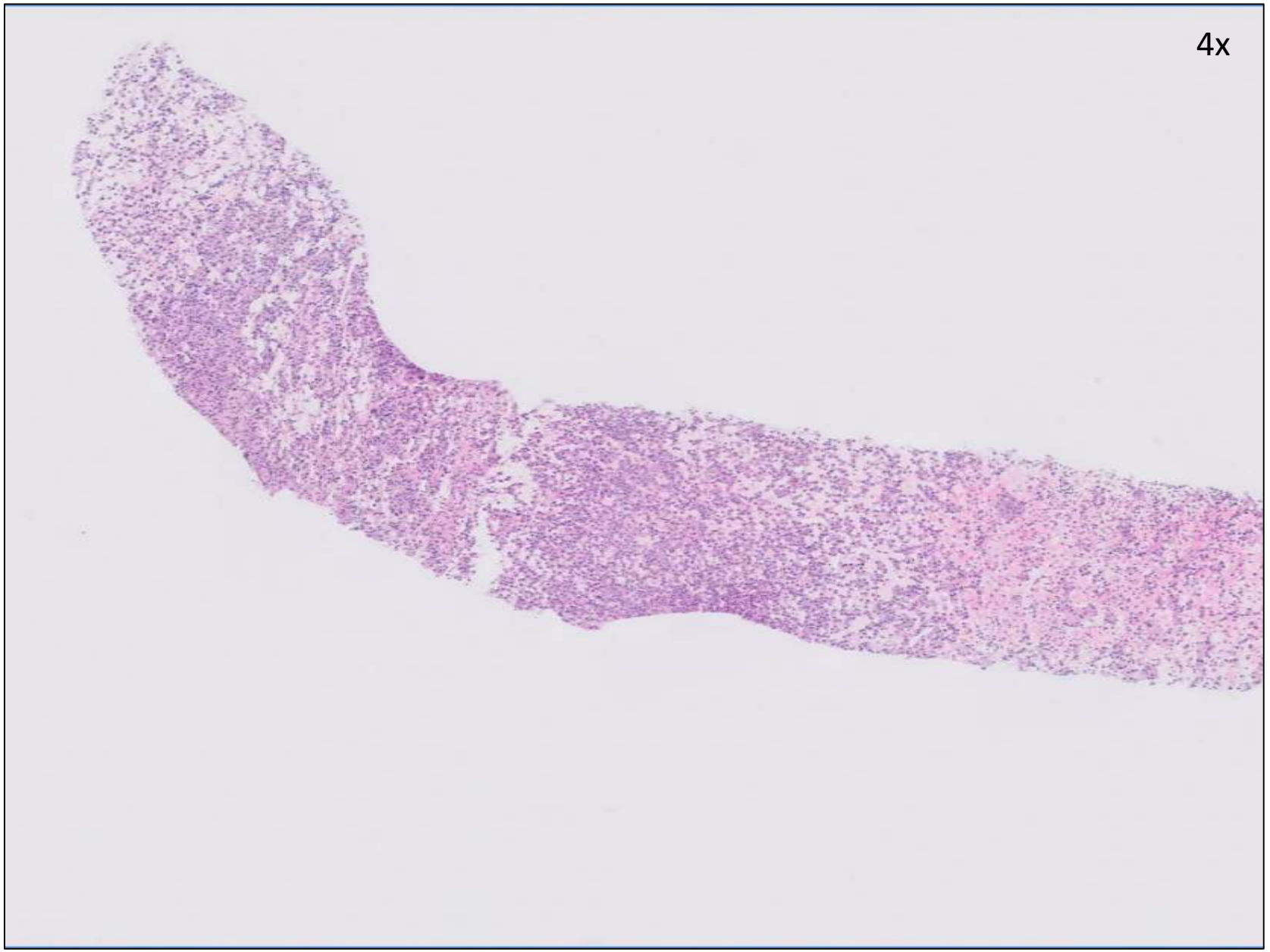
FEB 2023 DIAGNOSIS LIST

23-0201: juxtaglomerular cell tumor (kidney; GU path)
23-0202: solid pseudopapillary tumor (ovary; GYN path)
23-0203: Russell body gastritis (stomach; GI path)
23-0204: trabecular carcinoid tumor (ovary; GYN path)
23-0205: spindle cell lipoma (soft tissue; BST path)
23-0206: acinar cell carcinoma (pancreas; Gipath)
23-0207: papillary renal cell carcinoma, microcystic pattern (kidney; GU path)
23-0208: sloughed small blue rete testis cells (spermatocele; GU path)

23-0201

Kevin Lu/Emily Chan; UCSF

30ish F with 4cm left renal mass, biopsy performed.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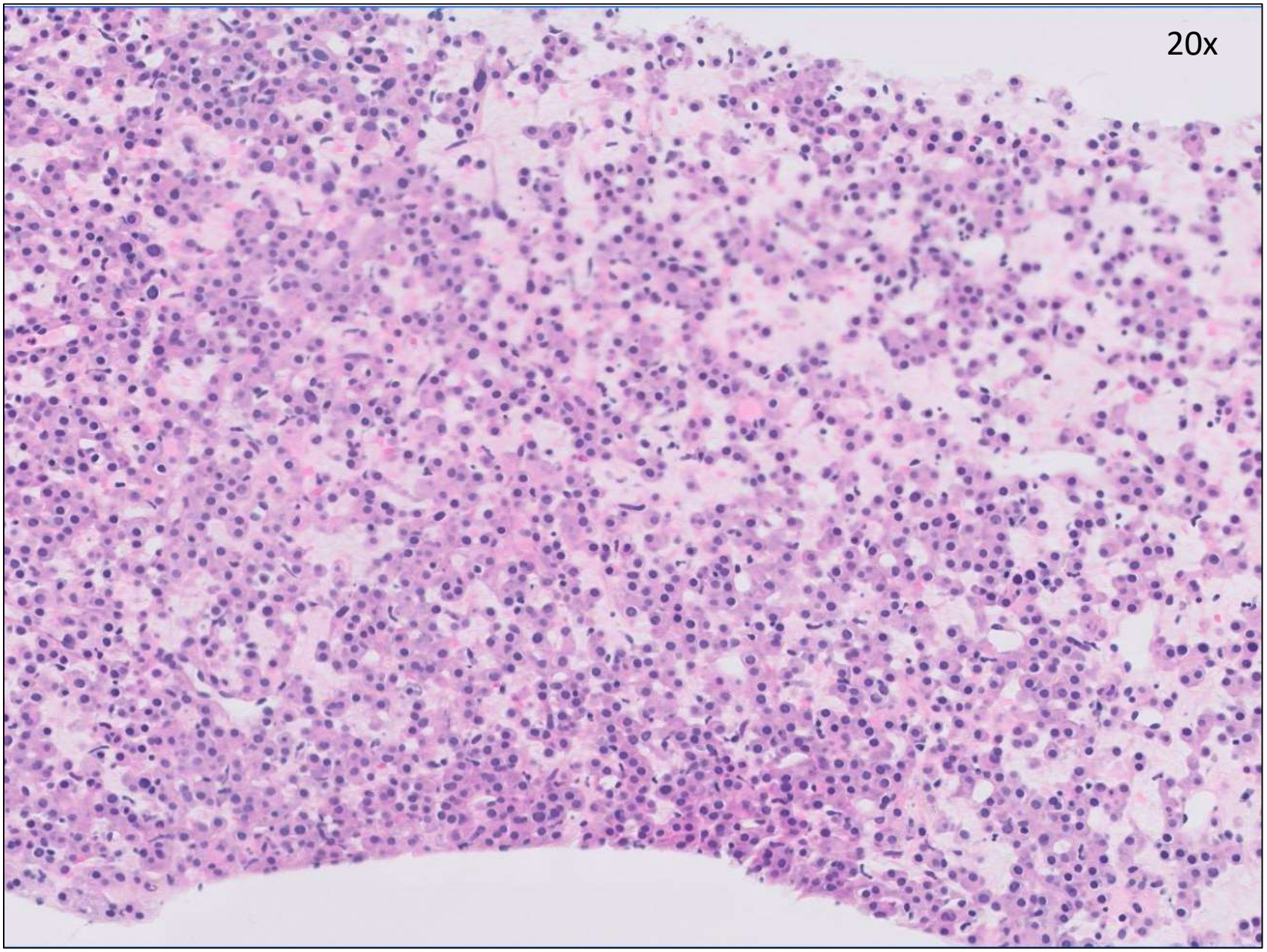
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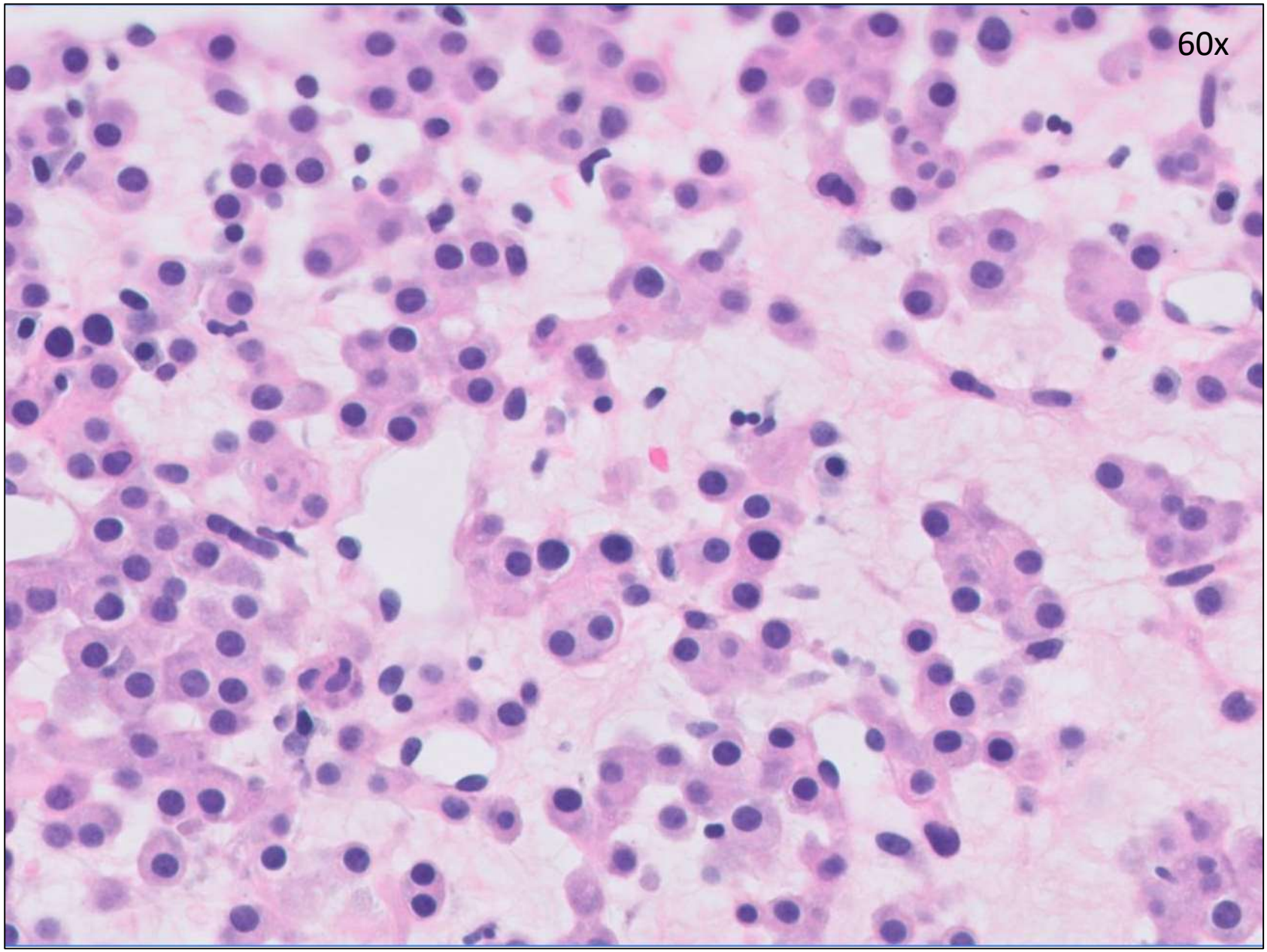

60x

DIAGNOSIS?

30ish woman with a 4 cm left kidney mass.

Kevin Lu, Emily Chan

UCSF

South Bay Pathology Society Monthly Meeting

February 6, 2023

Acknowledgements:

- Dr. Baorong Chen (Pathology Associates) and others who send us consults!
- Dr. Laura Brown (UCSF Hematopathology)

Differential diagnosis:

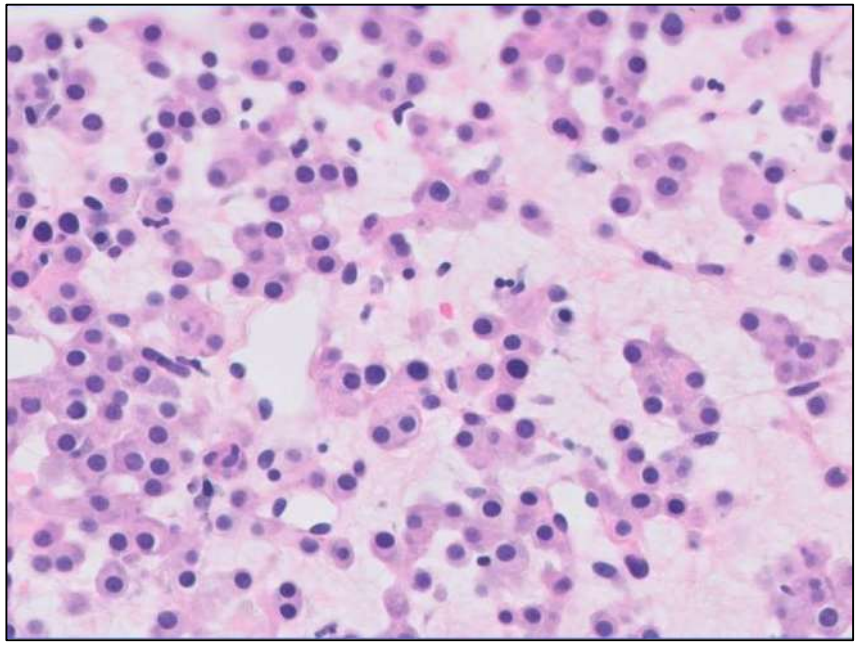
Discohesive, monotonous bland round cells, oncocytic, myxoid stroma, ?

- Renal tumor (Oncocytoma? RCC? Translocation?)
- Adrenocortical tumor
- Neuroendocrine tumor
- Paraganglioma
- Hematolymphoid

  …

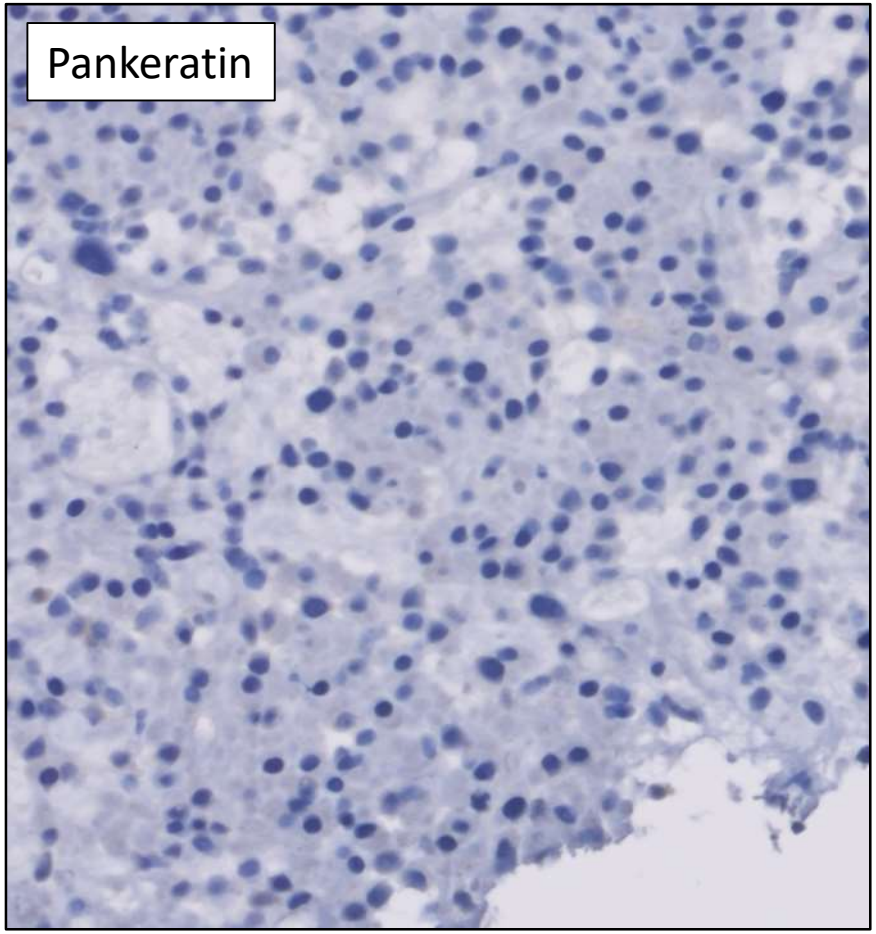
  …

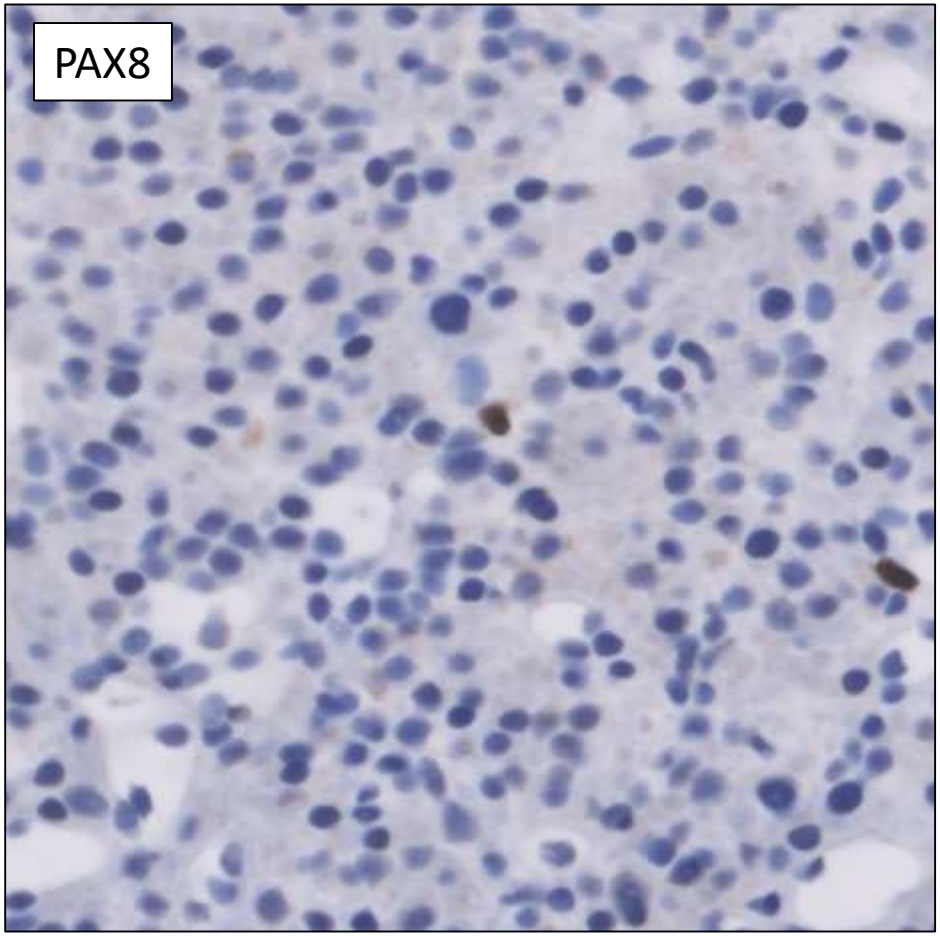
  …



- Soft tissue tumors? Pericytic tumor?

IHC Round 1 (provided)





Other negative IHC

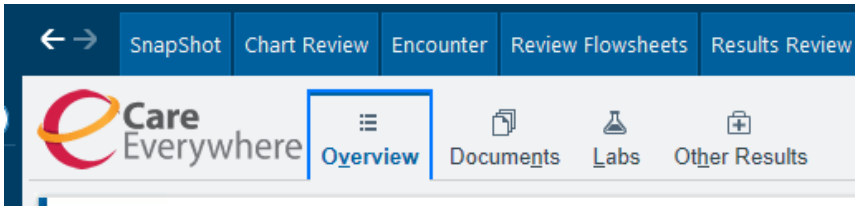
- CK7
- CD117 (scattered mast cells)
- MiTF
- HMB45, Melan A, S-100
- SMA
- Inhibin
- Chromogranin, synaptophysin (equivocal)

IHC Round 2:
Plasmacytoid? Lymphoma? Adrenocortical?

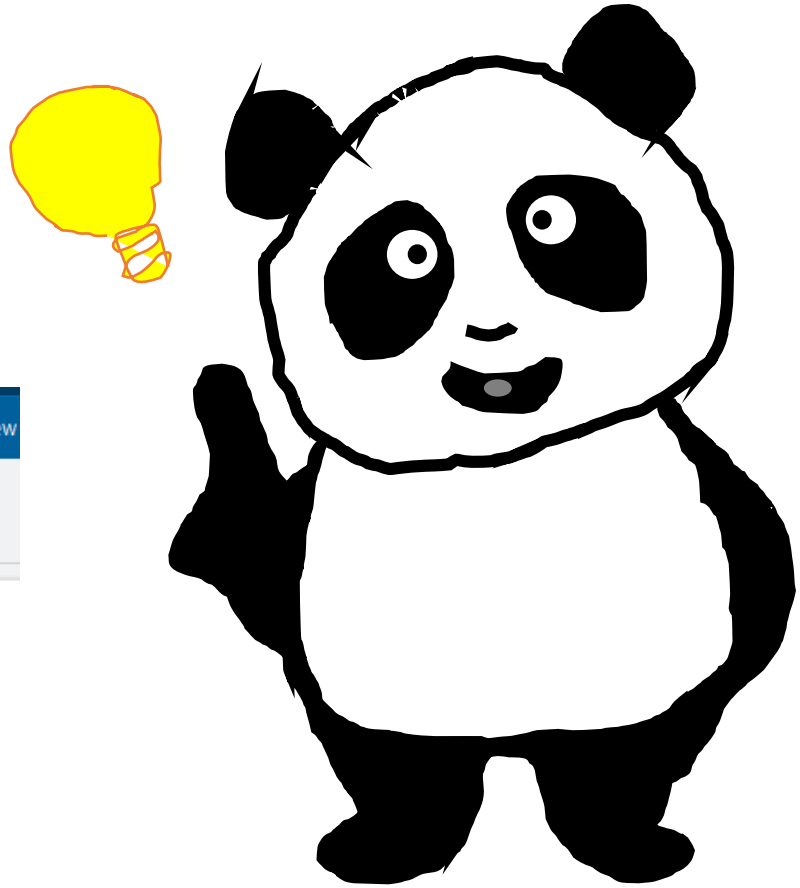
Additional negative IHC:

- CD45
- CD79a
- CD138, MUM1
- SF-1
- SALL4
- GATA3 +/-

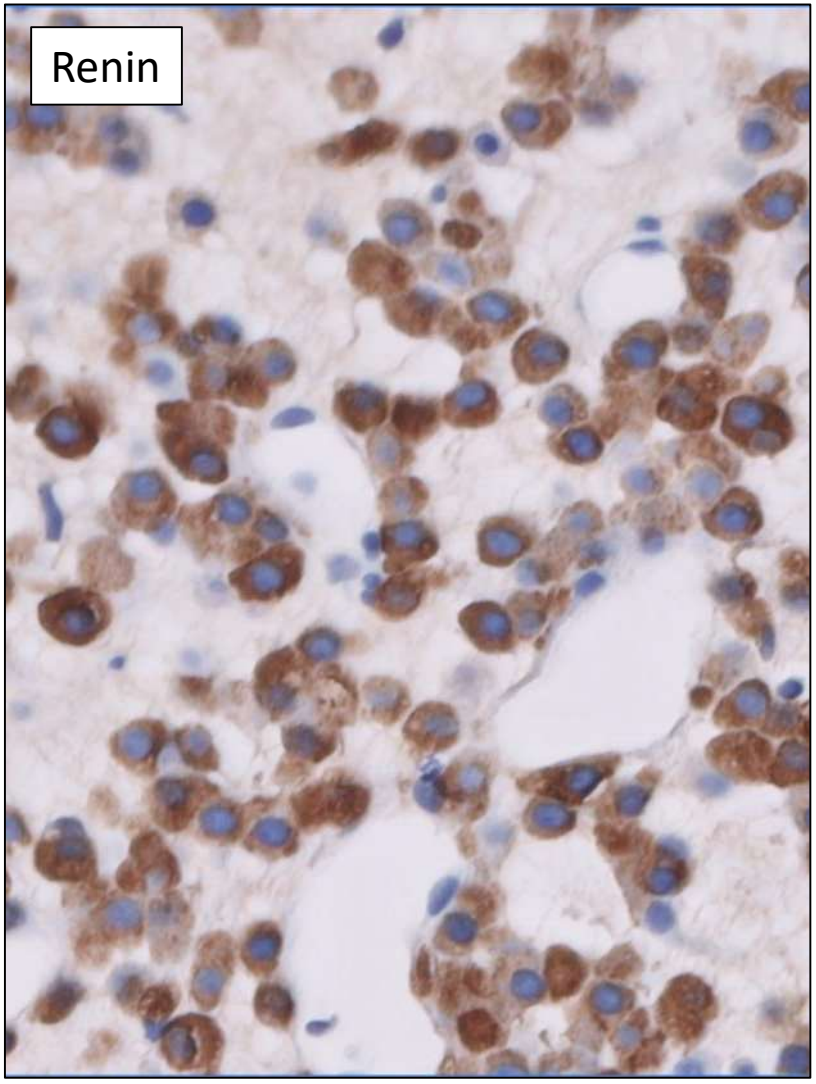
- Young patient
- Kidney tumor

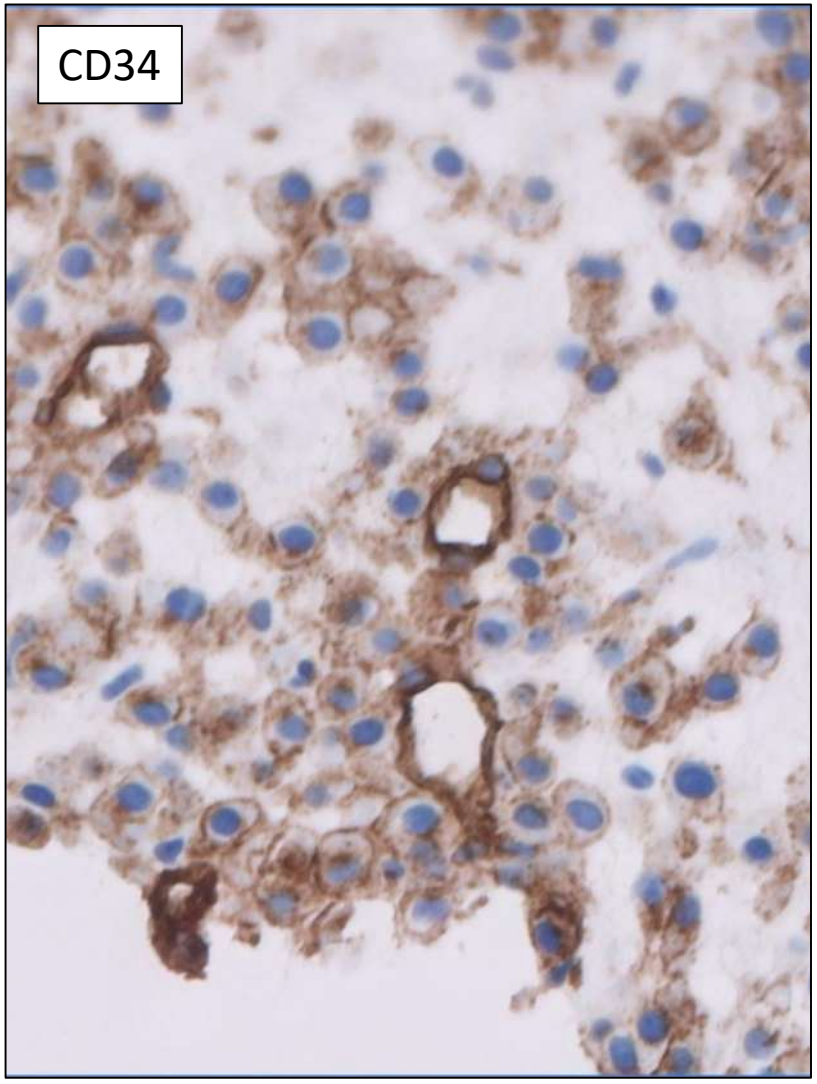


- Refractory HTN and electrolyte abnormalities



What about a renin/aldosterone secreting tumor?


Renin

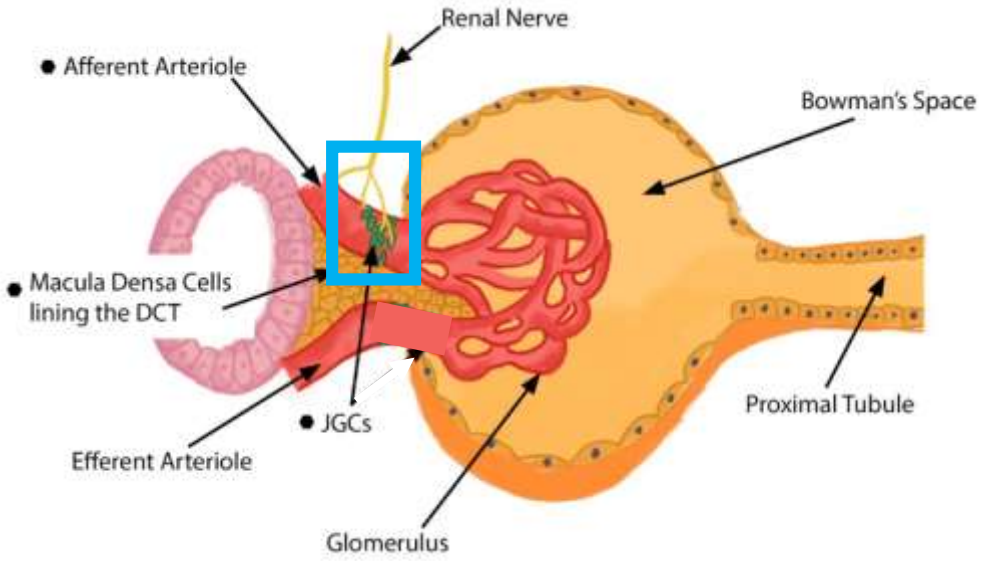

CD34

Diagnosis:

Juxtaglomerular cell tumor

Juxtaglomerular cell tumor (Reninoma)

- Pericytic renin-secreting tumor (perivascular mesenchymal cell origin, afferent arteriole)



Juxtaglomerular cell tumor (Reninoma)

- Pericytic renin-secreting tumor (perivascular mesenchymal cell origin, afferent arteriole)
- Clinical presentation is key:
  - Young (median age: 27)
  - Hyperreninism, hyperaldosteronism
  - Medically refractory HTN
  - Most clinical symptoms should resolve with resection

Human Pathology (2022) 128, 110–123

Human PATHOLOGY
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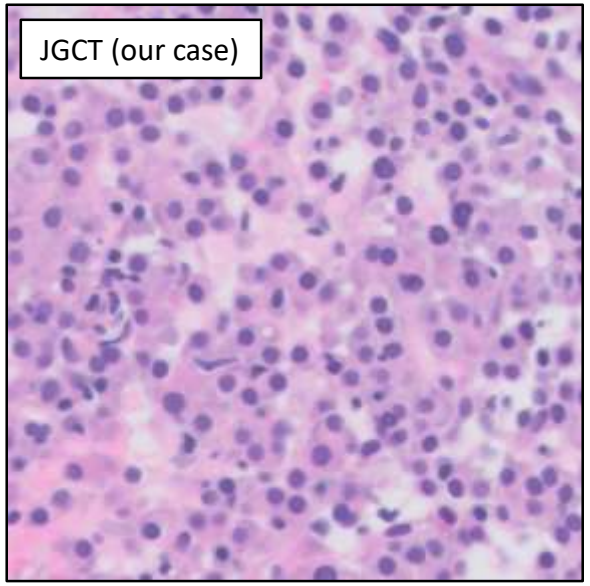
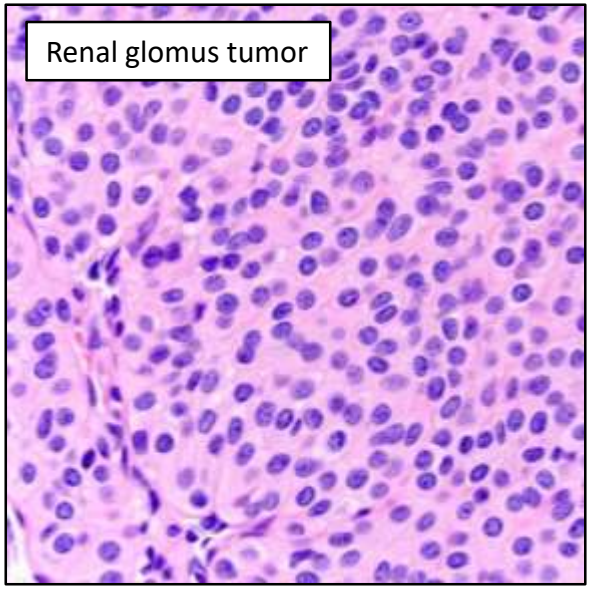
Original contribution

Immunohistochemical expression of renin and GATA3 help distinguish juxtaglomerular cell tumors from renal glomus tumors☆

Sounak Gupta MBBS, PhD[a,*], Andrew L. Folpe MD[a], Jorge Torres-Mora MD[a], Victor E. Reuter MD[b], Jonathan E. Zuckerman MD, PhD[c], Nadja Falk MD[d], Melissa L. Stanton MD[e], Selvaraj Muthusamy MD, PhD[f], Steven C. Smith MD[f], Vidit Sharma MD[g], Sanjeev Sethi MD, PhD[a], Loren Herrera-Hernandez MD[a], Rafael E. Jimenez MD[a], John C. Cheville MD[a]

[a]Department of Laboratory Medicine and Pathology, Mayo Clinic, Rochester, MN, 55905, USA





Human Pathology (2017) 64, 106–117

Human PATHOLOGY
www.elsevier.com/locate/humpath

Original contribution

Pericytic tumors of the kidney—a clinicopathologic analysis of 17 cases☆,☆☆

Deepika Sirohi MD[a,1], Steven C. Smith MD, PhD[b,1], Jonathan I. Epstein MD[c], Bonnie L. Balzer MD, PhD[a], Jeffry P. Simko PhD, MD[d], Dana Balitzer MD[d], Jamal Benhamida MD[d], Oleksandr N. Kryvenko MD[e], Nilesh S. Gupta MD[f], Swetha Paluru MD[c], Isabela Werneck da Cunha MD, PhD[g], Daniel N. Leal AAS,BS[a], Sean R. Williamson MD[f], Mariza de Peralta-Venturina MD[a], Mahul B. Amin MD[a,h,*]

[a]Department of Pathology and Laboratory Medicine, Cedars-Sinai Medical Center, Los Angeles, CA, 90048, USA

Diagnostic features of JGC tumors

- Round/polygonal monomorphic cells, bland nuclei, variable stroma, mast cell infiltrate
- Histologic ddx: Glomus tumor (GT)

	JGC	GT
Median age	27	51
Clinical symptoms	+	–
Renin	+++	+/-
CD34	+++	+
GATA3	+/-	–
SMA	+	+

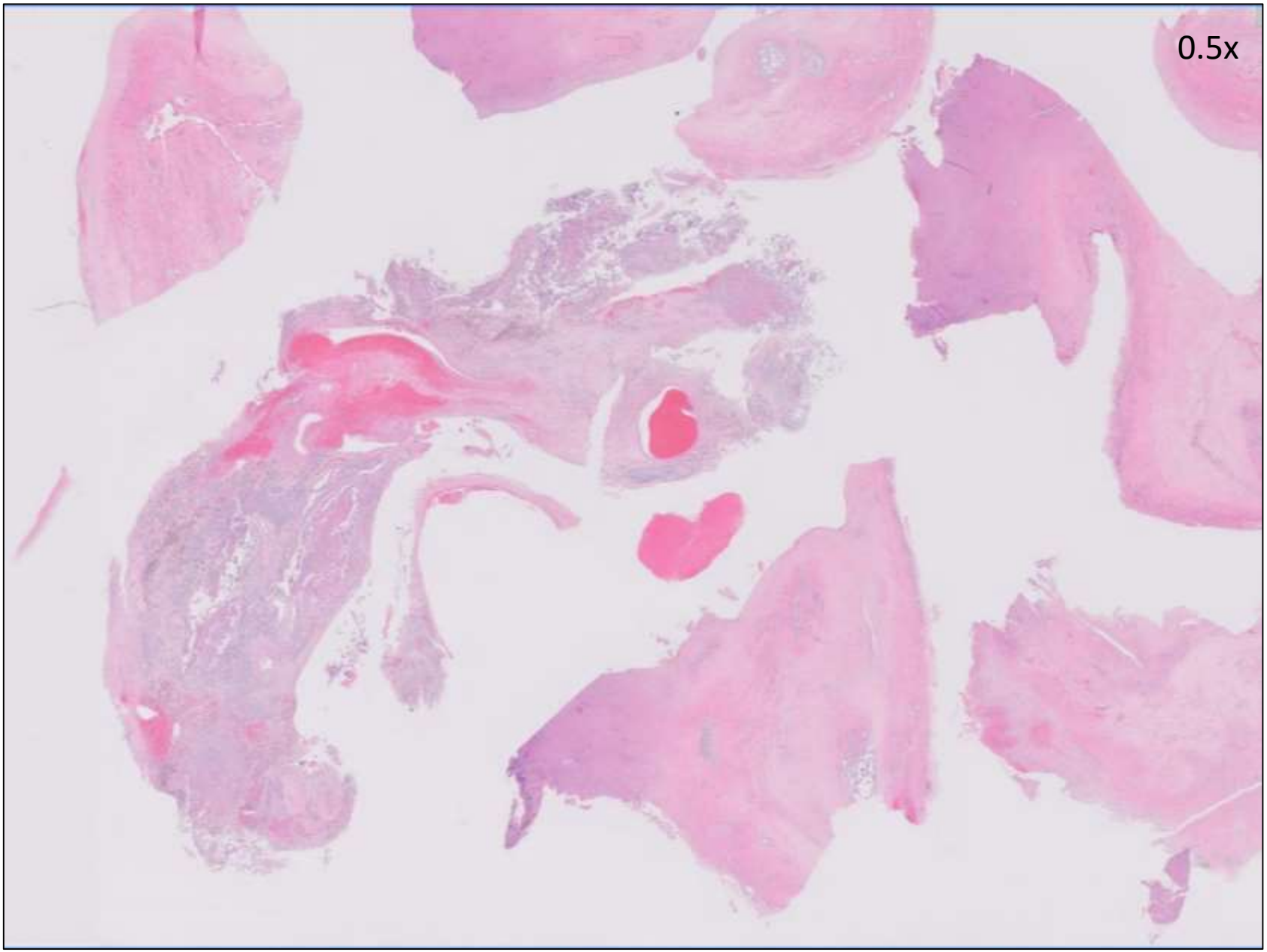
Take home points:

- In a young patient with refractory HTN, electrolyte abnormalities, serum renin/aldosterone…consider JGC tumors
- Clinical context is key!
- IHC:
  - **Strong** renin, CD34
  - Positive but variable SMA and GATA3

23-0202

Kevin Lu/Cynthia Gasper; UCSF

Teenage F with left ovarian cyst.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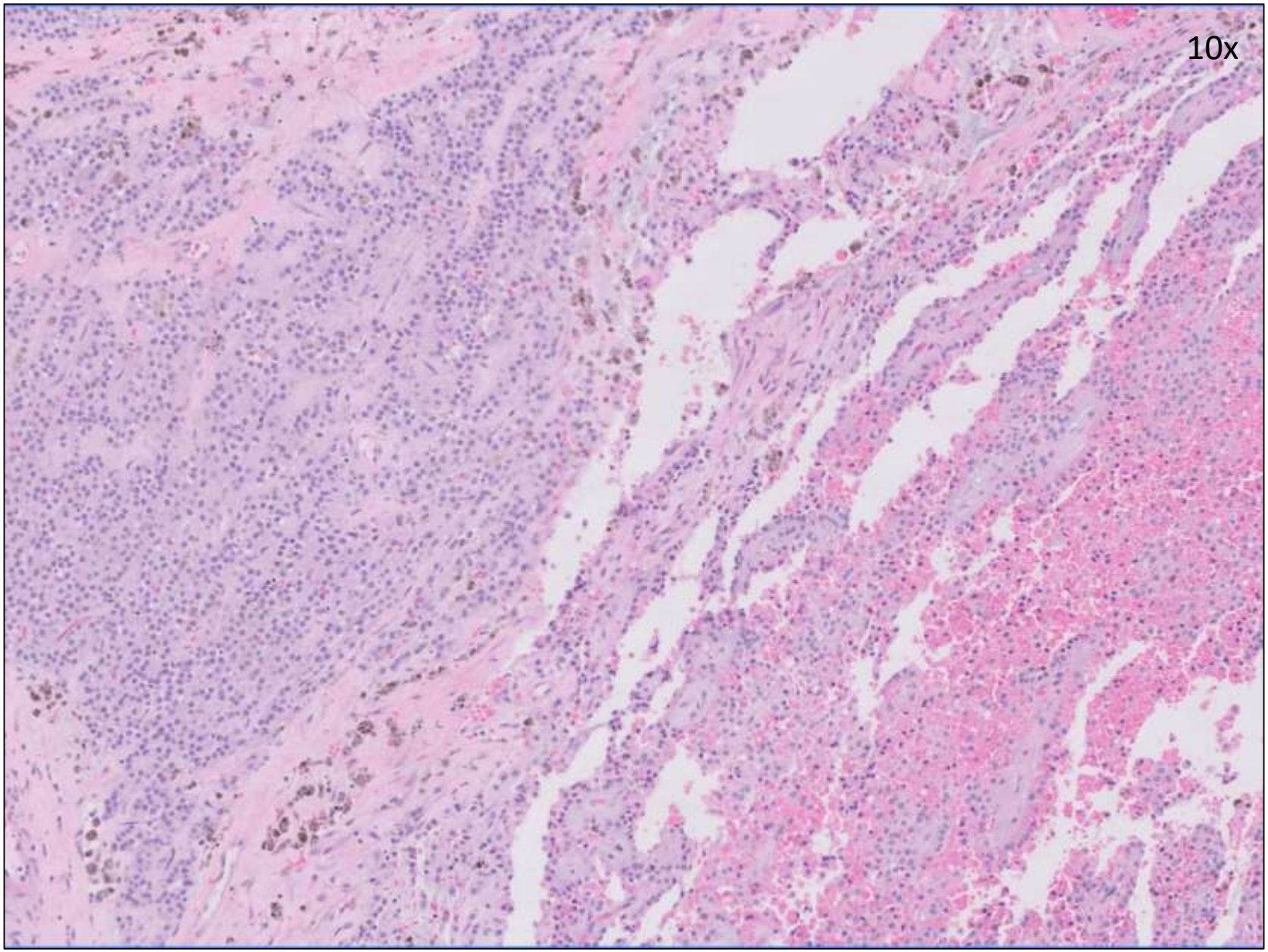
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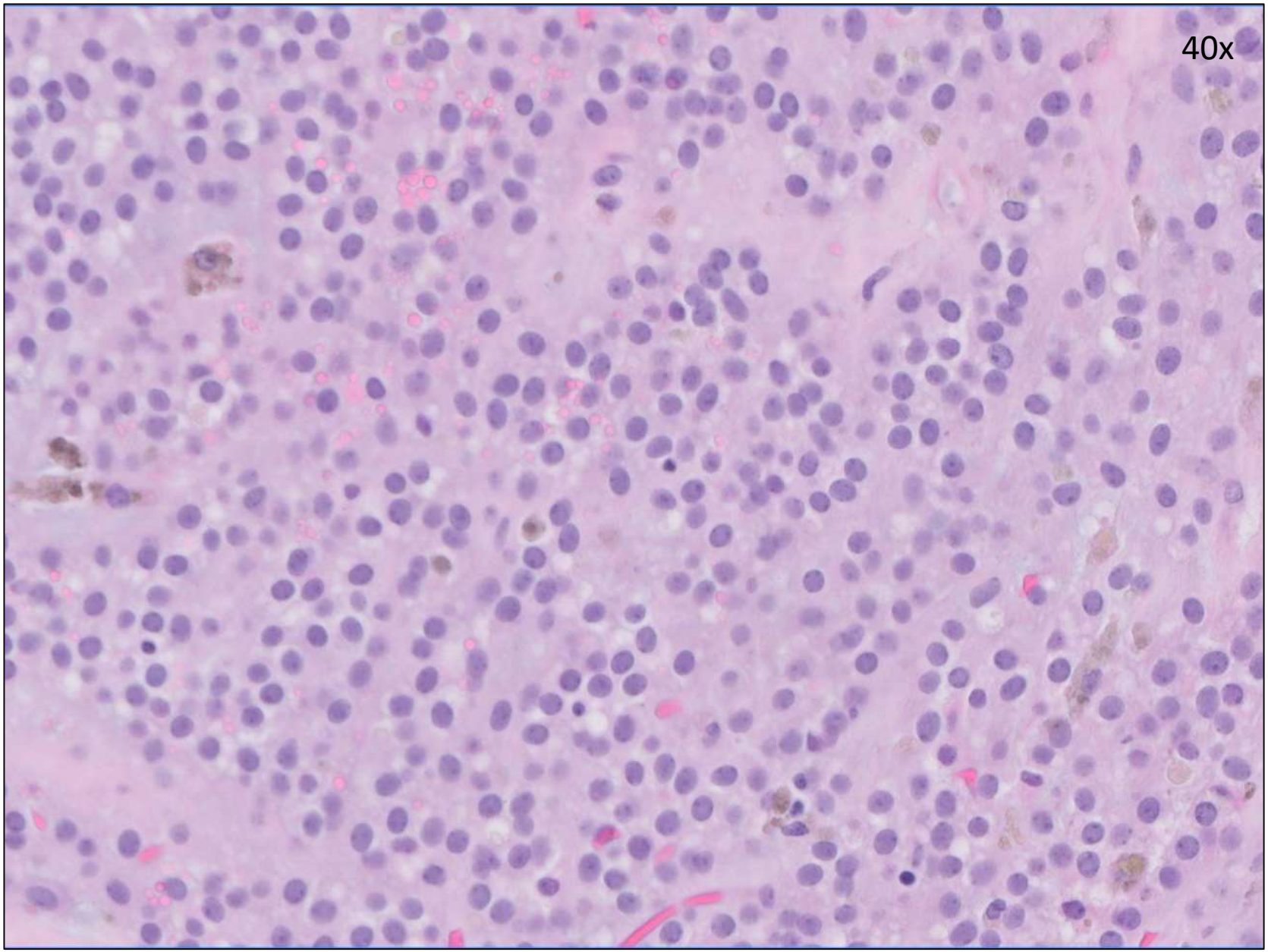
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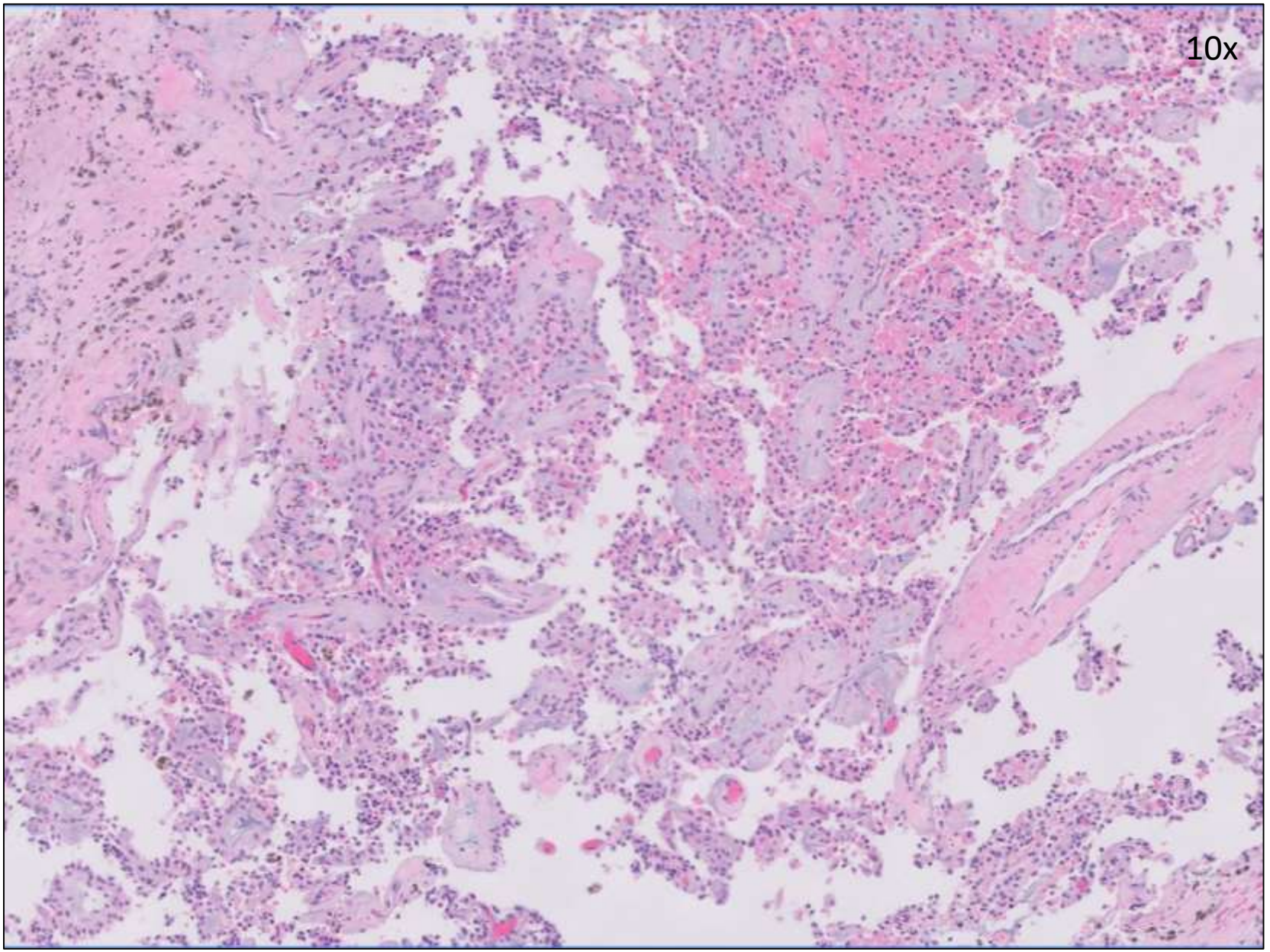
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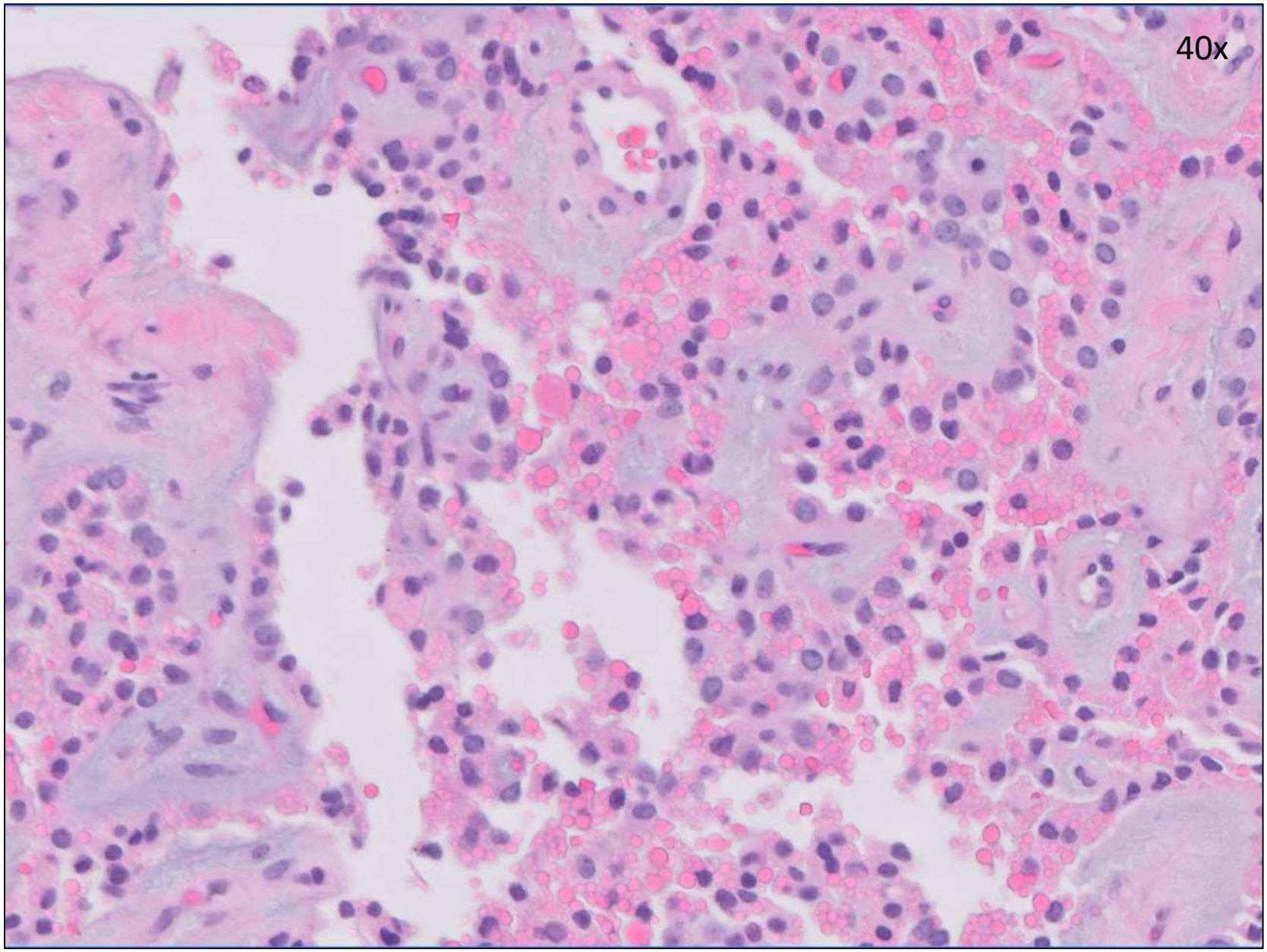
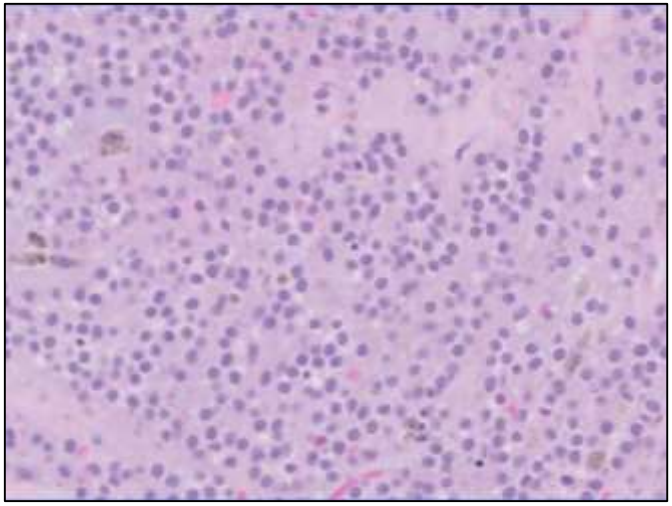
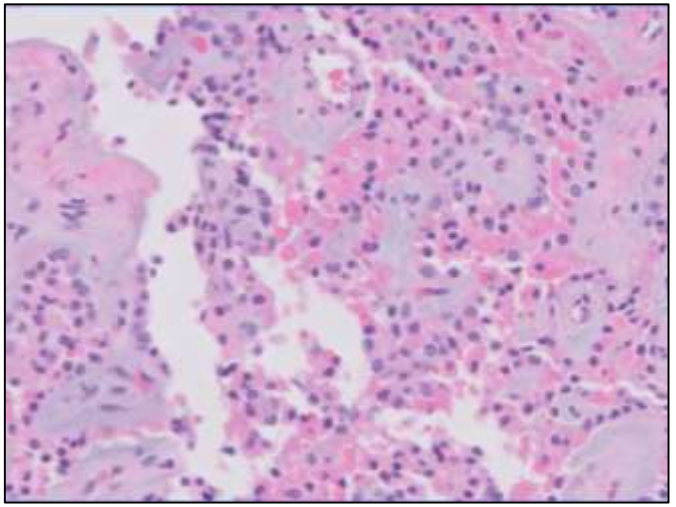

