Disclosures
July 10, 2017

Dr. Christian Kunder disclosed a financial relationship (spouse) with Gilead Sciences, Inc. (employee), which the activity planners have determined is not relevant to the case he will be presenting at the meeting.

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

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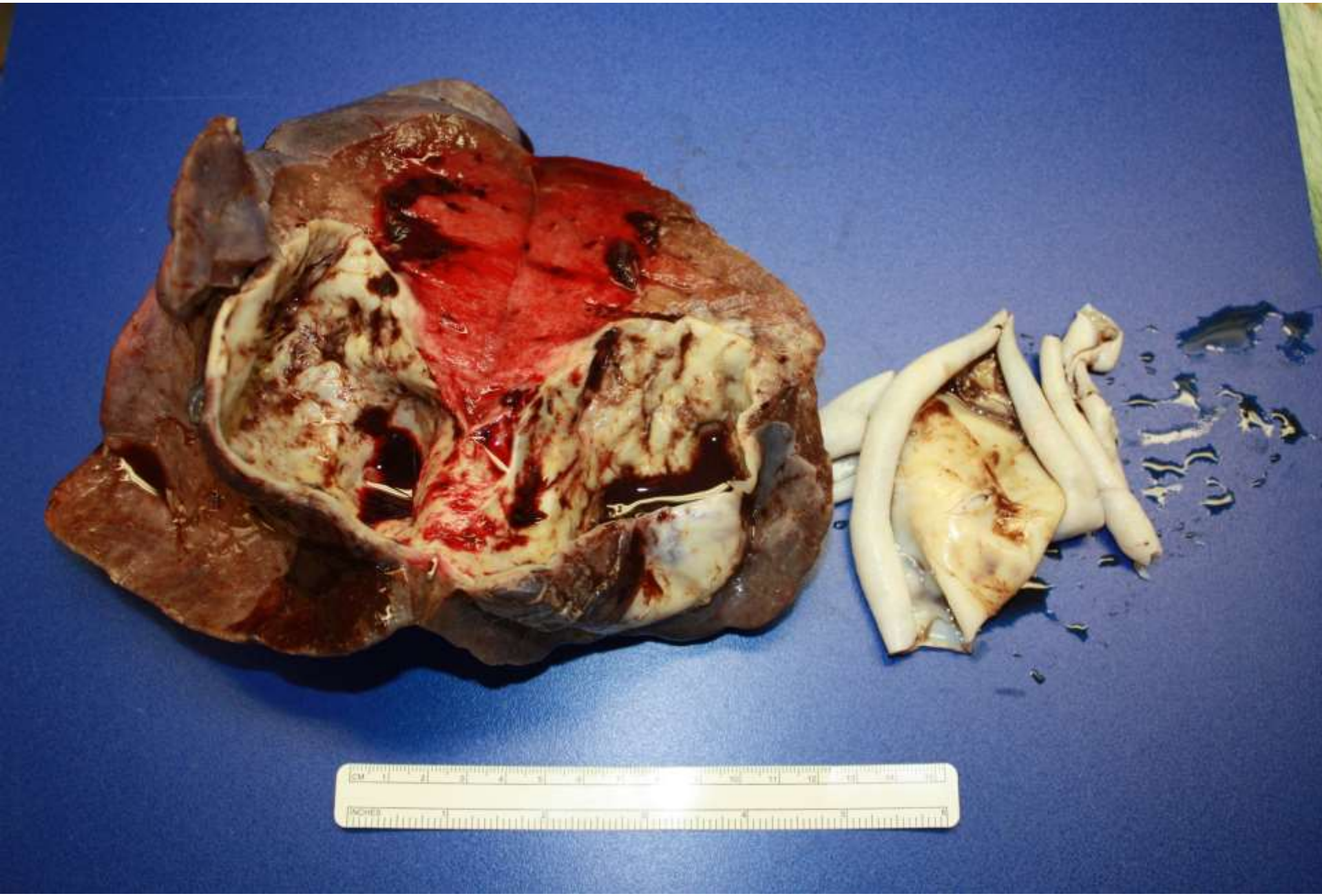
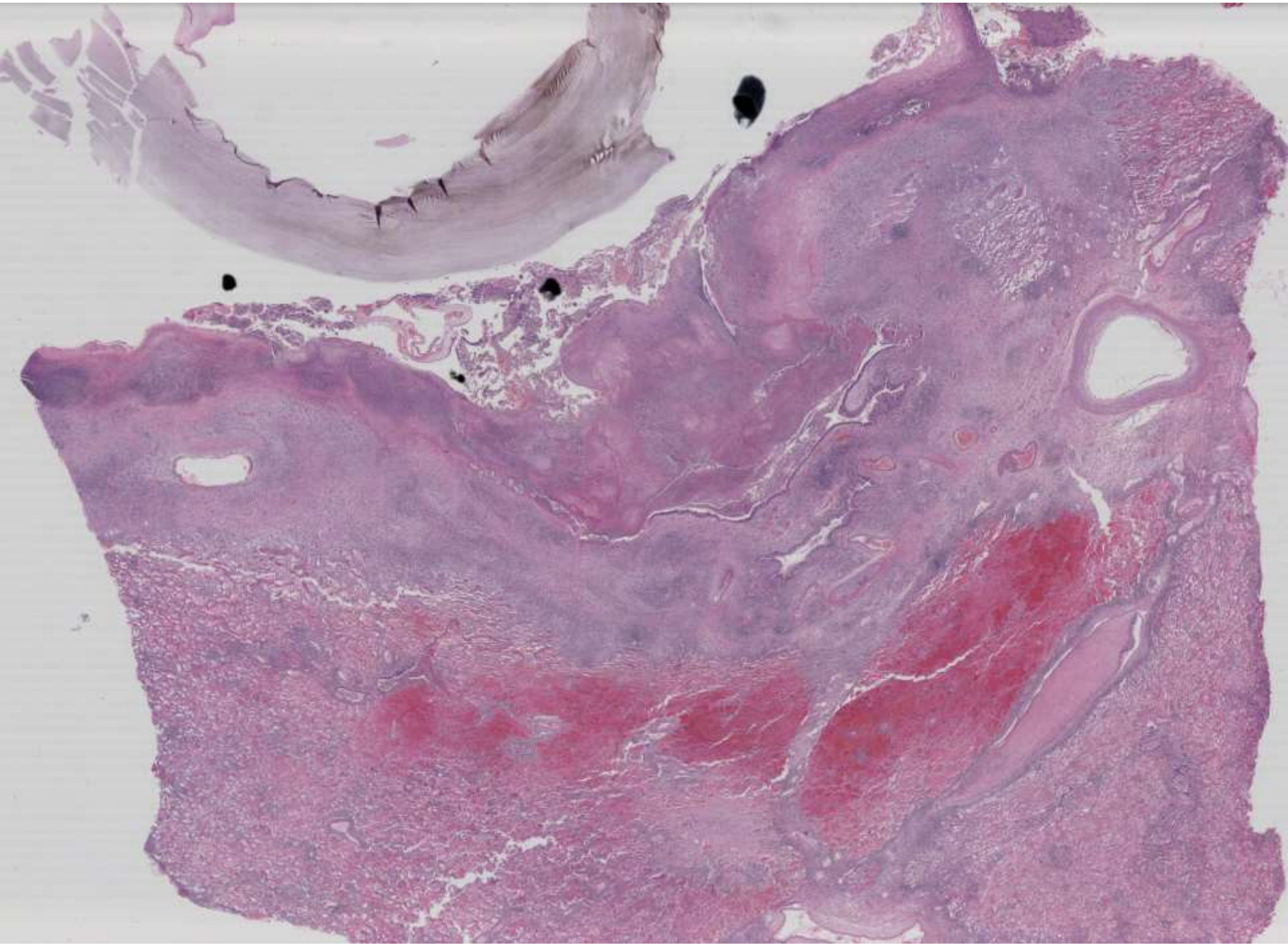
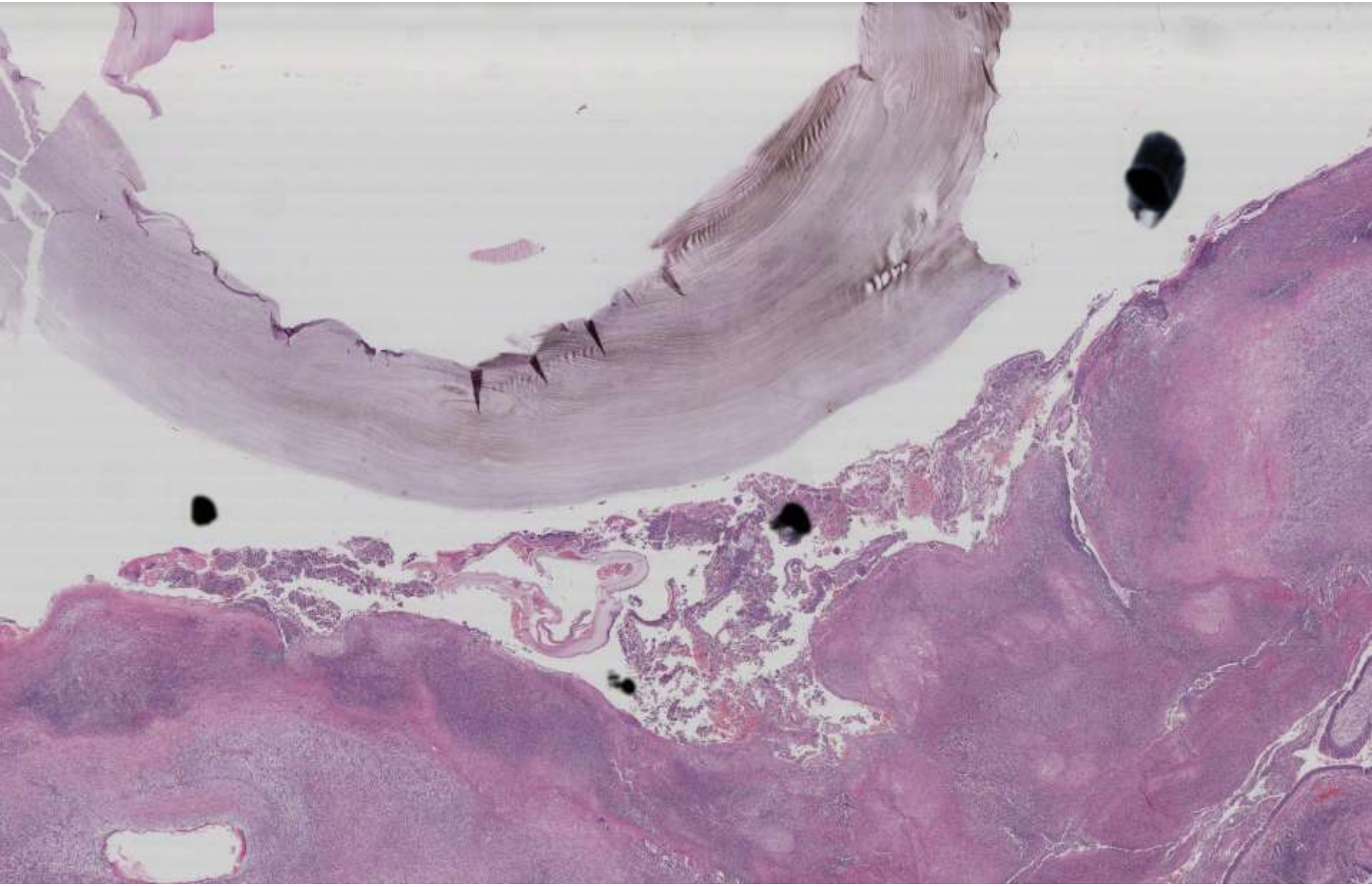
SB 6171
(scanned slide available)

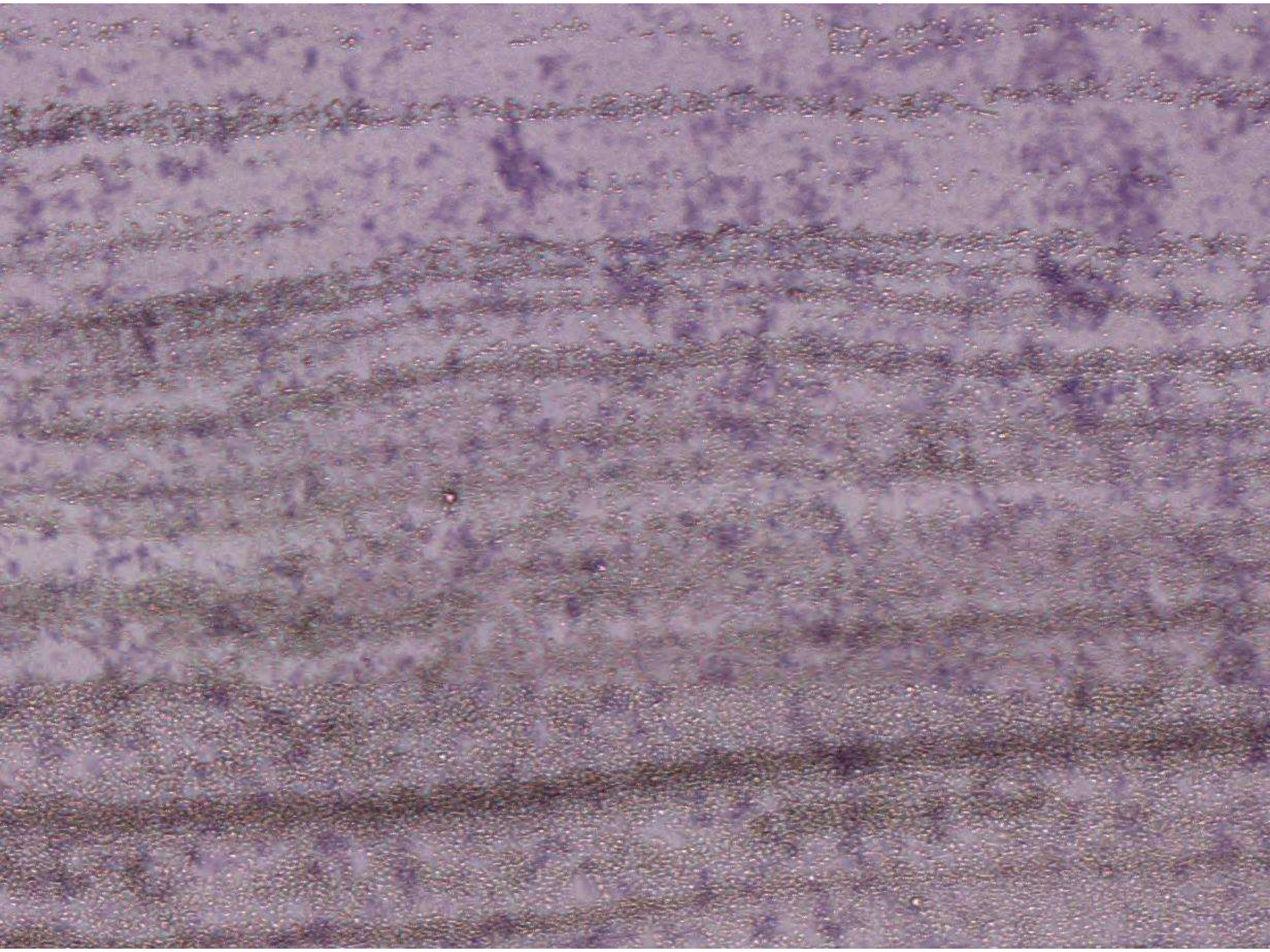
Charles Lombard; El Camino Hospital

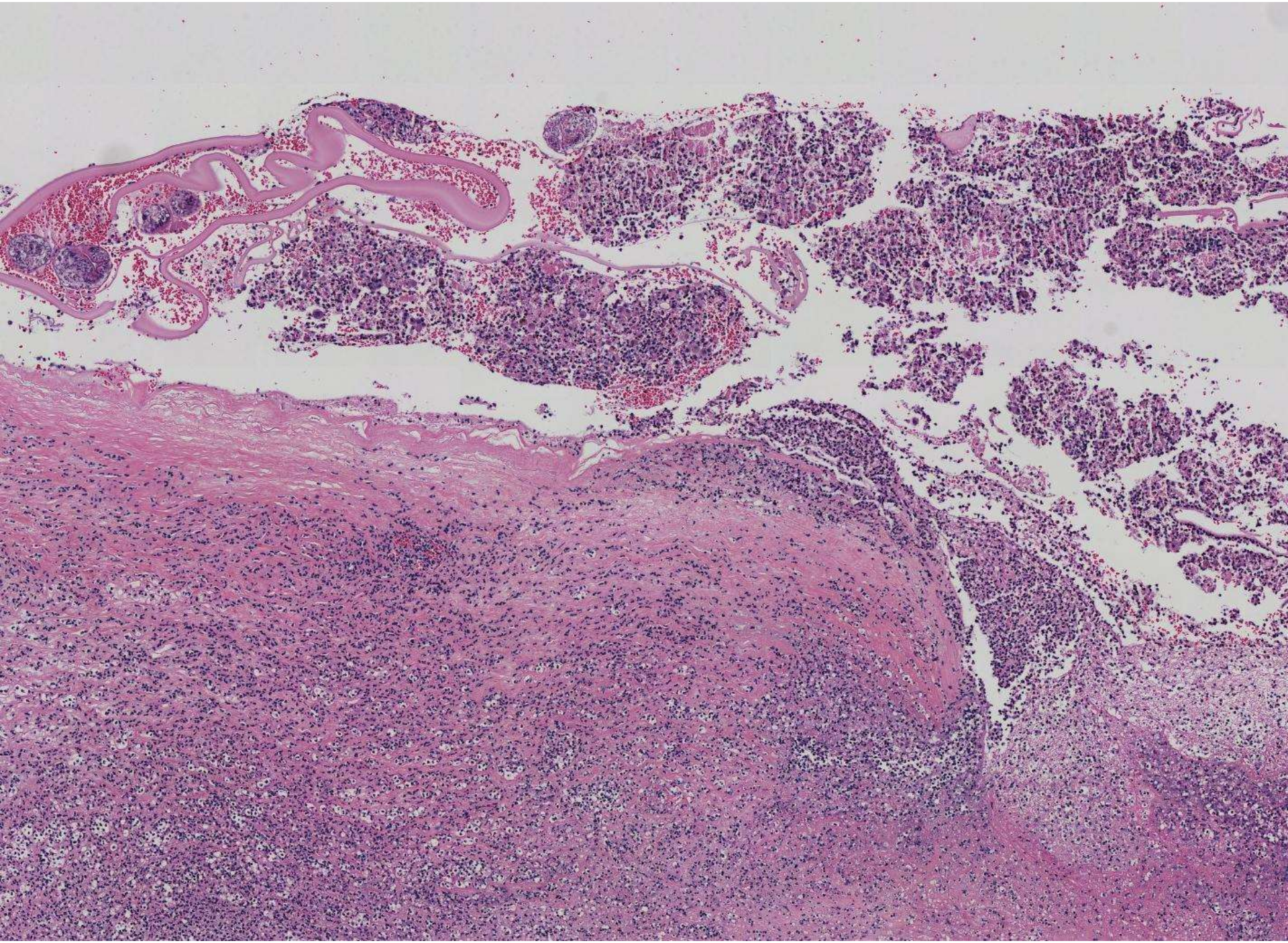
30-year-old woman with lung cyst, lobectomy performed.

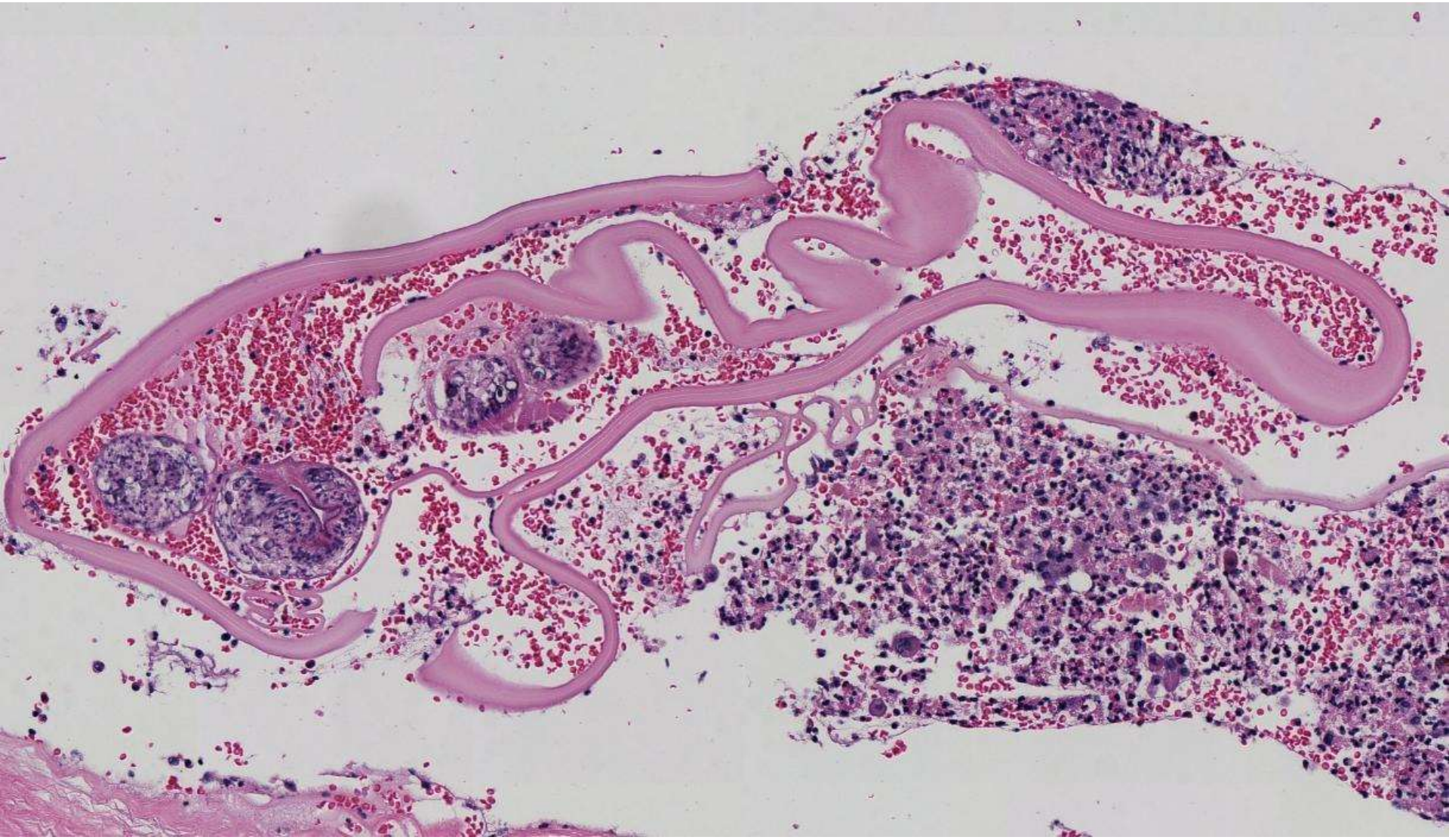


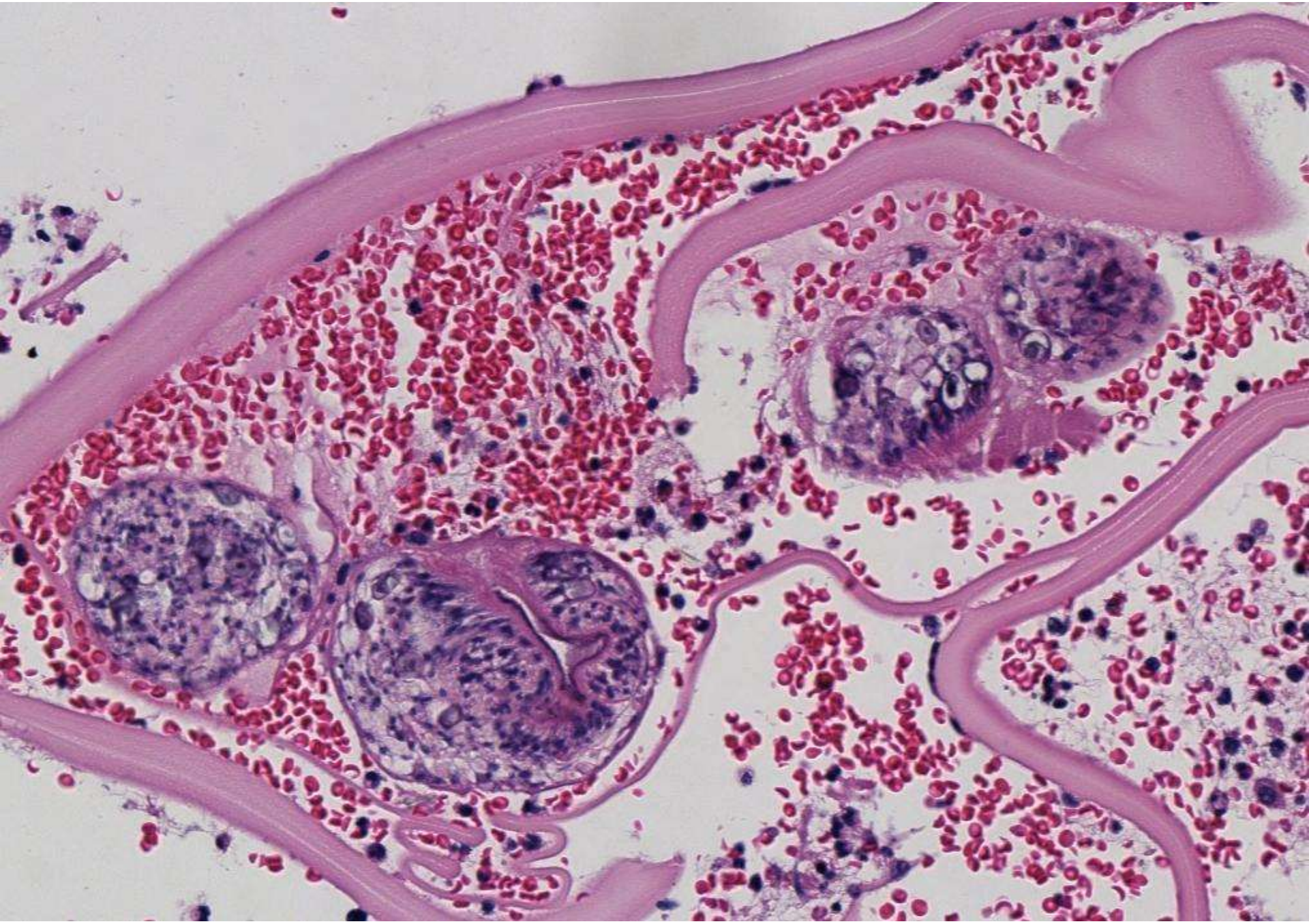


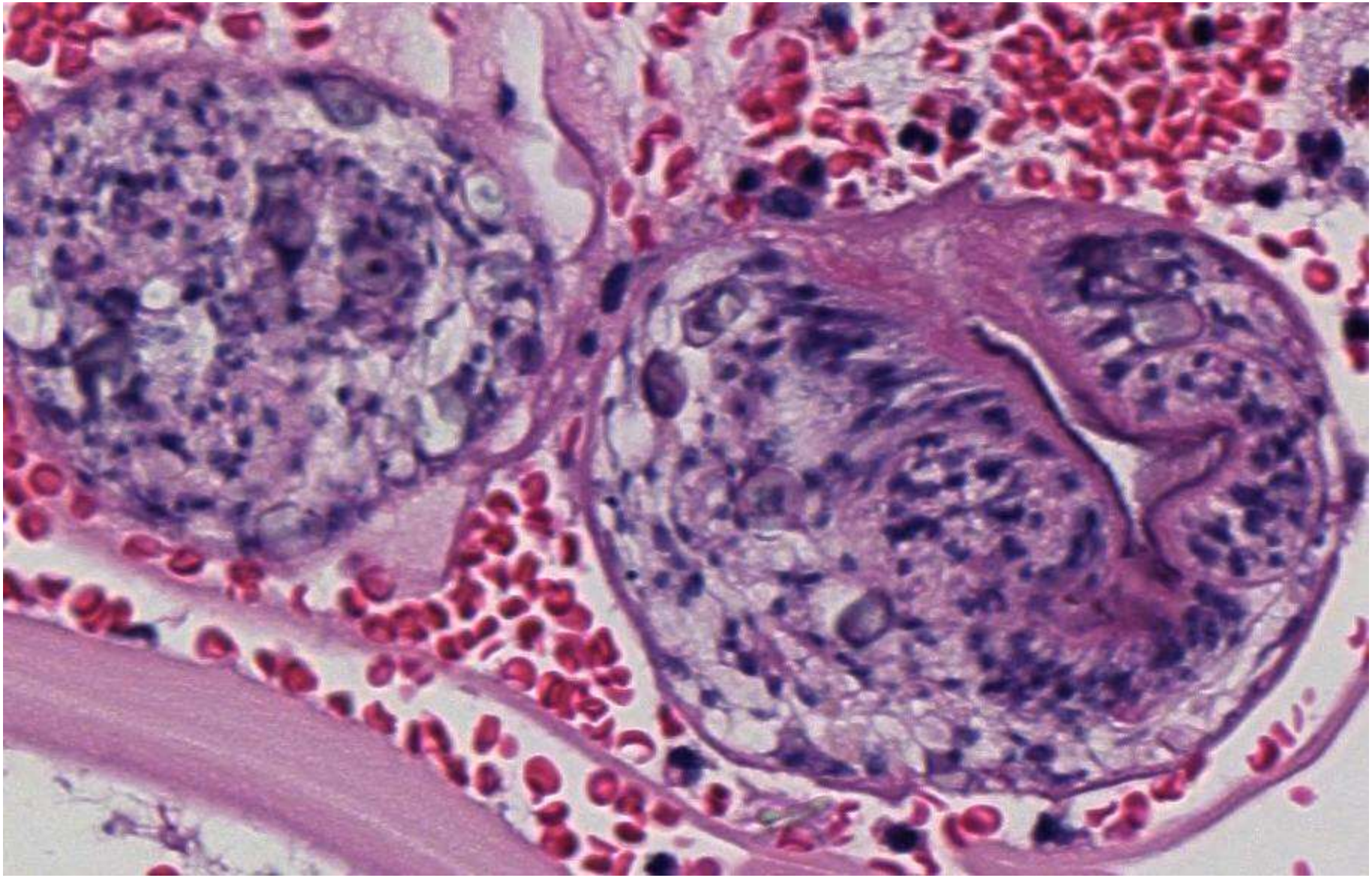


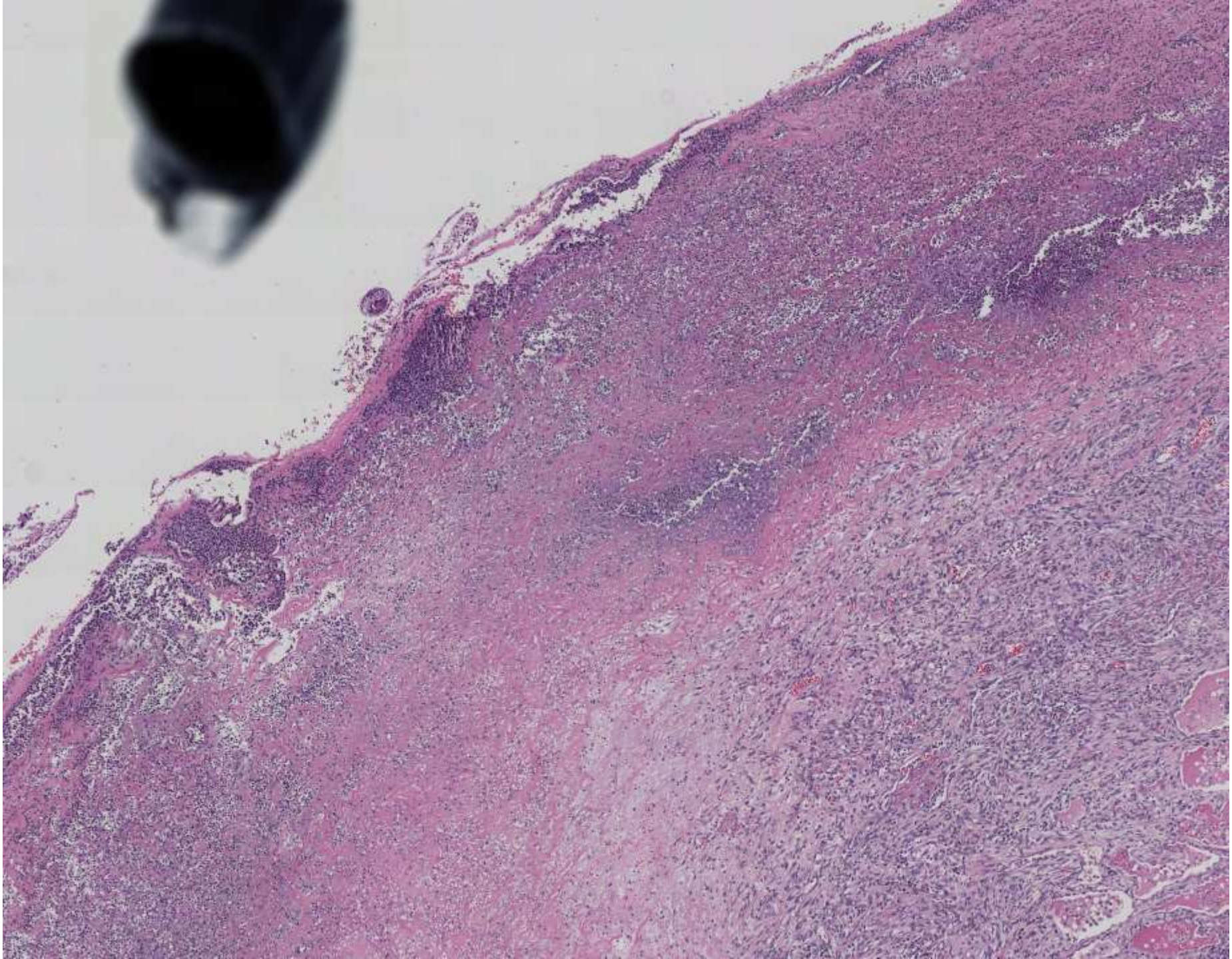


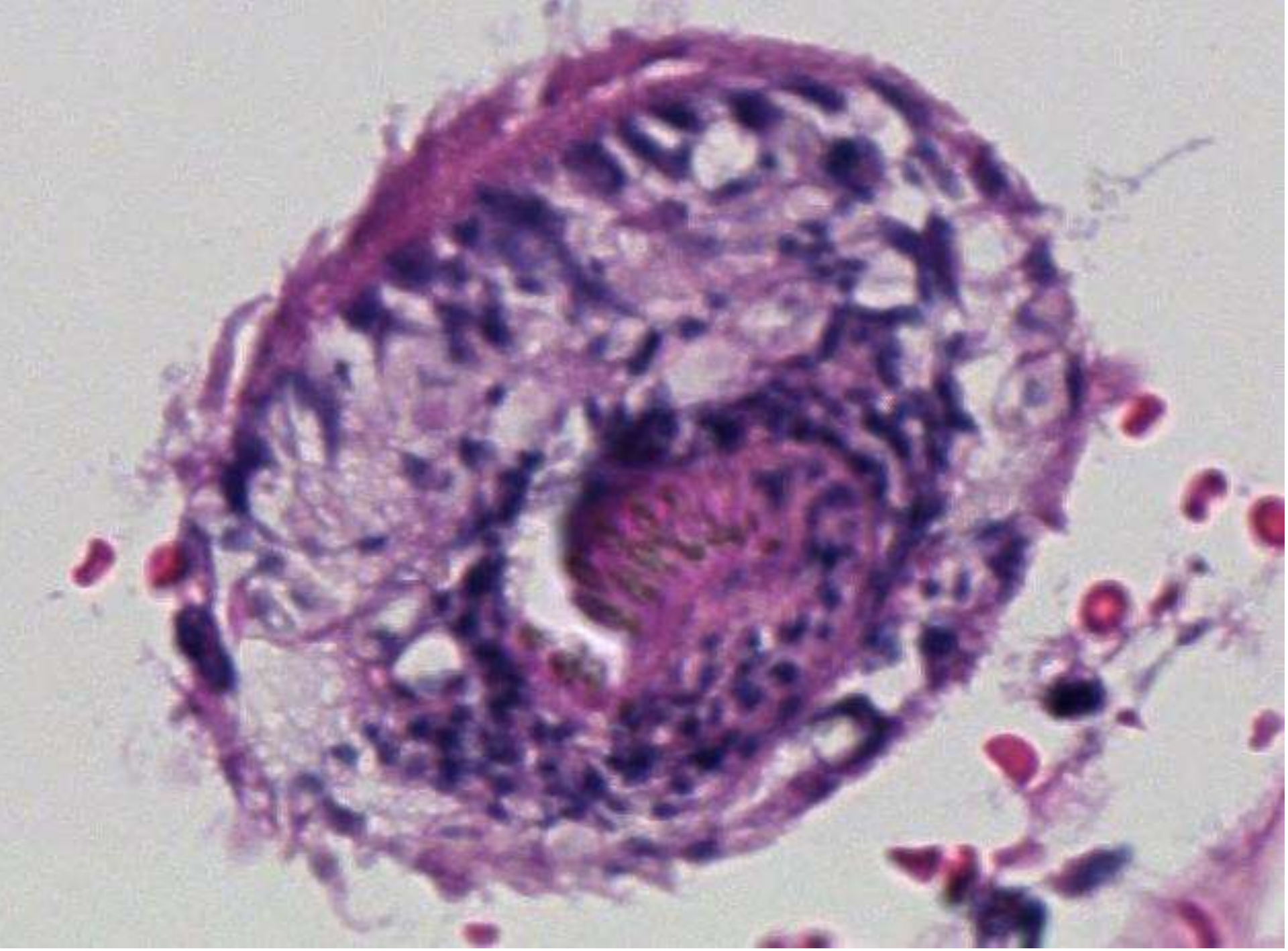








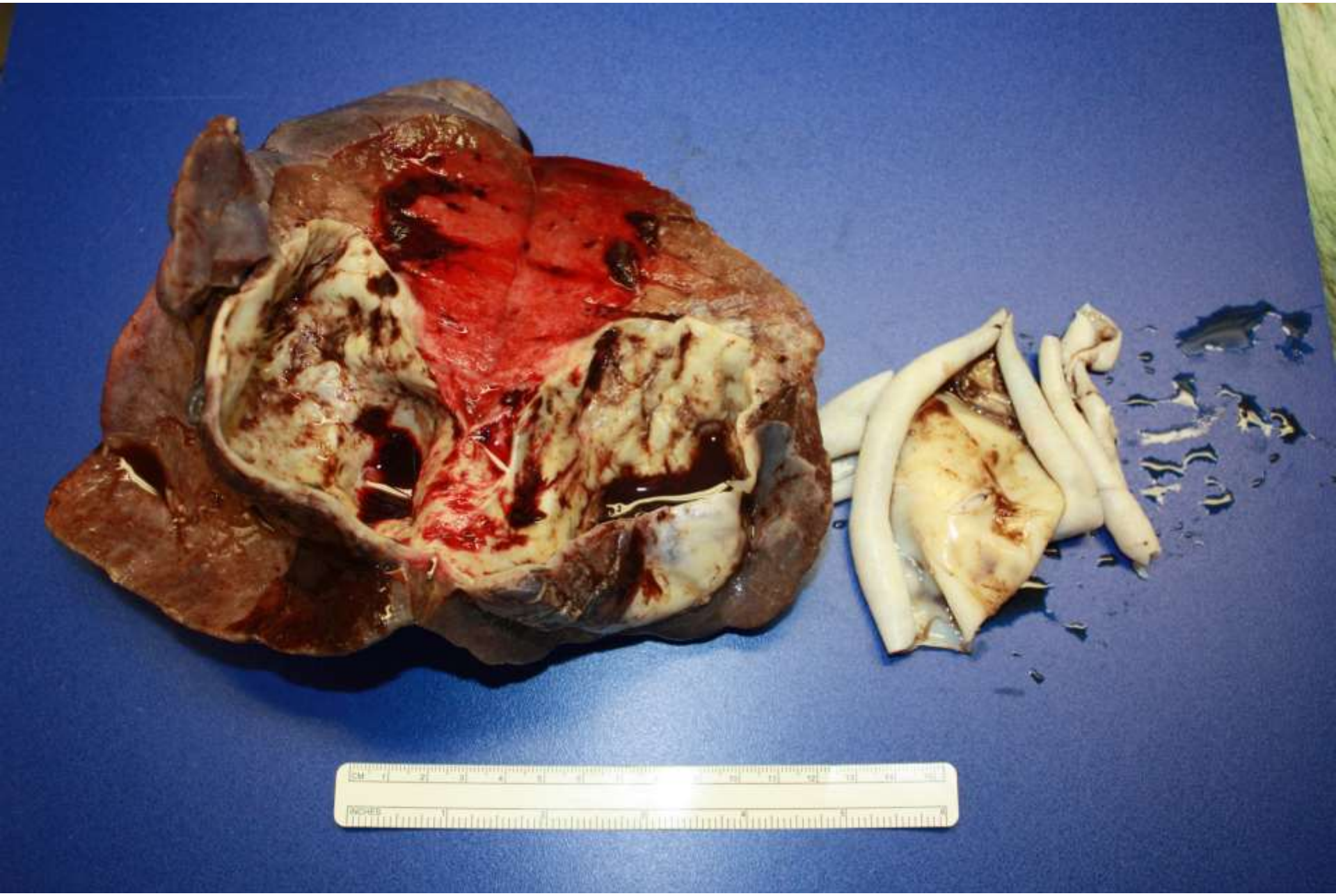


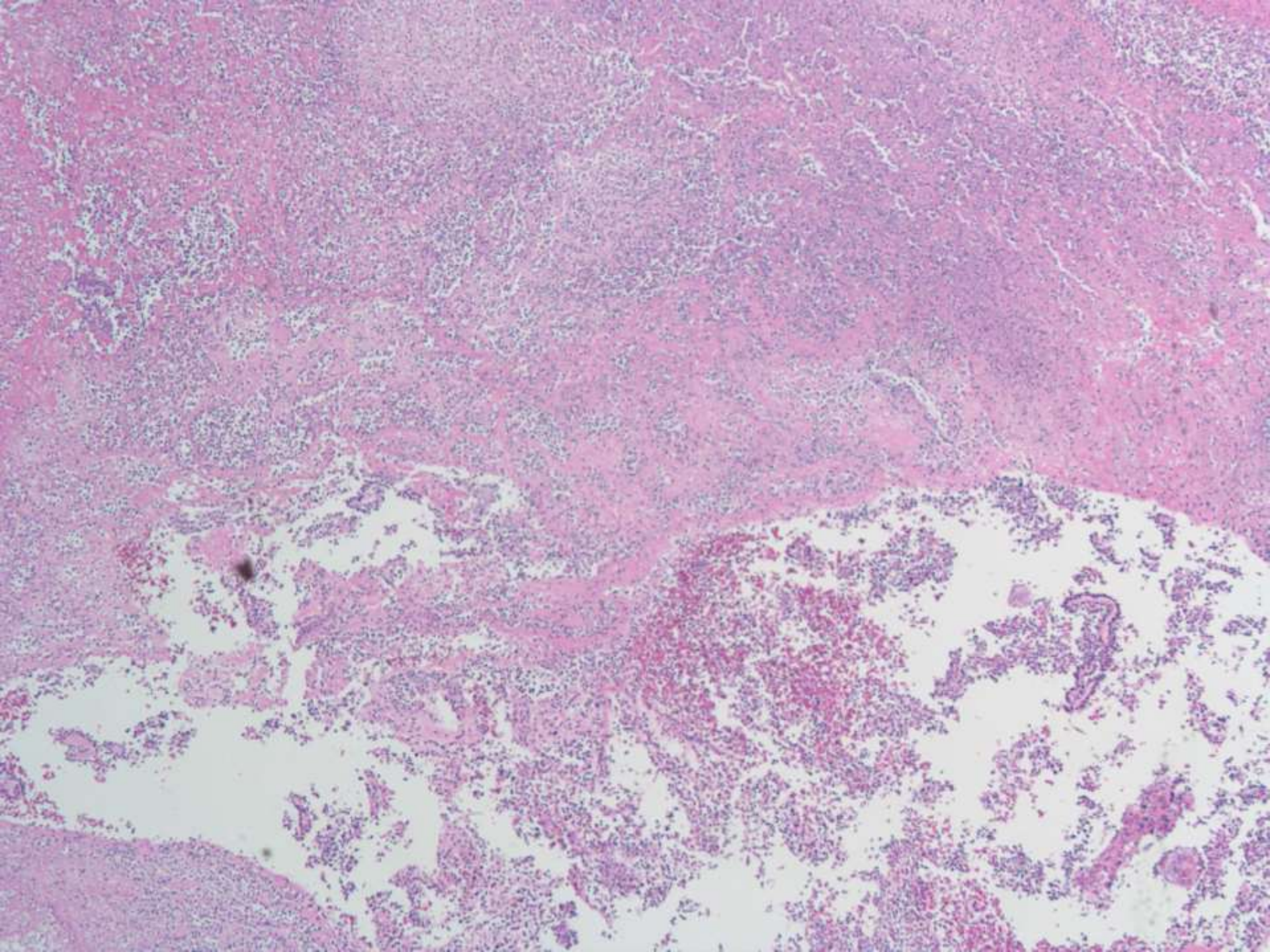


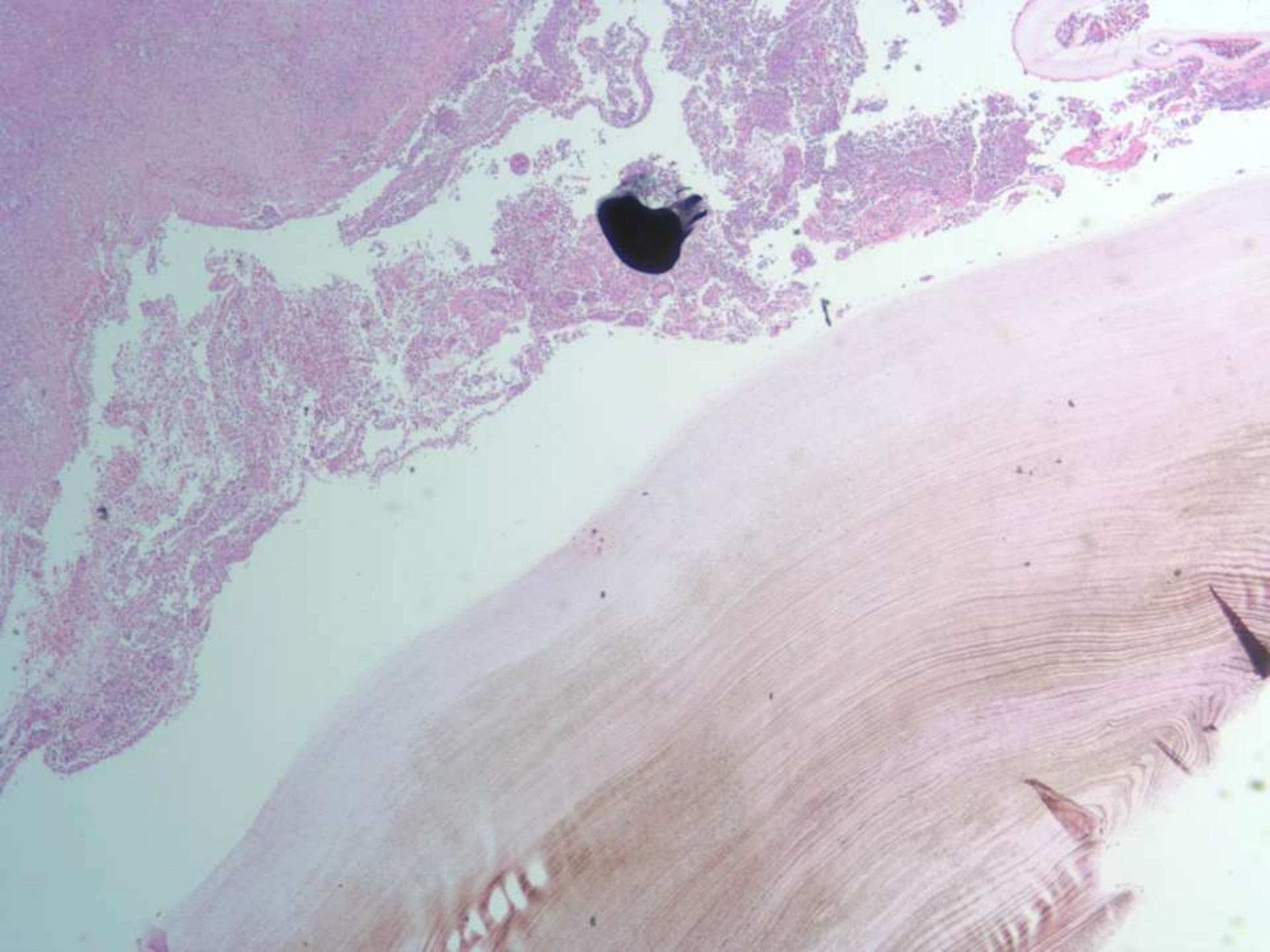
DIAGNOSIS?

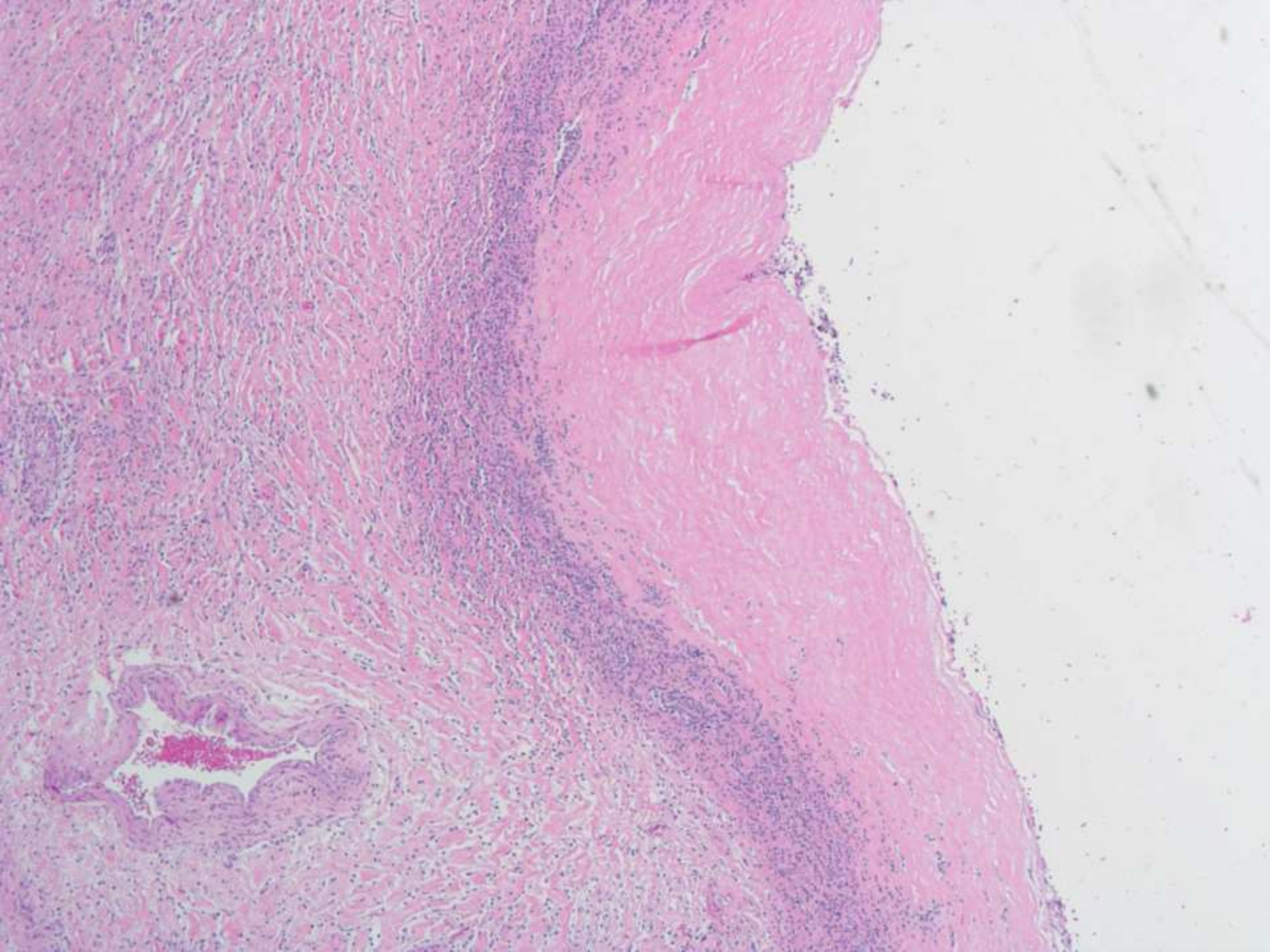
30 yo F

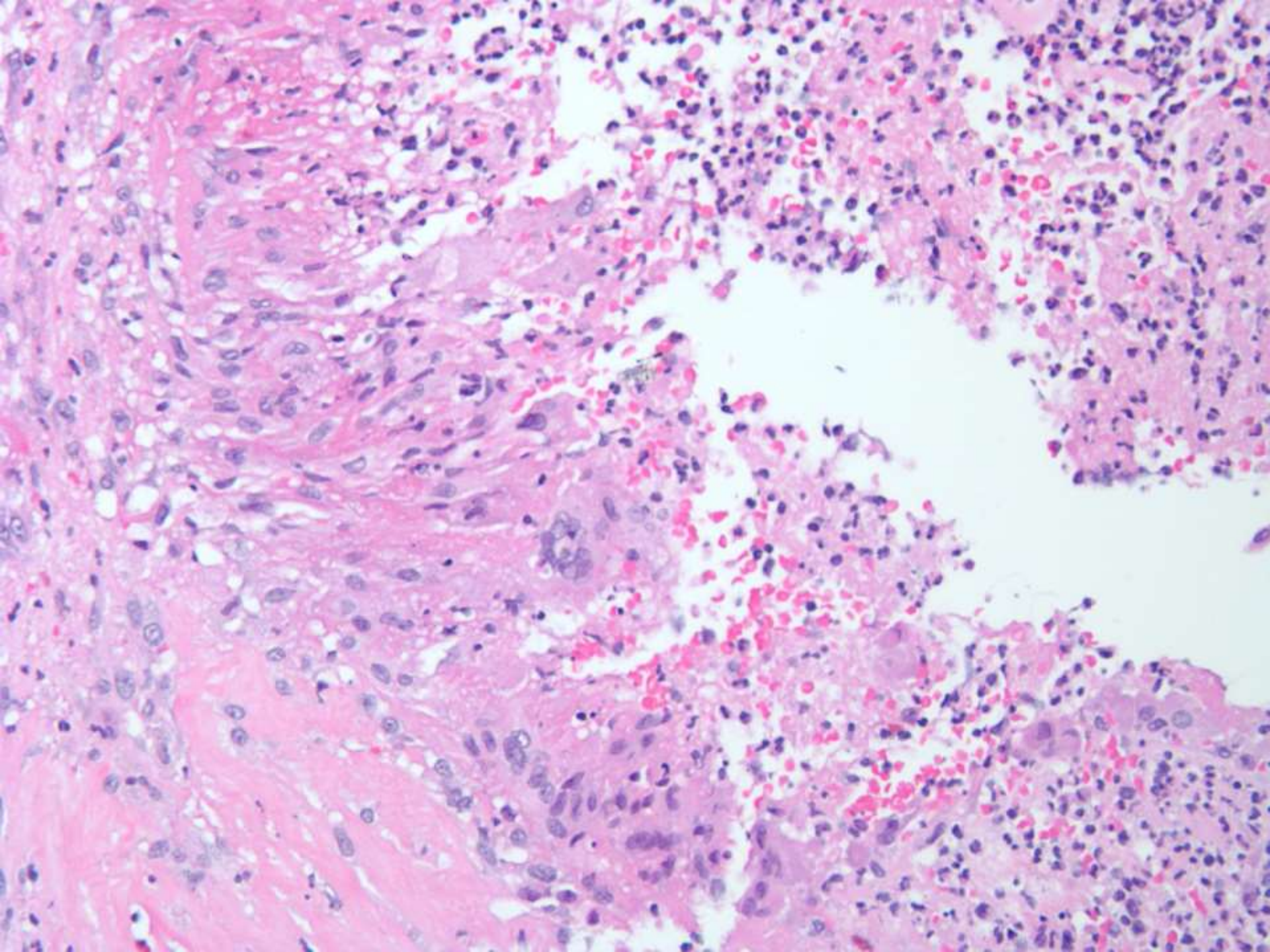
- Presents with dyspnea, hemoptysis
- History of cysts in lung and liver

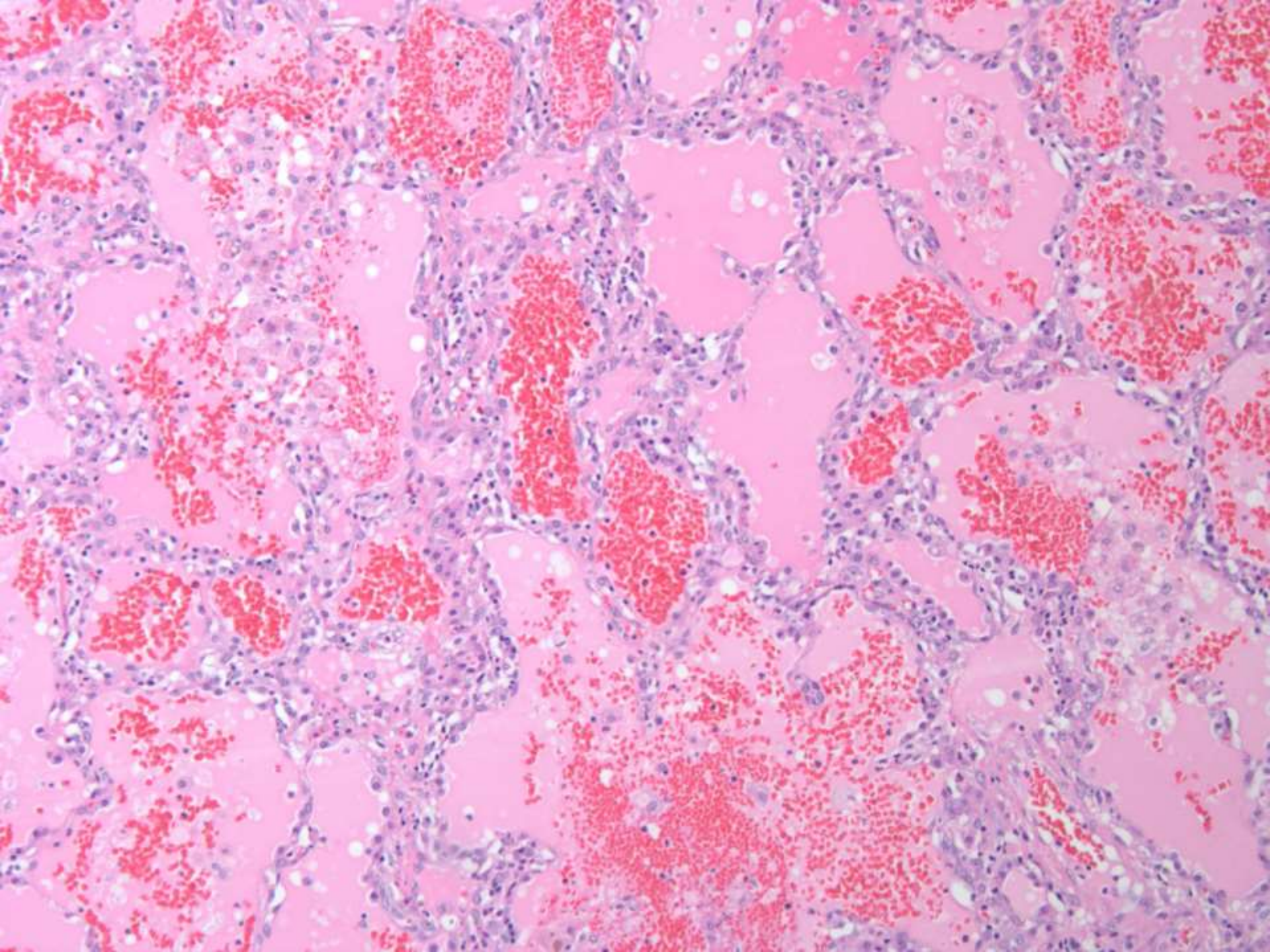

CM
INCHES

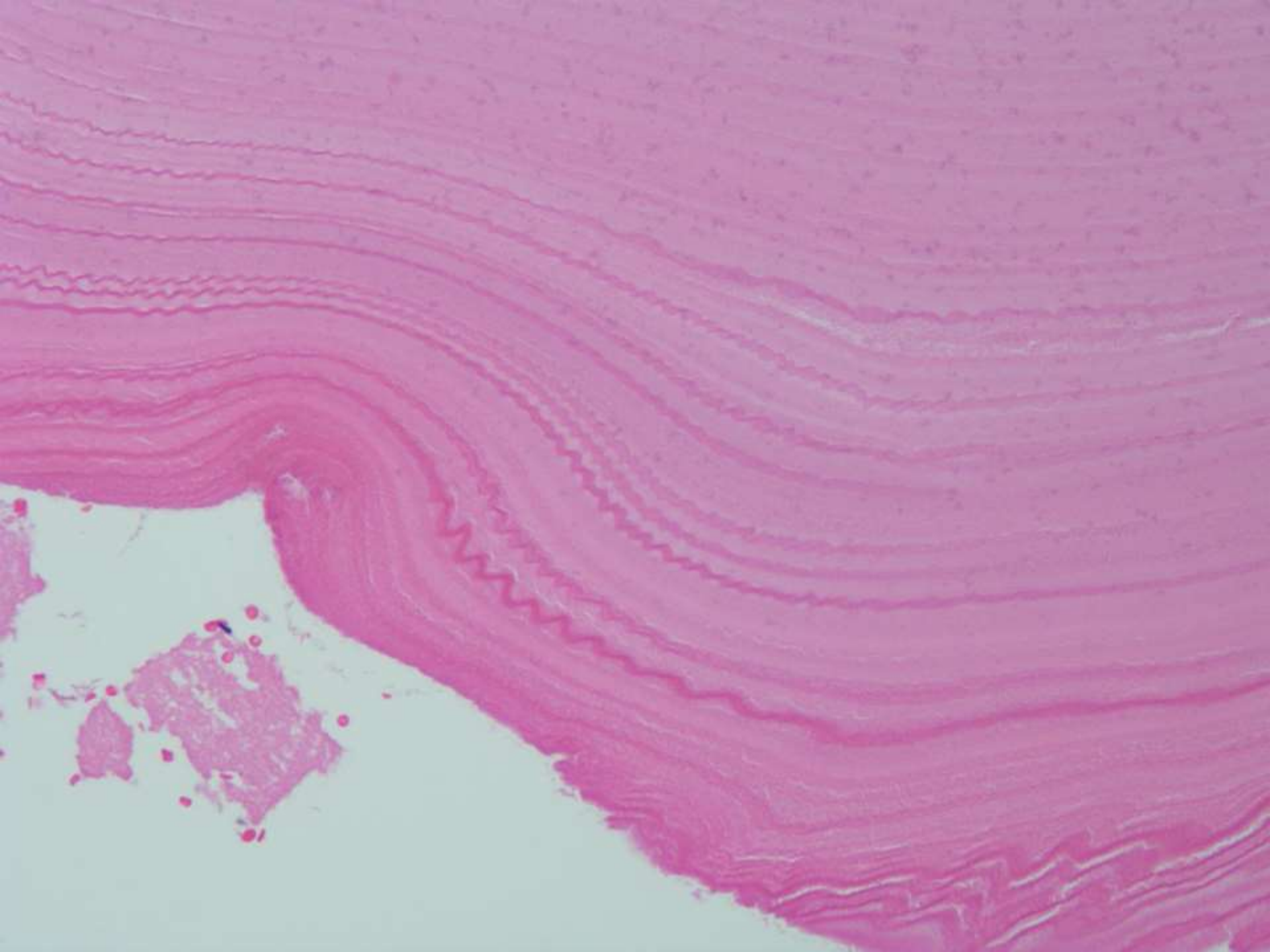


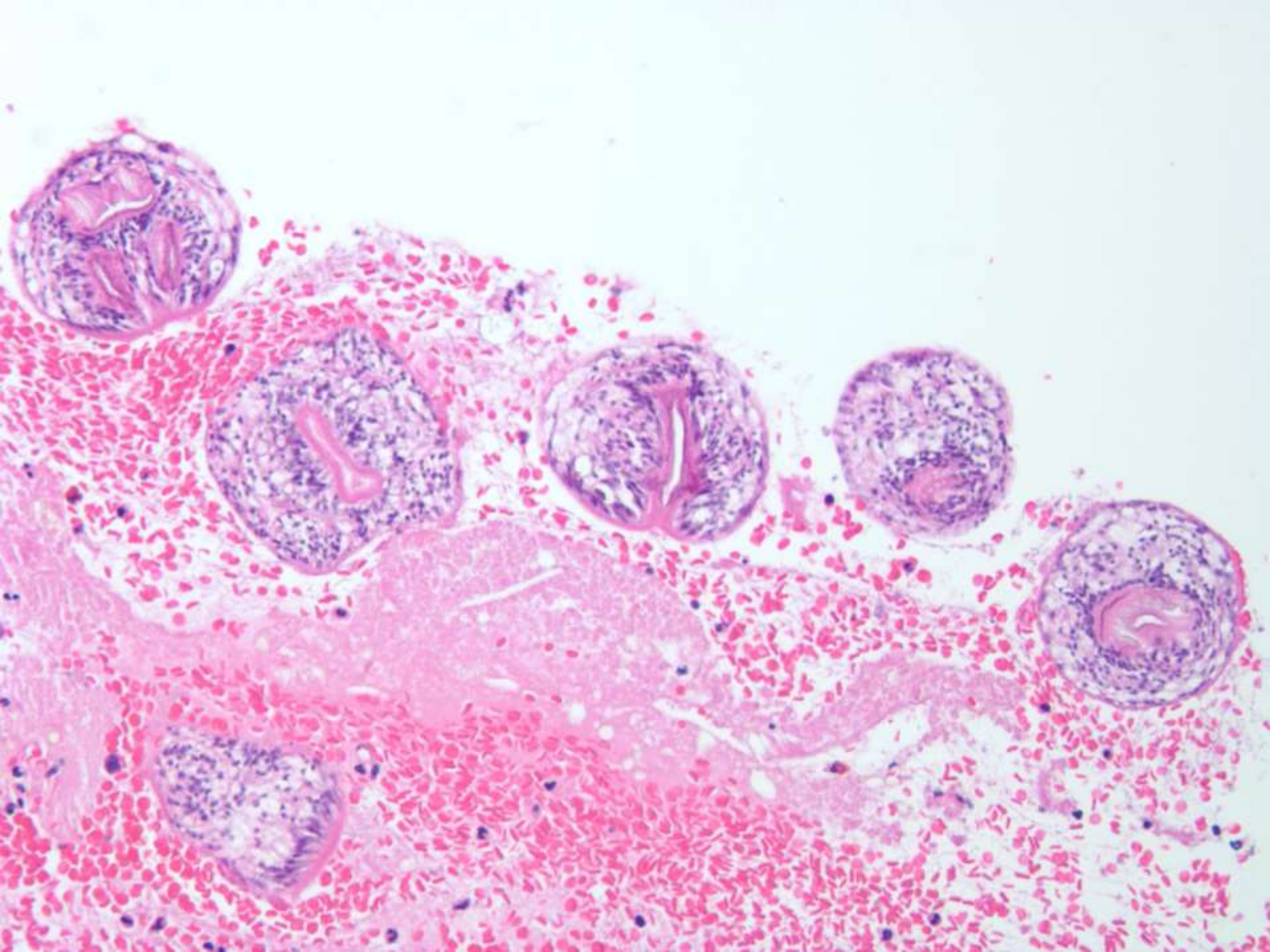


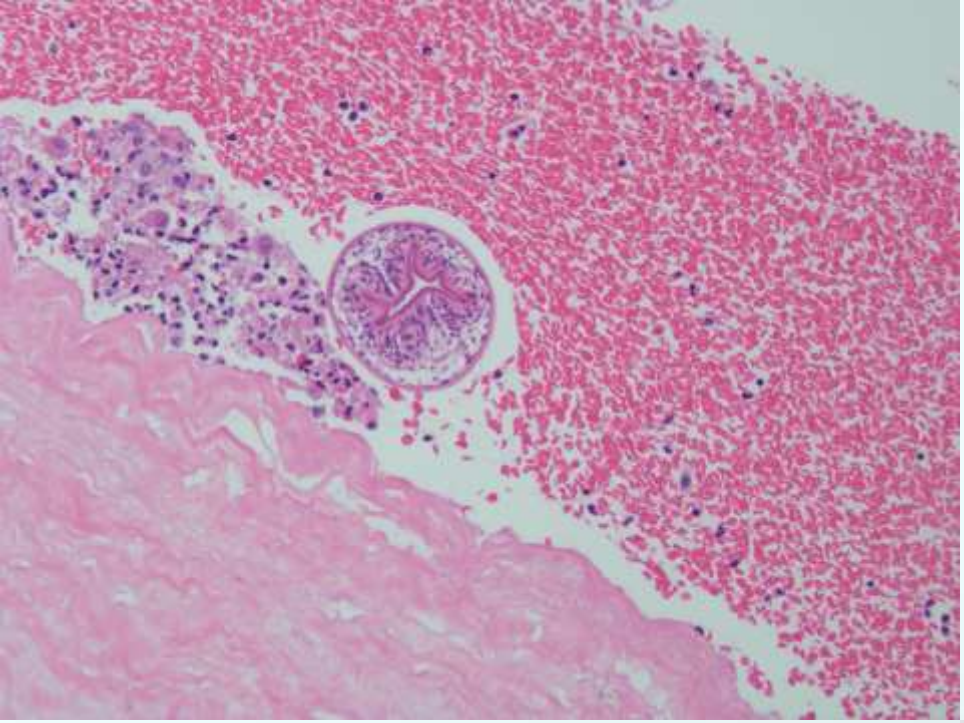
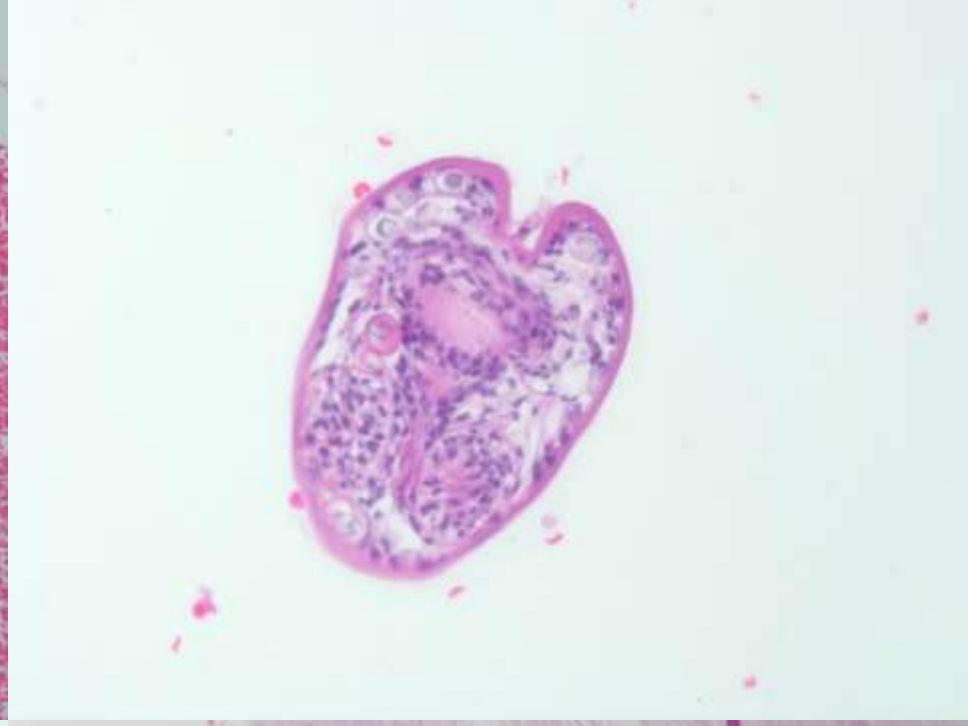
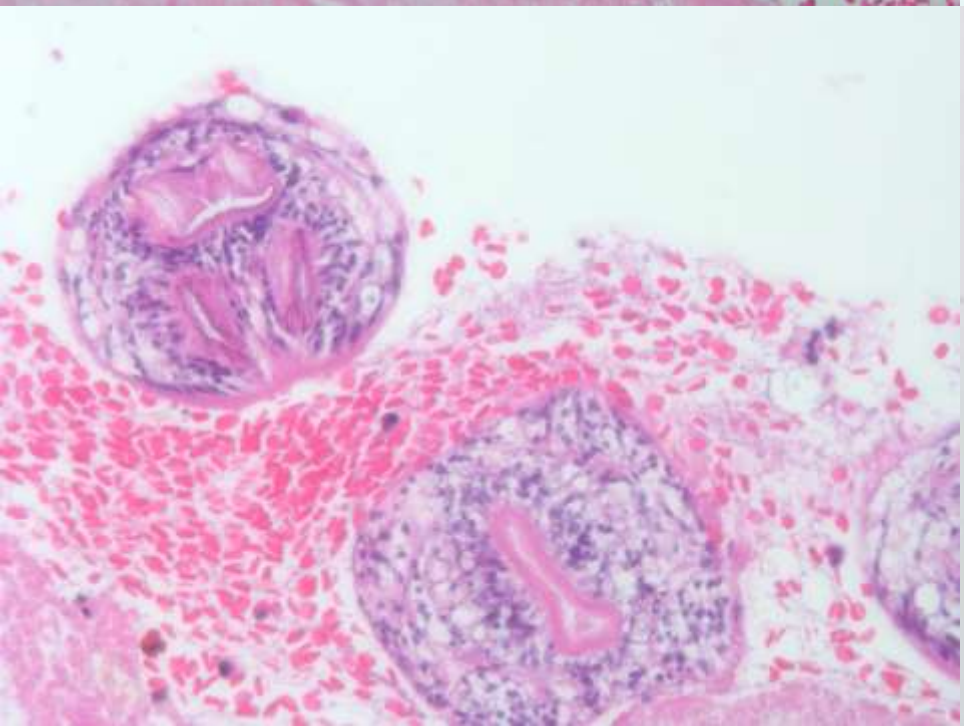
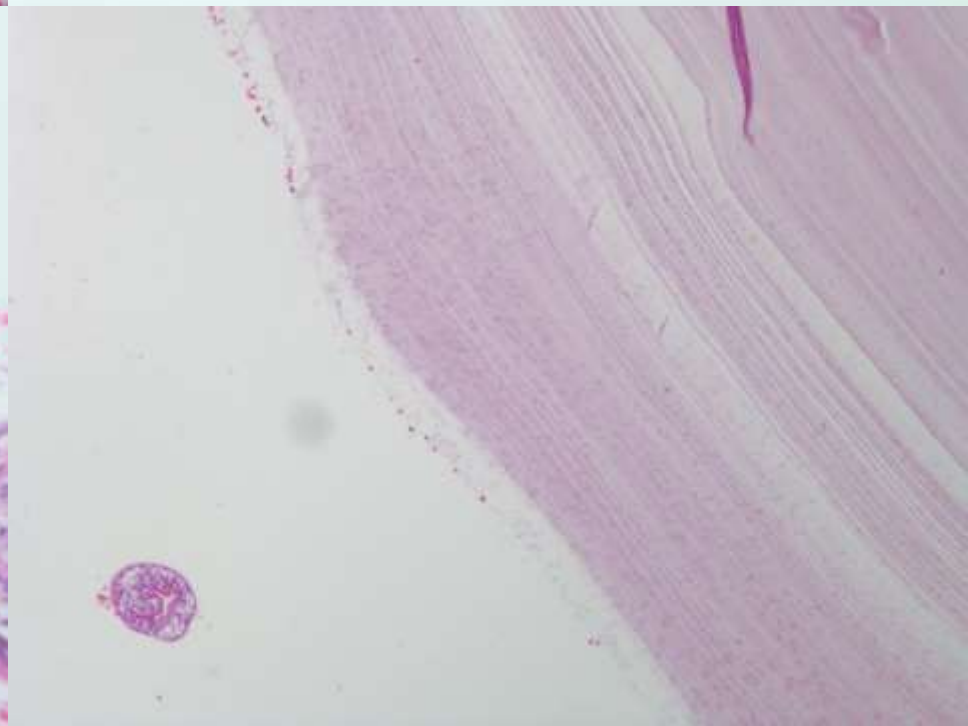


