

DSS 2014

Case #9

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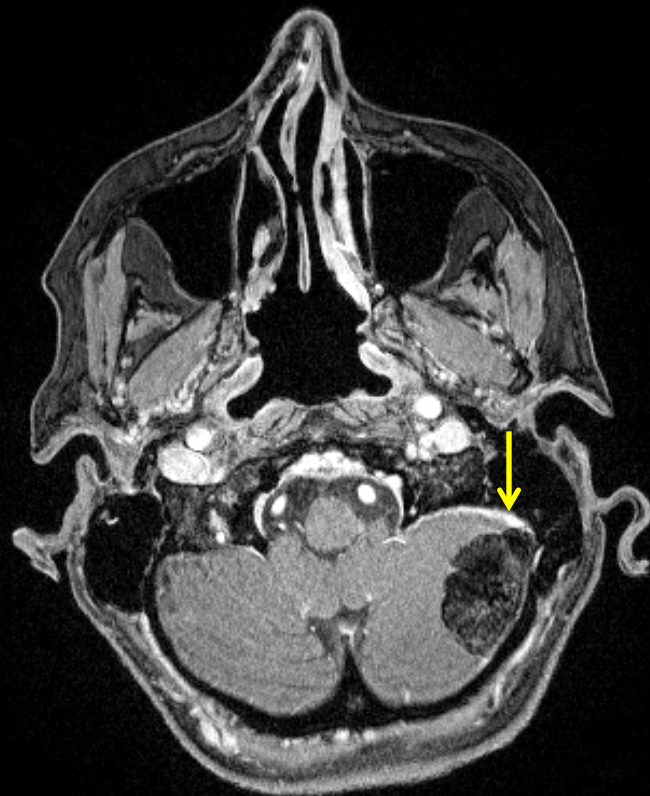
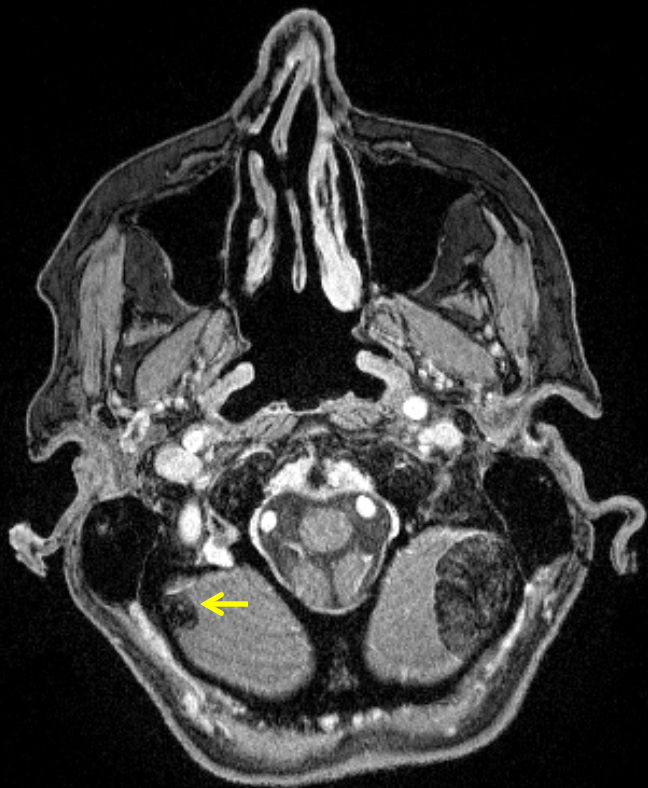
Disclosures

