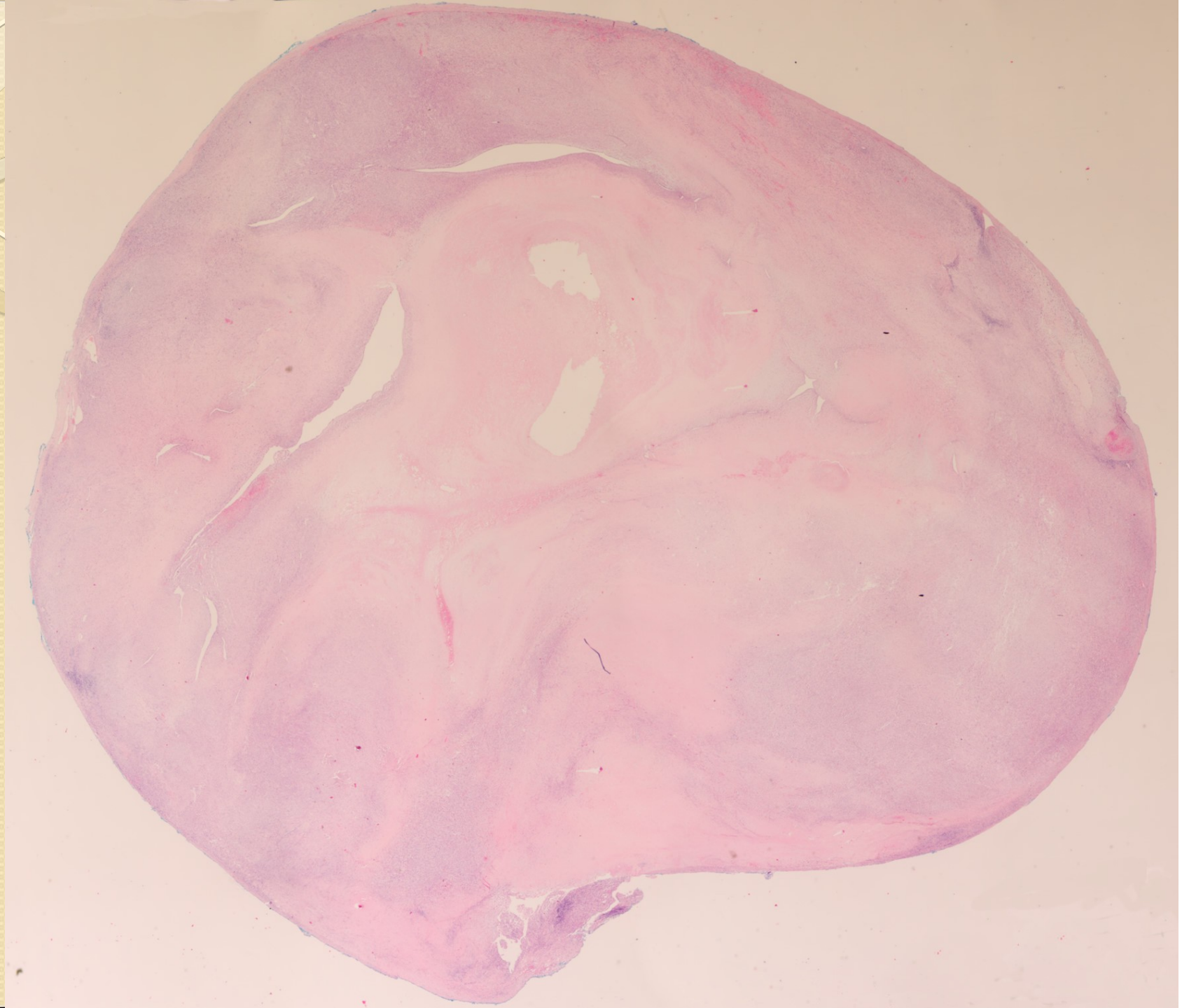
