Disclosures
June 1, 2015

Dr. Keith Duncan has disclosed that he/his group received a consultation fee from Abbvie (Redwood City) and Oxford Biotherapeutics (San Jose) for review of immunohistochemical stains. Dr. Sonam Prakash has disclosed that she received monetary benefits from Incyte Corporation in her role as advisor for the Hematopathology Publications Steering Committee. The activity planners have determined that these financial relationships are not relevant to the cases being presented.

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

Presenters:

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Activity Planners:

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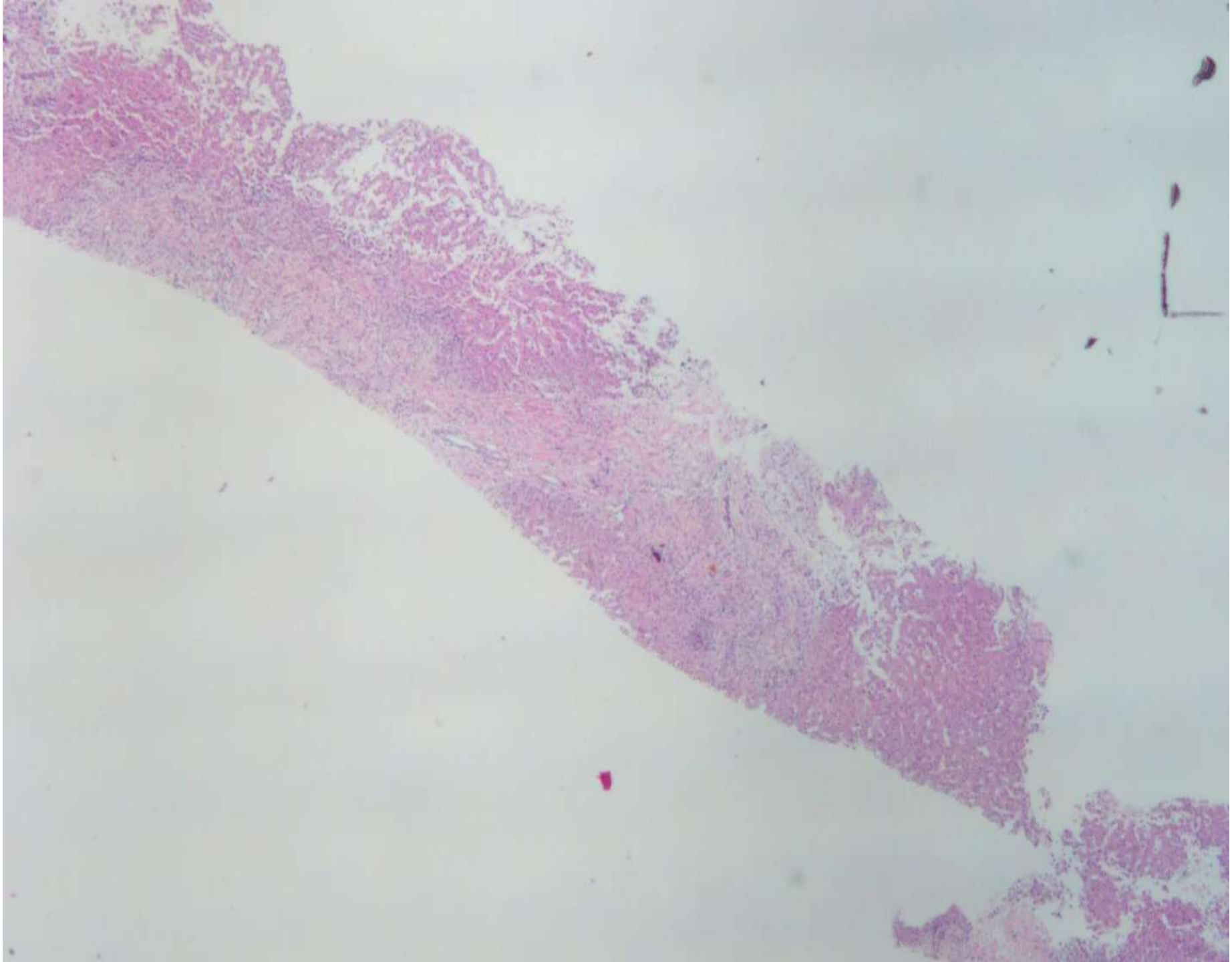
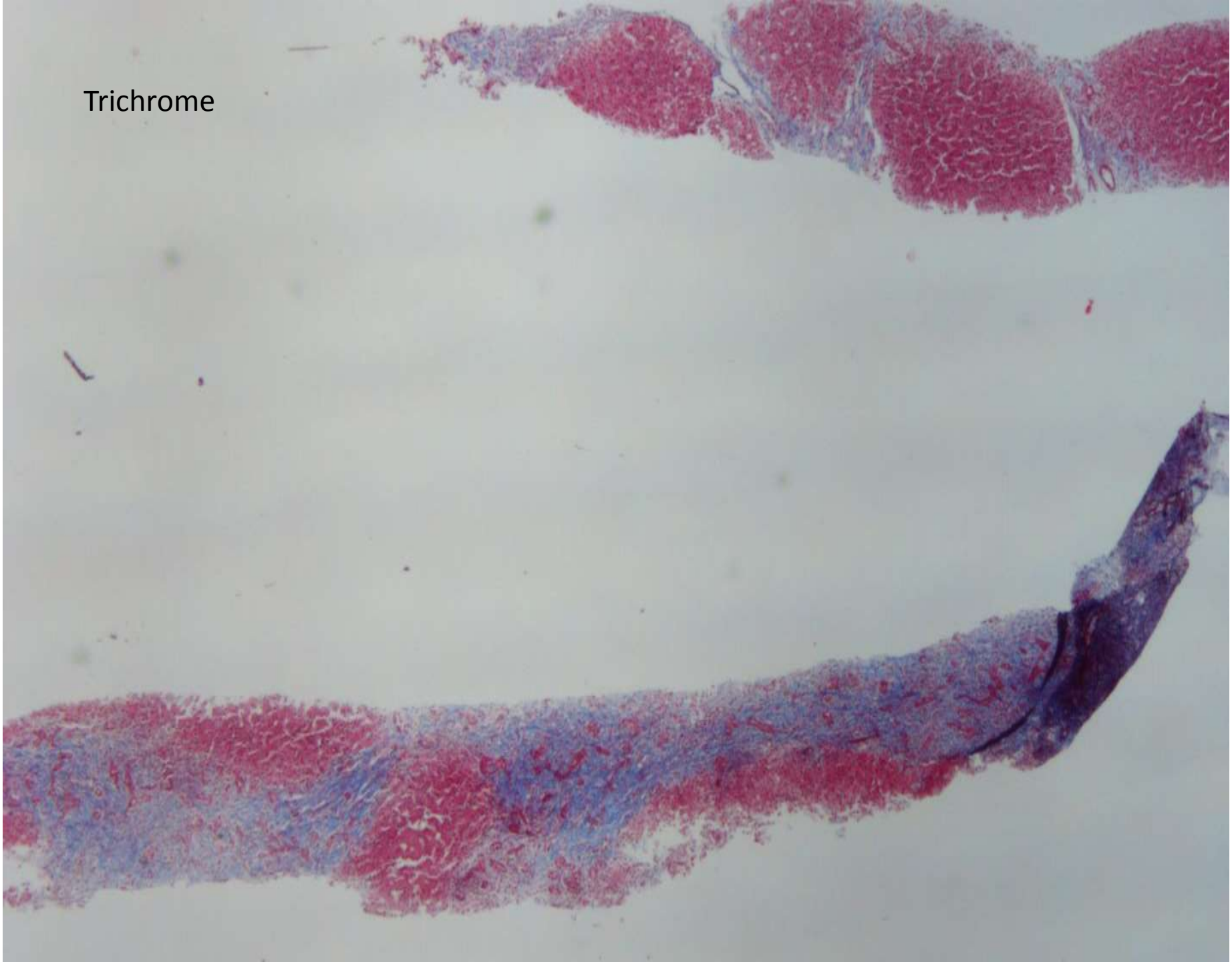
William Rogers, MD

SB 5941

- 15-year-old male incidentally discovered by imaging to have cirrhosis. Liver biopsy performed to determine etiology.

- **Walden Browne; Kaiser Antioch**

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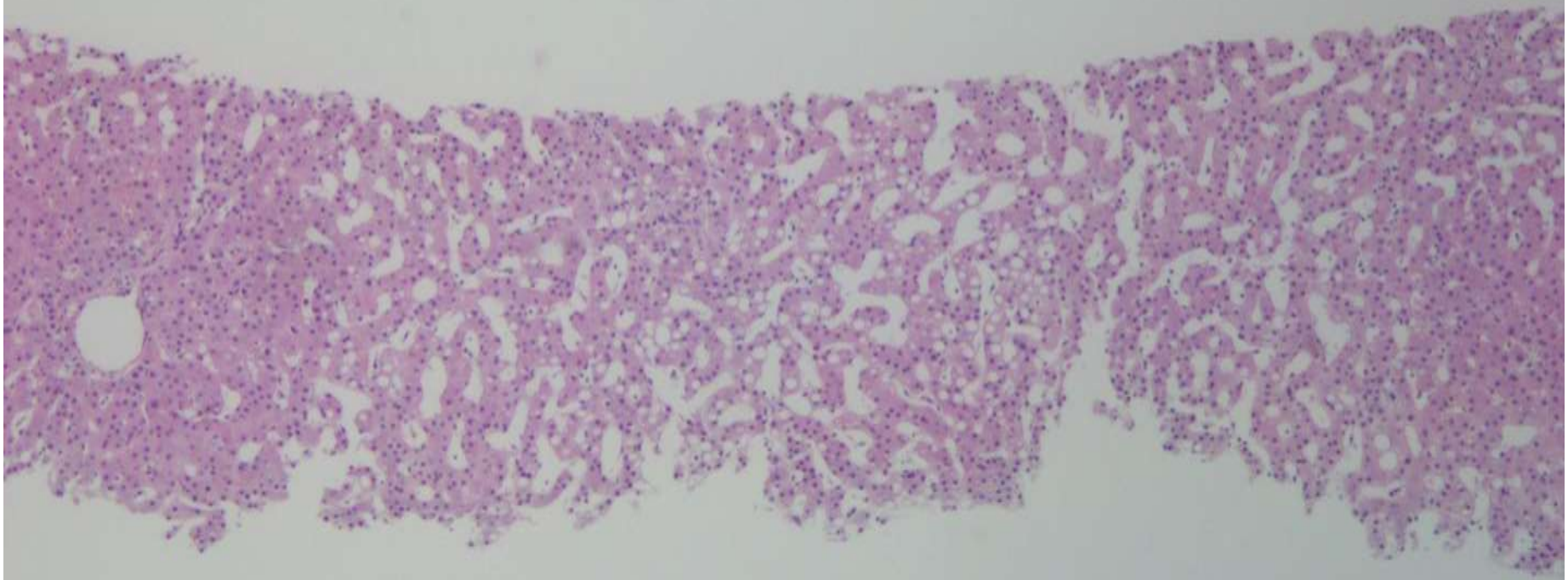

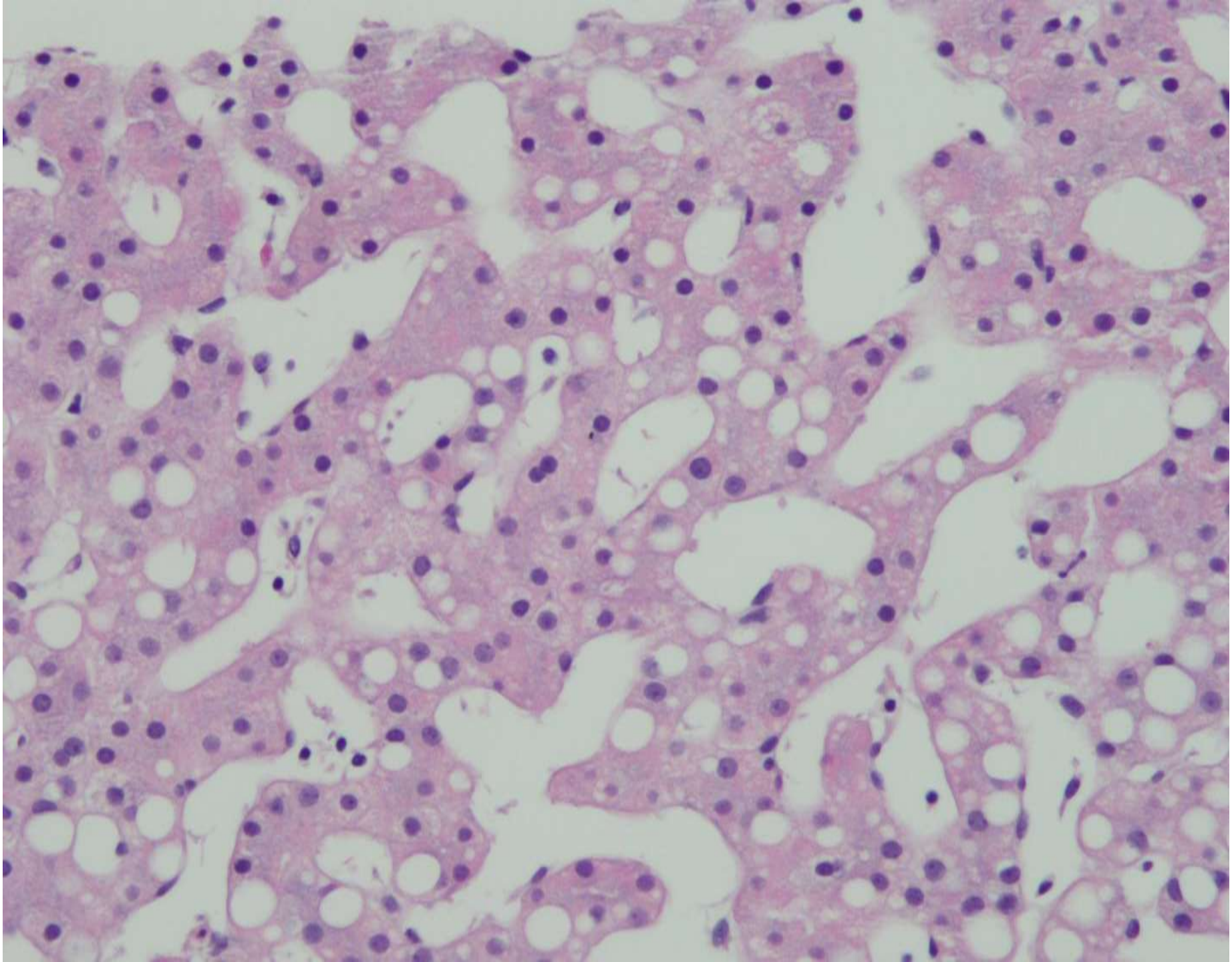

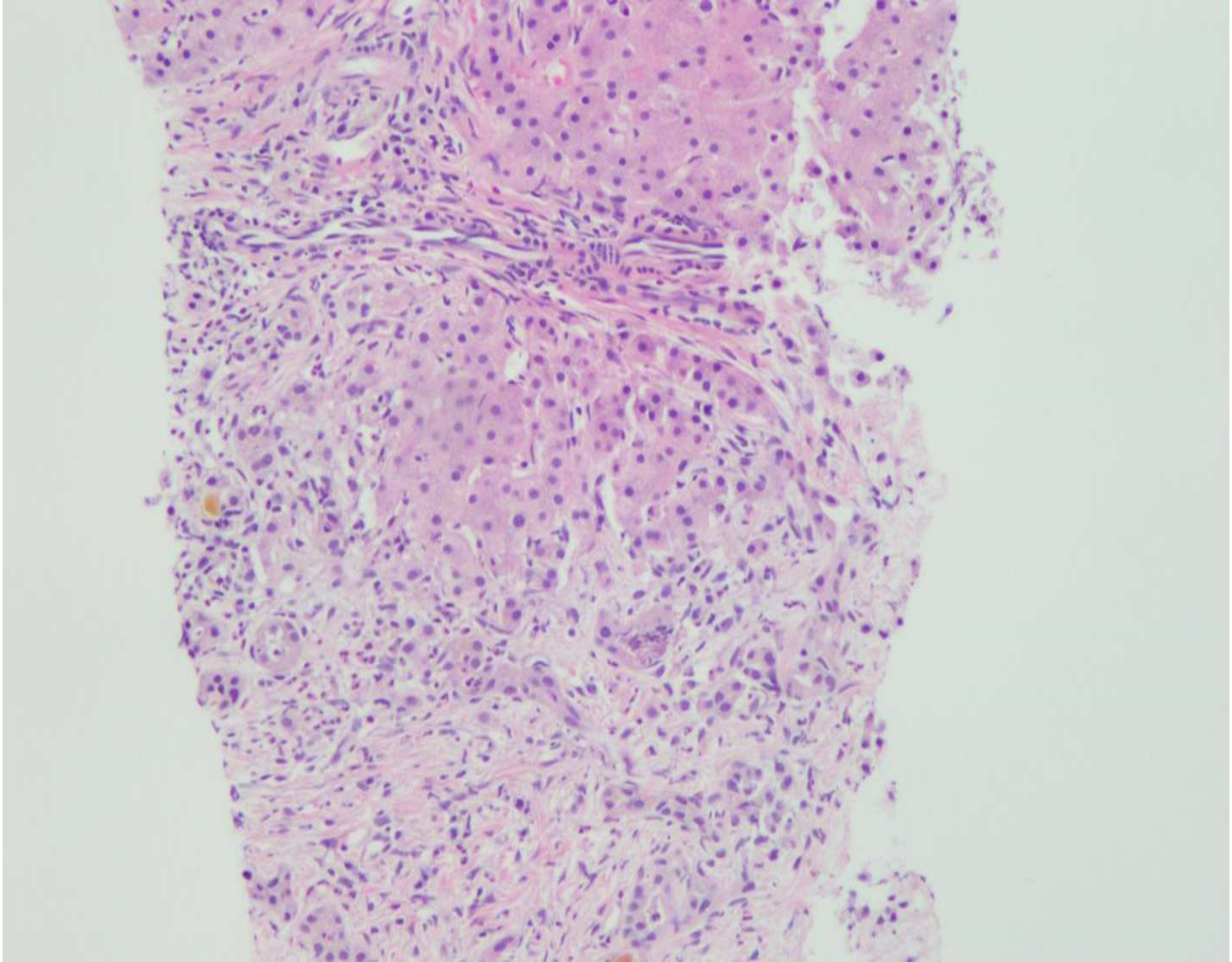

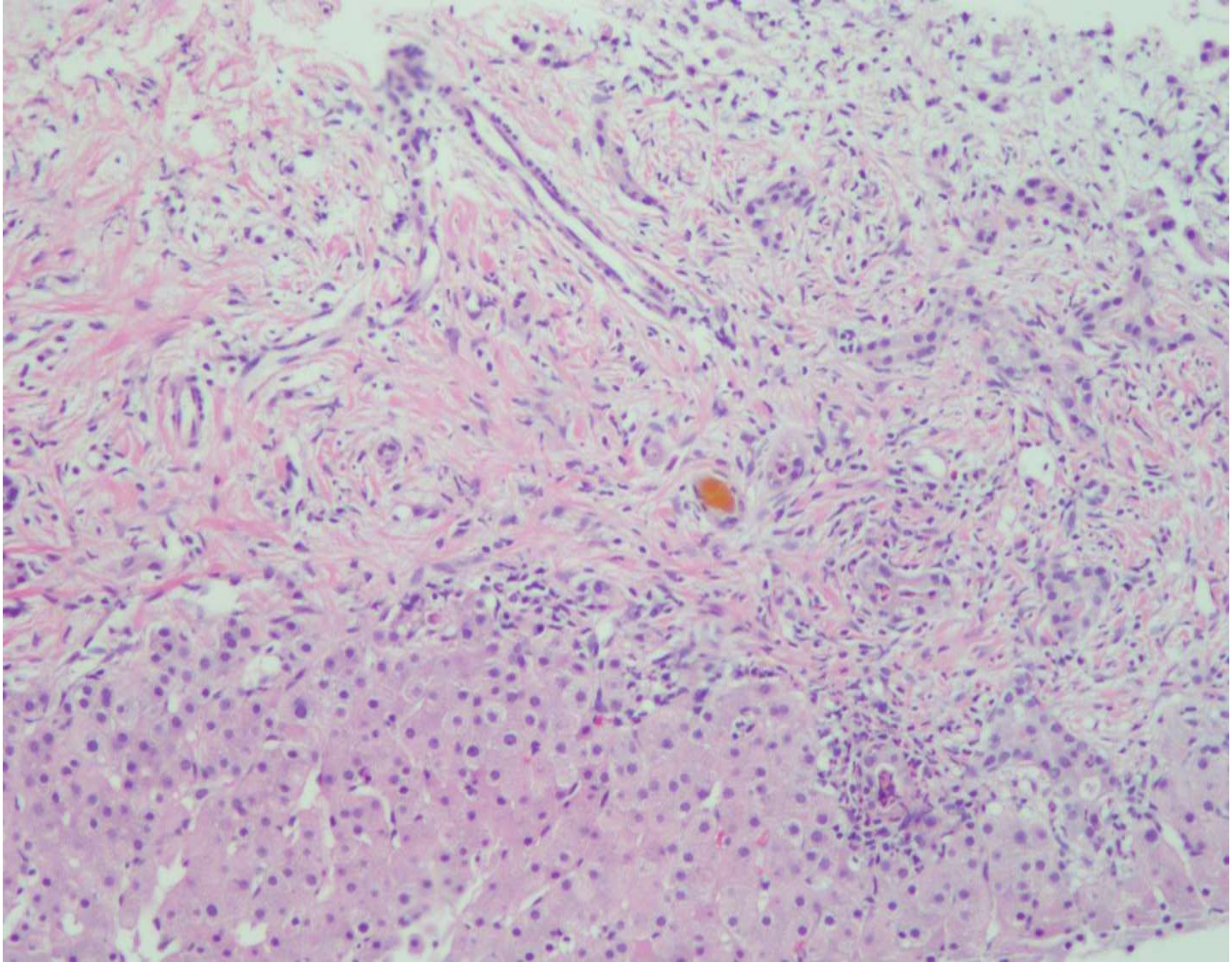

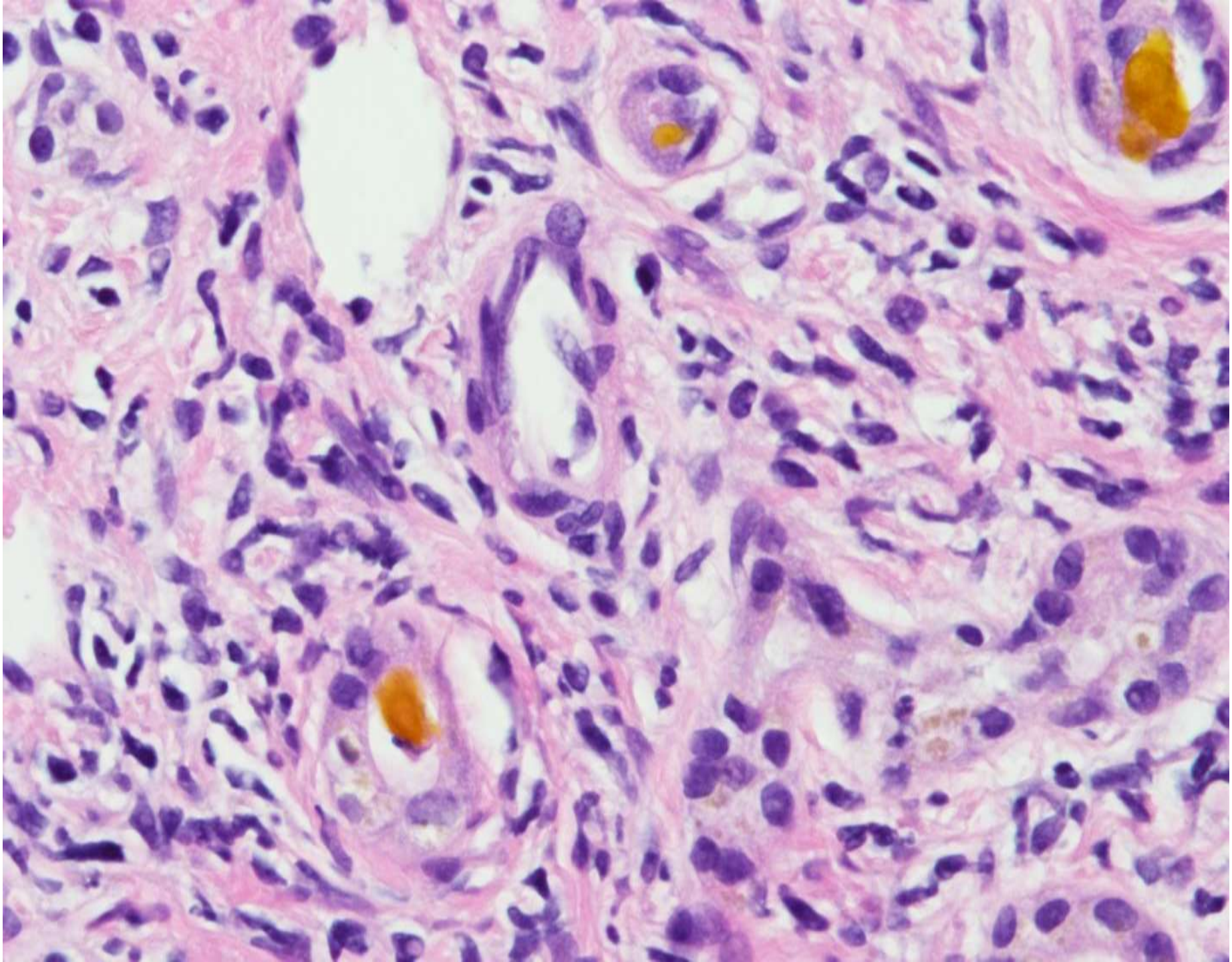

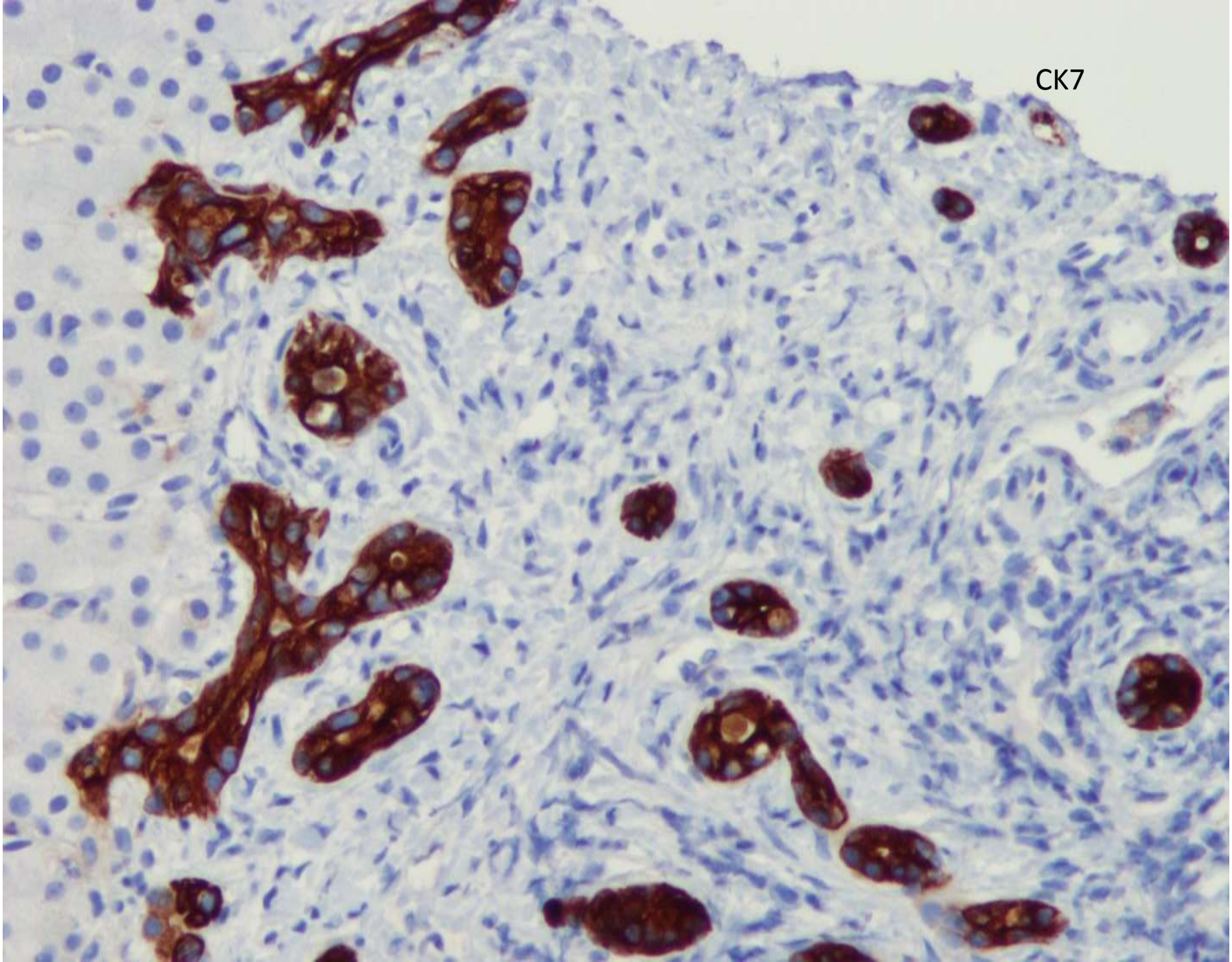
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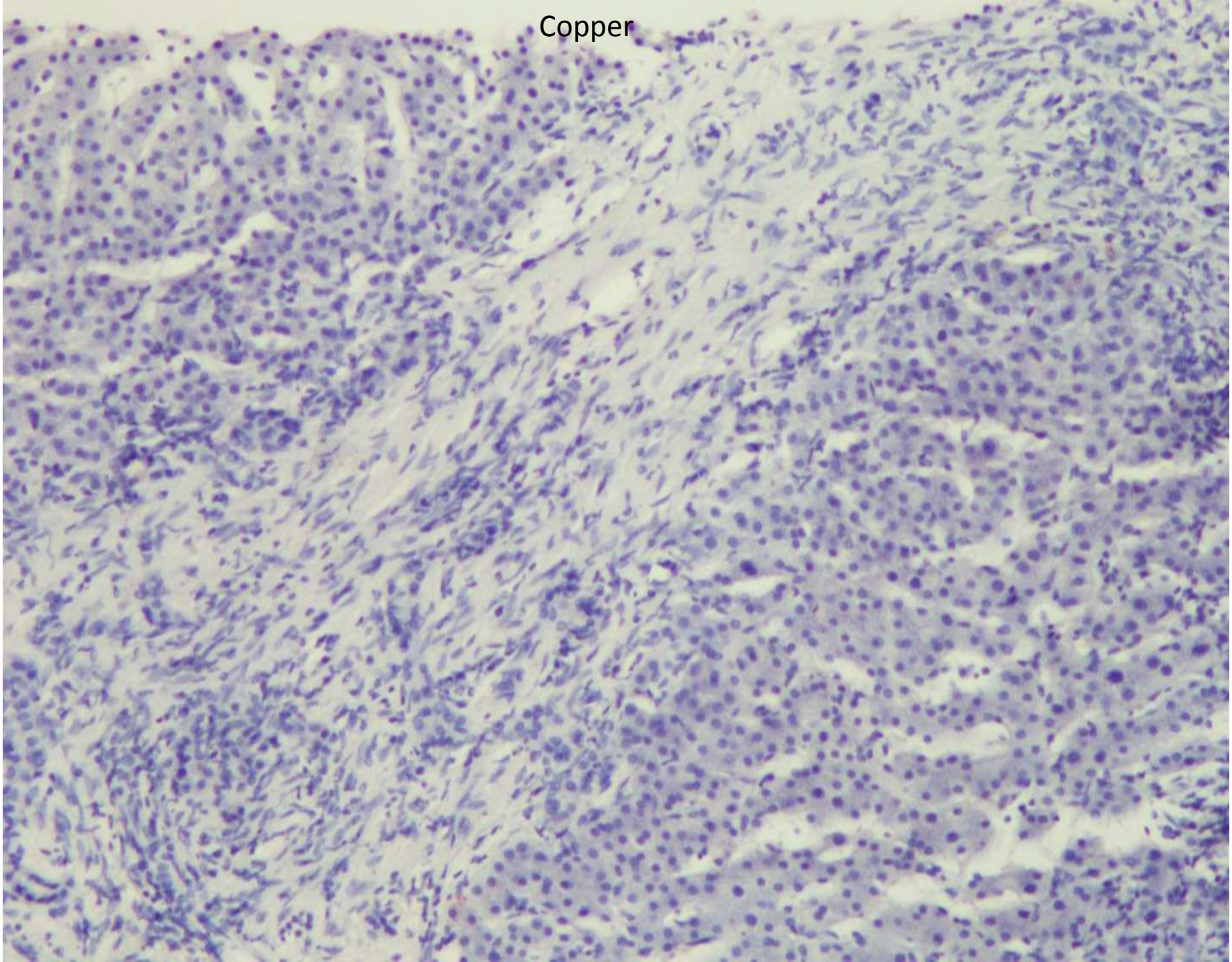
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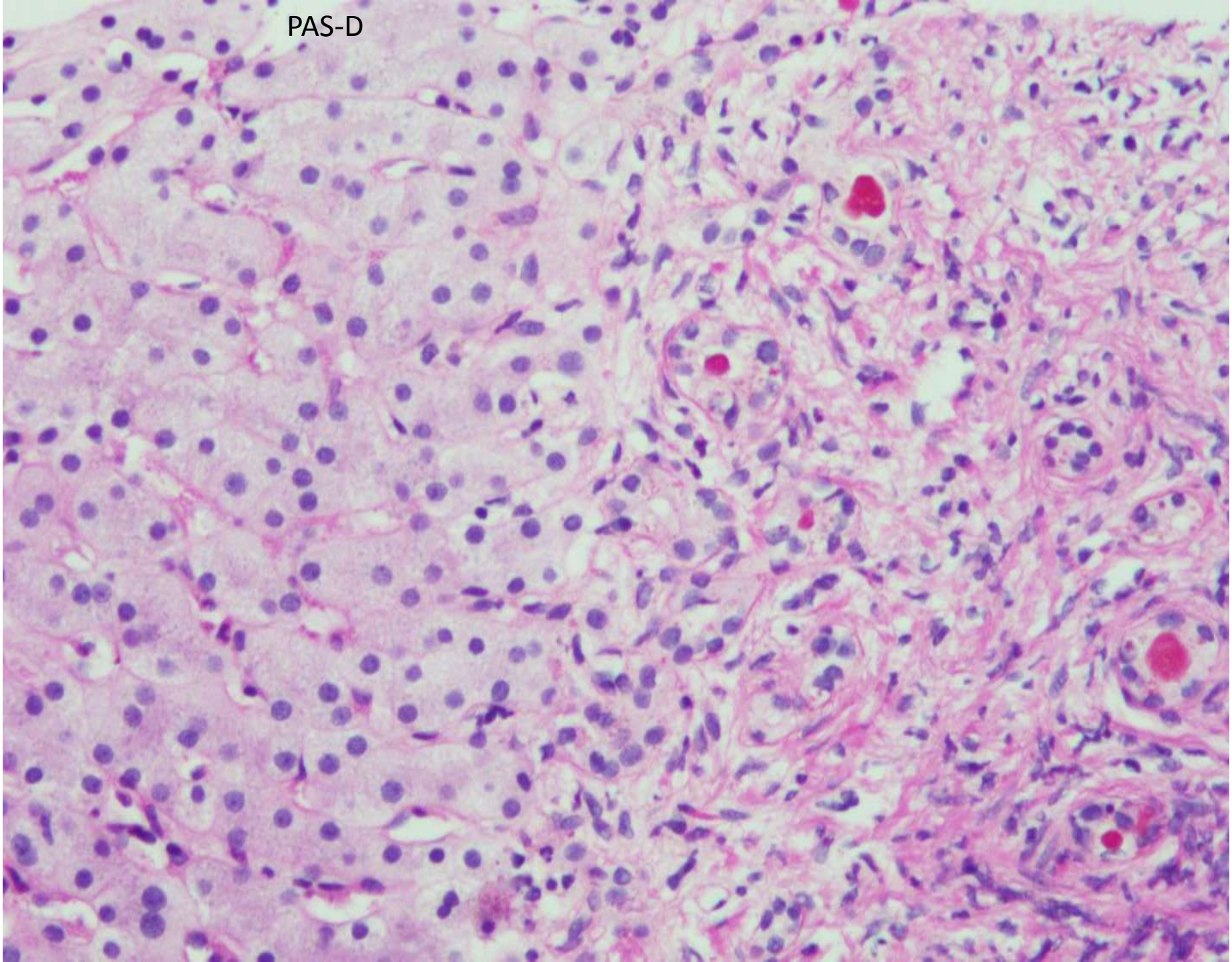
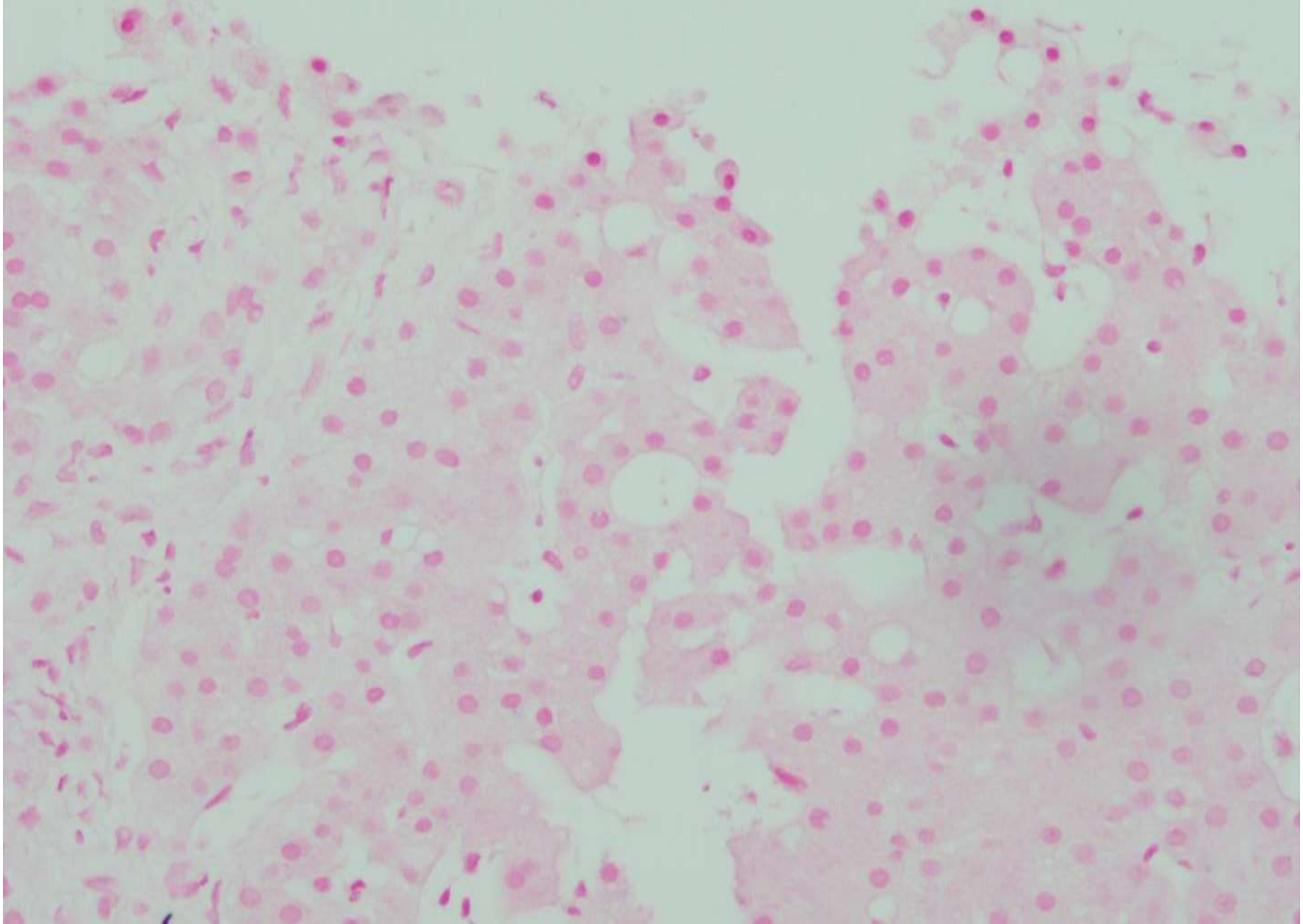

PAS-D

Prussian blue iron

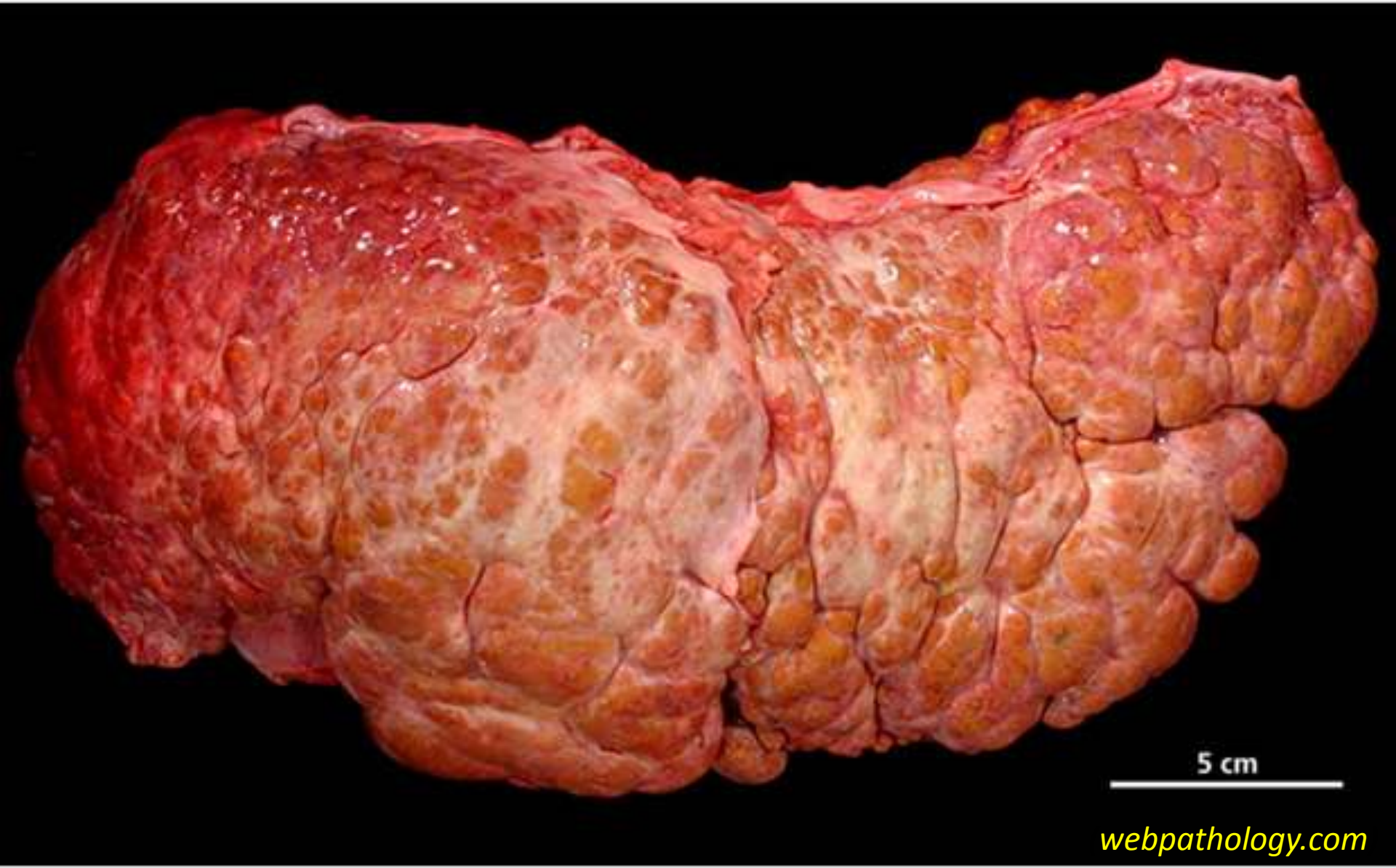


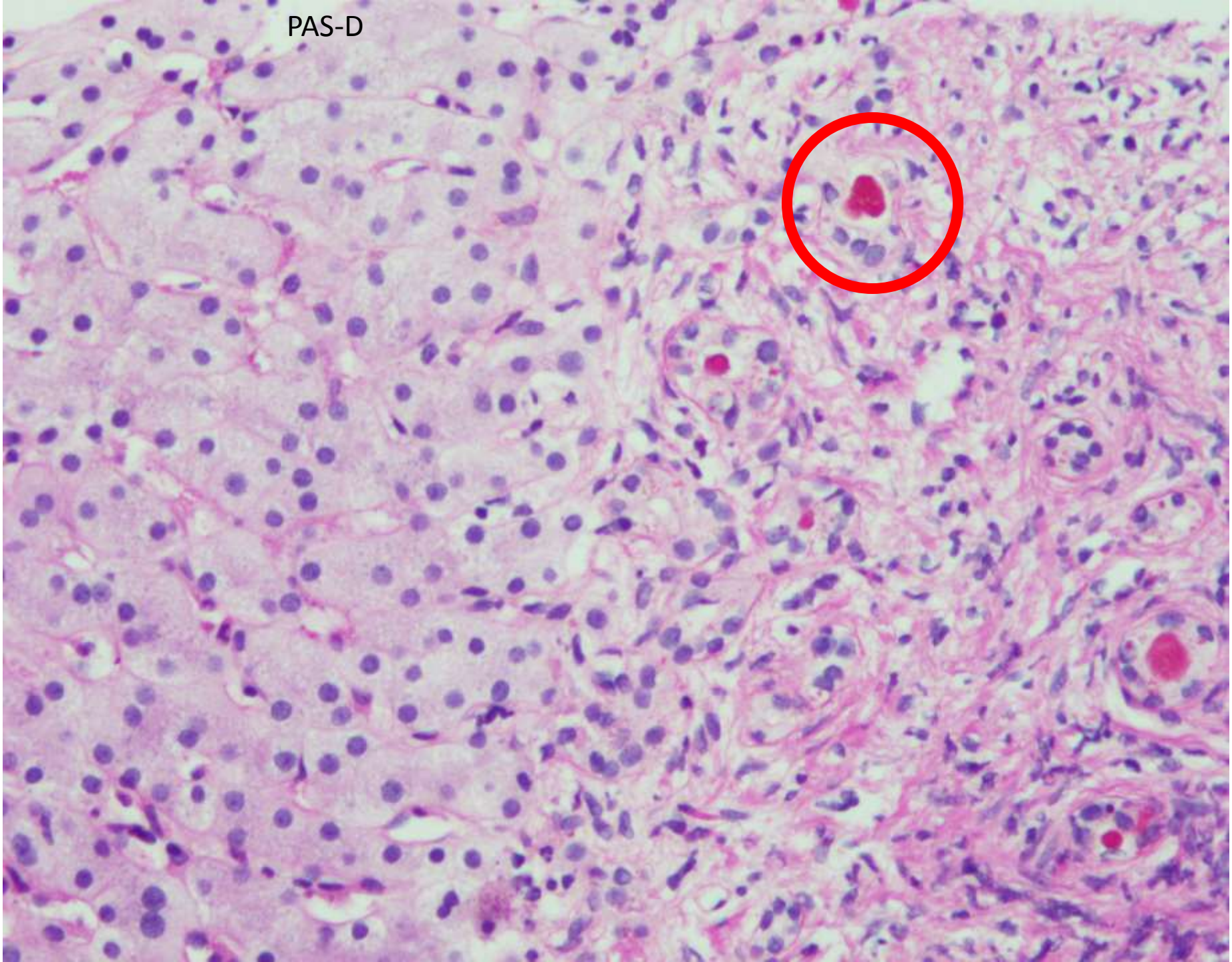
Diagnosis….???

Liver diseases in adolescents

- Nonalcoholic fatty liver disease
- Autoimmune hepatitis
- Viral hepatitis
- Wilson disease
- Alpha-1-antitrypsin deficiency
- Biliary atresia
- Cystic fibrosis
- Primary sclerosing cholangitis
- Alagille syndrome
- Progressive familial intrahepatic cholestasis

Cirrhosis as presentation of liver disease in adolescents is uncommon




PAS-D

Hepatic cystic fibrosis

- Liver disease peaks in late childhood / adolescence; Rarely presents as liver disease
- Histology:
  - Ductules with PASD+ eosinophilic secretions
  - Biliary fibrosis → “multilobular biliary cirrhosis”
  - Fatty change common
- Therapy: Ursodeoxycholic Acid (UDCA); potential transplant

Follow Up from 5/5/2015

Assessment:

15 y old with Chiari malformation, mild ASD, FTT, chronic loose stools admitted in January 2015 for MSSA PNA, later developed a pneumothorax. Found to have liver cirrhosis and pancreatic atrophy/insufficiency. Liver biopsy on 1/28 showed cholestatic cirrhosis with diastase resistant PAS staining, highly suggestive of cystic fibrosis. Eventually diagnosed with cystic fibrosis.

Since last clinic visit with me, pt has been doing well, stools have become more solid, now basically normal, though stools are very large. Still taking Actigall and Fe.

Plan:

- No need to recheck stool studies since pt's stools are now formed.
- Plan to repeat liver labs, CBC, and Fe studies.
- Given pt's diagnosis of cystic fibrosis and h/o cholestatic cirrhosis, recommend continuing Actigall indefinitely, and check liver labs every 6 months, annual US.
- No need for further GI clinic appointments unless other issues come up.

Adolescents: Special Liver Disease Population

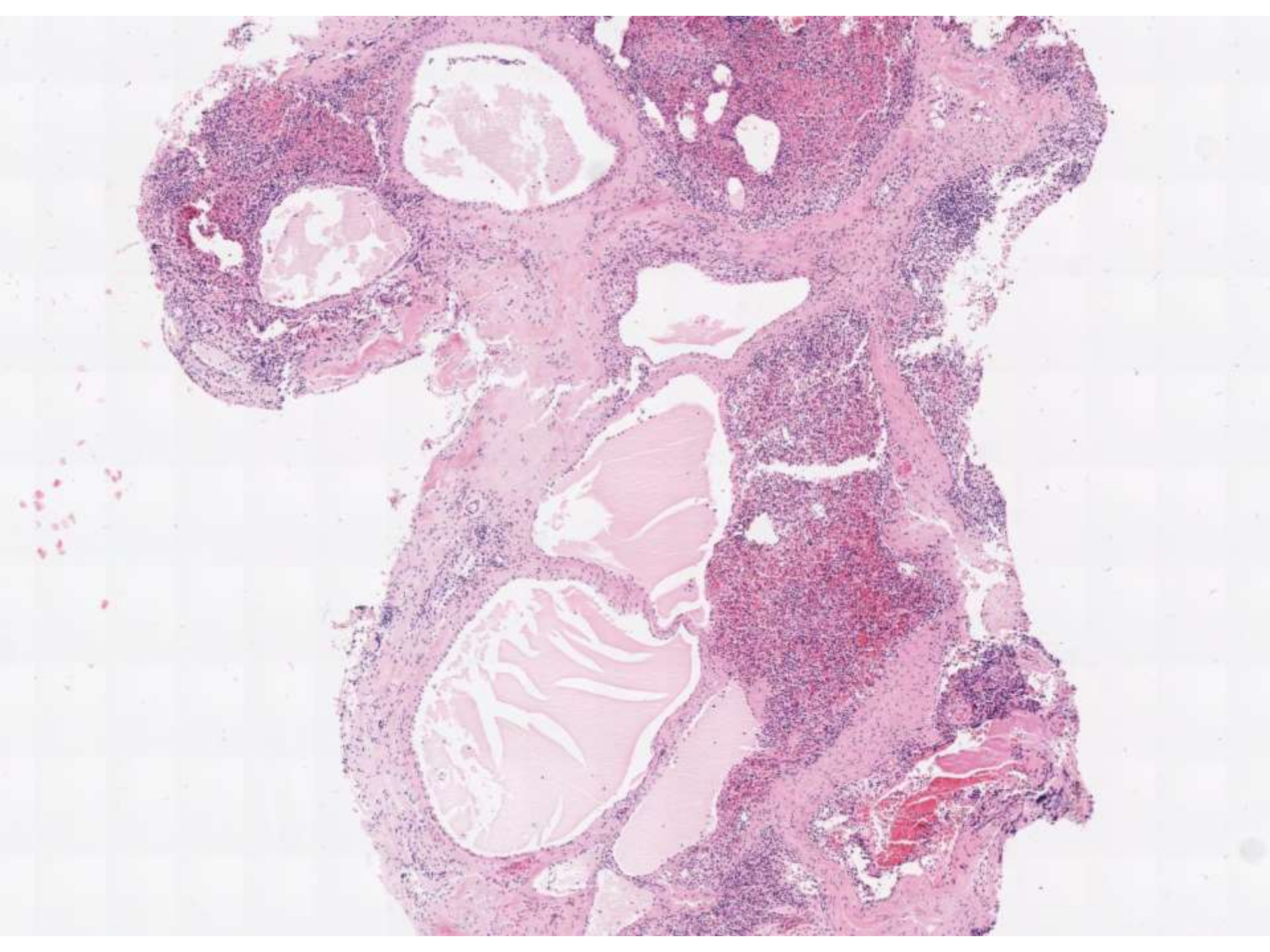
"Due to the development changes occurring in adolescence, monitoring for adherence, risk-taking behaviors, and signs of psychological diseases, such as anxiety and depression, is imperative when caring for an adolescent with liver disease."

