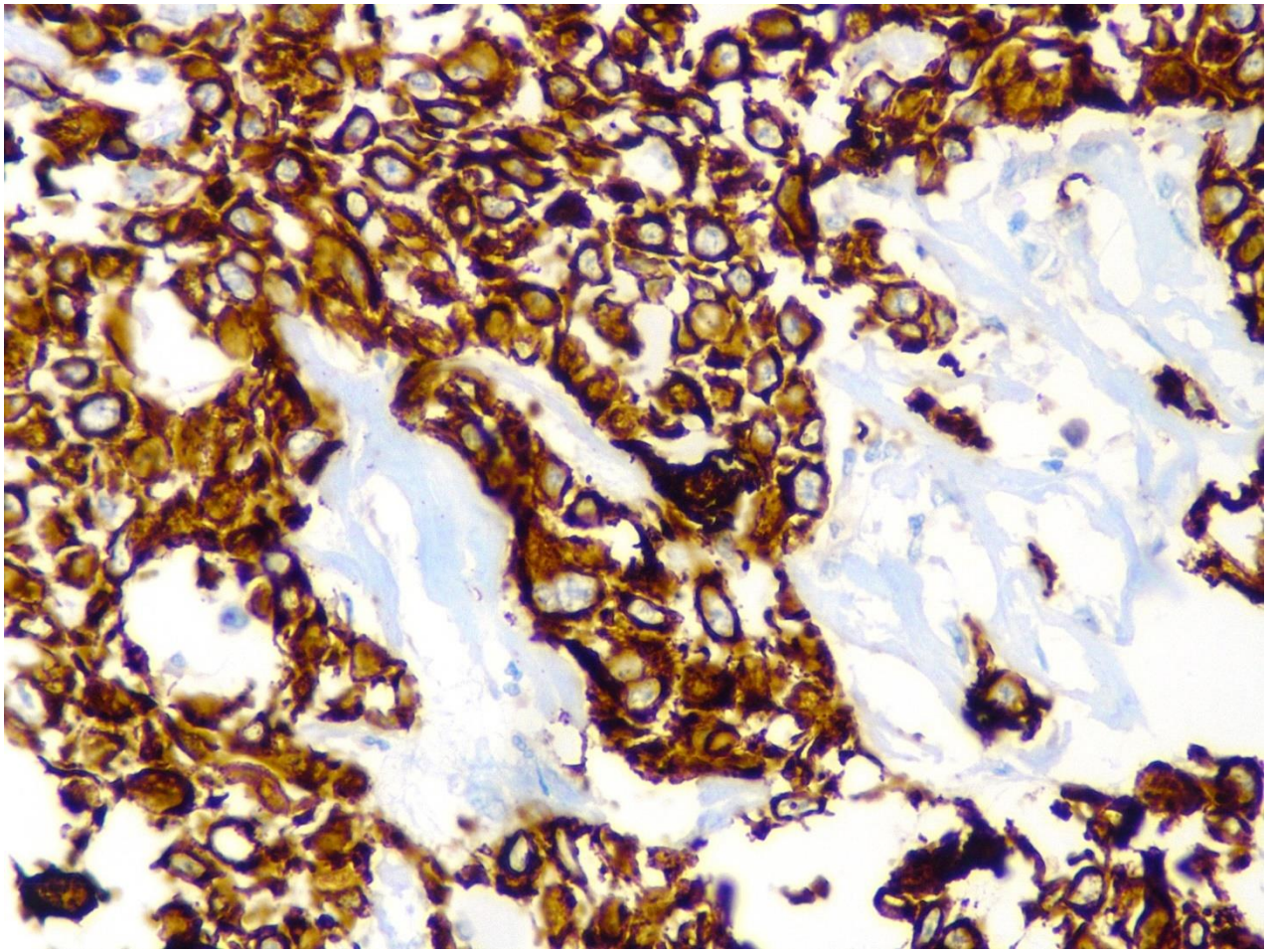
Summary of Histomorphological features

- Predominant pattern- nodular and reticular
- Cells are predominantly
  - Epithelioid
  - Spindled
  - Focal rhabdoid differentiation
- Brisk mitosis and necrosis evident
- Infiltrative pattern

