هفت سین در پاتولوژی

سین اول

- A 15 y/o man with recurrent flank pain, prior history of urinary tract infections and hematuria
- Unremarkable physical exam
- Renal ultrasound shows a staghorn stone filling the pelvis

Urine analysis

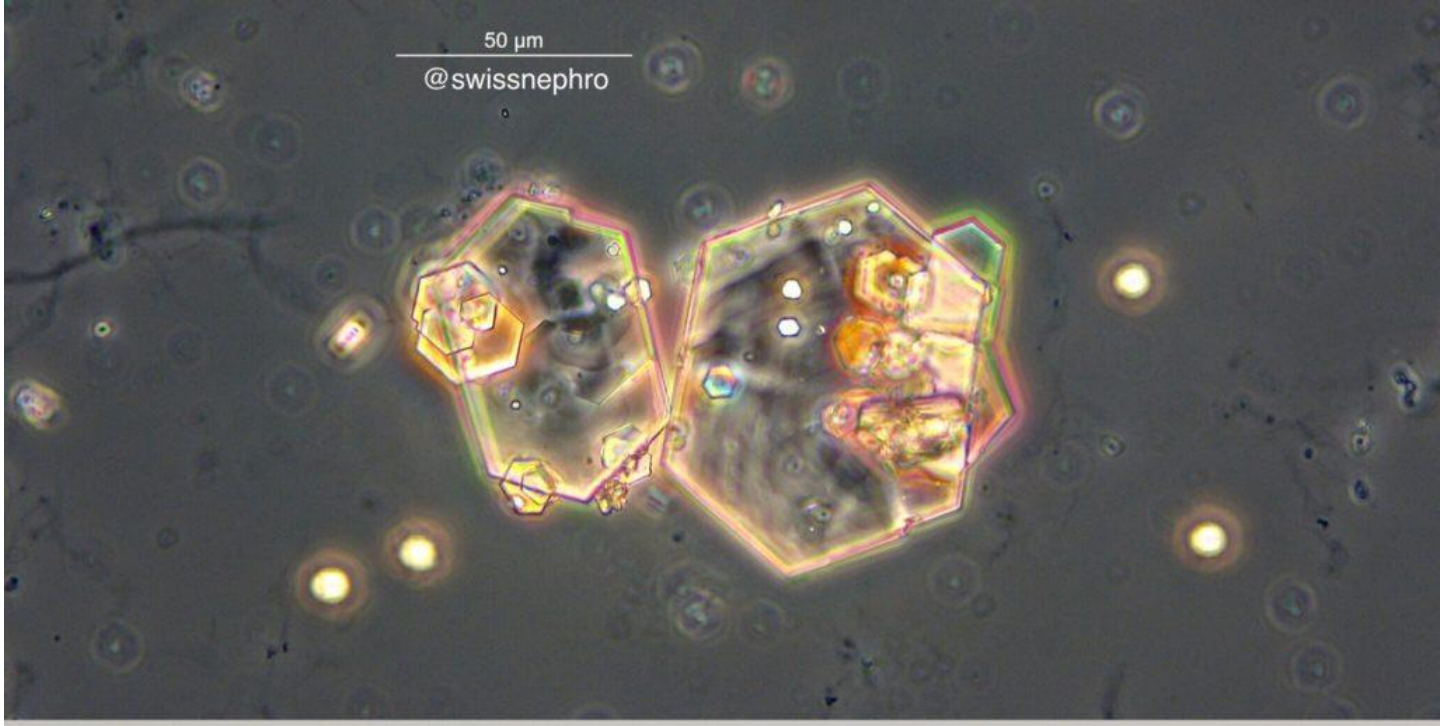


Figure credit: https://www.renalfellow.org/wp-content/uploads/2020/03/Fig-2-1024x1024.jpg

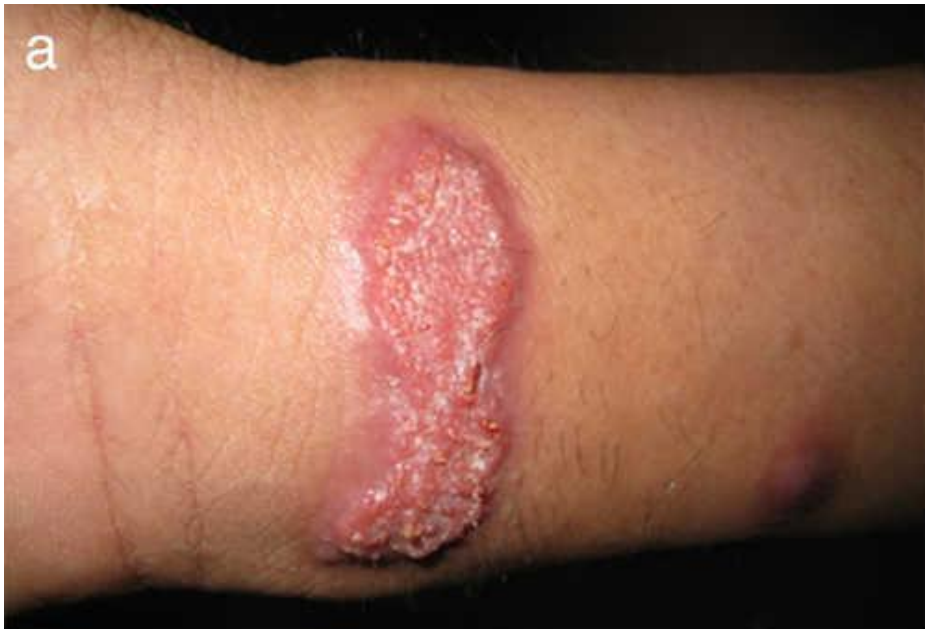
Cystinuria سیستینوری

- Cystinuria: autosomal recessive
  - Incidence: 1/10000
  - Defect in *SLC3A1* or *SLC7A9* (coding membrane receptor)
  - Lack of reabsorption of positive charged amino acids: cystine, lysine, ornithine and arginine
- Predilection to recurrent urinary stones, UTI and renal failure
  - Urine smells of rotten egg (due to cystine)
- Diagnosis:
  - Identification of crystals (non-polarizing) (unlike uric acid crystals)
  - Cyanide nitroprusside test
- Treatment:
  - Hydration, urine alkalinization, thiol drug therapy

سین دوم

- A 50 y/o female, complaining of several papules and pustules on upper extremity
- Teacher, florist by hobby
- Hx of alcohol consumption
- Scraping specimen