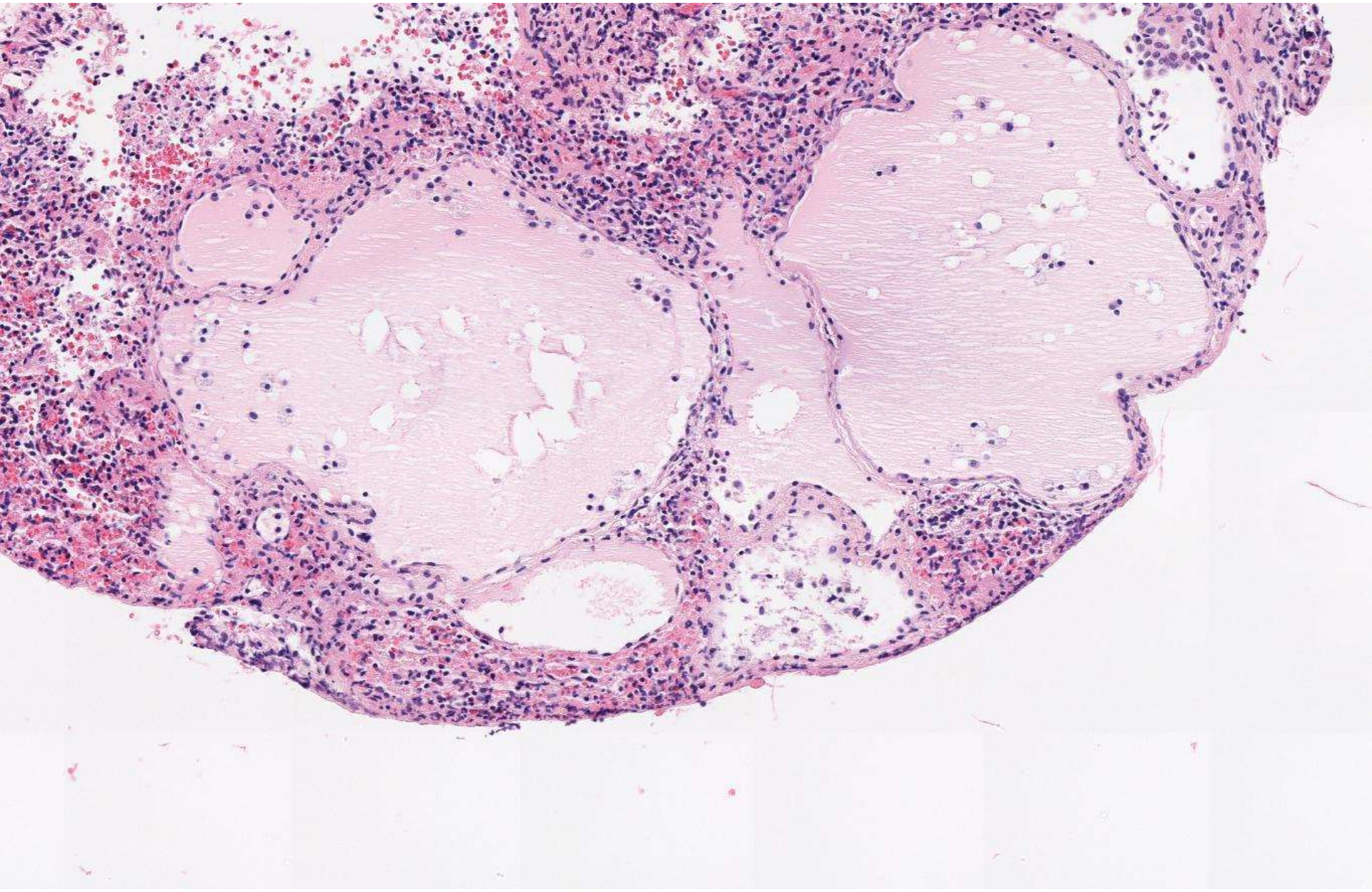
Mavis AM1, Alonso EM2. Liver disease in the adolescent. *Clin Liver Dis*. 2015 Feb;19(1):171-85.

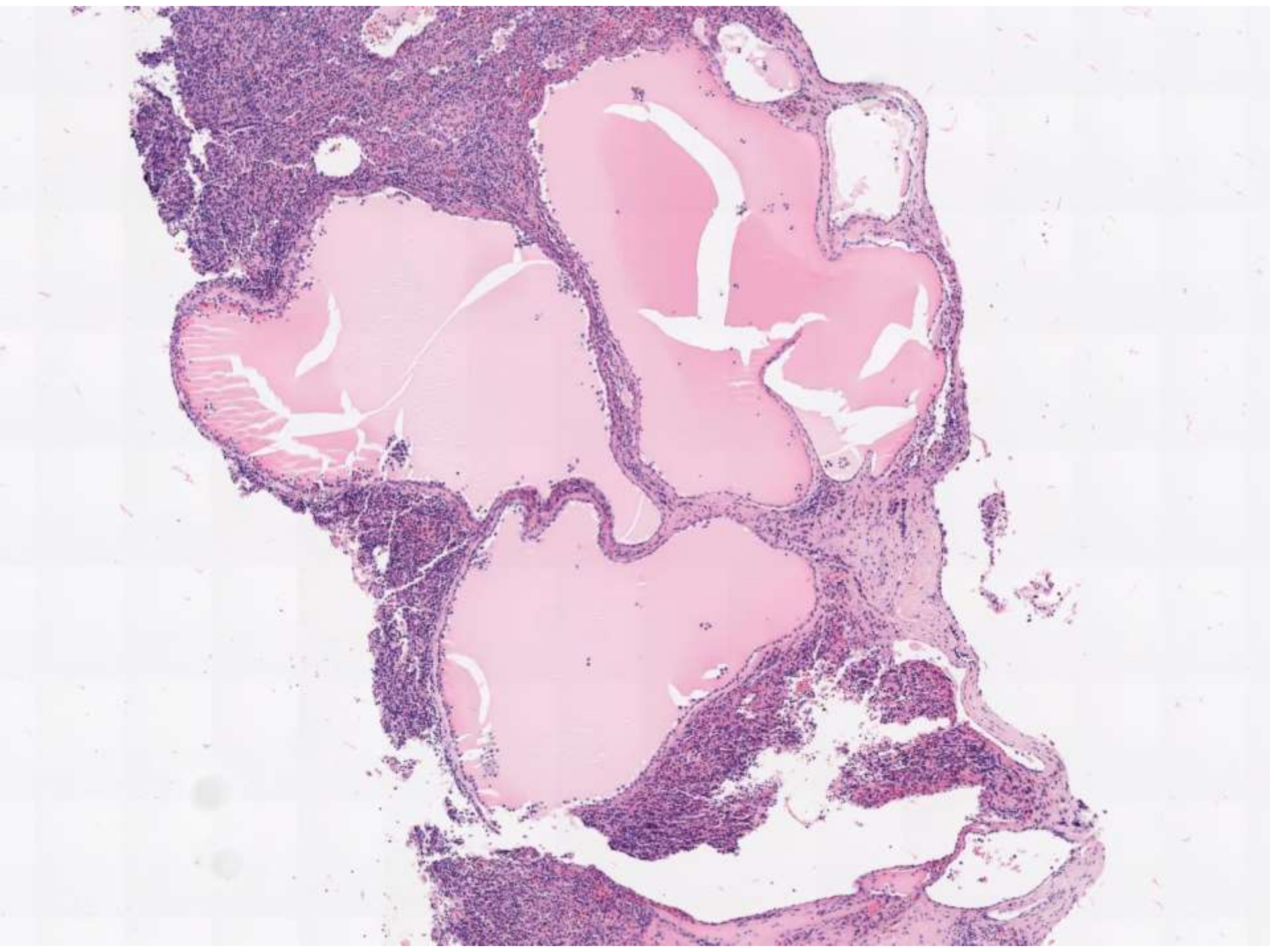
SB 5942

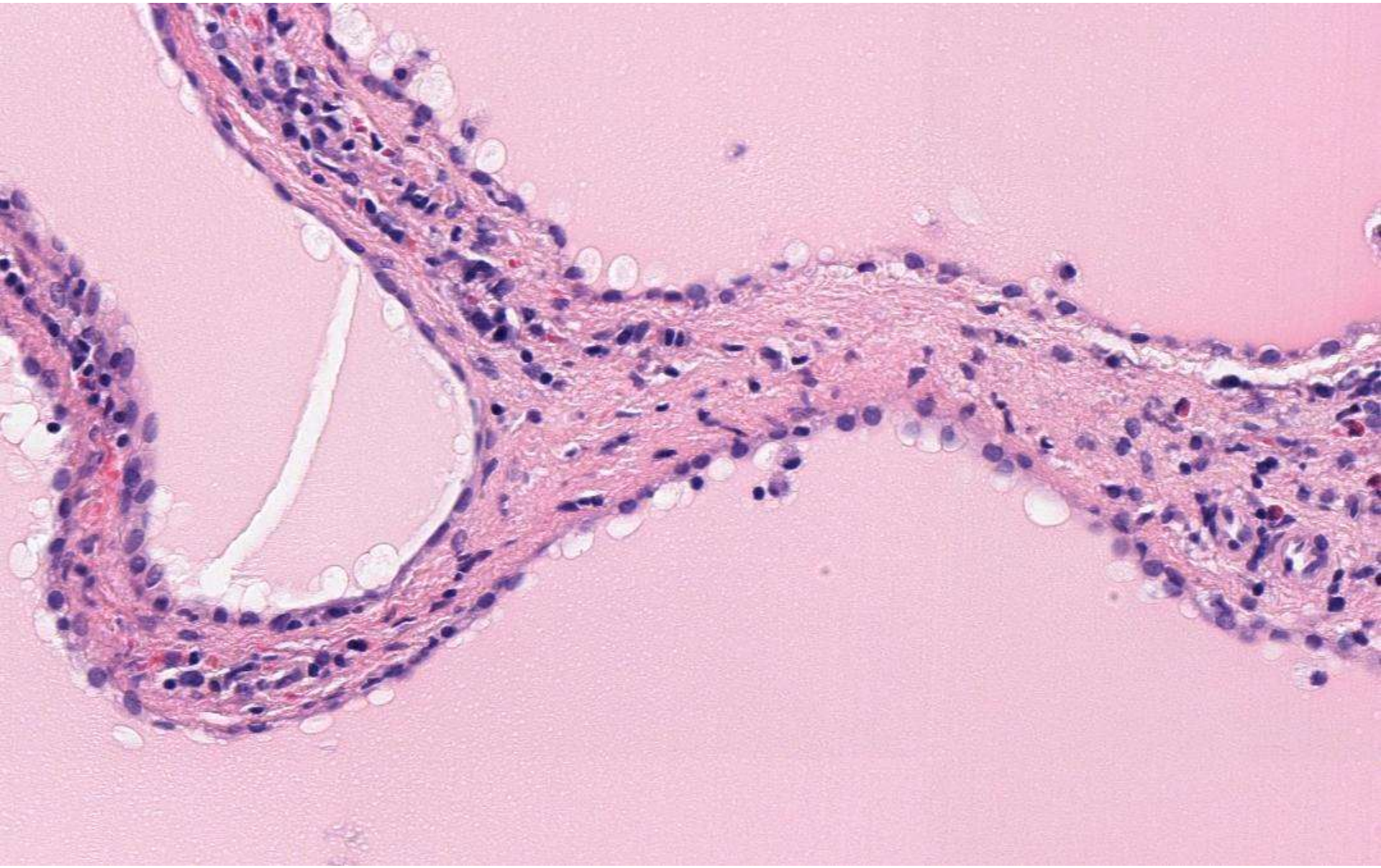
- 57-year-old woman with a history of GE reflux disease and bilateral dyskinesia who underwent cholecystectomy. A splenic capsular lesion was seen and biopsied.

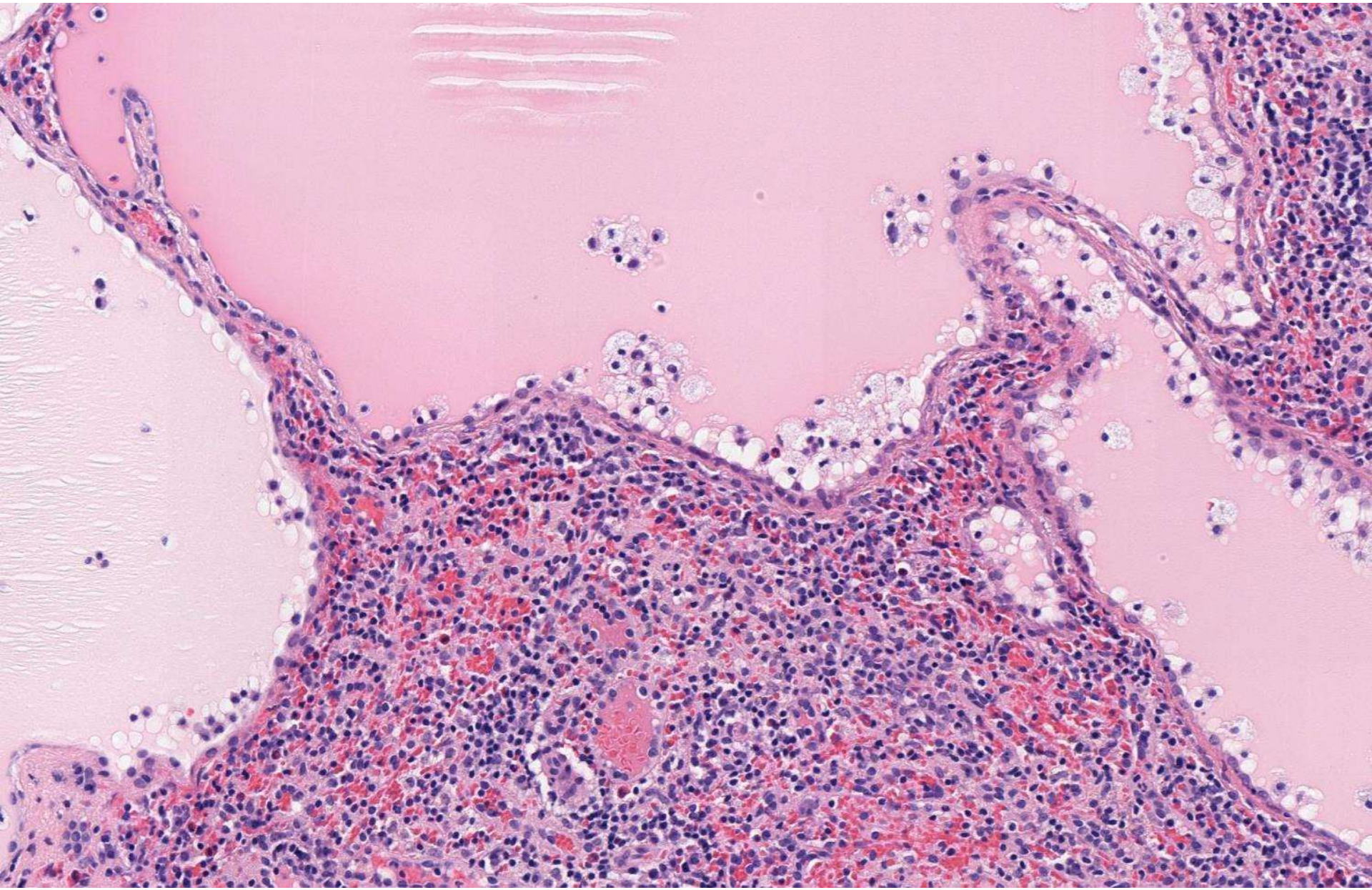
- **Charles Zaloudek; UCSF**



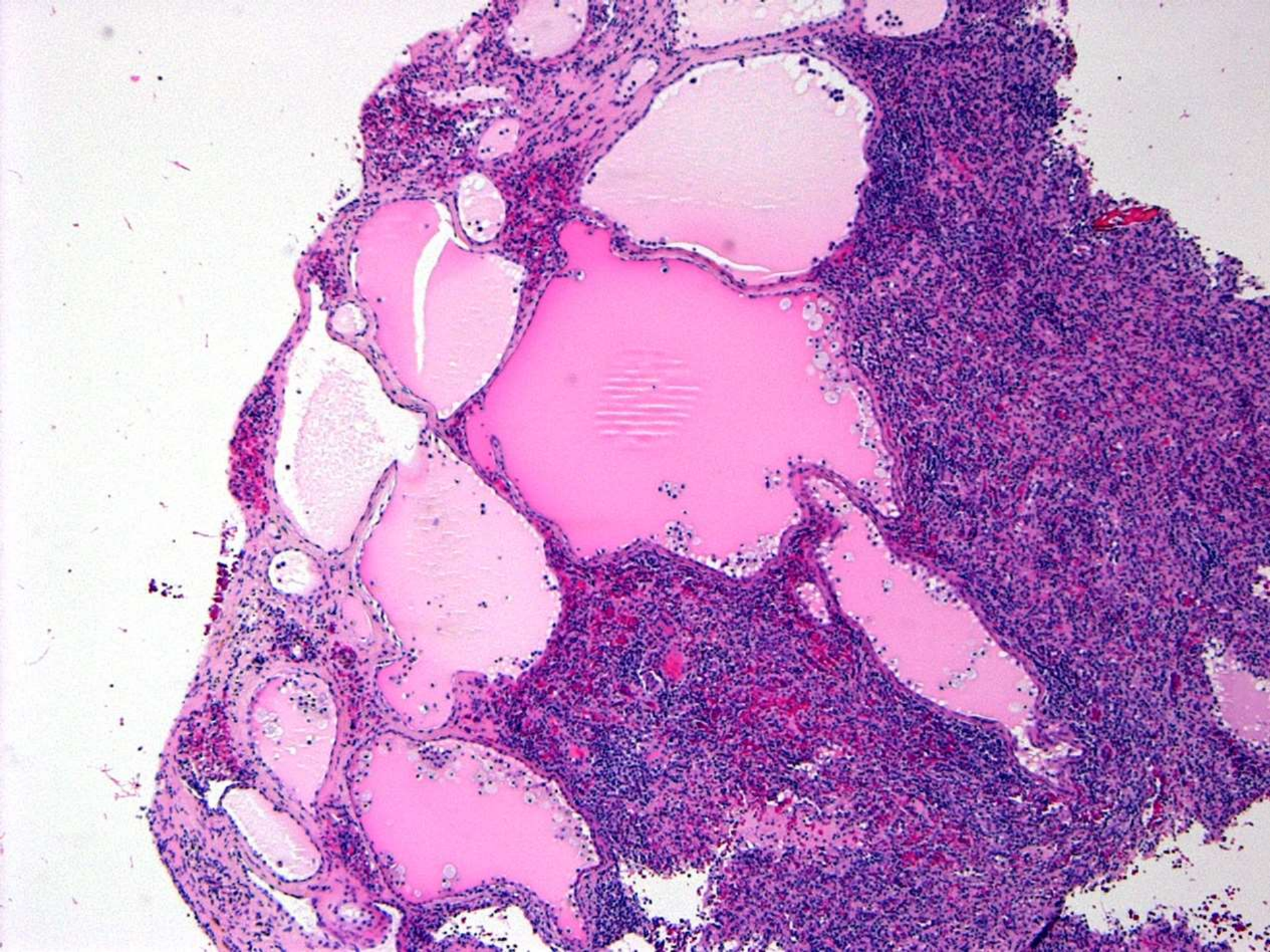


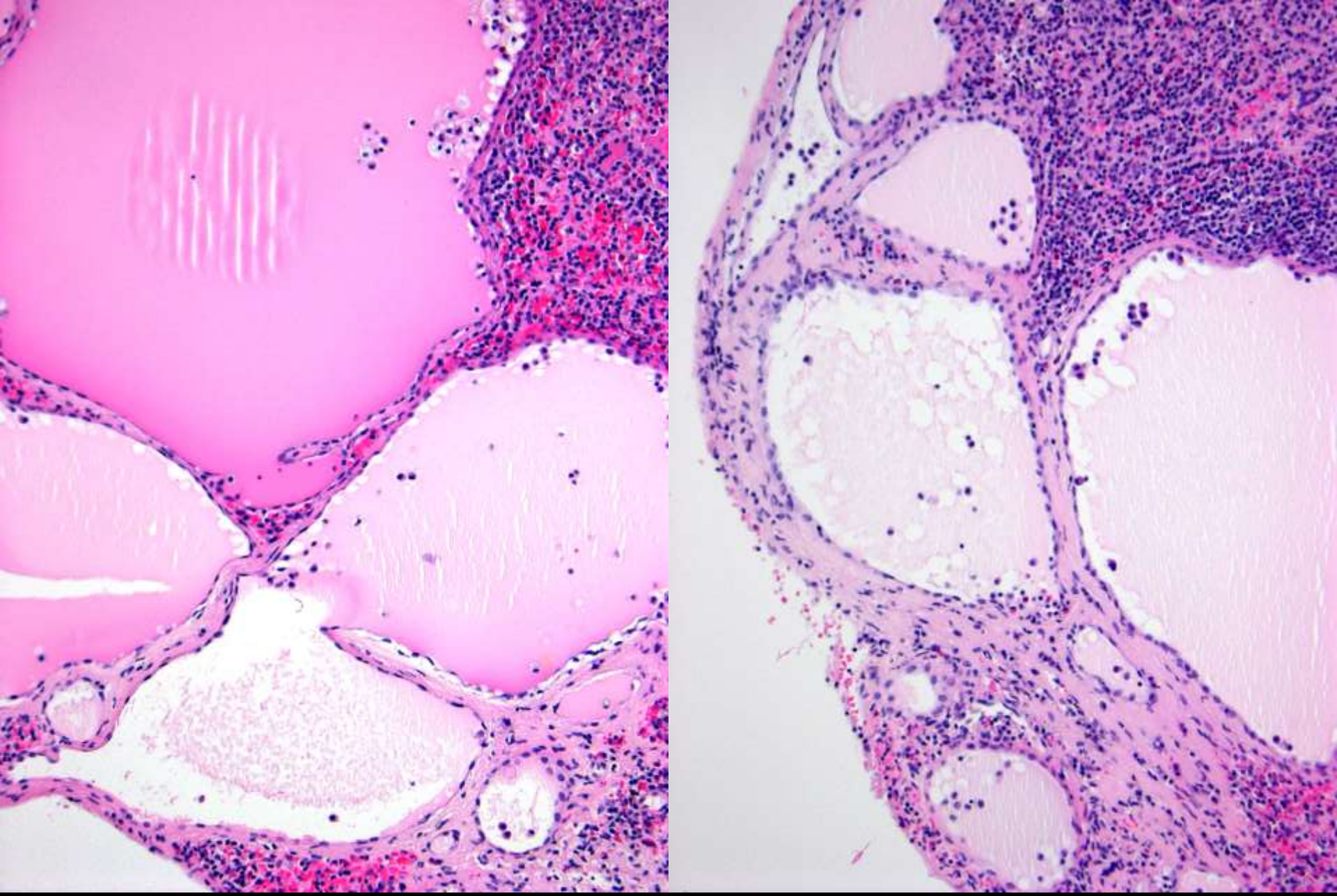


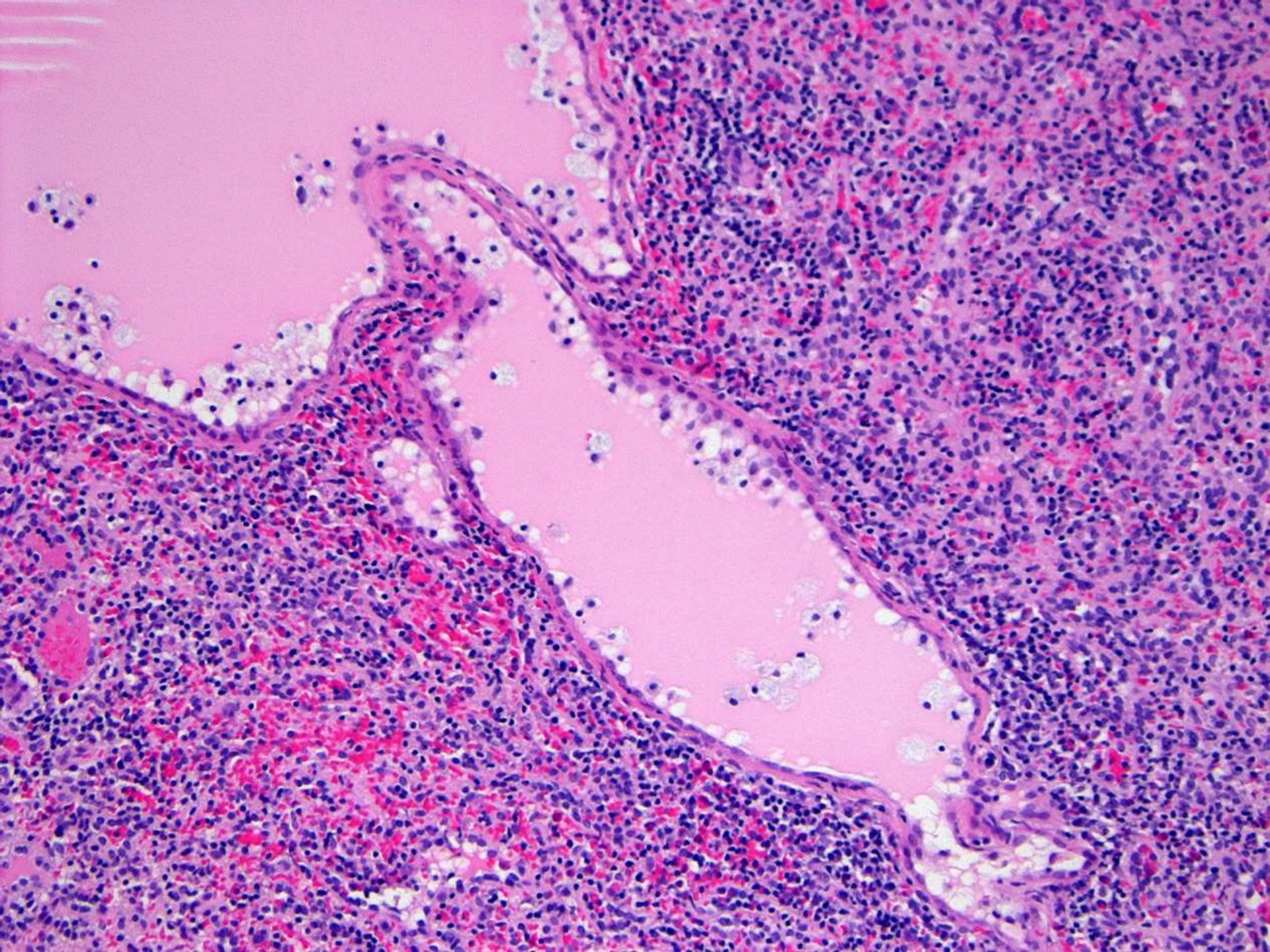


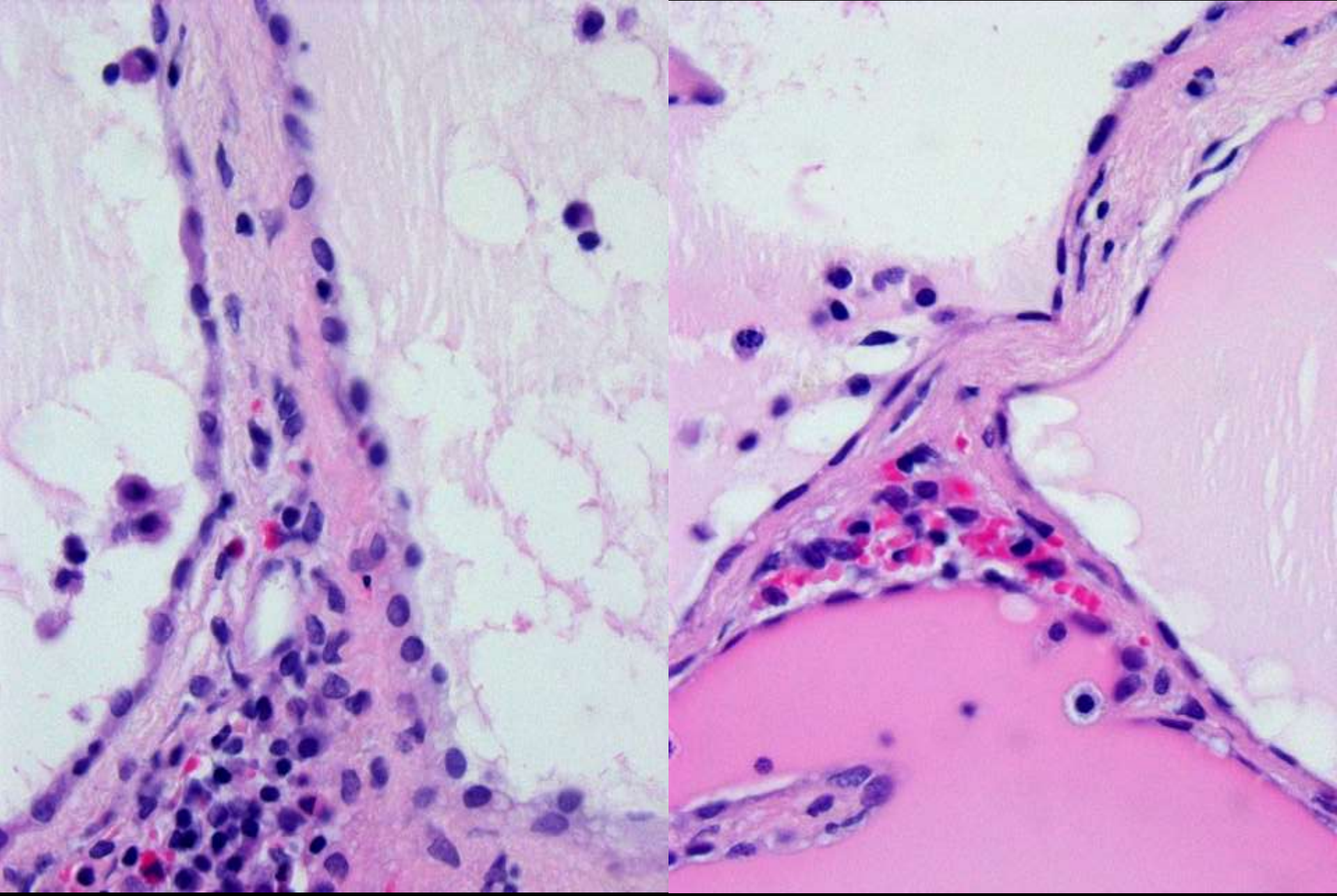


Diagnosis….???



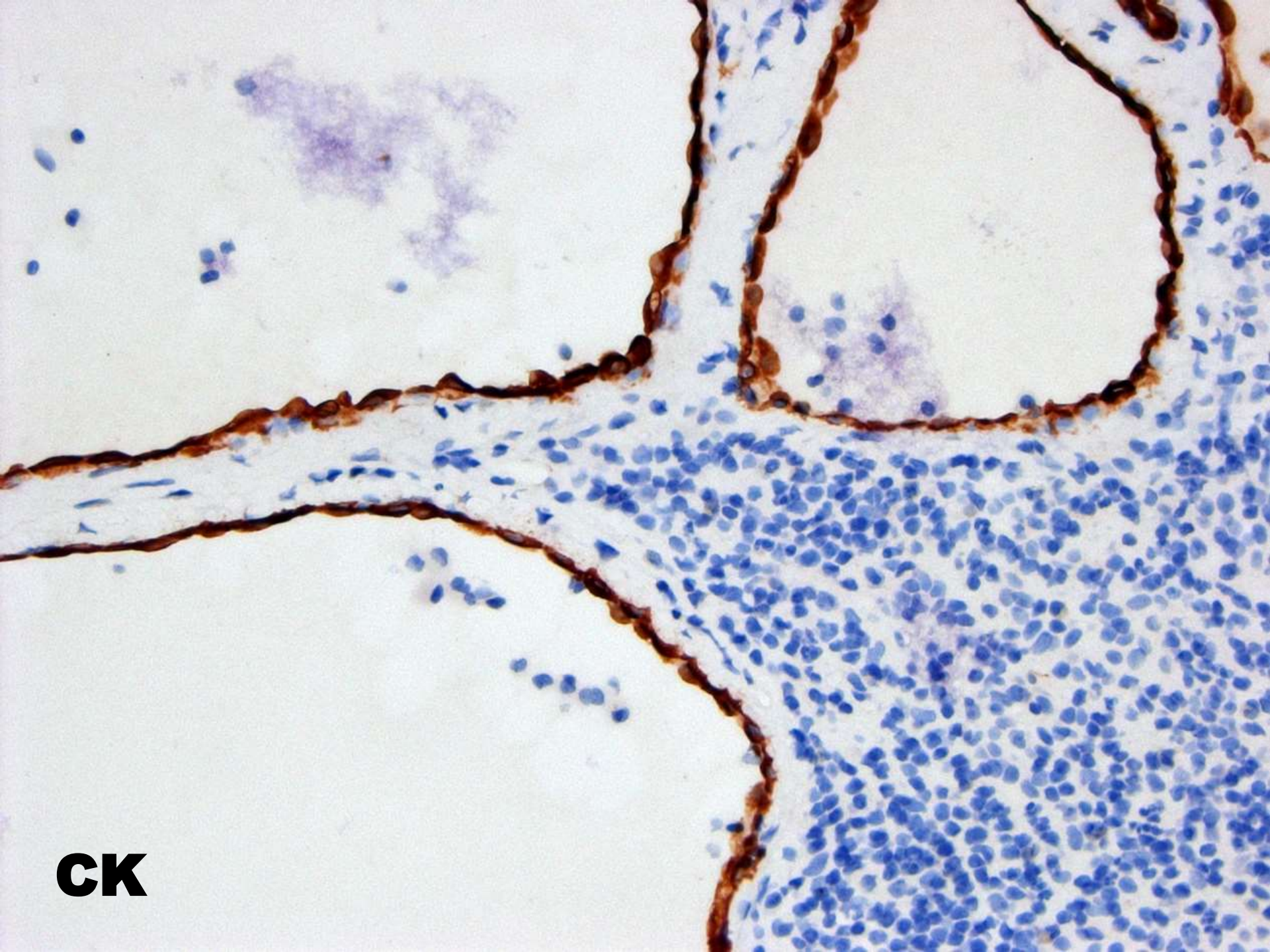






Differential Diagnosis

- **Hemangioma – CD31+, CD34+, CD8-**
- **Littoral cell angioma – CD31+, CD34-, CD8-, CD68+**
- **Lymphangioma – D2-40+**
- **Malignant vascular tumor**
- **Mesothelial cysts – CD31-, CD34-, CD8-, keratin +, D2-40+, calretinin +**


CK

Immunohistochemistry

- **Cytokeratin – positive**
- **D2-40 – positive**
- **Calretinin – positive**
- **CD31 – negative**
- **CD34 – negative**
- **CD8 - negative**

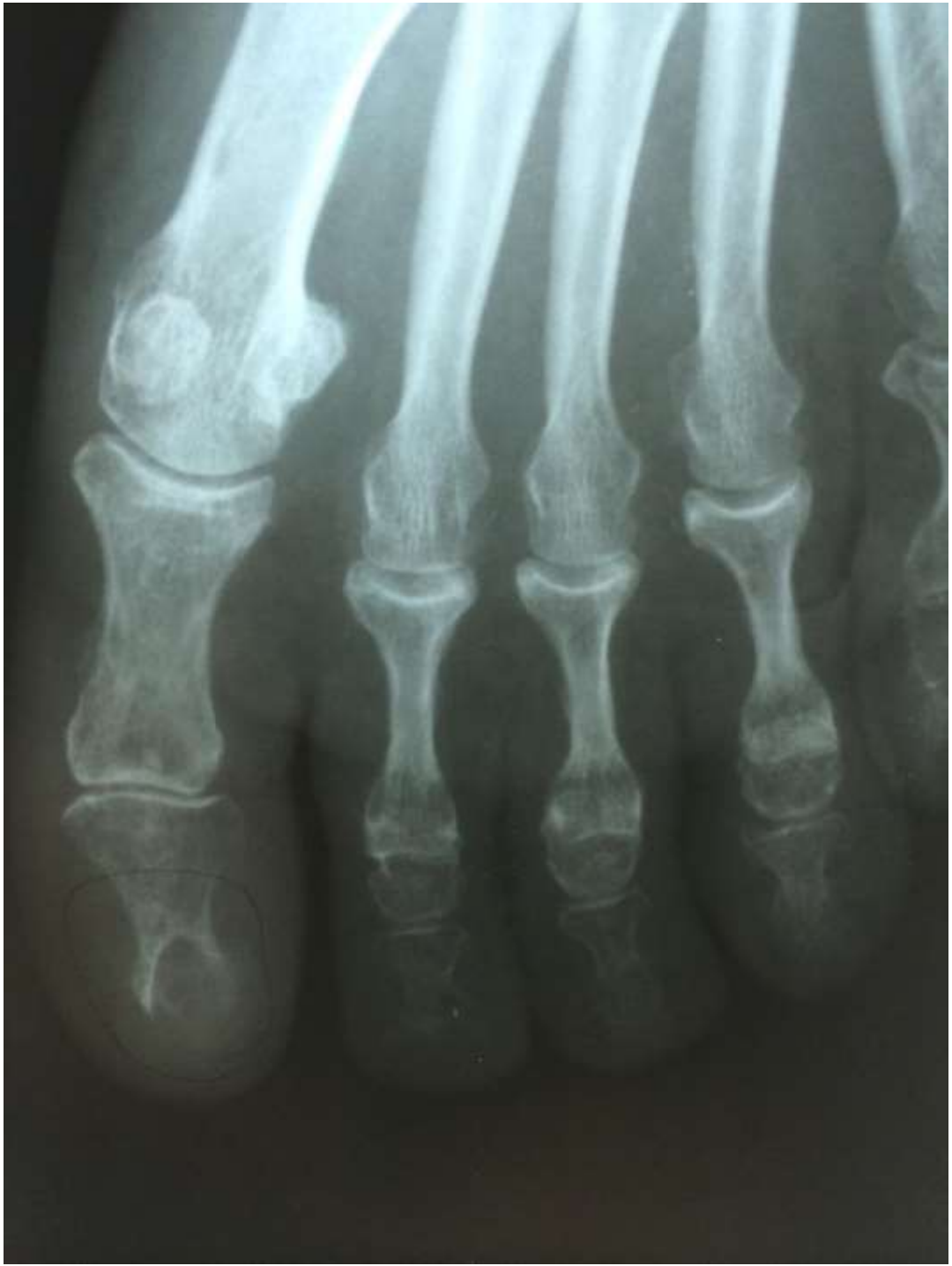
Arber, D. A., et al. (1997). "Splenic mesothelial cysts mimicking lymphangiomas." American Journal of Surgical Pathology 21(3): 334-338.

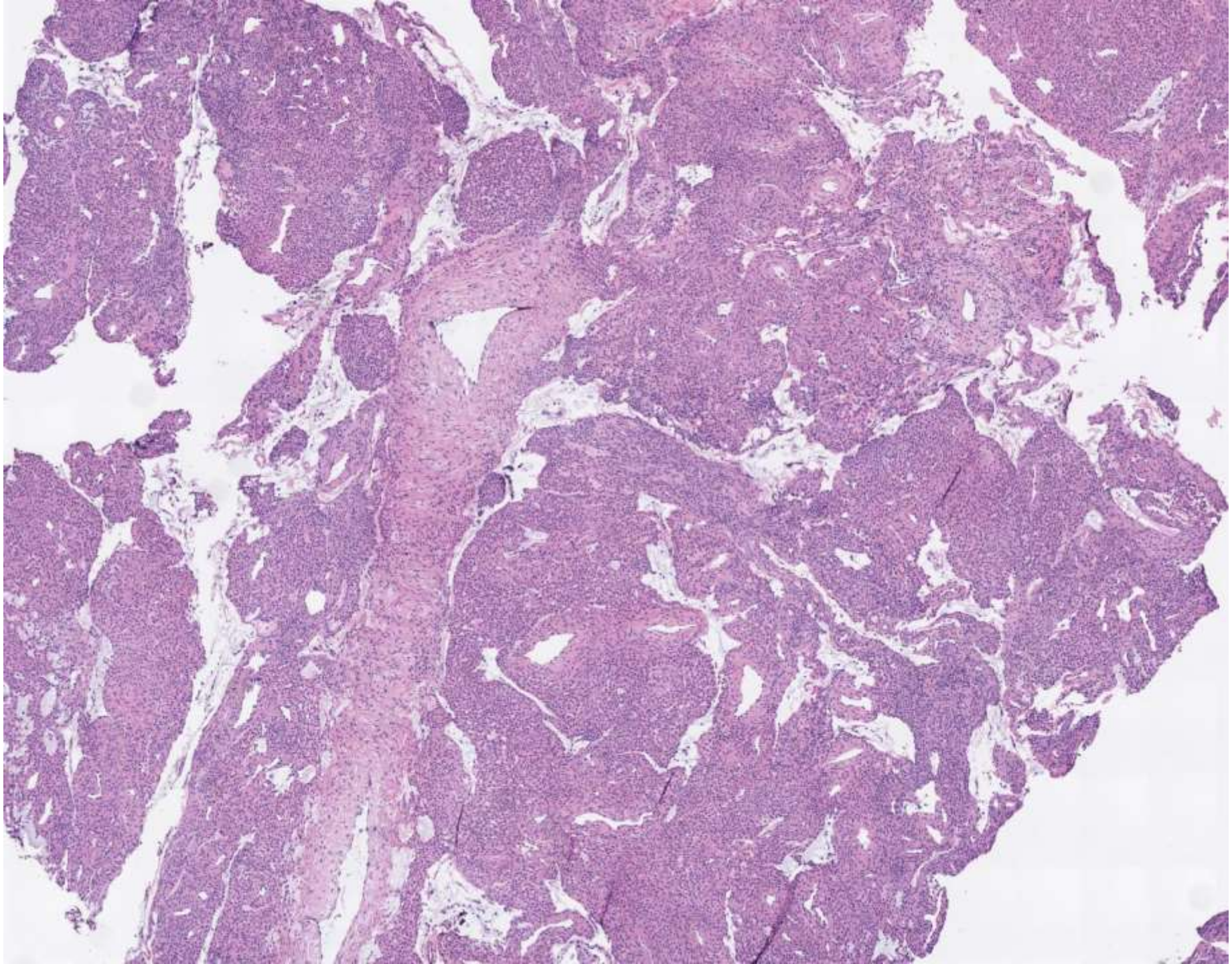
- **6 tumors with morphological features similar to those described for lymphangiomas**
- **All incidental findings in splenectomy specimens**
- **Lining cells + for keratin and HBME-1**
- **Lining cells – for CD31, CD34, F8**
- **Mesothelial derivation**

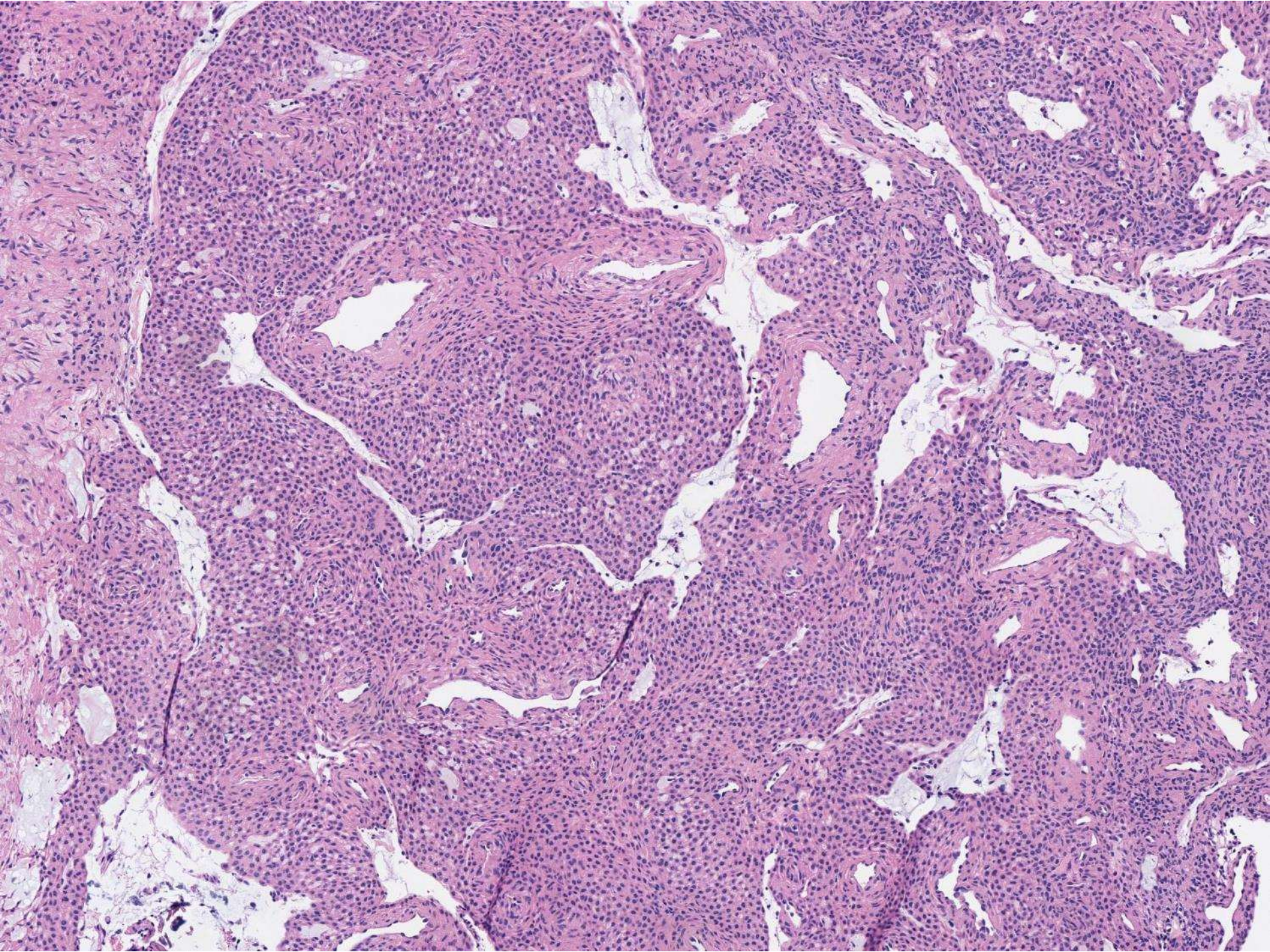
SB 5943

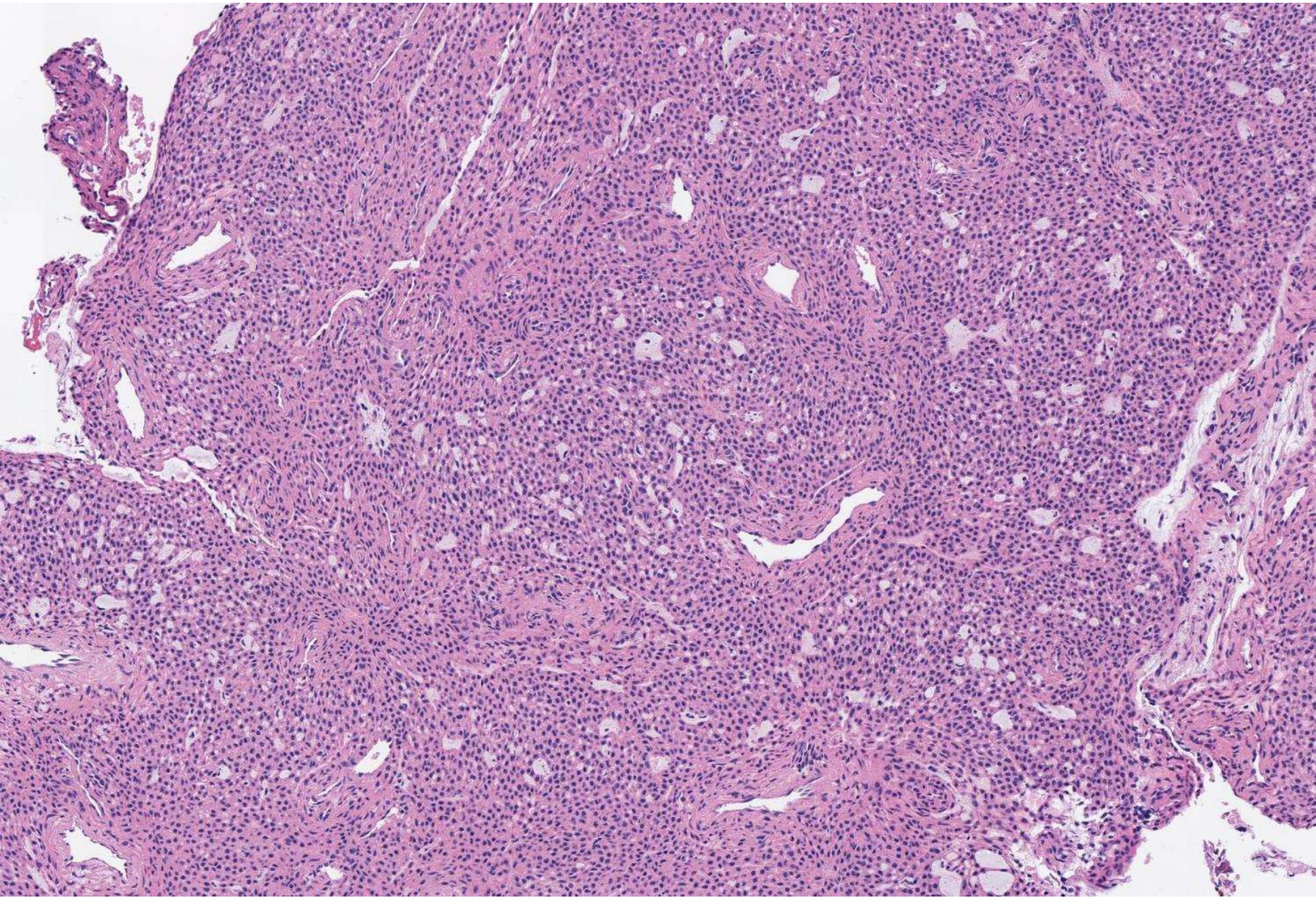
- 59-year-old woman with lytic lesion in distal phalanx of right big toe.