40x

Differential diagnosis

Bland monotonous nuclei, solid and papillary growth, eosinophilic hyaline globules

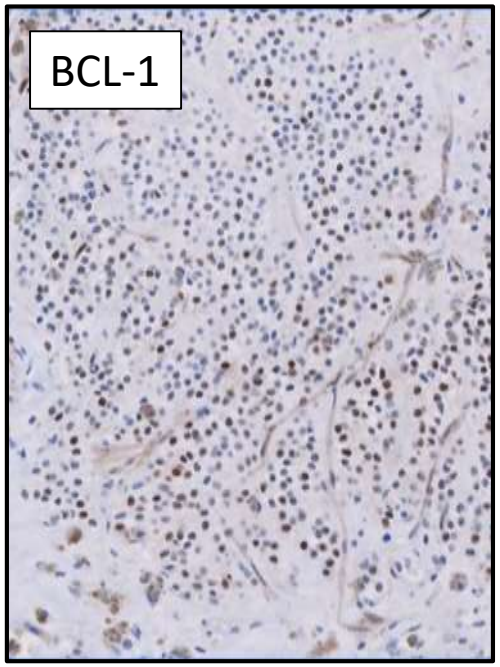
- Sex cord stromal tumor
  - Juvenile granulosa cell tumor
  - Microcystic stromal tumor
  - Signet ring stromal tumor
- Germ cell tumor
- Epithelial neoplasm
- Solid pseudopapillary tumor

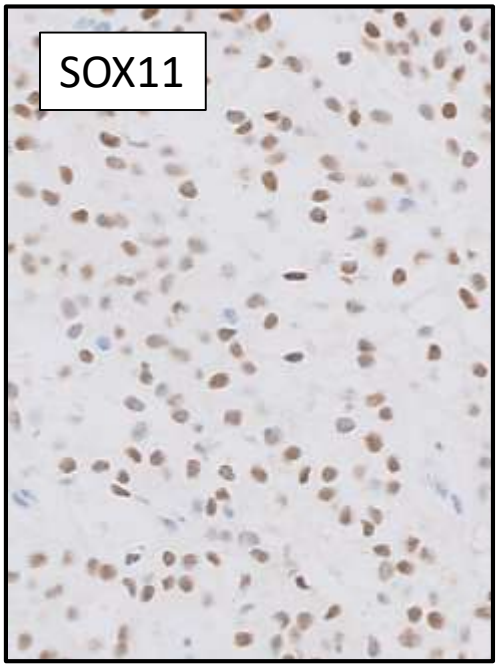


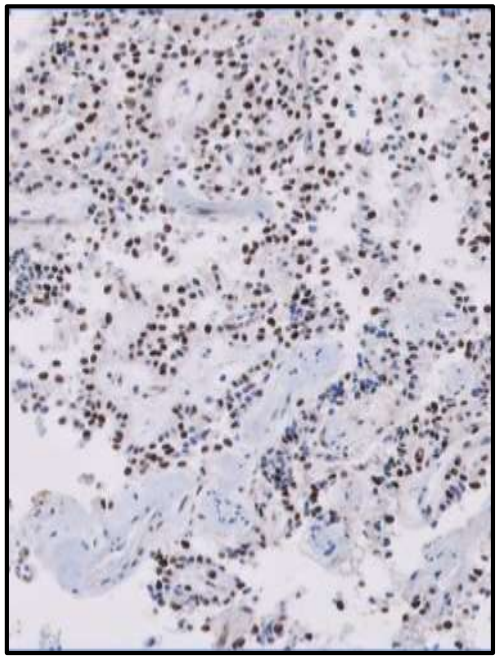


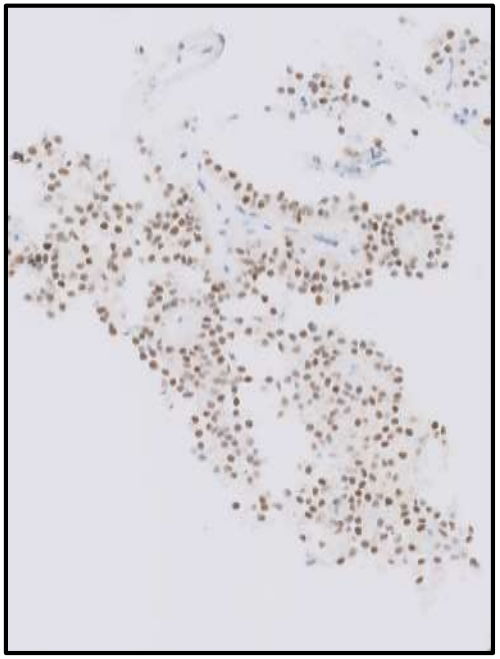
Negative IHC

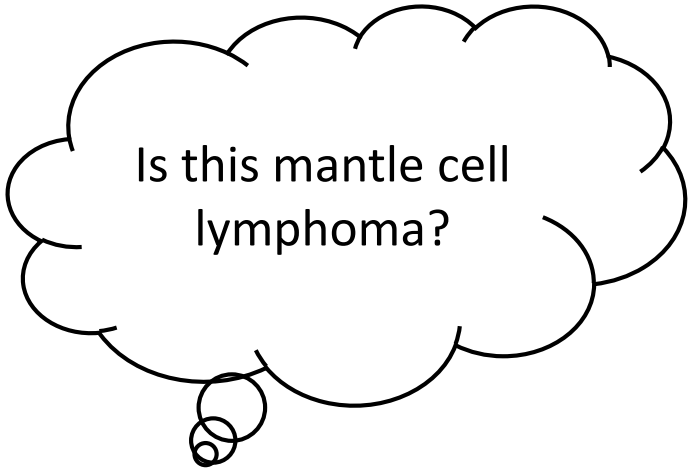
- Calretinin, inhibin, SF-1, FOXL2
- SALL4, OCT4, CD117, Glypican-3
- SOX10
- Pankeratin (focal staining)
- PAX8
- WT-1


BCL-1


SOX11



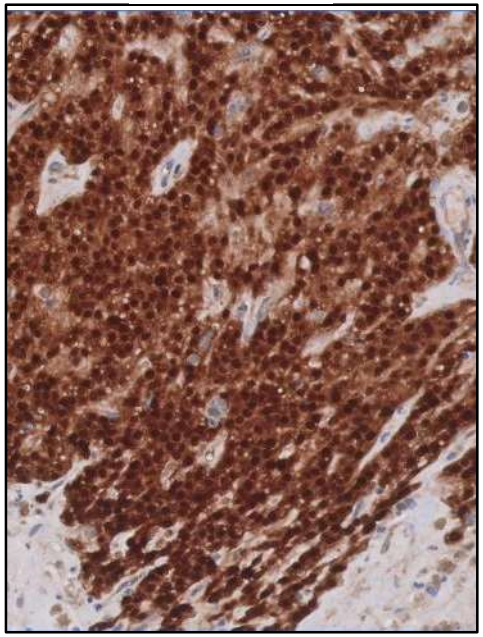


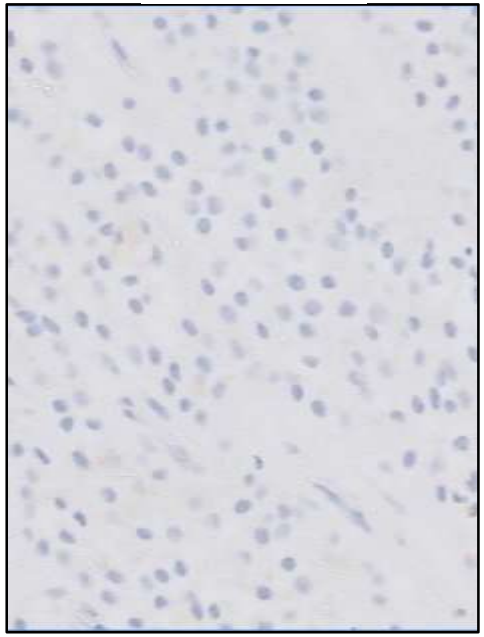

Is this mantle cell lymphoma?

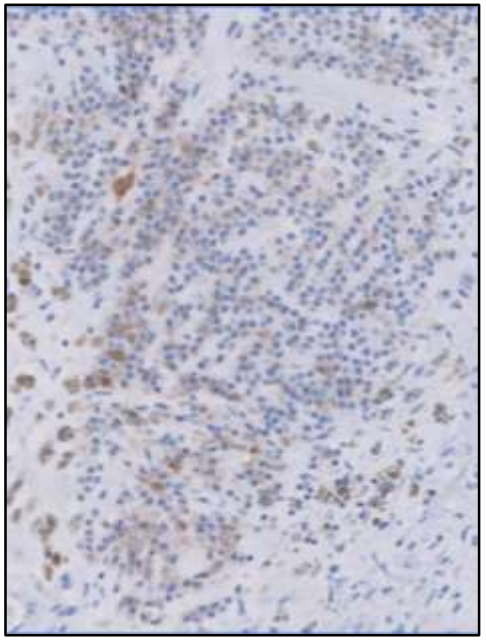
B-catenin

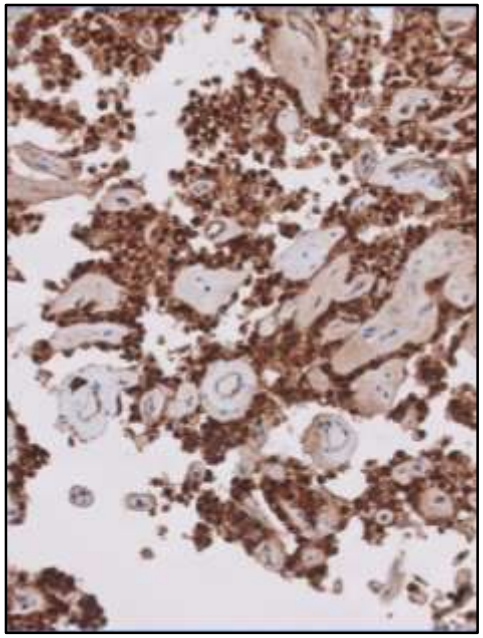
E-cadherin

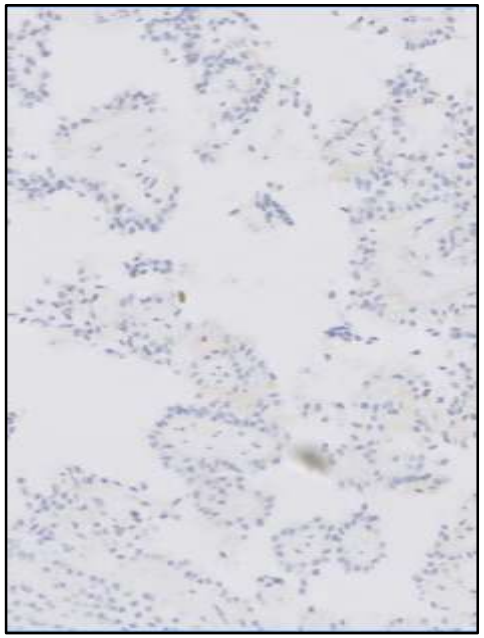
CD10

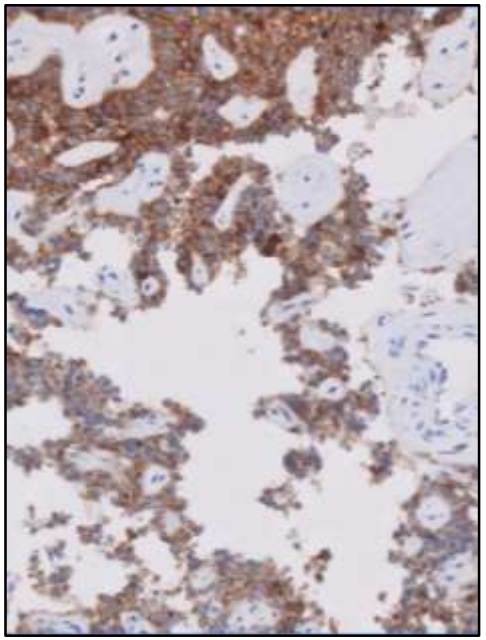












Diagnosis:

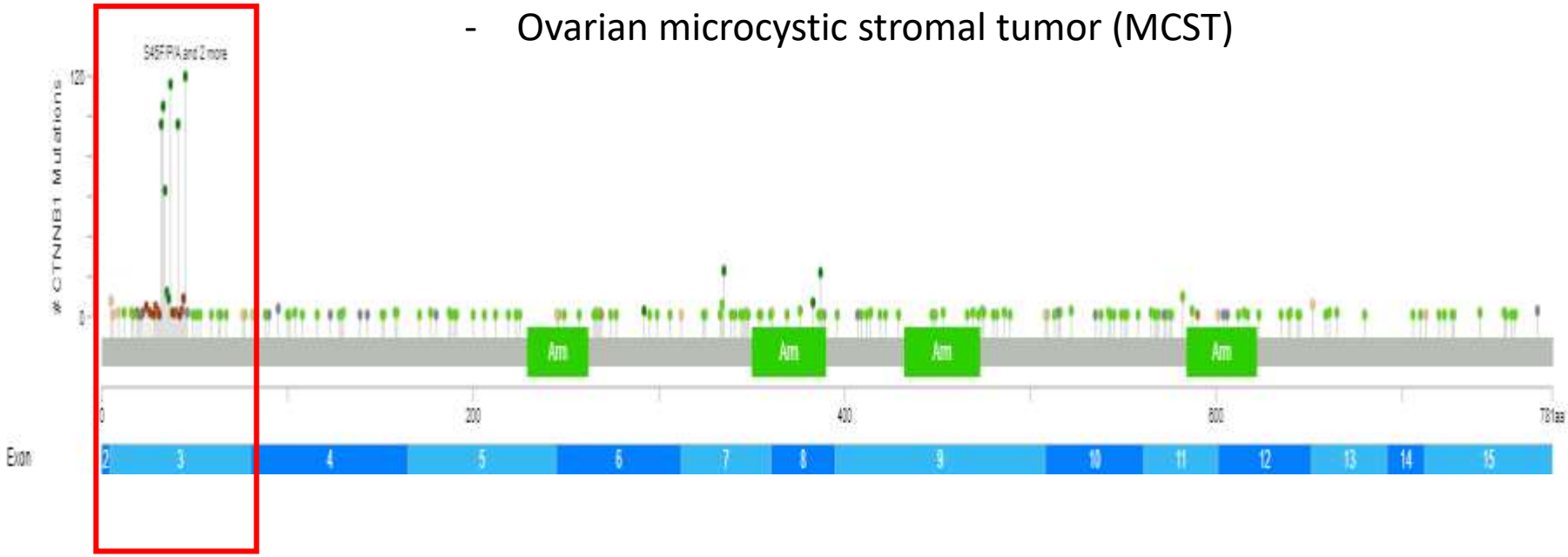
“Most c/w
solid pseudopapillary tumor”

Solid pseudopapillary tumor of the ovary

- Extremely rare
- Histomorphologically identical to pancreatic solid pseudopapillary tumors
  - Must rule out metastasis!
- Histologic features:
  - Growth patterns:
    1. Solid
    2. Nested
    3. Pseudopapillary (myxoid/hyalinized cores)
  - PAS-D+ hyaline globules (intra and extracellular)
- IHC: B-catenin (N+C), CD10, E-cadherin loss
- Controversial!

CTNNB1 exon 3 mutations

- Pancreatic solid pseudopapillary tumor (P-SPT)
- Ovarian solid pseudopapillary tumor (O-SPT)
- Ovarian microcystic stromal tumor (MCST)



ORIGINAL ARTICLE

Microcystic Stromal Tumor of the Ovary

Report of 16 Cases of a Hitherto Uncharacterized Distinctive Ovarian Neoplasm

Julie A. Irving, MD† and Robert H. Young, MD‡§*

Am J Surg Pathol • Volume 33, Number 3, March 2009

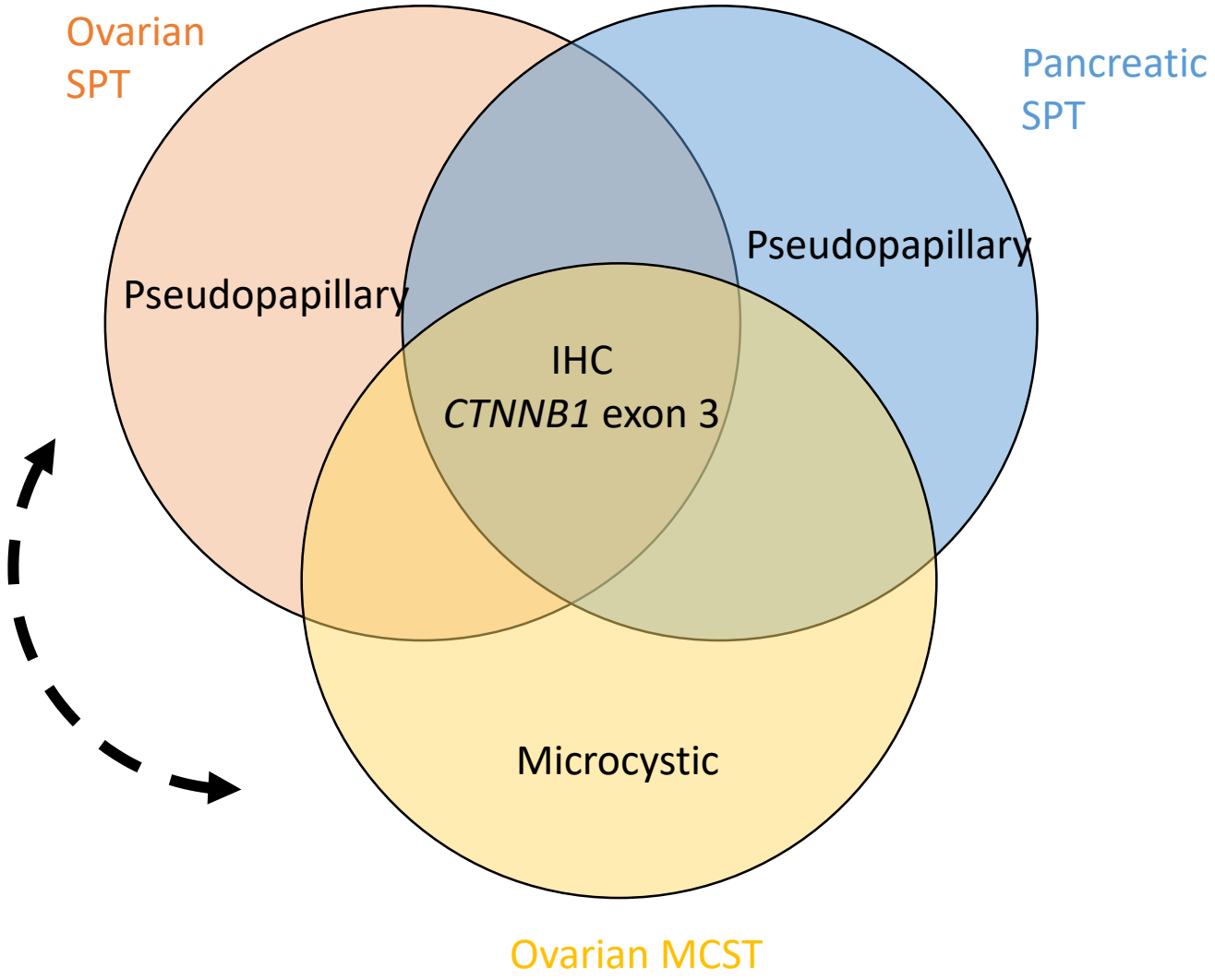
ORIGINAL ARTICLE

Solid Pseudopapillary Neoplasm of the Ovary: A Report of 3 Primary Ovarian Tumors Resembling Those of the Pancreas

Vikram Deshpande, MD, Esther Oliva, MD, and Robert H. Young, MD

Am J Surg Pathol • Volume 34, Number 10, October 2010

Immunohistochemical and molecular overlap



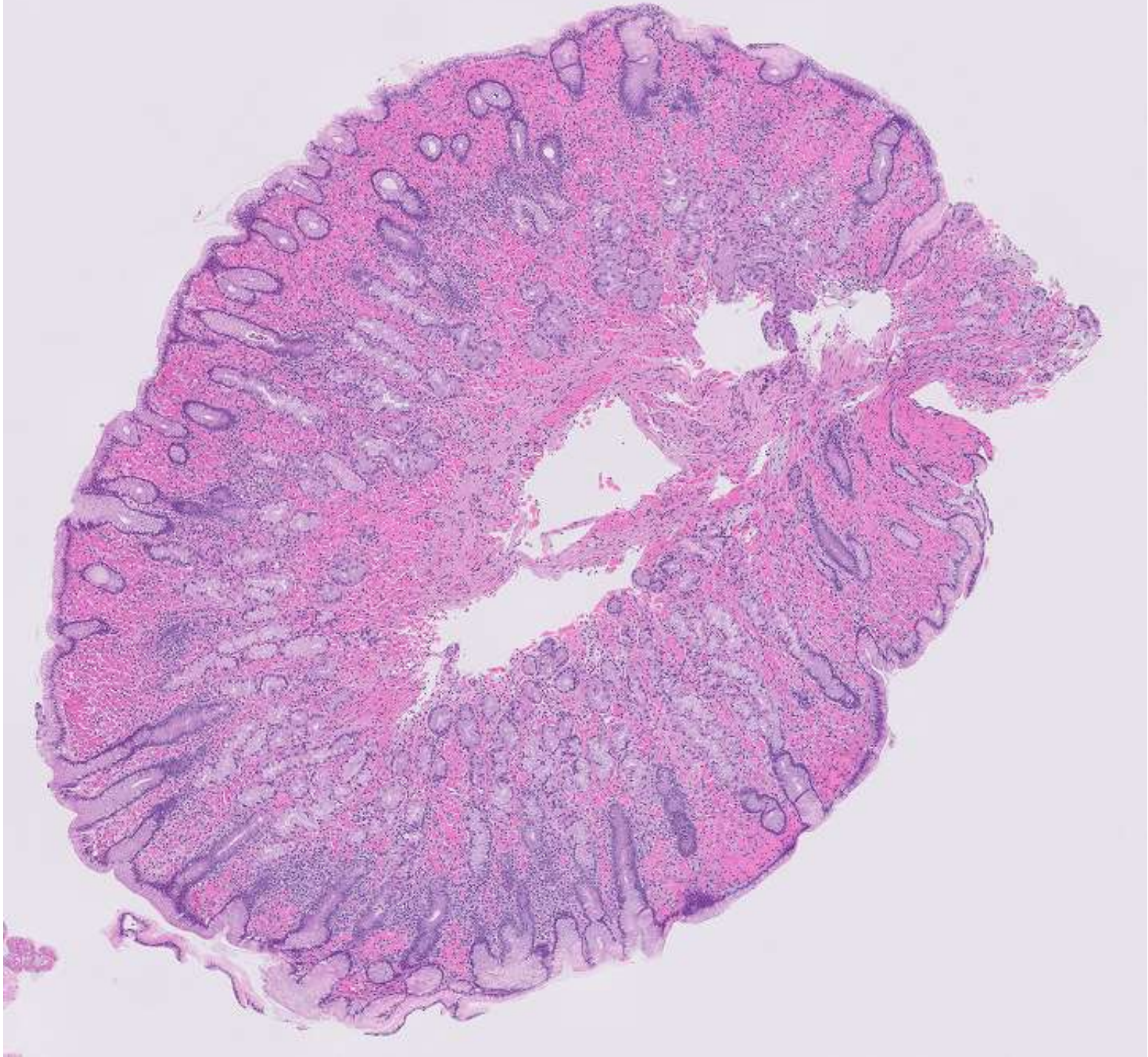
Take home points - Ovarian solid pseudopapillary tumor

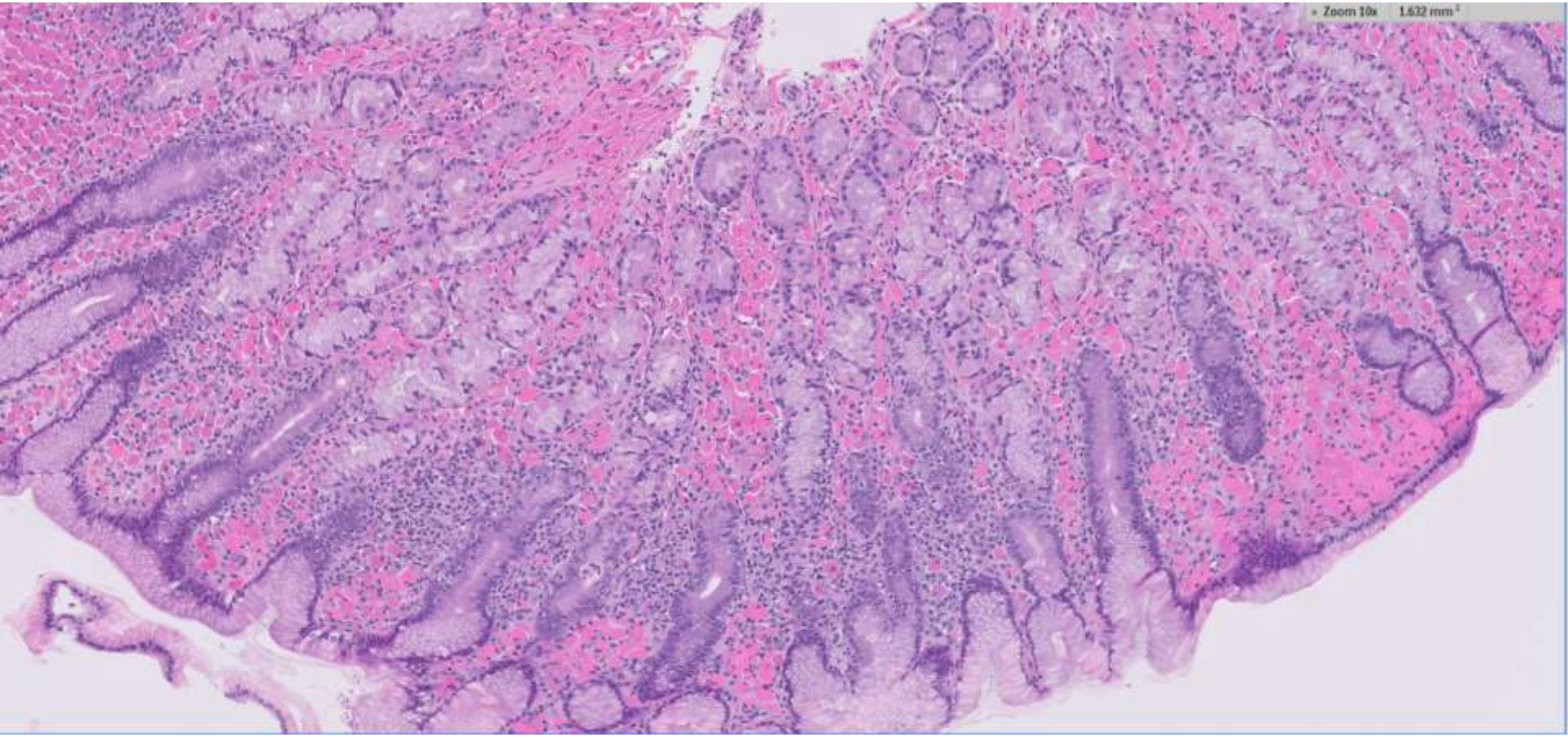
- Extremely rare tumors
- Features:
  - Growth pattern:
    1. Solid
    2. Nested
    3. **Pseudopapillary*****
  - PAS-D+ hyaline globules
  - *CTNNB1* exon 3 mutations
  - IHC: B-catenin (N+C), CD10, E-cadherin loss
- Significant histologic, IHC, and molecular overlap with ovarian microcystic stromal tumors and pancreatic SPTs!

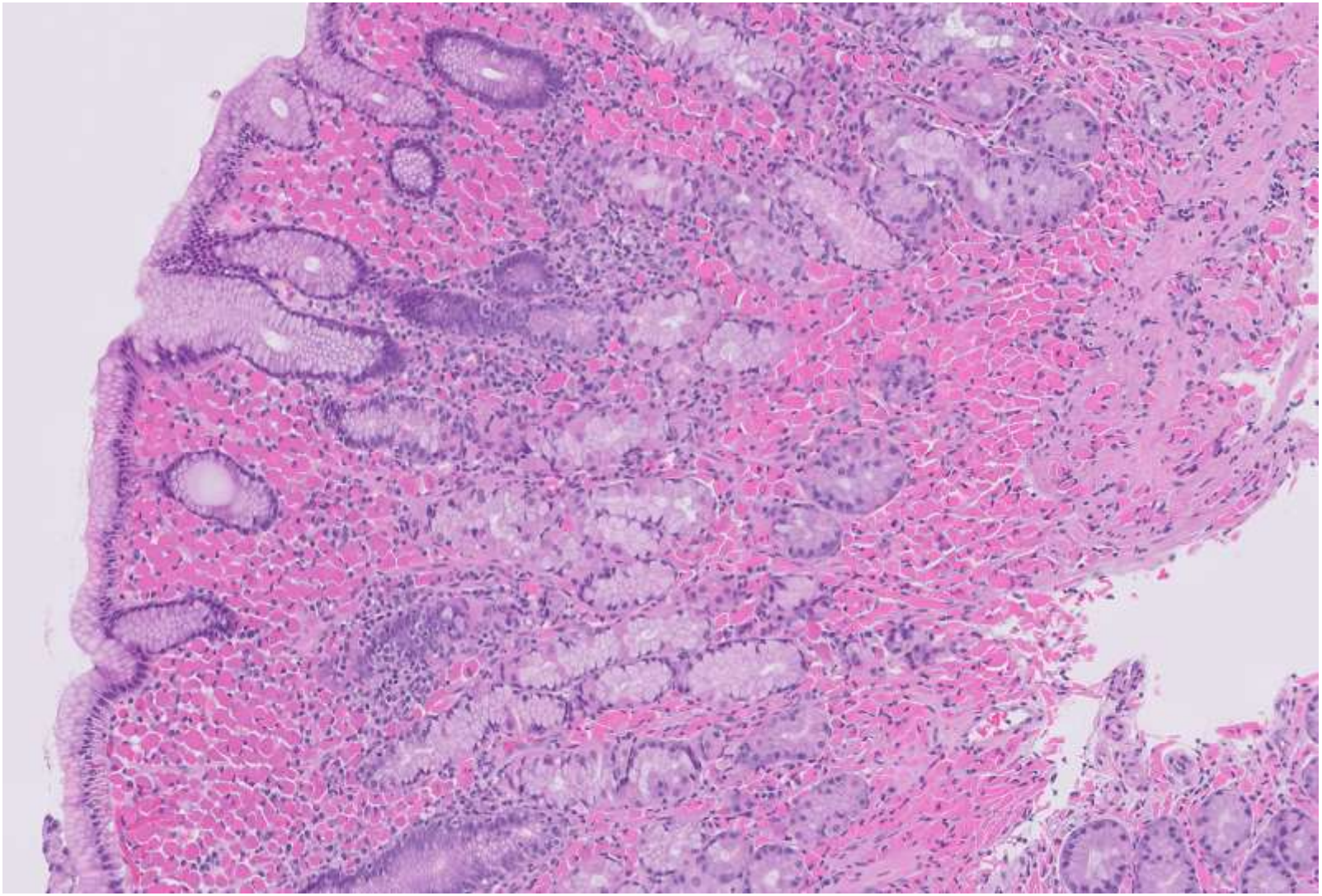
23-0203

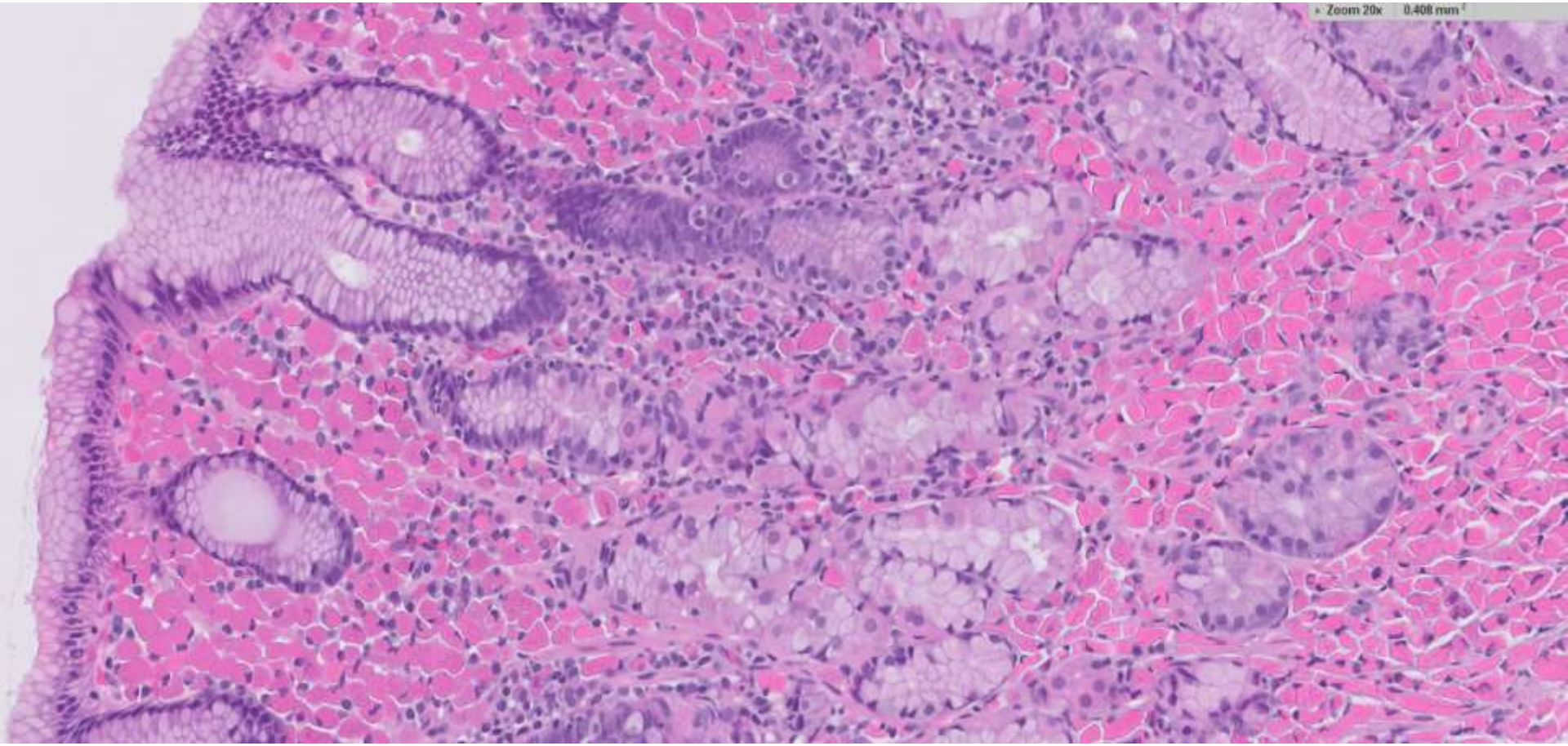
Eric Ollila/Megan Troxell; Stanford

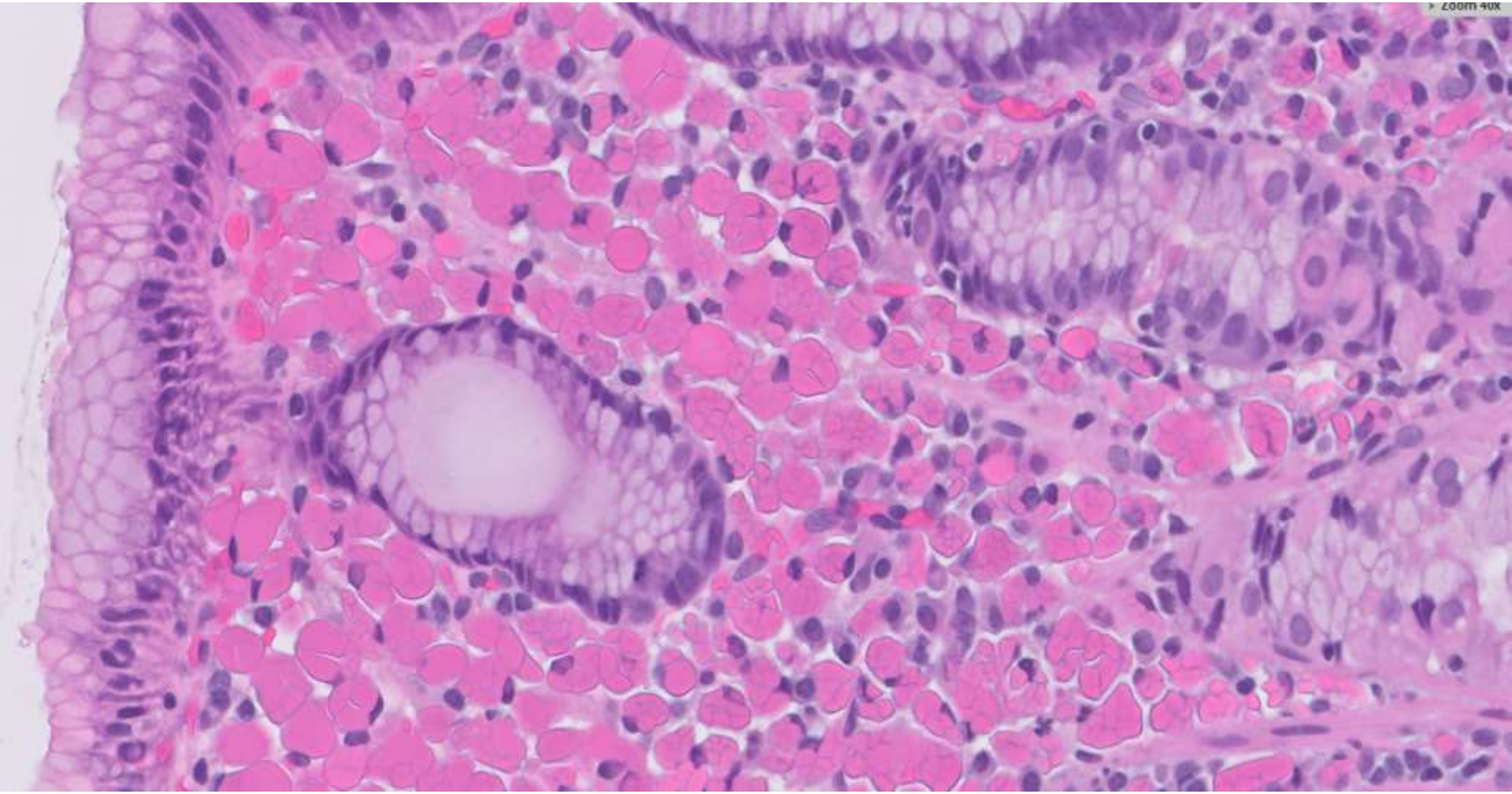
70ish M with h/o intrahepatic cholangiocarcinoma. EGD showed erosive gastritis in the antrum, incisura, and body.

