2003 PIP-D Cases

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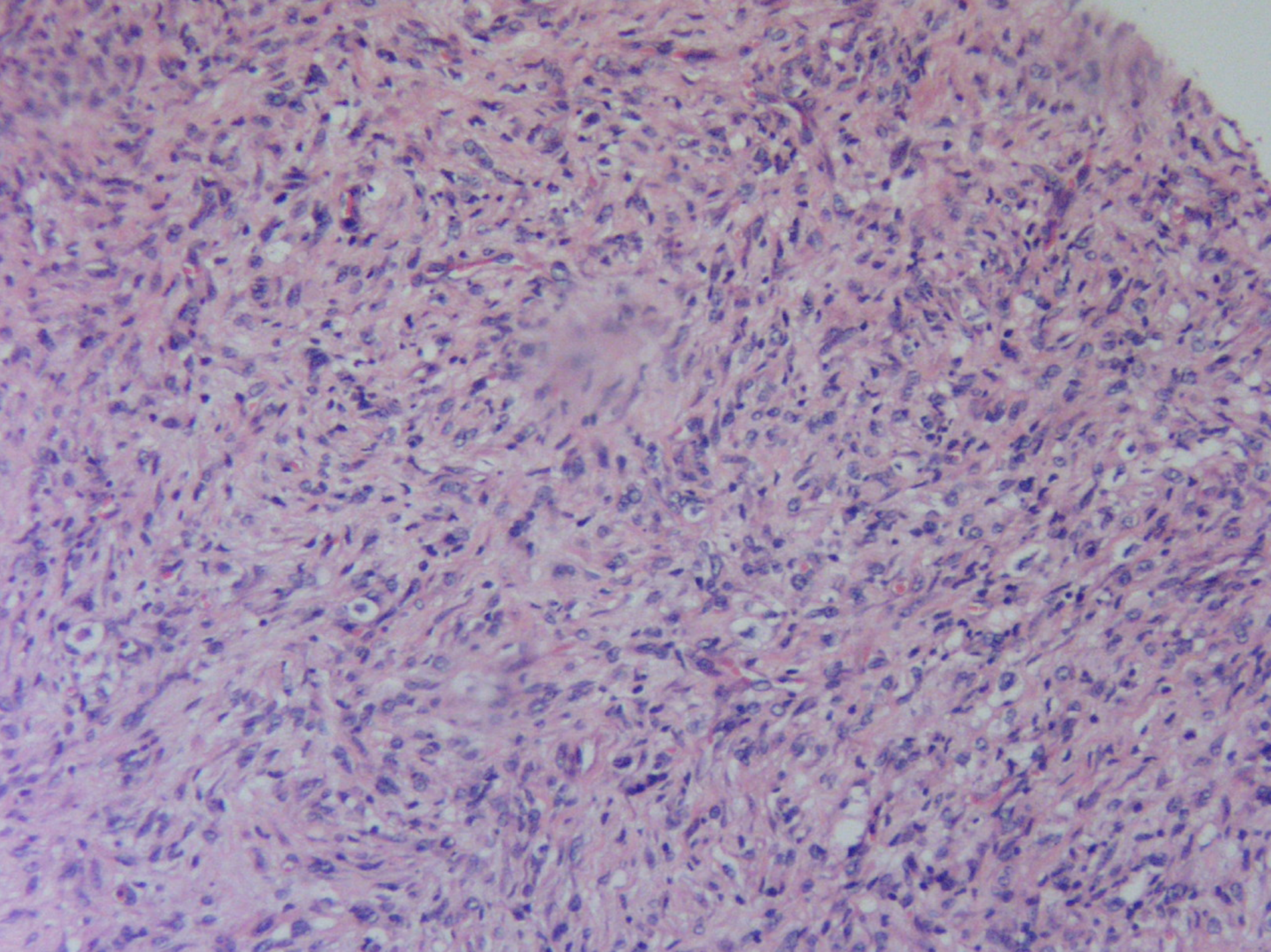
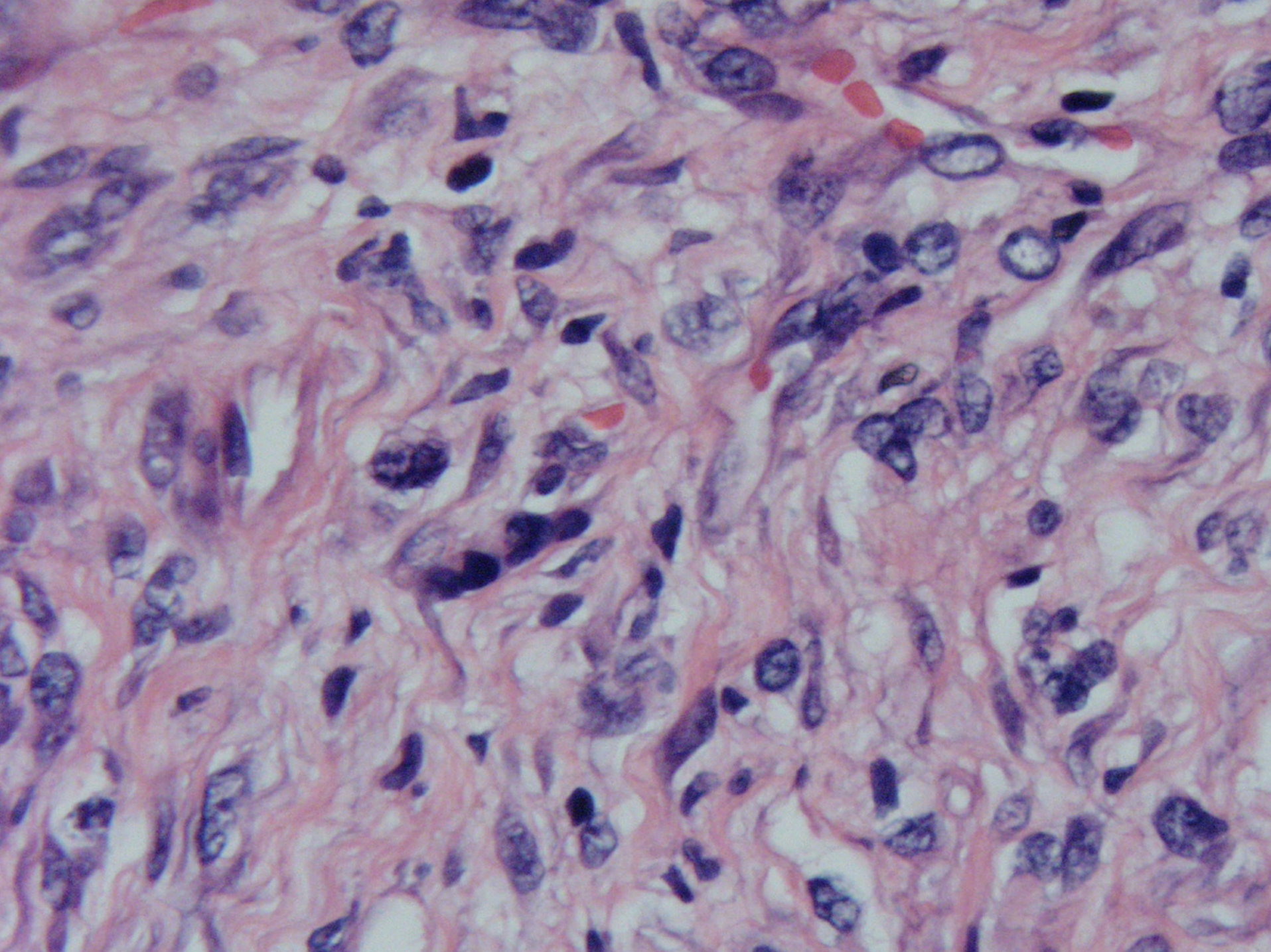
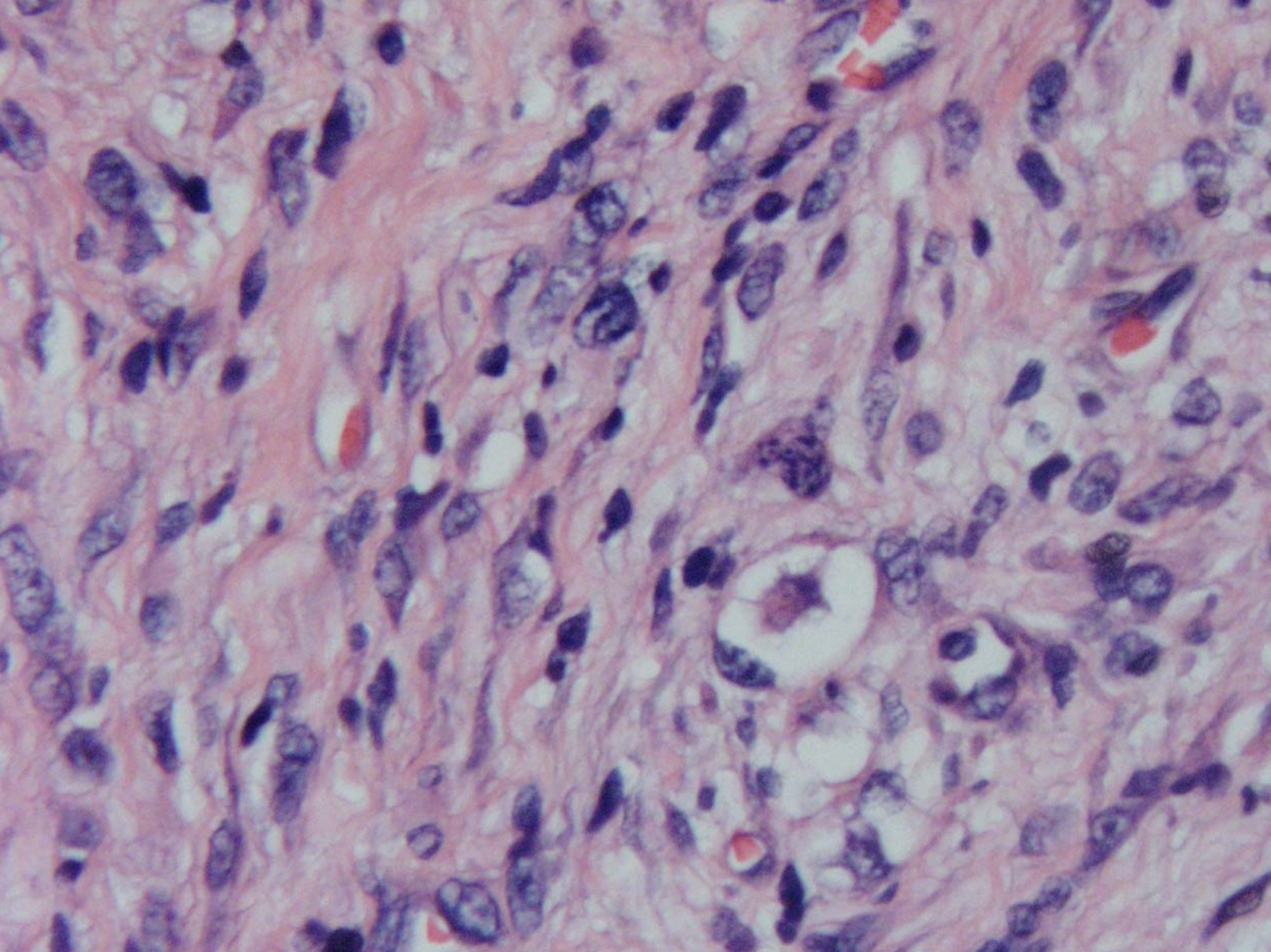
APMG

Case 31

- 4 mo boy with increasing head circumference, full fontanel, and vomiting
- Large enhancing medial temporal lobe lesion with adjacent cyst
- Angiogram with no tumor blush
- 3.5x2.5x2 cm lobulated reddish-gray firm mass with central cyst







Desmoplastic Infantile Astrocytoma/Ganglioglioma

DCA/G

- Temporal lobe with enhancing mural nodule
- Desmoplasia with spindle cells predominating
- Neuronal and astrocytic differentiation
- Increased cellularity with MF
- Occ necrosis with pseudopalisading
- Focal GFAP and +/- synaptophysin

Quick Facts

- Cerebrum most common location for CNS tumors in first 2 years of life
- Astrocytoma most common primary CNS tumor during first 2 years of life

Radiographic Features of Low Grade Tumors

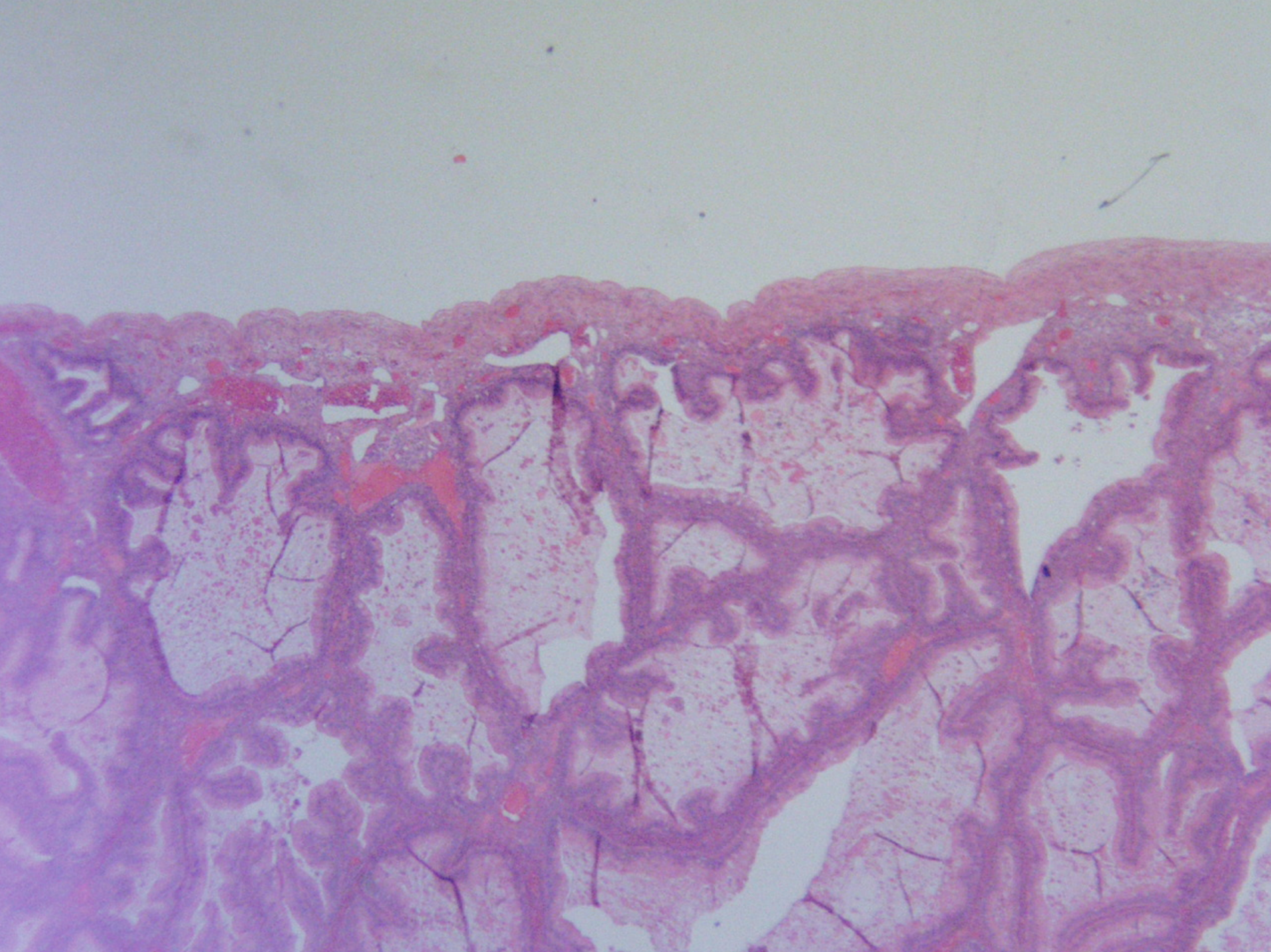
- Cystic architecture
- Intracortical location
- Skull deformation/erosion
- Solid enhancement