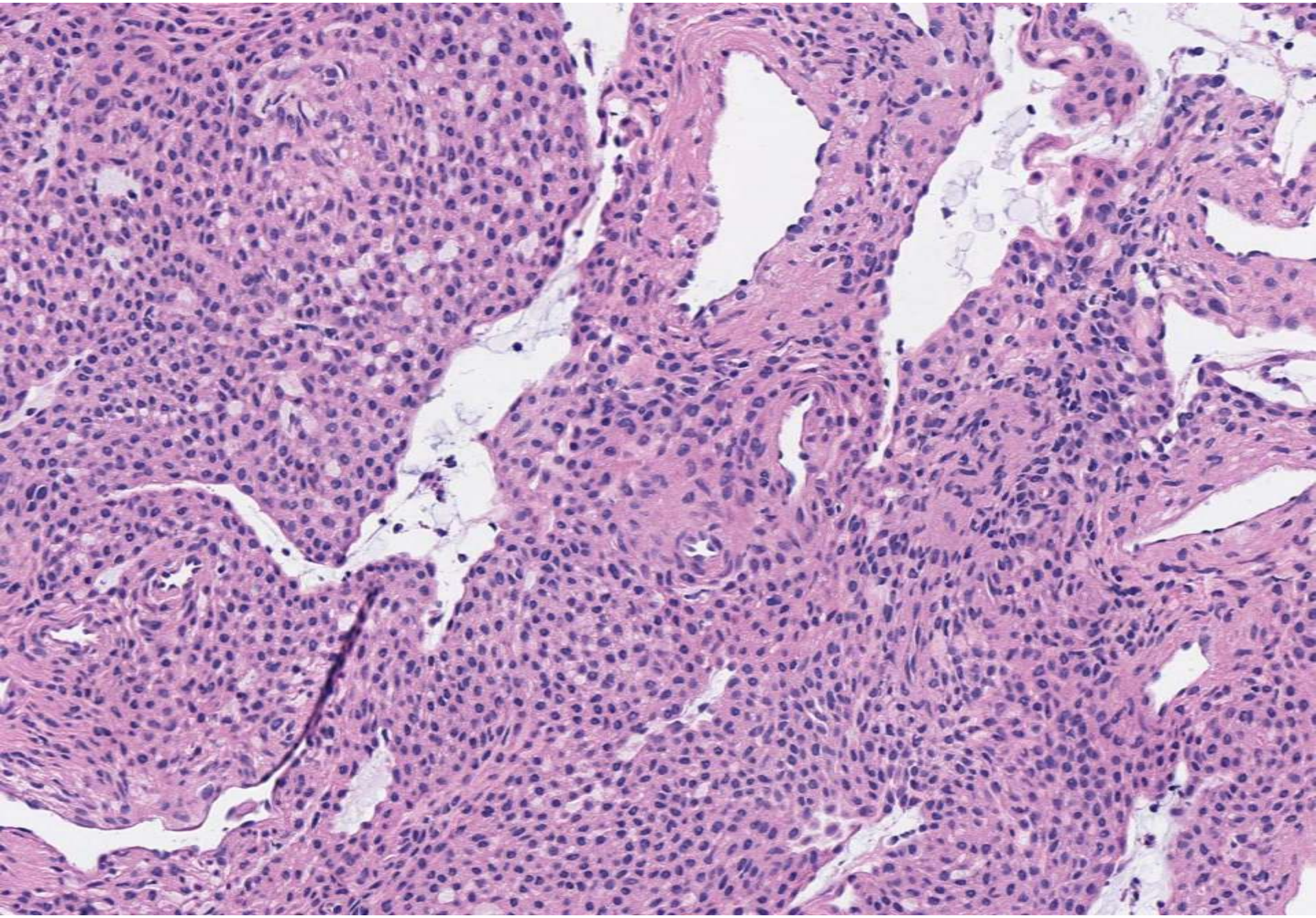
- **Mahendra Ranchod; Good Samaritan Hospital**

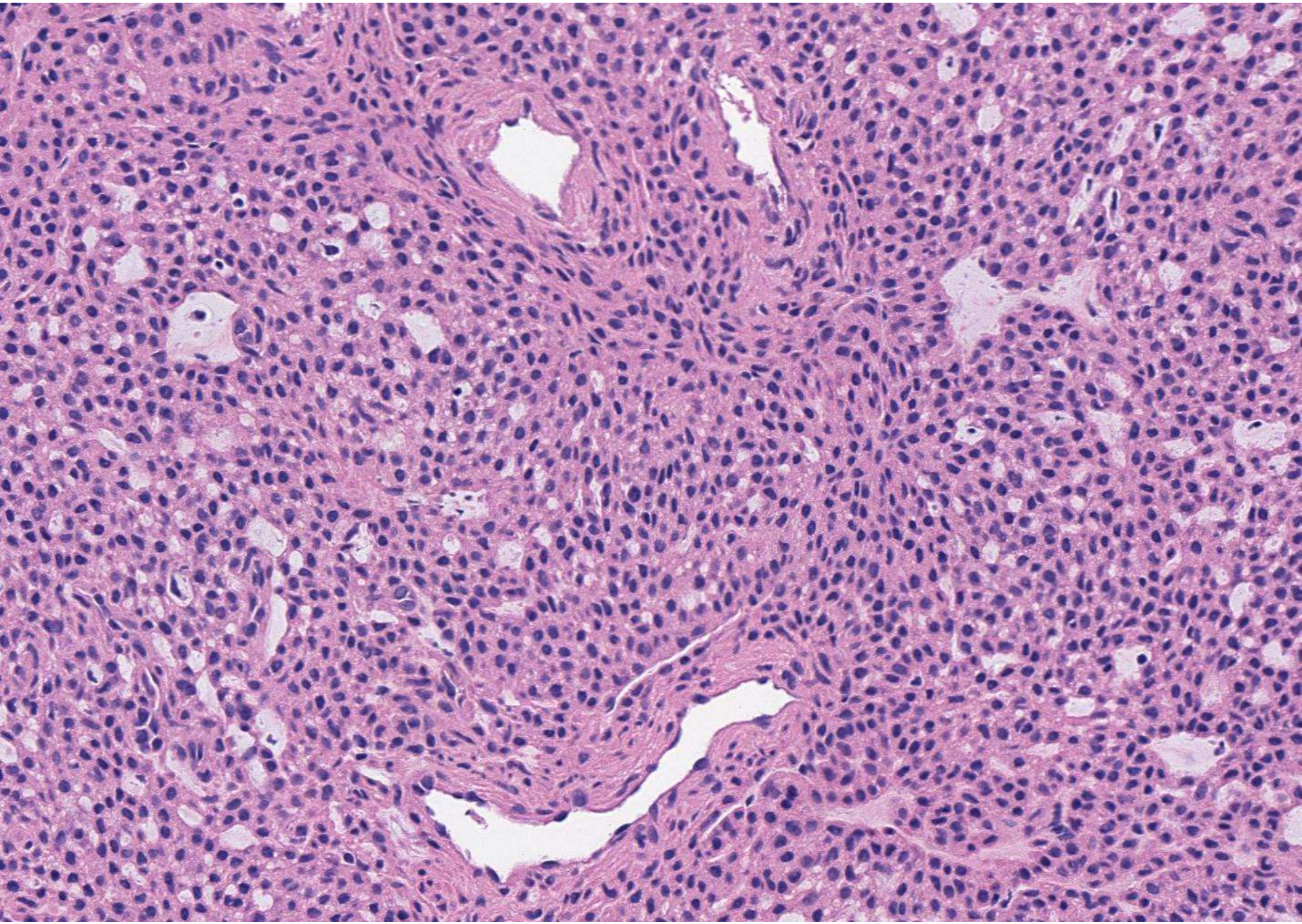




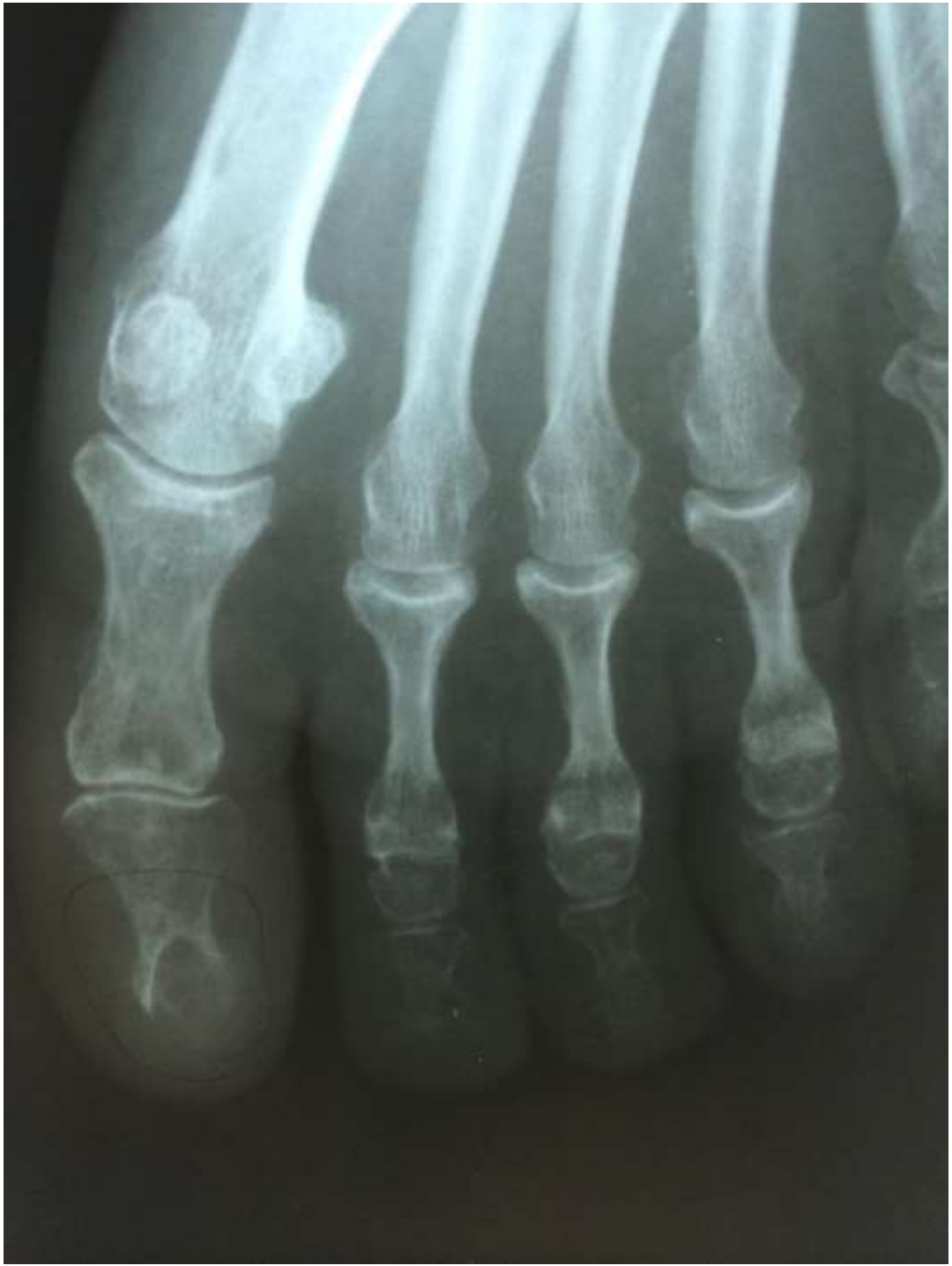


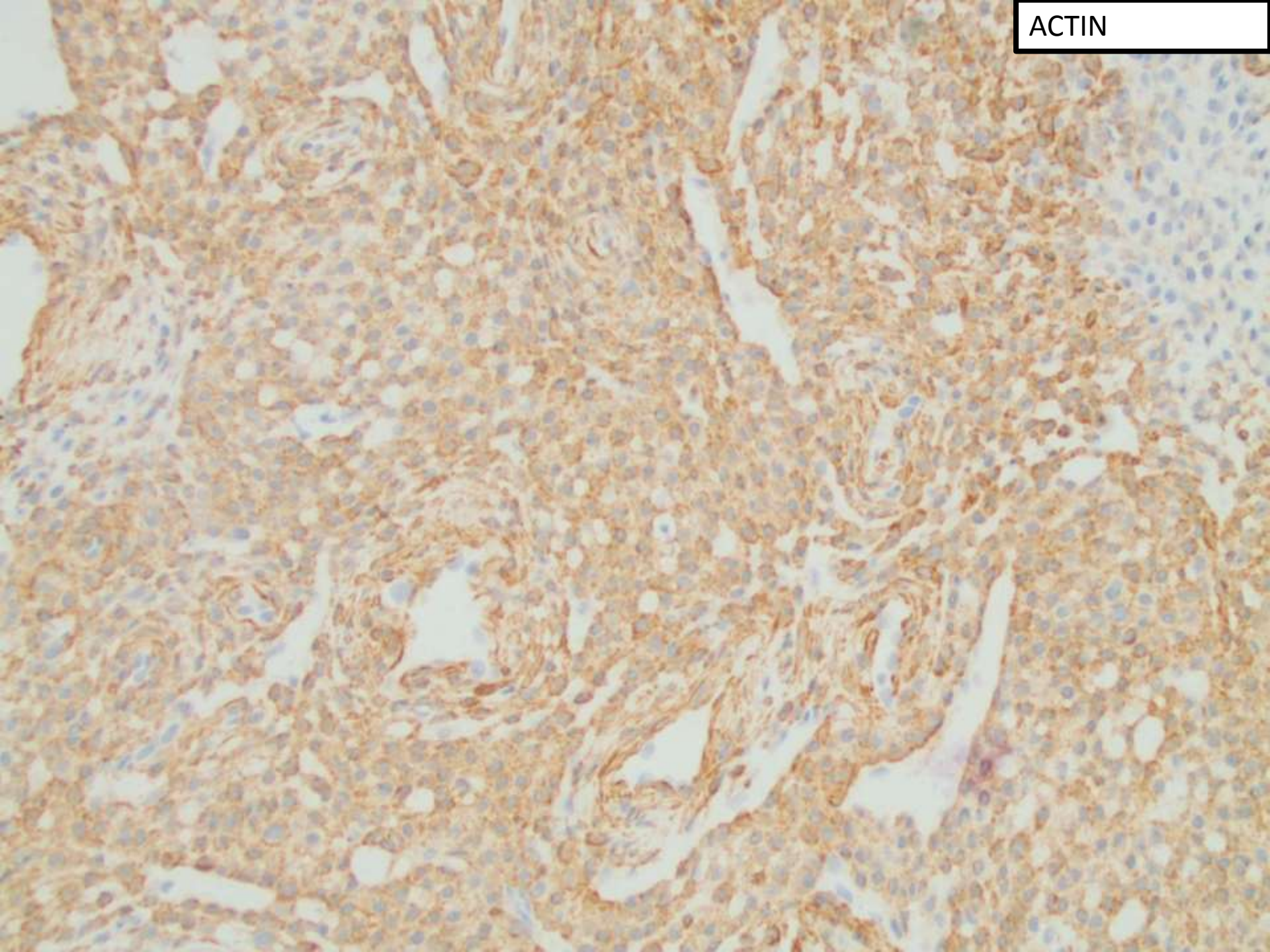






Diagnosis….???




ACTIN

Glomus tumors of Bone

- **Mainly distal phalanx of hands and feet**
- **Rarely other bones: coccyx, ulna**
- **Painful**
- **Lytic lesion with sclerotic margin**

Glomus tumors of distal phalynx

- Three possible scenarios:
  - Soft tissue lesion with compression deformity of bone
  - Soft tissue lesion with extension into bone
  - Primary neoplasm of bone

Immunohistochemistry

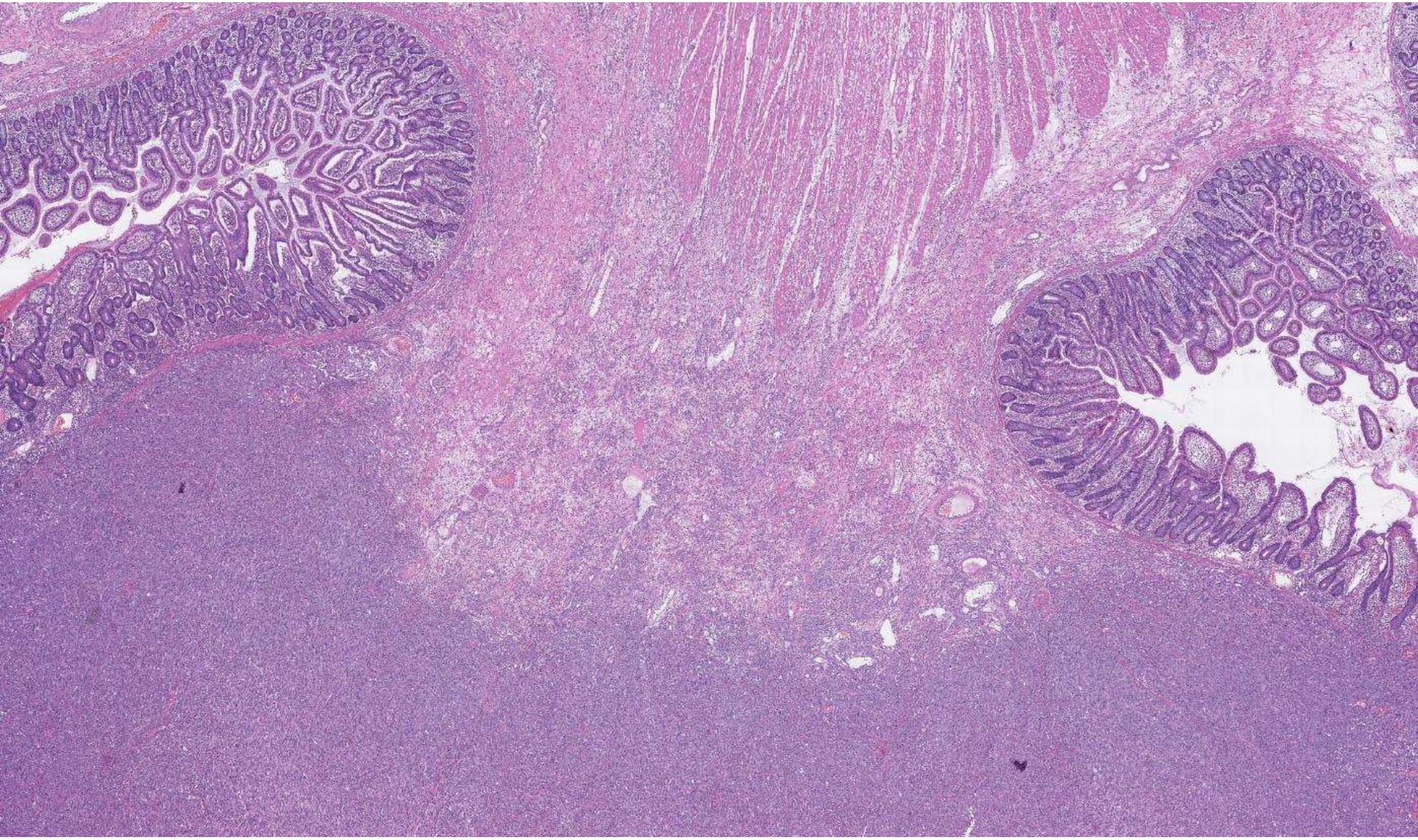
- **Smooth muscle actin**
- **Vimentin**
- **Desmin: variable**
- **Heavy-caldesmon: variable**

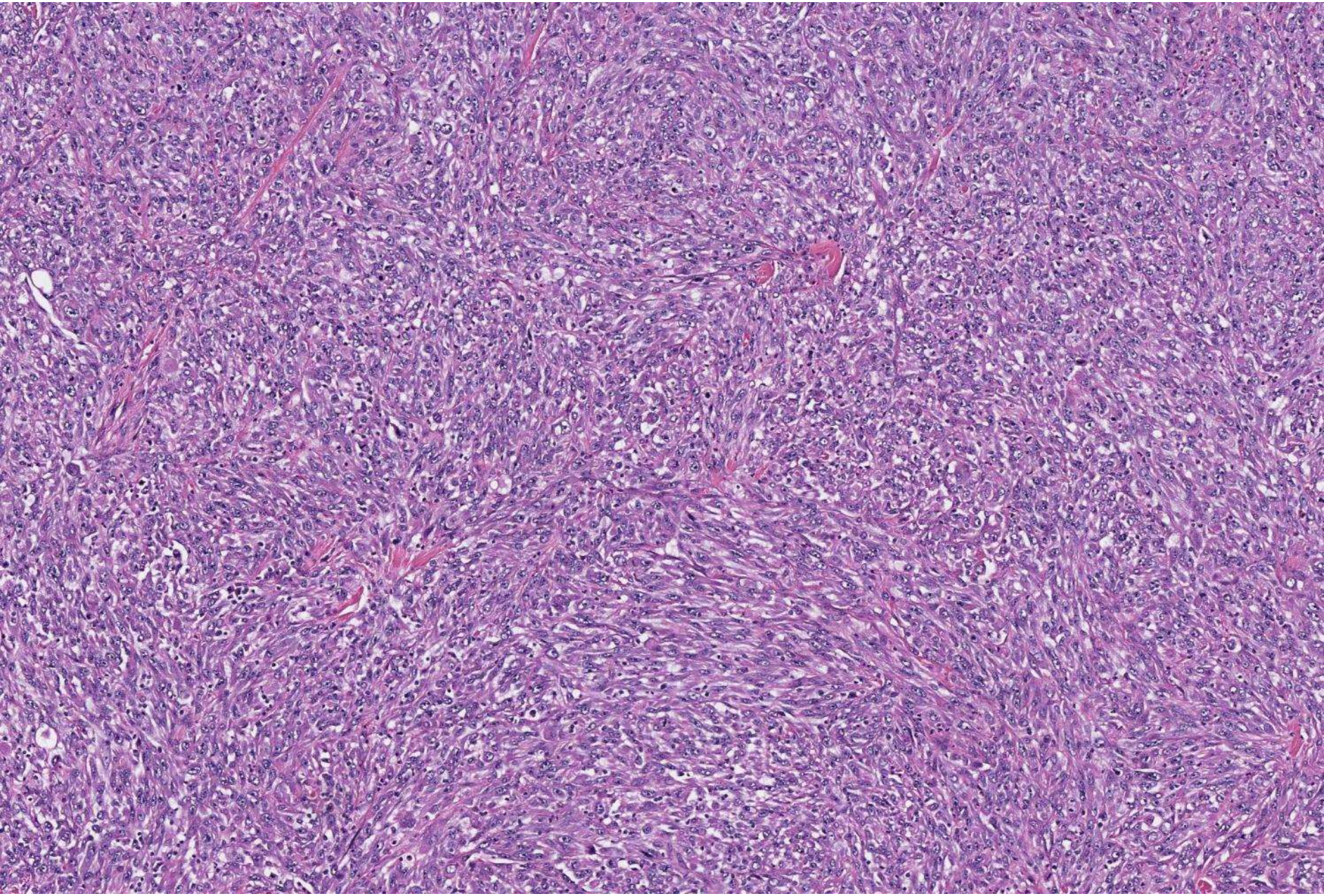
- **Laminin and type IV collagen outline single or groups of cells**

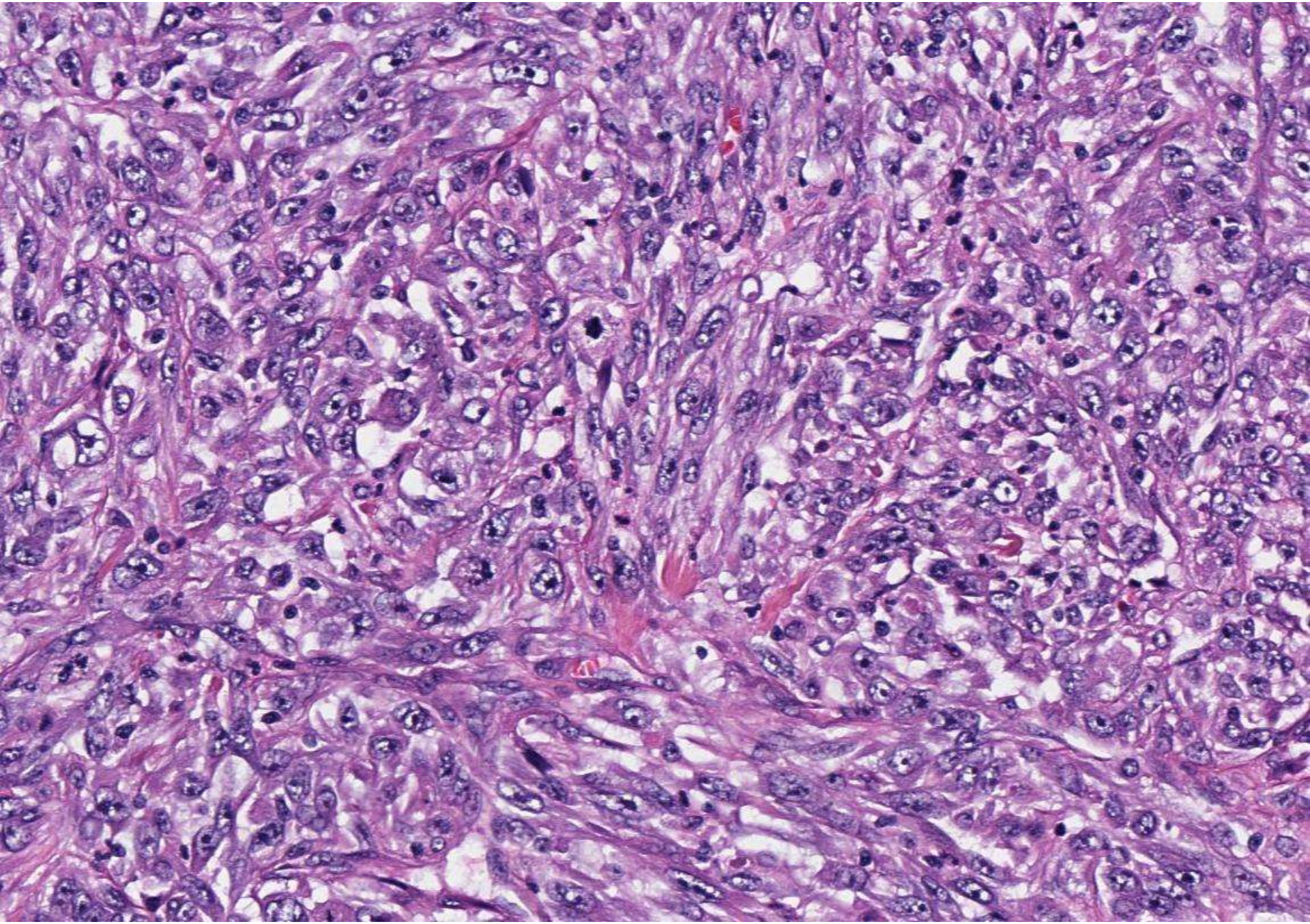
SB 5944

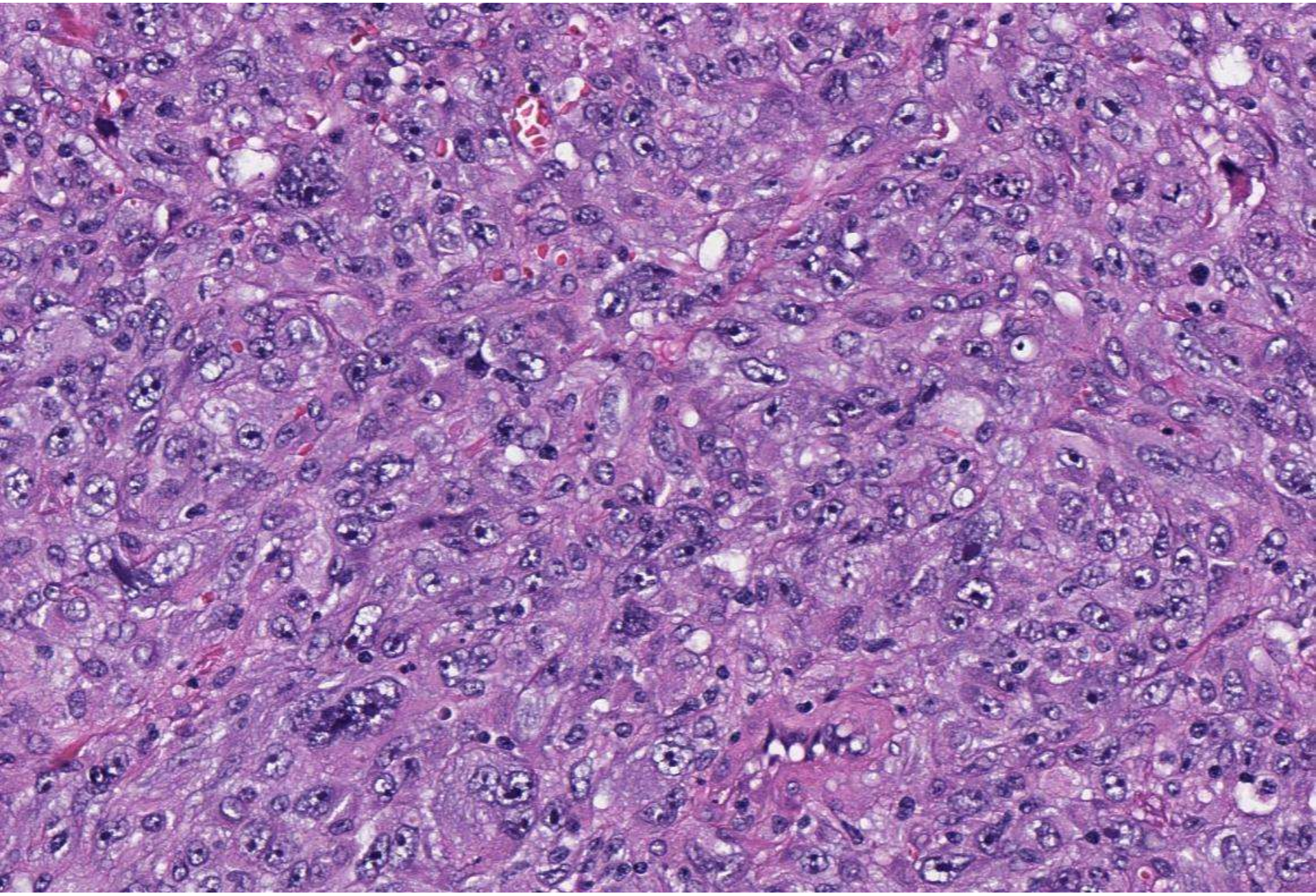
- 80-year-old male presenting with anemia and intussusception. Portion of small bowel resected.

- **Will Rogers; El Camino Hospital**

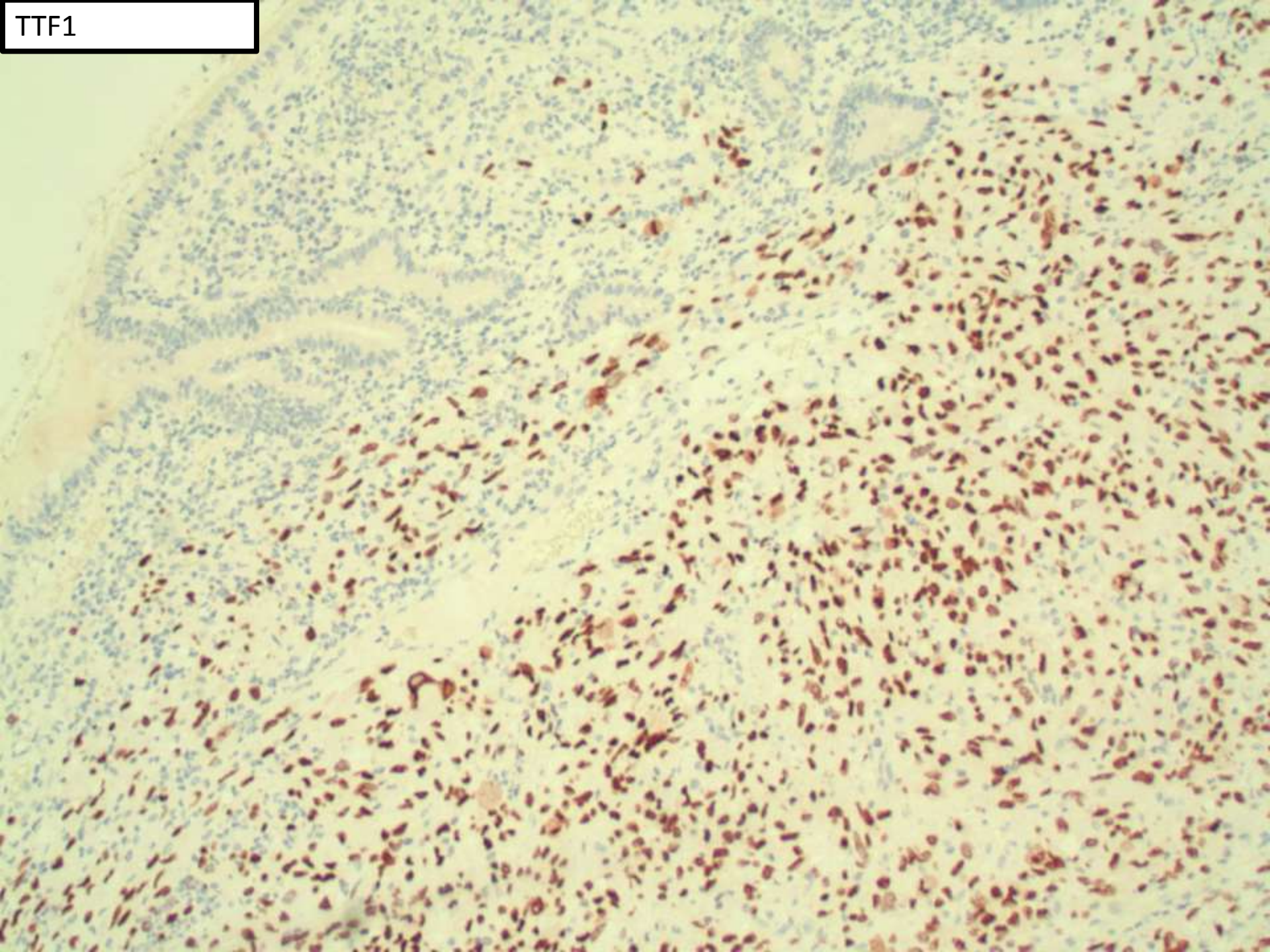








Diagnosis….???


TTF1

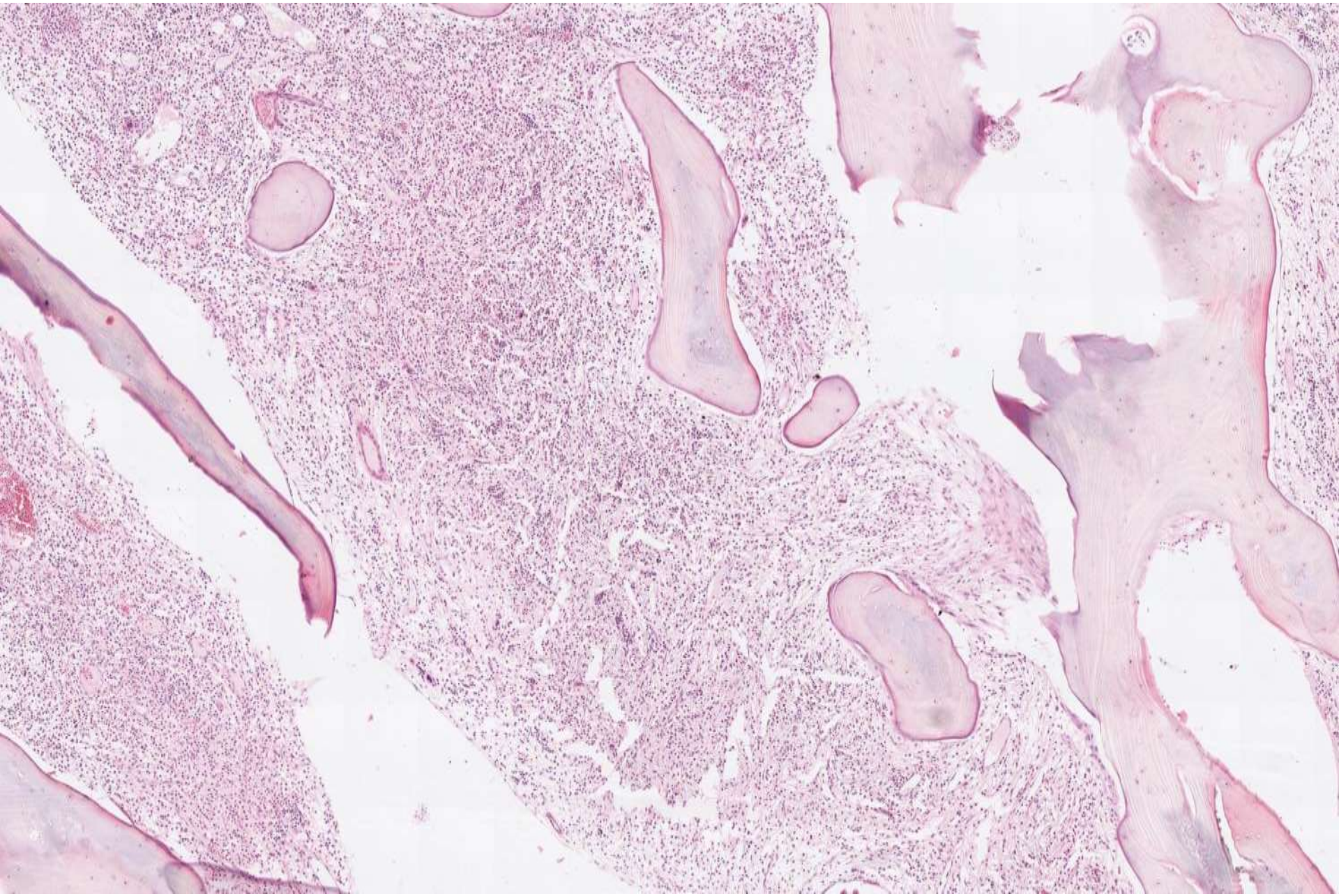
SB 5944

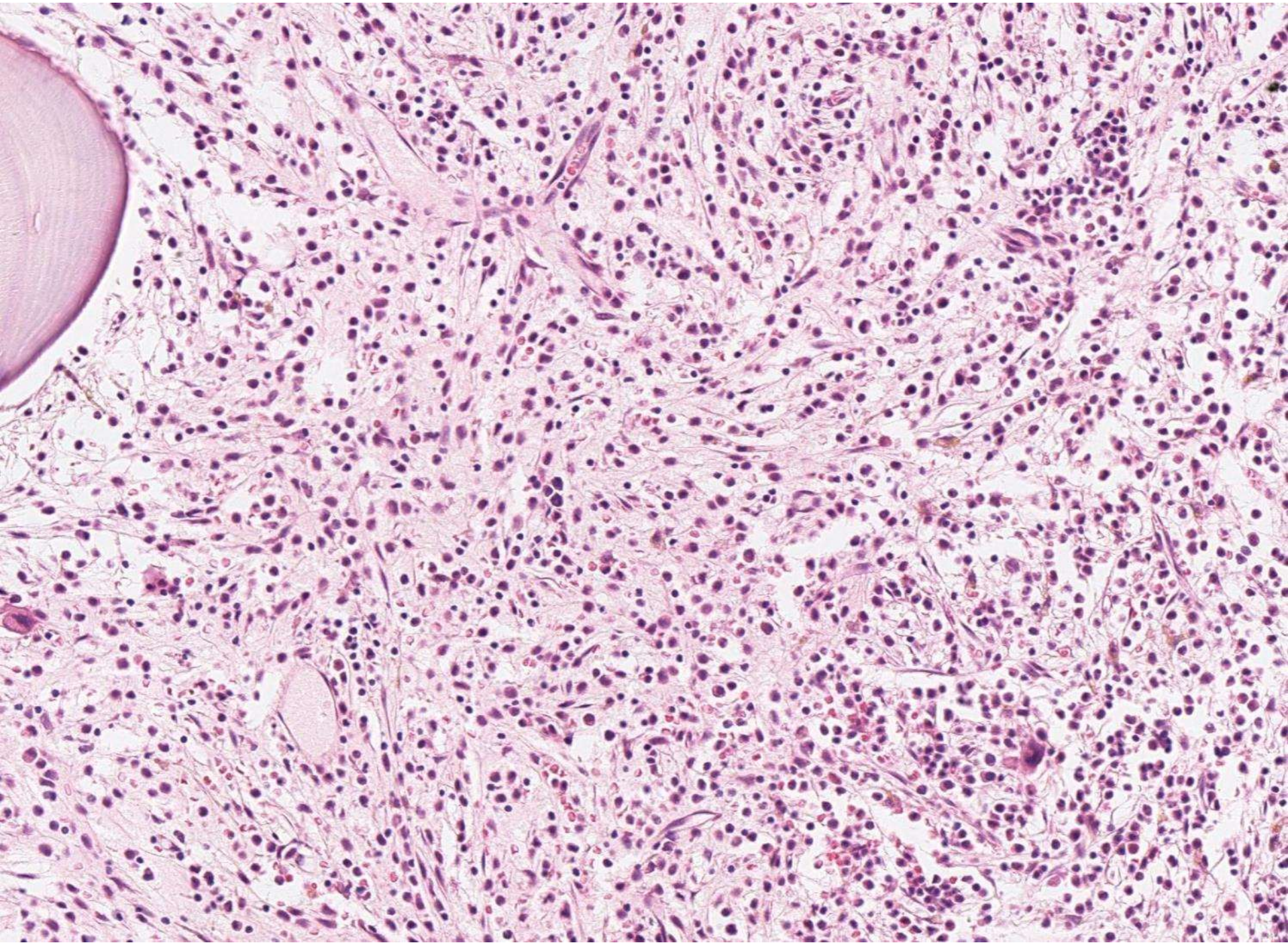
- Poorly differentiated carcinoma metastatic from Lung primary

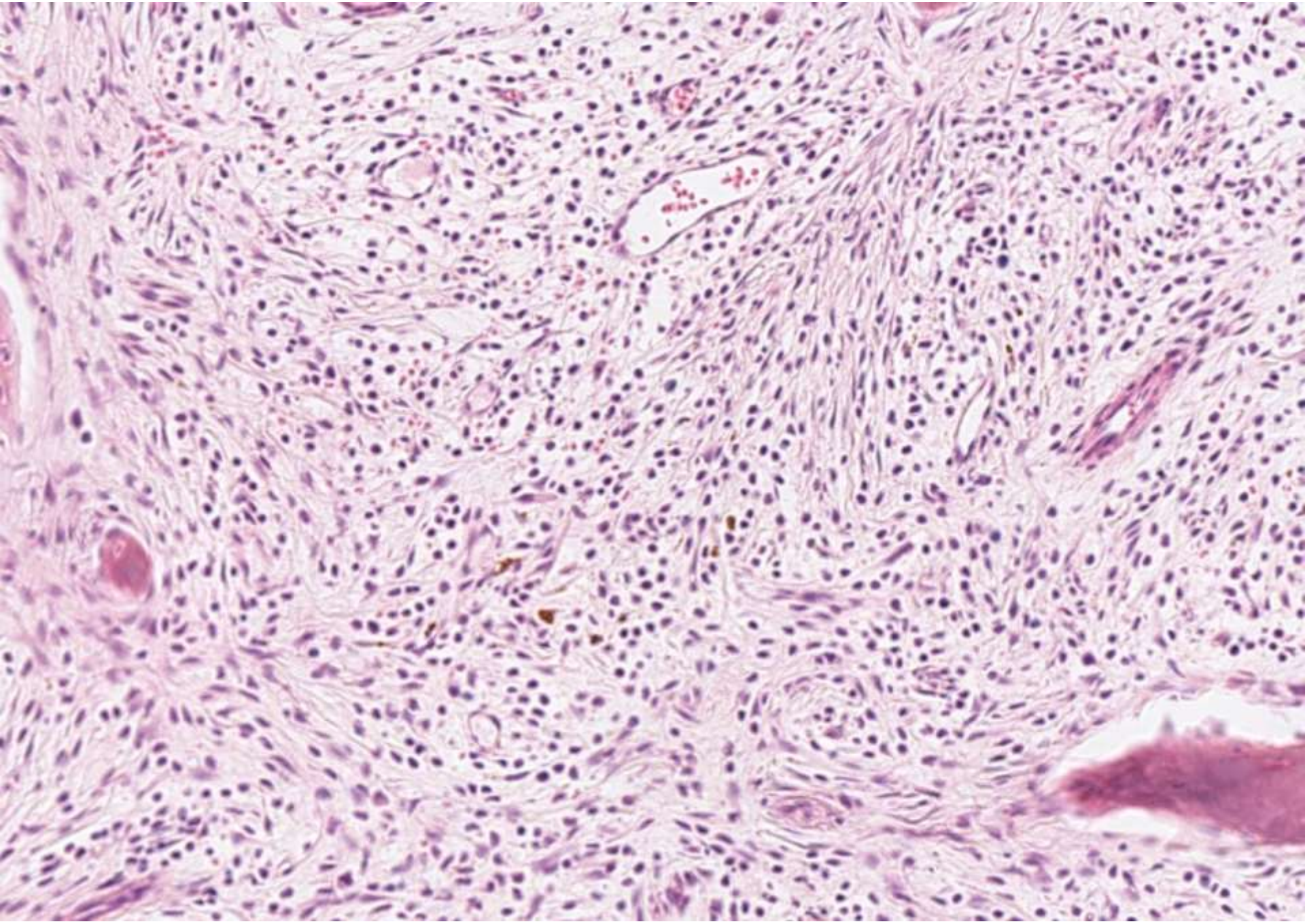
SB 5945

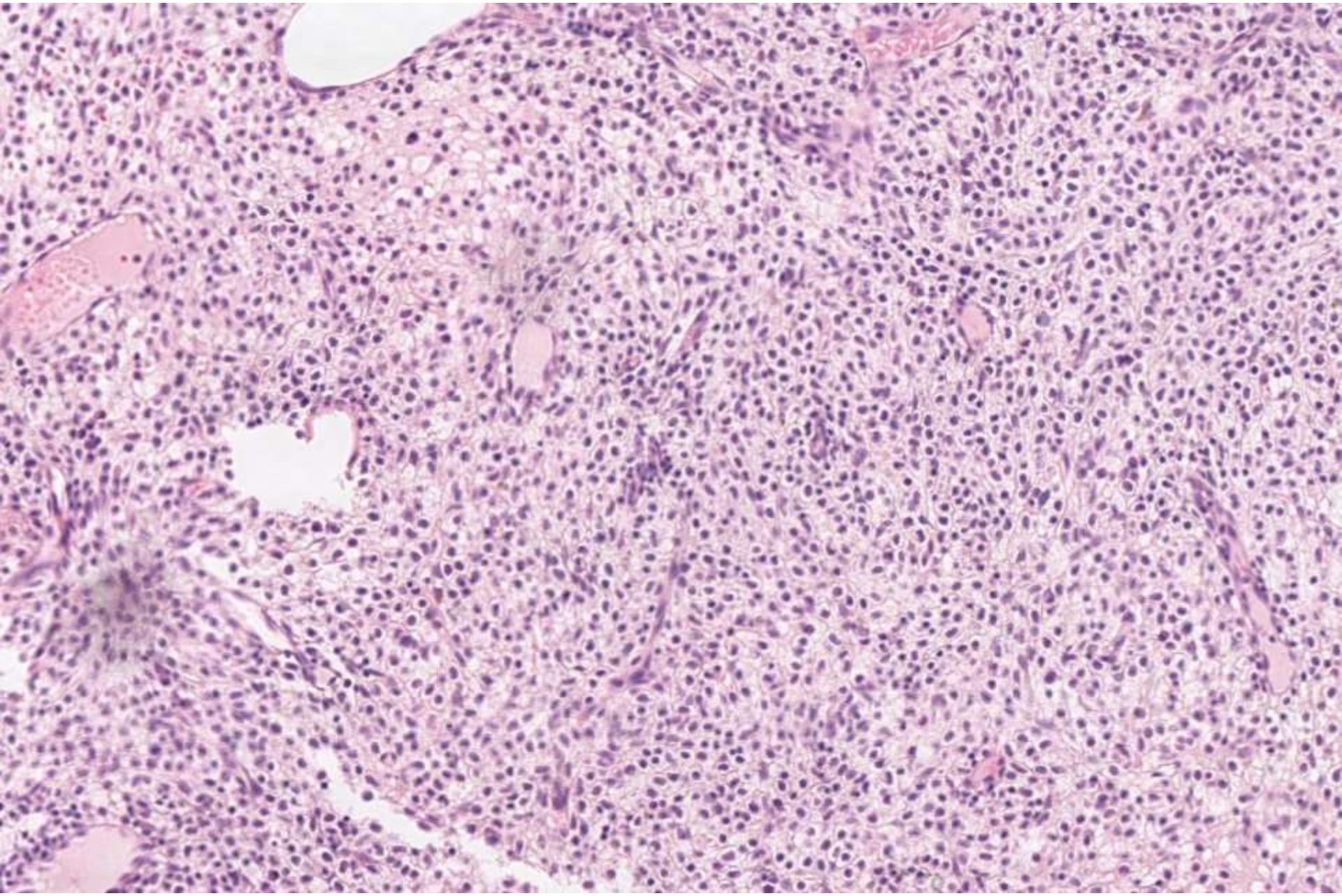
- 64-year-old male with reported osteoarthritis/DJD now with severe (9/10) pain who underwent left total hip arthroplasty. Operative findings included delamination of articular cartilage off the superior dome (gross photograph unavailable but necrosis not grossly observed).

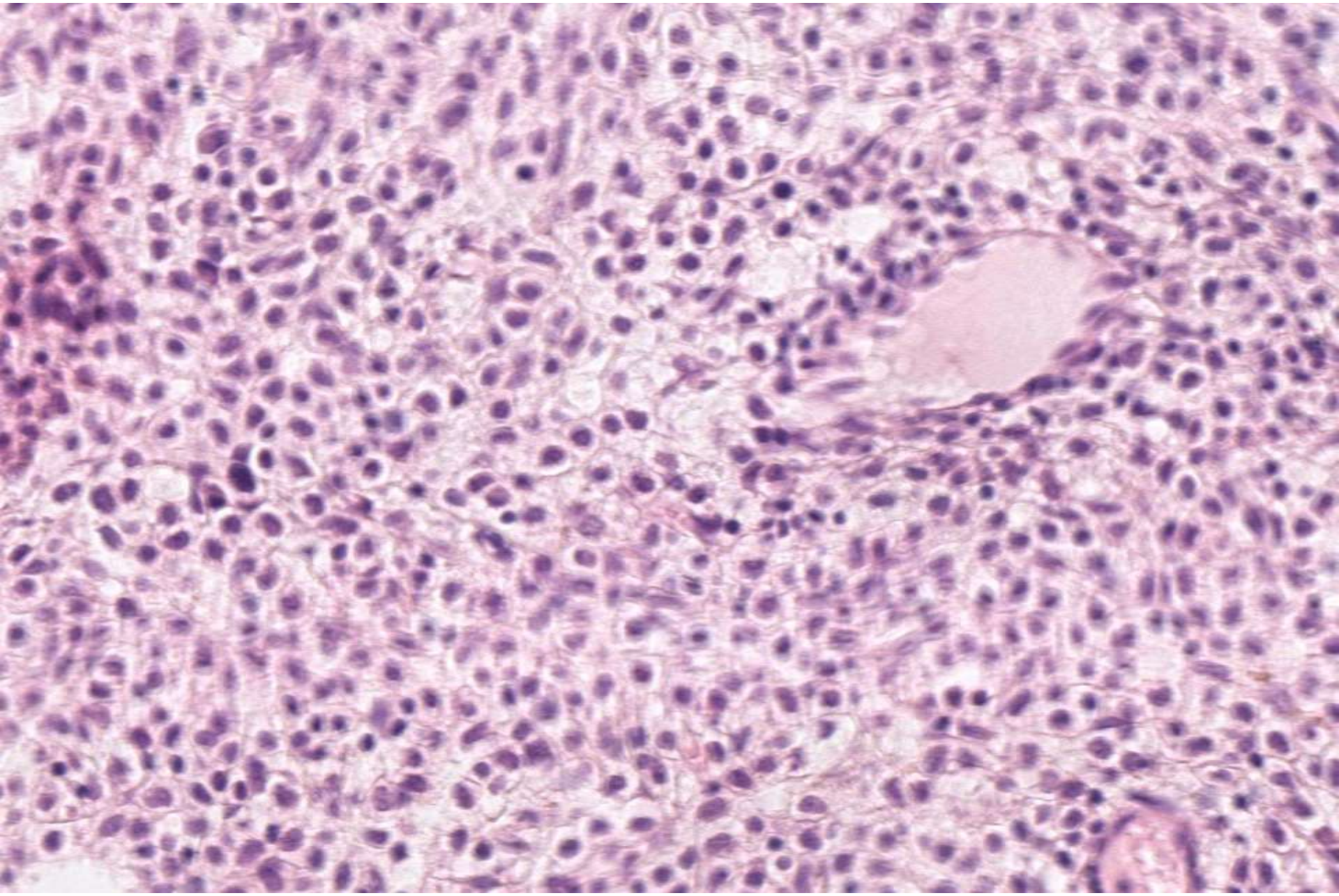
- **Ryan Johnson/Yaso Natkunam; Stanford**

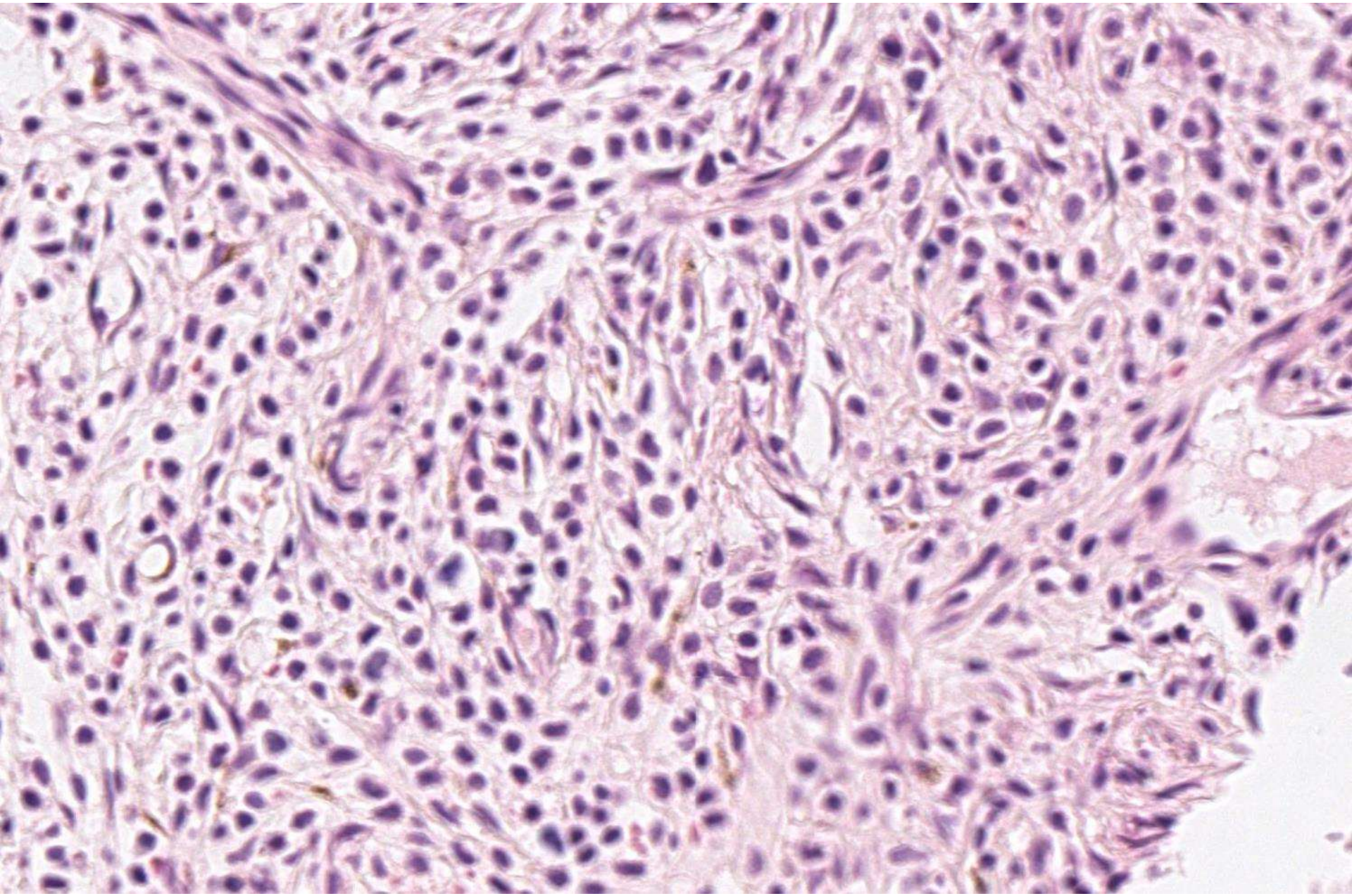










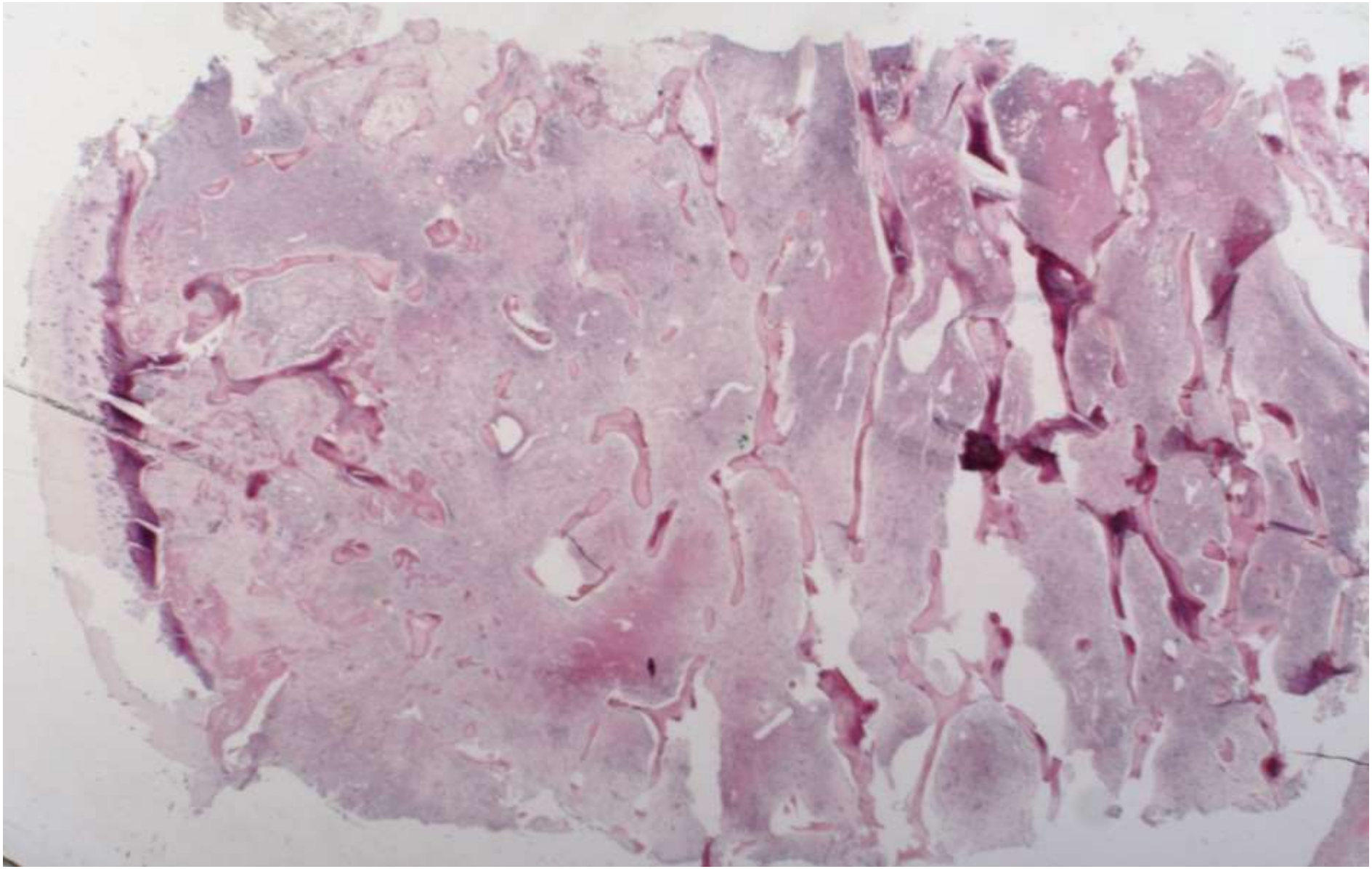


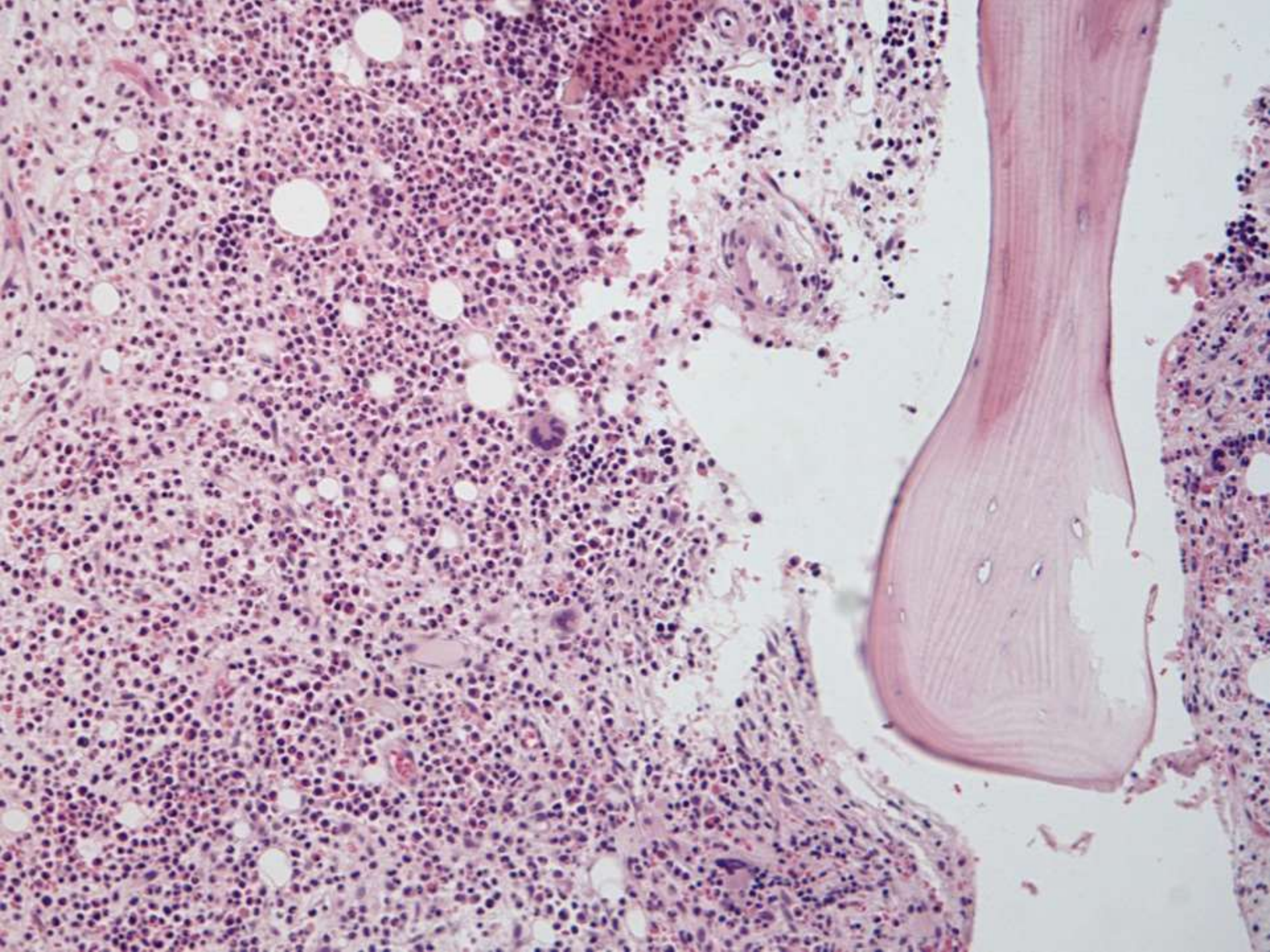
Diagnosis….???