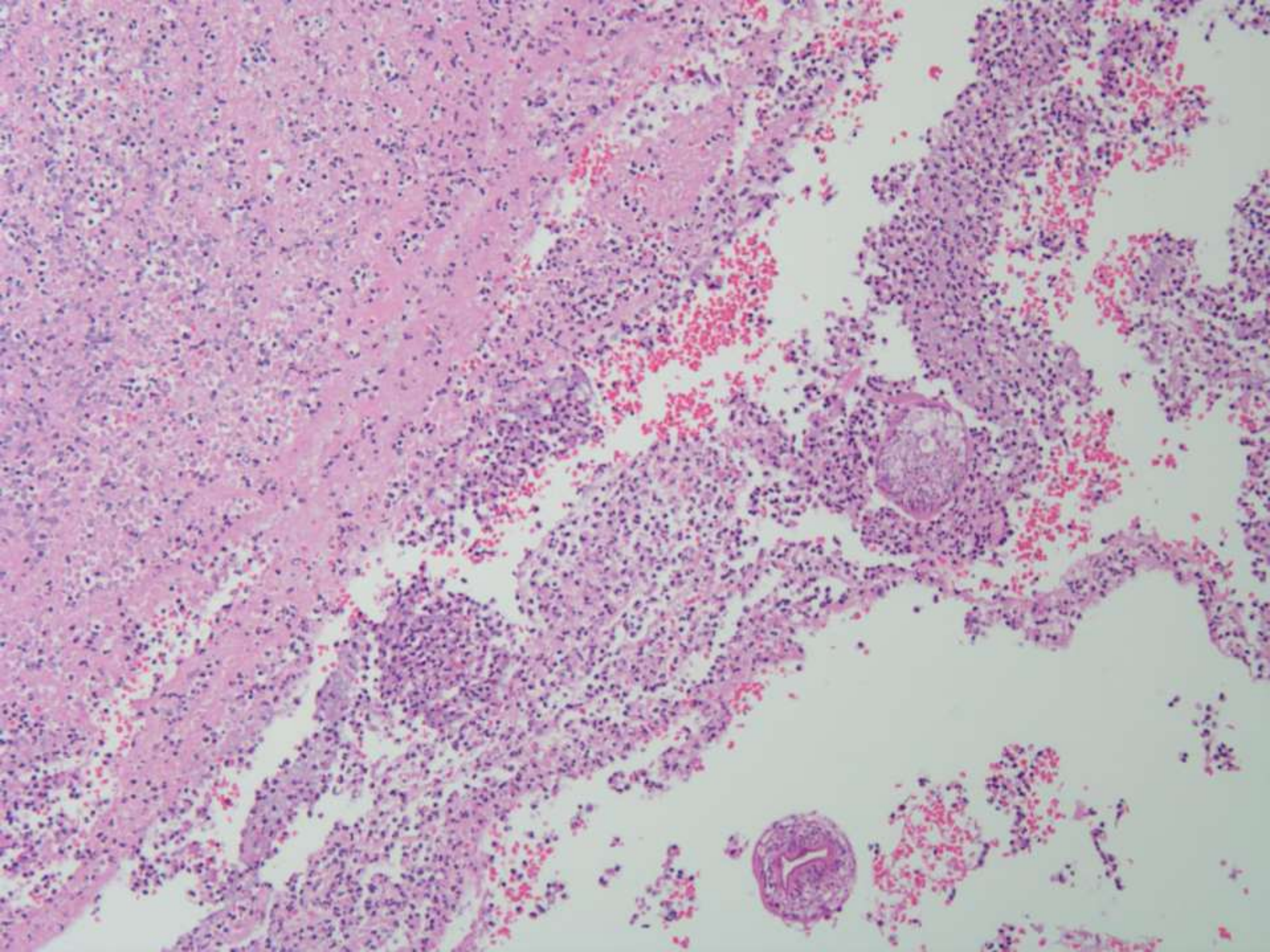


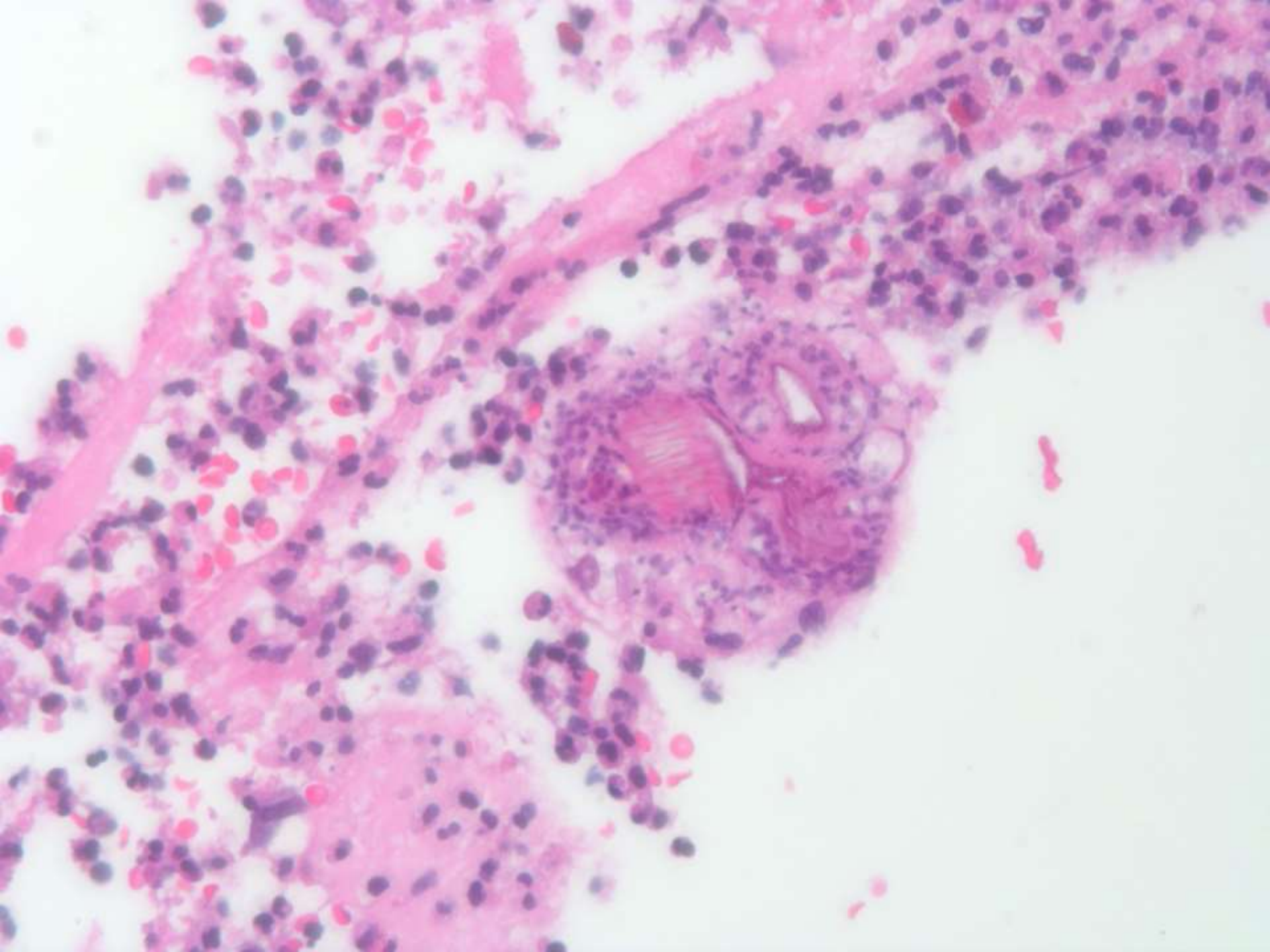


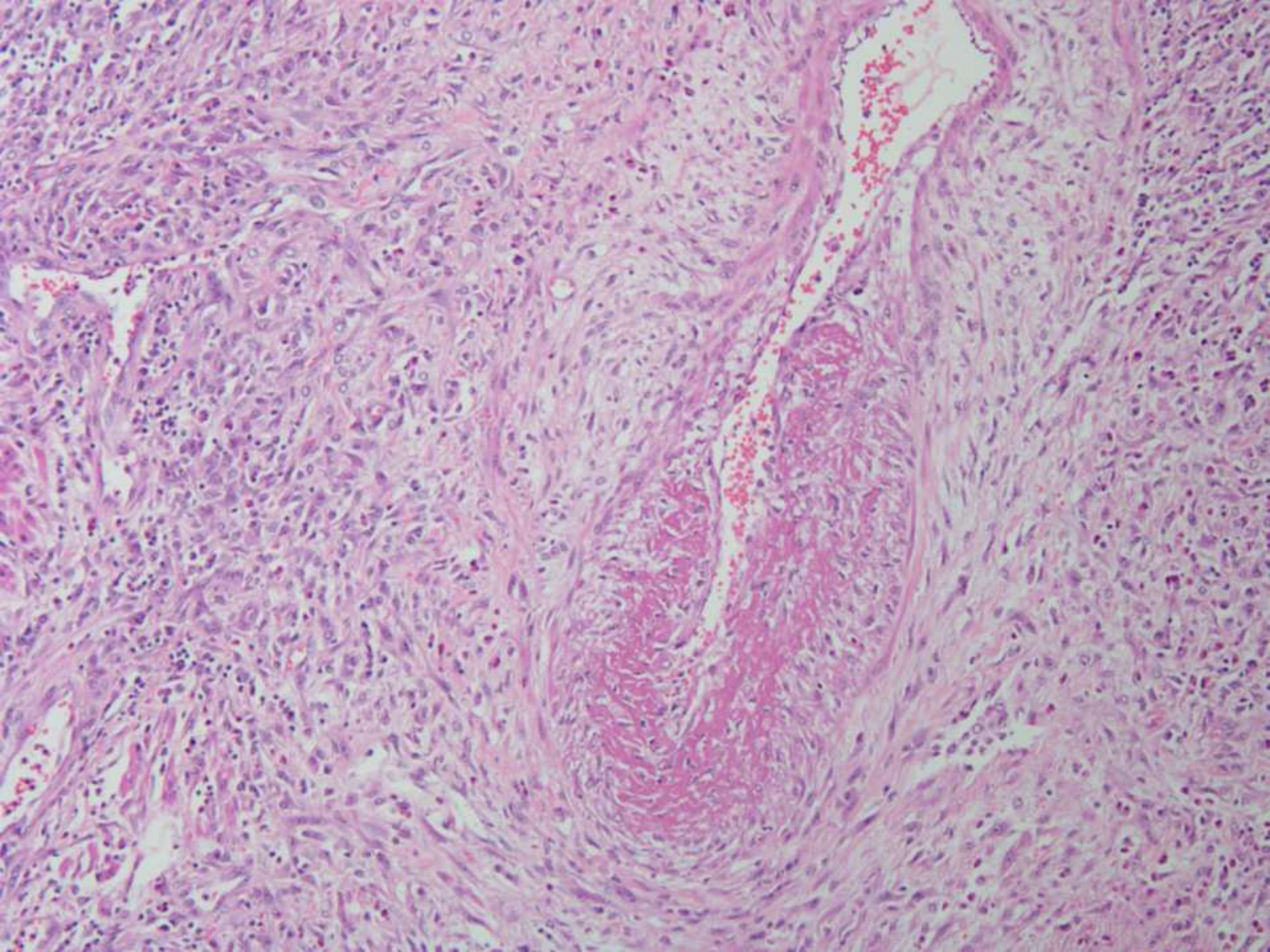










Diagnosis ???

Cystic Echinococcus (E. Granulosus)

History

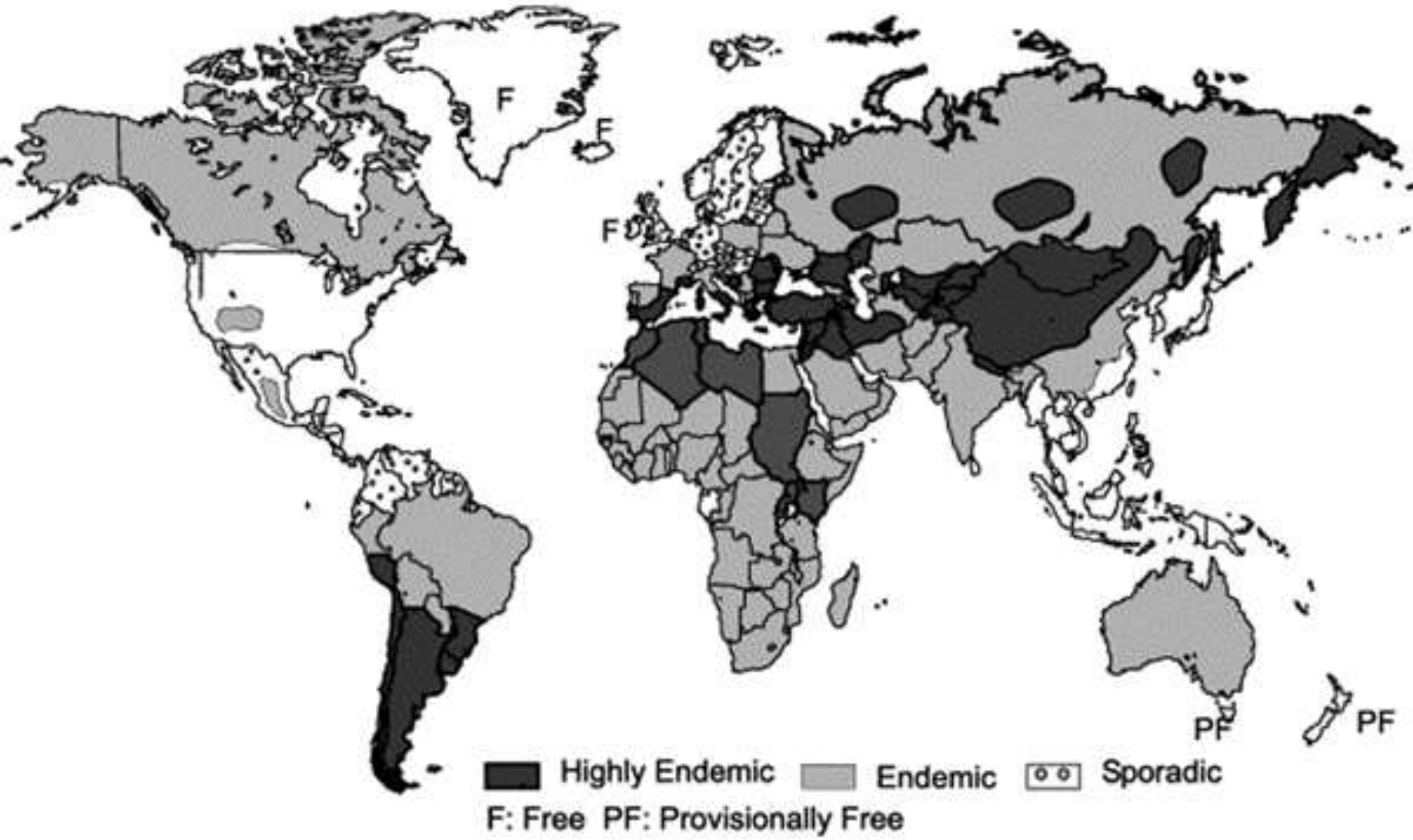
- 30 yo F born and raised in Iran
- Moved to Canada 2 y ago and here 3 mo ago
- Known cystic disease of lung and liver for some years. Seen in Iran and told it was “congenital”.
- Presented with Hemoptysis and dyspnea
- Started on prophylactic anti-parasitic agent
- Lobectomy followed by hepatic resections of cyts
- Serology positive for E. Granulosa

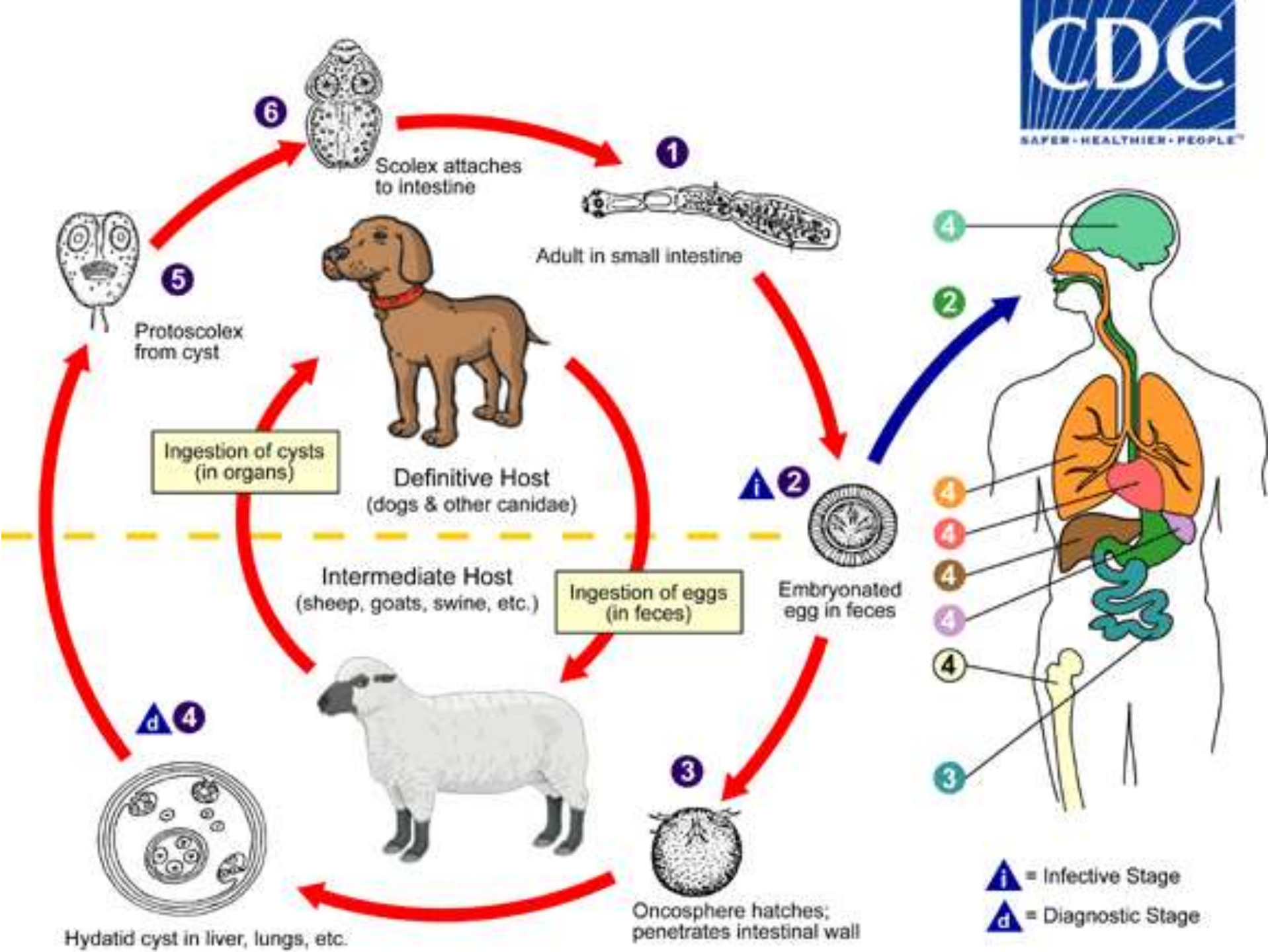
HISTORY

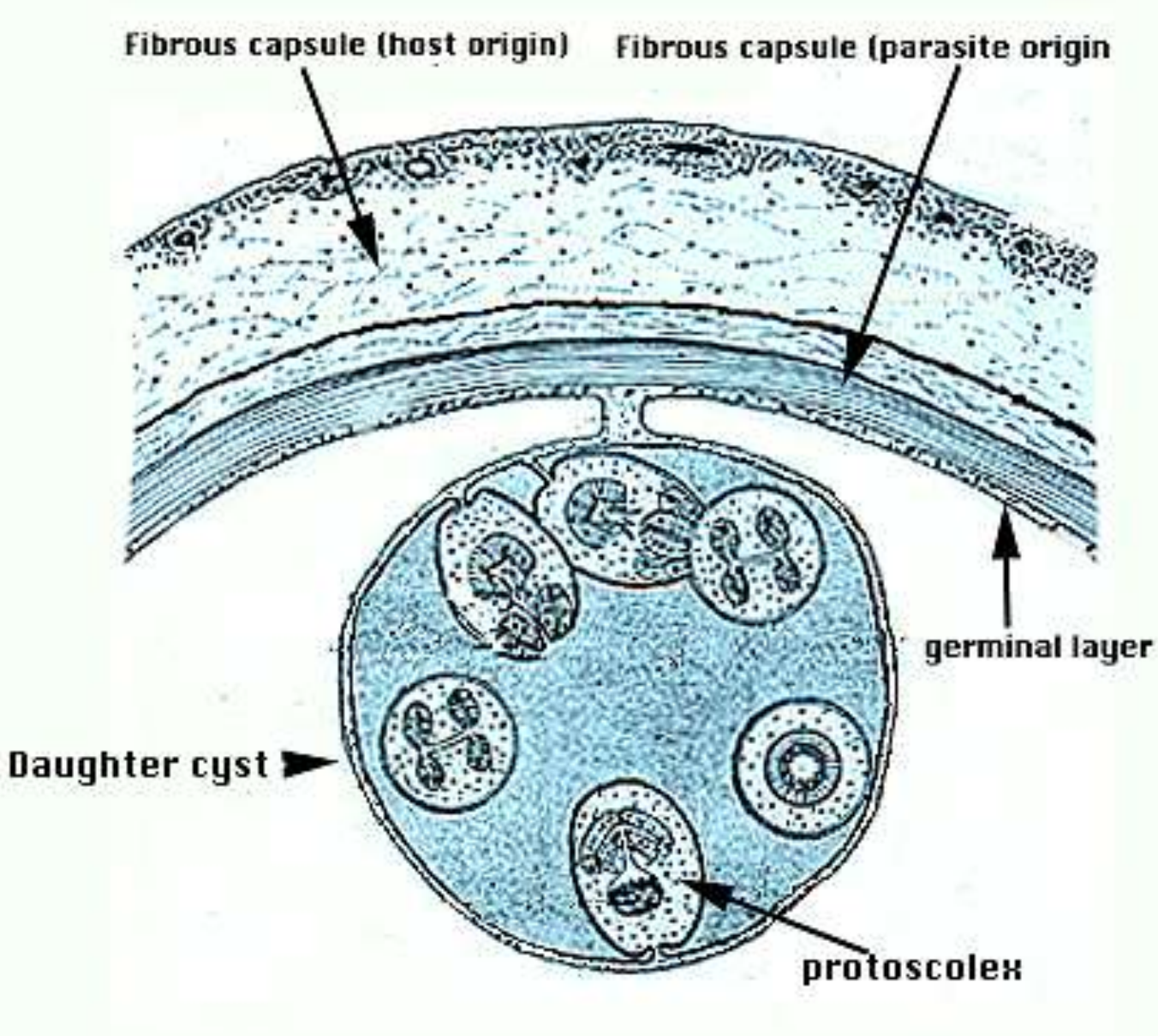
- Hydatid disease - one of the oldest diseases known to mankind, is a parasitic infestation caused by tapeworm of genus Echinococcus
- It was first described in the Talmud as a "Bladder full of water" .
- Hippocrates described the human hydatid disease more than two thousand years ago with a very interesting expression (liver filled with water) .

Epidemiology of E. Granulosus

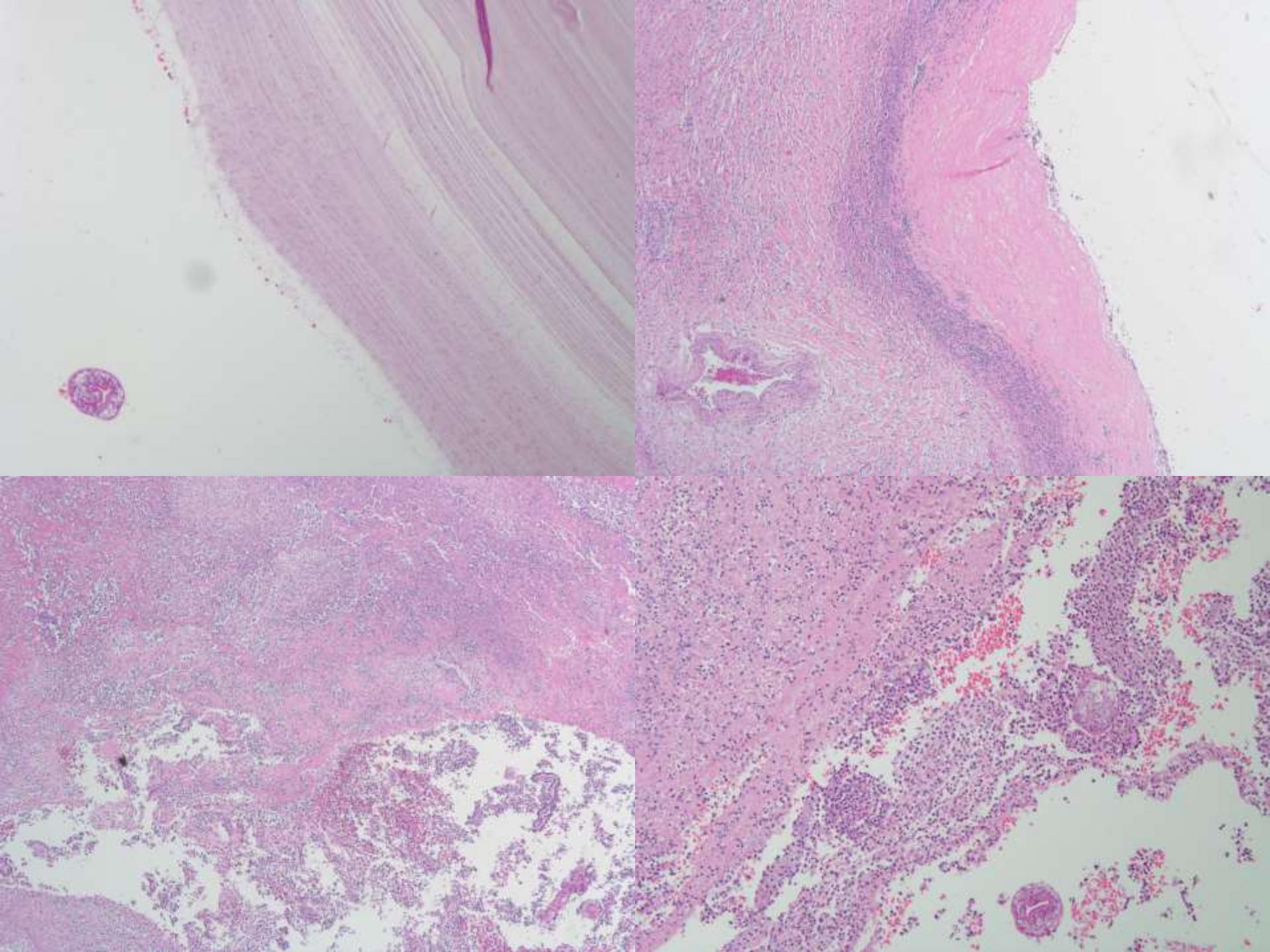
- Incidence has been falling for a number of decades but recently there appears to be a resurrgence of disease.
  - Reduction of control programs due to economic problems/lack of resources
- Geographic distribution depends on large numbers of nomadic or semi-nomadic sheep and goat flocks that represent the intermediate host and their close contact with dogs which are the primary host and who are thought to mostly provide the transmission of infection to humans

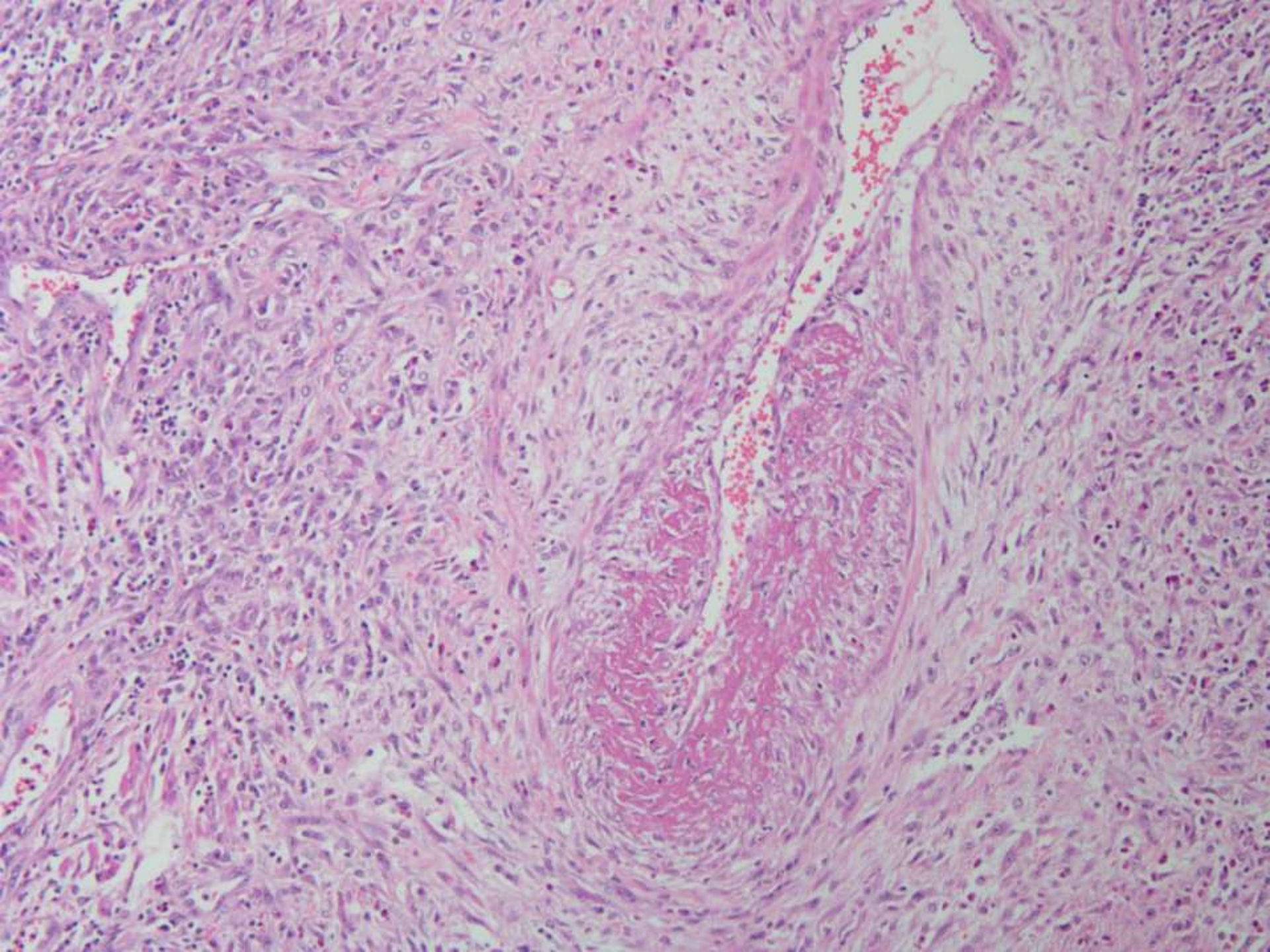

F
F
F
PF
PF
Highly Endemic
Endemic
Sporadic
F: Free PF: Provisionally Free


CDC
SAFER · HEALTHIER · PEOPLE™
6
Scolex attaches to intestine
1
Adult in small intestine
5
Protoscolex from cyst
Ingestion of cysts (in organs)
Definitive Host
(dogs & other canidae)
i 2
Embryonated egg in feces
Intermediate Host
(sheep, goats, swine, etc.)
Ingestion of eggs (in feces)
d 4
Hydatid cyst in liver, lungs, etc.
3
Oncosphere hatches; penetrates intestinal wall
4
2
4
4
4
4
4
3
i = Infective Stage
d = Diagnostic Stage



Hydatid cyst






Liver
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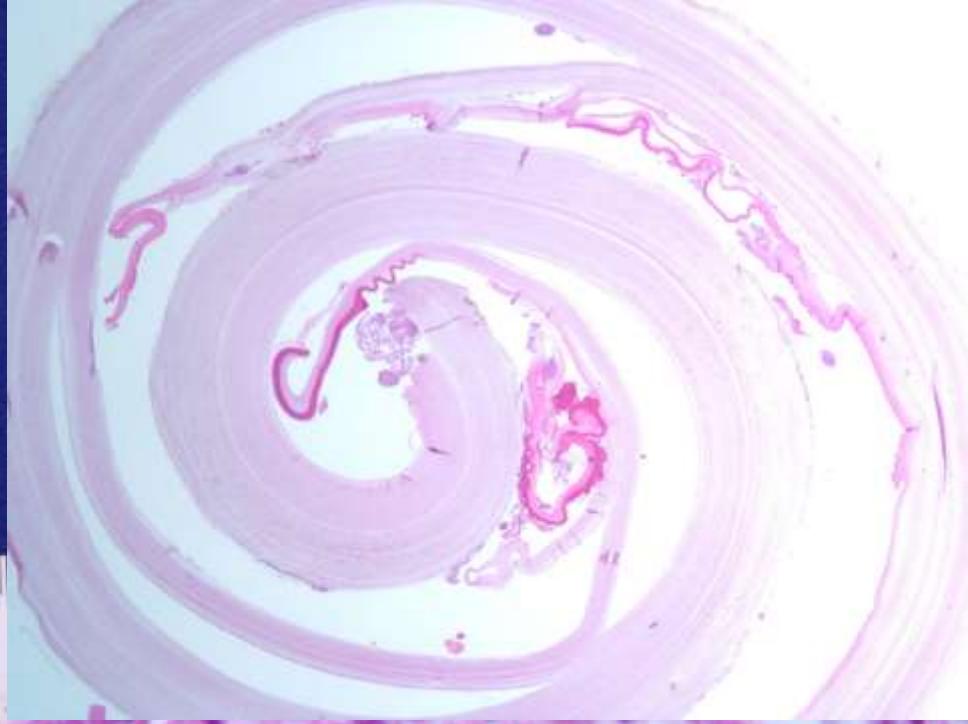
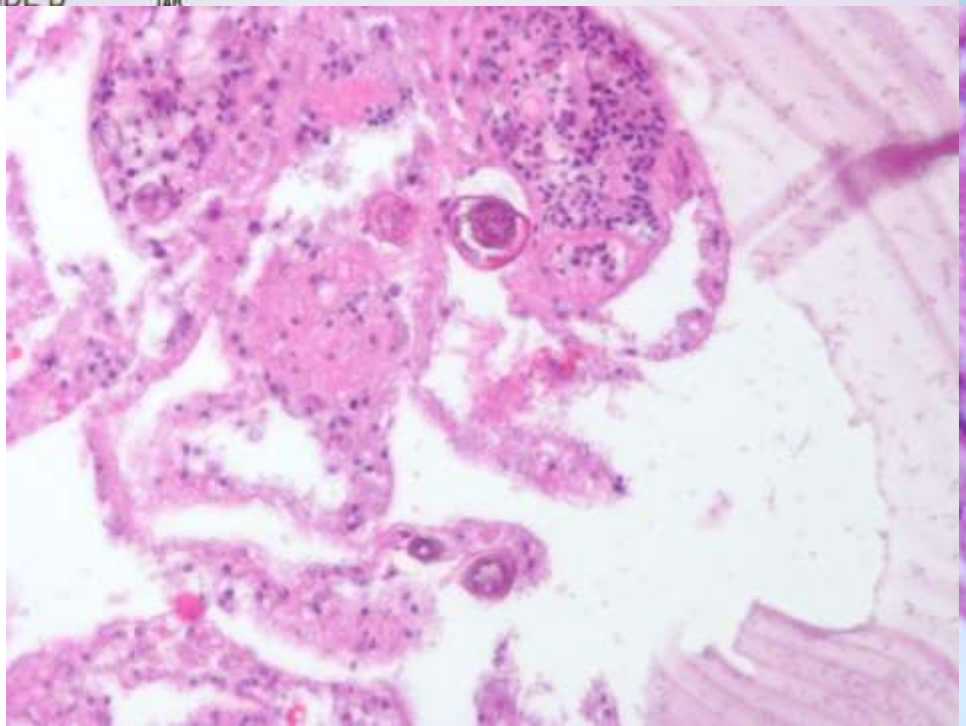
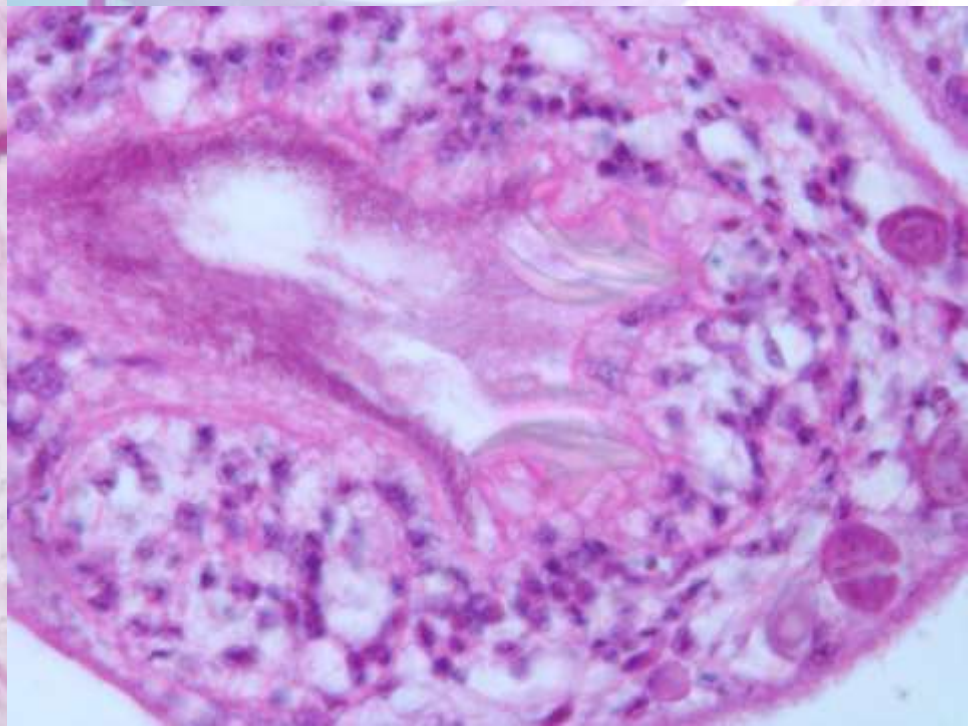


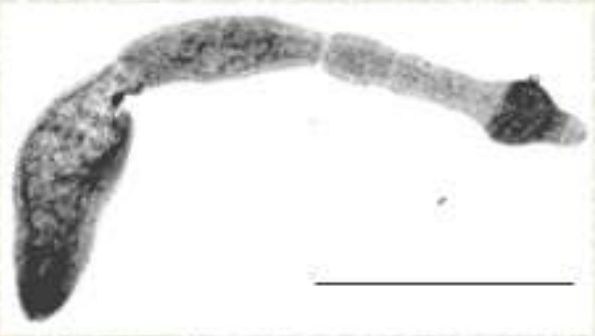




Echinococcosis

- A zoonotic infestation by a tapeworm causing hydatid disease

- *Echinococcus granulosus*
  - Cystic echinococcosis
  - Produces cystic lesions



- *Echinococcus multilocularis*
  - Alveolar echinococcosis
  - Invasive solid lesions of firm consistency, full of connective tissue and a jelly-like material

Echinococcus multilocularis

- Rare multilocular alveolar form
- Endemic in much of the upper Midwest of the United States, Alaska, Canada, Japan, central Europe, and parts of Russia
- Definitive hosts are foxes and, less commonly, cats and dogs
- Intermediate hosts are wild rodents
- Humans are infested either by direct contact with definitive hosts or indirectly by intake of contaminated water or contaminated plants such as wild berries

Caire Nail et al. Journal of Medical Case Reports (2017) 11:113
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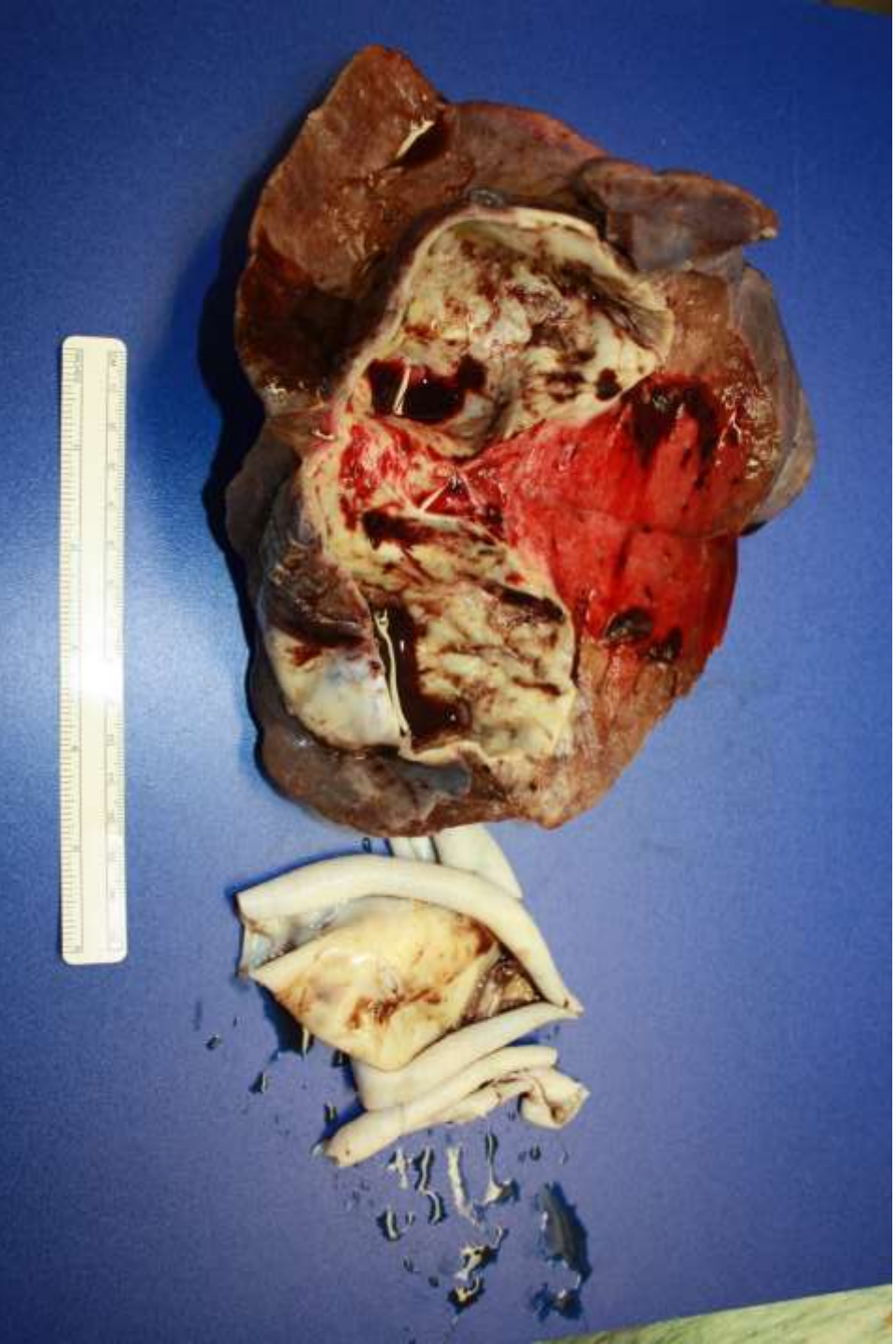
Journal of
Medical Case Reports

CASE REPORT Open Access

Disseminated alveolar echinococcosis resembling metastatic malignancy: a case report

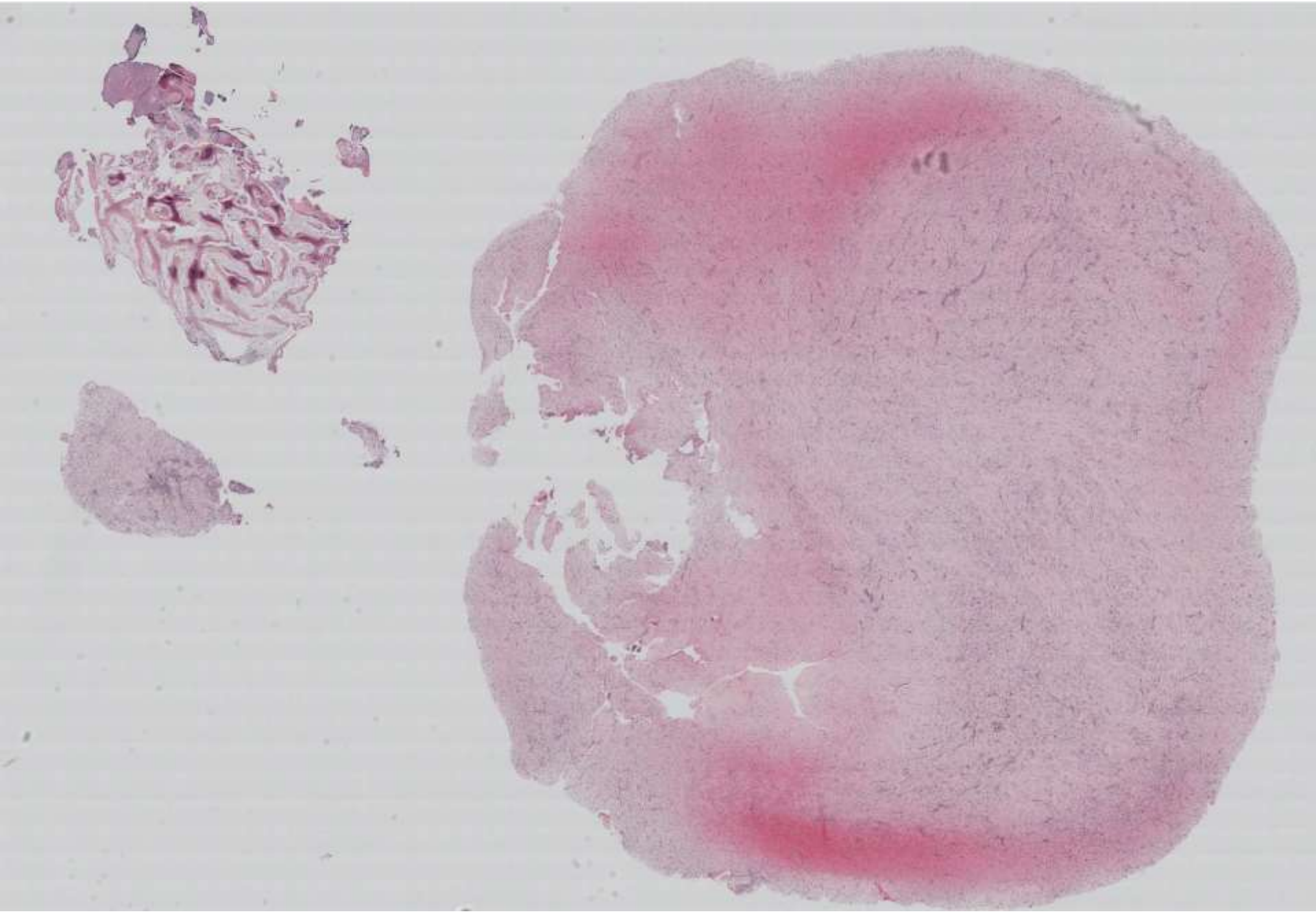
Laura Caire Nail[1], Ezequiel Rodríguez Reimundes[1*], Christelle Weibel Galluzzo[2], Dan Lebowitz[1], Yasmine Lucile Ibrahim[3], Johannes Alexander Lobrinus[3] and François Chappuis[2]

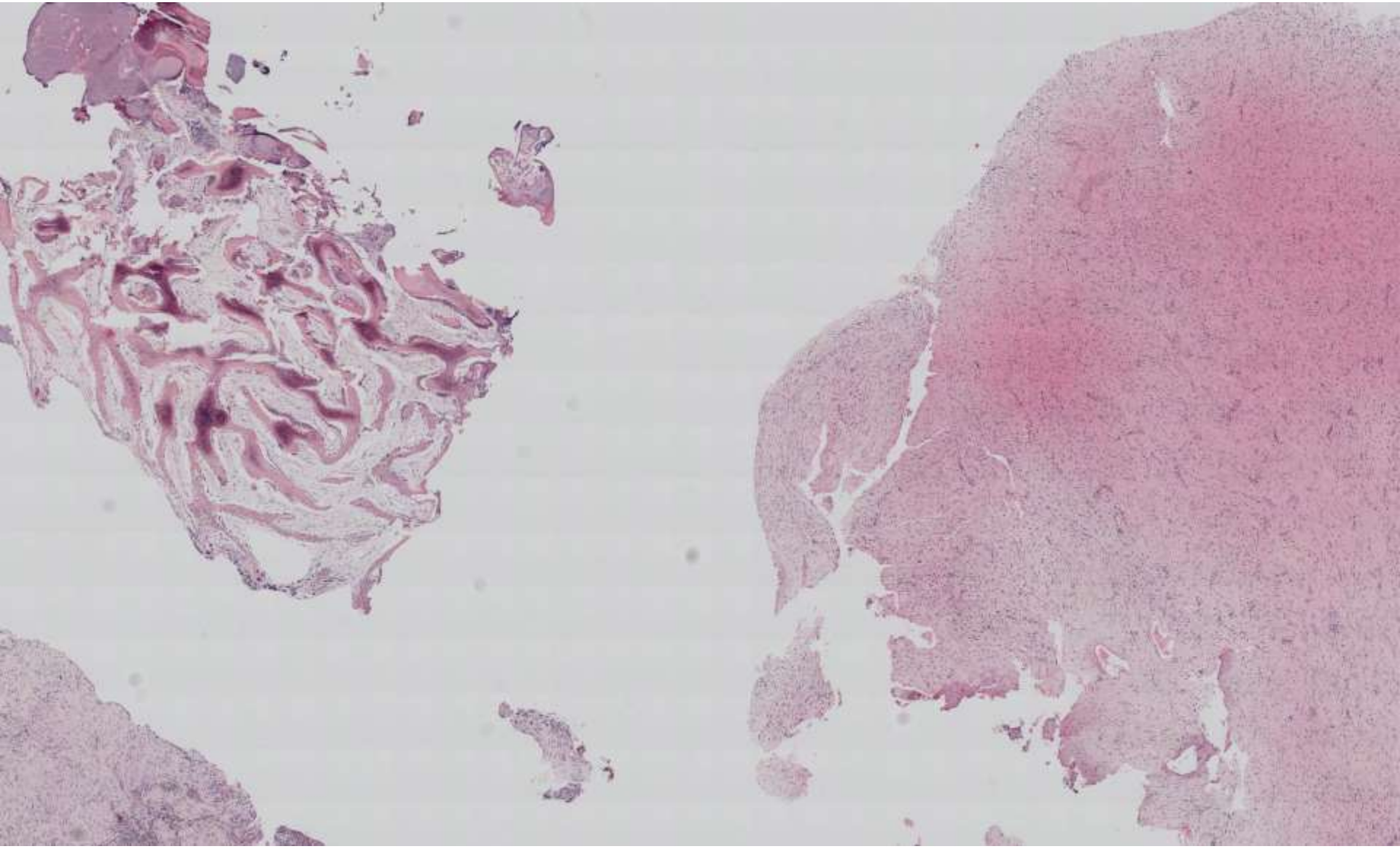


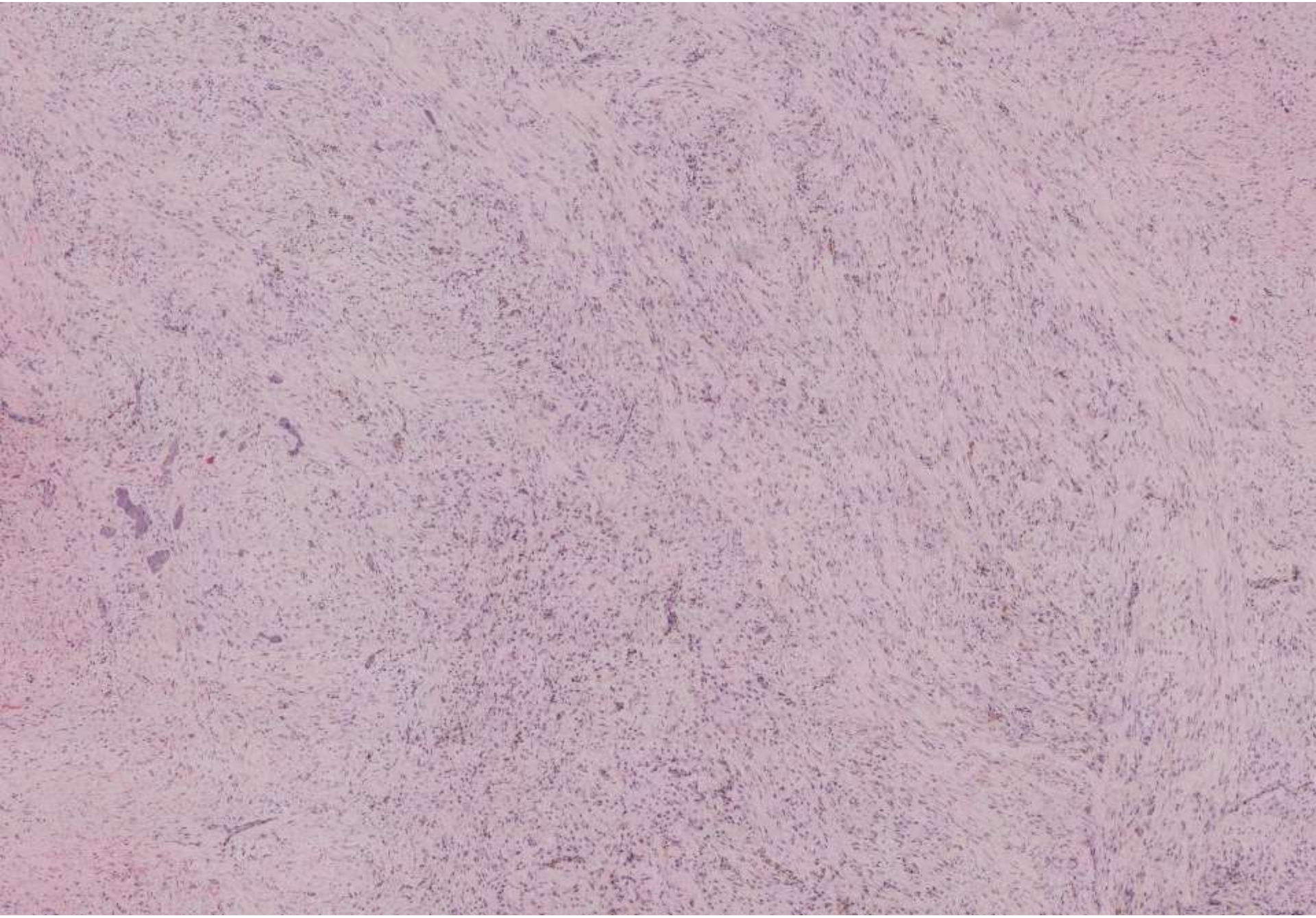
SB 6172
(scanned slide available)

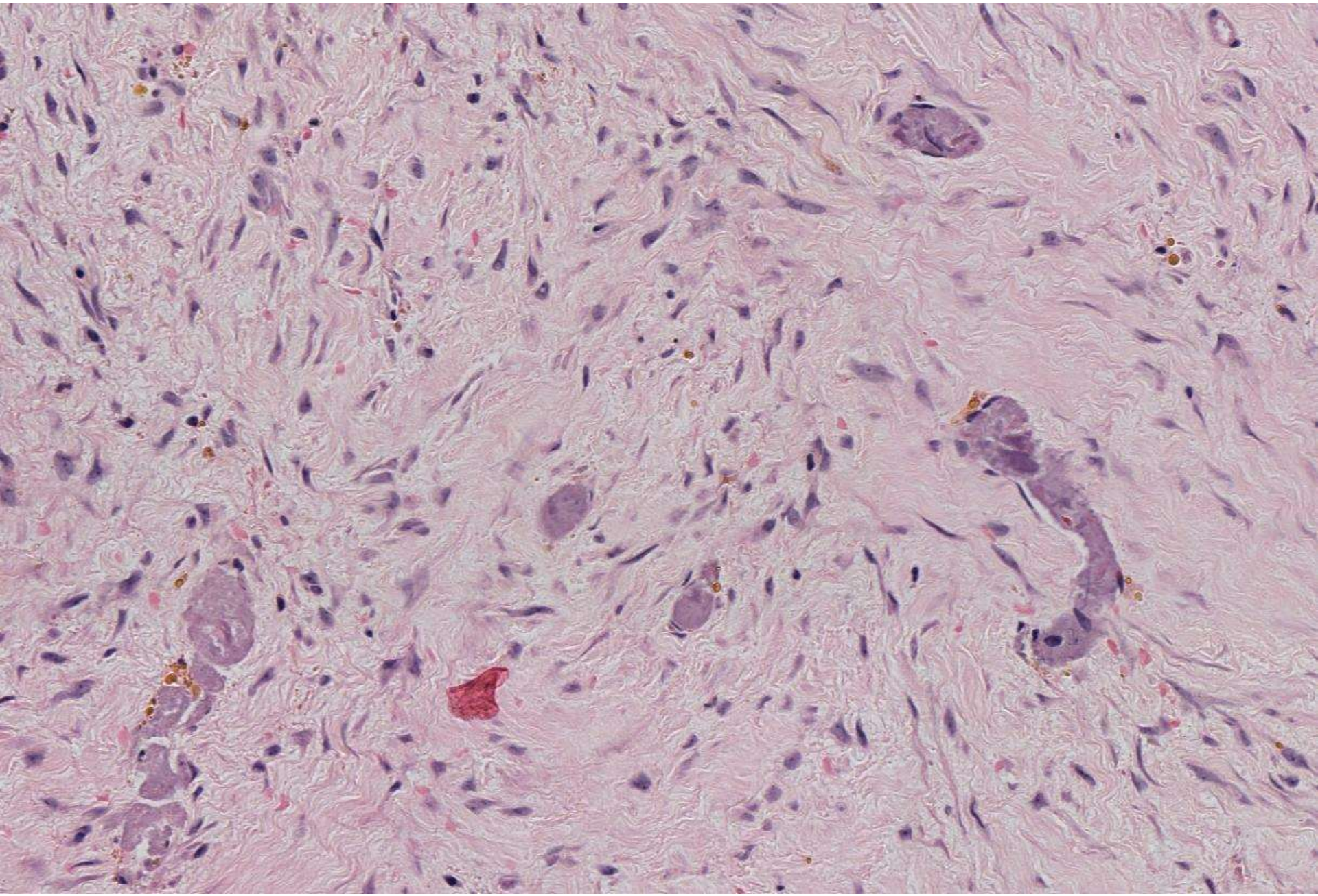
Balaram Puligandla; Kaiser Oakland

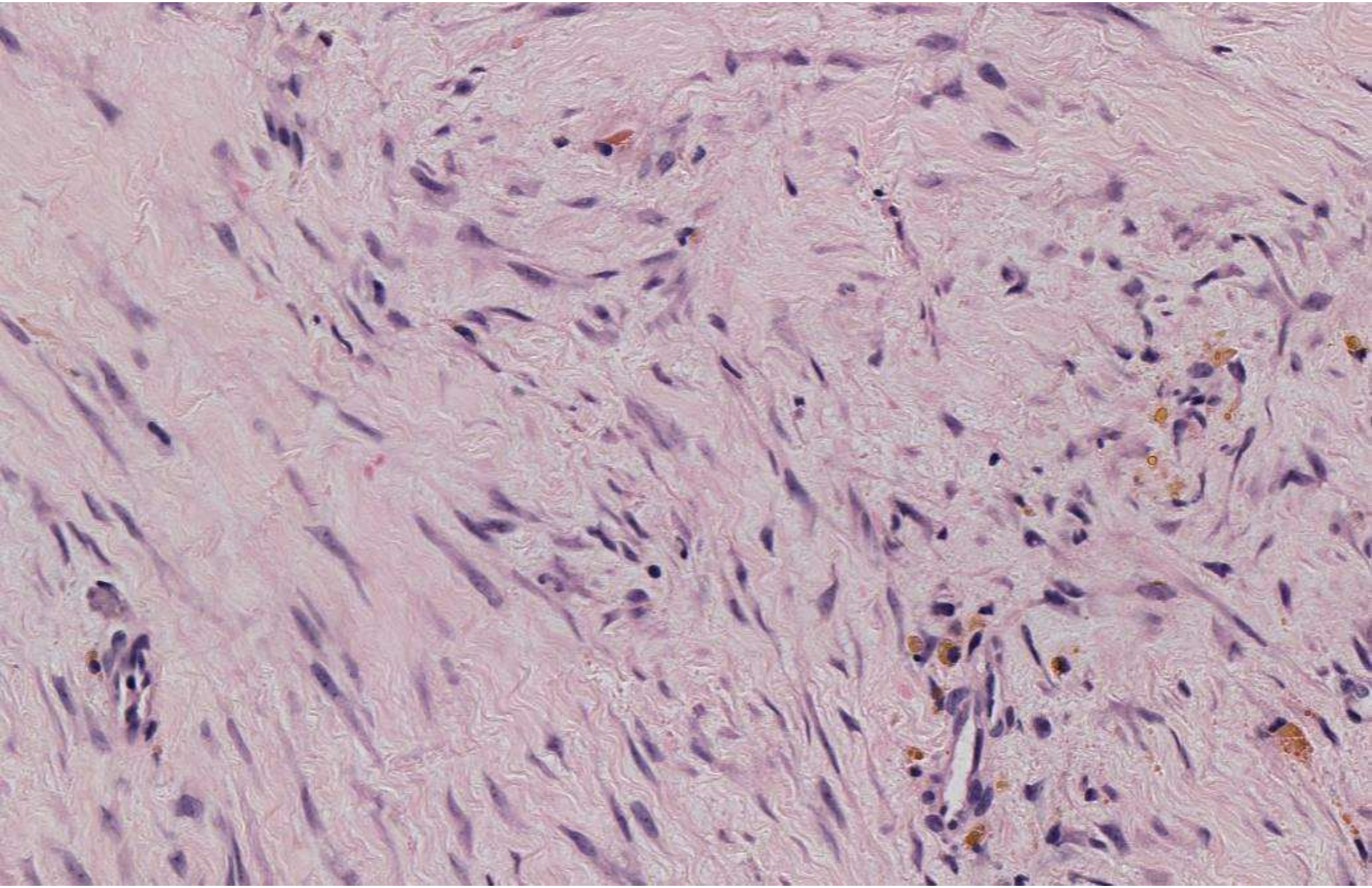
10-year-old girl with intra-bony mandibular lesion, rule out central giant cell lesion.