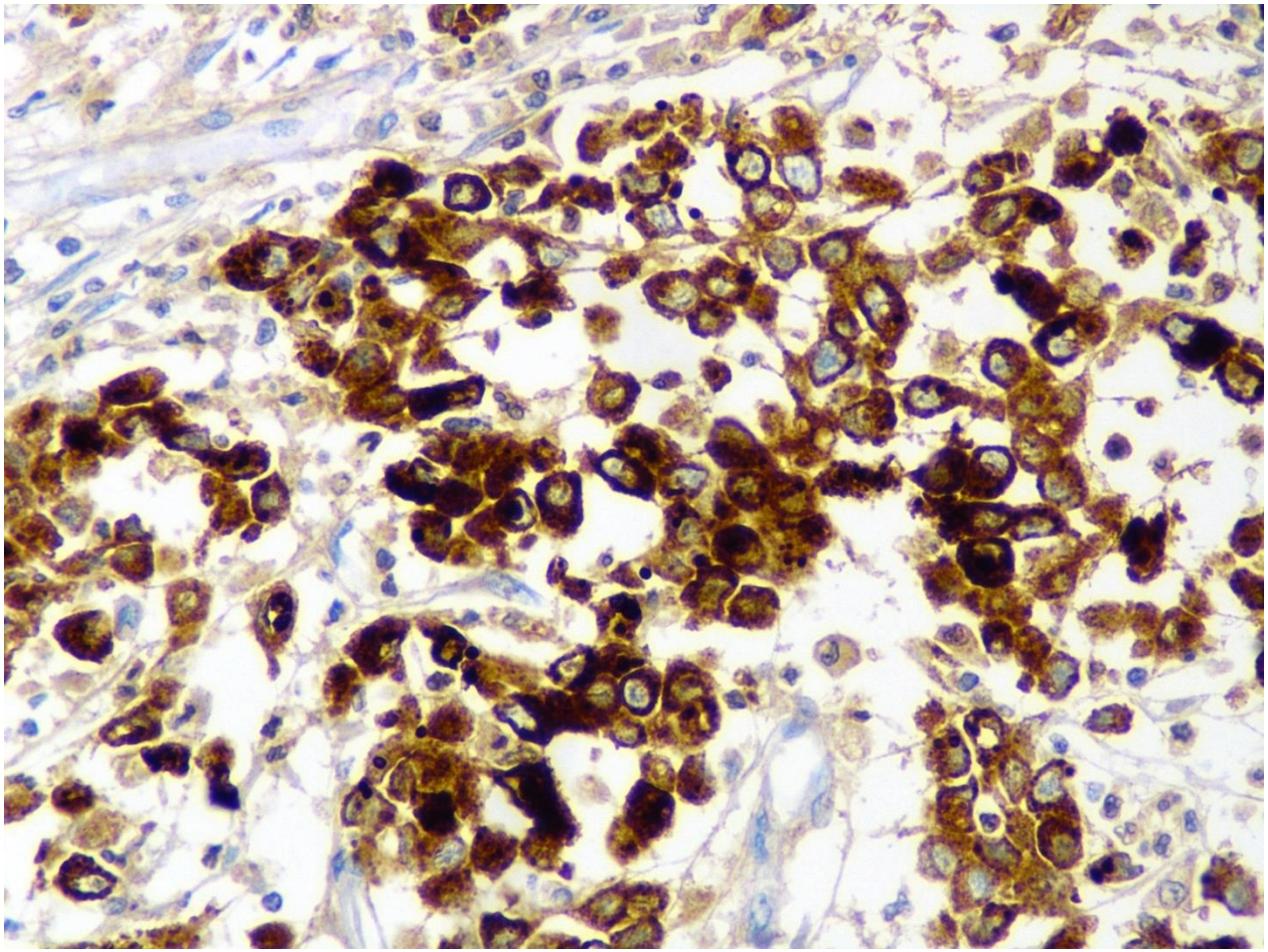
Differential diagnosis based on Histomorphology