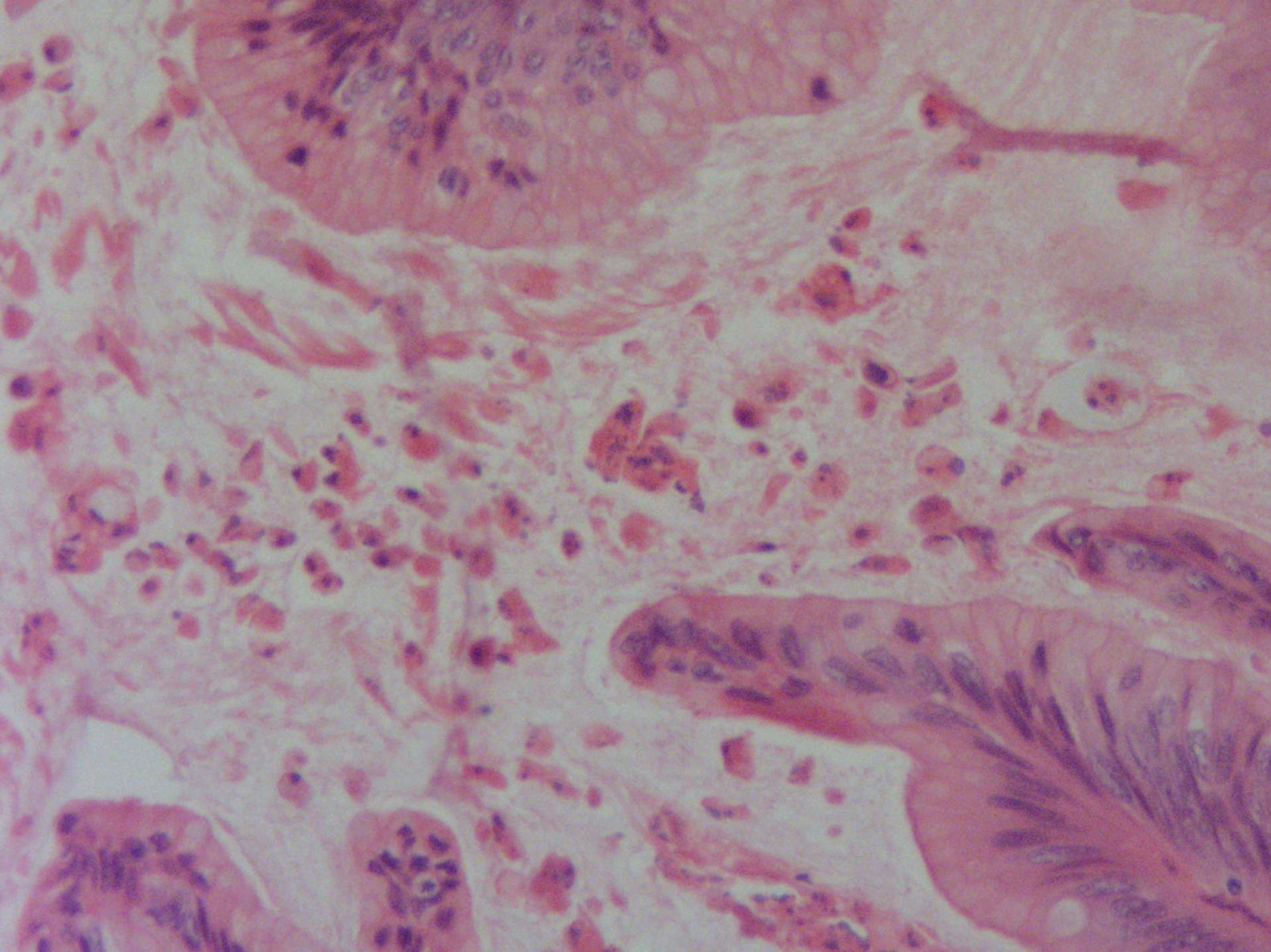
Pediatric Posterior Fossa Tumors

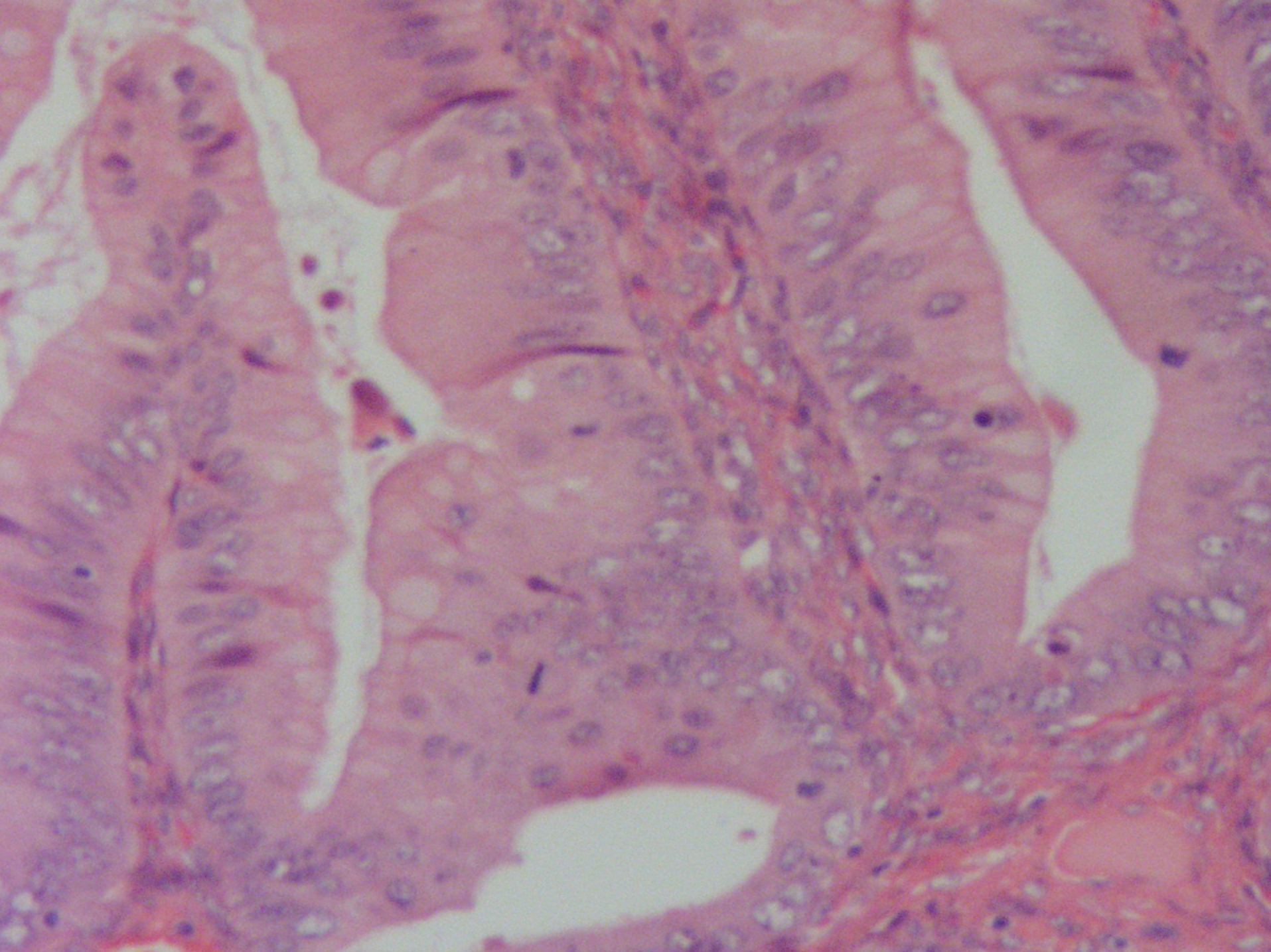
Age	Tumor
0-2 yrs	Teratoma (20% of all) Atypical teratoid/rhabdoid tumor Desmoplastic infantile astrocytoma/ganglioglioma
>2 yrs	Pilocytic astrocytoma Medulloblastoma Ependymoma

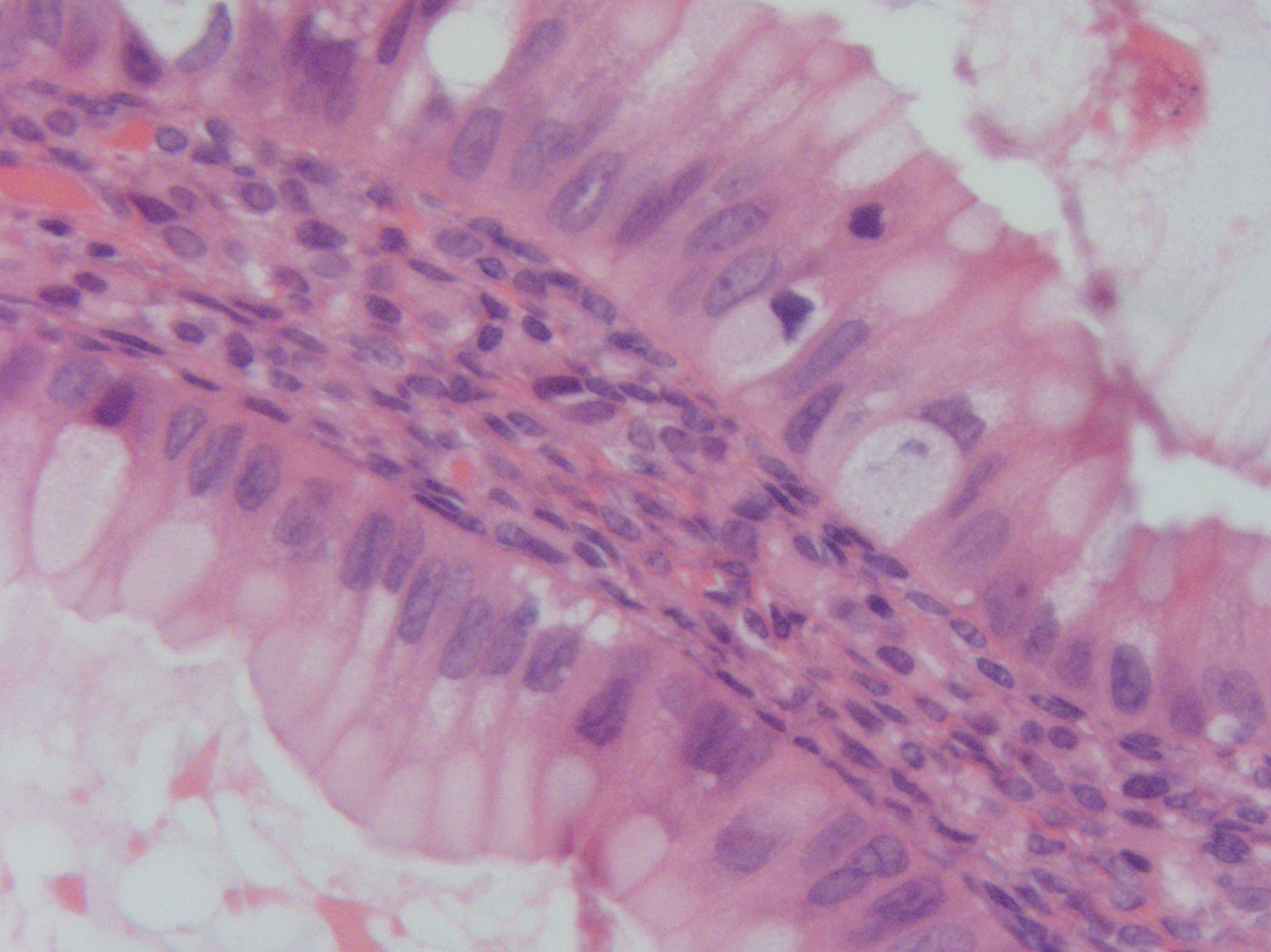
Case 32

- 88 F with diffuse abdominal pain and progressive nausea and vomiting
- 11 months s/p distal pancreactomy for Stg. I moderately differentiated adenoCA
- Exploratory laparotomy for lysis of adhesions and removal of right ovarian mass
- 140 gm 9x6x4.9 cm ovay with necrotic solid tumor
- Tumor cells + for CK7, CK20, Villin, CDX-2









Metastatic Pancreatic Adenocarcinoma

Ovarian Metastases

- 6-17% of all ovarian carcinomas
- Autopsy failed to reveal primary in 14%
- Laparotomy identified primary in 4/25 cases

Ovarian Metastases*

- Breast
- Colon
- Carcinoid
- Stomach

*Descending frequency

Favor Metastatic Carcinoma

- Surface implants
- Size <10 cm
- Multiple discrete tumor nodules
- Bilaterality
- Variation in growth pattern from each nodule
- Signet ring cells
- Vascular invasion
- Metastatic tumor may precede identification of extra-ovarian primary site by several years

Immunoperoxidase Not Helpful

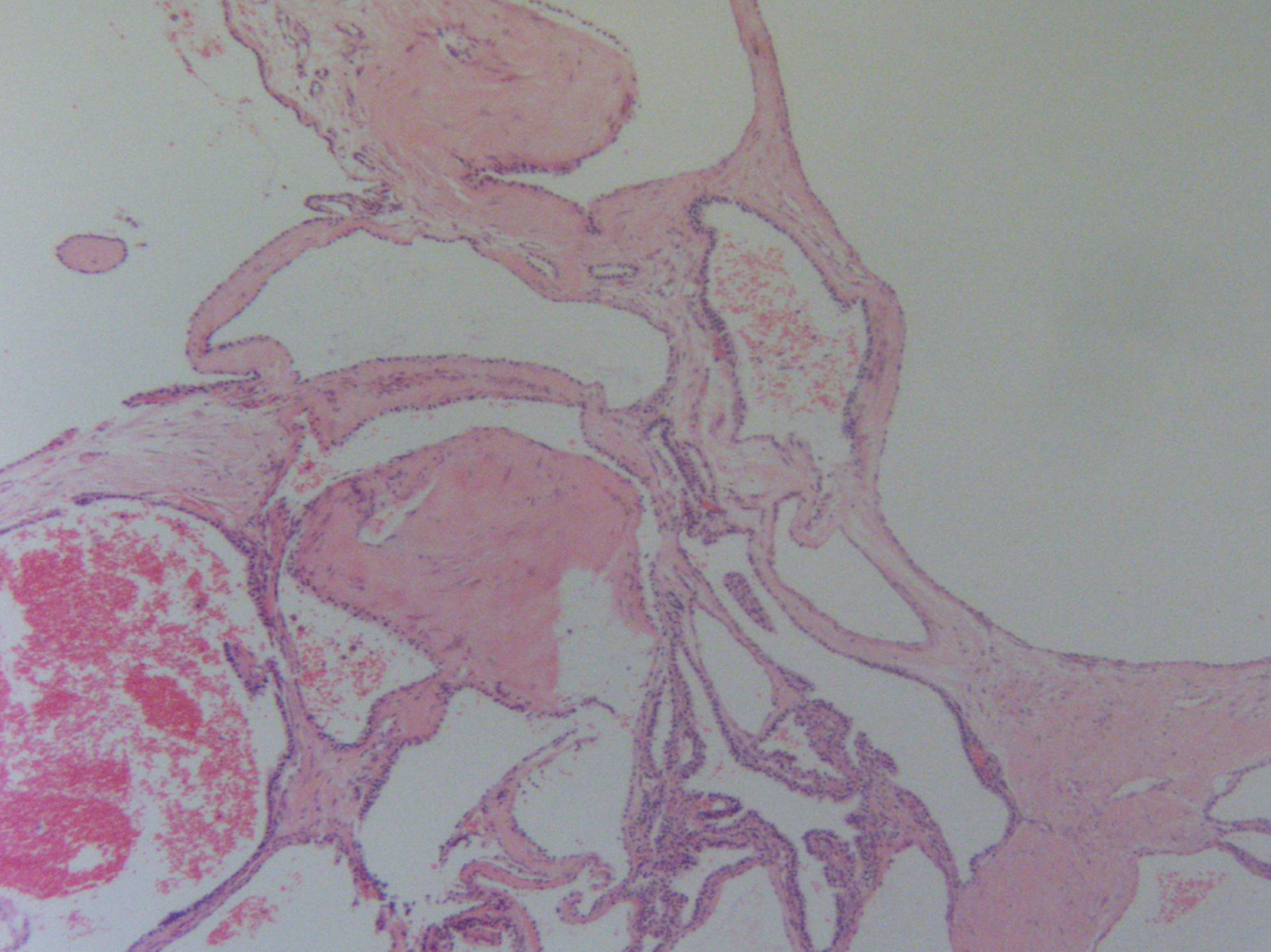
- Positive for both mucinous ovarian CA and metastatic pancreatic CA
  - CK7+
  - CK20+
  - CSX-2+
  - Villin+

