



EMORY
UNIVERSITY
SCHOOL OF
MEDICINE

**Department of
Pathology and
Laboratory
Medicine**



Georgia Association of Pathology

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2020 Virtual Pathology Course

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46 year-old female with adnominal mass

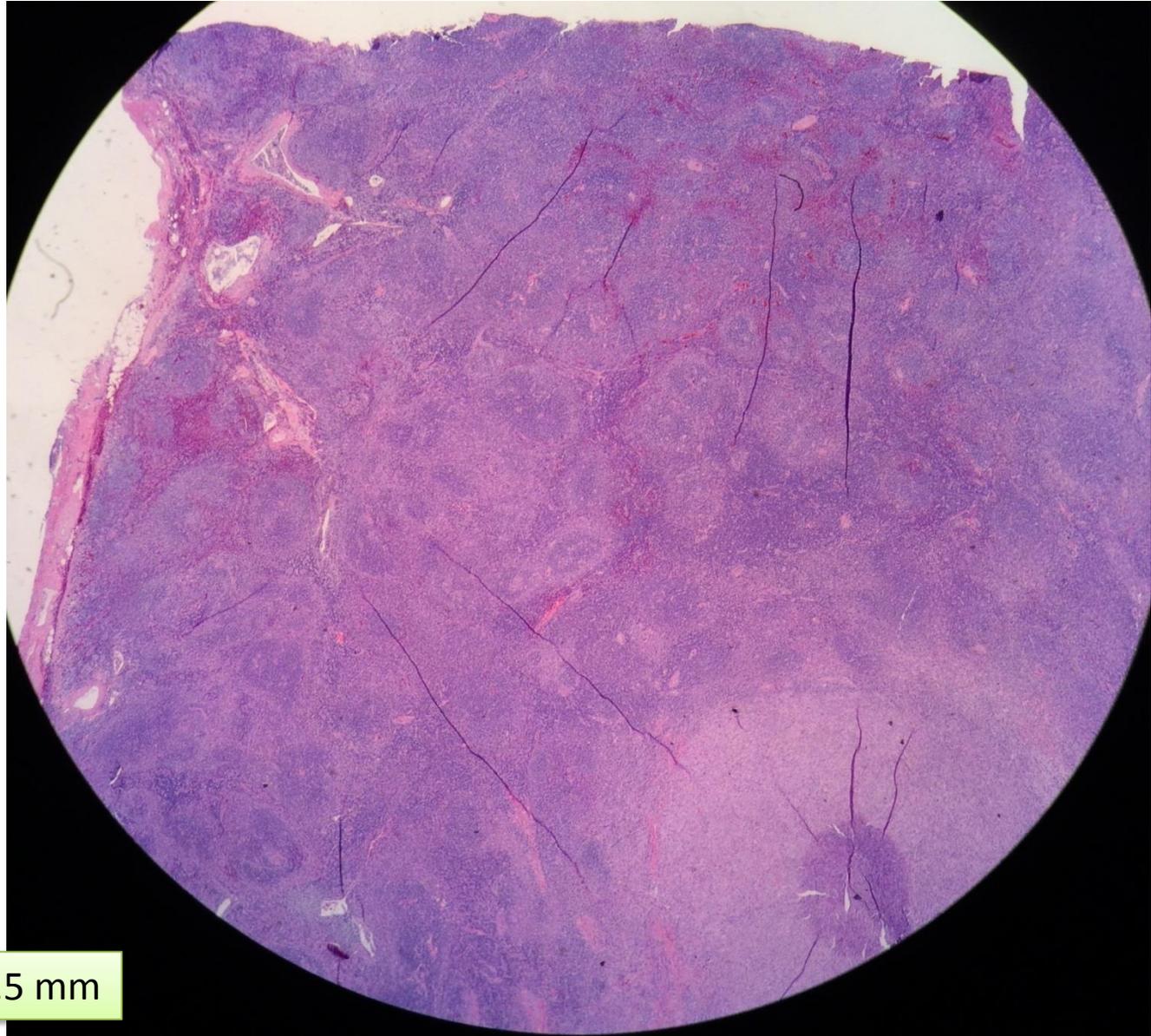
Linsheng Zhang, MD, PhD

Disclosure

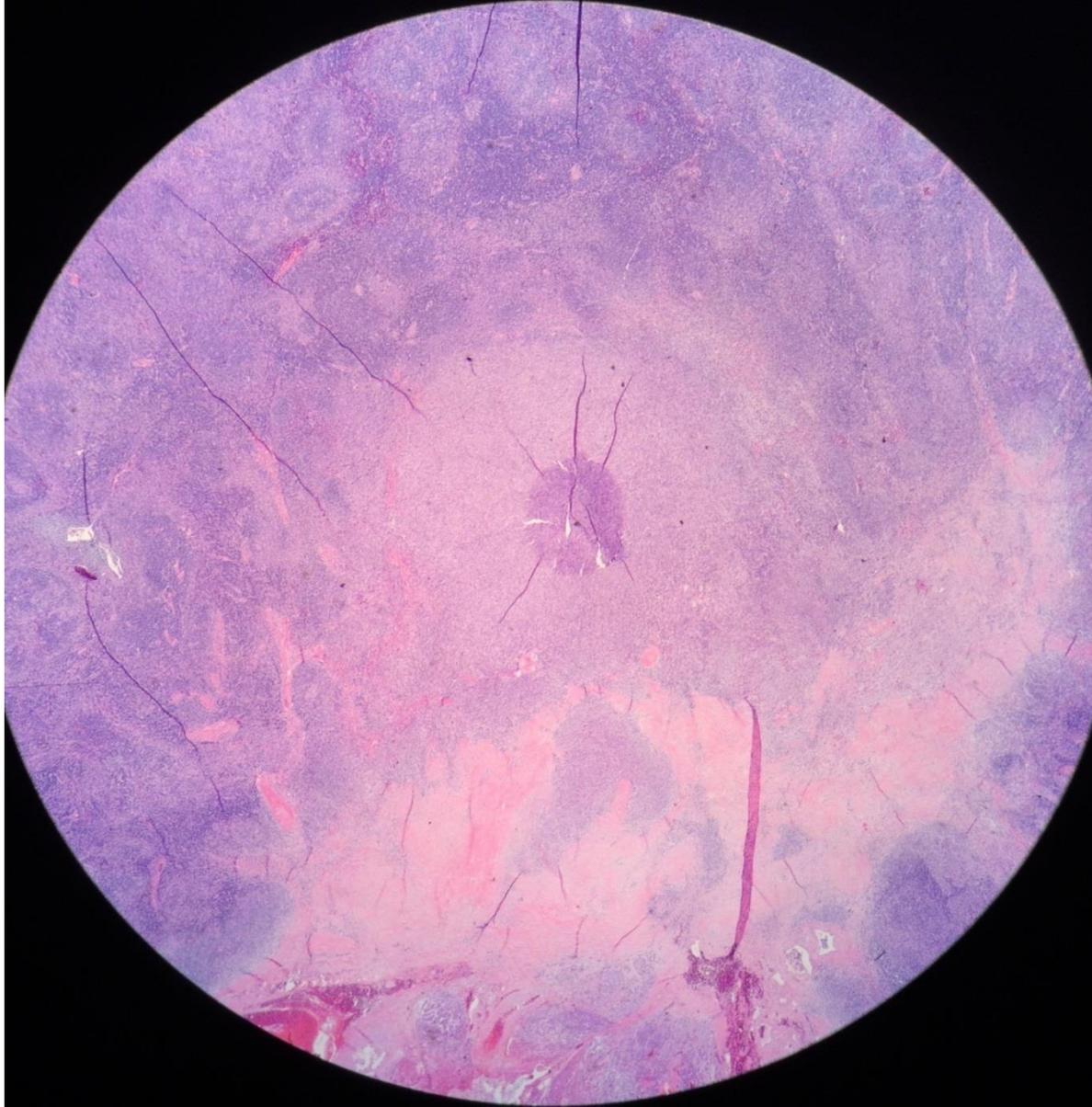
- No conflict of interest to disclose

Clinical history

- 46 year-old female with abdominal pain
- Untrasound of abdomen found a small mass
- Confirmed by MRI: a well circumscribed T2 hyperintense mass, 2.5 x 4.2 cm, cranial to an abutting junction of the pancreatic body and tail.
- Mass was resected (virtual slide).

