

# Disclosures

## February 6, 2017

Dr. Benjamin Buelow has disclosed that he is now an employee of TeneoBio, Inc., a commercial interest as defined by ACCME. He submitted his case prior to his employment of TeneoBio, Inc. and the case itself is not relevant to his employment and will be presented on his behalf by Dr. Julieann Lee of UCSF. Dr. Buelow has recused himself from any control in the content of the presentation.

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

### Presenters:

Kerri Rieger, MD  
Charles Lombard, MD  
Alana Shain, MD  
Sunny Kao, MD  
Mahendra Ranchod, MD  
Malti Kshirsagar, MD  
Seth Lummus, MD  
Don Born, MD  
Julieann Lee, MD  
Ankur Sangoi, MD

### Activity Planners/Moderator:

Kristin Jensen, MD  
Ankur Sangoi, MD

# SB 6131

**Kerri Rieger/Charles Lombard; Stanford/El Camino Hospital**

62-year-old male with one month history of cough and shortness of breath and one week history of widespread rash and mucosal erosions PMH: retroperitoneal Castleman disease Clinical ddx: EM/SJS, lichenoid drug eruption, r/o paraneoplastic pemphigus.

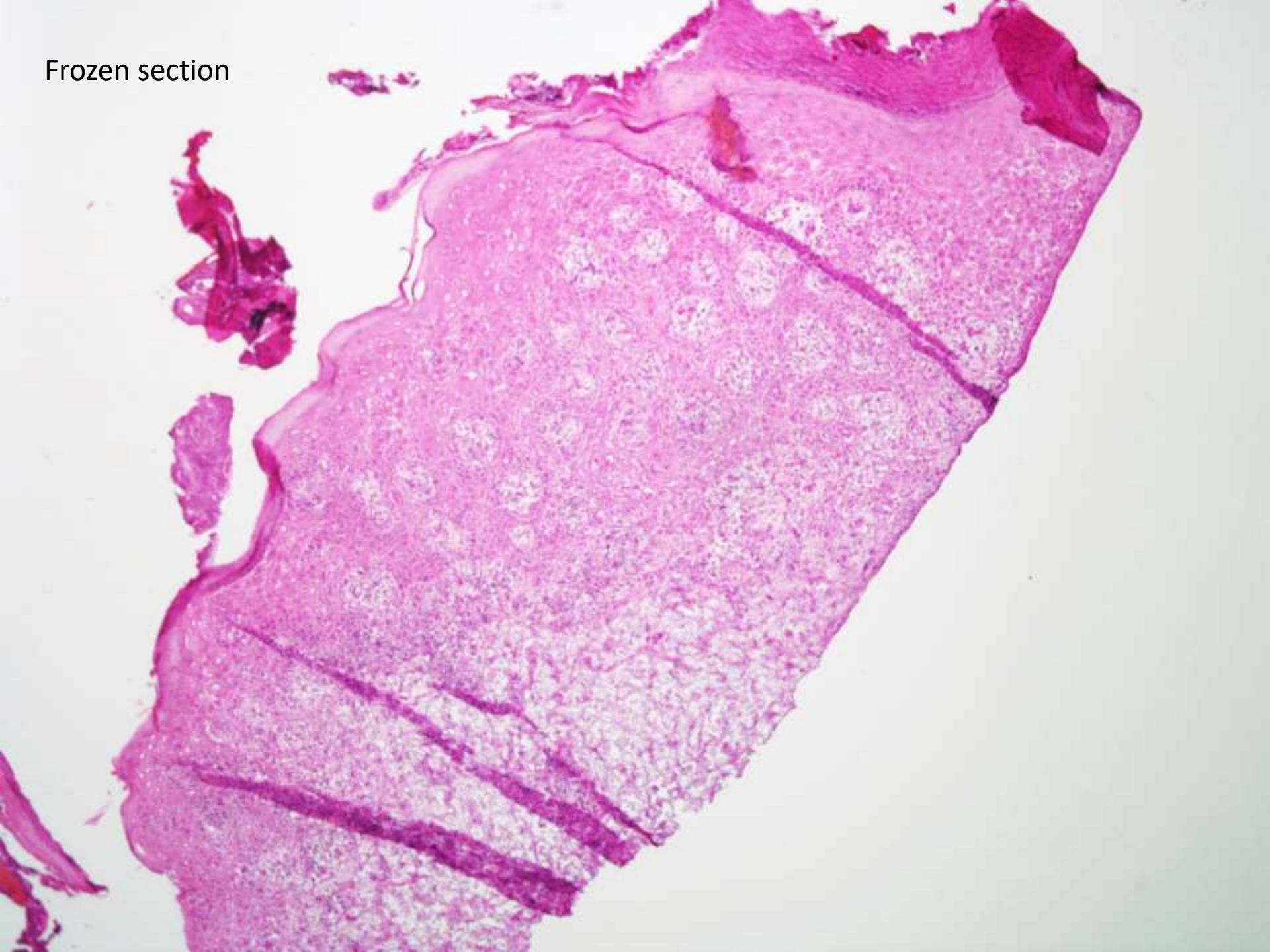






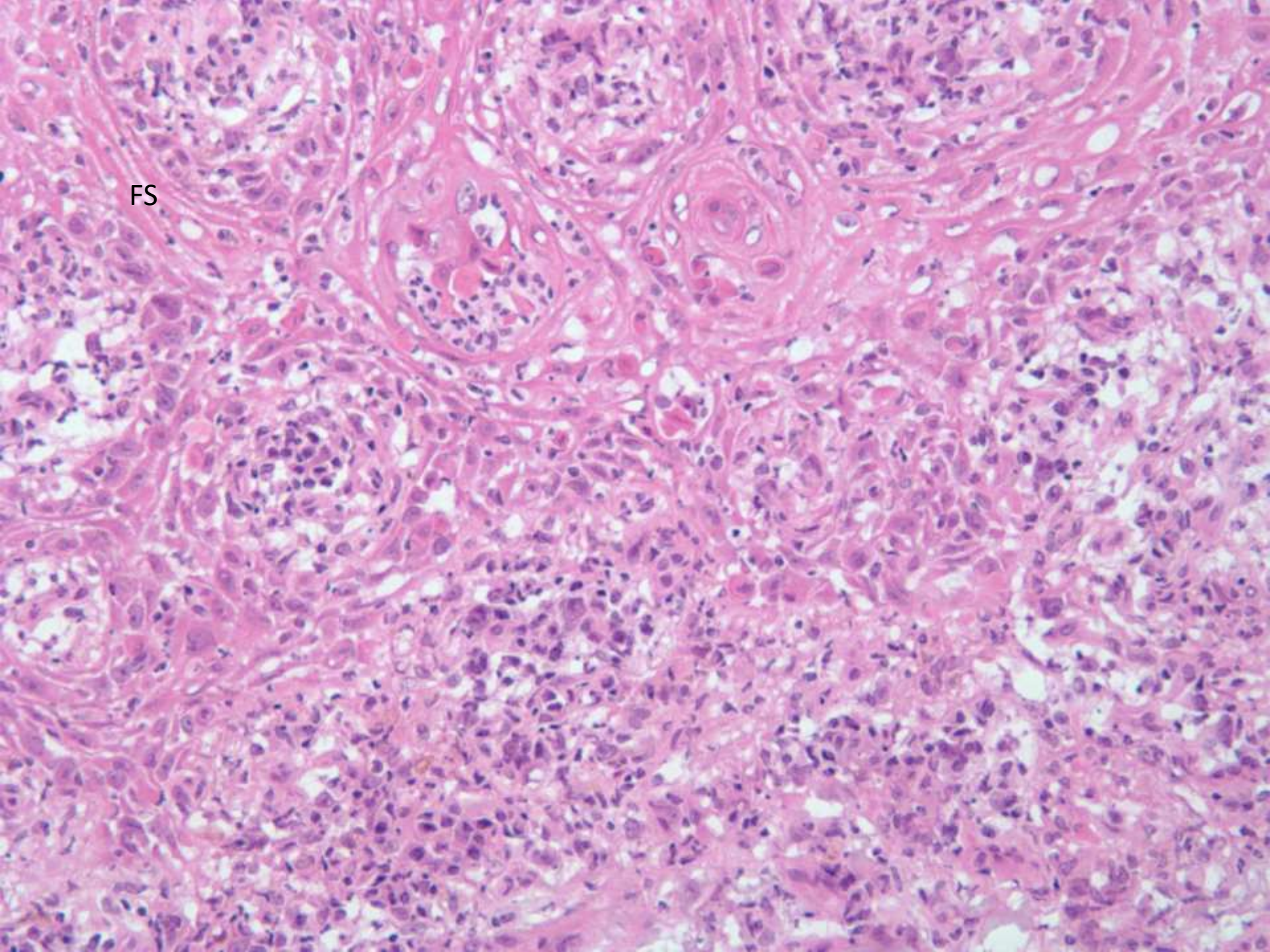


Frozen section



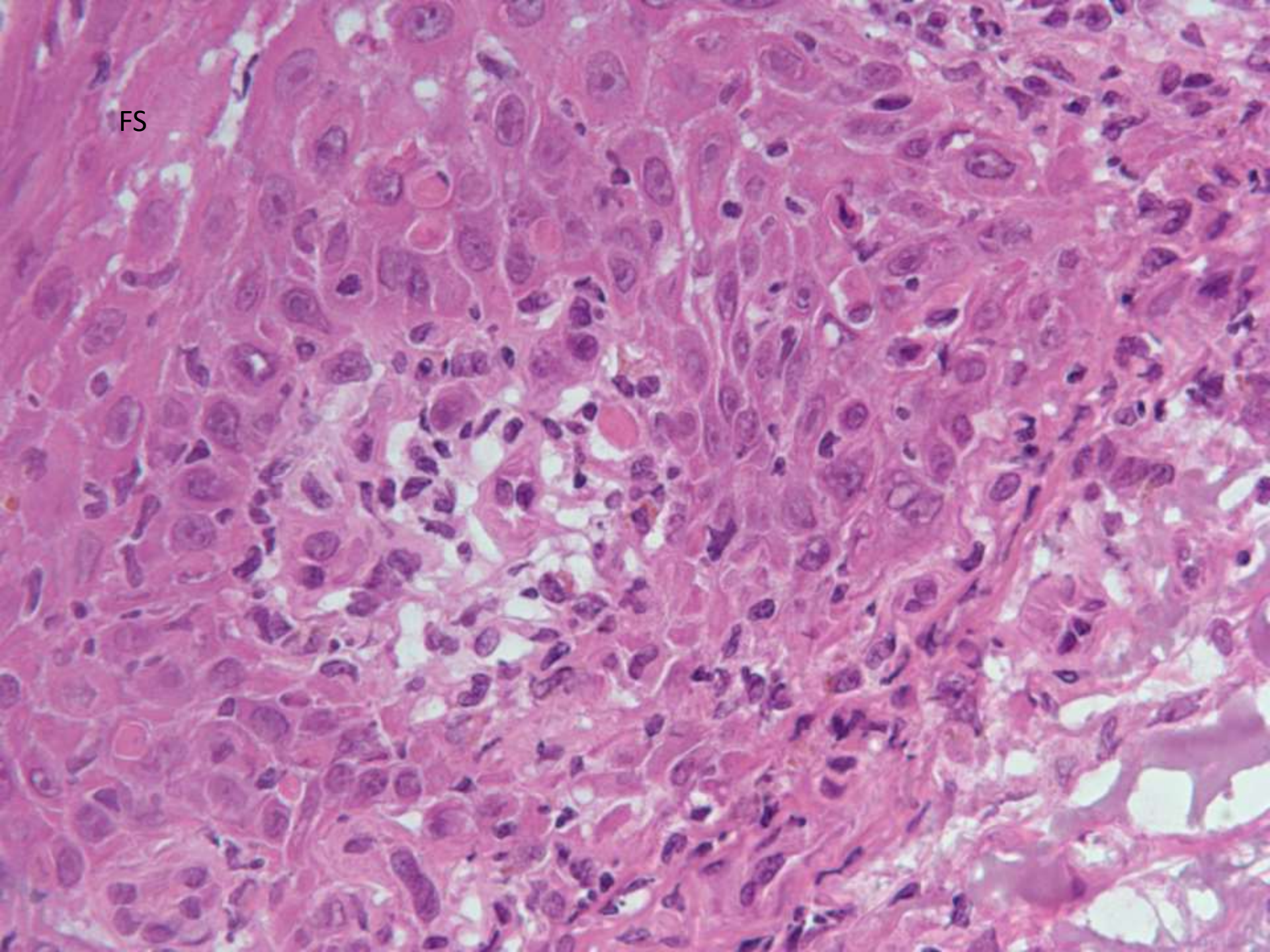


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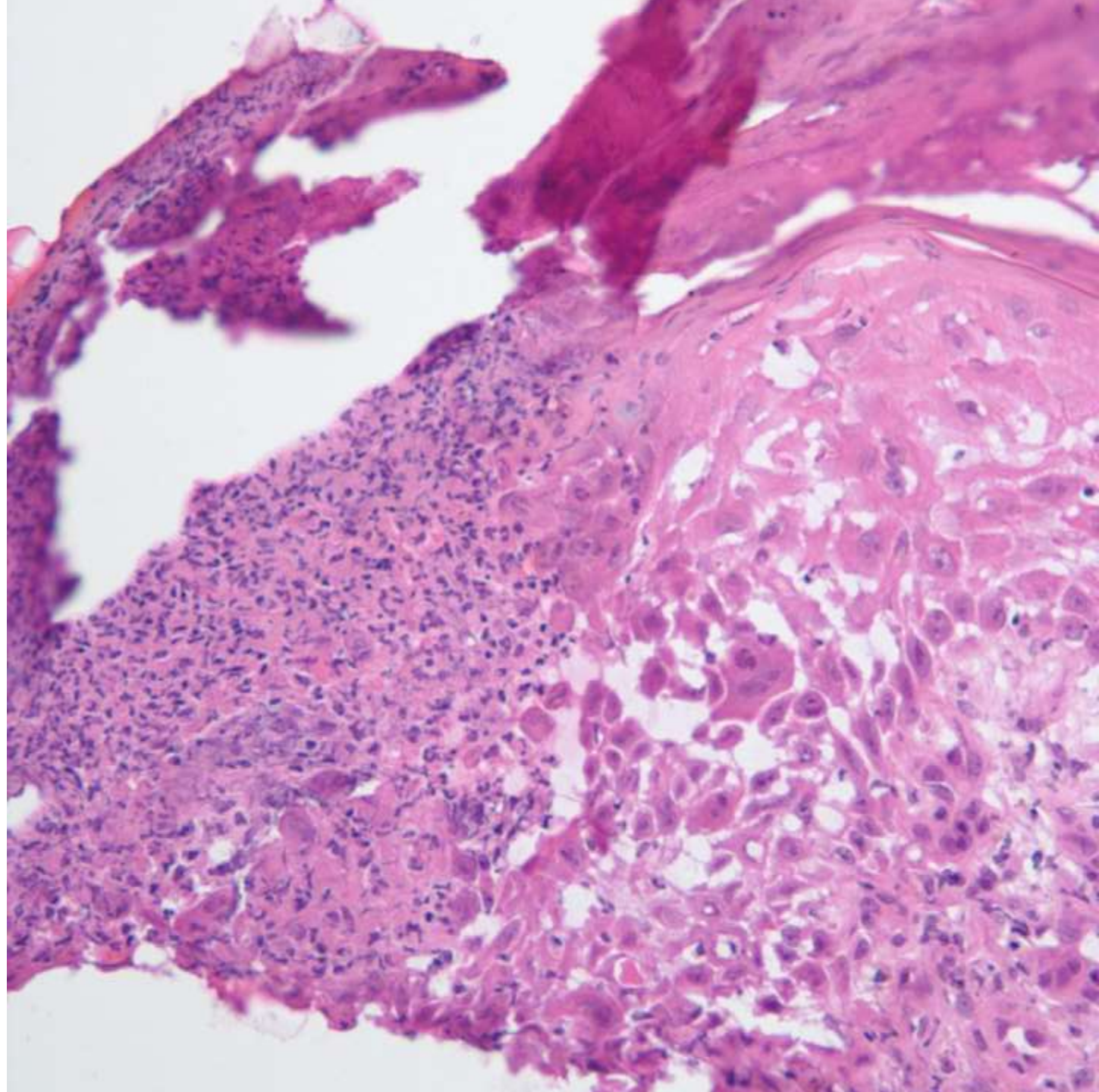




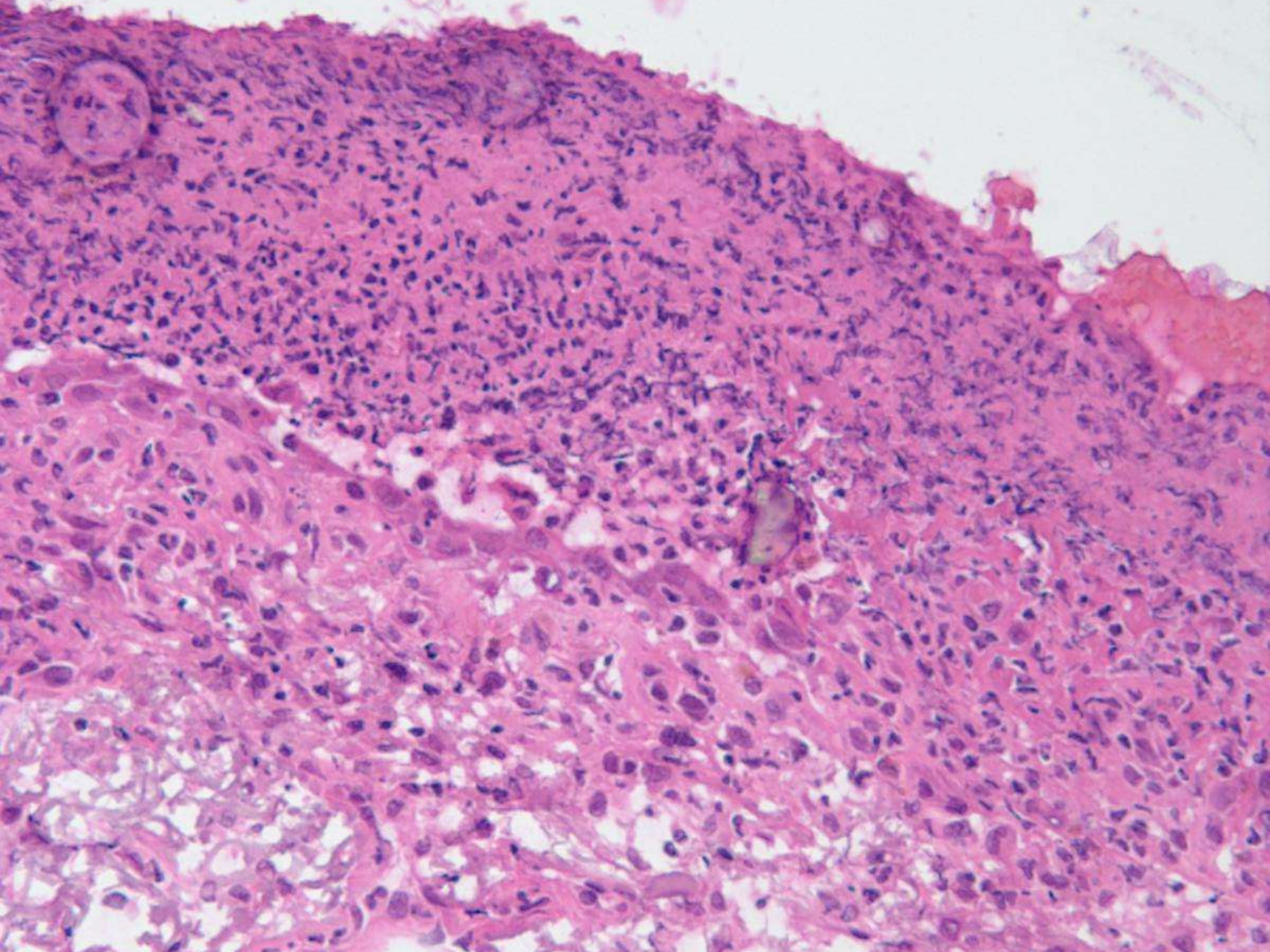
FS



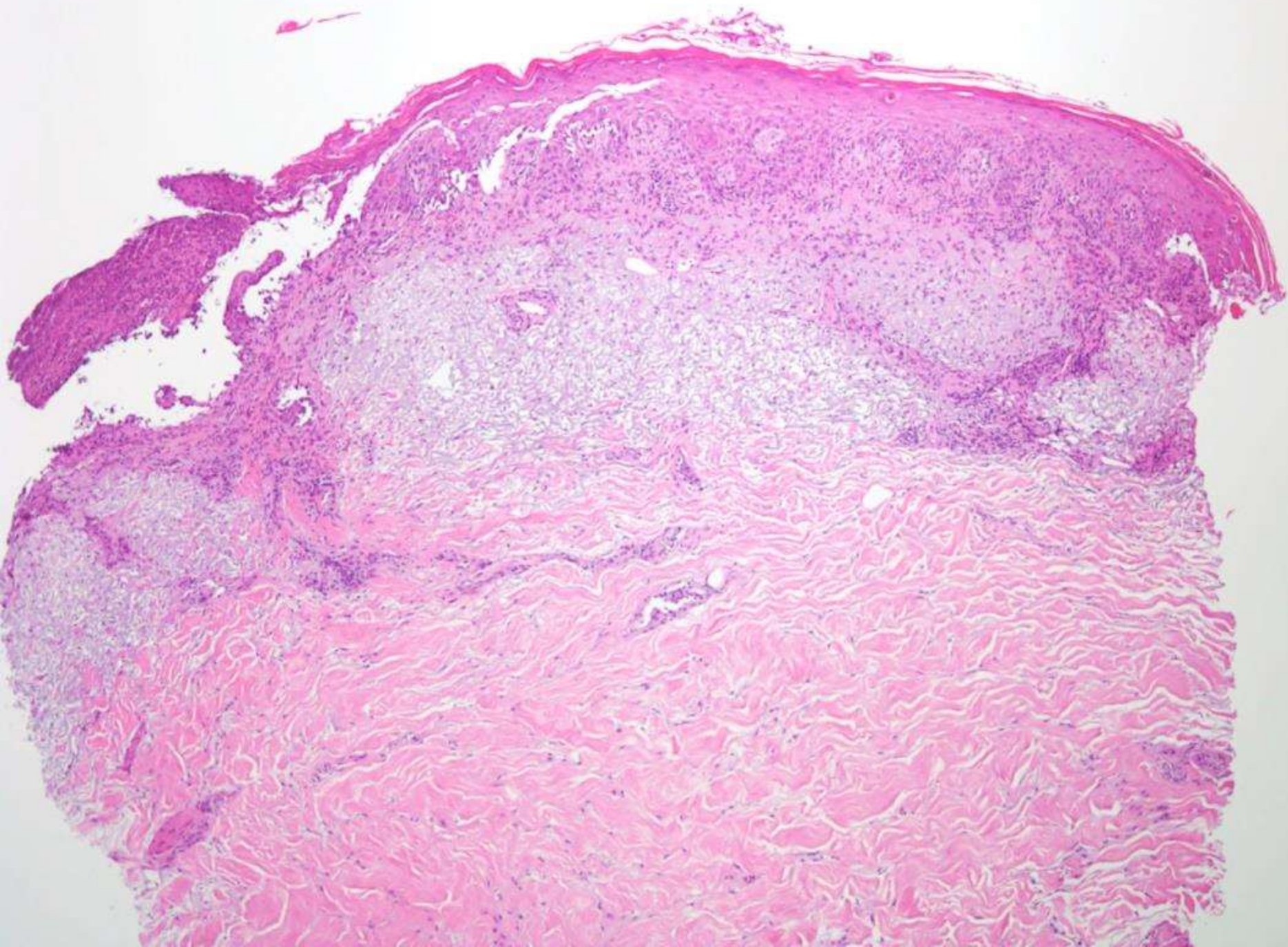




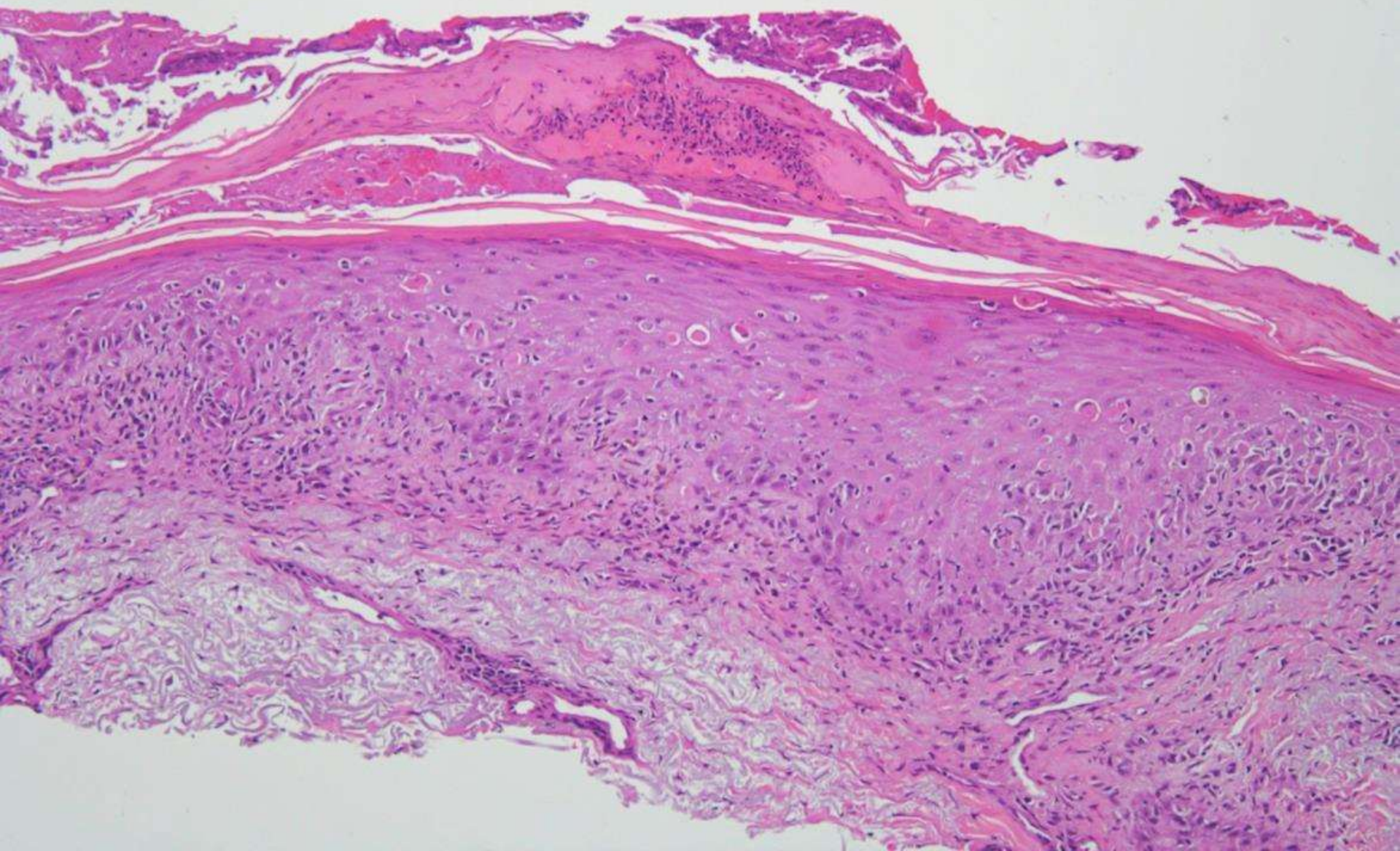




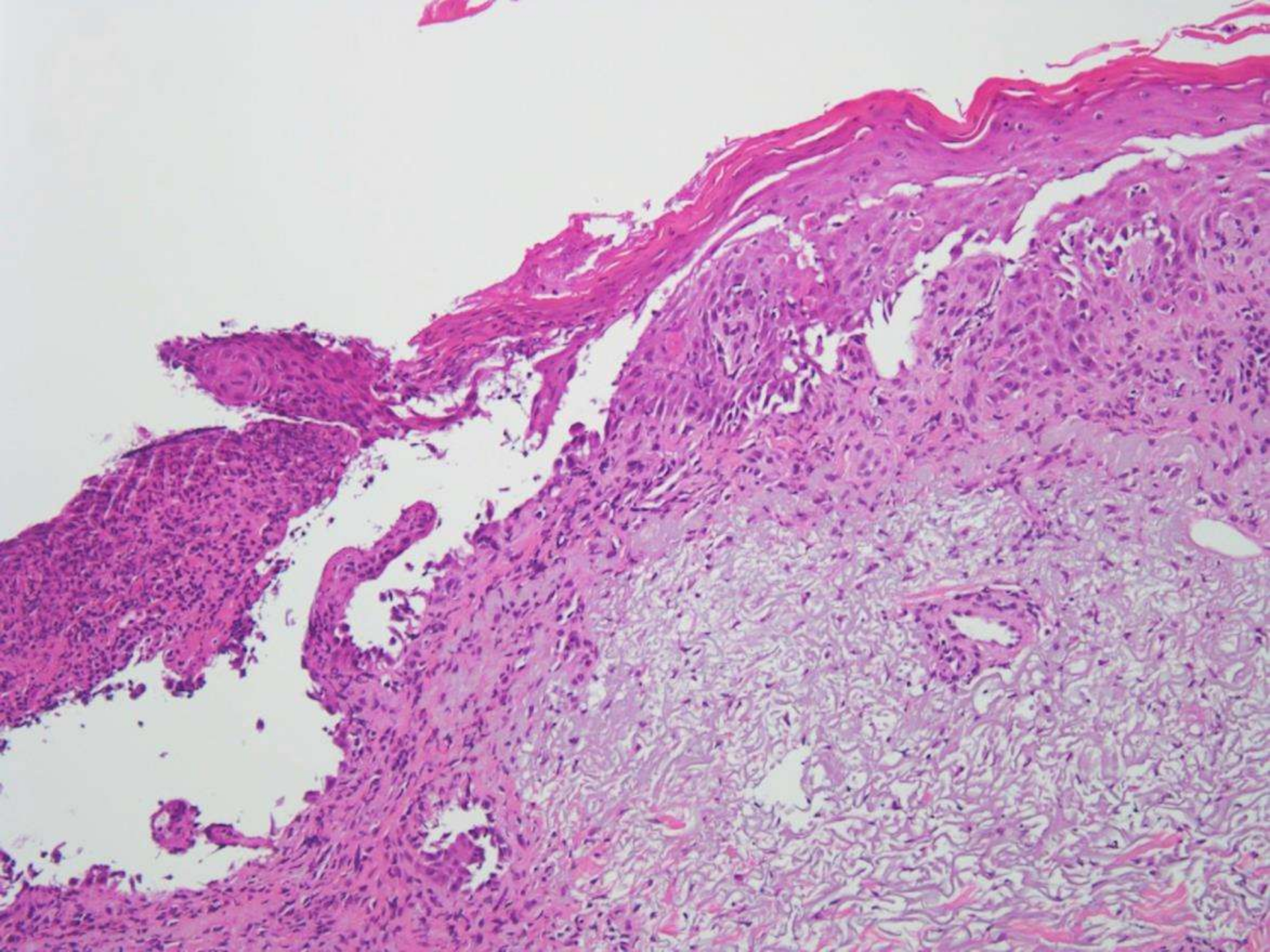




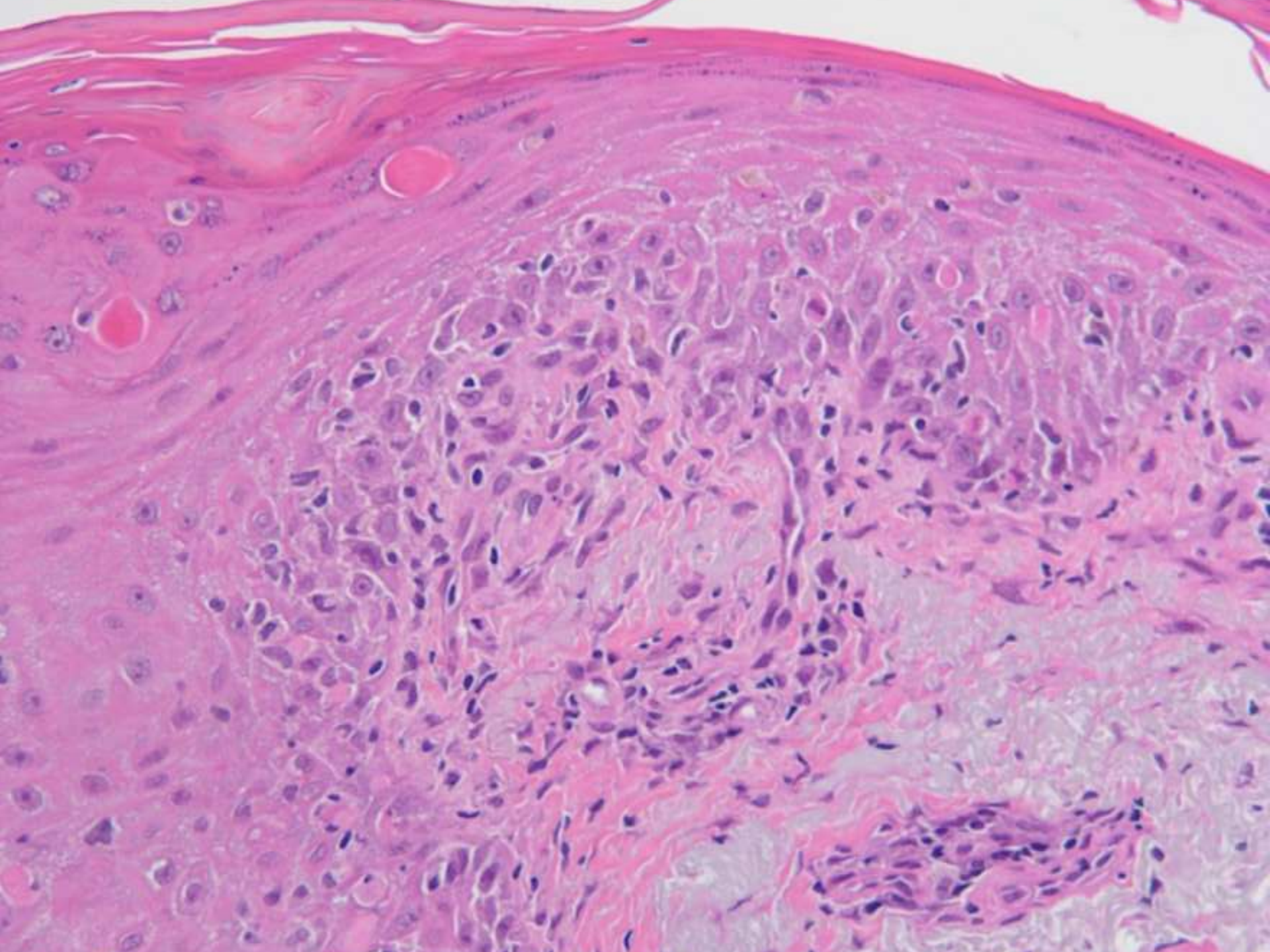




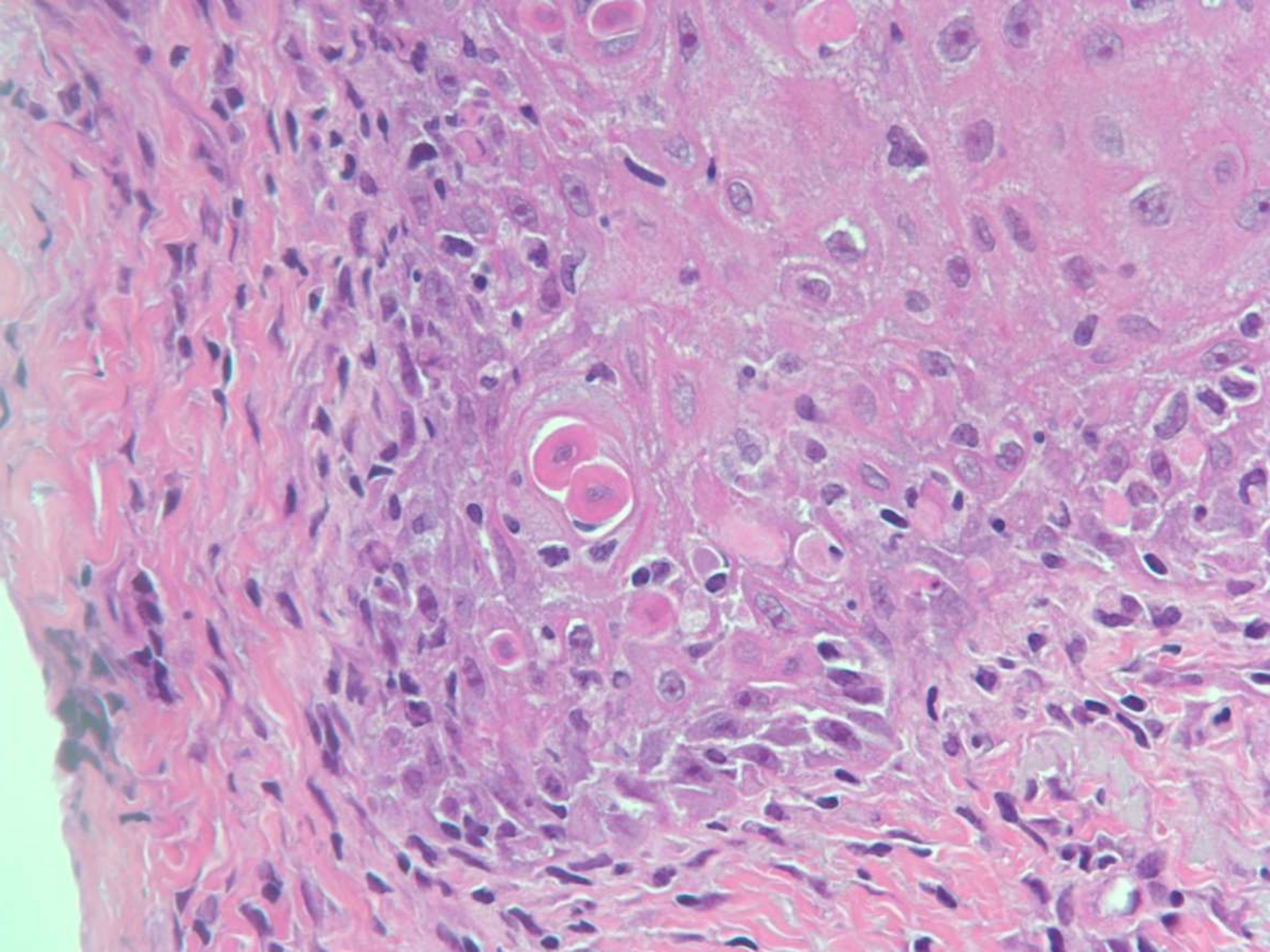










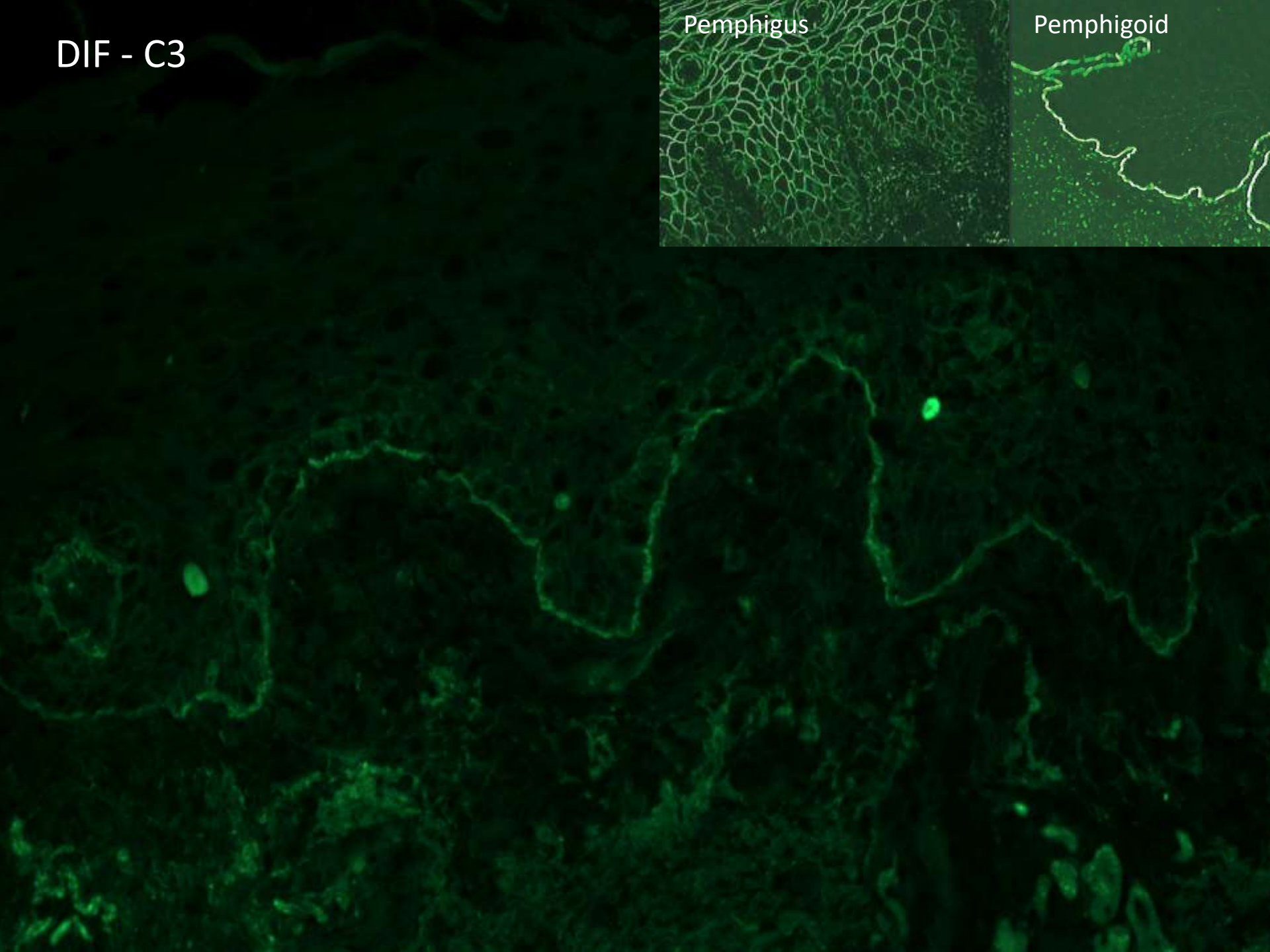




DIF - C3

Pemphigus

Pemphigoid



# Paraneoplastic Pemphigus (PNP)

- Autoimmune blistering disorder that occurs in the setting of lymphoproliferative disorders and other malignancies
- Clinical presentation typically begins with painful, severe stomatitis
- Cutaneous lesions are polymorphic and can resemble erythema multiforme, lichen planus, pemphigus vulgaris and bullous pemphigoid





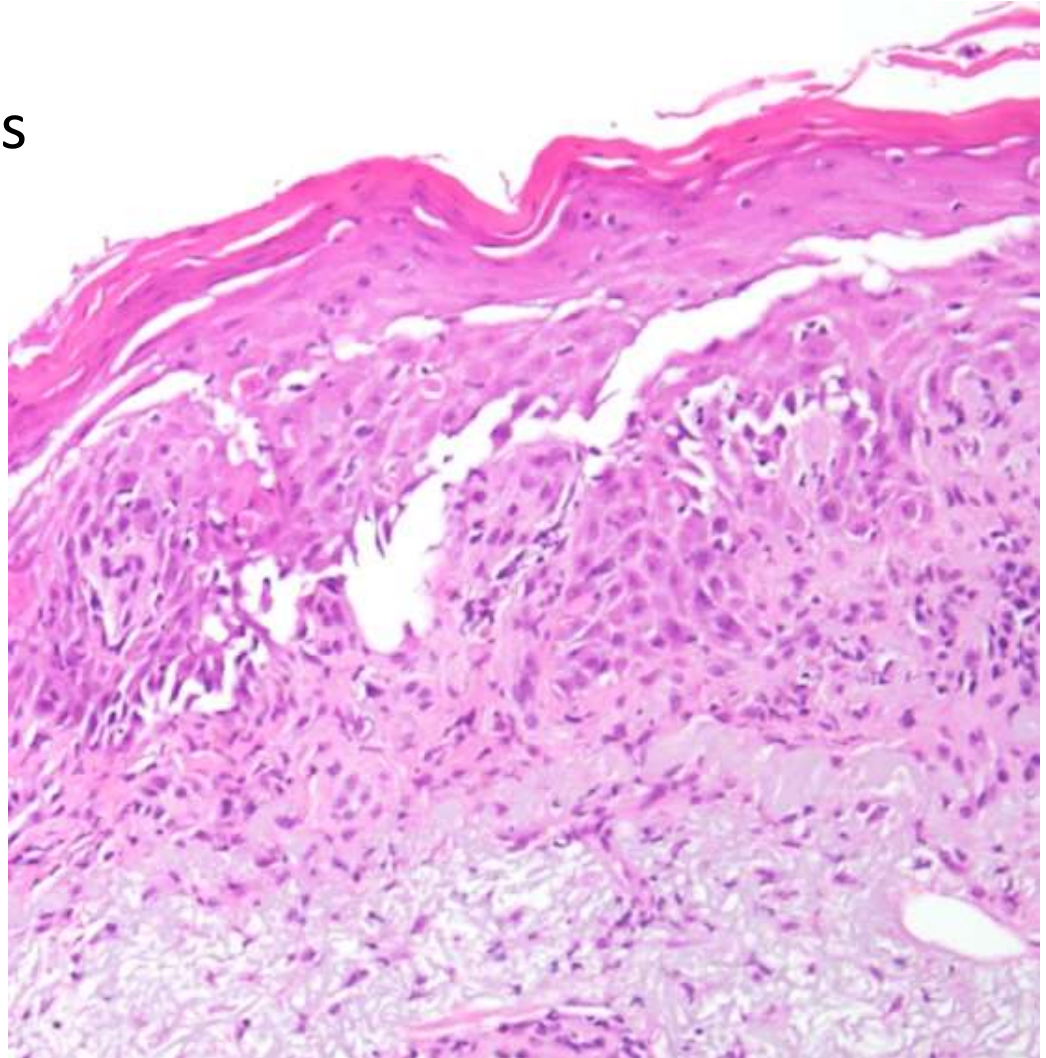
# Paraneoplastic Pemphigus (PNP)

- Mortality is high (up to 90%) and can be partly attributed to pulmonary involvement, usually in the form of bronchiolitis obliterans
- Disease results from autoantibodies that target a number of antigens, the most important of which are thought to be the plakin proteins
- Management:
  - Treatment of the underlying malignancy
  - Suppression of disease activity (systemic steroids and other immunosuppressants)

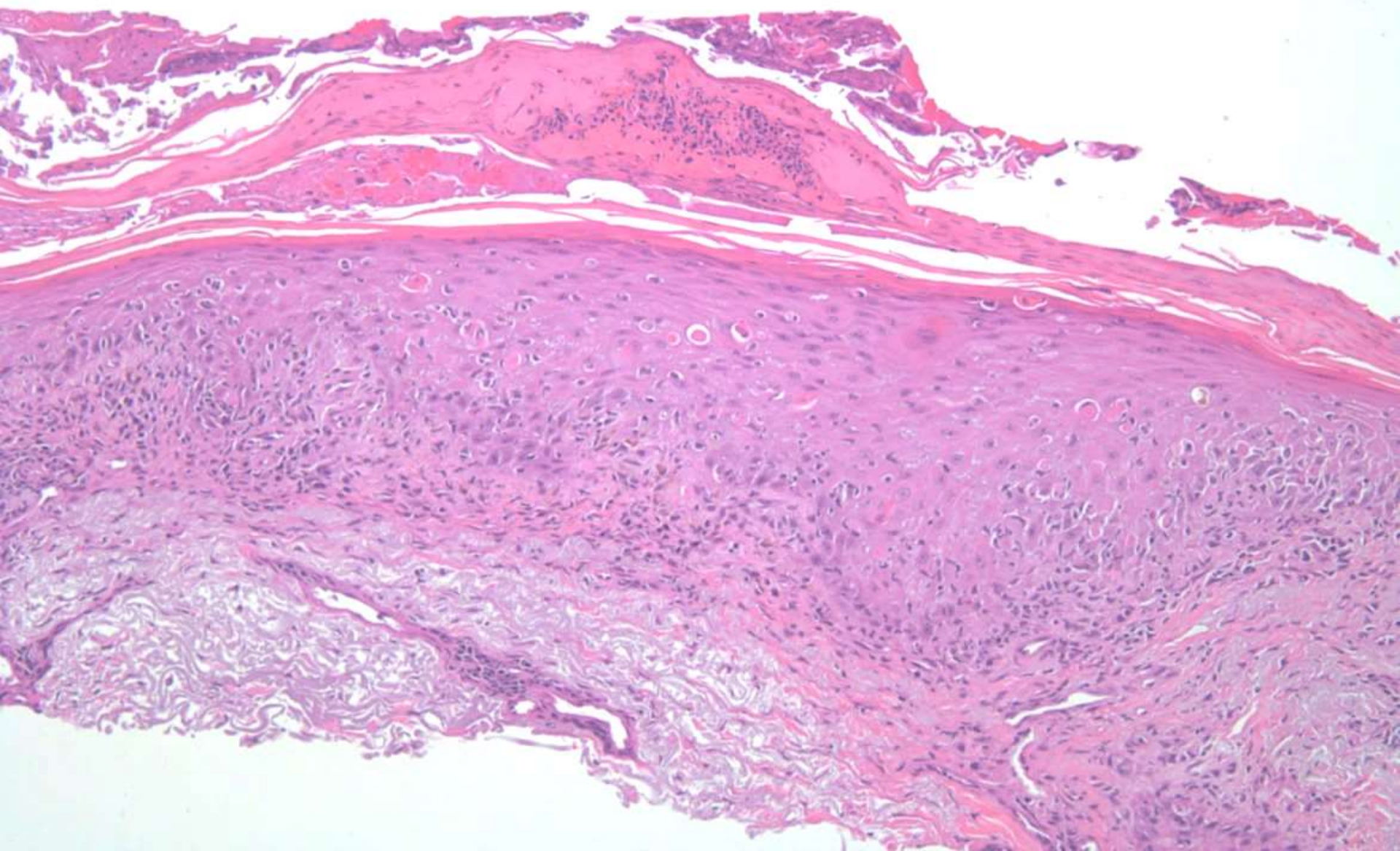


# PNP: histopathology

- Lichenoid/interface dermatitis with dyskeratosis, acantholysis
- +/- subepidermal bullae



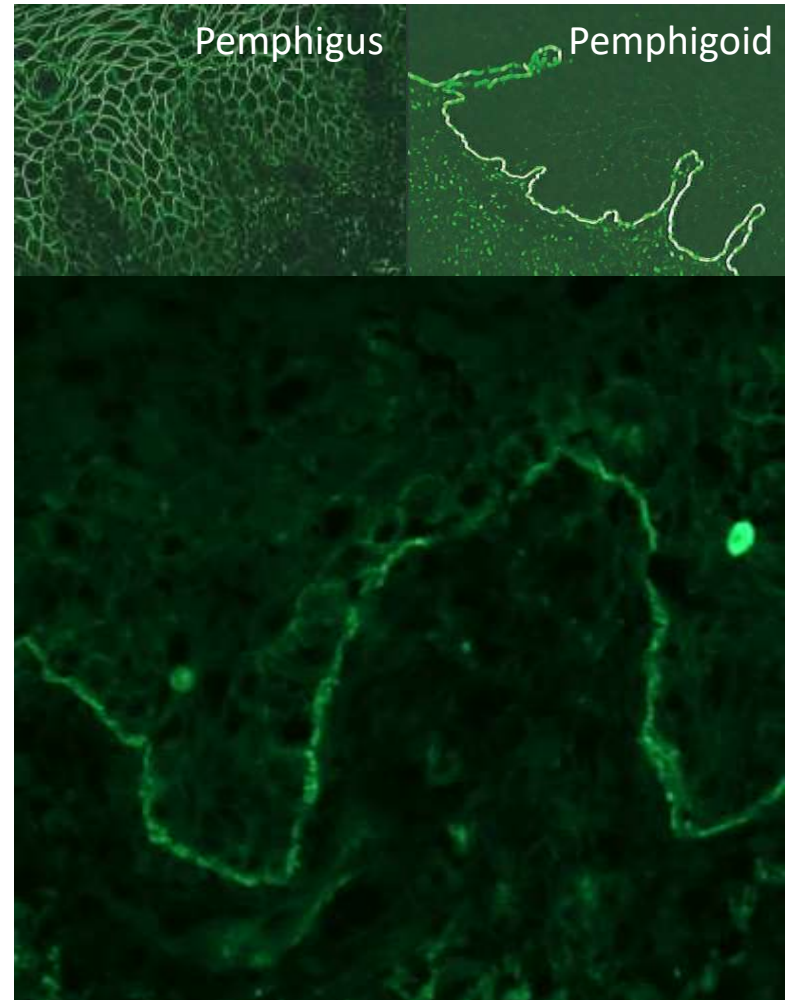






# PNP: immunofluorescence

- **Direct IF** shows IgG and C3 on epidermal cell surfaces and at BMZ
- 41% sensitivity; 87% specificity
- **Indirect IF** on transitional epithelial tissue (typically rat bladder)
- 86% sensitivity; 98% specificity



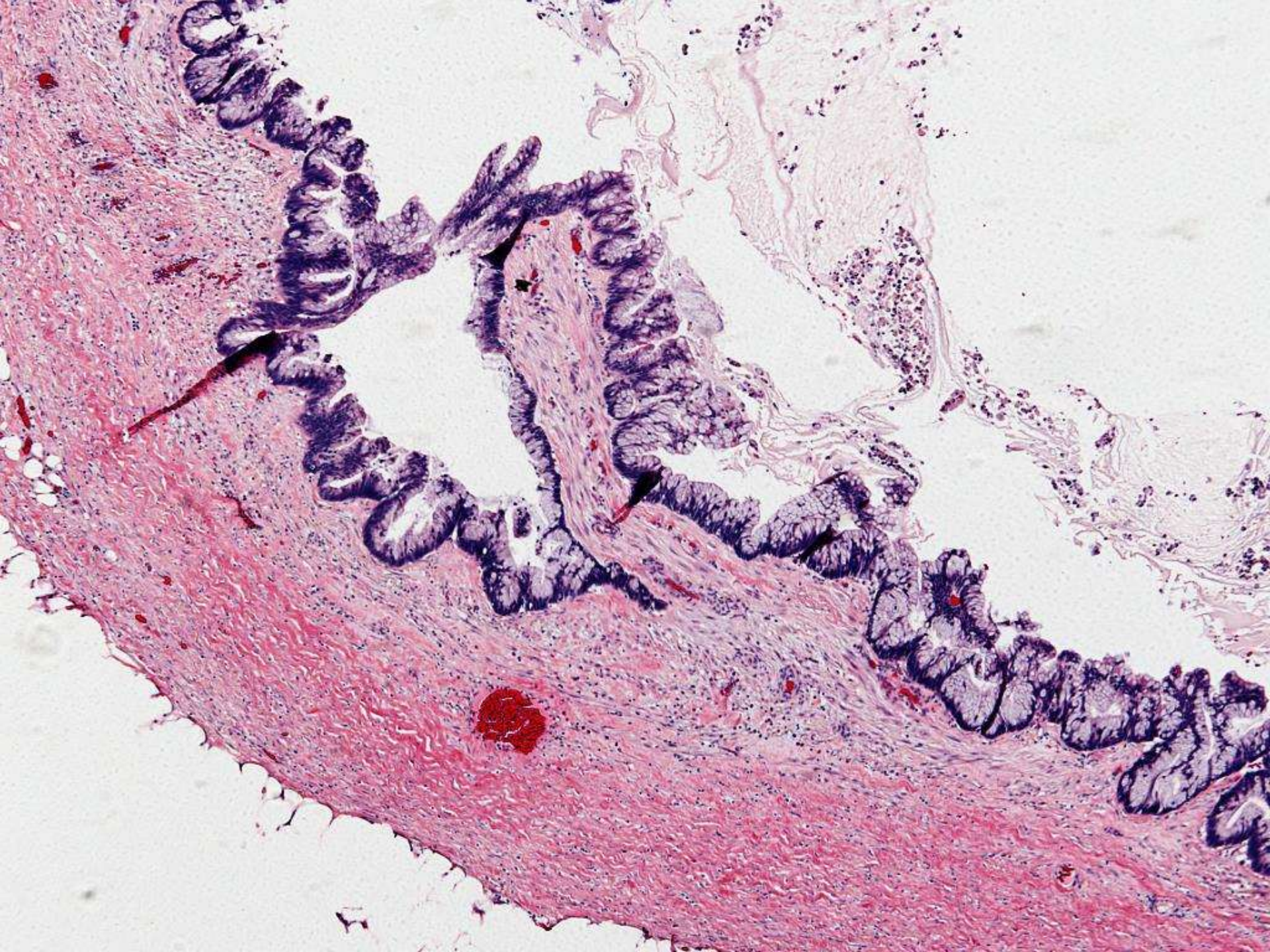


**SB 6133**

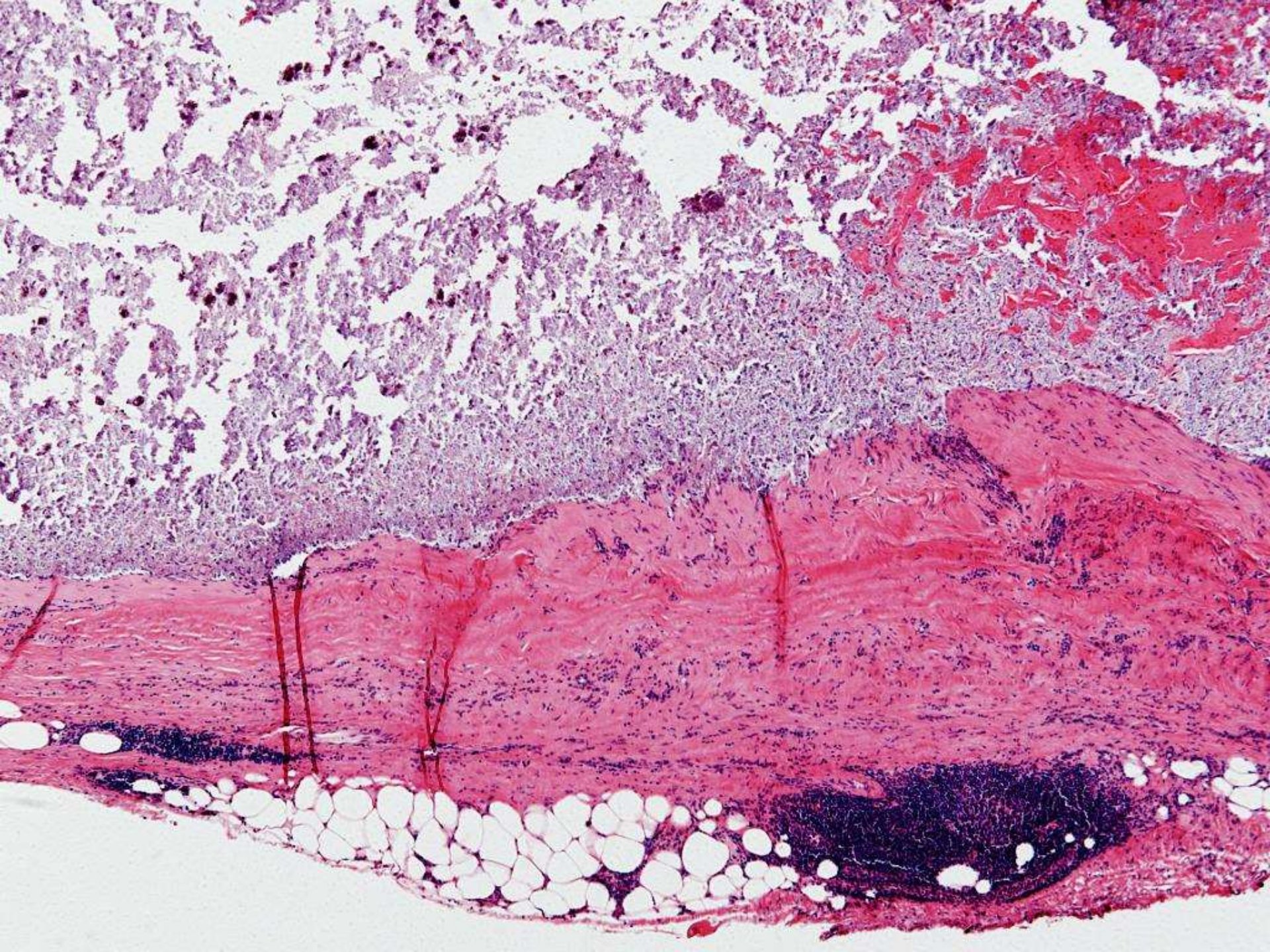
**Alana Shain/Sunny Kao; Stanford**

43-year-old male with hematuria. CT shows  
a partially calcified cystic urachal lesion.

