PIP-C 2003 Cases

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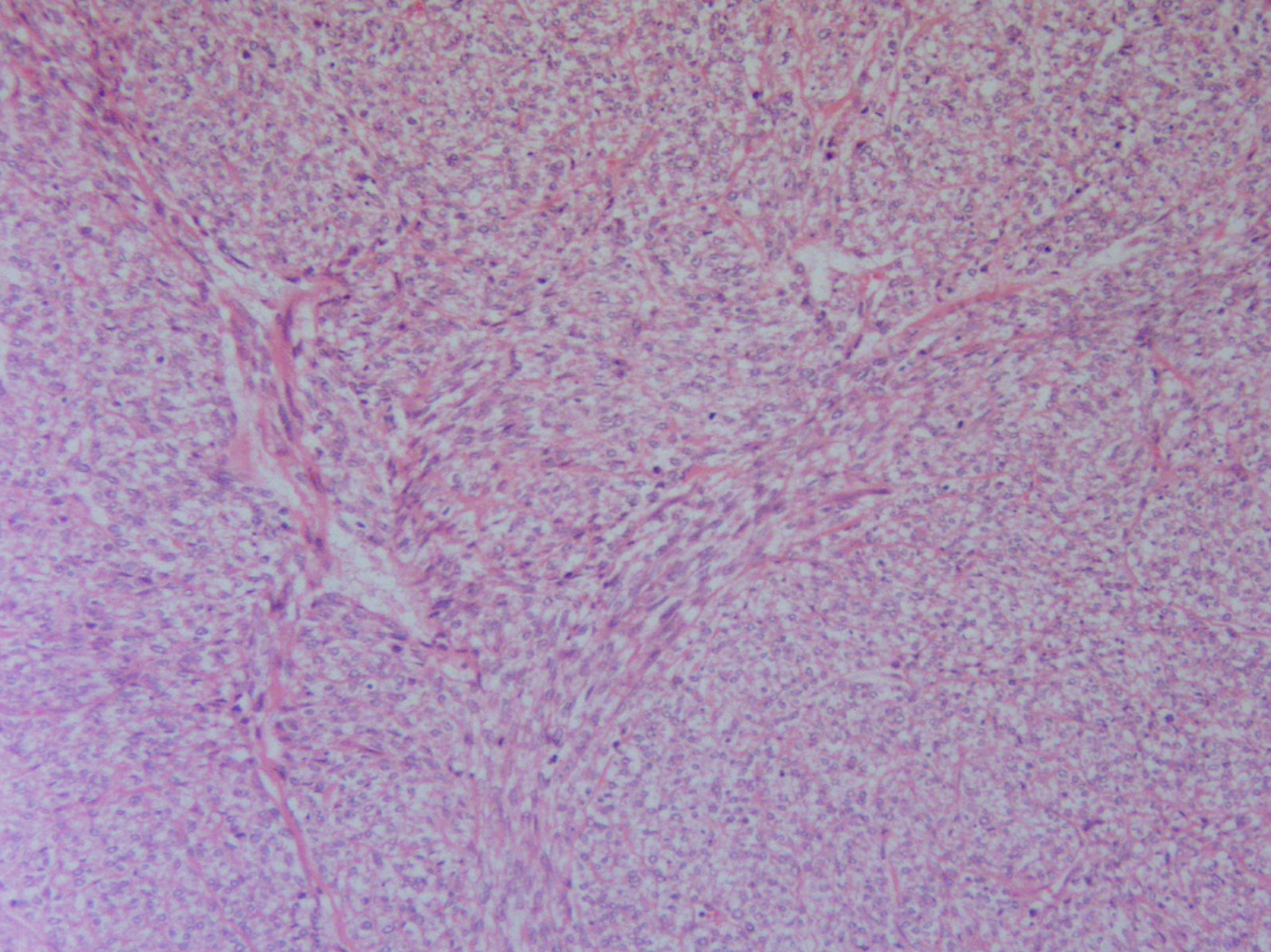
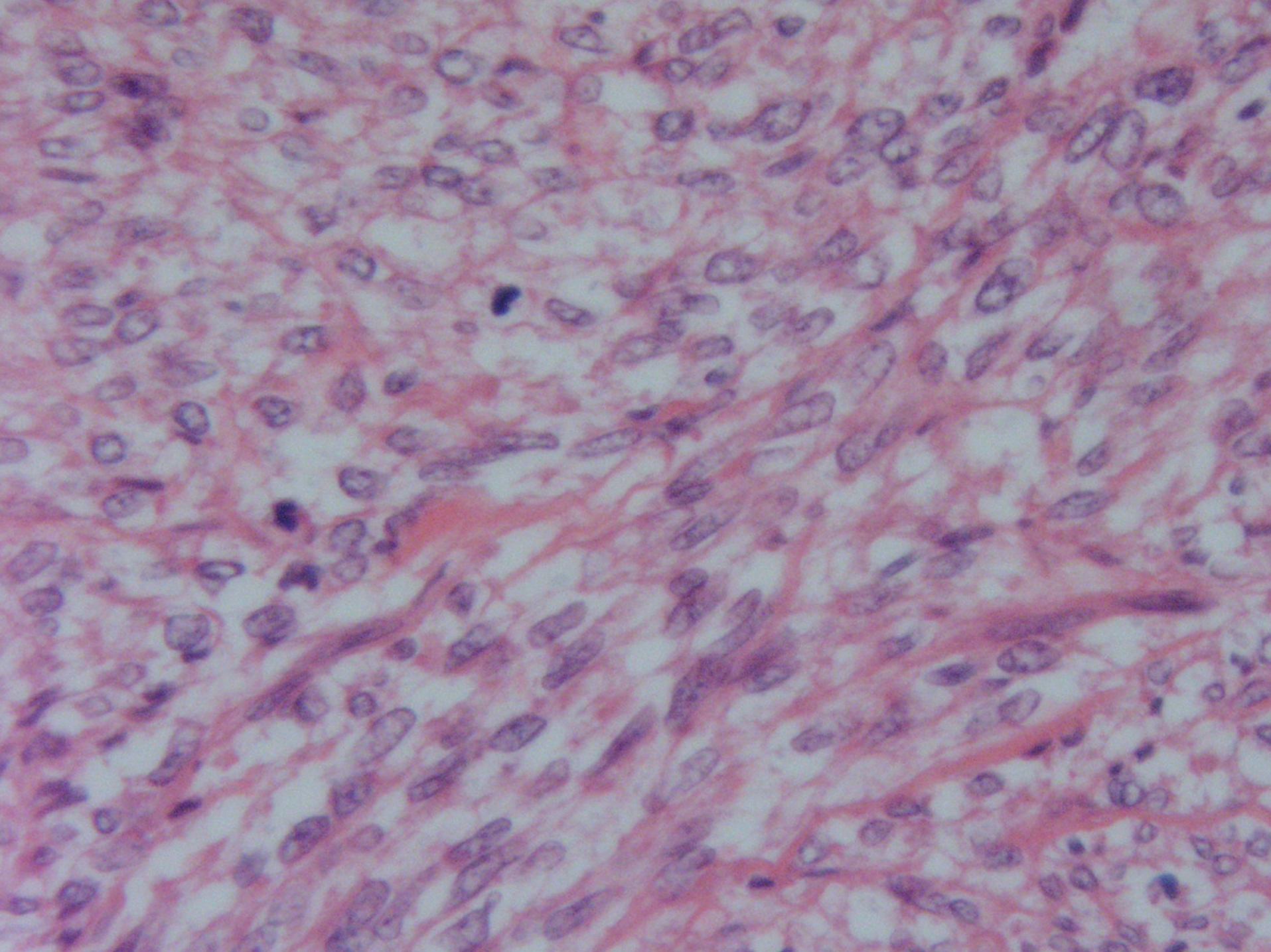
APMG

Case 21

- 39F
- Lower abdominal pain, LE swelling, palpable abdominal mass
- CT with tumor occluding IVC





Leiomyosarcoma of the Inferior Vena Cava

Clinical

- Rare
- 15-84 years (average 50 yrs)
- 80-90% women
- Symptoms vary with location
  - LE edema
  - Budd-Chiari syndrome
- Some retroperitoneal leiomyosarcomas may represent primary caval tumors
  - Only 25% are predominately intralumenal

Histopathology

- MF increased
- Pleomorphism, hemorrhage, necrosis not as pronounced as similar retroperitoneal tumors
- Positive for SMA, h-caldesmon, desmin

Prognosis

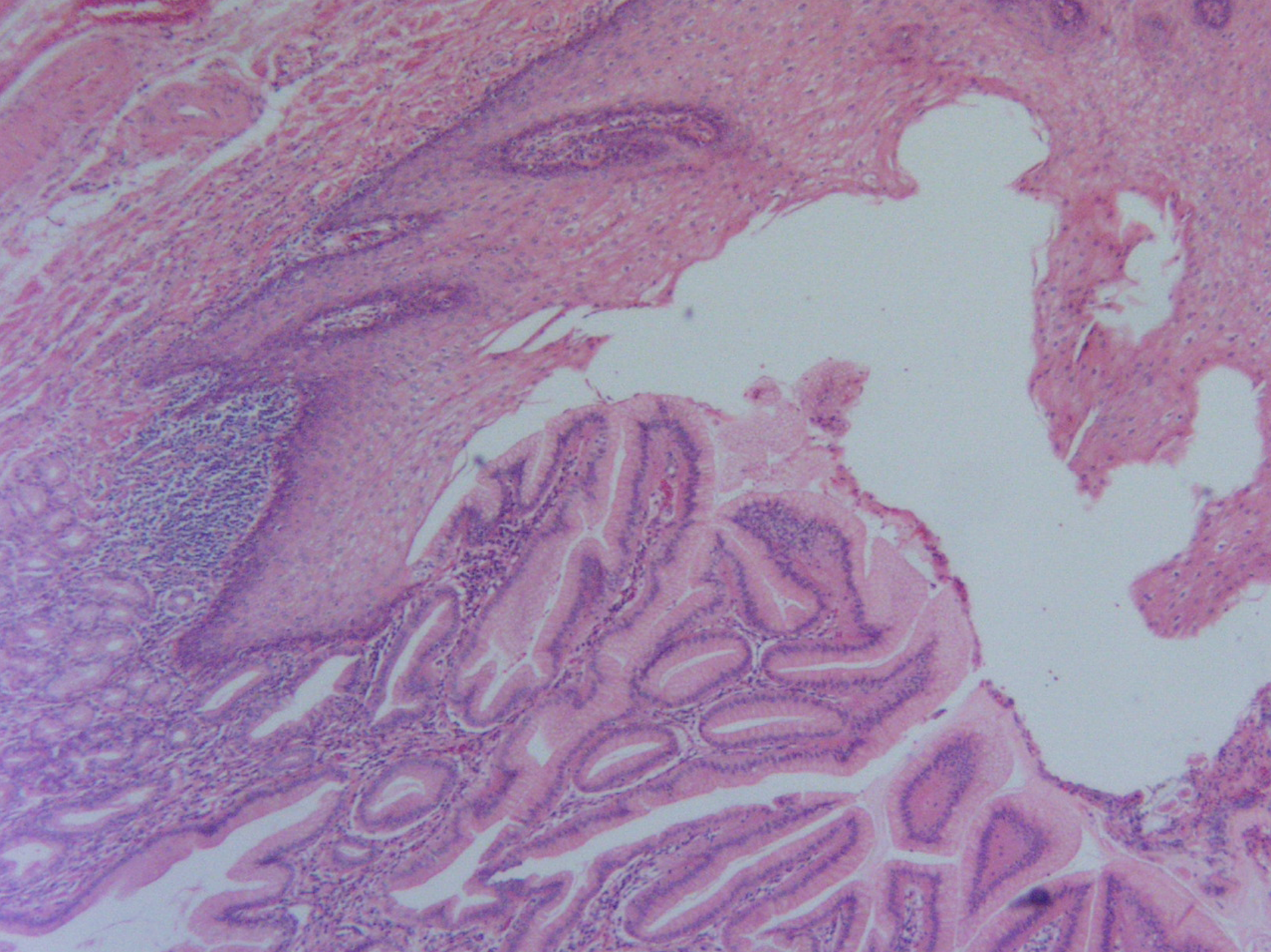
- 5YRS of 35%
- Mets to lungs, kidneys, pleura/chest wall, liver, and bone