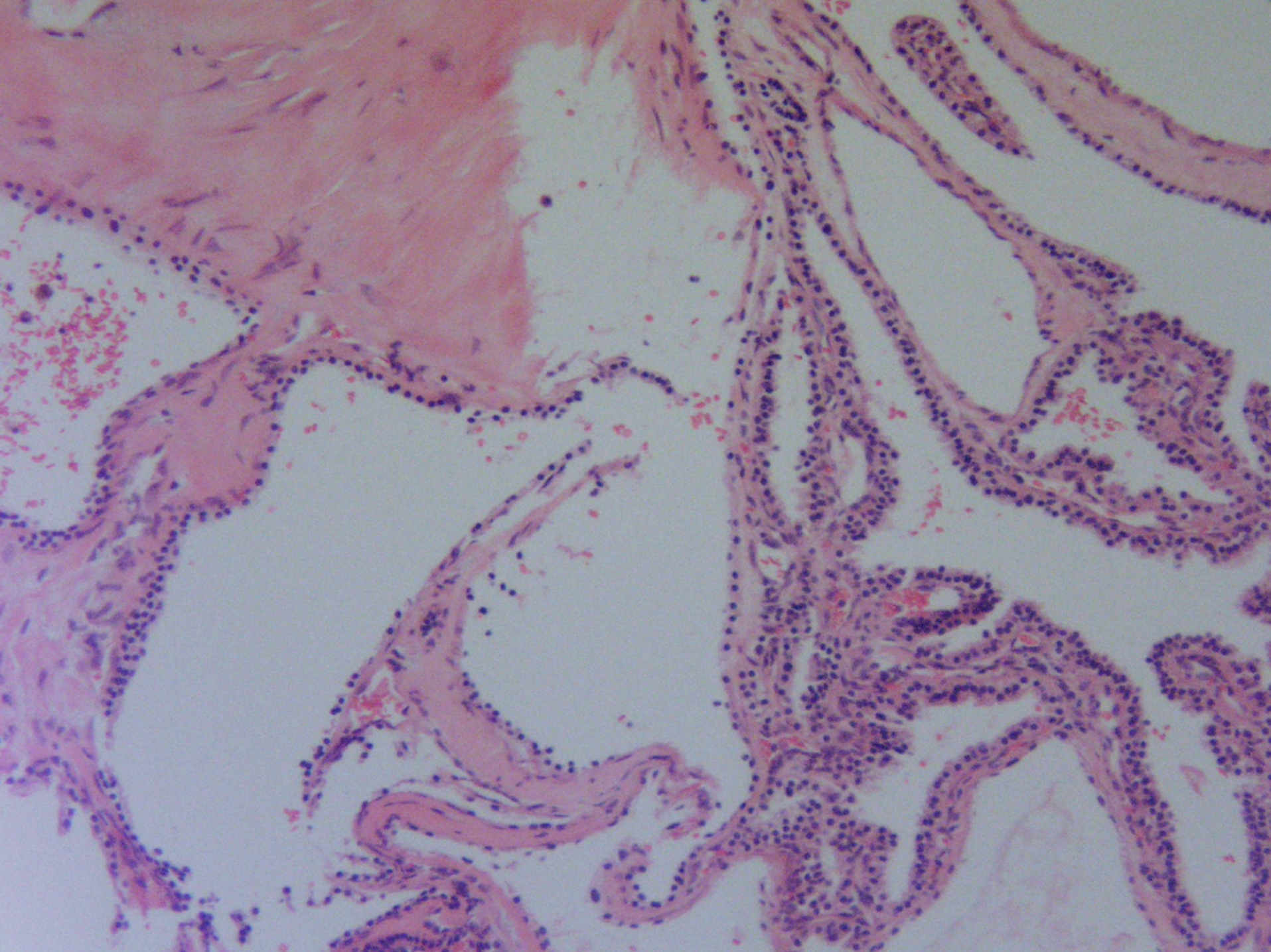
Mucinous Borderline Tumors

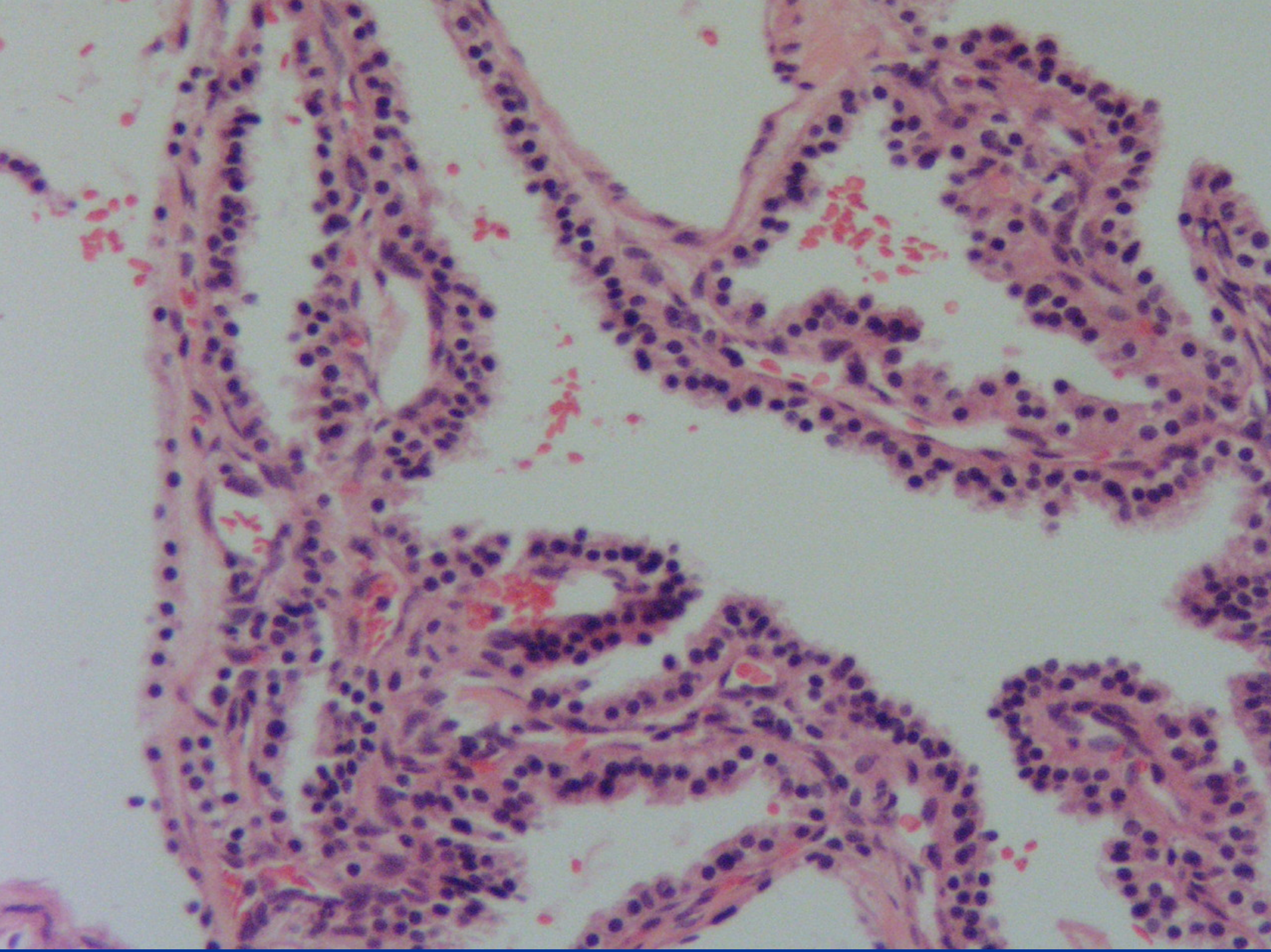
- May be indistinguishable from metastases
- Minimal atypia
- Overt features of invasion may be lacking

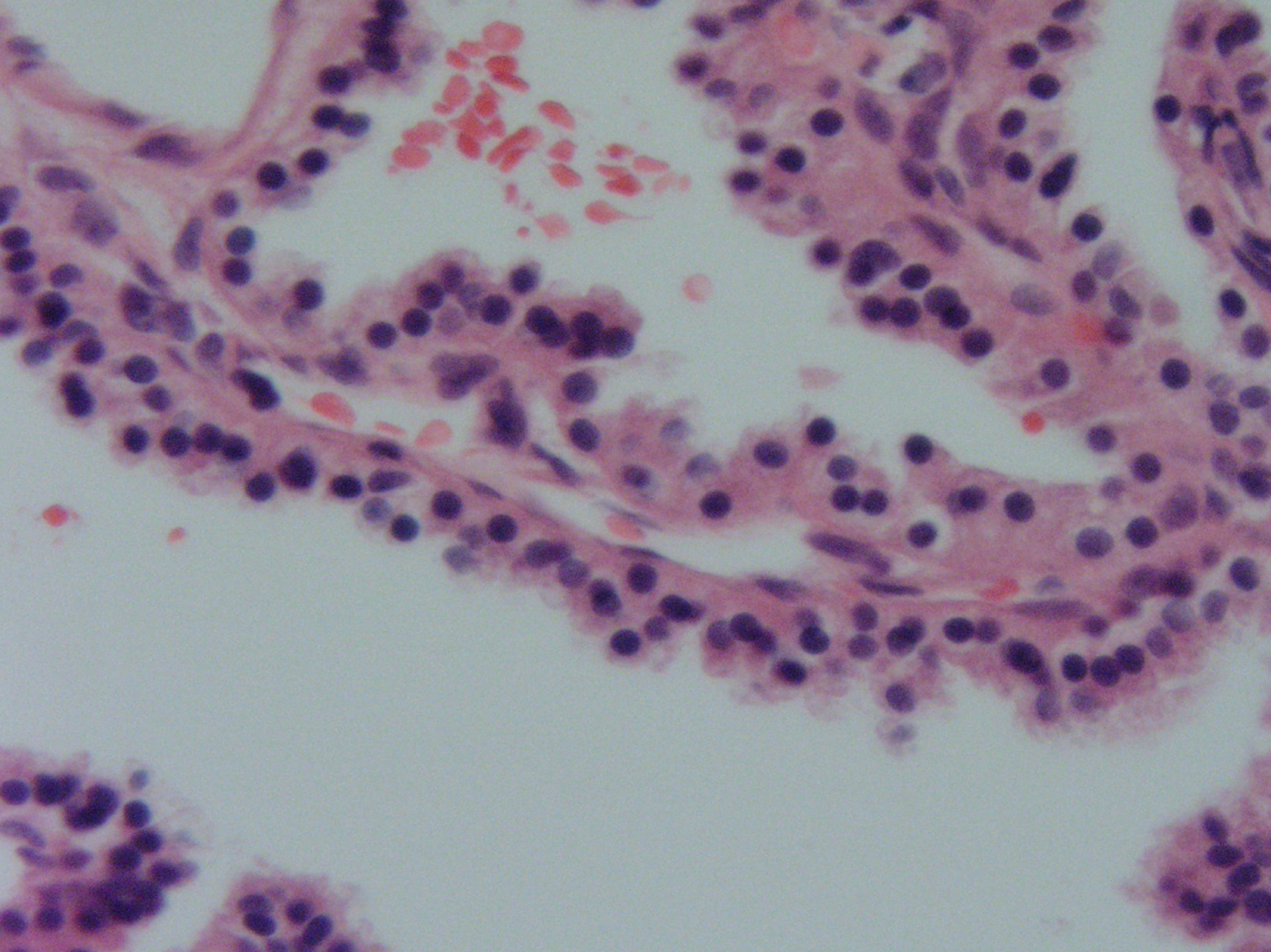
Case 33

- 65F with mild upper abdominal pain
- CT with large multicystic tumor in body and tail of pancreas
- FNA with light brown fluid of low viscosity with scattered polygonal cells with hyperchromatic nuclei- low levels of CEA and amylase
- Partial pancreatectomy with 25 cm tumor with thick white bosselated capsule
- Multicystic tumor with central radiating scar and cysts 1-2cm









Serous Microcystic Adenoma

Serous Microcystic Adenoma

- >60 yrs
- F>M
- Pancreas body and tail
- Multiloculated cyst, usually <2cm each
- Central sunburst scar and septal calcification
- Fluid low levels of CEA, contrast to mucinous
- Mutation of von Hippel-Lindau gene (3p)

Uncommon Variants

- Serous Oligocystic (Macrocystic) adenoma
  - VHL associated
  - Incompletely circumscribed
- Solid Serous Adenoma
  - Hyaline septa separate solid sheets of clear cells

DDX

- Mucinous cystic tumors
- Neuroendocrine tumors
- Solid-pseudopapillary tumor
- Lymphangioma
- Metastatic renal cell CA

Metastatic Renal Cell CA-Clear Cell Type

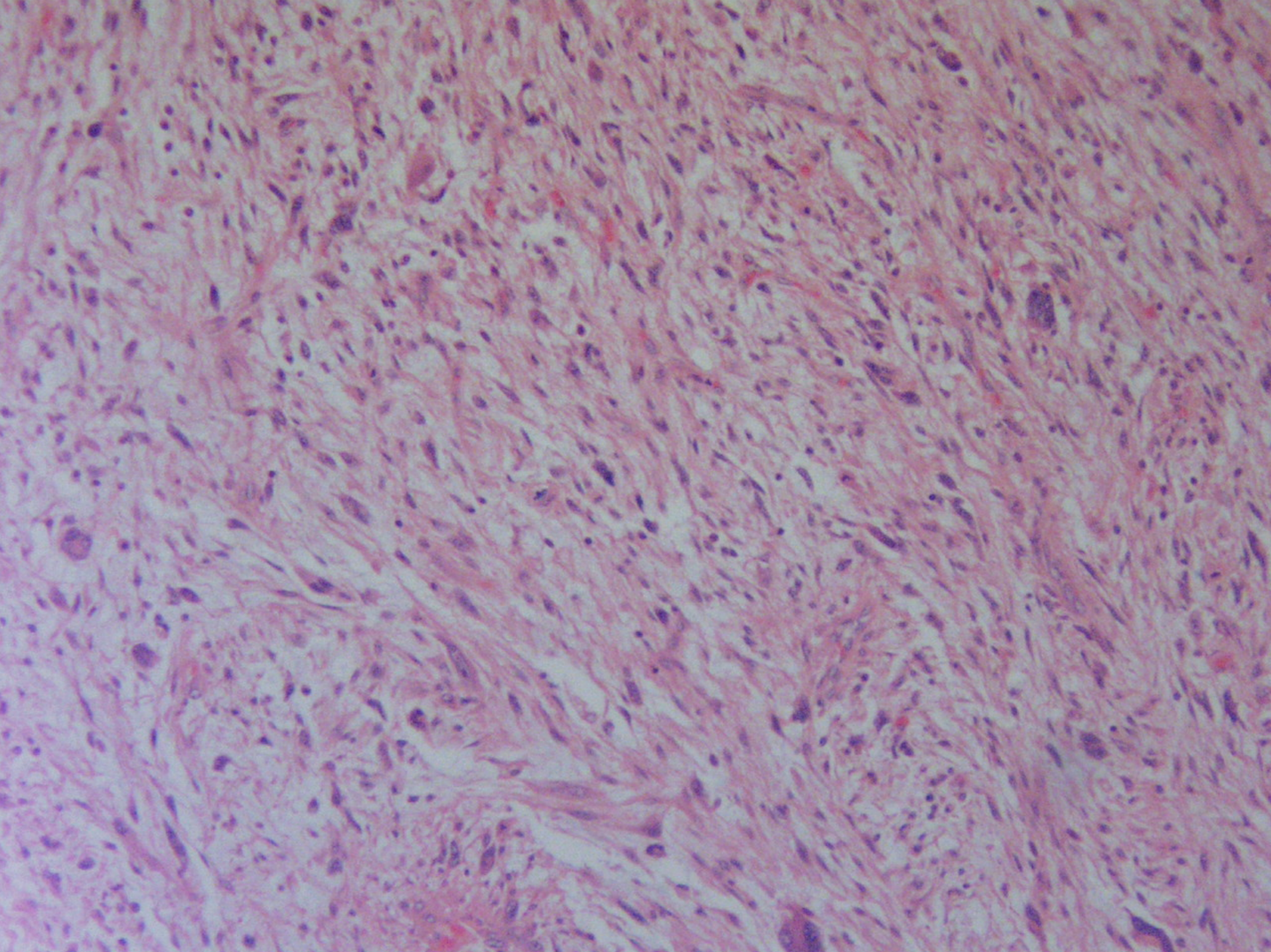
- Intracellular lipid
- PAS+
- PAS D-
- Chromogranin A-
- EMA+
- CEA-

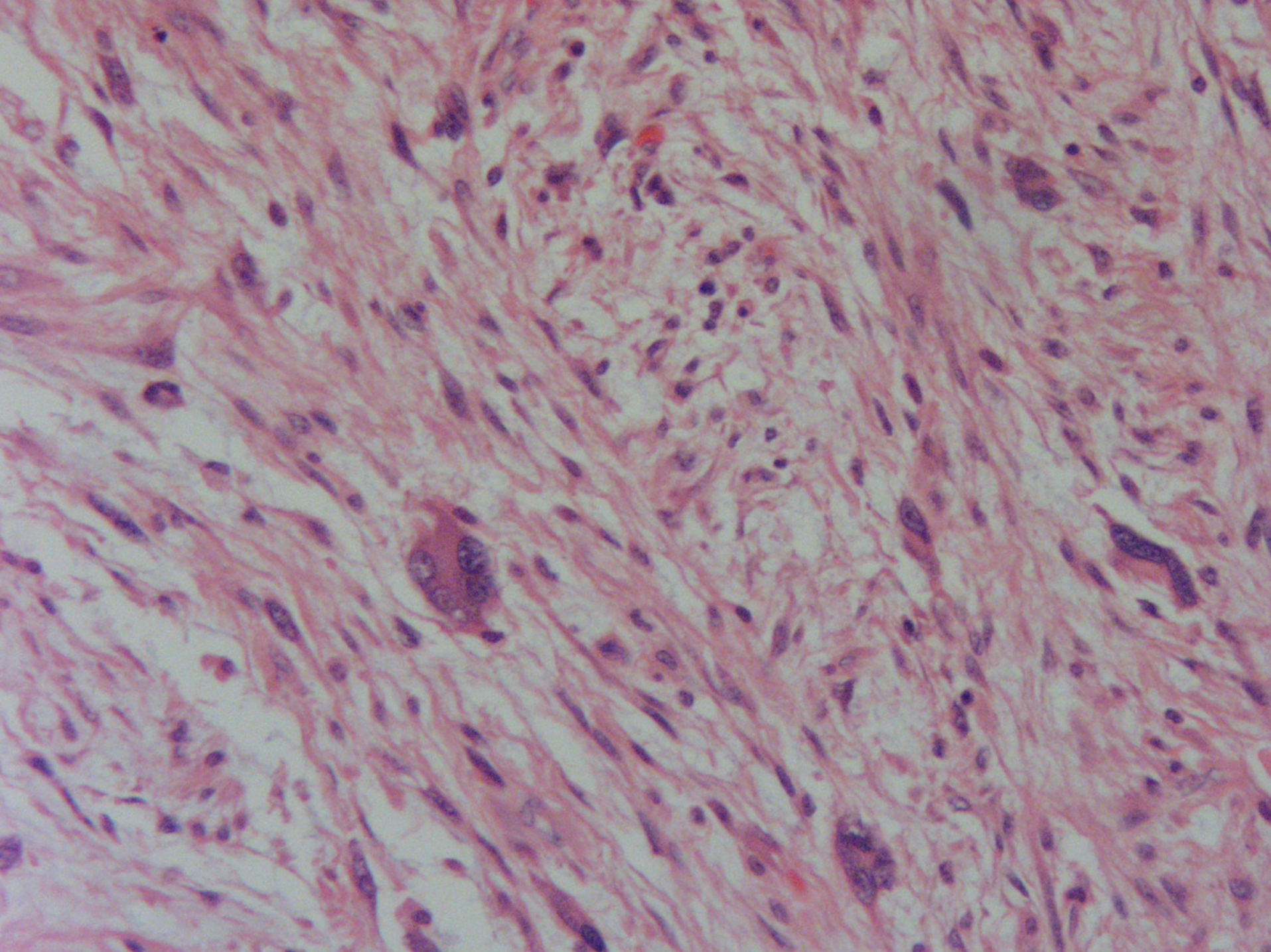
Solid-Pseudopapillary Neoplasm

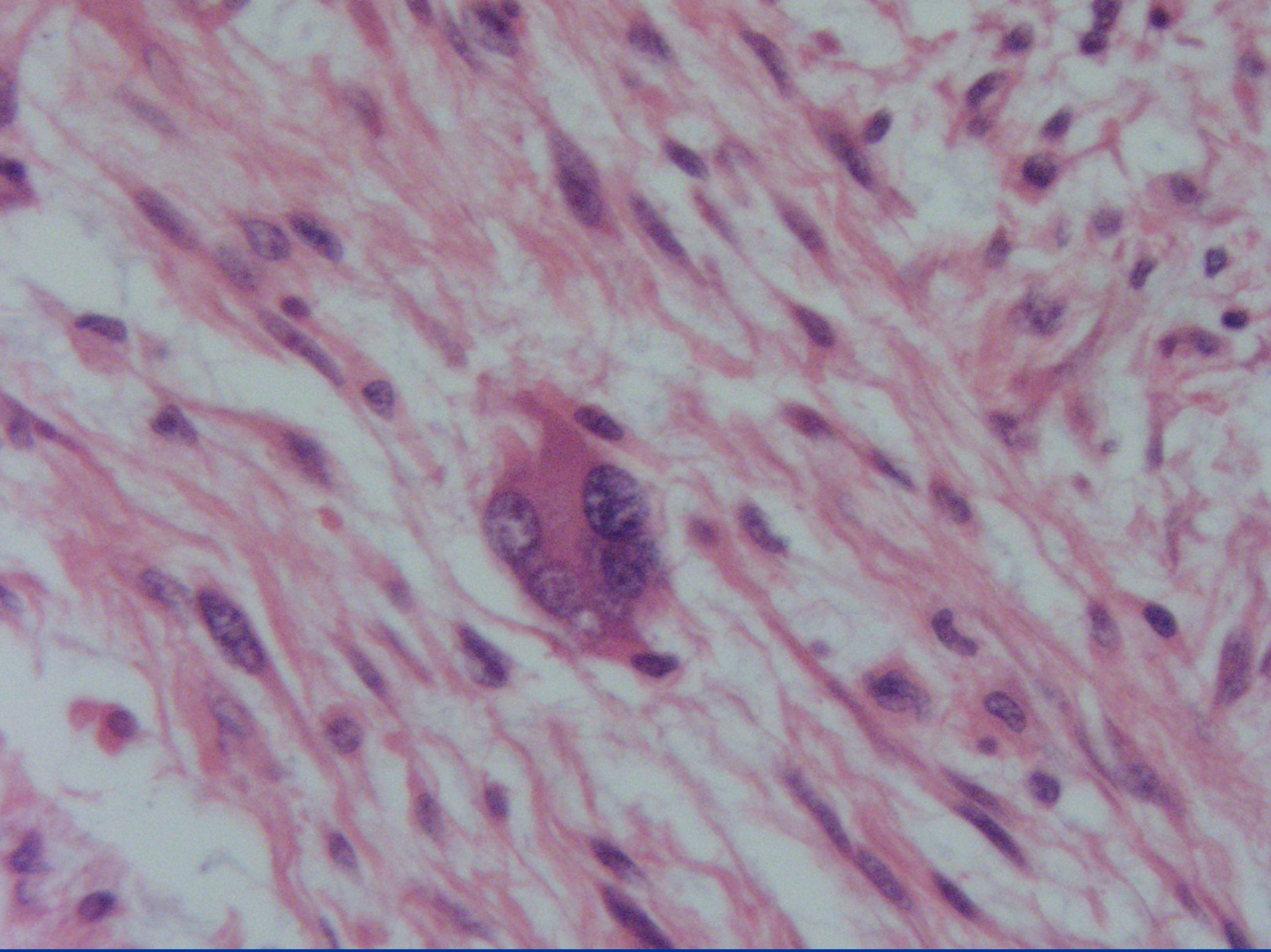
- Adolescent women
- Low grade malignancy
- A1AT+
- Progesterone Receptor+