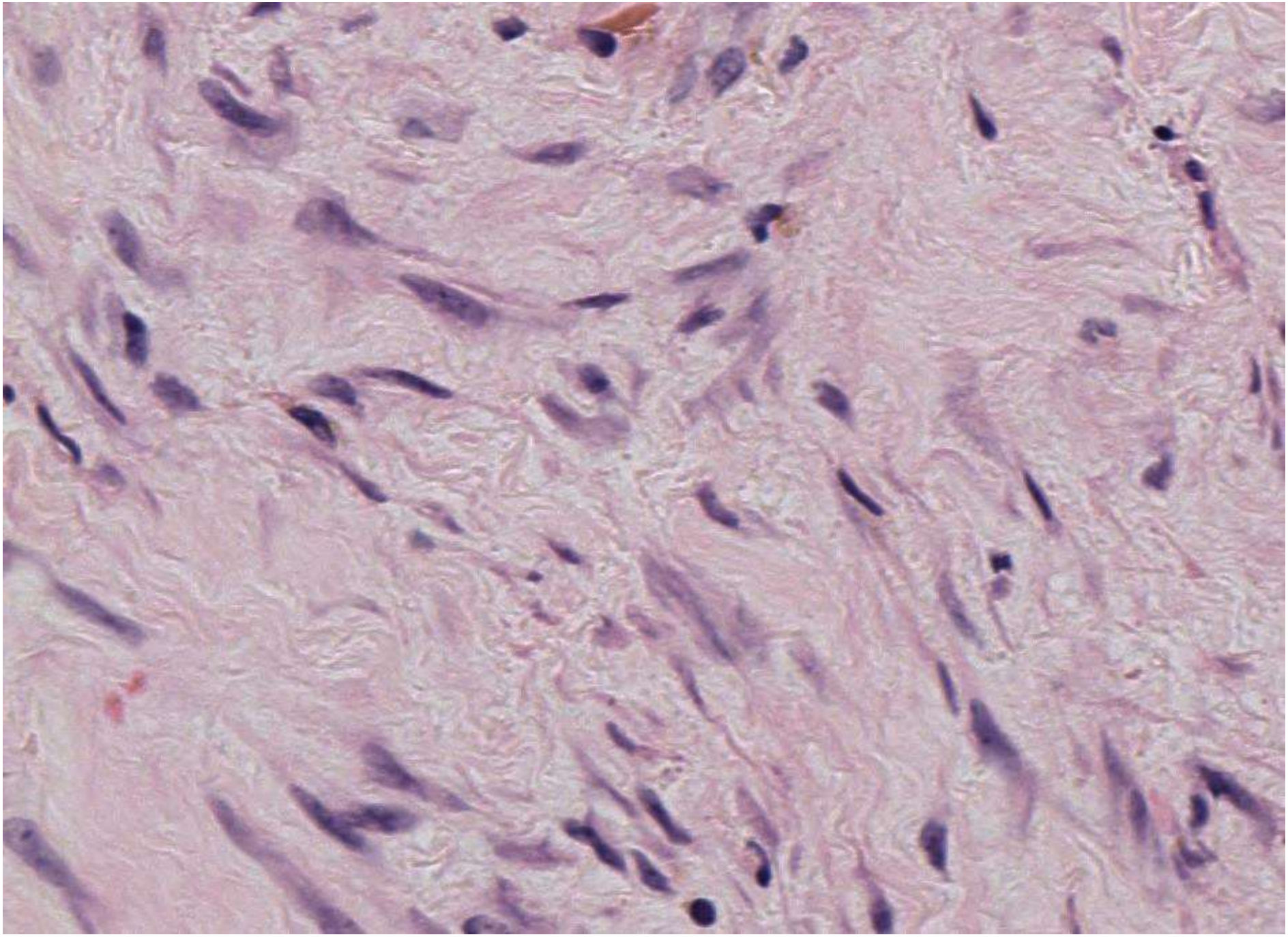












DIAGNOSIS?

Desmoplastic Fibroma

Desmoplastic Fibroma

- First described by Jaffe in 1958 as Desmoplastic Fibroma (DF) of Bone
- Rare, 0.1% of primary bone tumors
- First report of gnathic involvement in 1965
- WHO: Most common in mandible (22%), Femur (15%), Pelvic Bones(13%)

Gnathic Desmoplastic Fibroma

- >80% involve mandible, majority posteriorly
- Median age: 17 years (6 mos-60 years)
- Female predominance
- Asymptomatic swelling most common presentation
- Radiolucent with ill-defined non-sclerotic margins
- Head Neck Pathol 2015: Jun; 9(2): 196-204

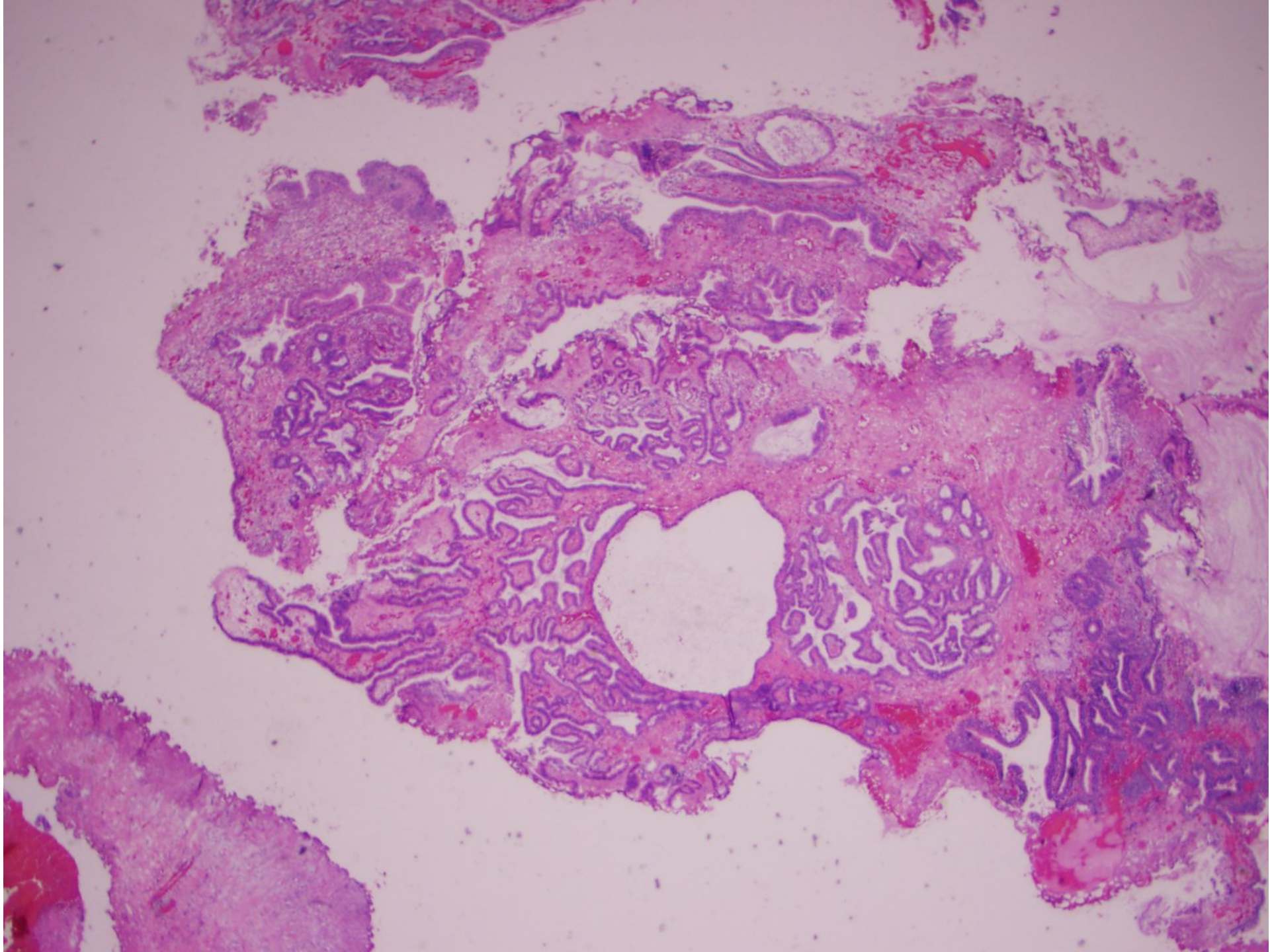
- Thought to be counterpart to fibromatosis
- No reliable IHC markers, Dx of exclusion
- No consistent B catenin staining seen in gnathic DF
- DDX: Nodular fasciitis, fibro-osseous lesions, odontogenic fibroma

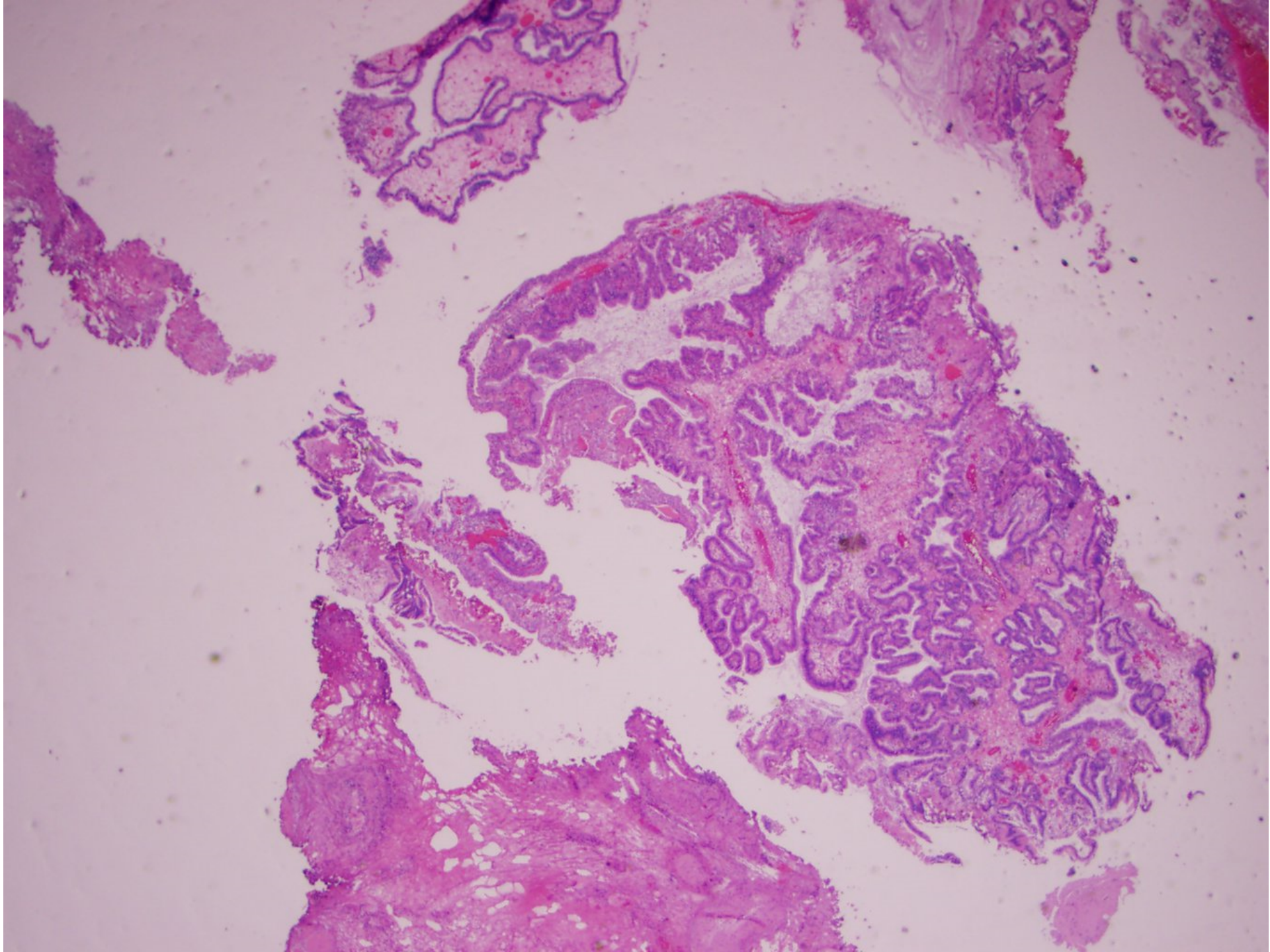
Summary

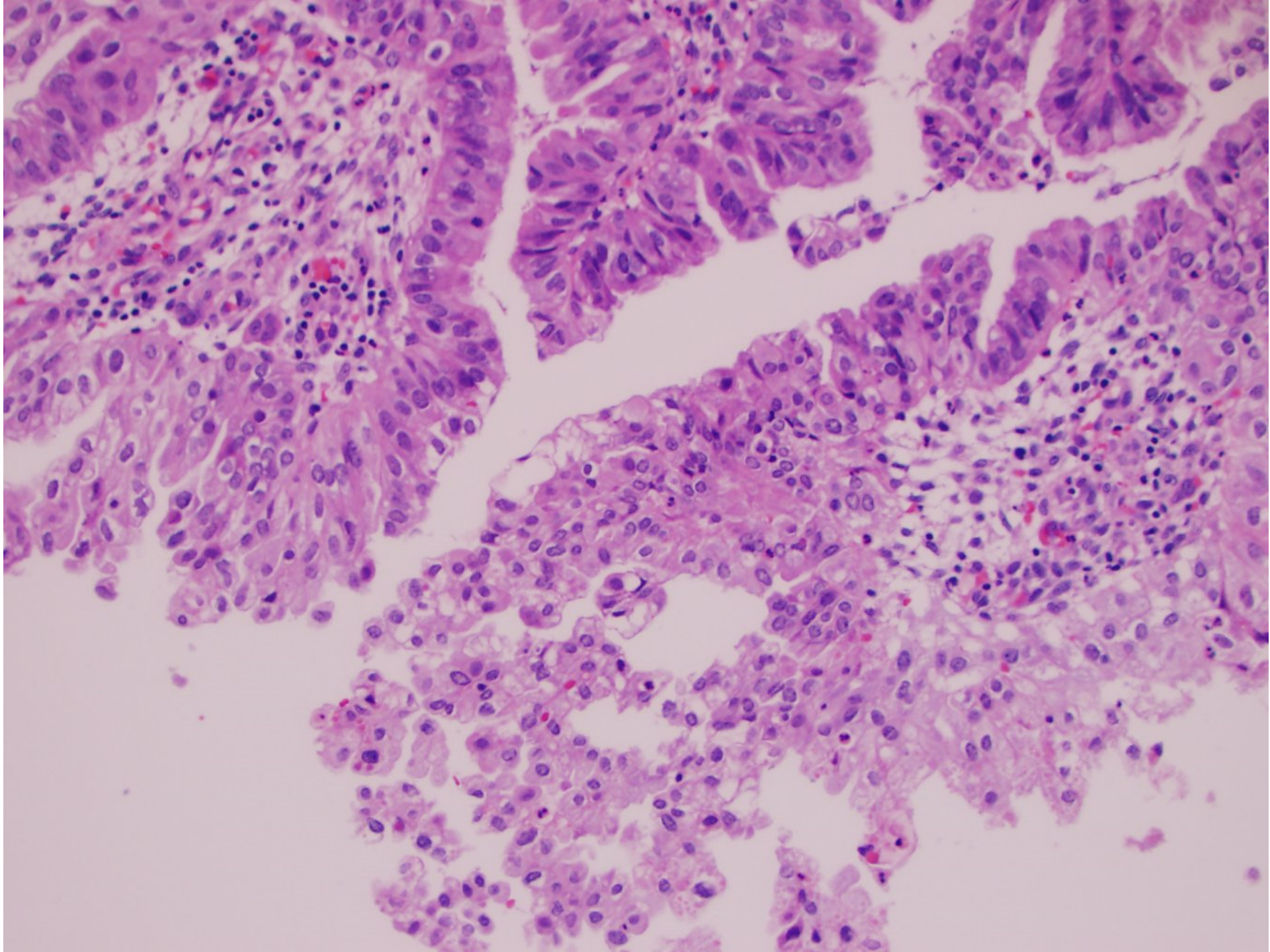
- DF Rare
- Seen most commonly in the mandible
- Young, female patients
- No reliable IHC markers
- B catenin most often negative

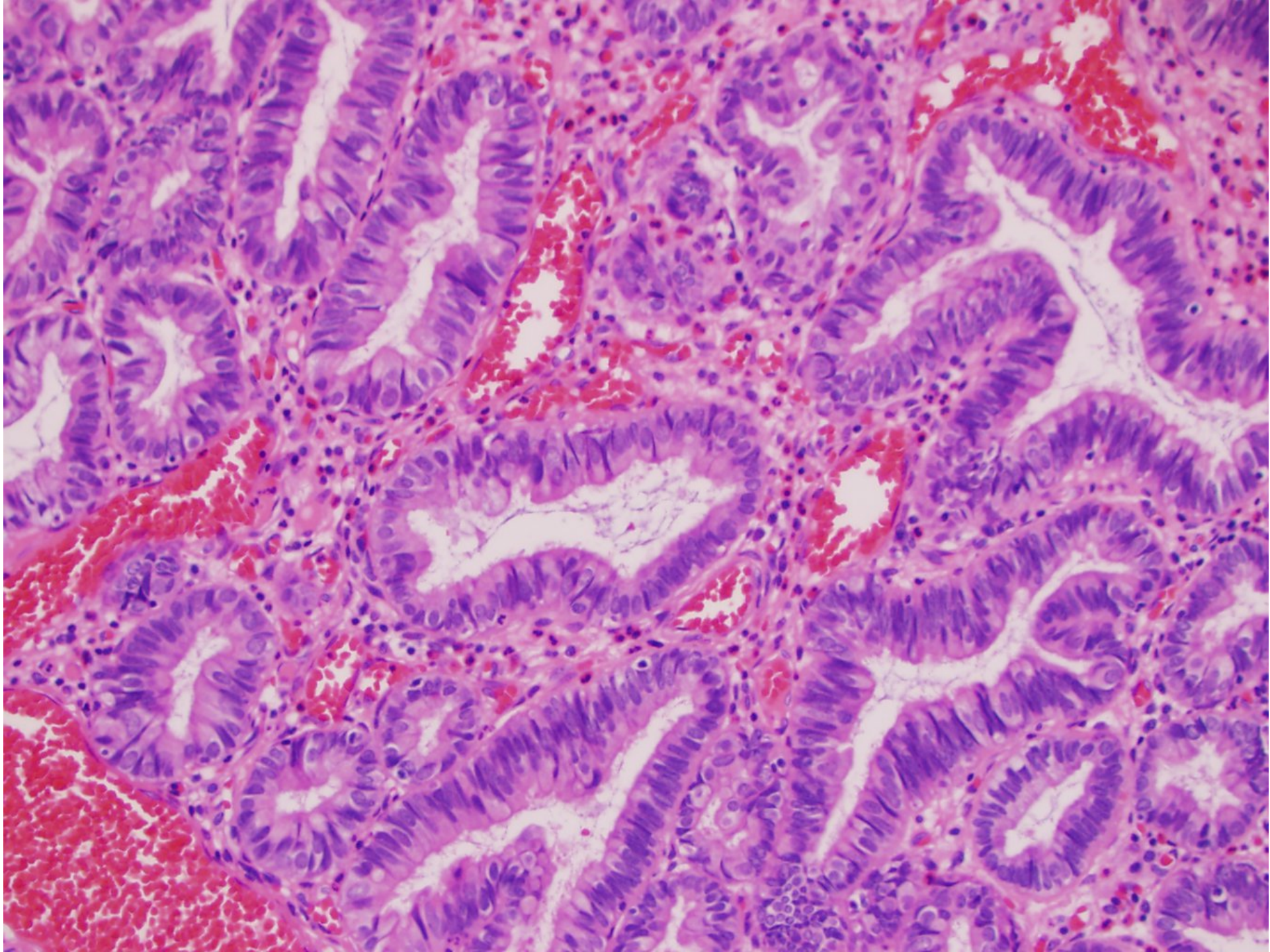
SB 6173

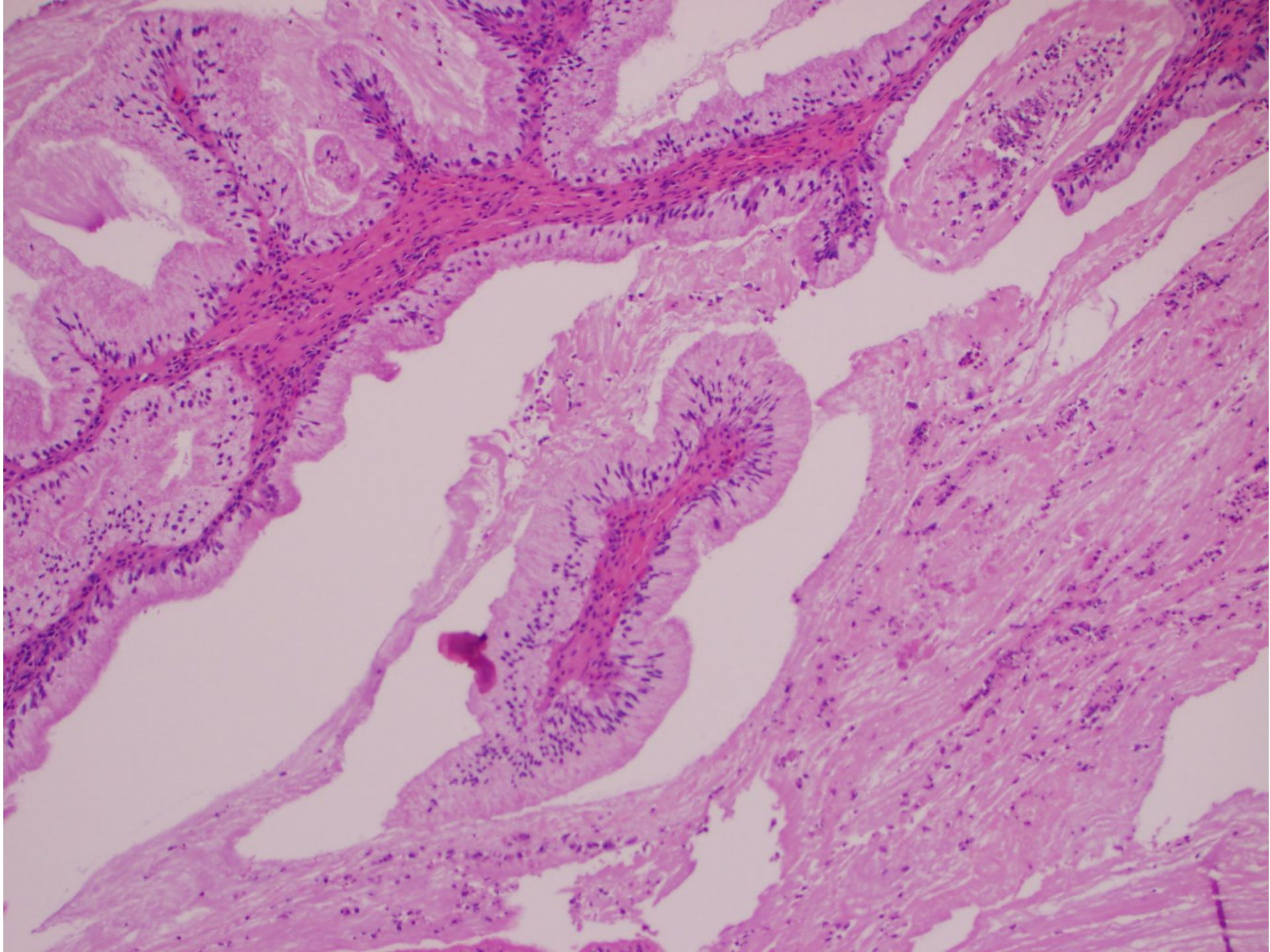
65-year-old woman with 5.2cm posterior bladder mass and right ureteral obstruction.

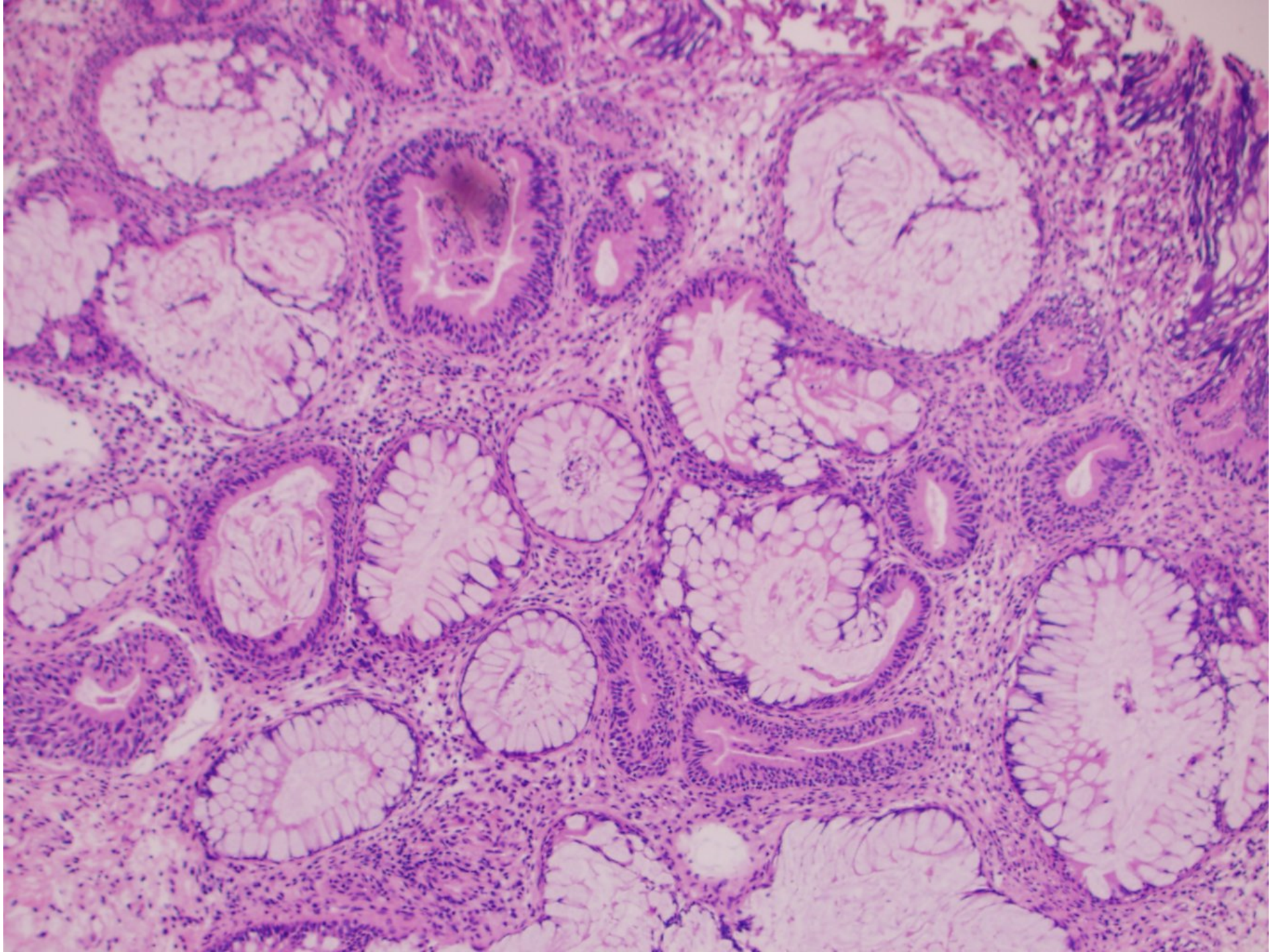


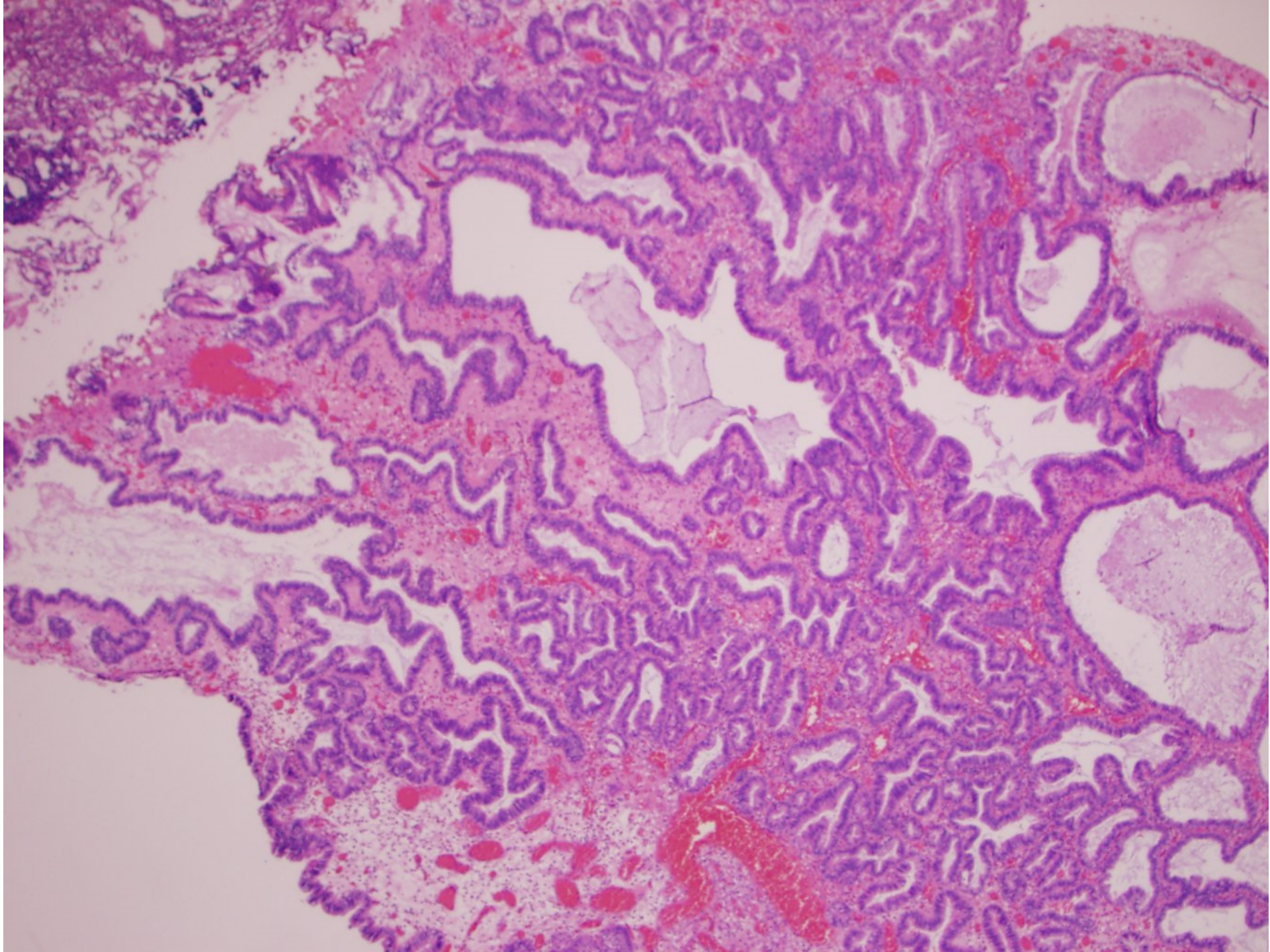


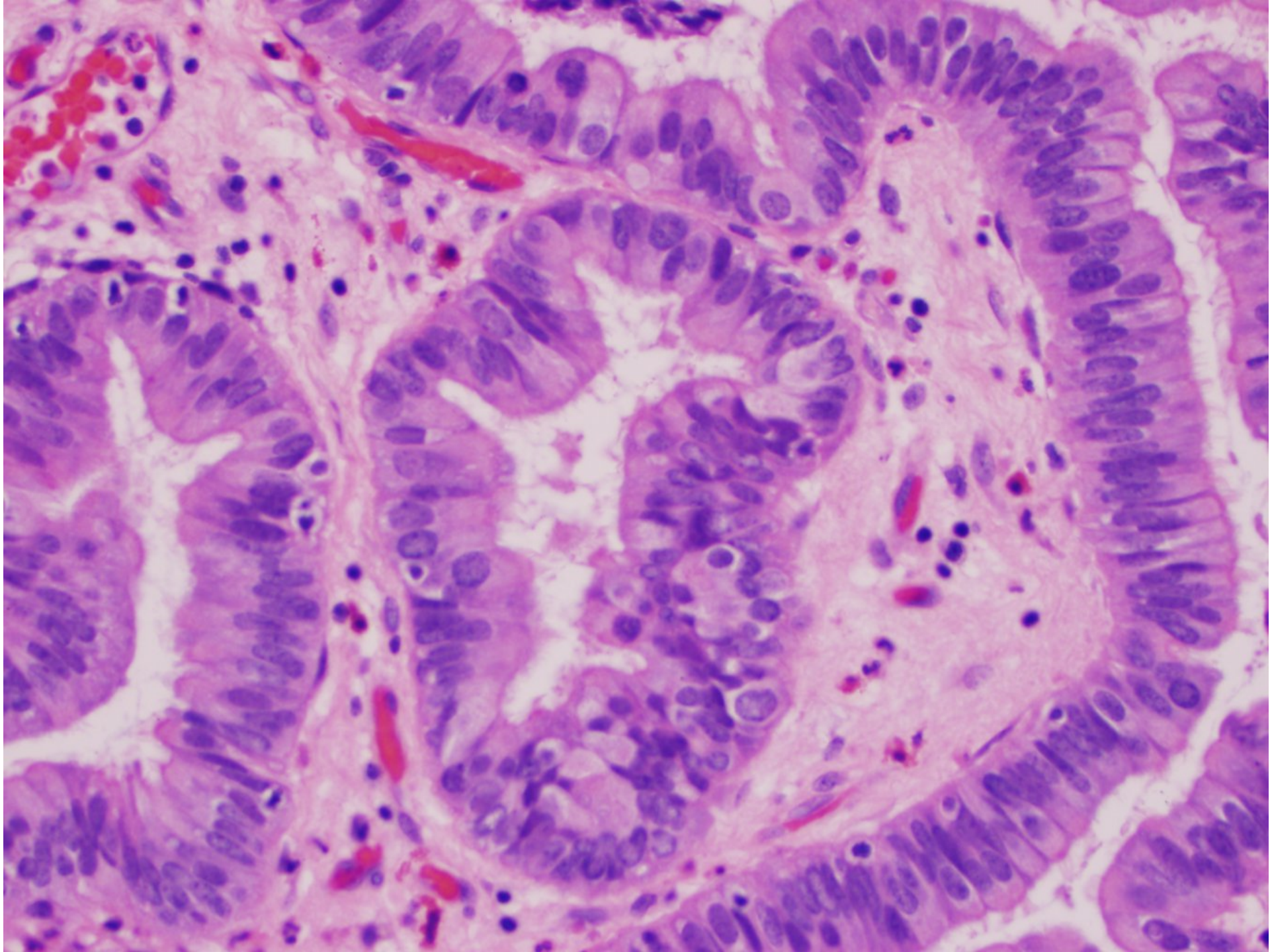


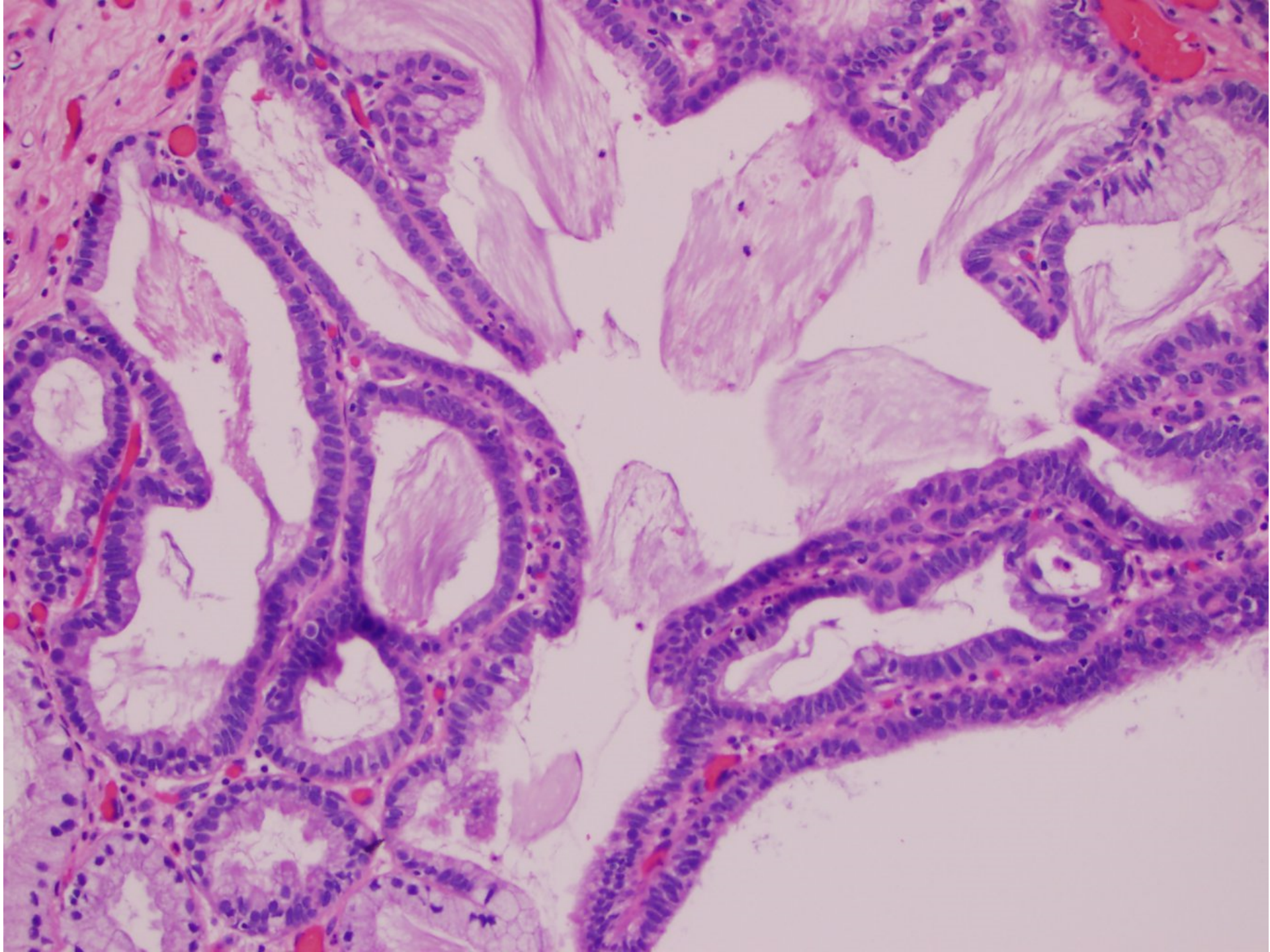


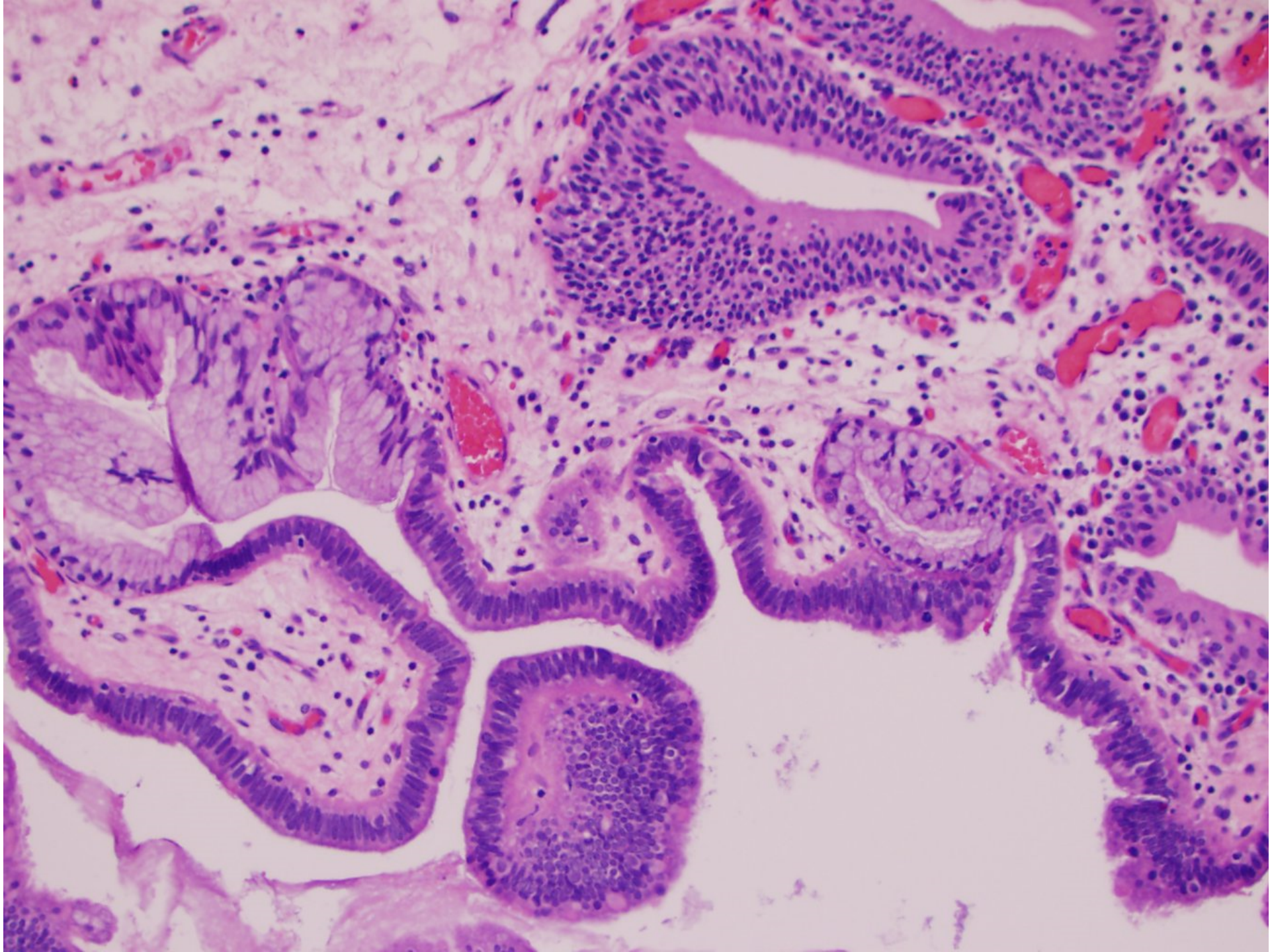


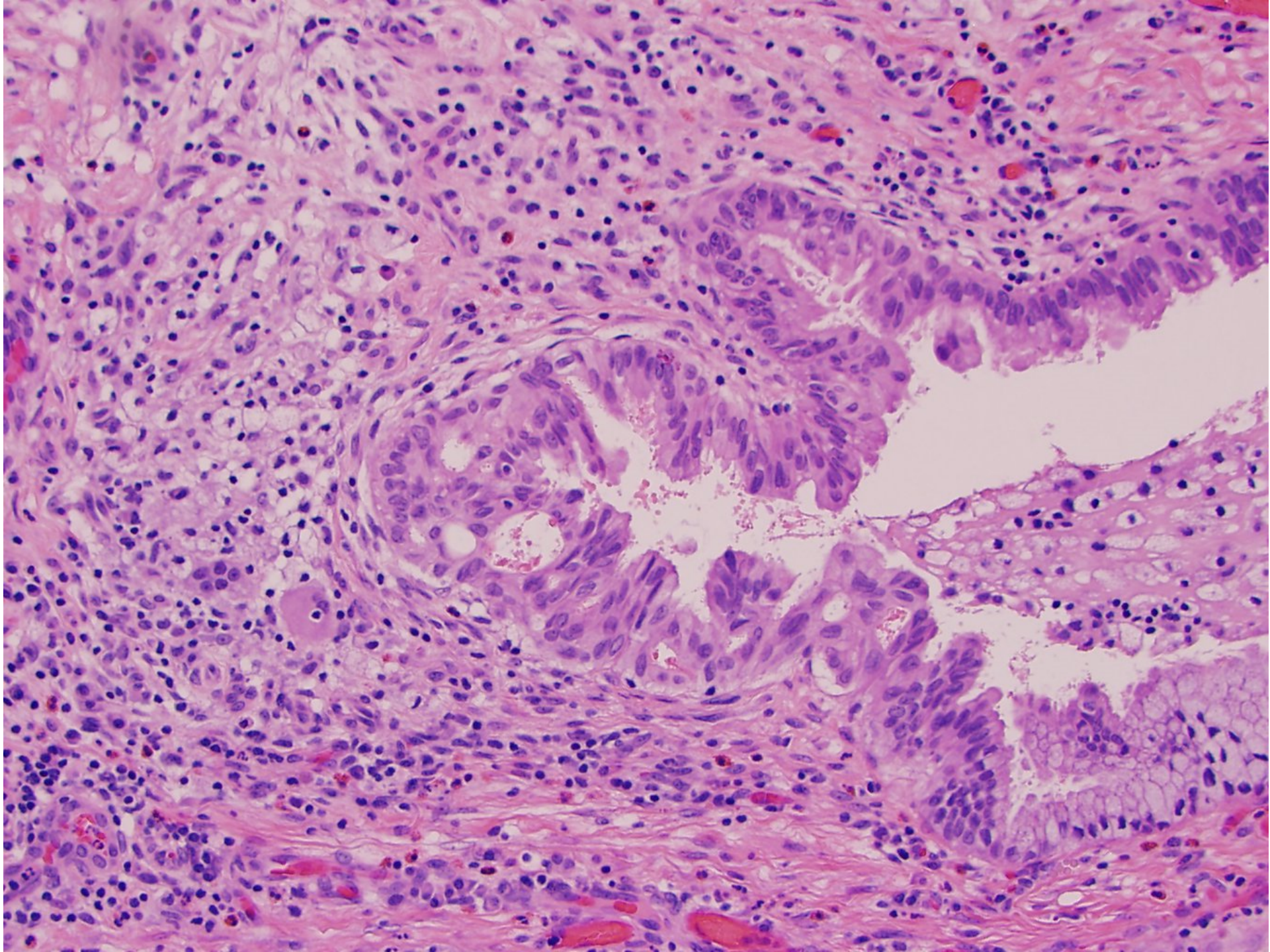


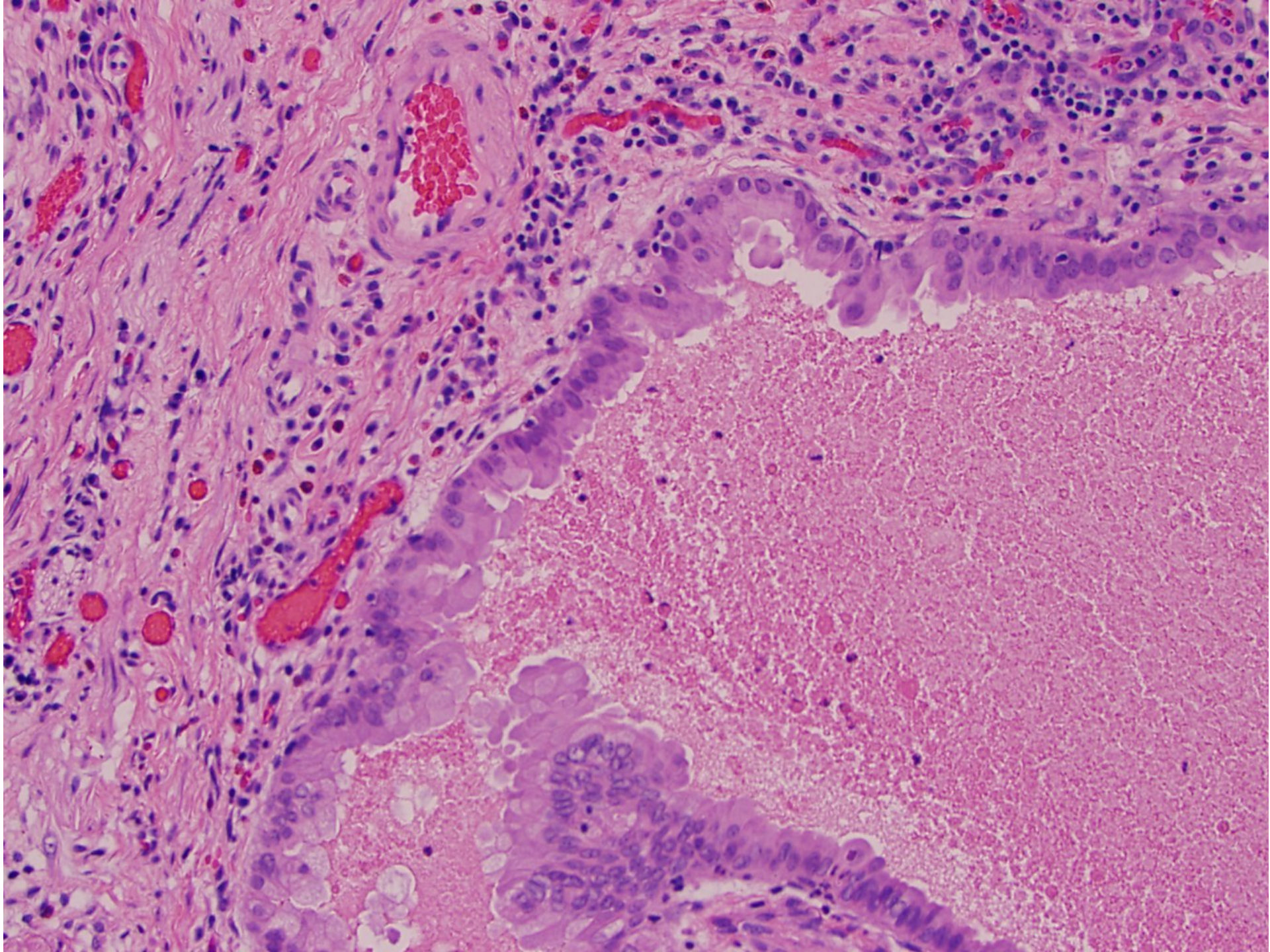


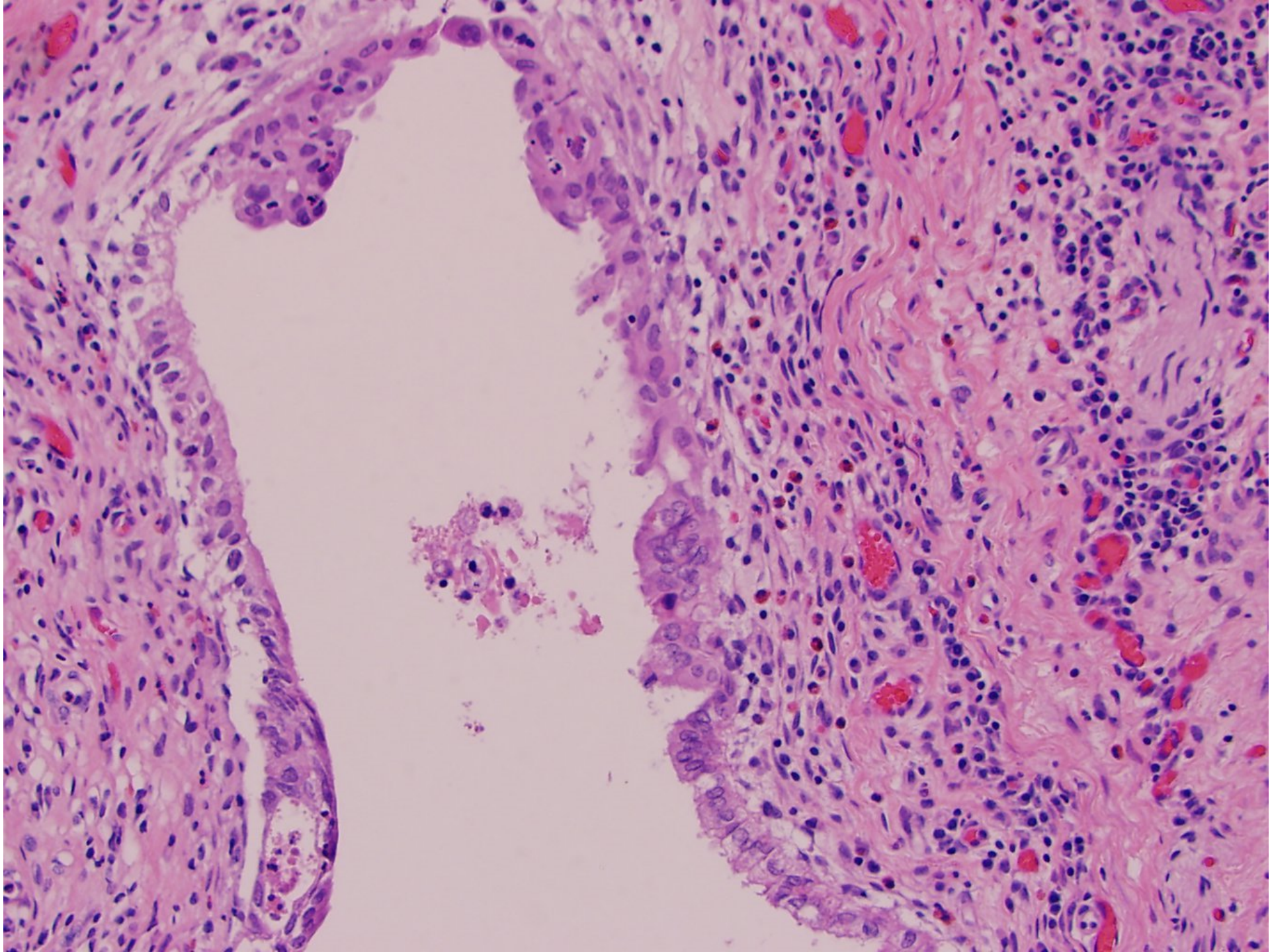


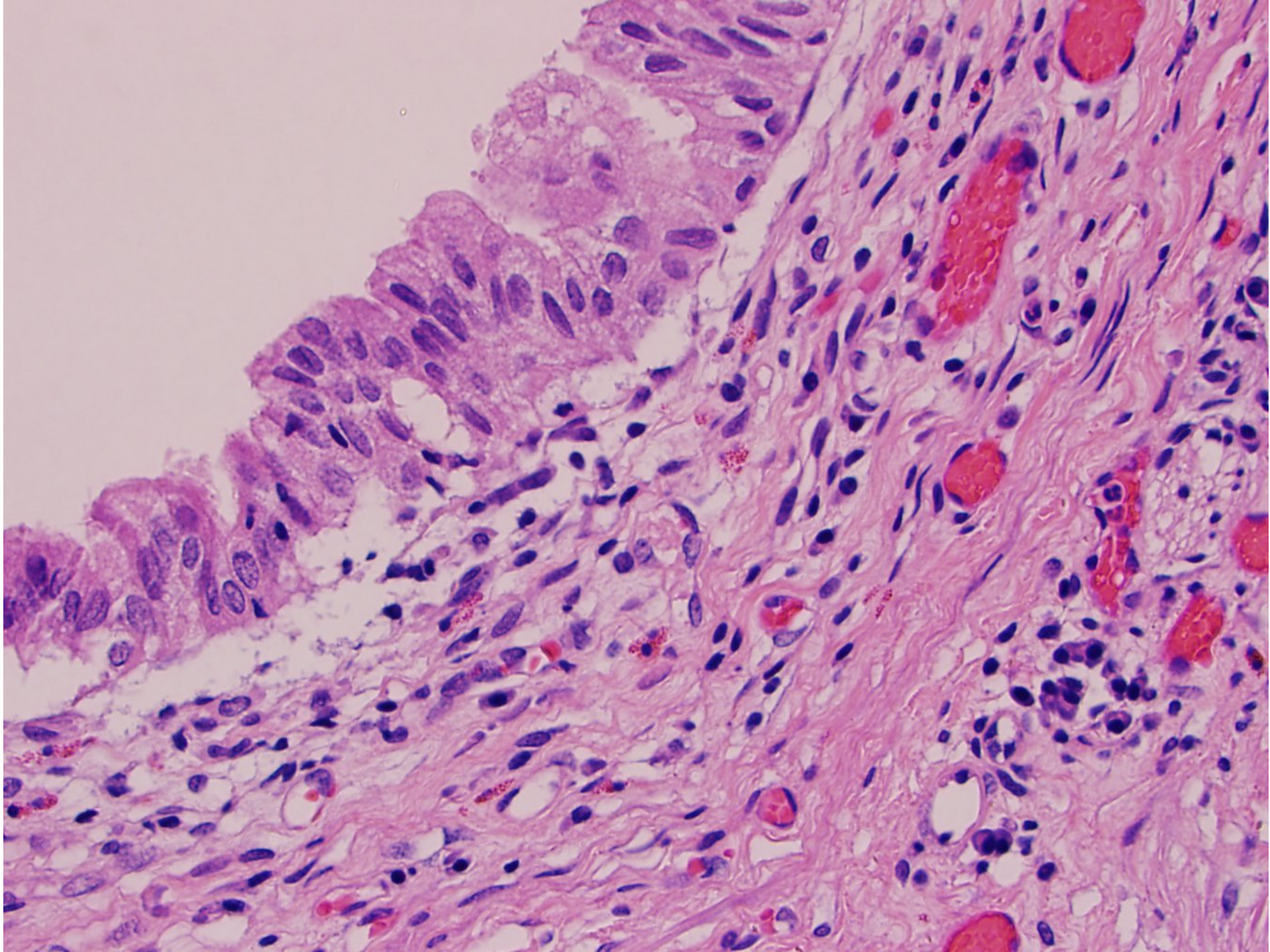












DIAGNOSIS?

Mullerianosis

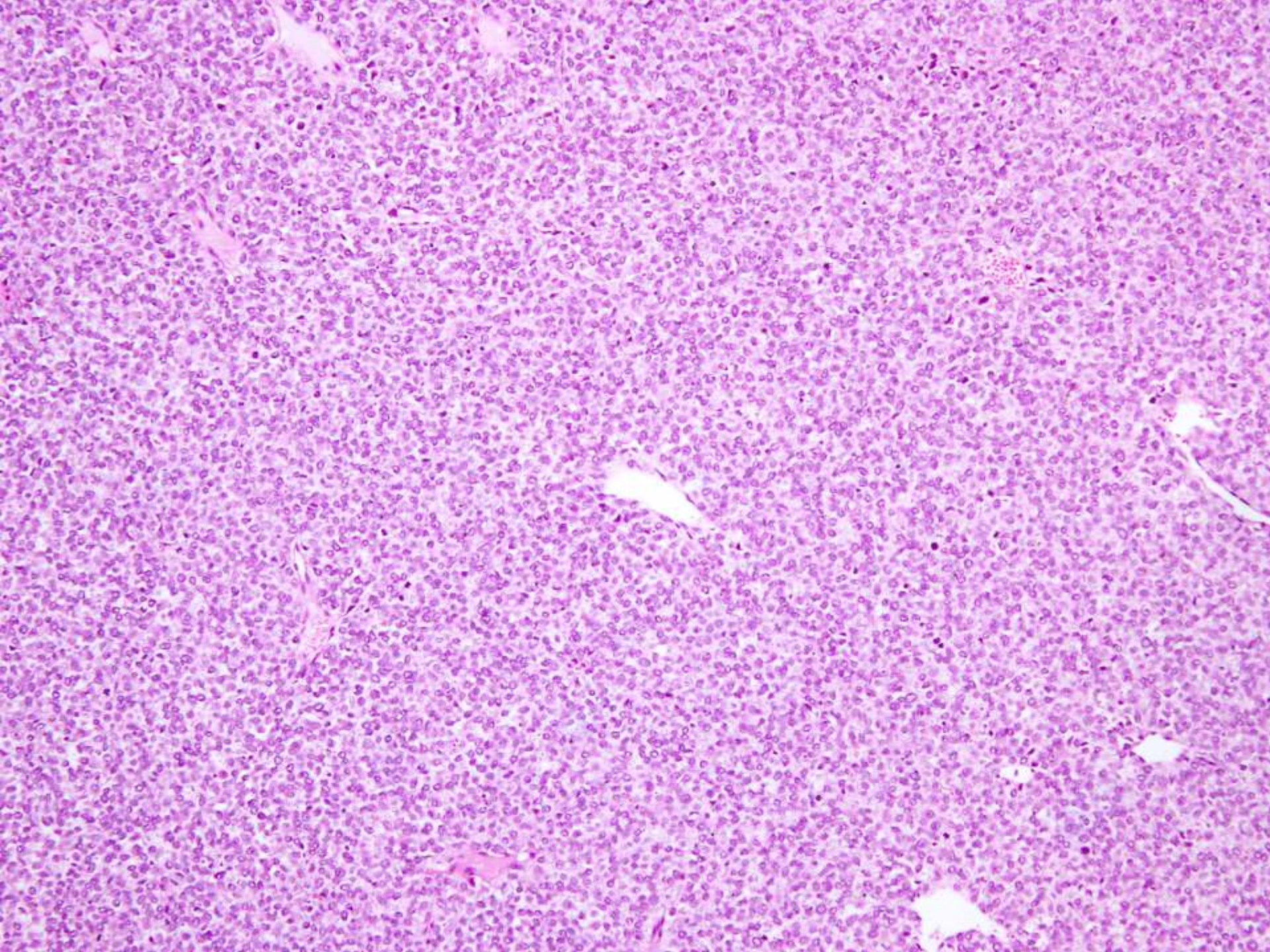
- Rare lesion of bladder mimicking tumor
- Usually seen in women of child-bearing age
- Includes endosalpingiosis (usually predominant component) endocervicosis, and endometriosis
- Differentiated from adenocarcinoma by architecture and cytology
- Implantative vs. metaplastic
- Benign process- treated hormonally

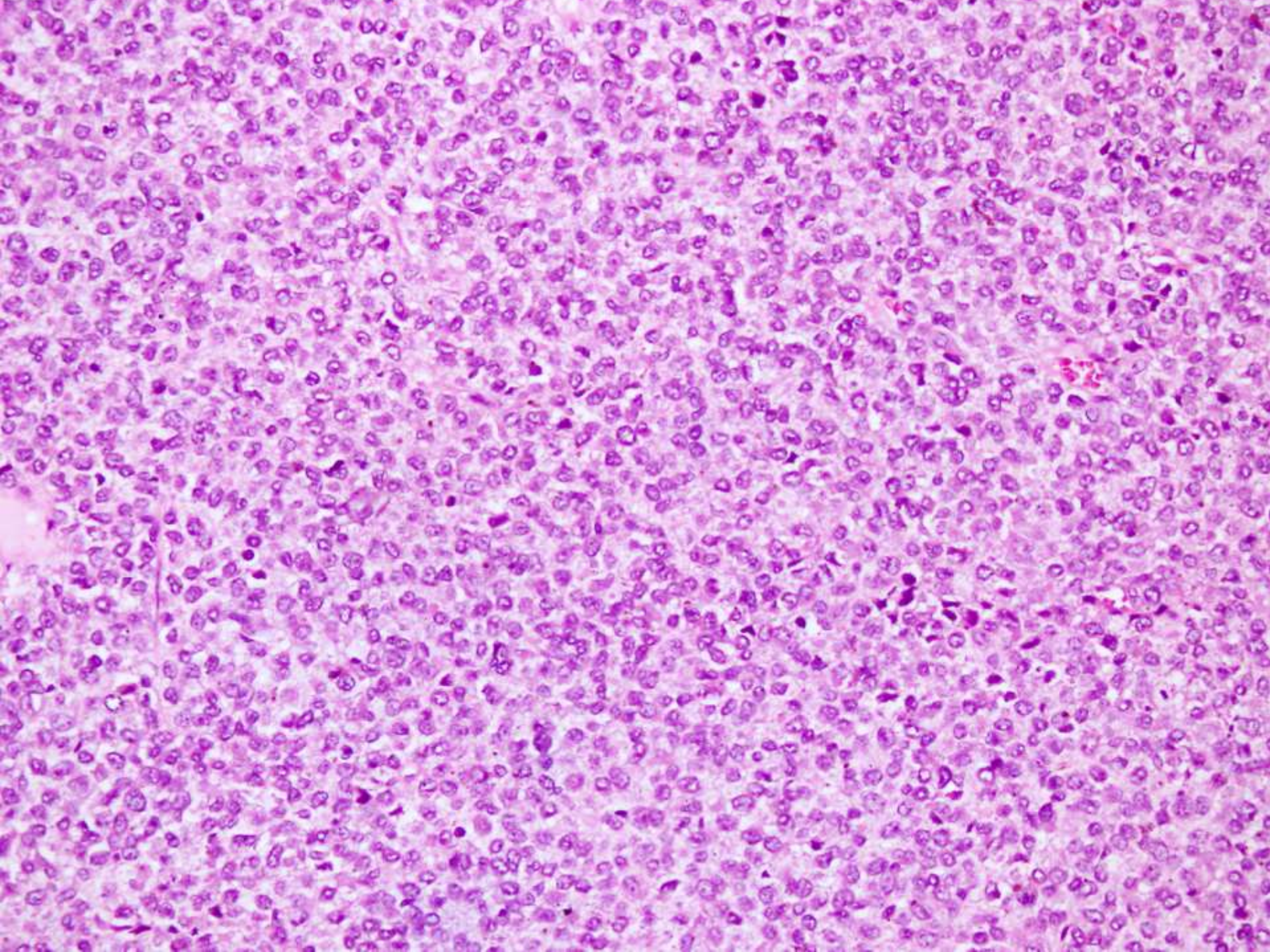
Reference: Young, RH and Clement, PB; Mullerianosis of the Urinary Bladder, Mod Path 1996 July;9(7):731-7

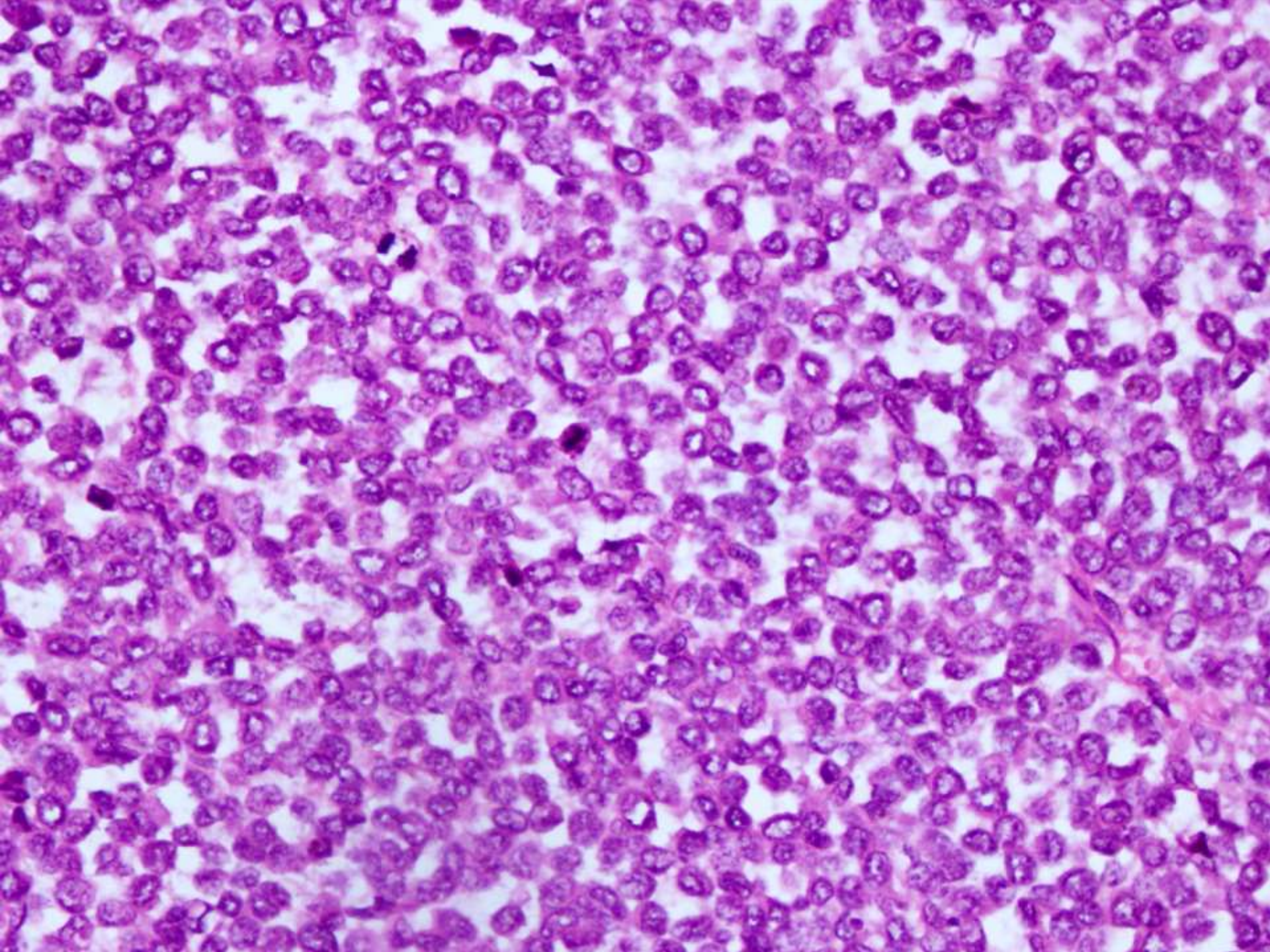
SB 6174

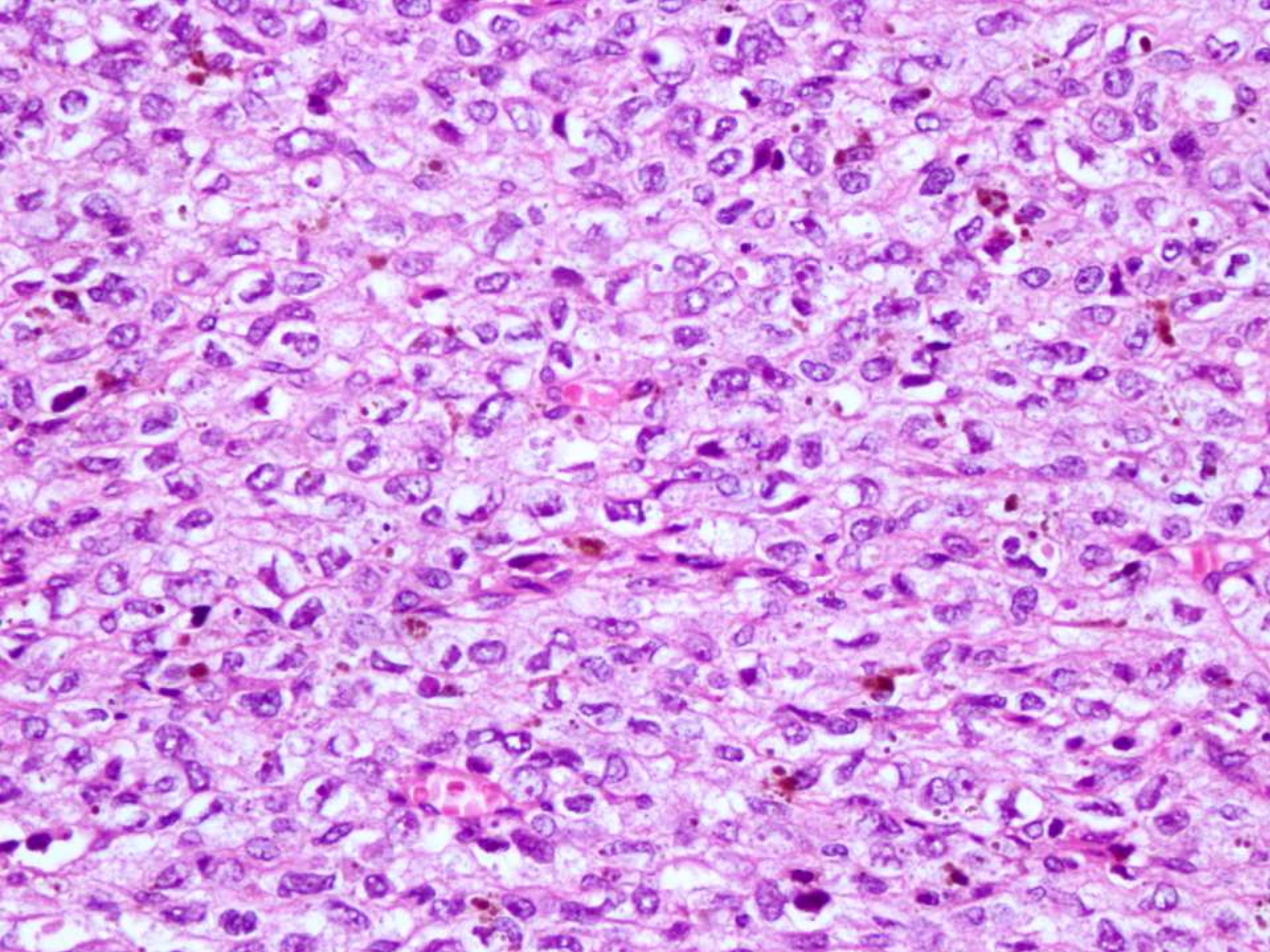
Nabeen Nayak; Sir Ganga Ram Hospital, New Dehli

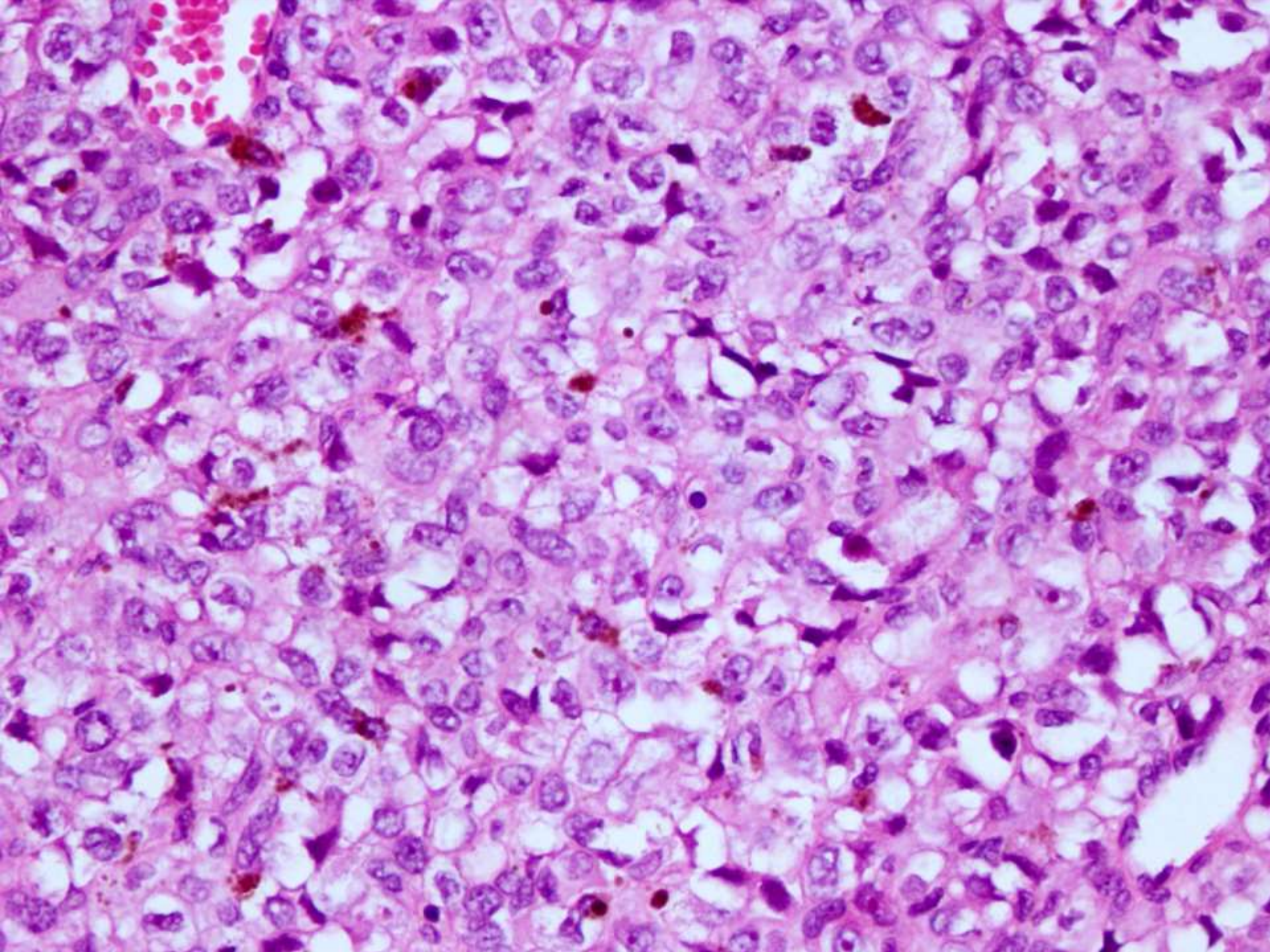
16-year-old boy with 5cm nodular mass in left side of neck deep to upper part of SCM muscle. Non-tender, no inflammation or sinus. He was euthyroid and had no significant general or systemic abnormalities.