- Myoepithelioma
- Hemangioendothelioma
- Mesothelioma
- Adenocarcinoma

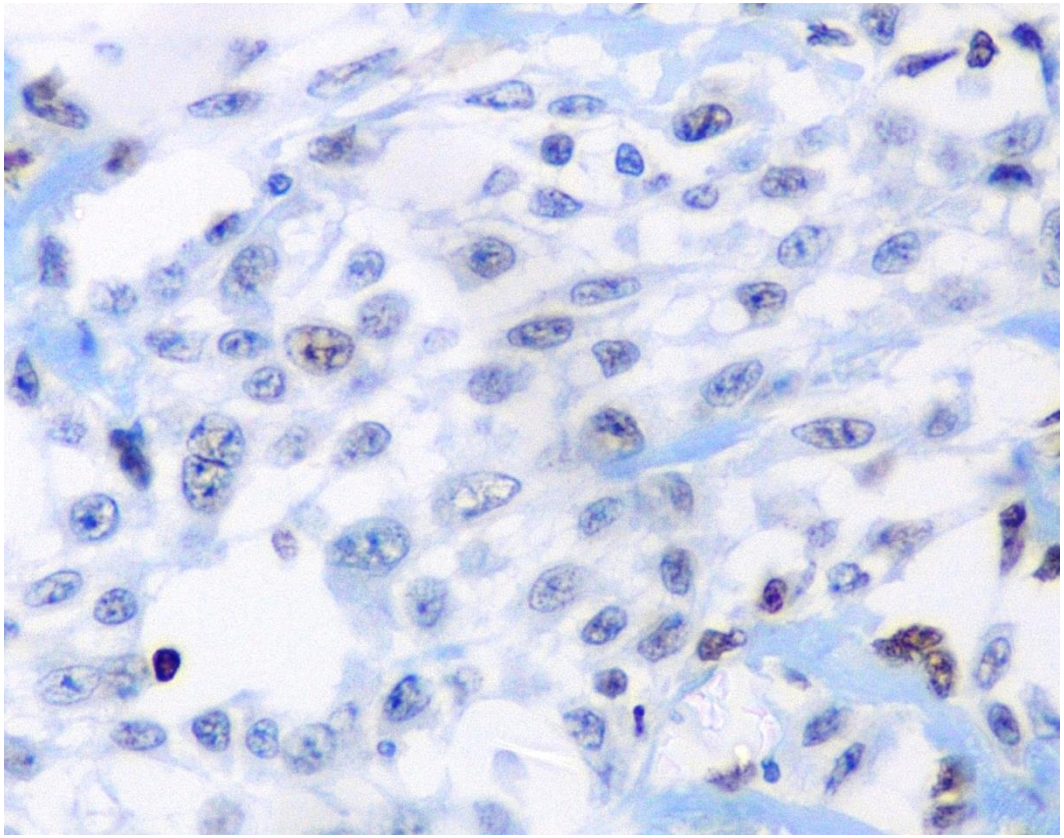
Immunohistochemistry (IHC)