- No financial relationships to disclose

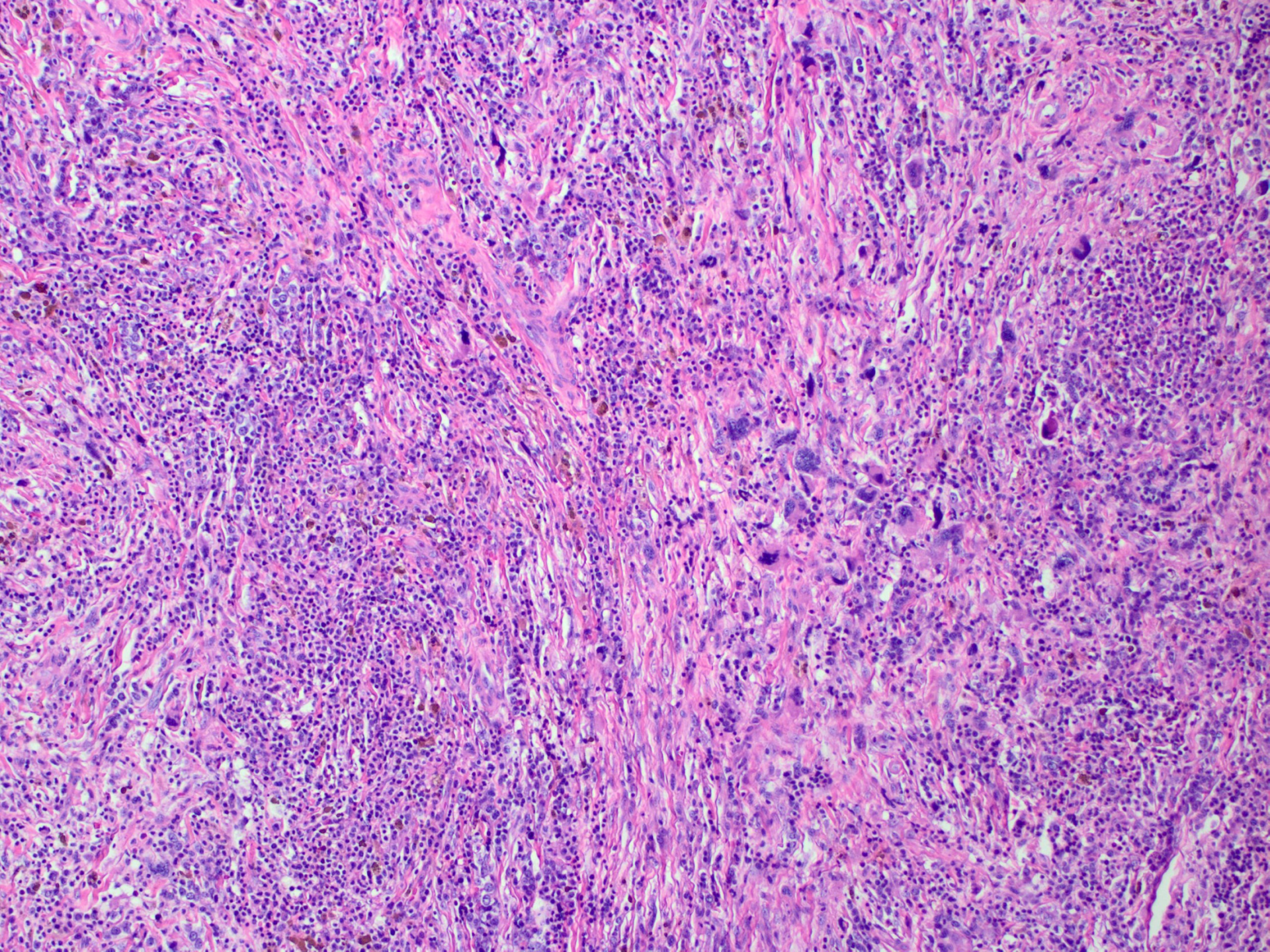
Clinical History

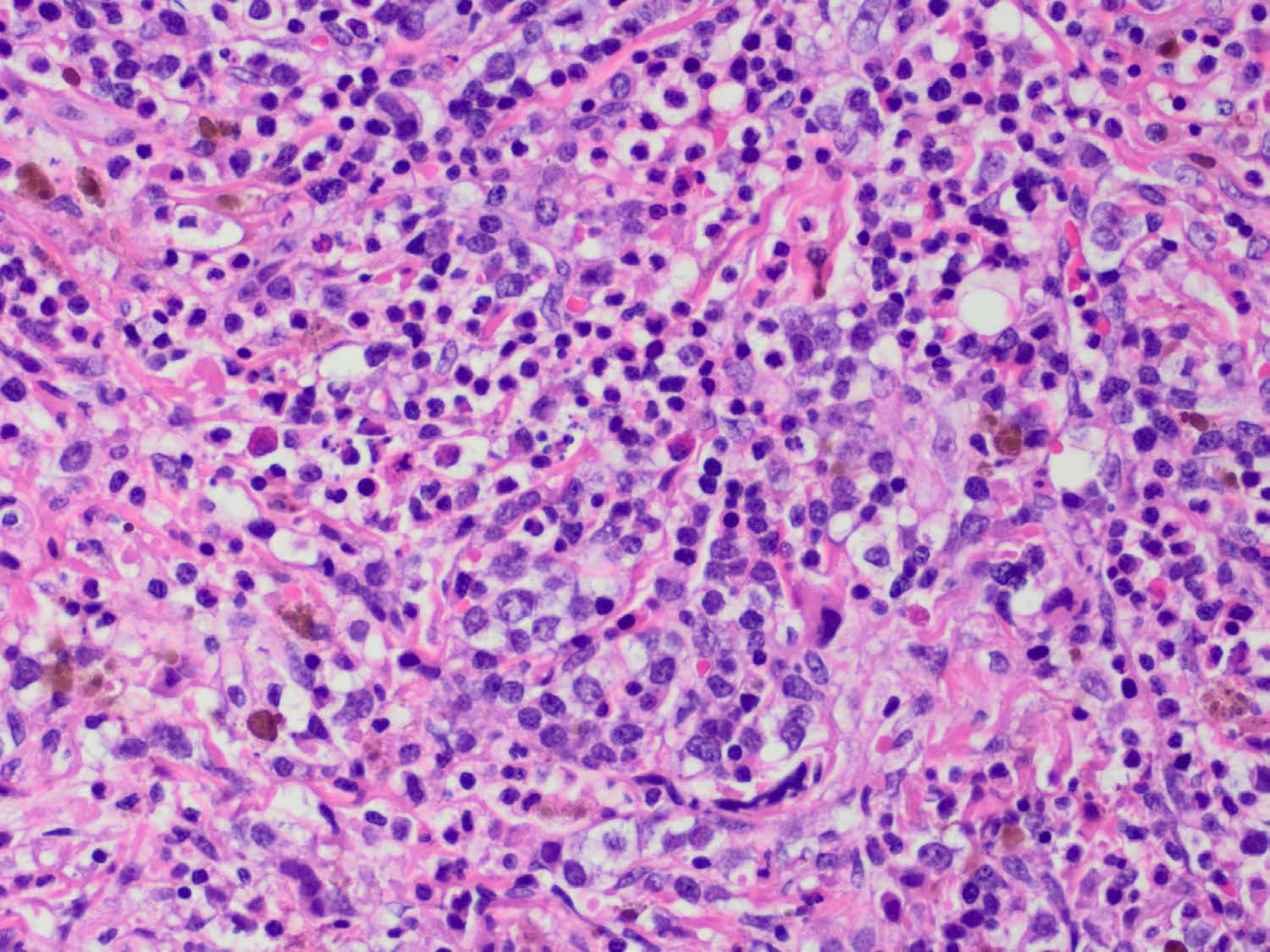
- 64-year-old female presents with constant right-sided frontal headache radiating to cheek and jaw for one month.
- She has a past medical history of cervical cancer, myelofibrosis secondary to polycythemia vera, splenomegaly, and GERD.
- She receives blood transfusions approximately every 3 weeks
- She was hospitalized 3 weeks prior for urosepsis secondary to nephrolithiasis