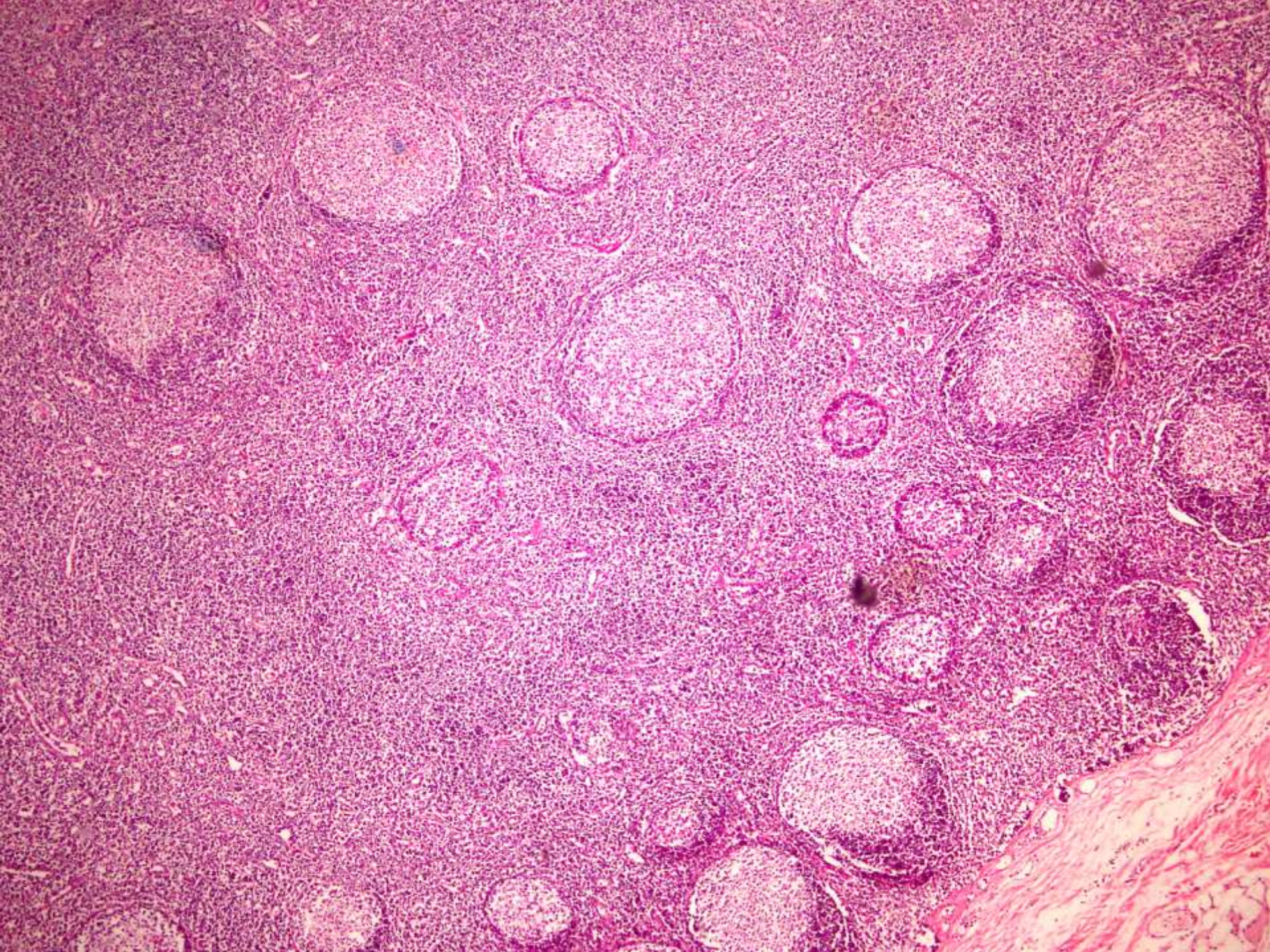




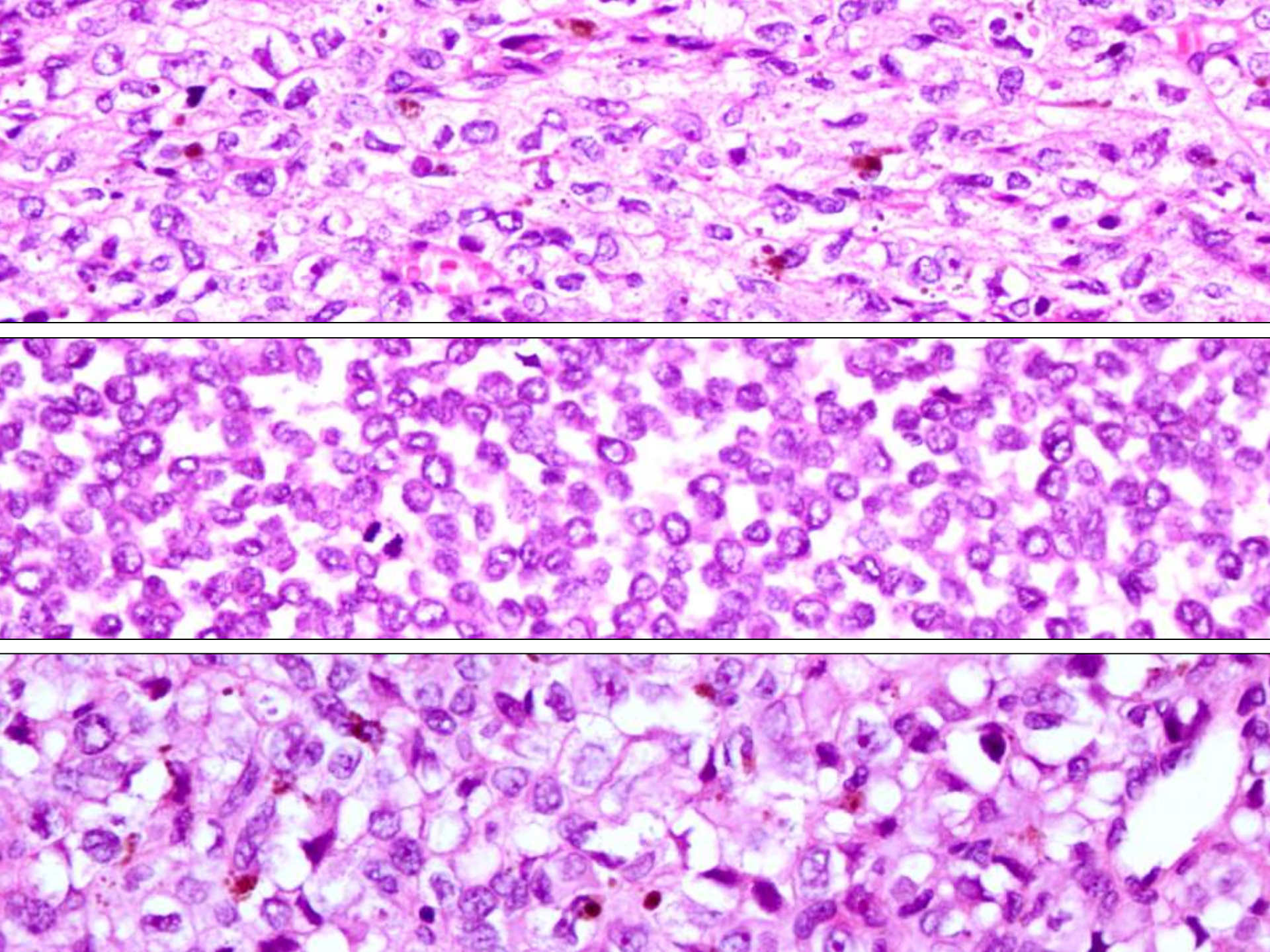


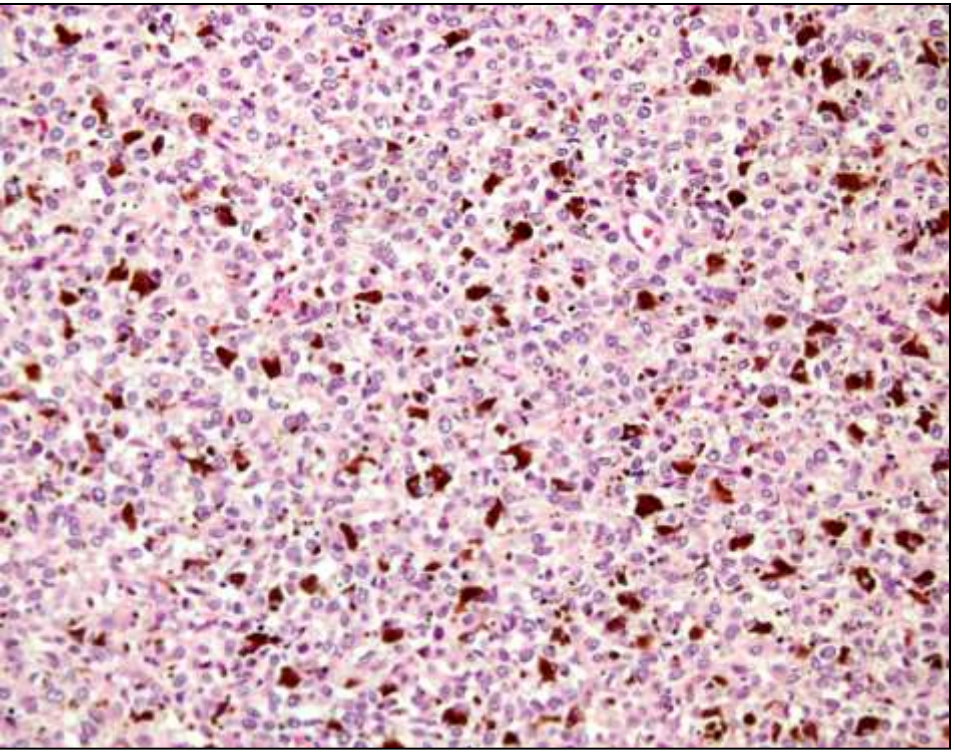


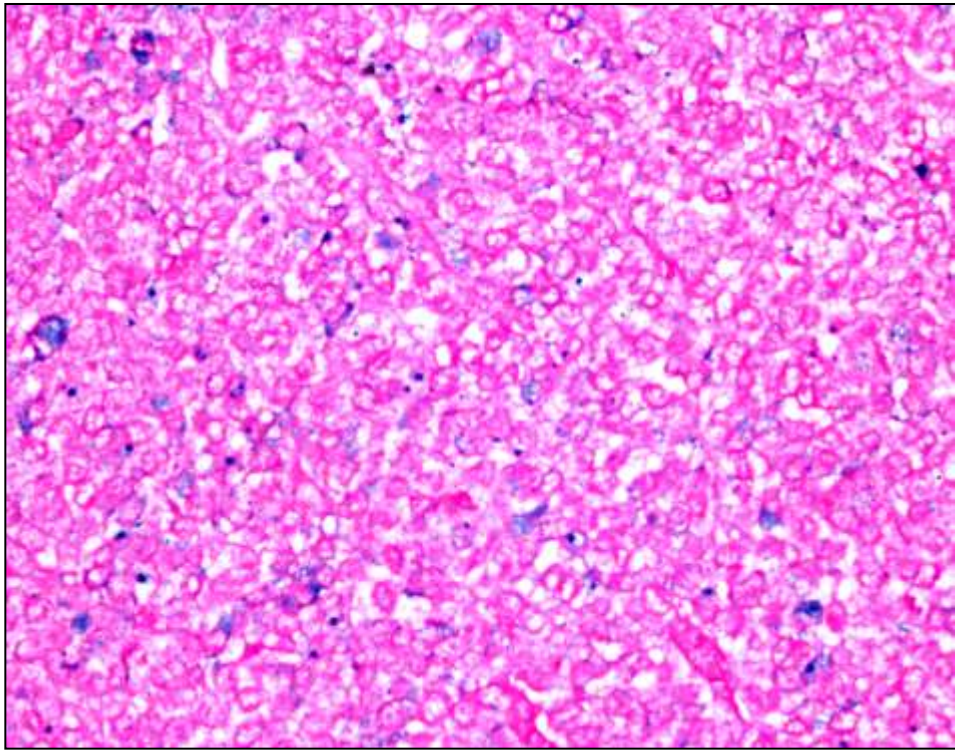




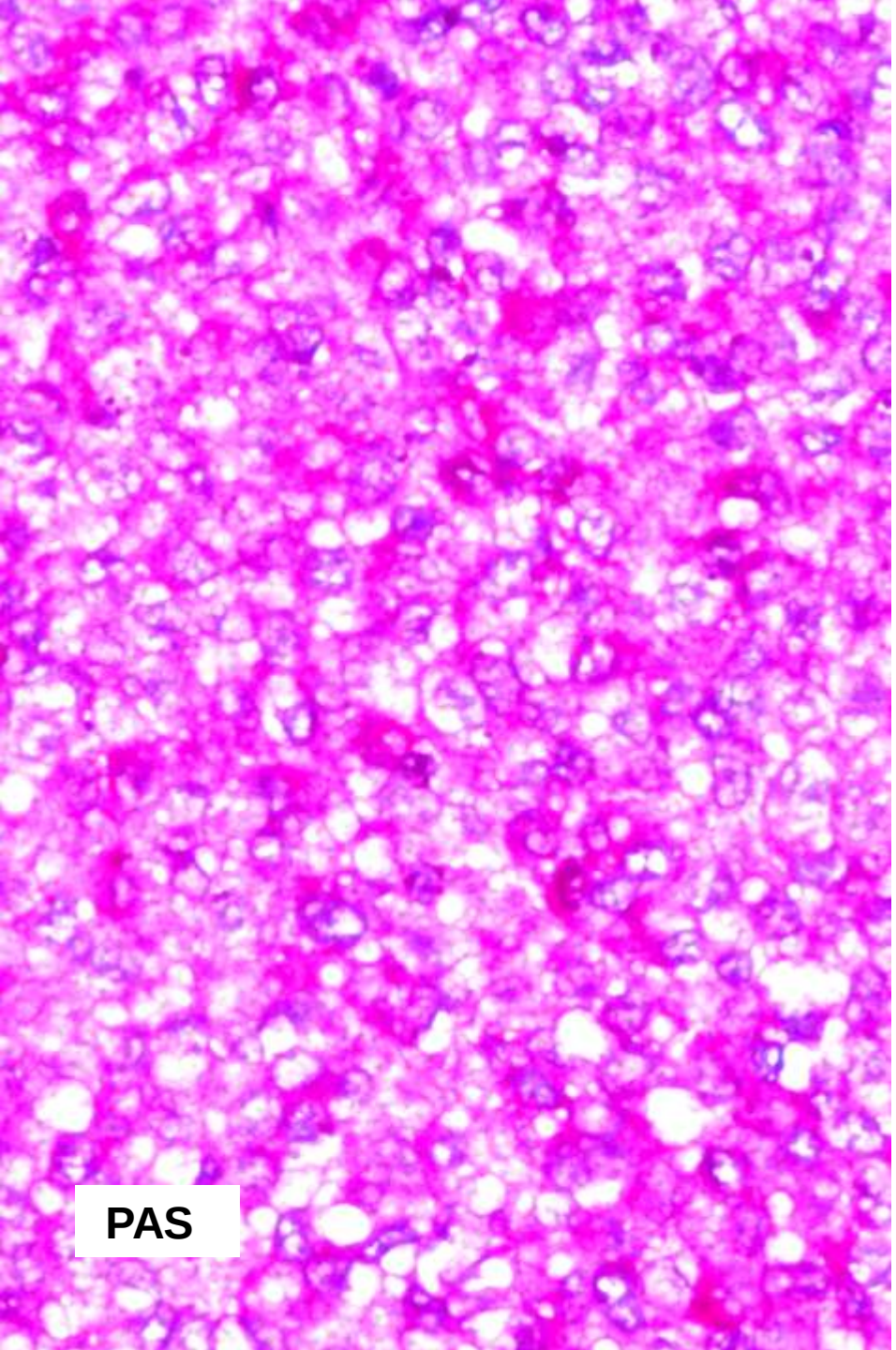
DIAGNOSIS?

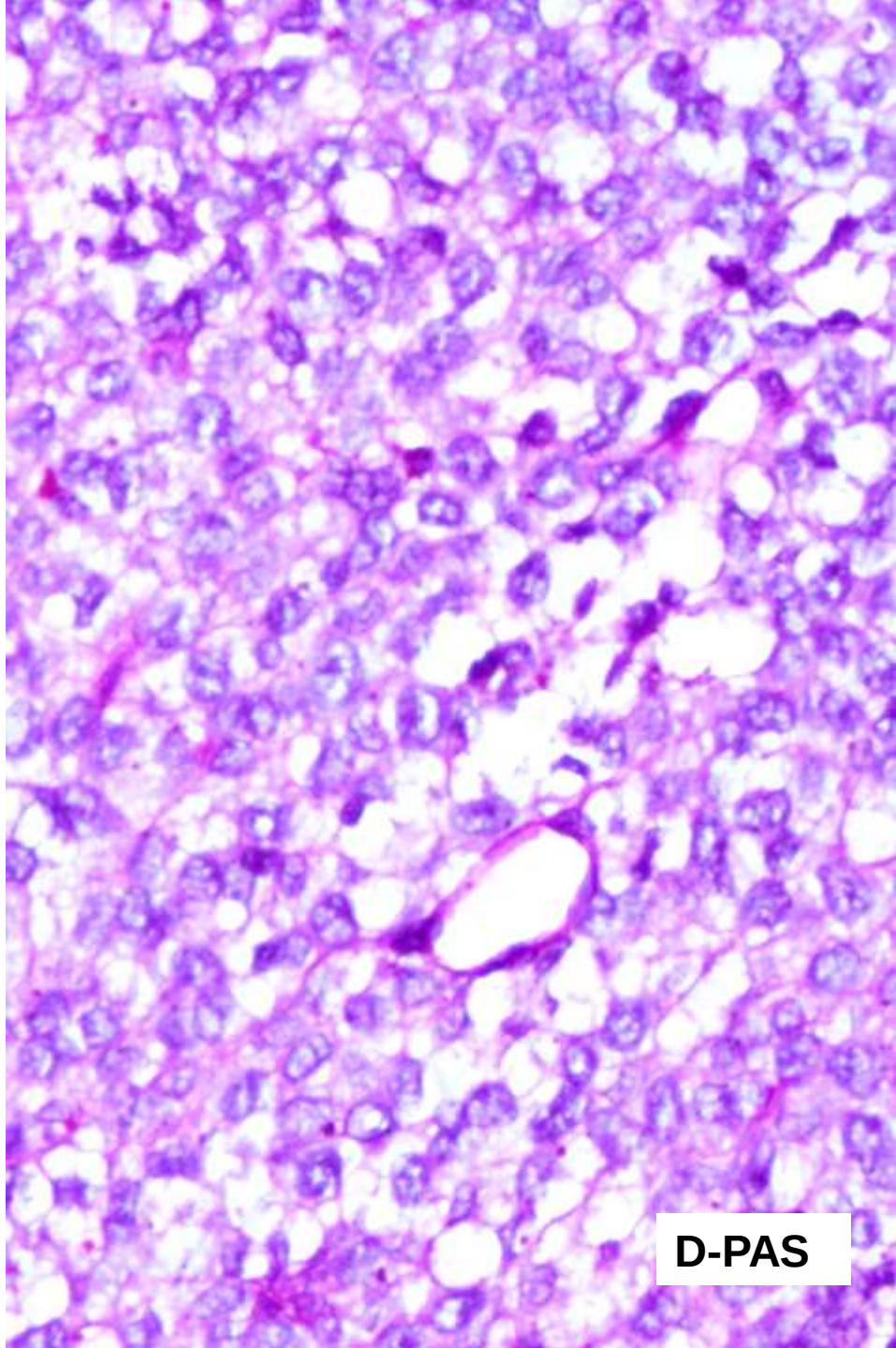






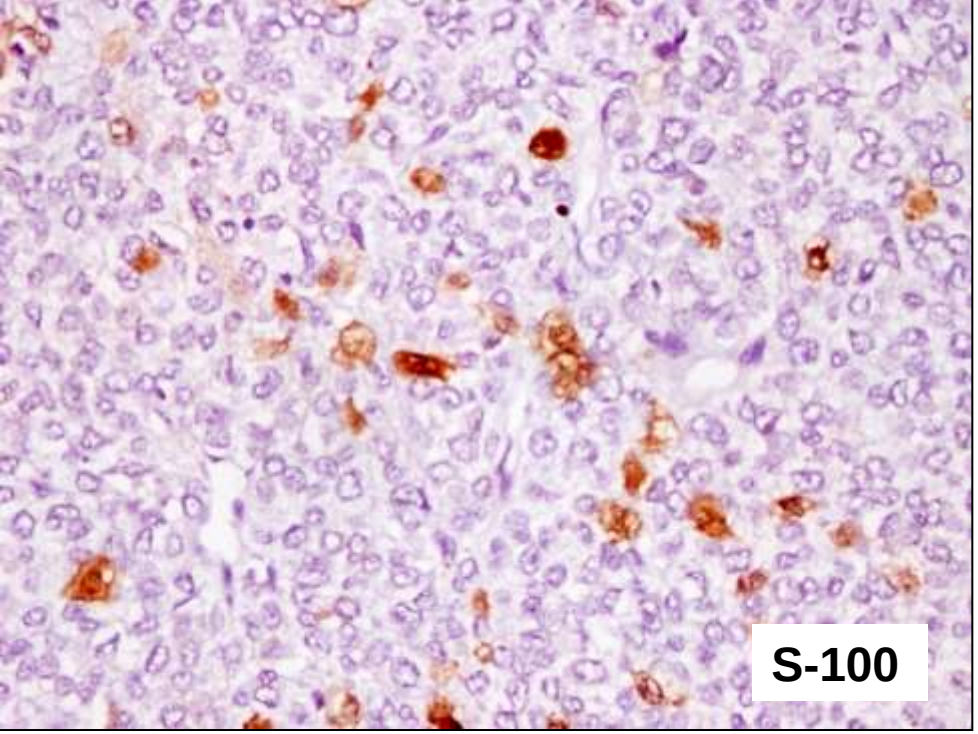
MELANIN STAINS

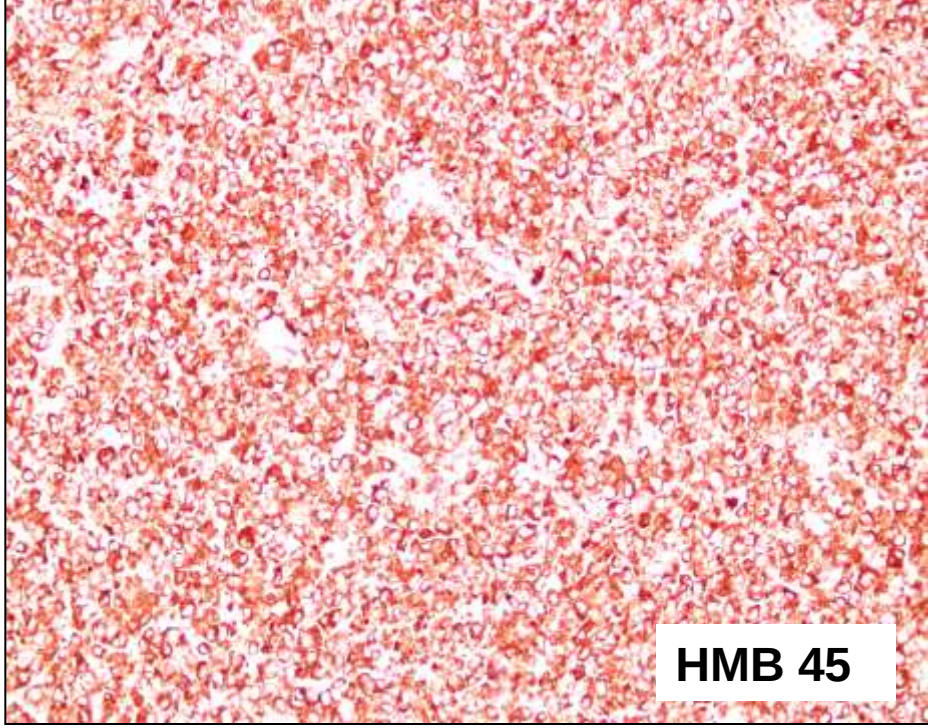

PAS

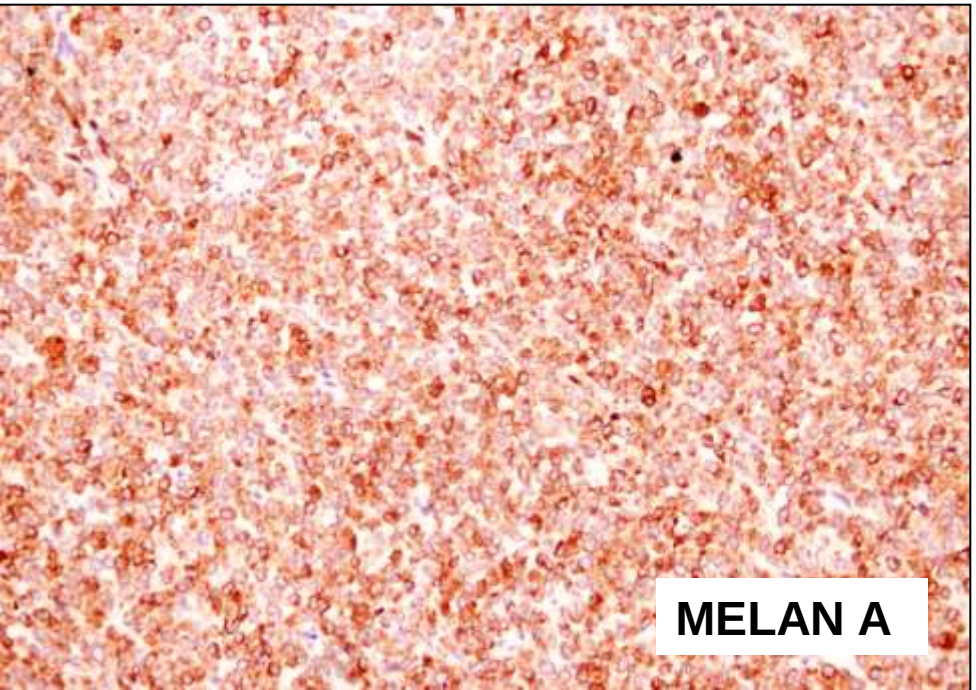

D-PAS

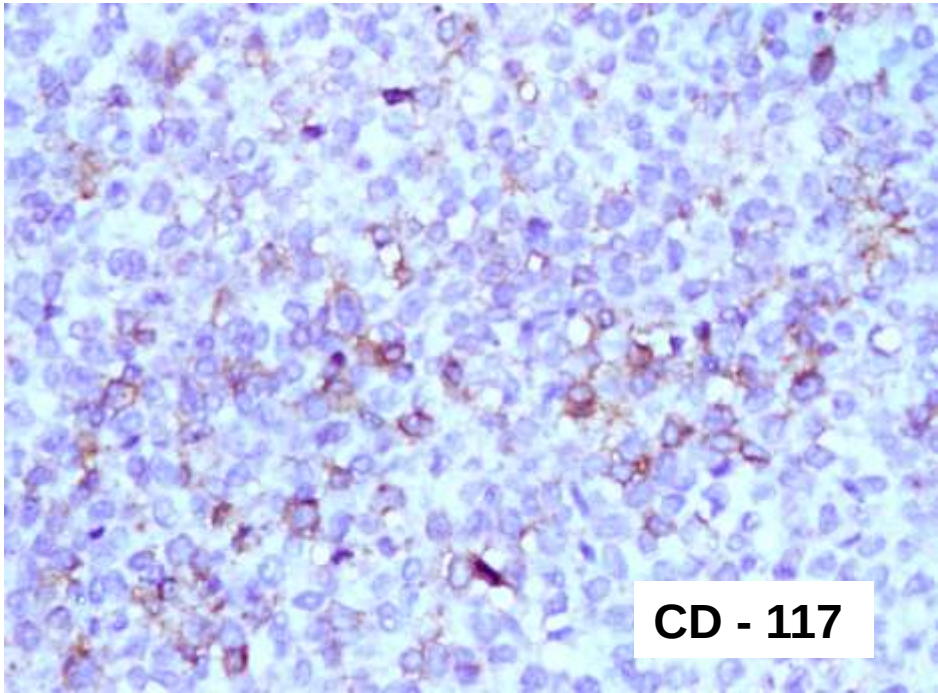
IMMUNOHISTOCHEMICAL PROFILE

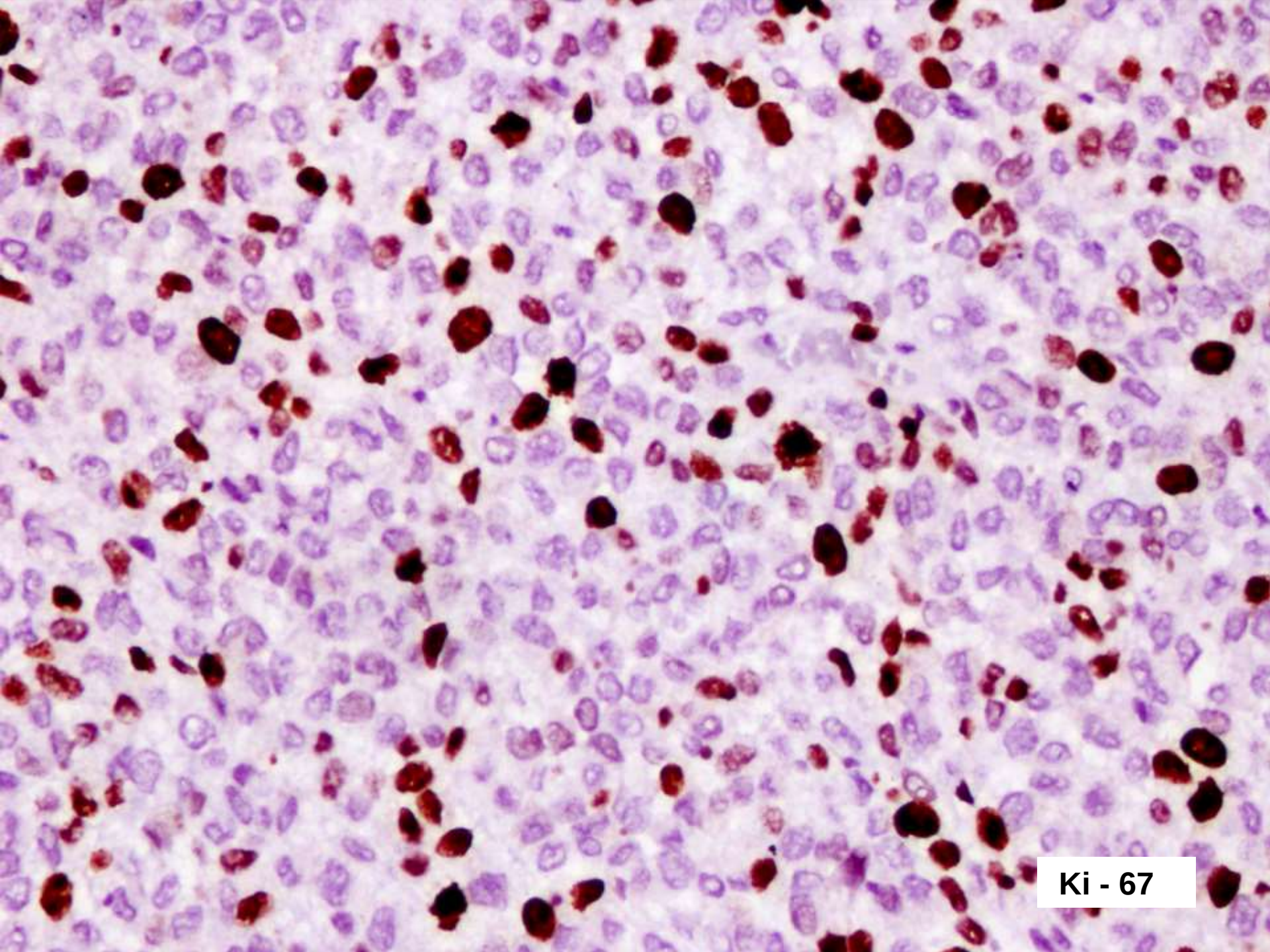
Negative Stains	**Positive Stains**
CYTOKERATIN	S –100 (10% Cells)
LCA	HMB – 45 (Diffuse)
SMA	MELAN - A (Diffuse)
SYNAPTOPHYSIN	CD – 117 (10% Cells)
CHROMOGRANIN A	Ki – 67 (25% Cells)


S-100


HMB 45


MELAN A


CD - 117


Ki - 67

DIAGNOSIS: Clear Cell Sarcoma of soft tissues, neck (Favored)

- Young age of patient
- Location: soft tissue cervical fascia, neck (though rare for CCS)
- No skin lesion anywhere, nor any other lesion in the body
- Variable cellular morphology with cytoplasmic glycogen
- Melanocytic differentiation & IHC features compatible with CCS
  (even though CD-117 if focally +ve – seen in about 15% cases)
- Lymph nodes are free of tumor

Later a FISH test done on FFPE block was negative for

EWS/ATF1 fusion translocation – t(12;22)(q13;q12)

This genetic mutation is reported +ve in 75-90% cases of CCS

Follow up;

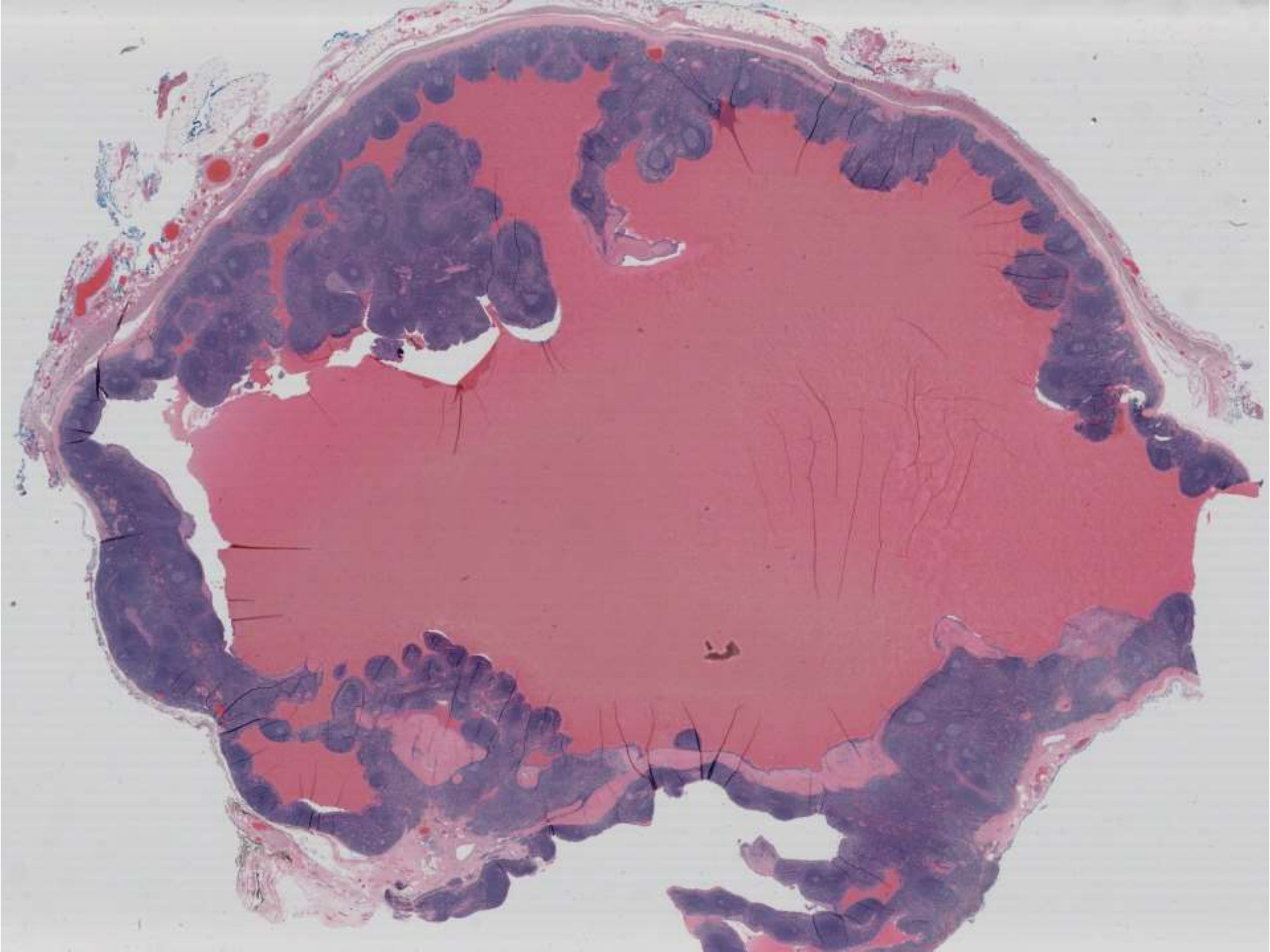
Patient had RT after wide resection of tumor which measured 4.6x4x3 cm.

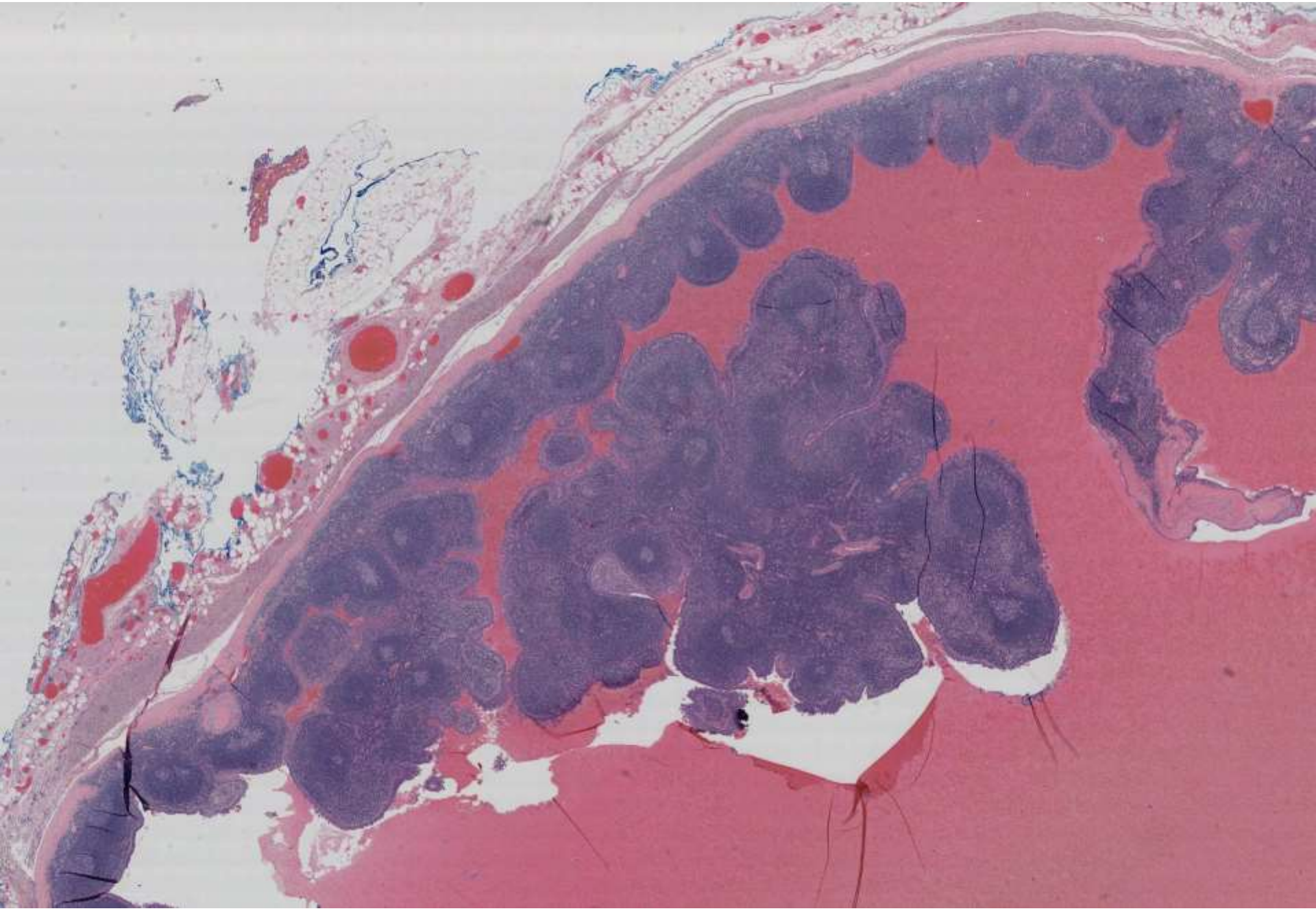
Only 7 months post-op now he is doing well with no local recurrence or any other lesion in the body. PET scan is negative

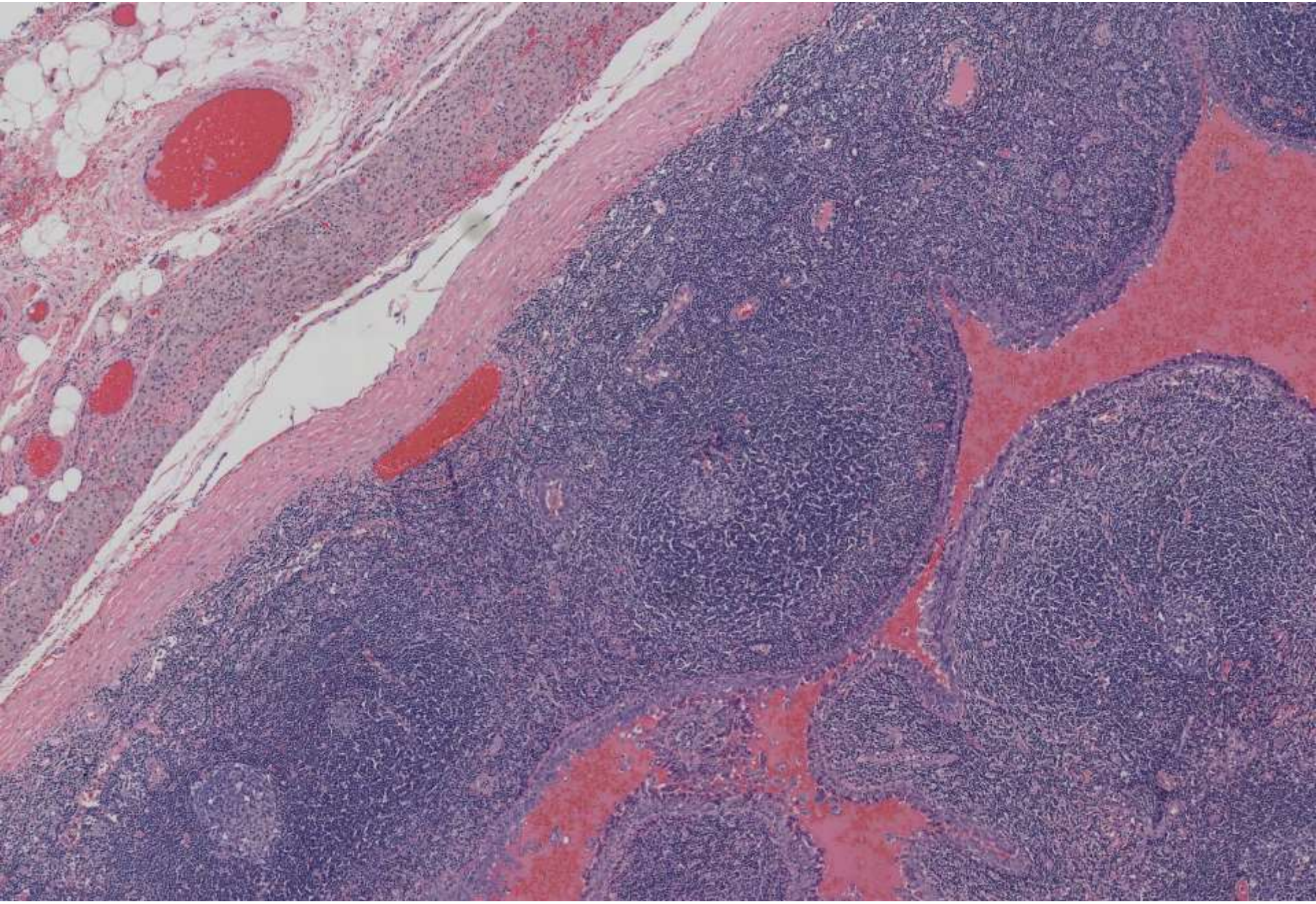
SB 6175
(scanned slide available)

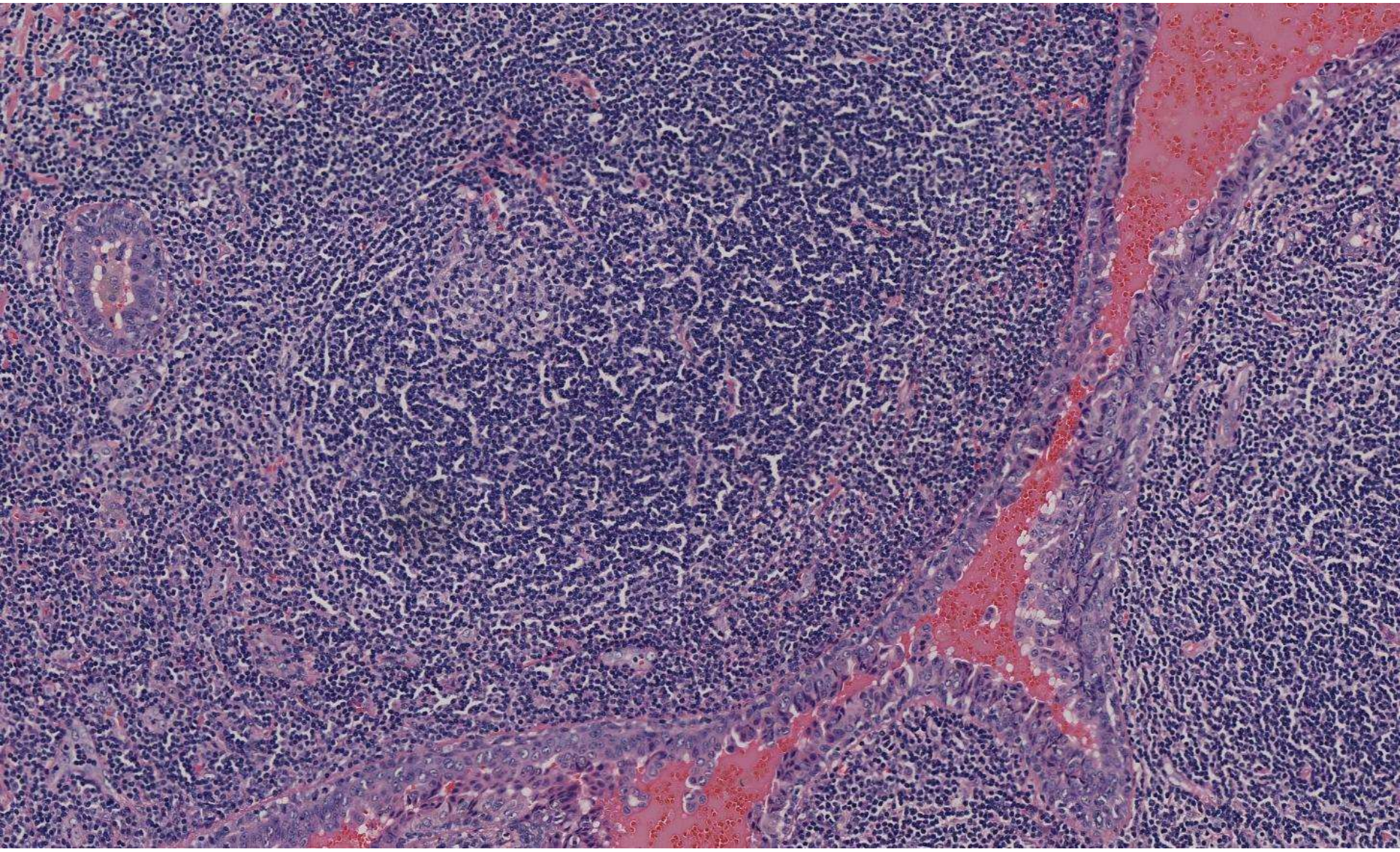
Erna Forgo/Christian Kunder; Stanford

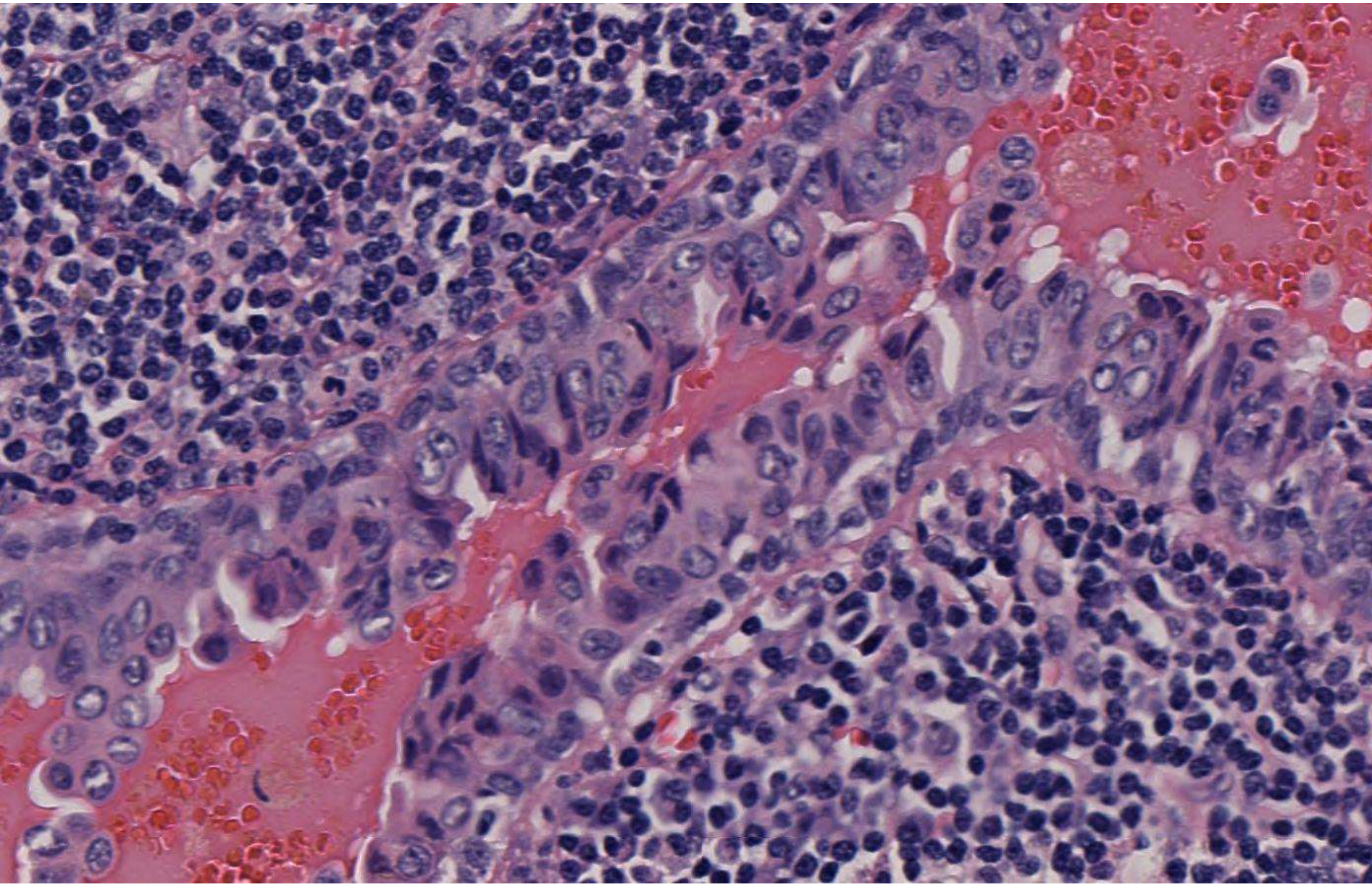
52-year-old man with neck mass for 2 years,
with slow increase in size recently.
Specimen: left superficial parotid gland.

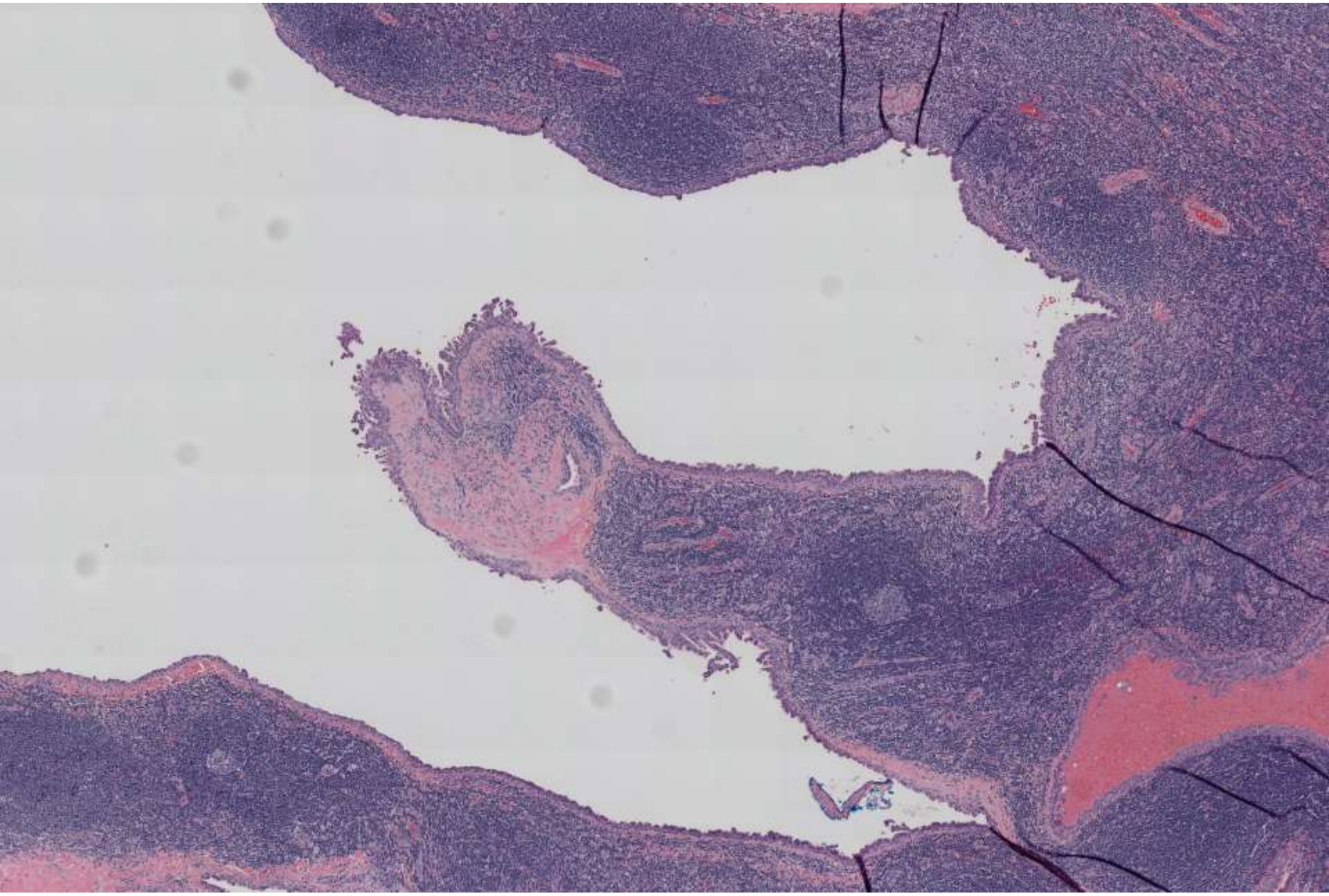


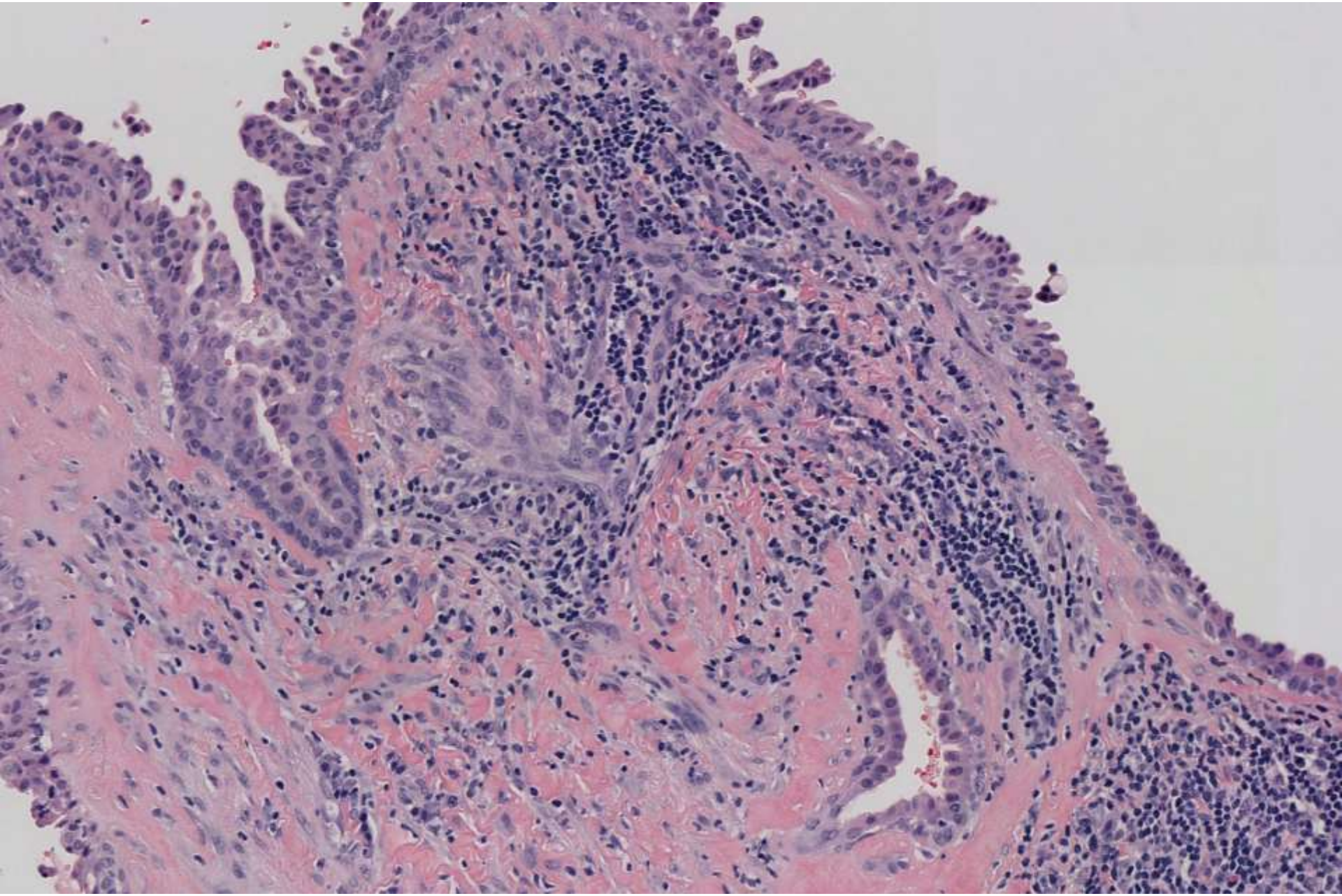


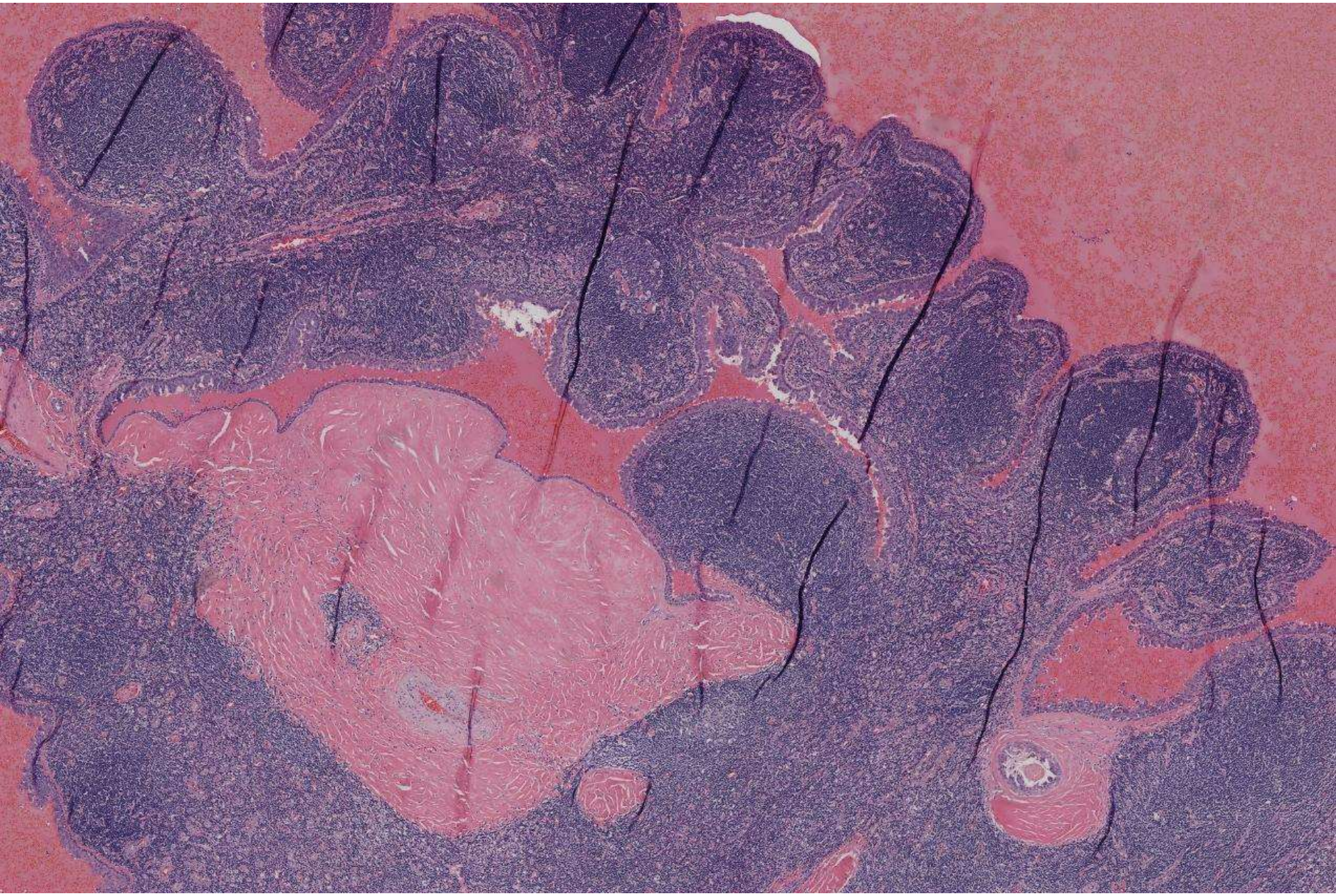


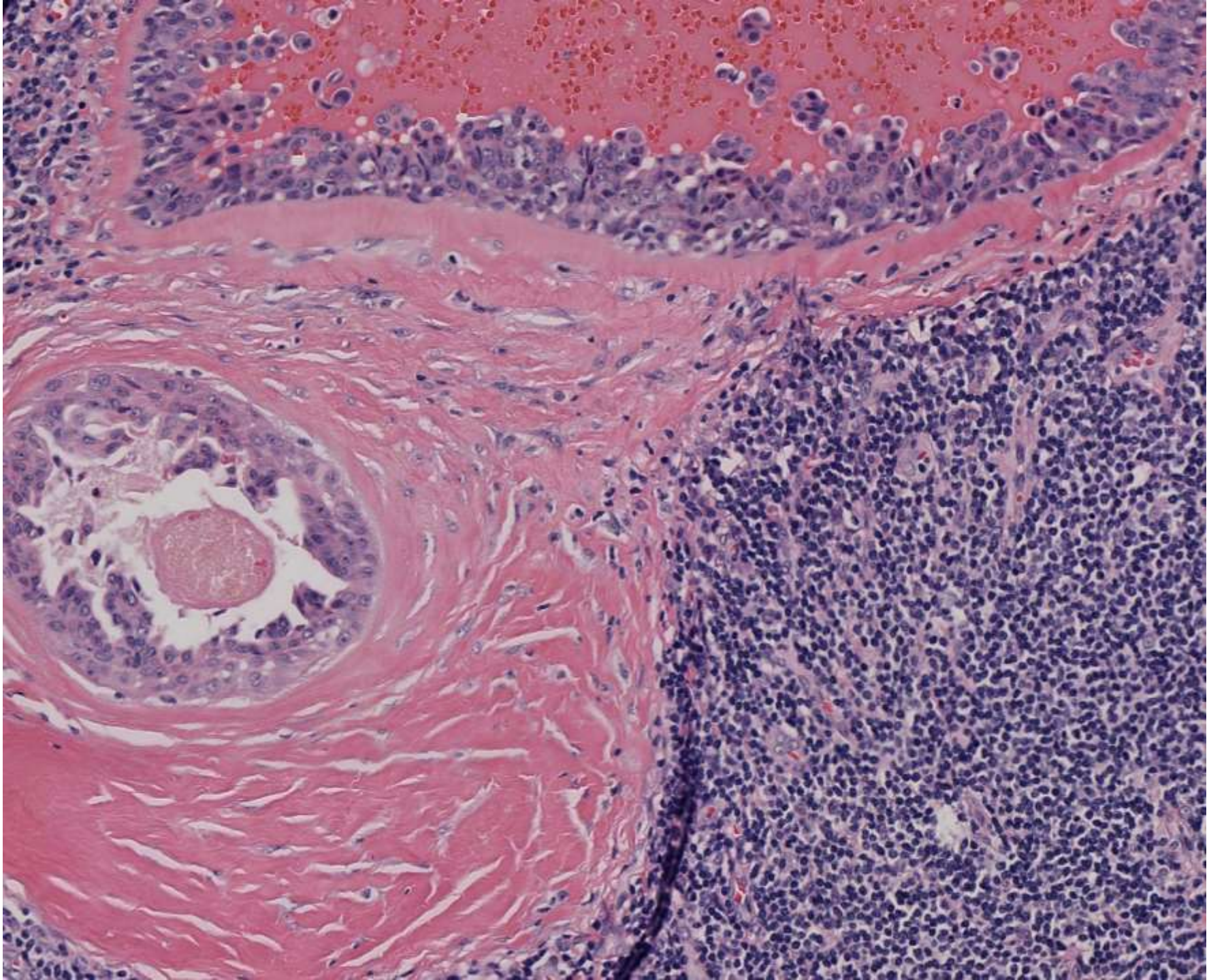












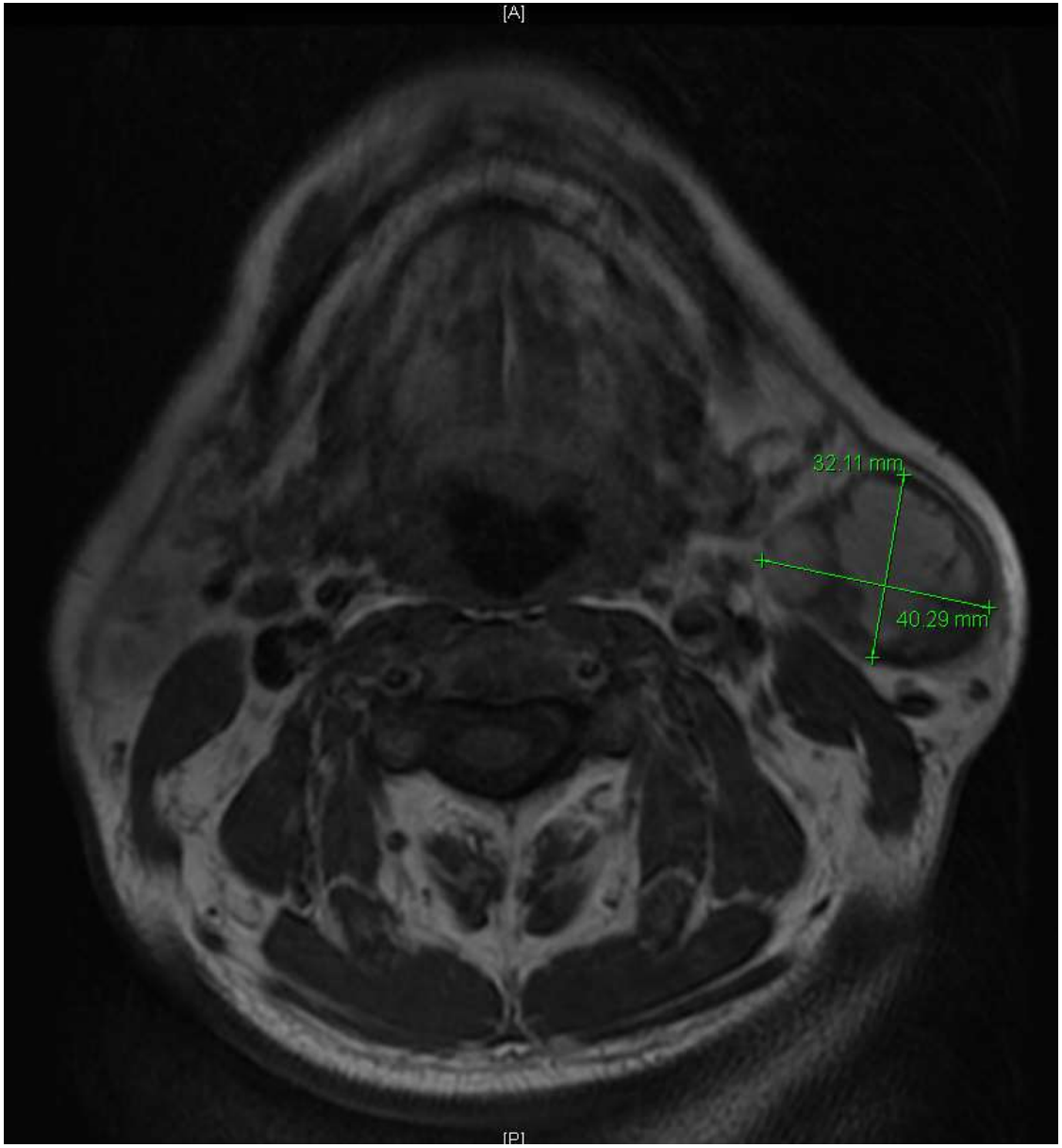
DIAGNOSIS?

