

Disclosures

February 1, 2016

The following planners and faculty have no financial relationships with commercial interests to disclose:

Presenters:

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Andrew Horvai, MD
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Sarah Cherny, MD
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John Higgins, MD
Ankur Sangoi, MD

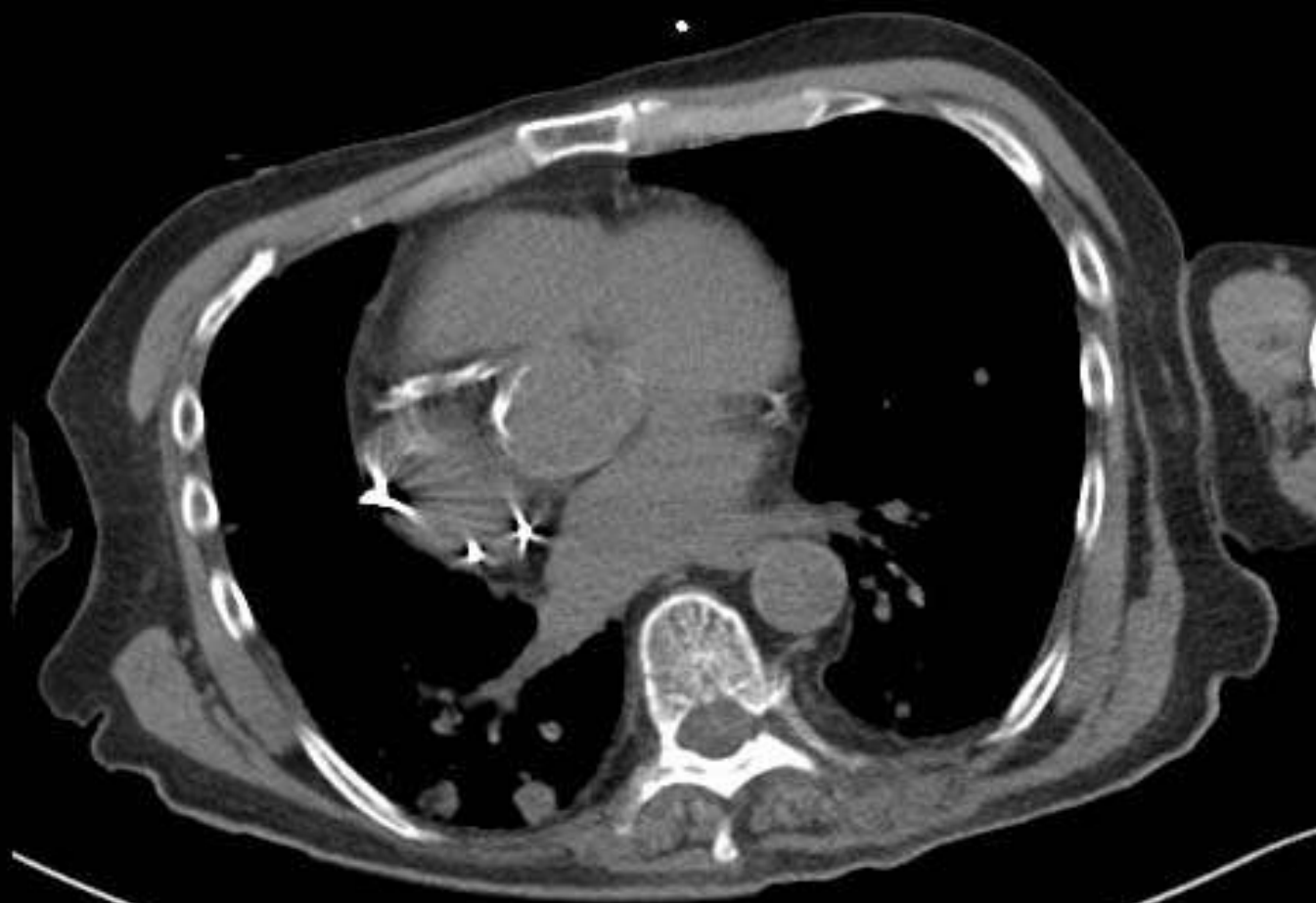
Activity Planners:

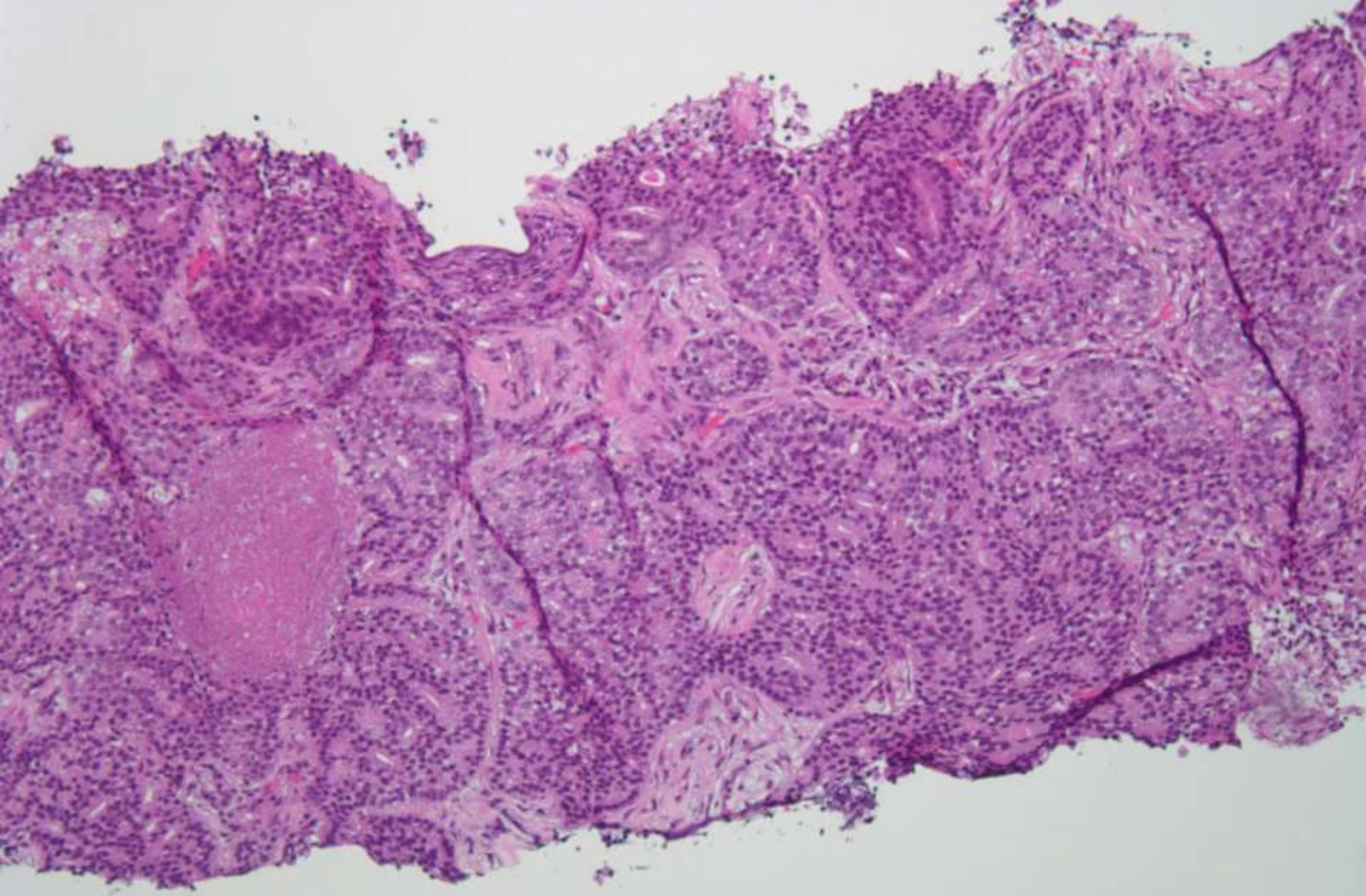
Kristin Jensen, MD
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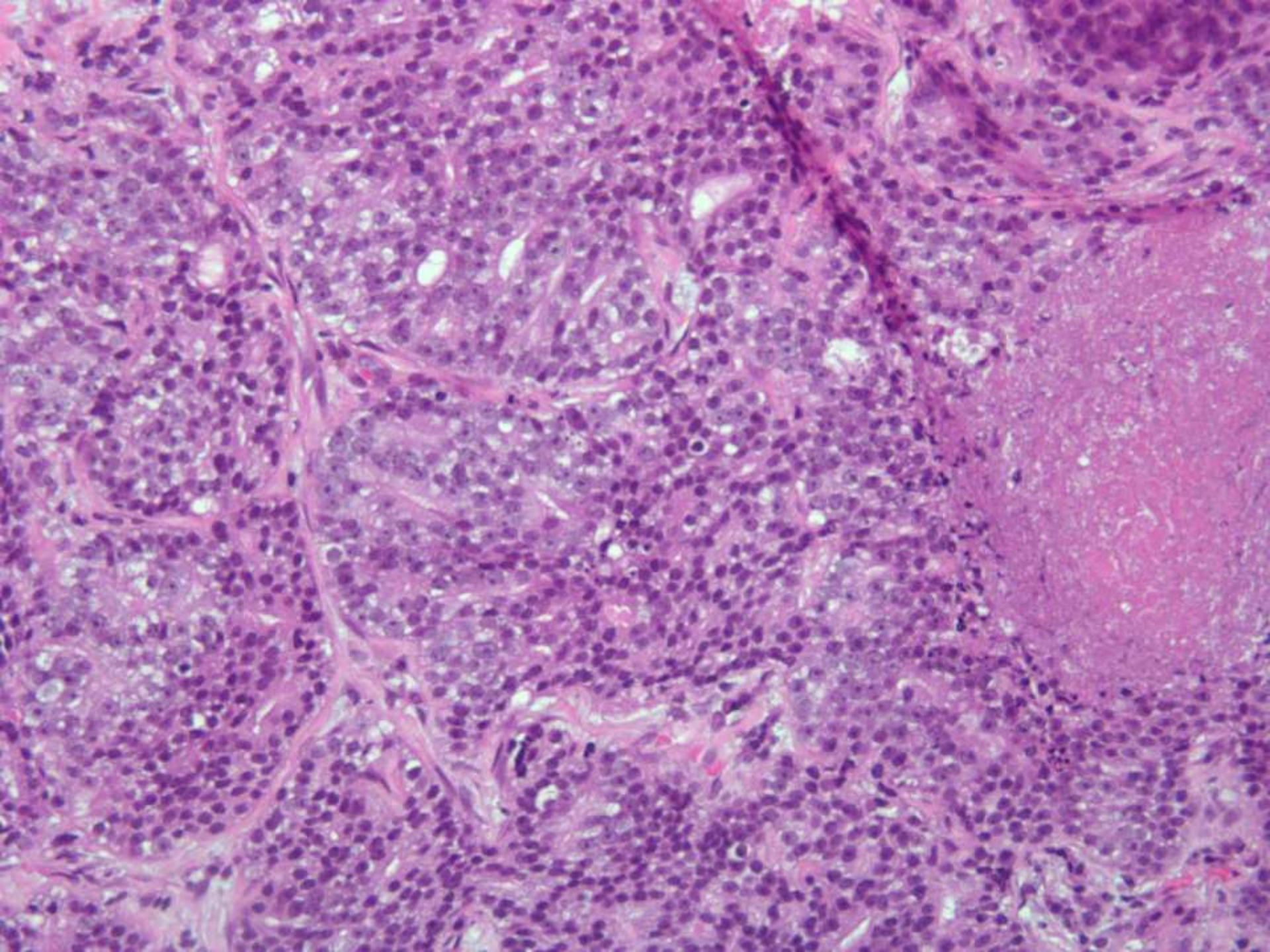
SB 6021

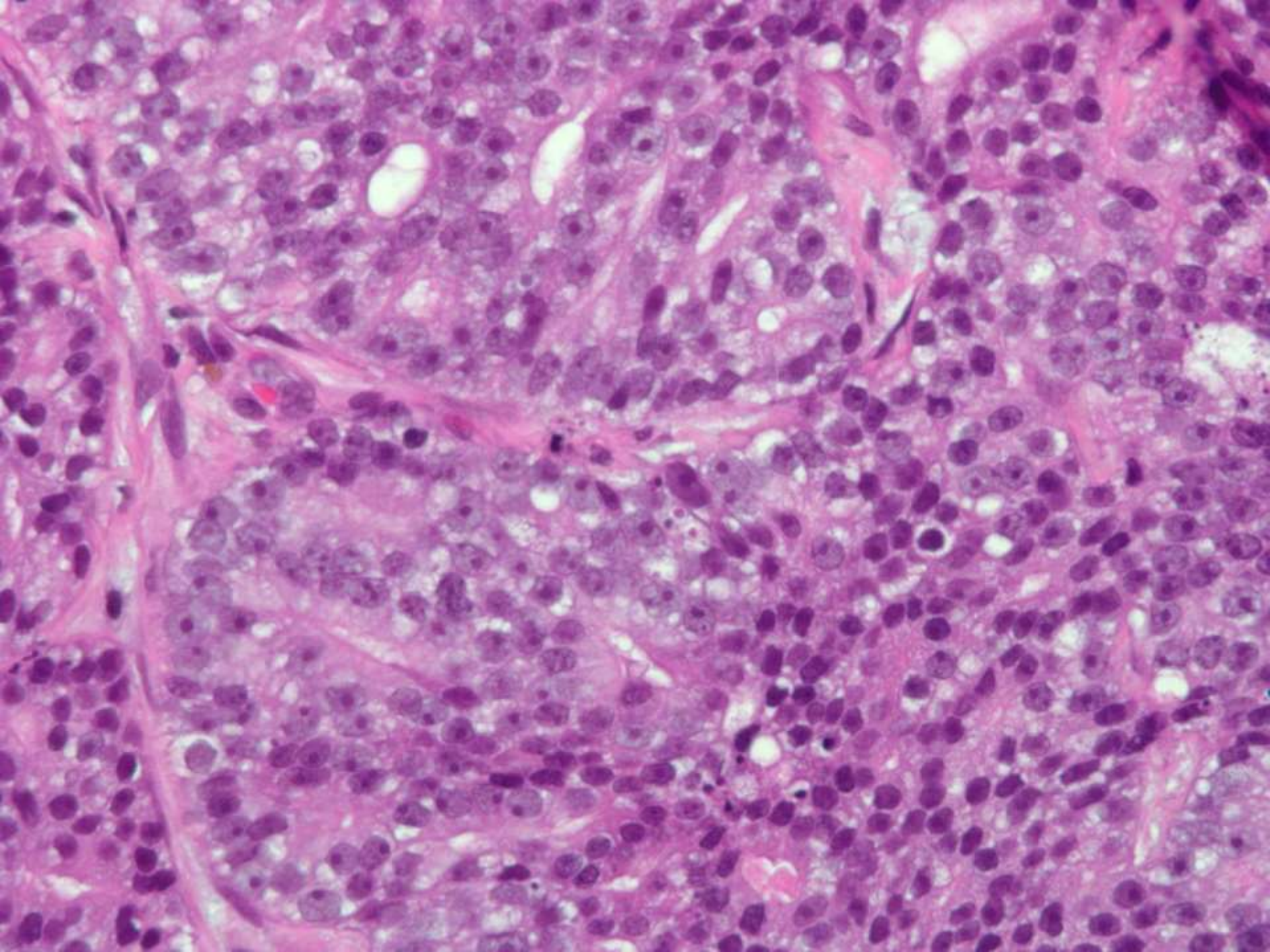
Charles Lombard; El Camino Hospital

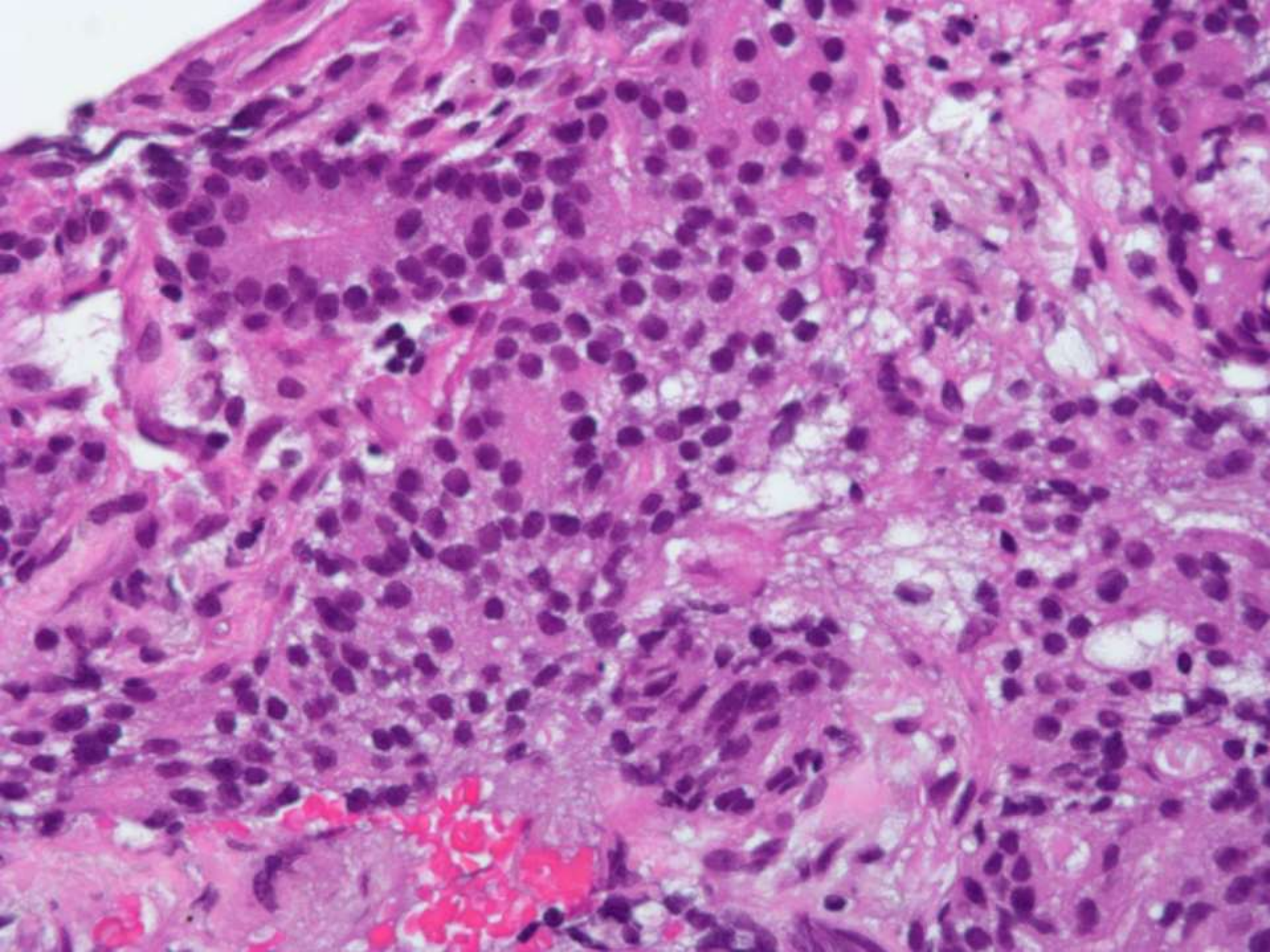
87-year-old man with a history of prostate cancer (normal serum PSA) and a lung mass which was biopsied.



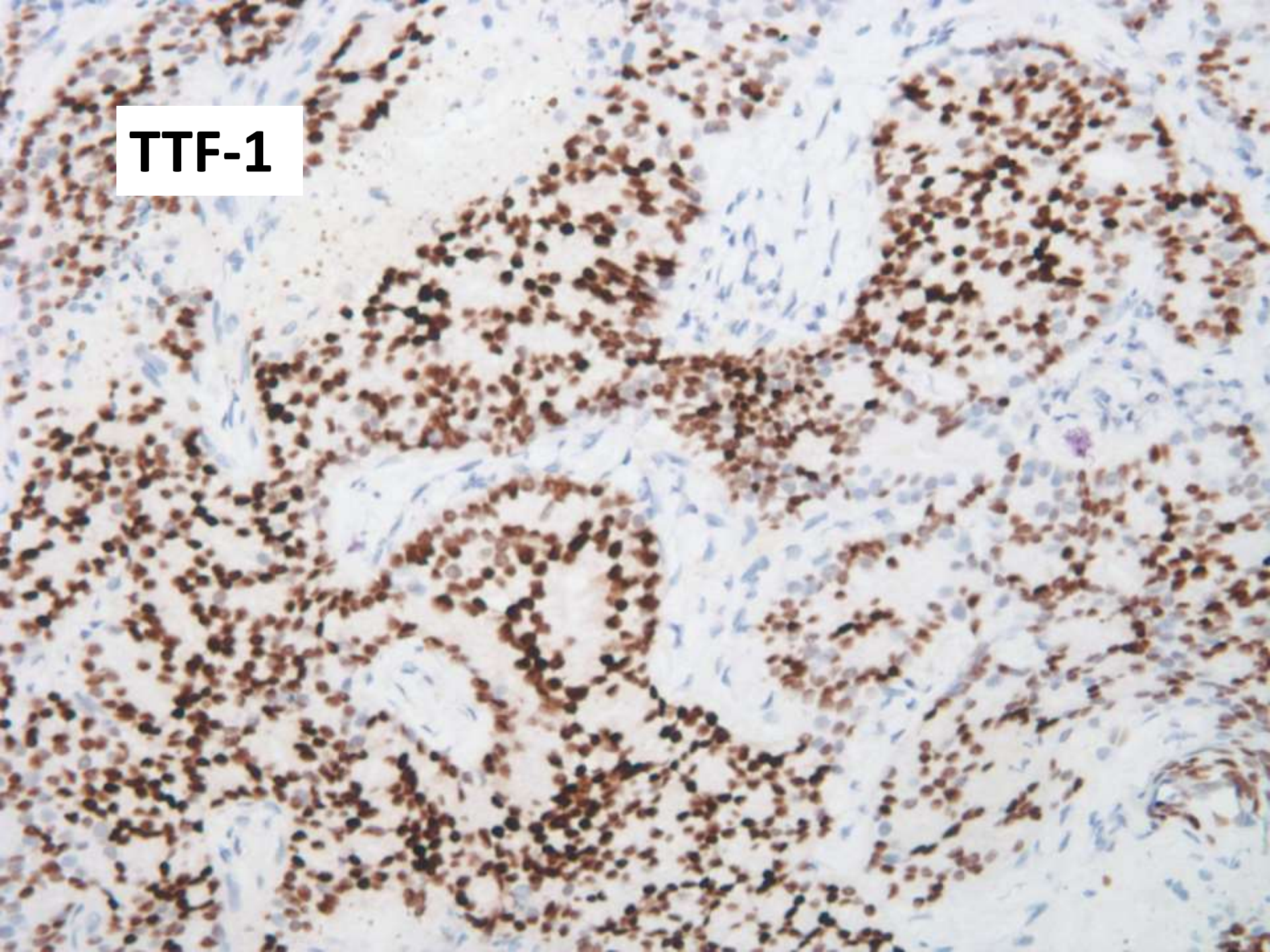








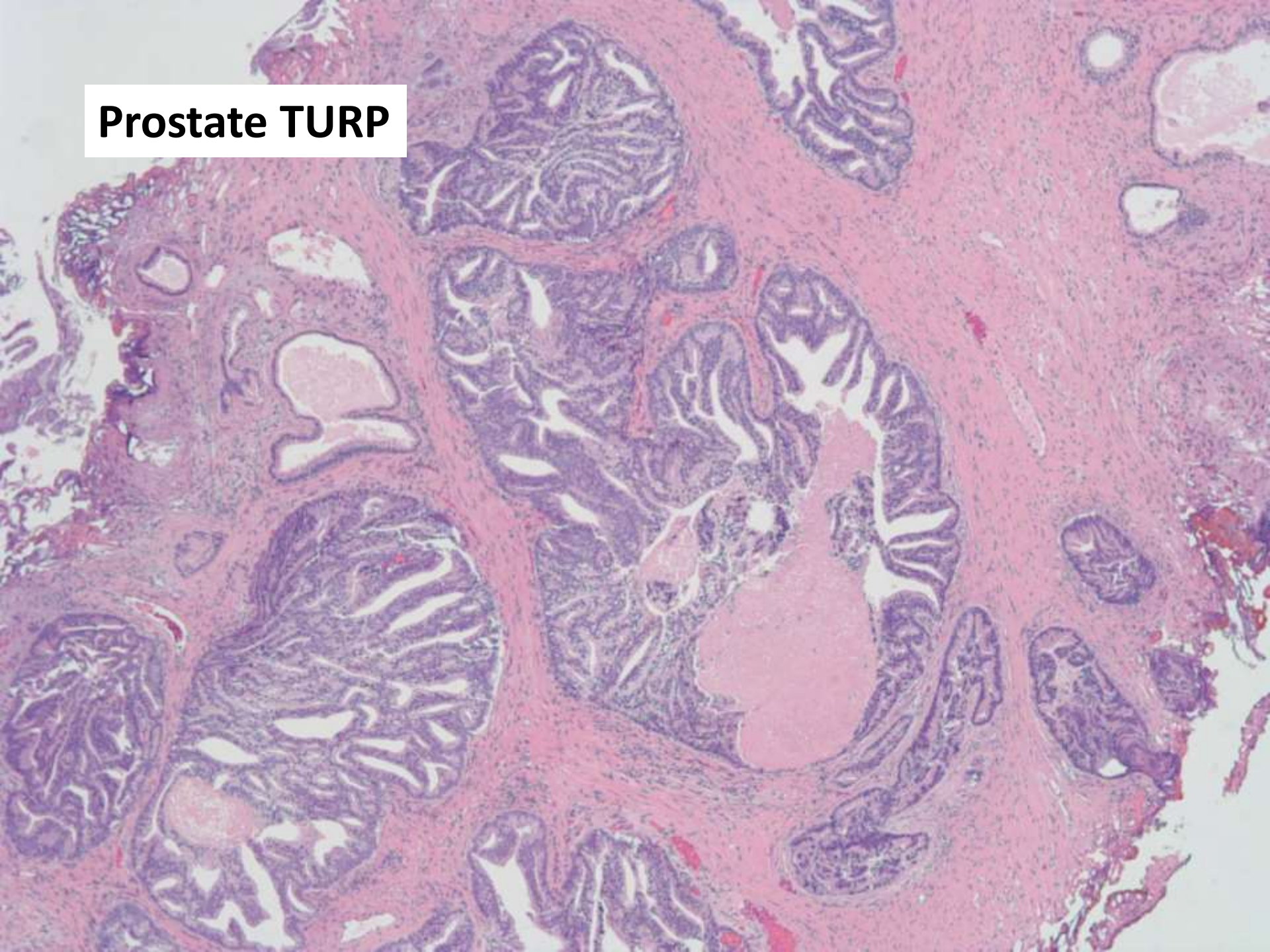
TTF-1

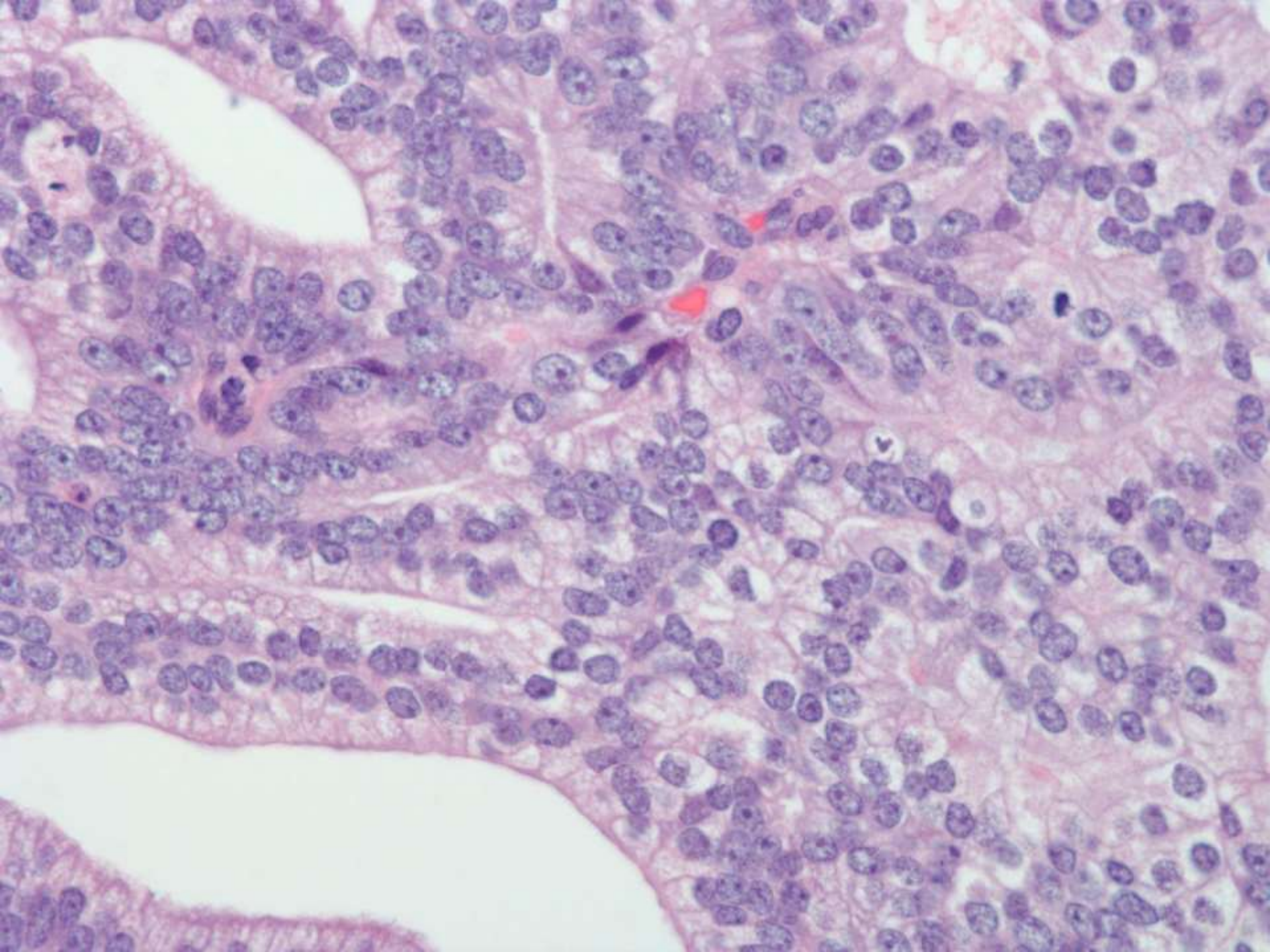


DIAGNOSIS?



Prostate TURP





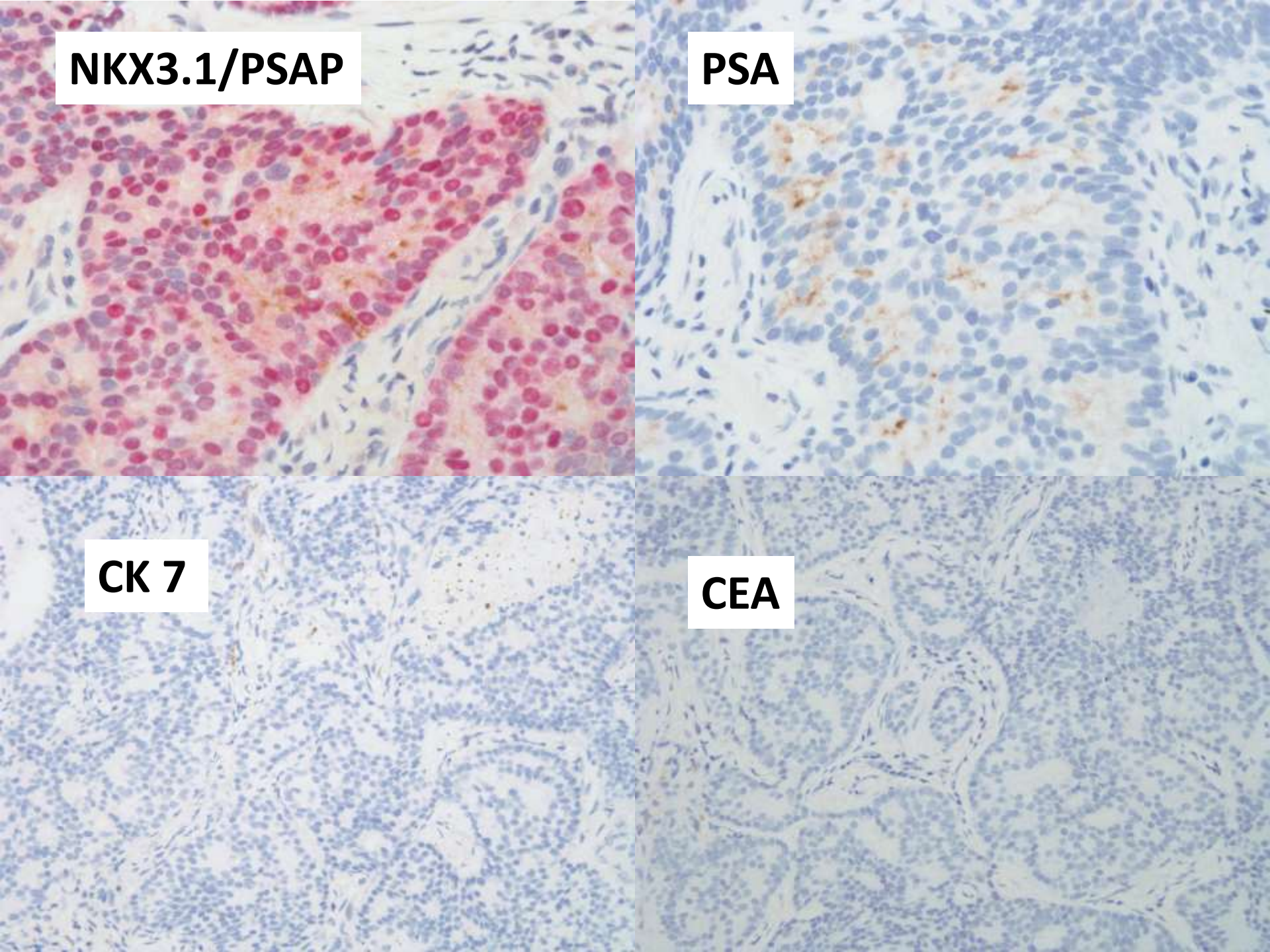
Lim et al: “TTF-1 may be expressed in ductal adenocarcinoma of the prostate: A potential pitfall.” J Clin Path 2007;60: 941-43.

NKX3.1/PSAP

PSA

CK 7

CEA



Morgan et al: “Ductal
adenocarcinoma of the prostate:
increased mortality risk and
decreased PSA secretion.”
J Urology 184(6): 2303-7;2010

Ductal Adenocarcinoma of Prostate

- 0.4-0.8% of prostate cancer (pure cases)
- Up to 5% Mixed cases
- Compared to Acinar type adenocarcinoma
 - Metastases more likely (12 vs 4%)
 - Higher Gleason score (usually 8)
 - Lower PSA with 2x increased odds $PSA < 4.0$
 - For cases with no mets initially 2.4 x increased disease specific mortality
- Comparison of 442,881 acinar cases vs 371 ductal
- National Cancer Registry

SB 6022

Yuna Kang/Andrew Horvai; UCSF

43-year-old man with left shoulder joint and soft tissue mass, prior history of synovial chondromatosis, excised 5 years ago.