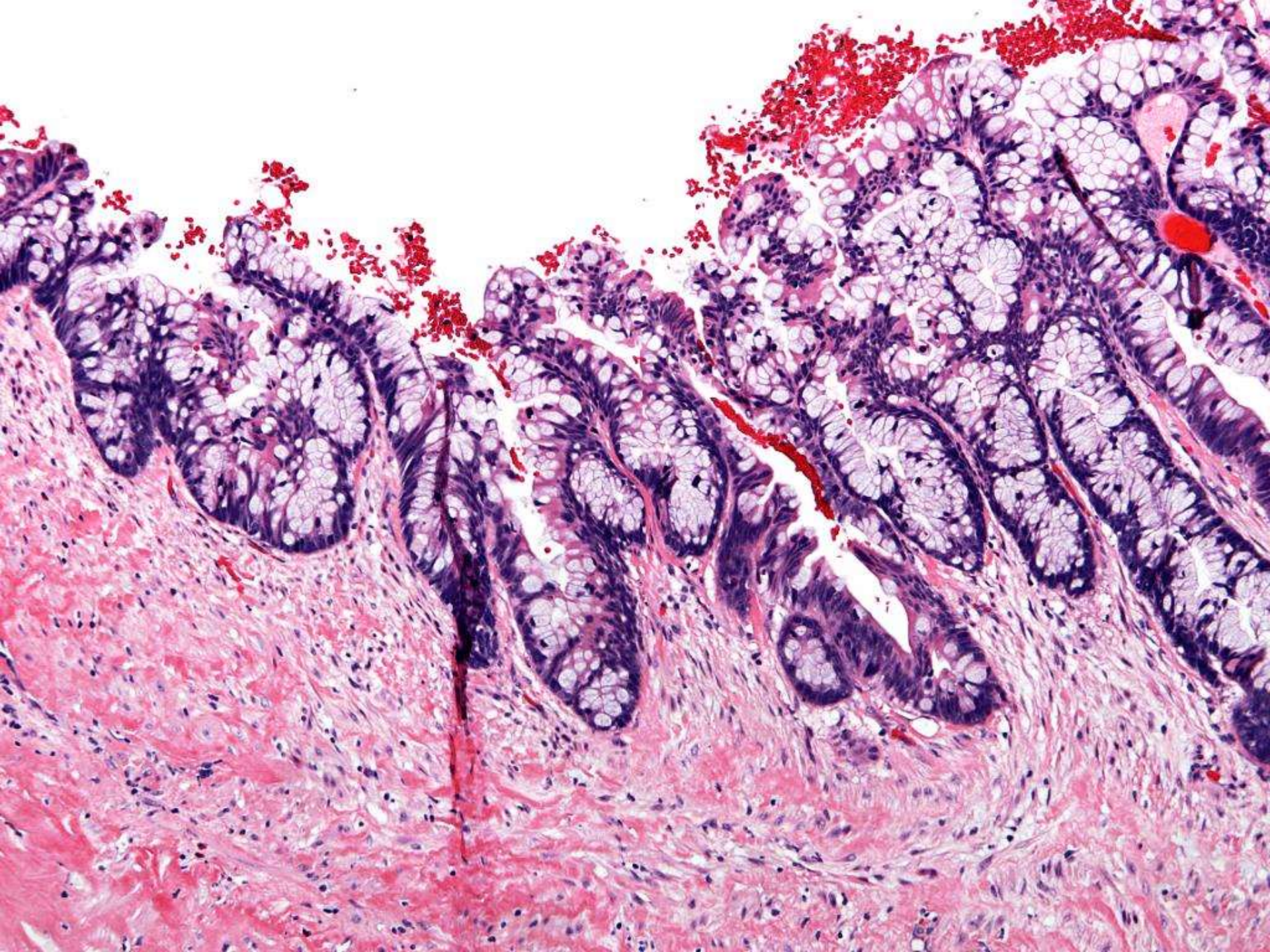




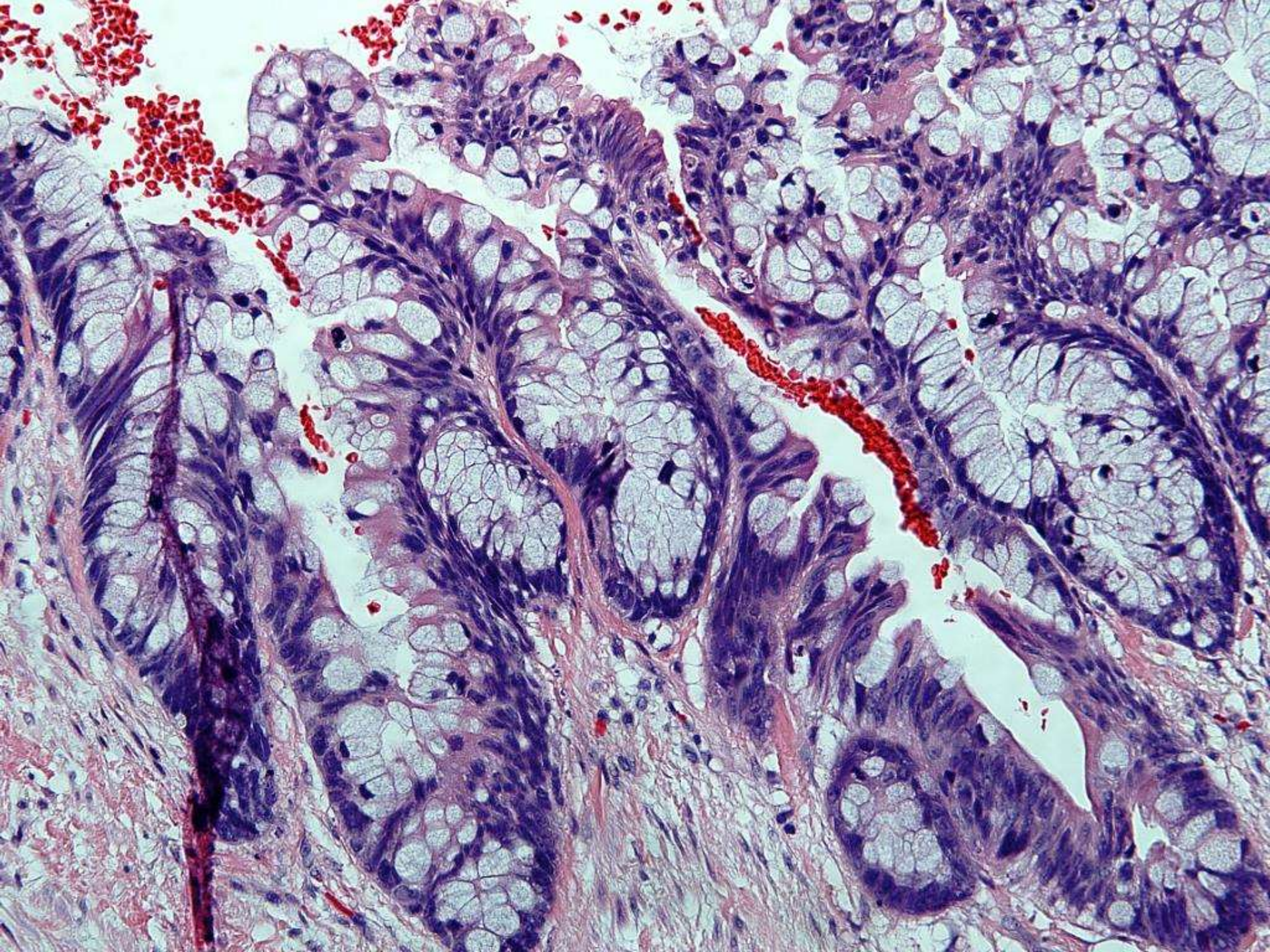




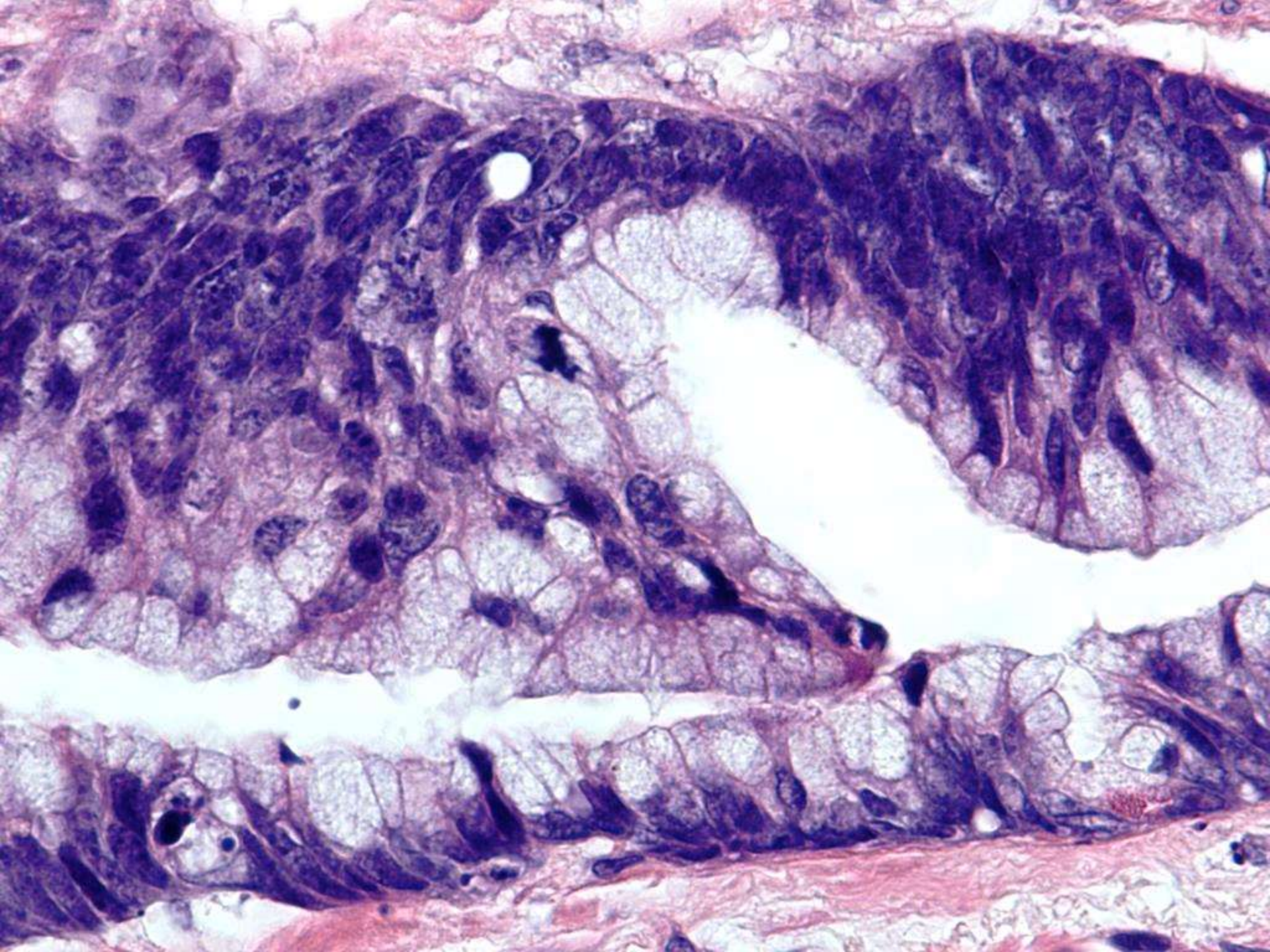










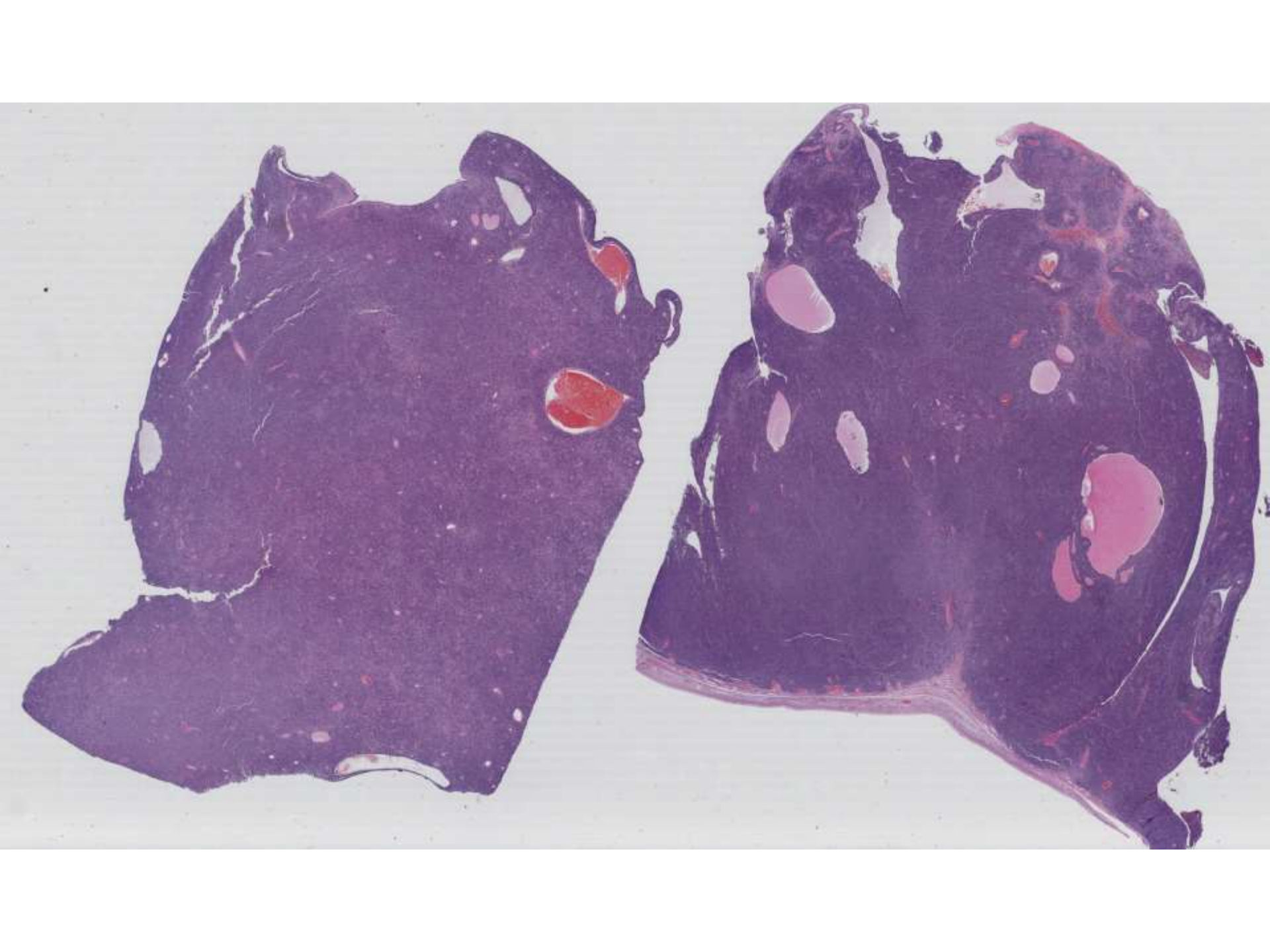




**SB 6132**  
**(scanned slide available)**

**Charles Lombard; El Camino Hospital**

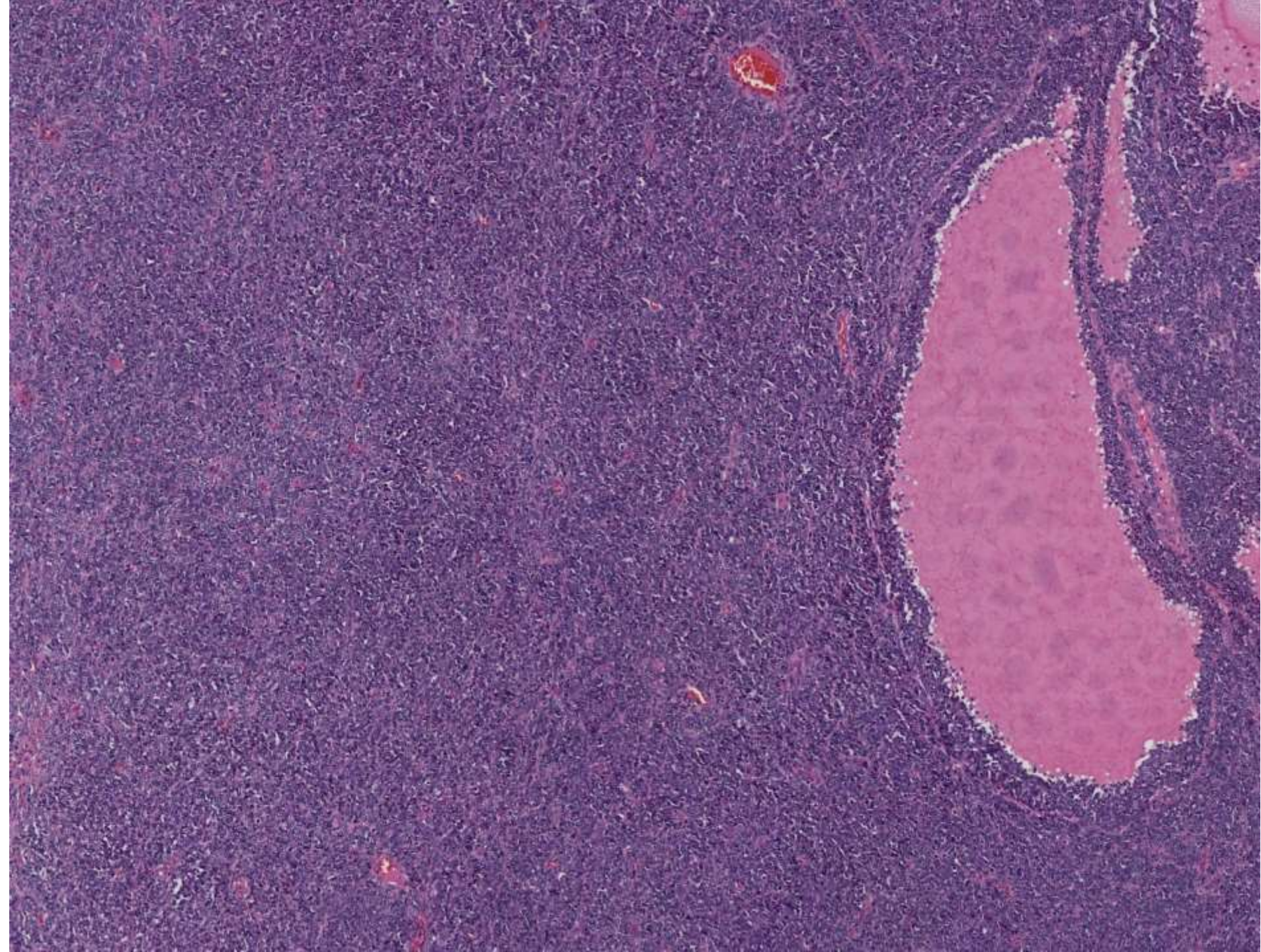
**27-year-old female with 13cm  
ovarian mass.**



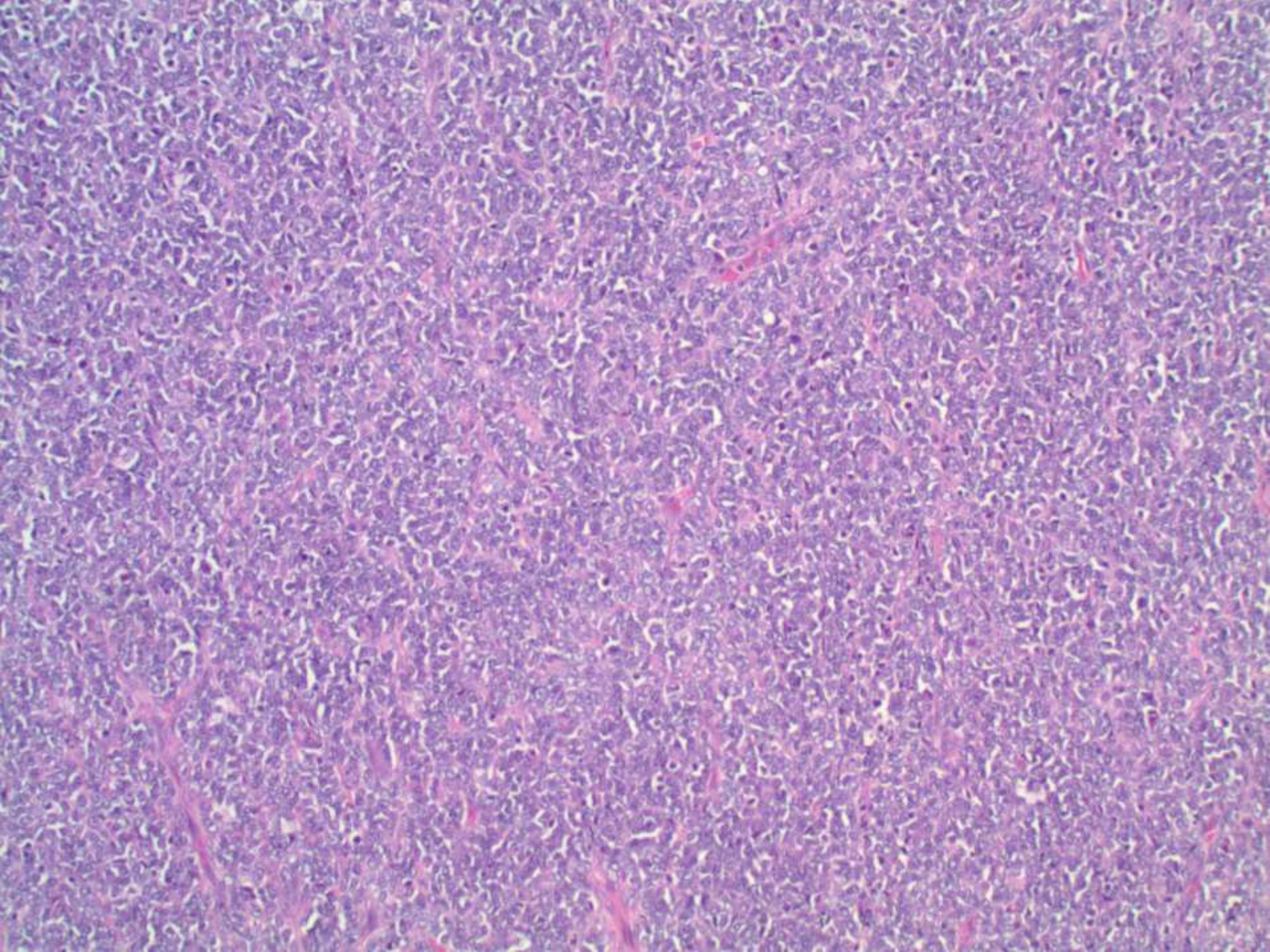




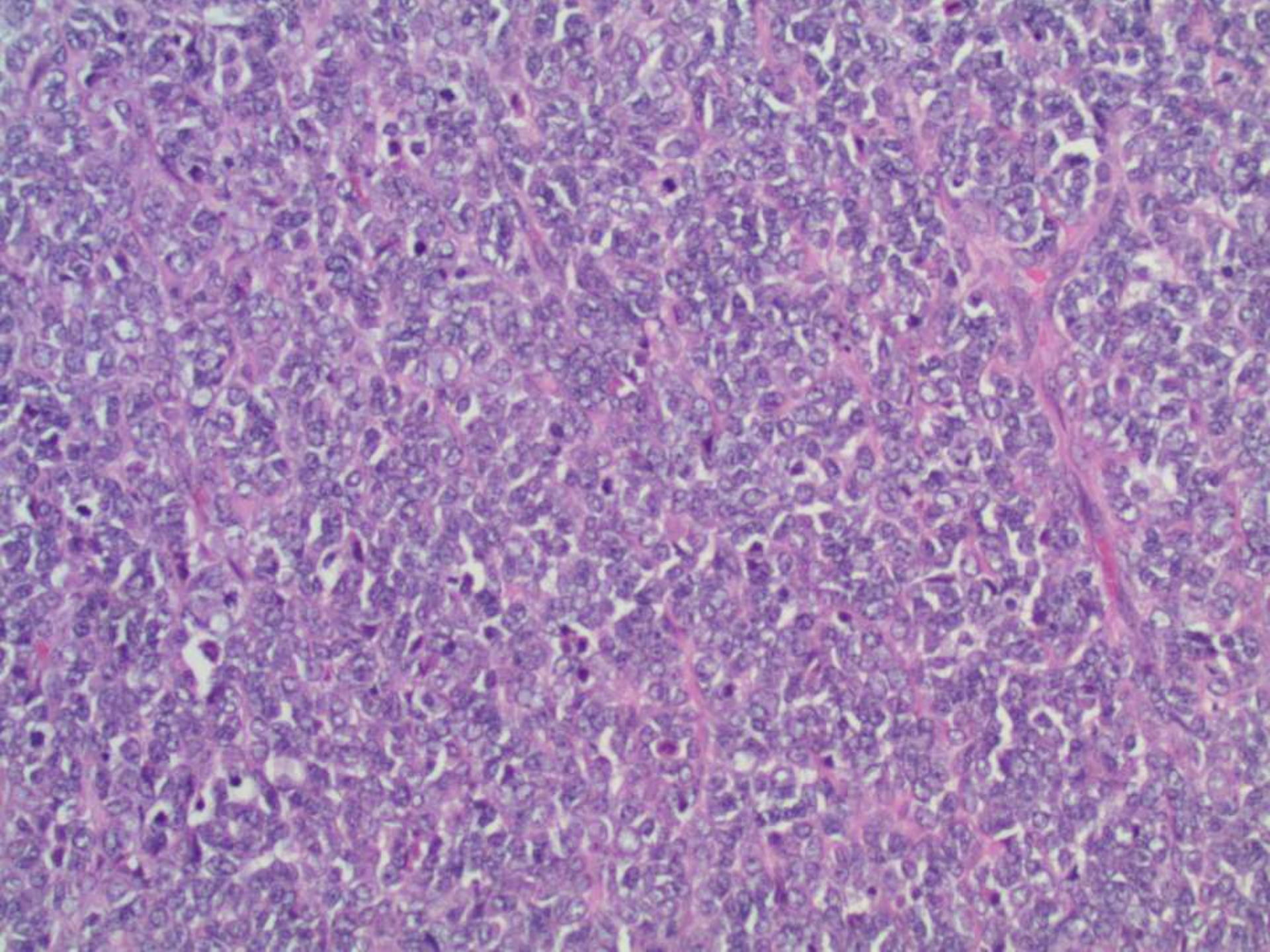




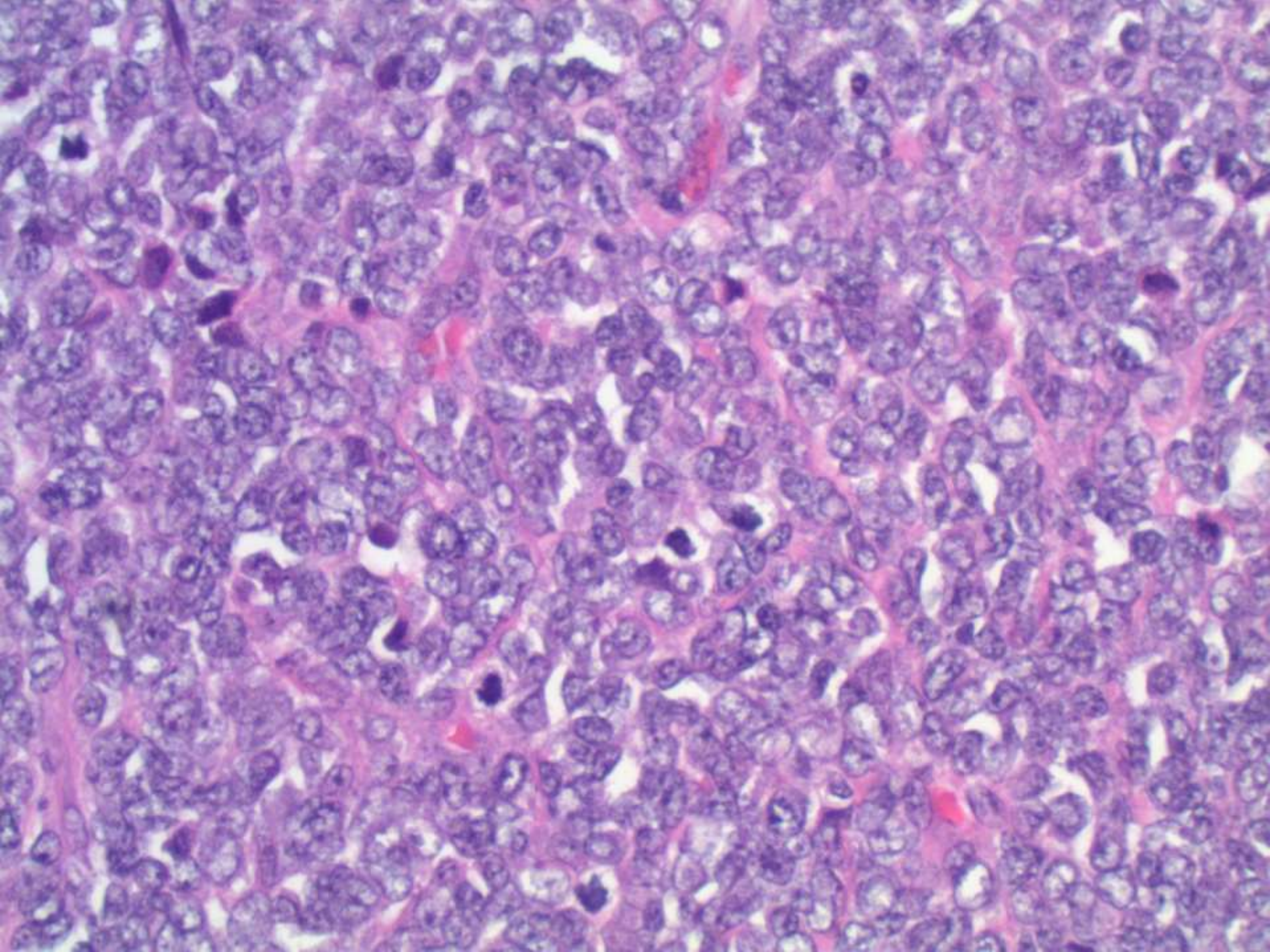










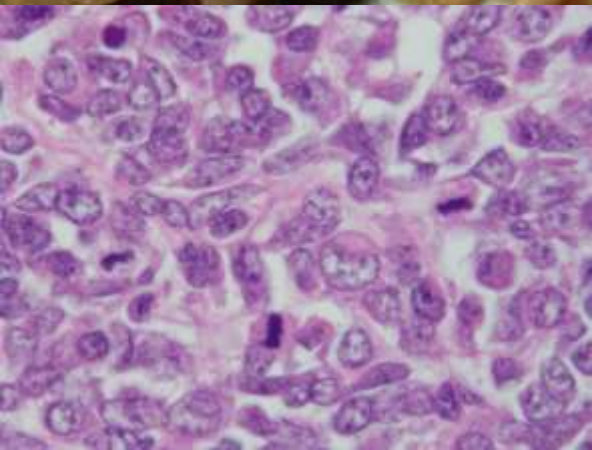
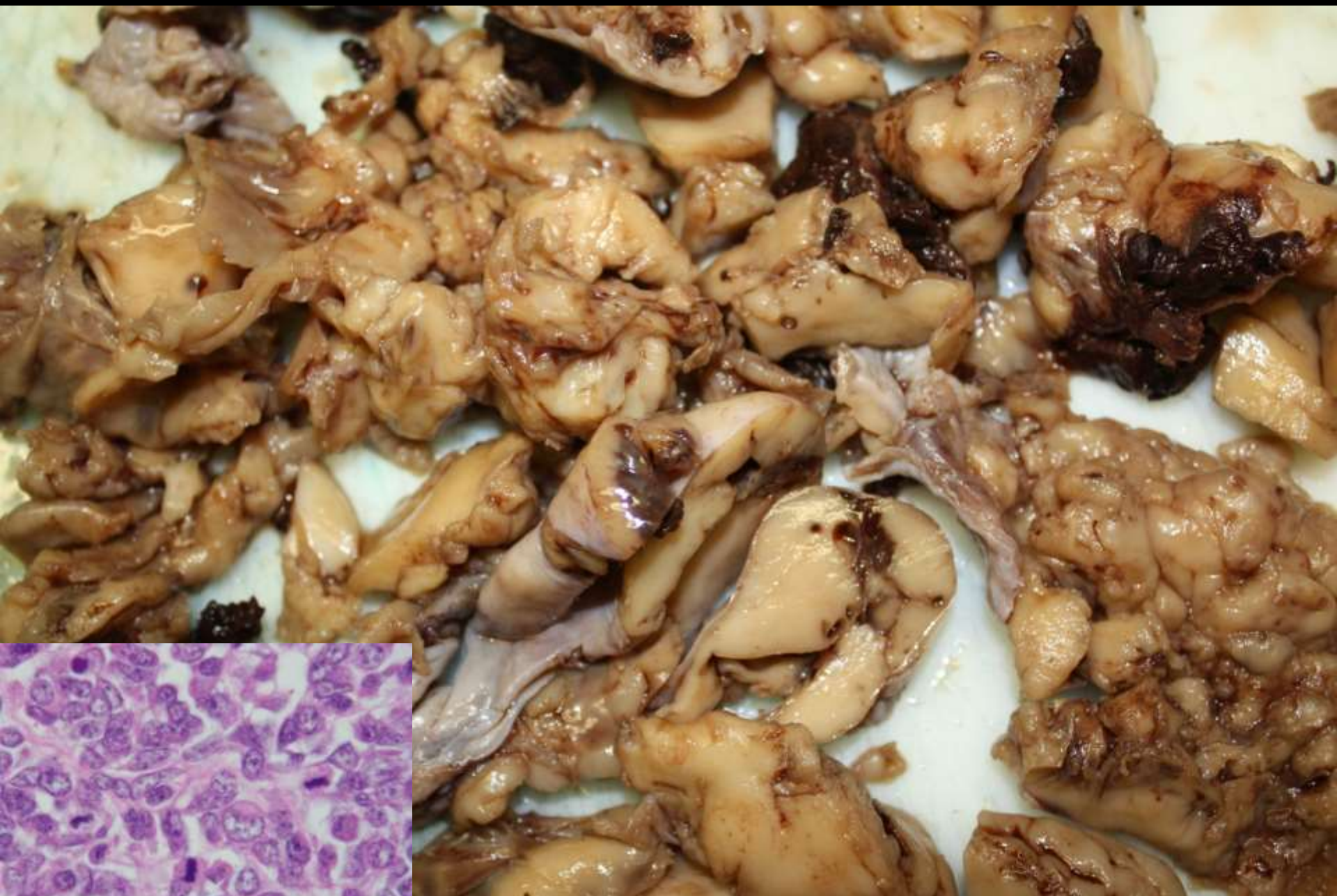




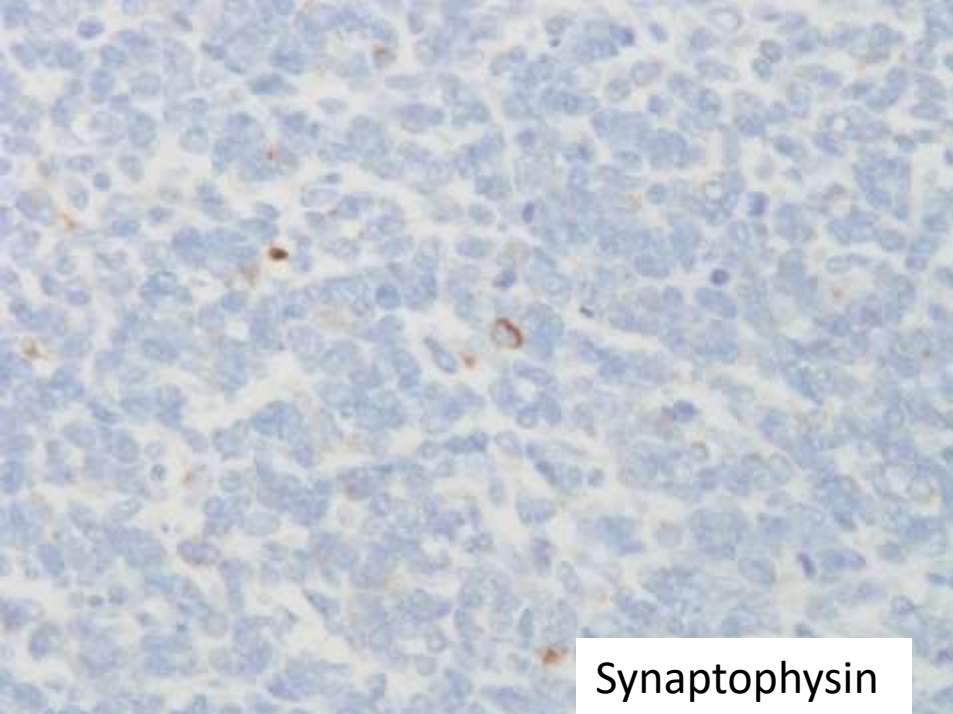
# 27 yo F with Ovarian mass and Hypercalcemia

Small cell carcinoma of the ovary  
Hypercalcemic type

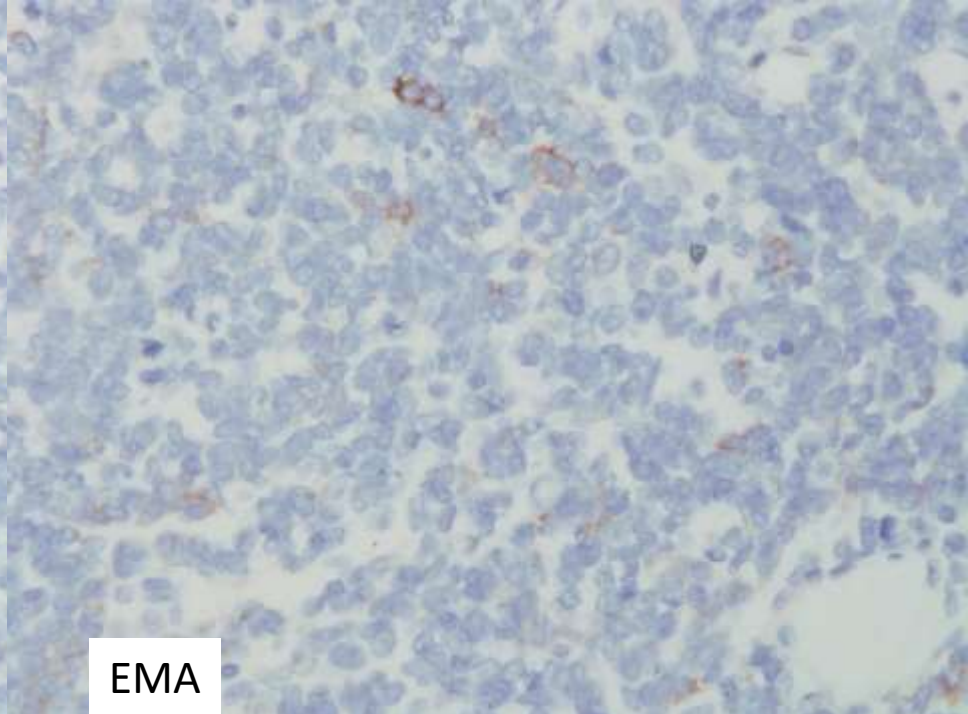




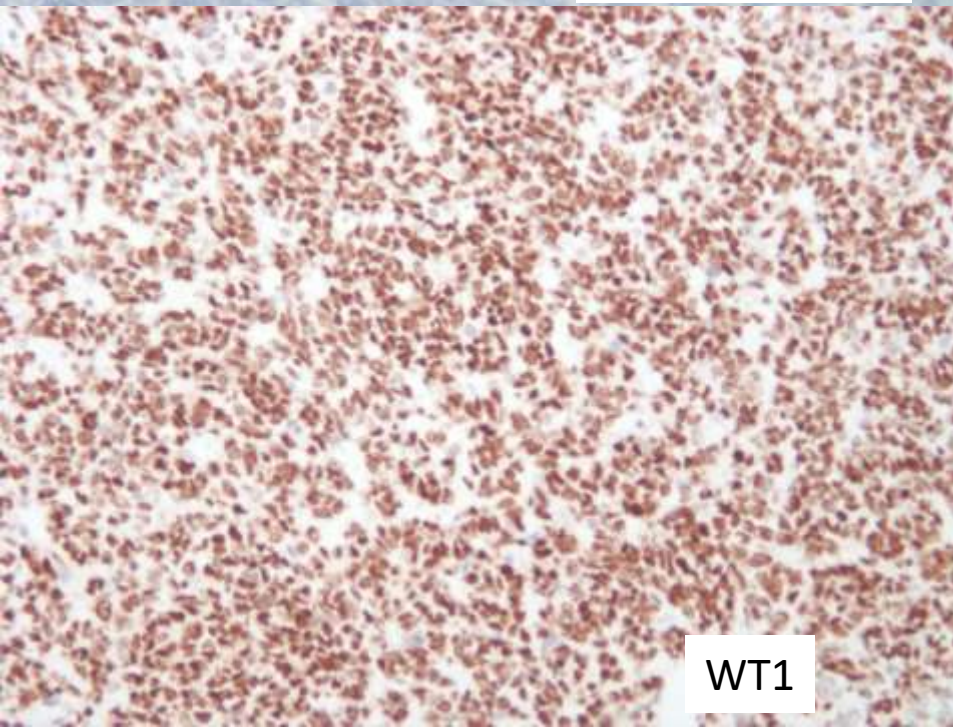




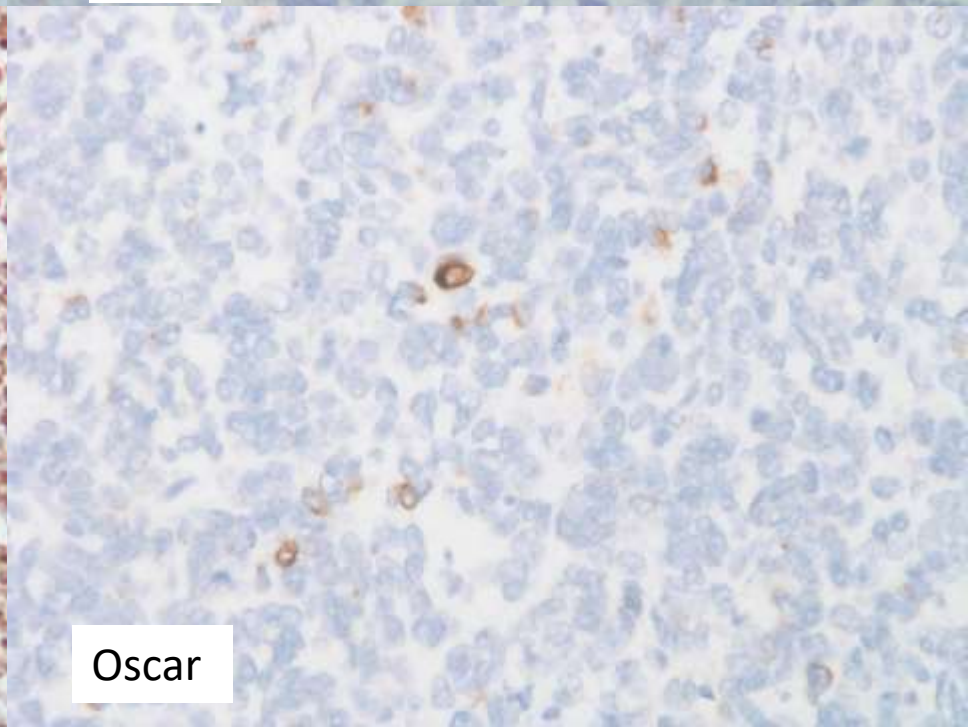
Synaptophysin



EMA



WT1



Oscar



# SCCO-HT

## Clinical features

- Age: 23 (1-71)
- Presentation
  - Pain 68%
  - Mass 55%
  - Hypercalcemia 68%
- Familial cases
  - Autosomal dominant pattern
  - SMARCA4 mutation



# SCCO-HT

## Histology

- Diffuse sheets of small undifferentiated cells
  - Nests, cords, clusters, follicle like spaces
  - Spindled areas, clear cell areas, hyaline globules
  - Rhabdoid features can be seen
  - Can have admixed large cell areas with enlarged nucleoli (Large cell variant– these predominate)
  - High mitotic rate
- Misdiagnosis rate in published series
  - 19-30%

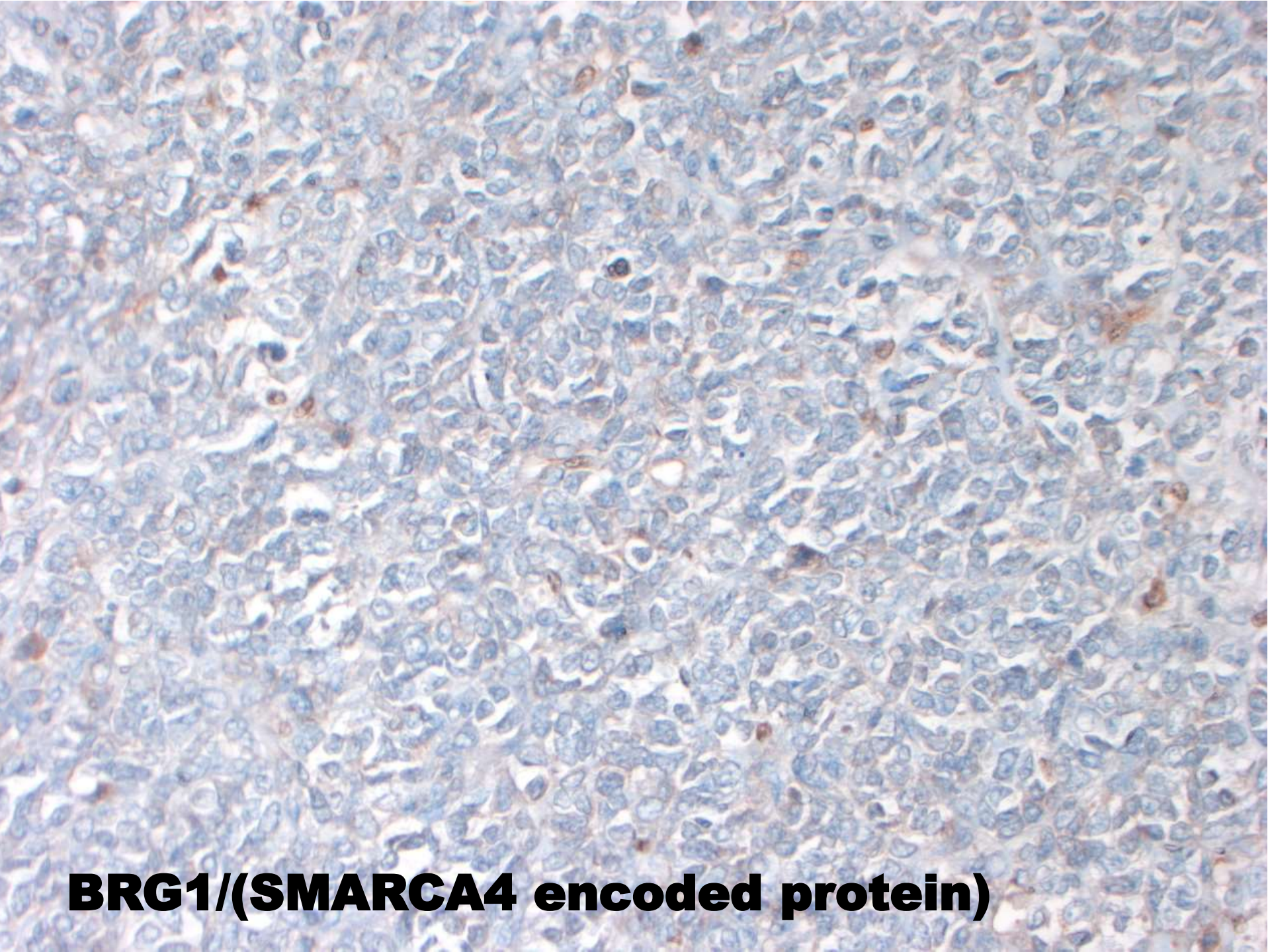


# SCCO-HT

## “Polyimmunophenotypic”

- WT1 95%
- P53 85%
- CD10: 95%
- Keratin variable 60-90%
- EMA variable 30-90%
- Calretinin variable 70%
- Synapto variable 15%
- But wait... there is a new kid on the block!!!