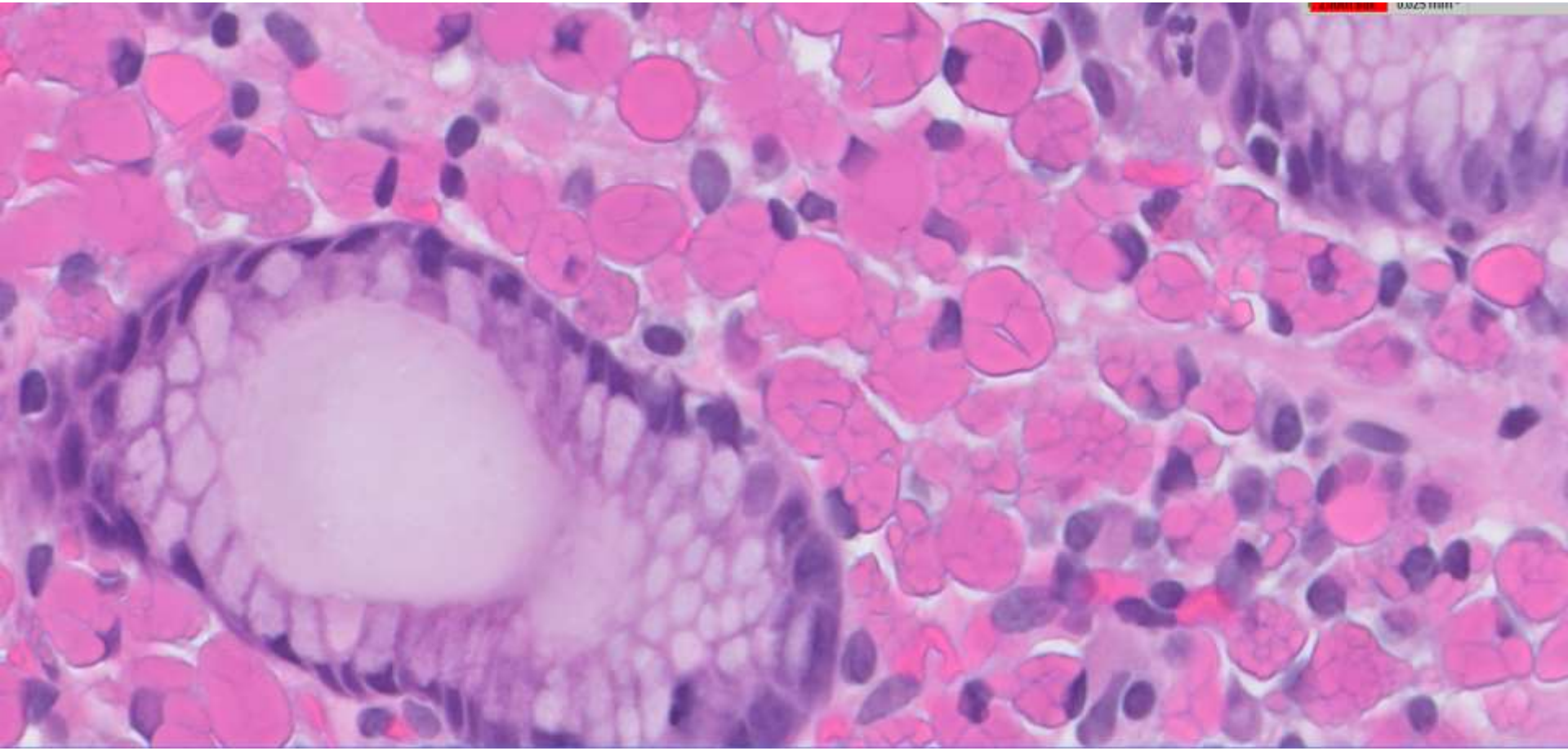



Zoom 10x 1.632 mm²




Zoom 20x 0.408 mm²


Zoom 40x



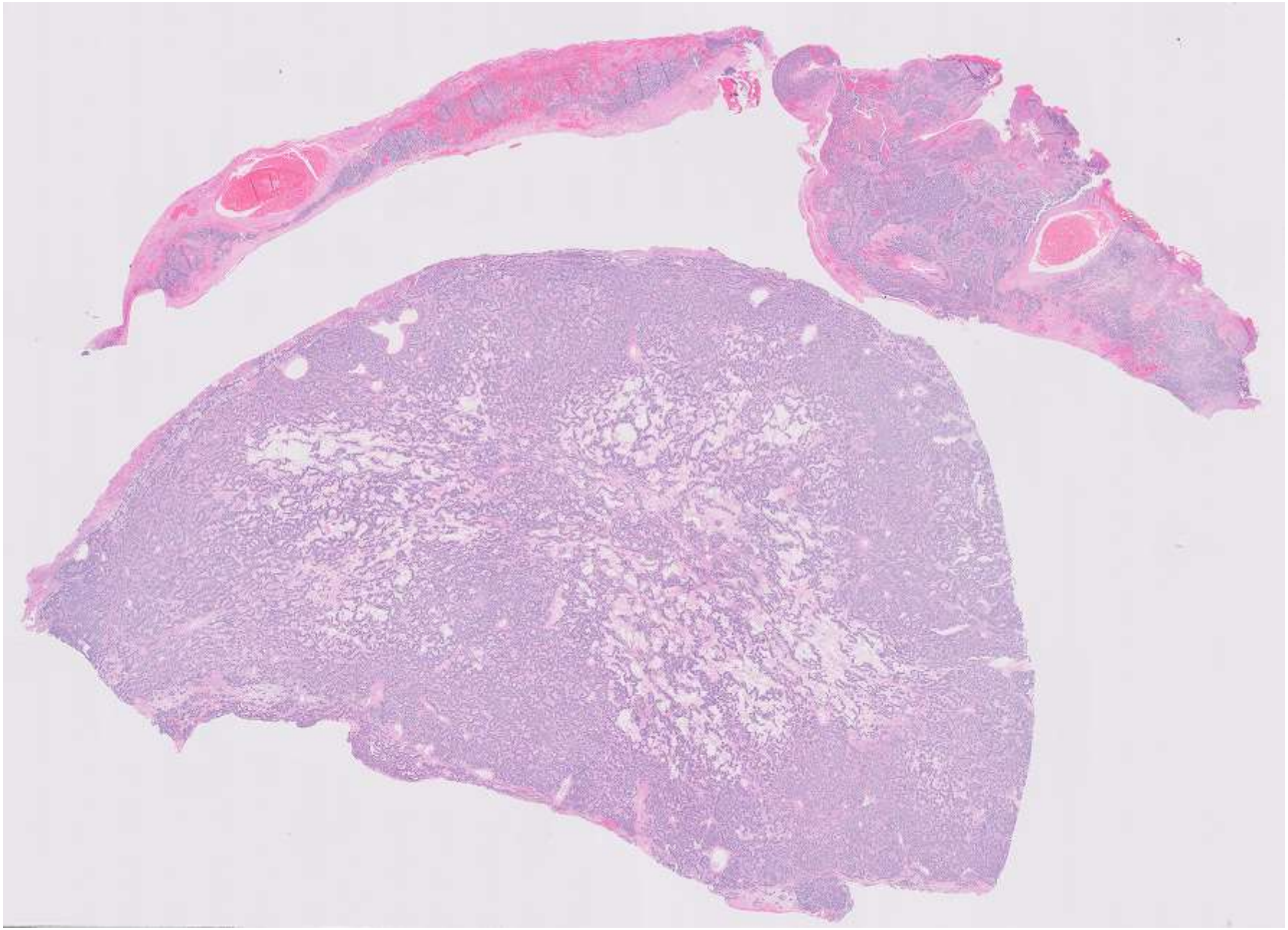
Russell body gastritis

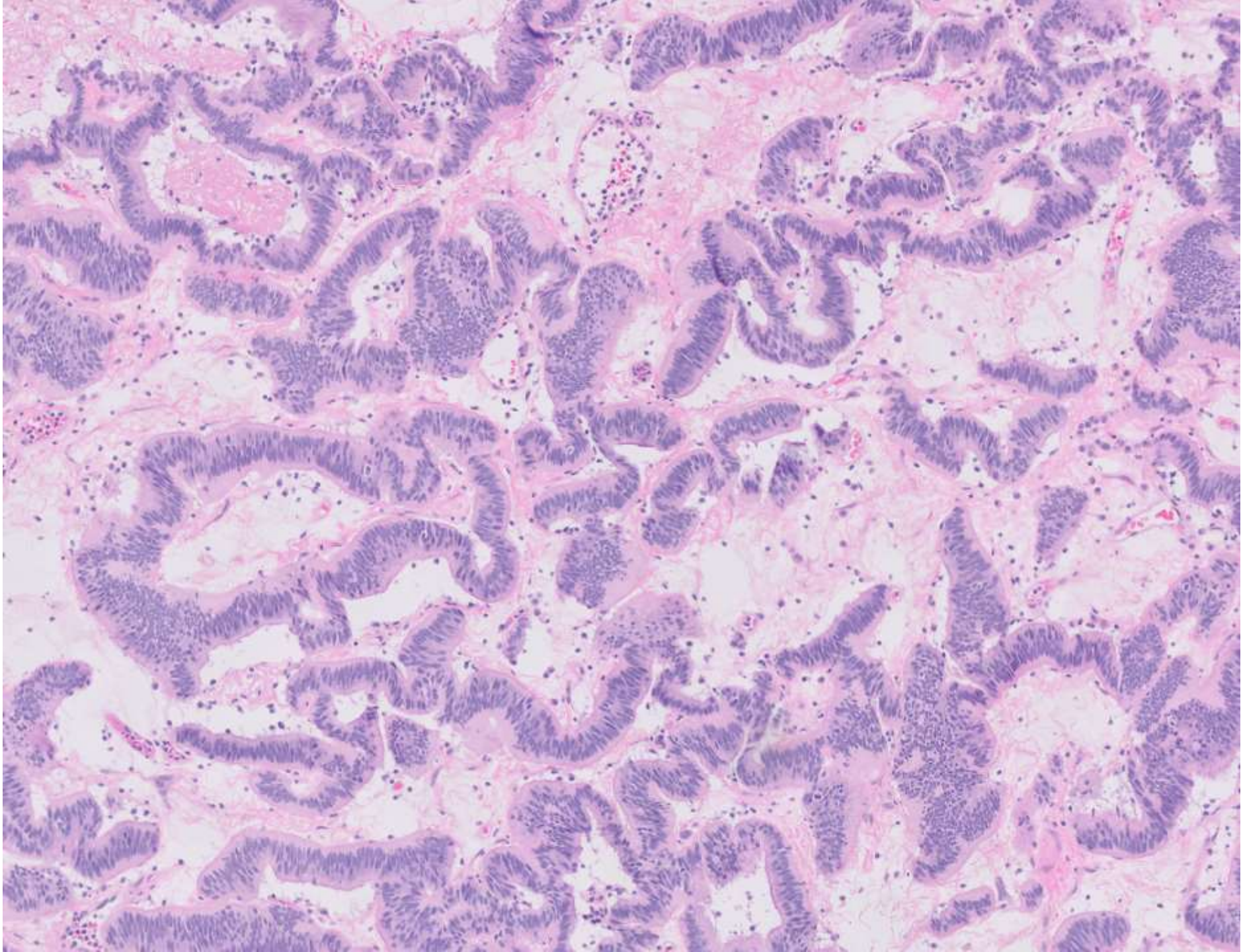
- Reactive gastric mucosal infiltration of plasma cells filled with cytoplasmic Russell bodies (Mott cells)
- Gross - mucosal congestion, erythema, scars, wall thickening, or mass
- Russell bodies - aggregates of immunoglobulins resulting from overstimulation and secretory disturbance in plasma cells
- Often associated with *H. pylori* infection and malignancies

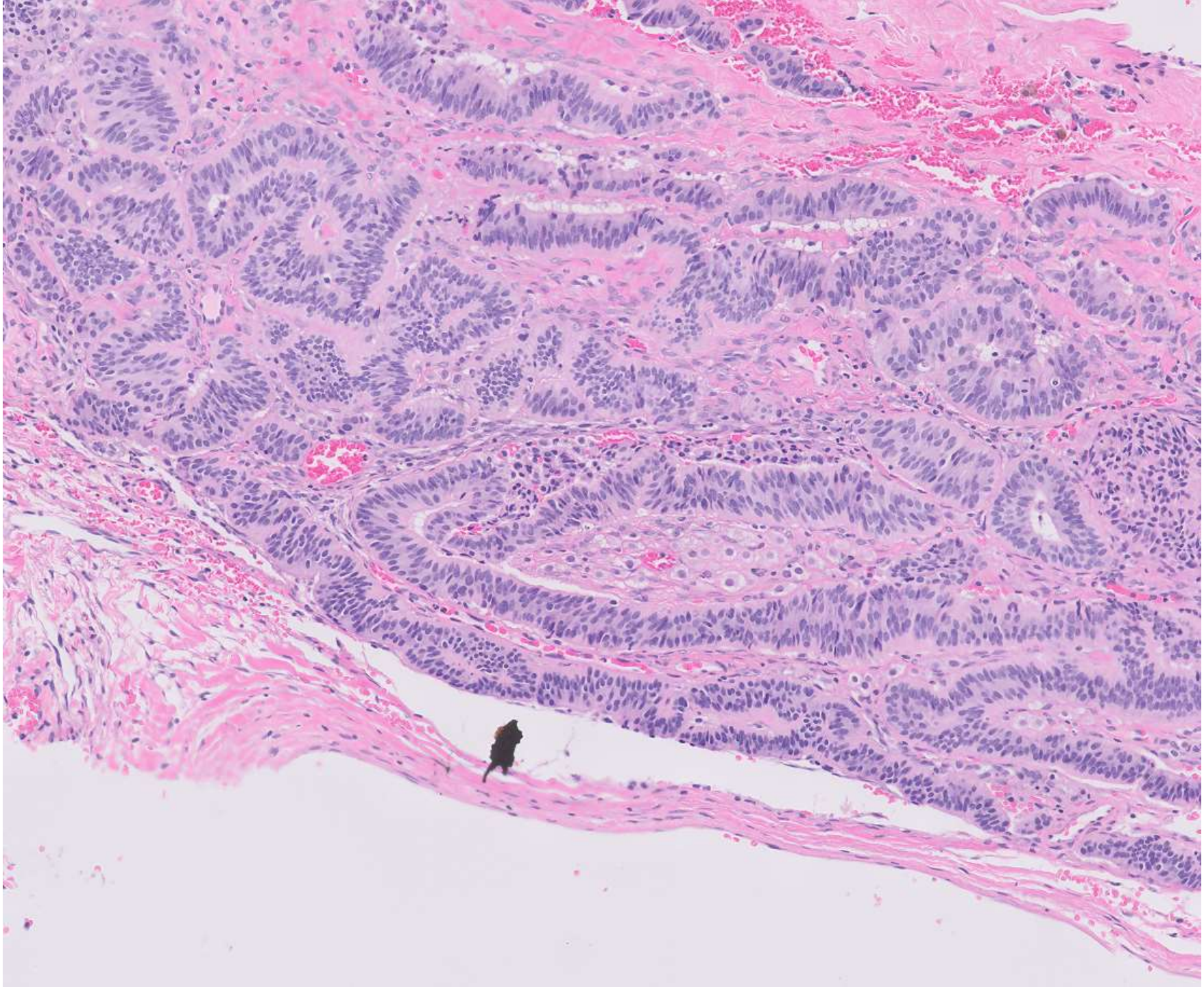
23-0204

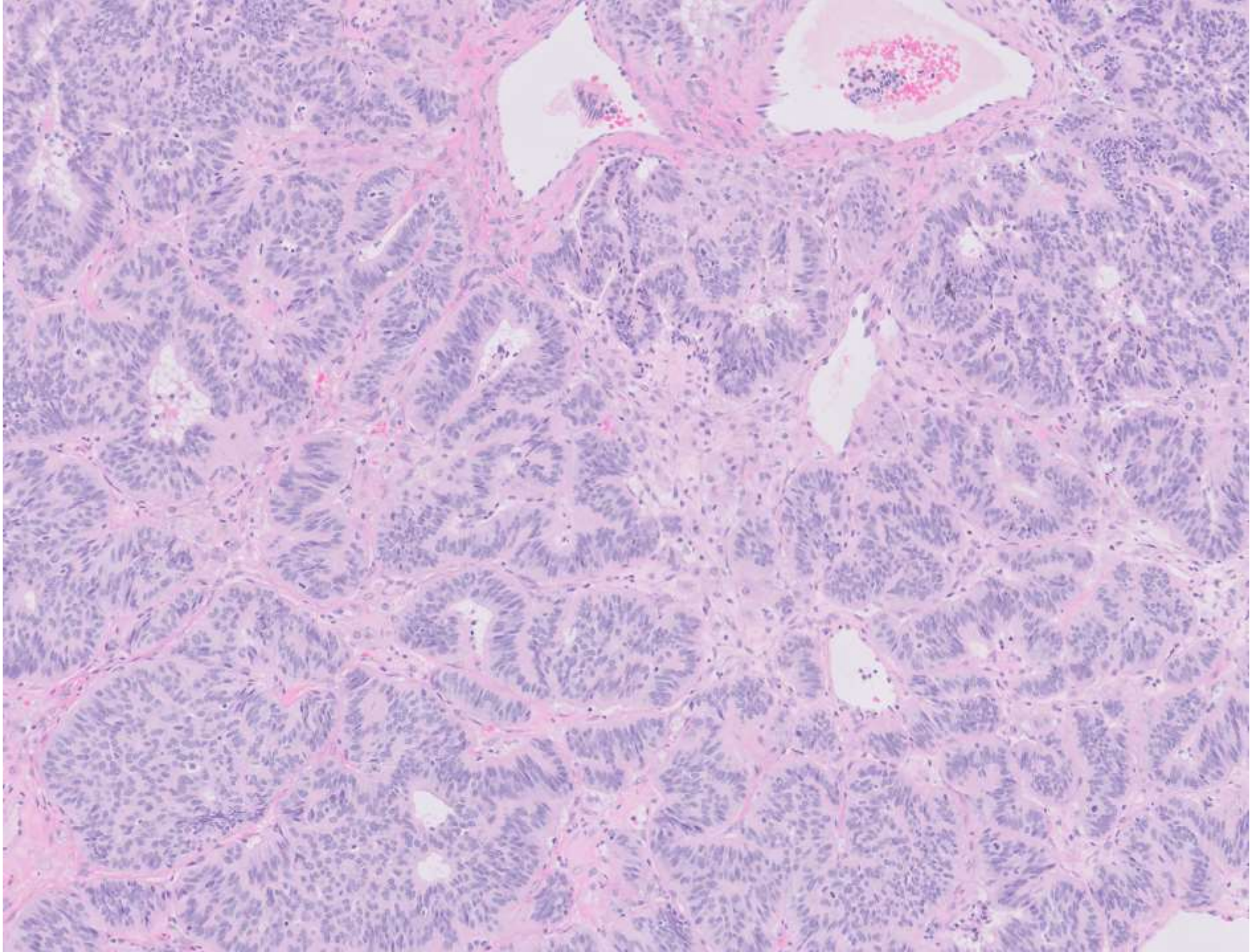
Ruth Zhang/Cynthia Gasper; UCSF

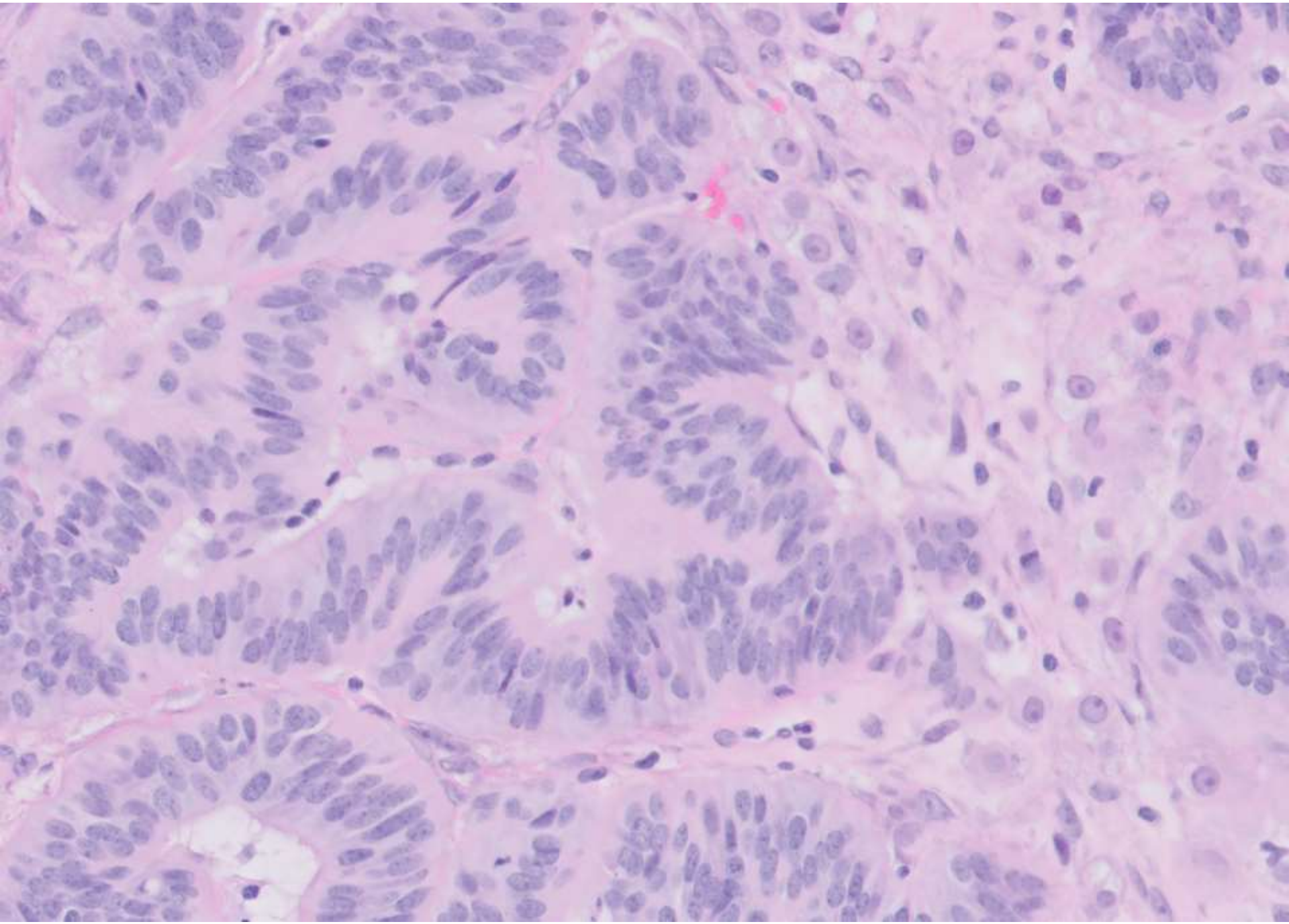
30ish F with left ovarian cyst, clinically thought to be dermoid cyst.





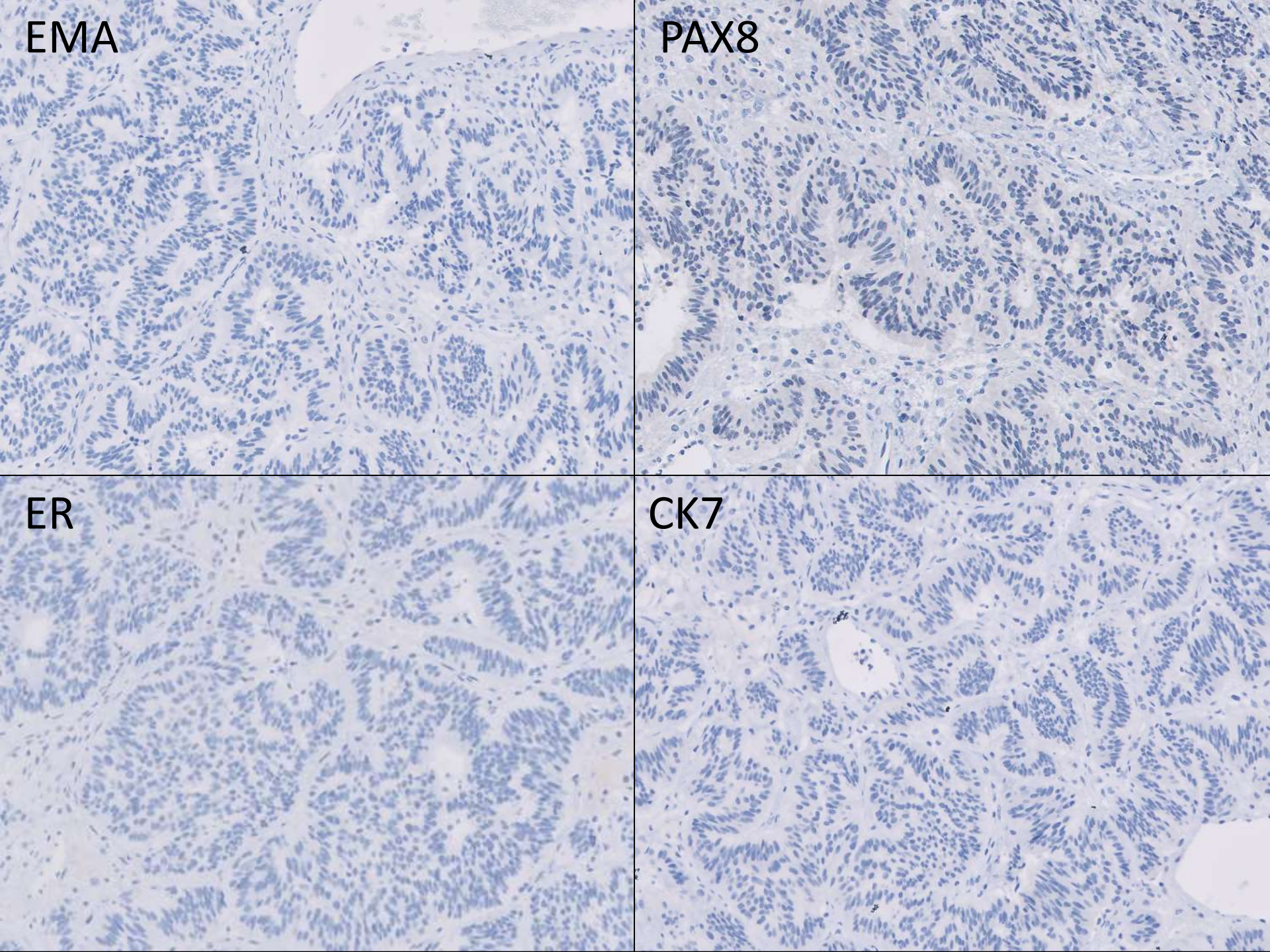


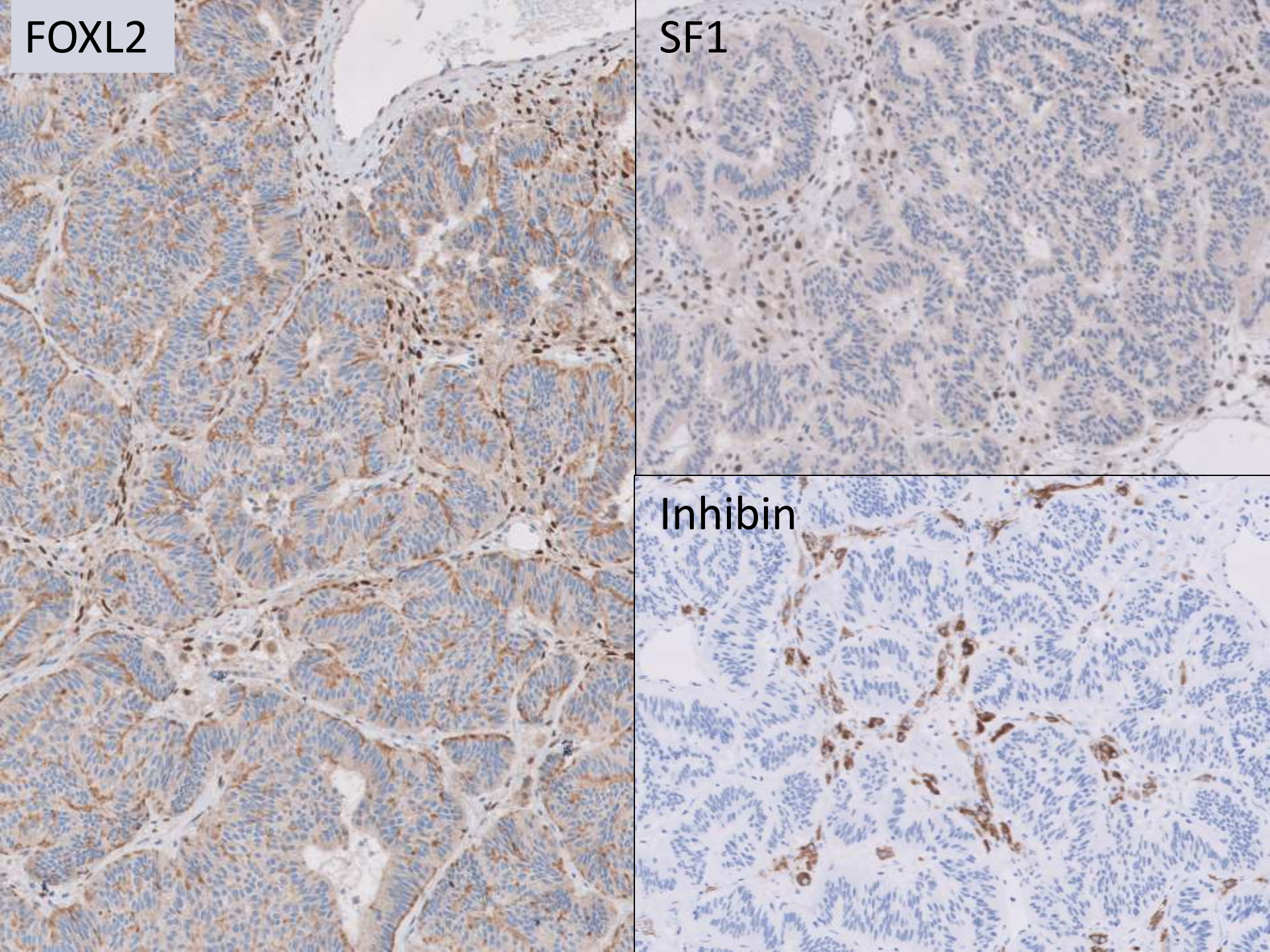


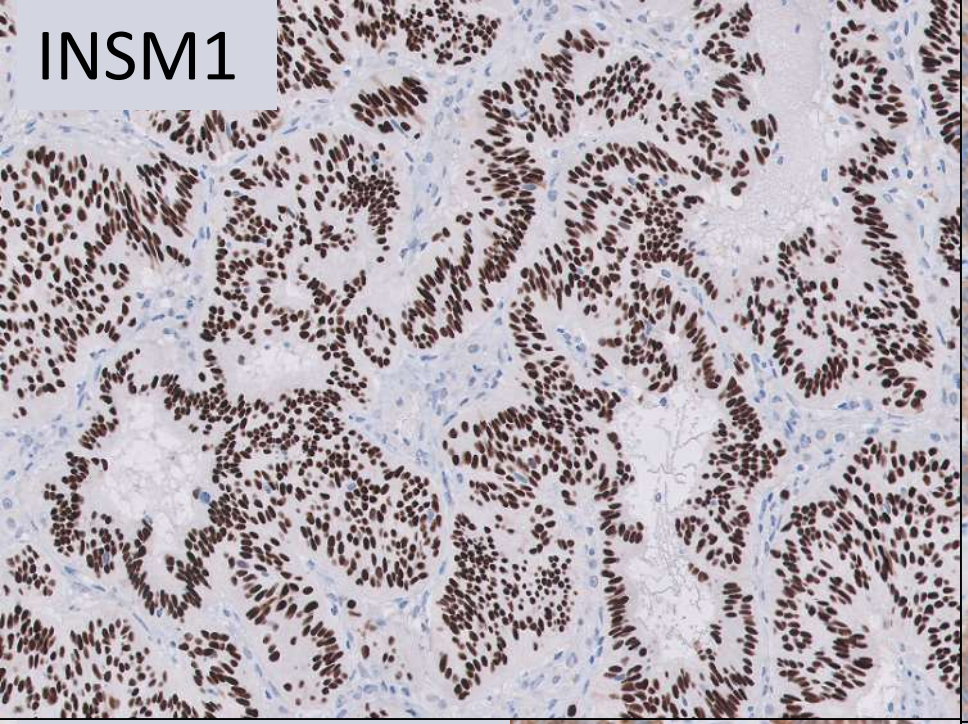


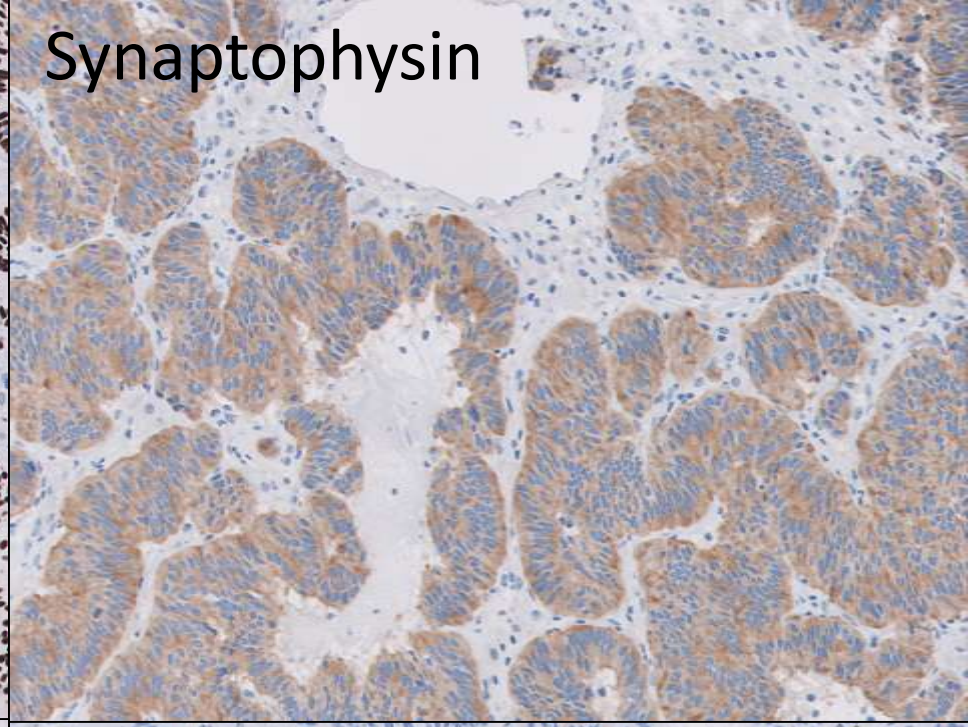
FS diagnosis

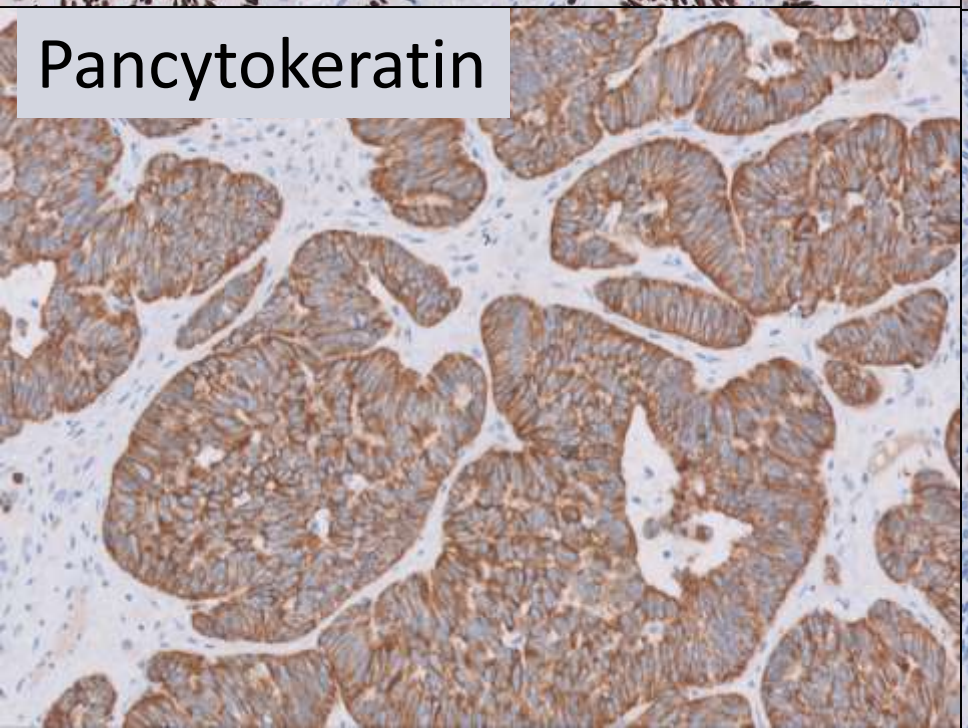
- Carcinoma, defer to permanent sections.

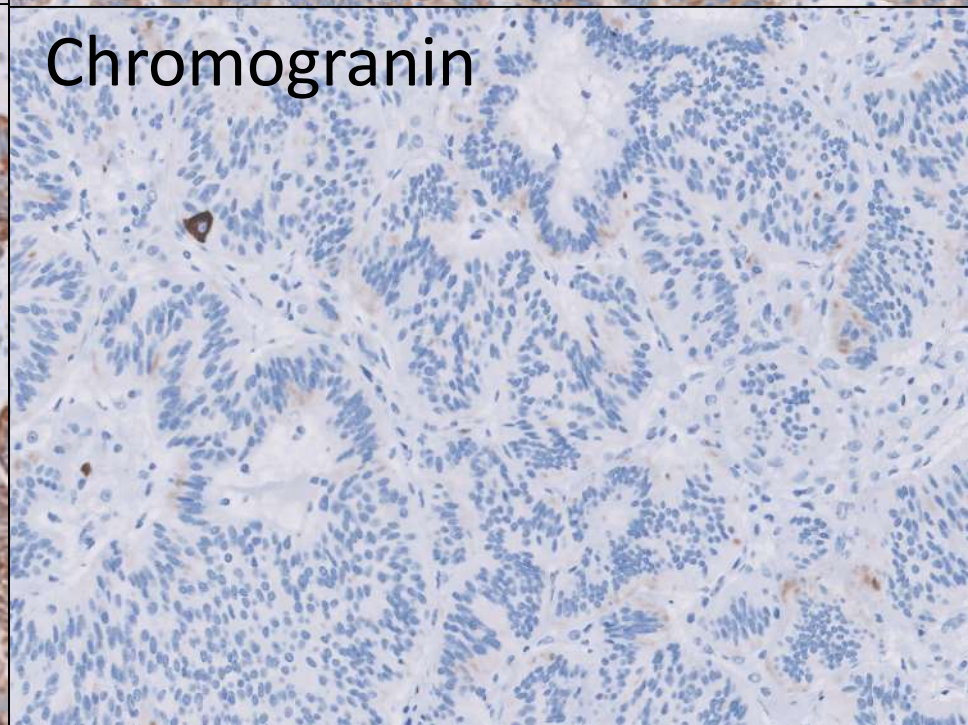

EMA
PAX8
ER
CK7


FOXL2
SF1
Inhibin


INSM1

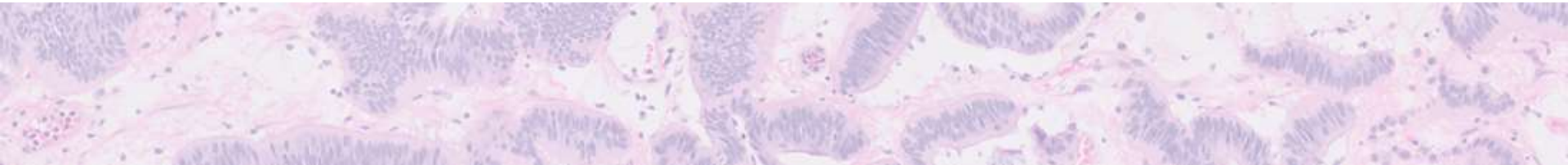

Synaptophysin


Pancytokeratin


Chromogranin

Trabecular carcinoid of ovary

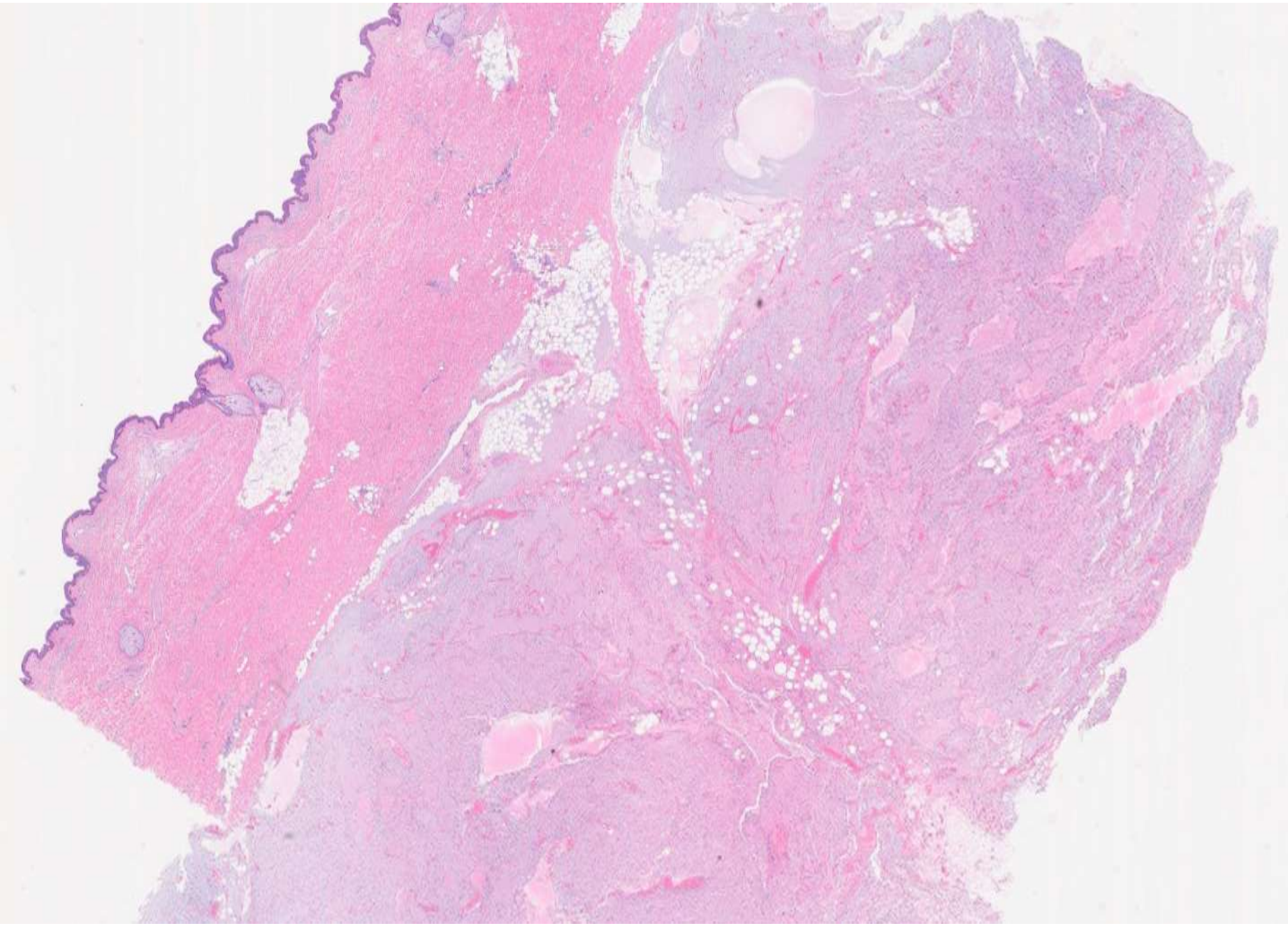
- Variant of well-differentiated neuroendocrine tumor of the ovary
  - Other variants include insular, stromal, and mucinous
- Trabecular/corded architecture
- Salt-and-pepper chromatin; cytoplasmic granules may be present
- Pitfalls: metastatic carcinoma, ovarian epithelial tumors, sex cord-stromal tumors

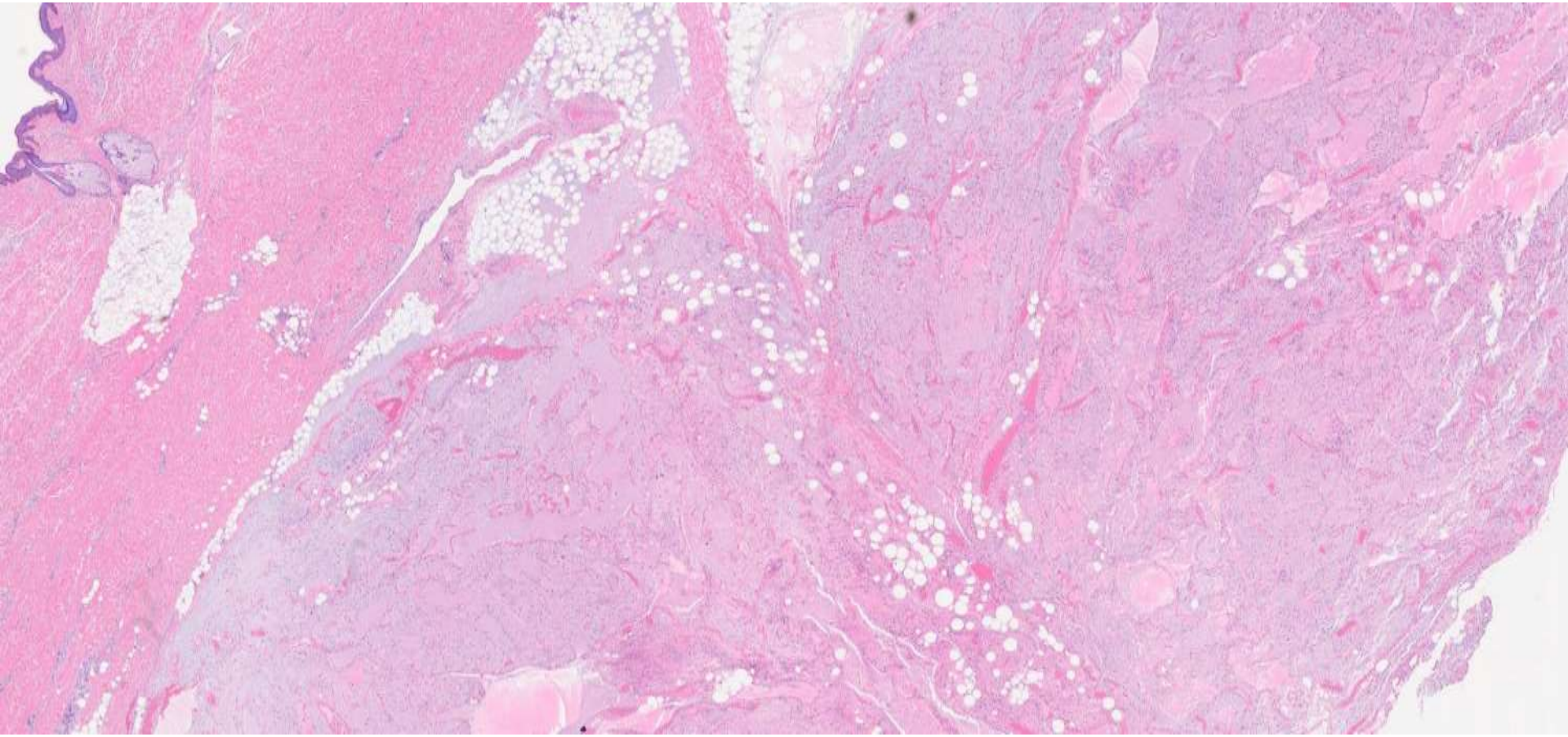


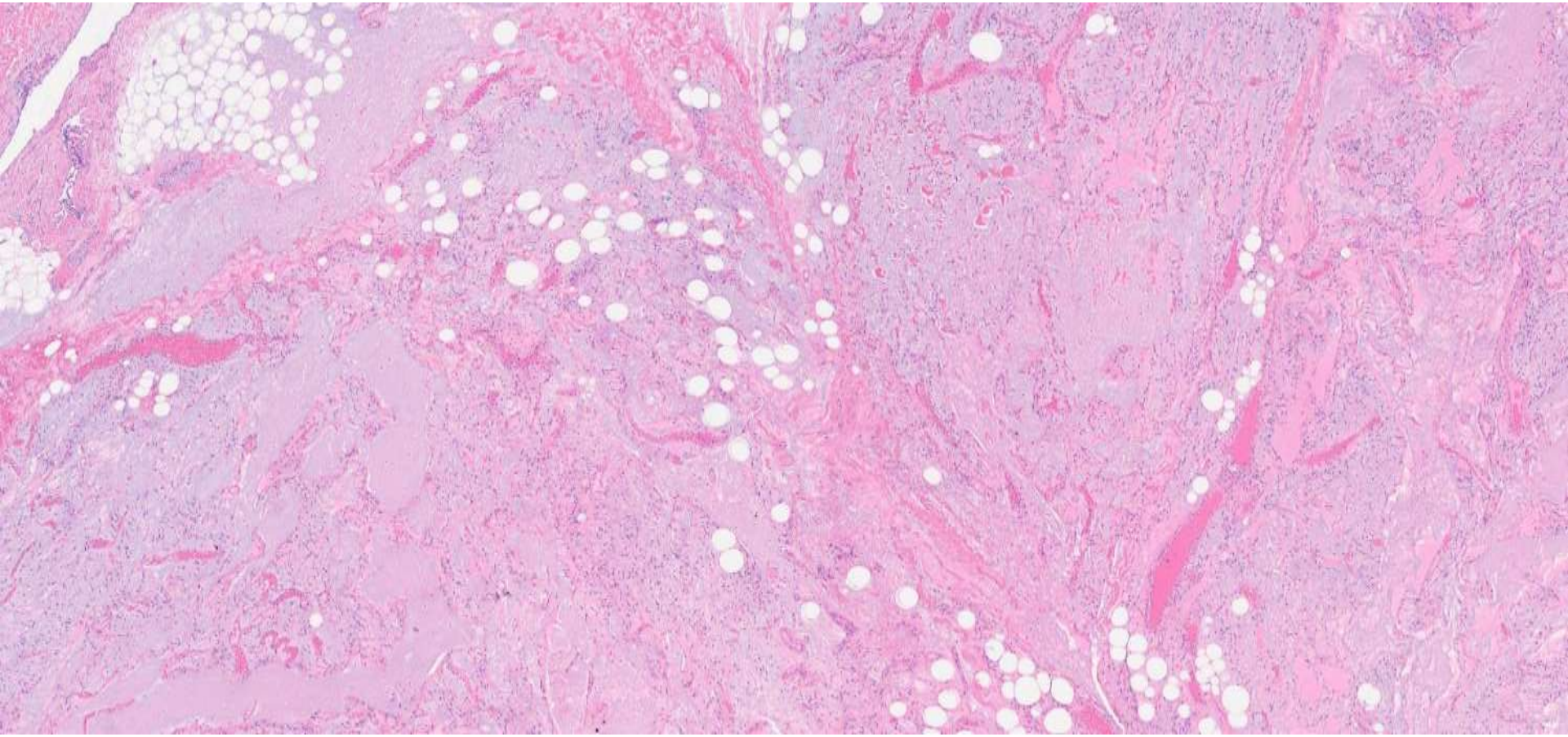
23-1205

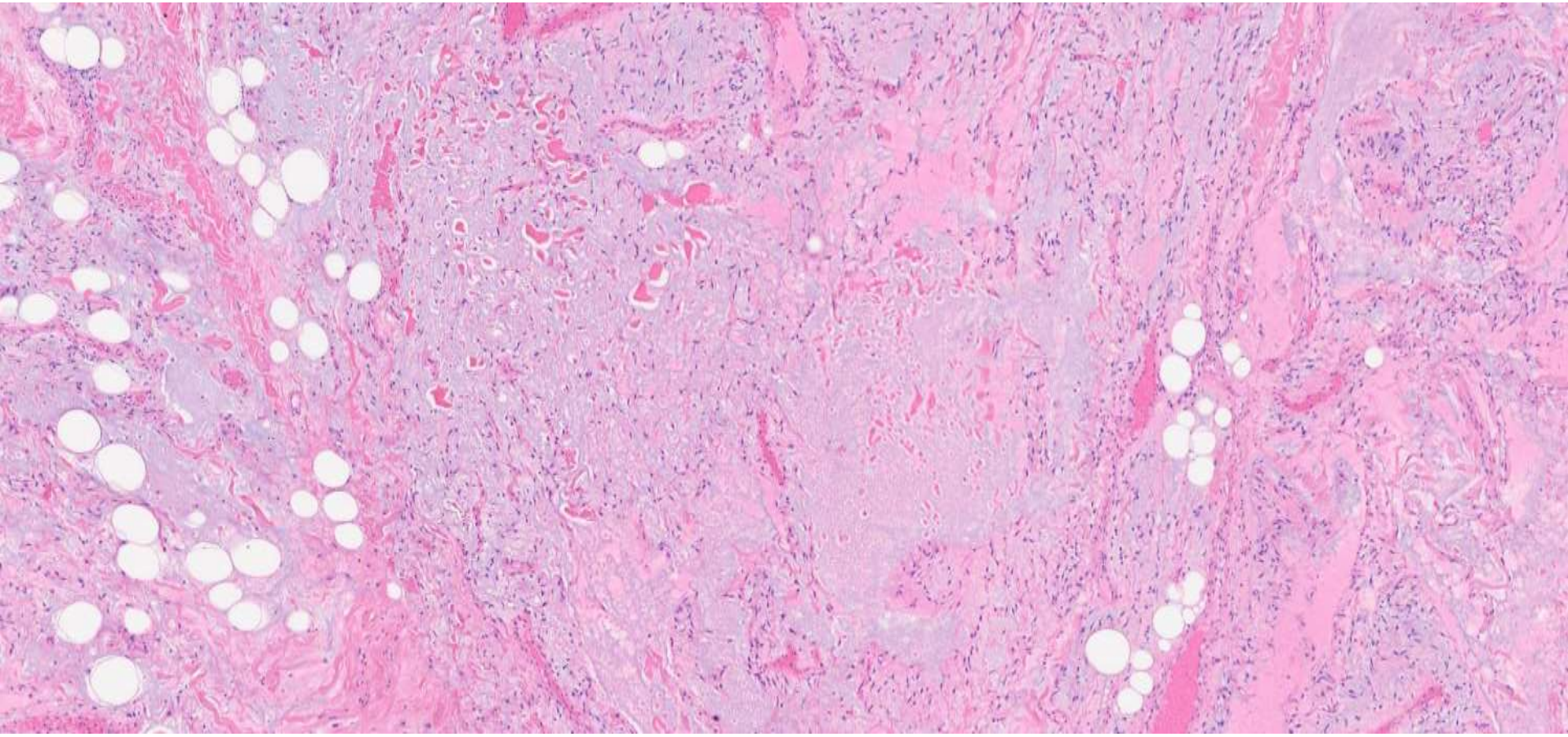
Harris Goodman; Alameda Health

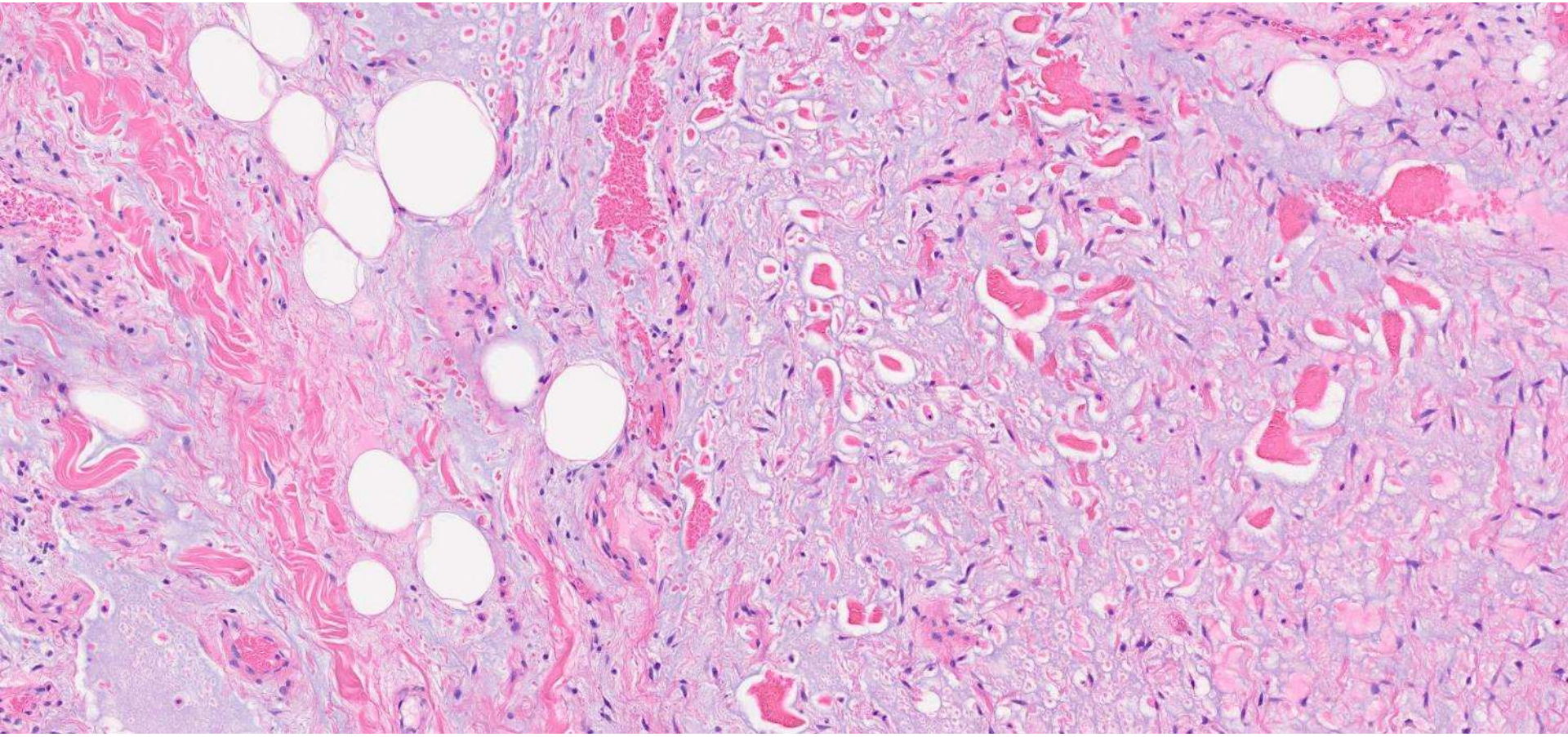
40ish M with right should mass noticed 6 months ago. He thought it arose from a spider bite. MRI showed a multilobulated mass.