2009 plain film



2010 plain film



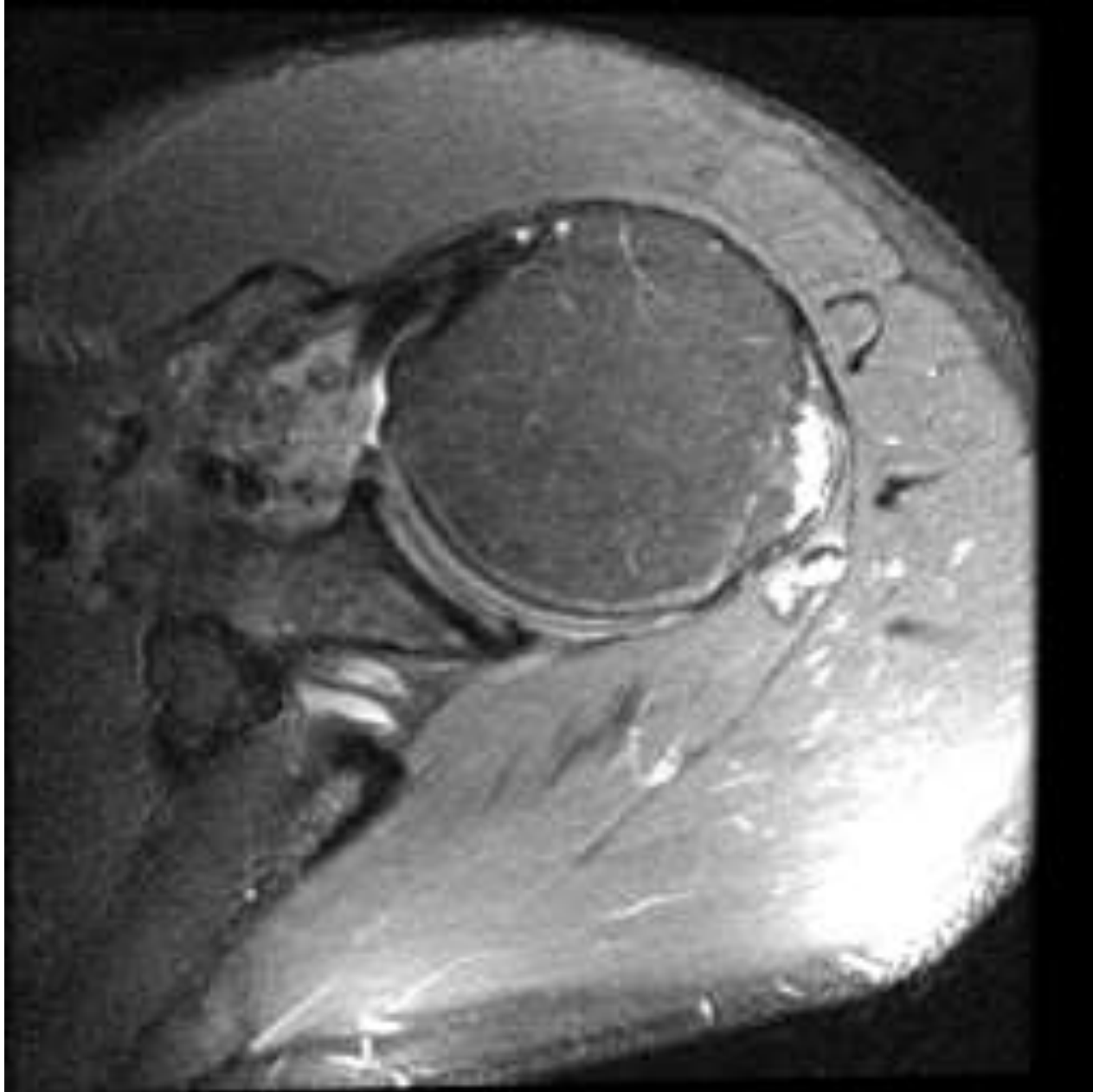
2013 plain film

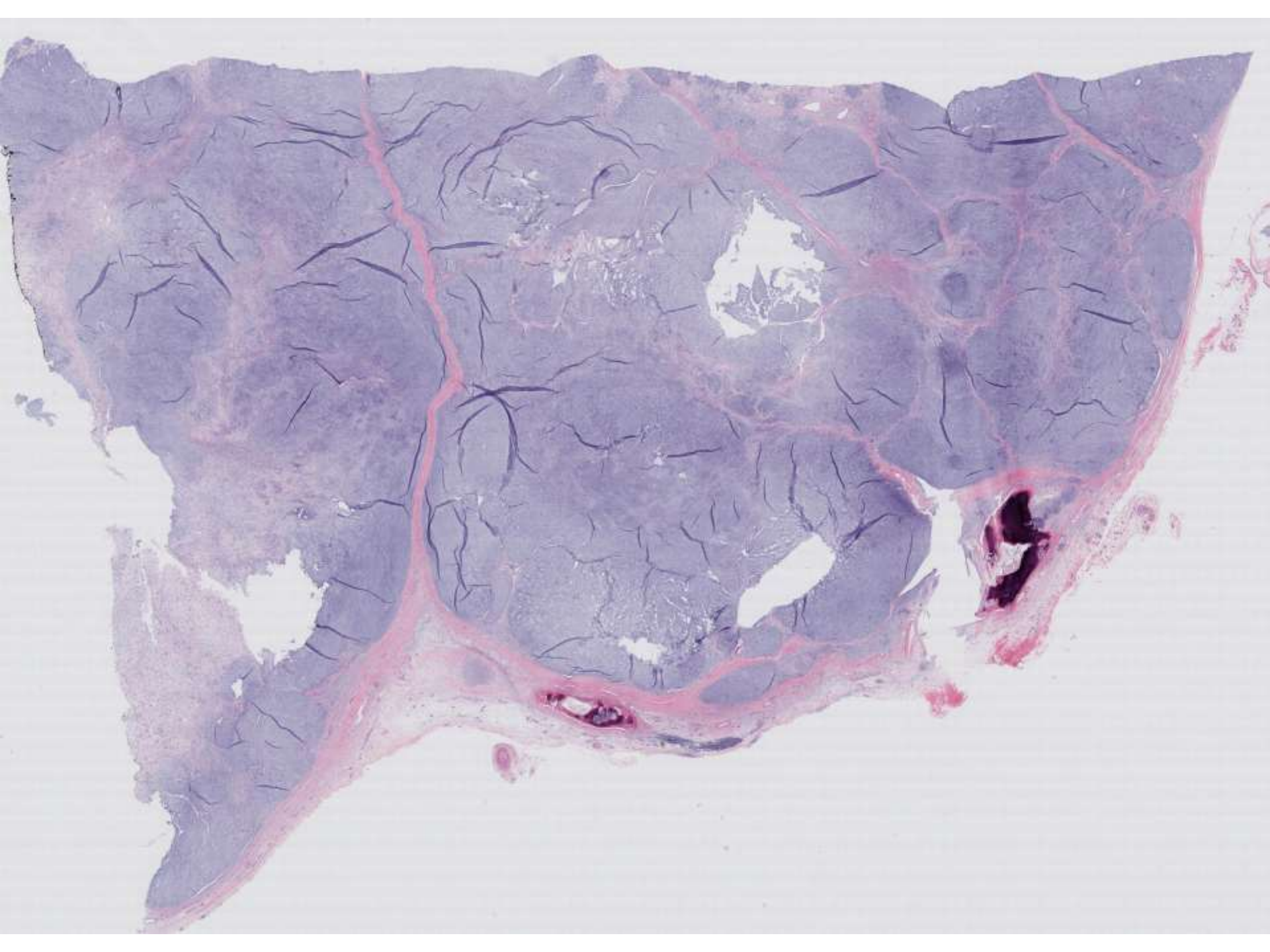


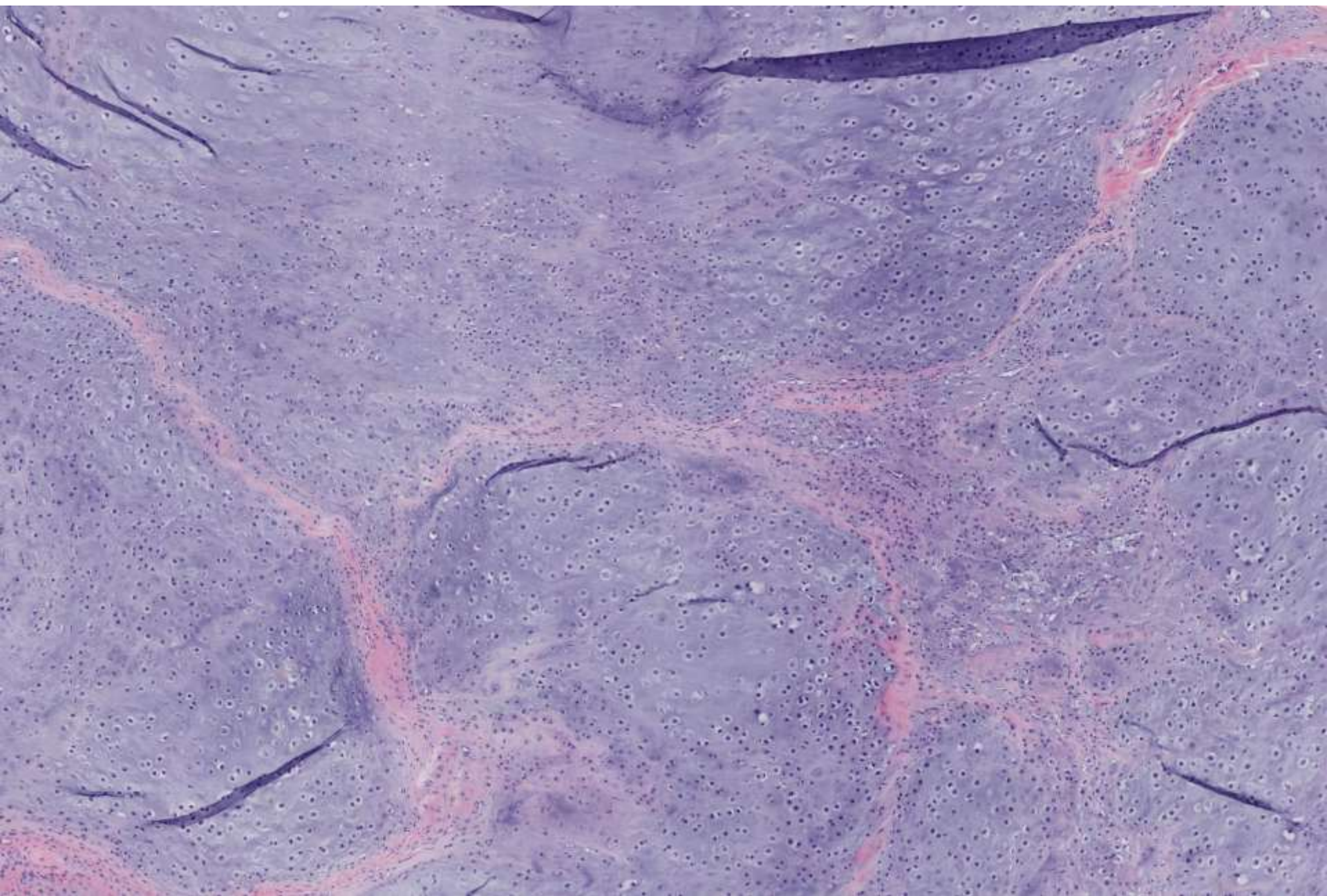
2015 plain film

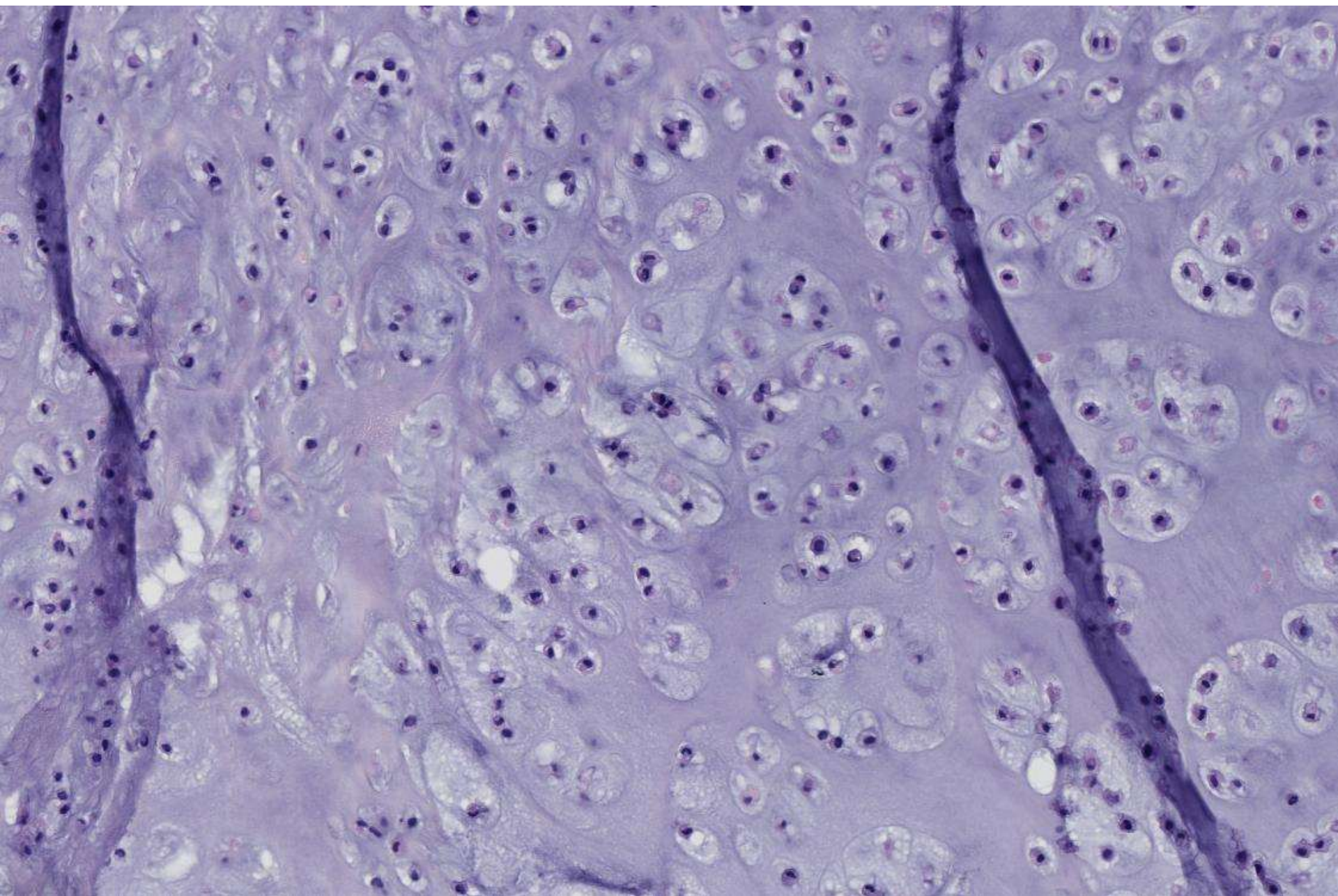


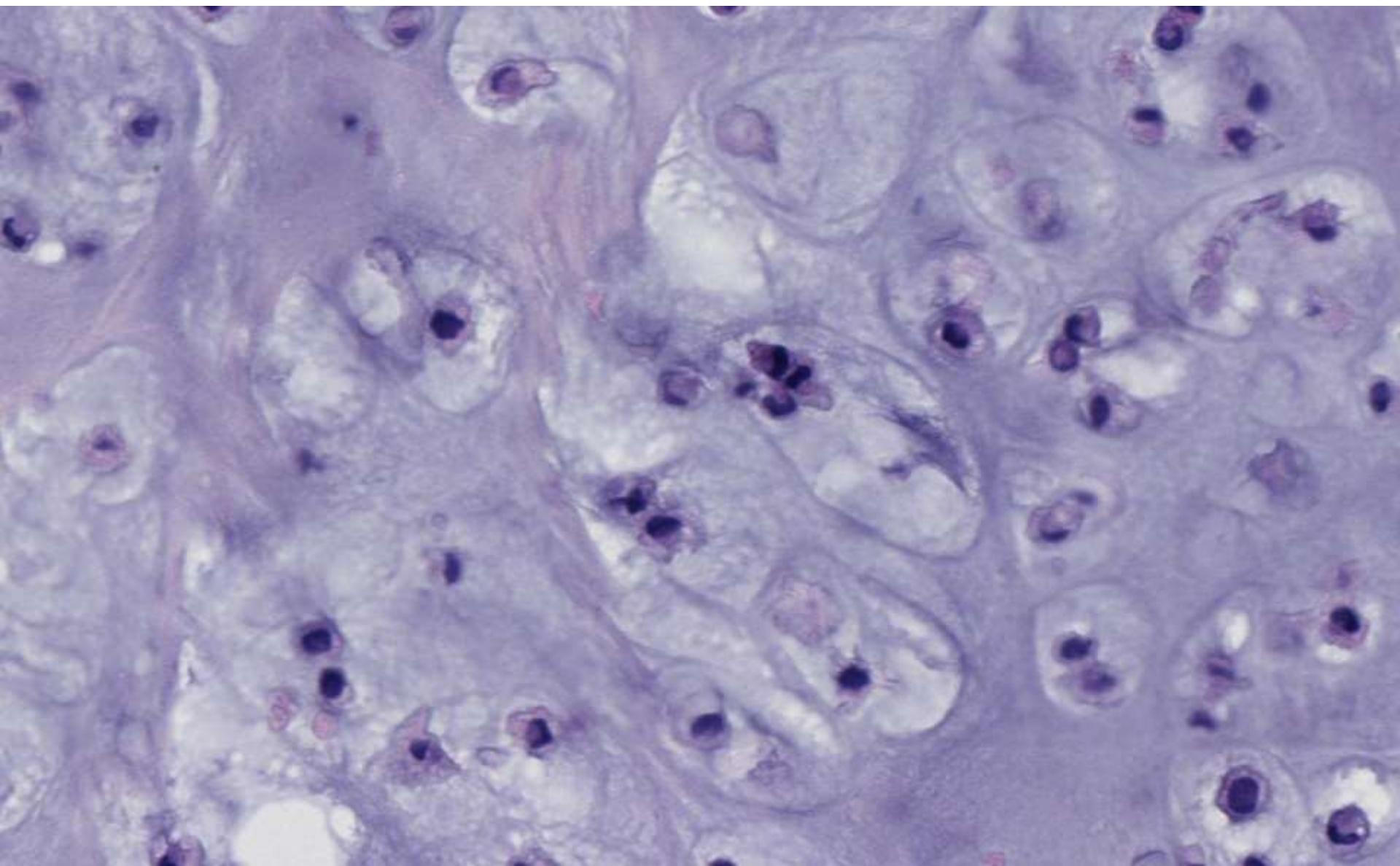
2015 MRI Axial Proton Density

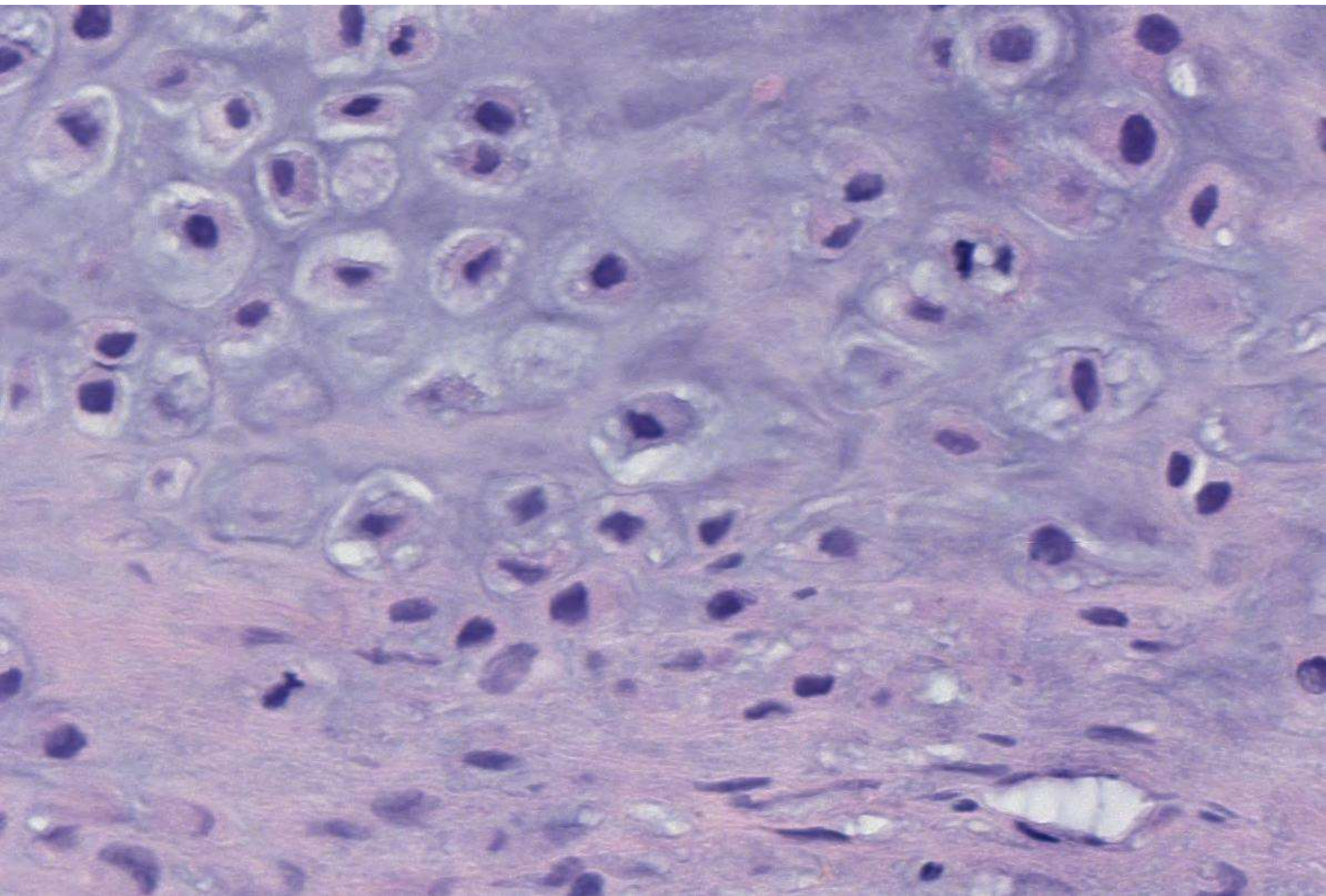












DIAGNOSIS?



43year old man

- 2009- Synovial chondromatosis, local excision
- 2010-2015- Radiographic evidence of recurrence
- 2015 – referred to UCSF

2009 Plain Film

SAR092400

S:247 L:34 - 40951220



Humerus, Lateral

Mar 20, 2009 8:50:47 AM

2010 Plain Film



2013 Plain Film

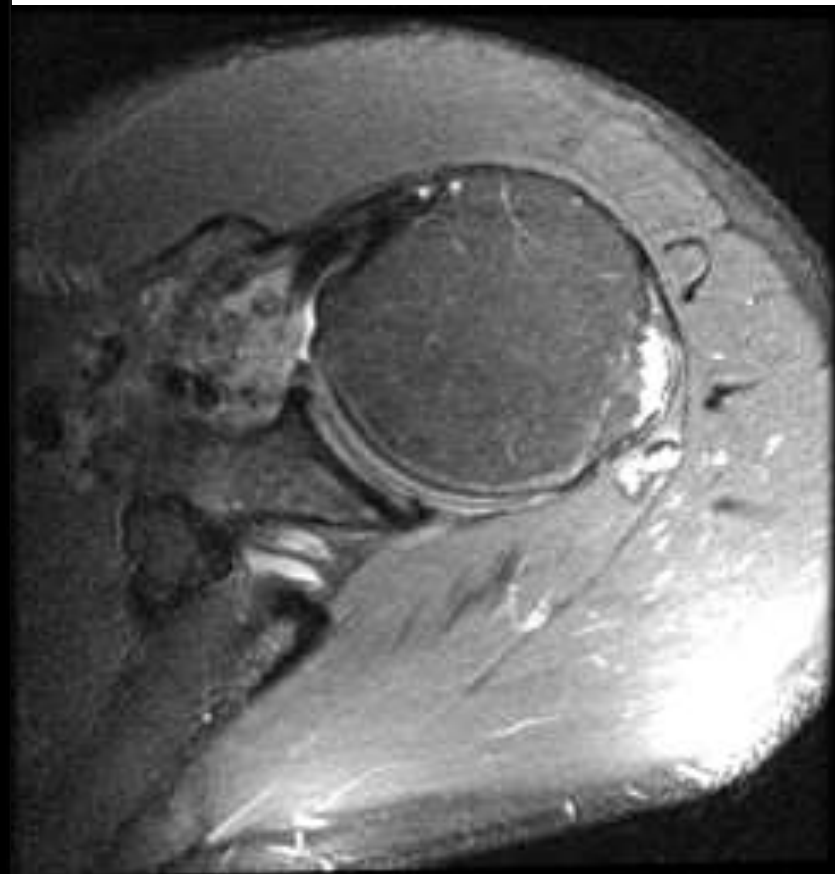


2015 Plain Film

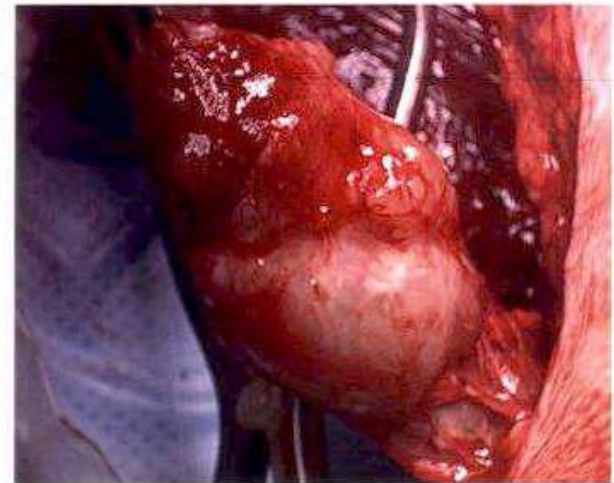
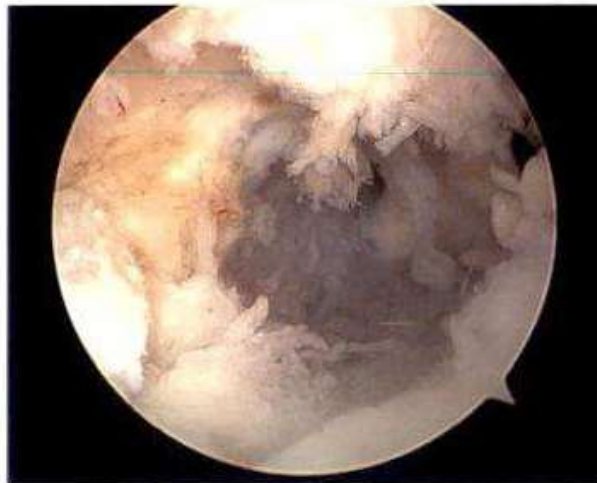
2015
JTA

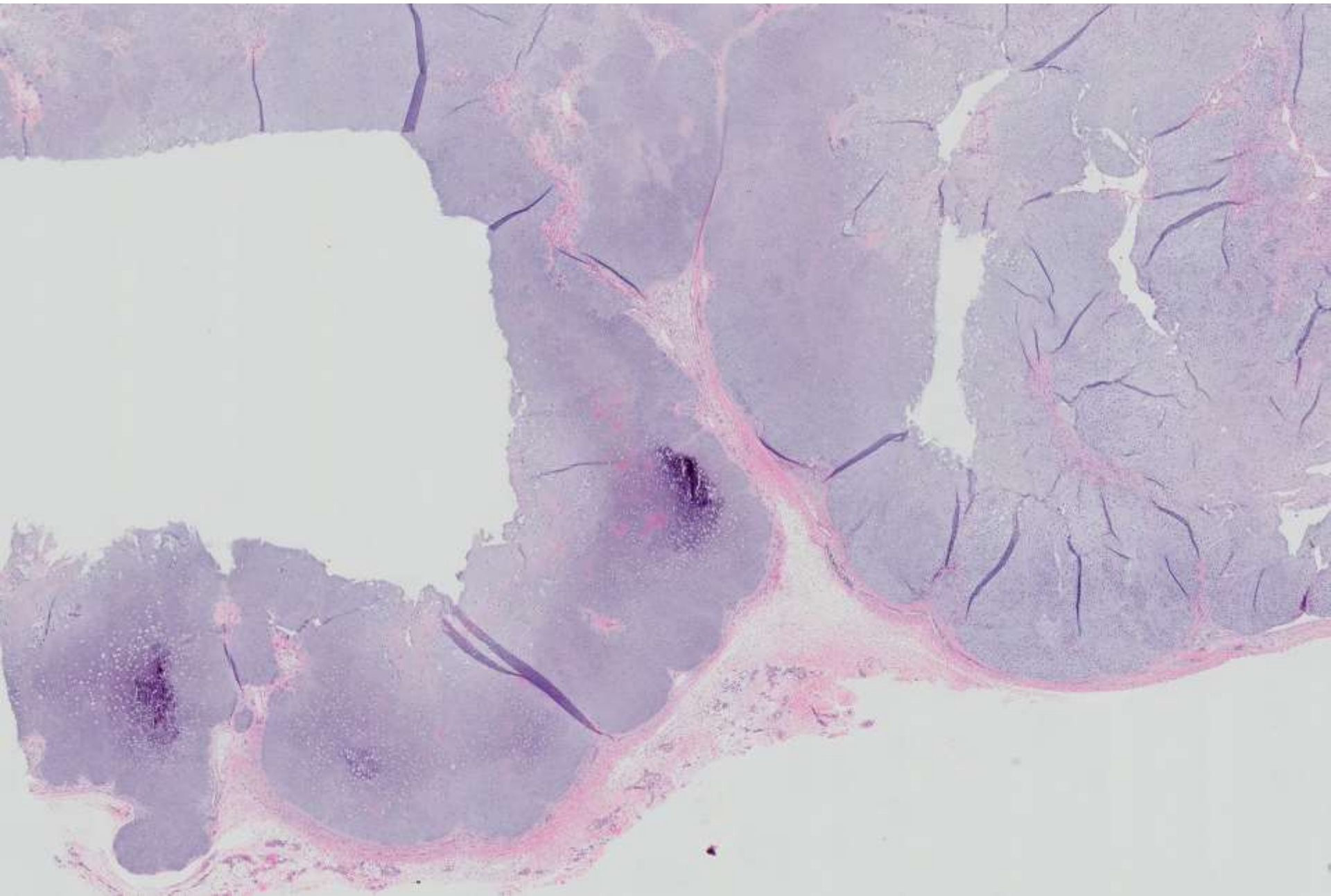


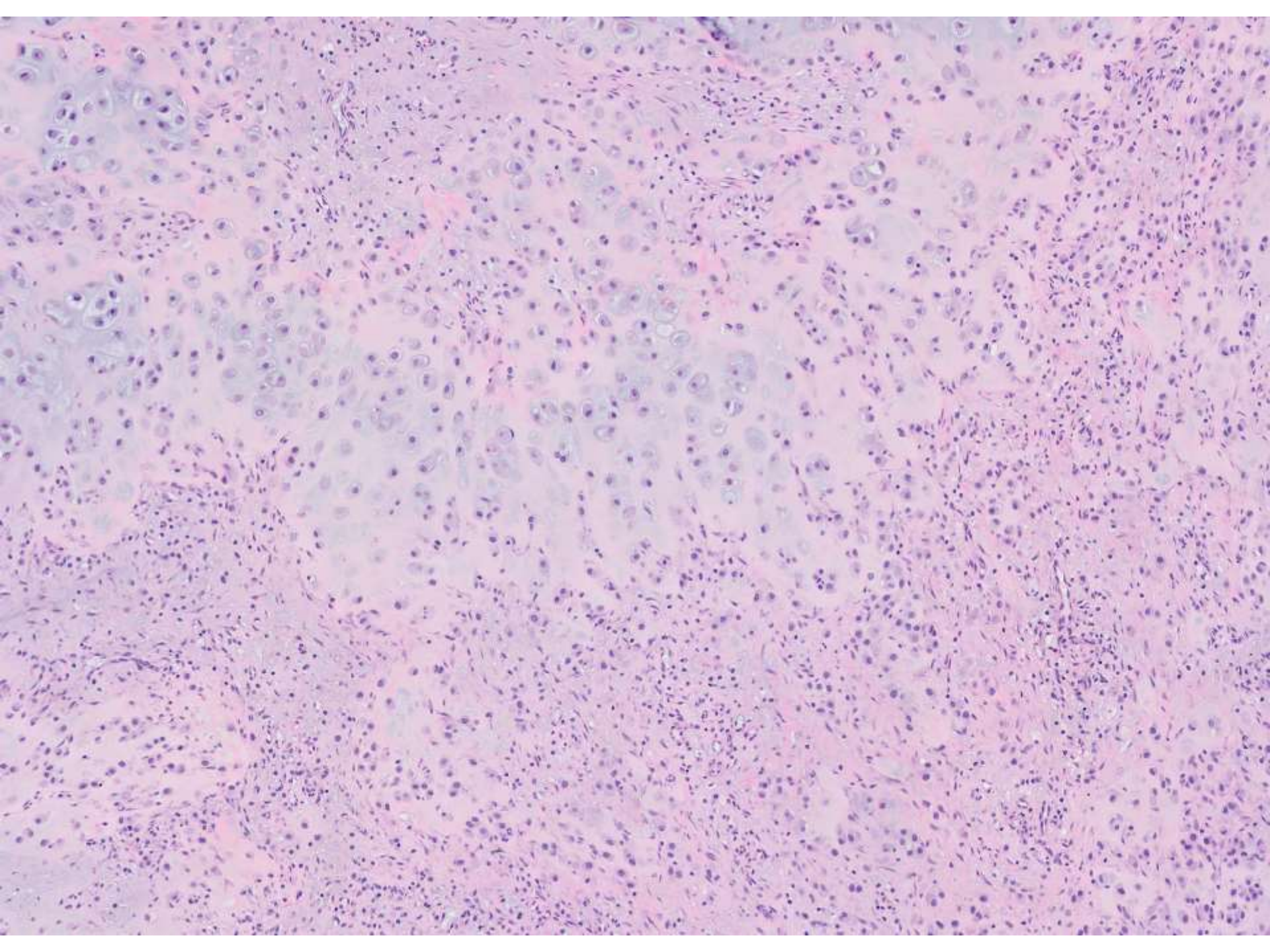
2015 MRI axial proton density

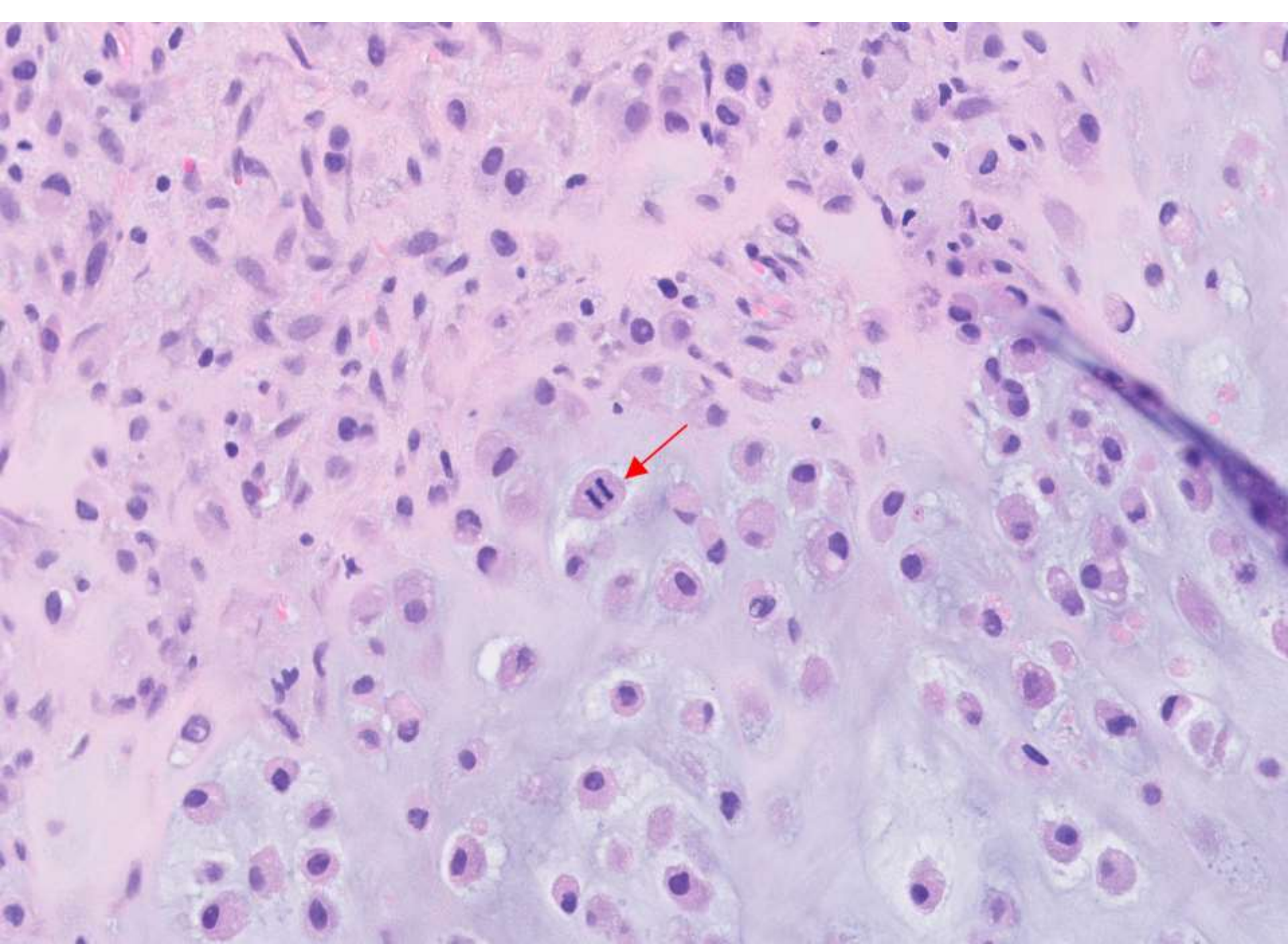


2015 Arthroscopy and intraoperative









Diagnosis:

- **Synovial chondromatosis with atypical features**
- **(Not quite enough for secondary chondrosarcoma)**

Secondary chondrosarcoma arising in synovial chondromatosis

- **Clinical**

- Rare complication (1-5%)
- Mean ~20 years after initial diagnosis, multiple recurrences
- Knee, hip most common
- Lung metastasis in ~30% of cases (!)

- **Radiographic**

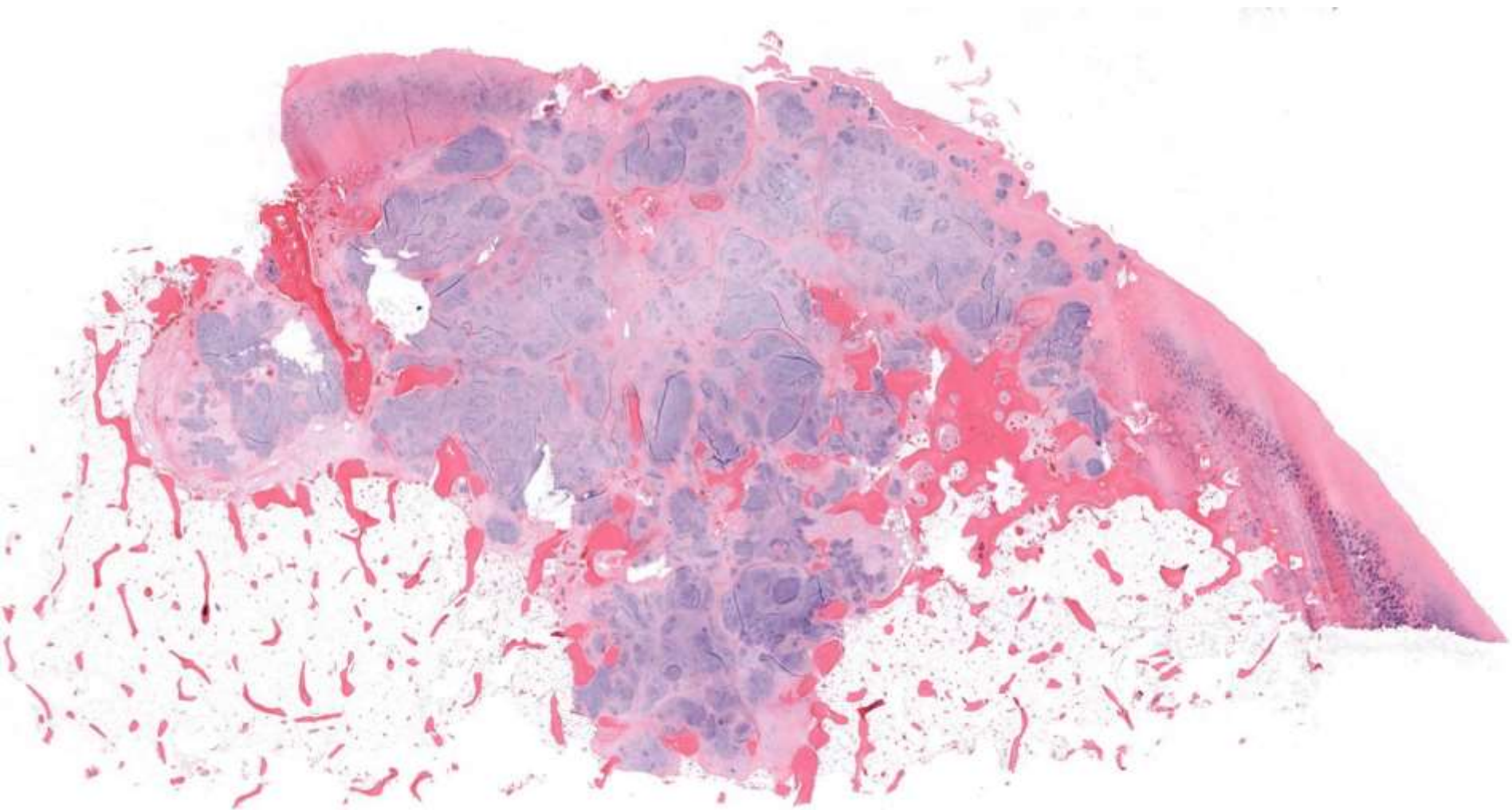
- Indistinguishable from synovial chondromatosis on imaging unless there is bone destruction

Secondary chondrosarcoma arising in synovial chondromatosis

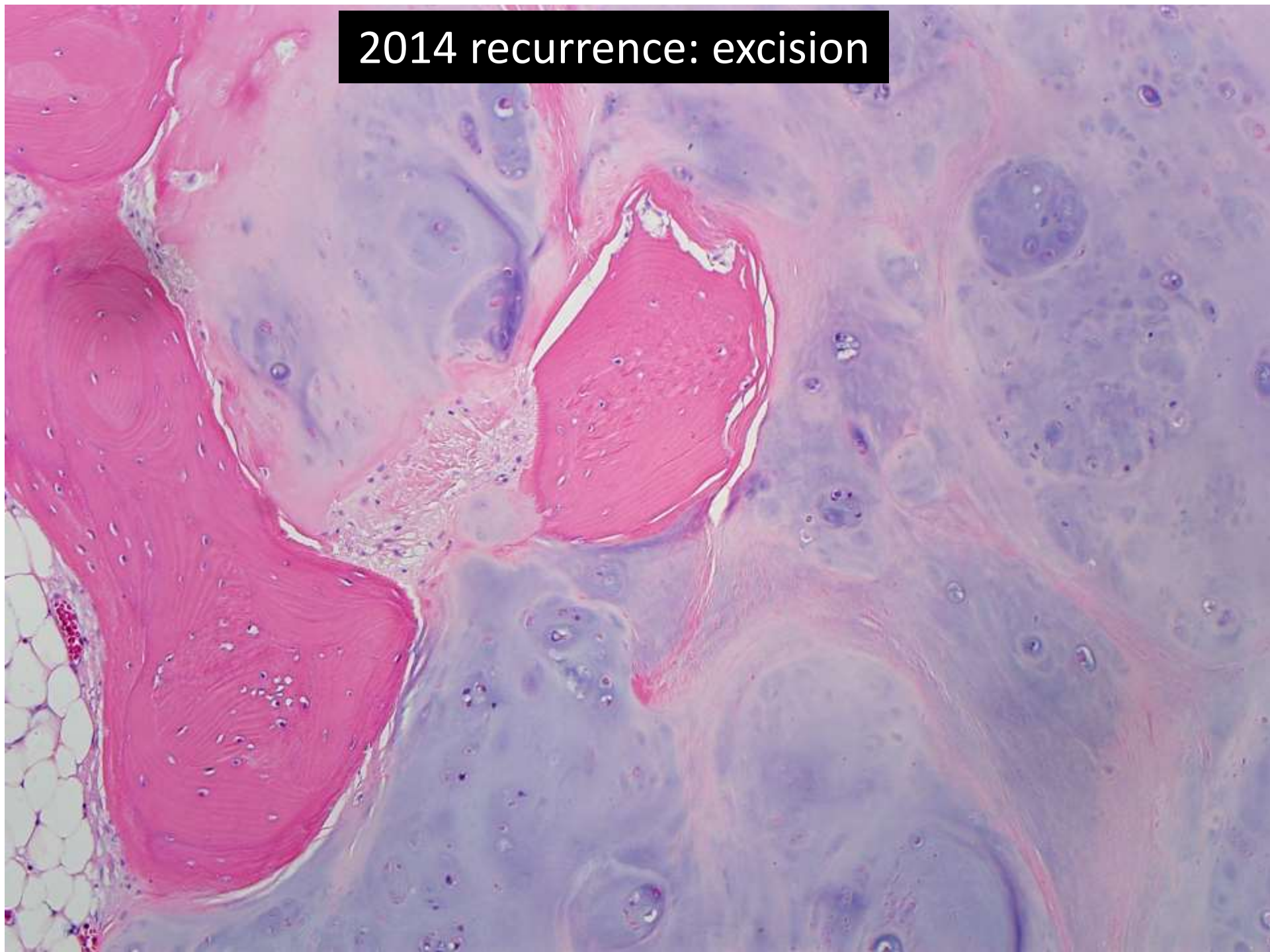
- **Pathology**

- Malignant features can be focal – biopsies may not be diagnostic
- Architectural: Myxoid change, sheets, entrapment of trabecular bone
- Cytologic: Marked nuclear atypia, spindling, mitotic activity
- Problems
 - How much atypia, cellularity and myxoid change is enough?
 - How many of above features required for malignancy?

Secondary chondrosarcoma



2014 recurrence: excision



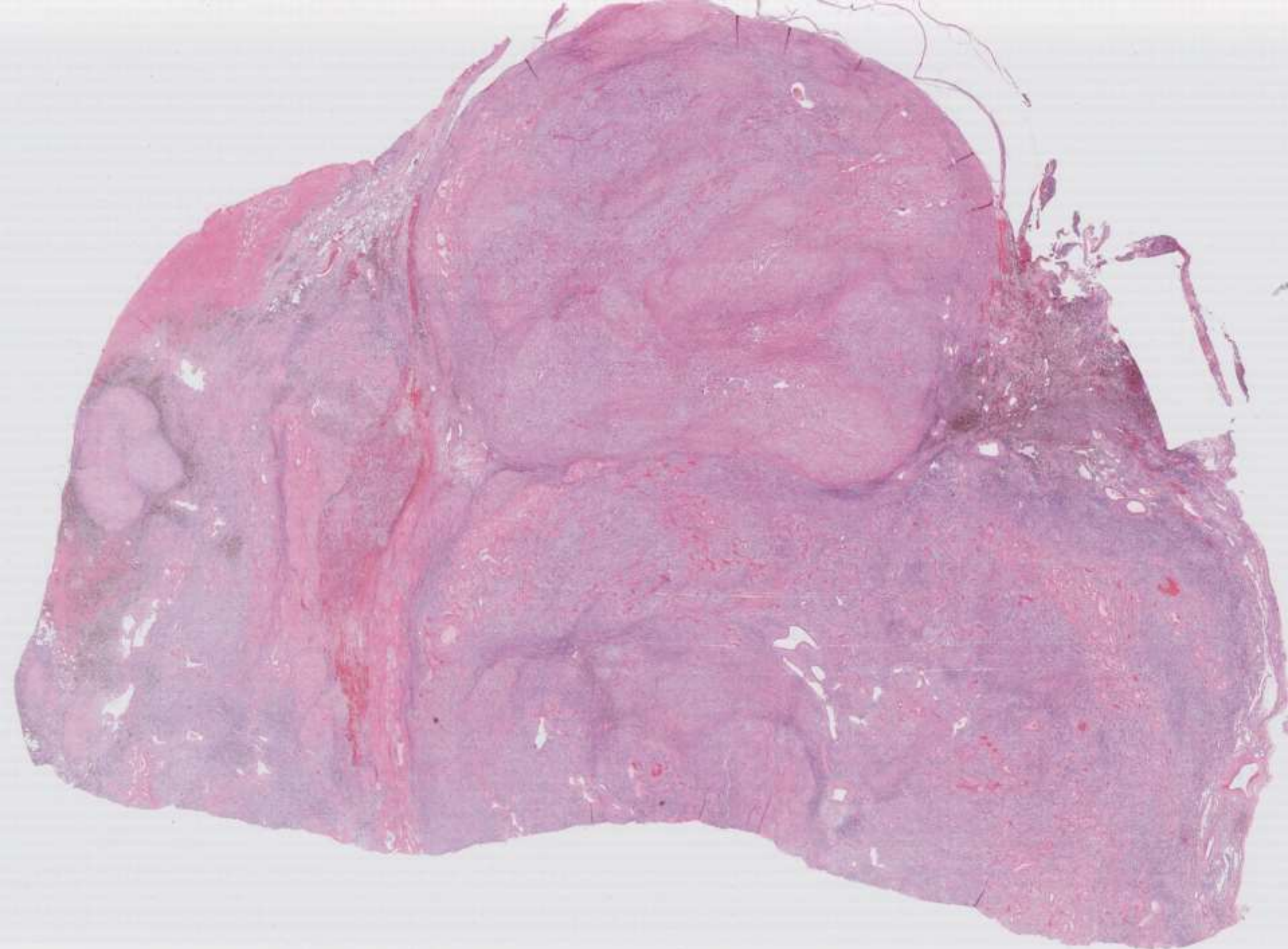
References

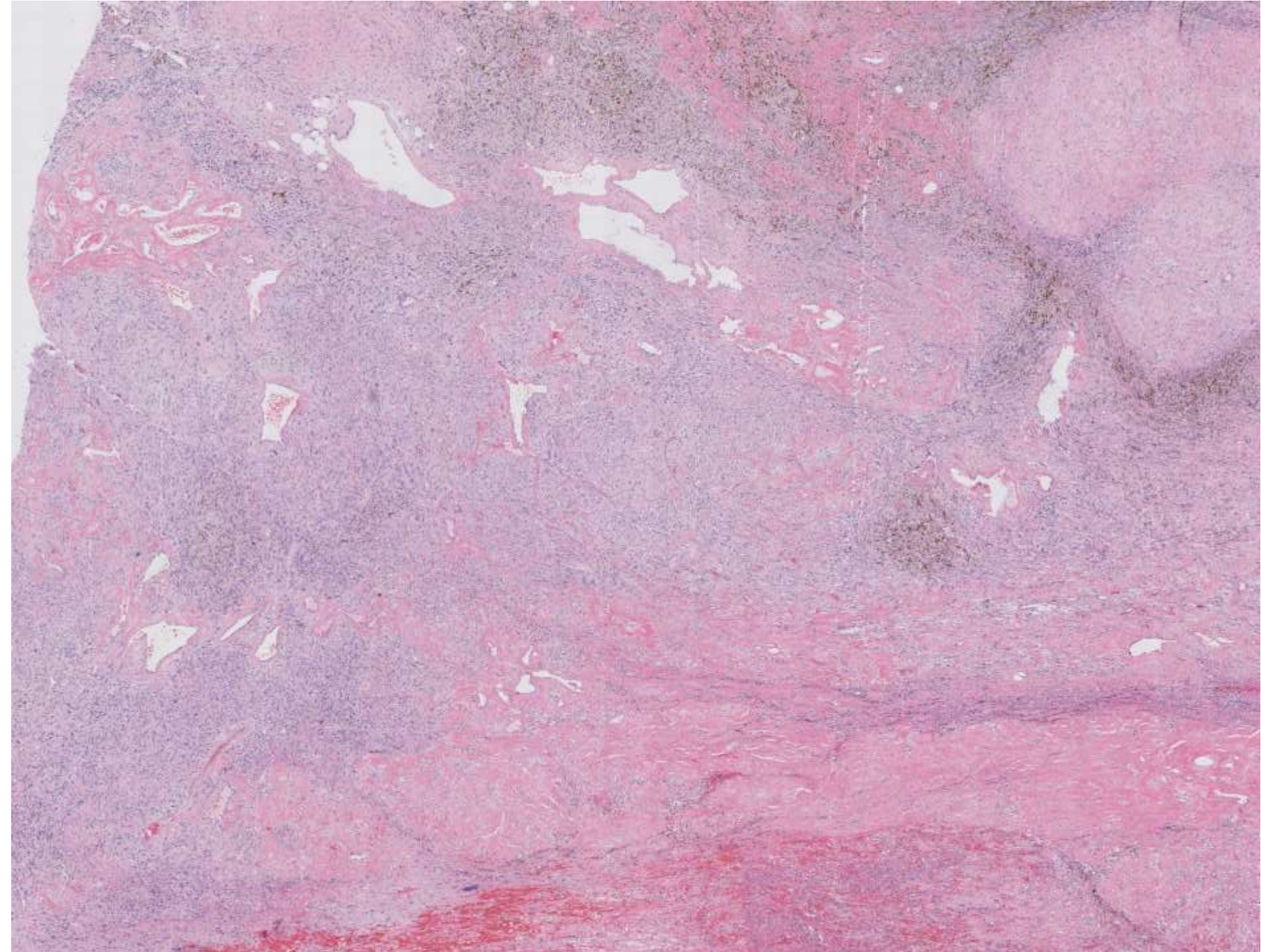
1. Bertoni F, Unni KK, Beabout JW, Sim FH. **Chondrosarcoma of the synovium**. *Cancer* 1991;67:155-162.
2. Kenan S, Abdelwahab IF, Klein MJ, Lewis MM. Case report 817: **Synovial chondrosarcoma secondary to synovial chondromatosis**. *Skeletal Radiol* 1993;22:623-6.
3. Davis RI, Hamilton A, Biggart JD. **Primary synovial chondromatosis: a clinicopathologic review and assessment of malignant potential**. *Human pathology*. 1998;29:683-688.
4. Blokx WA, Rasing LA, Veth RP, Pruszczynski M. **Late malignant transformation of biopsy proven benign synovial chondromatosis: an unexpected pitfall**. *Histopathology* 2000;36:564-6.
5. Campanacci DA, Matera D, Franchi A, et al. **Synovial chondrosarcoma of the hip: report of two cases and literature review**. *La Chirurgia degli organi di movimento*. 2008;92:139-144.
6. Hopyan S, Nadesan P, Yu C et al. **Dysregulation of hedgehog signaling predisposes to synovial chondromatosis**. *J Pathol* 2005; 206:143-150.
7. Wuisman PIJM, Noorda RJP, Jutte PC. **Chondrosarcoma secondary to synovial chondromatosis**. *Arch Orthop Trauma Surg* 1997;116:307-311
8. Wittkop B, Daview AM, Mangham DC. **Primary synovial chondromatosis and synovial chondrosarcoma**. *Eur Radiol* 2002;12:2112-2119.

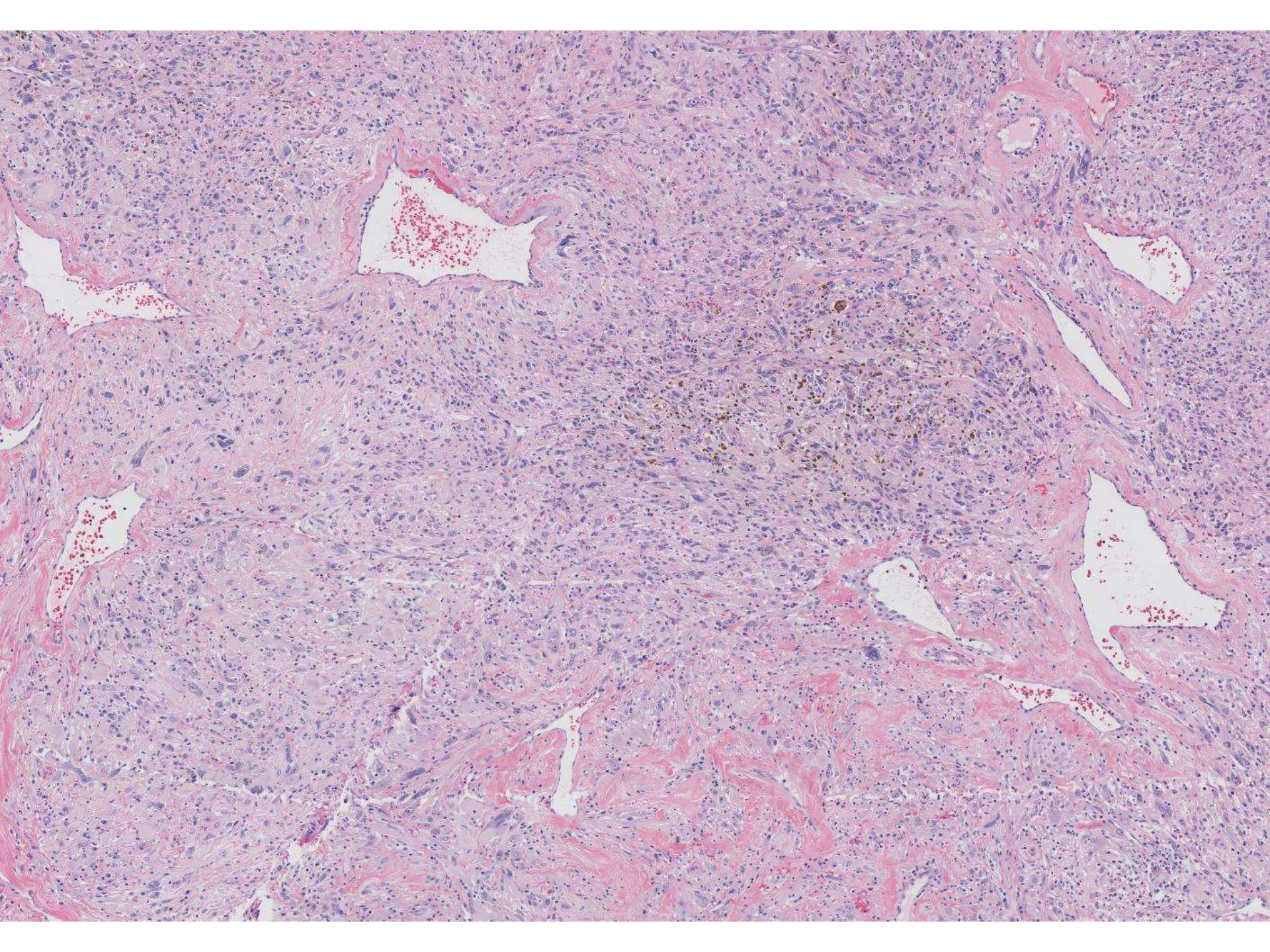
SB 6023

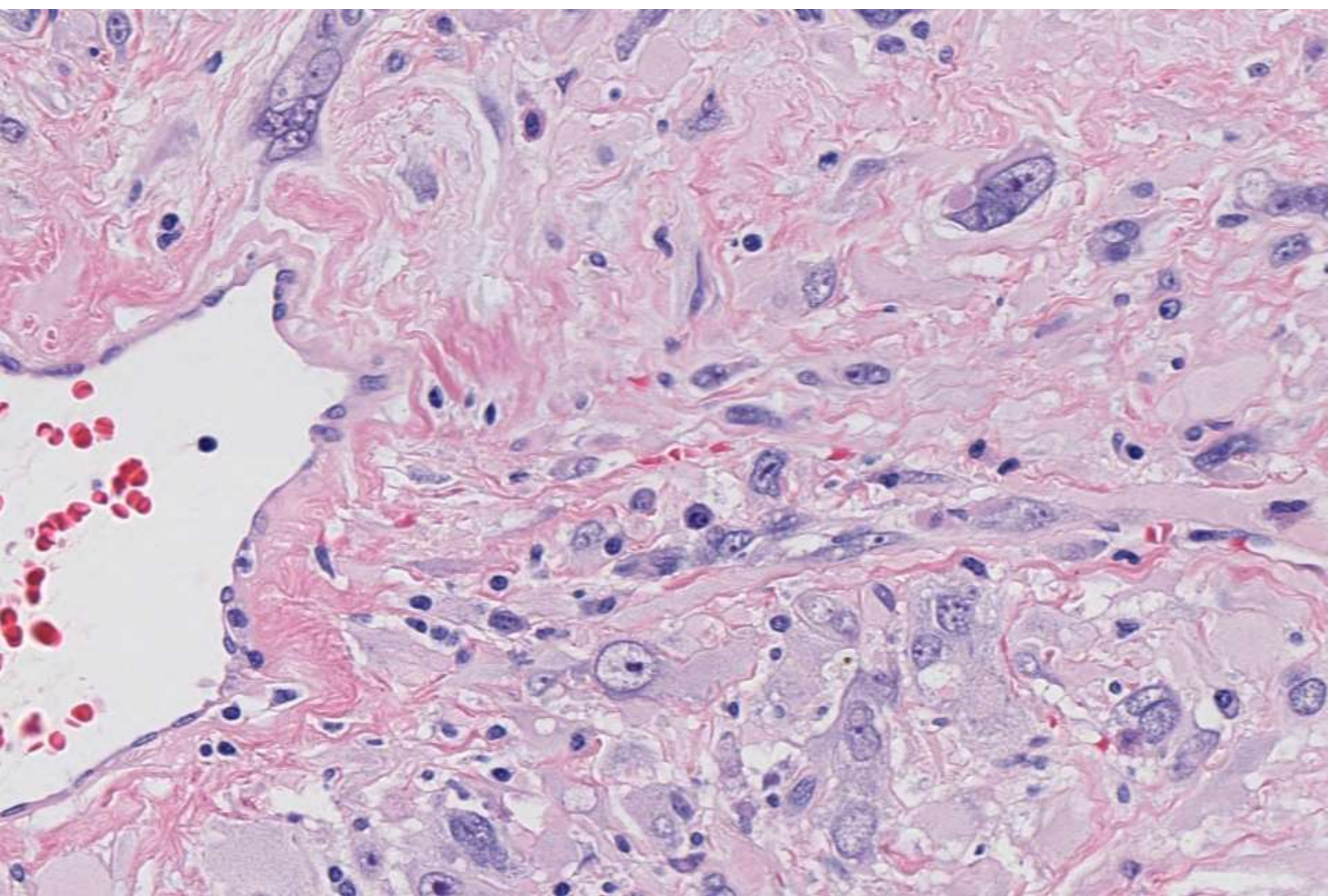
Ben Buelow/Andrew Horvai; UCSF

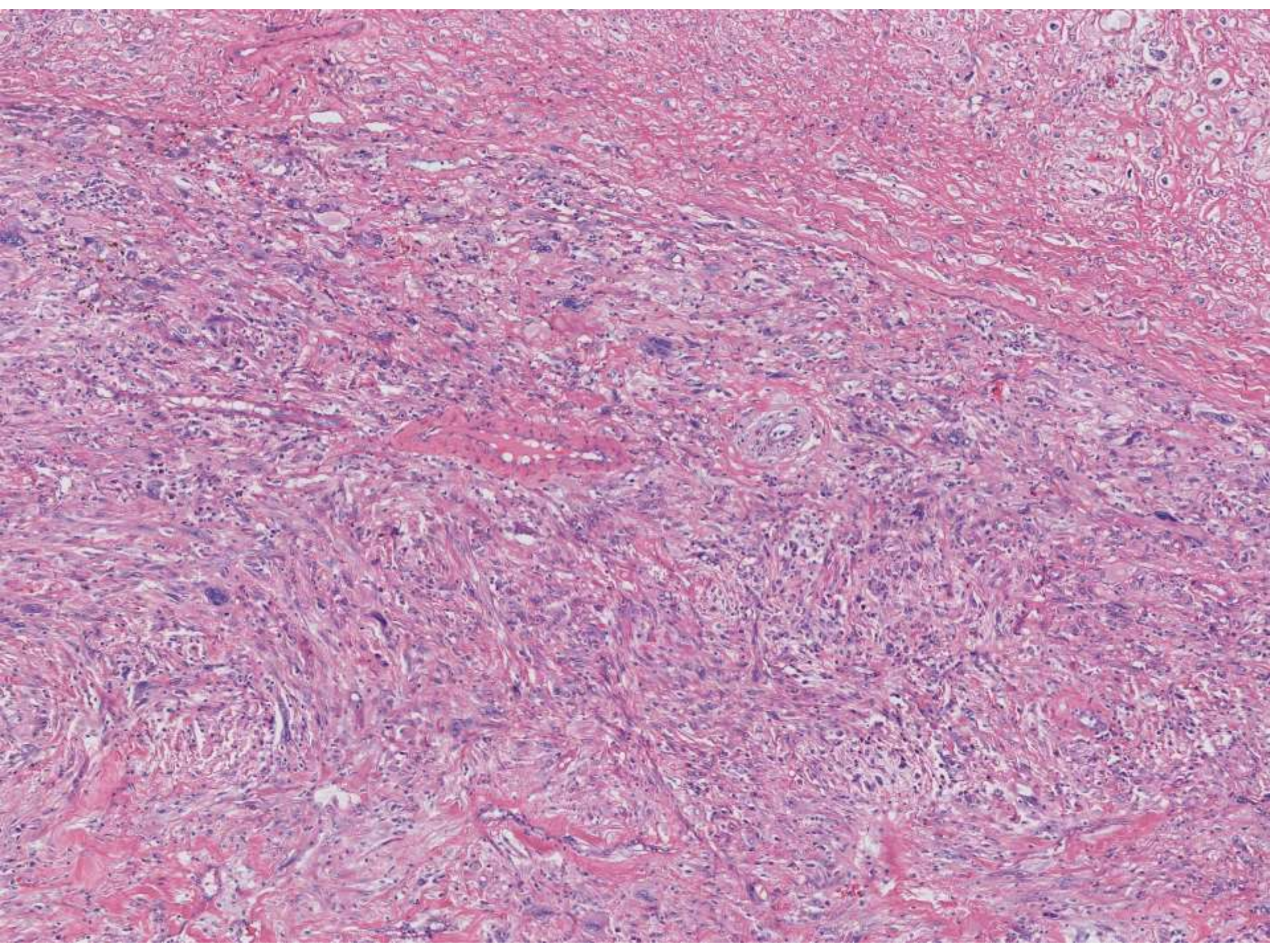
44-year-old man with superficial
lower leg mass.