**BRG1/(SMARCA4 encoded protein)**



# IHC for BRG1 protein

- BRG1 is encoded by SMARCA4
  - Mutation leads to loss of BRG1 expression by IHC
- Although some other tumors also lose this
  - Ovarian clear cell ca 4%; ESS 8%; EMCA rare
  - Most tumors in DDX lack this expression
    - SCC ovary pulmonary type, GCT, S-L tumor, Germ cell tumors, rhabdomyosarc, DSRCT, Ewings sarcoma, synovial sarcoma, lymphoma, melanoma
- In this case patient had loss of BRG1 and BRM by IHC
  - BRM loss thought to be 2° to epigenetic silencing not mutation to SMARCB2
- 92% of cases have SMARCA4 mutation; 86% have loss of BRG1 by IHC
  - One study: 43% of SCCO-HT mutations were GERMLINE mutations



# SCCO-HT

## What kind of tumor is this???

- ?? Carcinoma
  - EM: desmosomes, tight junctions, ...
  - IHC: some keratin, EMA, NE markers
- ?? Germ cell origin
  - EM: X-cellular BM-like material
  - LM: PAS+ globules
- ?? Sex cord stromal tumor
  - Follicle-like structures, calretinin positivity,...
- **Variant of malignant rhabdoid tumor**
  - Histologic similarity to
    - Renal rhabdoid tumor
    - CNS atypical teratoid rhabdoid tumor
  - Mutations in SW1/SNF pathway
    - SMARCB2 in rhabdoid tumors
    - SMARCA4 in SCCO-HT



# SCCO-HT

## Familial Cases

- Germline SMARCA4 mutations found in as many as 43% of cases of SCCO-HT.
  - But incidence of familial cases is low
    - Tumor of young females (esp germline cases)
    - Father may carry “silent” mutation
  - Recommend testing in all for germline mutation
    - Female Carriers are at risk for SCCO-HT
      - Prophylactic Oophorectomy??
      - Close follow up with U/S screening??
    - M/F children(< 4-5 y) may be at risk for RT's



# SCCO-HT

## Treatment

- Most are treated with primary surgery followed by chemotherapy
- 5 y survival rate for stage I disease: 50%
- Debatable role for RT
- REFERENCE:
- Hoang and Soslow: “SCCOHT: Past, Present, and Future.” AJSP: Reviews and Reports 21(4): 181-188; 2016.



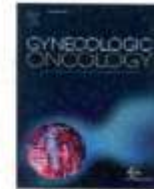
# Follow Up



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## Small cell carcinoma of the ovary-hypercalcemic type (SCCOHT): A review of 47 cases☆



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<sup>b</sup> Department of Gynecologic Oncology and Reproductive Medicine, The University of Texas MD Anderson Cancer Center, Houston, TX, USA

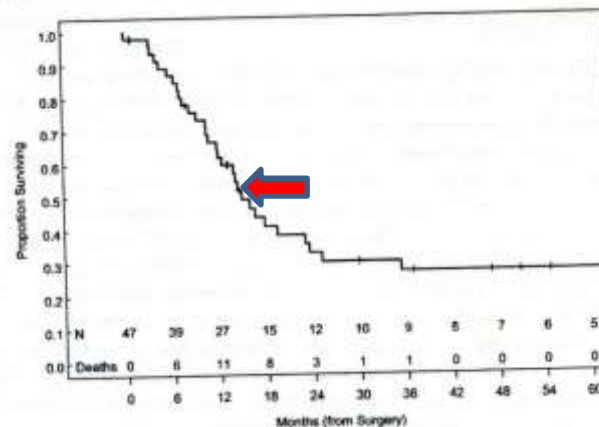
<sup>c</sup> Department of Biostatistics, The University of Texas MD Anderson Cancer Center, Houston, TX, USA

<sup>d</sup> Department of Radiation Oncology, The University of Texas MD Anderson Cancer Center, Houston, TX, USA

<sup>e</sup> Department of Pathology, The University of Texas MD Anderson Cancer Center, Houston, TX, USA

<sup>f</sup> Division of Gynecologic Oncology, Universidade Federal de São Paulo, São Paulo, Brazil

**Patient received VPCBAE Chemotherapy and is currently NED at 13 months.**





**SB 6133**

**Mucinous Cystic Tumor of  
Low Malignant Potential**

Case courtesy Dr. Peter Ames,  
Renown Health, Reno, NV

Alana Shain/Sunny Kao  
Stanford

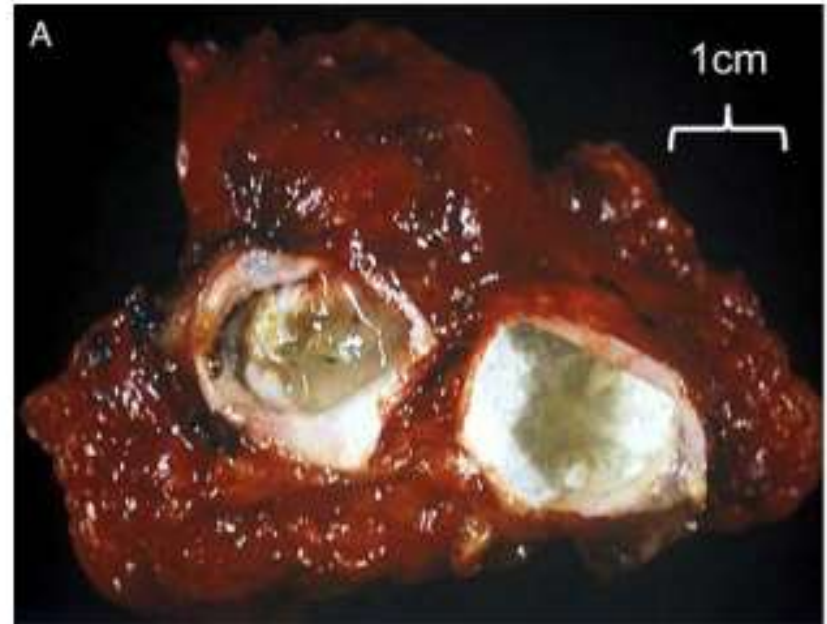
# Urachal Epithelial Neoplasms

- Glandular (the majority)
  - Noncystic invasive adenocarcinoma, 83%
  - Cystic tumors, 17%
- Non-glandular (rare!)
  - urothelial, squamous cell, neuroendocrine, mixed type



# Mucinous cystic tumors

- **Classification analogous to ovarian tumors**
- Cystic lumina (usually unilocular), copious mucin
- Presentation incidental or related to mass lesion (vs. hematuria for noncystic invasive adenocarcinoma)
- Reports of pseudomyxoma peritonei



Immunoprofile similar to primary ovarian tumors

CK7+ (60%), CK20+, CDX2+

Nuclear beta catenin negative, ER/PR negative

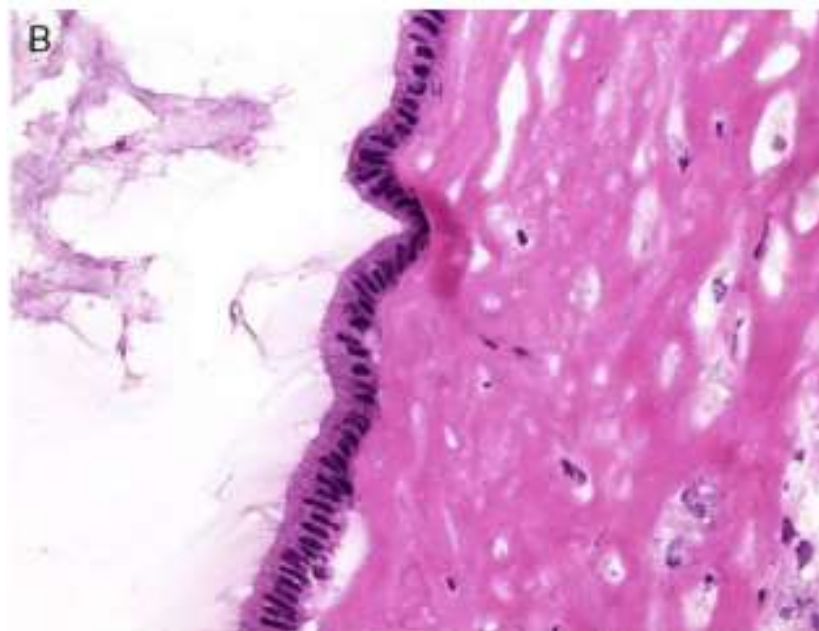
# Mucinous Cystic Tumors

Mucinous  
Cystadenoma

MCT of LMP

MCT of LMP with  
Intraepithelial  
carcinoma  
( $<2$  mm,  $<5\%$   
overall)

Mucinous  
cystadenocarcinoma  
(microinvasion or  
frank invasion)





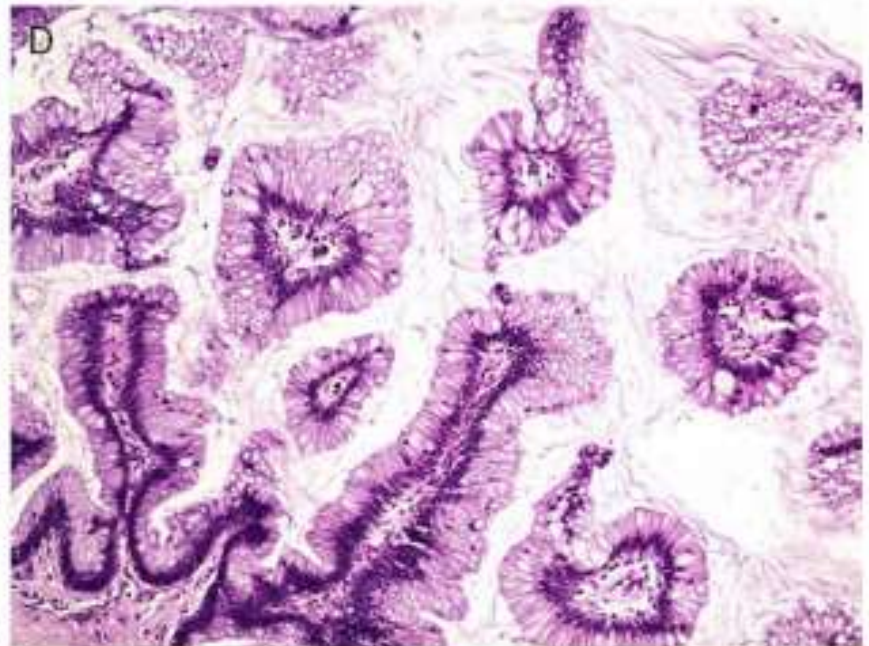
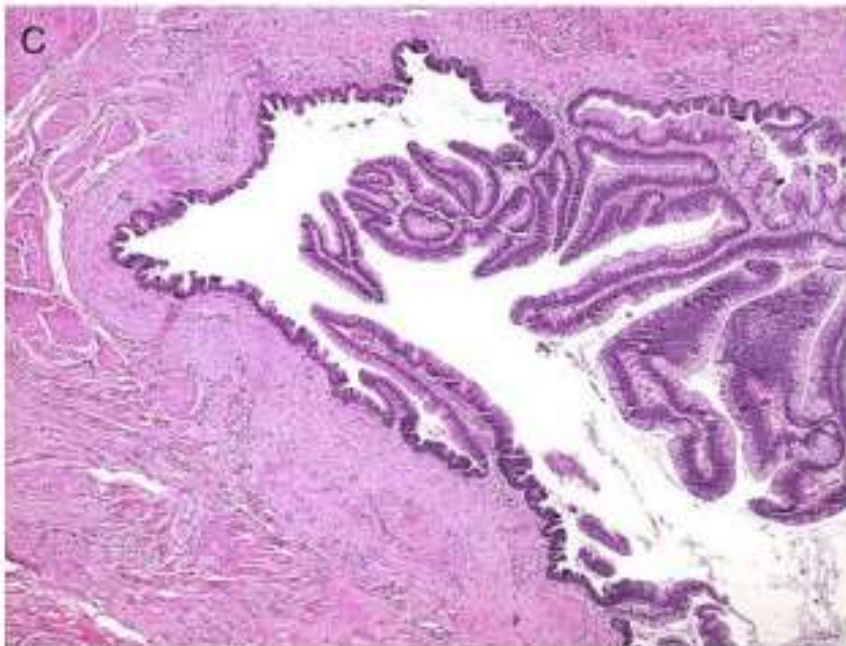
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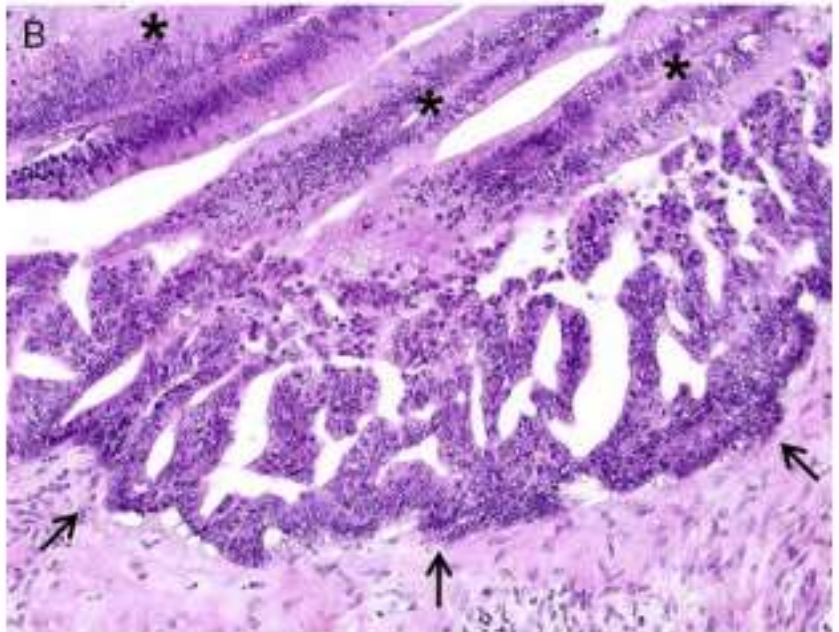
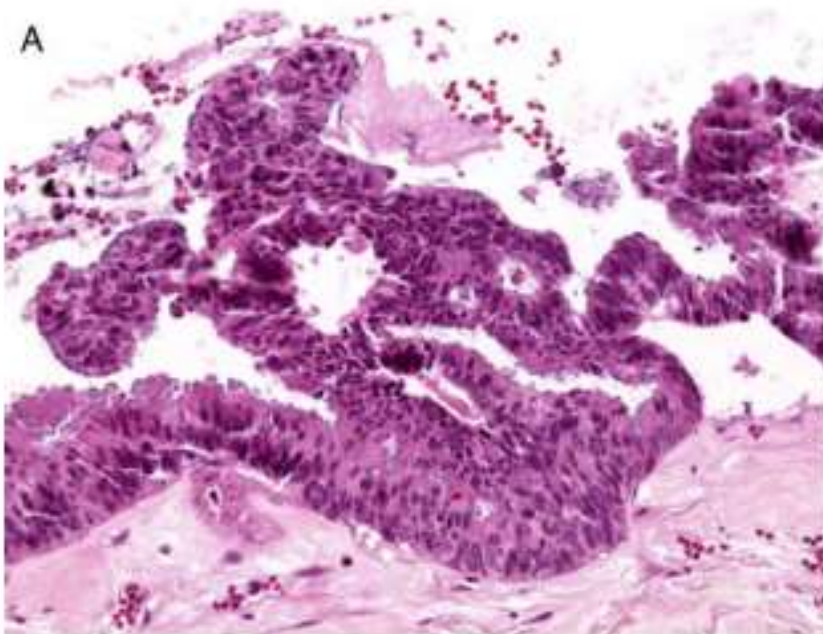
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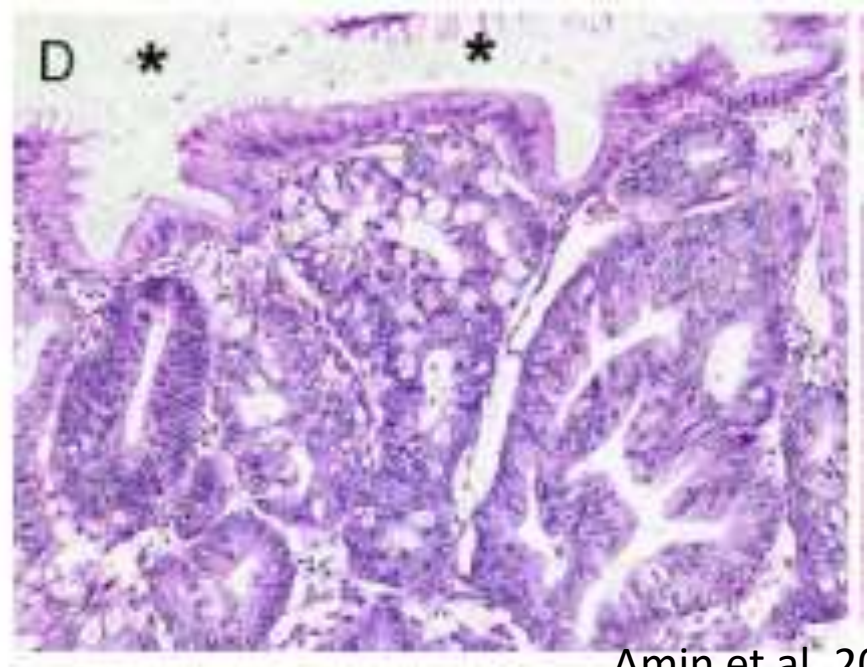
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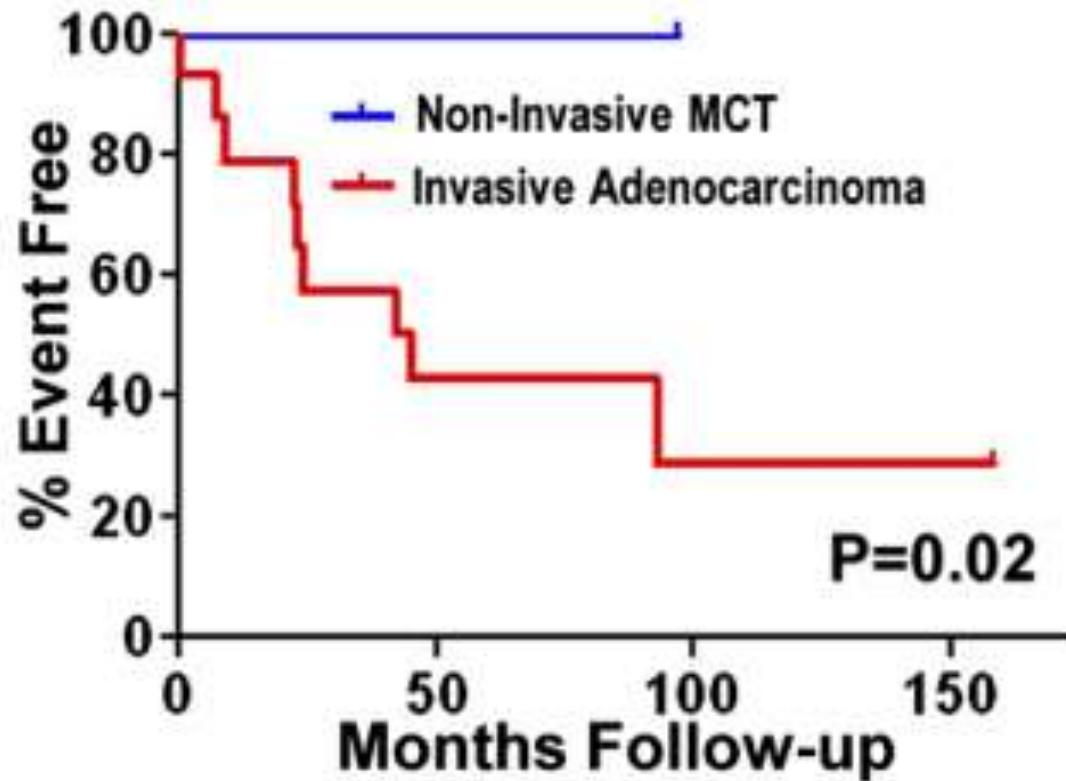
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overall)

Mucinous  
cystadenocarcinoma  
(microinvasion or  
frank invasion)



# Outcome Urachal Epithelial Neoplasms



Survival of **noninvasive mucinous cystic tumors** is **significantly better** than **noncystic adenocarcinomas**.



# Is it urachal?

## WHO Criteria for diagnosis of urachal adenocarcinoma (WHO 2016, page 113)

1. Location of the tumor in the bladder dome and/or anterior wall
  2. Epicenter of carcinoma in the bladder wall
  3. Absence of widespread cystitis cystica and/or cystitis glandularis beyond the dome or anterior wall
  4. Absence of known primary elsewhere
- Urachal vs. non-urachal adenocarcinoma prognosis controversial, but **tumors confined to urachus and bladder wall have better outcomes**
  - Other prognostic factors: grade, lymph nodes, metastasis
  - Partial cystectomy, urachectomy, umbilectomy

# References

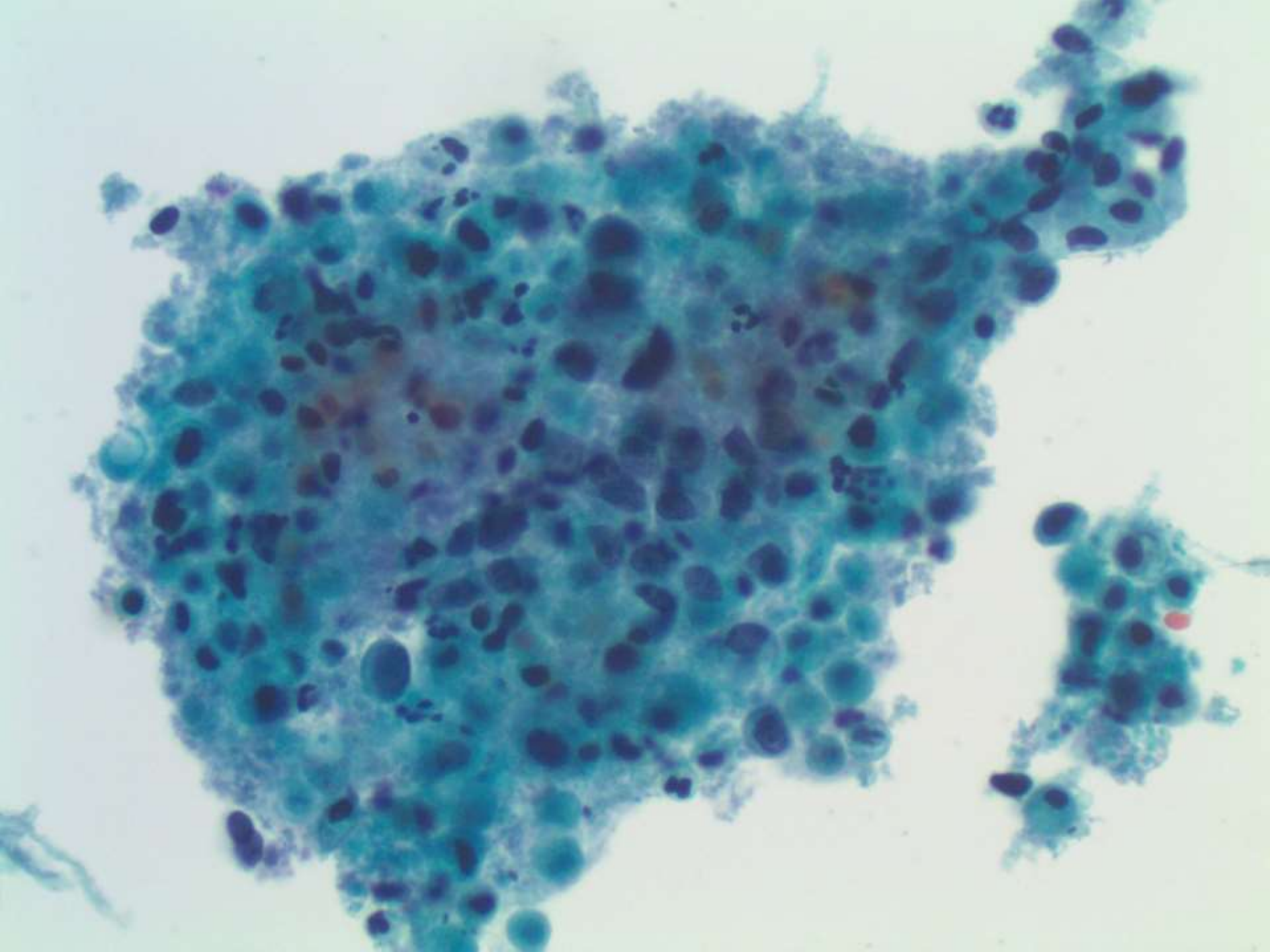
- The WHO, 2016
- Amin M B, Smith S C, Eble J N, Rao P, Choi W W, Tamboli P, Young R H. 2014. Glandular neoplasms of the urachus: a report of 55 cases emphasizing mucinous cystic tumors with proposed classification. The American journal of surgical pathology 38 (8): 1033-1045.
- Paner G P, Lopez Beltran A, Sirohi D, Amin M B. 2016. Updates in the Pathologic Diagnosis and Classification of Epithelial Neoplasms of Urachal Origin. Advances in anatomic pathology 23 (2): 71-83.



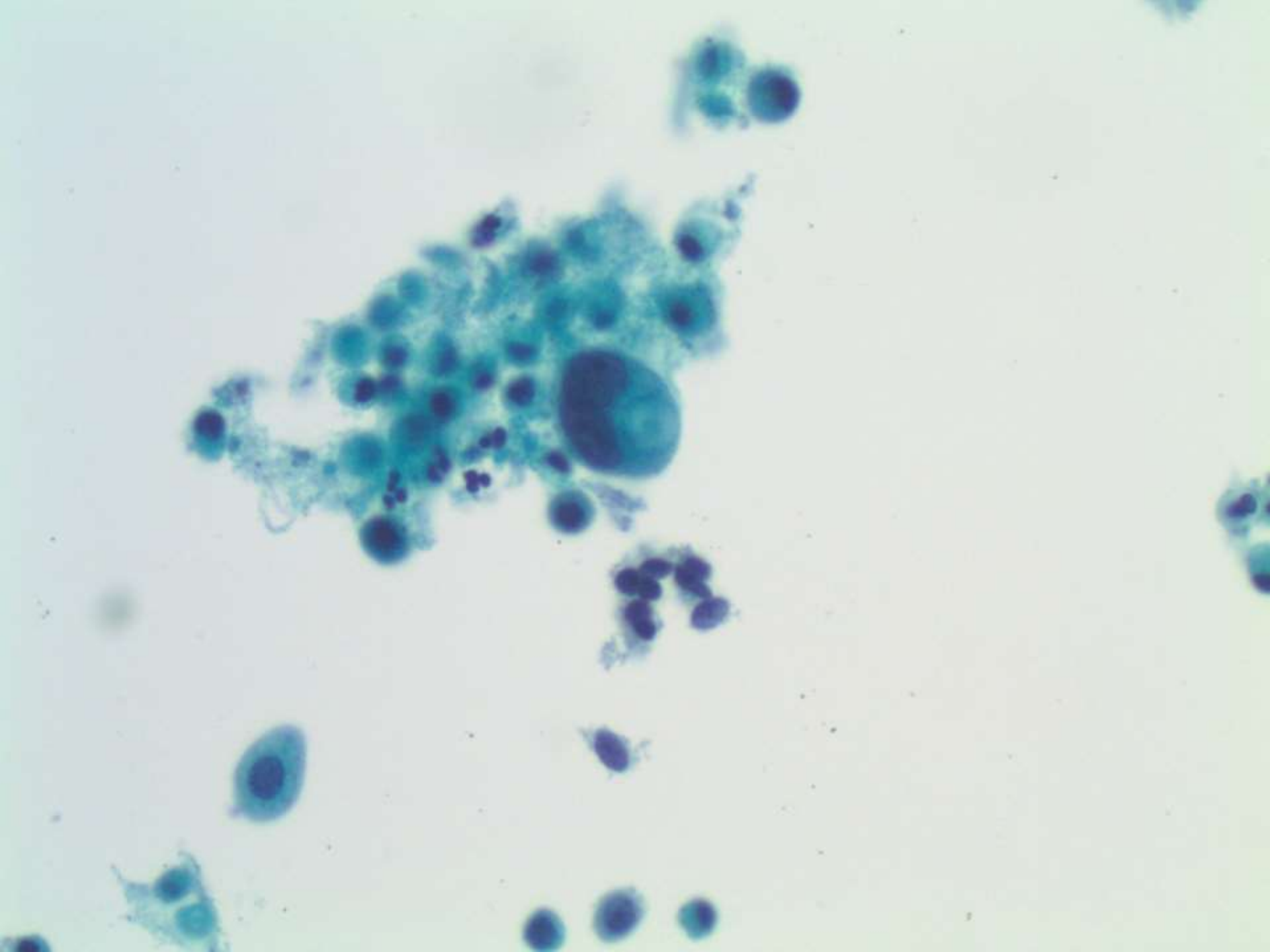
**SB 6134**

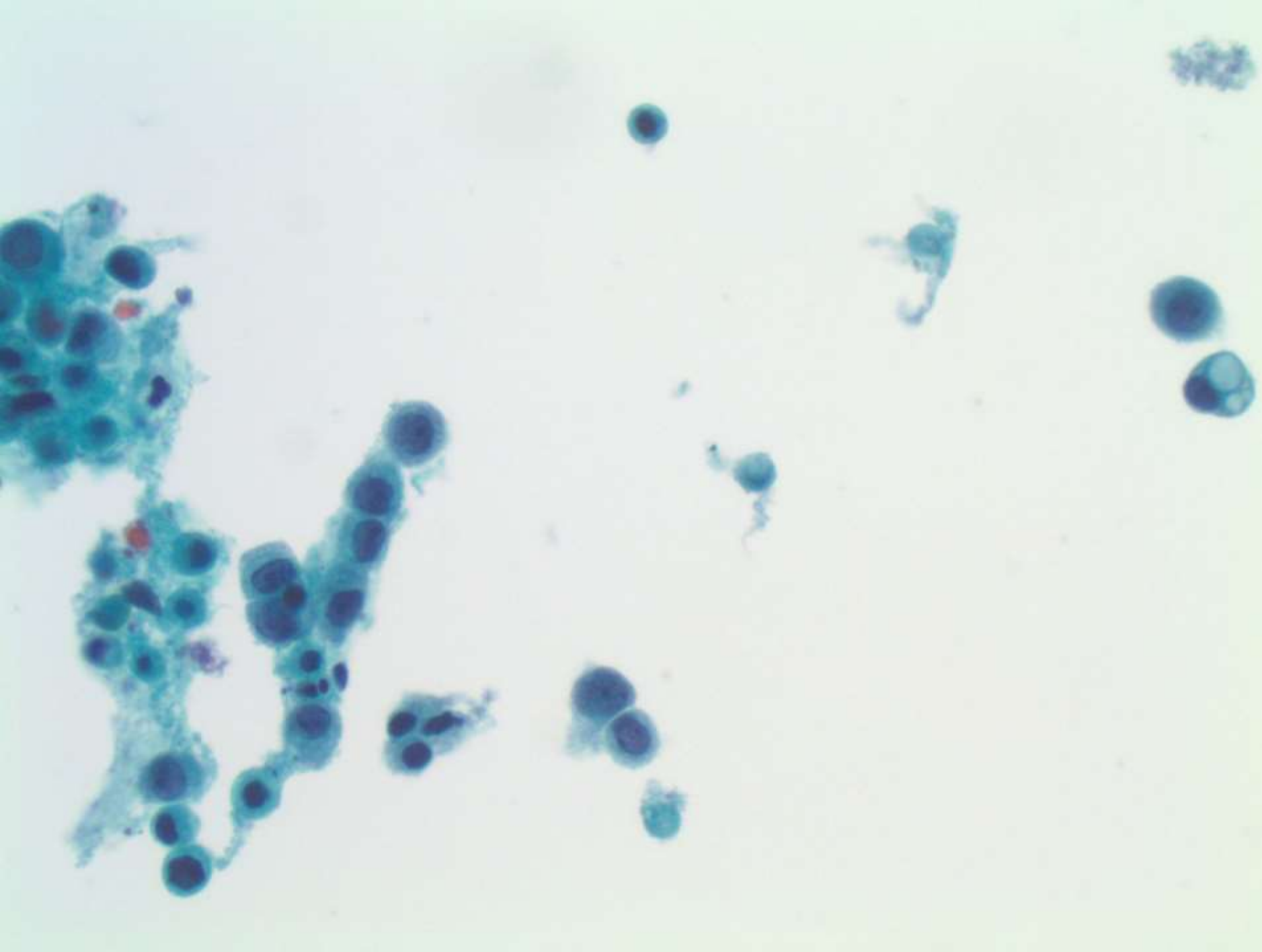
**Mahendra Ranchod; Good Samaritan  
Hospital**

56-year-old female, routine annual Pap  
smear submitted.

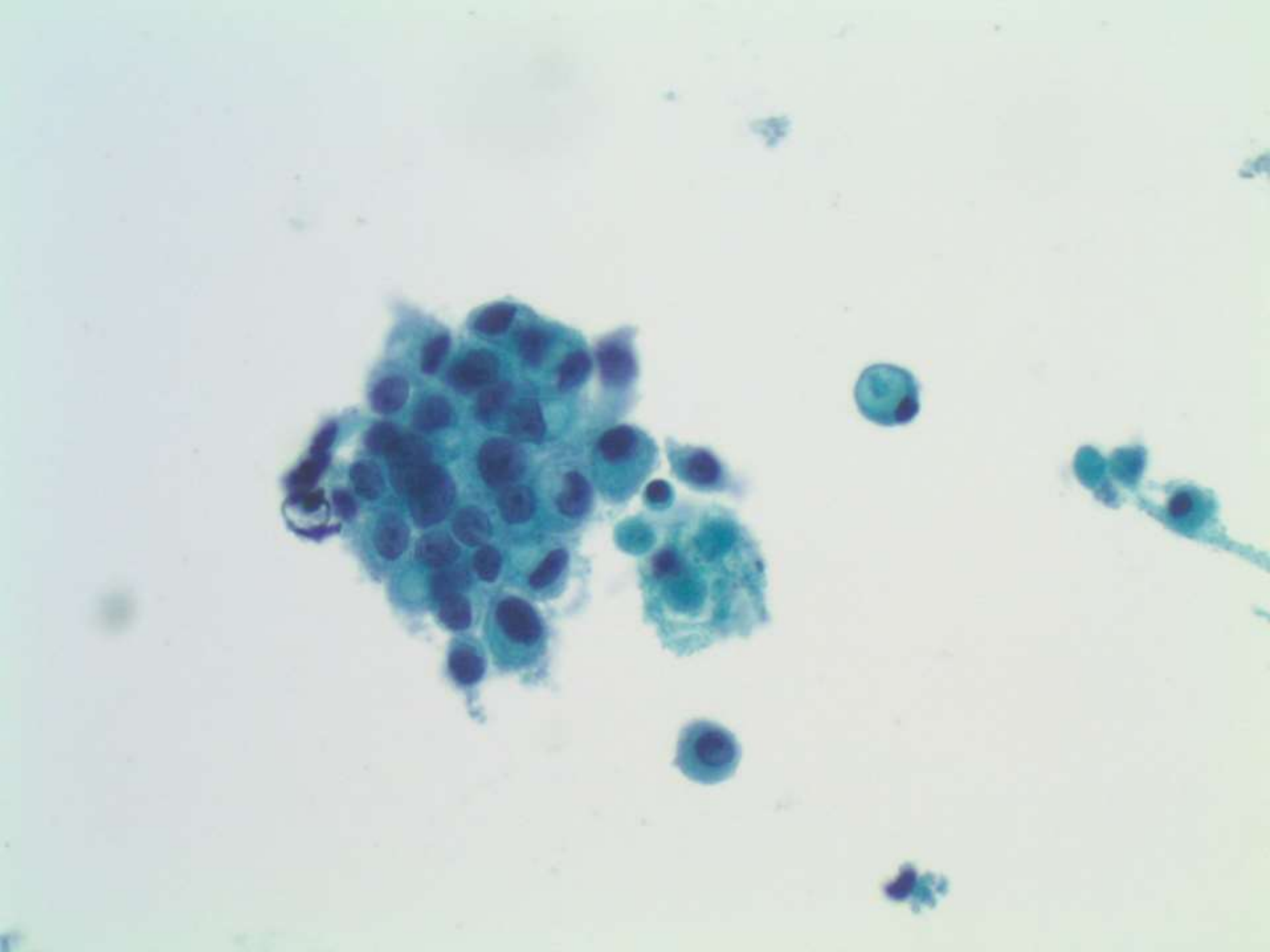


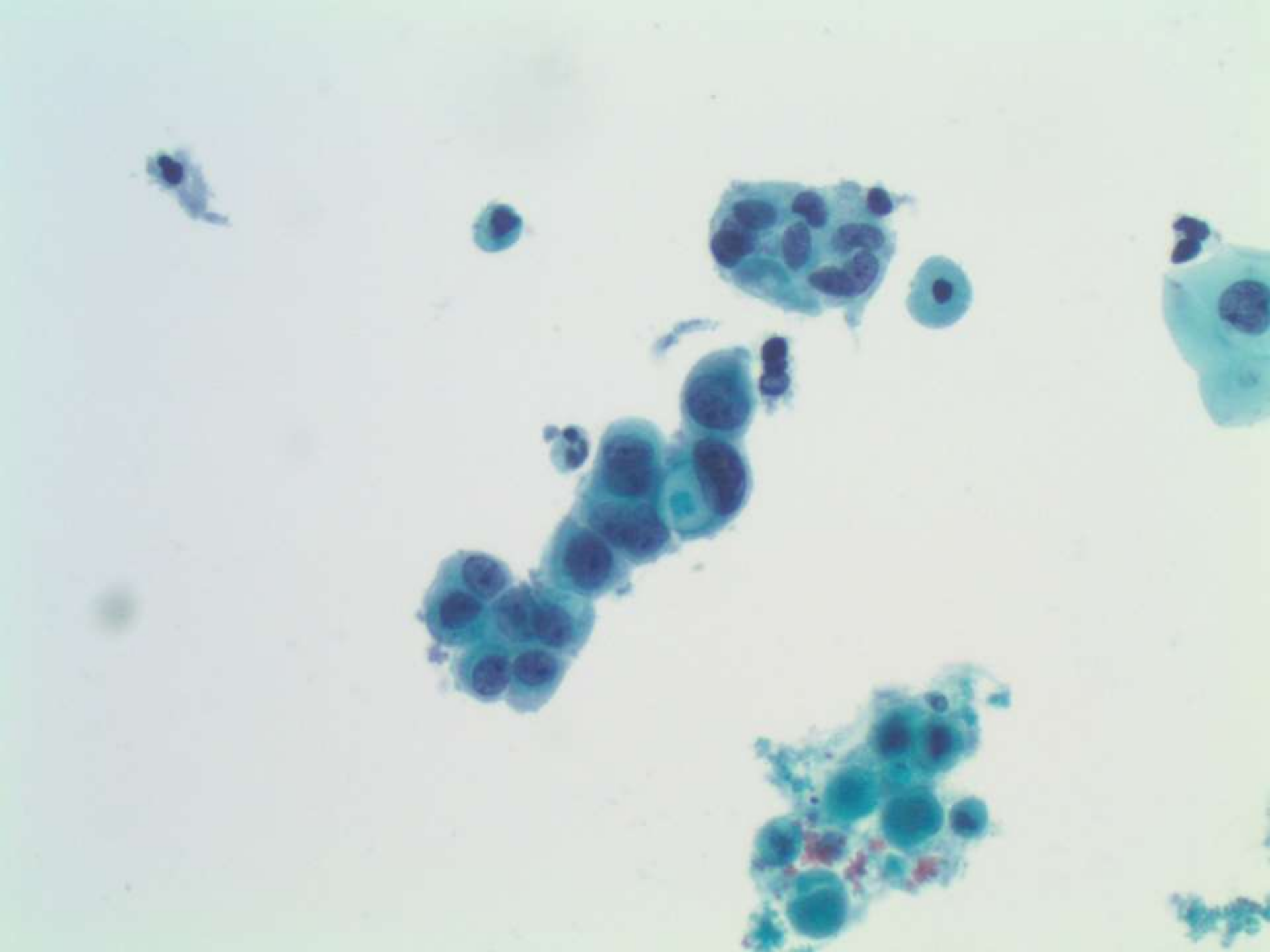














**GATA3**



**GATA3**





# **Metastatic Lobular Carcinoma: Uncommon sites**

**Gastrointestinal Tract**

**Peritoneum**

**Retroperitoneum**

**Meninges (carcinomatous meningitis)**

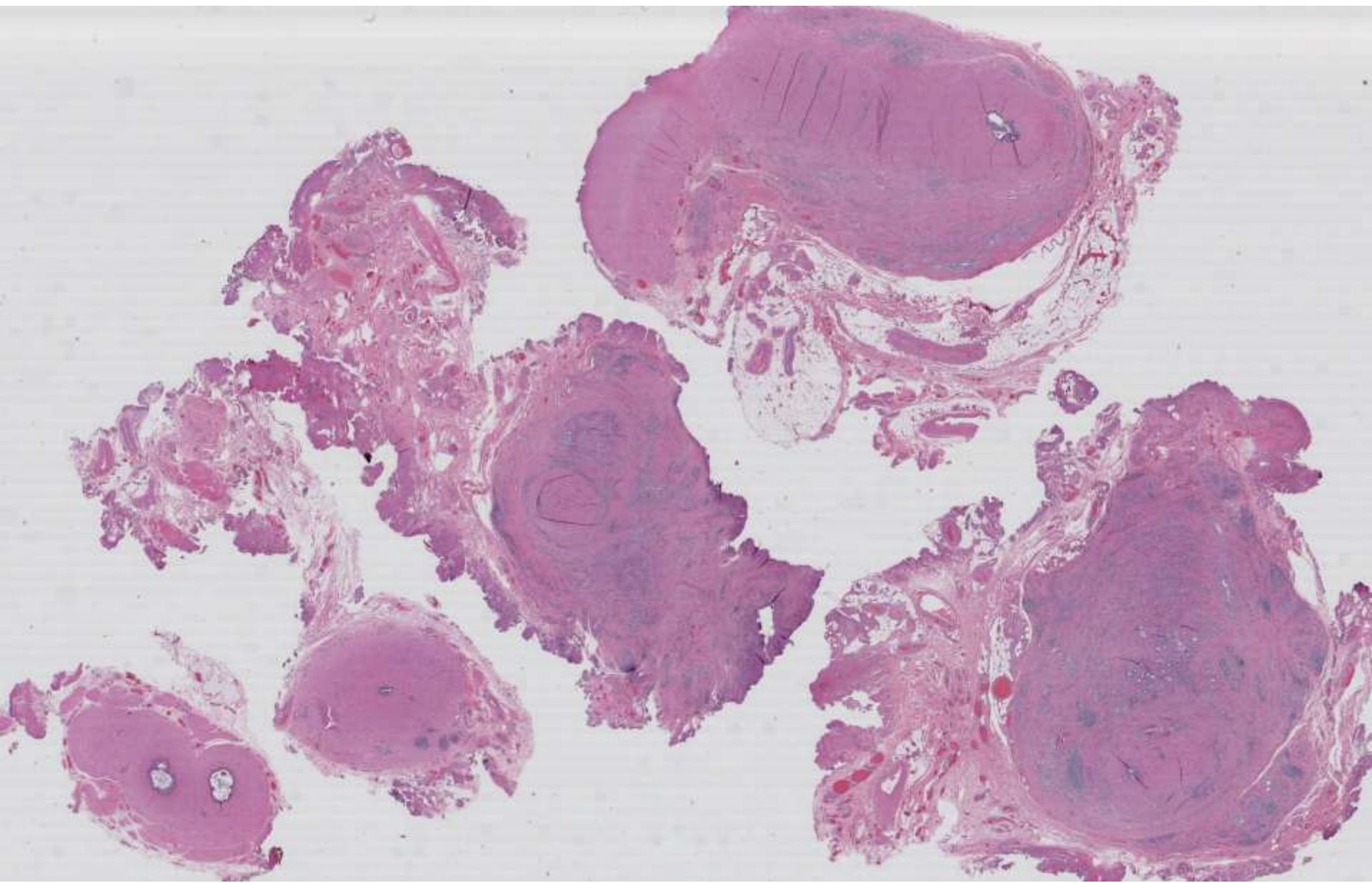
**Uterus**

# SB 6135

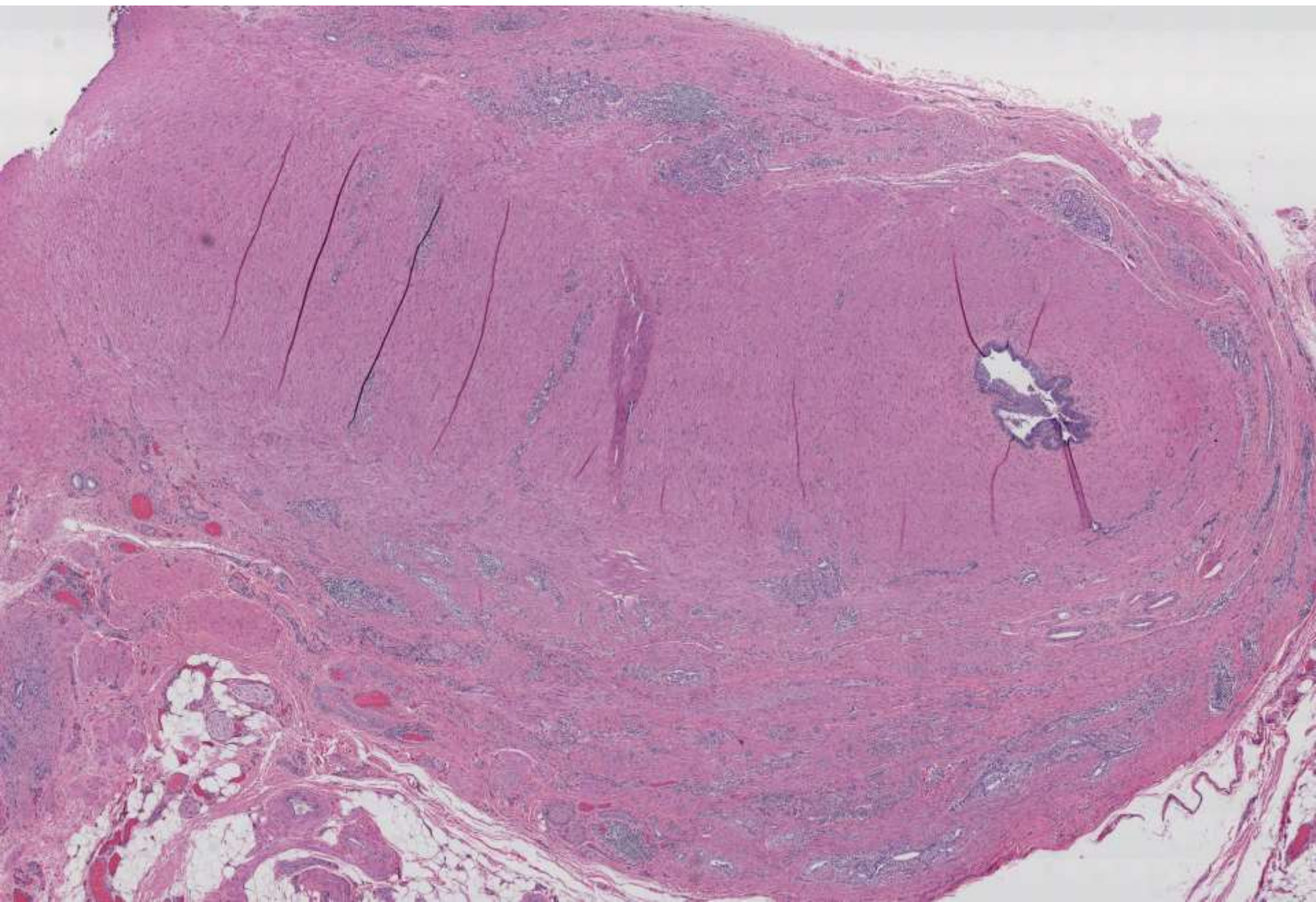
(scanned slide available)

**Malti Kshirsagar; El Camino Hospital**  
52-year-old male with “right spermatic  
granuloma” excisional specimen  
submitted.