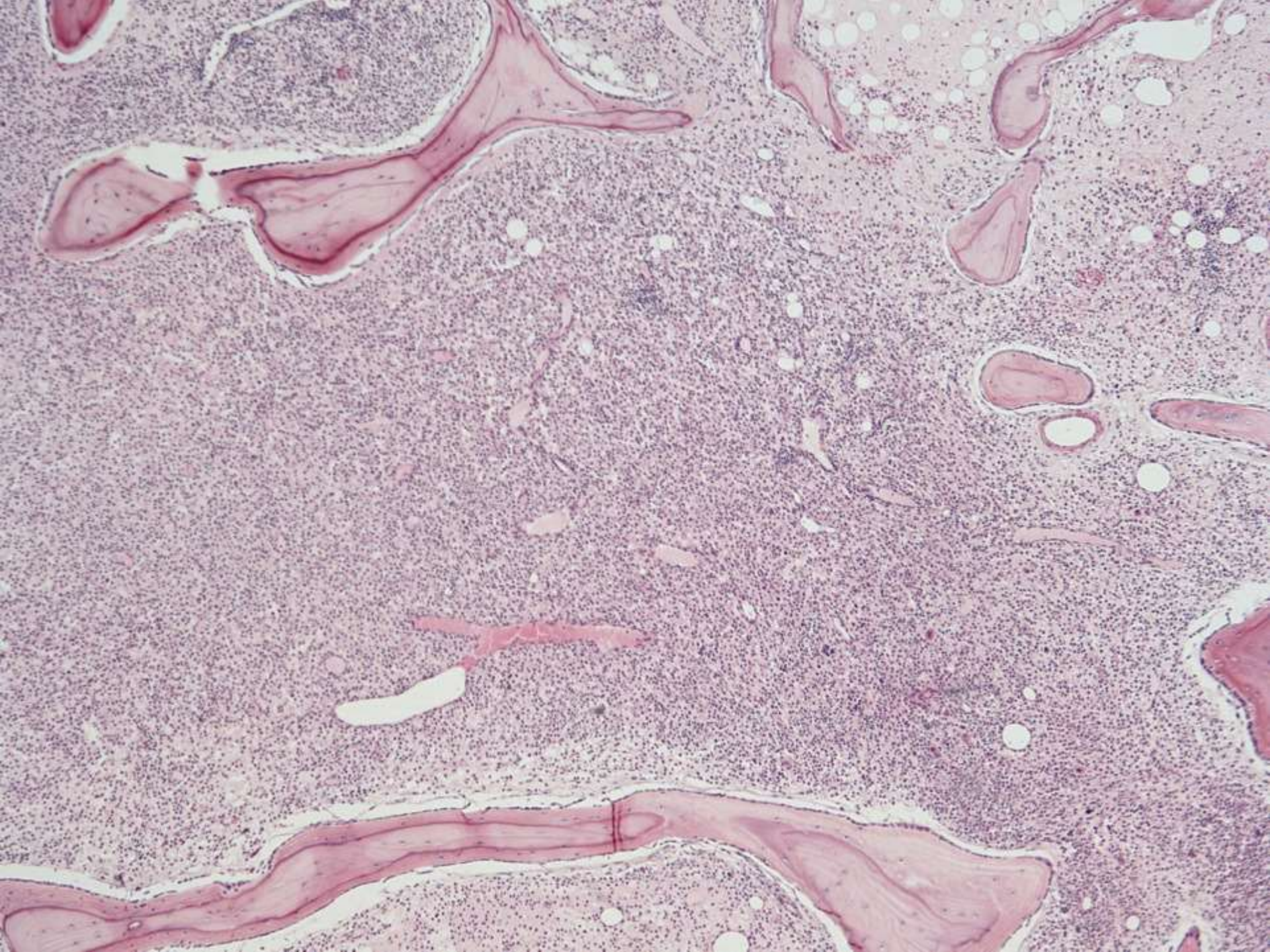
Case history

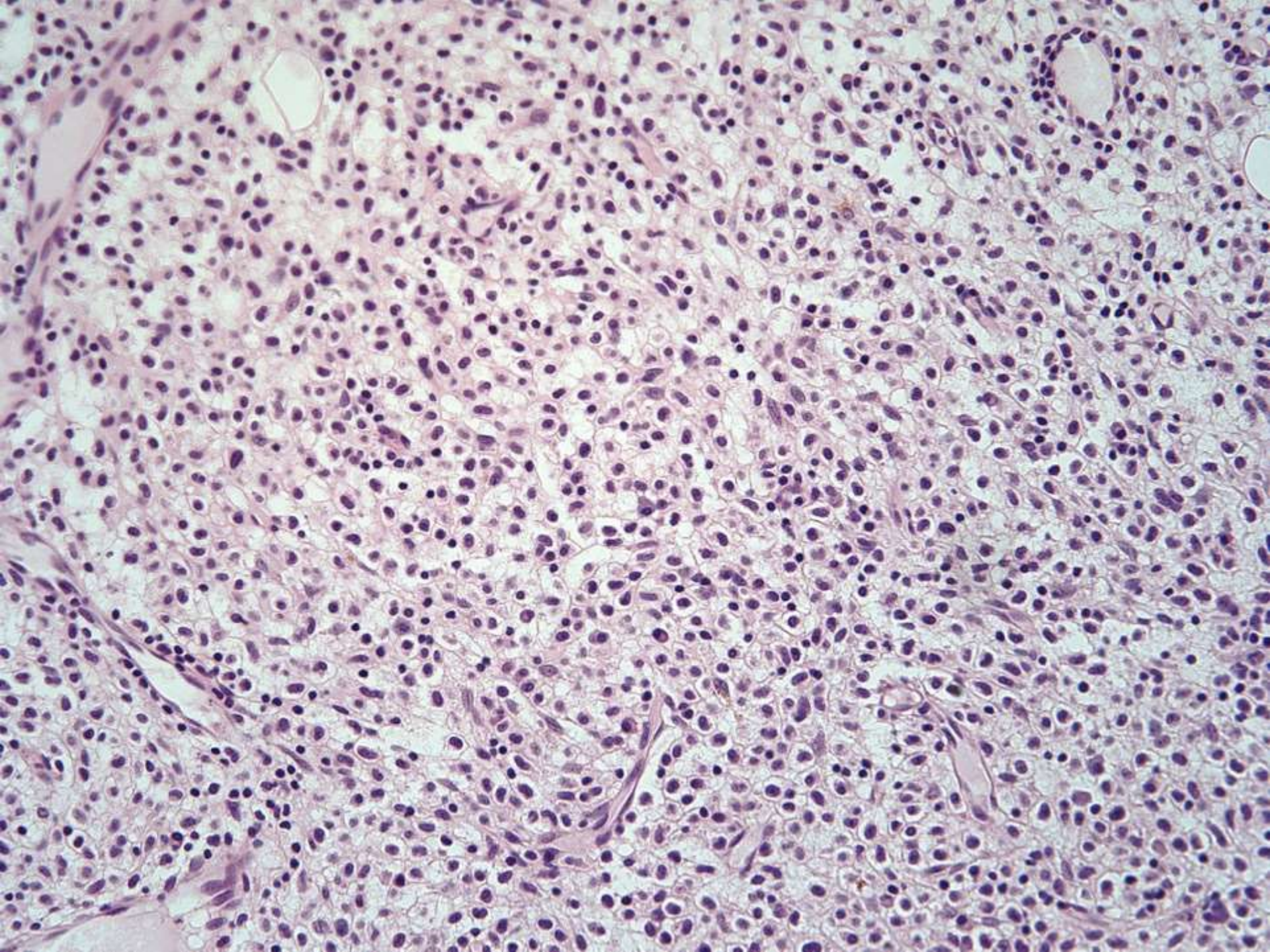
- 64 year old male with reported osteoarthritis/degenerative joint disease now with severe (9/10) pain who underwent left total hip arthroplasty. Operative findings included delamination of articular cartilage off the superior dome. (Gross photograph unavailable, but necrosis not grossly observed)

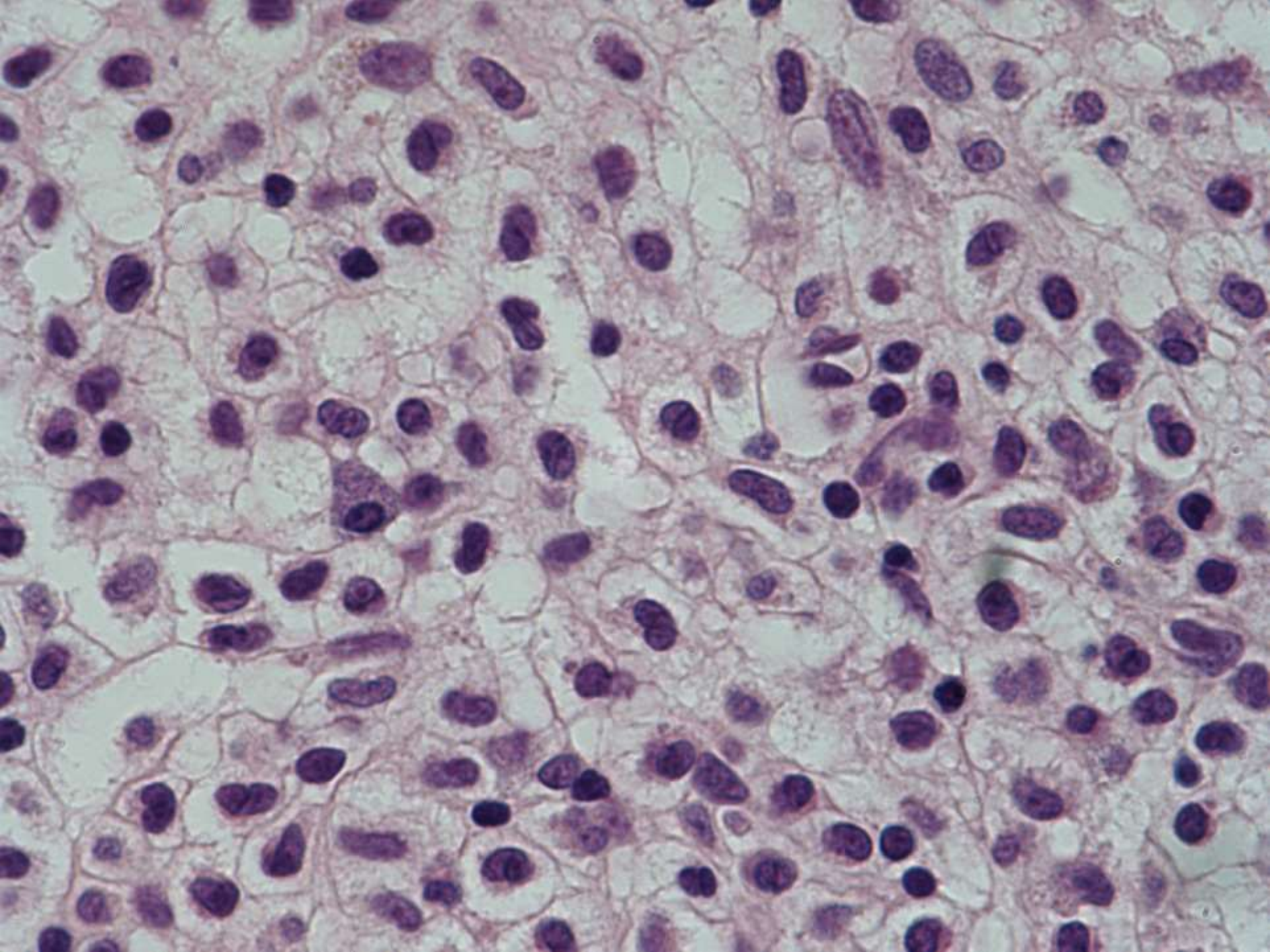
- WBC 5.9, H/H 7.6/22.5, PLT 222
  - No differential available

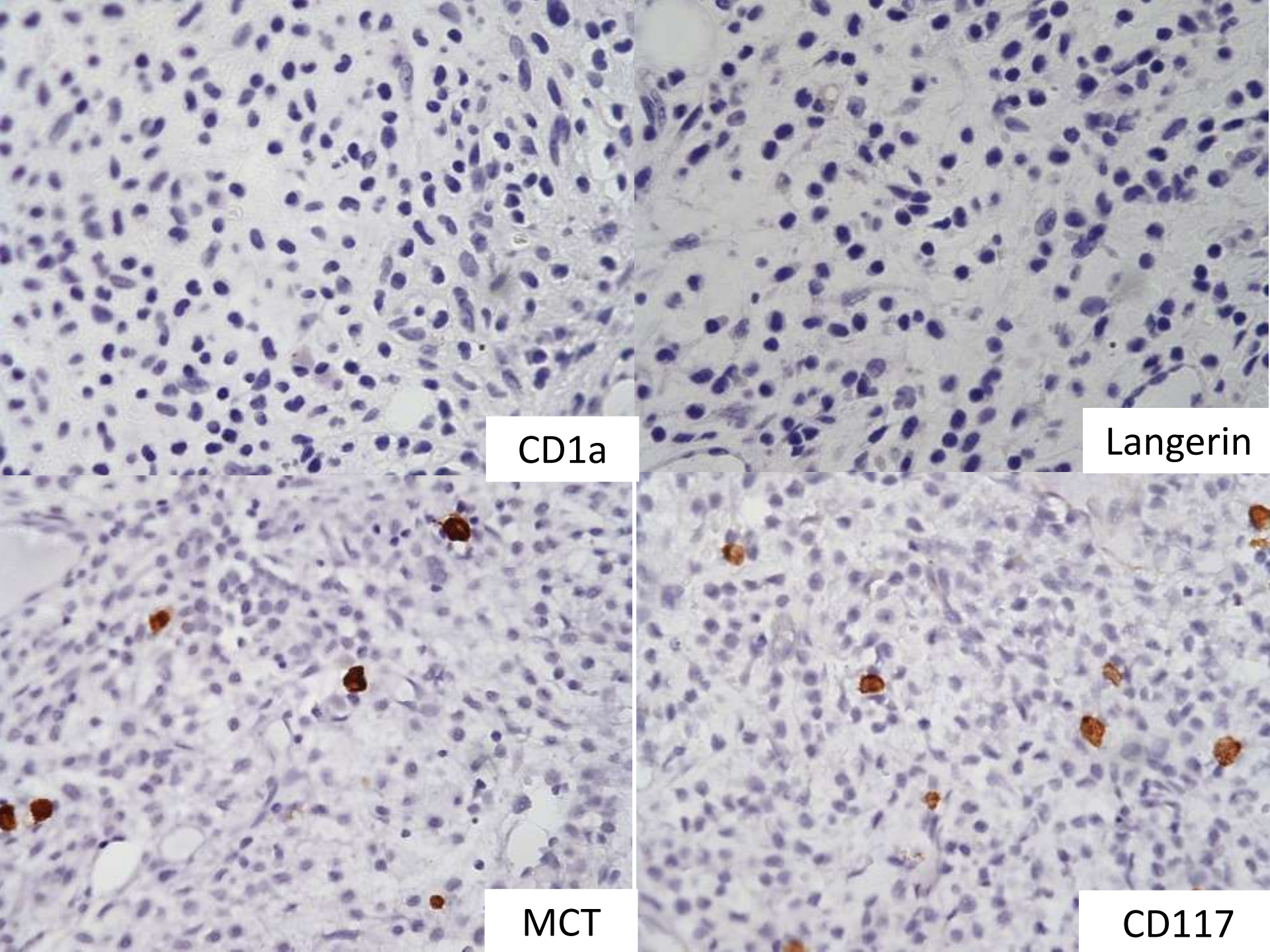


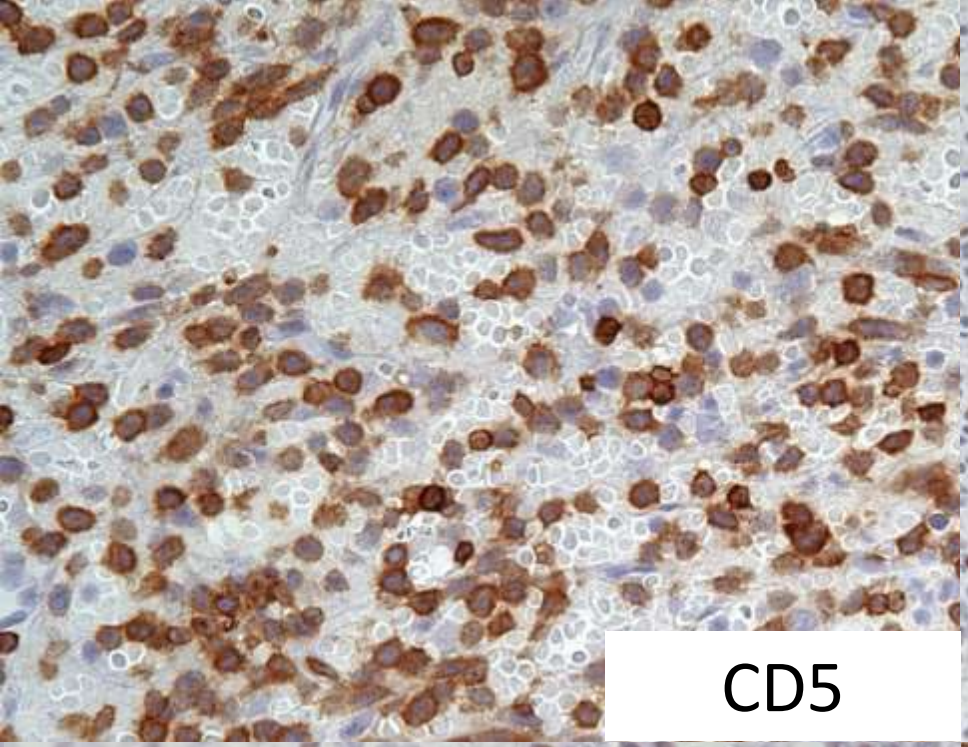


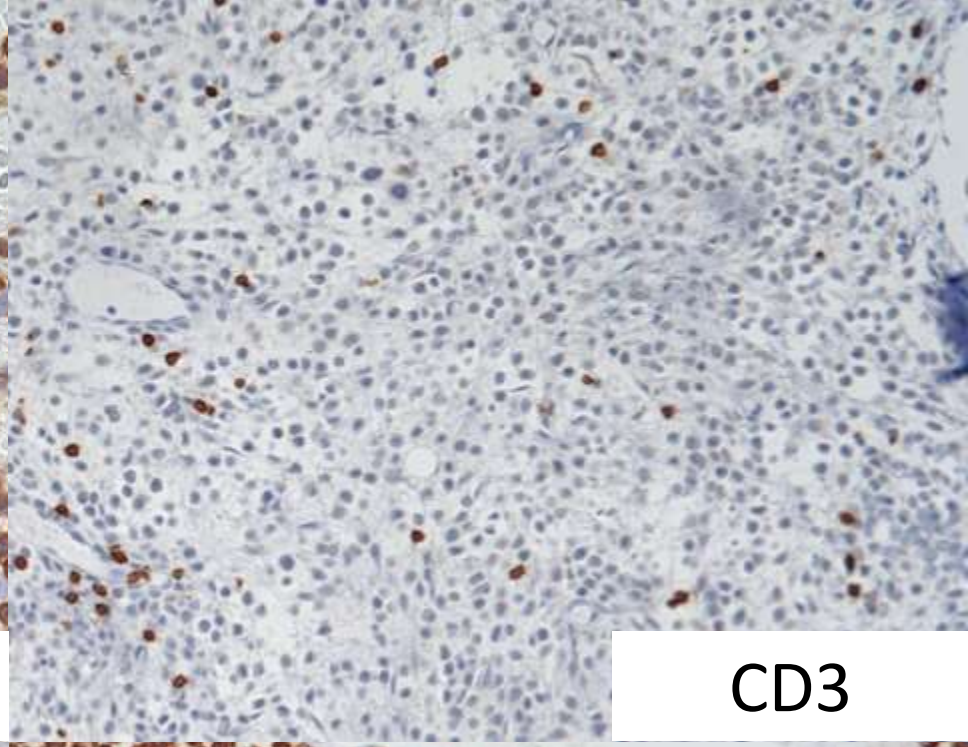







CD1a
Langerin
MCT
CD117


CD5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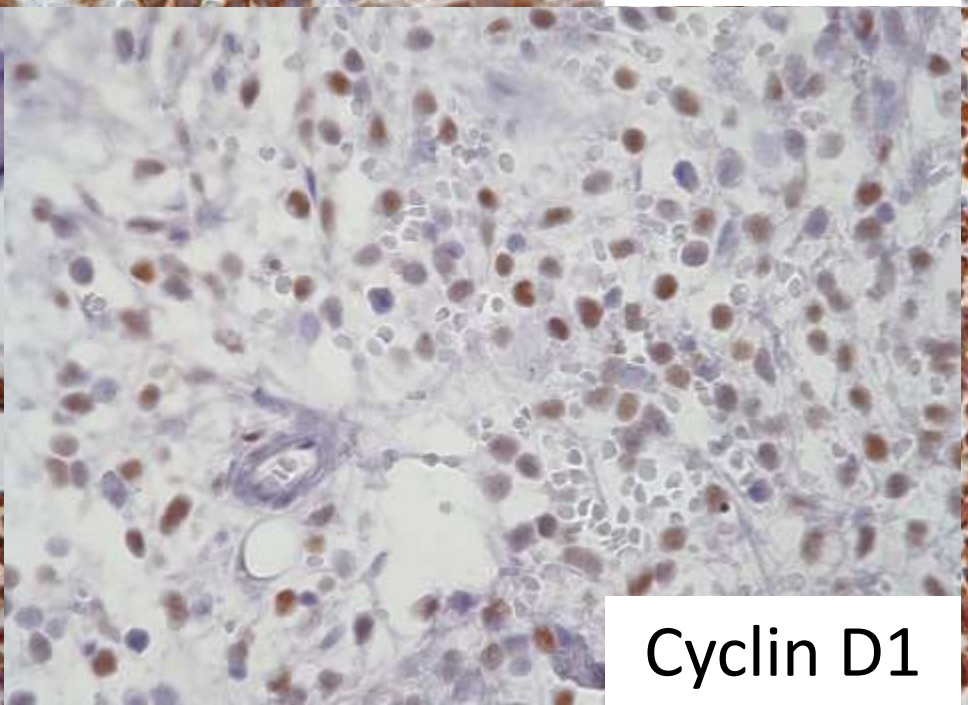
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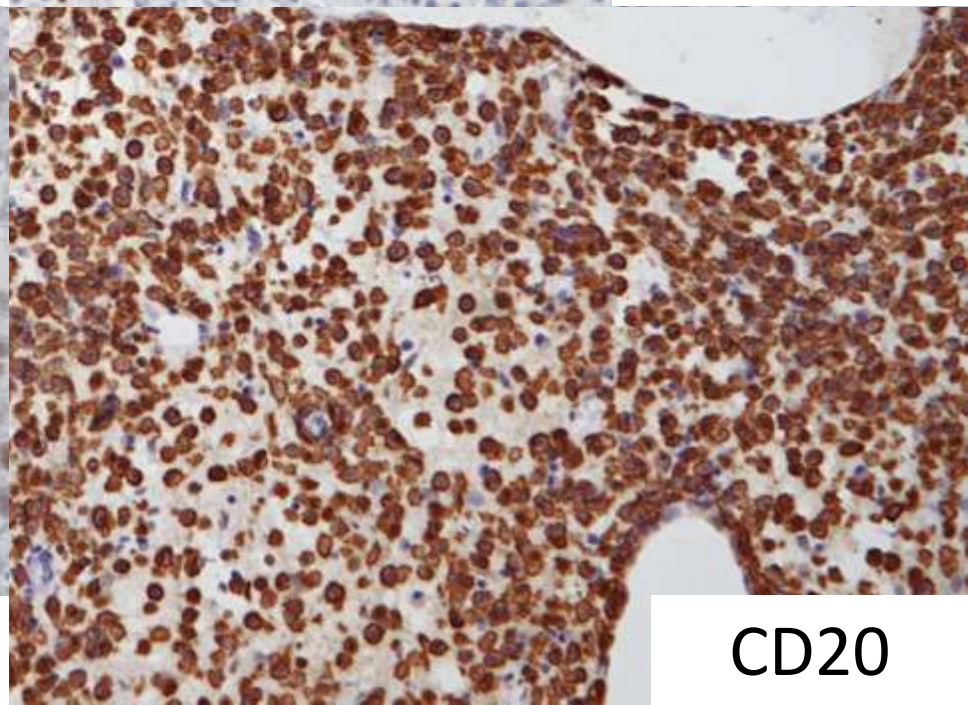
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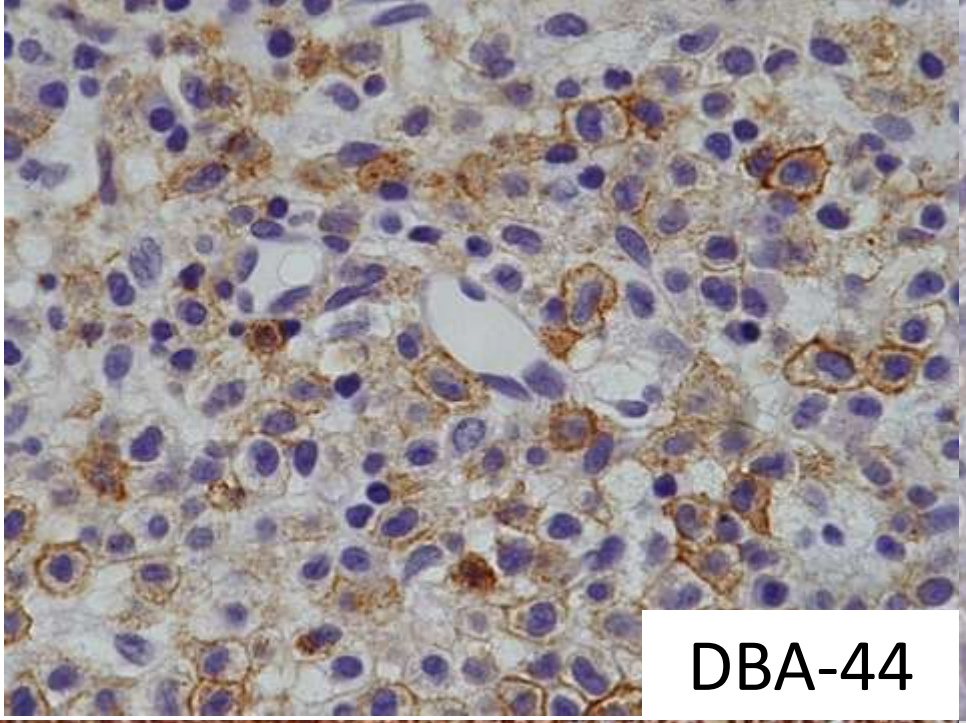
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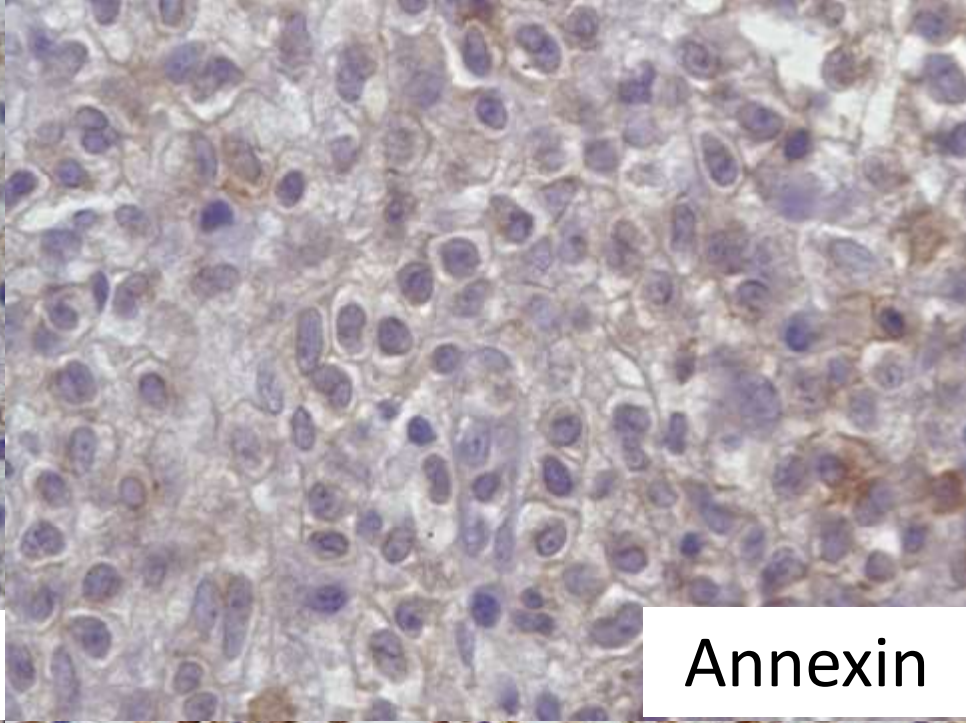
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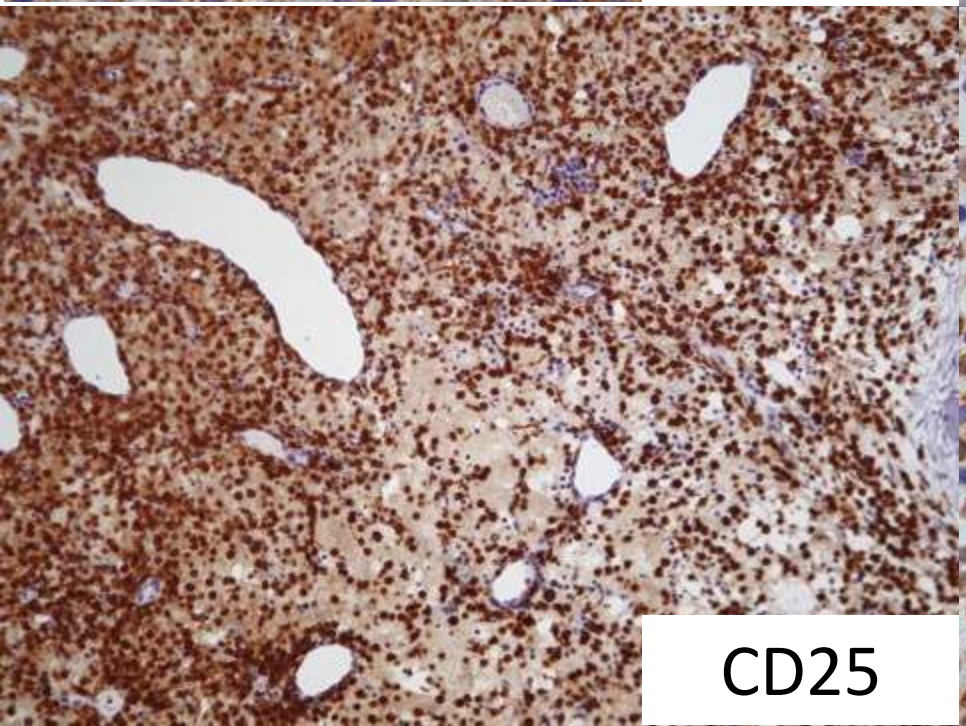
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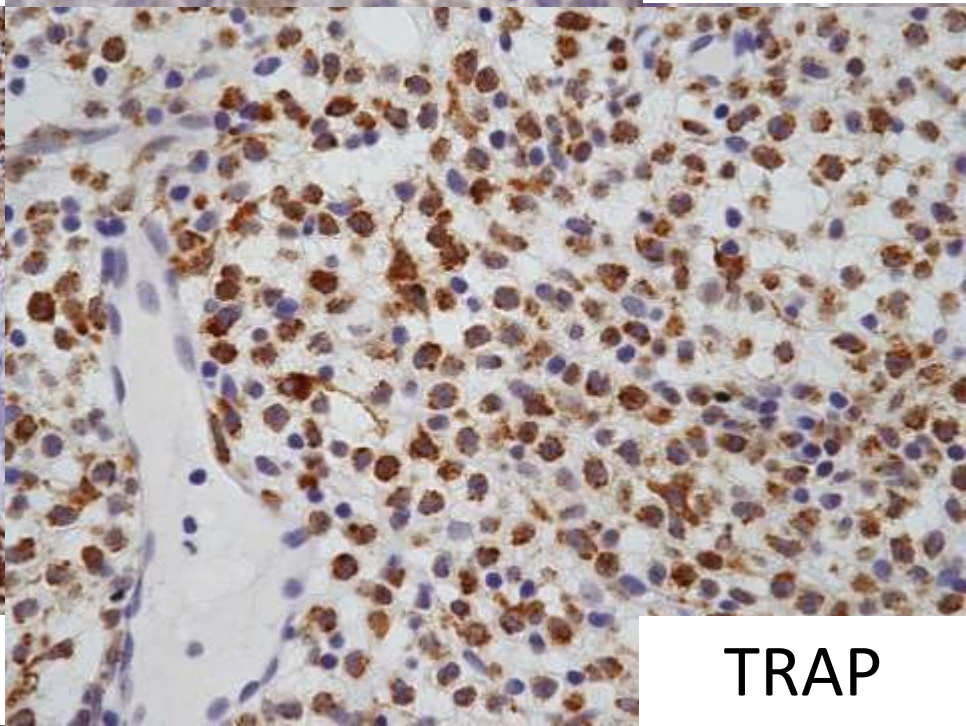

TRAP

Diagnosis:

- Hairy Cell Leukemia

- Typical symptoms of HCL
  - RUQ pain (splenomegaly), fatigue, bleeding, infections (cytopenias)
  - Lymphadenopathy less common (20%)
  - Bone mass lesions *as presentation* are rare (4-5 case reports in literature)
  - Cytopenias (monocytopenia) with variably involved (sometimes hypocellular) marrow

Skeletal complications of HCL

- Relatively rare complication of HCL
- Study of 267 patients with HCL – 8 had skeletal mass lesions (3% of cases*)
    - 7/8 had a prior dx with therapy
    - Most involved femoral head (other sites included ribs, vertebrae)
    - Lytic on imaging
    - Treated with local radiotherapy (+/- systemic chemo)

Lembersky et al *JCO* (1988)

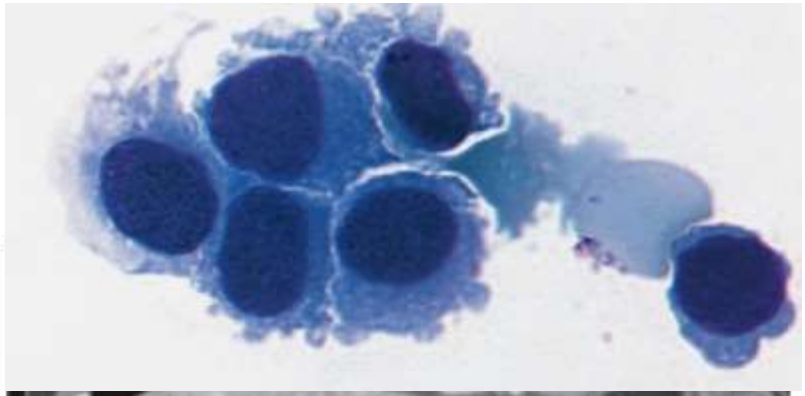
Lymphadenopathy in HCL

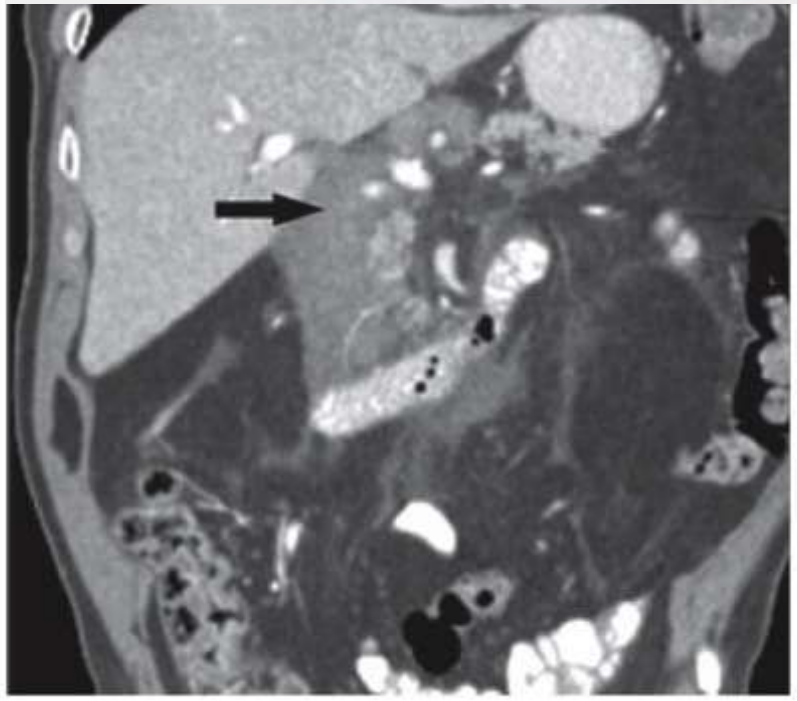
- Another uncommon complication/presentation of HCL
  - Usually associated with relapse in refractory patients
    - (n=88) 17% of patients at diagnosis, 56% of patients at relapse*
    - Overwhelmingly associated with bone marrow (leukemic) involvement
    - Generally associated with treatment failure
      - Prior to cladribine and Rituxan

Mercieca, *Leuk Lymphoma*. 1994;14 Suppl 1:79-83.

Soft tissue mass presentation

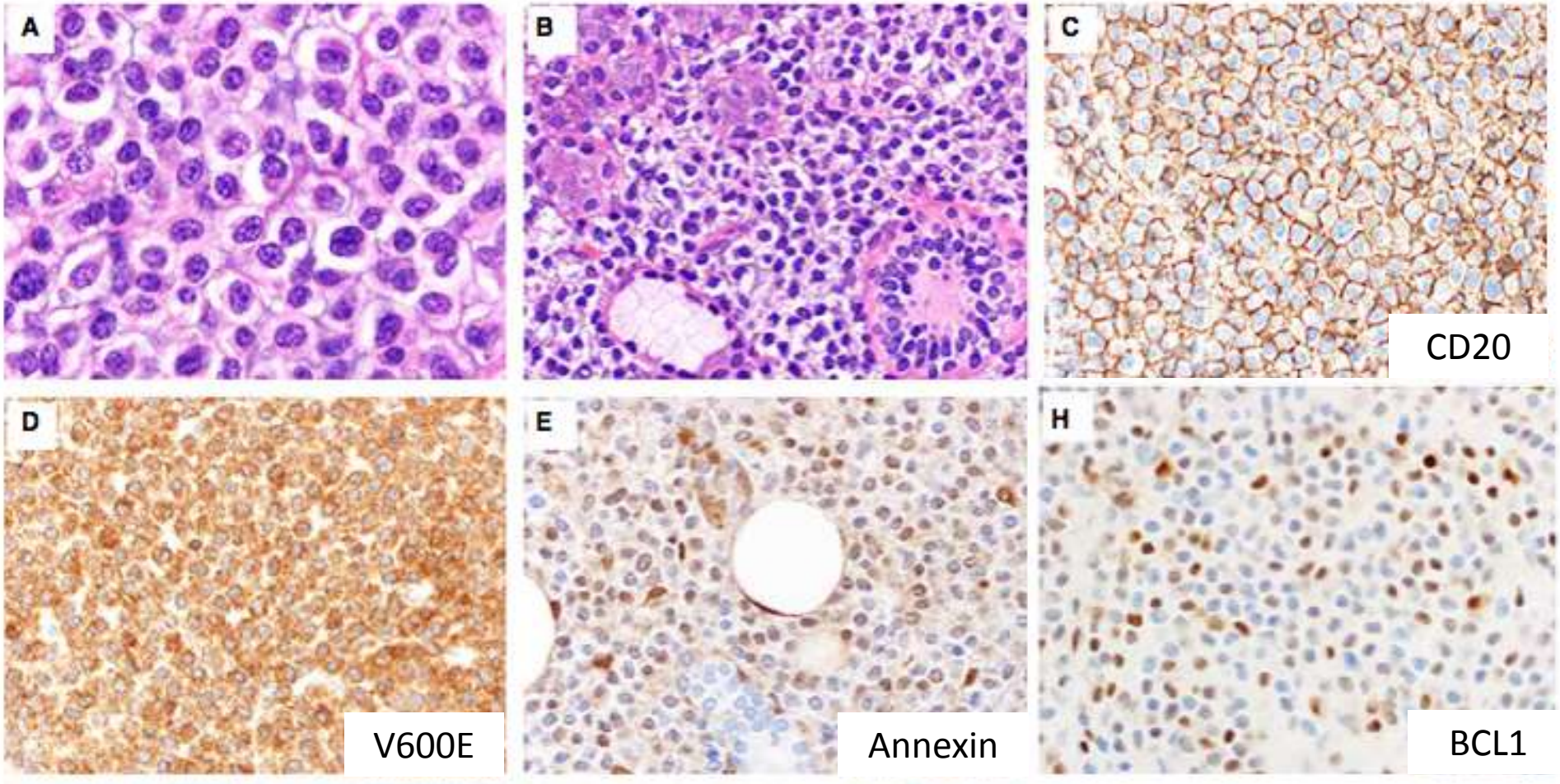
- ‘hairy cell lymphoma’
- Even more rare of a complication (case reports)
- Variety of sites, pancreatic, brain, skin, bone, joint, salivary gland
- Patients often go un(der)diagnosed
  - May lack cytopenias, circulating abnormal lymphocytes, splenomegaly





Subhawong et al Acta Cytologica 2012;56:463–466

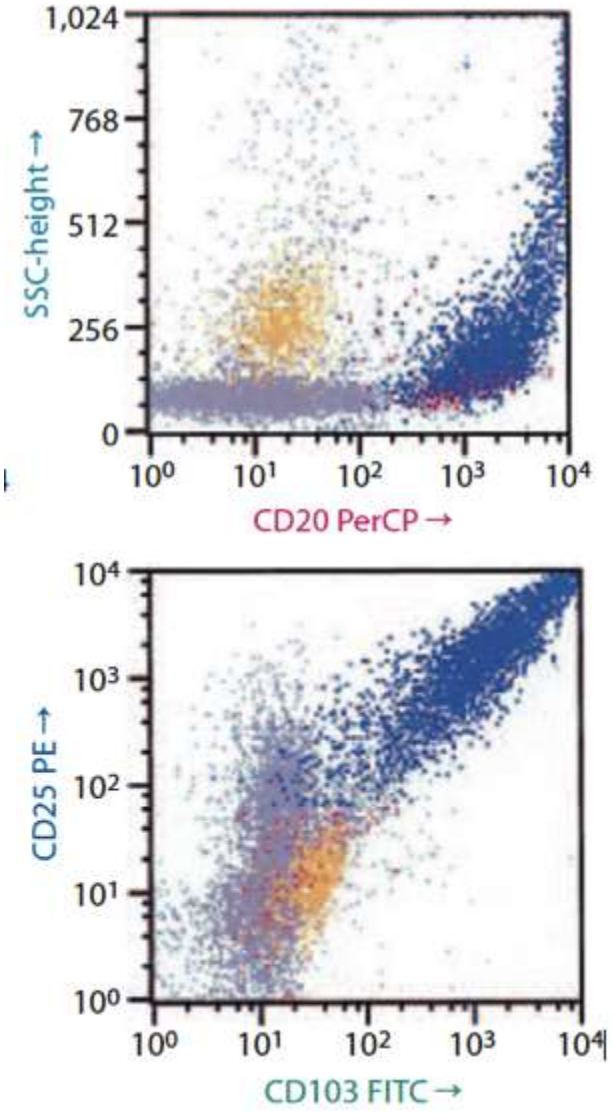
Soft tissue mass presentation



Liu et al, Histopathology 2015 Mar 21.
[Epub ahead of print]

Flow cytometric/immunohistochemistry review

- Bright CD20 and CD22
- Higher SSC/FSC (monocyte gate)
- Expression of CD25, CD103, CD123, CD11c
- Expression of TRAP*, CD25, cyclinD1, CD43, DBA.44, Tbet, Annexin, BRAF V600E

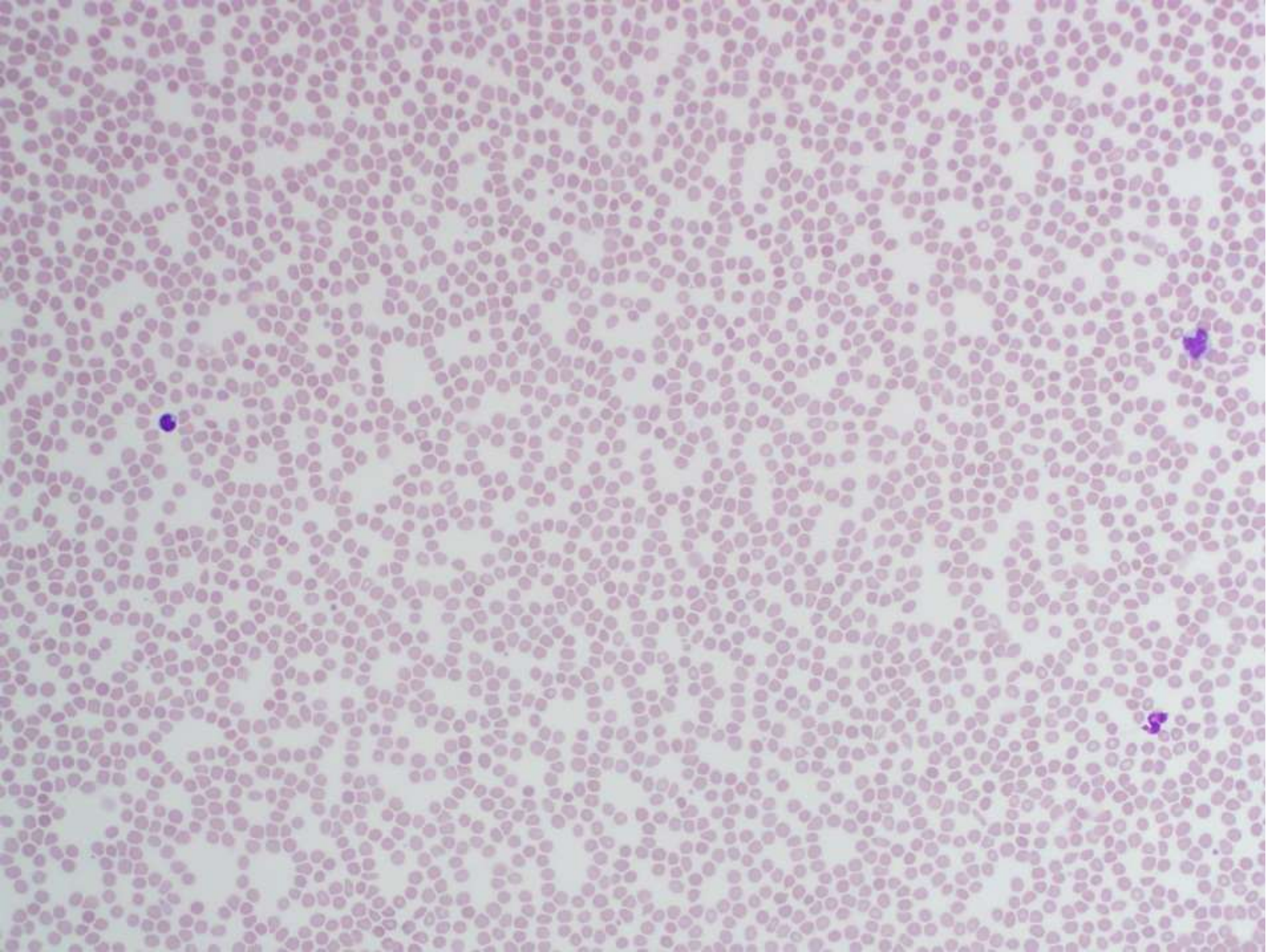


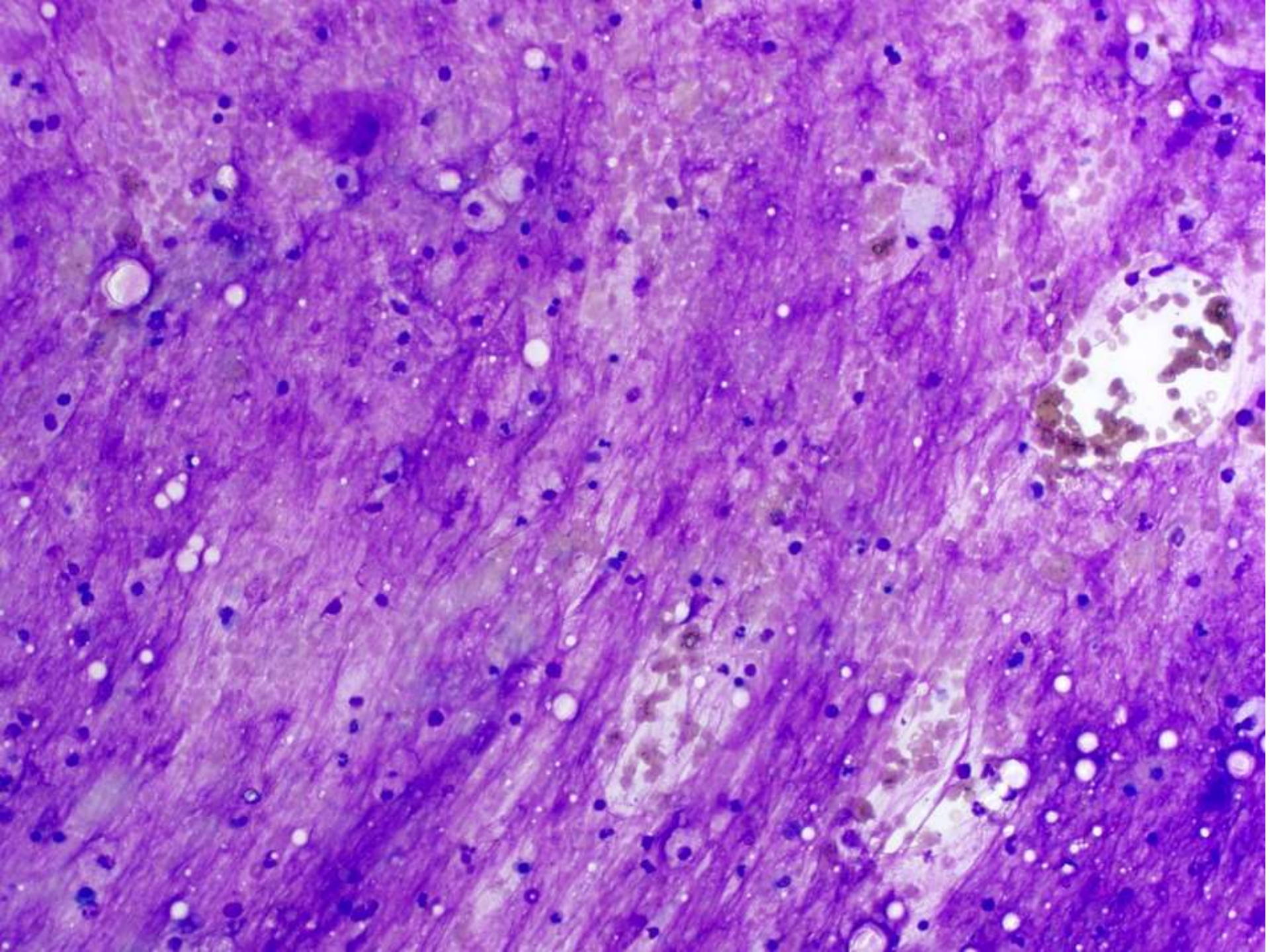
Subhawong et al Acta Cytologica 2012;56:463–466