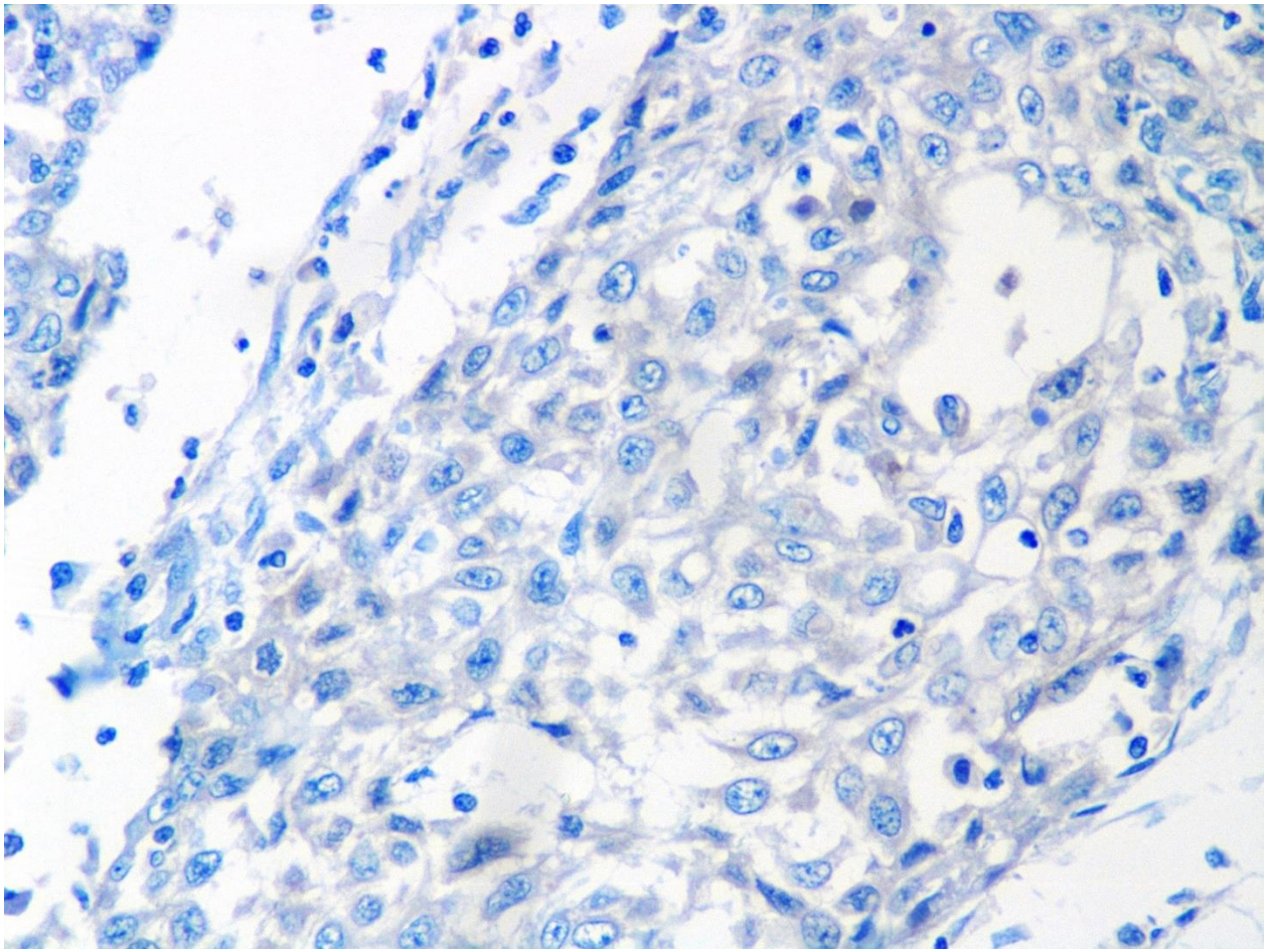
Cells are diffusely and strongly positive for CK



Cells are diffusely and strongly positive for EMA



Cells are showing
INI 1 Loss



Negative markers on IHC

Calponin , S-100, CK 5/6 , SMA, ERG, CD 31, FLI 1, SMA, GFAP, P40, BerEP4, TTF 1, CK 7, Calretinin, WT 1, D2-40, SOX 2

Summary of IHC

- Diffusely and strongly positive for CK and EMA
- INI 1 loss
- List of tumours showing INI1 loss
  - Myoepithelial carcinoma
  - Malignant rhabdoid and teratoid tumour
  - Epithelioid malignant MPNST
  - Extraskeletal myxoid chondrosarcoma
  - Epithelioid sarcoma