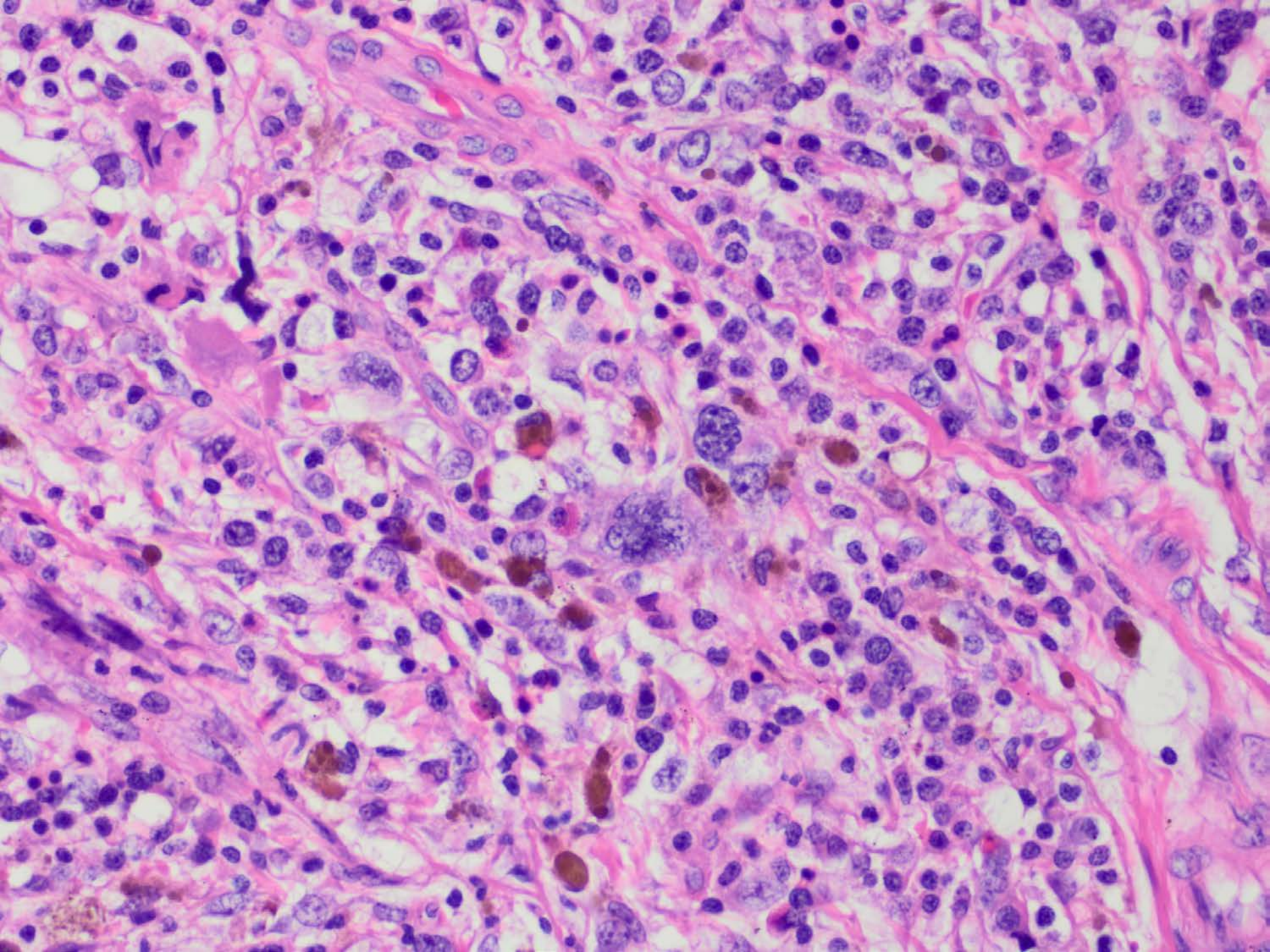
MRI Findings

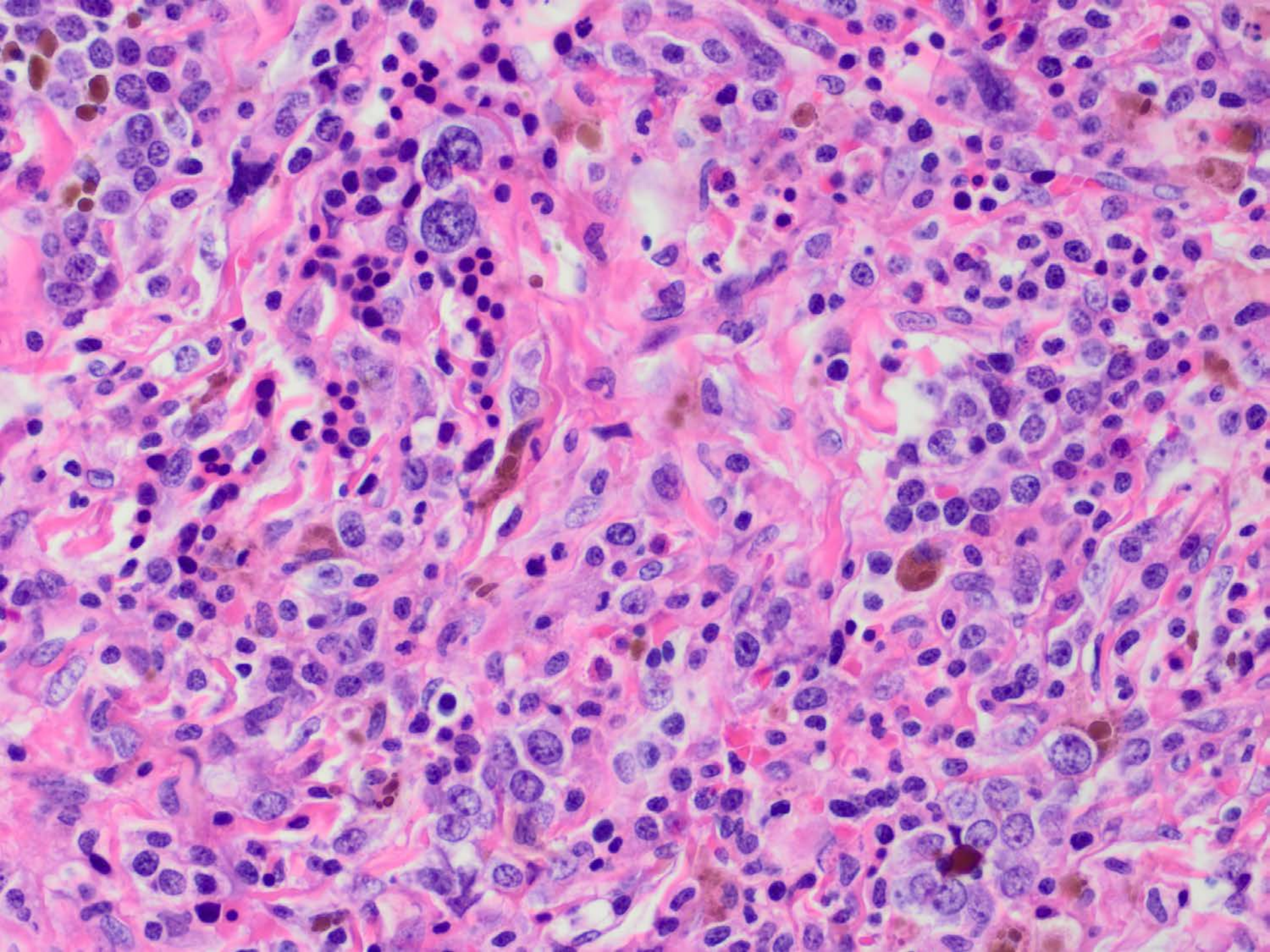