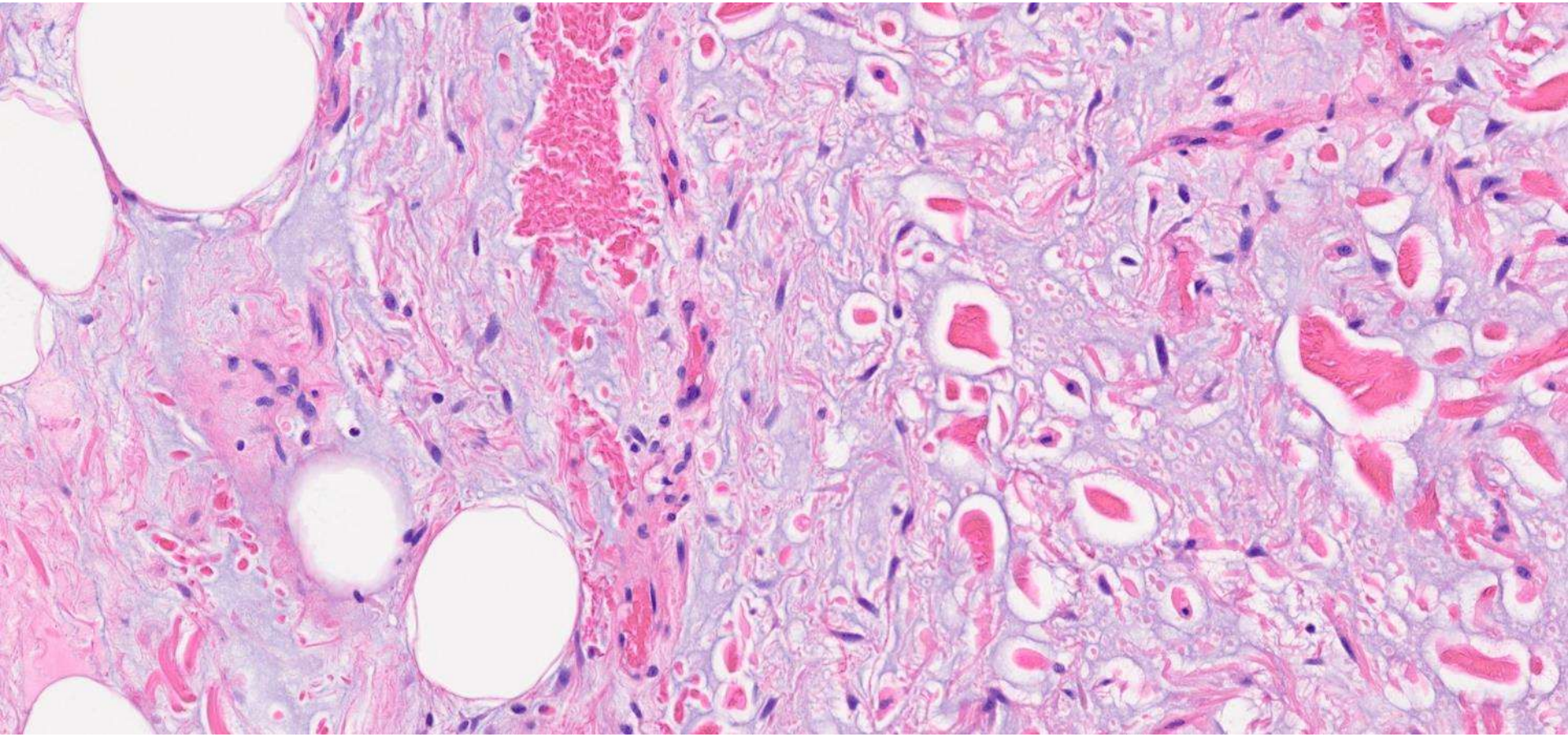


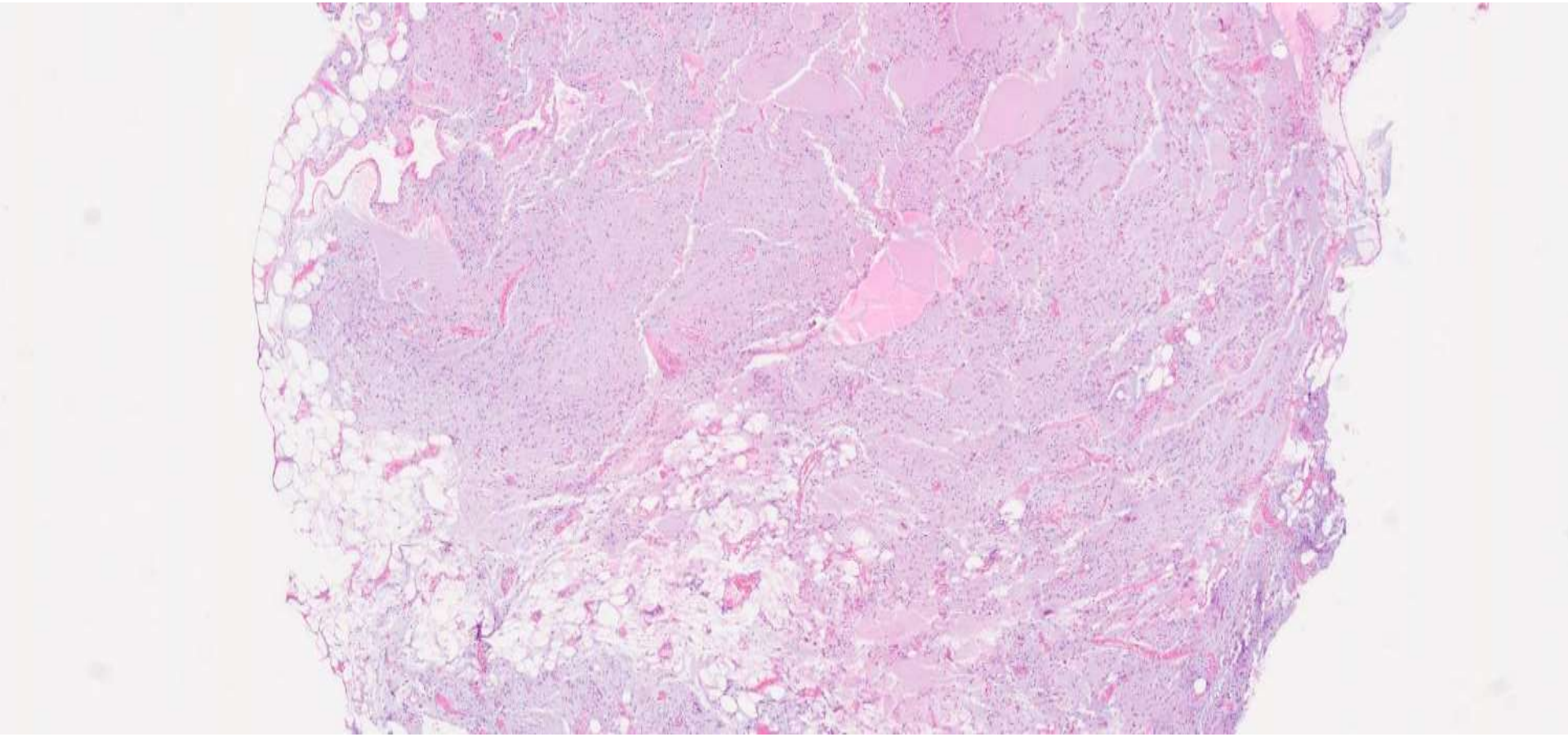


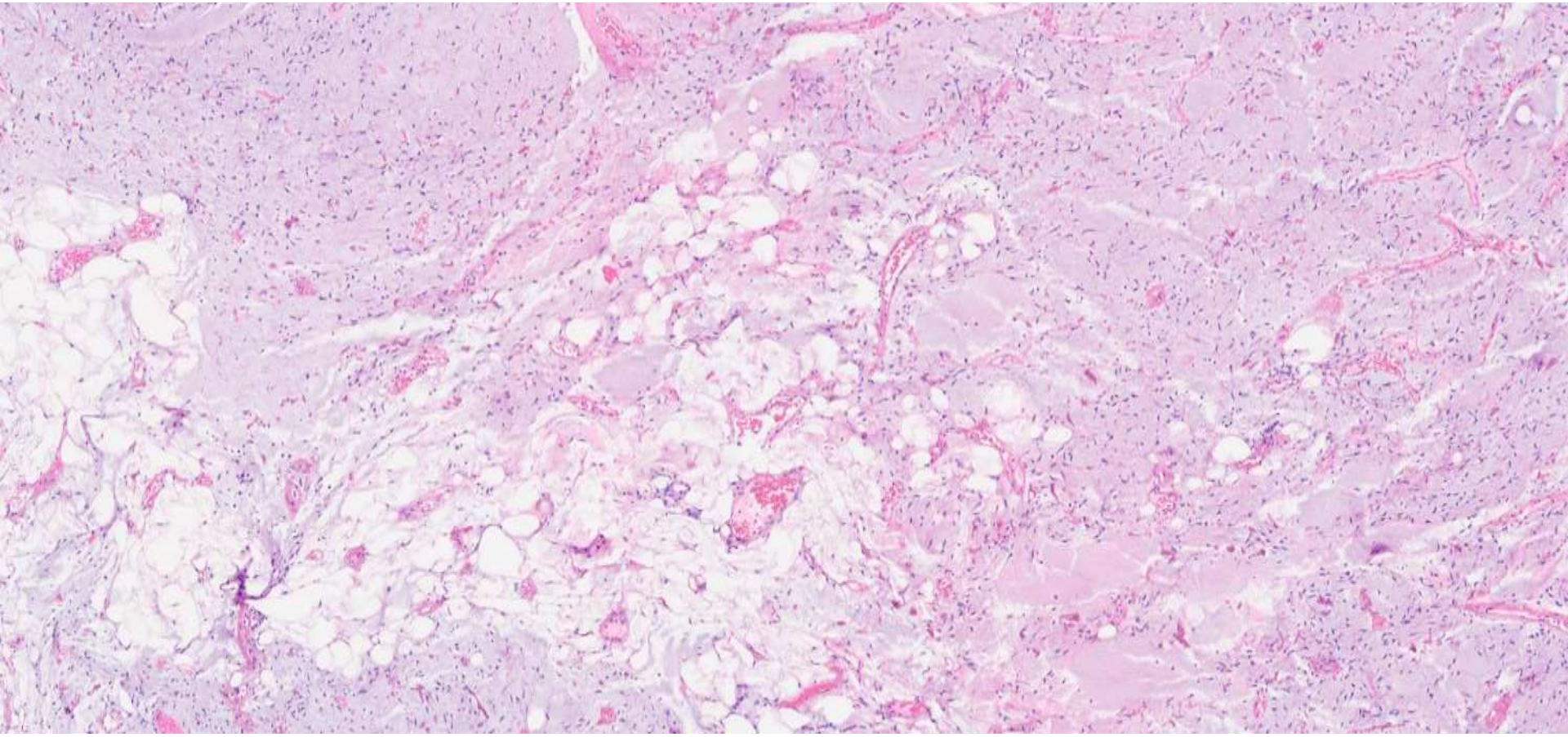


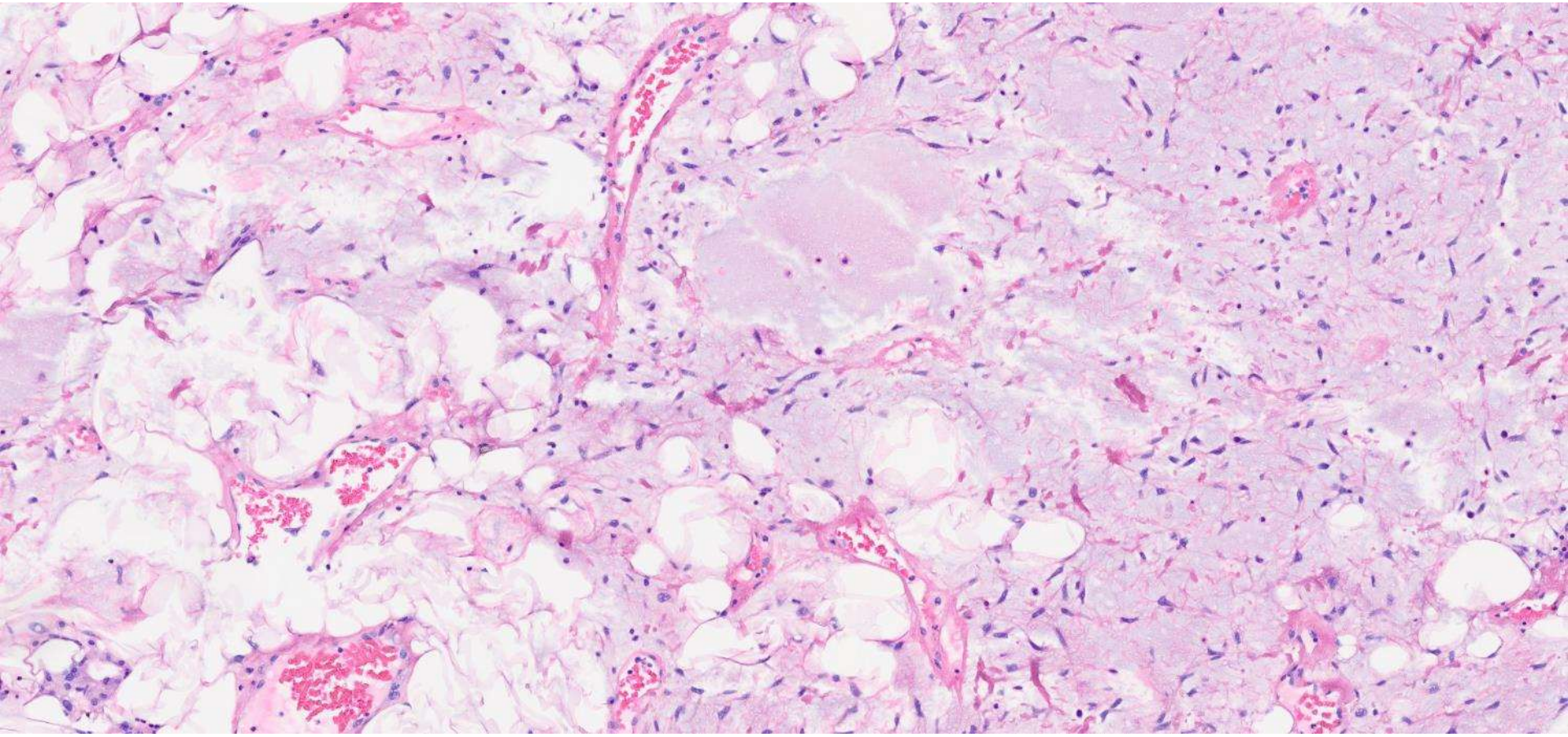




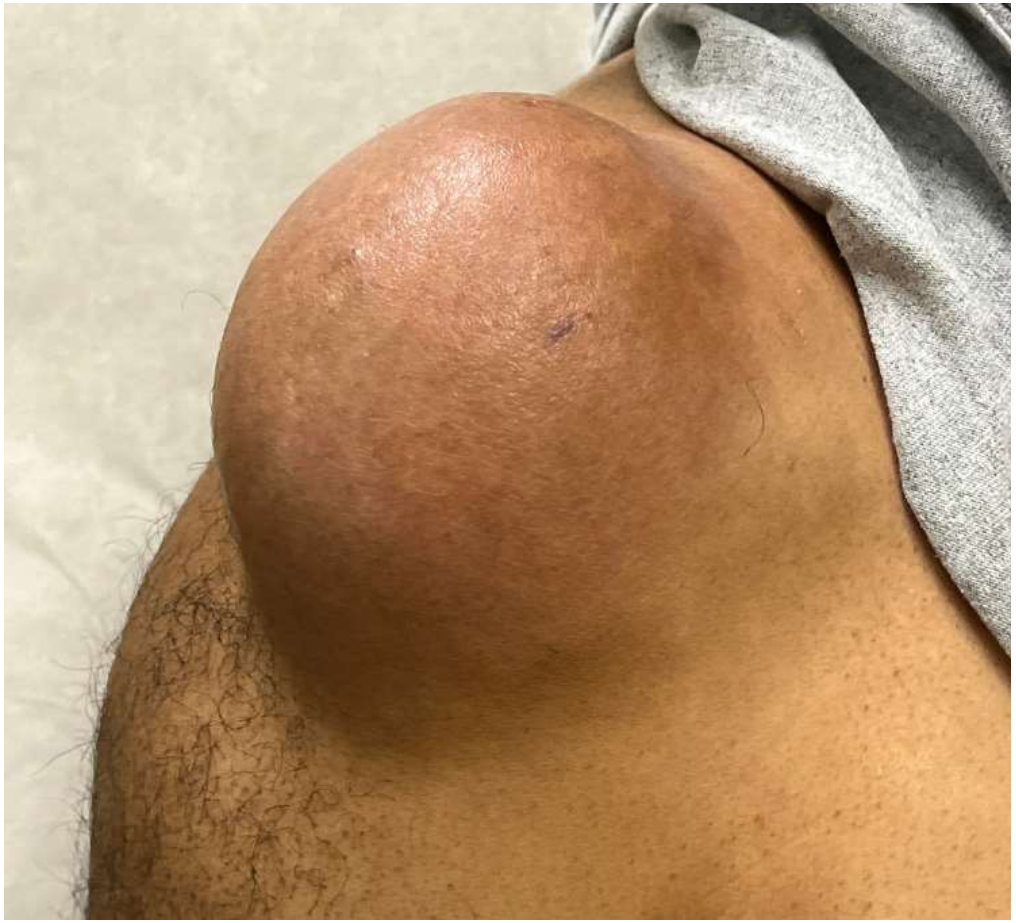








23-0205



23-0205

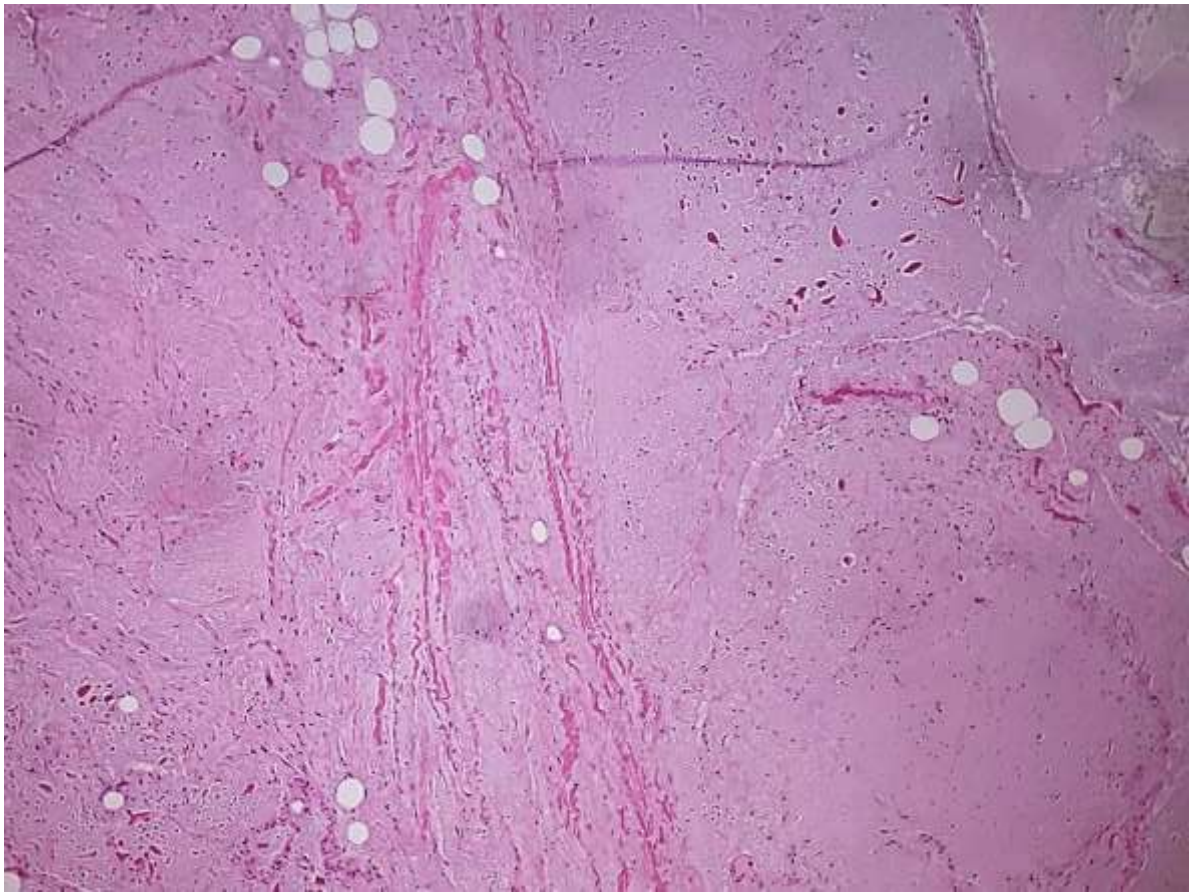
MRI SHOULDER

IMPRESSION:

Approximately 10.5 by 7.5 x 5.5 cm (ML x AP x CC) multilobulated multi-septated diffusely hyper-enhancing hyperintense T2 hypointense T1 mass in subcutaneous fat of right shoulder overlying deltoid muscle and AC joint. No fat density to suggest liposarcoma. No direct invasion to subjacent structures.

23-0205

23-0205



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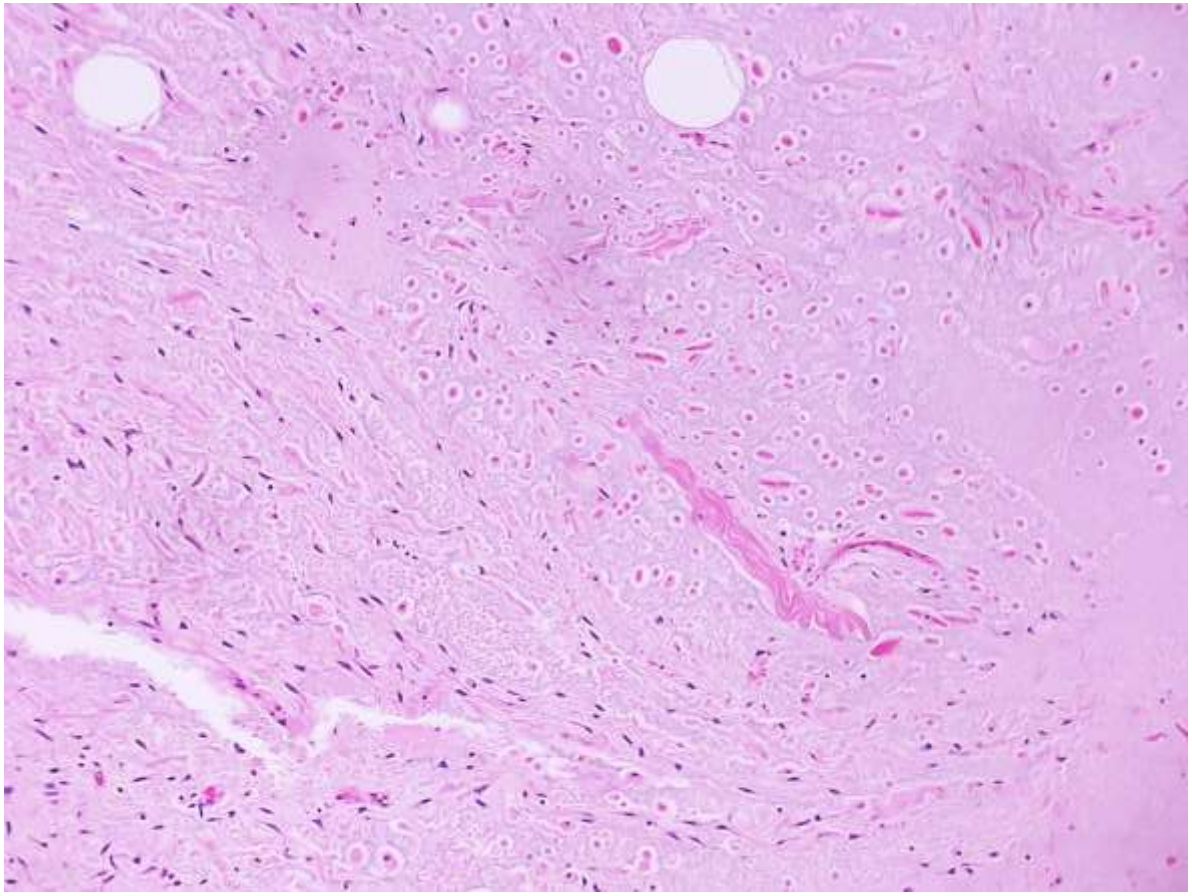

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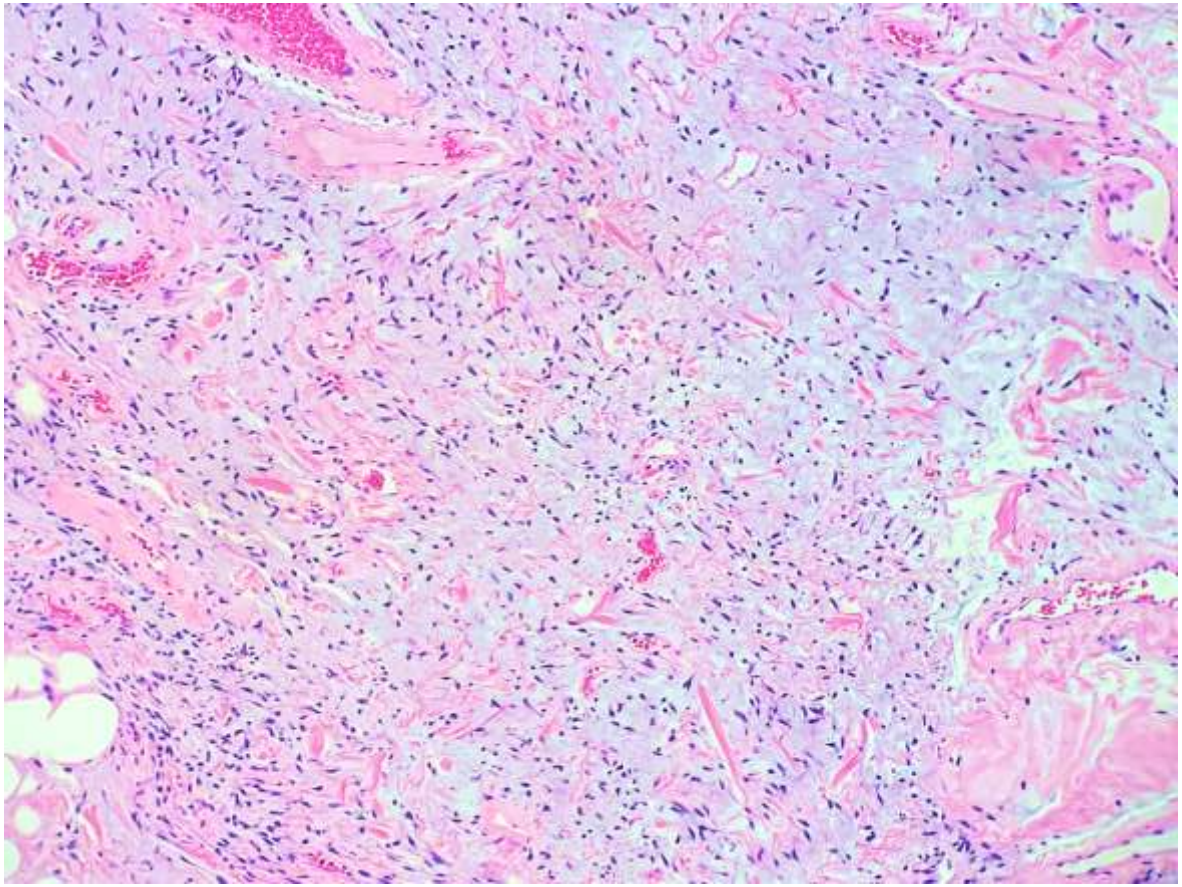

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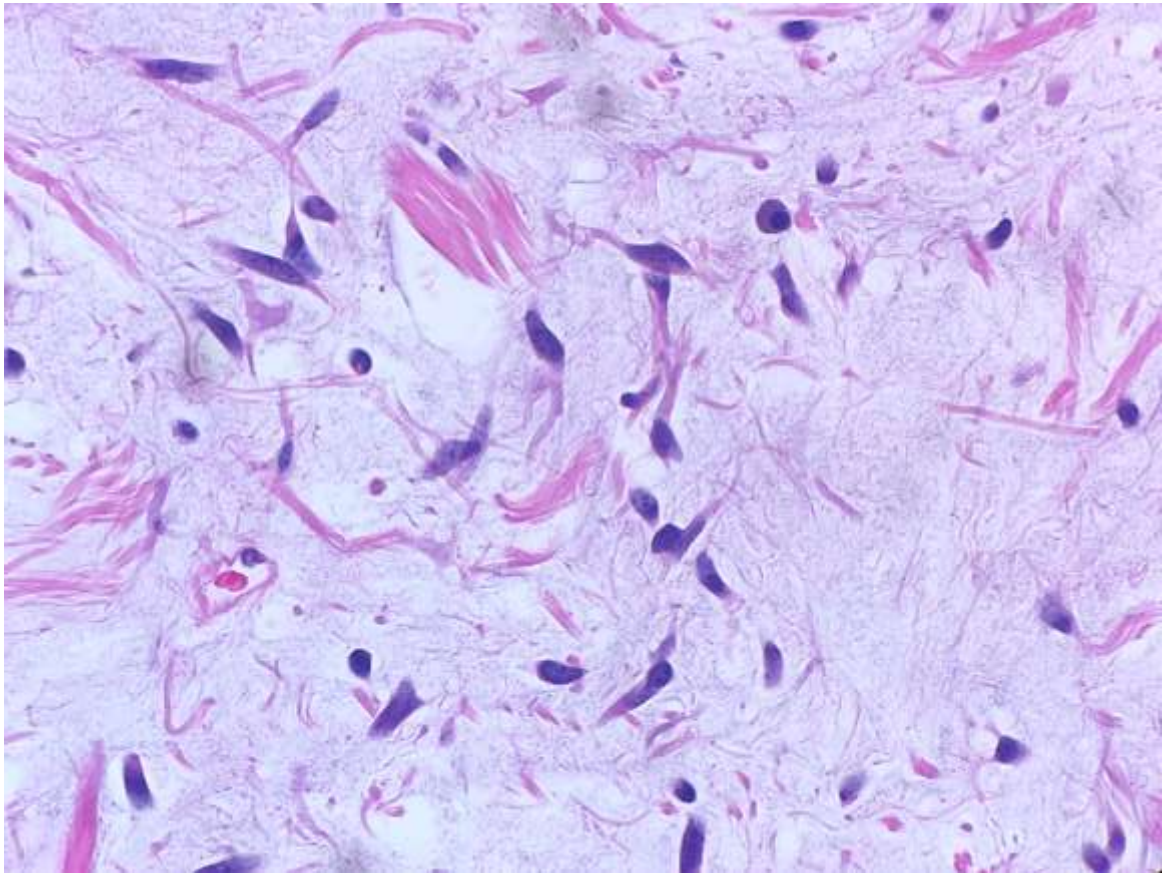

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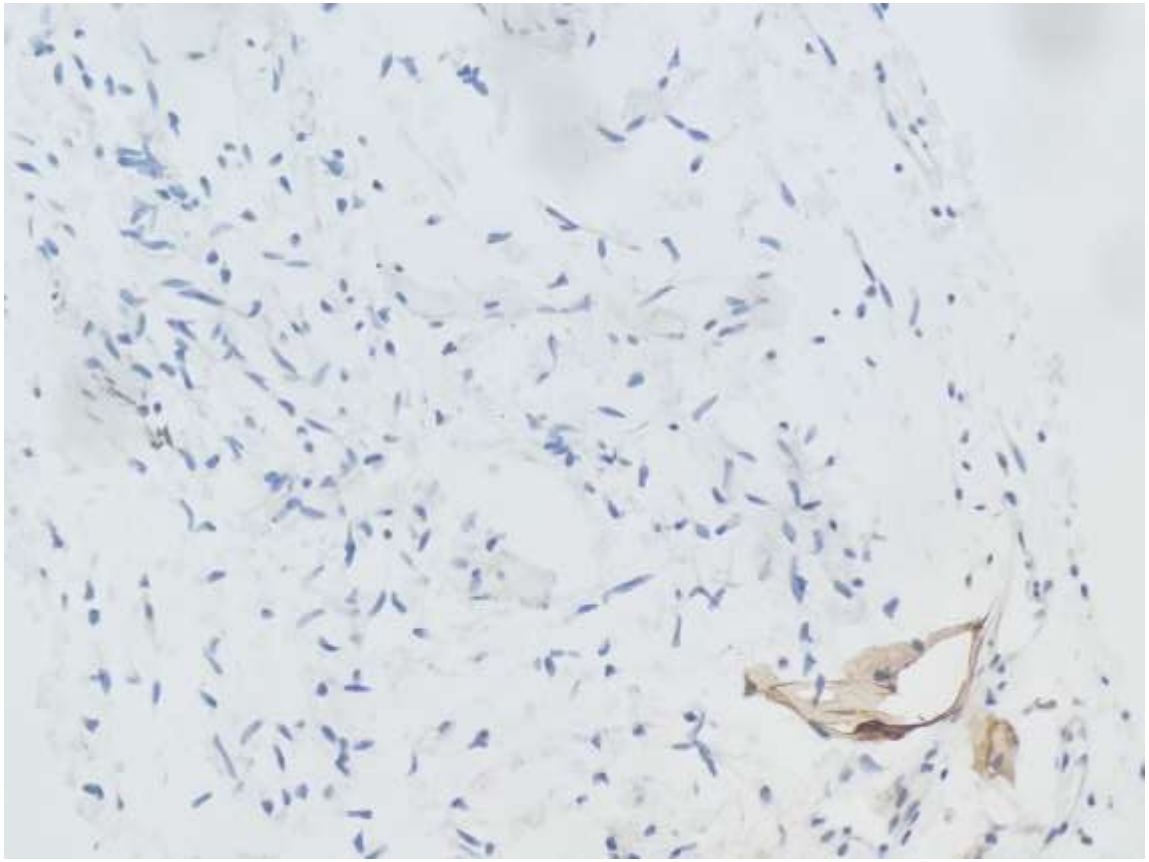
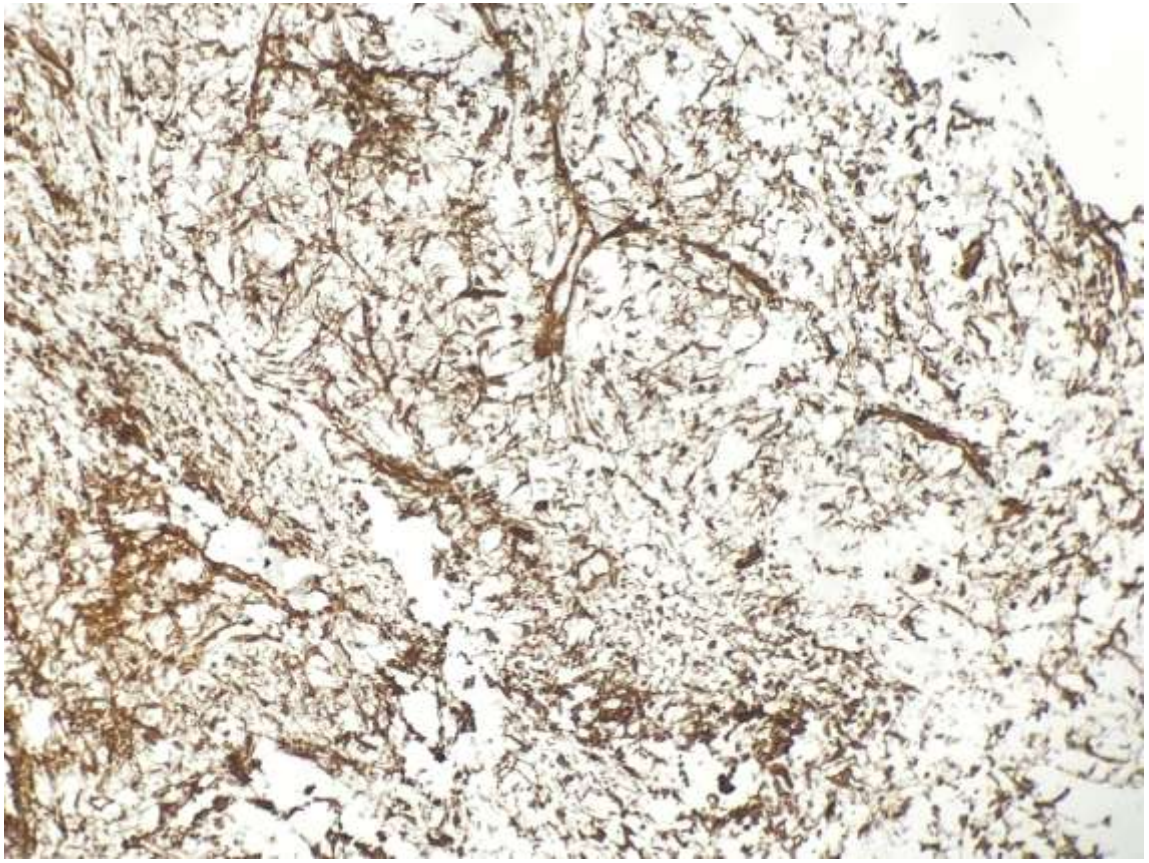


S-100

23-0205



CD34

23-0205

Spindle cell lipoma

- **Longstanding, mobile lesion in subcutaneous tissue; mean age 55 years; M>>>F (9:1).**
- **Shawl region of posterior neck / shoulder / upper back.**

- **Triad of mature adipocytes, bland spindle cells and hyalinized rope-like collagen fibers.**
- **Myxoid background with mast cells frequent.**
- **Infiltrative growth, necrosis, atypical spindle cells, pleomorphic lipoblasts and significant mitotic activity absent.**

- **CD34 positive, loss of nuclear RB1, negative for S-100, SOX10, SMA, desmin, STAT6.**

- **Characterized by partial or whole chromosome 13 or 16 deletions.**
- **Breakpoints cluster around 13q14 where the RB1 gene resides.**
- **Part of the so called 13q / RB1 family of tumors, which includes myofibroblastoma and cellular angiofibroma.**
- **Lacks MDM2 and CDK4 amplifications and FUS gene rearrangements.**

23-0205

Differential Diagnosis for a cytologically-bland spindle cell lesion with a myxoid matrix:

- **Spindle Cell Lipoma**
- **Atypical Lipomatous Tumor/Well-Differential Liposarcoma**
  - **MDM2 positive by IHC or MDM2 amplification by FISH**
- **Myxoid Liposarcoma**
  - **Small signet ring lipoblasts**
  - **FUS-DDIT3 gene rearrangement**
- **Peripheral Nerve Sheath Tumor**
  - **Positive for S-100 and SOX10**
- **Intramuscular Myxoma**
  - **Intramuscular, no fat or thick bundles of collagen; hypocellular**
- **Low-Grade Myxofibrosarcoma**
  - **Frequently in limbs and limb girdles**
  - **Elongated, curvilinear blood vessels**
  - **Pseudolipoblasts may be present**
  - **Varying cellularity and histologic grade**
- **Solitary Fibrous Tumor**
  - **STAT6, BCL2 positive**

23-0205

- **References:**

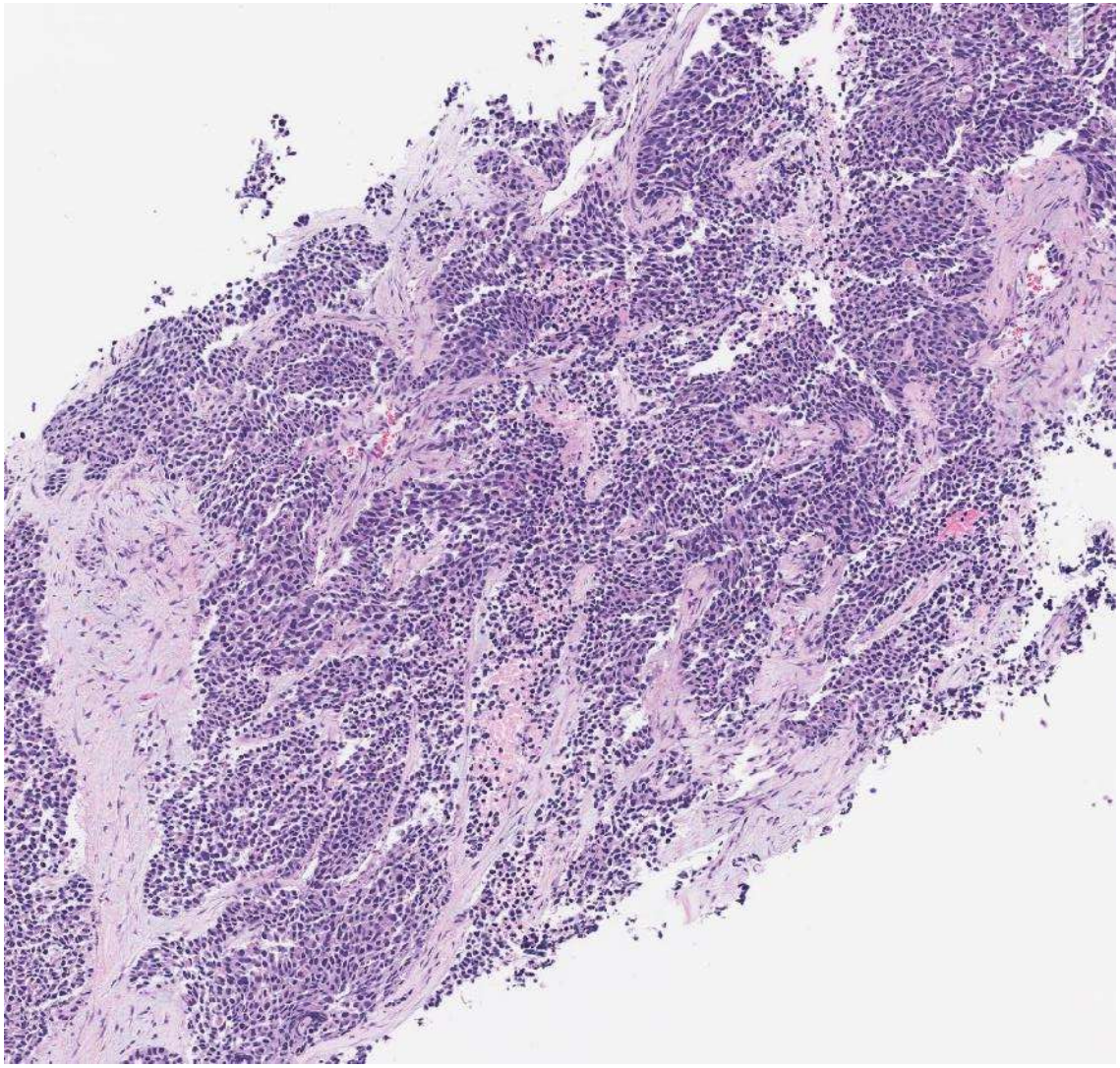
  - Wong YP, Chia WK, Low SF, Mohamed-Haflah NH, Sharifah NA. Dendritic fibromyxolipoma: a variant of spindle cell lipoma with extensive myxoid change, with cytogenetic evidence. Pathol Int. 2014 Jul;64(7):346-51.

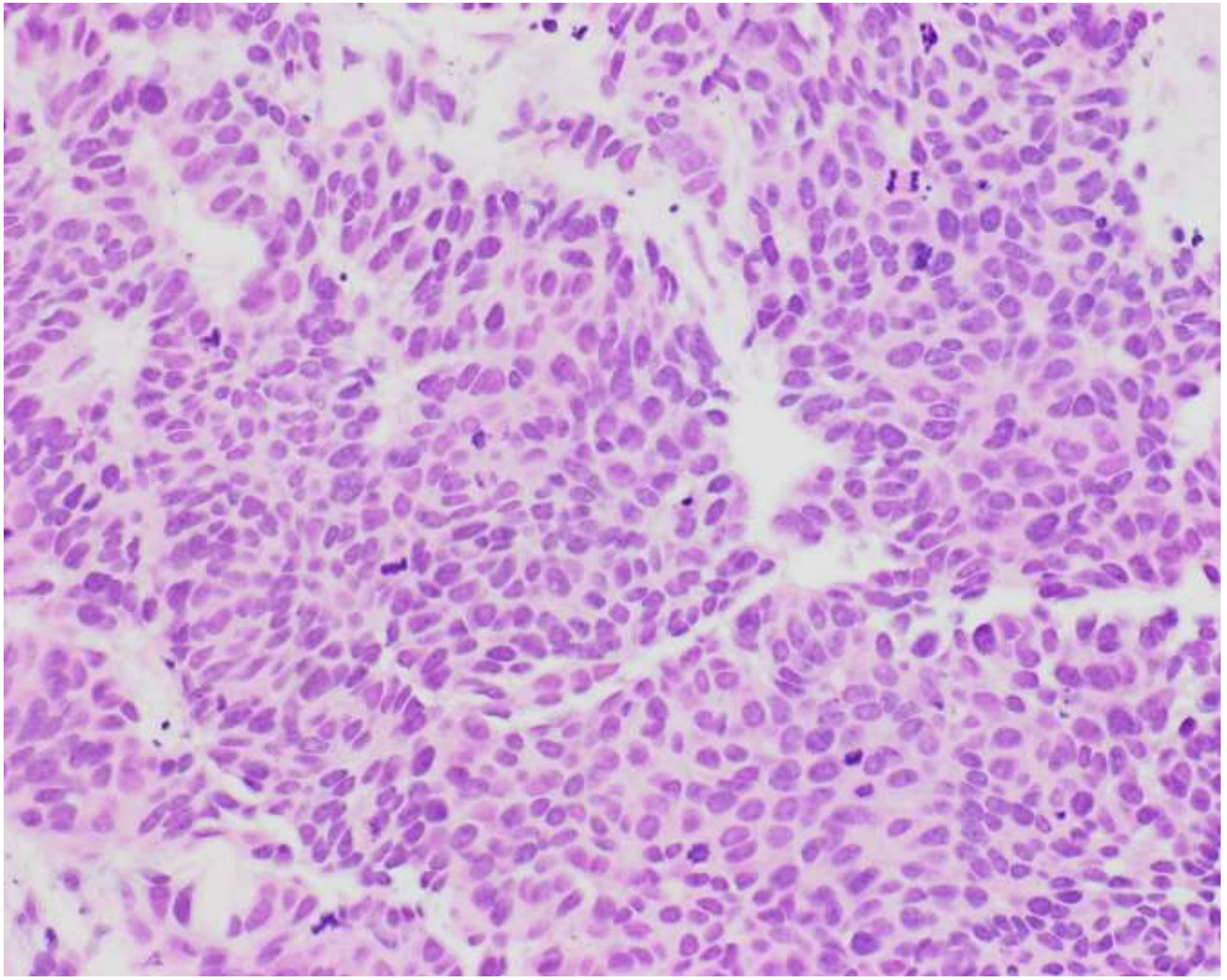
  - Hawley IC, Krausz T, Evans DJ, Fletcher CD. Spindle cell lipoma--a pseudoangiomatous variant. Histopathology. 1994 Jun;24(6):565-9.