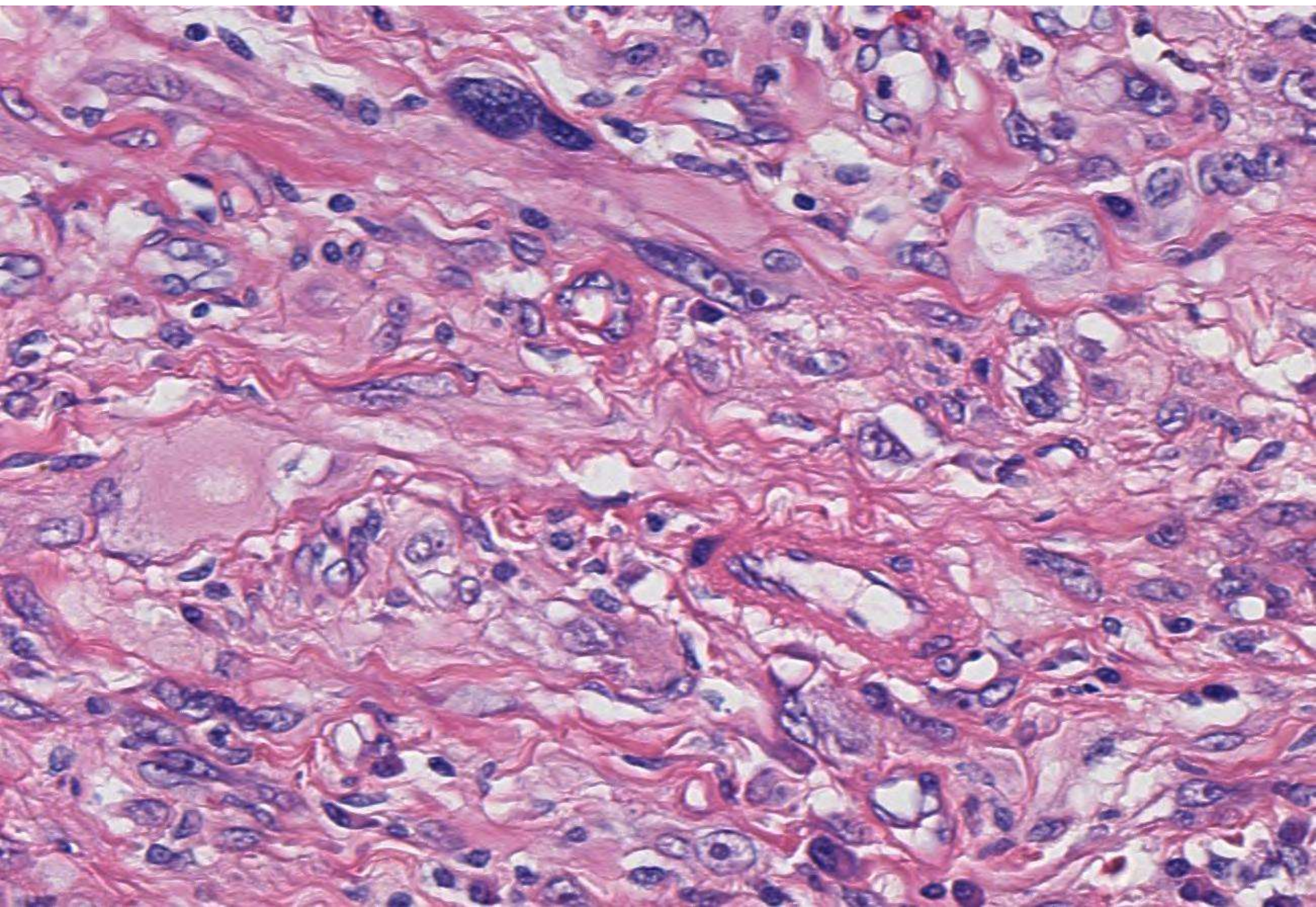








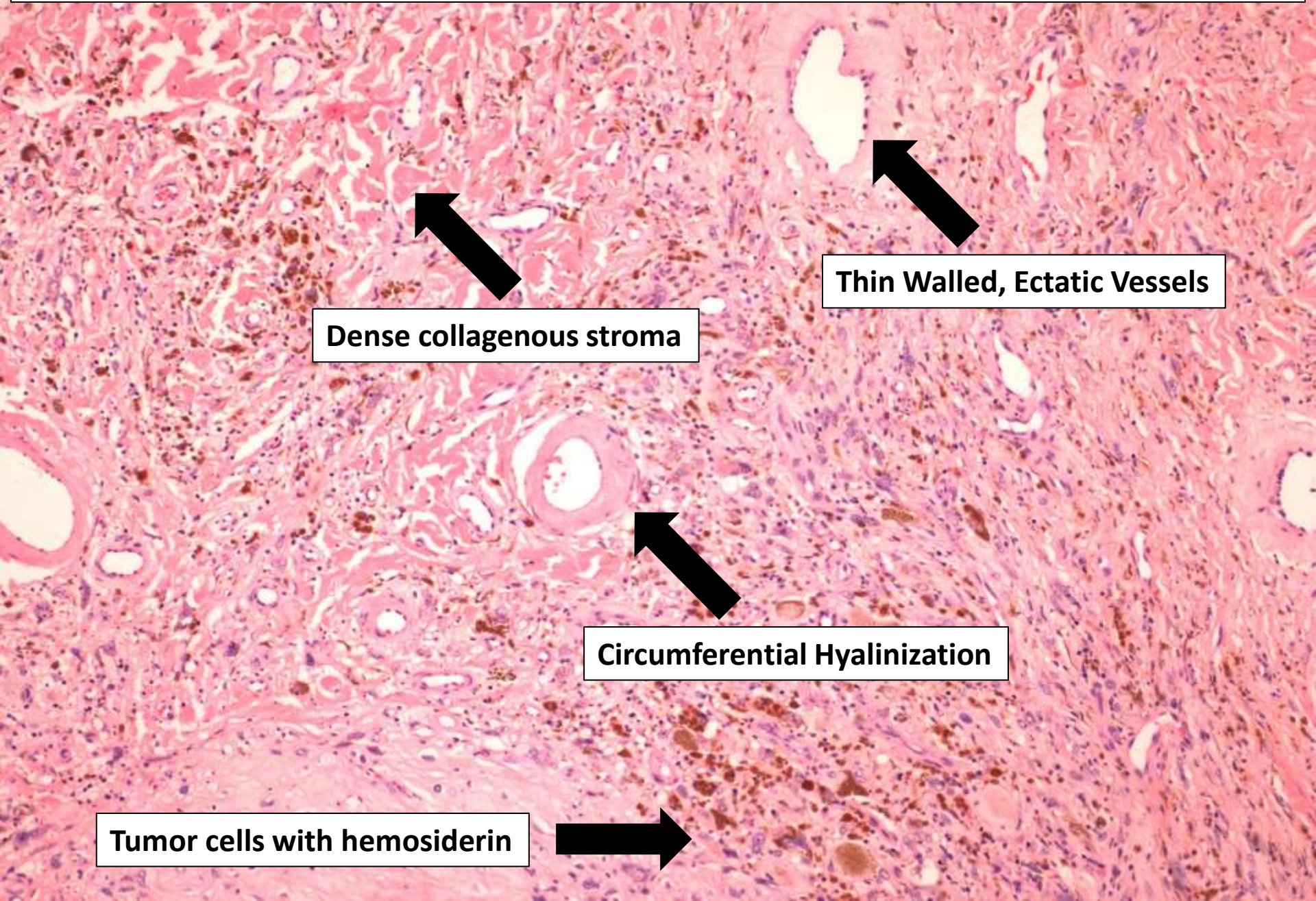




DIAGNOSIS?



Pleomorphic Hyalinizing Angioectatic Tumor (PHAT)

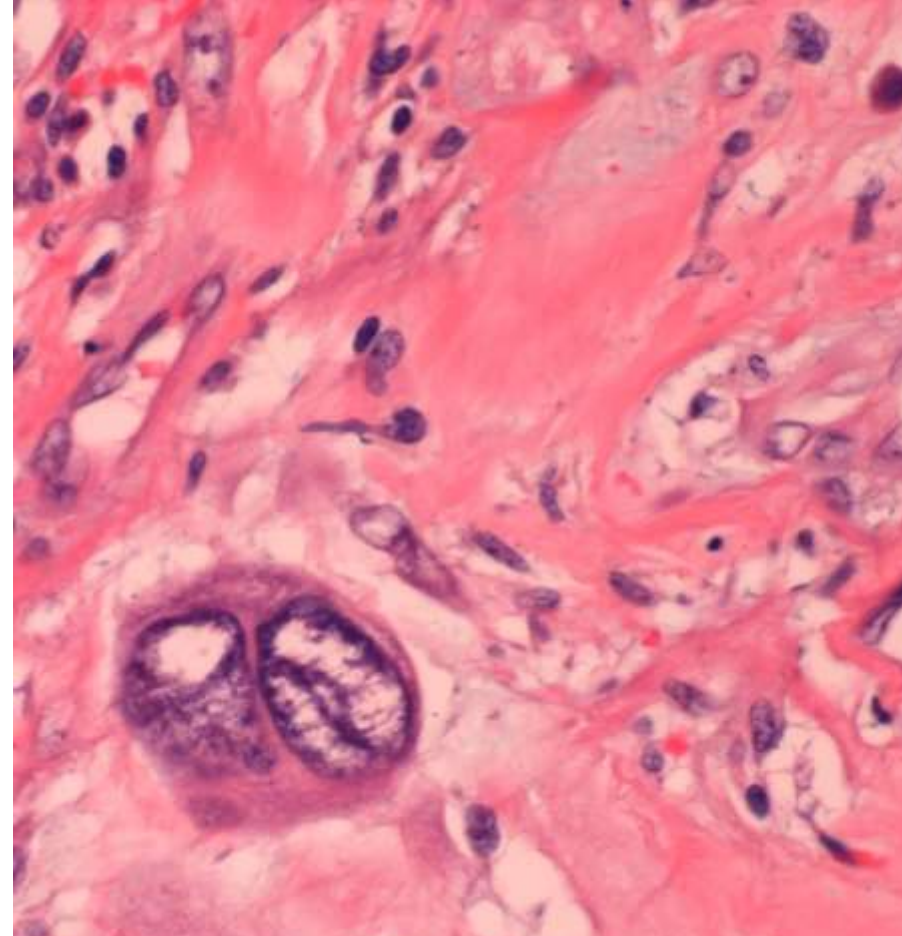
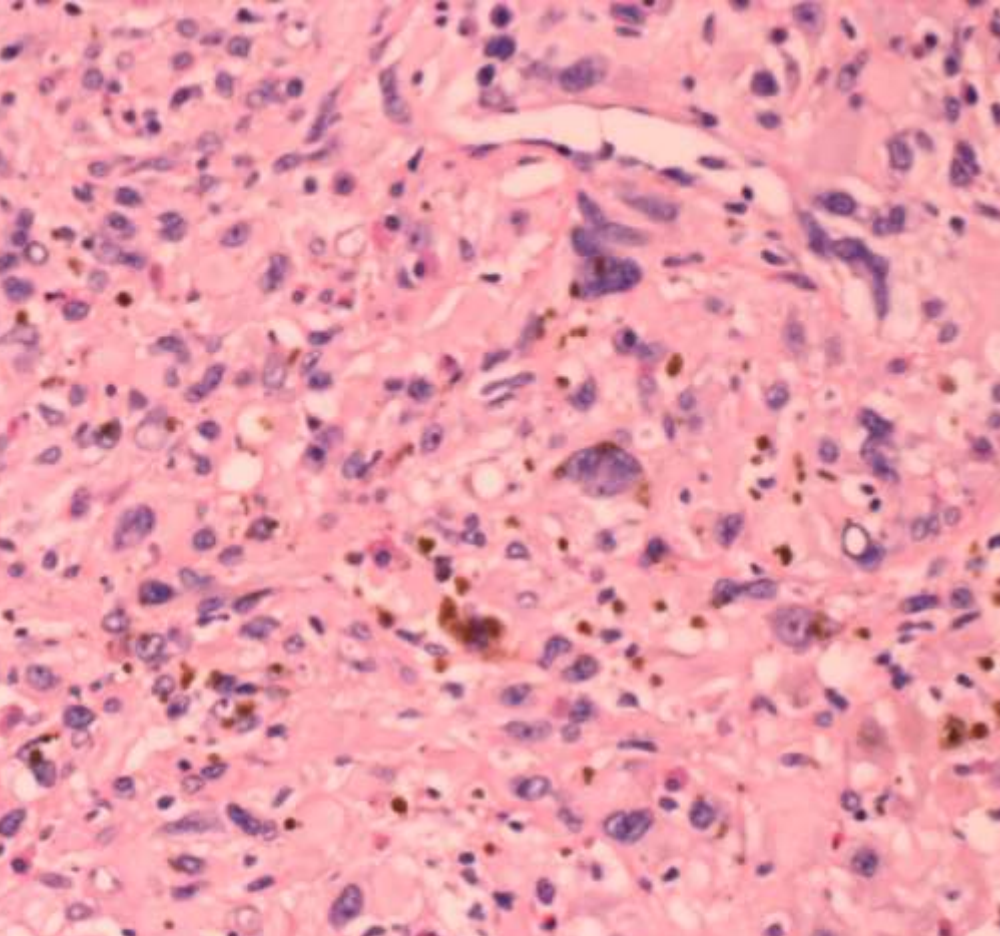


Dense collagenous stroma

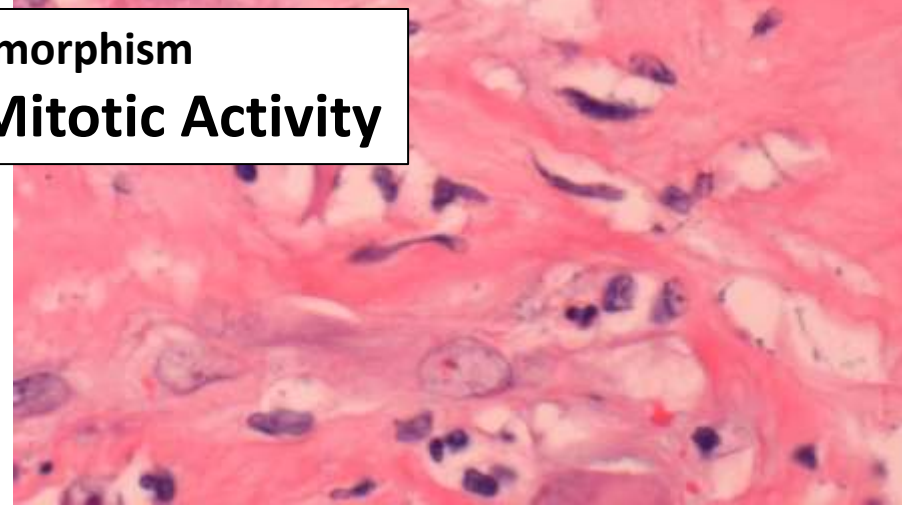
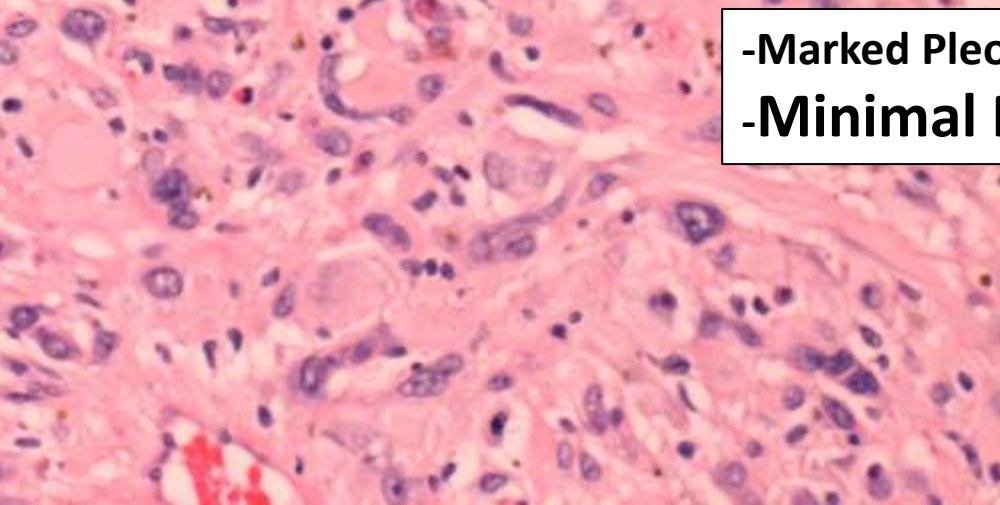
Thin Walled, Ectatic Vessels

Circumferential Hyalinization

Tumor cells with hemosiderin



-Marked Pleomorphism
-Minimal Mitotic Activity



Pleomorphic Hyalinizing Angioectatic Tumor (PHAT): Differential Diagnosis

- Pleomorphic Undifferentiated Sarcoma
 - Necrosis and Mitotic Activity
- Schwannoma
 - Immunohistochemistry
- Angiomatoid Fibrous Histiocytoma
 - Pleomorphism
- **Hemosiderotic Fibrohistiocytic Lipomatous Lesion (HFLT)**
- **MyxoInflammatory Fibroblastic Sarcoma (MIFS)**

Pleomorphic Hyalinizing Angioectatic Tumor (PHAT): Clinical

- Adults
- Slow growing
- Subcutaneous/Suprafascial
- Lower extremity
- Frequent (30-50%) recurrence
- No metastasis

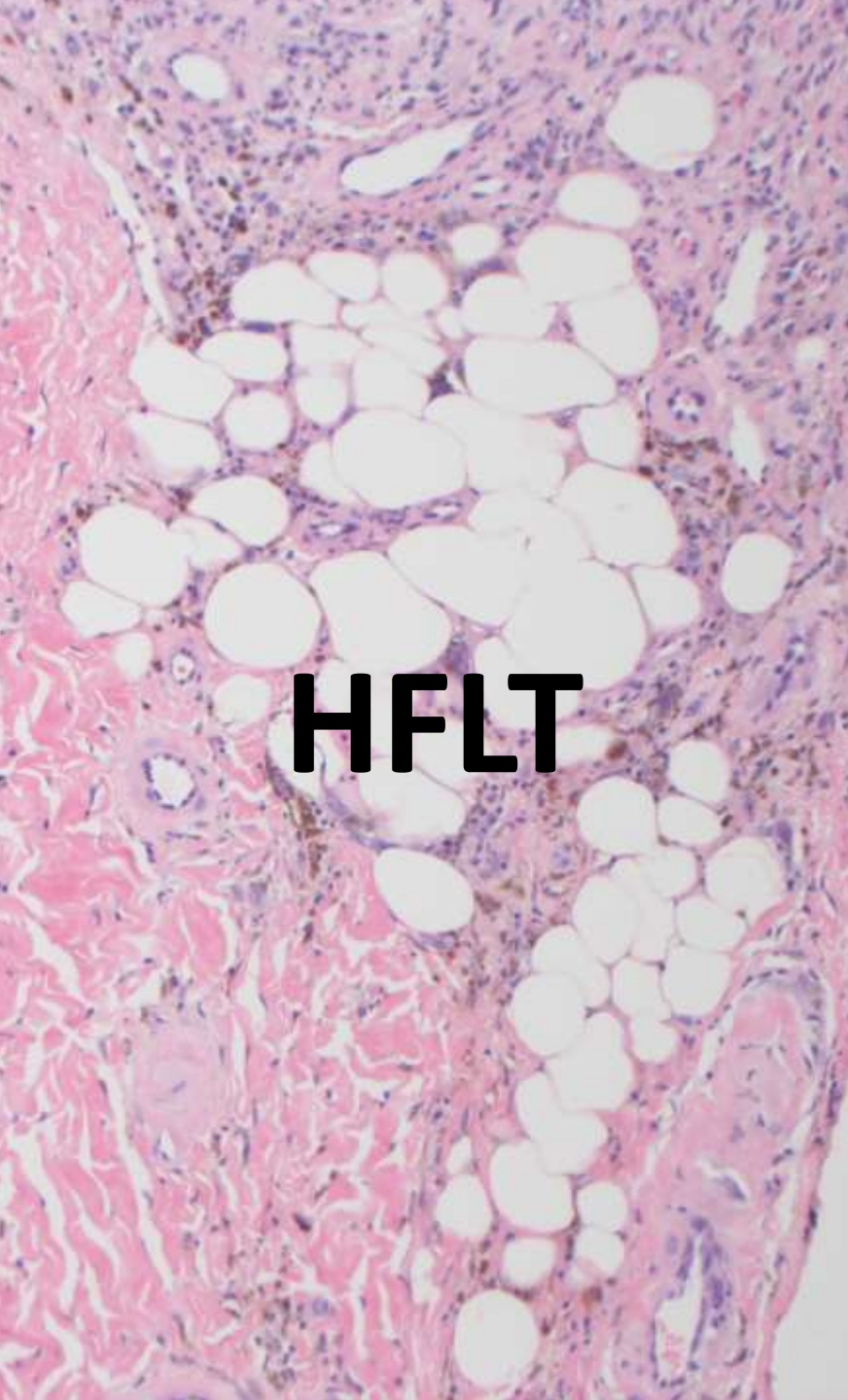
Pleomorphic Hyalinizing Angioectatic Tumor (PHAT): IHC

Positive

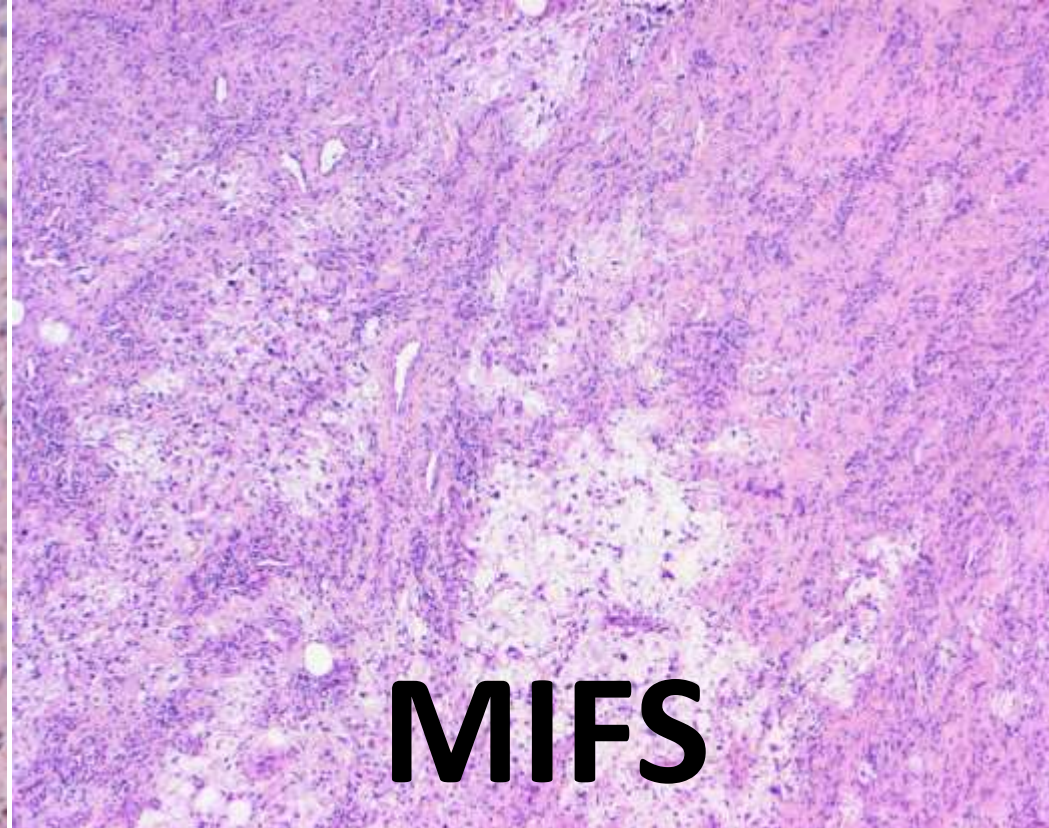
- CD34
- Vimentin
- +/- Factor 13a

Negative

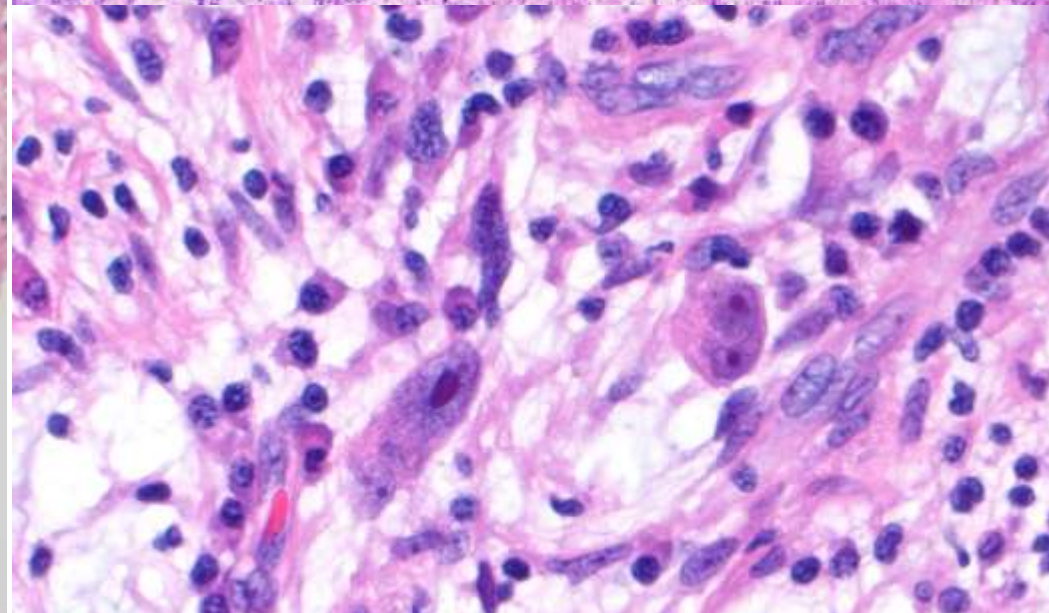
- S100
- Actin
- Desmin
- Keratin
- EMA
- CD31



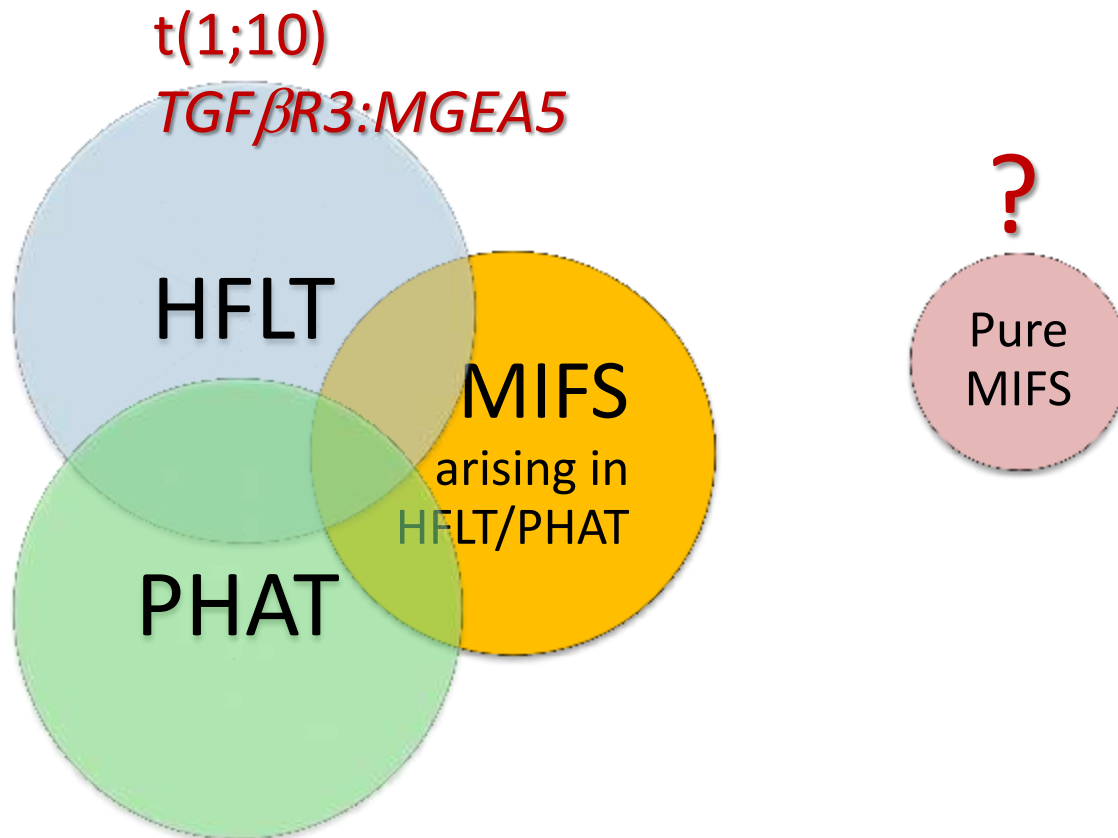
HFLT



MIFS



HFLT-PHAT-MIFS



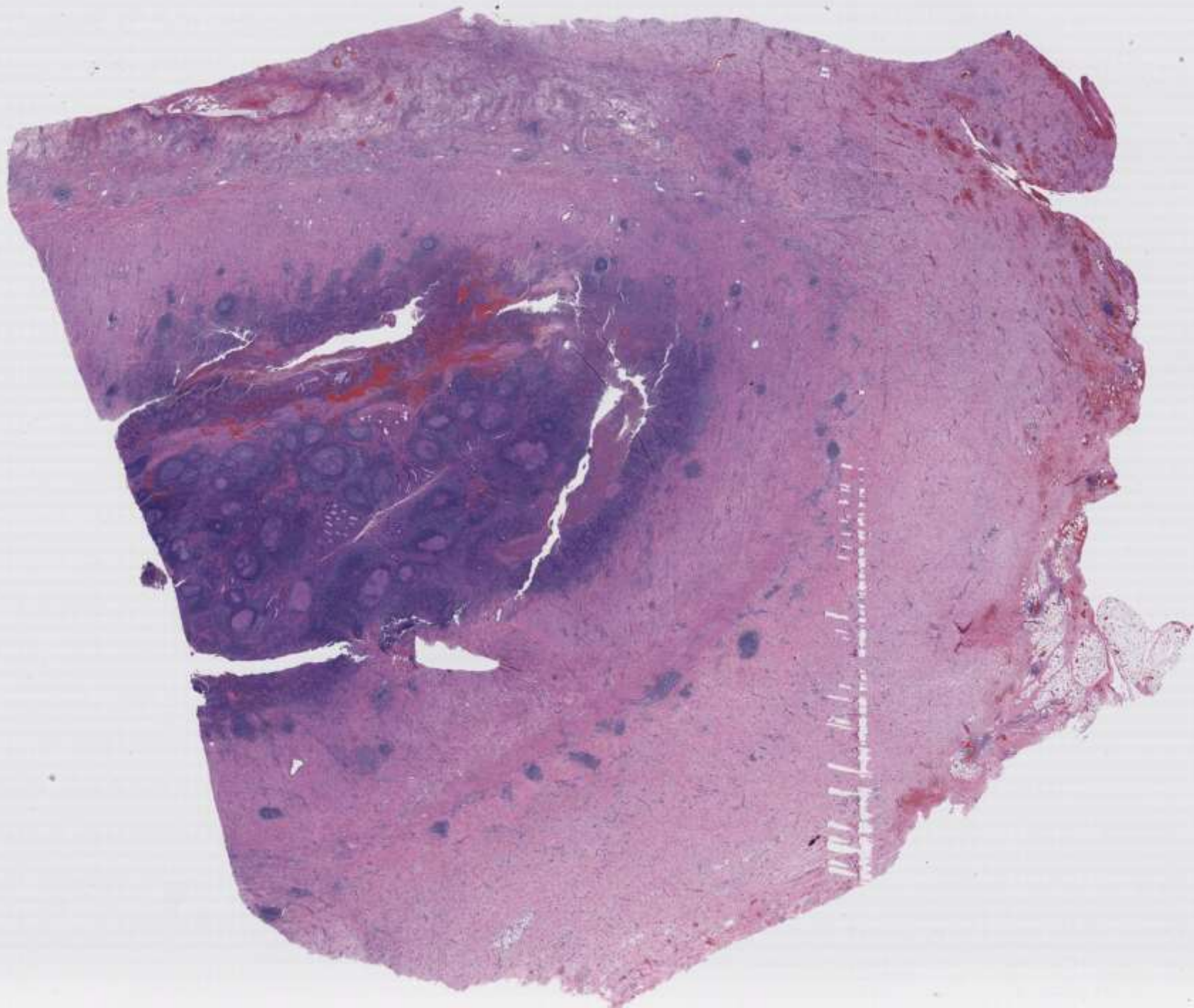
HFLT = Hemosiderotic fibrolipomatous tumor
PHAT = Pleomorphic hyalinizing angiectatic tumor
MIFS = Myxoinflamamatory fibroblastic sarcoma

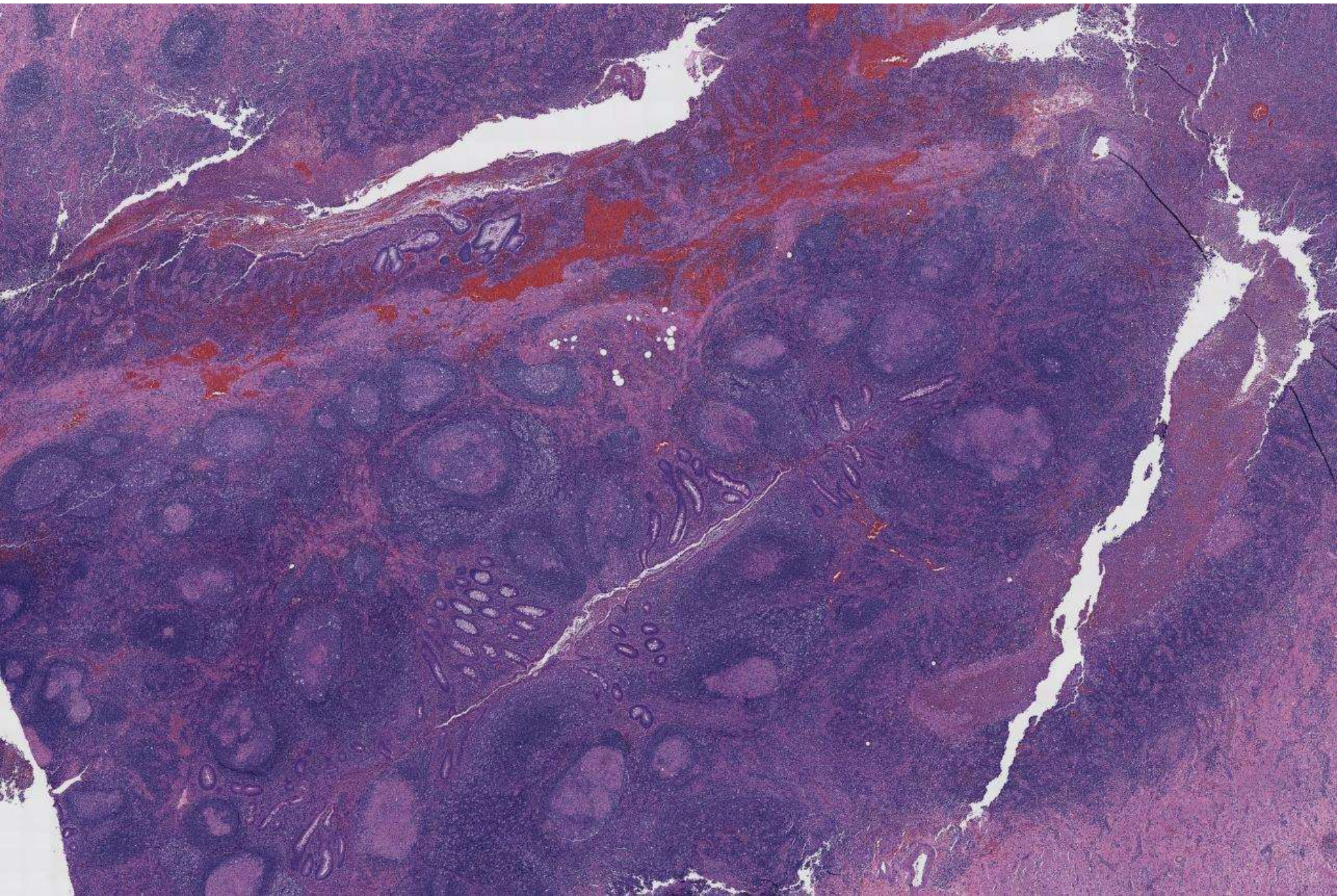
Pleomorphic Hyalinizing Angioectatic Tumor (PHAT)

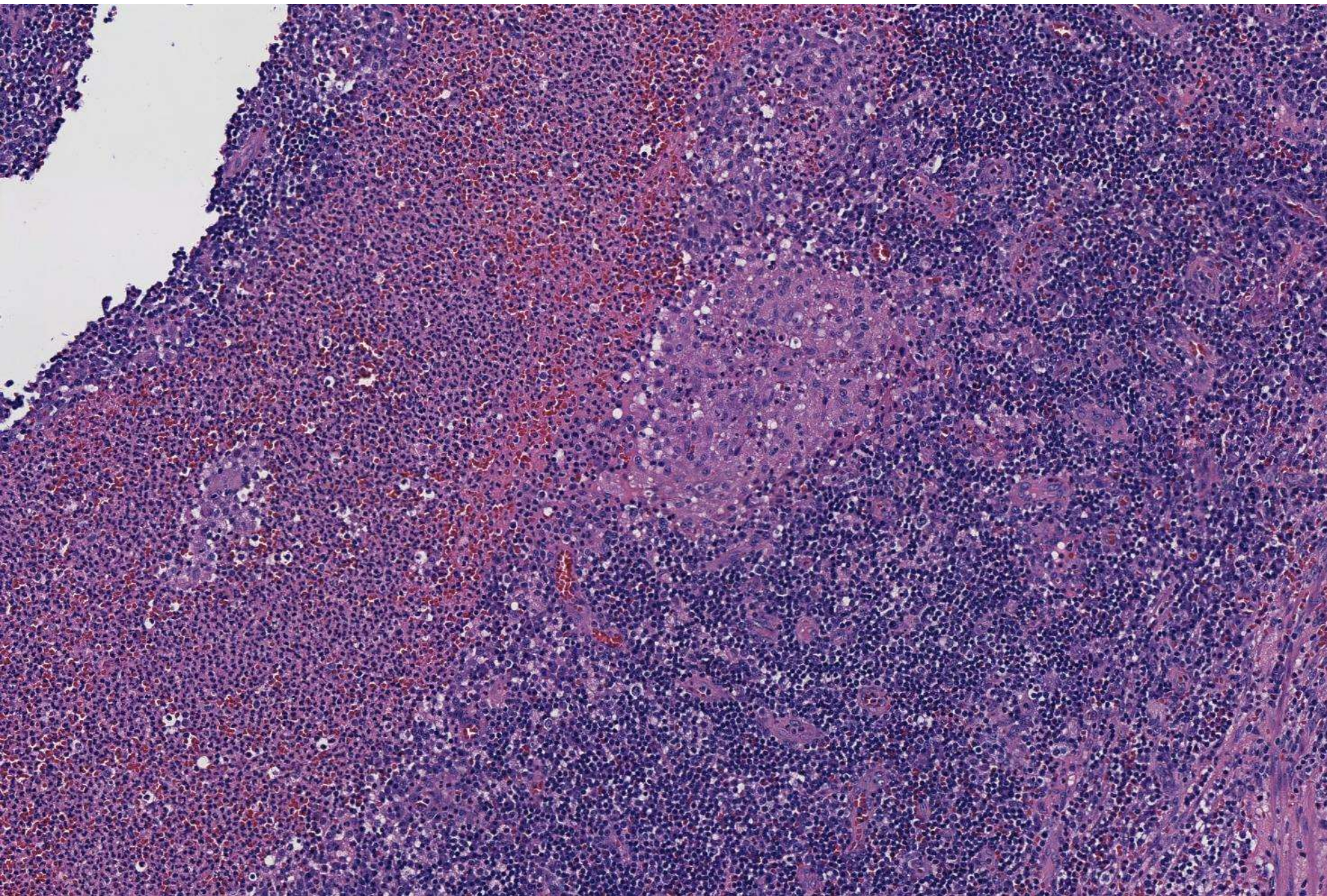
- **Clinical**
 - Lower extremity, slow growing
 - Frequent recurrence, no metastasis.
- **Morphology**
 - Ectatic, hyalinized small vessels, hemosiderin, marked pleomorphism, low mitotic rate
 - Consider this in cytology!
- **PHAT-HFLT-MIFS family**
 - T(1:10) TGFB3-MGEA5
 - HFLT: Prominent lipomatous component
 - MIFS: Myxoid/inflammatory areas, virocyte-like cells, infiltrative

SB 6024

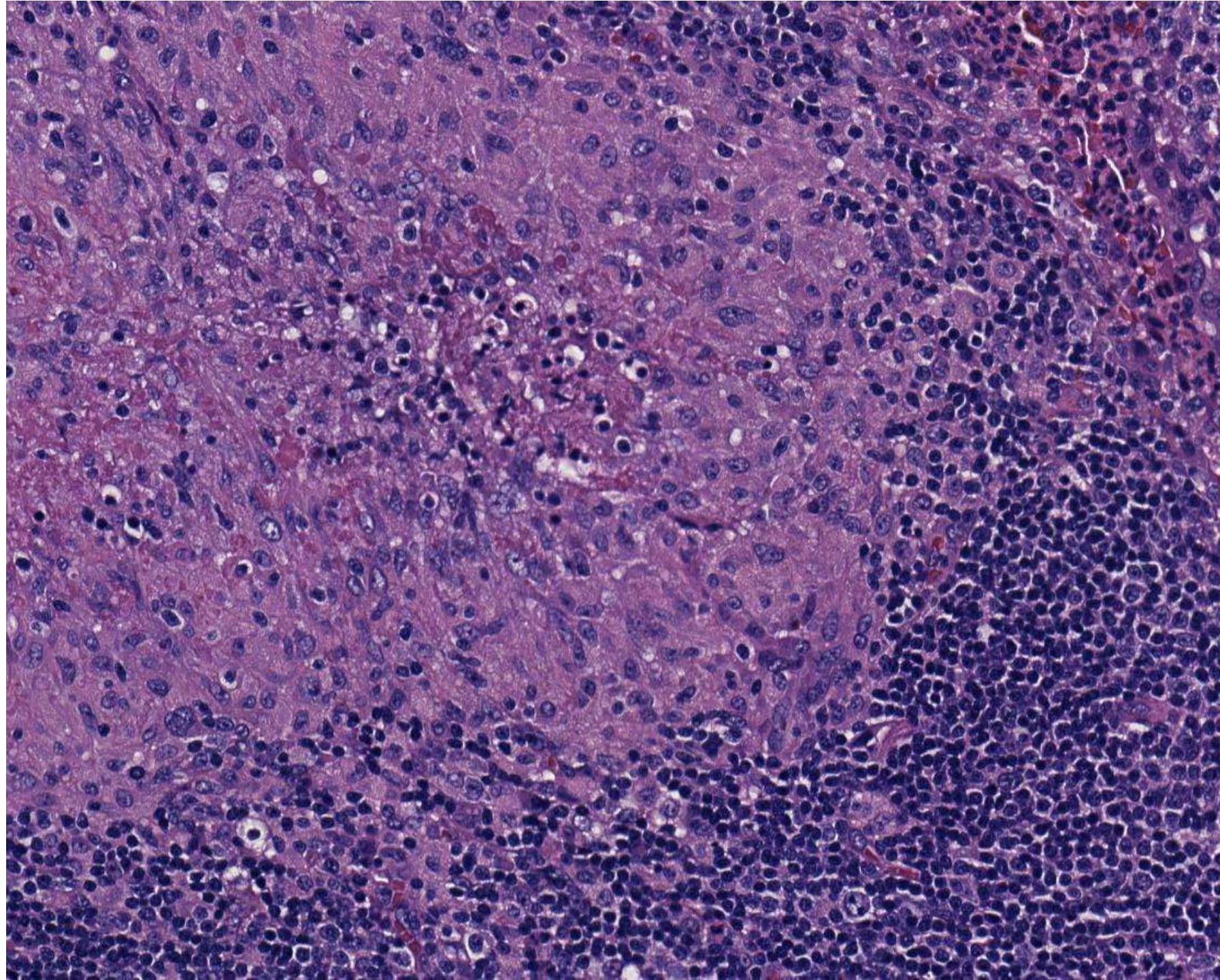
Mahendra Ranchod; Good Samaritan Hospital
7-year-old boy had an appendectomy for acute
appendicitis.











DIAGNOSIS?