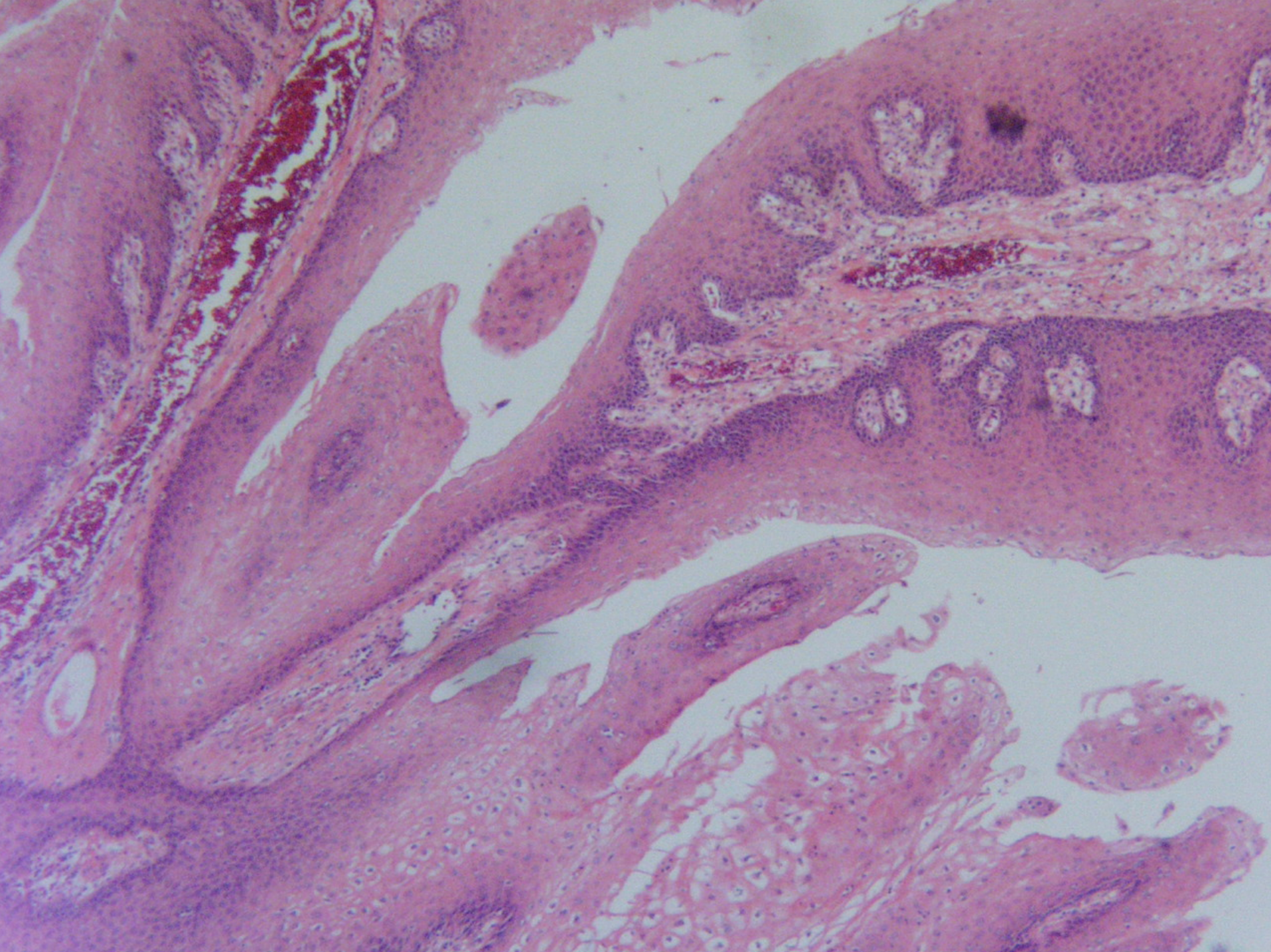
DDX

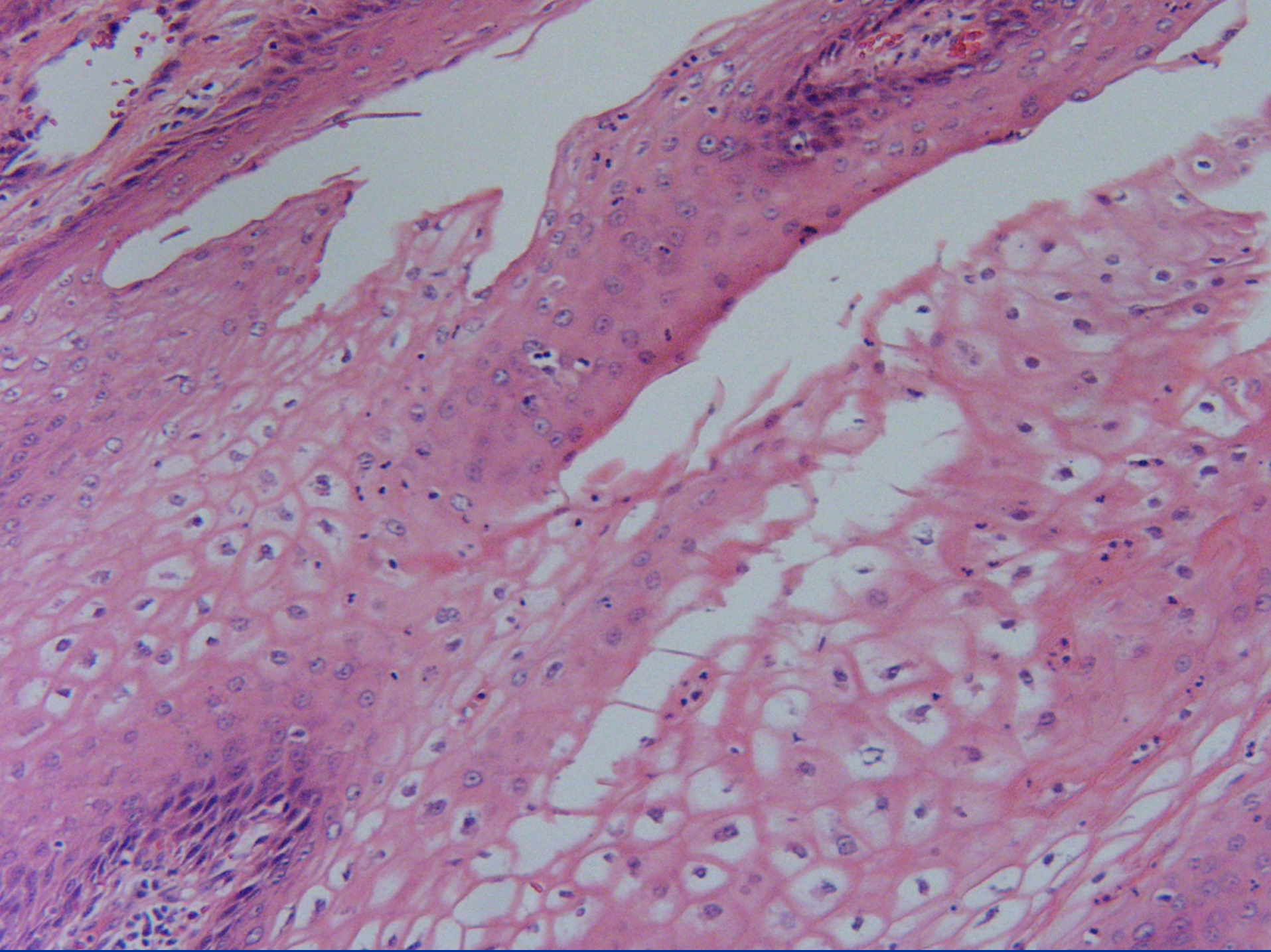
- Angiosarcoma
- Endometrial stromal sarcoma
- Intravenous leiomyomatosis
  - >5MF/10 hpf malignant
  - 1-4MF/10 hpf potentially malignant
- Renal cell carcinoma, sarcomatoid type

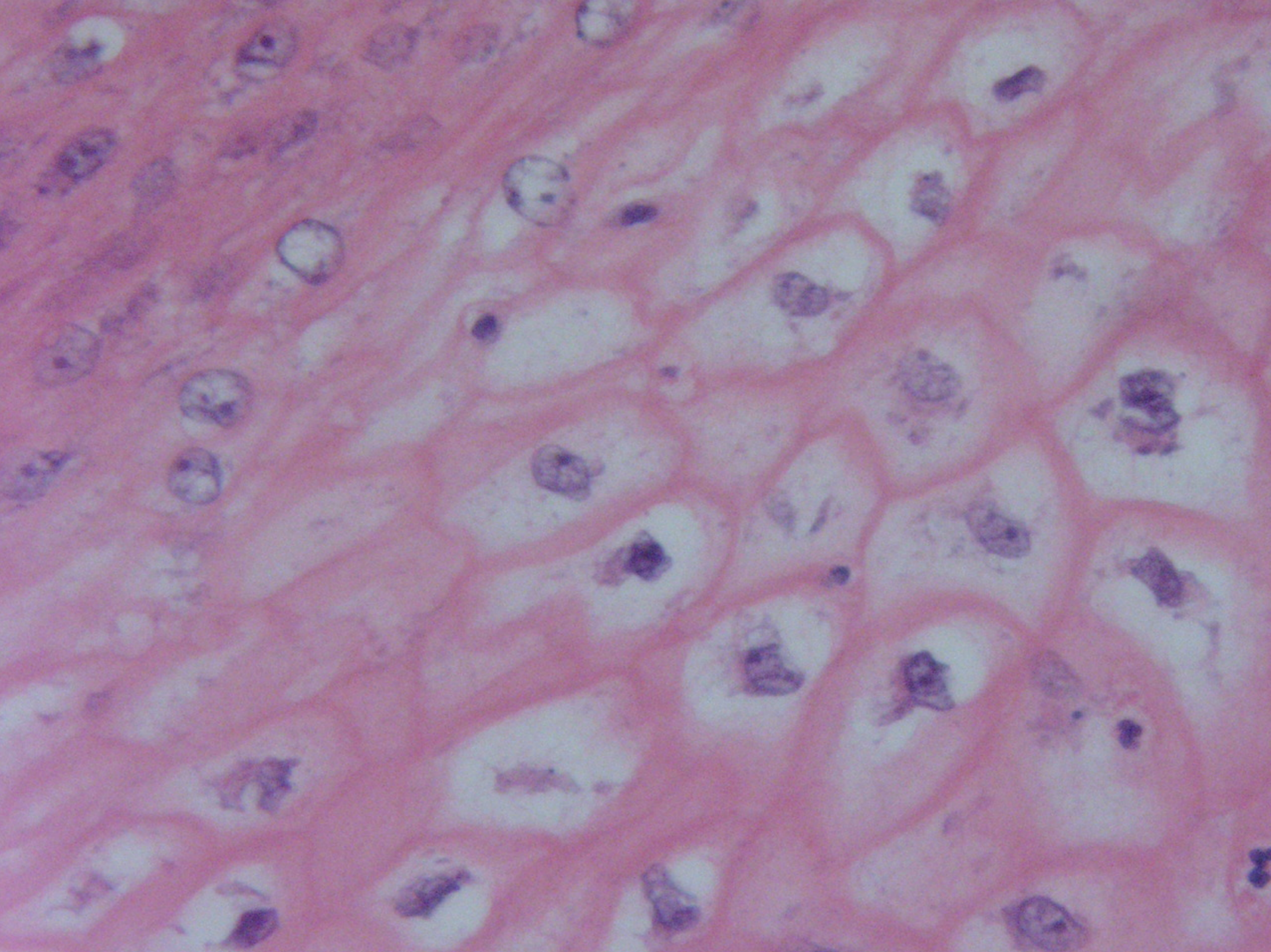
Case 22

- 61F
- Dysphagia, heartburn, hematemesis
- Exophytic lesion at distal esophagus









Squamous Papilloma of the Esophagus

Clinical

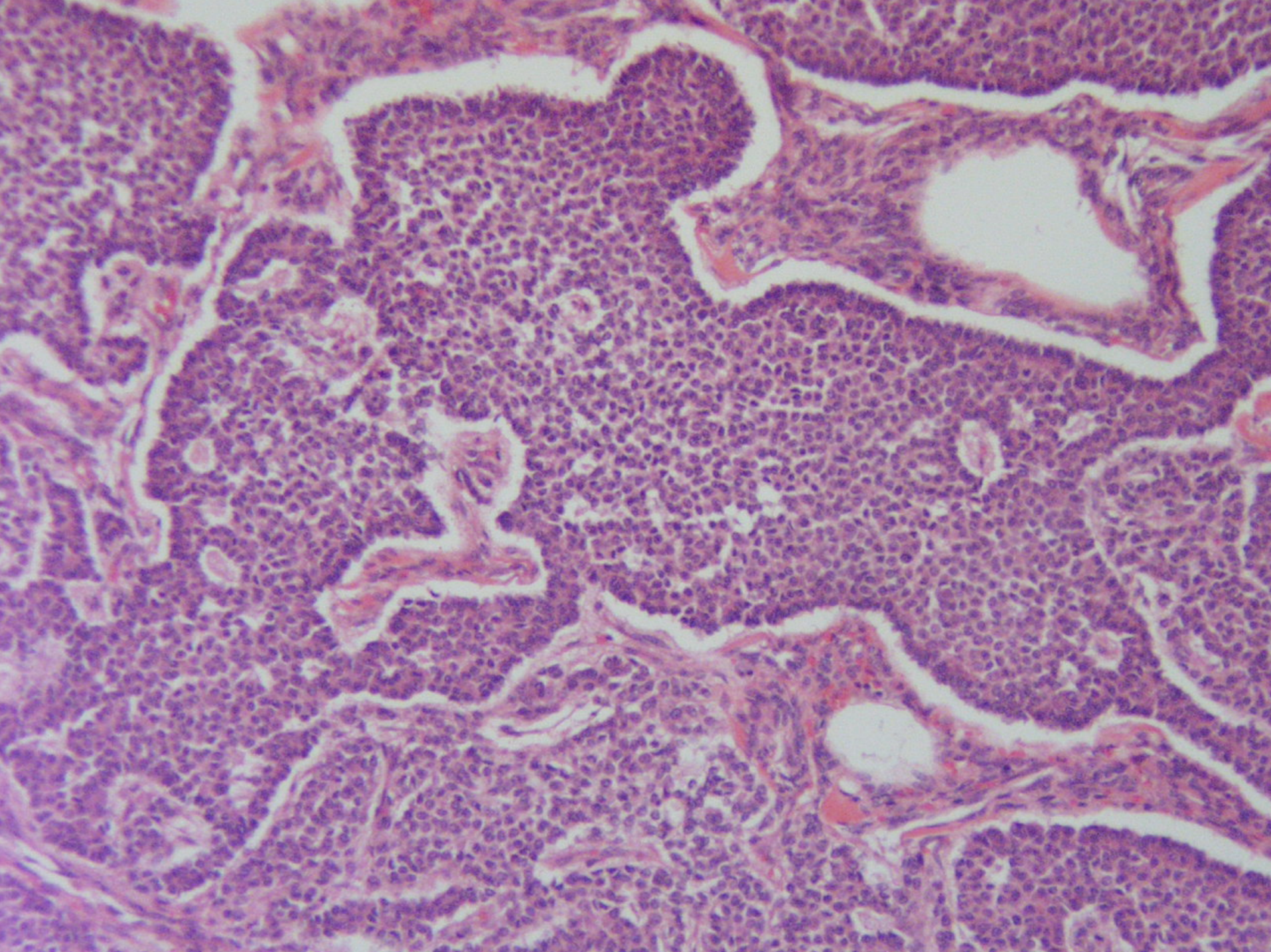
- <1% of esophageal tumors
- Usually solitary
- Middle aged men
  - Children with laryngeal papillomatosis (HPV 6/11)
- Dysphagia, heartburn, GERD
- Upper 5%
  Mid 24%
  Lower 71%
- Exaggerated regenerative response to GERD, trauma, or esophagitis
  - ?HPV

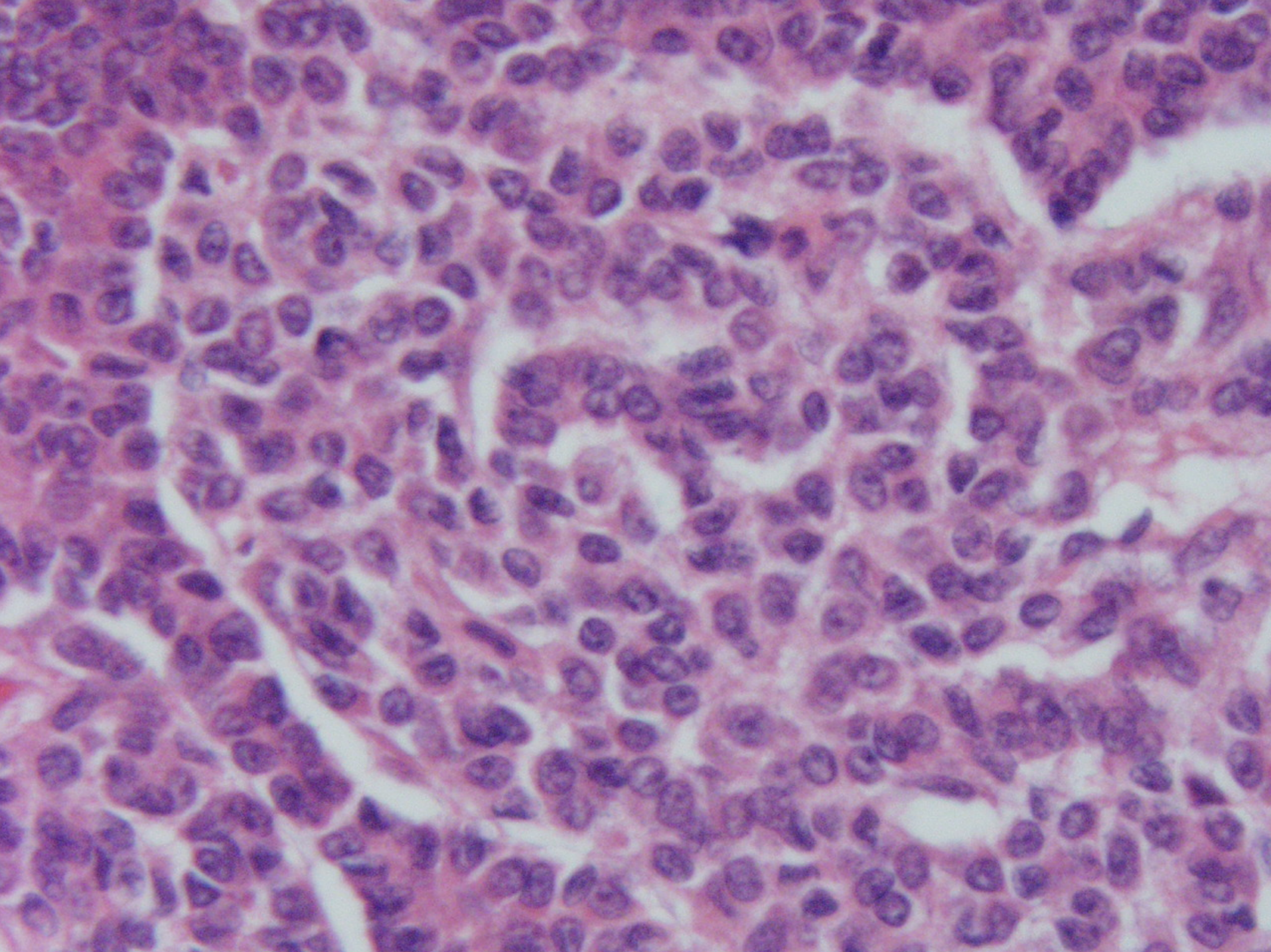
DDX

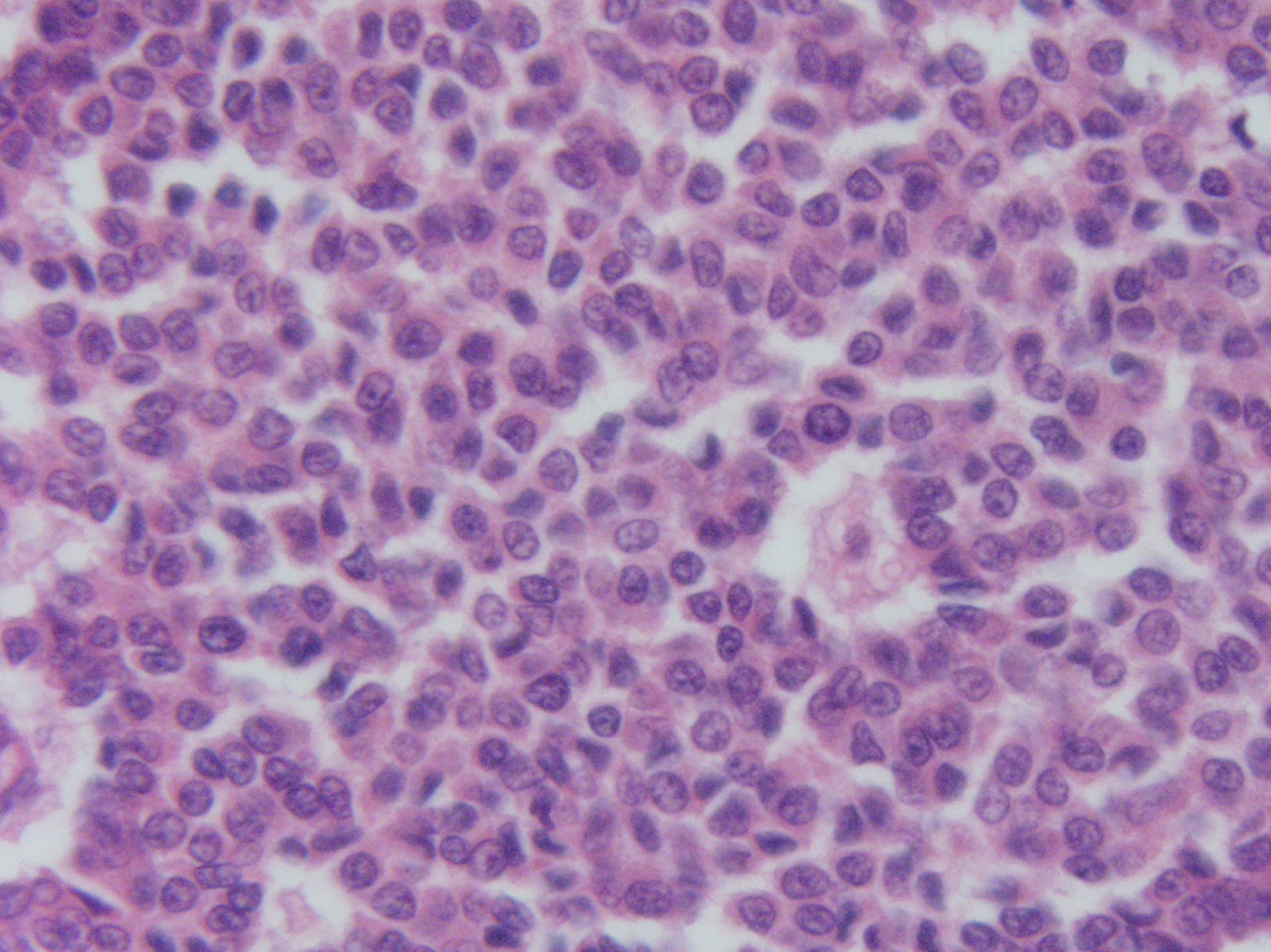
- Giant fibrovascular polyp
  - Upper 1/3, pedunculated, men
- Inflammatory fibroid polyp
  - Pedunculated with infiltrative borders
- Pseudoepitheliomatous hyperplasia
  - Often with granular cell tumors
- Squamous cell CA
  - 25% polypoid