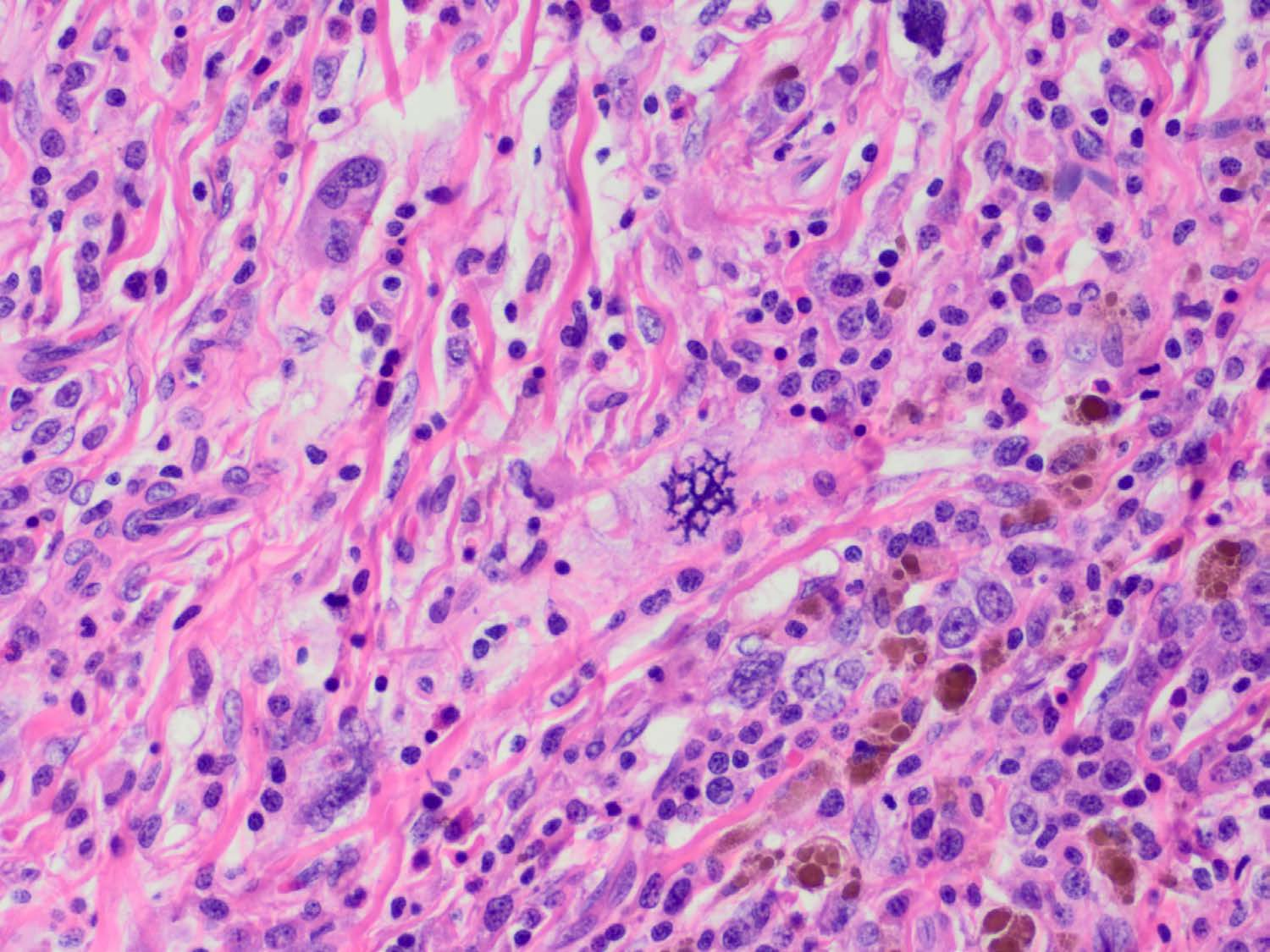
- A left suboccipital craniectomy was performed