Granulomatous appendicitis

- Crohn's disease
- Sarcoidosis
- Infections
 - Mycobacterial
 - Fungal
 - Parasitic
 - Yersinia
- Foreign body reaction
- Interval appendectomy
 - Granulomatous
 - Xanthogranulomatous
 - Crohn's-like
- Other

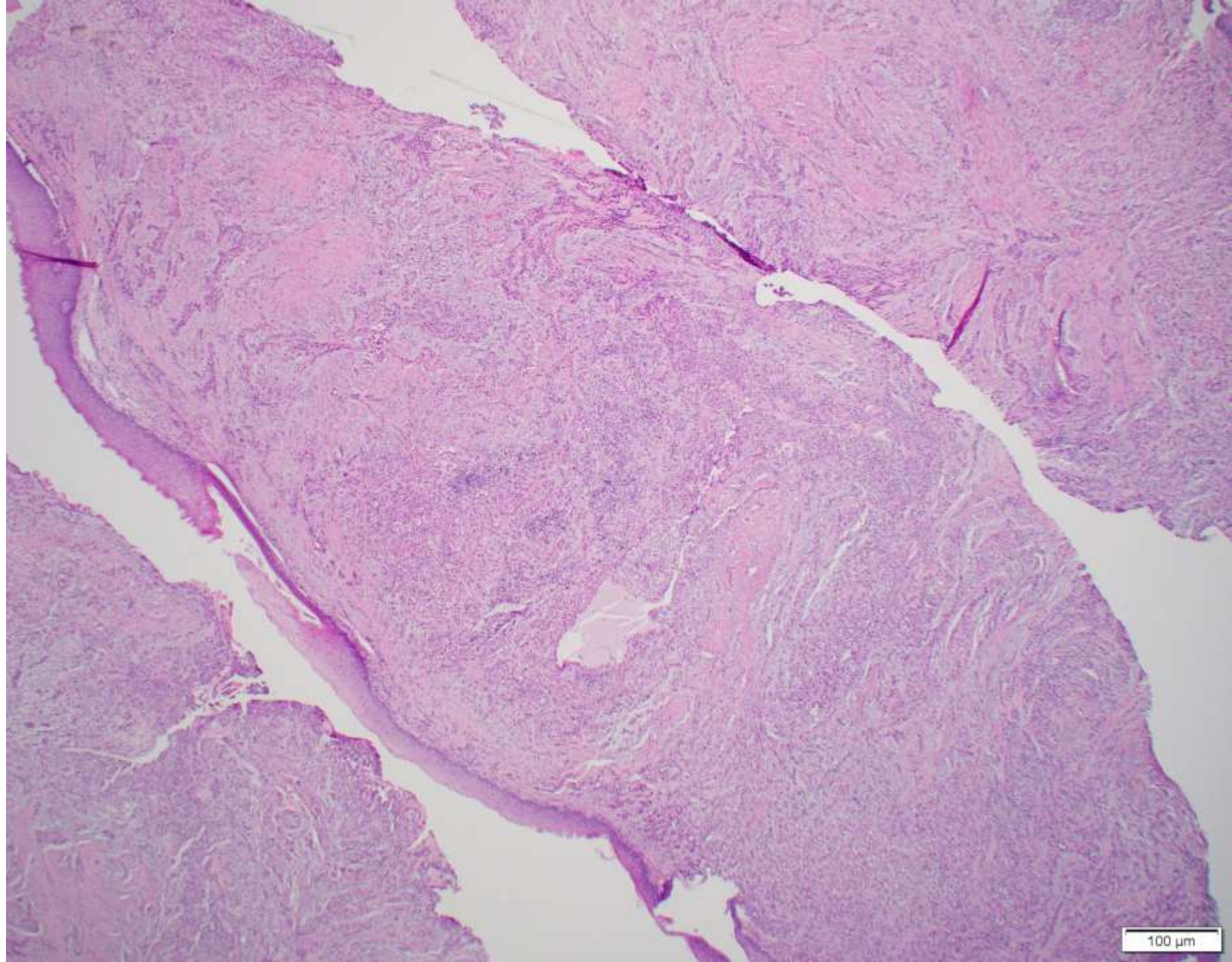
A few points about Isolated Granulomatous Appendicitis

- **Be circumspect about making a diagnosis of Crohn's disease**
- **Check on history: ? interval appendectomy**
- **Consider Yersinia when:**
 - **Short history of symptoms**
 - **Acute or subacute appendicitis + granulomas**
 - **Many granulomas**

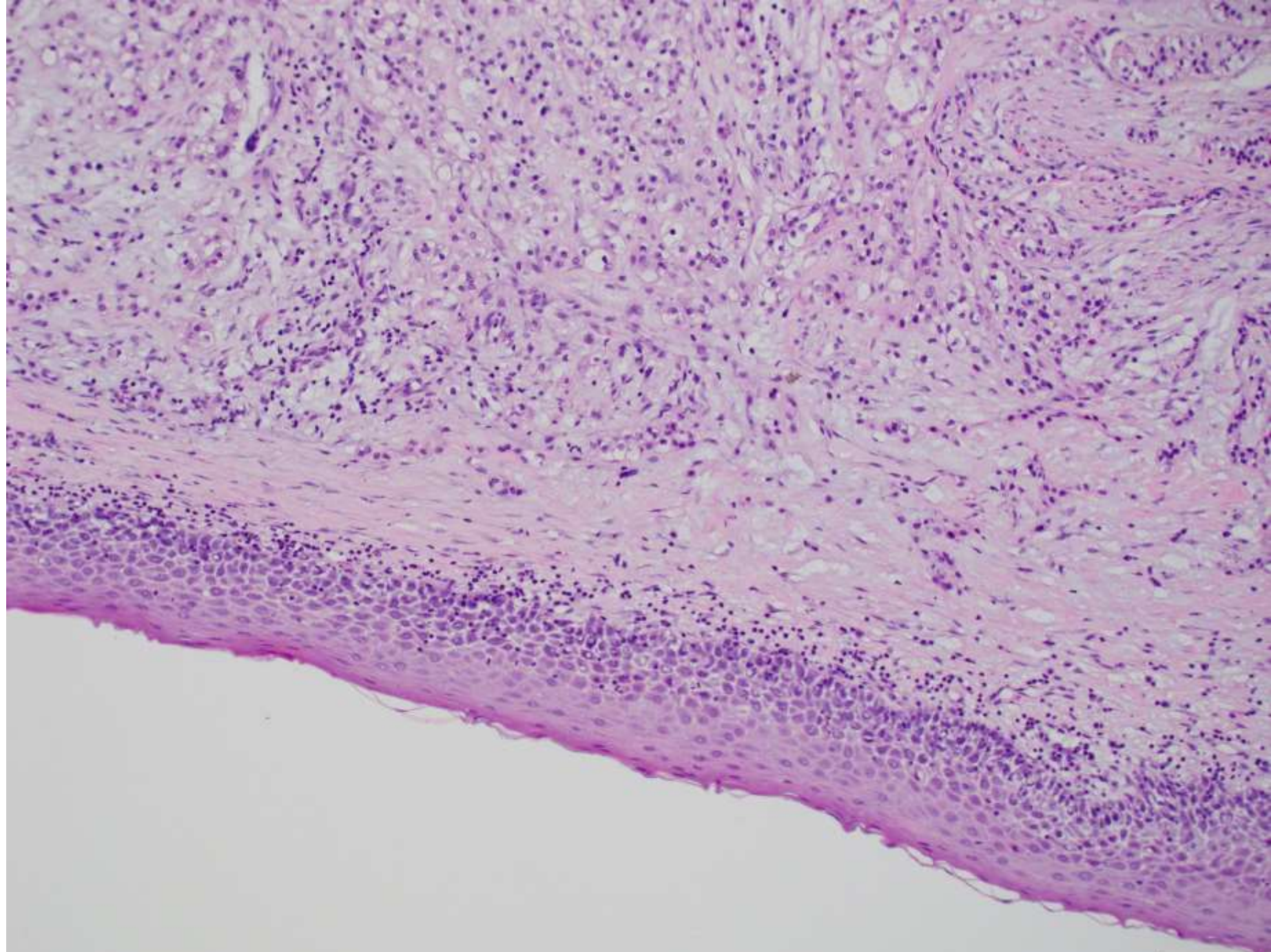
SB 6025

Sarah Cherny; Kaiser San Francisco

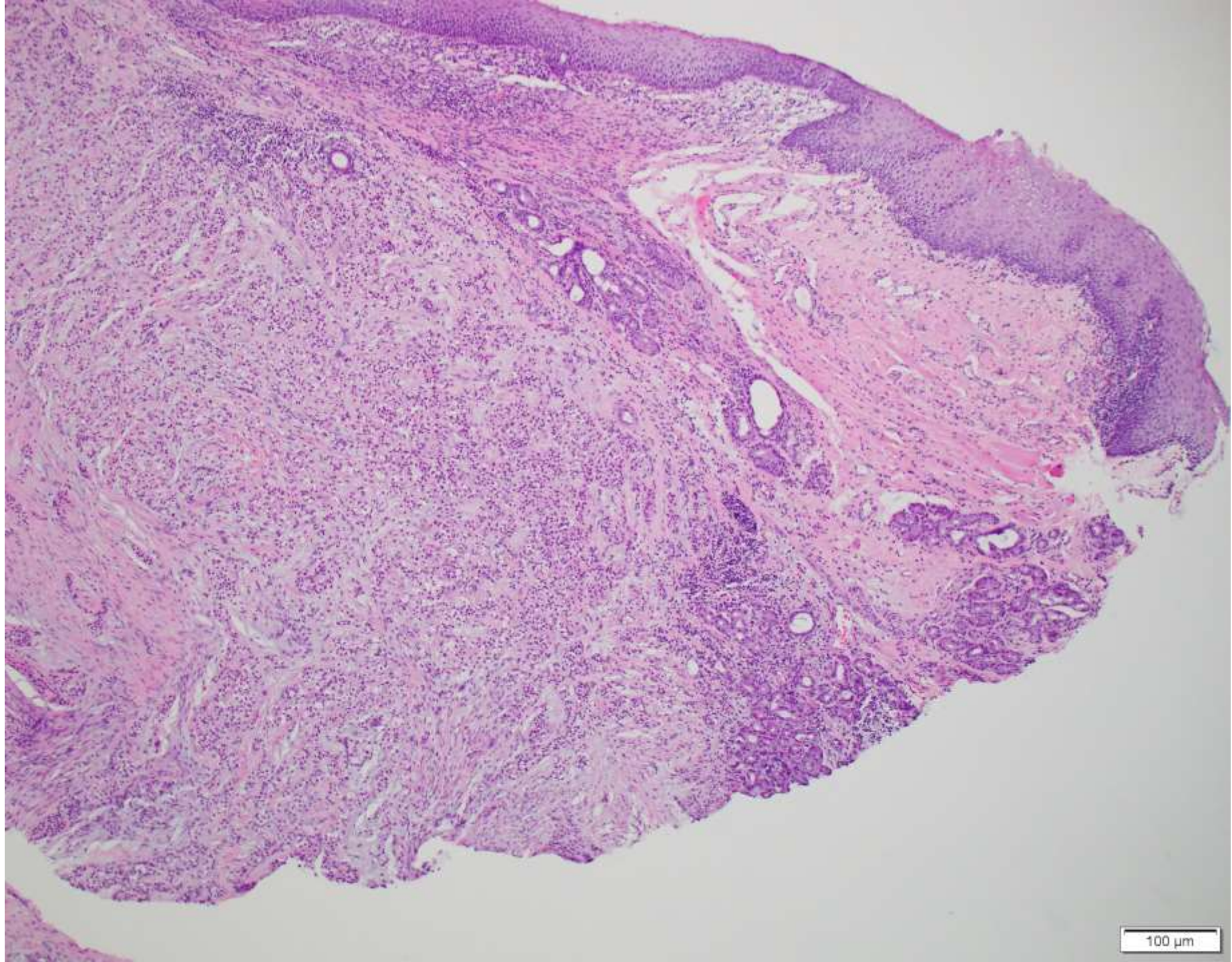
33-year-old pregnant woman (2nd trimester)
with new 1.2cm nodule on right posterolateral
tongue. Shave biopsy performed.

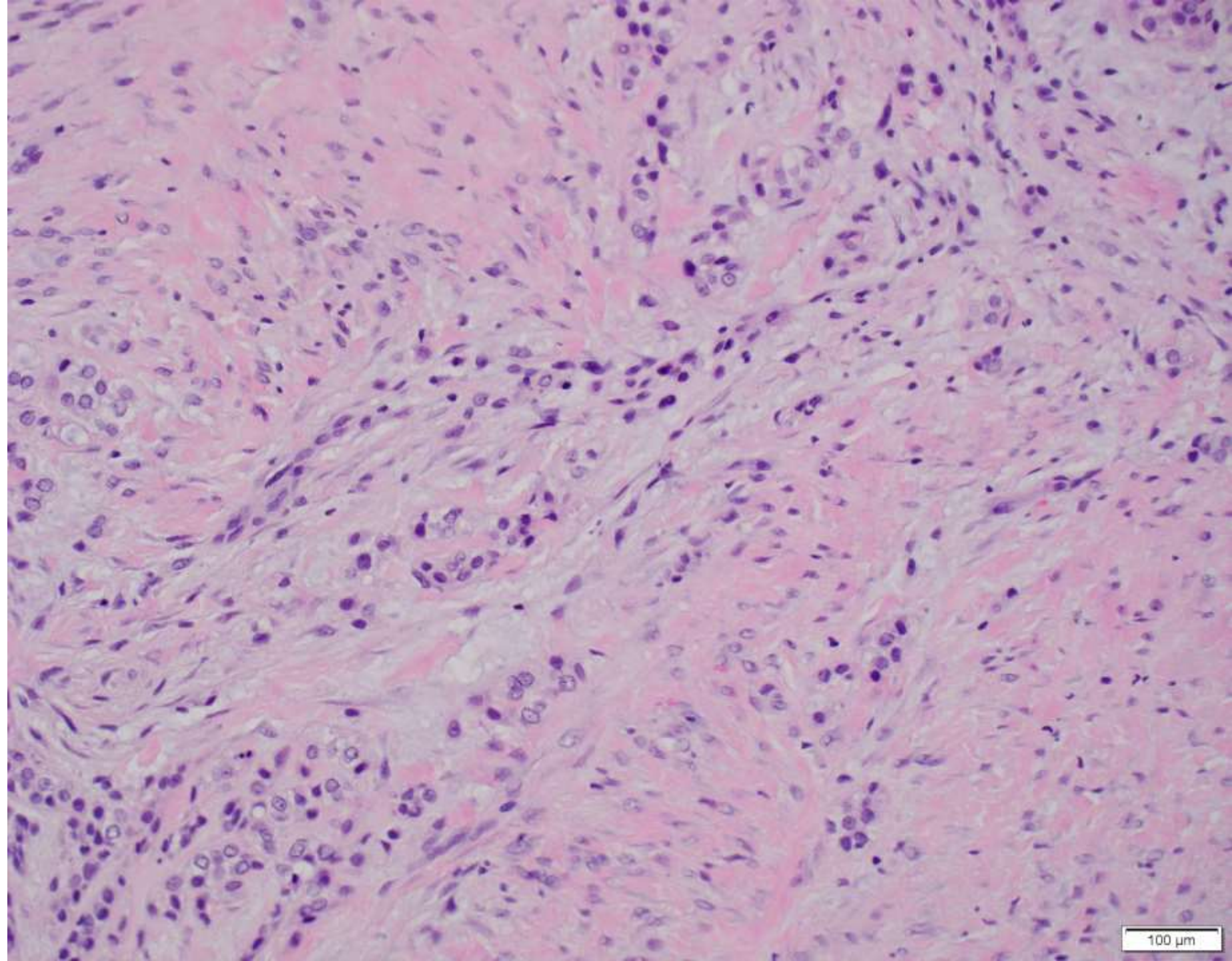


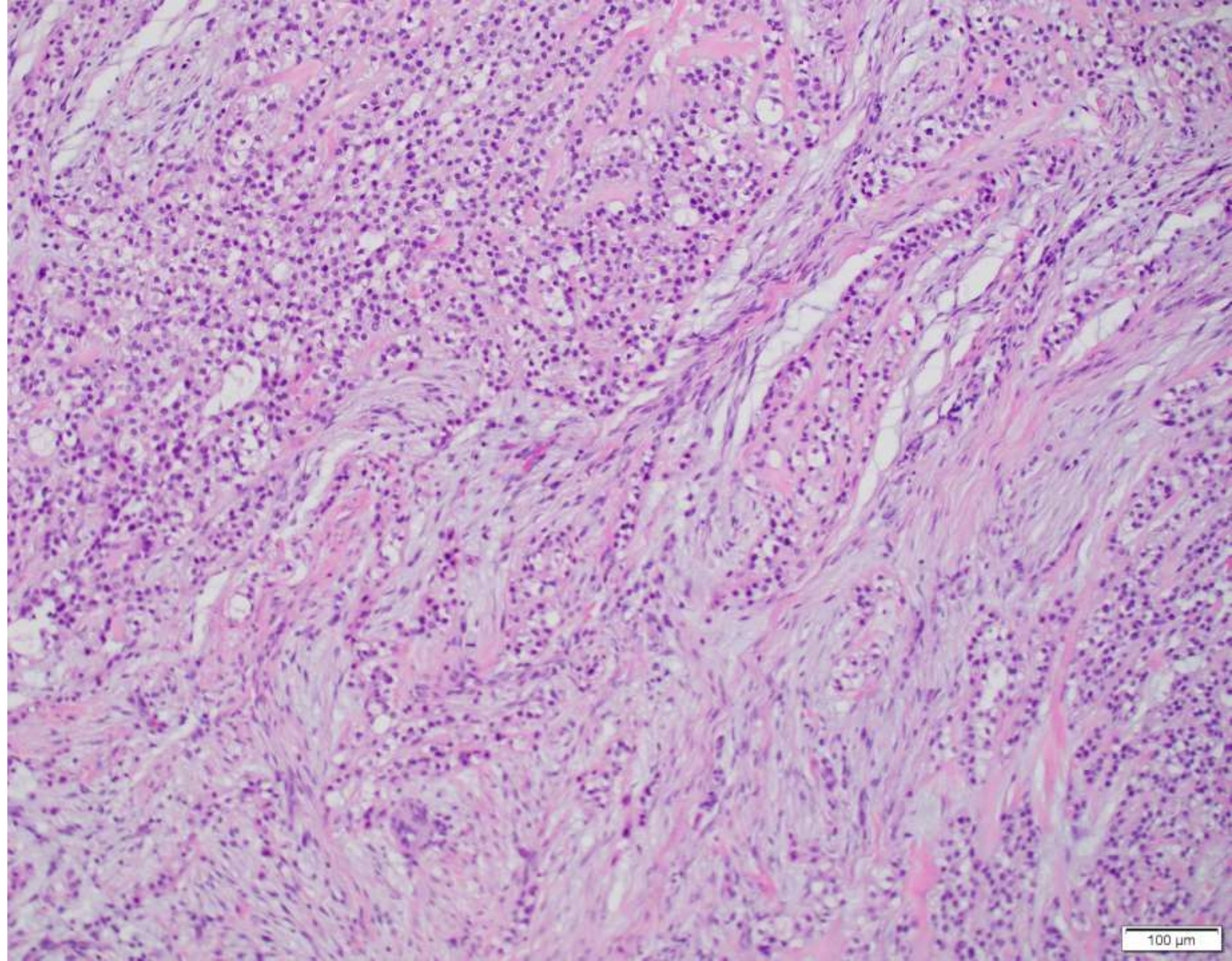
100 μm



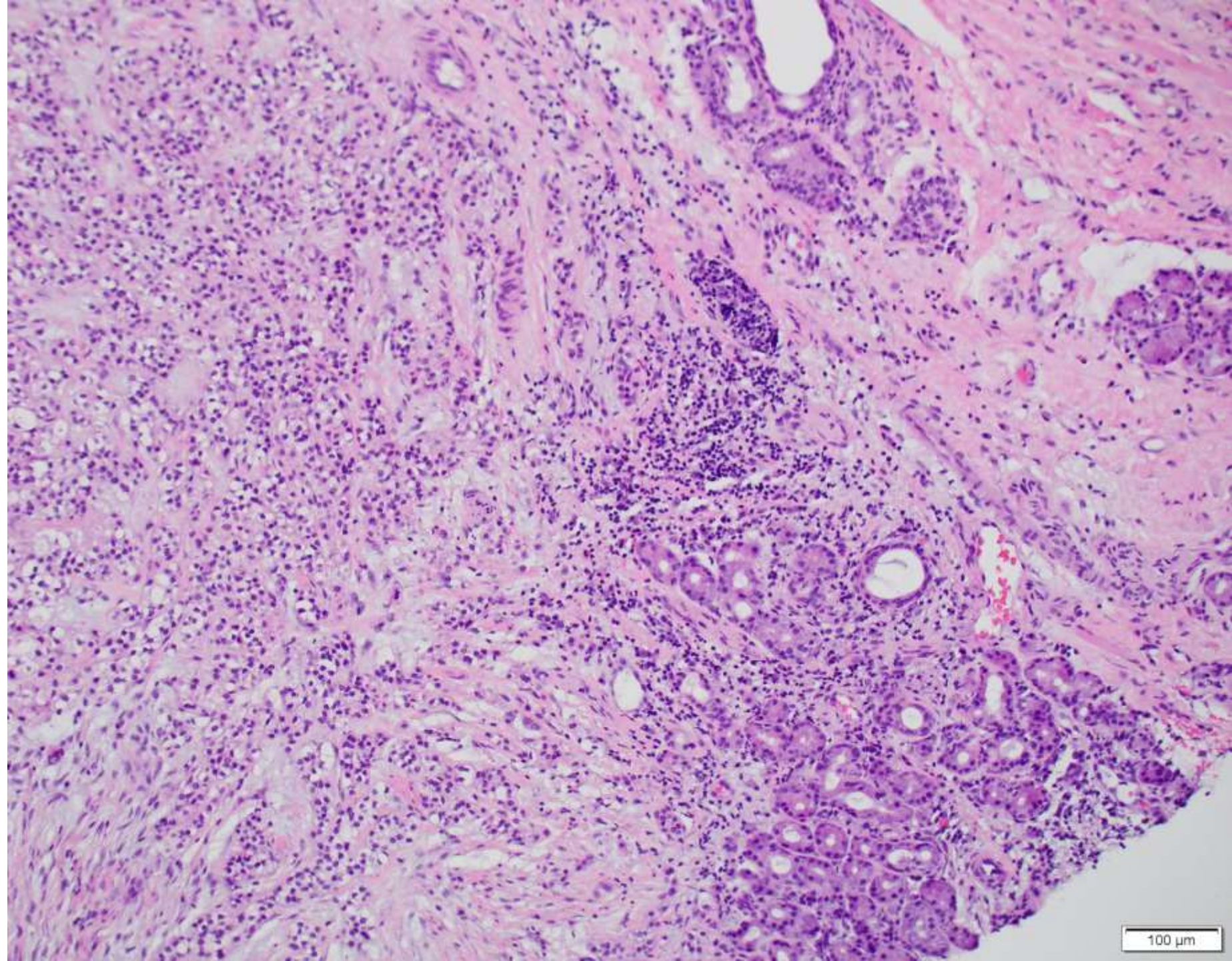
100 μ m

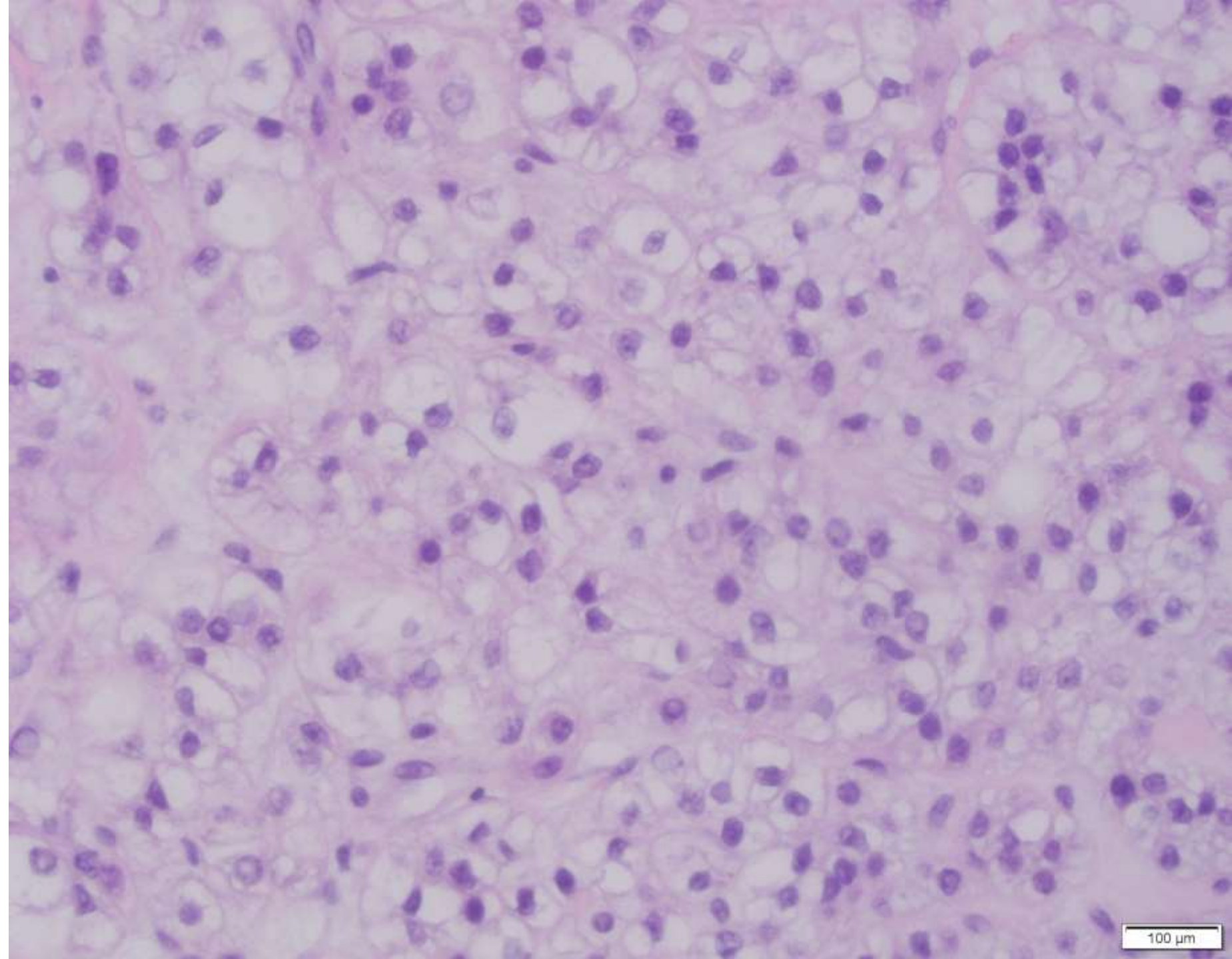






100 μ m





DIAGNOSIS?



Case 5:
33 yo woman, 17 weeks pregnant,
with tongue mass

Sarah Cherny
Kaiser SSF

Differential Diagnosis

- Hyalinizing Clear Cell Carcinoma
 - AKA Clear Cell Carcinoma, NOS (WHO)
 - AKA Clear Cell Adenocarcinoma (AFIP)
- Clear cell change in a salivary gland tumor (e.g., clear cell variants of myoepithelioma, myoepithelial CA, mucoepidermoid CA)
- Much less likely (given young age and lack of relevant history): metastasis

Hyalinizing Clear Cell Carcinoma

- Rare, infiltrative, low-grade salivary gland neoplasm, first described in 1993 (AJSP, 1994; 18:74-82)
- Intraoral minor salivary glands >> major salivary glands
 - Palate > buccal mucosa, tongue, floor of mouth, lip, retromolar and tonsillar areas
- Grows as nests, sheets, cords of clear cells within a desmoplastic and hyalinized stroma
 - Ductal structures are not seen
 - Unencapsulated, infiltrative borders
 - ALI not characteristic, low mitotic counts, and PNI in ~40%

Hyalinizing Clear Cell Carcinoma

- Peak occurrence in patients in 40-70 year age range; M=F
- Swelling > ulceration and/or pain, 1 month to 15 years
- Size usually <3 cm, but poorly circumscribed and infiltrative

Stains...

- Immunoprofile – not specific:
 - Positive: keratin, p63
 - Negative to focal & weak: S100, actin, vimentin
- Clear cells contain glycogen (PAS+)
- Mucin stains negative

EWSR1 rearrangements

- Described in variety of tumors
 - Ewing sarcoma, DSRCT, myxoid liposarcoma, extraskeletal myxoid chondrosarcoma, clear cell sarcoma, angiomatoid fibrous histiocytoma, soft tissue myoepithelial tumors
- Characterizes large majority of HCCC
 - EWSR1-ATF fusion transcript t(12;22)
 - Not identified in other salivary gland tumors
 - However, also present in clear cell odontogenic carcinoma
 - Differentiate by location

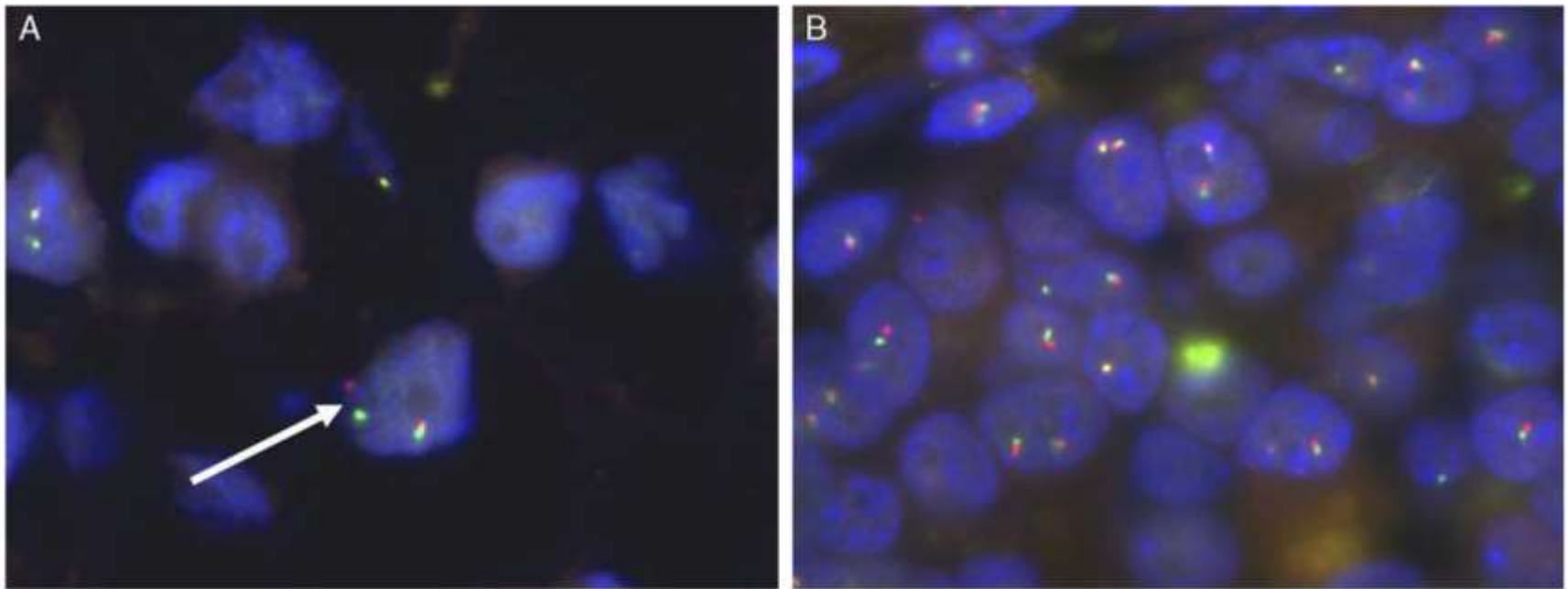


FIGURE 3. Interphase nuclei demonstrating intact and disrupted (arrow) *EWSR1* signals. The majority of HCCCs showed *EWSR1* rearrangements by FISH. An HCCC with disrupted *EWSR1* (A) and a myoepithelioma with intact *EWSR1* (B).

EWSR1 Genetic Rearrangements in Salivary Gland Tumors A Specific and Very Common Feature of Hyalinizing Clear Cell Carcinoma

Akeesha A. Shah, MD, Robin D. LeGallo, MD,* Annemieke van Zante, MD, PhD,†
Henry F. Frierson, Jr, MD,* Stacey E. Mills, MD,* Kenneth W. Berean, MD,‡
Mark J. Mentrikoski, MD,* and Edward B. Stelow, MD**

Am J Surg Pathol 2013;37:571–578

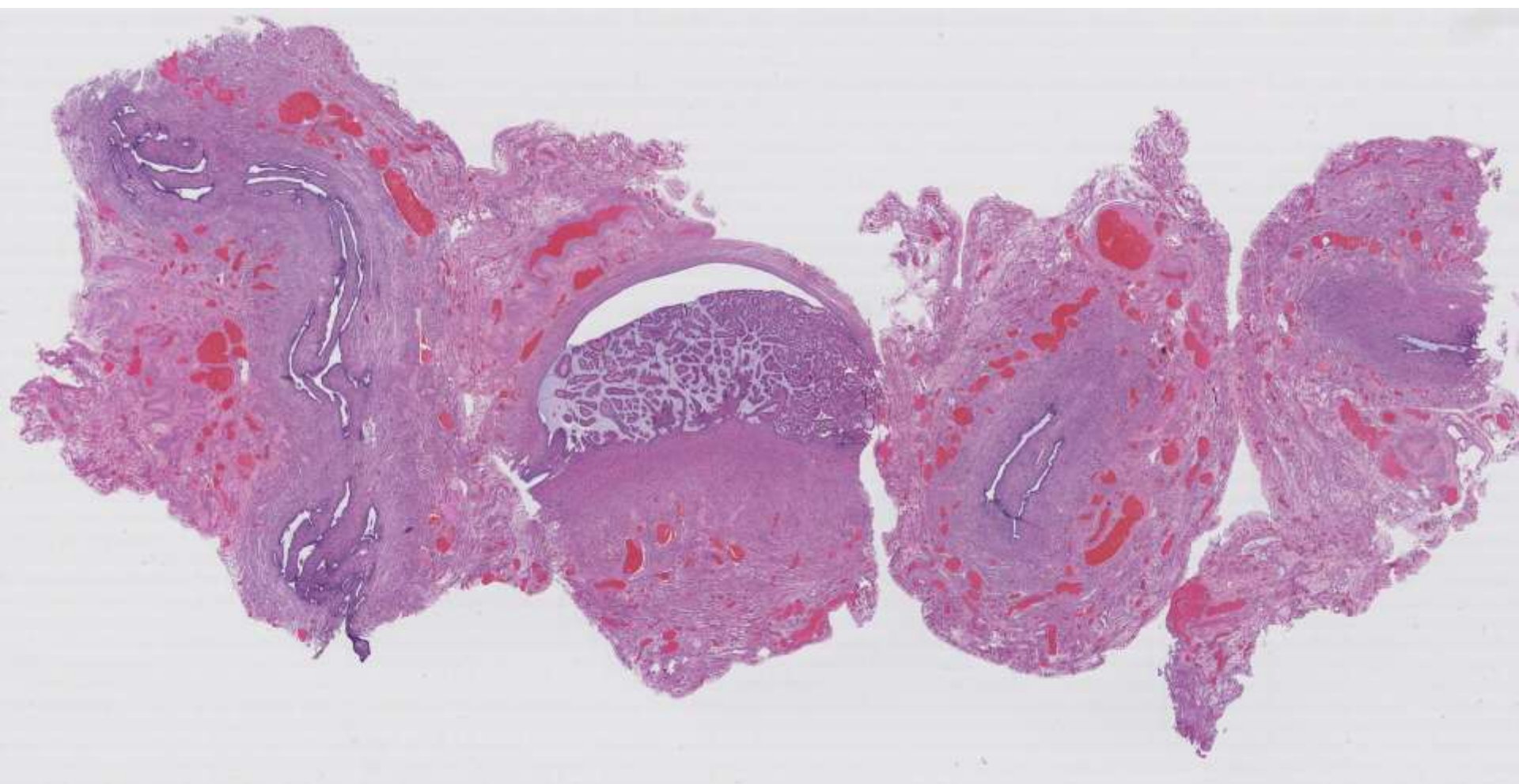
Prognosis

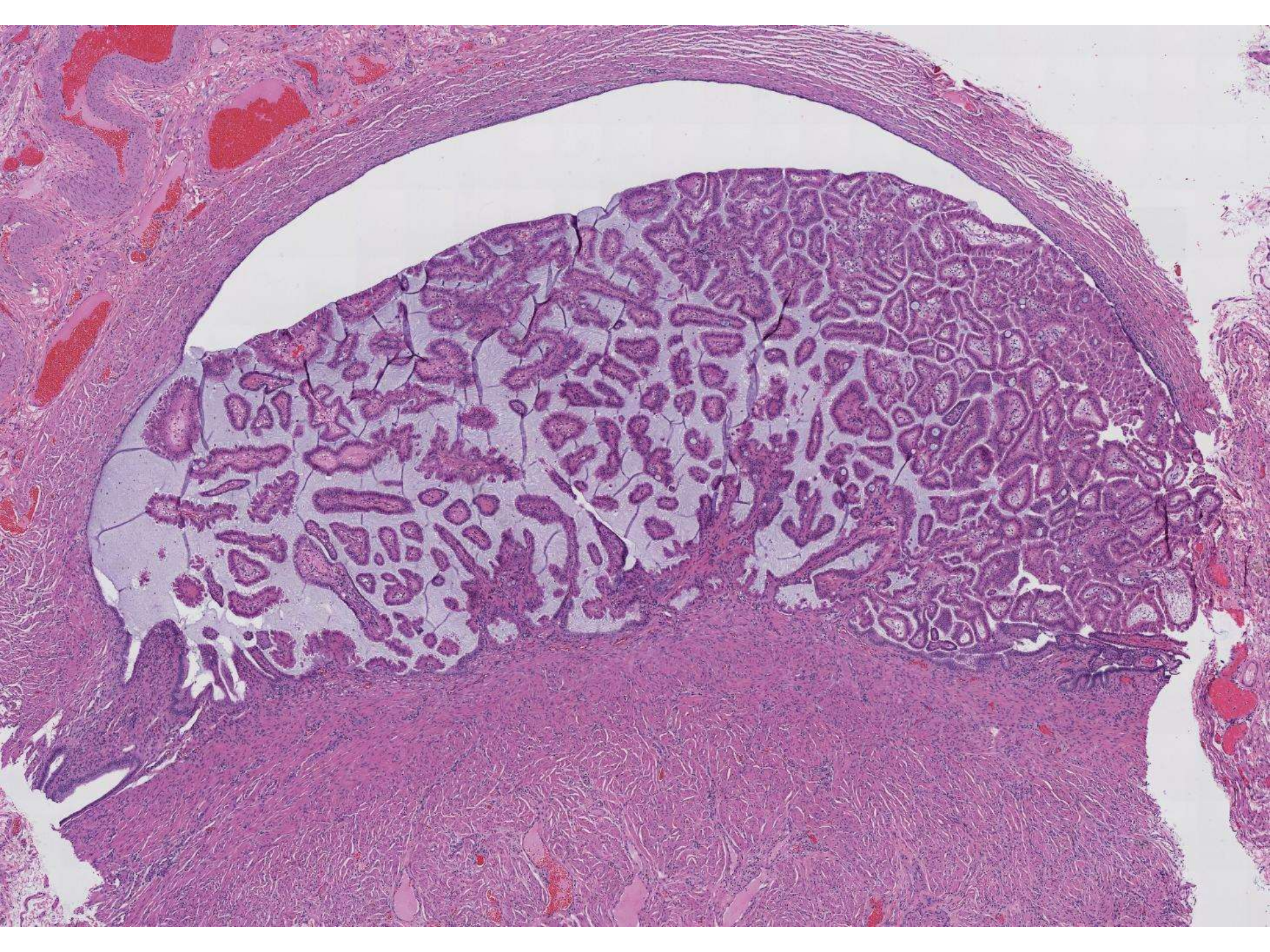
- Generally excellent
 - Local recurrence in 12-17% of reported cases
 - Only infrequent reports of metastatic spread
- Patient underwent wide local excision with negative margins (at 22 weeks)
- Patient and 6-month-old baby girl doing well

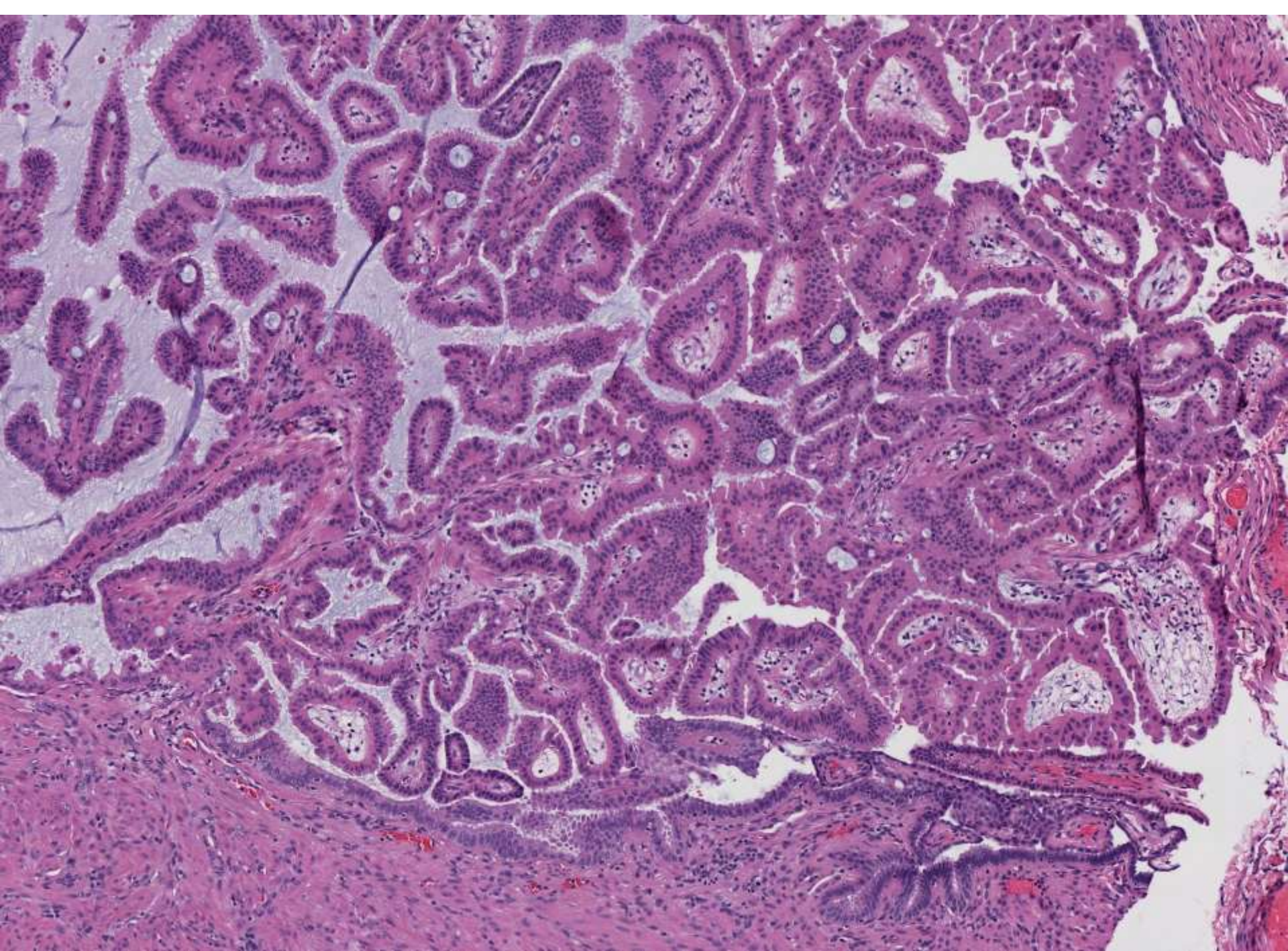
SB 6026

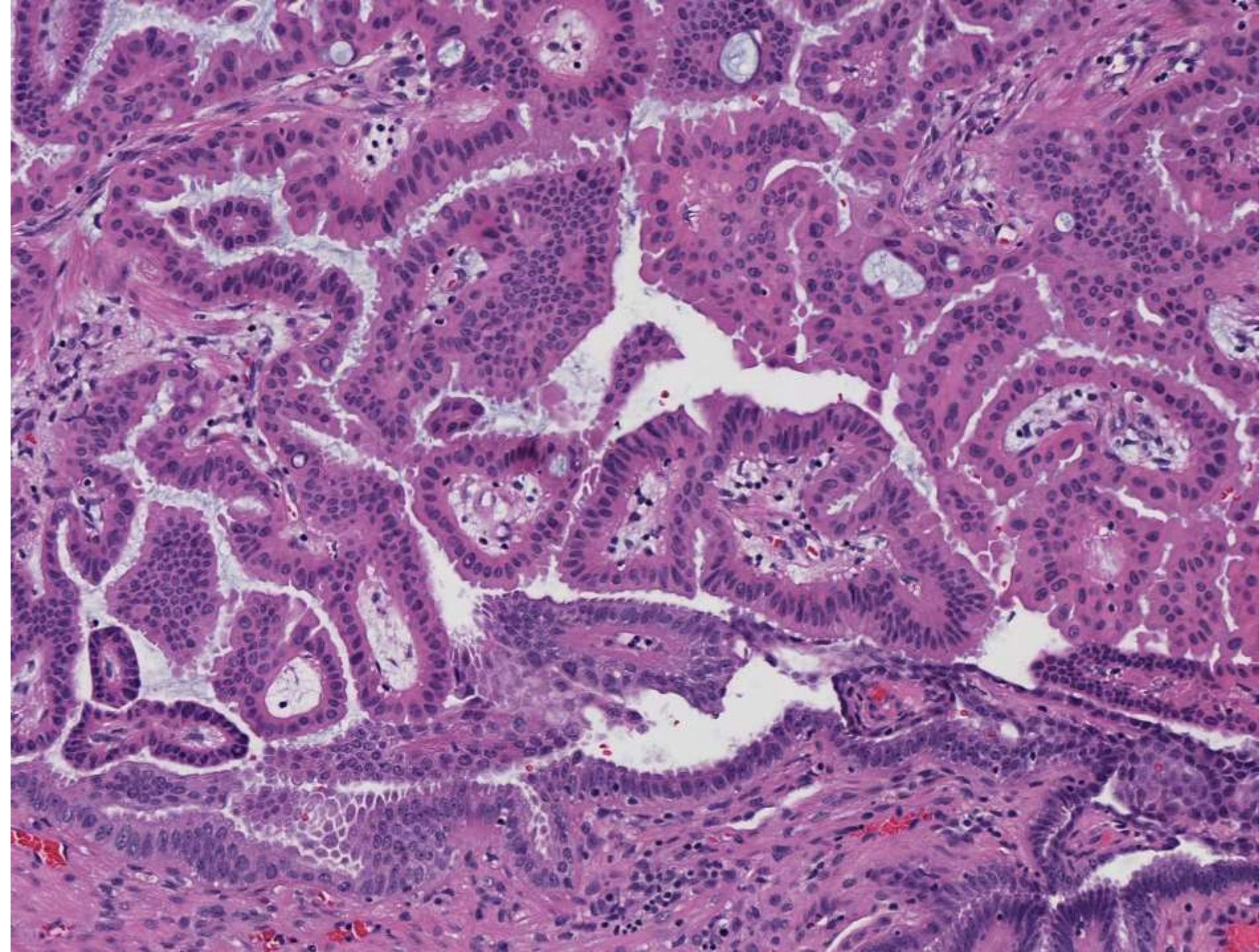
Malti Kshirsagar; El Camino Hospital

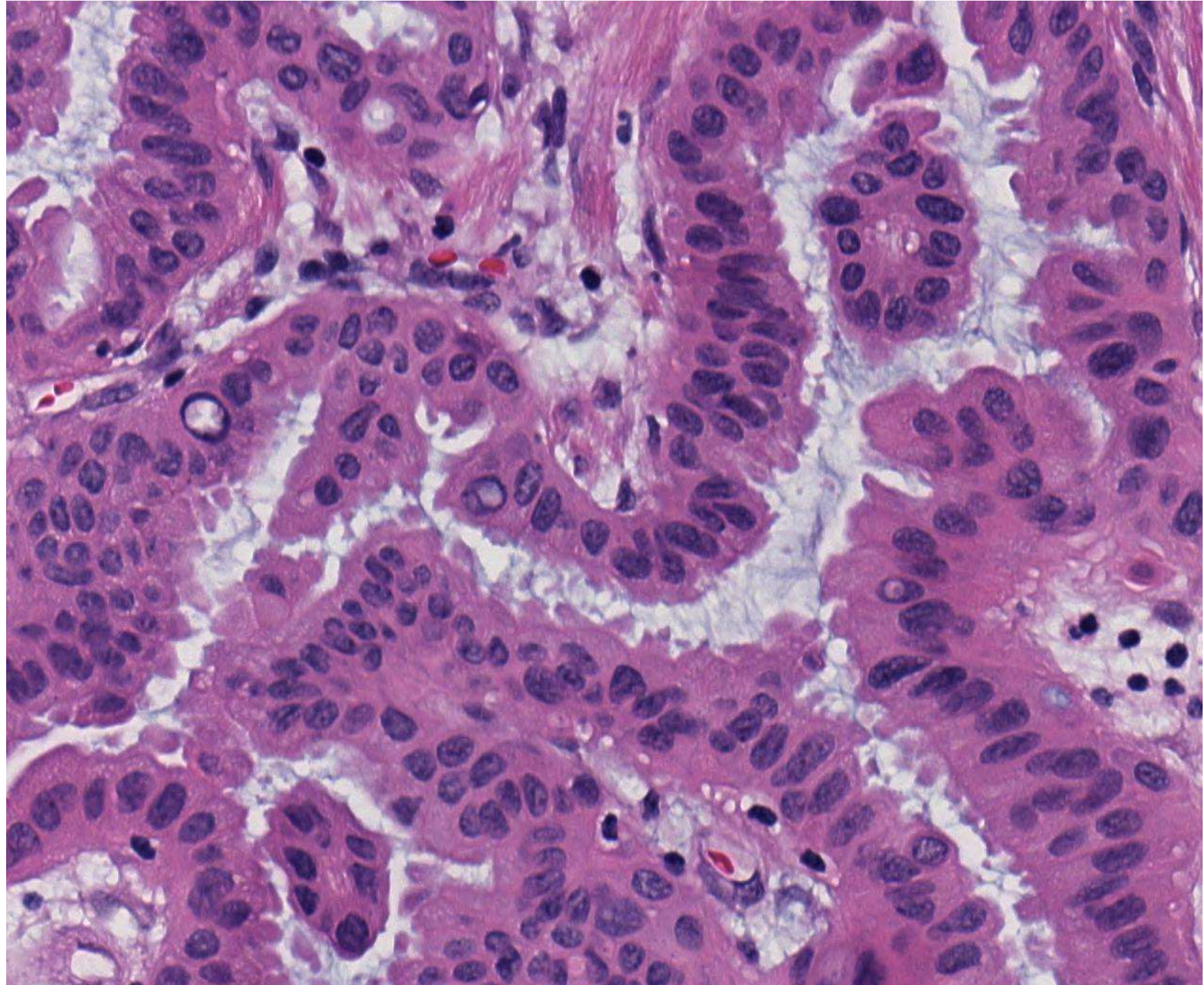
27-year-old female undergoes bilateral
tubal ligation status post cesarean section.