Case 23

- 53F
- Left adnexal mass, 1310 gm, 19x15x9 cm ovary
- Cut surface with pale yellow to gray, solid with focal myxoid areas







Adult Granulosa Cell Tumor of the Ovary

Clinical

- 1-2% ovarian neoplasms
- Adult-95% Juvenile 5%
- Perimenopausal (Mean 45-55 yrs)
- Abdominal discomfort and endocrine manifestations
  - AUB due to endometrial hyperplasia
  - Rarely androgen excess
  - Tumor rupture rare with acute abdomen
- Unilateral 95%
- Mean 12 cm, varying solid to cystic

Histopathology

- Round to ovoid angulated nuclei (coffee-bean)
- Call-Exner bodies (microfollicular) in 50%
- IPOX
  - Inhibin ~100%
    SMA~50%
    S100~50%
    Keratin~20-40%

Treatment/Prognosis

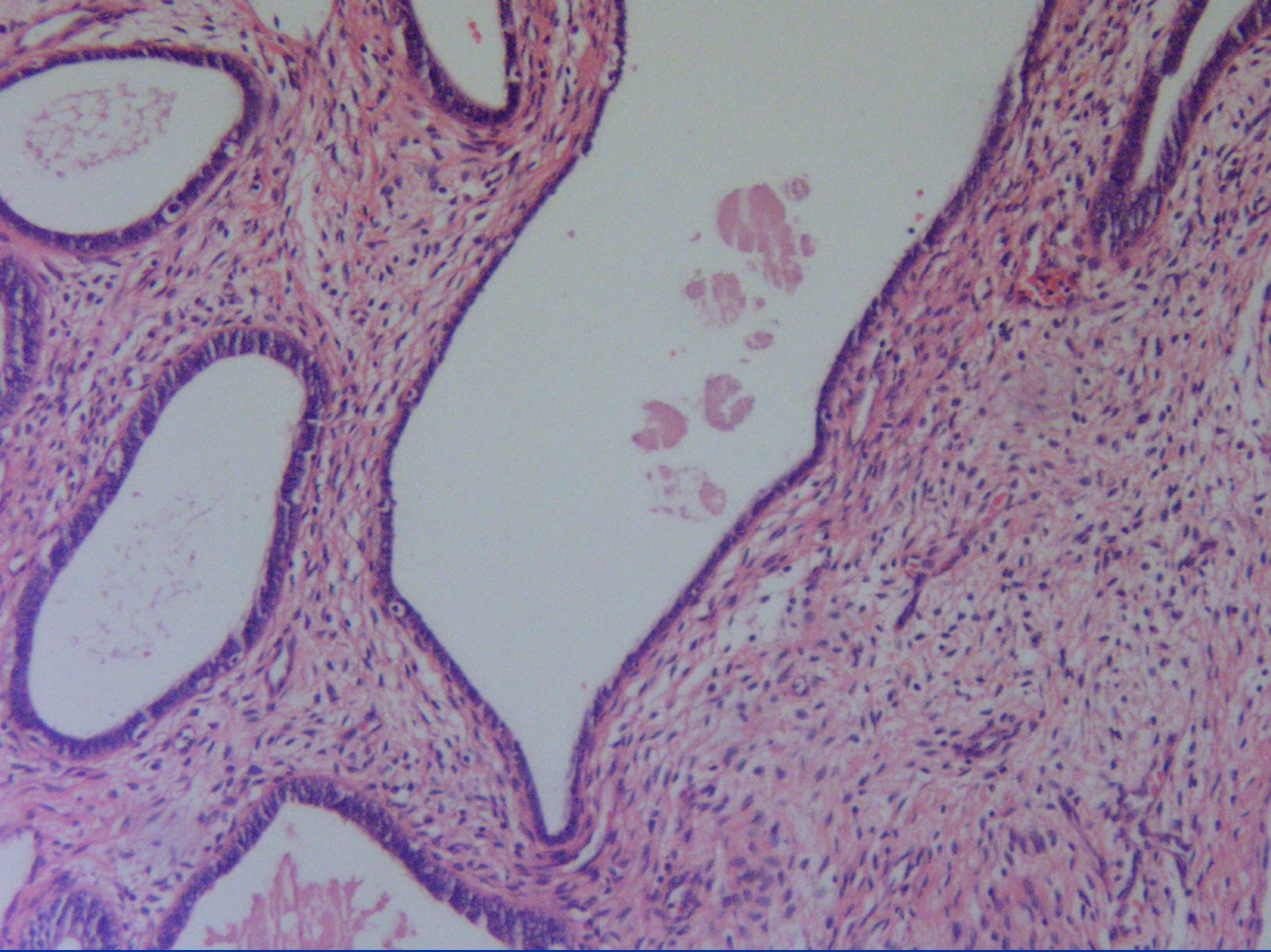
- TAH-BSO, younger consider USO
- 10YRS 60-90%
  20YRS 30-50%
- Recurrences >20 yrs, usually abdomen
- Mets to lungs, brain, bone, liver
- Chemotherapy with 35% response to cis-platinum based regimens with local radiotherapy
- Higher tumor stage, tumor size, and rupture adversely affect prognosis

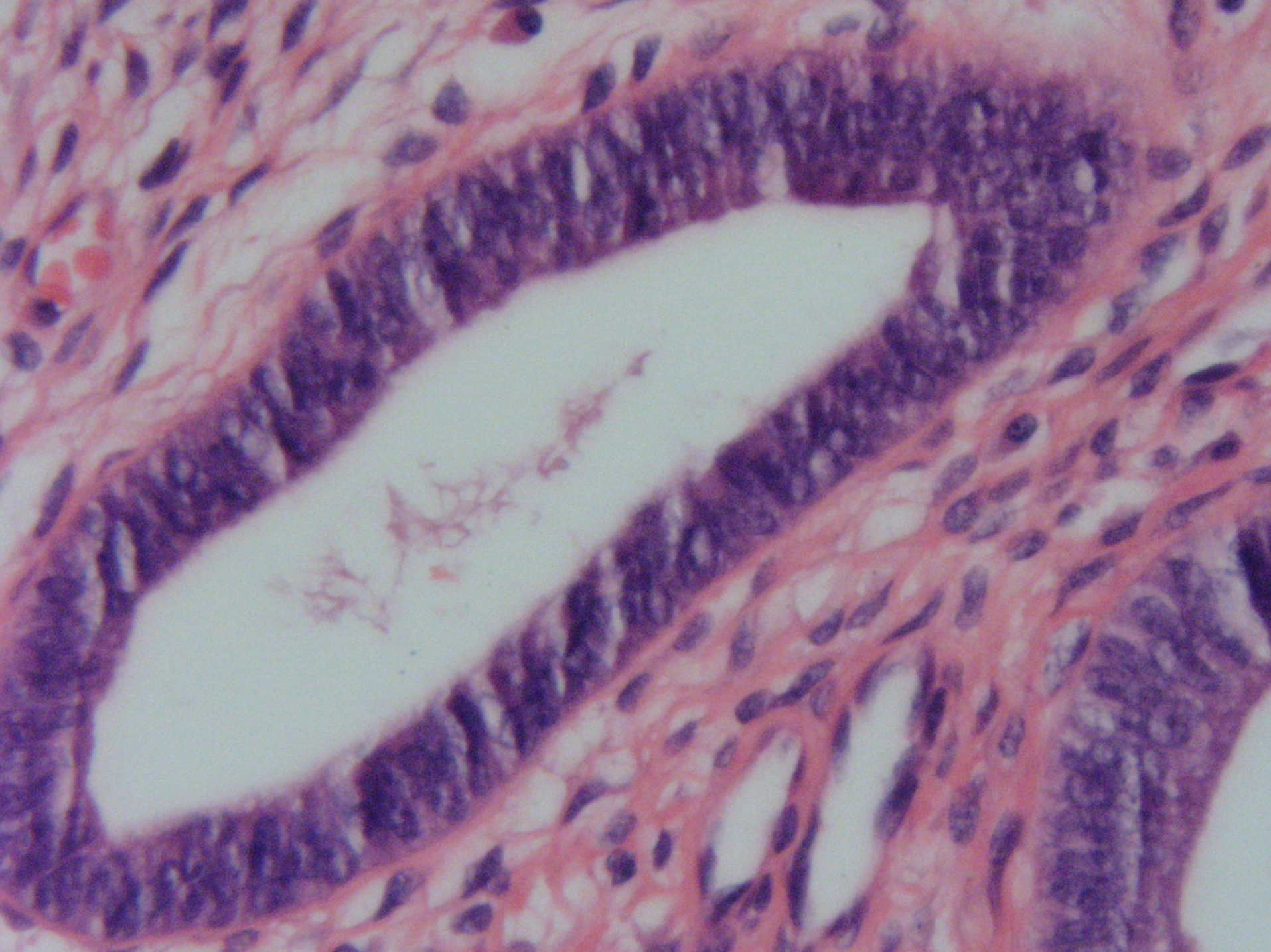
DDX

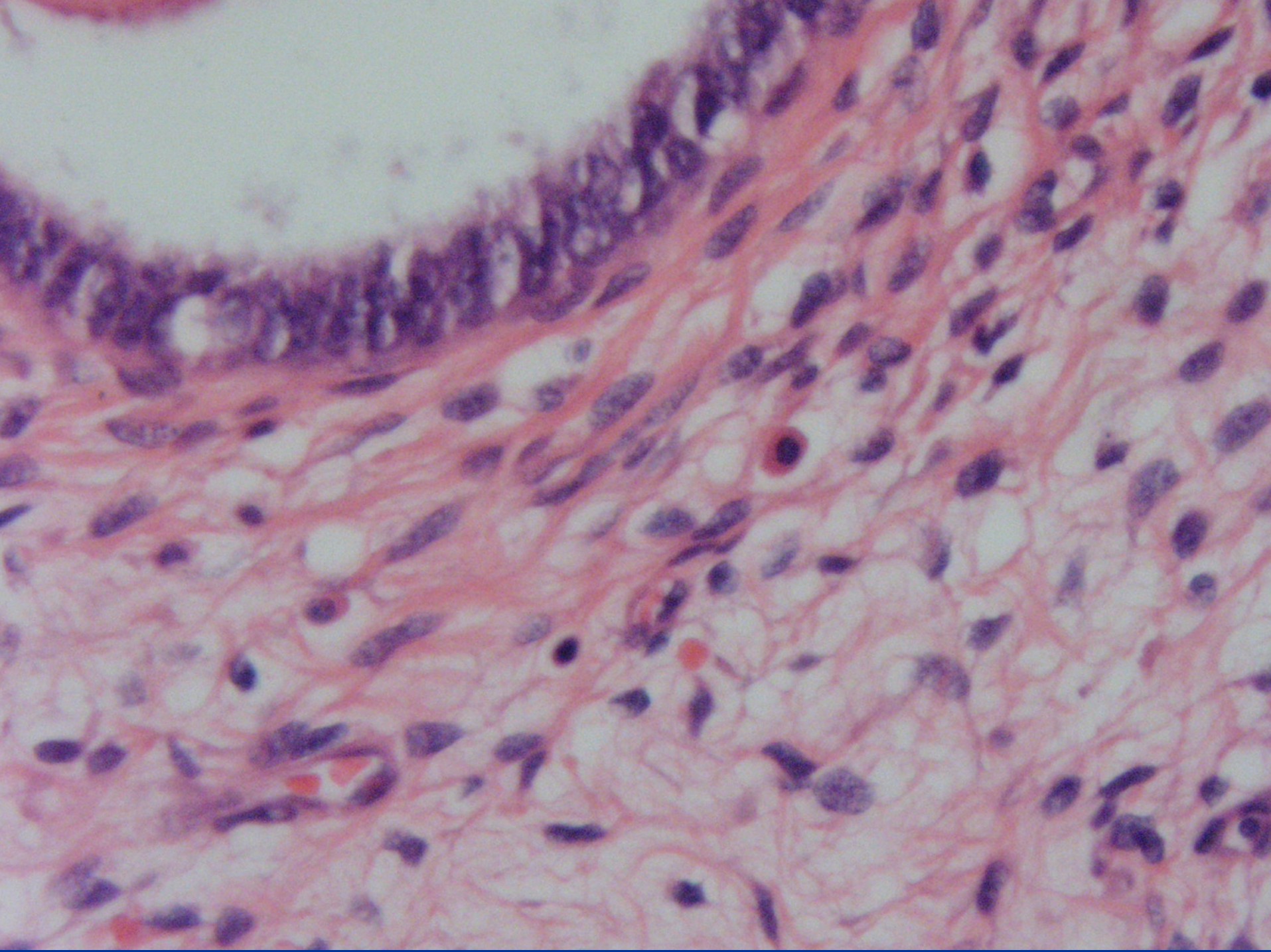
- Carcinoid tumor
- Cellular fibroma
- Endometrioid carcinoma
- Endometrial stromal sarcoma
- Juvenile granulosa cell tumor
  - Larger cell size with variable follicles with mucin
    Numerous MF
- Metastatic breast carcinoma
- Small cell carcinoma
- Thecoma

Case 24

- 81F
- Irregular vaginal bleeding
- Tamoxifen for 4 months after tx of breast CA
- TAH/BSO
- 230 gm uterus, 10x7x5 cm uterus with 8cm polypoid lesion filling cavity







Endometrial Polyp

Clinical

- Peri-menopausal with irregular vaginal bleeding
- Uterus usually not enlarged
- Occasionally may prolapse through cervical os

