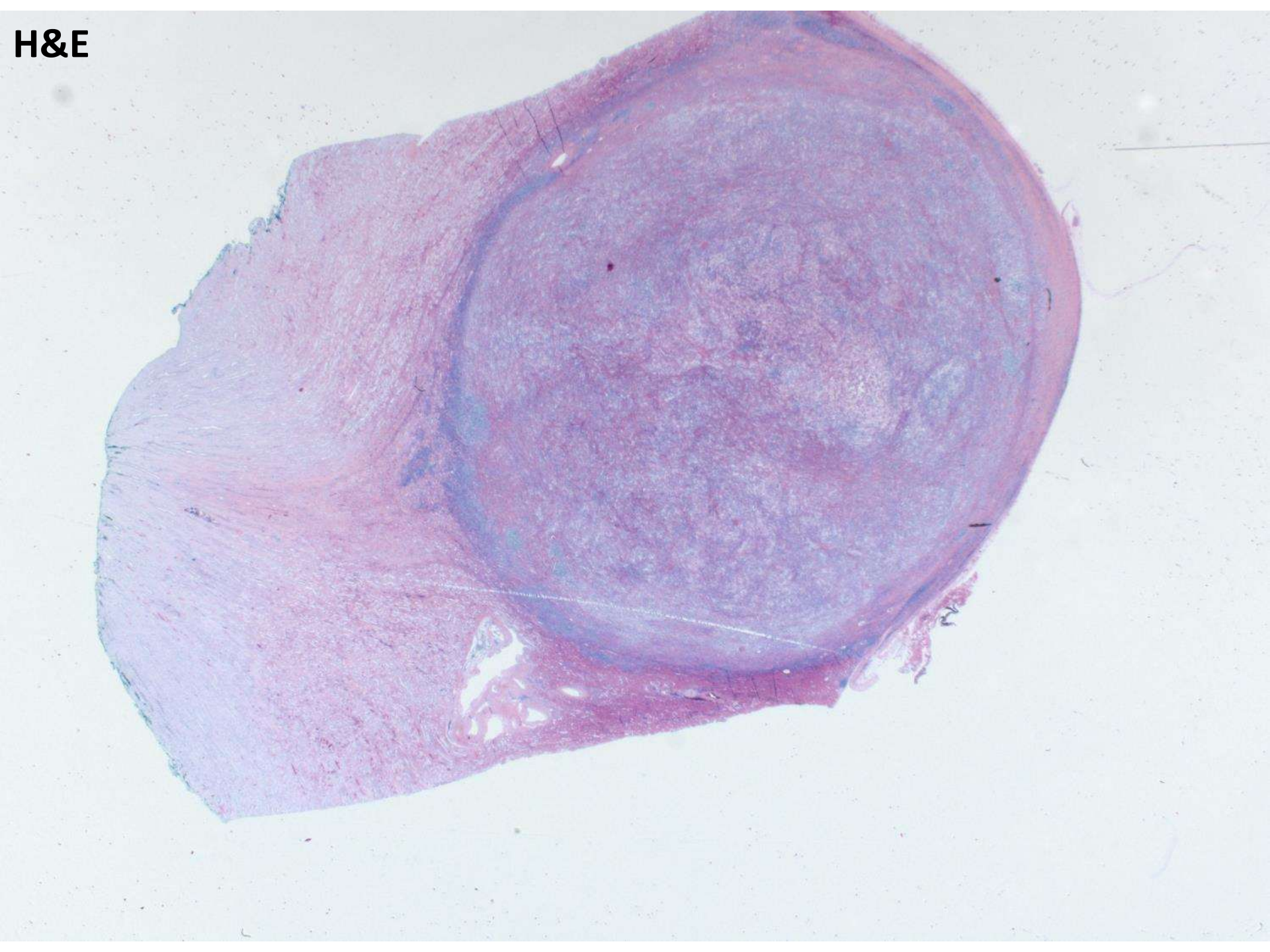
MAY 2020 DIAGNOSIS LIST

20-0401: Extranodal Rosai-Dorfman disease (kidney; GU pathology)
20-0402: Juxtaglomerular cell tumor (kidney; GU pathology)
20-0403: metastatic neuroendocrine tumor (ovary; GI and GYN pathology)
20-0404: plasmacytoid urothelial carcinoma (ureter; GU pathology)
20-0405: amyloidosis (bladder; GU pathology)
20-0406: mesonephric-like carcinoma (ovary; GYN pathology)
20-0407: clear cell ependymoma (brain; neuropathology)
20-0408: echinococcal cyst (ovary; infectious disease pathology)
20-0409: gliomatosis peritonei (ovary; GYN pathology)
20-0410: prostatic ductal adenocarcinoma (urethra; GU pathology)

20-0401

Julia Rewerska/Sunny Kao; Stanford

55-year-old F presenting with an incidental 3cm right renal mass on imaging.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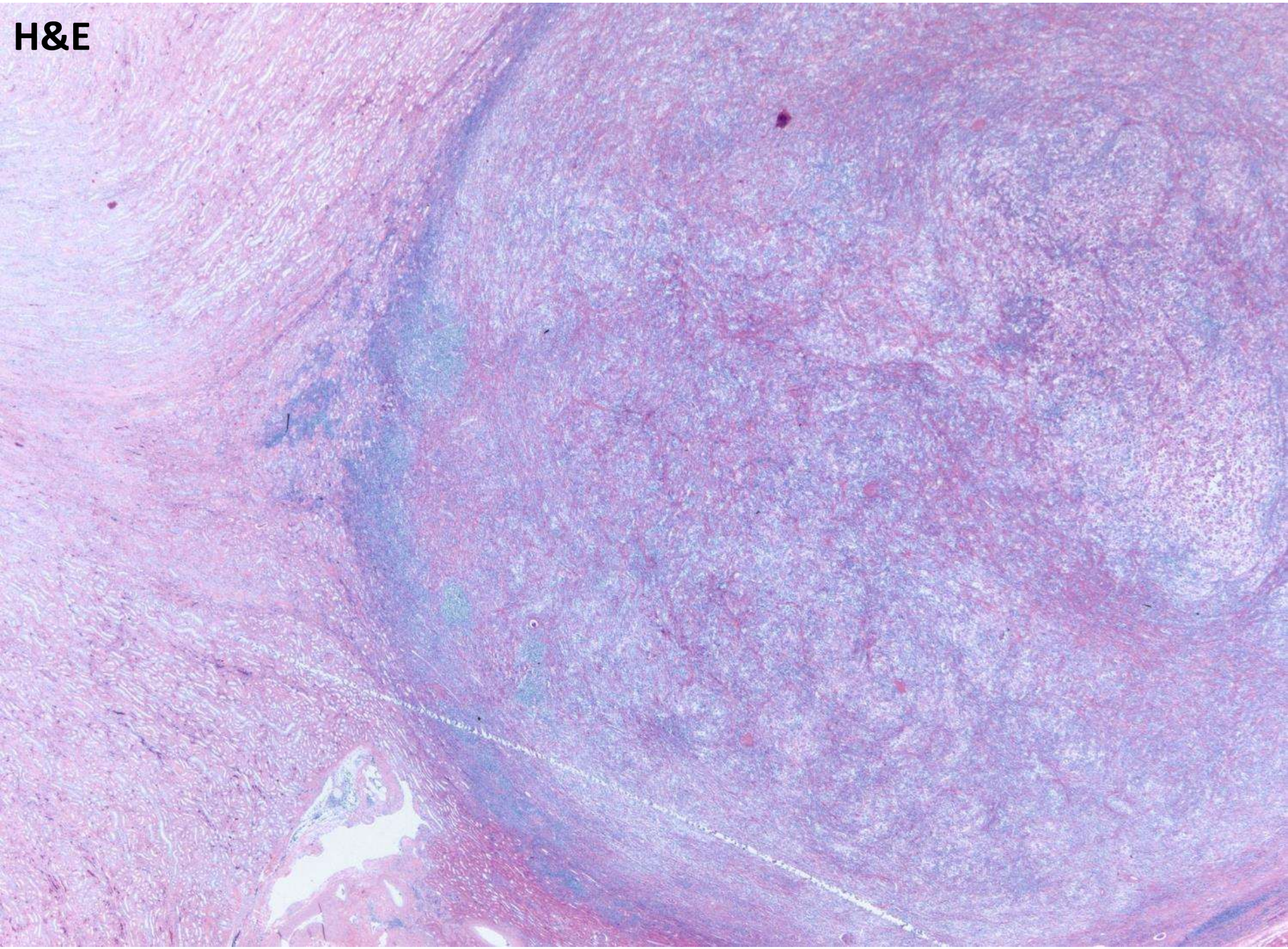
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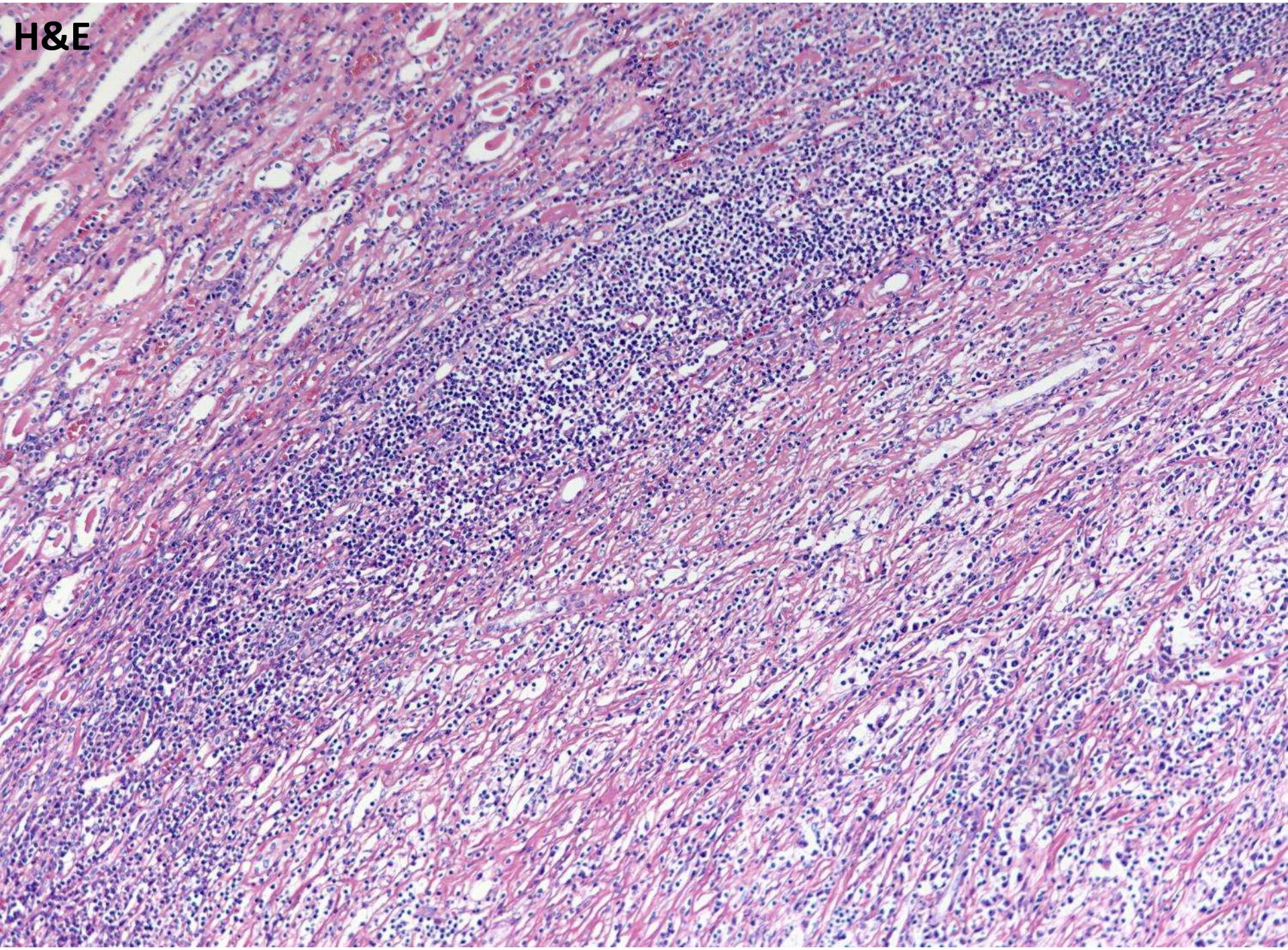
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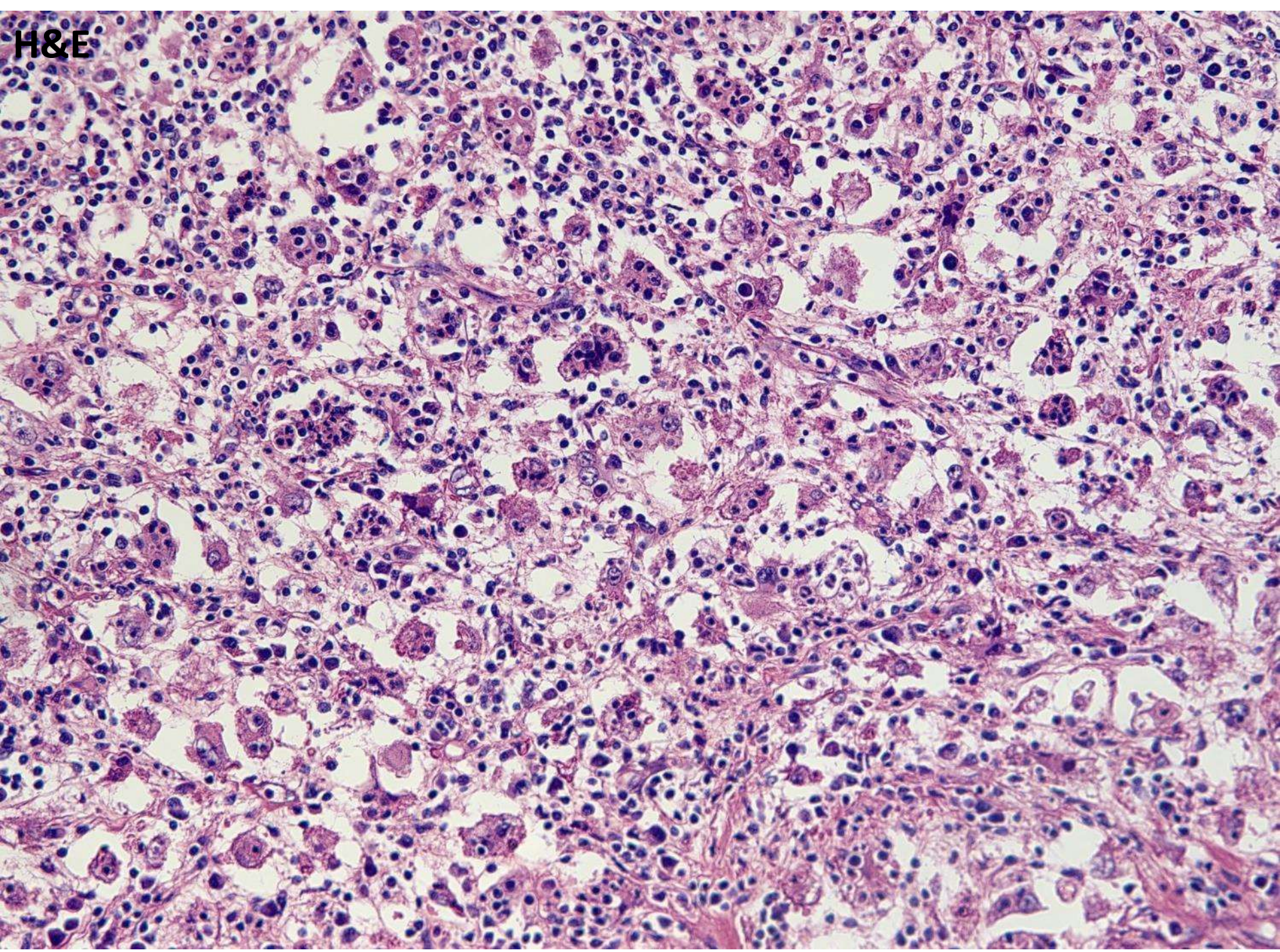
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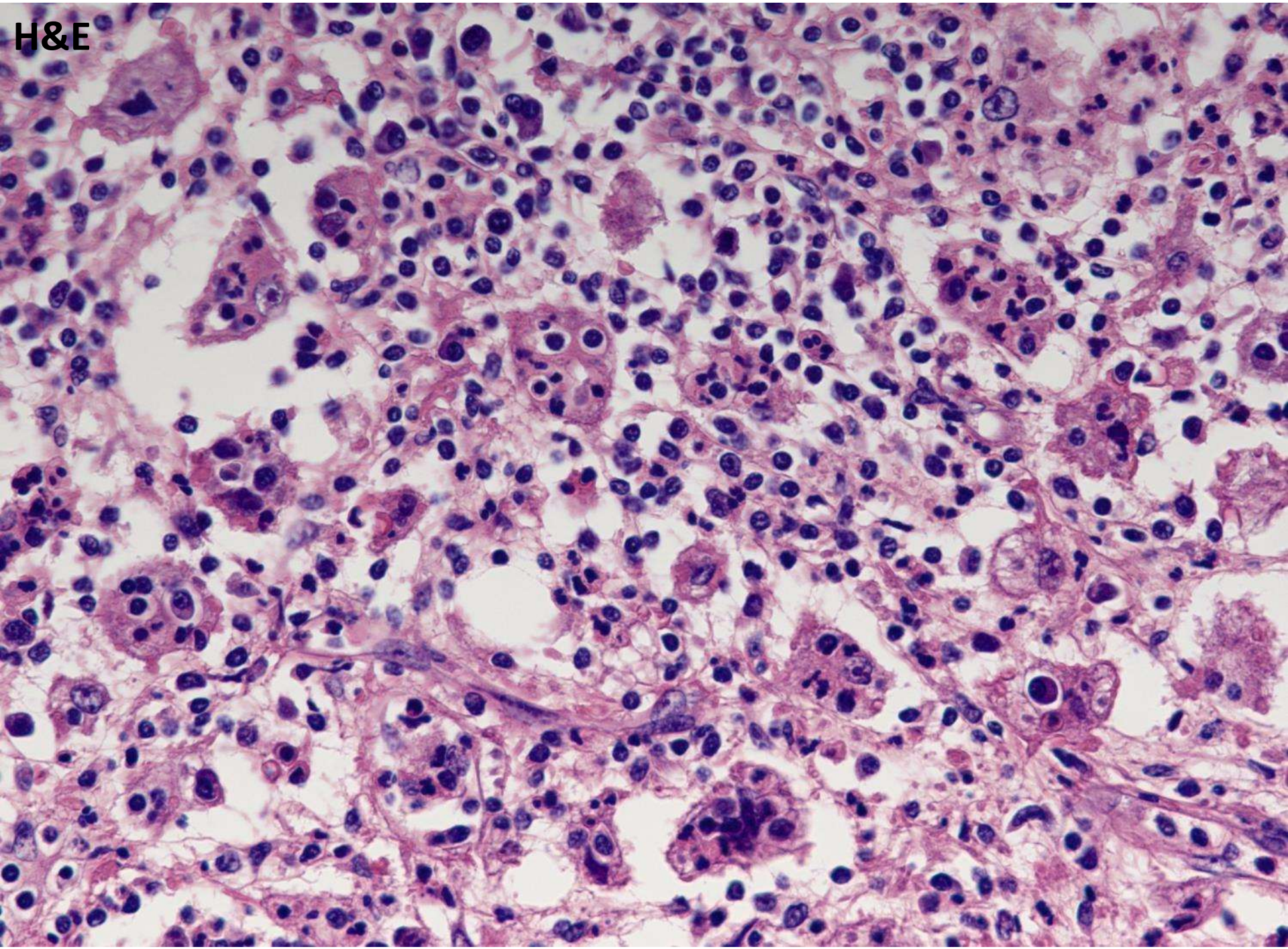
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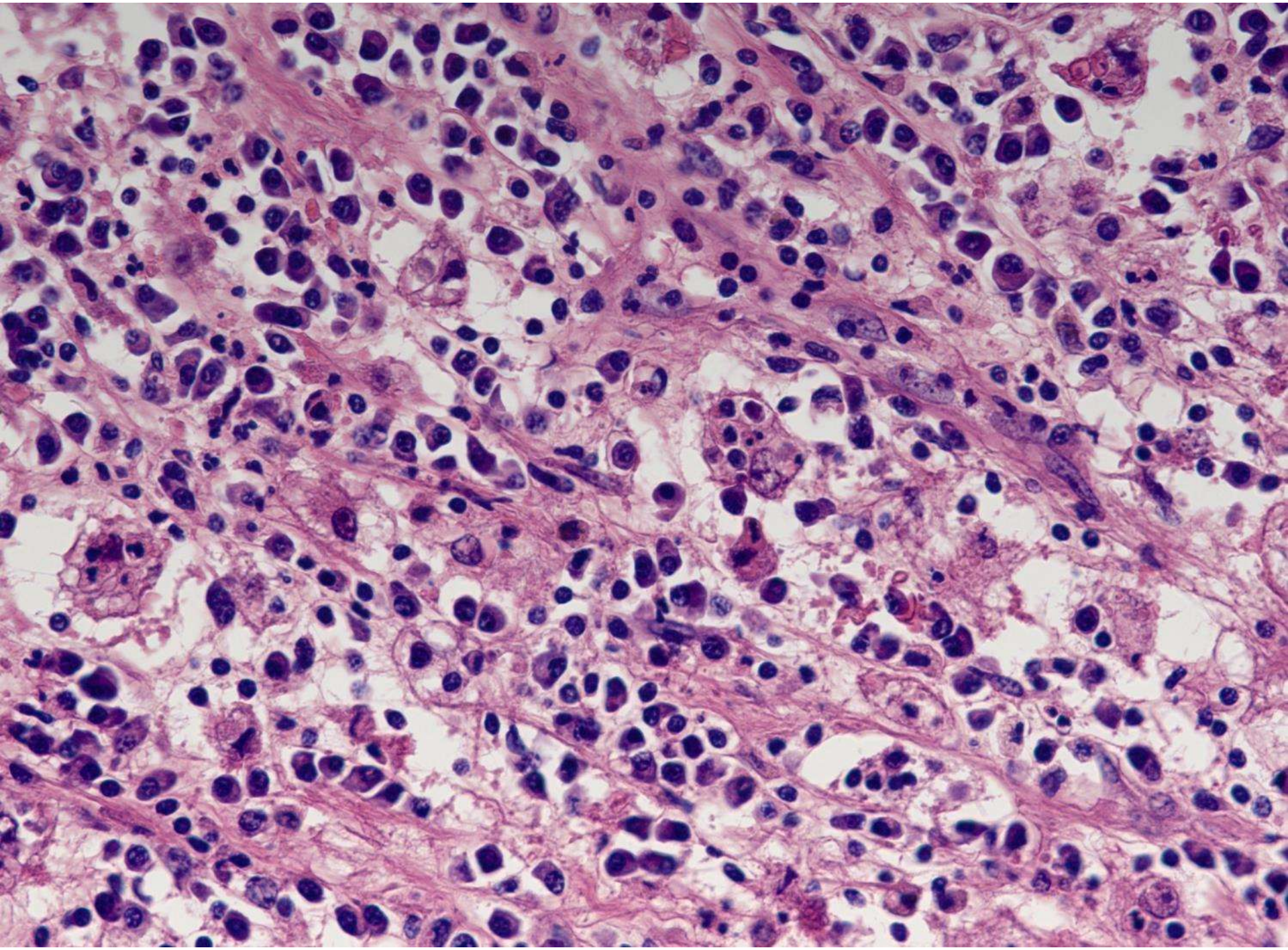

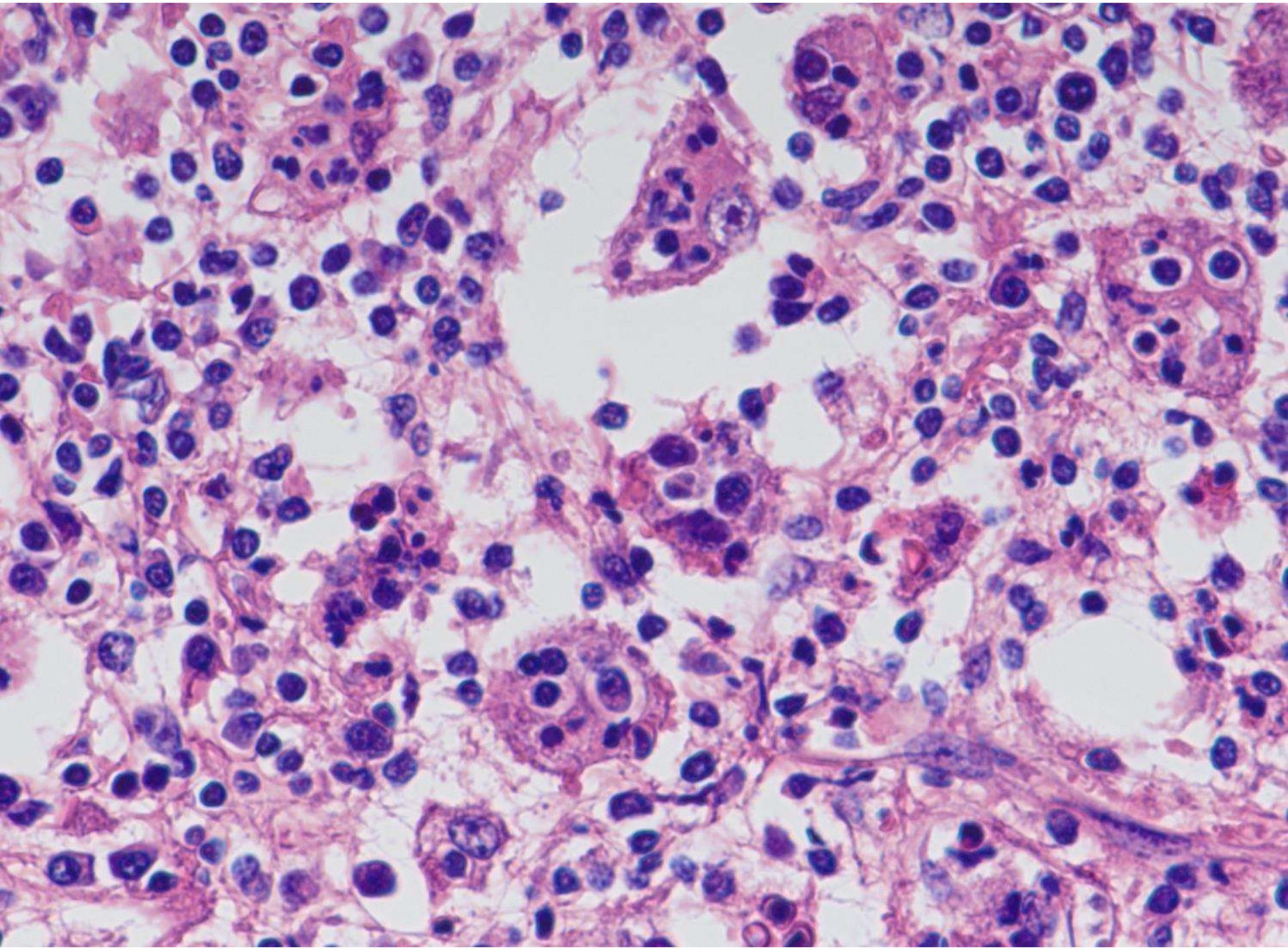

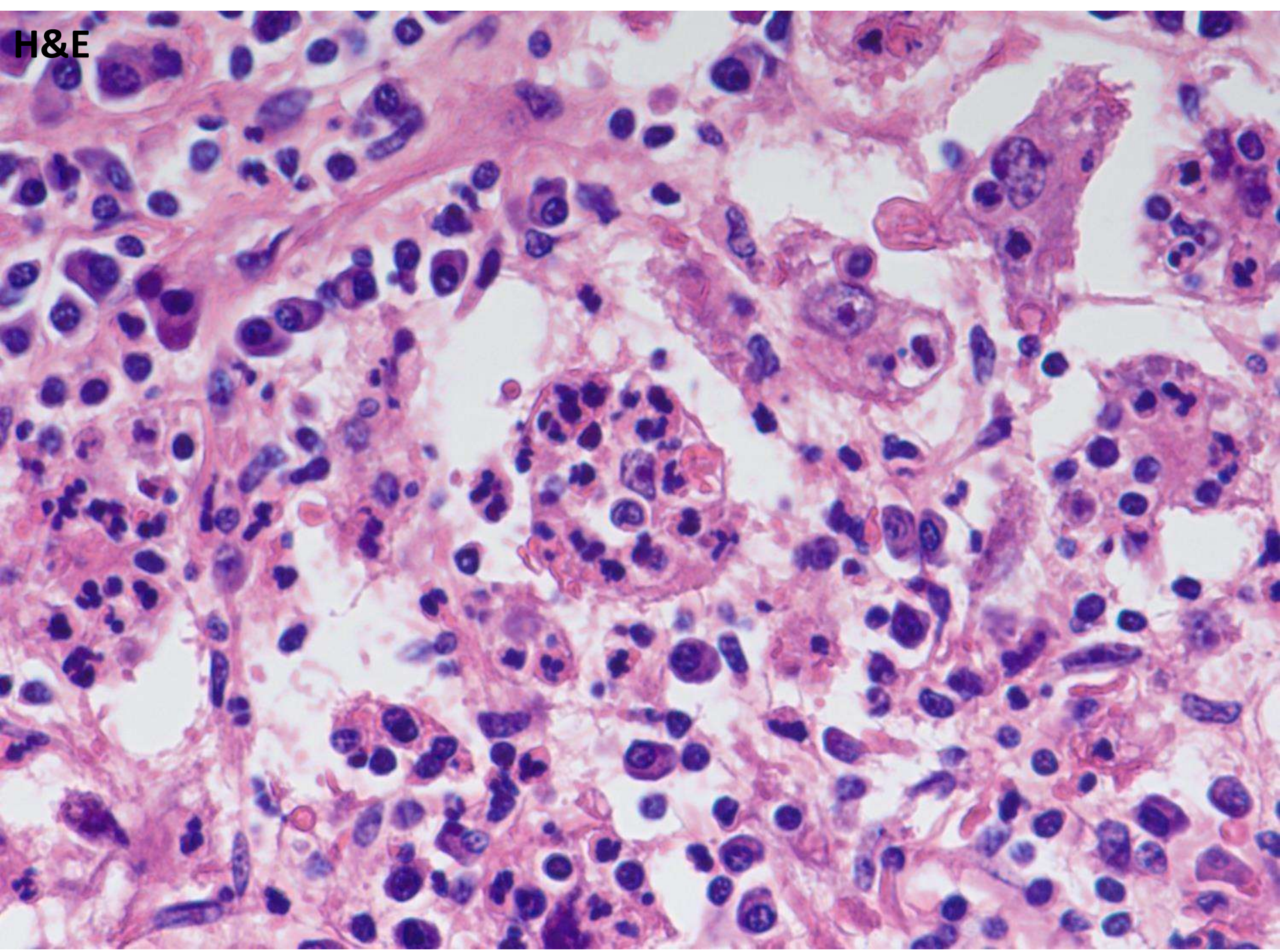
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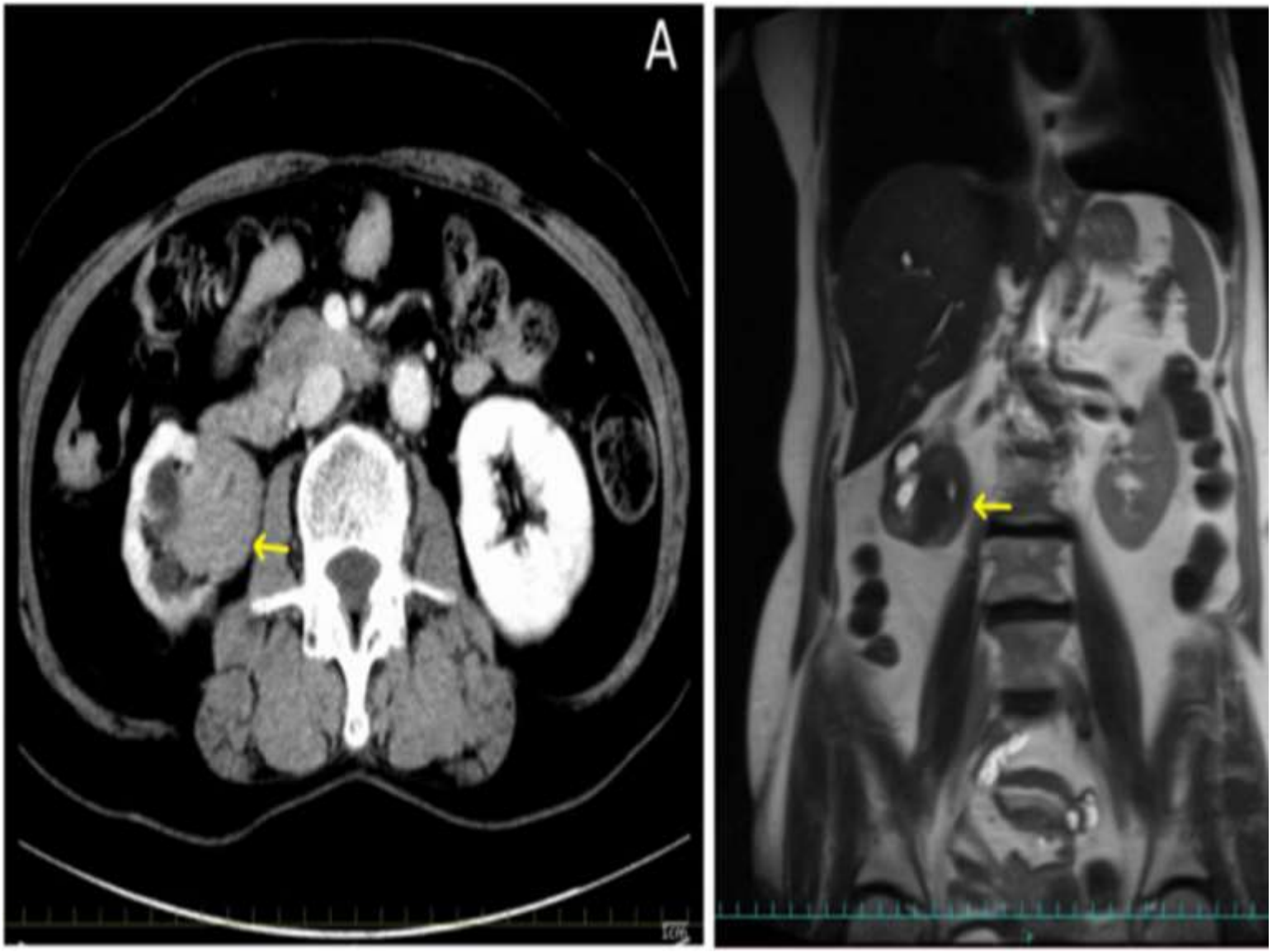
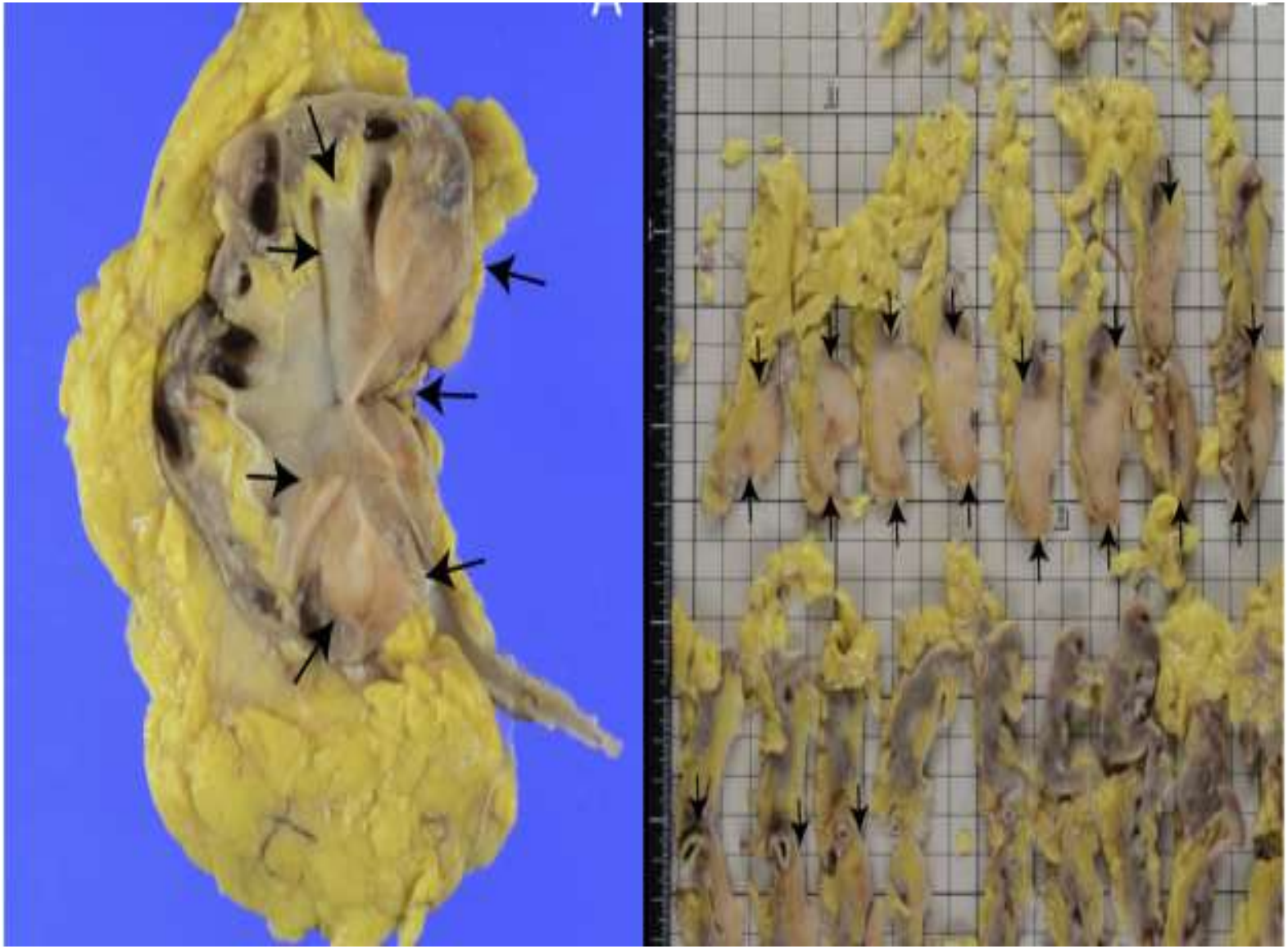


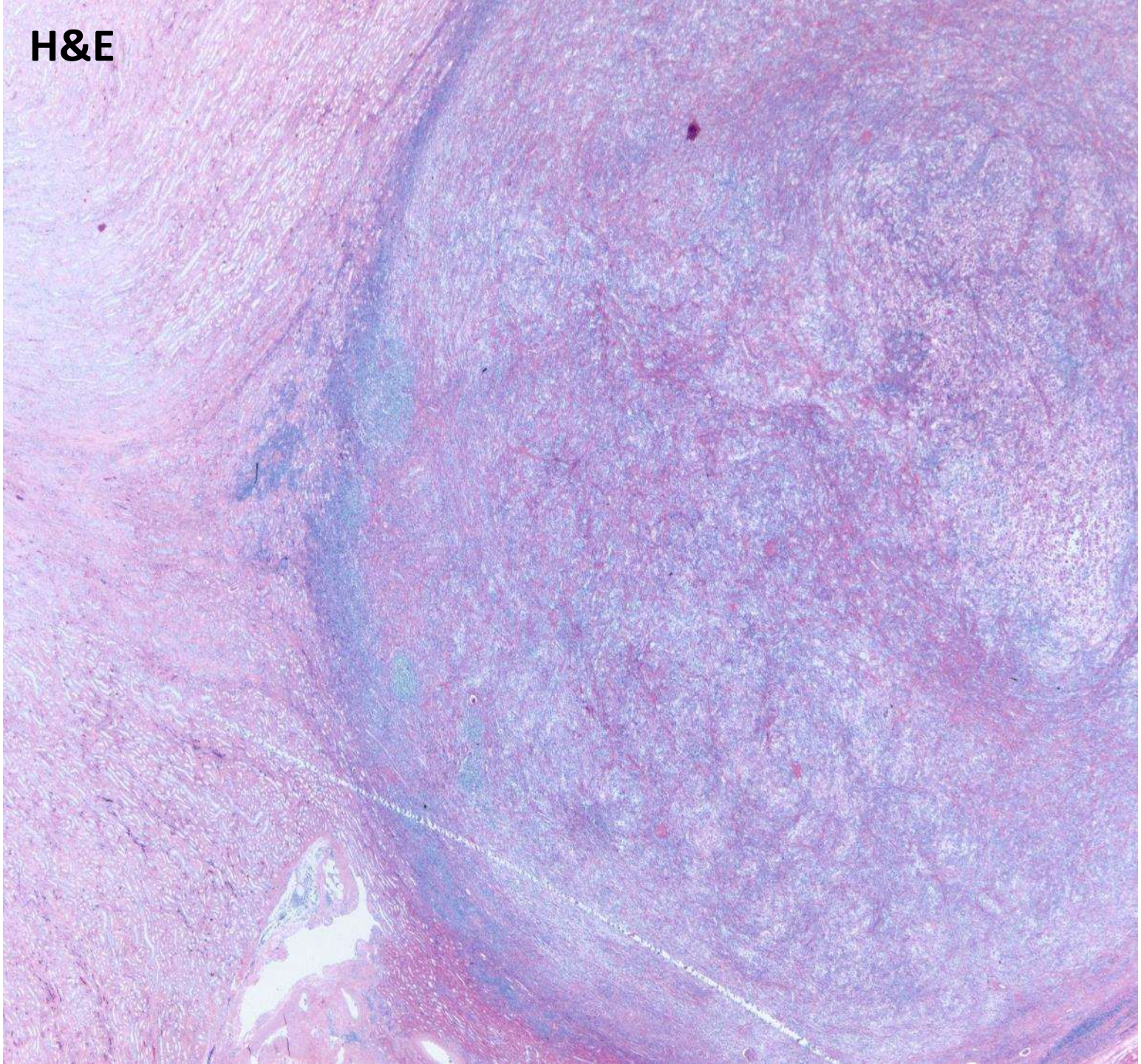
Tanaka H; Mukai S; Kamoto T; Kataoka H. Extranodal Rosai-Dorfman disease of the kidney: A case report. Human pathology: case reports: Volume 17; Article 200306- September 2019



Tanaka H; Mukai S; Kamoto T; Kataoka H. Extranodal Rosai-Dorfman disease of the kidney: A case report. Human pathology: case reports: Volume 17; Article 200306- September 2019

Differential Diagnoses

- Renal cell carcinoma
- Lymphoma
  - Especially when accompanied by lymphadenopathy
- IgG4- related kidney disease
- Extranodal Rosai-Dorfman disease
- Langerhans cell histiocytosis
- Infection
  - Tuberculosis
- Xanthogranulomatous pyelonephritis
- Metastatic tumor
  - Malignant melanoma
- Storage disease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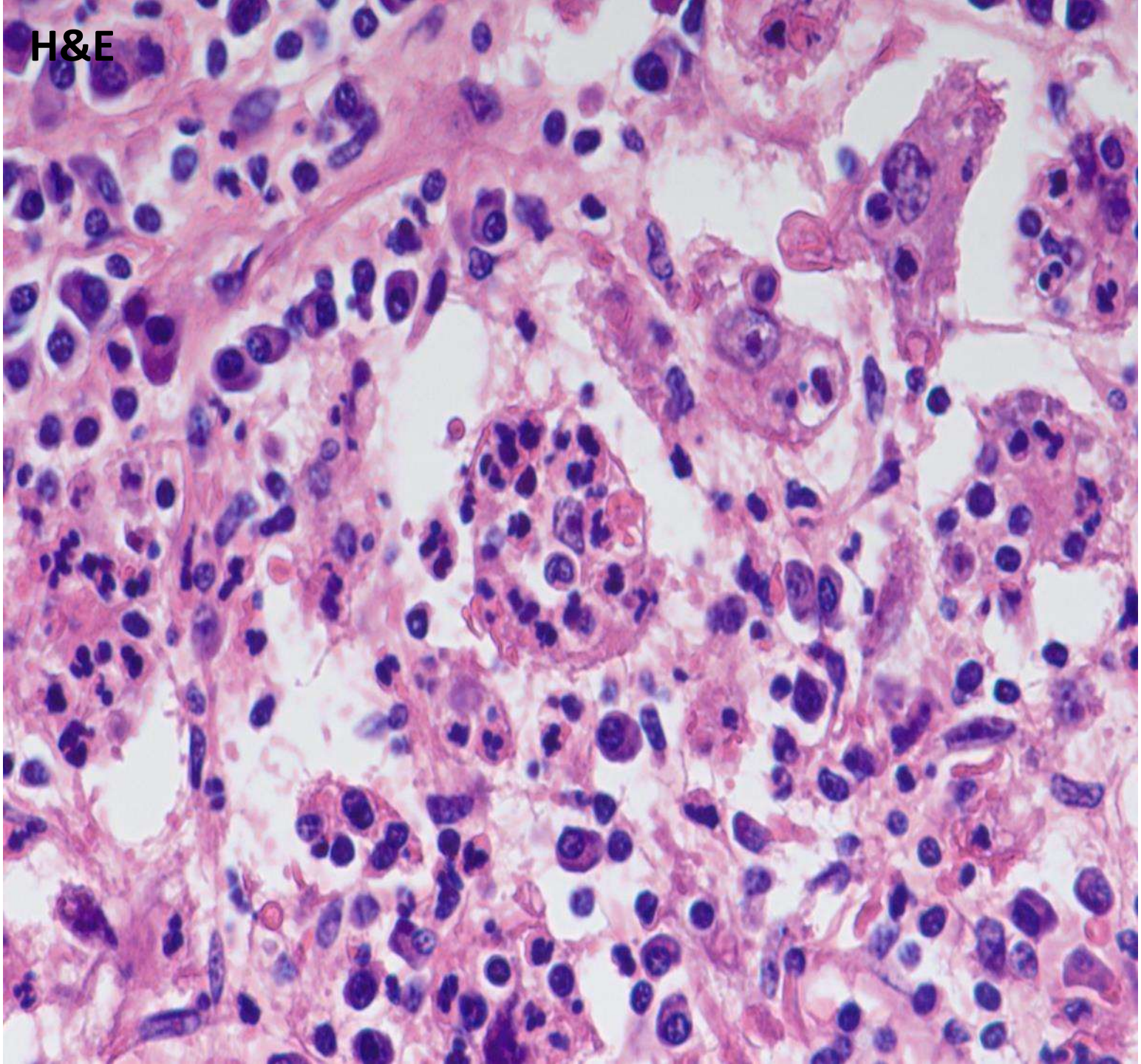
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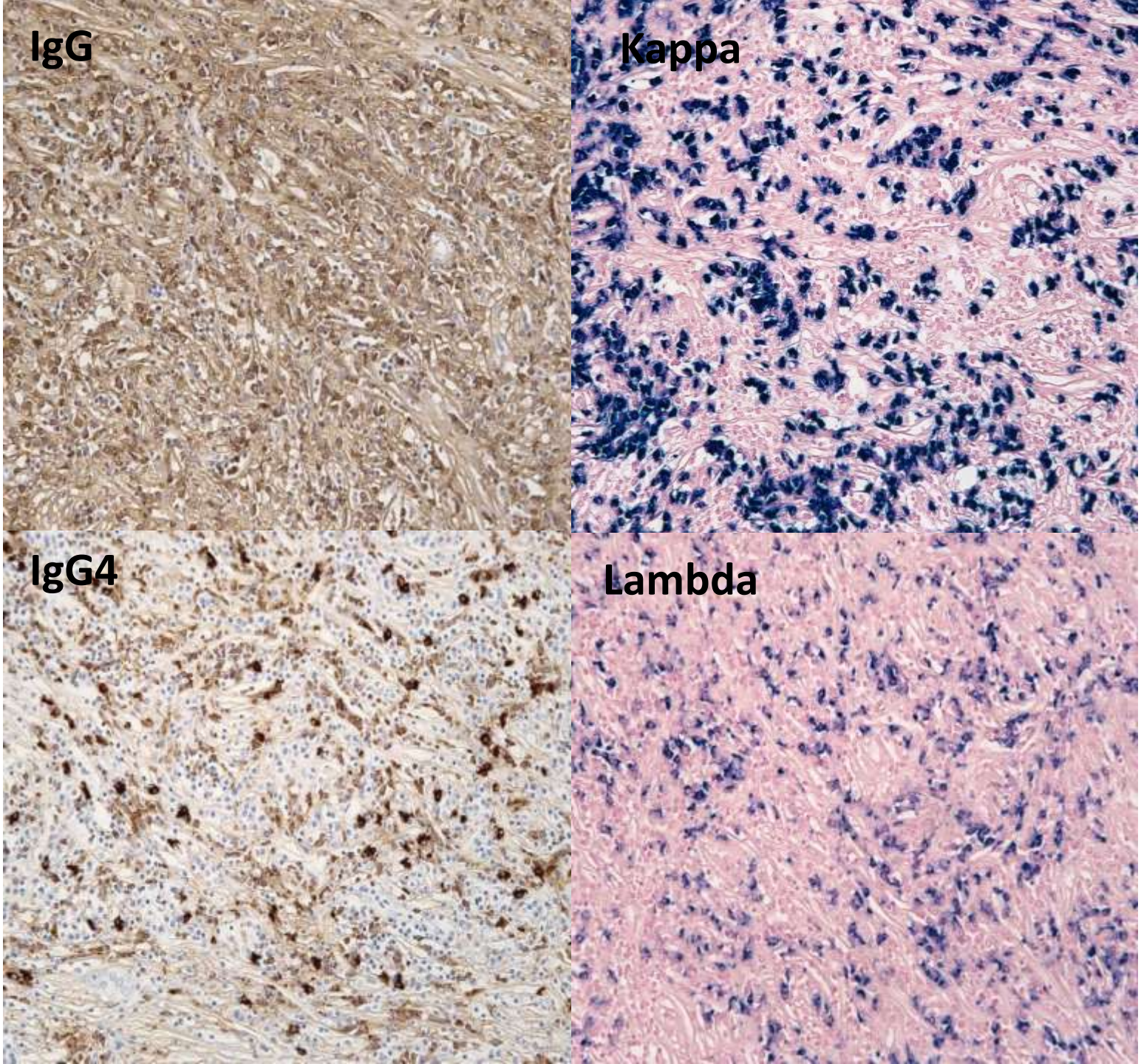
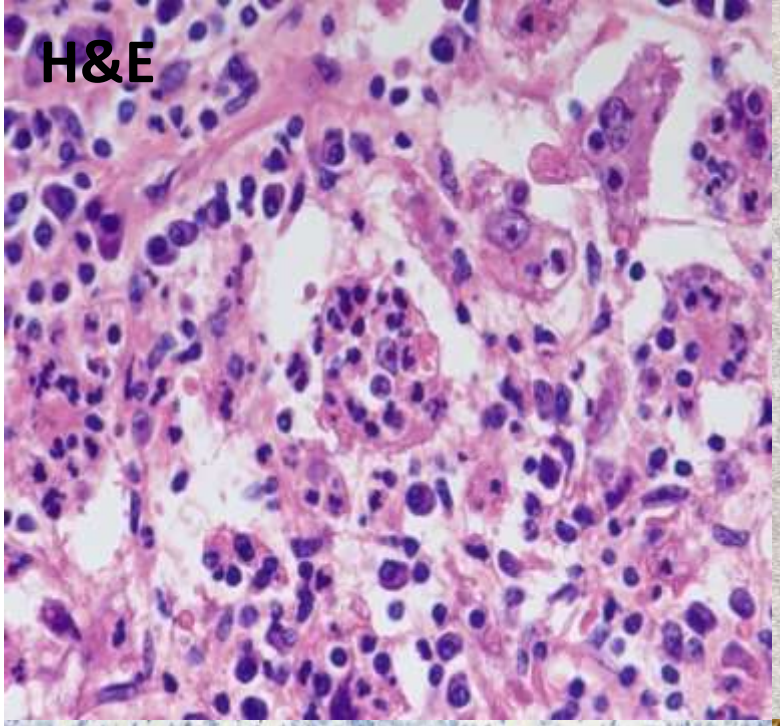

IgG
Kappa
IgG4
Lambda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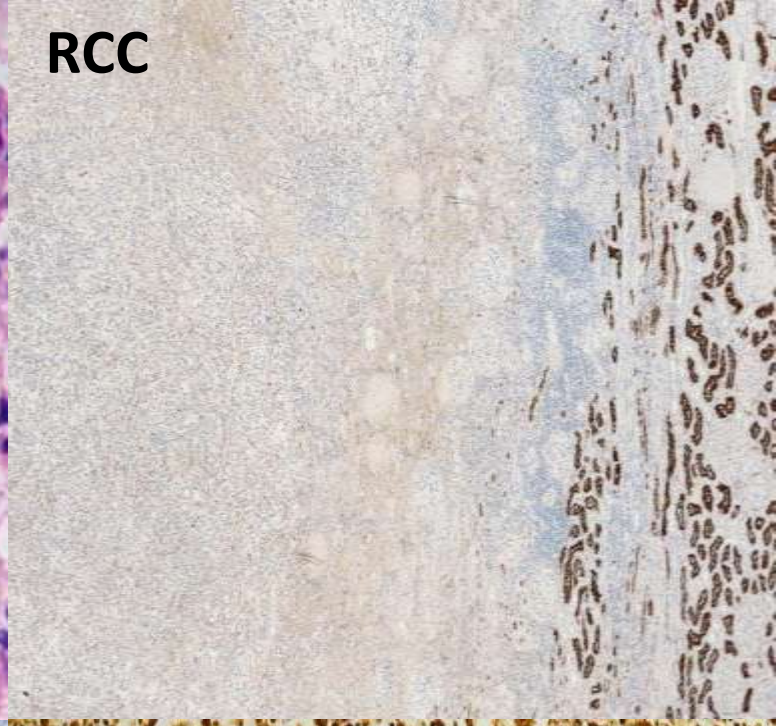
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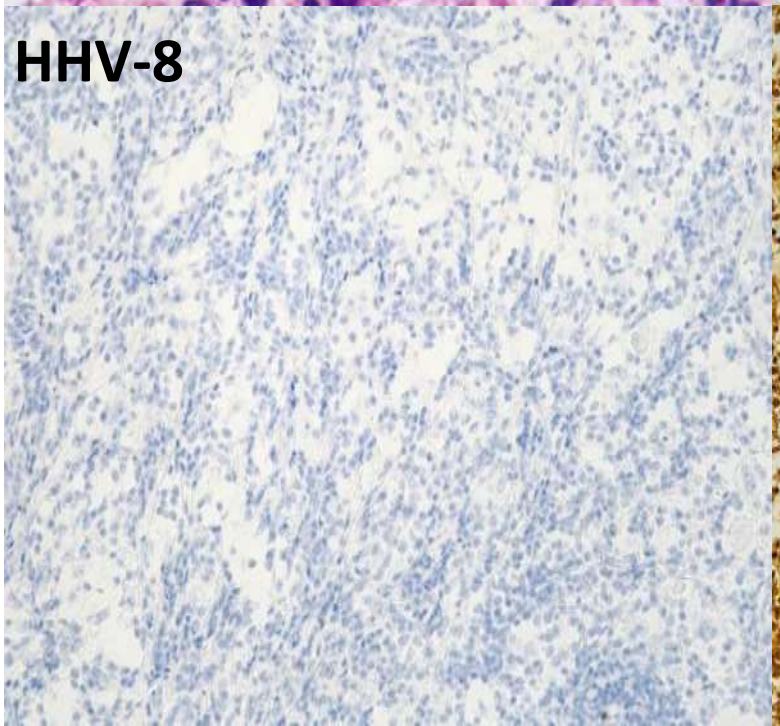
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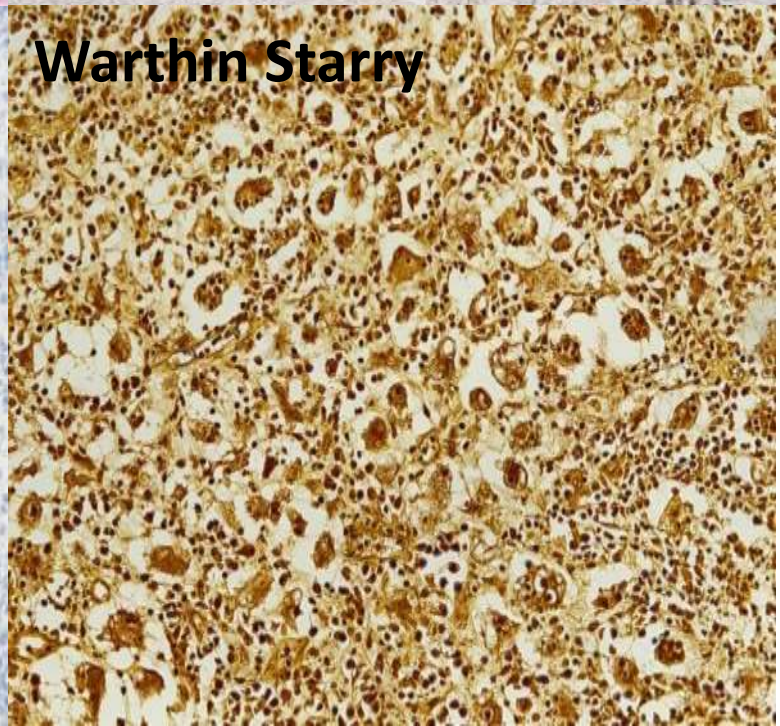
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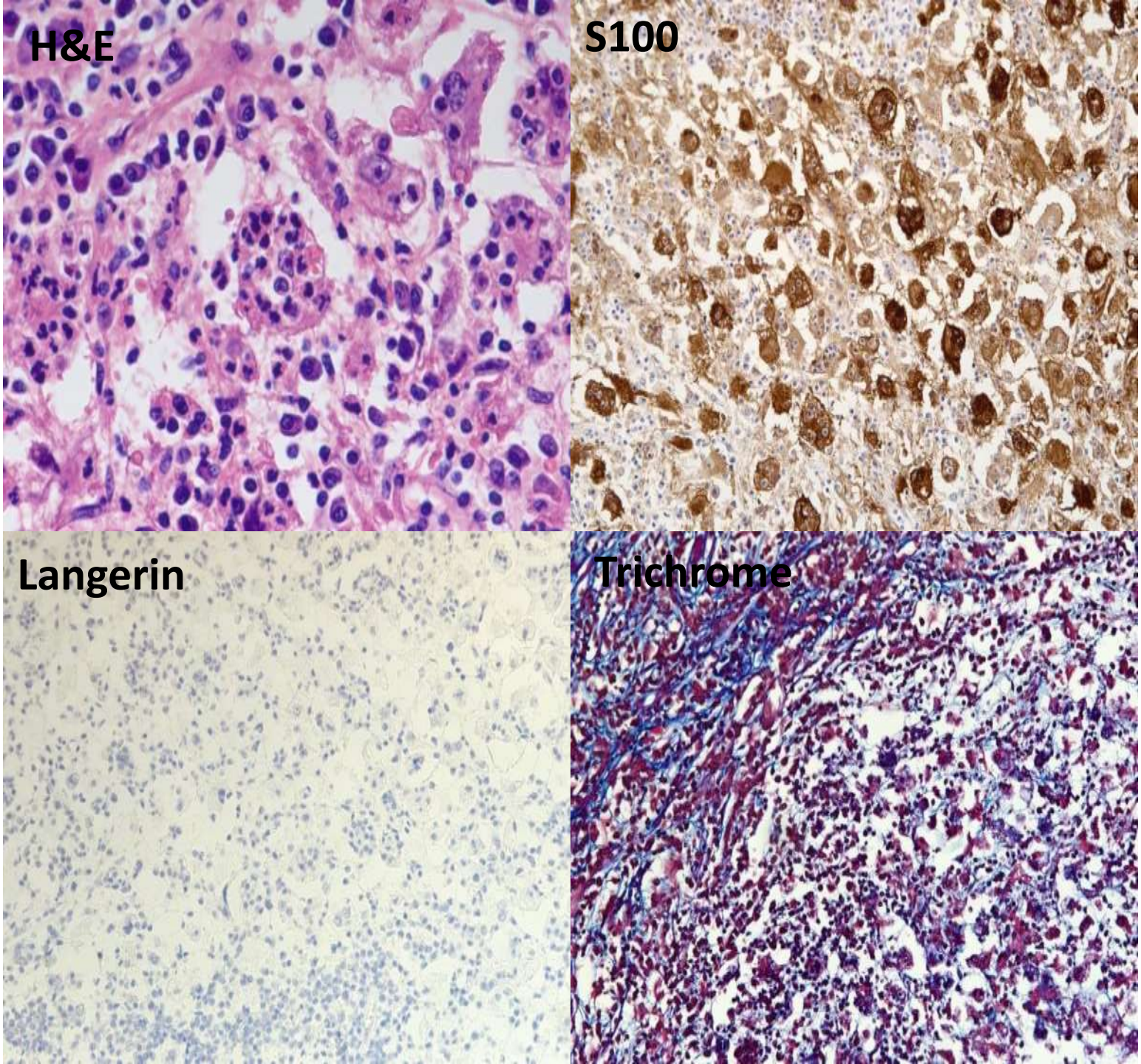
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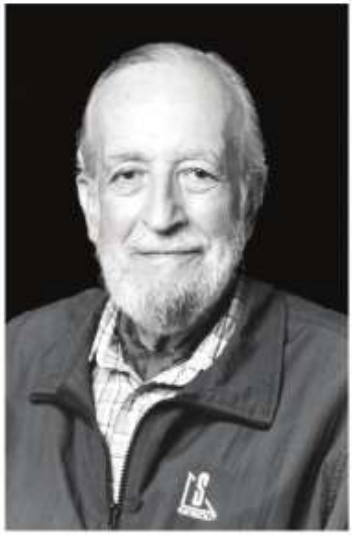
S100
Langerin
Trichrome

Extranodal Rosai-Dorfman Disease

Introduction

- Described in 1969 by Rosai and Dorfman
  - Sinus histiocytosis with massive lymphadenopathy
- Manifests in children and young adults most commonly
  - Mean age 20 years old
  - Can occur at any age
- At presentation
  - Cervical lymphadenopathy (87%)
  - Extranodal sites with or without involvement of lymph nodes (up to 43%)
  - Constitutional symptoms: fever, weight loss
  - Organ specific symptoms



Credit to Stanford School of Medicine

Extranodal Rosai-Dorfman Disease

Clinical Course

- Heterogeneous clinical course
- Benign, self limited
  - Younger age
  - Limited number of lymph nodes involved
- Aggressive, chronic relapses
  - Older age
  - Underlying immunologic abnormalities
  - Numerous lymph nodes involved
  - Extranodal organ involvement

Etiology

- Unknown
  - Florid reactive process vs. clonal proliferation
  - Immune-mediated disorder vs. virus-mediated disorder
- Macrophage colony-stimulating factor (M-CSF)
  - Induces chemotaxis of blood monocytes to site of lesion
  - Induces differentiation of monocytes to histiocytes
  - Induces differentiation of monocytes to mature macrophages
    - Phagocytosis
  - Monocytes secrete more M-CSF
    - Cycle continues

Extranodal Rosai-Dorfman Disease

Renal manifestations

- Extranodal disease with associated lymphadenopathy and systemic disease
  - Approximately 4%
- Extranodal disease without associated lymphadenopathy and systemic disease
  - Very Rare (~ 4 cases reported)

Our Patient

- Recent infection with Influenza A
- Treated for pericarditis at time diagnosis
- Imaging showed both renal and pancreatic mass but no lymphadenopathy

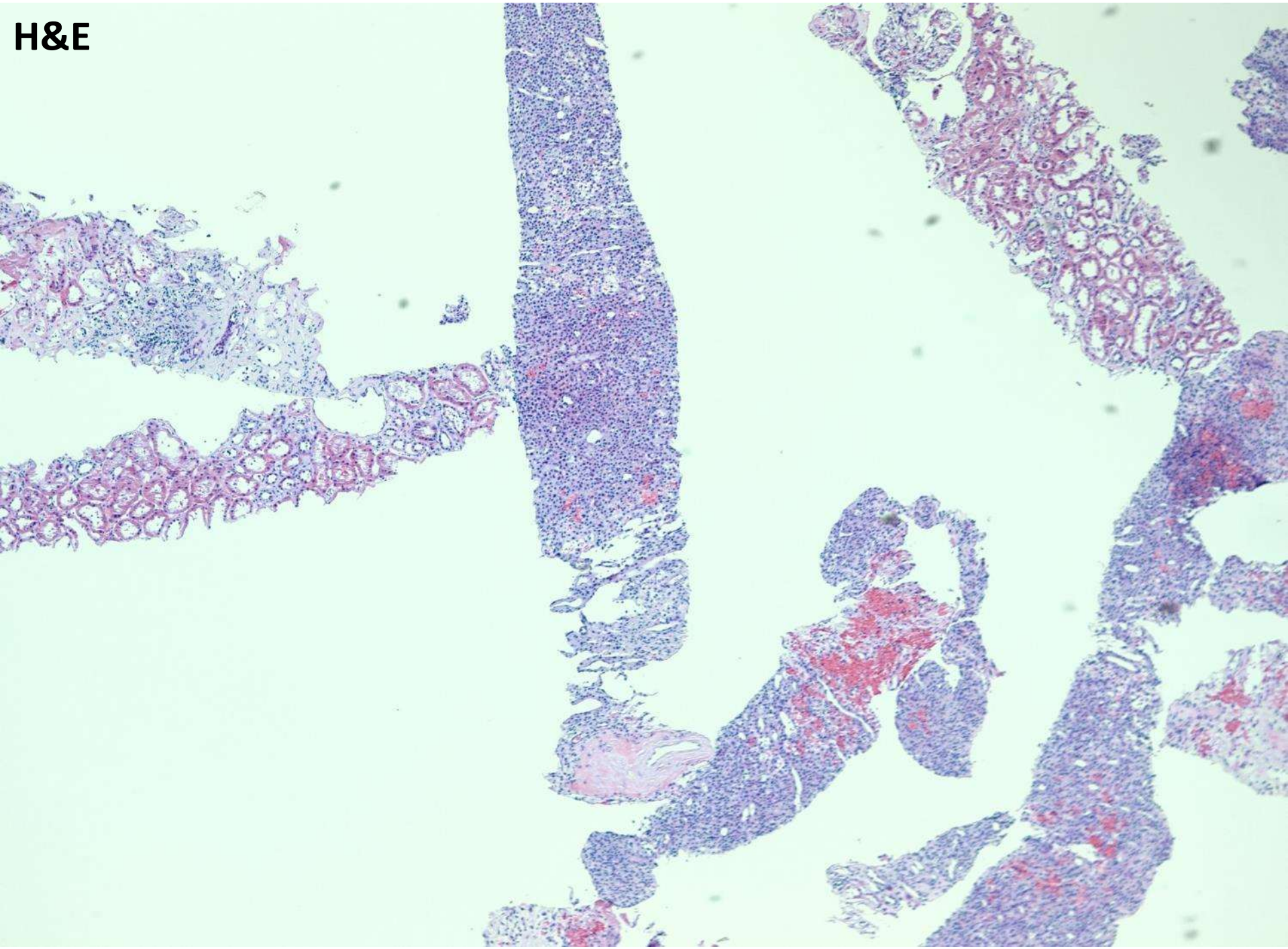
Extranodal Rosai-Dorfman Disease

- Take away points
  - Imaging
    - Rosai-Dorfman disease may overlap with neoplastic and non-neoplastic disease
    - Is not frequently considered in the differential diagnosis of an infiltrative renal mass
  - Diagnosis
    - Based on morphology and immunohistochemistry
      - Infiltrate composed of lymphocytes, plasma cells and histiocytes
      - Emperipolesis
      - Positive for S100, CD68 and CD163 on immunohistochemistry (negative for CD1a and Langerin)