obtained



Picture credit: Rev Iberoam Micol 2016: 33:38-42

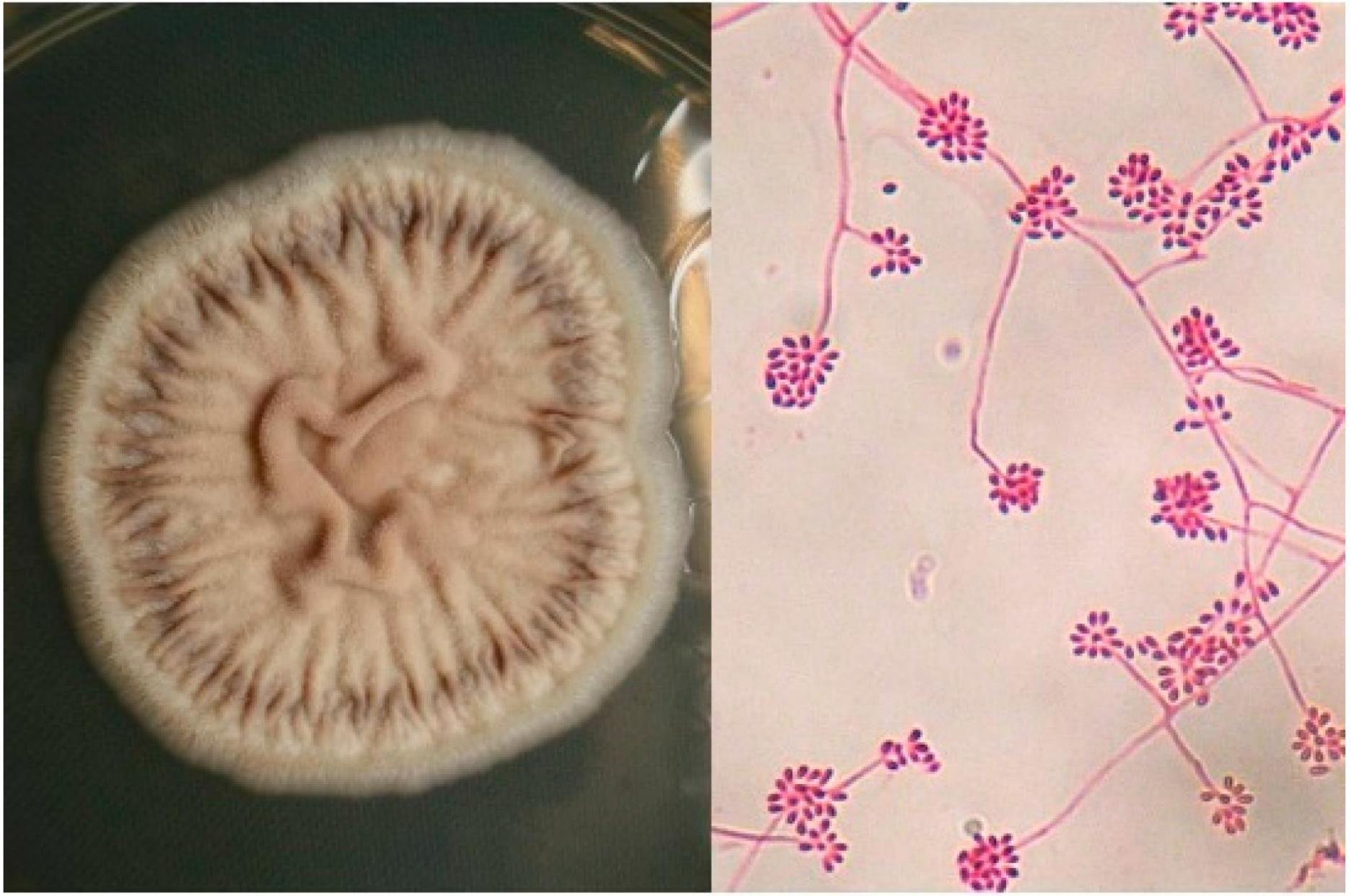


Figure credit: J. Fungi 2017, 3(1), 6

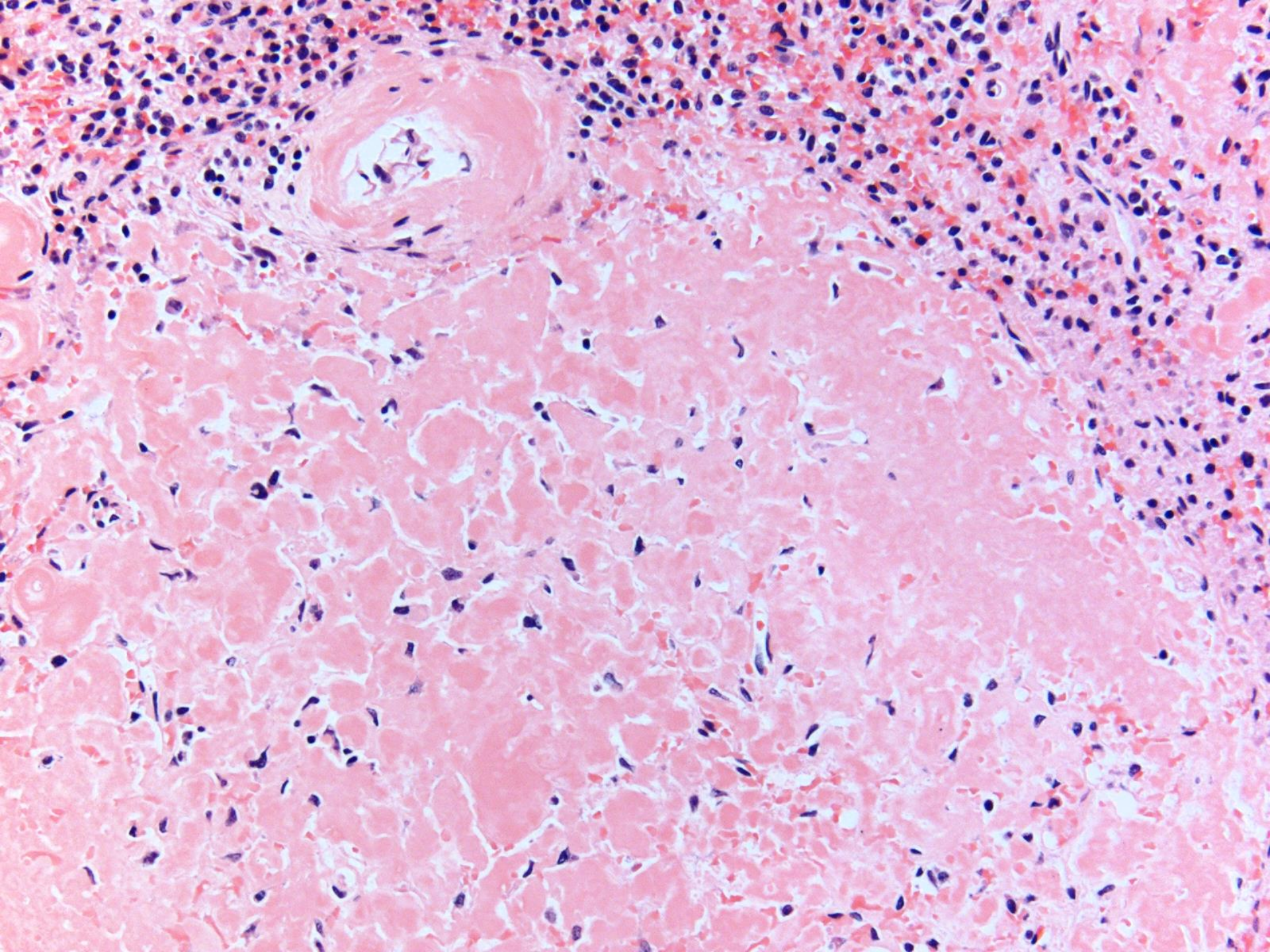
Sporothricosis اسپوروتریکوسیس

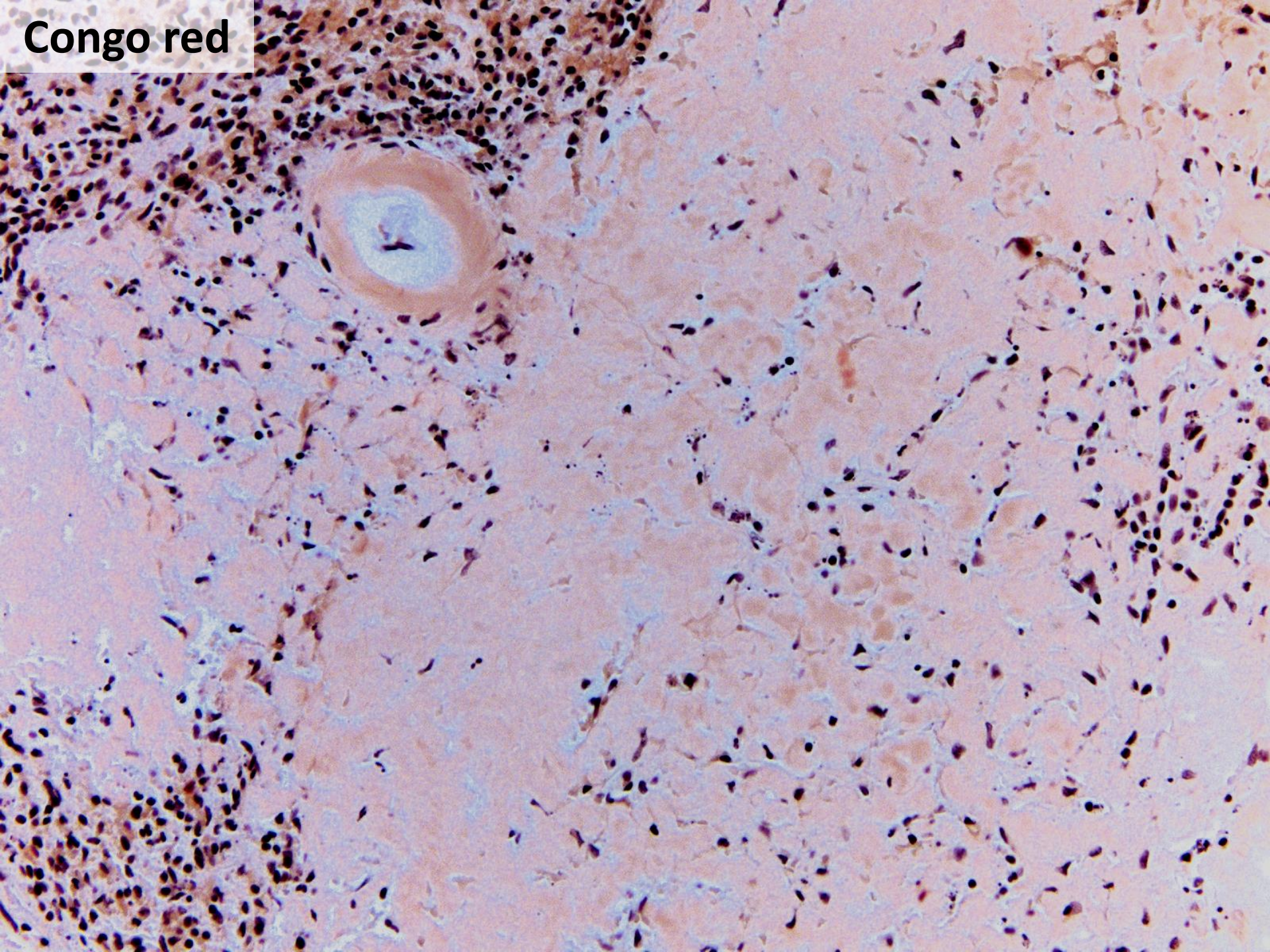
- Etiology: Sporothrix Schenckii, a dimorphic fungus commensal on vegetations (yeast-form in 37ºC, hypha in 25ºC)
- Subacute infection of skin and subcutis
  - Granulomatous inflammation with/out suppuration
- Treatment:
  - Itraconazole (3-6 months) (oral)
  - Supersaturated potassium iodide (SSKI) (topical)

سین سوم

- A 57 y/o male with medical history of HTN, peripheral vascular disease, DVT and pulmonary embolism (on Coumadin) was admitted for increasing respiratory difficulty
  - Within 4 hours he developed respiratory distress, decreased saturation and became unresponsive
  - Intubation, started pressors for fluctuation of blood pressure
  - Initially saturation improved (extubated) but later deteriorated (intubated again)
  - Echo: no thrombus
- Passed away after two weeks in ICU

Spleen

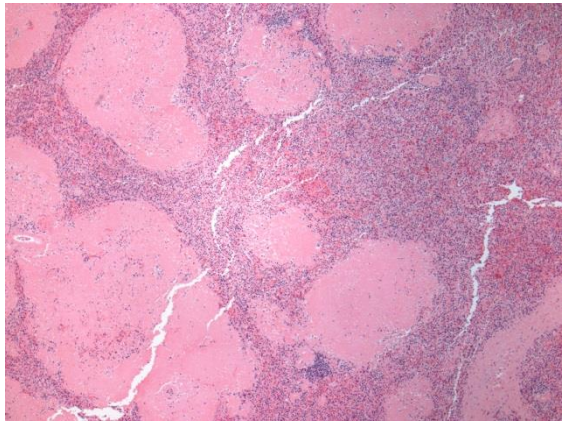



Congo red

Sago spleen طحال ساگو

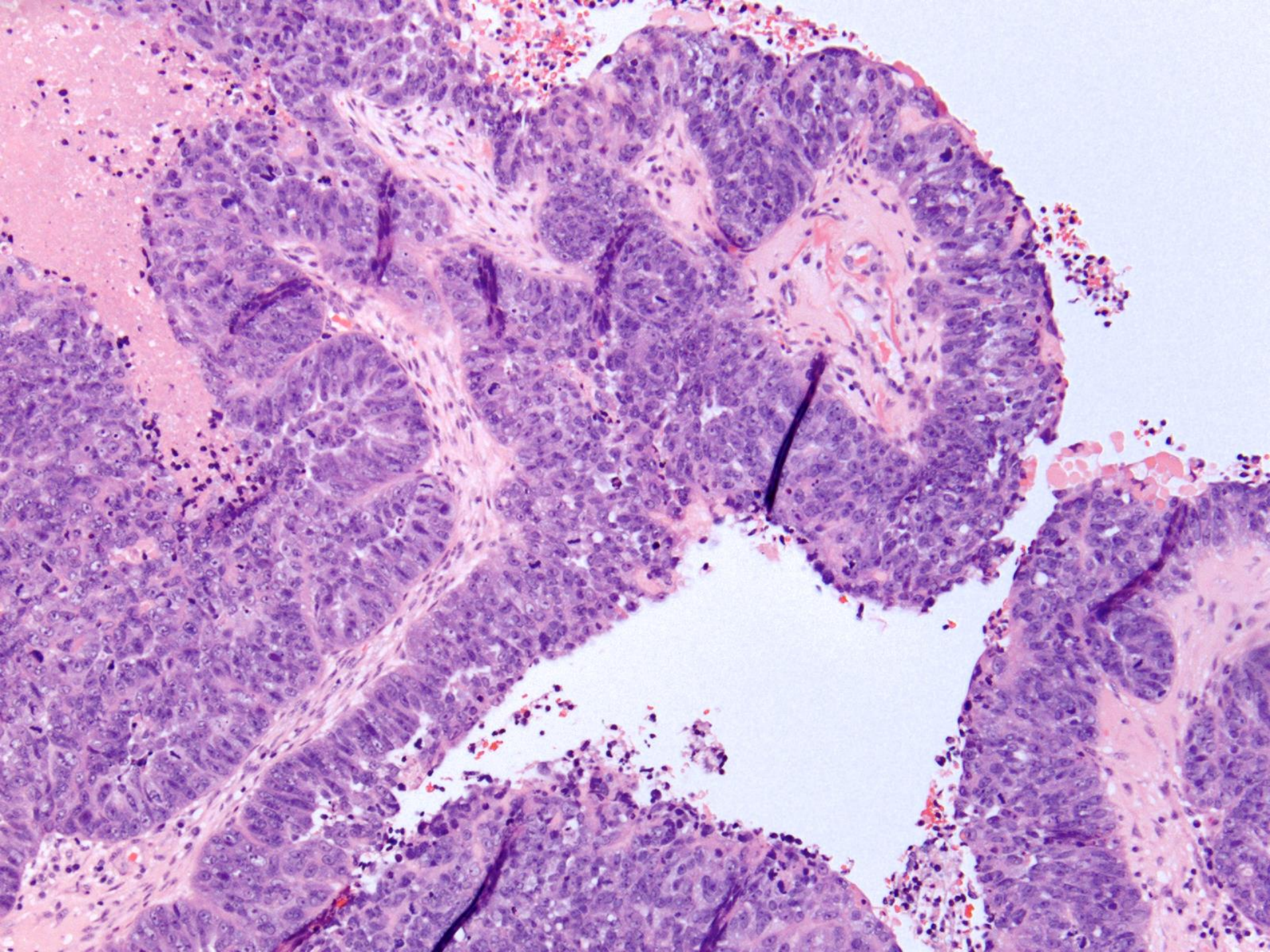
- Secondary amyloidosis of spleen:
  - Follicular pattern: Sago spleen
  - Diffuse pattern: Lardaceous spleen

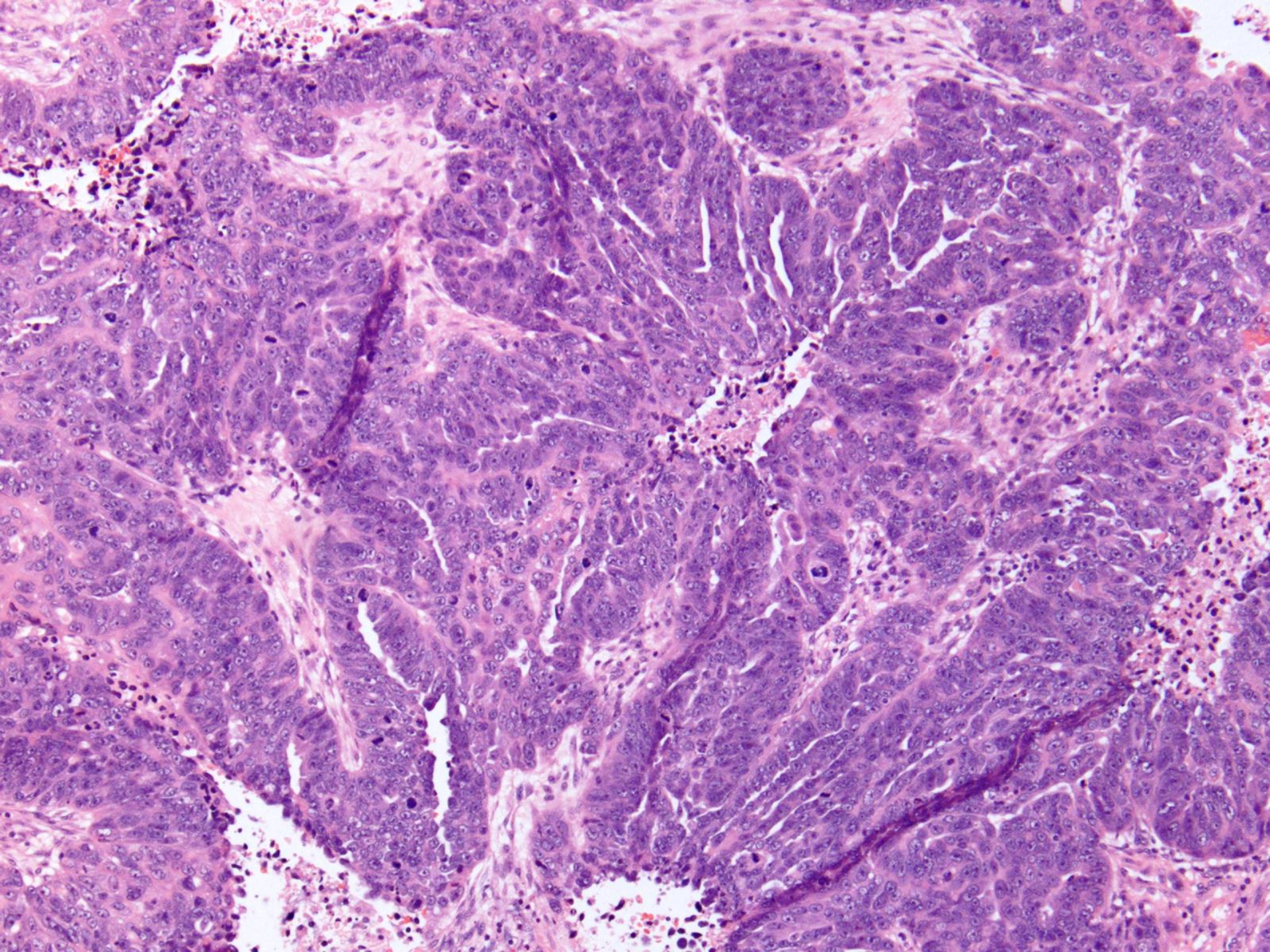
- Cause of death: systemic amyloidosis with multiple organ failure