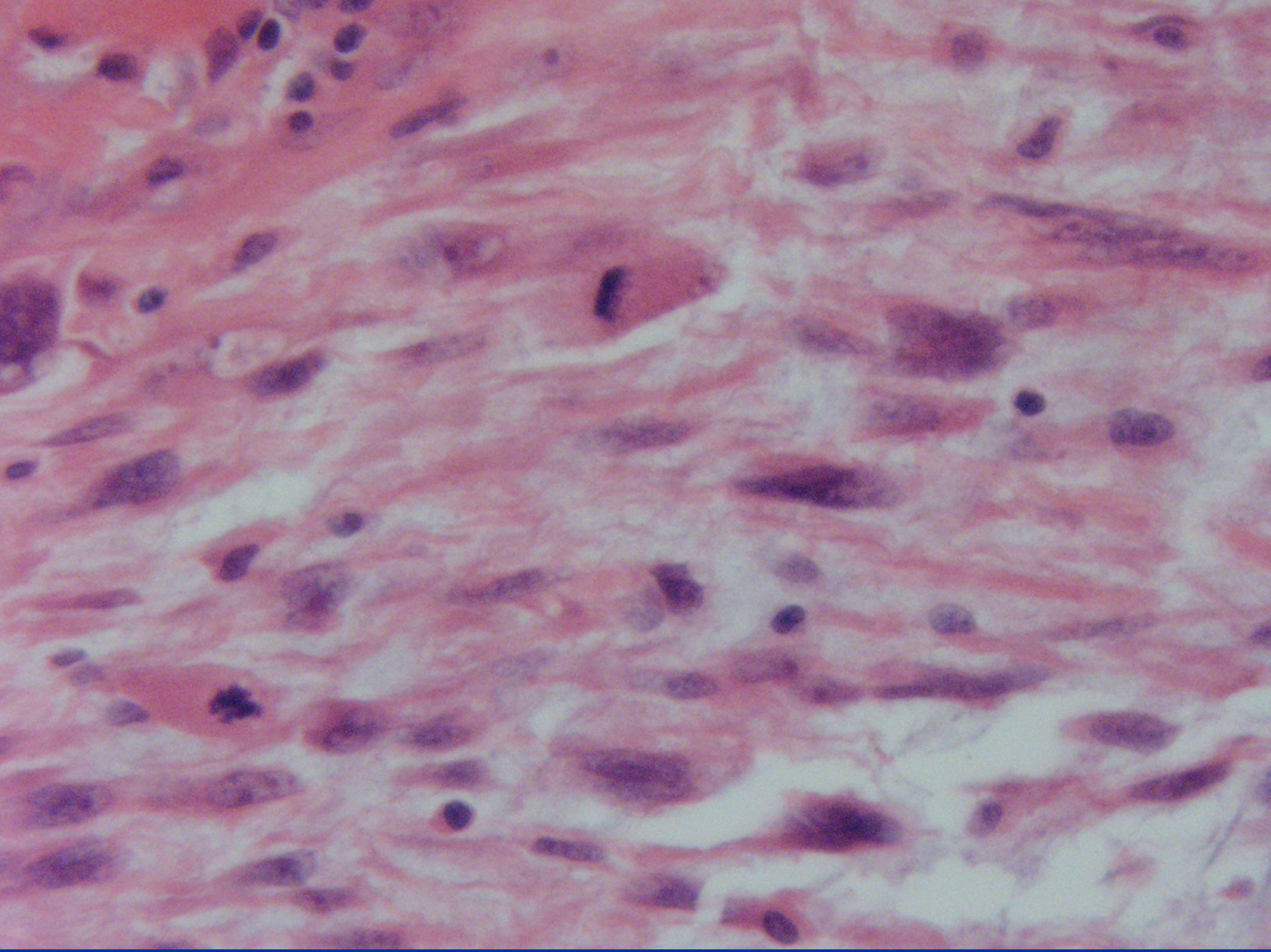
Case 34

- 46F with lower abdominal pain, pelvic mass, menometrorrhagia
- Hysterectomy with solitary, poorly circumscribed 8cm mass
- Soft, variegated, gray yellow tumor with necrosis









Leiomyosarcoma

Leiomyosarcoma

- 1% malignant uterine corpus tumors
- Postmenopausal (average 52 yrs)
- Unrelated to endometrial CA risk factors
- Signs and symptoms similar to leiomyoma
- Usually solitary
- 2/3 intramural
- Range 6-9 cm

Prognosis

- Intraperitoneal spread to LN, lung, liver, bone, kidney
- Recurrence
  - 57% hematogenous
  - 20% pelvic

Necrosis

Necrosis	Defn.	LM	LMS
Hyaline (Infarctive)	Necrosis separated from viable tumor by zone of connective tissue	+++	+/-
Tumor cell	Abrupt transition viable cells to necrotic cells without interposed zone	Rare	+++
Ulcerative	Ulcerated surface of tumor	+++	+++

LM vs LMS

Tumor Cell Necrosis	Atypia	MF/10 hpf	DX
Present	Diffuse moderate to severe	Any level	LMS
Present	None to mild	>10	LMS
Present	None to mild	<10	LMS (r/o LM infarct)

LM vs LMS

Tumor Cell Necrosis	Atypia	MF/10 hpf	DX
Absent	Diffuse moderate to severe	>10	LMS
Absent	Diffuse moderate to severe	<10	Atypical LM with LRR
Absent	None to mild	<10	LM
Absent	None to mild	>10	LM-mitotically active
Absent	Focal moderate to severe	>10	LM with limited experience
Absent	Focal moderate to severe	>15	STUMP

Adenosarcoma

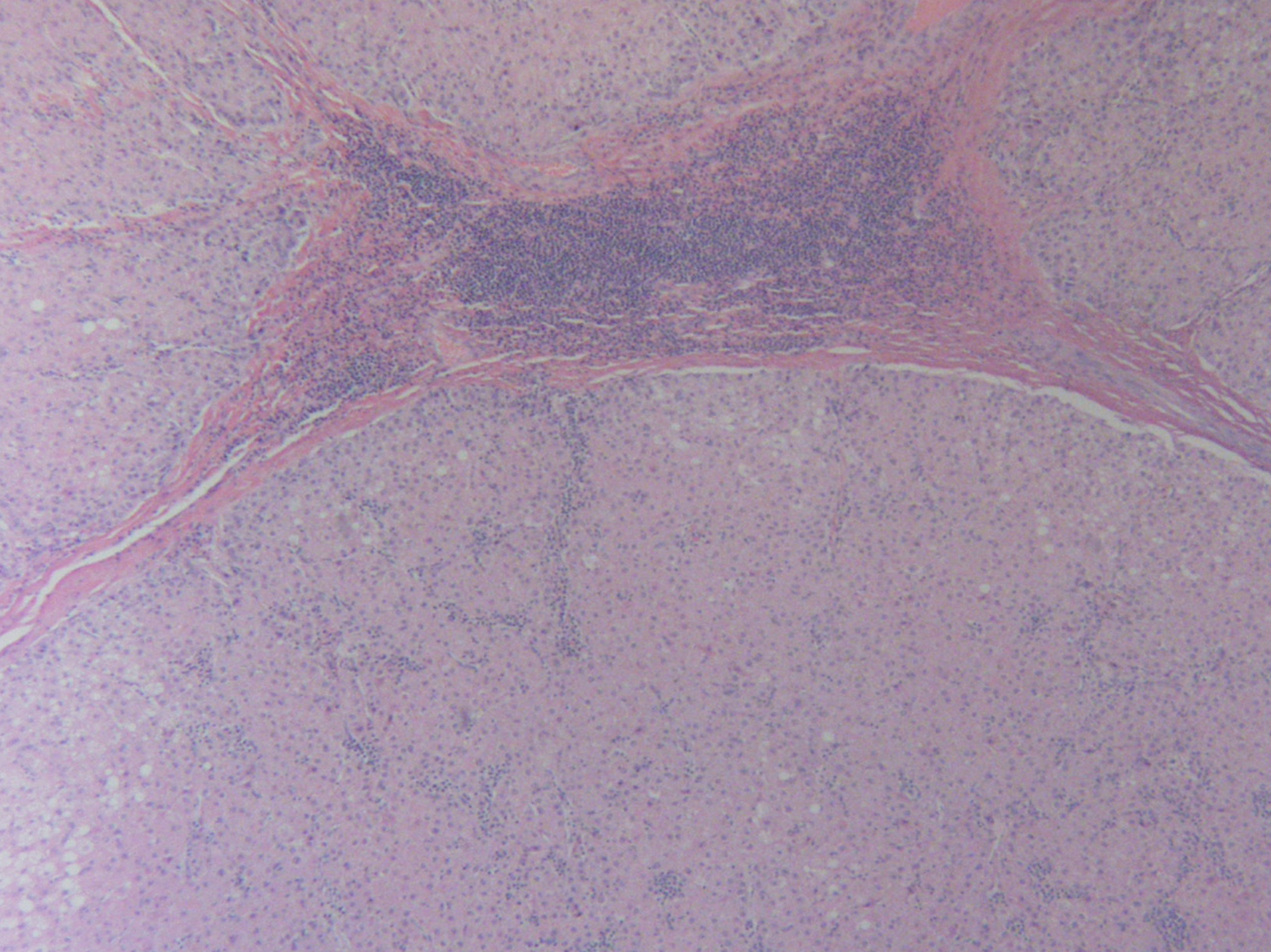
- Biphasic neoplasm
- Recurrence 25-40%
- Metastasis rare <5%

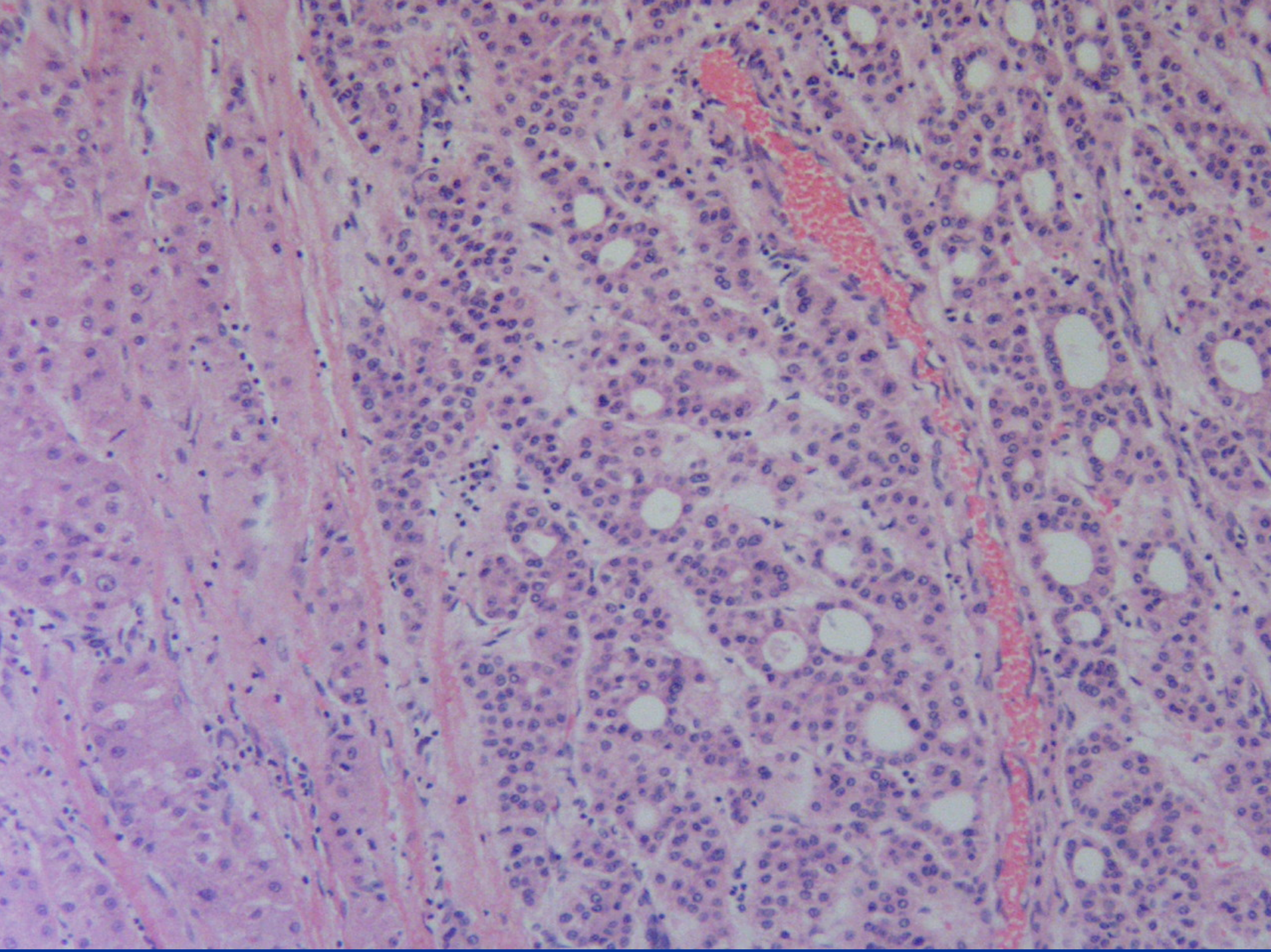
Atypical Polypoid Adenomyoma

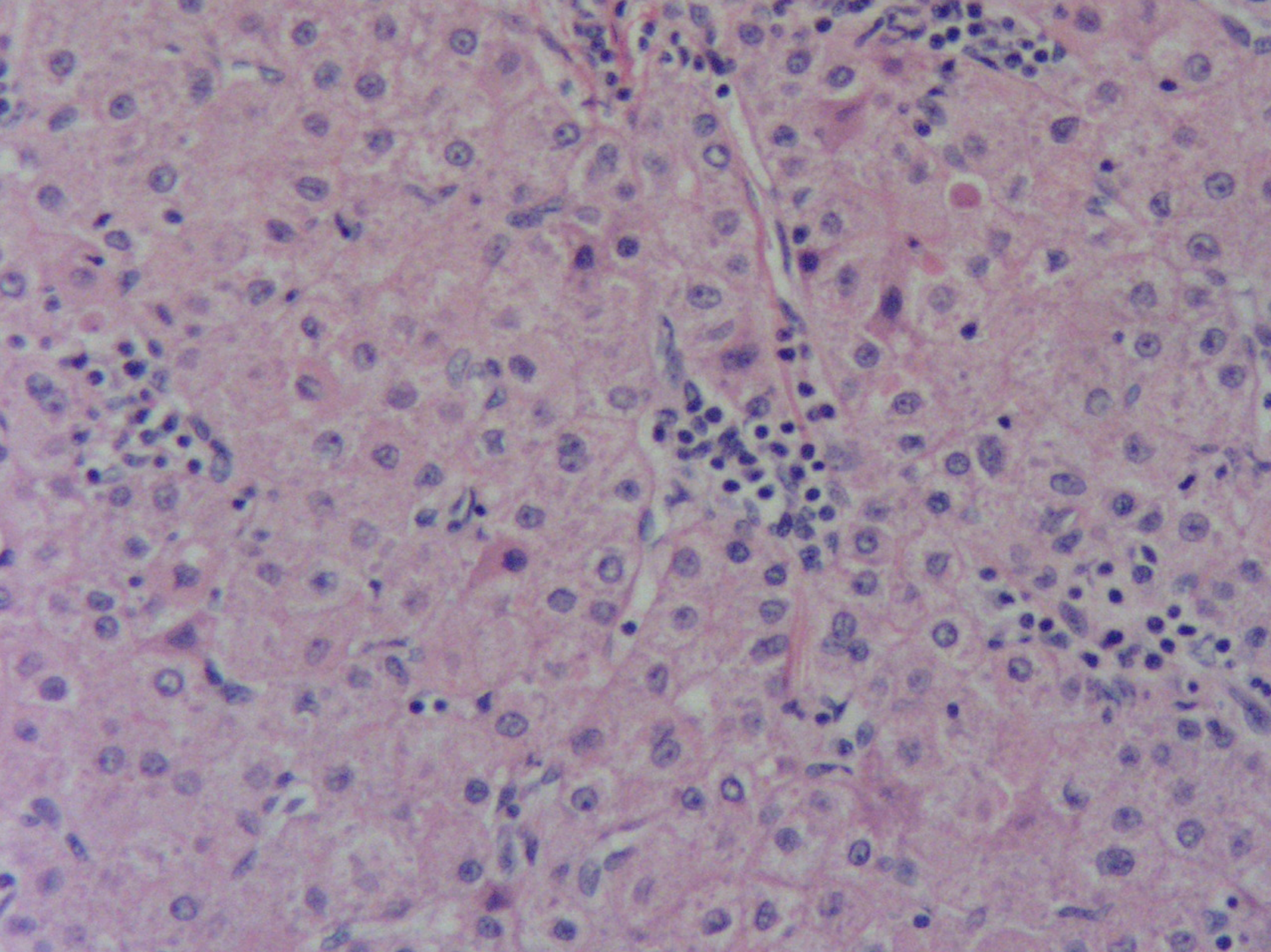
- Benign
- Usually diagnosed on EMC
- Mimics endometrioid adenocarcinoma with myometrial invasion