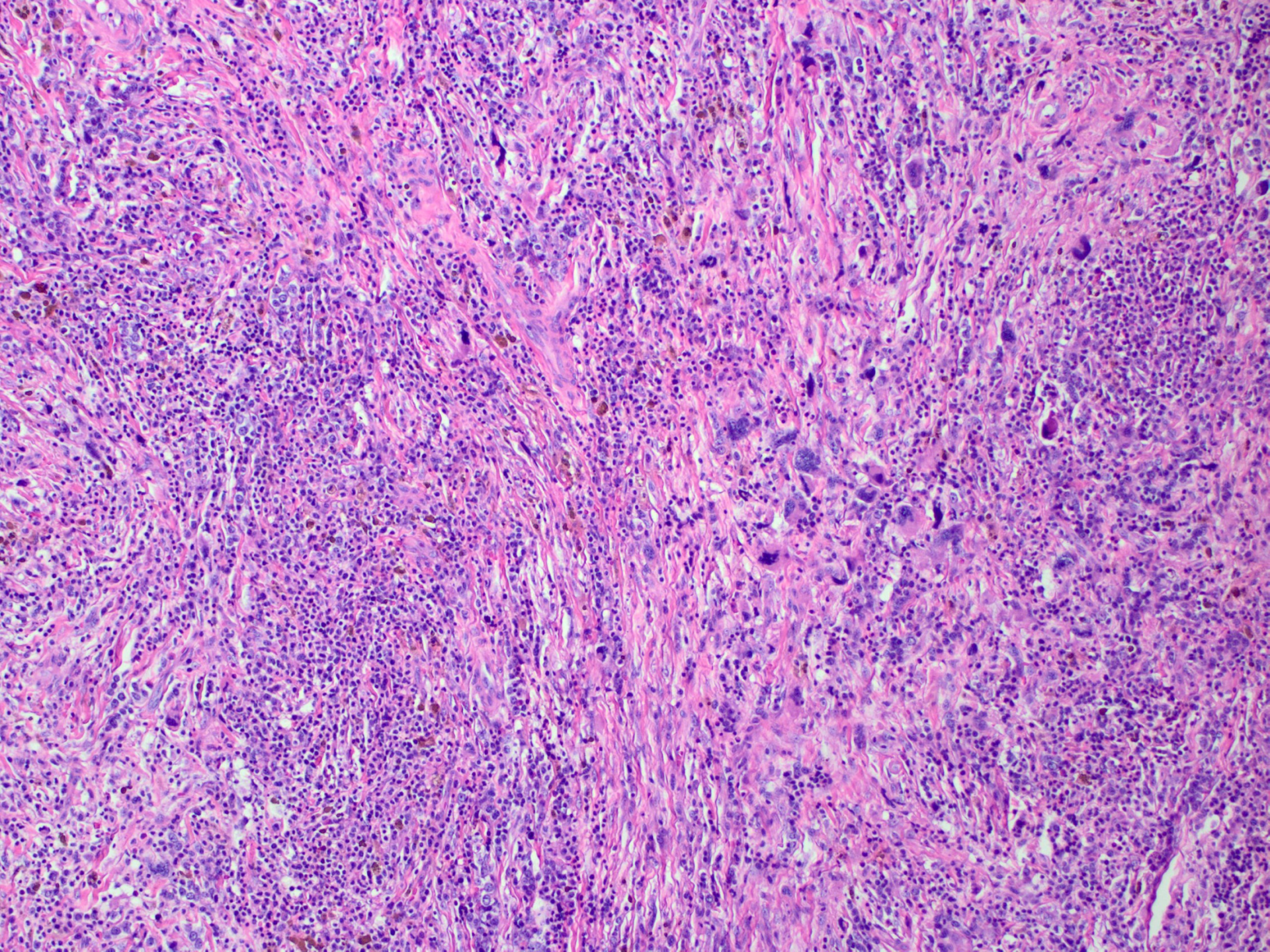


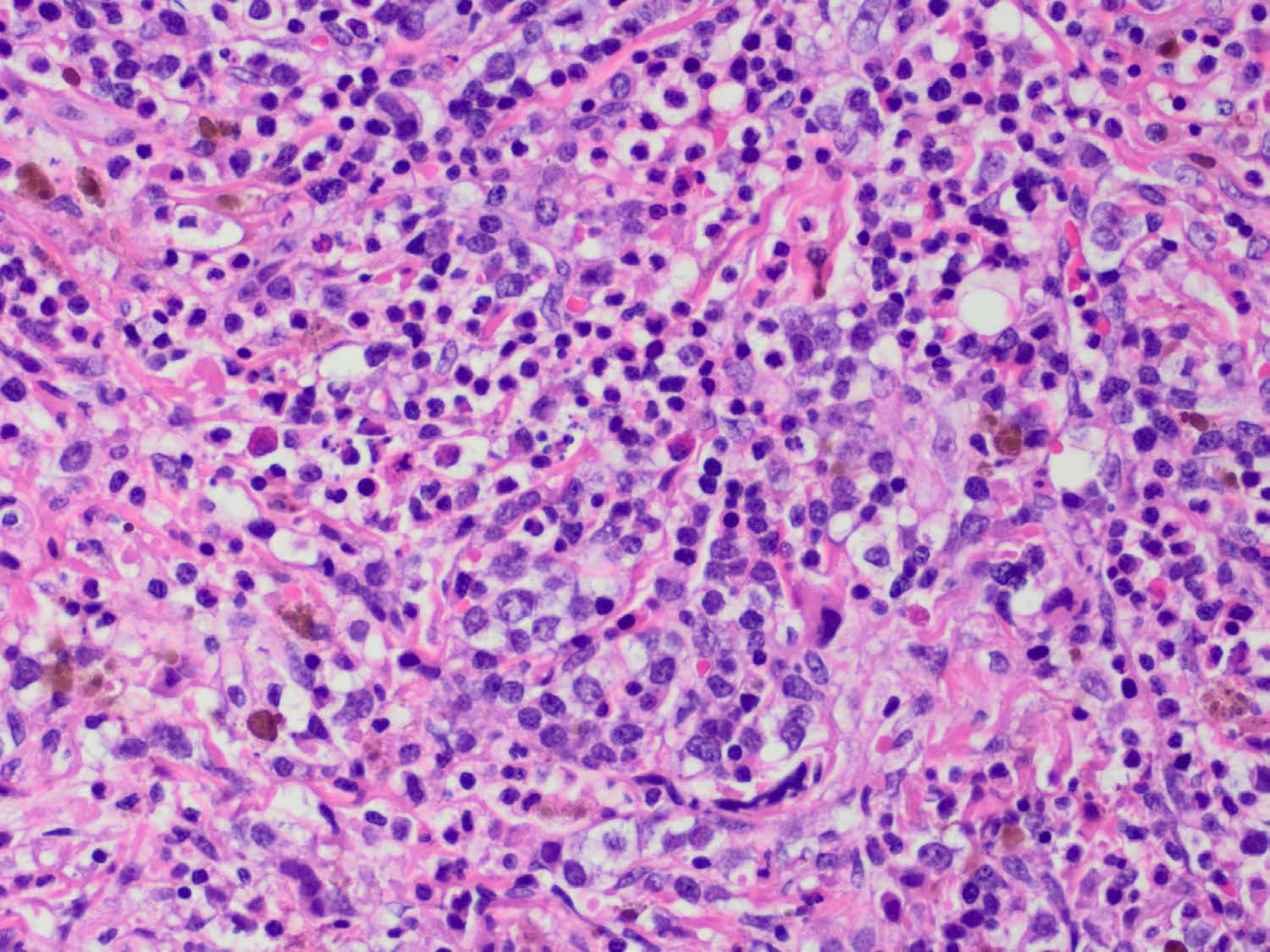