Histopathology

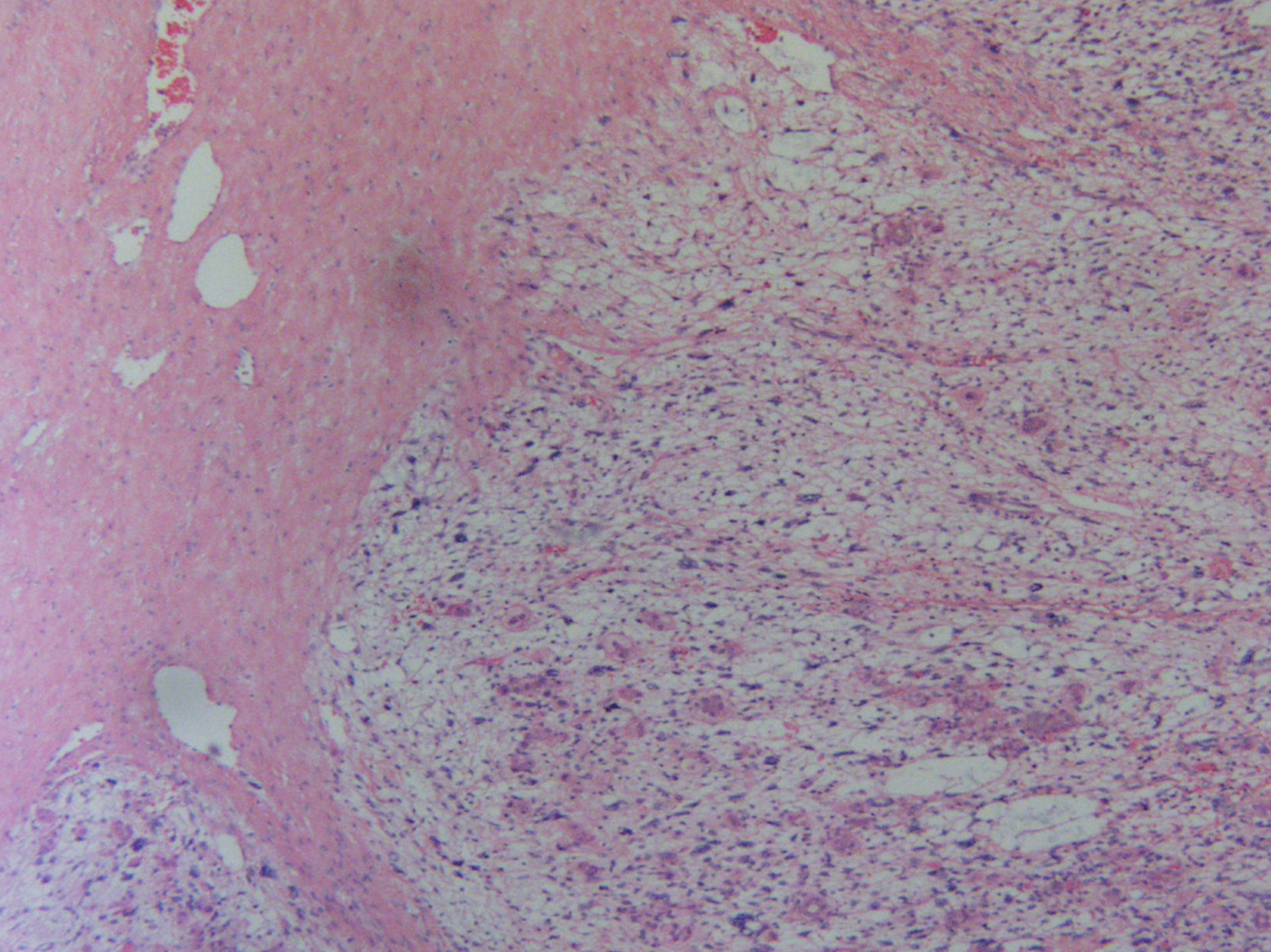
- Tamoxifen-induced polyps
  - Larger
  - Glands parallel to long axis of polyp (similar to normal endometrium), but may be staghorn, irregular, or small
  - More frequent stromal fibrosis, periglandular stromal condensation (cambium layer), and mucinous metplasia

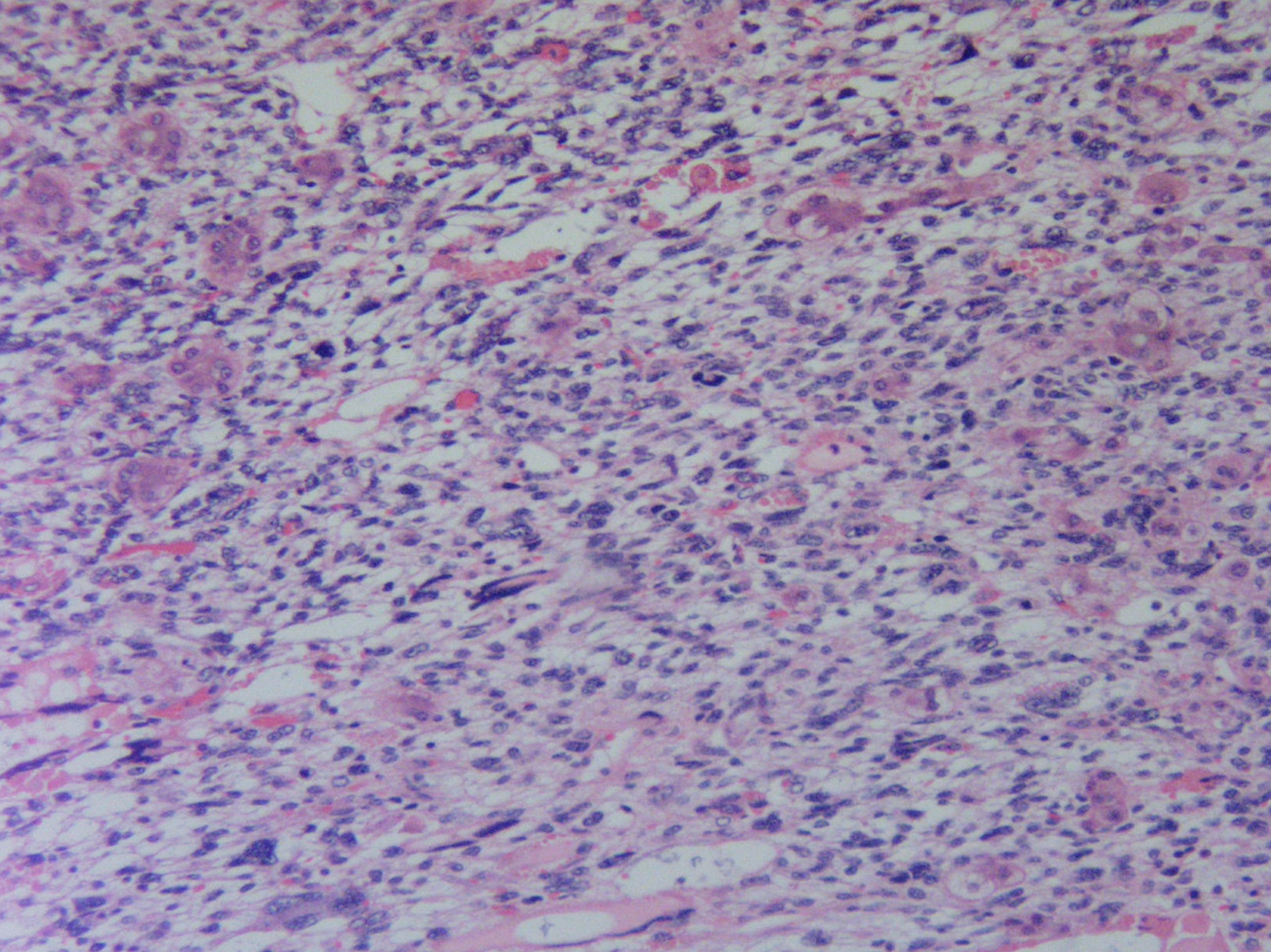
DDX

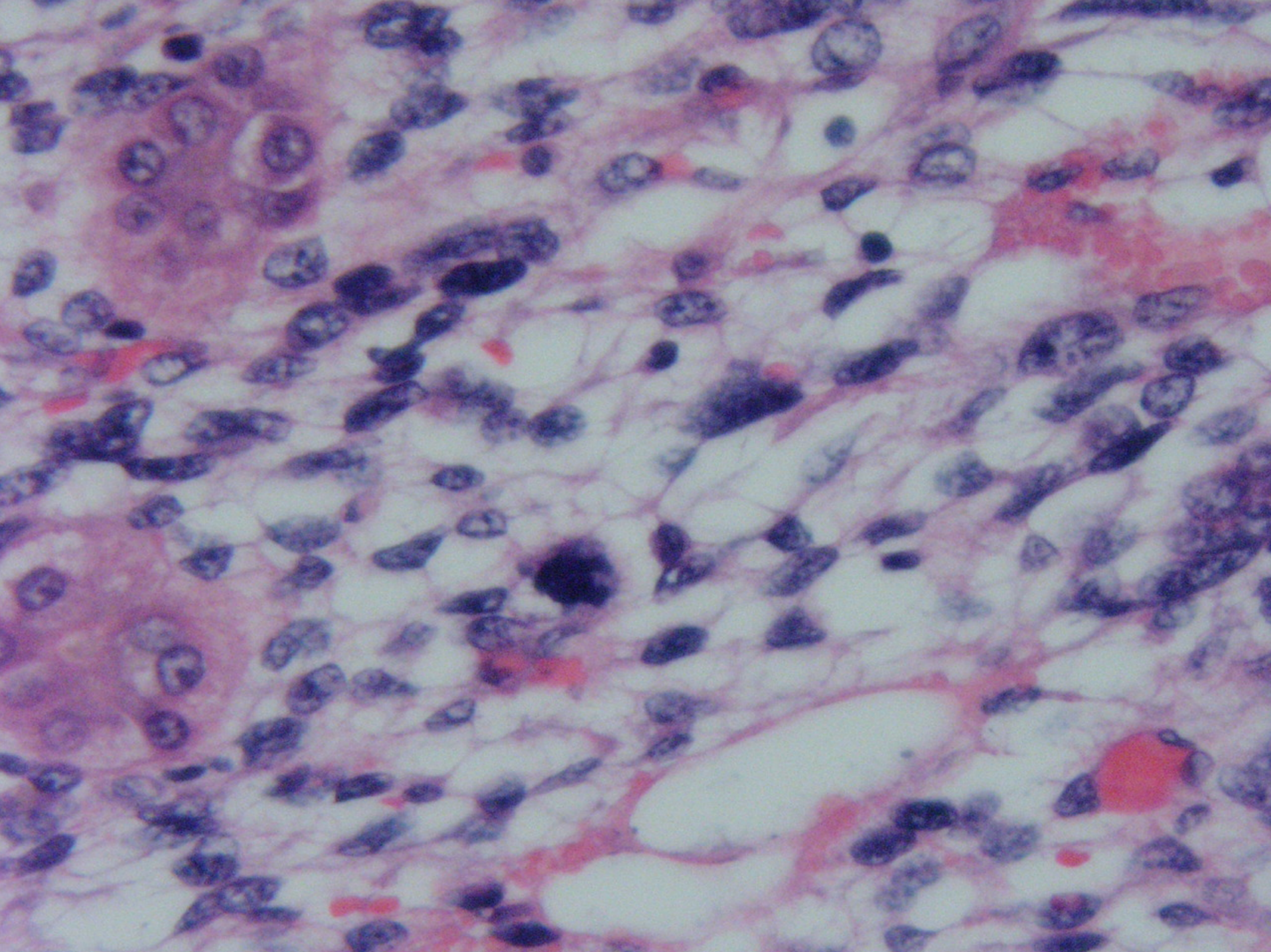
- Adenosarcoma
- Atypical polypoid adenomyoma
- Endometrial hyperplasia
- Endometrial adenocarcinoma
- Leiomyoma
- Placental polyp

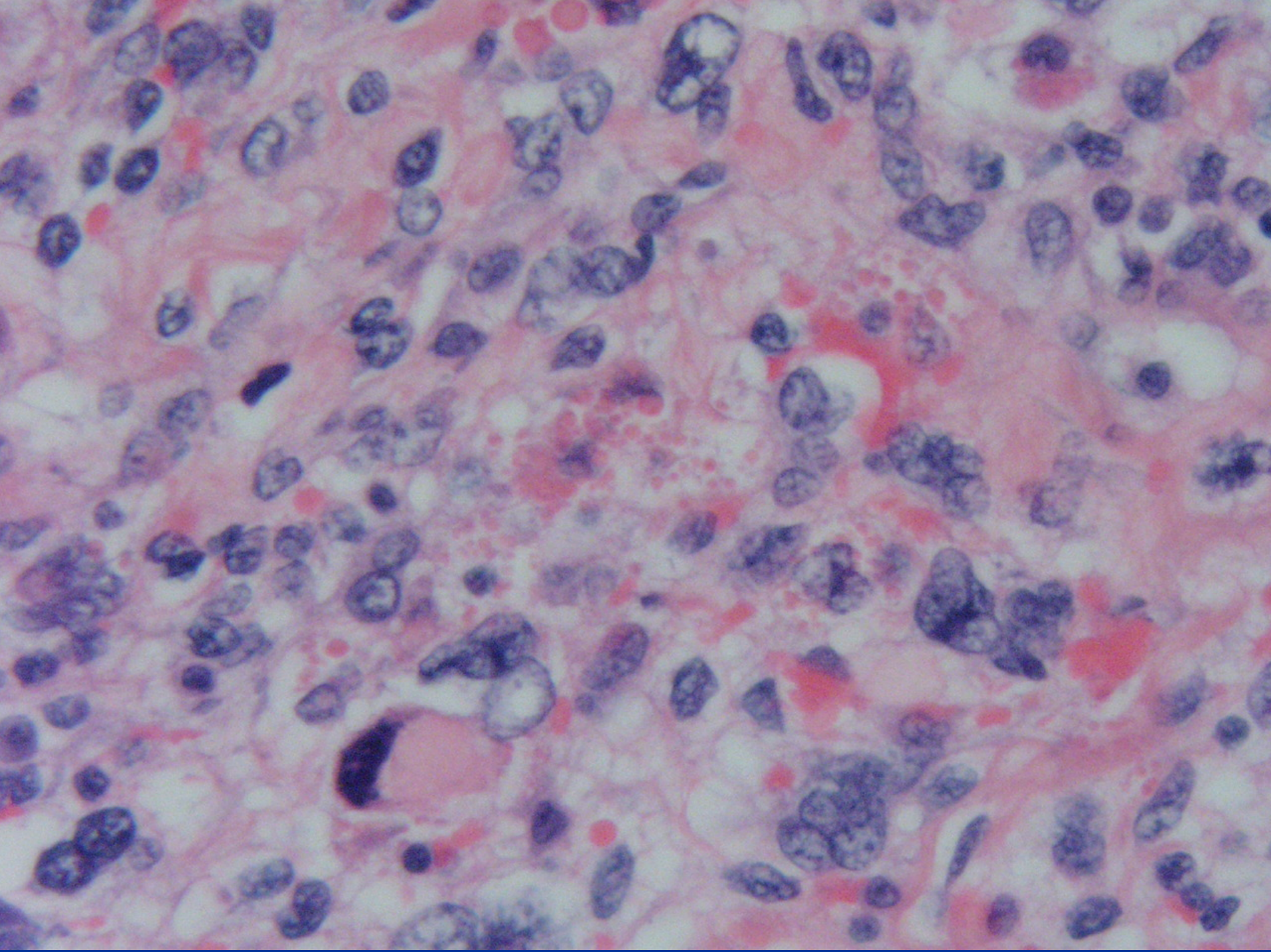
Case 25

- 16F
- Abdominal pain
- CT with subcapsular hepatic hematoma
- Several month hx of right flank and shoulder pain
- Later CT showed mass in right lobe of liver
- LFT and AFP normal
- Pseudoencapsulated 20cm mass
- Weak VIM, negative CK, CD31, FVIII, actin, desmin
- PAS+DR intracytoplasmic inclusions









Embryonal (Undifferentiated) Sarcoma of the Liver

Clinical

- Late childhood
- Second most common malignant tumor in children between ages 5-20 yrs
- M=F
- Abdominal mass and/or pain
- LFTs may be normal, AFP normal
- Large or multiple masses, usually right lobe
  - May extend to diaphragm or IVC

Prognosis

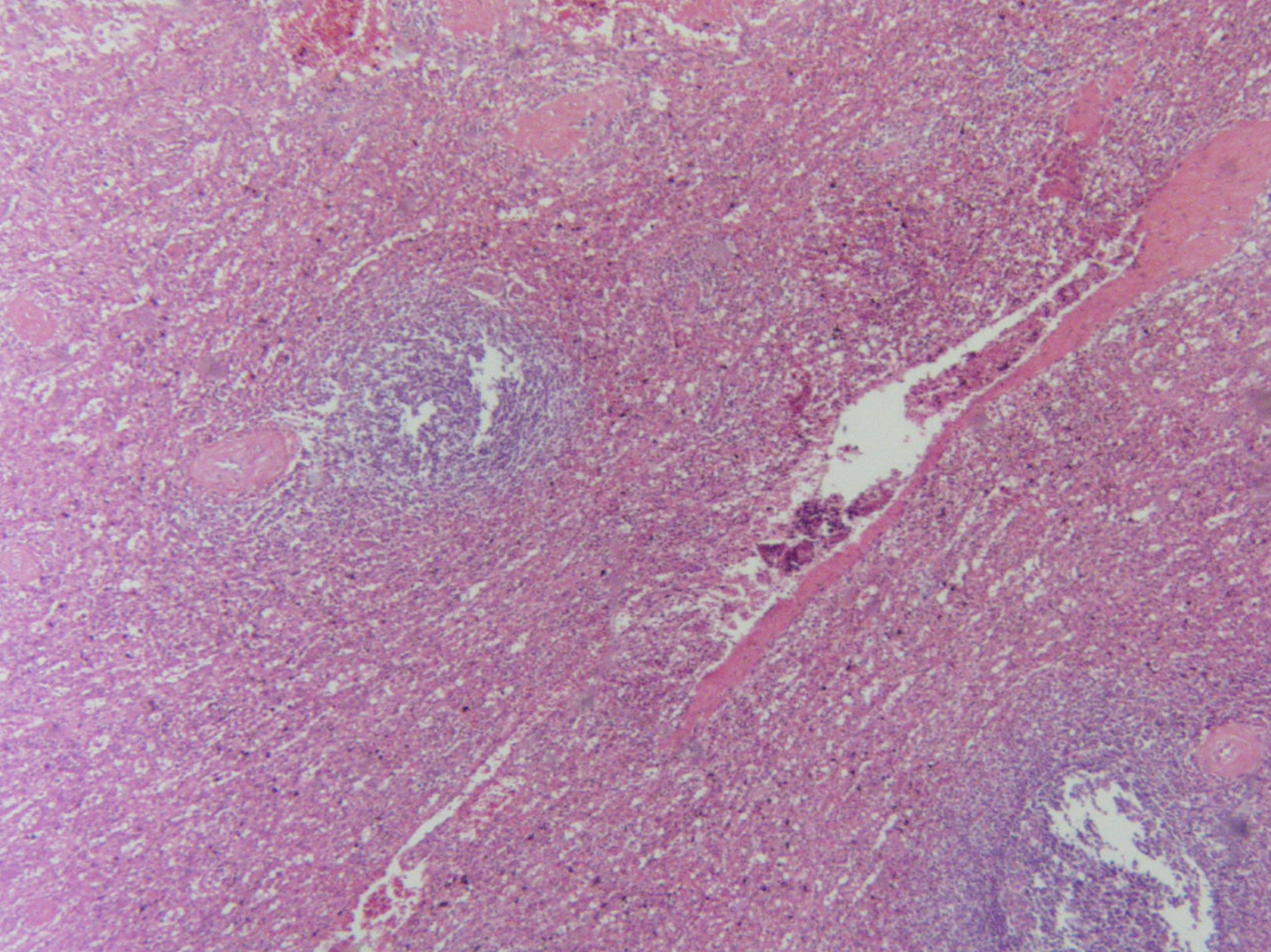
- 9% survival
- Local recurrence common after surgery
- Distant mets common-lungs
- Liver transplant with chemotherapy may offer disease free survival in 70-80%