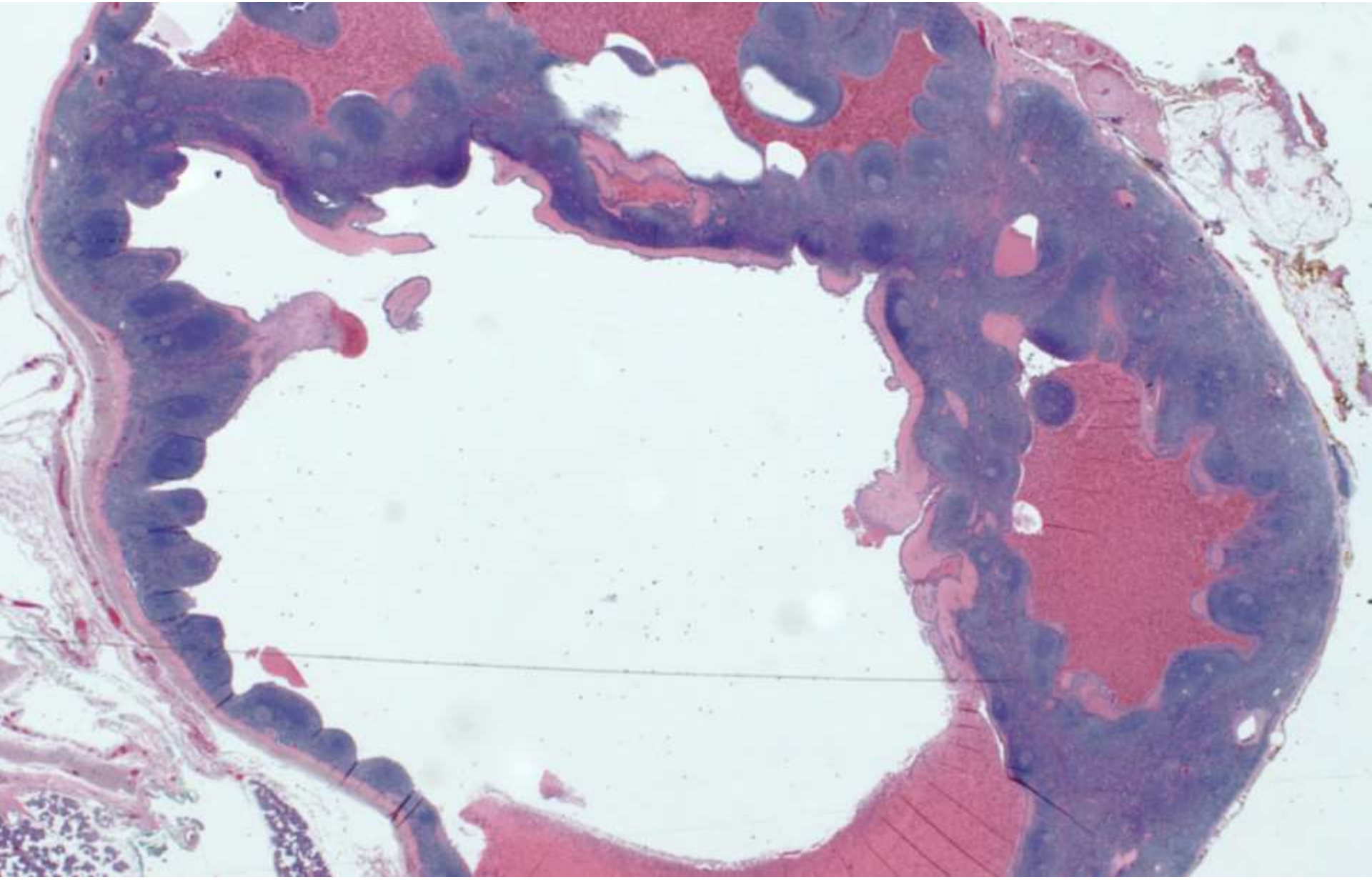
52 year-old male with a neck mass for 2 years, with slow increase in size recently

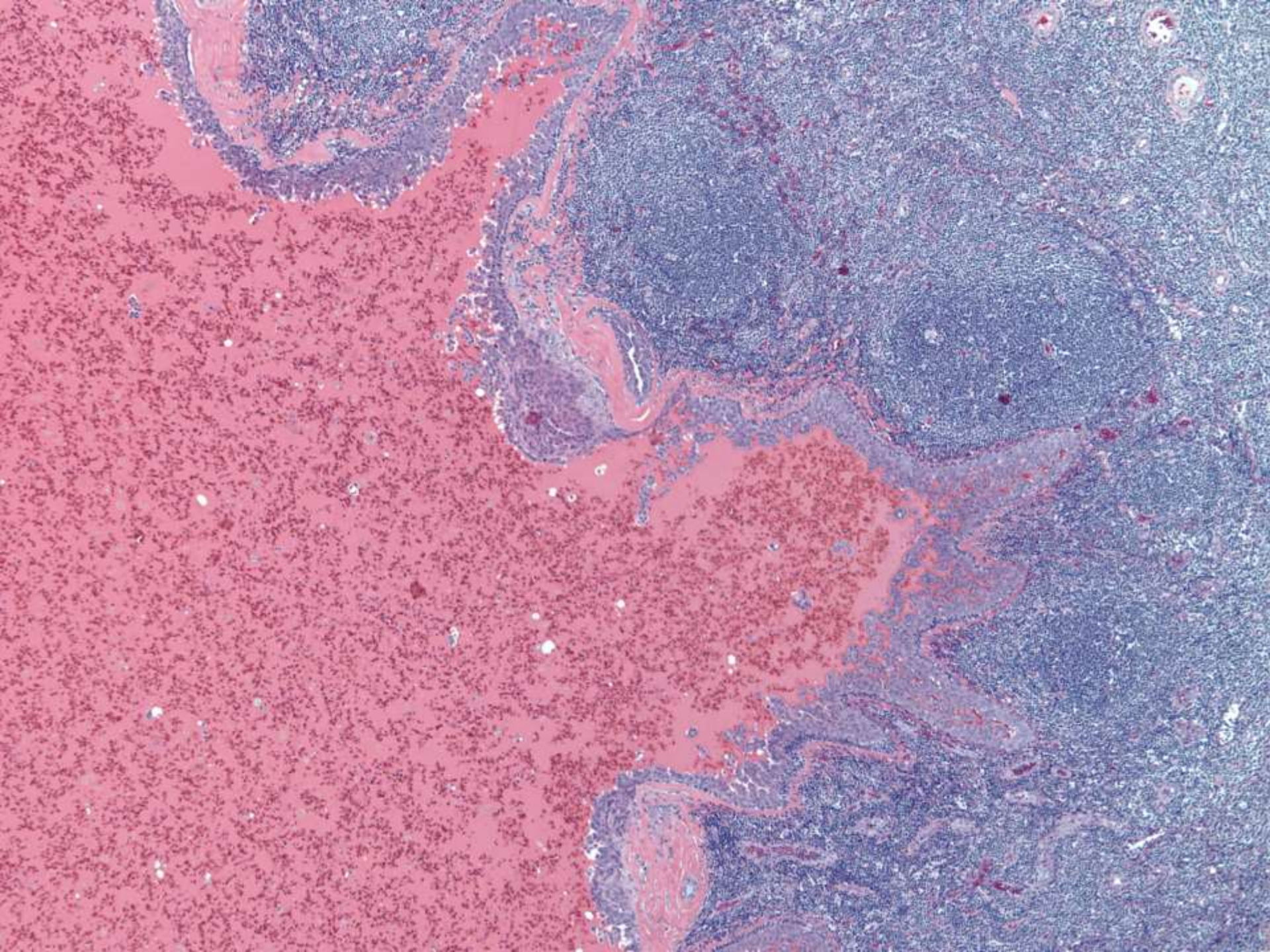
Erna Forgó/Christian Kunder

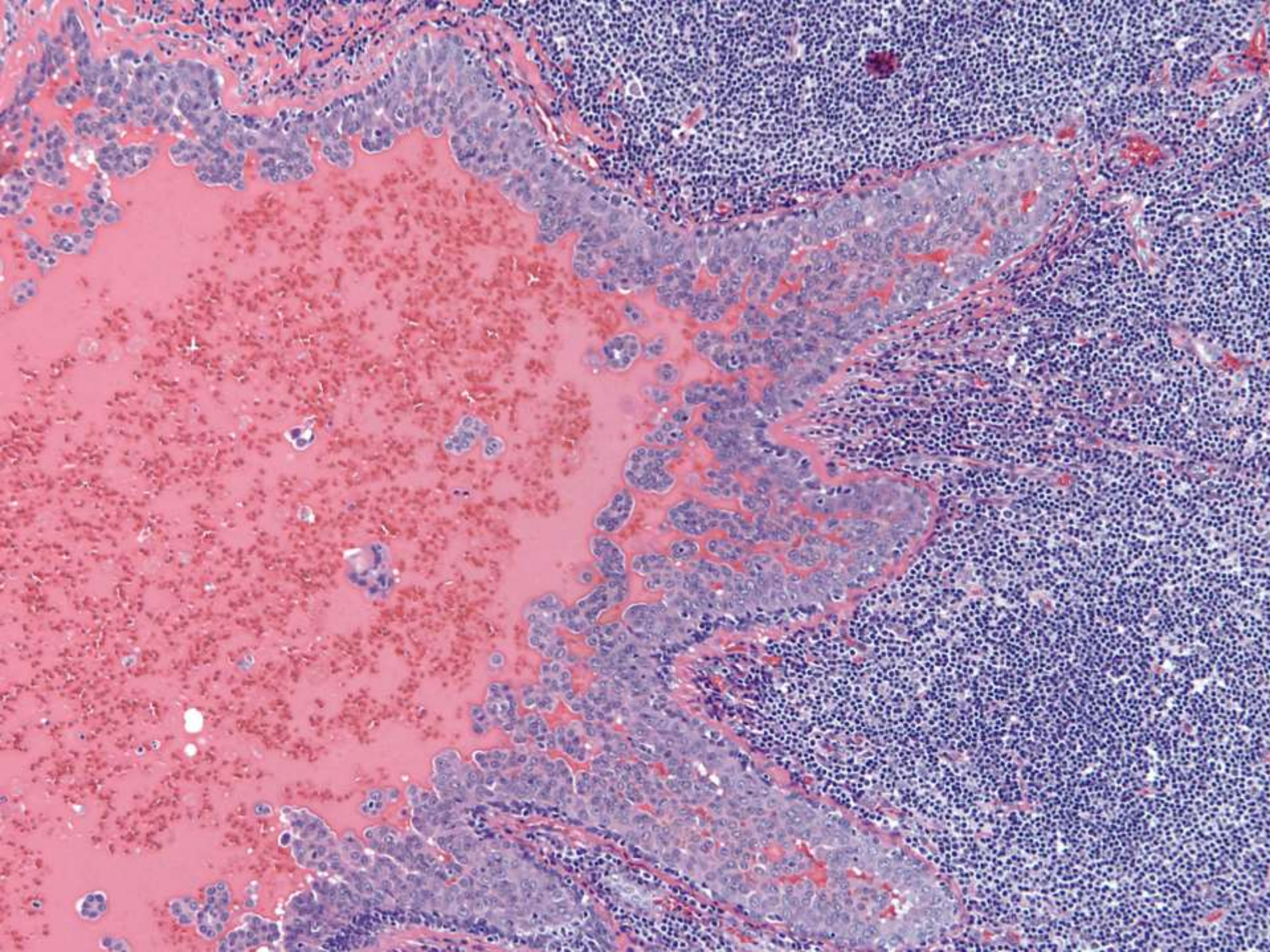
7/10/2017

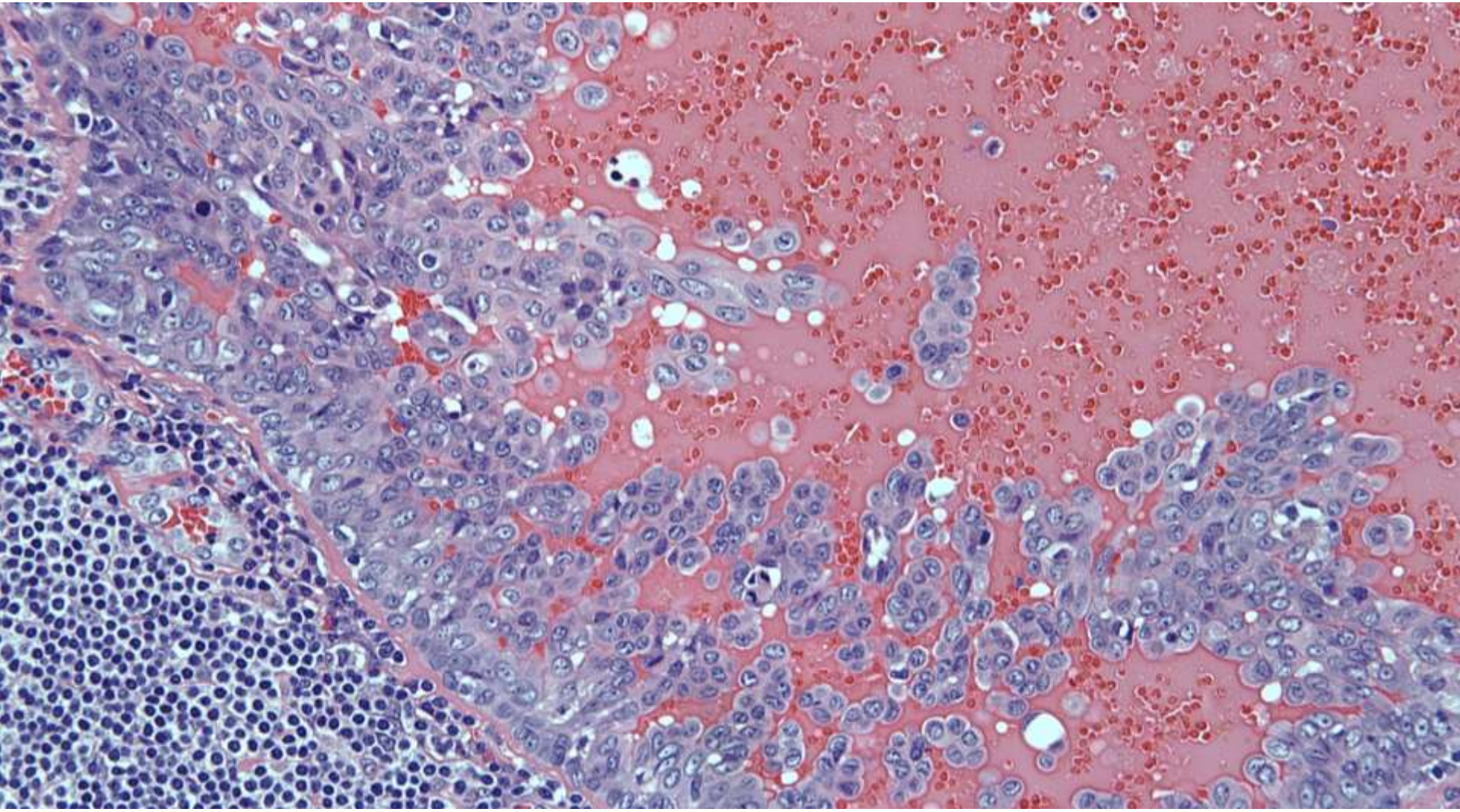
South Bay

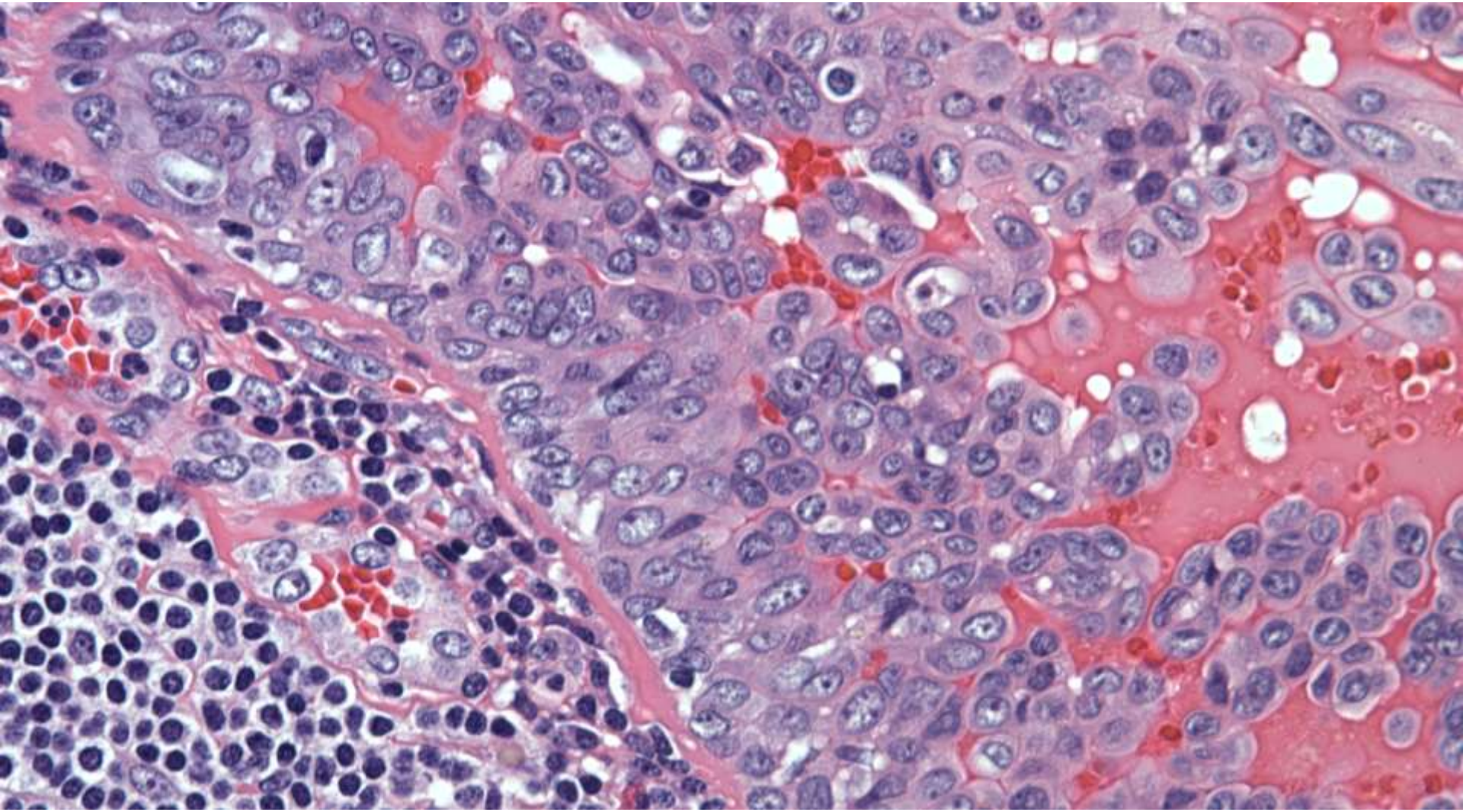

[A]
32.11 mm
40.29 mm
[P]











Staining patterns

- **PASd**: (-) for zymogen granules in the lining cells
- **SOX10**: (+) in both the lining cells and the basal layer
- **DOG1**: (+) in luminal cells, in some areas in a canalicular pattern (normal salivary acini and in acinic cell carcinoma)
- **CD20 and CD3**: highlight B cells and T cells within the germinal centers, consistent with a reactive lymphoid process

Dr. Stacey Mills at UVA

- “The parotid mass bears a vague resemblance to a Warthin tumor, with a cystic epithelial component in a lymphoid background, however in this case the epithelium is not overtly oncocytic. We would best classify this as a cystic lymphadenoma which is related to Warthin tumor in the sense that it arises in a salivary gland rest entrapped in a periparotid lymph node.”

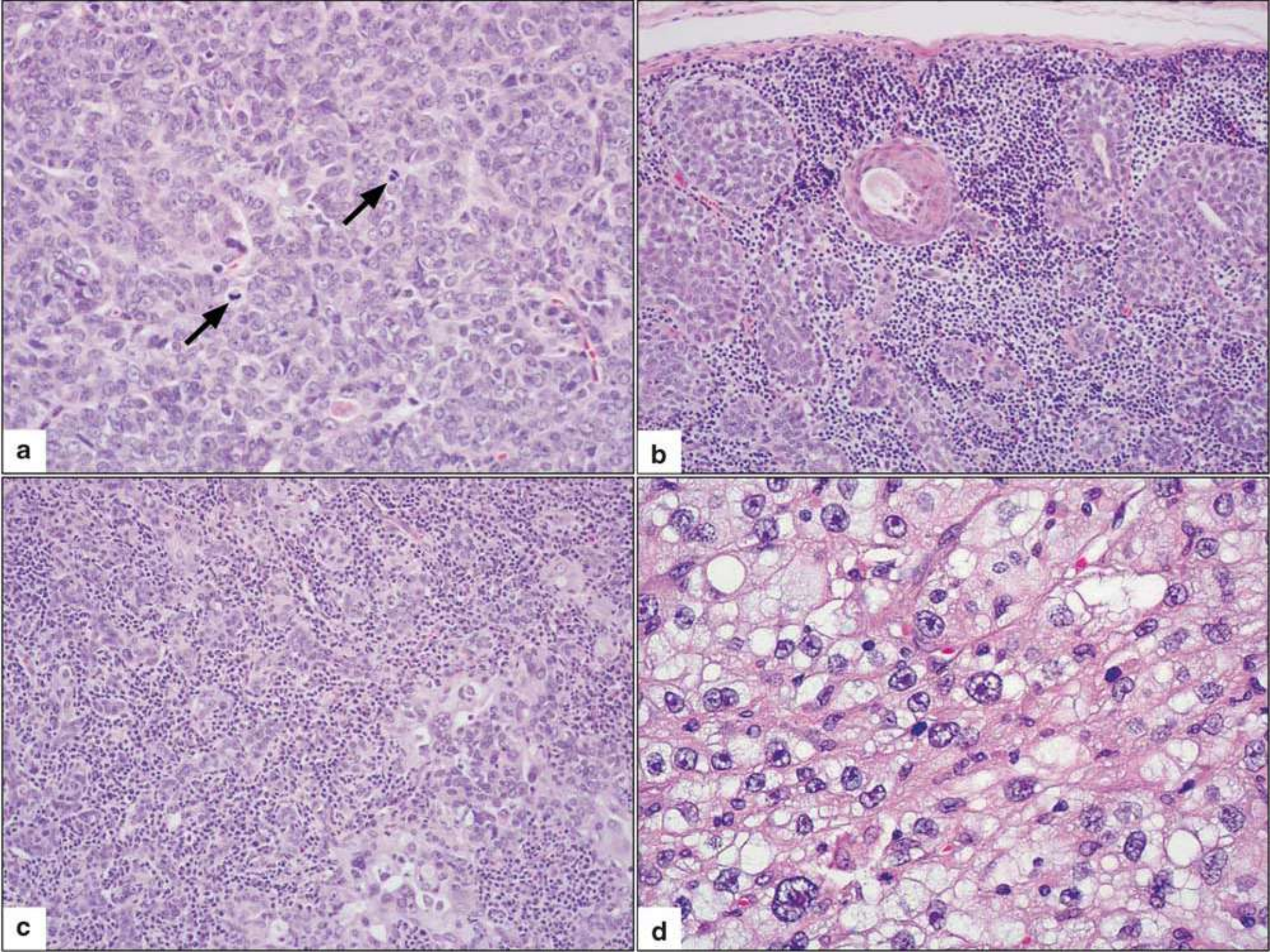
Cystic Lymphadenoma

- Rare (<0.2%) cystic salivary gland neoplasm composed of epithelial nests and cysts distributed within dense hyperplastic lymphoid tissue
- Well circumscribed, typically encapsulated
- Pushing, yet non-infiltrative border
- Mean: 6th-8th decades, range: 11-79 years
- Equal M:F distribution
- Treatment: Conservative surgical excision
- Rare malignant transformation reported

Malignant Transformation

- 33 cases diagnosed at 10 different institutions
  - 22 sebaceous and 11 non-sebaceous
- Two sebaceous lymphadenomas showed malignant transformation:
  - 74 year-old woman developed basal cell adenocarcinoma
  - 76 year-old man developed sebaceous carcinoma
- Malignancy was based on mitoses and increased cellularity in the basal cell adenocarcinoma where the tumor cells remained monomorphic
- In the sebaceous carcinoma, there was significant cytologic atypia, nuclei were large and vesicular with prominent nucleoli, and foamy cytoplasm

Modern Pathology (2012) 25, 26–35

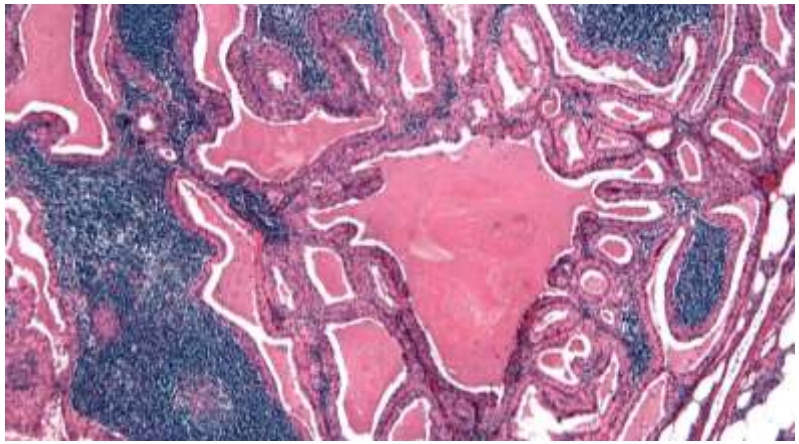

a
b
c
d

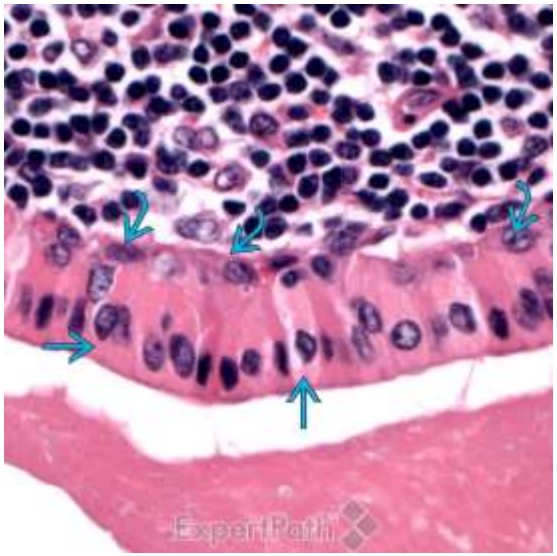
Differential Diagnosis

- Warthin tumor
- Acinic cell carcinoma
- Mammary analogue secretory carcinoma

Warthin Tumor

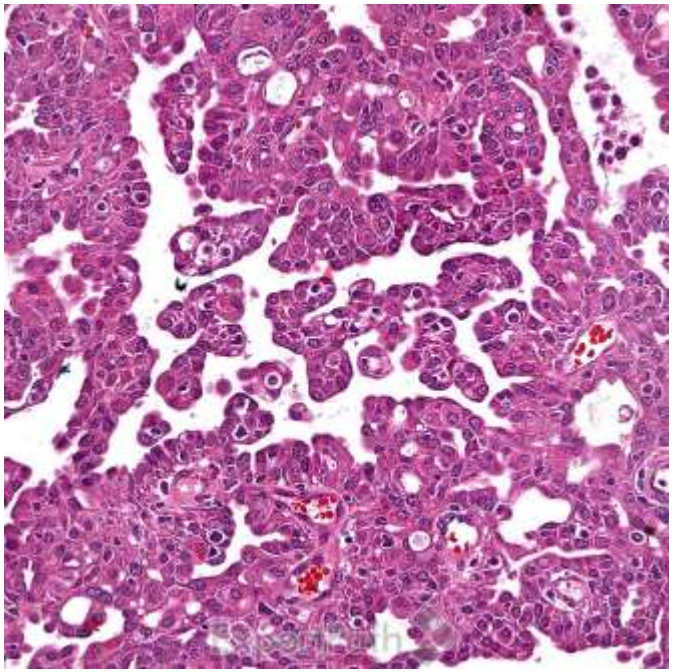
- Papillary projections into cystic spaces surrounded by lymphoid stroma
- Epithelium: double cell layer: 1. luminal cells and 2. basal cells
- Stroma: mature lymphoid follicles with germinal centers
- SOX10 negative





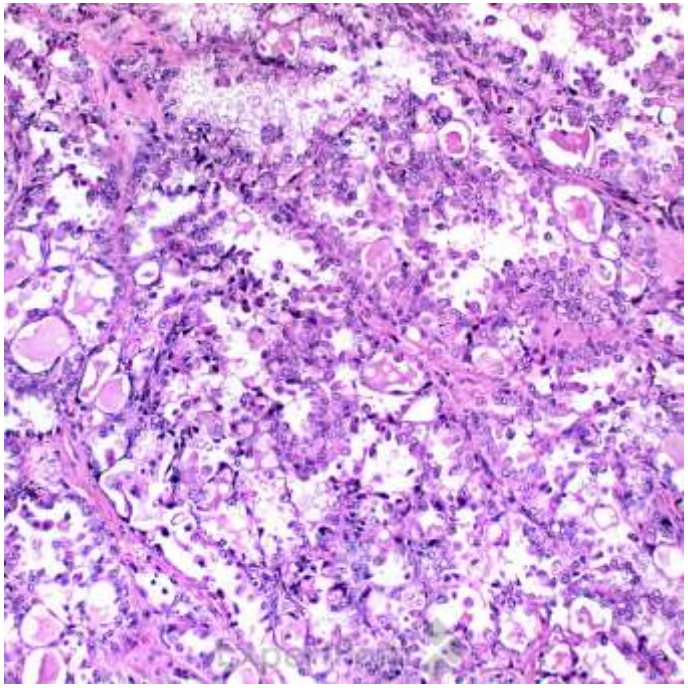
Acinic Cell Carcinoma

- Malignant epithelial salivary gland neoplasm of mixture of cell types:
  - serous acinar, intercalated duct-like, hobnail, vacuolated, clear, and nonspecific glandular cells
- Cytoplasmic zymogen secretory granules PAS(+), diastase-resistant
- DOG1 positive



Mammary Analogue Secretory Carcinoma (MASC)

- Uniform cells with vacuolated cytoplasm, microcystic to cystic and papillary architecture, intraluminal secretions, and scalloping
- Absence of basophilic granules
- Mammaglobin (+), S100 (+), CK7 (+)
- *ETV6* translocation (+)



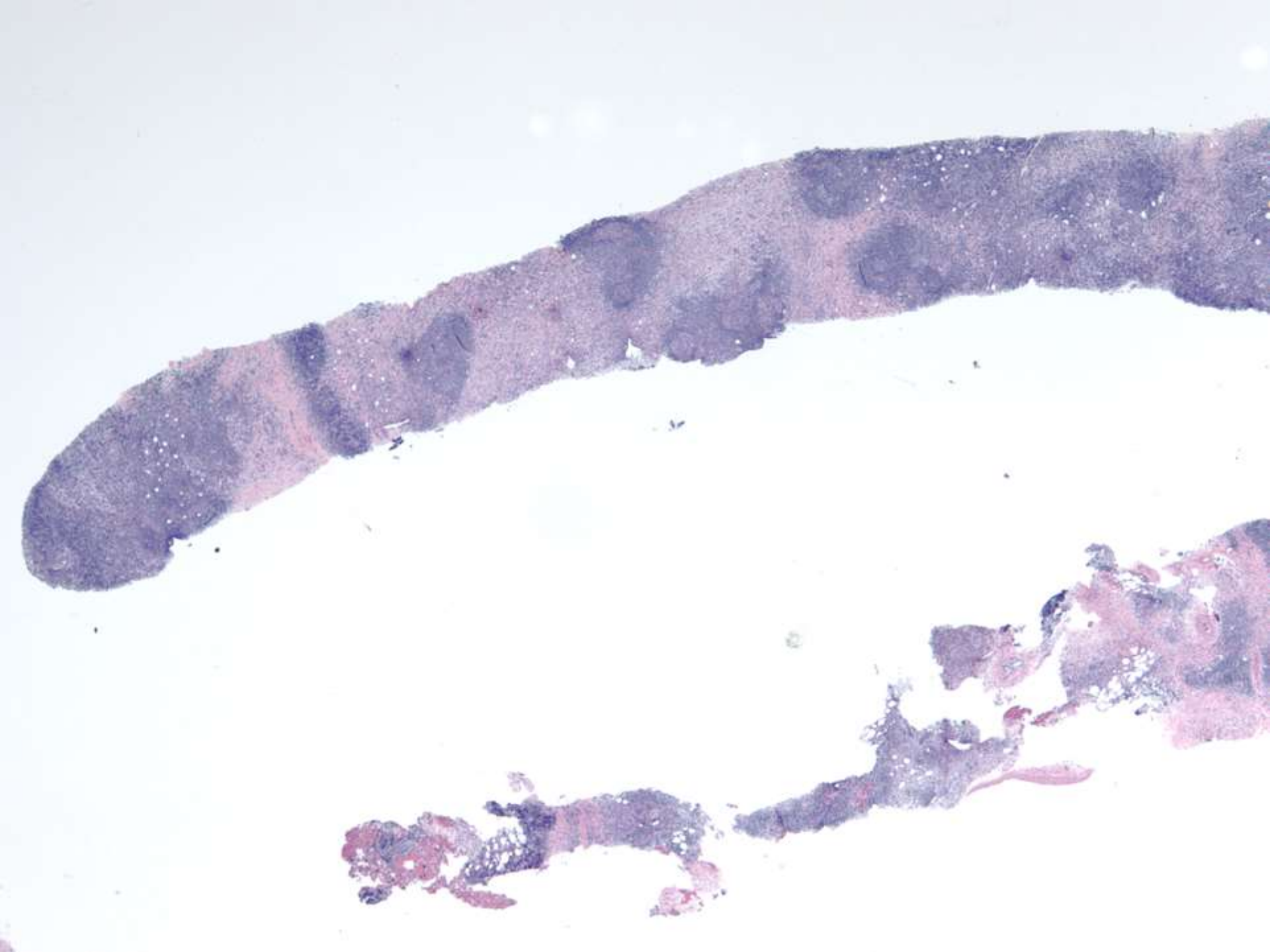
Follow Up

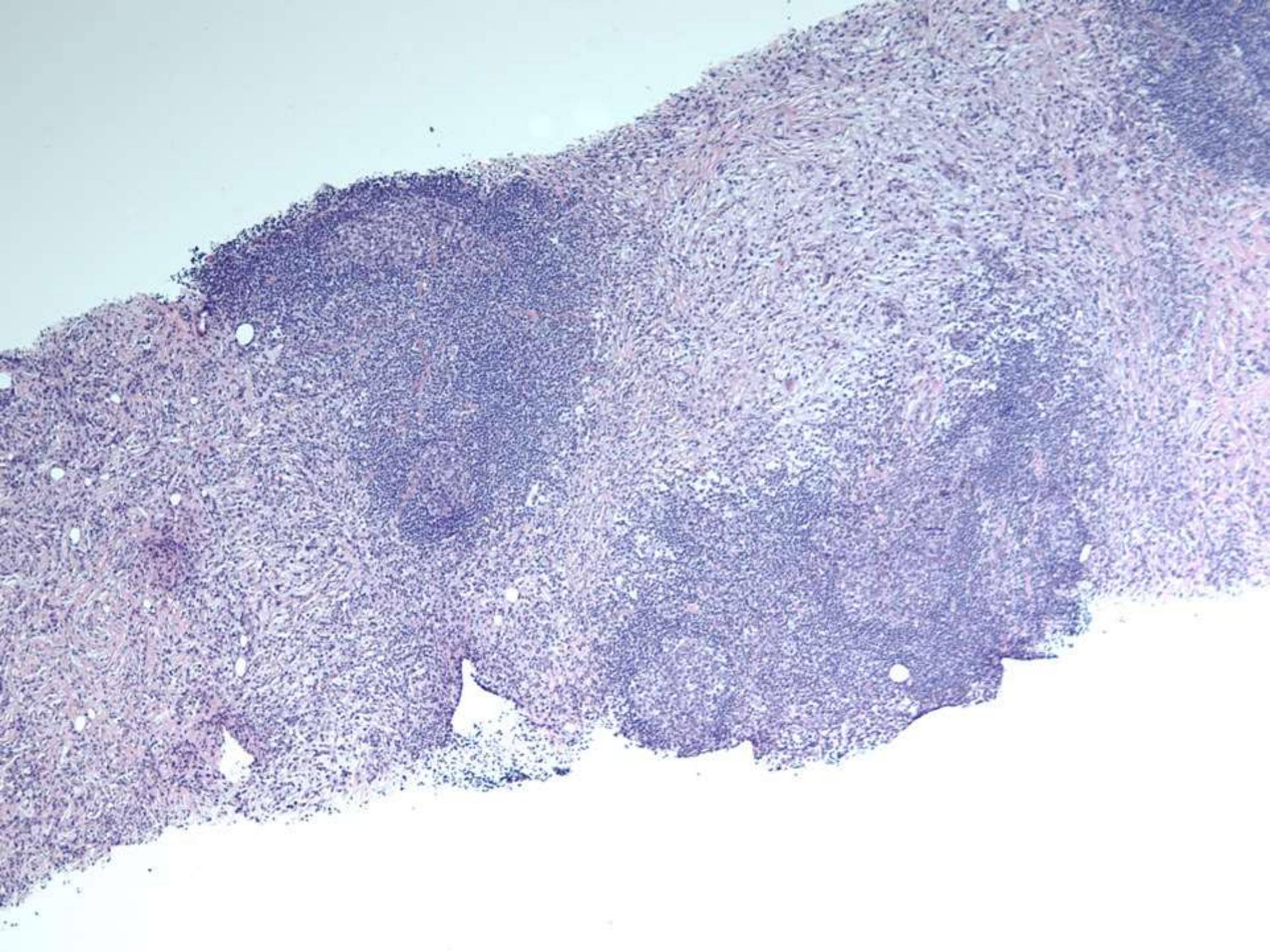
- Doing well since surgery, no pain
- Patient had numbness to left ear, face and upper neck at post-op visit
- The patient is without facial weakness or new neck mass

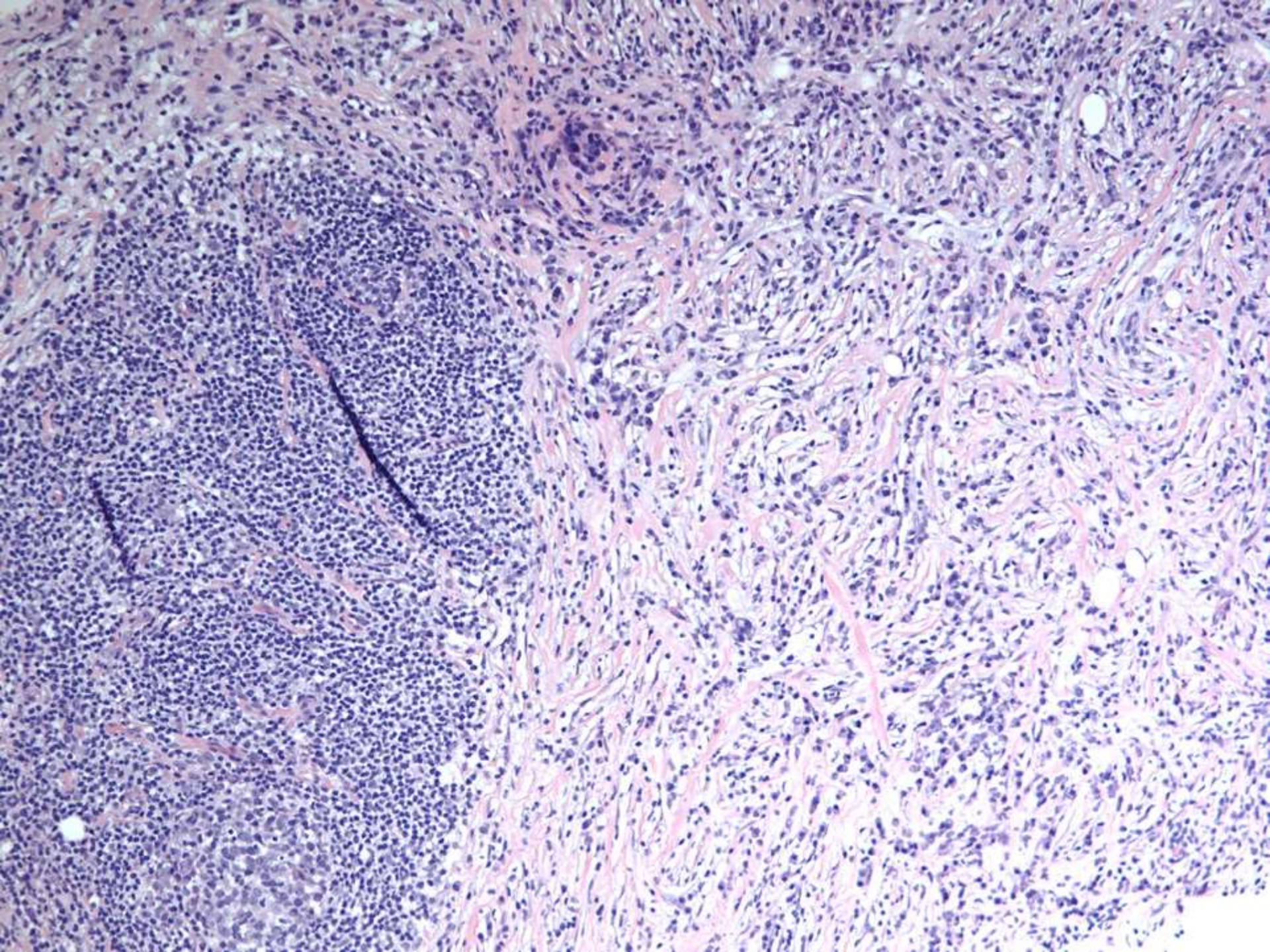
SB 6176

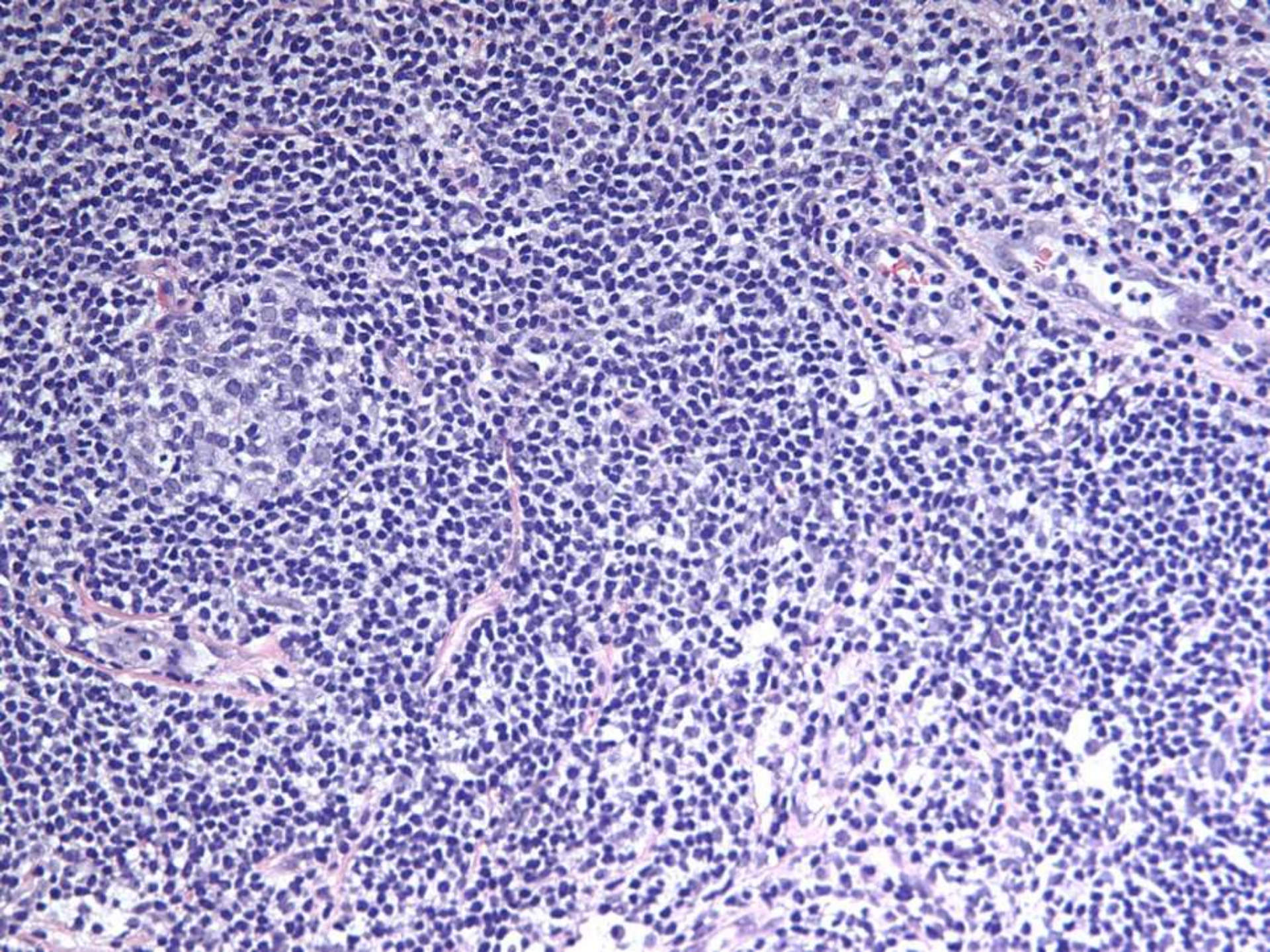
Jenny Hoffmann/Robert Ohgami; Stanford

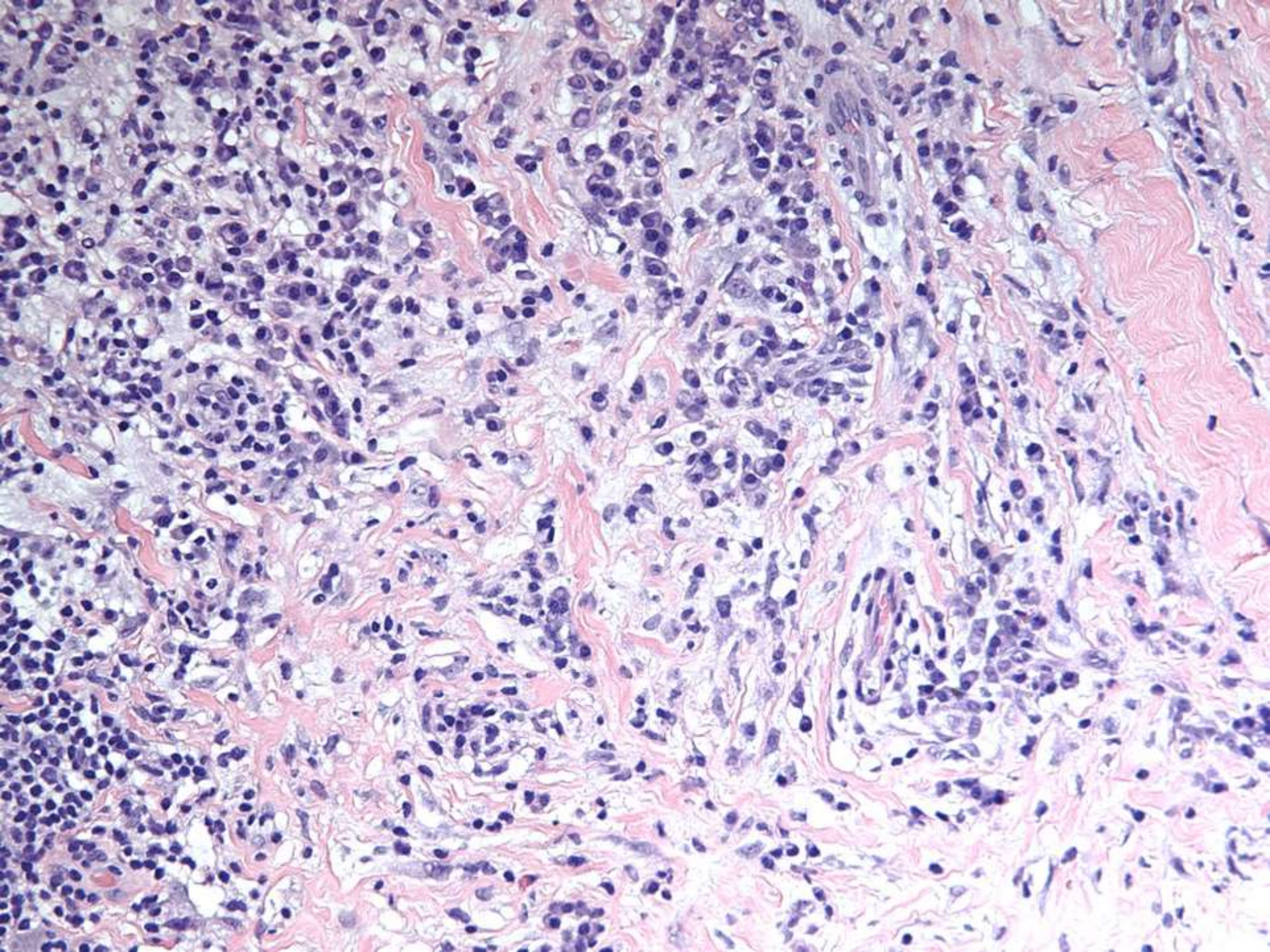
56-year-old woman with ill-defined 1.9cm mass at edge of breast with possible invasion into pectoralis muscle.

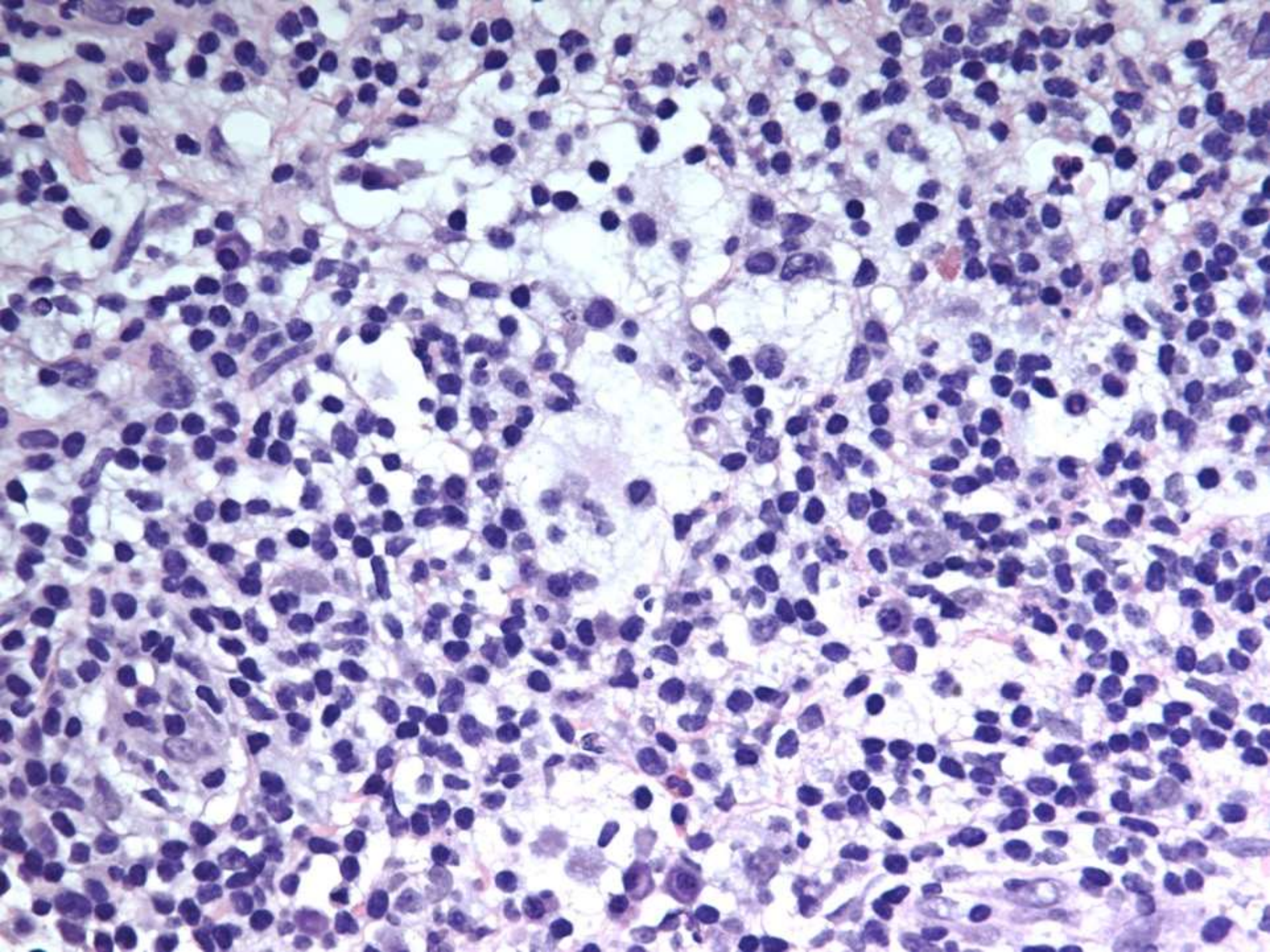


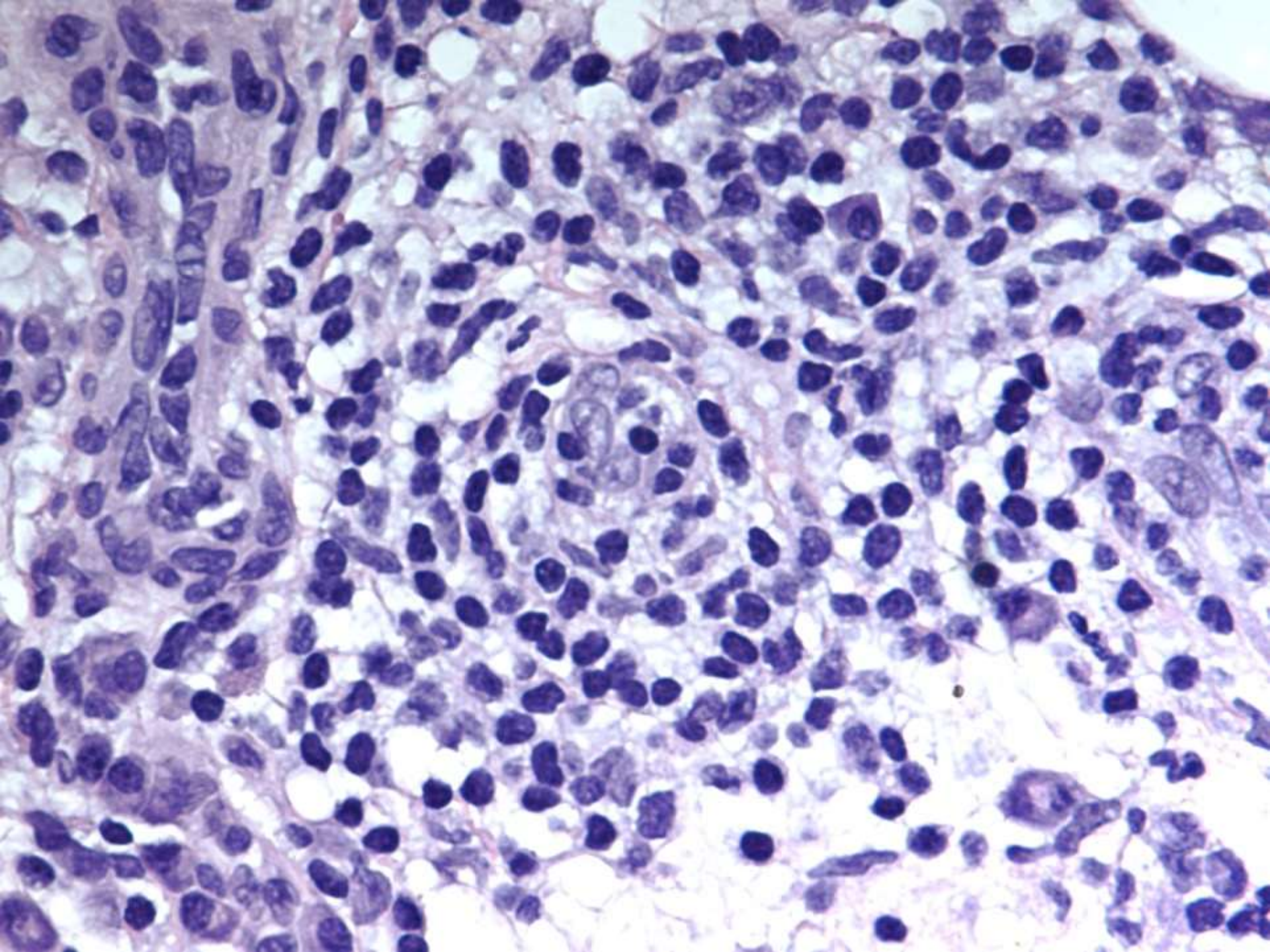












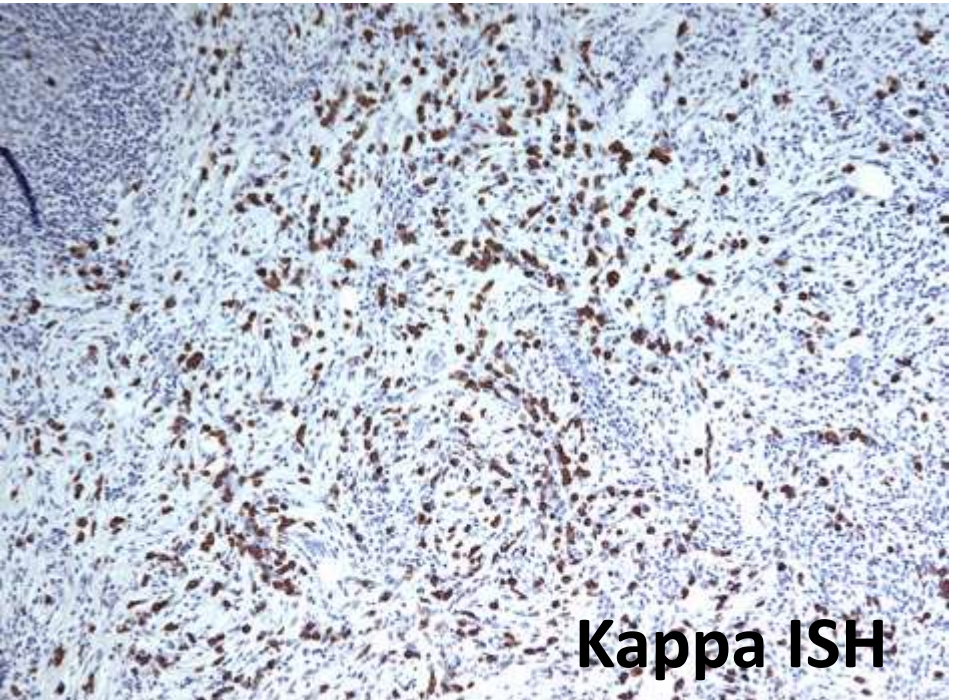
DIAGNOSIS?