20-0402

Julia Rewerska/Christian Kunder; Stanford

39-year-old F with slow growing 1.9cm left renal mass. PMH significant for dissecting aortic aneurysm.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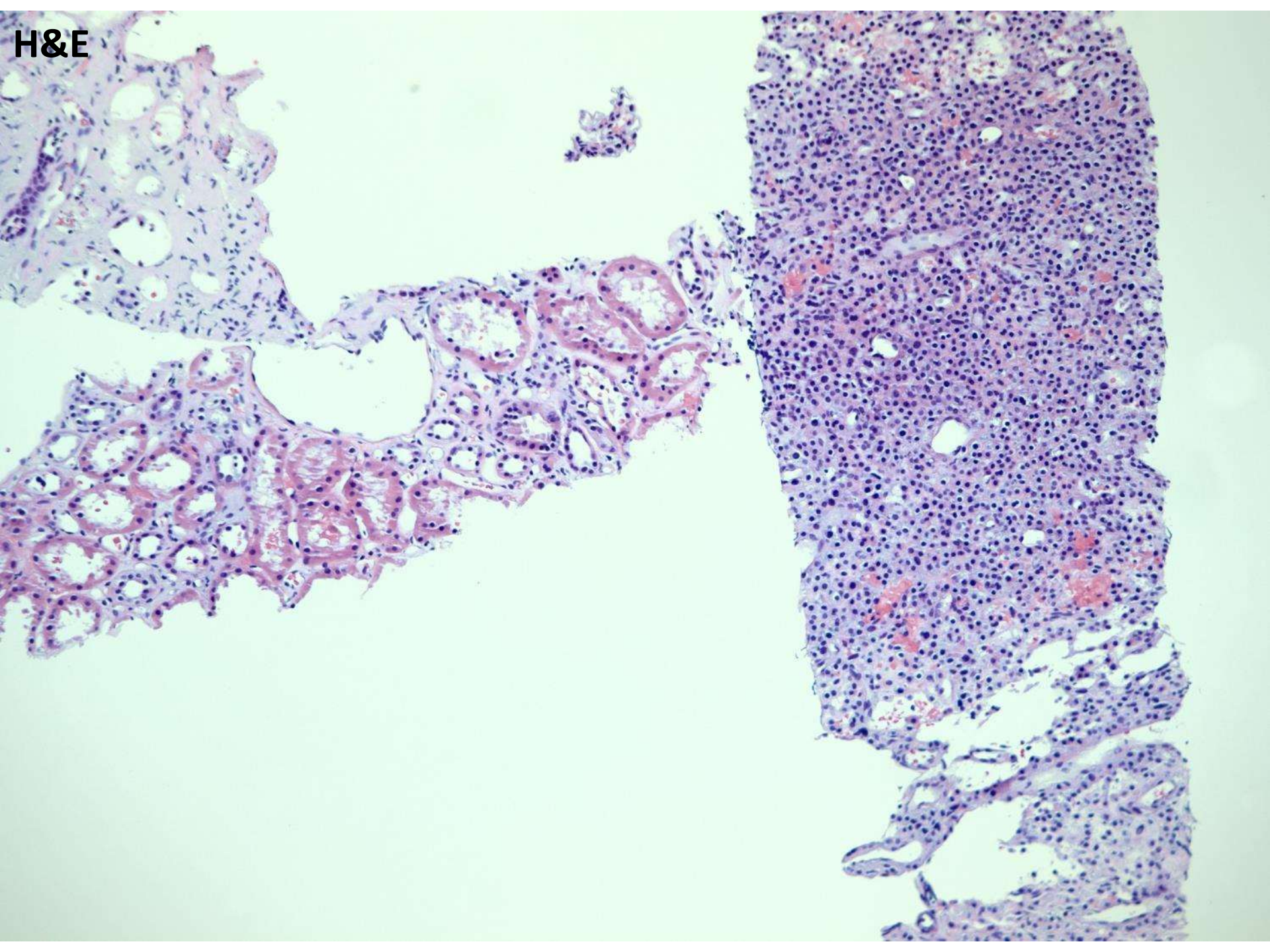
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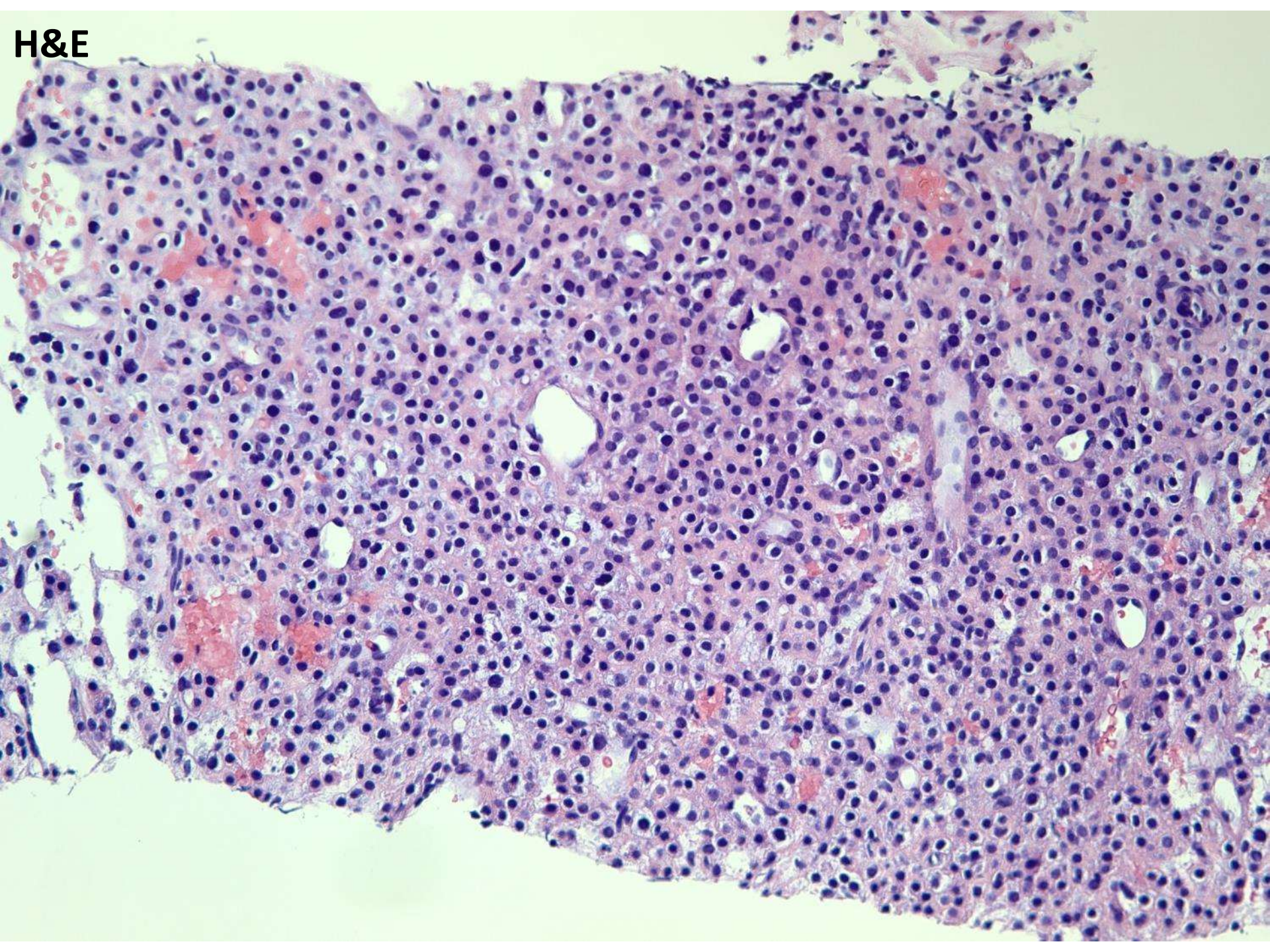
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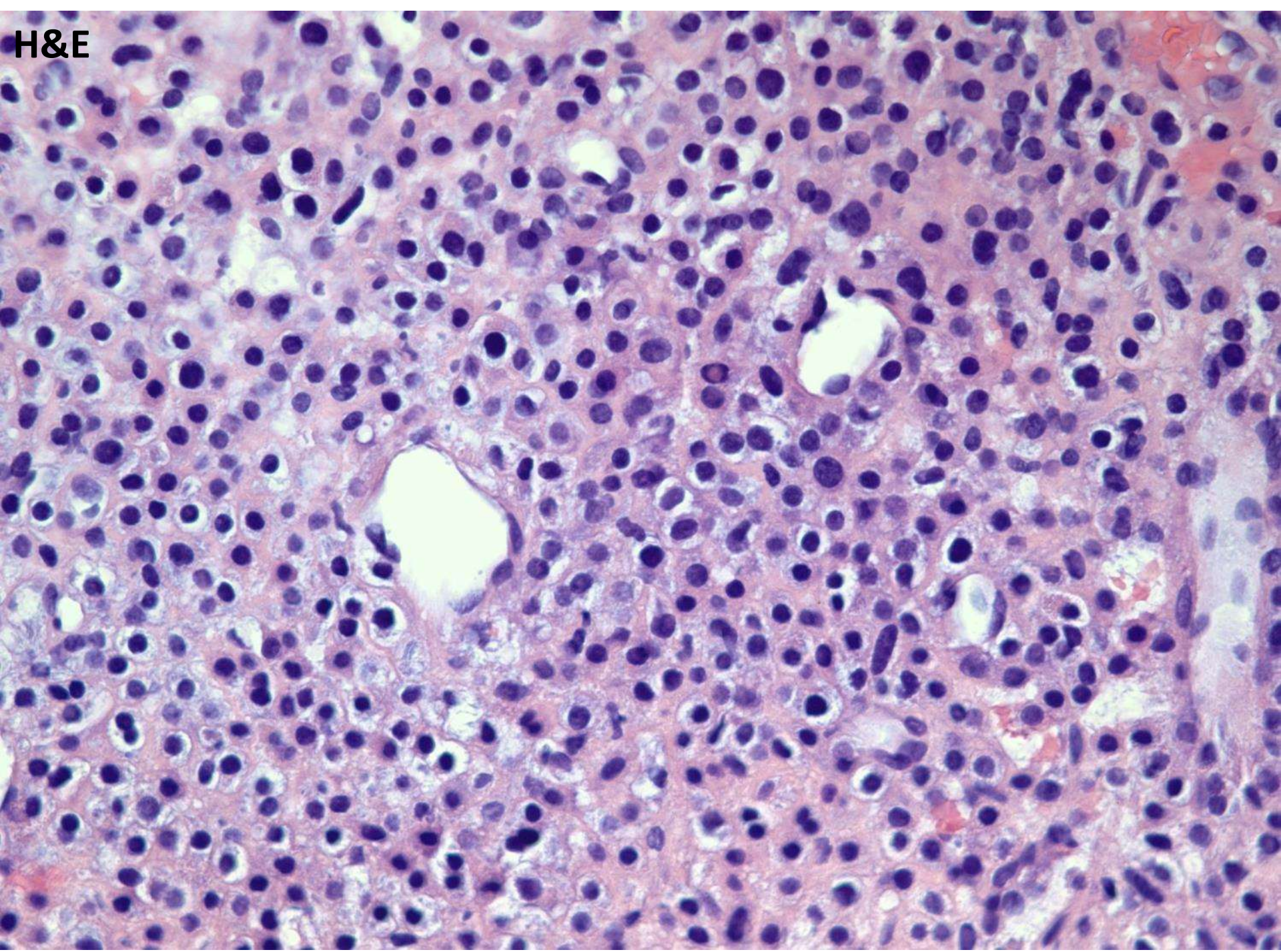
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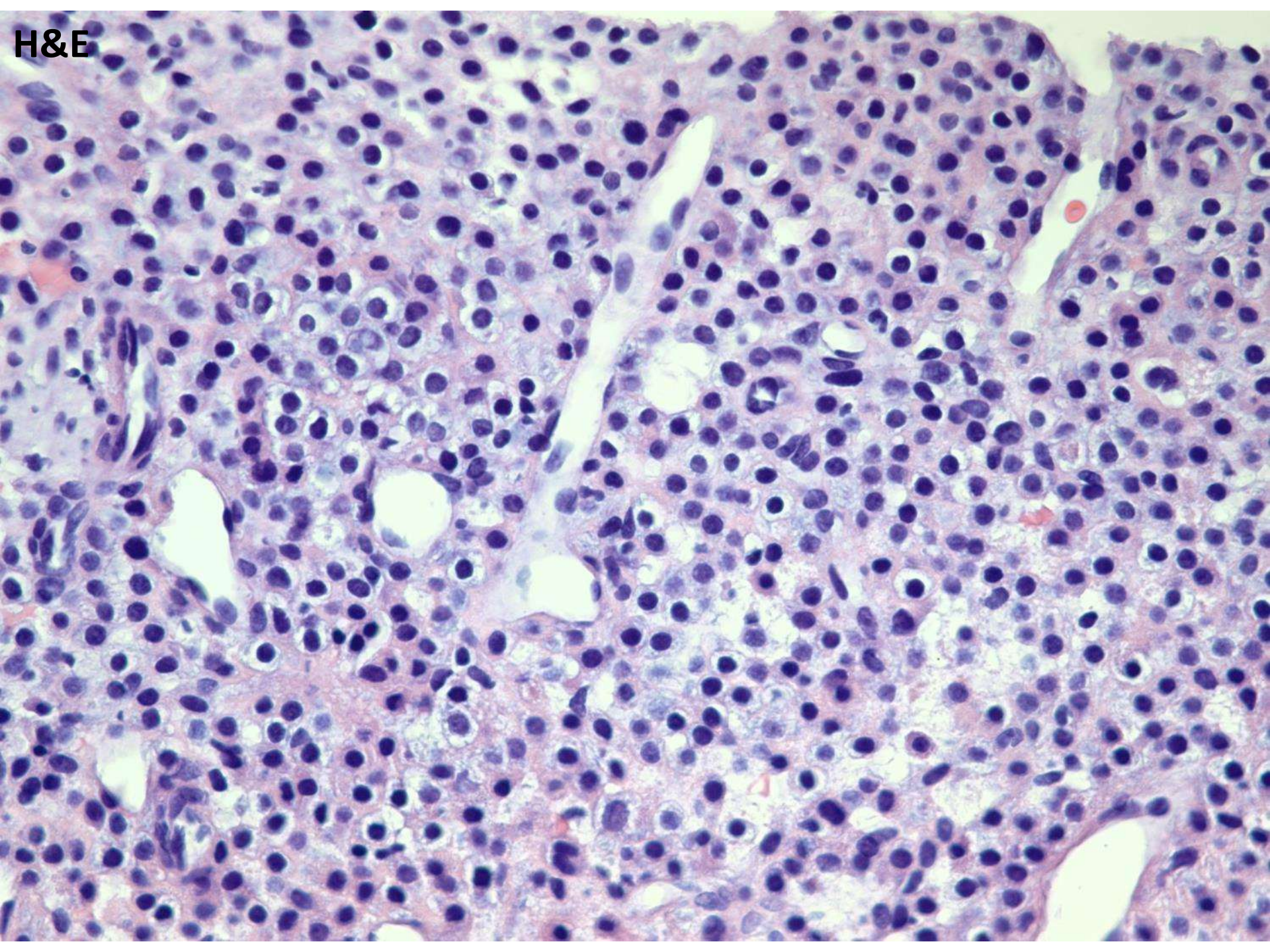
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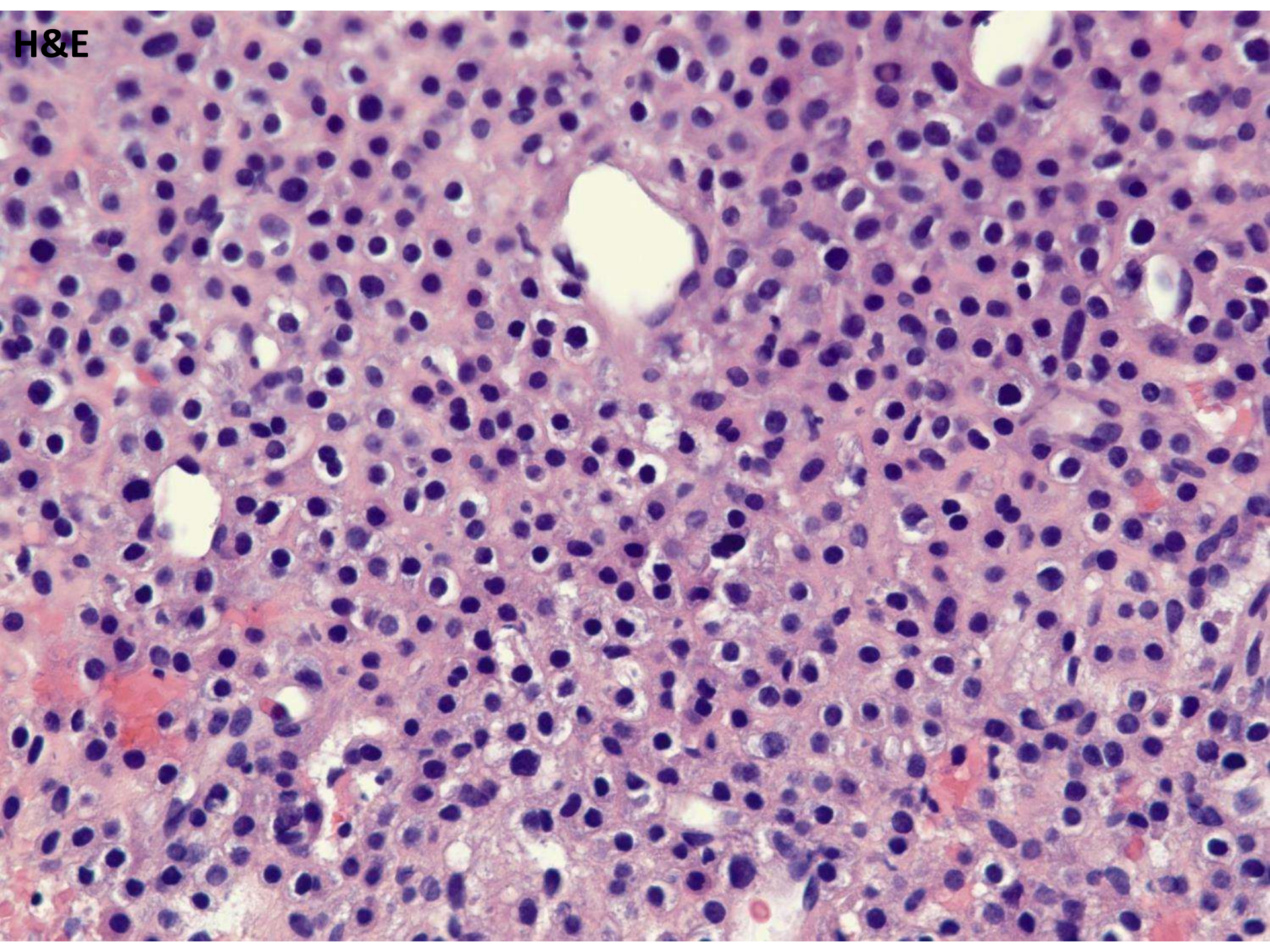
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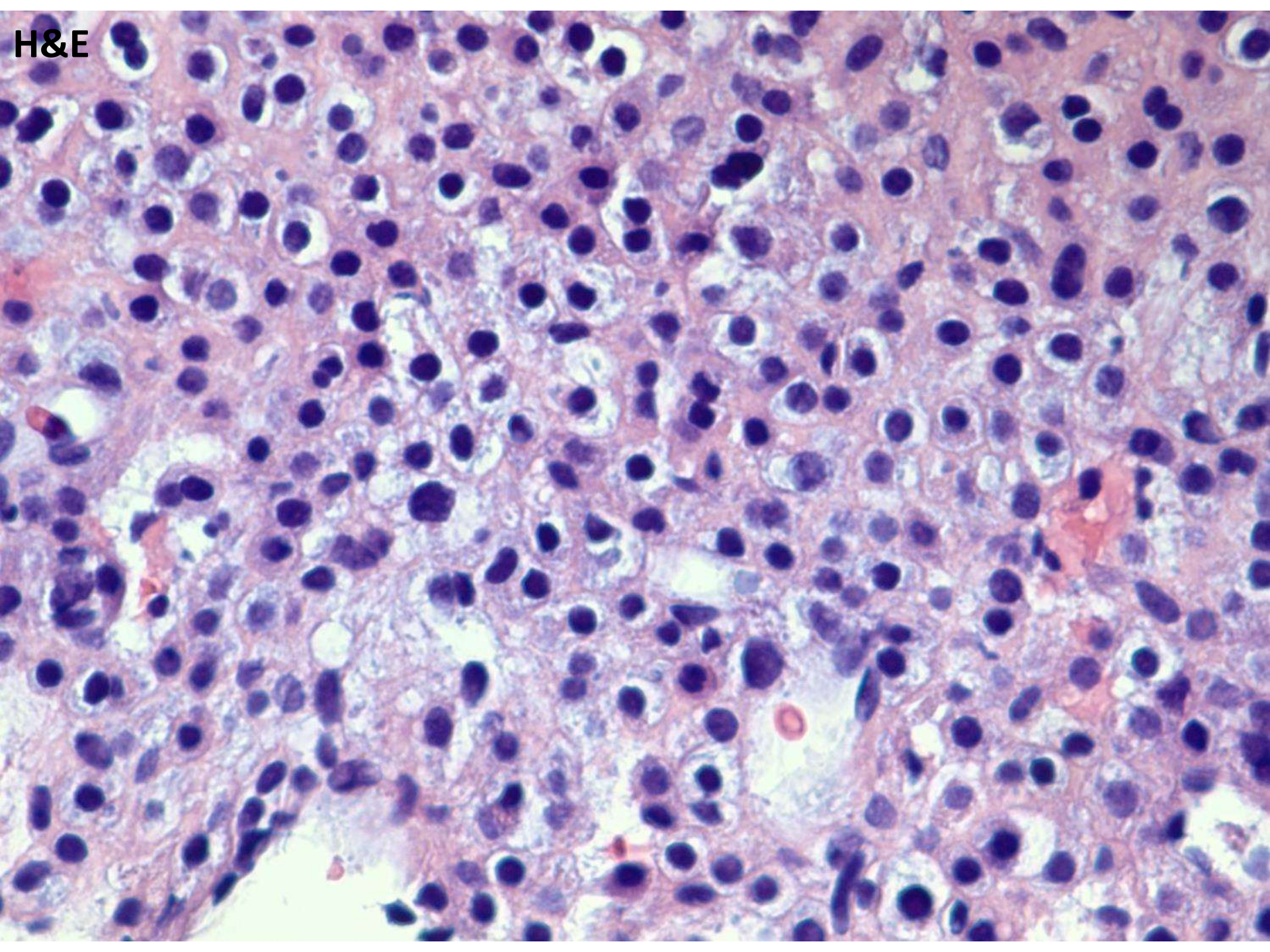
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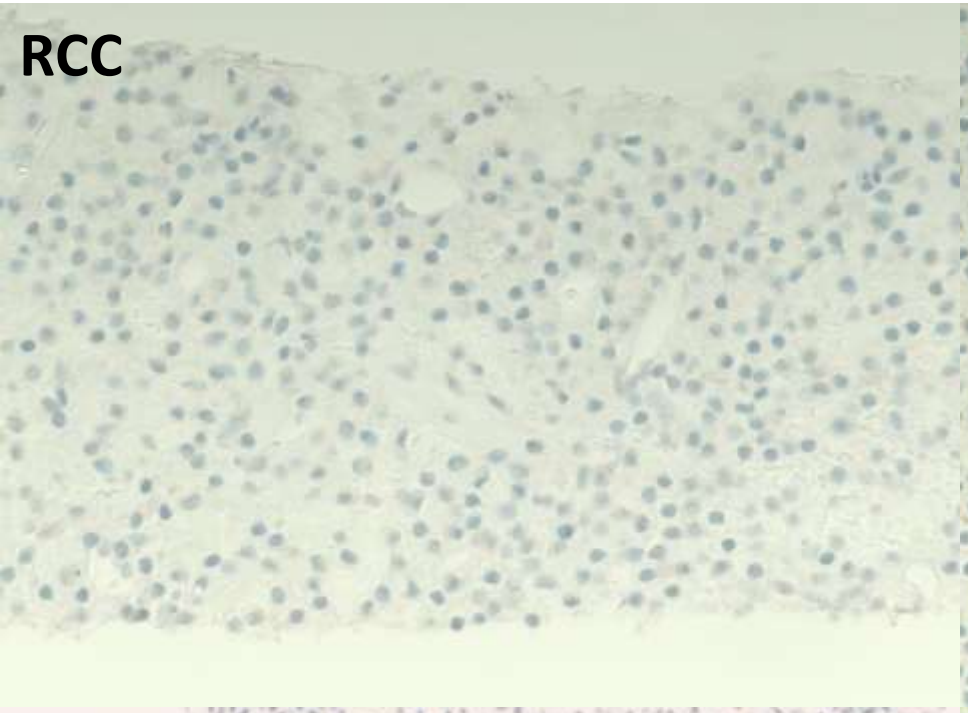
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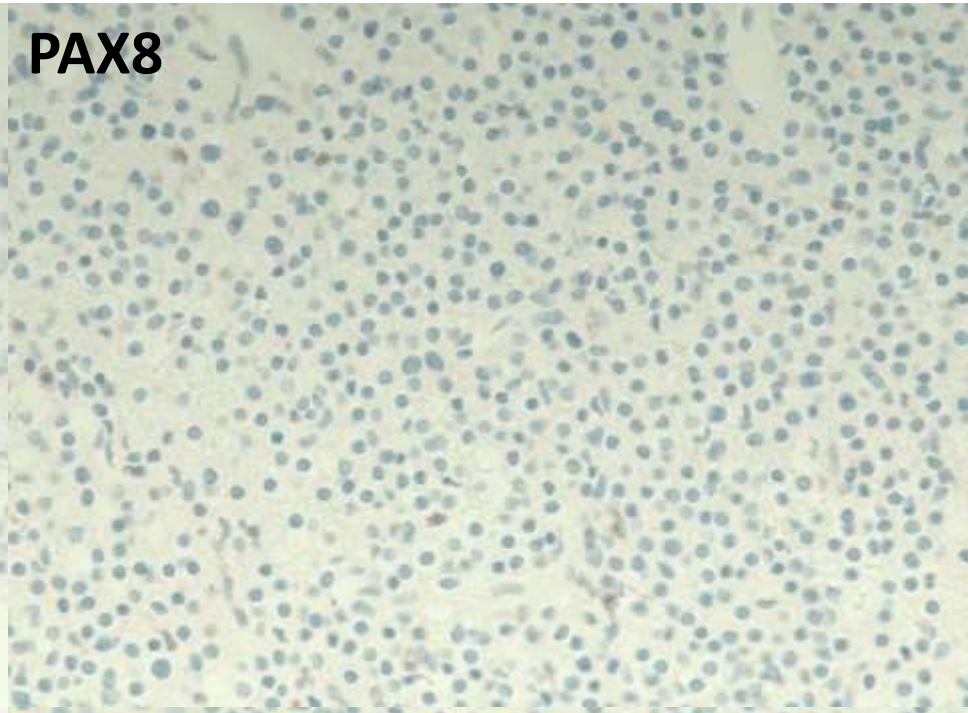
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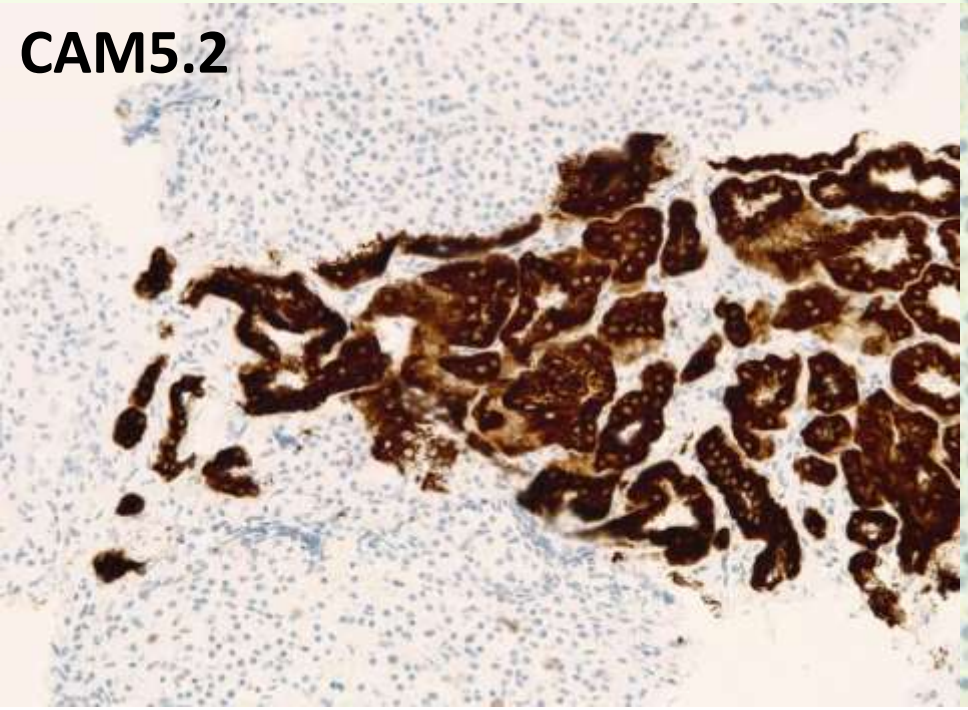
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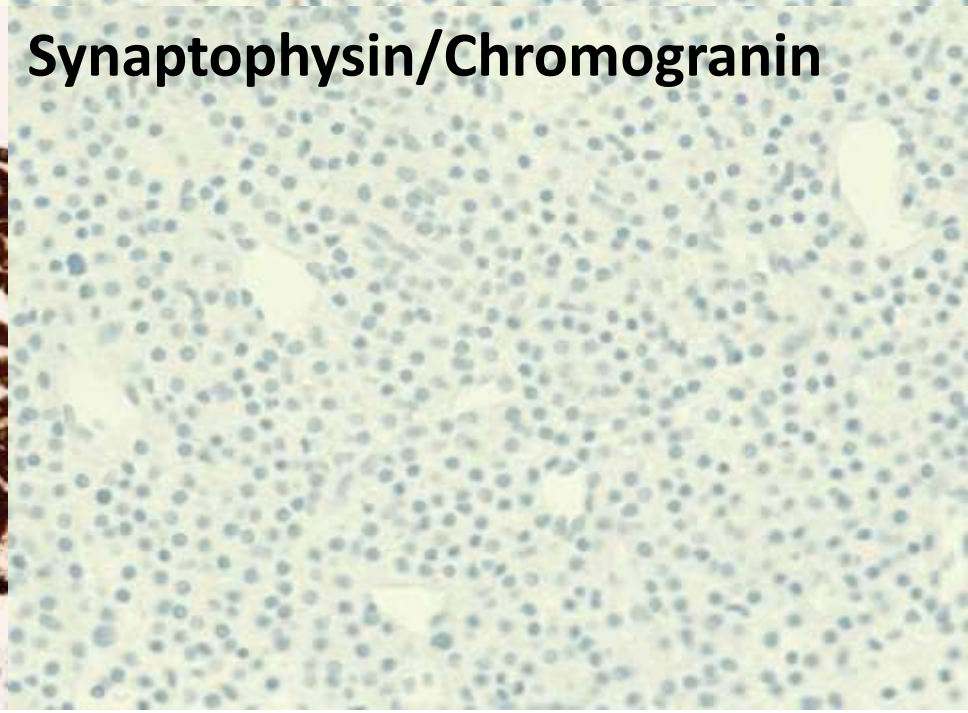
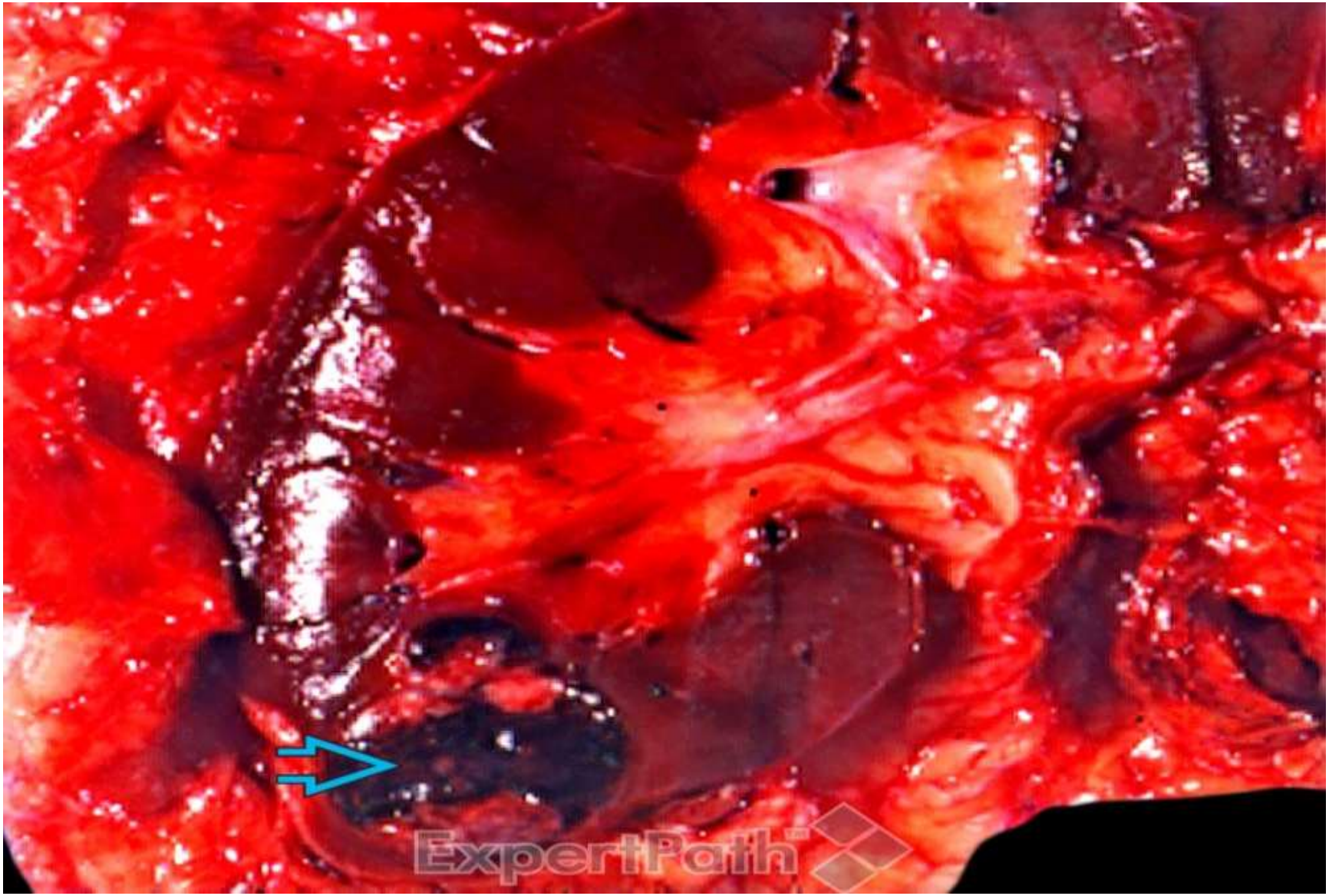
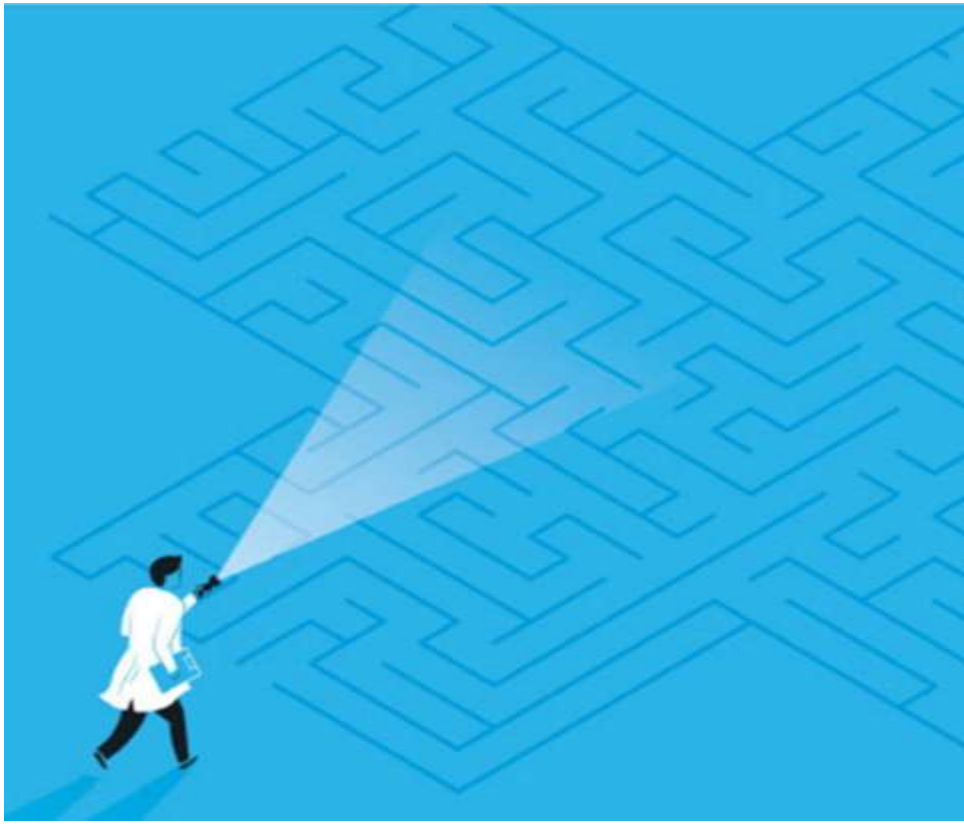

Synaptophysin/Chromogranin

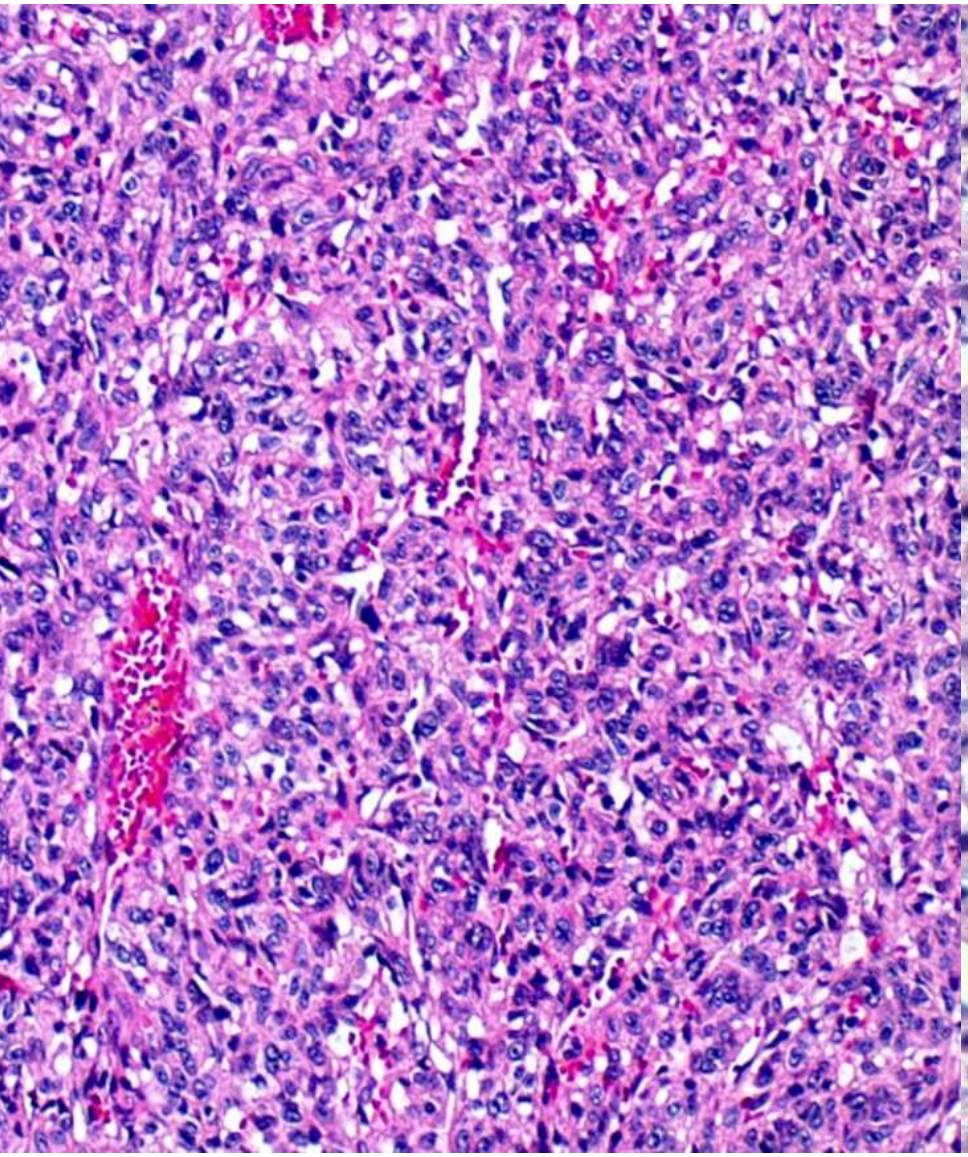

ExpertPath

Differential Diagnoses

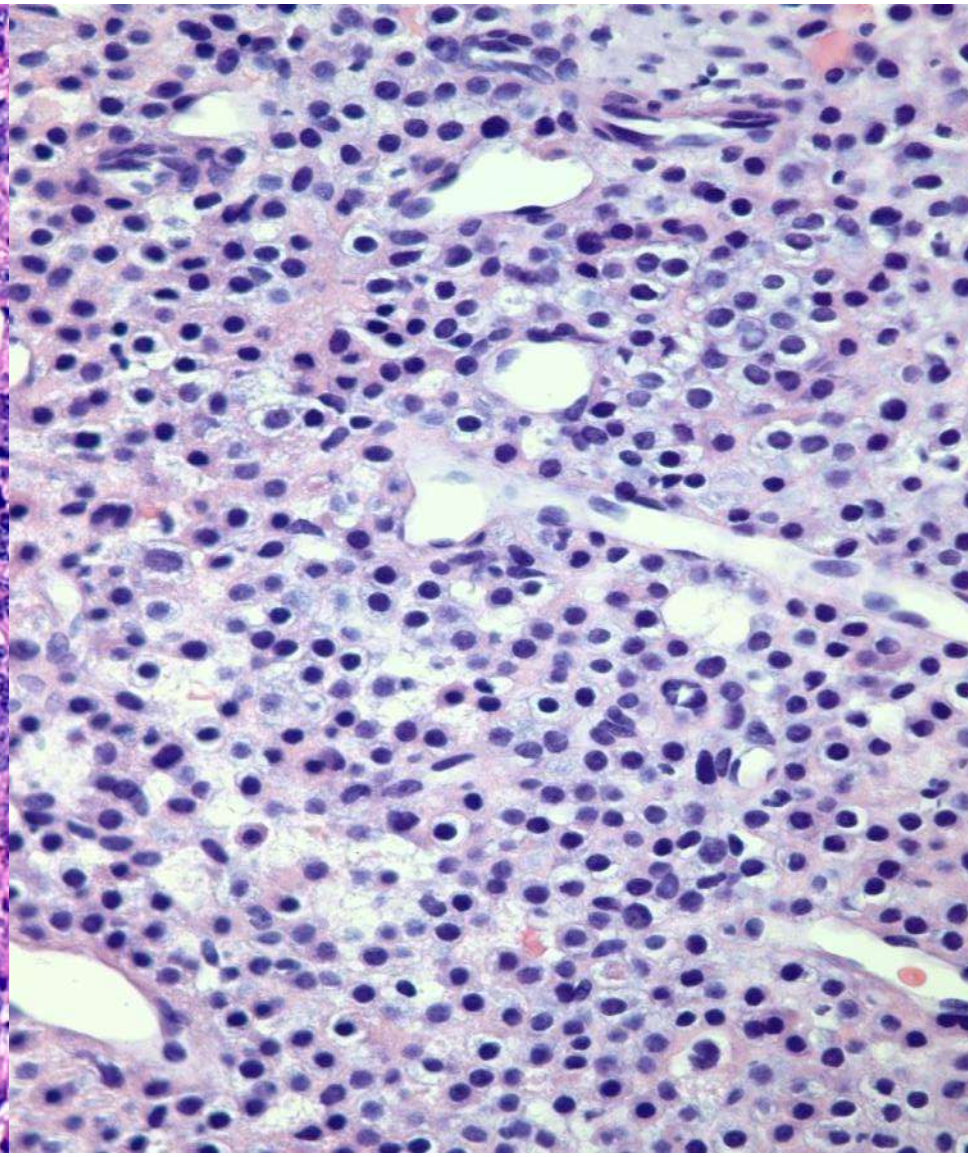
- Glomus Tumor
- Juxtaglomerular cell tumor
- Oncocytic renal cell neoplasm
- Solitary fibrous tumor/hemangiopericytoma

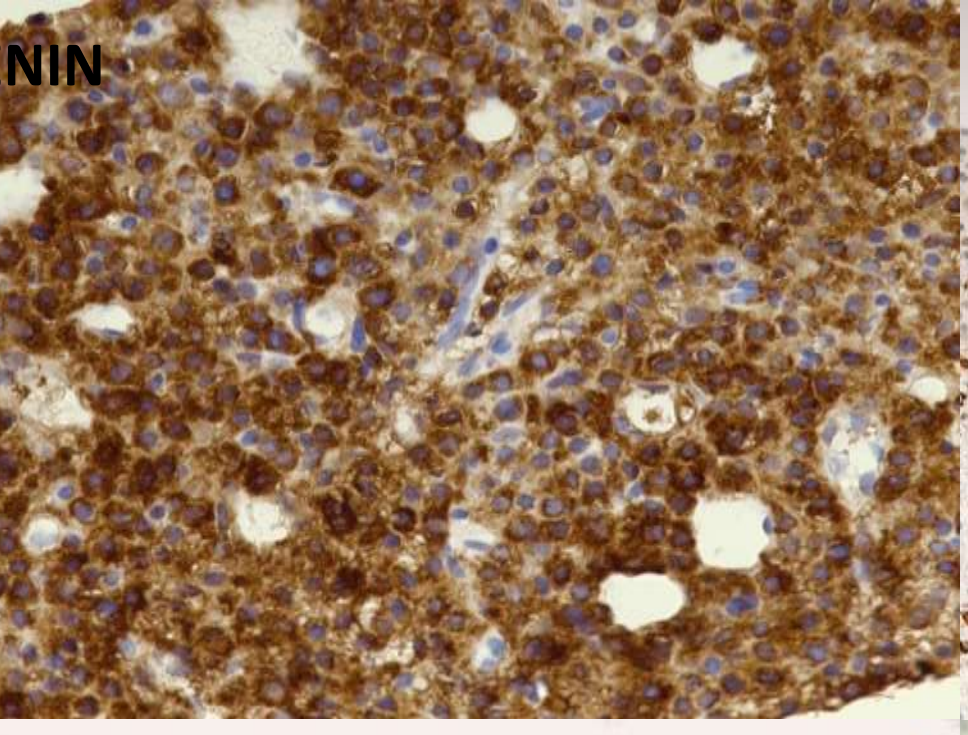


omus Tumor



Our Tumor

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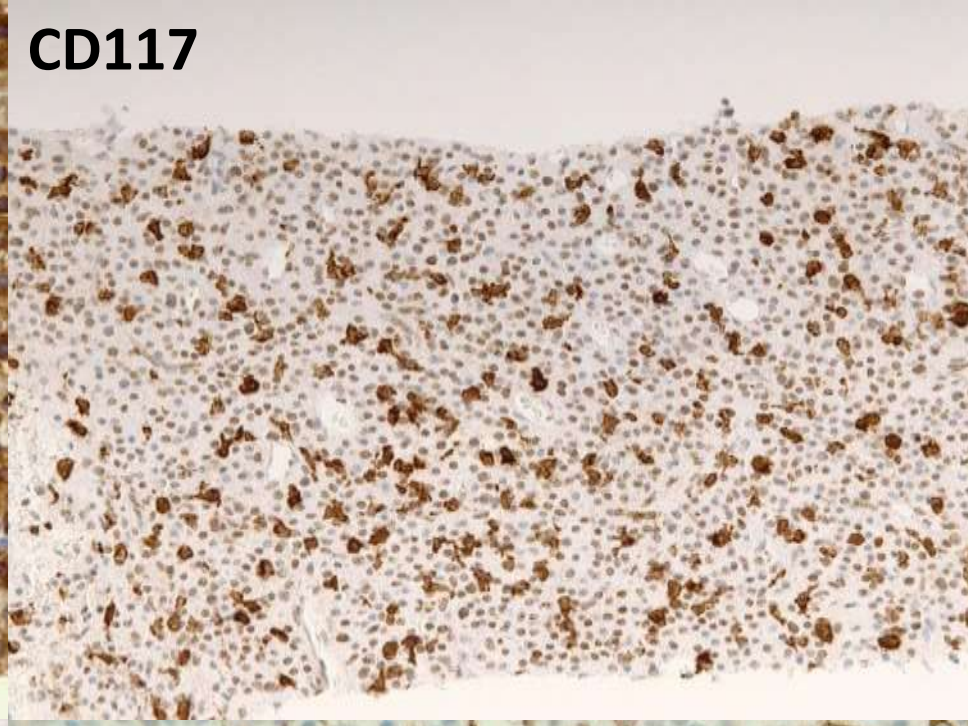
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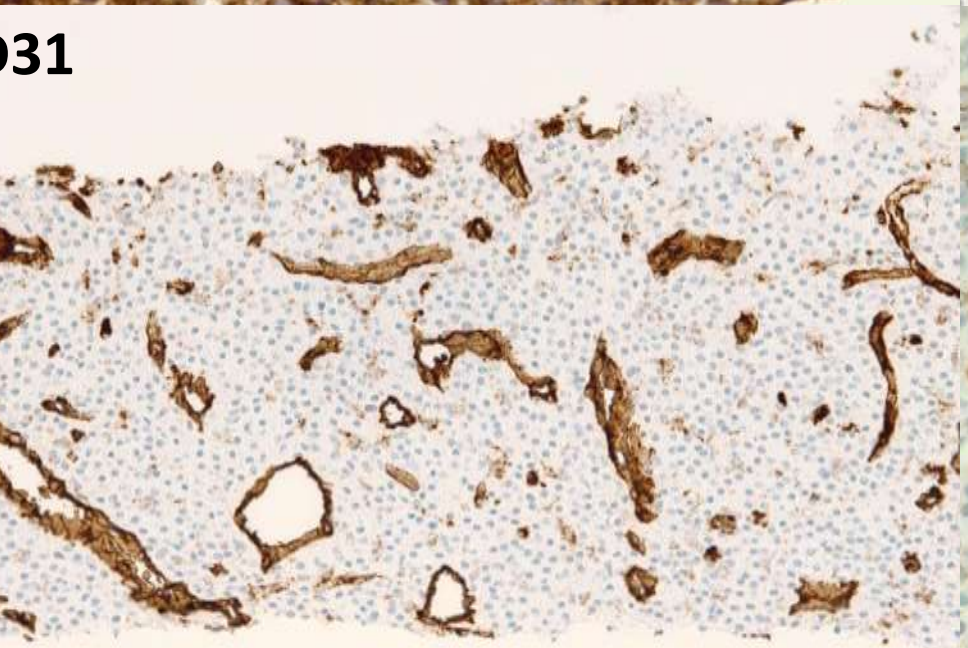
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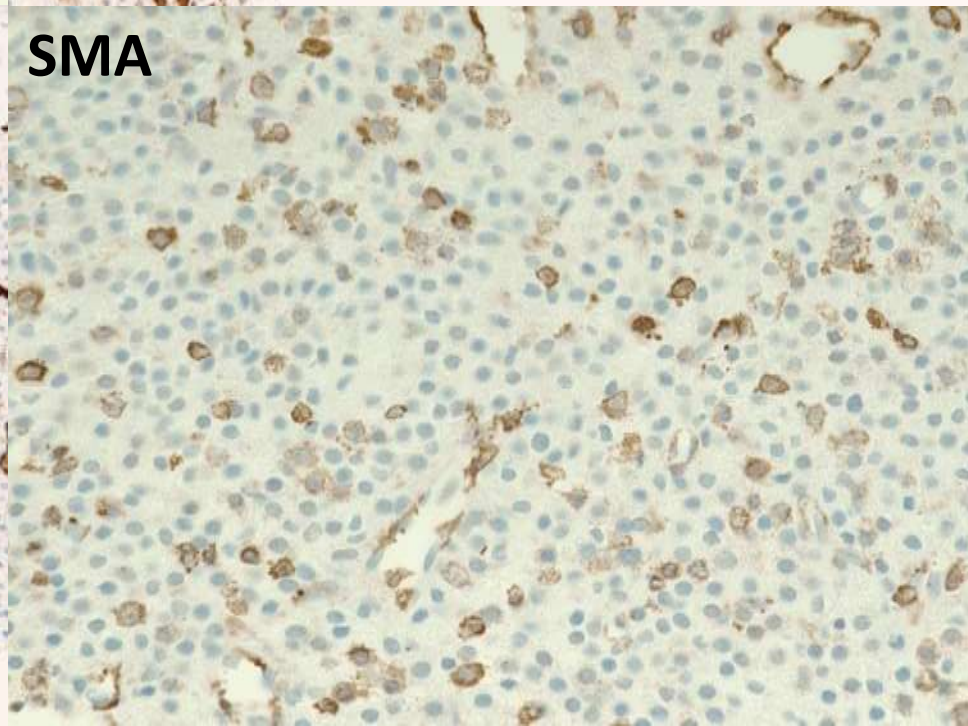
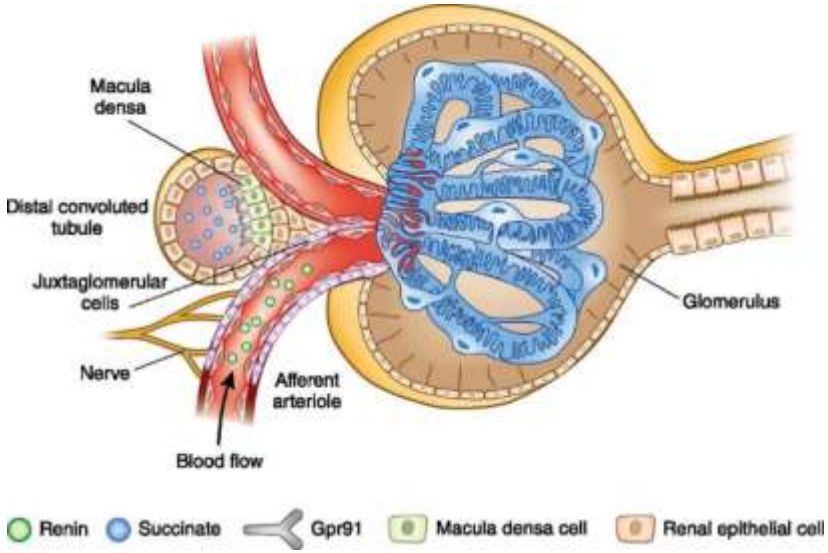

SMA

Juxtaglomerular Cell Tumor

Introduction

- Rare <100 cases reported
- Tumor derived from specialized smooth muscle cells of the juxtaglomerular apparatus
  - Hyperreninism
  - Hypokalemia
  - Hyperaldosteronism
- Usually manifests in young adults
  - 2nd to 3rd decade
  - Range 6 - >80 years of age
- Female to Male ratio 1.5:1

Juxtaglomerular Apparatus



Juxtaglomerular Cell Tumor

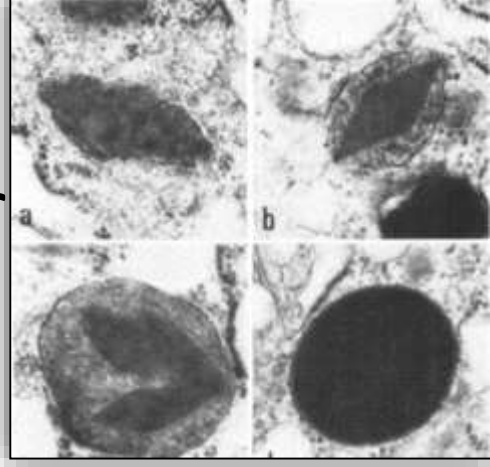
Clinical Course

- At presentation
  - Severe hypertension
  - Minimal response to medical therapy
- Treatment
  - Partial or total nephrectomy
- Prognosis
  - Benign outcome
    - One known case of lung metastasis
  - Minority of patients (<10%) with persistent hypertension
    - Secondary to hypertensive angiopathy

Etiology

- Loss of chromosomes 9 and 11
- LOH of same chromosomes
- Recurrent chromosomal abnormality
- Very few tumors tested

Juxtaglomerular Cell Tumor

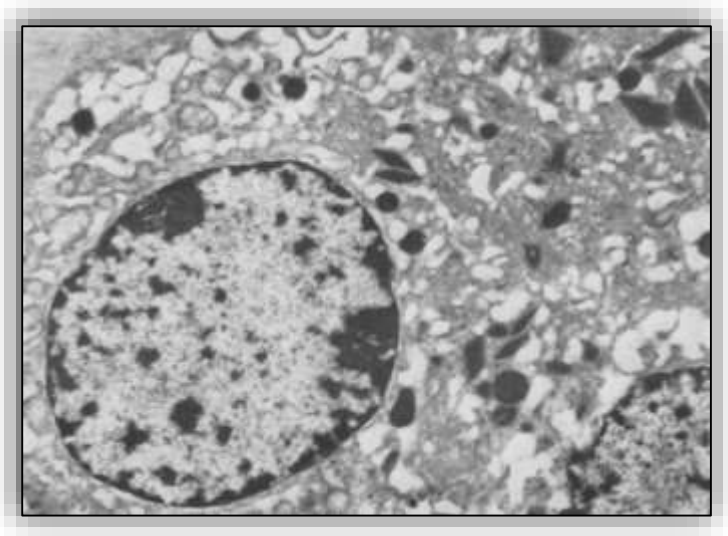


Our Patient

- 39 year-old female history of type A thoracic aortic dissection
- Systolic blood pressure
  - Average 160-180 mmHg
  - Prior to treatment 180-200 mmHg
- Scheduled surgery cancelled due to hypertension
  - Underwent cryoablation
  - BP 131/67 at hospital discharge

Take Away Points

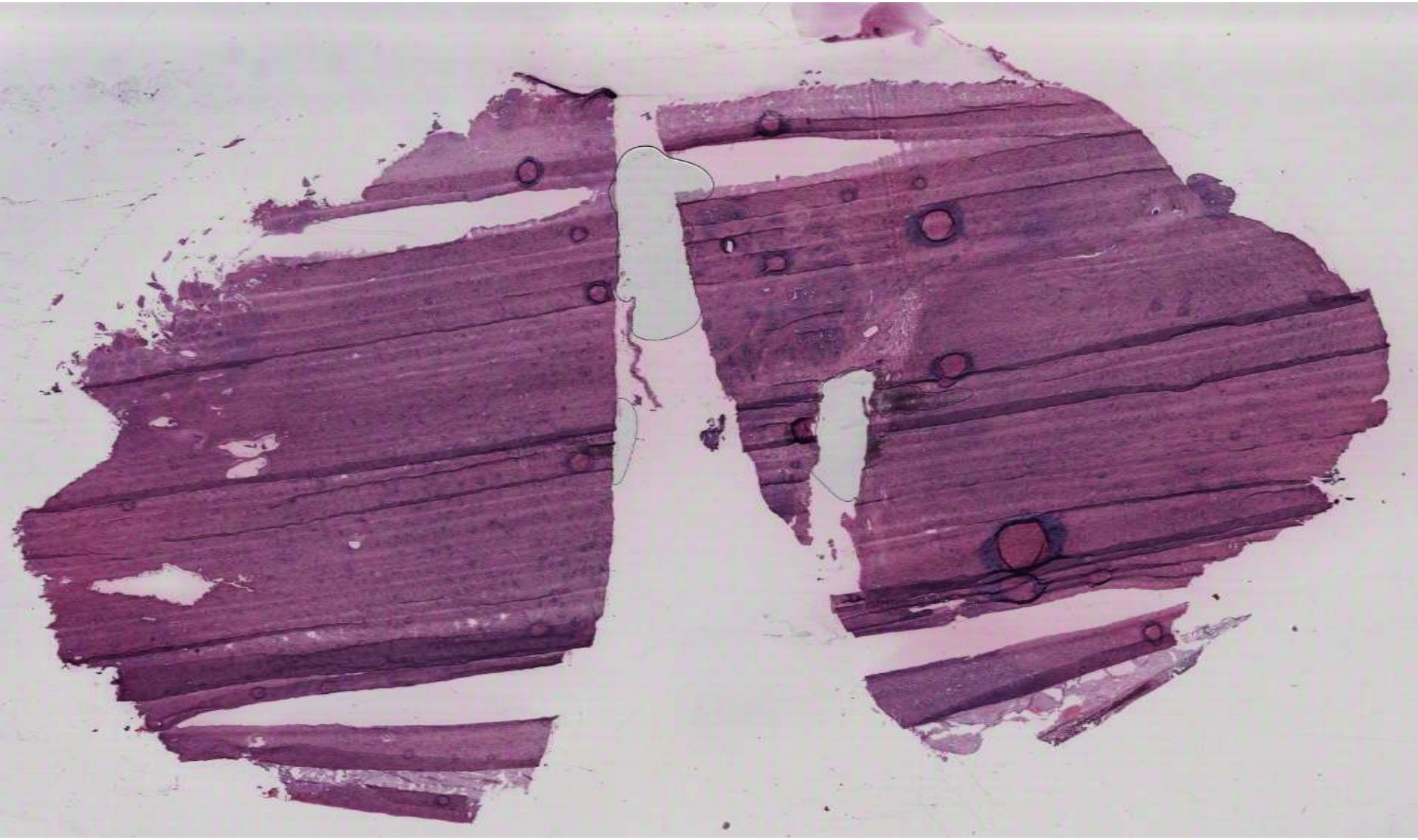
- Rare tumor
- Should prompt imaging
  - Young adults with resistant hypertension

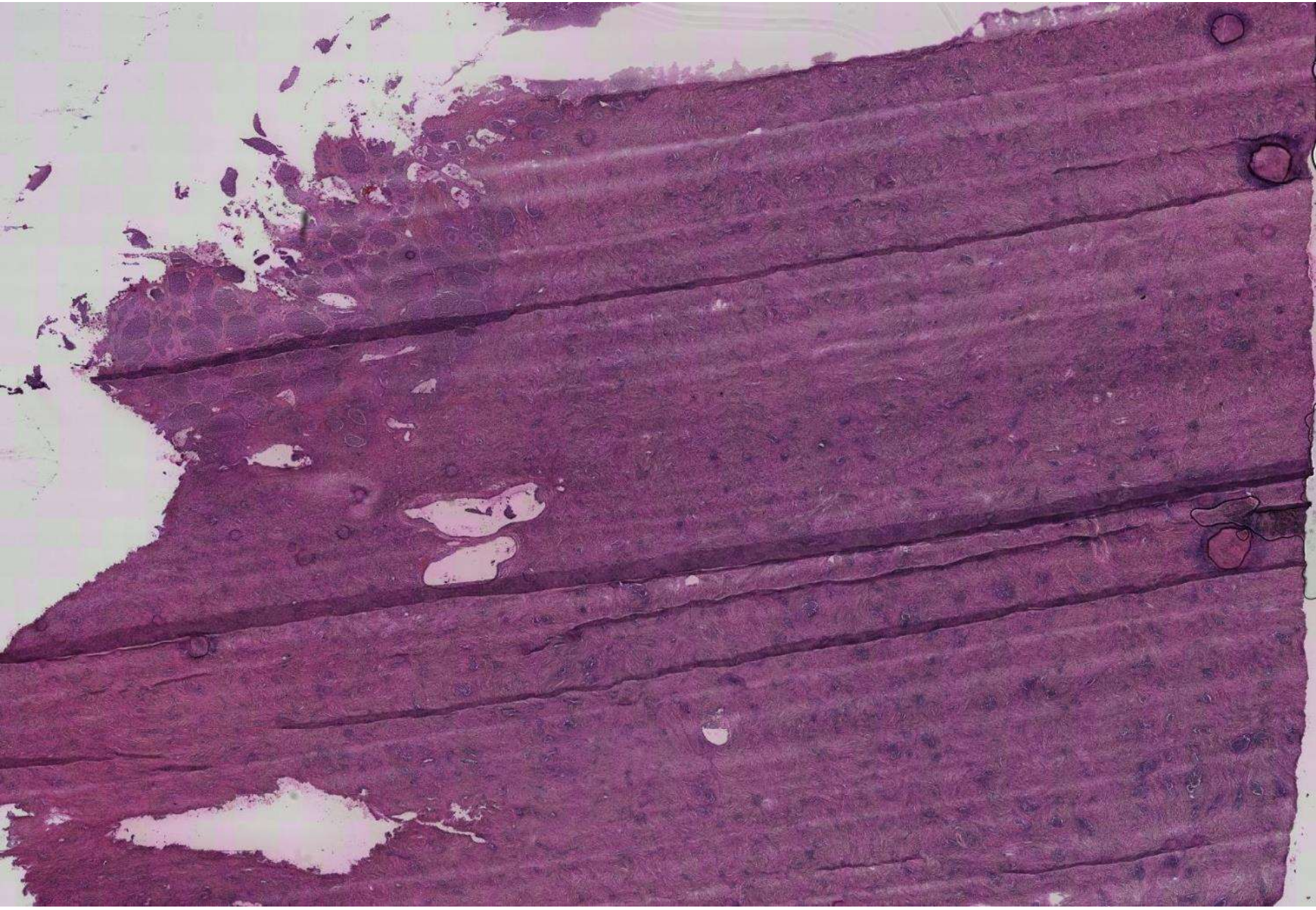


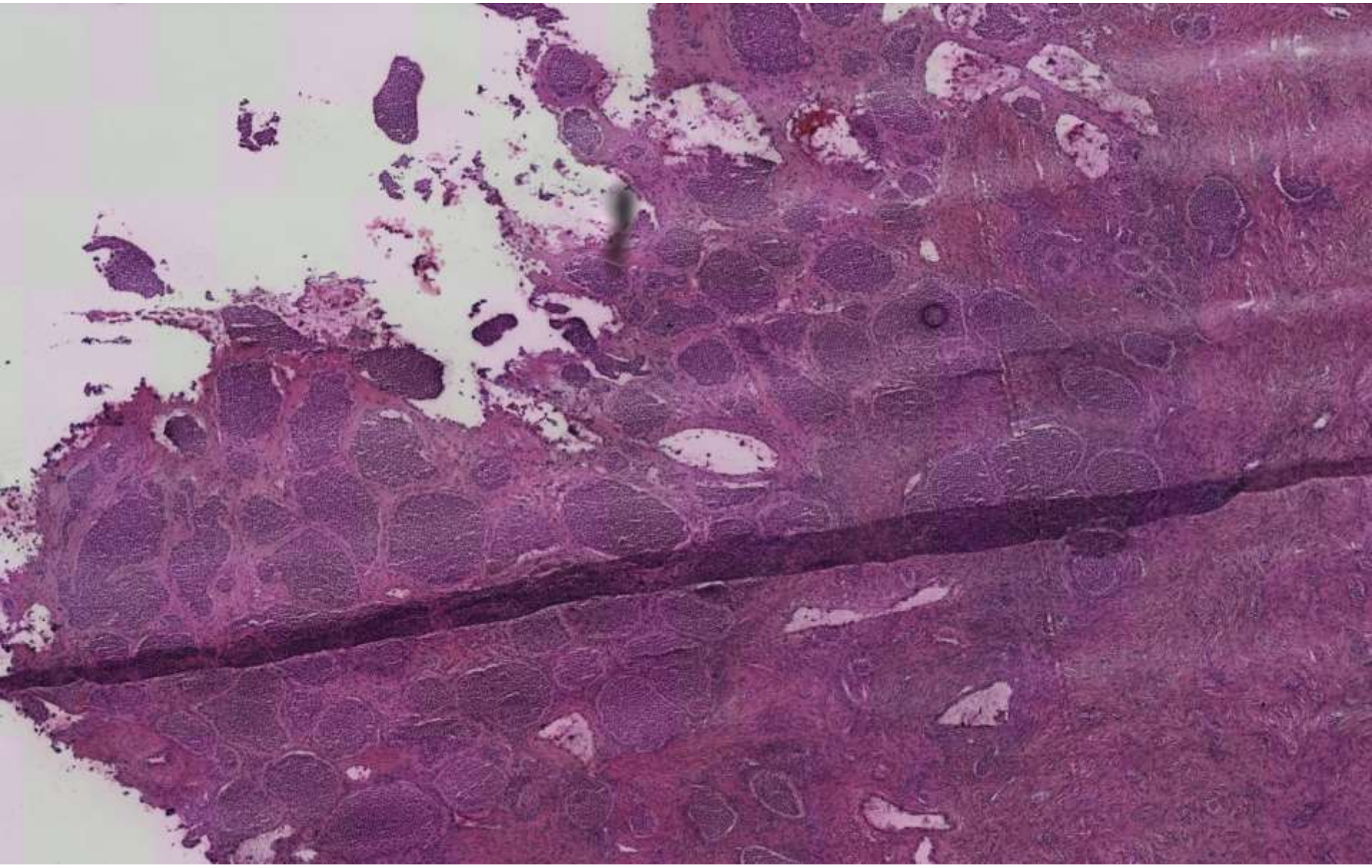
20-0403
SCANNED SLIDE AVAIL!

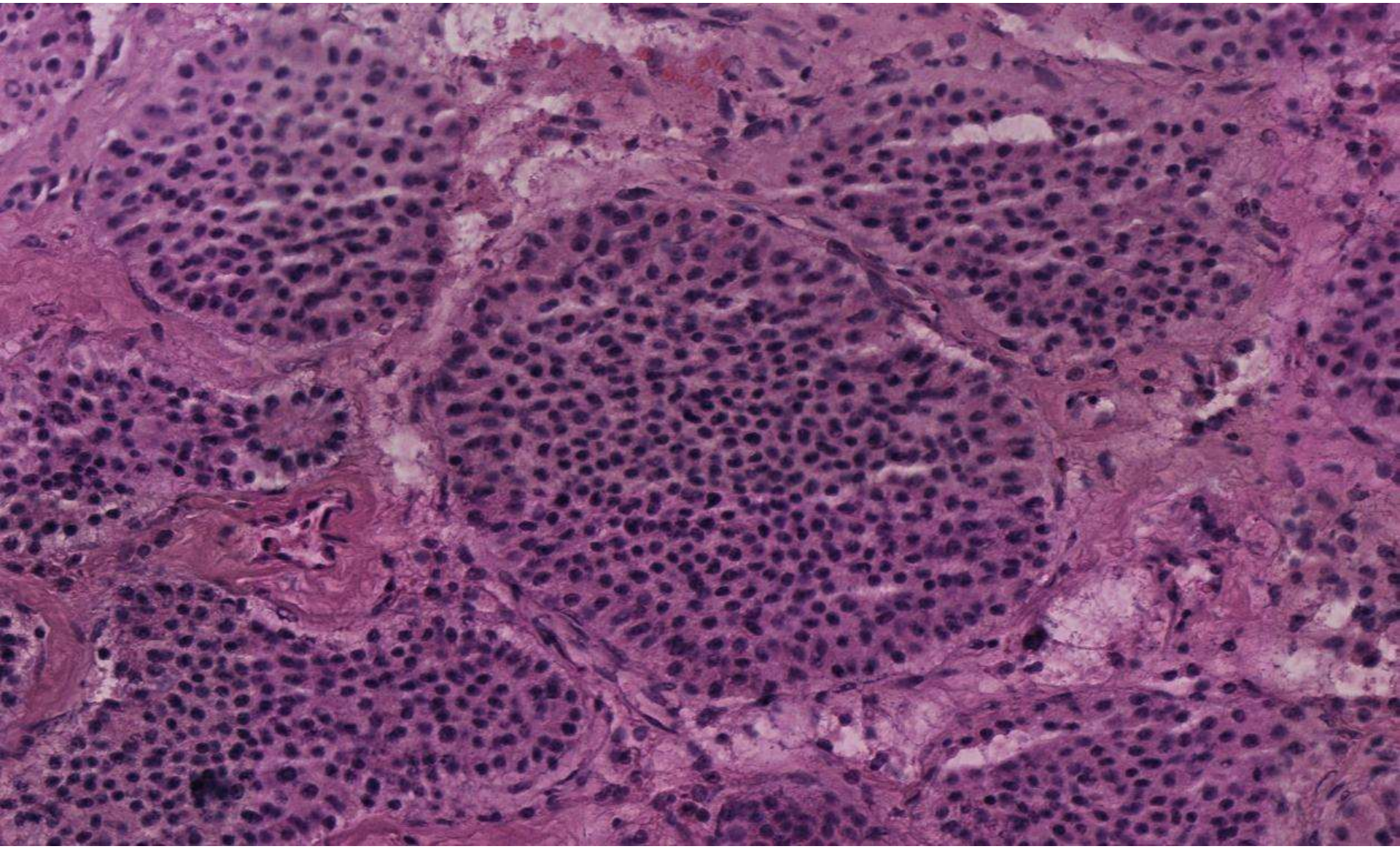
Natalie Patel; El Camino Hospital

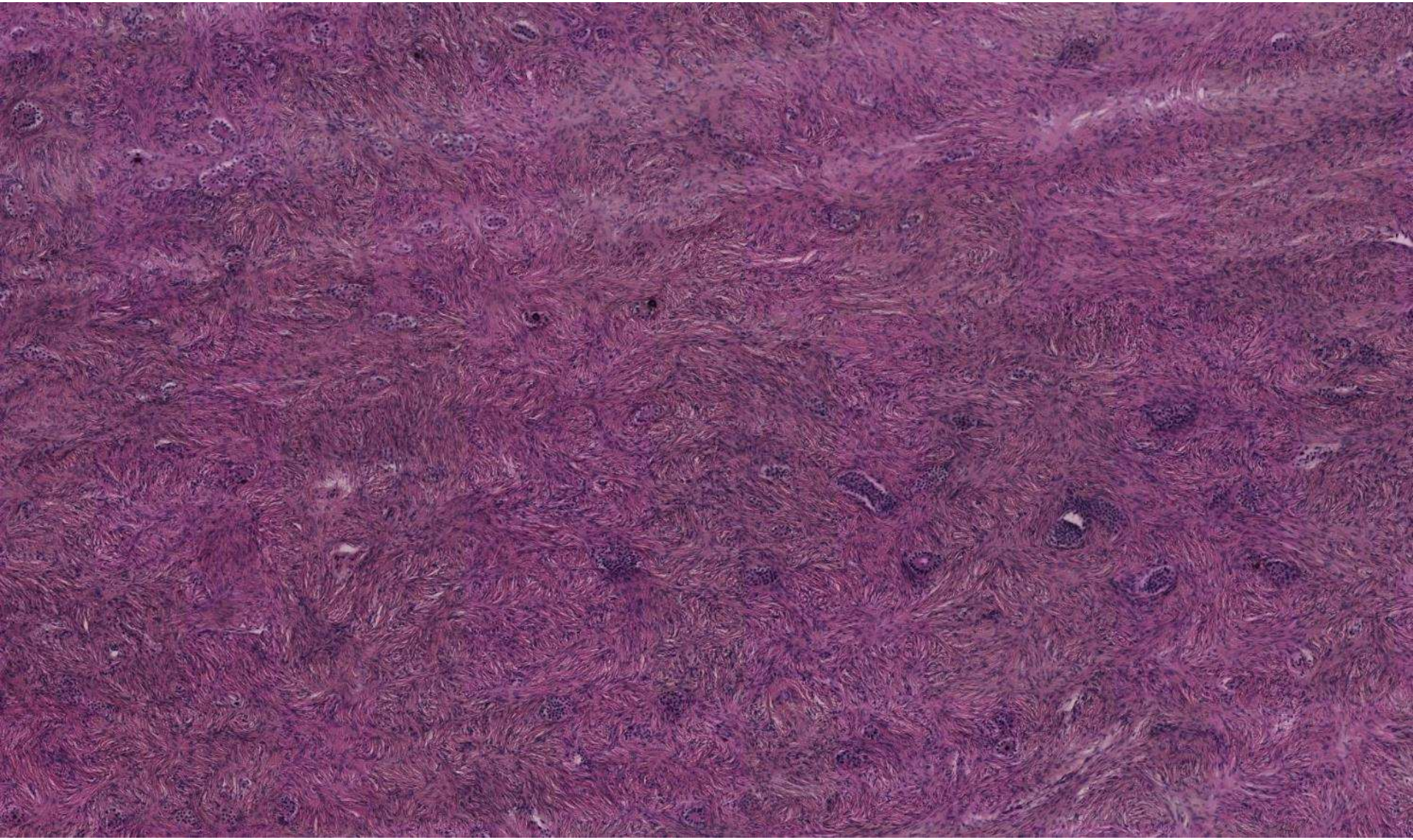
66-year-old F with 5cm right ovarian mass, submitted for frozen section.