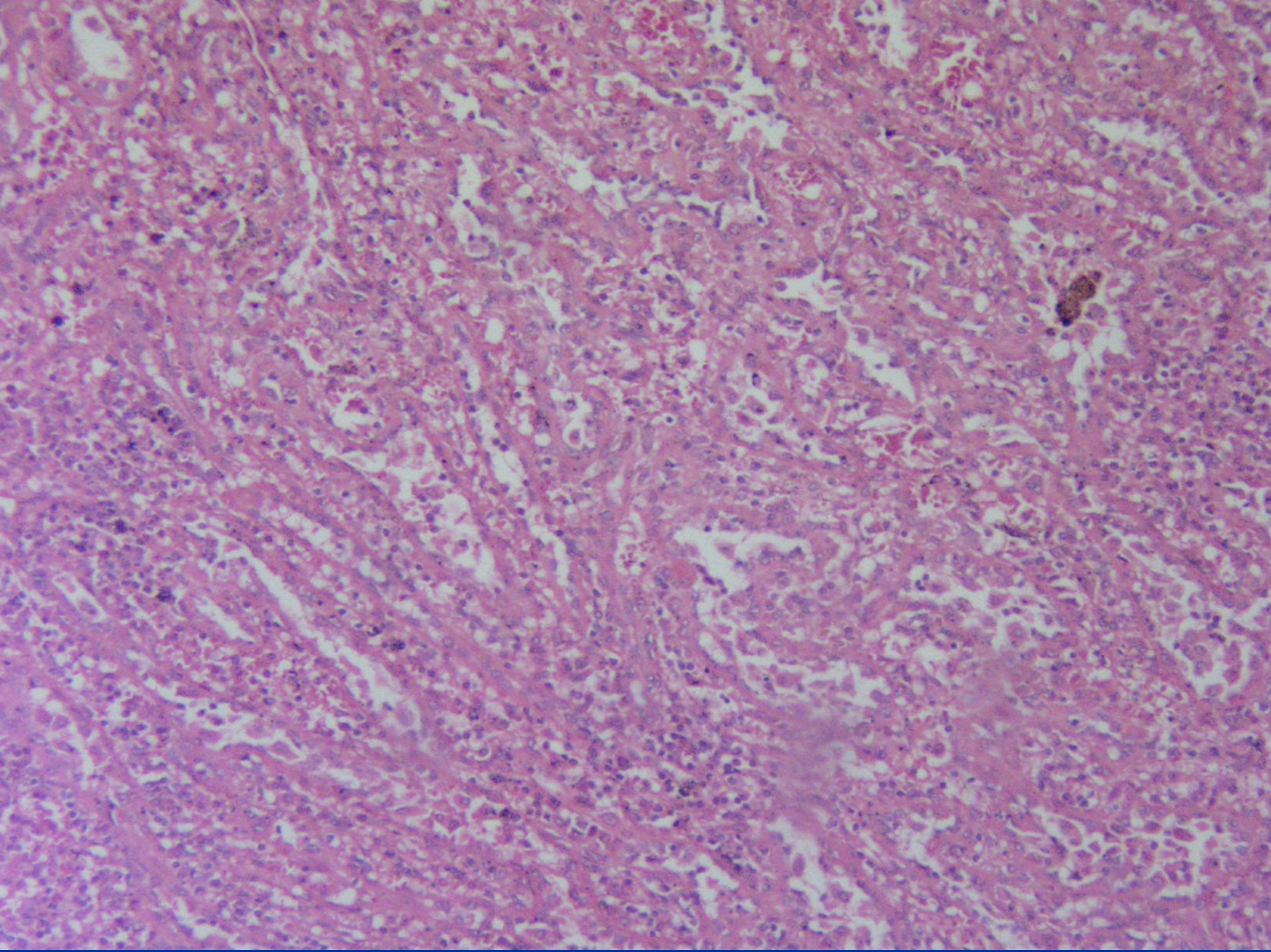
DDX

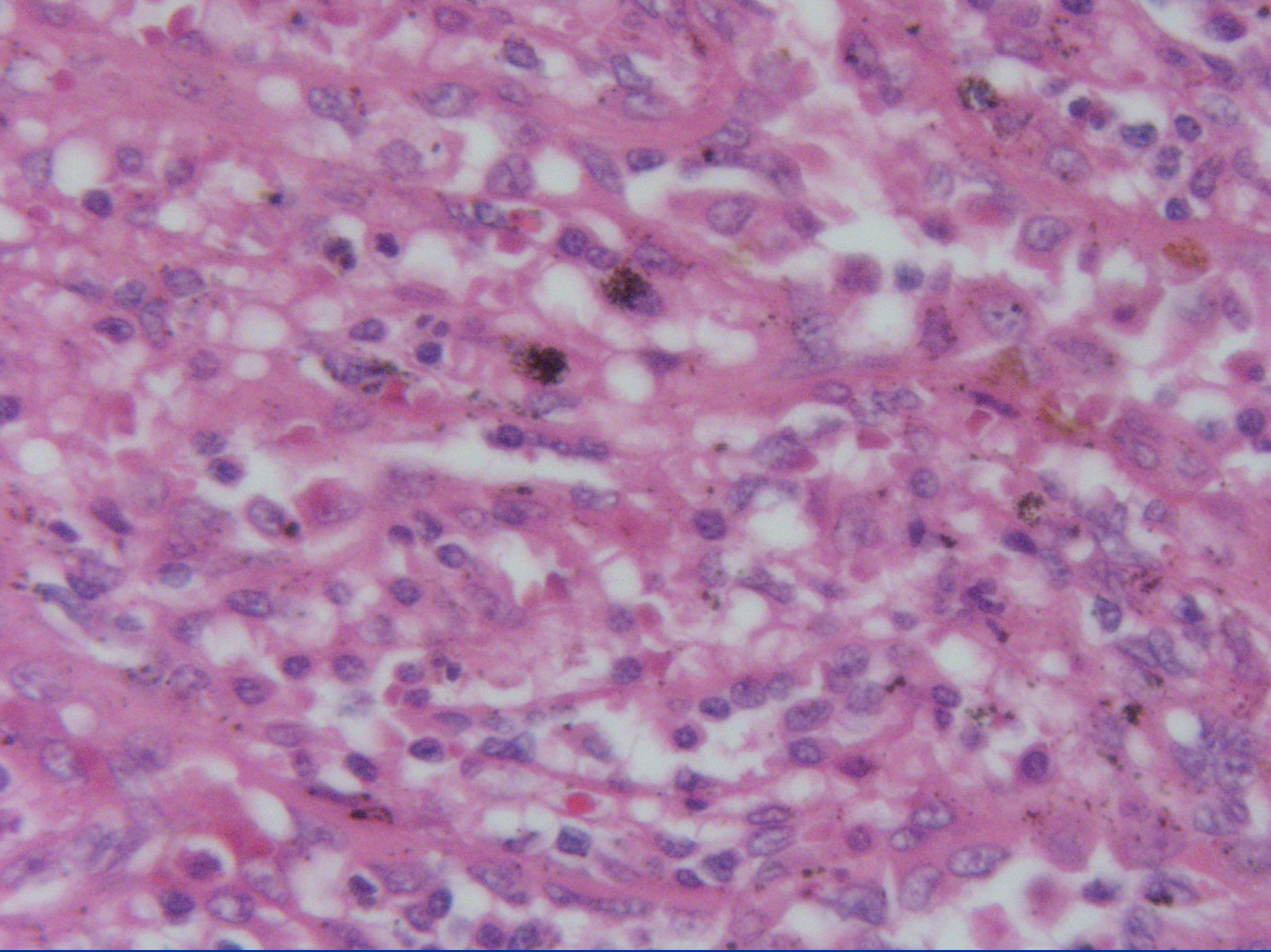
- Angiosarcoma
- Embryonal rhabdomyosarcoma
- Hepatocellular carcinoma
- Mesenchymal hamartoma
  - M>F
  - Bland cells, low MF
- Metastatic sarcoma

Case 26

- 63M abdominal pain
- CT with multinodular spleen
- 497g spleen 14x13.5x5.3 cm
- Capsular surface multinodular and hemorrhagic







Littoral Cell Angioma of the Spleen

Clinical

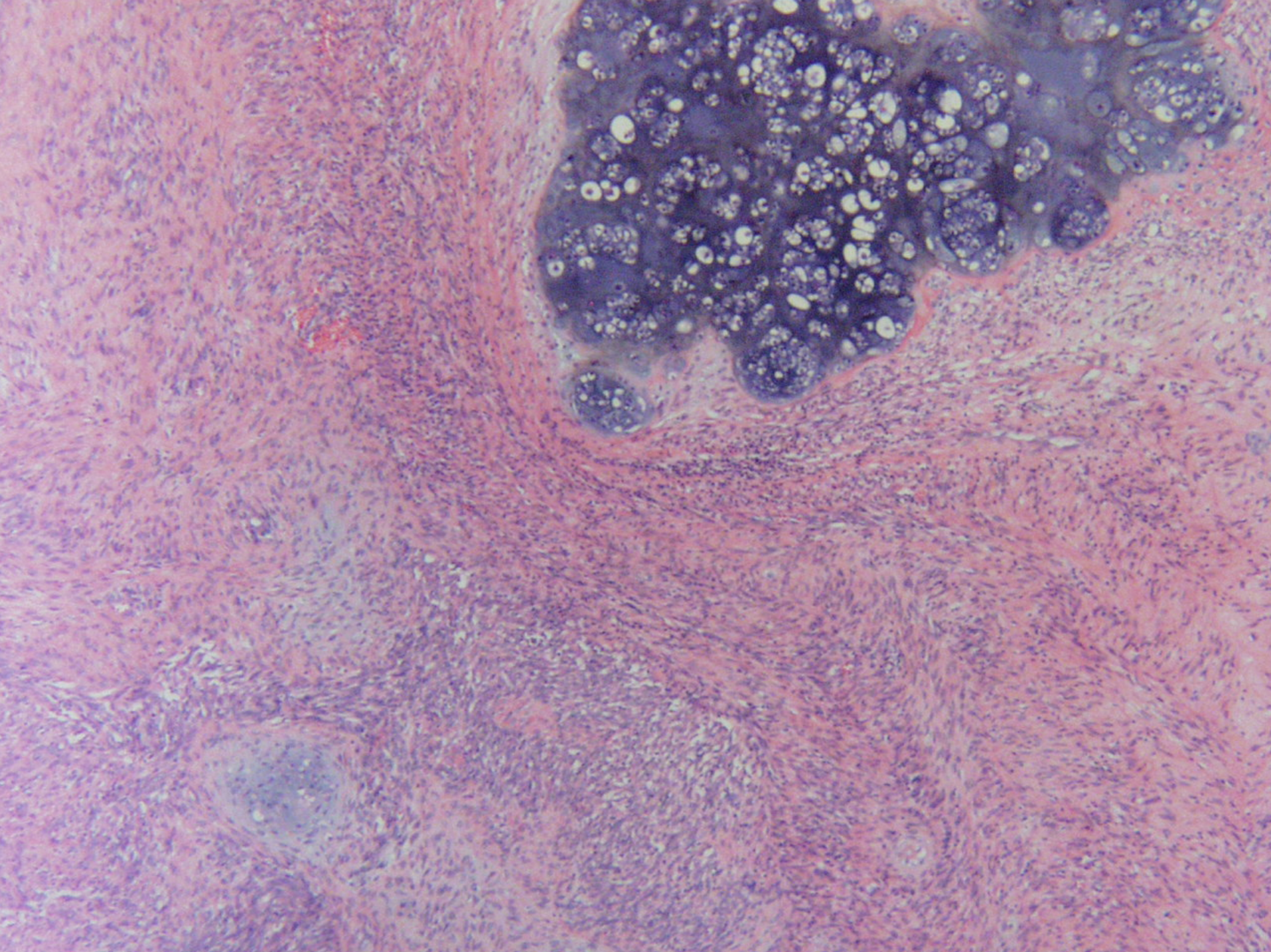
- Neoplastic counterpart of Littoral cells lining the splenic sinus channels of the red pulp
- M=F
- Usually splenomegaly, possible rupture
- Possible association with visceral malignancies
  - Colon, renal, and pancreas