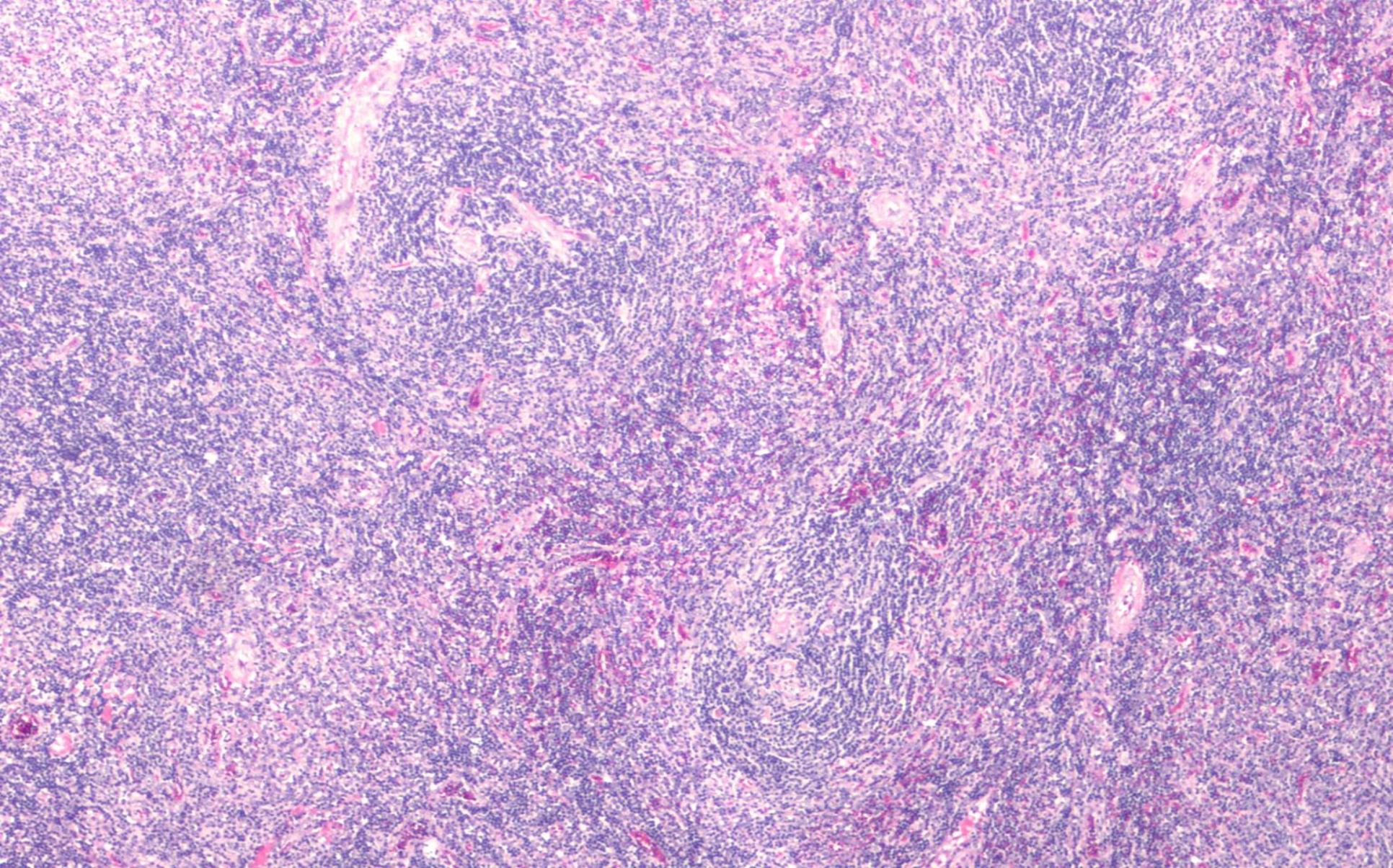


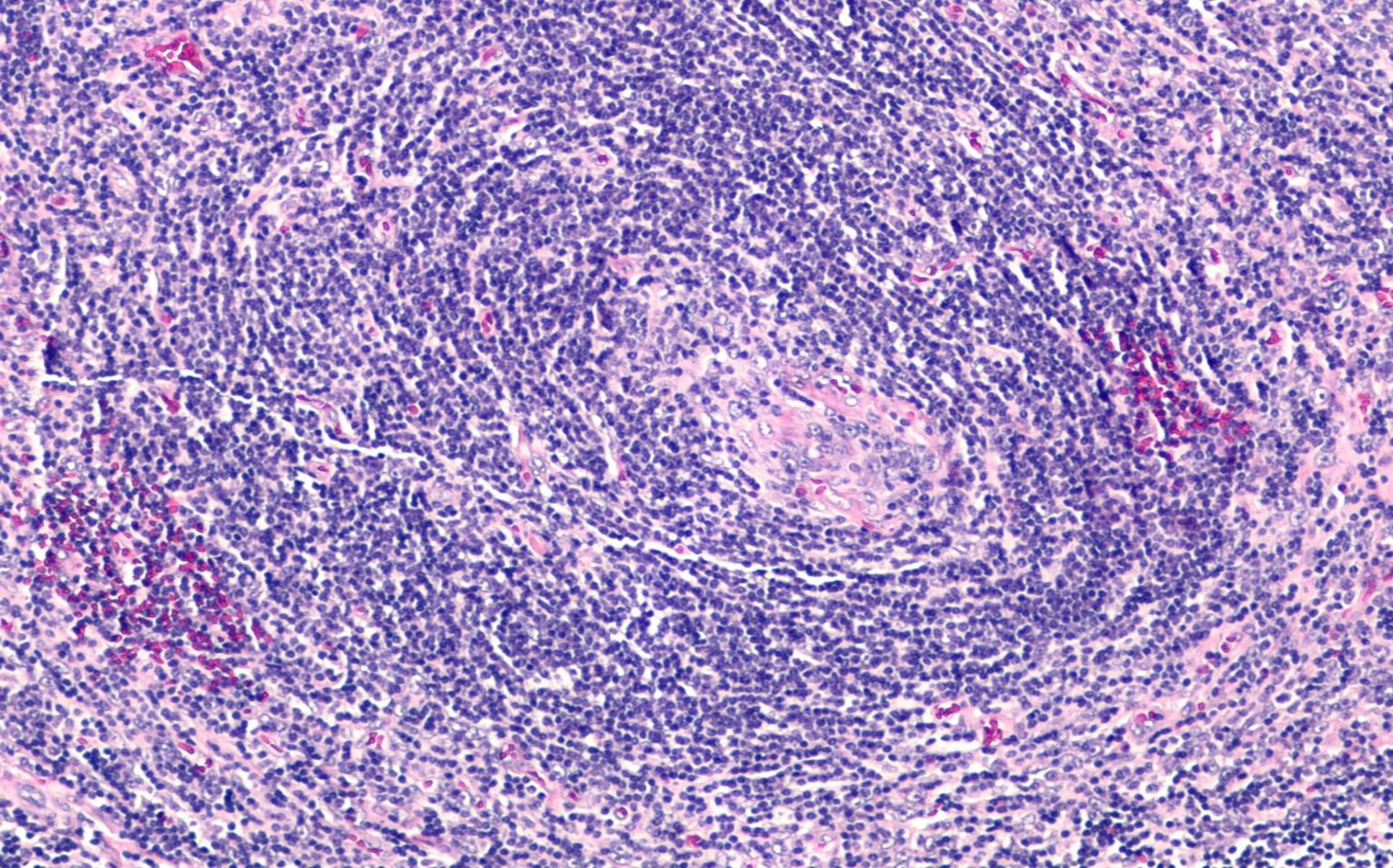
Maximal size 7.5 mm

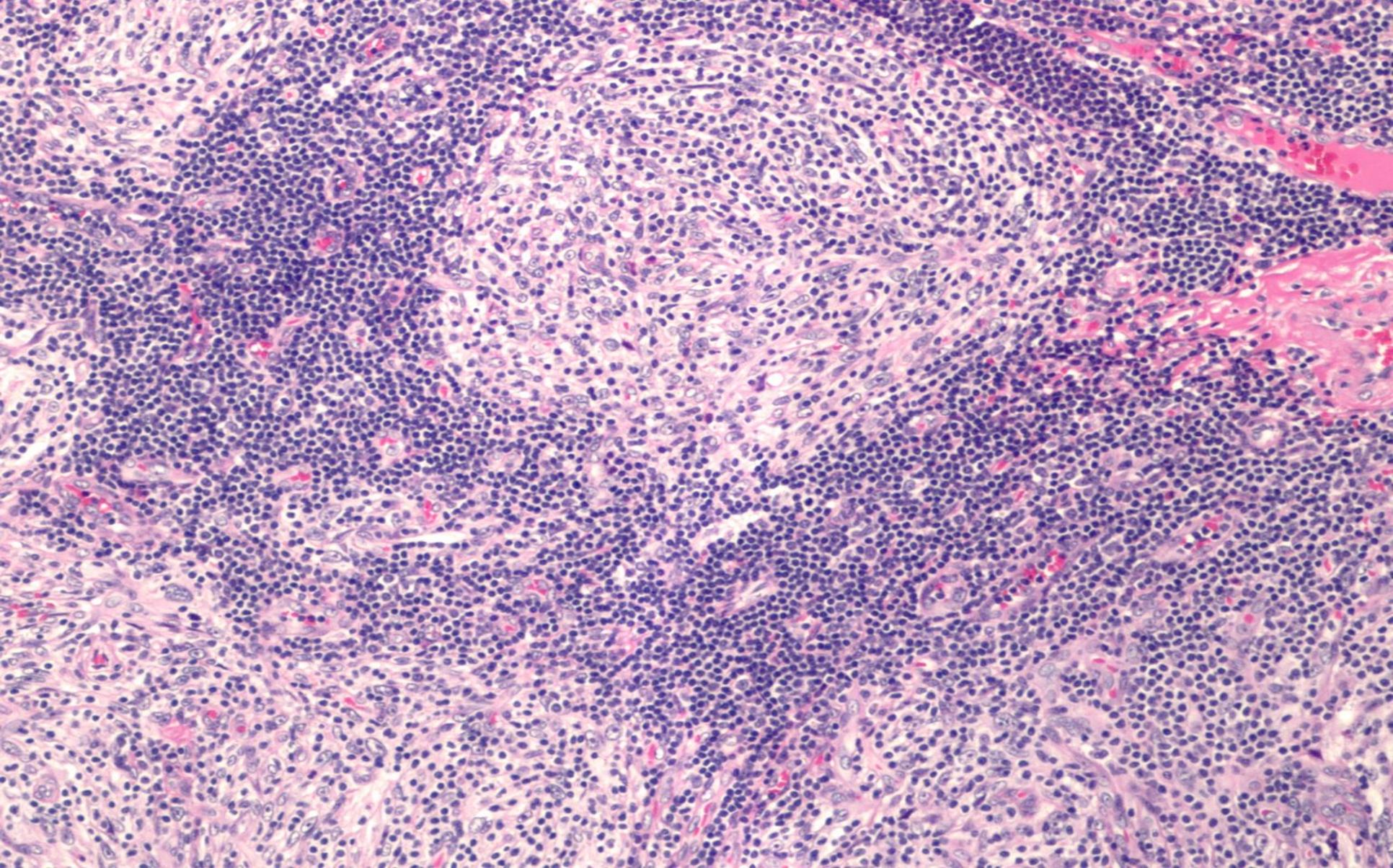


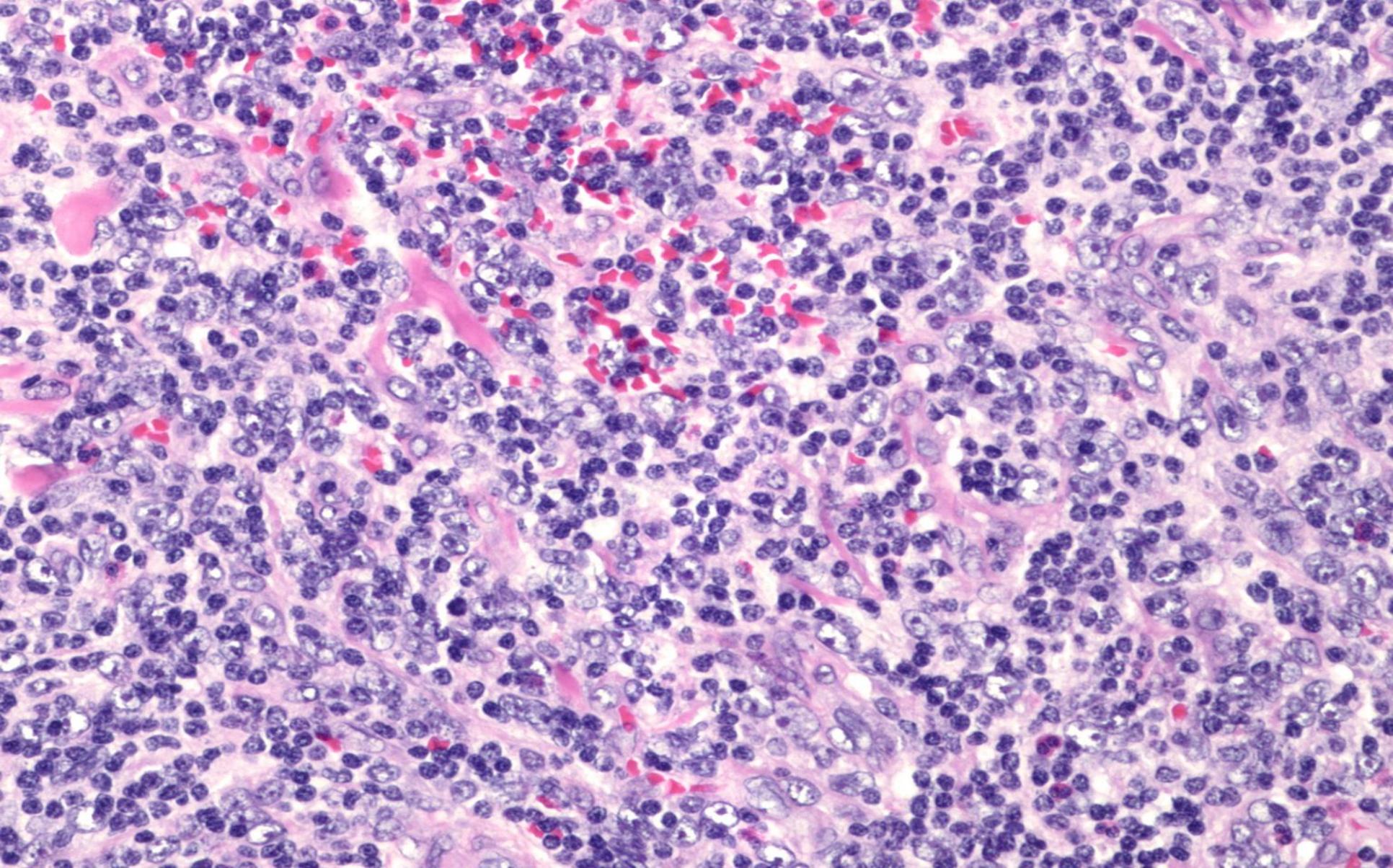
The specimen is entirely submitted for histology exam.

