

USCAP 2008 Specialty Conference:

NEUROPATHOLOGY: Case #1



Anthony T. Yachnis, MD

Professor

Director of Anatomic Pathology

Chief, Neuropathology Section

Department of Pathology, Immunology

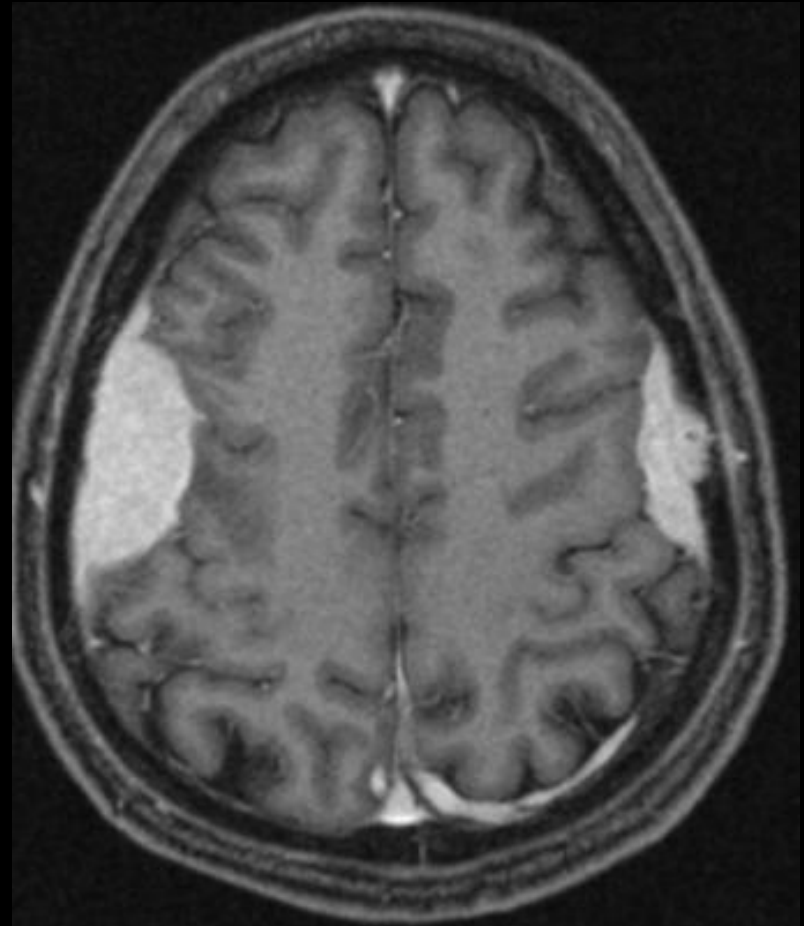
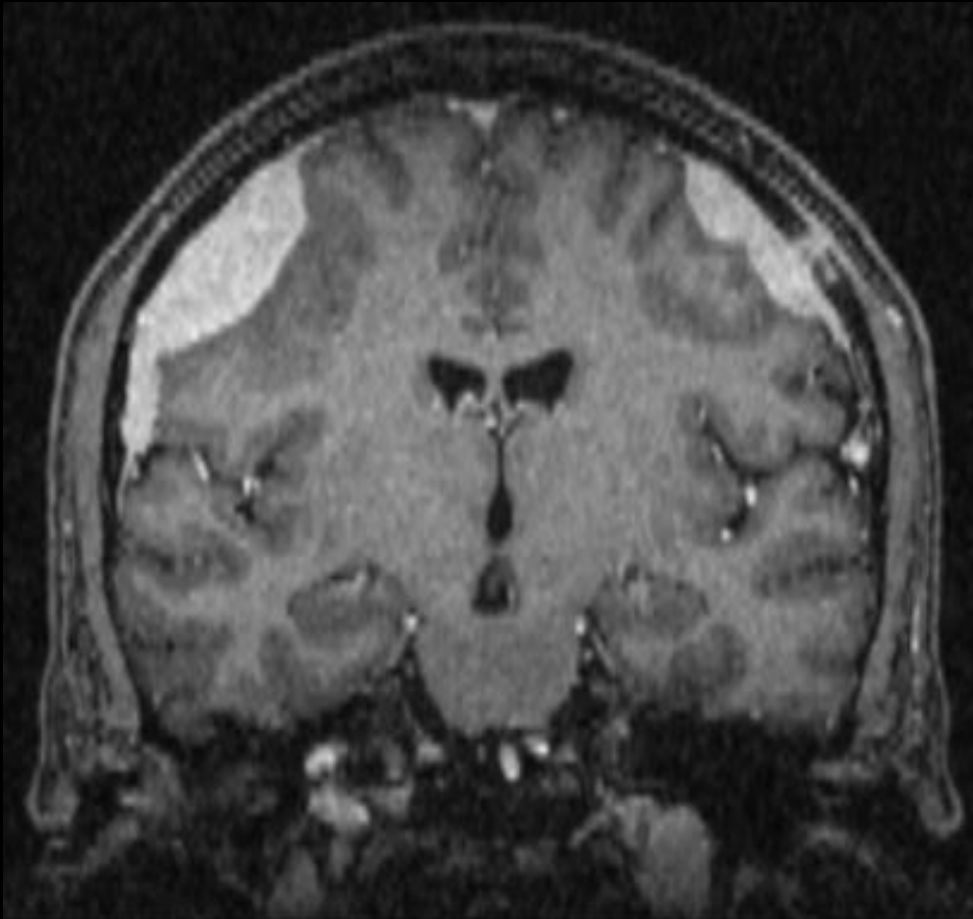
and Laboratory Medicine