23-0206

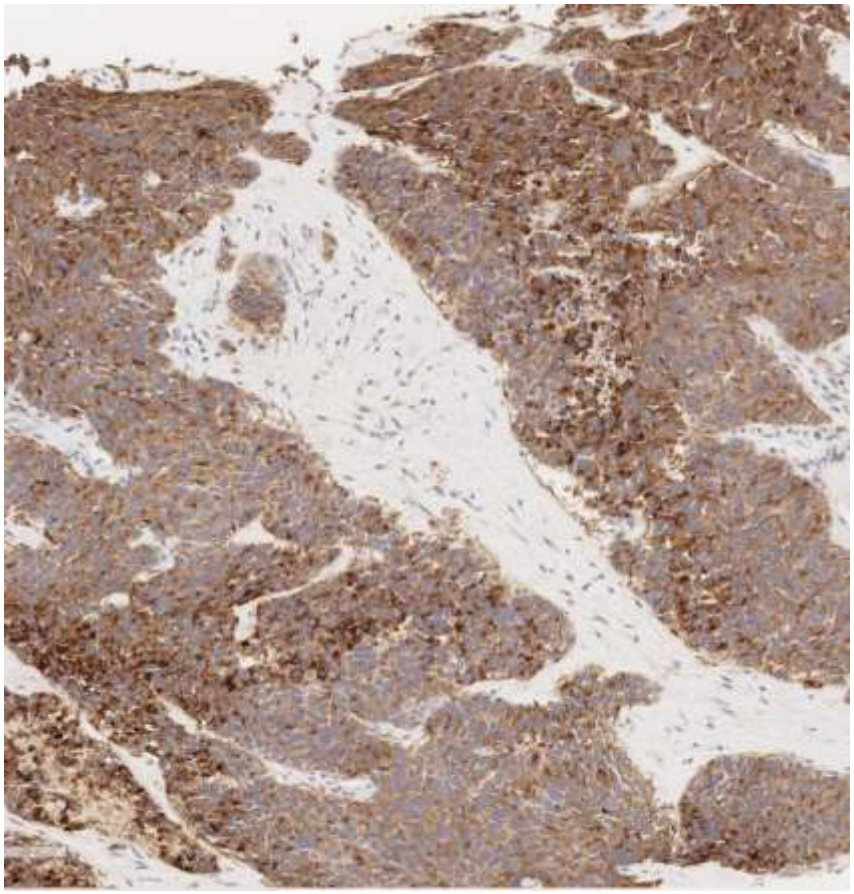
Nancy Joseph, UCSF

60ish F with pancreatic and liver masses. Undergoes liver mass biopsy.

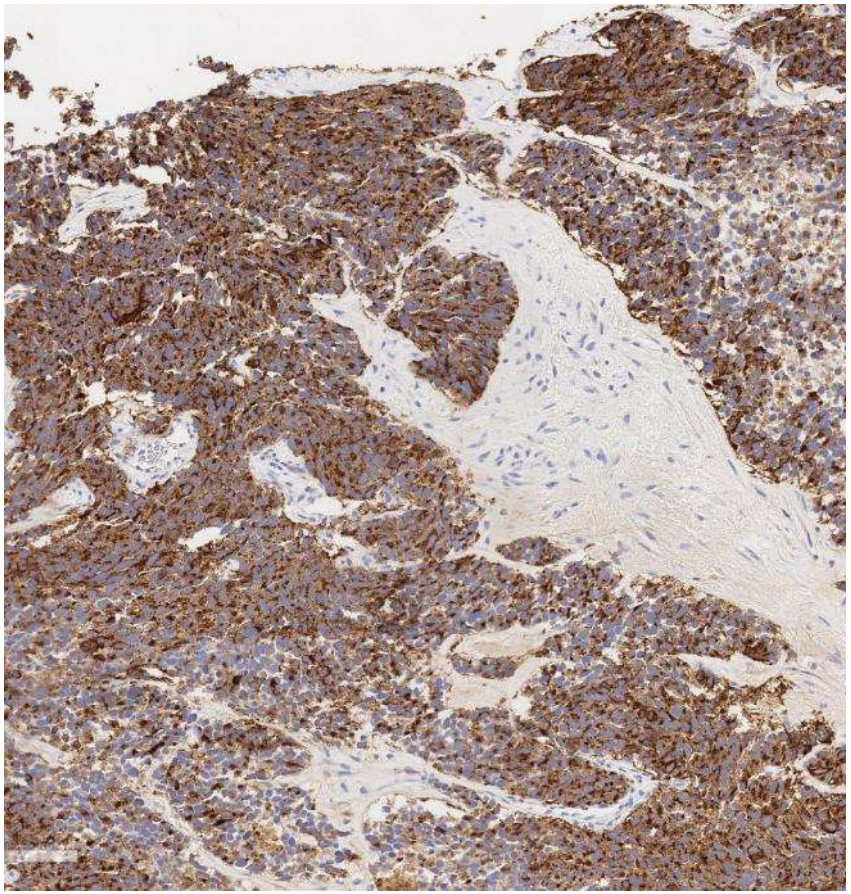




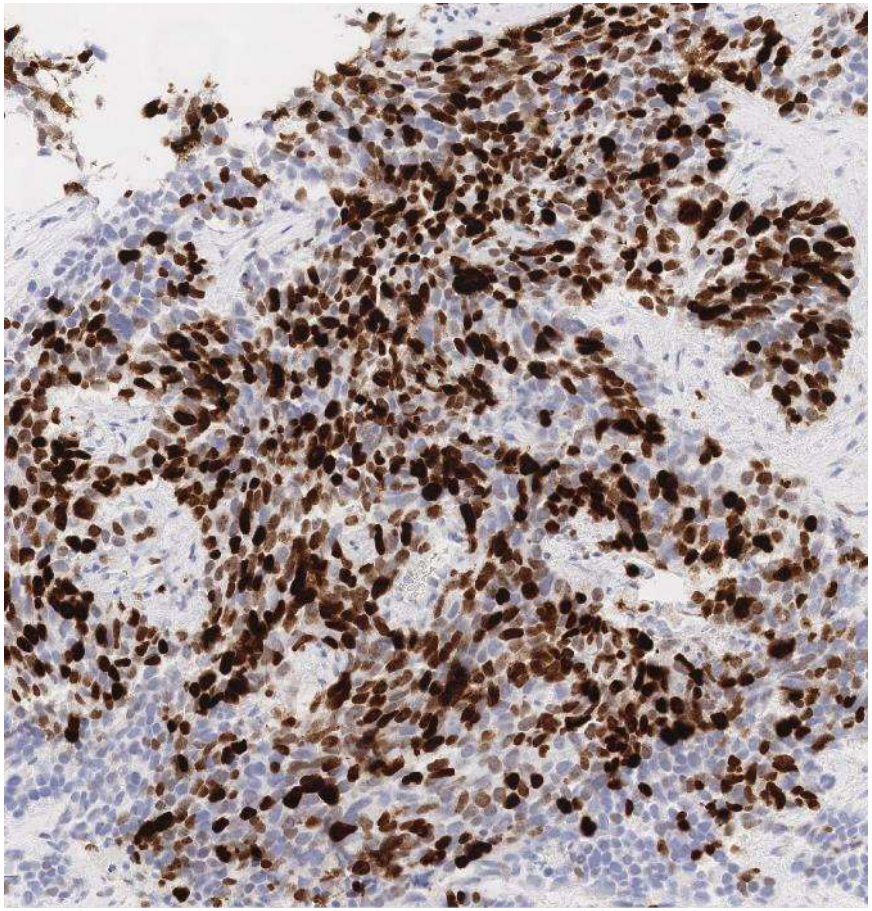
Synaptophysin



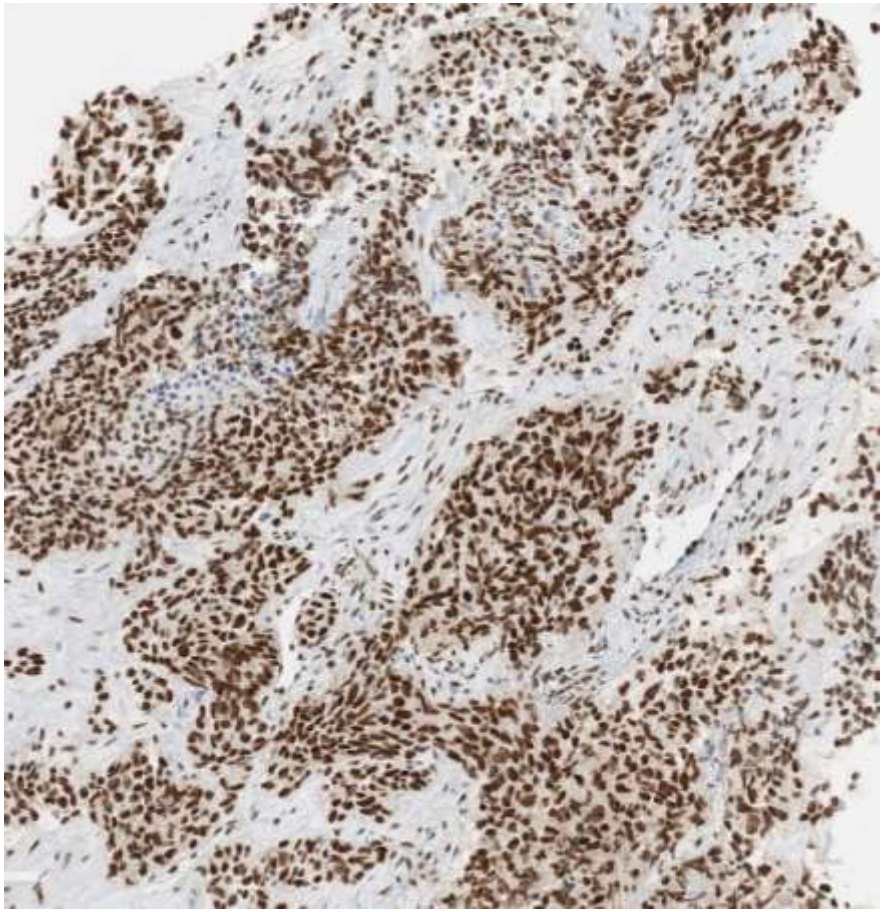
Chromogranin



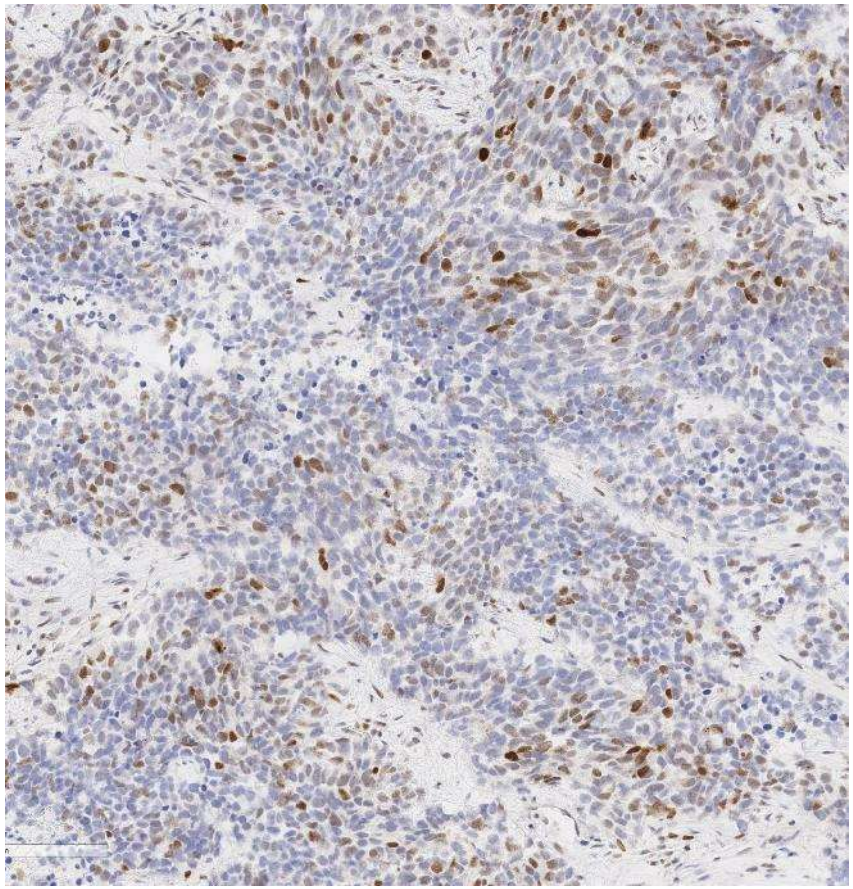
Ki-67



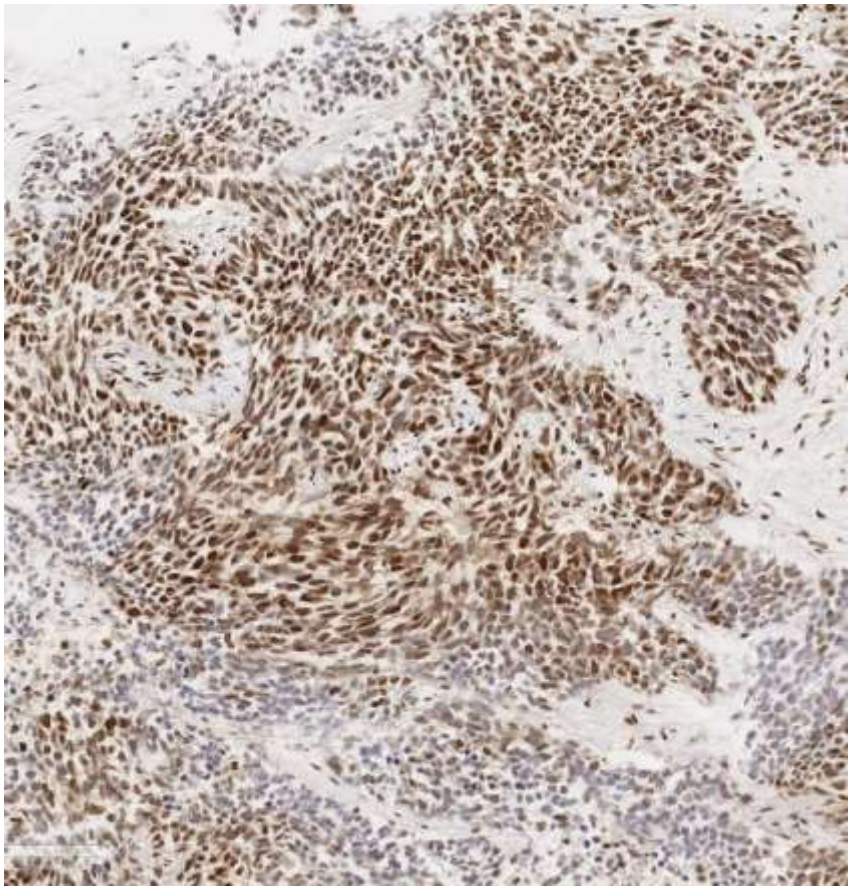
ATRX



p53



Rb

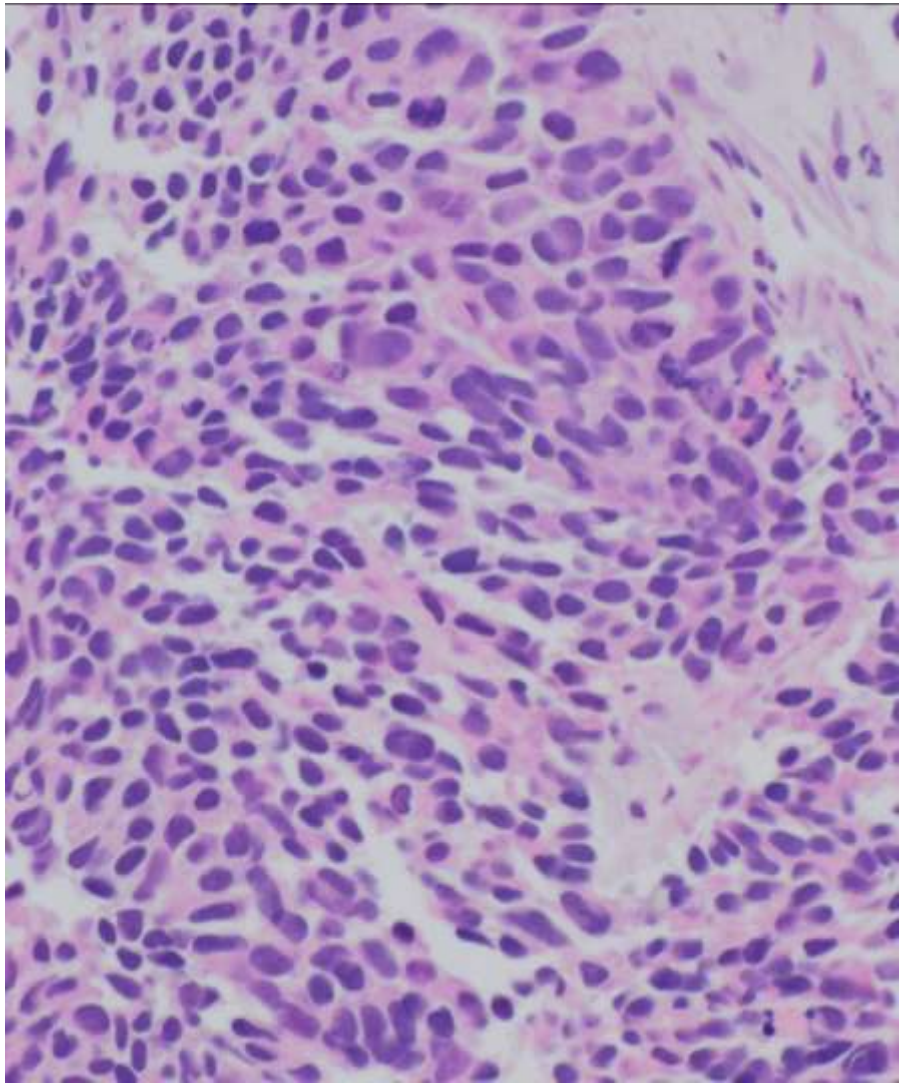


DIAGNOSIS?

Differential Diagnosis?

- Neuroendocrine Carcinoma (NEC)
- Grade 3 neuroendocrine tumor (NET)

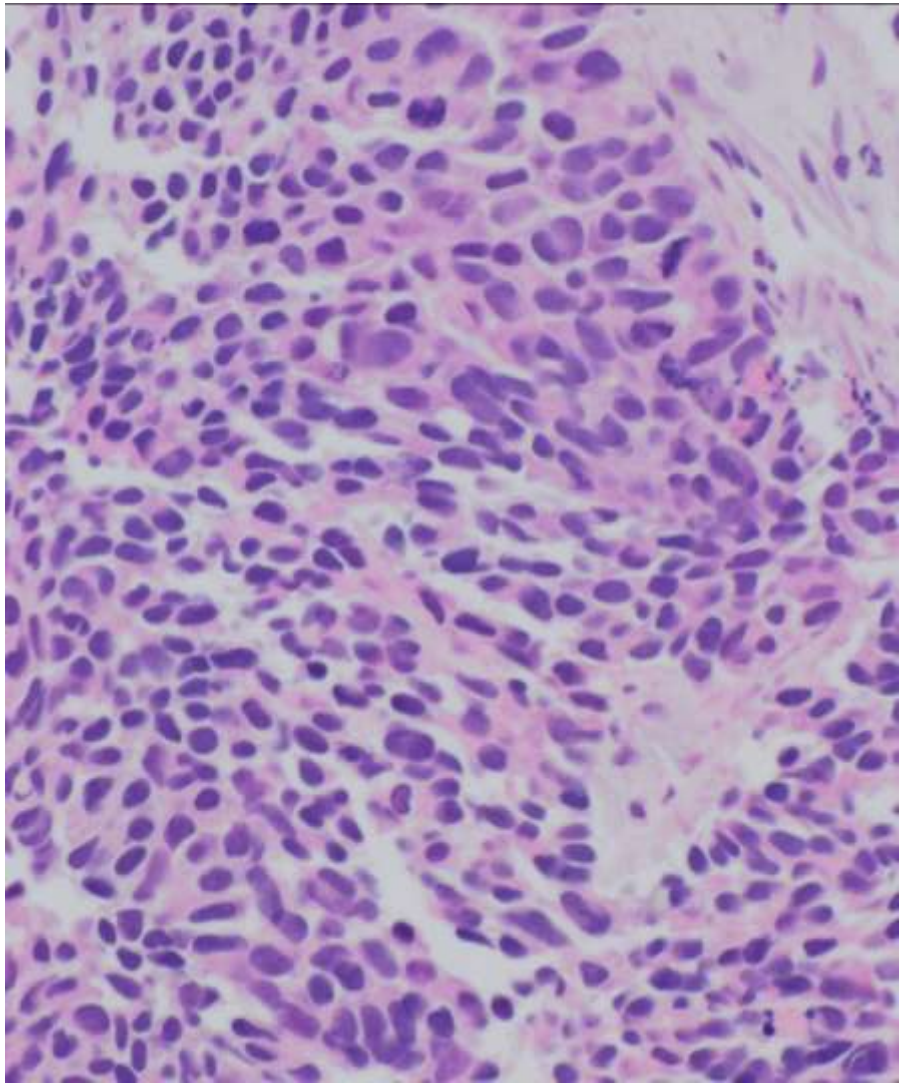
Anything else?



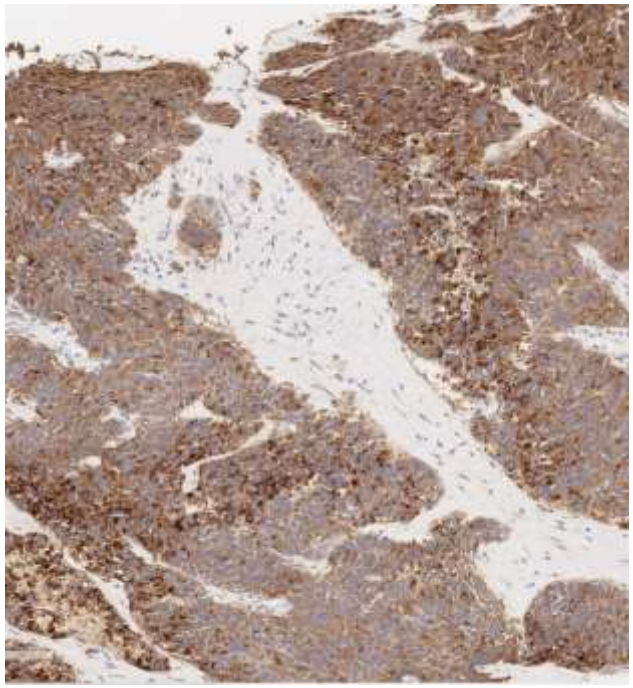
Differential Diagnosis?

- Neuroendocrine Carcinoma (NEC)
- Grade 3 neuroendocrine tumor (NET)

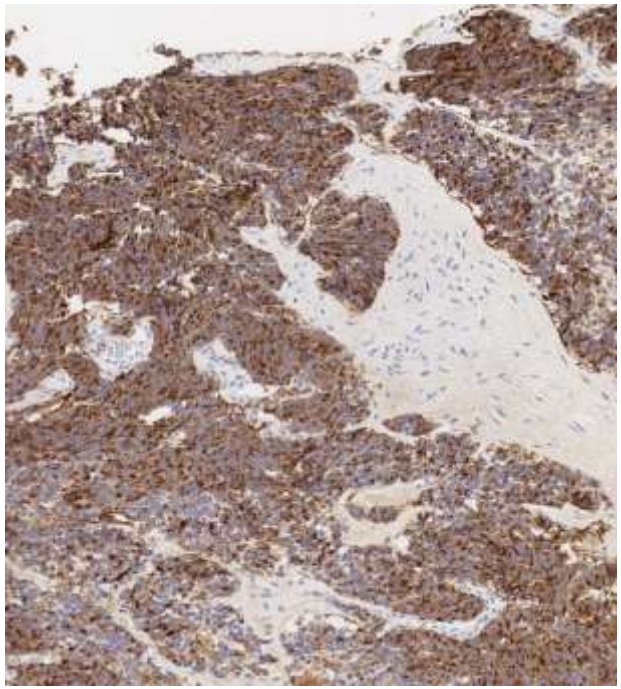
- Acinar cell carcinoma with NE expression



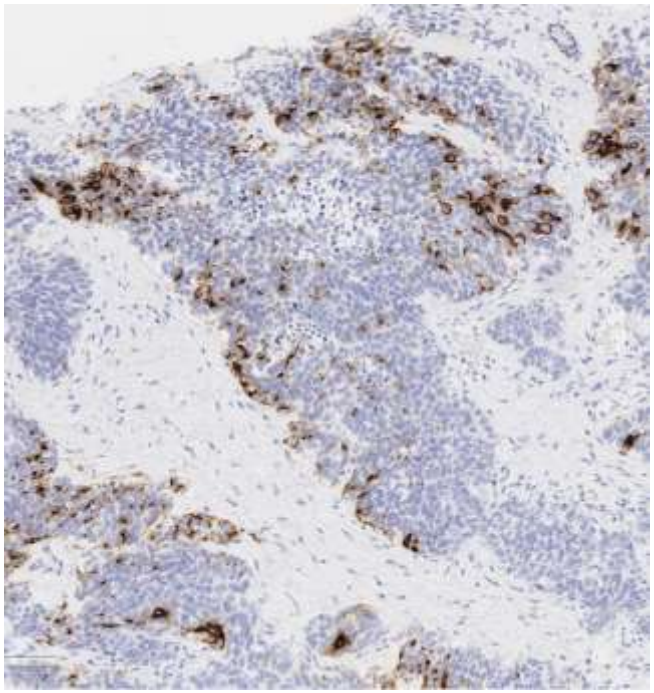
Acinar Cell Carcinoma with NE expression
(mixed acinar cell carcinoma-neuroendocrine carcinoma, ACC-NEC)



Synaptophysin



Chromogranin



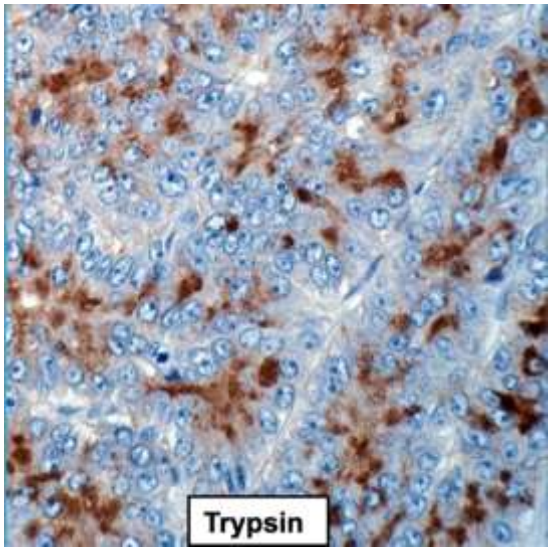
Trypsin

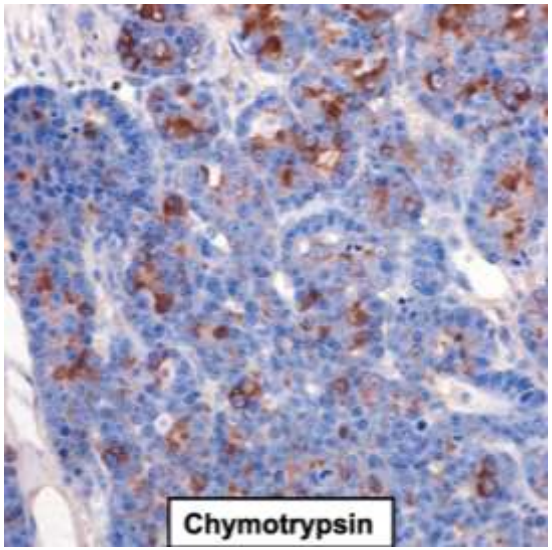
NGS Results: mutations in ***APC*** and ***ATM***

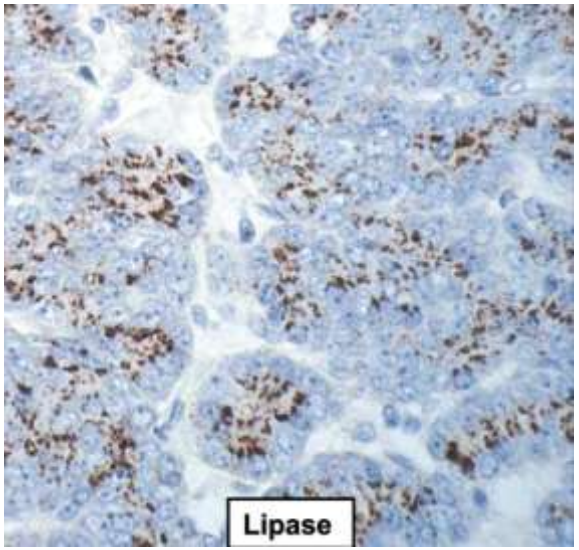
	NET	NEC	Acinar cell carcinoma	ACC with NE expression (mixed ACC-NEC
Pancytokeratin	+	+	+	+
Synaptophysin	+	+	+/-	+
Chromogranin	+	+	+/-	+
ACC markers (trypsin, lipase, chymotrypsin)	-	-	+	+
Genetics	*MEN1, ATRX, DAXX, SETD2, TSC1/2* (up to 35% of G3 NET have *TP53)*	*TP53, RB1*, site-specific adenocarcinoma genes (*KRAS, SMAD4*)	*APC, STK11, PRKAR1A, ATM, BRAF in-dels/fusions*	Same as ACC
Prognosis	Better than NEC, but G3 NET can be similar to NEC	Worst	Better than NEC and G3 NET	Same as ACC

Take Home Point

Stain all high-grade pancreatic neuroendocrine neoplasms (NET and NEC) with two ACC markers (trypsin, chymotrypsin, or lipase or BCL10 clone that cross-reacts with lipase)







Chmielecki J, et al. Cancer Discov. 2014;4(12):1398-405.

MODERN PATHOLOGY

USCAP

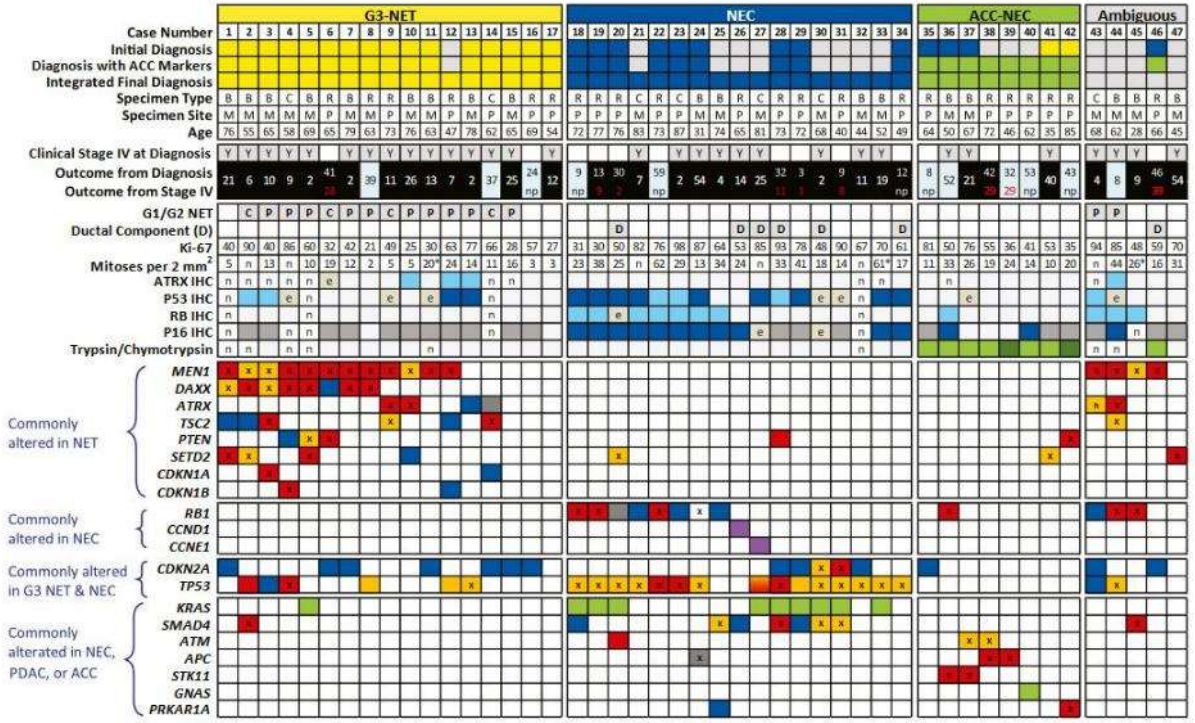
Journal homepage: https://modernpathology.org/

Research Article

Integrated Genomic and Clinicopathologic Approach Distinguishes Pancreatic Grade 3 Neuroendocrine Tumor From Neuroendocrine Carcinoma and Identifies a Subset With Molecular Overlap

Sarah E. Umetsu[a], Sanjay Kakar[a], Olca Basturk[b], Grace E. Kim[a], Kwun Wah Wen[a], Gillian Hale[d], Nafis Shafizadeh[e], Soo-Jin Cho, Ryan M. Gill[a], Kirk D. Jones[a], Pooja Navale[f], Emily Bergsland[g], Nancy M. Joseph[a,*]

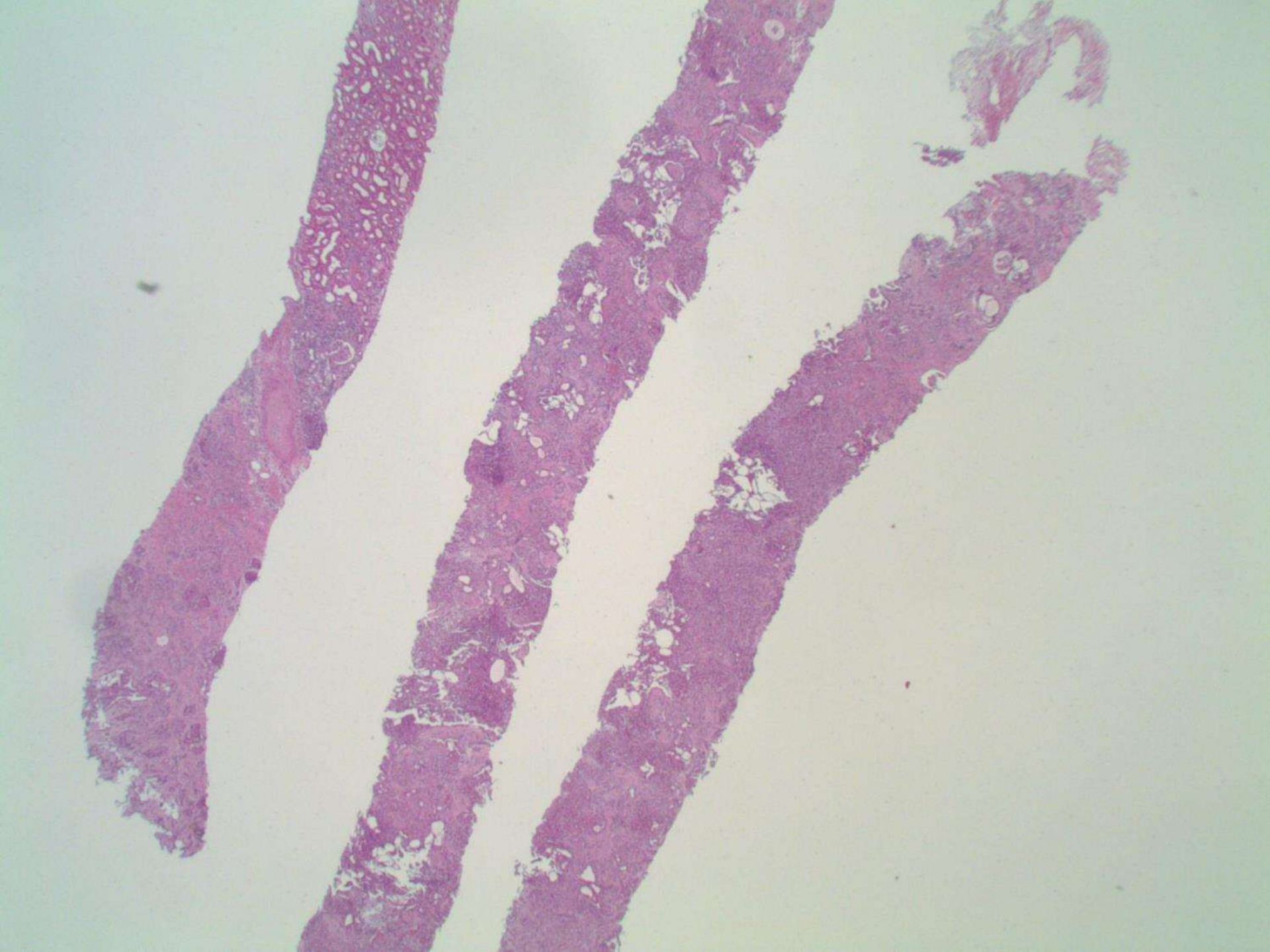
Recent study shows 8/47 (17%) of cases diagnosed as either pancreatic NEC or G3 NET are actually ACC with NE expression

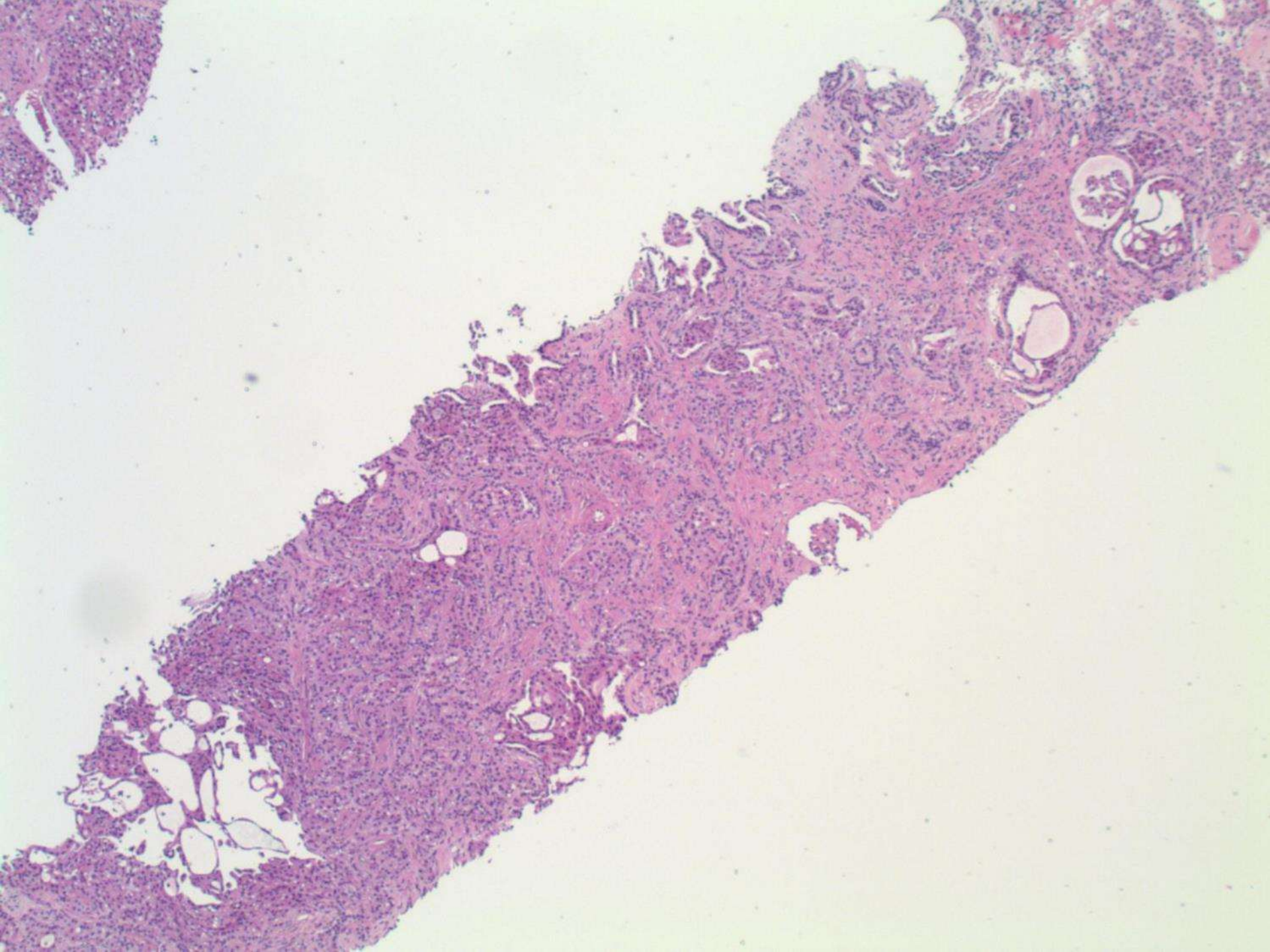


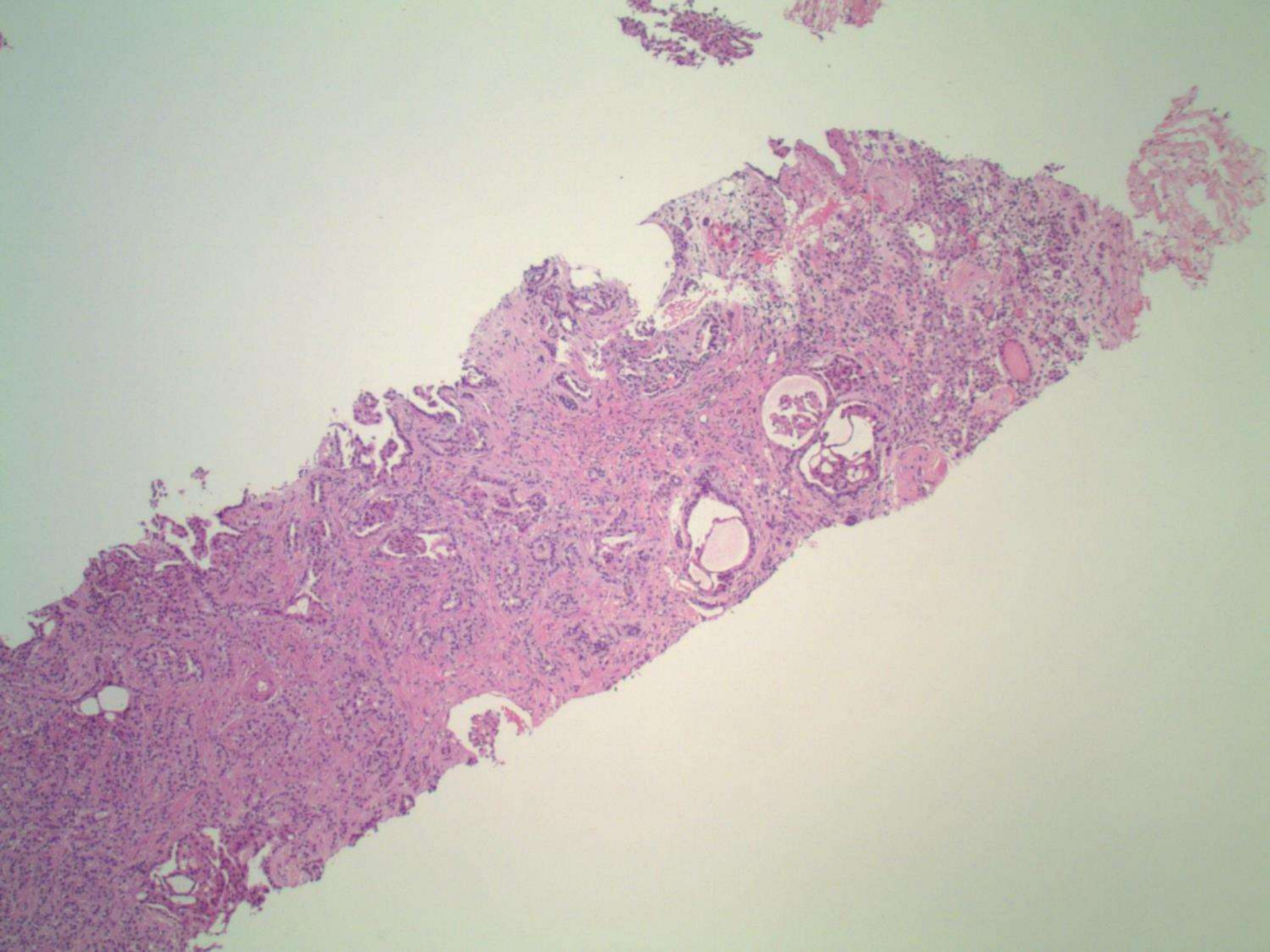
23-1207

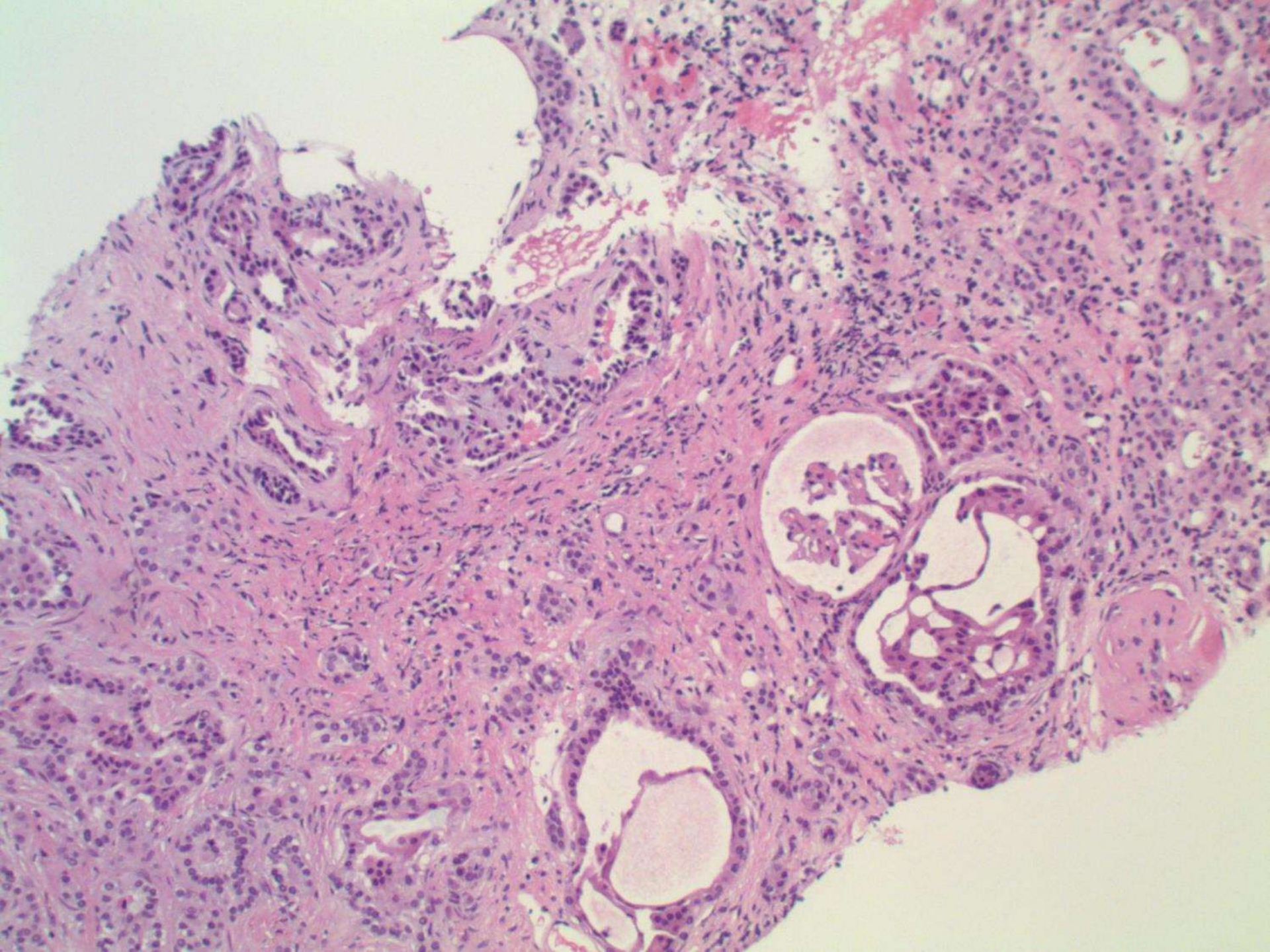
Ankur Sangoi; El Camino Hospital

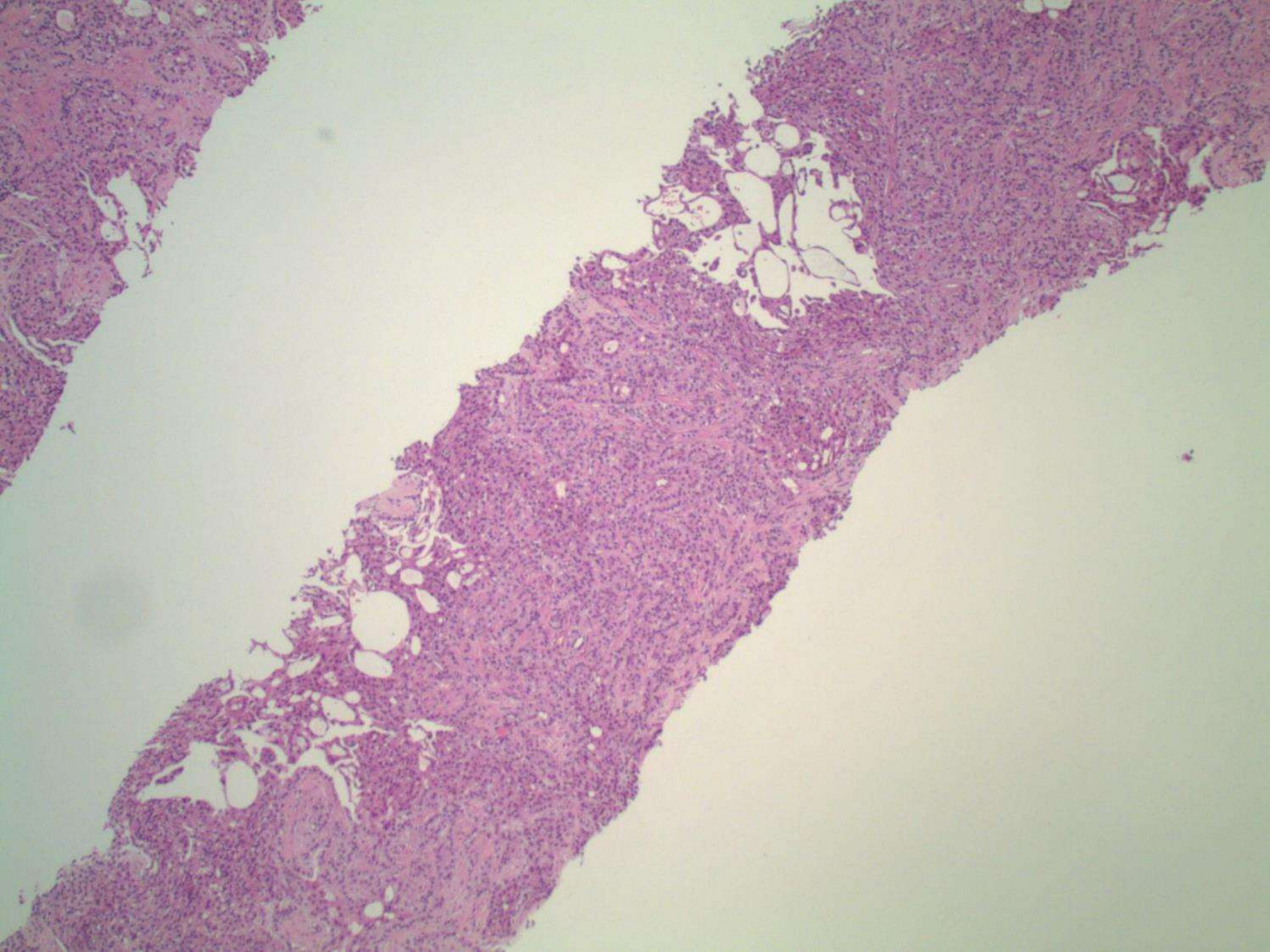
60ish M presenting with 4.5cm renal mass, undergoes biopsy.