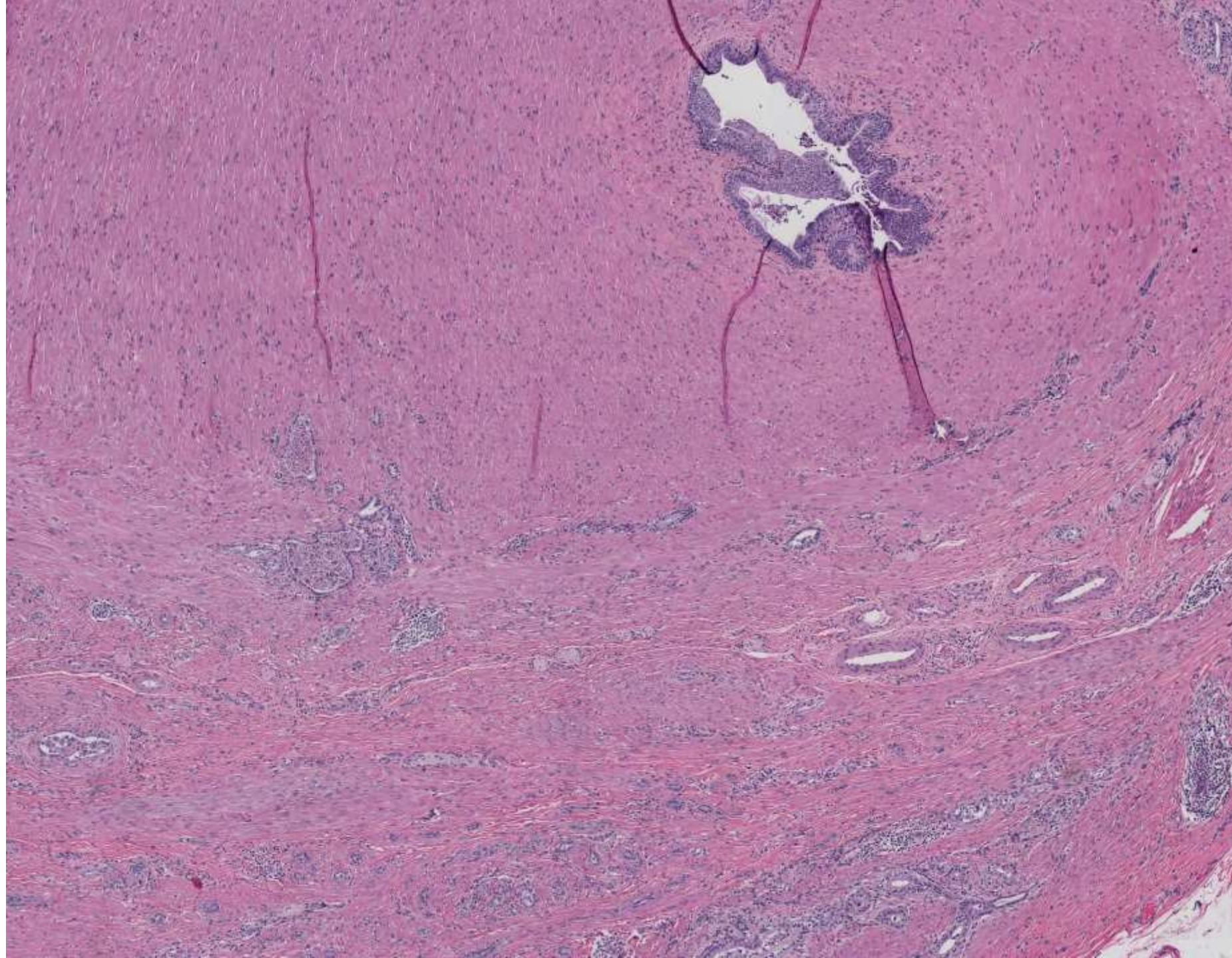




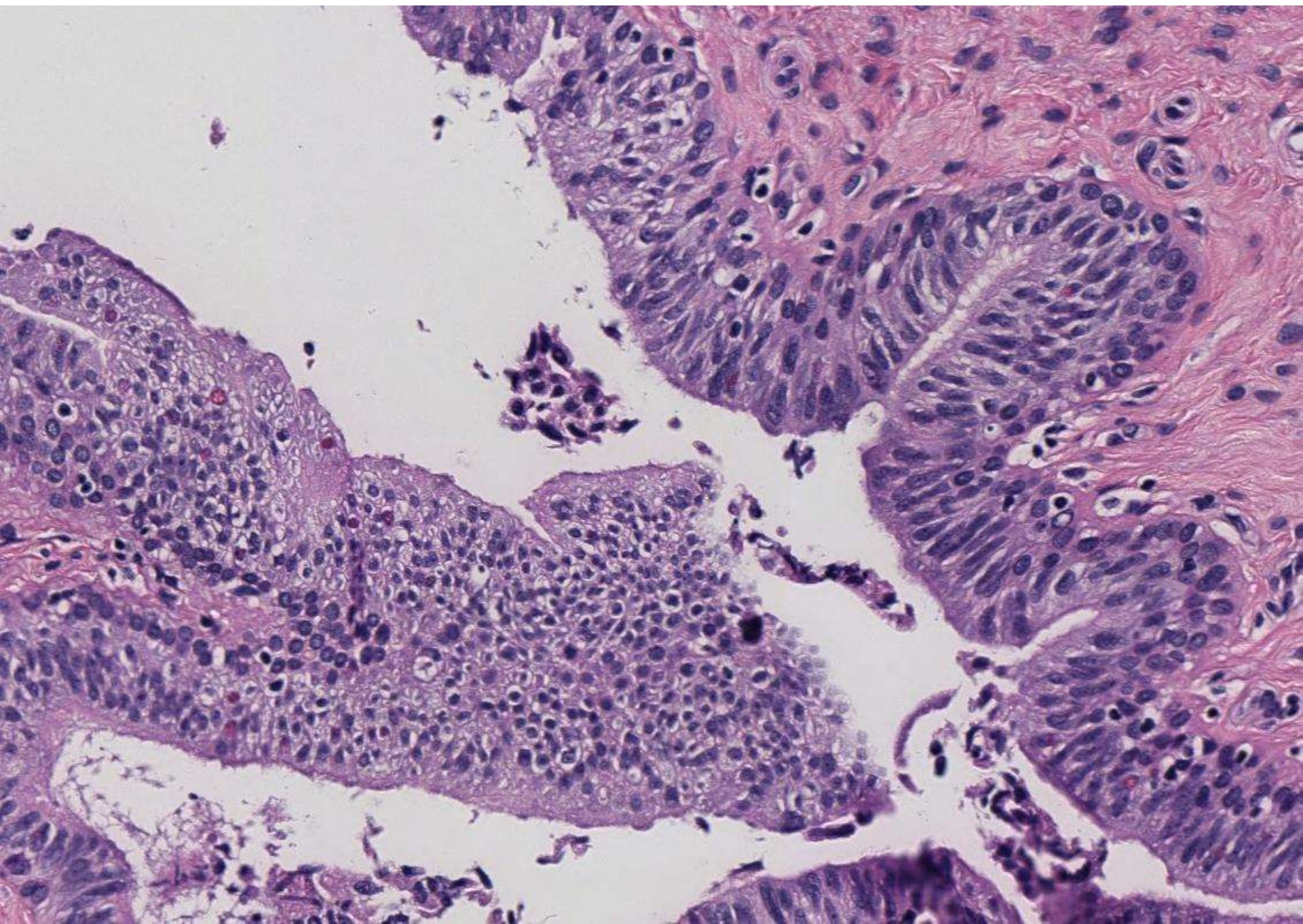




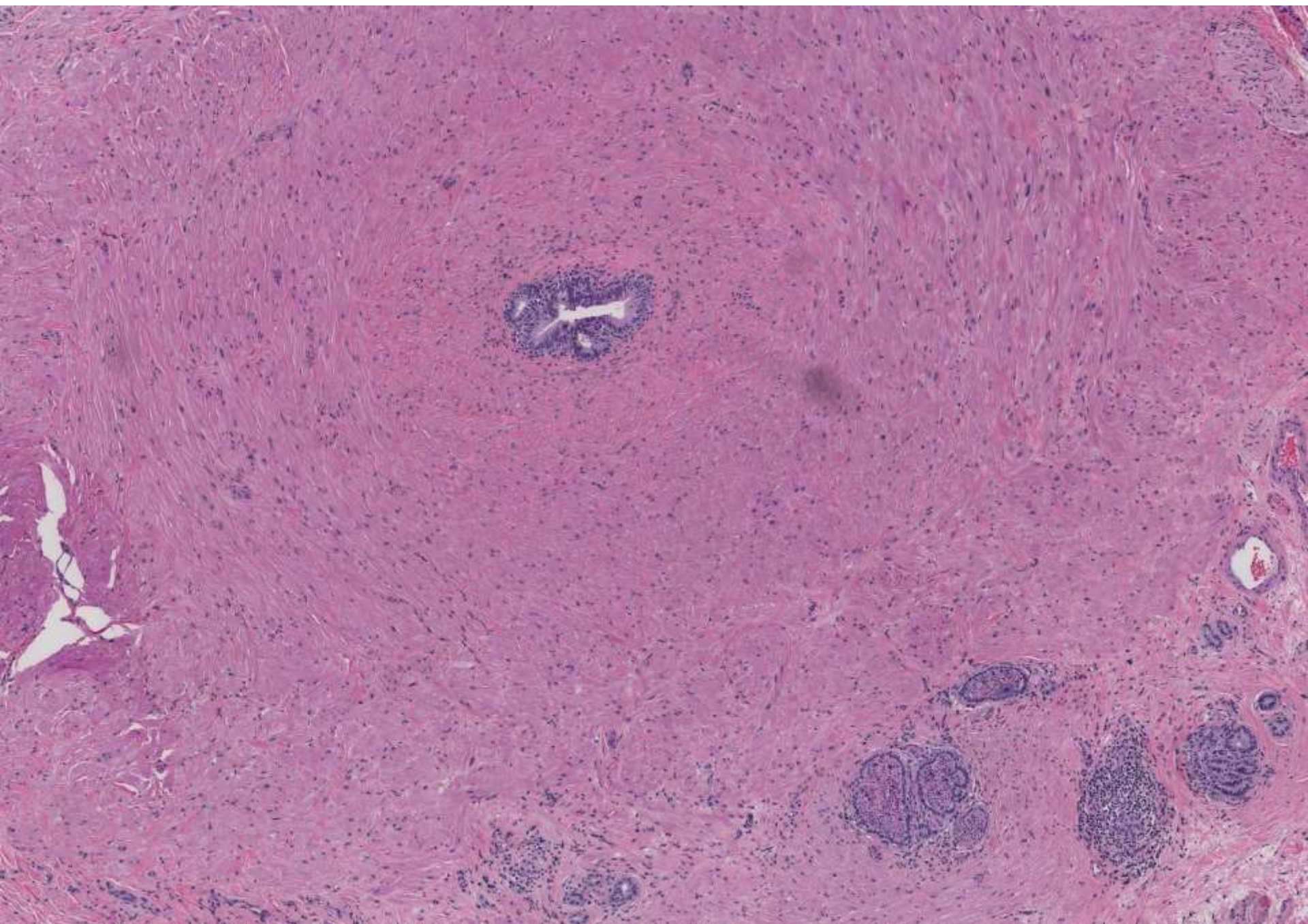




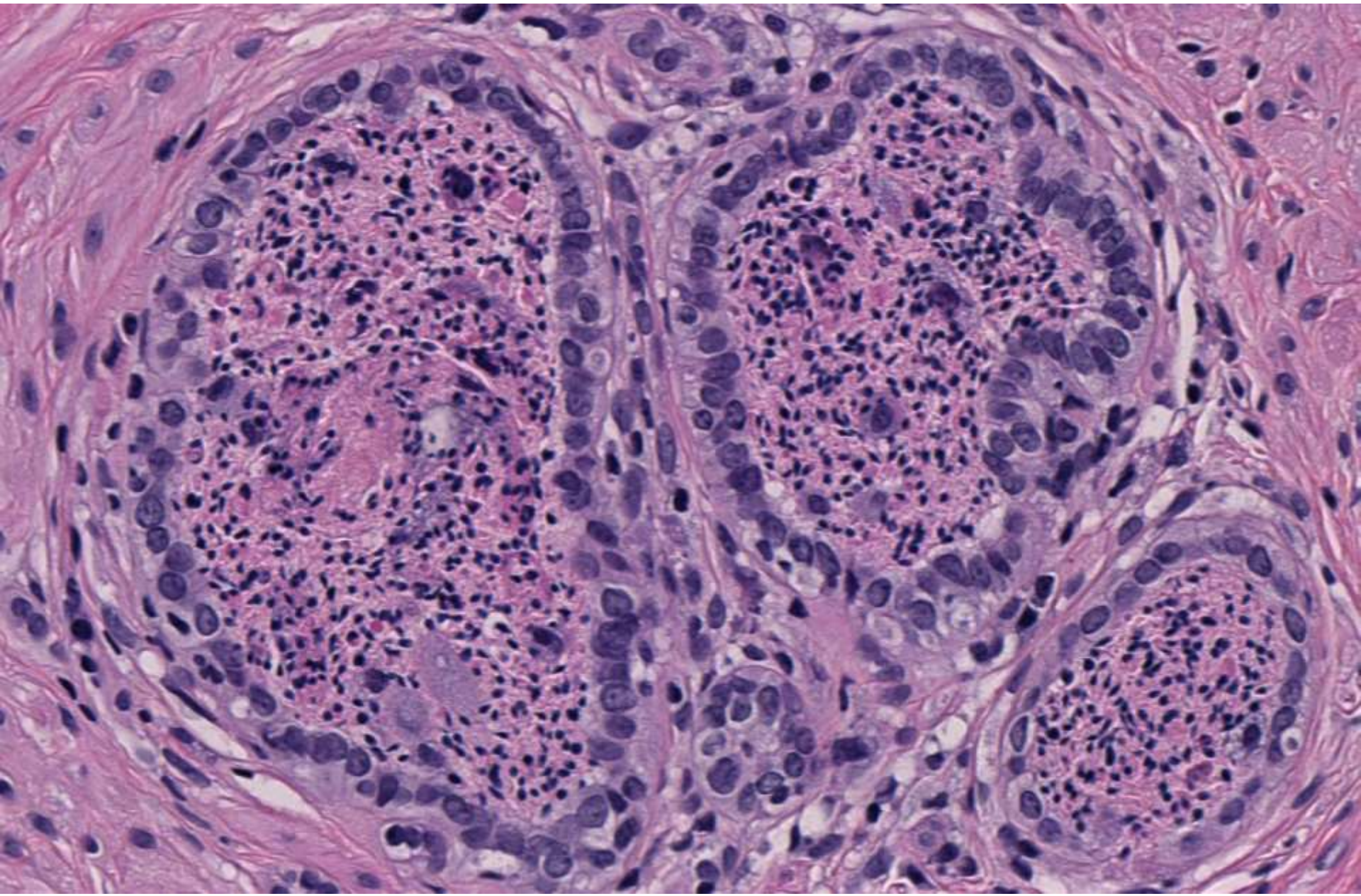




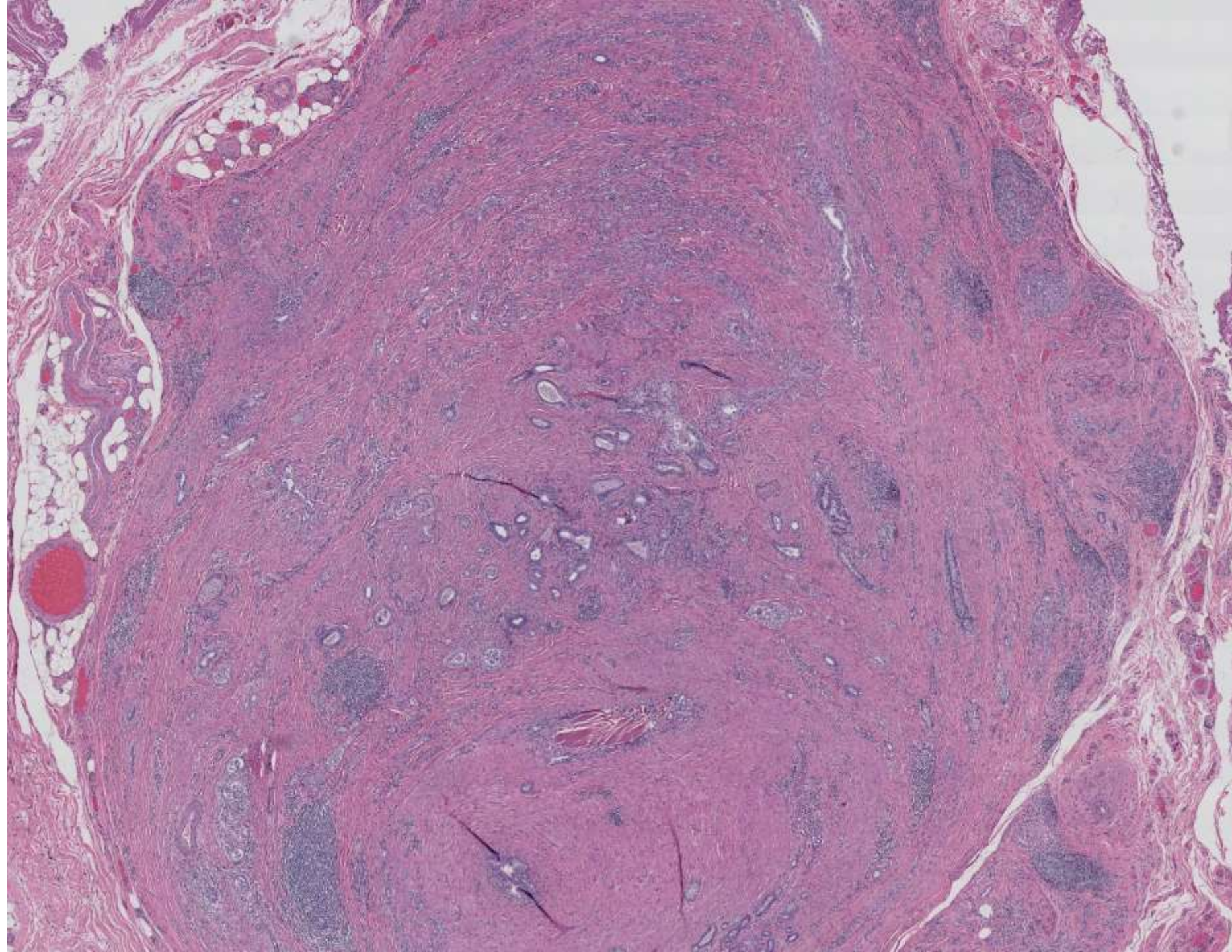




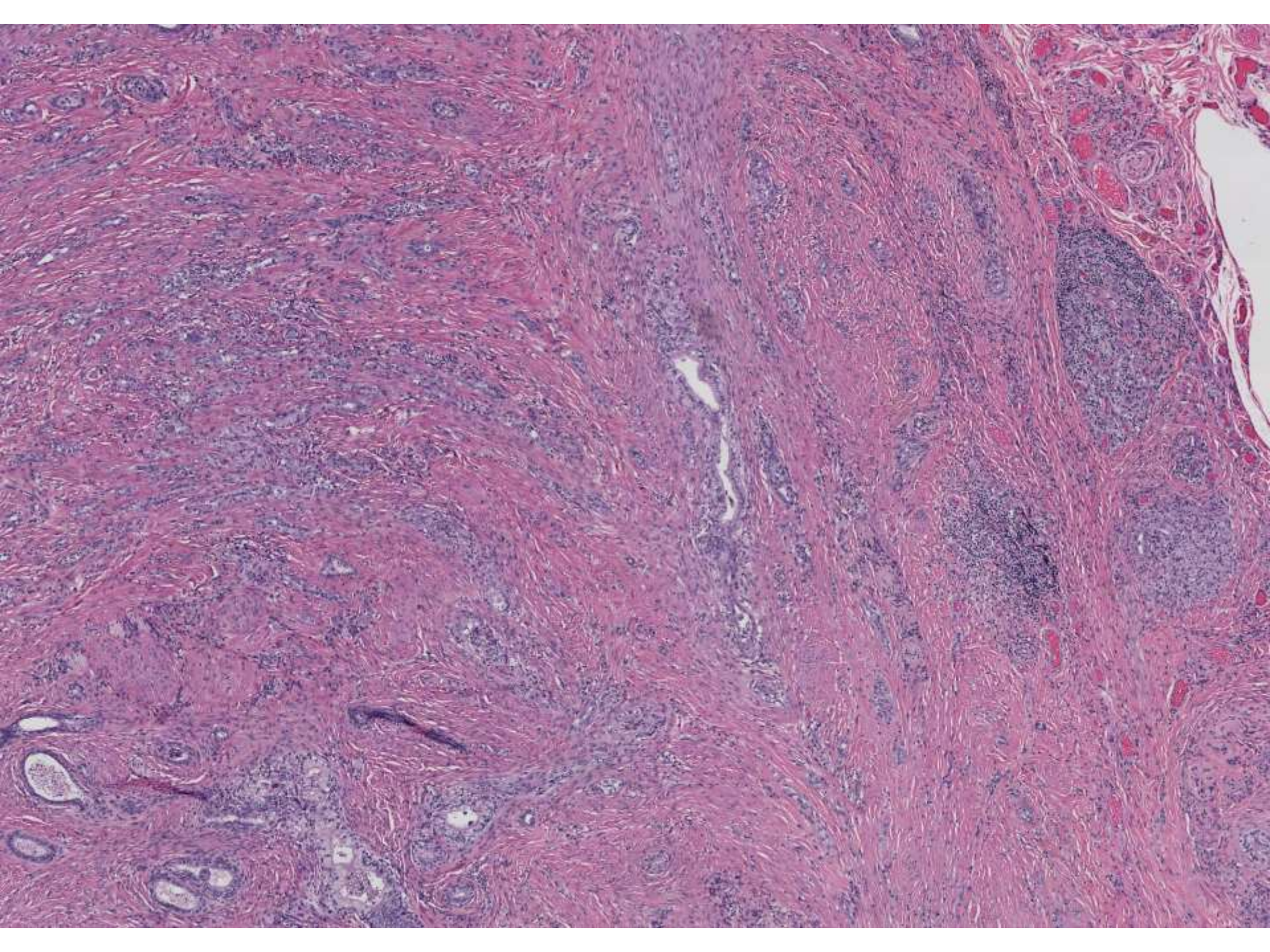




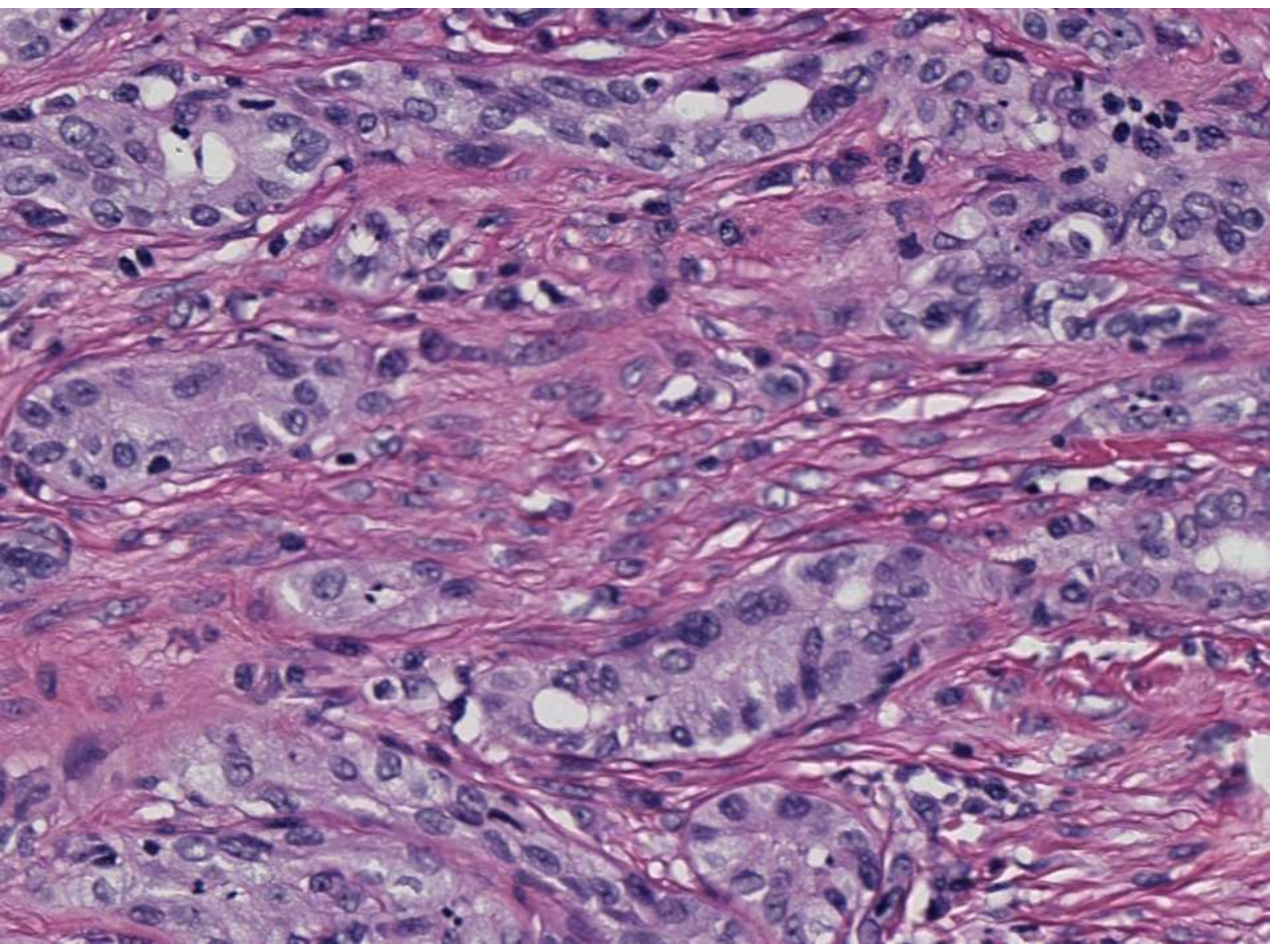




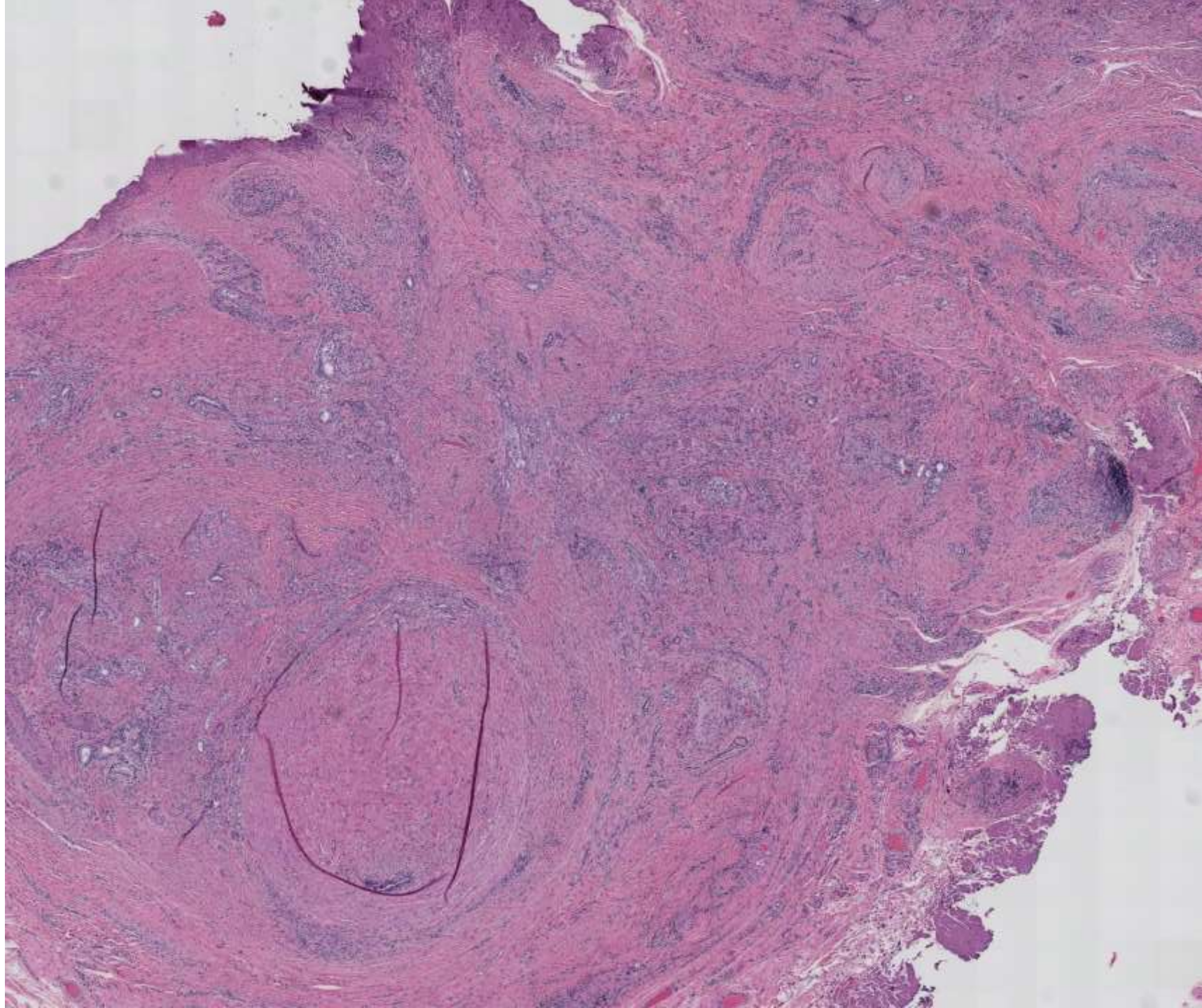




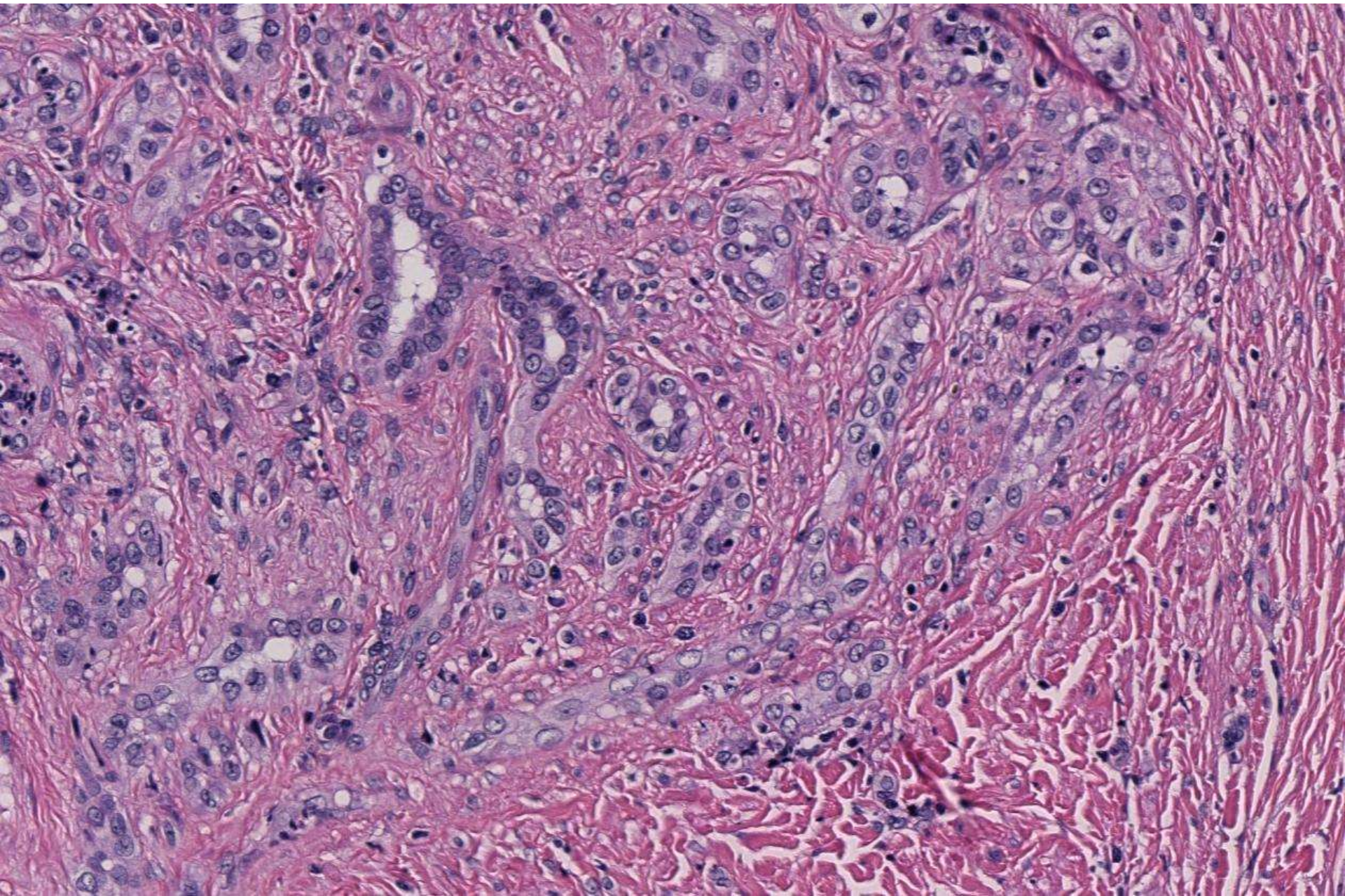




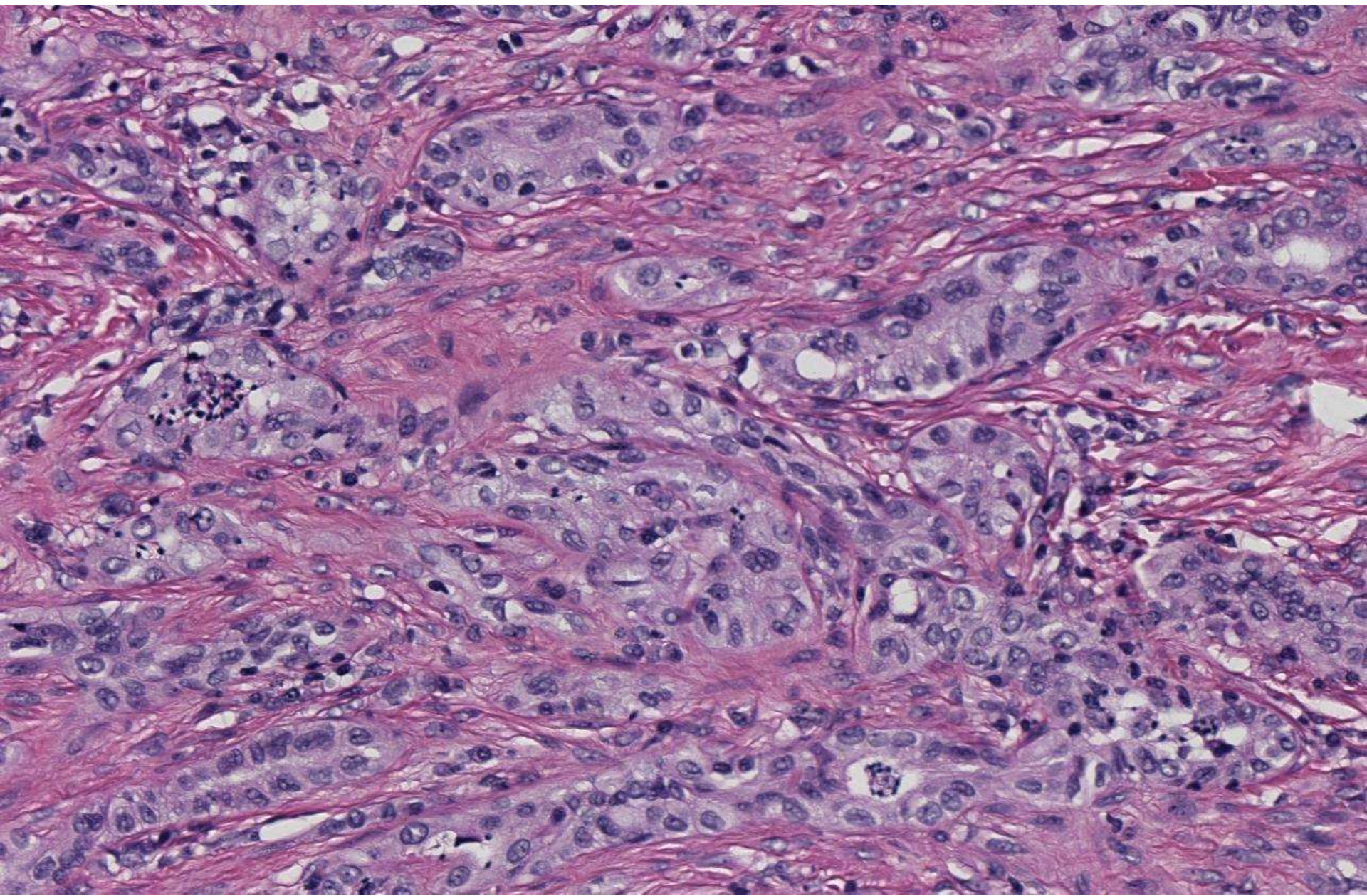










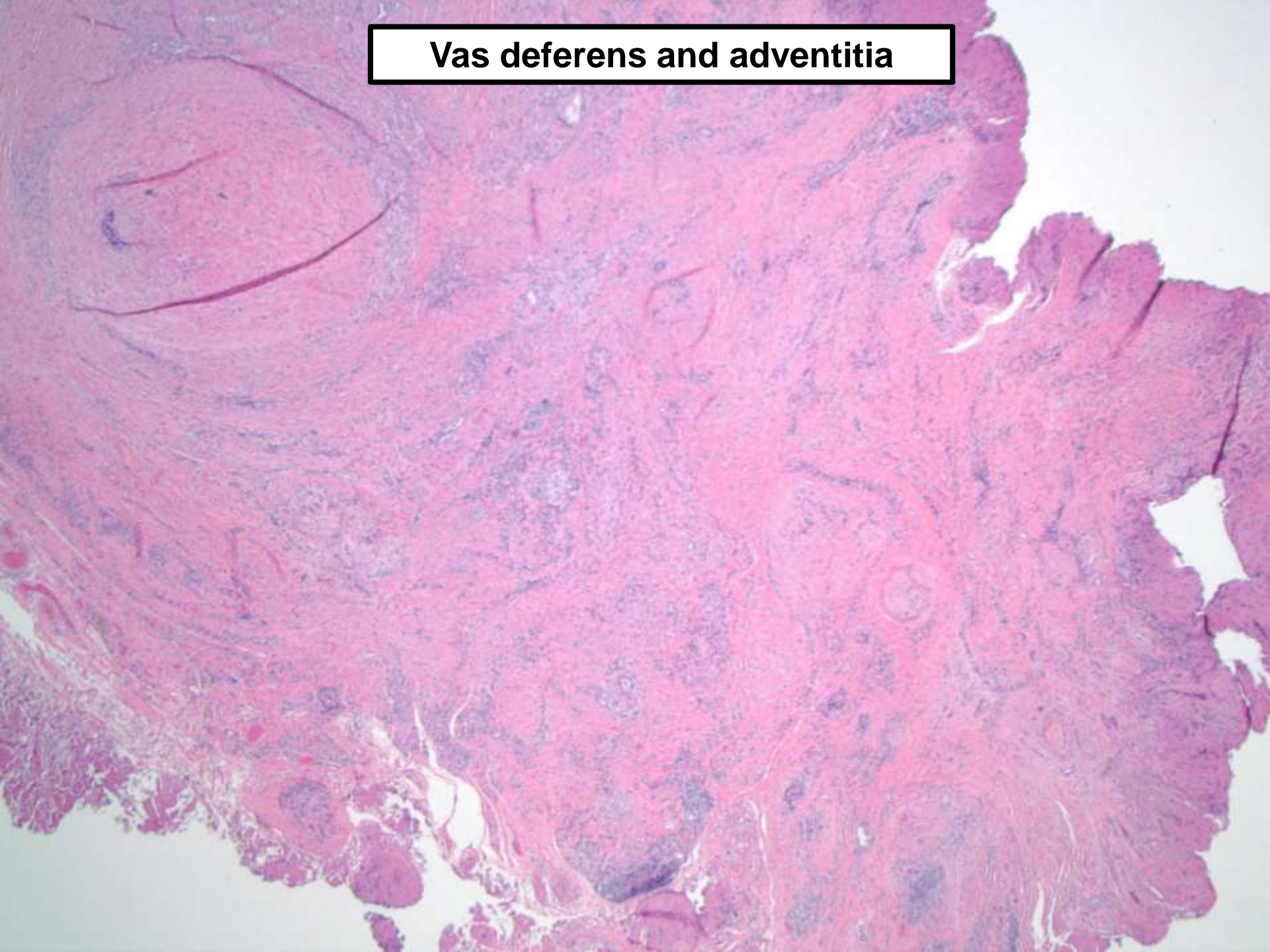




## **Differential diagnosis:**

1. Vasitis nodosa
2. Metastatic adenocarcinoma
3. Adenomatoid tumor – arising in  
spermatic cord or adjacent epididymis
4. Malignant mesothelioma

## Vas deferens and adventitia



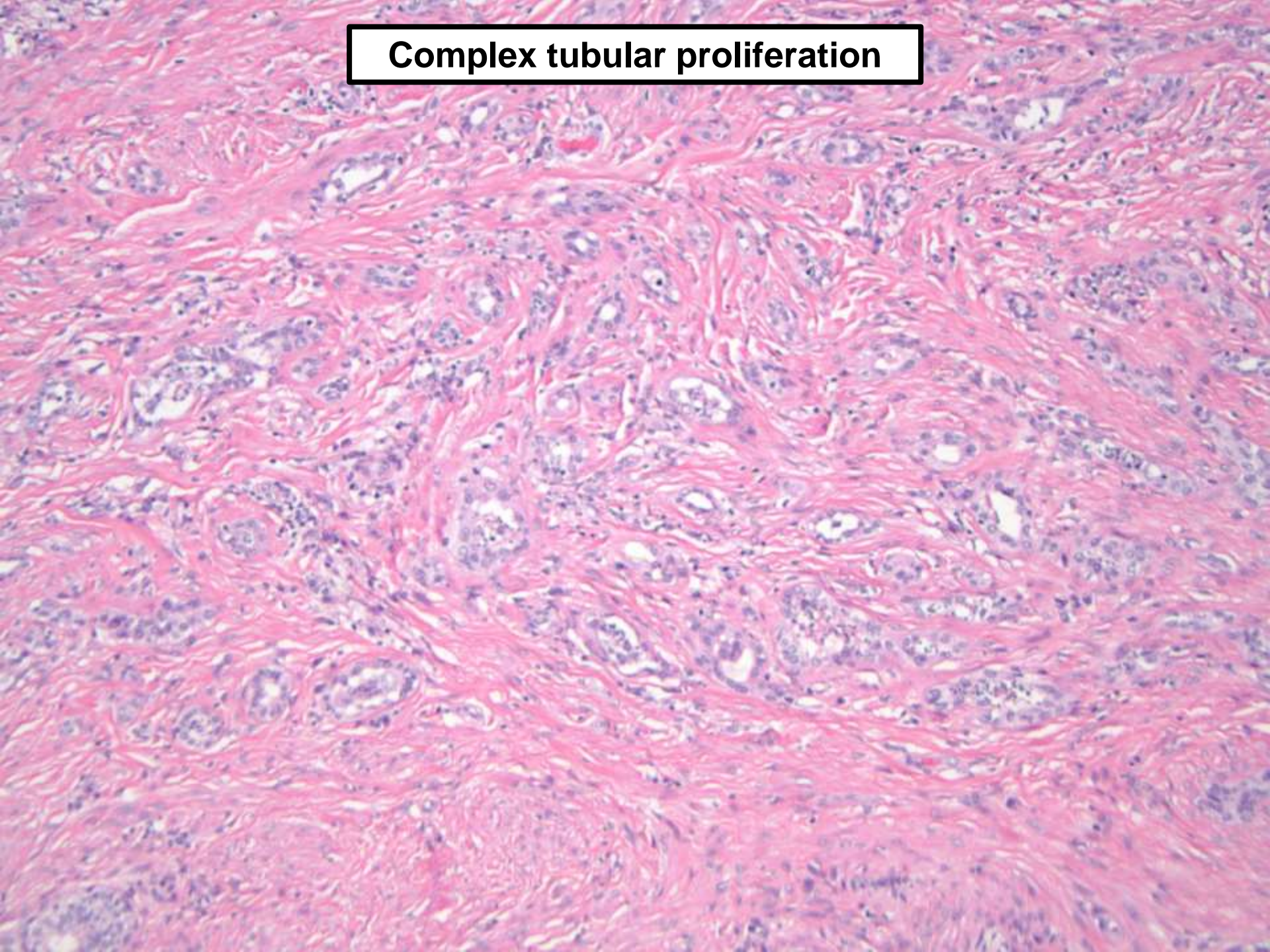


**Vas deferens with “tubular proliferation”**



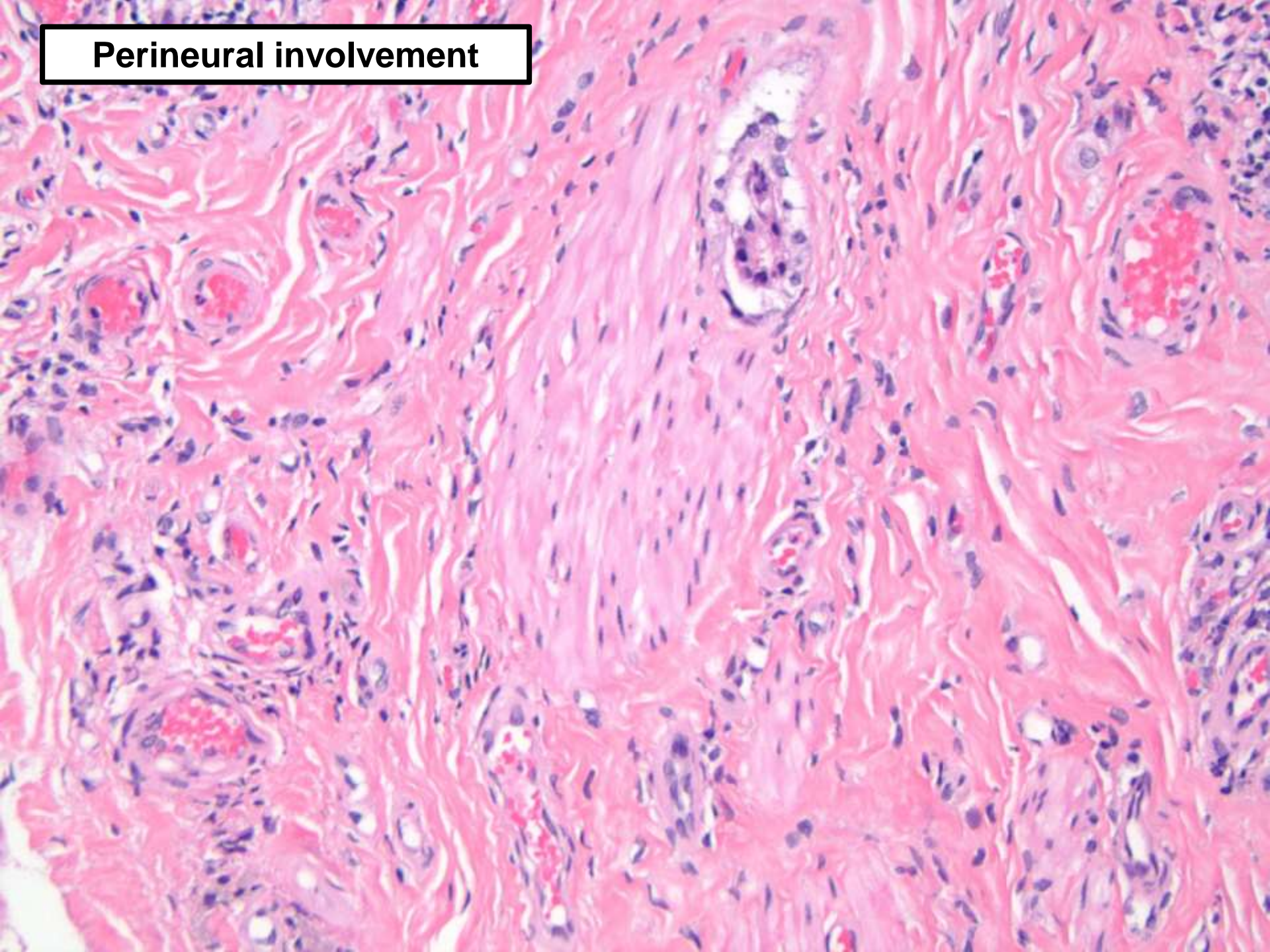


## Complex tubular proliferation



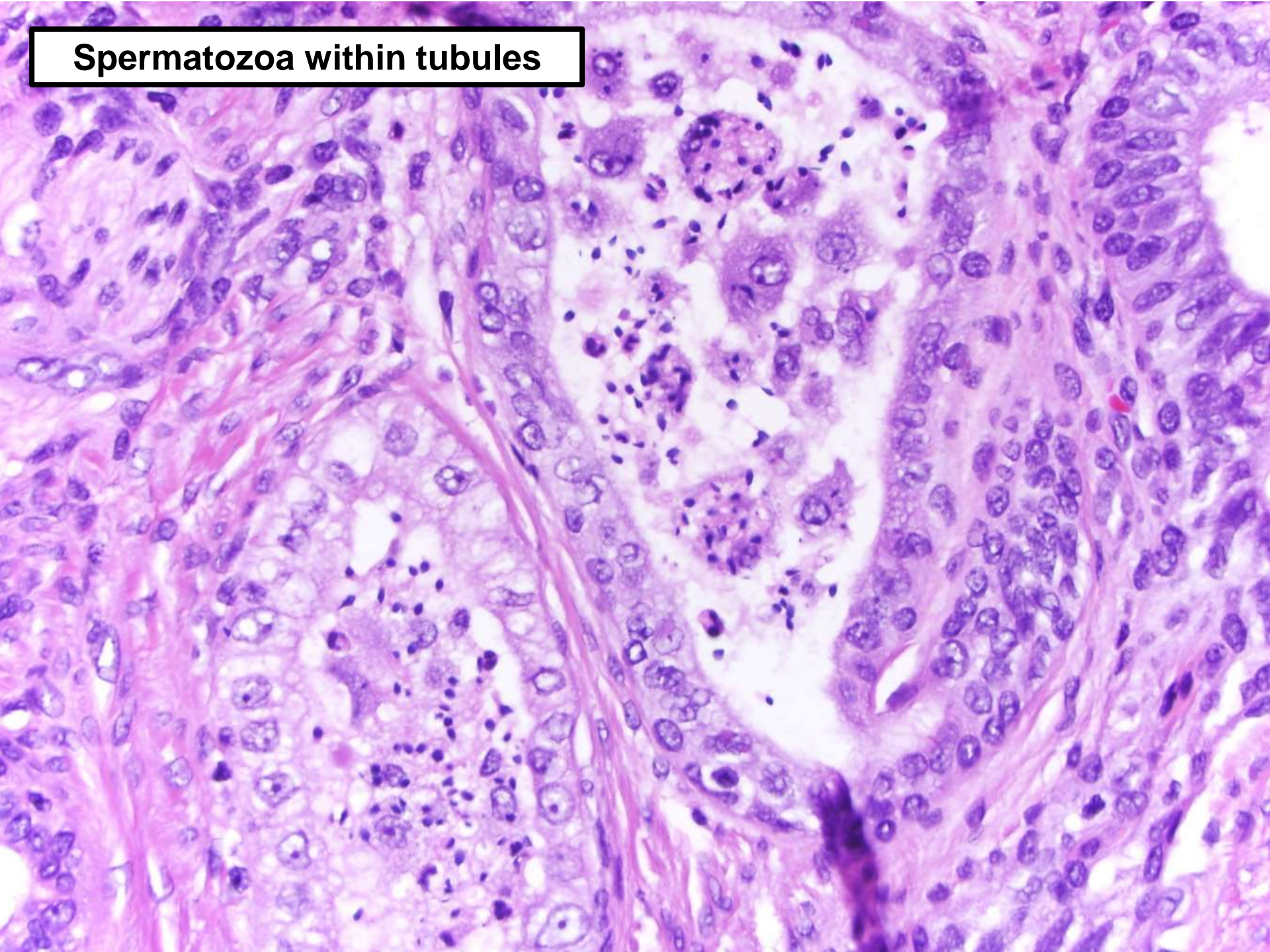


## Perineural involvement



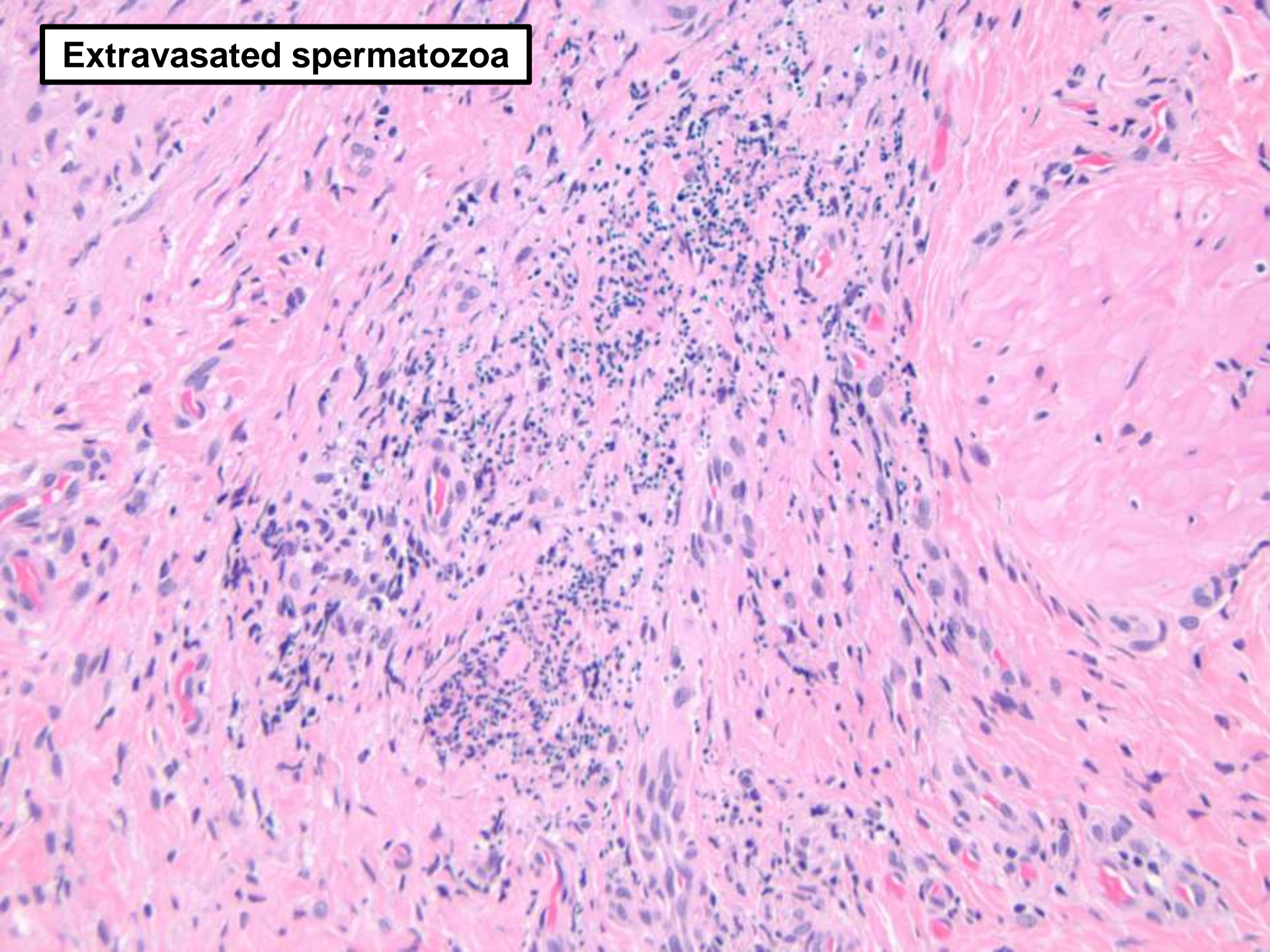


**Spermatozoa within tubules**



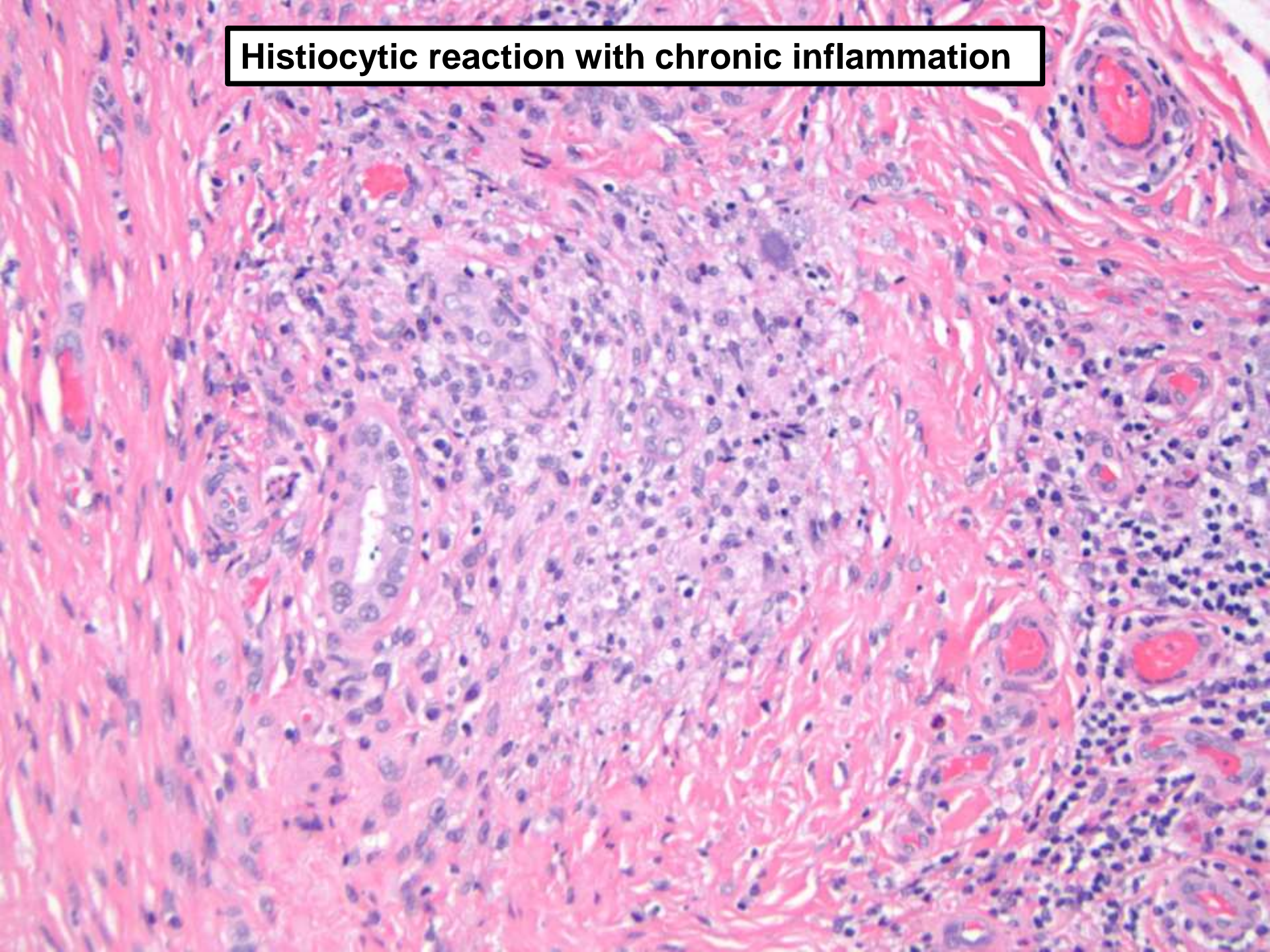


**Extravasated spermatozoa**



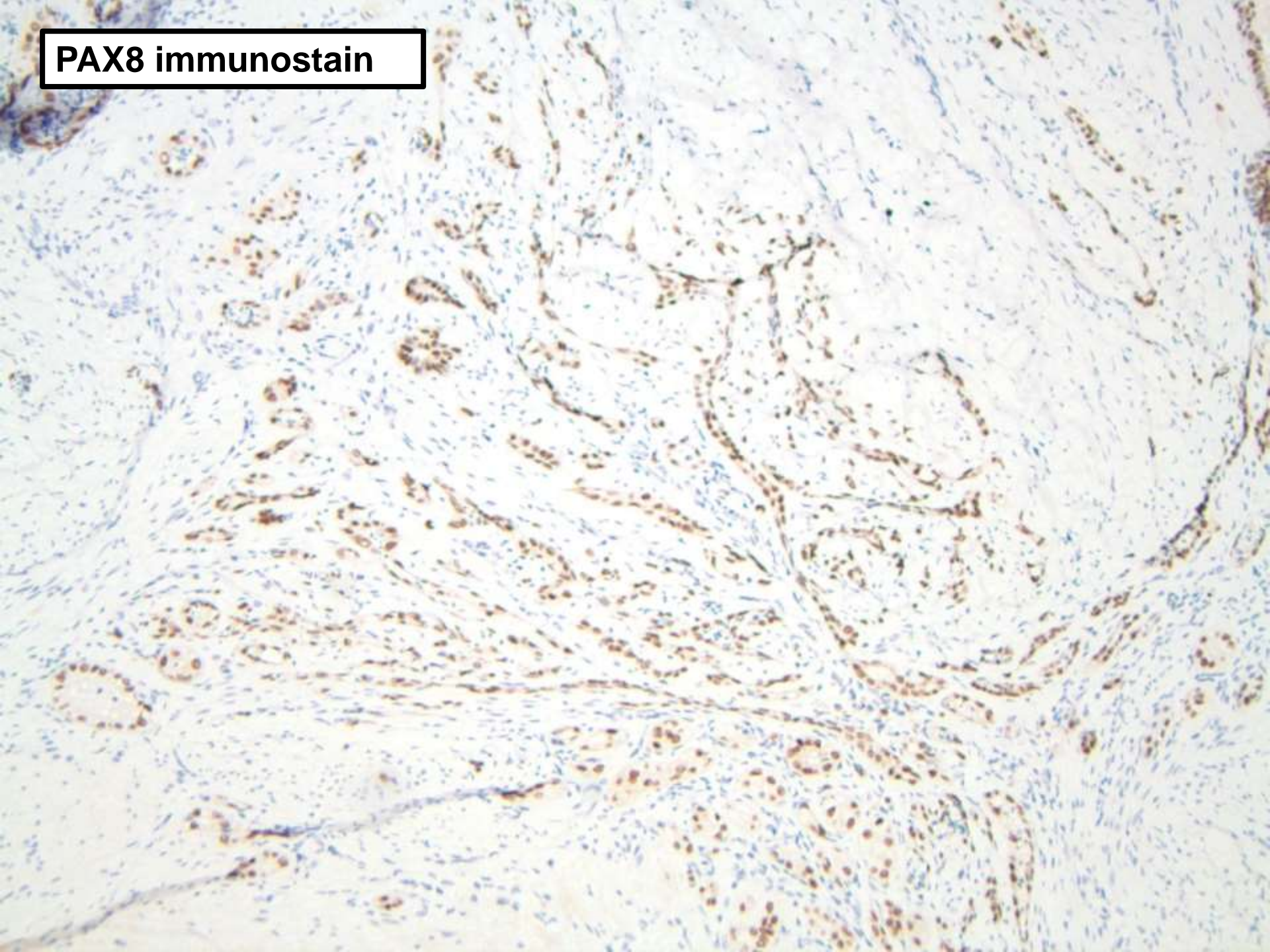


## Histiocytic reaction with chronic inflammation





**PAX8 immunostain**



# **Diagnosis: Vasitis nodosa**

**Per patient's wife:**

**“Oh yeah, he had a vasectomy years ago”**



Term "Vasitis nodosa" coined  
in 1943 by JA Benjamin et al

Vasitis nodosa: A new clinical  
entity simulating  
tuberculosis of the vas  
deferens. J Urol 49: 575-  
582, 1943

Subsequent reports  
(1954-1967) referred to  
these lesions as:

1. "Unusual vas deferens  
tumor"
2. "Spermatic cord tumor"
3. Vas deferens lesion  
"simulating salpingitis  
isthmica nodosa"
4. "Mesonephric  
hamartoma"

## Vasitis Nodosa

ALBERT L. OLSON, M.D.