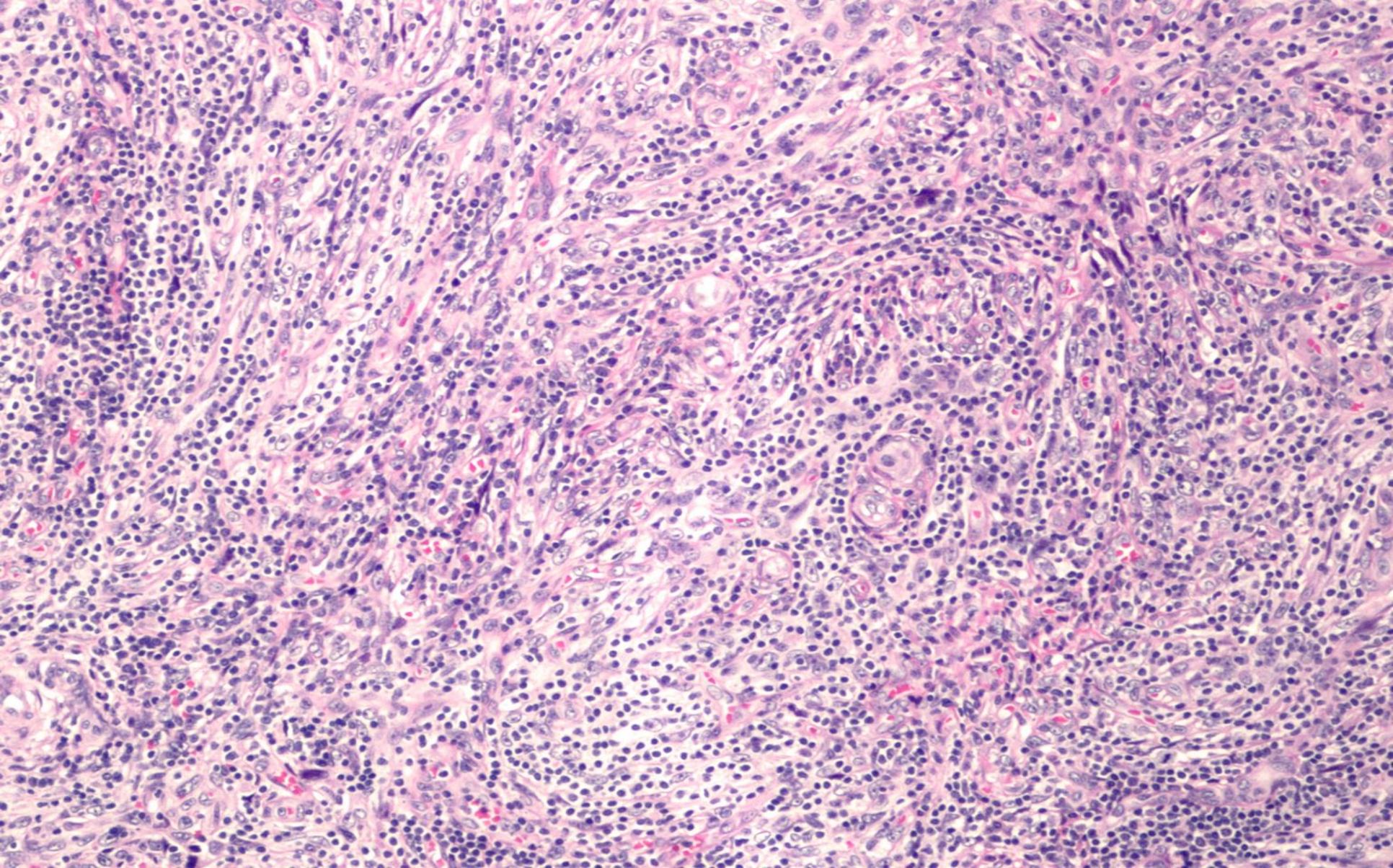
Maximum size of the nodule: 7.5 mm

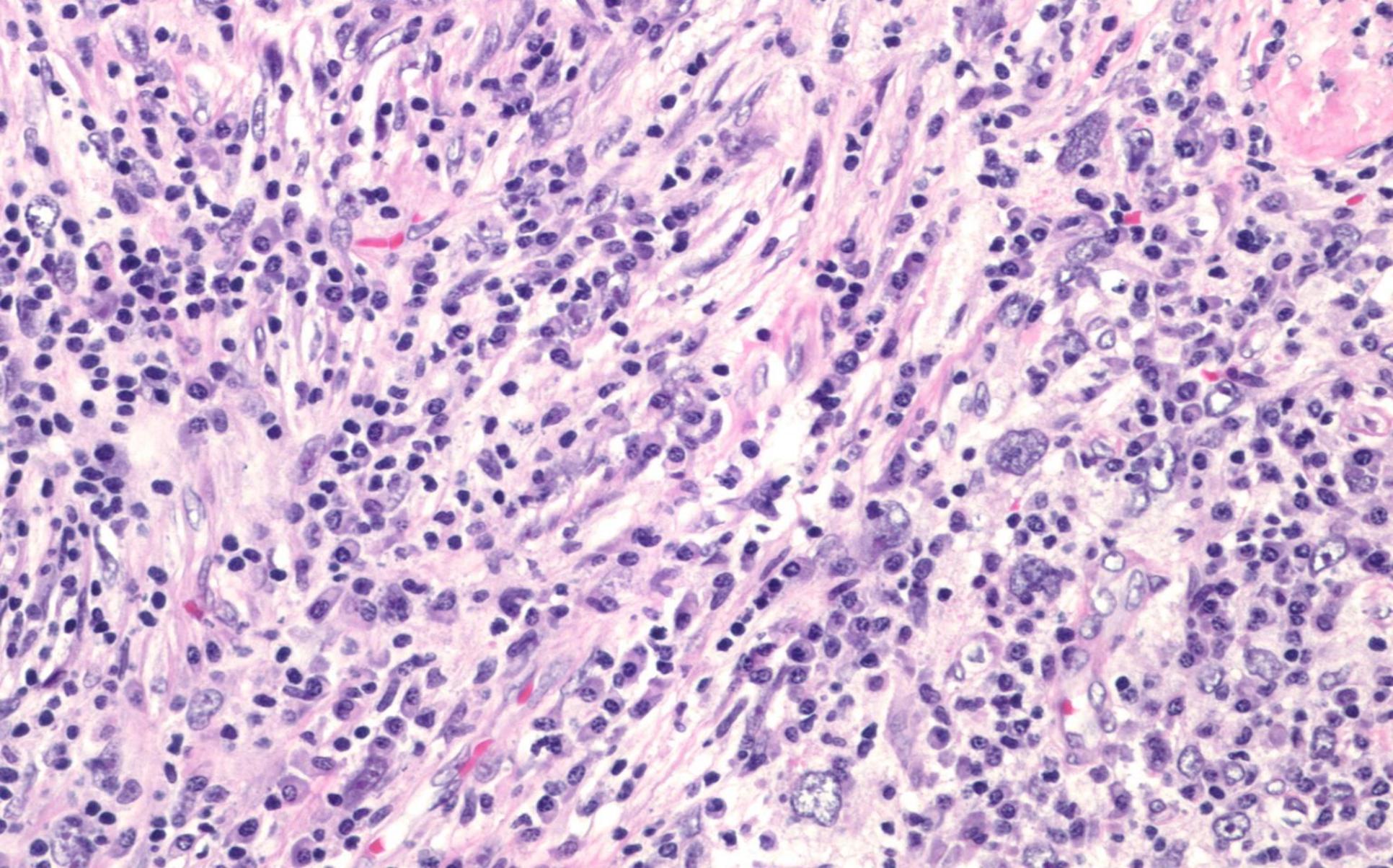


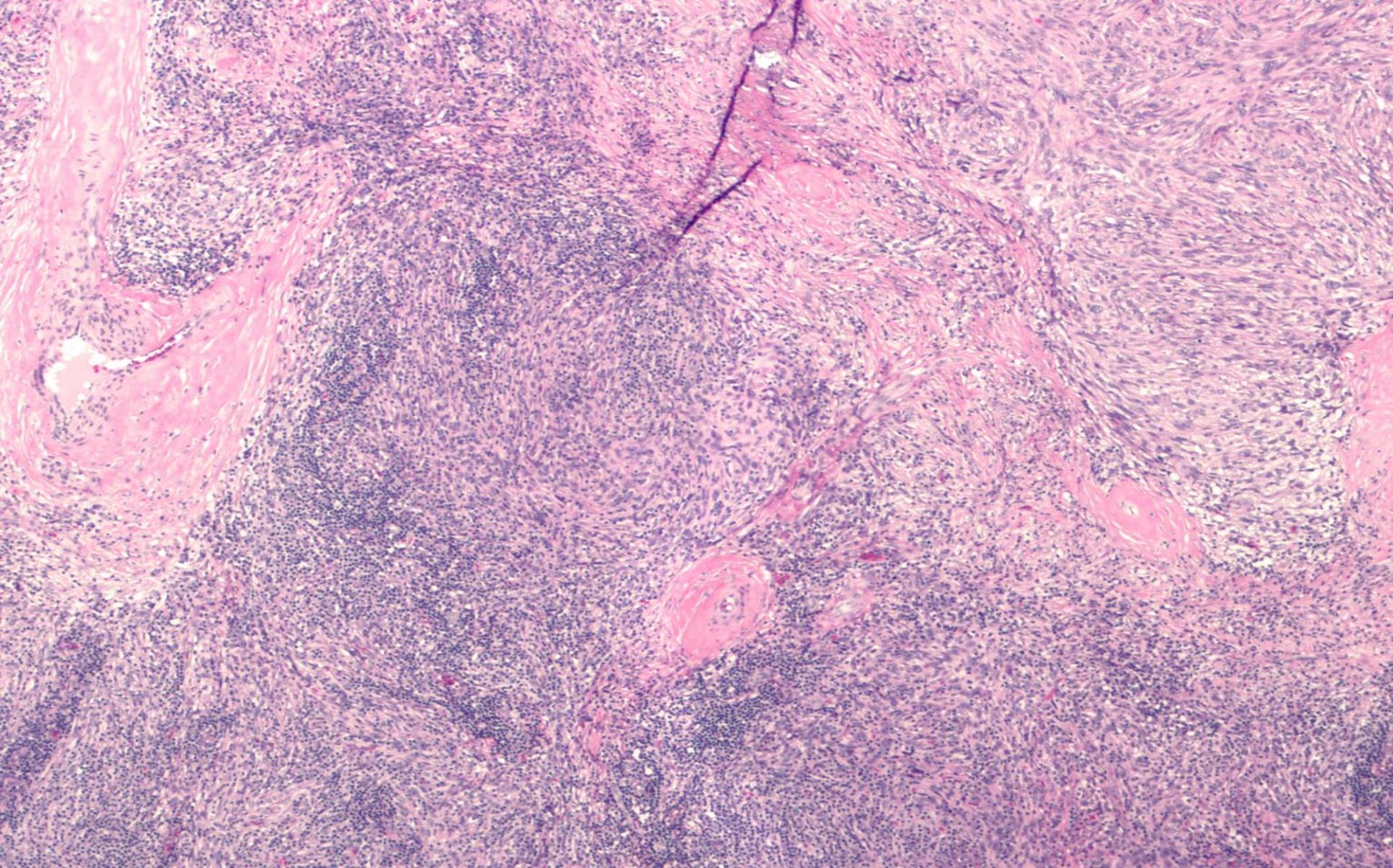


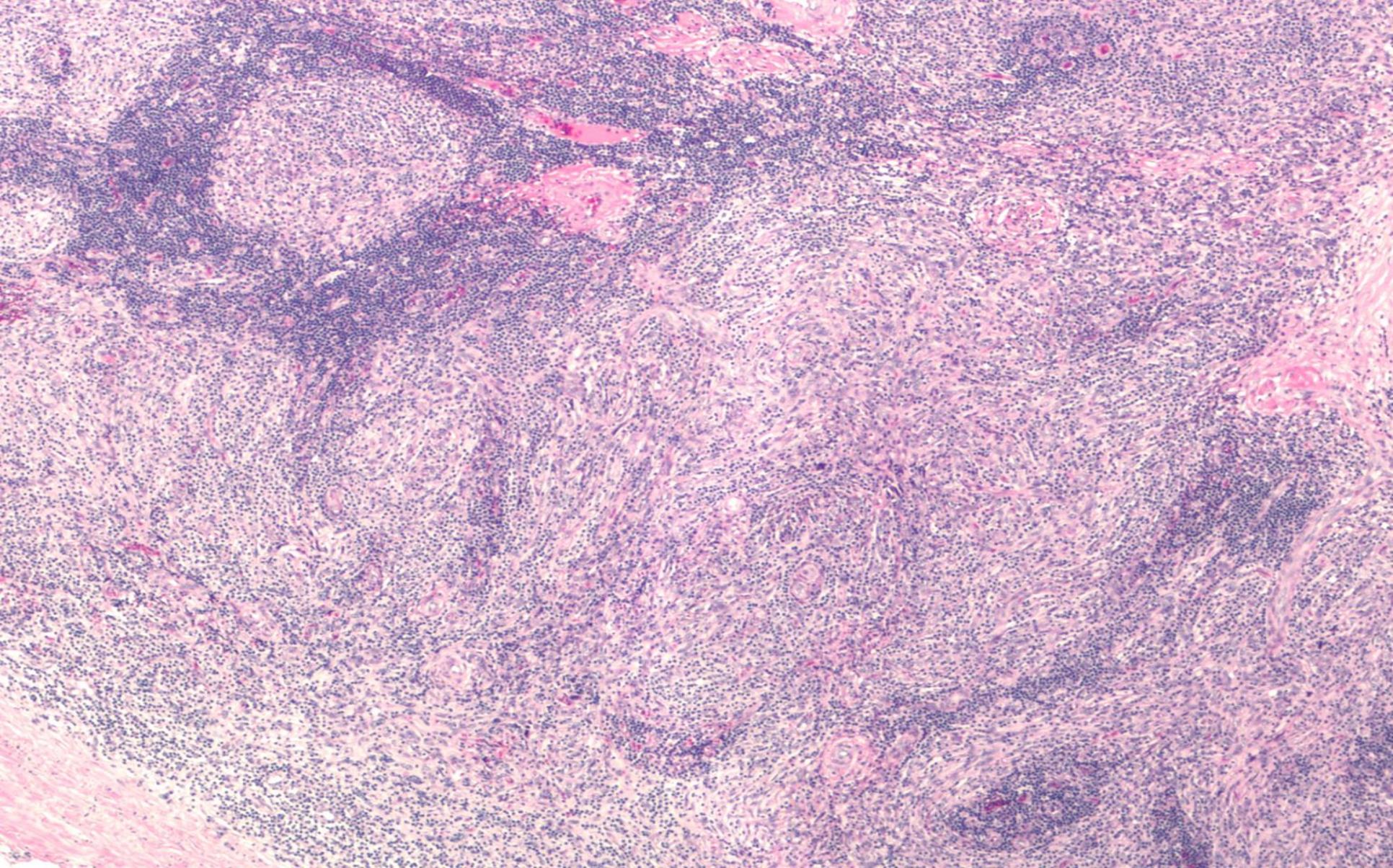


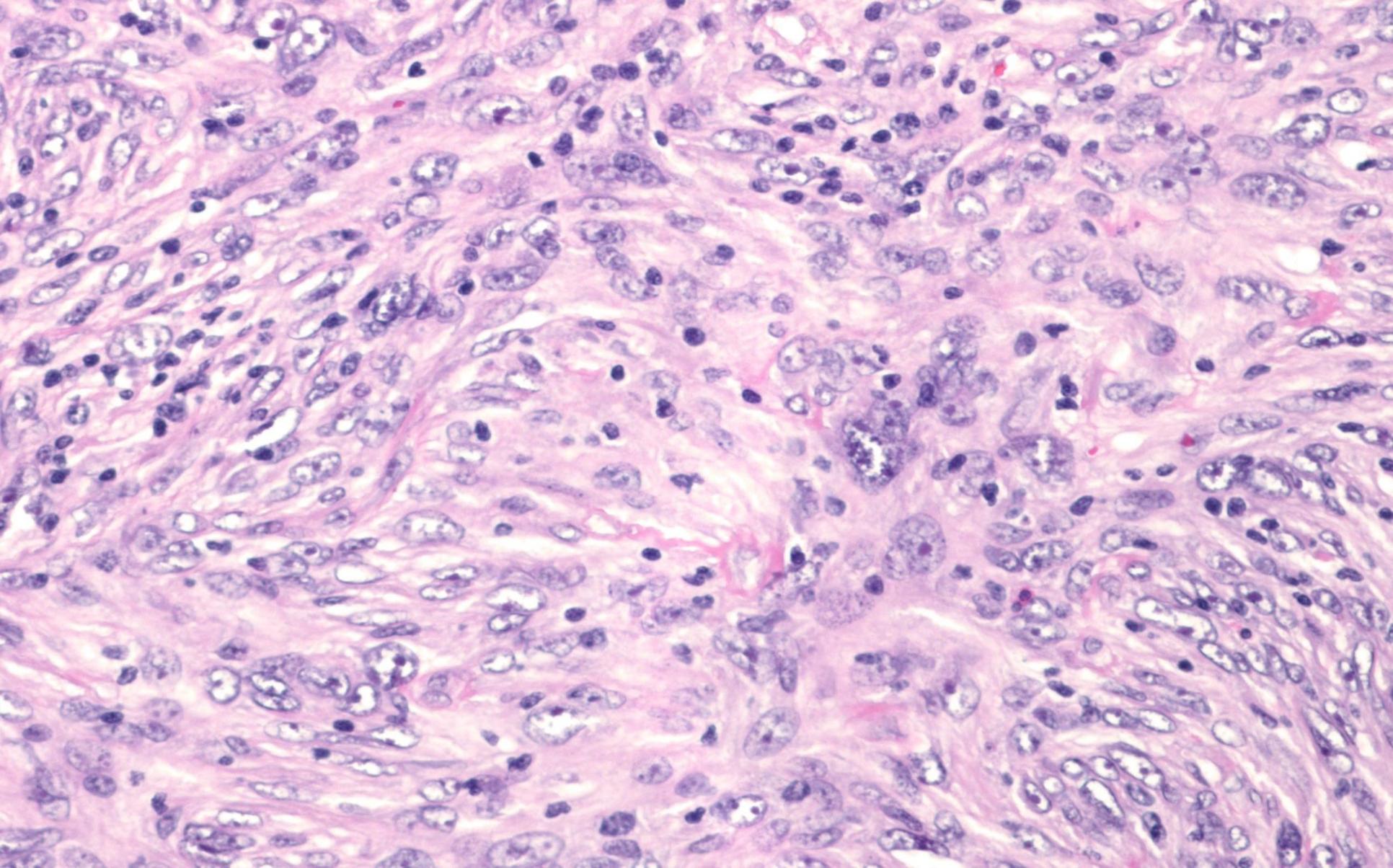


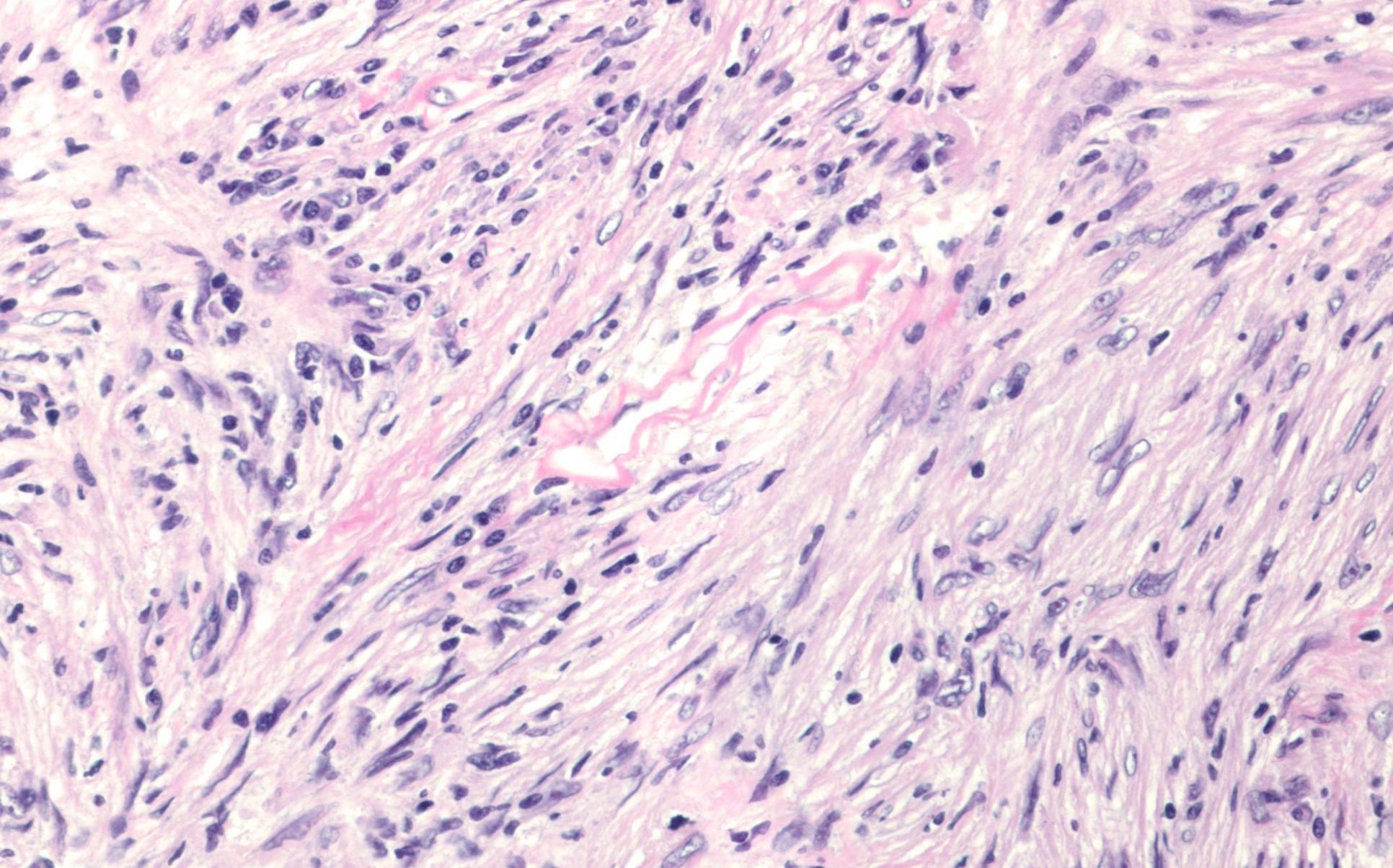


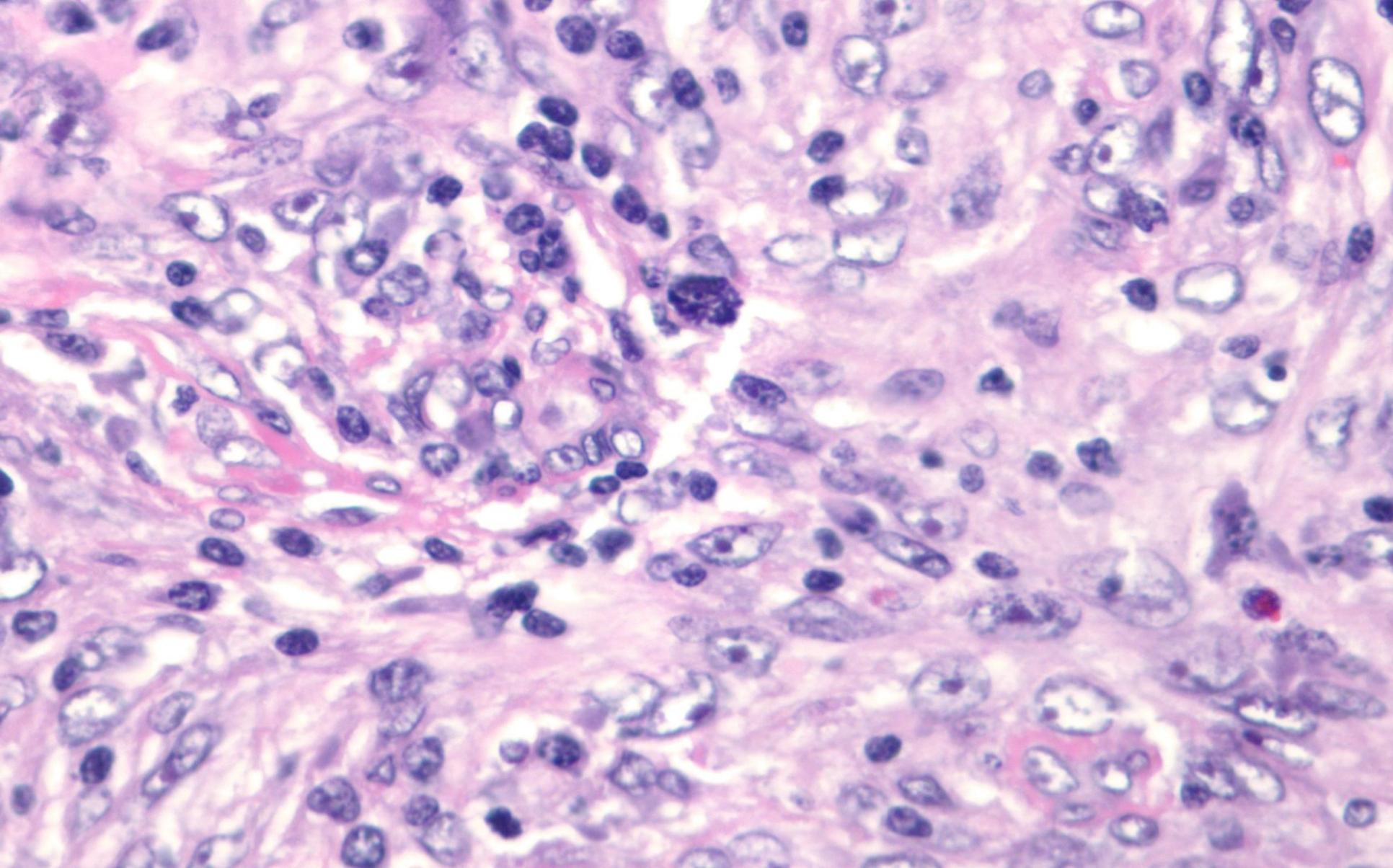