Differential Dx

- Rosai-Dorfman disease (RDD)
- IgG4 related sclerosing mastitis
- Inflammatory pseudotumor/inflammatory myofibroblastic tumor
- Granulomatous mastitis/infective granulomas
- Langerhans cell histiocytosis
- Erdheim-Chester


CD3


CD20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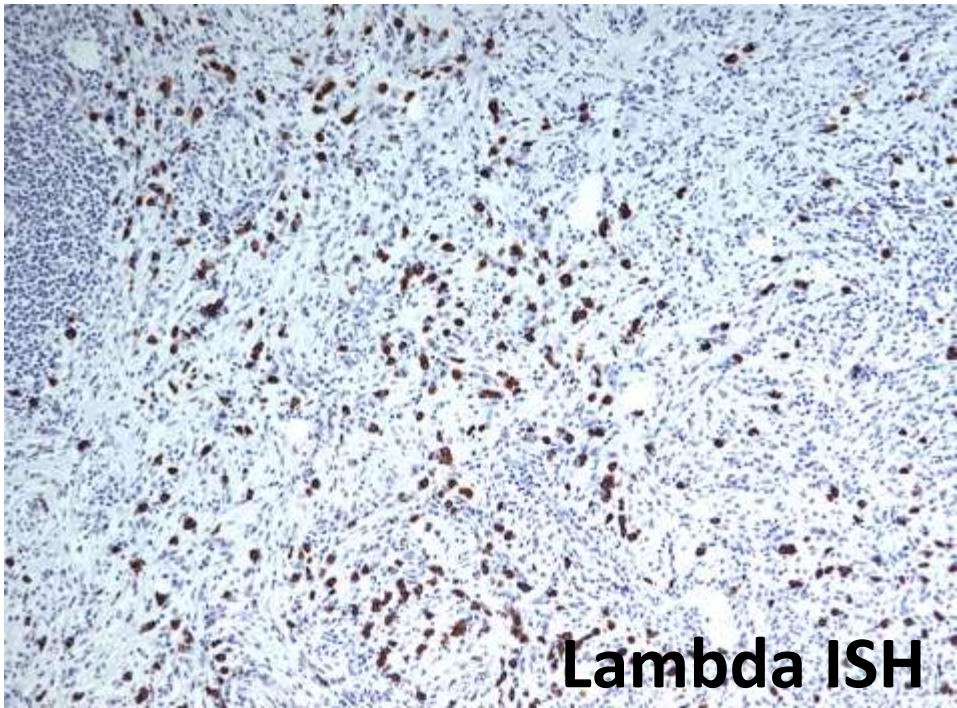
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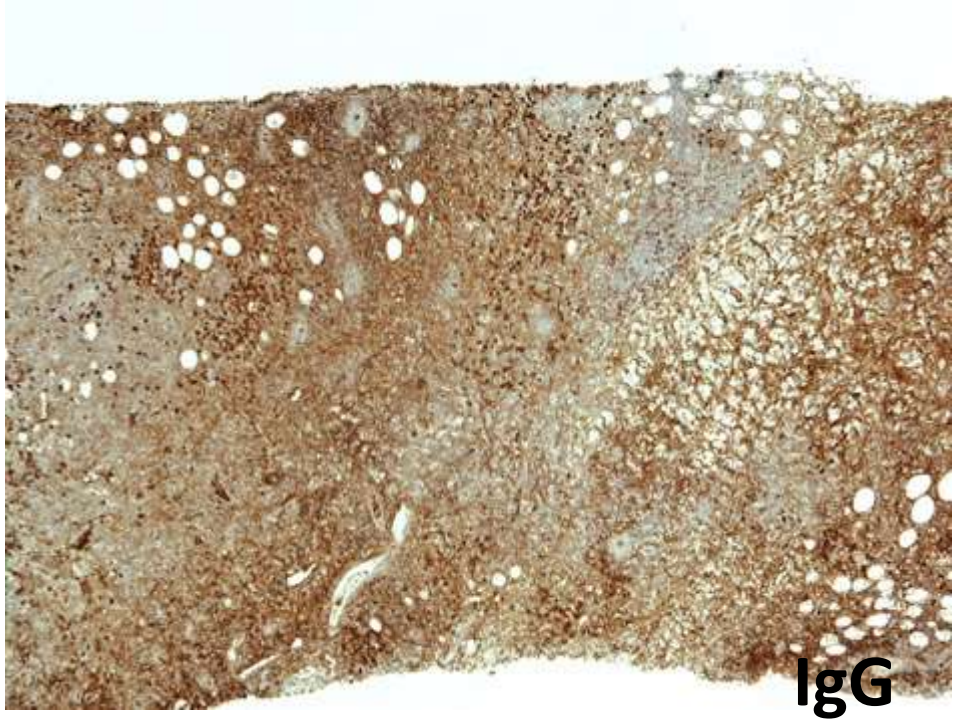
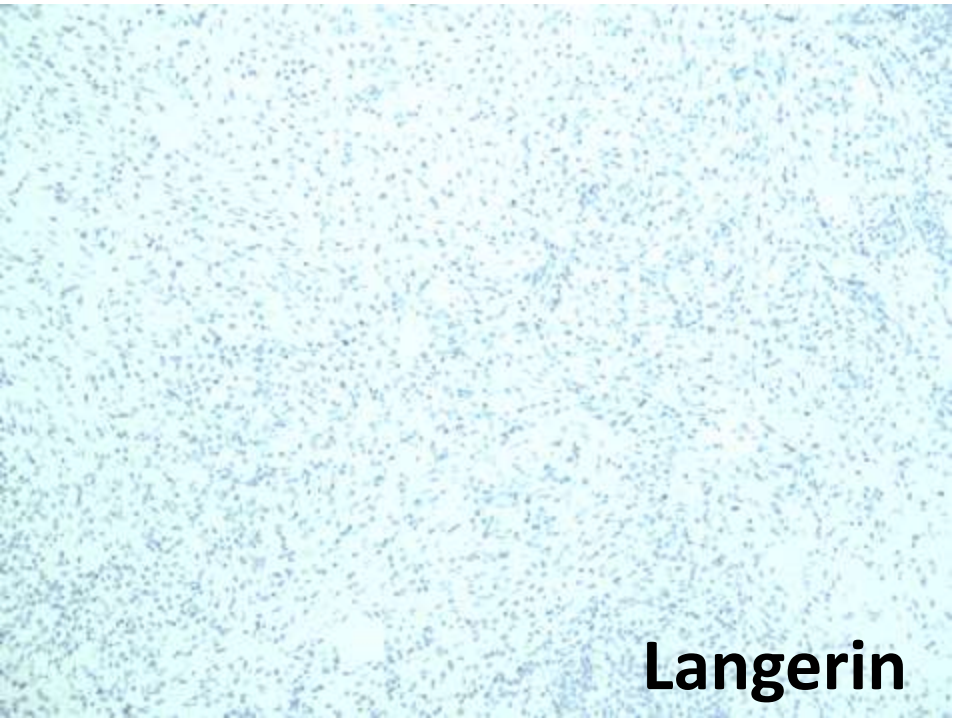
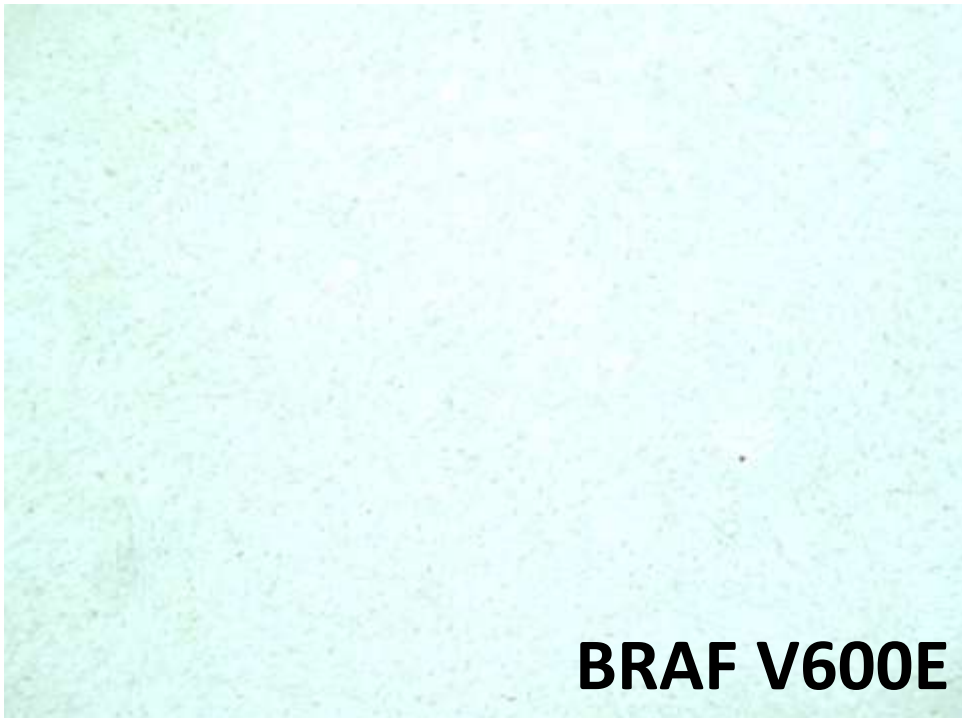
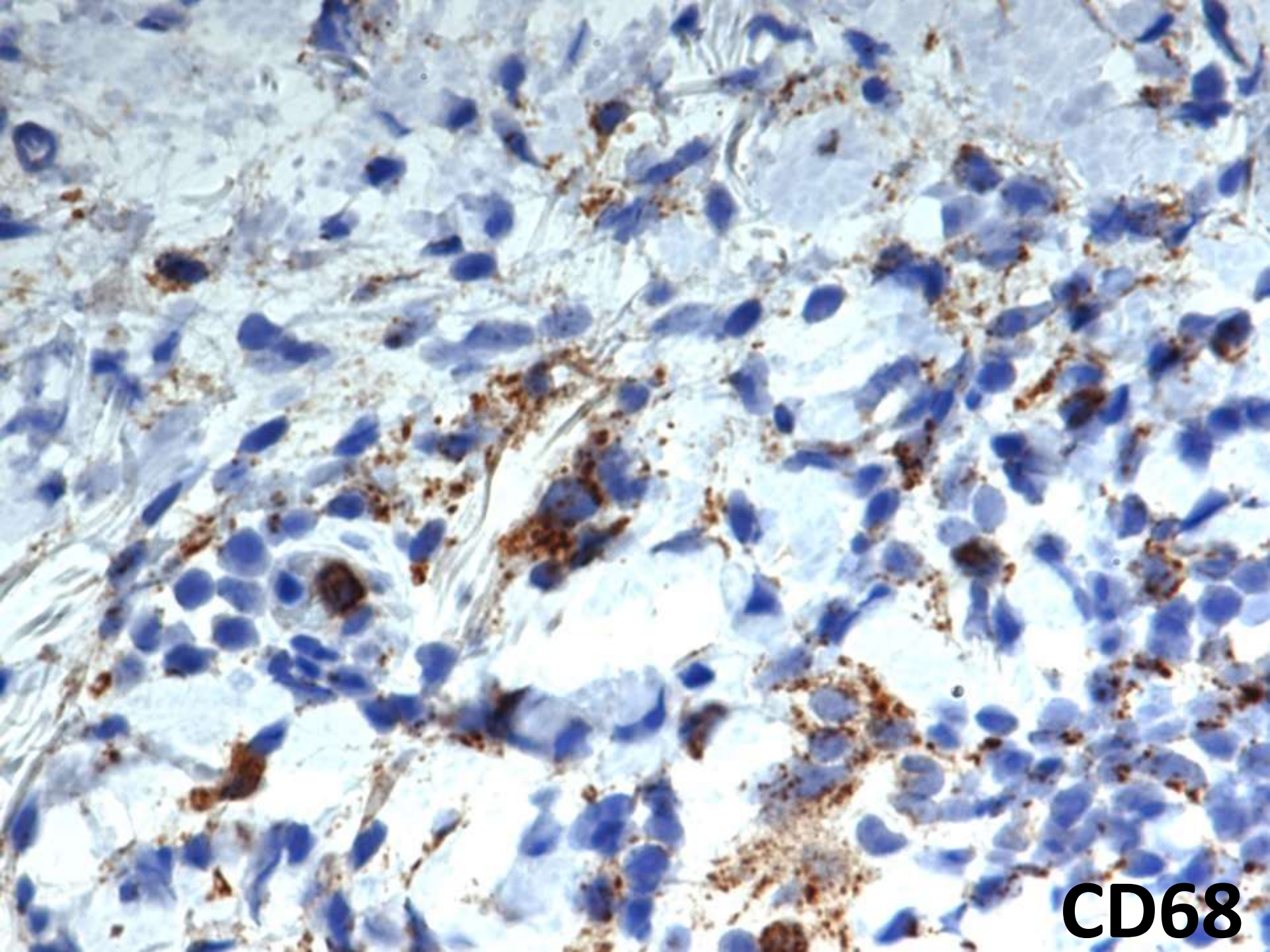
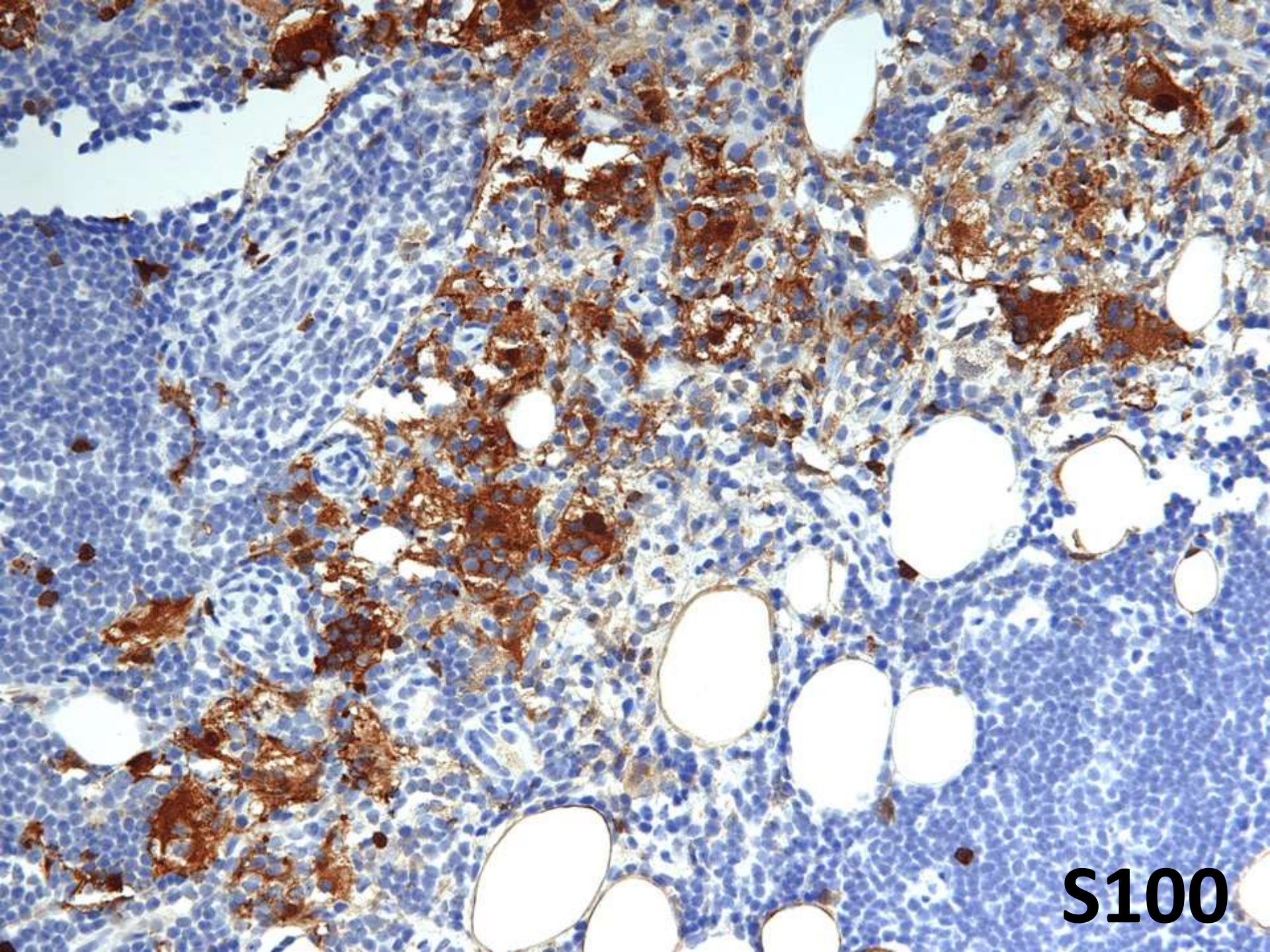

IgG

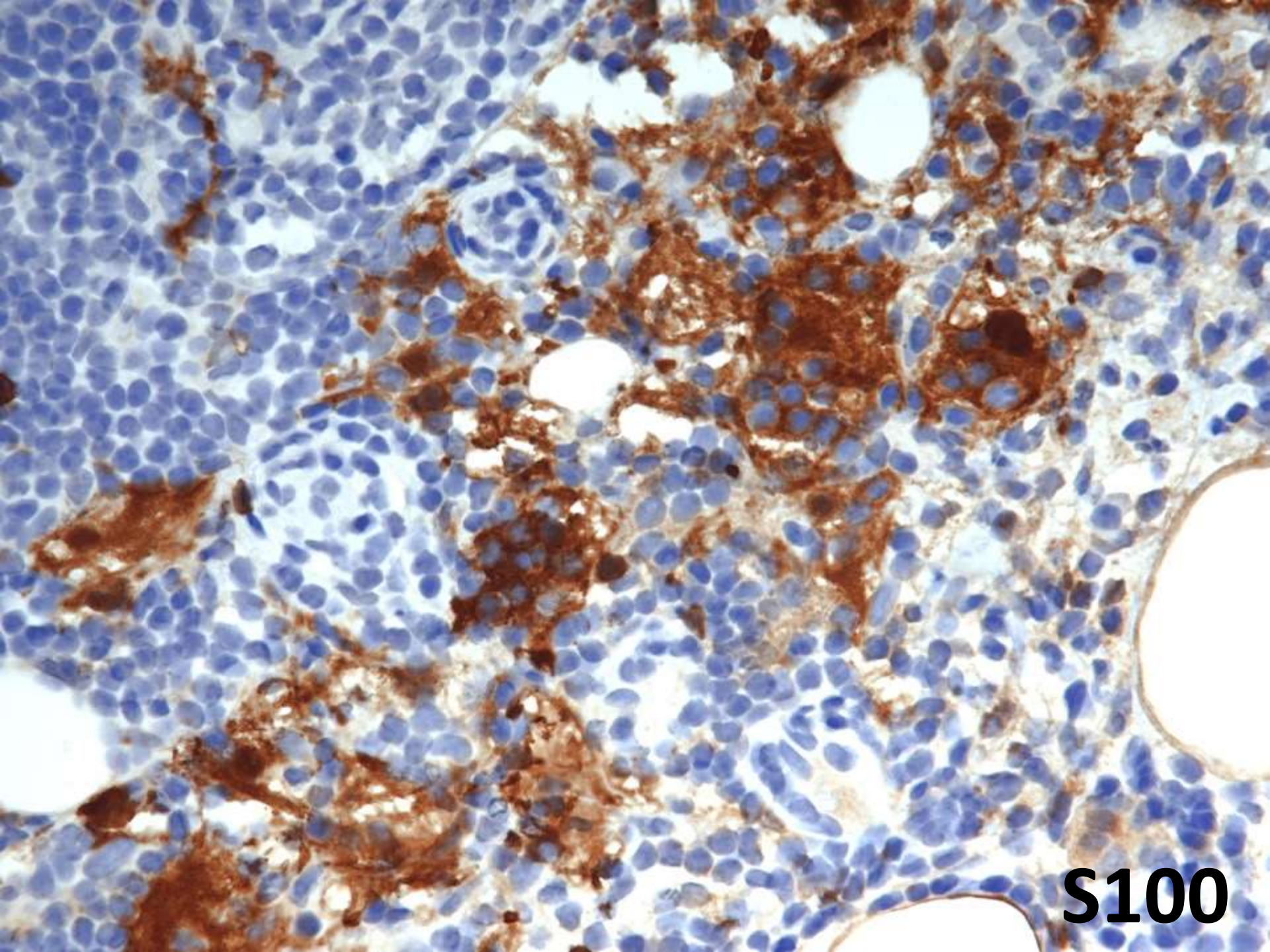

IgG4


Langerin


BRAF V600E


CD68


S100


S100

Diagnosis

BREAST, LEFT, 12 O'CLOCK, CORE BIOPSY

-- EXTRANODAL ROSAI-DORFMAN DISEASE

Rosai-Dorfman Disease of the Breast

- Very rare, usually in females – 3 cases reported in literature in males
- Clinical presentation
  - Most commonly as asymptomatic breast mass
  - Can involve ipsilateral axillary LN, or be bilateral or systemic
- Morphology
  - Ill-defined aggregates of histiocytes in background of lymphocytes and plasma cells
  - May have dense fibrosis
  - Histiocytes w/ engulfed lymphs (rarely RBCs, PCs, neutrophils)

A review of 16 cases of RDD of breast

- All cases had prominent lymphocytic inflammation (16/16)
  - 10/16 cases lymphocytes formed nodules some with GCs
  - 6/16 lymphocytes, lymphocytic inflammation diffuse
- Sclerosis moderate to prominent (12/16)
  - Multiple patterns: Storiform, thick collagen bands dissecting into fat, wispy collagen bands
  - Histiocytes frequently located in these areas, can be obscured and mimic myofibroblastic process (emperipoiesis can be indistinct)
- Numerous plasma cells (13/16)
- IgG4:IgG increased in 1 case (1/3)

Unpublished data

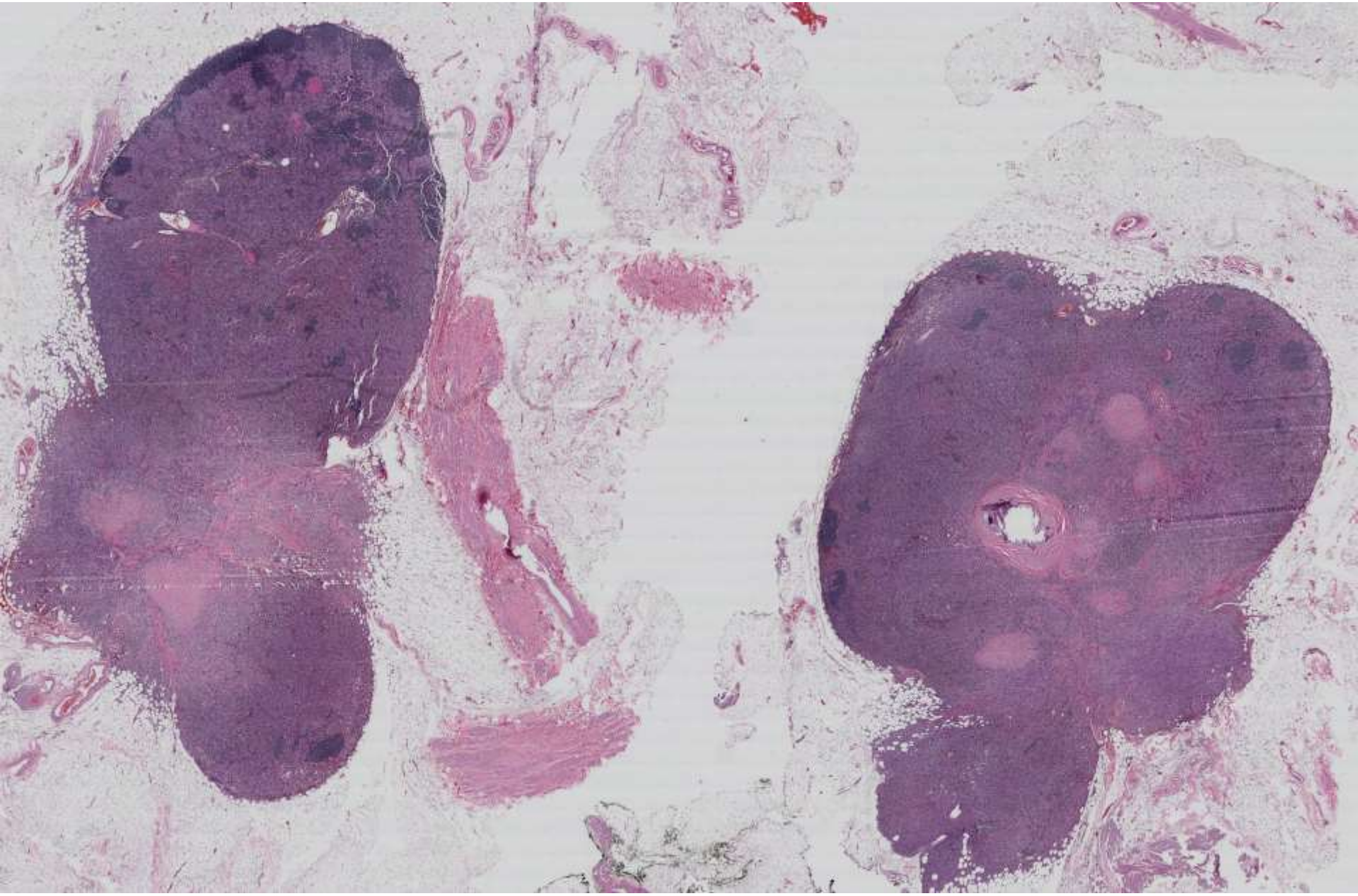
References

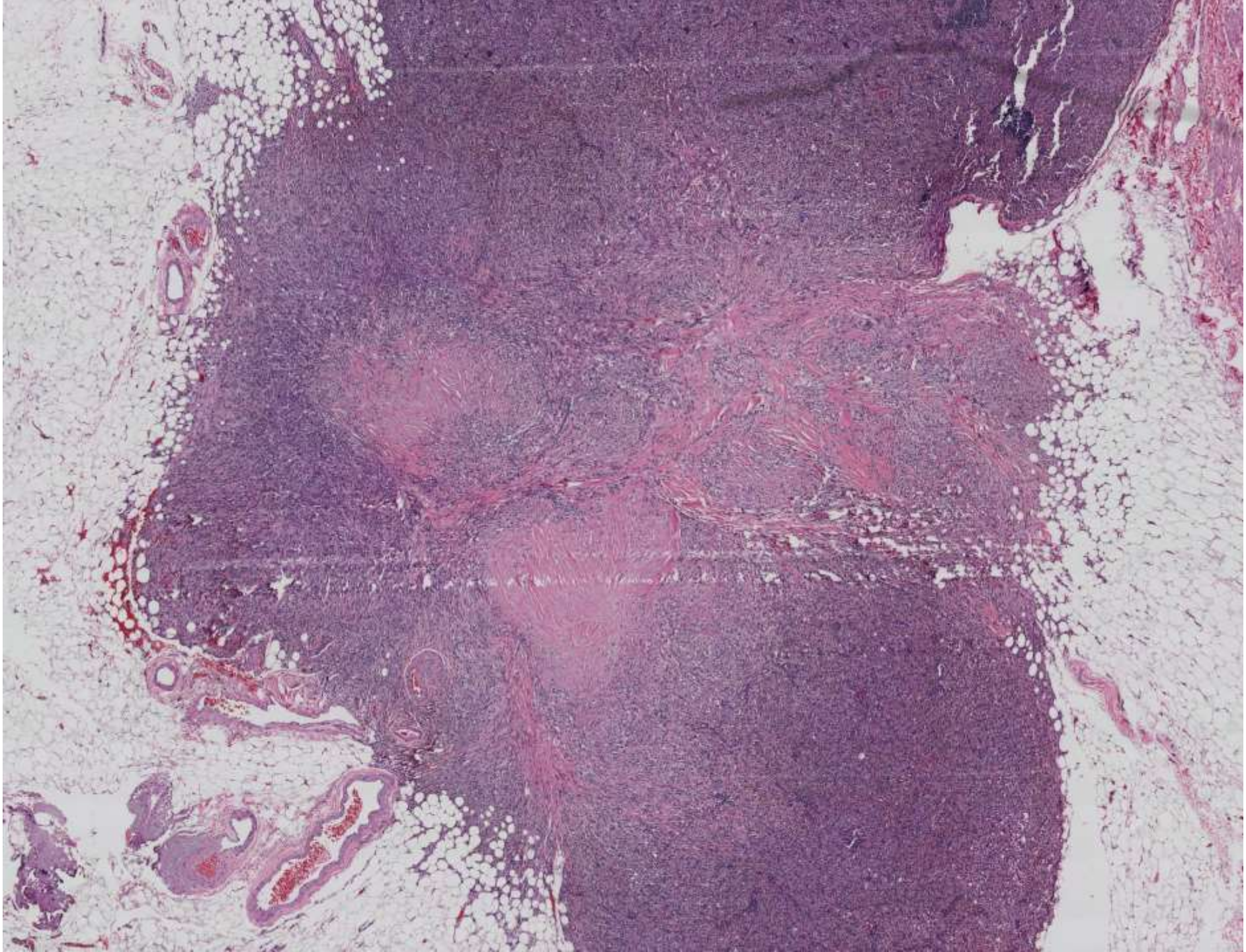
1) Green I, Dorfman RF, Rosai J. Breast involvement by extranodal Rosai–Dorfman disease: report of seven cases. Am. J. Surg. Pathol. 21 (1997) 664–668.
2) Chen YP, Jiang XN, Lu JP, et al. Clinicopathologic analysis of extranodal Rosai-Dorfman disease of the breast: a report of 12 cases. Zhonghua Bing Li Xue Za Zhi. 2016;45(8):556-60
3) Liu M, Li X, Li Y, et al. Rosai-dofrman disease with features of IgG4-related disease in the breast: cases report and literature review. Asian Pac J Allergy Immunol. 2017: doi10.12932.
4) Mantilla JG, Goldberg-Stein S, Wang Y. Extranodal Rosai-Dorfman Disease: Clinicopathologic Series of 10 patients with radiologic correlation and review of the literature. Am J Clin Pathol. 2016:145(2):211-21.
5) Delaney EE, Larkin A, MacMaster S, et al. Rosai-Dorfman Disease of the breast. Cureus. 9(4):e1153 DOI 10.7759/cureus.1153
6) El-Attrache B, Kapenhas E, Morgani J, et al. A rarity in breast pathology: a male case of Rosai-Dorfman disease and literature review. Int J Surg Case Rep. 2017;37:1-3.

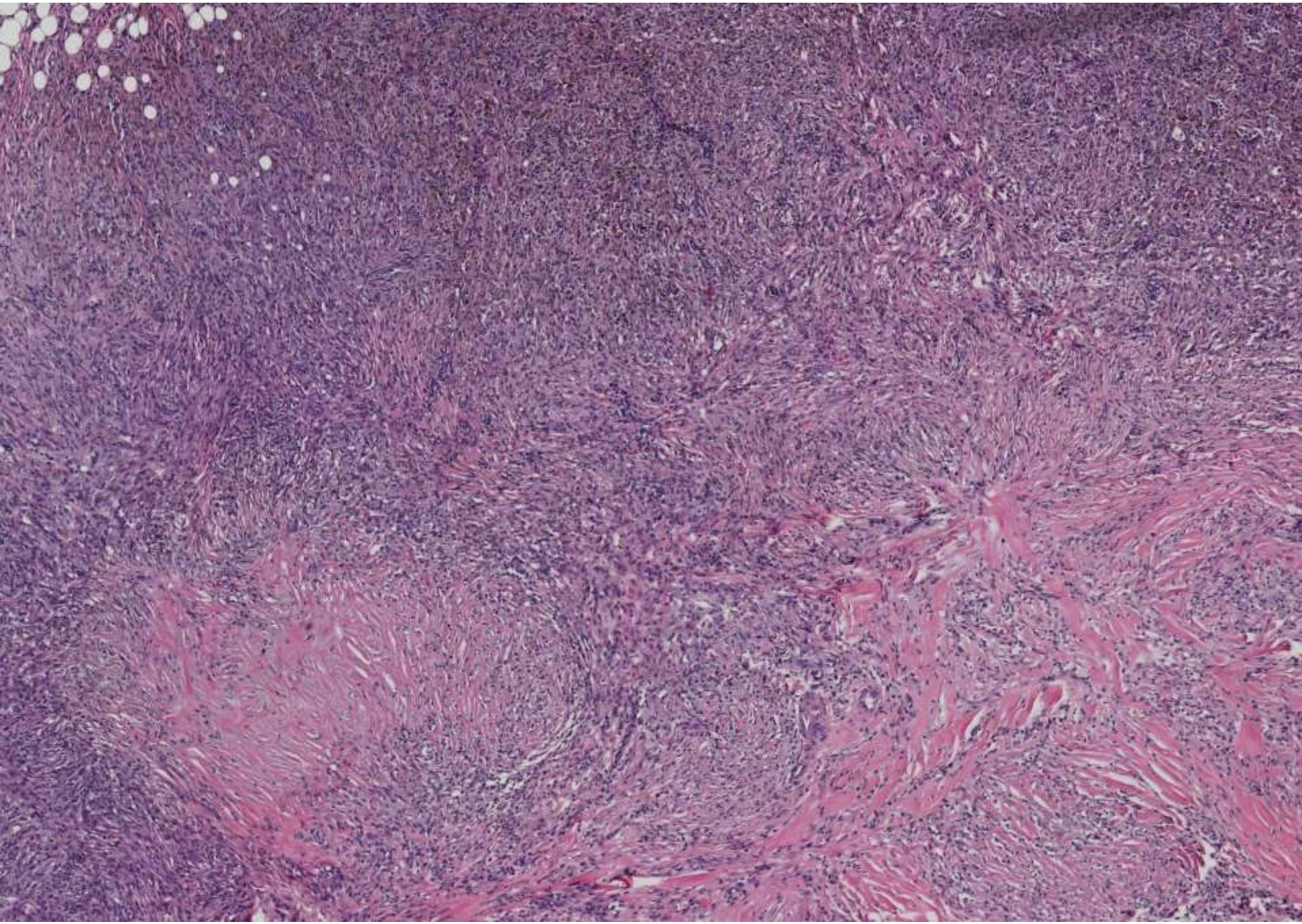
SB 6177
(scanned slide available)

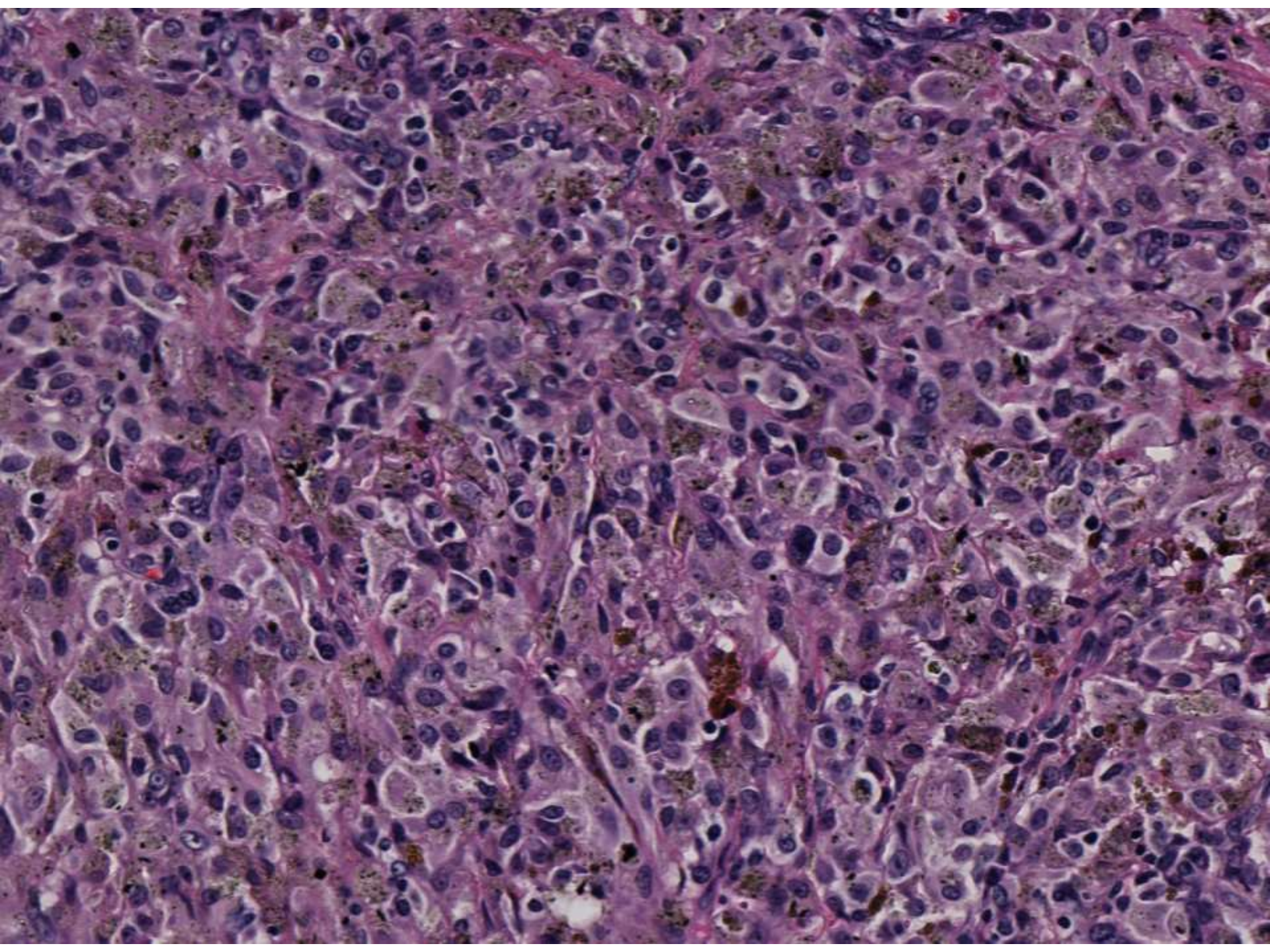
Ankur Sangoi; El Camino Hospital

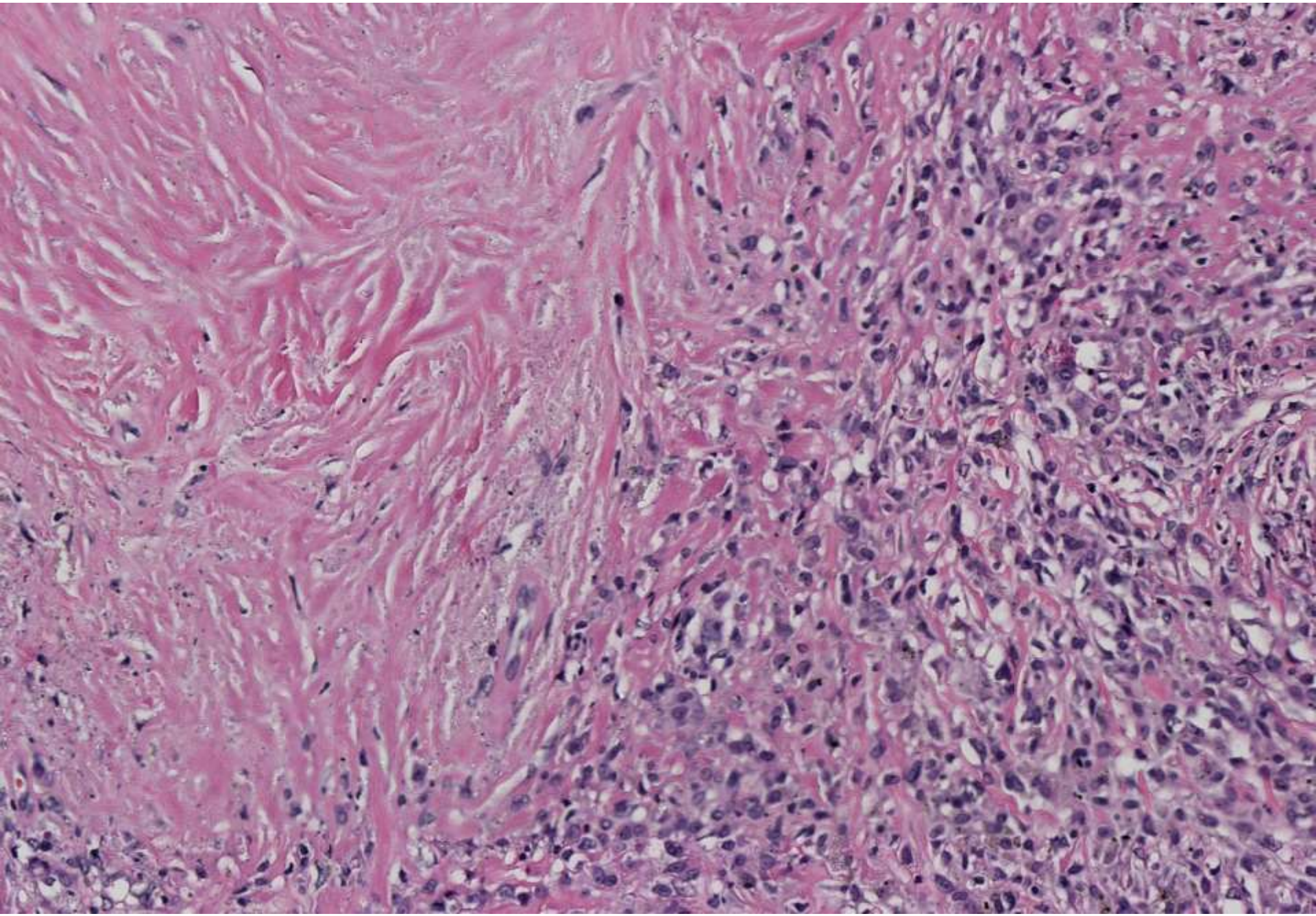
76-year-old man with enlarged mediastinal lymph node removed at time of aortic valve repair.

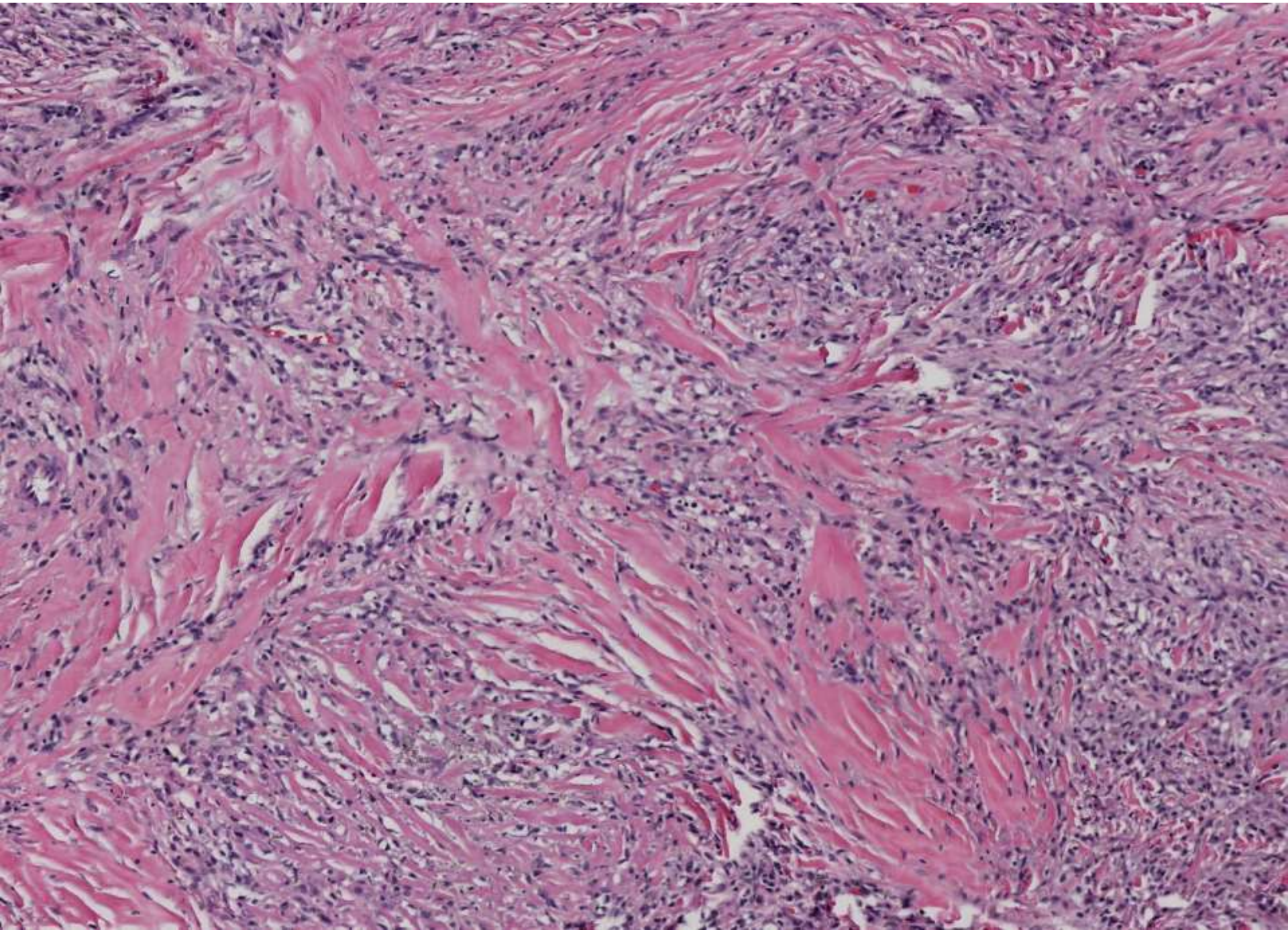


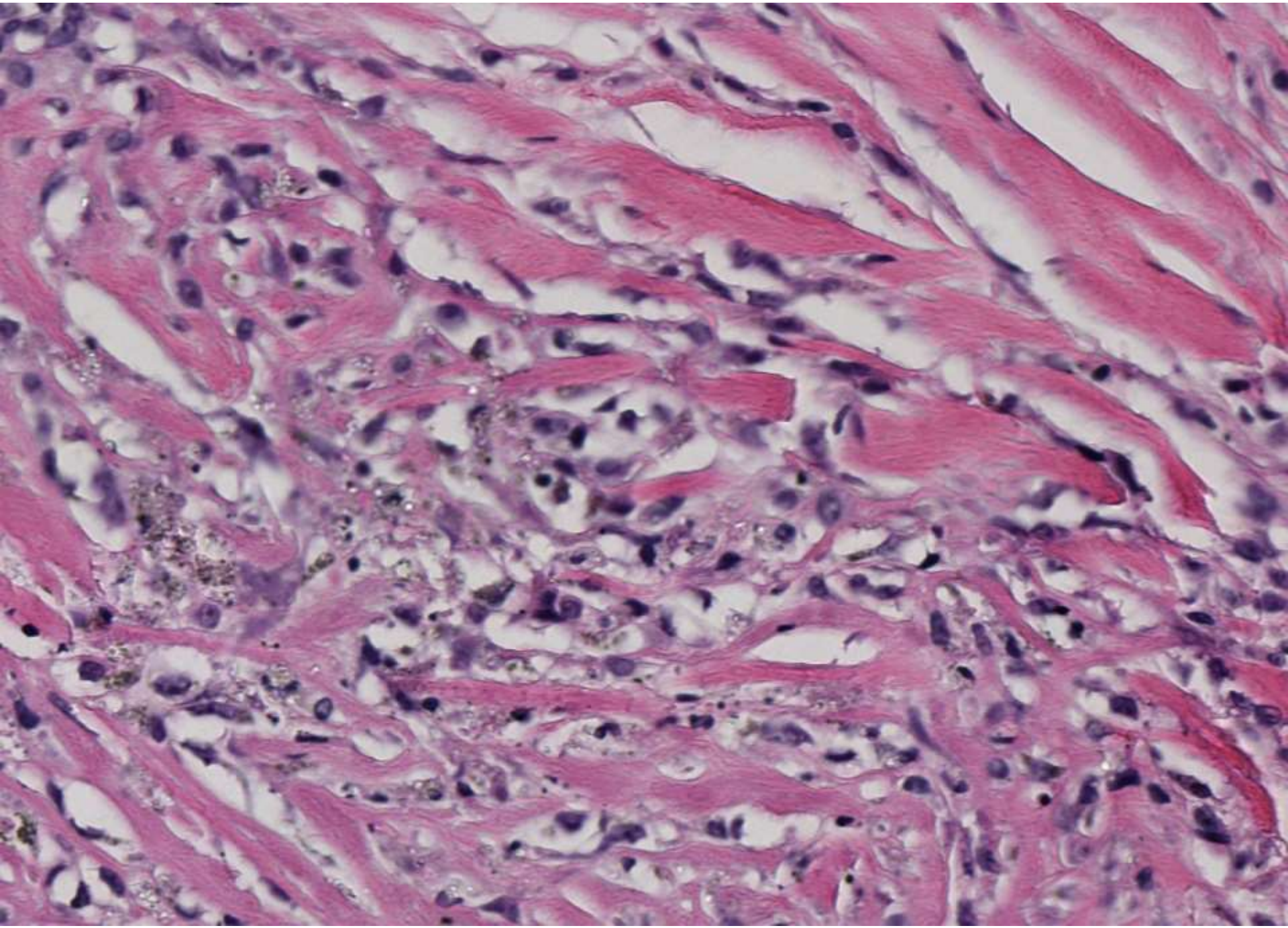


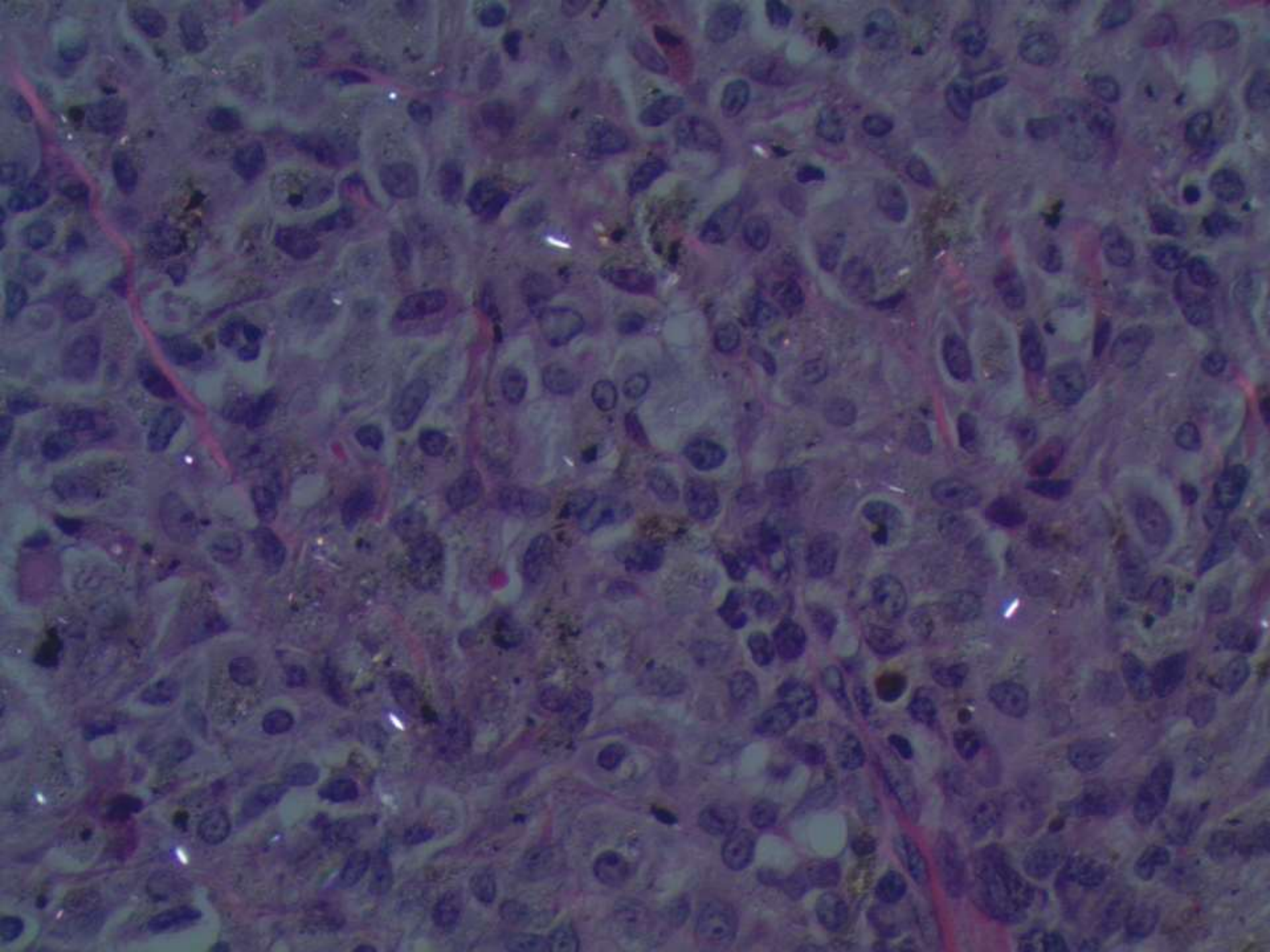












DIAGNOSIS?

DDx

- Follicular dendritic cell tumor
- Spindle cell melanoma
- Kaposi sarcoma
- Fibrohistiocytic neoplasms (MFH)
- Lymphoma (Hodgkin)
- Palisaded myofibroblastoma
- Inflammatory pseudotumor
- Infectious (mycobacterial pseudotumor)
- Reactive (crystalline pseudotumor)

FINAL DIAGNOSIS:

Anthracotic and Anthracosilicotic Spindle Cell Pseudotumors of Mediastinal Lymph Nodes:

Report of Five Cases of a Reactive Lesion That Simulates Malignancy

PEDRAM ARGANI, MD, RONALD GHOSSEIN, MD,
AND JUAN ROSAI, MD

HUMAN PATHOLOGY Volume 29, No. 8 (August 1998)

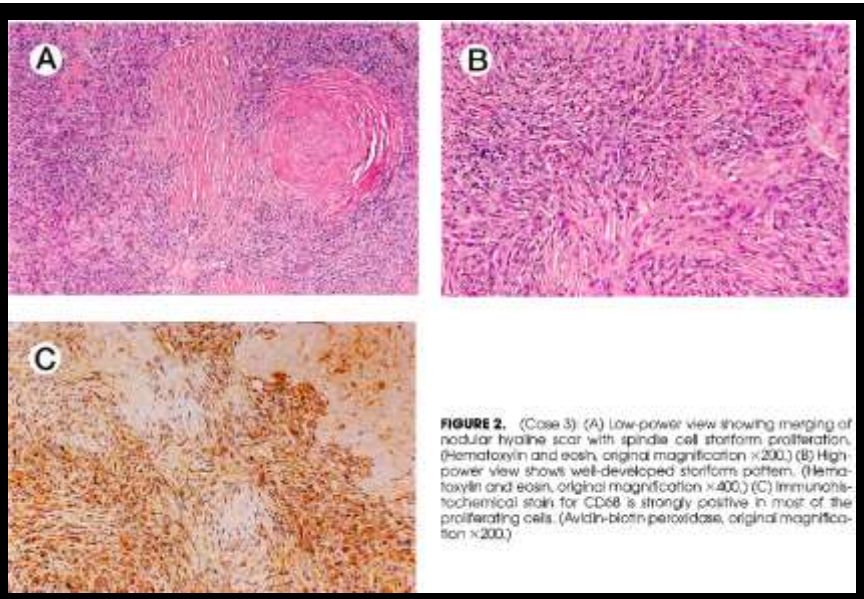
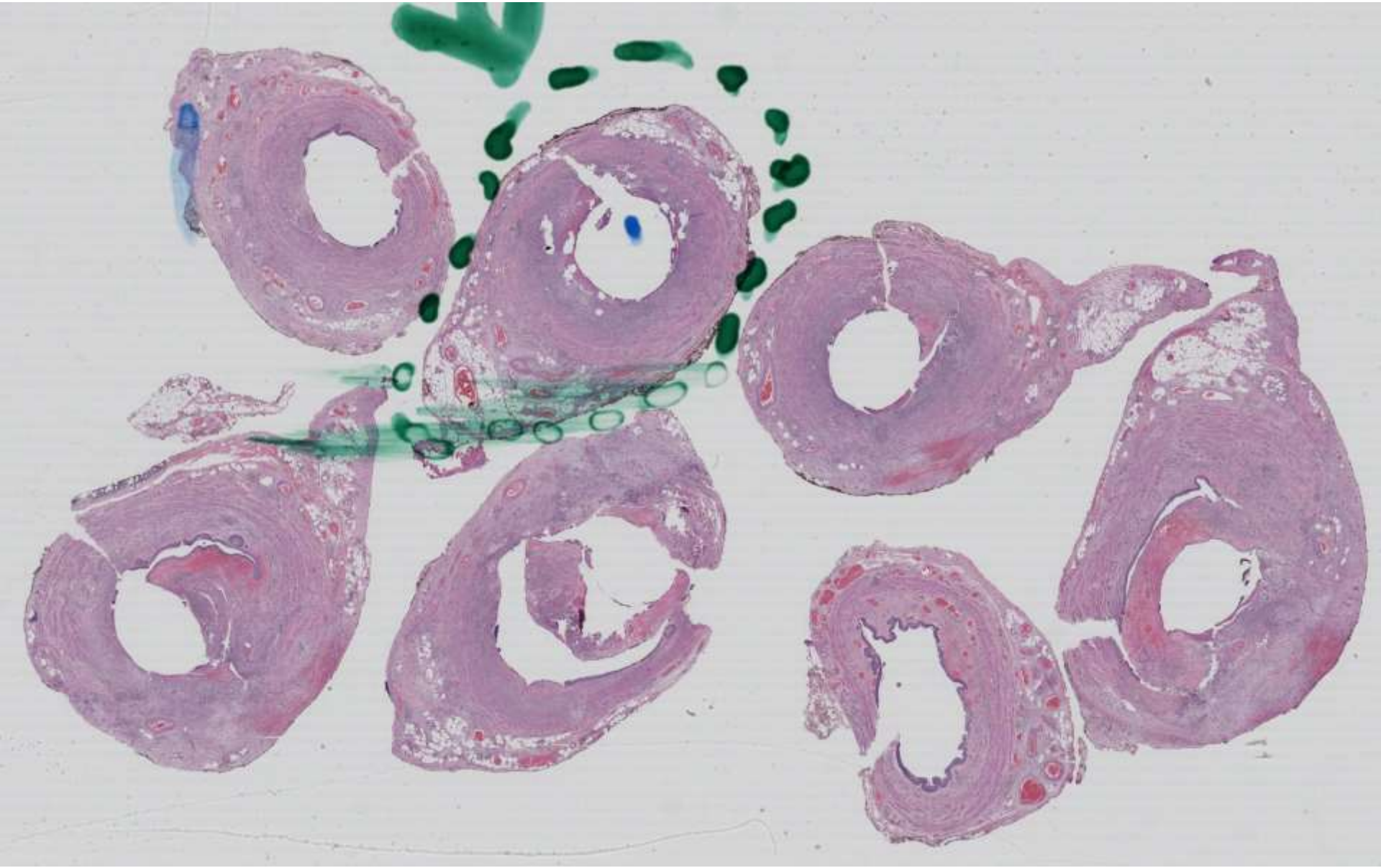


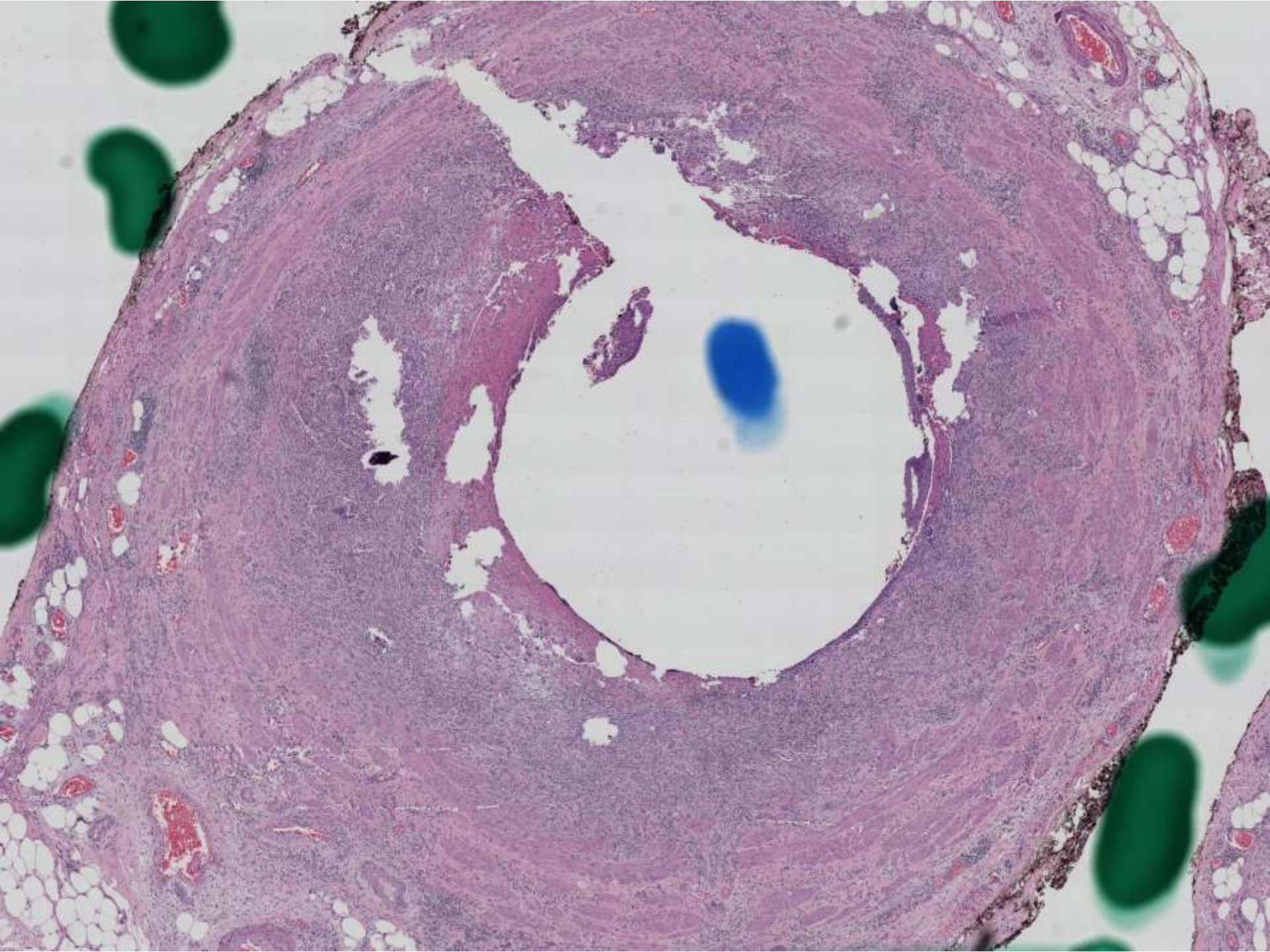
FIGURE 2. (Case 3) (A) Low-power view showing merging of nodular hyaline scar with spindle cell storiform proliferation. (Hematoxylin and eosin, original magnification ×200.) (B) High-power view shows well-developed storiform pattern. (Hematoxylin and eosin, original magnification ×400.) (C) Immunohistochemical stain for CD68 is strongly positive in most of the proliferating cells. (Avidin-biotin peroxidase, original magnification ×200.)

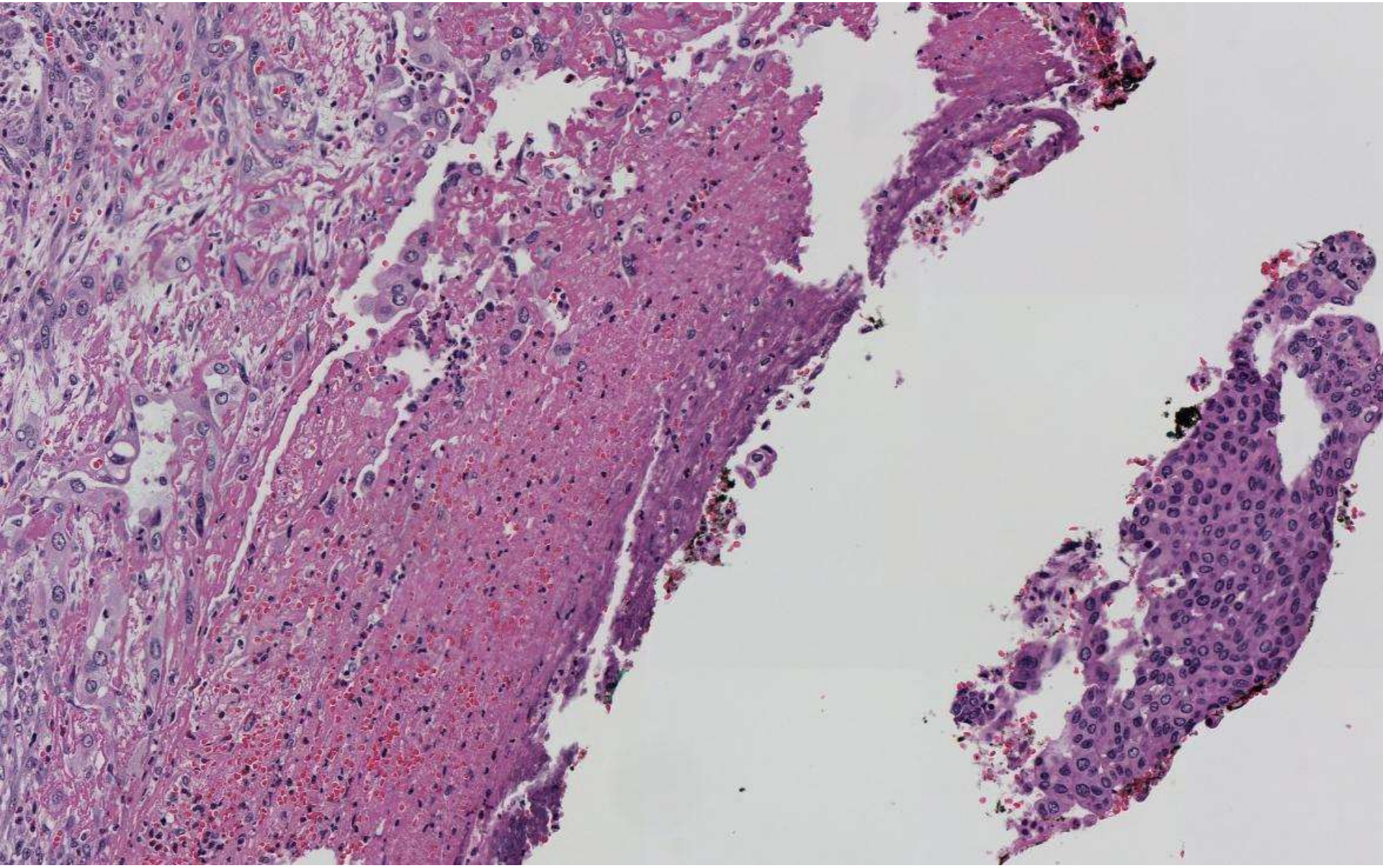
SB 6178
(scanned slide available)

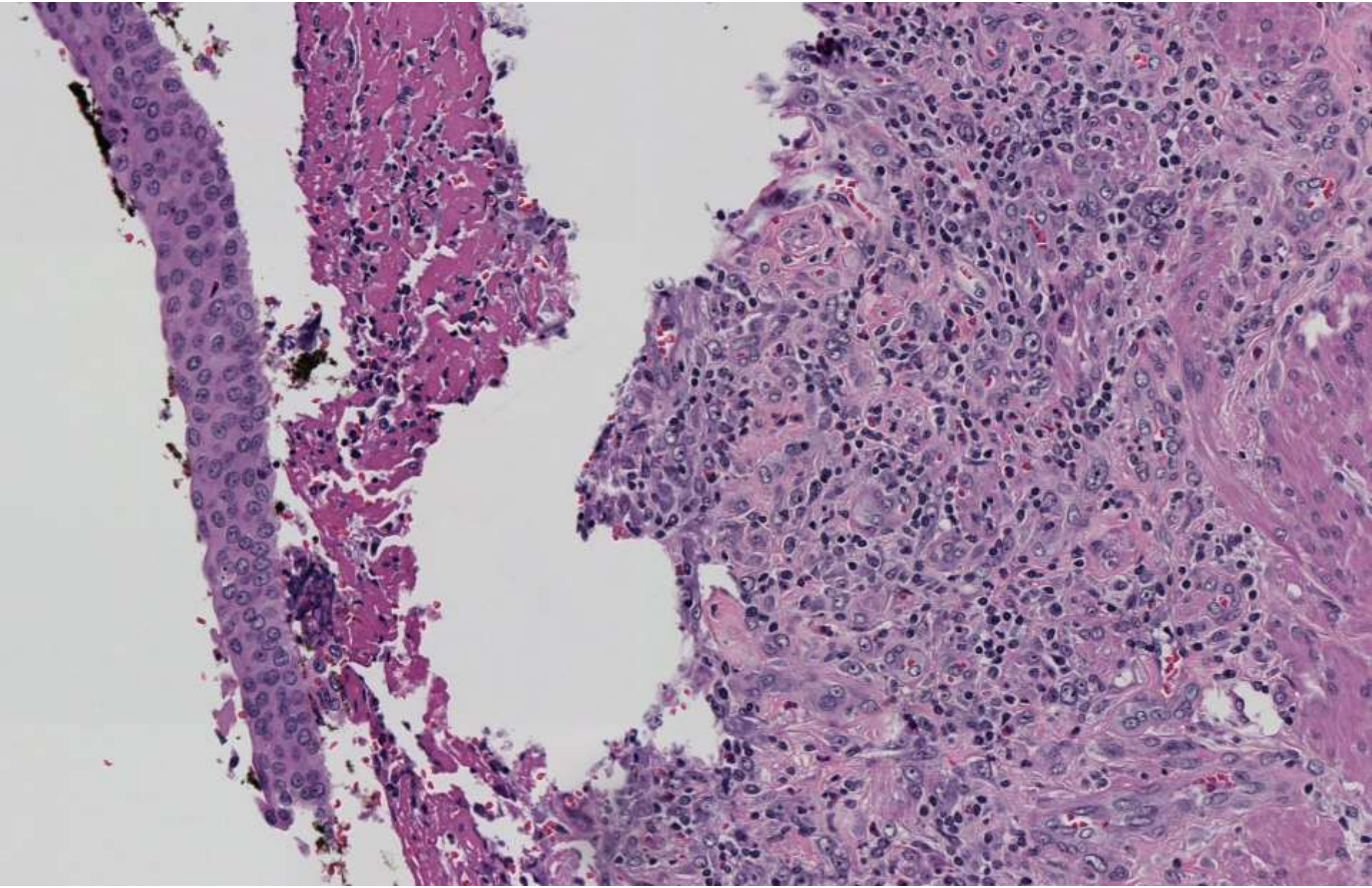
Ankur Sangoi; El Camino Hospital

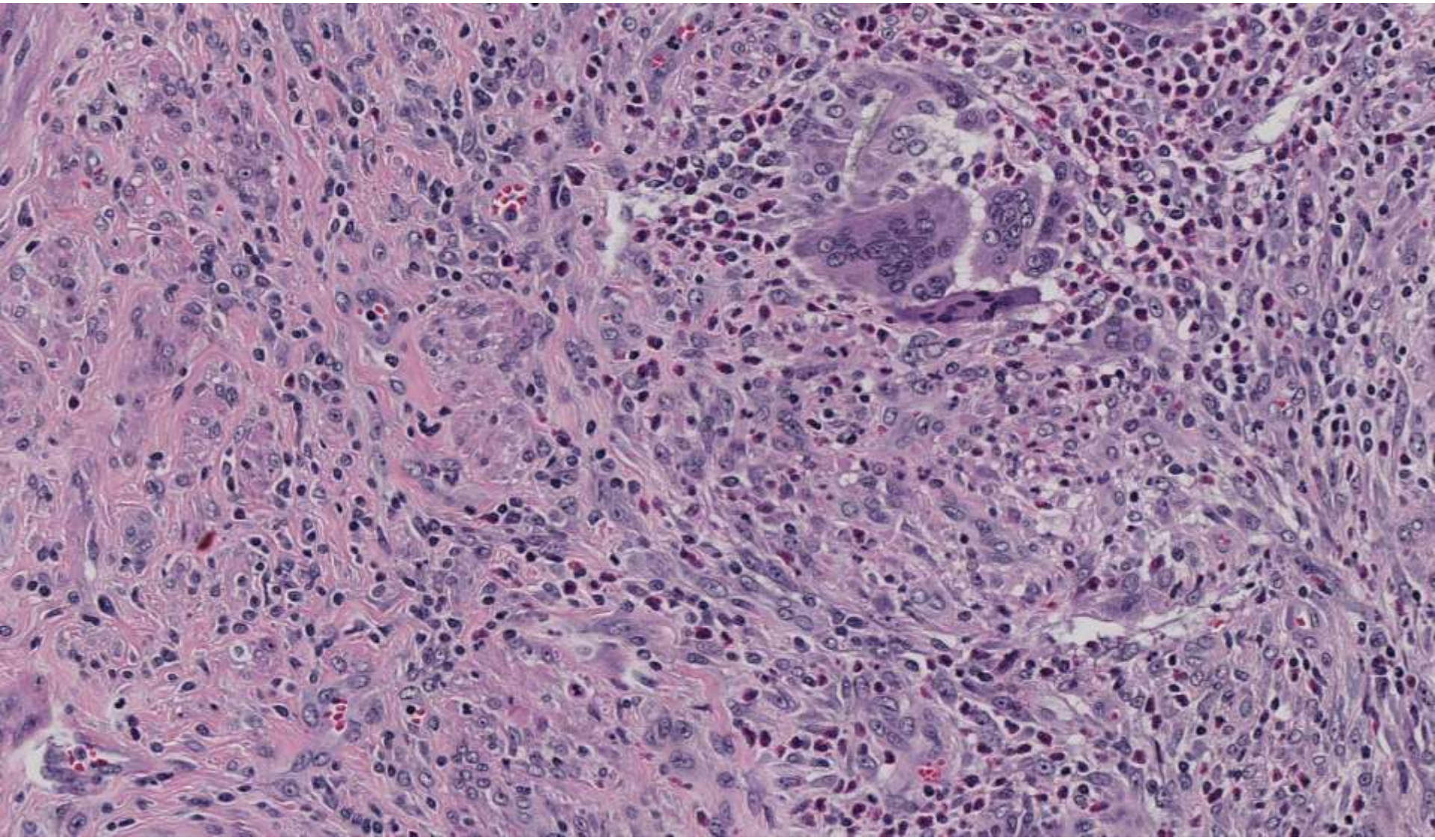
66-year-old man with nephrouretectomy performed for high grade non-invasive papillary renal cell carcinoma at renal pelvis. Section of mid ureter submitted.