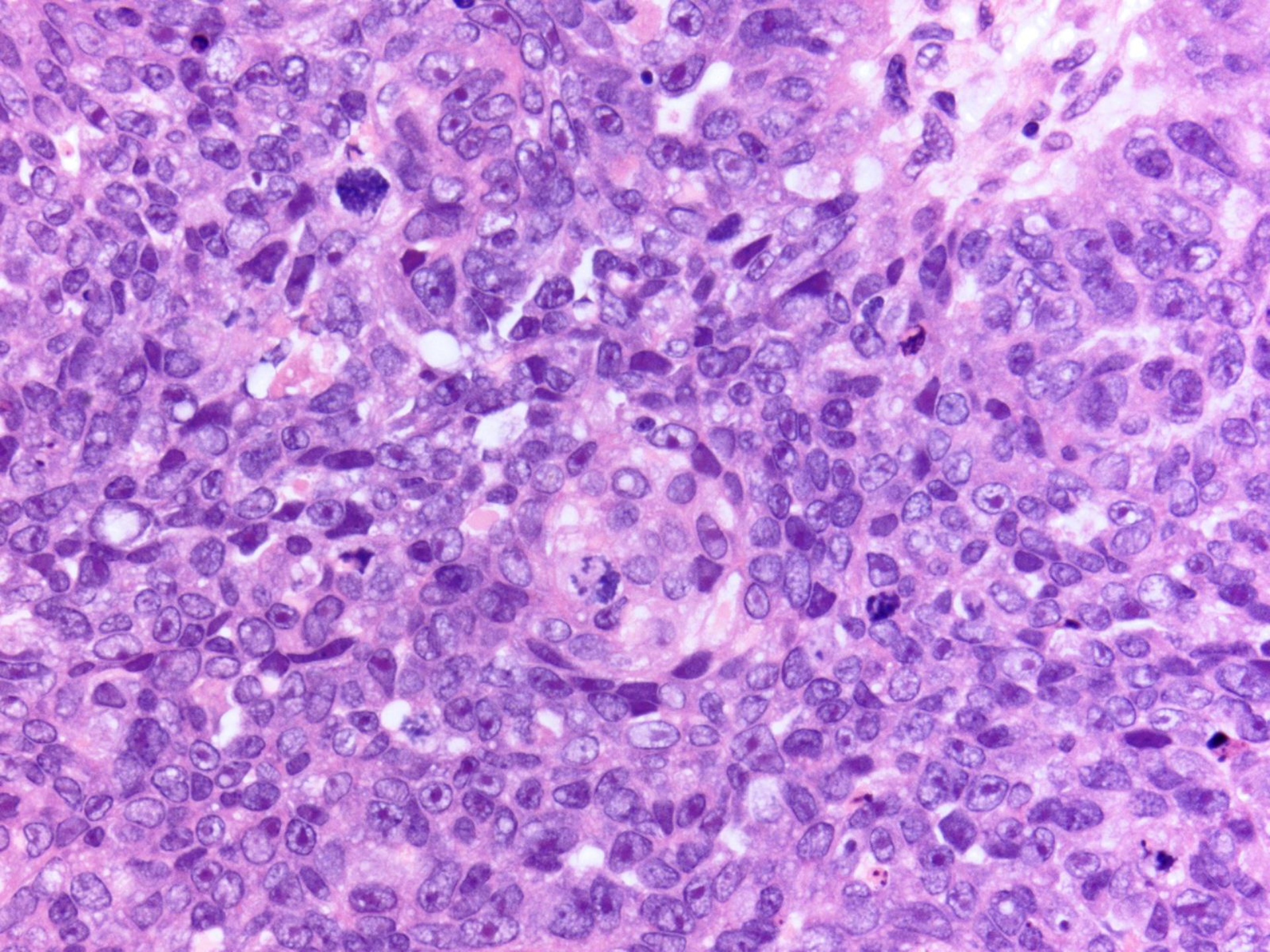


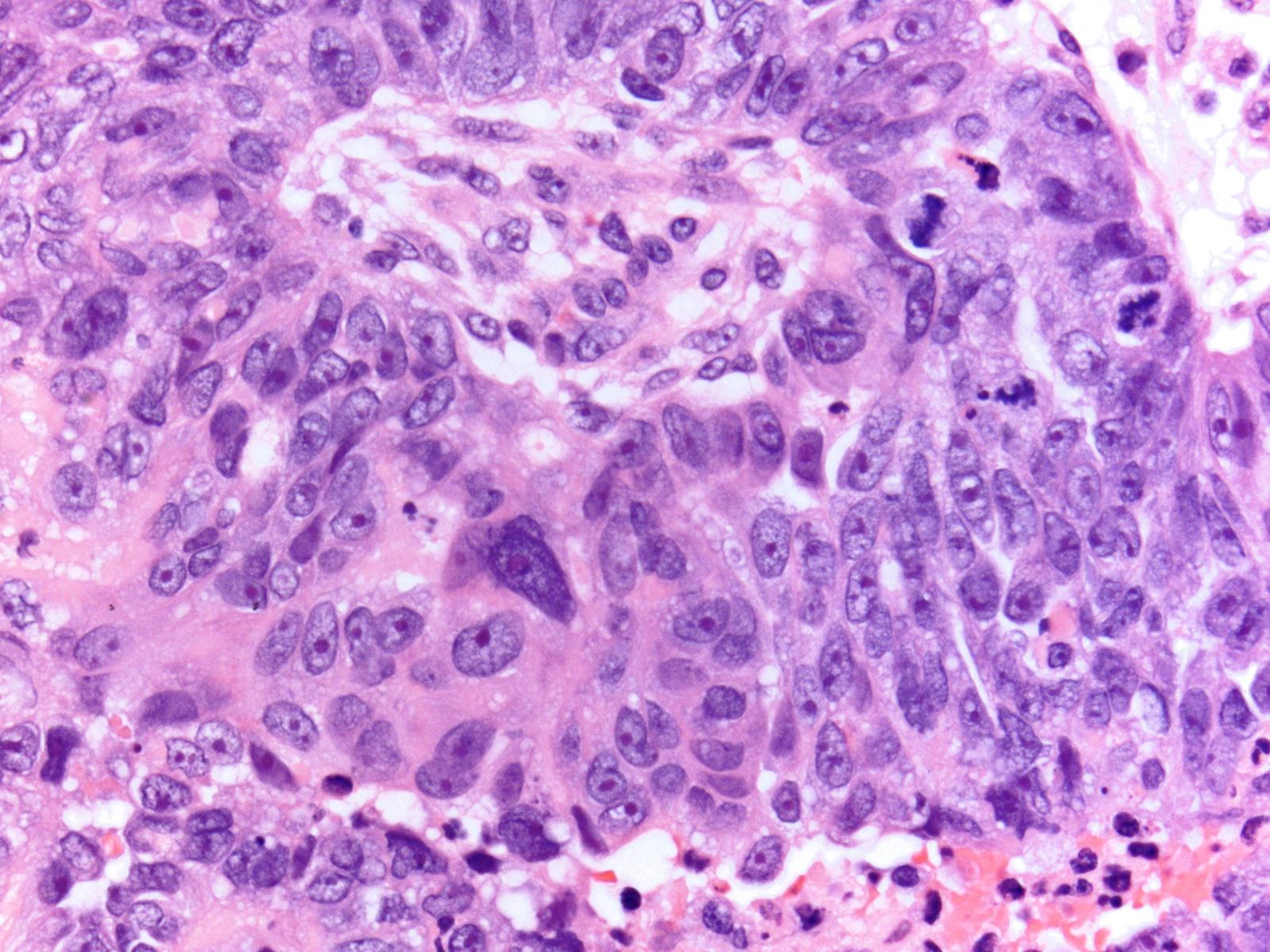
سین چهارم

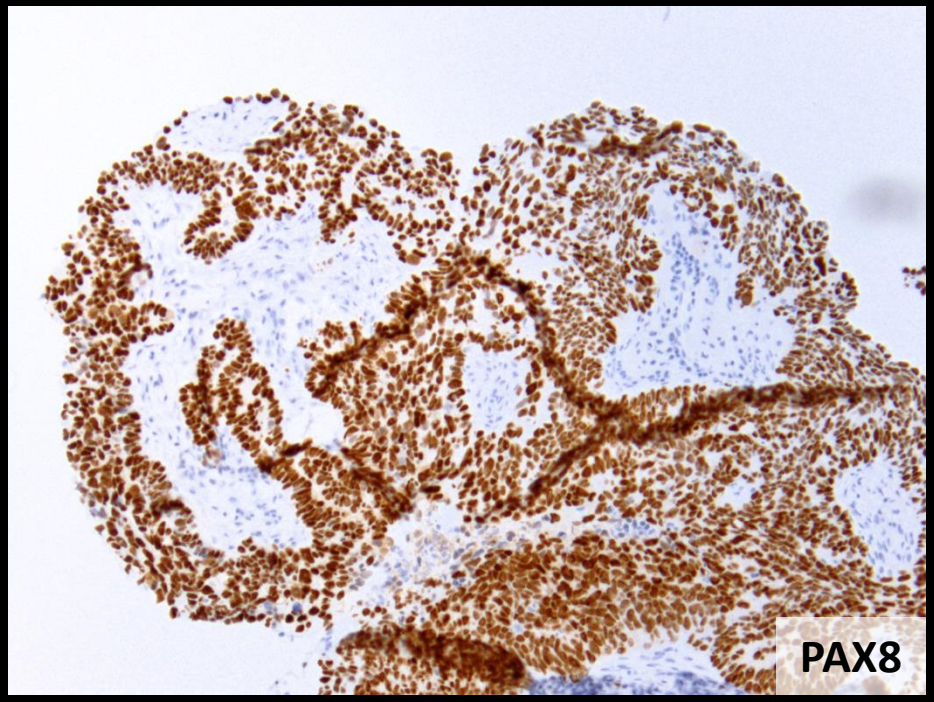
- An 82 year old female with history of intermittent abdominal pain
- CT abdomen reveals large sigmoid mass, ascites and peritoneal carcinomatosis
- Findings at surgery: firm omental lesions, several nodules studding the peritoneal surfaces, firm adherent mass in the pelvis (appeared to be the left ovary - firmly adherent to the sigmoid colon)