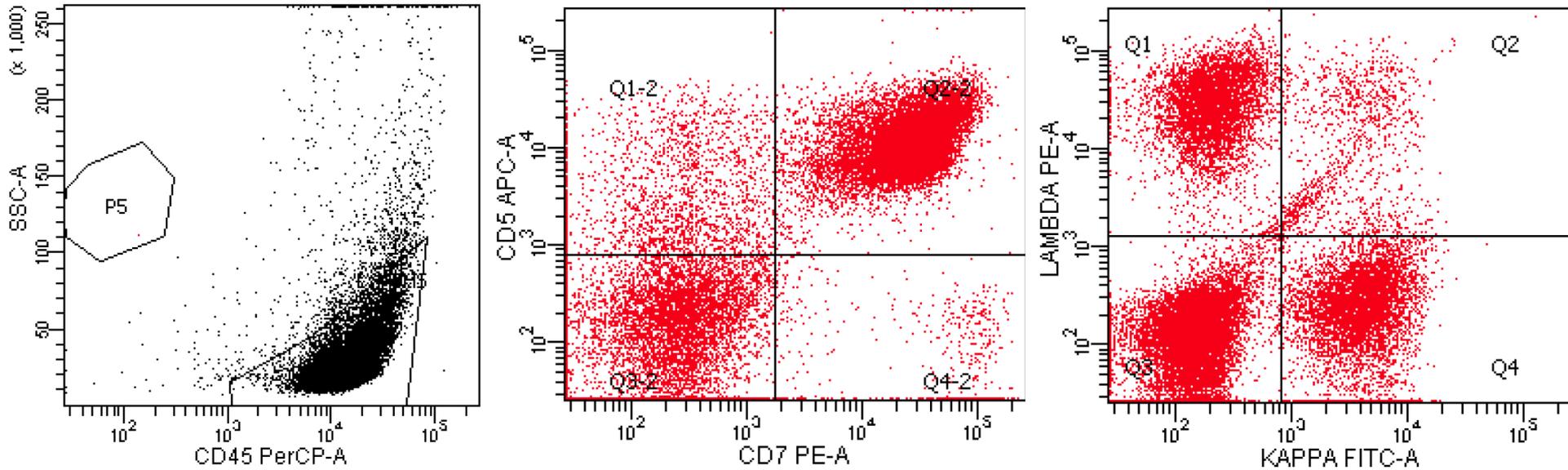








Flow Cytometry

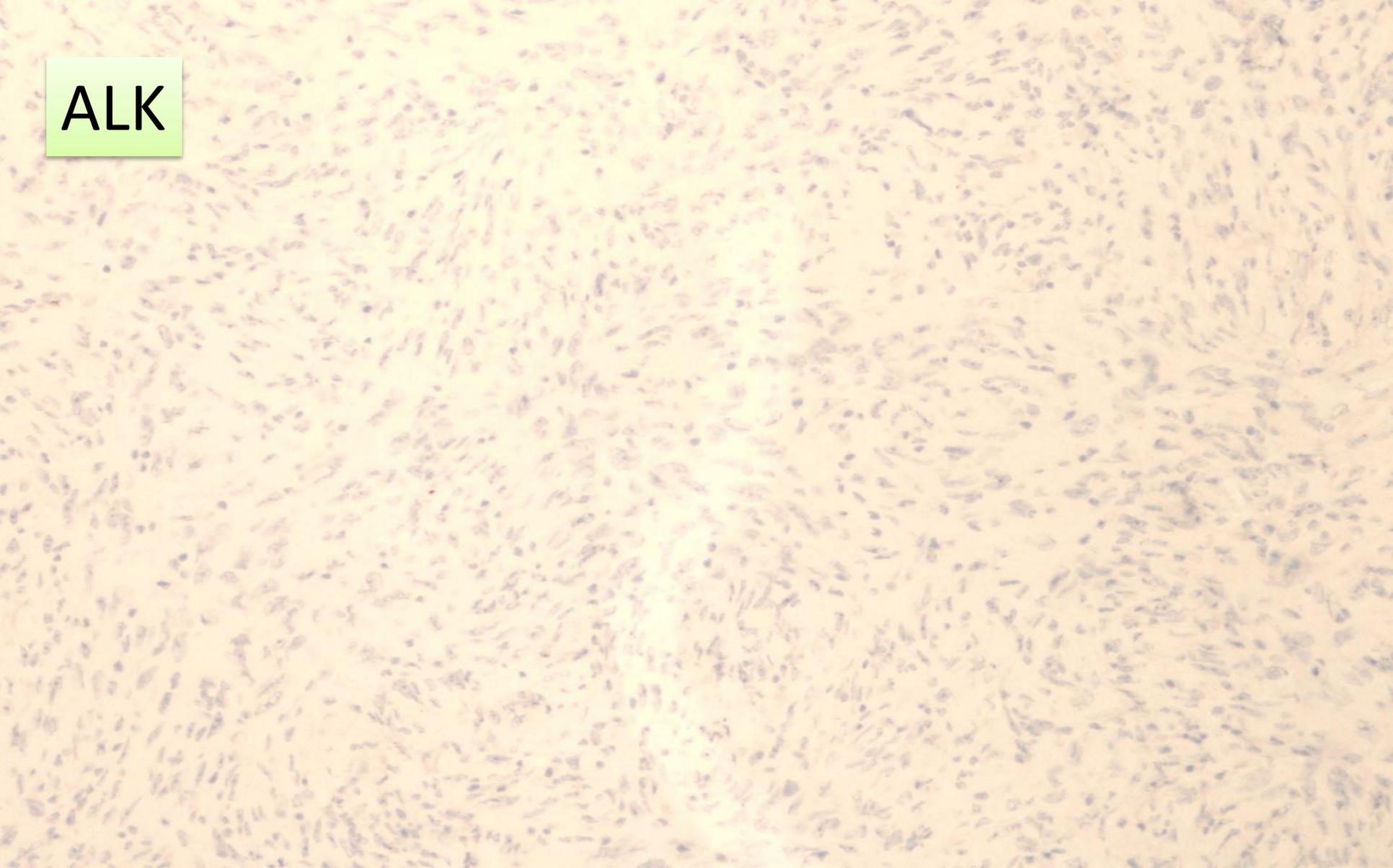


Mature lymphocytes > 97%. T-cells show no aberrant phenotype; B-cells are polytypic.

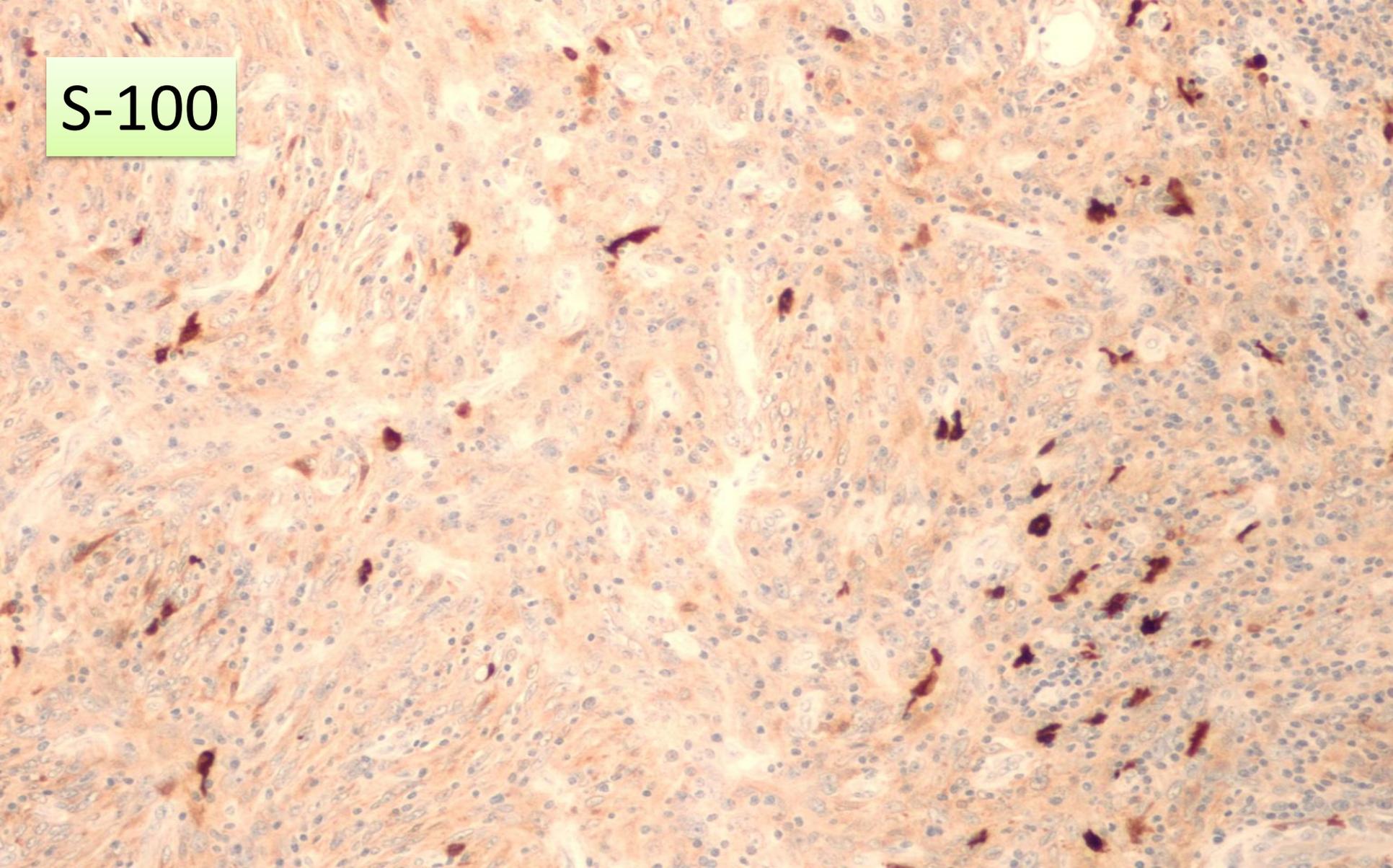
Differential Diagnosis

- **Follicular dendritic cell sarcoma**
- **Interdigitating dendritic cell sarcoma**
- Intranodal palisaded myofibroblastoma
(prominent hemorrhage, amianthoid fibers)
- Inflammatory myofibroblastic tumor
- Angiomatoid fibrous histiocytoma
(Circumscribed, fibrous pseudocapsule)
- Lymphoepithelioma-like carcinoma
- Metastatic malignancies:
 - Melanoma
 - Spindle cell carcinoma
 - Gastrointestinal stromal tumor (GIST).
 - Malignant peripheral nerve sheath tumors