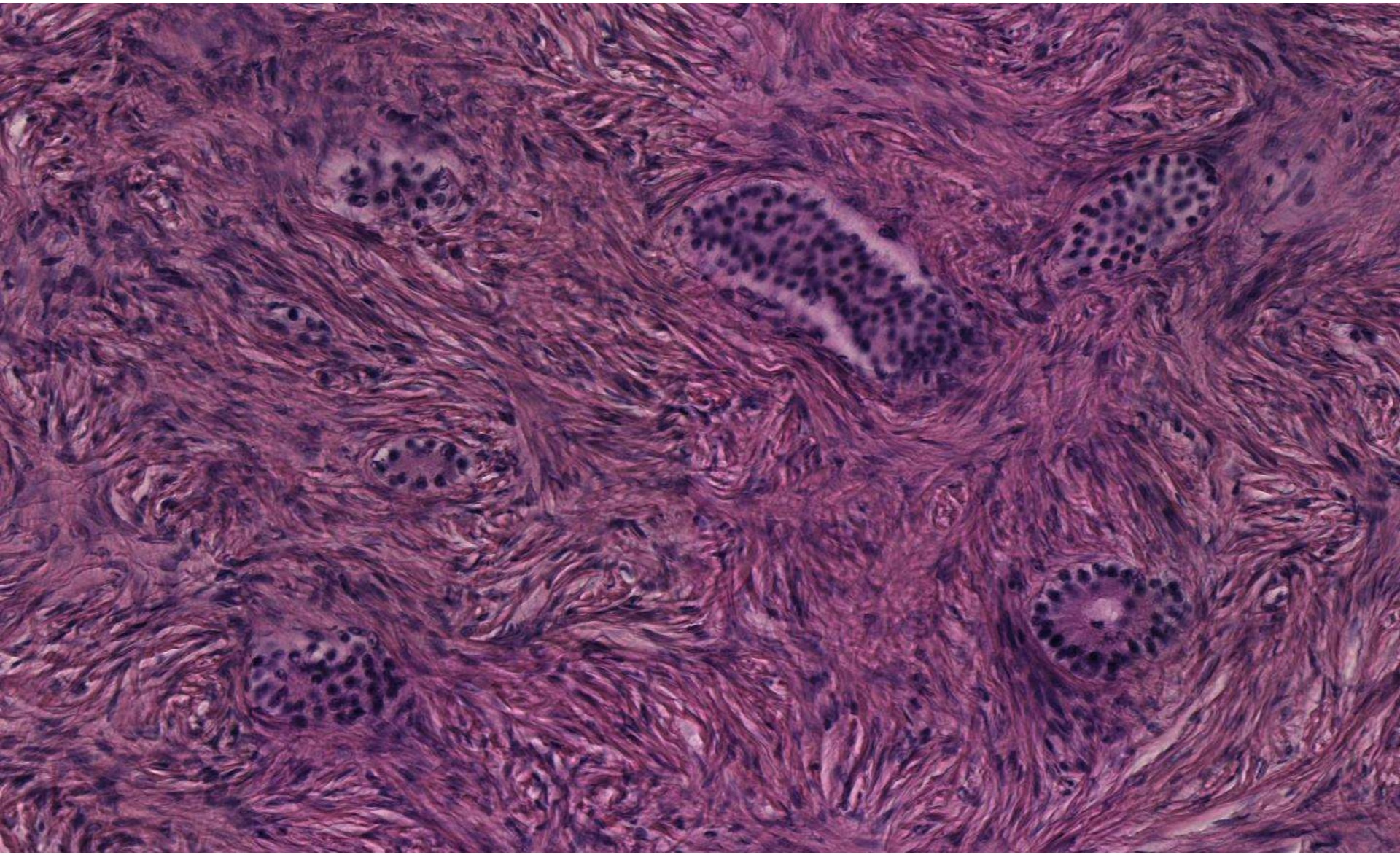


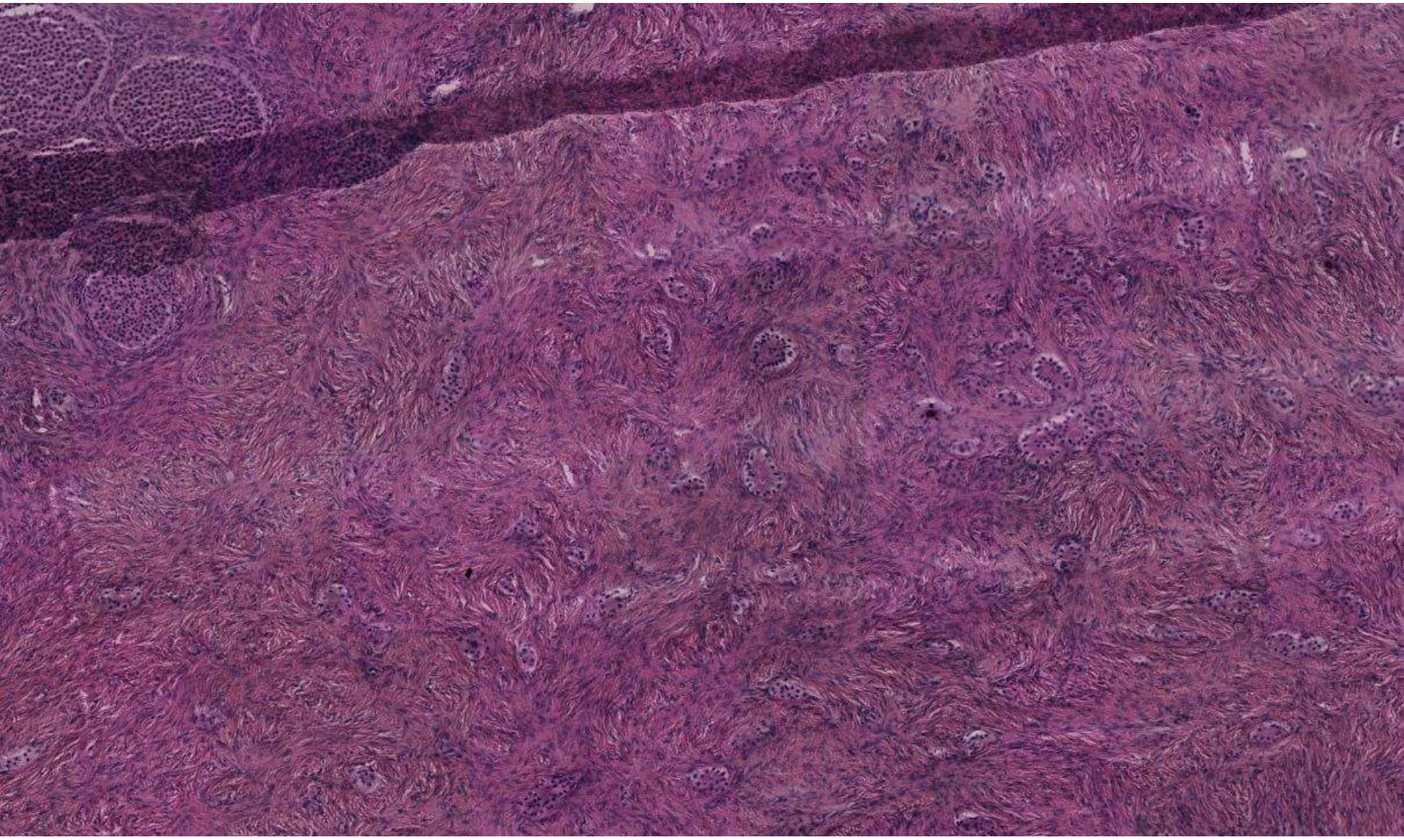


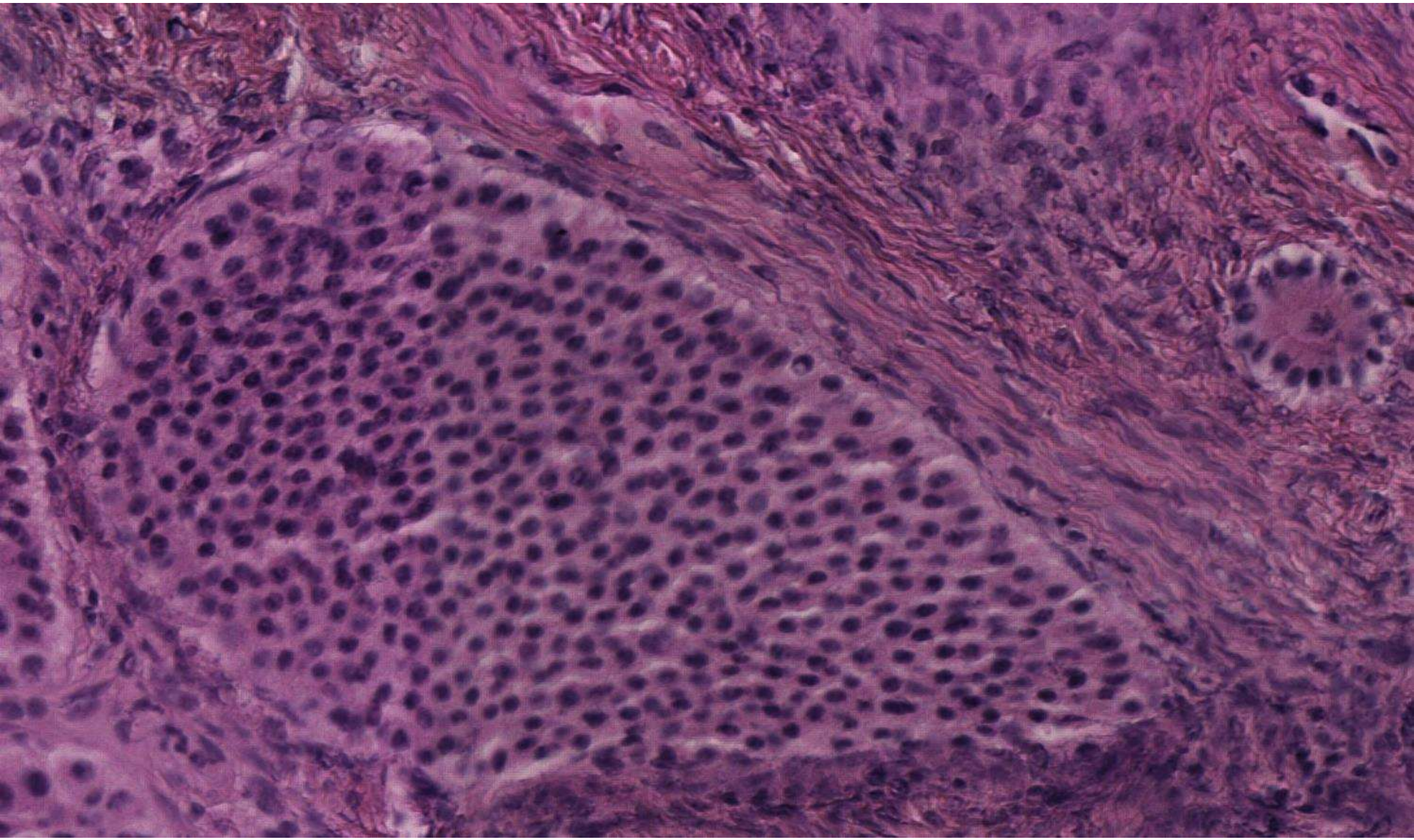


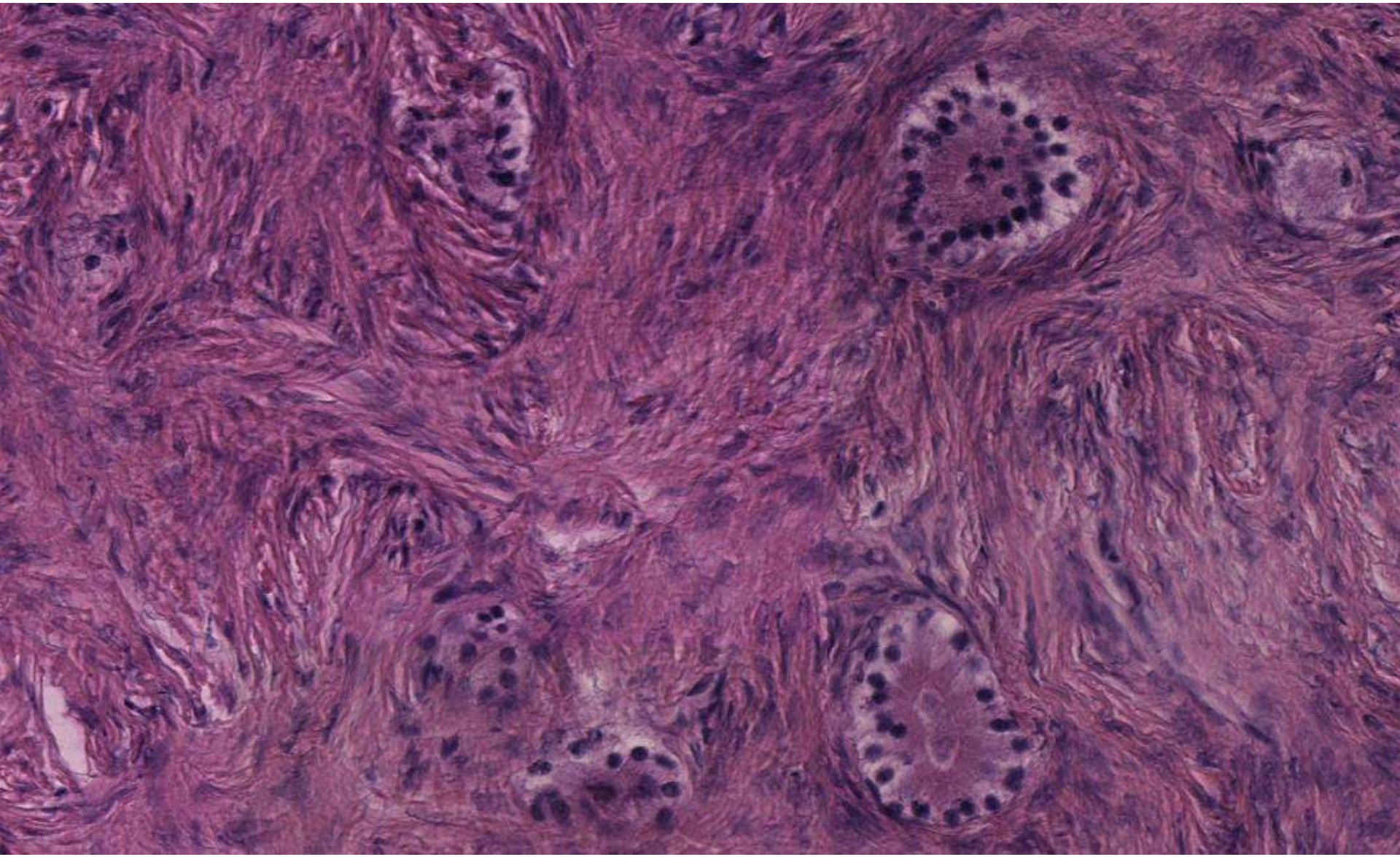


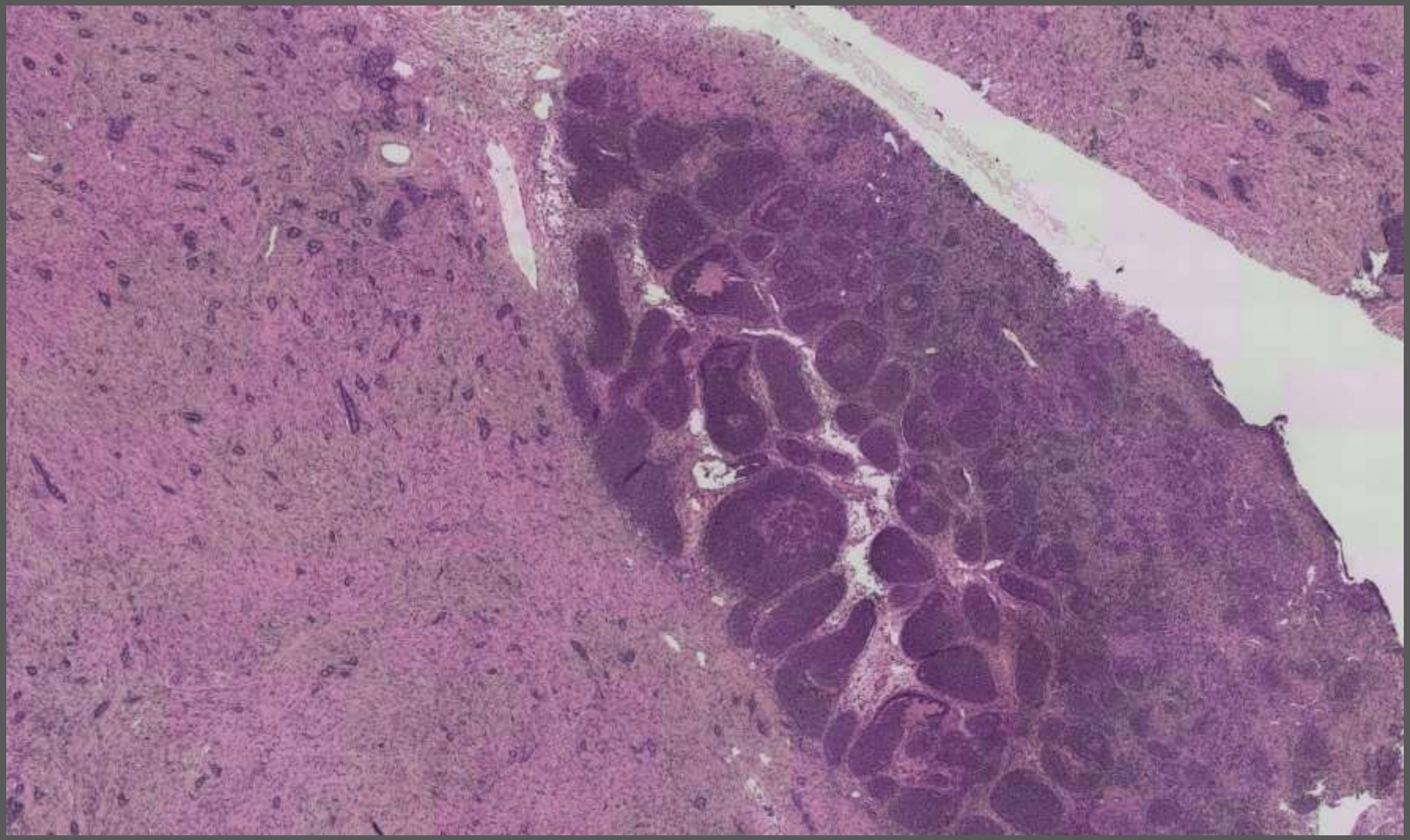


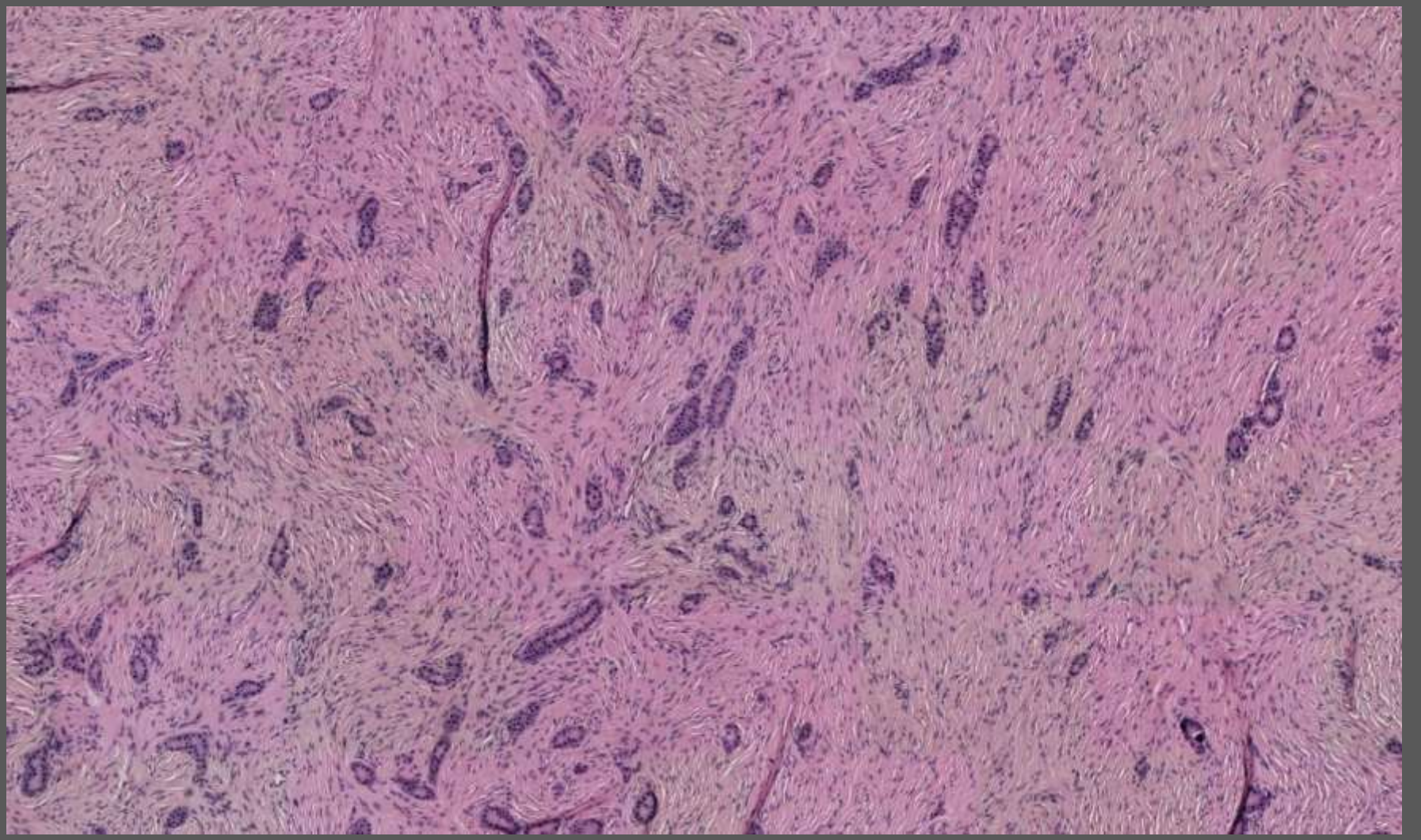


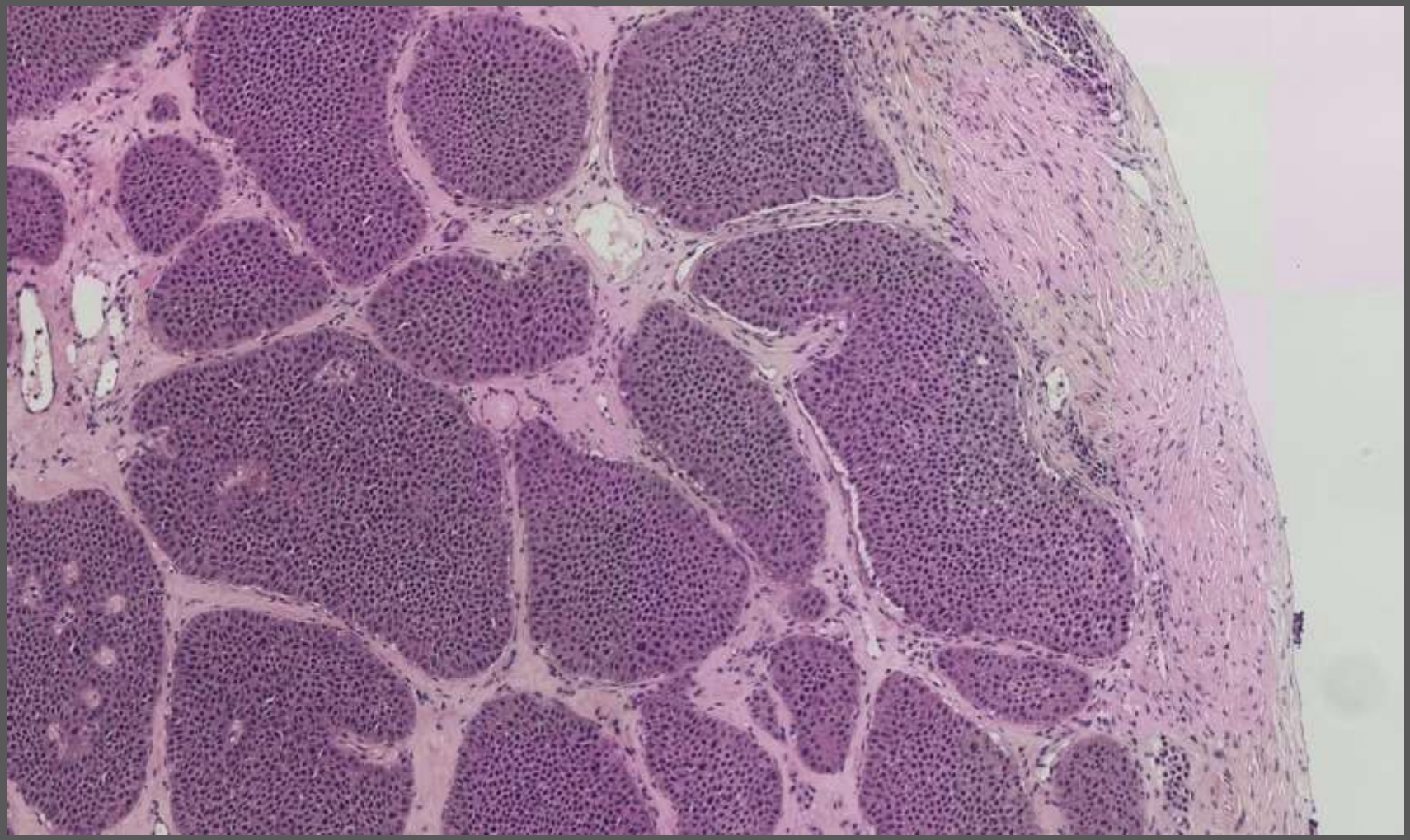


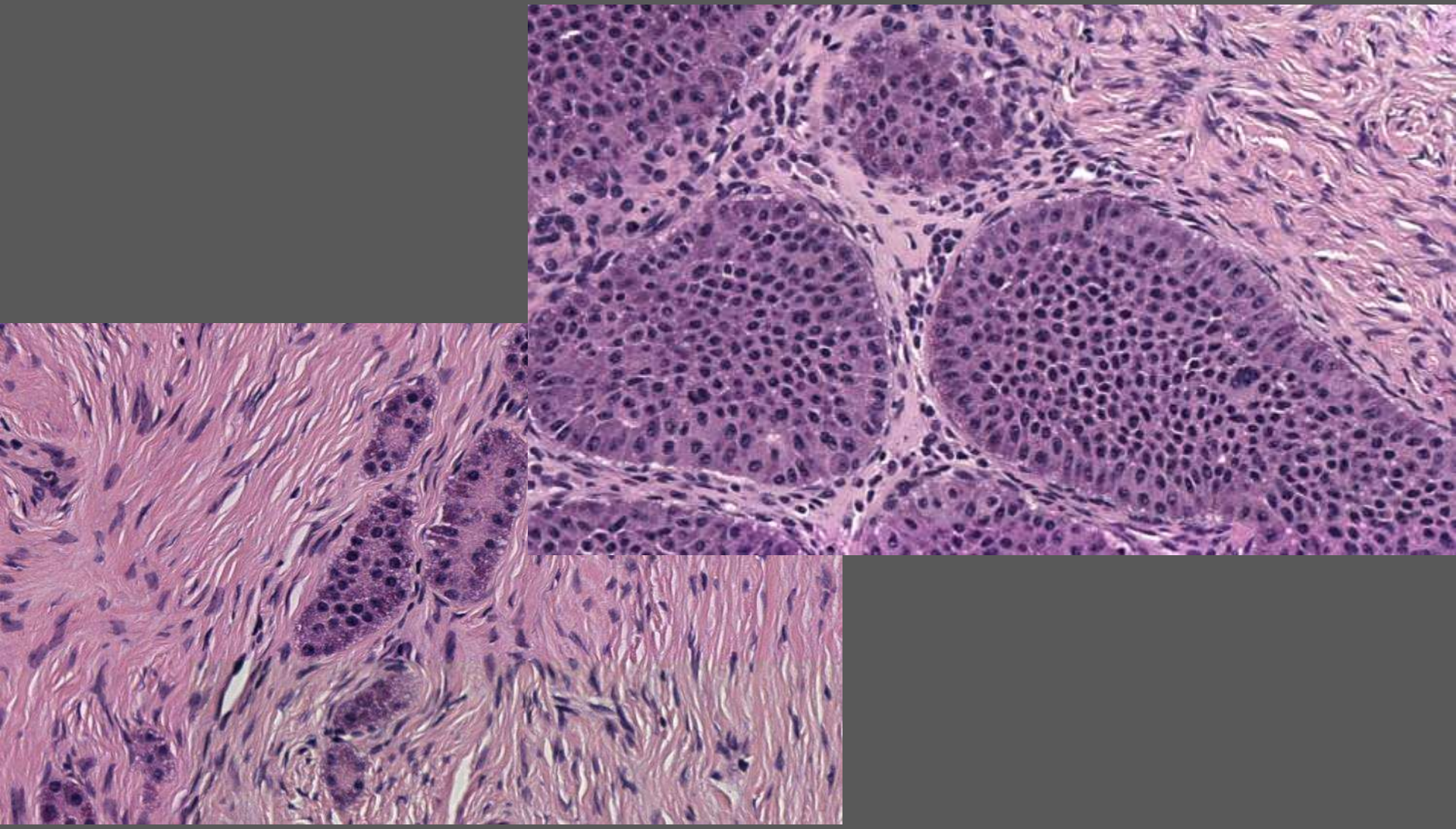












Differential diagnosis

- Primary ovary:
  - Granulosa cell tumor
  - Brenner tumor
  - Primary carcinoid tumor
  - Endometroid adenocarcinoma
- Metastatic carcinoma

Pertinent clinical history and gross

- Age – 66
- Bilateral ovary masses
- Yellow—tan tumor lesion
- Size: 5cm and 4cm
- Fallopian tubes: surface nodules
- Peritoneal lesions
- History of small intestine neuroendocrine tumor (2010)

Ovary metastases

- 15% of ovarian malignancies
- Approach to frozen: Primary tumor site (Mullerian) vs. Mets
- This will drive surgical staging and prognosis

Metastatic NET

- Originate primarily from distal terminal ileum
- Other less common sites: Cecum, appendix , jejunum, & pancreas
- Typically have synchronous metastasis
- Poor prognosis

Primary ovarian NET

- Pure carcinoid is rare (<0.1%)
- Can be a/w teratoma (struma carcinoid or mucinous tumor)
- Most useful for distinction:
  - Unilateral, gross (lacks multinodular appearance), size (typically smaller)
- IHC (CDX-2 etc.) has controversial utility

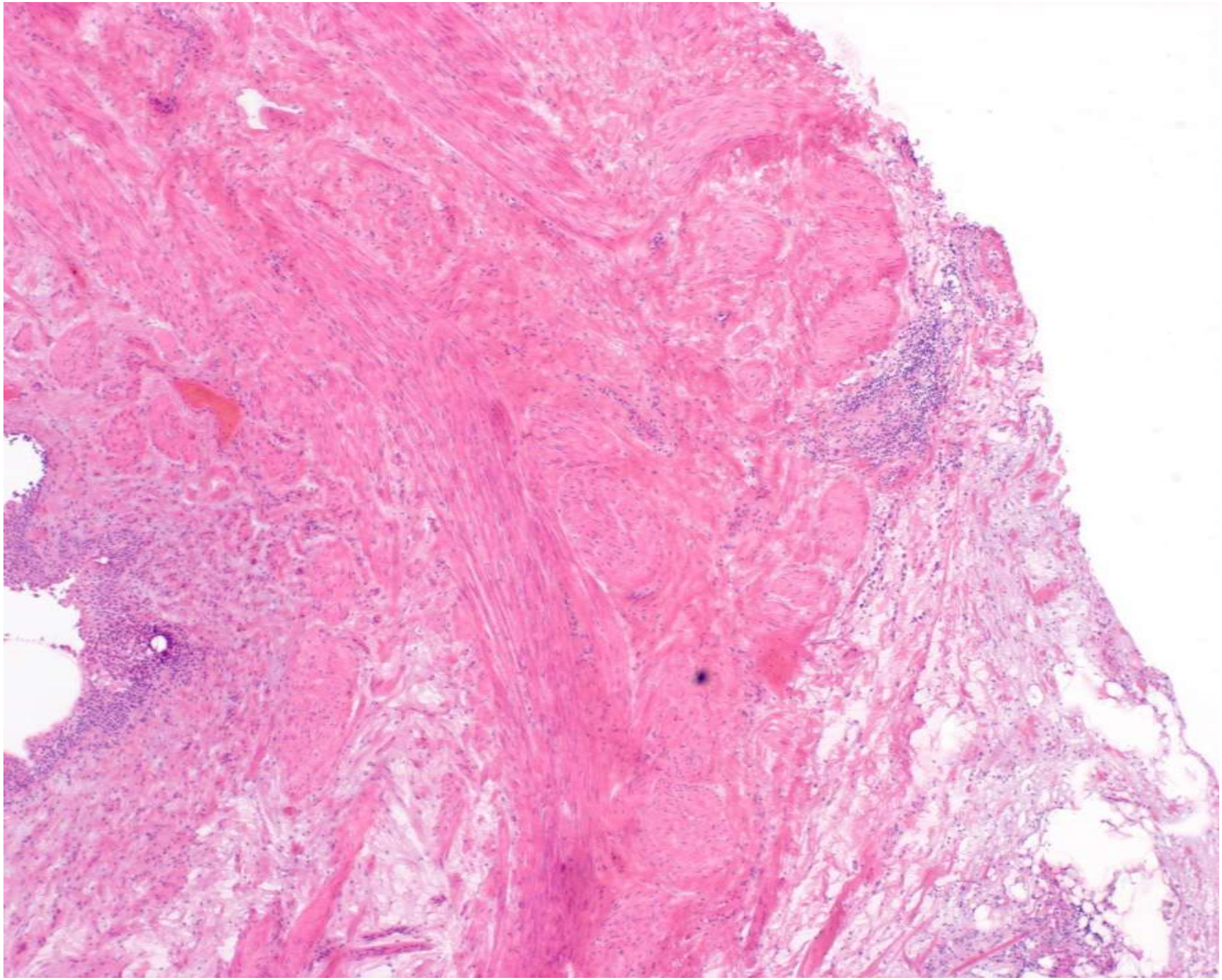
References

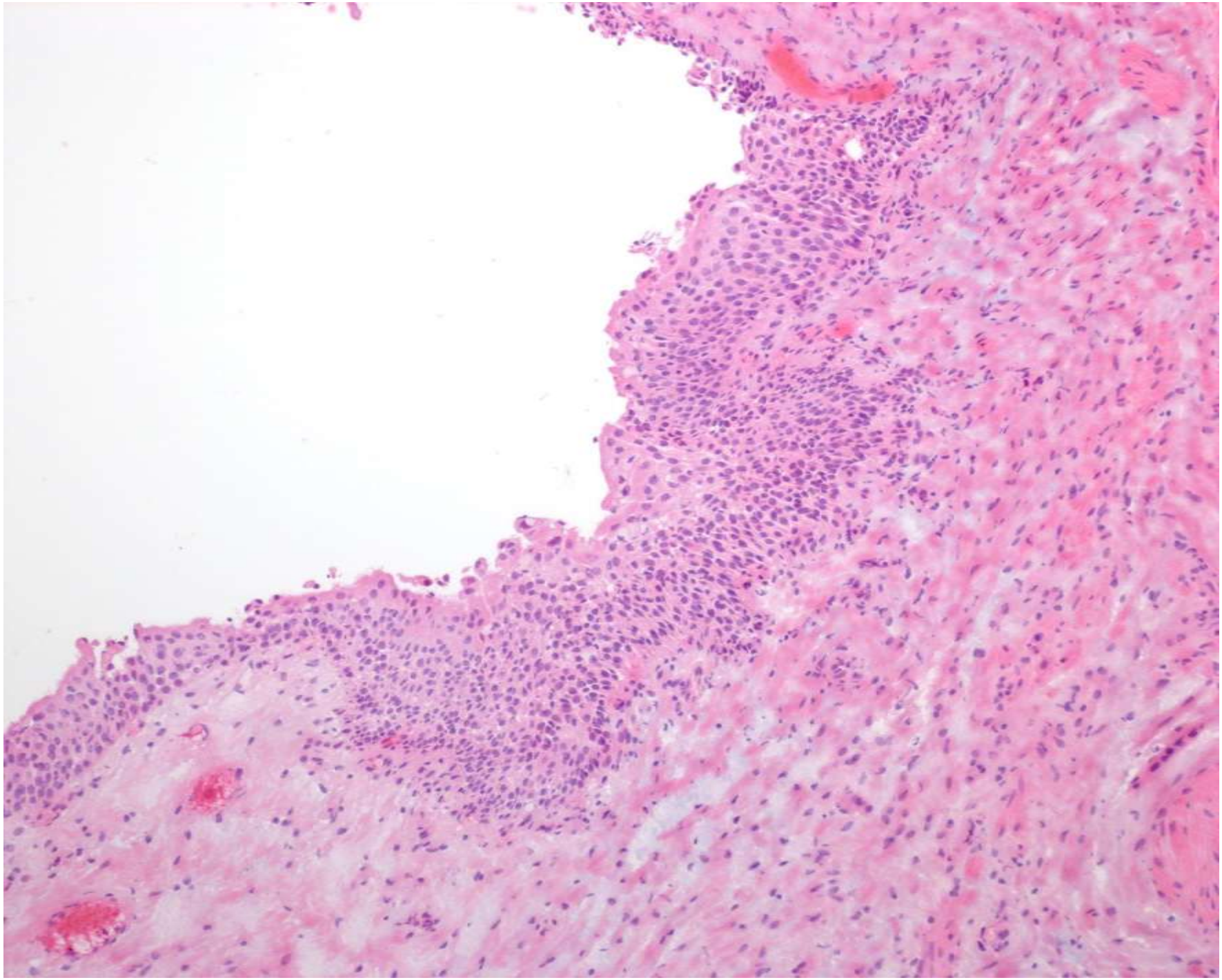
- 1. Rabban, J., et al. "Primary ovarian carcinoid tumors may express CDX-2: a potential pitfall in distinction from metastatic intestinal carcinoid tumors involving the ovary." *International journal of gynecological pathology* 28.1 (2009): 41-48.
- 2. Desouki, M., et al. "CDX2 may be a useful marker to distinguish primary ovarian carcinoid from gastrointestinal metastatic carcinoids to the ovary." *Human pathology* 44.11 (2013): 2536-2541.
- 3. Strosberg, J., et al. "Metastatic carcinoid tumor to the ovary: a clinicopathologic analysis of seventeen cases." *Gynecologic oncology* 106.1 (2007): 65-68.
- 4. Robboy, Stanley J., Robert E. Scully, and Henry J. Norris. "Carcinoid metastatic to the ovary. A clinicopathologic analysis of 35 cases." *Cancer* 33.3 (1974): 798-811.

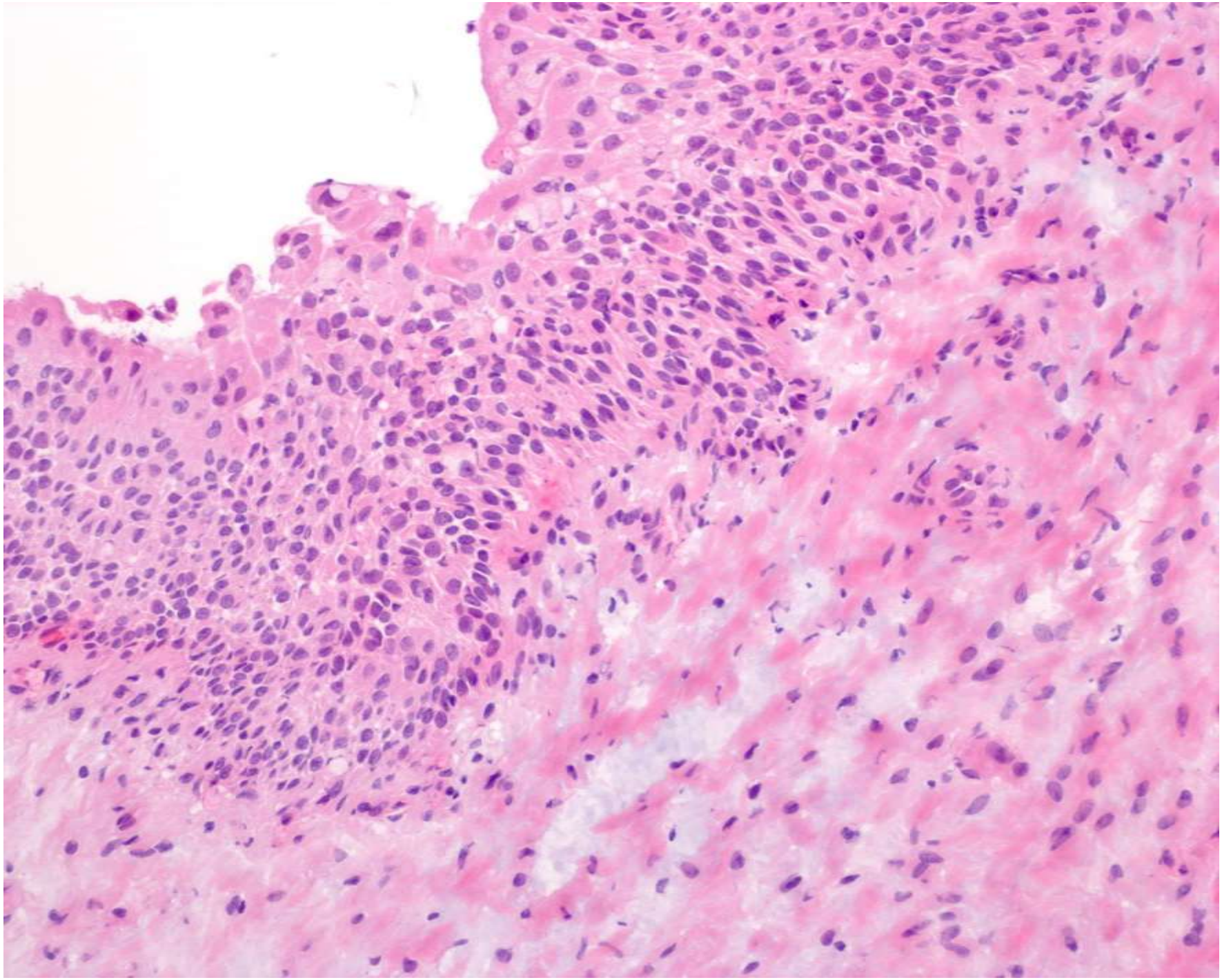
20-0404

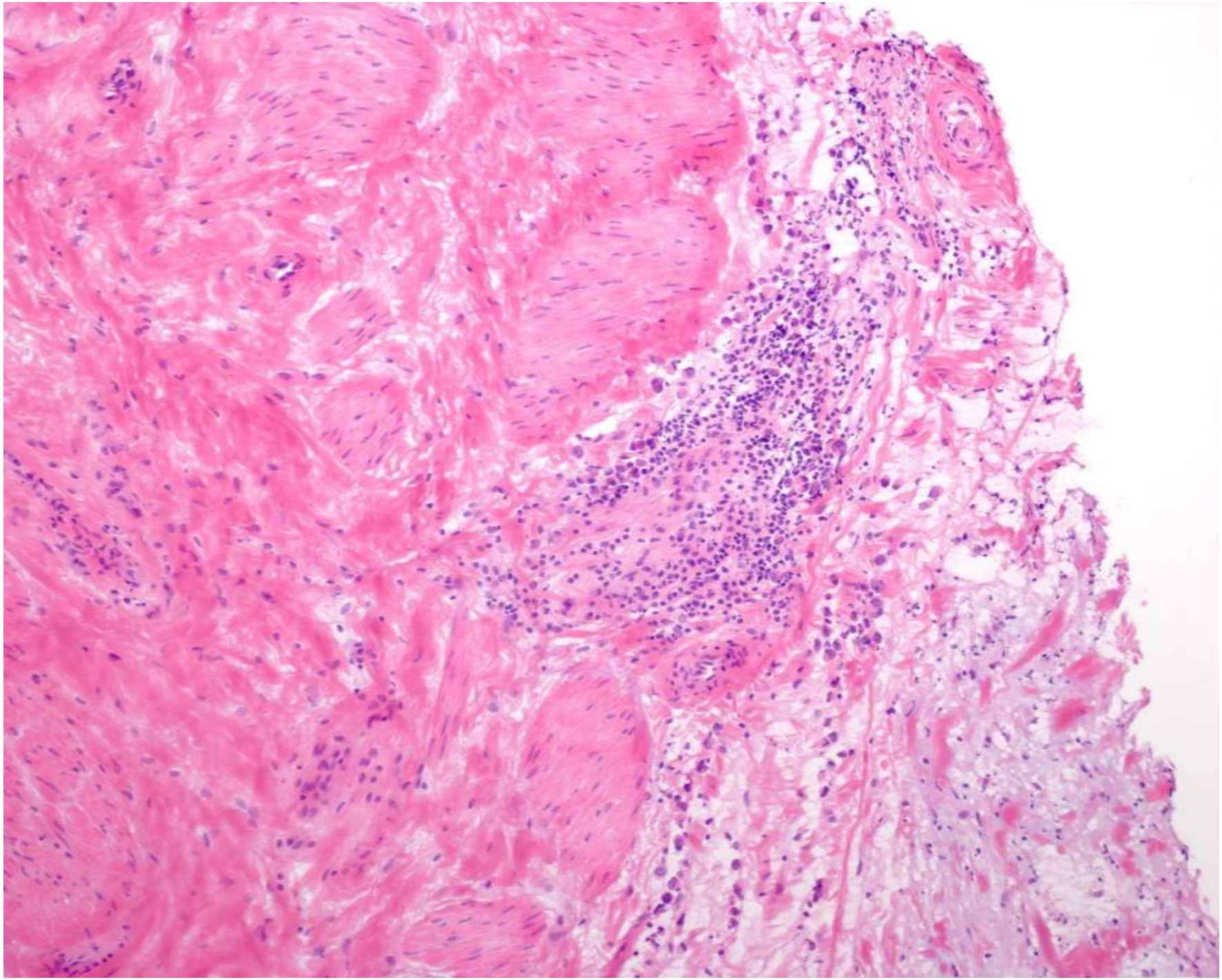
Ruth Zhang/Nicholas Ladwig/Emily Chan; UCSF

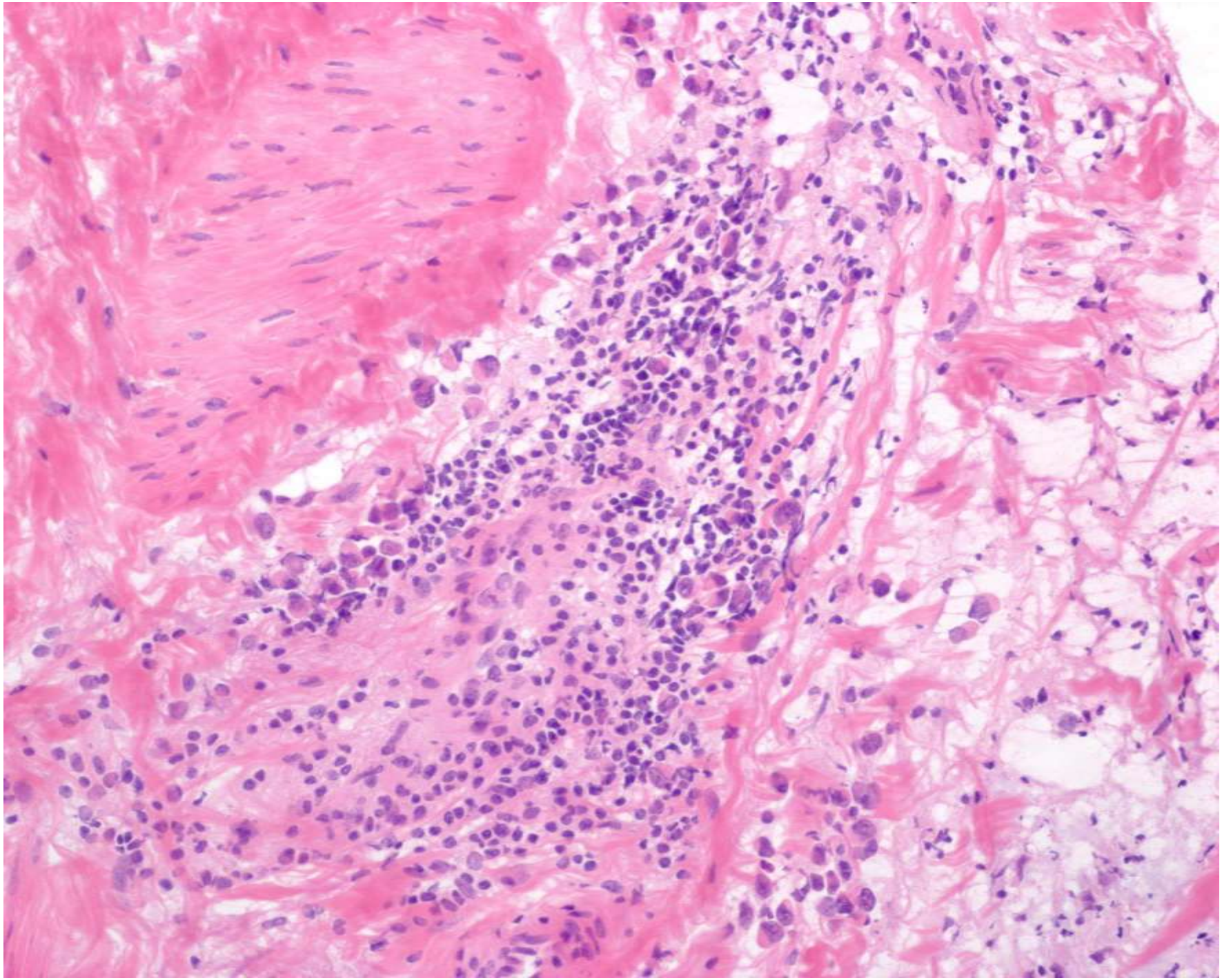
60-year-old M with bladder cancer. Ureter margin submitted for frozen section at the time of radical cystectomy.

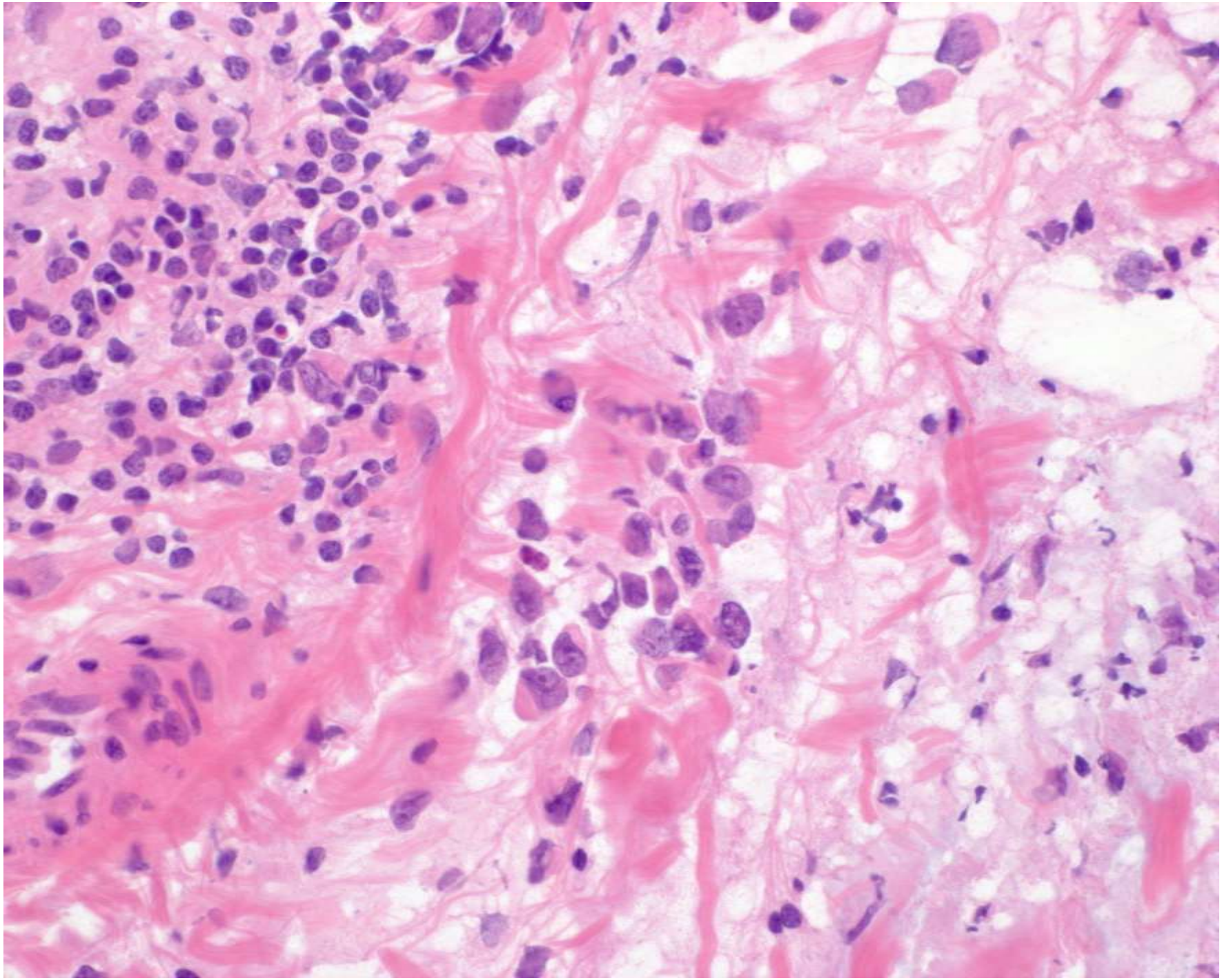


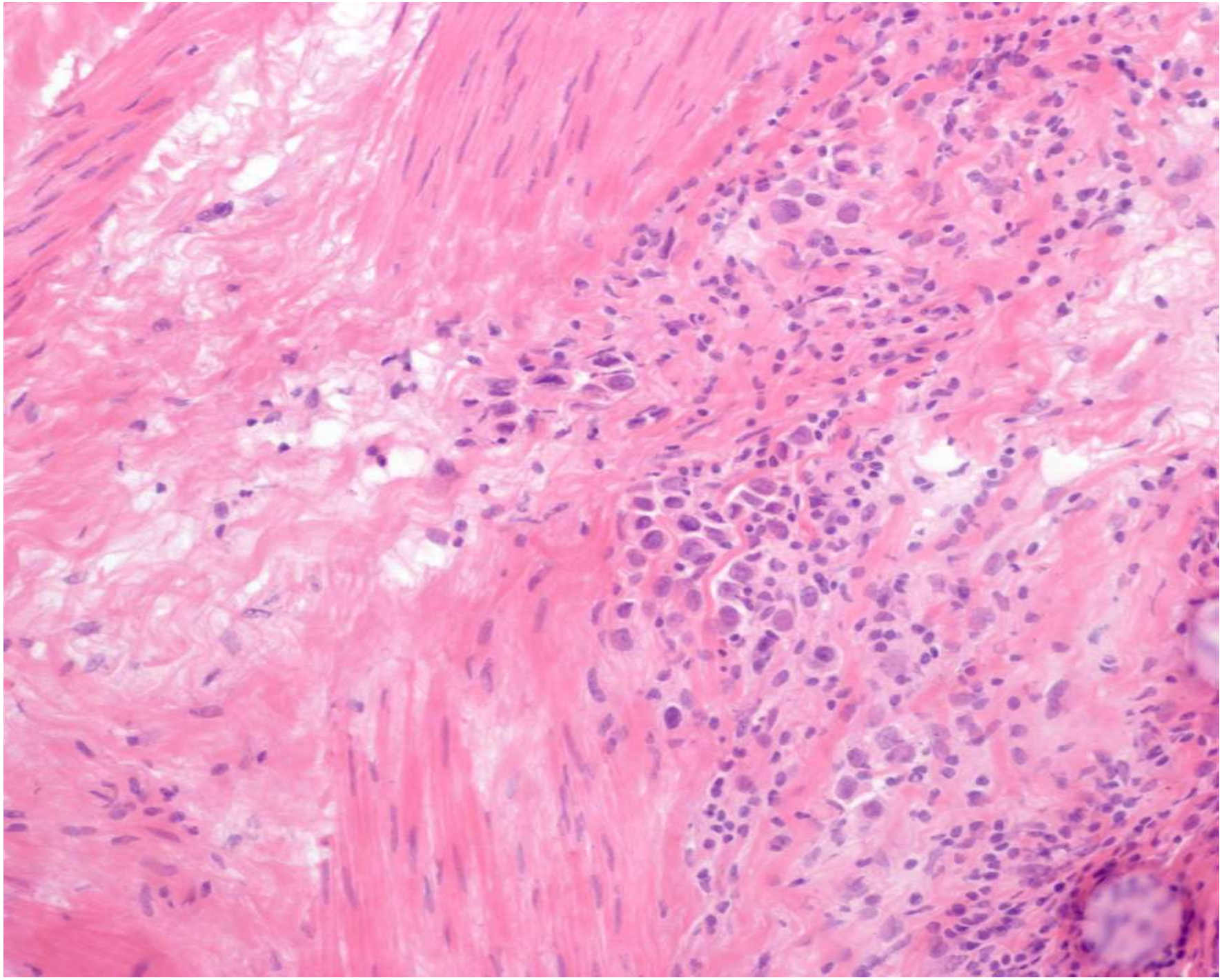






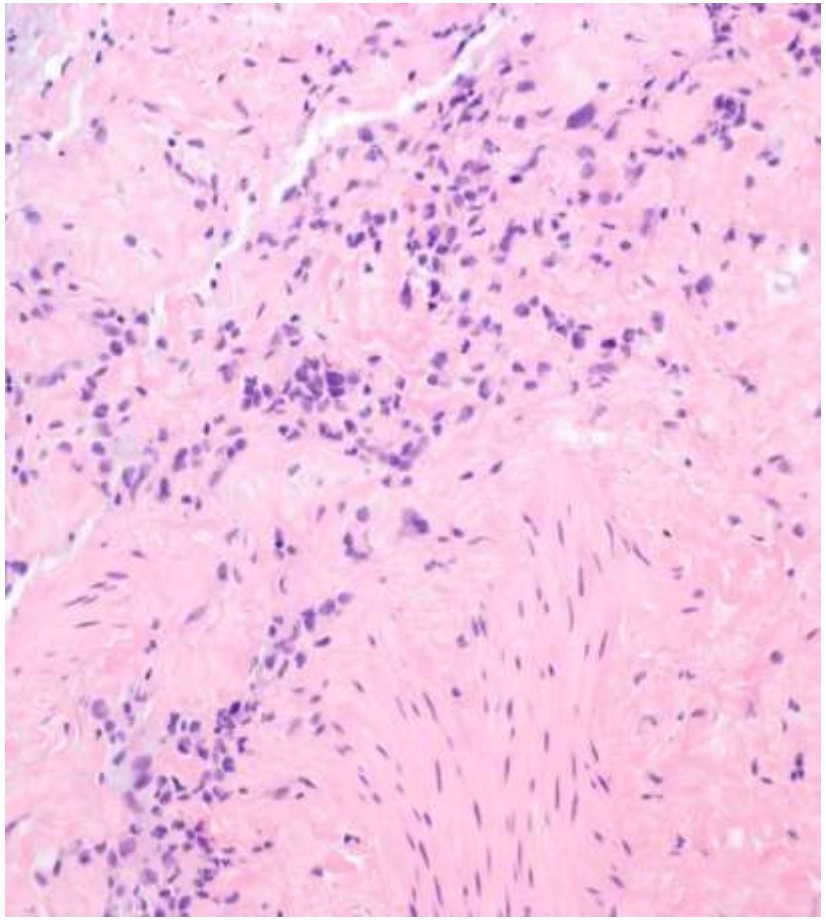


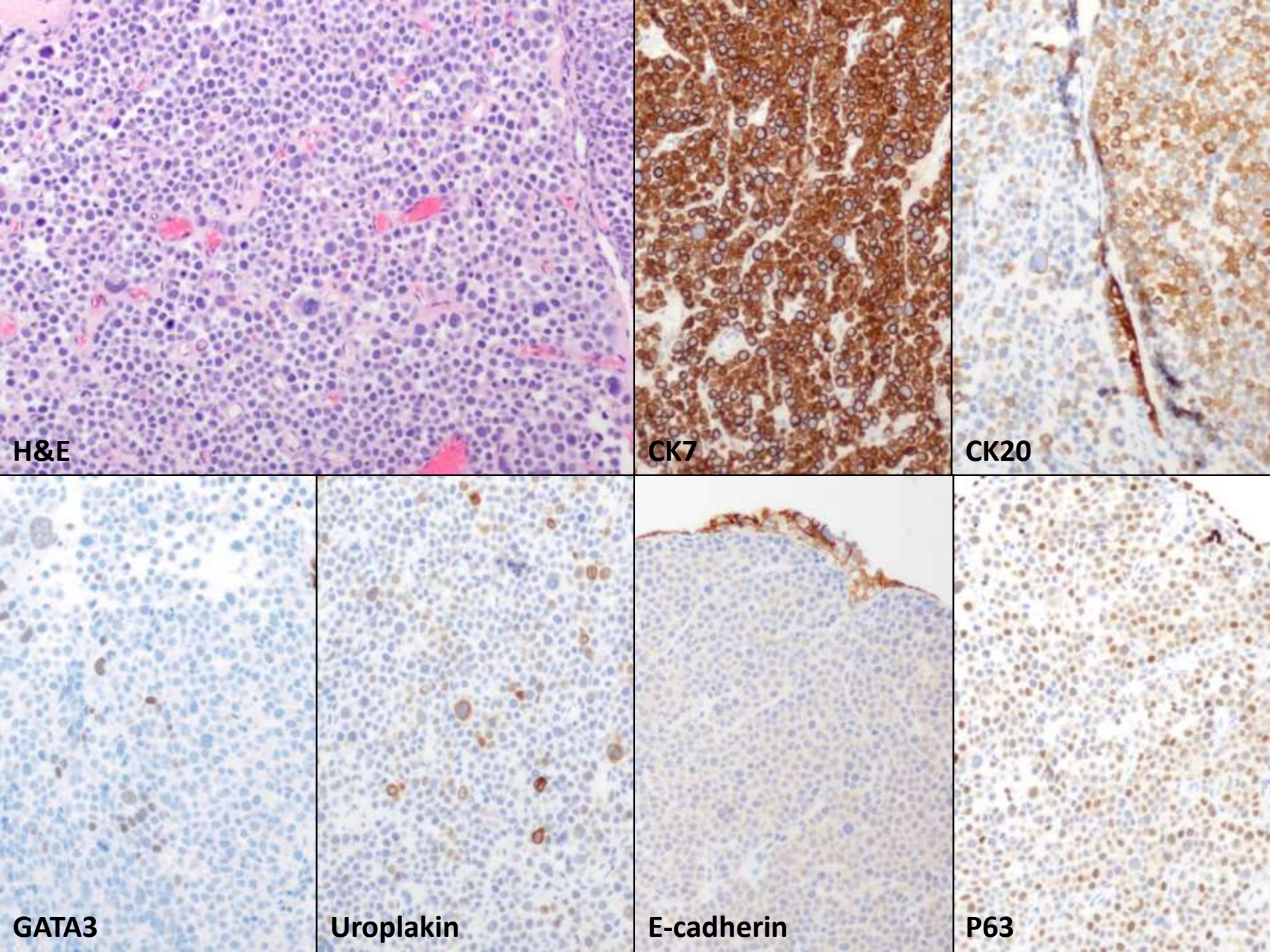




Differential diagnosis?

- Chronic inflammation
- Plasma cell neoplasm or lymphoma
- Variant of invasive urothelial carcinoma
- Melanoma
- Metastatic invasive lobular carcinoma




H&E
CK7
CK20
GATA3
Uroplakin
E-cadherin
P63

Plasmacytoid urothelial carcinoma

- Discohesive cells with eccentric nuclei and abundant eosinophilic cytoplasm
  - May show rhabdoid or signet ring cell-like morphology
- Discontinuous, infiltrative growth
- Advanced stage at presentation

› J Urol. 2012 Mar;187(3):852-5. doi: 10.1016/j.juro.2011.10.145. Epub 2012 Jan 15.

Plasmacytoid Carcinoma of the Bladder: A Urothelial Carcinoma Variant With a Predilection for Intraperitoneal Spread

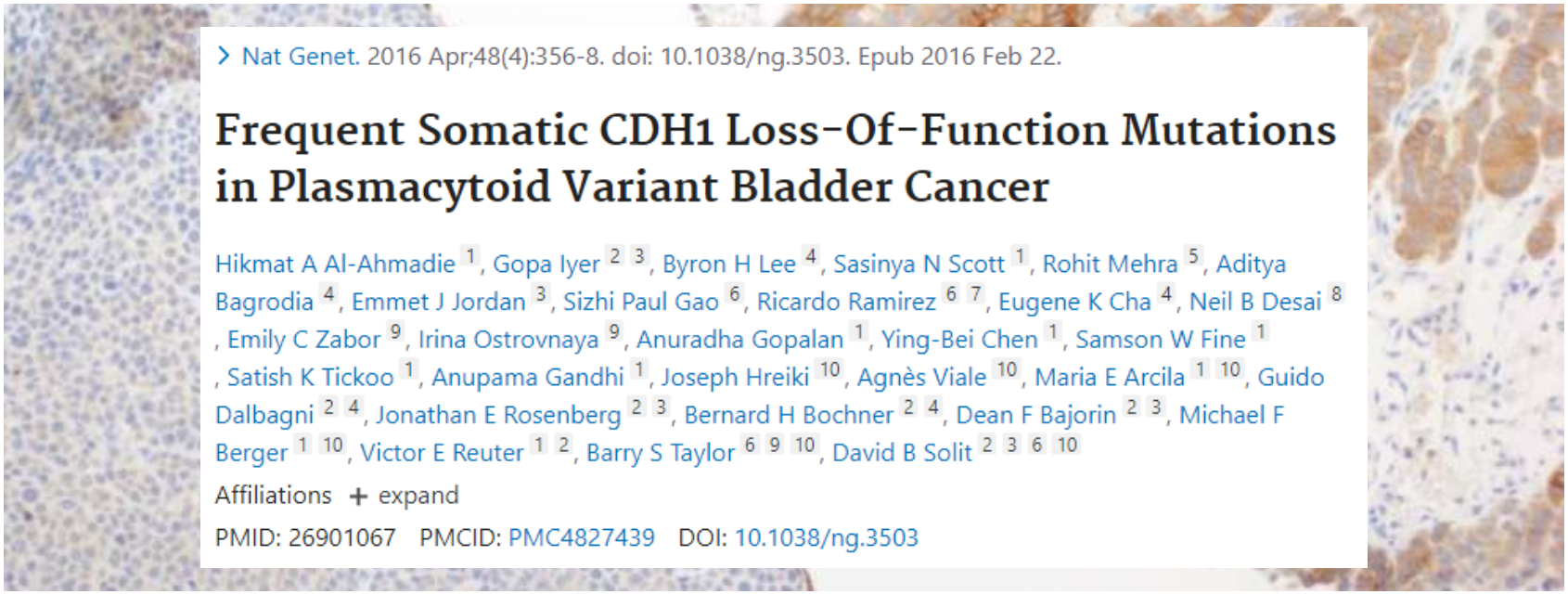
Roberto Rafael Ricardo-Gonzalez [1], Michael Nguyen, Neriman Gokden, Ankur R Sangoi, Joseph C Presti Jr, Jesse K McKenney

Affiliations + expand

PMID: 22245324 DOI: 10.1016/j.juro.2011.10.145

Plasmacytoid urothelial carcinoma

- Loss of E-cadherin (CDH1)
  - Cell adhesion molecule
  - Sequencing: CDH1 truncating somatic mutation (28/31) or promoter hypermethylation (4/5)
  - IHC: loss of E-cadherin (31/31)



› Nat Genet. 2016 Apr;48(4):356-8. doi: 10.1038/ng.3503. Epub 2016 Feb 22.

Frequent Somatic CDH1 Loss-Of-Function Mutations in Plasmacytoid Variant Bladder Cancer

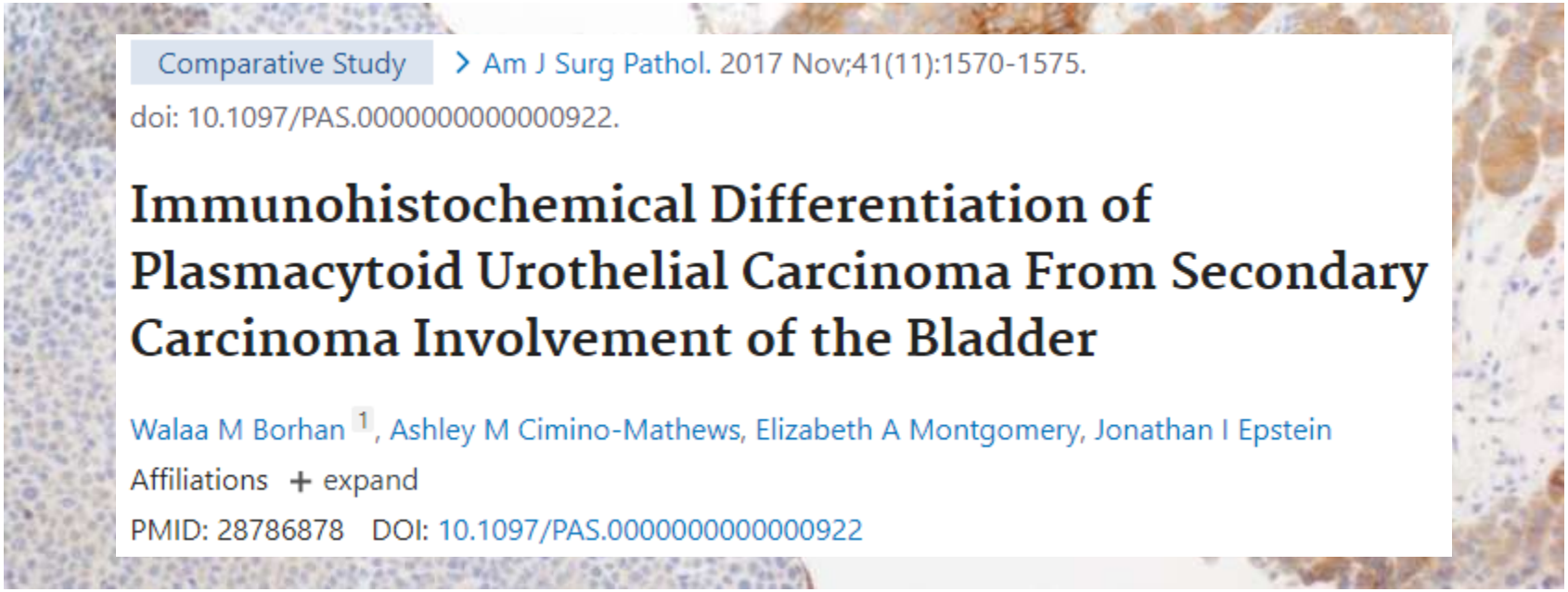
Hikmat A Al-Ahmadie [1], Gopa Iyer [2 3], Byron H Lee [4], Sasinya N Scott [1], Rohit Mehra [5], Aditya Bagrodia [4], Emmet J Jordan [3], Sizhi Paul Gao [6], Ricardo Ramirez [6 7], Eugene K Cha [4], Neil B Desai [8], Emily C Zabor [9], Irina Ostrovnaya [9], Anuradha Gopalan [1], Ying-Bei Chen [1], Samson W Fine [1], Satish K Tickoo [1], Anupama Gandhi [1], Joseph Hreiki [10], Agnès Viale [10], Maria E Arcila [1 10], Guido Dalbagni [2 4], Jonathan E Rosenberg [2 3], Bernard H Bochner [2 4], Dean F Bajorin [2 3], Michael F Berger [1 10], Victor E Reuter [1 2], Barry S Taylor [6 9 10], David B Solit [2 3 6 10]

Affiliations + expand

PMID: 26901067 PMCID: PMC4827439 DOI: 10.1038/ng.3503

Plasmacytoid urothelial carcinoma

- Loss of E-cadherin (CDH1)
  - Lost also in invasive lobular carcinoma and hereditary diffuse gastric cancer
  - Differentiating IHC: Uroplakin II, mammaglobin, ER



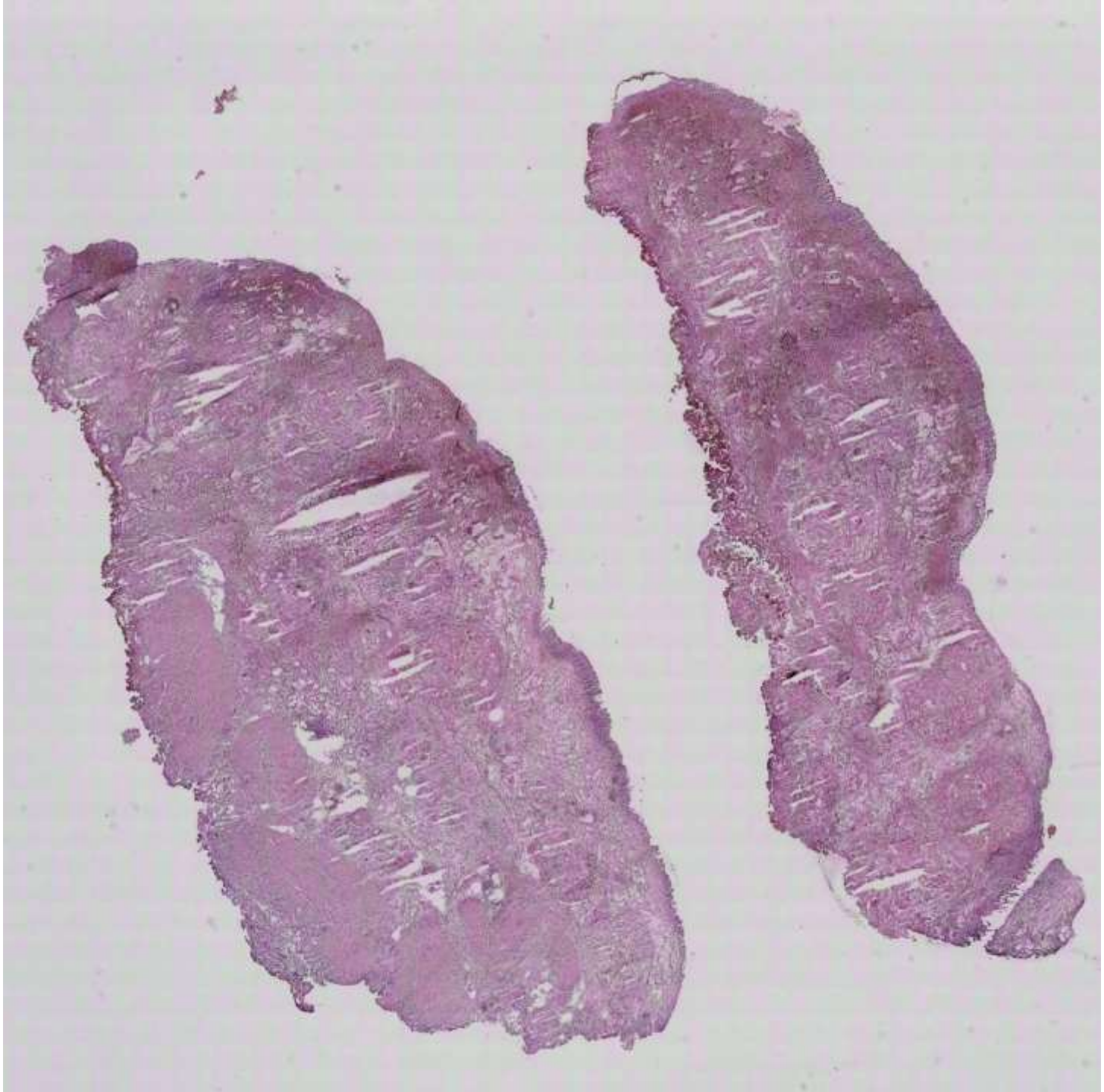
PUC at time of frozen section

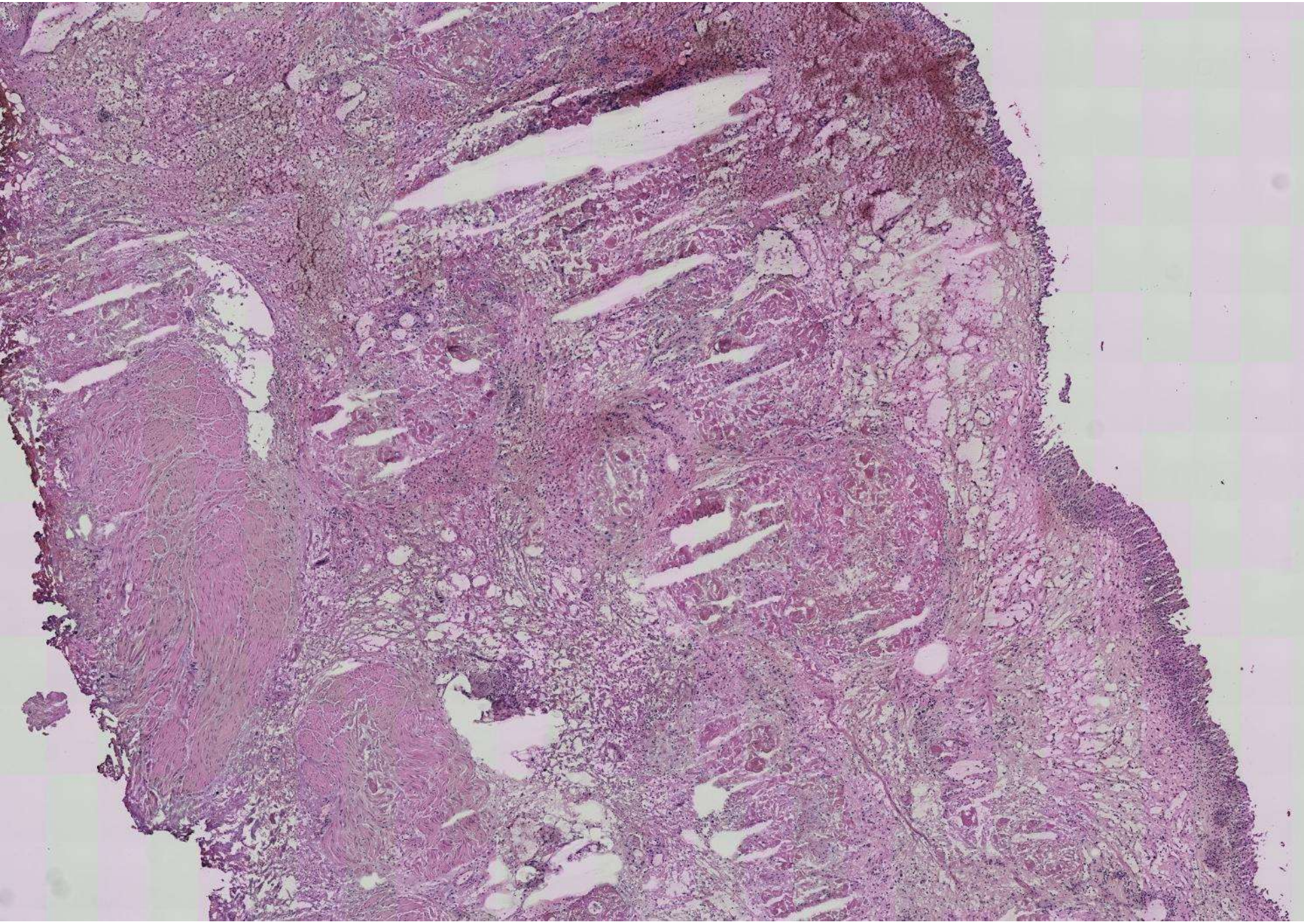
- In biopsy/TUR, important to state if a variant is present (even if muscle invasion not seen)
  - Guide surgery/oncology planning
  - Facilitate subsequent pathologic diagnosis
- Remember to look away from the mucosa!
  - Periureteral soft tissue (adventitia)
  - Serosal surfaces

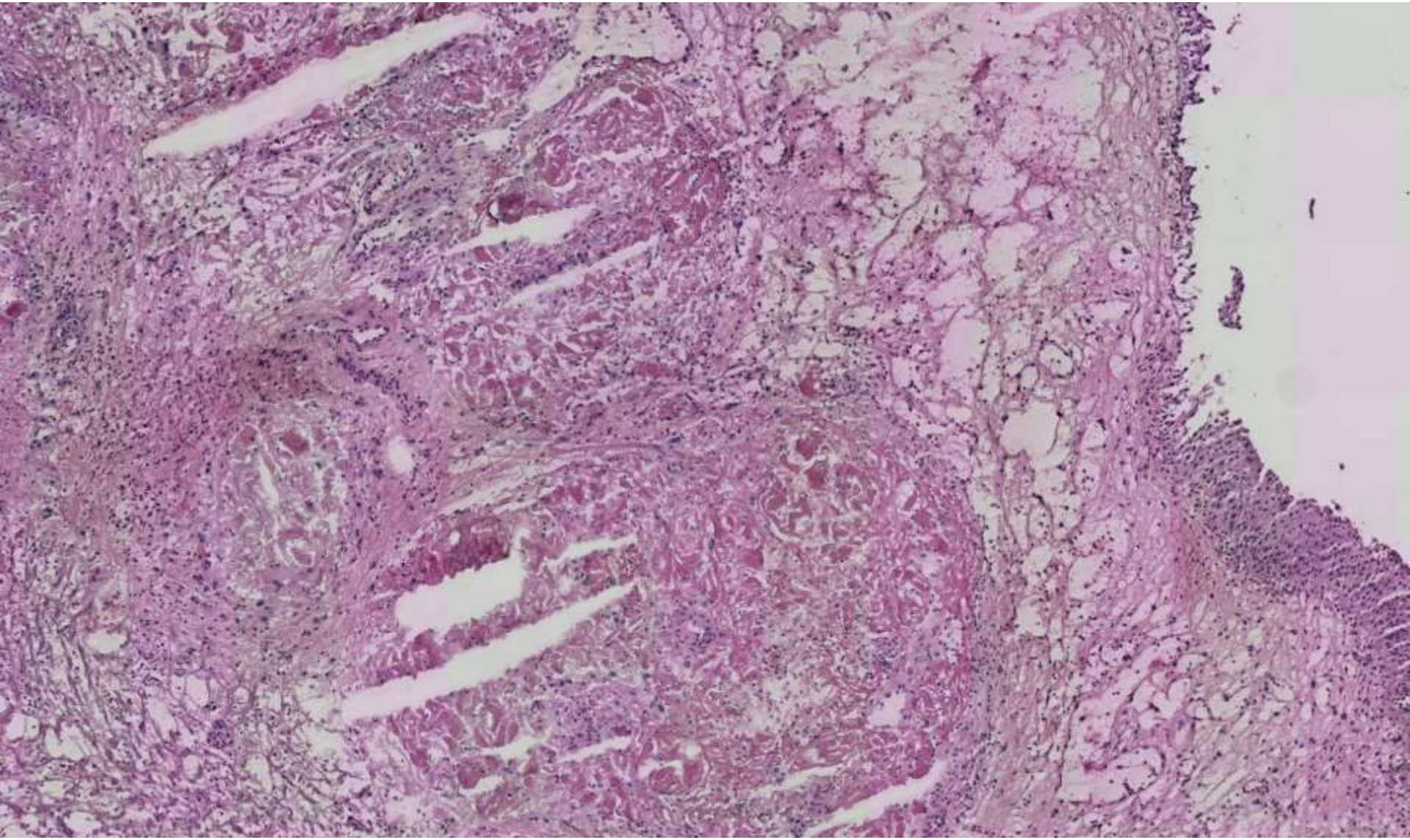
20-0405
SCANNED SLIDE AVAIL!