*Department of Pathology, Loma Linda University Hospital,  
Loma Linda, California 92354*

### ABSTRACT

Olson, Albert L.: Vasitis nodosa. *Amer. J. Clin. Path.* 55: 364-368, 1971. Three cases in which extravasation of spermatozoa in the wall of the vas deferens occurred, accompanied by formation of epithelial-lined spaces within the wall, are reported. All of the patients previously had undergone vasectomies. Review of the literature reveals several cases with similar lesions but different names. The author suggests that the lesions be designated "vasitis nodosa," partly because what apparently was the first case reported was described by that name and partly because of the resemblance to salpingitis isthmica nodosa. Obstruction may be the most significant factor in pathogenesis.

DURING THE PAST TWO DECADES there have been several reports of nodules of the vas deferens which contained intramural epithelial structures. However, because of different etiologic concepts various names have been applied to these lesions. This paper will present three similar cases, with suggestions as to pathogenesis and nomenclature.

### Report of Cases

**Case 1.** A 31-year-old man had a repeat vasectomy because of failure of a vasectomy done 19 months earlier. One of the segments from the second operation appeared normal grossly, but the other was a firm, triangular specimen measuring 2.5 by 1.8 by 0.7 cm.

**Case 2.** A 56-year-old man underwent a transurethral resection of the prostate accompanied by a bilateral vasectomy, although the history recorded a previous bilateral vasectomy in 1951. The surgeon noted that the left vas was enlarged and contained yellow mucoid fluid. The larger

of the two segments excised was 0.3 cm. in diameter.

**Case 3.** A 38-year-old man had pain in his groin for 5 or 6 days. Past history disclosed a vasectomy 10 years previously. A tender, firm nodule was palpated in the region of the right globus major, resulting in surgical exploration which revealed a nodule in the vas deferens. The submitted specimen was 3 cm. long and had an oval cross section measuring 1.4 by 0.5 cm.

### Microscopic Findings

**Case 1.** There was loss of the normal muscle pattern except in one eccentrically located region where a circular zone of smooth muscle surrounded a group of epithelial-lined spaces interpreted as remnants of the original lumen (Fig. 1). Elsewhere there was a mixture of fibrous connective tissue, smooth muscle fibers, numerous spermatozoa, histiocytes, and epithelial cells with pale-staining nuclei and cytoplasm. These epithelial cells occurred in randomly situated groups of small nests, acini, and ducts (Fig. 2). Spermatozoa were present in the lumina of a few of these

Received May 4, 1970; accepted for publication  
June 24, 1970.

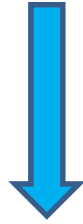
# **Vasitis Nodosa & Epididymis Nodosa**

- 1. Occur most commonly as late post-vasectomy changes (in up to 50% of men) – usually an incidental finding**
- 2. May also occur following trauma, herniorrhaphy, and prostatectomy**
- 3. Lesions typically small (less than 1 cm)**
- 4. Most lesions asymptomatic, though patients may present with scrotal swelling/pain/tenderness**
- 5. Most cases require no treatment. In patients who are symptomatic, surgical resection is generally curative**

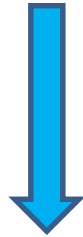


# Why does it happen?

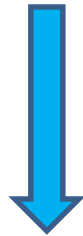
**Tubular obstruction**



**Increased luminal pressure**



**“Blow out” injury with leakage of spermatozoa**



**Inflammatory reaction with excessive regeneration  
of the epithelial lining**



# **Vasitis Nodosa & Epididymitis nodosa- Histology**

- 1. Proliferation of small ducts and gland-like structures in walls of vas deferens (and epididymis) – may not communicate with lumen of vas deferens**
- 2. Glands/tubules lined by bland cuboidal or low columnar cells**
- 3. May see perineural and possibly vascular invasion**
- 4. Intraluminal spermatozoa are a reassuring finding**
- 5. Extravasation of sperm with inflammatory response**
- 6. 70% cases show associated sperm granulomas**



*References:*

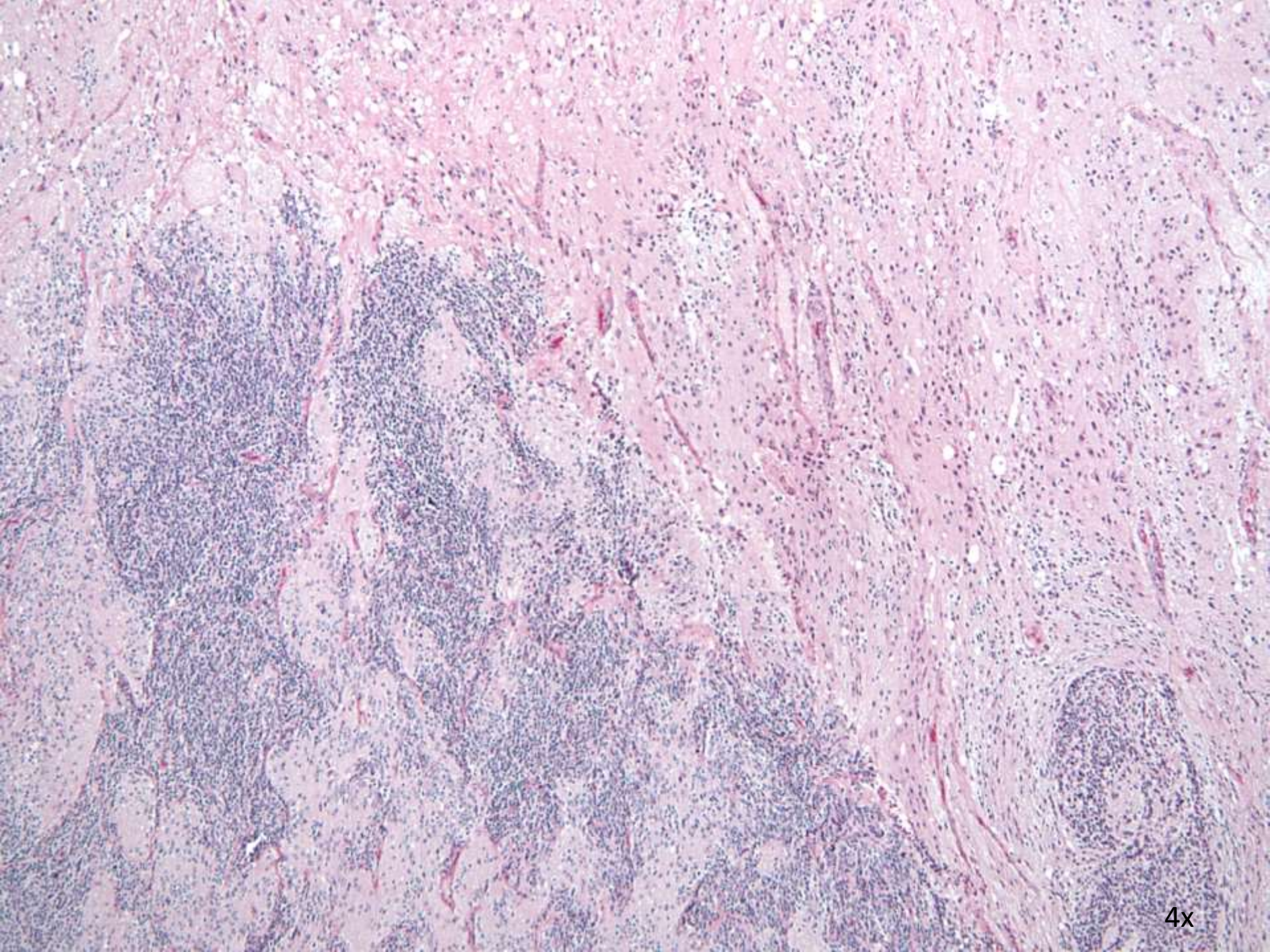
1. S Easley and G T MacLennan. Vasitis and epididymitis nodosa. 2006 Apr; J Urol 175: 1502.
2. A Olson. Vasitis Nodosa. Am J Clin Pathol. 1971 Mar; 55(3): 364-8.
3. JA Benjamin et al. Vasitis nodosa: A new clinical entity simulating tuberculosis of the vas deferens. 1943; J Urol 49: 575-582.

# SB 6136

**Seth Lummus/Don Born; Stanford**

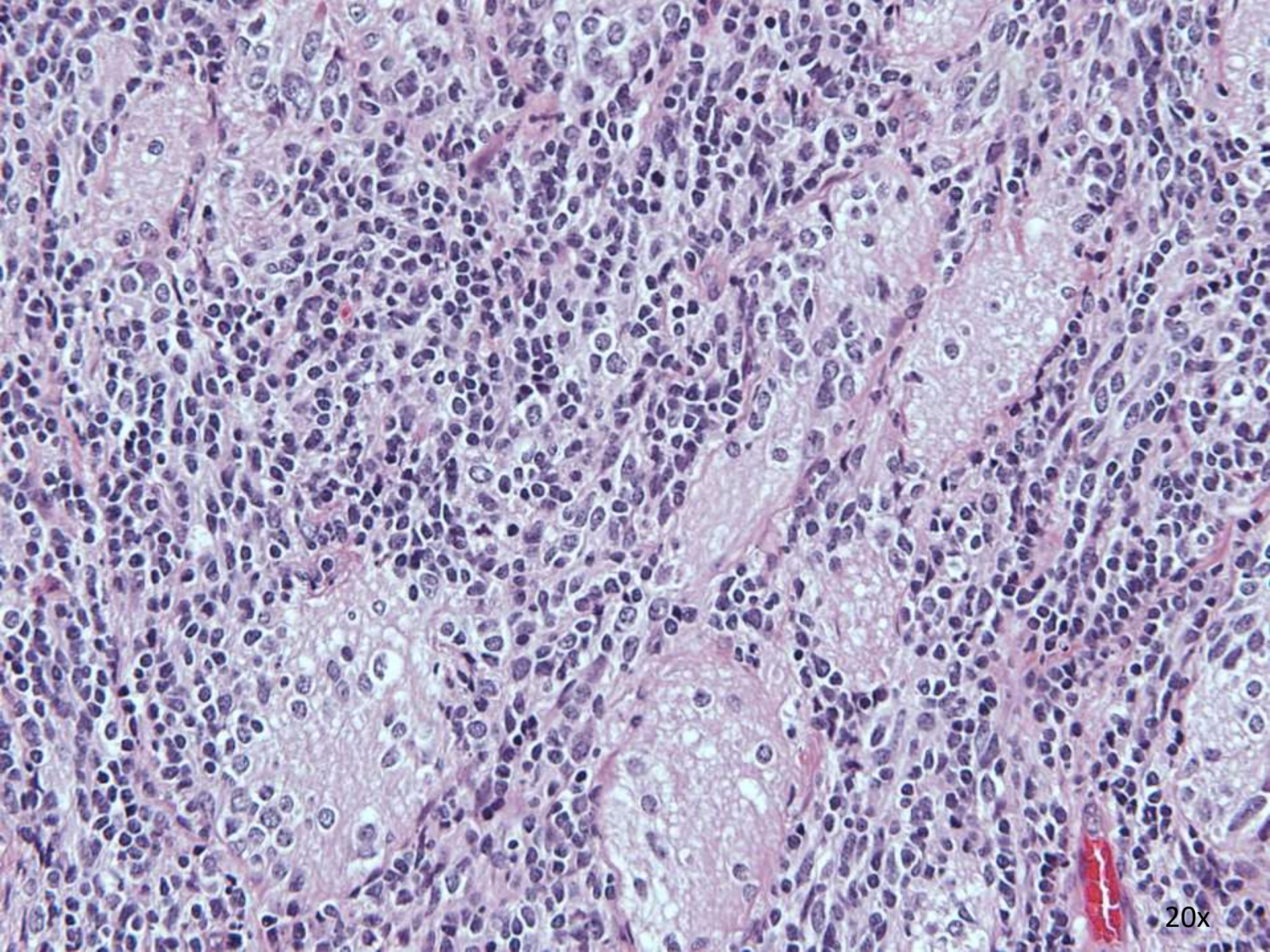
2-year-old 11 month female, previously healthy, who fell backwards from her high chair and struck her head on the wooden floor. An X-ray demonstrated a skull fracture, and a subsequent neuroimaging showed a large heterogenous mass in the left parietal, occipital, and frontal lobes.





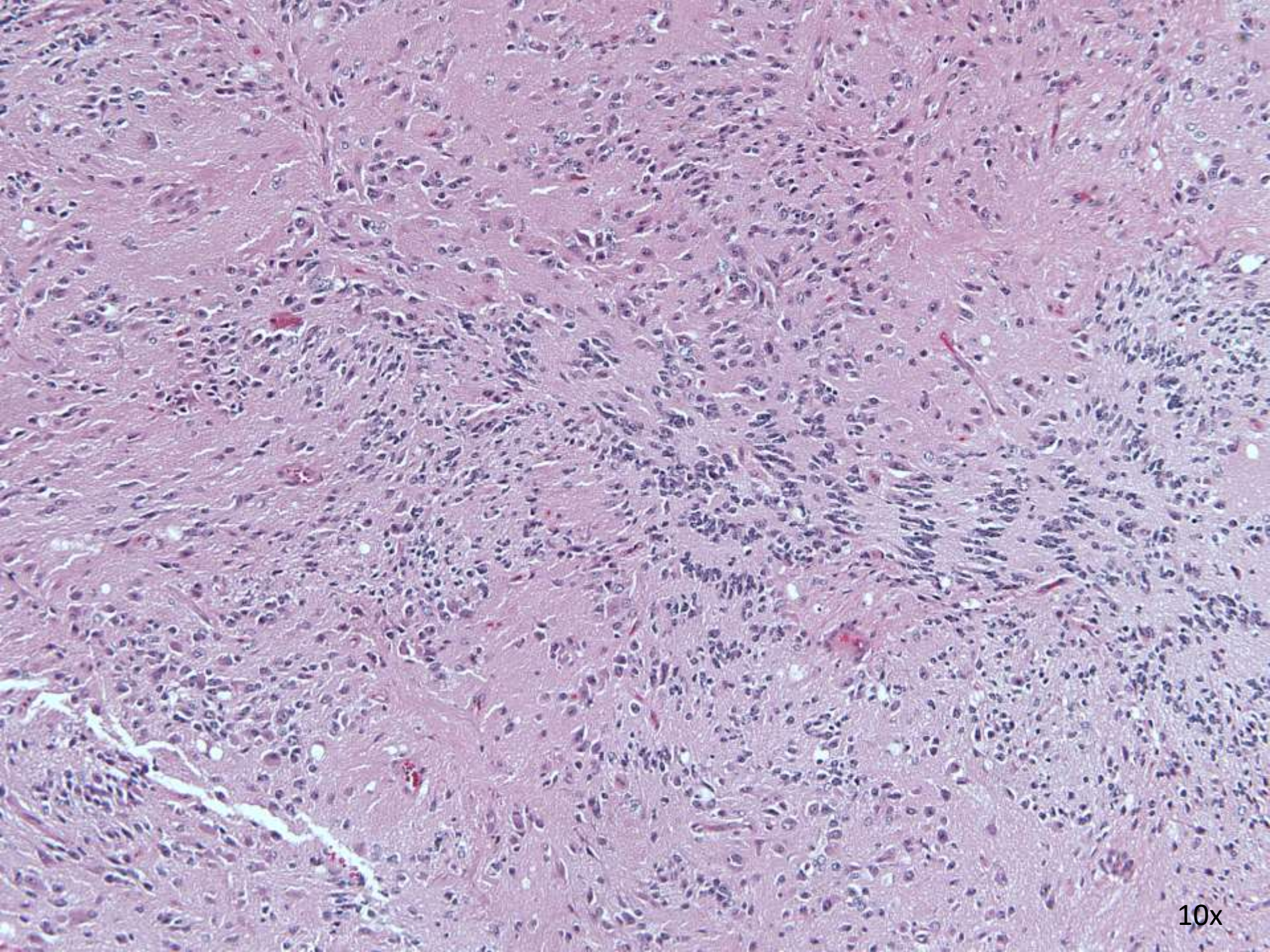
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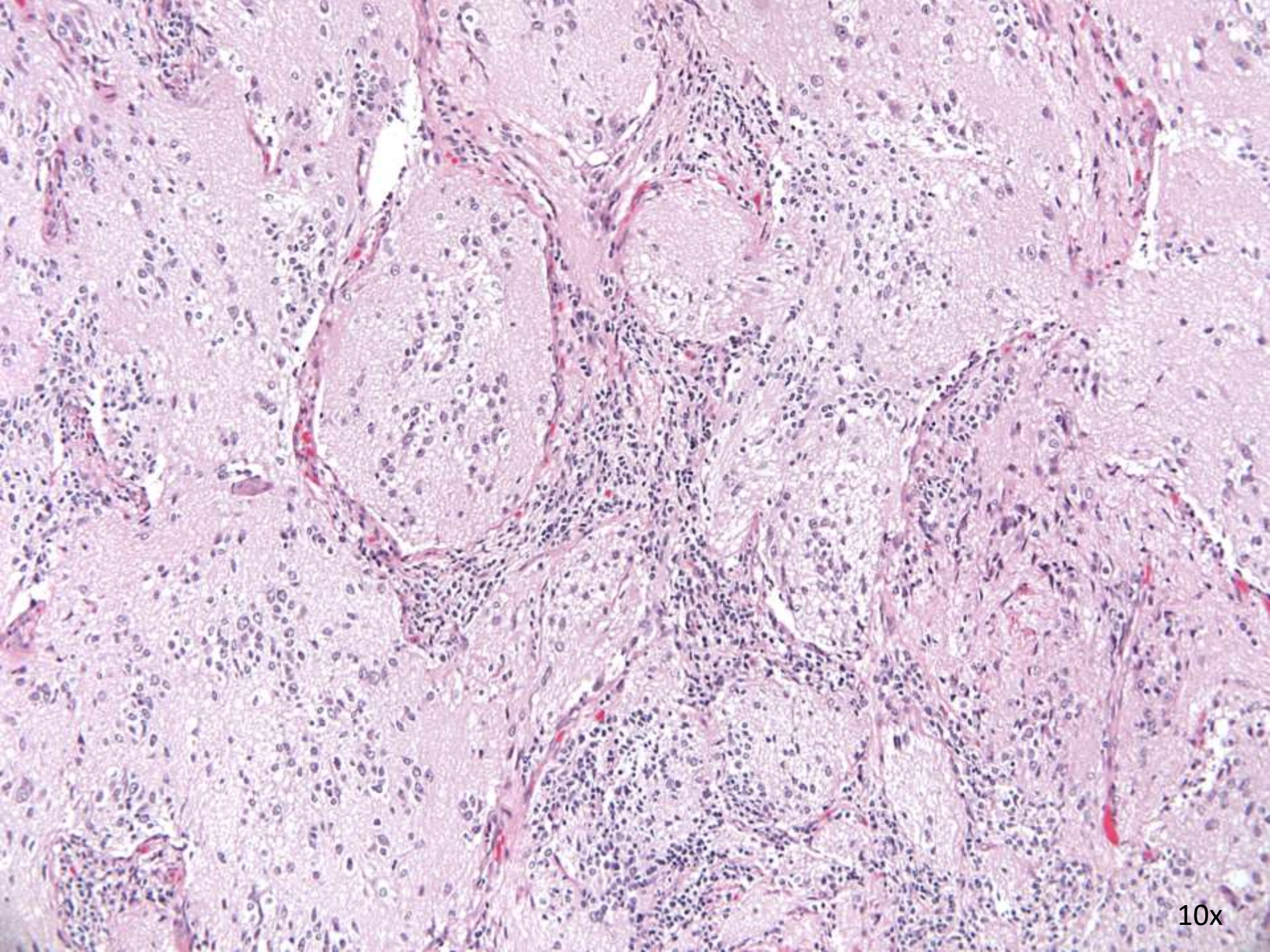
20x





10x

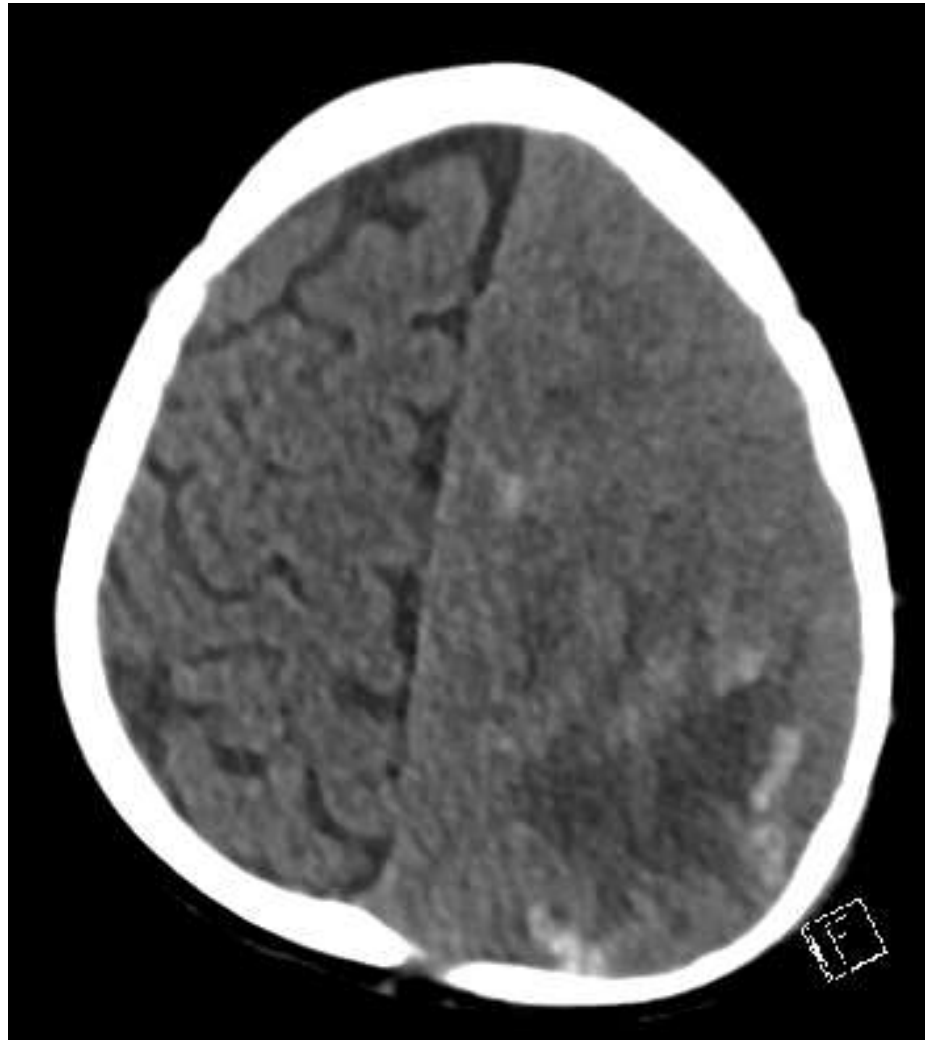




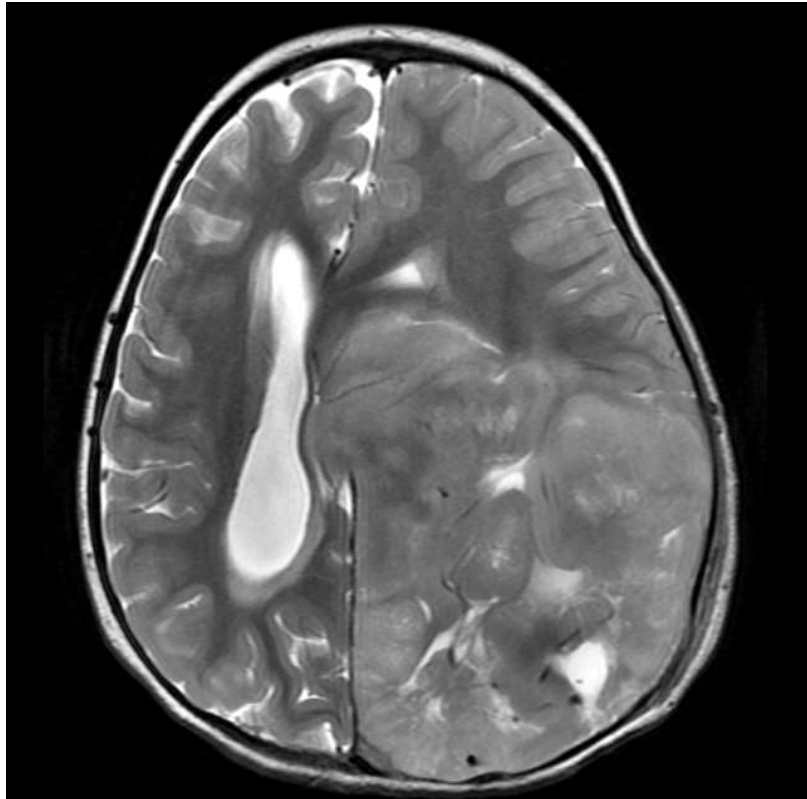
10x



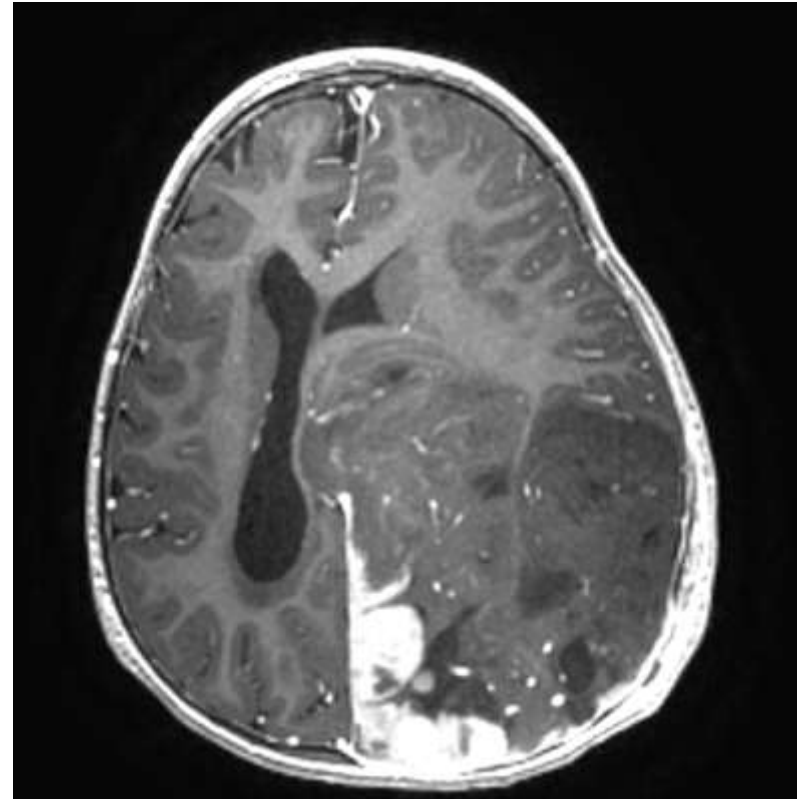
# Initial CT



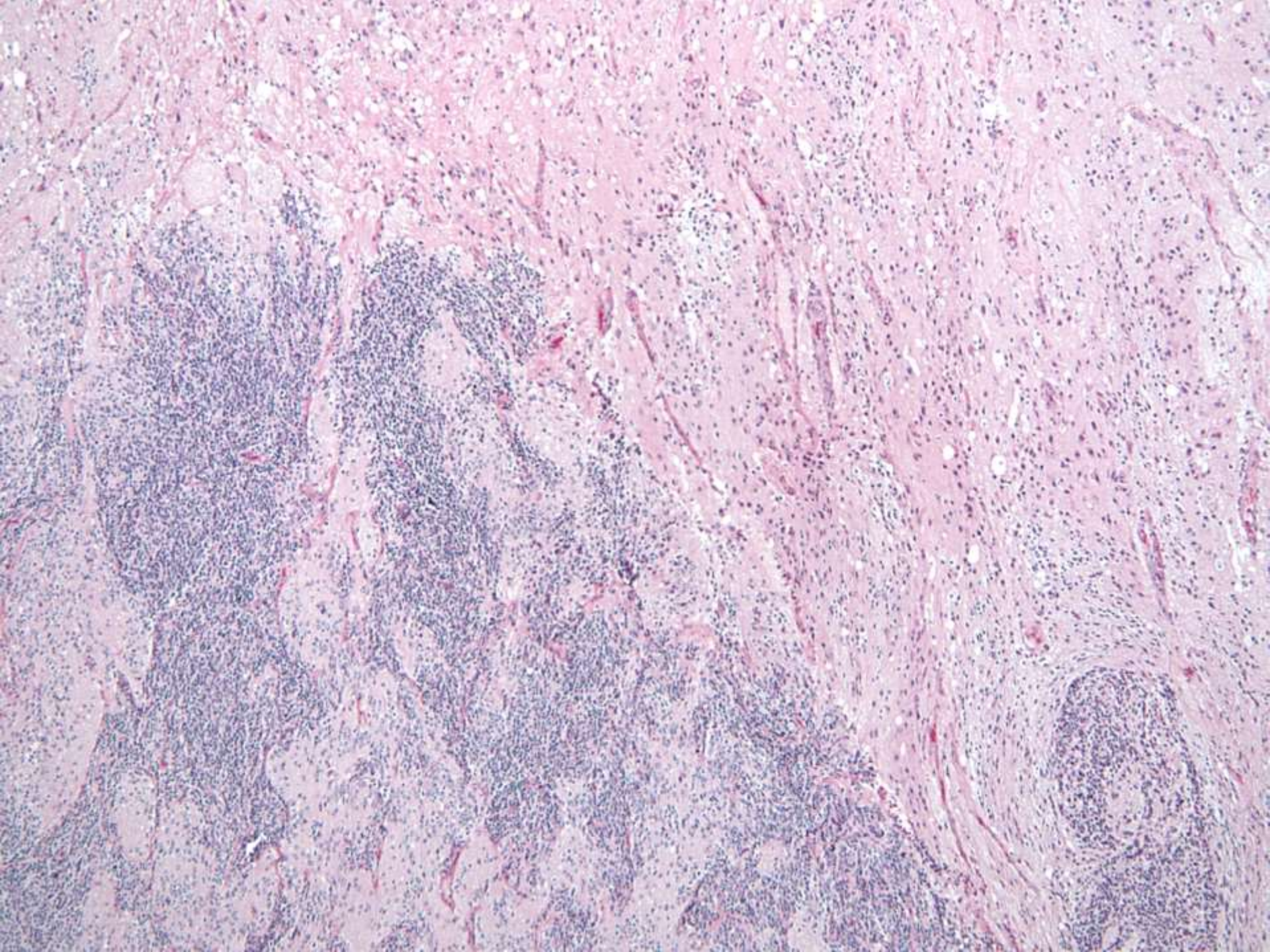
T2 FLAIR



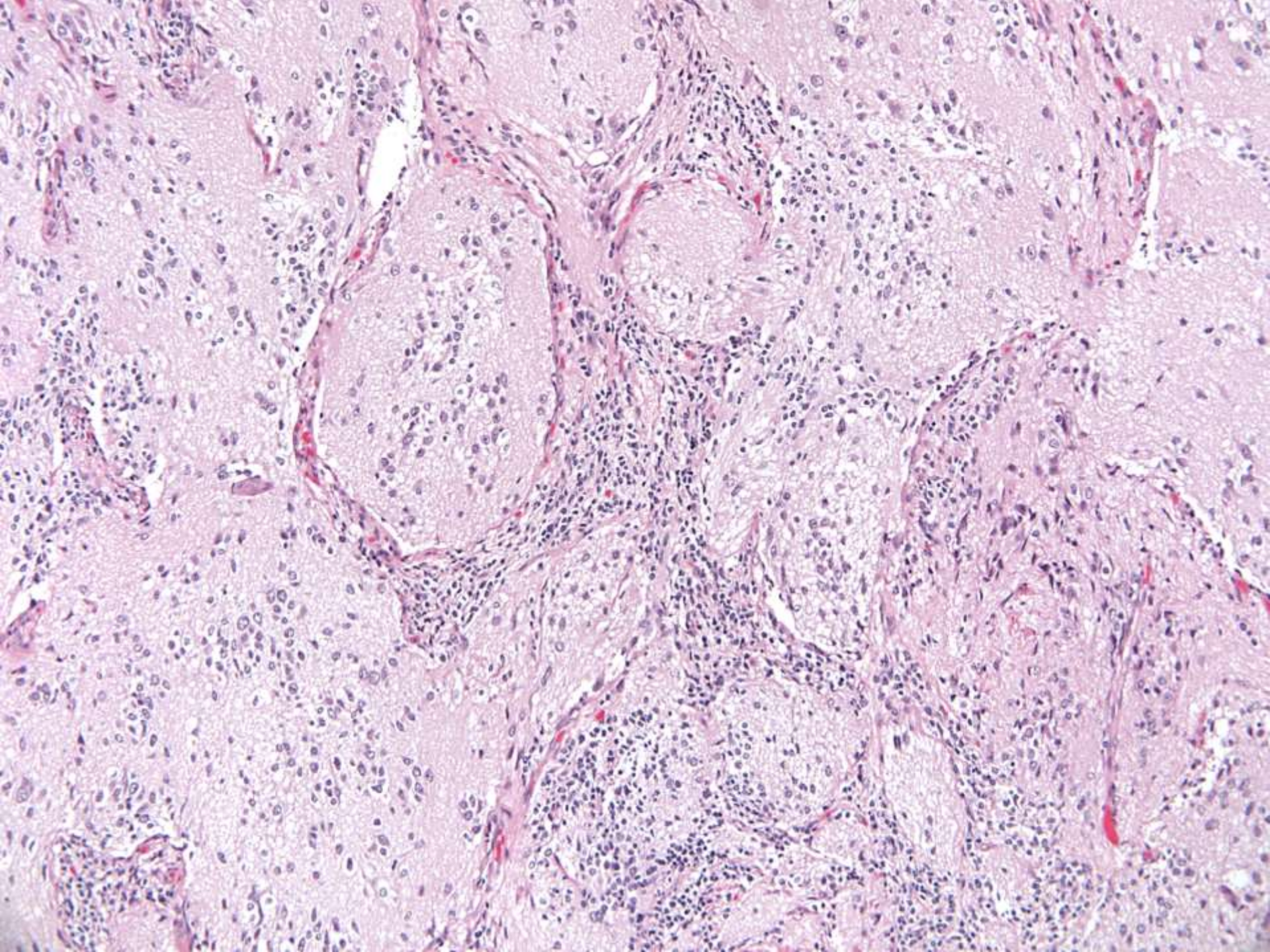
T1 Gad



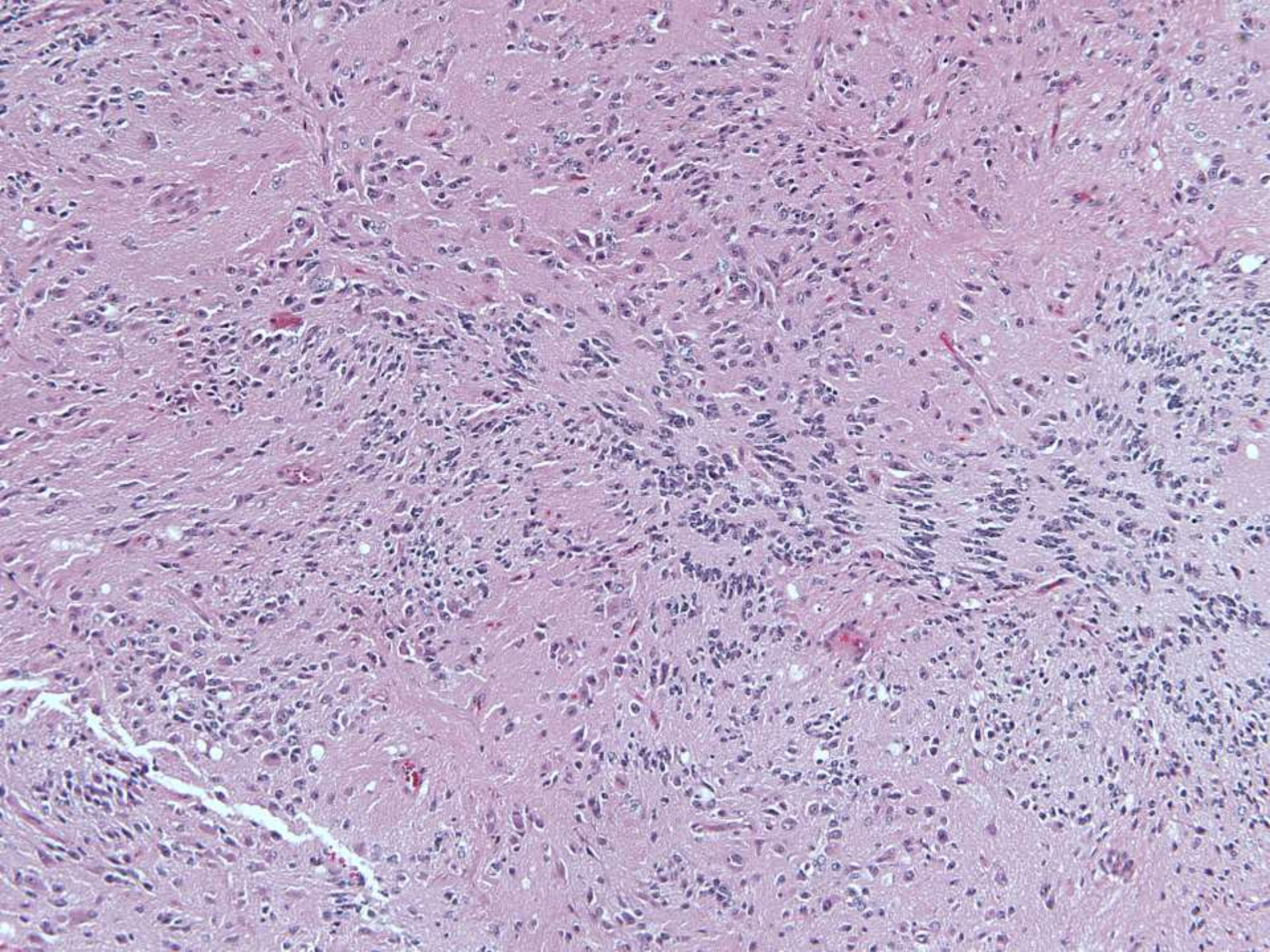




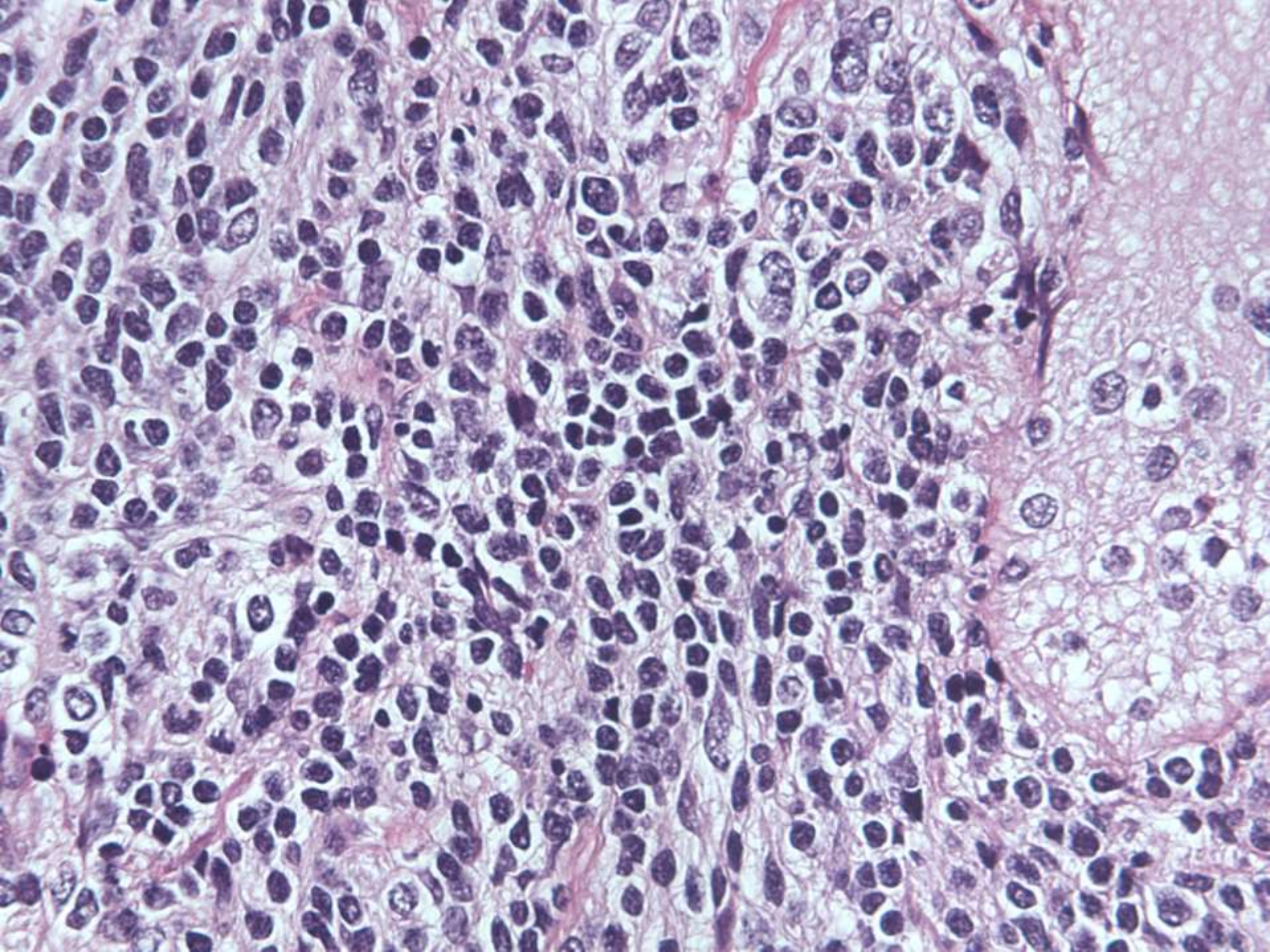




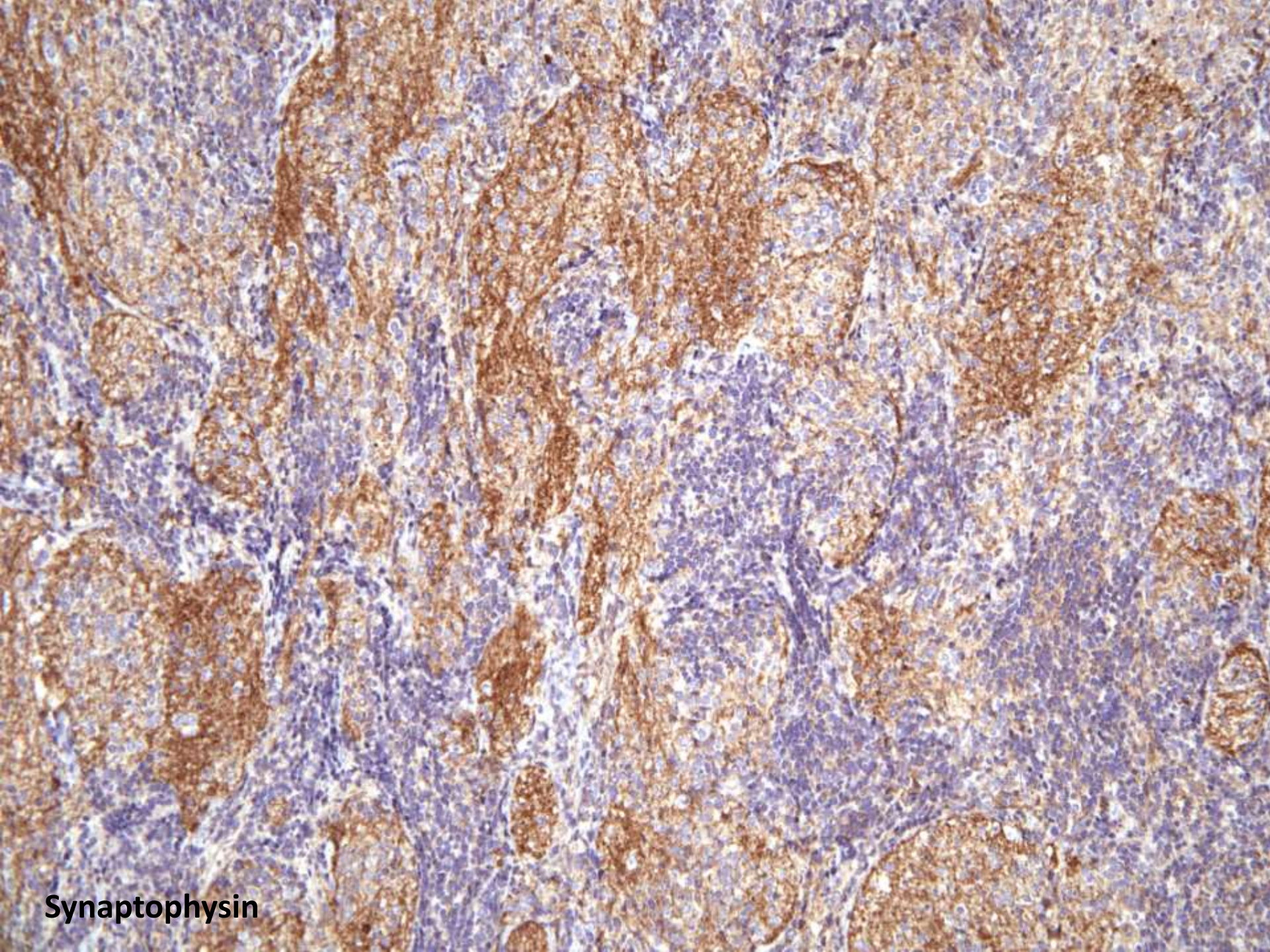






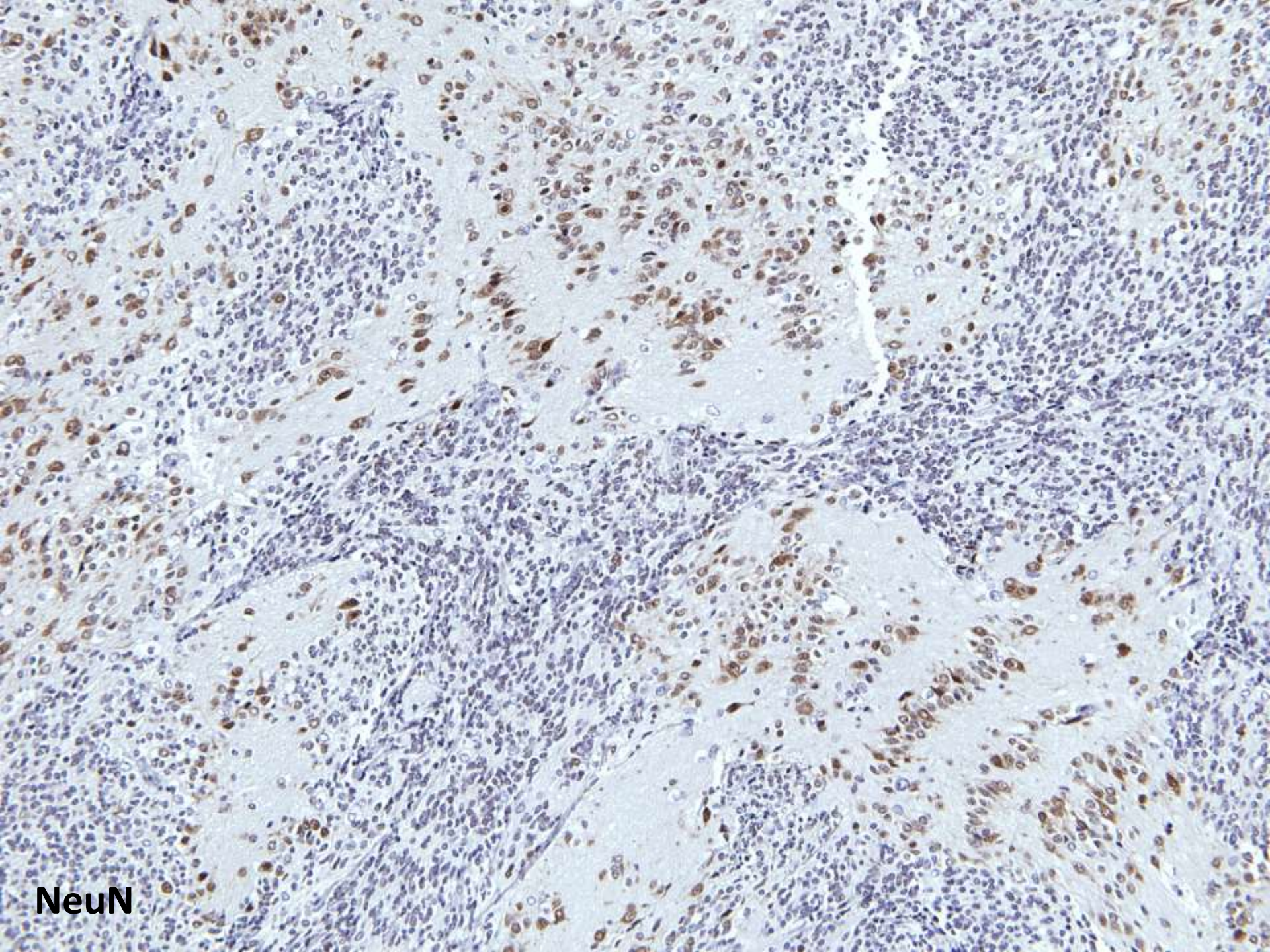






**Synaptophysin**





NeuN



# CNS Ganglioneuroblastoma

- One of the “other” embryonal tumors
- WHO grade IV
- Very rare (~10 cases reported)
- Surgery/chemo/radiation
- Survivals were reported in three of the 10 previously reported patients with the range of survival after initial symptoms was 0.7–36 months (median 11 months), which is substantially less than the 60% 3 year survival in pediatric patients with cerebral neuroblastomas

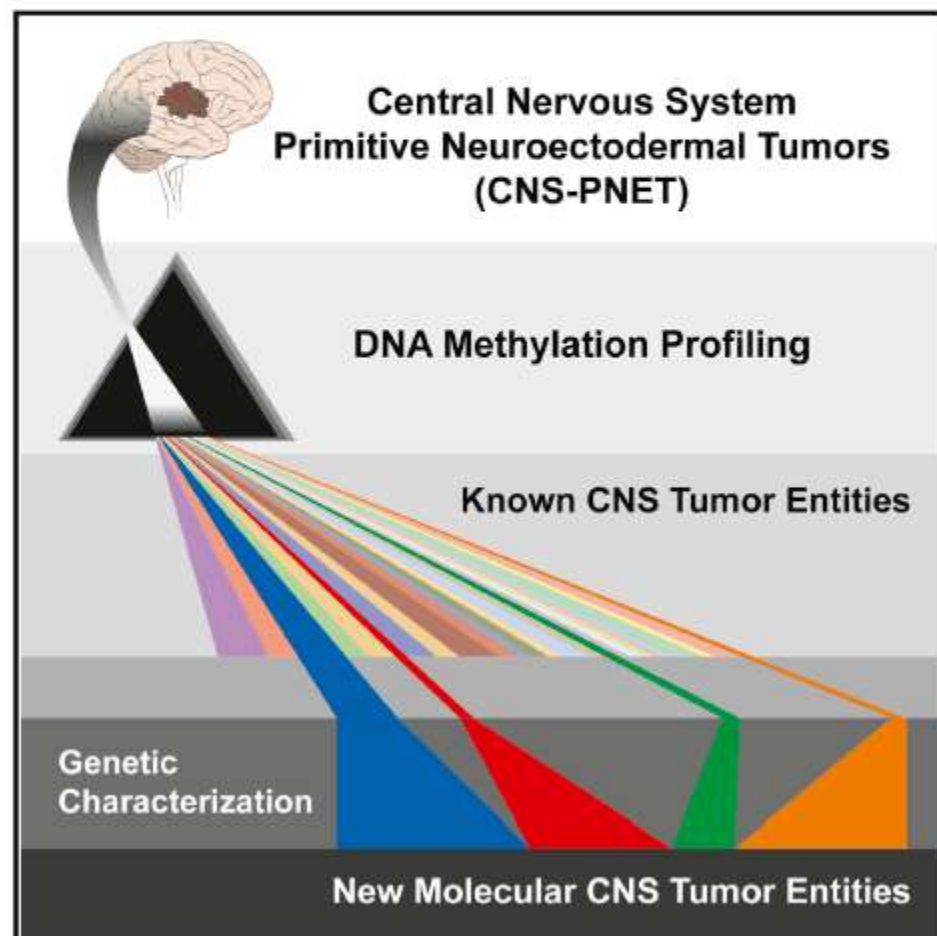
# Key Diagnostic Points

- An embryonal tumor with divergent morphologies and varying degrees of neuronal differentiation – poorly differentiated neuroepithelial cells, groups of neurocytic cells, and ganglion cells
- Dysplastic ganglion cells are a prominent element in small groups rather than dispersed



# New Brain Tumor Entities Emerge from Molecular Classification of CNS-PNETs

## Graphical Abstract



## Authors

Dominik Sturm, Brent A. Orr, Umut H. Toprak, ..., David W. Ellison, Andrey Korshunov, Marcel Kool

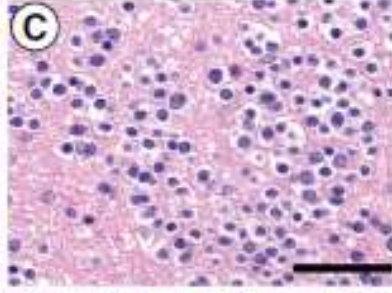
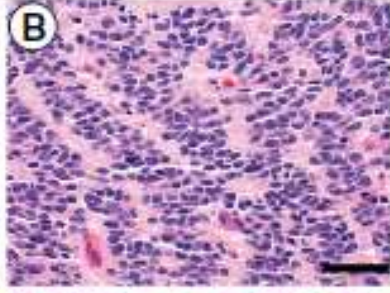
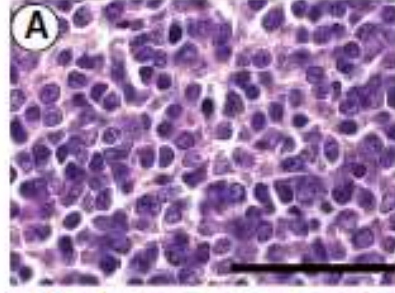
## Correspondence

m.kool@dkfz.de

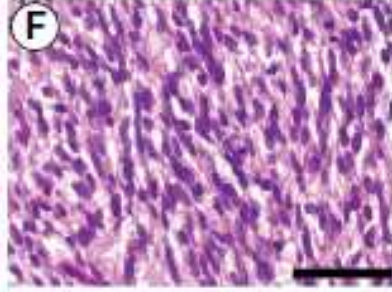
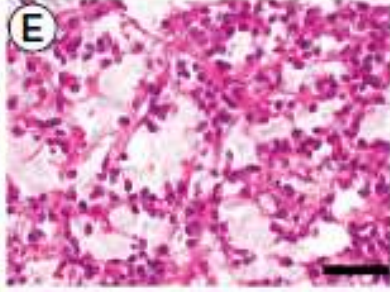
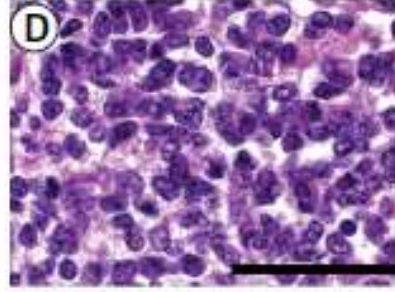
## In Brief

Highly malignant primitive neuroectodermal tumors of the CNS (CNS-PNETs) have been challenging to diagnose and distinguish from other kinds of brain tumors, but molecular profiling now reveals that these cancers can be readily classified into some known tumor types and four new entities with distinct histopathological and clinical features, paving the way for meaningful clinical trials.