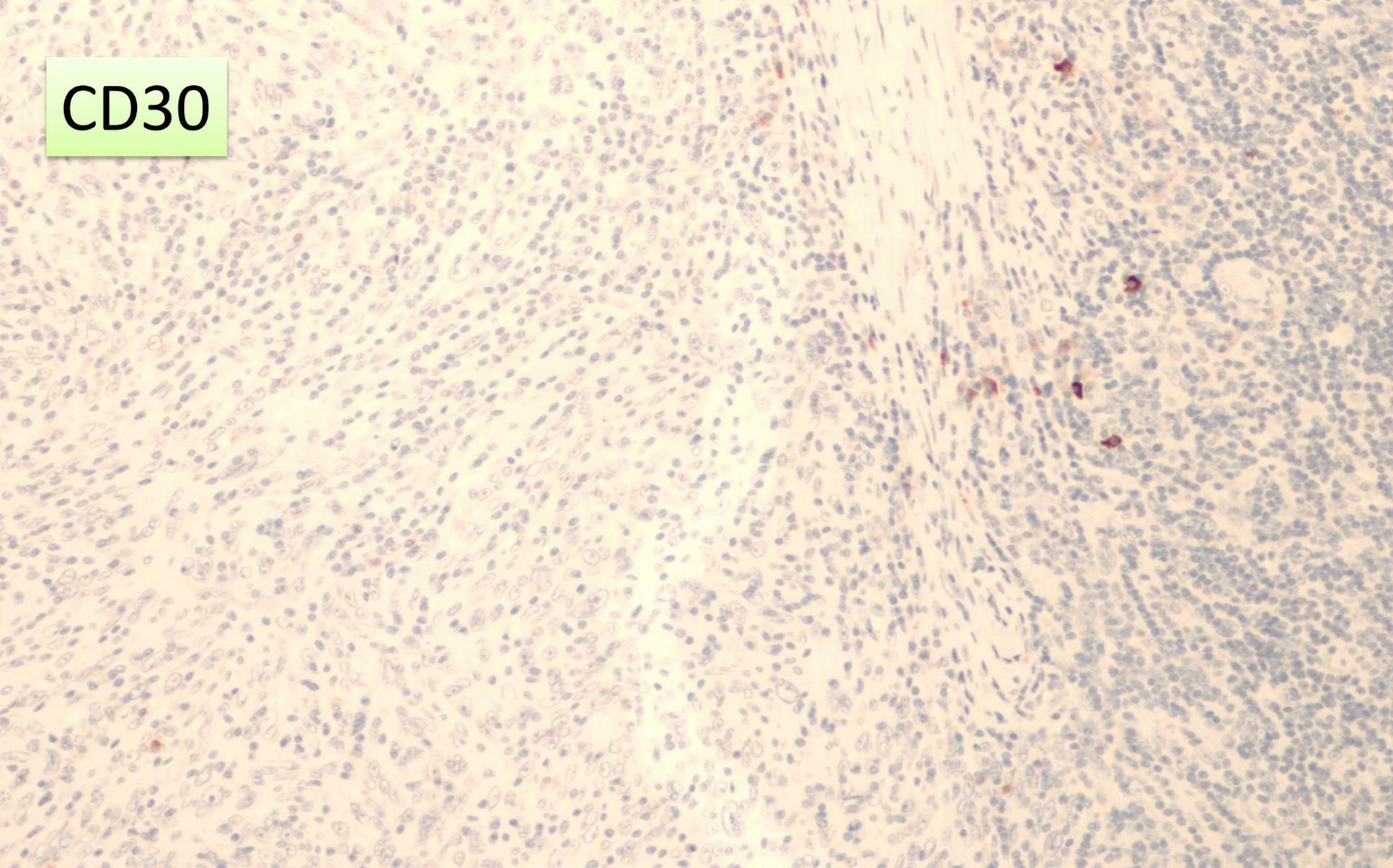
ALK



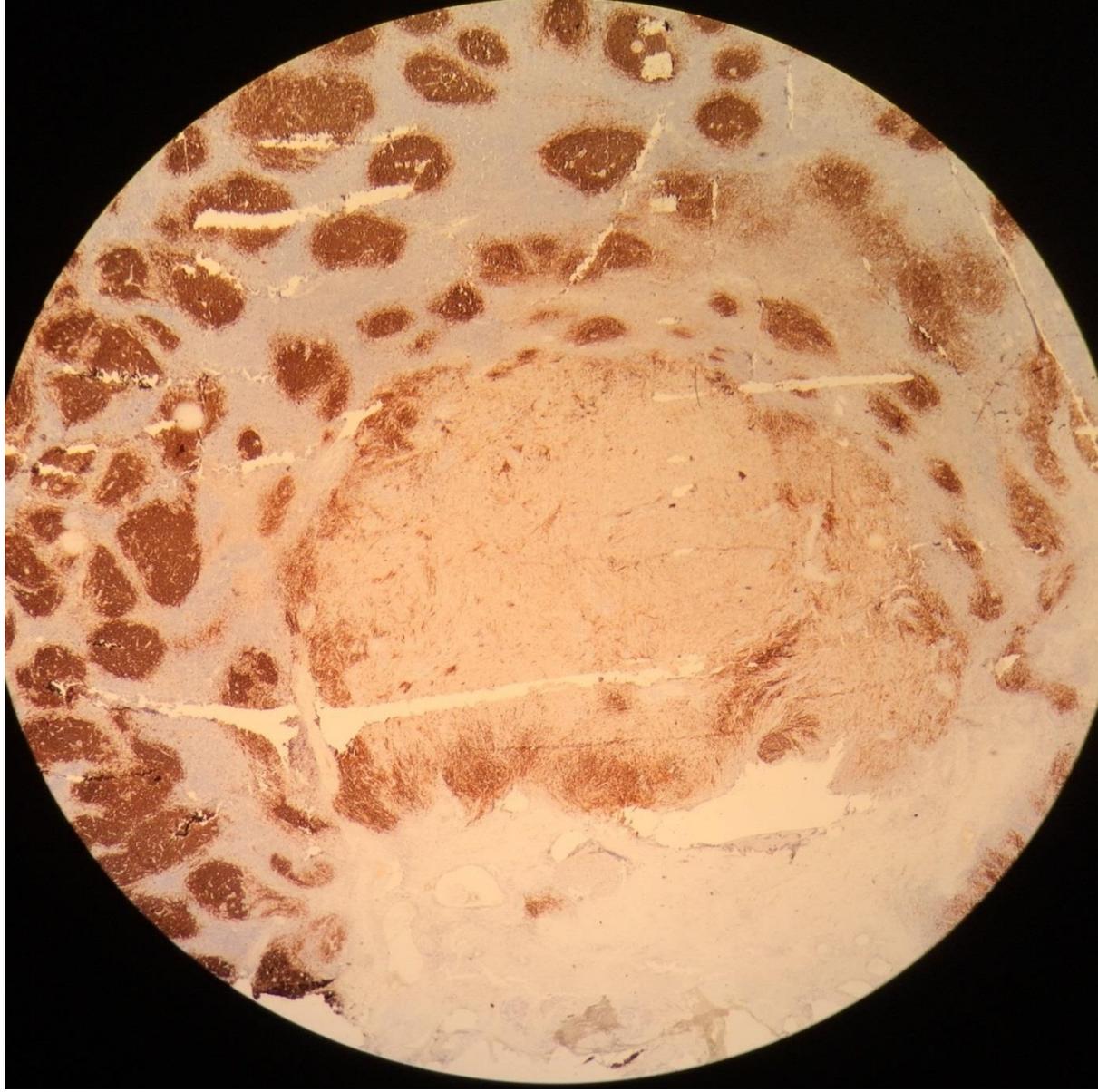
S-100

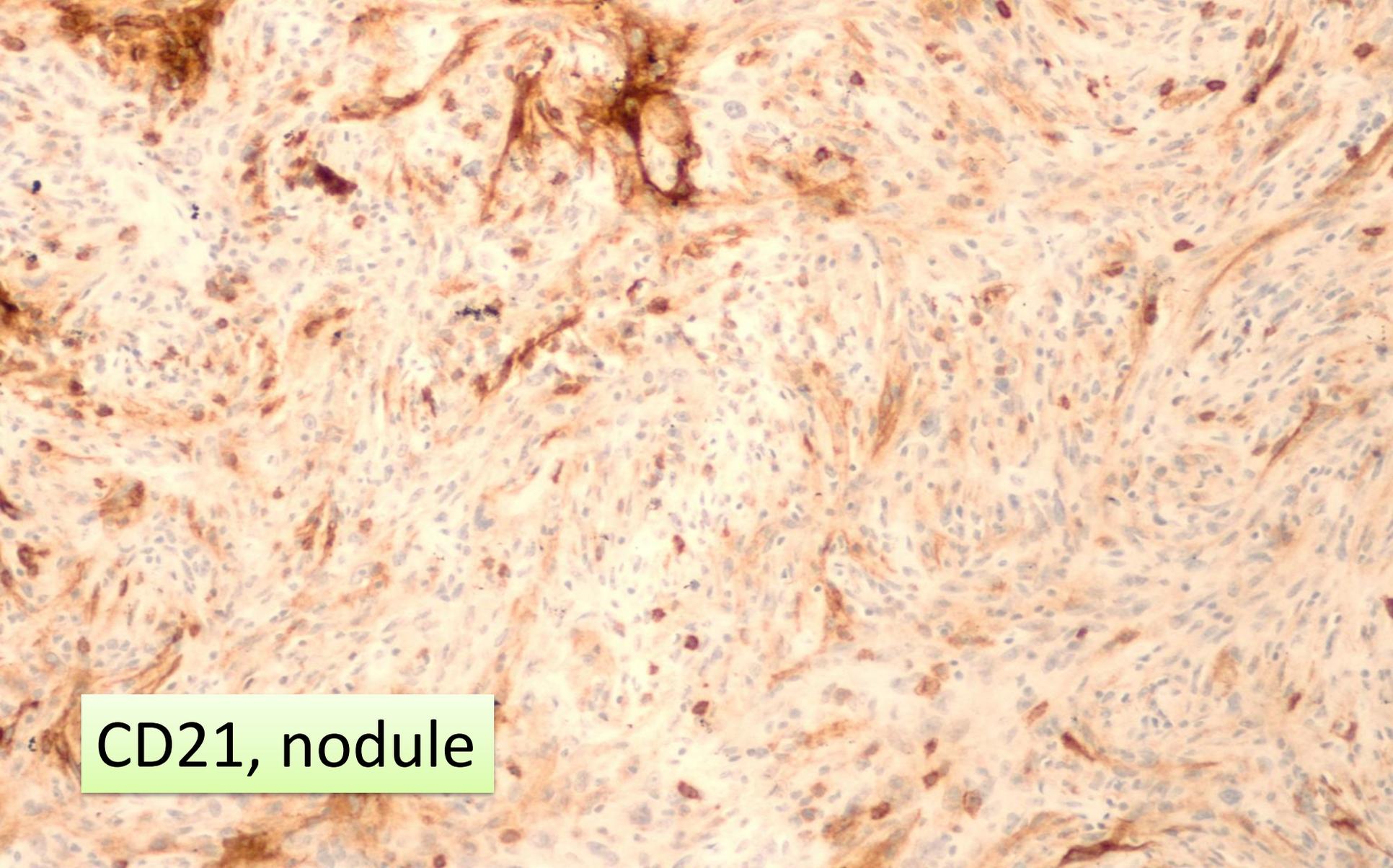


CD30



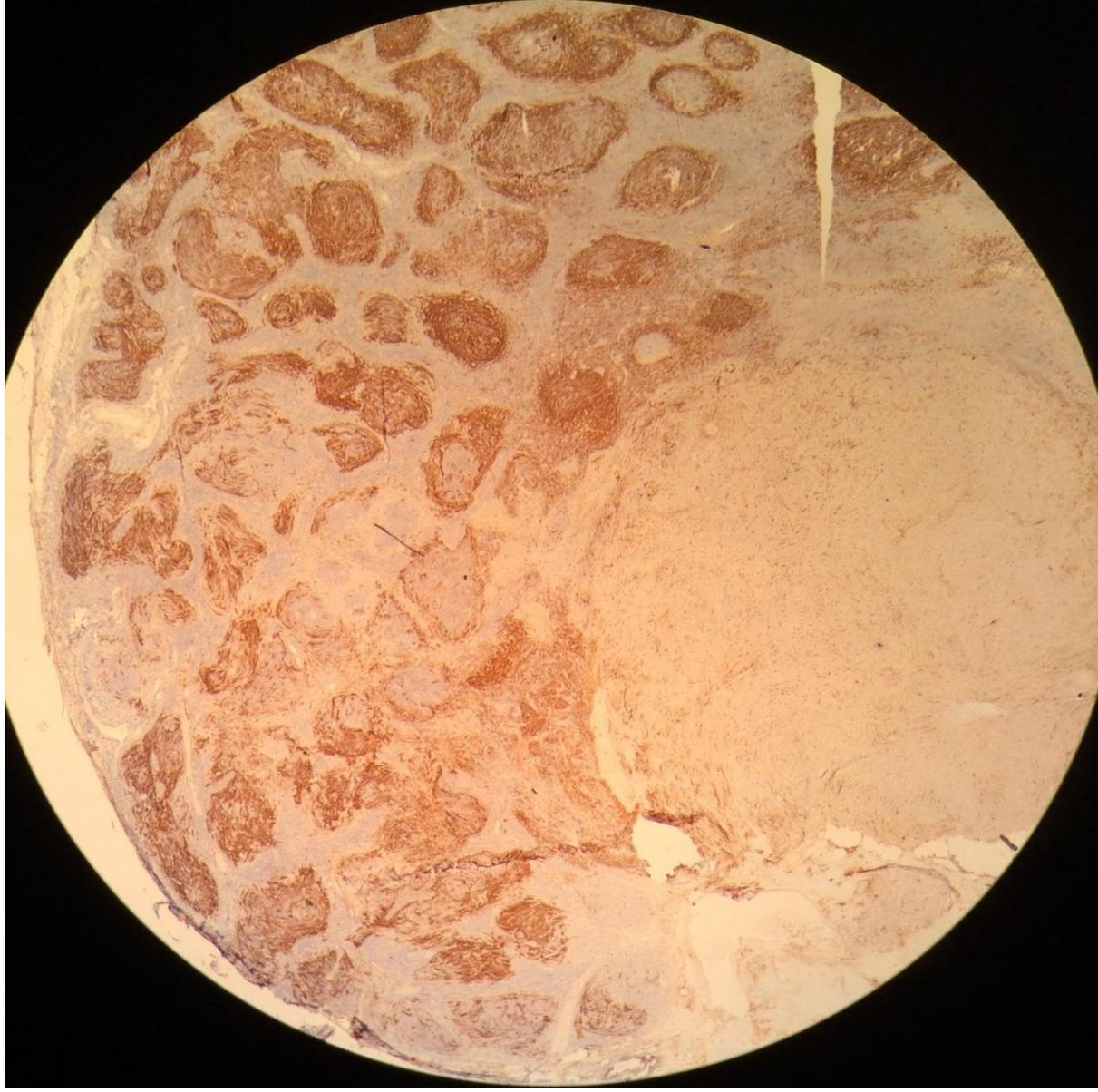
CD21



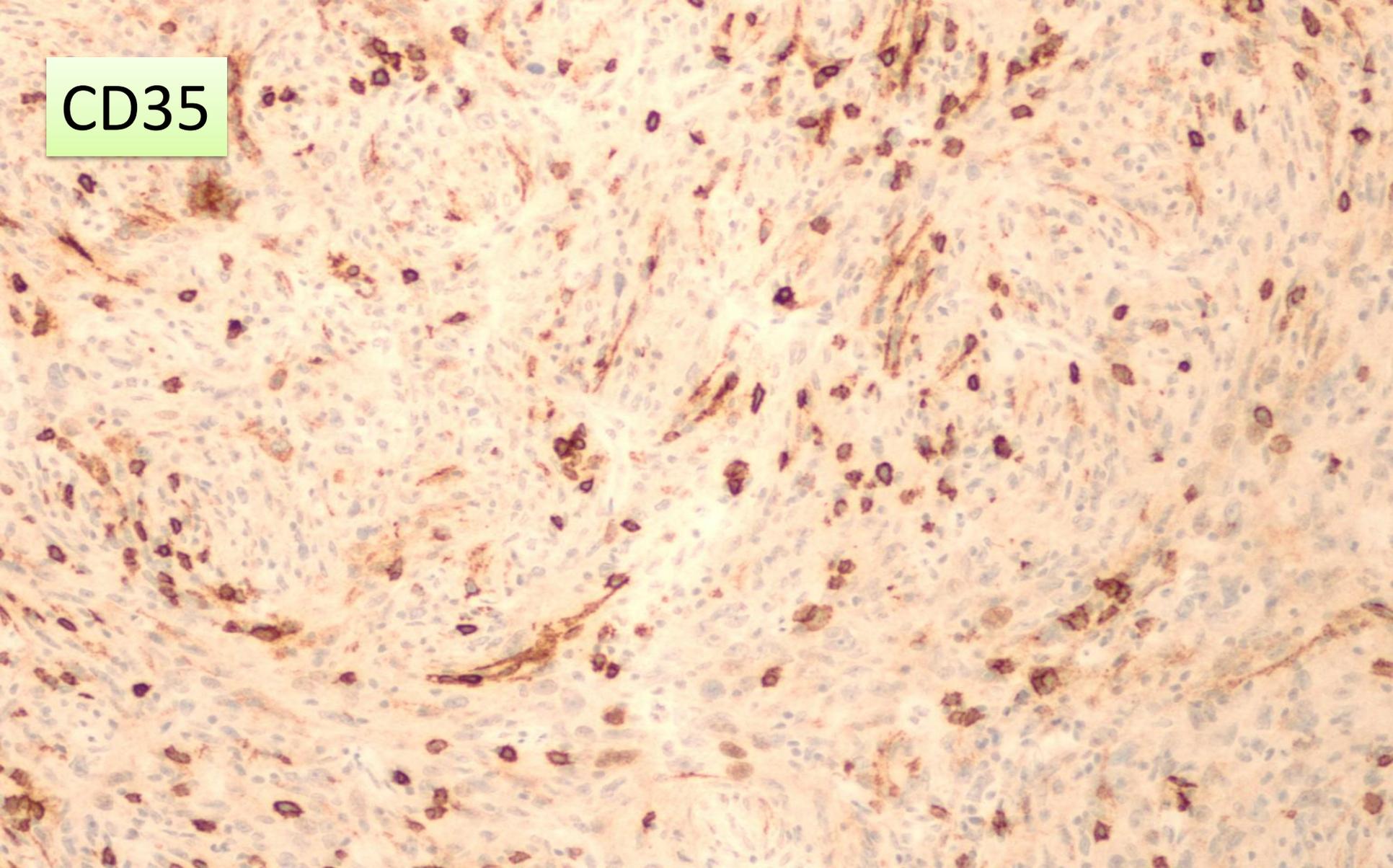


CD21, nodule

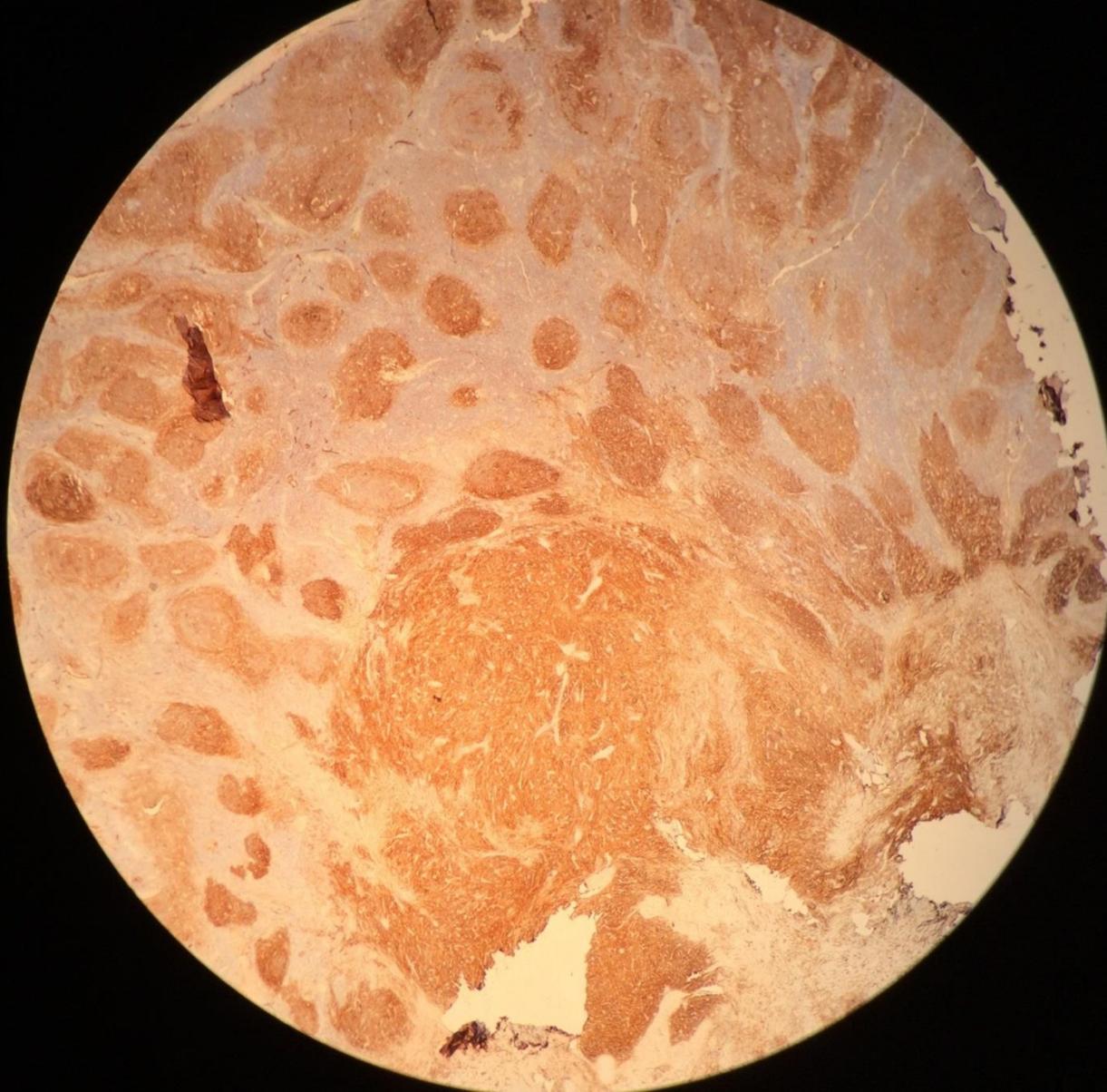
CD35



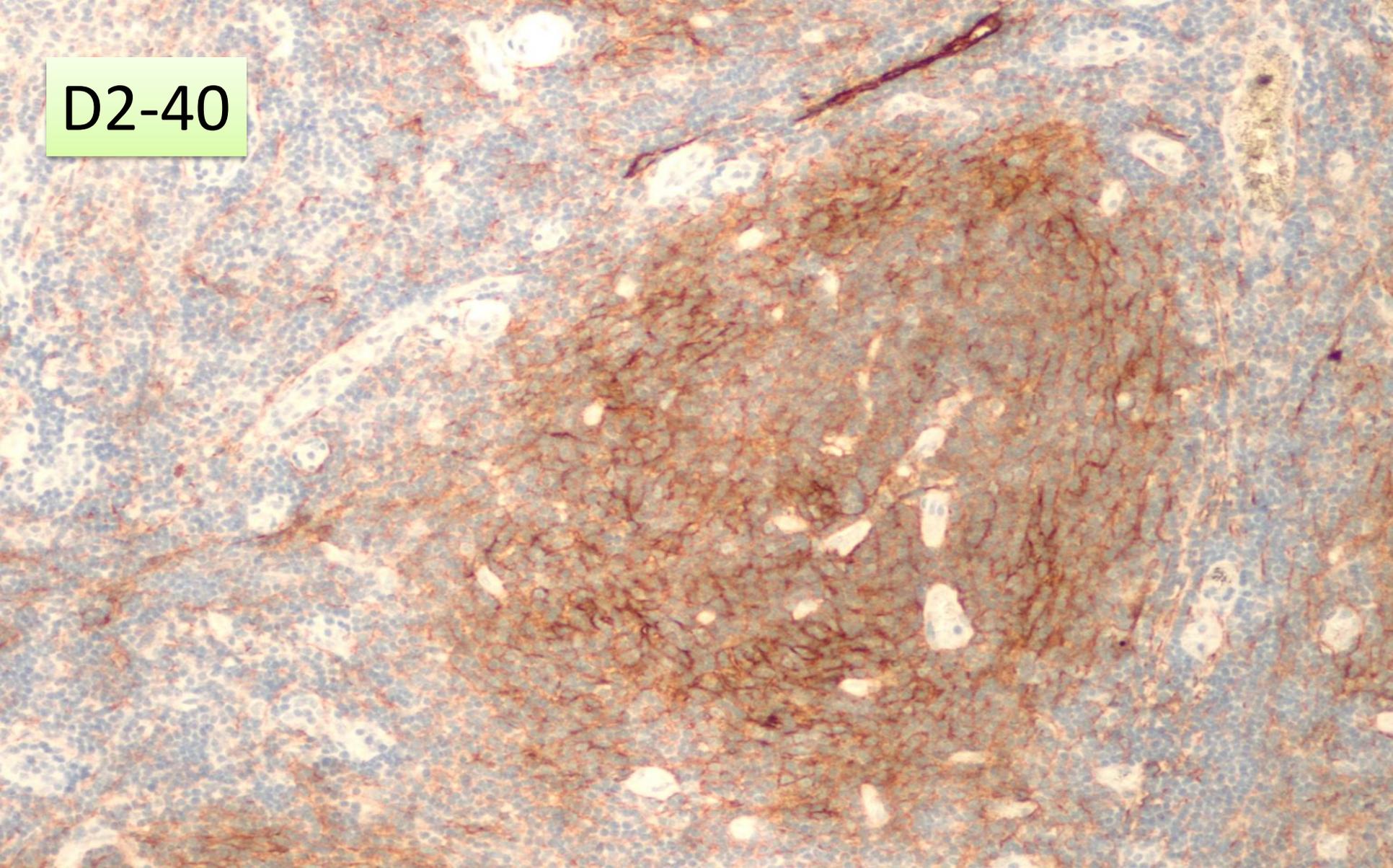
CD35



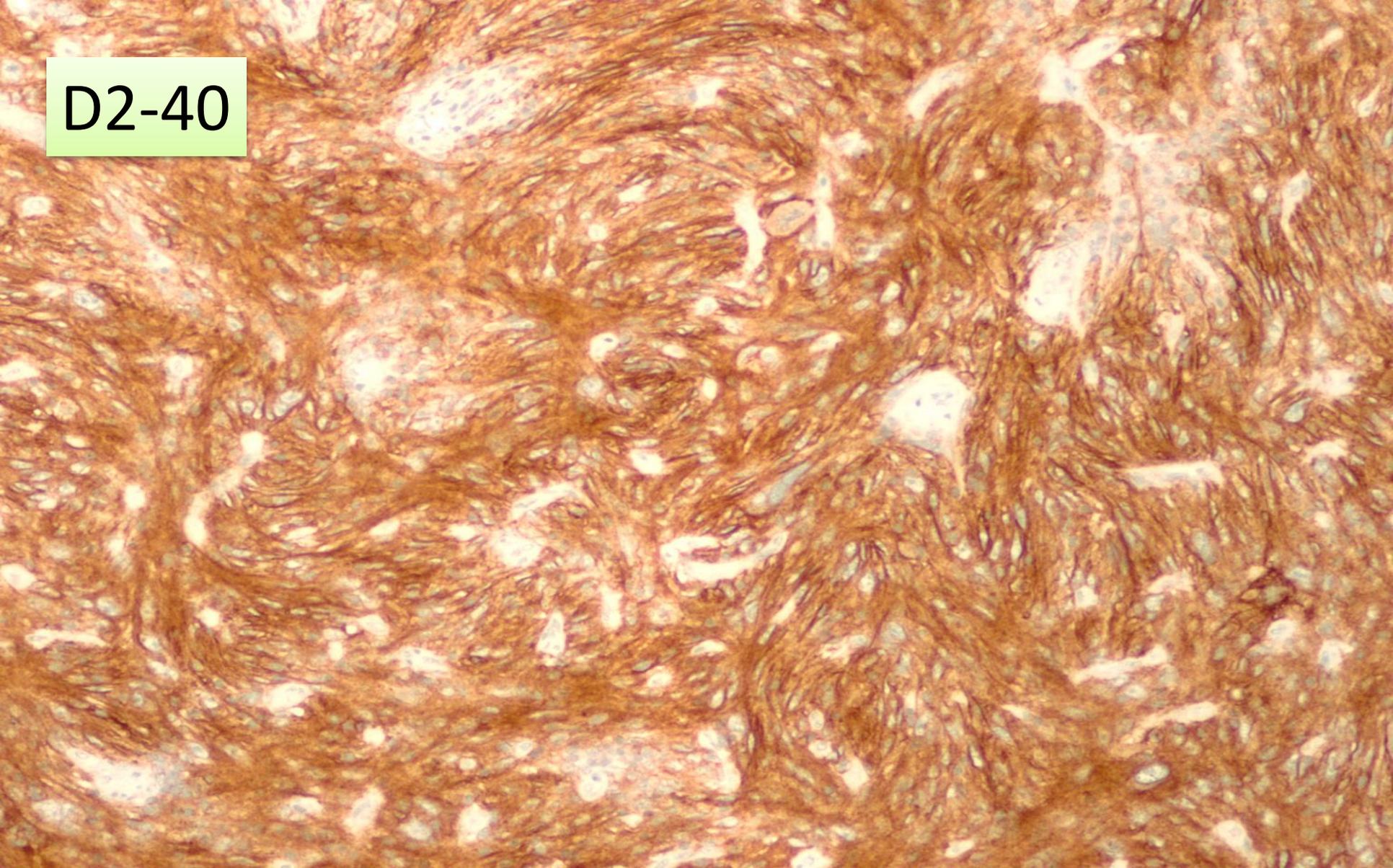
D2-40



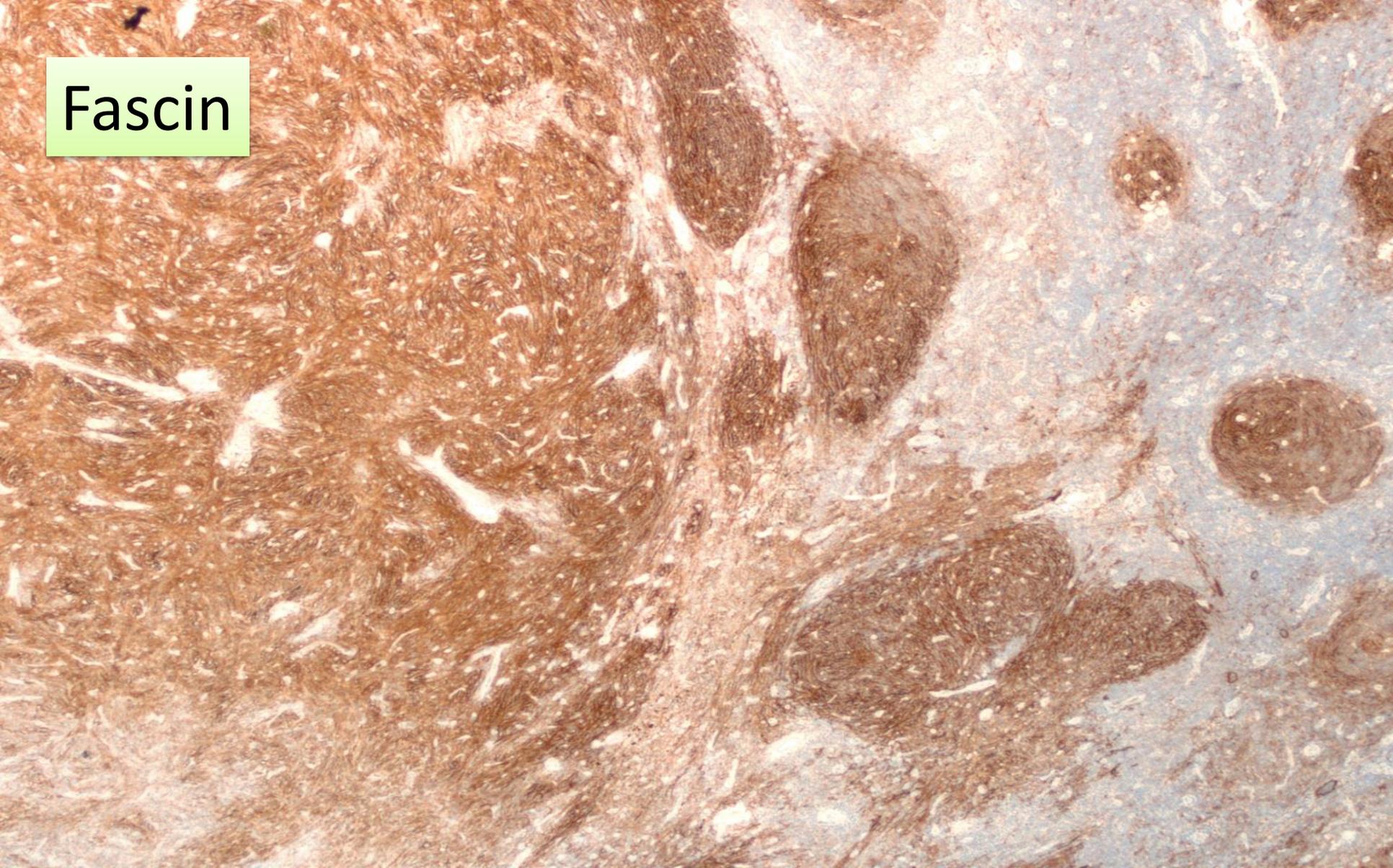
D2-40



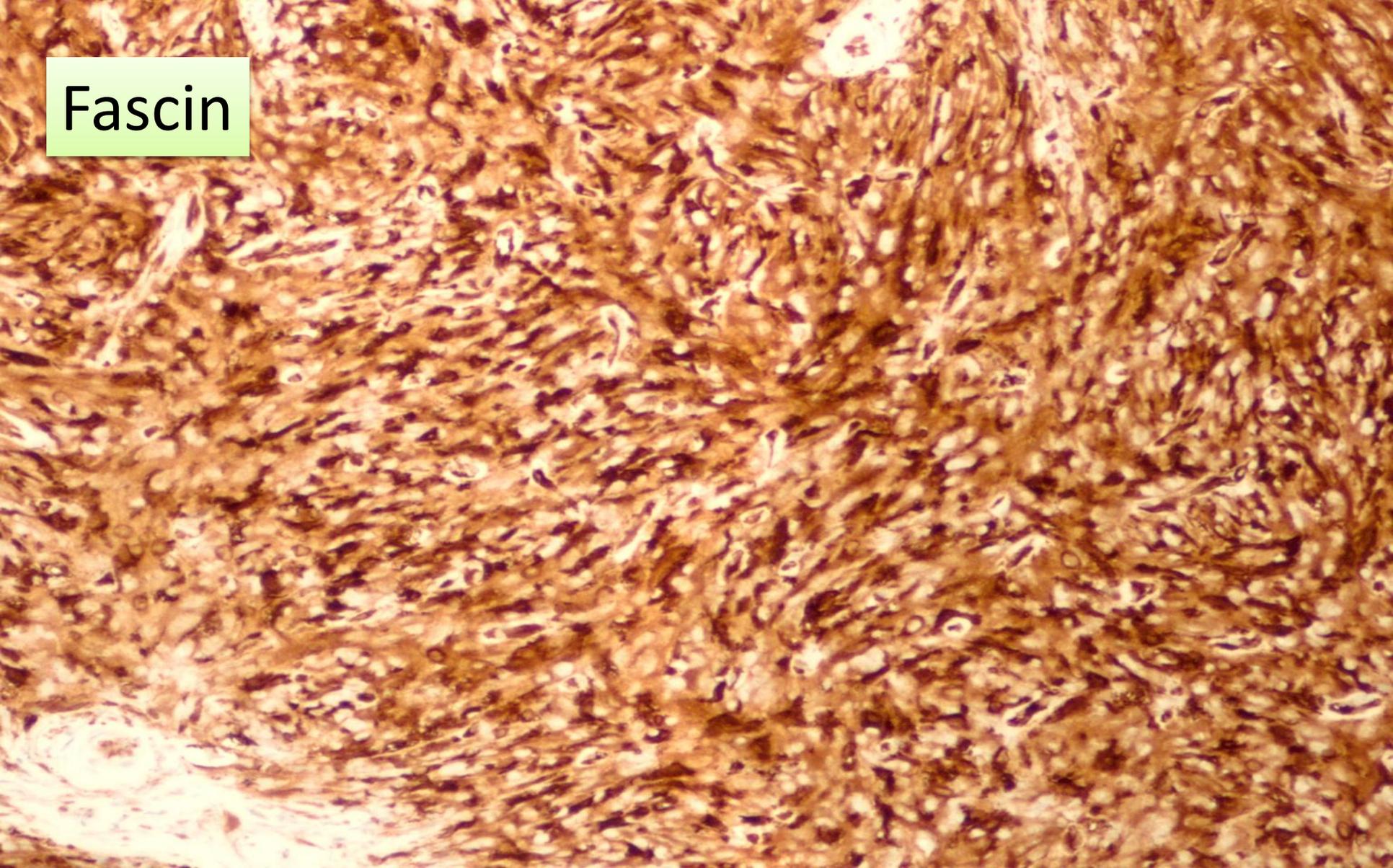
D2-40



Fascin



Fascin



Follicular Dendritic Cell Sarcoma

- Uncommon neoplastic proliferation of spindled to ovoid cells.
- Most FDCS arise from lymph nodes, at least one-third occur in extranodal sites.