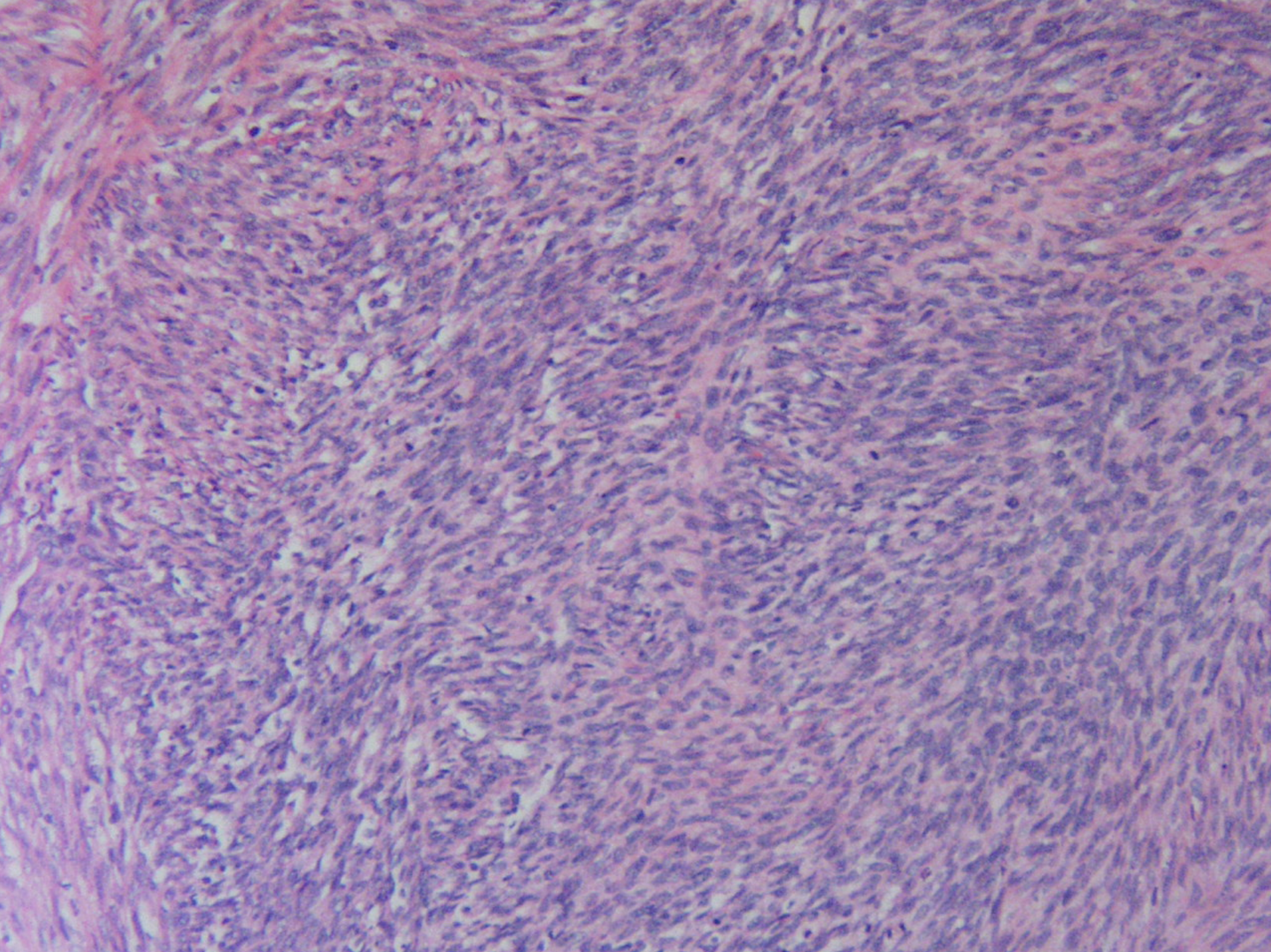
DDX

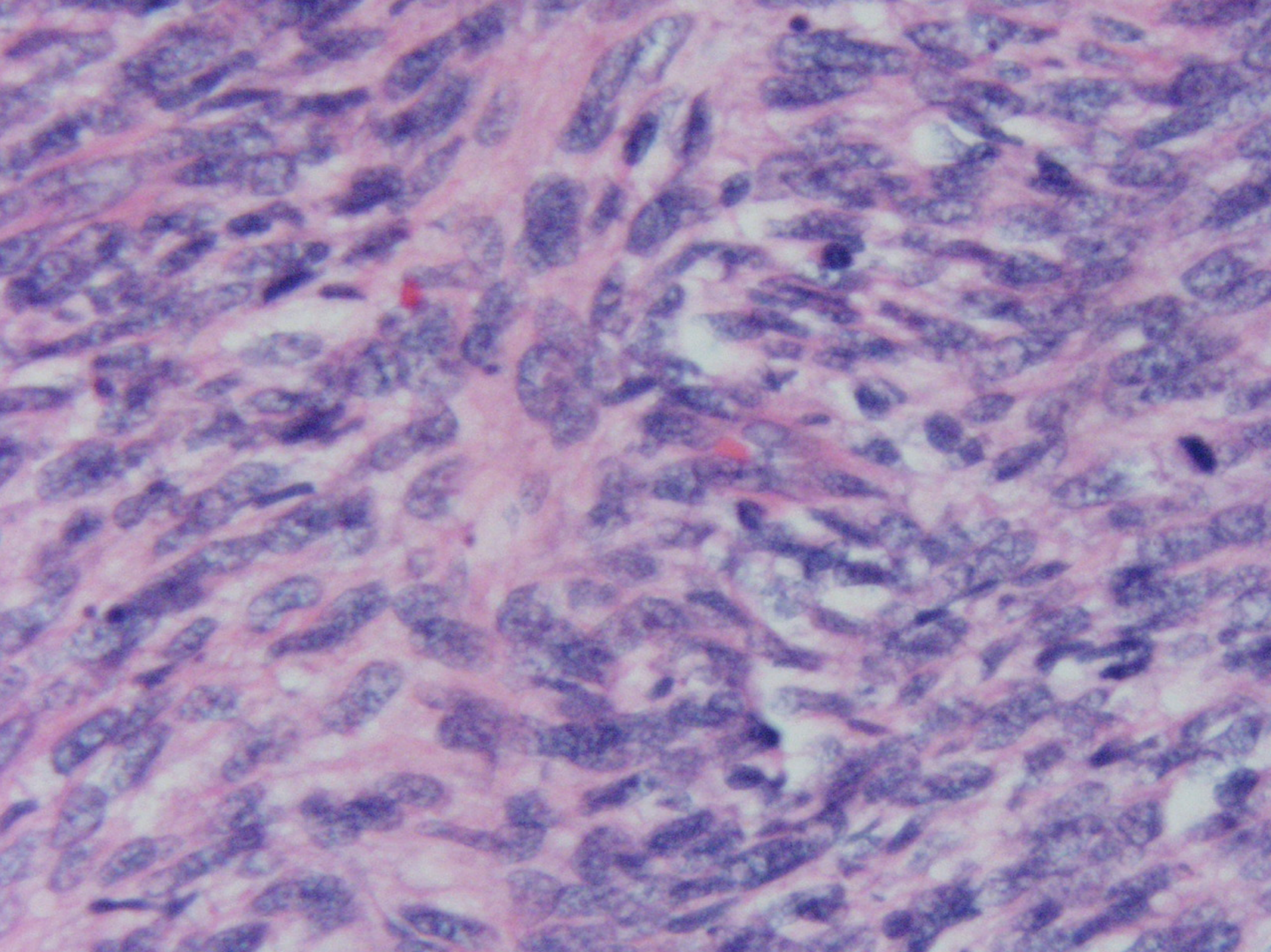
- Angiosarcoma
- Bacillary angiomatosis
- Lymphangioma
- Peliosis
- Splenic hamartoma
  - Collapsed spaces lined by slightly plump cells without atypia, similar to surrounding red pulp
  - Lining cells CD8+
- Splenic hemangioma

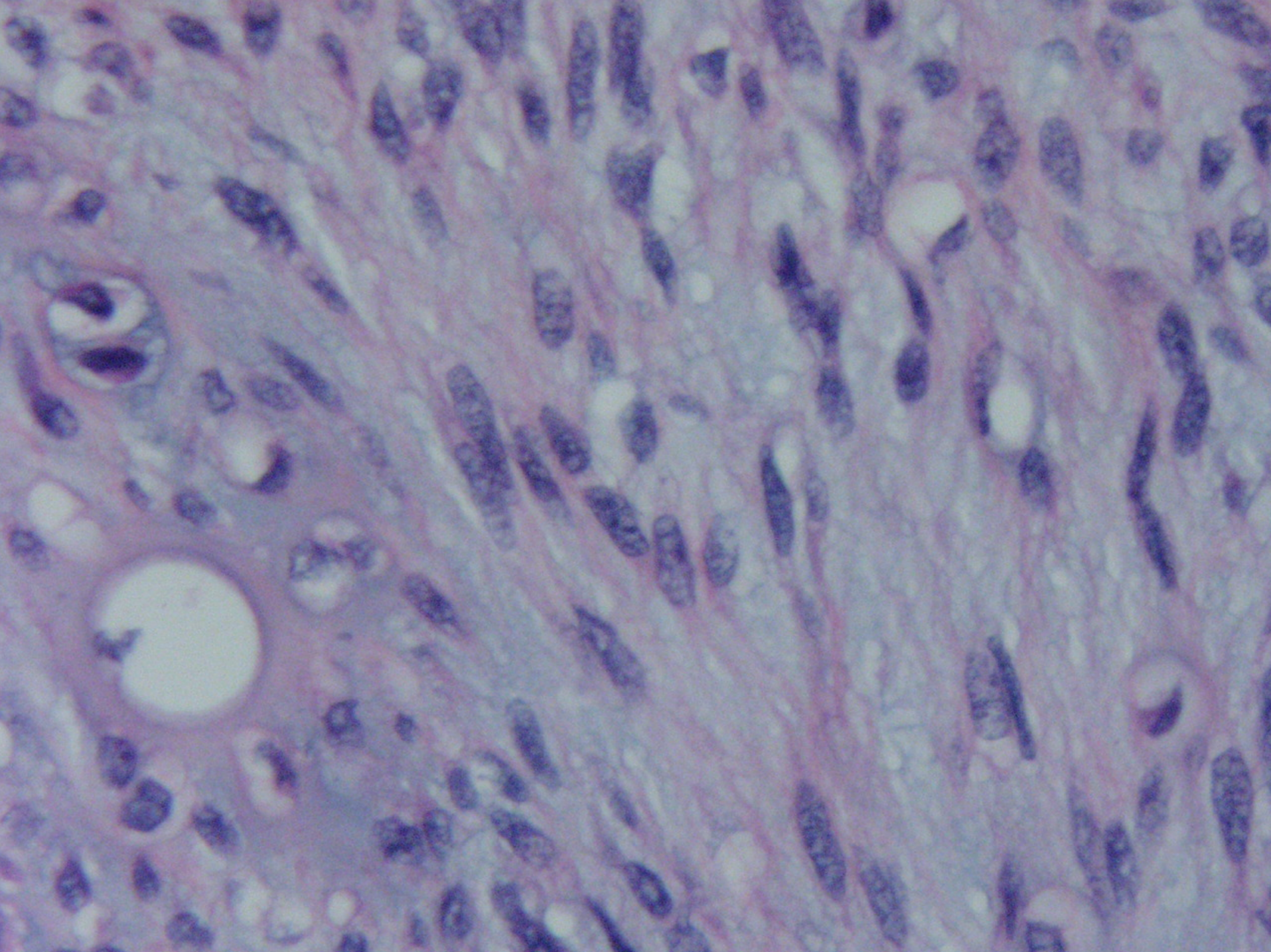
Case 27

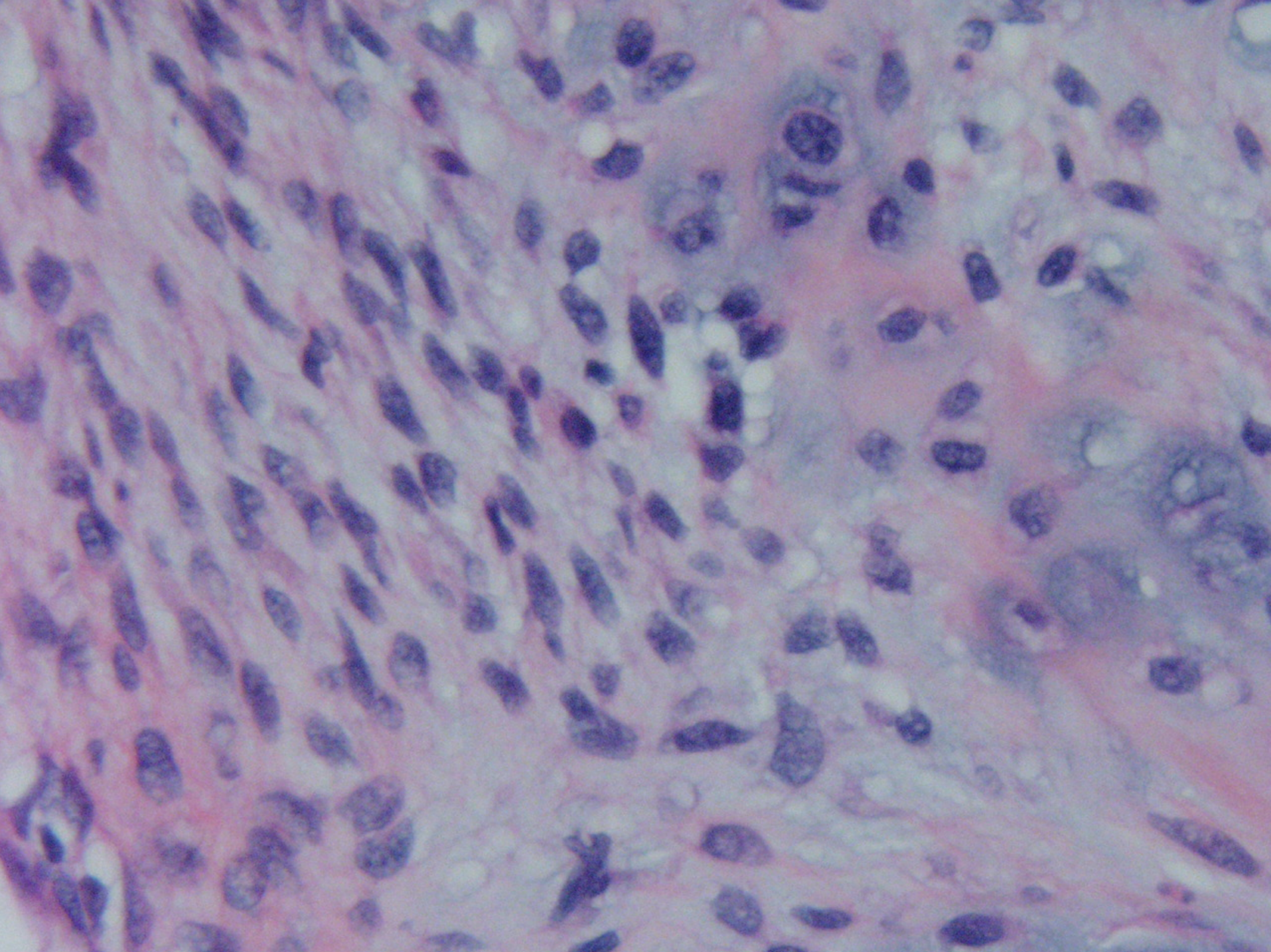
- 20M hx neurofibromatosis I
- Painful enlarging mass in right thigh
- 14x4x4.5 cm firm yellow mass
- VIM and S100+
  Negative for CD34, actin, EMA











Malignant Peripheral Nerve Sheath Tumor of Soft Tissue

Clinical

- 25-50% with NF1 patients, usually >30 yrs
  3-5% patients with NF1 develop MPNST
- 80% of NF1 pts. will have adjacent neurofibroma
- Prox. Upper or LE
- 5YRS of 52%
  Local recurrence 40-65%
  Mets in 40-68%, lung, bone, pleura
  Prior rdx may give rise to 10-15% of tumors

Histopathology

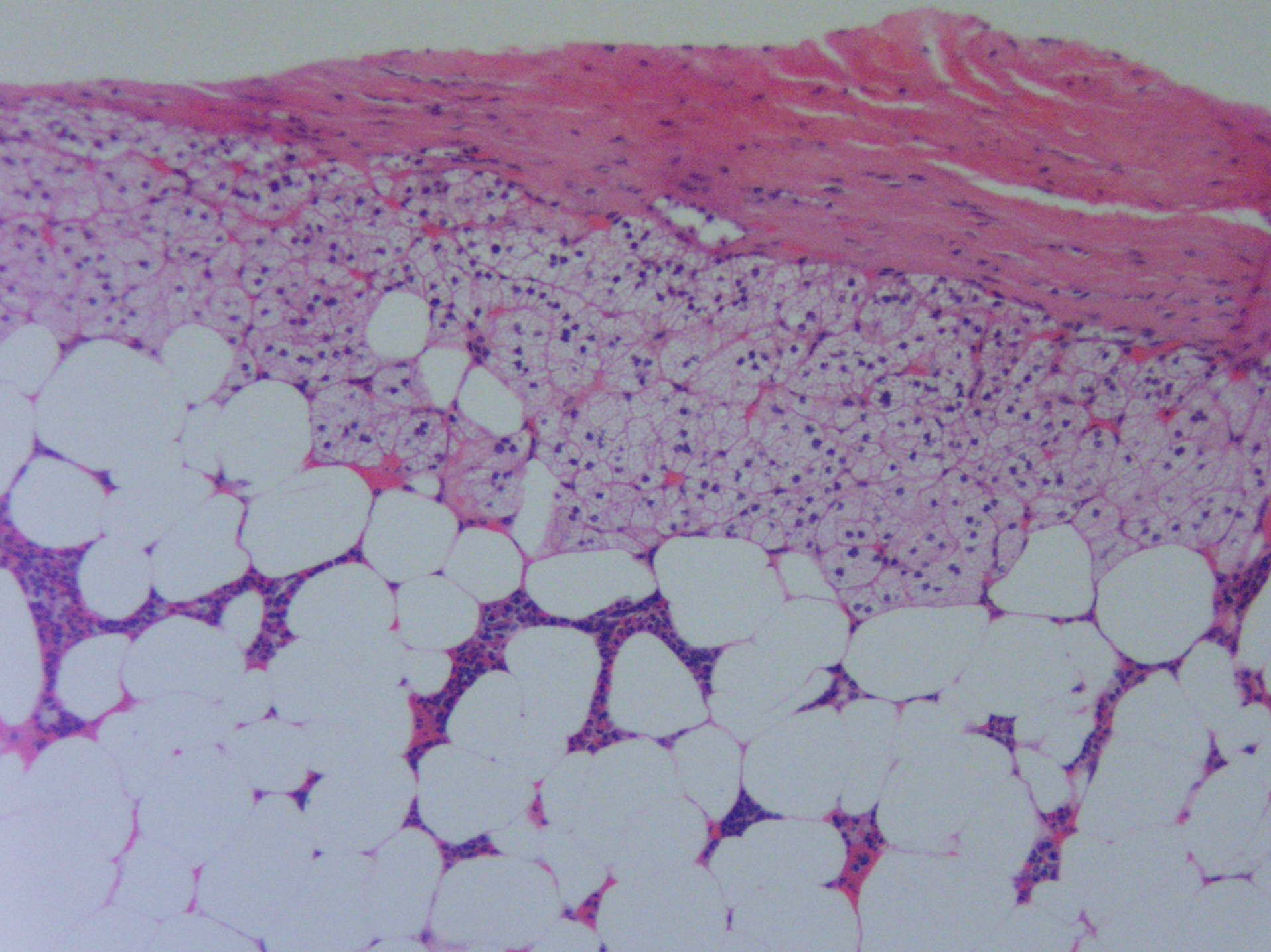
- NF1 deletion (monosomy 17) in 76% of cases
  - NF 31%
    Hemangiopericytomas 17%
    Fibrosarcomas 17%
- Homozygous p16 del in 45%
- EGFR amplification in 26%