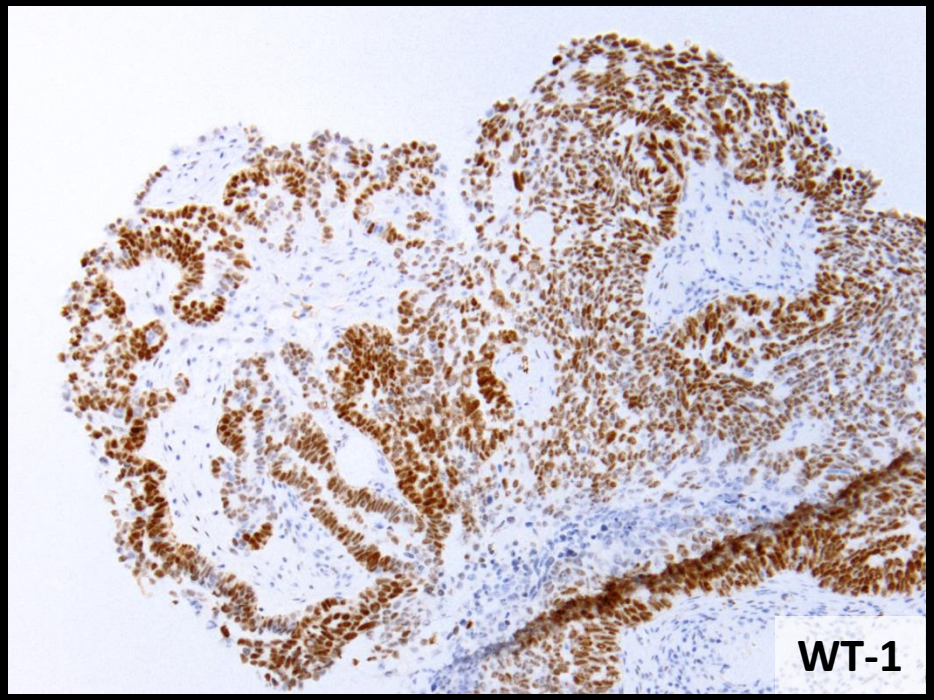


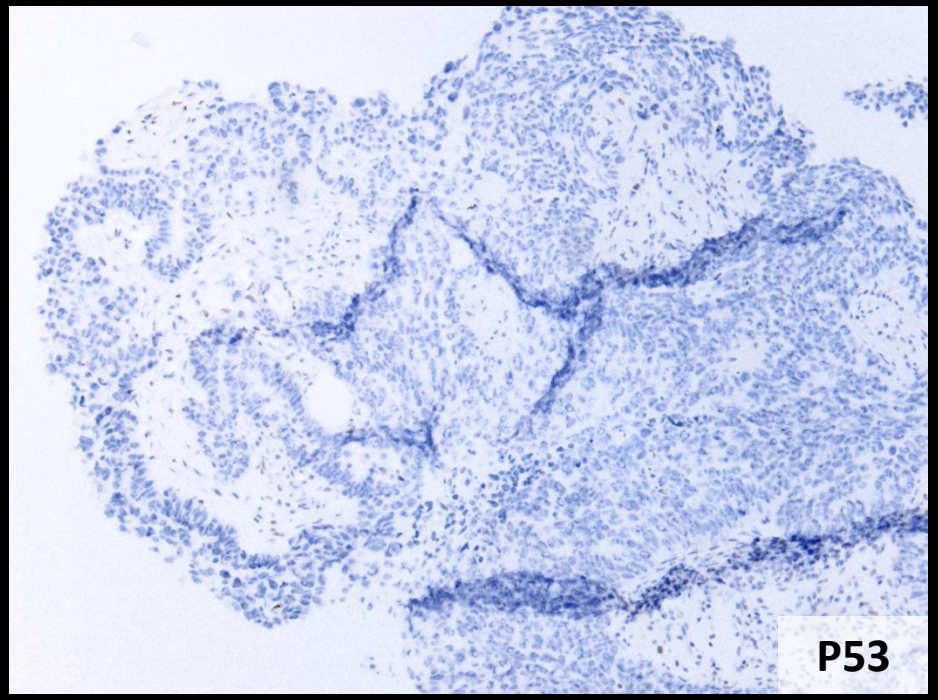







PAX8


WT-1

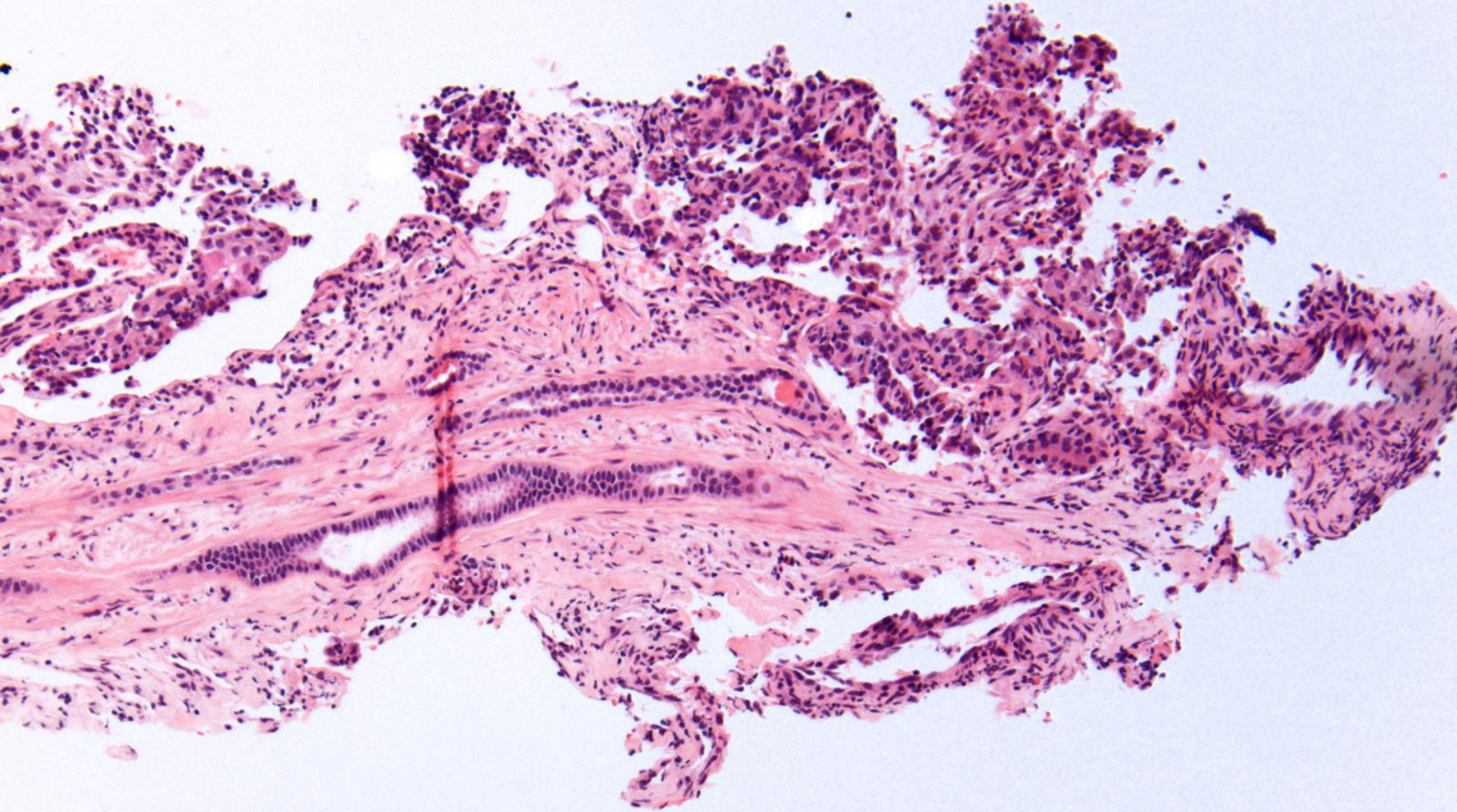

P53

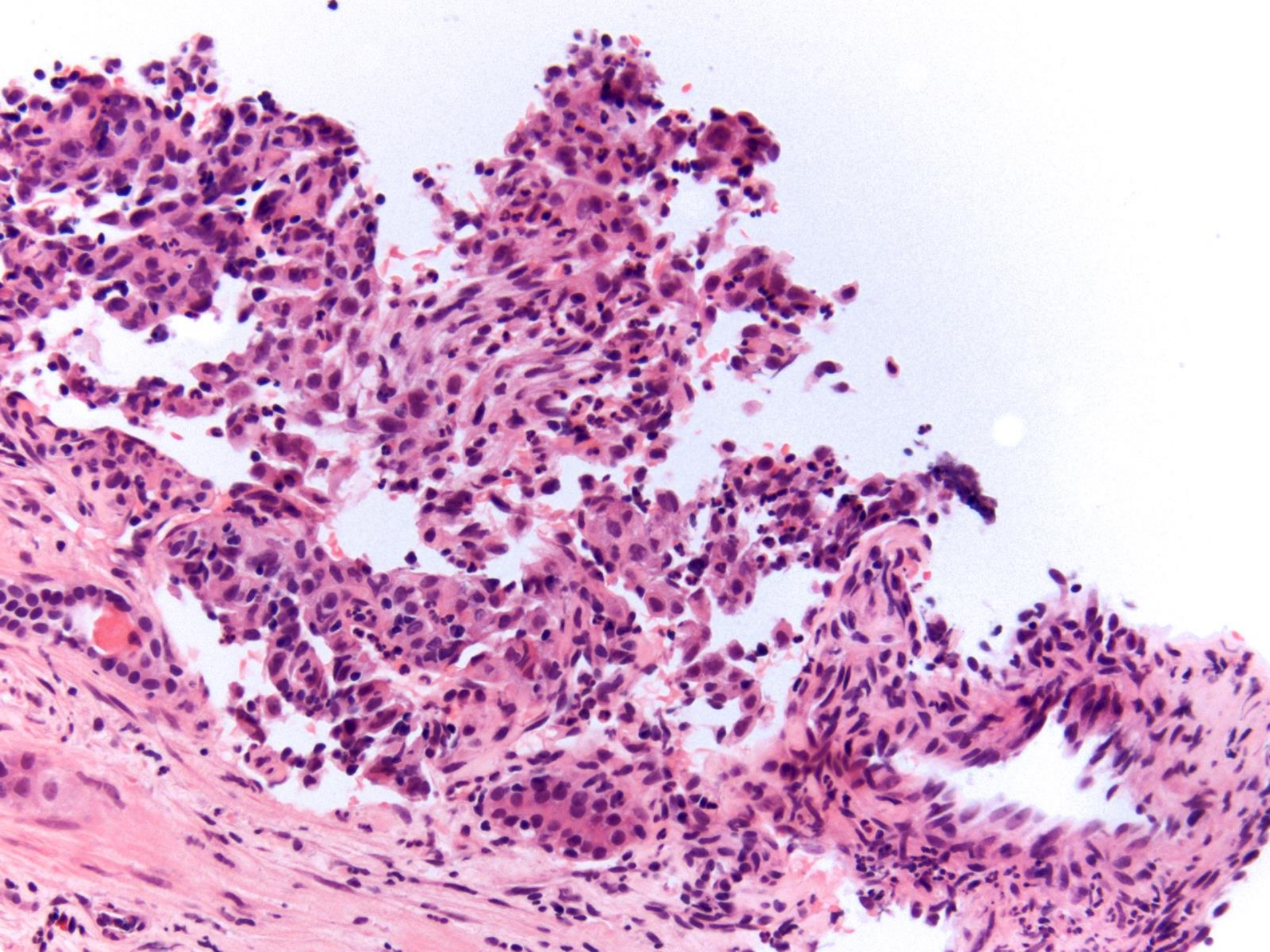
Serous carcinoma سروس کارسینوما

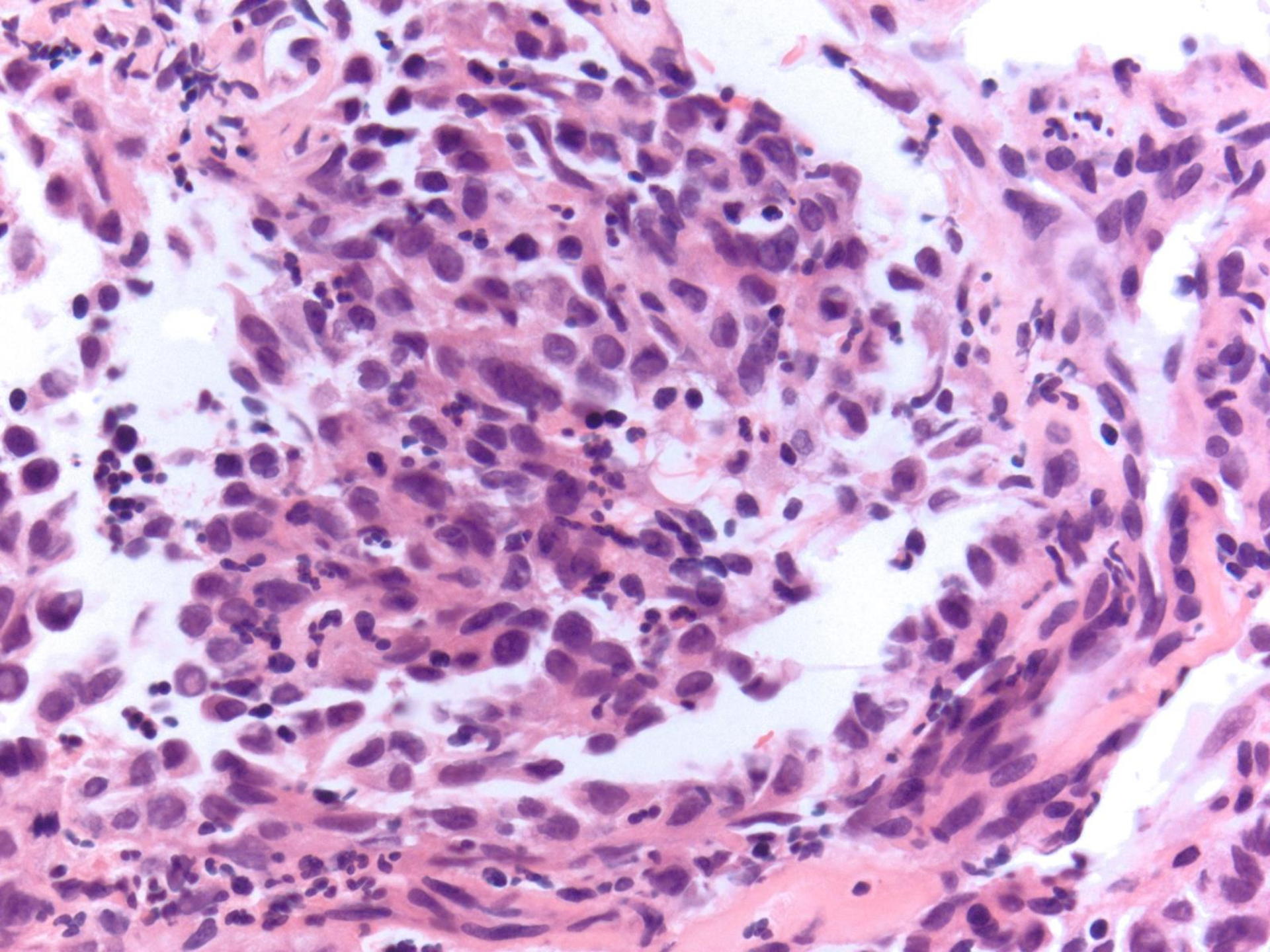
- Serous carcinoma: most common extrauterine female genital tumor
  - Almost always harbors *TP53* mutation
  - Commonly identified at advanced stages
  - Serum CA125 is usually elevated
  - Precursor: serous tubal intraepithelial carcinoma (STIC)
  - Chemotherapy is the mainstay of treatment
    - Treatment refractory is seen in *CCNE1* amplification and reversion of *BRCA1/2* mutation
    - Most important prognostic factor: stage