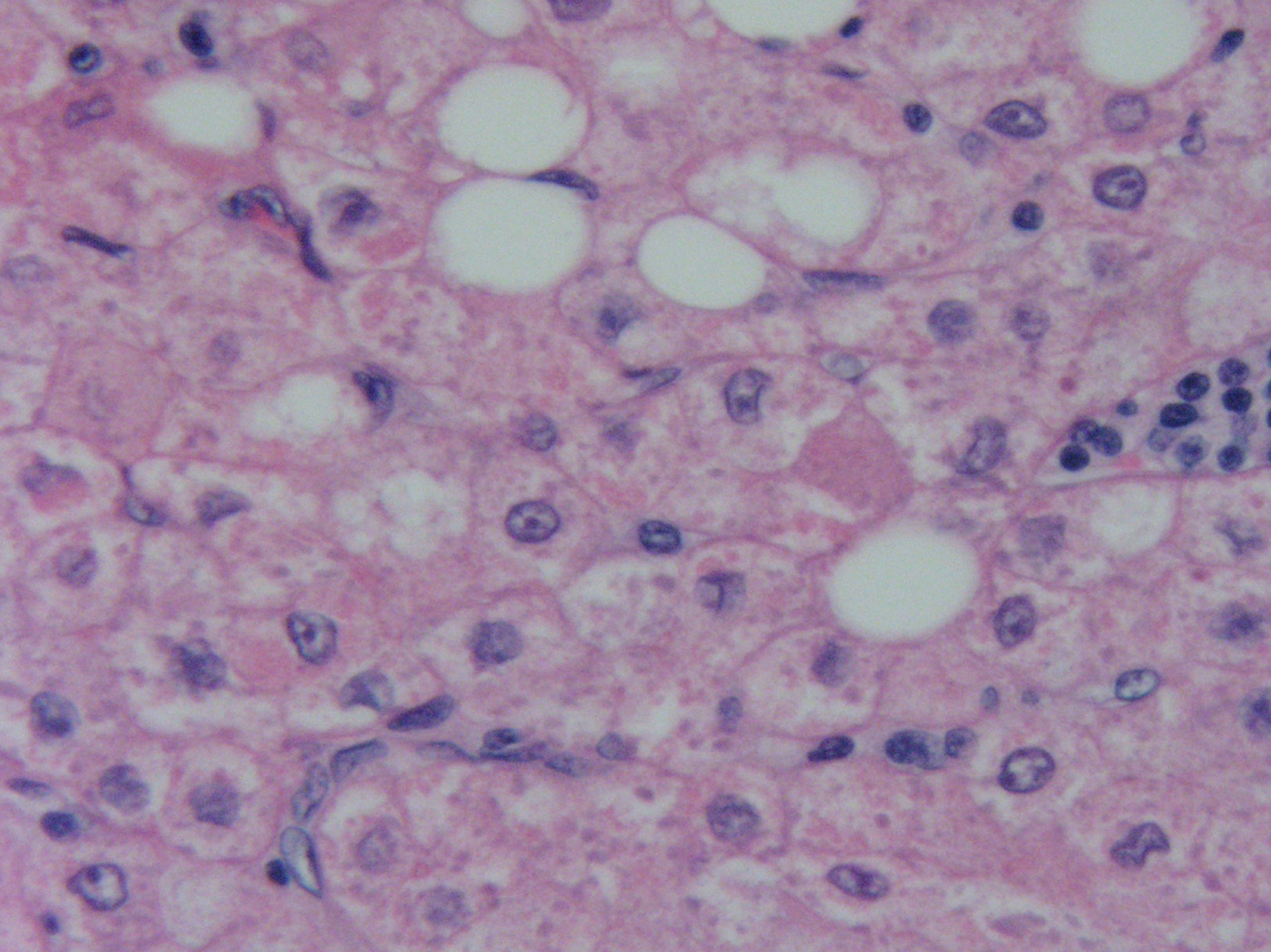
Case 35

- 67M with abdominal pain
- Mass in right lobe of liver
- Partial hepatectomy with 7cm well circumscribed nodule, tan, focally hemorrhagic









Hepatocellular Carcinoma

	Incidence	Age Decade	Sex M:F
China	80/100,1000	4-5th	8:1
USA	5/100,000	7-8th	2:1

Tumor	Hepatocytes	Other
HCC	Uniform >3 layers	Acinar growth occ.
Adenoma	Uniform <3 layers	No bile ducts
FNH	Nodular aggregates with thin fibrous septae	
Fibrolamellar HCC	Large polygonal cells with eos. granular cytoplasm	Lamellar fibrosis

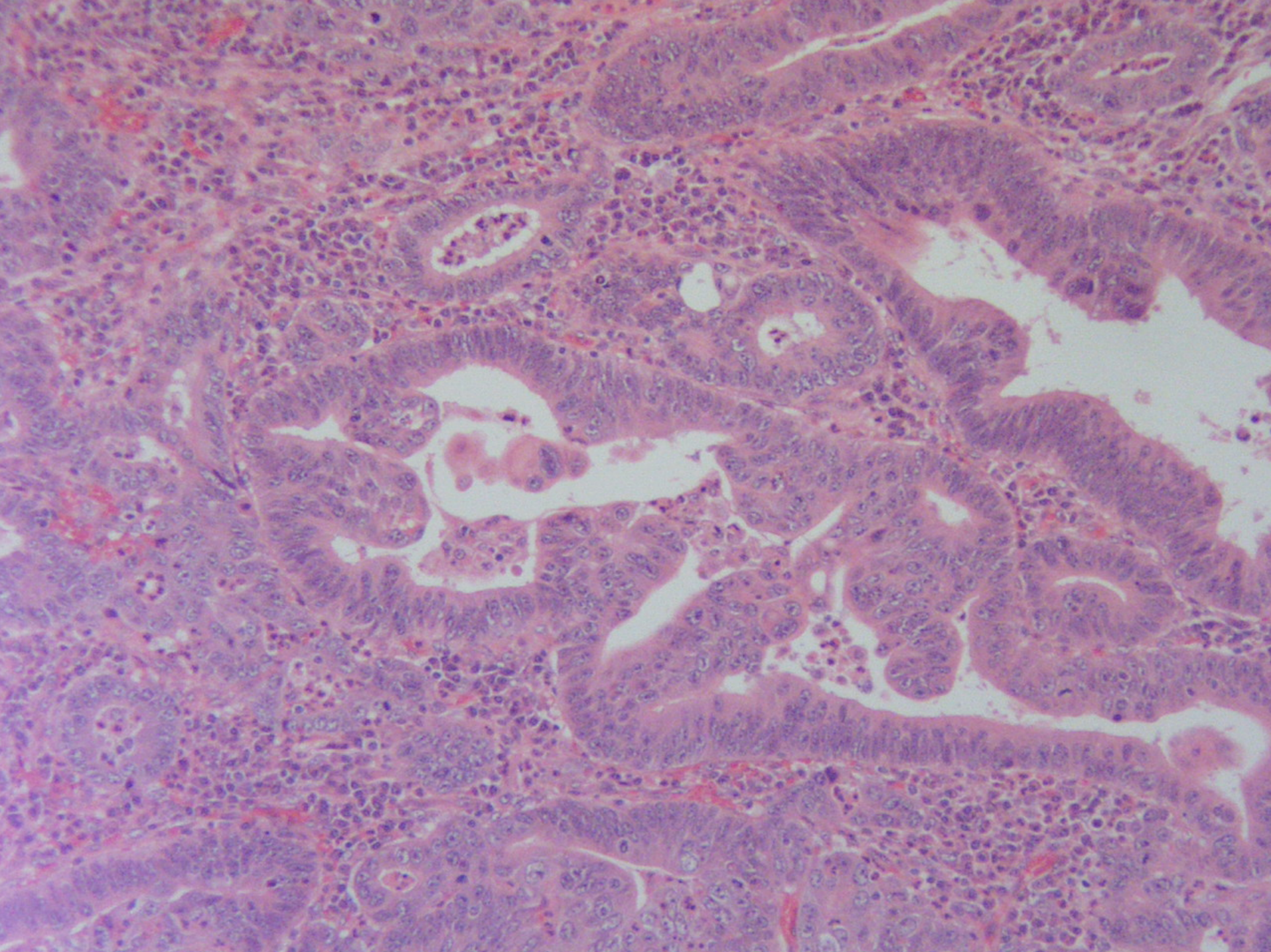
Quick Facts

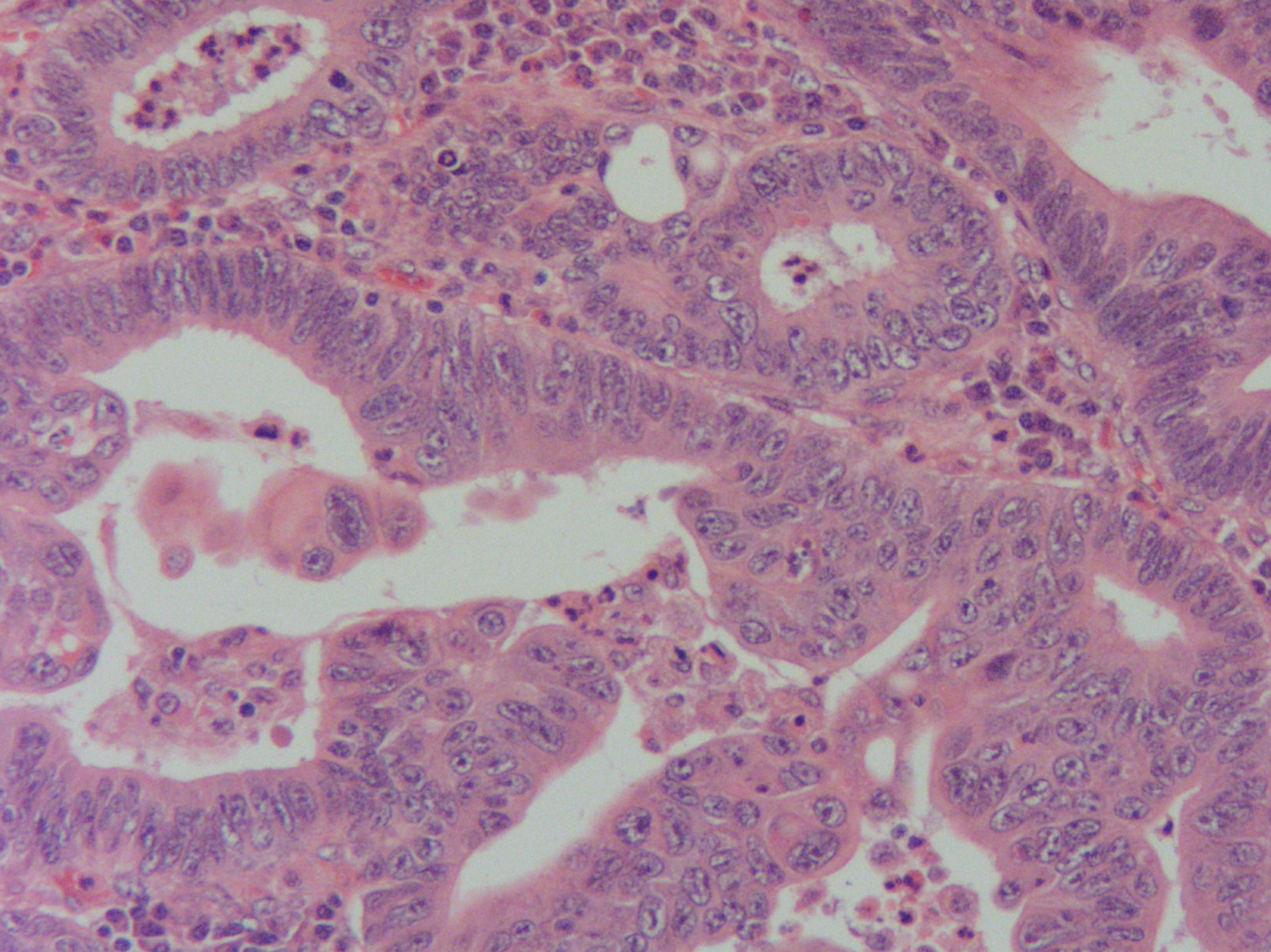
- Hepatocellular adenoma associated with steroid use

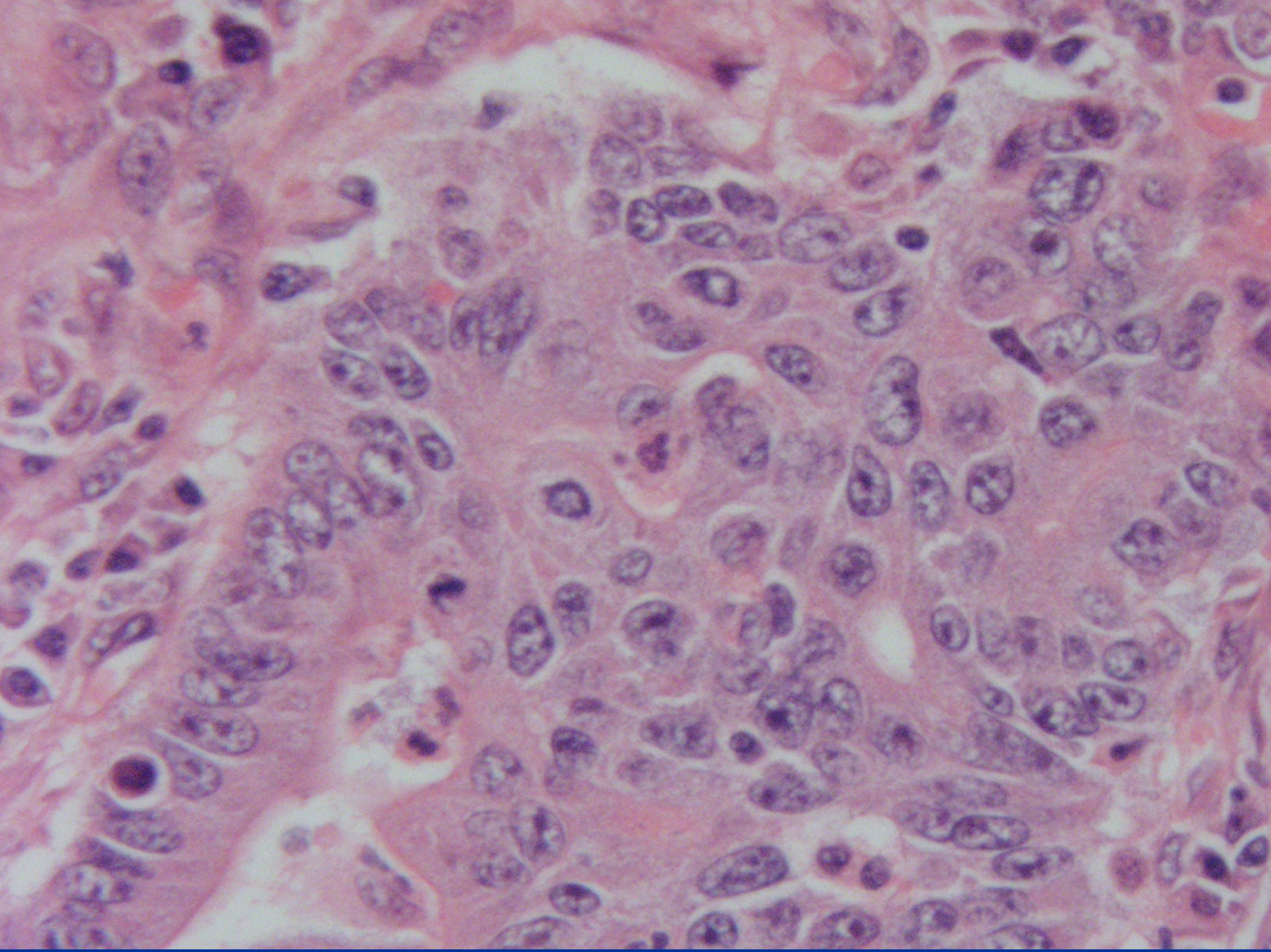
- Fibrolamellar hepatocellular carcinoma usually in non-cirrhotic liver young adult

Case 36

- 62M with 30lb weight loss and early satiety for 5 months
- Ulcerated mass in distal lesser curvature
- 5cm elevated, ulcerated mass in antrum with grossly involved lymph node