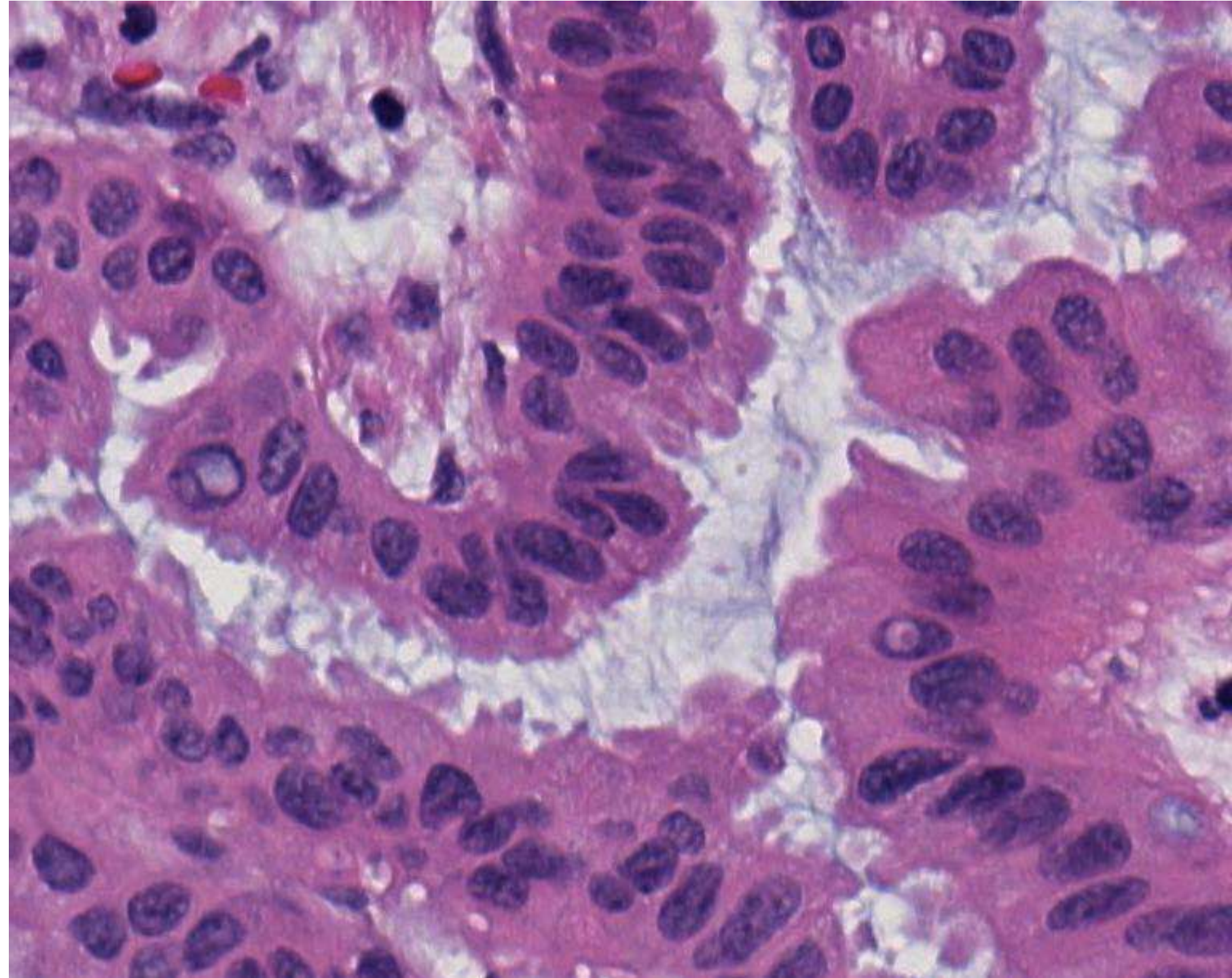










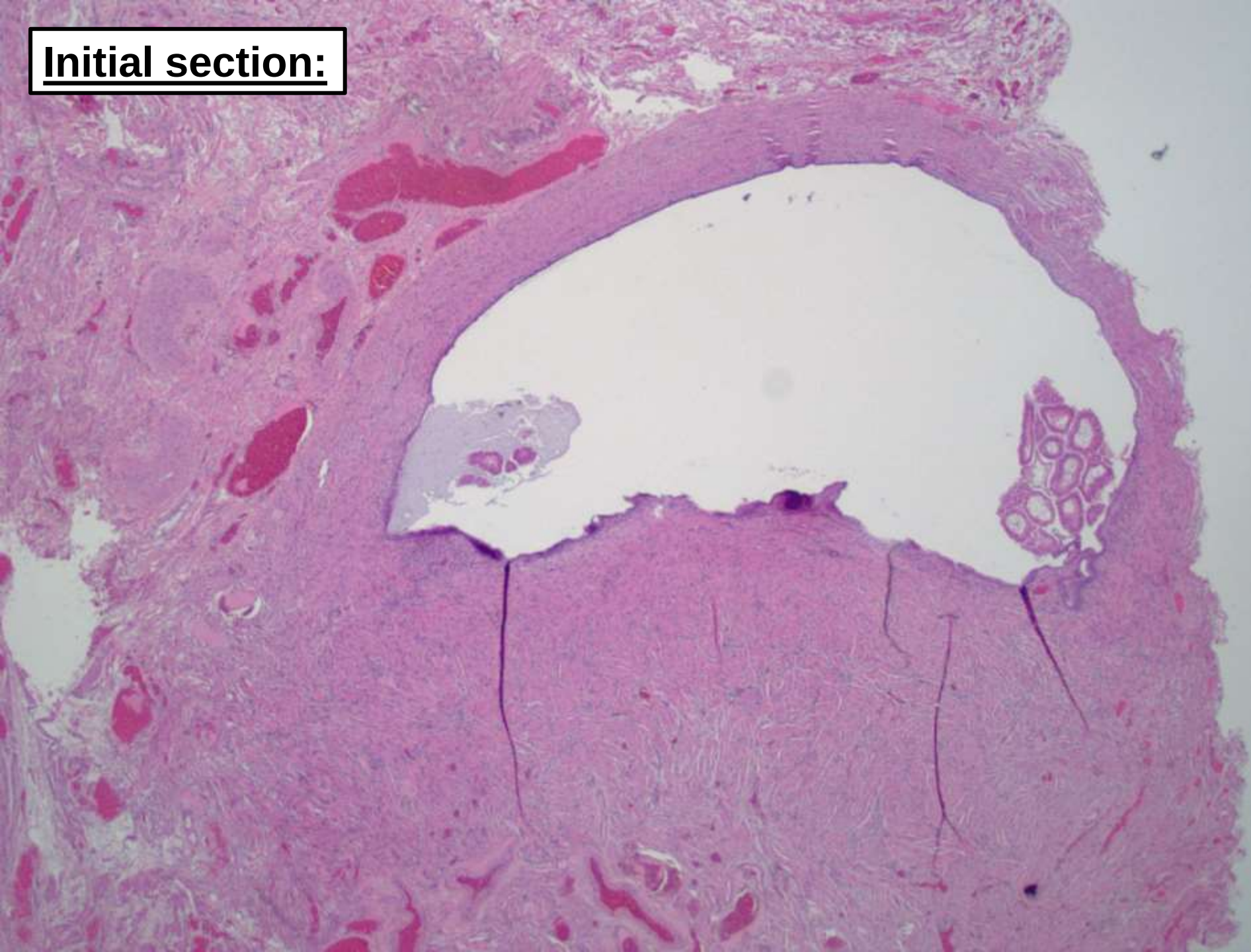


DIAGNOSIS?

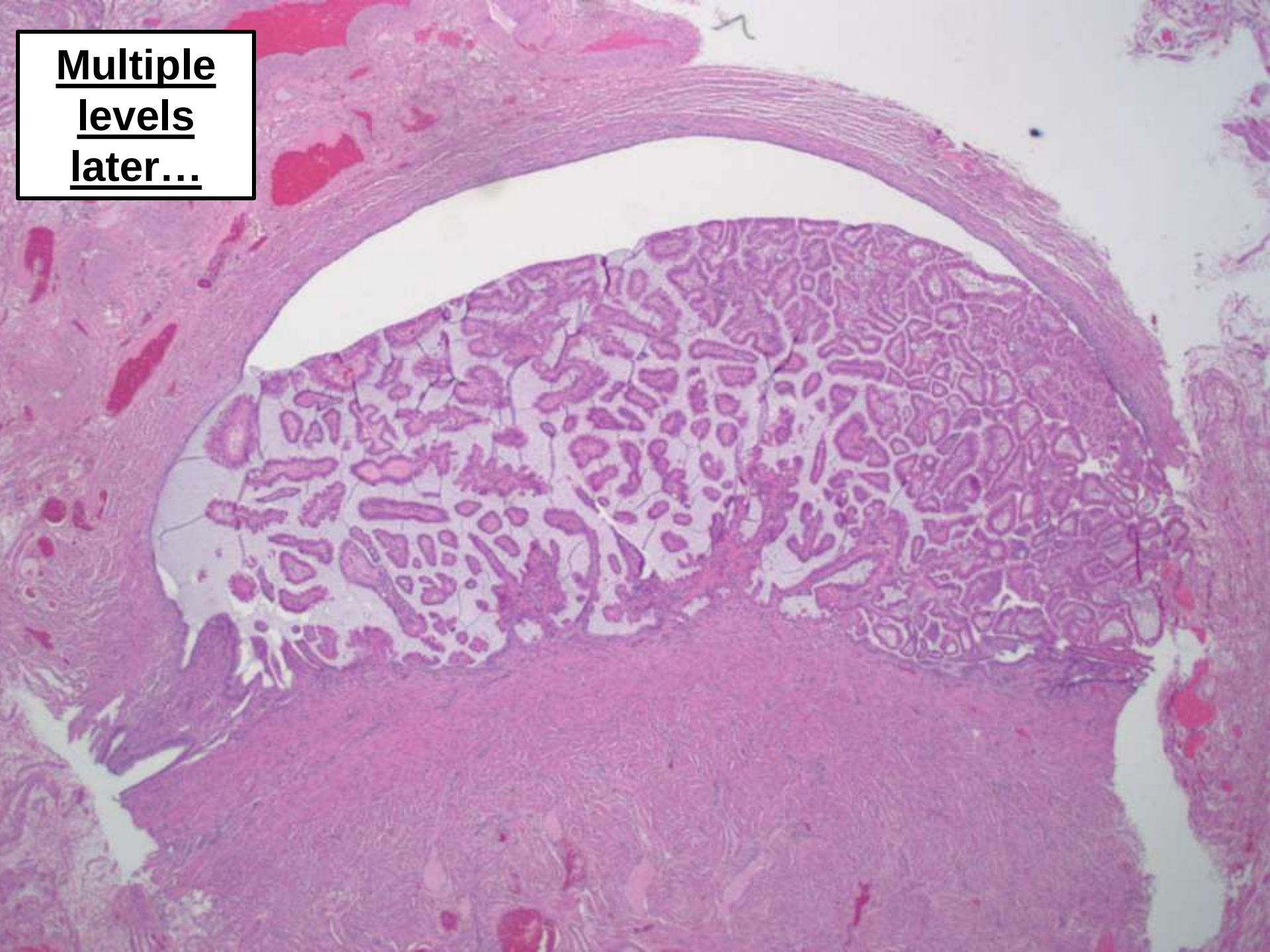


**Diagnosis: Metaplastic Papillary
Tumor of the Fallopian Tube**

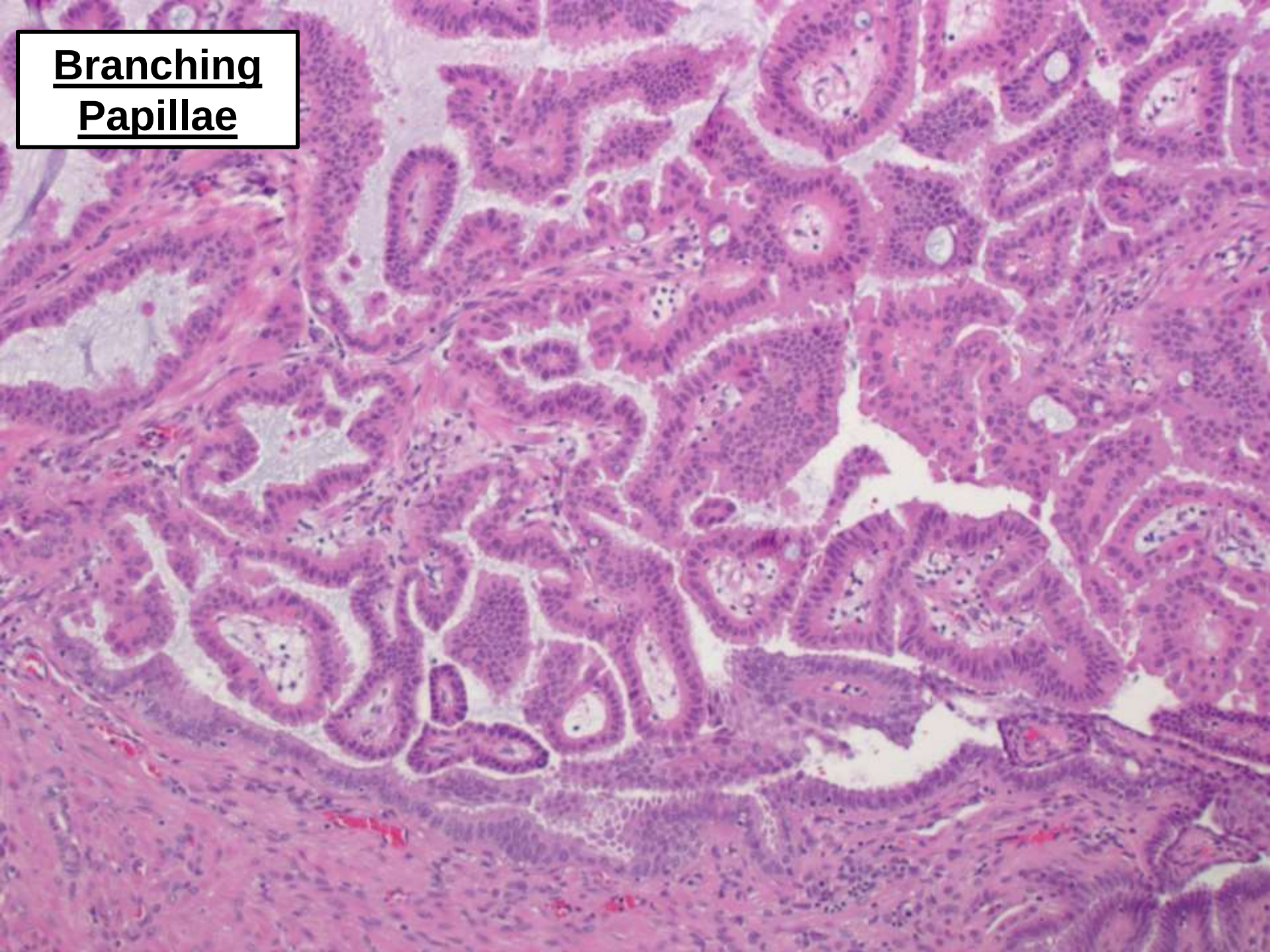
Initial section:



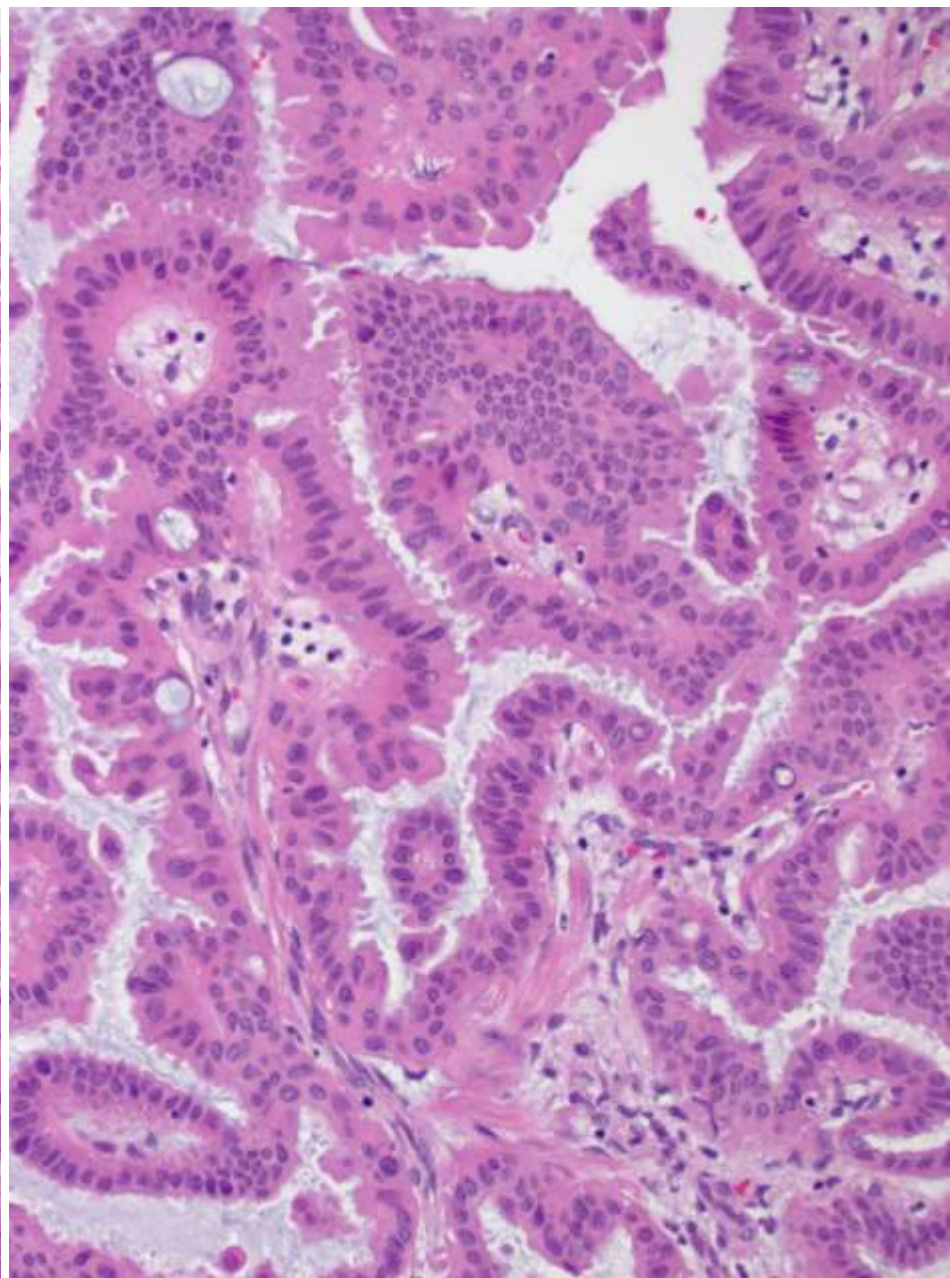
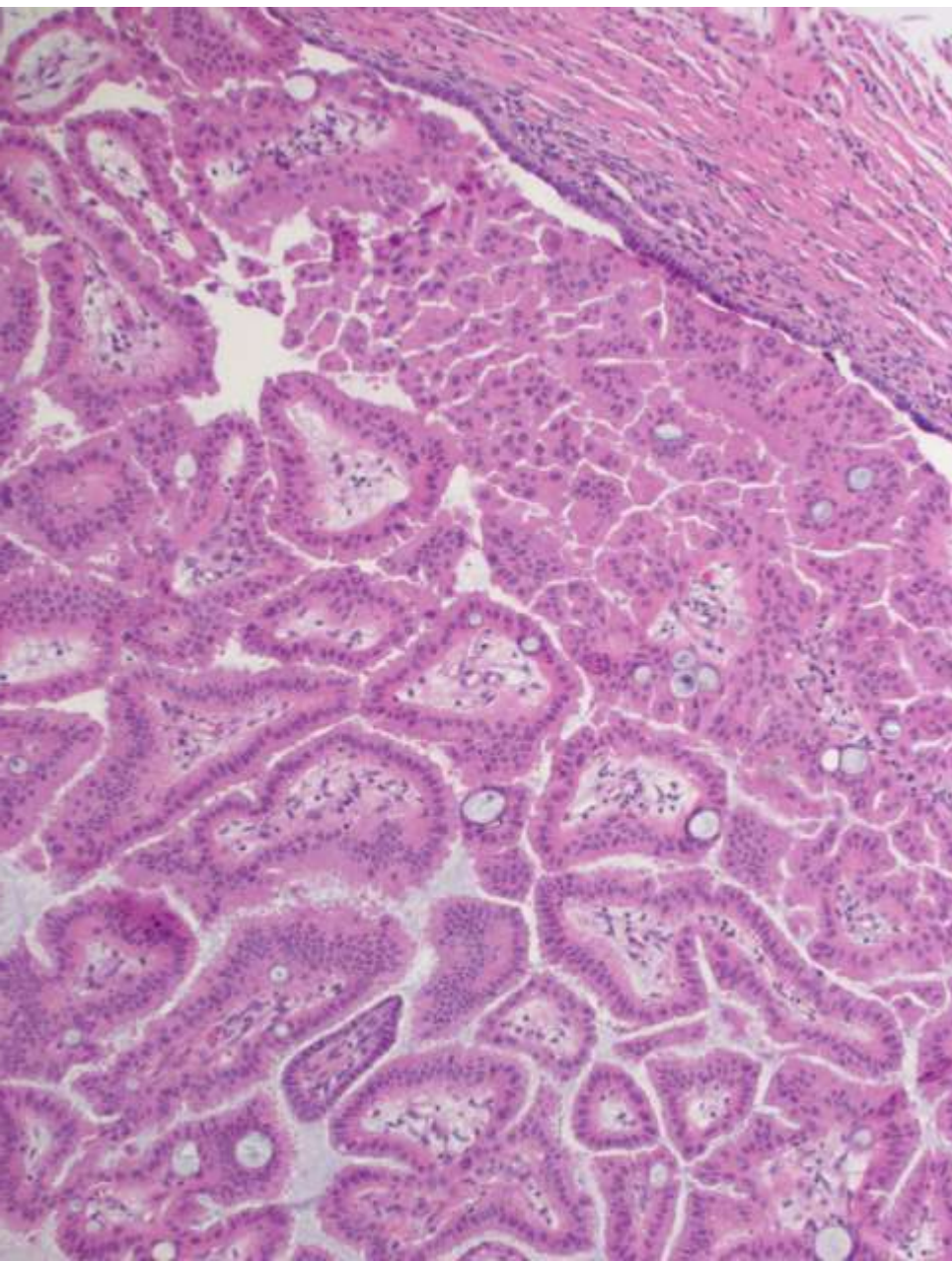
Multiple
levels
later...



Branching
Papillae



Detached epithelial tufts



Saffos, Rhatigan, & Scully, 1980:

“In all cases, the lesions were incidental microscopic findings in grossly normal fallopian tube segments”

“Exophytic papillary configuration”

“Majority of the cells had abundant bright eosinophilic cytoplasm”

“Epithelial budding was common in areas where the cells were pseudostratified”

Metaplastic Papillary Tumor of the Fallopian Tube— A Distinctive Lesion of Pregnancy

ROSILIE O. SAFFOS, RONALD M. RHATIGAN and ROBERT E. SCULLY

Departments of Pathology, University Hospital of Jacksonville, Jacksonville, and University of Florida, Gainesville, Florida; and Massachusetts General Hospital, Harvard Medical School, Boston, Massachusetts

Am. J. Clin. Pathol. 74: 232, 1980

During routine examination of fallopian tube segments removed postpartum for sterilization, four examples of a distinctive intraluminal papillary epithelial tumor have been encountered. The clinical and pathological features of the lesion are described in this article.

Four women, 27 to 33 years of age, underwent abdominal tubal ligations after uneventful pregnancies and deliveries. All were multiparous, and none had a history of gynecological or hormonal abnormalities. Only one patient had used oral contraceptives several years before the discovery of the tubal lesion. None of the patients was taking other hormonal medications, and none had a history of exposure to diethylstilbestrol.

Two women subsequently had total abdominal hysterectomies with bilateral salpingo-oophorectomies because of a suspicion that the tubal lesions were malignant. Additional disease was not detected in either of these patients, and they are well 6 and 2 years postoperatively. A third woman, who received no further therapy, is well 1.5 years after the tubal ligation. The fourth patient was seen only in the immediate postpartum period and was subsequently lost to follow up.

In all cases, the lesions were incidental microscopic findings in grossly normal fallopian tube segments. All were strikingly similar. They involved only part of the circumference of the mucosa and had an exophytic, papillary configuration due, in part, to their origin from the tubal plicae (Fig. 1). One lesion, in addition, contained small cysts deep to the papillae. The cores of the papillae were thin, sometimes edematous, and rounded and consisted of loose fibrovascular tissue that contained small numbers of inflammatory cells, mostly lymphocytes.

The neoplastic epithelium was confined to the surfaces of the papillae and the small cysts. It was made up of columnar, nonciliated cells arranged either as a single layer of uniform cells with basal nuclei or as a pseudostratified layer of cells with nuclei at various levels within them. Epithelial budding was common in areas where the cells were pseudostratified (Fig. 2). Generally, the nuclei were large and oval and ranged from pale and vesicular to moderately hyperchromatic, occasionally they contained prominent nucleoli (Fig. 3). A single mitotic figure was seen in one lesion; none was observed in the other three. The majority of the cells had abundant bright eosinophilic cytoplasm. Sparsely interspersed among

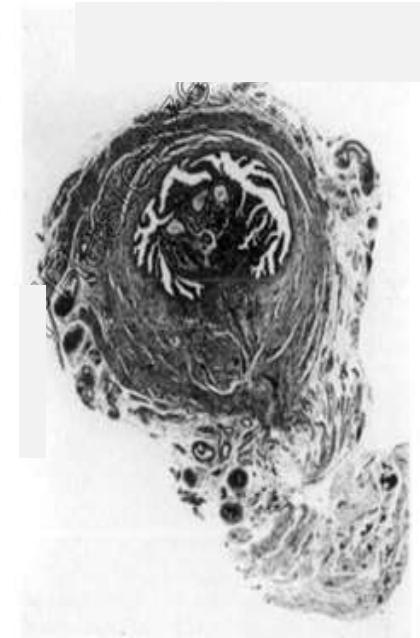


Fig. 1. Metaplastic papillary tumor. The papillary and adenomatous configuration is apparent. The remainder of the circumference of the tubal mucosa appears normal. Hematoxylin and eosin. $\times 15$.

these cells in all cases were other cells with intracytoplasmic mucin that could be demonstrated with the mucicarmine stain. Occasionally the mucin-containing cells were distended and resembled goblet cells (Fig. 3). In two lesions a moderate amount of dense extracellular mucin was also present on the free surfaces of the papillae, within the tubal lumina, and in the lumina of the small cysts.

The remainder of the tubal segments were normal; there was no associated inflammation.

When the authors first encountered the lesion just

1st attempt at molecular analysis (2011):

- 1 case of MPT, 4 ovarian BOT, 2 ovarian LGSC

**MPT showed
NO alterations
in the 8
investigated
chromosomal
regions... a
finding similar
to that seen in
the BOTs**

International Journal of Gynecological Pathology
30:532-535, Lippincott Williams & Wilkins, Baltimore
© 2011 International Society of Gynecological Pathologists

Case Report

Metaplastic Papillary Tumor of the Salpinx: Report of a Case Using Microsatellite Analysis

Tiziana D'Adda, B.Sc.D., Silvia Pizzi, Ph.D., Lorena Bottarelli, Ph.D., Cinzia Azzoni, Ph.D.,
Stefania Manni, and Giovanna Giordano, M.D., Ph.D.

Summary: Metaplastic papillary tumor (MPT) of the salpinx is a rare lesion found in the lumen of fallopian tubes during the postpartum period. This lesion is very small and is composed microscopically of papillae lined by stratified epithelium. Similar to serous borderline ovarian tumors (BOTs), epithelial elements of MPT show a budding, abundant, dense, and eosinophilic cytoplasm, bland nuclei or with mild atypia. It is not clear whether this lesion is a papillary metaplastic proliferation or a small atypical proliferative (borderline) serous tumor associated with pregnancy. Owing to its rarity, MPT has never been investigated in molecular studies and compared with ovarian serous neoplasms. In this study, a case of tubal MPT was molecularly examined and compared with 4 BOTs and with 2 low-grade ovarian carcinomas, using microsatellite analysis with 13 markers at 8 chromosomal regions involved in ovarian carcinogenesis. The tubal MPT and one of the BOTs showed no alterations in the investigated chromosomal regions. The remaining 3 BOTs showed only single allelic imbalances. Instead, low-grade serous carcinomas showed a higher frequency of alterations, including allelic imbalance at chr10q23, 1p36, 9p22, and 17. In conclusion, this study provides, for the first time, molecular data on an MPT of the fallopian tube, indicating that this entity might share both morphologic and molecular similarities with a subset of minimally altered BOTs, termed atypical proliferative serous tumors, which behave in a benign manner. However, in our opinion, further molecular studies should be conducted on other cases of MPTs to confirm this hypothesis. **Key Words:** Metaplastic papillary tumor—Borderline ovarian tumor—Ovarian low-grade carcinoma—Microsatellite analysis.

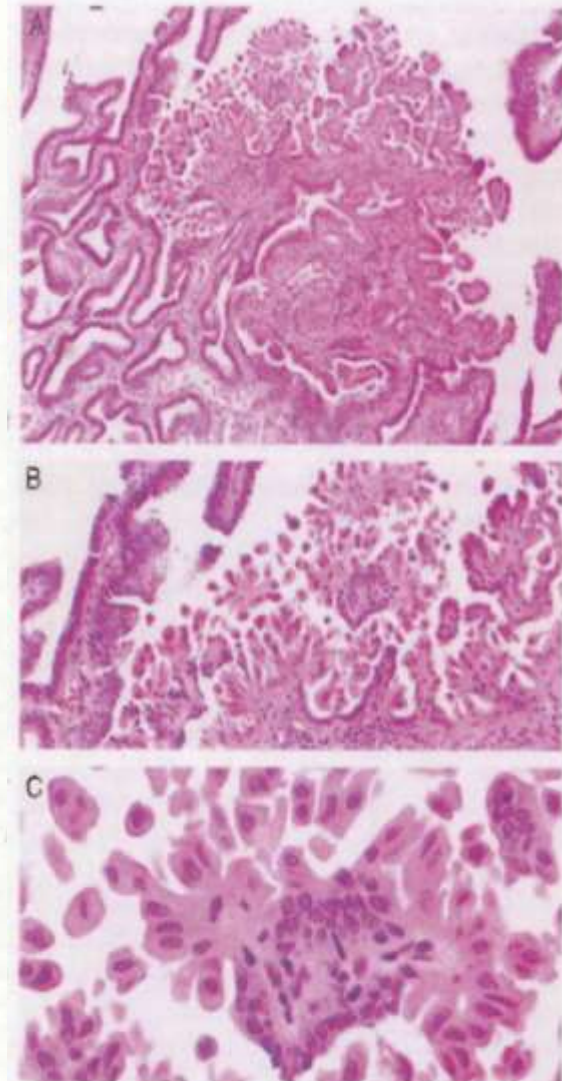
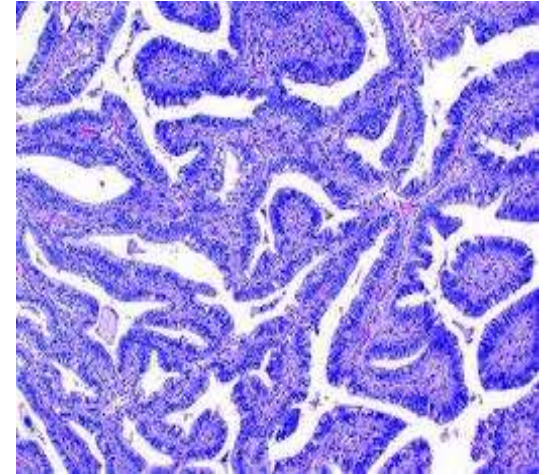


FIG. 1. Tubal metaplastic papillary tumor with small papillary formations (A, H&E: 100 ×). Budding of epithelial elements (B, H&E: 200 ×) showing abundant eosinophilic cytoplasm and slight nuclear pleomorphism (C, H&E: 630 ×). H&E indicates hematoxylin and eosin.

Differential diagnosis:

1. Fallopian tube papilloma

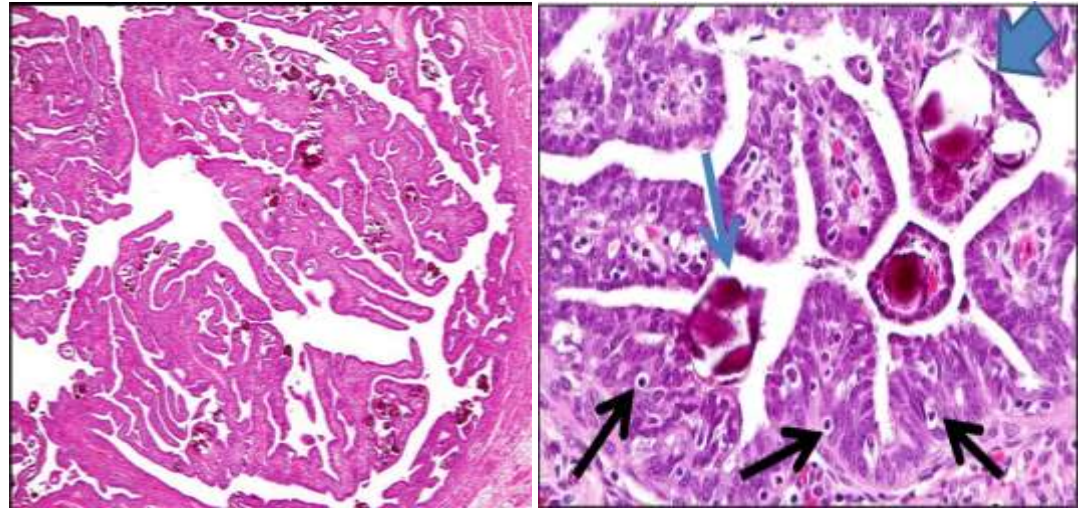
- Intraluminal mass of complex papillae resembling an exaggerated pattern of normal tubal mucosa with fine fibrovascular cores
- Maintains normal tubal cell types



Blaustein's, 6th Ed

2. Papillary tubal hyperplasia

- Small papillae and rounded clusters of epithelial cells with associated psammoma bodies



Kurman RJ, Vang R, et al. Am J Surg Pathol. 2011; 35(11):1605-1614

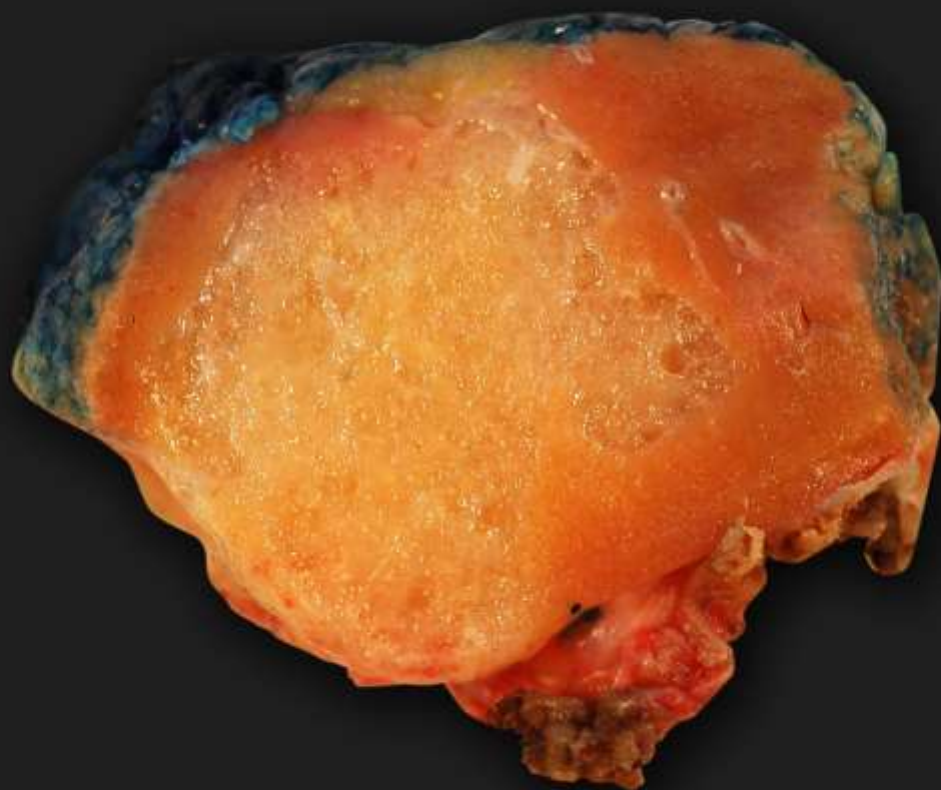
Take home points:

1. This is an uncommon lesion – clinically occurs in patients with a recent pregnancy
2. Some consider this lesion as a “low grade borderline tumor associated with pregnancy;” others suggest it may represent a metaplastic proliferation related to pregnancy
3. Histological features include microscopic size, papillary architecture, pseudostratified cells with eosinophilic cytoplasm lining stromal cores, and occasional detached epithelial tufts
4. All cases reported in the literature have shown indolent behavior, with a lack of recurrence or metastasis

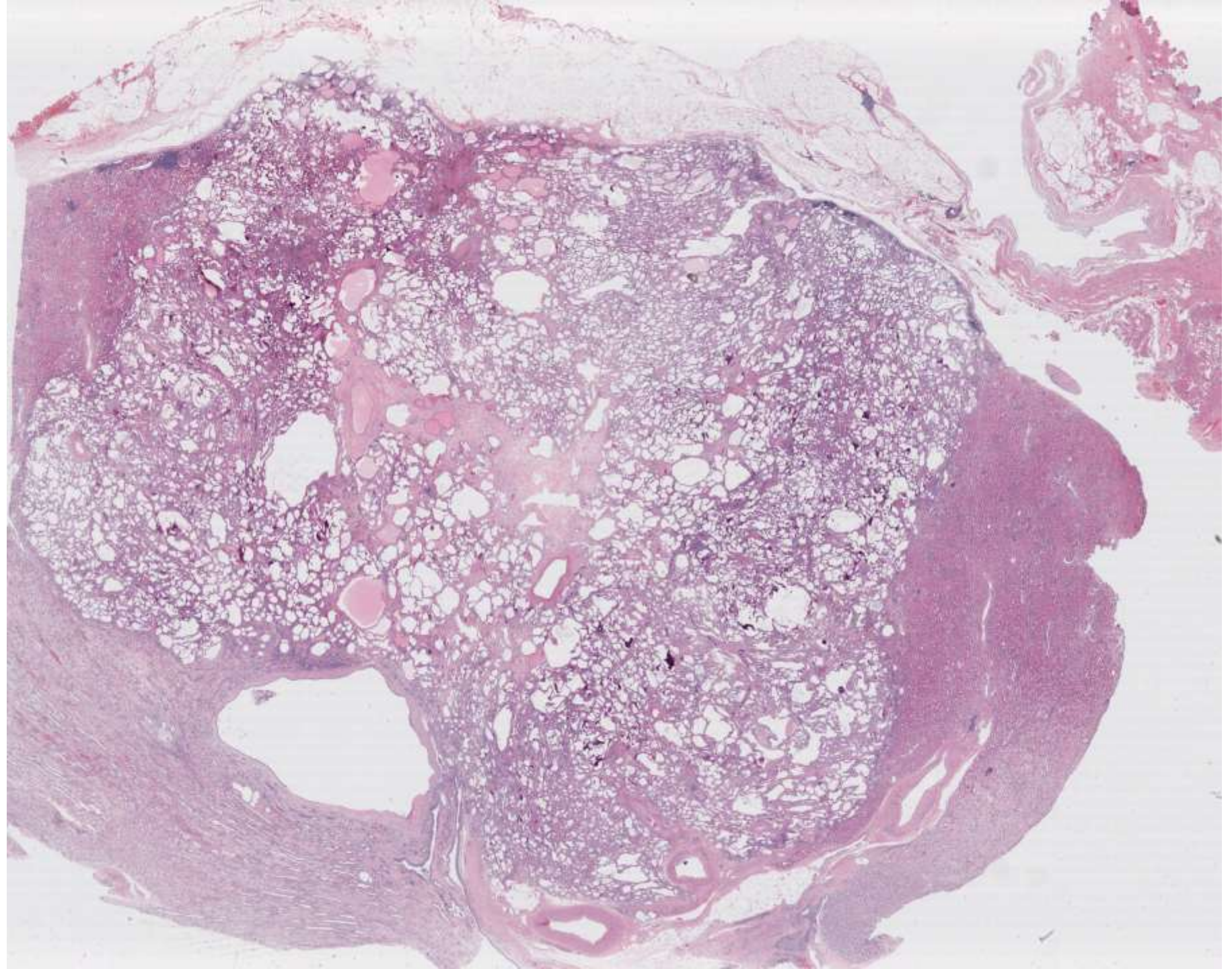
SB 6027

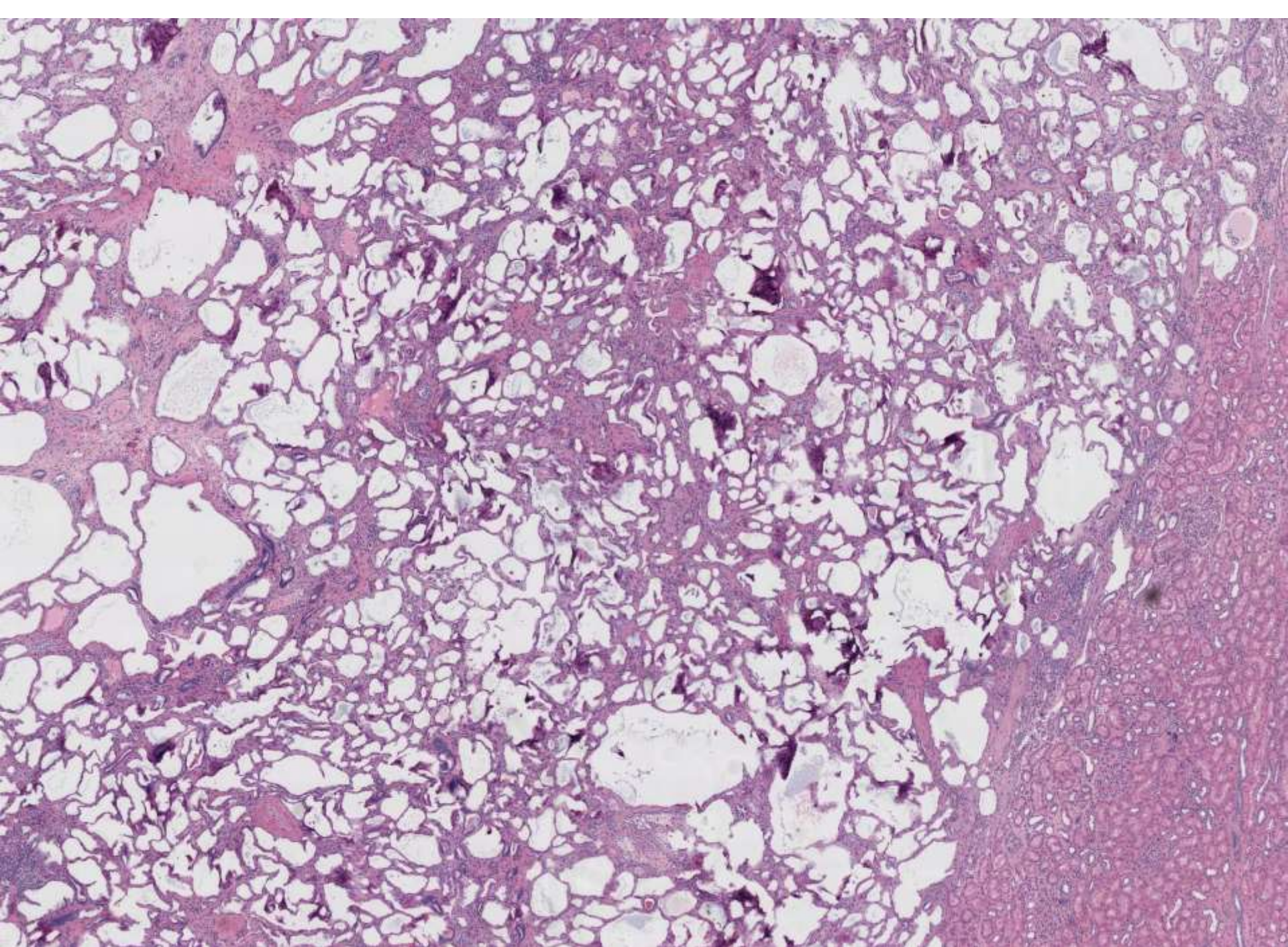
John Higgins; Stanford

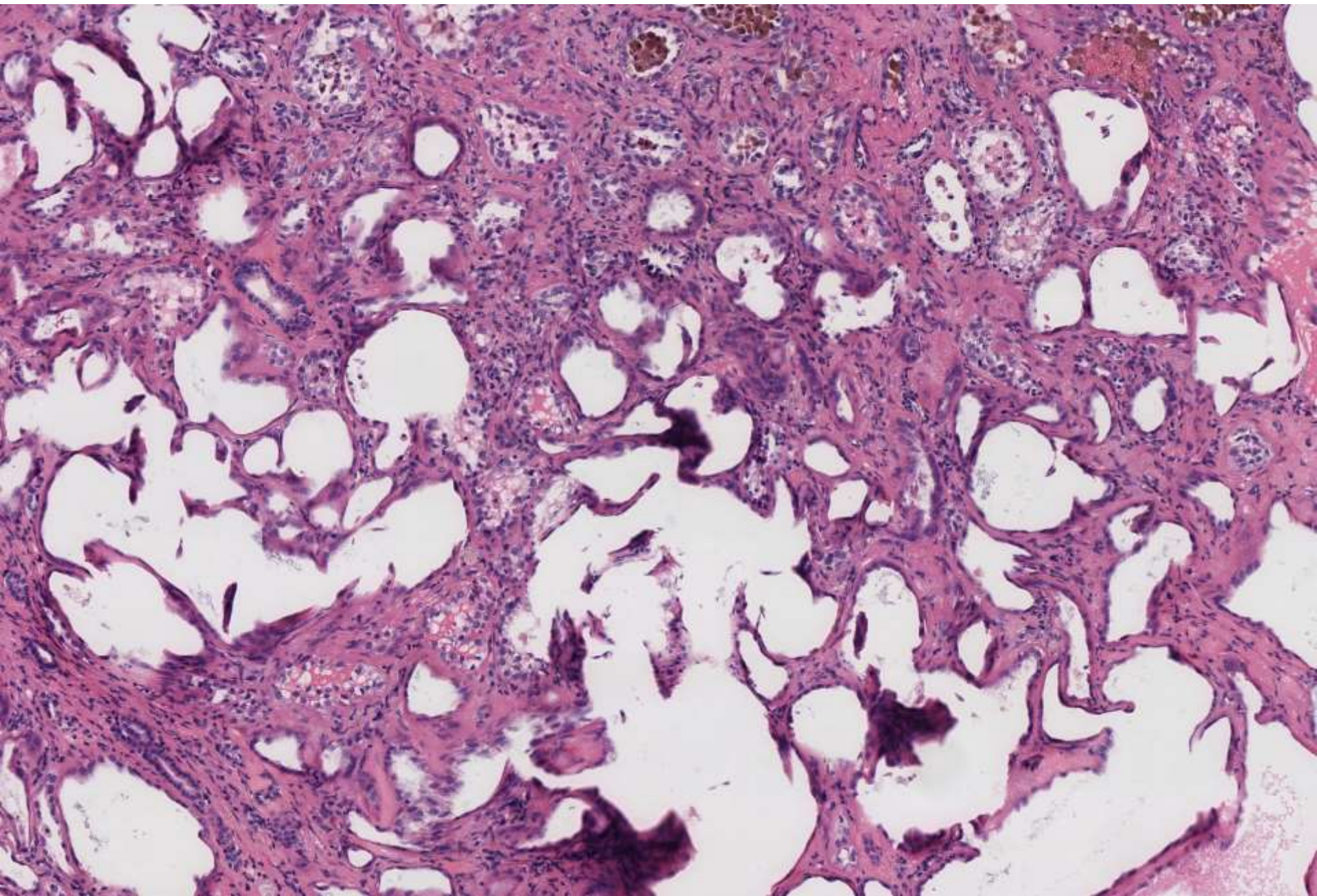
62-year-old male with renal mass.