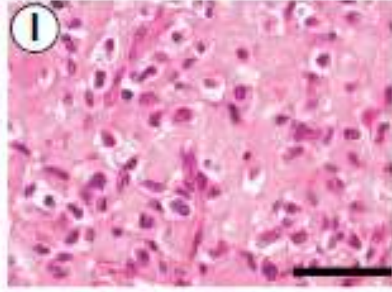
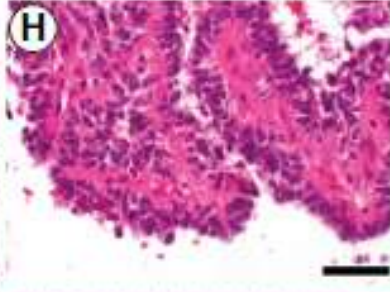
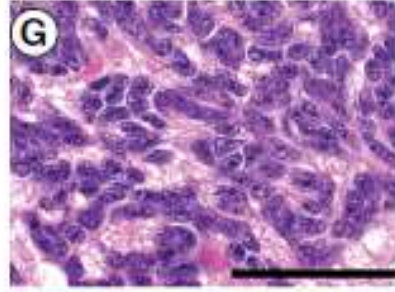
CNS  
NB-FOXR2



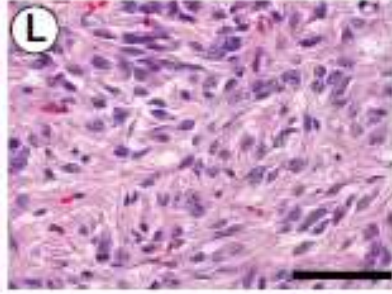
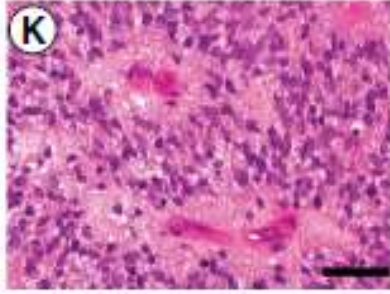
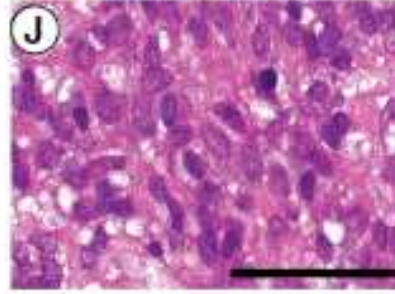
CNS  
EFT-C/C



CNS  
HGNET-MN1



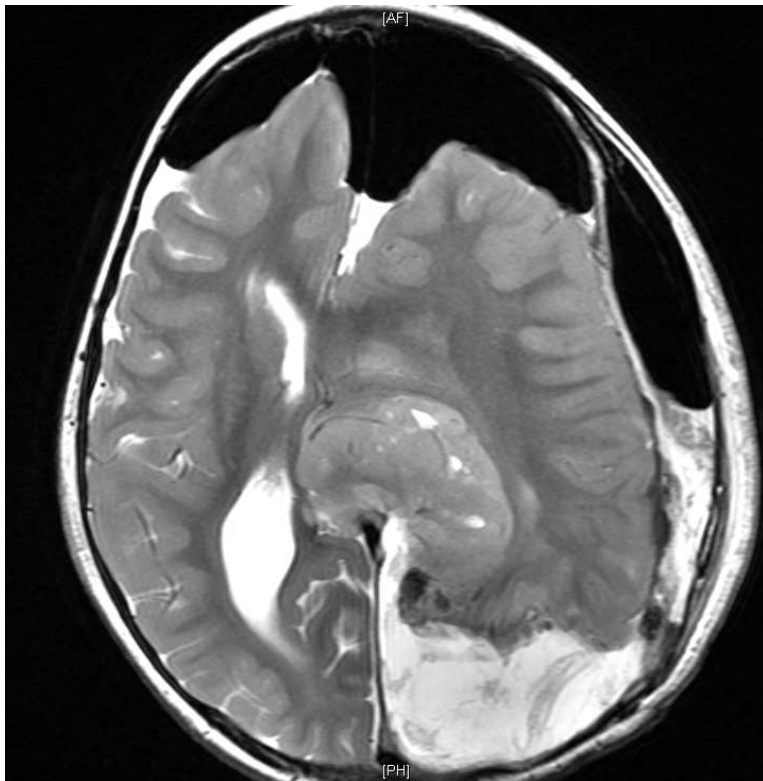
CNS  
HGNET-BCOR





# Clinical follow-up

Post-op 9/10



12/5



# Clinical follow-up

- Doing well overall
- CSF from an LP on 9/19/16 was negative for metastatic disease. Post operatively Valeria had right facial paresis and right hemiparesis, which improved over the last few months.
- Enrolled on St. Jude study SJYC07 and began induction therapy on 9/23/16 and completed therapy on 1/3/17.
- Second resection last week

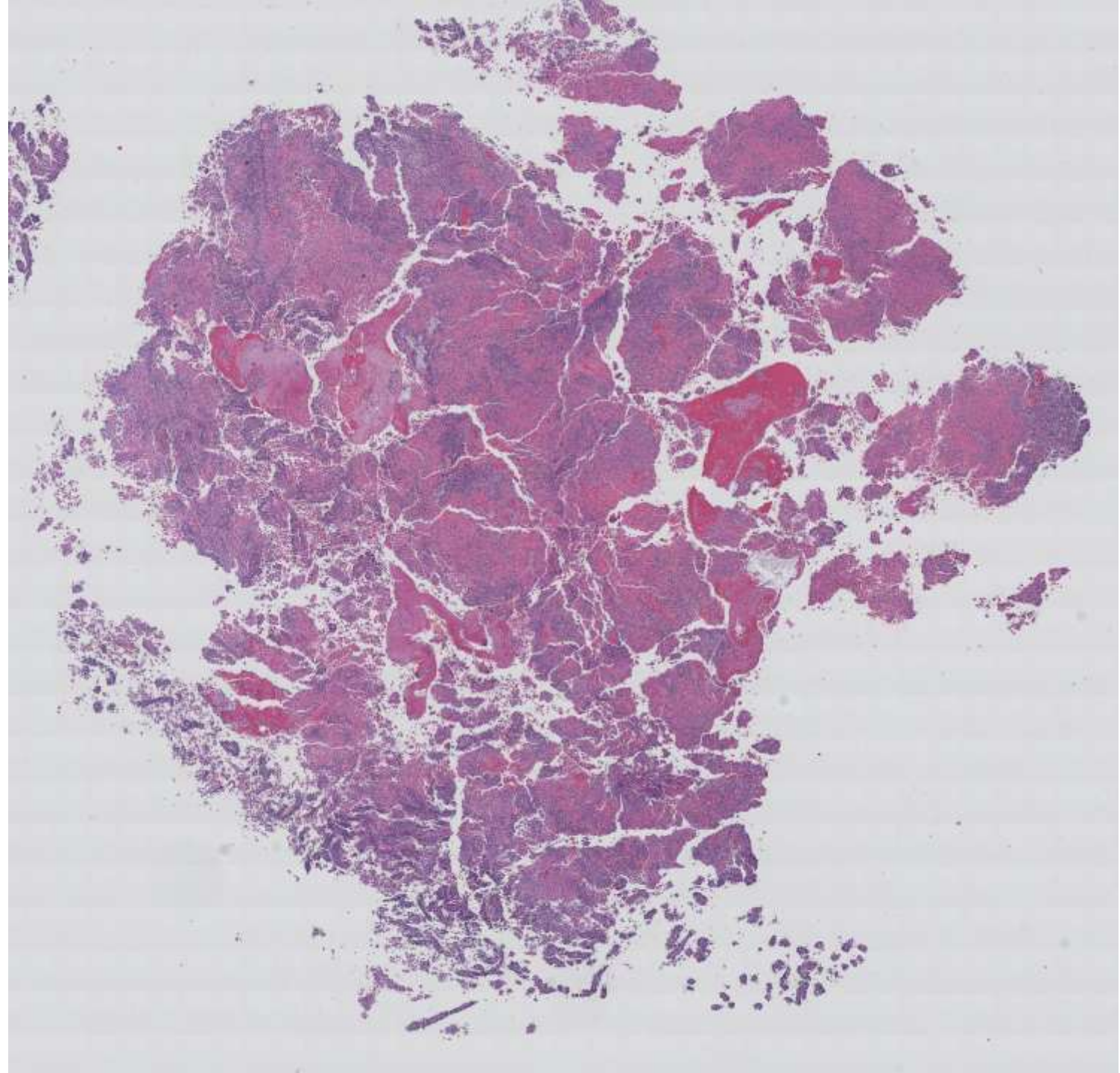


# SB 6137

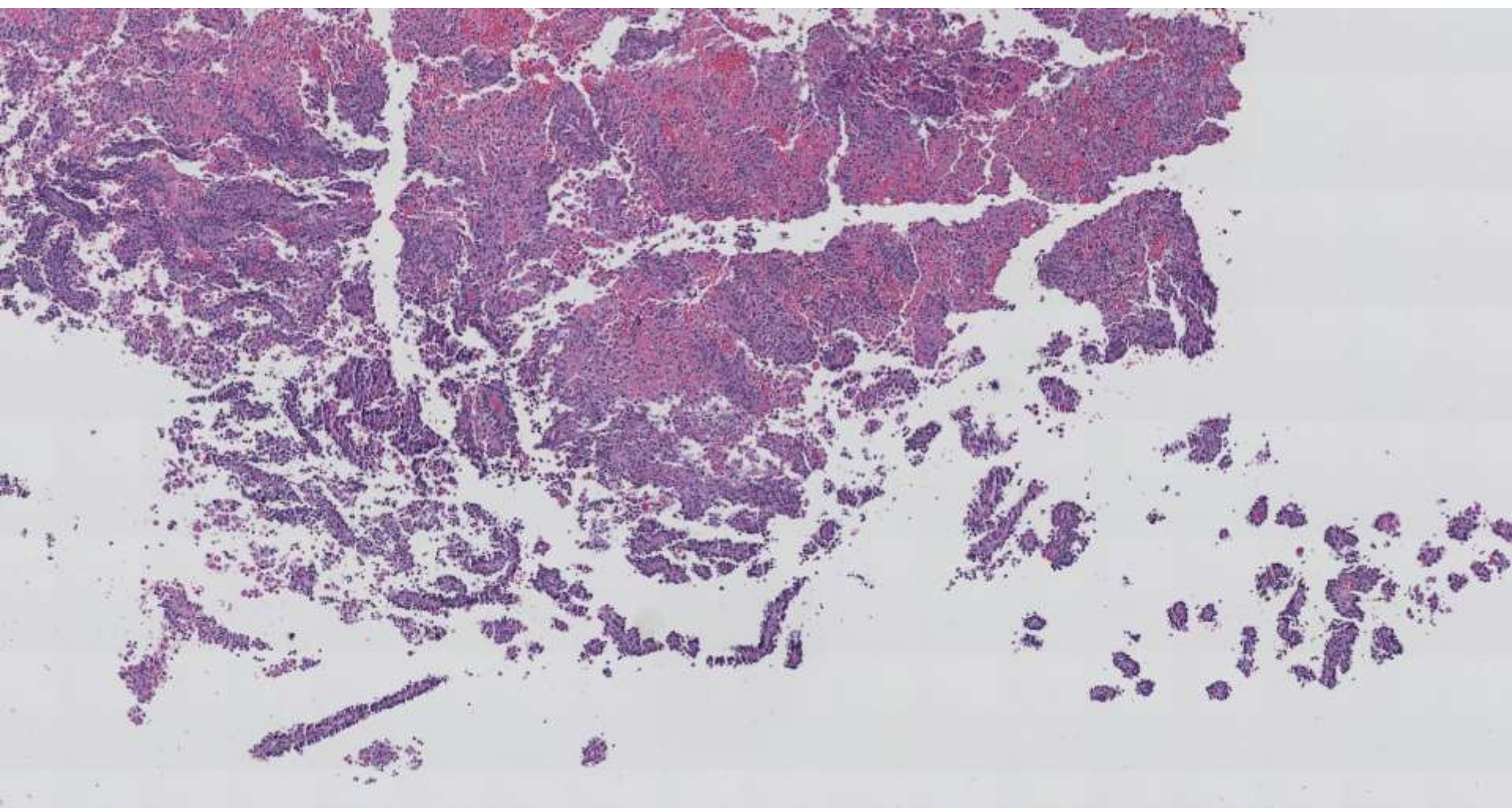
## (scanned slide available)

**Ben Buelow/Julieann Lee; UCSF**

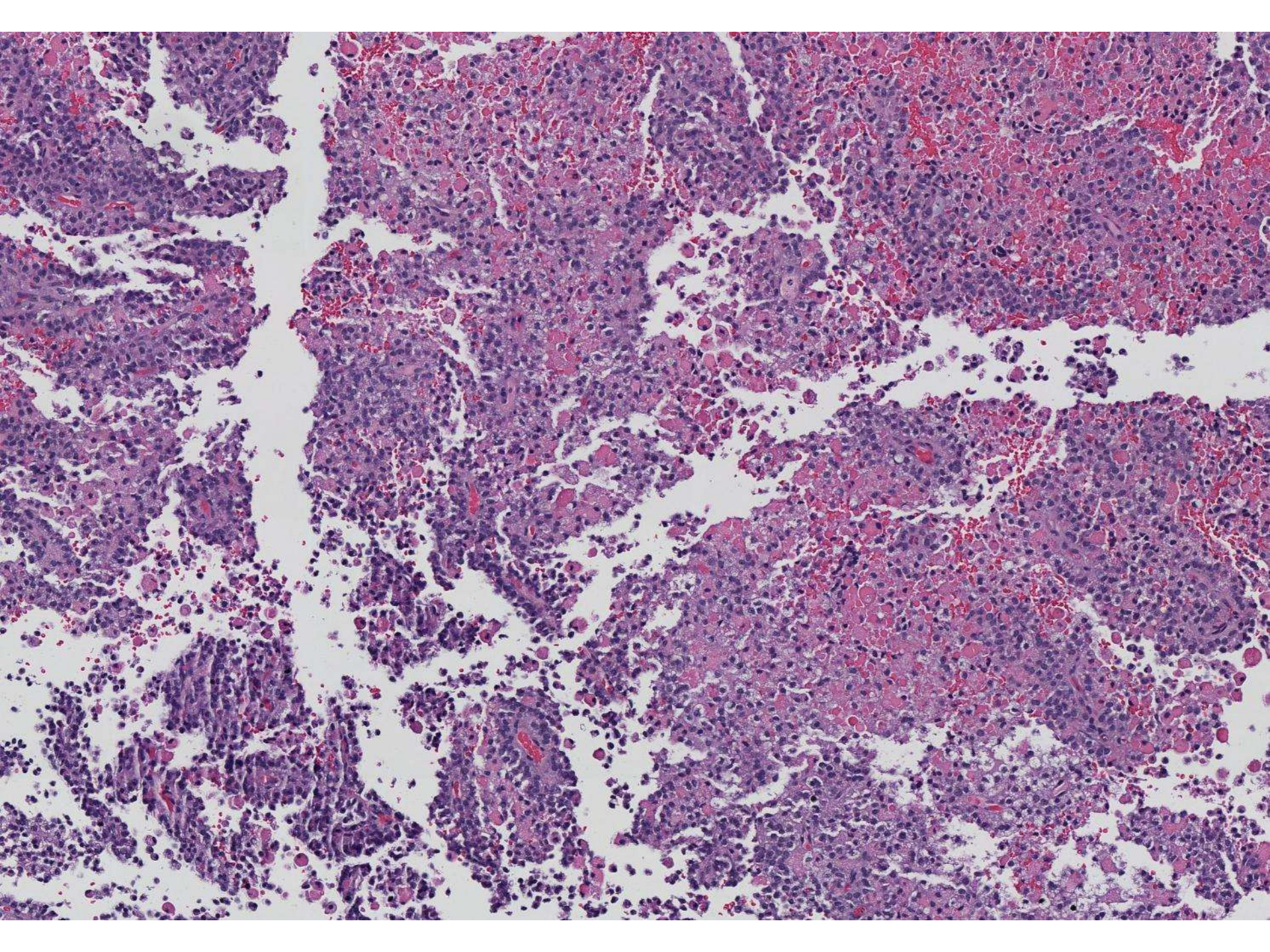
16-year-old female who presented with an abdominal mass. Intraoperatively she had an 18cm smooth ovarian mass composed of homogenous tan-white focally hemorrhagic tissue. The patient's AFP and B-HCG were markedly elevated. Frozen section of the main mass showed sheets of monotonous large moderately cohesive polygonal cells with nucleomegaly, coarse chromatin, and variably prominent nucleoli. A brisk lymphocytic infiltrate was present. A periaortic LN submitted for frozen section was negative. A second peripancreatic LN was submitted for permanent section and is shown here.



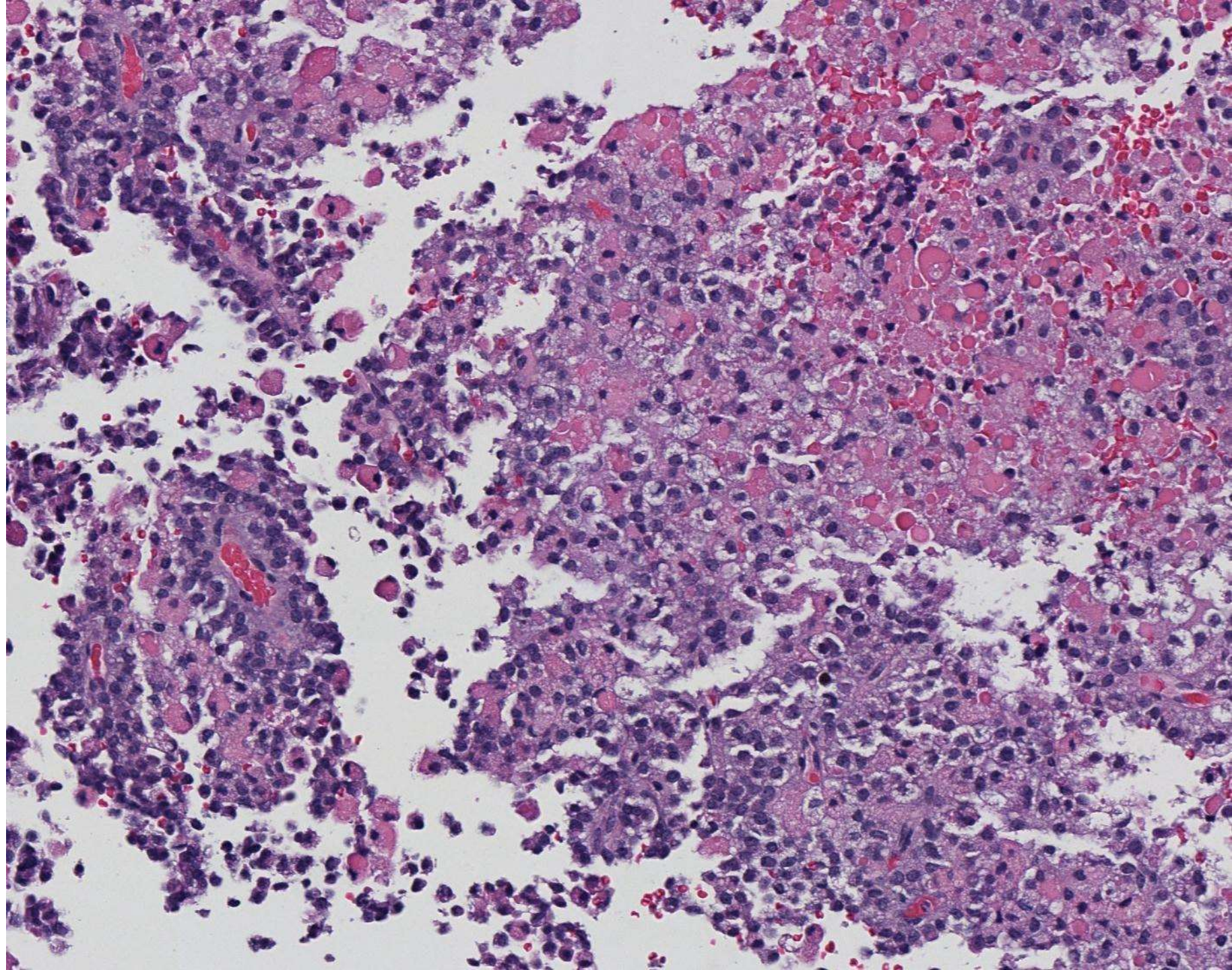




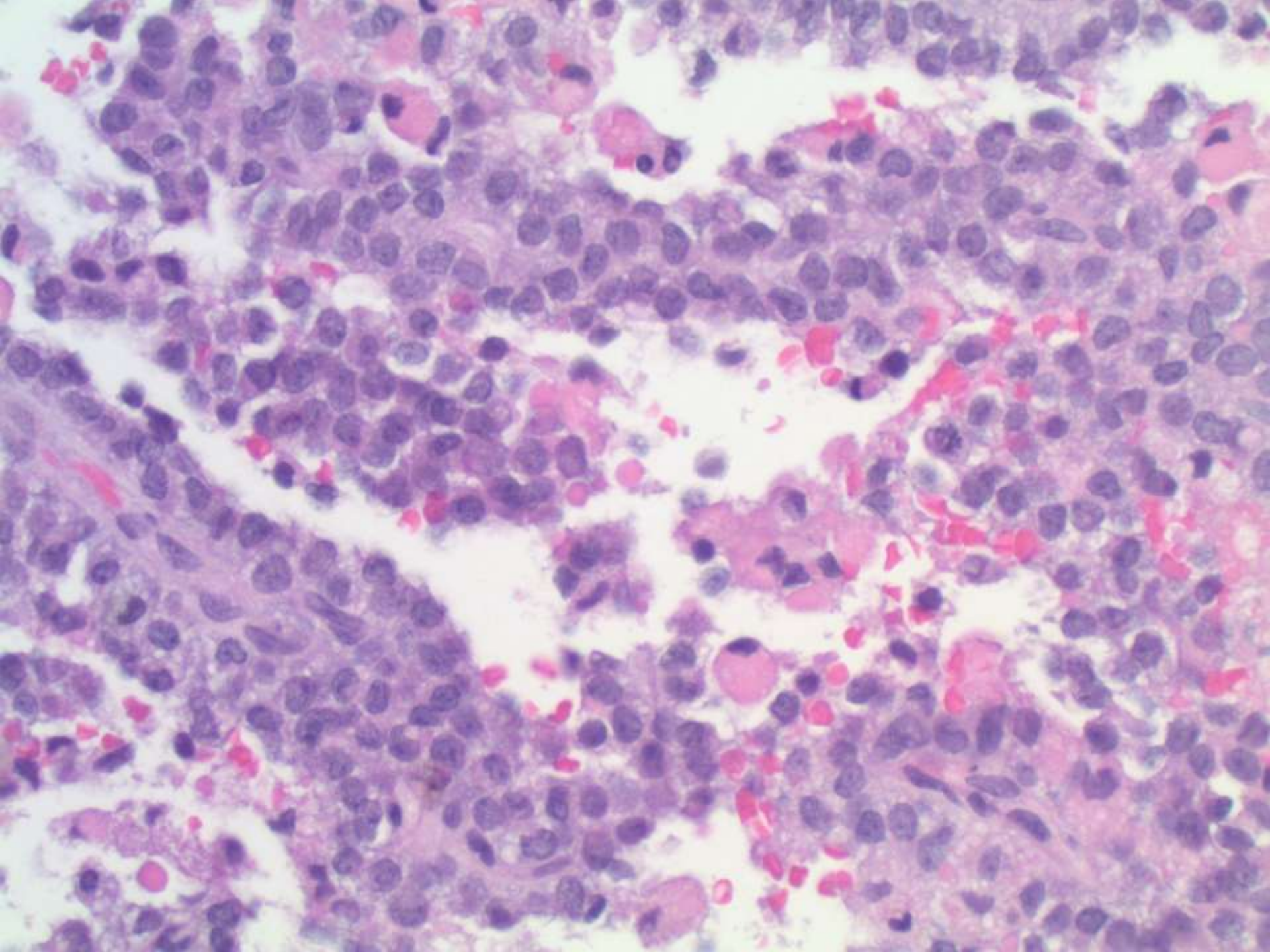












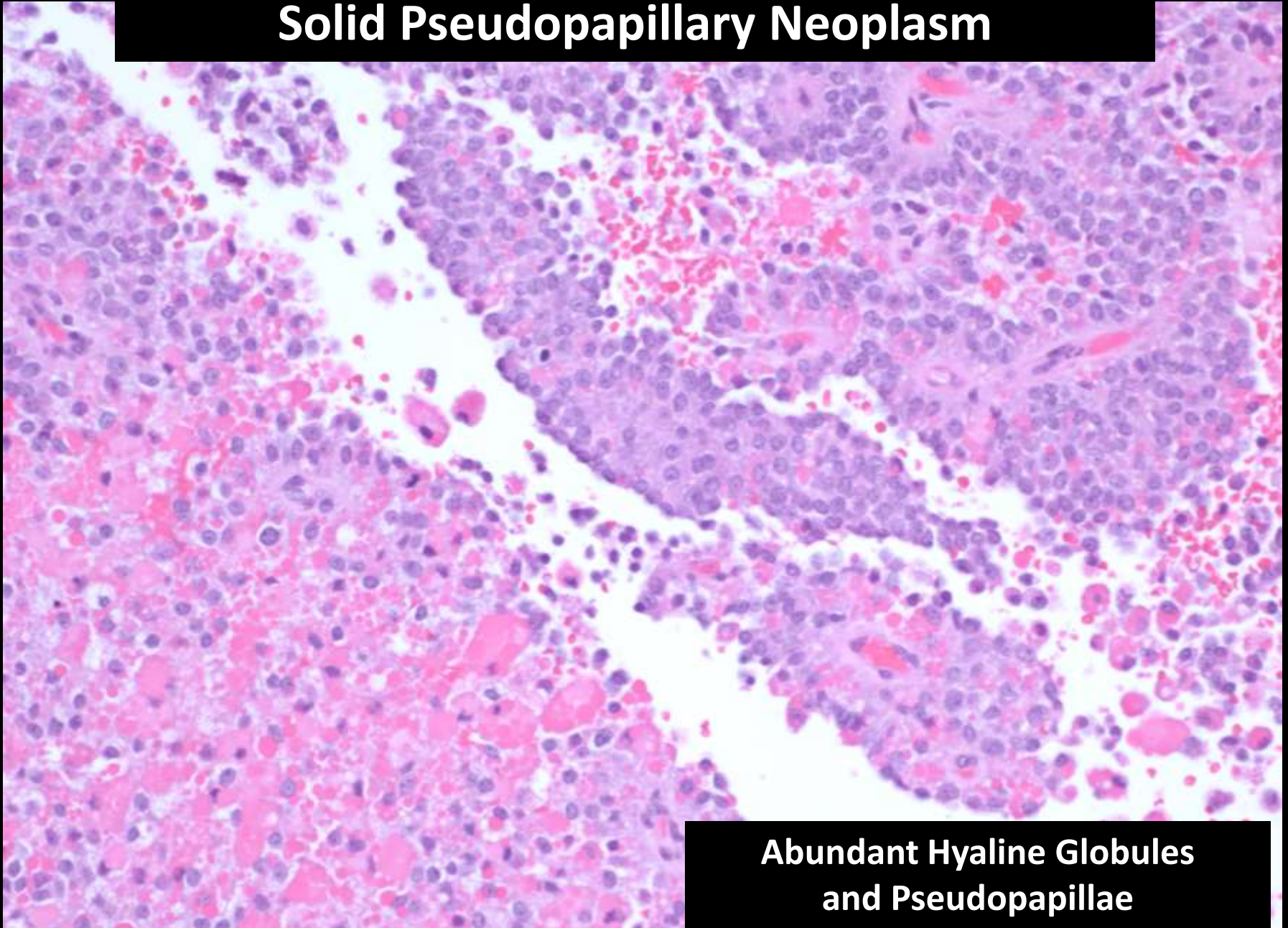


# **16-year-old female with a 16 cm right ovarian mass, and “peripancreatic lymphadenopathy”**

Julieann Lee & Ben Buelow  
UCSF

South Bay Pathology Society  
February 6, 2017

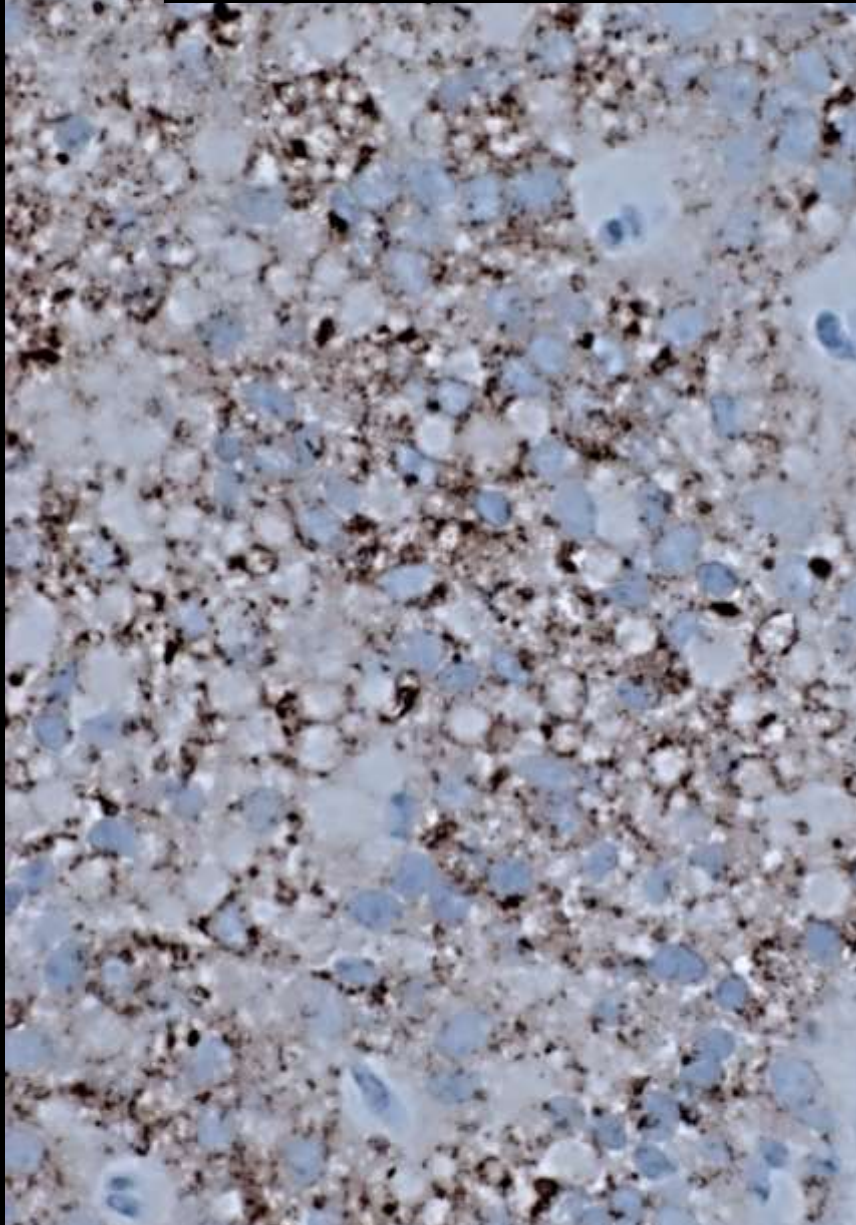
# Solid Pseudopapillary Neoplasm



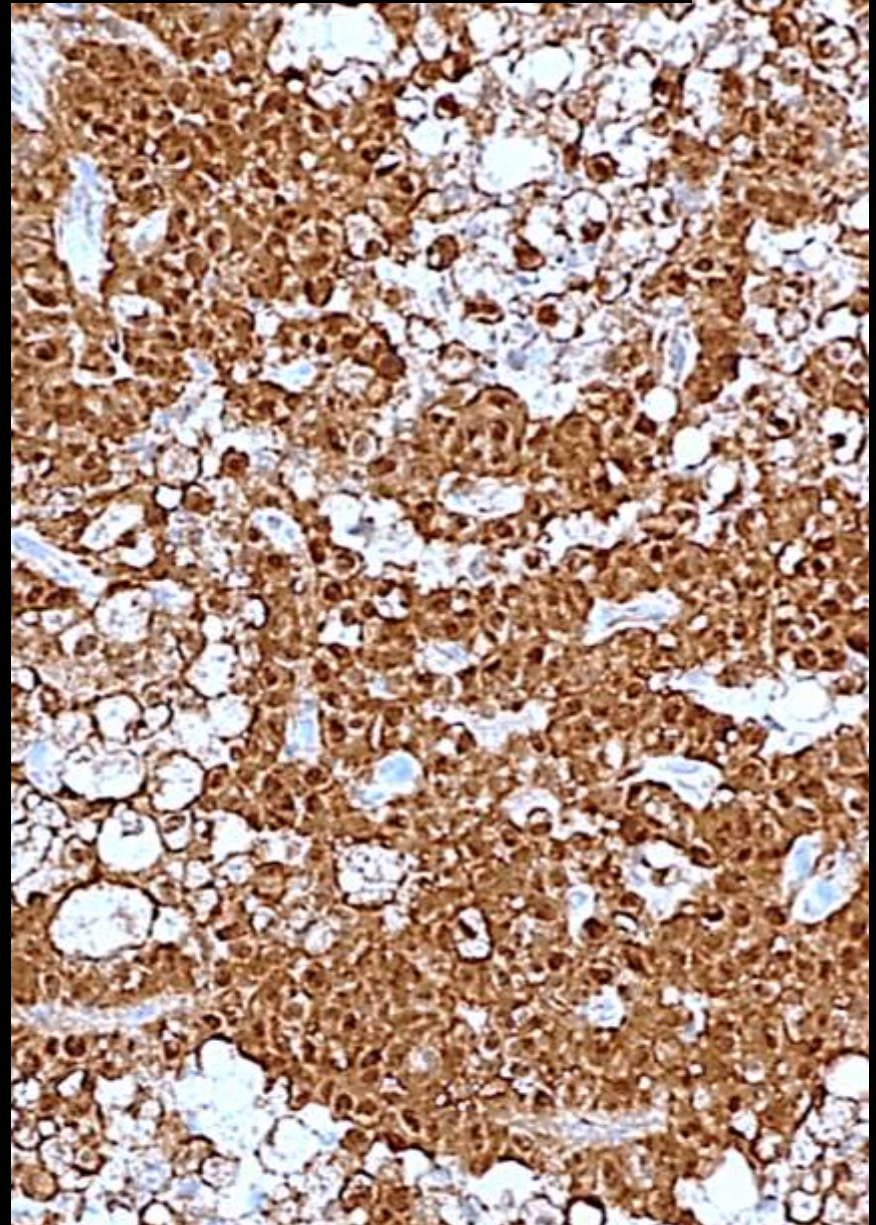
**Abundant Hyaline Globules  
and Pseudopapillae**



# Solid Pseudopapillary Neoplasm

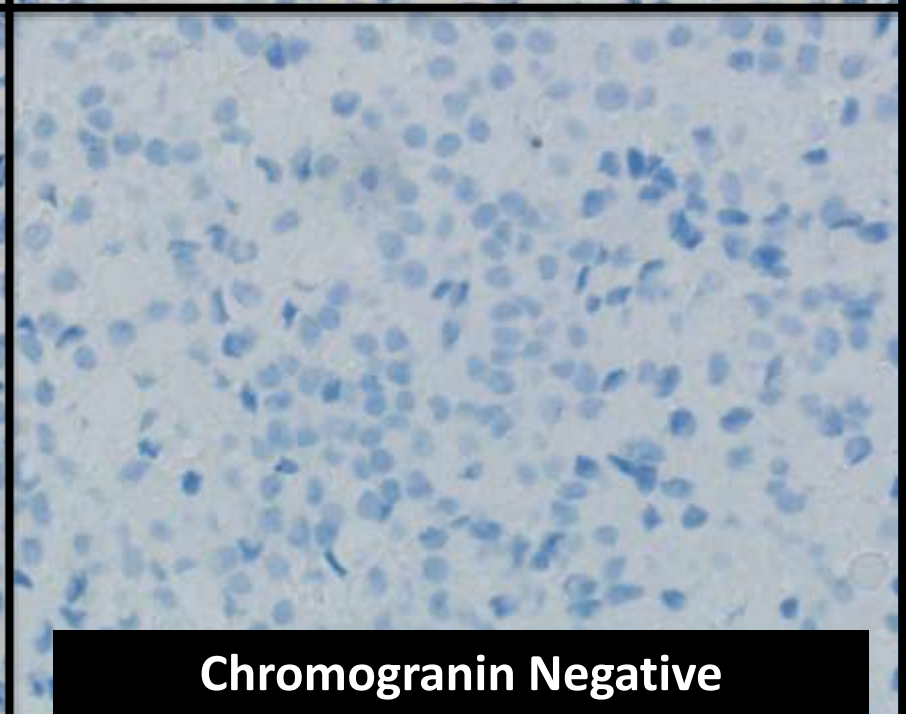
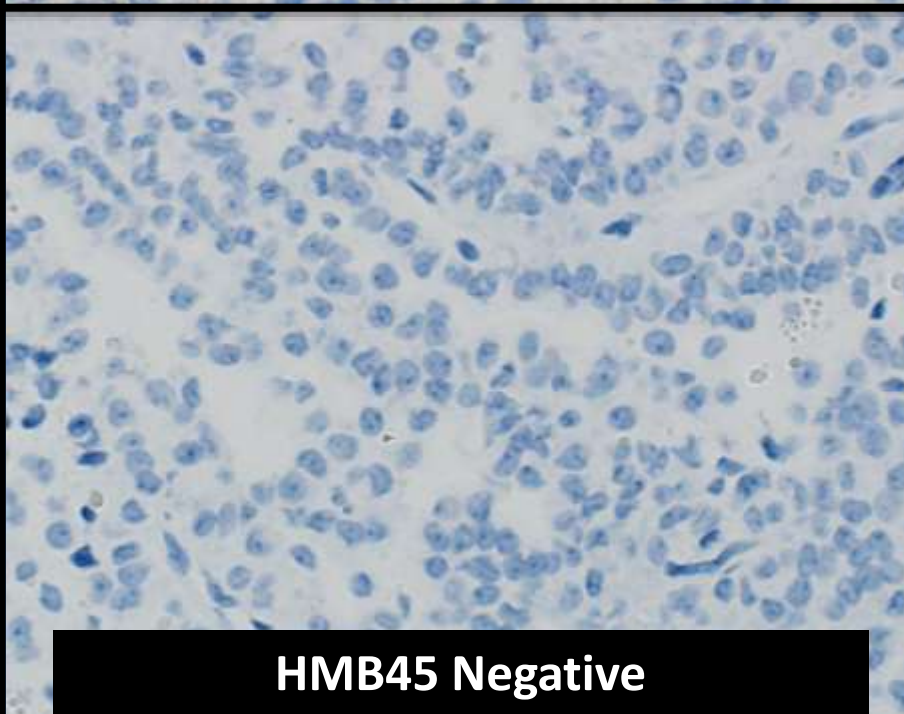
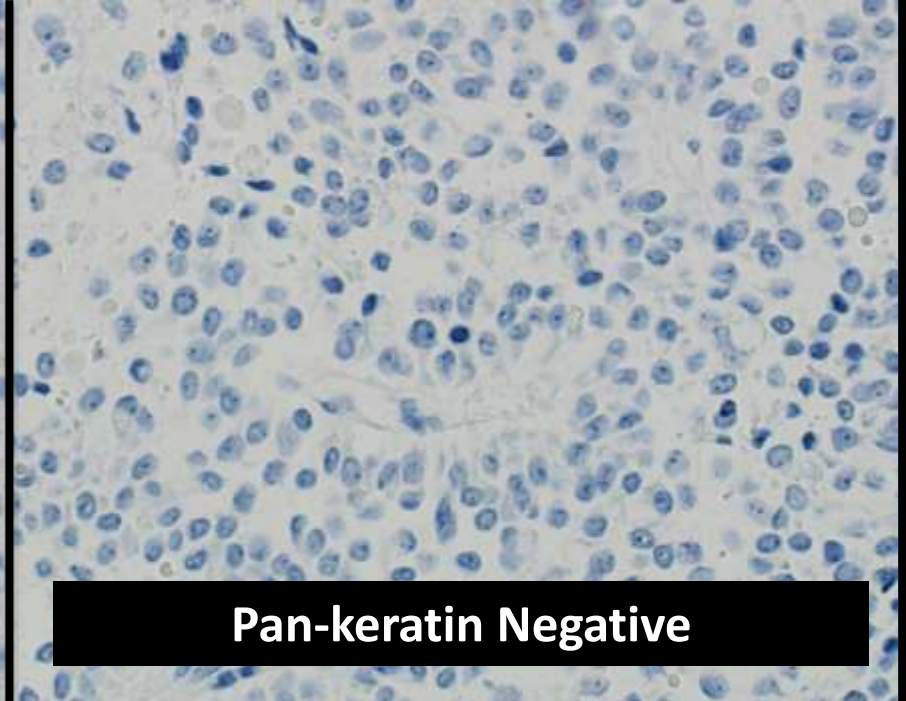
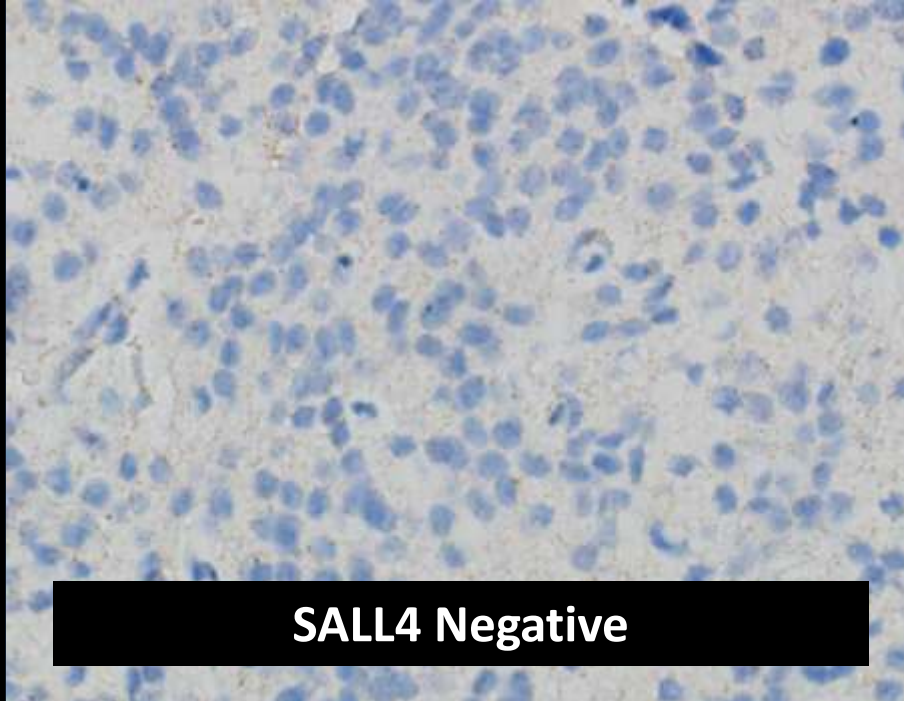


**CD10**

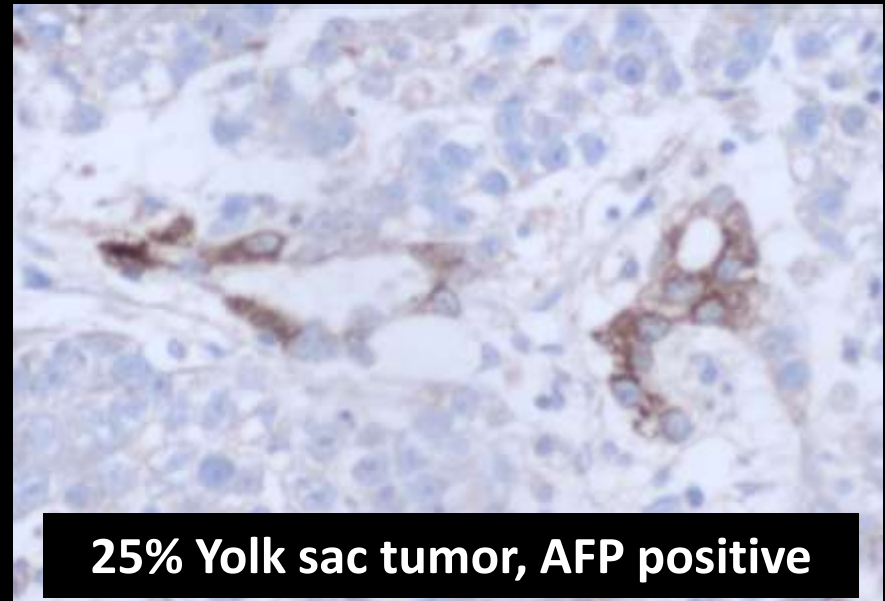
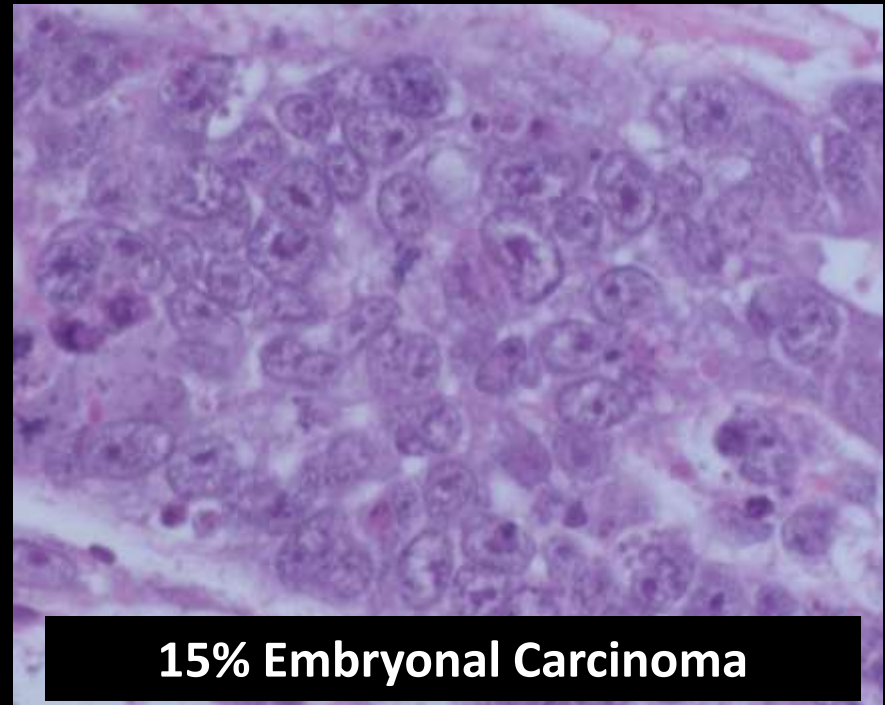
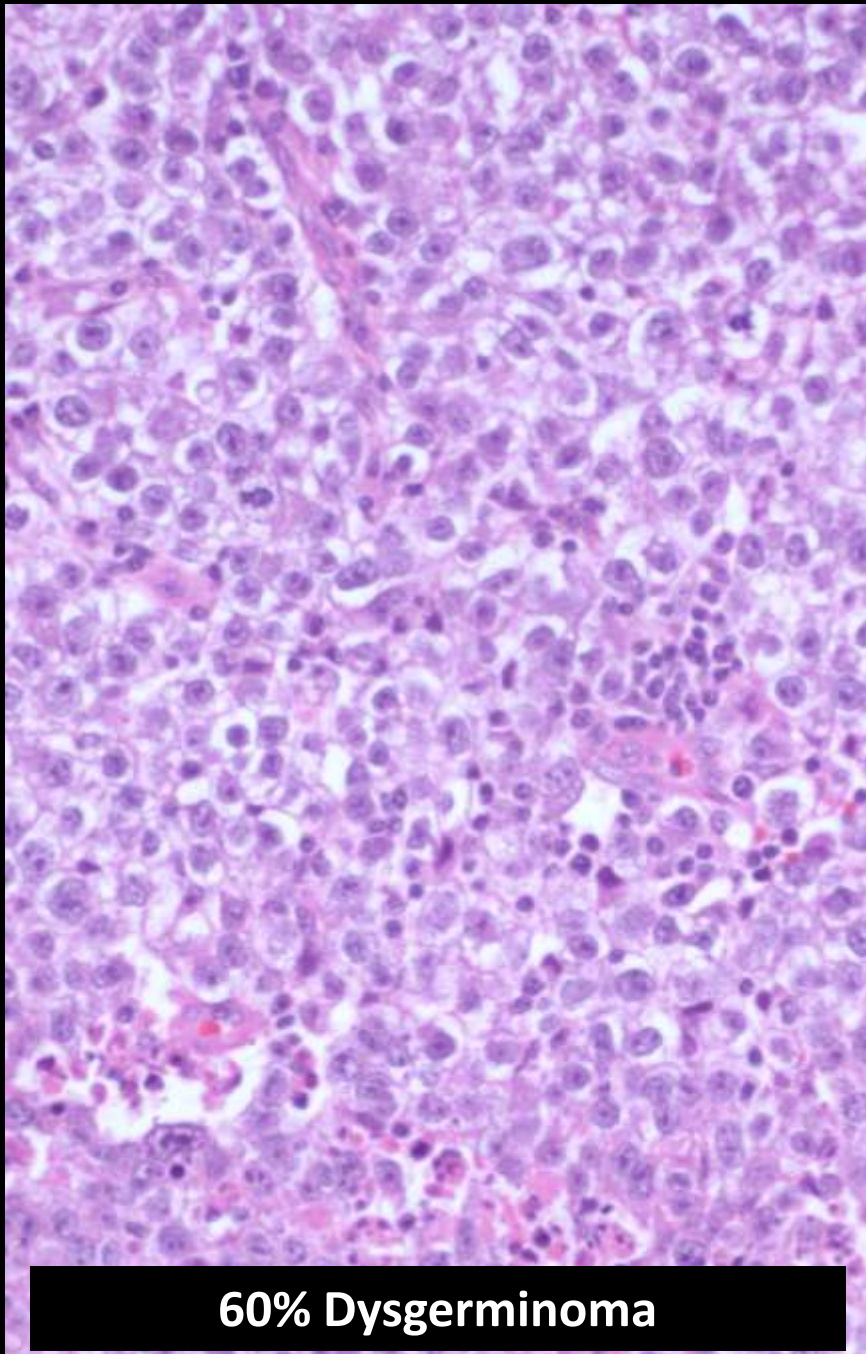


**B-catenin**









# Solid Pseudopapillary Neoplasm of the Pancreas

- Occur predominantly in young women
- Usually discovered incidentally
- Rare: ~2% of all exocrine pancreatic neoplasms
- Metastasis in ~10% of cases
- Rarely can directly extend into stomach, duodenum, and spleen
- 85% cases cured with complete surgical resection
- Origin outside of the pancreas is uncommon, has been reported in retropancreatic tissue



# Solid Pseudopapillary Neoplasm of the Pancreas

- Solid to cystic, often mixed
- Poorly cohesive monomorphic cells
- Pseudopapillae
- Eosinophilic hyaline globules
- Often have nuclear grooves
- No morphologic predictors of outcome
- Nuclear/cytoplasmic B-catenin, and CD10 positive

# SB 6138

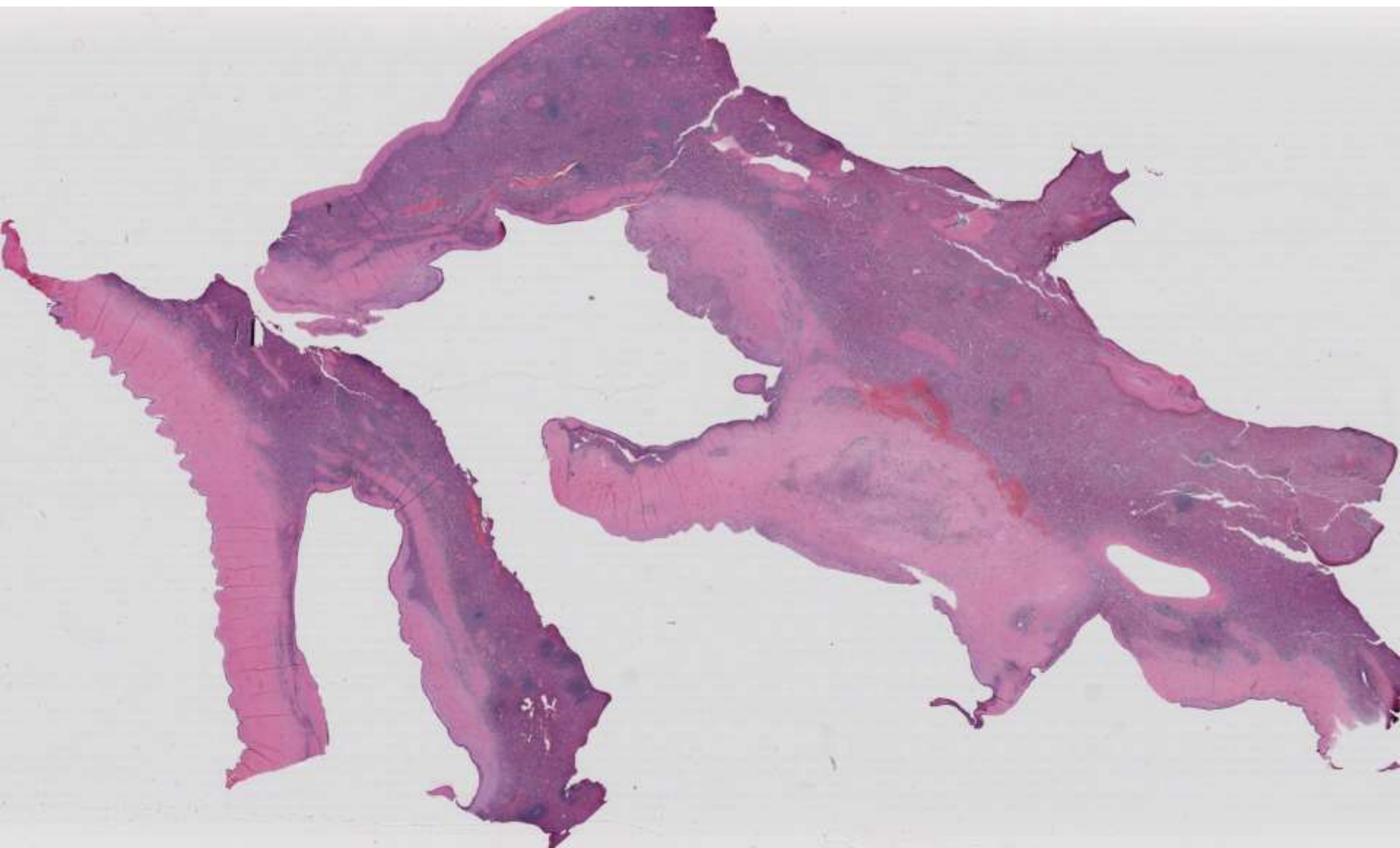
## (scanned slide available)

**Ankur Sangoi; El Camino Hospital**

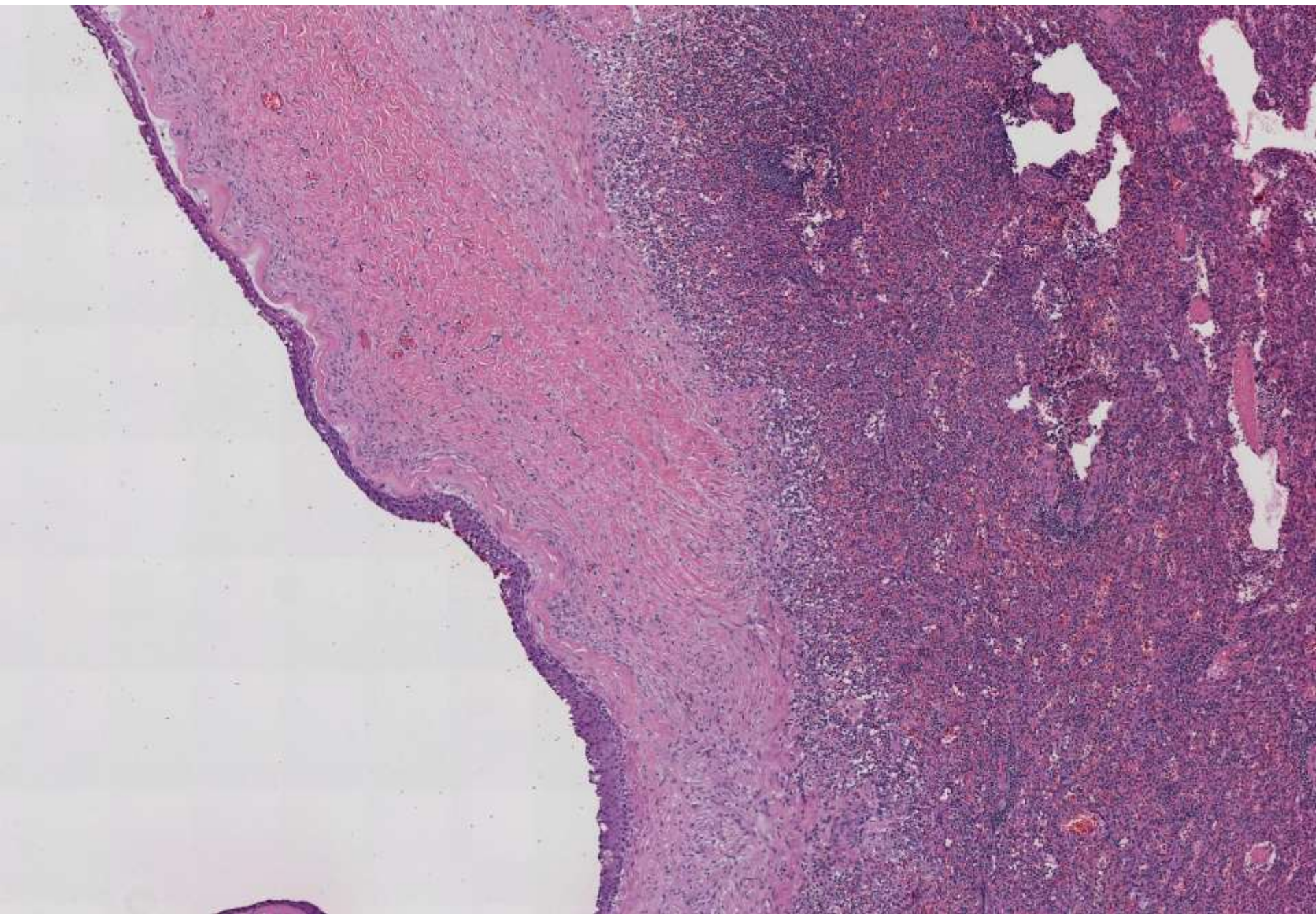
25-year-old female pancreas mass and elevated CA19-9. Distal pancreatectomy with partial resection of surrounding colon/stomach/spleen performed.