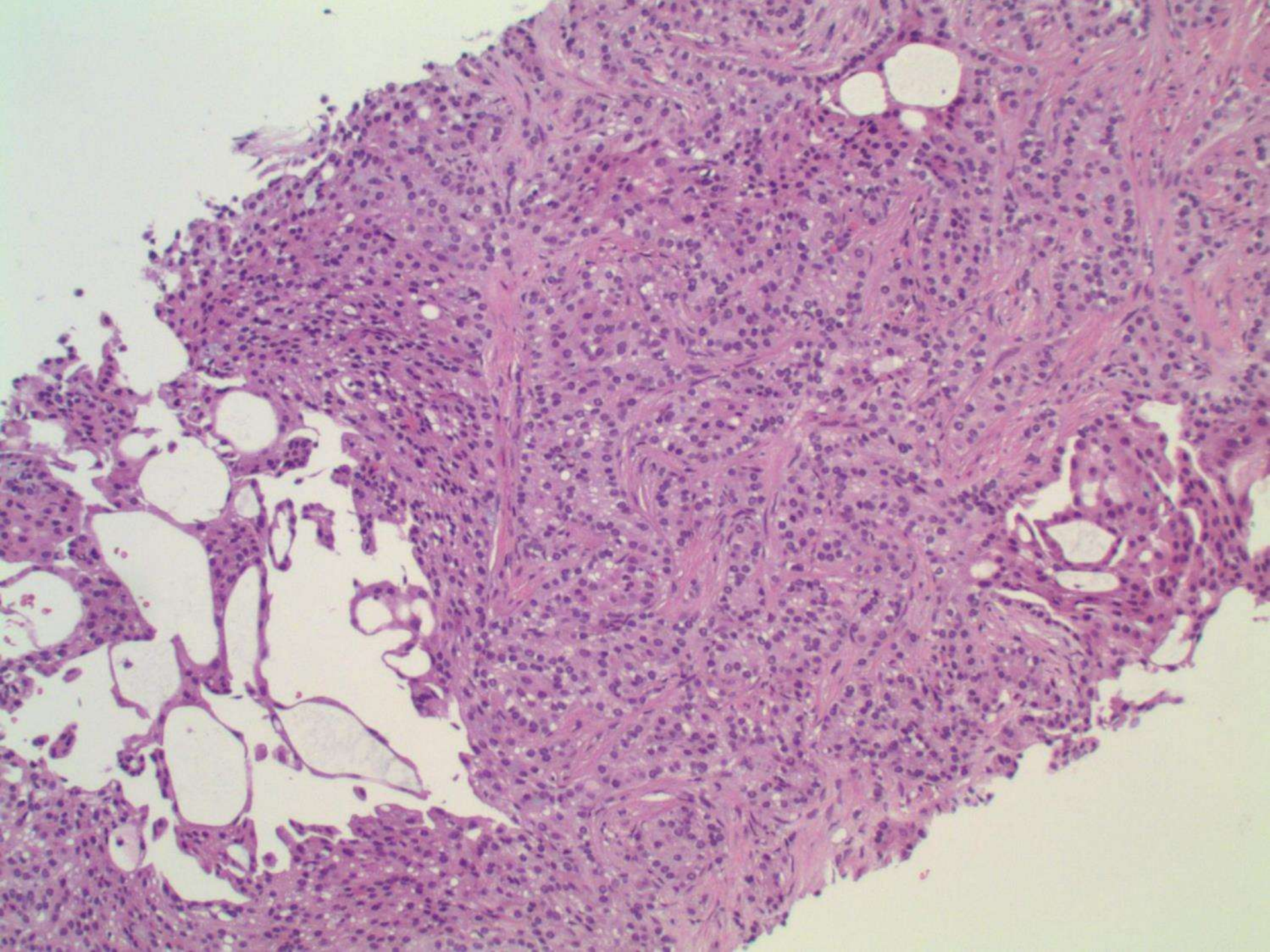


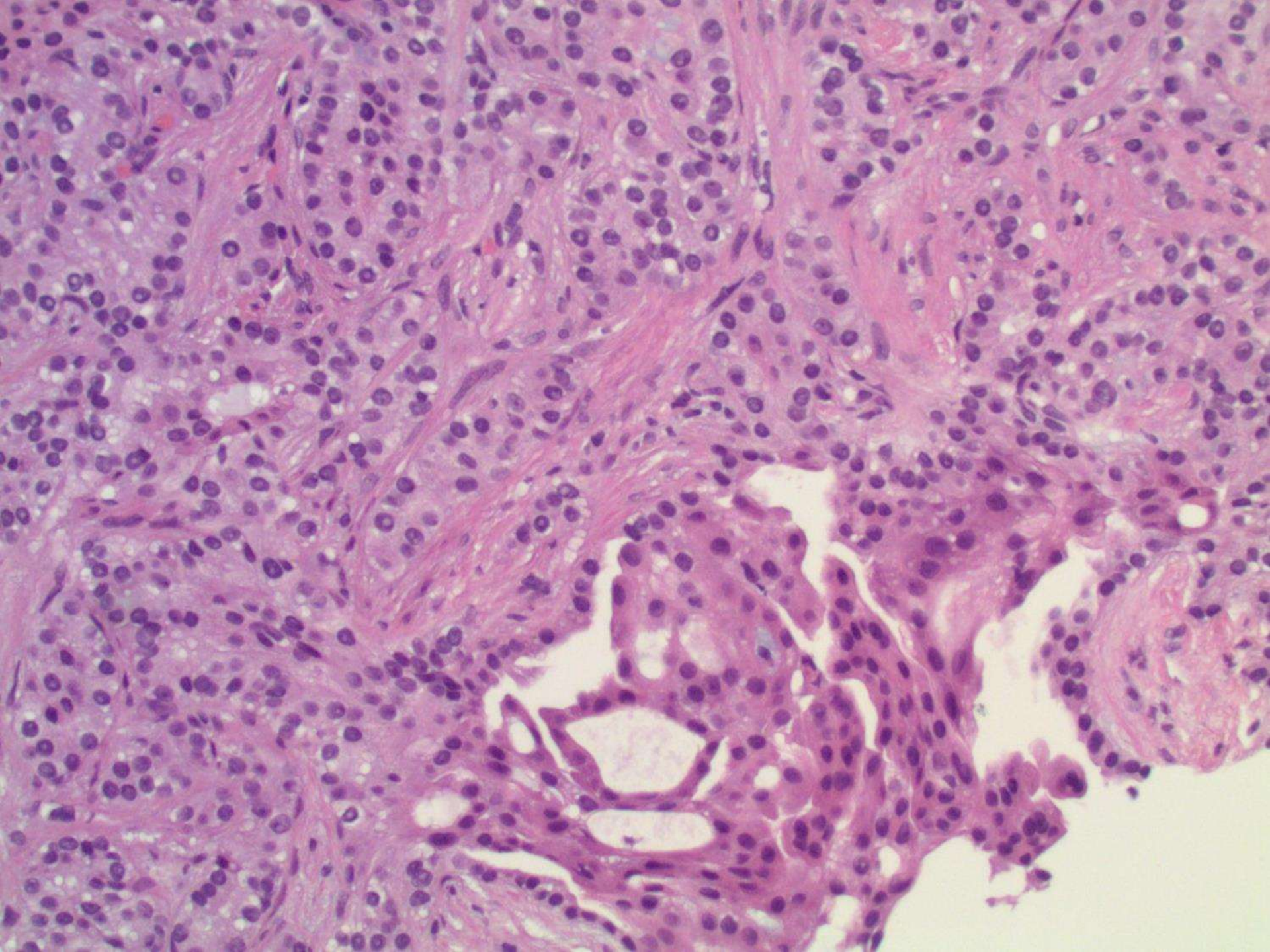


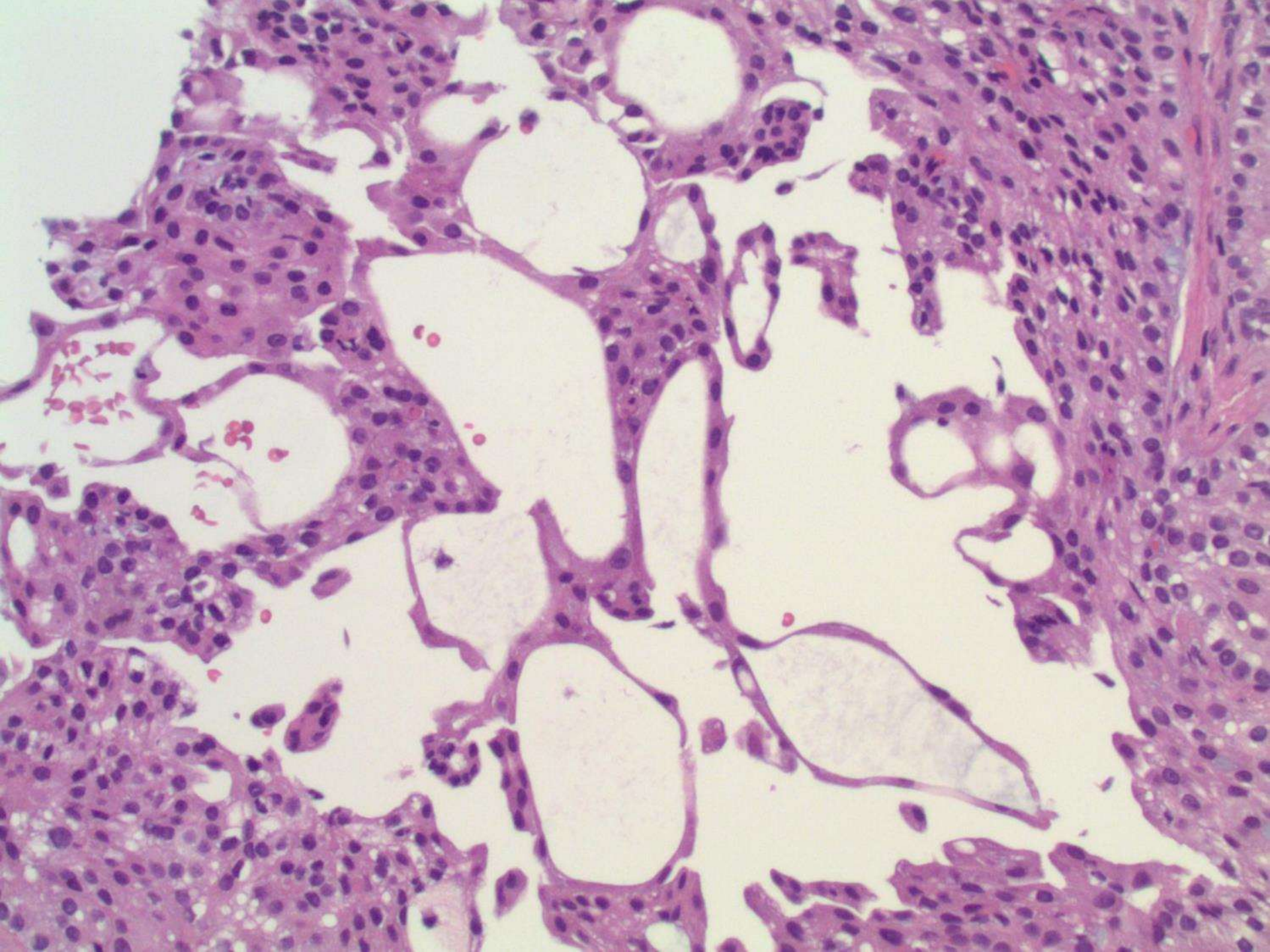


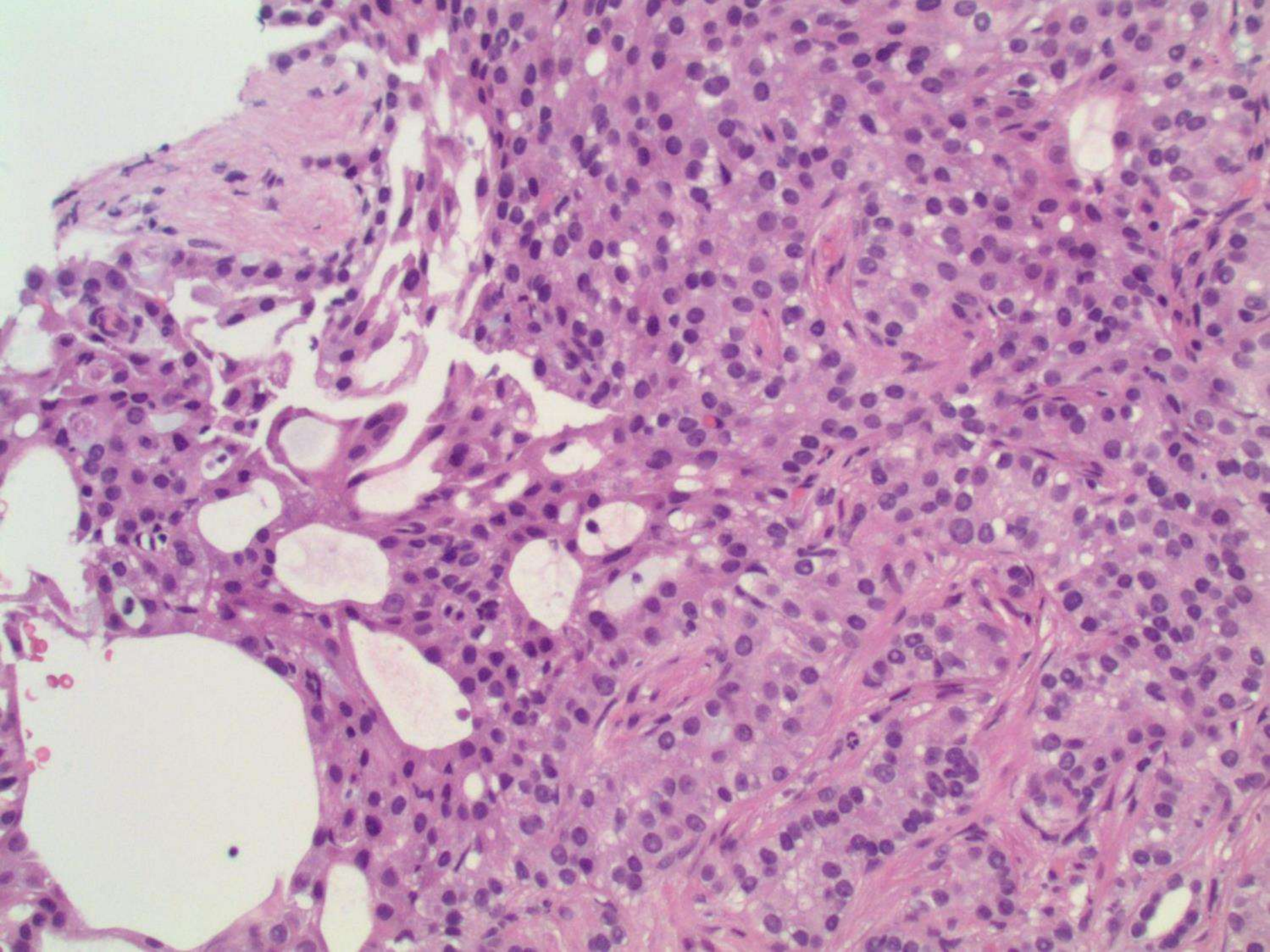


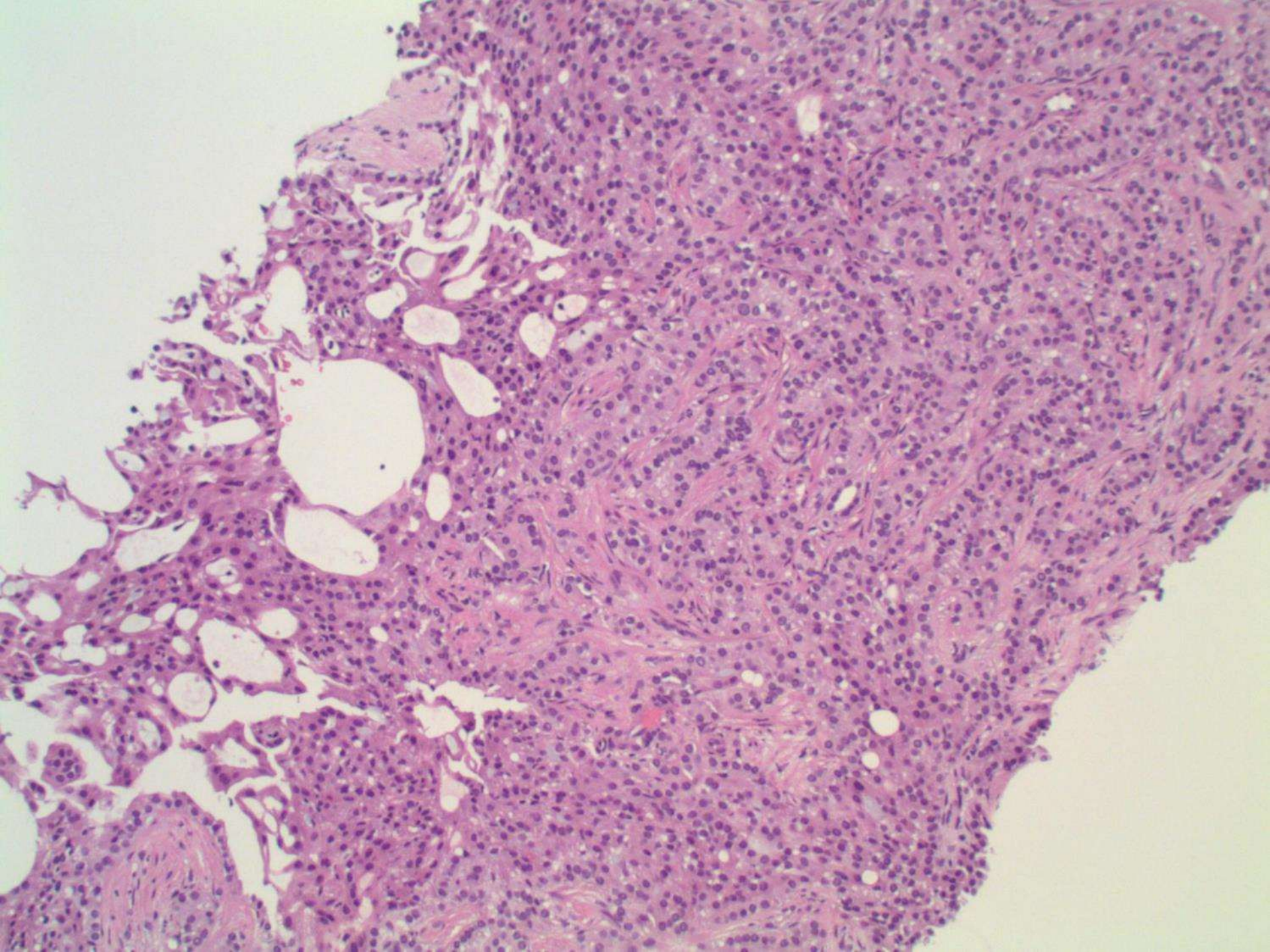


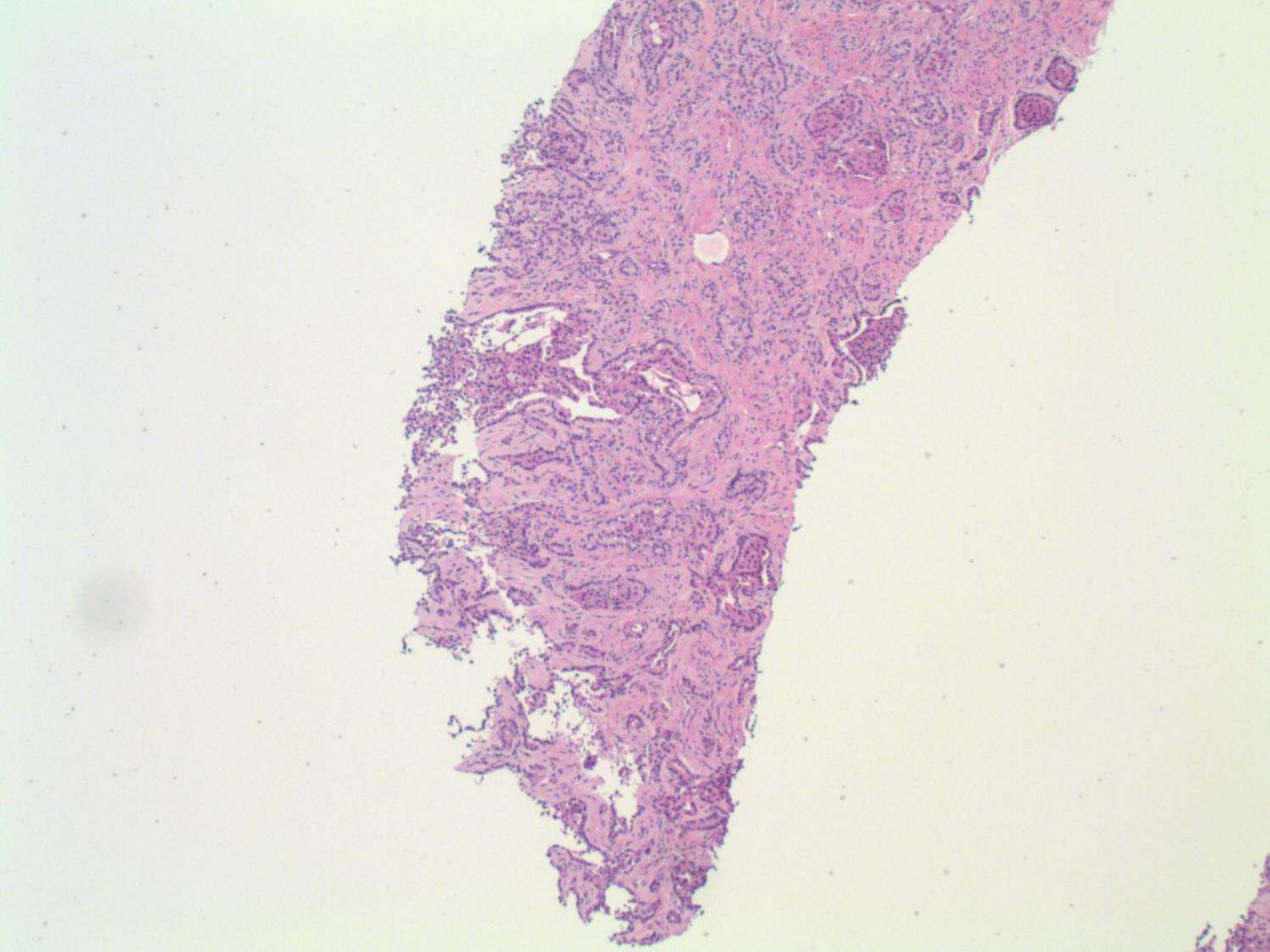


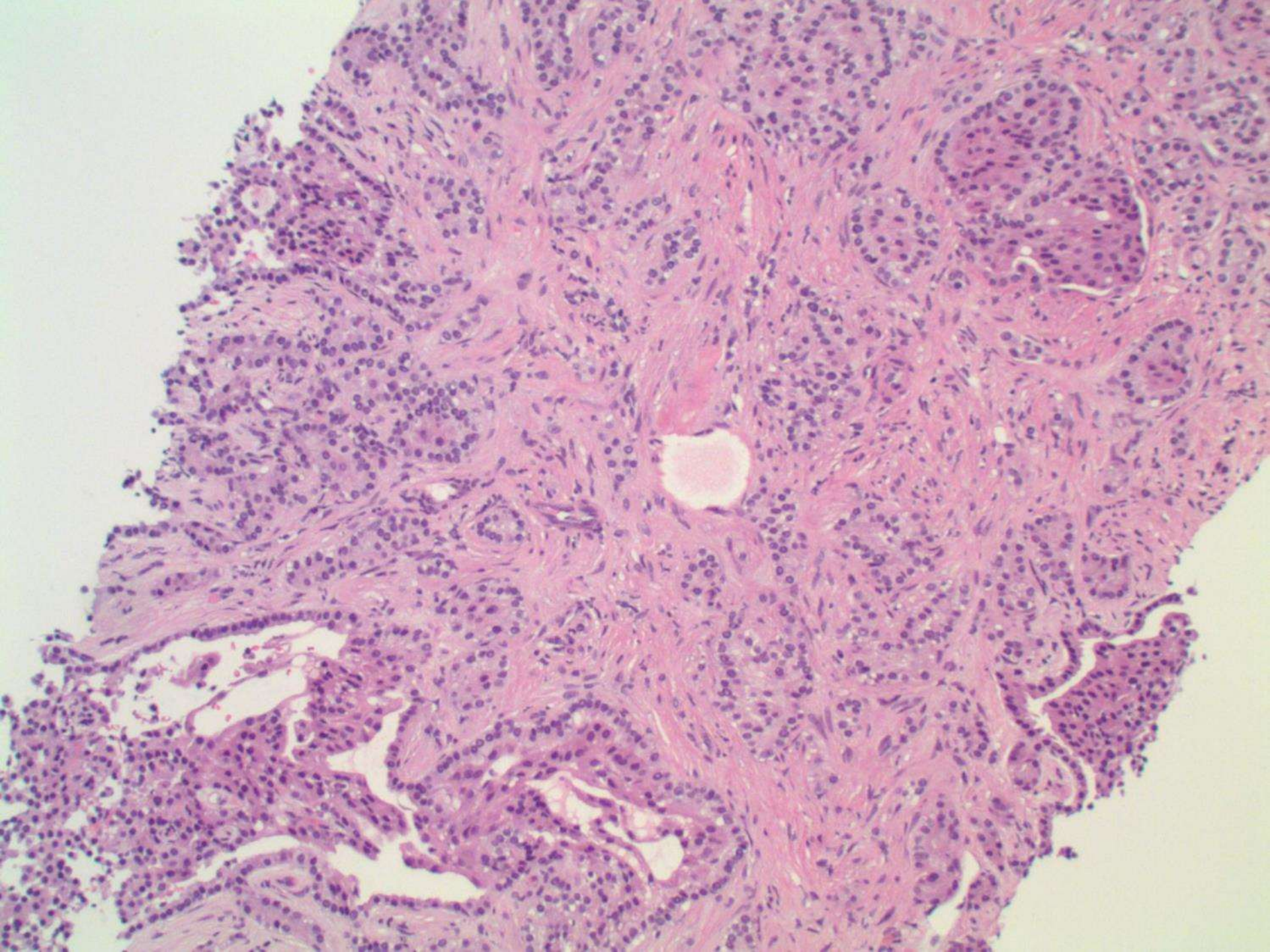


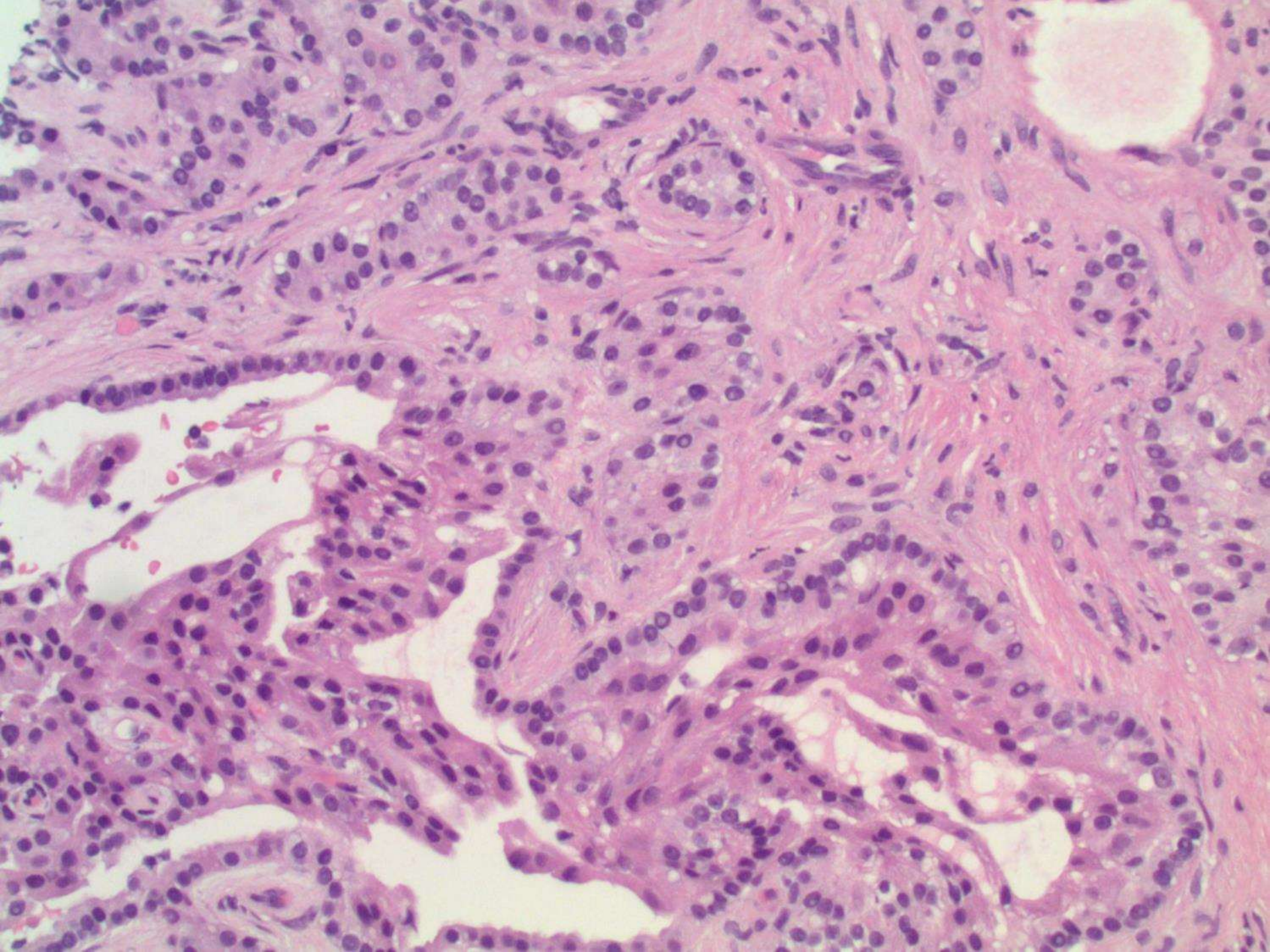












DDx

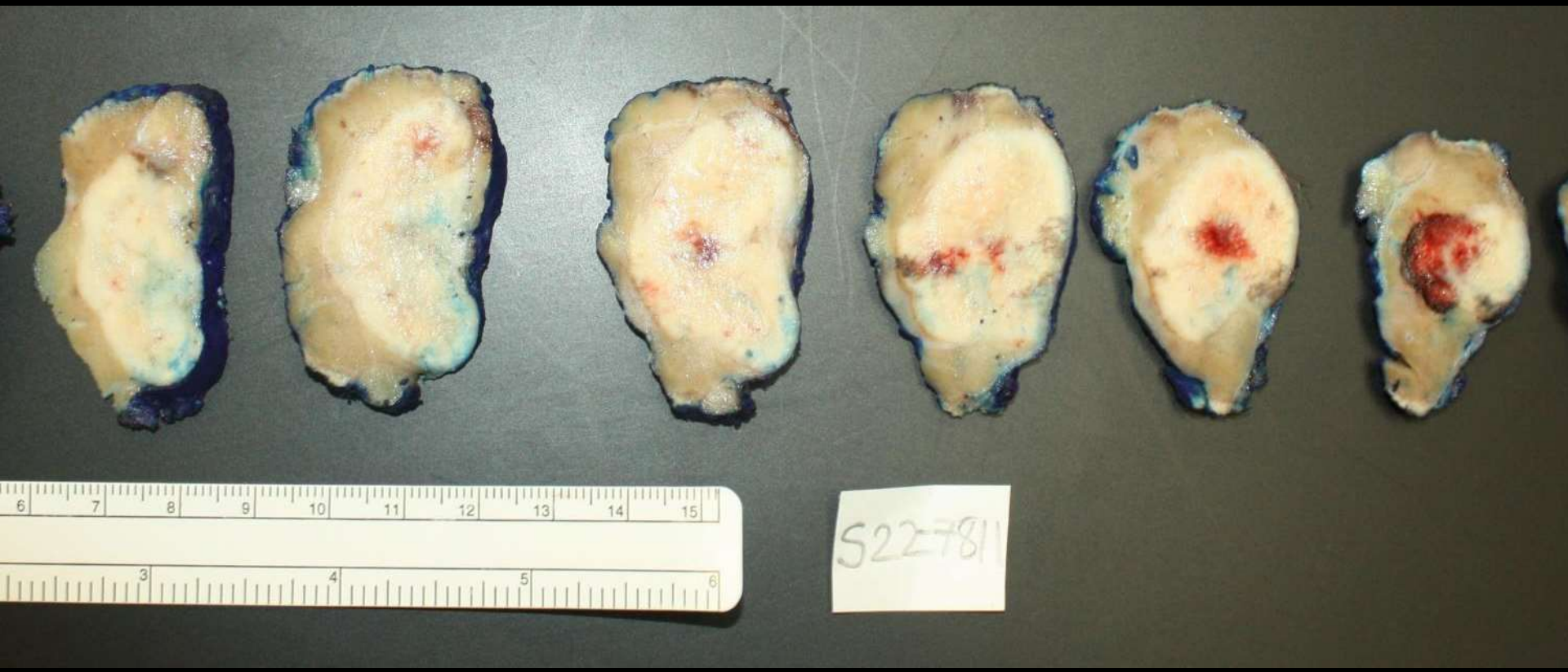
- Papillary RCC
- ACD-RCC
- MiTF RCC
- Tubulocystic RCC
- FH-def RCC
- Clear cell RCC
- Urothelial carcinoma
- metastasis

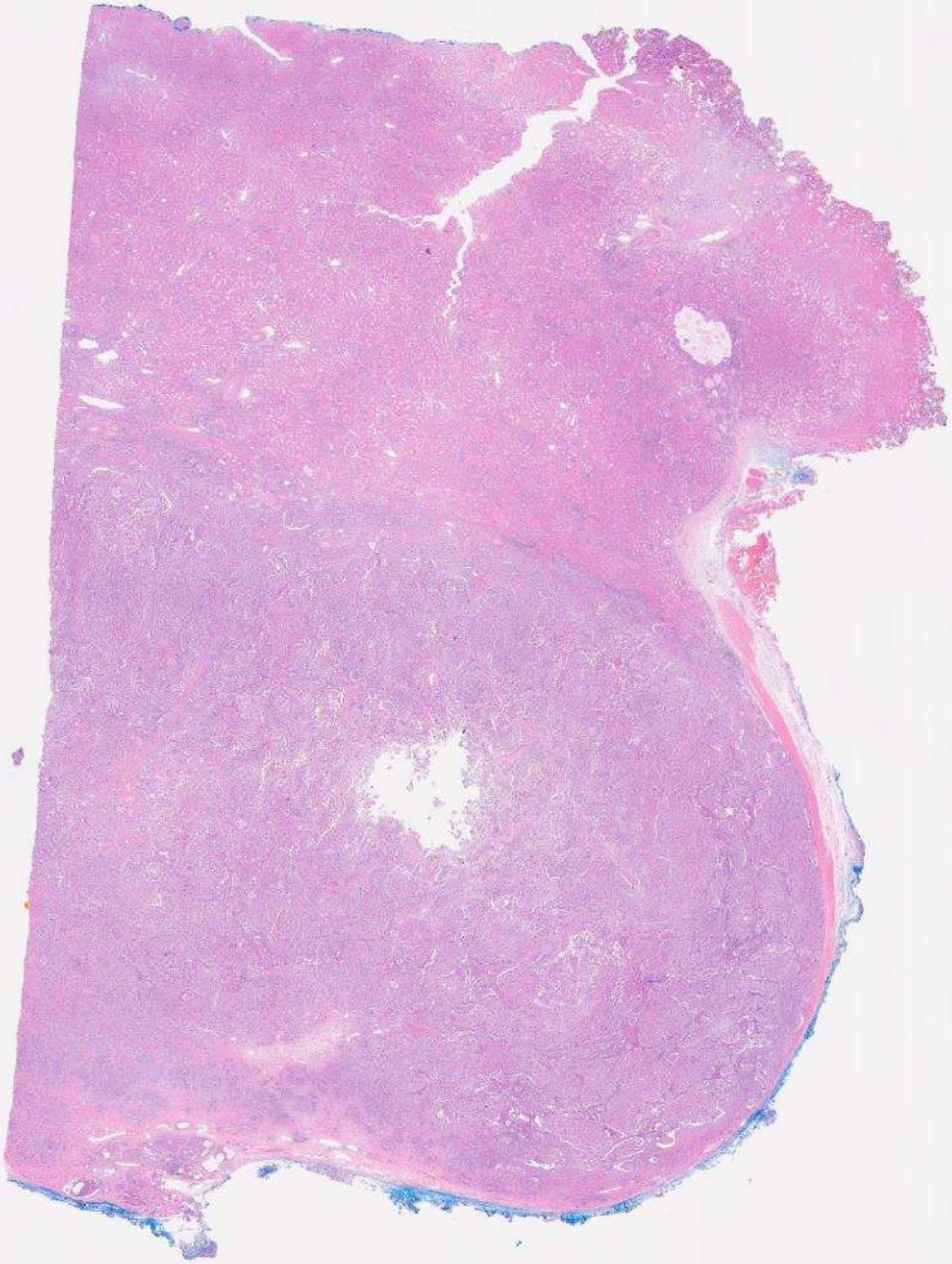
IHC summary

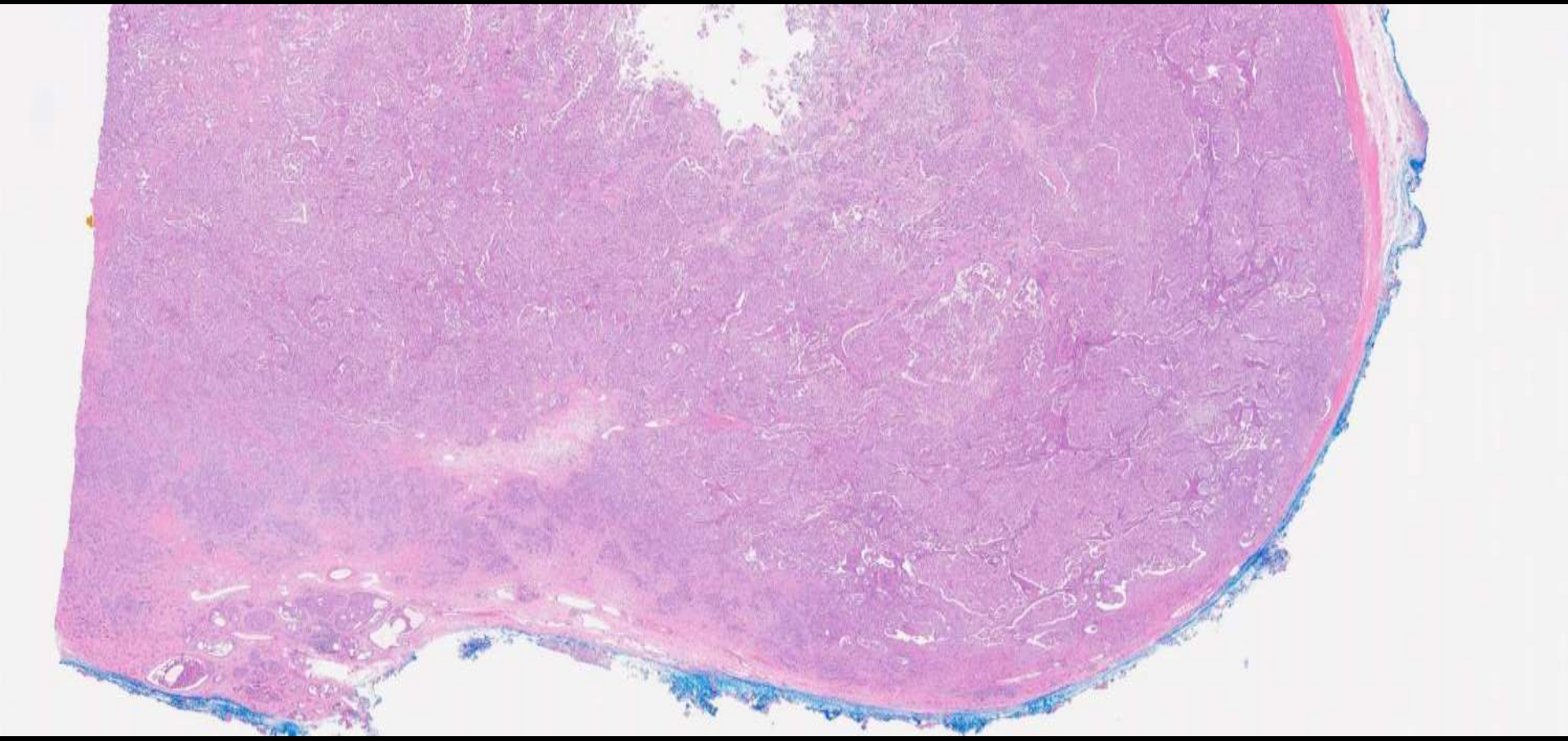
- PAX8+
- CK7+
- AMACR+
- Vimentin+
- CA9 neg
- CD117 neg
- GATA3 neg

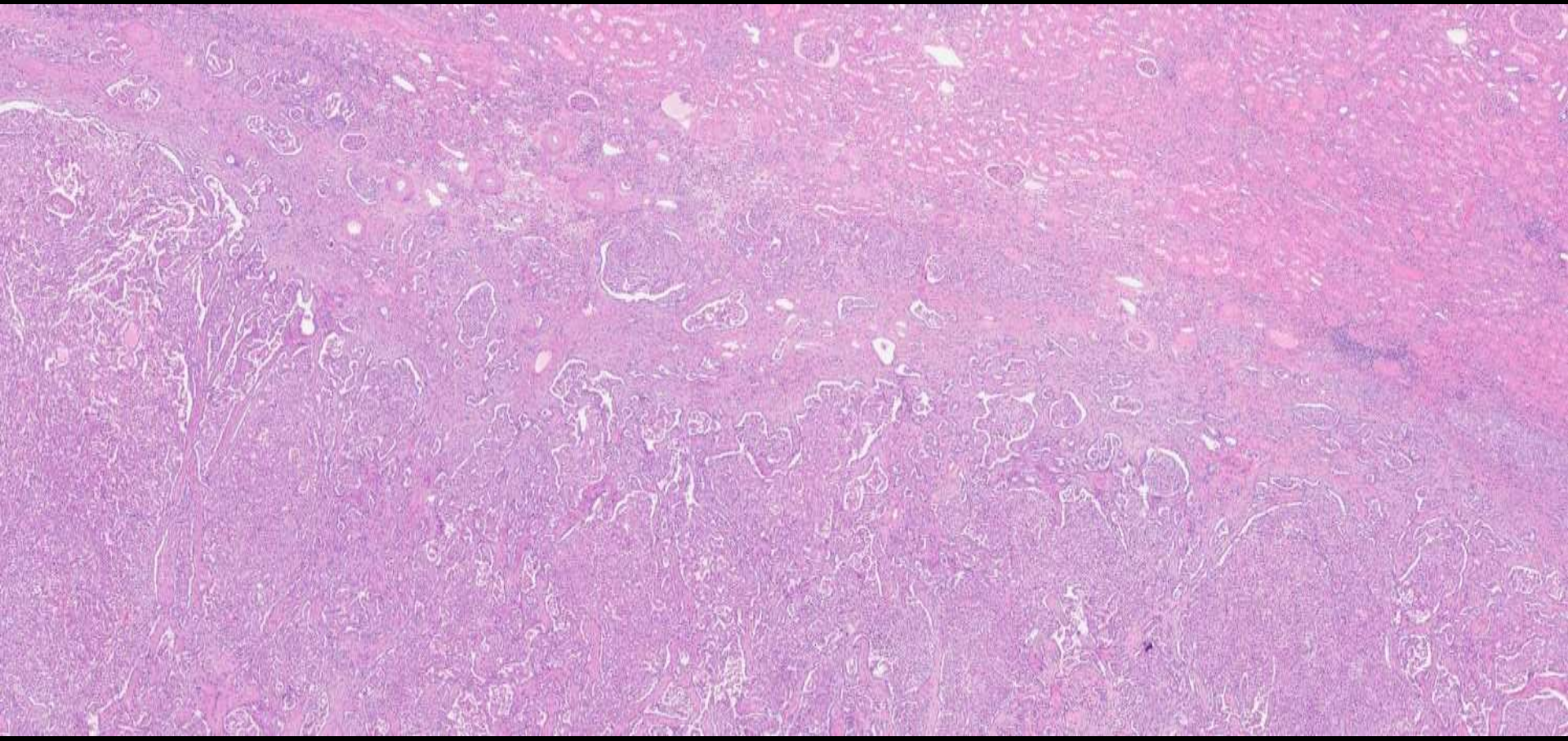
DDx

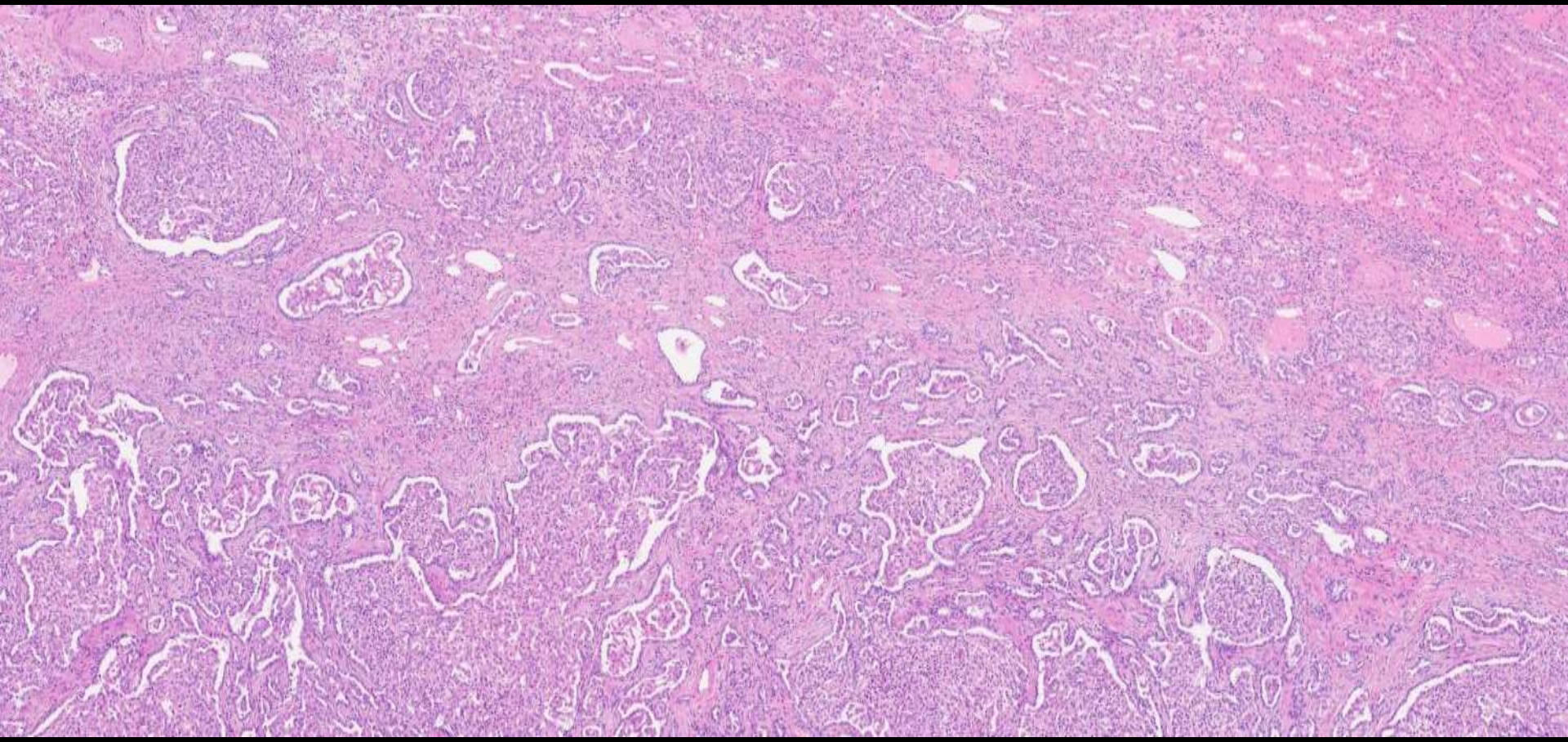
- **Papillary RCC**
- **ACD-RCC**
- **MiTF RCC**
- **Tubulocystic RCC**
- **FH-def RCC**
- **Clear cell RCC**
- **Urothelial carcinoma**
- **metastasis**