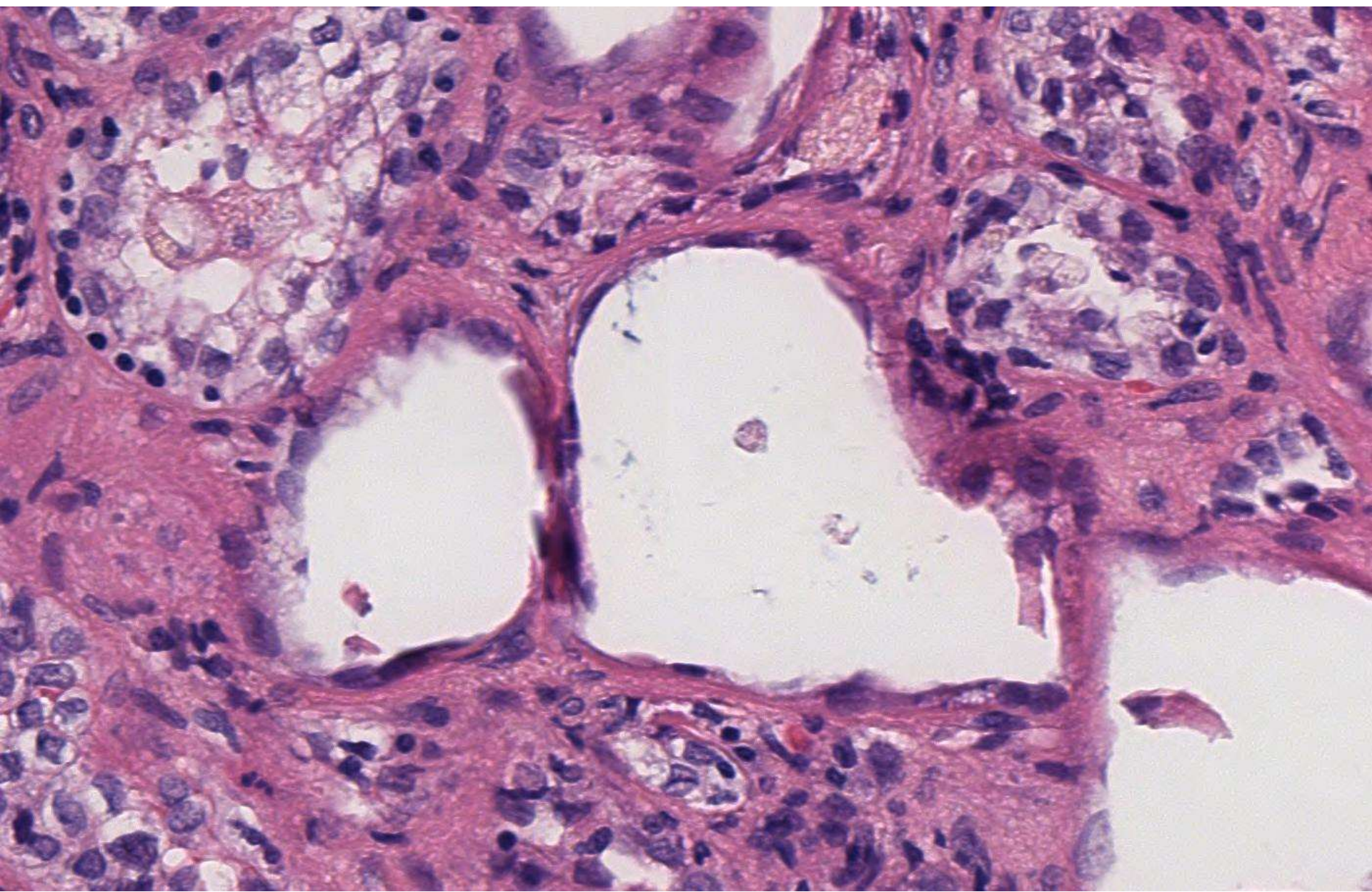


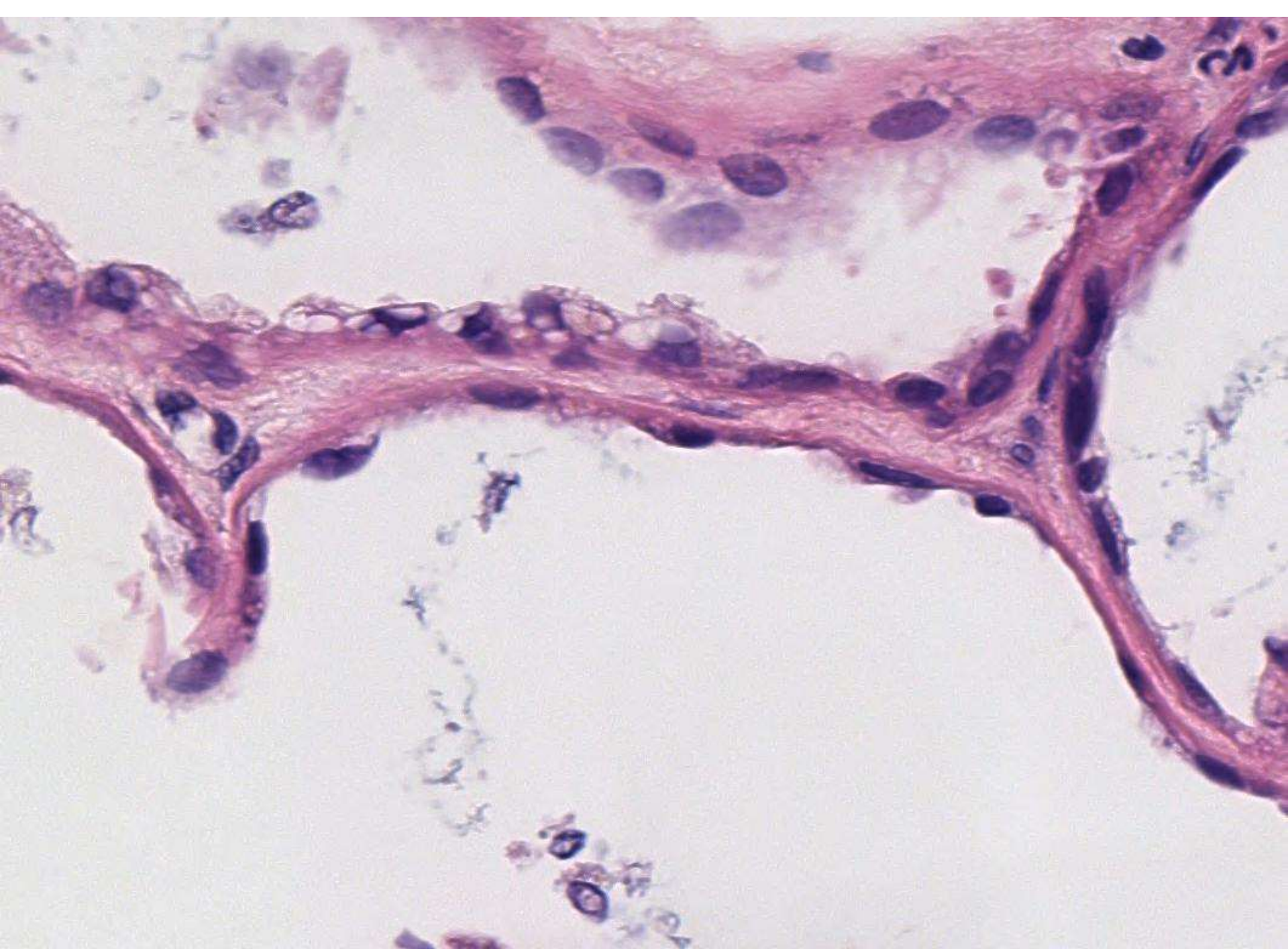
4 cm

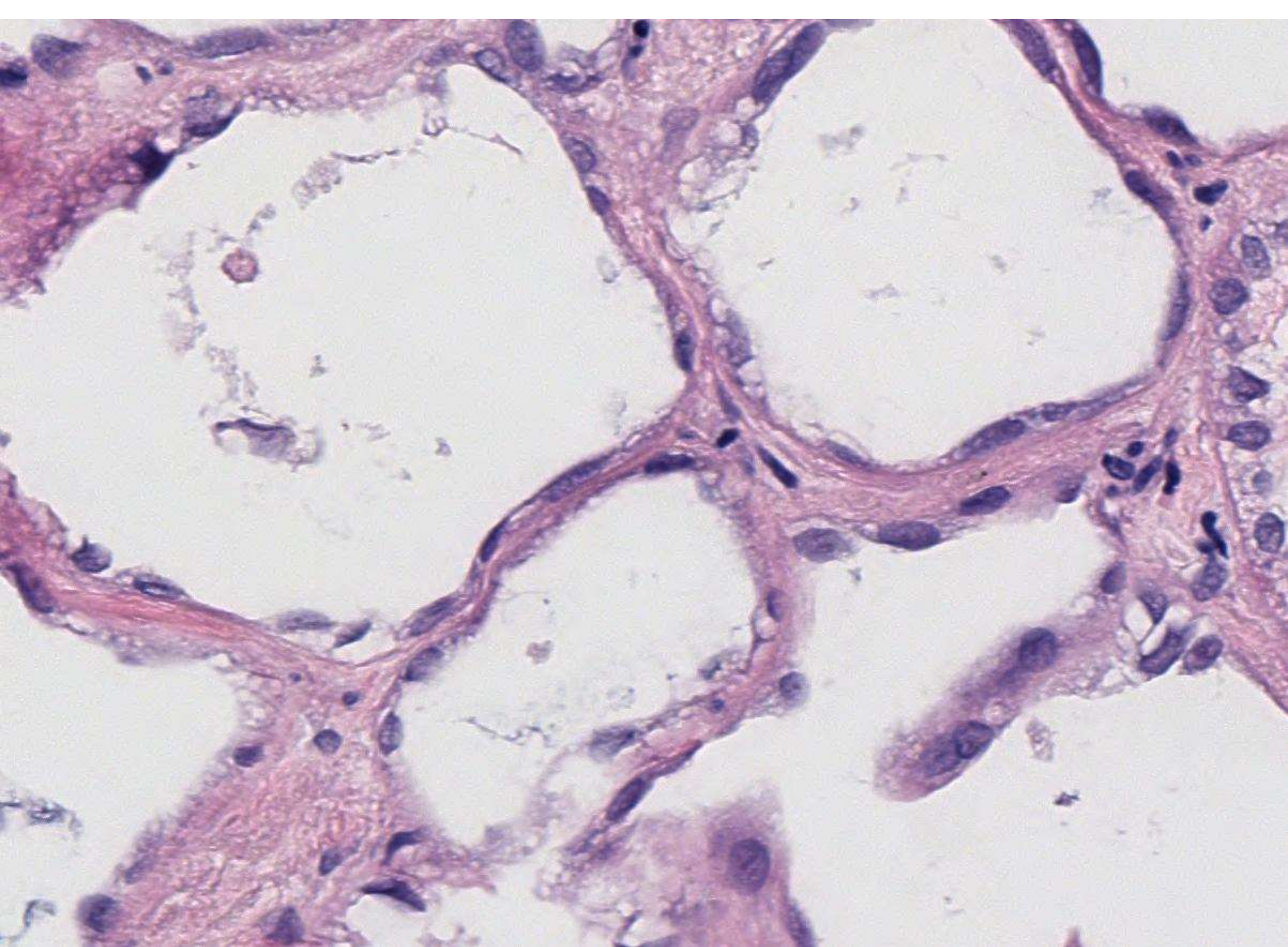










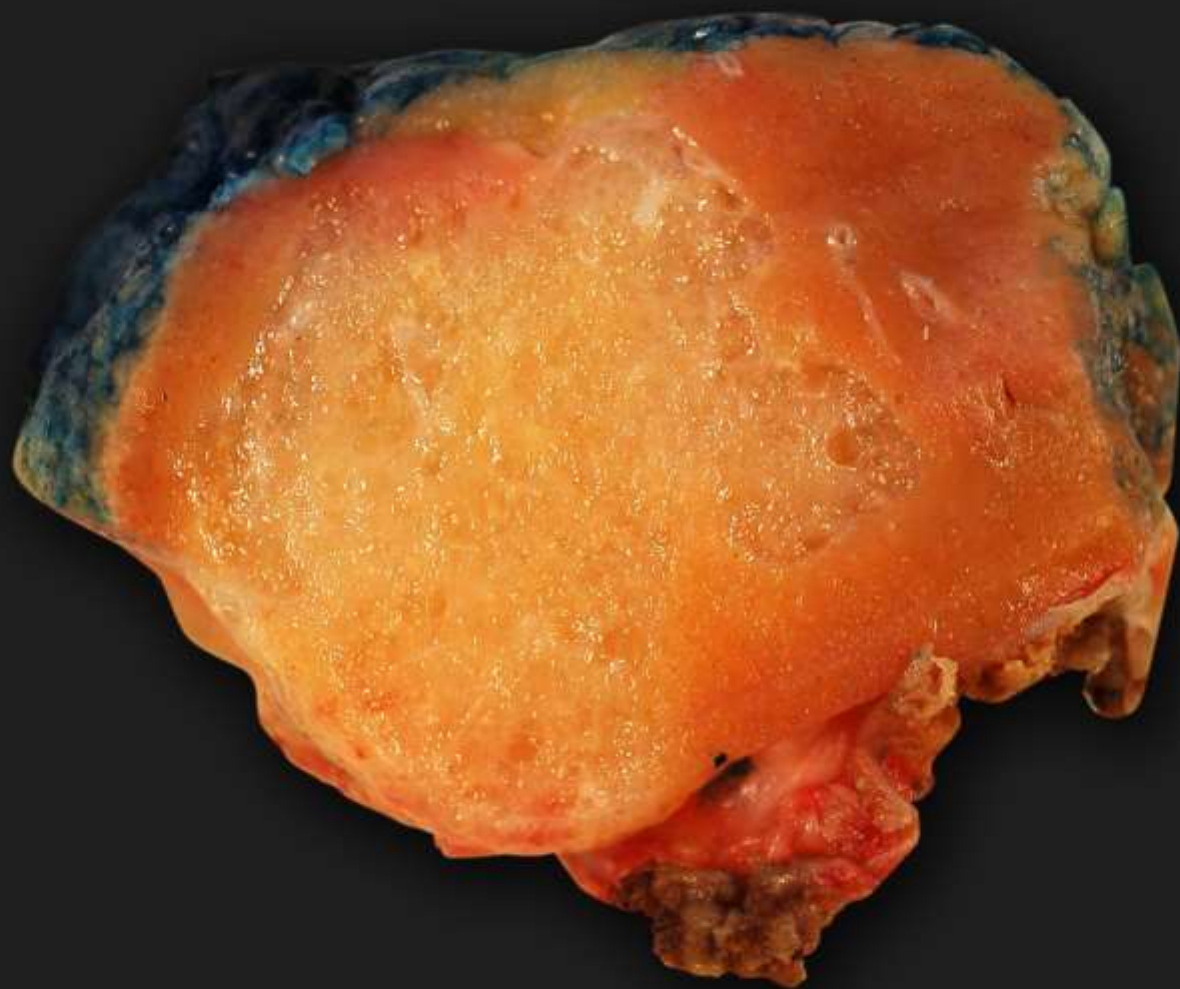


DIAGNOSIS?

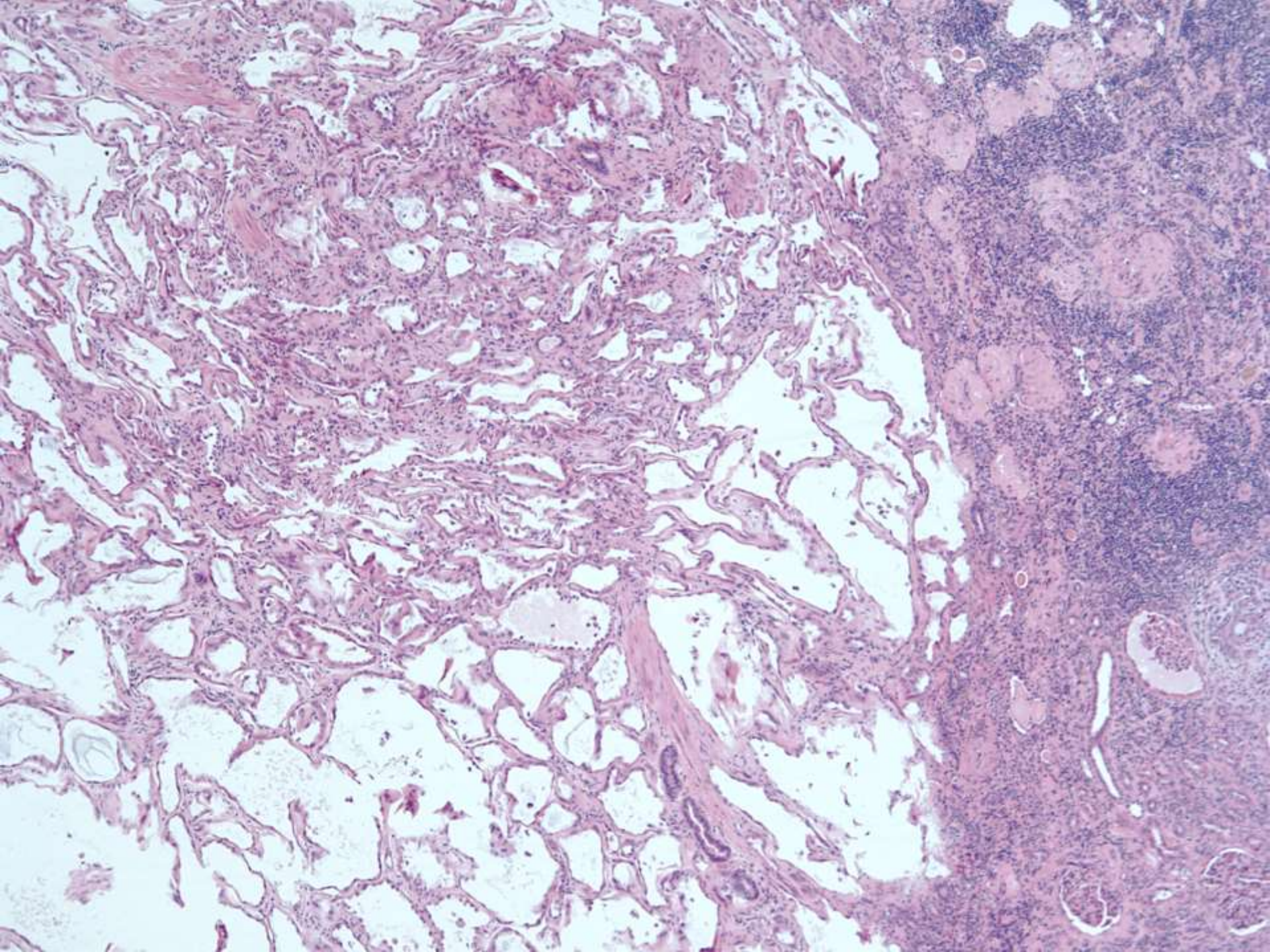


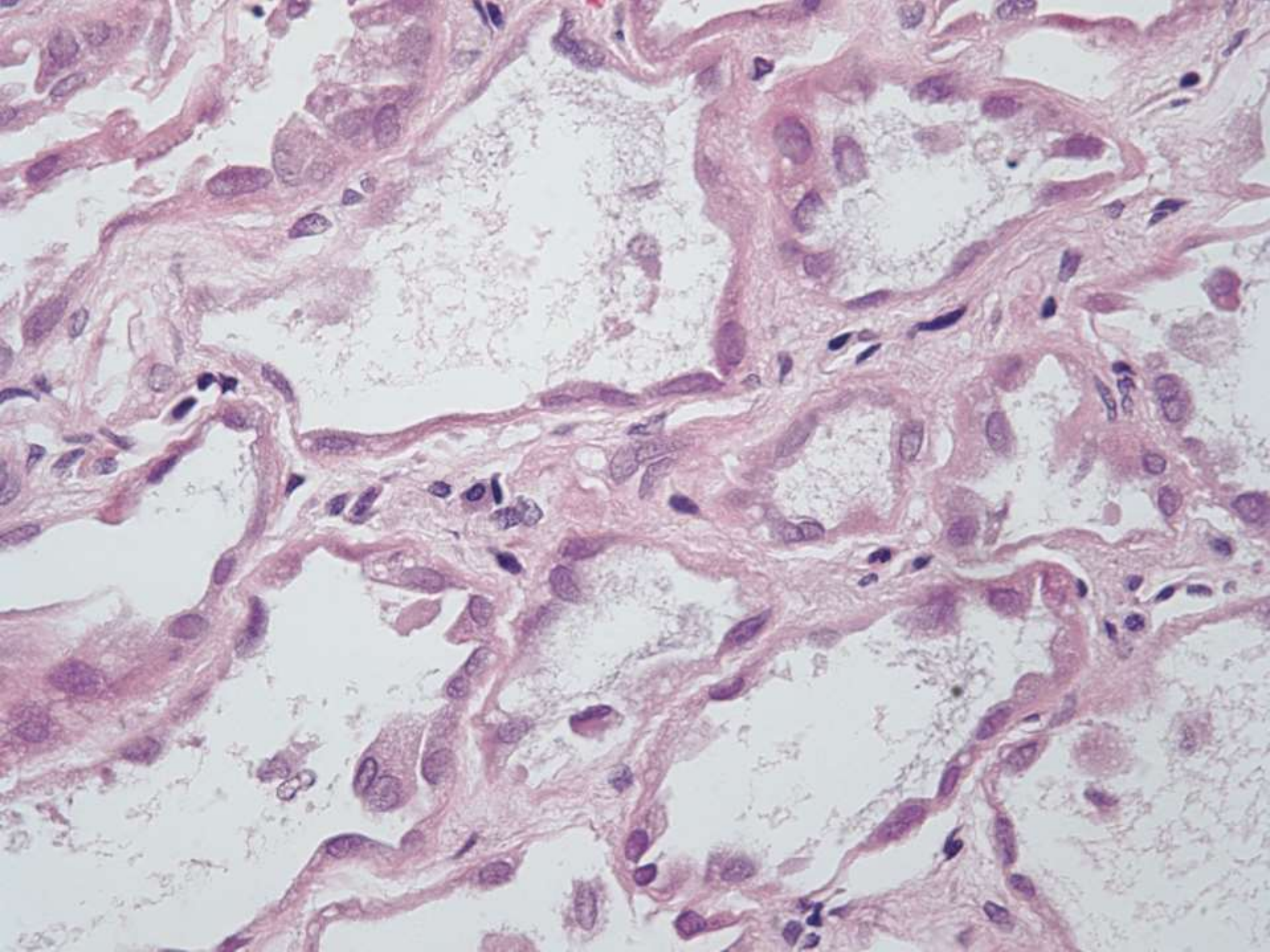
Tubulocystic carcinoma of the kidney: clinicopathologic analysis of 31 cases of a distinctive rare subtype of renal cell carcinoma. Amin MB et al.
Am J Surg Pathol. 2009 Mar;33(3):384-92.

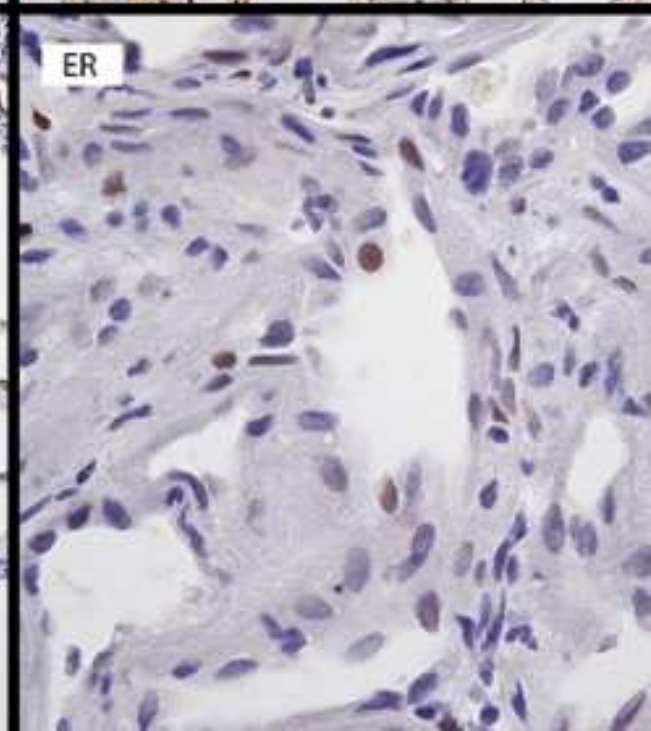
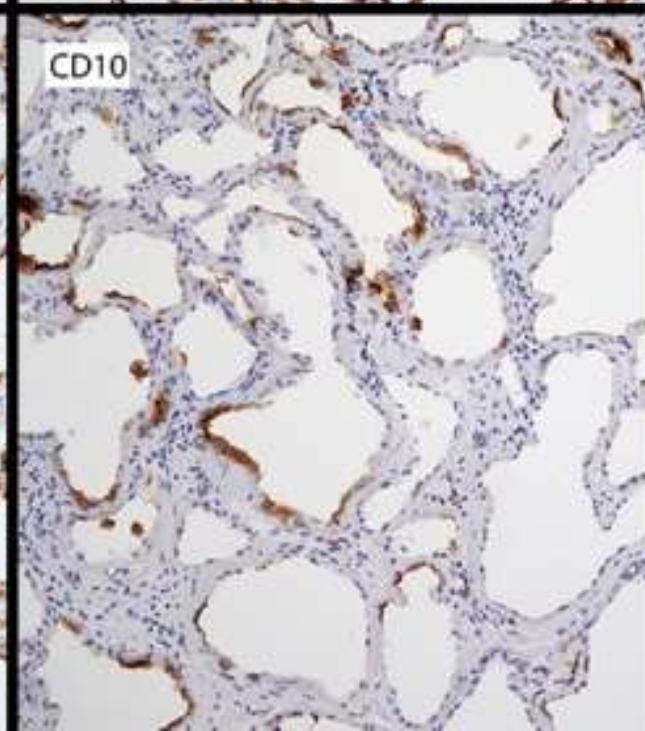
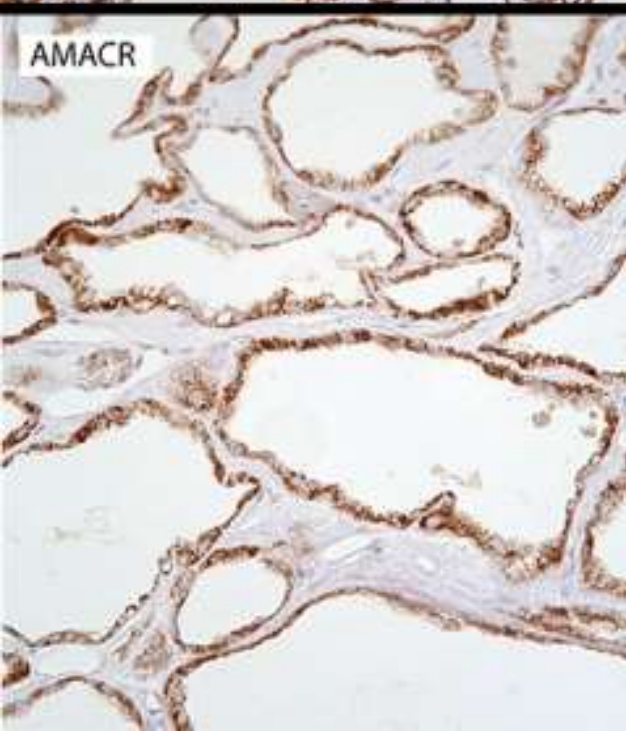
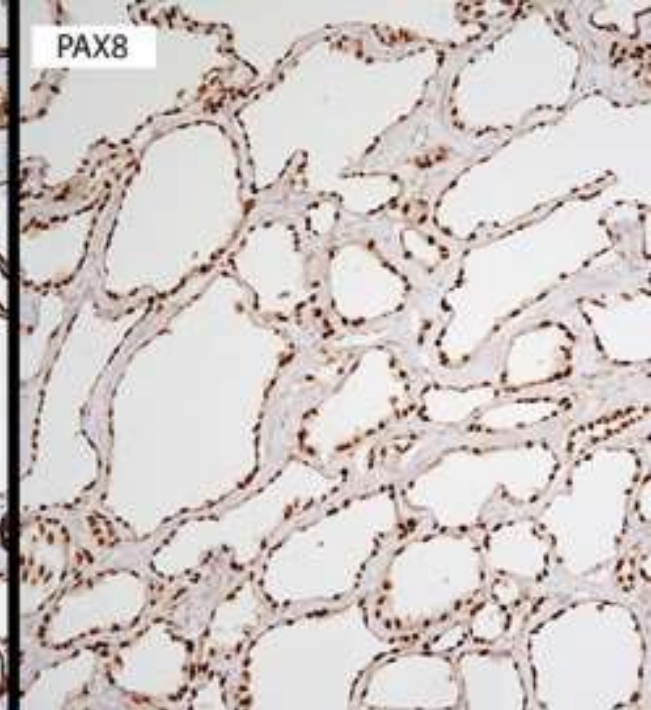
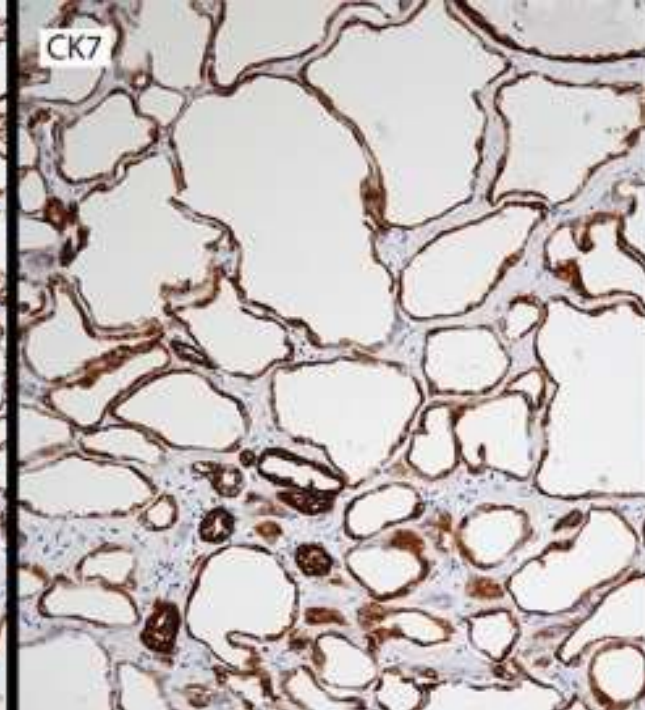
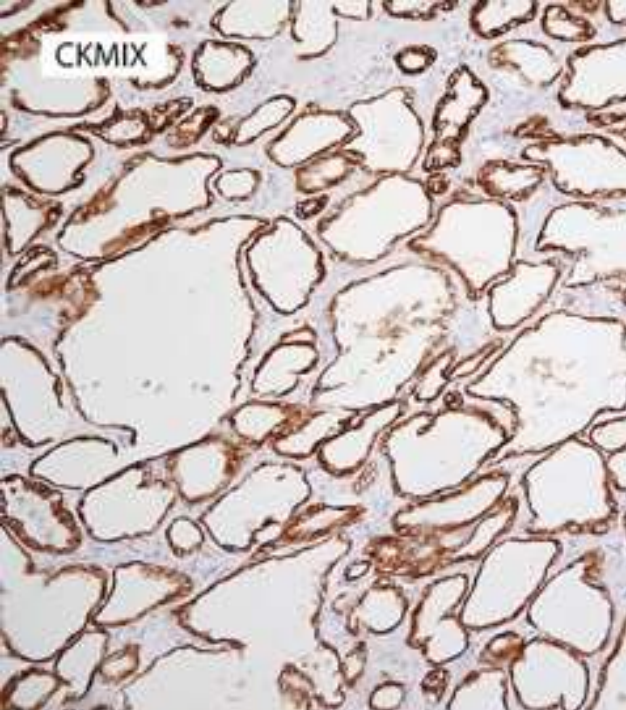
- Bellini duct carcinoma and low-grade collecting duct carcinoma now called tubulocystic RCC
- Strong male predominance (7:1)
- Stage break down
 - pT1 (24)
 - pT2 (4)
 - pT3 (3)
- Disease progression (median follow-up of 56 months) occurred in 3 patients
 - 1 with local recurrence
 - 2 with distant metastasis to bone and liver



4 cm







Tubulocystic carcinoma

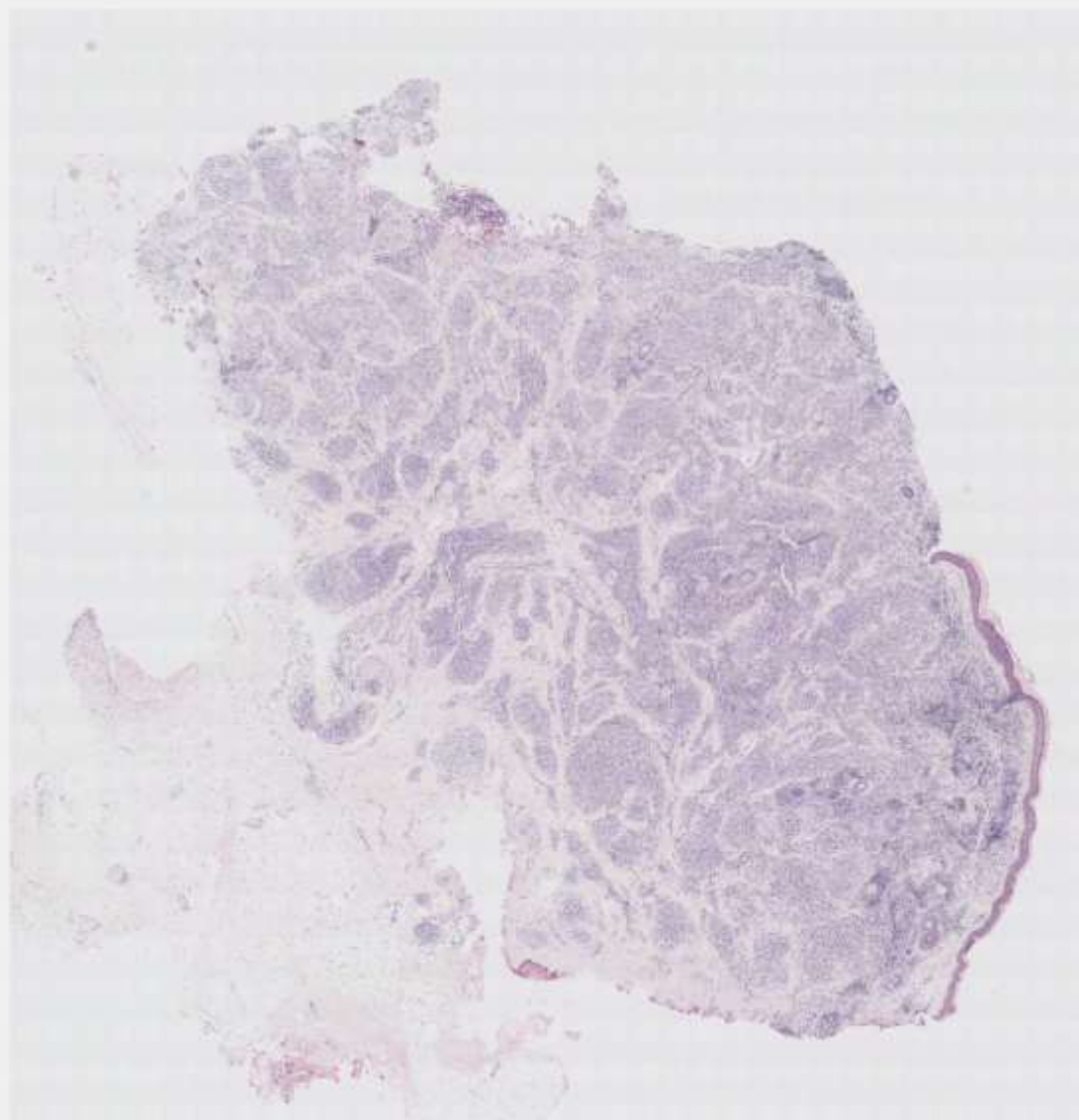
DDX

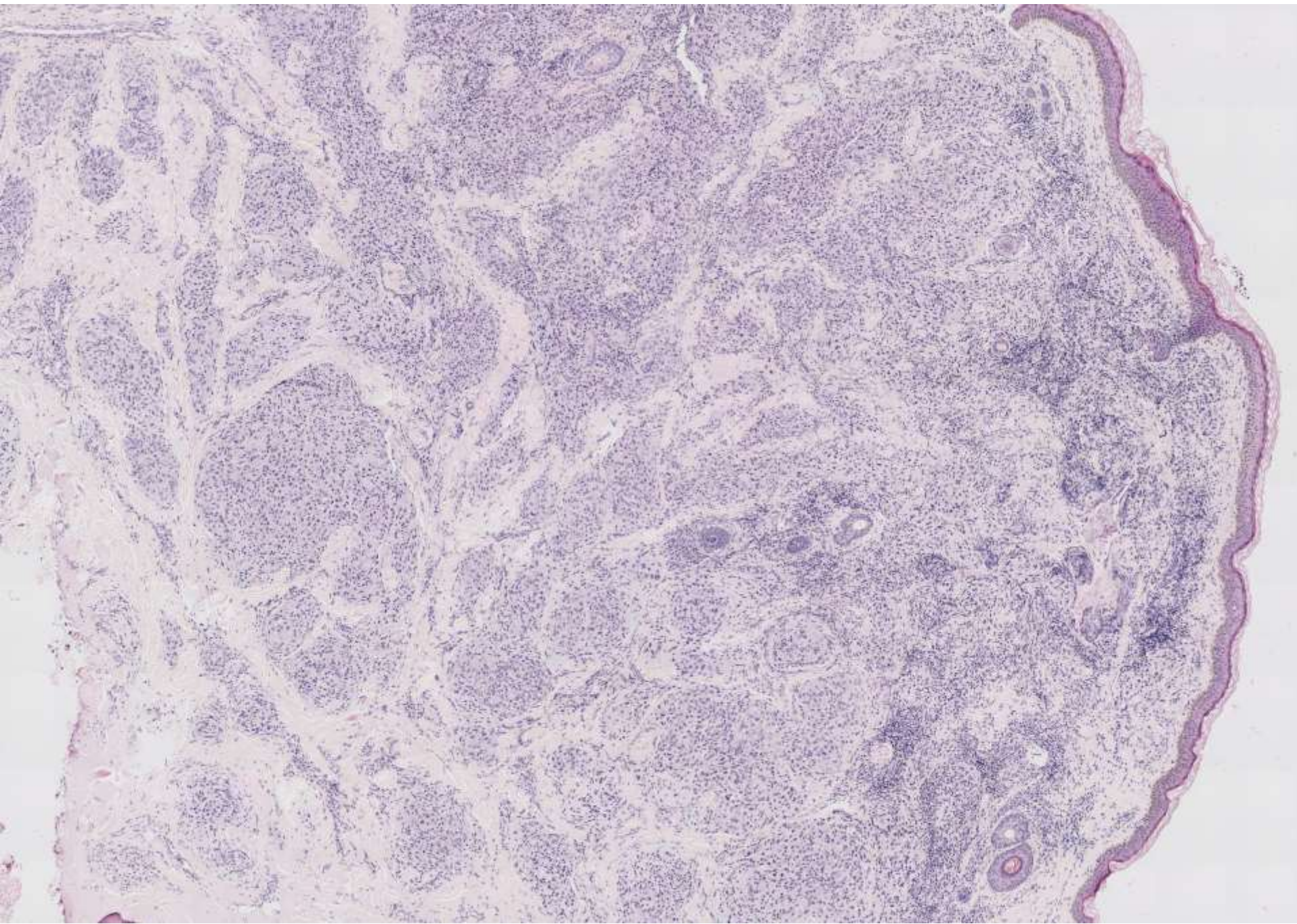
- Cystic kidney lesions
 - Cystic nephroma
 - multilocular cystic renal cell carcinoma
- Oncocytoma with prominent cysts
- Mixed epithelial and stromal tumor
- Collecting duct carcinoma
- Papillary renal cell carcinoma

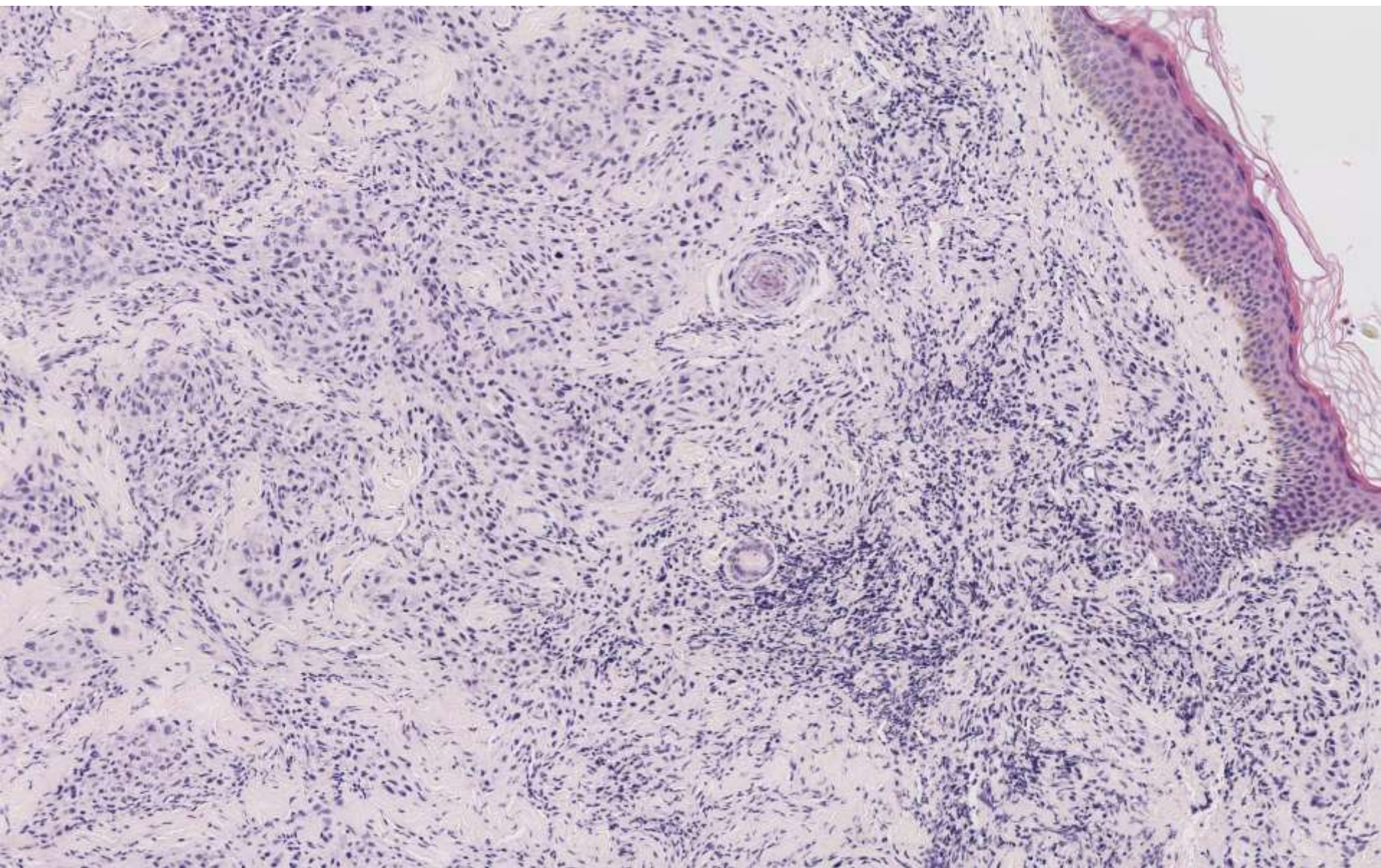
SB 6028

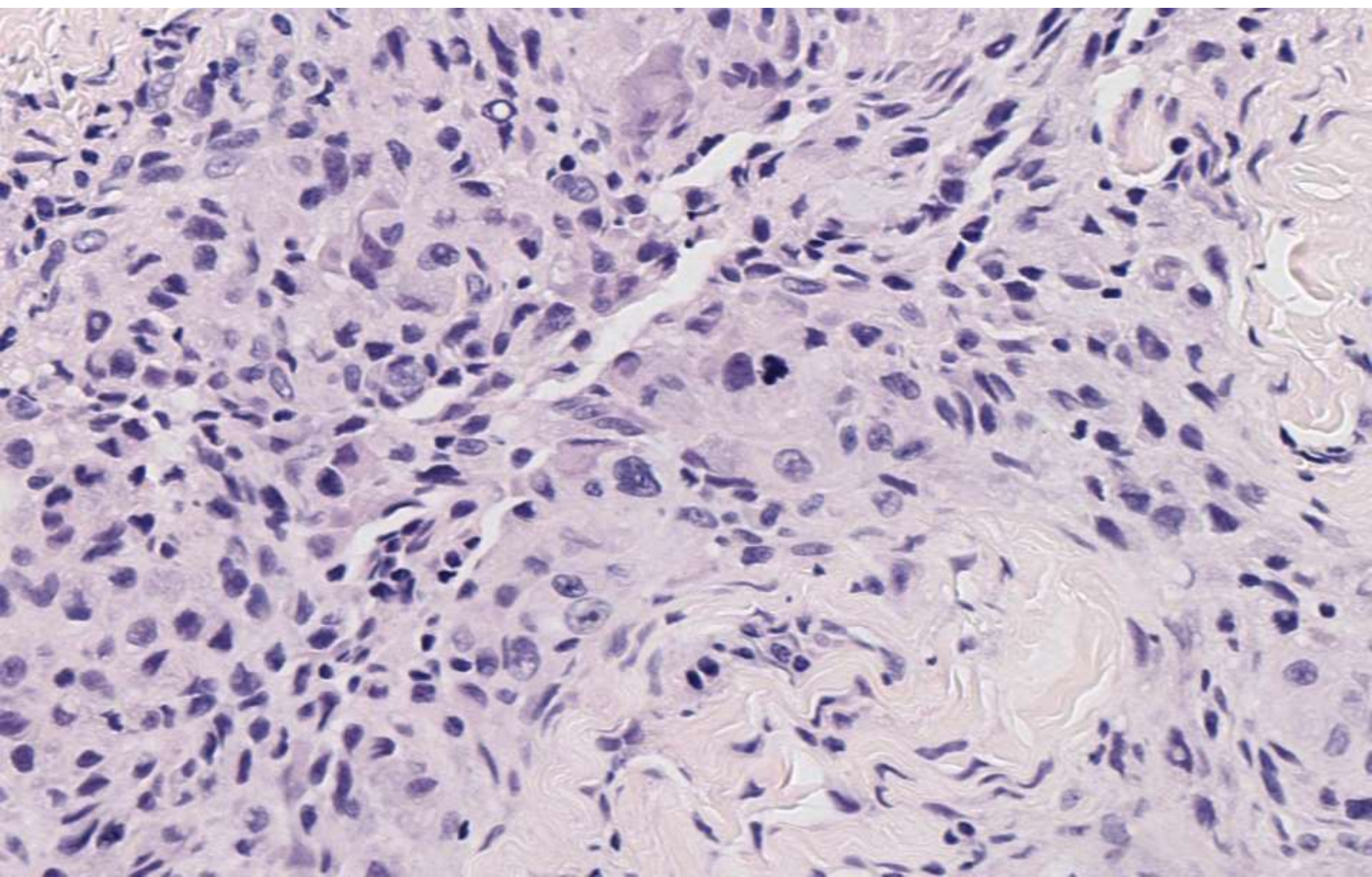
Andrew Horvai; UCSF

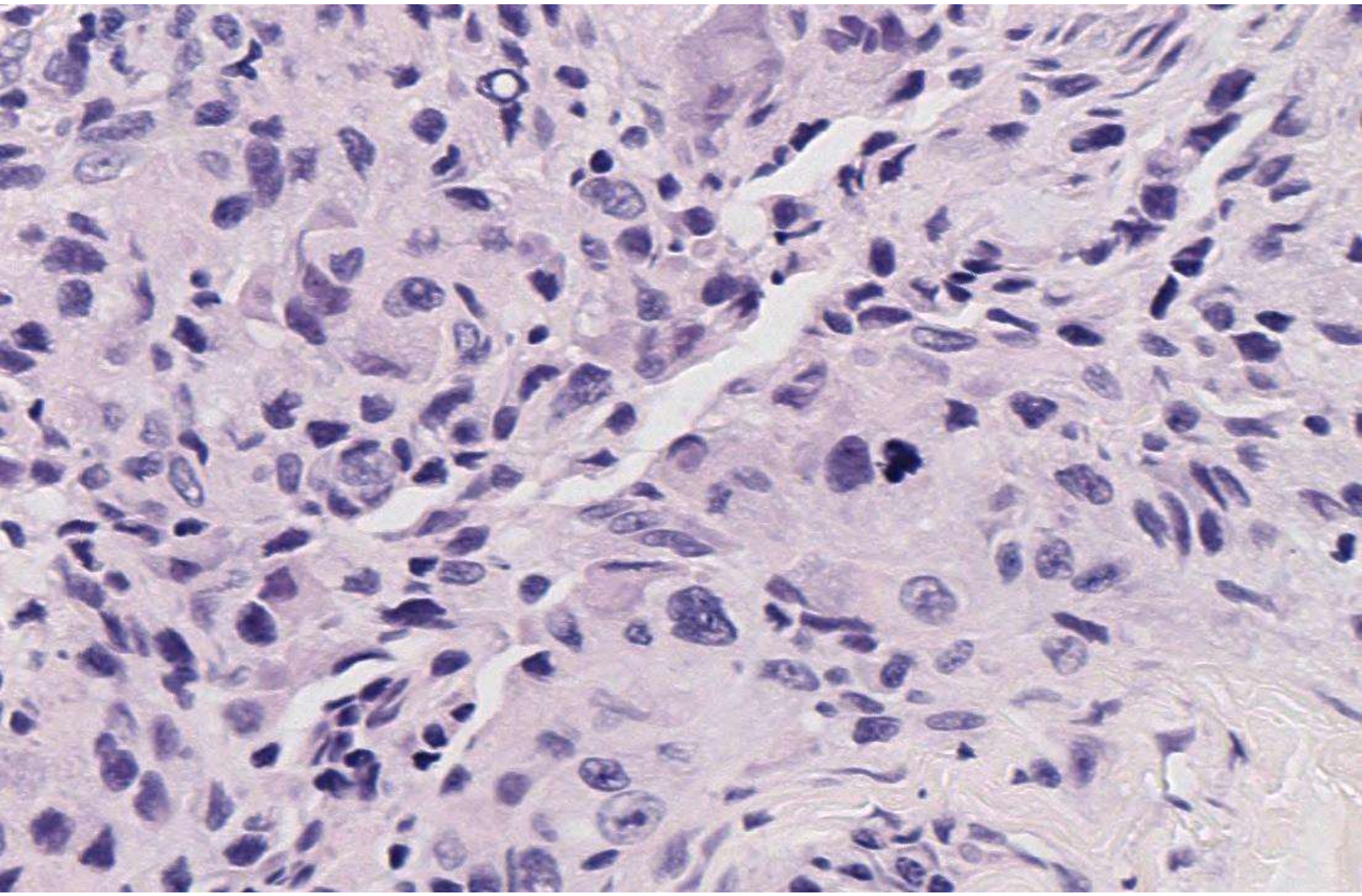
11-year-old boy with nodule on chin.