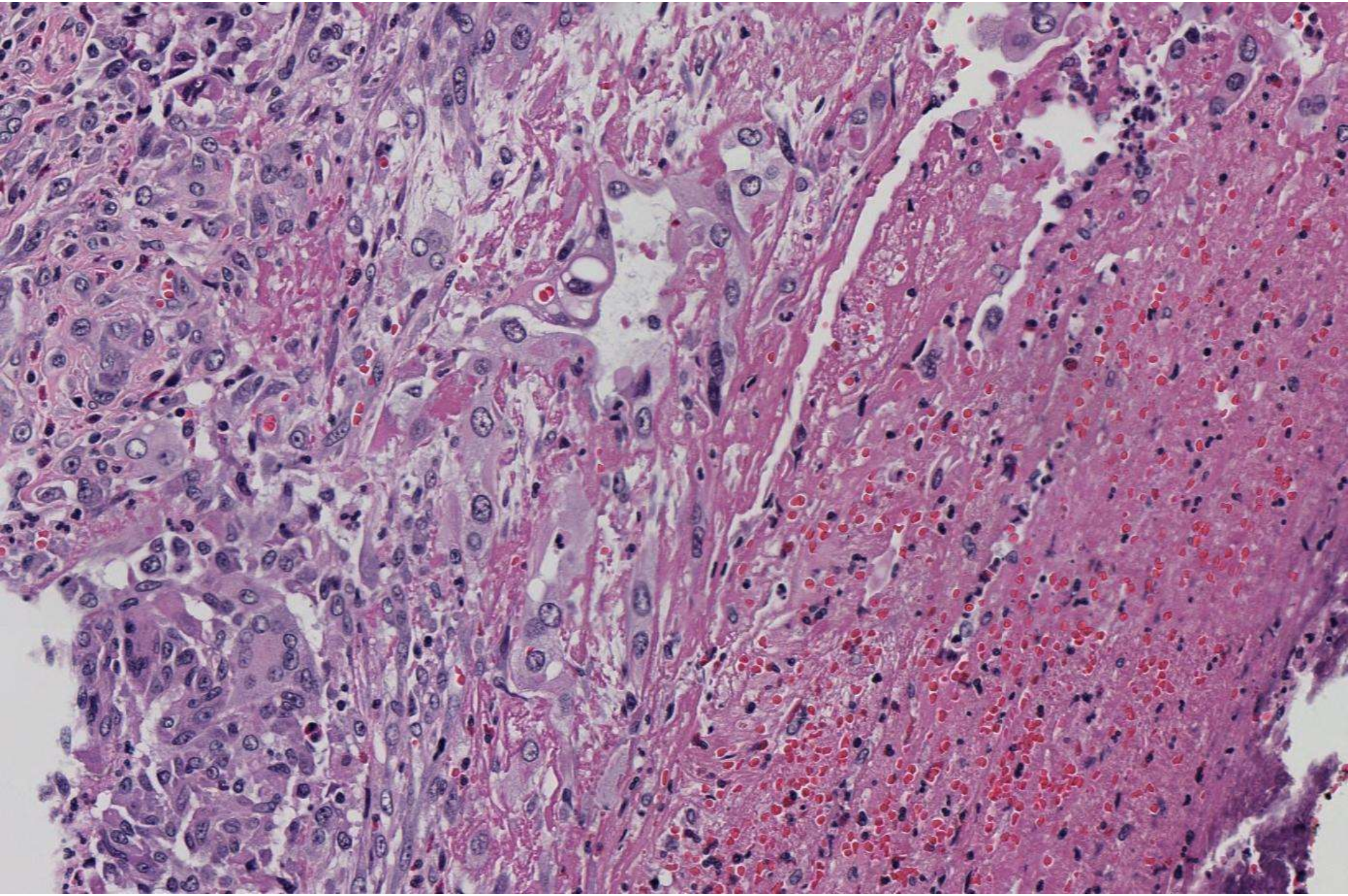


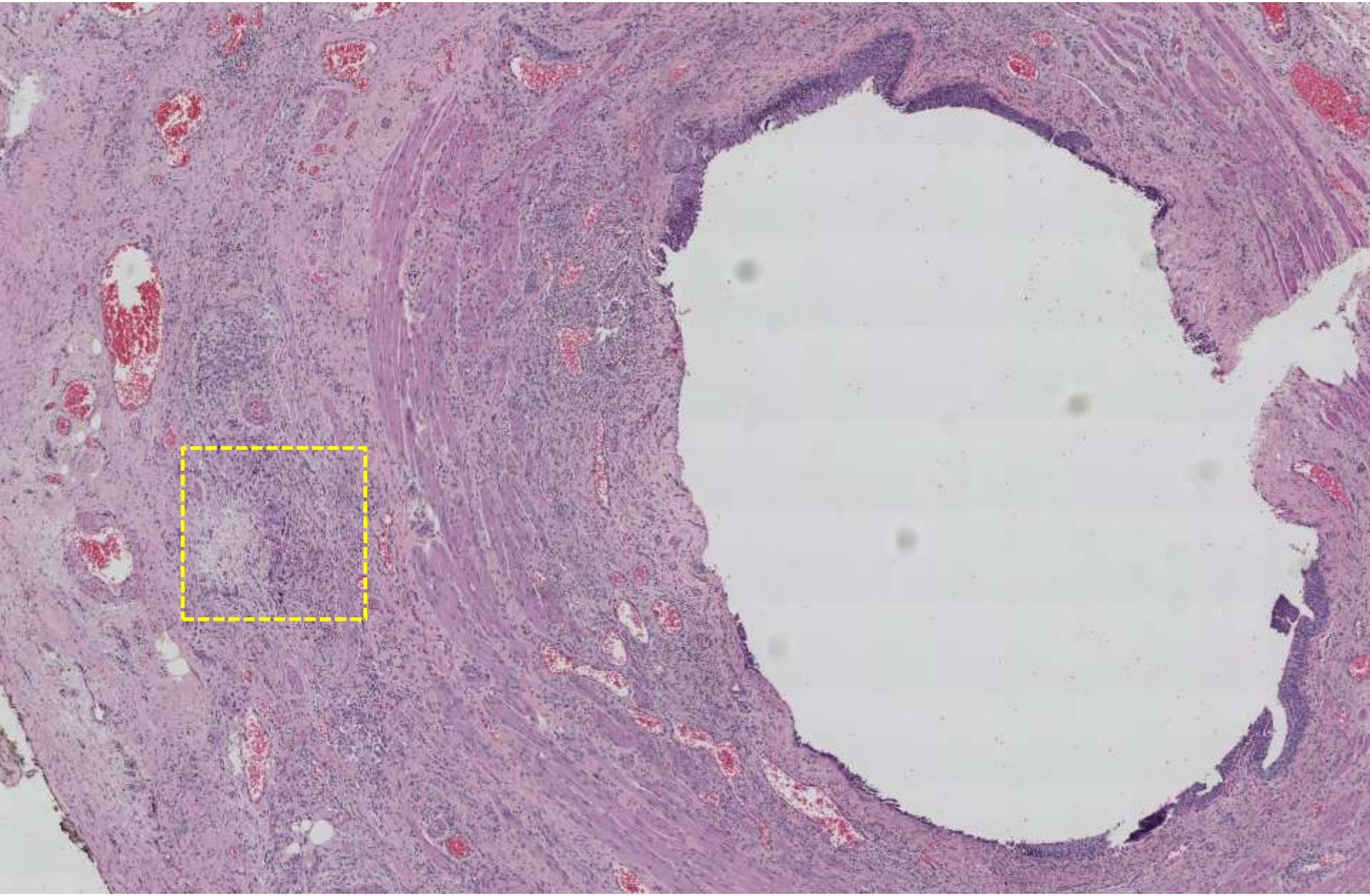


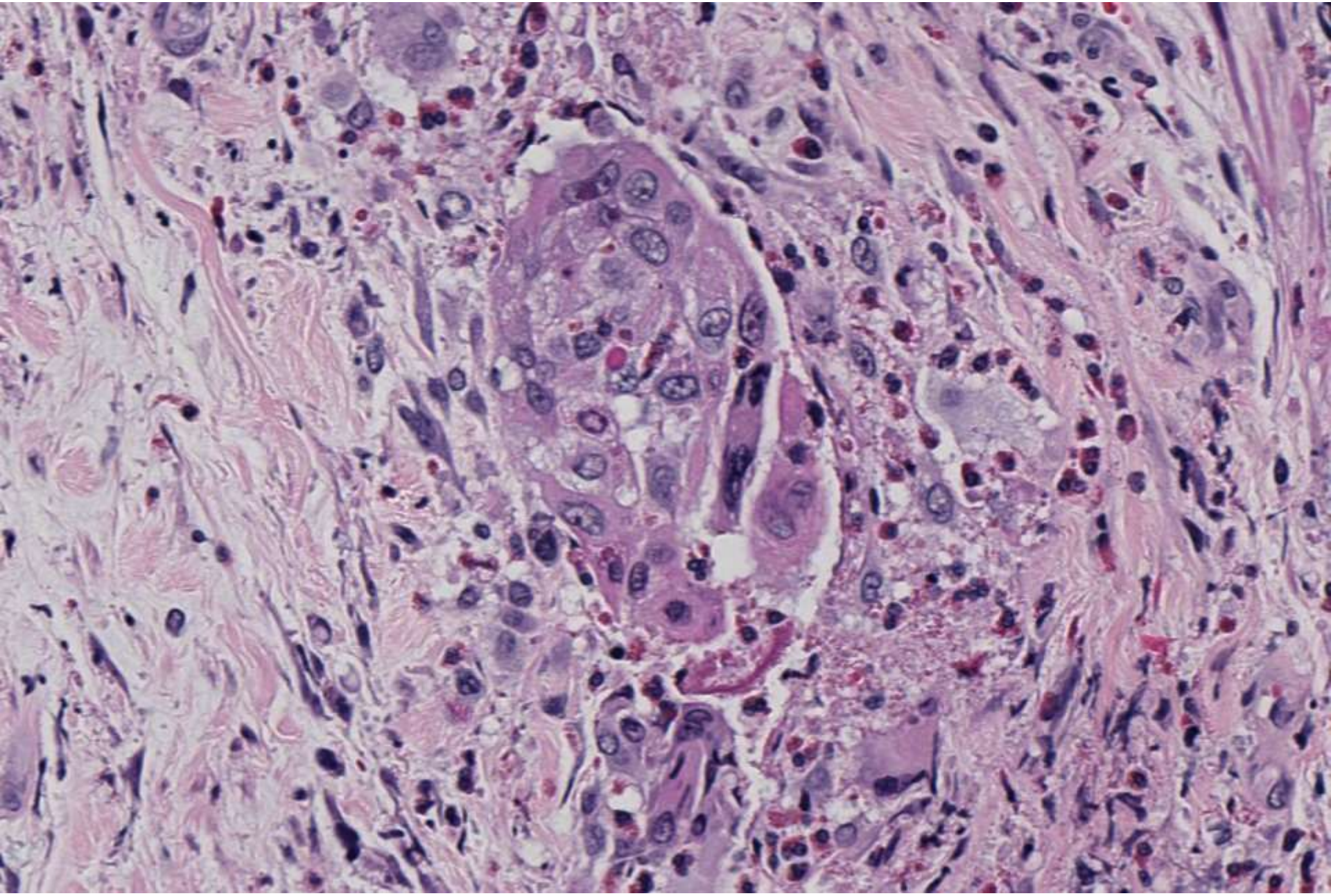




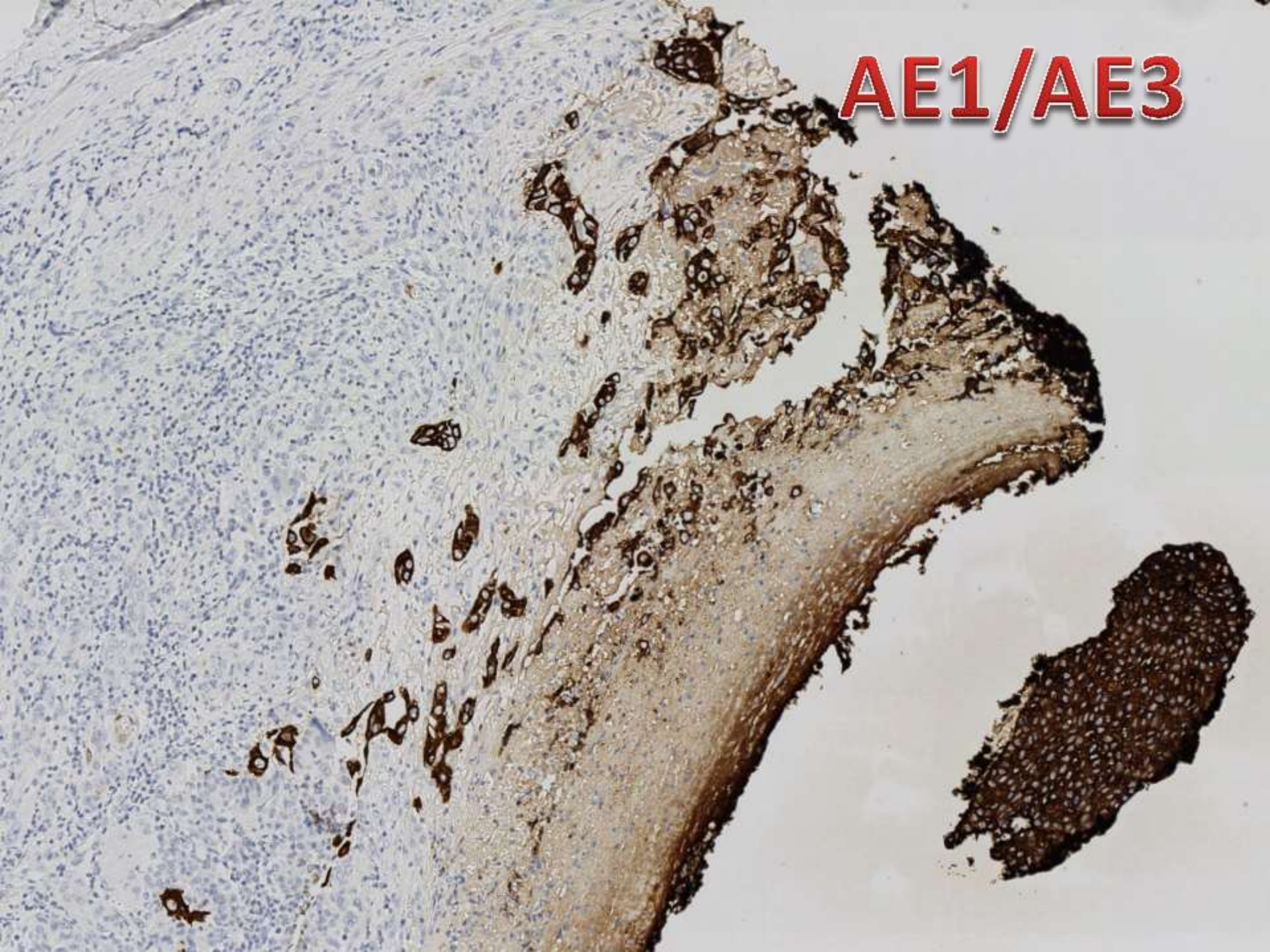









AE1/AE3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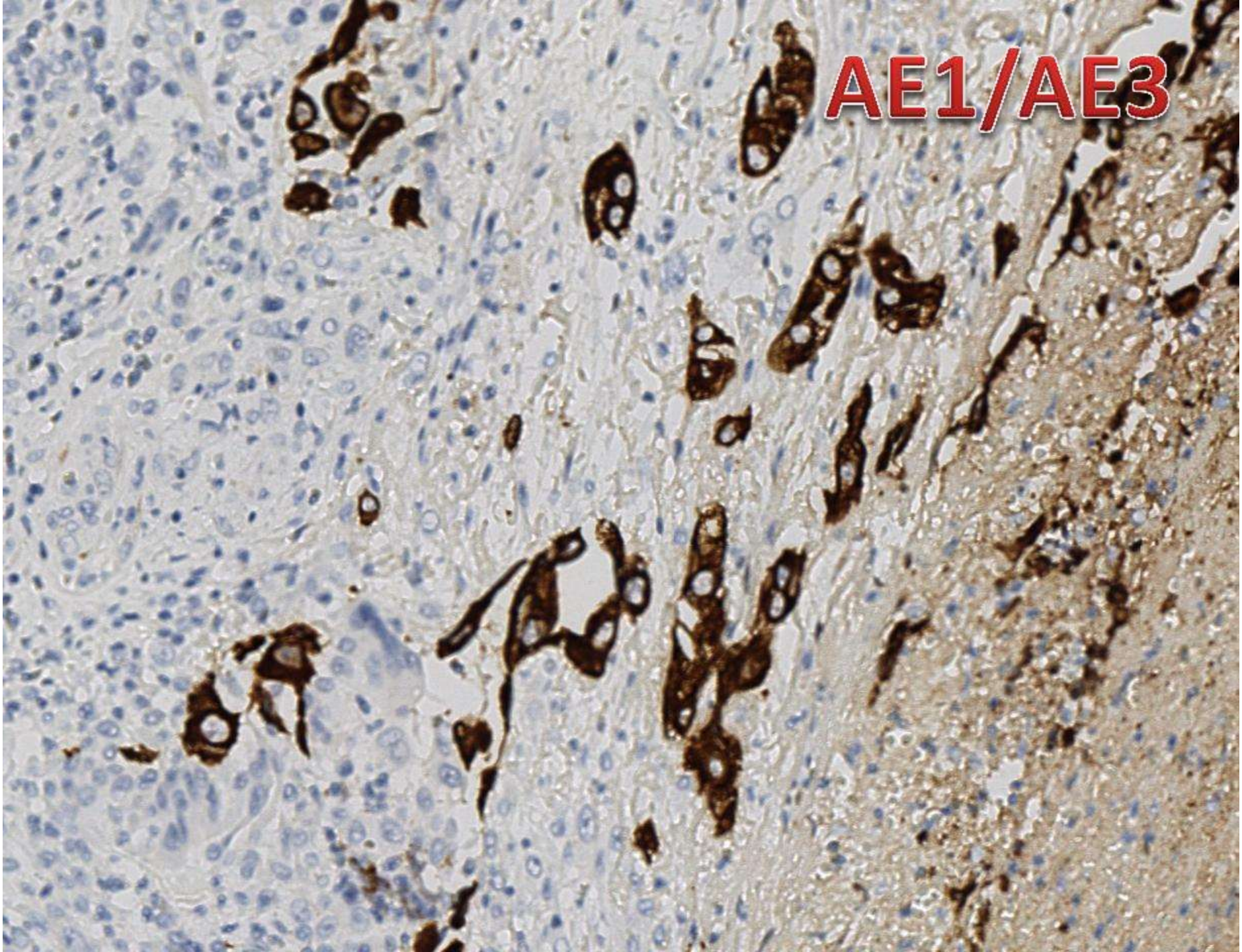
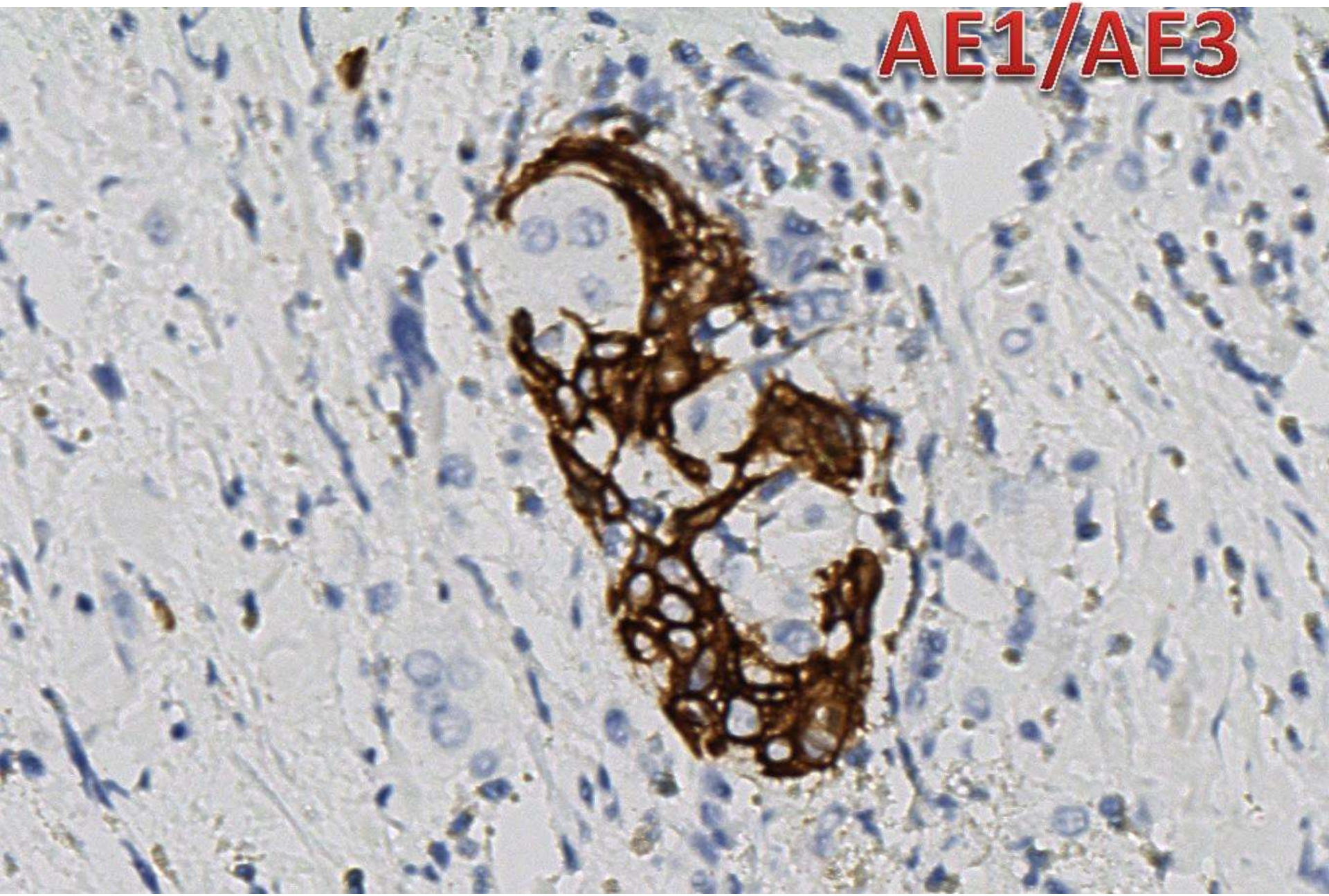

AE1/AE3


AE1/AE3


AE1/AE3

DIAGNOSIS?

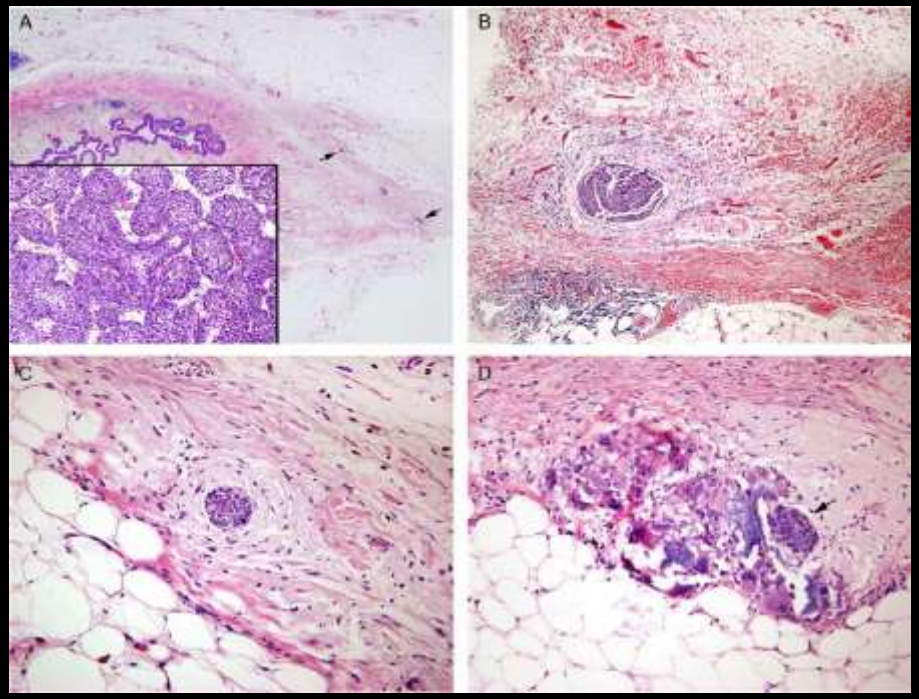
FINAL DIAGNOSIS:

Mechanical Implantation of Urothelium Into Periureteral Soft Tissue

A Series of 4 Cases Mimicking High-stage Urothelial Carcinoma

Sara E. Wobker, MD, MPH, Manju Aron, MD,† and Jonathan I. Epstein, MD‡§‖*

Am J Surg Pathol • Volume 40, Number 11, November 2016



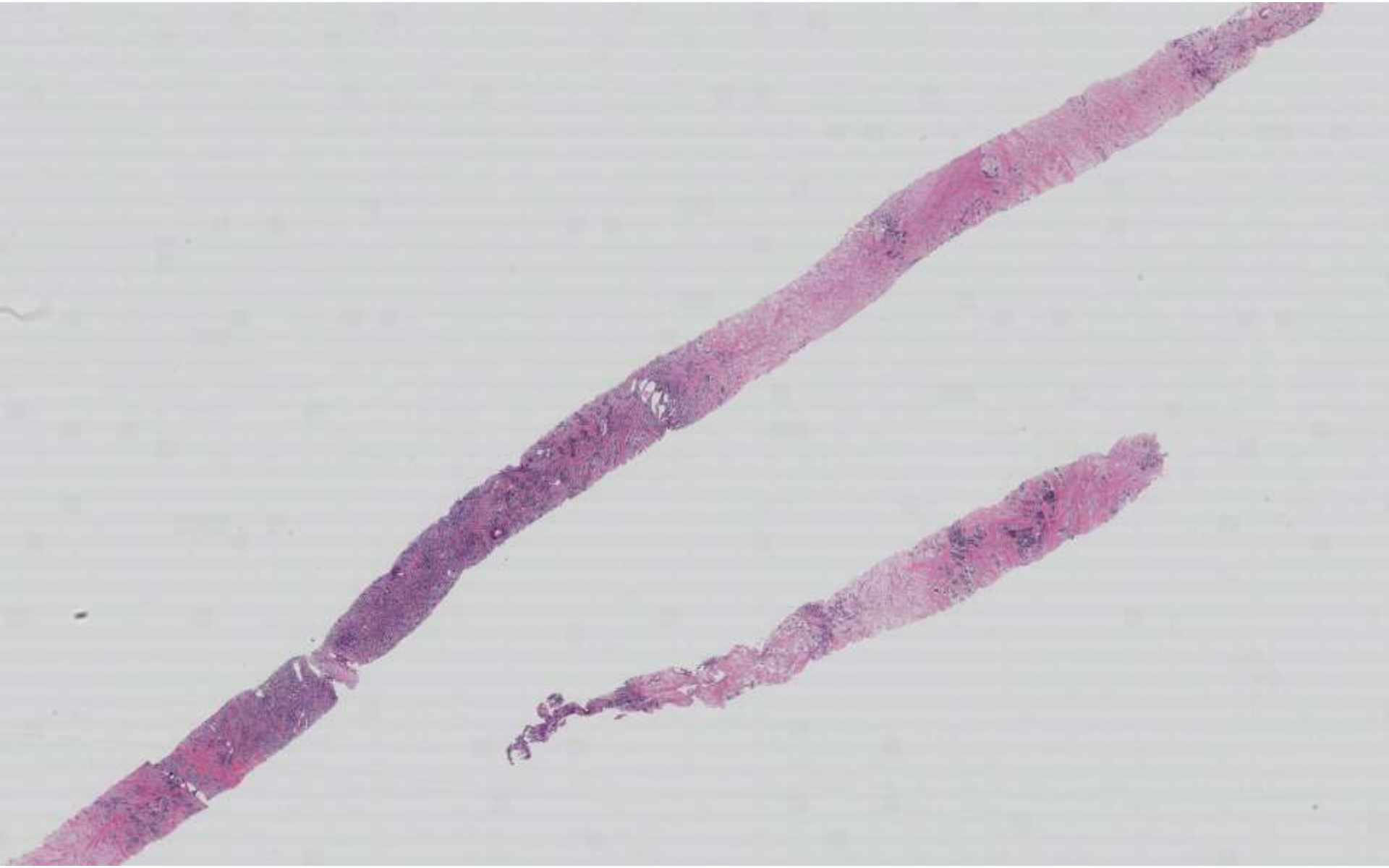
TAKE HOME POINTS

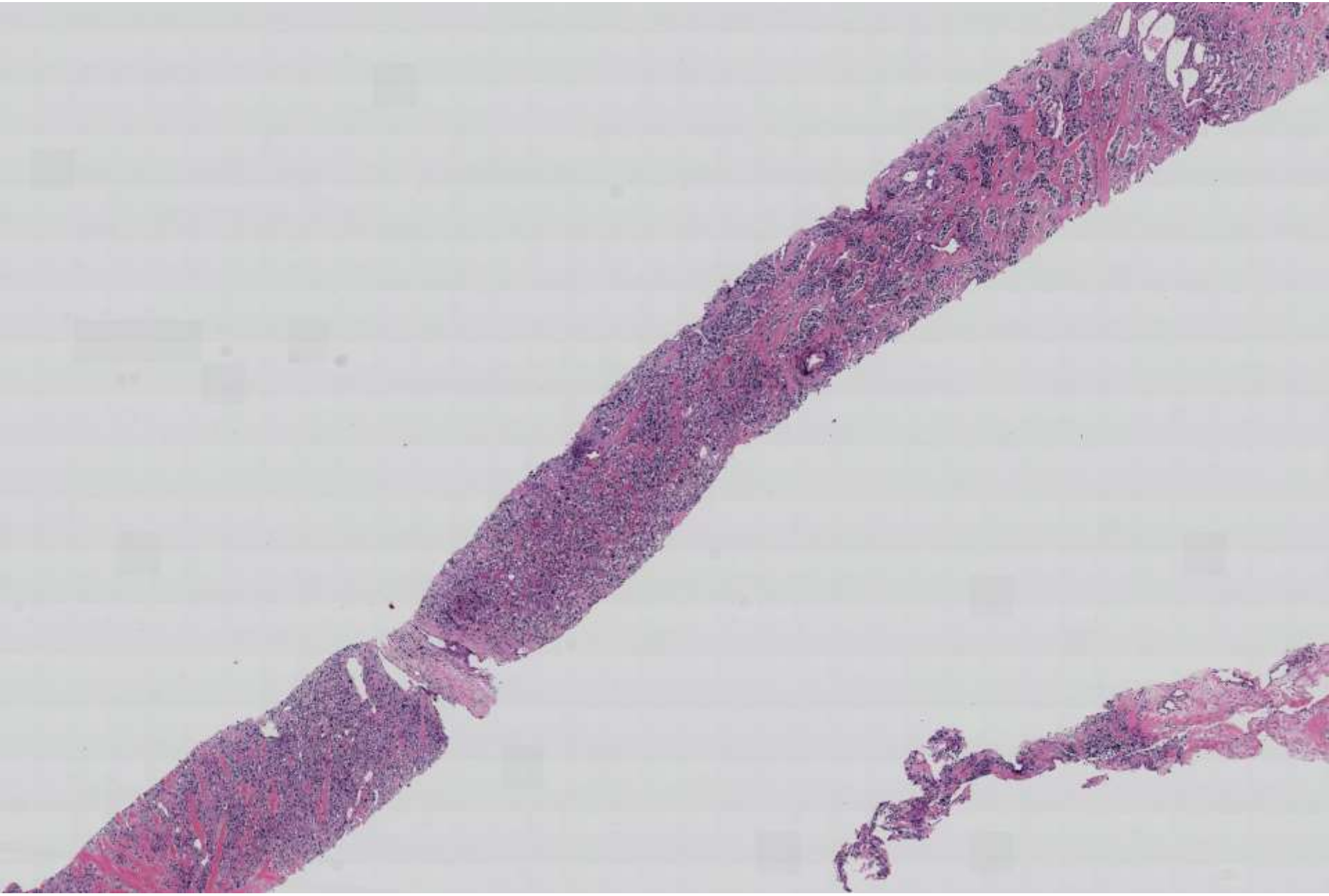
- **In setting of low grade upper tract tumors with bland nests of urothelium beyond muscularis**
  - Inquire about prior ureteral biopsy or stent
    - “mechanical implantation” vs pT3 disease

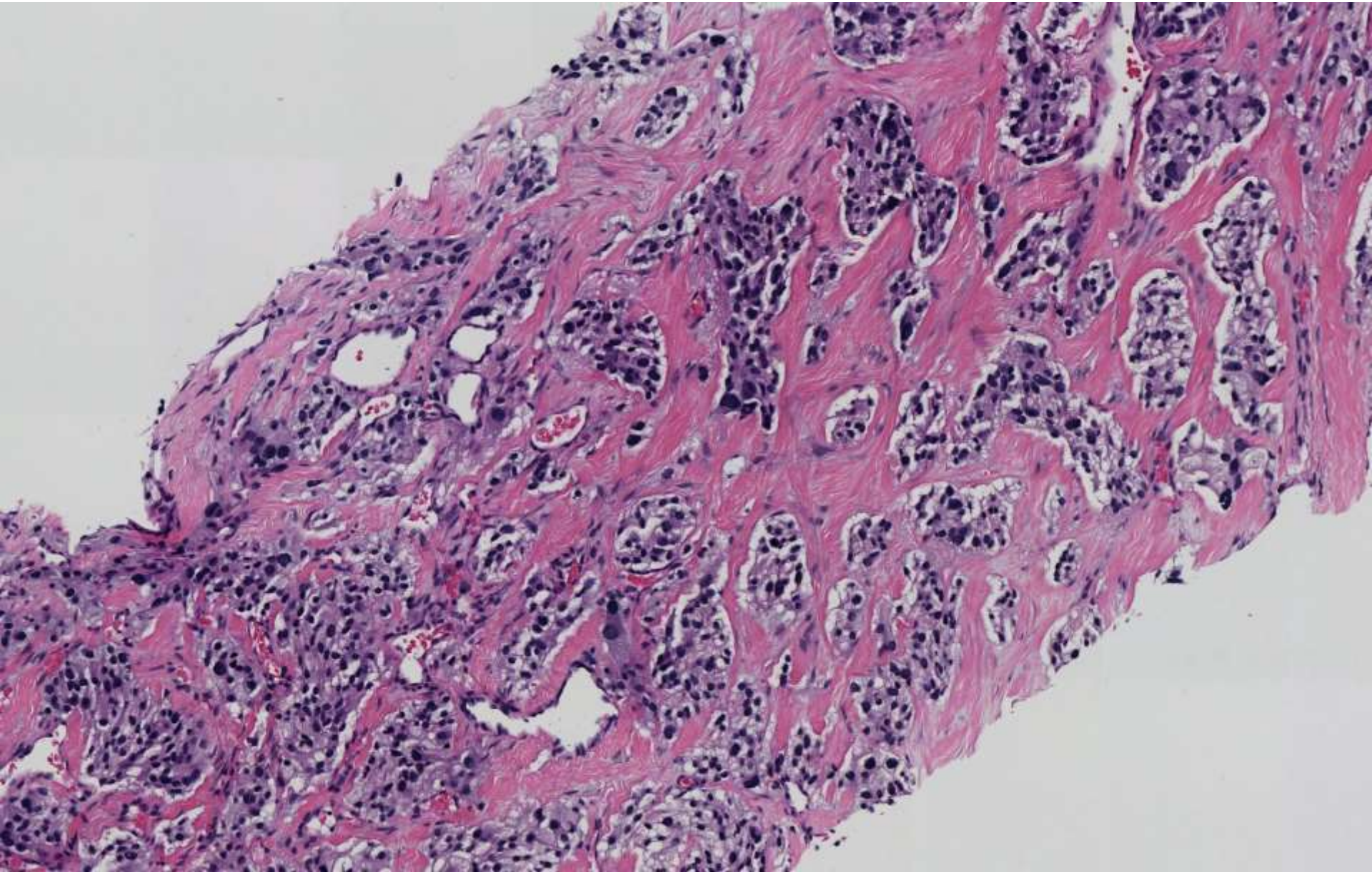
SB 6179
(scanned slide available)

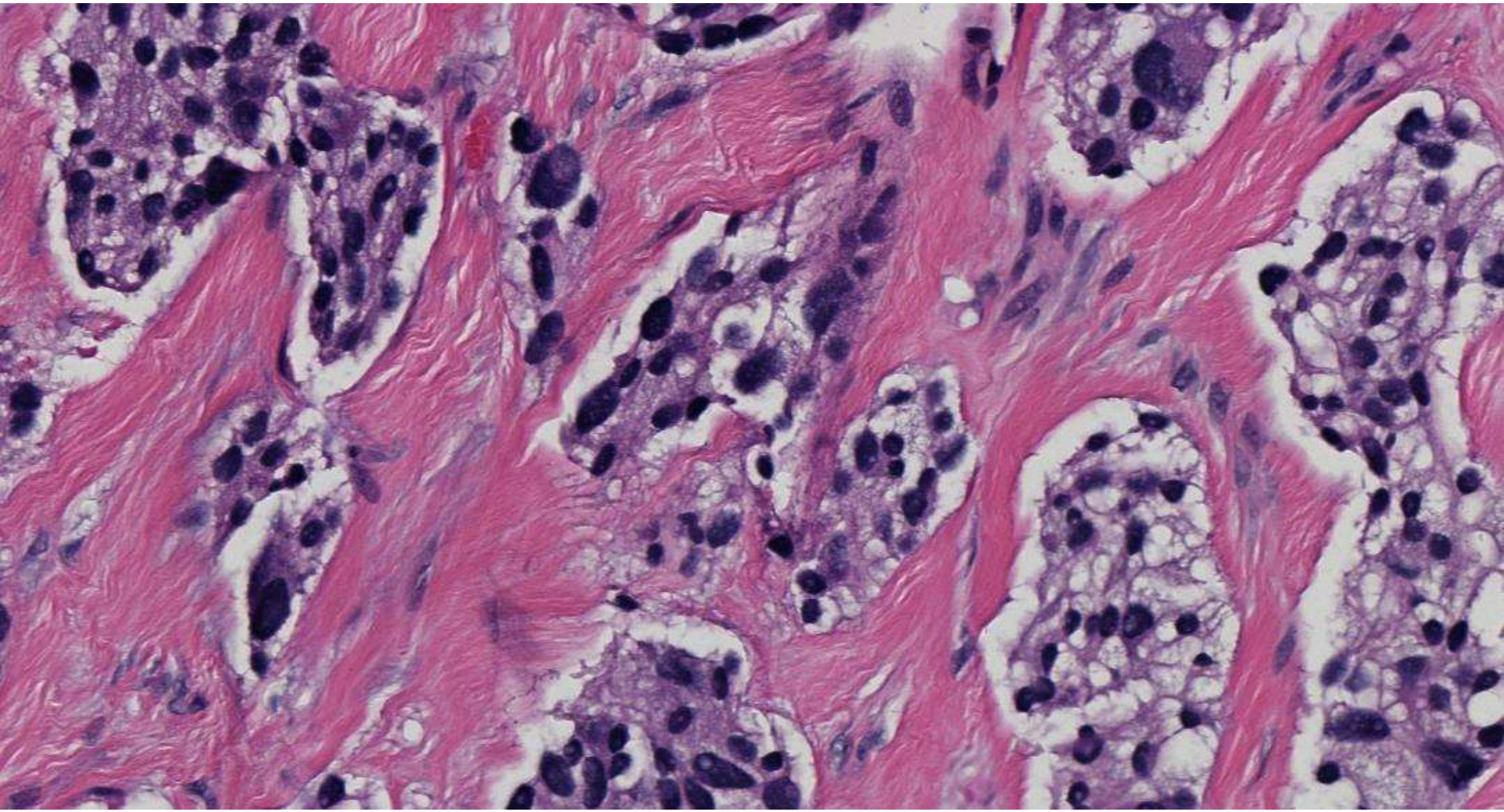
Ankur Sangoi; El Camino Hospital

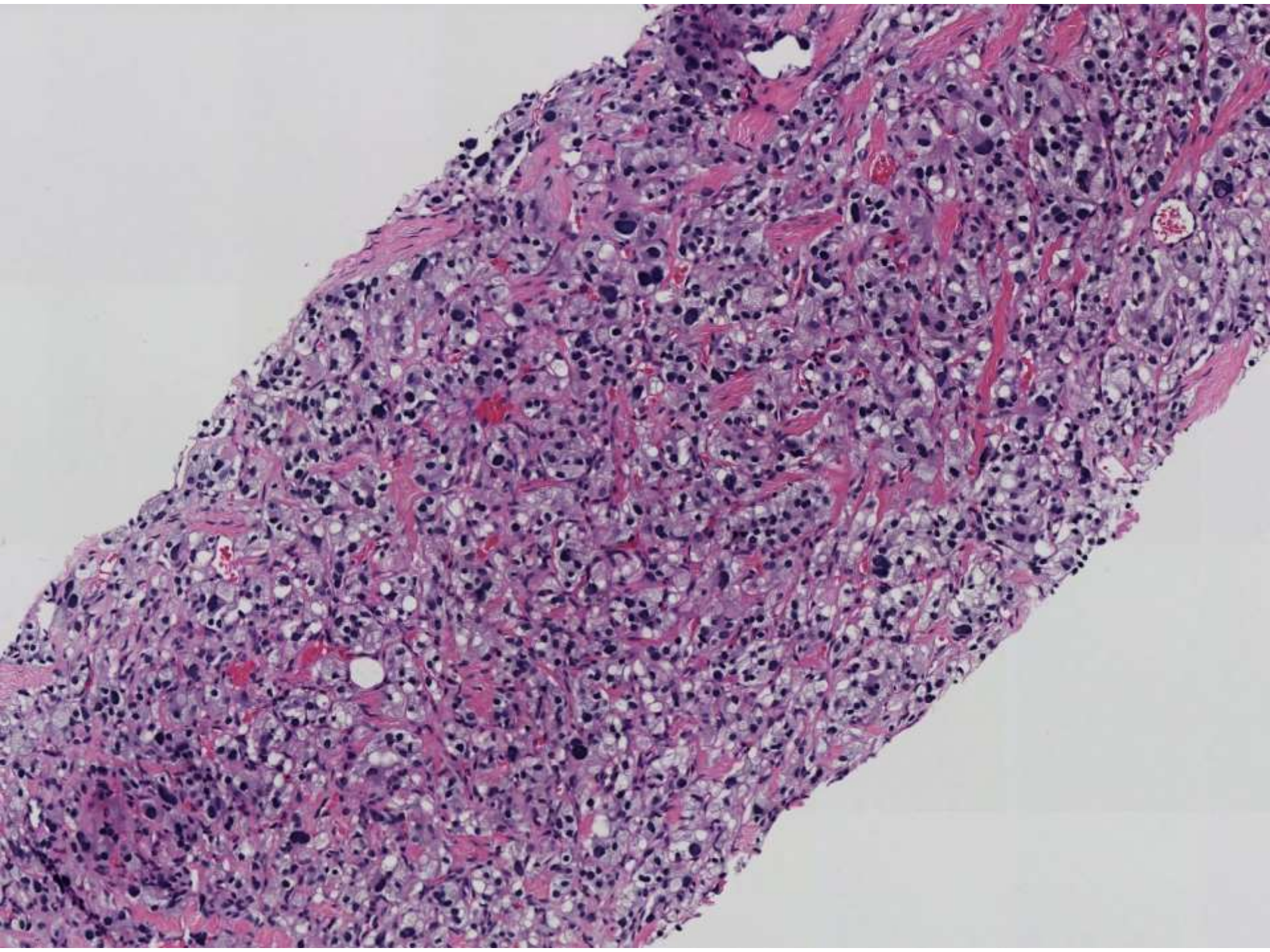
83-year-old woman with history of breast cancer, now presents with adrenal gland mass.

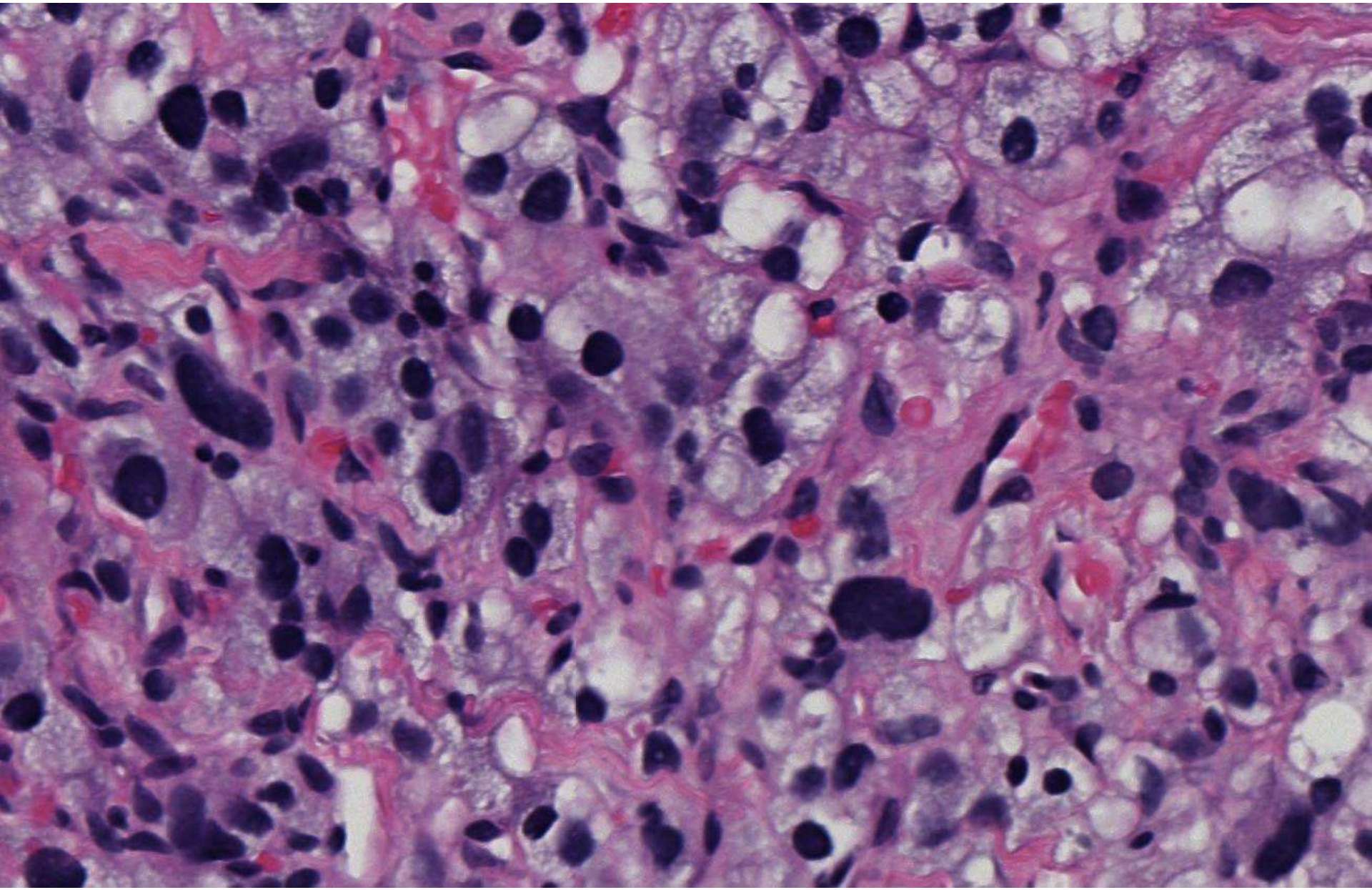








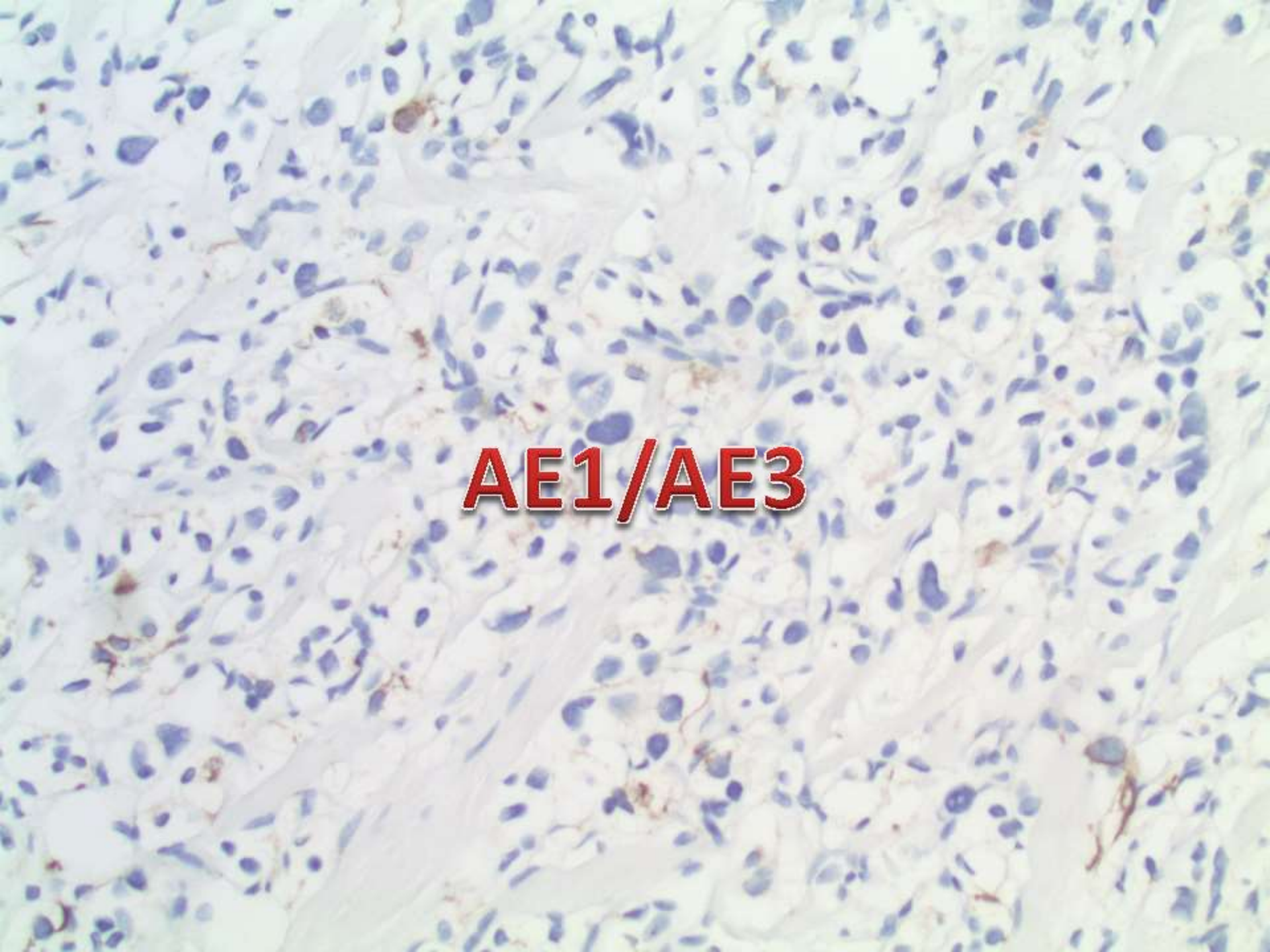


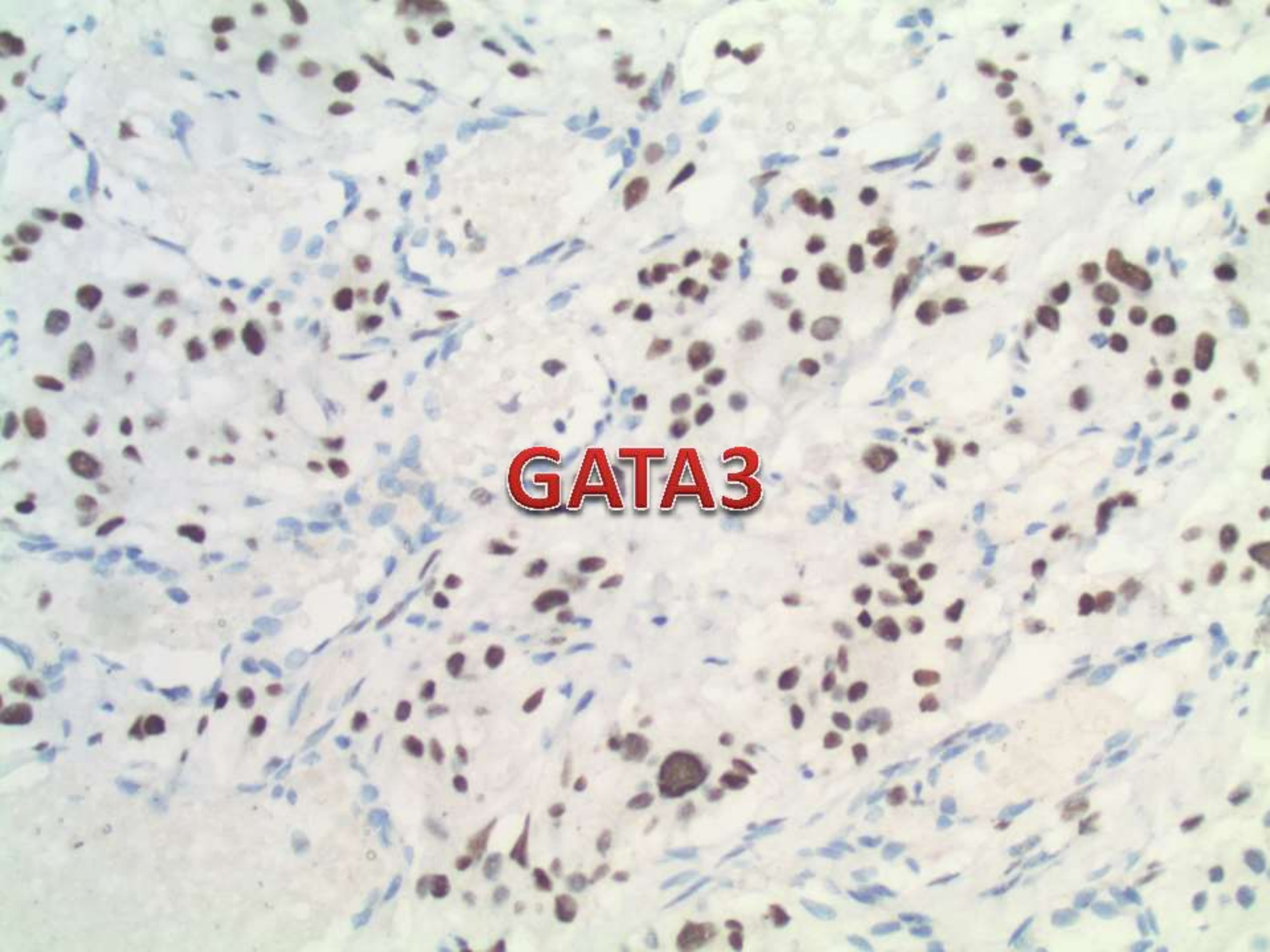


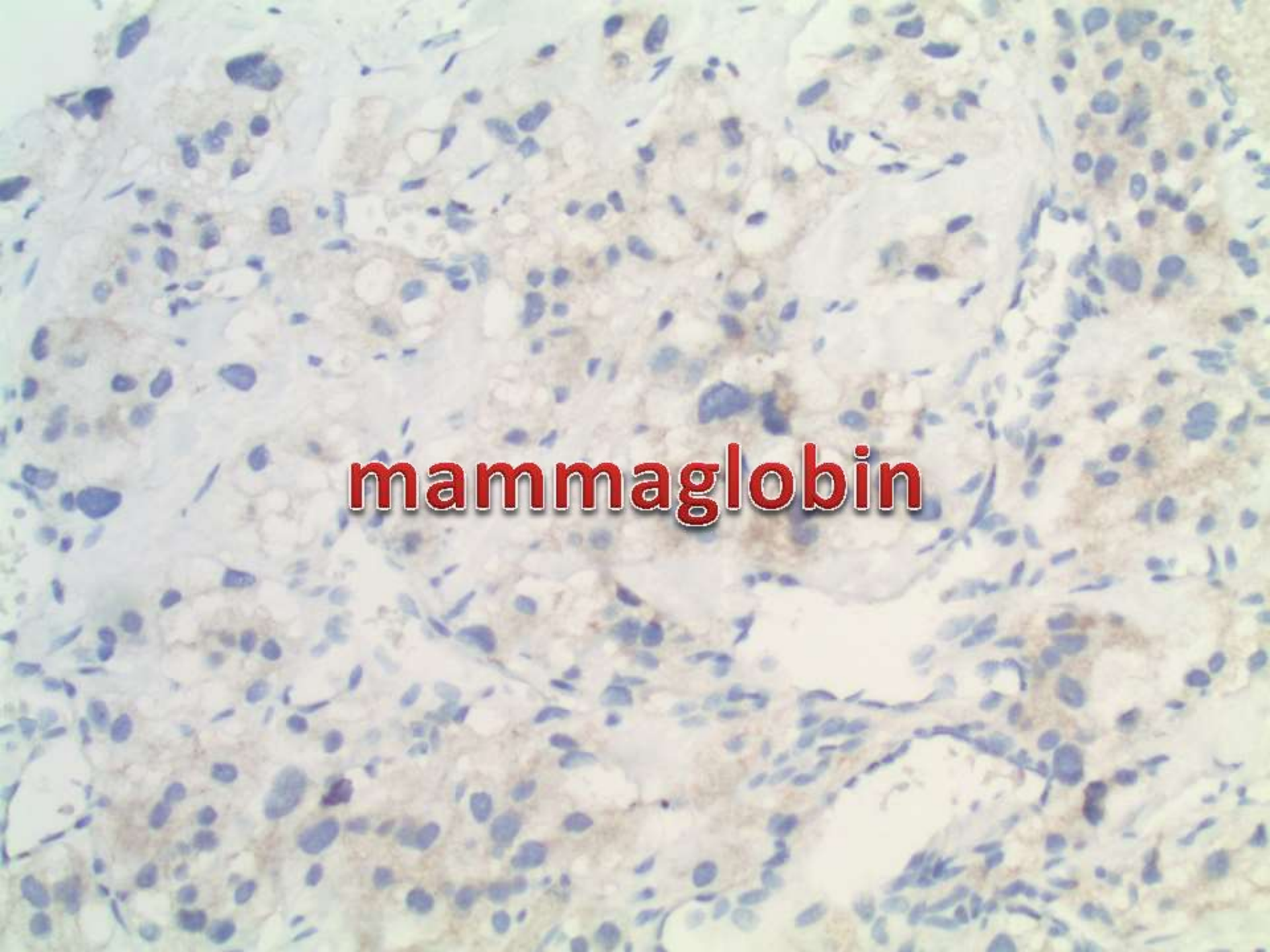
DIAGNOSIS?

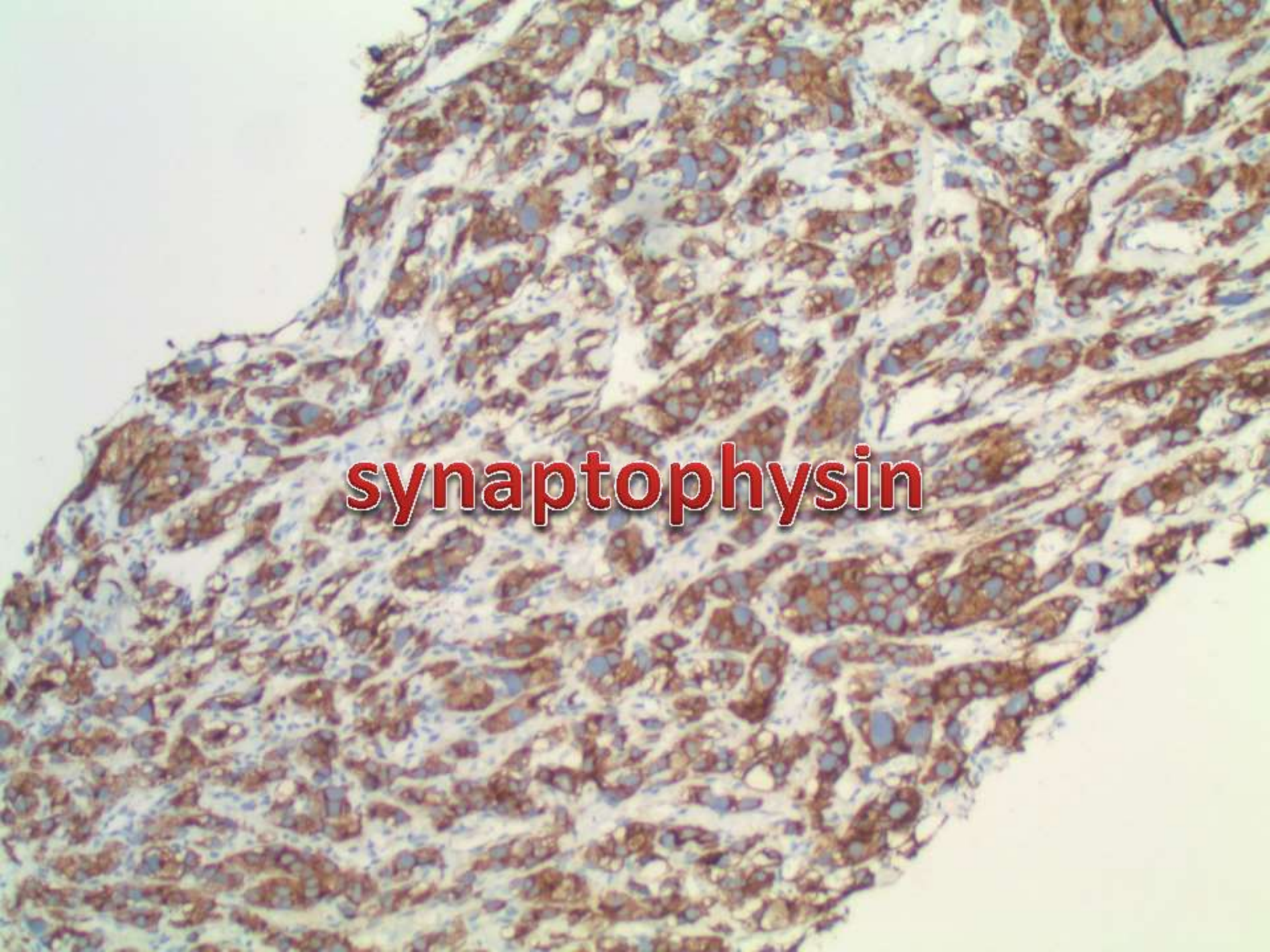
DDx

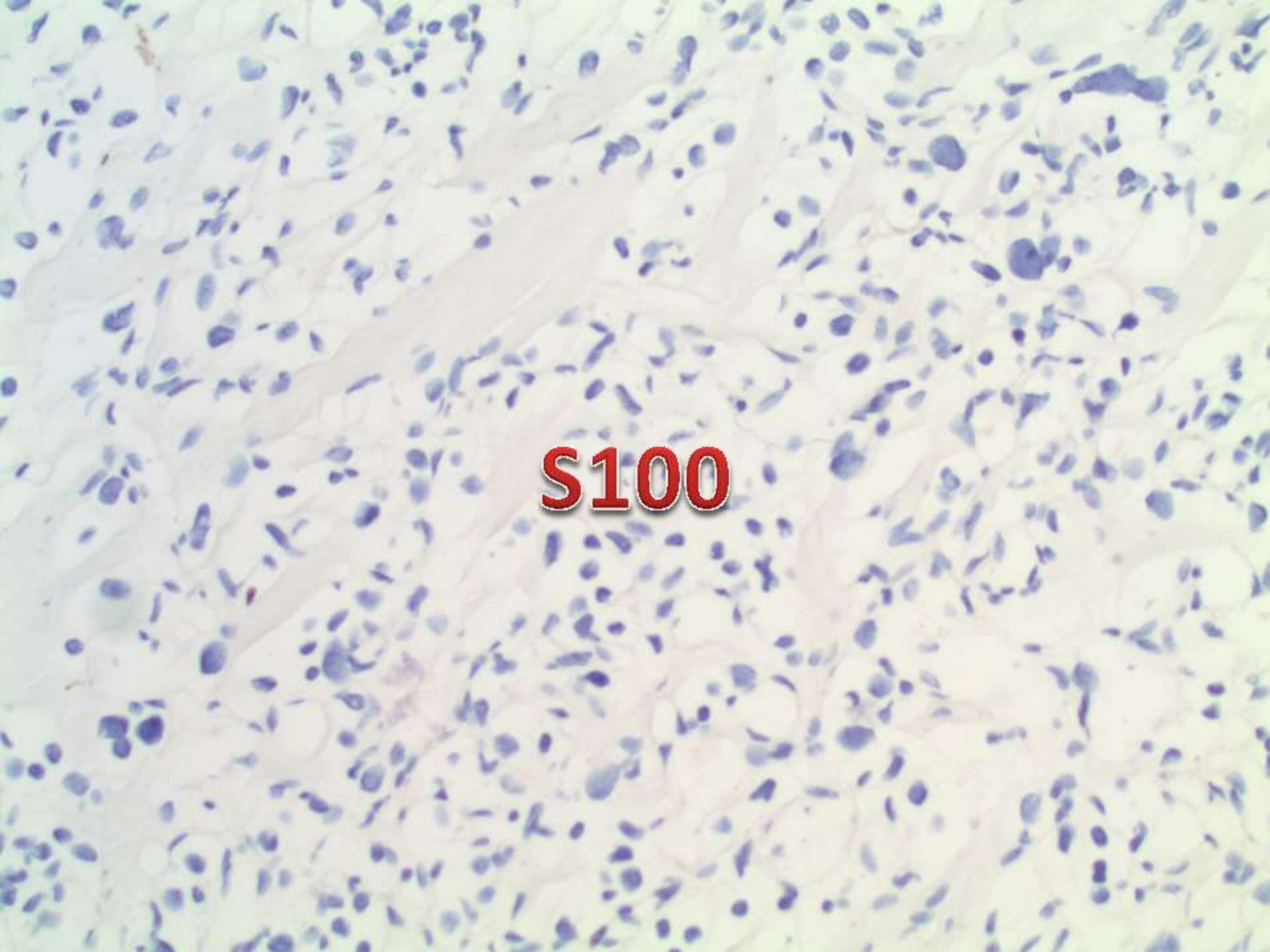
- Metastatic carcinoma
- Adrenal cortical neoplasm
- Pheochromocytoma
- Melanoma
- PEComa


AE1/AE3


GATA3


mammaglobin

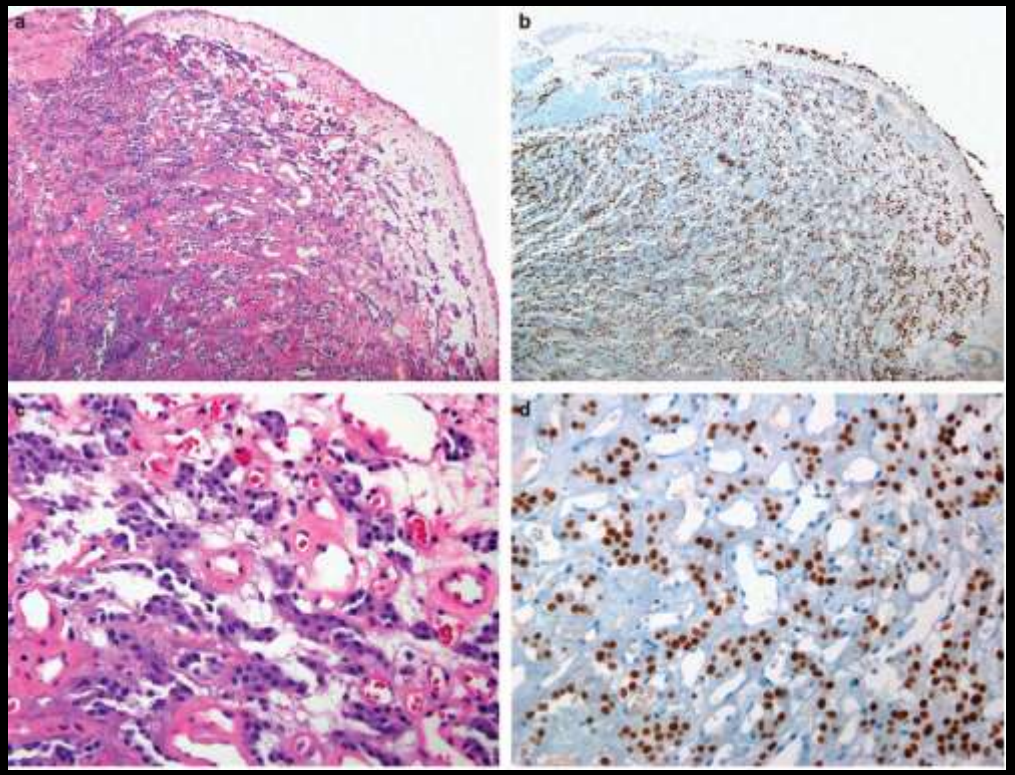

synaptophysin


S100

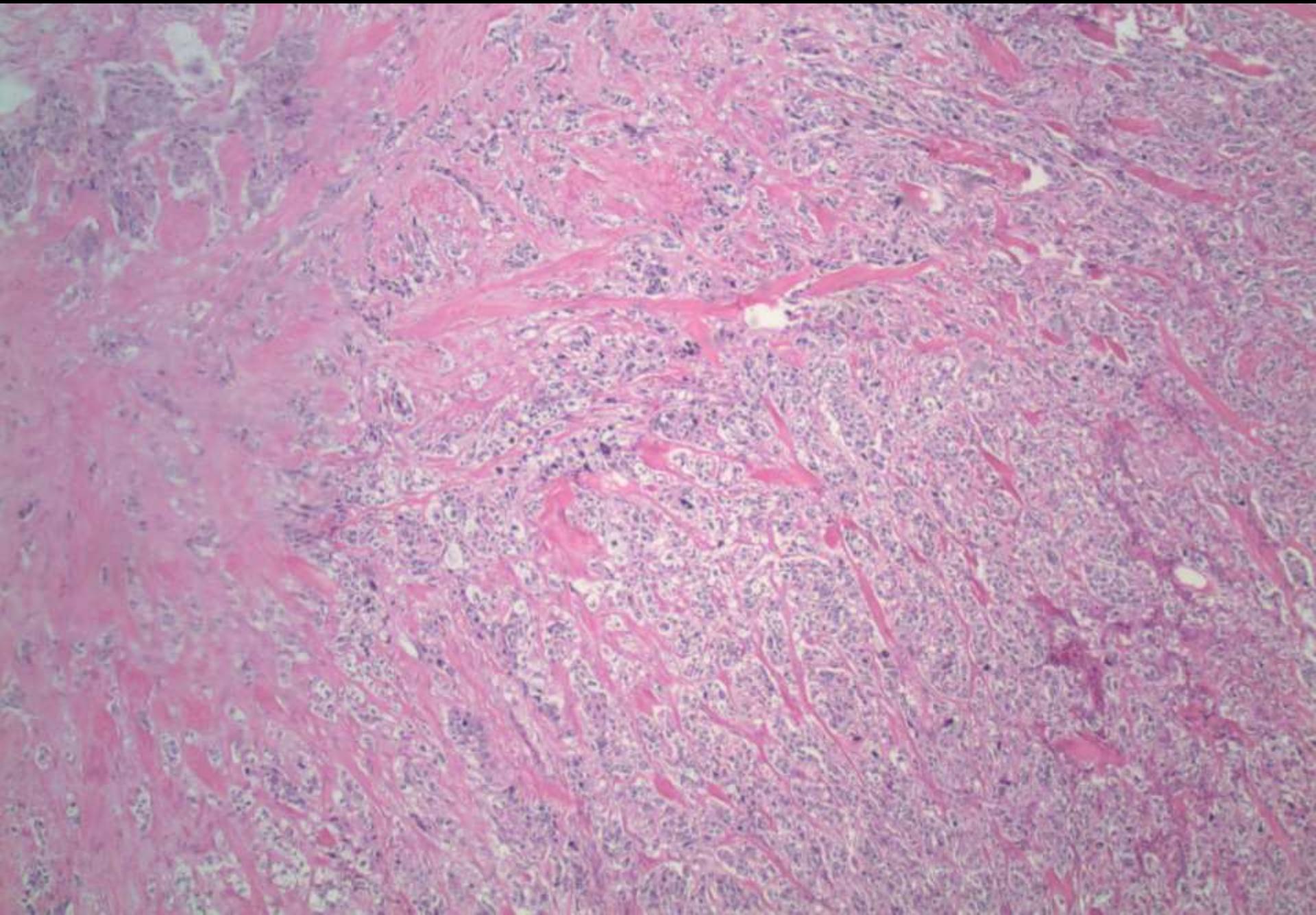
DIAGNOSIS?