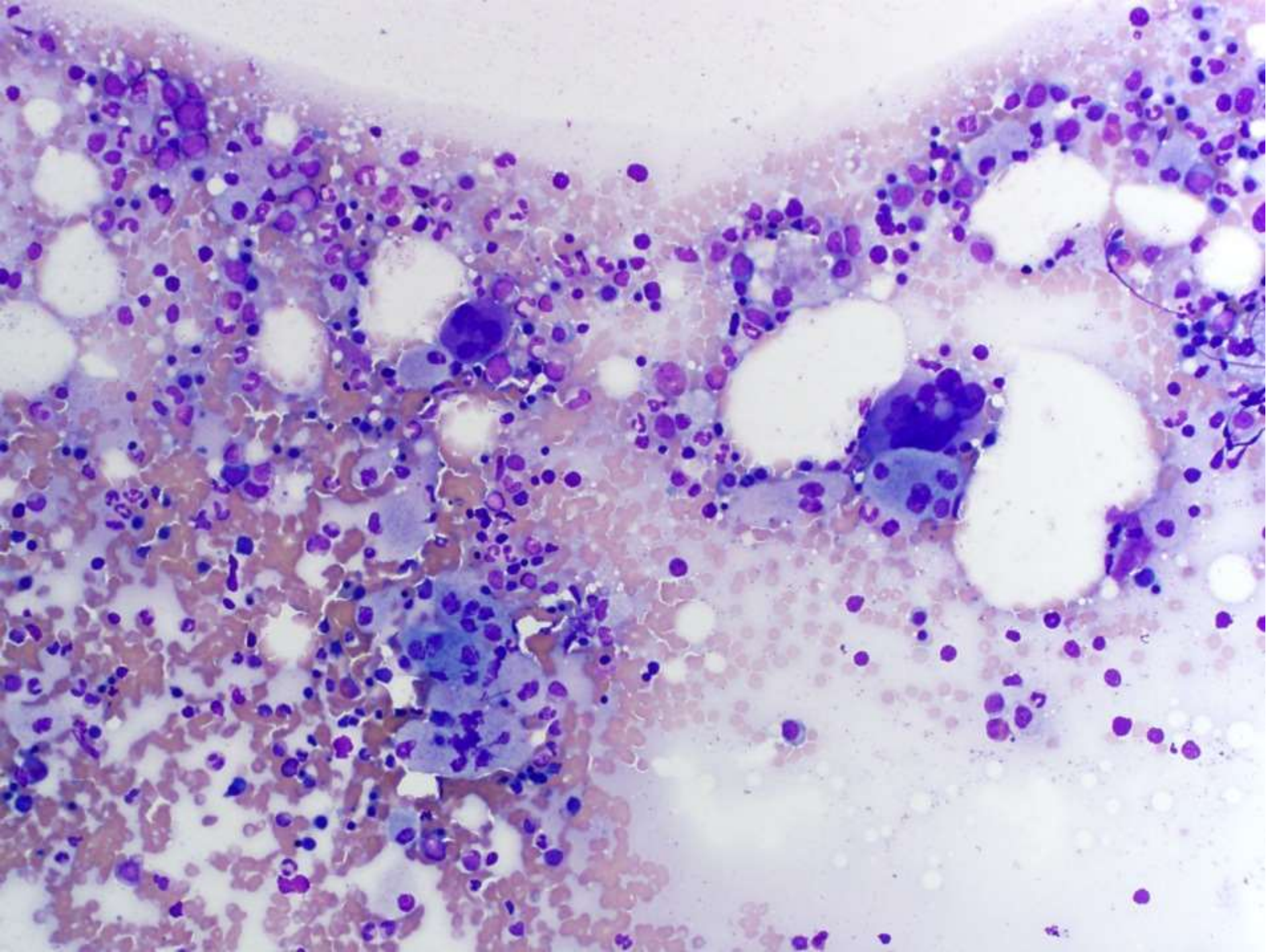
SB 5946

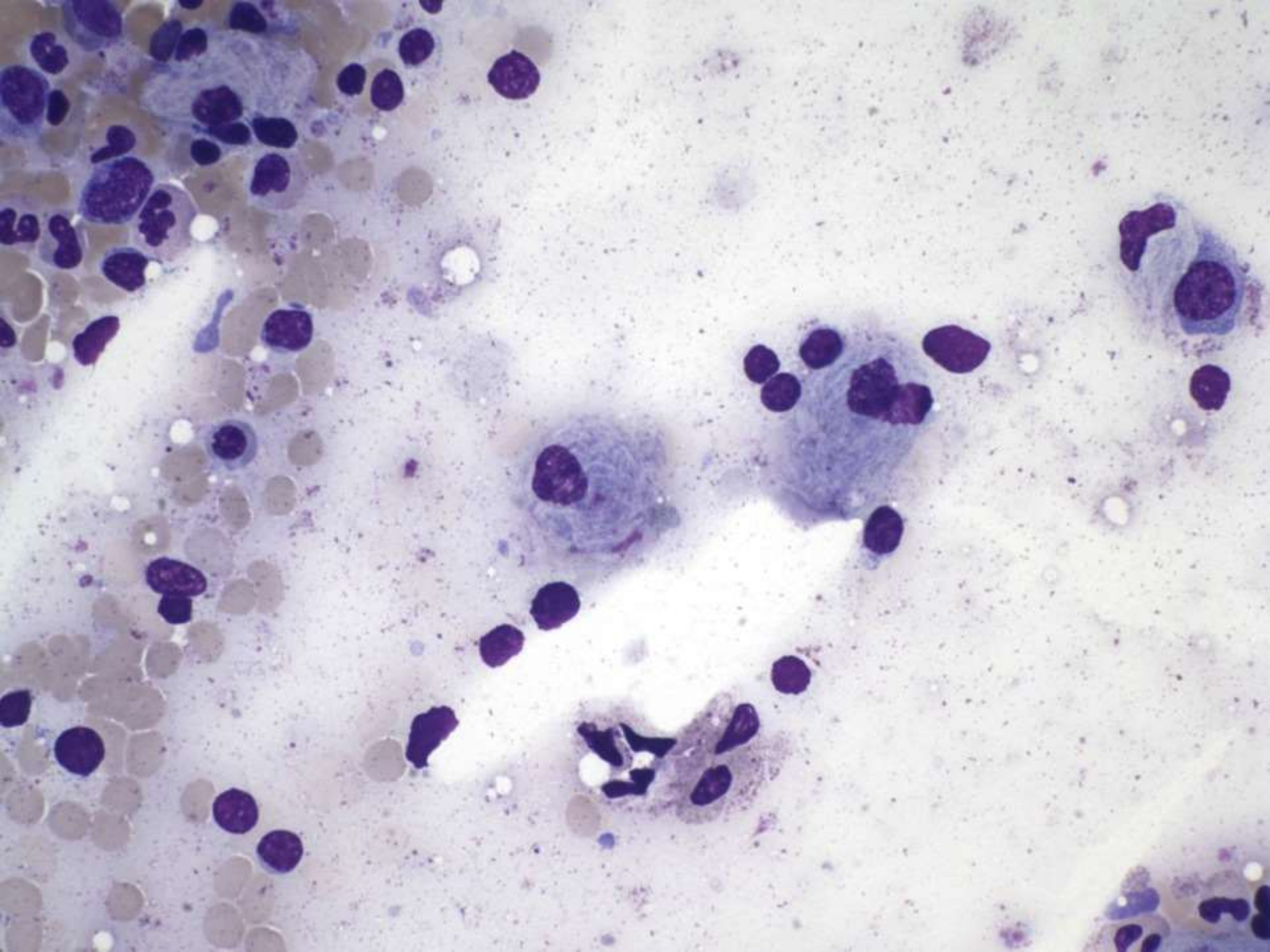
- 32-year-old male who presented with ras and with recent complains of early satiety and nausea. Was noted to have splenomegaly on physical exam. Later ultrasound imaging also showed hepatomegaly. CBC revealed thrombocytopenia. Bone marrow aspirate and biopsy performed.

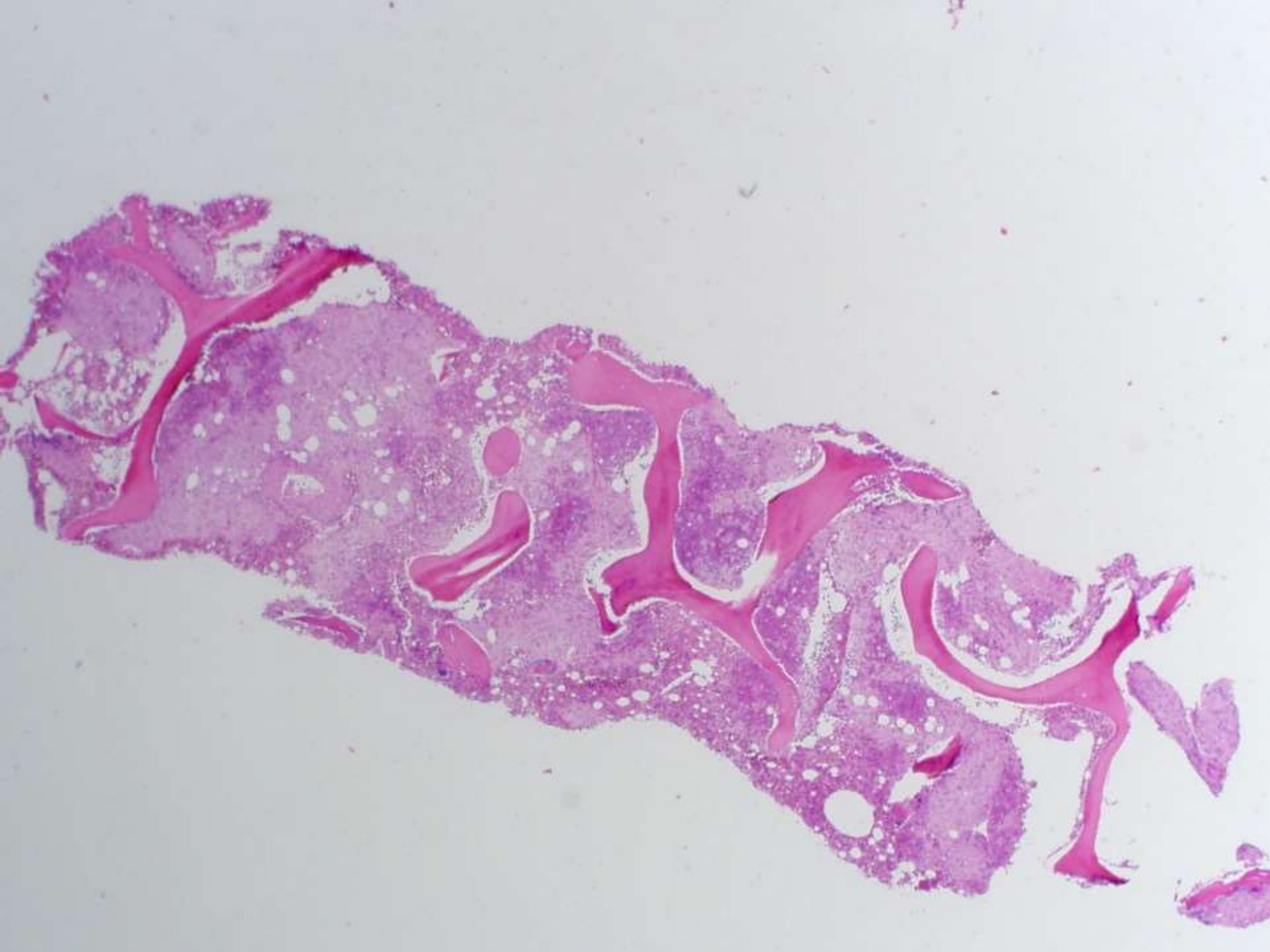
- **Ryan Johnson/Yaso Natkunam; Stanford**

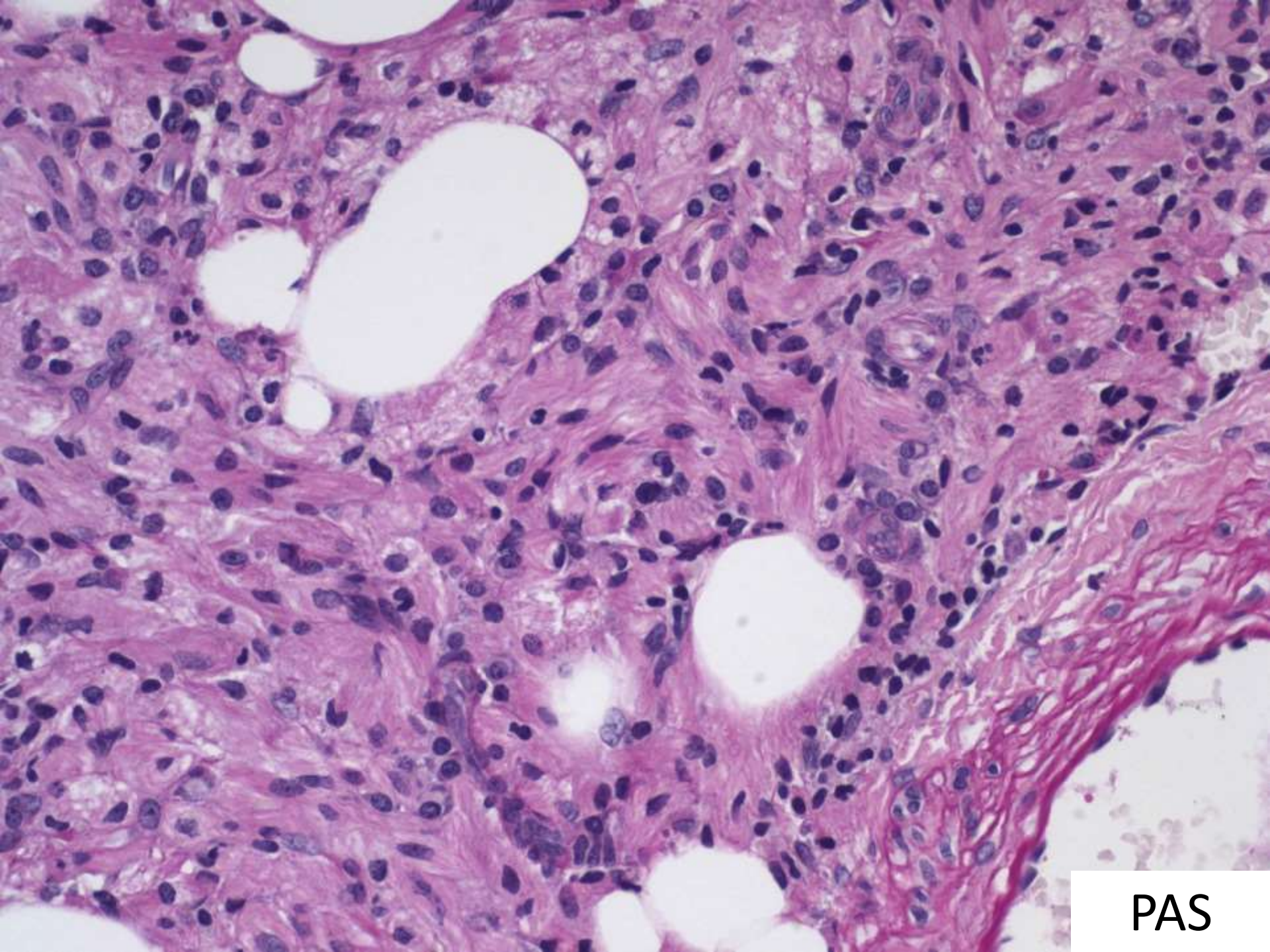








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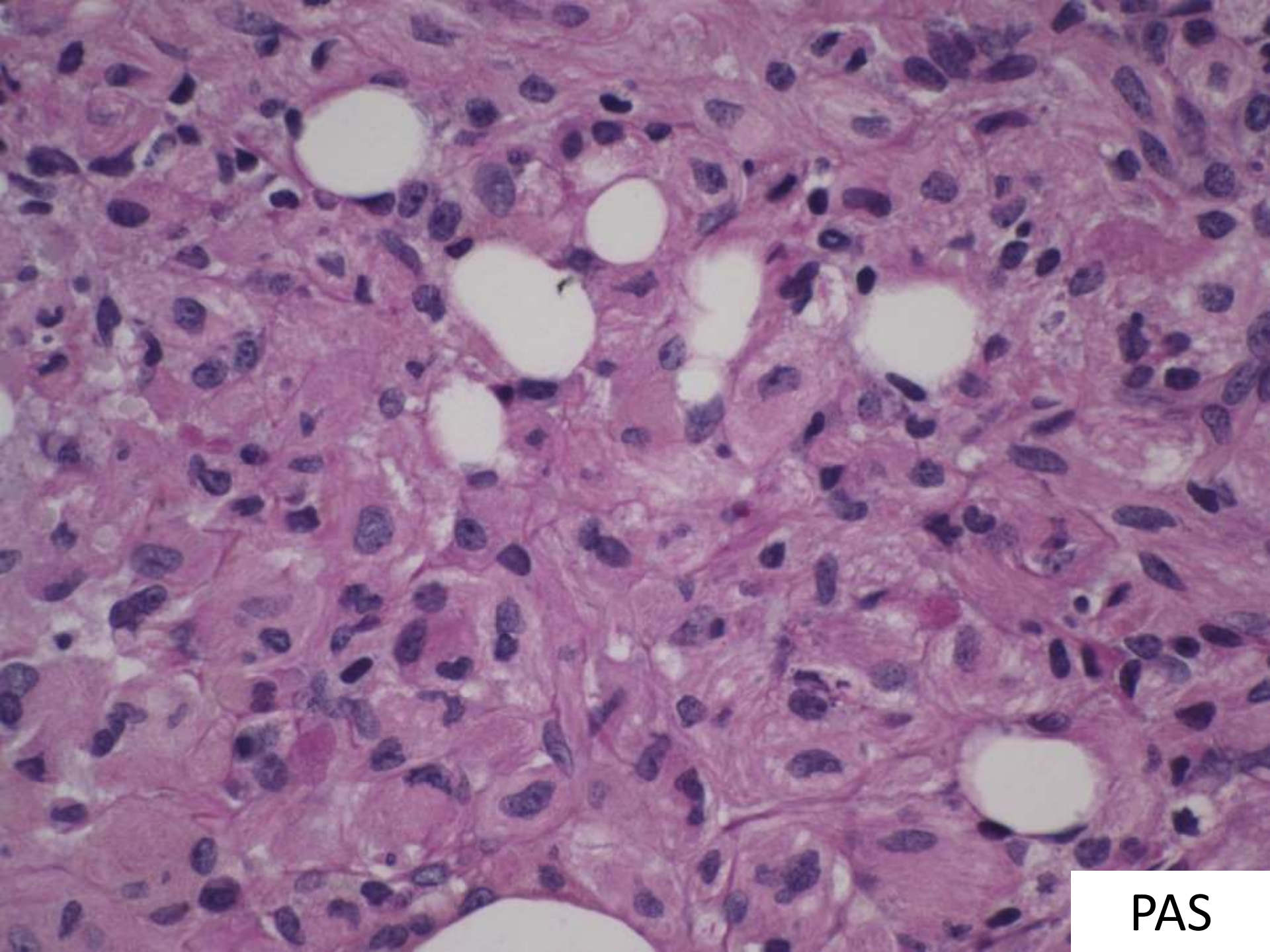
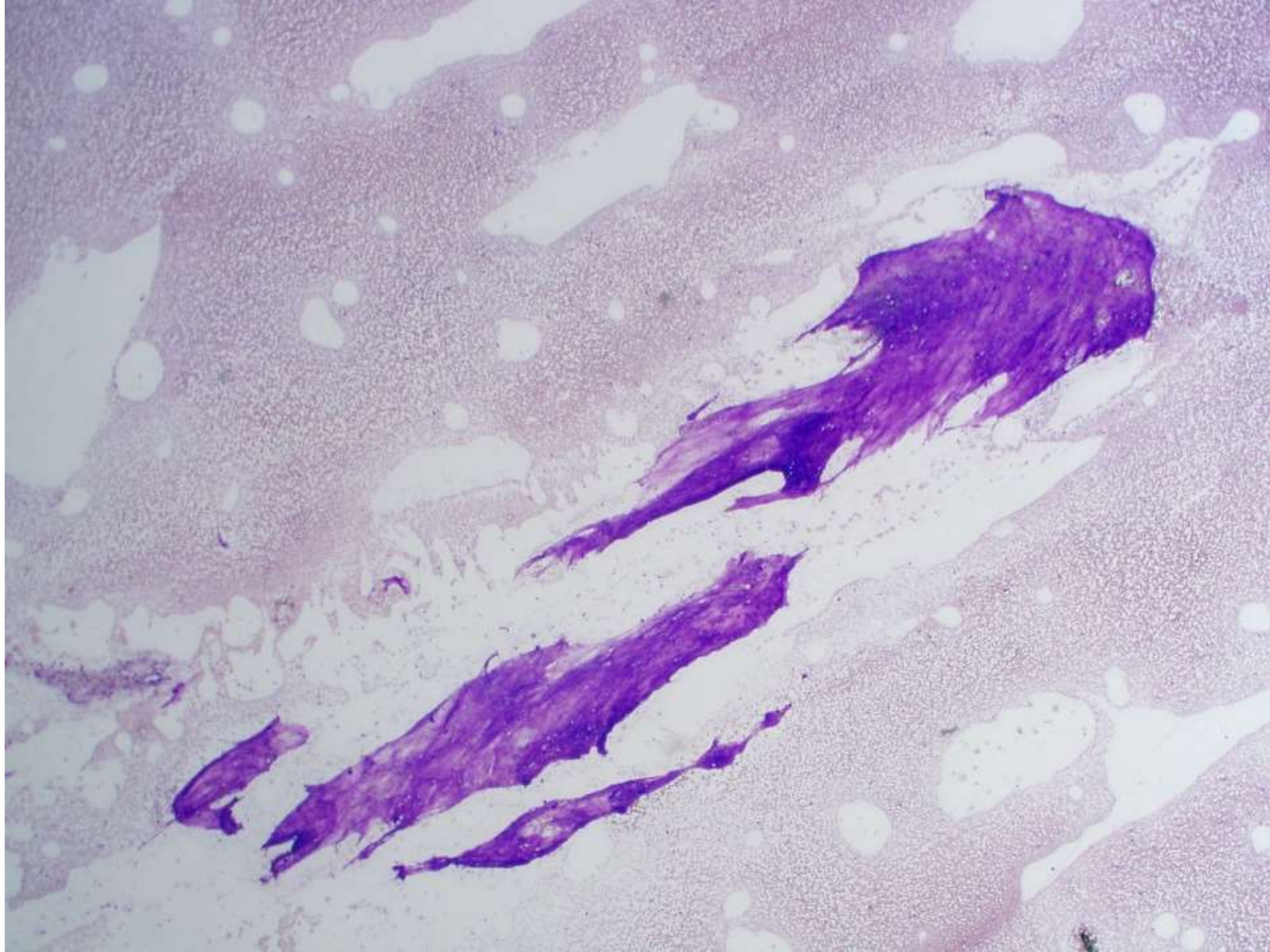

PAS

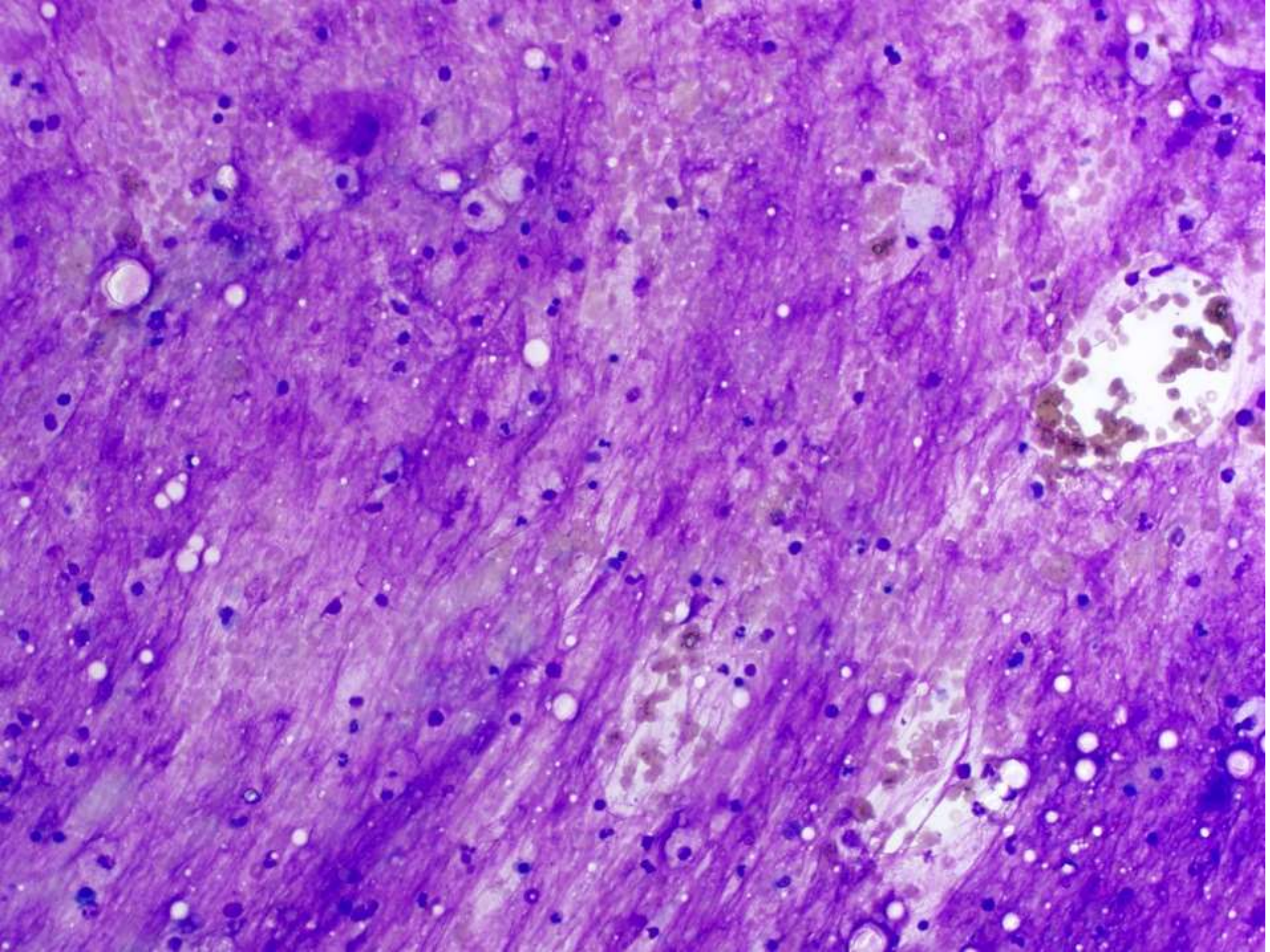
Diagnosis….???

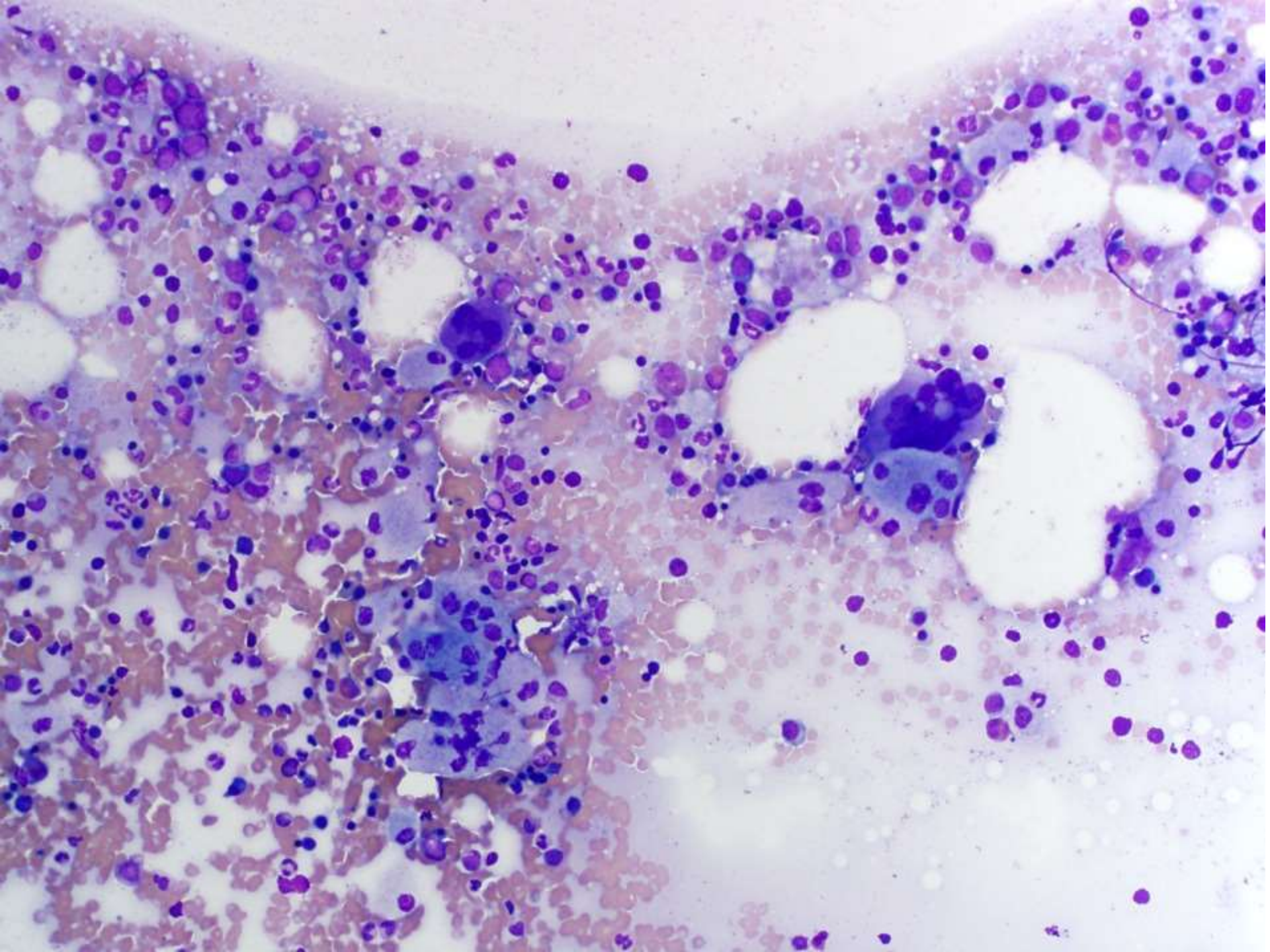
H&P and Labs

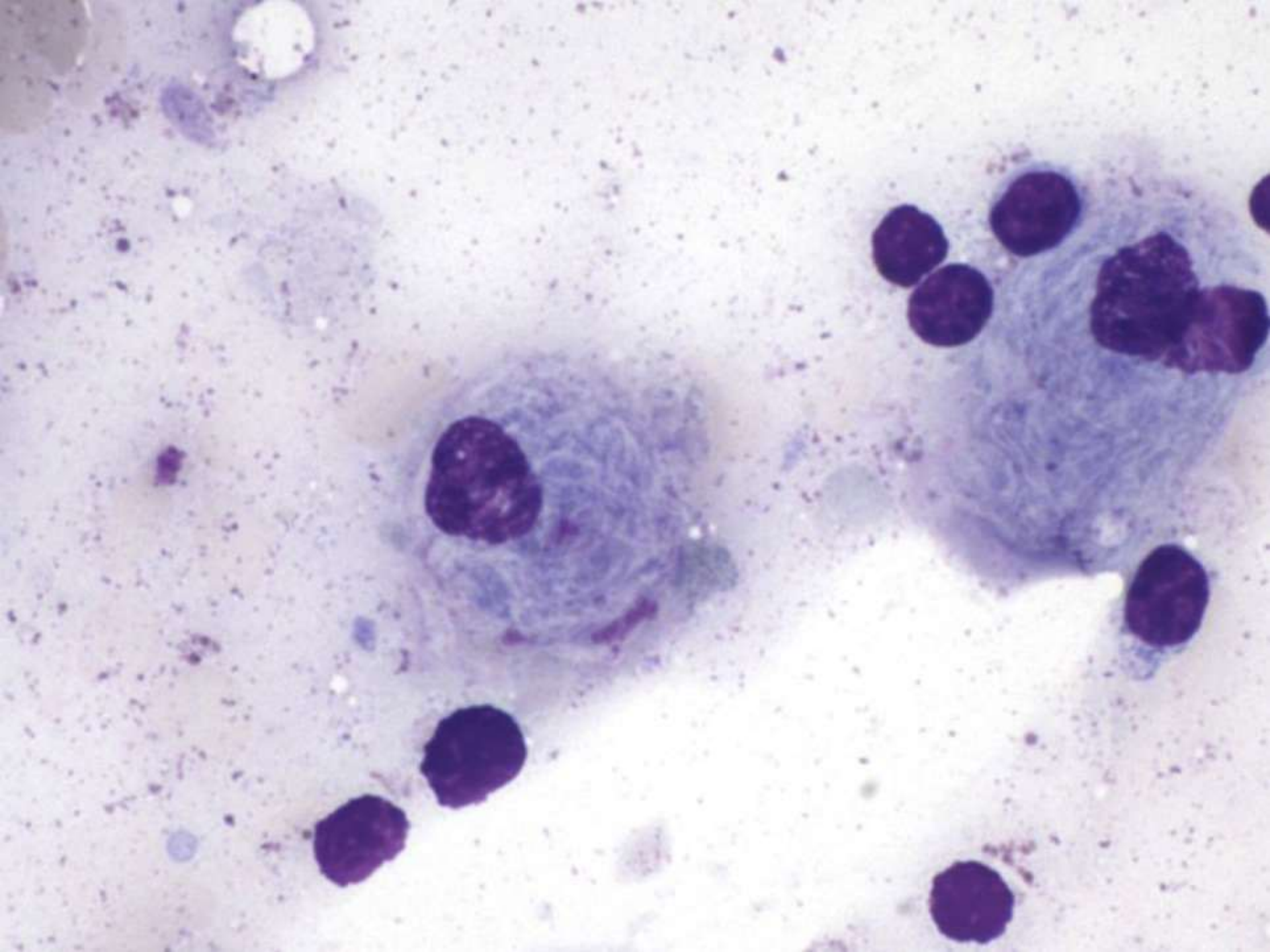
- 32 year old male who presented with rash and with recent complaints of early satiety and nausea
  - Was noted to have splenomegaly on physical exam
  - Later U/S imaging also showed hepatomegaly
  - CBC revealed thrombocytopenia
  - *Recently emigrated from Western Russia*

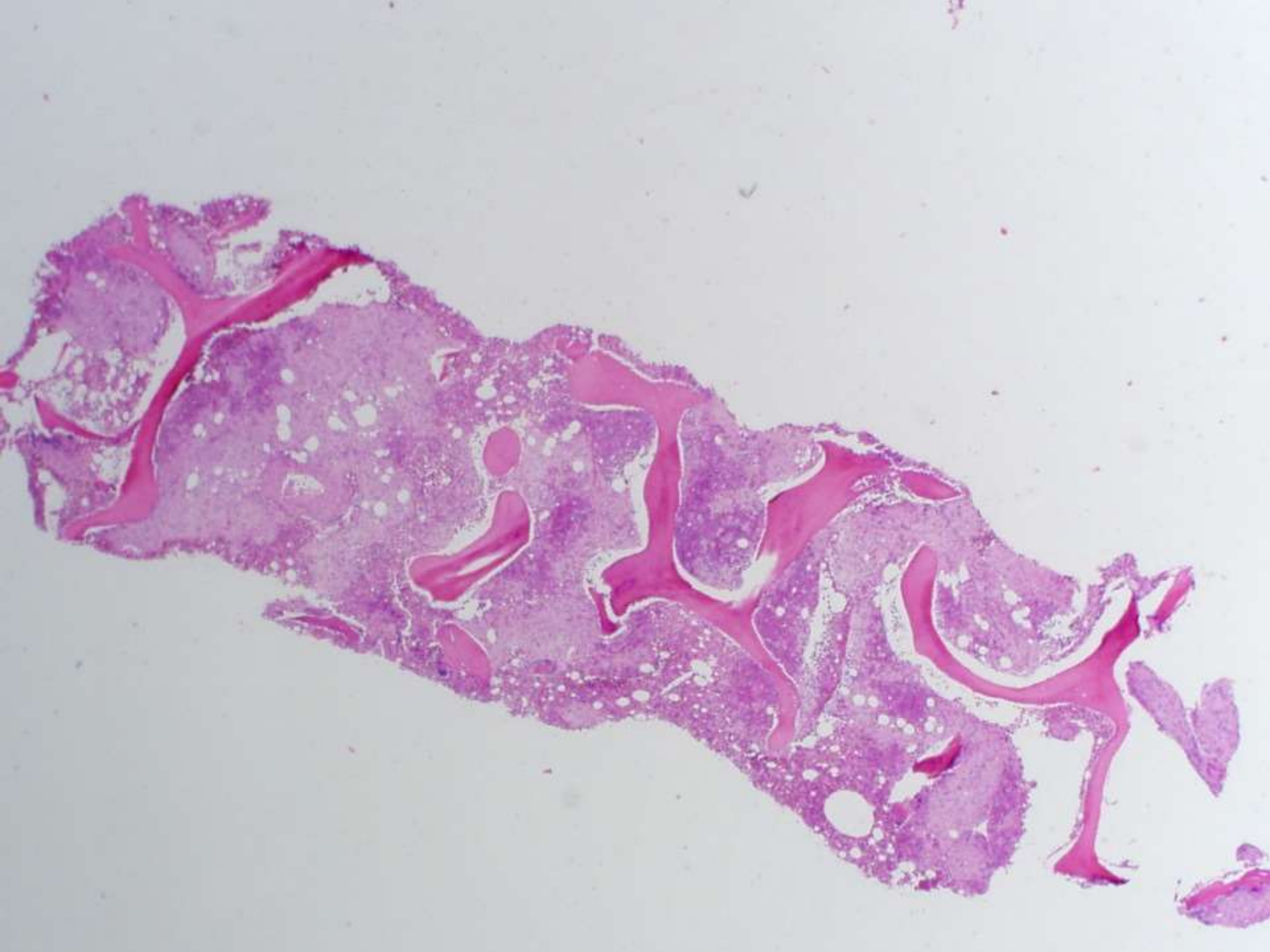
- CBC: WBC 5.4 K/uL; Hgb 14.1 g/dL; PLT 63 K/uL; MCV 89.3

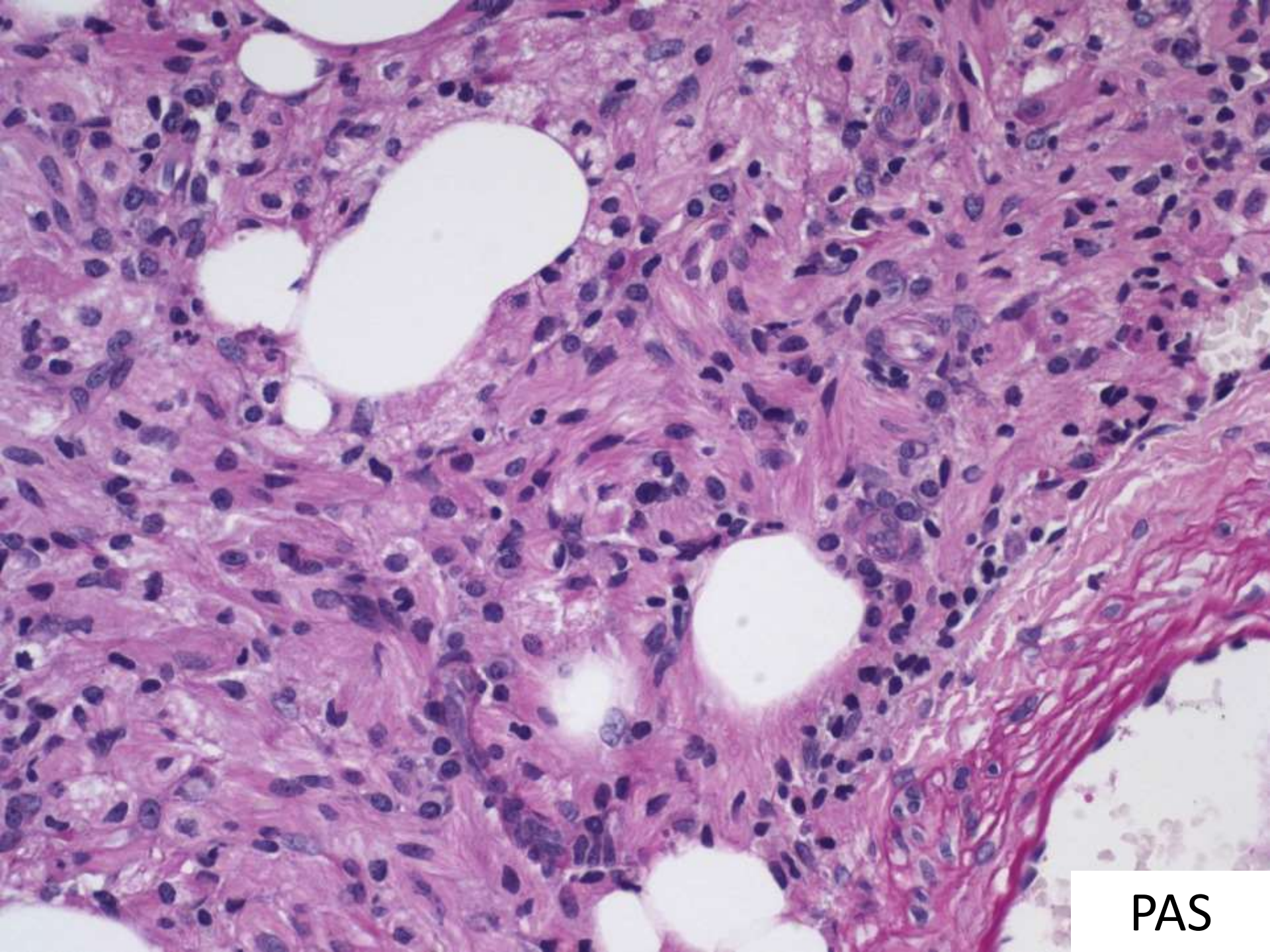








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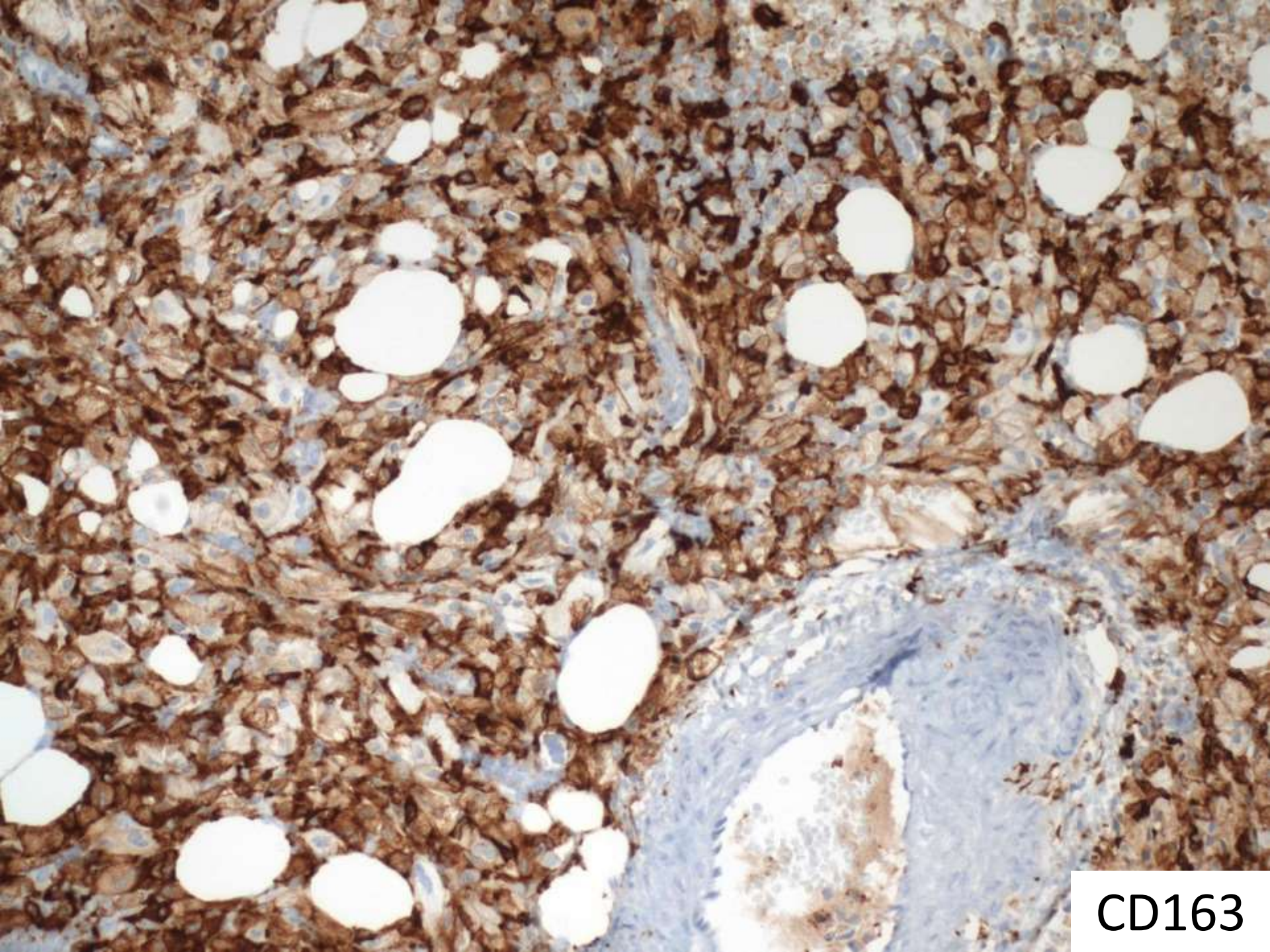

CD163

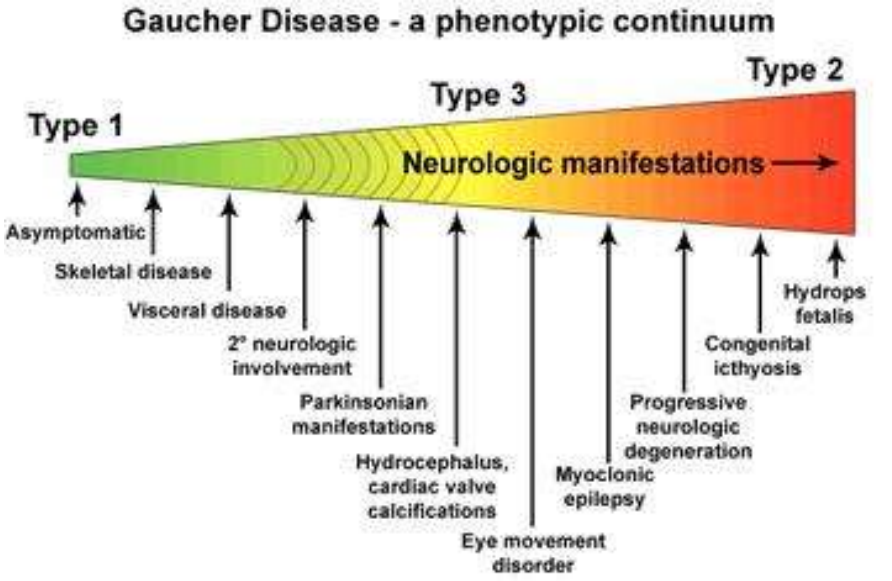
Additional Studies

- AFB, Fite, and GMS studies were negative

- Patient was noted to be heterozygous for the N370S mutation in glucocerebrosidase (GSA) gene
  - Consistent with Carrier status for Gaucher
  - Pending sequence analysis of rare mutations
  - Asymptomatic so withholding enzyme therapy at present

Gaucher disease

- Lysosomal storage disorder (glucocerebrosidase A)
- Autosomal recessive
- Three types (1,2,3) – type 1 is most common and usually presents at adult age
  - Ashkenazi Jewish population (carrier rate is 1 in 10)
  - Symptoms depend on subtype
- Increased risk of myeloma (5x) and Parkinsonism later in life
- Treatment with enzyme replacement therapy is mainstay



Patients with Gaucher disease can have a spectrum of symptoms, ranging from mild to severe neurological effects. The classic categories of types 1, 2 and 3 have blurry edges along this continuum.