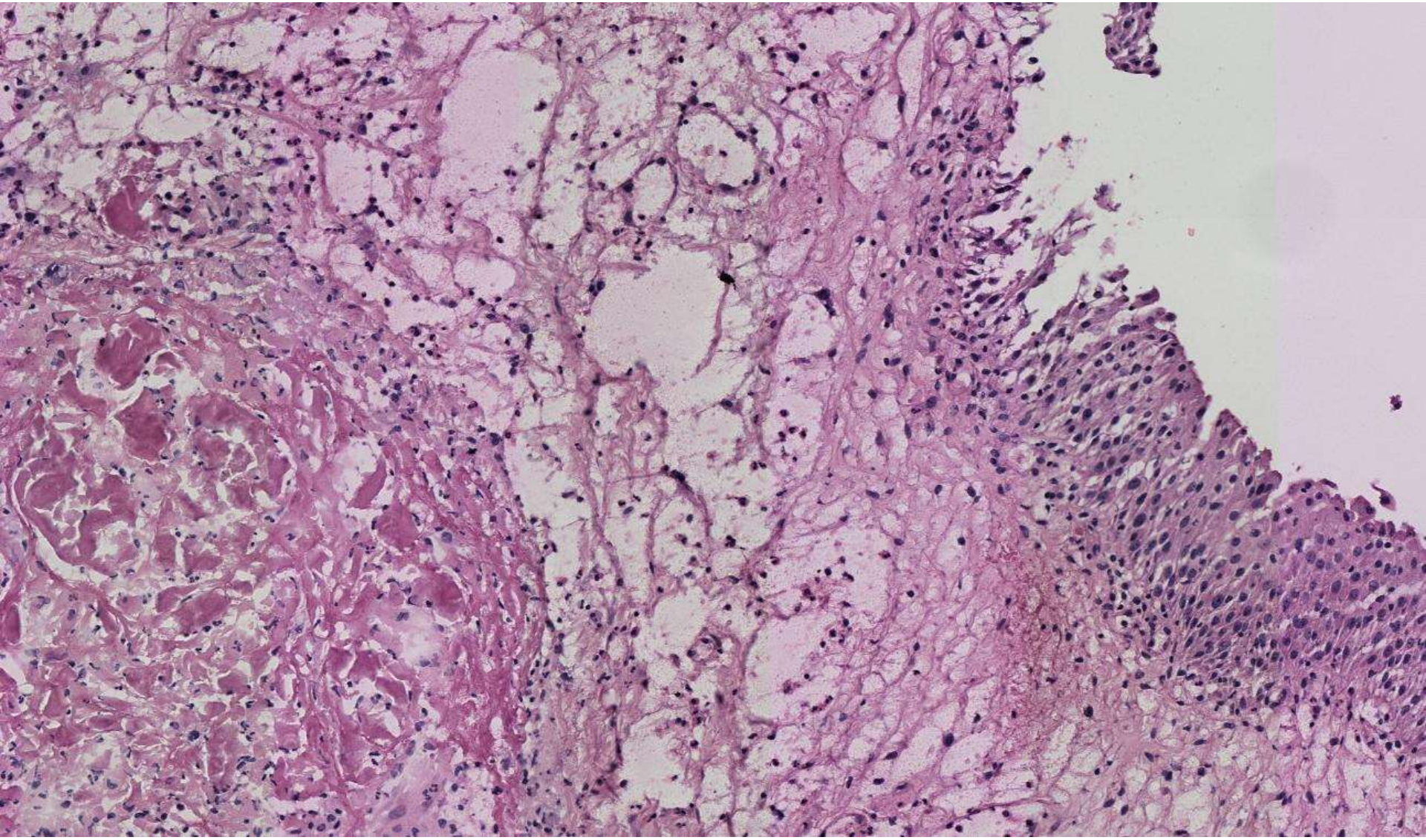
Ankur Sangoi; El Camino Hospital

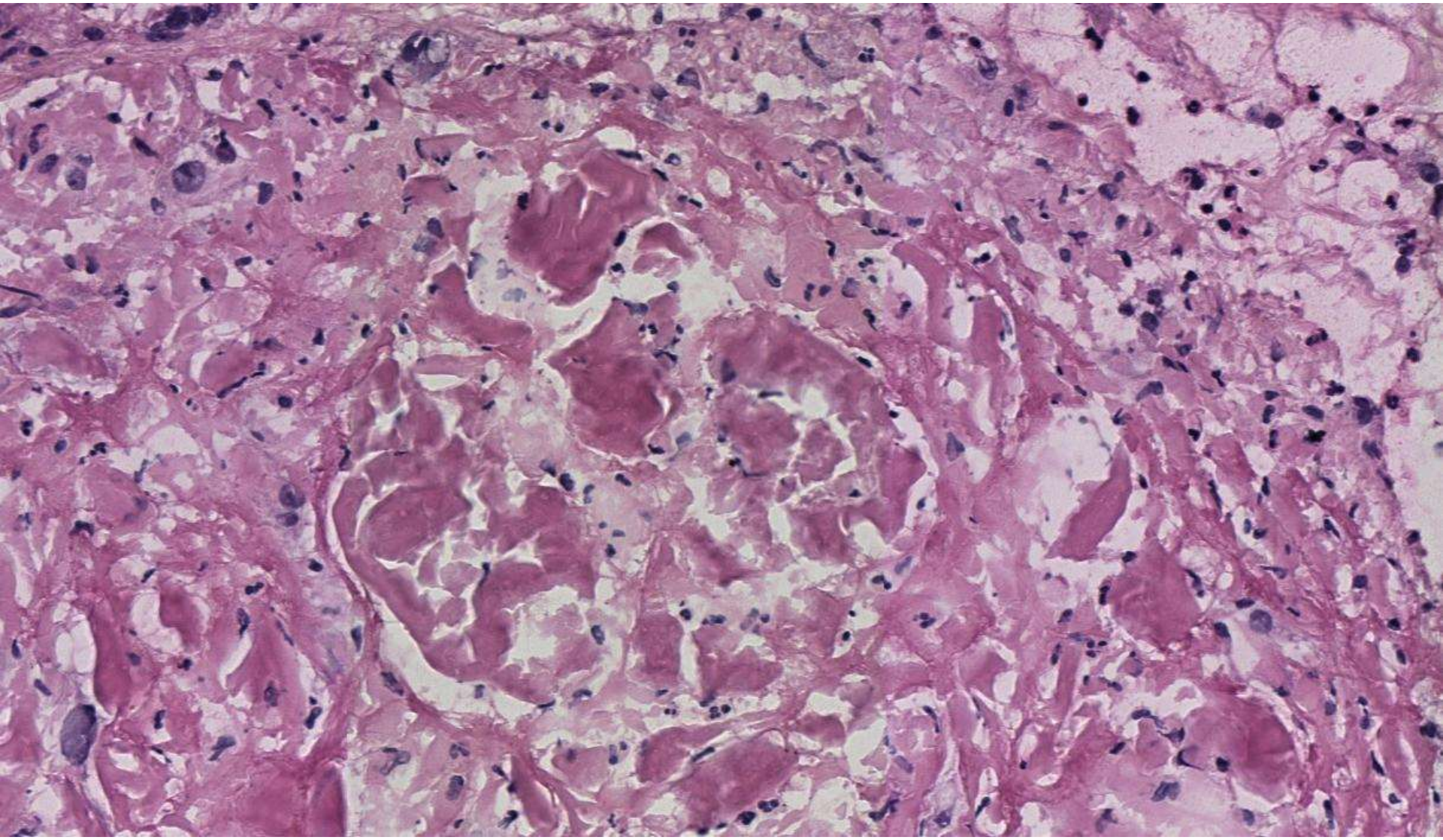
71-year-old M with bladder mass. Portion of TURBT specimen submitted for frozen section.

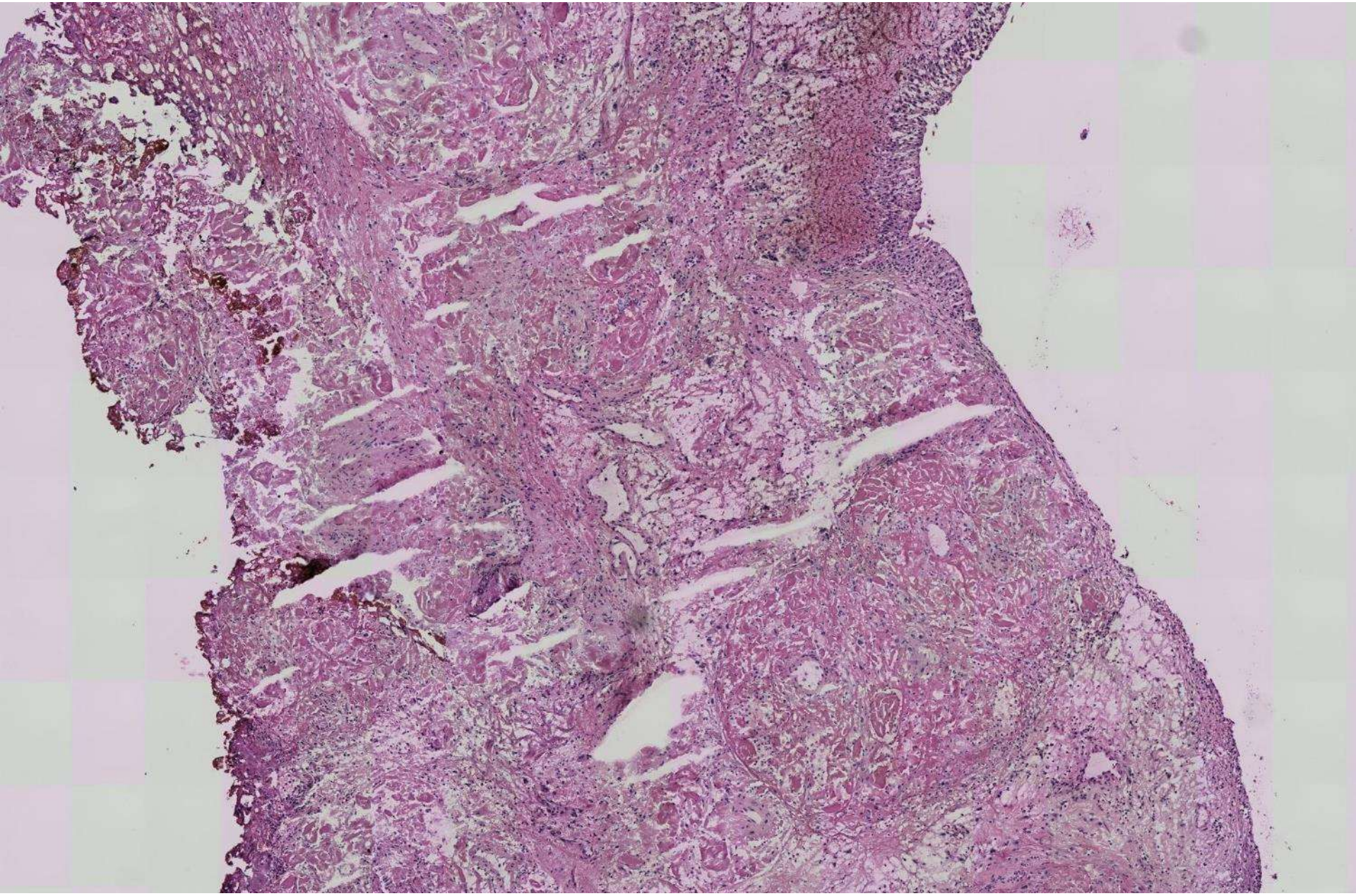


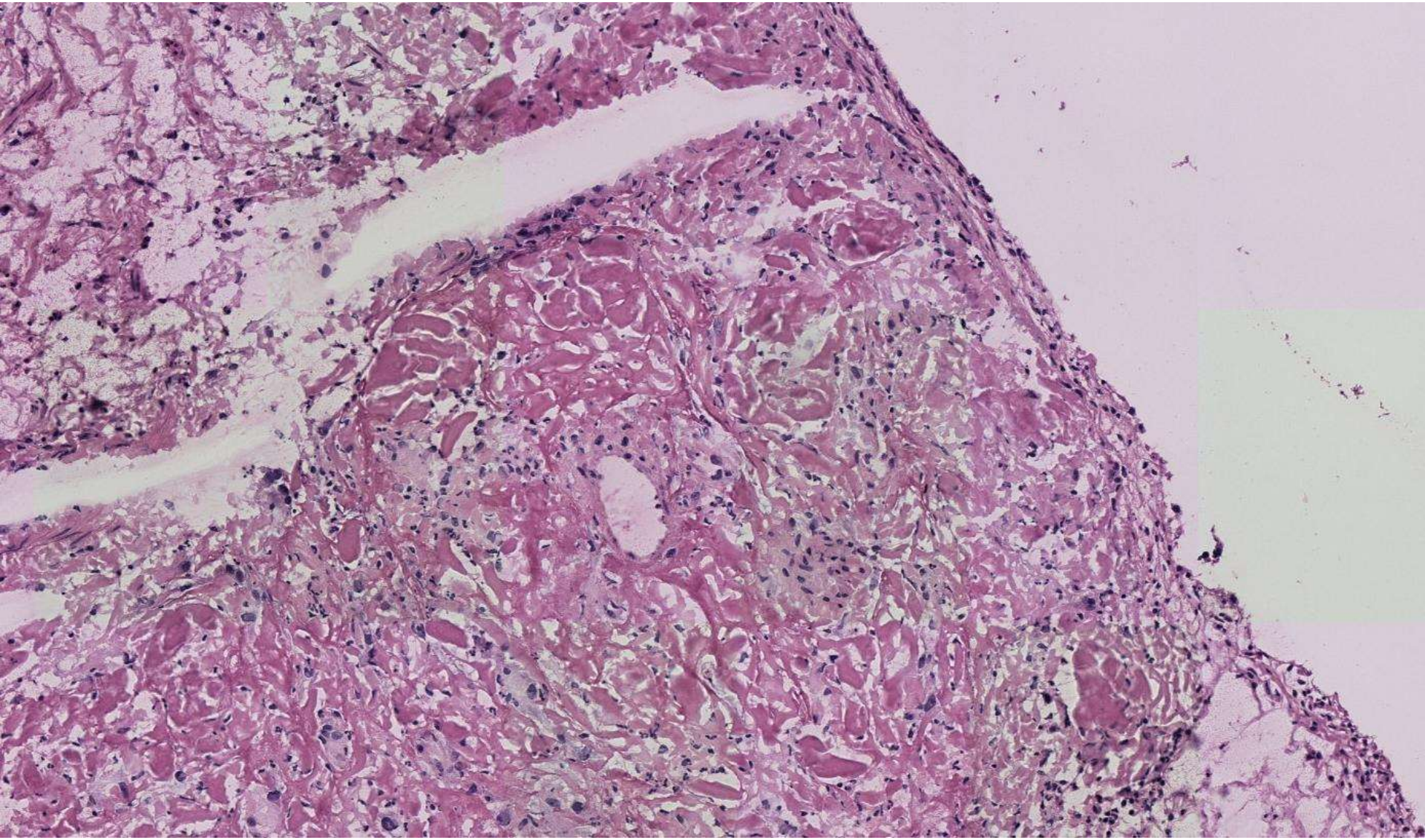


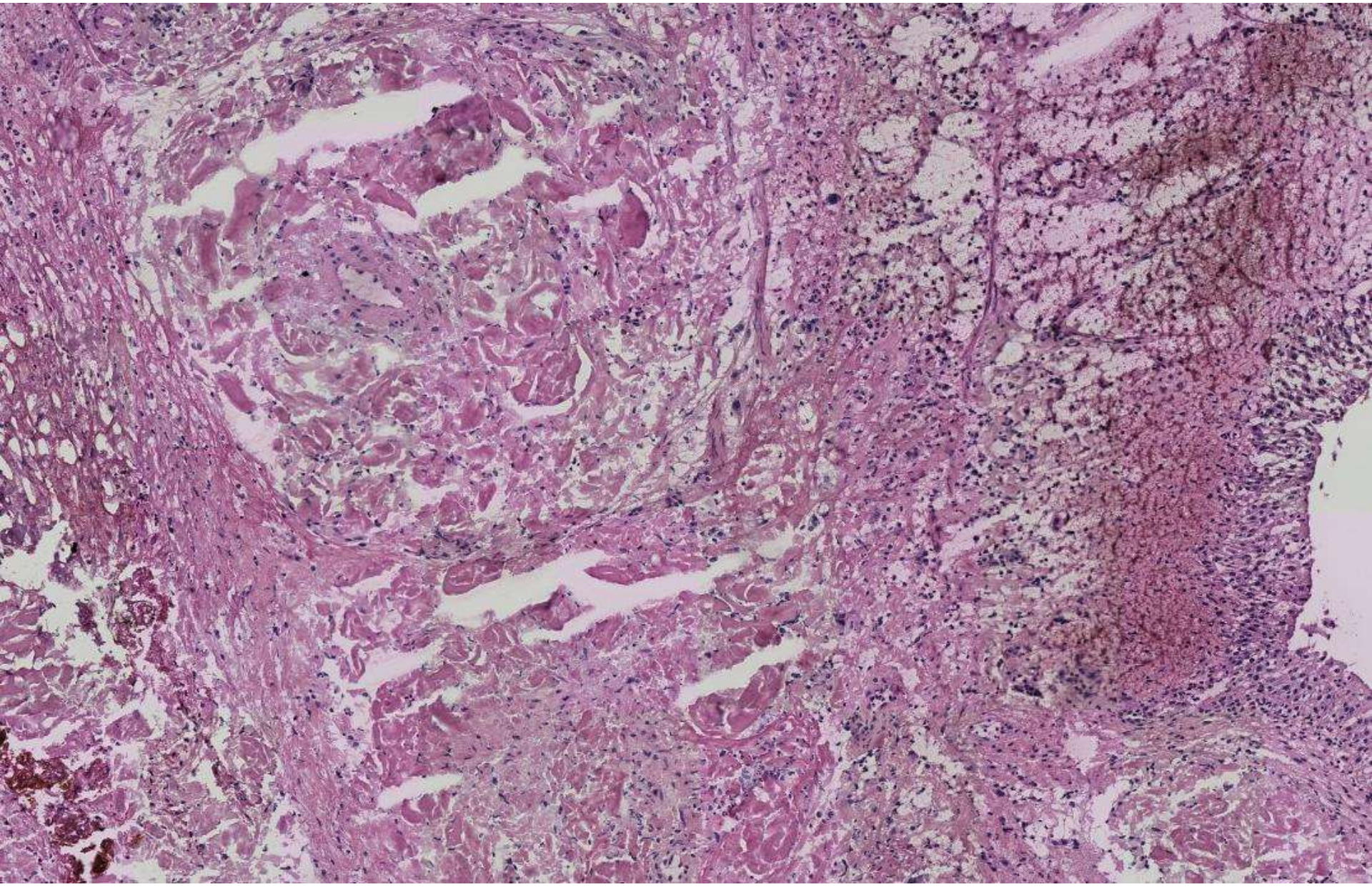


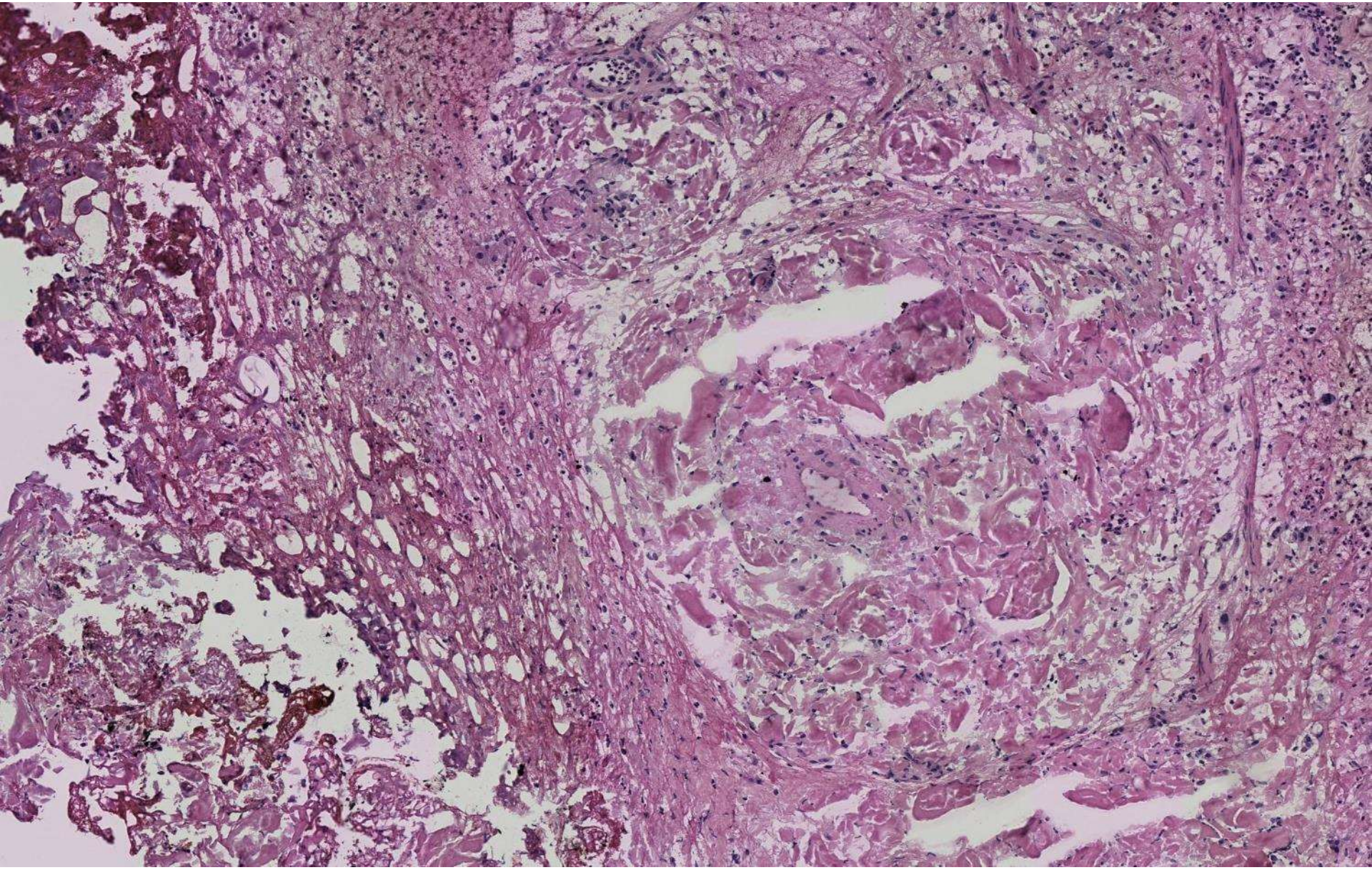


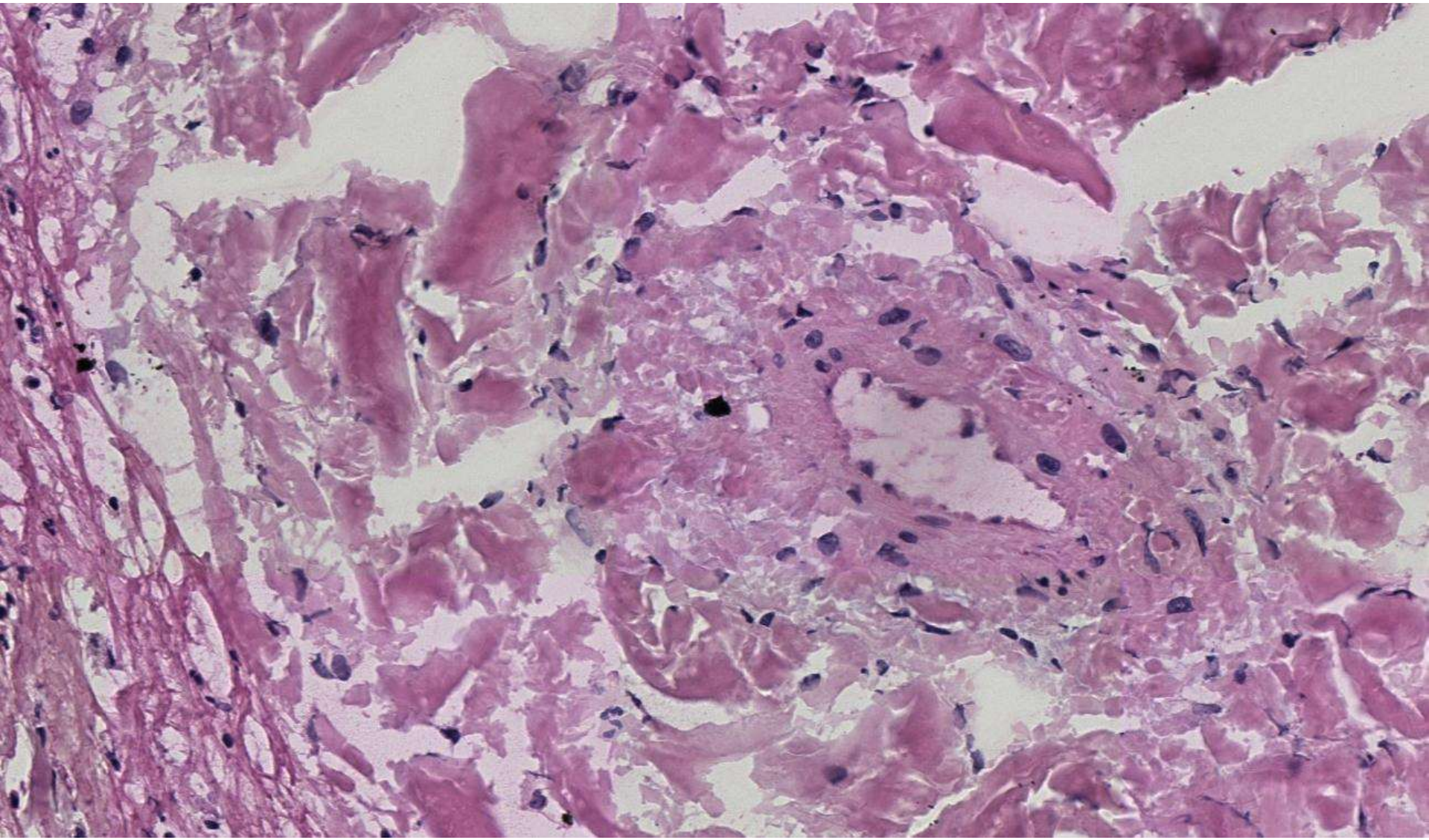








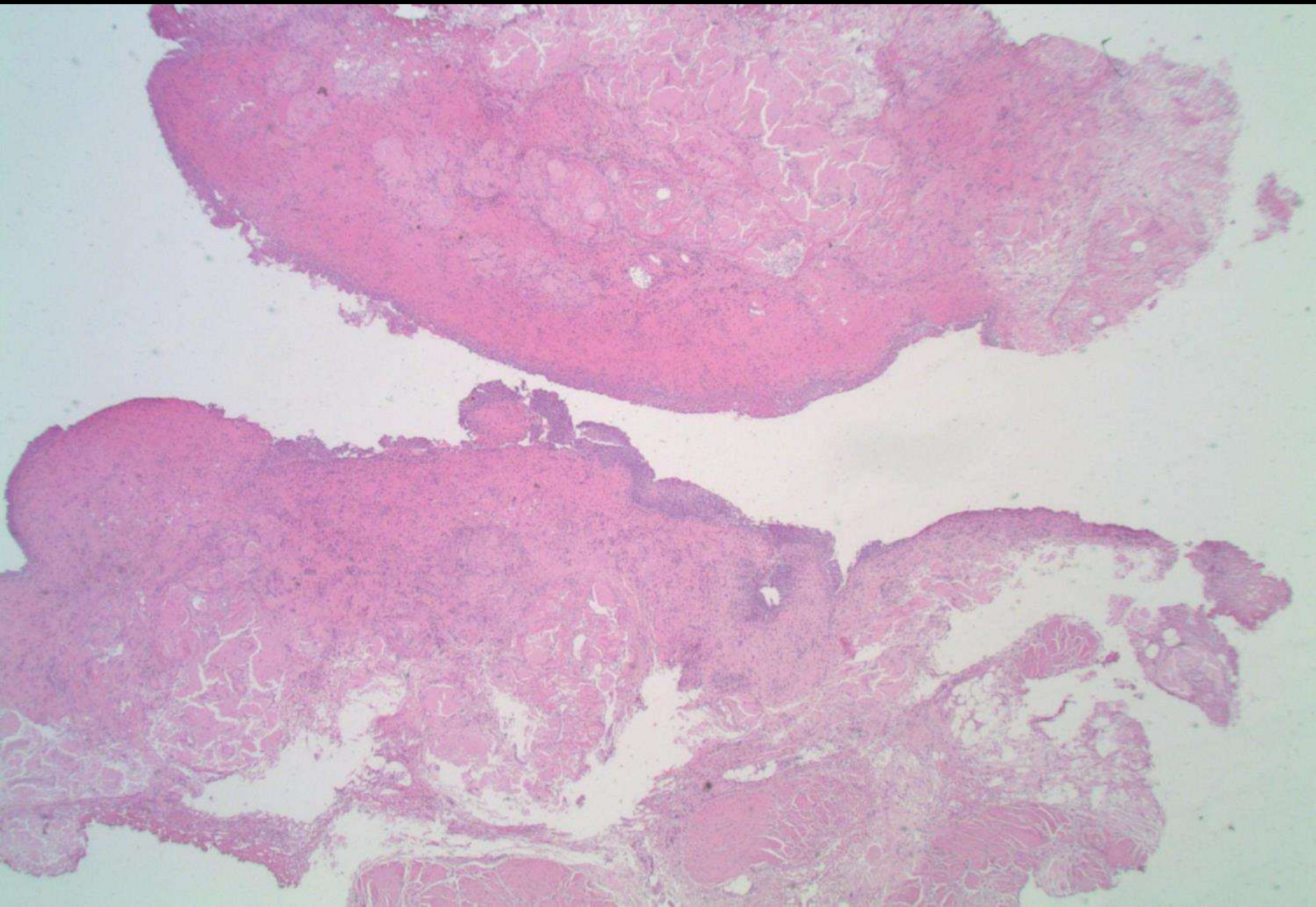




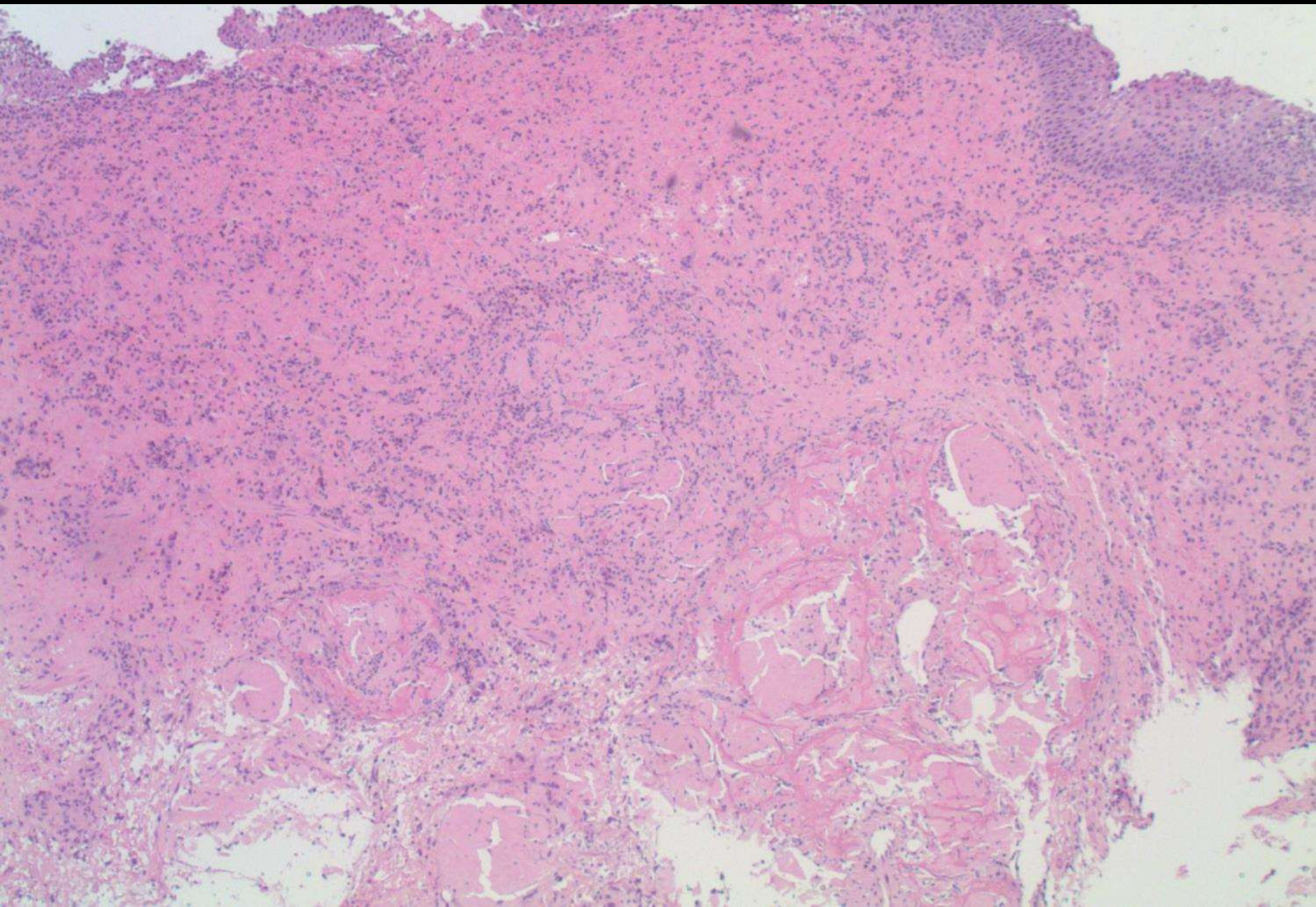
DDx

- Amyloidosis
- Fibrosis/granulation tissue
- Granuloma
- Vasculitis
- Hyalinized schwannoma

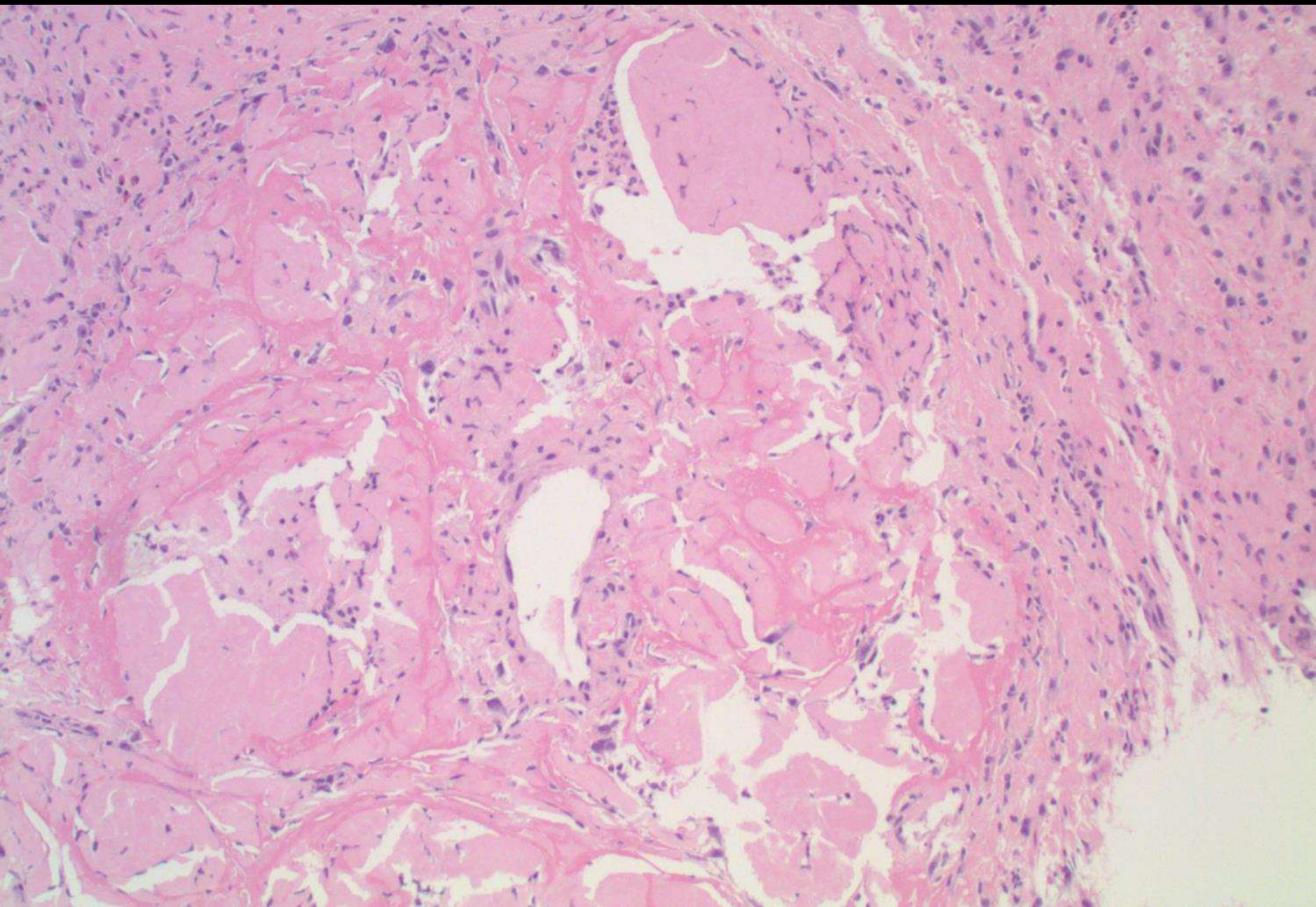
permanent sections of frozen

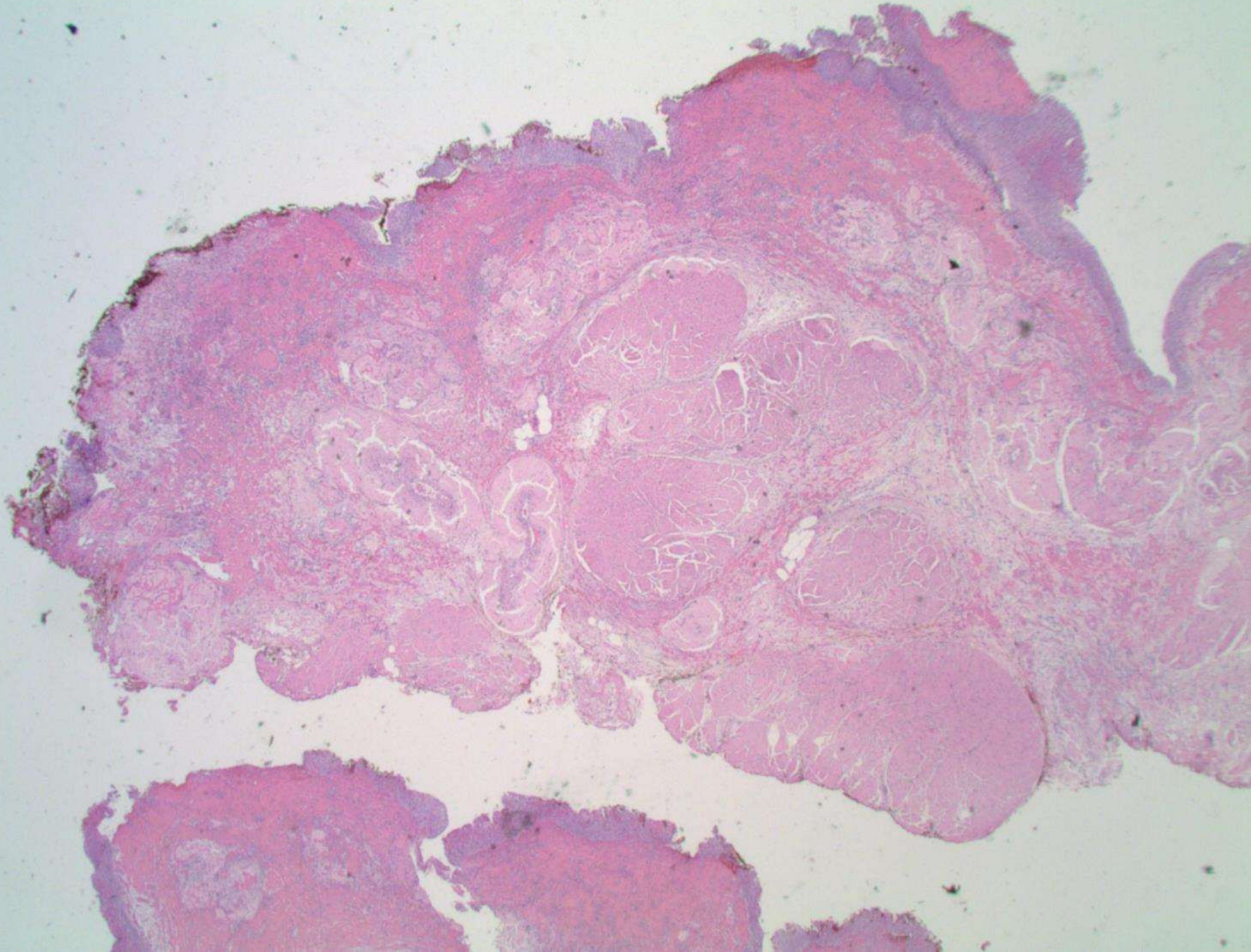


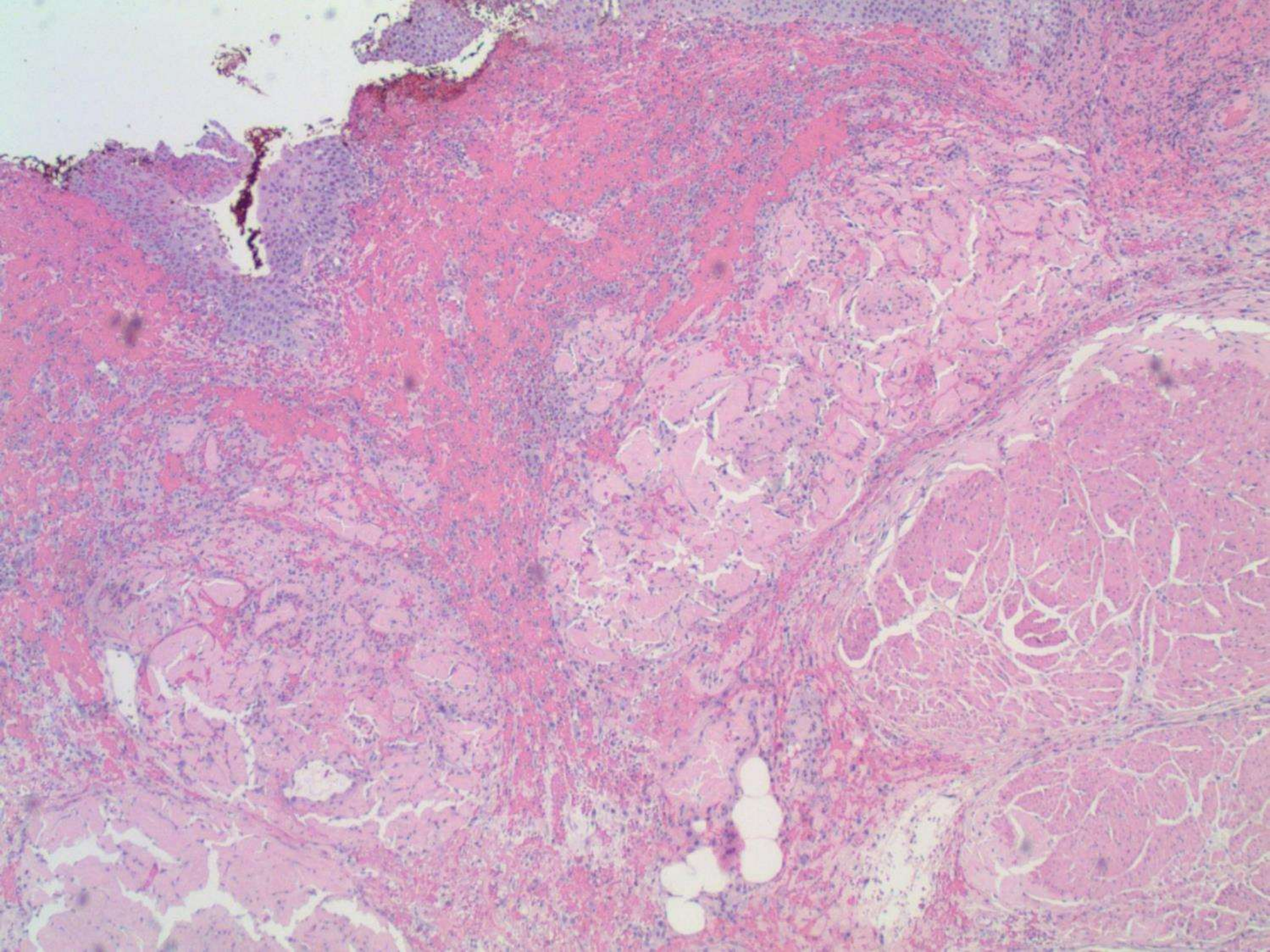
permanent sections of frozen

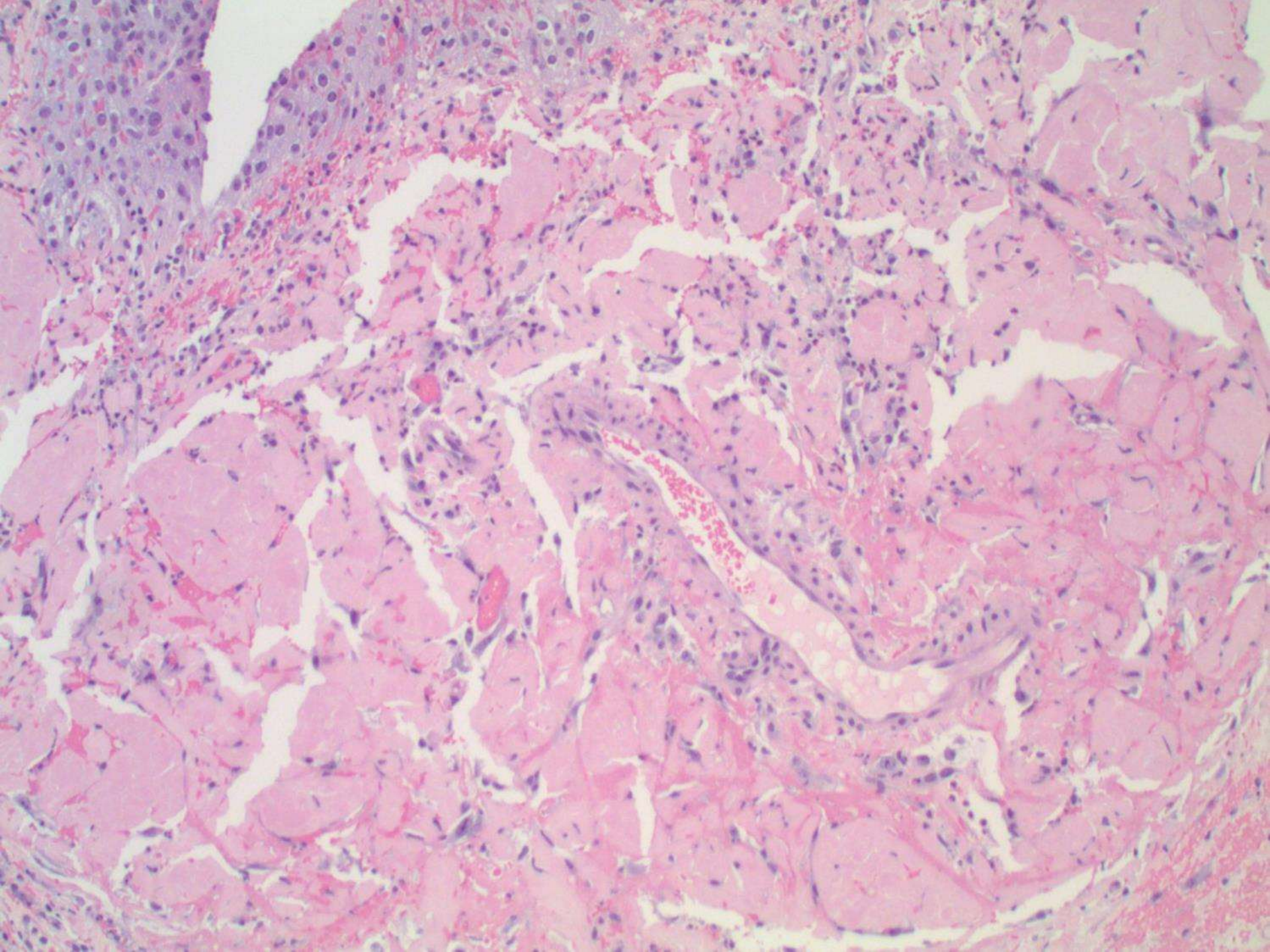


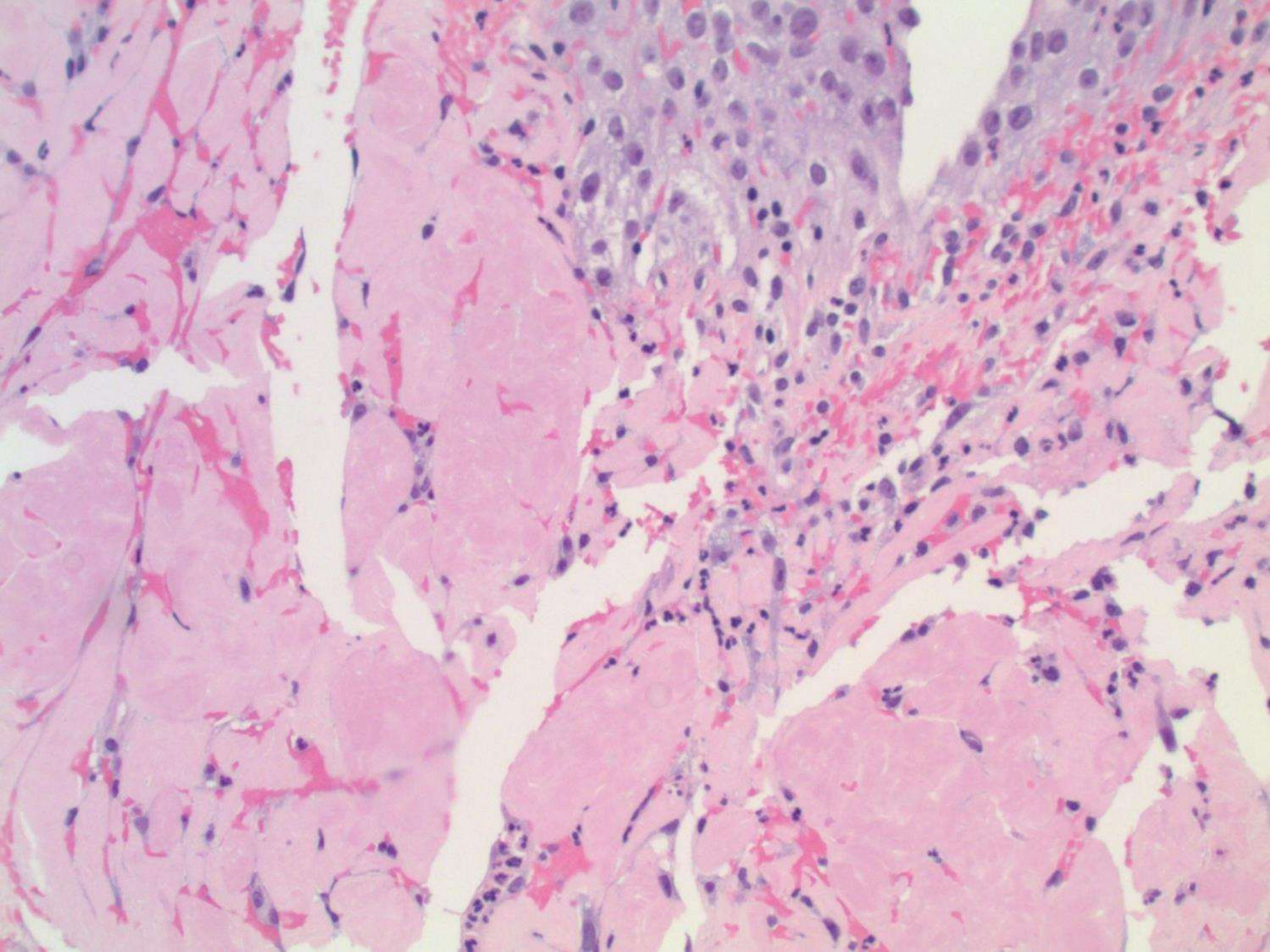
permanent sections of frozen

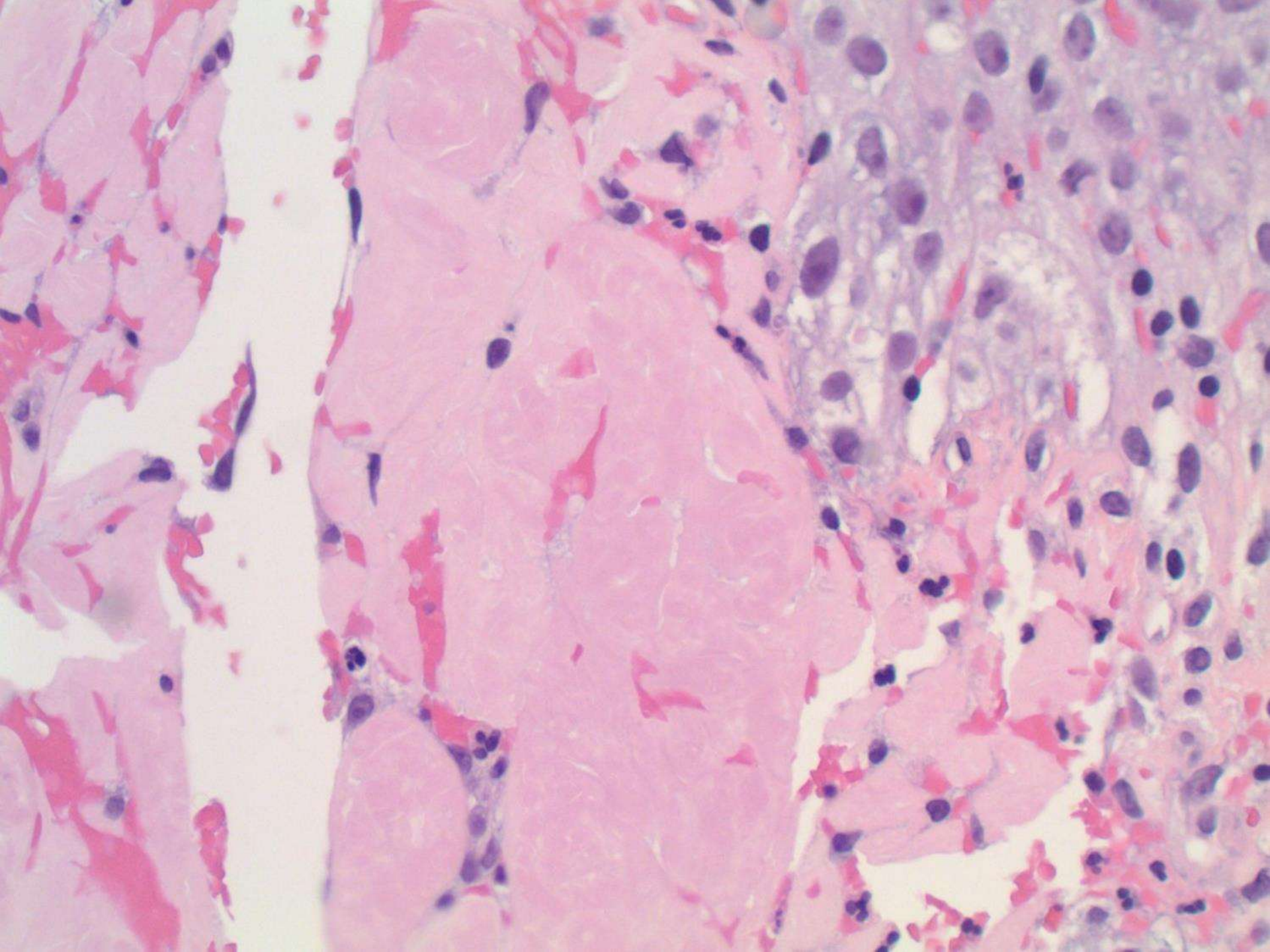




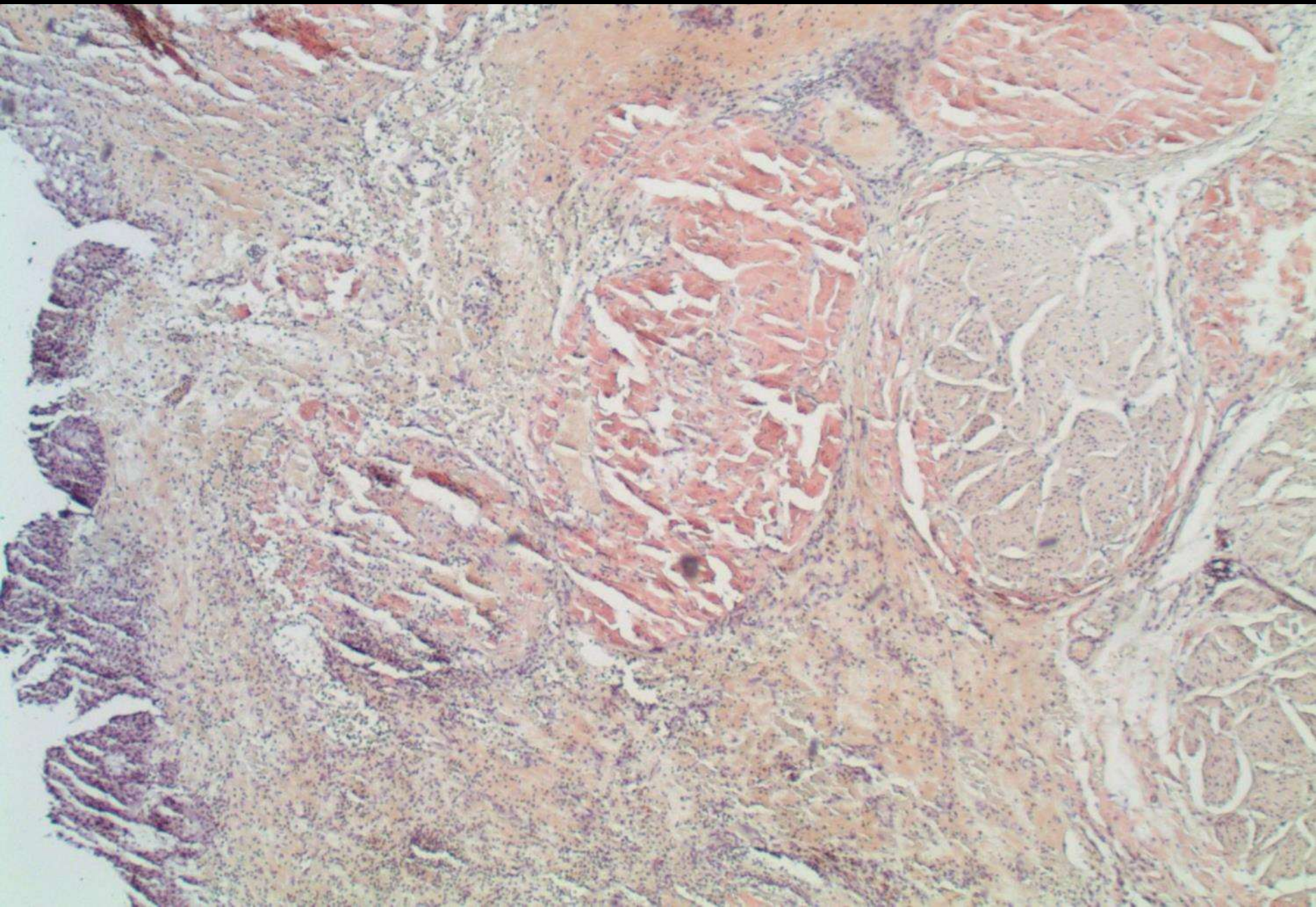




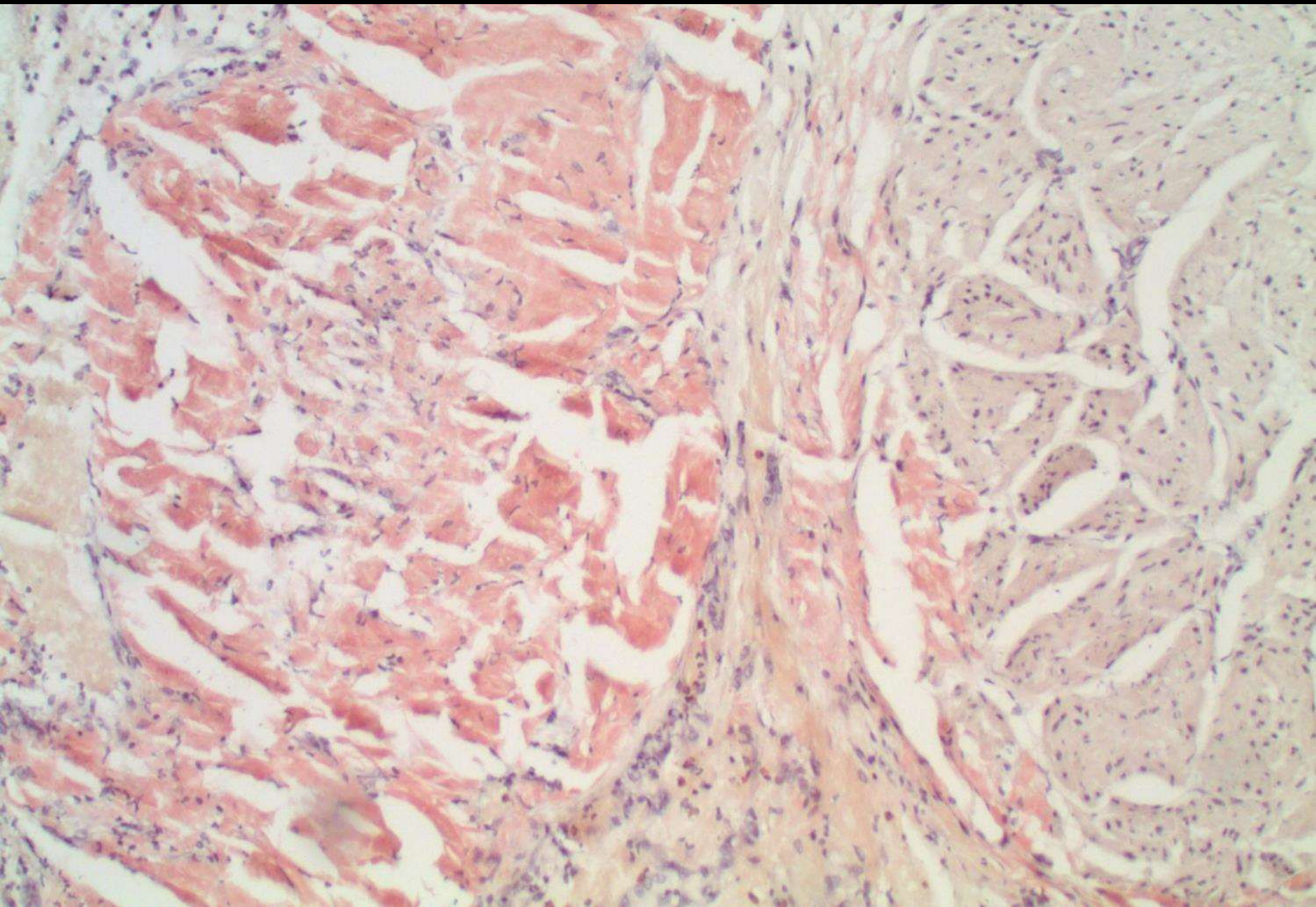




Congo red



Congo red



Congo red (polarized)

Additional work-up

- **History**
  - no prior malignancy or systemic amyloidosis
- **IHC**
  - polyclonal plasma cells
- **Mass spec testing**
  - ATTR (transthyretin) type amyloid

Urinary bladder amyloidosis

- **CLINICALLY**
  - Rare (Southbay!); usually >50 y/o
  - Presents as hematuria
  - May mimic neoplasm cystoscopically
  - Amyloid subtyping key for clinical work-up
  - Primary amyloidosis has high local recurrence rate
    - Partial cystectomy may be required

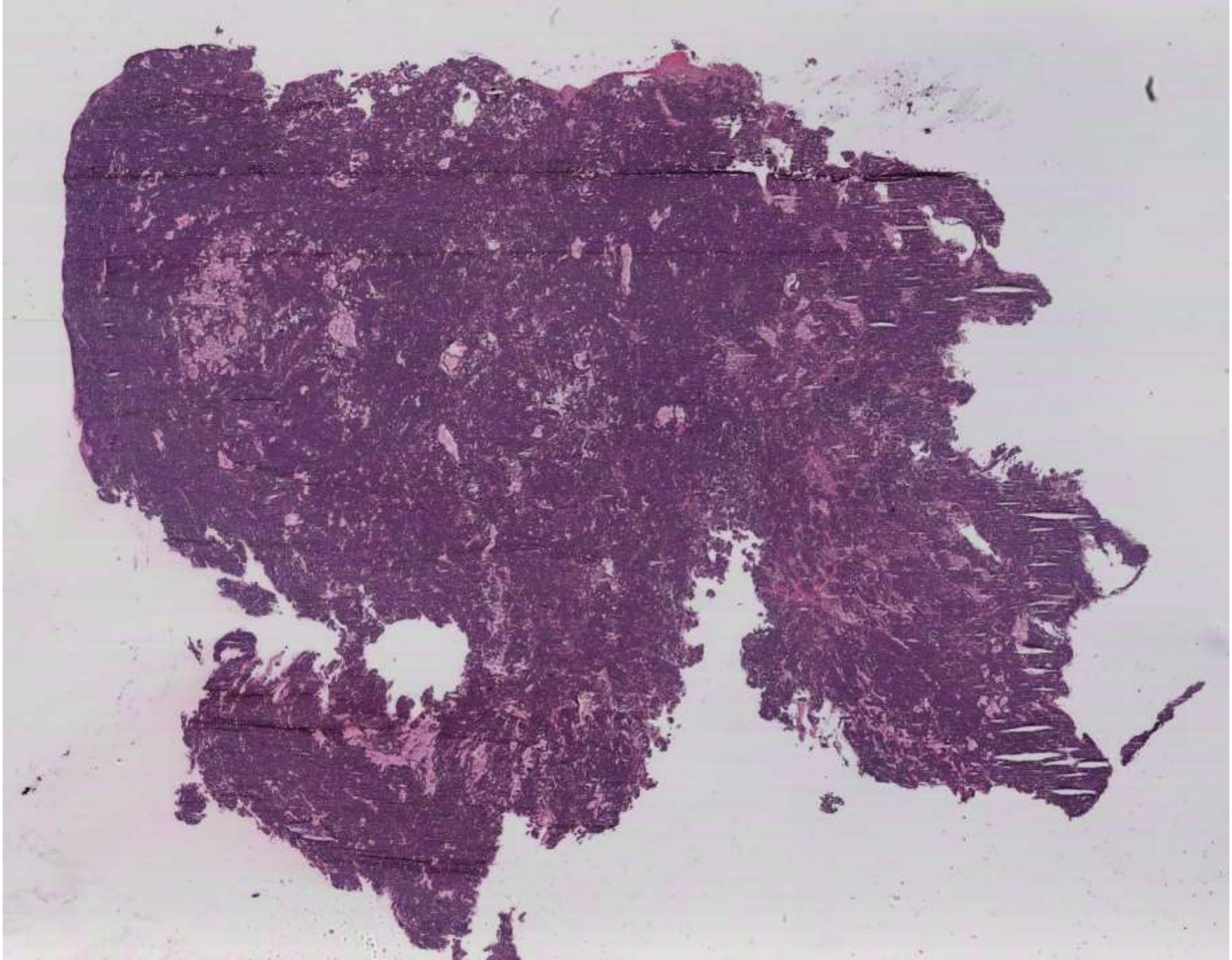
Urinary bladder amyloidosis

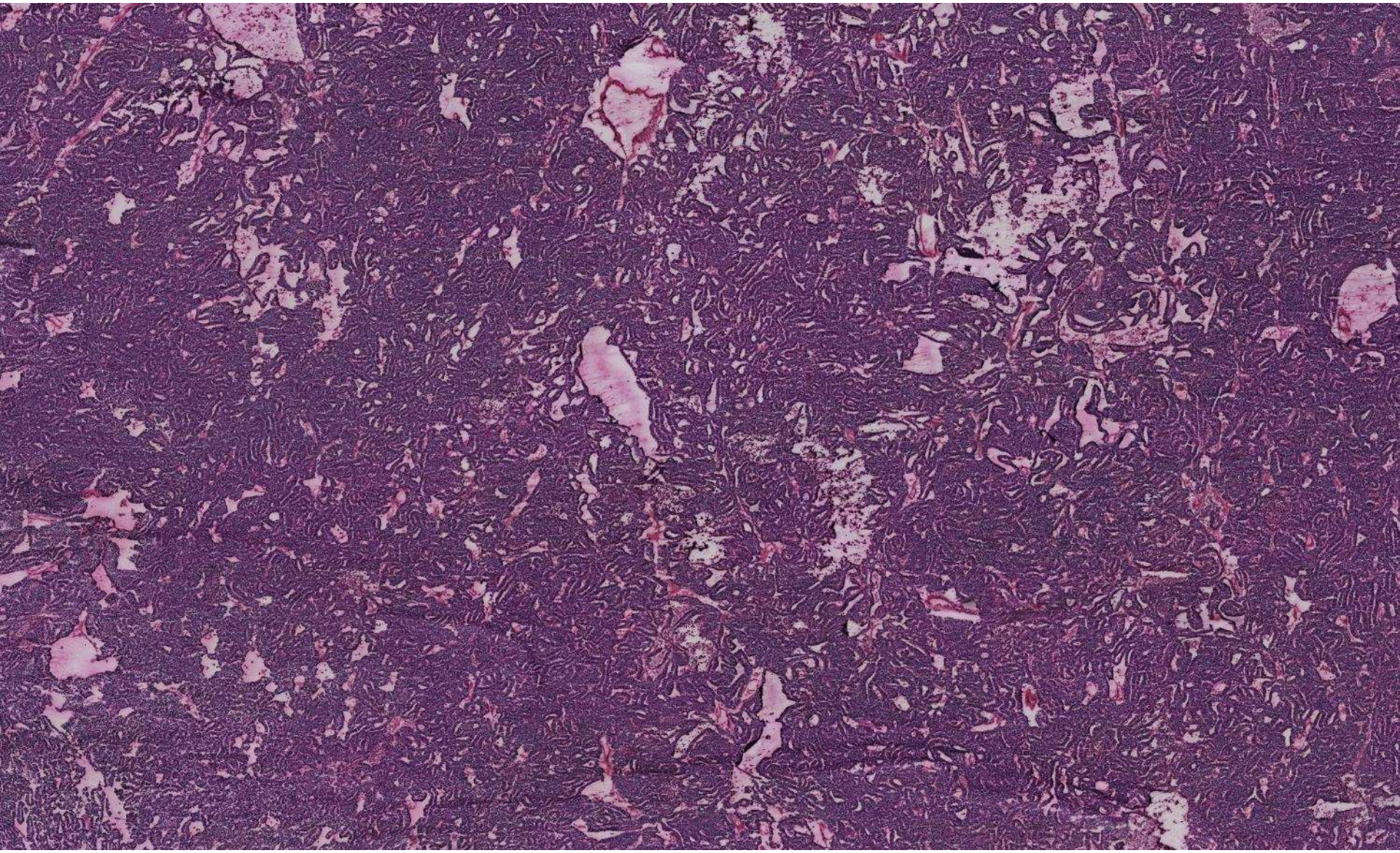
- **HISTOLOGICALLY**
  - Because uncommon, often overlooked as organizing fibrosis
  - Dx doesn’t necessarily indicate systemic process!

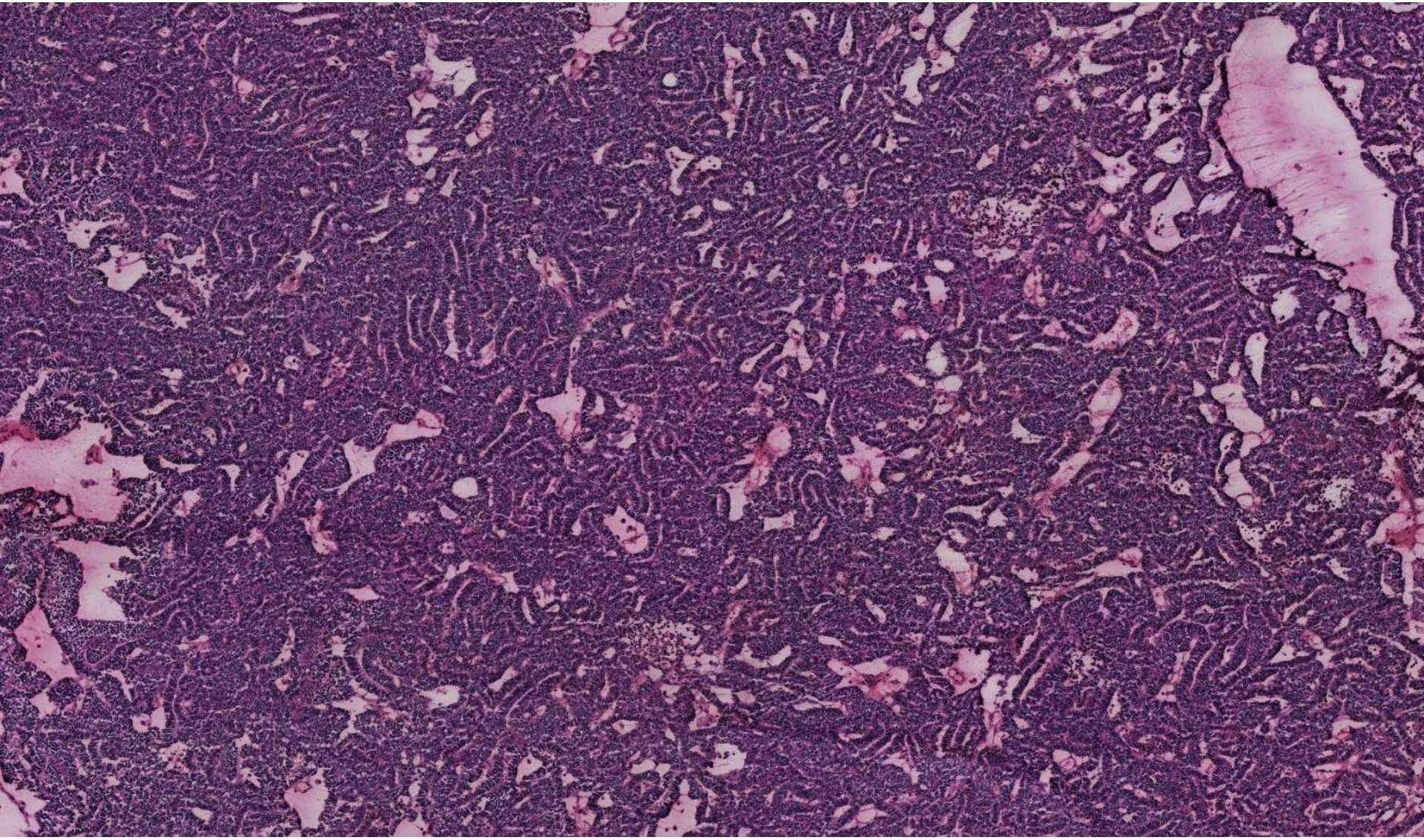
20-0406
SCANNED SLIDE AVAIL!