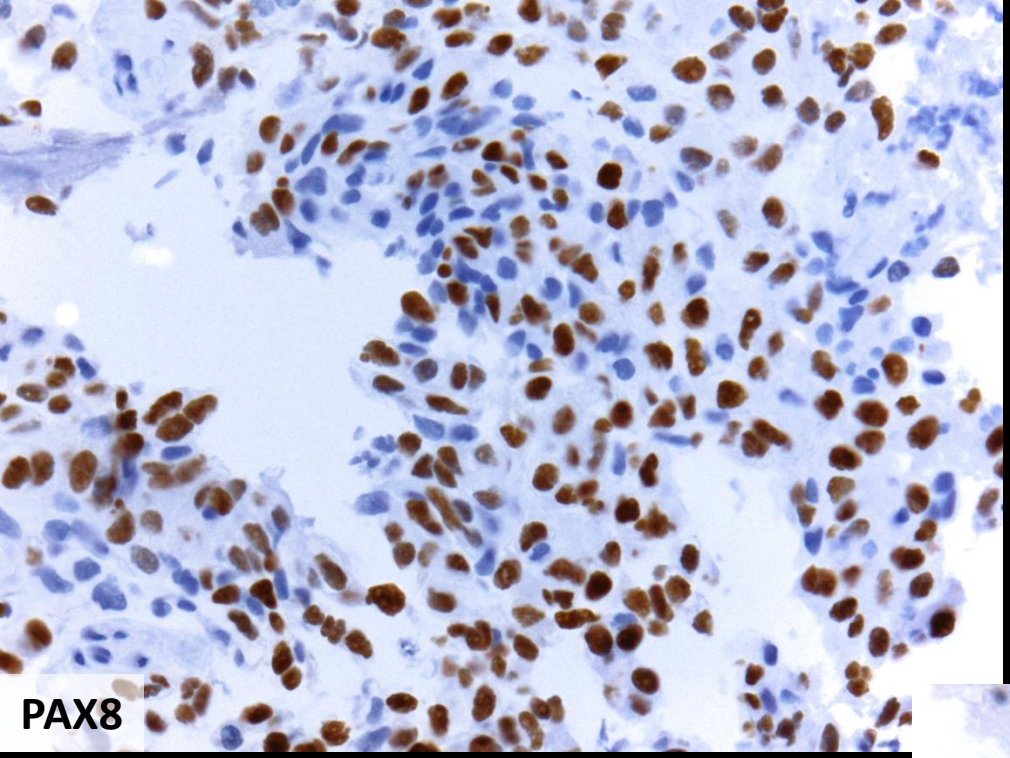
سین پنجم

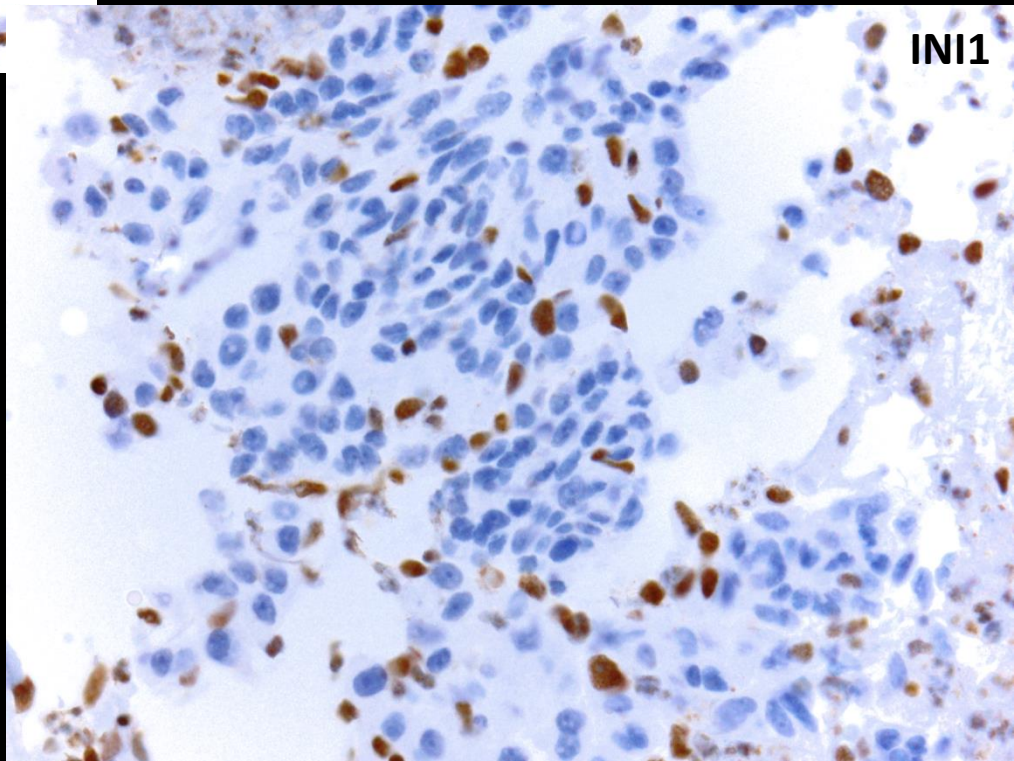
- A 35 y/o African-American male, previously healthy, complains of hematuria and right flank pain
- Reports a similar self-remitting episode a few years ago when visiting overseas
- Physical exam: mild flank tenderness, otherwise unremarkable
- CBC: normal; smear: rare target RBSs; U/A: specific gravity = 1.005 containing 15 RBC and 2 PMN, no bacteria
- Imaging: 5 cm mass in corticomedullary junction of the right kidney
- Kidney needle biopsy performed.







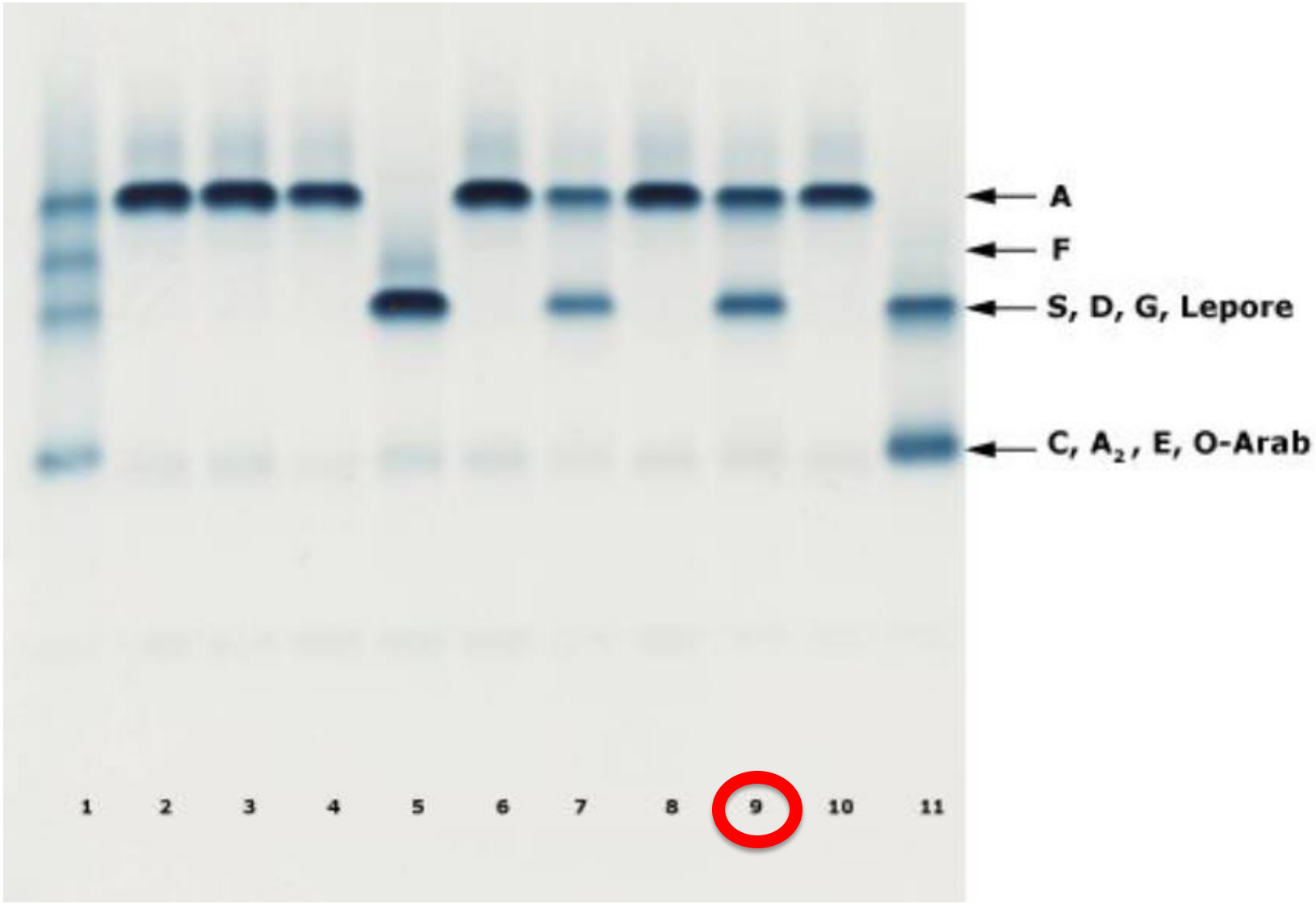

PAX8


INI1

Biopsy report

Poorly differentiated carcinoma with loss of INI1, favor renal medullary carcinoma