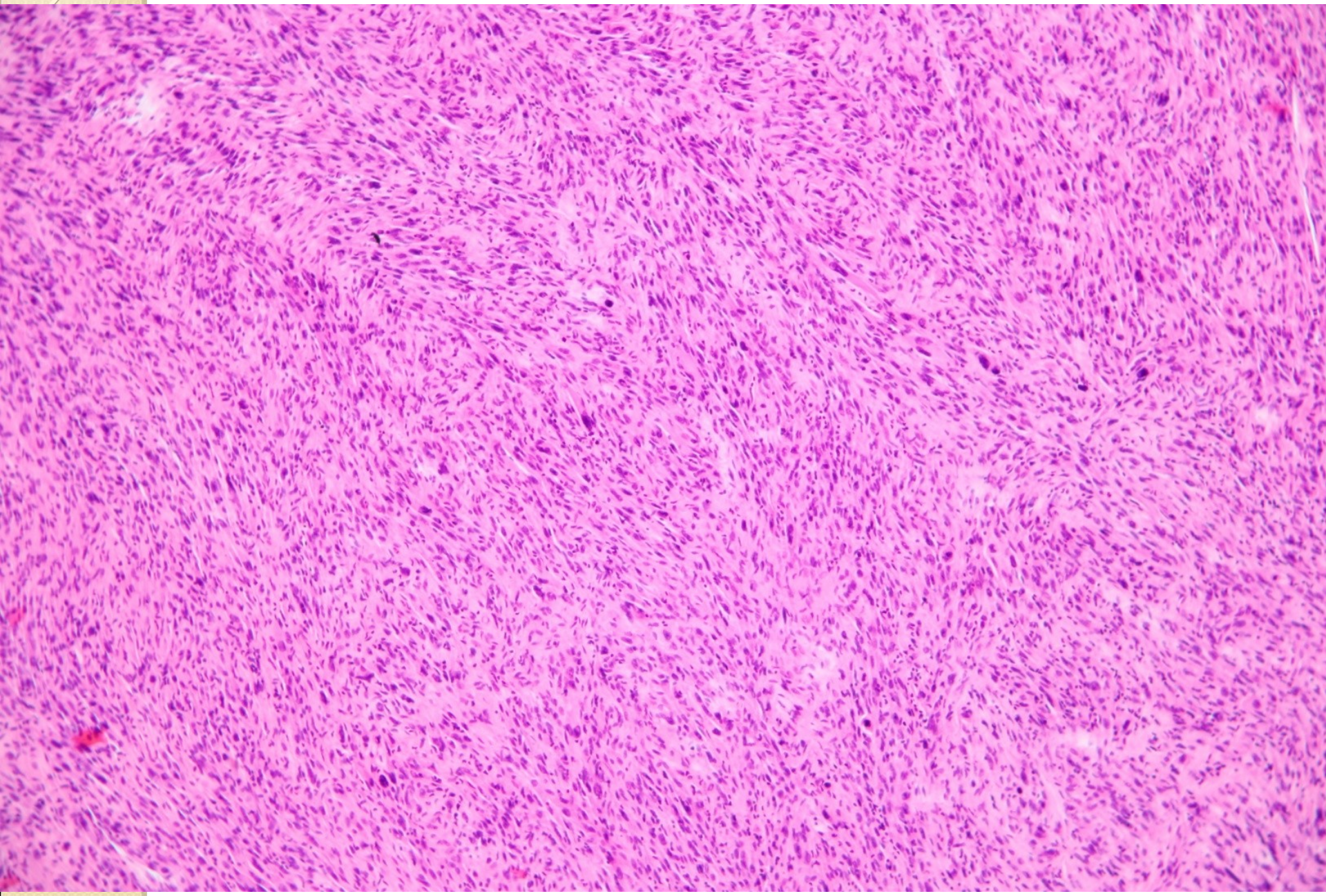
CPC