University of Florida

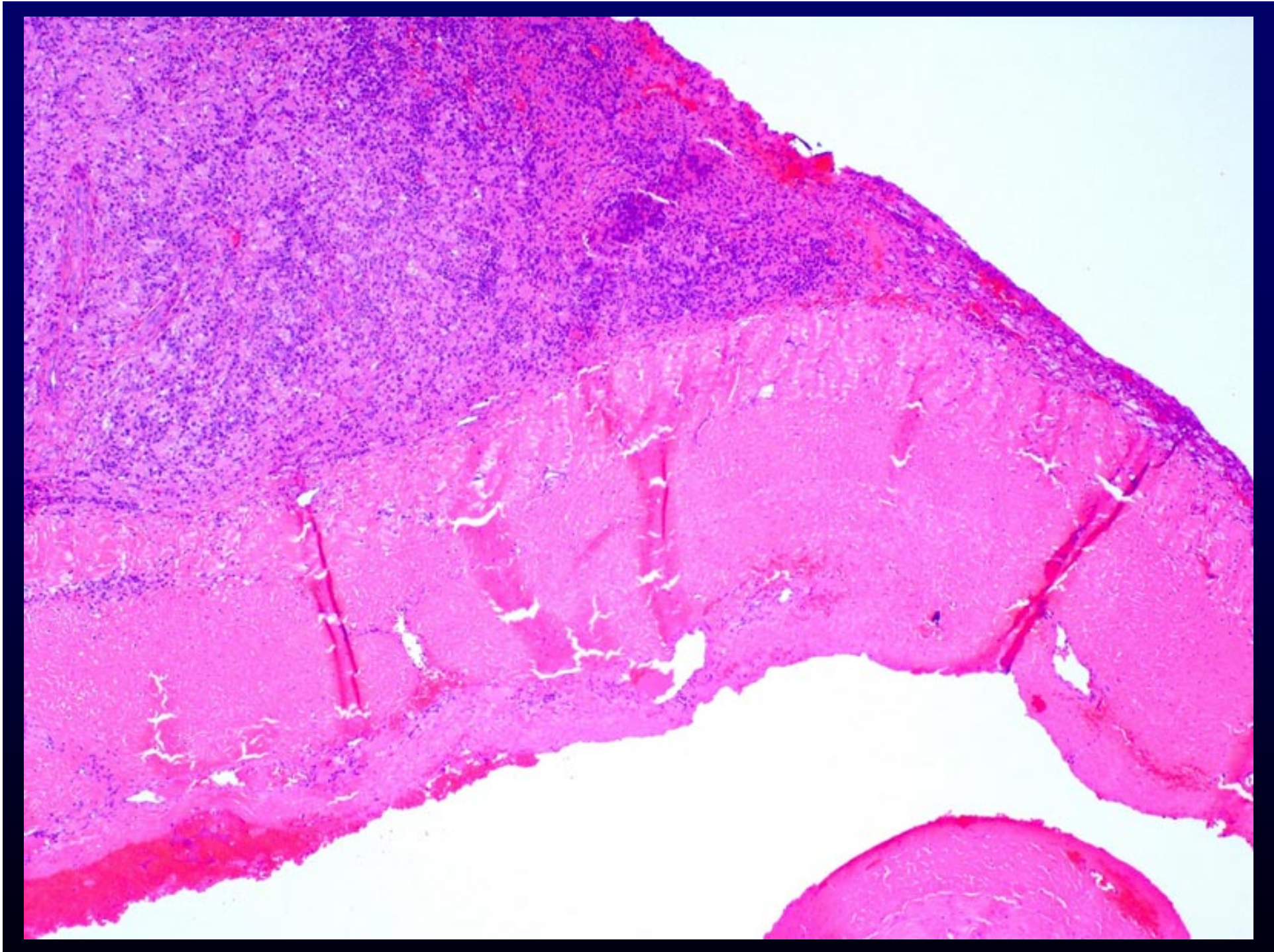