Hb electrophoresis

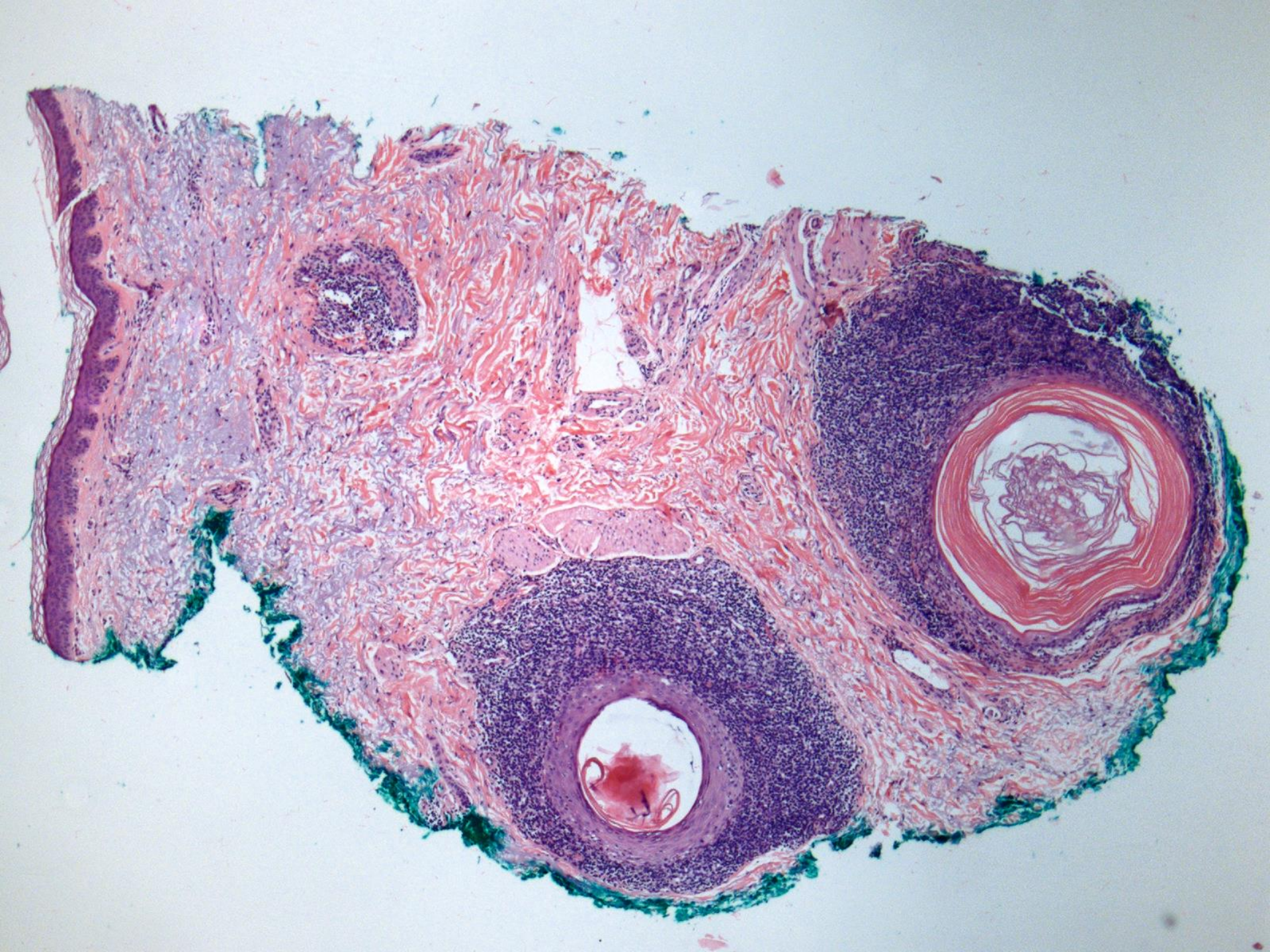


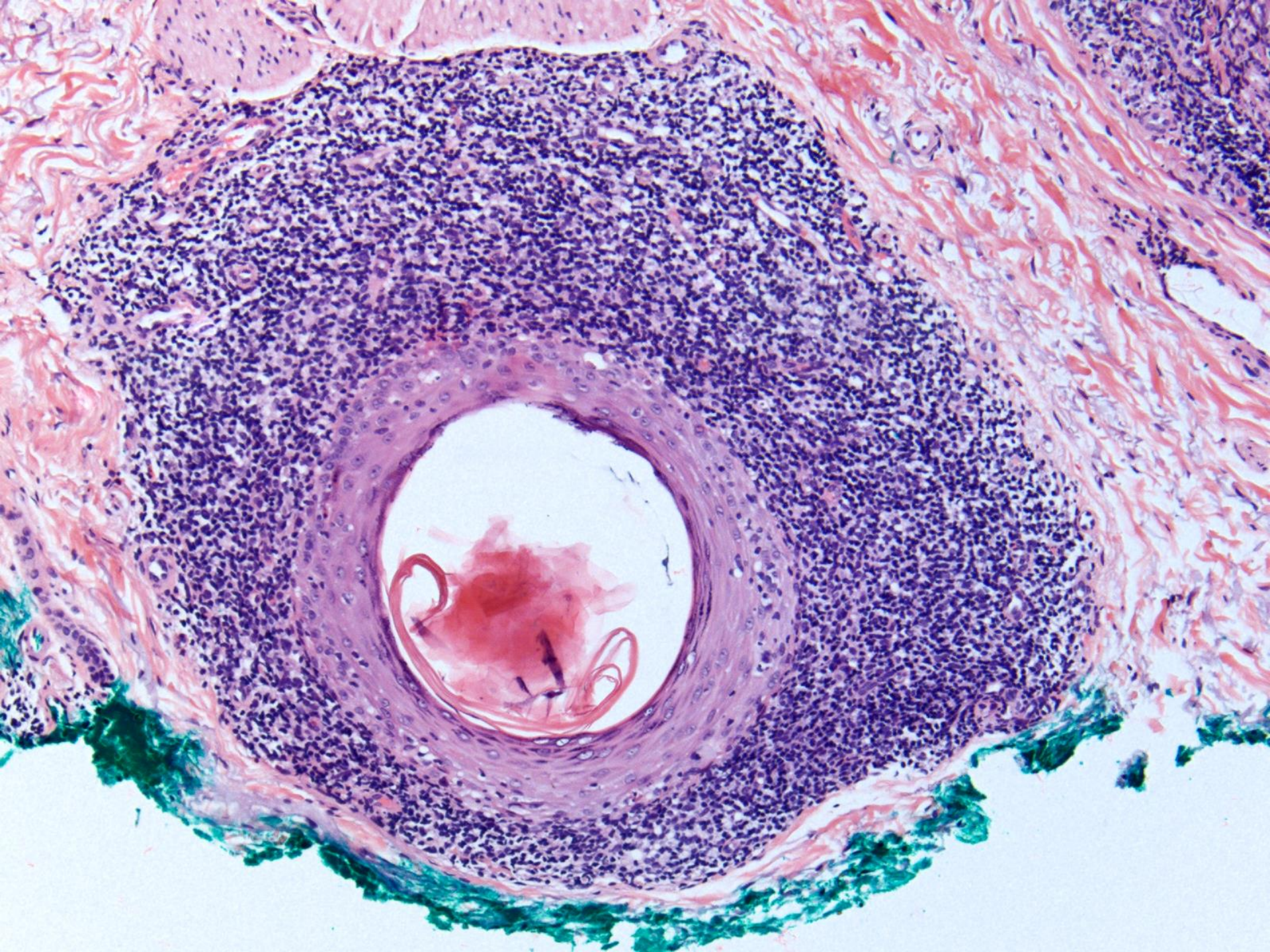
Sickle cell trait
بیماری سیکل سل

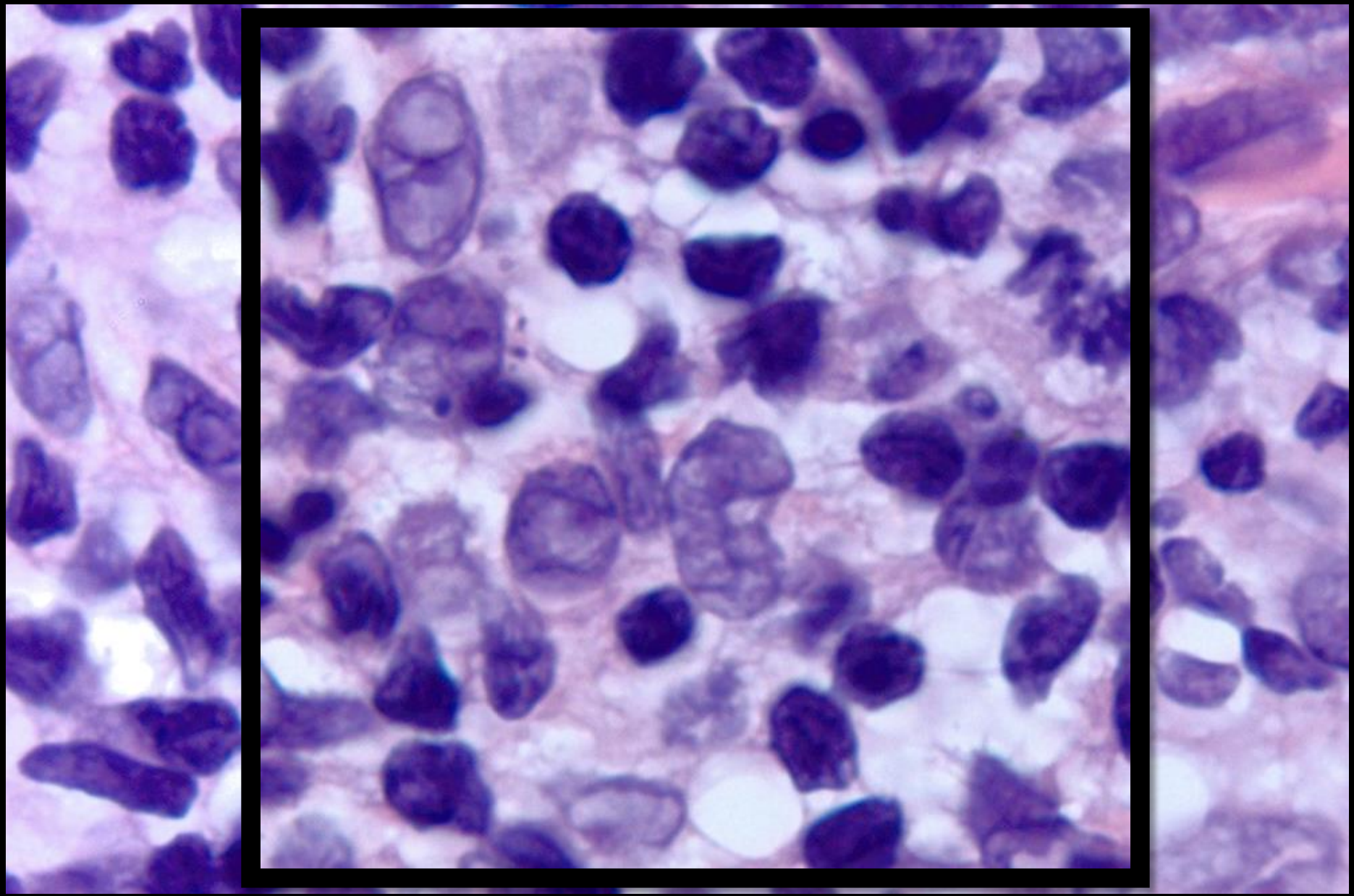
- Sickle cell trait (Hb AS)
  - Heterozygous state for Hb S is the most common hemoglobinopathy in the USA (8% in African-American population)
  - Usually asymptomatic unless extreme low oxygen state (may cause splenic infarct in high altitude or air flight)
  - Provides protection from falciparum malaria
  - Electrophoresis: Hb A >Hb S

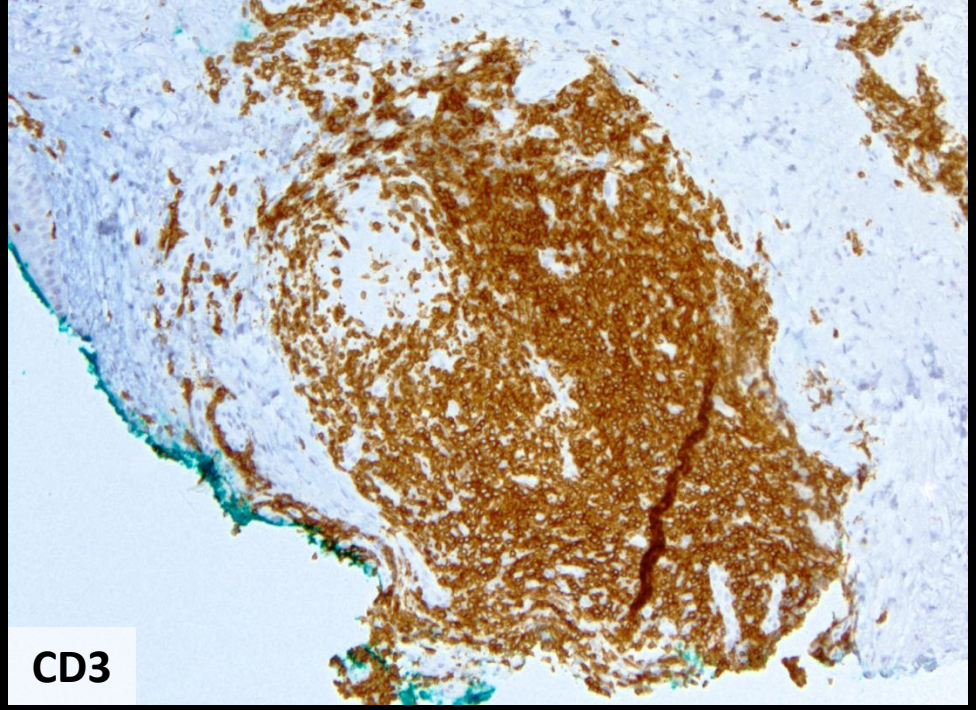
سین ششم

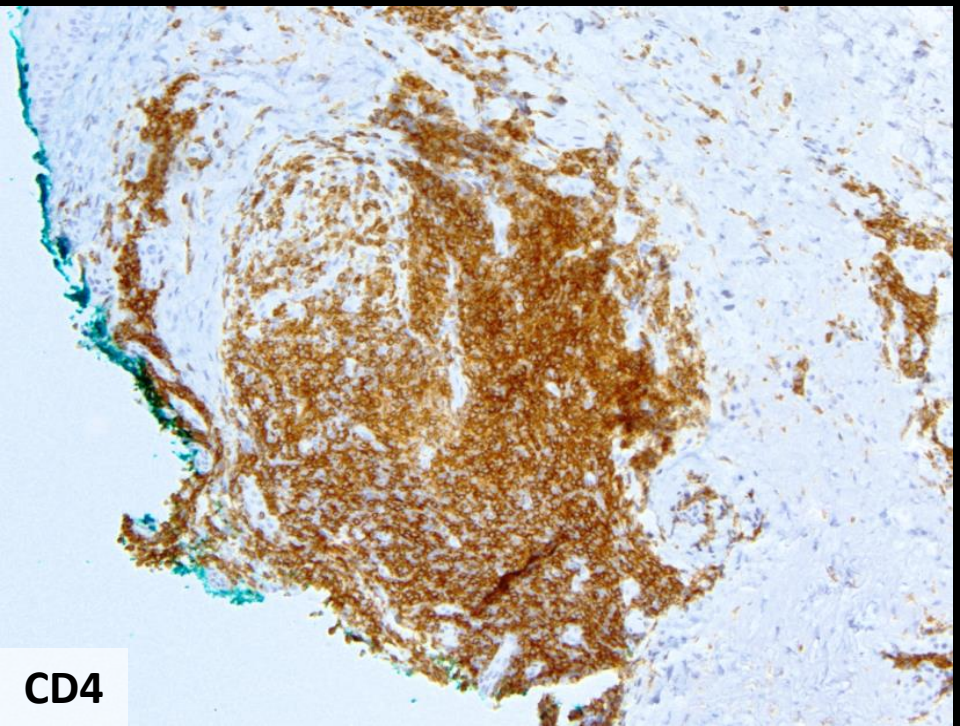
- 64 y/o male with prior skin cancers (SCC, BCC and AK) visited for annual skin surveillance for itchy skin lesions
- Erythematous patches on both forearms, non-scaling, with hair loss
- No change in shape or color, non-responsive to topical therapy for AK
- No personal or family history of melanoma
- A punch biopsy was obtained