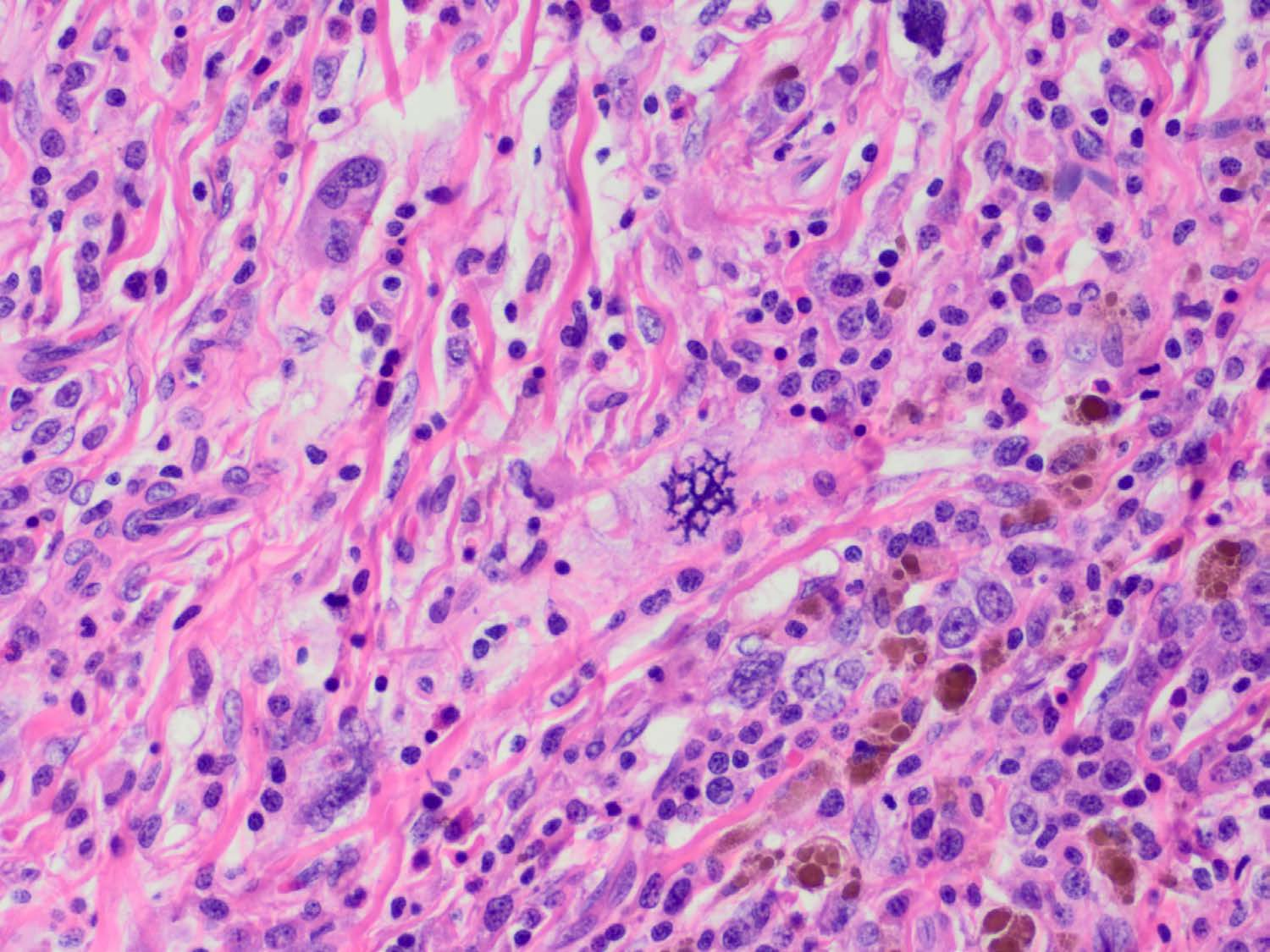




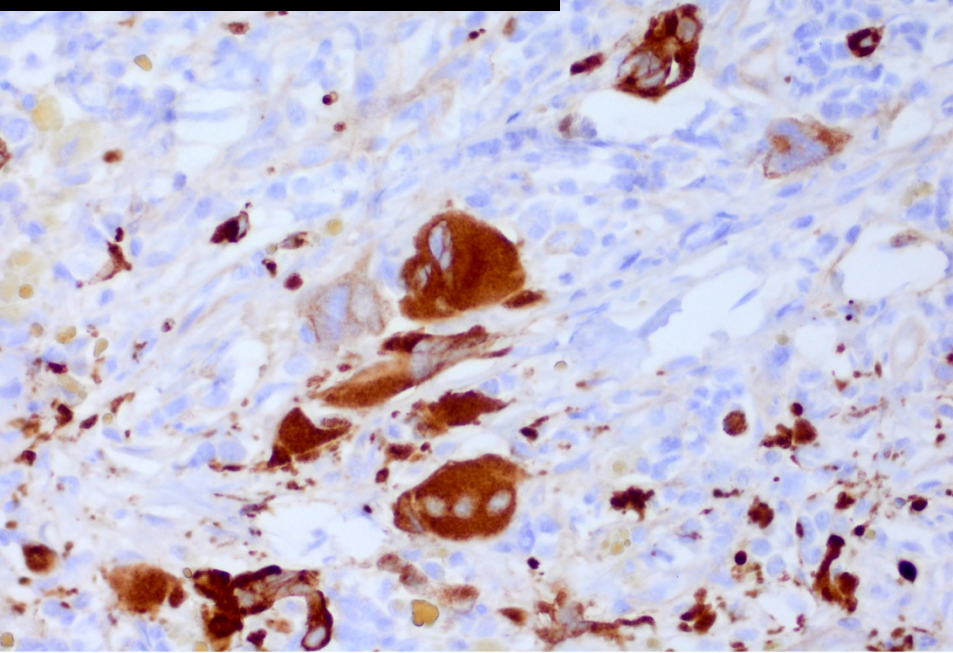
Diagnosis?