GATA3 expression in paragangliomas: a pitfall potentially leading to misdiagnosis of urothelial carcinoma

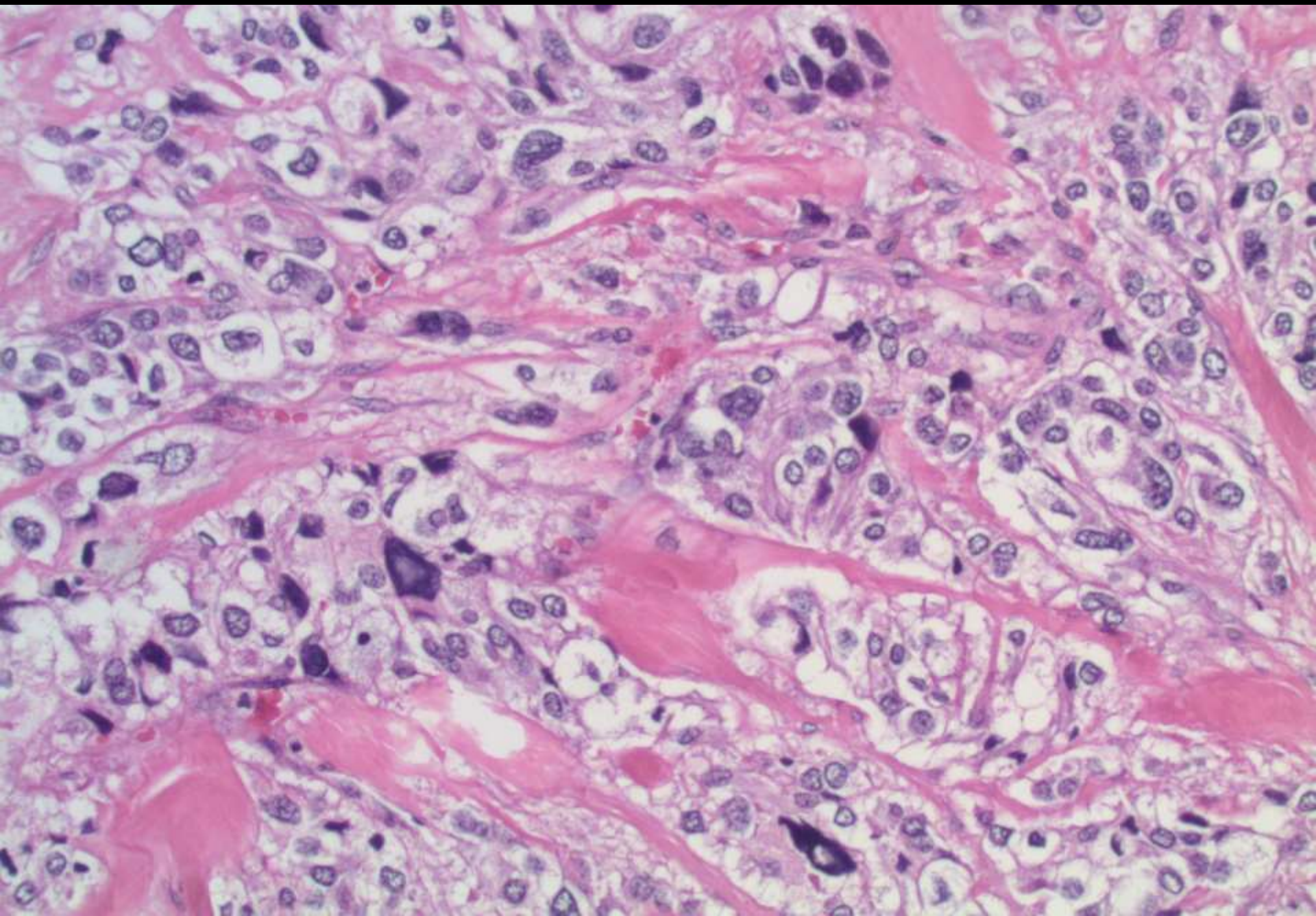
Modern Pathology (2013) 26, 1365–1370



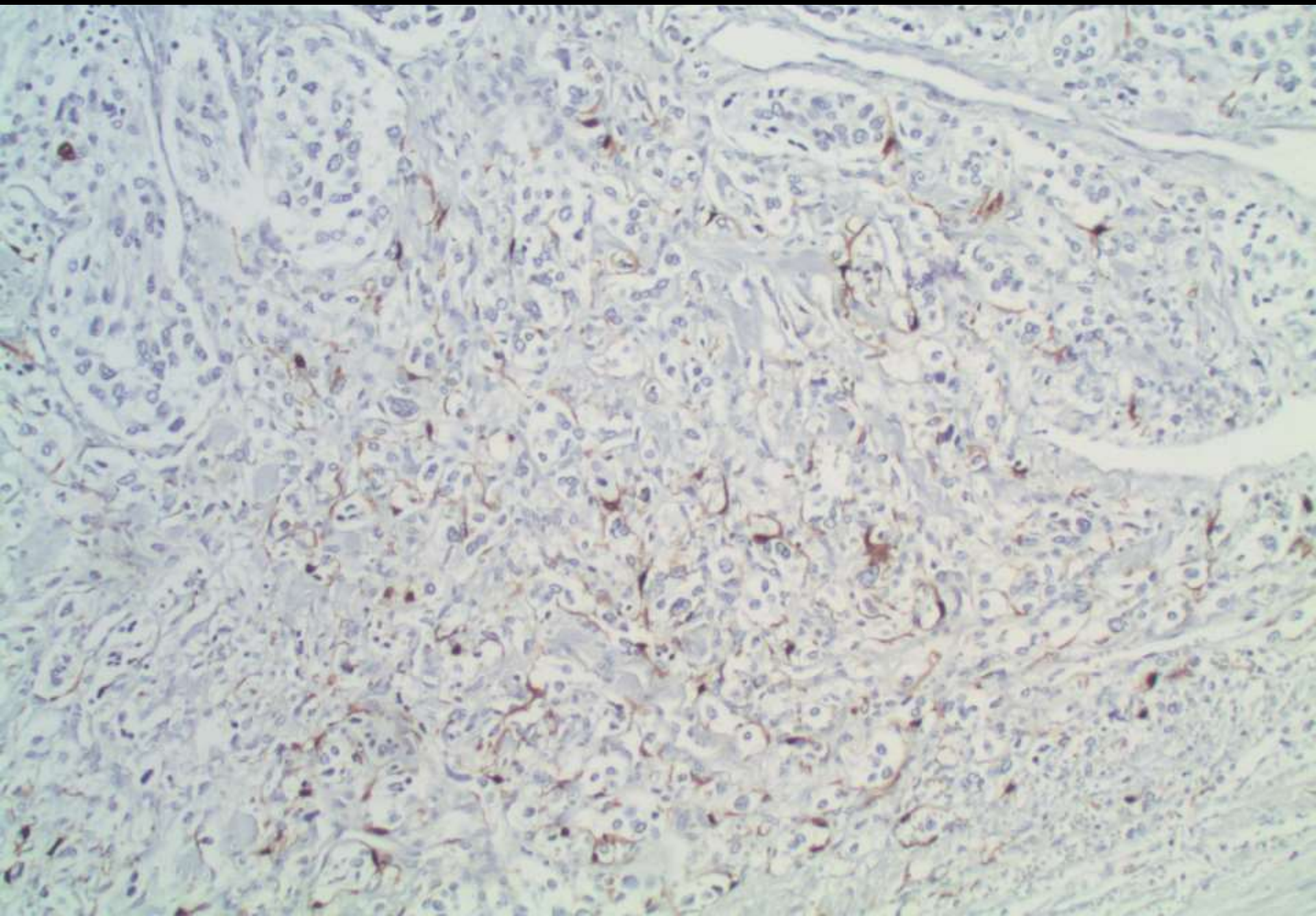
Adrenalectomy specimen: Pheochromocytoma



Adrenalectomy specimen: Pheochromocytoma



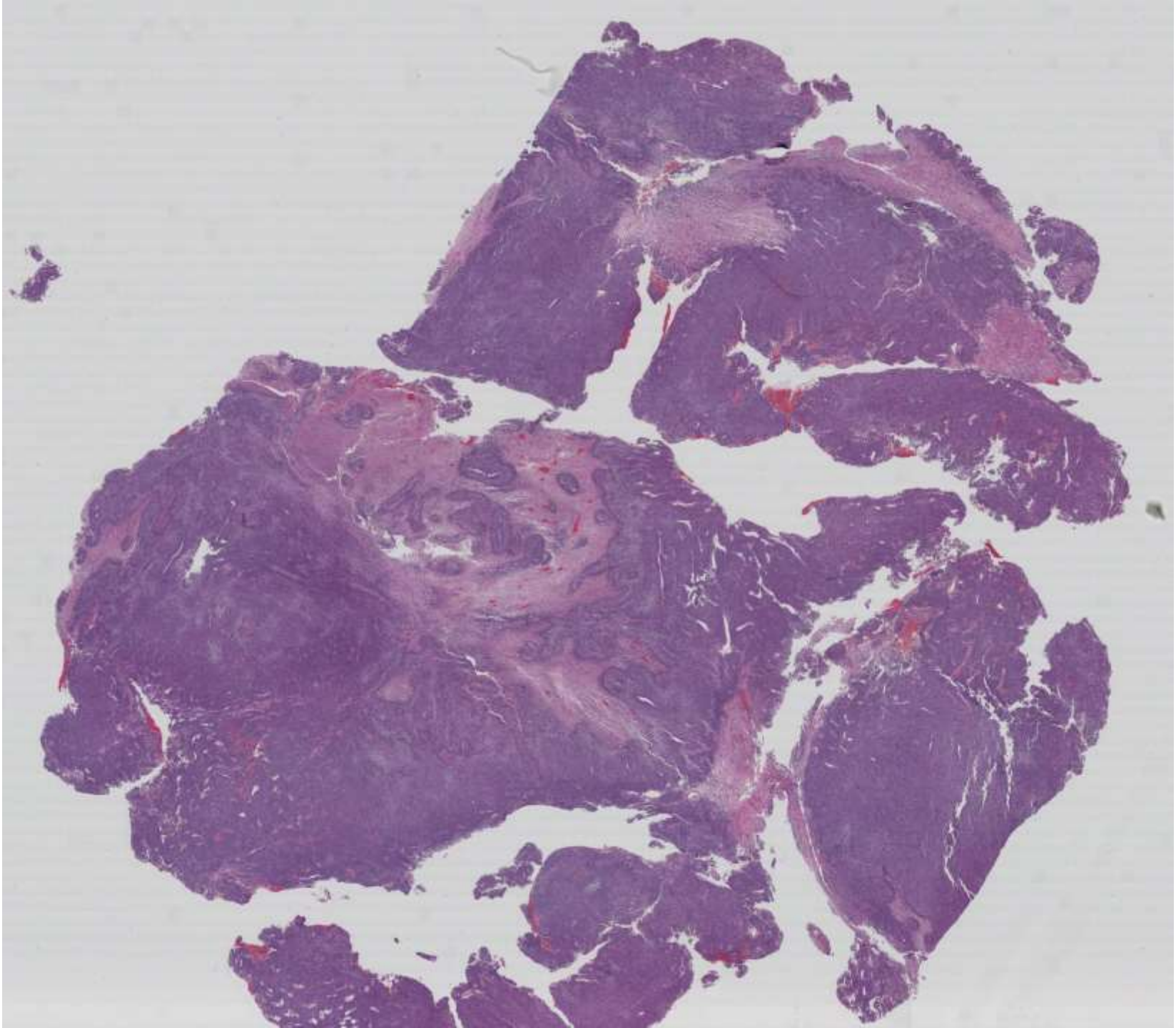
S100 on Adrenalectomy specimen: Pheochromocytoma

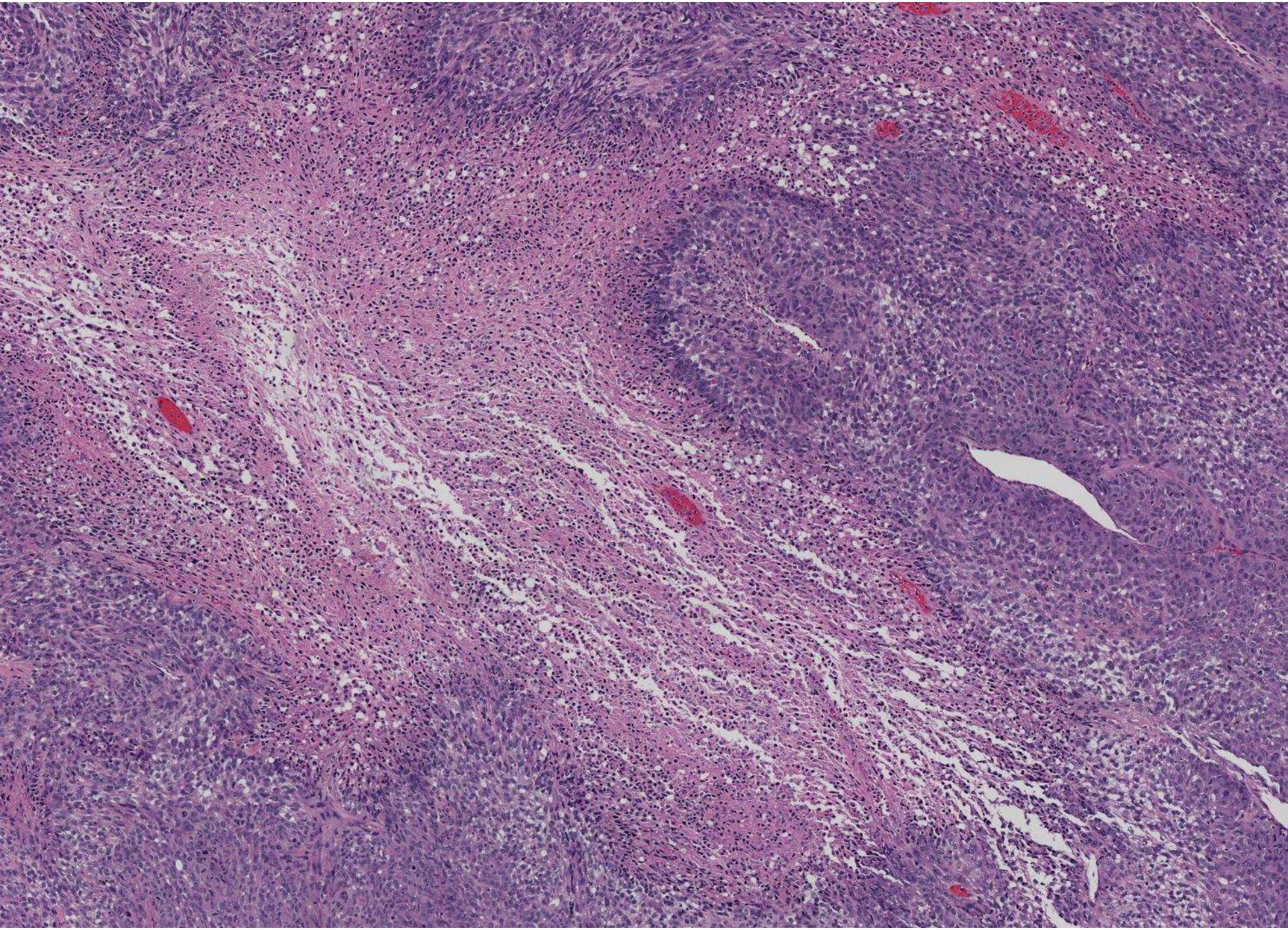


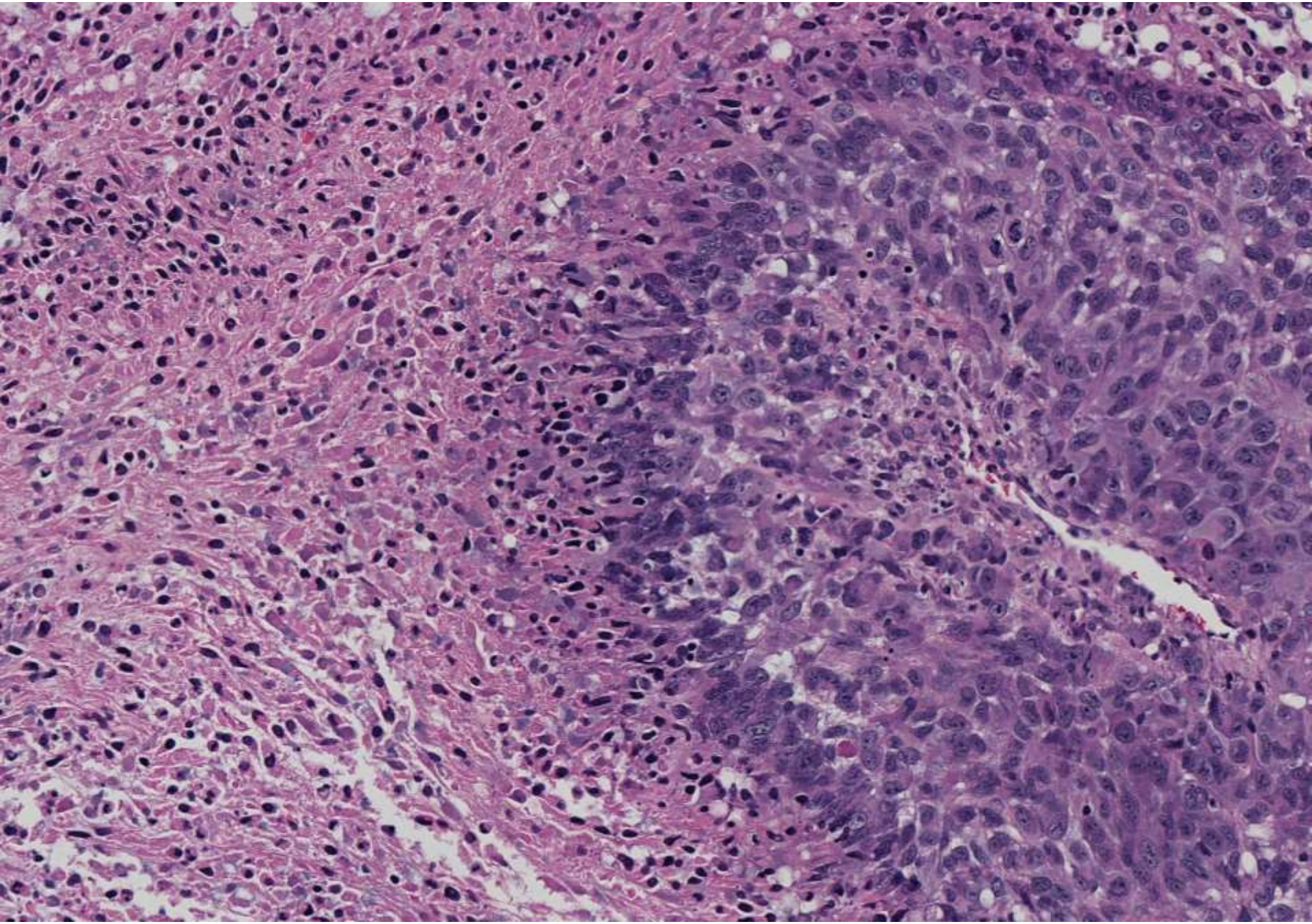
SB 6180
(scanned slide available)

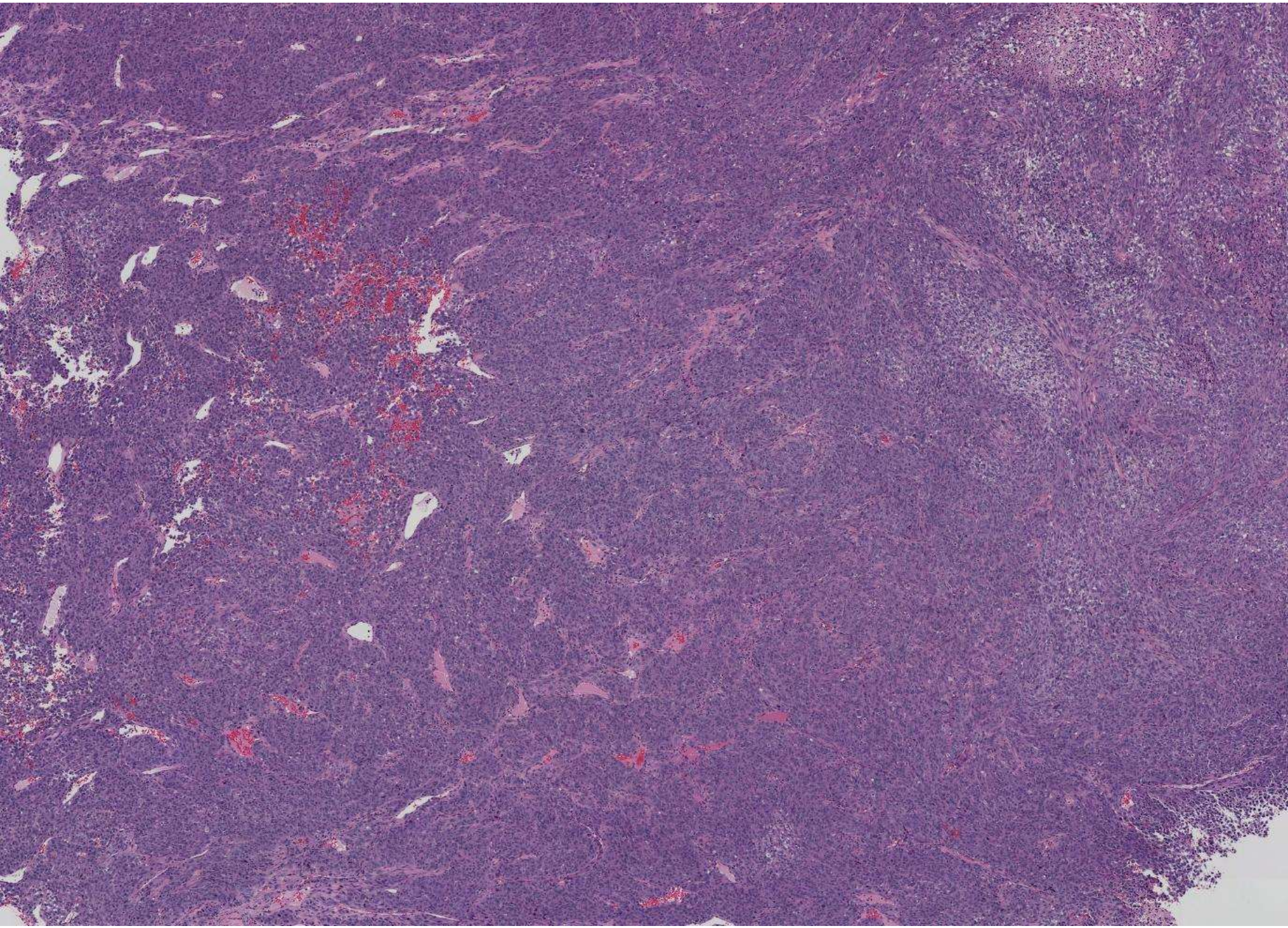
Ankur Sangoi; El Camino Hospital

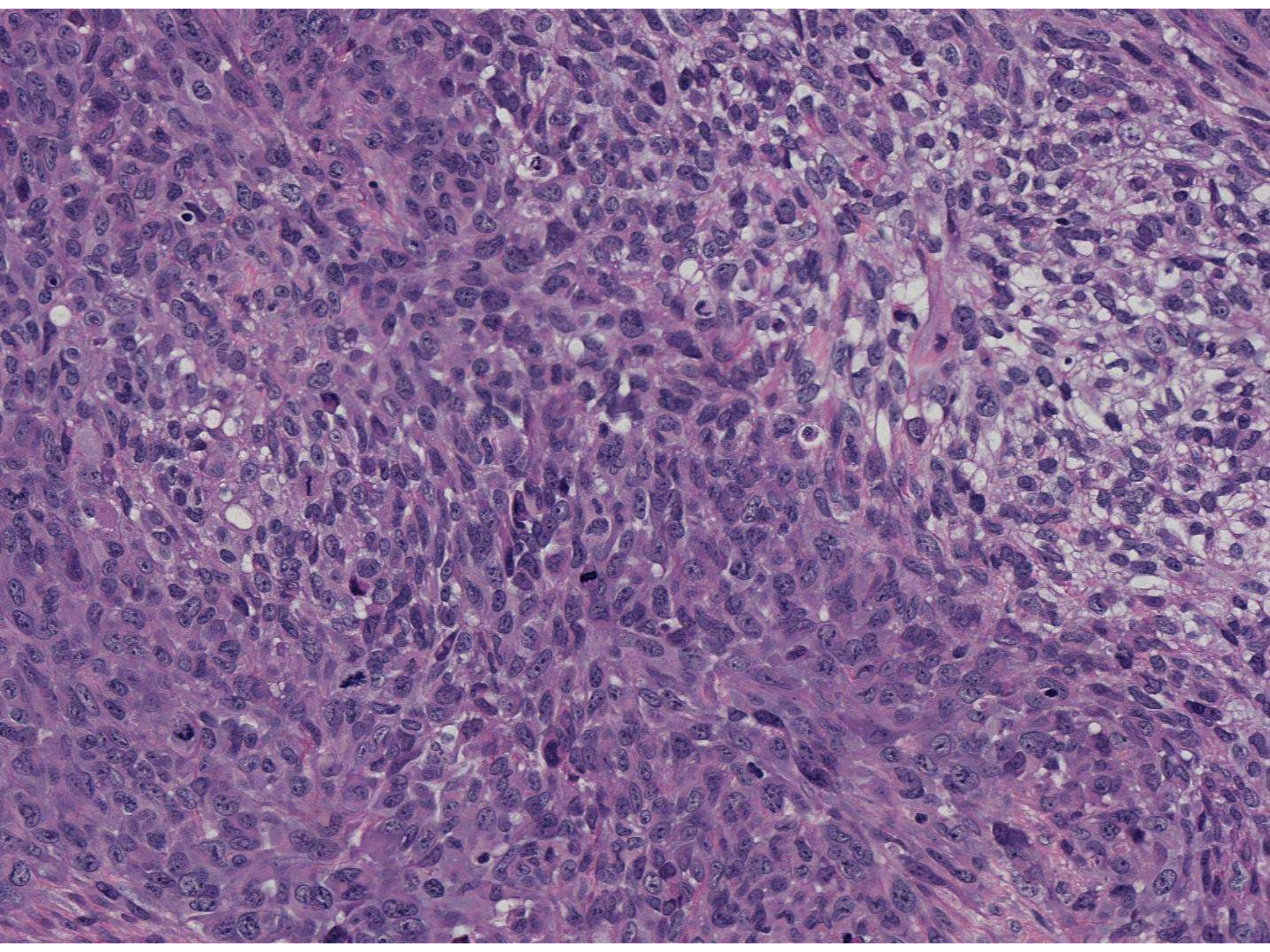
44-year-old man with pleural mass. One month prior he had a groin mass core biopsy of “poorly differentiated large cell carcinoma”.

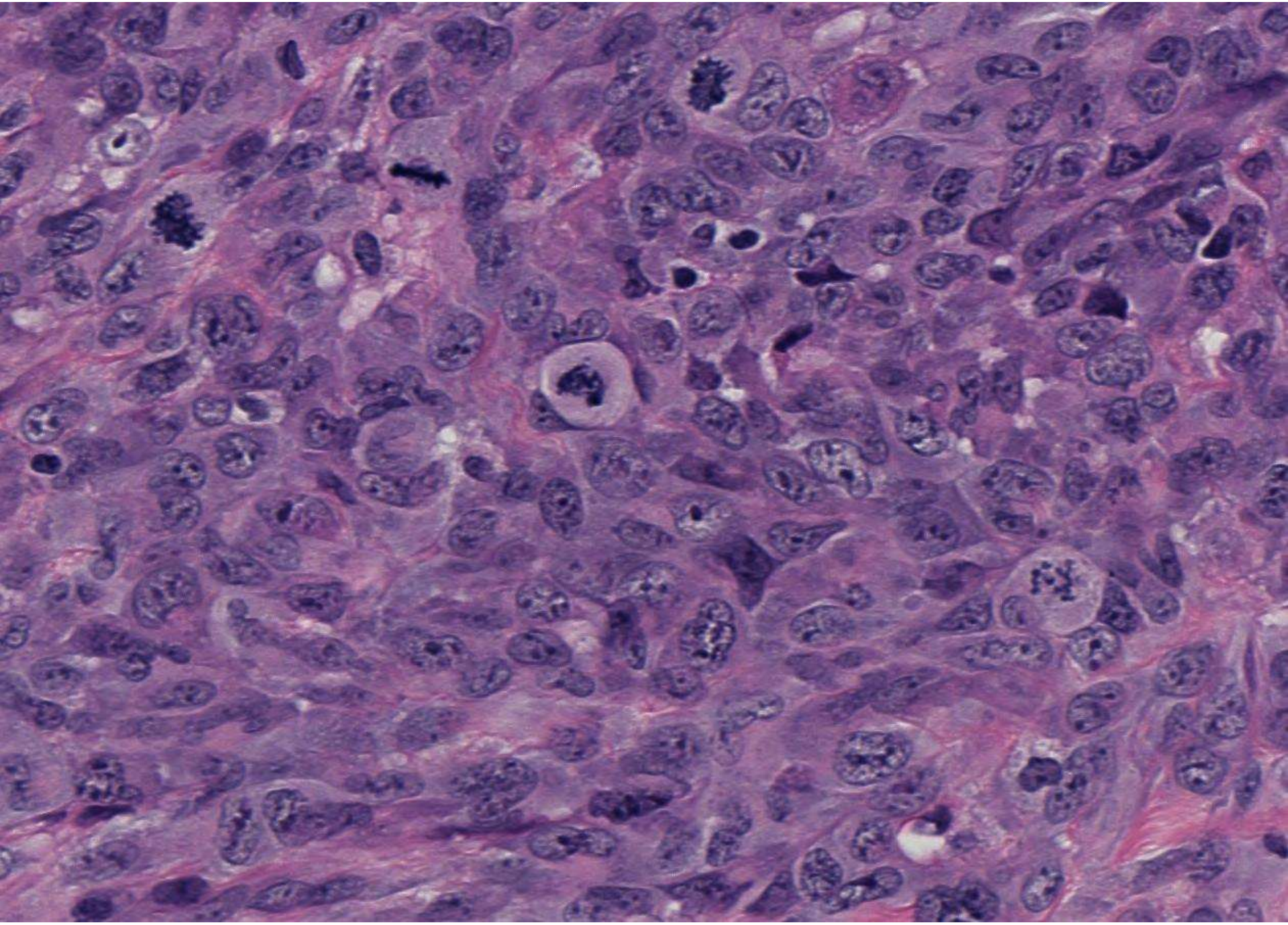


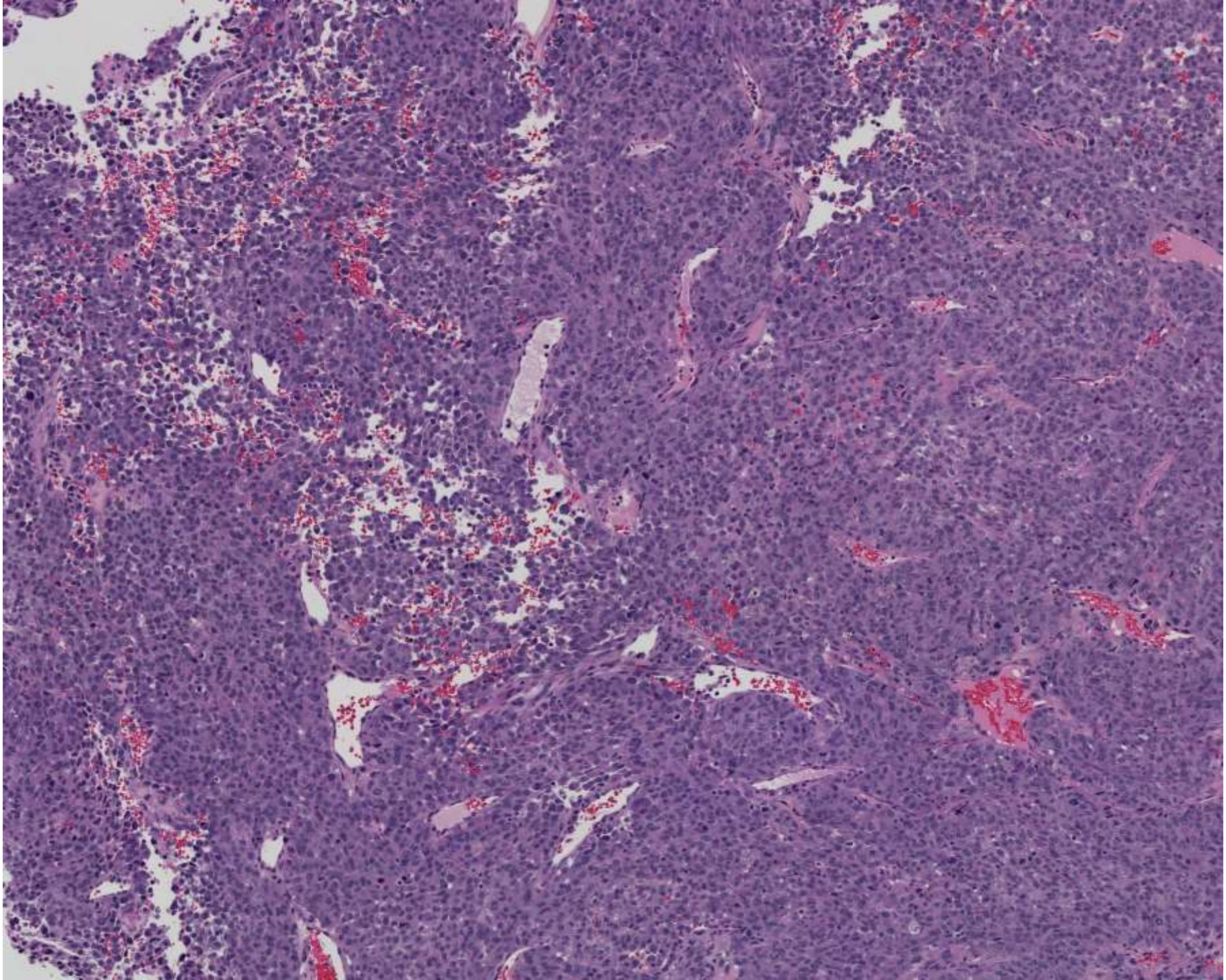


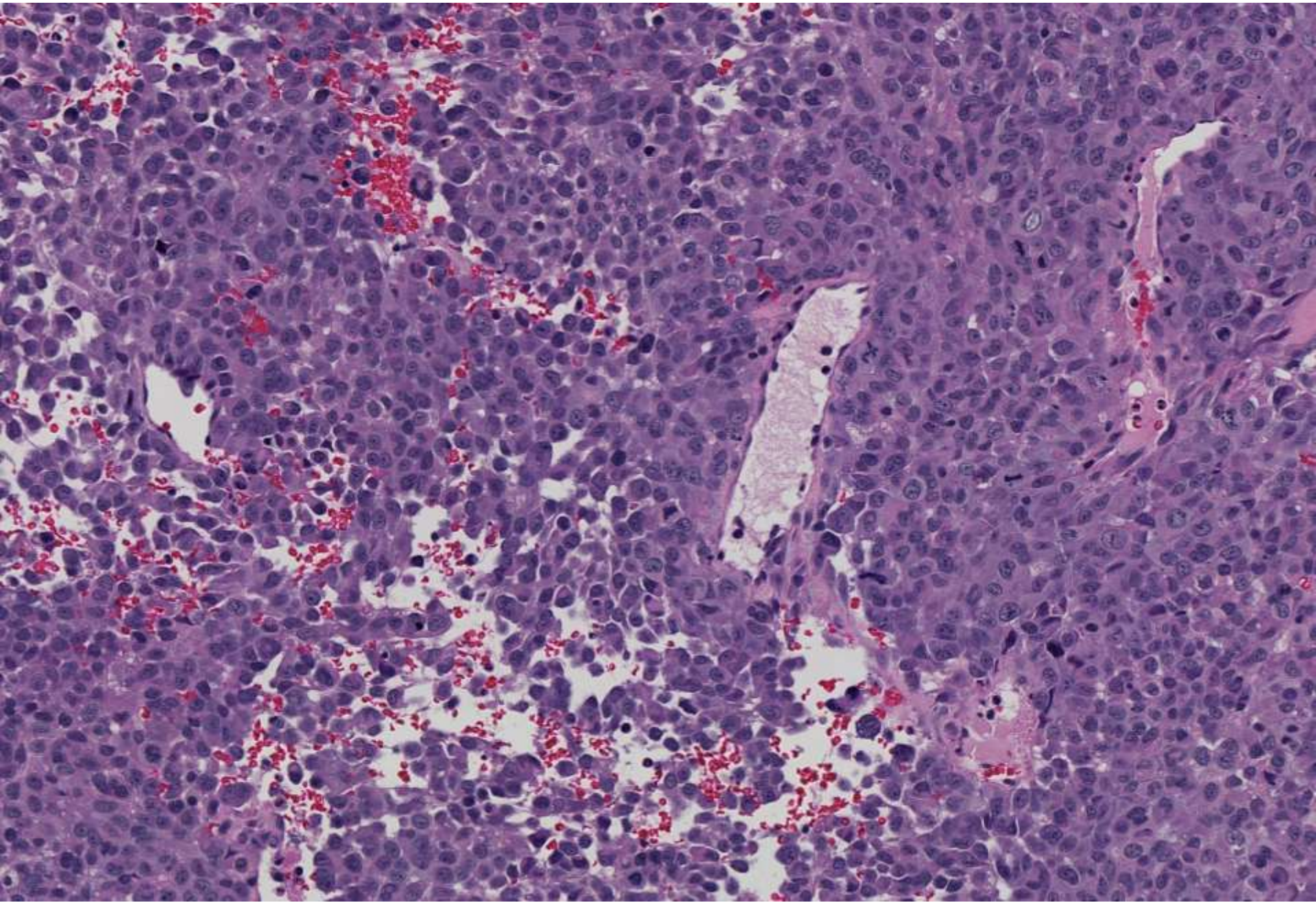


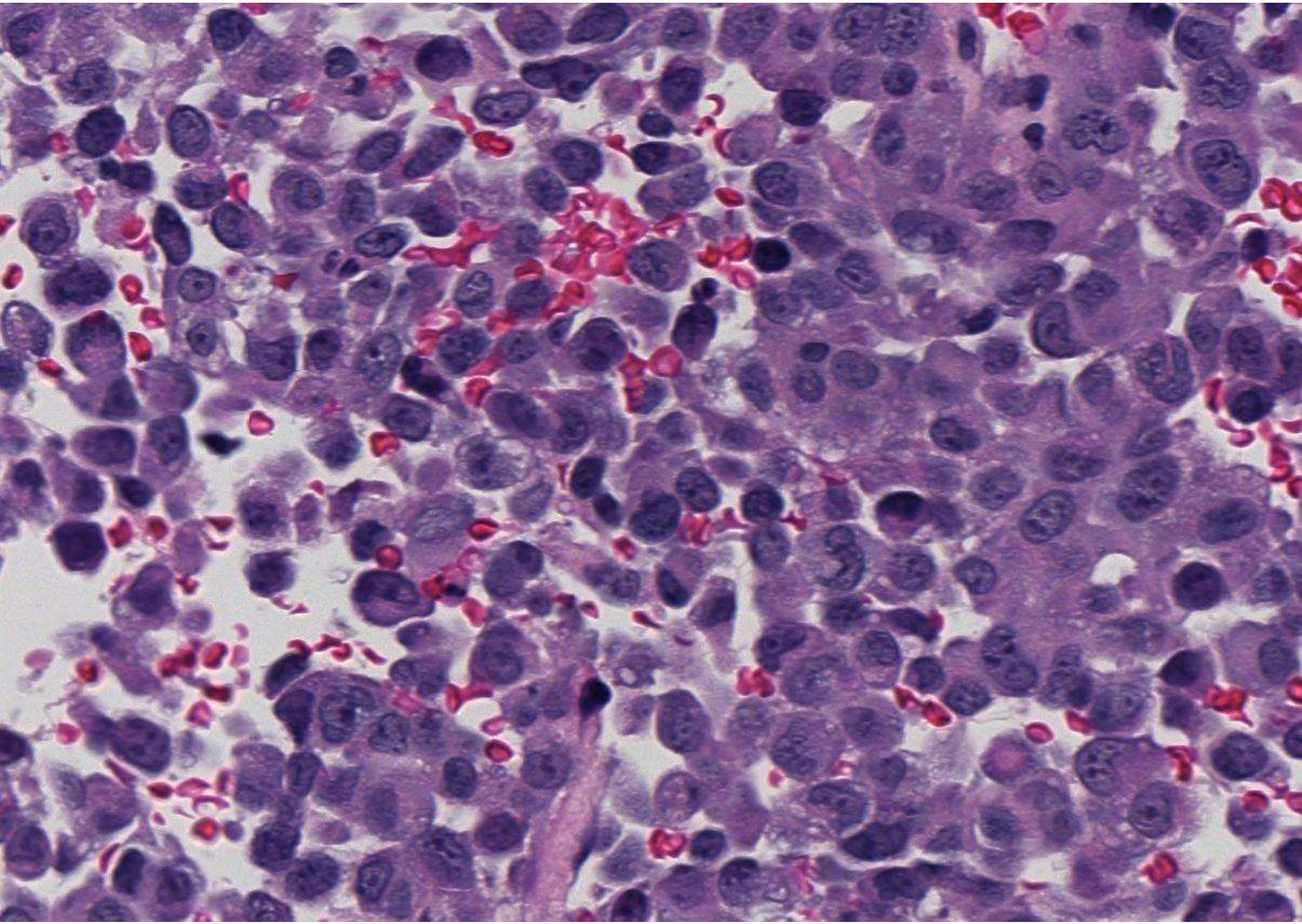






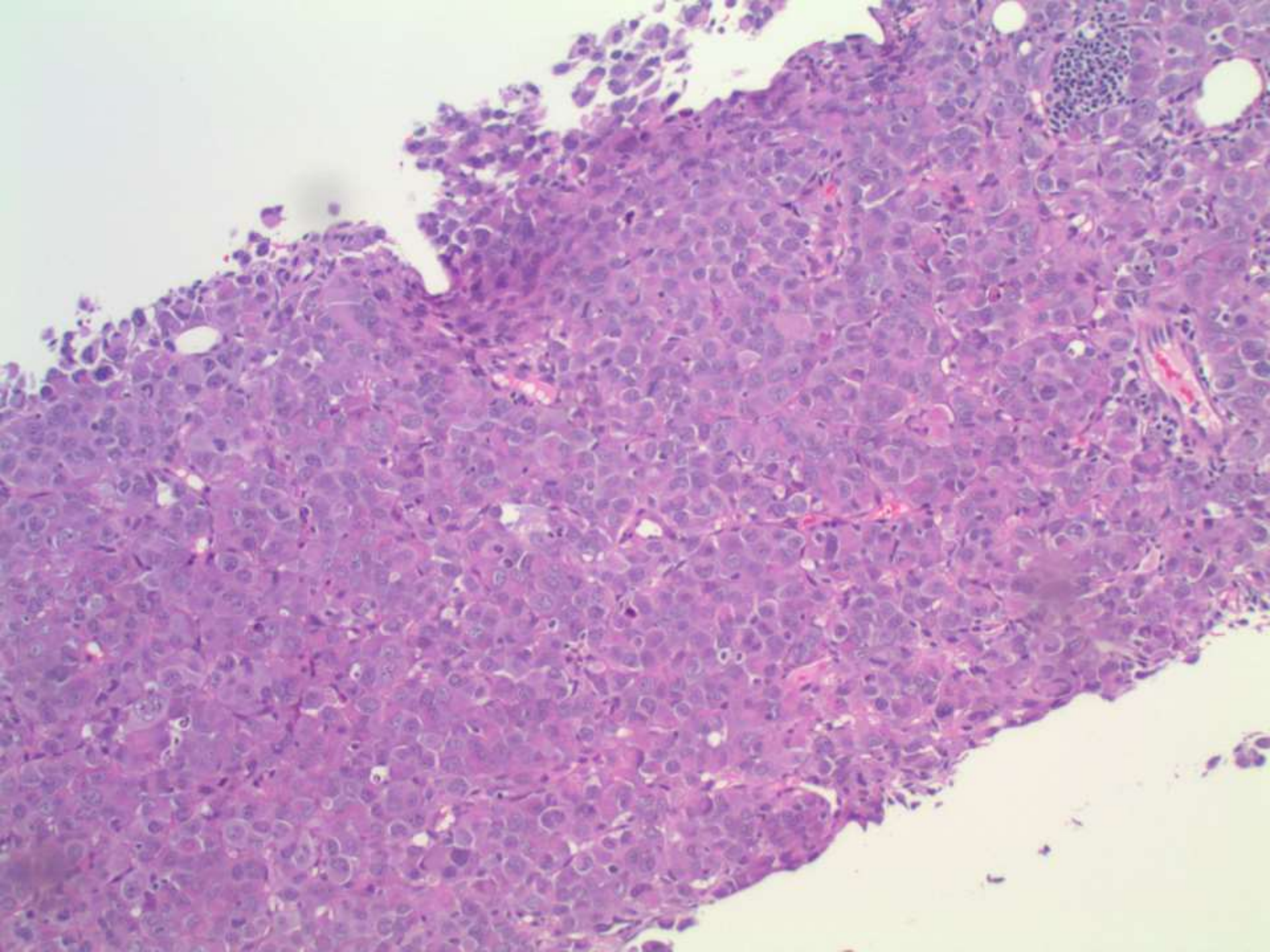


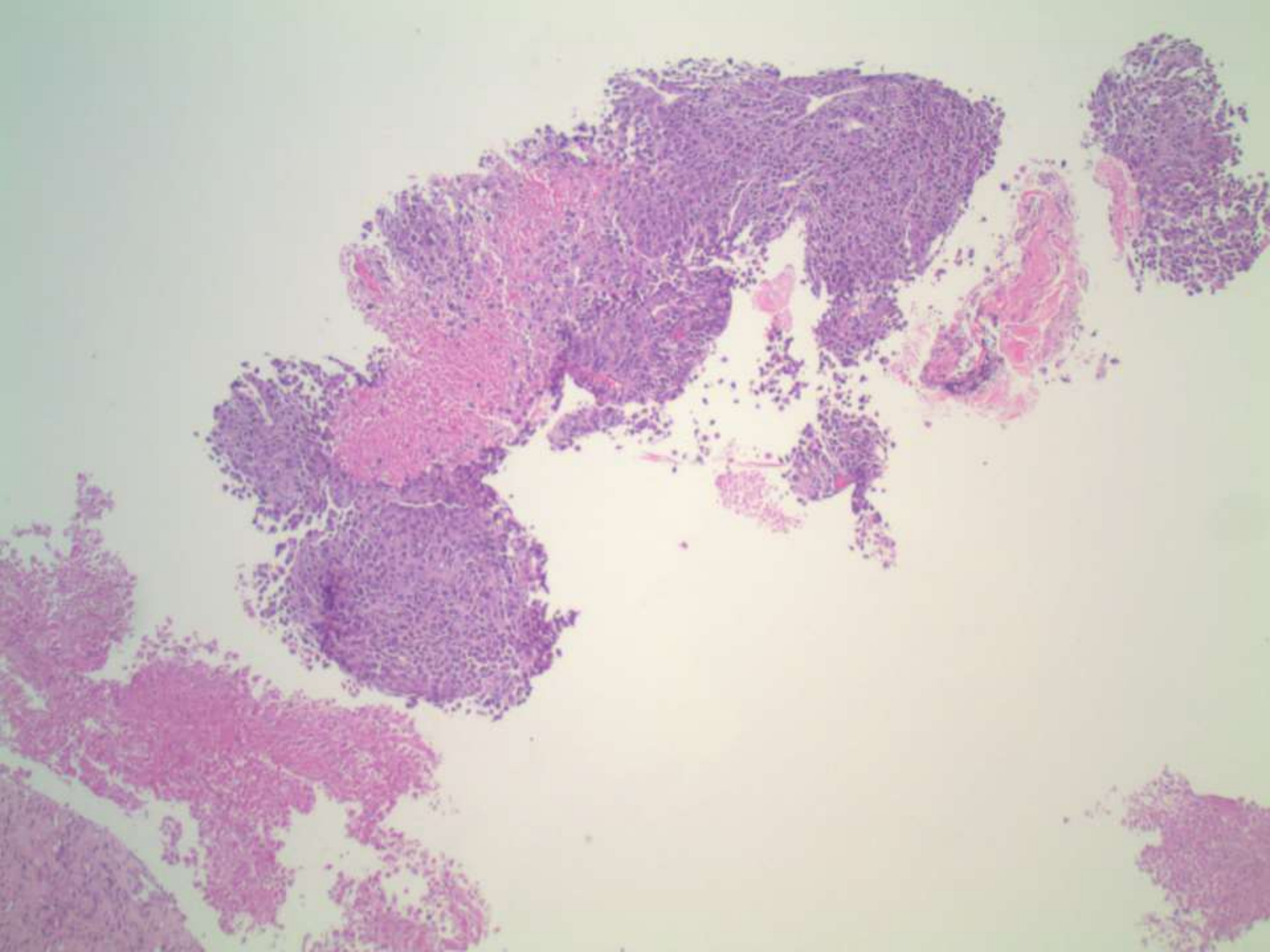


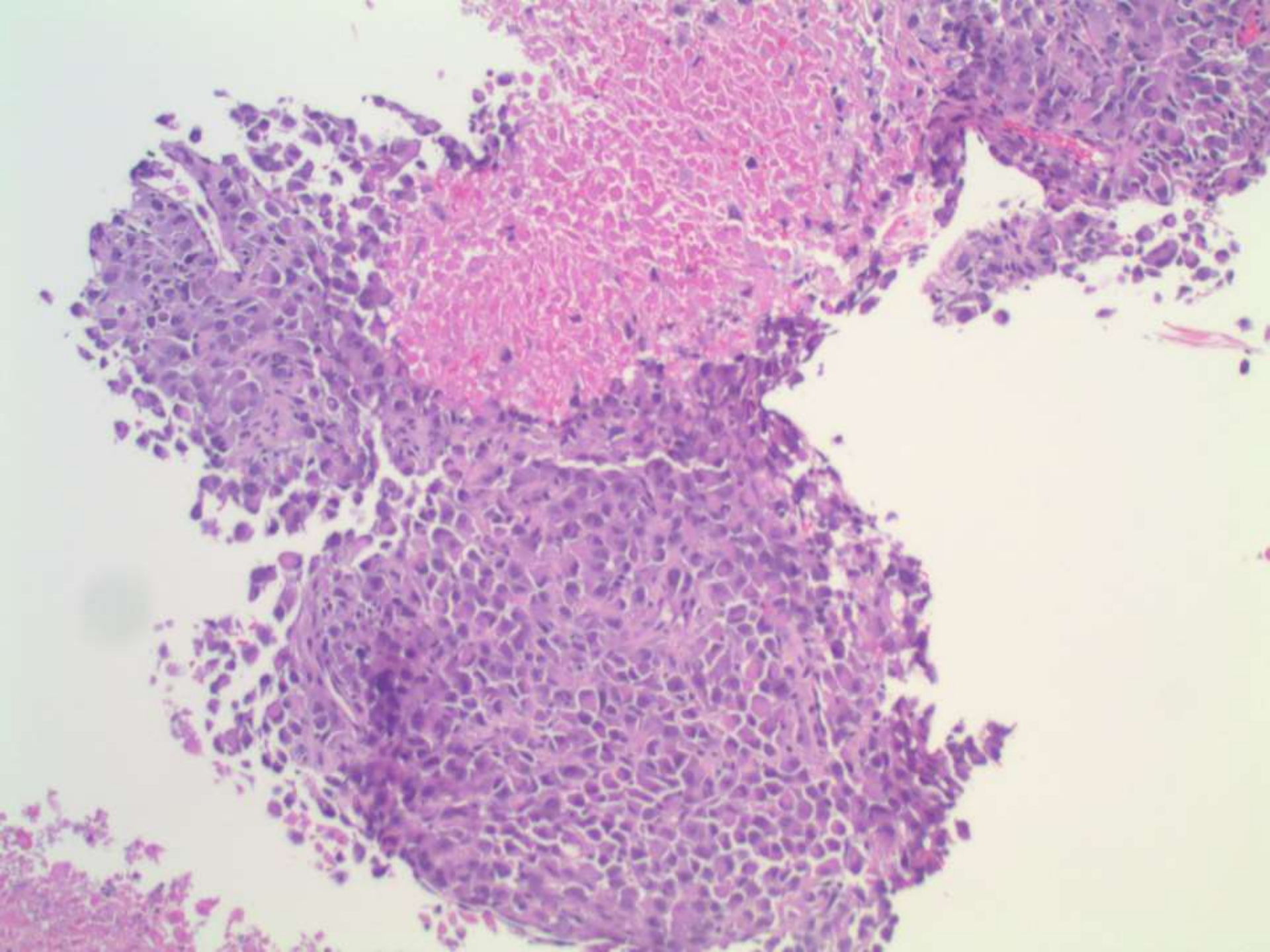


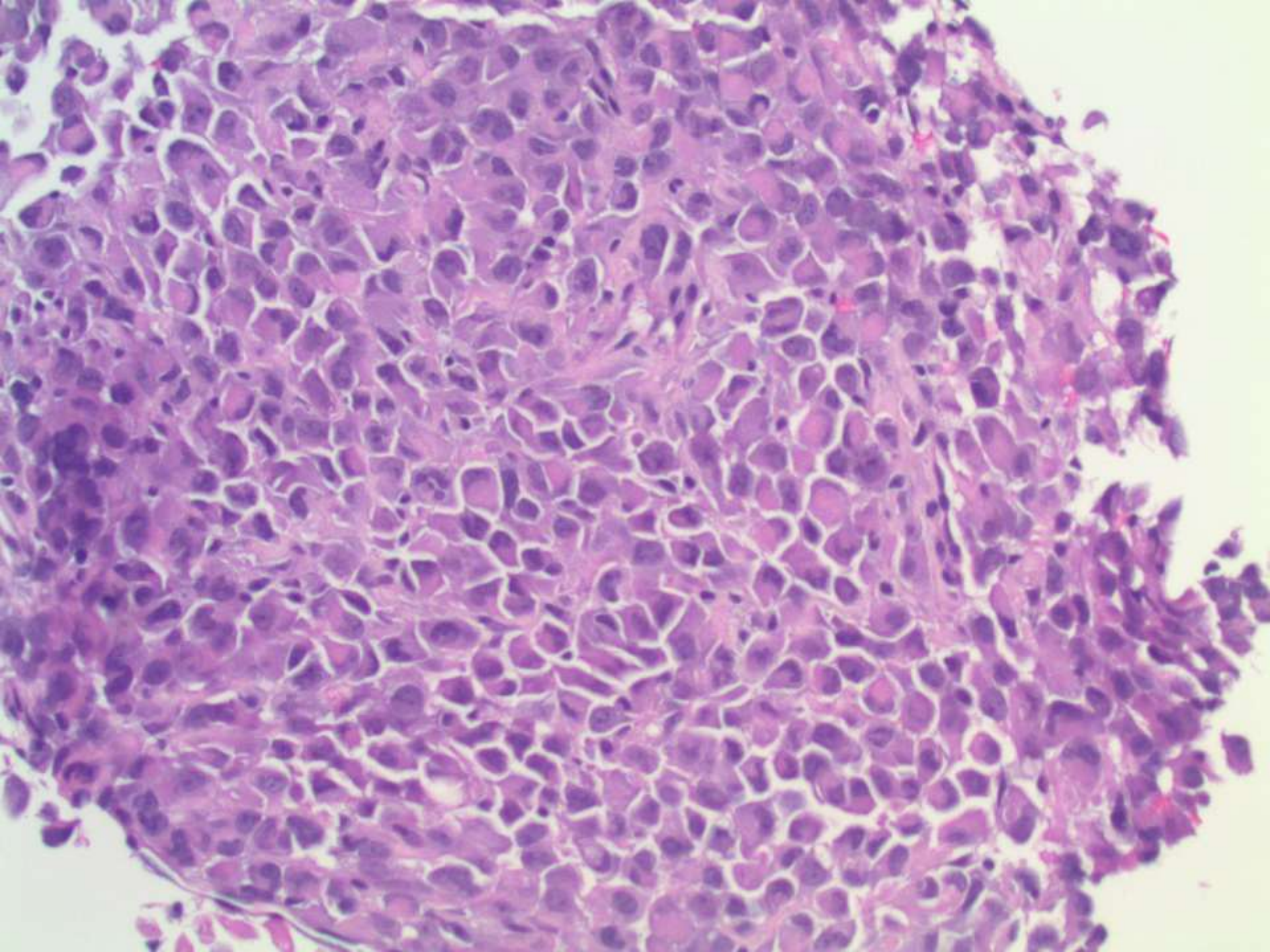
DIAGNOSIS?

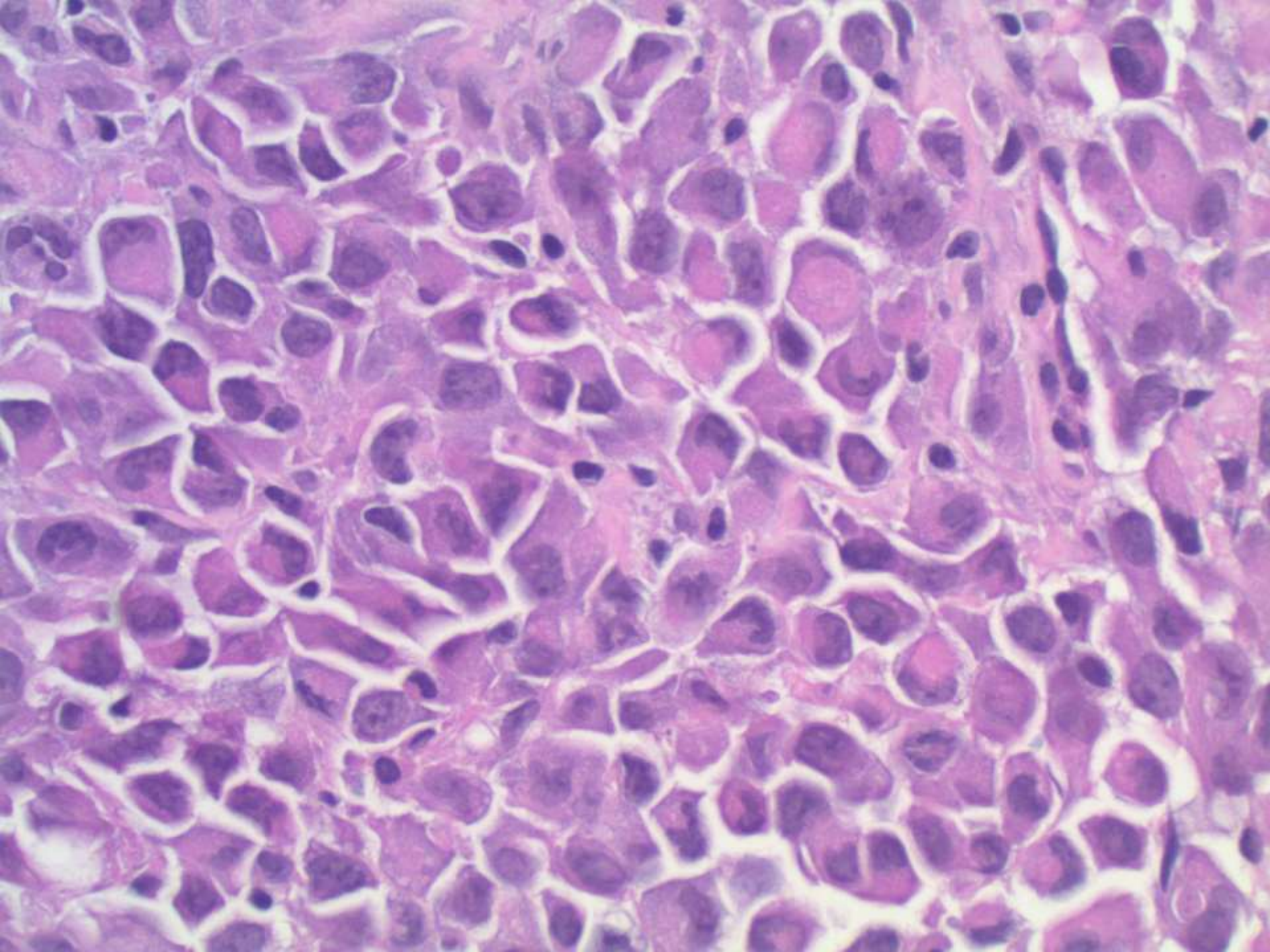
About that prior groin mass bx…

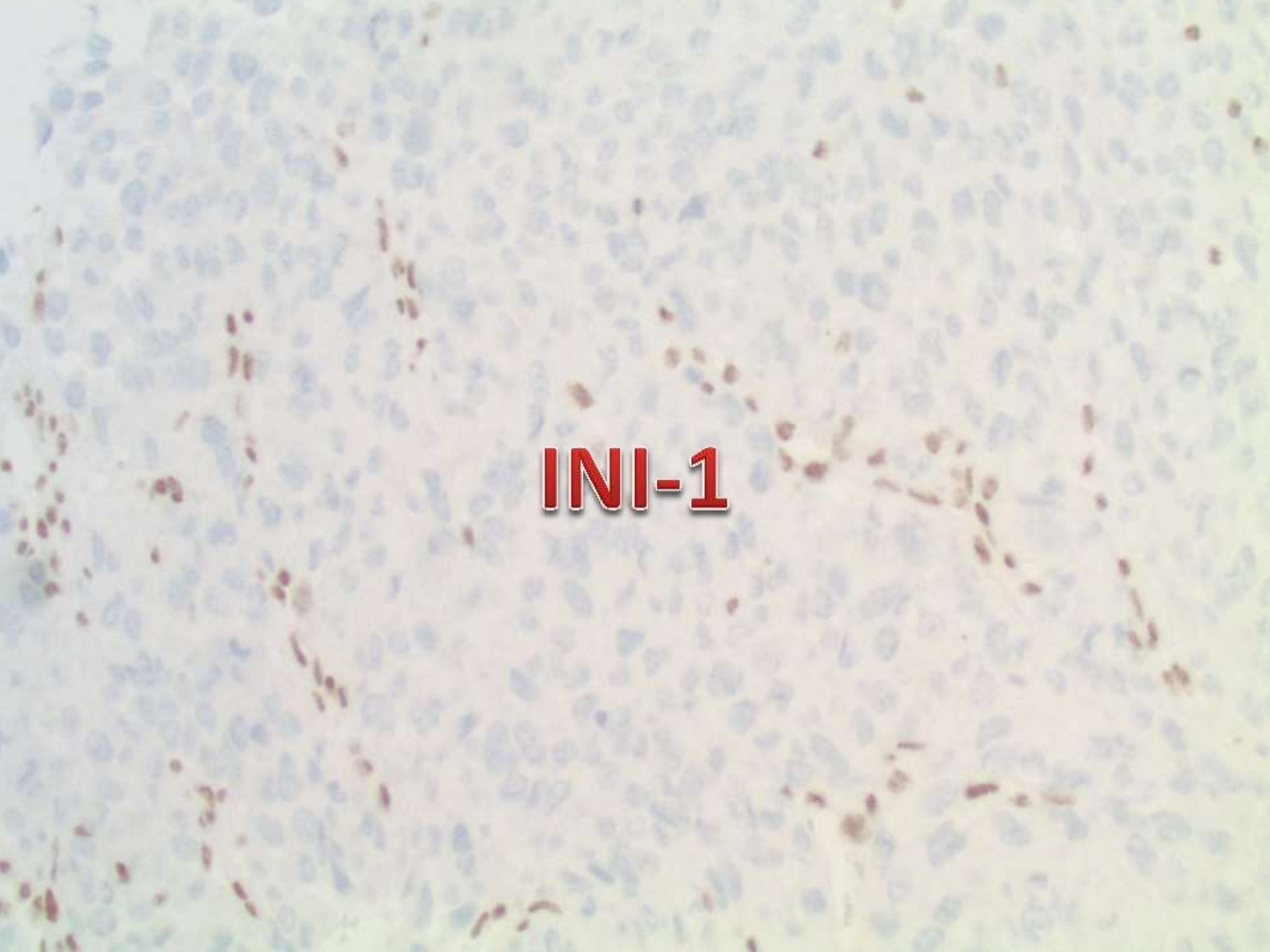











INI-1

FINAL DIAGNOSIS

- **Right groin, core biopsy:**
  - Proximal-type epithelioid sarcoma
- **Pleural mass, excision:**
  - Metastatic proximal-type epithelioid sarcoma

TAKE HOME POINTS

- **When encountering funny-looking tumor with "rhabdoid" features**
  - Consider staining for INI-1 (SMARCB1)
    - "positive" result is ABSENT staining
    - Growing list of INI-1 deficient neoplasms
  - Also consider staining for BRG1 (SMARCA4)
    - "positive" result is ABSENT staining
    - smaller list of BRG1 deficient neoplasms

PROXIMAL TYPE EPITHELIOID SARCOMA

- Keratin, EMA usually positive
- CD34 (50%)
- INI-1 usually lost
- Usually negative for specific vascular markers
- Usually negative for p63/p40

The Expanding Family of SMARCB1(INI1)-deficient Neoplasia: Implications of Phenotypic, Biological, and Molecular Heterogeneity

Abbas Agaimy, MD

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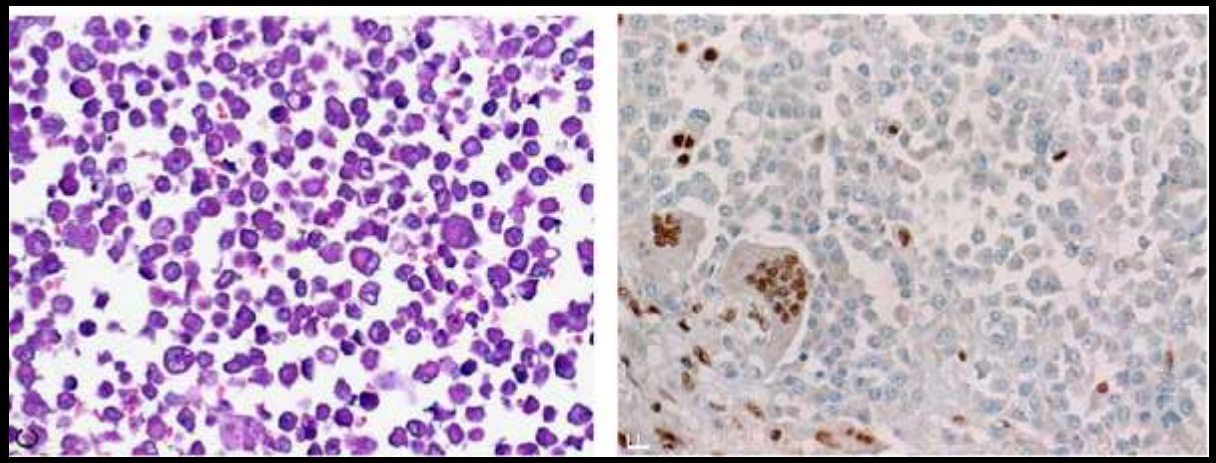

TABLE 1. Reported SMARCB1-deficient Neoplasms

Neoplasm	Typical/Frequent Site/s	Age Group	Rhabdoid Features	% Cases With SMARCB1 Loss	Other Characterized Genetic Alterations Other Than SMARCB1 Loss
AT/RT	CNS, mainly posterior cranial fossa	Children <5 y	Prominent	98%	None or rare
Malignant rhabdoid tumors	Kidney, soft tissue, neck, rarely any organ	Children <5 y	Prominent	100%	None or rare
SMARCB1-deficient sellar tumors	Sellar and perisellar region, skull base	Mainly young adults (20-40 y)	Variable	100%	No data
Cribriform neuroepithelial tumor	Within 3rd/4th ventricle, periventricular	Children	No	100%	No data
Renal medullary carcinoma	Kidney, mainly right	Children + young adults	Subtle or none	100%	None or rare
Collecting duct carcinoma	Kidney	Adults and elderly	None or subtle	15%	No consistent aberrations
Epithelioid sarcoma, classical (distal) type	Distal extremities	Young adults	Less prominent	90%-100%	No consistent aberrations
Epithelioid sarcoma, proximal type	Proximal extremities and axial	Young adults	Prominent	100%	No consistent aberrations
Epithelioid MPNST	Soft tissue extremities	Adults, children	Not prominent	50%	No consistent data
Synovial sarcoma	Deep soft tissue, extremities	Young adults, wide age range	Usually subtle (9%)	6%-100%	SYT translocation in >90%
Myoepithelial carcinoma	Soft tissue	Wide age children + adults	Variable, not prominent	10%-40%	Up to 50% EWSR1-fusion+
Extraskeletal myxoid chondrosarcoma	Deep soft tissue, extremities	Variable adults	Not consistent	16%	4 SMARCB1-deficient cases lacked EWSR1 fusion
Typical/poorly differentiated chordoma	Sacral, skull base, others	Variable, adults	Not prominent	1.4%/100%	Variable
Osteosarcomas	Bones	Mainly children and young adult but any age	No	0.7%	Variable genetics
Primary bone tumors all	Bones	Variable	No	0.6%	Variable genetics
Ossifying fibromyxoid tumors	Soft tissue extremities, head & neck & trunk	39-63 (median 52)	No	74%, mosaic pattern (hemizygous deletions)	Not clear
GIST	Small intestine, stomach	50-70 y	Rare	up to 70%, mosaic pattern	*KIT* or *PDGFRA* mutations
Gastrointestinal carcinoma	All sites, mainly right colon, jejunum, stomach	Mean, 65 y	Prominent in most cases	75% of tested cases	75% CIMP phenotype (MSI)
Undifferentiated pancreatic rhabdoid carcinoma	Head ± body and tail	44-96 y (mean, 65 y)	Prominent	28% of cases	*KRAS* alterations in 25%
Sinonasal carcinoma	Nasal cavity and sinuses	28-52 y	Subtle	2.7% of all sinonasal carcinomas	No genetic data