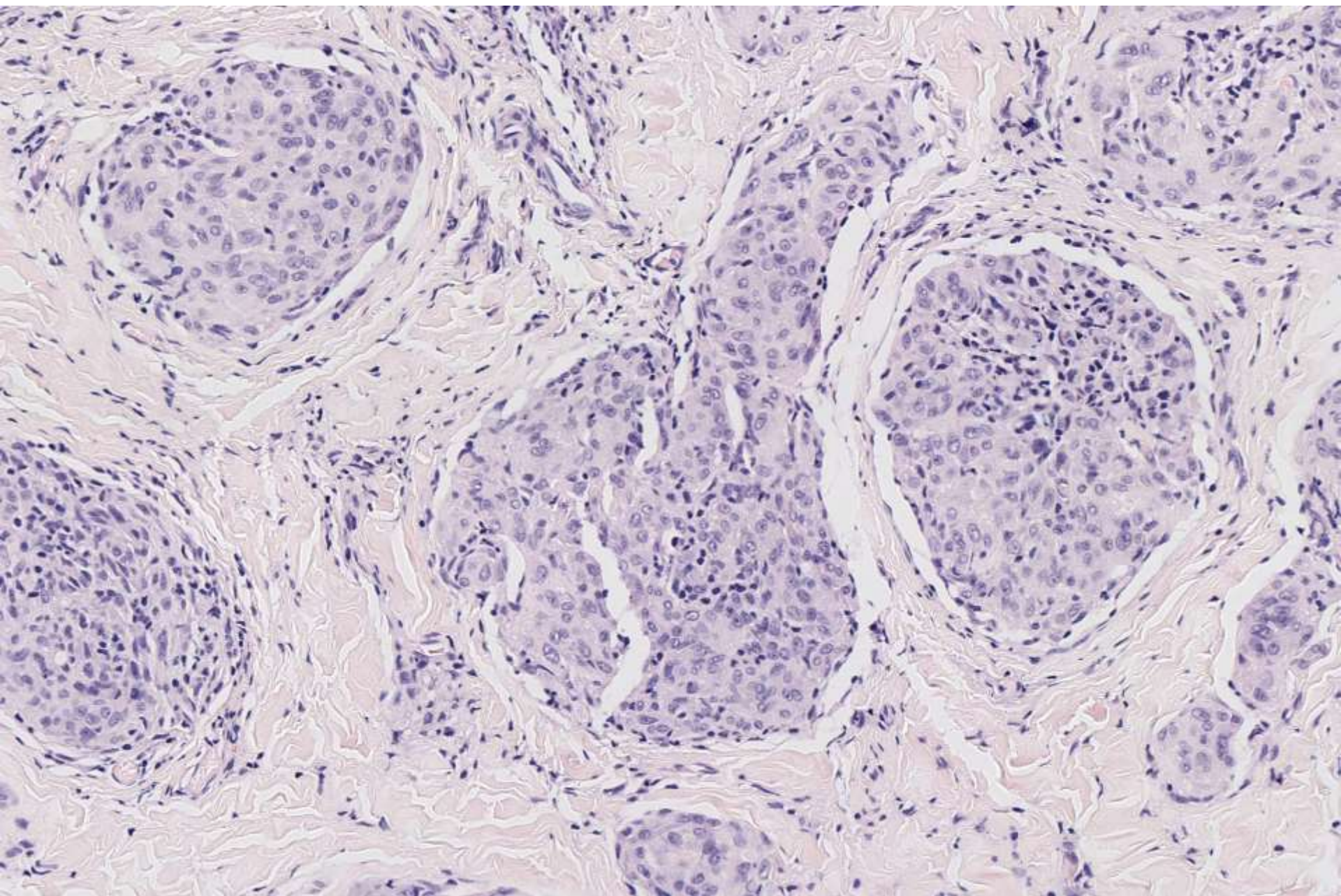


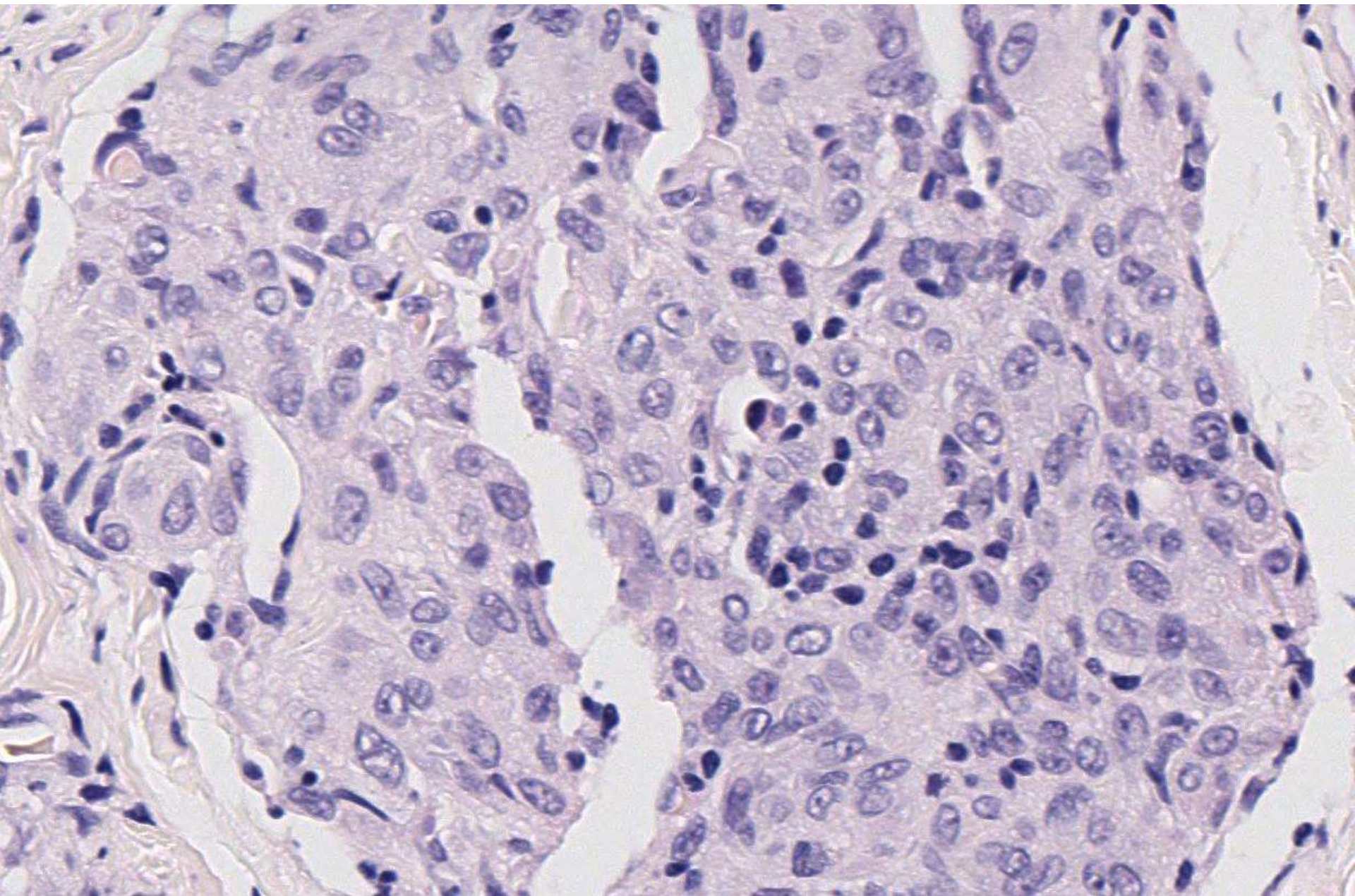






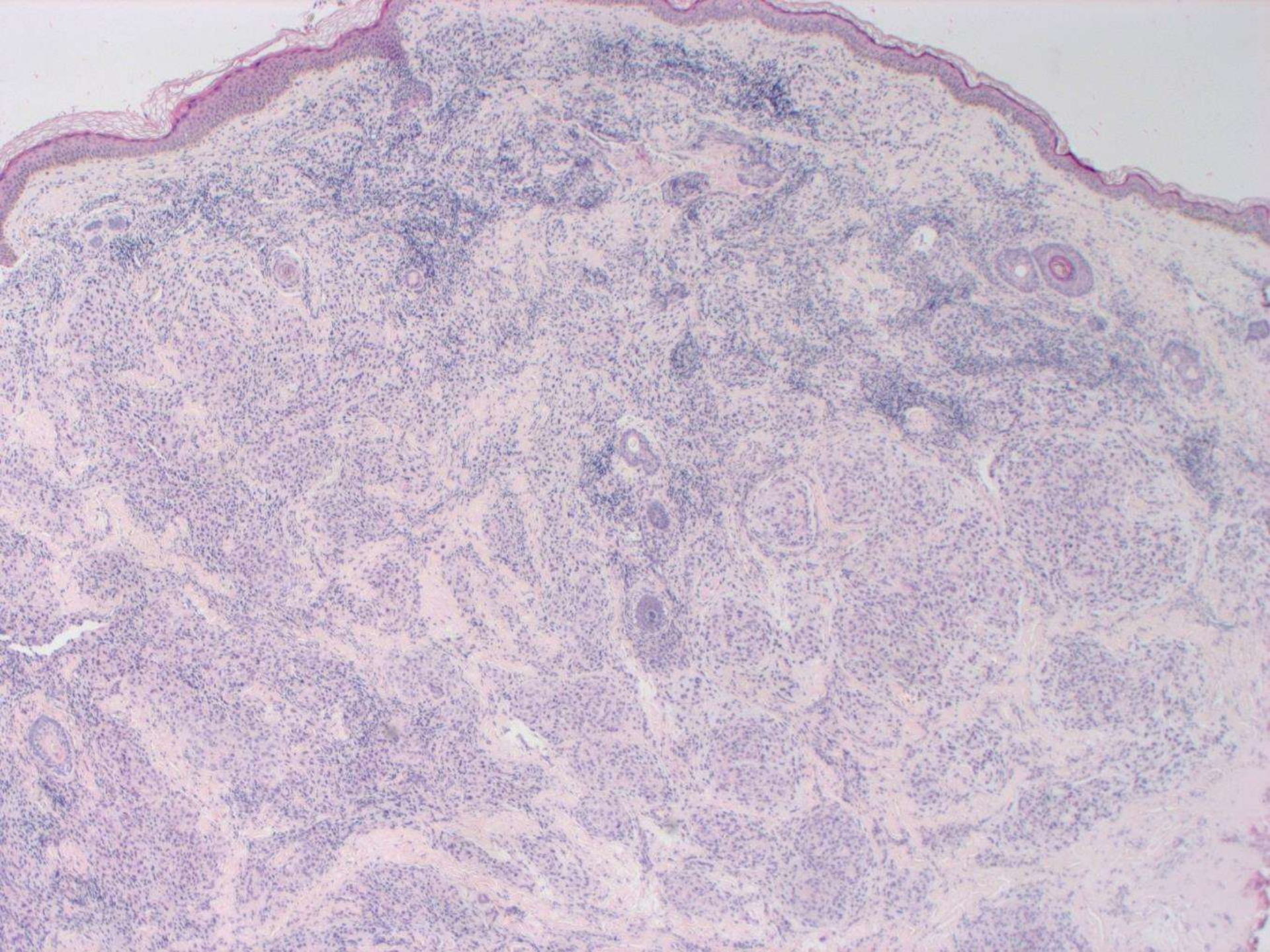


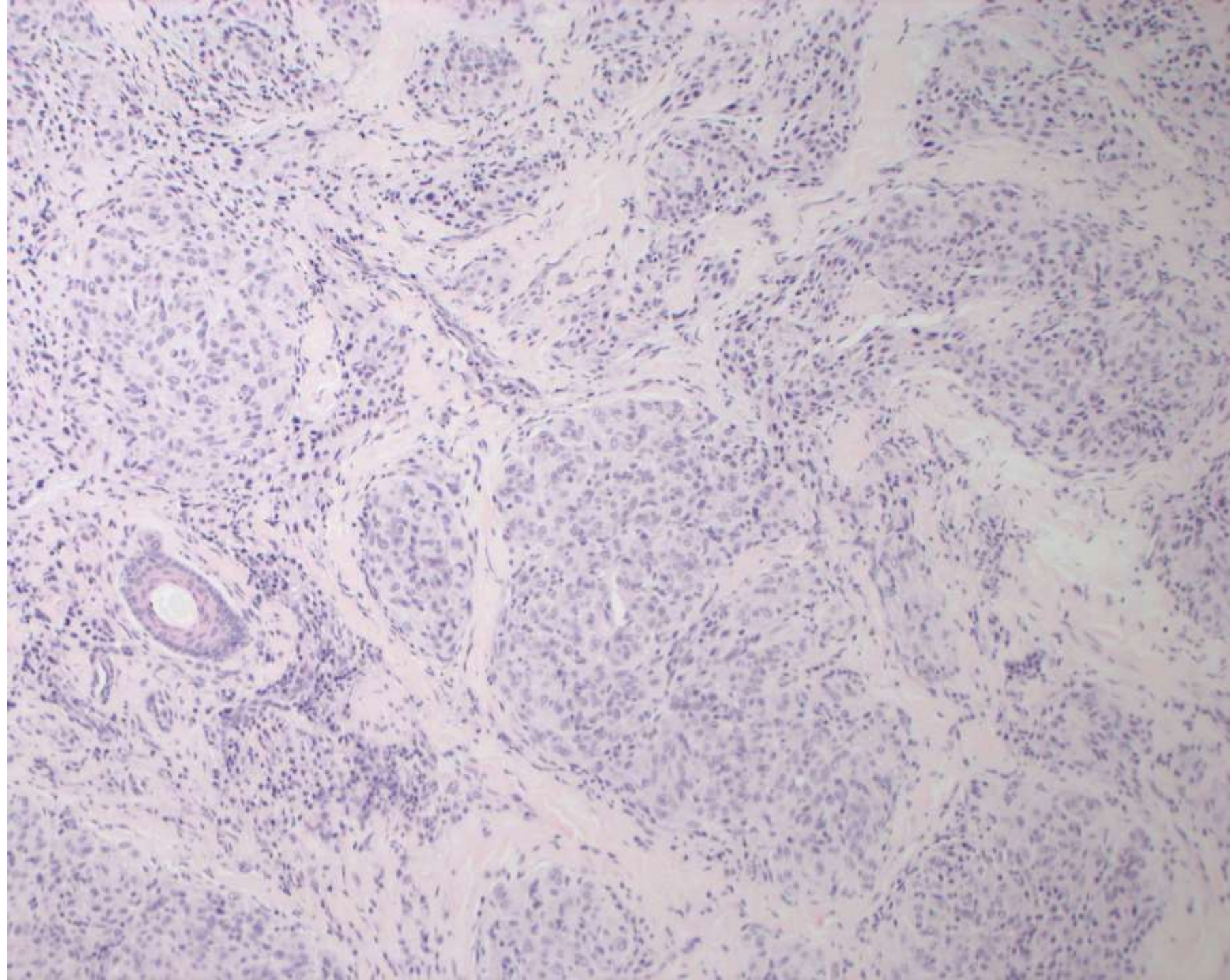


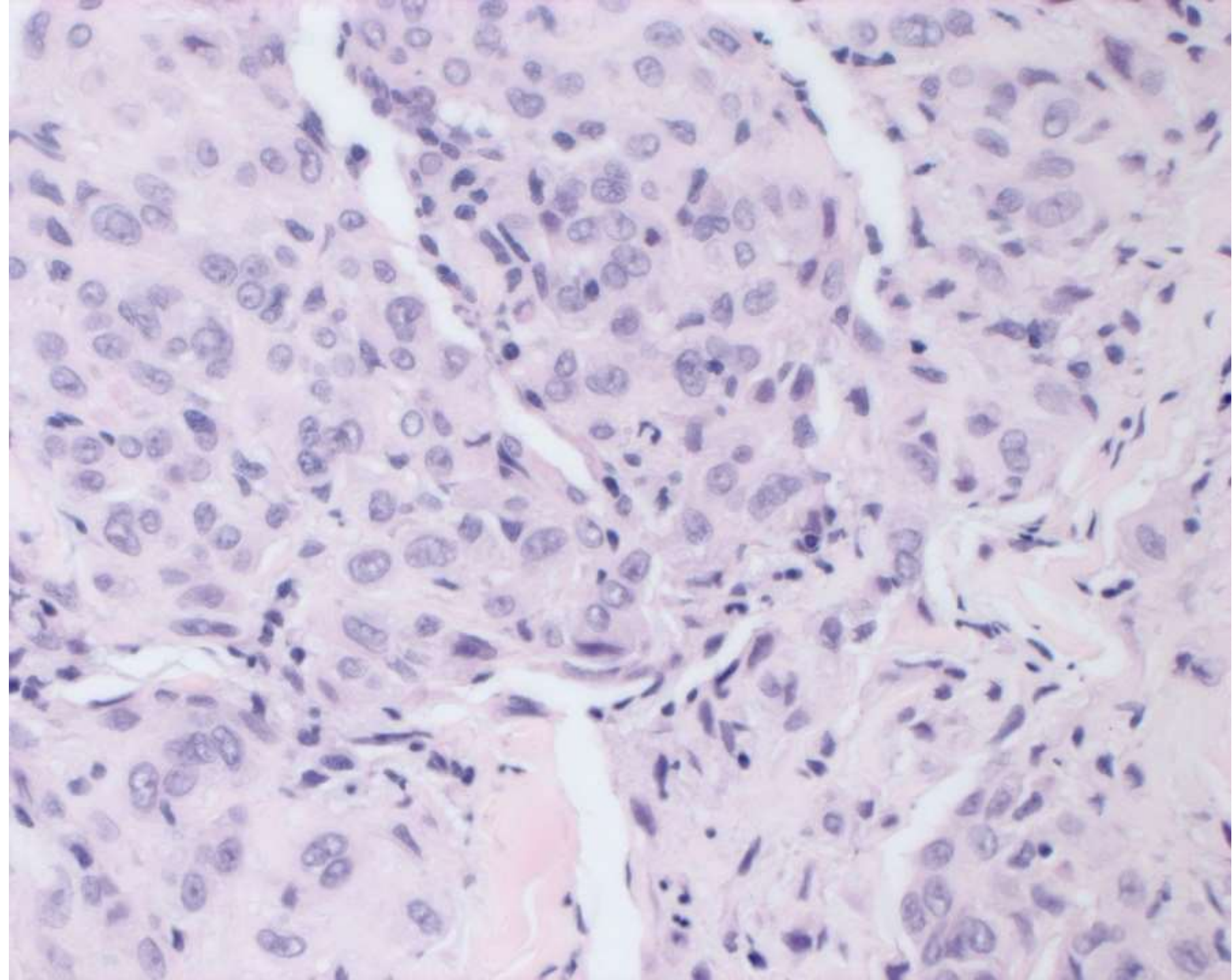


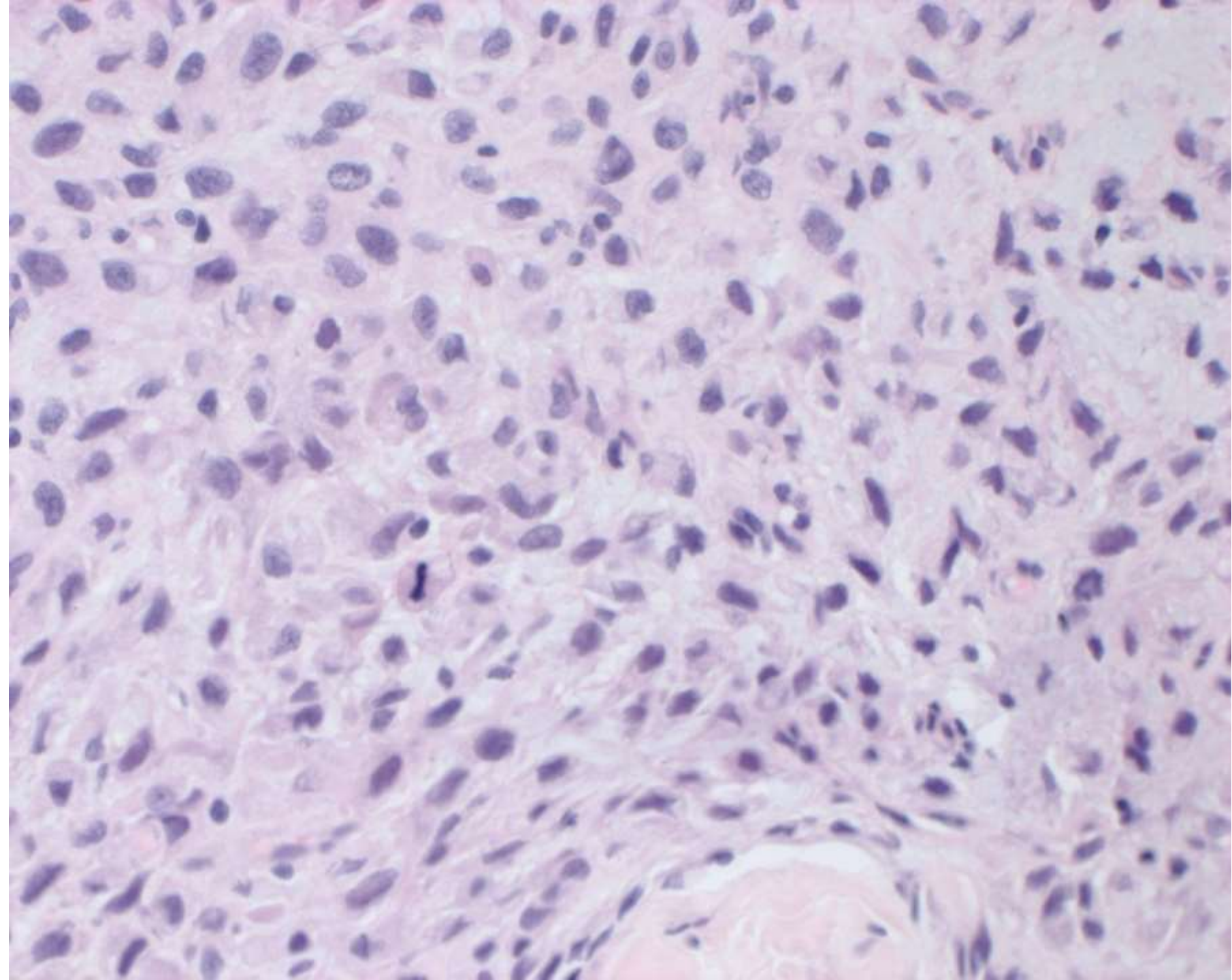
DIAGNOSIS?



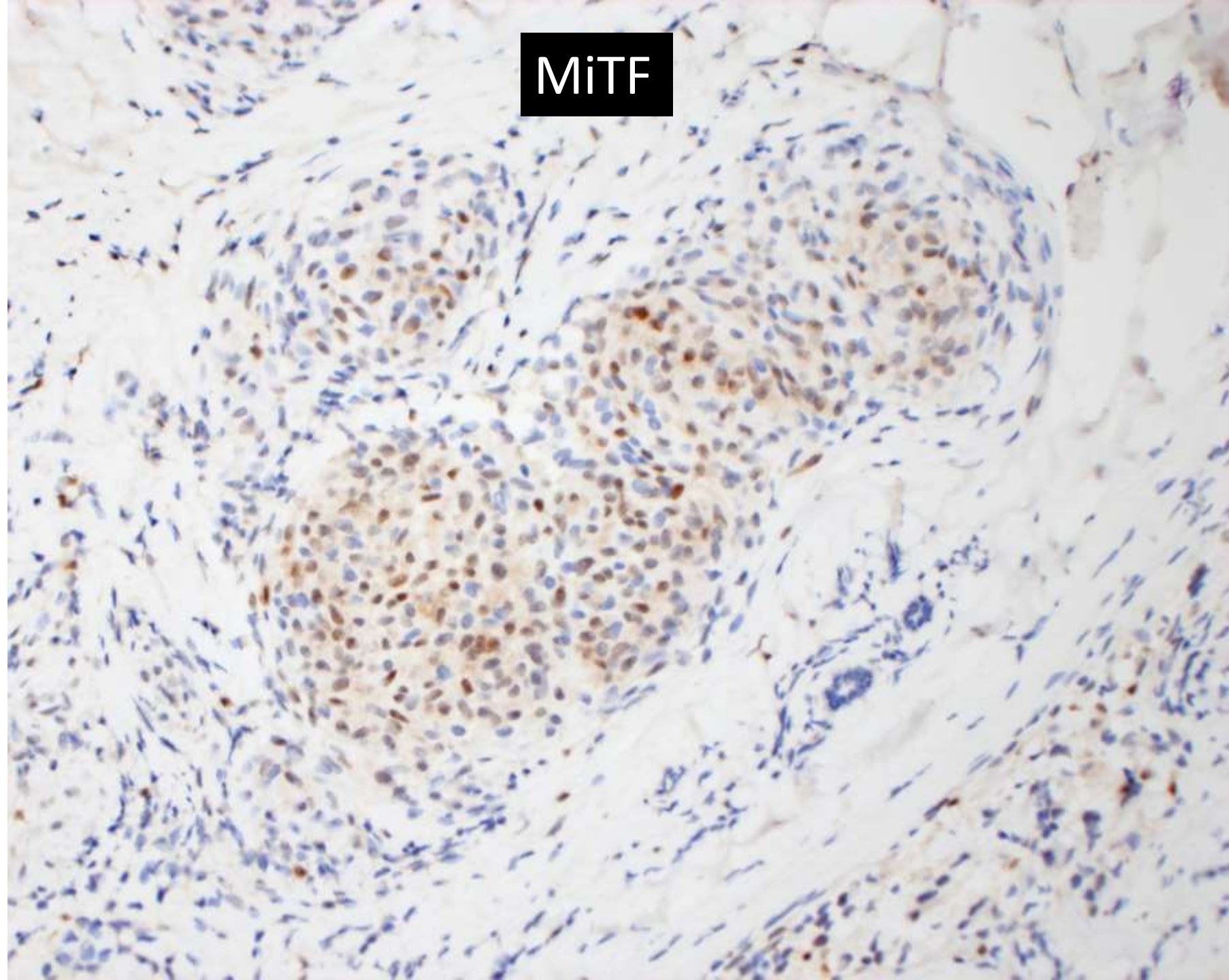








MiTF



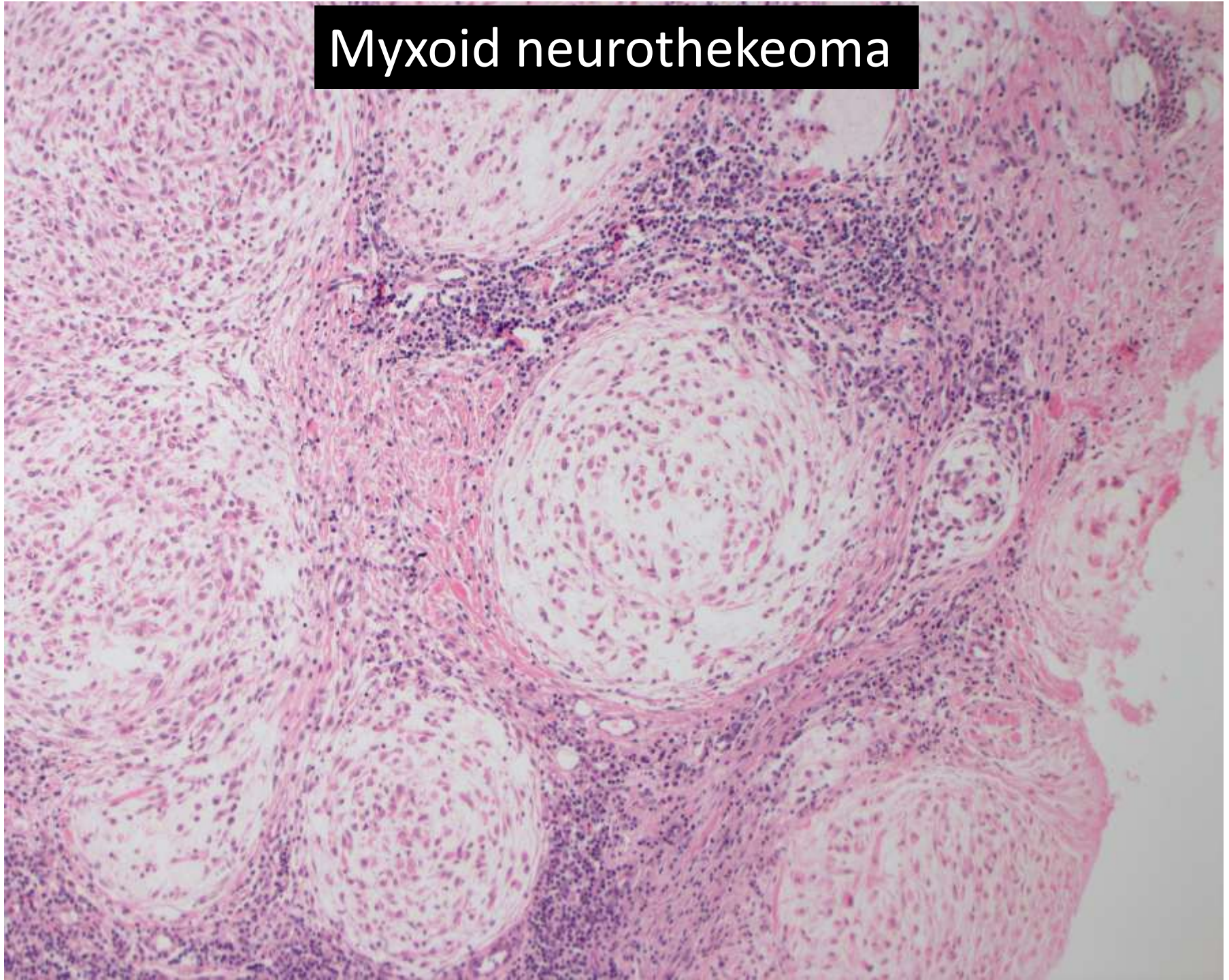
IHC results summary

Positive	Negative
CD68	S100
MiTF	SOX10
CD10	GFAP
NKIC3	Desmin
	SMA
	EMA
	CD34

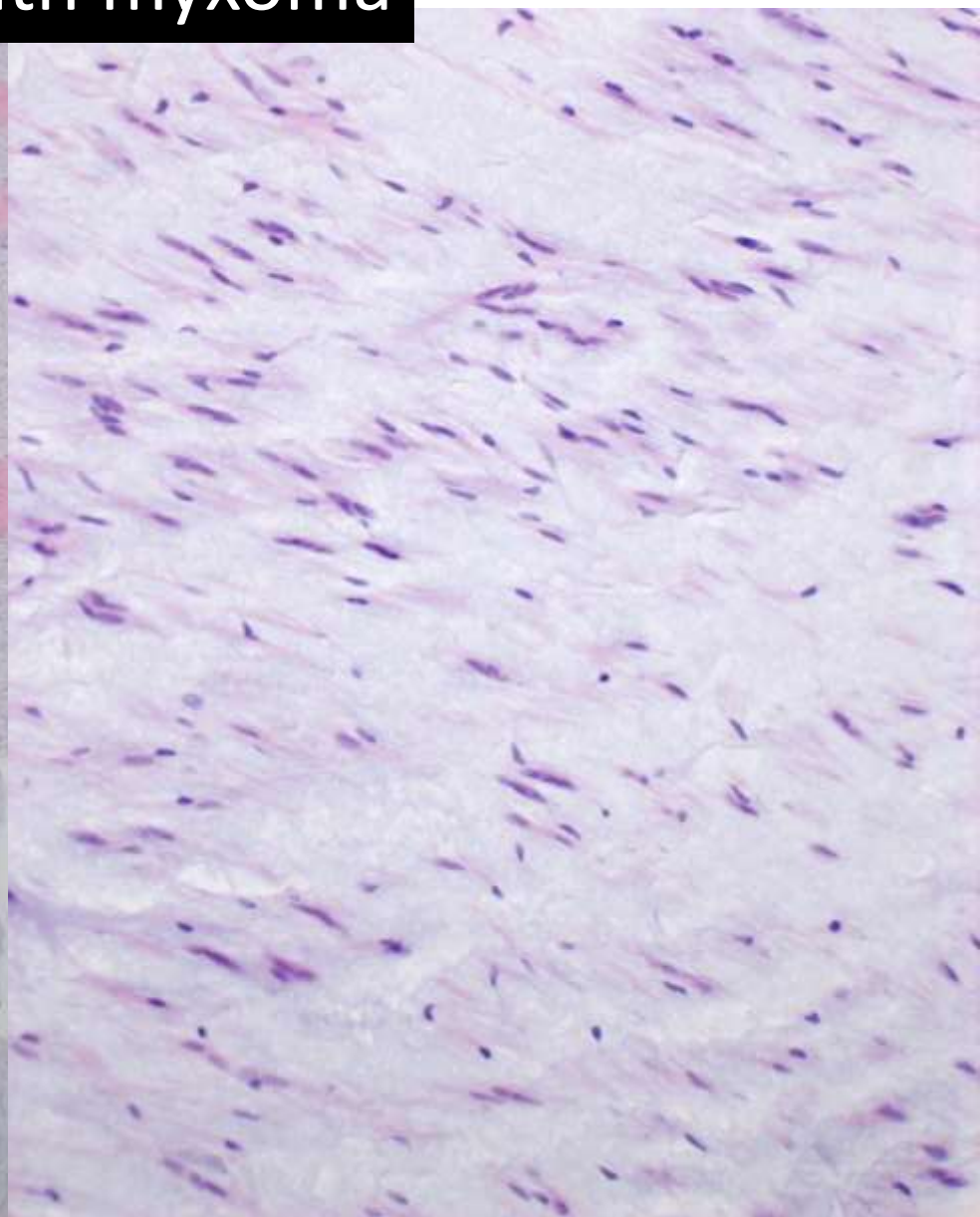
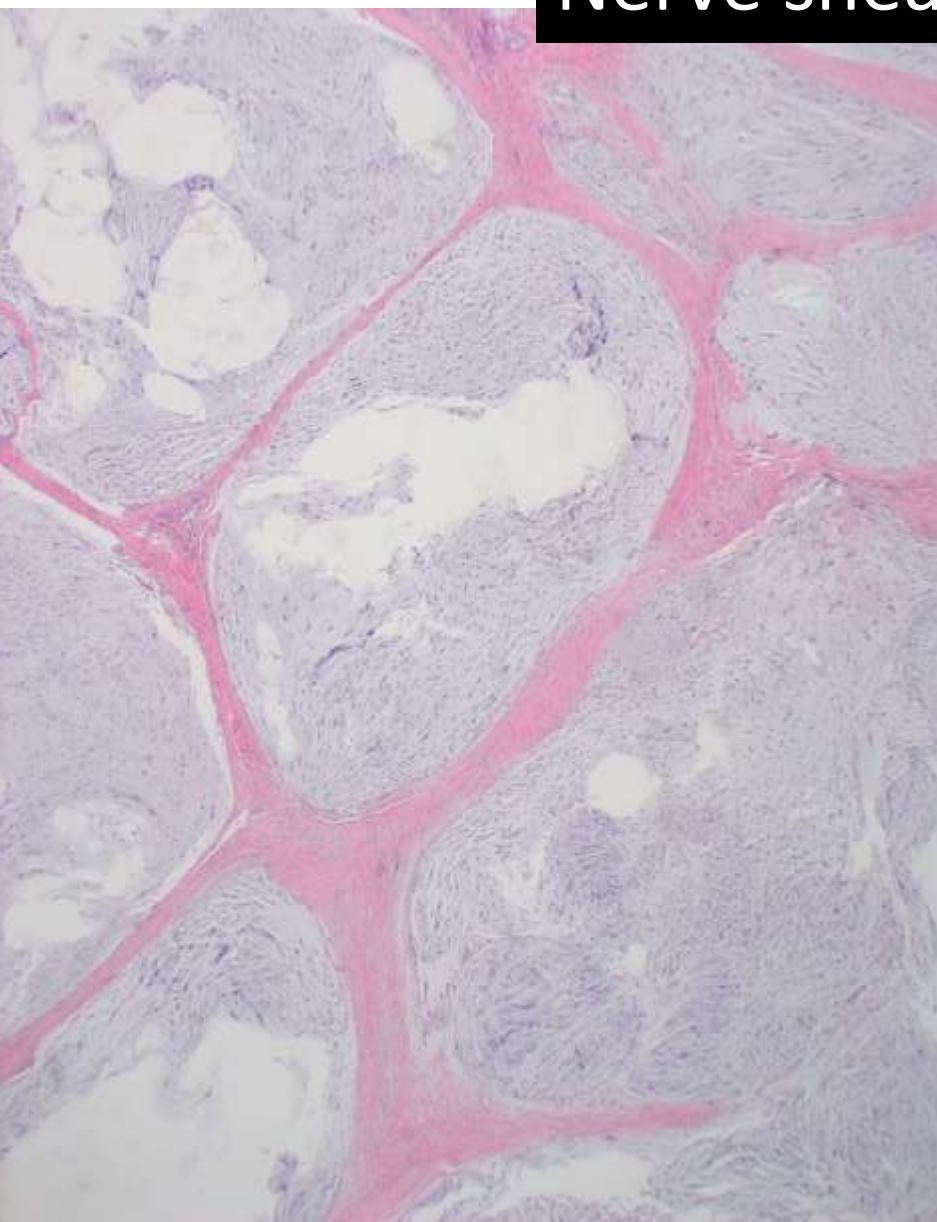
Diagnosis

- Neurothekeoma

Myxoid neurothekeoma



Nerve sheath myxoma

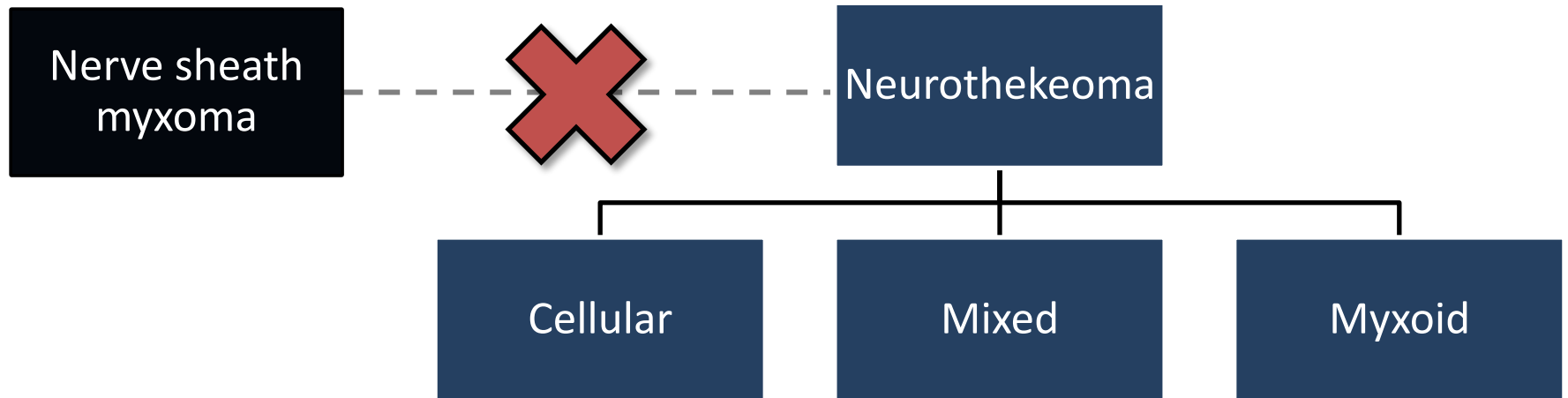


Neurothekeoma vs. nerve sheath myxoma

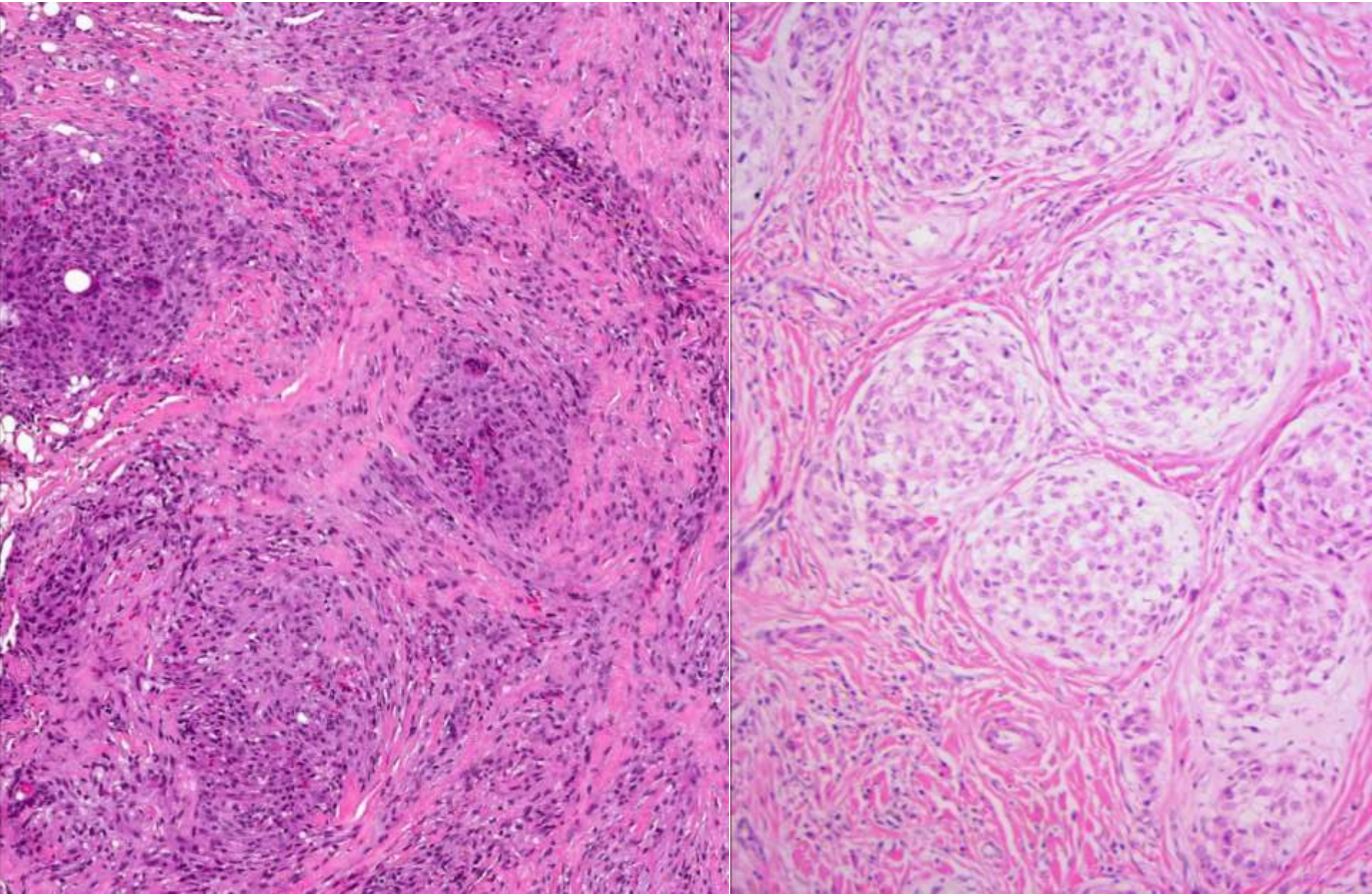
	Neurothekeoma	Nerve sheath myxoma
Clinical	Children, young adults Rare recurrence	Young to mid adult ~50% recurrence
Location	Proximal upper extremity, face	Distal extremity
Growth pattern	Multinodular, poorly circumscribed	Sharply circumscribed
Myxoid	None to focal	Diffuse
Cells	Plump, epithelioid, stellate Atypia common	Bipolar, long processes Minimal atypia
Mitoses	>5 / 10 hpf possible	<1 / 10 hpf

S100, SOX10	-	+
NKIC3, MiTF	+	-/+
SMA	+/-	-

Classification



Plexiform fibrohistiocytic tumor



Neurothekeoma vs. Plexiform FH tumor

	Neurothekeoma	Plexiform FH tumor
Clinical	Children, young adults, rare recurrence	
Location	Proximal upper extremity, face	Proximal upper extremity
Growth pattern	Multinodular, plexiform	
Myxoid	None to focal	
Cells	Plump, epithelioid, stellate Atypia common	Biphasic: epithelioid and spindled, osteoclasts
Mitoses	>5 / 10 hpf possible	<1 / 10 hpf

MitF	+	-
NKIC3	+	+
SMA	+/-	+/-

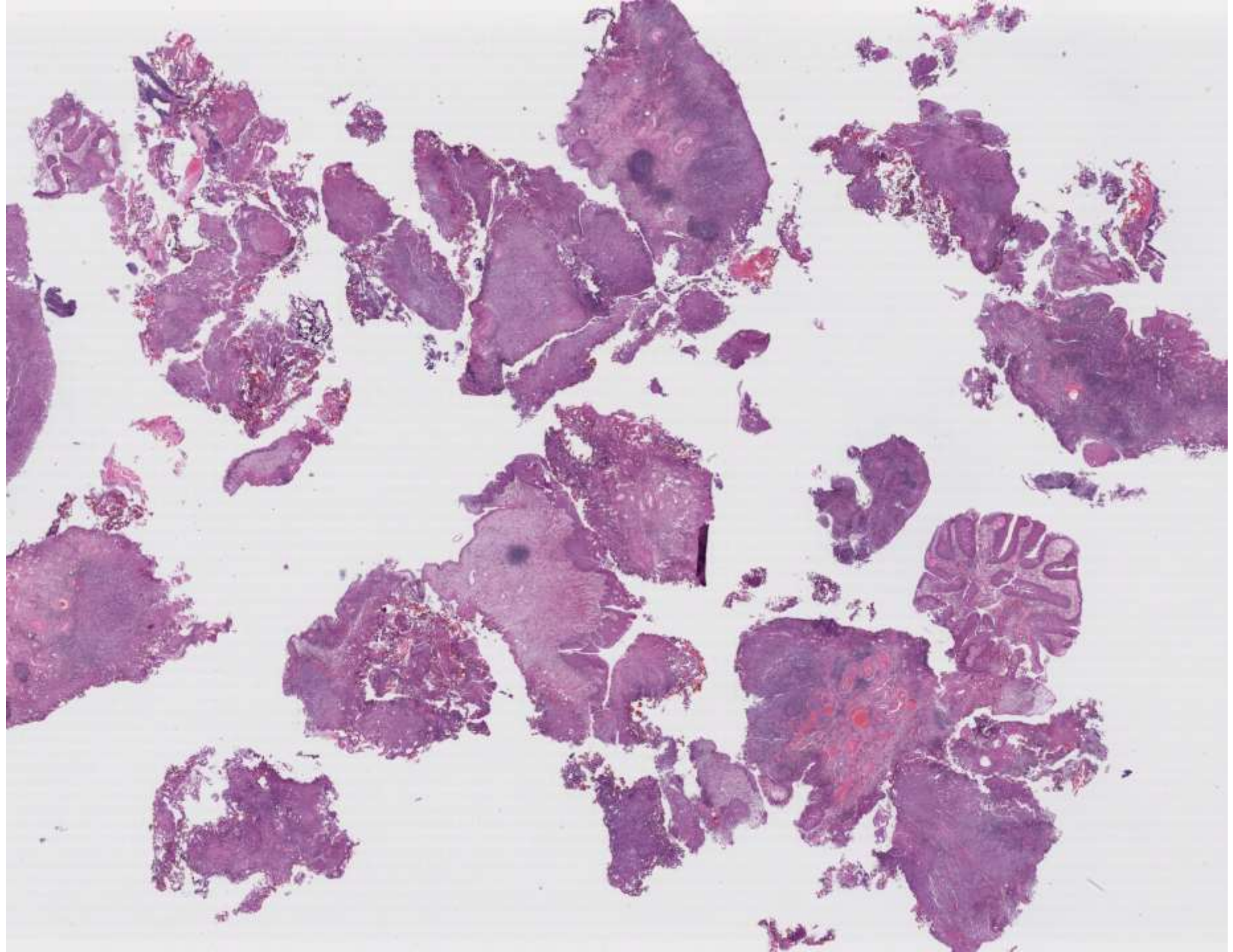
References

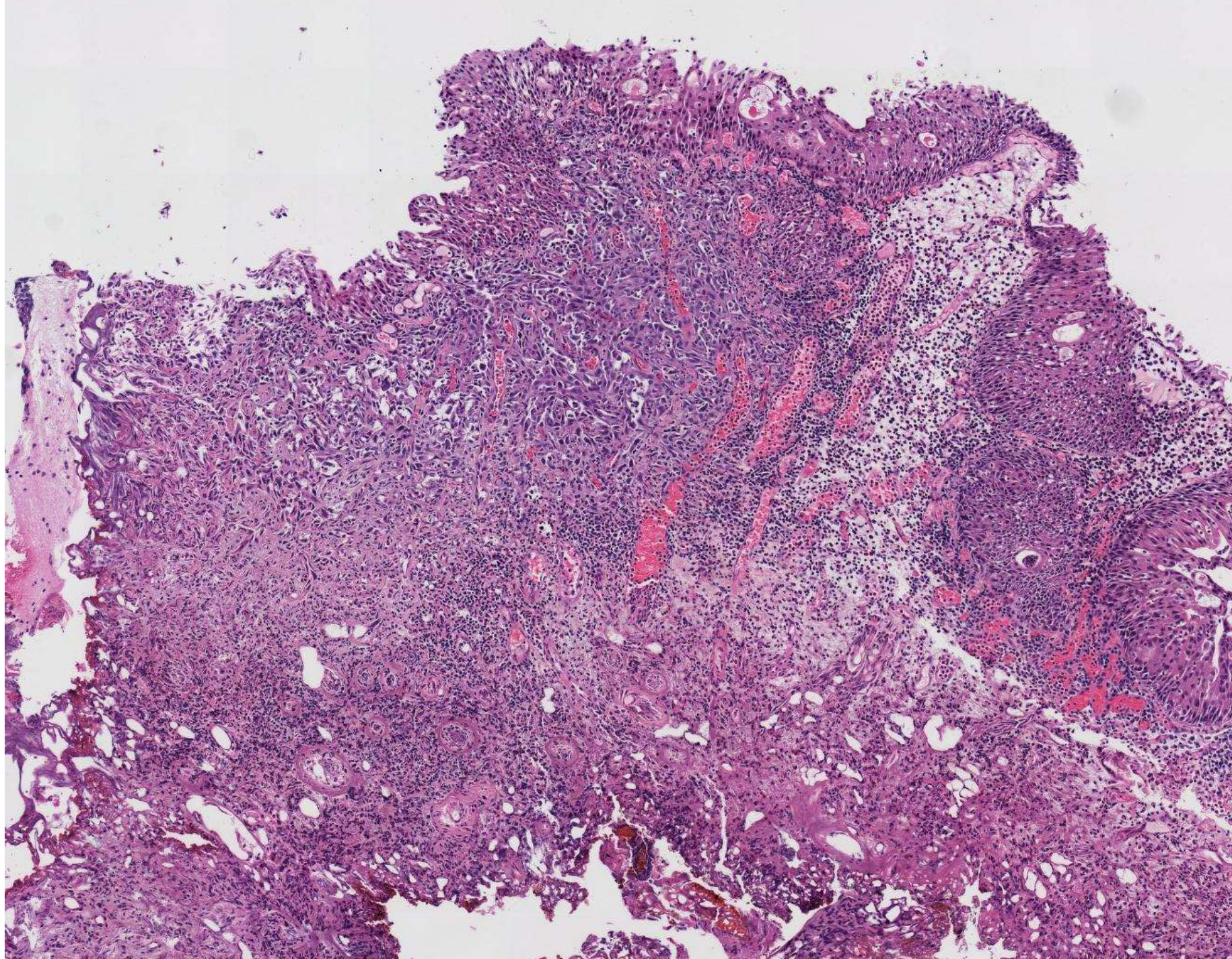
- Fox MD et al. Am J Dermatopath 2012; 34:157-60
- Hornick JL and Fletcher CD. Am J Surg Pathol 2007; 31:329-40.