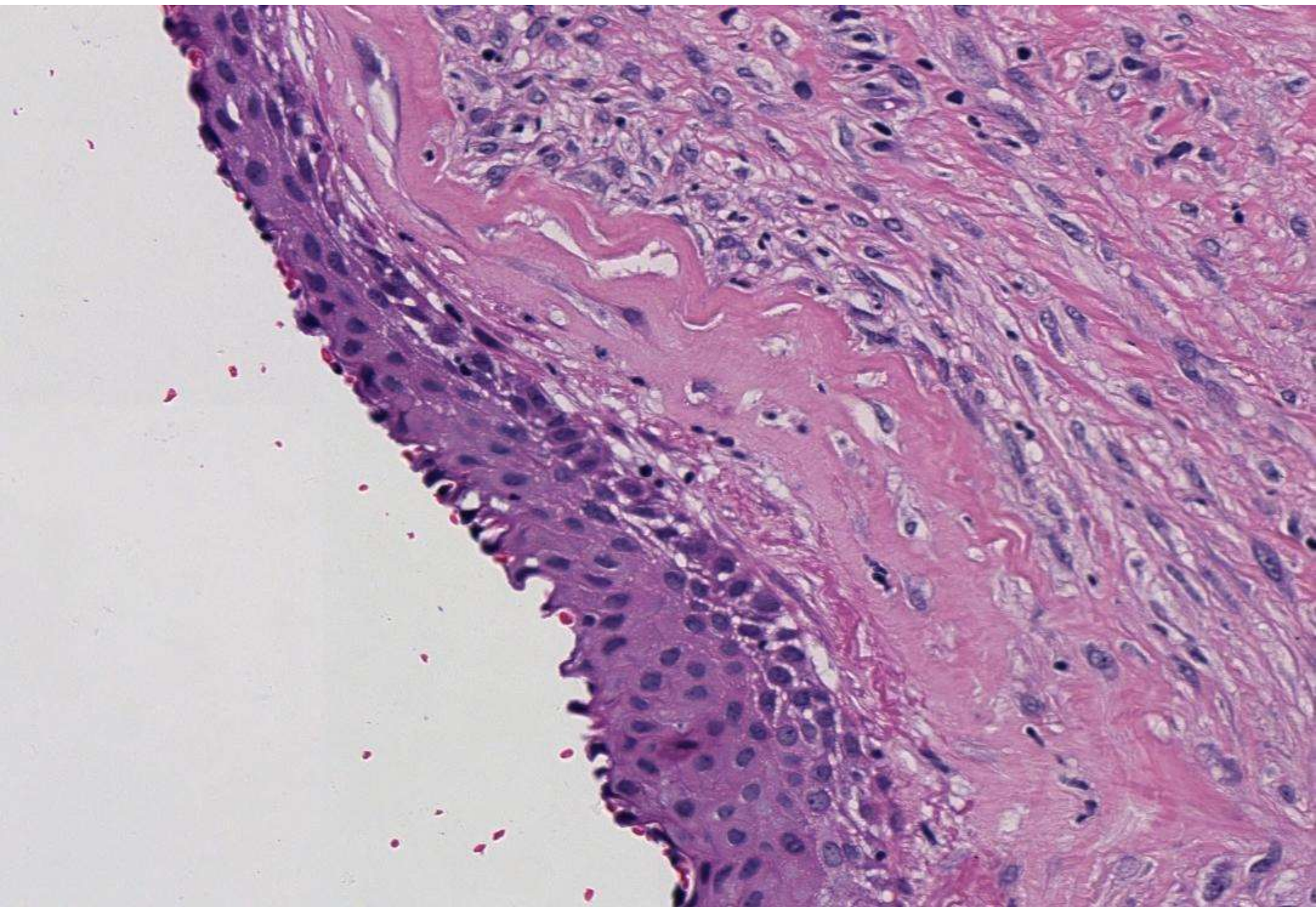




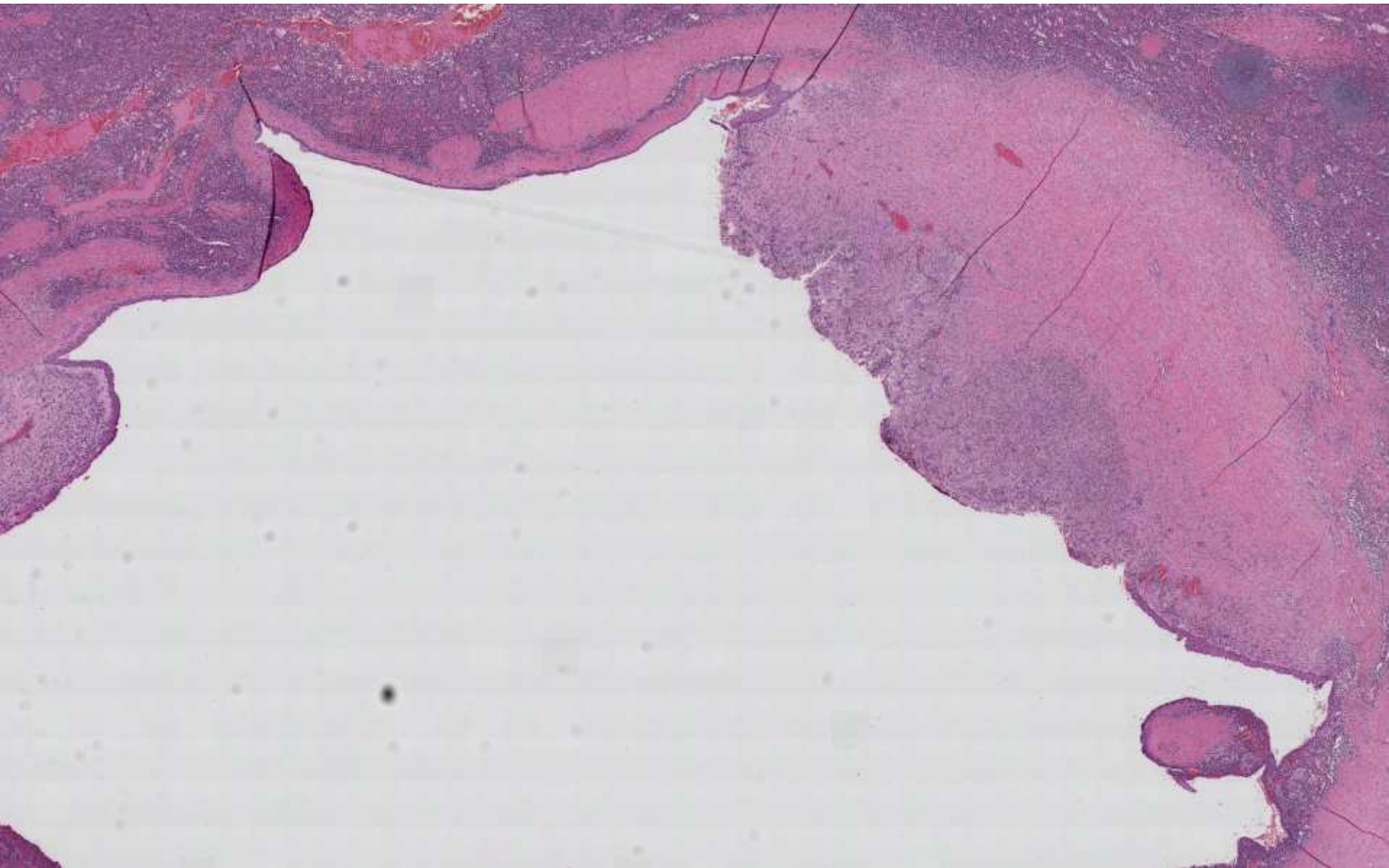




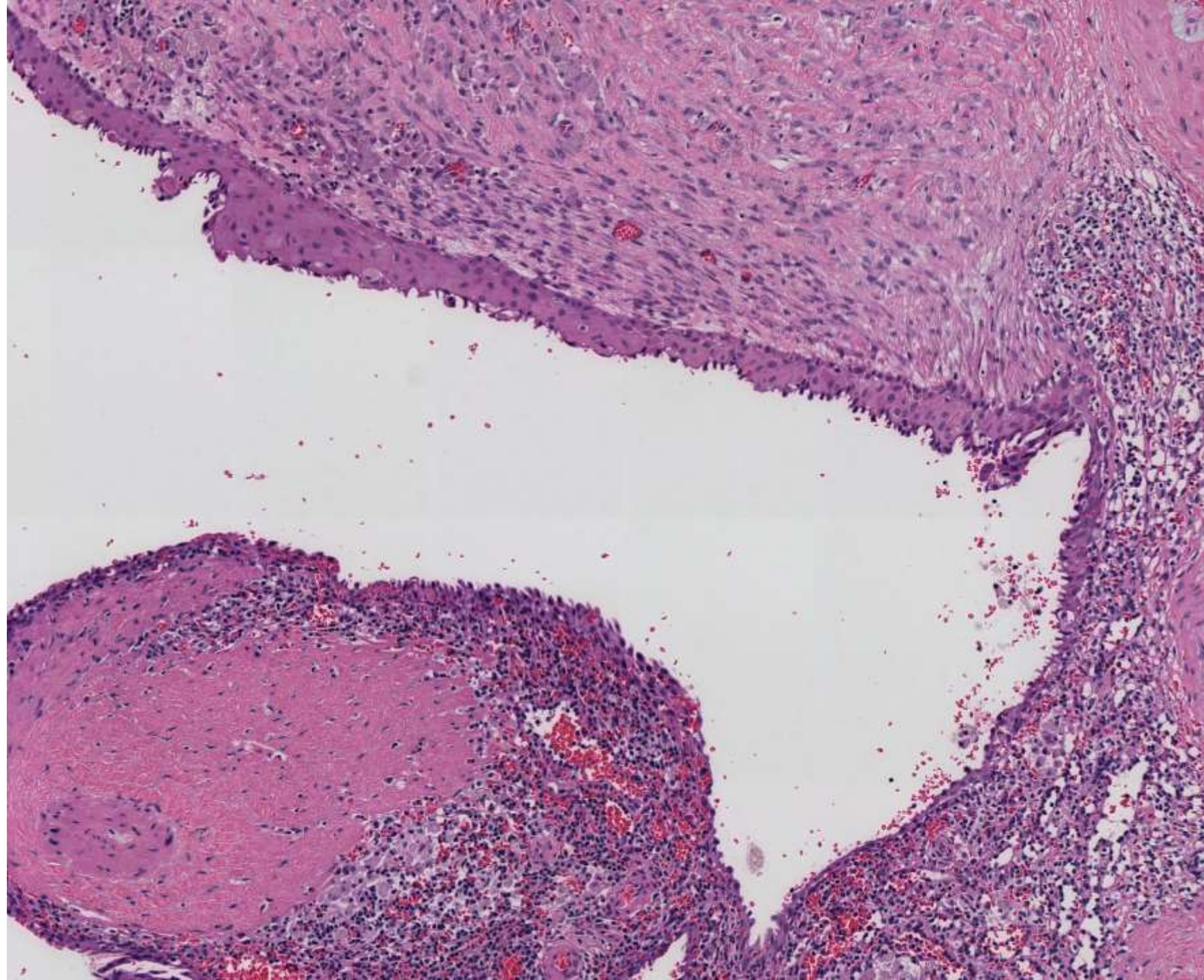




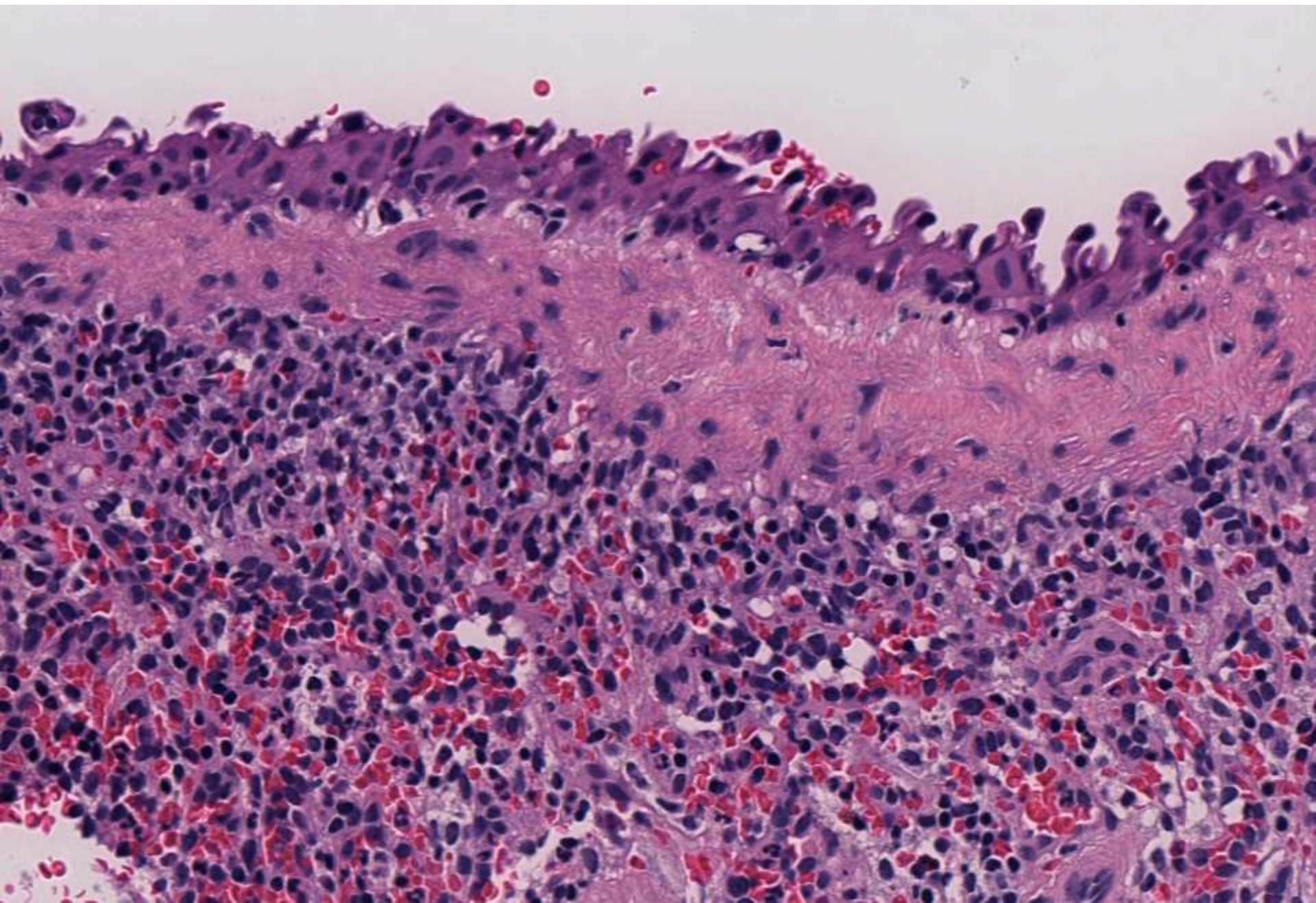












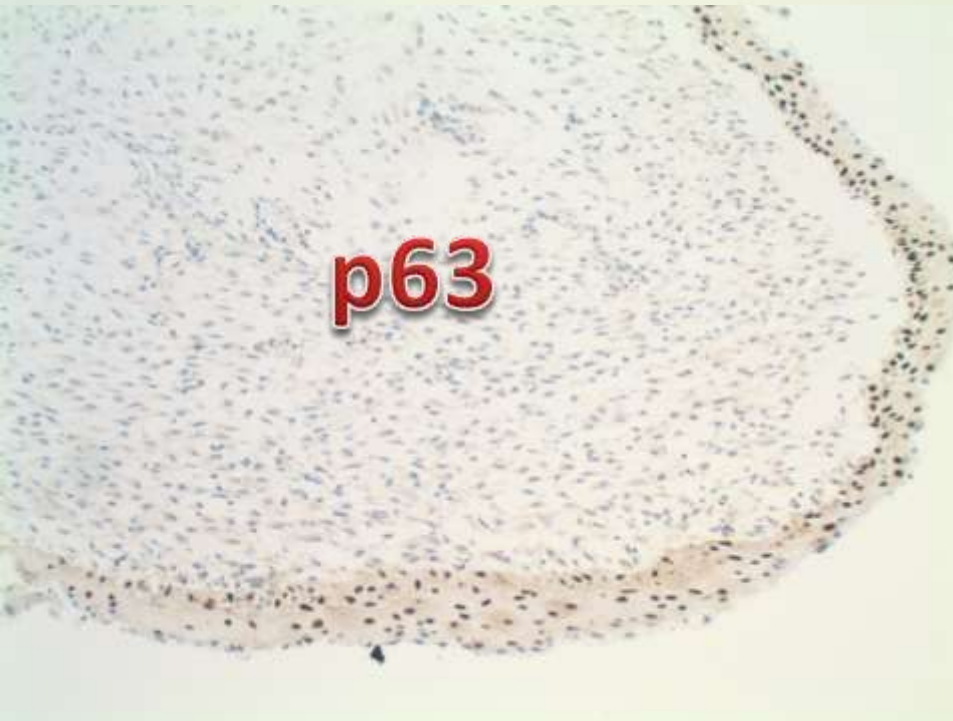
# DIAGNOSIS?





# DDx of splenic cysts

- Echinococcal cyst
- epithelial cyst
- dermoid cyst
- mesothelial cyst
- pseudocyst
- Vascular lesion (lymphangioma)





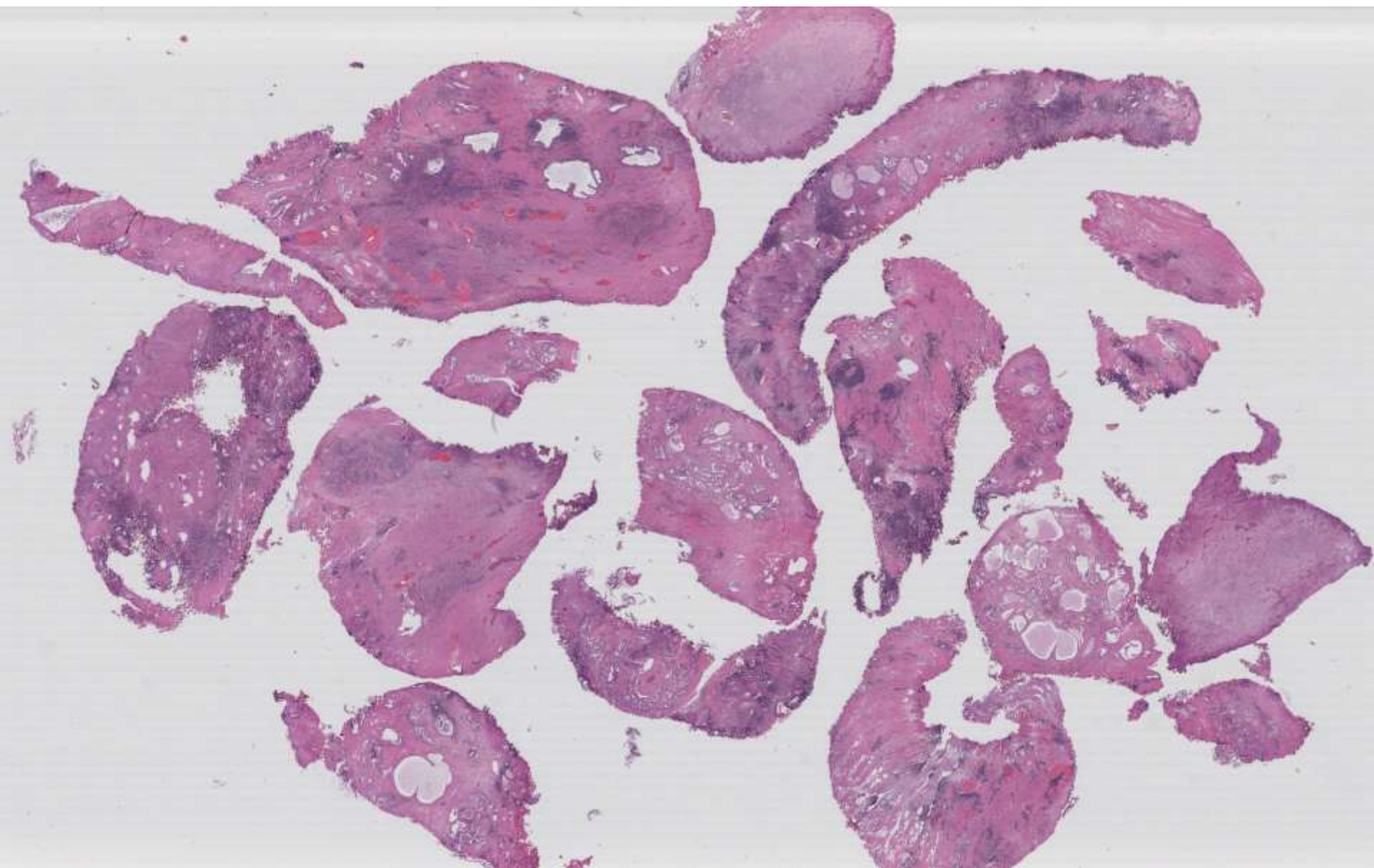
# DX: SPLENIC EPITHELIAL CYST ("splenic epidermoid cyst")

- Uncommon
- Etiology unknown
  - May arise from metaplasia in mesothelial cysts
- Usually children/young adults
- Solitary or multiple
  - May be associated with accessory spleen
- Often large and require splenectomy
- Can be associated with elevated serum CEA and CA19-9 levels

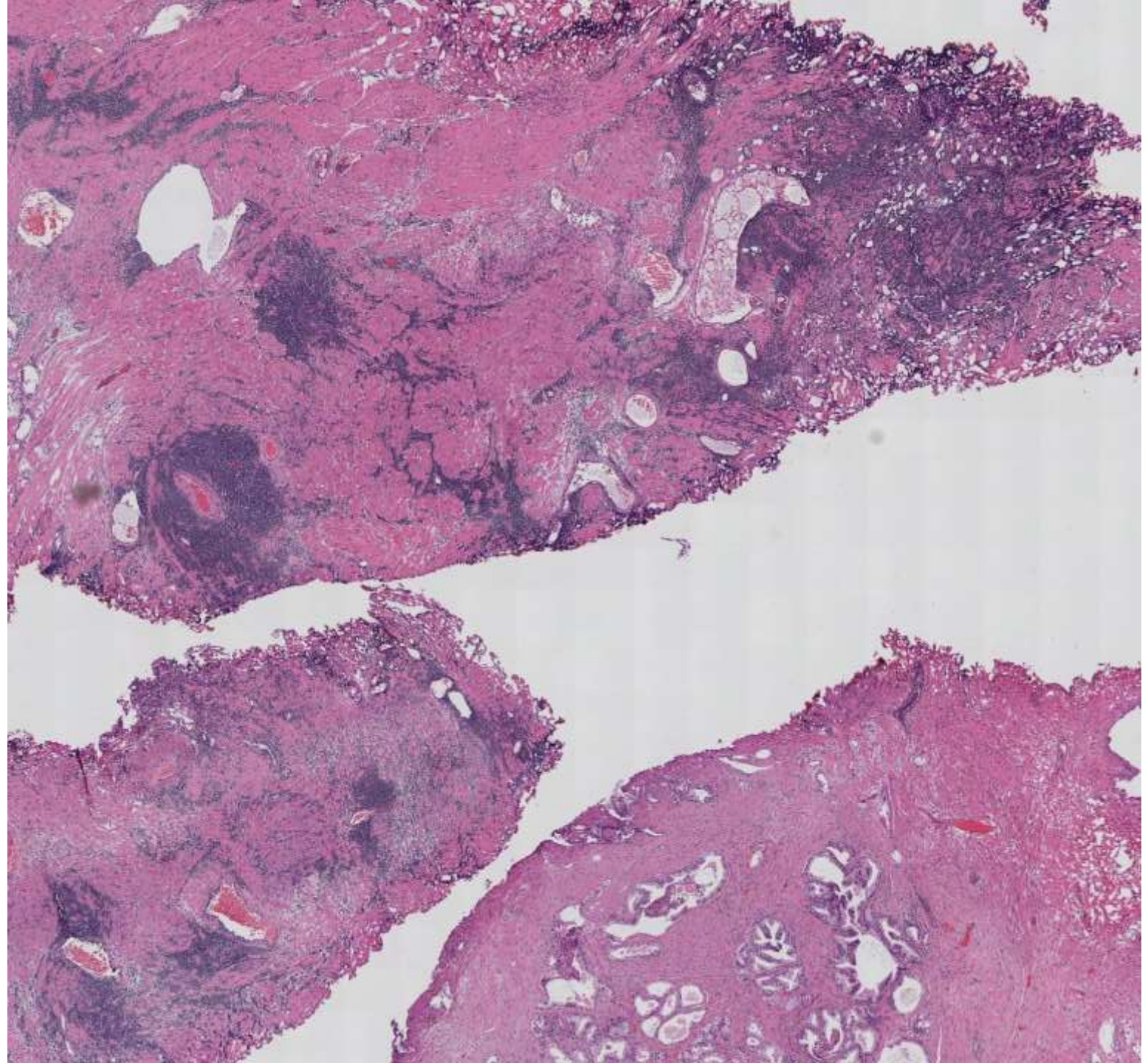
**SB 6139**  
**(scanned slide available)**