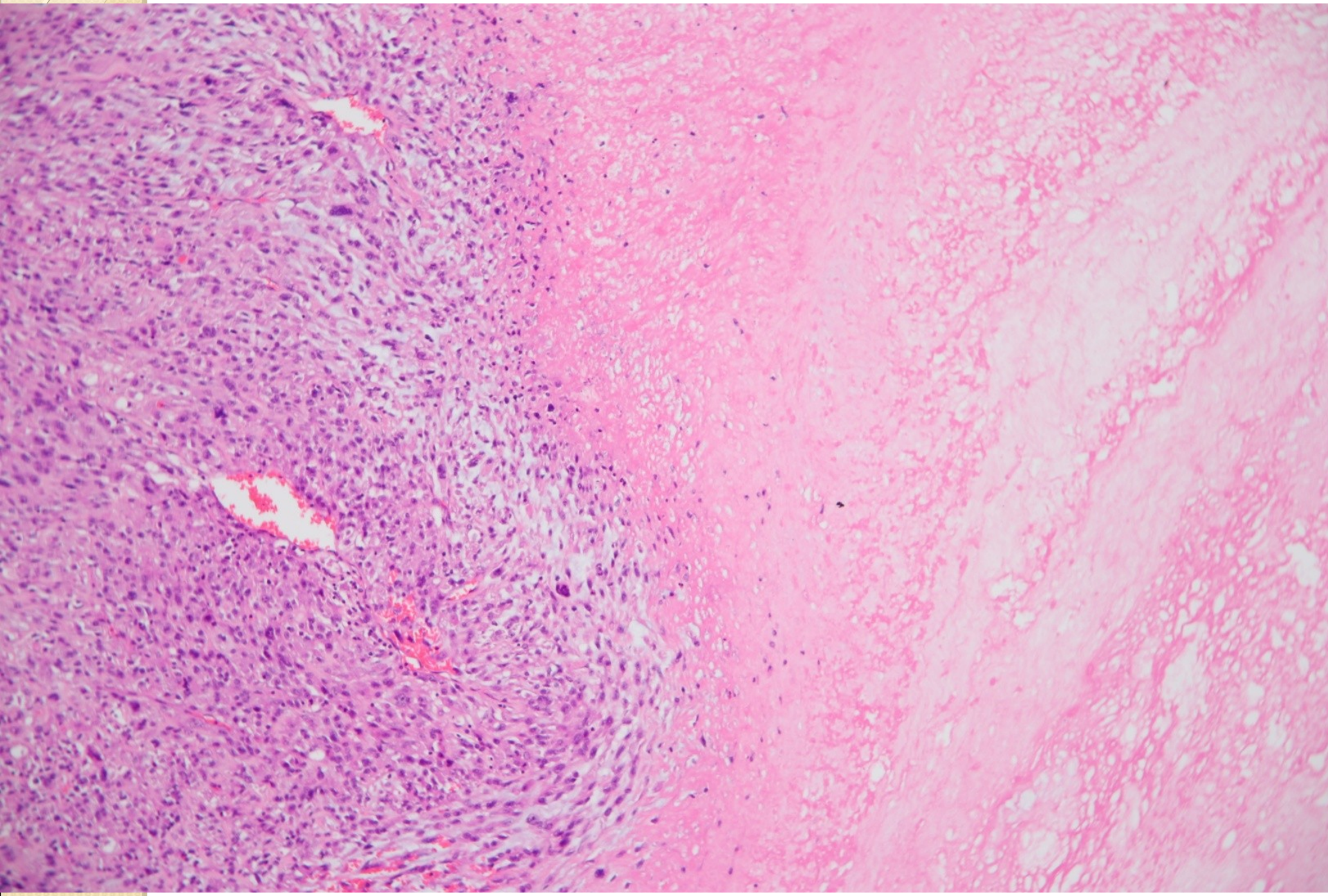
2019/12/12

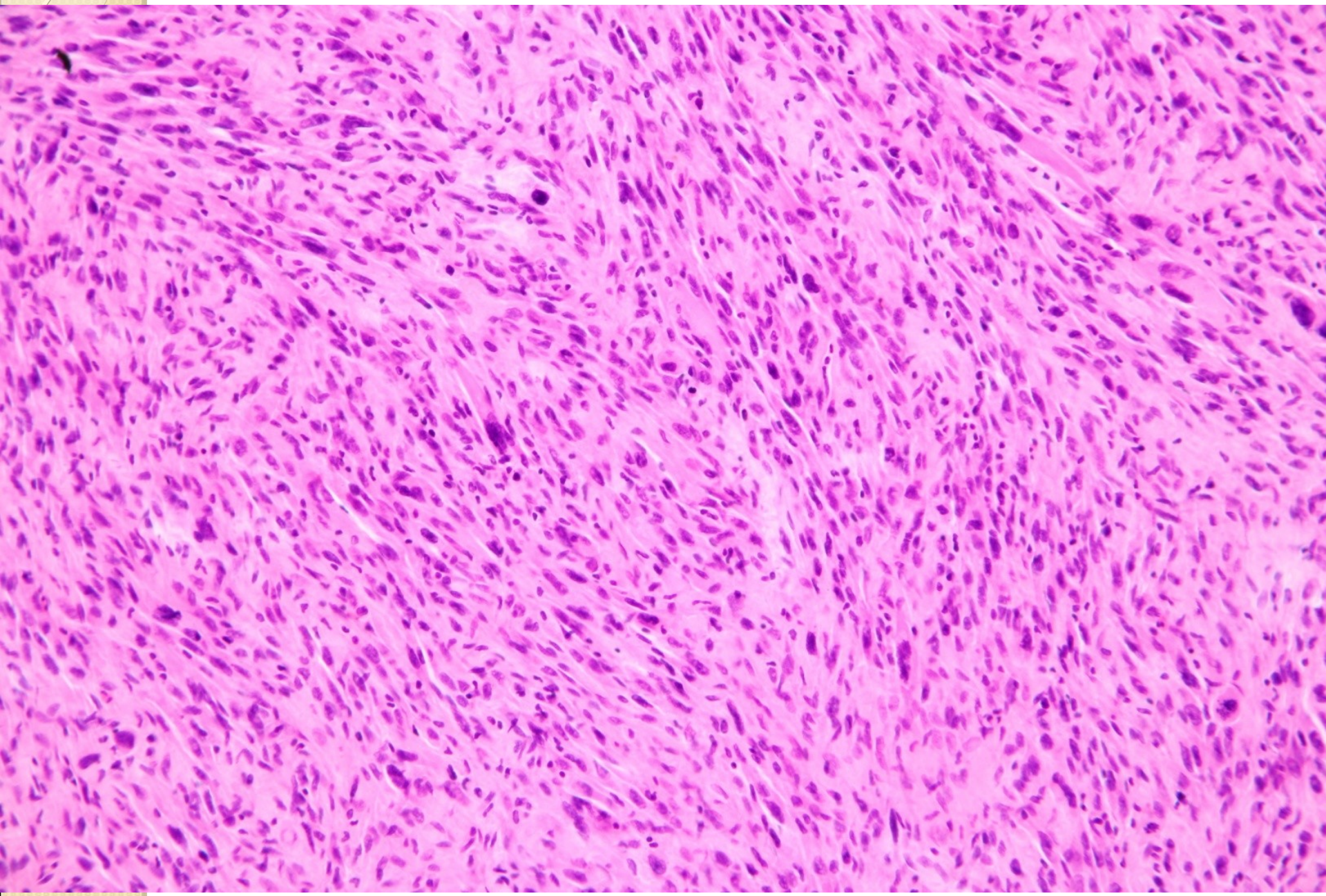
Gross

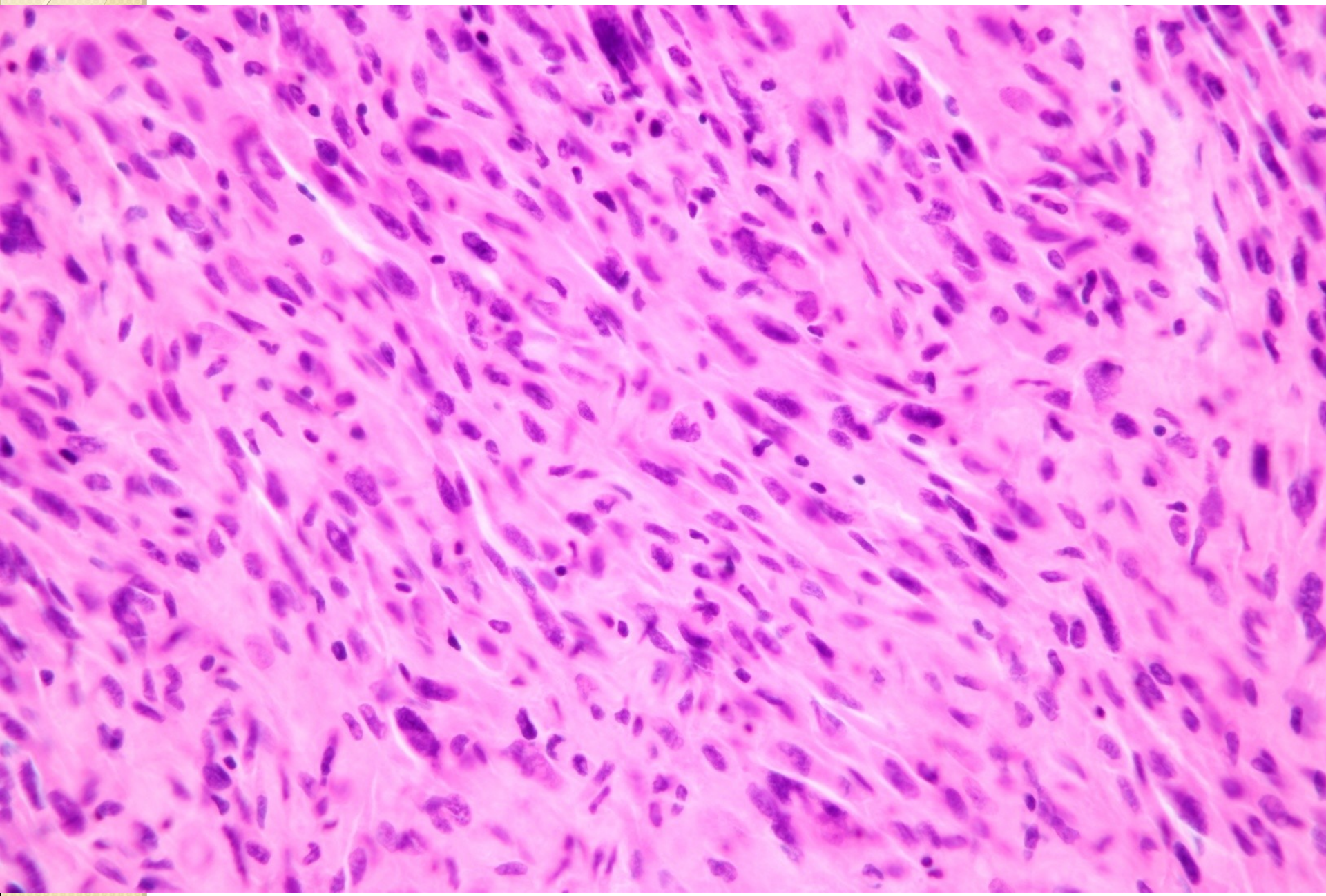
- Three pieces of gray white solid soft tissue tumor
- Labelled pulmonary artery solid tumor
- 10.1x5.2x3.1 cm