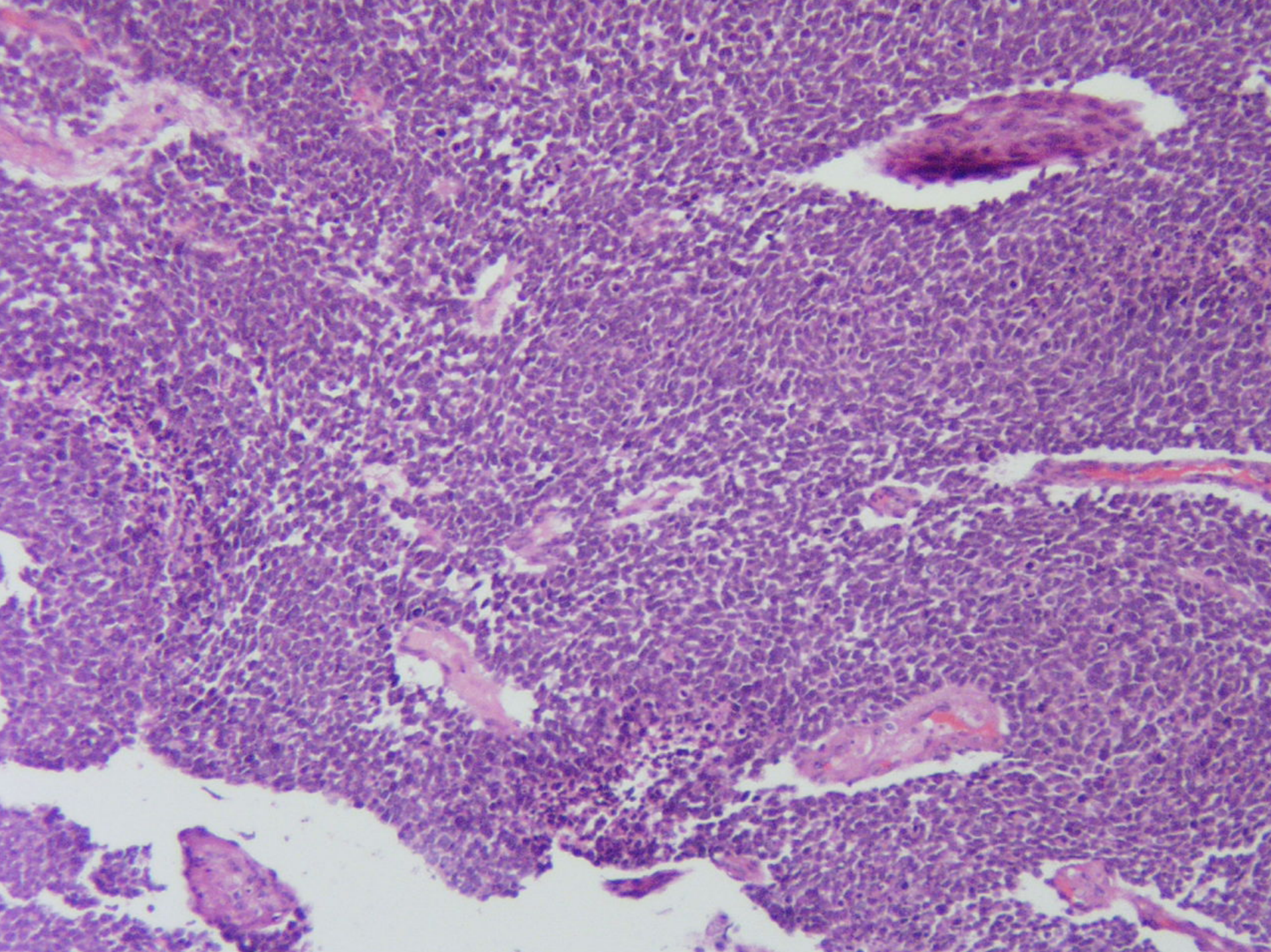
Adenocarcinoma, Intestinal Type

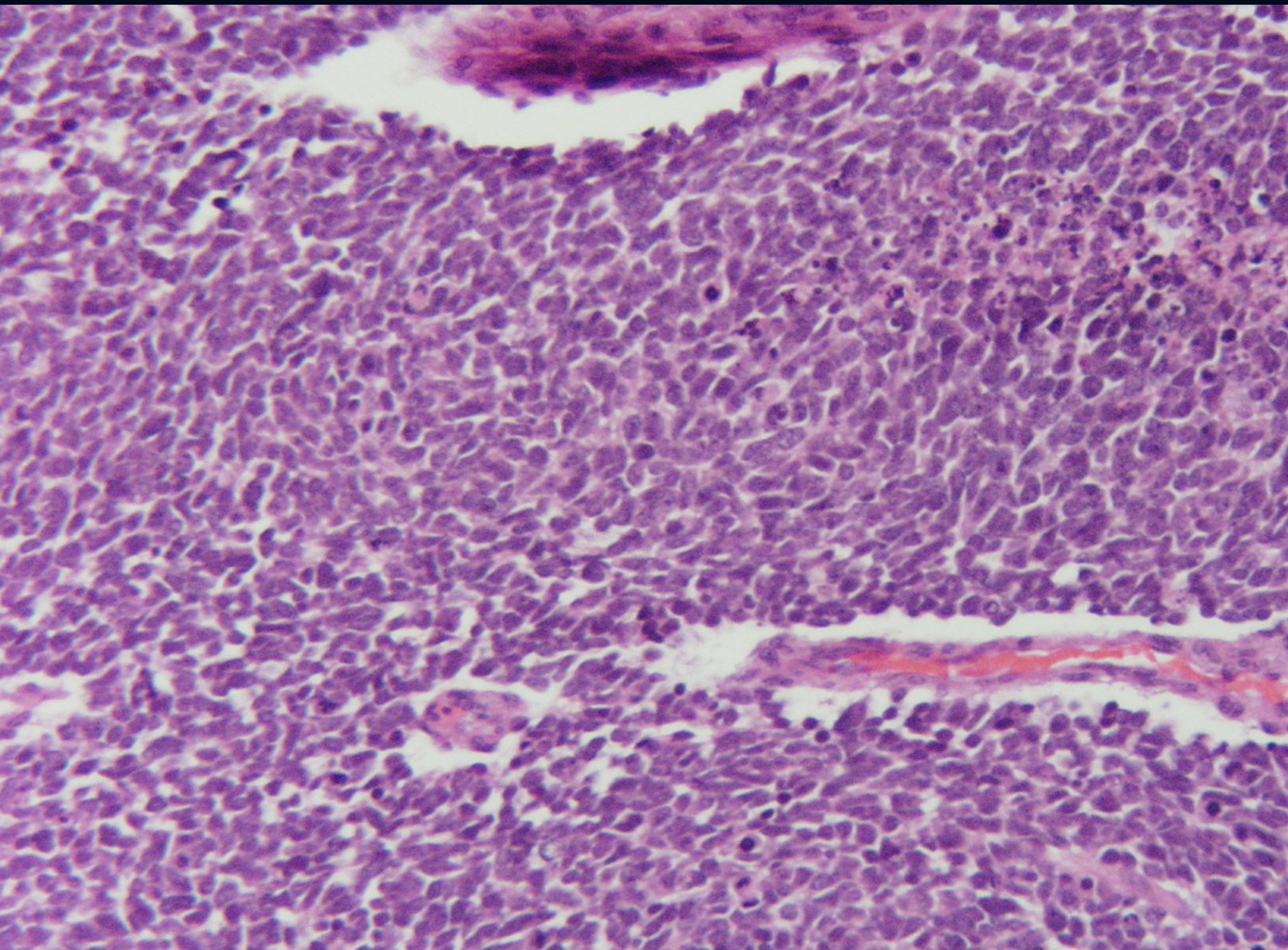
Quick Facts

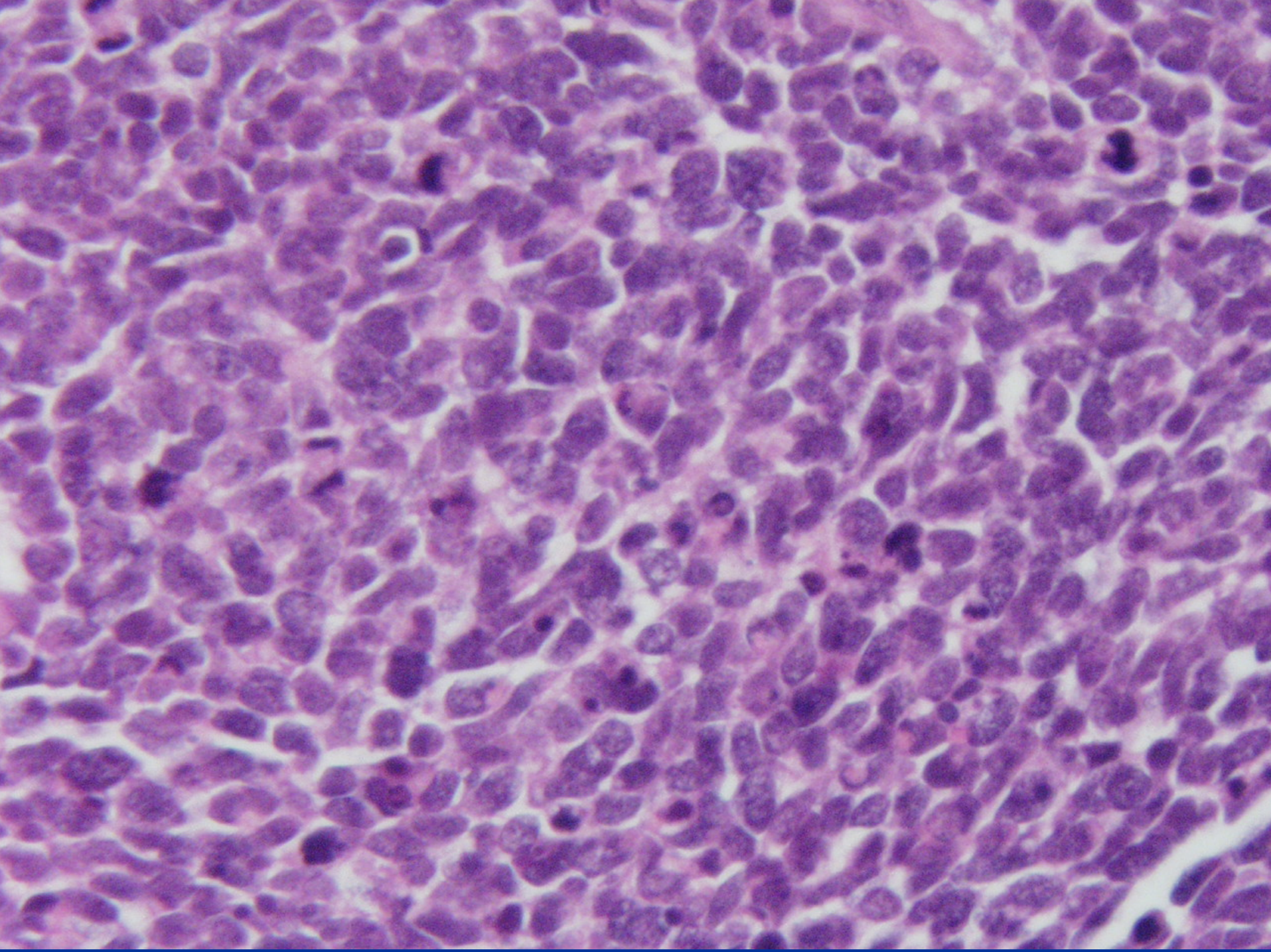
- Carcinoid tumors are indolent when arising in autoimmune gastritis vs. sporadic
- Adenocarcinoma, diffuse type, may be familial due to germline E-cadherin mutations
- Adenocarcinoma, intestinal type, most strongly linked to H. pylori

Case 37

- 2yo boy with palpable right sided abdominal mass
- 13cm tumor in upper pole of right kidney with variegated, focally necrotic cut surface







Wilm's Tumor
Blastema predominant

Nephroblastoma

- Most common GU childhood tumor
- 1-3 yrs
- Stage and histology most important prognostic features
- Anaplasia is unfavorable histology

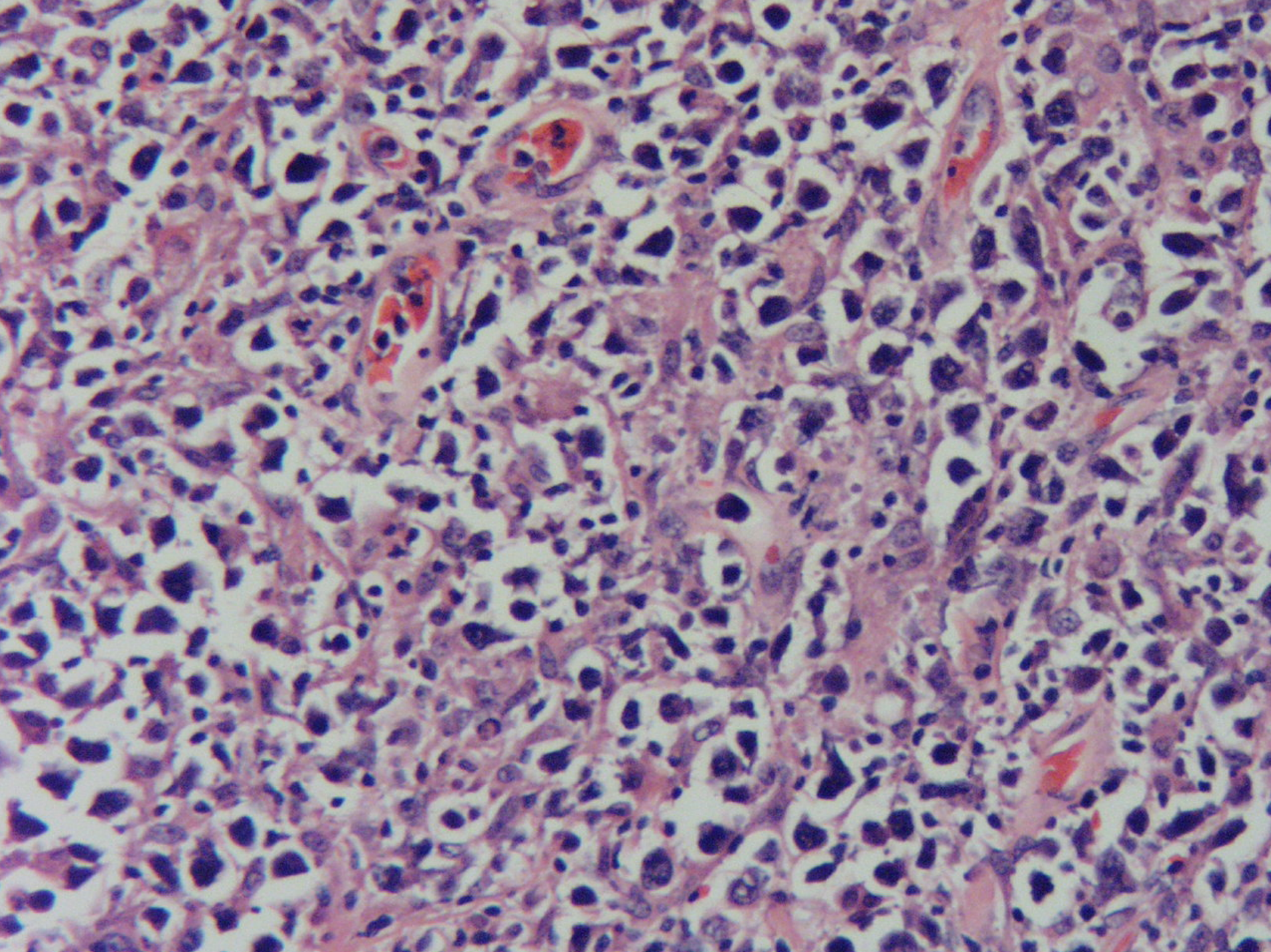
Histopathology

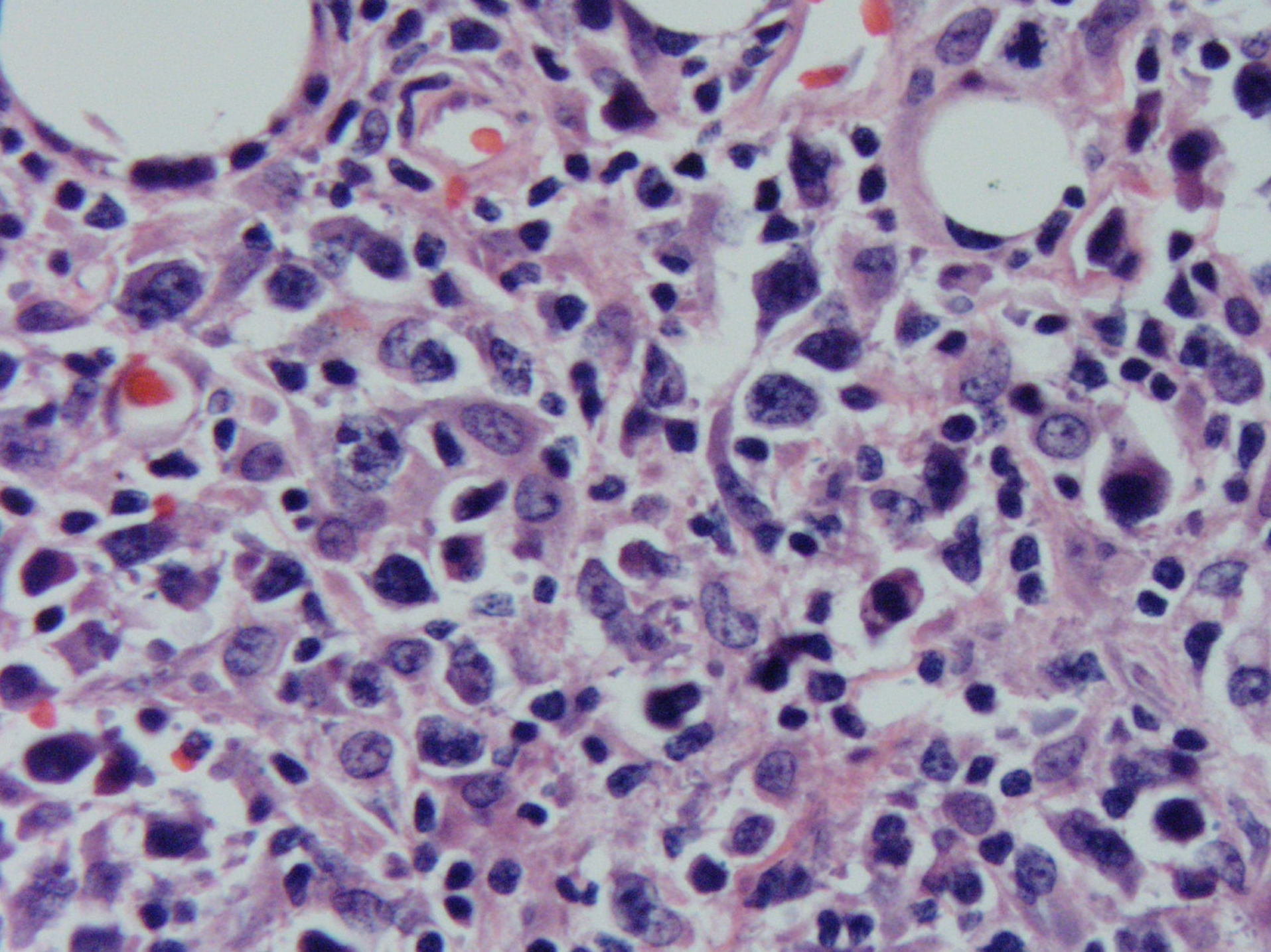
- Triphasic epithelial, stromal, blastemal
  - Blastemal predominance if >33%
- VIM+, CK+/-
- MIC2 negative