**Ankur Sangoi; El Camino Hospital**  
73-year-old male with BPH. TURP  
performed.

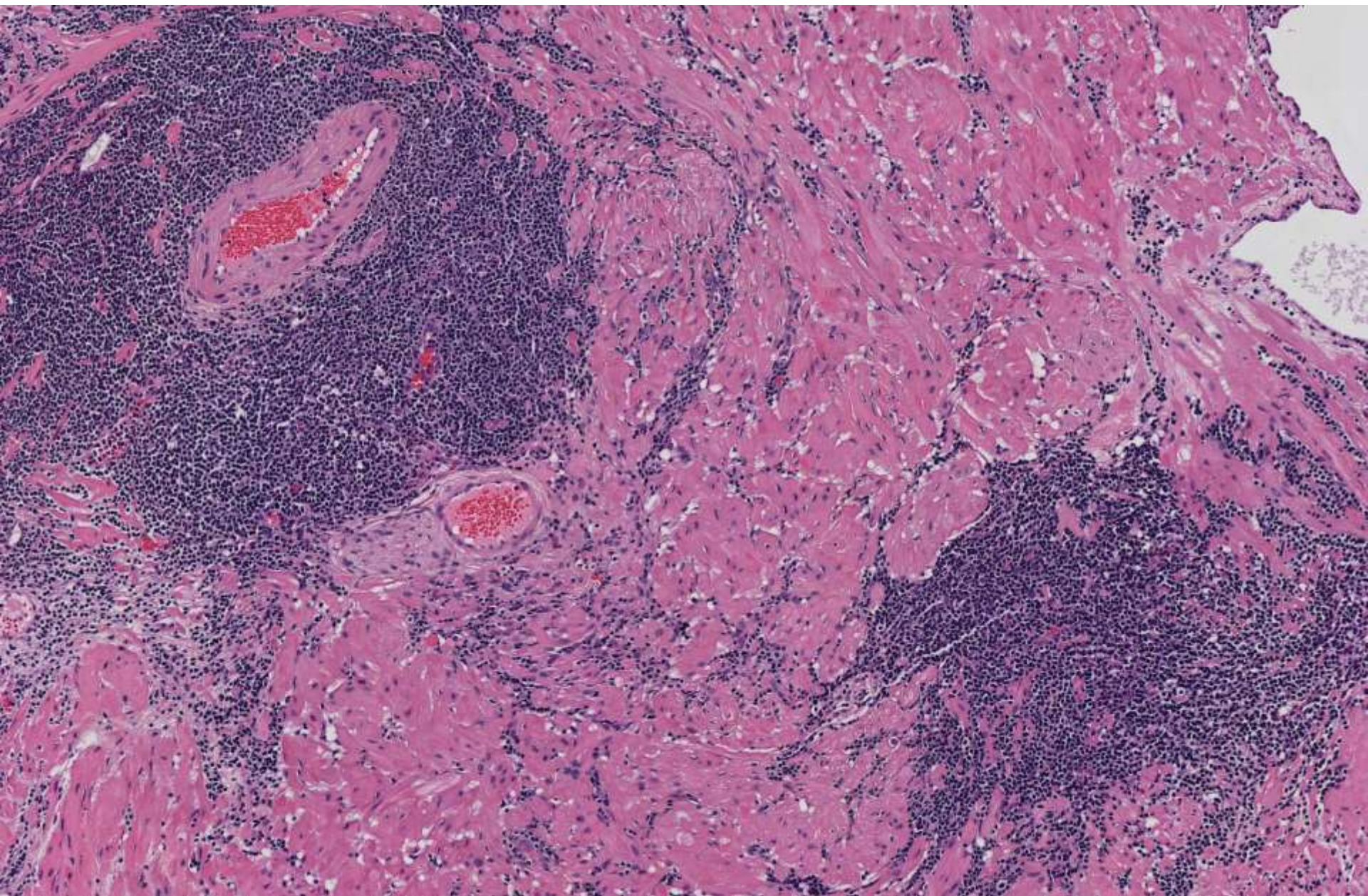




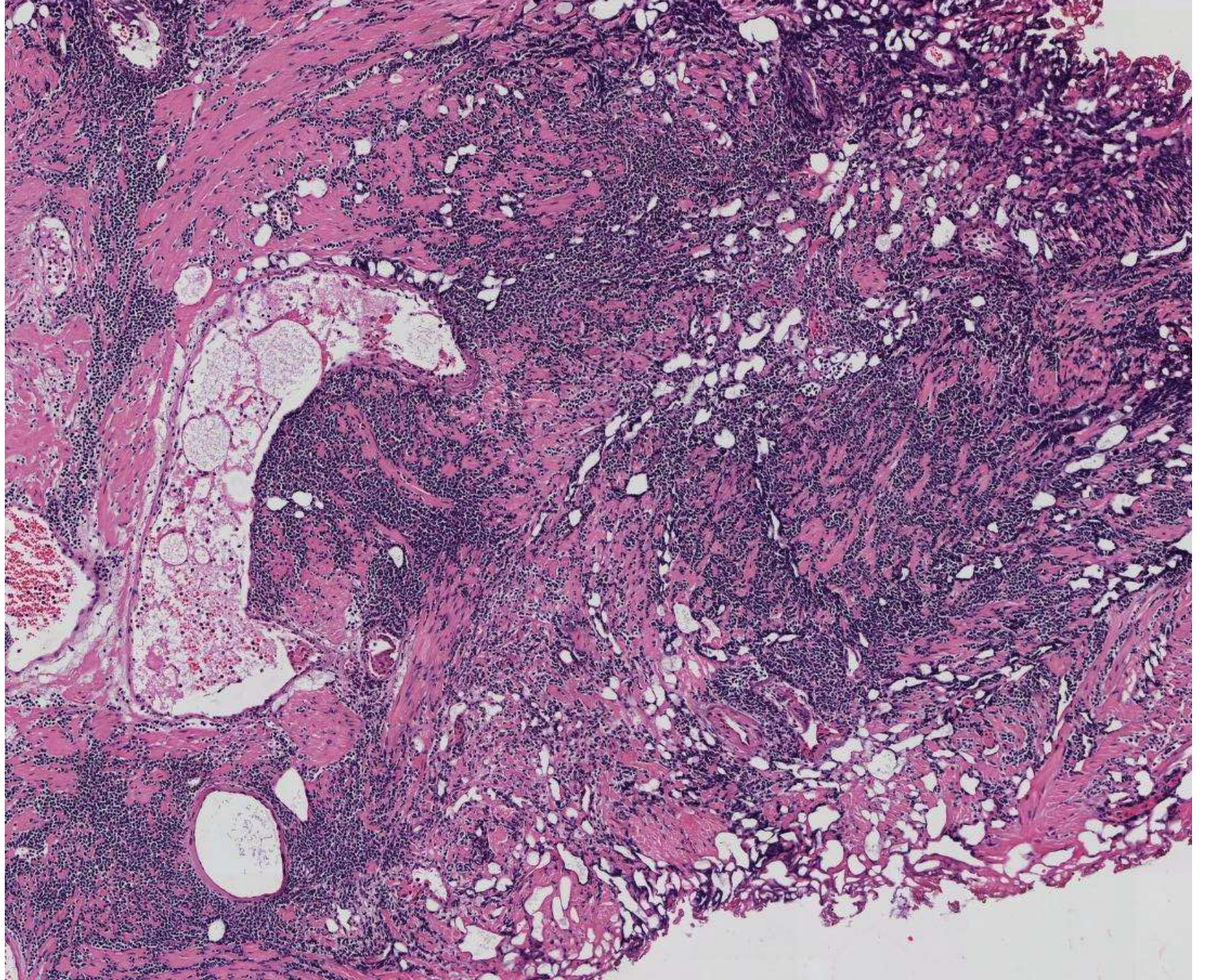




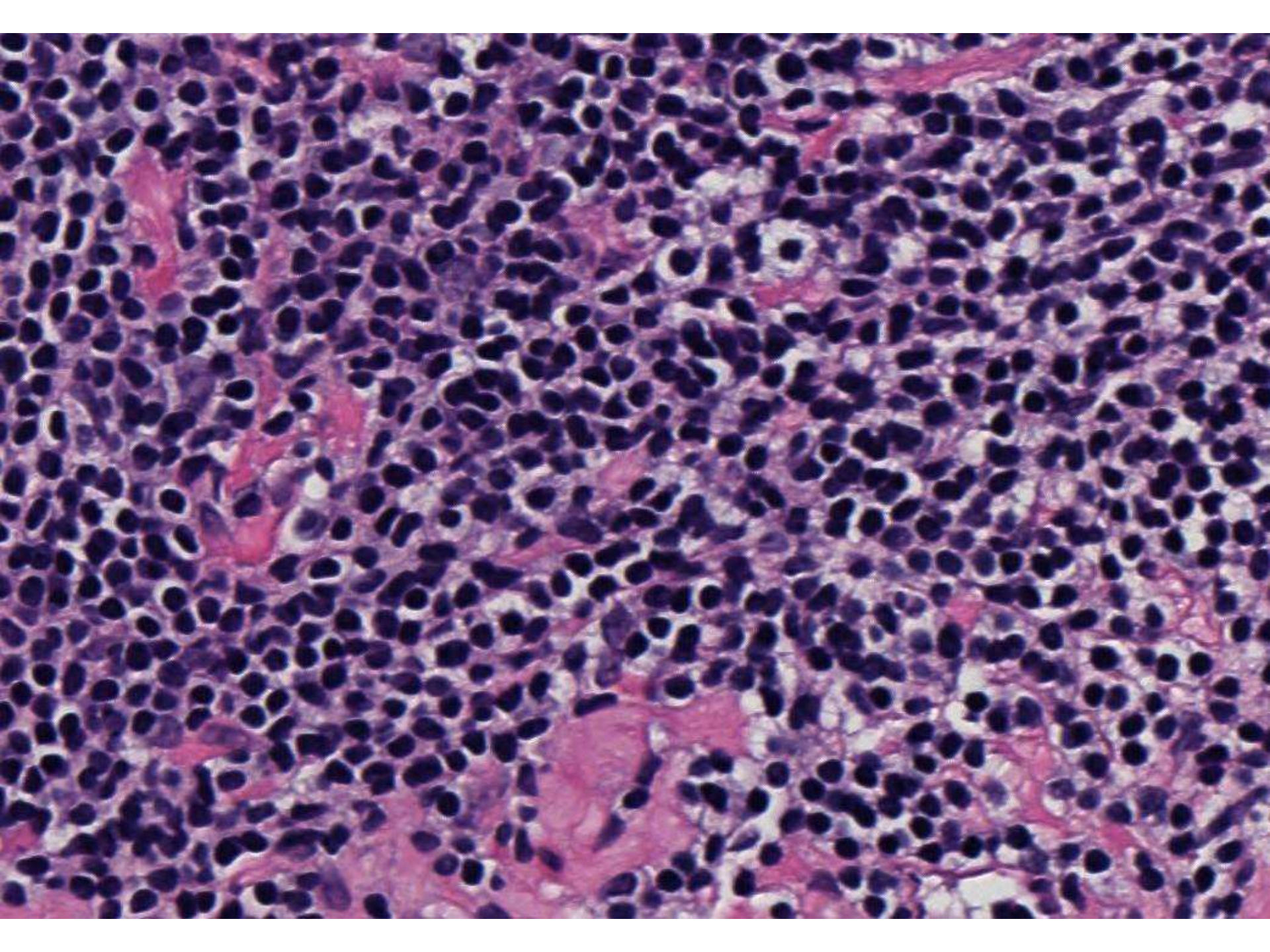




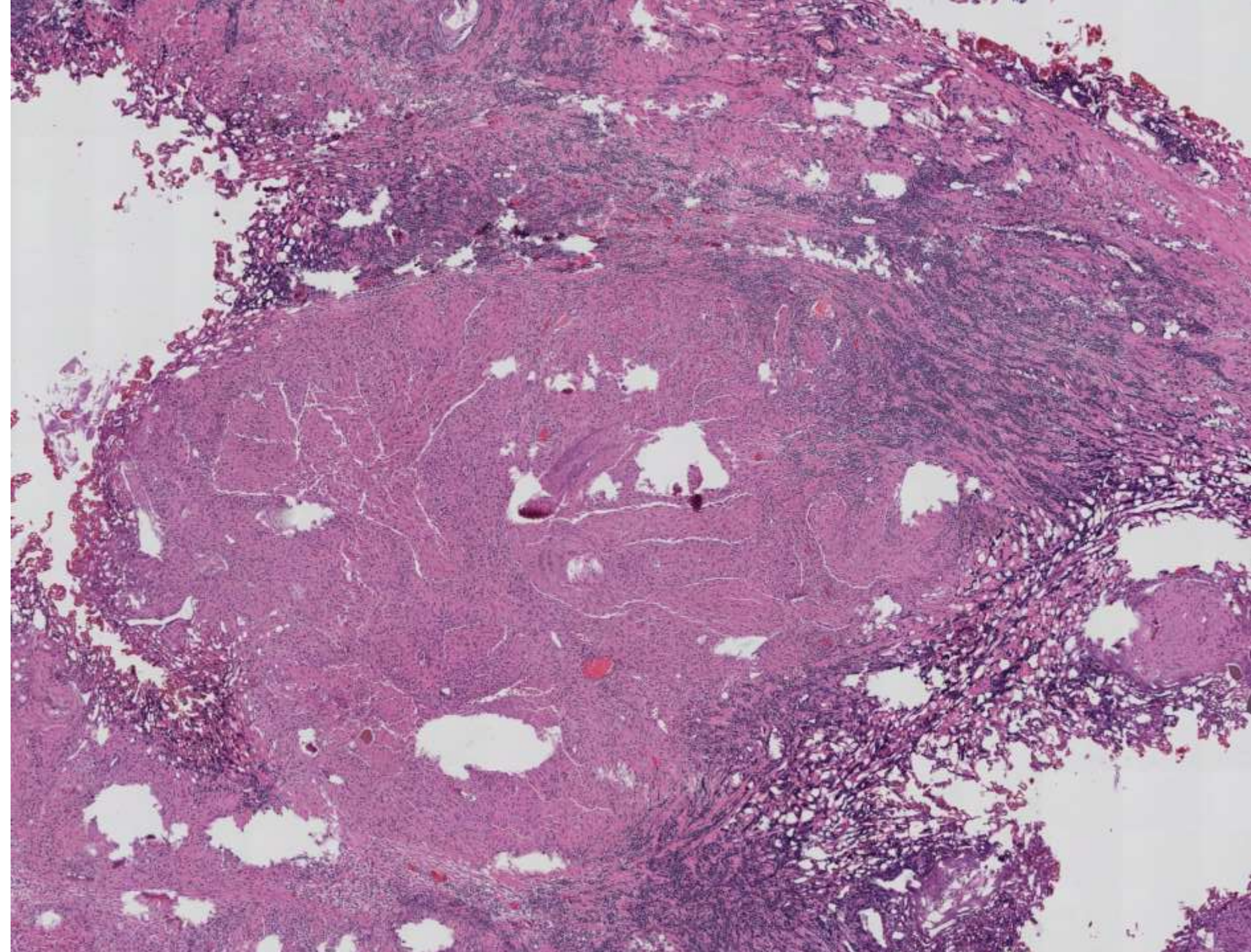




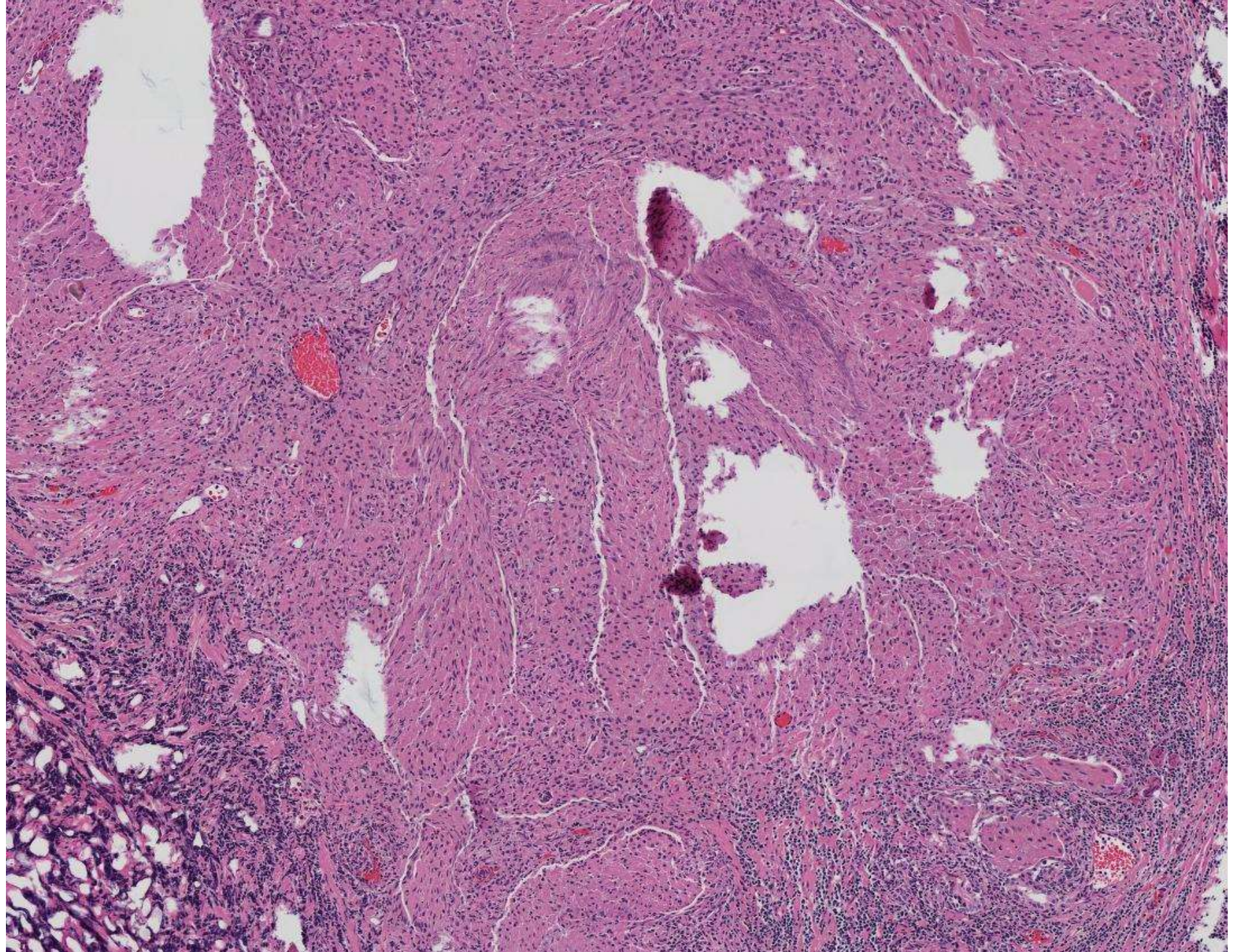




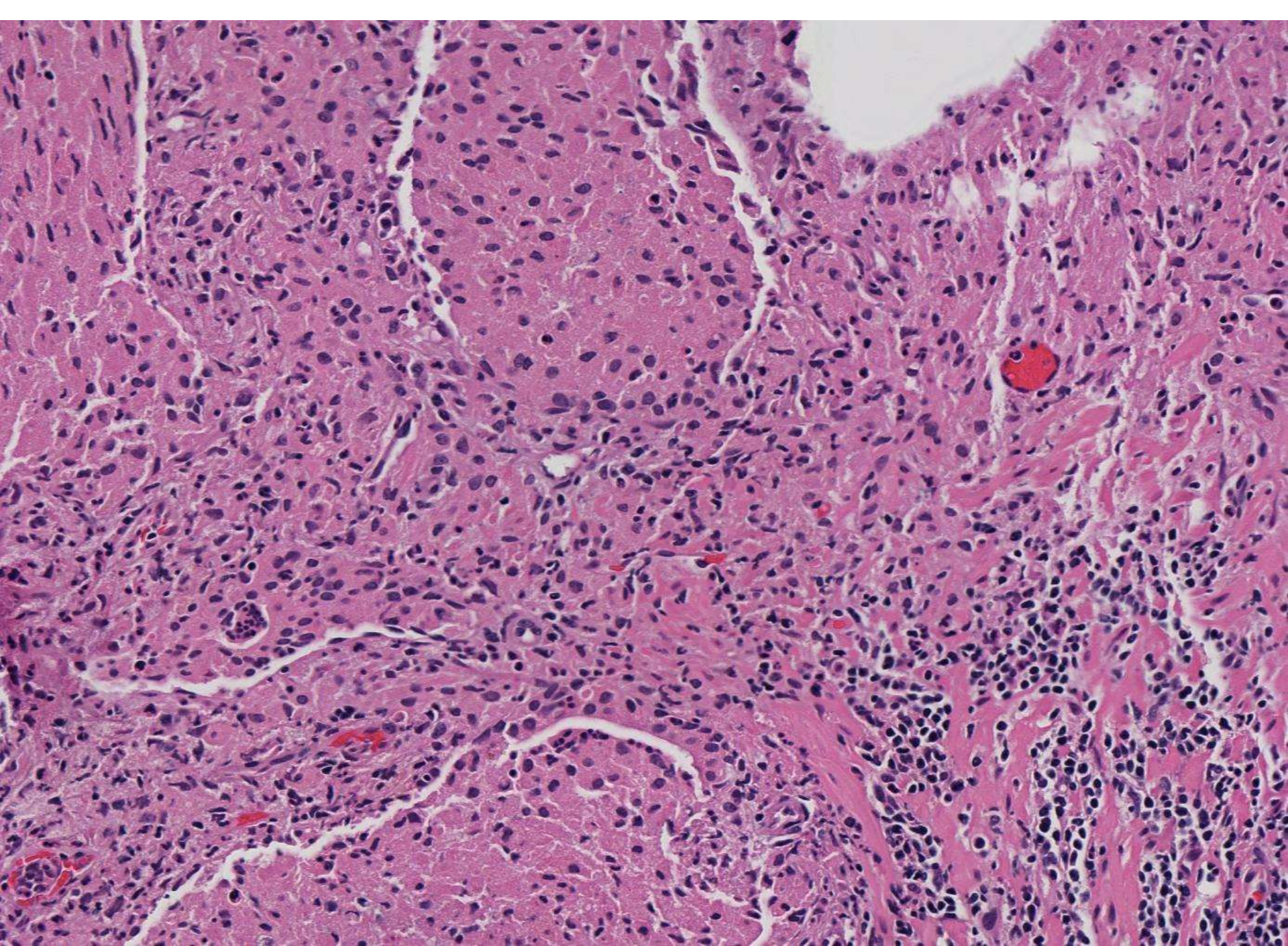




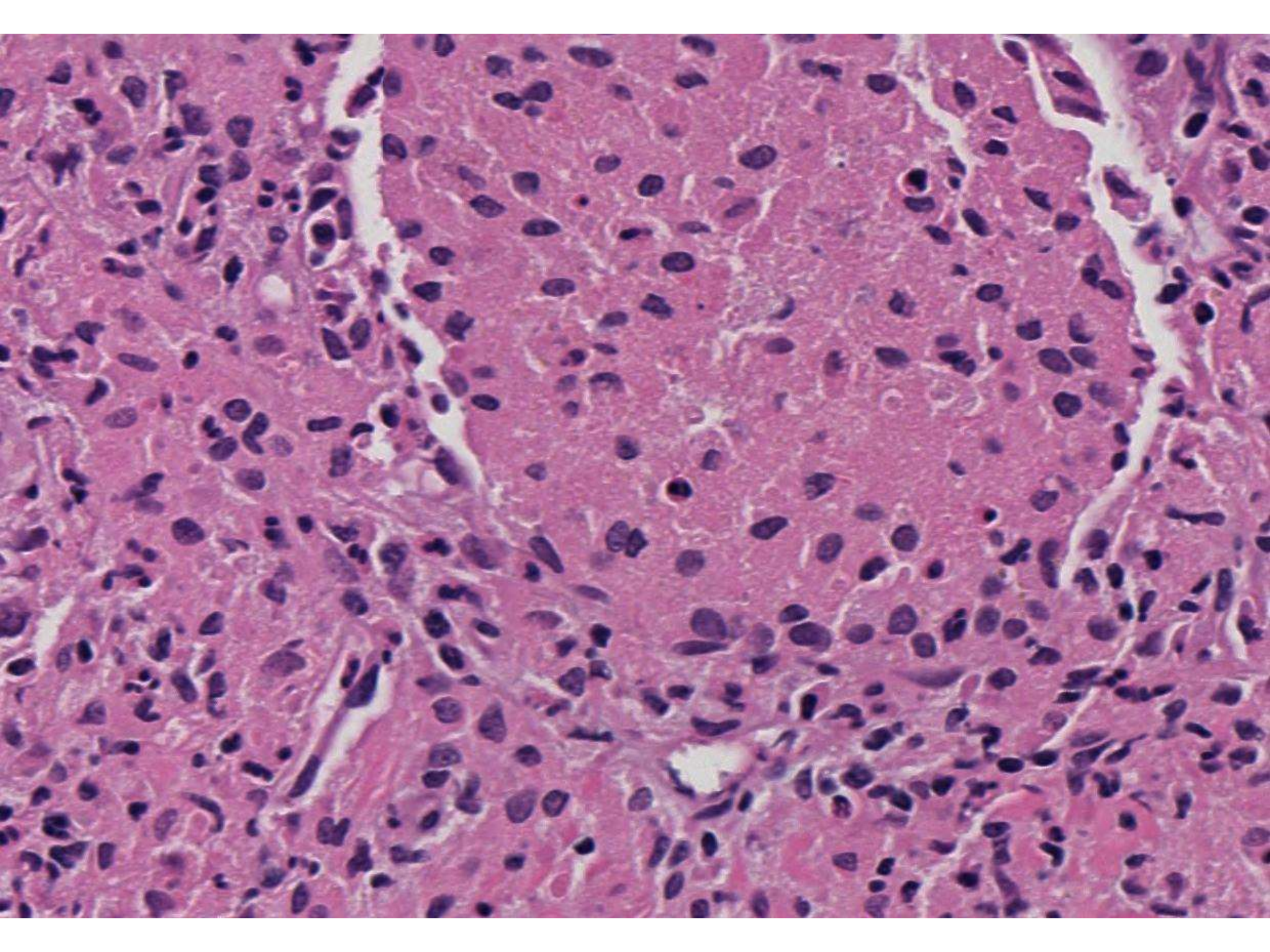




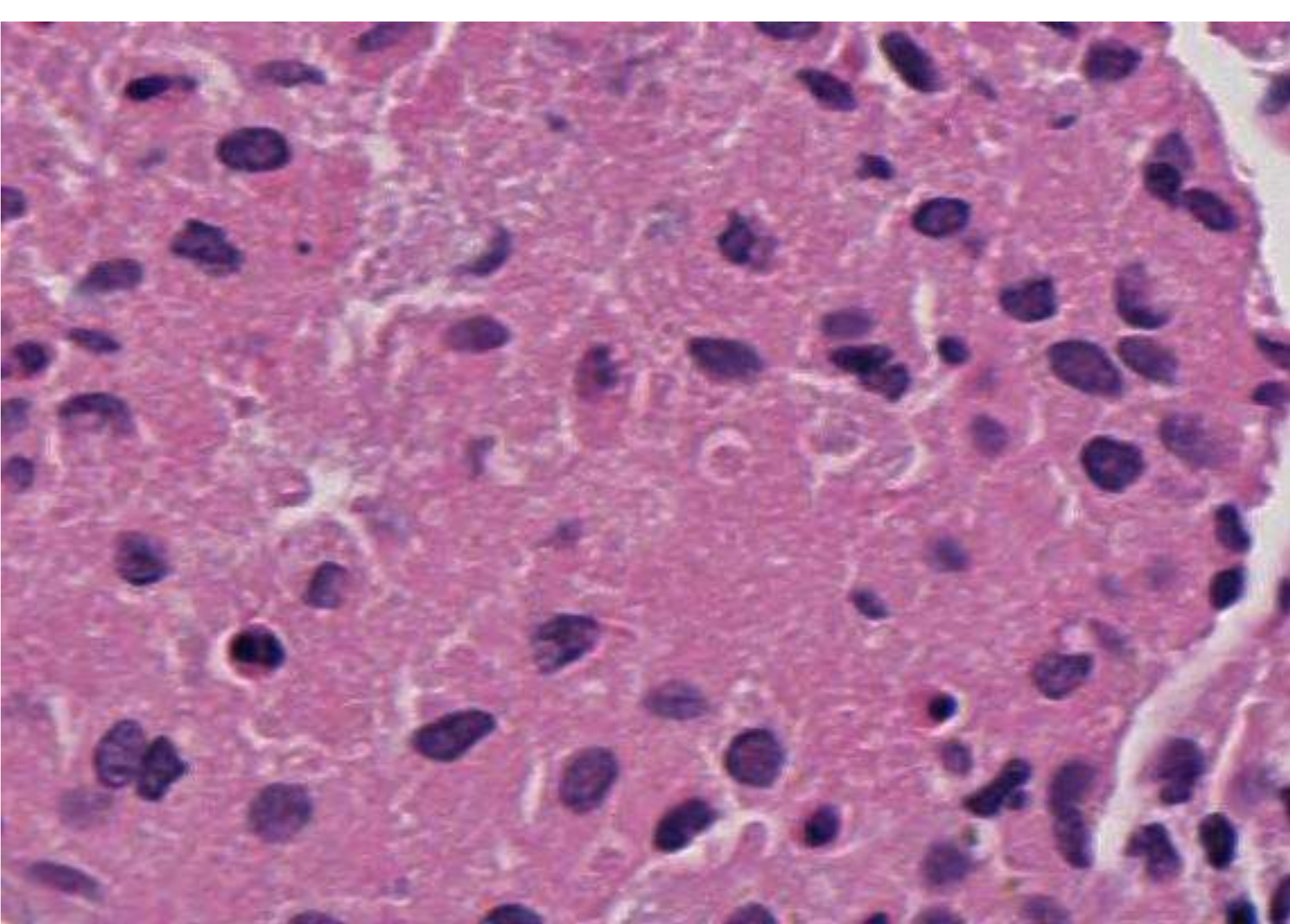






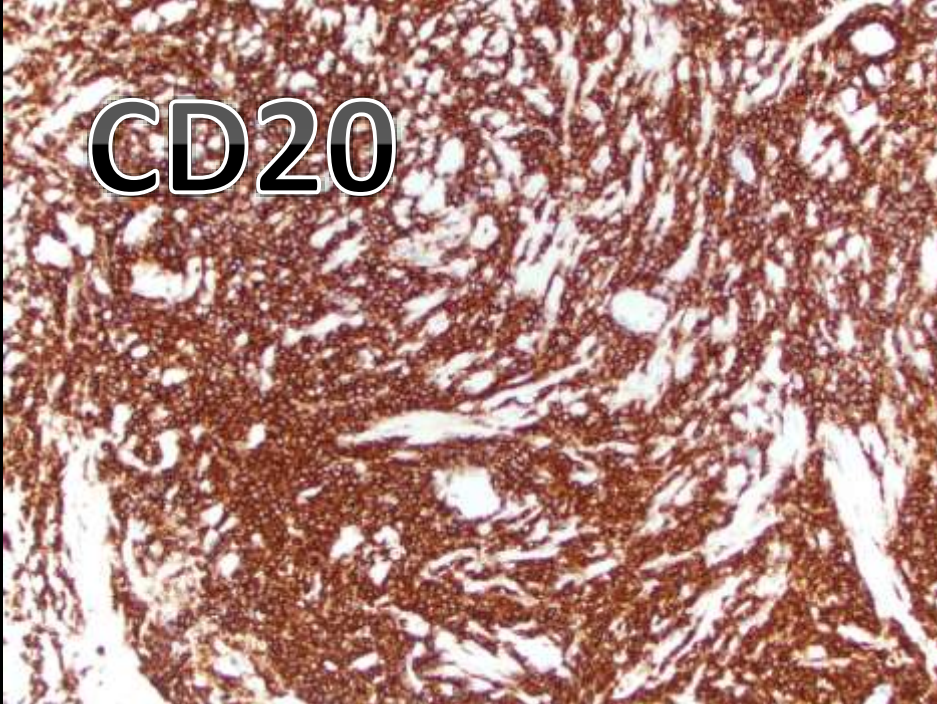




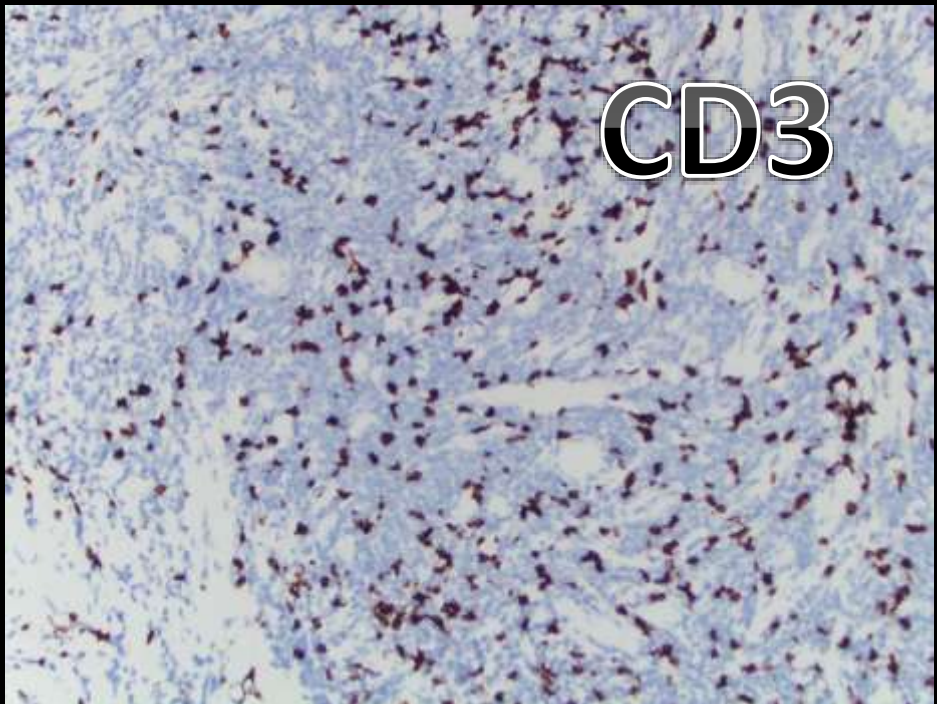




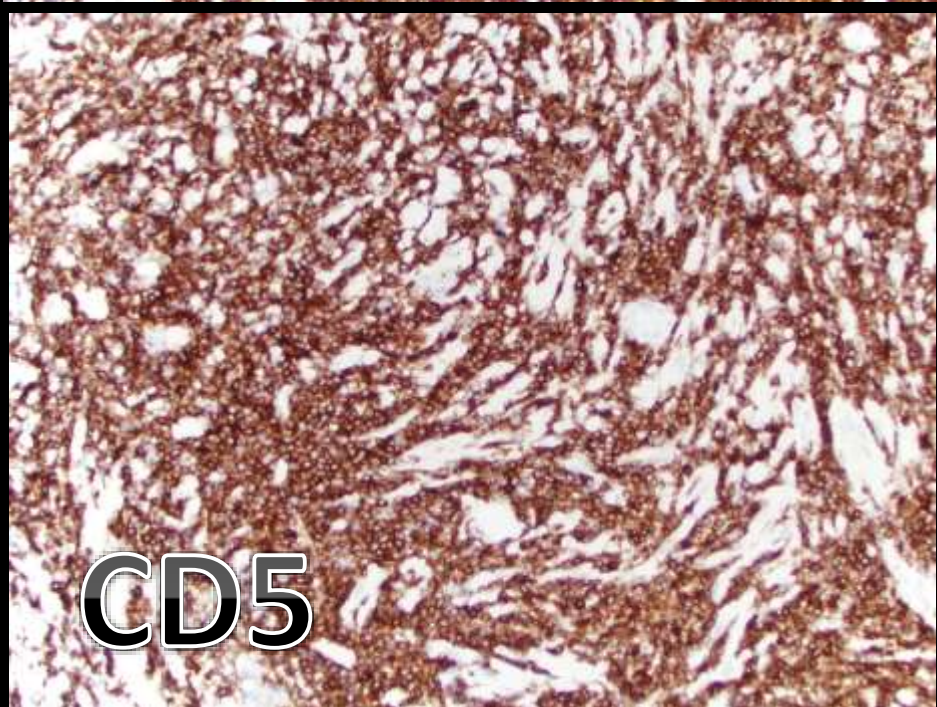
**CD20**



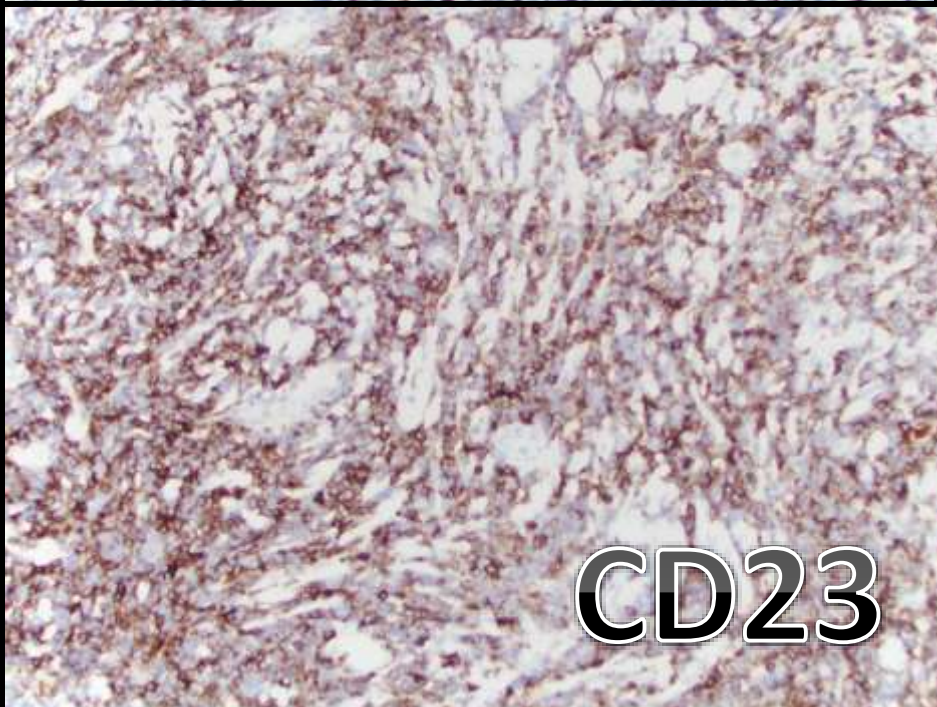
**CD3**



**CD5**

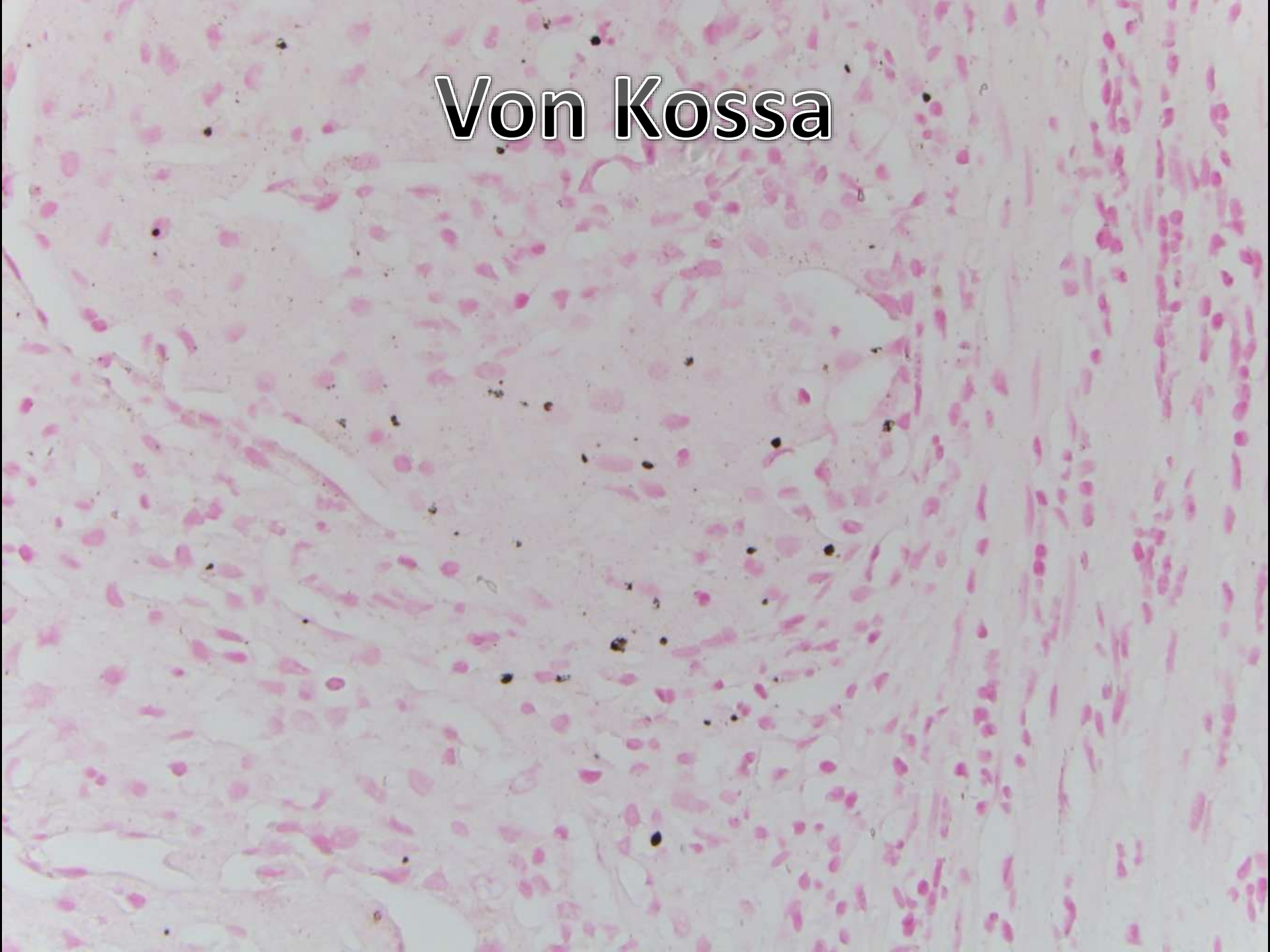


**CD23**



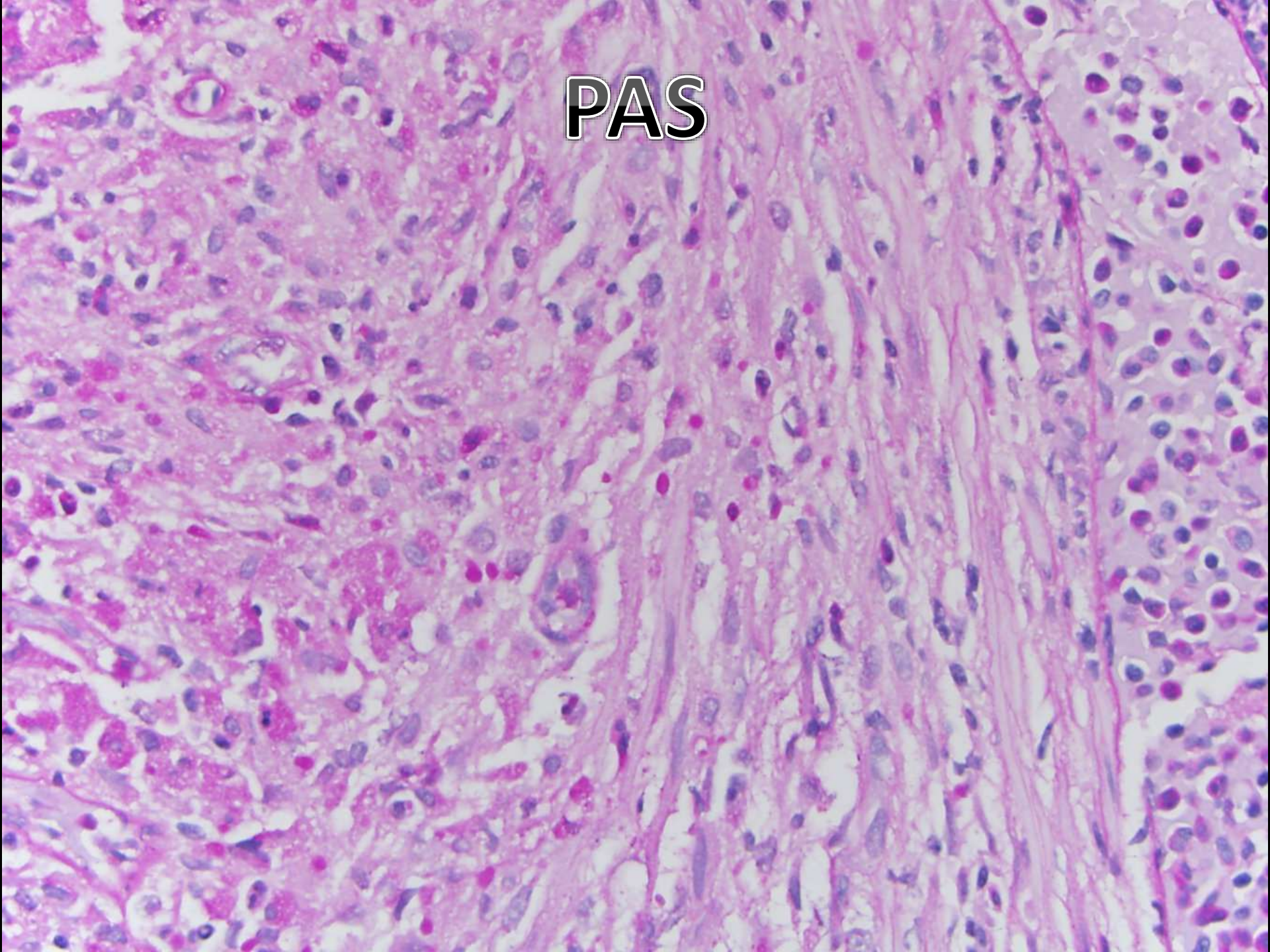


# Von Kossa





PAS



# DIAGNOSIS

- Prostate, TURP:
  - Nodular prostatic hyperplasia
  - Chronic lymphocytic leukemia/small lymphocytic lymphoma (CLL/SLL)
  - Malakoplakia



# Incidental and Concurrent Malignant Lymphomas Discovered at the Time of Prostatectomy and Prostate Biopsy *A Study of 29 Cases*

*Peiguo G. Chu, MD, PhD, Qin Huang, MD, PhD, and Lawrence M. Weiss, MD*

*Am J Surg Pathol • Volume 29, Number 5, May 2005*

- From 4381 prostate specimens
  - 29 lymphomas (<1%) in prostate specimens or pelvic LNs
    - 18 incidental cases
    - 11 with concurrent known lymphoma
    - Subtype: mostly CLL/SLL; less often marginal zone lymphoma, mantle cell lymphoma, follicular lymphoma, DLBCL

# Malakoplakia

- Rare histiocytic disease that occurs in all organs
  - Common in GU tract, particularly bladder
- Caused by defects in phagocytic or degradative functions of histiocytes in response to gram negative coliforms (*E. coli* or *Proteus*) that results in chronic inflammatory state, followed by intracellular deposition of iron and calcium (known as Michaelis-Gutmann bodies)
- Often misdiagnosed clinically as a malignant condition



# SB 6140

## (scanned slide available)

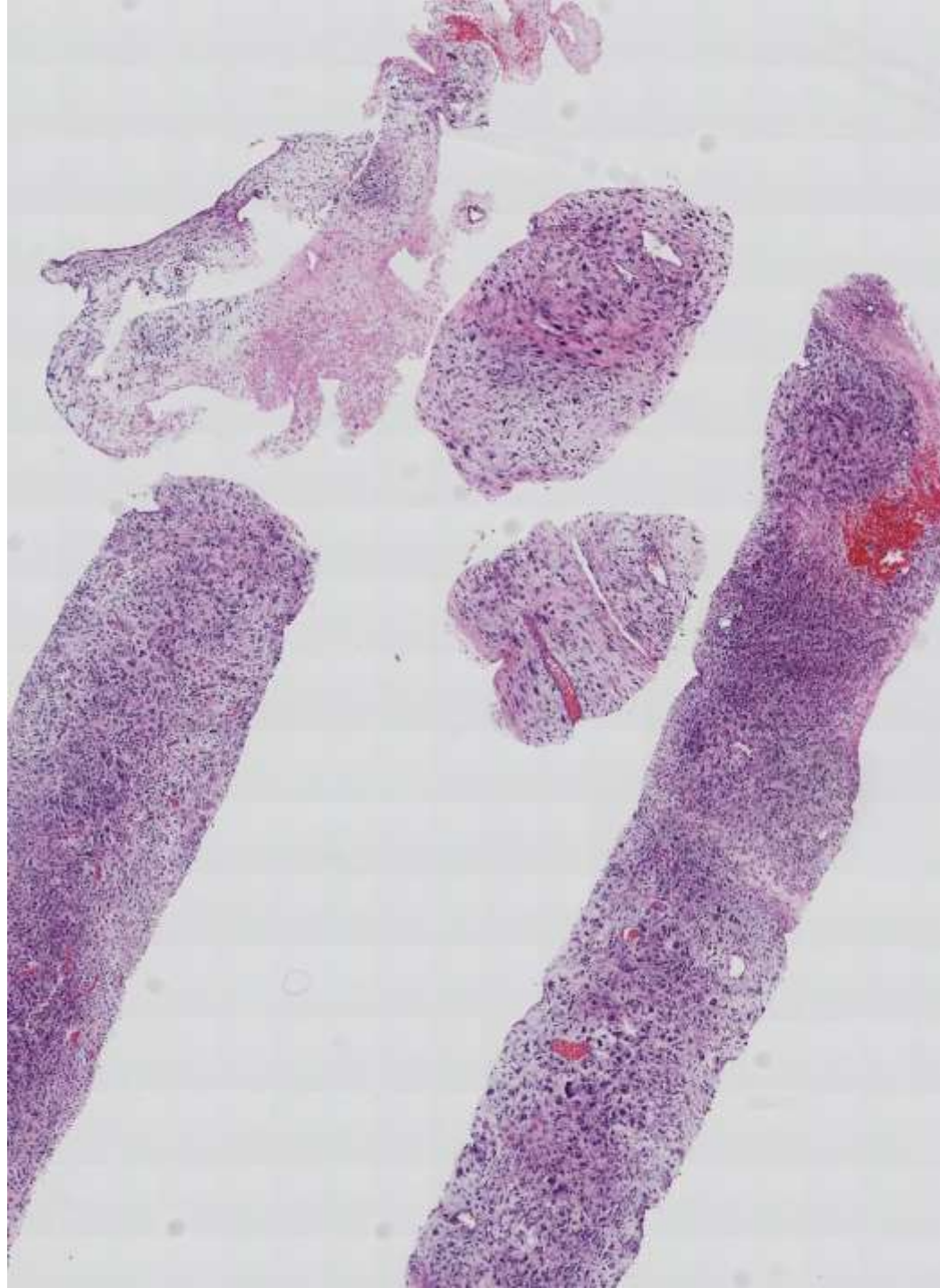
**Ankur Sangoi; El Camino Hospital**

37-year-old male with reported history of stage II mixed nonseminomatous germ cell tumor (80% embryonal carcinoma/15% teratoma/5% yolk sac tumor) status post radical orchiectomy and chemotherapy with residual teratomatous elements on retroperitoneal lymph node dissection in 2013.

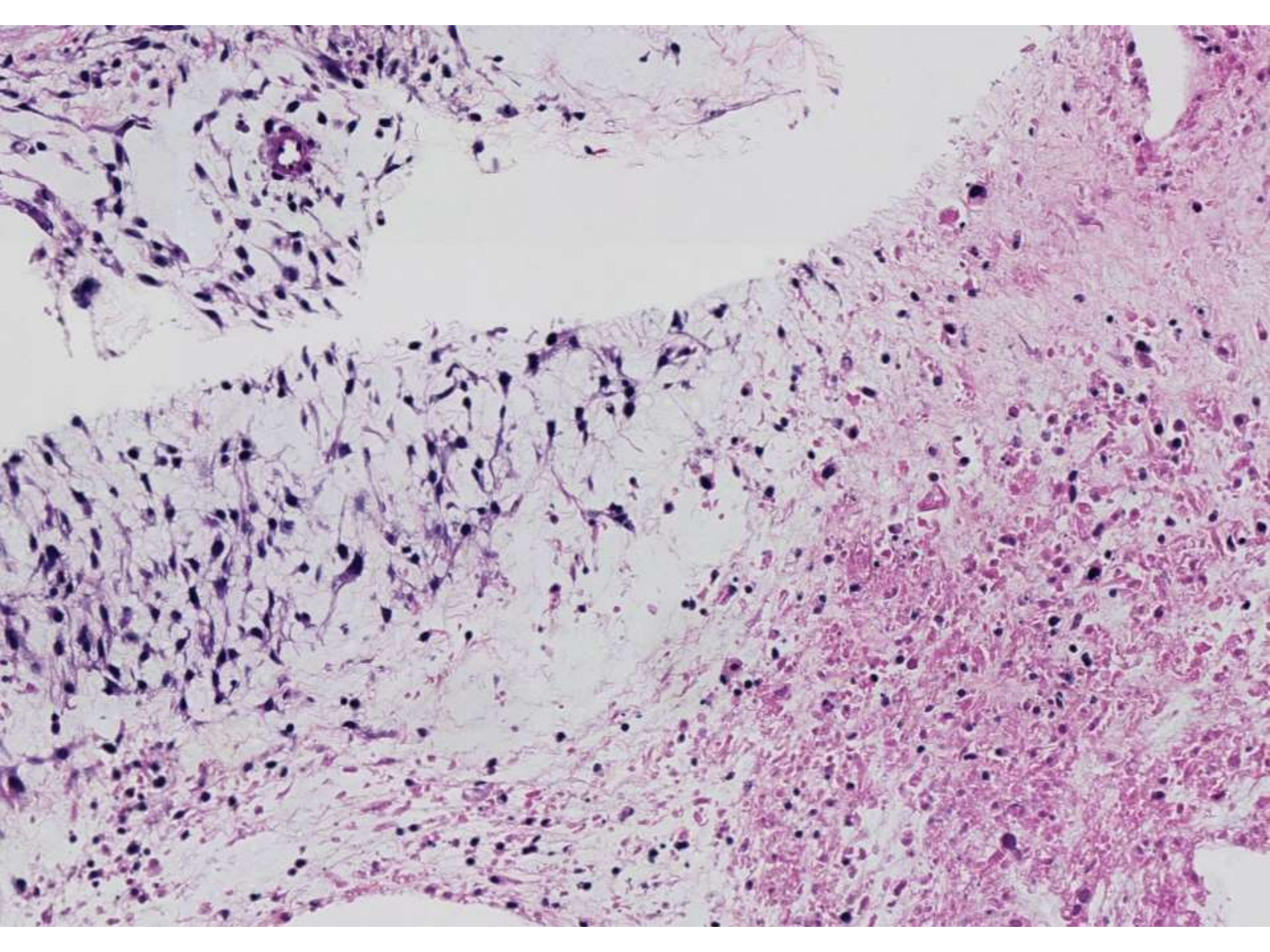
He now presents with an 11.5 cm abdominal mass and bulky paratracheal lymphadenopathy with elevated serum AFP, beta-HCG, and LDH.



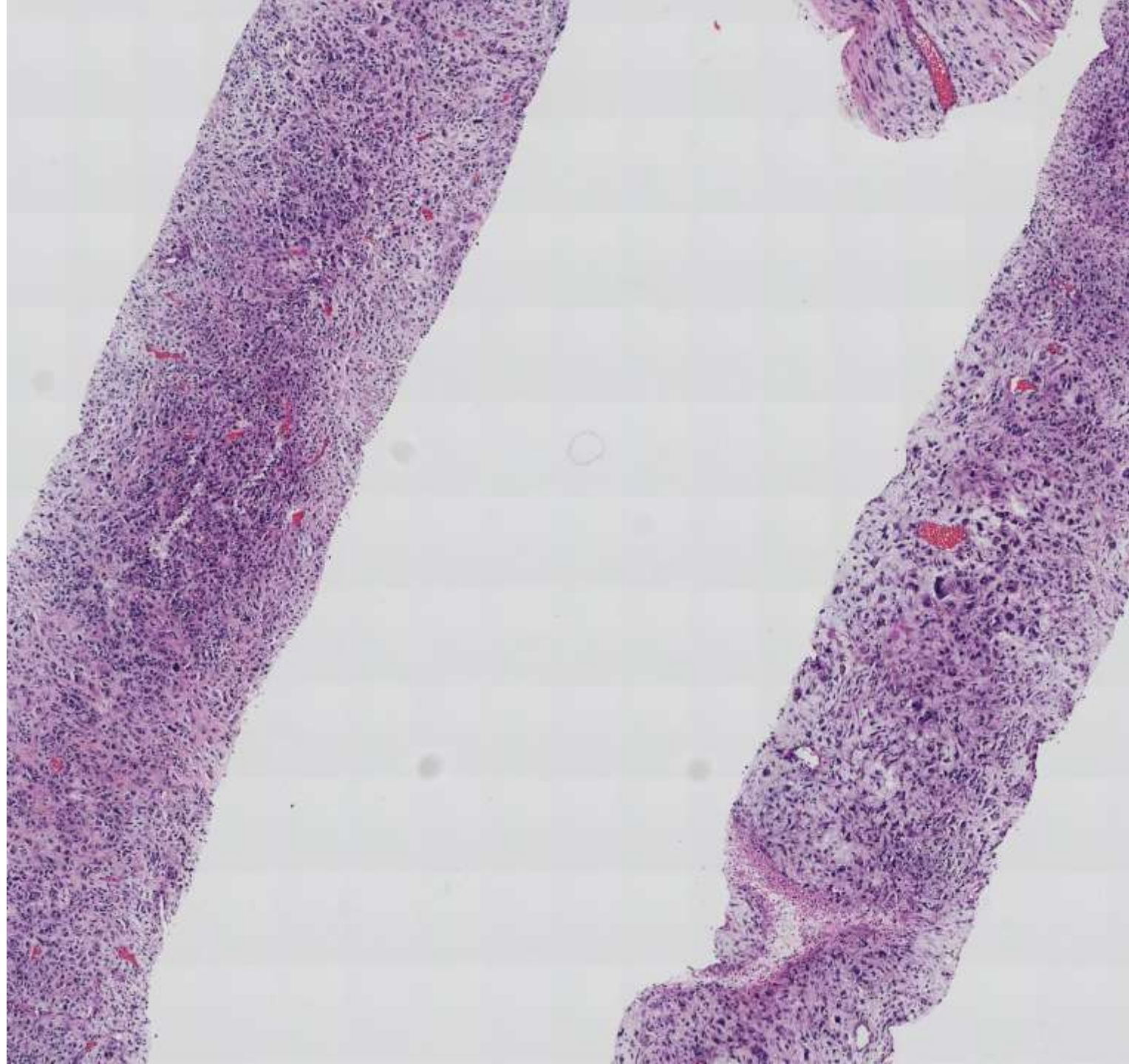




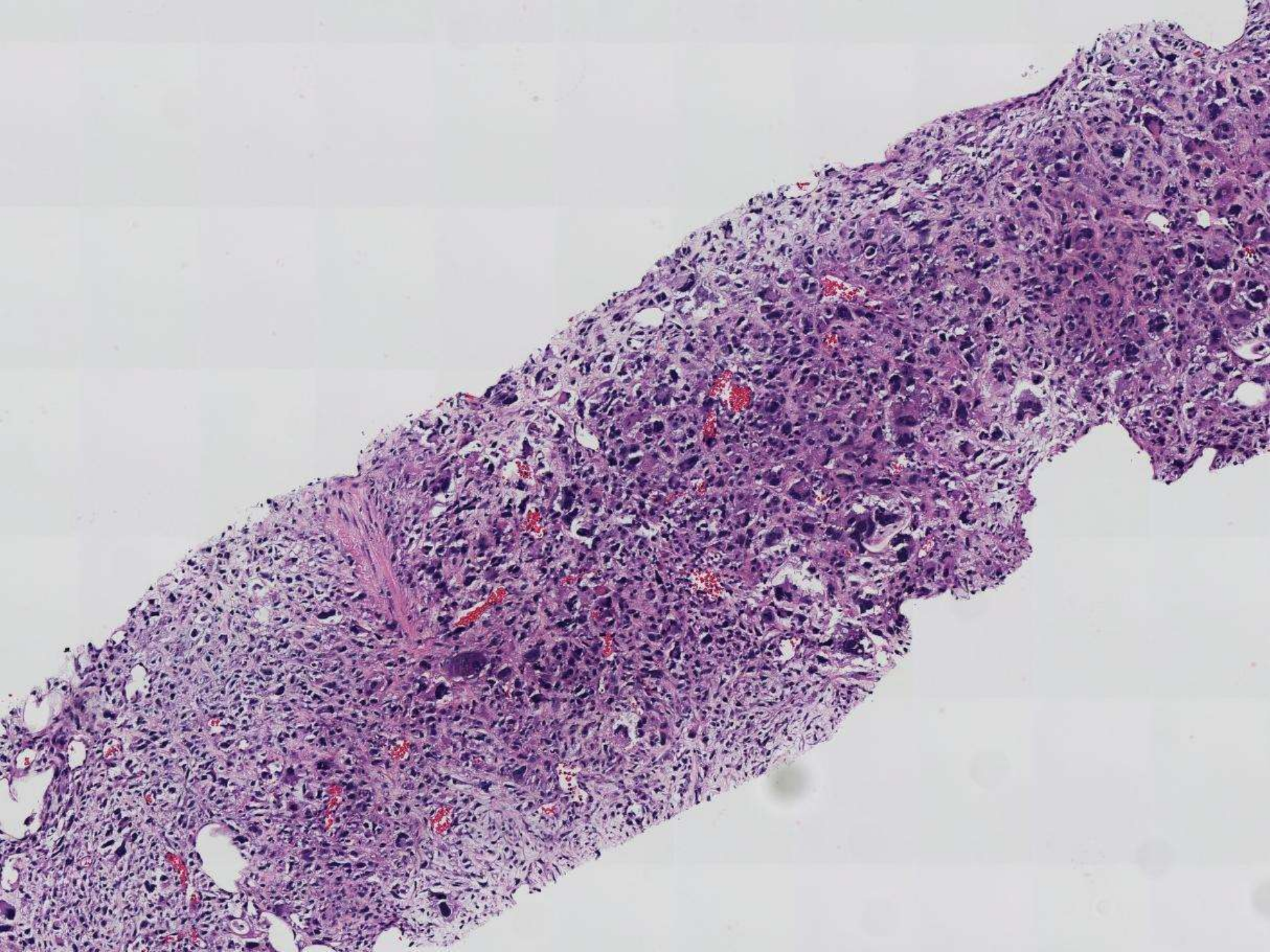




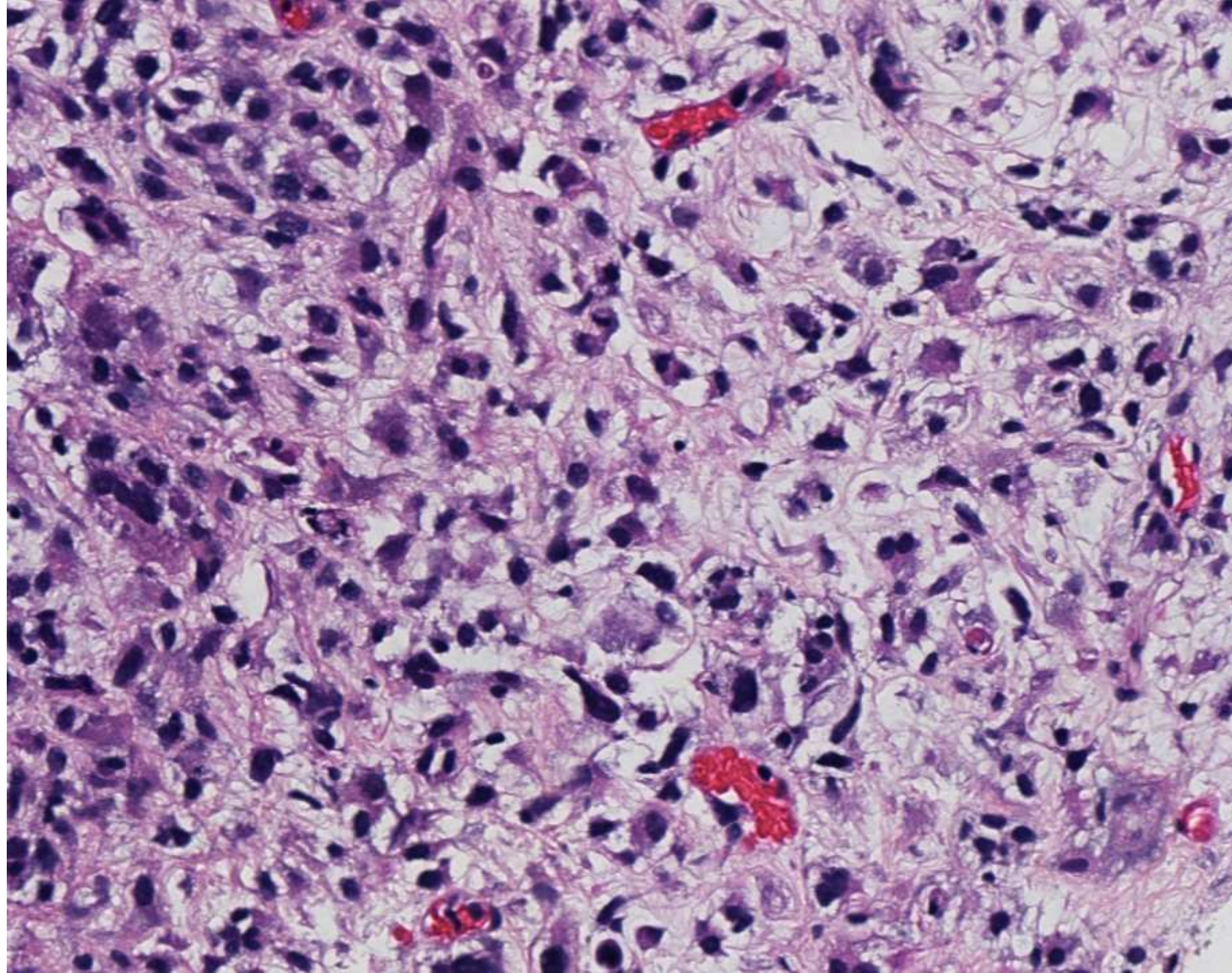




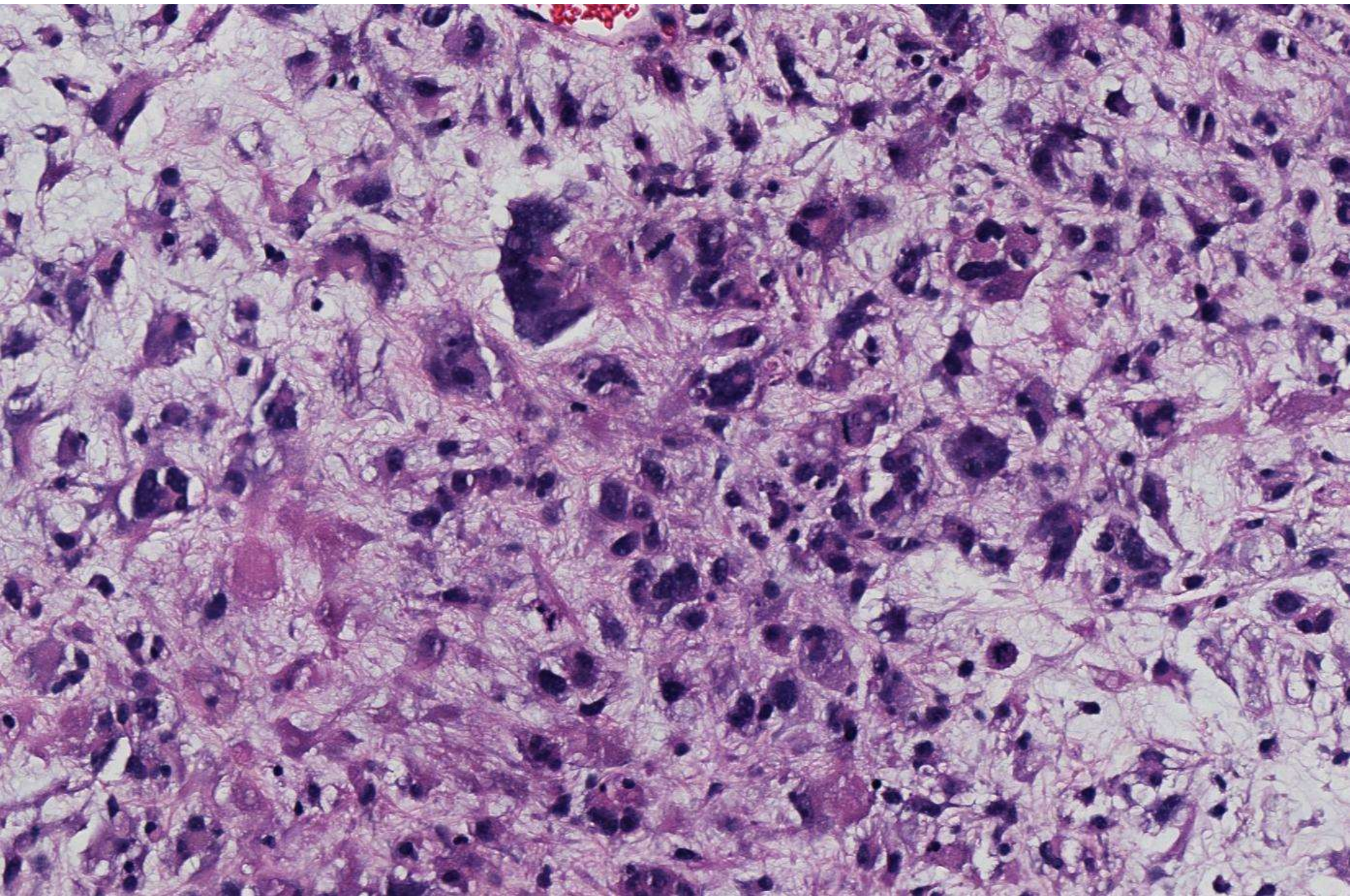




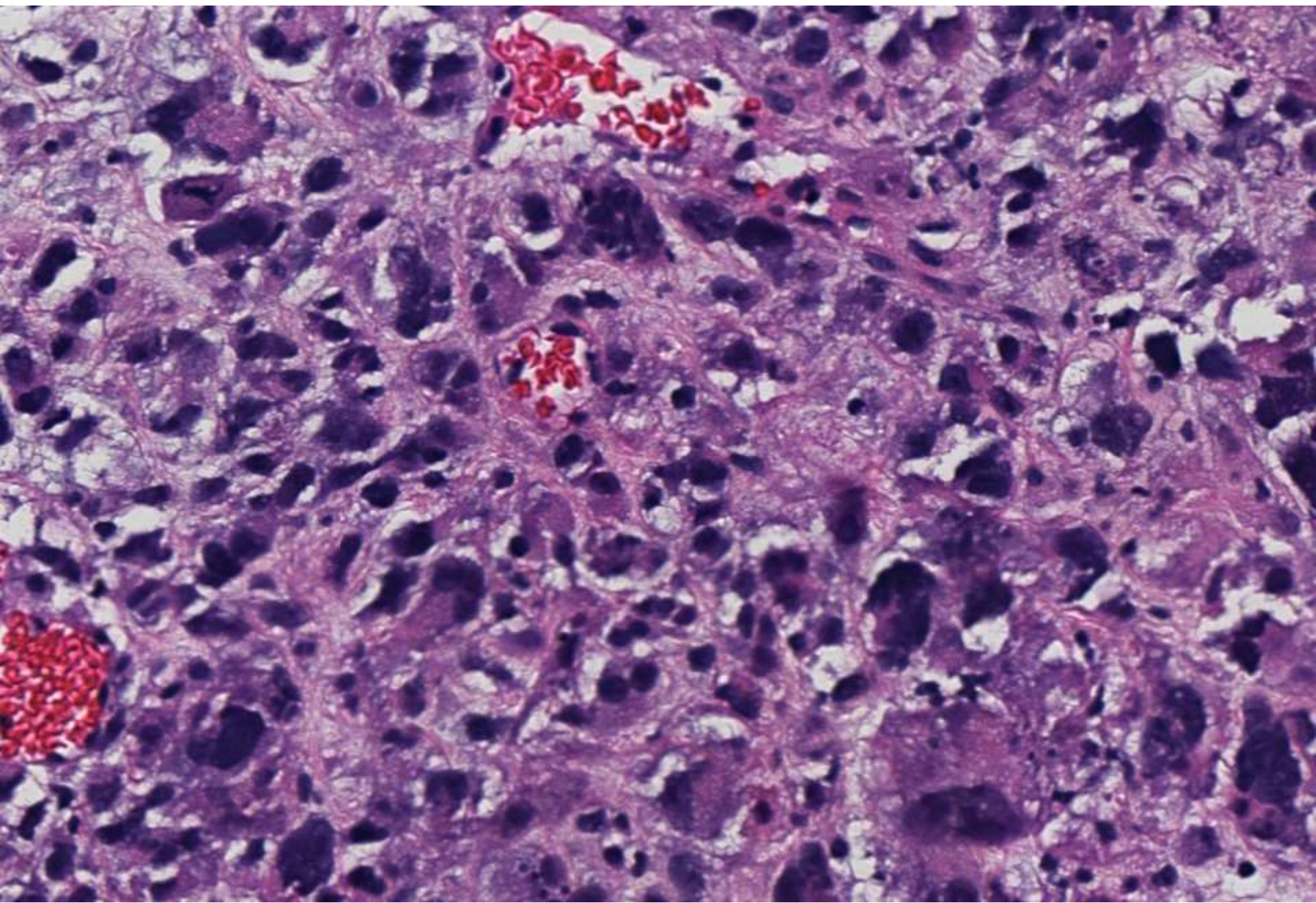
















A histological section of tissue stained with AE1/AE3 (H&E stain). The tissue shows a dense population of cells with prominent nuclei stained blue (hematoxylin) and cytoplasm/extracellular matrix stained pink (eosin). The overall architecture is disorganized, with many cells appearing atypical and crowded, suggesting a neoplastic process. The text "AE1/AE3" is overlaid in red.

**AE1/AE3**



A histological section of tissue, likely liver, stained for glypican 3. The tissue shows a dense population of cells with brown staining, indicating the presence of glypican 3. The staining is localized to the cytoplasm and cell membranes of the cells. The text "glypican3" is overlaid in red with a white outline.

glypican3

# IHC SUMMARY

- AE1/AE3: positive
- Glypican3: positive
- SALL4: positive
  
- OCT3/4: negative
- CD30: negative
- Desmin: negative
- SOX11: negative
- CD43: negative



# DIAGNOSIS

- Yolk sac tumor
  - Predominantly sarcomatoid variant
  - Focal microcystic variant
- No evidence of somatic malignancy (no transformation to frank sarcoma)

# Malignant “transformation” of testicular GCTs

- Uncommon overall
- Usually sarcomatoid transformation
  - Less often: carcinomatous transformation
  - Occurs in late recurrences
  - Subtype frequency: rhabdomyosarcoma, angiosarcoma, leiomyosarcoma, chondrosarcoma
    - Can also be sarcoma NOS subtype

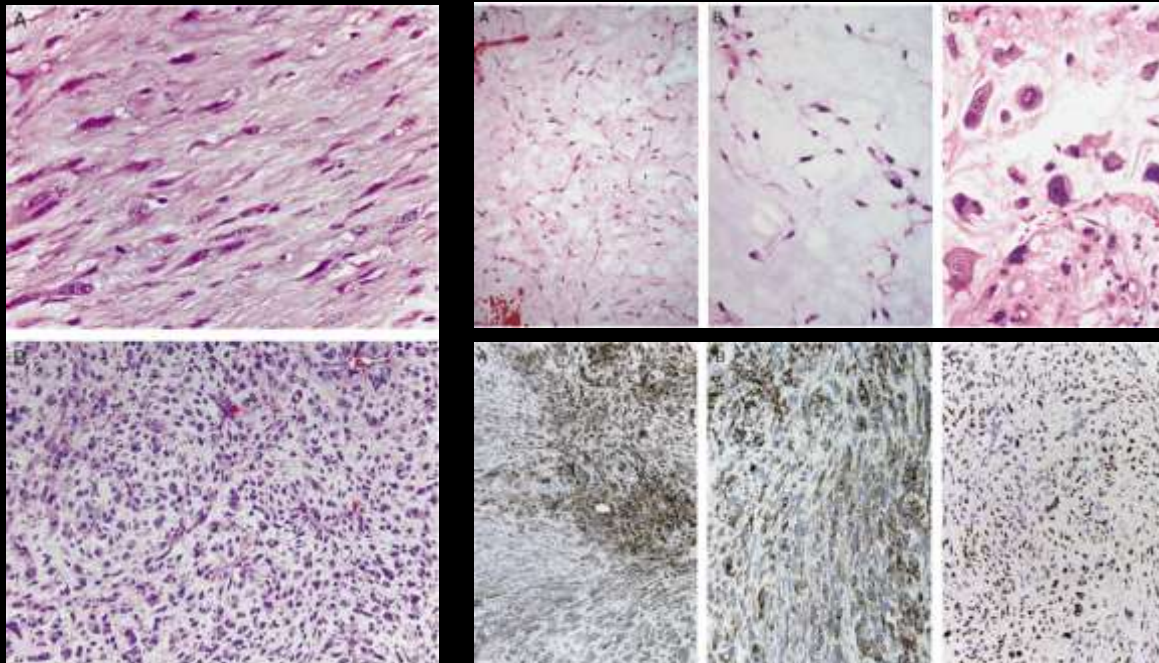


# Many Postchemotherapy Sarcomatous Tumors in Patients With Testicular Germ Cell Tumors Are Sarcomatoid Yolk Sac Tumors

## *A Study of 33 Cases*

*Brooke E. Howitt, MD,\* Martin J. Magers, MD,† Kevin R. Rice, MD,‡  
Cristina D. Cole, MD,† and Thomas M. Ulbright, MD†*

*Am J Surg Pathol • Volume 39, Number 2, February 2015*



# Postchemotherapy sarcomatous tumors in testicular GCTs

- 33 sarcomatoid tumors lacking specific sarcoma subtype (NOS)
- Yolk sac tumor criteria:
  - Moderate intensity and >10% staining with both AE1/AE3 and glypican3
- 22 of 33 (67%) tumors met this criteria
  - 50% cases desmin positive but myogenin negative
  - 68% SALL4 positive; all MUC4 negative
    - Sarcomatoid yolk sac tumors
- Typically occur several years after initial Dx with normal to mildly elevated AFP
- Behave aggressively when high grade