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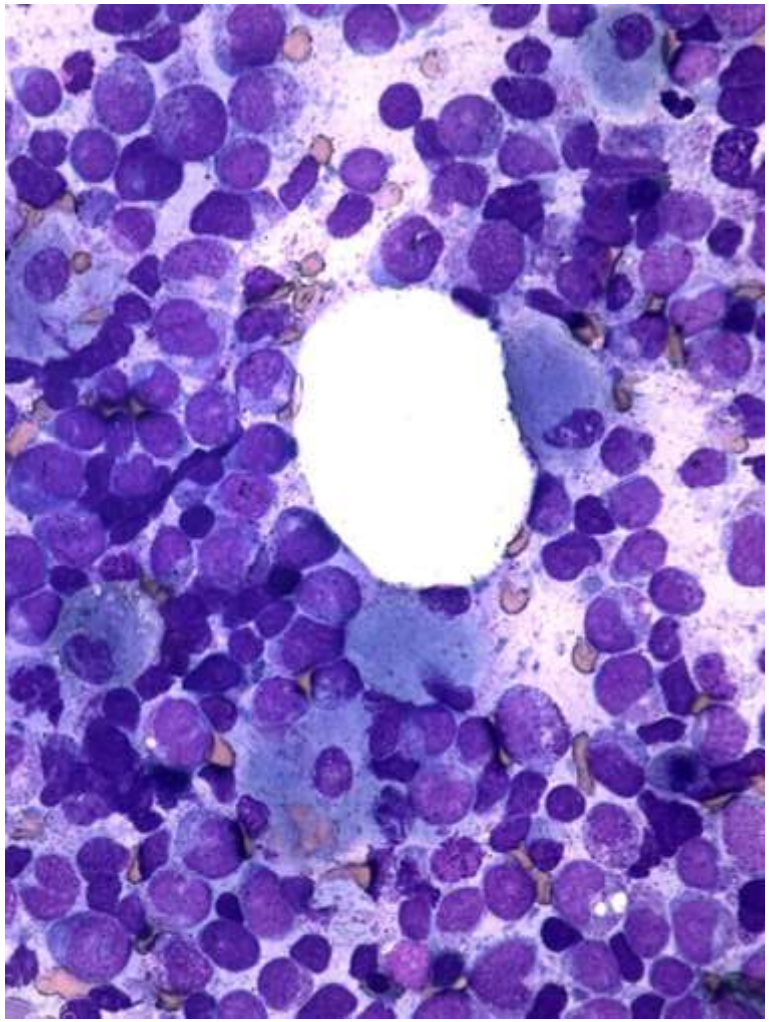
Differential diagnosis of ‘sea blue histiocytes’

- Gaucher disease
- ‘states of high bone marrow turnover’
  - Myeloproliferative neoplams (e.g. CML)
  - Myelodysplastic syndrome
  - Idiopathic/immune thrombocytopenic purpura
  - LBL, non-Hodgkin lymphomas
  - Beta thal major
- Neiman-Pick disease (sphingomyelinase def.)
- Total parenteral nutrition (high lipid emulsion)



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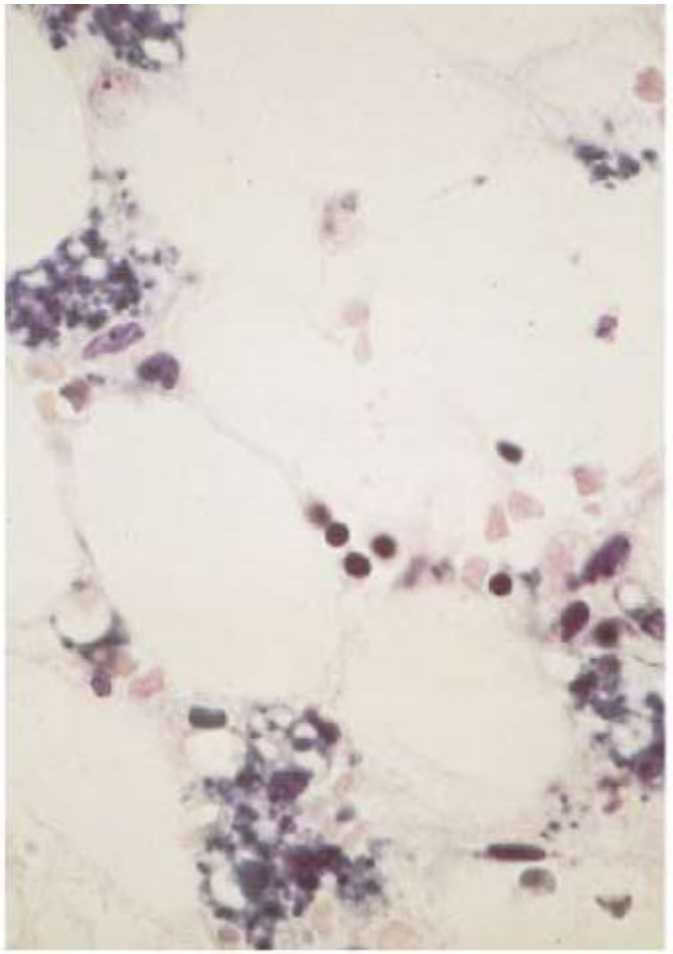

Bigorgne et al. BJH 1996; 95, 258-262

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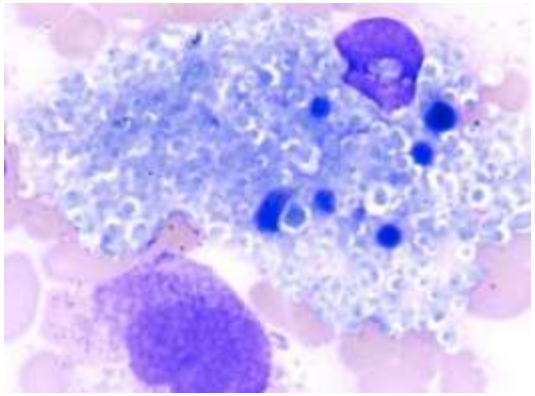
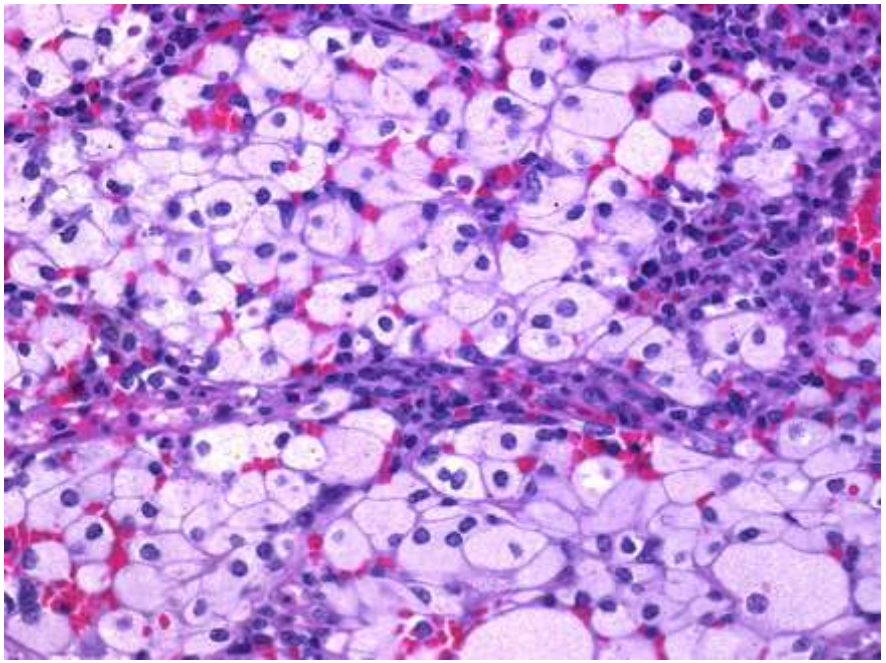
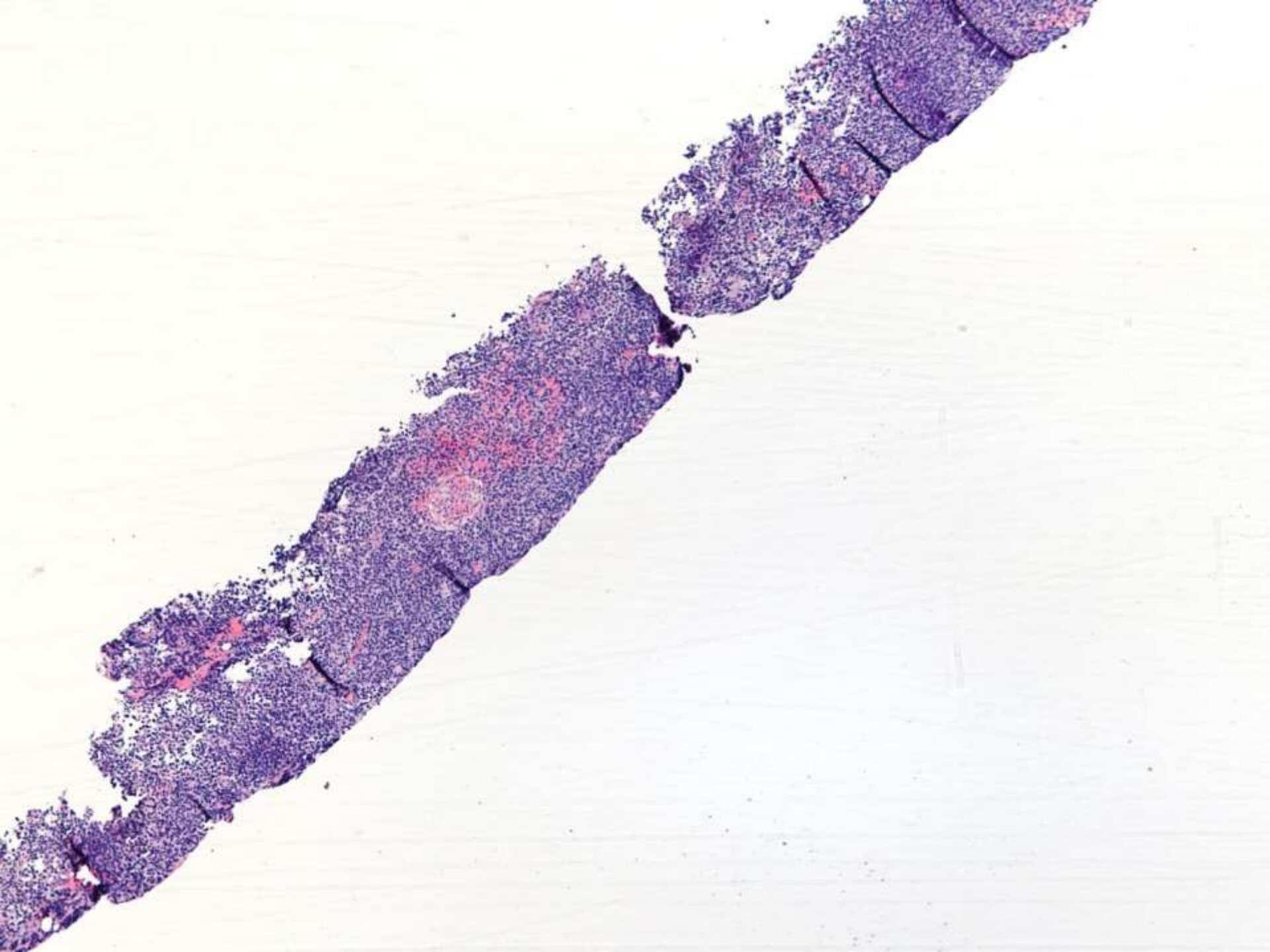


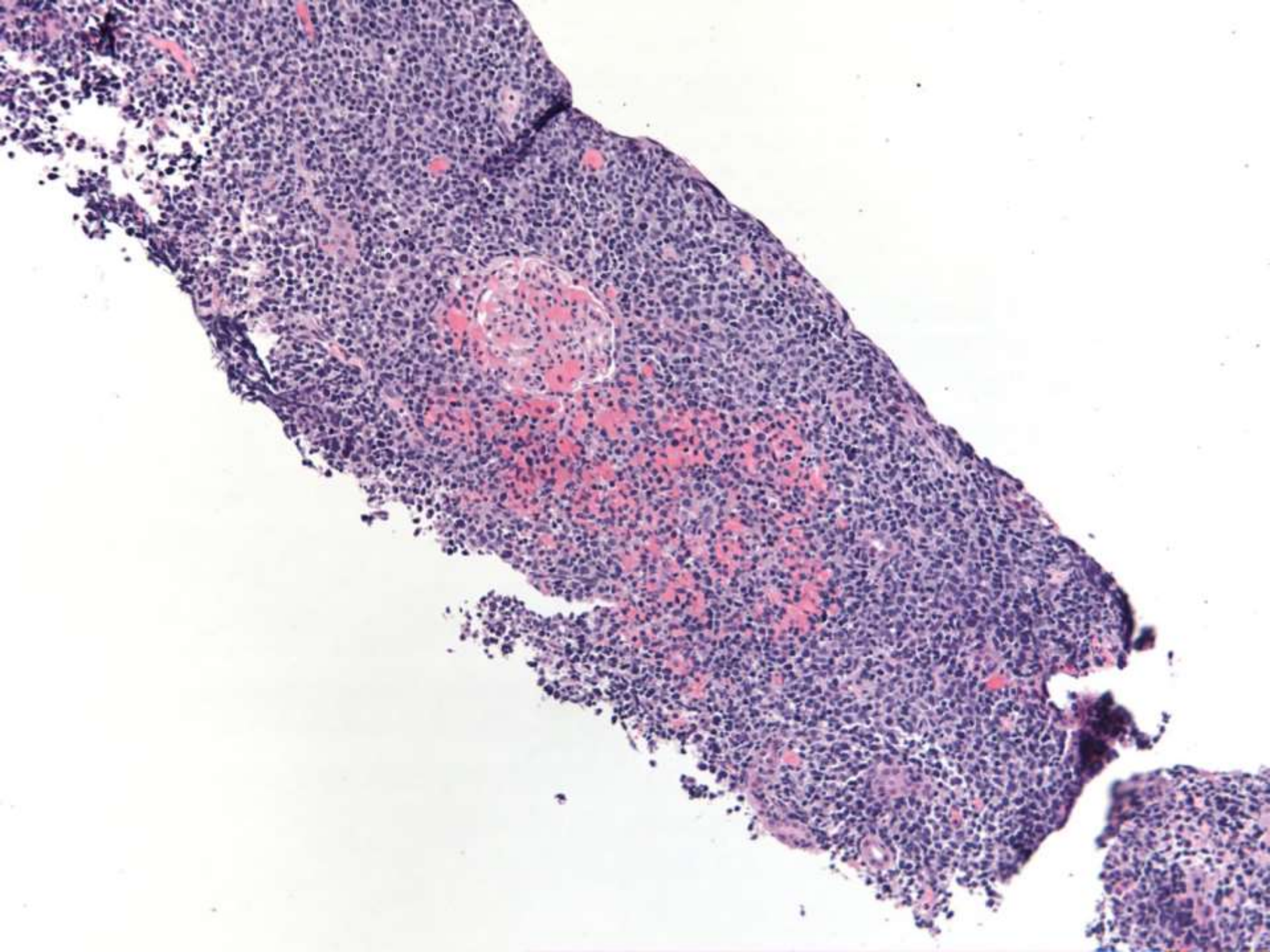


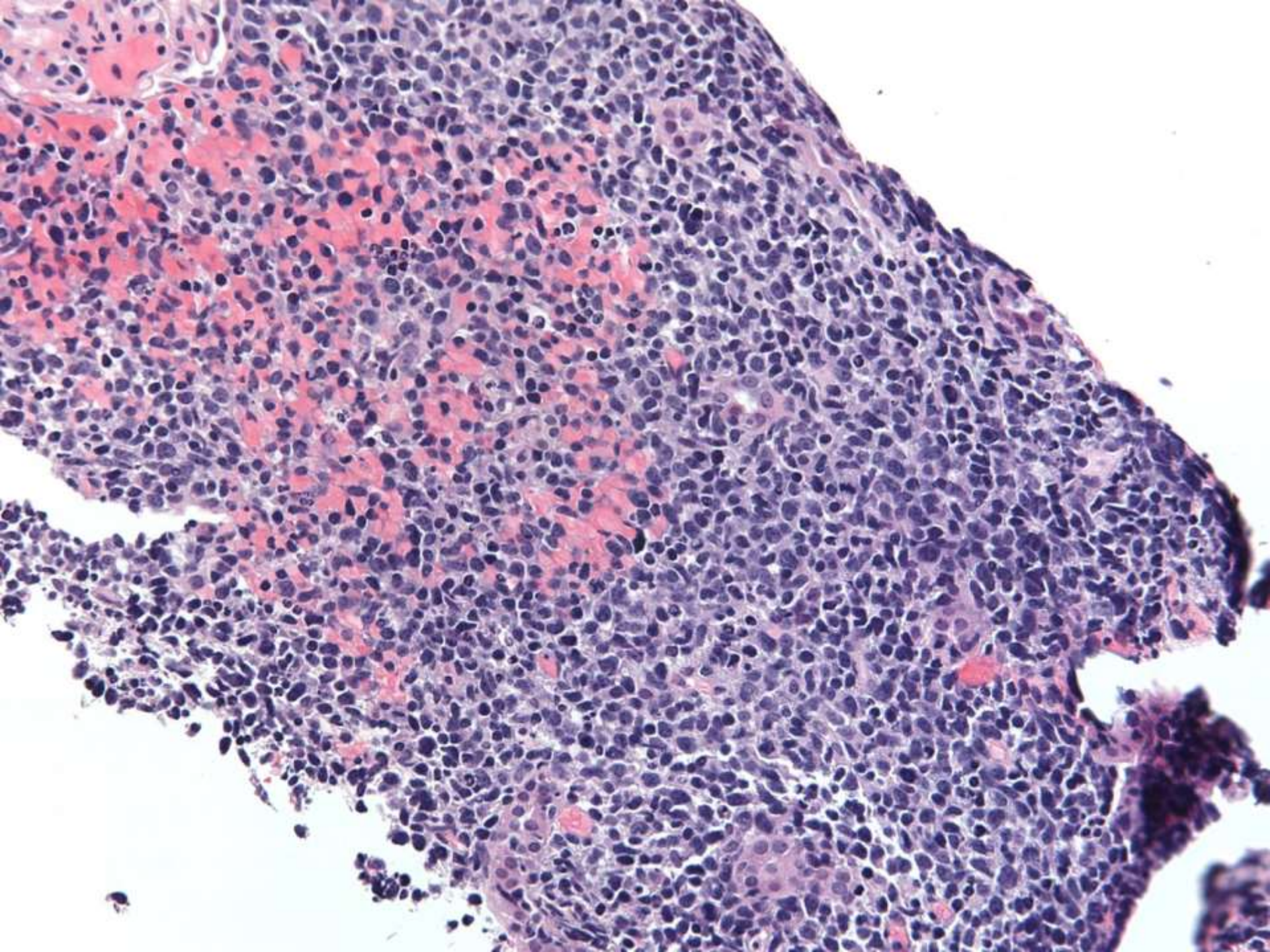
SB 5947

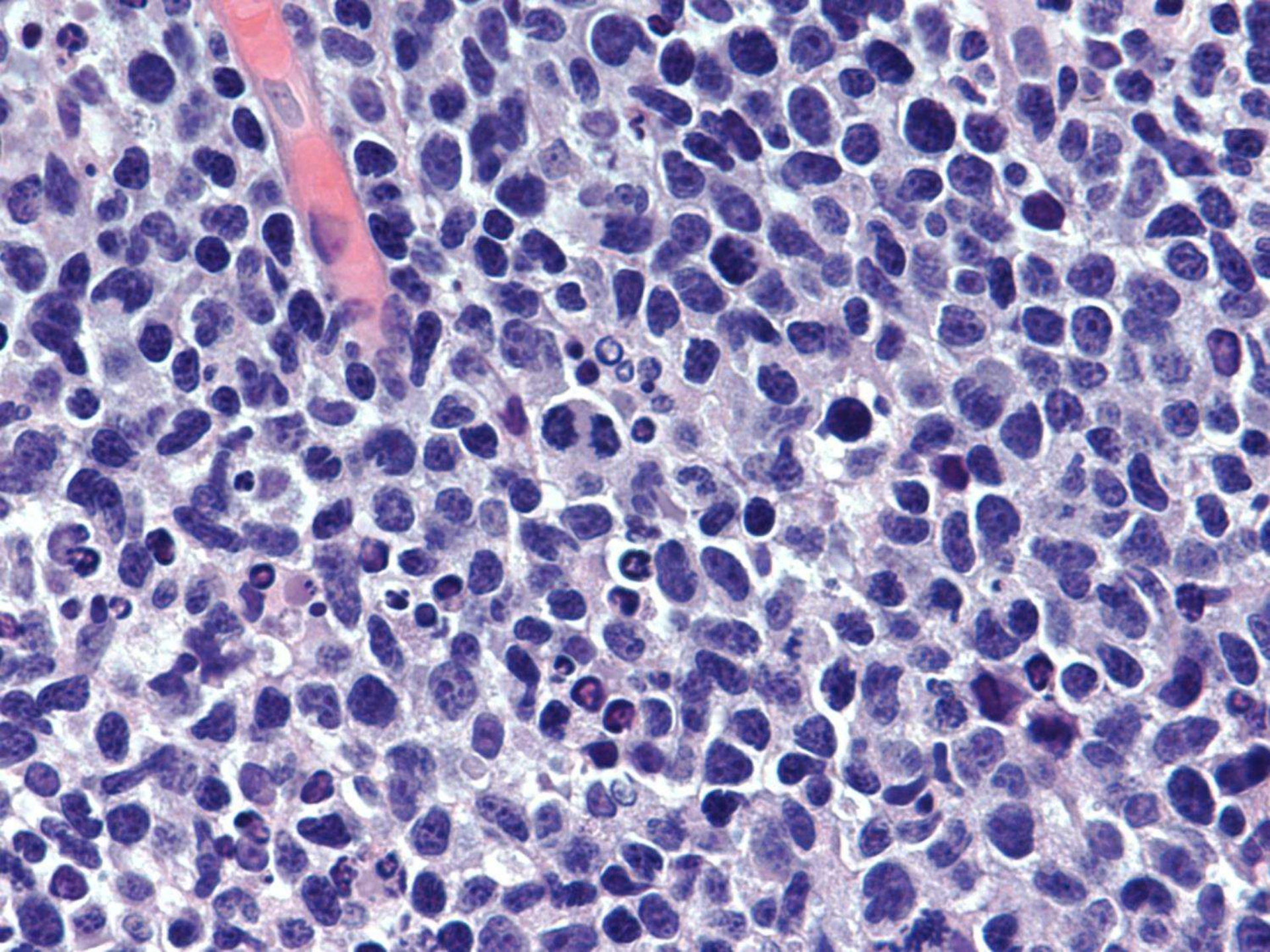
- 56-year-old Asian woman with bilateral renal and adrenal masses. Right kidney mass was biopsied.

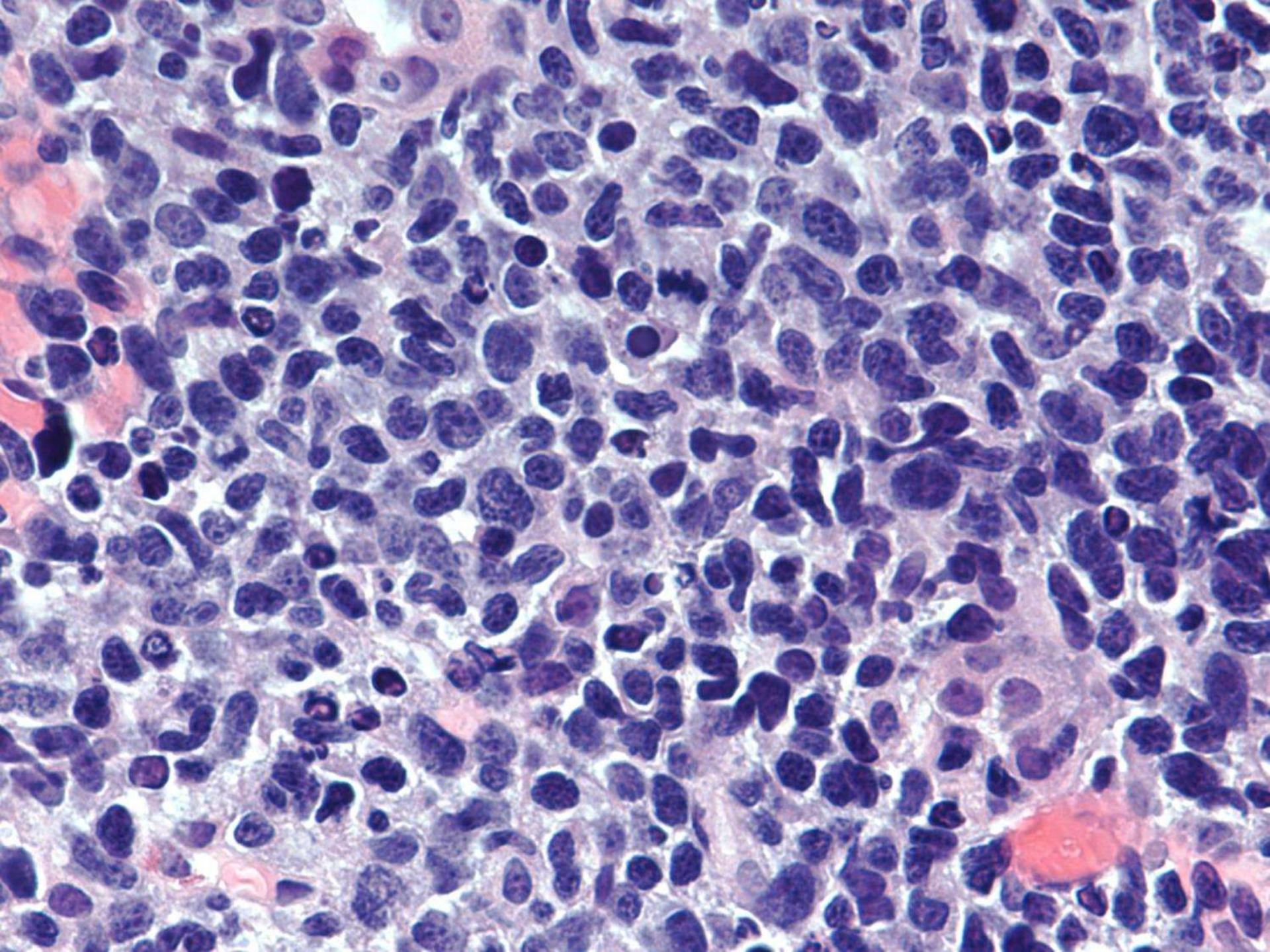
- **Linlin Wang/Daniel Arber; UCSF/Stanford**

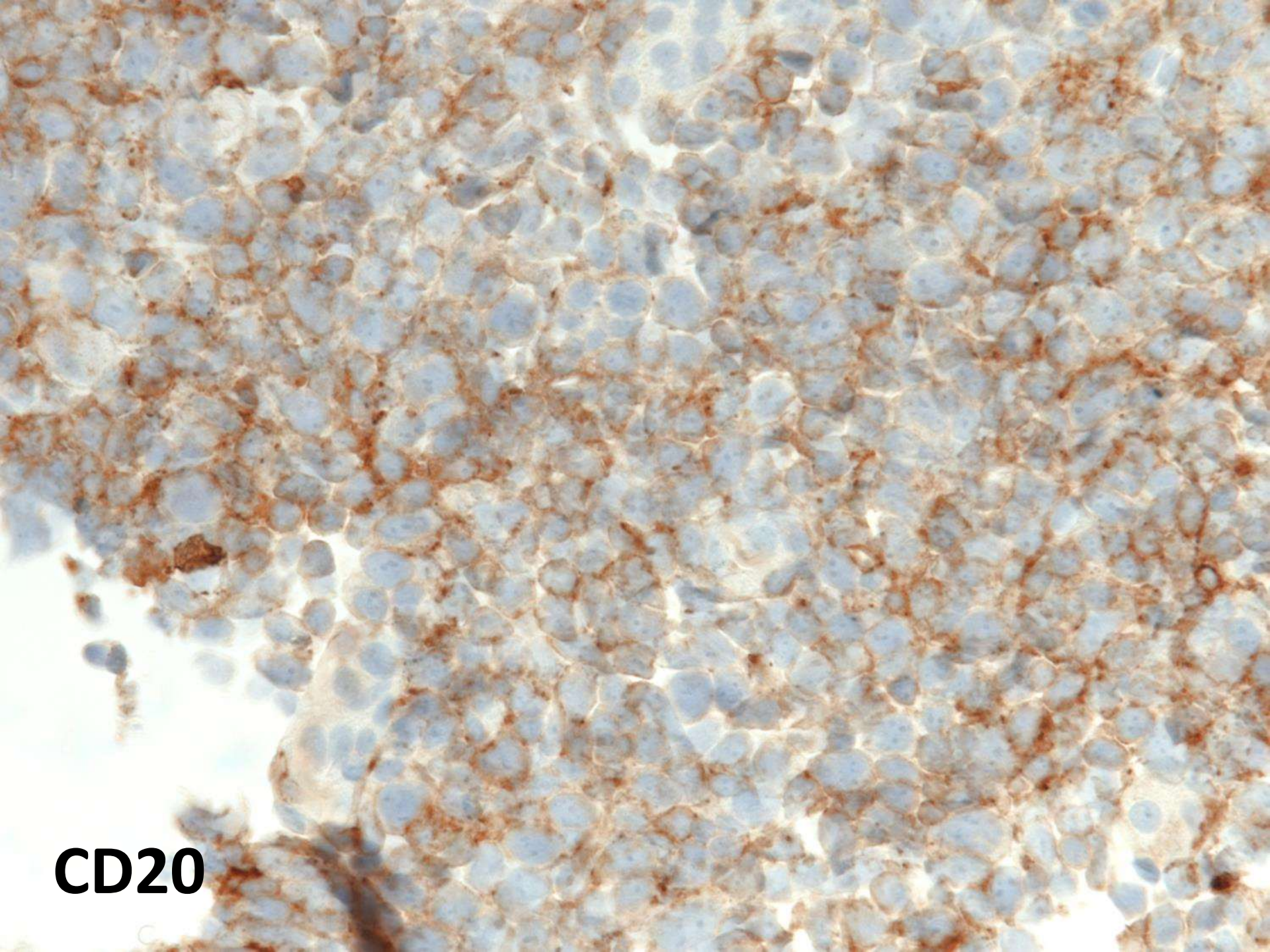








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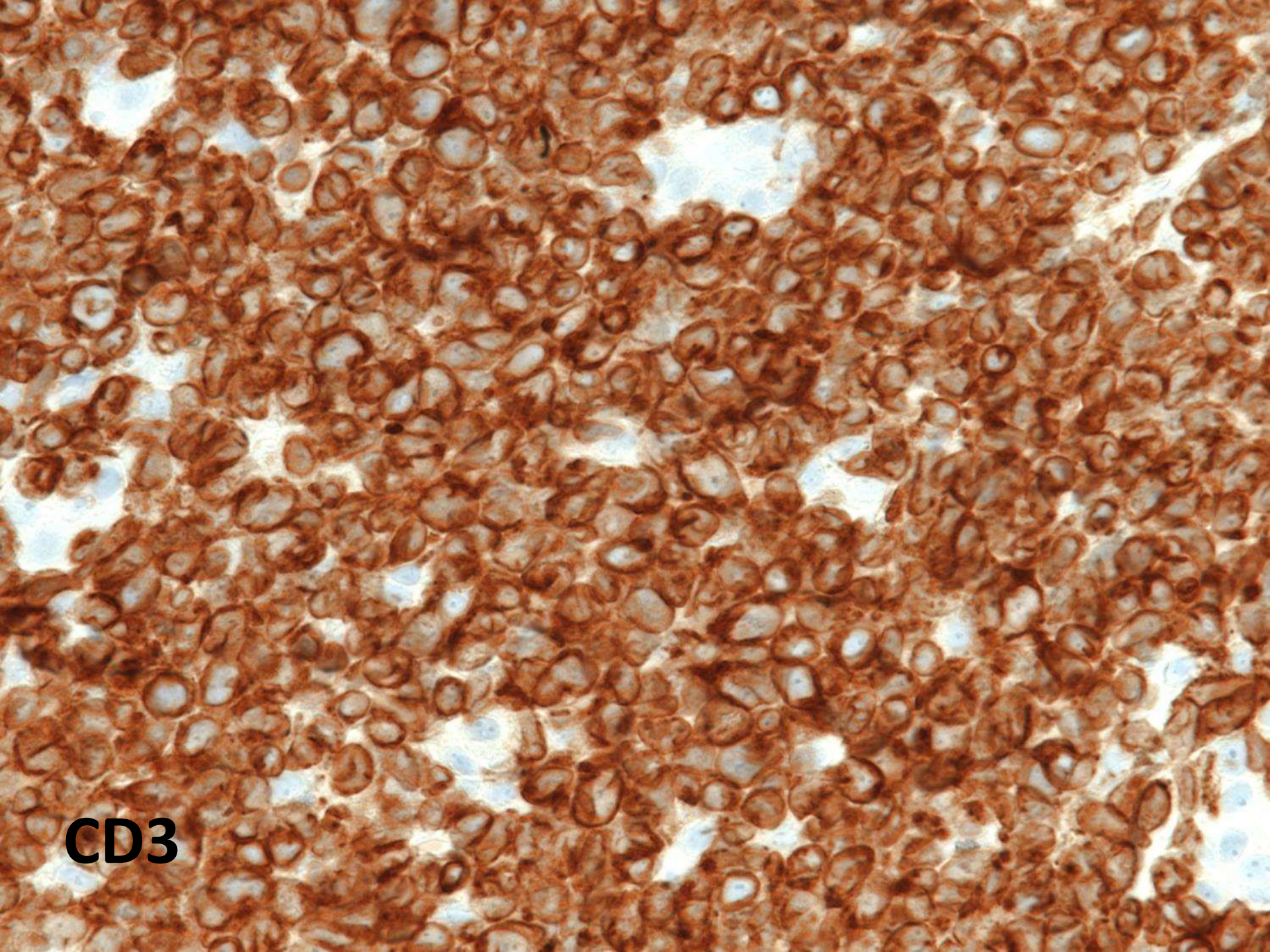

CD3

Ancillary Studies

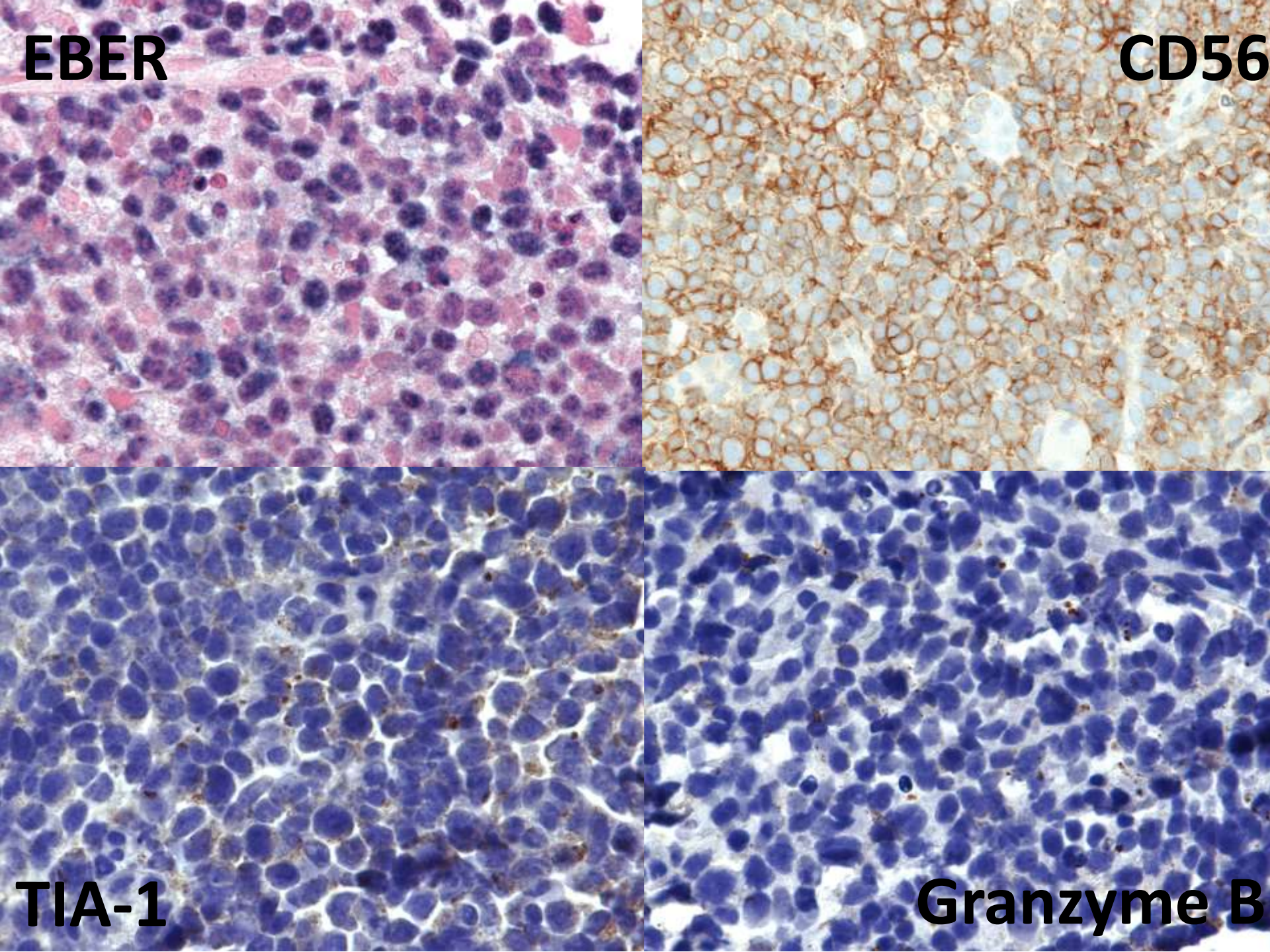
- Flow cytometry:

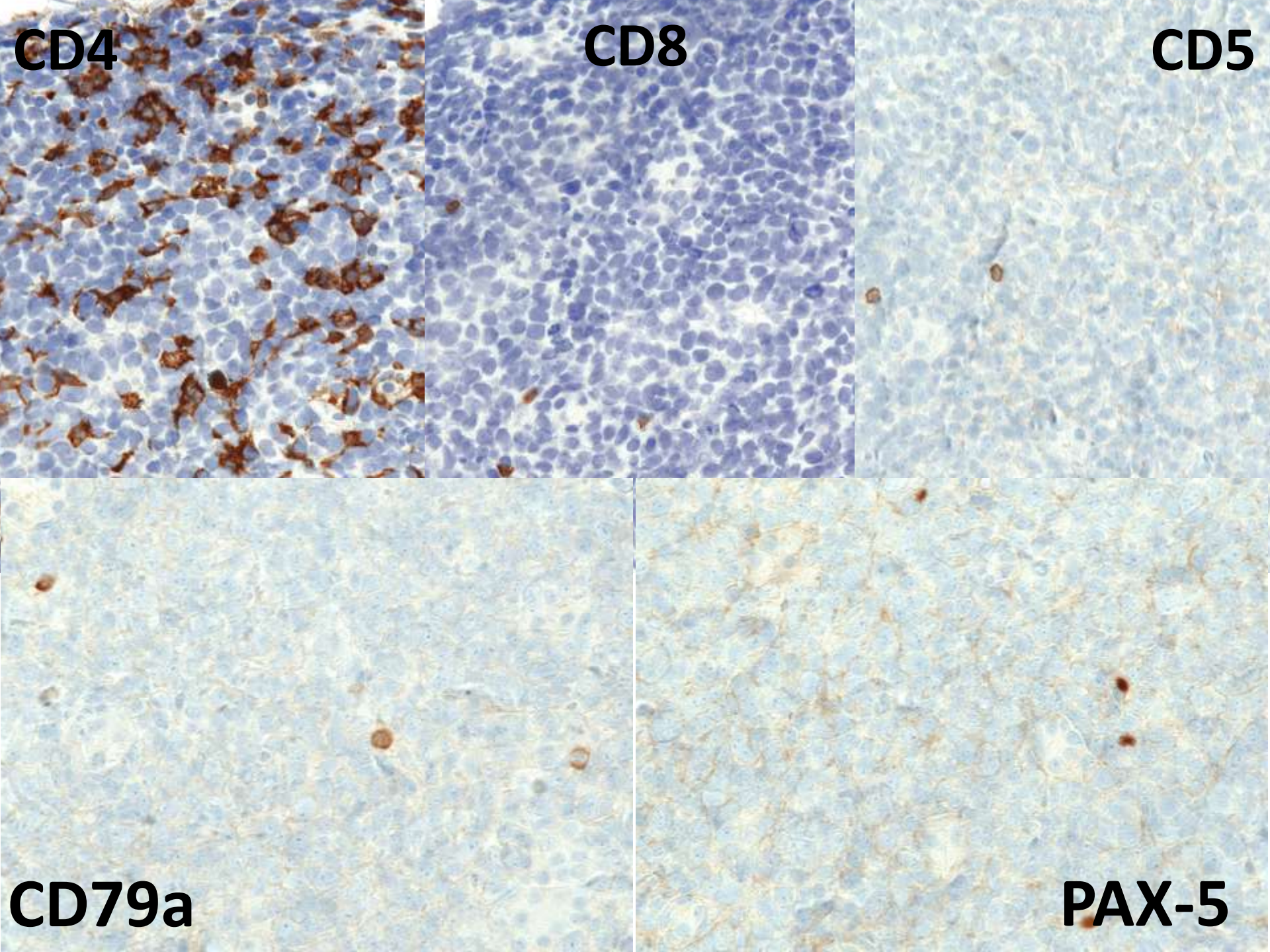
  Predominance of T-cells with loss of surface CD3

- Molecular study:

  No evidence of a clonal TCR gene rearrangement

Diagnosis….???


EBER
CD56
TIA-1
Granzyme B


CD4
CD8
CD5
CD79a
PAX-5

Diagnosis:

Extranodal NK/T-cell lymphoma

Extranodal NK/T-cell Lymphoma

- Most prevalent in East Asia, Mexico, Central and South America
- Commonly occurs in nasal cavity, may occur at any site (skin, GI, testis, soft tissue)
- Associated with EBV
- Aggressive disease with variable prognosis (5-year survival rate: 40%-78%)

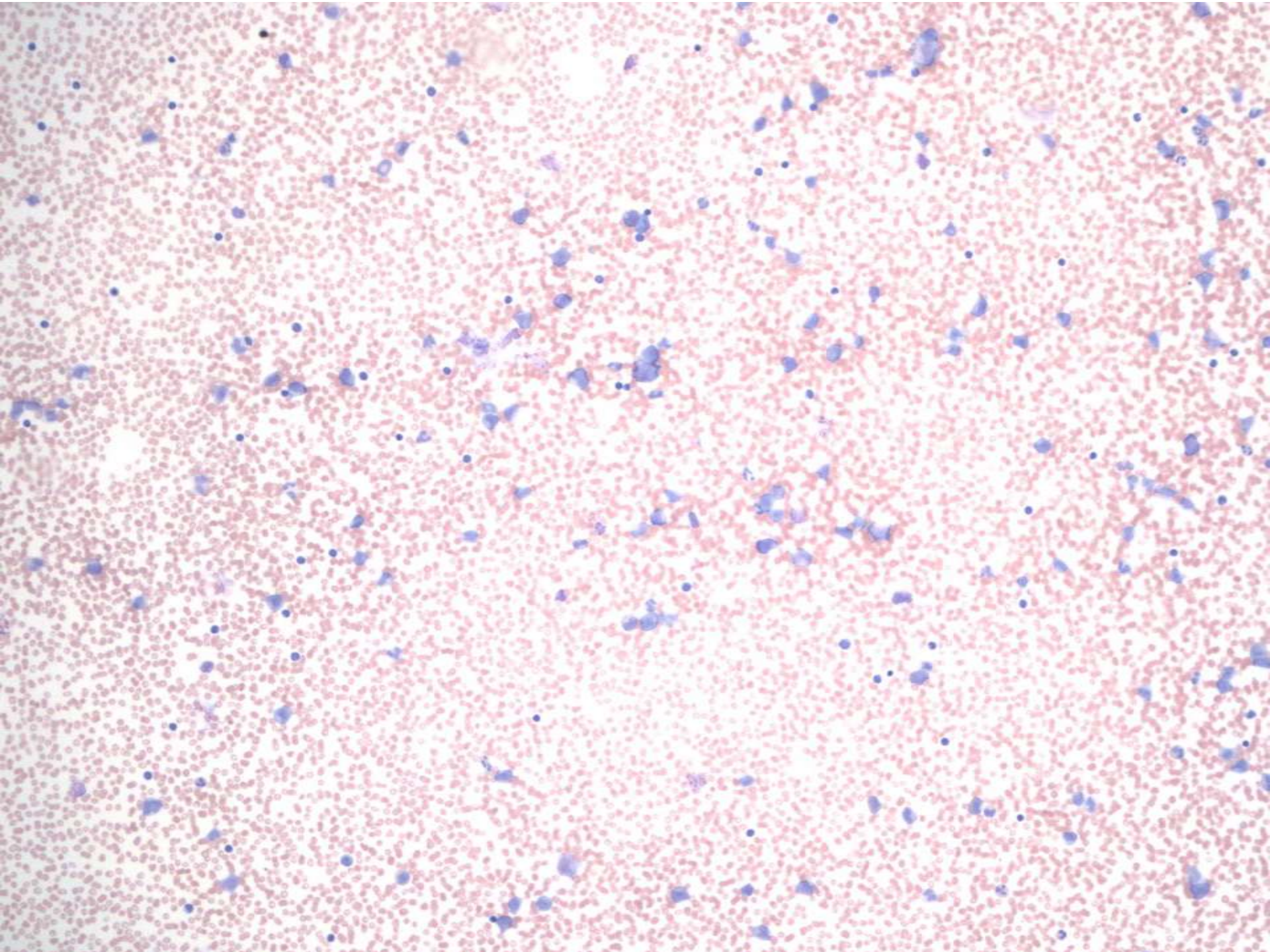
Extranodal NK/T-cell Lymphoma

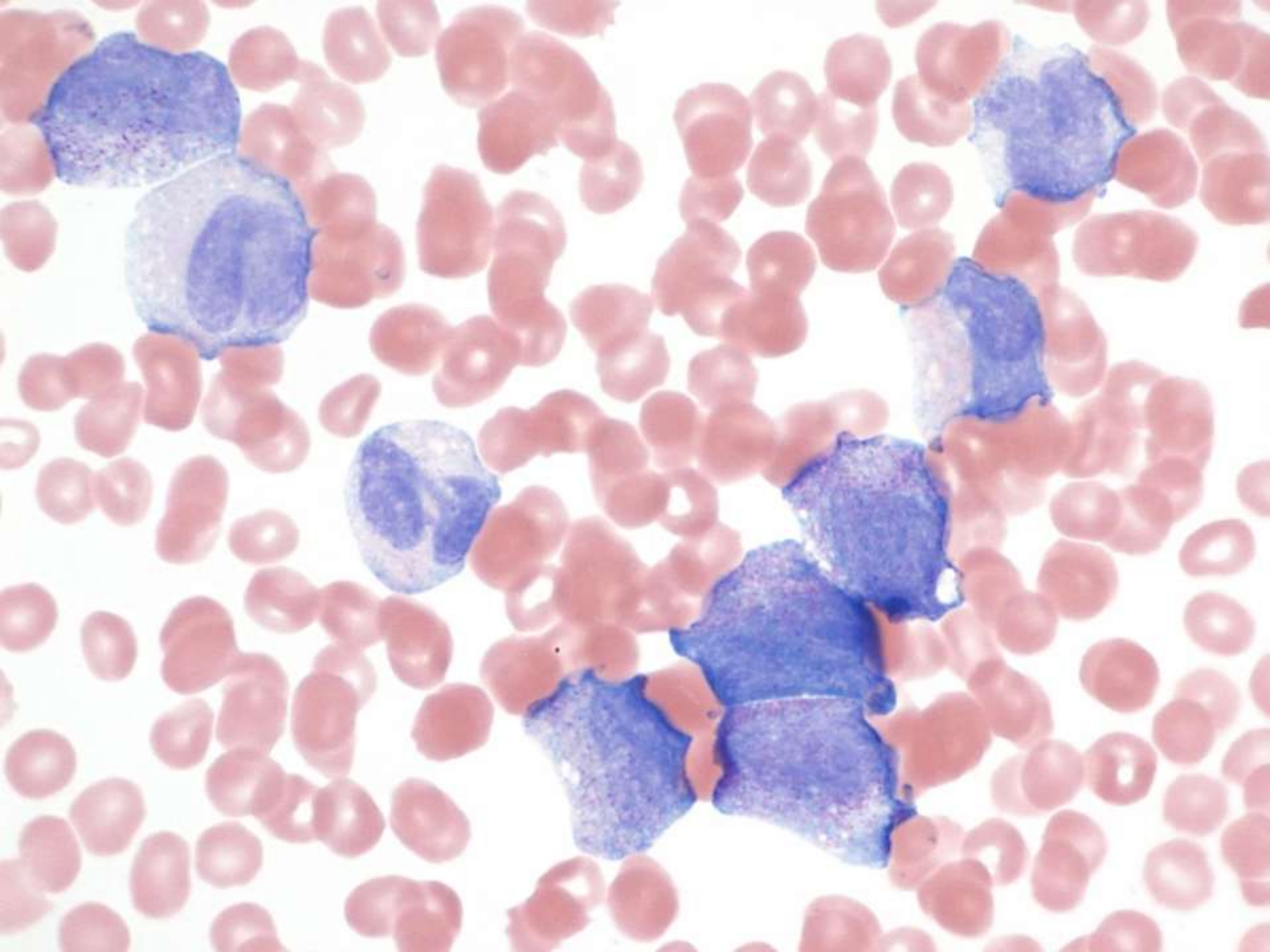
- Morphology
  - Angioinvasive infiltrate of atypical lymphocytes with varying size
  - Necrosis
- Immunophenotype: EBV+
  - NK cells: surface CD3-, cCD3+, CD56+, CD2+ TIA-1+, granzyme B+, perforin +
  - T-cells: surface CD3+, TCR+, TIA-1+, granzyme B+, perforin +
- Molecular study
  - NK cells: clonal TCR rearrangement negative
  - T-cells: clonal TCR rearrangement positive

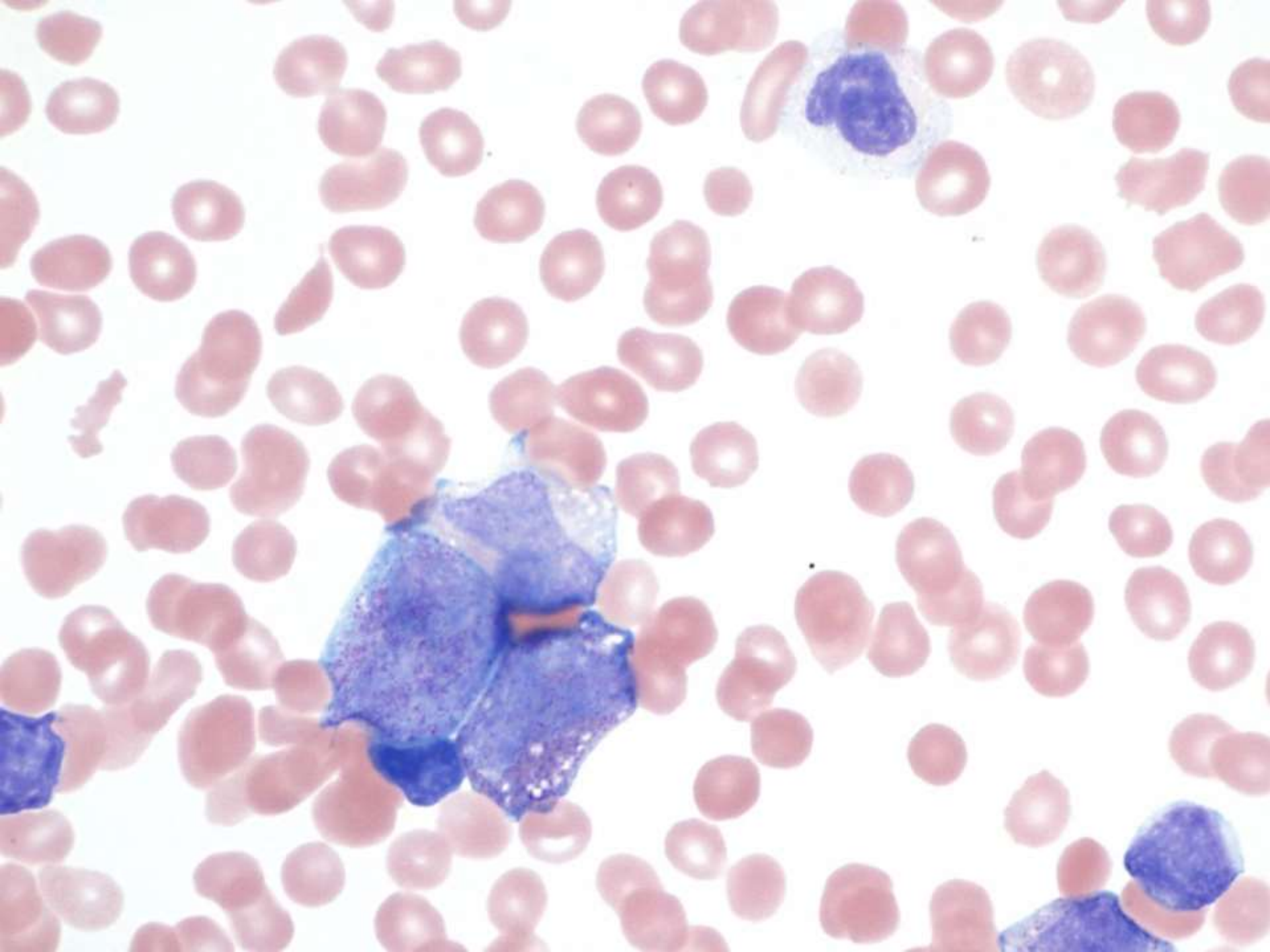
SB 5948

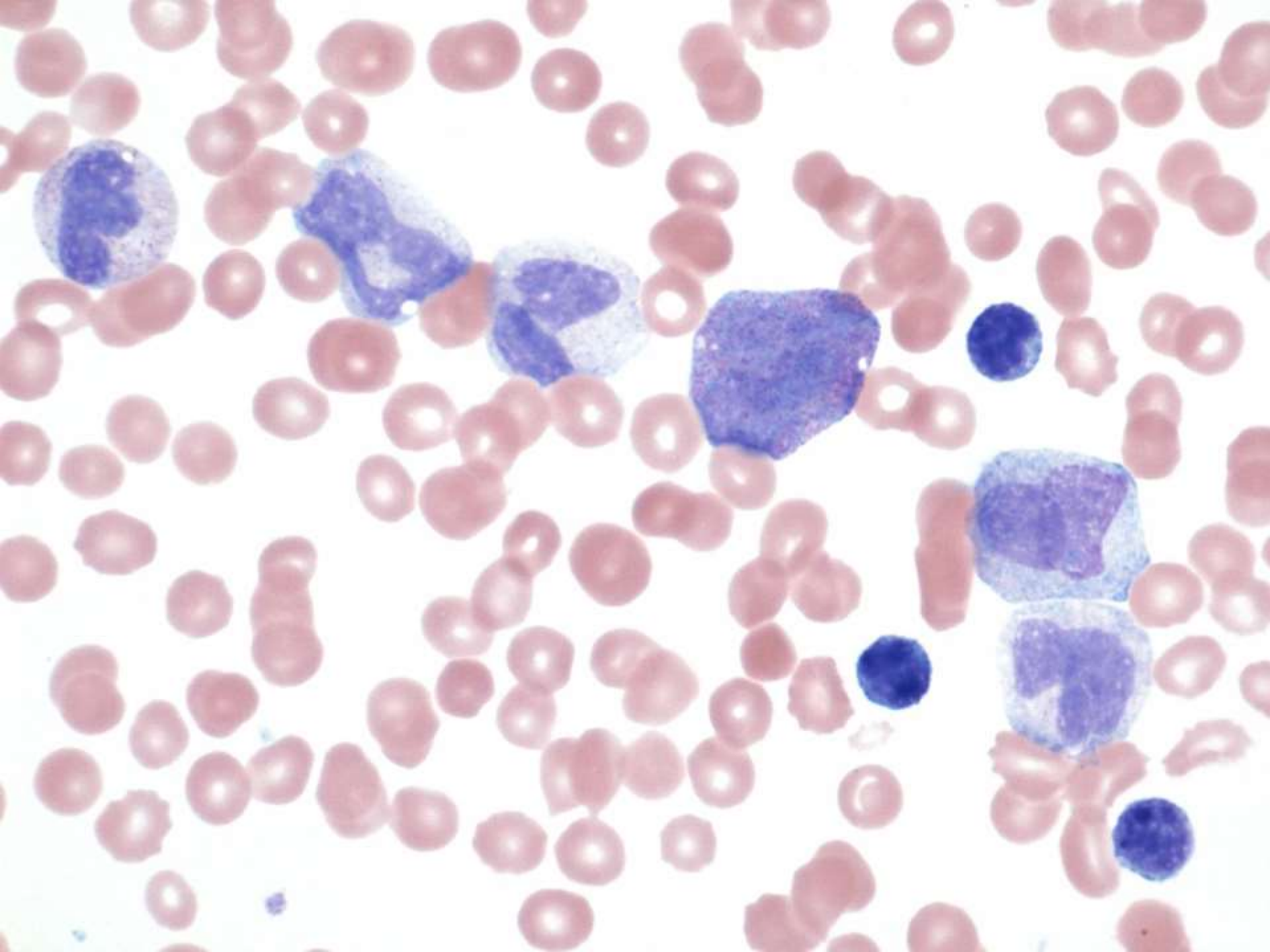
- A 10-month-old boy who was presented with high fever and was found to have anemia and thrombocytopenia with a prominent leukocytosis. CBC: WBC 82.8 x 10E9/L (NEUT 18 x10E9/L, LYMPH 23 x10E9/L, MONO 5.8 x10E9/L), HgB 9.5 g/dL, MCV 81 fl, PLTS 23 x10E9/L, HgF <1.0%