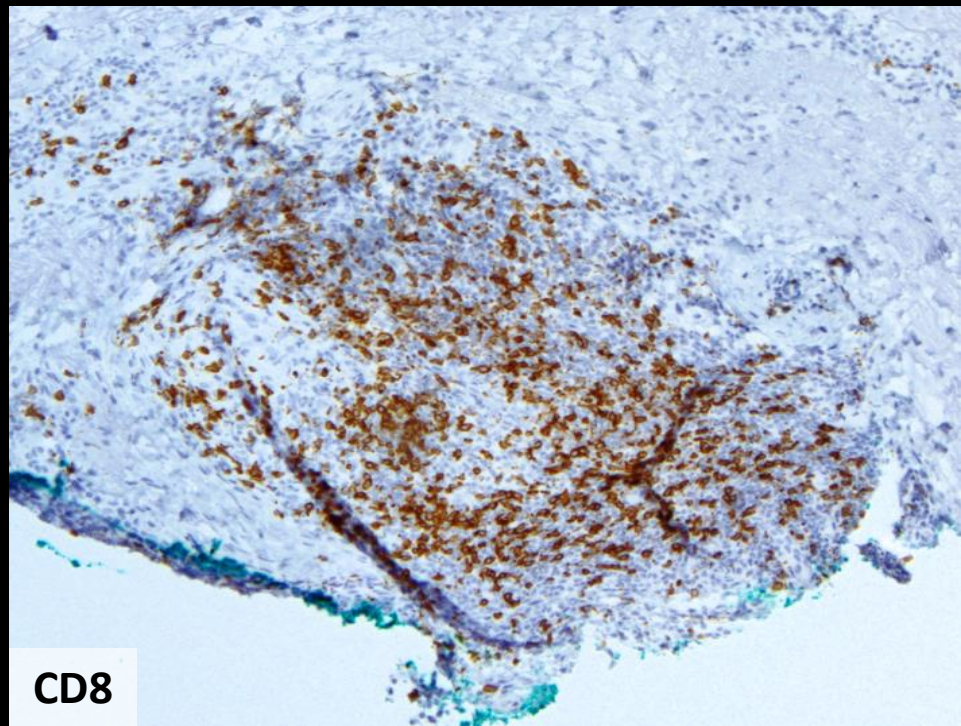







CD3


CD4

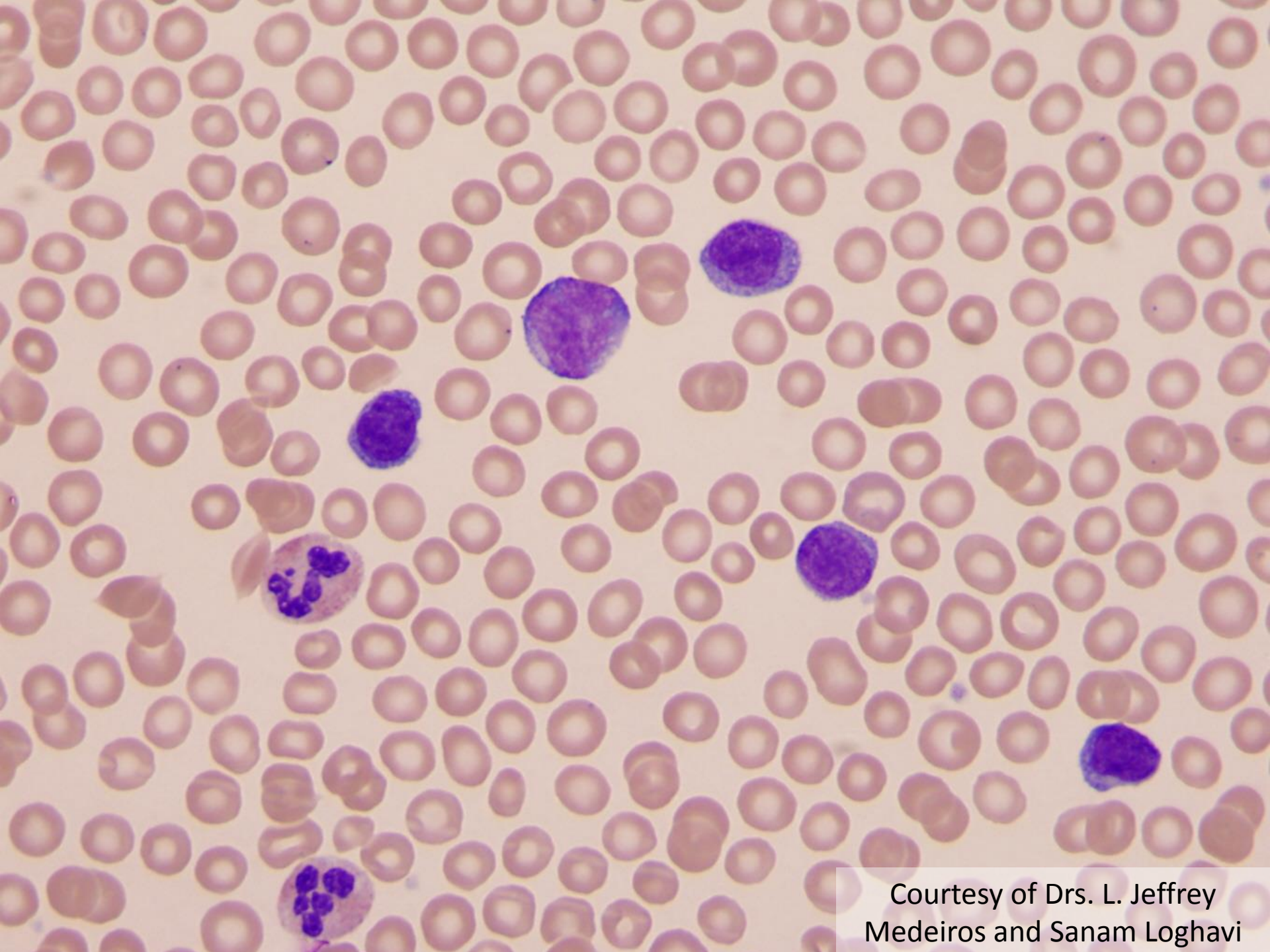

CD8

Biopsy report

Atypical clonal T cell infiltrate, highly suspicious for folliculotropic mycosis fungoides

(loss of CD7, clonal expansion of T cell receptor B)

Course:
Non-responsive to topical therapy, alopecic patches expanded, developed lymphocytosis and generalized LAP



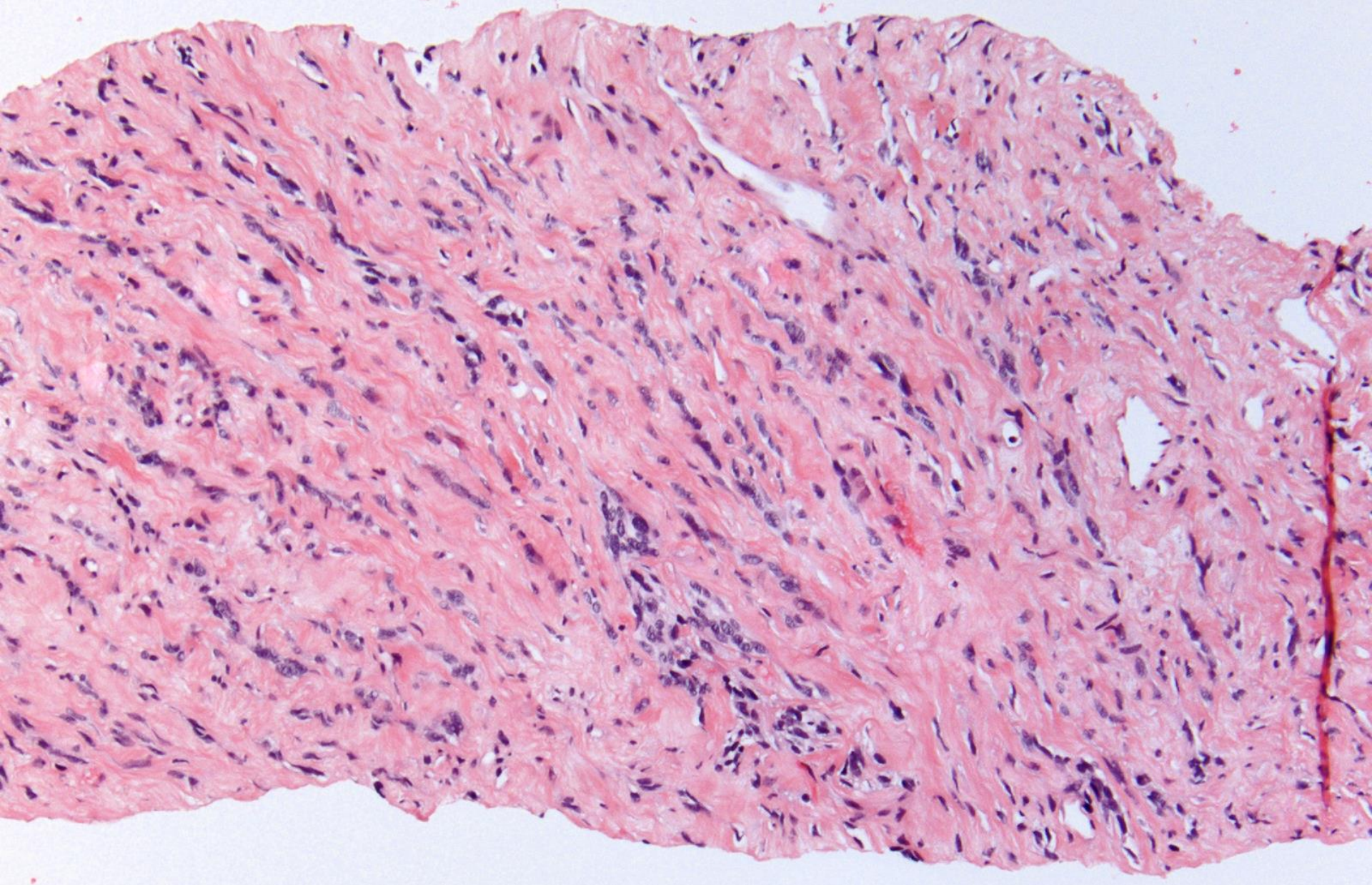
Courtesy of Drs. L. Jeffrey Medeiros and Sanam Loghavi

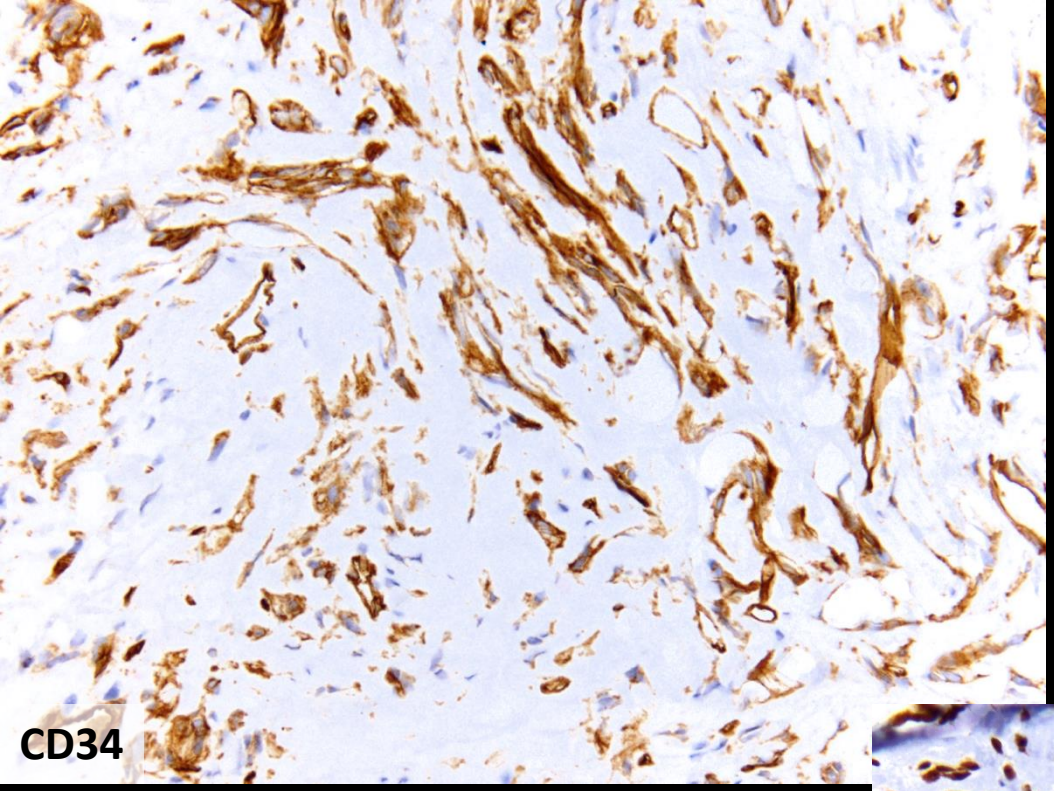
Sézary syndrome
سندرم سزاری

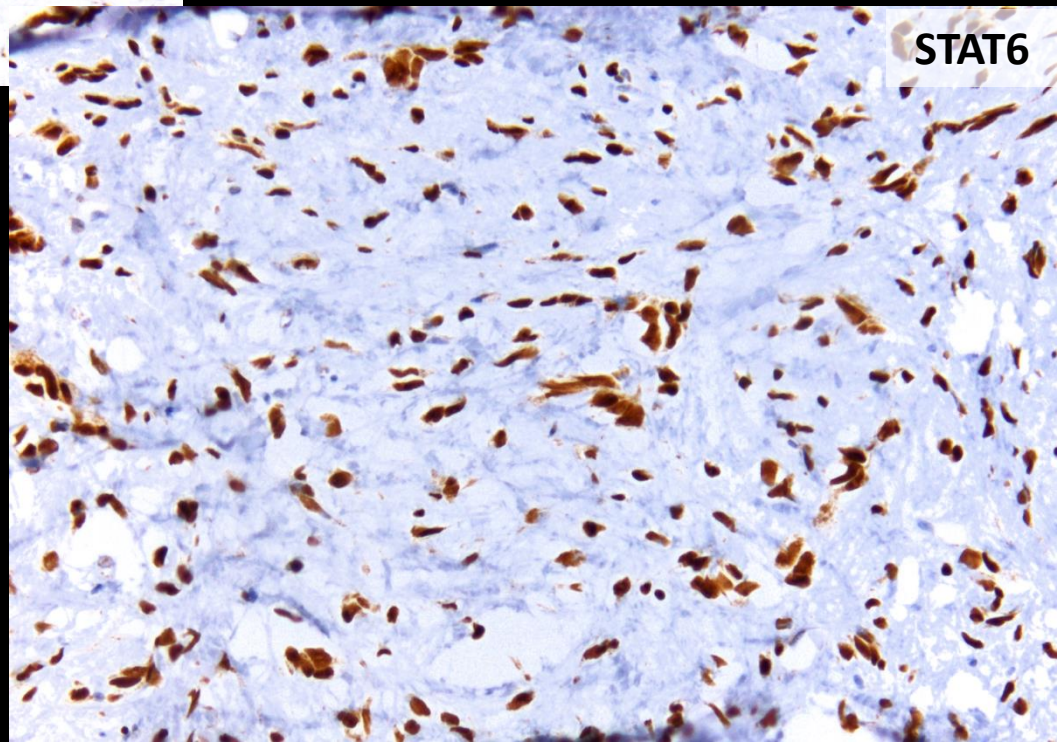
- Sézary syndrome:
  - rare (<5% of cutaneous lymphomas)
  - Triad: erythroderma, LAP, presence of Sézary cells (neoplastic T cells) in skin, LN & peripheral blood
  - Neoplastic cells are CD4+, CD4/CD8 ratio is expanded (>10)
  - Loss of other T cell antigens(±)
  - Aggressive course with 5 year survival of 10-20%
    - Prognostic factors: extent of LN and peripheral blood involvement