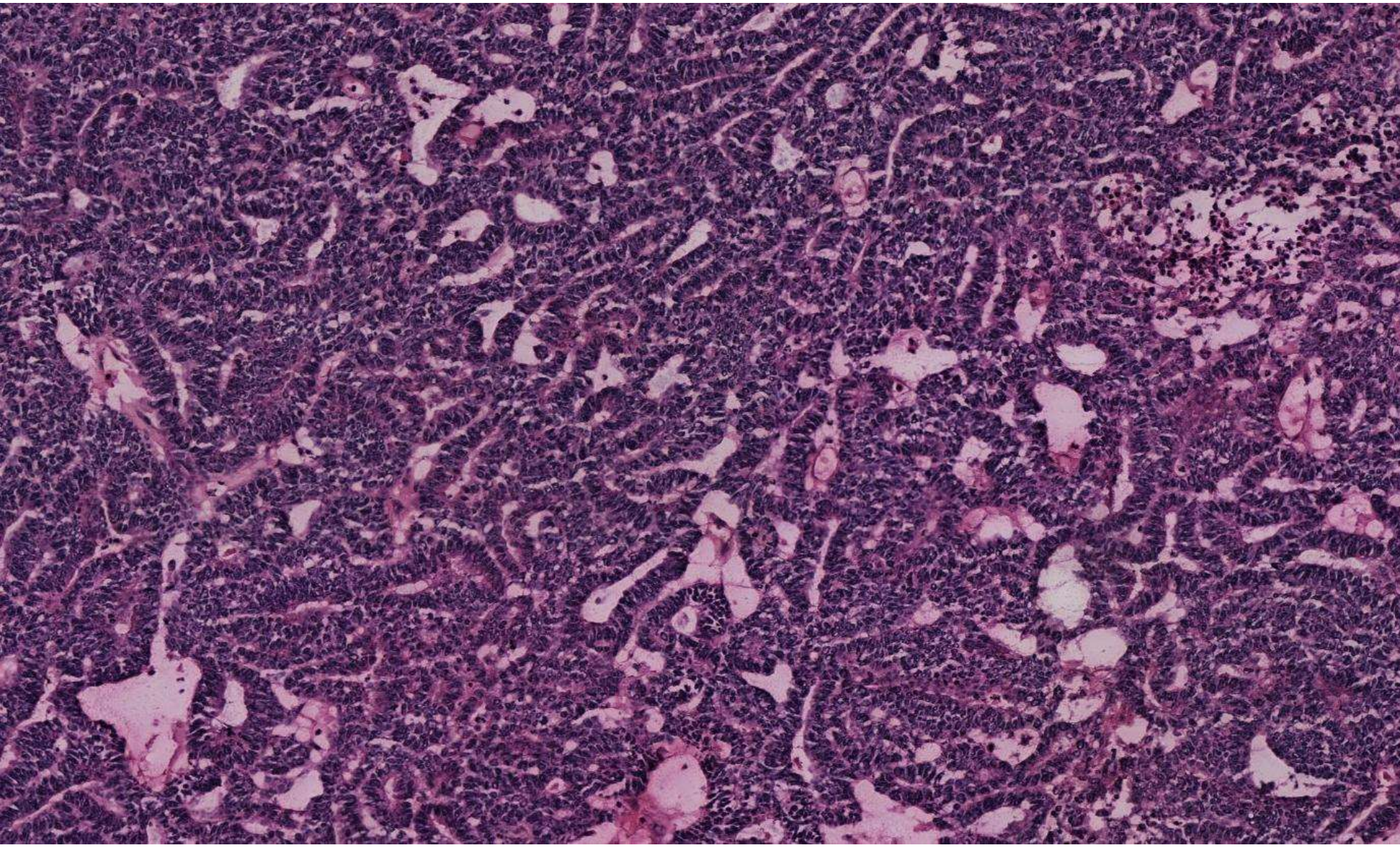
Saba Ali; El Camino Hospital

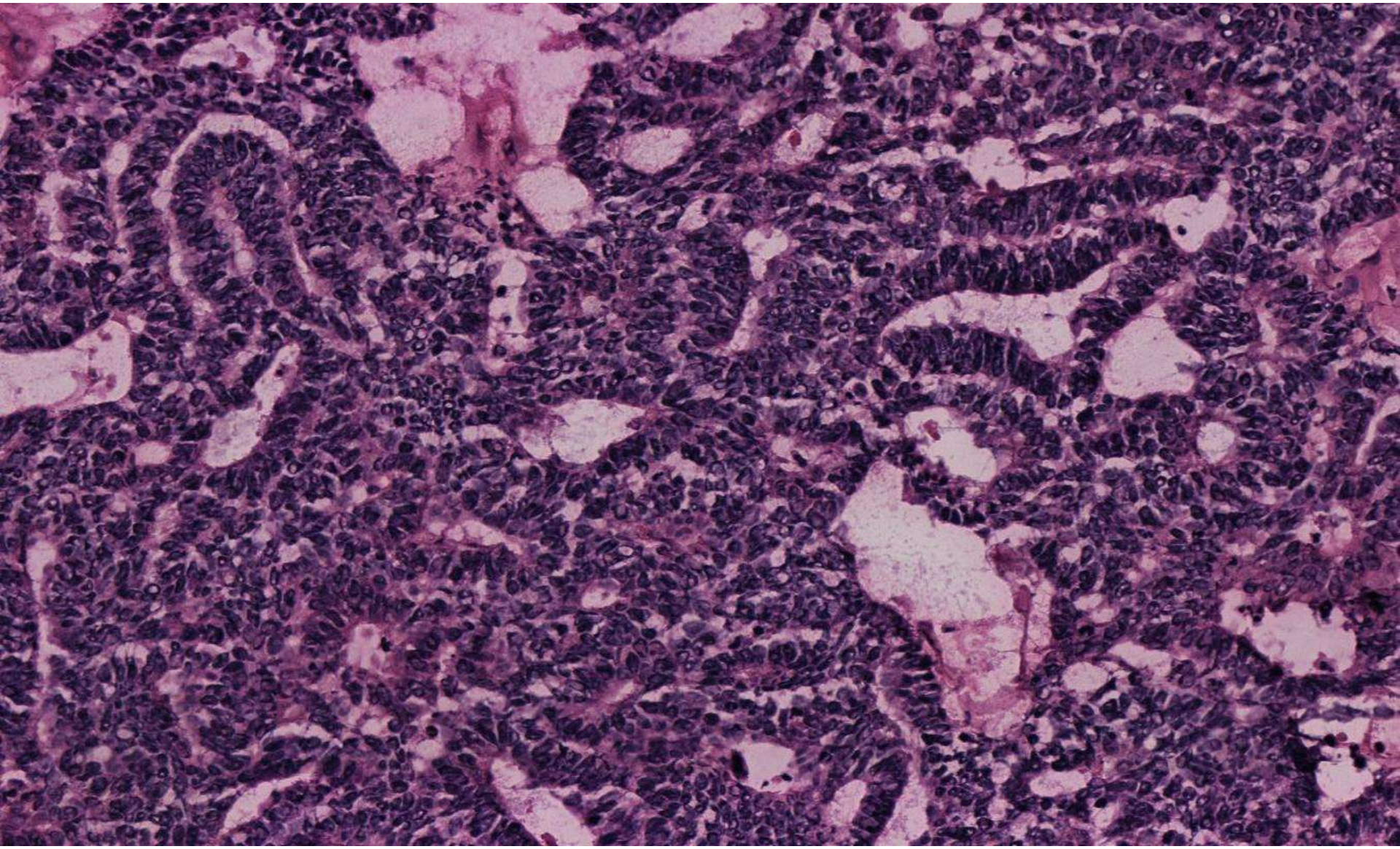
70-year-old F with 12cm firm tan left ovarian mass, submitted for frozen section.

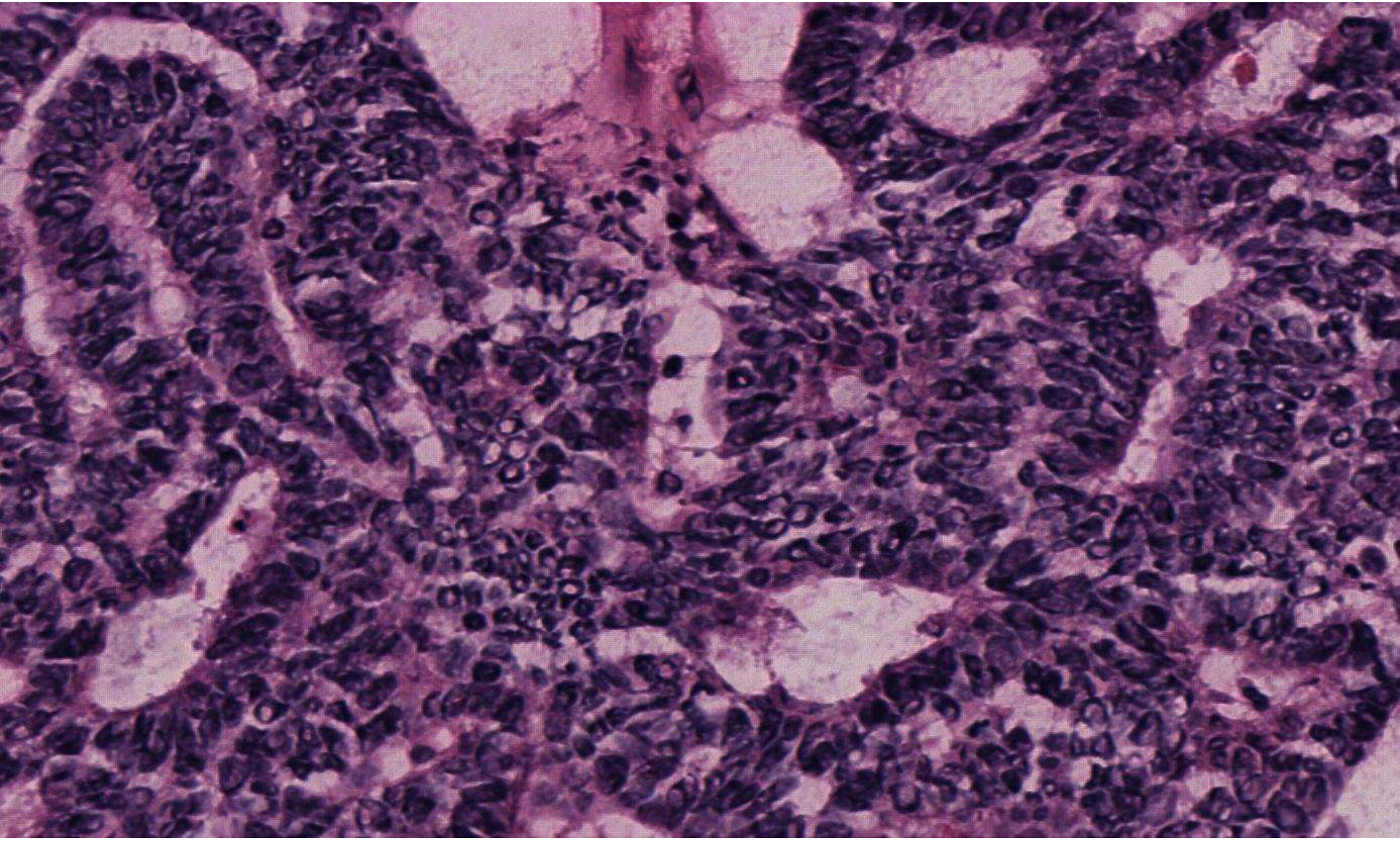


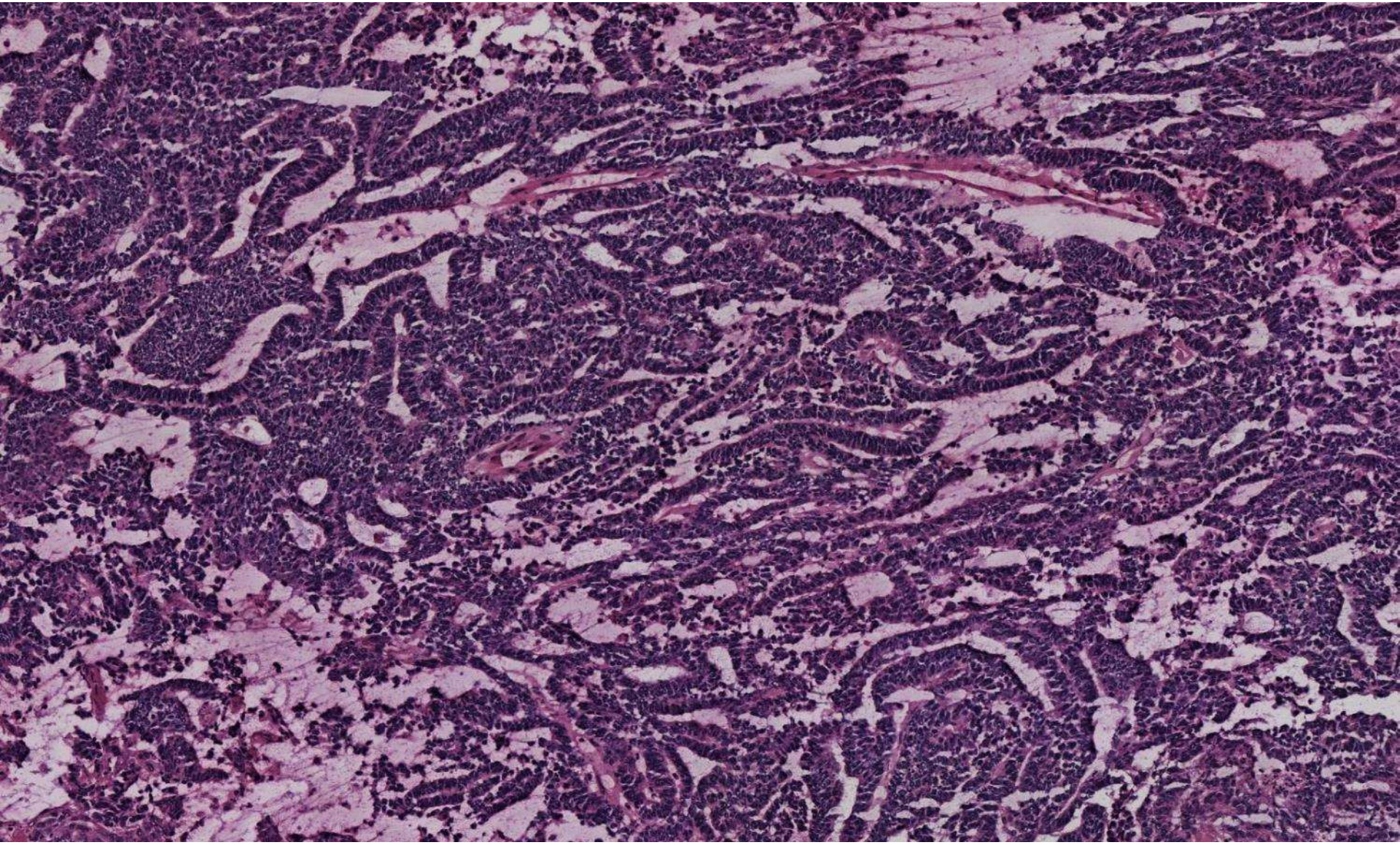


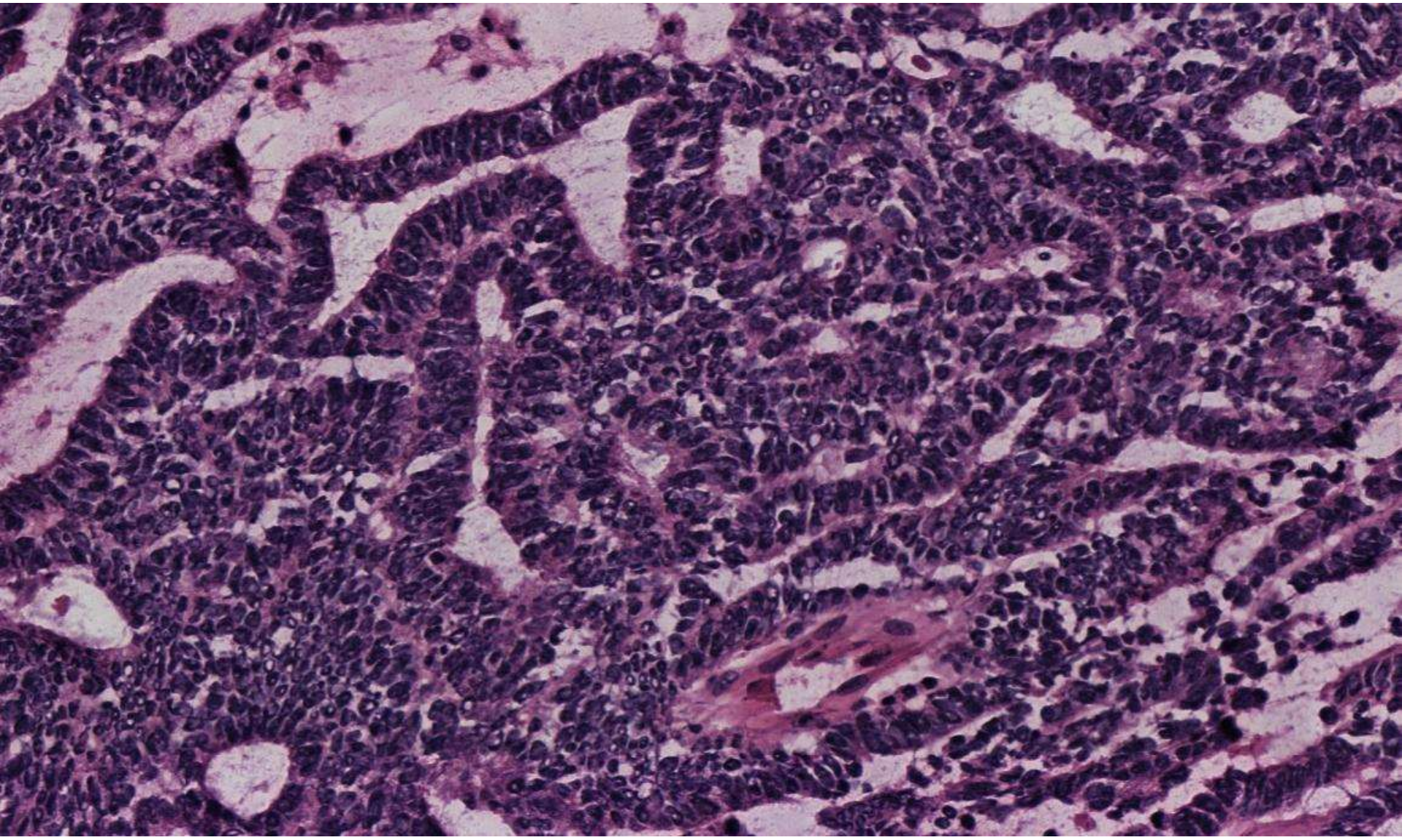


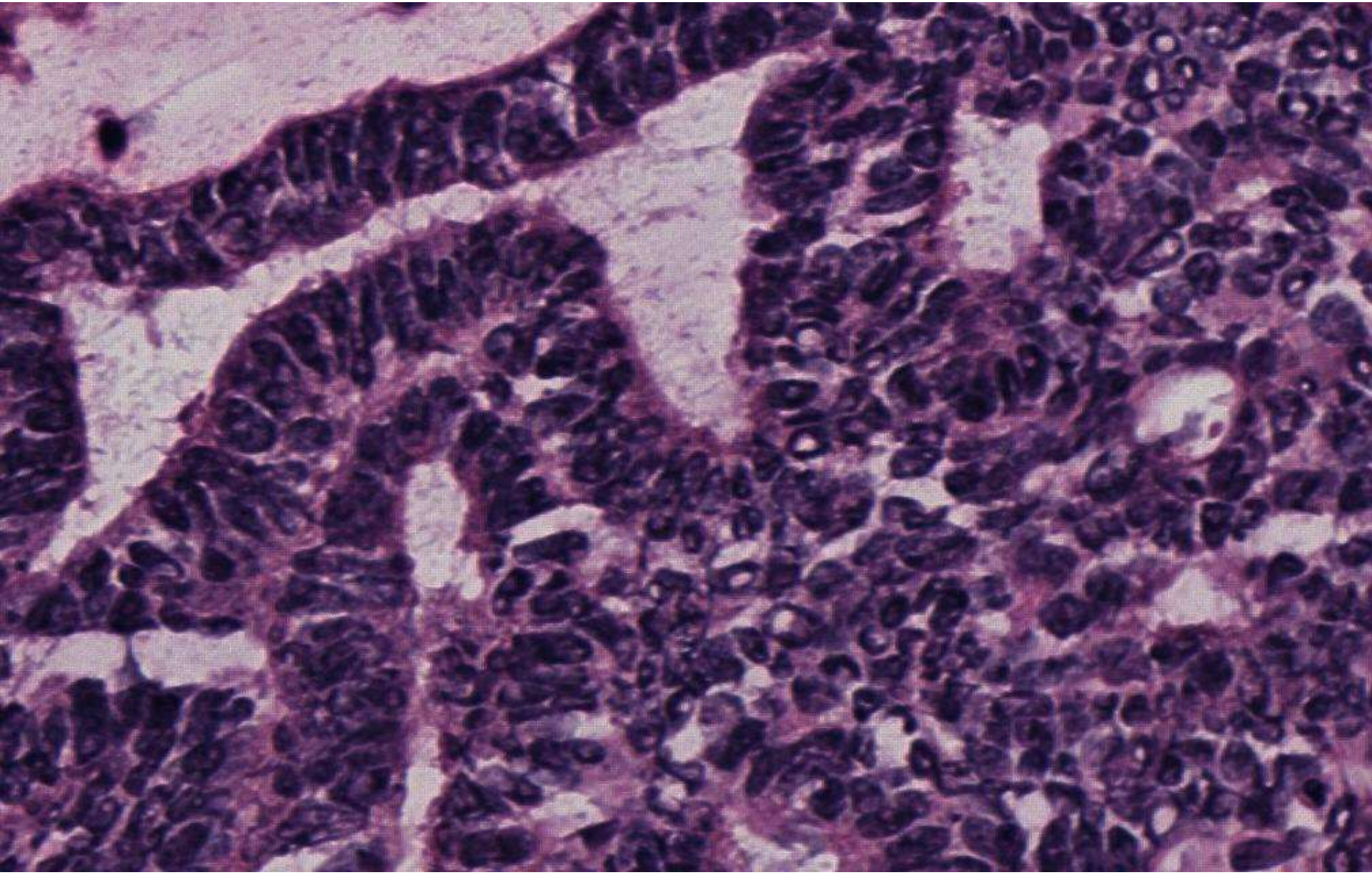






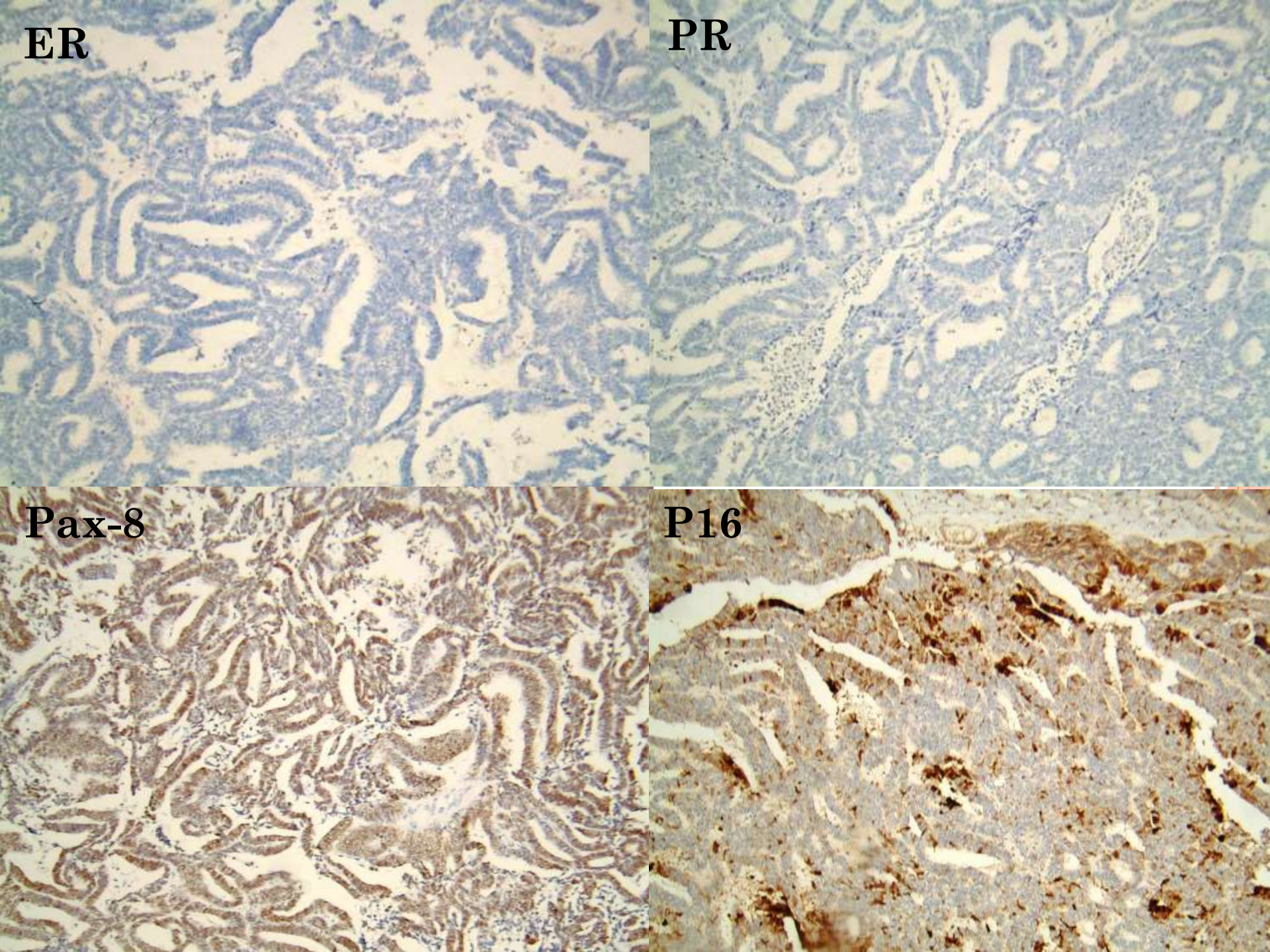


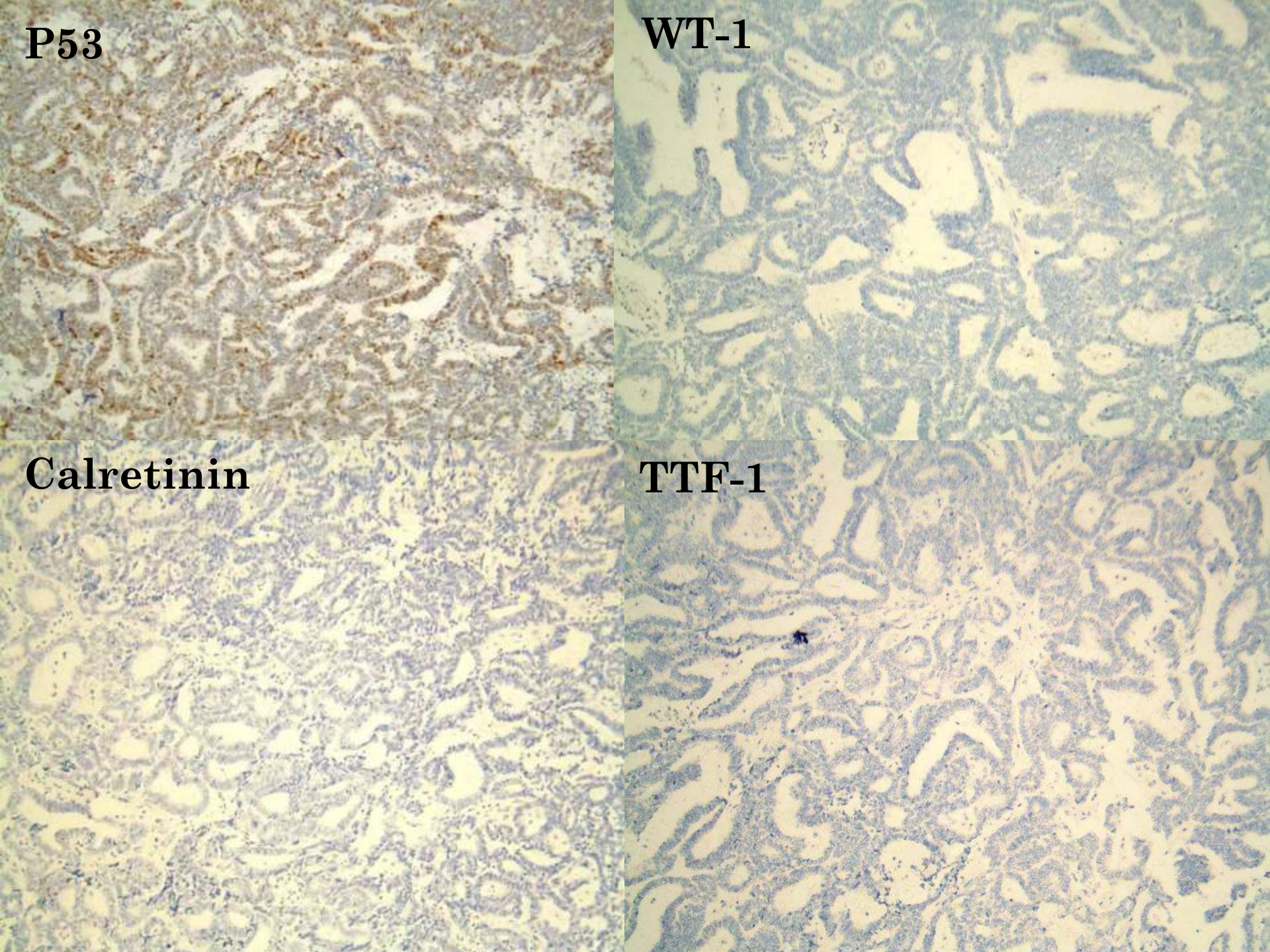


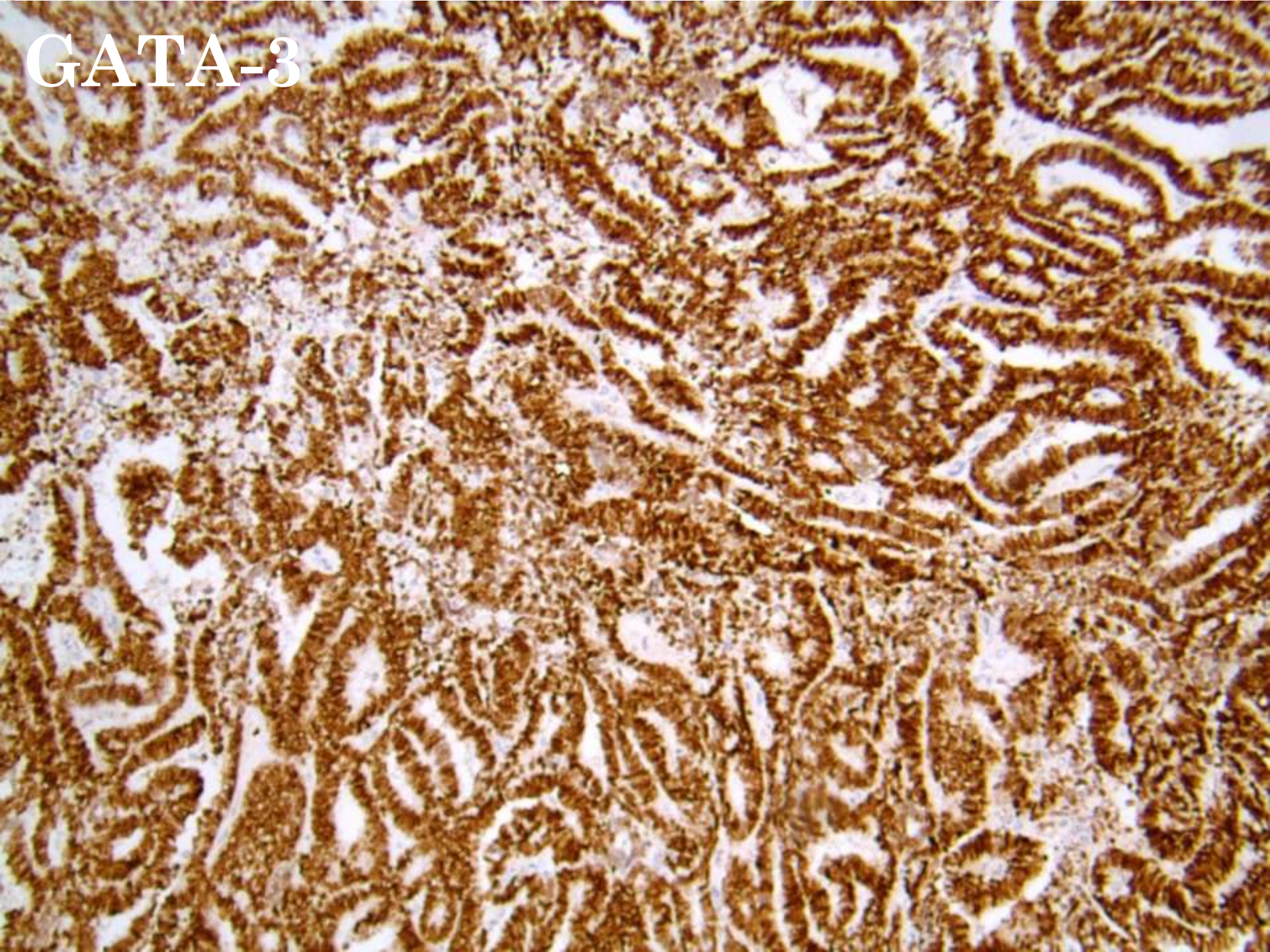


Differential diagnosis?

- Sex cord stromal
- Endometrioid adenocarcinoma
- Low-grade serous carcinoma
- Mesonephric-like carcinoma
- Germ cell tumor
- Usual endocervical type adenocarcinoma

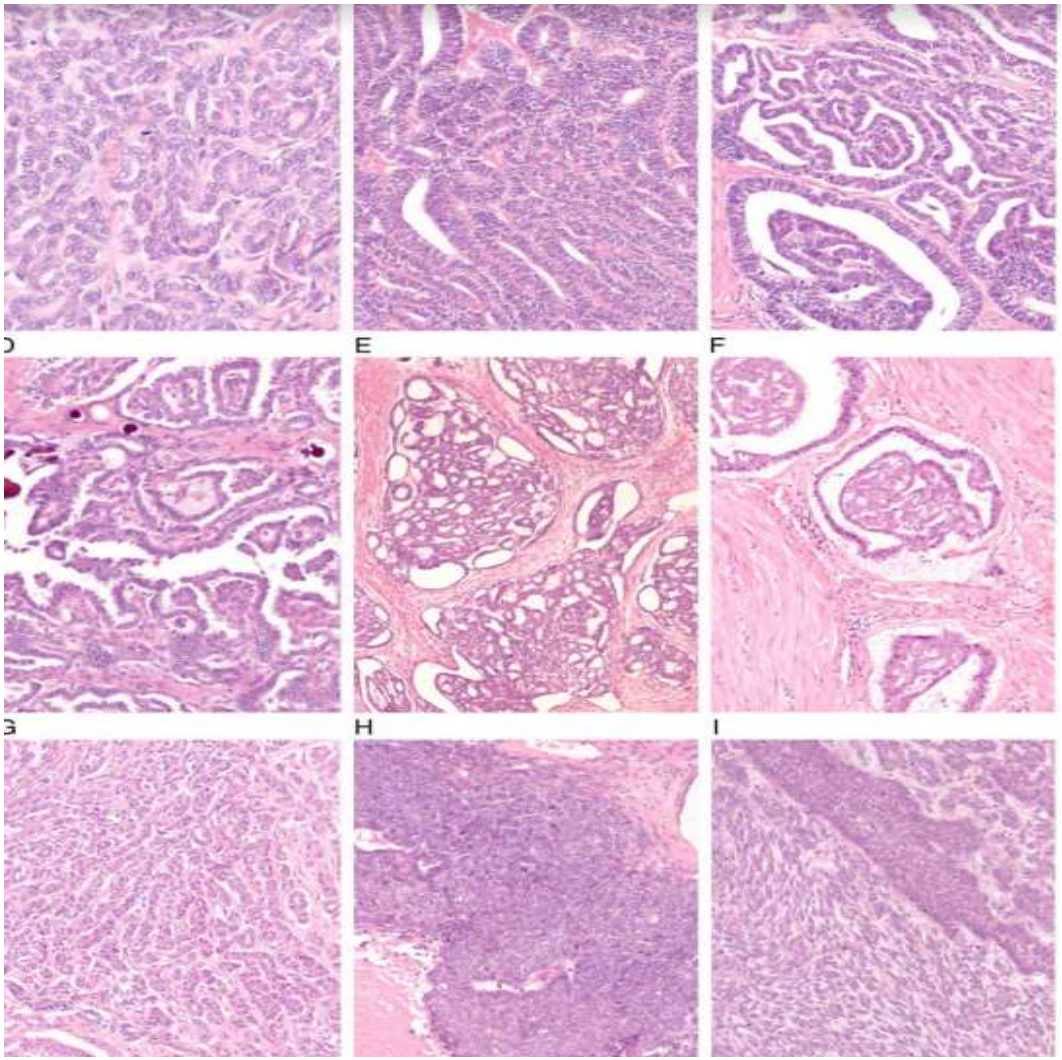

ER
PR
Pax-8
P16


P53
WT-1
Calretinin
TTF-1


GATA-3

MESONEPHRIC-LIKE CARCINOMA OF THE OVARY

- Derived from mesonephric/Wolffian remnants
- Most commonly originate in cervix “mesonephric carcinoma”
- Originate in uterine corpus and ovary termed “mesonephric-like carcinoma”, no remnants seen adjacent
- Can have sarcomatous component (malignant mixed mesonephric tumor)



A Comparison of GATA3, TTF1, CD10, and Calretinin in Identifying Mesonephric and Mesonephric-like Carcinomas of the Gynecologic Tract

Jennifer Pors, MD, Angela Cheng, BSc,† Joyce M. Leo, MD,‡ Mary A. Kinloch, MD,§ Blake Gilks, MD,*† and Lynn Hoang, MD*†*

(*Am J Surg Pathol* 2018;42:1596–1606)

Mesonephric Adenocarcinomas of the Uterine Cervix and Corpus

HPV-negative Neoplasms That Are Commonly PAX8, CA125, and HMGA2 Positive and That May Be Immunoreactive With TTF1 and Hepatocyte Nuclear Factor 1-β

Kenny, Sarah L. MB*; McBride, Hilary A. FIBMS*; Jamison, Jackie FIBMS†; McCluggage, W. Glenn FRCPath*

Author Information

The American Journal of Surgical Pathology: June 2012 - Volume 36 - Issue 6 - p 799-807
doi: 10.1097/PAS.0b013e31824a72c6

Marker	Negative	Focal	Diffuse
CD10	2	6	0
EMA	0	2	6
vimentin	0	3	5
calretinin	1	6	1
inhibin	4	4	0
CEA	5	2	1
ER	6	2	0
p16	3	4	1
PAX8	1	0	7
HMGA2	1	5	2
CA125	0	6	2
TTF1	5	1	2
WT1	8	0	0
HNF1-β	5	0	3

A Comparison of GATA3, TTF1, CD10, and Calretinin in Identifying Mesonephric and Mesonephric-like Carcinomas of the Gynecologic Tract

Jennifer Pors, MD, Angela Cheng, BSc,† Joyce M. Leo, MD,‡ Mary A. Kinloch, MD,§ Blake Gilks, MD,*† and Lynn Hoang, MD*†*

(*Am J Surg Pathol* 2018;42:1596–1606)

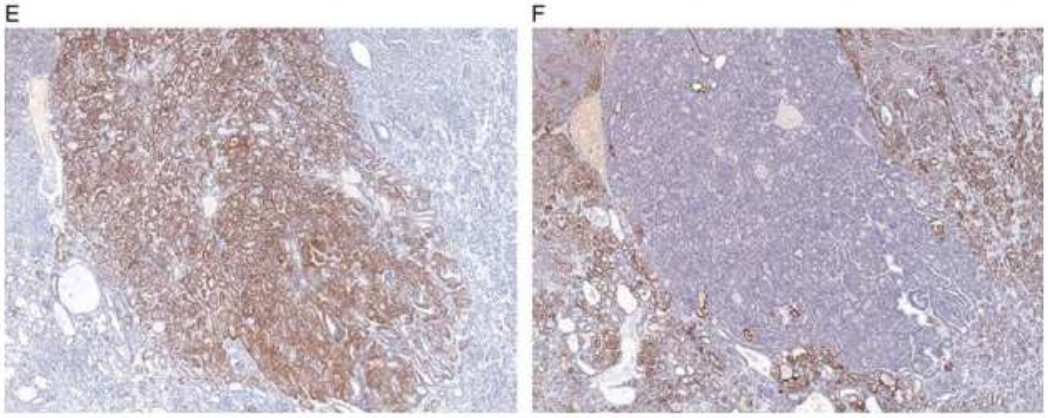
TABLE 1. Summary of Clinical and Immunohistochemical Features of Mesonephric and Mesonephric-like Carcinomas

				Mesonephric Carcinoma Immunohistochemistry (Intensity*; Proportion)						MNR
Case	Site	Age (y)	FIGO Stage	GATA3	TTF1	PAX8	ER	CD10	Calretinin	Present/ Absent
1	Endometrial	65	pT4b	++; 5%	+++; 90%	++/+++; 95%	+/++; 55%	+++; 20% (L)	–	Absent
2	Endometrial	31	pT3a	++/+++; 20%	+++; 60%†	+++; 100%	–	+++; 50% (L)	–	Absent
3	Endometrial	75	pT1b	++/+++; 40%	++/+++; 60%†	+++; 100%	–	+++; 10% (L)	–	Absent
4	Endometrial	91	pT3a	++/+++; 20%	++/+++; 90%†	+++; 100%	+; <10%	–	–	Absent
5	Cervix‡	49	pT2b	–	–	+/++; 80%	–	+++; 50% (L)	–	Present
6	Cervix	78	pT1b1	+++; 90%	–	++/+++; 95%	–	–	++; 50%	Present
7	Cervix	64	pT1b1	+; <5%	–	+/++; 95%	–	++; 10% (L)	++; 10%	Present
8	Cervix	50	pT3a	+/++; 10%	–	+++; 100%	–	++; 10% (L)	++; 10%	Present
9	Cervix‡	49	pT2b	++/+++; 90%	+++; 10%	+/++; 95%	–	++; 10% (L)	++; 10%	Absent
10	Cervix	62	pT1b2	++/+++; 80%	–	+++; 100%	–	+; 30% (L)	–	Present
11	Cervix‡	59	pT2b	+++, 50%	–	+++, 50%	–	–	–	Present
12	Ovary	67	pT1c	++/+++; 70%	++/+++; 20%†	++/+++, 90%	–	+++, 50% (L)	–	Absent
13	Vagina§	66	pT3	+++; 95%	–	+++/100%	–	+++, 40% (NL)	–	Absent

TABLE 4. Comparison of GATA3, TTF1, CD10, and Calretinin Performance in Distinguish Mesonephric Carcinomas From Other Endometrial and Endocervical Adenocarcinomas*

	GATA3 (%)	TTF1 (%)	CD10 (%)	Calretinin (%)
Sensitivity	91	45	73	36
Specificity	94	99	83	89

*Excludes mesonephric carcinomas occurring in the vagina and ovary.



MOLECULAR

A Combined Morphologic and Molecular Approach to Retrospectively Identify *KRAS*-Mutated Mesonephric-like Adenocarcinomas of the Endometrium

David L. Kolin, MD, PhD, Danielle C. Costigan, MBBCh,† Fei Dong, MD,†*
Marisa R. Nucci, MD, and Brooke E. Howitt, MD*‡*

2019;43:389–398)

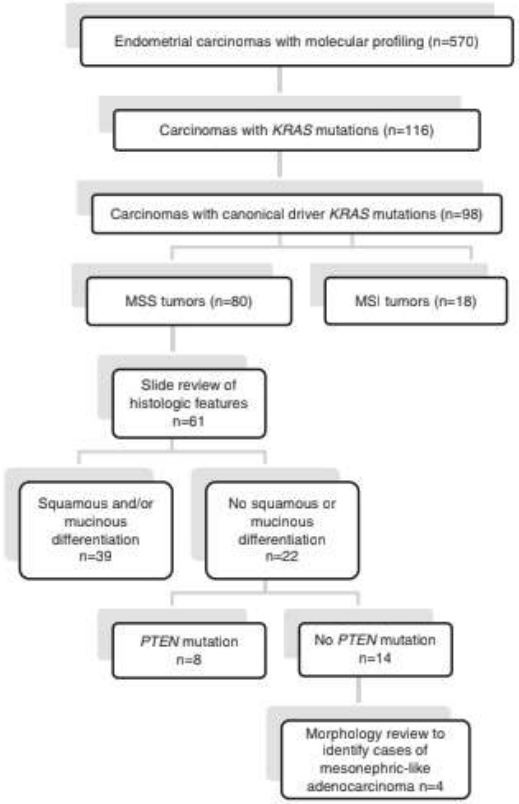


FIGURE 1. Algorithm used to identify cases of mesonephric-like adenocarcinoma amongst a cohort of endometrial carcinomas.

TABLE 3. Cases of Retrospectively Identified Mesonephric-like Adenocarcinoma, Their Original Diagnoses, Clinical Follow-up, and Molecular Features

Case	Original Diagnosis	Age (y)	FIGO Stage	Follow-up Period (y)	Outcome	Immunohistochemistry	DNA Variants	CNV
1	Carcinosarcoma	64	IB	12.5	AWD (lung metastases)	POS: TTF-1, GATA-3 (focal), CD10 (patchy, luminal), PAX8, ER (patchy, weak) NEG: PR, synaptophysin, chromogranin, p63	*KRAS* c.35G>C (p.G12A)	1p loss, 1q gain, 10p gain, 10q loss, 21 q loss
2	Endometrioid, grade 1	57	IA	1.6	NED	POS: TTF-1, CD10 (luminal), ER (heterogenous), PR (heterogenous) NEG: GATA-3 p53: wild-type	*KRAS* c.35G>T (p.G12V)	1p loss, 1q gain, 4p loss, 4q loss, 11p loss, 11q loss, 21q loss
3	Endometrioid, grade 2	58	IVB	2.5	AWD (local disease)	POS: p16 (patchy), TTF-1 (patchy), CD10 (patchy, luminal), Napsin A (scattered cells) NEG: ER, PR, GATA-3 p53: wild-type	*KRAS* c.35G>A (p.G12D) *PIK3R1* c.1351_1353delGAA (p.E451del)	1q gain, 11p loss, 11q loss, 13q loss, 17p loss, 22q loss
4	Endometrioid, grade 2	62	IIIC	8.4	DOD, with lung metastases	POS: PAX8, TTF-1 NEG: ER, PR, NapsinA, WT-1, chromogranin, synaptophysin, and thyroglobulin p53: wild-type	*KRAS* c.35G>T (p.G12V)	ND

AWD indicates alive with disease; DOD, died of disease; ND, not done; NED, no evidence of disease.

c-KIT Analysis and Targeted Molecular Sequencing of Mesonephric Carcinomas of the Female Genital Tract

Pors, Jennifer MD*; Ho, Julie BSc†; Prentice, Leah PhD‡; Thompson, Emily MBChB*; Cochrane, Dawn PhD§; Gibbard, Evan BSc‖; Huntsman, David MD‡,§; Gilks, Blake MD*,†,¶; Hoang, Lynn N. MD*,†,‖,¶

Author Information

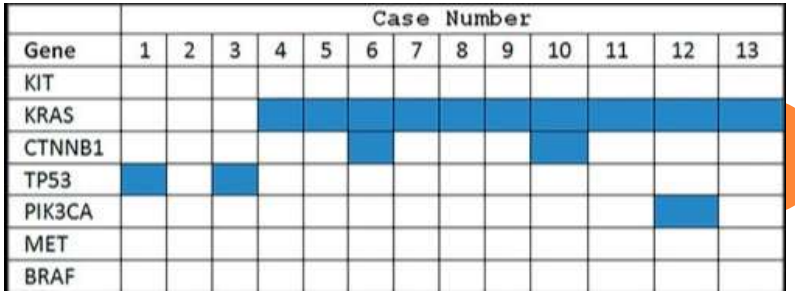
The American Journal of Surgical Pathology: April 2020 - Volume 44 - Issue 4 - p 495-502
doi: 10.1097/PAS.0000000000001403

Case	Age (y)	TNM Stage	c-KIT Immunohistochemistry (Intensity; Proportion)‡	c-KIT Mutation Status
Mesonephric carcinoma				
Cervical				
1*	49	pT2b	Negative	Negative
2	64	pT1b1	++, 80%	Negative
3	50	pT3a	++, 70%	Negative
4	62	pT1b2	++, 5%	Negative
5*	59	pT2b	+, 10%	Negative
Vaginal/ pelvic mass				
6	66	pT3	+++, 40%	Negative
Mesonephric-like carcinoma				
Endometrial				
7	65	pT4b	+, 10%	Negative
8	31	pT3a	+++, 20%	Negative
9	75	pT1b	++, 5%	Negative
Ovarian				
10	67	pT1c	+++, 50%	Negative
11	59	pT2b	Negative	Negative
12	52	pT3	NA	Negative
13	54	pT1c	++, 30%	Negative

*Carcinosarcoma.
‡Intensity scored as: +, weak; ++, moderate; +++, strong.
NA indicates not available.

Case	Missense Mutations KIT	KRAS	CTNNB1	TP53	PIK3CA
Mesonephric carcinoma					
Cervical					
1*	—	—	—	c.841G > A p.Asp281Asn	—
2	—	—	—	—	—
3	—	—	—	c.818G > A p.Arg273His	—
4	—	c.35G > T p.Gly12Val c.149C > T p.Thr50Ile	—	—	—
5*	—	c.35G > A p.Gly12Asp	—	—	—
Vaginal/pelvic mass					
6	—	c.35G > T p.Gly12Val	c.110C > T p.Ser37Phe	—	—
Mesonephric-like carcinoma					
Endometrial					
7	—	c.34G > T p.Gly12Cys	—	—	—
8	—	c.35G > T p.Gly12Val	—	—	—
9	—	c.35G > A p.Gly12Asp	—	—	—
Ovarian					
10	—	c.34G > T p.Gly12Cys	c.94G > A p.Asp32Asn c.98C > G p.Ser33Cys c.110C > T p.Ser37Phe	—	—
11	—	c.35G > A p.Gly12Asp	—	—	—
12	—	c.35G > A p.Gly12Asp	—	—	c.3132T > G p.Asn1044Lys
13	—	c.35G > T p.Gly12Val	—	—	—

*Carcinosarcoma.



PROGNOSIS

Malignant Mesonephric Tumors of the Female Genital Tract

A Clinicopathologic Study of 9 Cases

Bagué, Silvia MD; Rodríguez, Ingrid M MD; Prat, Jaime MD, FRCPATH **Author Information**

The American Journal of Surgical Pathology: May 2004 - Volume 28 - Issue 5 - p 601-607

tumor. Immunostainings are not helpful. Mesonephric ACs often present in early stage and have better prognosis than their müllerian counterparts. Surgery alone appears to be the treatment of choice. In contrast, MMMTs may present in advanced stage and are agressive tumors, similar to malignant mixed müllerian tumors.

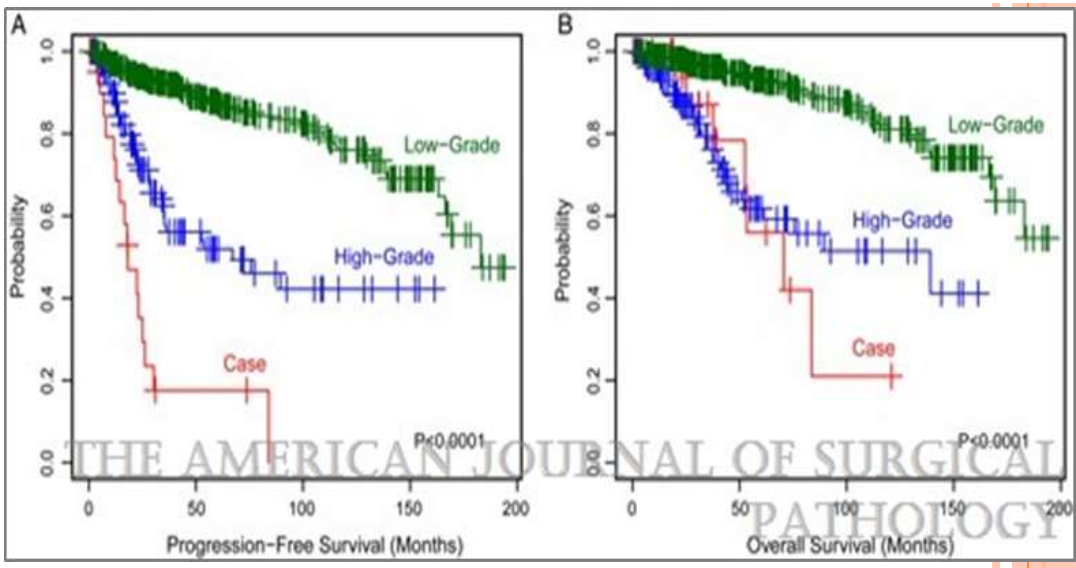
Mesonephric-like Carcinoma of the Endometrium

A Subset of Endometrial Carcinoma With an Aggressive Behavior

Euscher, Elizabeth D. MD*; Bassett, Roland†; Duose, Dzifa Y. PhD*; Lan, Chieh*; Wistuba, Ignacio MD*; Ramondetta, Lois MD‡; Ramalingam, Preetha MD*; Malpica, Anais MD* **Author Information**

The American Journal of Surgical Pathology: April 2020 - Volume 44 - Issue 4 - p 429-443
doi: 10.1097/PAS.0000000000001401

median progression-free survival for all patients was 169.3 months: 183 months for low-grade ECa, 67.1 months for USC and 18.2 months for MLCa. The median overall survival for cases of MLCa was 70.6 months and for USC was 139.1 months. The median survival for low-grade ECa was not reached. Univariate Cox regression



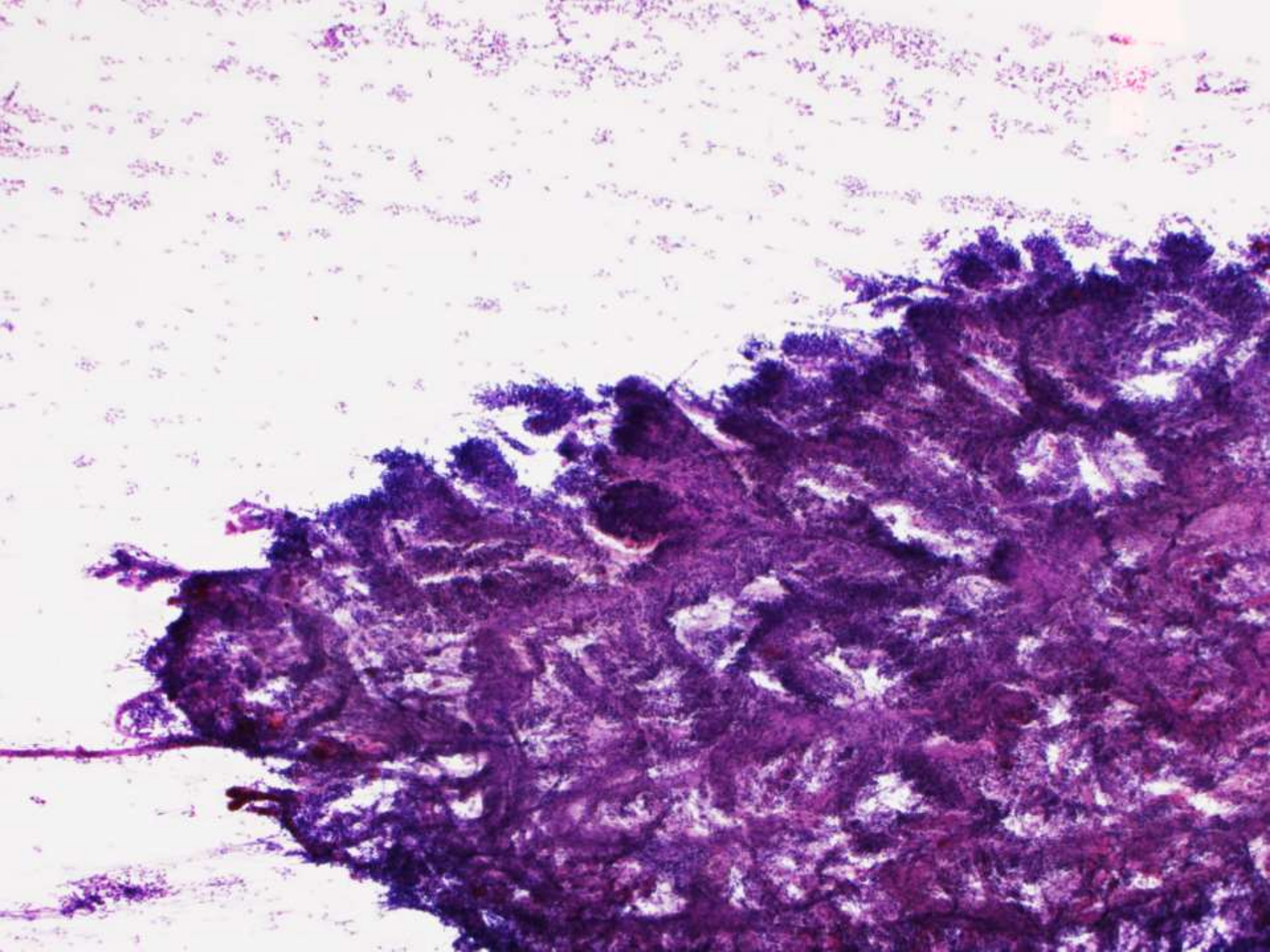
Take home points

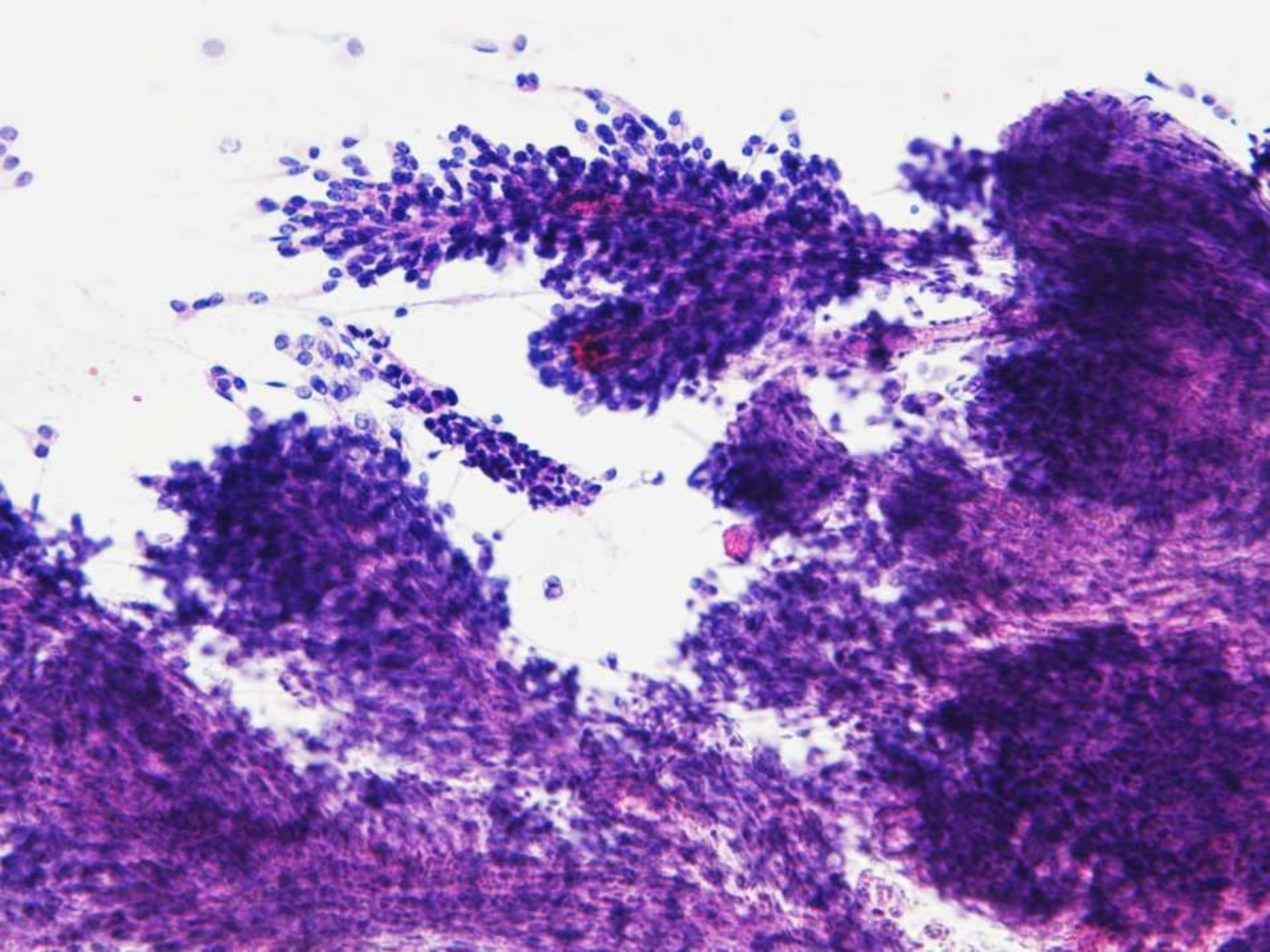
- Mesonephric-like carcinoma has many morphological patterns
- Immunohistochemistry can help guide
  - ER/PR negative
  - PAX-8 positive
  - P53 Wild-type
  - GATA-3/TTF-1: Inverse staining, DO BOTH!
    - GATA-3 CAN BE POSITIVE IN CARCINOSARCOMA
- Molecular mutations:
  - KRAS in majority
  - CTNNB1, TP53, PIK3CA in few
  - Despite CD117 IHC positivity, C-KIT mutations are not present
- Prognosis:
  - Recent studies show poor prognosis in endometrial mesonephric-like carcinoma

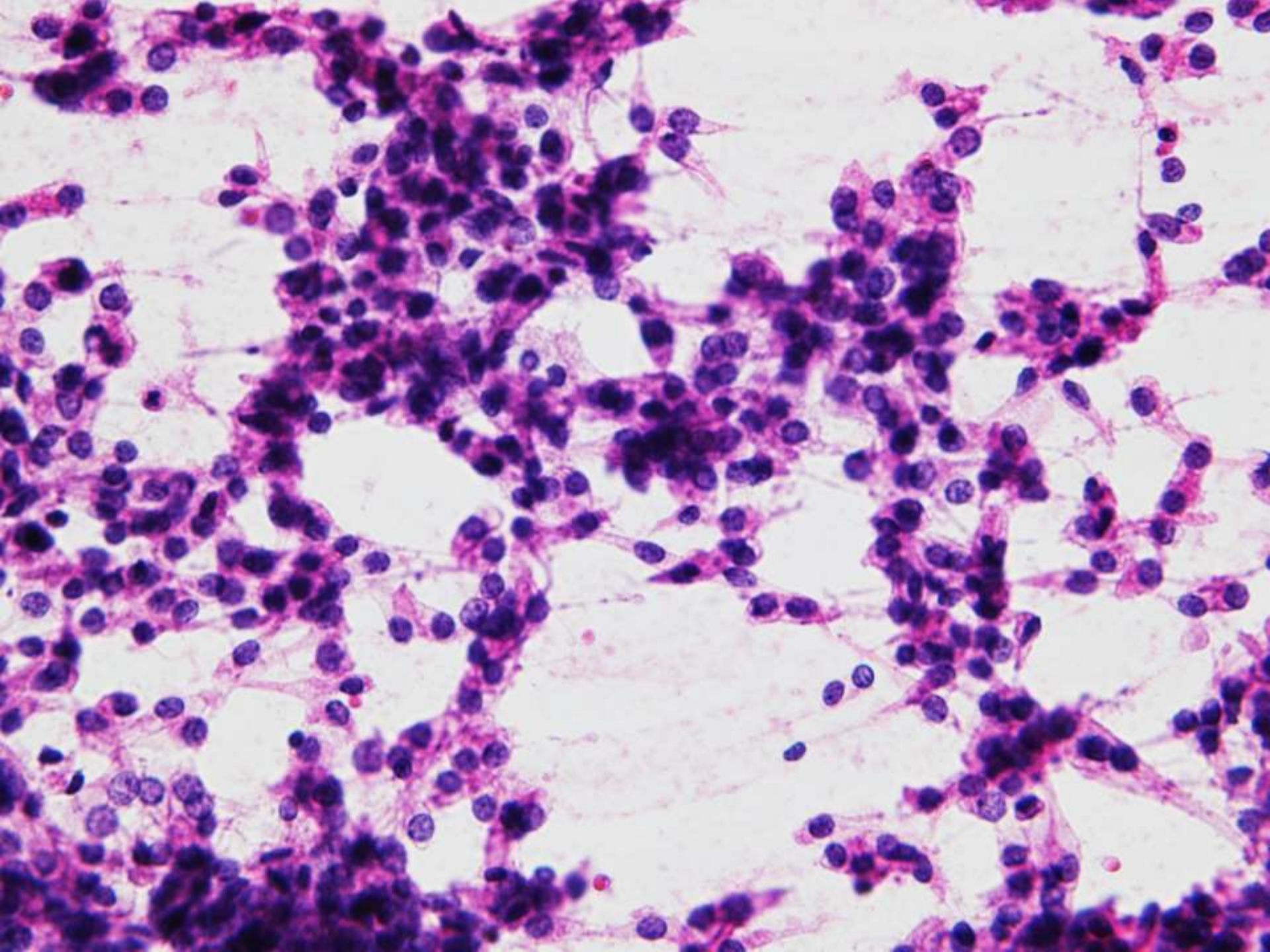
20-0407
SCANNED SLIDES AVAIL!

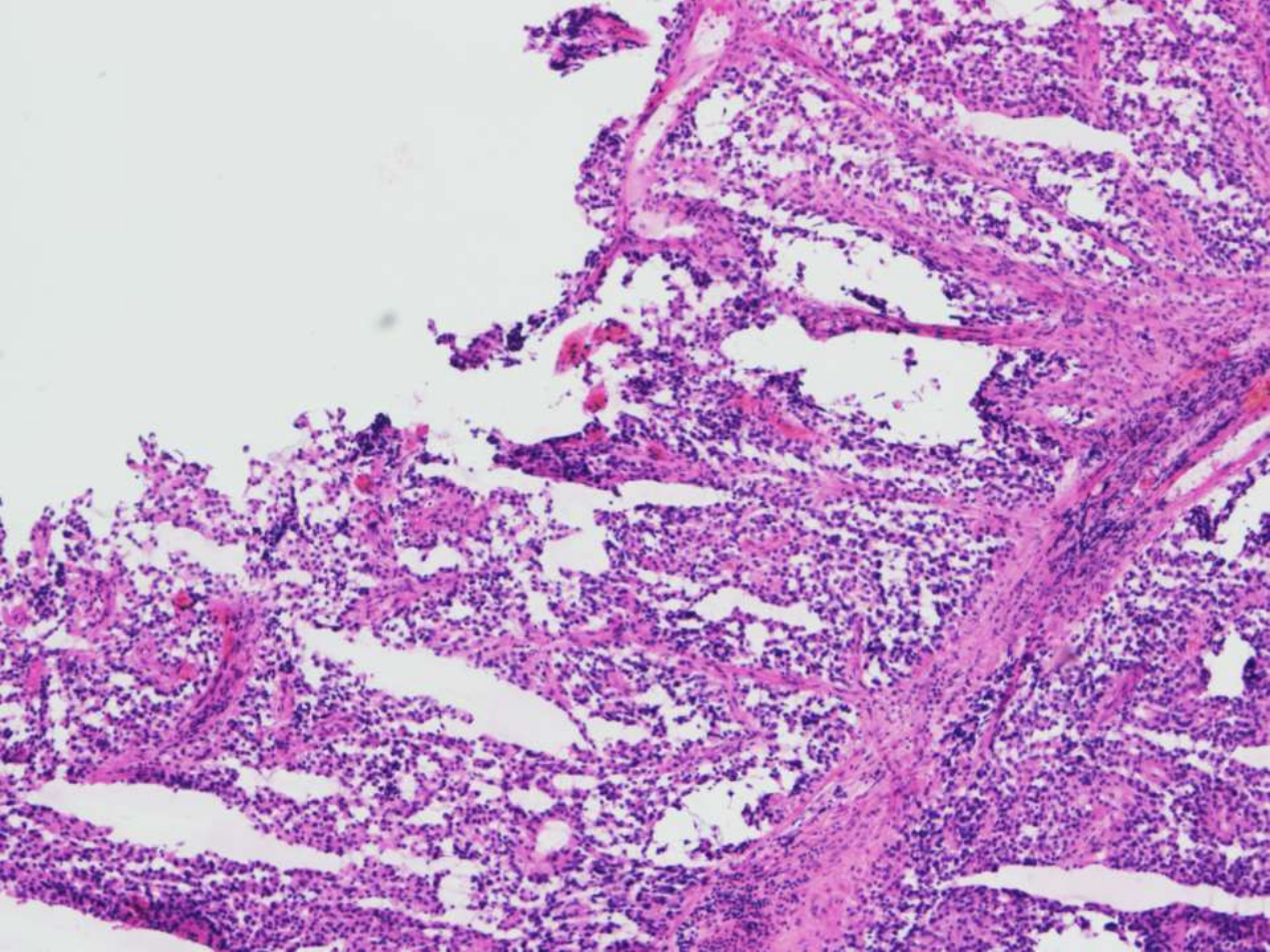
Romain Cayrol/Hannes Vogel; Stanford

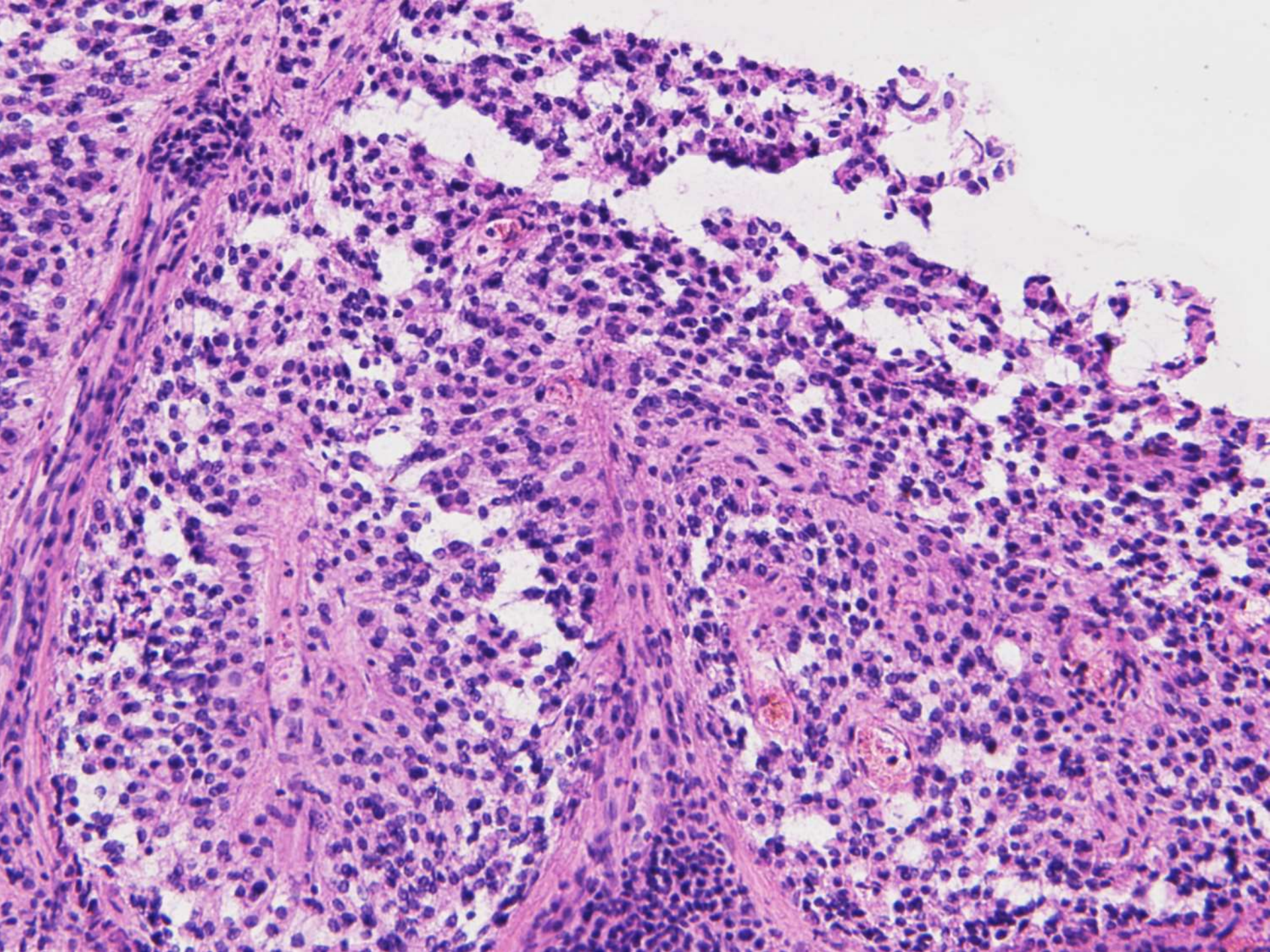
62-year-F with 7.5mm enhancing 4th venticular tumor, submitted for frozen section.