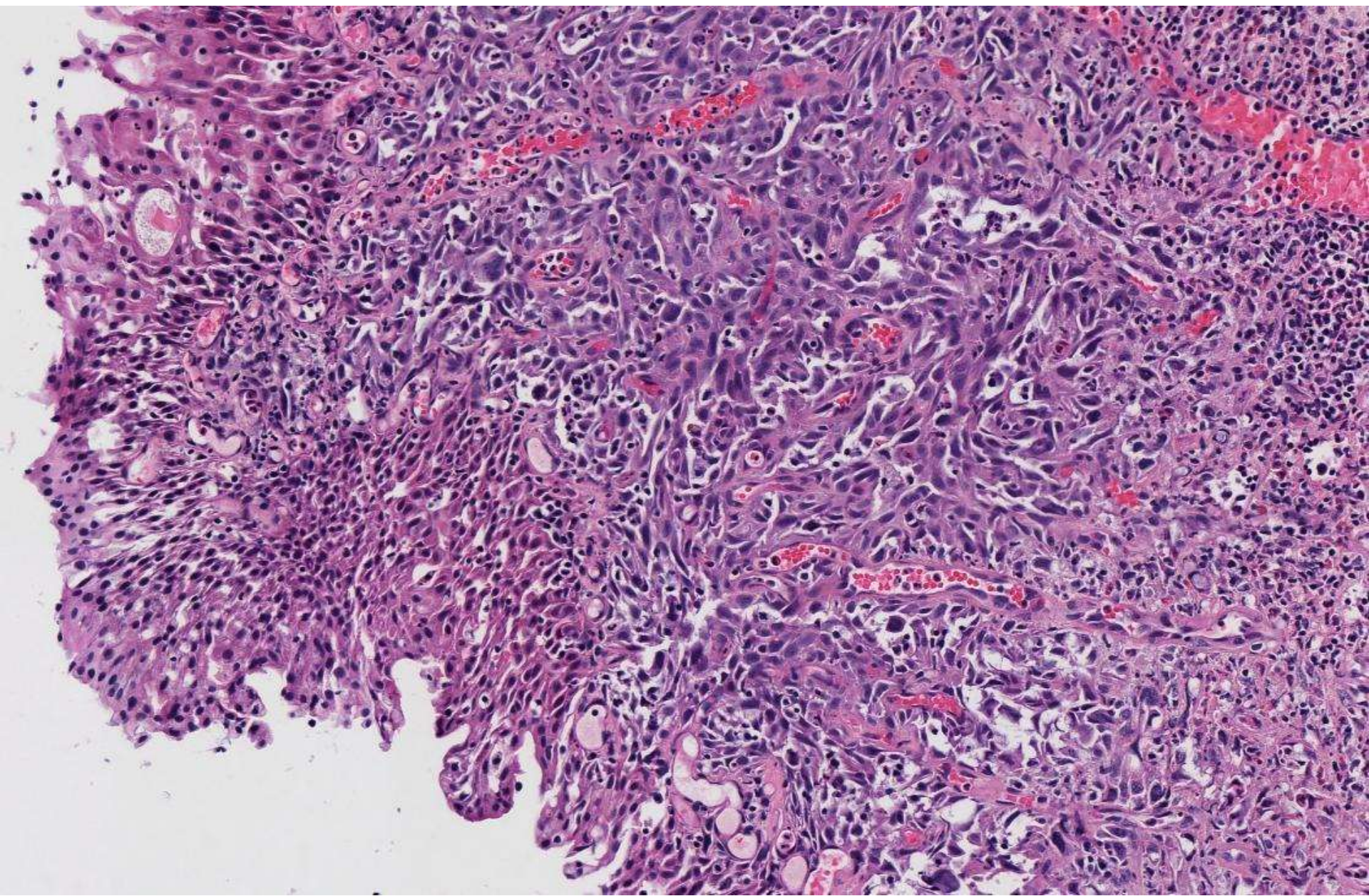
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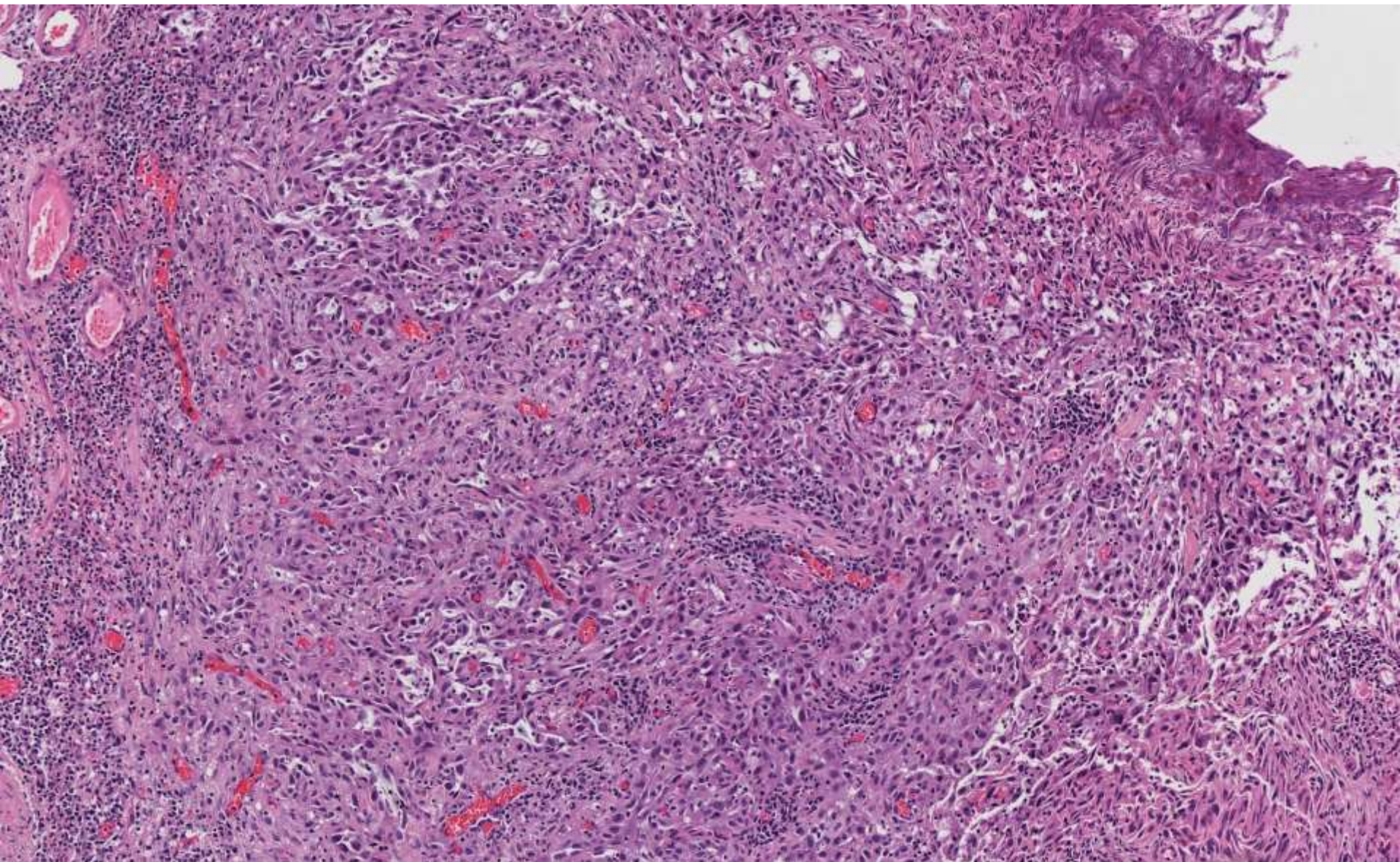
Ankur Sangoi; El Camino Hospital

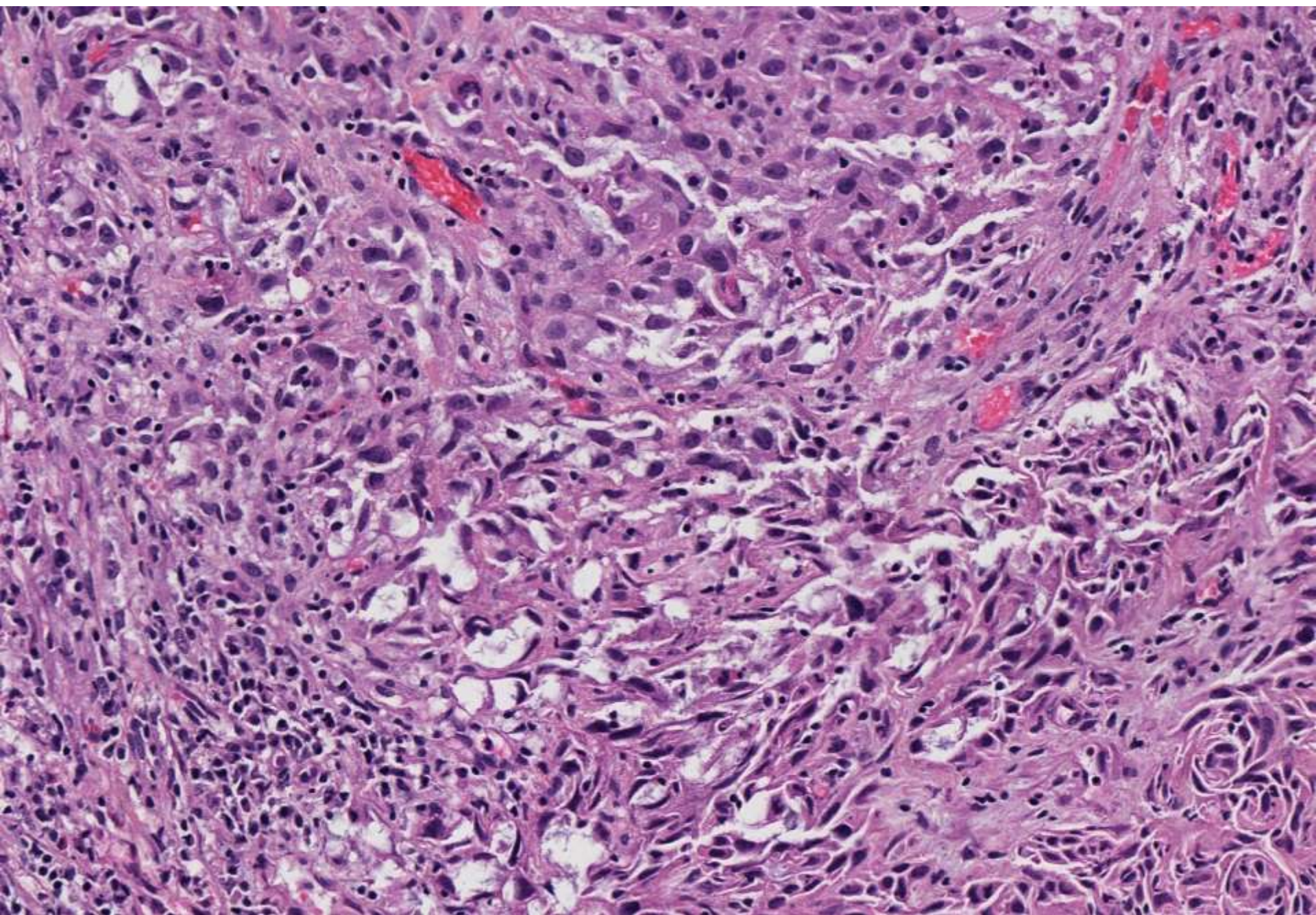
79-year-old female with bladder
tumor resection.

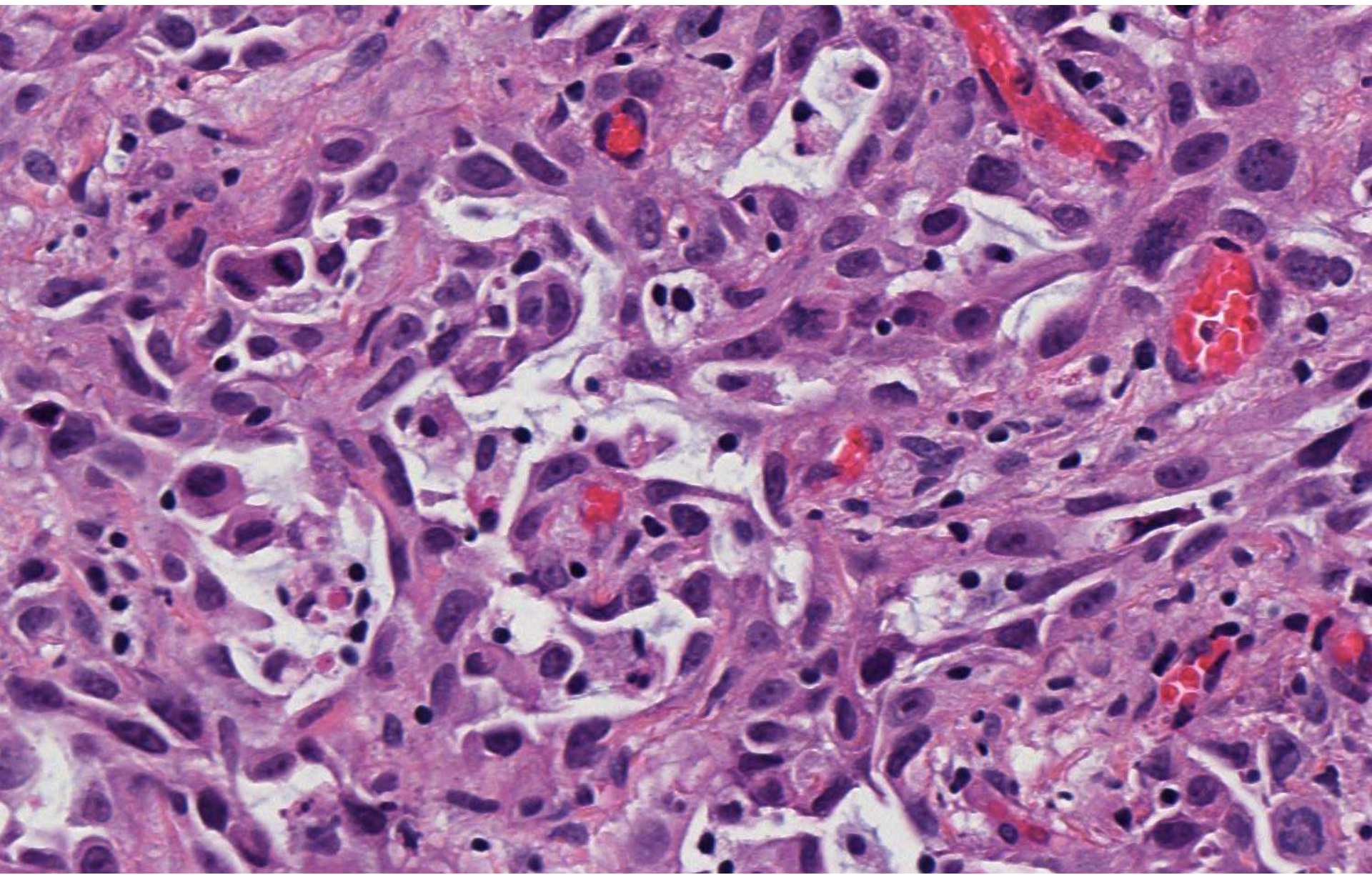


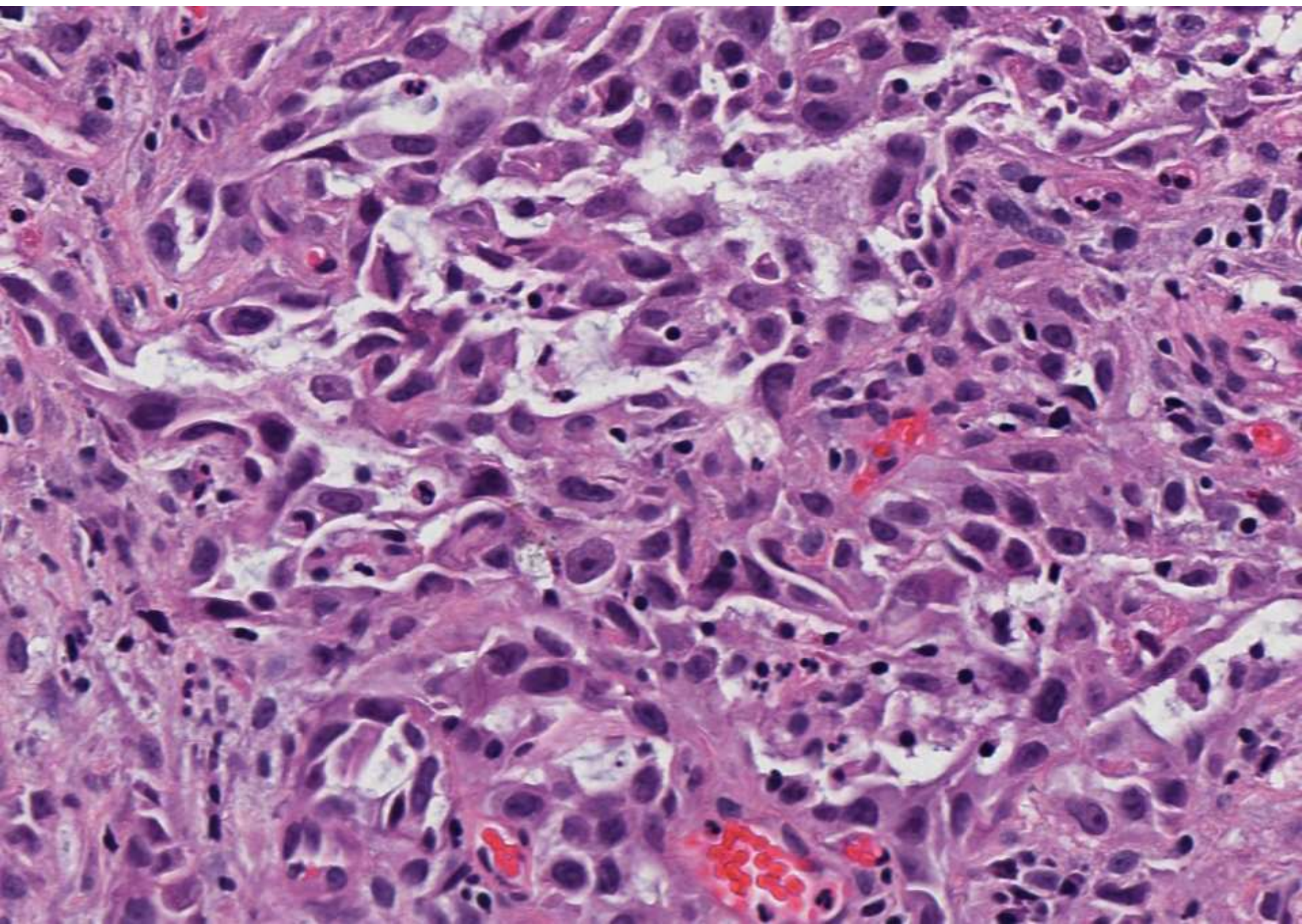












DIAGNOSIS?



DDx

- **Angiosarcoma/epitheloid angiosarcoma**
 - Can be keratin+
 - Usually not CK7/CK20/GATA3/p63+
 - + for vascular markers
 - History of radiation
- **Sarcomatoid urothelial carcinoma (carcinosarcoma)**
 - Unequivocal spindle cell morphology
 - Often decreased/negative keratin expression
- **Clear cell adenocarcinoma**
 - Usually has admixed more classic patterns
 - Tubular/tubulopapillary
 - Lack pseudoluminae with inflammatory cells
 - Admixed endometriosis (females)
 - PAX8+, usually CK20/GATA3 negative

Pseudoangiosarcomatous Urothelial Carcinoma of the Urinary Bladder

Gladell P. Paner, MD,† Roni Michelle Cox, MD,‡ Kyle Richards, MD,† Ashwin Akki, MD, PhD,*
Neriman Gokden, MD,‡ Antonio Lopez-Beltran, MD,§ Thomas Krausz, MD,*
Jesse K. McKenney, MD,|| and Gary D. Steinberg, MD†*

Am J Surg Pathol • Volume 38, Number 9, September 2014

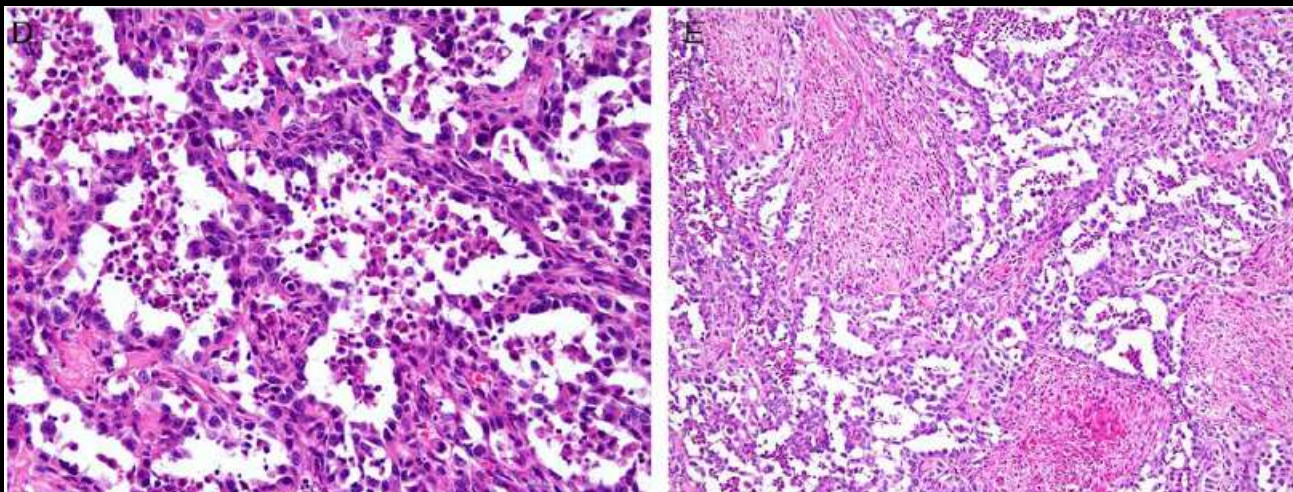


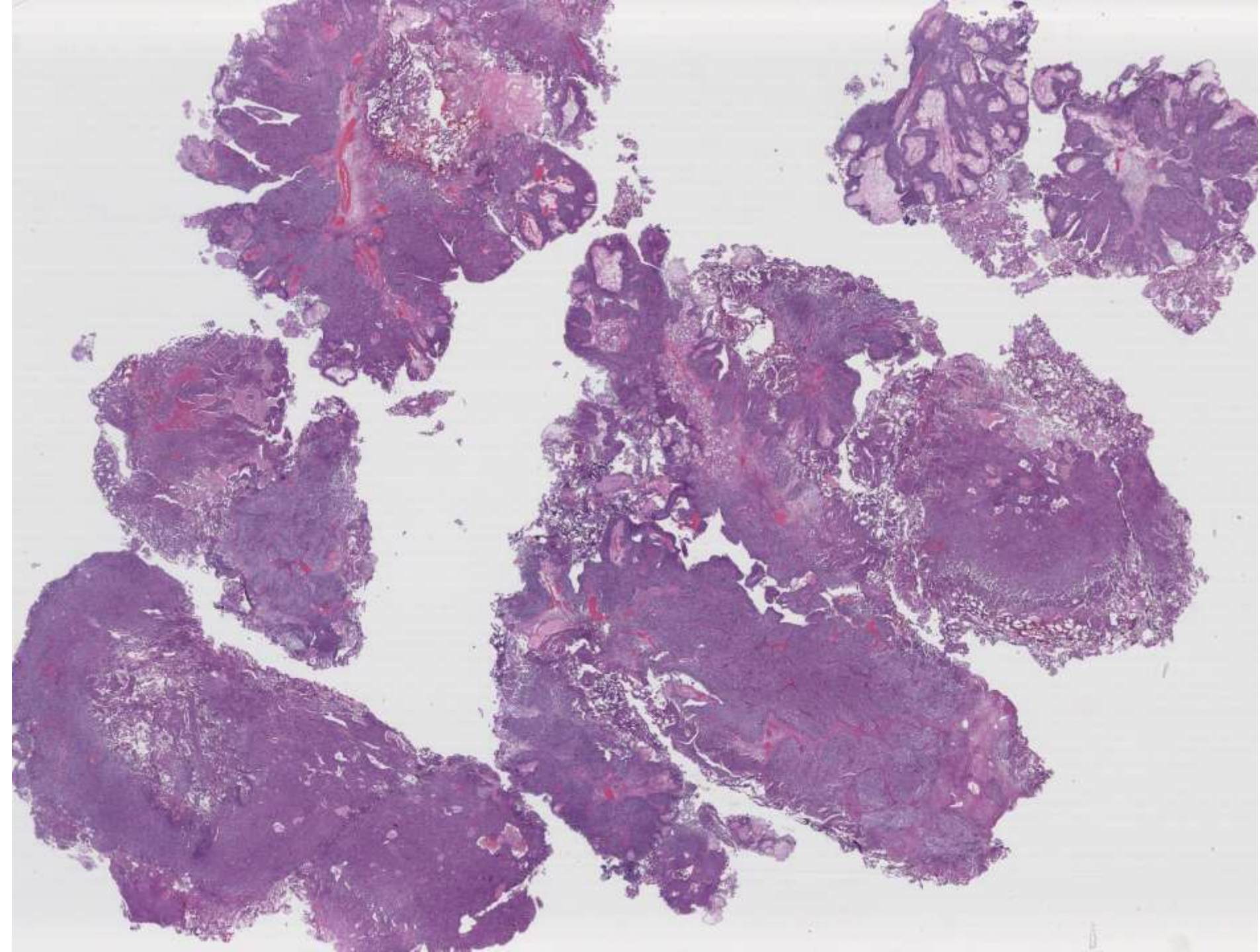
TABLE 2. Clinicopathologic Features of Pseudoangiosarcomatous Carcinoma of the Urinary Bladder

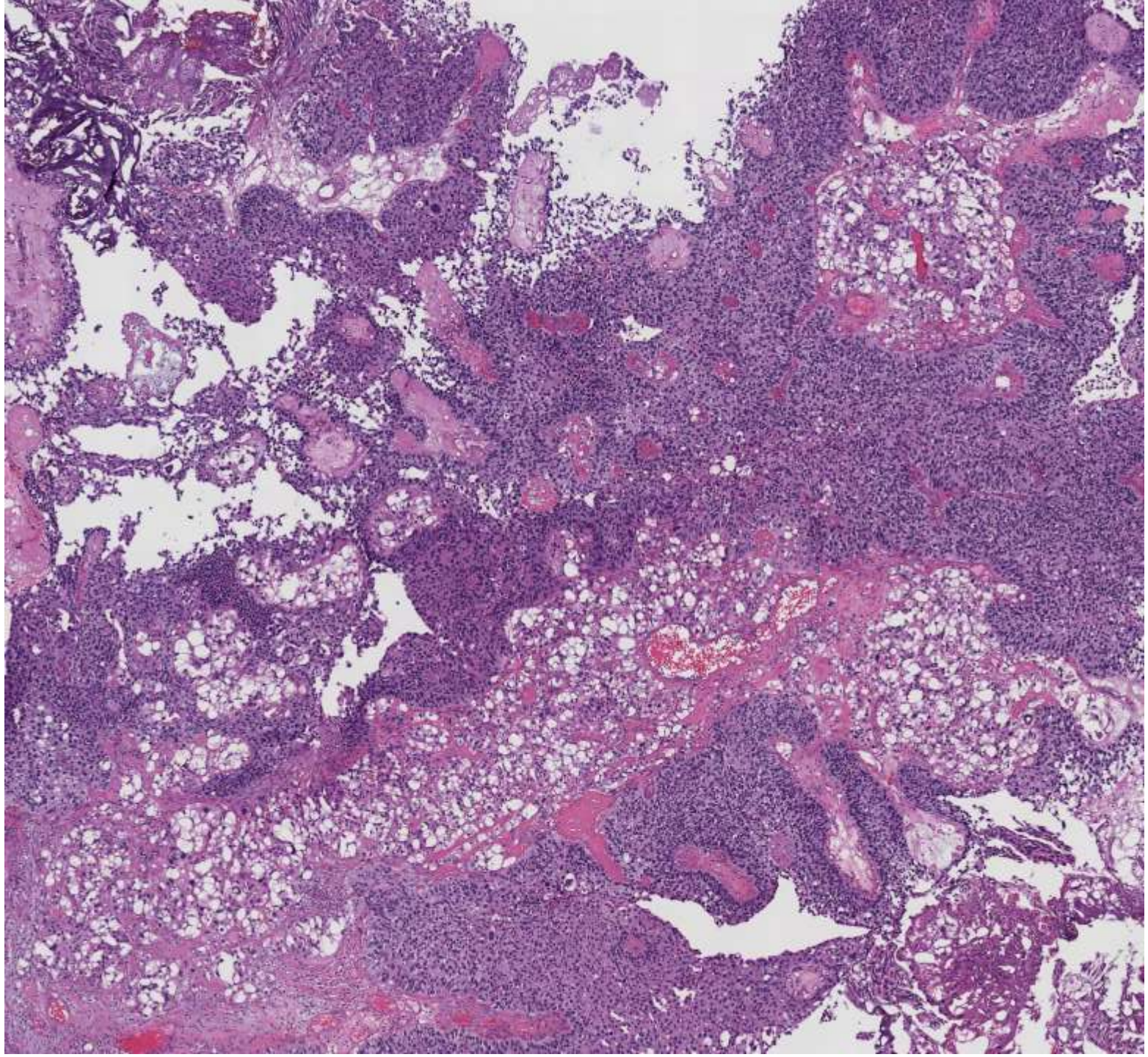
Case	Age (y)/ Sex	Location	Treatment	Pseudoangiosarcomatous Component (%)	Nonpseudoangiosarcomatous Component (%)	Stage at Cystectomy	Outcome
1	55/M	Trigone and anterior wall	Cystoprostatectomy, pelvic lymph node dissection, and postoperative chemotherapy	Urothelial carcinoma (70%)	Conventional urothelial carcinoma (25%), sarcomatoid spindle cell carcinoma (5%)	pT4a,N0	Metastasis to bone at 9 mo, DOD at 12 mo
2	70/M	Posterior and left lateral walls	Cystoprostatectomy, pelvic lymph node dissection, and postoperative chemotherapy	Urothelial carcinoma (65%)	Small cell carcinoma (30%), micropapillary carcinoma (~3%), glandular differentiation (~3%)	pT4a,N2	Metastasis to retroperitoneum at 2 mo, DOD at 22 mo
3	61/M	Trigone, lateral and anterior walls	Preoperative radiation, cystoprostatectomy, and pelvic lymph node dissection	Urothelial carcinoma (60%), squamous differentiation (15%)	Urothelial carcinoma (5%), squamous differentiation (15%)	pT3b,N0	DOD at 5 mo
4	72/M	Extensive infiltration of bladder wall	Cystoprostatectomy and pelvic lymph node dissection	Urothelial carcinoma (55%), squamous differentiation (35%)	Urothelial carcinoma (5%), squamous differentiation (5%)	pT3b,N0	DUC at 3 mo; concomitant pT3a,N0 Gleason 4 + 5 = 9 prostatic adenocarcinoma
5	78/M	Hutch diverticulum at left postero-lateral wall	Cystoprostatectomy and pelvic lymph node dissection	Urothelial carcinoma (35%)	Sarcomatoid spindle cell carcinoma (40%), conventional urothelial carcinoma (25%)	pT3a,N0	DOD at 11 mo
6	47/F	Dome, posterior, and lateral walls	Anterior exenteration, TAHBSO, and pelvic lymph node dissection	Urothelial carcinoma (25%), squamous differentiation (50%)	Urothelial carcinoma (10%), squamous differentiation (15%)	pT4a,N2	NA*
7	87/M	NA	Adjuvant chemotherapy, cystoprostatectomy, and pelvic lymph node dissection	Urothelial carcinoma (60%)	Squamous differentiation (40%)	pT4a,N1	DOD at 6 mo

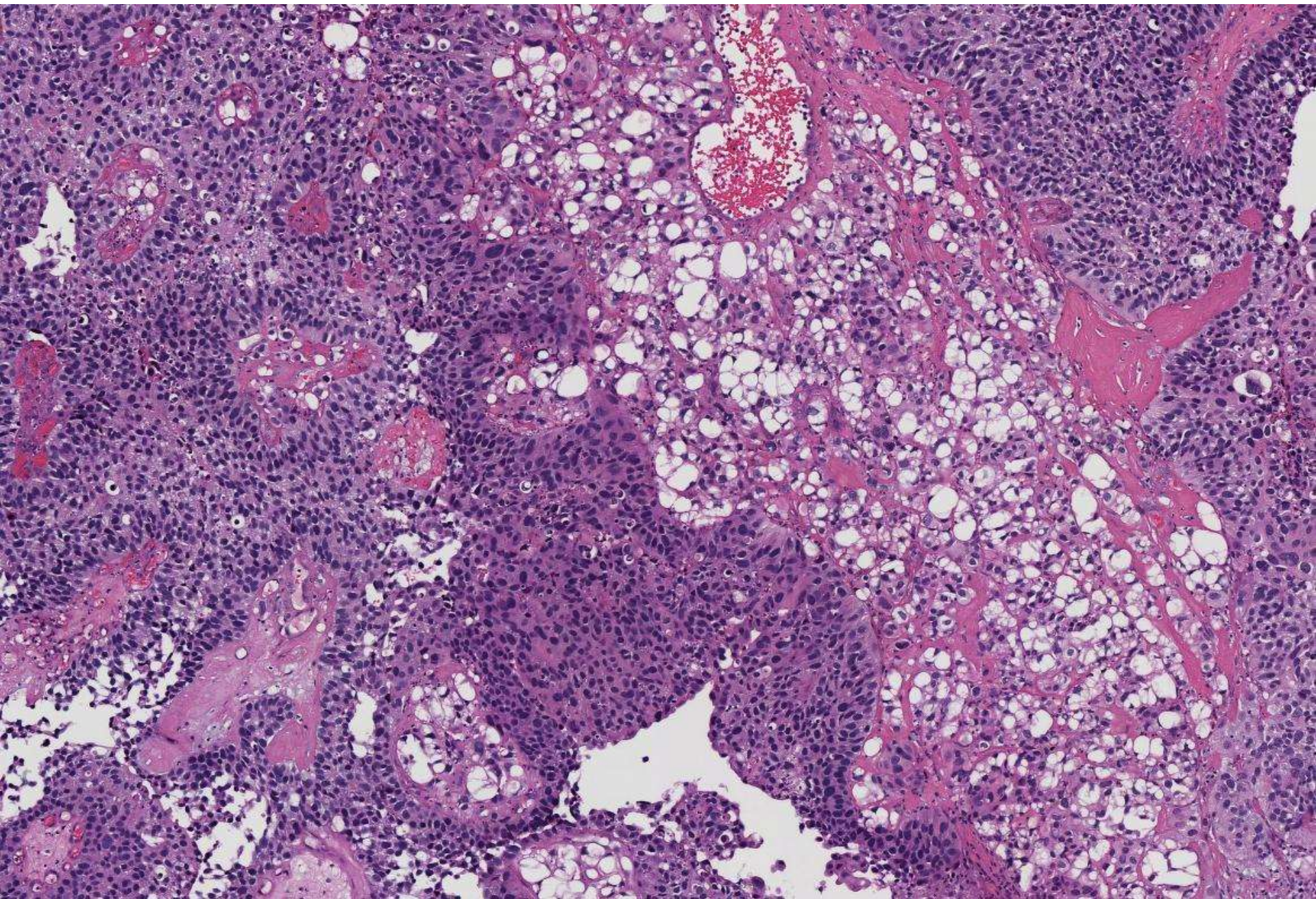
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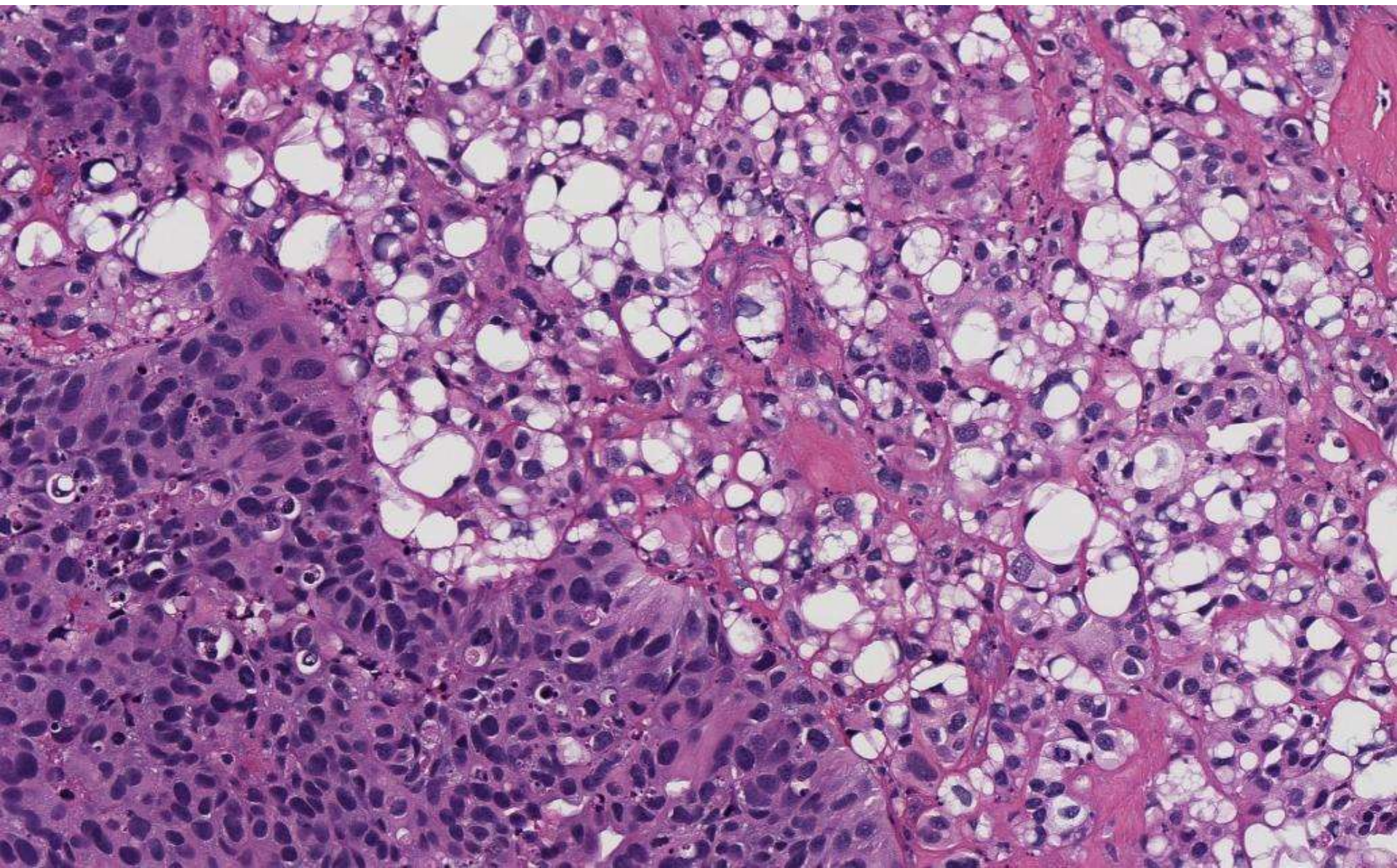
Ankur Sangoi; El Camino Hospital

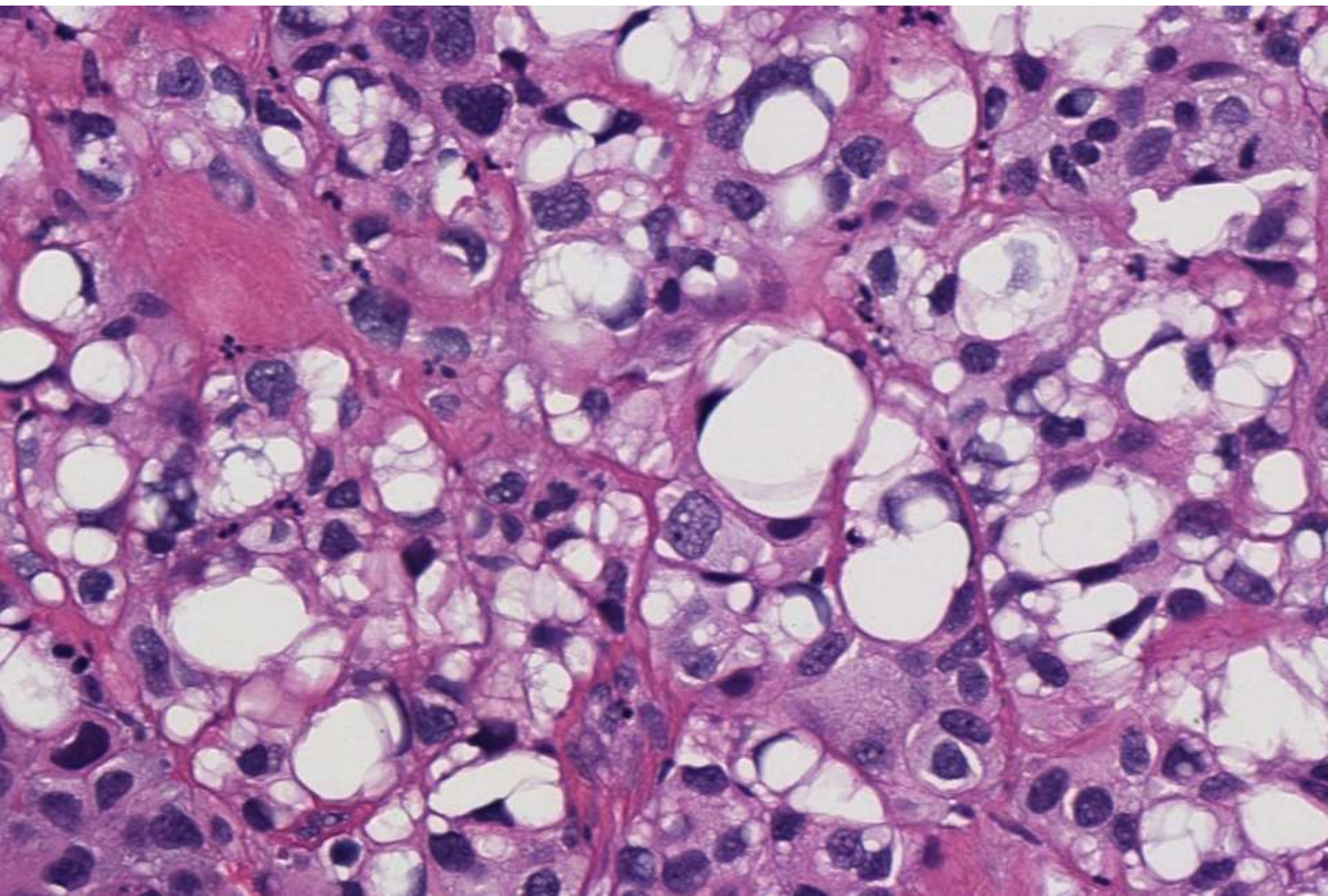
70-year-old female with bladder
tumor resection.

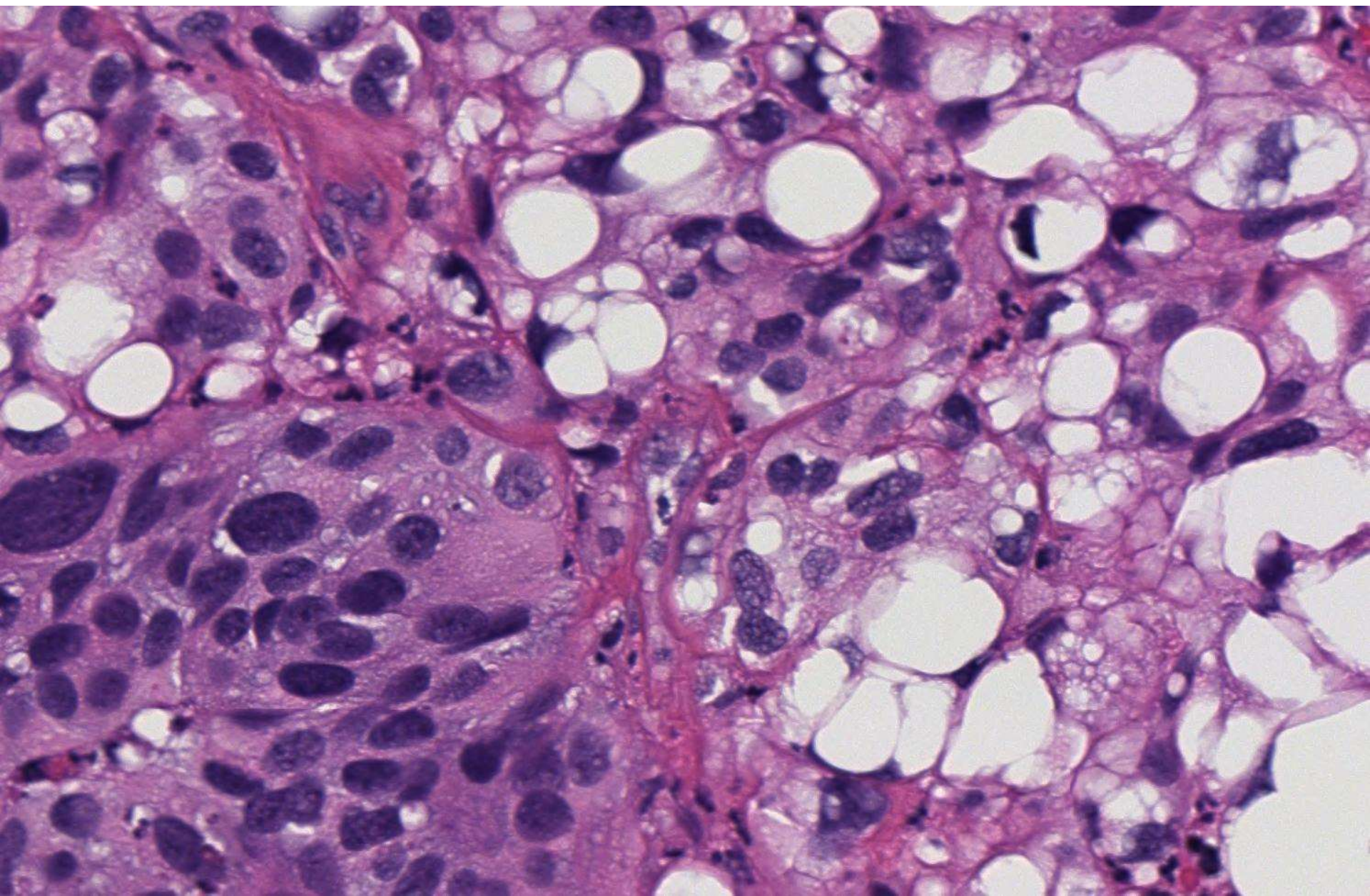












DIAGNOSIS?



DDx

- **Liposarcoma**
 - Keratin negative; MDM2/CDK4/S100+
- **Sarcomatoid carcinoma (carcinosarcoma)**
 - Unequivocal spindle cell morphology
 - Often decreased/negative keratin expression
- **Signet-ring adenocarcinoma (primary or metastatic)**
 - Site-specific IHCs for metastatic disease
 - Mucin+
 - Lack lipoblast-like cells

Urothelial Carcinoma of the Bladder, Lipid Cell Variant: Clinicopathologic Findings and LOH Analysis

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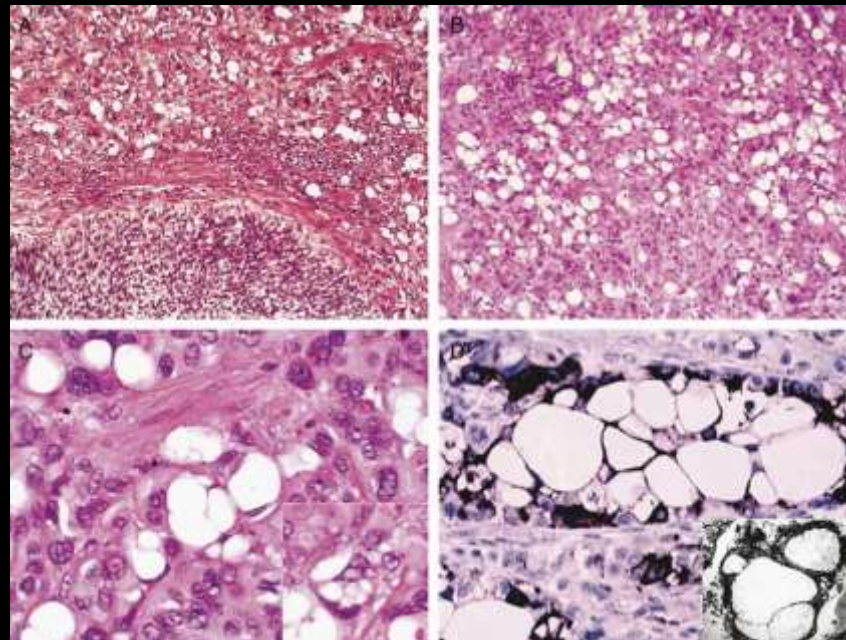


TABLE 1. Clinical and Pathological Features of the 27 Cases of Bladder Carcinoma With Lipid Cell Features

Characteristics	No. Cases (%) N = 27
Age in years (range) (mean; median)	42 to 94 (70;74)
Gender	
Male	25 (93)
Female	2 (7)
Initial symptoms	
Hematuria	27 (100)
Obstructive symptoms	2 (7)
Fever	1 (4)
Urinary retention	2 (7)
Anemia	2 (7)
Treatment	
Cystoprostatectomy	13 (48)
Cystoprostatectomy and chemotherapy	3 (11)
Cystoprostatectomy and radiochemotherapy	1 (4)
TURB	7 (26)
TURB and BCG	3 (11)
TNM stage	
Ta	1 (4)
T1	2 (7)
T2, at least	7 (26)
T3a	4 (15)
T3b	8 (30)
T4a	5 (18)
Lymph node status	
Nx	14 (52)
N1	12 (44)
N0	1 (4)
Lipid cell component	
10%	6 (22)
20%	10 (38)
30%	5 (18)
40%	2 (7)
50%	4 (15)
Associated urothelial carcinoma	
Conventional, high grade	22 (81)
Papillary, high grade	3 (11)
Other	
-Micropapillary	2 (7)
-Plasmacytoid	1 (4)
-Glandular differentiation	1 (4)
-Squamous differentiation	1 (4)
Follow-up (mo)	
DOD, range 16 to 58 (mean, 33)	16 (59)
AWD, range 8 to 25 (mean, 22)	8 (30)
DOC, range 6 to 15 (mean, 10)	3 (11)