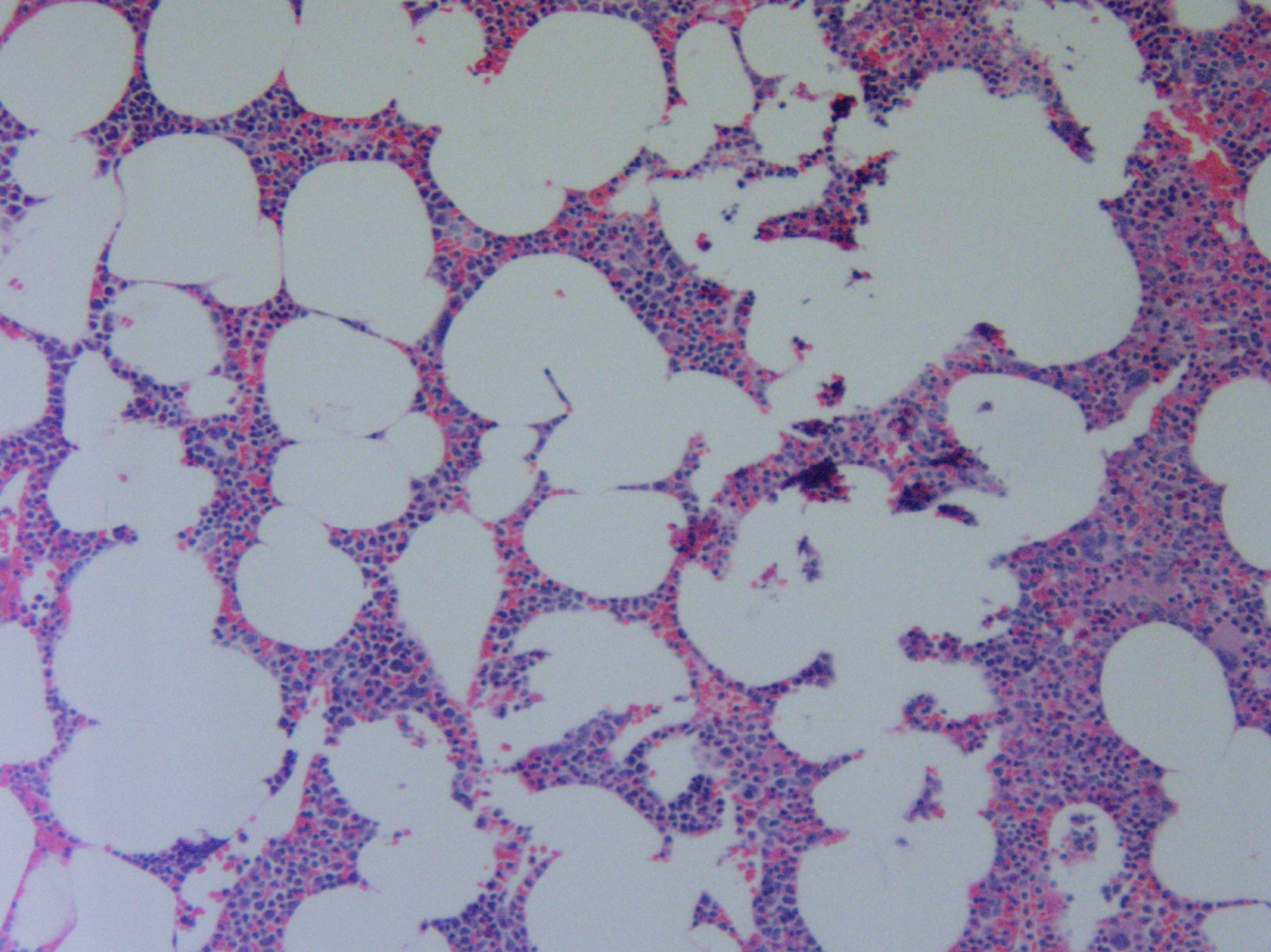
DDX

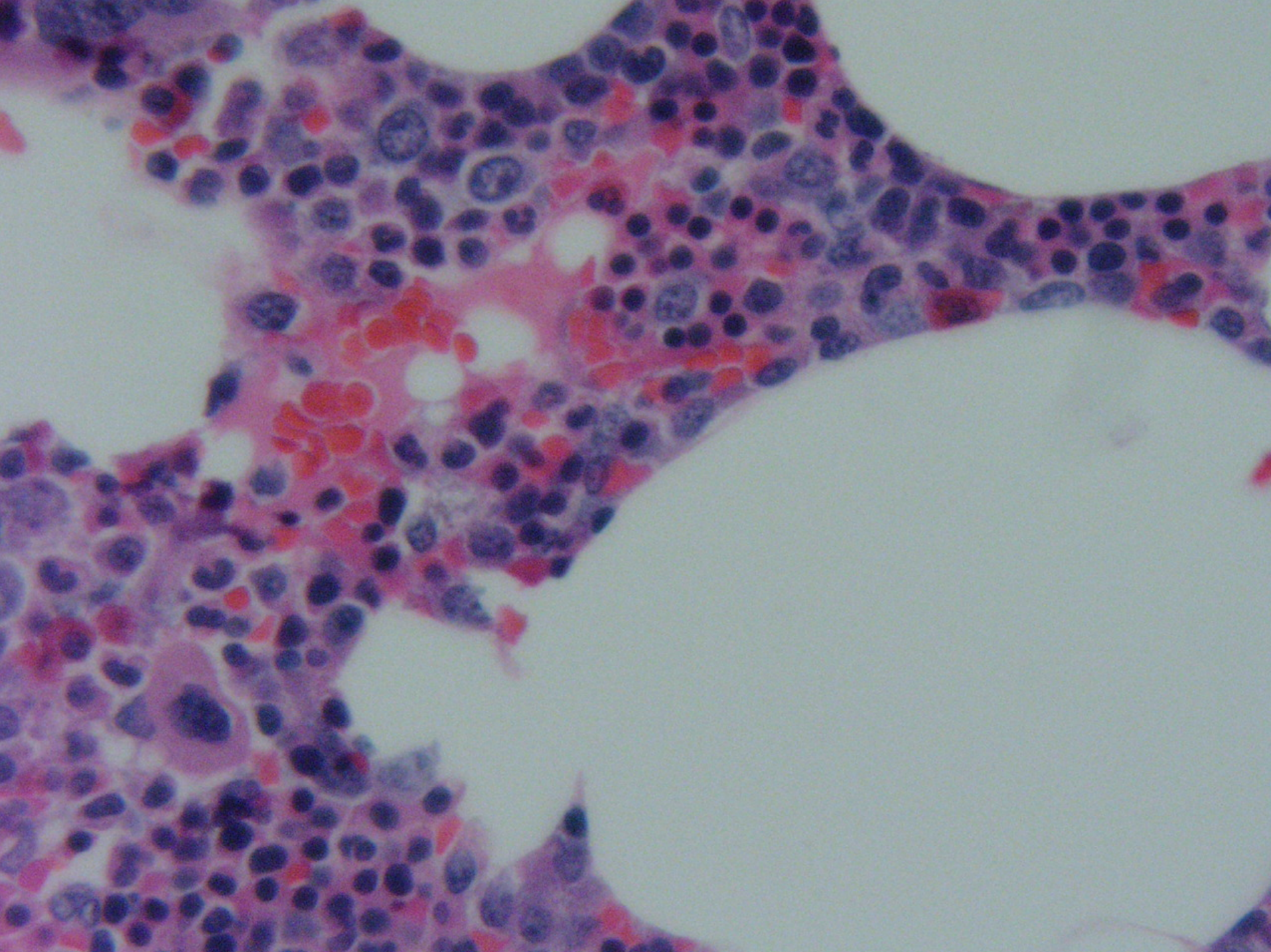
- Cellular schwannoma
- Fibrosarcoma
- Hemangiopericytoma
- Leiomyosarcoma
- Monophasic synovial sarcoma
  - CK, t(X;18)(p11.2, q11.2) in 90%
- Plexiform neurofibroma
- Spindle cell melanoma

Case 28

- 34M hx. Idiopathic cardiomyopathy
- CT solitary mass of adrenal gland
- 8cm adrenal mass with lobulated yellow to gray soft mass with fatty hemorrhagic cut surface







Myelolipoma of the Adrenal Gland

Clinical

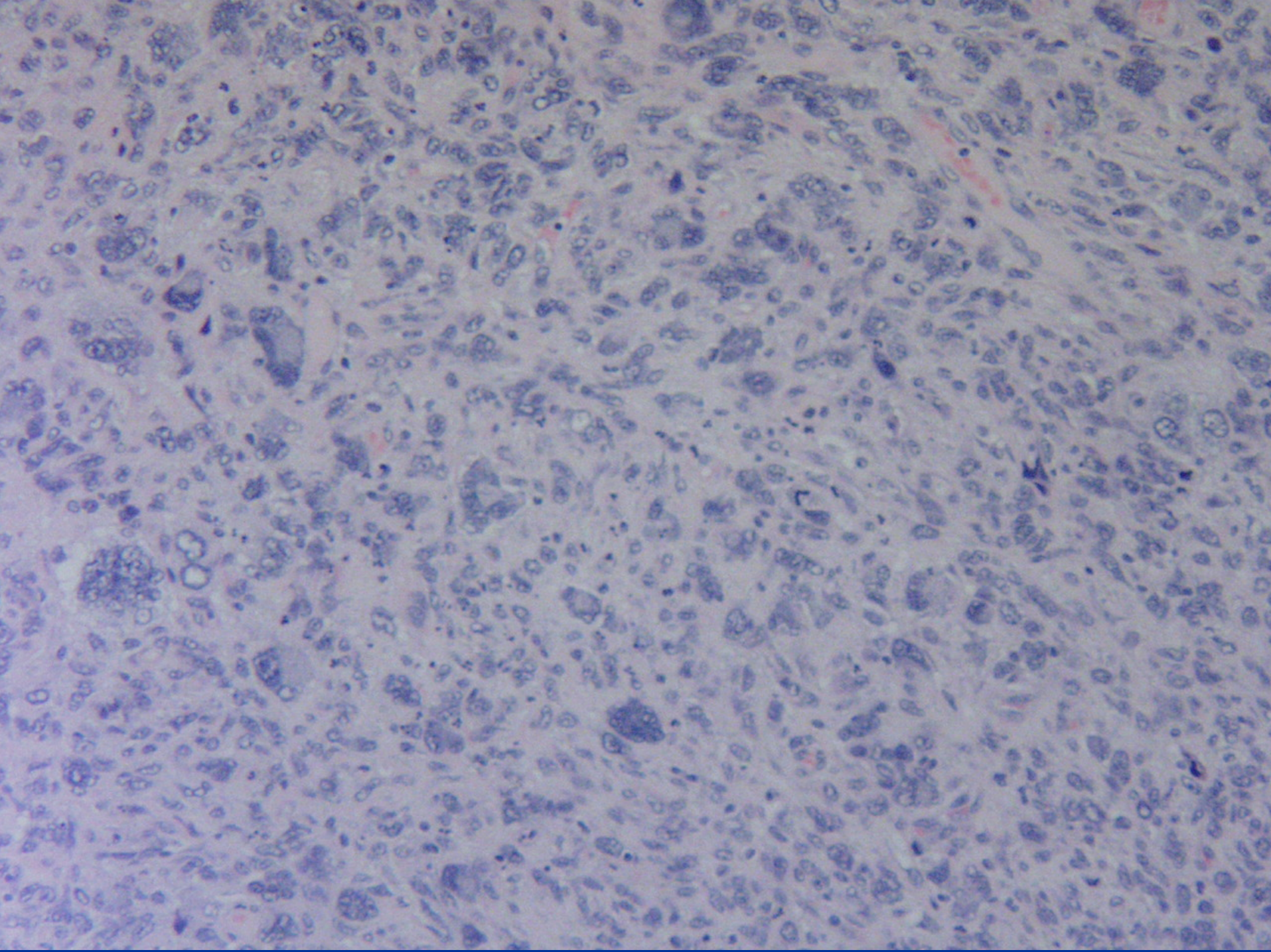
- >40 yrs
- Usually asymptomatic and incidental findings
  - Larger tumors may displace kidney and cause abdominal pain
- Radiolucent masses
- Rare association with hormonally active tumors
  - Adrenocortical adenomas, carcinomas, pheochromocytomas
- ? Hormone drive metaplasia of adrenal stromal cells

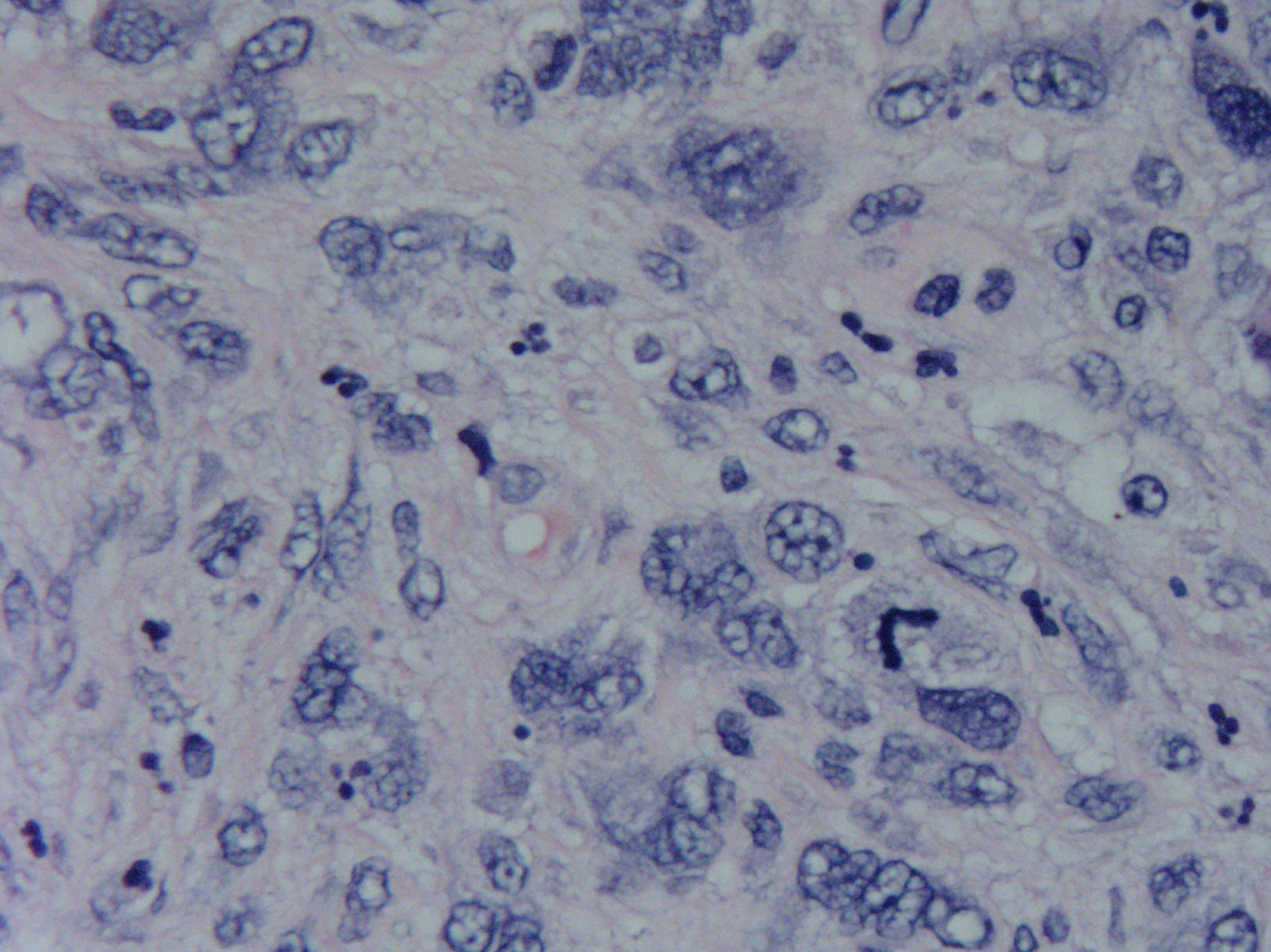
DDX

- Adrenal cortical carcinoma
- Angiolipoma
- Extramedullary hematopoietic tumor associated with essential thrombocythemia
- Extramedullary plasmacytoma
- Liposarcoma
- Myeloid sarcoma
  - No adipose tissue

Case 29

- 68M with hematuria
- Sessile bladder neck lesion
- TURB with later cystoprostatectomy
- 2.5cm ulcerated non-papillary mass involving bladder base extending into muscularis
- VIM+, focally + for CK, CEA, CK7, SMA, MSA
- Negative for EMA, CK20, S100, CD15, and CD34