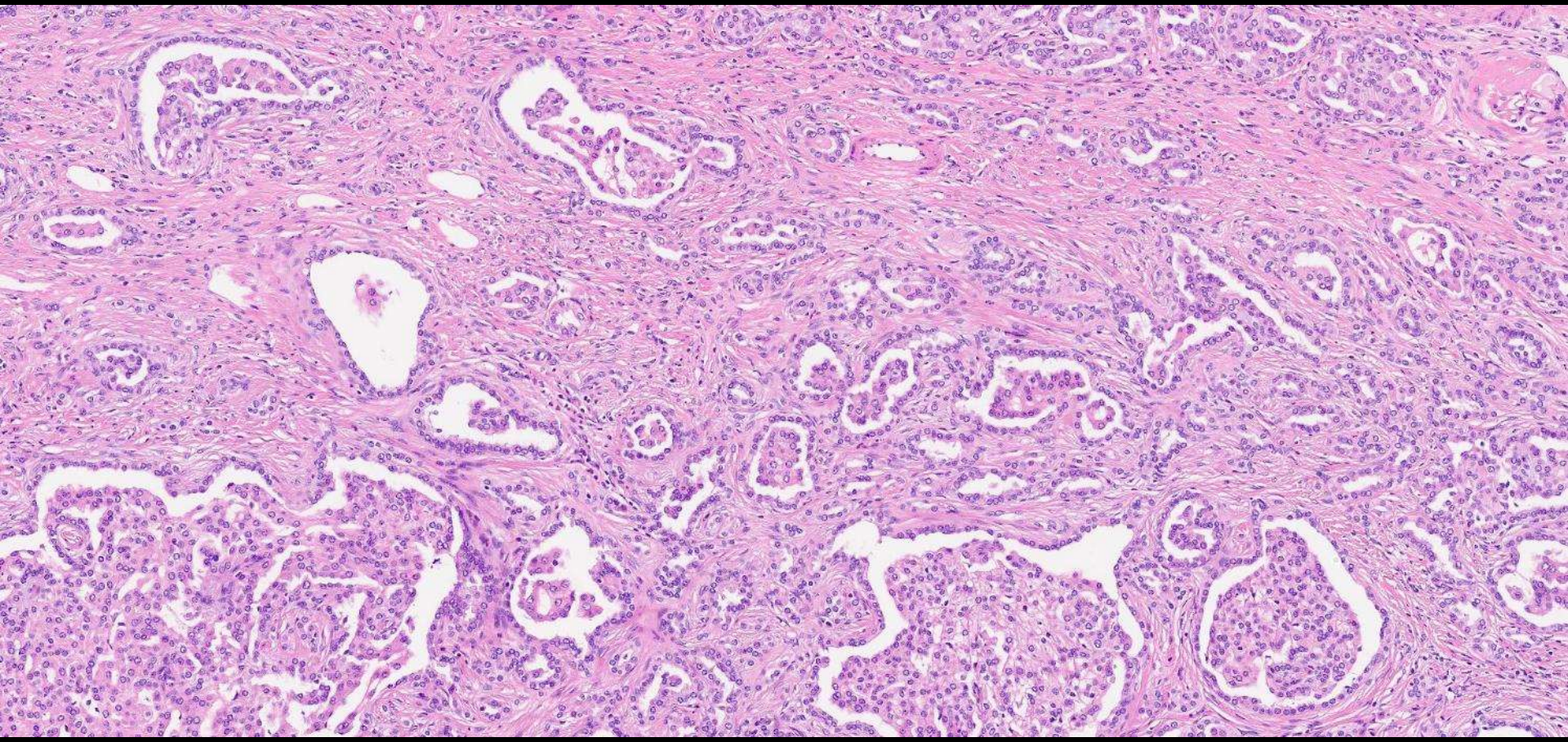

S22-7811

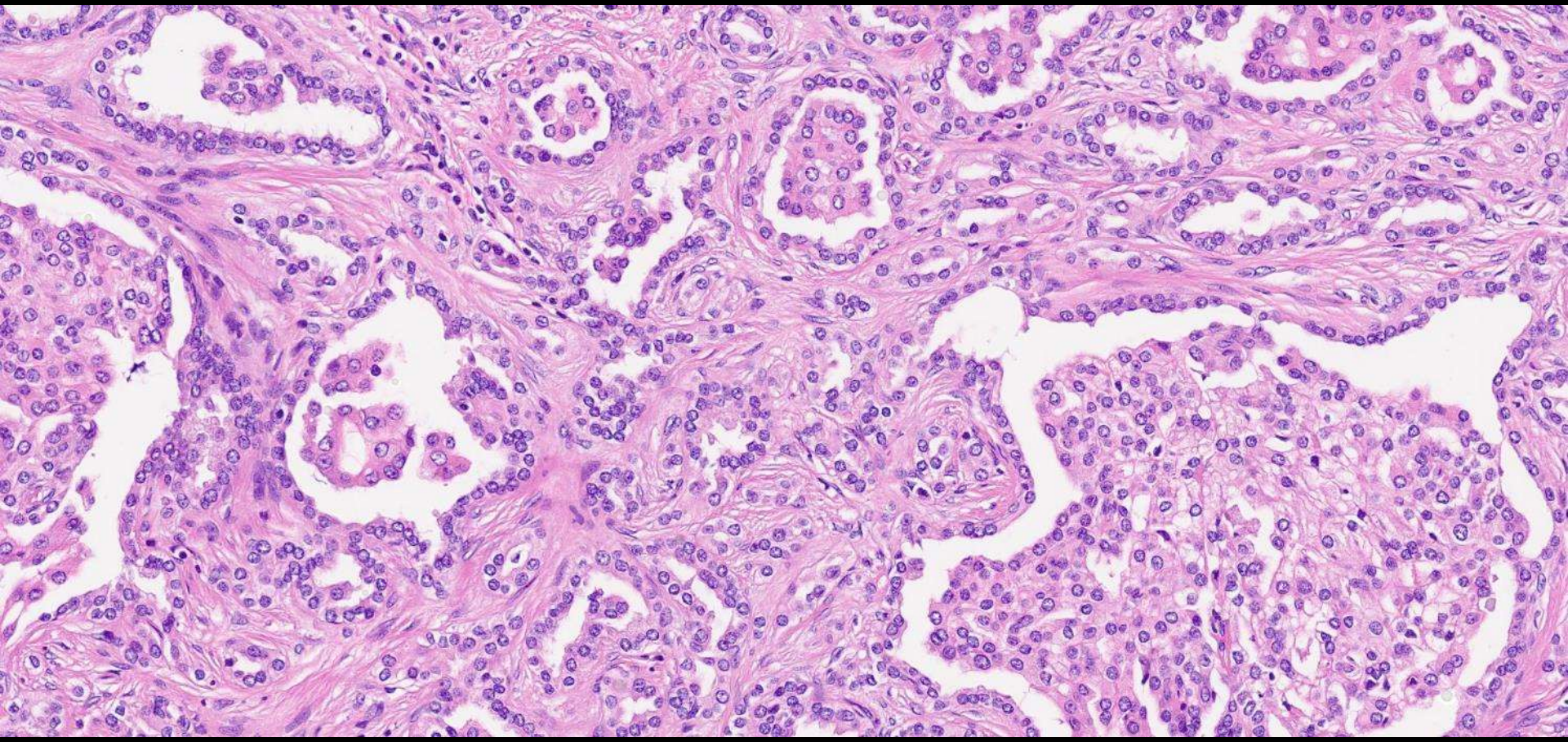


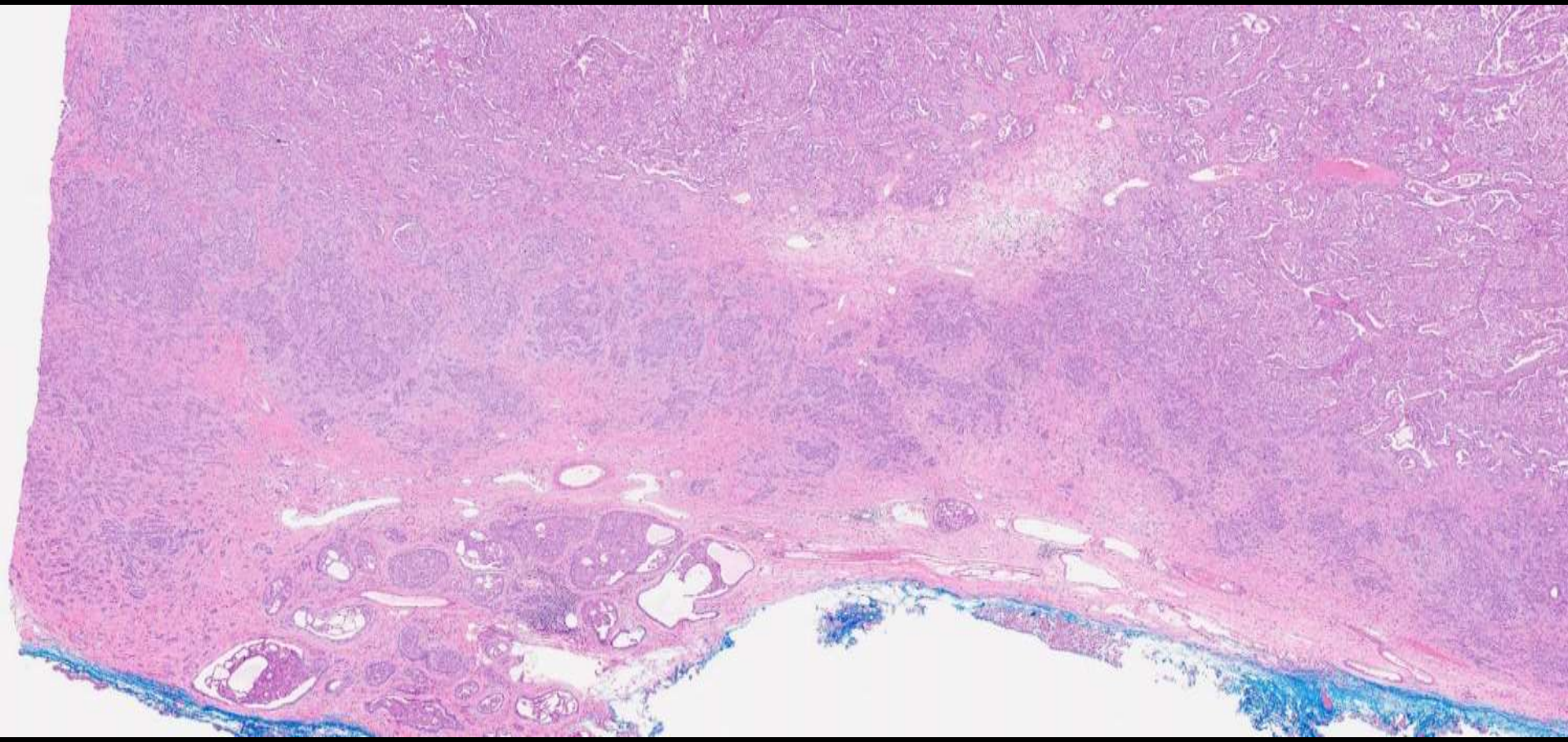


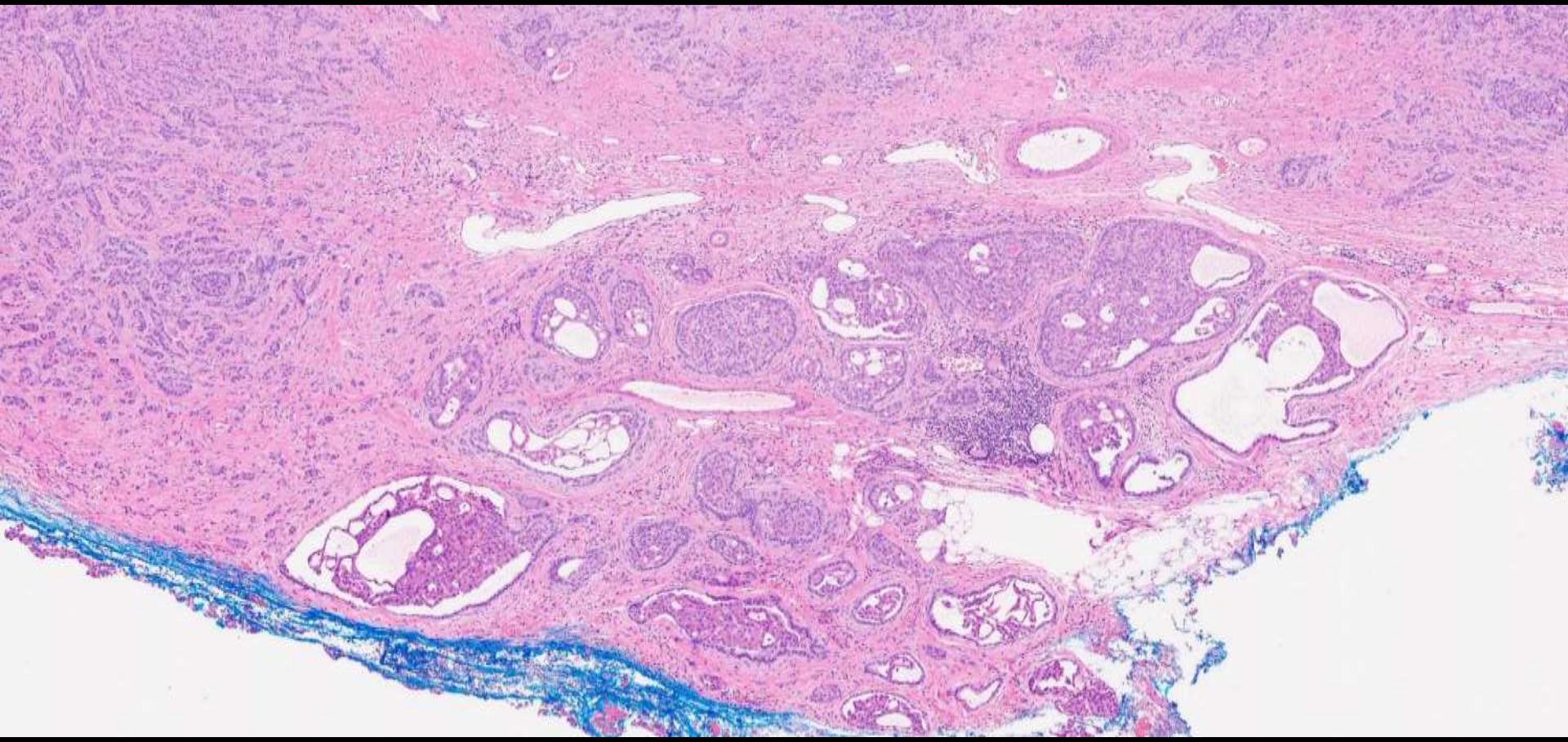


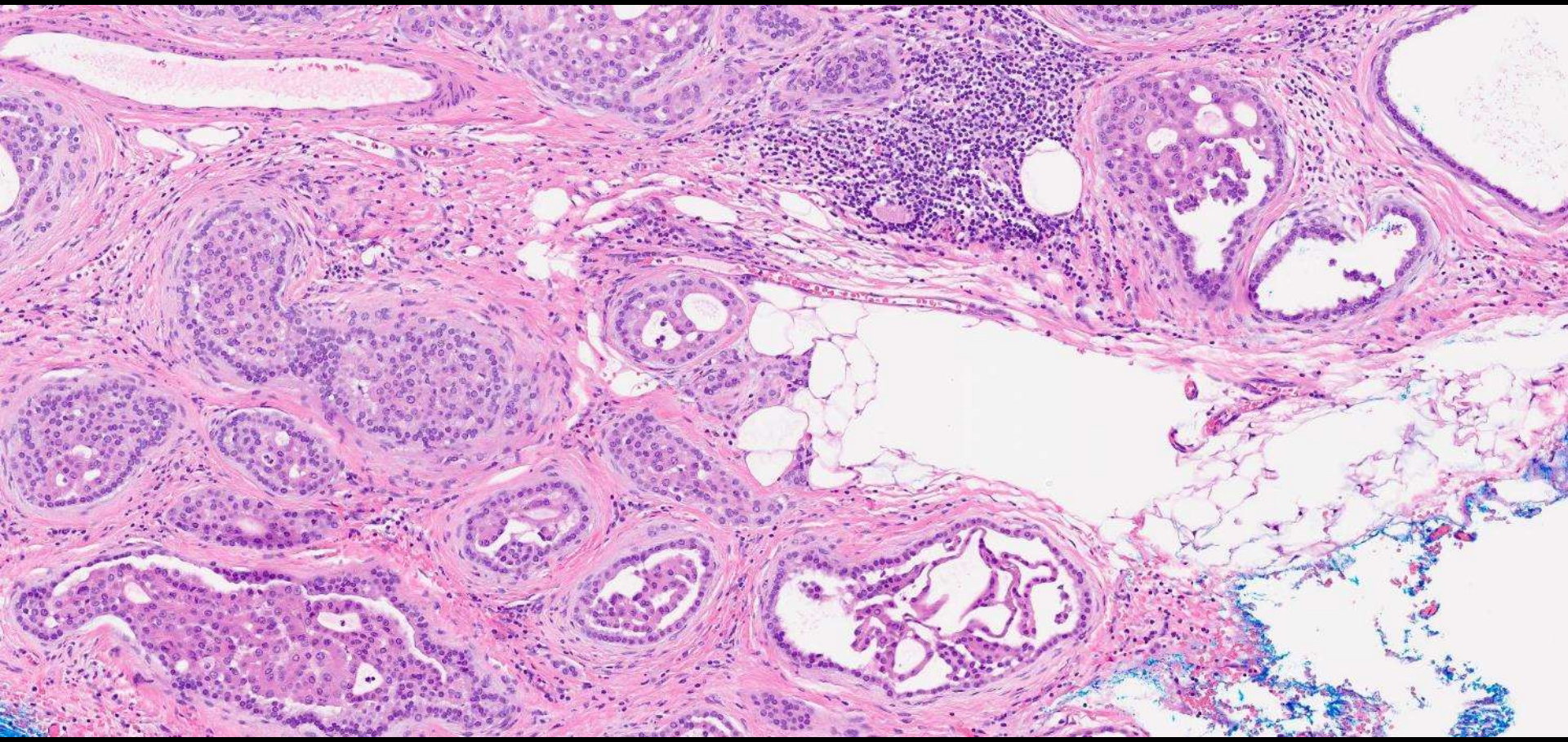


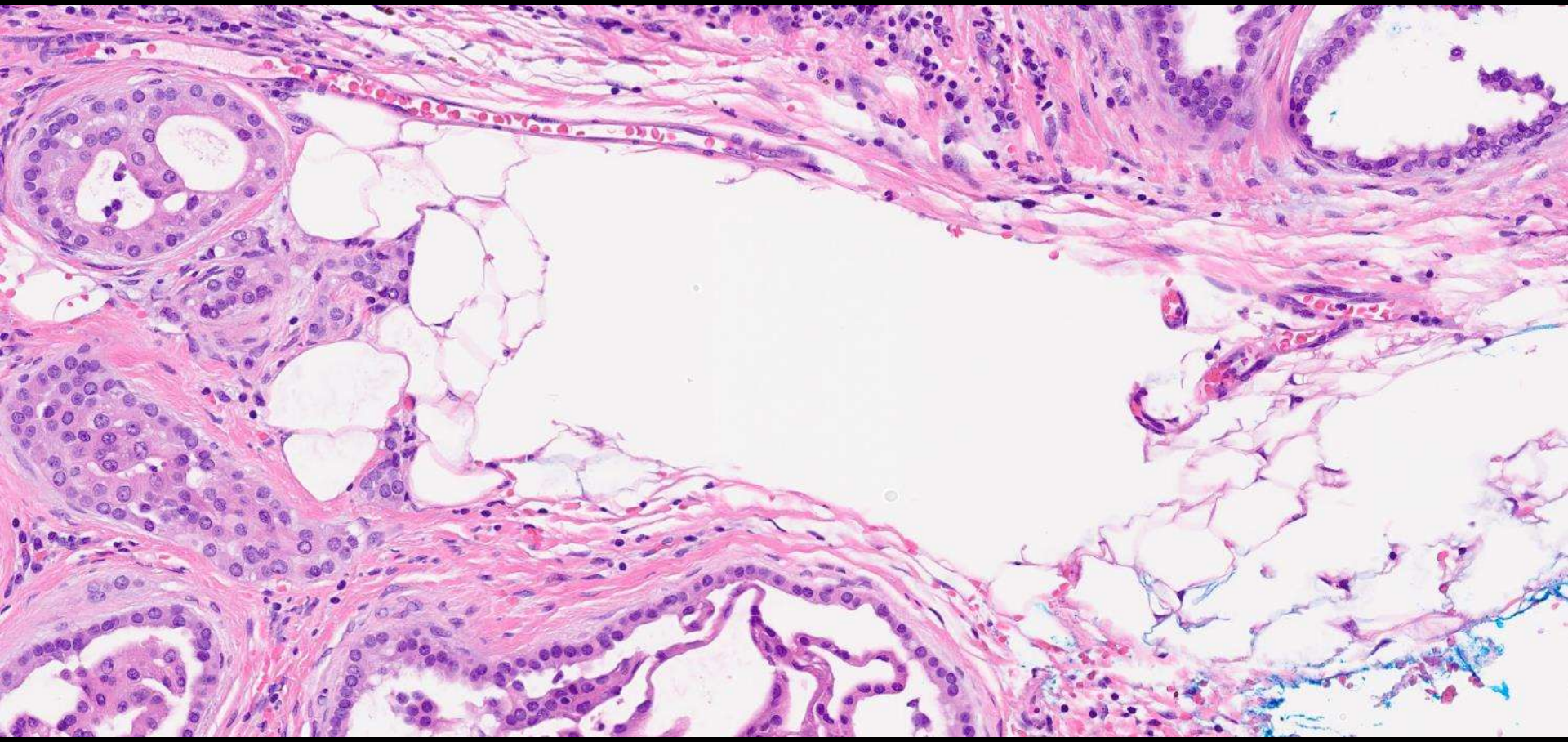








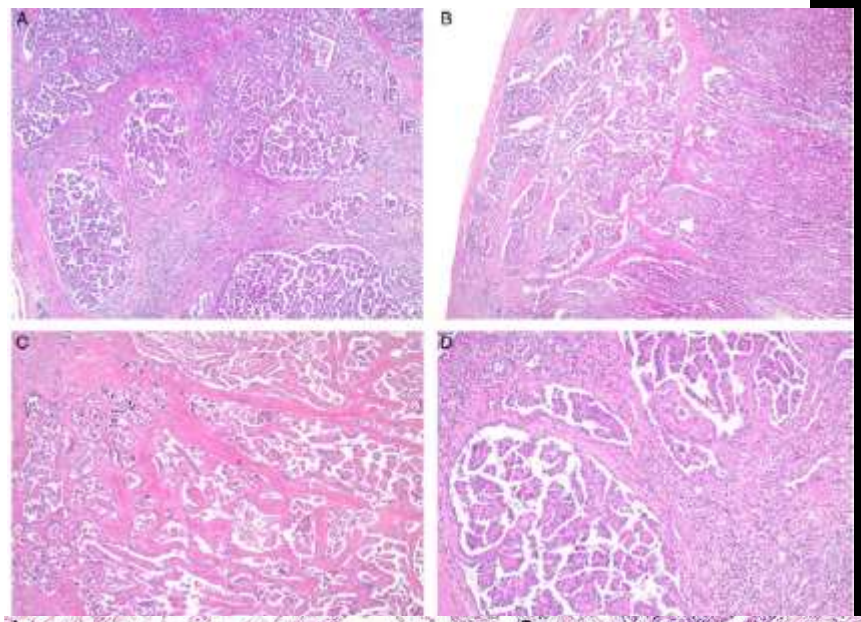




Papillary Renal Cell Carcinoma With Microcystic Architecture Is Strongly Associated With Extrarenal Invasion and Metastatic Disease

Emily Chan, MD, Bradley A. Stohr, MD, PhD,* Robert S. Butler, MS,† Roni M. Cox, MD,‡ Jonathan L. Myles, MD,‡ Jane K. Nguyen, MD, PhD,‡ Christopher G. Przybycin, MD,‡ Jordan P. Reynolds, MD,‡ Sean R. Williamson, MD,‡ and Jesse K. McKenney, MD‡*

Abstract: Papillary renal cell carcinoma (PRCC) is well-recognized as a morphologically and molecularly heterogenous group of kidney tumors with variable clinical behavior. Our goal was to analyze a unique histologic pattern of PRCC we have observed in routine practice to evaluate for potential clinical significance or distinct molecular signature. We identified 42 cases of PRCC showing a morphologically distinct architecture characterized by numerous epithelial-lined cysts containing the papillary tumor (herein called "microcysts"), which are typically separated by fibrous stroma. Of the initial 42 case test set with microcystic features, 23 (55%) were stage pT3a or higher. Most tumors had strong and diffuse cytoplasmic immunoreactivity for CK7 (93%, 37/40) and AMACR (100%, 40/40). Fumarate hydratase staining was retained in all cases tested (39/39). We performed next-generation sequencing on 15 of these cases with available tissue and identified chromosomal alterations commonly reported in historically "type 1" PRCC, notably multiple chromosomal gains, particularly of chromosomes 7 and 17, and *MET* alterations. However, alterations in pathways associated with more aggressive behavior (including SETD2, CDKN2A, and members of the NRF pathway) were also identified in 6 of 15 cases tested (40%). Given this molecular and immunophenotypic data, we subsequently reviewed an additional group of 60 consecutive pT2b-pT3 PRCCs to allow for comparisons between cases with and without microcysts, to assess for potential associations with other recently described histologic patterns (ie, "unfavorable architecture": micropapillary, solid, and hobnail),



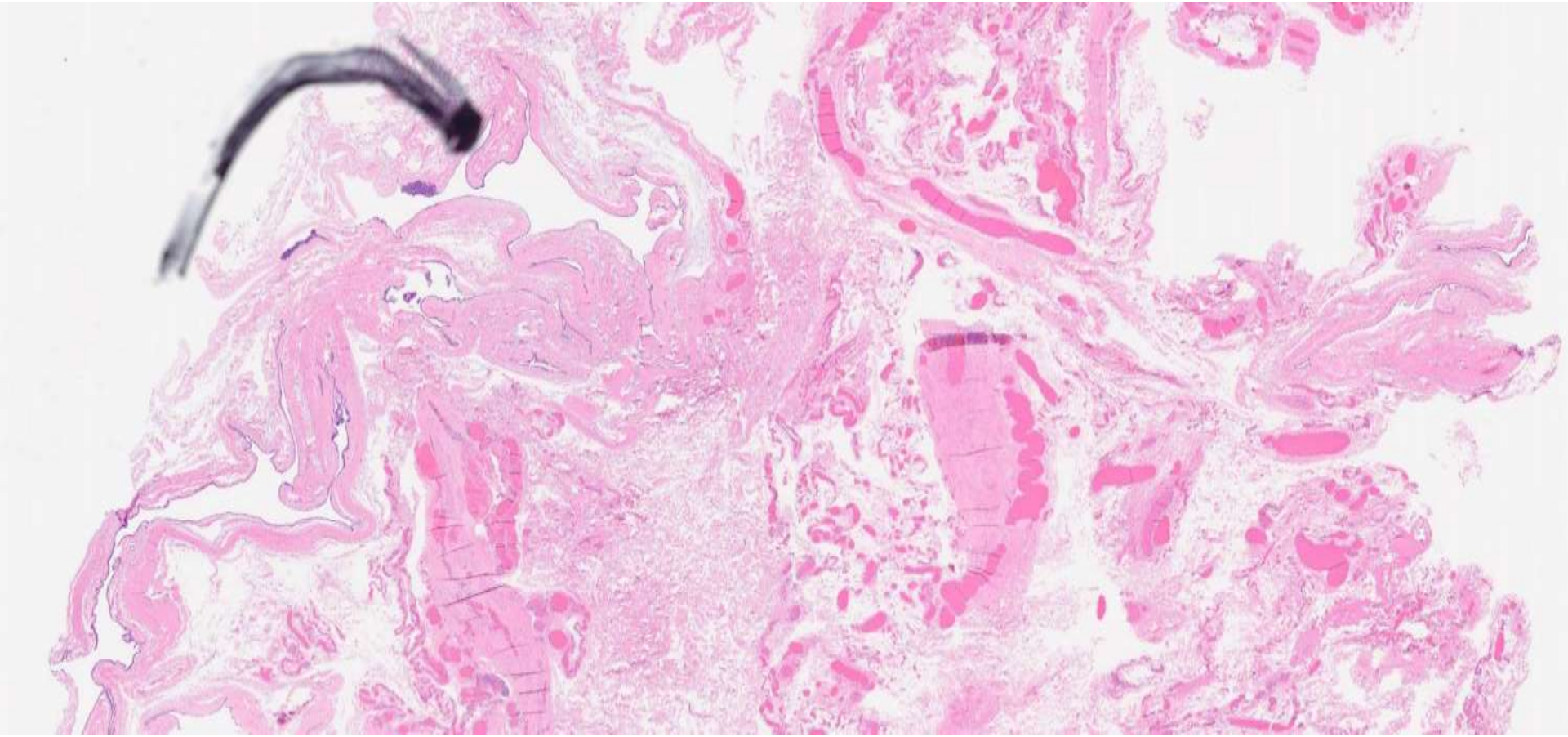
From the *Department of Pathology, University of California San Francisco (UCSF), San Francisco, CA; †Department of Quantitative Health Sciences, Cleveland Clinic; and ‡Department of Pathology,

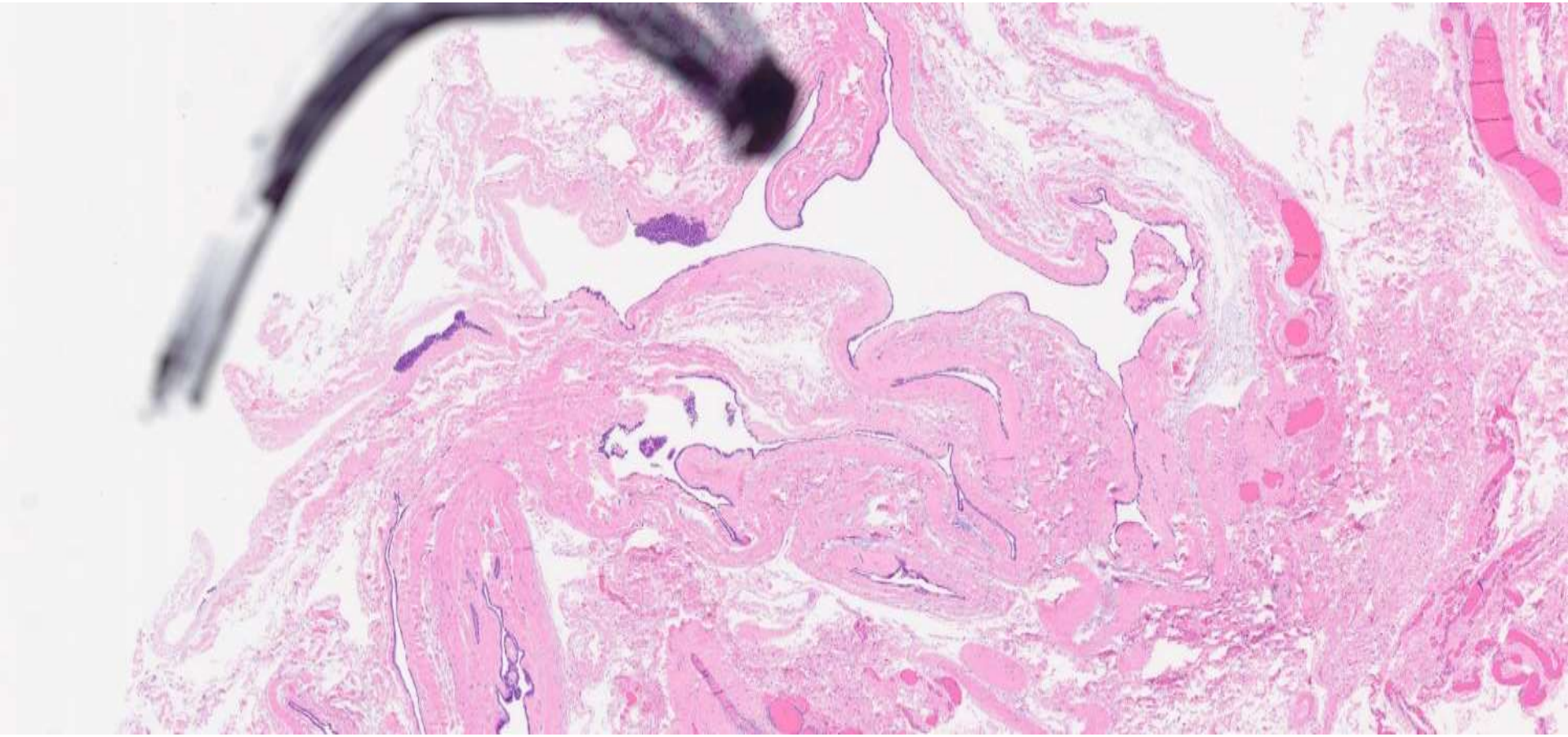
(*Am J Surg Pathol* 2022;46:392–403)

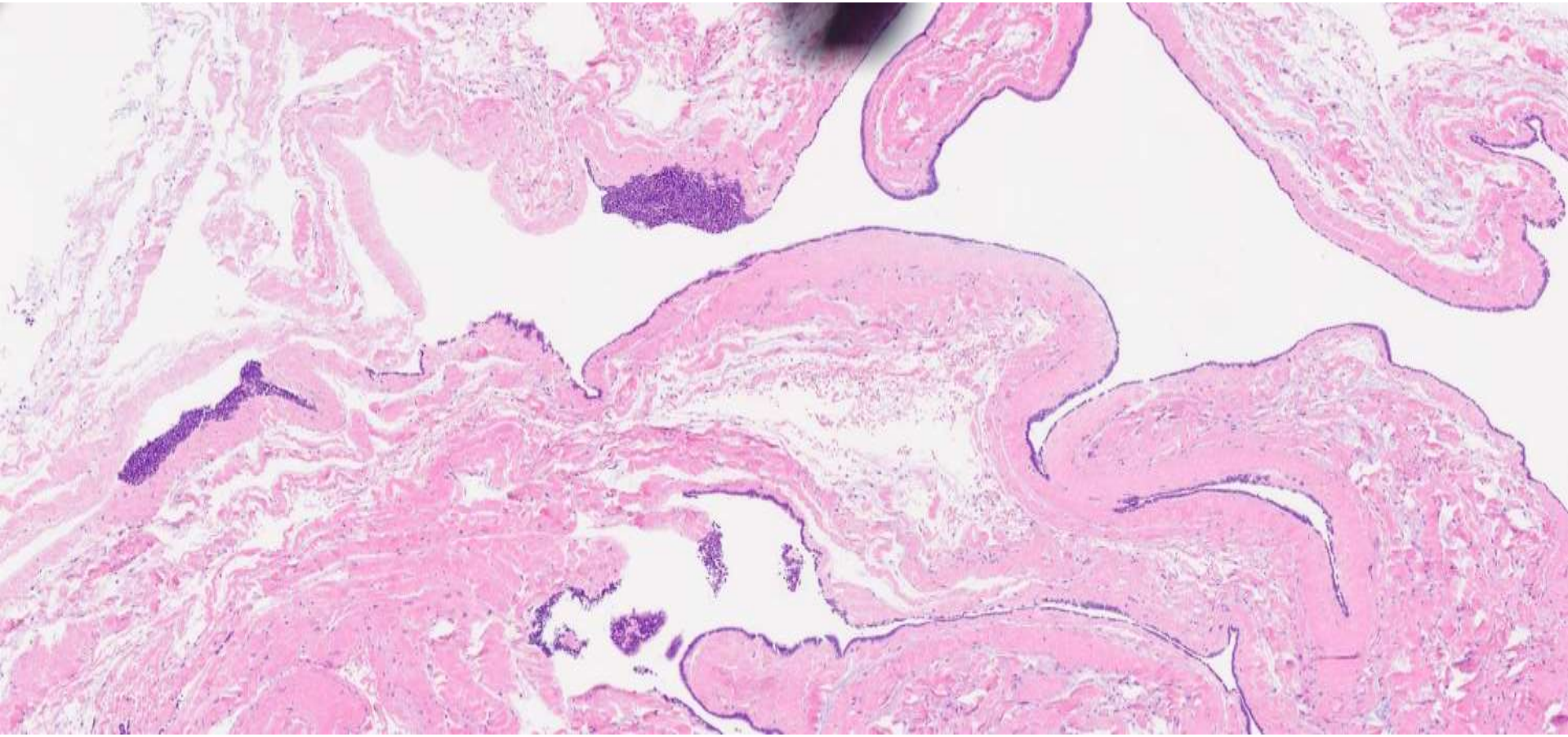
23-0208

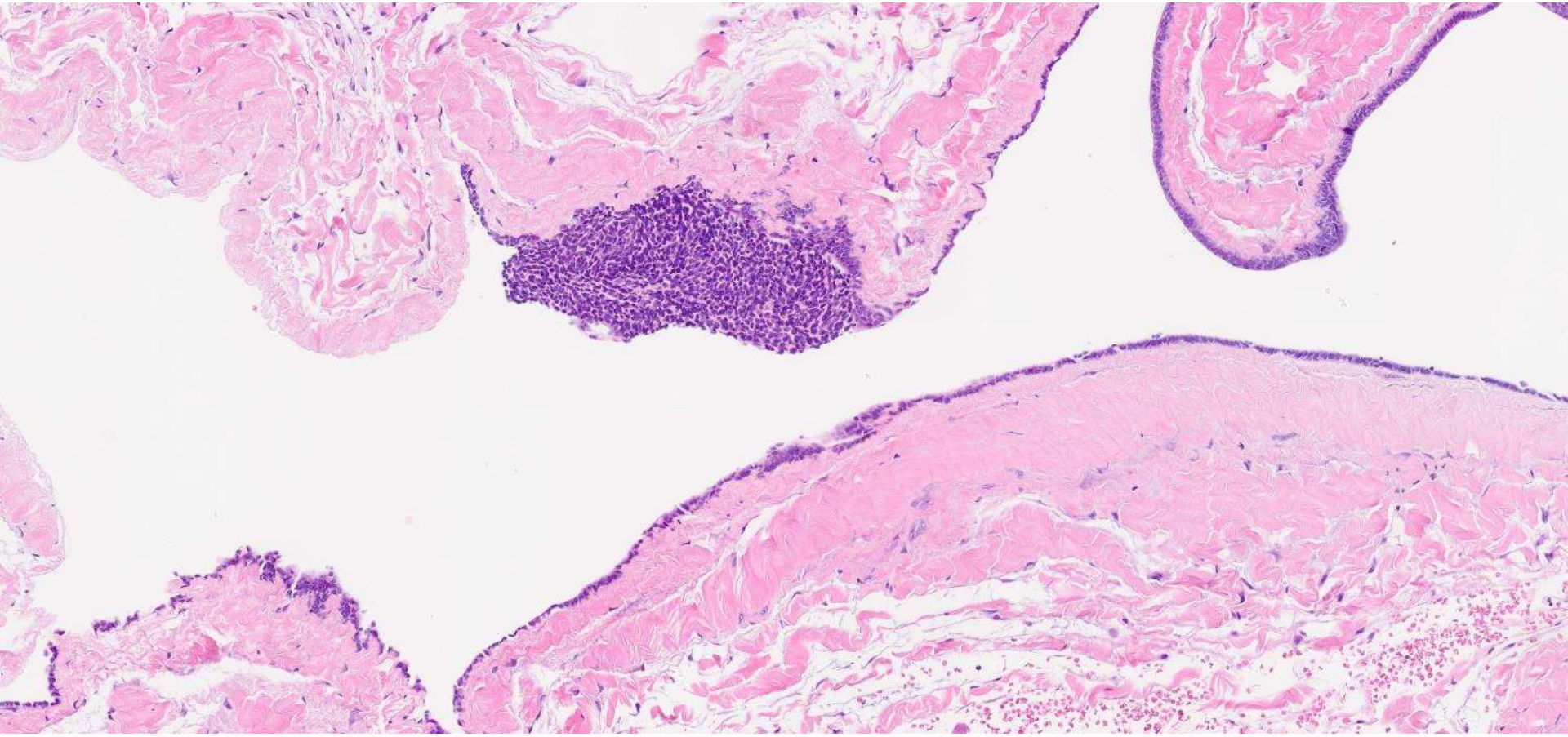
Ankur Sangoi; El Camino Hospital

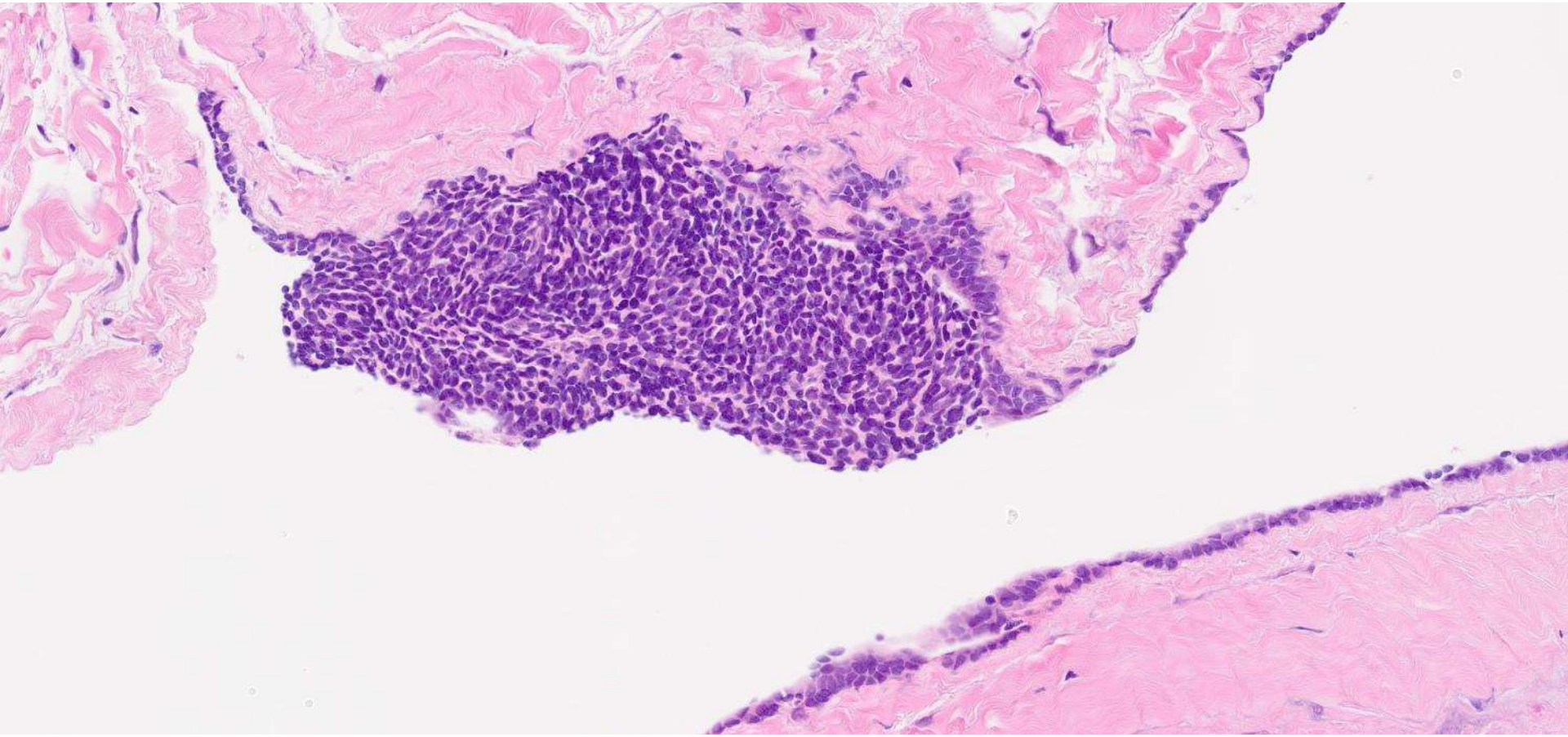
50ish M with h/o lung mass, now presents with spermatocele and undergoes spermatocelectomy.