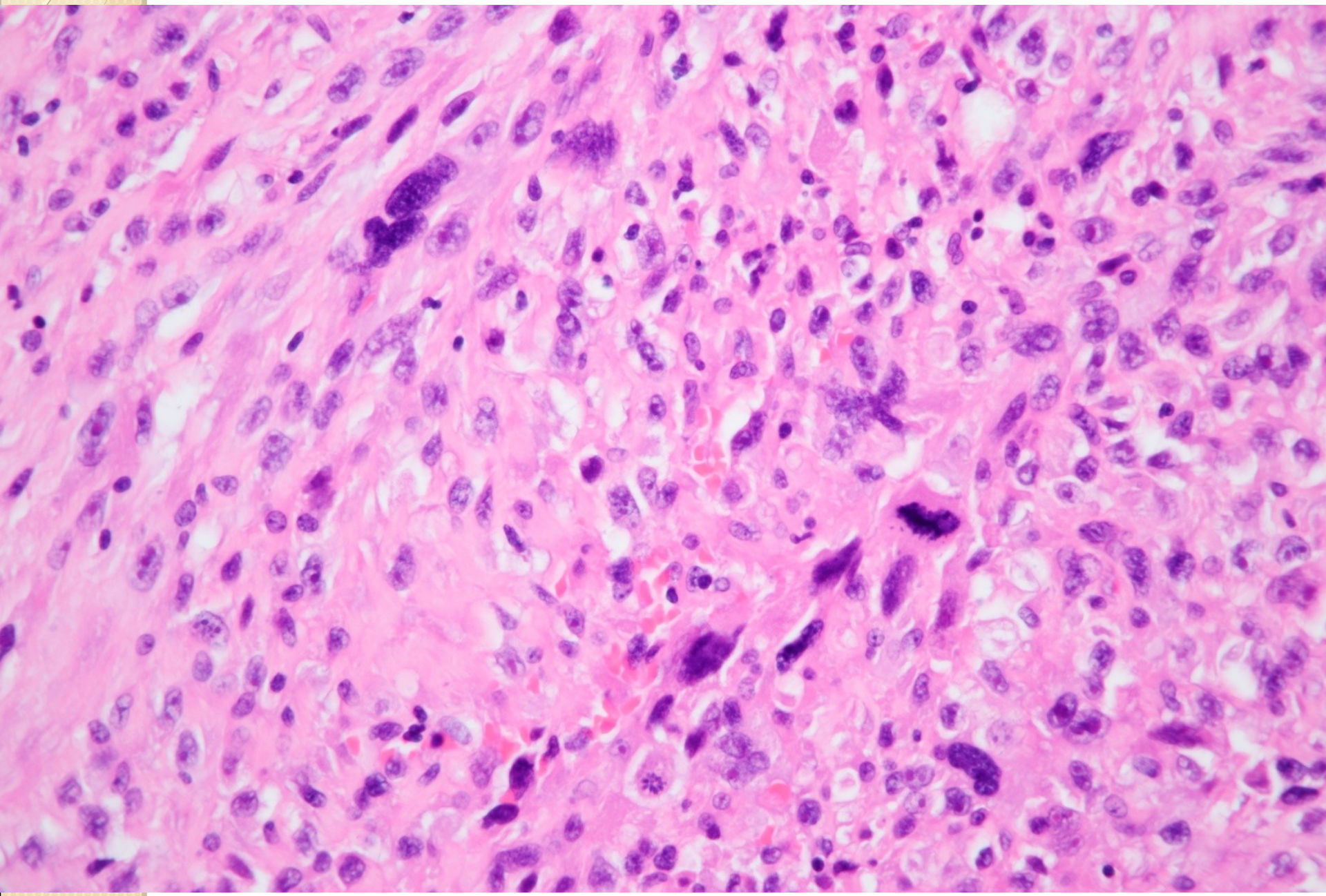






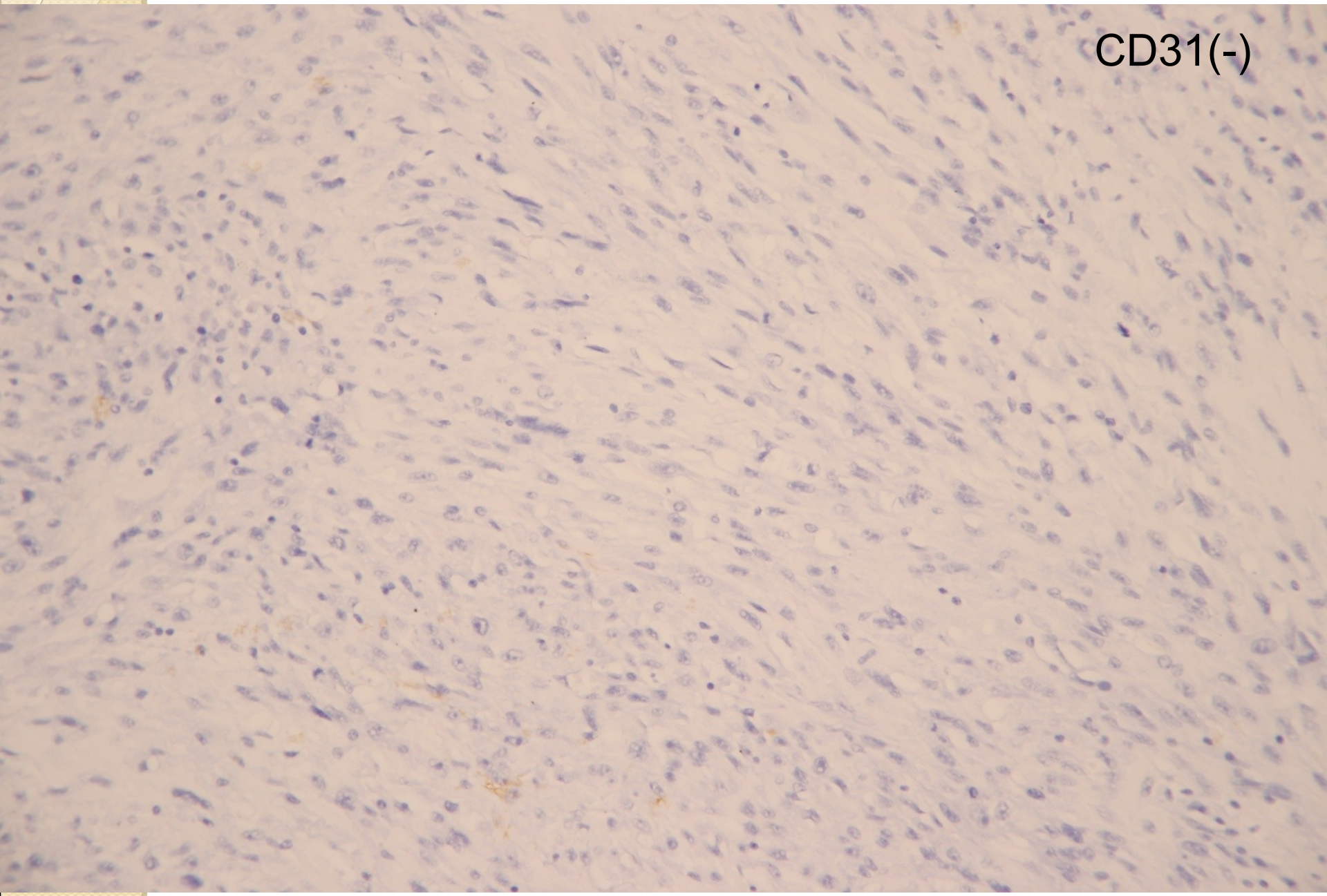


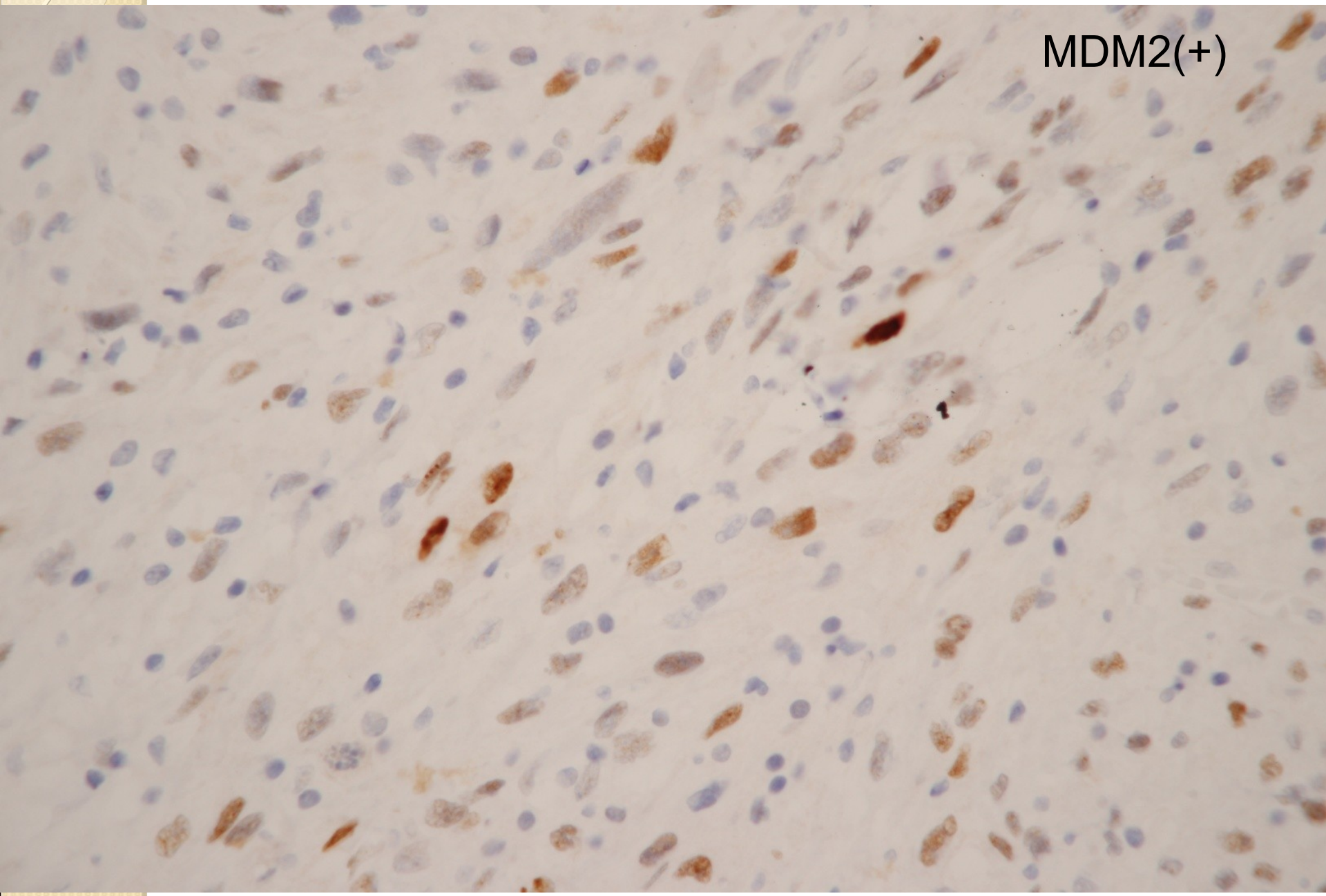




Microscopic Findings

- Sheet-like and focal fascicular growth patterns
- Epithelioid to spindled neoplastic cells
- Hyperchromatic pleomorphic nuclei
- Frequent mitosis, including atypical mitosis


CD31(-)


MDM2(+)

Immunohistochemistry

- MDM2(+)
- CD31(-)
- SMA(-)

Pathological Diagnosis

Intimal sarcoma

Definition