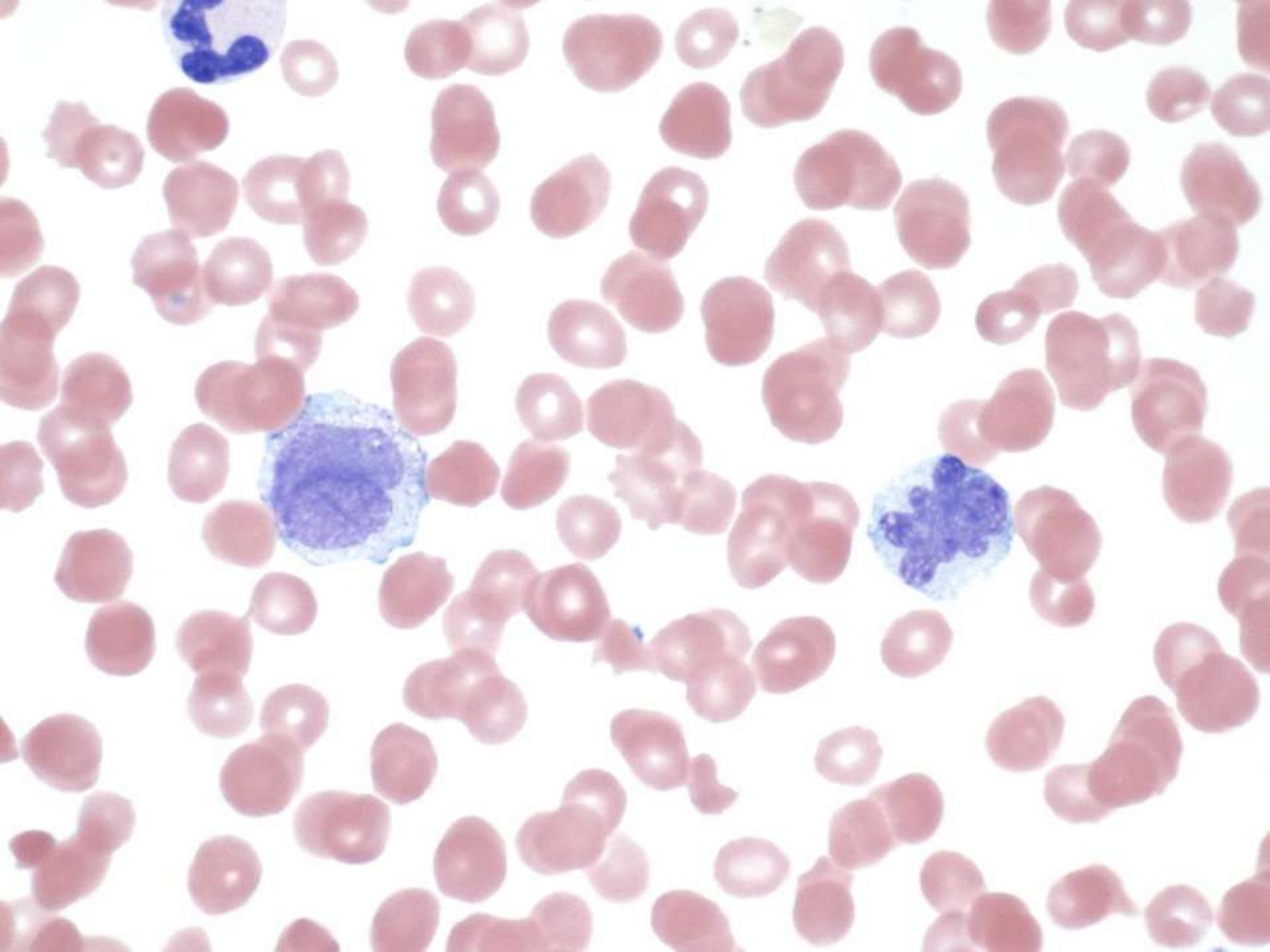
- **Linlin Wang/Sonam Prakash; UCSF**

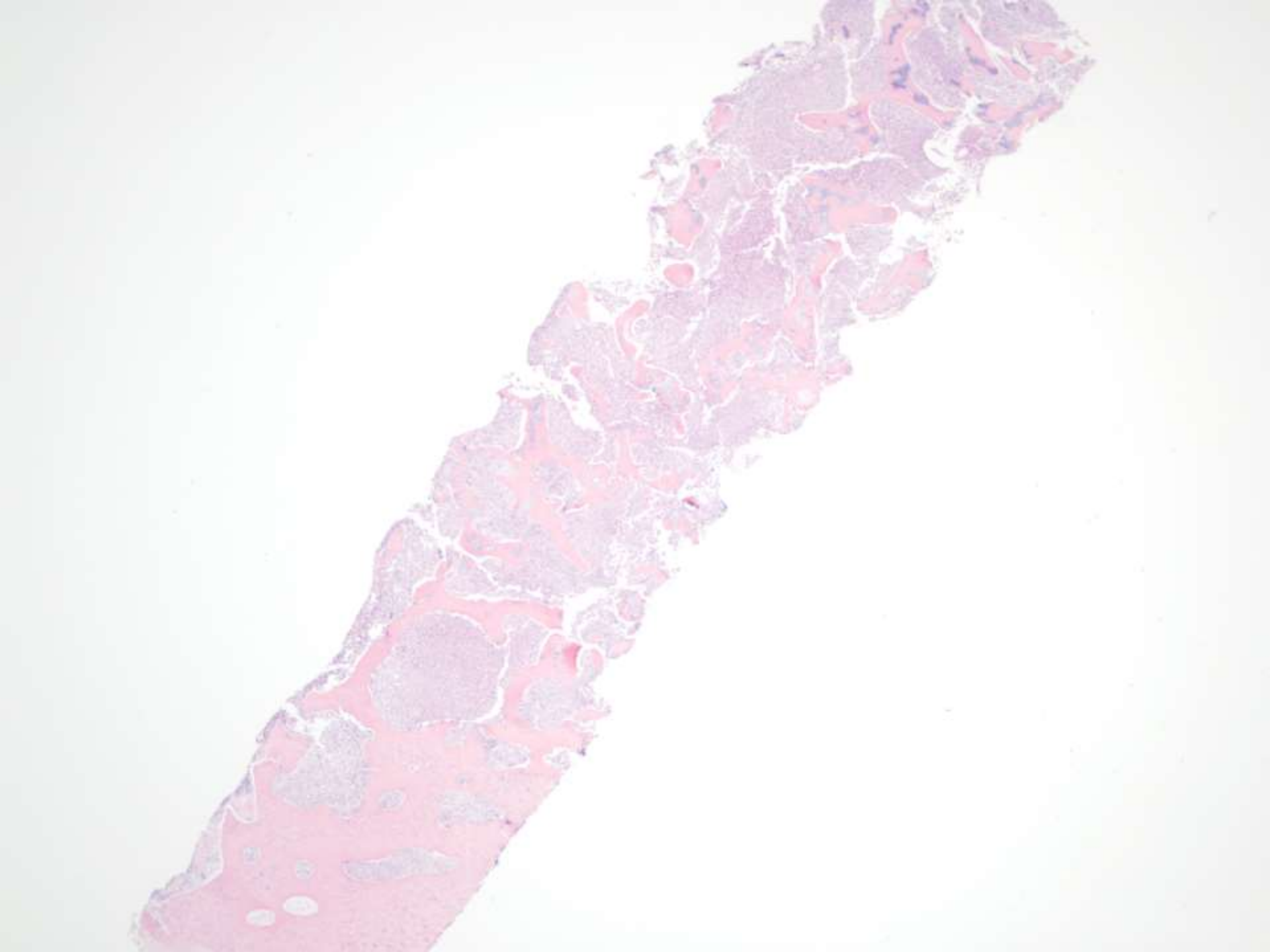


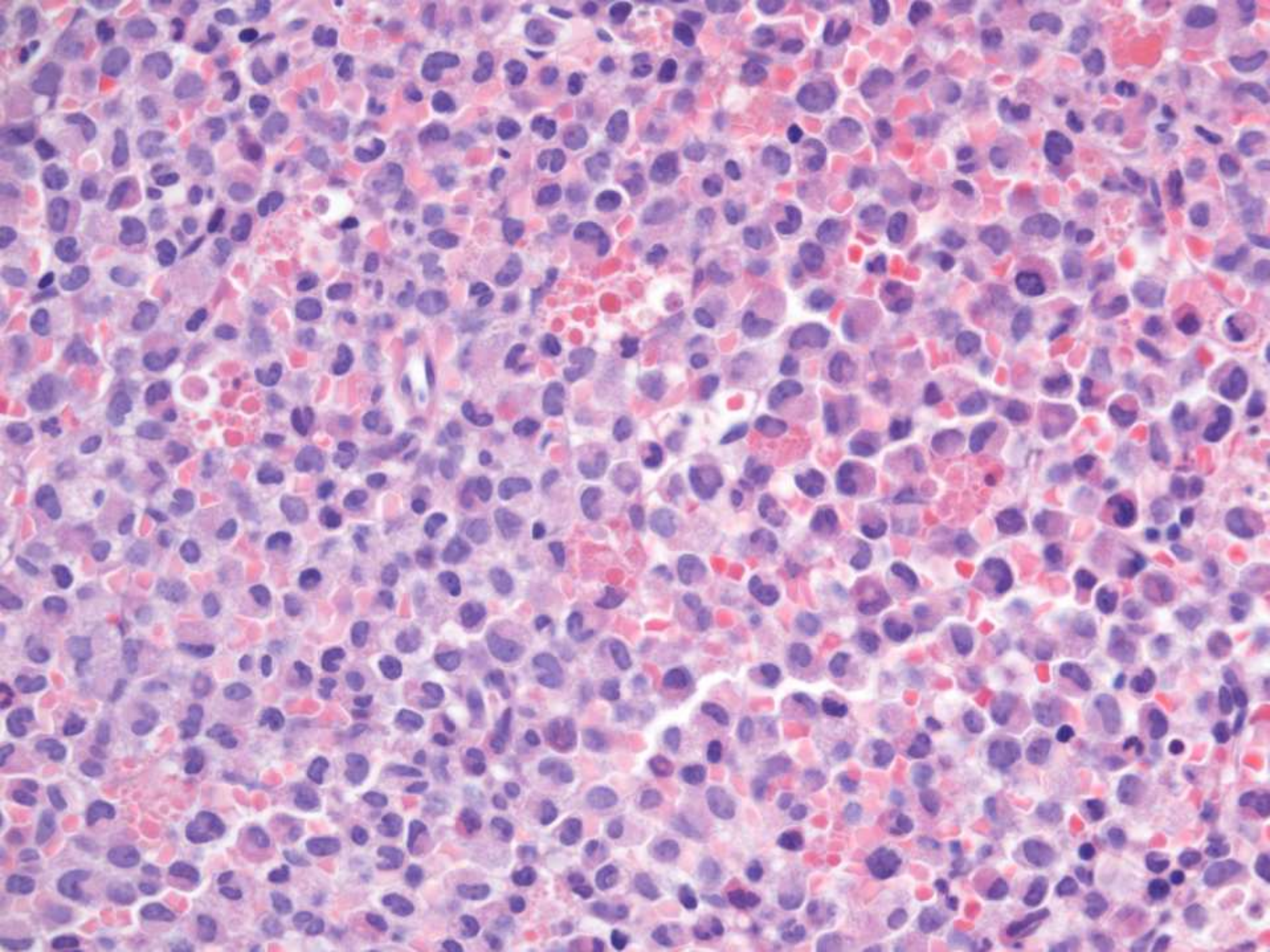




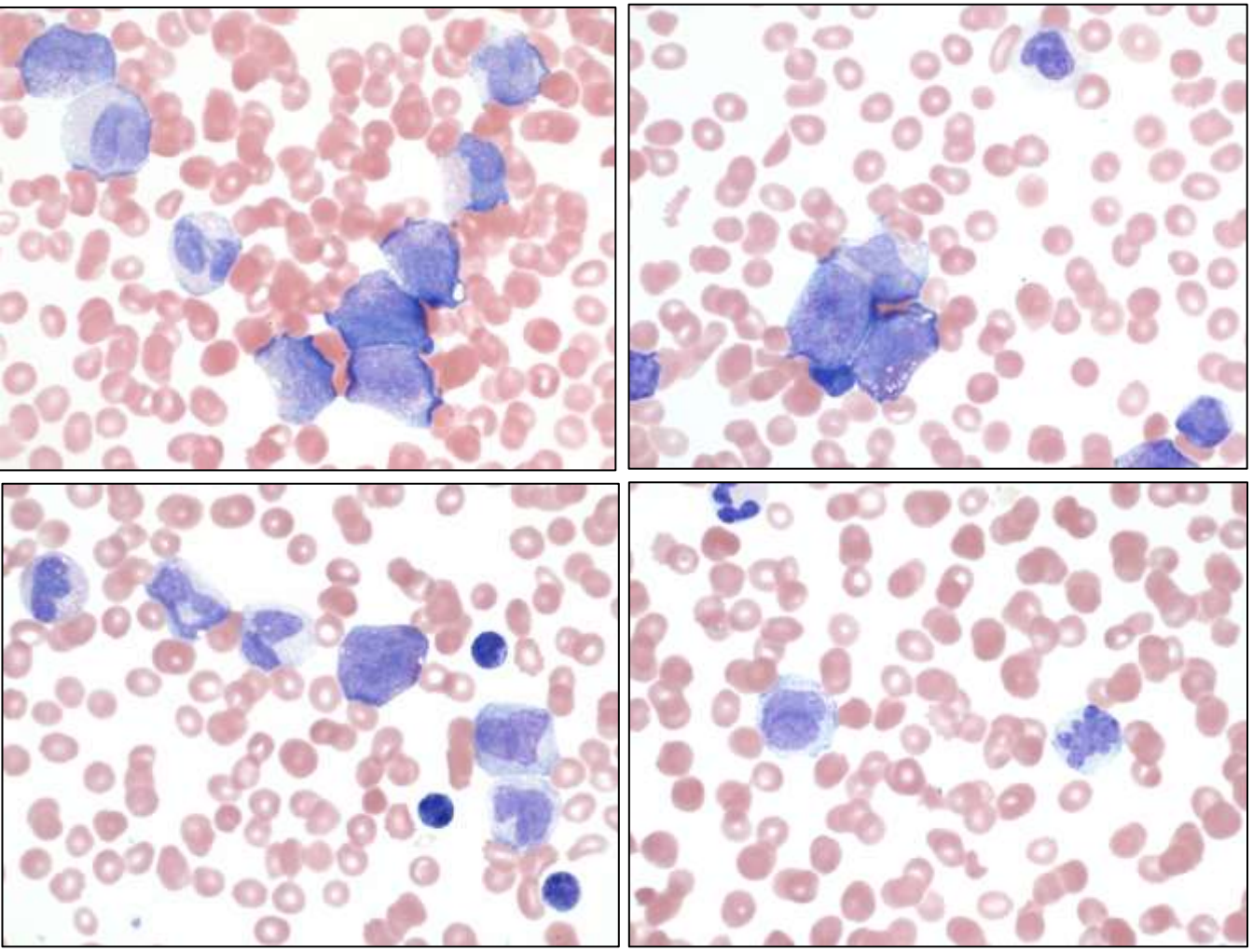


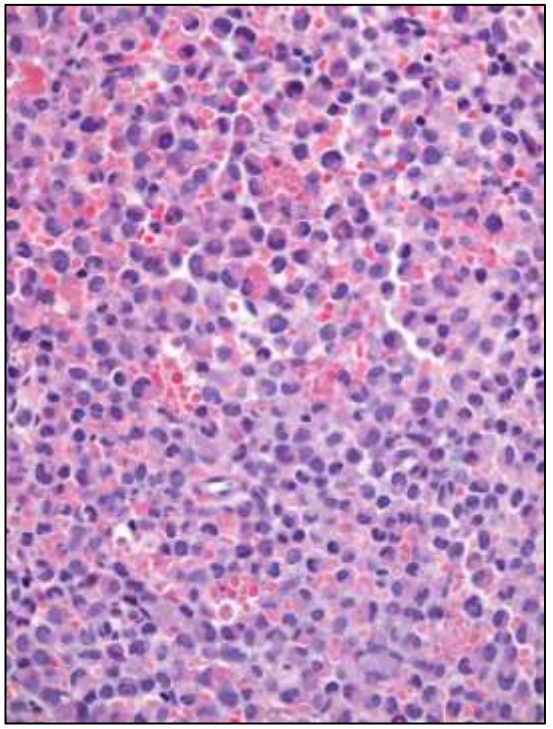






Diagnosis….???





Differential diagnosis:
APL
AML
JMML
Reactive

WBC 82.8 (MONO 5.8), HgB 9.5, PLTS 23

Ancillary Studies

IMMUNOPHENOTYPING:

1. No evidence of a lymphoproliferative disorder.
2. No increase in myeloid blasts (blasts are 0.1% of total events).

CYTOGENETICS: 46,XY[20]

FISH Analysis:

Negative for CHIC2/PDGFRA, PDGFRB, FGFR1, 7q31, PML/RARA, and BCR-ABL gene rearrangements.

Diagnosis Criteria for JMML

Table 4.03 Diagnostic criteria of juvenile myelomonocytic leukaemia*.

1. Peripheral blood monocytosis >1x10⁹/L
2. Blasts (including promonocytes)** are <20% of the leukocytes in the blood and of the nucleated bone marrow cells
3. No Ph chromosome or *BCR-ABL1* fusion gene
4. Plus two or more of the following:
   - Haemoglobin F increased for age
   - Immature granulocytes in the peripheral blood
   - WBC count >10x10⁹/L
   - Clonal chromosomal abnormality (may be monosomy 7)
   - GM-CSF hypersensitivity of myeloid progenitors *in vitro*

*Modified from {1596}

**In this classification, promonocytes are equivalent to blasts.

Category 1	Category 2	Category 3
ALL of the following	**At least 1 of the following**	**At least 2 of the following**
Splenomegaly*	Somatic mutation in *RAS* or *PTPN11*	Circulating myeloid precursors
AMC >1000/uL	Clinical diagnosis of NF1 or *NF1* gene mutation	WBC >10,000/uL
Blasts in PB/BM < 20%	Homozygous mutation in *CBL*	Increased Hgb F for age
Absence of the t(9;22) or *BCR/ABL* fusion gene	Monosomy 7	Clonal cytogenetic abnormality
		GM-CSF hypersensitivity

**For the 7-10% of patients without splenomegaly, the diagnostic criteria must include all other features in category I AND one of the parameters in category II OR*
No features in category II but three features from category 3, including a clonal cytogenetic abnormality.

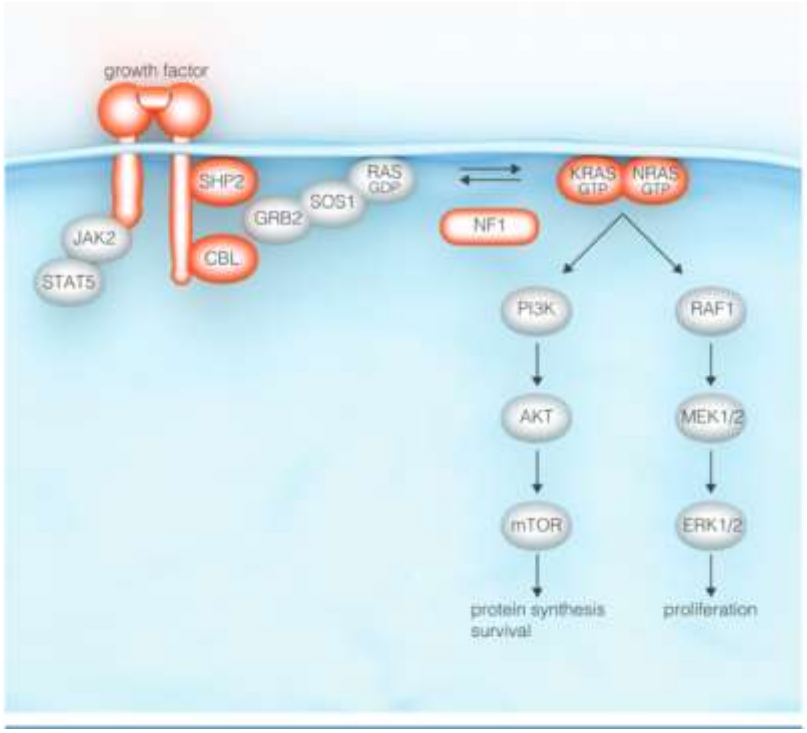
Haematologica 2010; 95: pp 179-82

Diagnosis: JMML

- Positive for KRAS c.35G>T (p. Gly12Val) mutation.
- Features are unusual for JMML:
  - Relatively mild increase in monocytic forms
  - Predominant dysplastic immature myeloid elements in the peripheral blood
  - Mild hepatosplenomegaly
  - Lack of elevation of hemoglobin F

JMML

- WHO classification: Mixed MPN/MDS of young children
- Median age at diagnosis 1.7 years, Boys > girls
- Splenomegaly, fever, thrombocytosis, monocytosis, dysplastic myeloid cells, increased HbF
- Poor prognosis
- Treatment: HSCT
- 90% JMML patients have largely mutually exclusive mutations in one the 5 genes: PTPN11 (35%), NRAS/KRAS (20-25%), NF1 (11%), CBL (12%)



Blood Spotlight

JMML and RALD (Ras-associated autoimmune leukoproliferative disorder): common genetic etiology yet clinically distinct entities

Katherine R. Calvo,[1] Susan Price,[2,3] Raul C. Braylan,[1] Joao Bosco Oliveira,[4] Michael Lenardo,[2,3] Thomas A. Fleisher,[1] and V. Koneti Rao[2,3]

[1]Department of Laboratory Medicine, Clinical Center, [2]Laboratory of Immunology, National Institute of Allergy and Infectious Diseases (NIAID), [3]NIAID Clinical Genomics Program, National Institutes of Health, Bethesda, MD; and [4]Instituto de Medicina Integral Prof. Fernando Figueira, Recife, Brazil

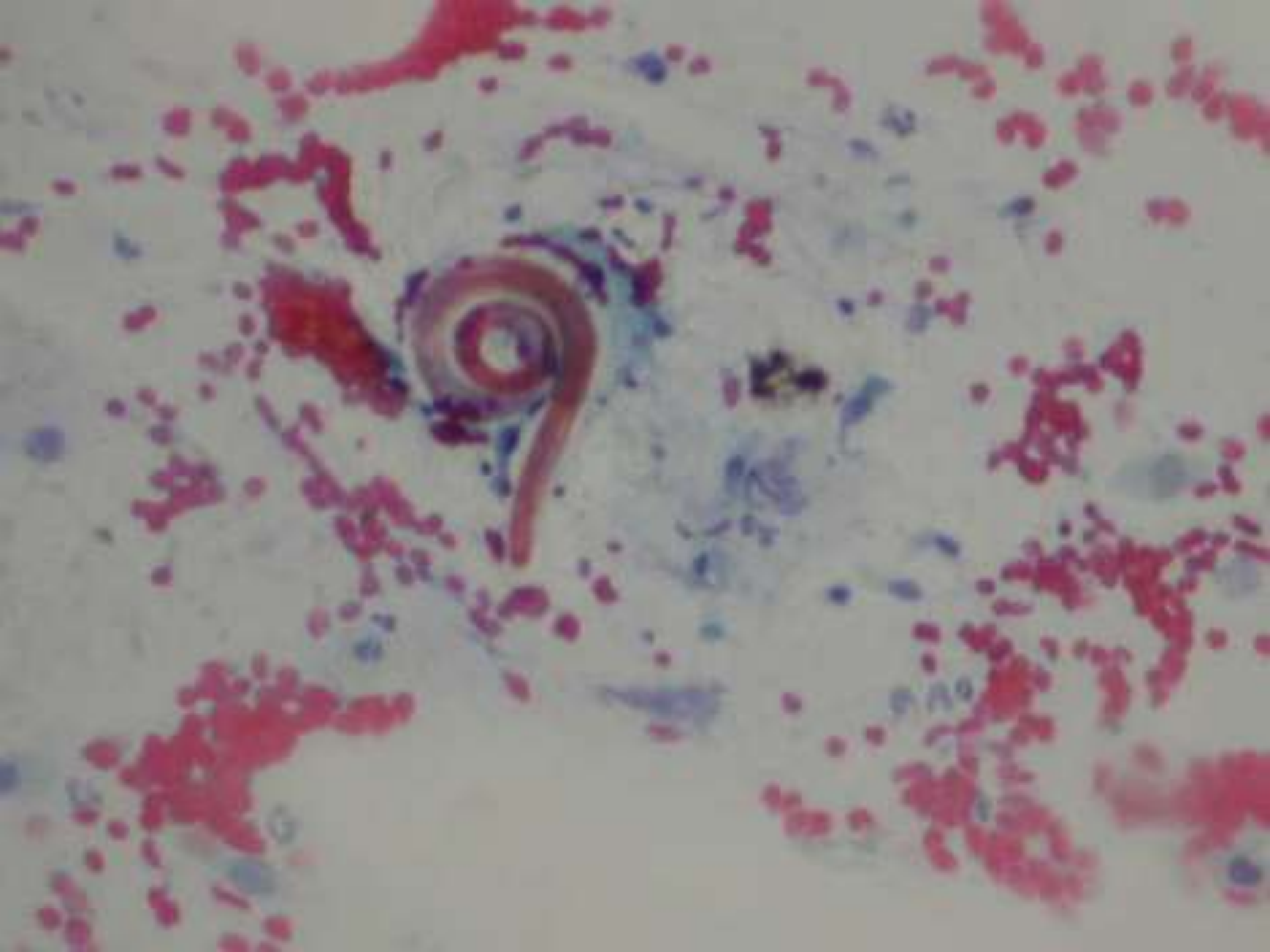
RALD

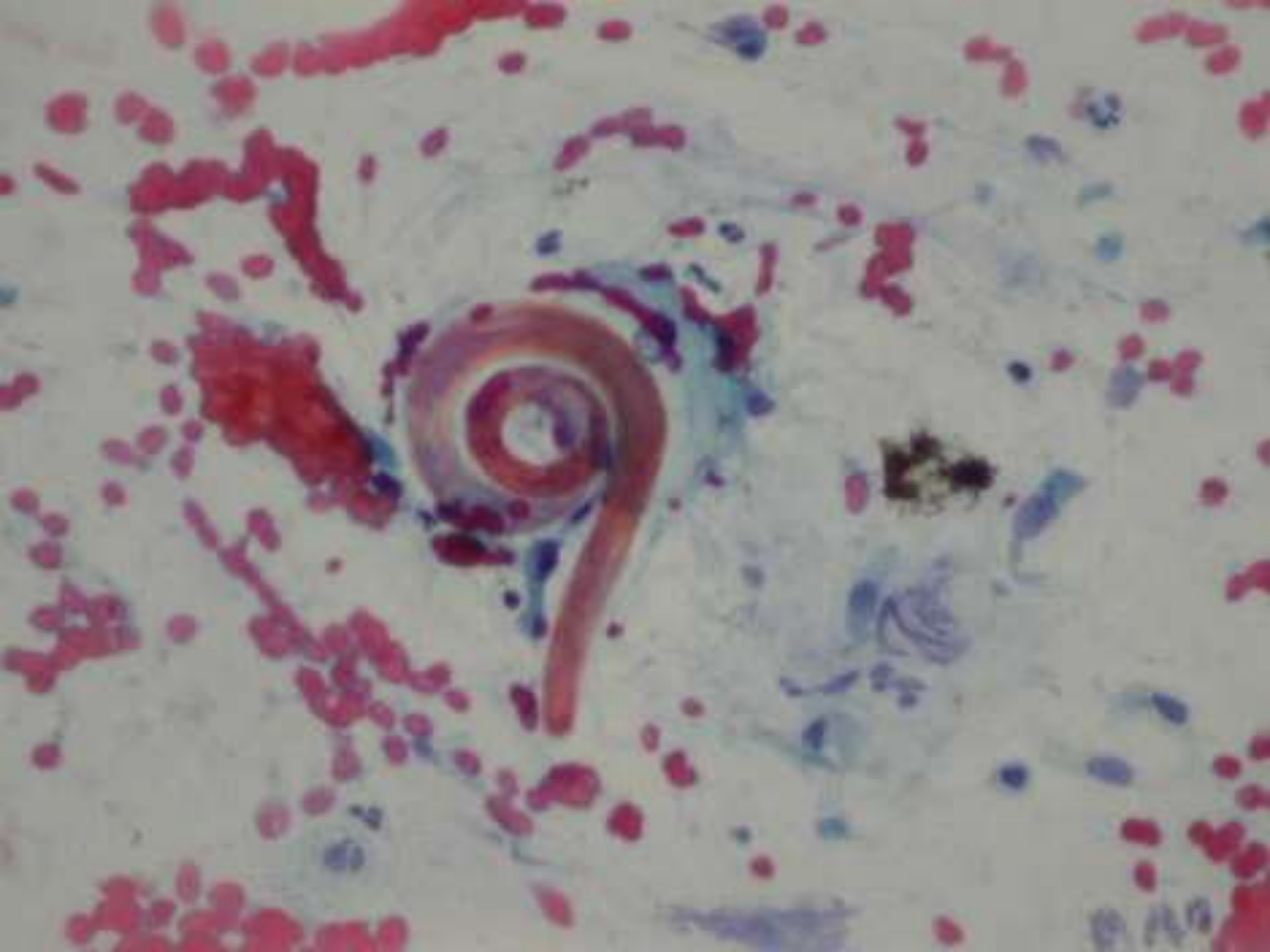
- Persistent monocytosis, leukocytosis, lymphadenopathy, splenomegaly and hypergammaglobulinemia
- Mutation in KRAS and NRAS
- Indolent clinical course

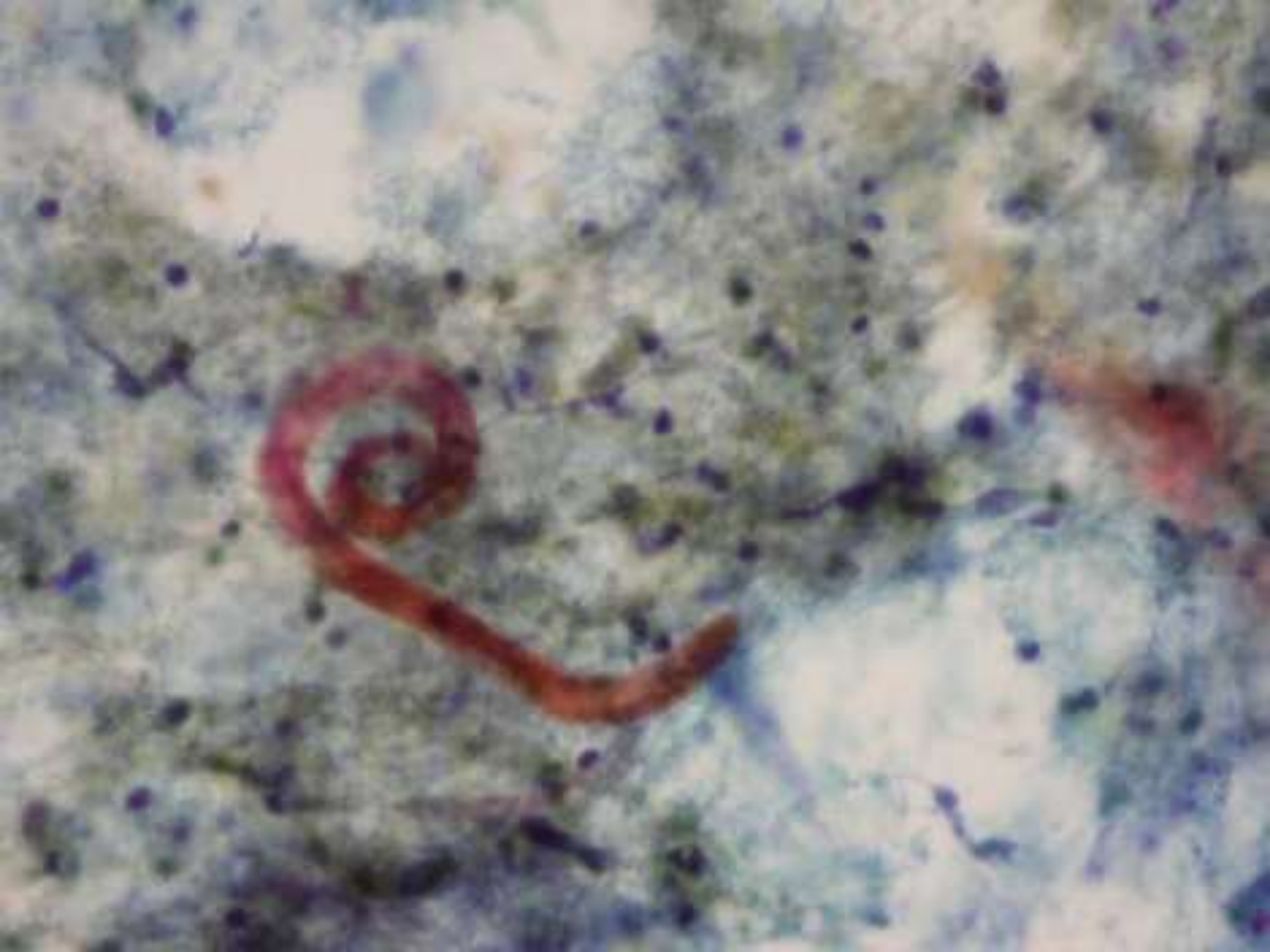
SB 5949

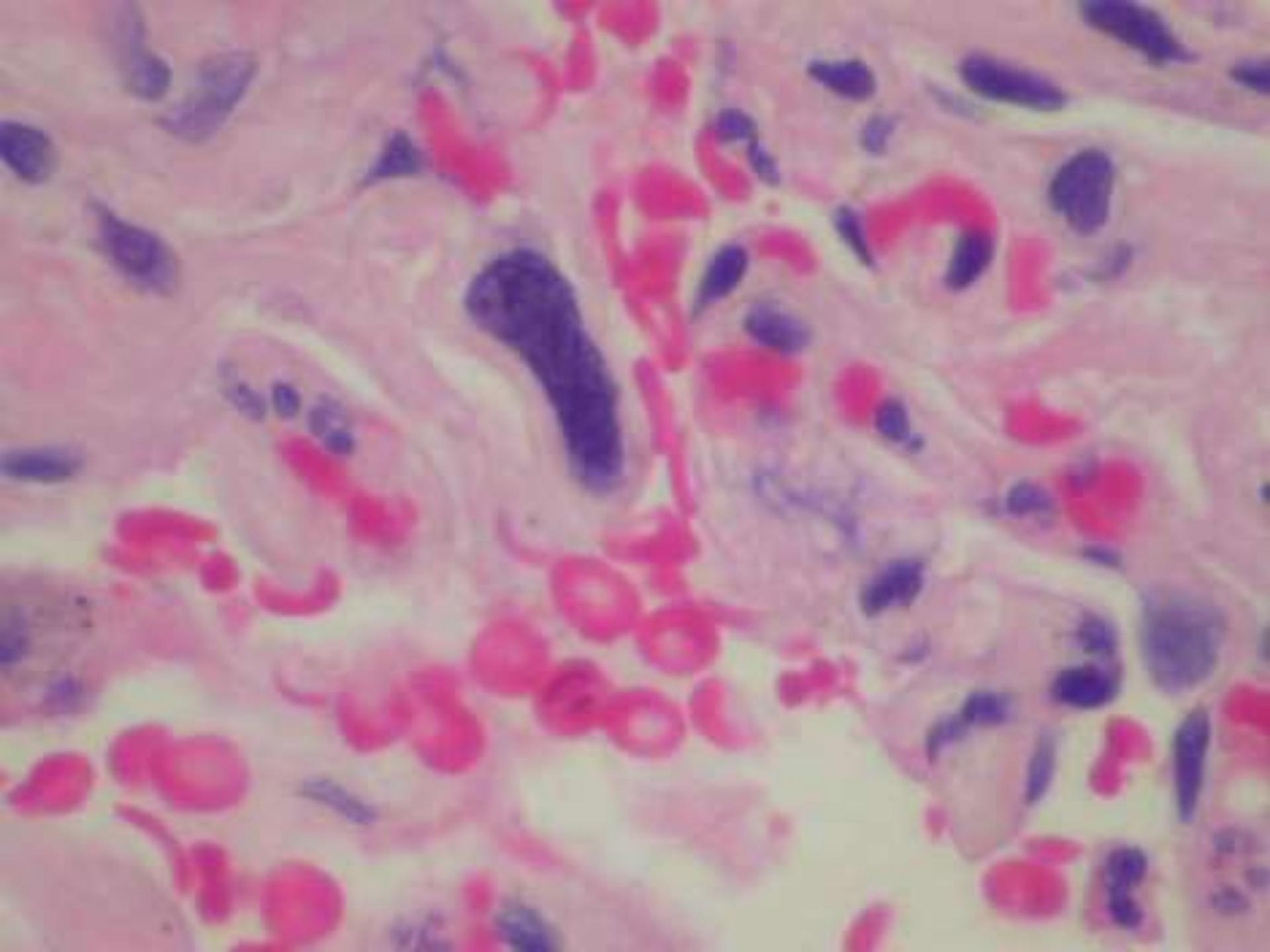
- 82-year-old male with a history of asthmatic bronchitis, hypothyroidism, and diabetes was admitted after presenting with cough and respiratory distress with sputum production. Chest X-ray revealed right pneumothorax. He received broad-spectrum antibiotics and had negative influenza studies. A chest tube was placed for management of the pneumothorax and had VATS procedure and then bronchoscopy with washings.

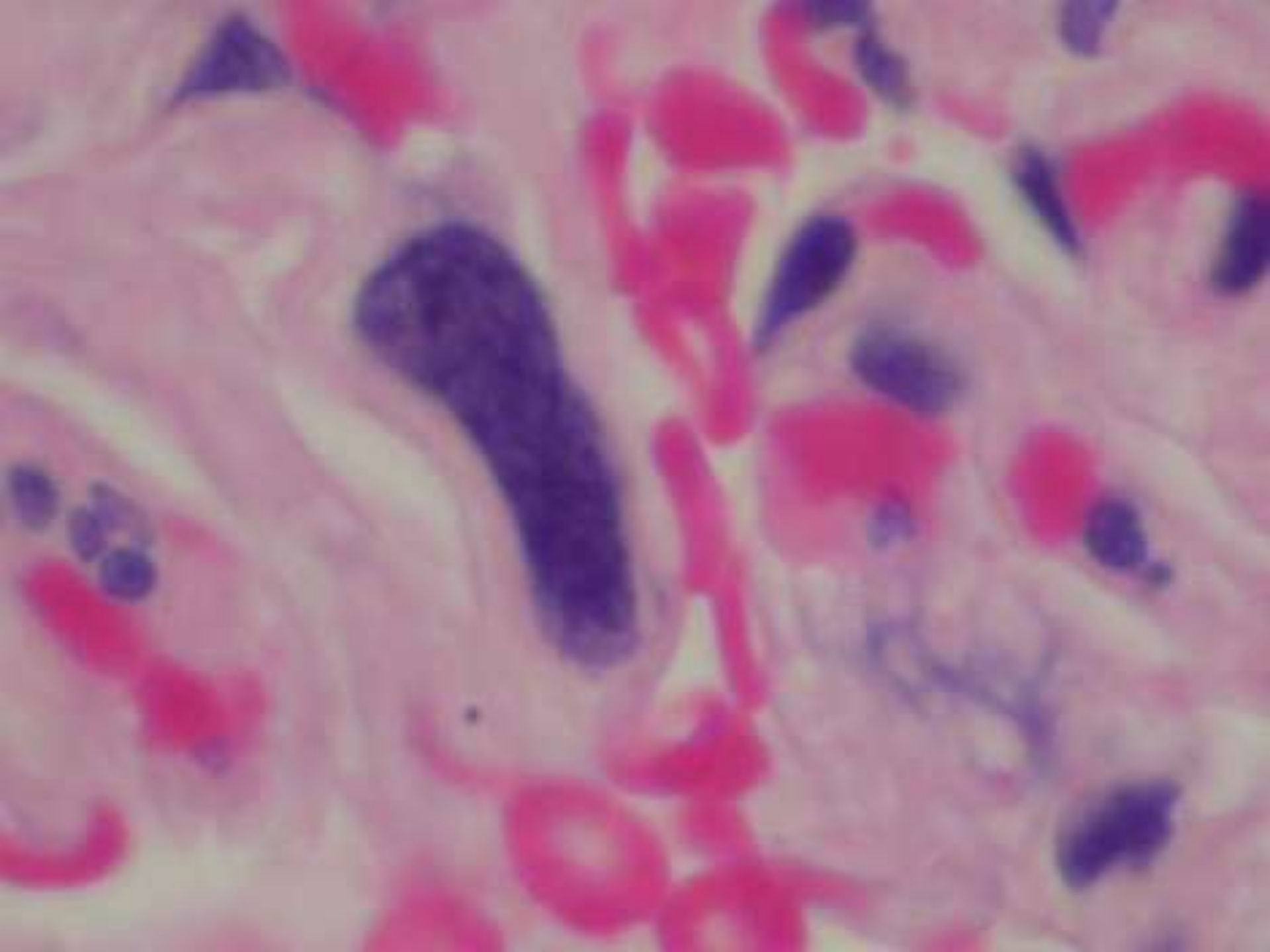
- **Keith Duncan; Mills-Peninsula**











Strongyloides stercoralis

Strongyloidiasis was first described in French troops stationed in modern day Vietnam during the late 19th century who were suffering from severe, persistent diarrhea.

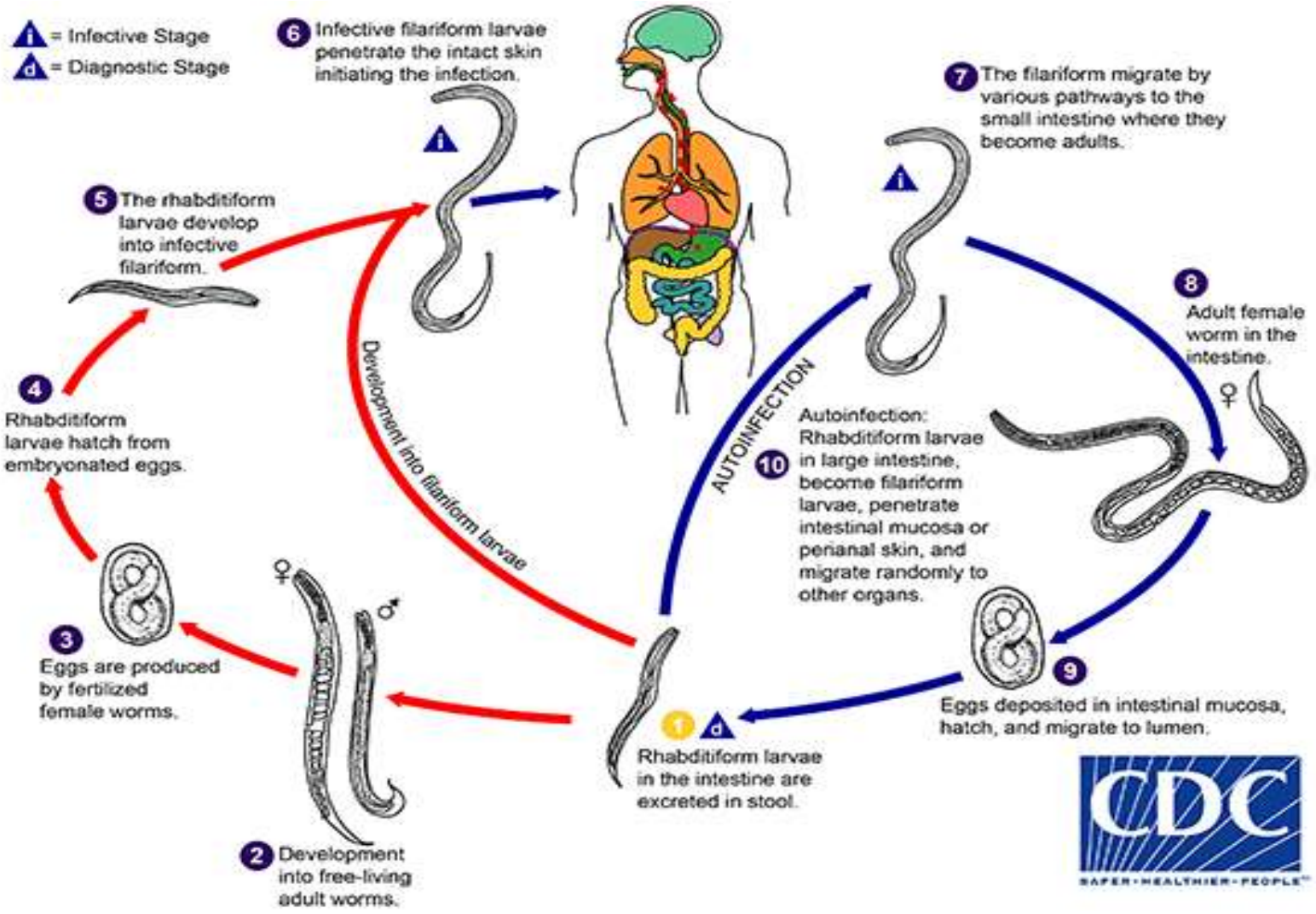
Parasitic disease caused by nematodes, or roundworms (2 mm long), in the genus *Strongyloides* that enter the body through exposed skin (bare feet).

Unlike most parasitic worms, *Strongyloides* has a heterogonic life cycle (has a separate free-living cycle where it lives and reproduces without a host in the soil).

***M*ost common in tropical or subtropical climates.**

Most people who are infected with *Strongyloides* are asymptomatic; but may develop a severe form and, if untreated, become critically ill.

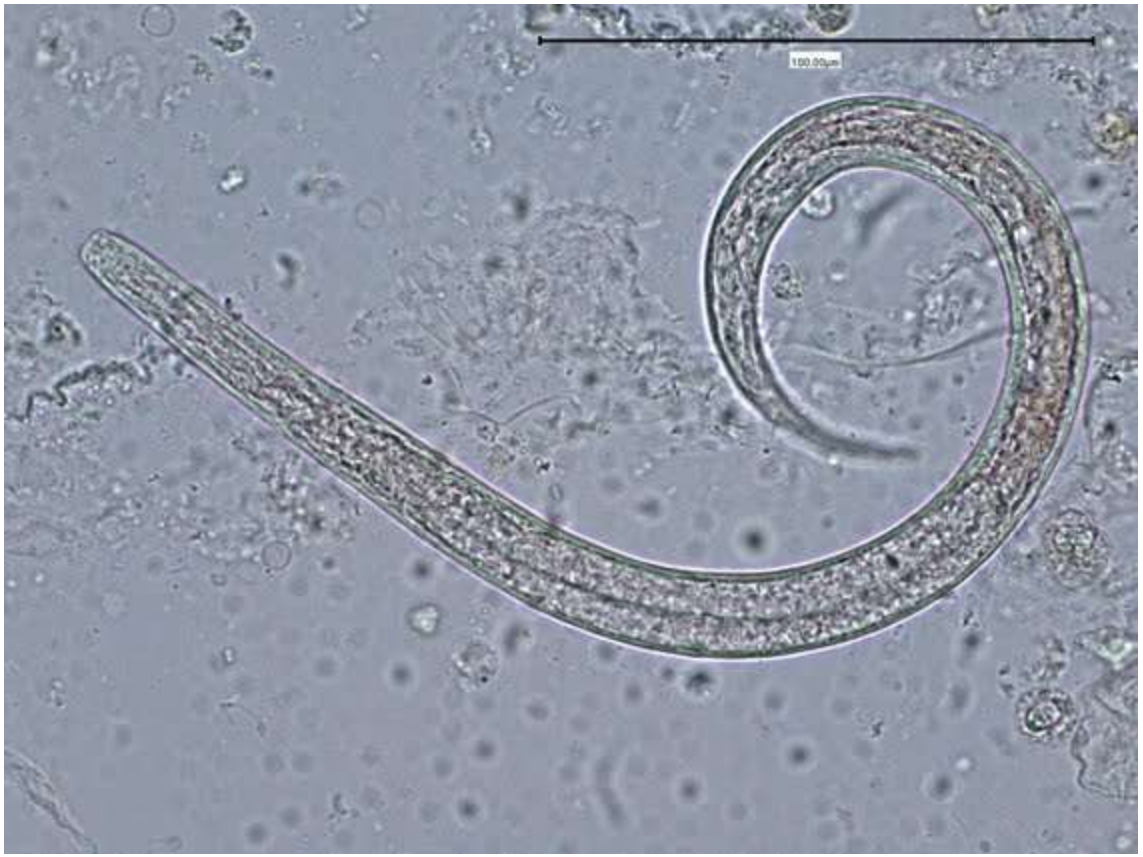
LIFE CYCLE

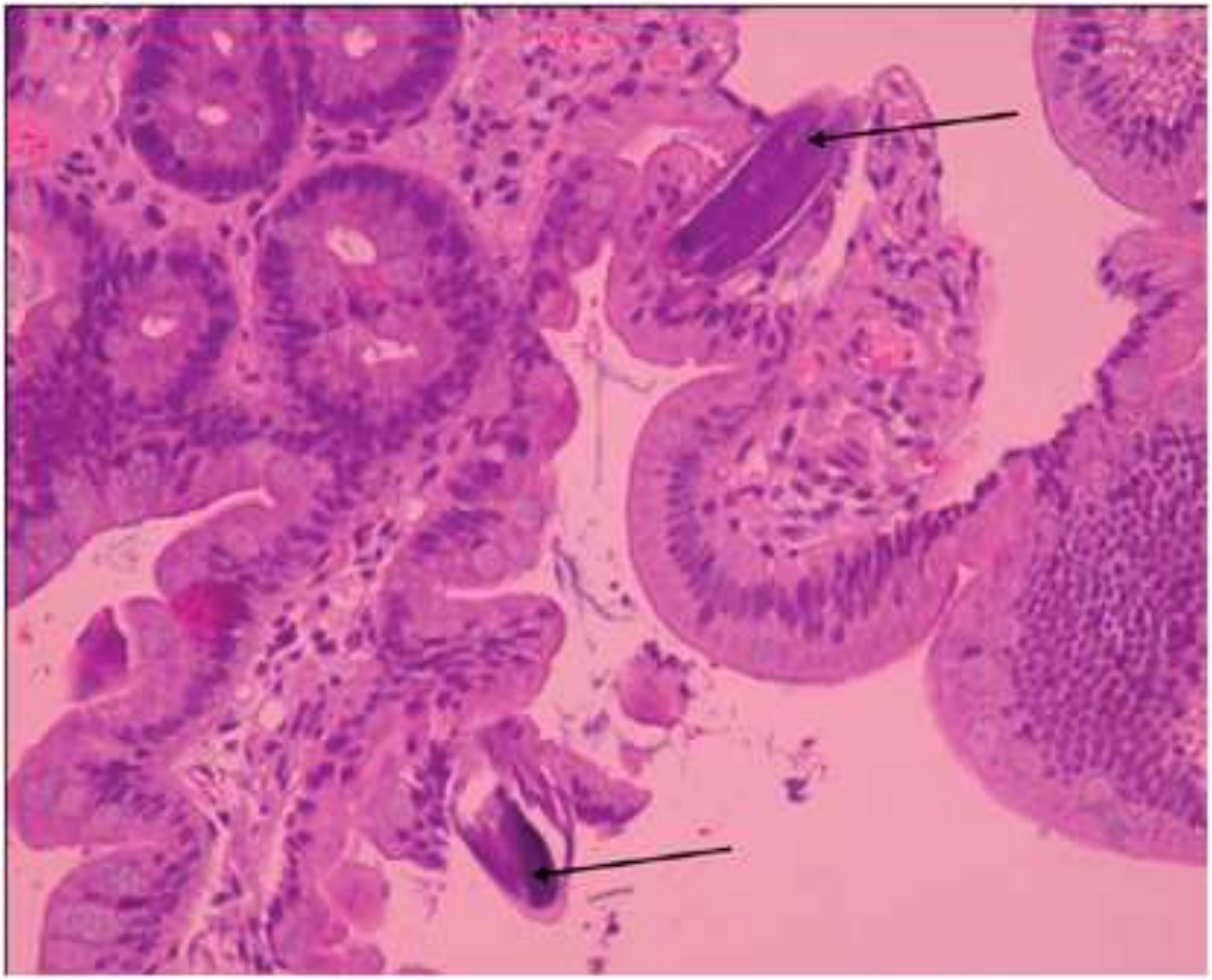

i = Infective Stage
d = Diagnostic Stage
1 d Rhabditiform larvae in the intestine are excreted in stool.
2 Development into free-living adult worms.
3 Eggs are produced by fertilized female worms.
4 Rhabditiform larvae hatch from embryonated eggs.
5 The rhabditiform larvae develop into infective filariform.
6 Infective filariform larvae penetrate the intact skin initiating the infection.
7 The filariform migrate by various pathways to the small intestine where they become adults.
8 Adult female worm in the intestine.
9 Eggs deposited in intestinal mucosa, hatch, and migrate to lumen.
10 Autoinfection: Rhabditiform larvae in large intestine, become filariform larvae, penetrate intestinal mucosa or perianal skin, and migrate randomly to other organs.
Development into filariform larvae
AUTOINFECTION
CDC
SAFER·HEALTHIER·PEOPLE™

Diagnosis

Locating juvenile larvae, either rhabditiform or filariform, in recent stool samples will confirm the presence of this parasite.

Other techniques used include direct fecal smears, culturing fecal samples on agar plates, serodiagnosis through ELISA, and duodenal fumigation. Still, diagnosis can be difficult because of the varying juvenile parasite load on a daily basis.


100.00µm



Diagn Cytopathol. 2009 Dec;37(12):903-5. doi: 10.1002/dc.21125.

Strongyloides stercoralis in a bronchial washing specimen processed as conventional and Thin-Prep smears: report of a case and a review of the literature.

Grapsa D[1], Petrakakou E, Botsoli-Stergiou E, Mikou P, Athanassiadou P, Karkampasi A, Ioakim-Liossi A.