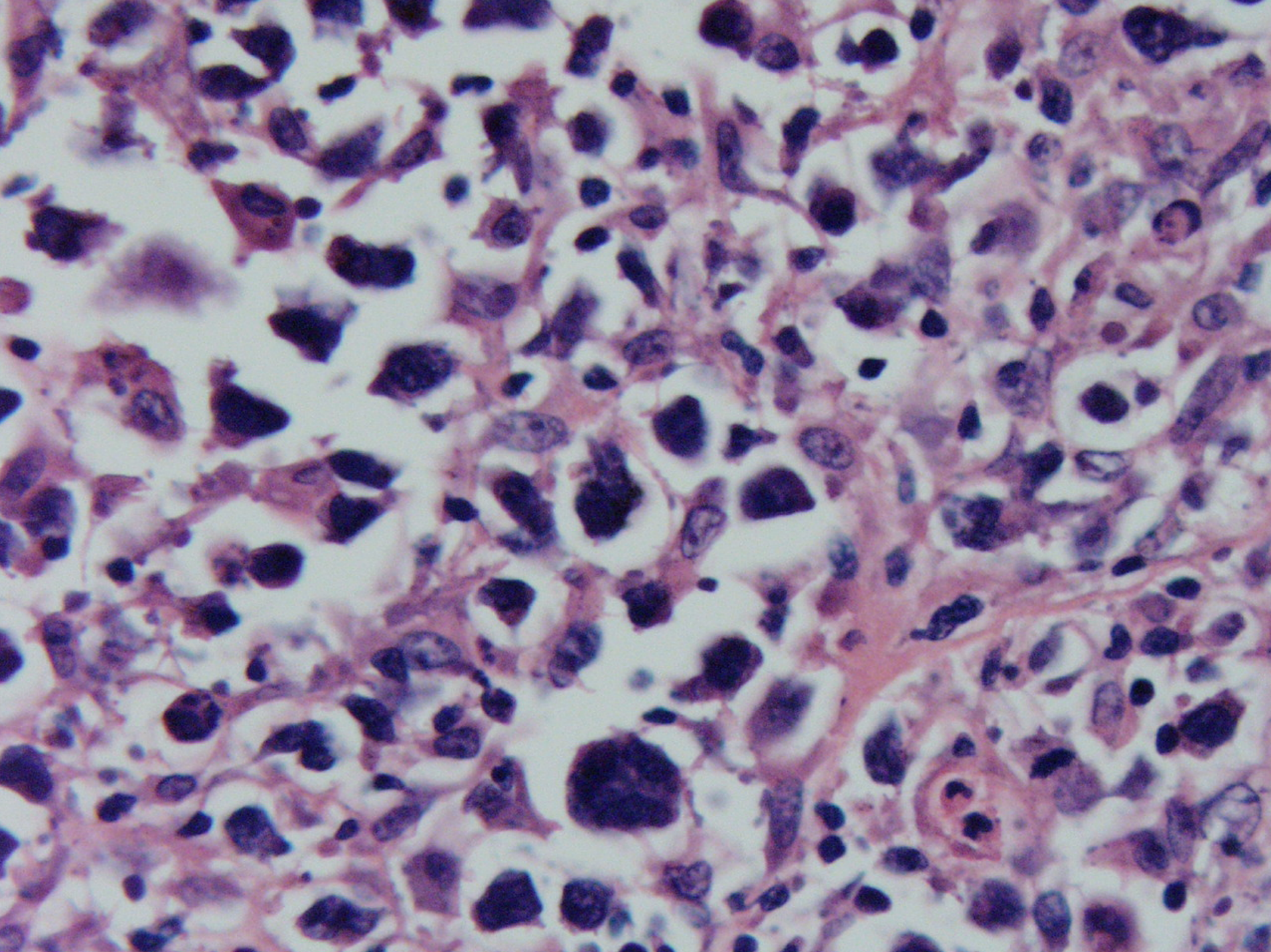
Tumor	Histology	Other
CCSK	Round to spindled nuclei, optically clear cytoplasm, stroma with prominent arborizing capillaries	Mets to bone
MRTK	Prominent eosinophilic cytoplasm with eccentric nuclei	EMA and desmin+ Central PNET association
CMN	Uniform spindle cells with macro/microcysts	t(12;15)

Case 38

- 69F s/p grade 3 endometrioid endometrial adenocarcinoma, presents with abdominal pain
- Right adrenal mass by CT scan
- 3 cm tan nodule in adrenal gland
- Tumor
  - + CK20 and CD45
  - - EMA, pankeratin, S100, MART-1, calretinin, synaptophysin







Diffuse Large B-cell Lymphoma (Anaplastic variant)

Diffuse Large B-cell Lymphoma (Anaplastic variant)

- 30-40% of adult NHL
- 6-7th decades

Immunoperoxidase

- Pan B cell markers
- Surface Ig 75%
- Bcl-2 +
- Ki-67+ but usually <99%

Mind Your Ts and Bs

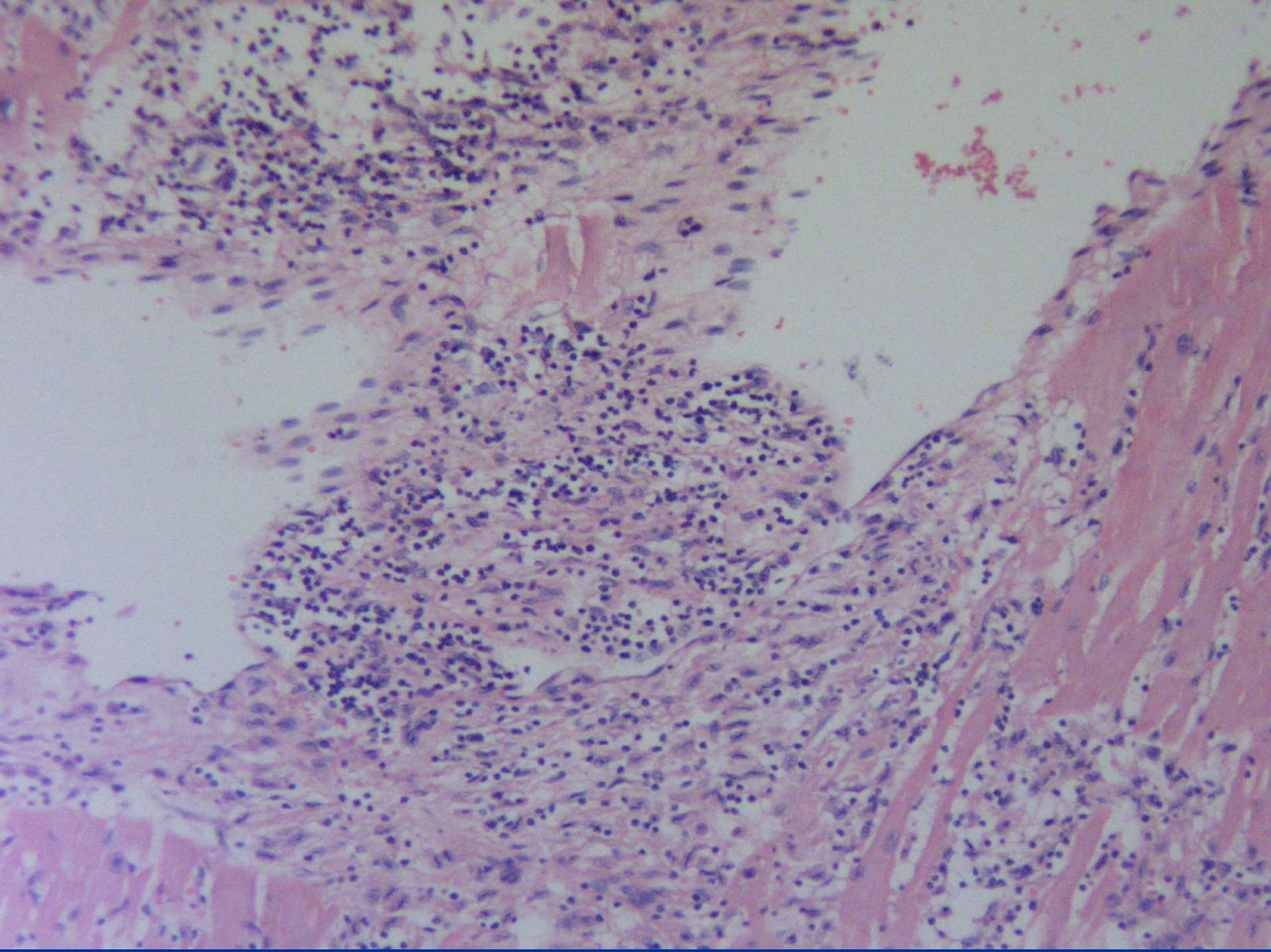
- B cell lymphomas with CD30 are not anaplastic large cell lymphomas
- Distinction from anaplastic T or null cell lymphomas

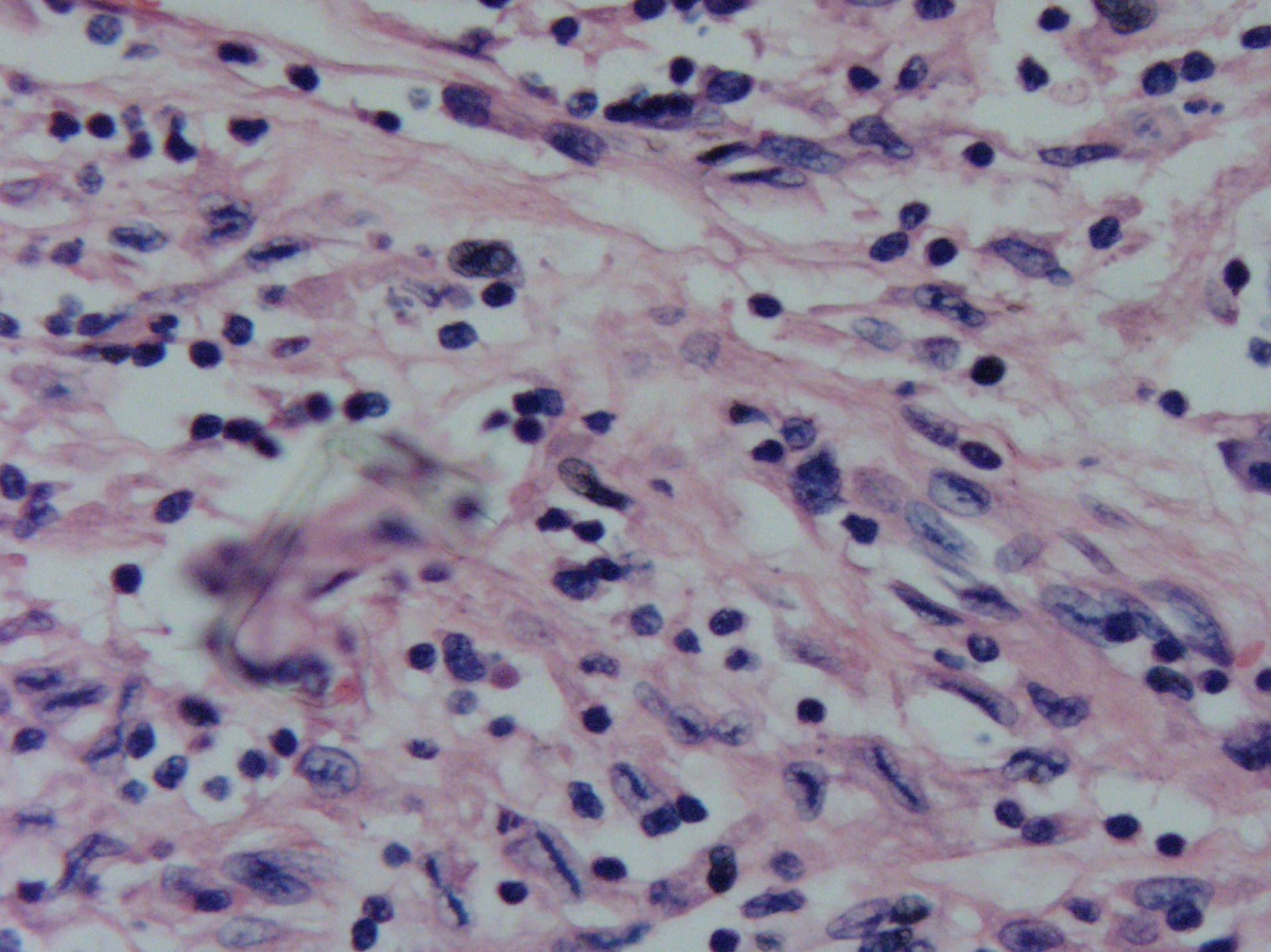
DDX

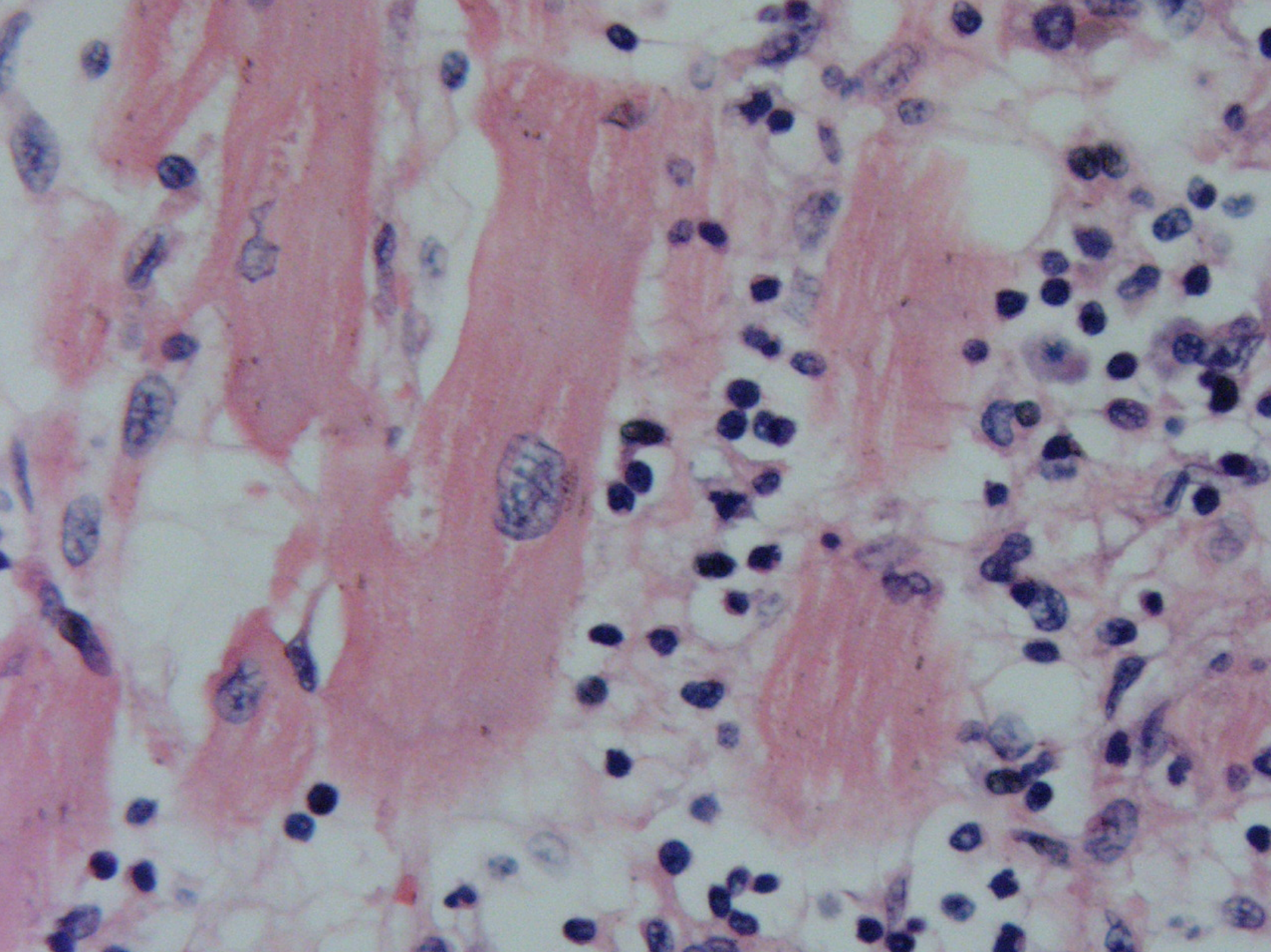
HD-lymphocyte depleted	CD15+ CD30+ CD45-
DLBCL-anaplastic var.	CD20+ CD45+ EMA-
Adrenocortical CA	Inhibin+ VIM+
Metastatic melanoma	S100+ VIM+

Case 39

- 43M with second heart transplant
- First heart transplant occurred after idiopathic illness leading to cardiac failure
- 740gm 15x13x9 cm heart
- Epicardium diffusely granular with adhesions
- RV 0.9cm, LV 2.0cm
- Coronary arteries uniformly narrowed and patent