- At least some morphologic features of normal FDCs.
- A broad differential diagnosis: spindle cell proliferation/neoplasm
- Characteristic immunophenotypic profile.
 - Relatively specific (may have partial loss): CD21, CD23, CD35, clusterin
 - Sensitive but not specific: D2-40, Fascin
 - Misleading markers (variably positive): CD68, S100, EMA
- Ki-67 usually low, 1-25%
- ~20% harbors *BRAF* V600E mutation

Some Clinical Associations

- Castleman disease
- Angioimmunoblastic T-cell lymphoma (AITL)
- Follicular lymphoma
- Dysregulated immune system:
 - Paraneoplastic pemphigus
 - Myasthenia gravis

Prognosis of FDCCS

- Local recurrences are common, occurring in approximately 40% to 50% of cases.
- Common metastatic sites: liver, lung, and lymph nodes.
- The mortality rate is approximately 20%, usually after a protracted course.

Prognostic Factors of FDCS

- Tumors arising in lymph nodes are often indolent, with low rate of metastases (approximately 10%).
- Unfavorable prognostic factors:
 - intra-abdominal location,
 - large tumor size (greater than 6 cm)
 - Coagulative necrosis,
 - mitotic count greater than 5 mitoses per 10 highpower fields,
 - Significant cellular atypia
- Intraabdominal location is the single most important unfavorable prognostic (relapse rate as high as 80%).

Follow up 4 years later

No clinical presentation;

No new adenopathy.