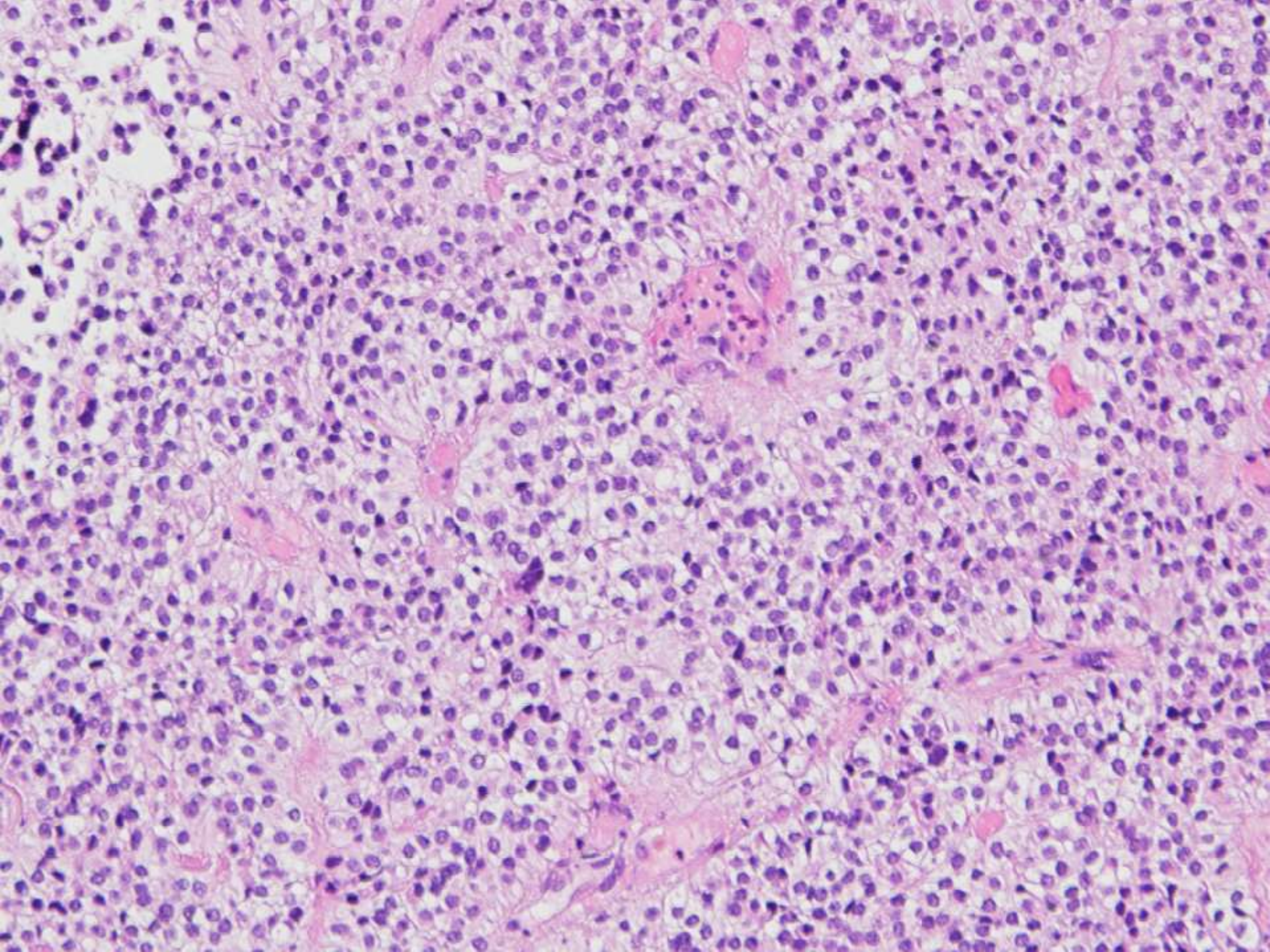


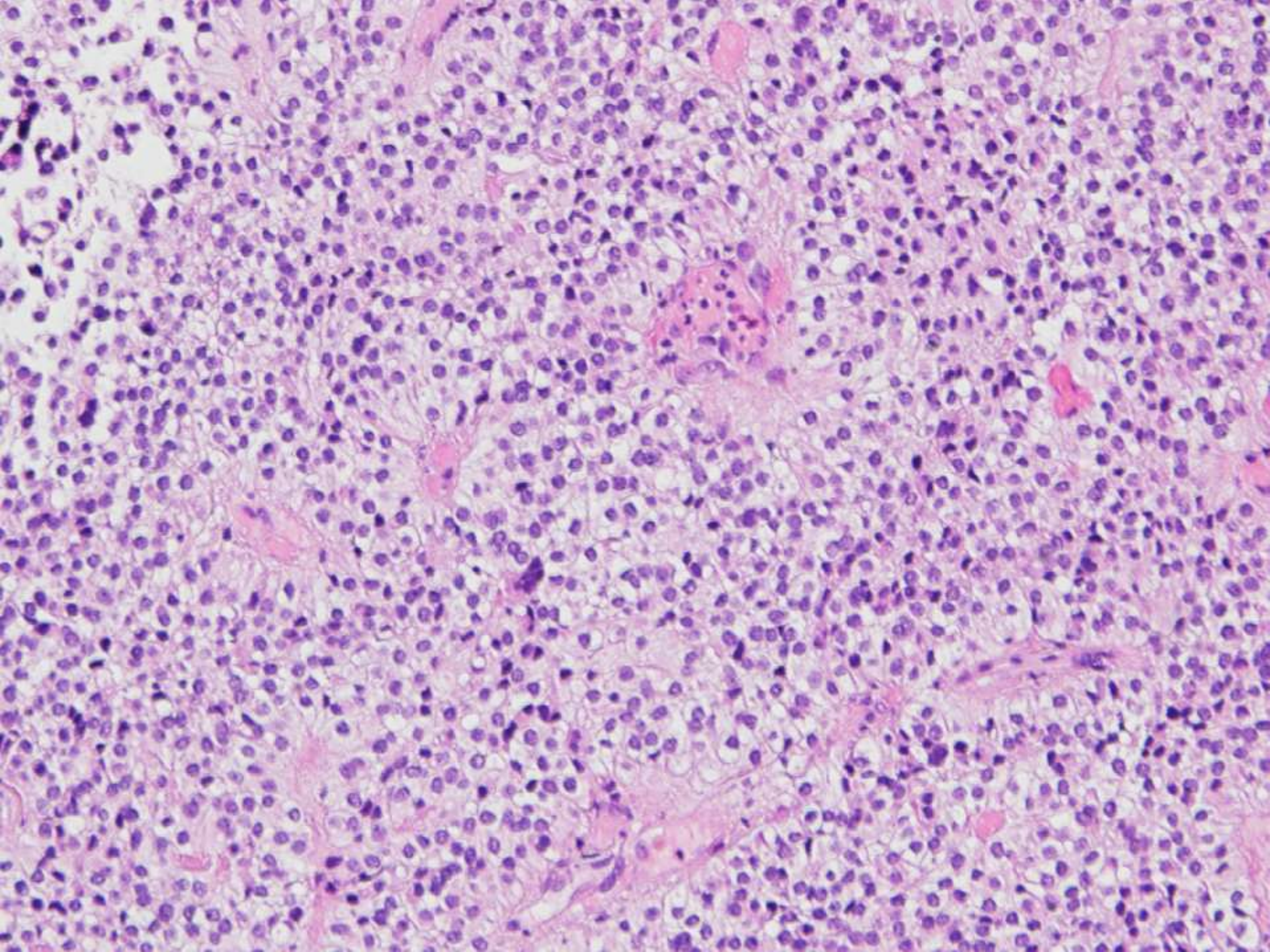


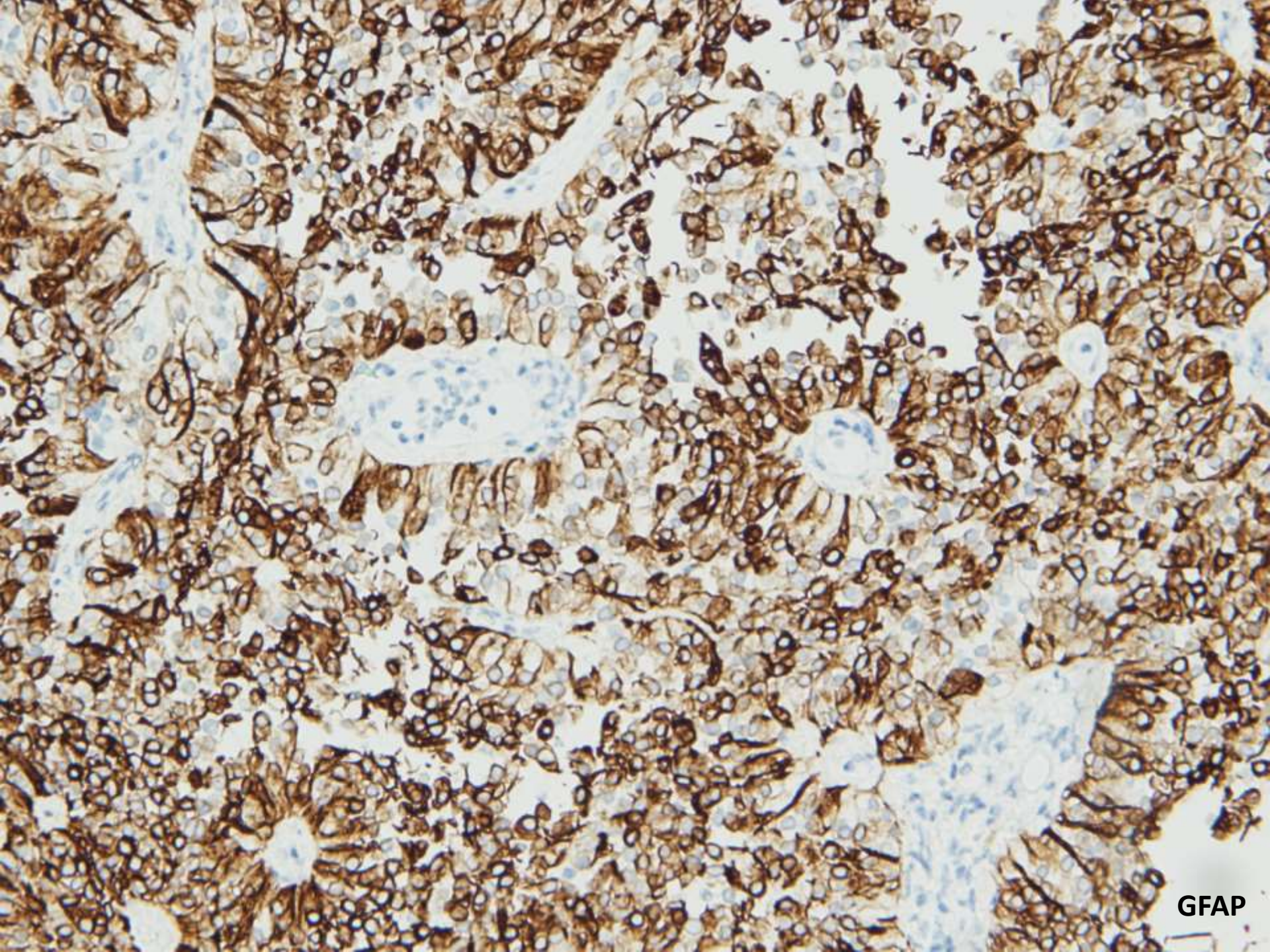


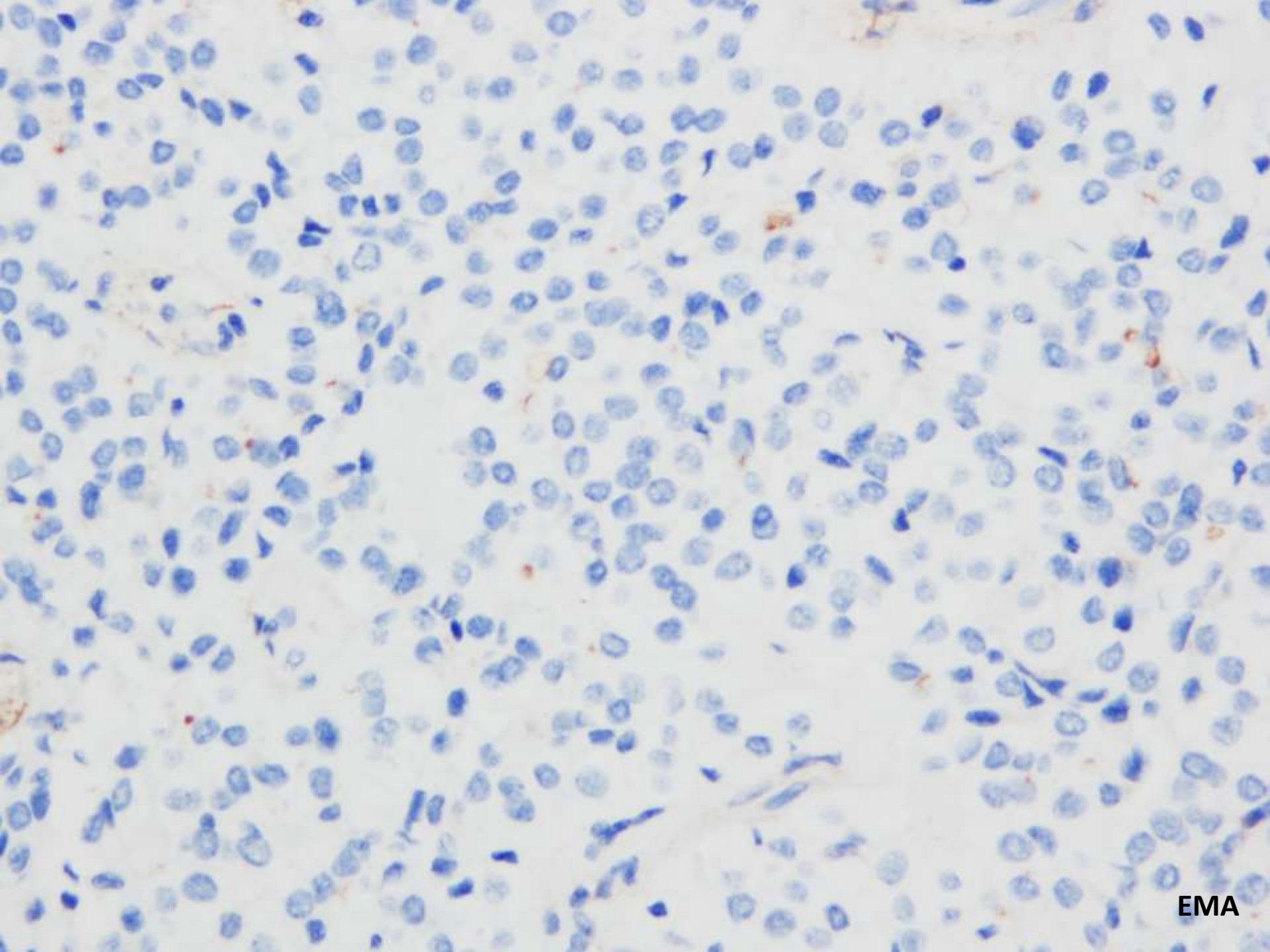









GFAP

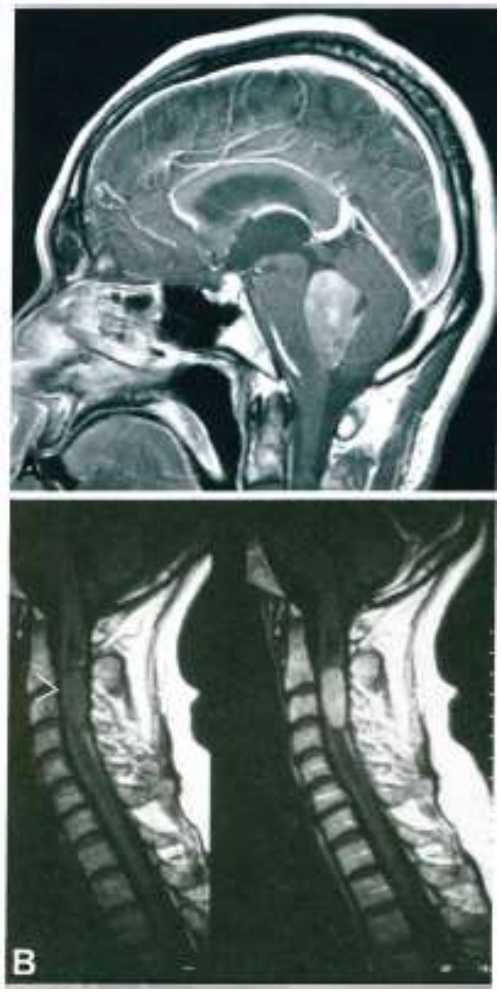

EMA

Diagnosis

- **BRAIN, RIGHT CEREBELLAR MEDULLARY TUMOR, RESECTION**
  - **EPENDYMOMA WITH CLEAR CELL FEATURES, WHO GRADE II**

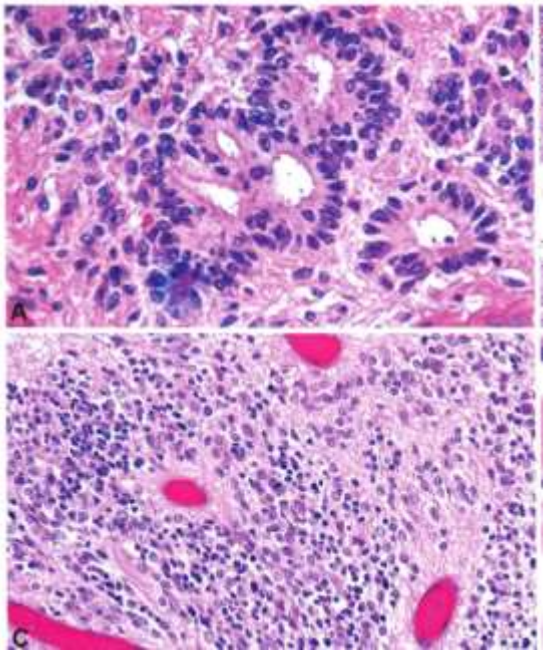
Ependymoma

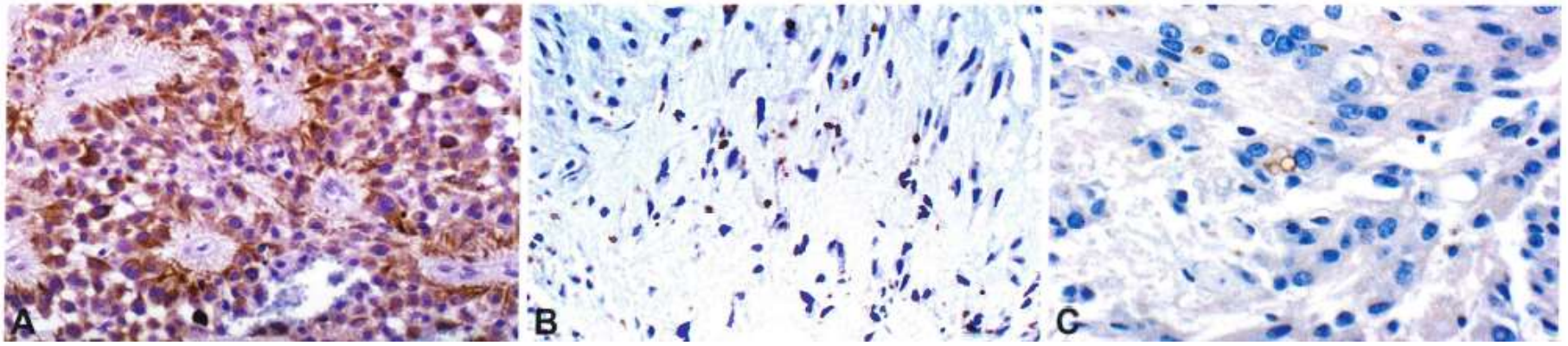
- 6.8% of neuroepithelial neoplasms
- 60% posterior fossa, 30% supratentorial, 10% spinal cord
- Posterior fossa are often pediatric, spinal often young adults (30-40s) and supratentorial affect both pediatric and adult patients
- Well circumscribed on imaging with various degree of enhancement, often secondary ventricular obstruction, +/- cystic, +/- calcifications, CSF spread



Ependymoma

- Derived from ependymal cells (cilia and microvilli on EM)
- Circumscribed glioma with uniform small cells characterized by perivascular anucleate zones (perivascular pseudo-rosettes), +/- ependymal rosettes with a central canal
- WHO grade II or grade III (increased mitoses, microvascular proliferation)
- GFAP positive, S100 positive, EMA in most cases dots and rings



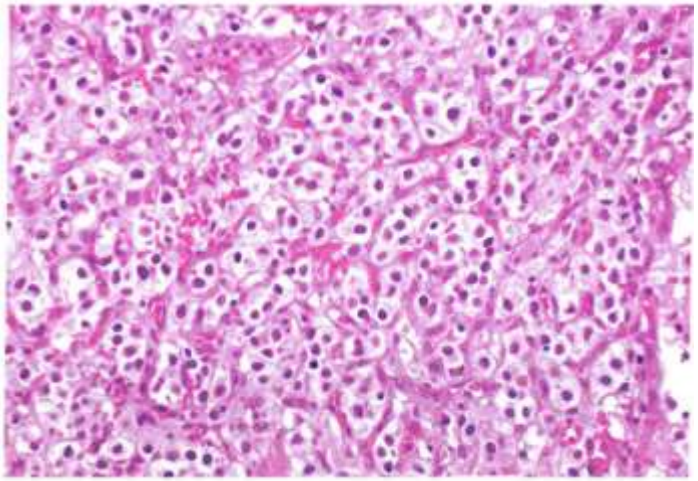


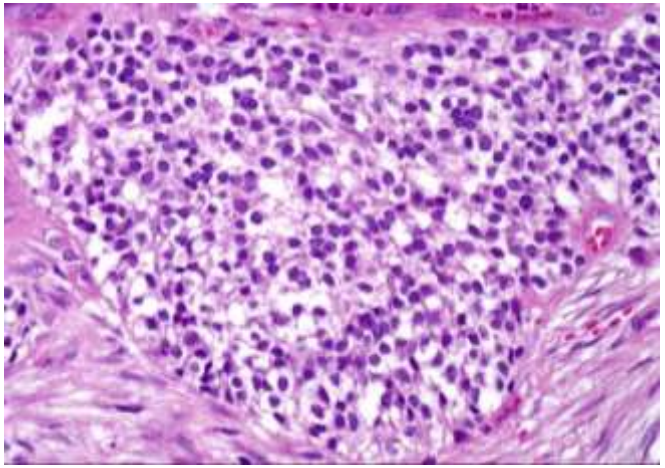
The 2016 World Health Organization Classification of Tumors of the Central Nervous System. Louis DN et al. 2016

Ependymoma variants

- Histologic variants
  - Papillary, tanycytic and clear cell

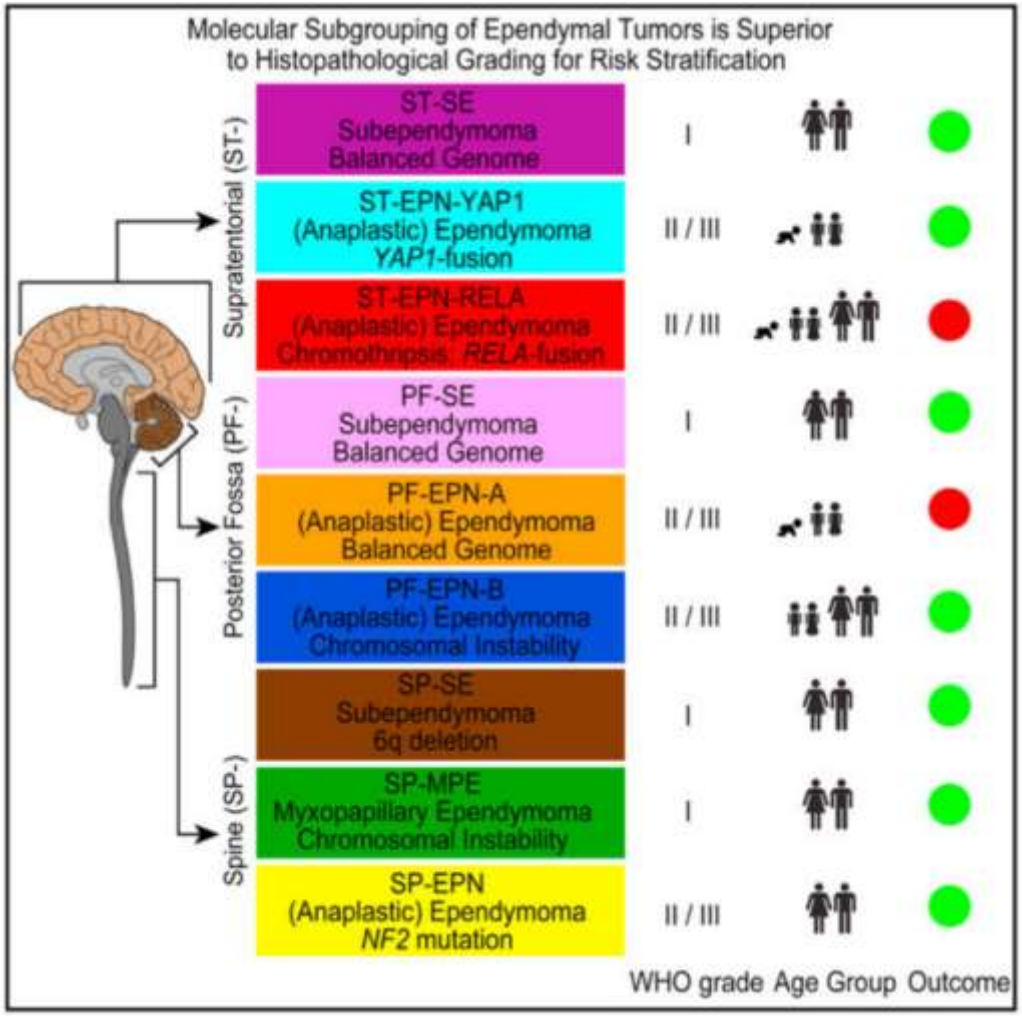
- Clear cell variant
  - Oligodendroglial phenotype with perinuclear halo
  - Mostly supratentorial in the pediatric population
  - Some reports describe a more aggressive clinical course compared to other histologic variants
  - Differential diagnosis includes oligodendroglioma, central neurocytoma, clear cell renal carcinoma, hemangioblastoma, choroid plexus neoplasm





Ependymoma molecular variants

- Molecular subtypes
  - 9 different ependymoma subgroups
  - Prognostic implication (age, location, molecular subgroup and *grade*)
  - Immunohistochemical and FISH studies can be used to subtype ependymomas



Molecular Classification of Ependymal Tumors across All CNS Compartments, Histopathological Grades, and Age Groups. Pajtler KW, et al. Cancer Cell. 2015 May 11;27(5):728-43

Conclusions

- Clear cell ependymoma is a histologic variant of ependymomas characterized by perinuclear halos and rare rosettes/pseudo-rosettes
  - Mostly supratentorial in the pediatric population
  - No definite prognostic implication but some reports of a more aggressive clinical course
  - Major prognostic factors for ependymomas: age, location, molecular subtype, *grade*
  - Differential diagnosis includes oligodendroglioma, central neurocytoma, clear cell renal carcinoma, hemangioblastoma, choroid plexus neoplasm

References

- The 2016 World Health Organization Classification of Tumors of the Central Nervous System: a summary. Louis DN, Perry A, Reifenberger G, von Deimling A, Figarella-Branger D, Cavenee WK, Ohgaki H, Wiestler OD, Kleihues P, Ellison DW. Acta Neuropathol. 2016 Jun;131(6):803-20

- Molecular Classification of Ependymal Tumors across All CNS Compartments, Histopathological Grades, and Age Groups. Pajtler KW, Witt H, Sill M, Jones DT, Hovestadt V, Kratochwil F, Wani K, Tatevossian R, Punchihewa C, Johann P, Reimand J, Warnatz HJ, Ryzhova M, Mack S, Ramaswamy V, Capper D, Schweizer L, Sieber L, Wittmann A, Huang Z, van Sluis P, Volckmann R, Koster J, Versteeg R, Fults D, Toledano H, Avigad S, Hoffman LM, Donson AM, Foreman N, Hewer E, Zitterbart K, Gilbert M, Armstrong TS, Gupta N, Allen JC, Karajannis MA, Zagzag D, Hasselblatt M, Kulozik AE, Witt O, Collins VP, von Hoff K, Rutkowski S, Pietsch T, Bader G, Yaspo ML, von Deimling A, Lichter P, Taylor MD, Gilbertson R, Ellison DW, Aldape K, Korshunov A, Kool M, Pfister SM. Cancer Cell. 2015 May 11;27(5):728-43

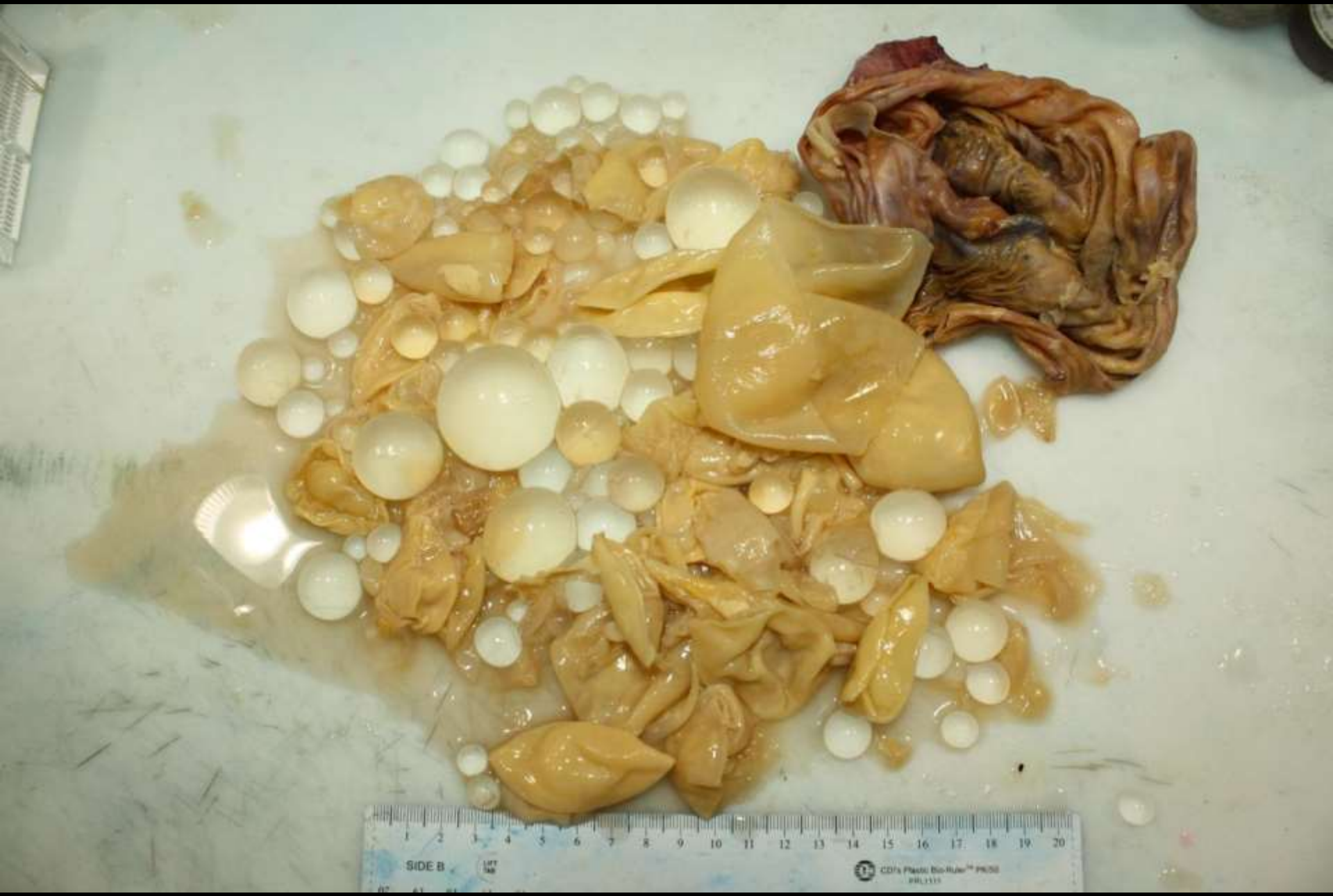
- Surgical resection of fourth ventricular ependymomas: case series and technical nuances. E Winker *et al*. *J. Neurooncology* 2016; 130:341-349.

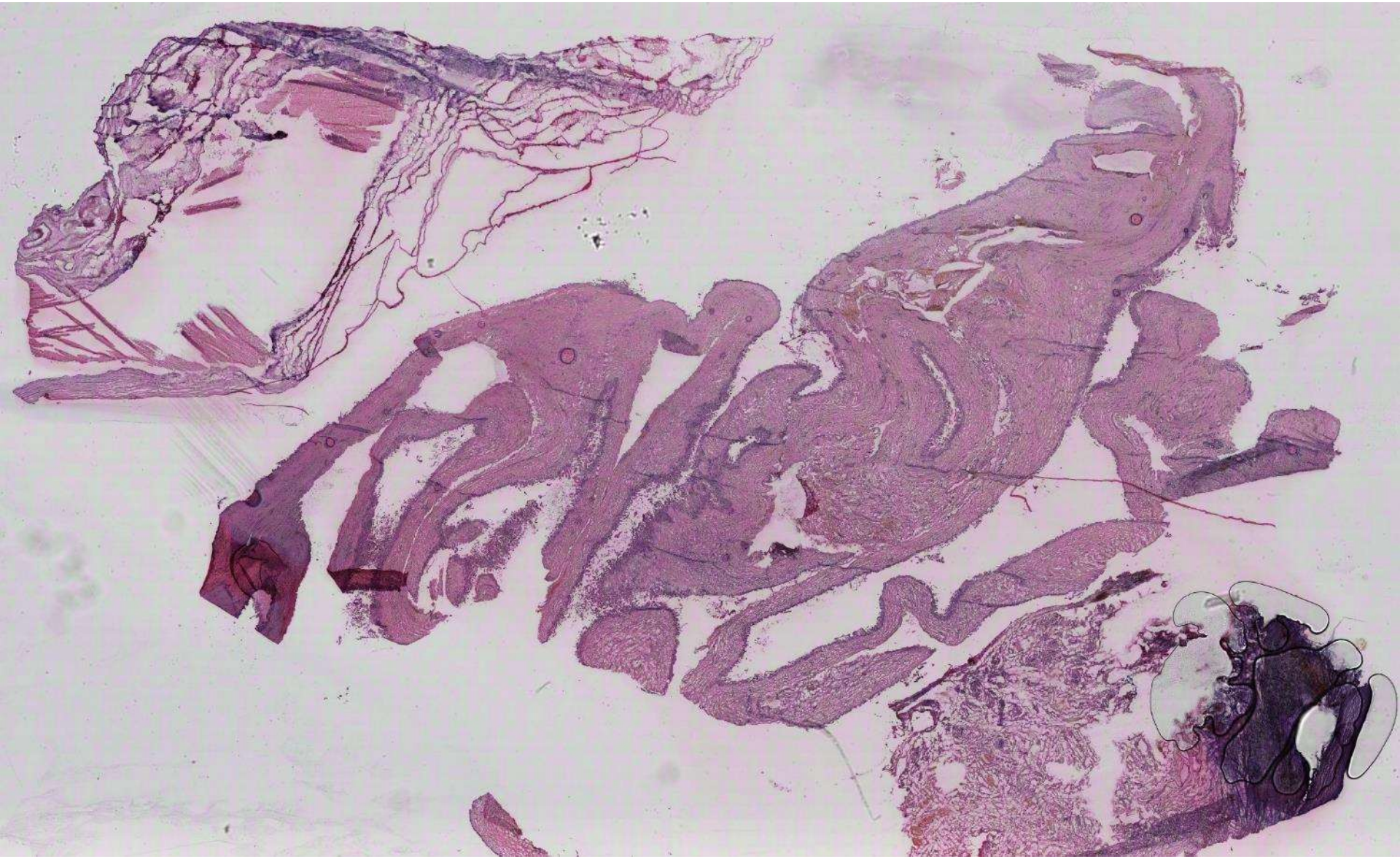
- Clear Cell Ependymoma: A Clinicopathologic and Radiographic Analysis of 10 Patients. M Fouladi, *et al*. *Cancer* 2003; 98(10): 2232.

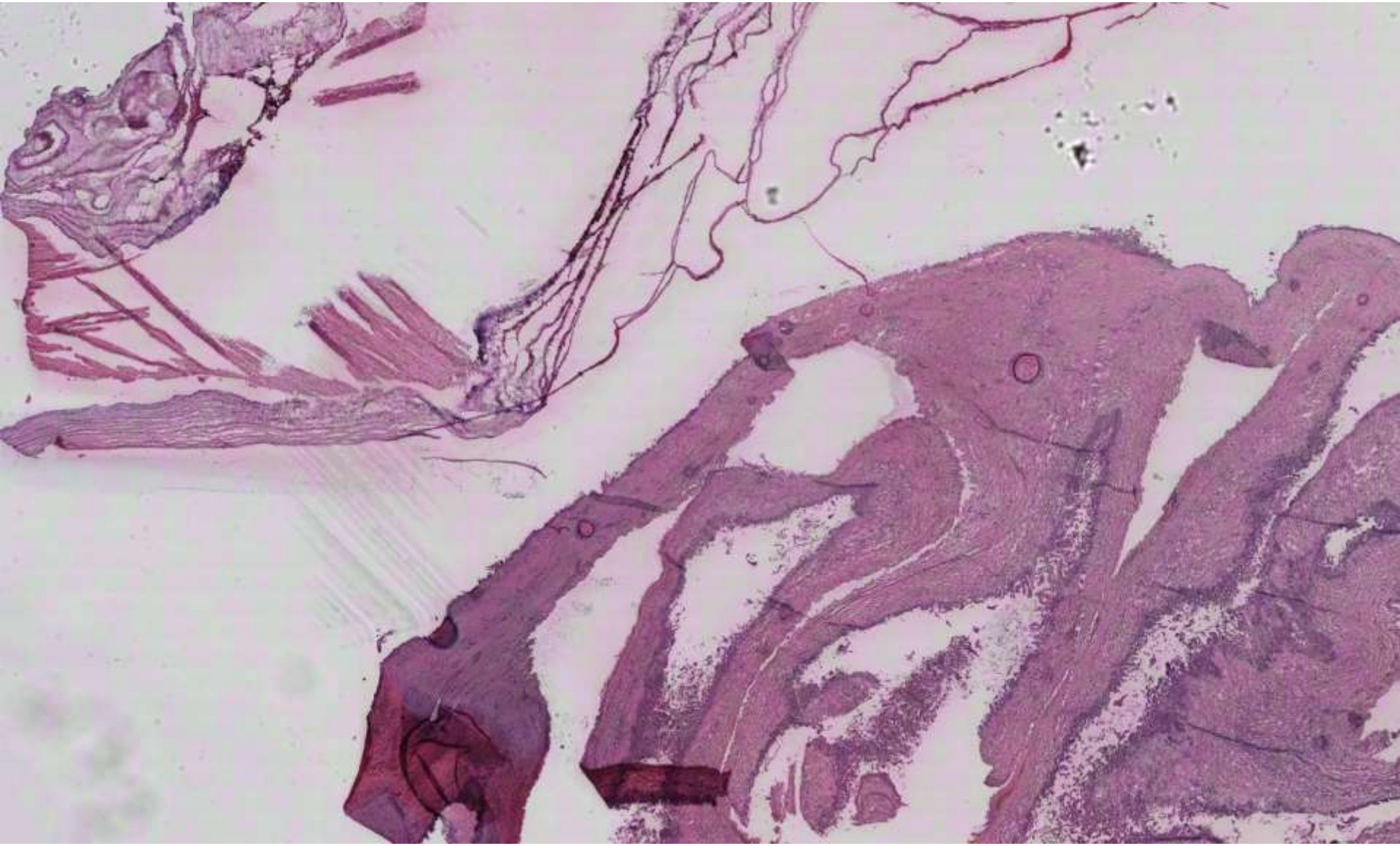
20-0408
SCANNED SLIDE AVAIL!

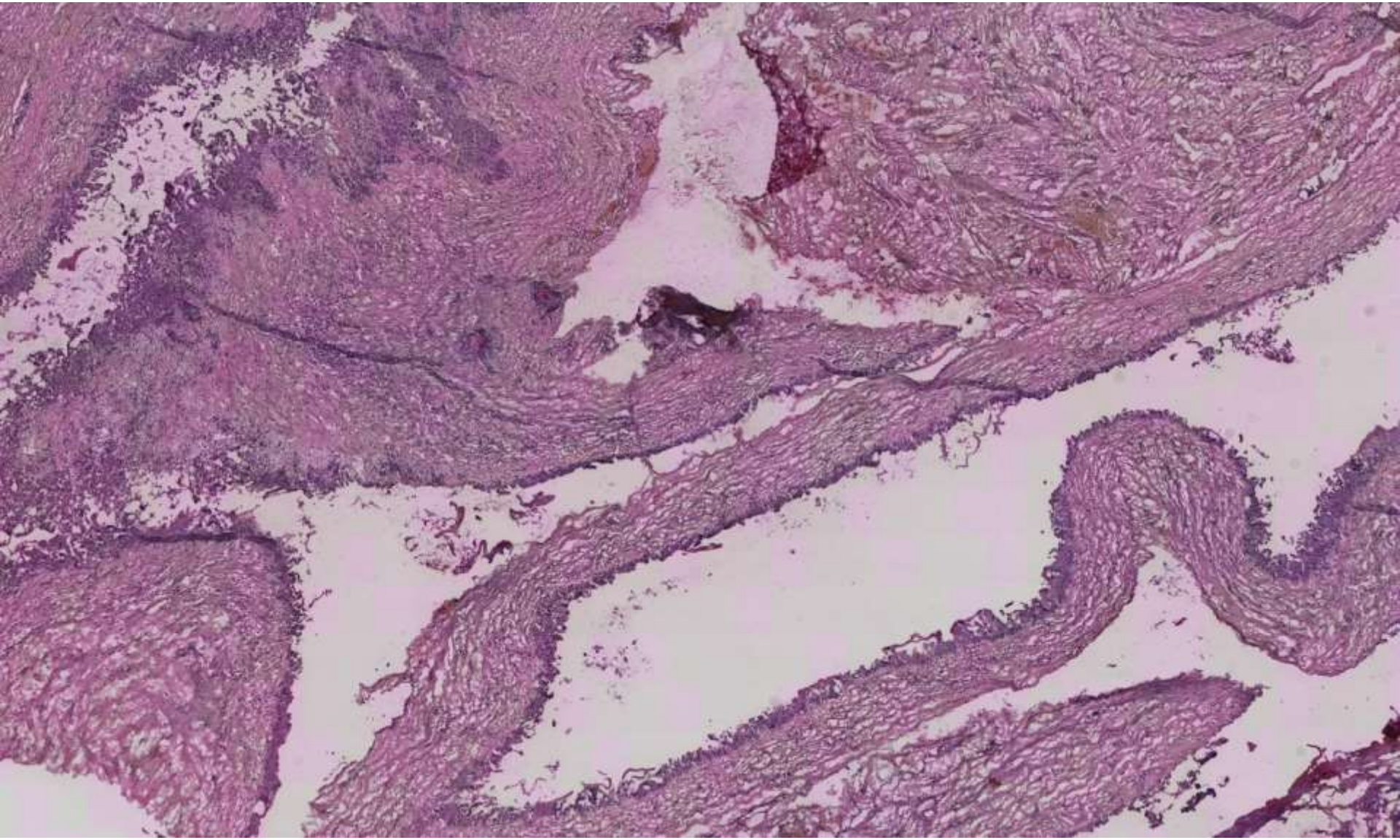
Ankur Sangoi; El Camino Hospital

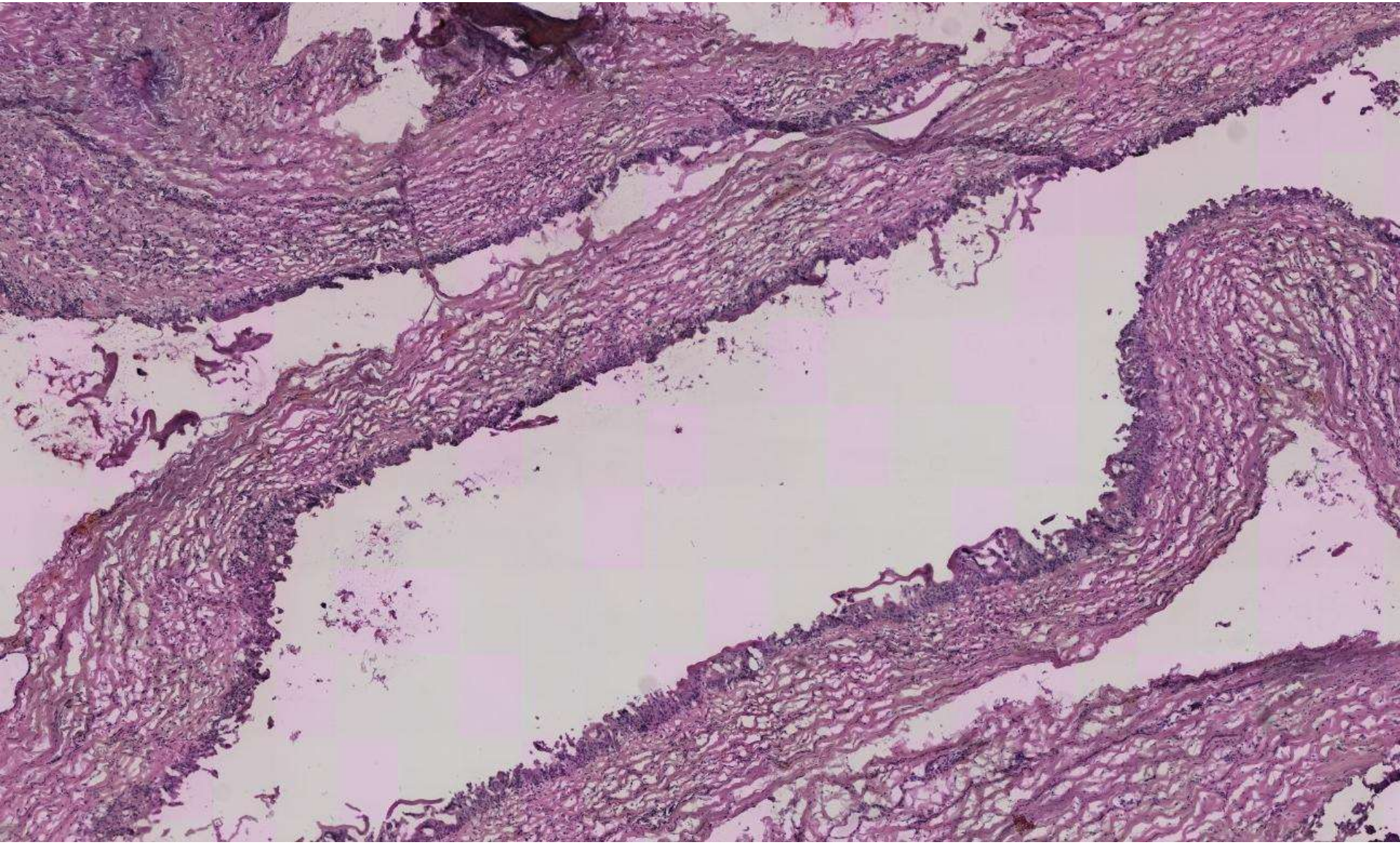
22-year-old F with 26cm right adnexal mass composed of coalescing fluid-filled cysts, submitted for frozen section.

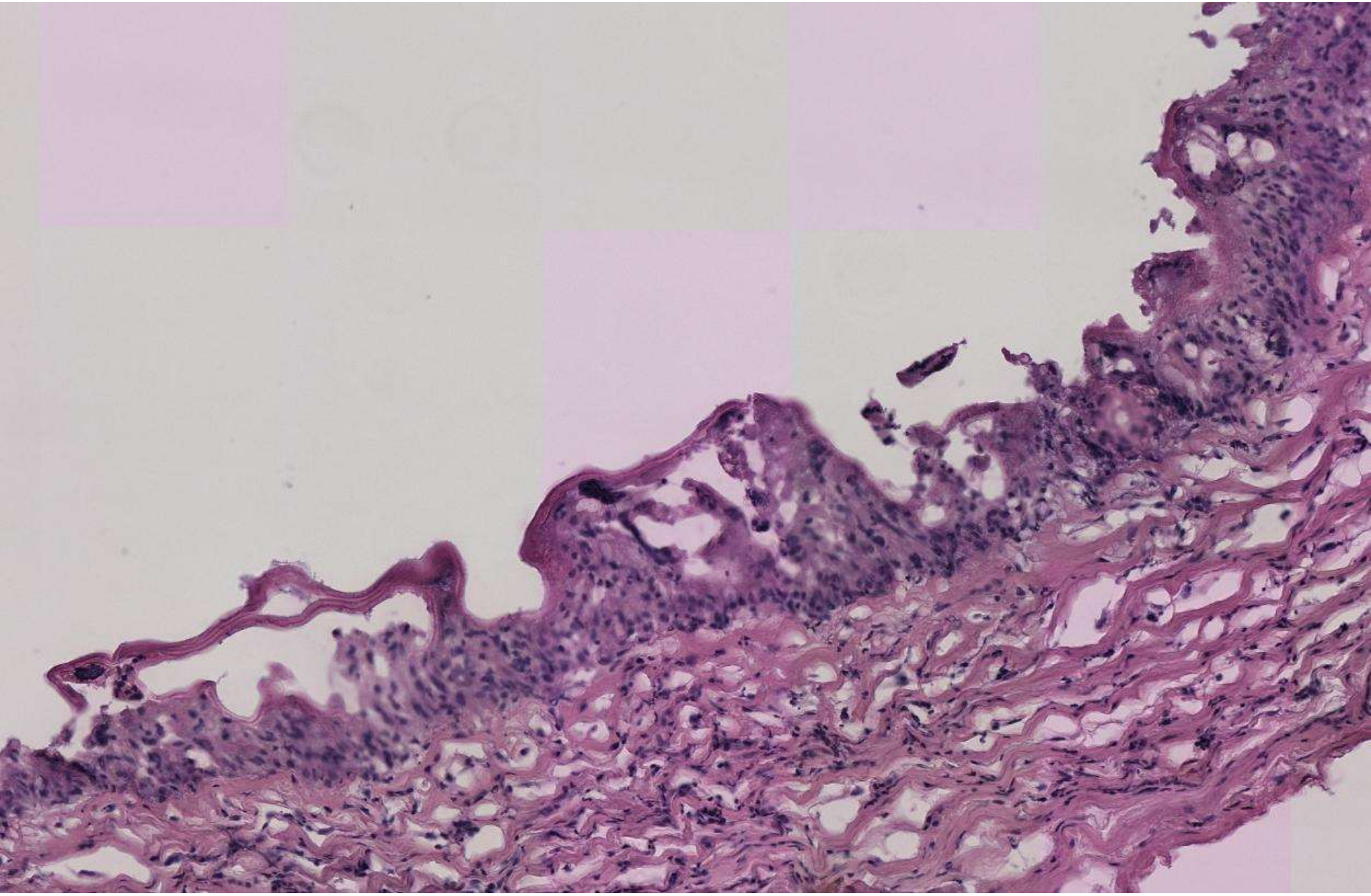

SIDE B

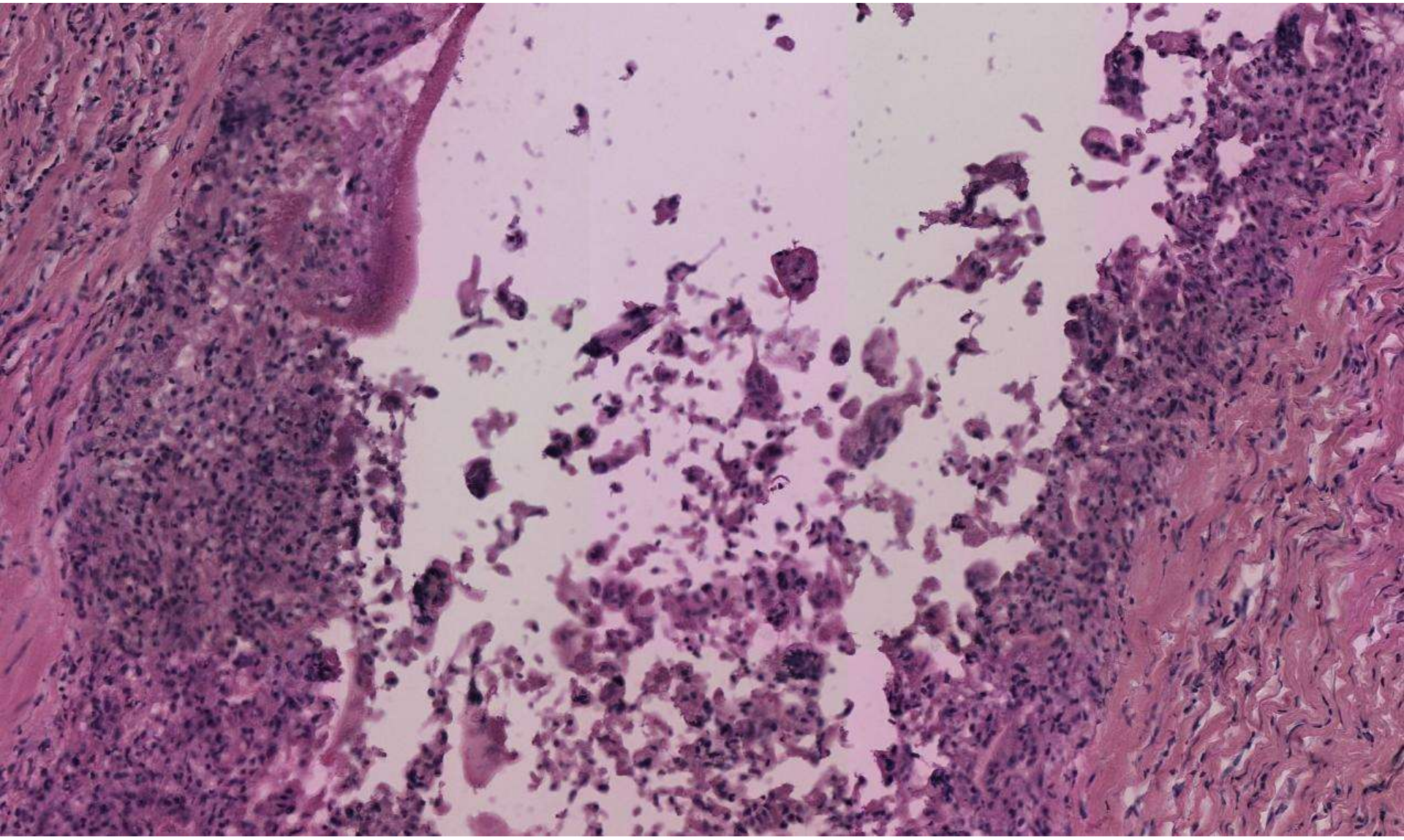


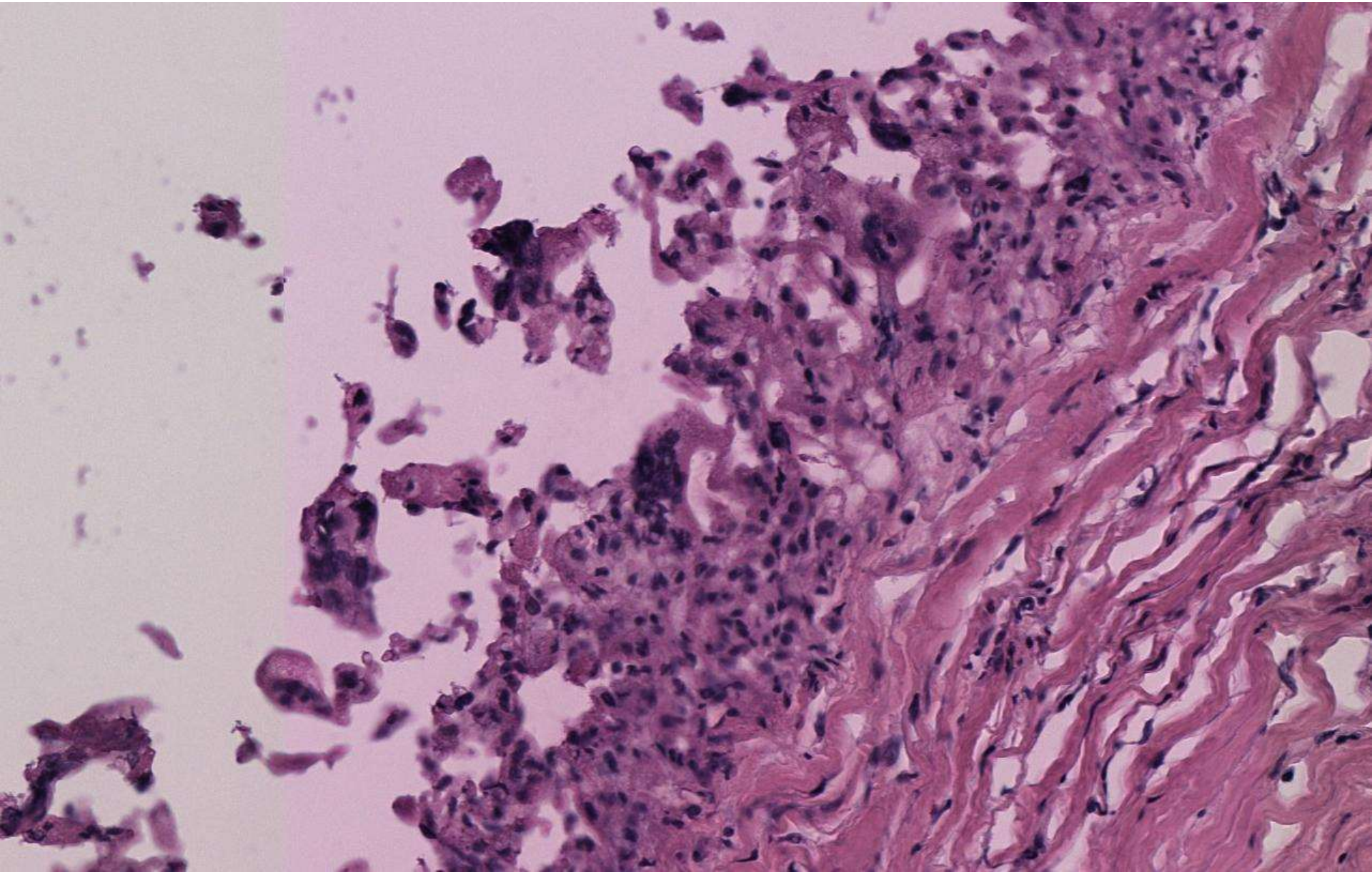


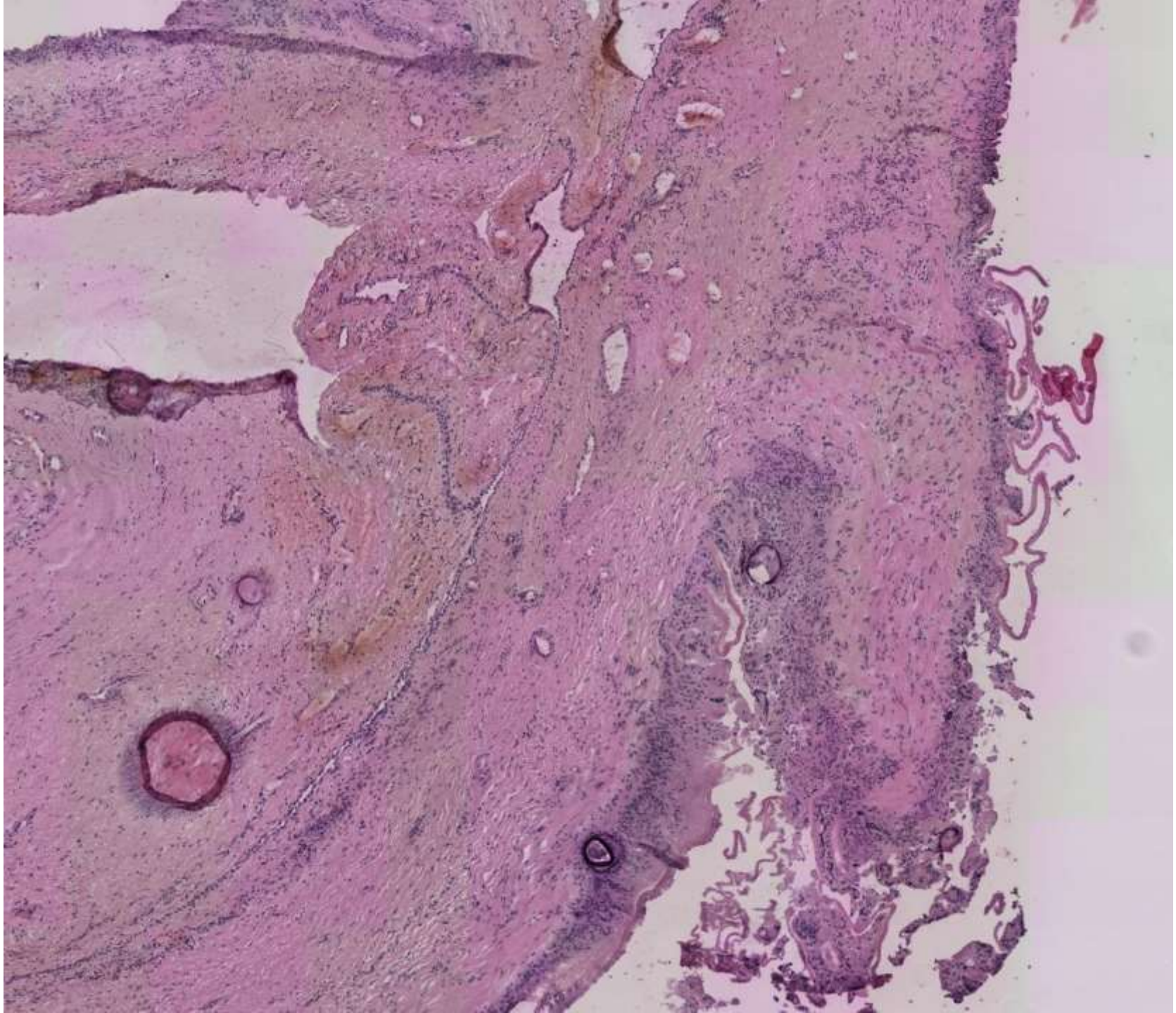


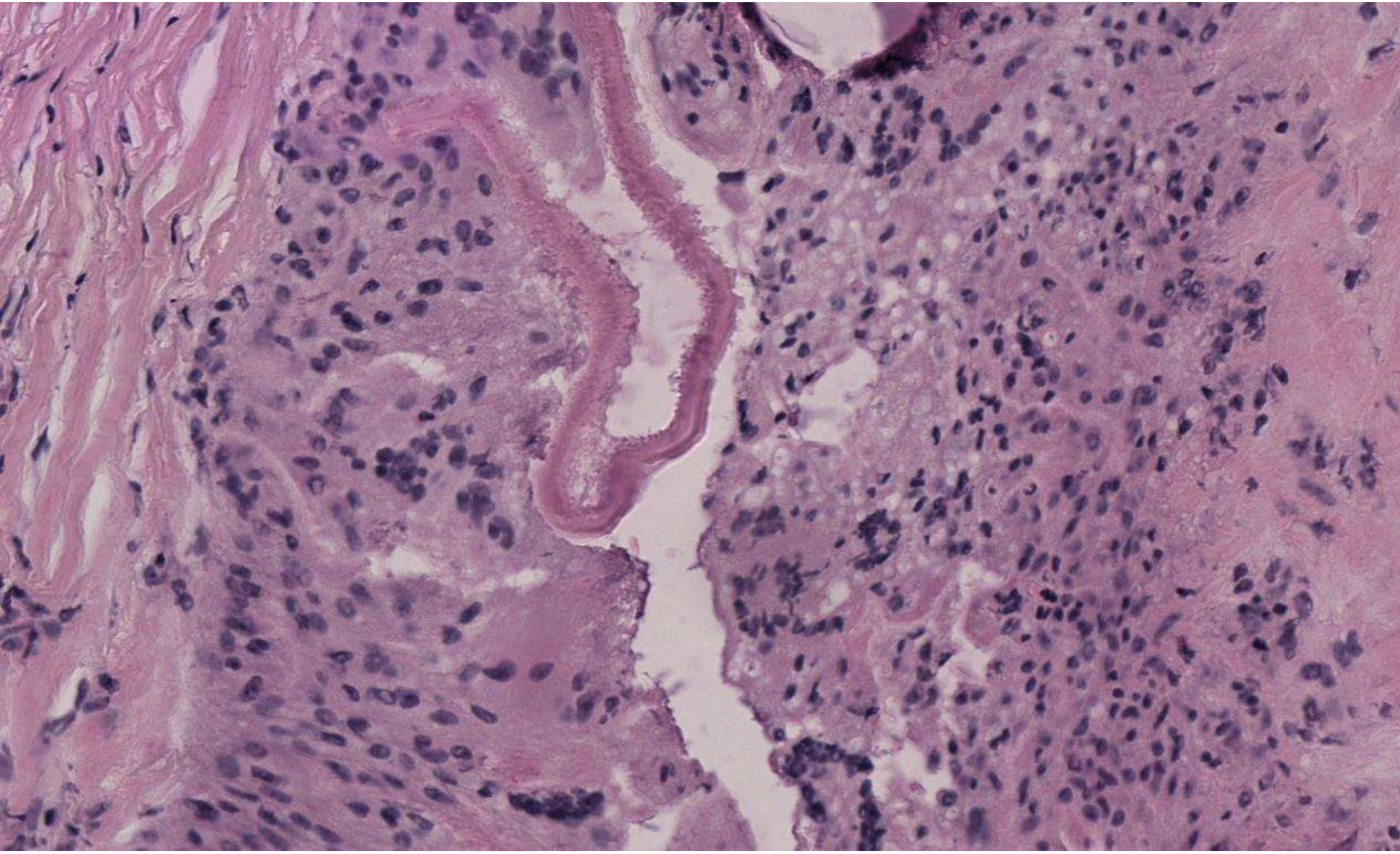










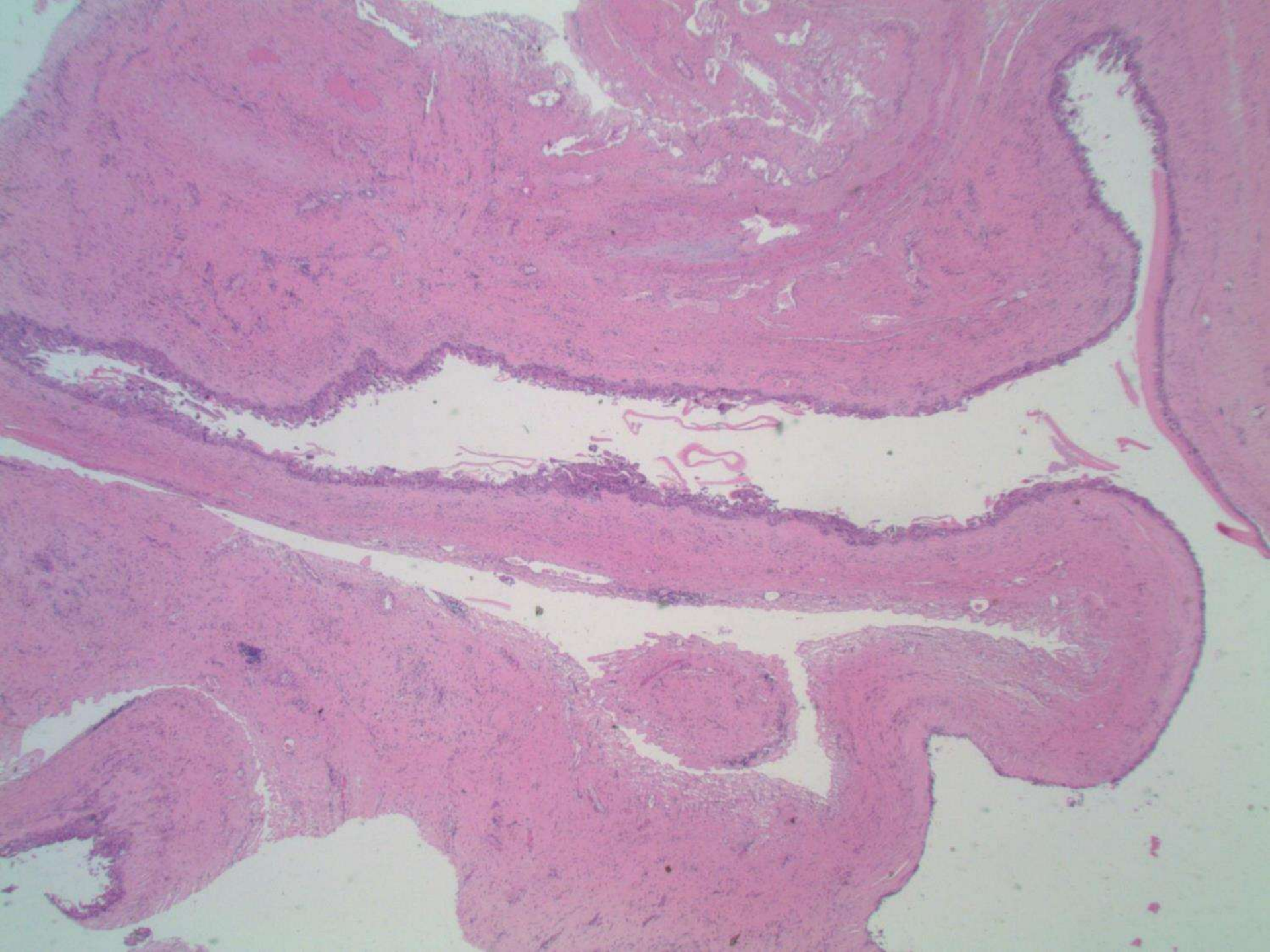


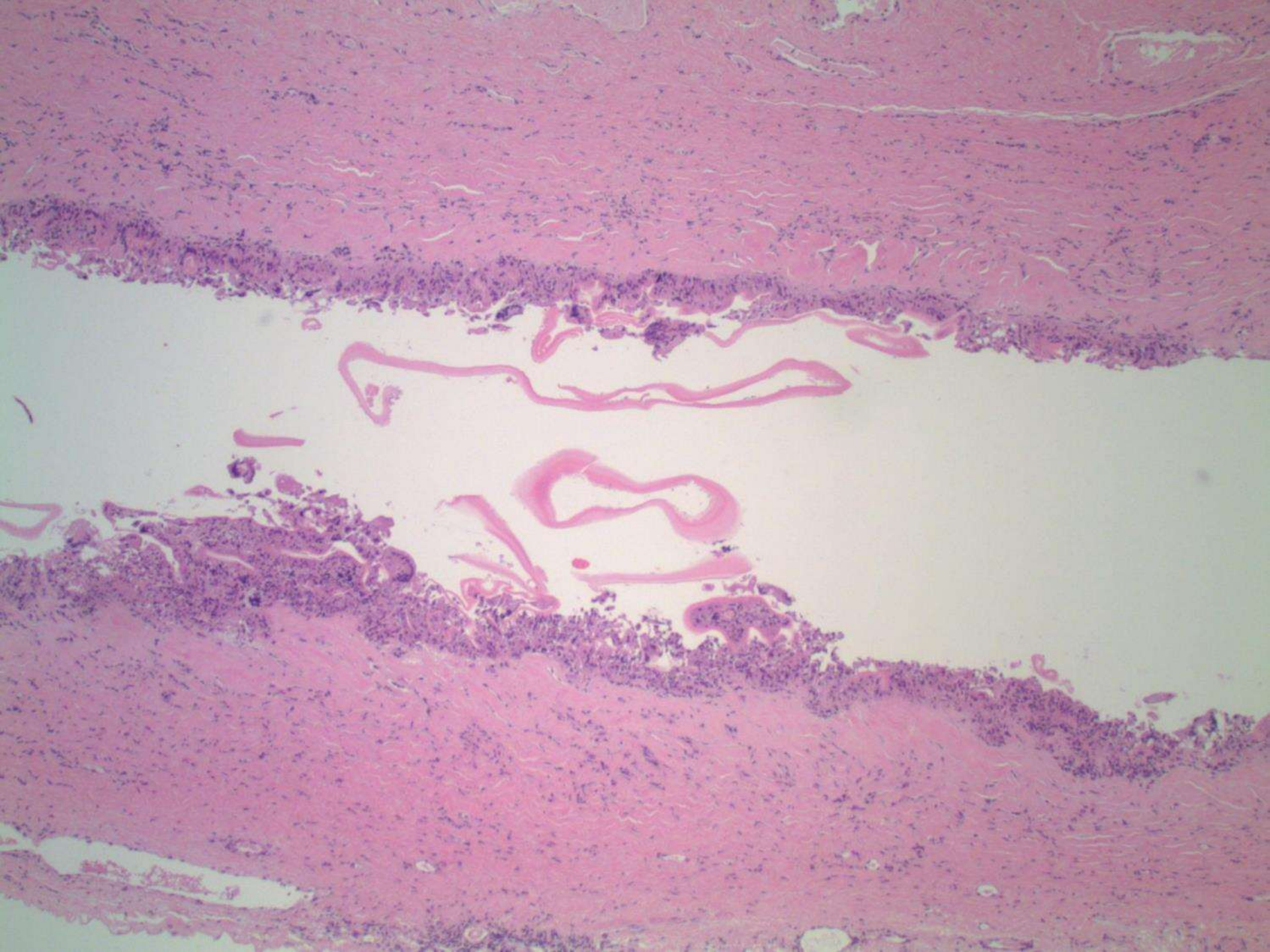
DDx

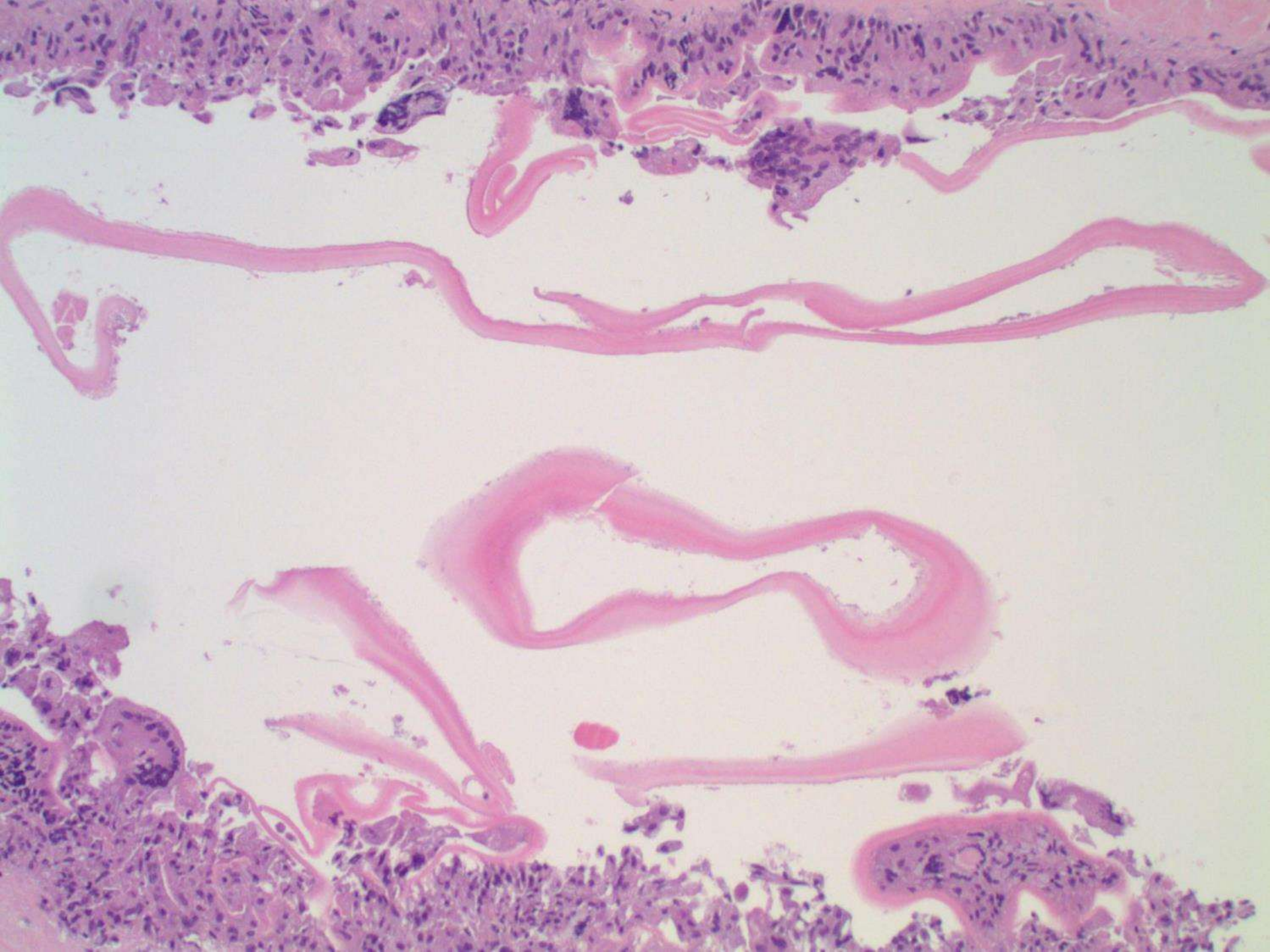
- Peritoneal inclusion/mesothelial cyst
- Inflammatory pseudocyst
- Parasitic cyst
- Mucinous cystadenoma
- Serous cystadenoma

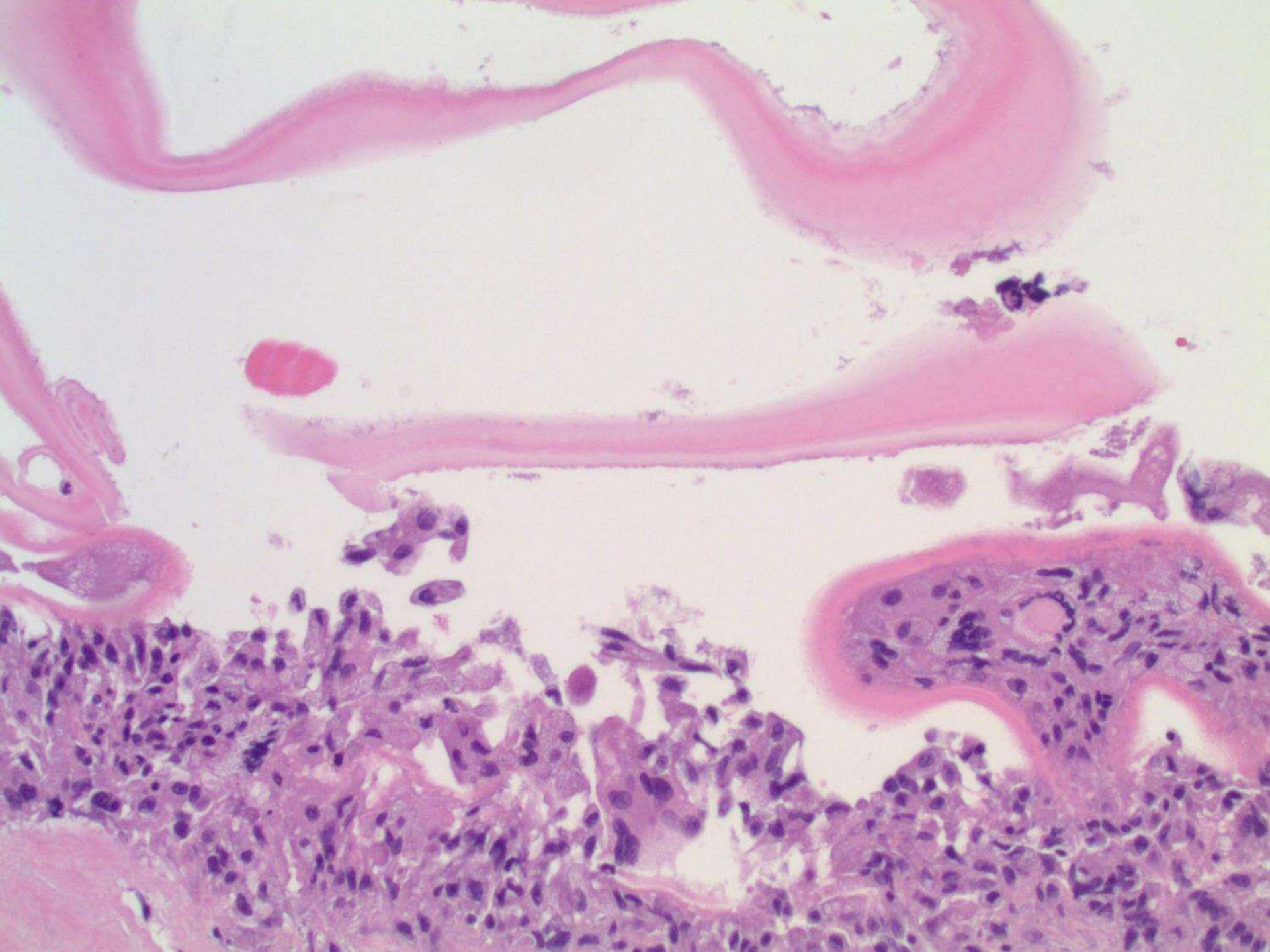
DDx
(after seeing gross image)