Differential diagnosis

Tumour with CK and EMA positivity and INI1 loss

- Epithelioid malignant MPNST
- Myoepithelial carcinoma
- Epithelioid sarcoma

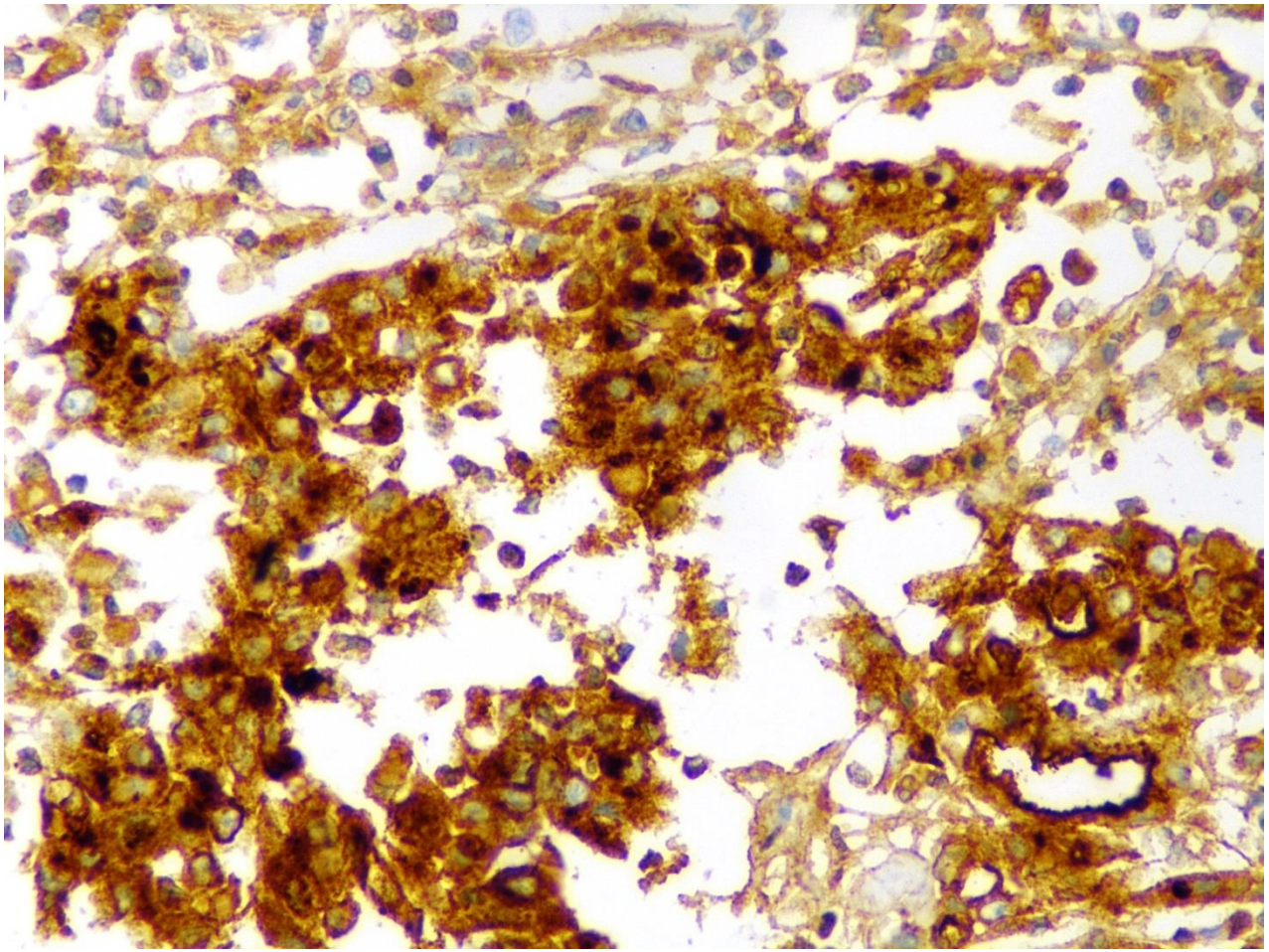
Ruling out differentials according to Negative markers on IHC

- Calponin
- CK 5/6
- S-100

Myoepithelial carcinoma

- S-100

Epithelioid MPNST



Cells are diffusely and strongly positive for CA 125

Final diagnosis

Primary Proximal epithelioid sarcoma of lung

Discussion

- This case has been presented due to rarity of this tumour and there are many morphological and immunohistochemical differential diagnosis for rhabdoid phenotype

- This case was previously diagnosed as mesothelioma

Primary proximal epithelioid sarcoma of lung

- Primary proximal epithelioid sarcoma of lung is a rare entity
- They are found commonly in the extremities.
- Lung as a primary site is very rare.
- Epithelioid sarcomas are divided according to location into proximal type or axial type when found above the elbow.
- Occur in young age and has poor prognosis than conventional type.

- Histologically
  - Cells are epithelioid with some rhabdoid features
  - Significant lymphohistiocytic infiltration
  - Brisk mitoisis
  - Necrosis
- IHC
  - Positive for CK, EMA, CD 34 (50% cases)
  - CA 125 positive
  - INI 1 loss

- Diagnosis is challenging due to varied histomorphological features
- It should be aided by immunohistochemistry to rule out all the differential diagnosis.
- Surgical resection and adjuvant chemotherapy may be a reasonable curative treatment option for stage III PES