Author information

[1]Cytopathology Department, LAIKO Athens General Hospital, Medical School, University of Athens, Athens, Greece. dimgrap@yahoo.gr

Abstract

Strongyloidiasis is an opportunistic infection which may result in a fatal hyperinfection syndrome in immunocompromised patients. We report the case of a pulmonary infection with Strongyloides stercoralis in a 61-year-old male with a history of a long-term administration of corticosteroids. Cytologic examination of a bronchial washing specimen, processed both as conventional and as Thin-Prep smears, revealed an abundance of the typical larvae of Strongyloides stercoralis, amidst a cellular population comprising several acute inflammatory cells as well as bronchial epithelial cells with features of basal cell hyperplasia or regenerative atypia. To the best of our knowledge there is only one previous report describing Strongyloides stercoralis in thin-layer smears, and there are no previous studies comparing its morphology in conventional and thin-layer preparations.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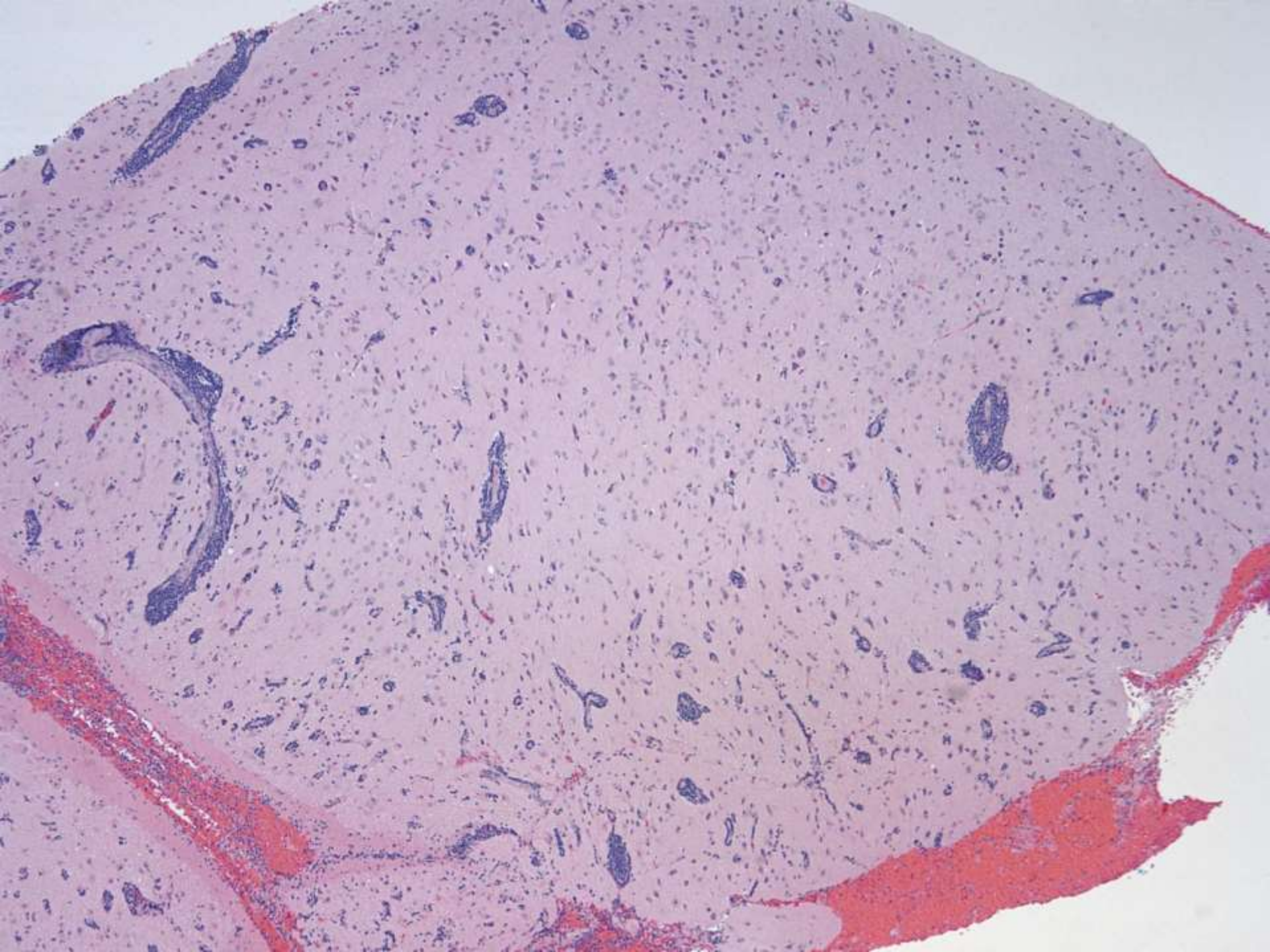
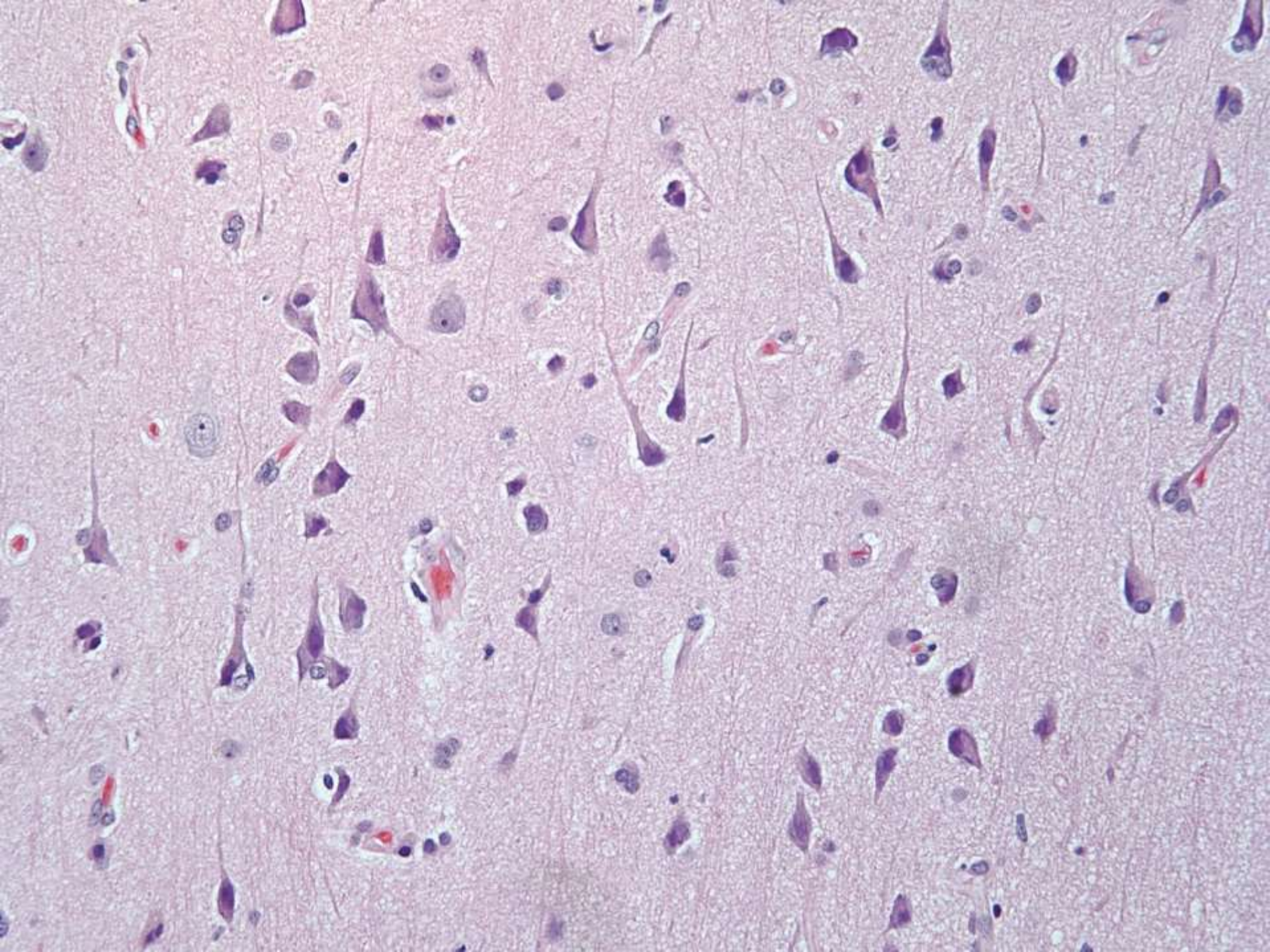
www.martybucella.com
"Would walking into McDonald's instead of using the drive-thru be considered more exercise?"

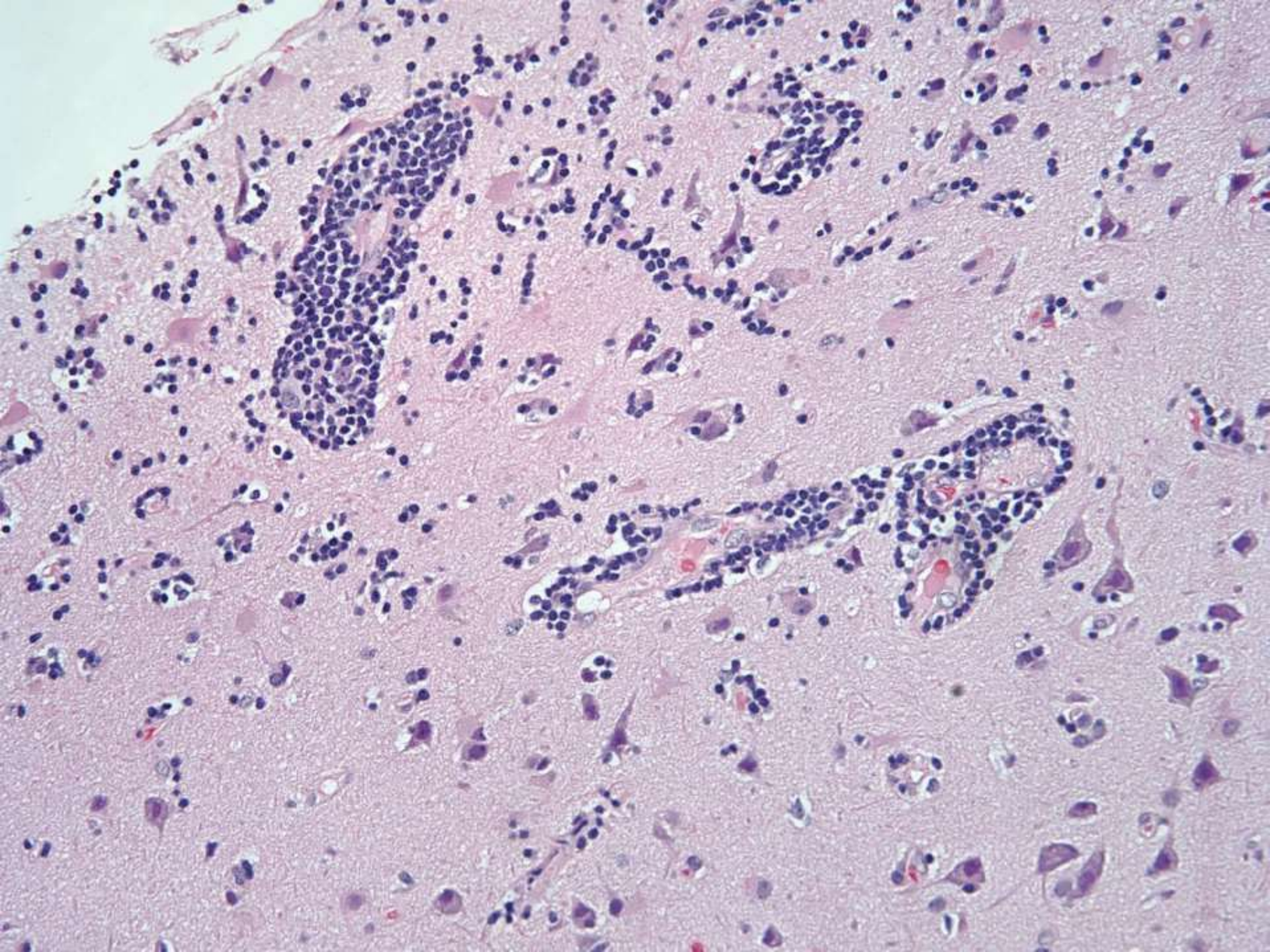
SB 5950

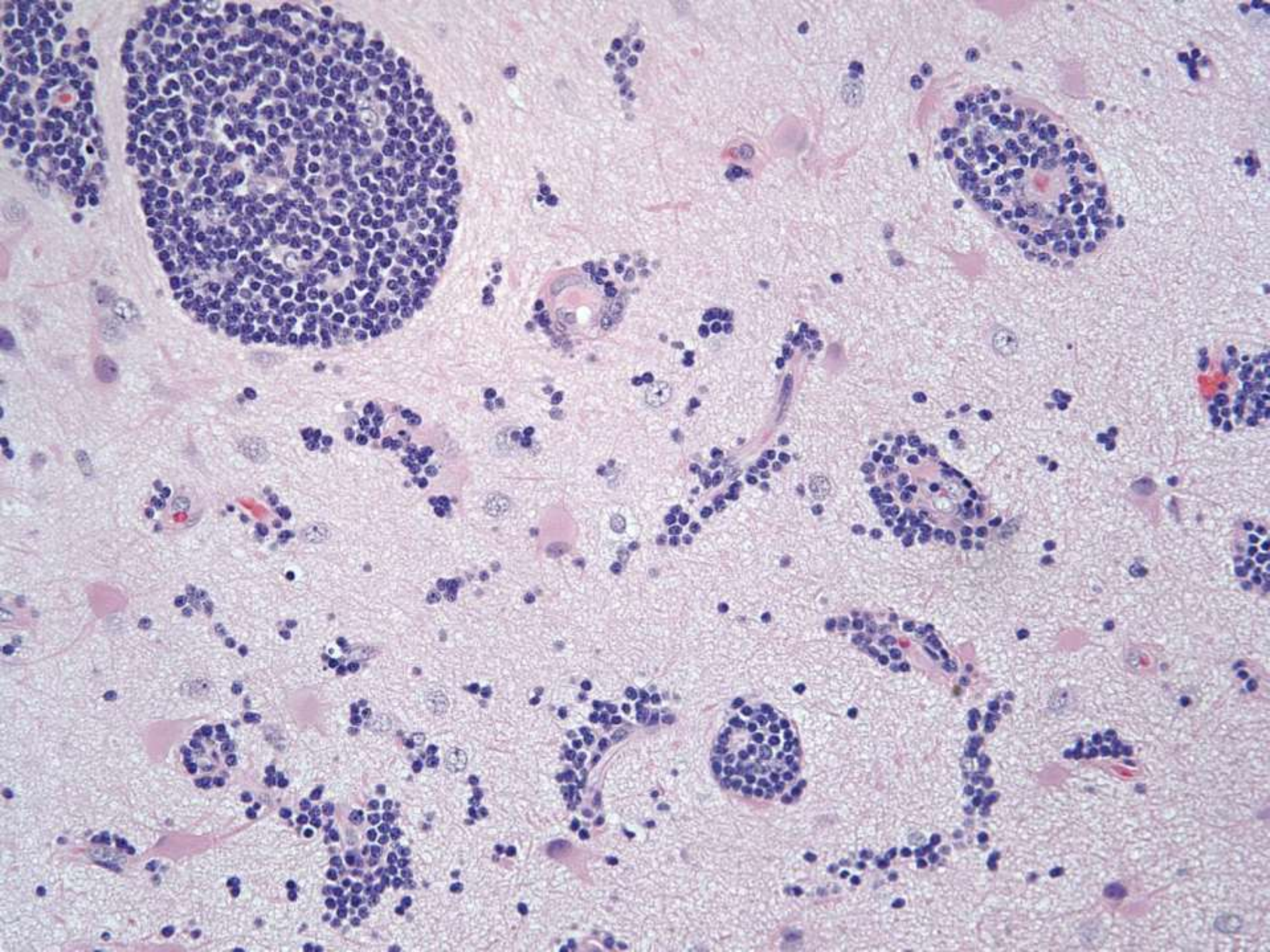
- 44-year-old male with right frontal subdural empyema.

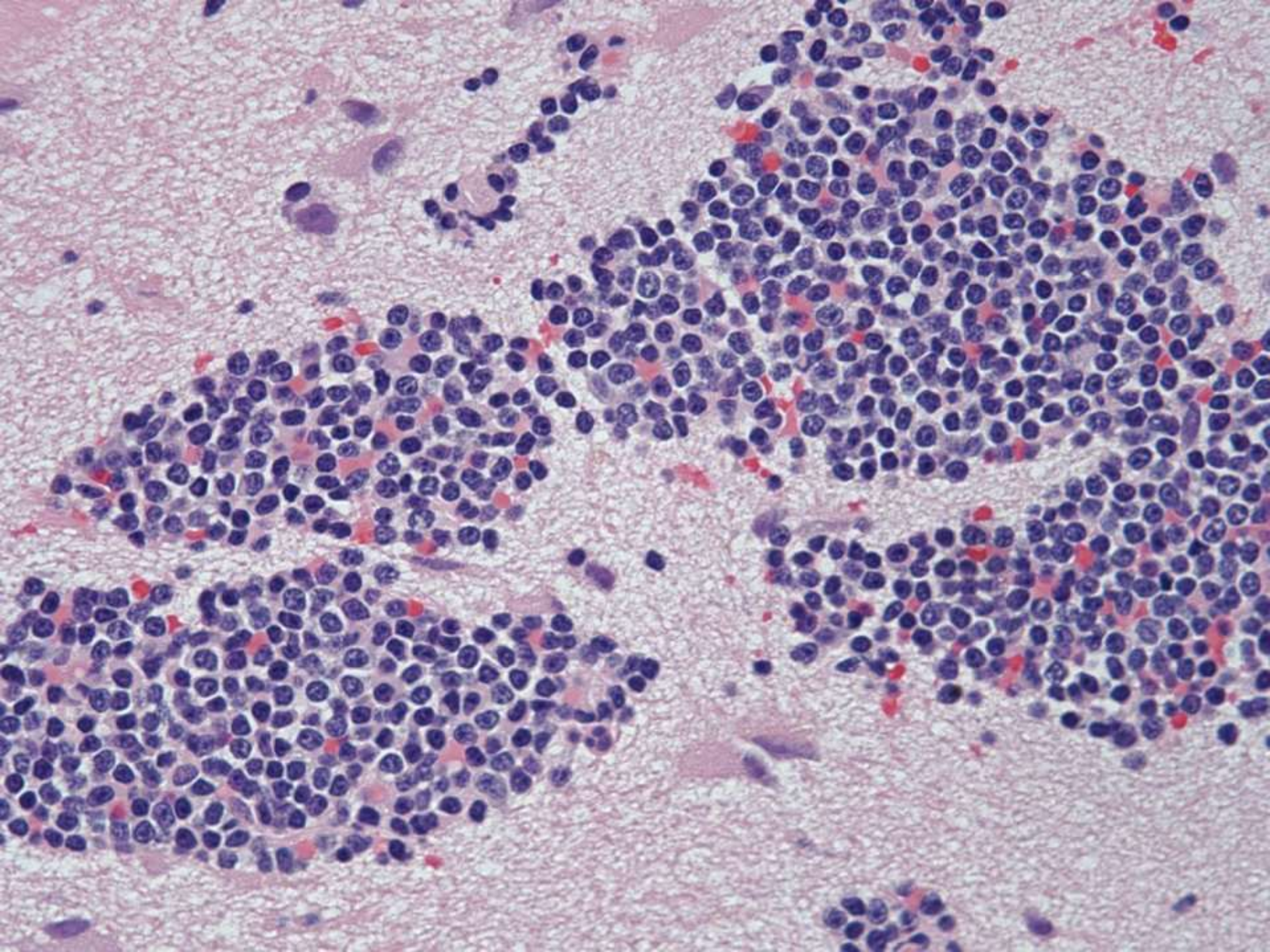
- **Chieh-Yu Lin/Hannes Vogel; Stanford**







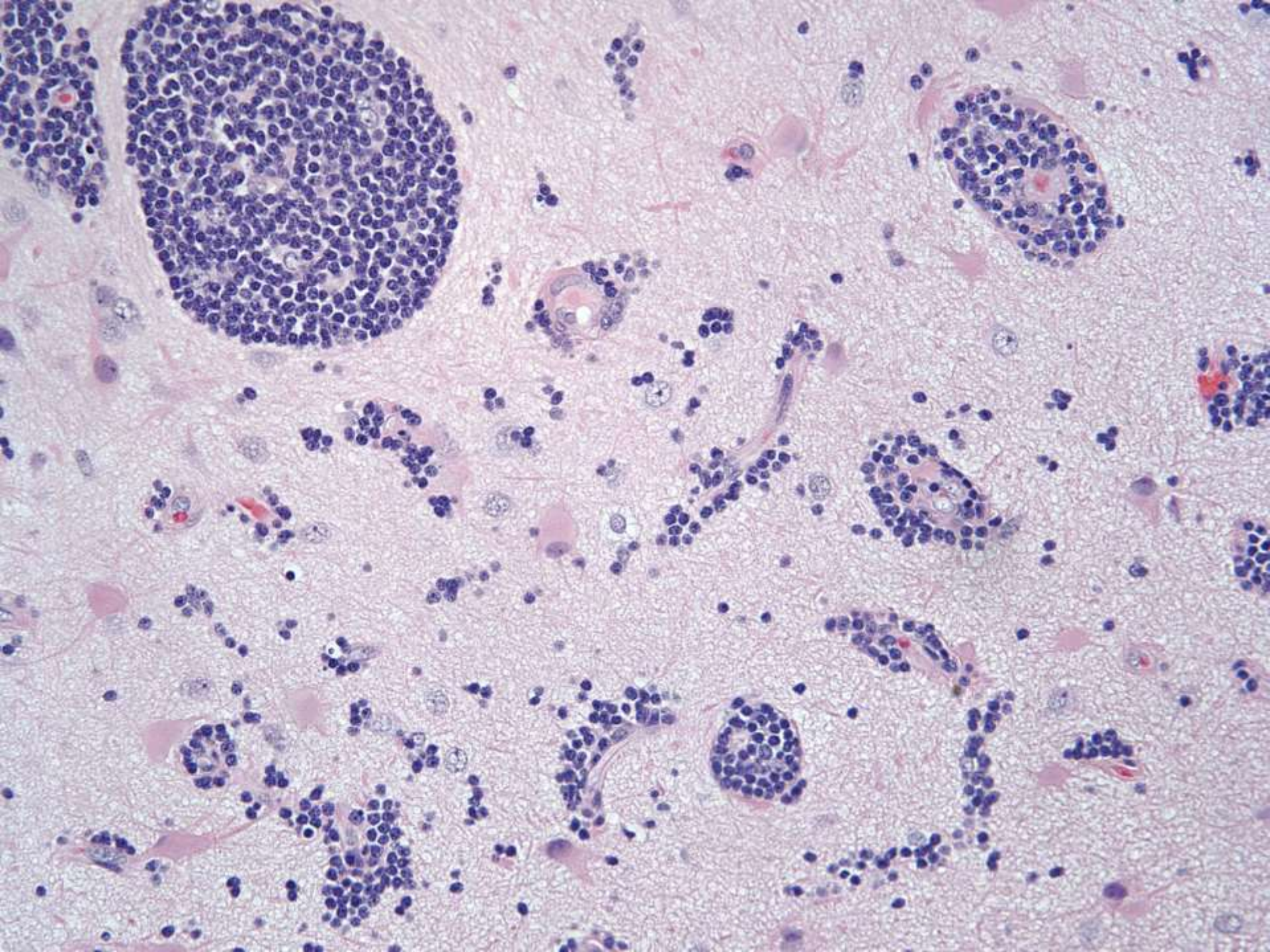


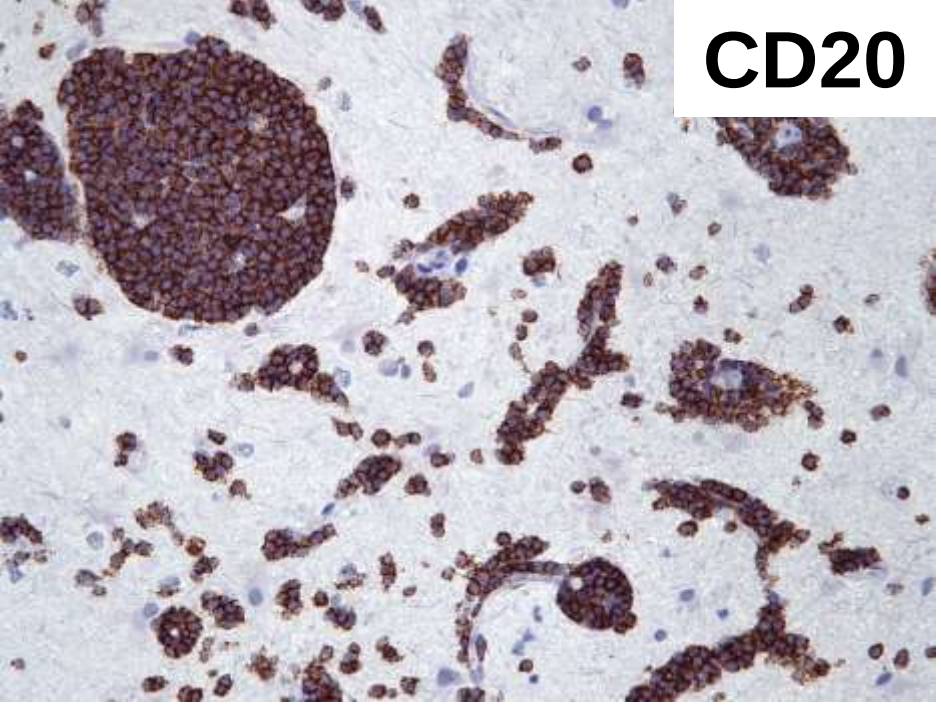


Diagnosis….???

Brief history and imaging findings

- Headache and left-side weakness
- MRI findings:
  - 1cm peripheral and ringed enhancement with vasogenic edema and subdural fluid collection; can’t exclude infection
- Clinical impression: neurocysticercosis

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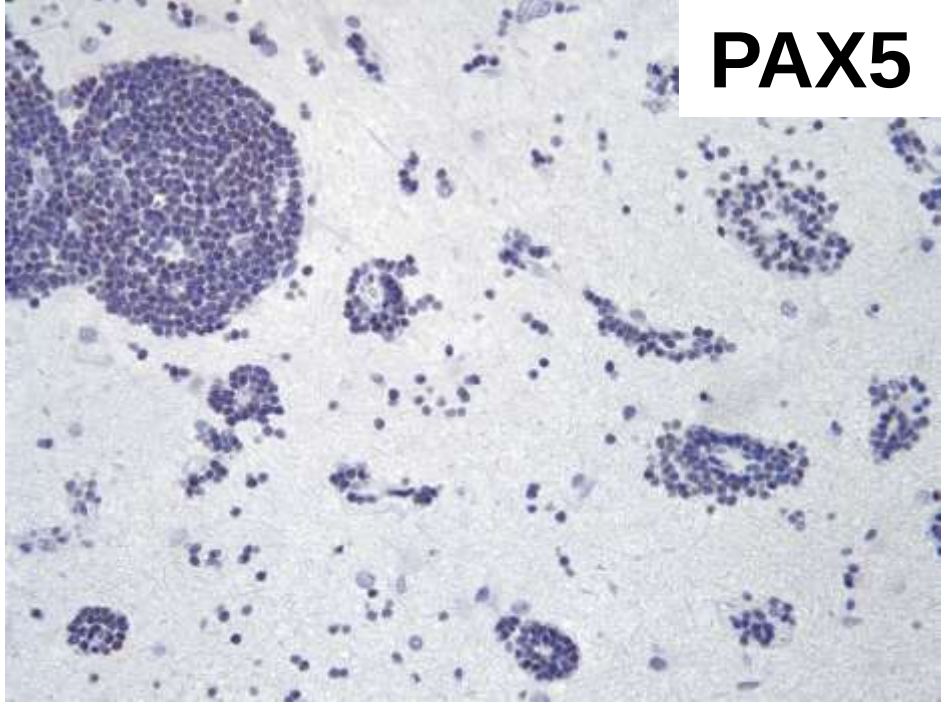
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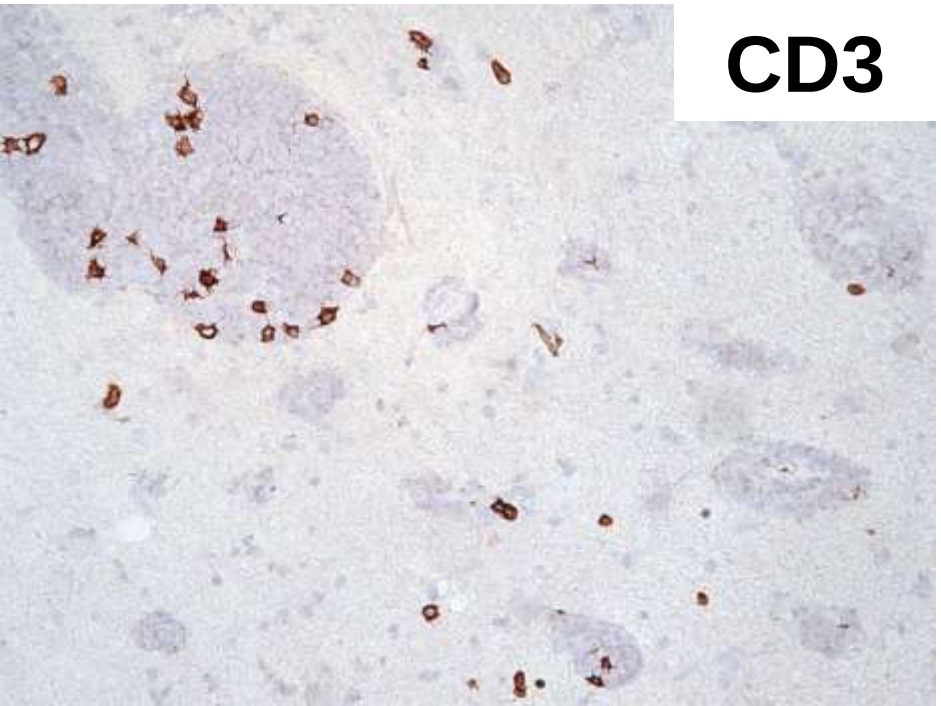
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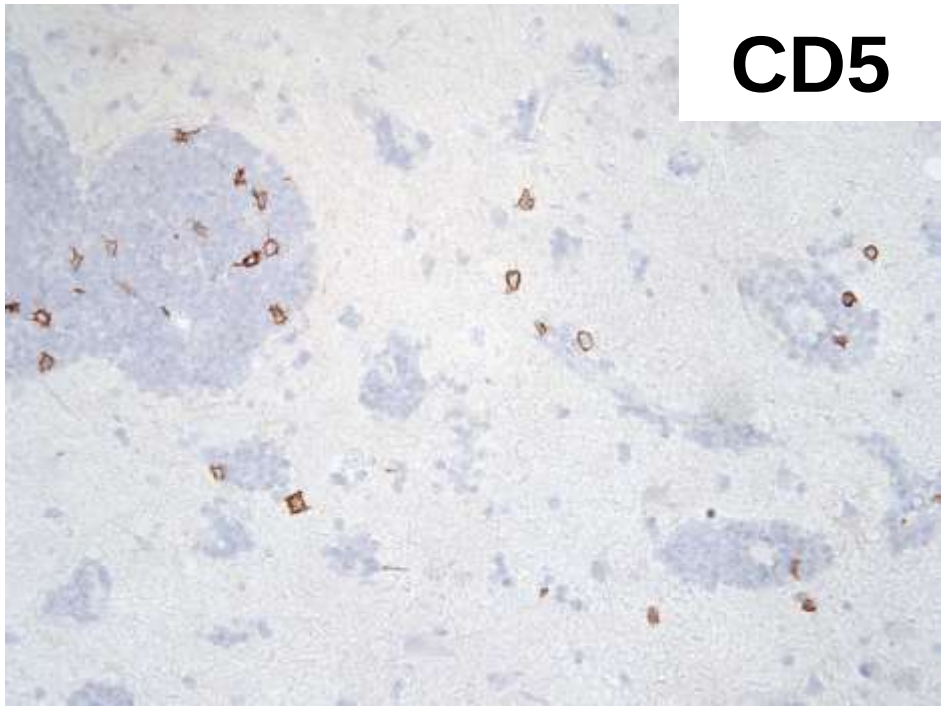
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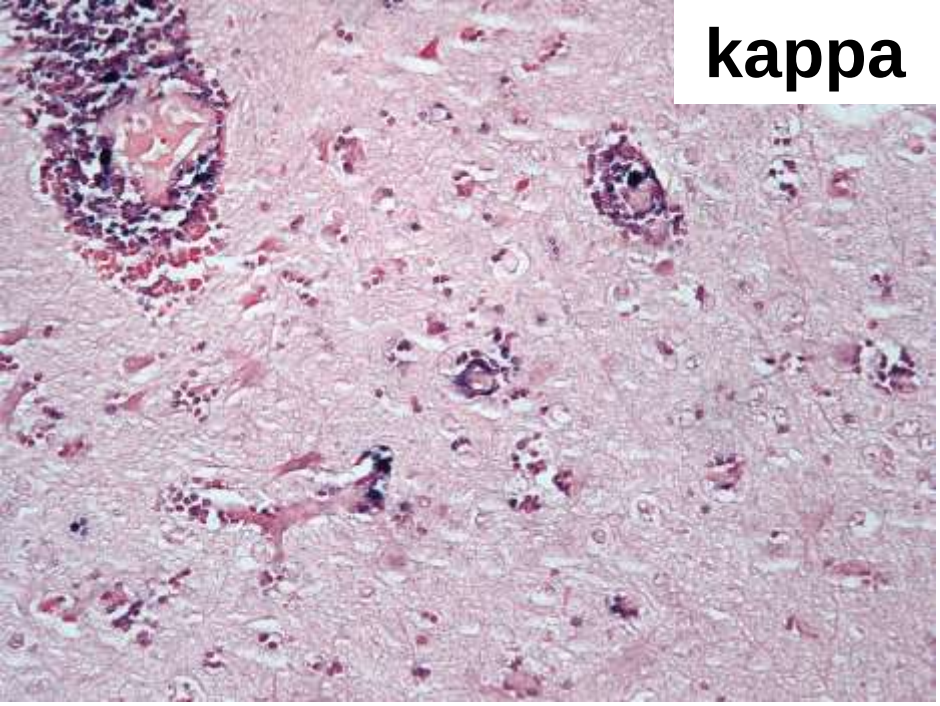
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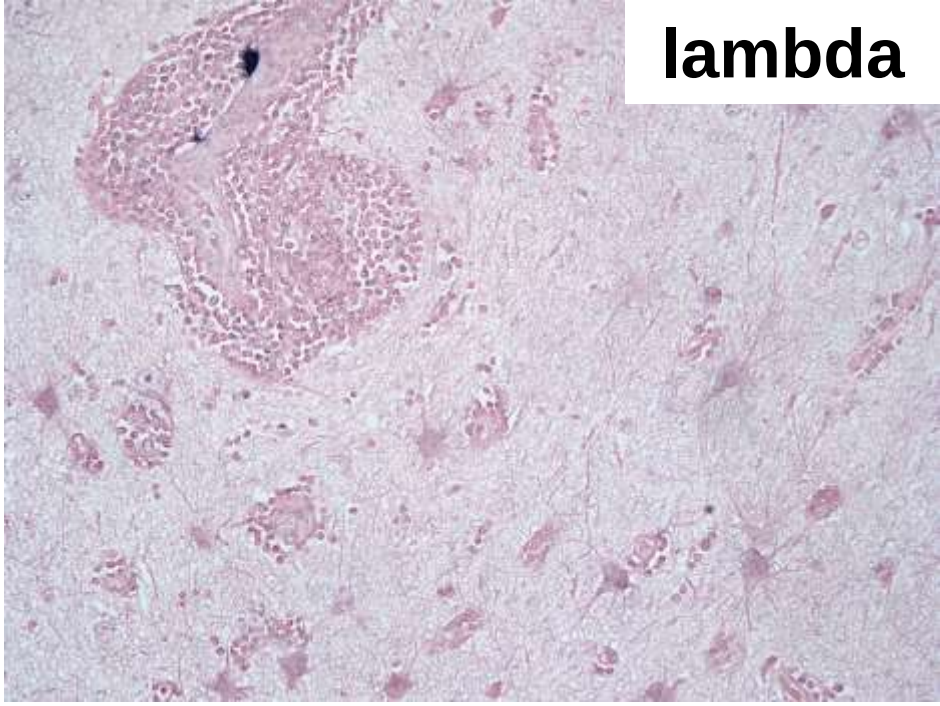
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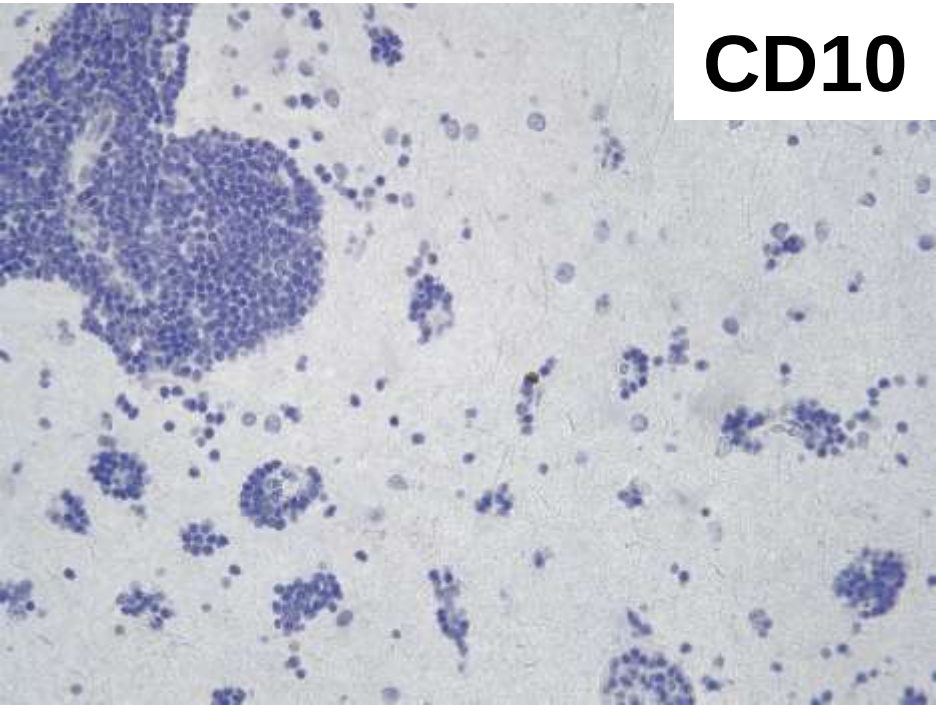
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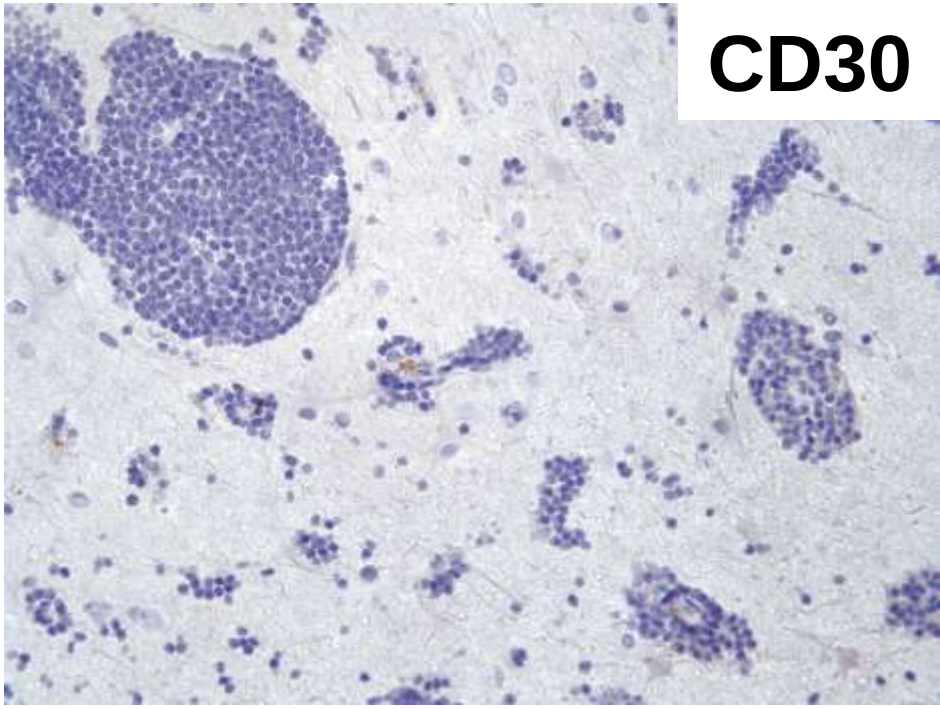
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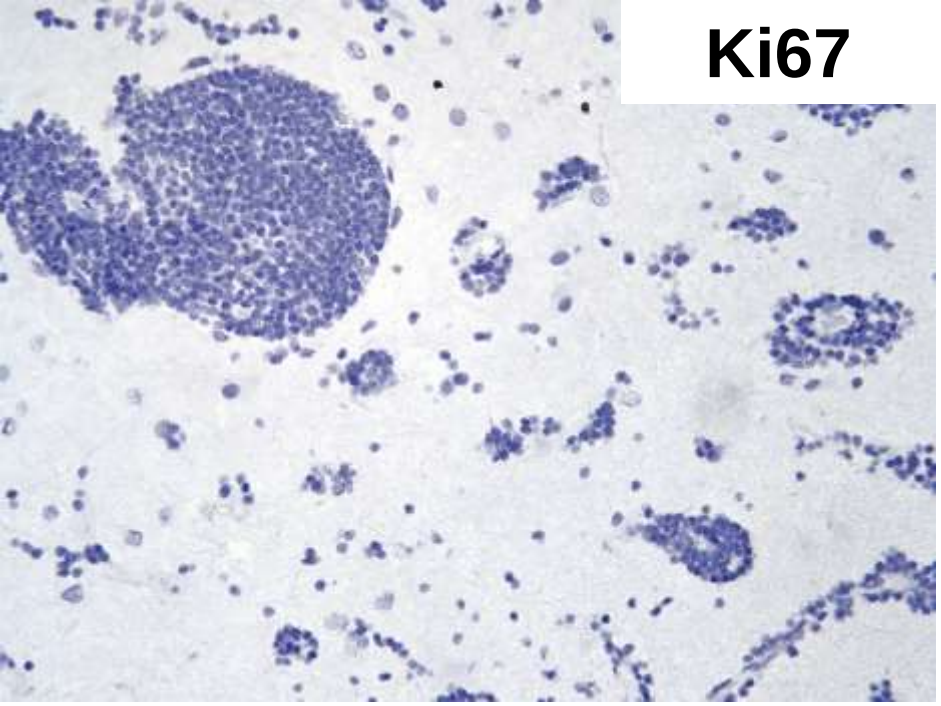
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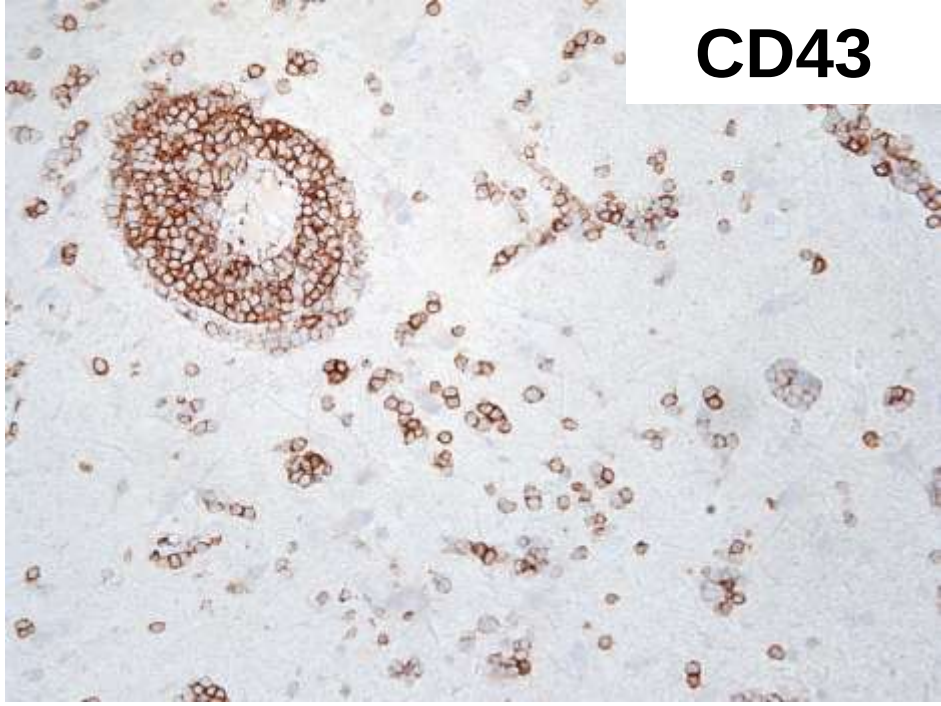
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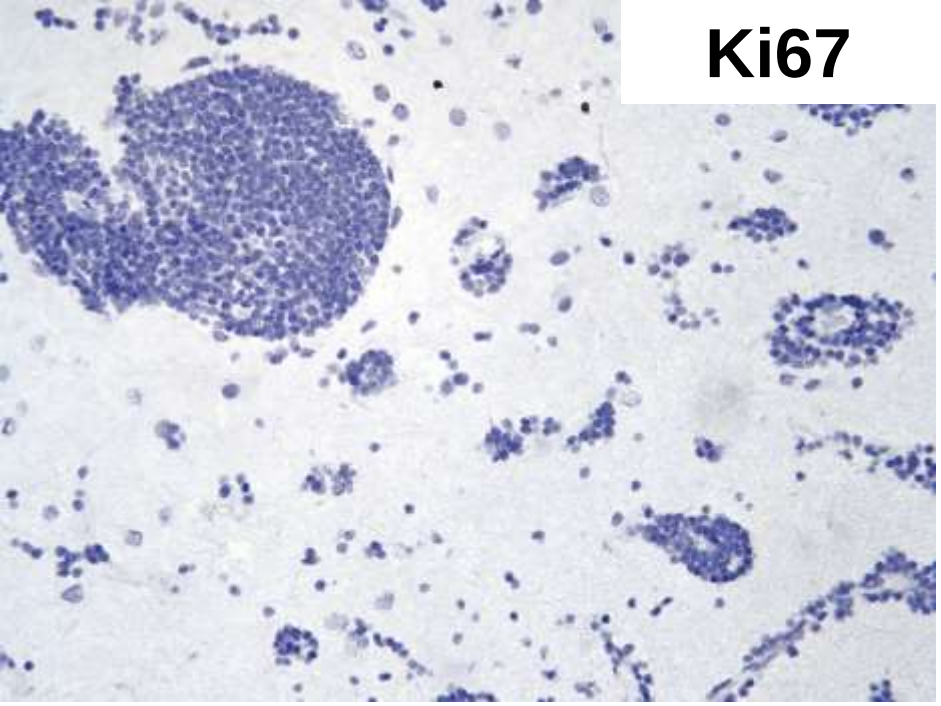
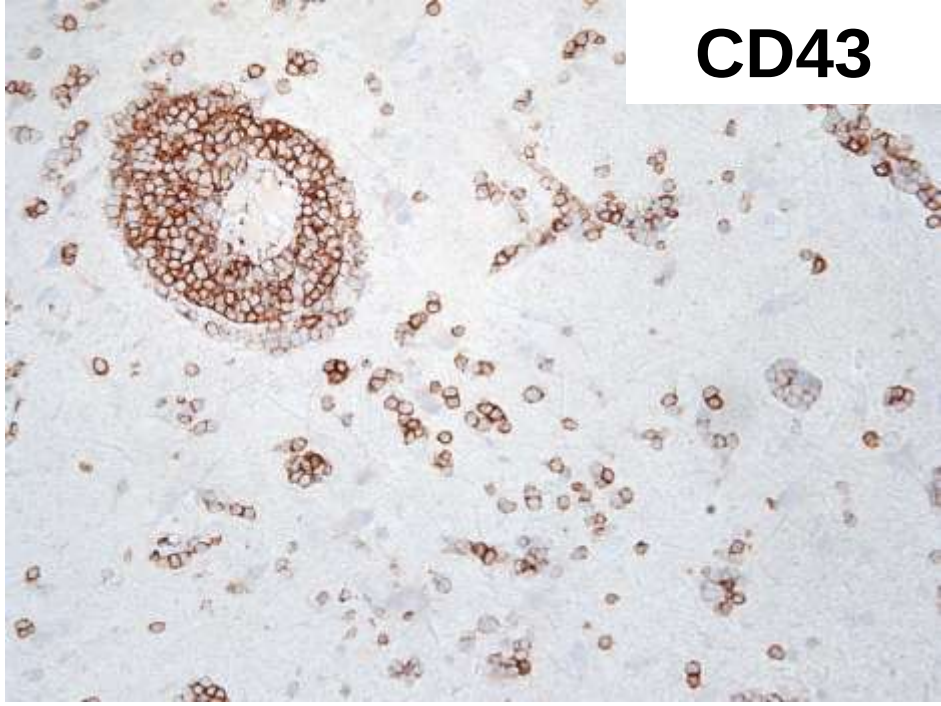




RIGHT FRONTAL SUBDURAL “EMPYEMA”, BIOPSY

-- EXTRANODAL MARGINAL ZONE LYMPHOMA

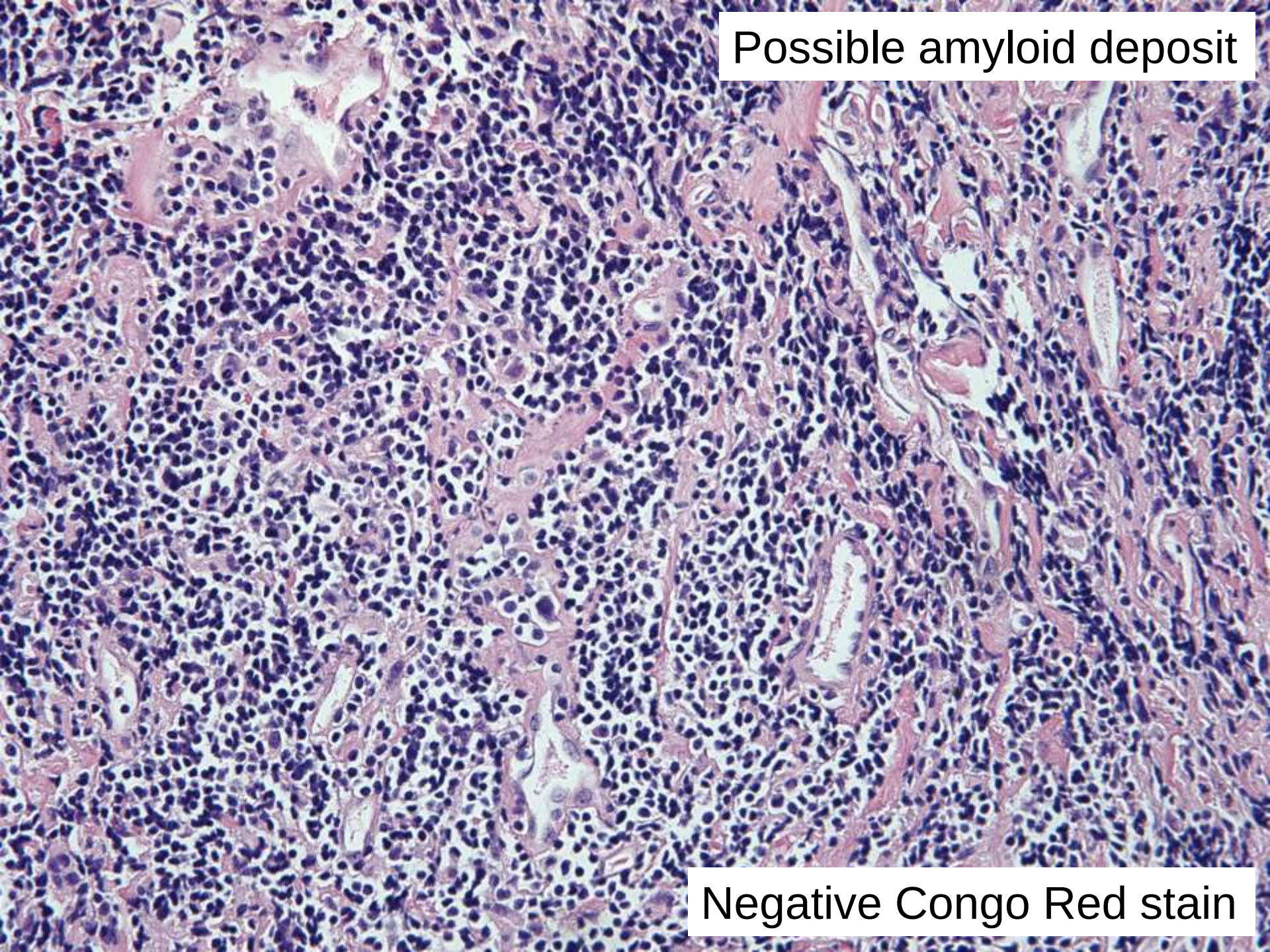
Dural marginal zone lymphoma

- Middle-aged women
- Radiographic findings: mimic meningioma or subdural hematoma
- D/D: meningioma, CLL/SLL, Castleman disease, dural metastasis, rheumatoid nodules
- Indolent disease course
- No standard treatments
  - surgery, local radiation therapy, chemotherapy

Table 1. Reported cases of intracranial extranodal marginal zone B-cell lymphomas

Case	Reference	Age	Sex	Location	Symptoms	Treatment and outcome	Immunohistochemistry
1	18	39	F	Left CP angle (dura)	Hearing loss, facial pain/weakness	Subtotal excision; AWD after 4 yr	CD20+, CD79a+, CD21+ germinal centers, kappa LCR
2	18	62	F	Left parietal-occipital area	Headaches	Radiation; AWD after 6 months	CD20+, CD79a+
3	17	40	F	Right cavernous sinus	Numbness, visual field defects	Radiation; NED at 63 months	CD20+, CD3+ reactive T cells; lambda LCR
4	17	62	F	Biparietal dural thickening	Seizures	Systemic fludarabine; NED at 22 months	CD20+, CD3+ reactive T cells; lambda LCR
5	17	52	F	Left frontal dural mass	Seizures and numbness	Radiation/chemo; NED at 9 months	CD20+, CD3+ reactive T cells; kappa LCR
6	17	43	F	Left tentorial mass	Dizziness, headaches, blurred vision, numbness	Radiation; NED at 7 months	CD20+, CD3+ reactive T cells, CD43+; lambda LCR
7	17	57	F	Left anterior falx cerebri mass	Seizures	Radiation; NED at 14 months	CD20+, CD3+ reactive T cells, CD43+; equivocable LCR
8	19	66	F	Right parietal dura	Syncope, seizures	Radiation; NED at 12 months	Not described
9	14	28	F	CP angle	Tinnitus, nausea, headache, bilateral papilledema	Excision; NED at 2 yr	CD20+, CD10+ (follicular center cells), bcl2+ in some follicular centers, CD43+, CD3+ (50% cells)
10	20	64	F	Right fronto-parietal dura	Left hemiparesis, headache	Subtotal excision, radiation; NED at 3 months	IgD+/CD20+ small lymphocytes, IgD-/CD20+ lymphoplasmacytoid cells, CD20-/CD138+ plasma cells; kappa LCR
11	Current	47	M	Left tentorial mass	Seizure, visual field defects, memory loss	Subtotal excision, radiation; NED at 8 months	CD20+, CD79a+, CD43+; kappa LCR

LCR, light chain restriction; CP cerebellopontine; AWD, alive with disease; NED, no evidence of disease.


Possible amyloid deposit
Negative Congo Red stain

Take home points

1. Marginal zone lymphoma is the most common primary dural lymphoma

2. Small bland lymphocytes +/- amyloid deposition

3. Indolent disease course; no standard treatment

References

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