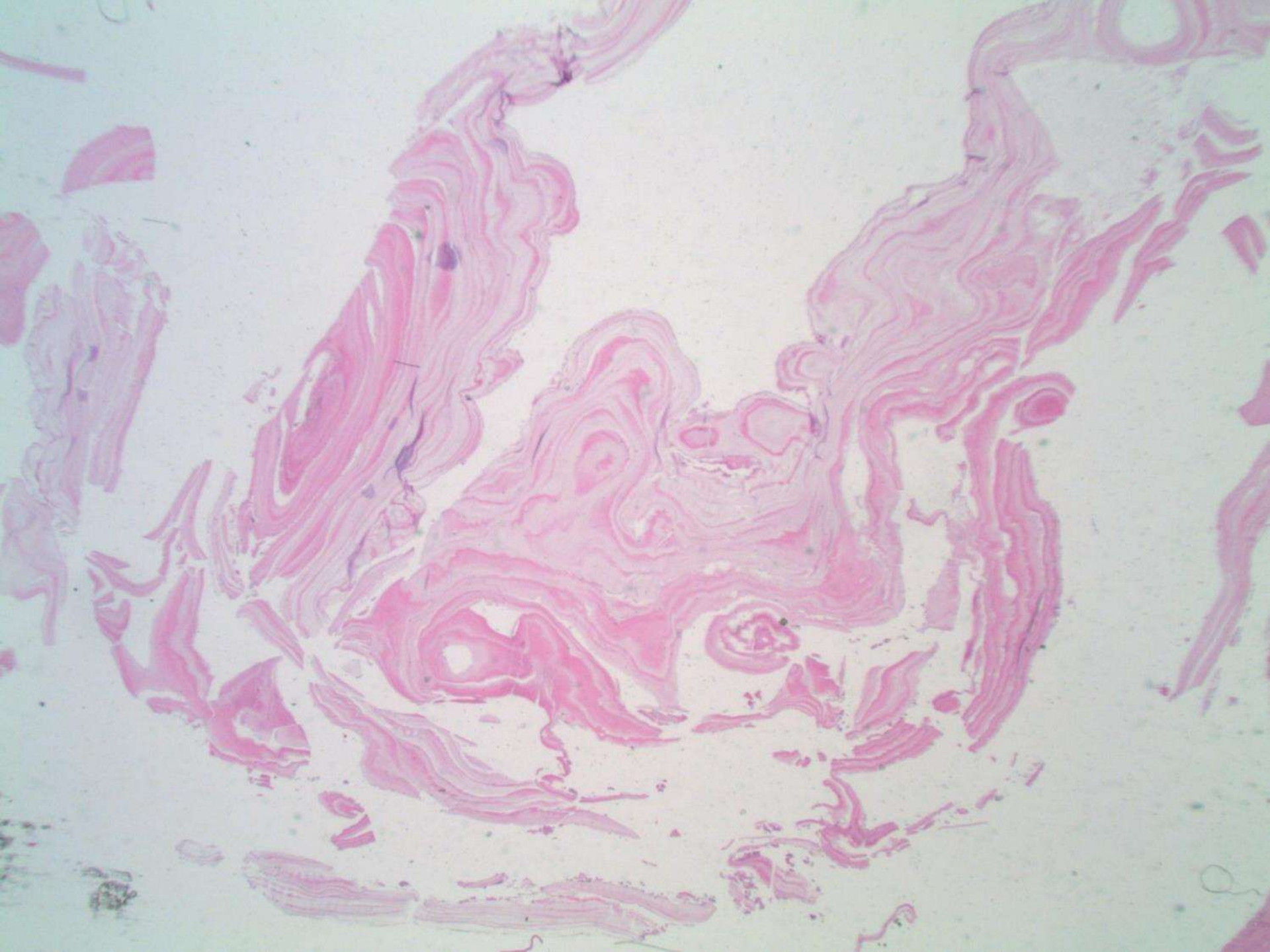
- Peritoneal inclusion/mesothelial cyst
- Inflammatory pseudocyst
- Parasitic cyst
- Mucinous cystadenoma
- Serous cystadenoma

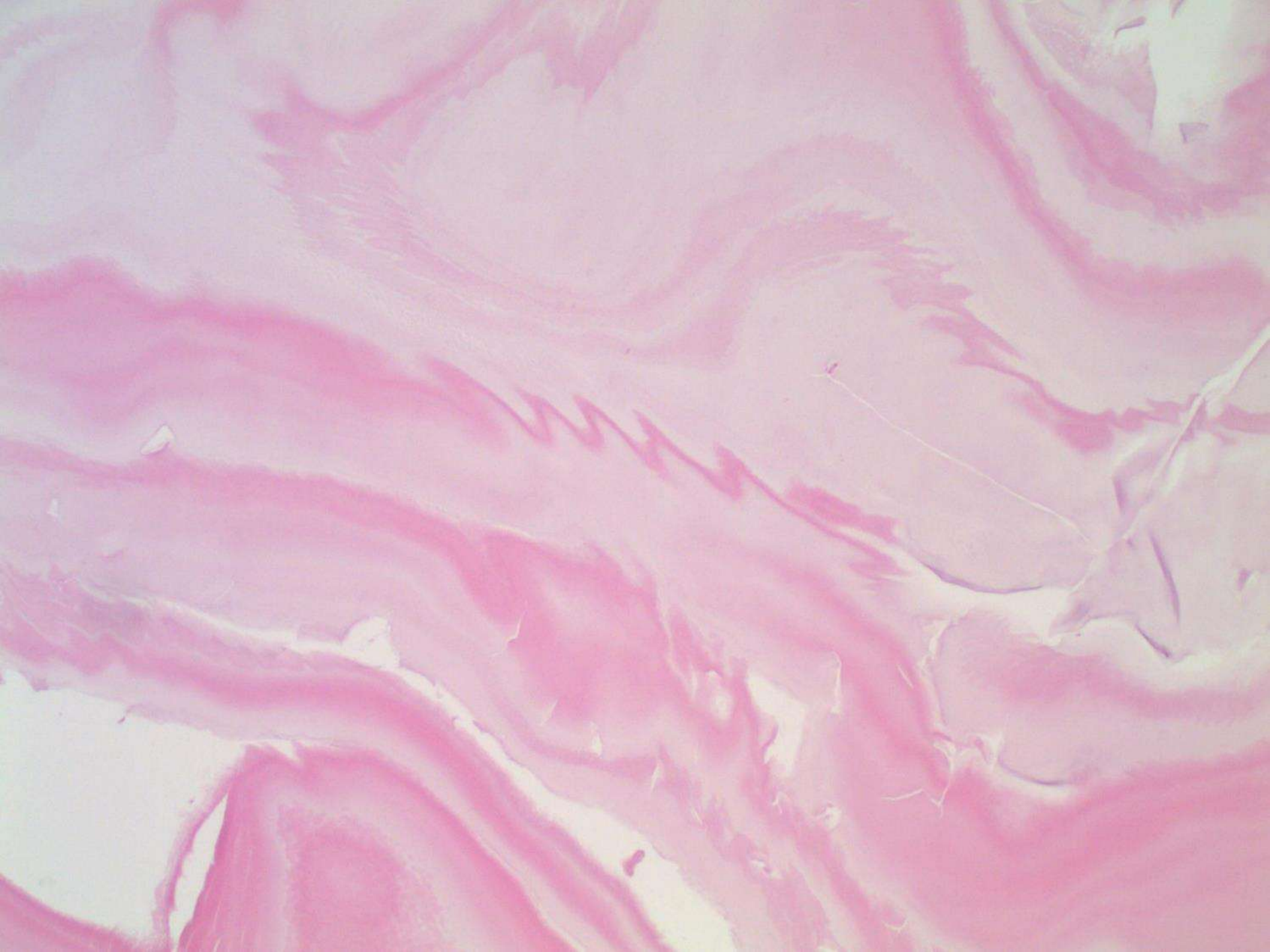


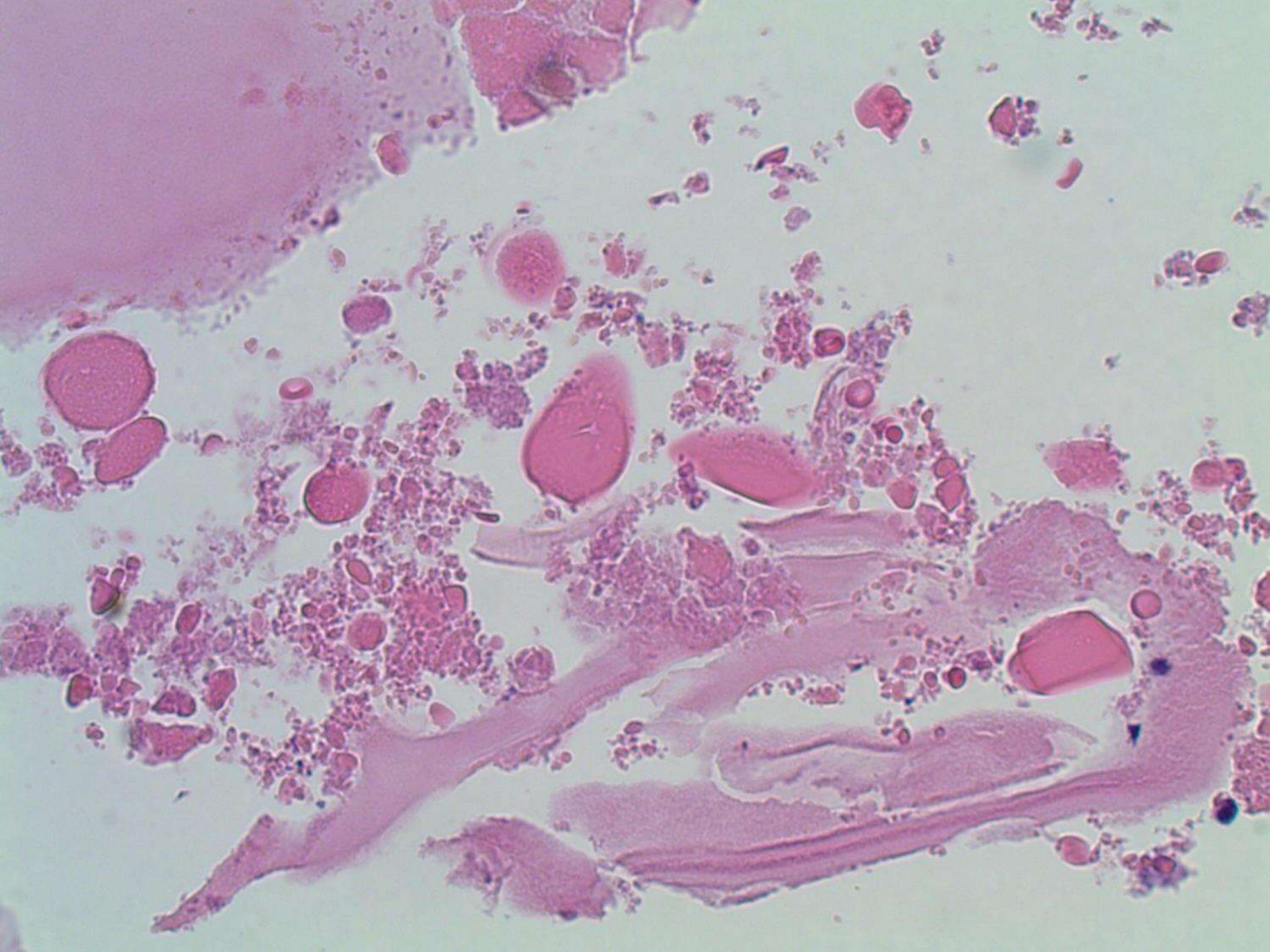


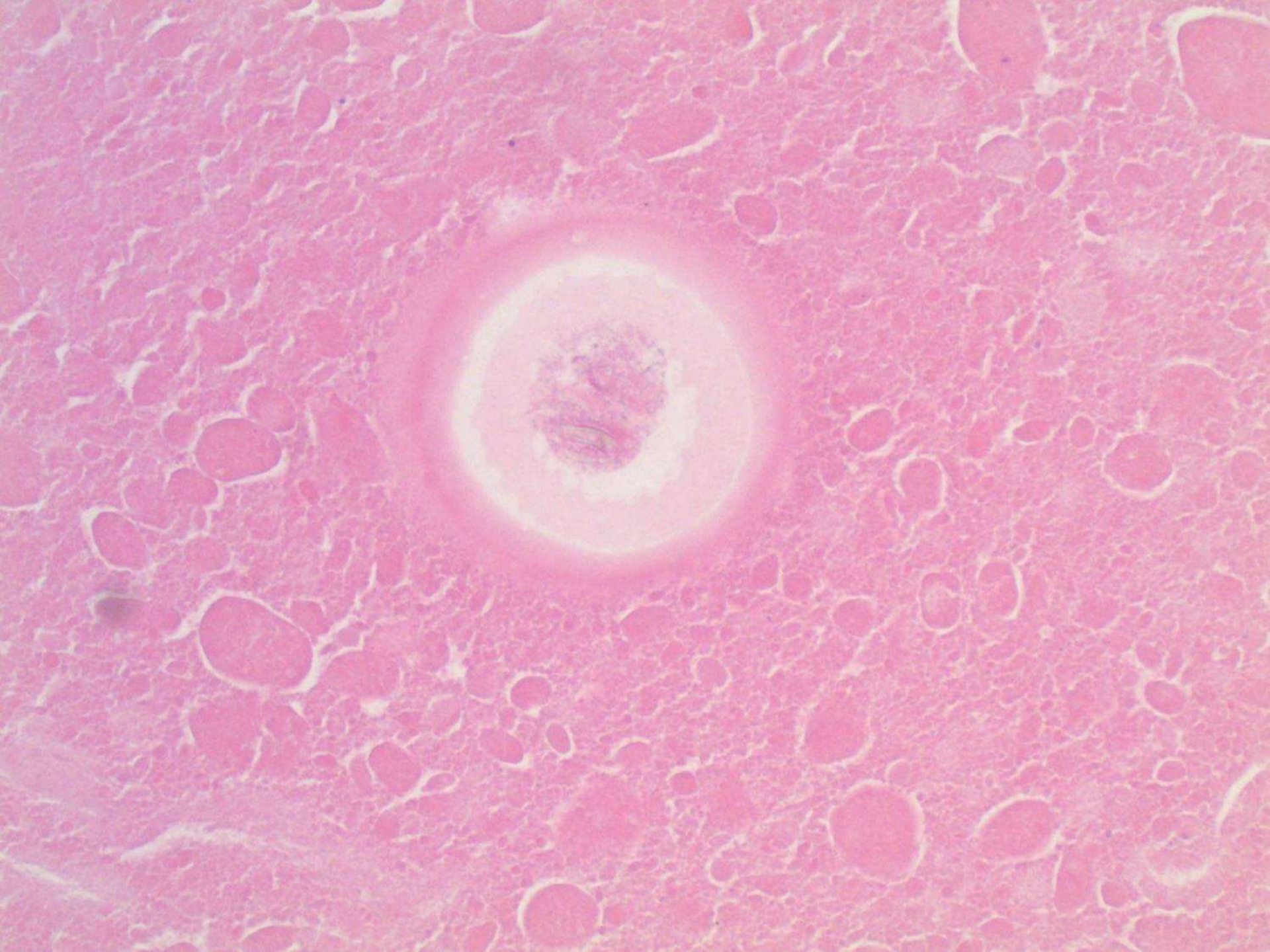


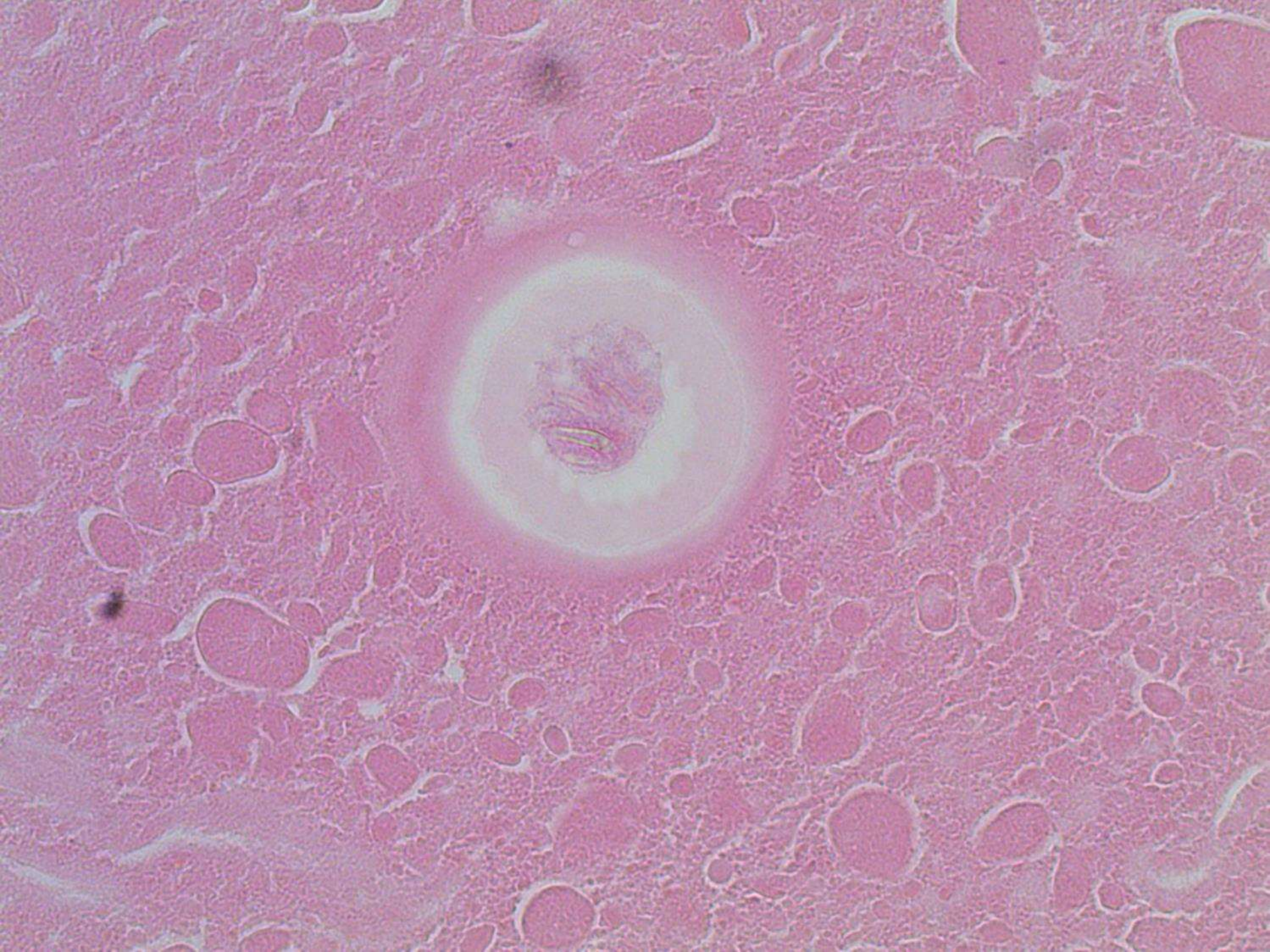


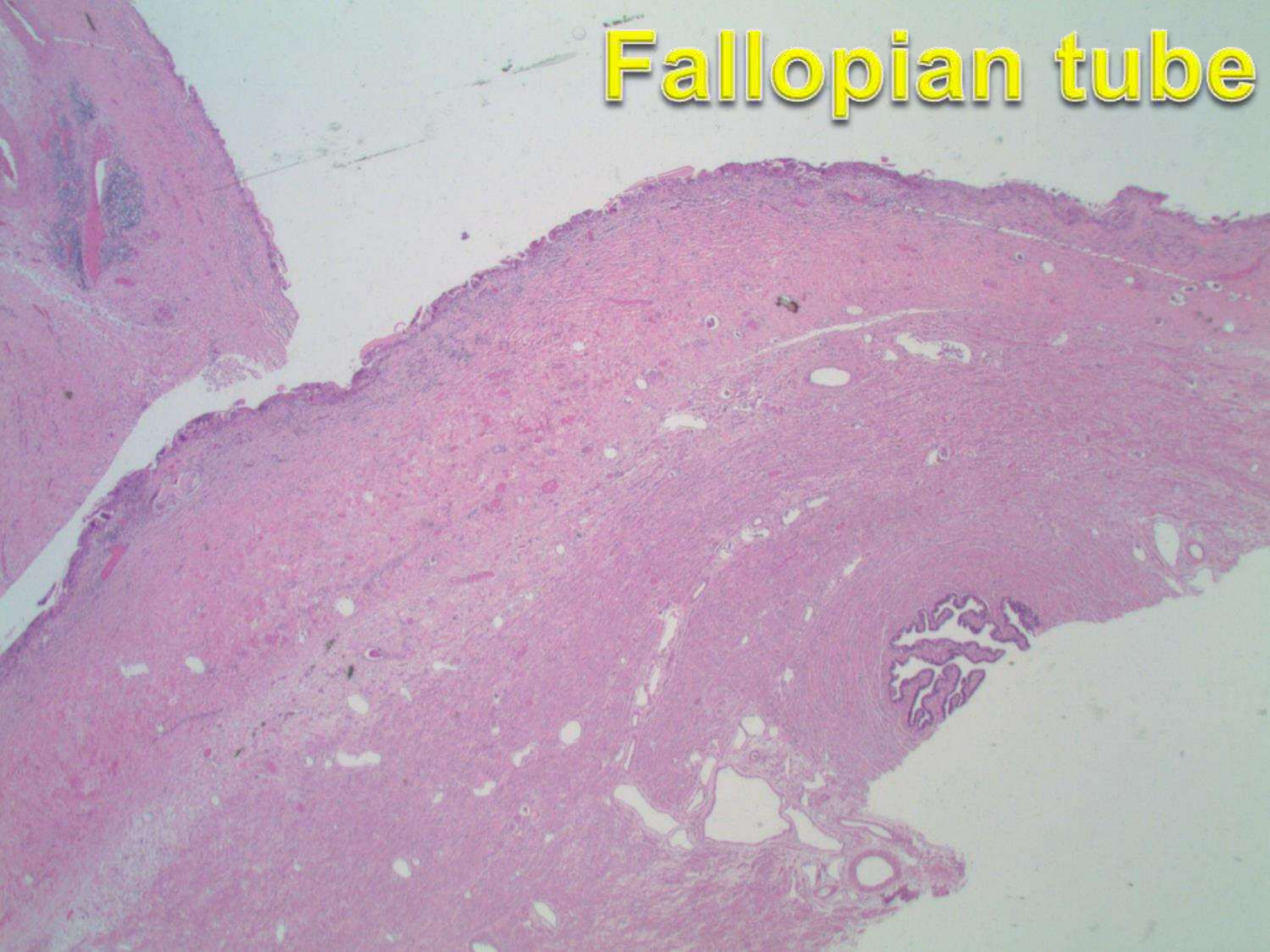


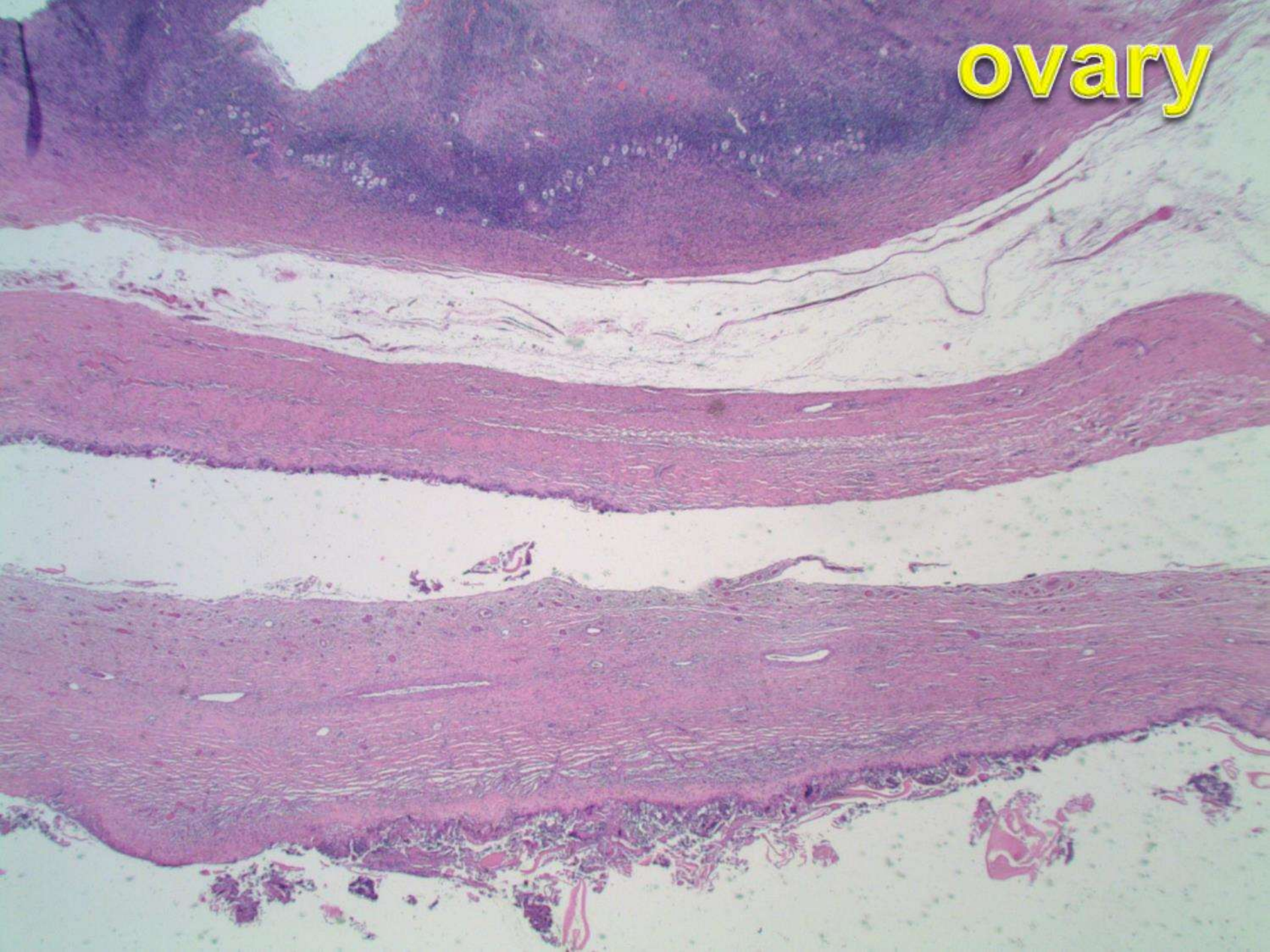


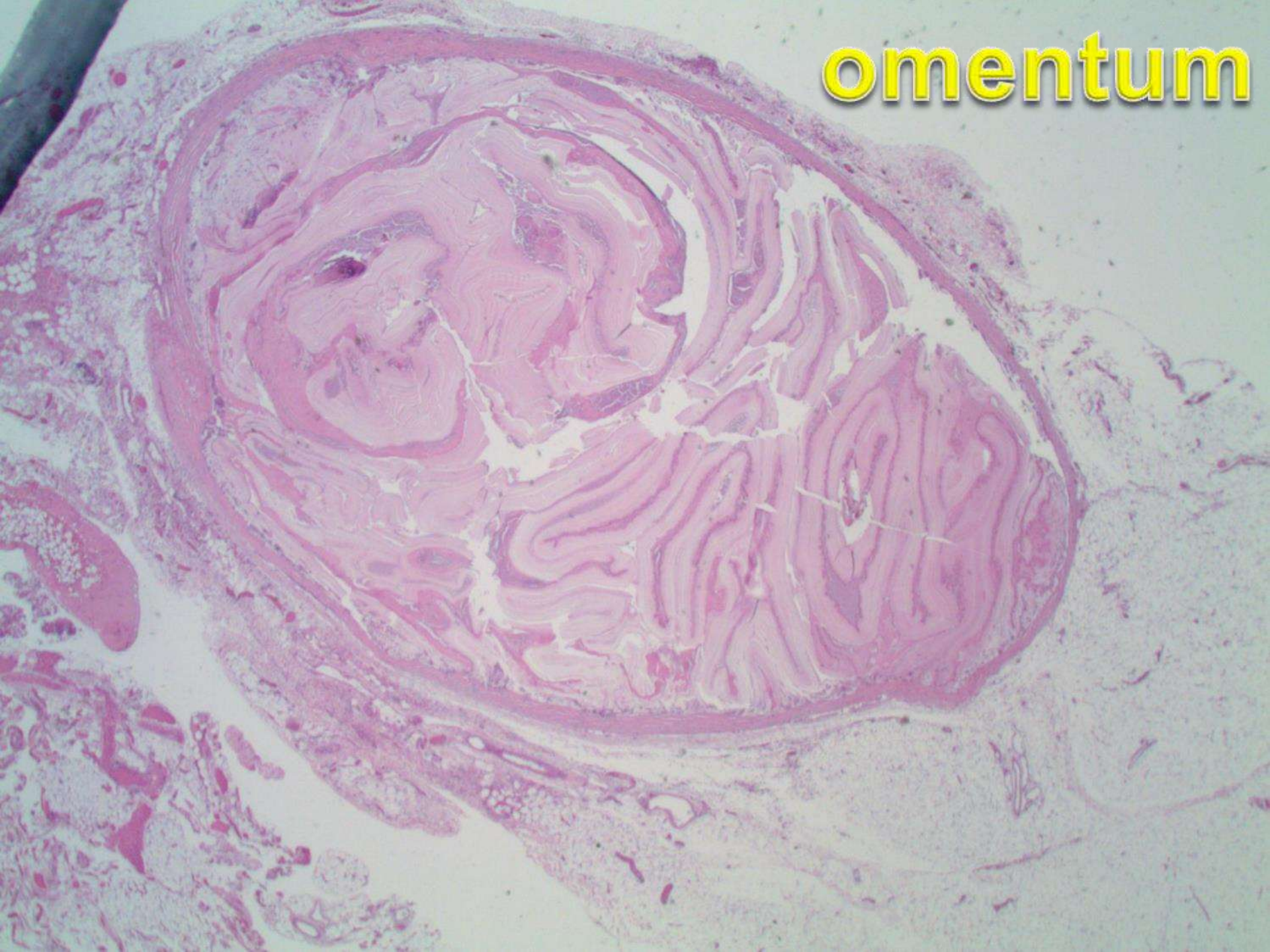


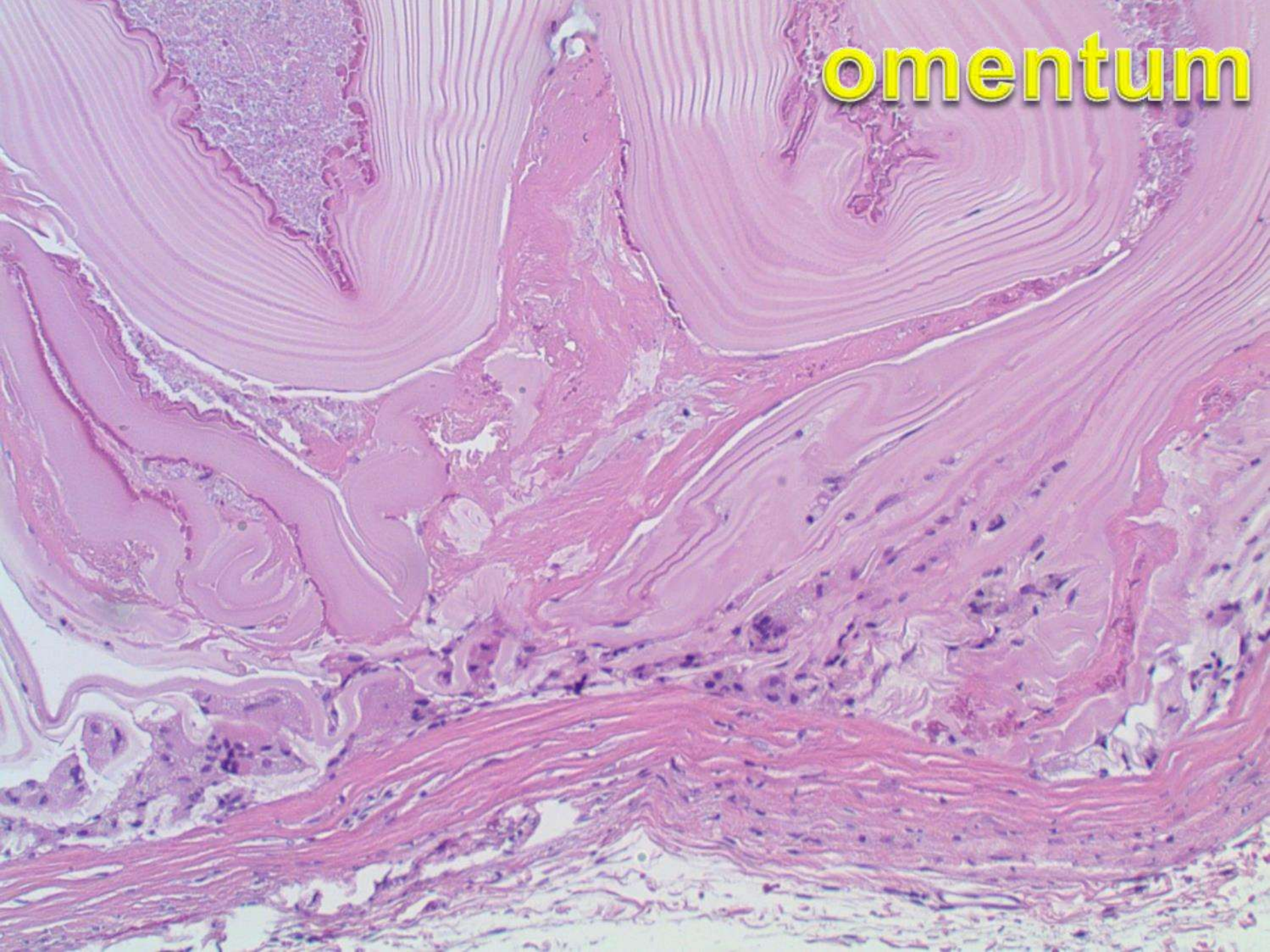





Fallopian tube


ovary


omentum


omentum

Final diagnosis

- **Right pelvic Echinococcal cyst**
  - Involving right ovary/fallopian tube
  - Omentum
- **Serology**
  - Positive for *Echinococcal granulosus*

International Journal of Gynecological Pathology
37:301–304, Lippincott Williams & Wilkins, Baltimore

Case Report

Coexisting Primary Ovarian and Omental Hydatid Disease Mimicking an Ovarian Neoplasm: A Case Report

Emre E. Tas, Ph.D., Gulin F. Yegin Akcay, Ph.D., Fatma Yildirim, Ph.D., and Filiz Yavuz, Ph.D.

Summary: Hydatid disease is a parasitic infection that most commonly affects the liver and lungs, although the disease can arise in any part of the body. Cysts may mimic many benign and malignant conditions. The diagnosis cannot be confirmed preoperatively in all cases. A 44-yr-old menopausal woman was admitted to the department of gynecology with complaints of abdominal distention. A fixed abdominopelvic mass was identified. Radiology revealed a 20-cm mass with branched septations and solid components. CA-125 level was 55 kU/L, and Risk of Malignancy Index-2 score was 880. These findings suggested the presence of an ovarian neoplasm, and laparotomy was performed. Cystic masses measuring 22 cm and 4 cm, originating from the omentum majus and left ovary, respectively, were found during surgery. Frozen-section analysis revealed hydatid disease. Infracolic omentectomy and total abdominal hysterectomy with bilateral salpingo-oopherectomy were performed. Results of a serum *Echinococcus* hemagglutination test performed immediately after surgery were negative. The patient was prescribed albendazole for 6 mo and discharged on the third postoperative day with no complaints. The incidence of hydatid disease in the female reproductive system is very rare; however, clinicians must be aware of this disease and take necessary precautions while operating because any spillage may lead to anaphylactic shock and increased risk of recurrence. **Key Words:** Echinococcus—Ovarian neoplasms—Omentum.

Updates Surg (2015) 67:279–282
DOI 10.1007/s13304-015-0291-6

ORIGINAL ARTICLE

Unusual locations of hydatid disease: a 33 year's experience analysis on 233 patients

Georgios D. Lianos[1] · Avrilios Lazaros[1] · Konstantinos Vlachos[1] · Georgios K. Georgiou[1] · Haralampos V. Harissis[1] · Alberto Mangano[2] · Stefano Rausei[2] · Luigi Boni[2] · Francesco Frattini[2] · Antonio Biondi[3] · Gianlorenzo Dionigi[2] · Christos Katsios[1]

Table 1 Extrahepatic hydatid disease

Location	No of cysts	%
Spleen	11	17
Pelvis	9	14
Lungs	2	3
Diaphragm	2	3
Colon	9	14
Mesentery	1	1.6
Omentum	7	11
Peritoneum	22	34.4
Bladder	1	1.6
Total	64	100

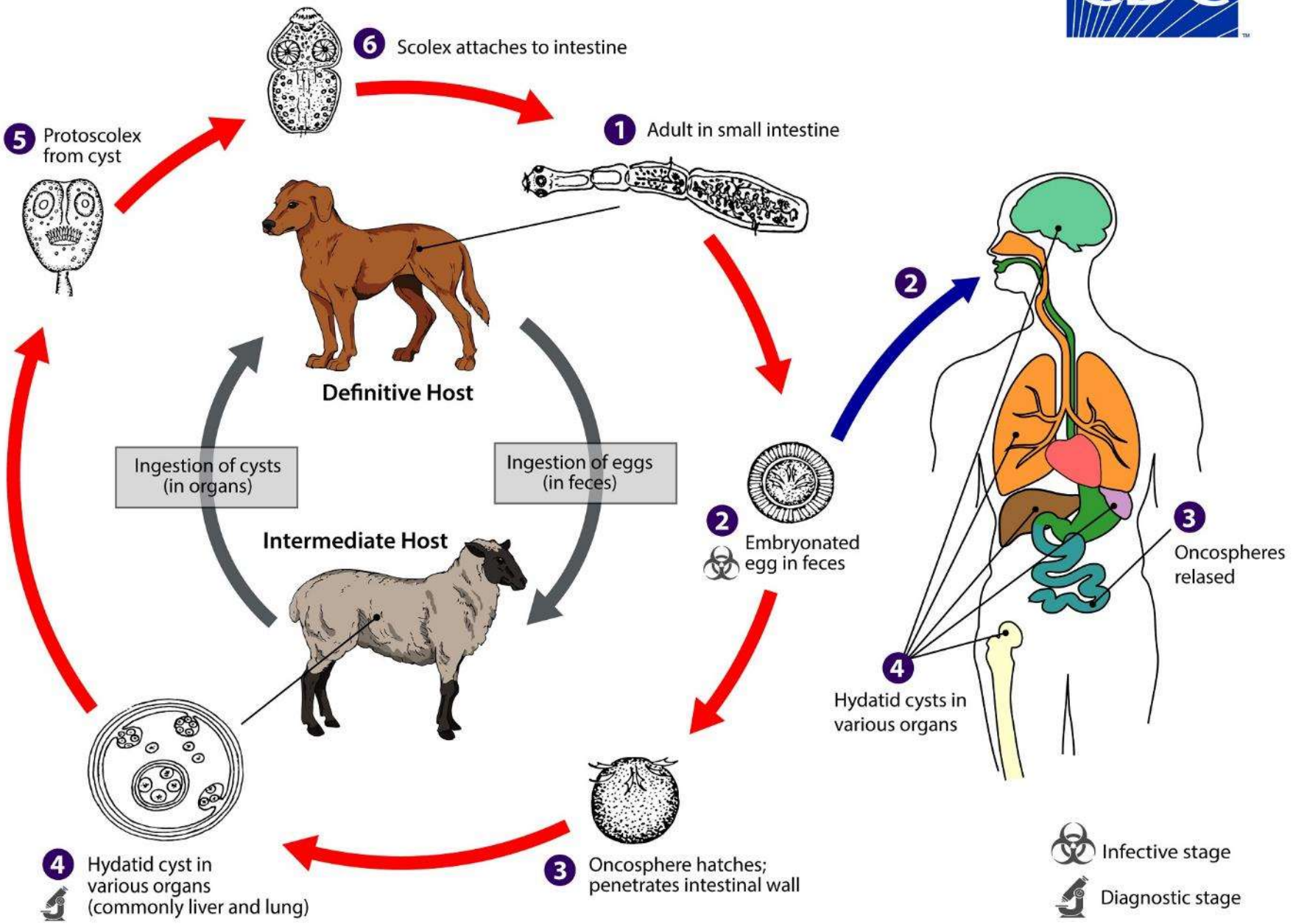
Table 1: The published cases of the hydatid cyst with unusual locations from Iran

Locations	Numbers	Most common clinical manifestations	References
Intracranial Spinal Orbit	256	Headache Low back pain Visual impairment	14-37
Musculoskeletal	57	Swelling Pathologic fracture	38-66
Cardiovascular	42	Angina, dyspnea and palpitation, pressure effect	67-82
Kidney and Urinary Tract	31	Flank pain	83-90
Spleen	20	Left upper quadrant pain	91-94
Ovary Uterus Fallopian Tube	11	Ovarian mass Lower abdominal pain	95-103
Pancreas	6	Epigastric pain	104-109
Salivary Gland	9	Painless swelling	110-118
Breast	8	Breast mass	119-125
Thyroid	4	Thyroid enlargement	126-129
Adrenal	2	Flank pain	130-131
Appendix	1	Abdominal pain	132
Mediastinum	7	Pressure effect on adjacent organs	133-139
Omental, Mesenteric, Retroperitoneal	7	Mostly asymptomatic	140-146
Parapharyngeal	1	Nonspecific	147
Nasolabial	1	Nonspecific	148
Total	463	-	-

Iran J Med Sci March 2013; Vol 38 No 1

Cystic Echinococcosis

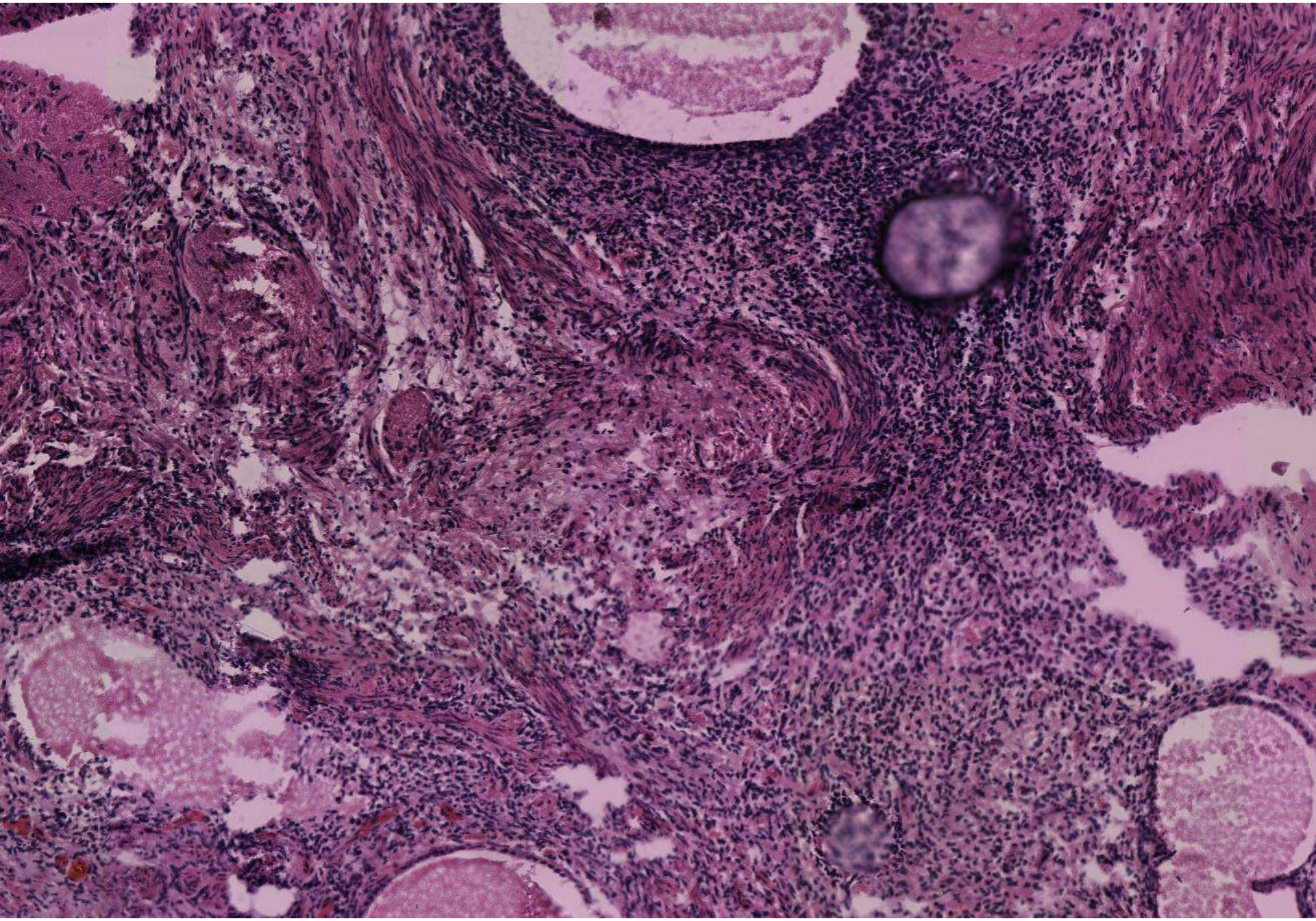
Echinococcus granulosus sensu lato

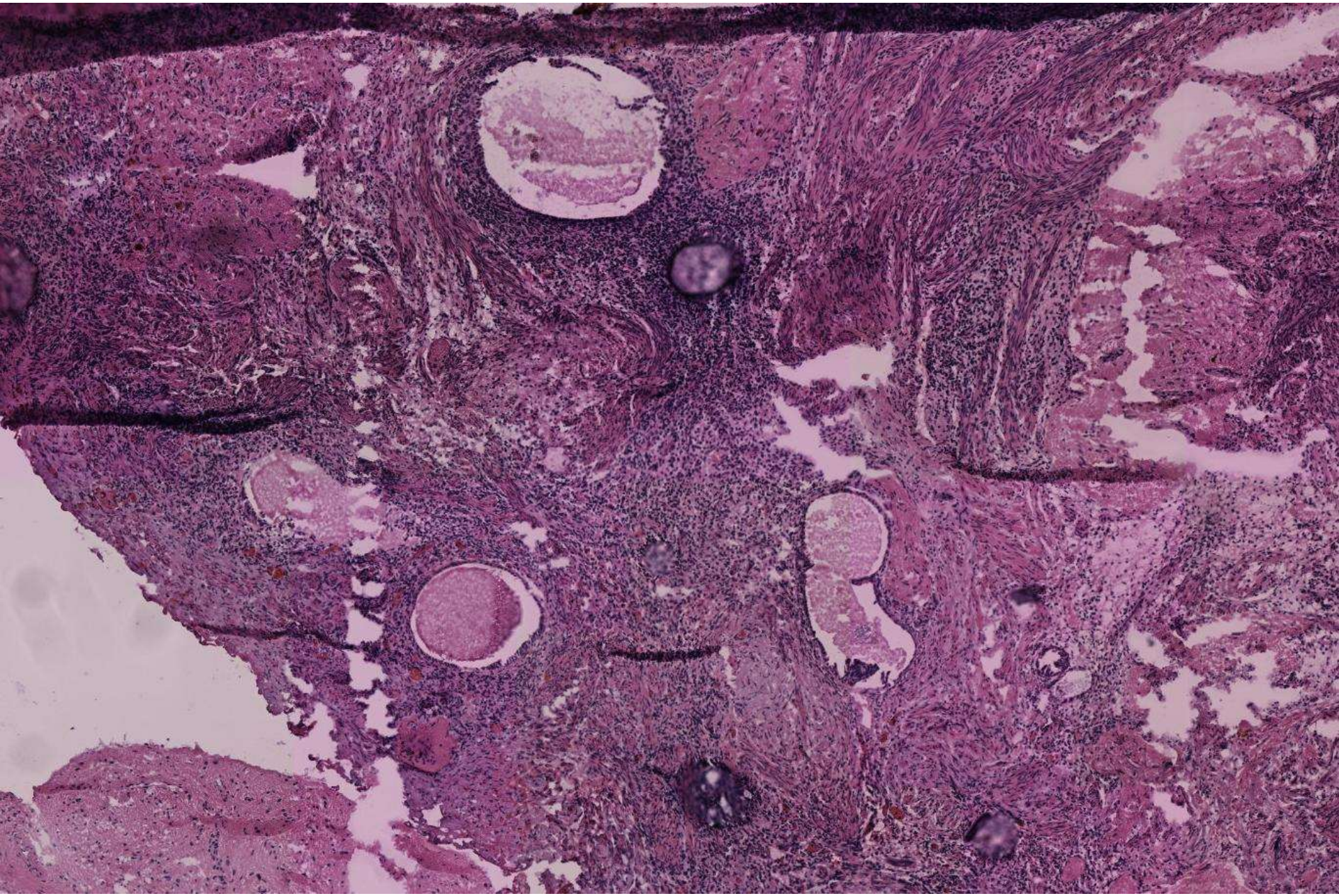


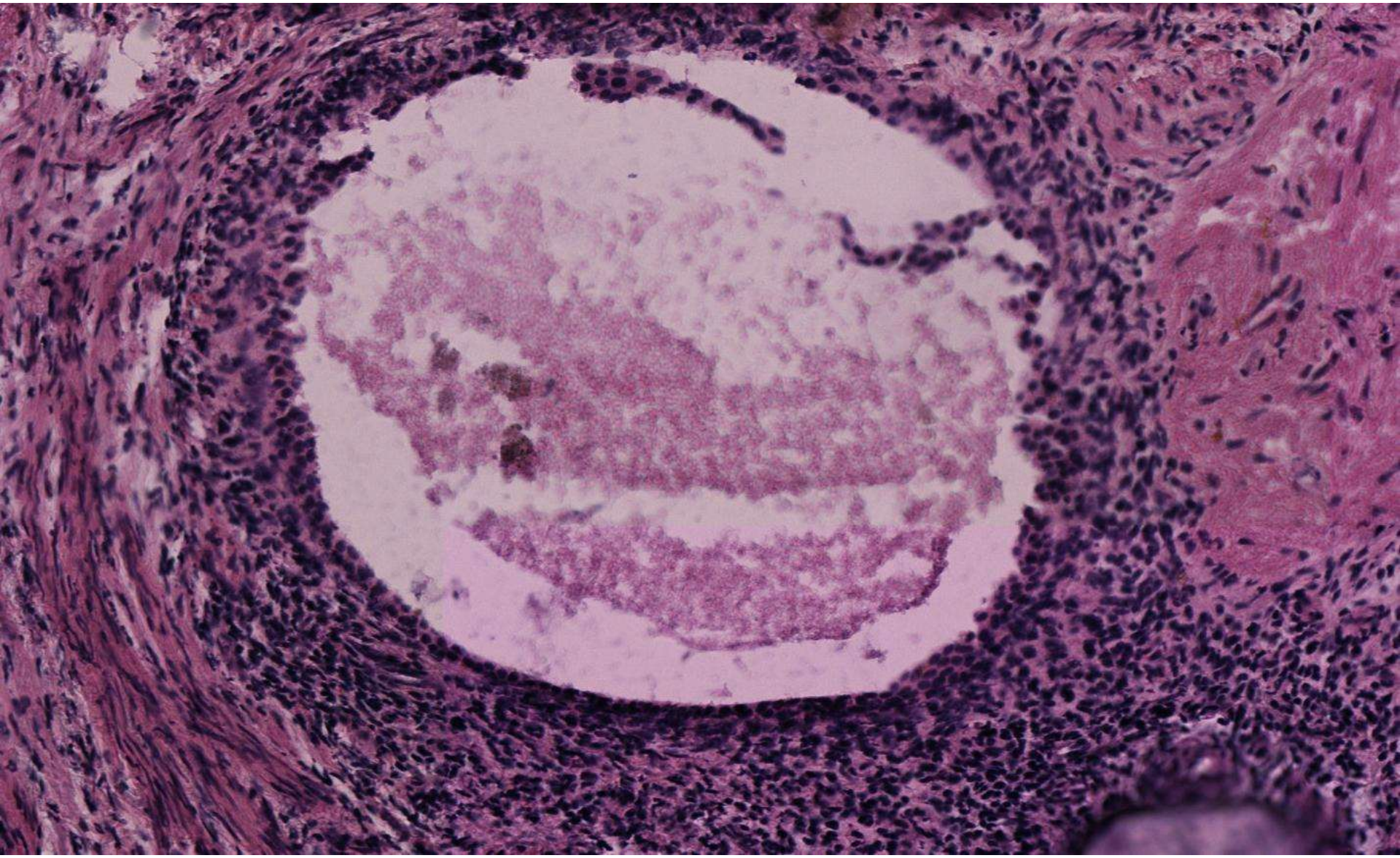
20-0409
SCANNED SLIDE AVAIL!

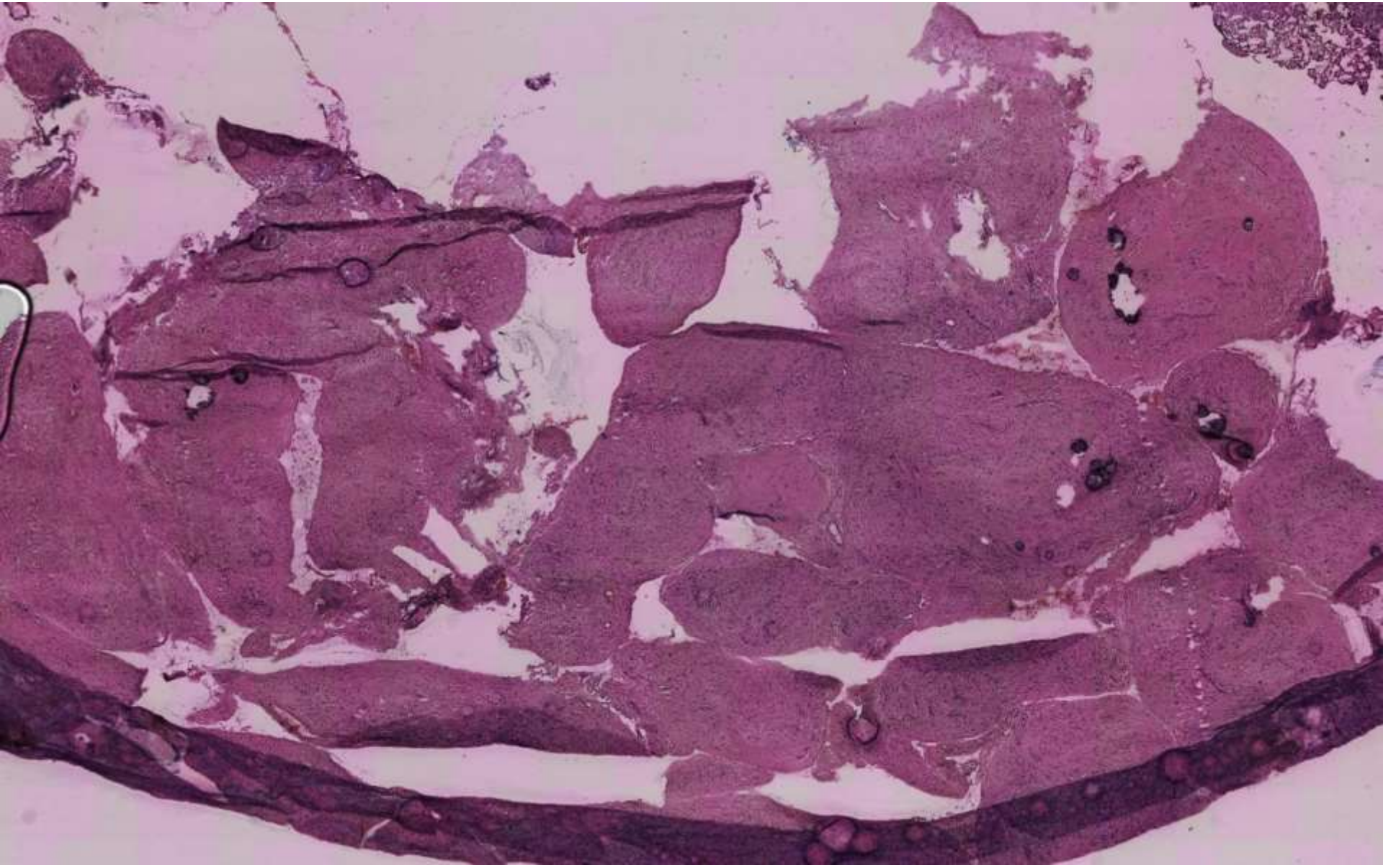
Ankur Sangoi; El Camino Hospital

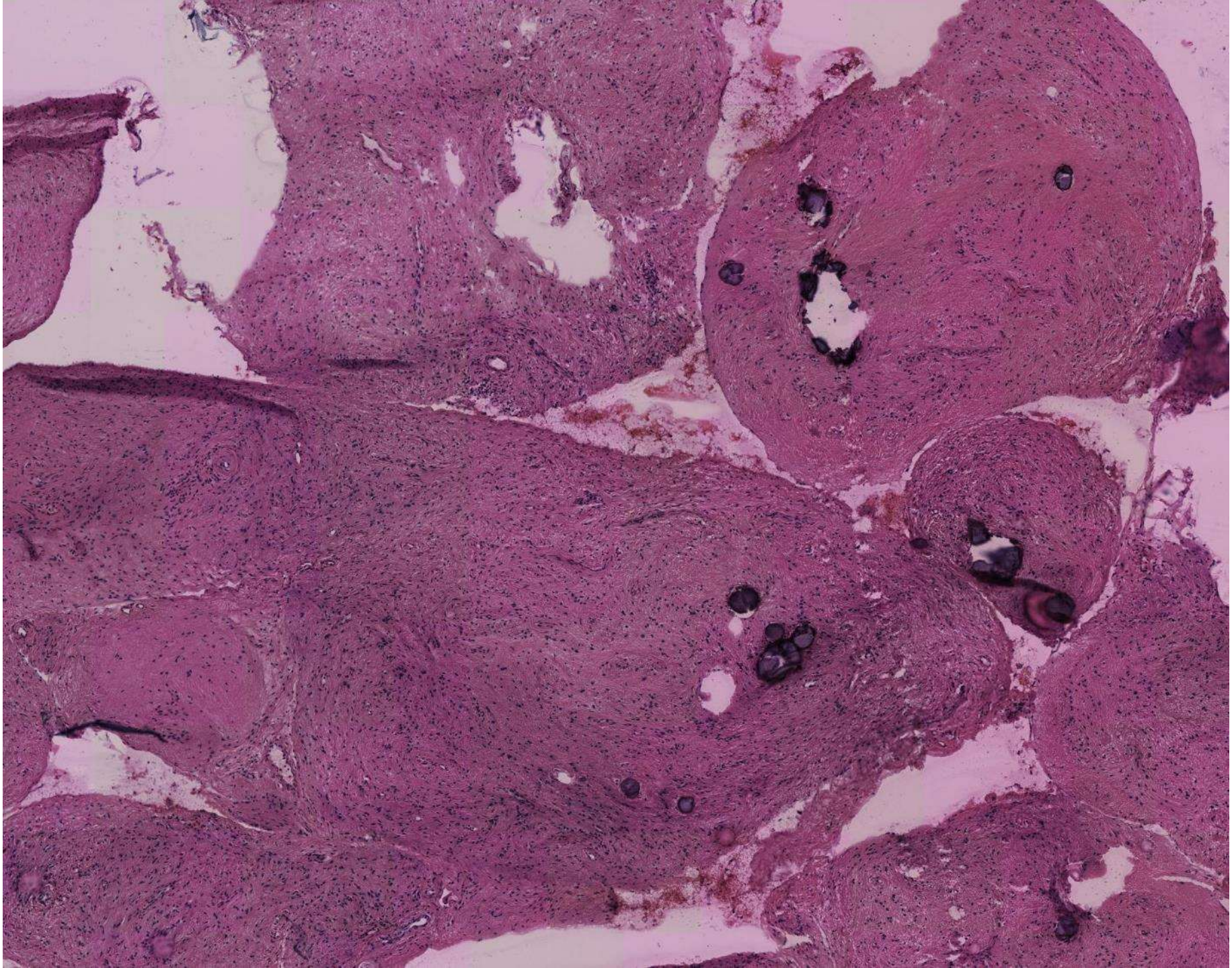
67-year-old F with “lacy” peritoneal lesions found on CT imaging. Section of omentum submitted for frozen section.