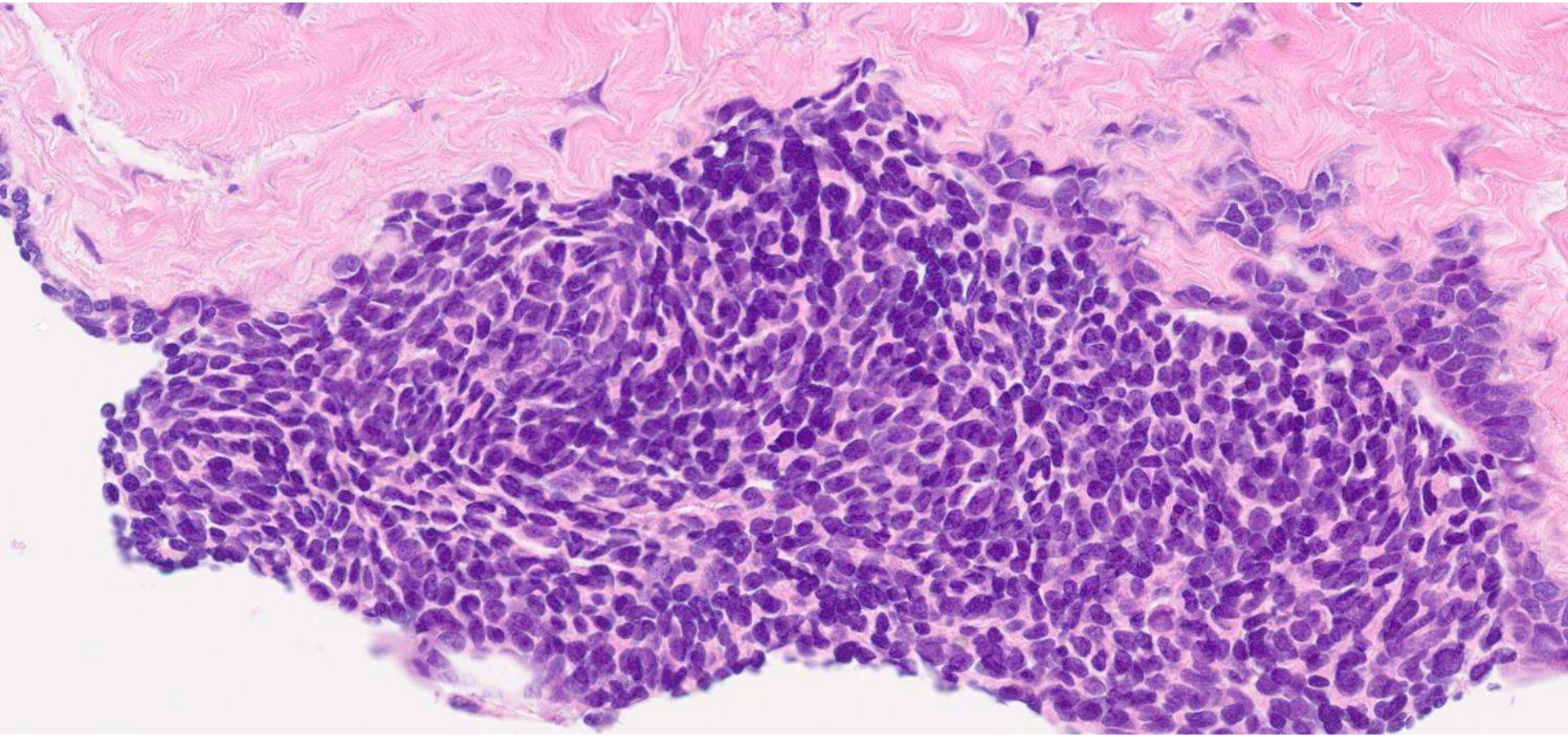


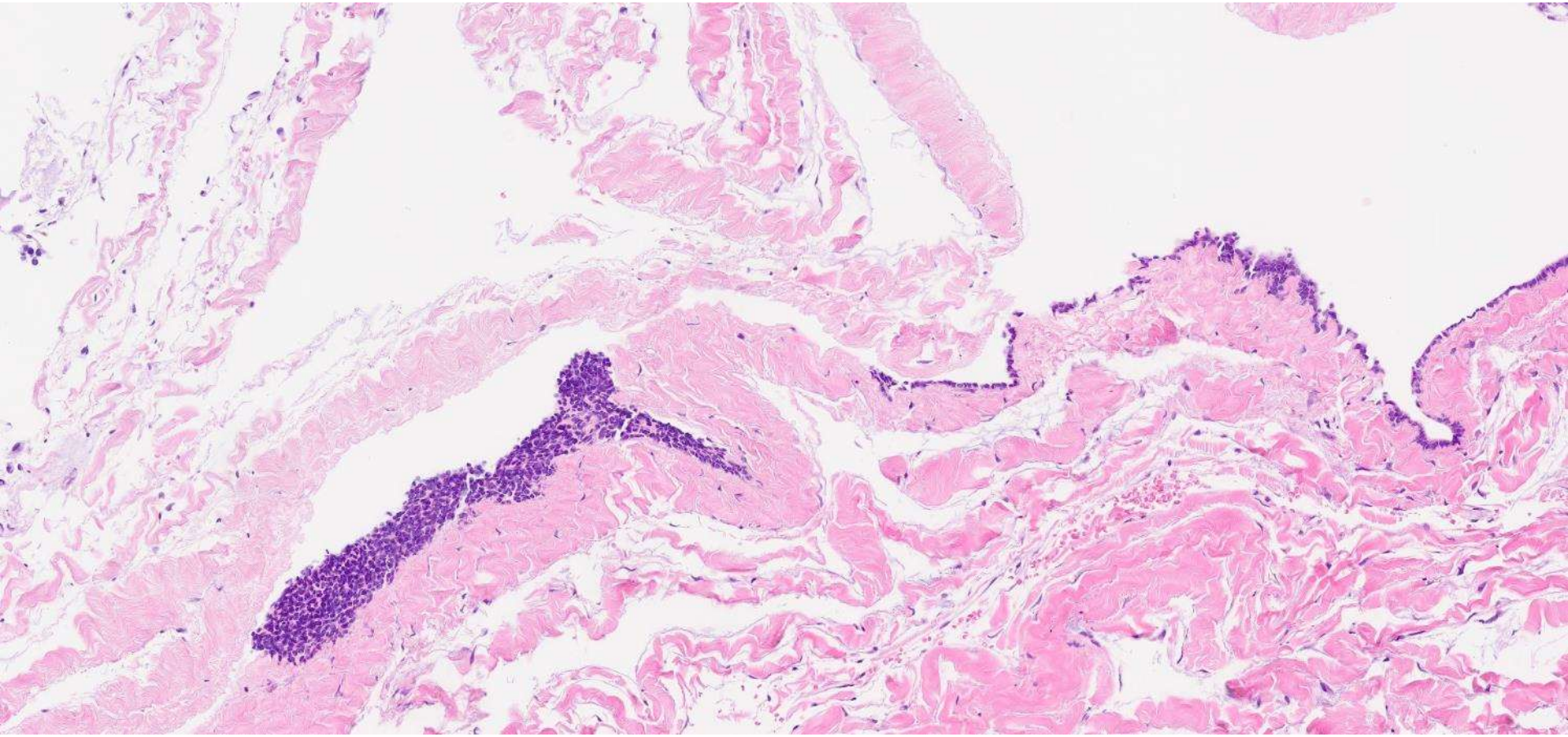


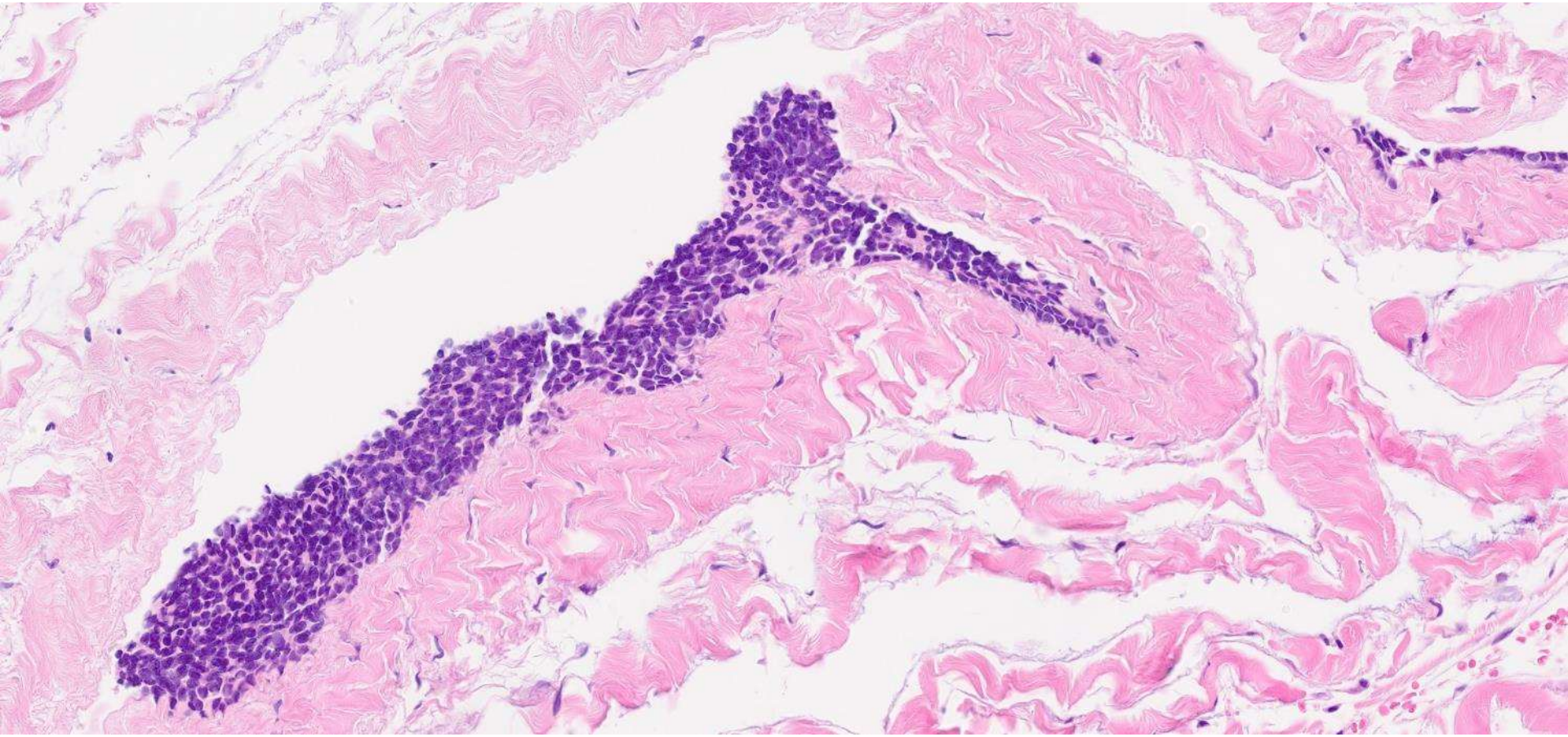


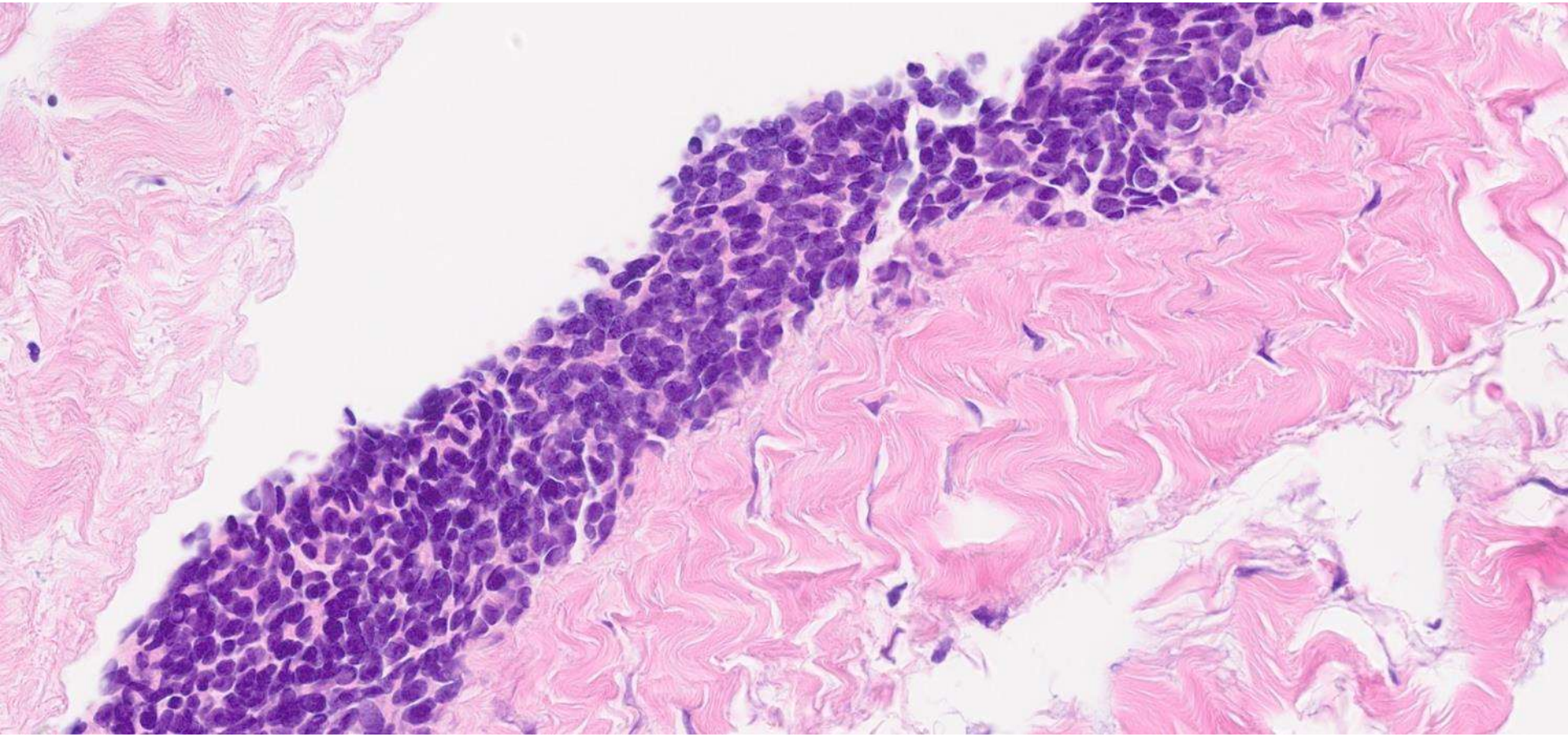


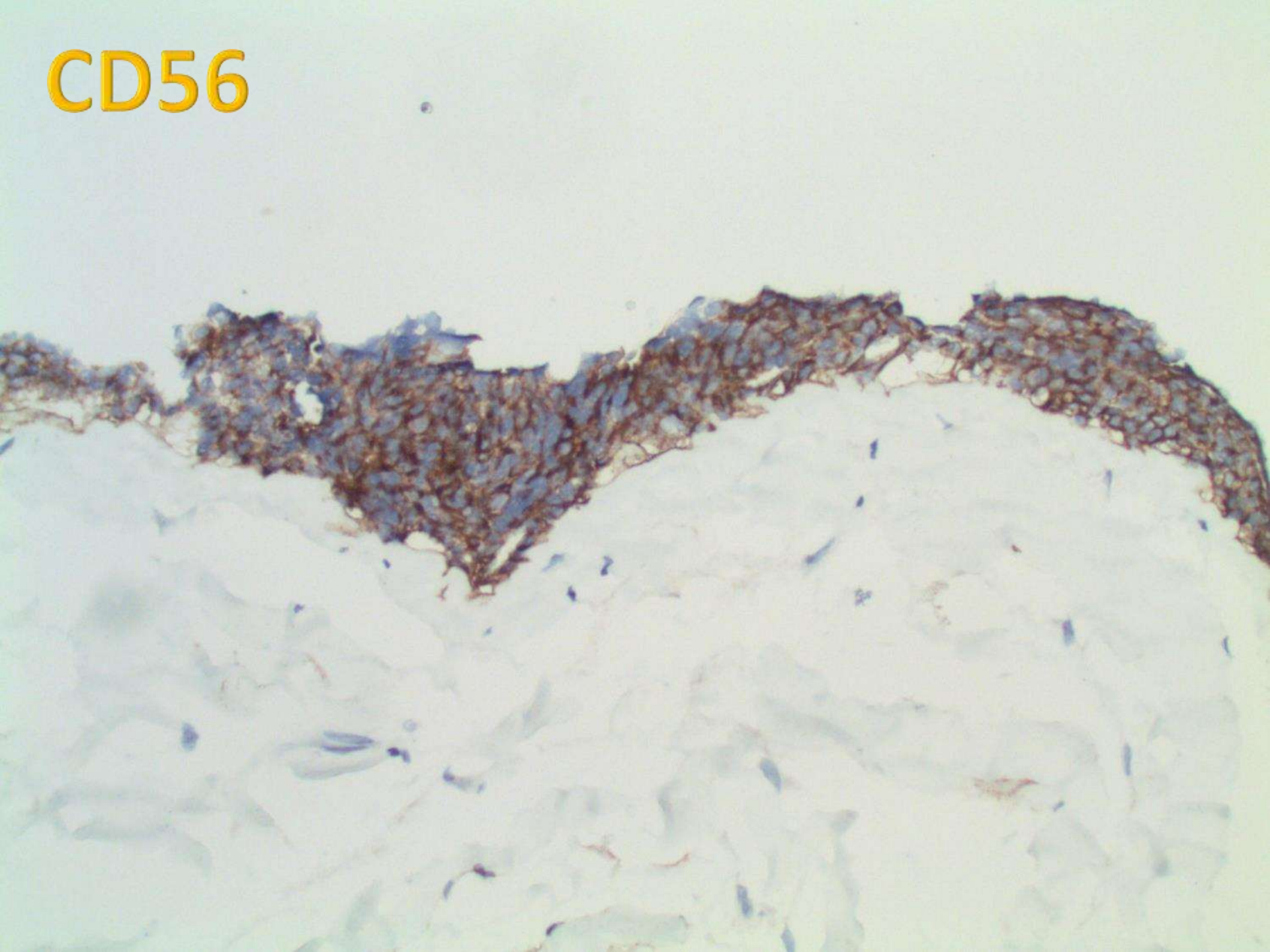




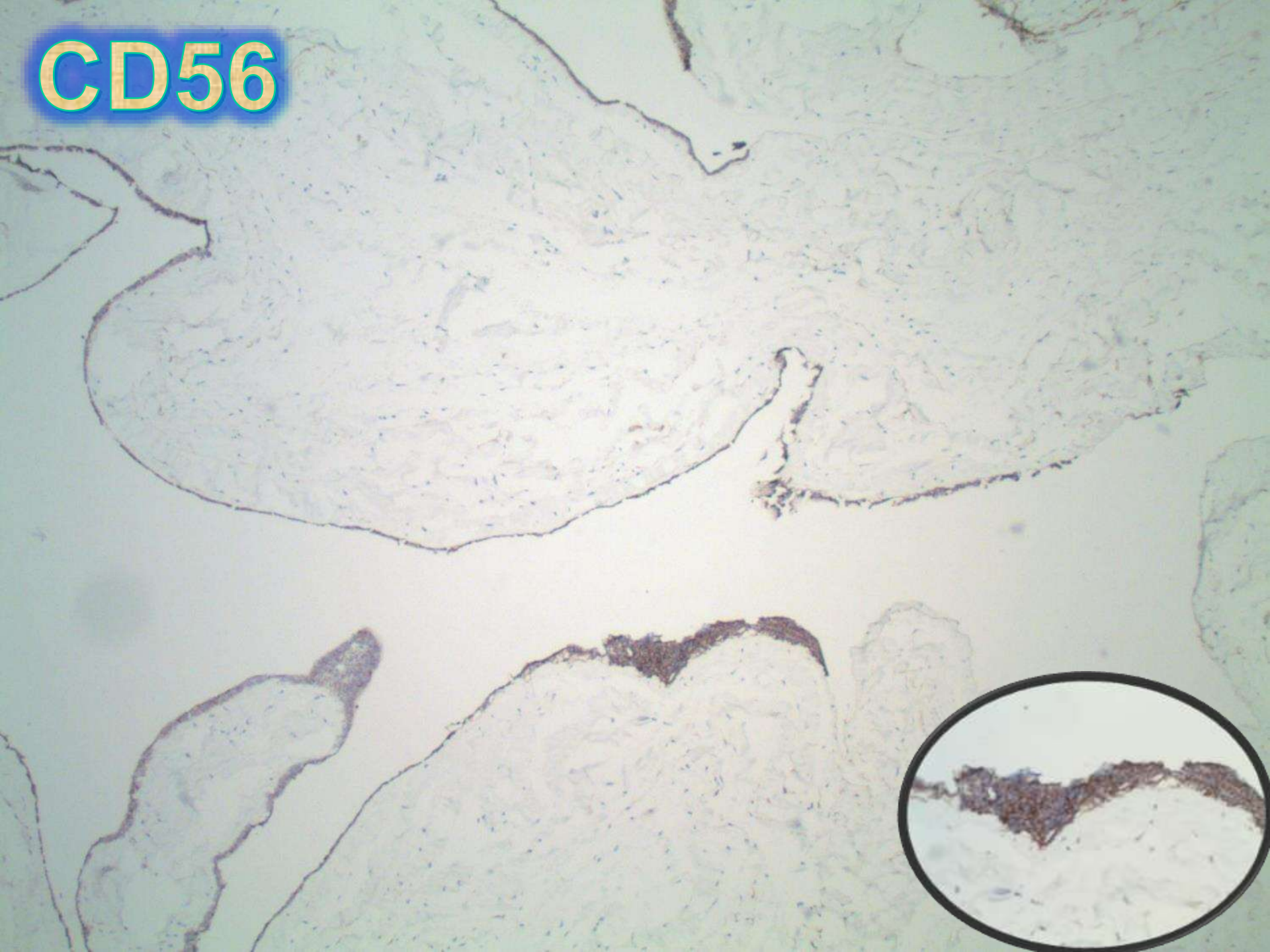


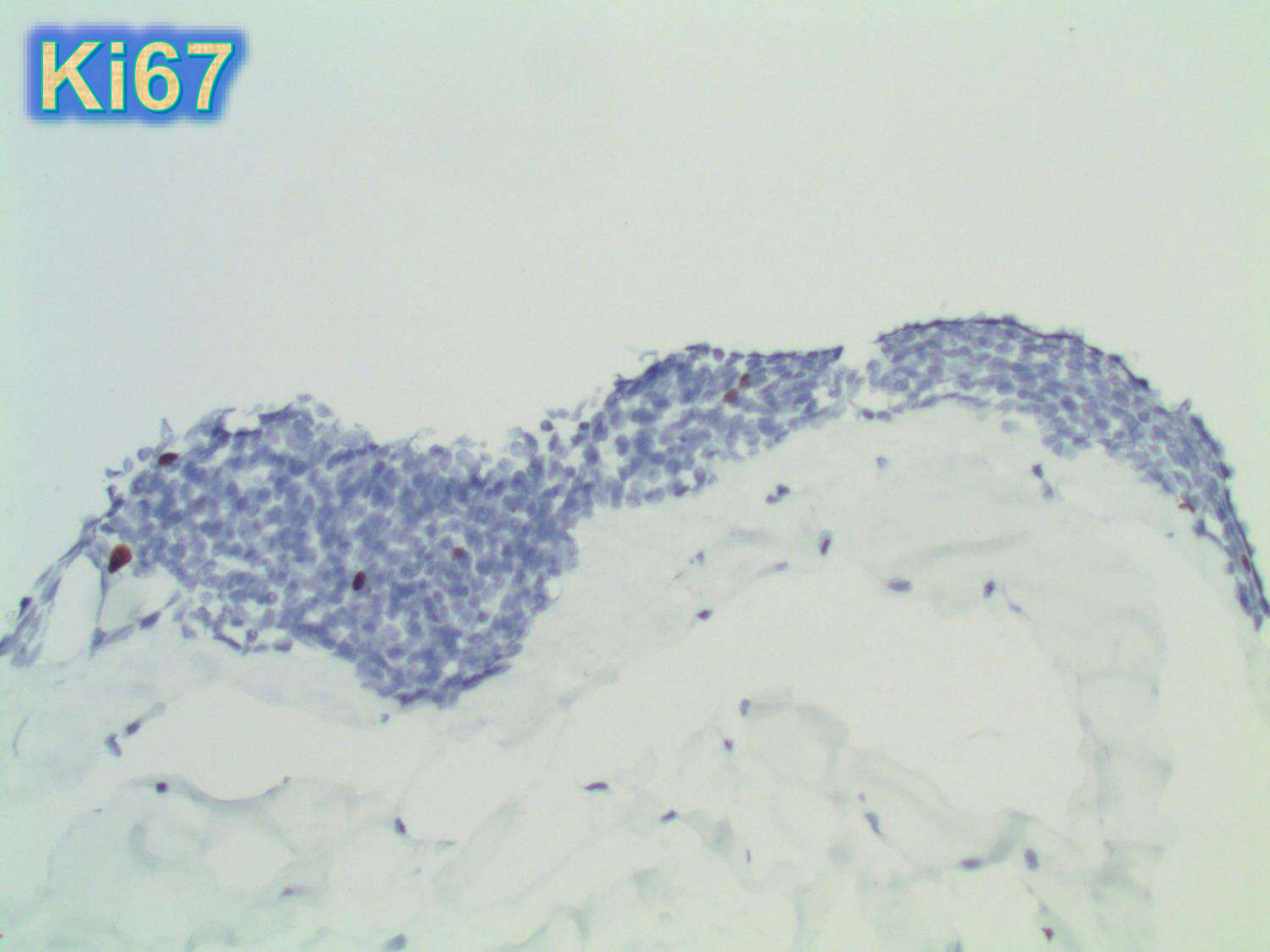


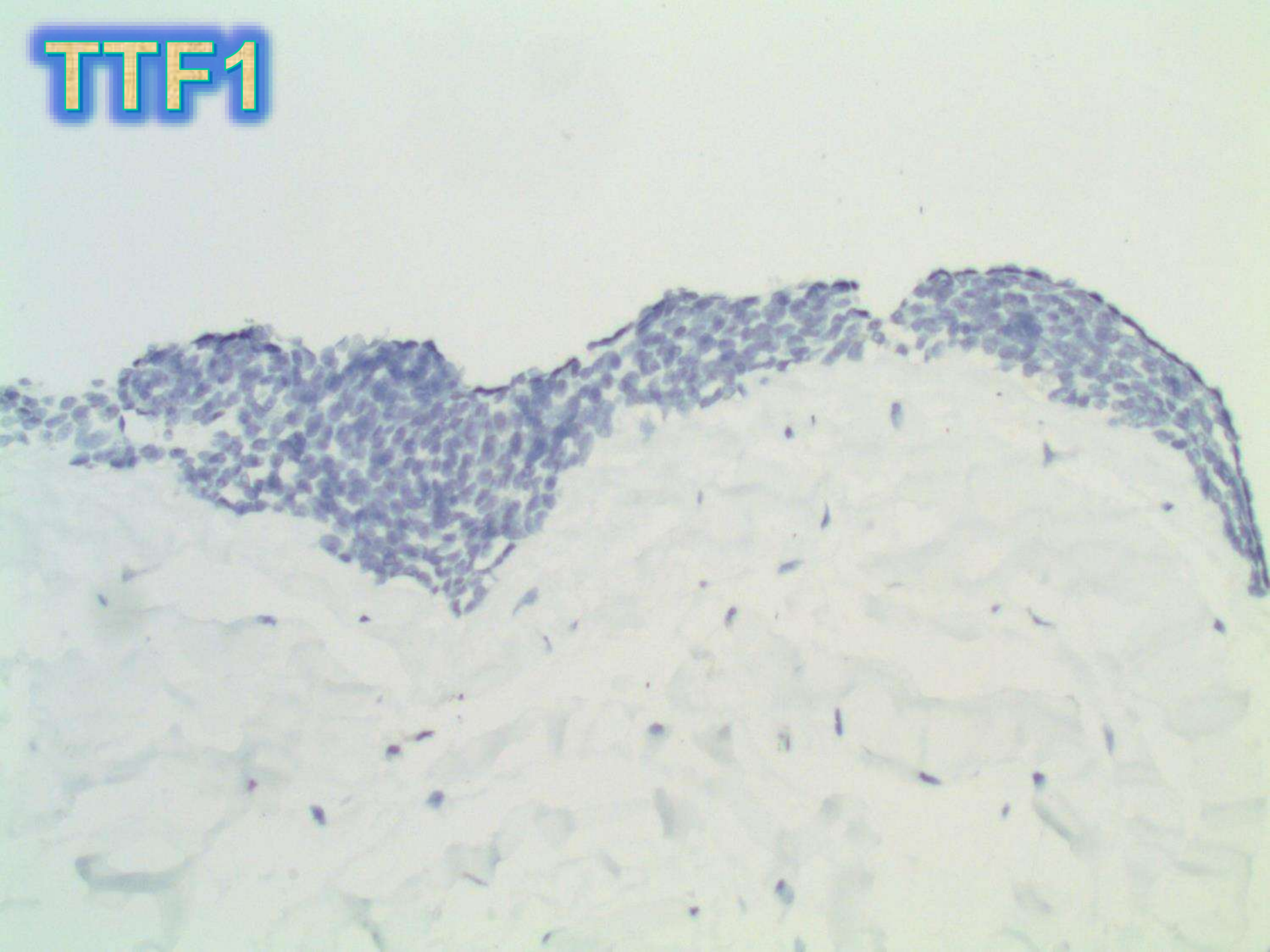



CD56

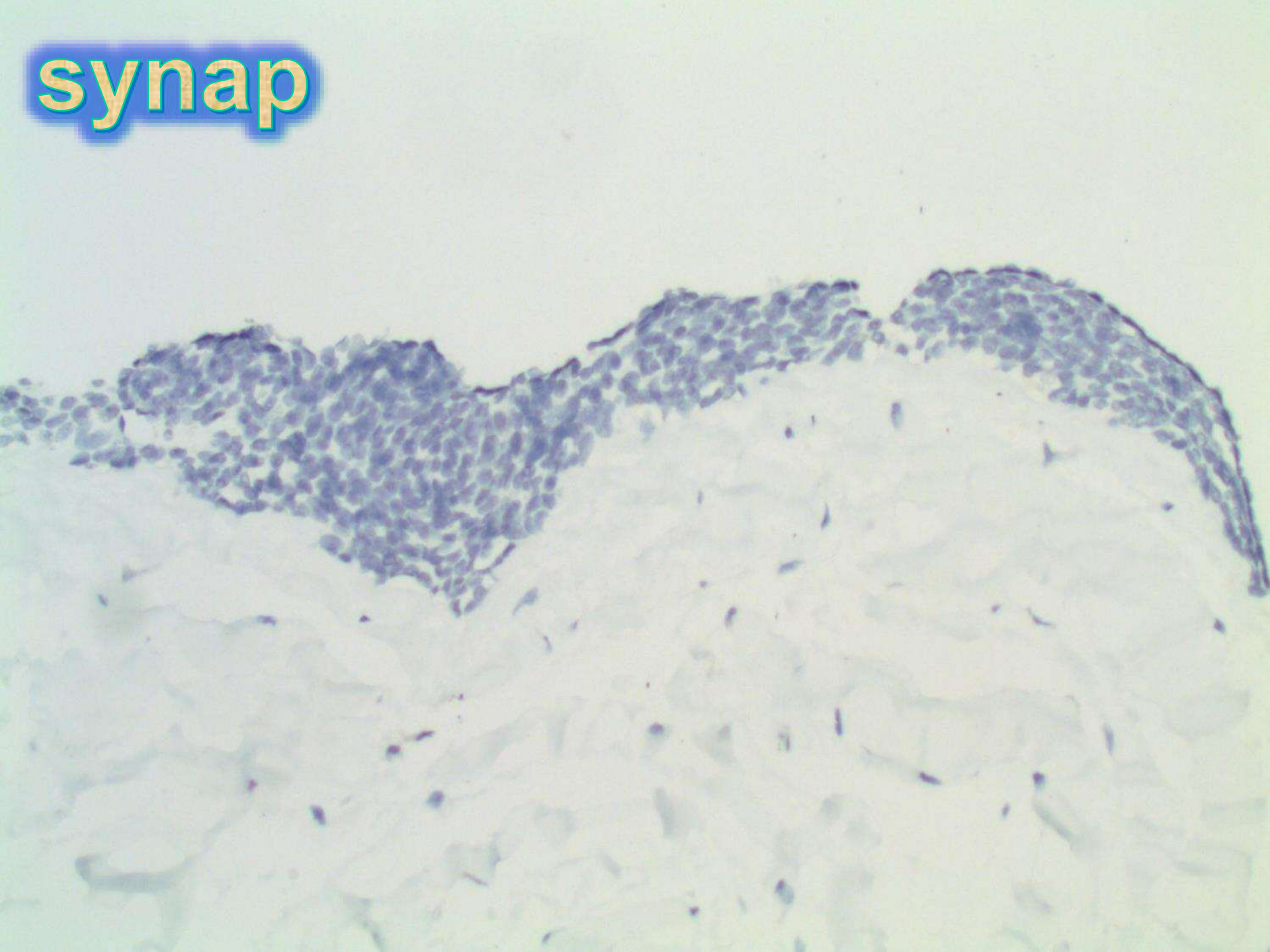
DDx

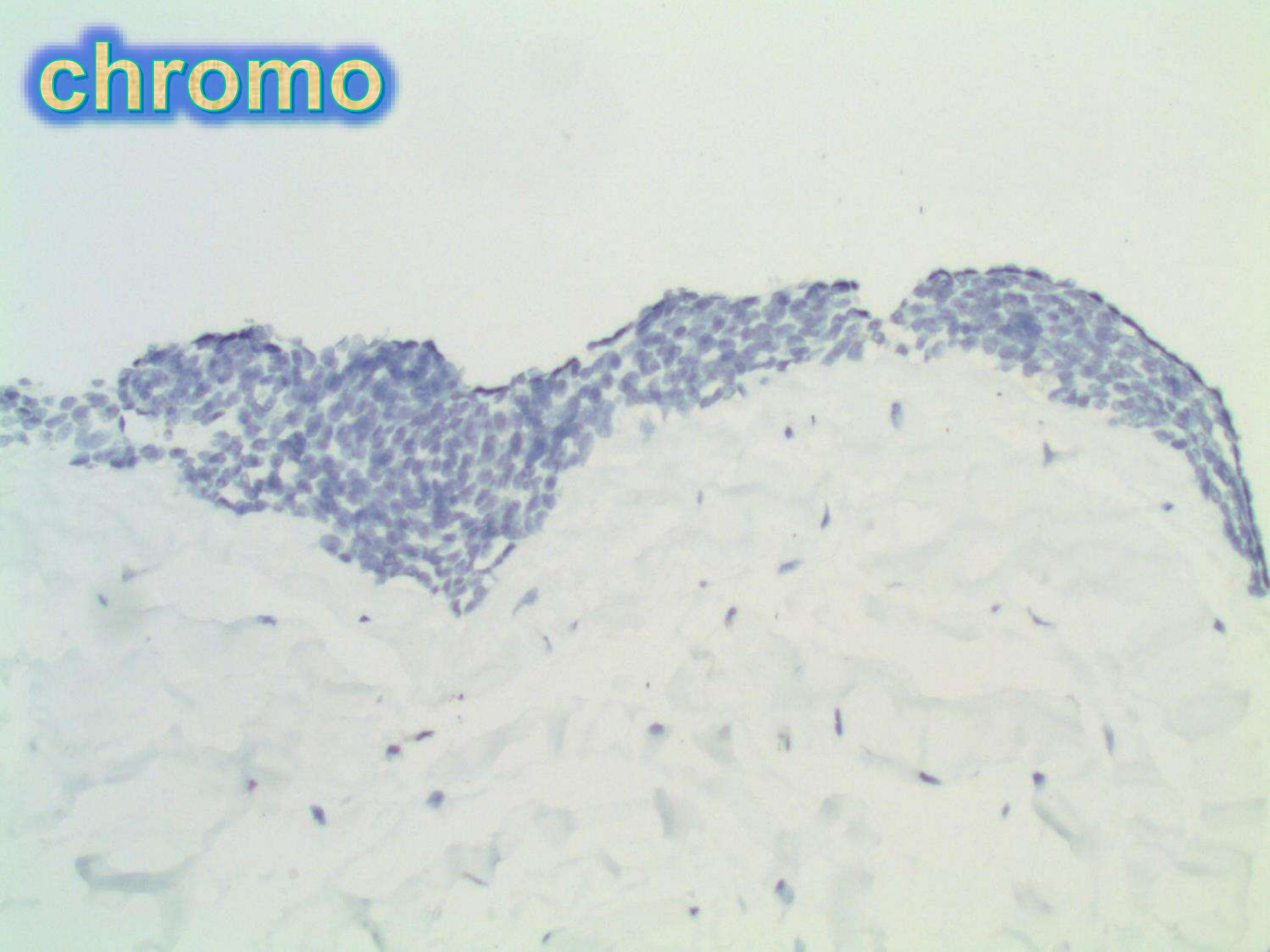

CD56

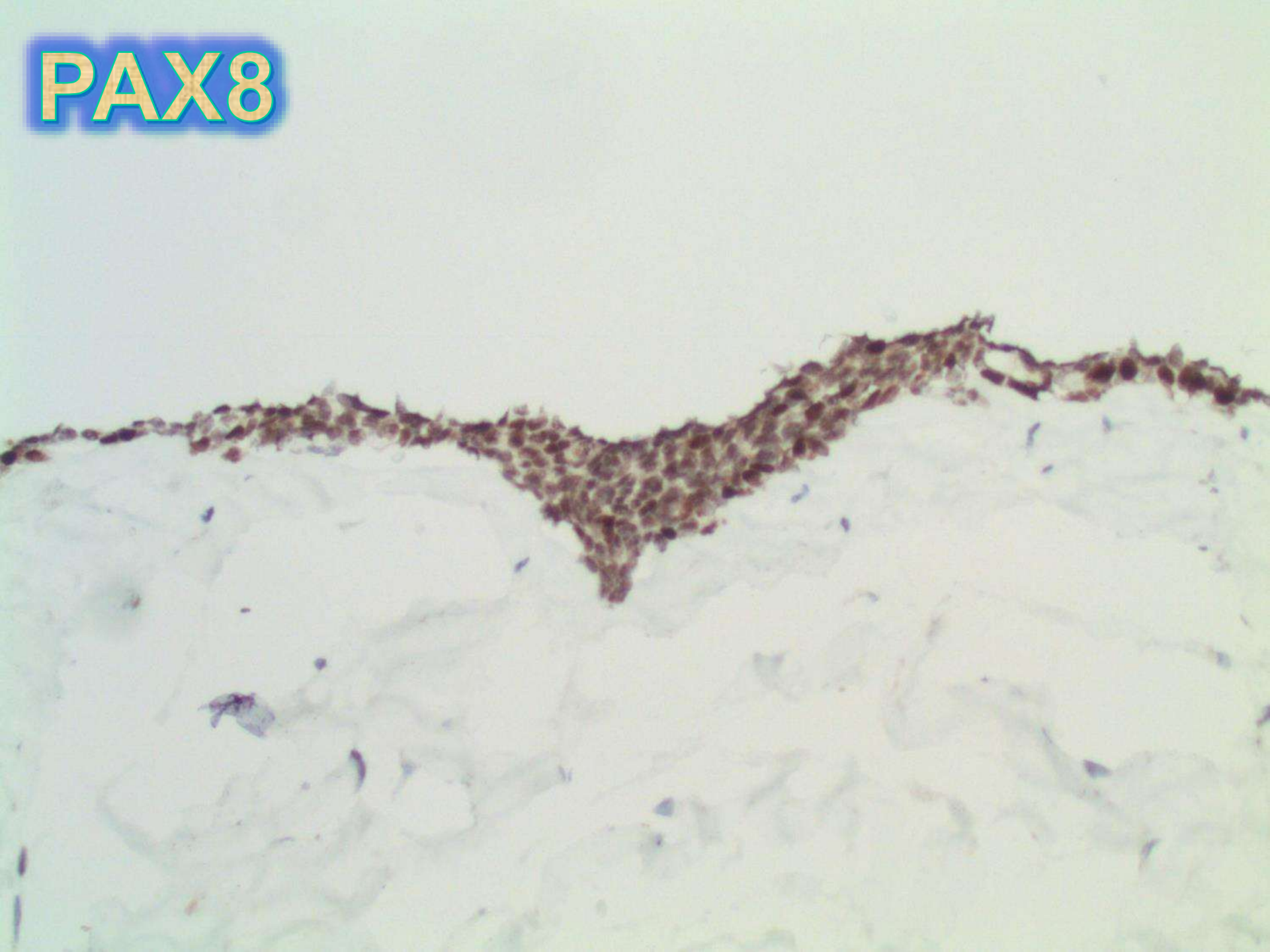

Ki67

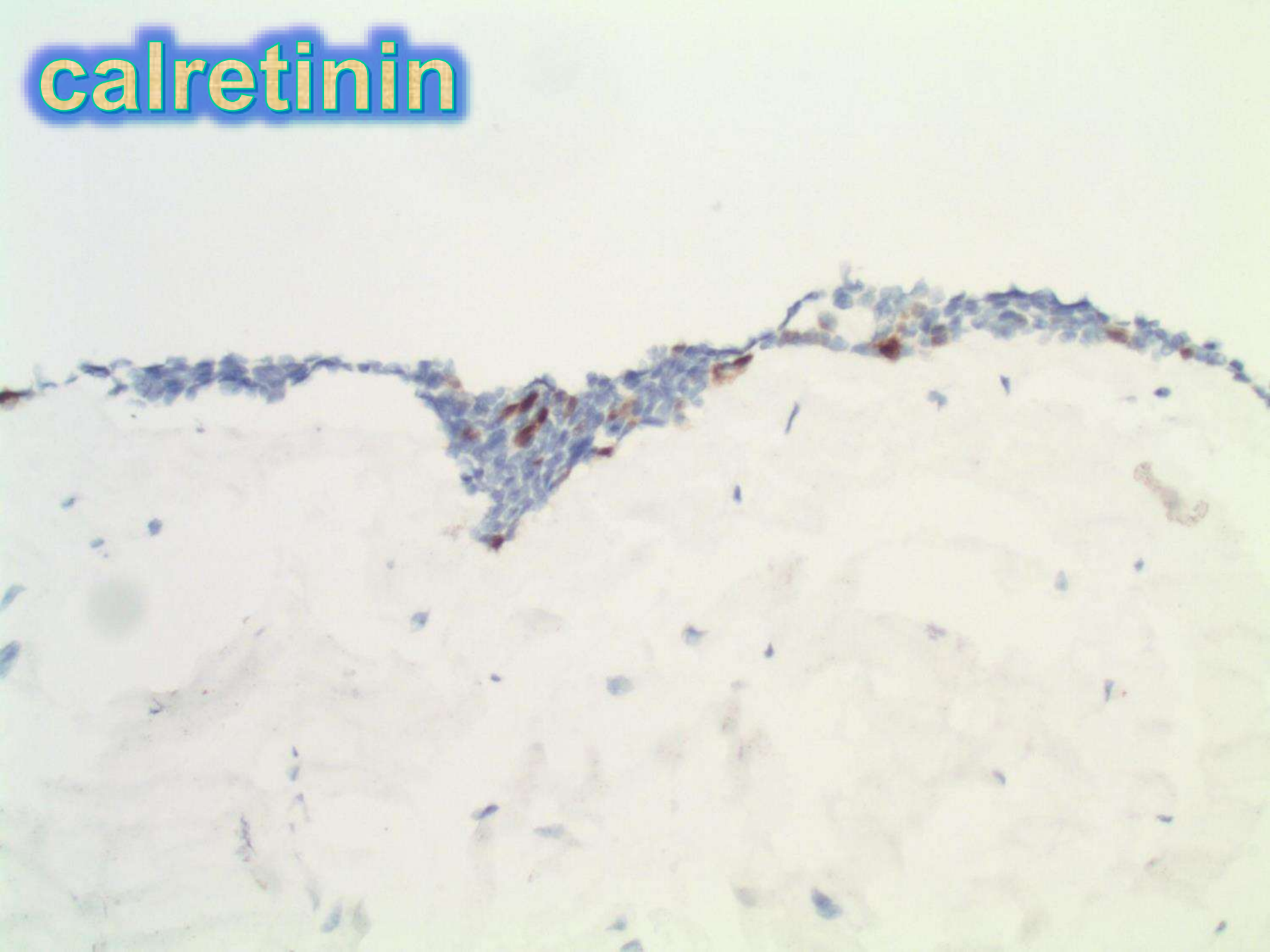

TTF1

synap




chromo


PAX8


calretinin

Human Pathology (2010) 41, 88–93

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Original contribution

Small blue cells mimicking small cell carcinoma in spermatocele and hydrocele specimens: a report of 5 cases

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Received 28 May 2009; revised 29 June 2009; accepted 30 June 2009

Keywords:
Hydrocele;
Spermatocele;
Small cell carcinoma

Summary We identified 5 cases of hydrocele and spermatocele resections containing detached small cellular "blue" clusters, raising questions of small cell carcinoma by contributors to our consult service. Patients were 37, 39, 52, 67, and 70 years old. None of the 4 patients with follow-up developed small cell carcinoma. On routine stained sections, there were multiple clusters of detached hypercellular cells with focal streaming, high nuclear-to-cytoplasmic ratios, and hyperchromatic nuclei without prominent nuclei. There were no mitotic figures, apoptotic bodies, or necrosis. In 4 of 5 cases, there was sufficient tissue to perform immunohistochemistry along with 10 cases each of normal rete testis and epididymis. CD56 was positive in 4 of 4 cases of the "blue cells" and in 9 of 10 of normal rete testis; yet, it was positive in only 2 of 10 normal epididymis. Synaptophysin and chromogranin were negative in all cases of "blue cells." PAX2 was negative in all cases of "blue cells" similar to the 1 of 9 positive staining in rete testis and in contrast to the positivity seen in 9 of 9 cases of normal epididymis. Ki-67 was negative or showed only rare positive cells in all of the cases of the "blue cells." Clusters of blue cells suggestive of sloughed rete testis cells can mimic small cell carcinoma in hydrocele and spermatocele specimens based on their low power appearance and positive CD56 staining. Closer examination of the cells' bland morphology, low expression of Ki-67, and lack of chromogranin and synaptophysin, along with recognition of this entity, can prevent a misdiagnosis of malignancy.

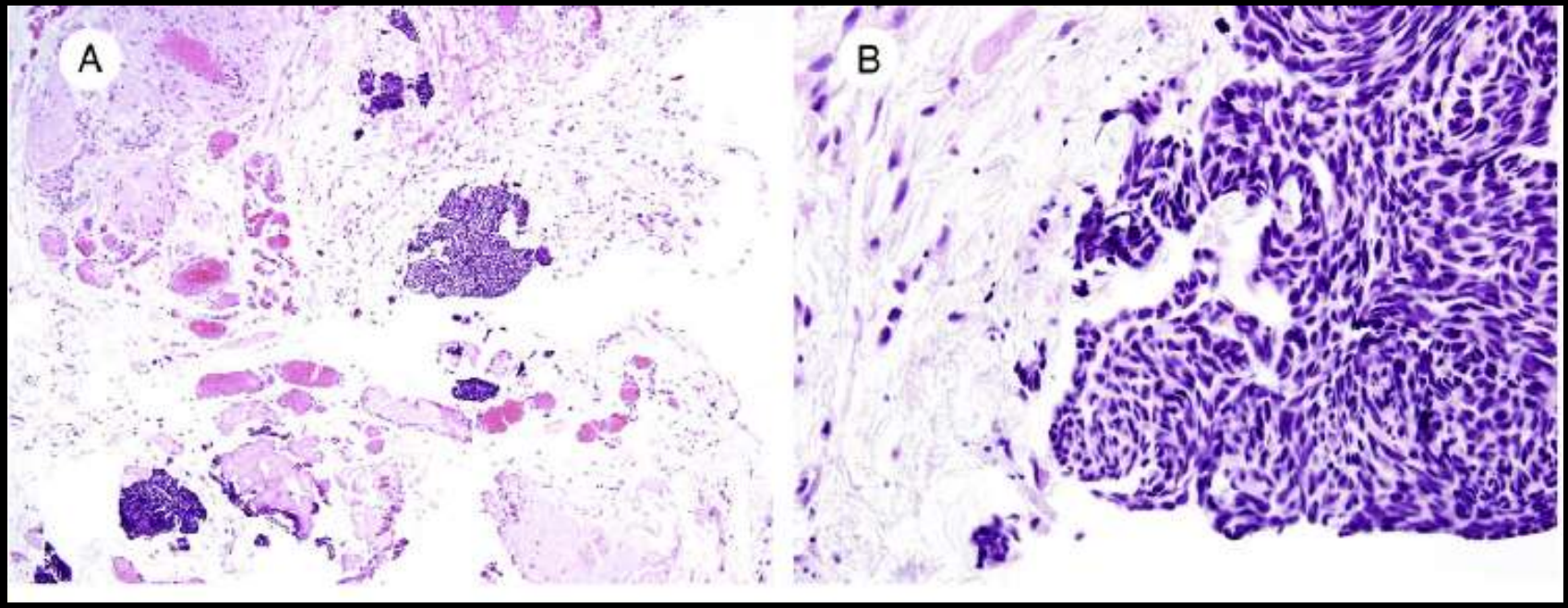

Table 1 Immunohistochemistry

	CK7/CK20	Calretinin	CK5/6	Synaptophysin	Chromogranin	CD56	PAX2
Blue cells	3/4:0/4	2/4	0/4	0/4	0/4	4/4	0/4
Rete testis	10/10:0/10	9/9	1/10	1/10	0/10	9/10	1/9
Epididymis	10/10:0/10	4/9	1/10	7/10	0/10	2/10	9/9

Human Pathology (2010) **41**, 88–93

PAX8 and PAX2 Immunostaining Facilitates the Diagnosis of Primary Epithelial Neoplasms of the Male Genital Tract

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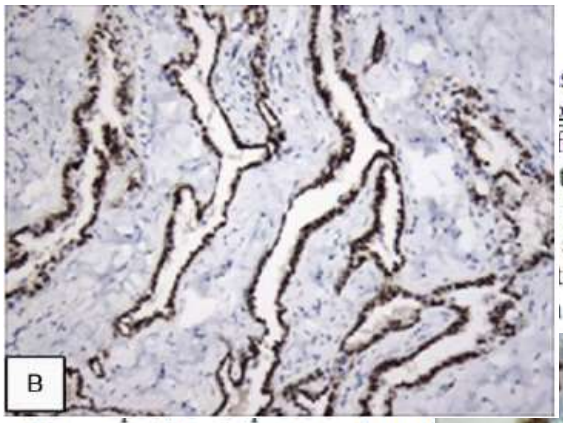
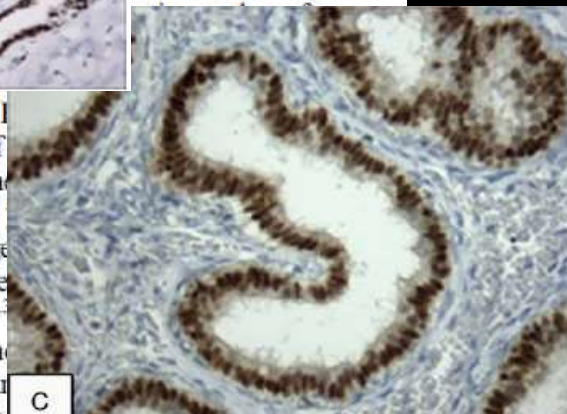
Abstract: PAX8 and PAX2 are cell-lineage-specific transcription factors that are essential for the development of Wolffian and Müllerian ducts and have recently emerged as specific diagnostic markers for tumors of renal or Müllerian origin. Little is known about their expression in the Wolffian duct-derived human male genital tract. We report our findings of PAX8 and PAX2 expression in the epithelium of the normal male genital tract and in epithelial tumors derived therefrom using immunohistochemistry (IHC). We found that PAX8 and PAX2 were expressed in the epithelium of the male genital tract from the rete testis to the ejaculatory duct. Rare glands in the prostatic central zone, a tissue of purported Wolffian duct origin, were focally positive for PAX2, but no PAX8 was detected in this area, a finding that may warrant further study. We found diffuse expression of PAX8 and PAX2 in 1 case each of serous cystadenoma of the epididymis, carcinoma of the rete testis, Wolffian adnexal tumor of the seminal vesicle, and endometrioid carcinoma of the seminal vesicle. Neither PAX8 nor PAX2 was detected in the seminiferous tubules and interstitium of the normal testis, nor in Leydig cell tumors (n = 6), Sertoli cell tumors (n = 2), or 48 of 49 germ cell tumors. One pediatric yolk sac tumor showed focal and weak staining for PAX8. Tumors of mesothelial origin, that is, adenomatoid tumors (n = 3) and peritoneal malignant mesotheliomas (n = 37) in men, were negative for PAX2 and PAX8. Neither PAX2 nor PAX8 was present in other areas of the prostate. Expression of PAX8 and PAX2 in these primary epithelial neoplasms of the male genital tract is due to their histogenetic relationship with Wolffian or Müllerian ducts. PAX8 and PAX2 IHC may facilitate the diagnosis of these tumors and should be included in the differential diagnostic IHC panel.

Key Words: PAX8 and PAX2, epithelial tumor, male genital tract

(*Am J Surg Pathol* 2011;35:1473–1483)





scription factors
gan development
fic patterns.[22,25]
the development
in Wolffian and
stages; they are
these ducts, such
tubes.[3,9,10,39,40]

However, little is known about
or neoplastic epithelial cells of
tract, which is a derivative of th
Although rare,[11,17,37,46,54]
of the Wolffian-derived male g
they can be a diagnostic challe
specific histologic features.[1,17,18,
the male genital tract is a diagn
thorough clinical investigation a