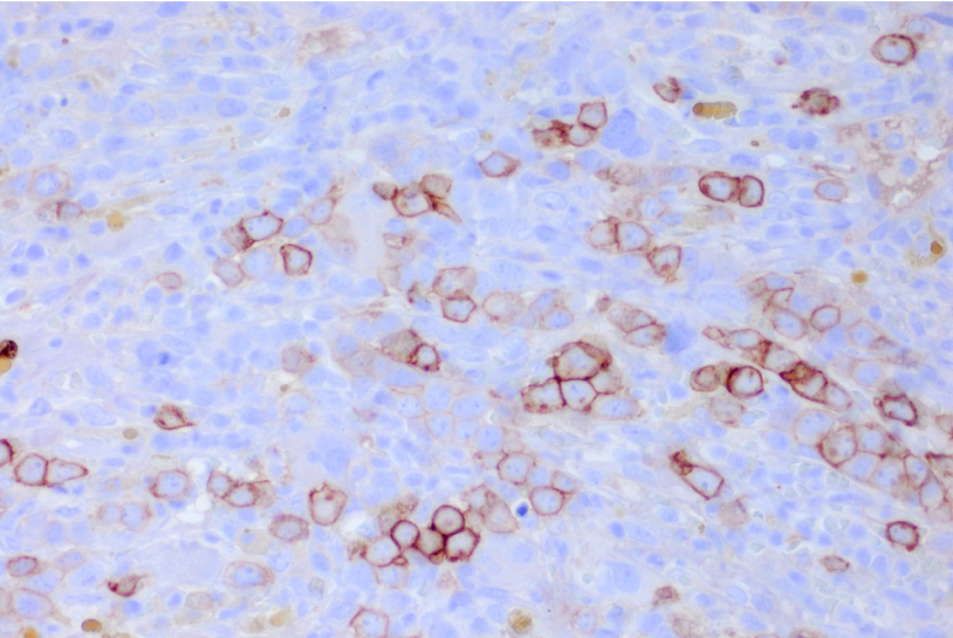
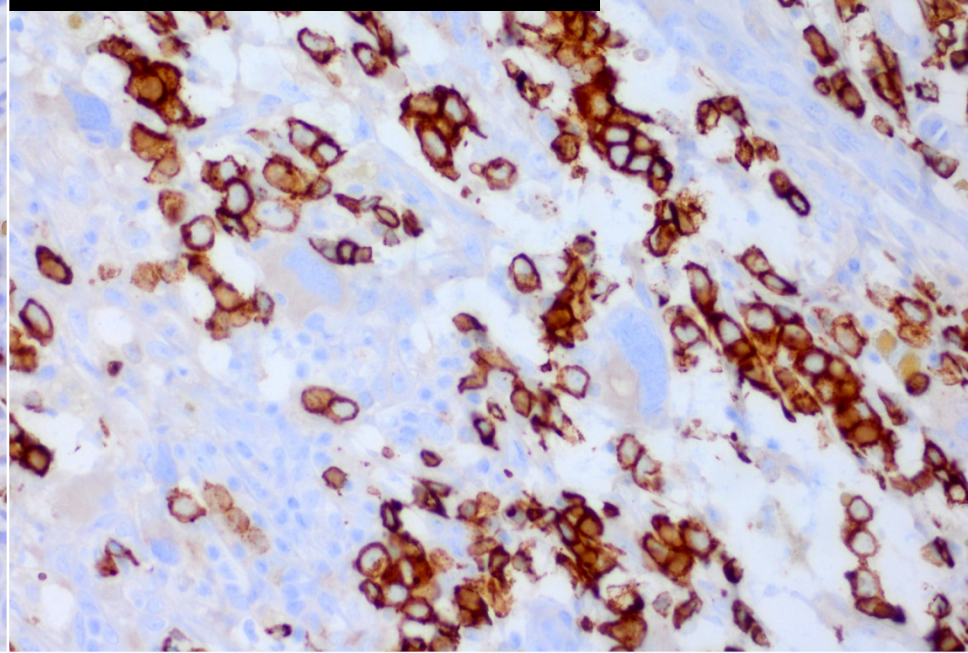