سین هفتم

- A 56 y/o male with recent diagnosis of large left retroperitoneal mass
- CT scan: 20 cm left retroperitoneal mass, displacing the left kidney and spleen
- A needle biopsy was performed




CD34

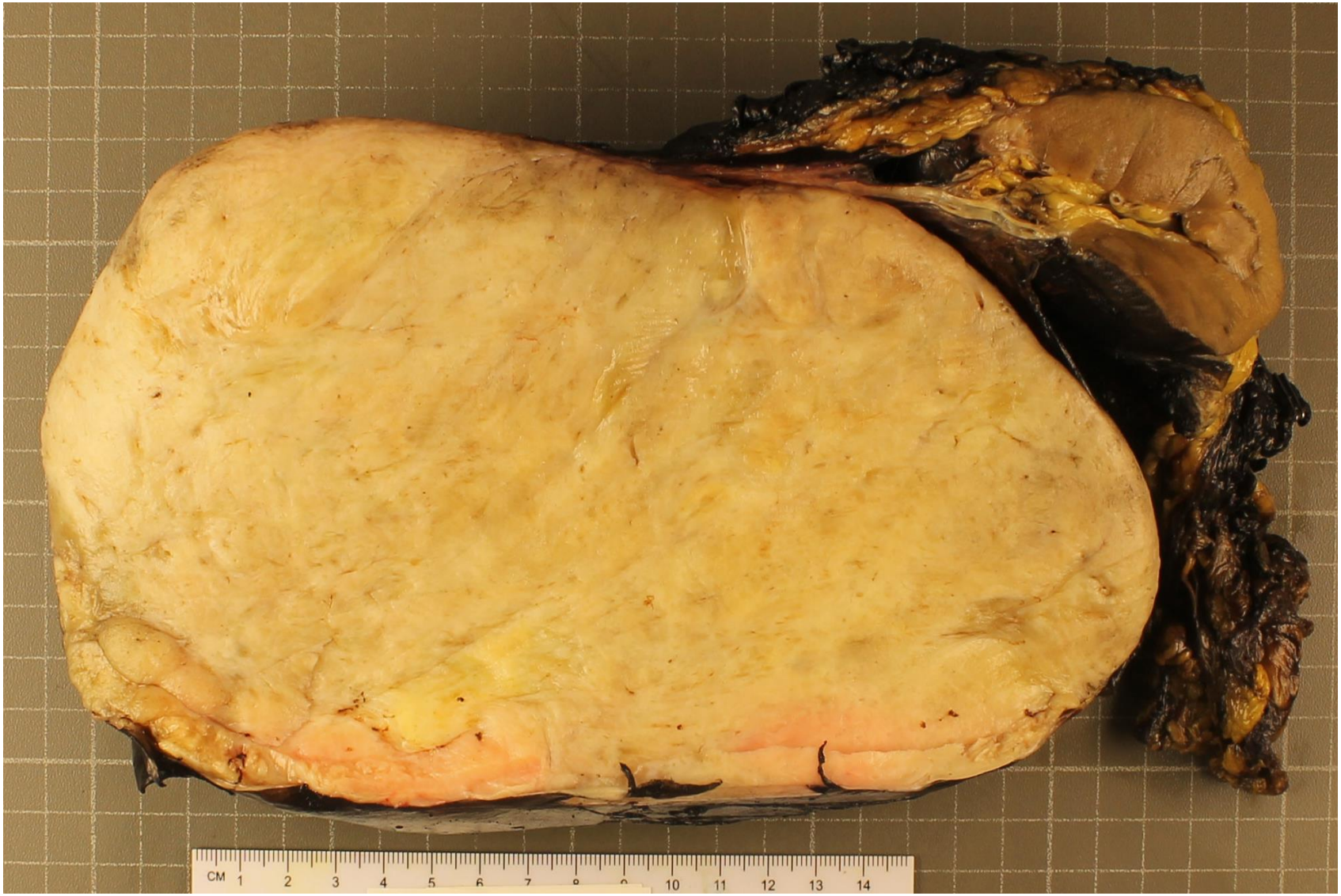

STAT6

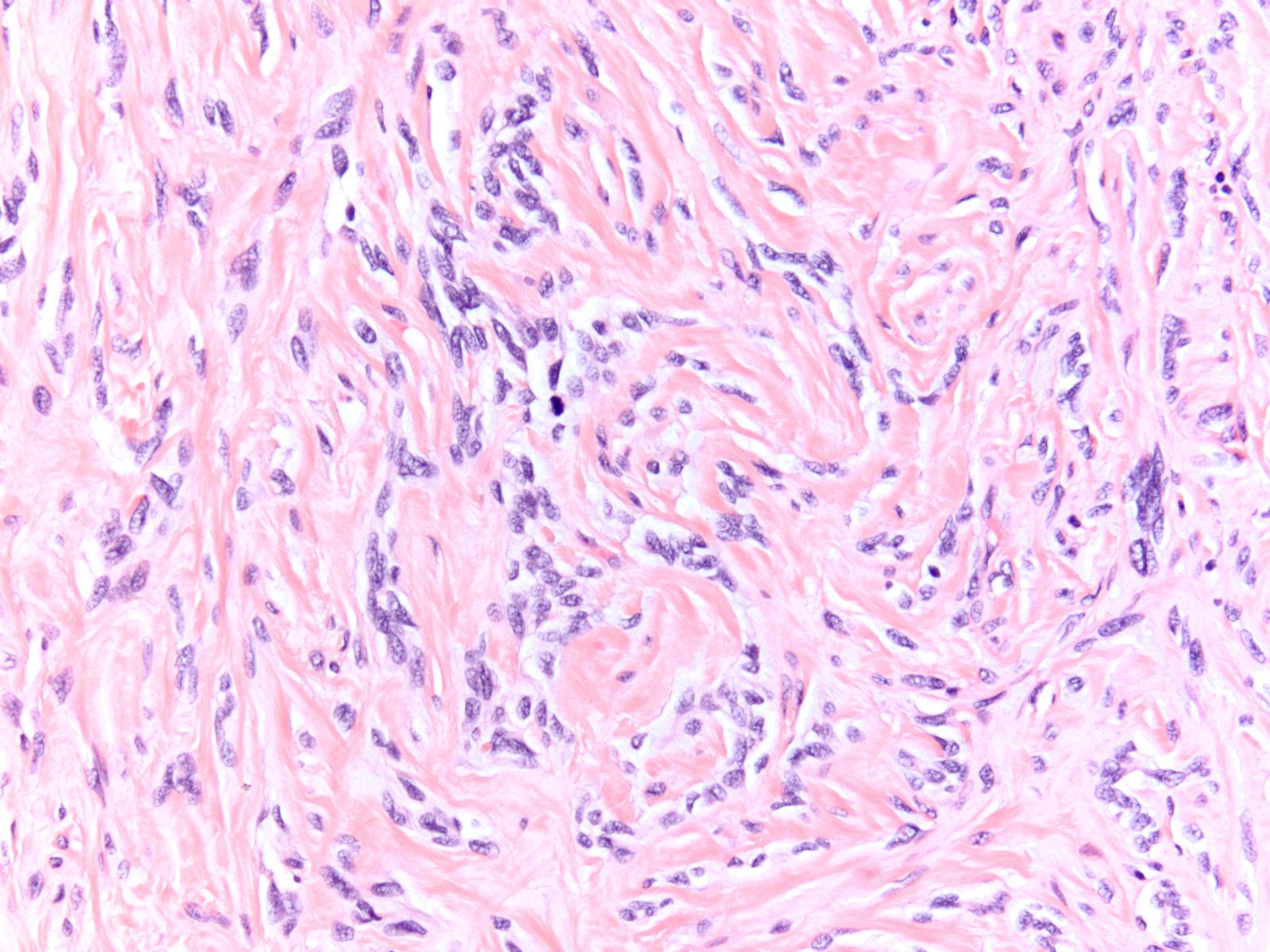
Biopsy result

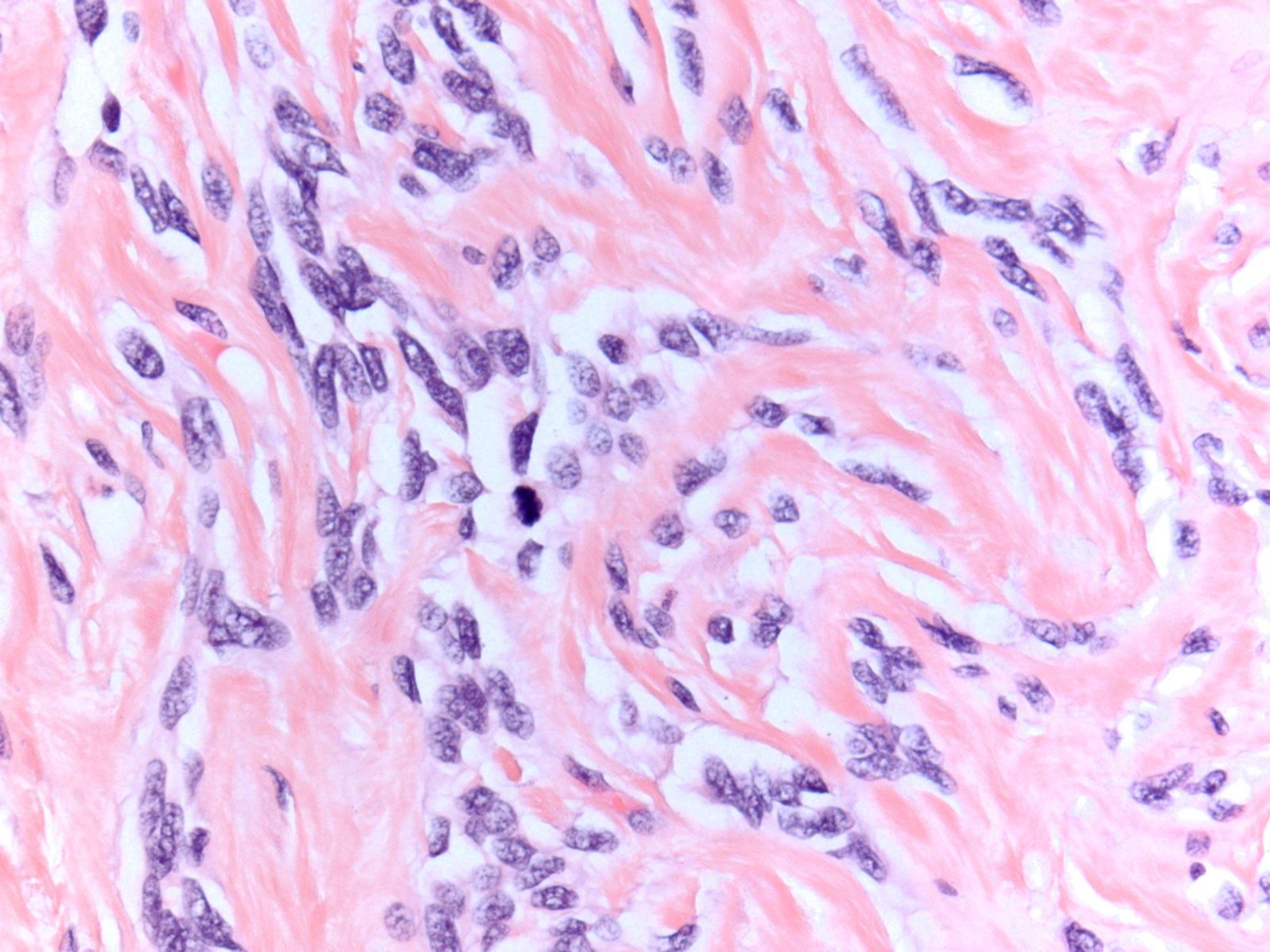
Spindle cell neoplasm, favor solitary fibrous tumor

Negative for: CK, MDM2, Desmin, SMA, myogenin, S100

At laparotomy, a 25cm mass in the kidney was removed.







Solitary fibrous tumor

- Rarely seen as a spindle cell tumor in kidney, of uncertain origin
- Usually presents as large renal tumor
- Histology: hemangiopericytoma-like vessels, patternless growth of spindle cells and fibrocollagenous stroma
- Novel fusion transcript: *NAB2-STAT6*
- Usually follows a benign clinical course; however aggressive behavior is reported.

نوروز پیروز!