Sarcomatoid Urothelial Carcinoma of the Urinary Bladder

Prognosis

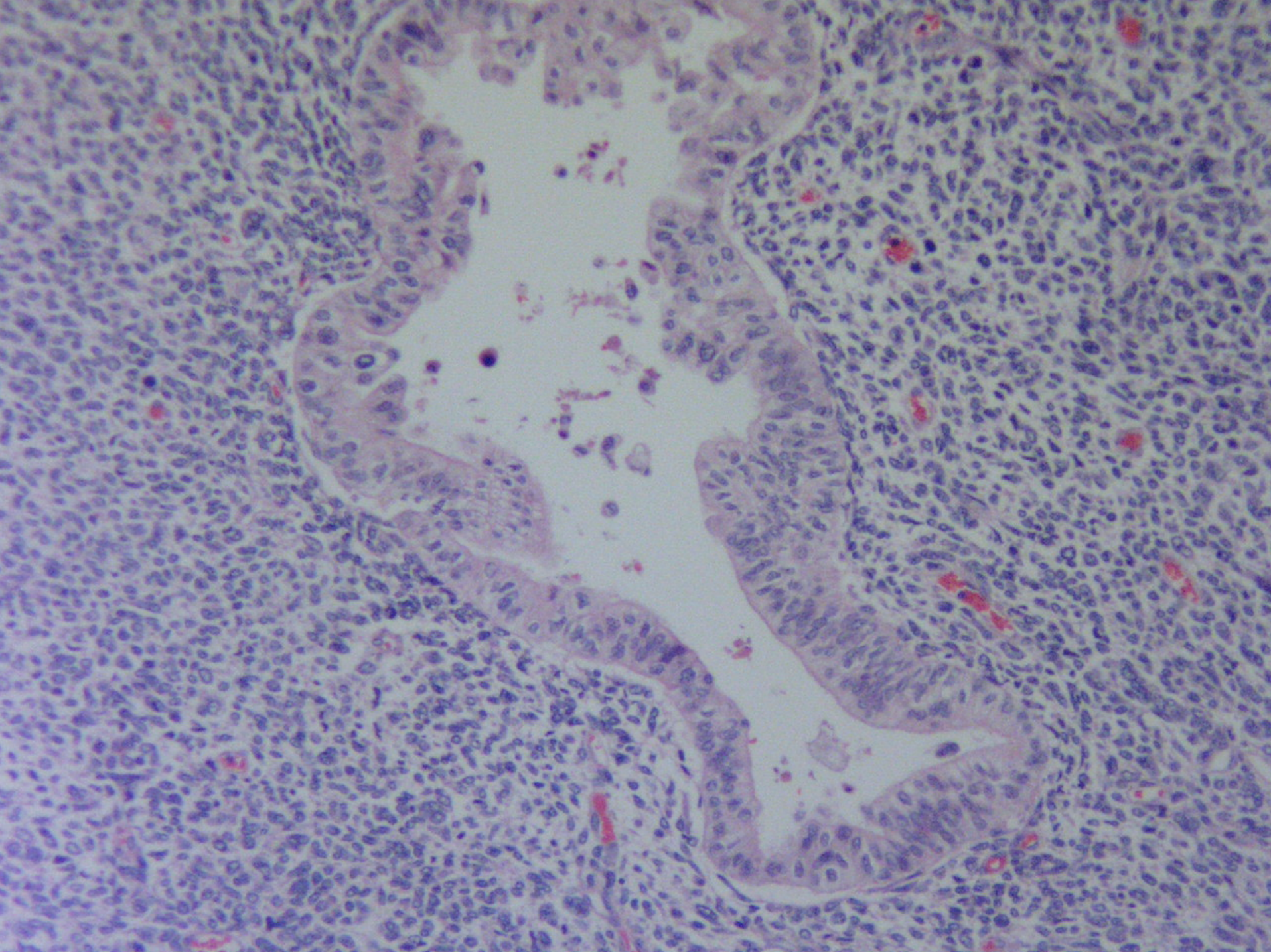
- Median survival 12-16 months
- 2YRS of 28%
  5YRS 20%
- Stage is best prognostic factor
  - No difference compared to classic urothelial CA

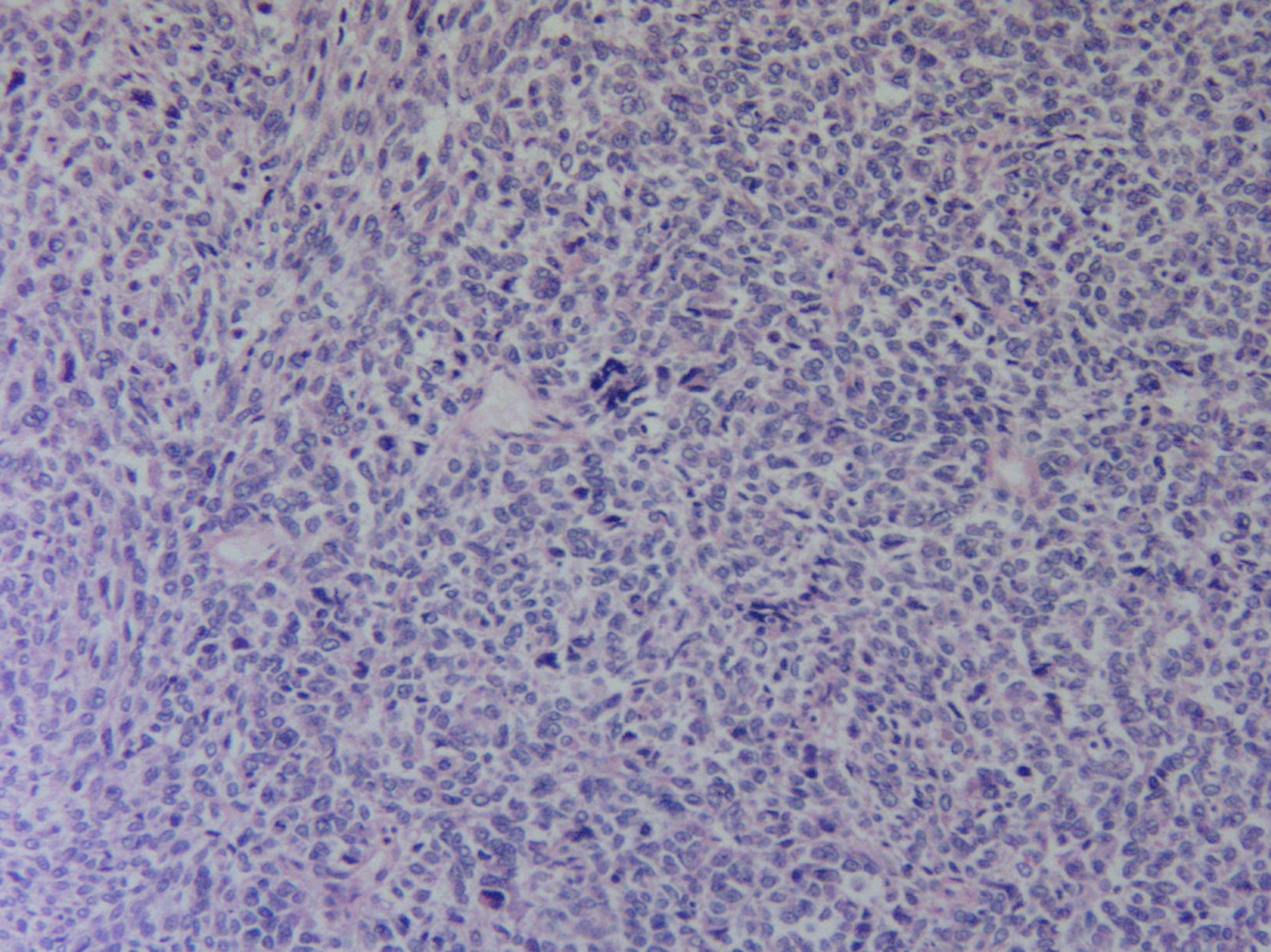
DDX

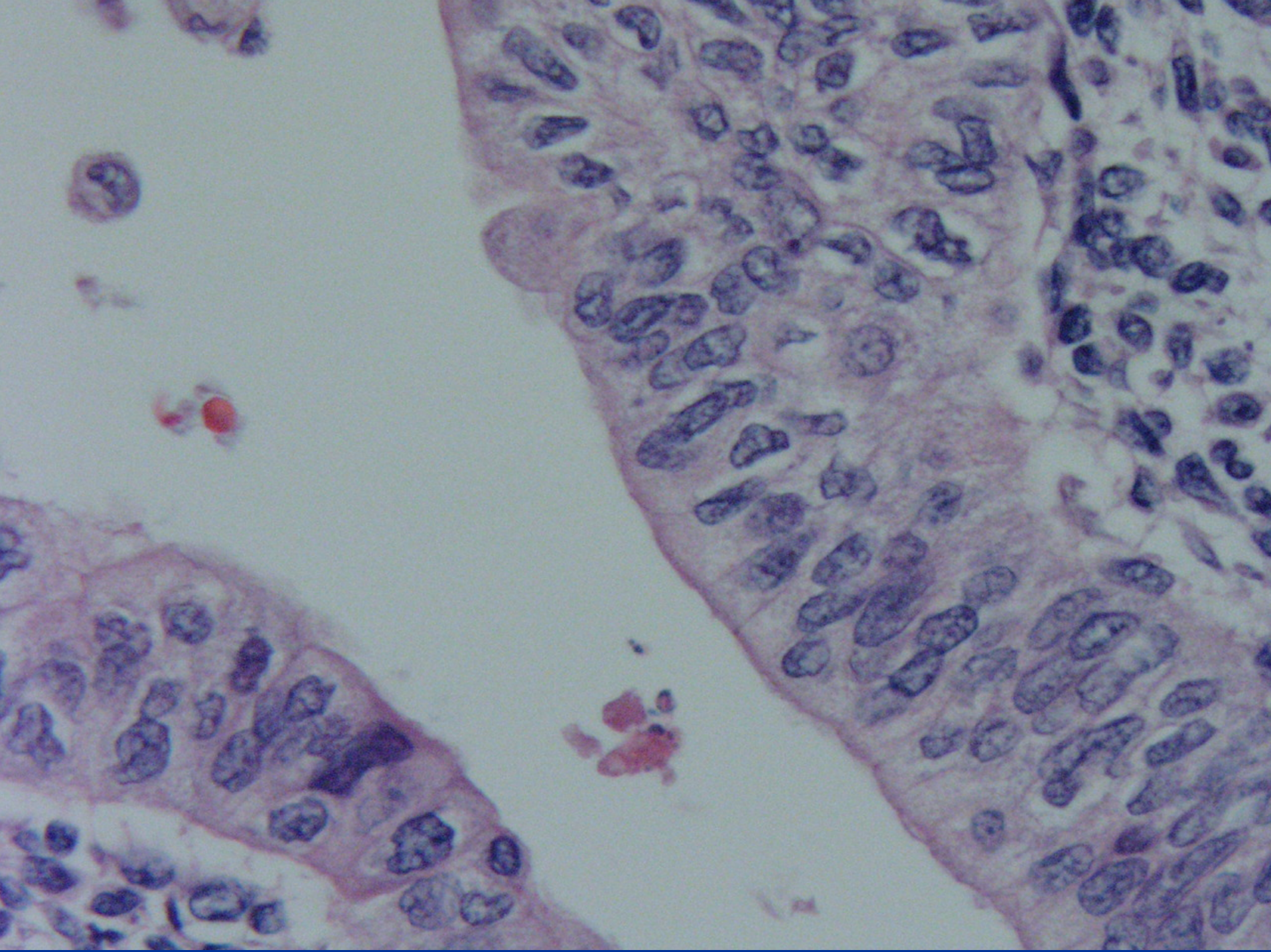
- Inflammatory pseudotumor
  - Young adults with antecedent trauma
    Variable cellularity with myxoid matrix
    MF <2 10/hpf
    +MSA, VIM, less for SMA, Des
    Rare cases of +CK, EMA
- Invasive urothelial carcinoma with pseudosarcomatous stroma reaction
- Leiomyosarcoma
- Postoperative spindle cell nodule
  - <3 months of procedure
    Numerous MF
    + for VIM,MSA, Des, CK
    - for EMA

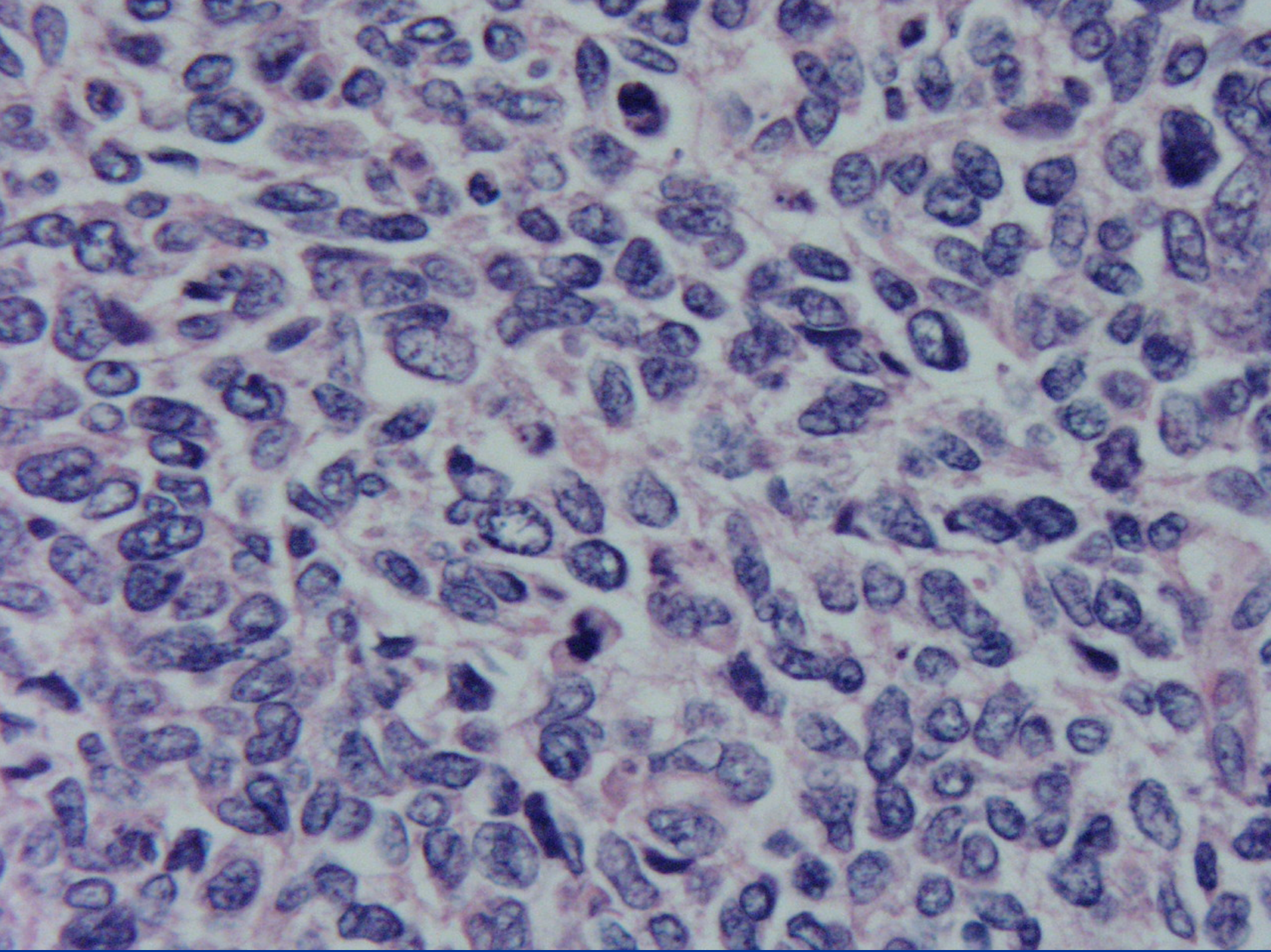
Case 30

- 36M smoking history with left upper lobectomy for tumor
- 520gm lung lobe with 21 cm friable necrotic mass
- Spindle cells: VIM+, desmin-
- Epithelial cells: CAM5.2+, EMA+, CEA+, PAS+
- Both cells: negative for CK20, chromogranin, S100, HMB45









Biphasic Pulmonary Blastoma

Clinical

- Rare 0.25-0.5% primary lung tumors
- Pulmonary symptomatology
- Mean and median age 4th decade
- M=F
- Smoking history
- Large peripheral lesion (average 10 cm), rarely multiple
- 5YRS of 16%
- Mets to nodes, lung, pleura, and brain

Histopathology

- Malignant glandular component admixed with malignant primitive mesenchymal stroma
- Glands resemble pseudoglandular phase of fetal lung development
  - Subnuclear or supranuclear vacuoles
  - Squamous morules and pearls
- Stroma may contain heterologous elements
  - Chondrosarcoma, osteosarcoma, rhabdomyosarcoma

Pulmonary Blastomas-Former

- Adult type
  - Biphasic pulmonary blastomas
  - Well-differentiated fetal adenocarcinomas
- Childhood type
  - Pleuropulmonary blastoma

WHO Classification

- *Well-differentiated fetal adenocarcinoma
  - Subtype of adenocarcinoma
- *Biphasic pulmonary blastoma
  - Carcinomas with pleomorphic, sarcomatoid, or sarcomatous elements
- Pleuropulmonary blastoma
  - Soft tissue neoplasm of the lung

*May represent a spectrum

DDX

- Leiomyosarcoma
- Malignant melanoma
- Mesothelioma, biphasic
- Pleuropulmonary blastoma
  - First 2 decades
    Malignant embryonal stroma without epithelial
- Pulmonary adenocarcinoma, acinar pattern
- Synovial sarcoma
- Well-differentiated fetal adenocarcinoma
  - Stroma is benign