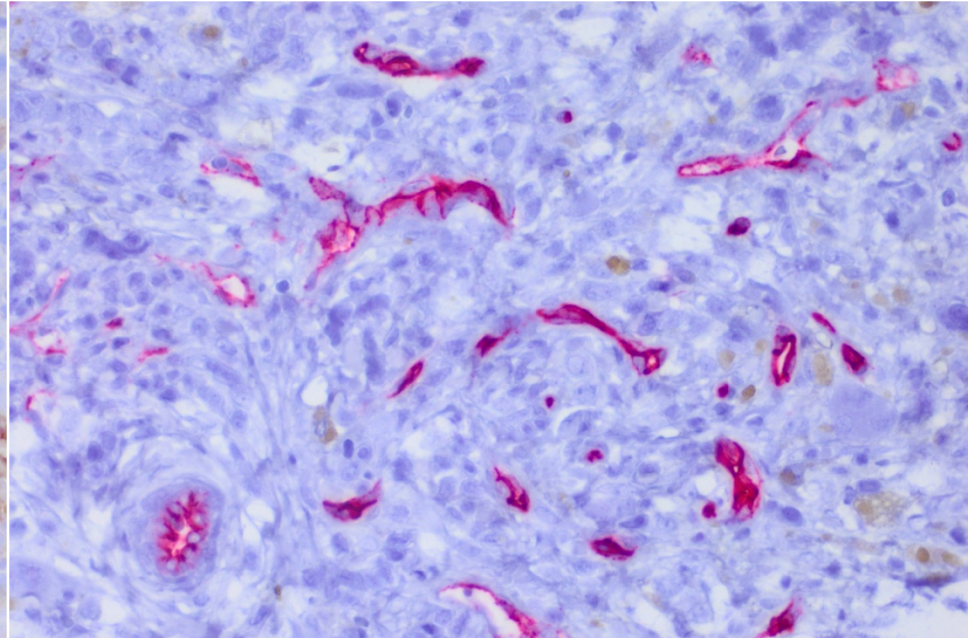
CD61 - megakaryocytes



CD71 - erythroid precursors

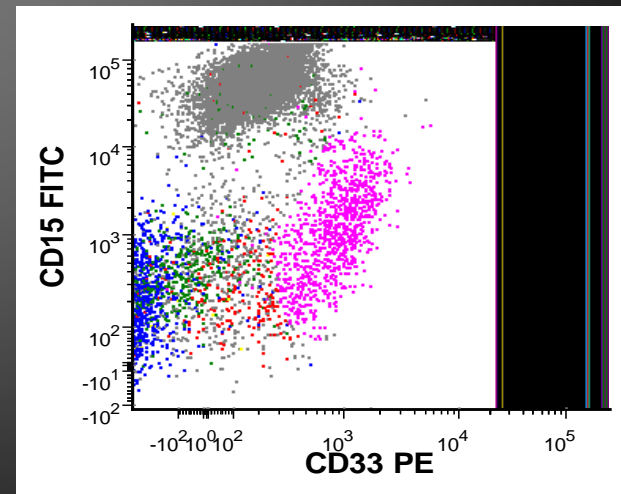
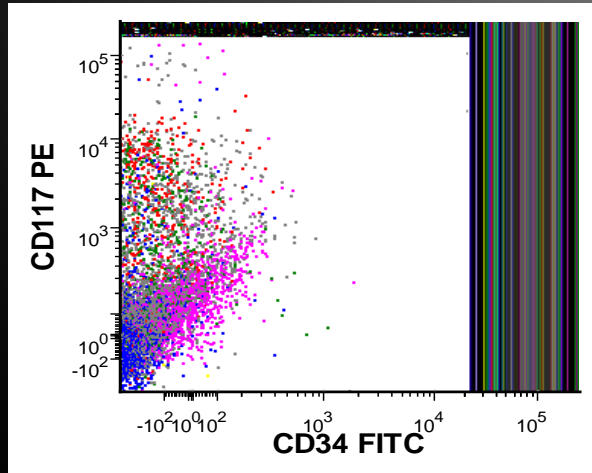
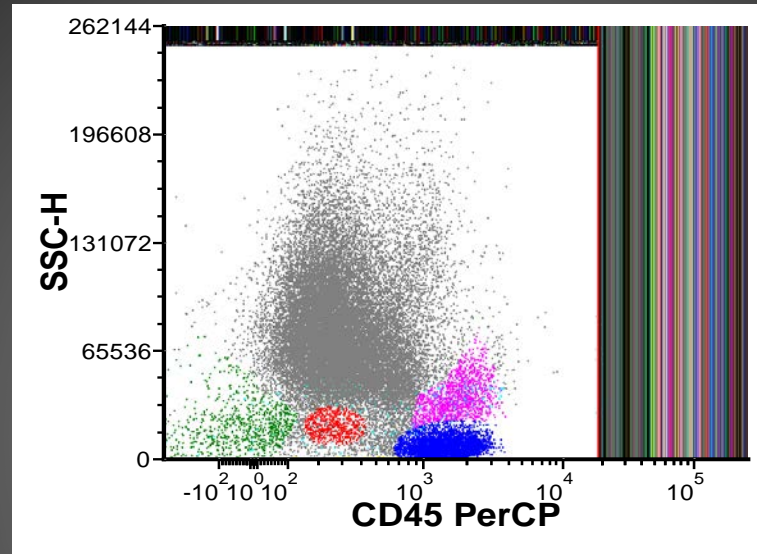


c-KIT - early erythroid & myeloid cells



CD34 - hematopoietic stem cells

Flow Cytometry



Diagnosis

- **Extramedullary involvement by chronic myeloproliferative neoplasm with extensive fibrosis**
 - Consistent with prior bone marrow biopsy, 2011
 - Multilineage hematopoietic elements with erythroid and megakaryocytic hyperplasia with atypia and fibrosis

Chronic Myeloproliferative Neoplasm

- Polycythemia vera
- Essential thrombocytopenia
- Primary myelofibrosis
- Chronic myeloid leukemia

Polycythemia Vera

- Clonal proliferation of myeloid cells with the presence of an elevated red blood cell mass
- Occurs in all age groups, mean age 60
- Survival when treated exceeds 10 years, untreated 18 months
- 95 to 100% of patients with PV have a JAK2 mutation, usually V617F

Treatment

- Jakafi (ruxolitinib) is a JAK1/2 inhibitor
 - Improvement in debilitating symptoms and quality of life
 - Reduction in splenomegaly
 - ? Overall survival benefit

Secondary Myelofibrosis

- PV transforms into post-PV myelofibrosis in approximately 10% of patients
 - Risk factors : disease duration, age > 60
 - Non-clonal proliferation of fibroblasts caused by inappropriate secretion of growth factors by megas or platelets
- Significantly increases the risk for:
 - Acute leukemia
 - “Extramedullary hematopoiesis”

EMH Terminology

The American Journal of Surgical Pathology 24(1): 51-55, 2000

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Sclerosing Extramedullary Hematopoietic Tumor in Chronic Myeloproliferative Disorders

Ellen D. Remstein, M.D., Paul J. Kurtin, M.D., and
Antonio G. Nascimento, M.D.

- Predominant sclerotic component and atypical megakaryocytes
- May be multiple or solitary
- Marker of advanced disease

EMH Tumor

- “Fibrous hematopoietic tumor”
- “Extramedullary myelofibrosis”

EXTRAORDINARY CASE REPORT

Cutaneous Myelofibrosis With *JAK2* V617F Mutation:
Metastasis, Not Merely Extramedullary Hematopoiesis!

*Garth R. Fraga, MD** and *Samuel K. Caughron, MD†*

Extramedullary hematopoiesis involving the central nervous system and surrounding structures

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Human Pathology; 2011; 42 (10): 1524–1530

CASE OF MONTH MAY 2011

doi:10.1111/j.1750-3639.2011.00521.x

37-YEAR-OLD WOMAN WITH MULTIPLE INTRACRANIAL MASSES

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² Department of Radiology, Tongji Hospital, Tongji University School of Medicine, Shanghai, China

Brain Pathology; 2011, 21 (5): 607–610

CASE REPORT

Marie E. Beckner · John Y. K. Lee
Sydney S. Schochet Jr. · Charleen T. Chu

Intracranial extramedullary hematopoiesis associated with pilocytic astrocytoma: a case report

Acta Neuropathology; 2003, 106: 584-587

Intracranial EMH - Treatment

- Radiation

American Journal of Hematology 78:151–152 (2005)

**Intracranial Extramedullary Hematopoiesis:
Brief Review of Response to
Radiation Therapy**

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- Varying regimens 10-30 Gy
- Good neurologic recovery
- Surgery - for decompression only
- Myelosuppression - already myelosuppressed

Patient Summary

- Low dose radiation
- Patient suffered a fall at home leading to left cerebellar hemorrhage
- Procedures were withheld due to thrombocytopenia
- Transferred to hospice and died one month later

Summary

- Patient with bilateral cerebellar masses
- History of myelofibrosis secondary to Jak2+ PV
- Dx: Extramedullary site of chronic myeloproliferative neoplasm
- Rare complication of hematopoietic dyscrasias
 - Thalassemia > myelofibrosis
- Metastatic vs reactive/reparative response
- Radiation alone