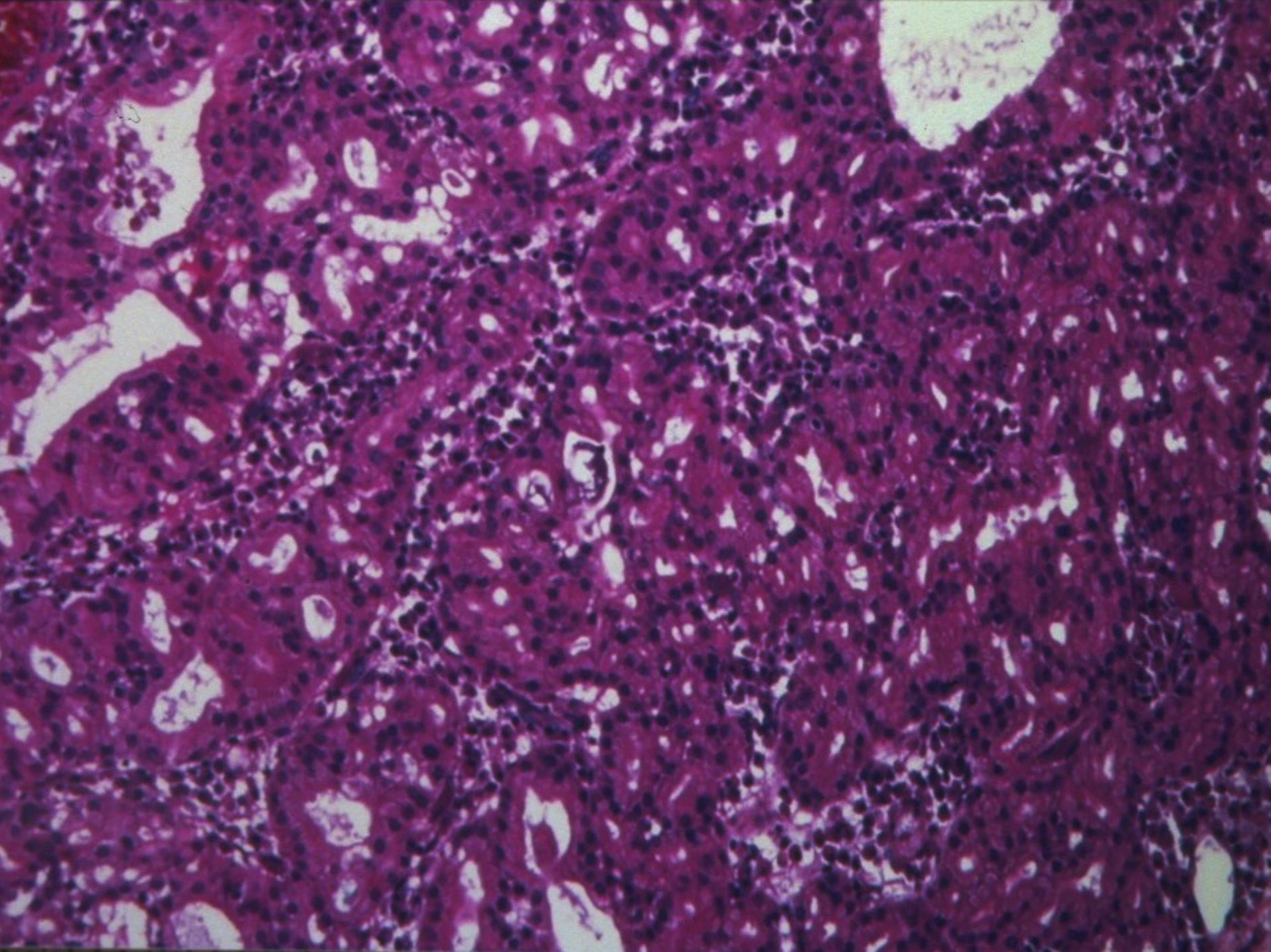
Severe Acute Cellular Rejection and Chronic Vasculopathy

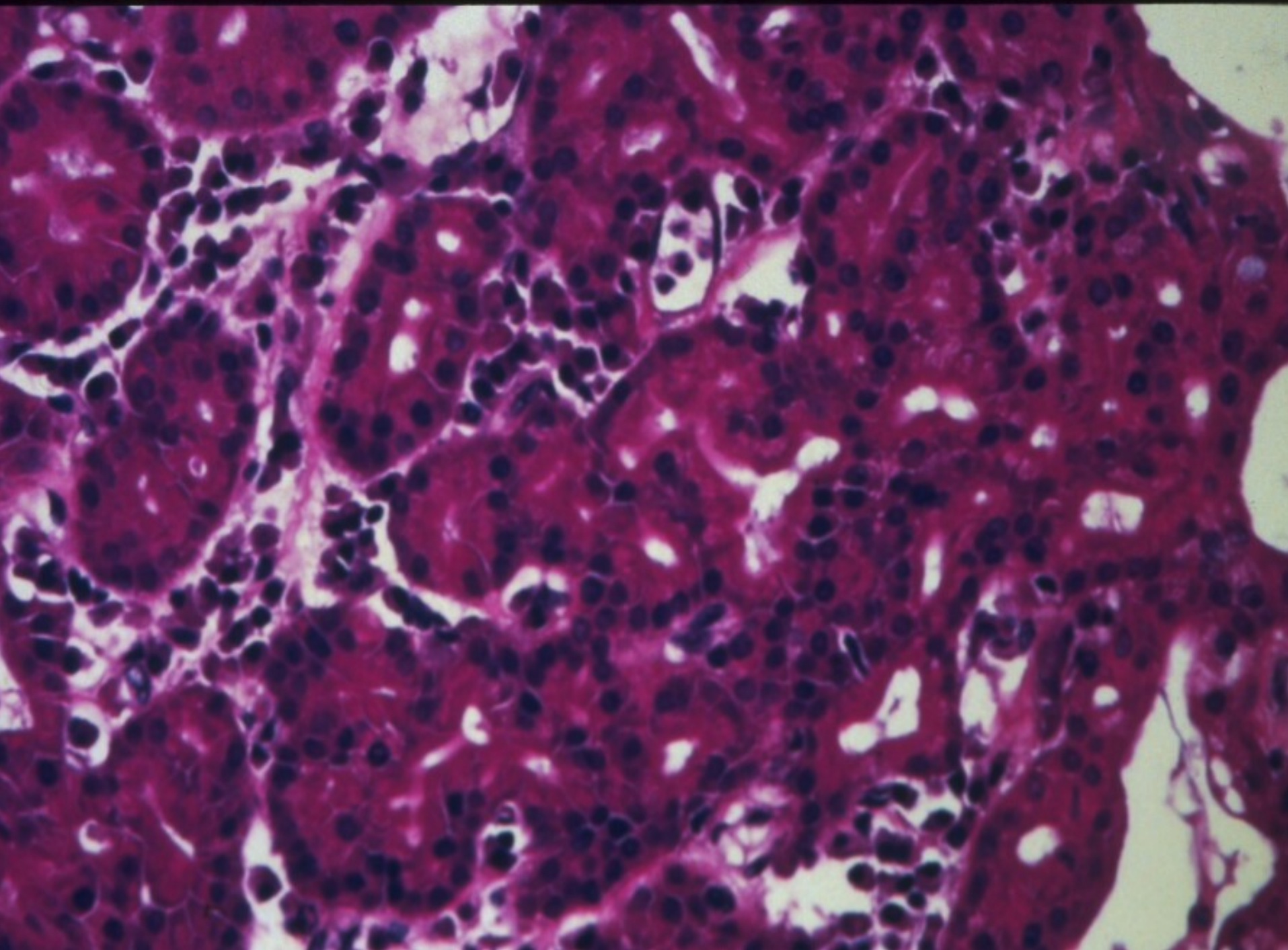
DDX

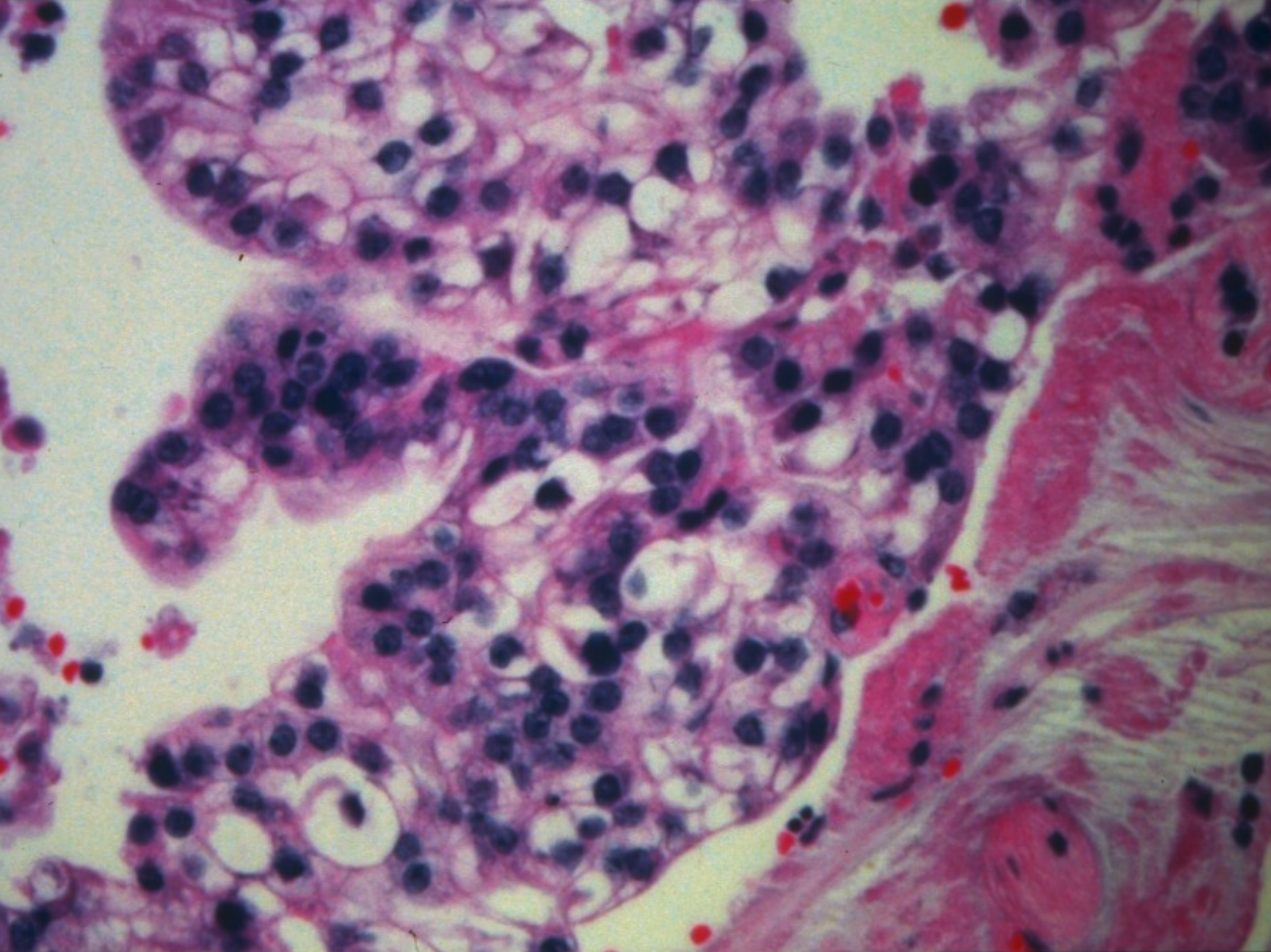
Acute cellular rejection	Most common Cell mediated First 3 months post transplant
Humoral rejection	DIF shows complement or Ig in vessel walls Beyond immediate post-op period
PTLD	Preneoplastic/neoplastic Viral oncogenesis and immunosuppression (EBV)

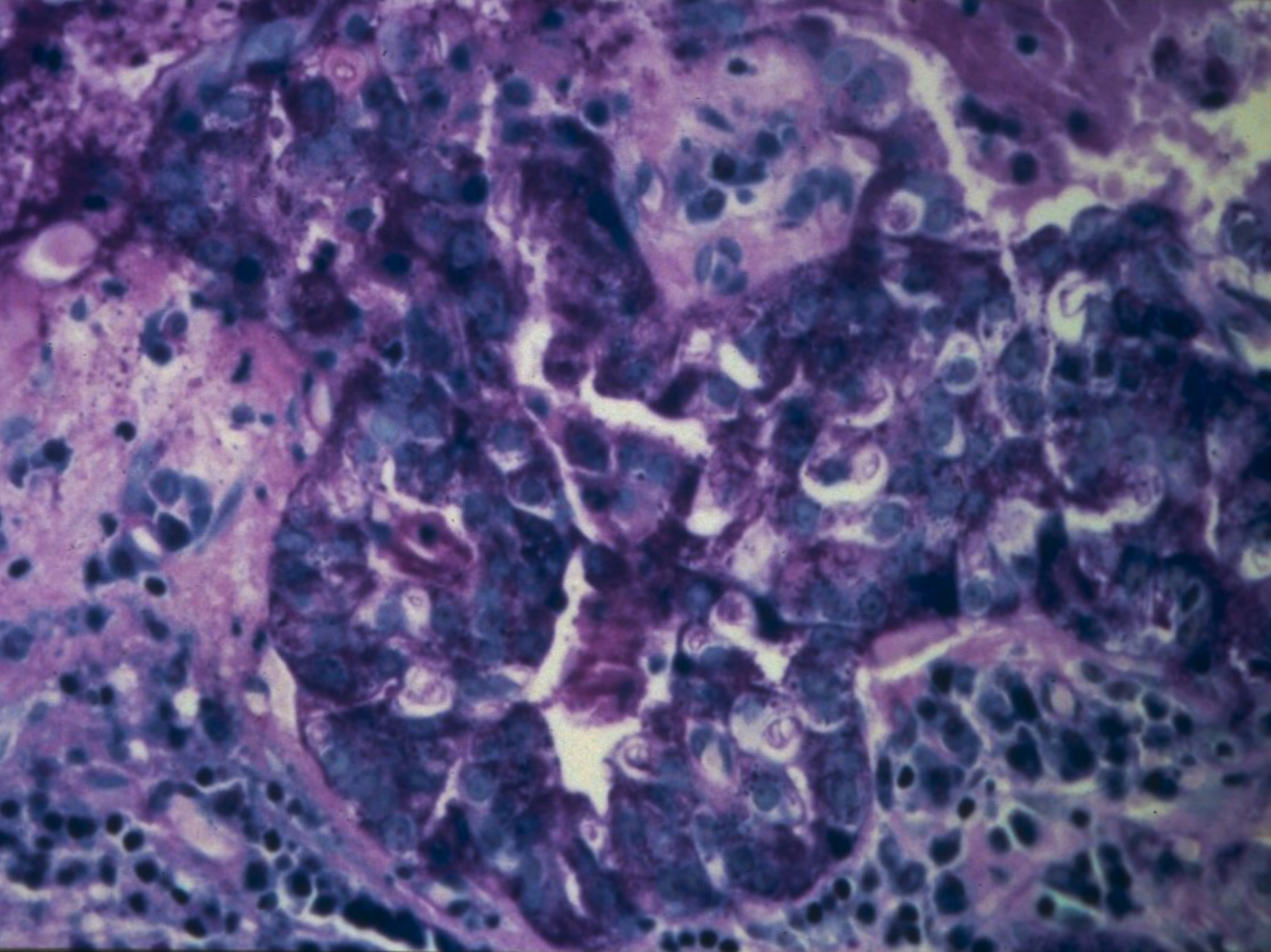
Case 40

- 13F with type I diabetes mellitus presents with right infra-auricular mass gradually enlarging for previous 2 years
- Mass more prominent during episodes of upper respiratory infection but otherwise asymptomatic
- Parotidectomy with 1.9 cm circumscribed tan nodule









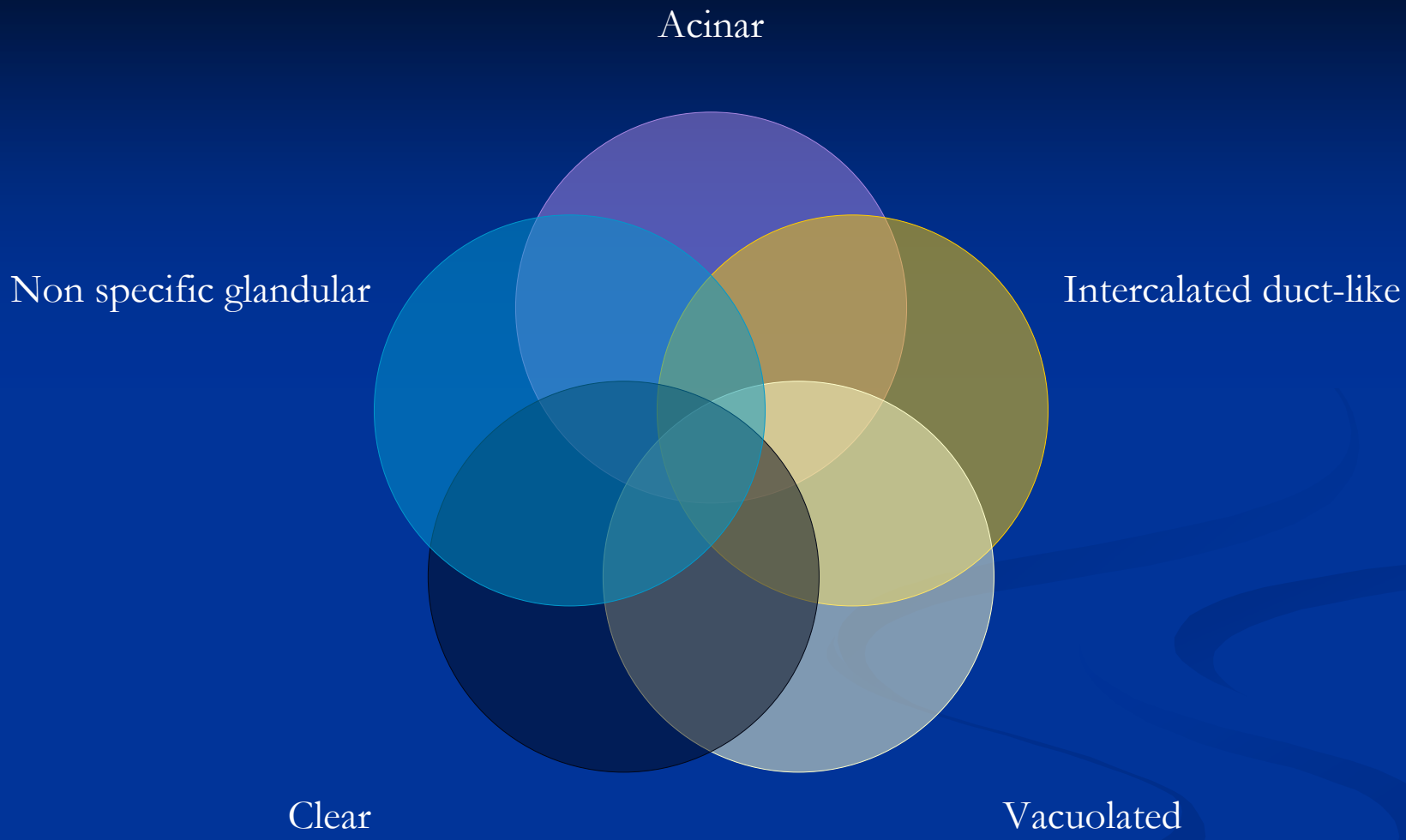
Acinic Cell Adenocarcinoma

Acinic Cell CA

	Overall	Children
Tumor type	AdenoCA, NOS Mucoepidermoid Acinic cell CA	Mucoepidermoid Acinic cell CA

Acinic Cell CA

- Parotid gland most common site
- F:M 3:2
- 2-8th decades
- 3% bilateral but most common bilateral malignant neoplasm
- Recurrence in 1/3
- Death 15%


Acinar
Non specific glandular
Intercalated duct-like
Clear
Vacuolated

Histopathology

- Solid
- Microcystic
- Papillary-cystic
- Follicular
- Abundance of lymphocytic stroma
- Undifferentiated

DDX

Acinic Cell CA	May be bilateral
Polymorphous LGAC	Monomorphous cells in cords and trabeculae Minor salivary glands
Mucoepidermoid CA	Most common malignant salivary gland in children