- Malignant mesenchymal tumors
- Large blood vessels (systemic and pulmonary circulation)
- Predominantly intraluminal growth

Epidemiology

- Very rare tumors
- Pulmonary tumors twice as common as tumors of aortic origin
- Pulmonary tumors are slightly more common in females (sex ratio, 1.3)
- Aortic tumors occur equally frequently in males and females
- Mean age:
  - 48 years for pulmonary tumors
  - 62 years for aortic tumors

Sites of involvement

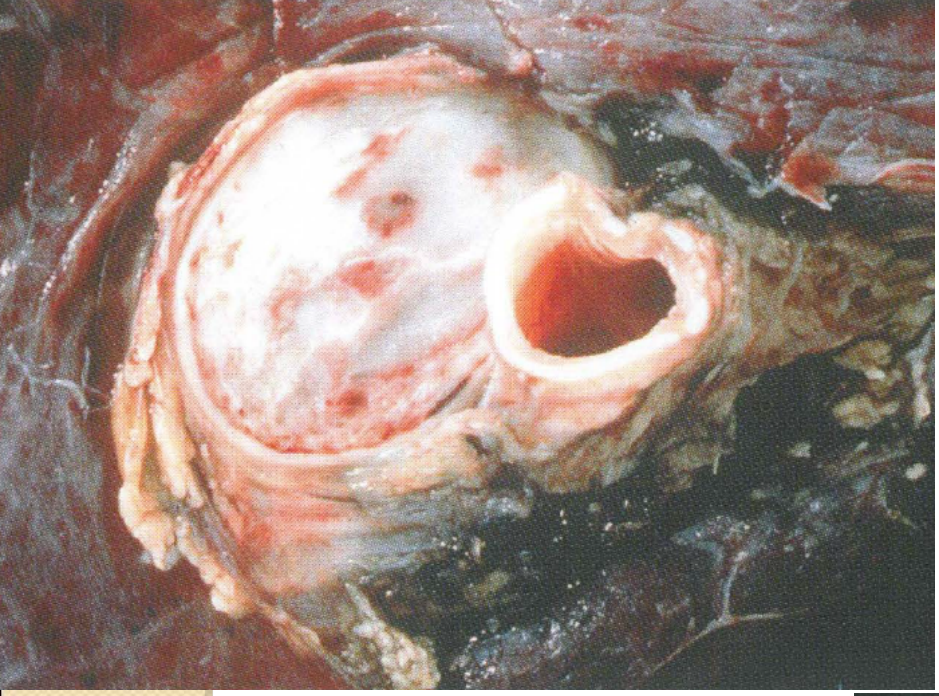
- Pulmonary: pulmonary trunk (80%), right or left main pulmonary arteries (50-70%), or both (40%)
  - ✓Some involve the pulmonary valve or extend into the right ventricular outflow tract
  - ✓Direct infiltration or lung metastases (40%)
  - ✓Extrathoracic spread: 20% of cases (lungs, kidneys, lymph nodes, brain and skin)
- Aorta: mostly in abdominal aorta (between the celiac artery and the iliac bifurcation), 30% in the descending thoracic aorta