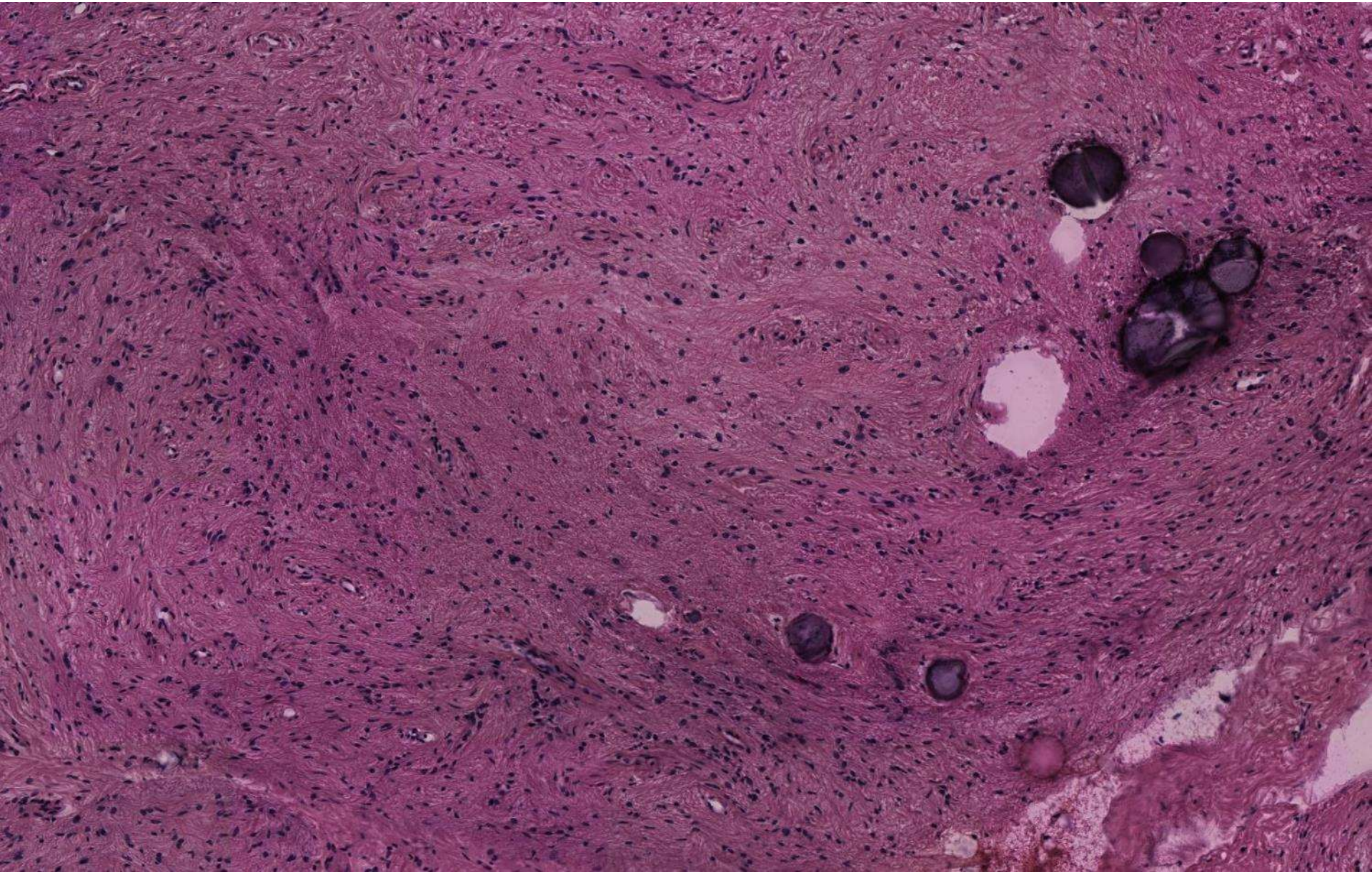


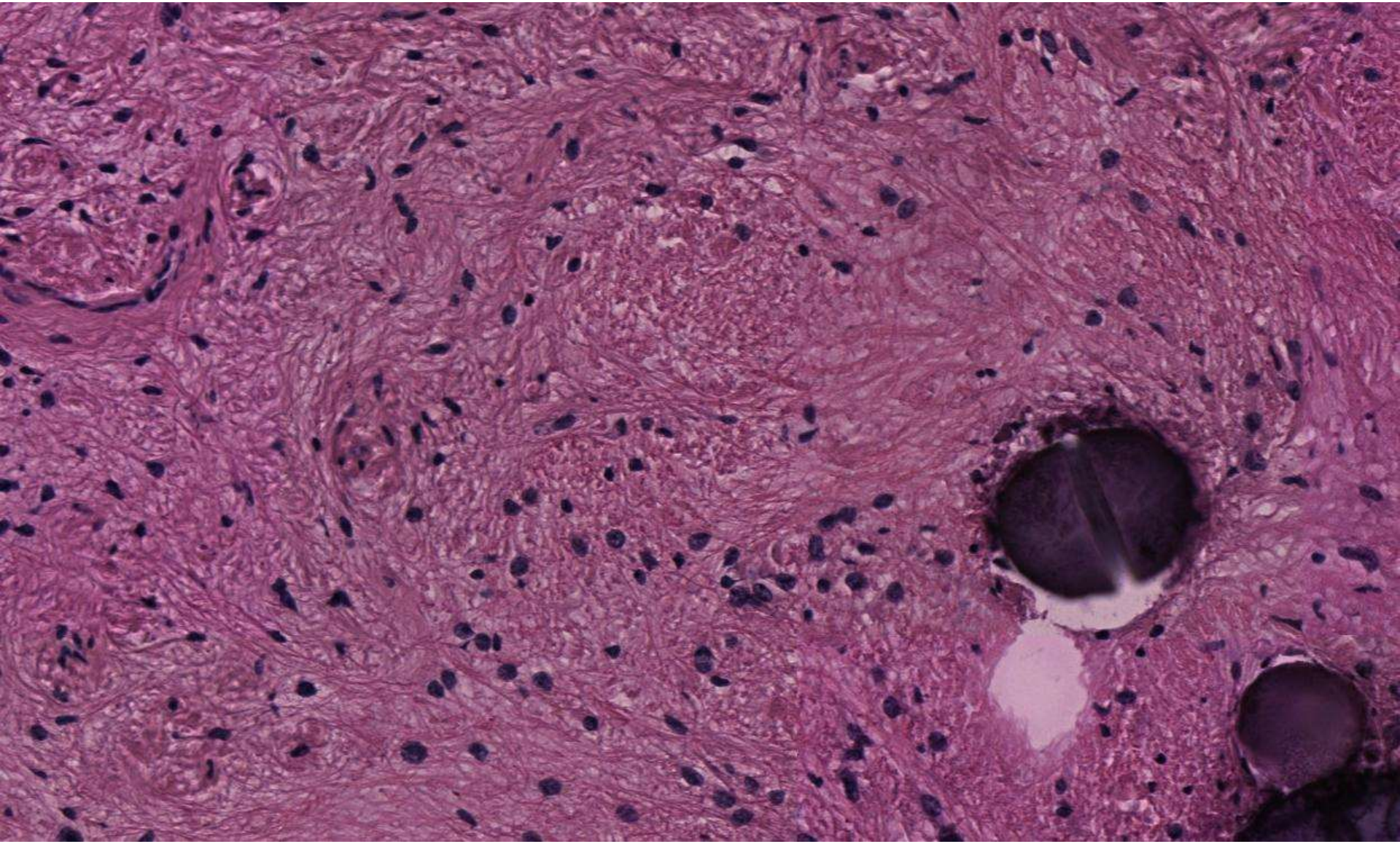


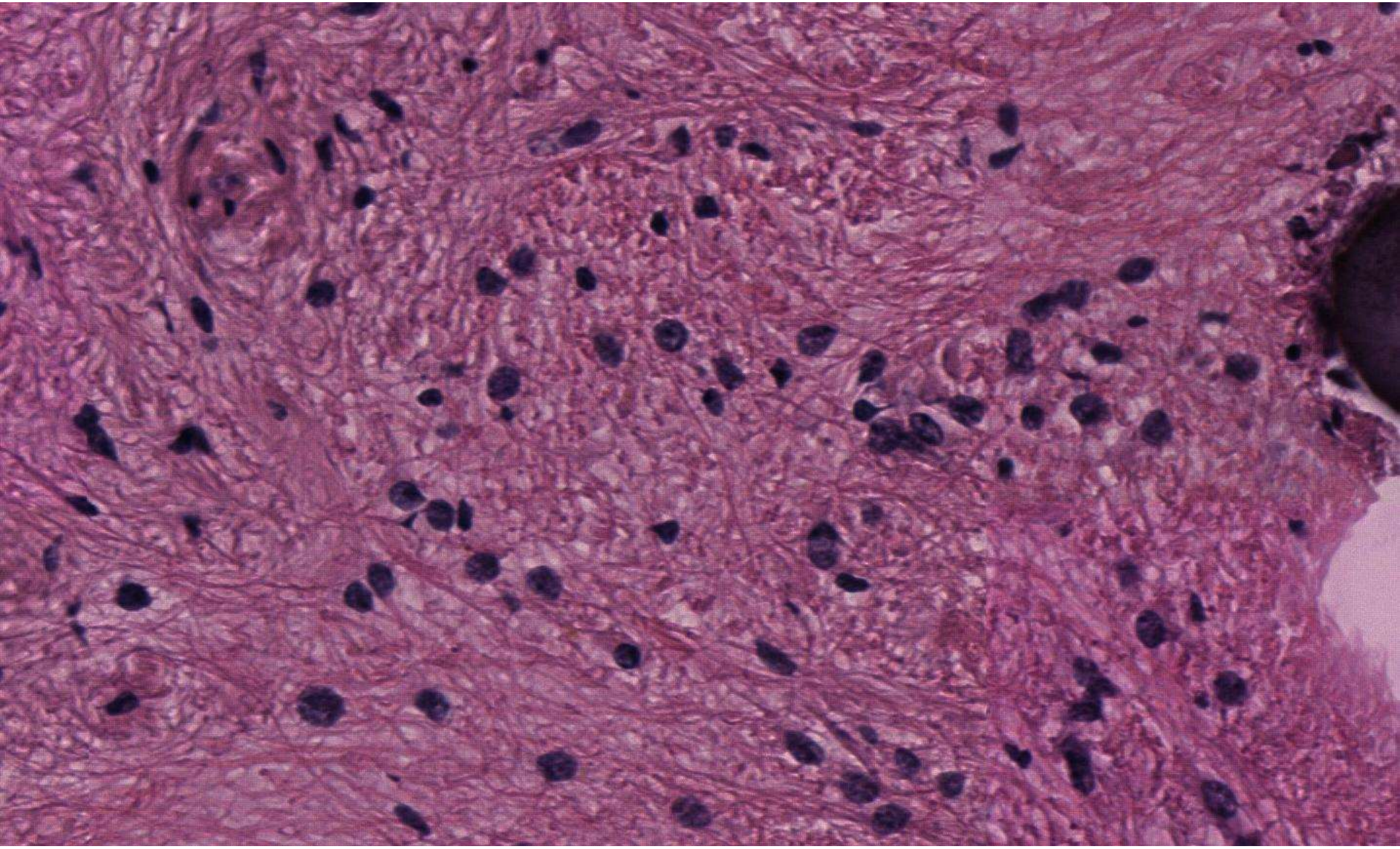






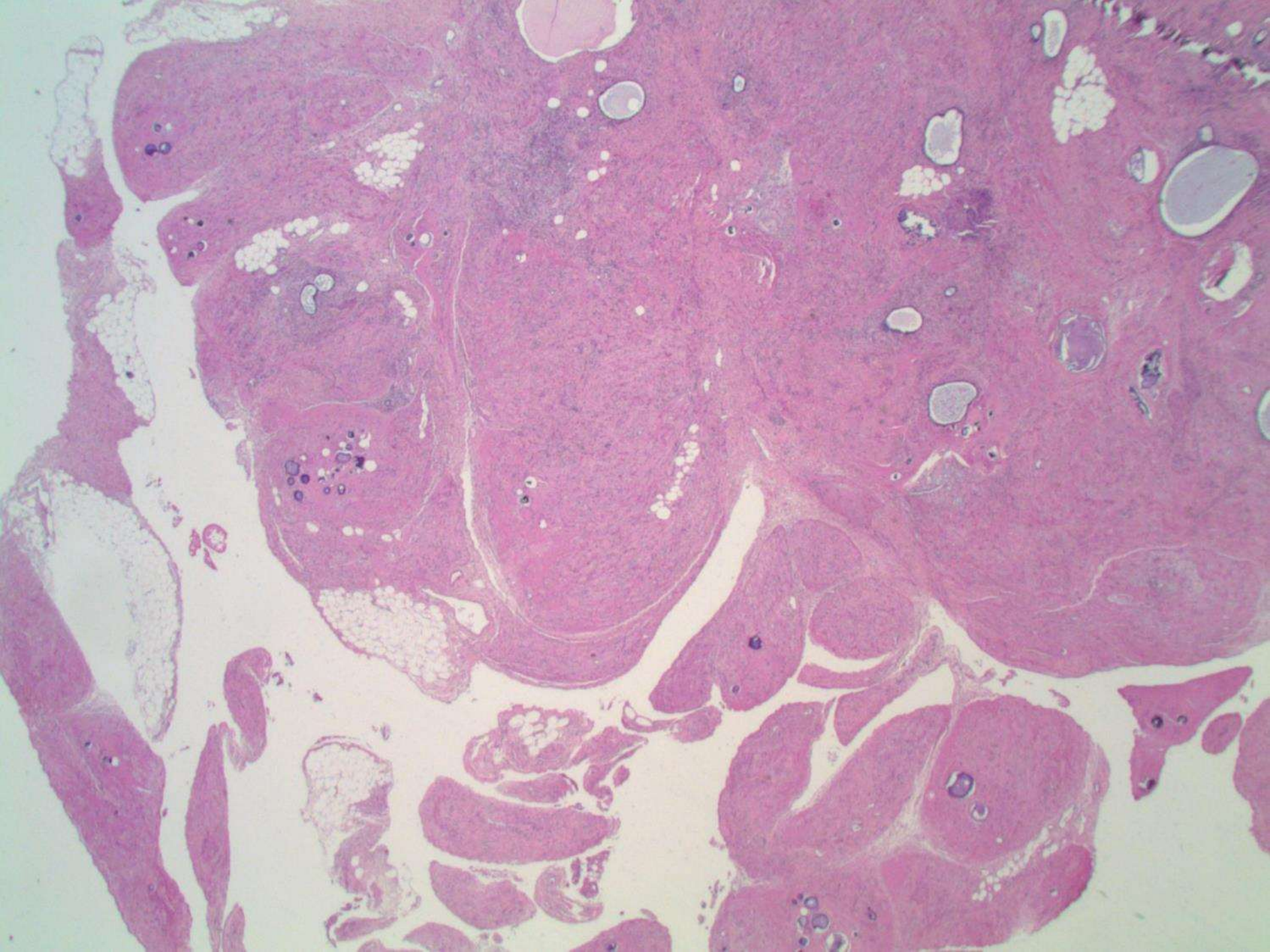


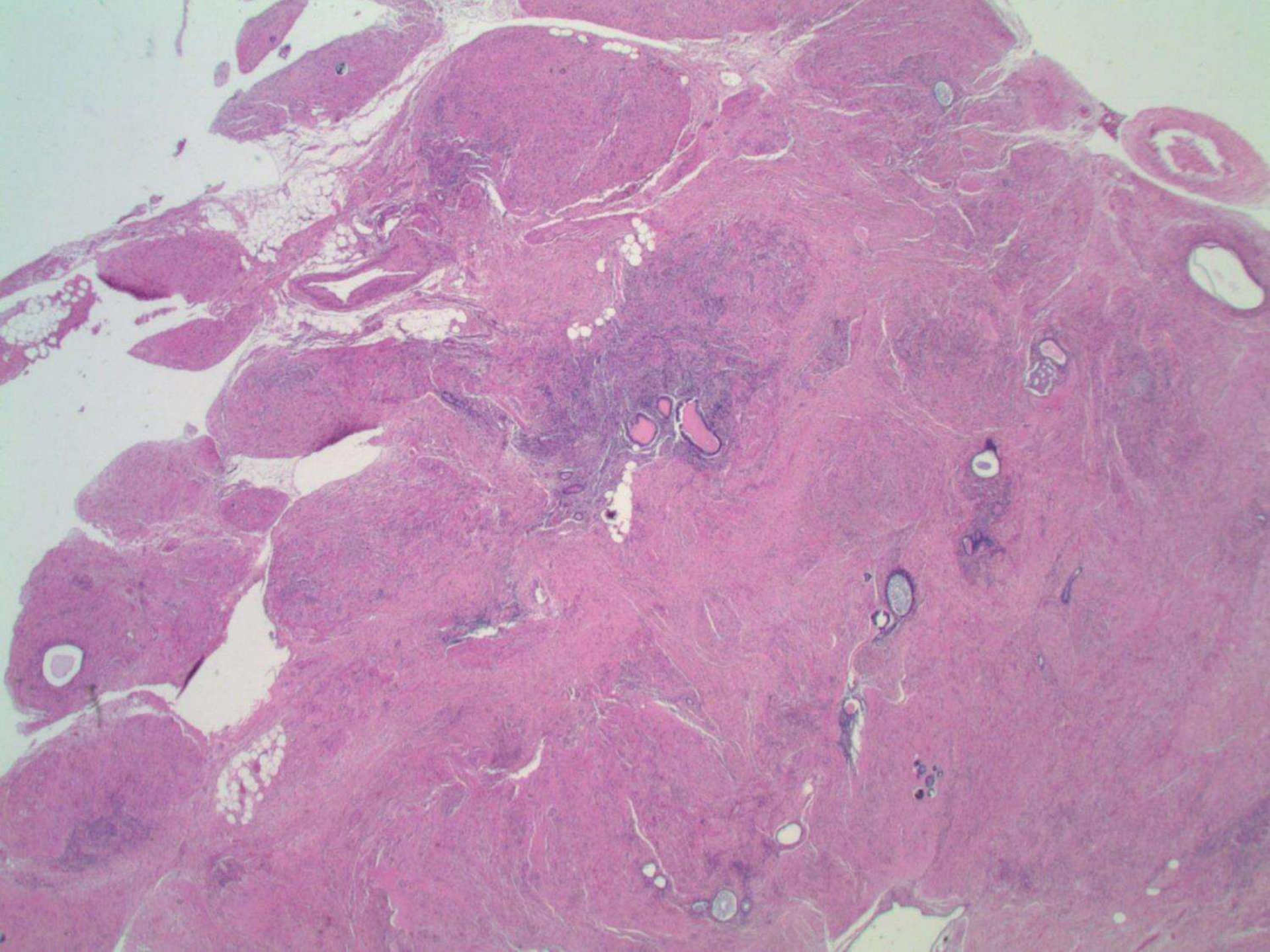


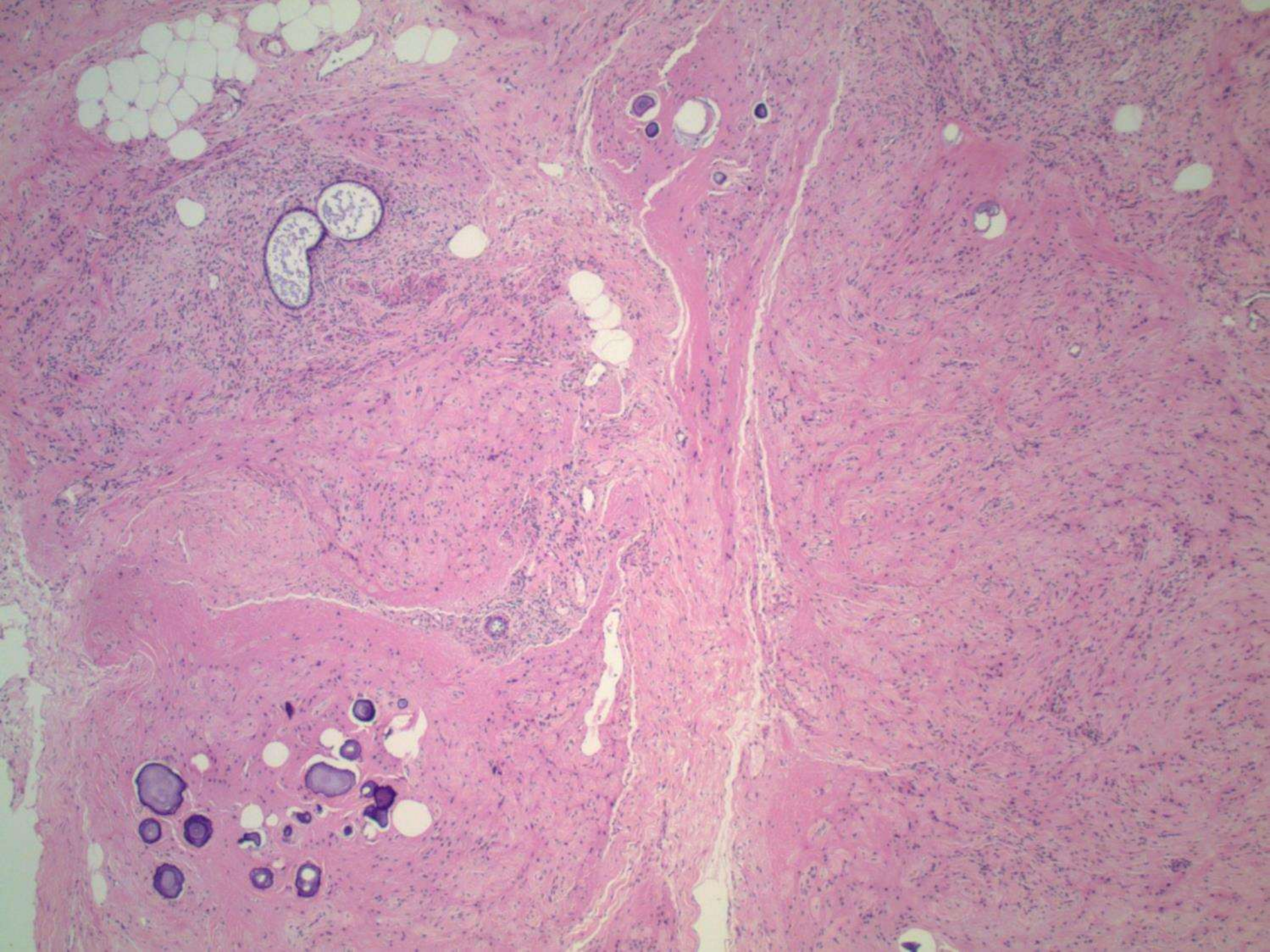


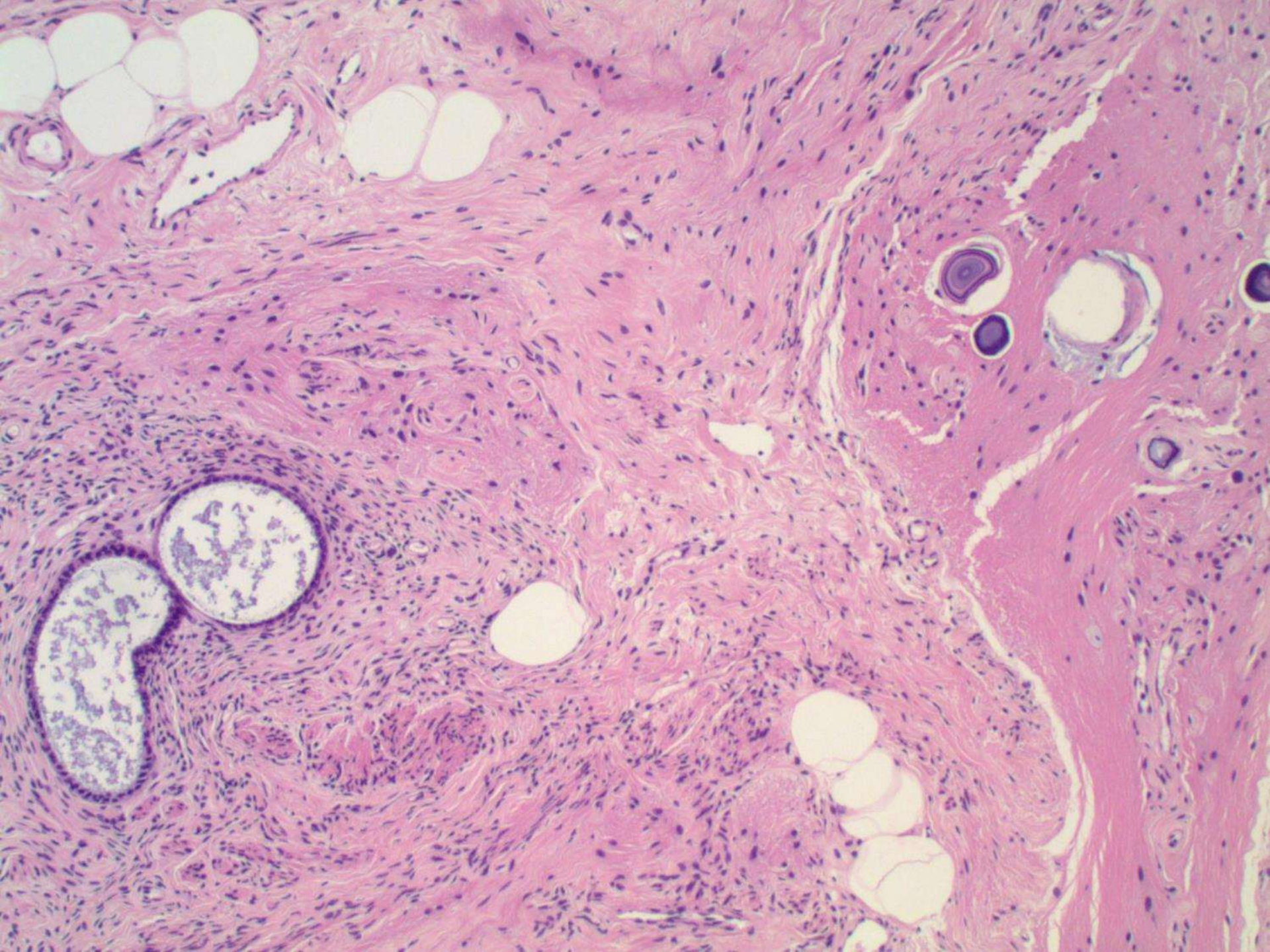
DDx

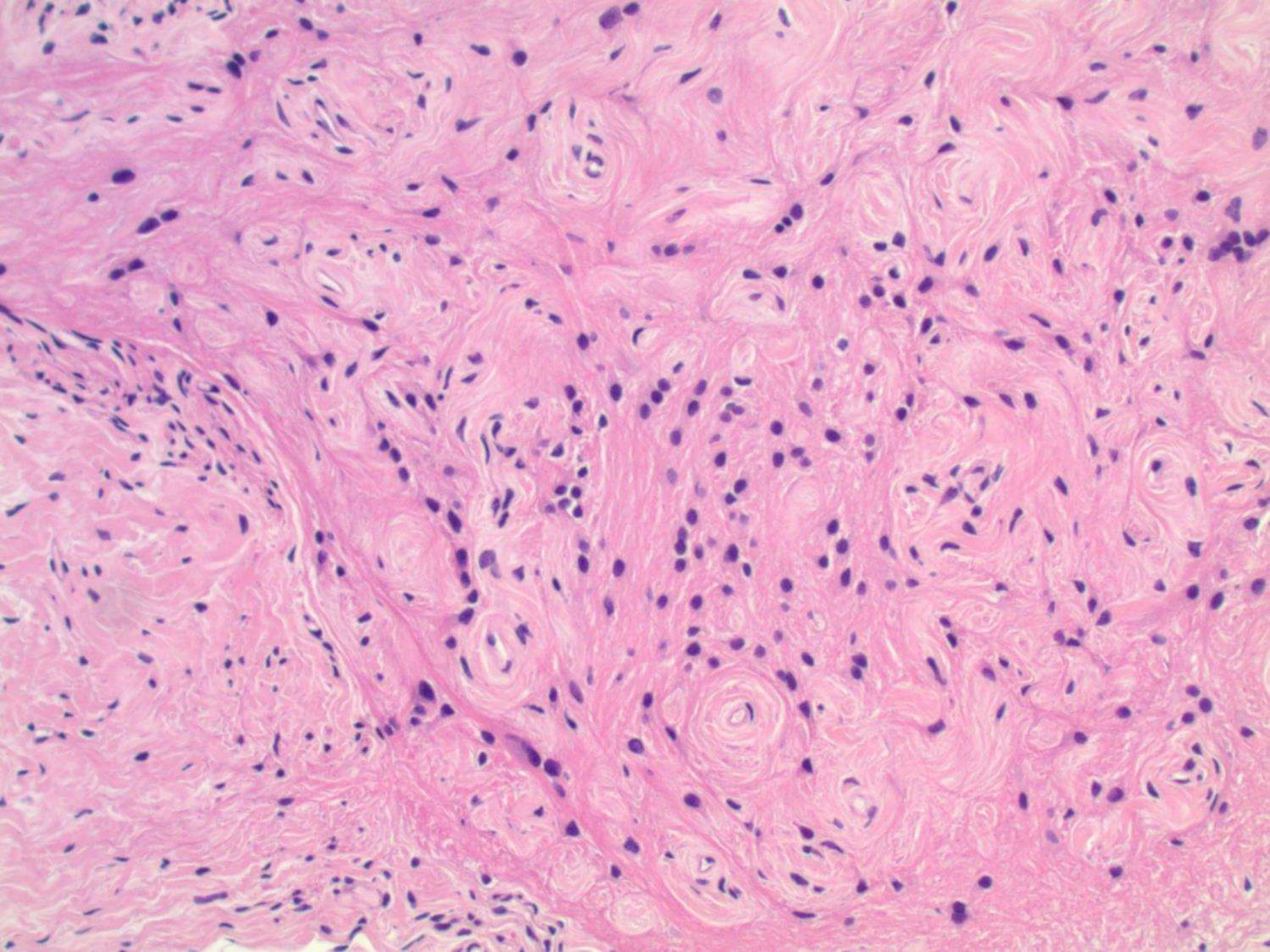
- Endometriosis +/- treatment
- Sclerosing peritonitis
- Inflammatory myofibroblastic tumor
- Adenosarcoma
- Psammocarcinoma
- Gliomatosis peritonei
- Mesothelioma

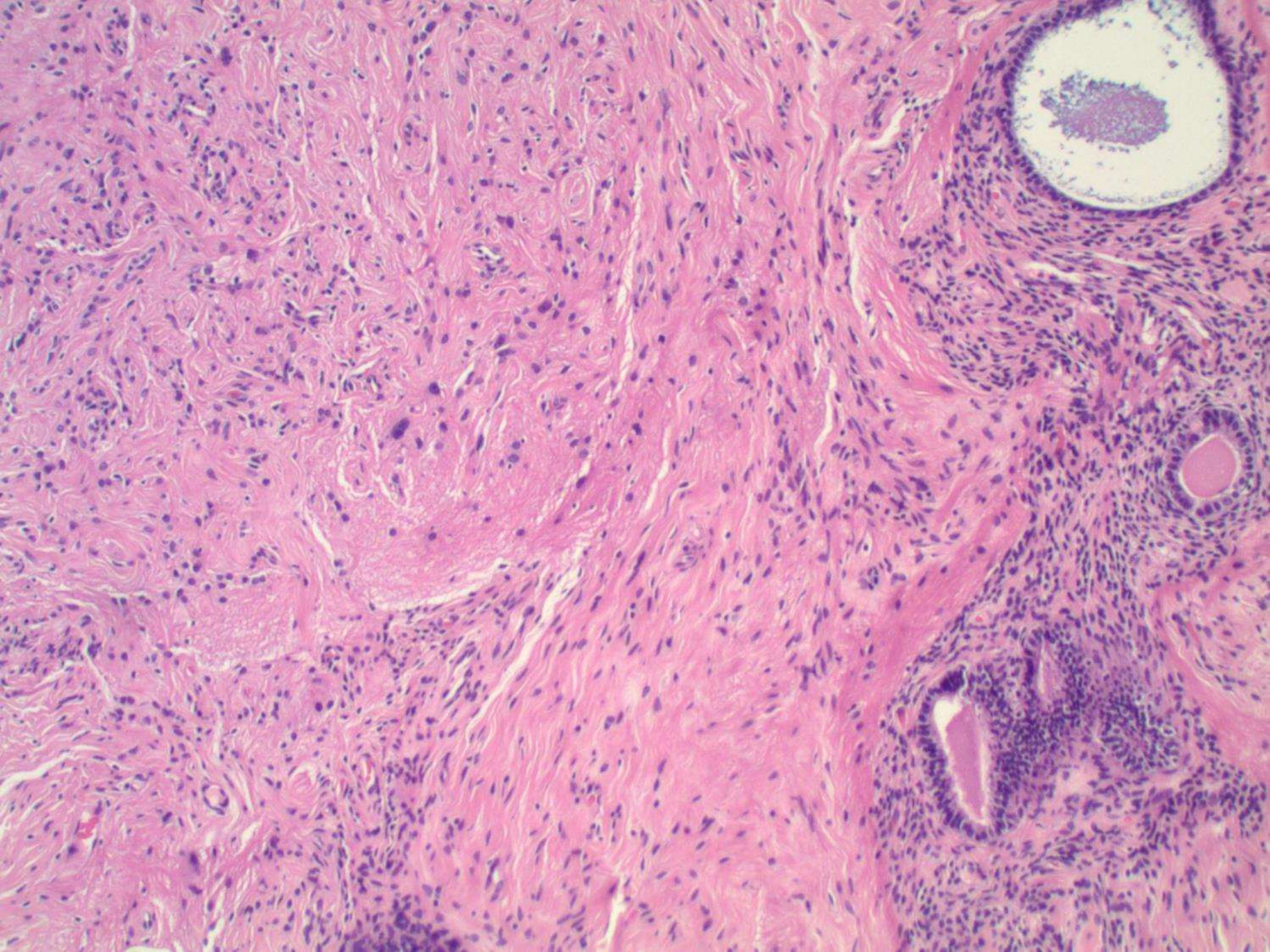


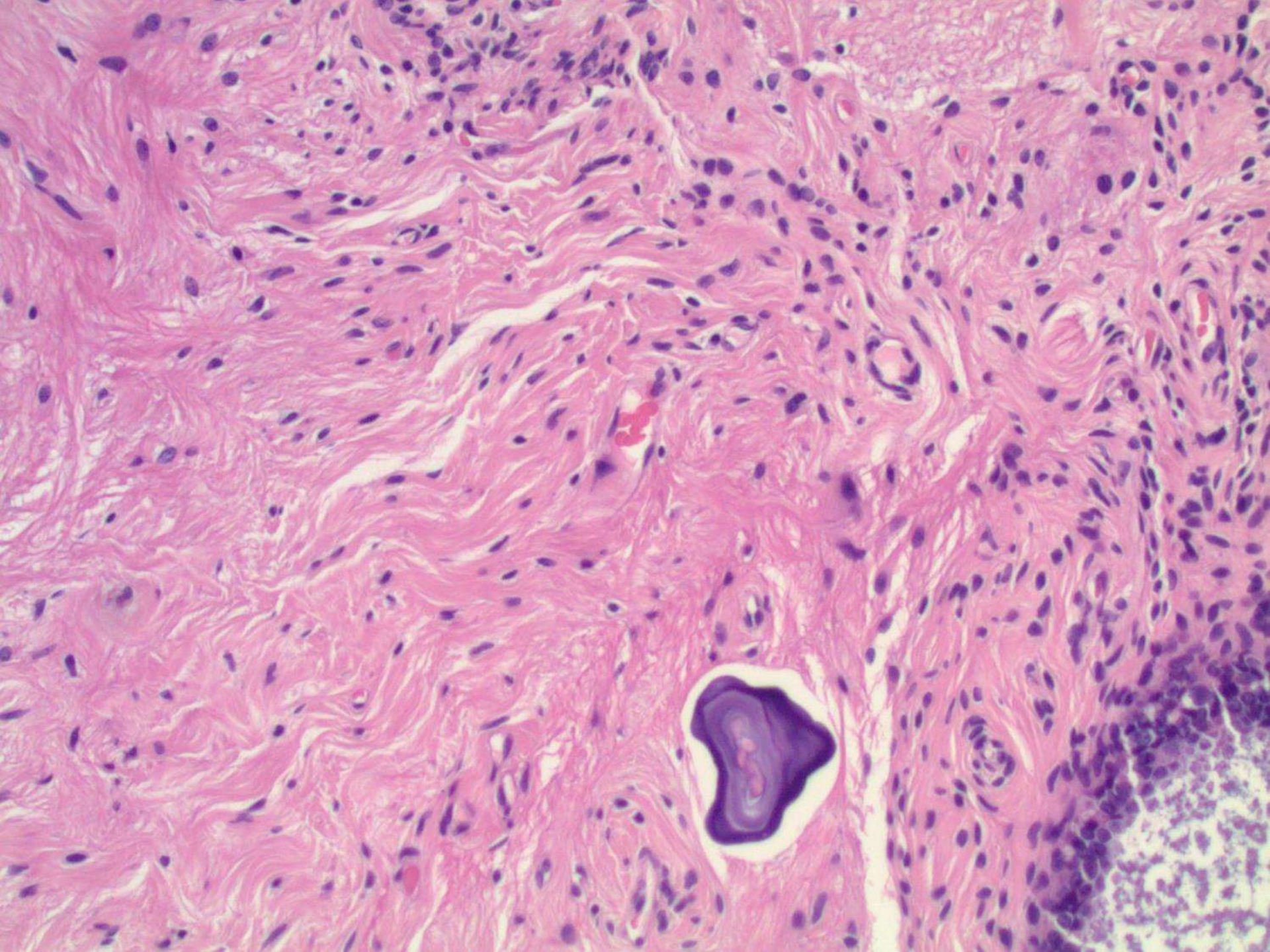


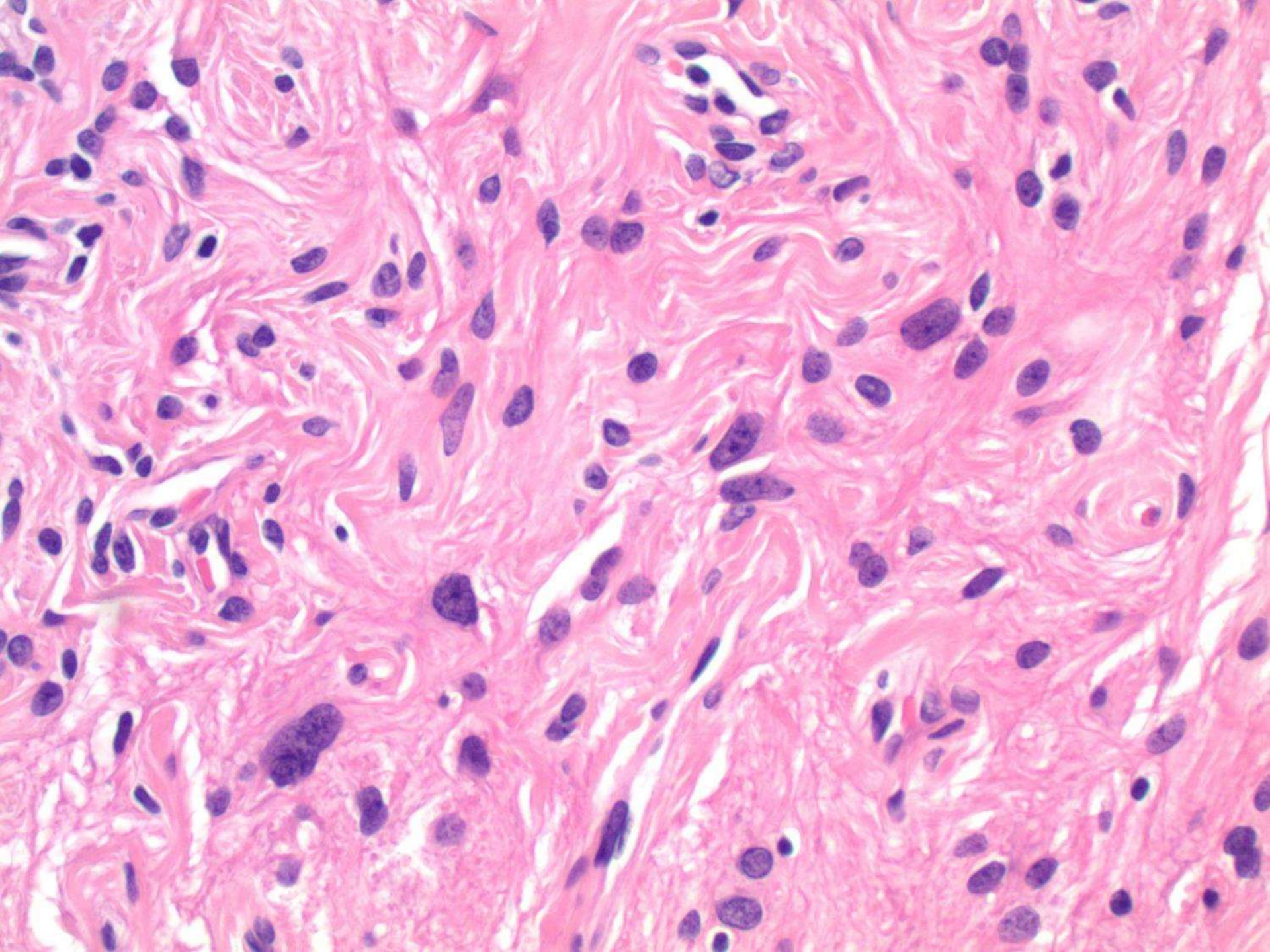


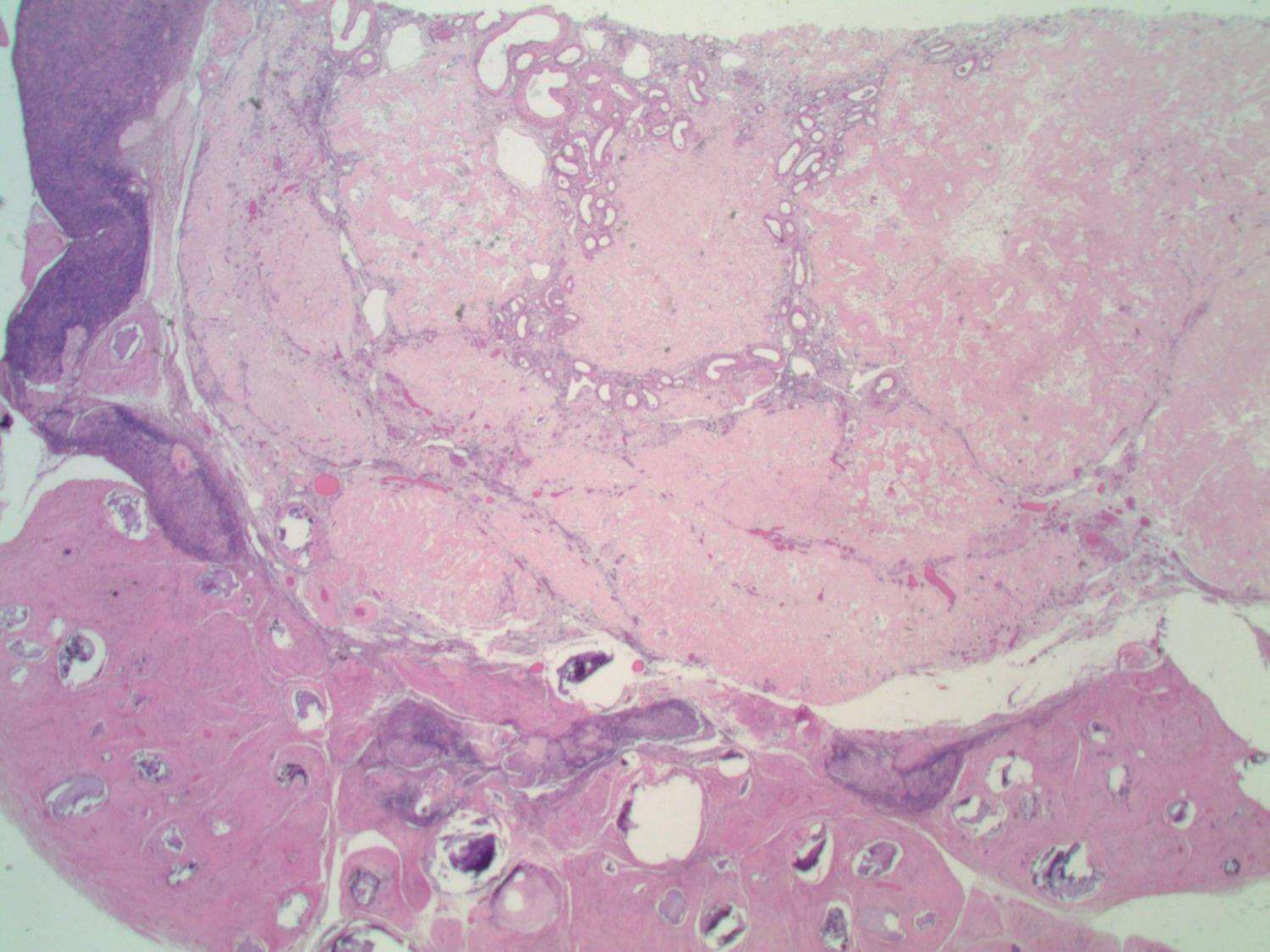


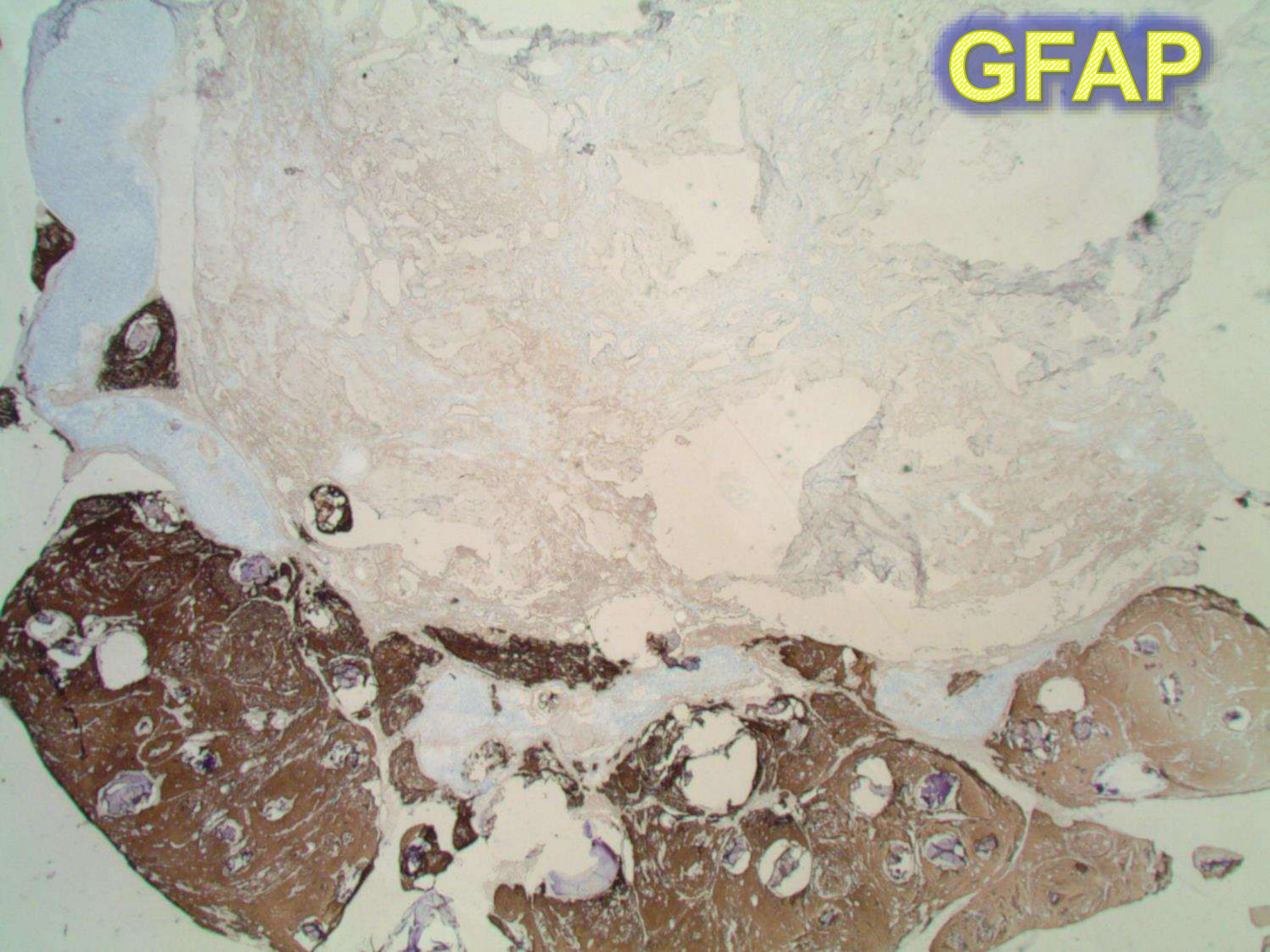


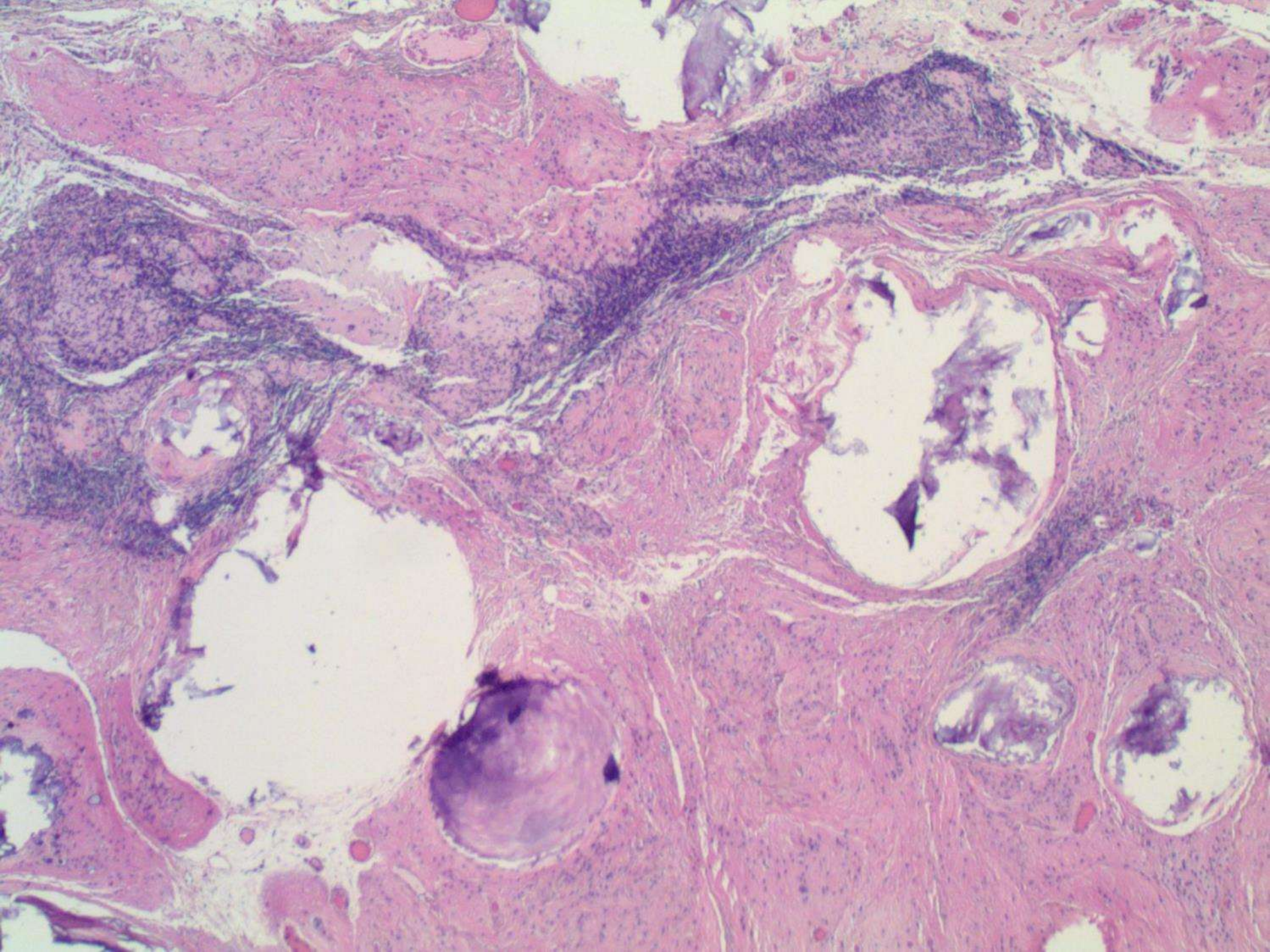


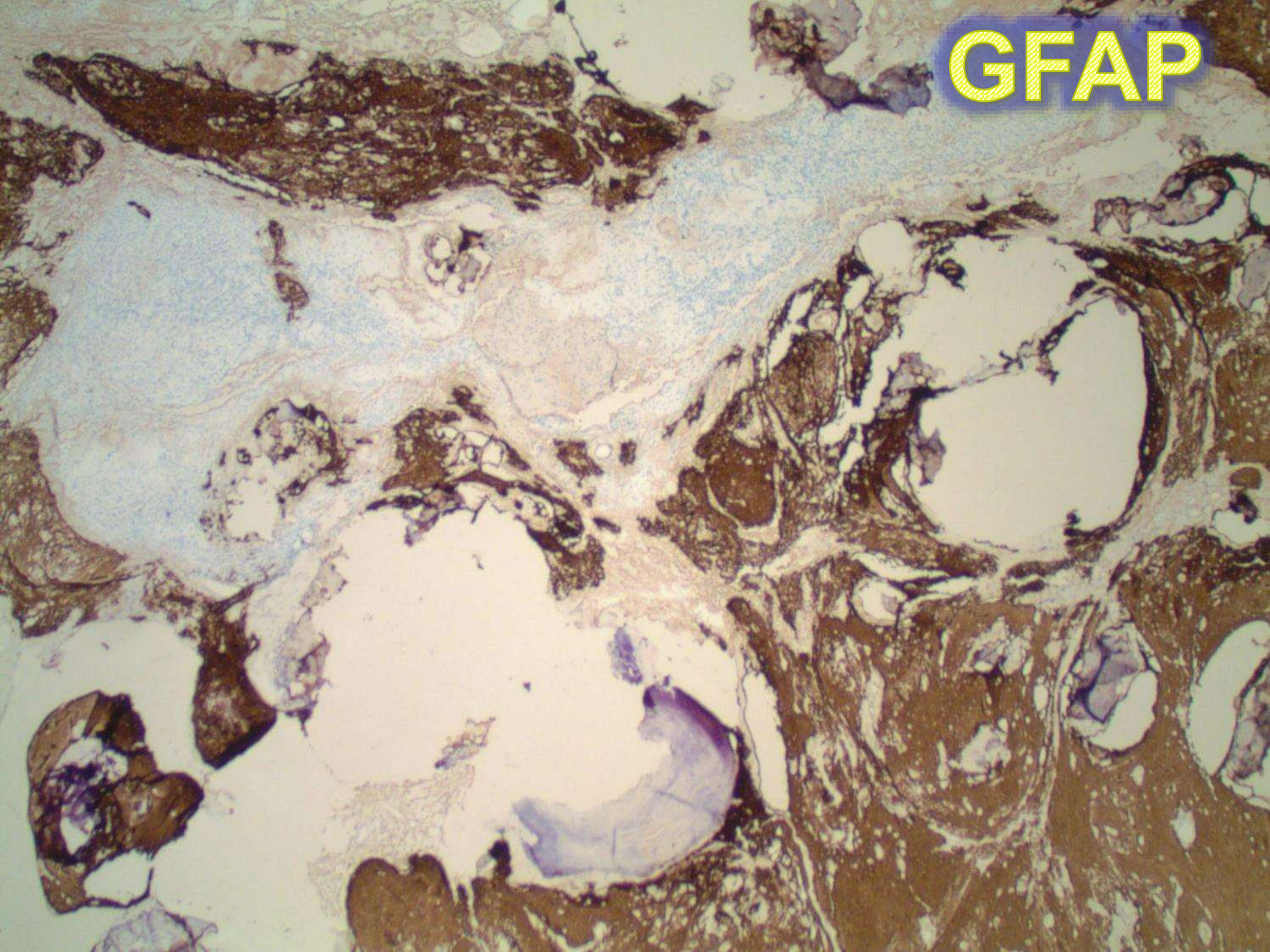







GFAP




GFAP

patient has h/o teratoma!

Dx:
gliomatosis peritonei

- **Rare condition often associated with immature ovarian teratoma or germ cell tumor**
- **Presence of mature glial tissue in peritoneum**
- **Considered grade 0 teratoma per WHO grading for immature teratoma**
- **Presence not usually associated with adverse outcomes**
    - Rarely can transform into malignant glial neoplasms

Gliomatosis peritonei: a clinicopathologic and immunohistochemical study of 21 cases

Li Liang[1], Yifen Zhang[1,2], Anais Malpica[1], Preetha Ramalingam[1], Elizabeth D Euscher[1], Gregory N Fuller[1] and Jinsong Liu[1,3]

[1]Department of Pathology, The University of Texas MD Anderson Cancer Center, Houston, TX, USA; [2]Department of Pathology, Nanjing Drum Tower Hospital, Nanjing University Medical School, Nanjing, People's Republic of China and [3]Department of Pathology, The First Affiliated Hospital of Nanjing Medical University, Nanjing, People's Republic of China

Gliomatosis peritonei, a rare condition often associated with immature ovarian teratoma, is characterized by the presence of mature glial tissue in the peritoneum. We retrospectively evaluated 21 patients with gliomatosis peritonei and studied their clinicopathologic features and immunophenotype. The patients' ages ranged from 5 to 42 years (median, 19 years). Their primary ovarian tumors consisted of immature teratoma ($n=14$), mixed germ cell tumors ($n=6$), and mature teratoma with a carcinoid tumor ($n=1$). Gliomatosis peritonei was diagnosed at the same time as primary ovarian neoplasm in 16 patients and secondary surgery in 5 patients. Also, 11 of 21 patients had metastatic immature teratoma ($n=4$), metastatic mature teratoma ($n=2$), or both ($n=5$). One patient developed glioma arising from gliomatosis peritonei. Seventeen patients had follow-up information and were alive with no evidence of disease ($n=13$), alive with disease ($n=3$), or alive with an unknown disease status ($n=1$). The follow-up durations ranged from 1 to 229 months (mean, 49 months; median, 23 months). Immunohistochemistry results demonstrated that SOX2 was expressed in all cases of gliomatosis peritonei and glioma with tissue available (nine of nine cases), whereas OCT4 and NANOG were negative in all cases with available tissue (eight of eight cases). In conclusion, both gliomatosis peritonei and glioma arising from it show a SOX2+/OCT4−/NANOG− immunophenotype. These findings demonstrated that gliomatosis peritonei is associated with favorable prognosis, although it is important to rule out potentially associated immature teratoma and malignant transformation. SOX2 may have an important role in the development of gliomatosis peritonei.

Modern Pathology (2015) **28**, 1613–1620; doi:10.1038/modpathol.2015.116; published online 13 November 2015

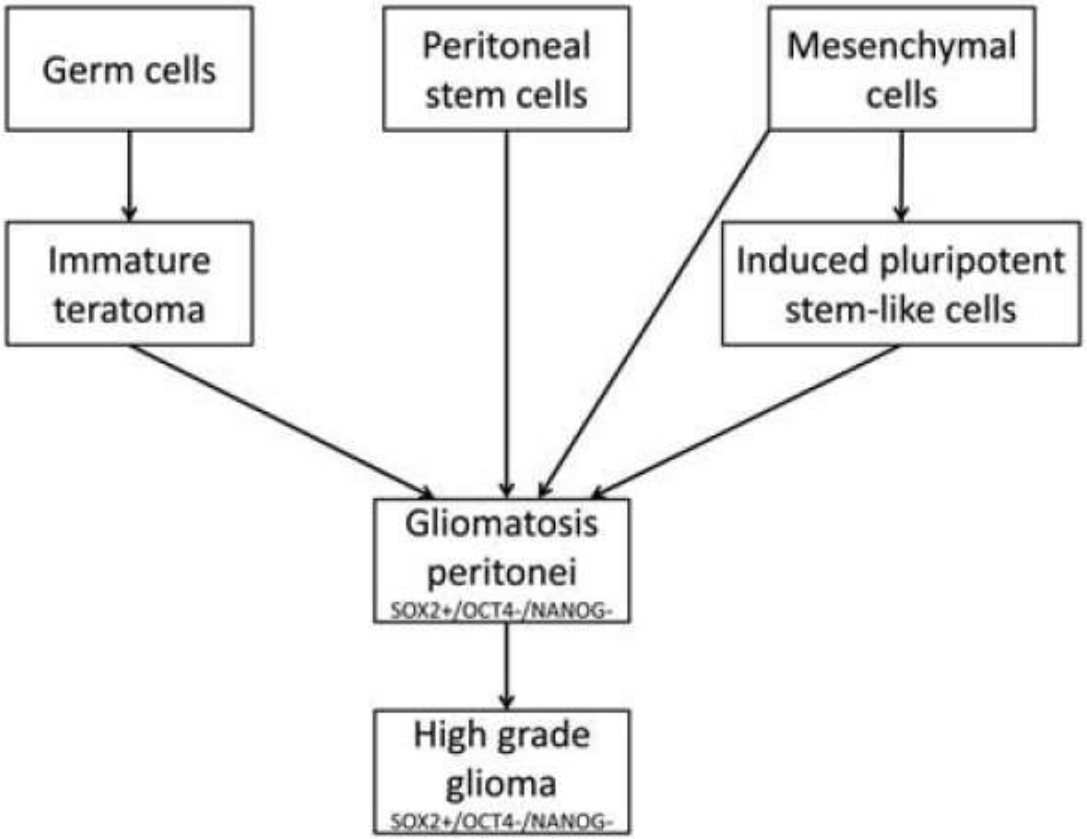


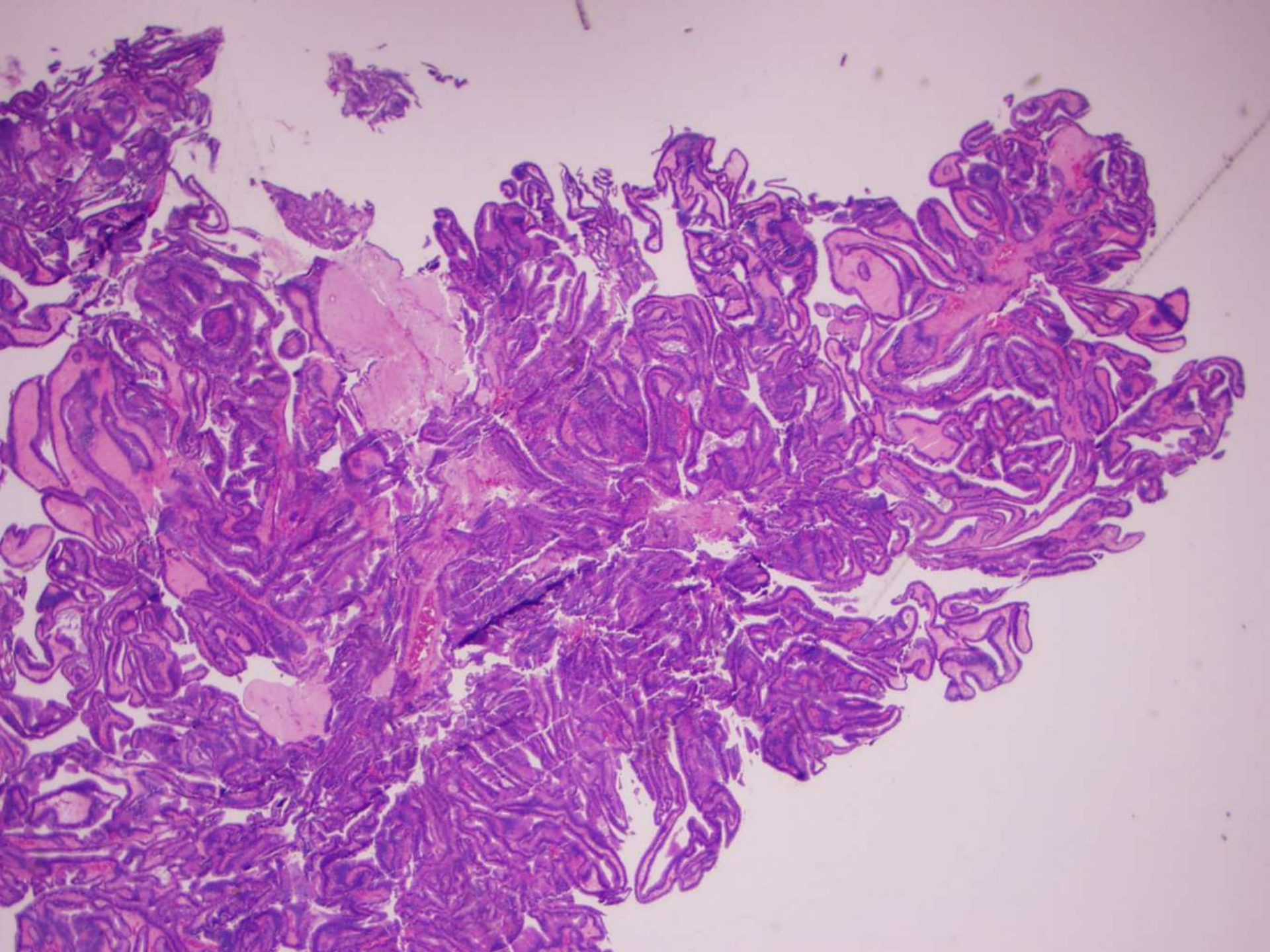
Figure 3 Hypotheses regarding the origin of gliomatosis peritonei. First, it may derive from an immature teratoma that undergoes maturation or from cancer stem cells within an immature teratoma. Second, it may derive from peritoneal stem cells that differentiate toward the neural lineage induced by factors secreted by teratoma. Third, it may derive from subperitoneal mesenchymal cells that transdifferentiate into glial cells either directly or through an intermediate stage of induced pluripotent stem-like cells.

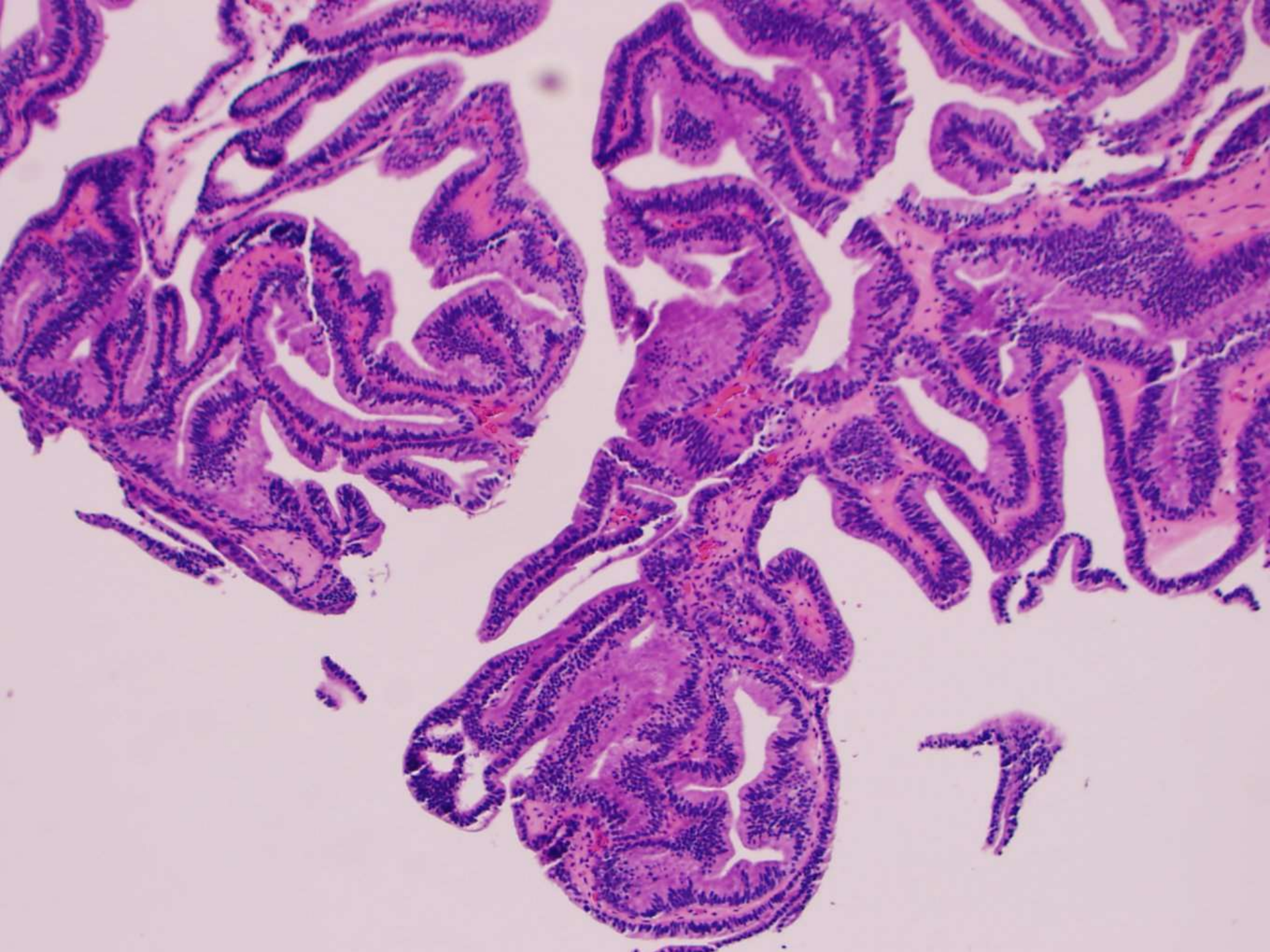
MODERN PATHOLOGY (2015) 28, 1613–1620

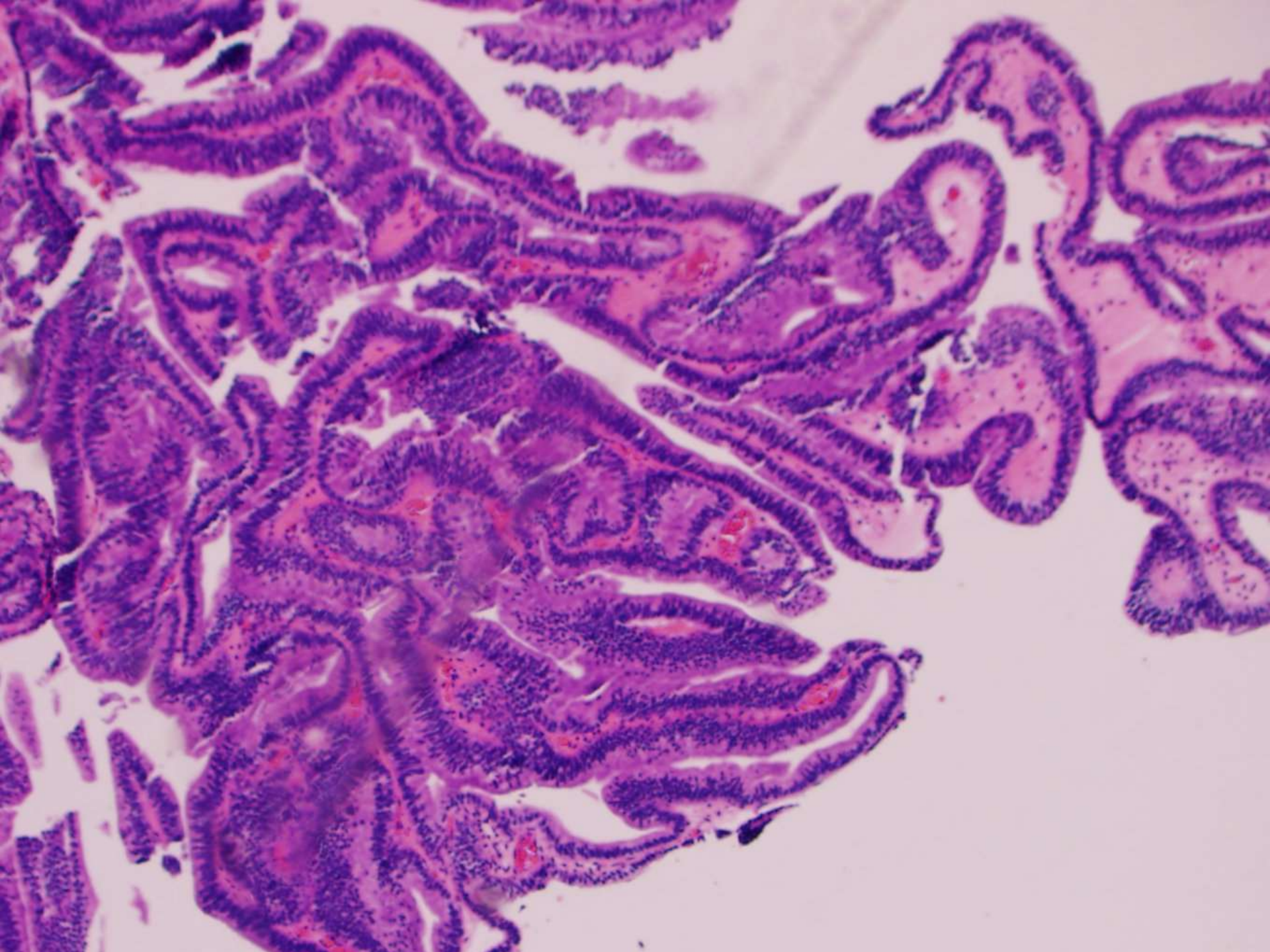
20-0410

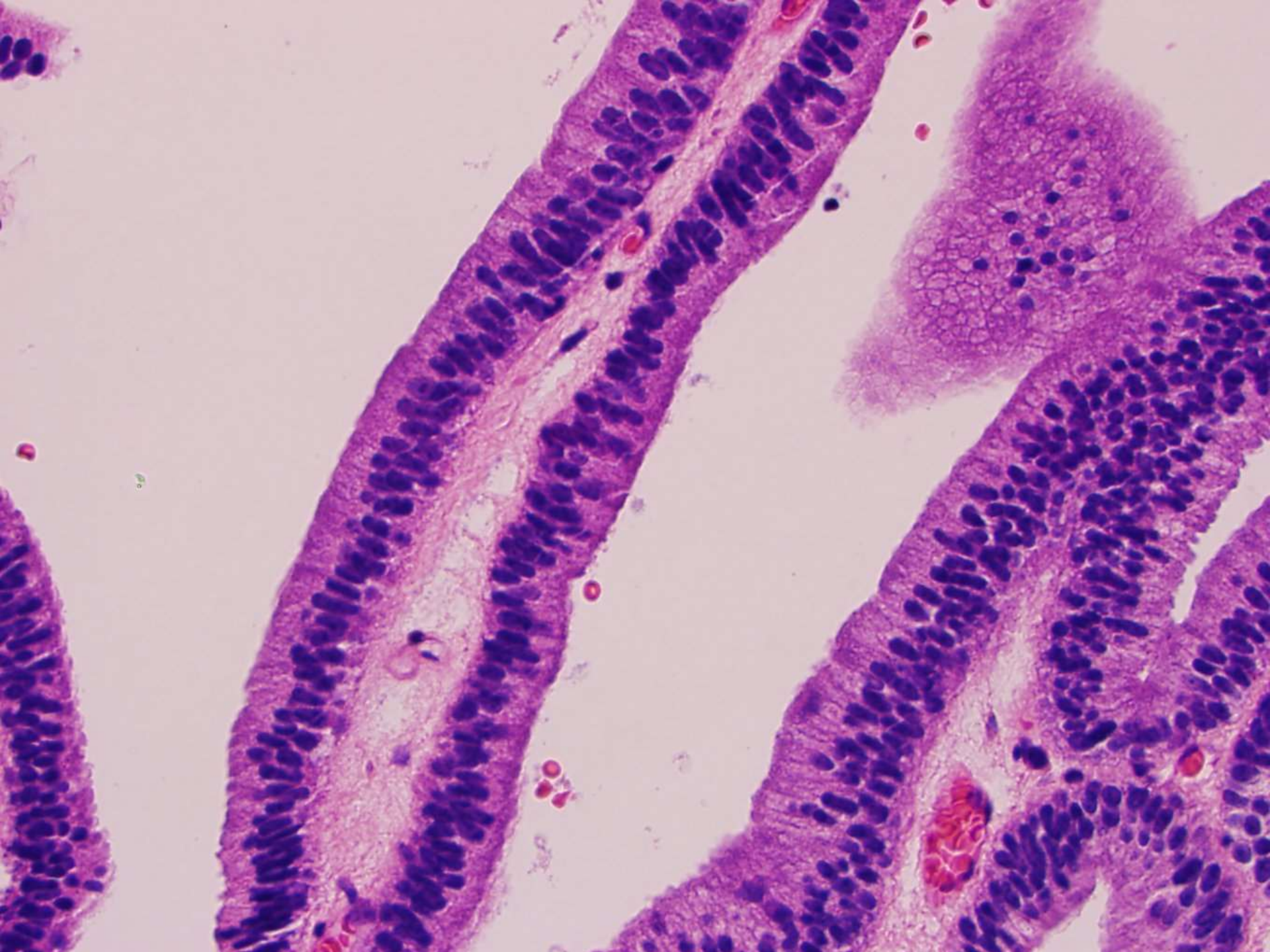
Greg Rumore; Kaiser Diablo

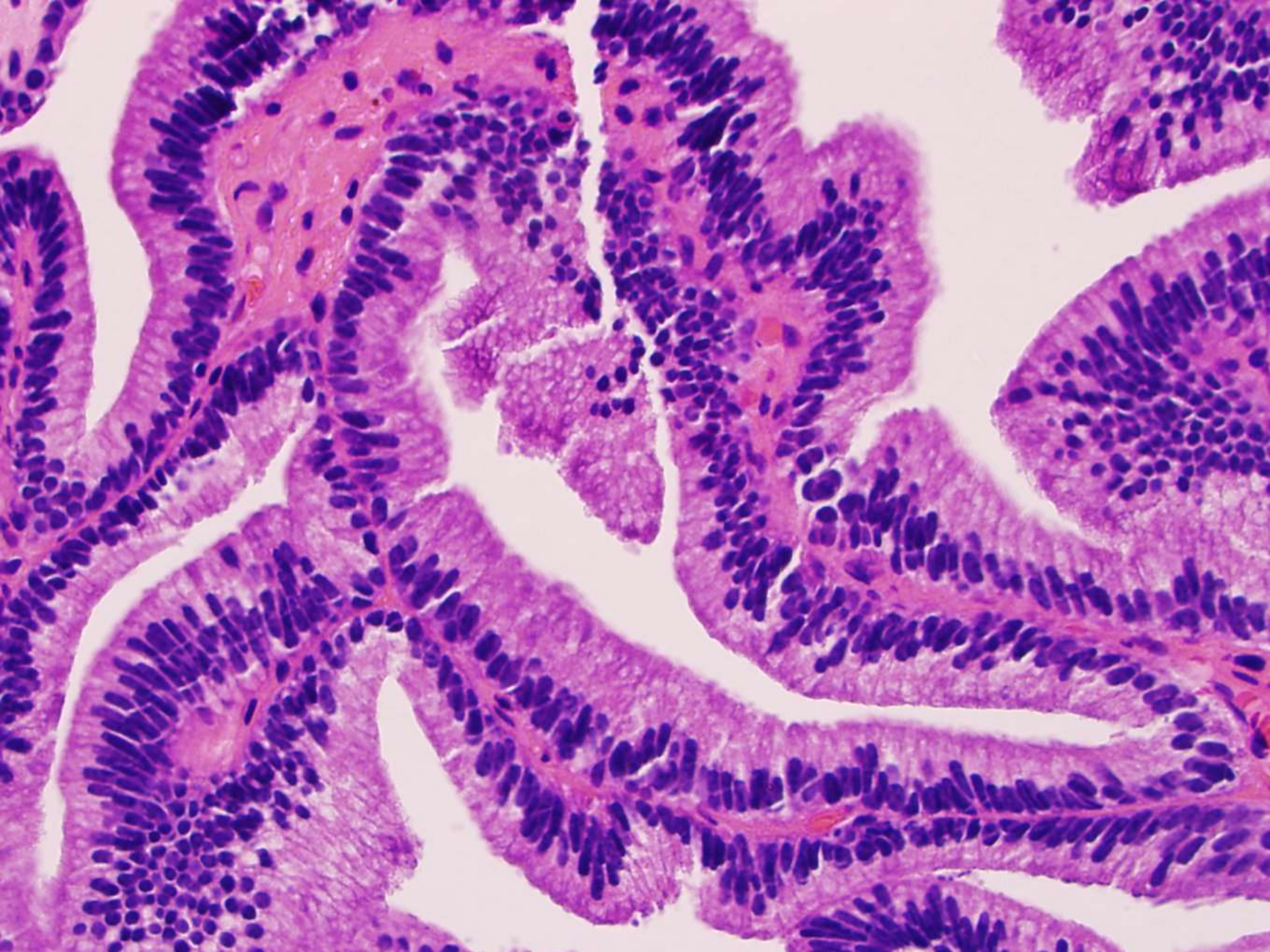
84-year-old M with h/o 3+3 prostate cancer, now has 0.5cm papillary lesion with “fronds” in prostatic urethra. TURP performed.

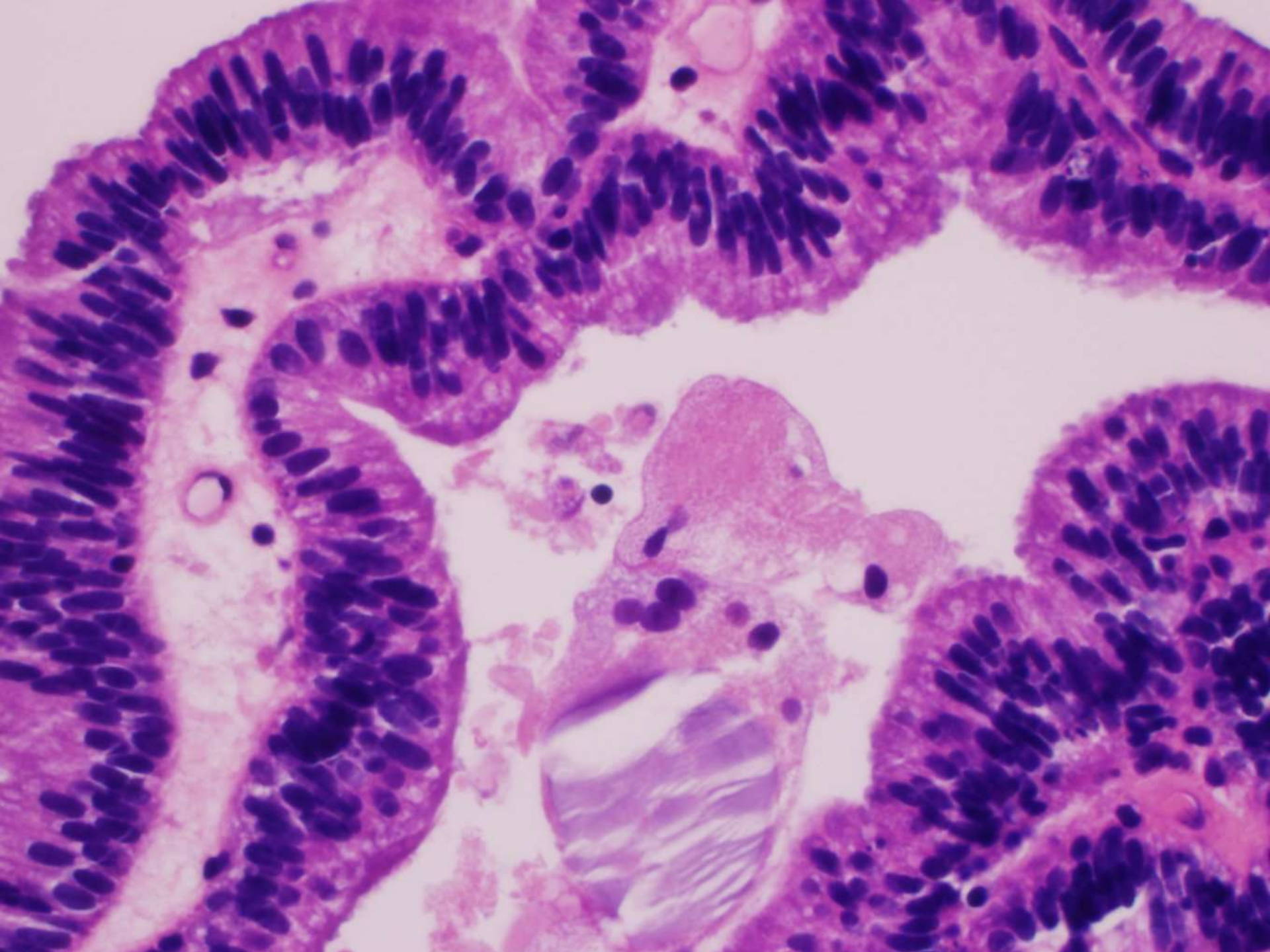






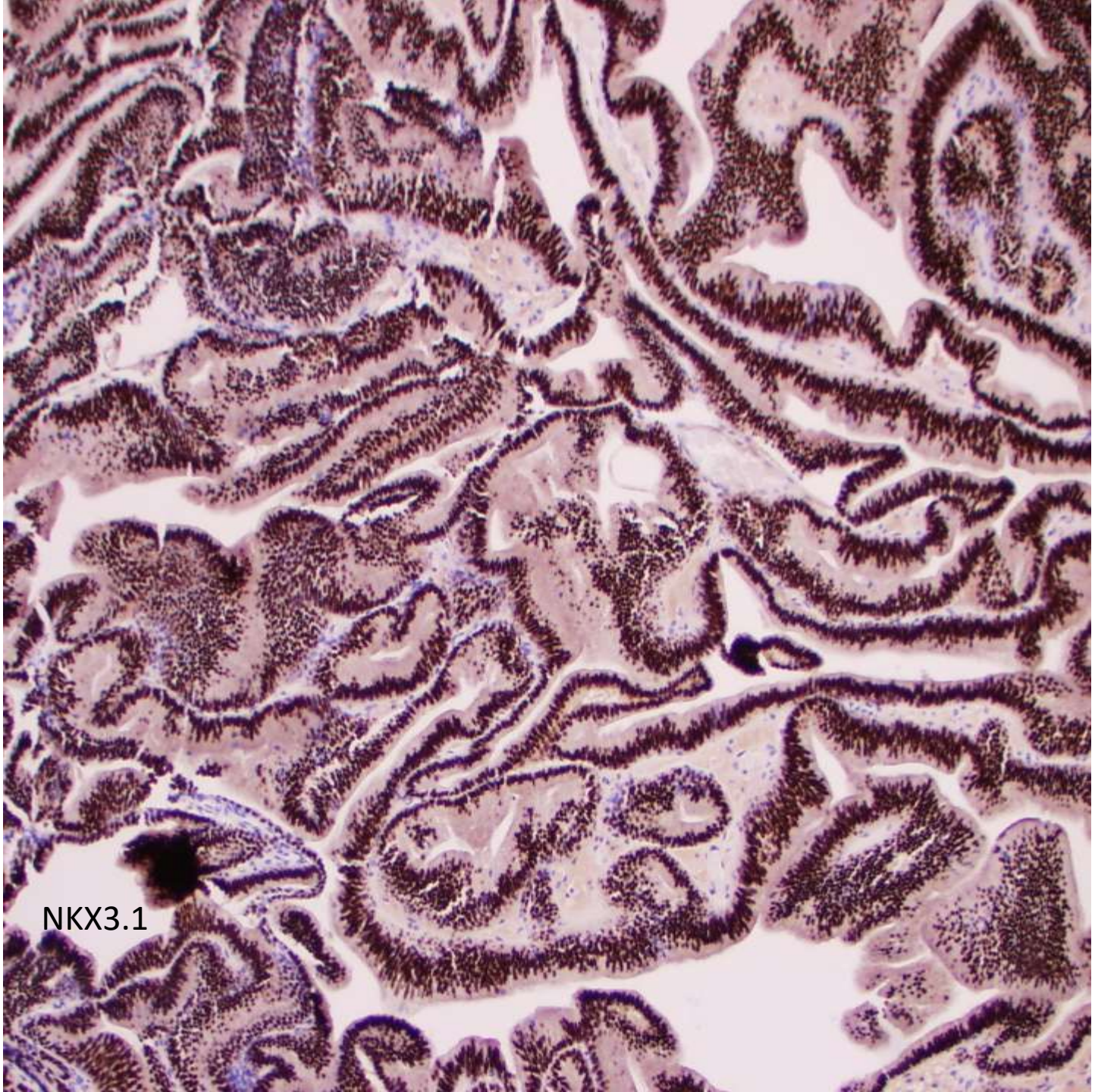


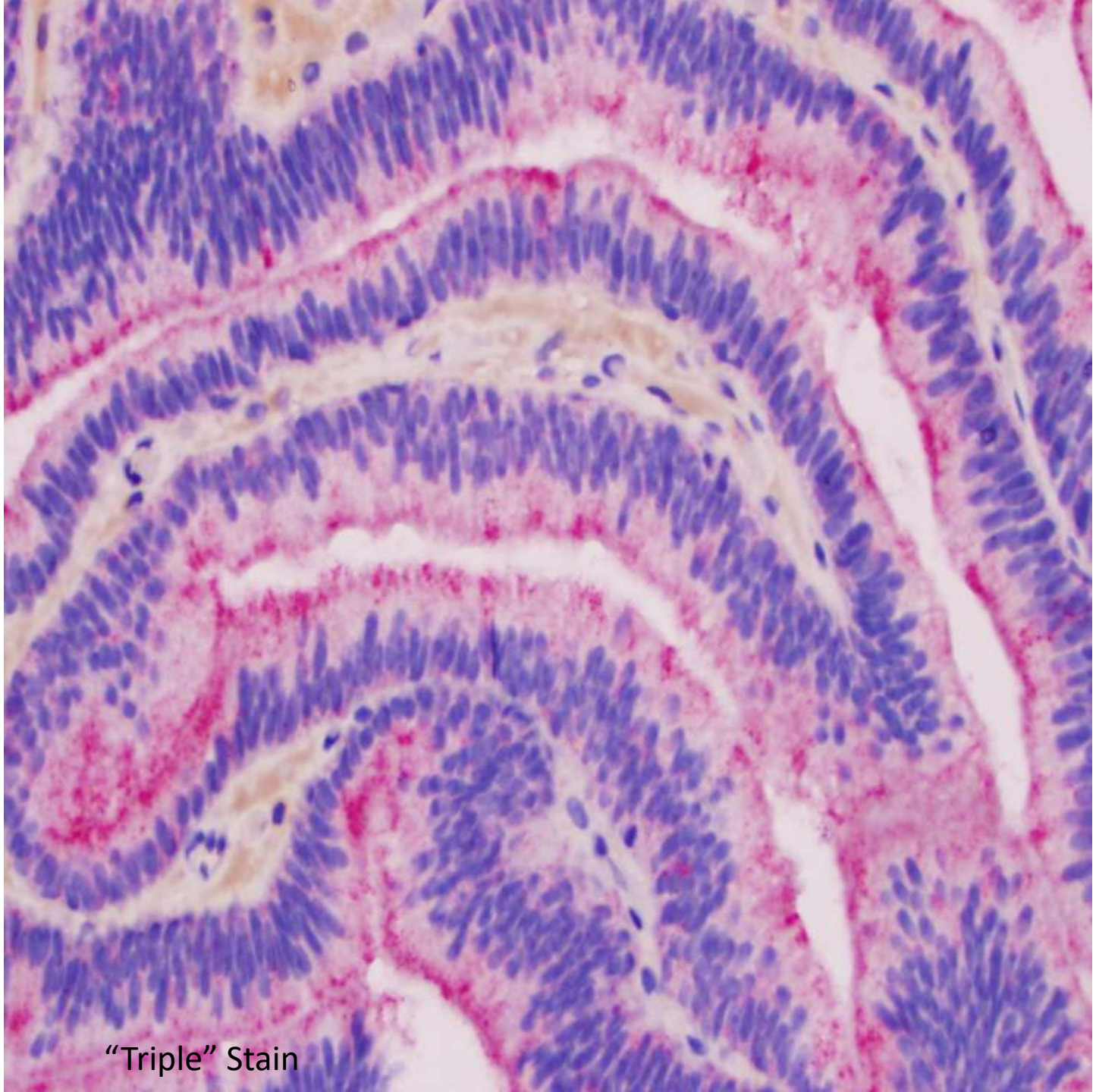




Differential Diagnosis

- Urethral Polyps-fibrous or prostatic
- Adenoma
- Primary Urothelial Carcinoma
- Prostate Ductal Carcinoma


NKX3.1


“Triple” Stain

Prostate Duct Carcinoma of Urethra

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Ductal adenocarcinoma of the prostate diagnosed on transurethral biopsy or resection is not always indicative of aggressive disease: implications for clinical management

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Study Type – Prognosis (case series)
Level of Evidence 4

OBJECTIVE

To report the clinicopathological characteristics of 23 cases of ductal adenocarcinoma of the prostate (DCP) and discuss the implications for clinical management, as DCP is considered an aggressive subtype of prostate adenocarcinoma (PA).

PATIENTS AND METHODS

transurethral resection (TURP) in the present era of screening with prostate-specific antigen (PSA) is unclear. The study included 23 cases of pure DCP without acinar PA diagnosed on UB or TURP. Demographic information, serum PSA level, follow-up surgical procedures (RP, TURP or TRUSB) and outcome data were collected.

RESULTS

The mean age of the men was 67.5 years and the mean PSA level before the procedure was 12.5 ng/mL; 14 cases were detected on UB and nine were diagnosed on TURP. The mean

Gleason score ≥8 PA. Extraprostatic extension, seminal vesicle invasion and regional lymph node metastasis were present in seven, six and two cases, respectively.

CONCLUSIONS

Most DCP diagnosed on UB or TURP in this contemporary series was associated with aggressive PA, but a subset presented as a small periurethral tumour with no concomitant acinar PA, and was eradicated by the initial biopsy/TURP alone. We recommend that patients with a diagnosis of

Prostate Duct Carcinoma

- Usually regarded as aggressive tumor-diagnosed on TRB
- Can also arise in and around verumontanum-friable polypoid lesions
- If it arises from periurethral glands with no acinar component, may behave less aggressively
- May be eradicated by biopsy alone
- Recommend repeat TUR and TRB prior to radical prostatectomy on tumors diagnosed on TUR and Urethral biopsies