Clinical features

- Nonspecific and related to tumour emboli
- Pulmonary: recurrent pulmonary embolic disease
- Aorta: consequences of emboli (claudication, absent pulses, back pain, abdominal angina, malignant hypertension, rupture of aneurysm formed by the tumor)

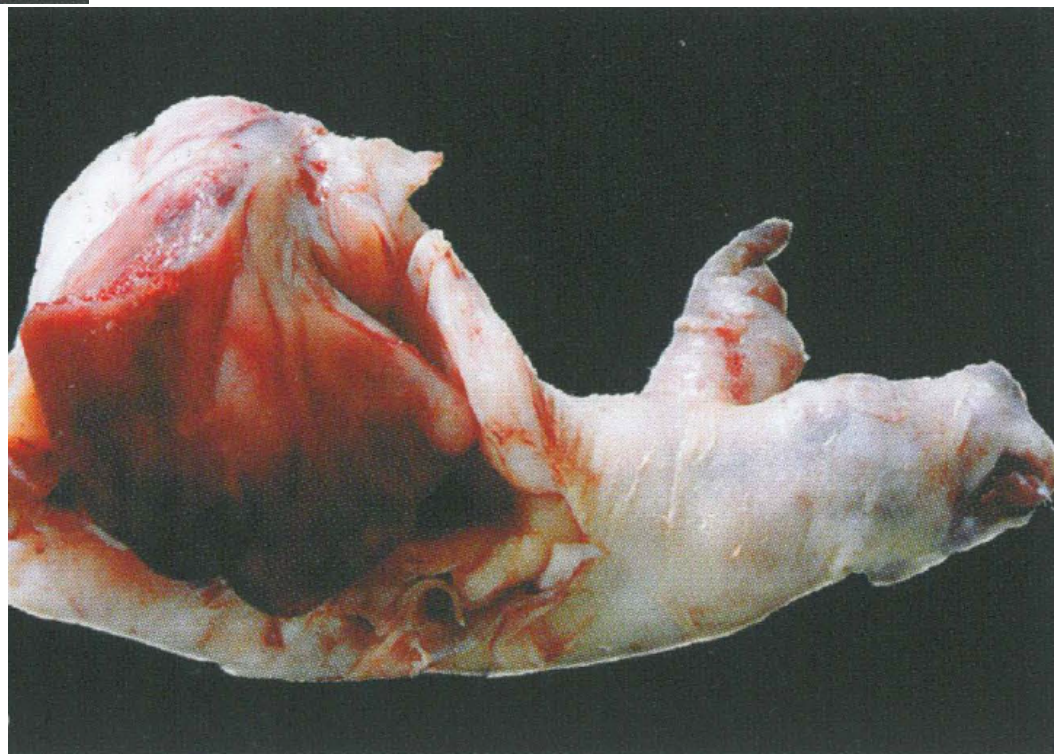
Macroscopy

- Mostly intravascular polypoid masses attached to the vessel wall
- Harder, bony areas corresponding to osteosarcomatous differentiation
- Some of the aortic tumors may cause thinning and aneurysmal dilatation of the vessel wall



Pulmonary artery occluded by tumor

Endarterectomy specimen



Histopathology

- Poorly differentiated mesenchymal malignant tumors
- Atypical spindle cells
- Varying degrees of mitotic activity, necrosis and nuclear polymorphism
- Rare cases may contain areas of rhabdomyo- or osteosarcomatous differentiation

Immunophenotype

- Nuclear expression of MDM2 in 70% of cases
- Endothelial markers are negative
- Variable positivity for SMA

Genetics

- PDGFRA (81 %)
- EGFR (76%)
- MDM2 (65%)

Prognosis and Treatment

- The prognosis is poor
- Mean survival
  - Aortic sarcomas: 5-9 months
  - Pulmonary sarcomas: 13-18 months
- Treatment: Surgery, radiotherapy, chemotherapy

Thank you for your attention