College of Medicine

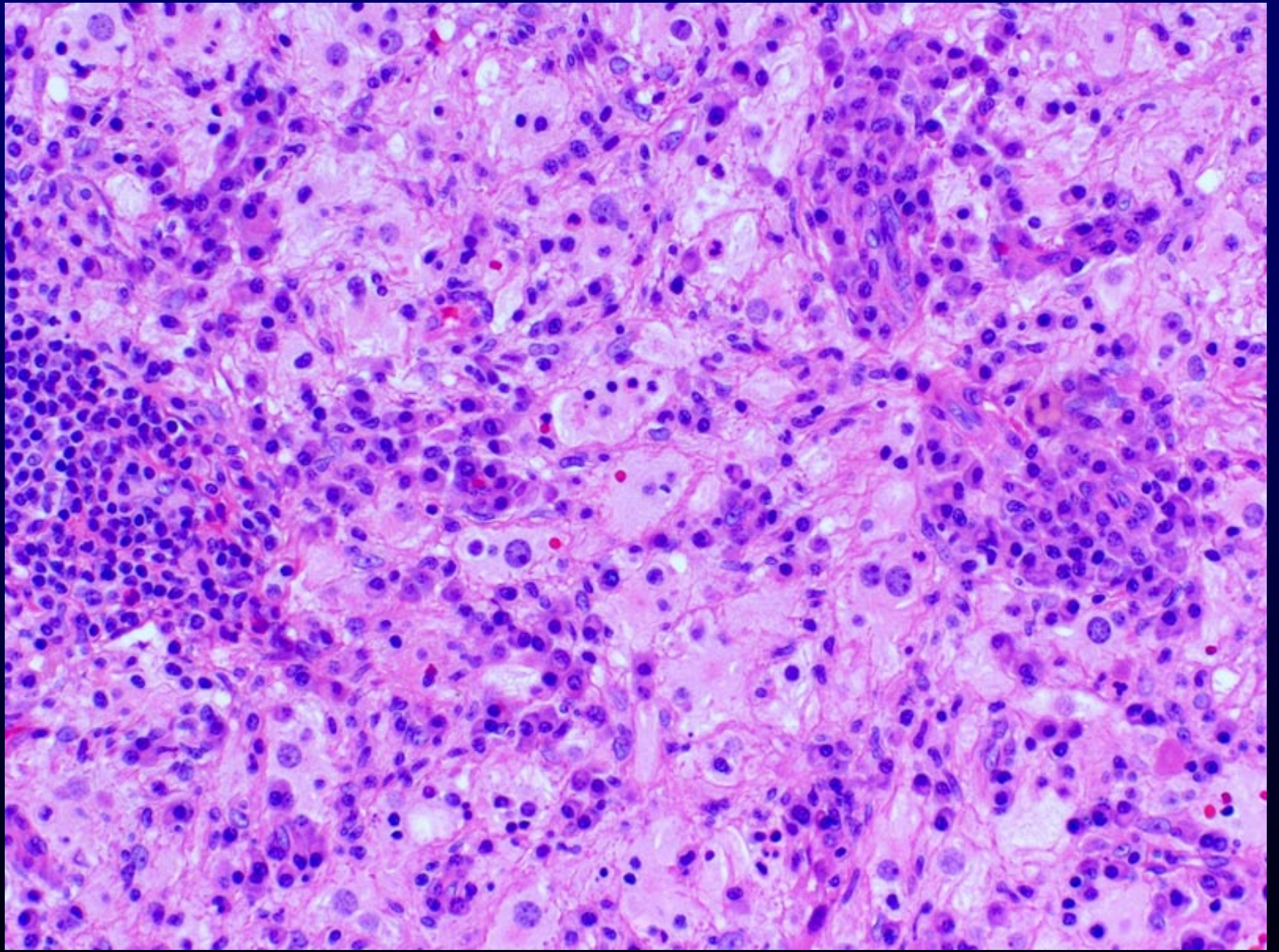
Clinical History

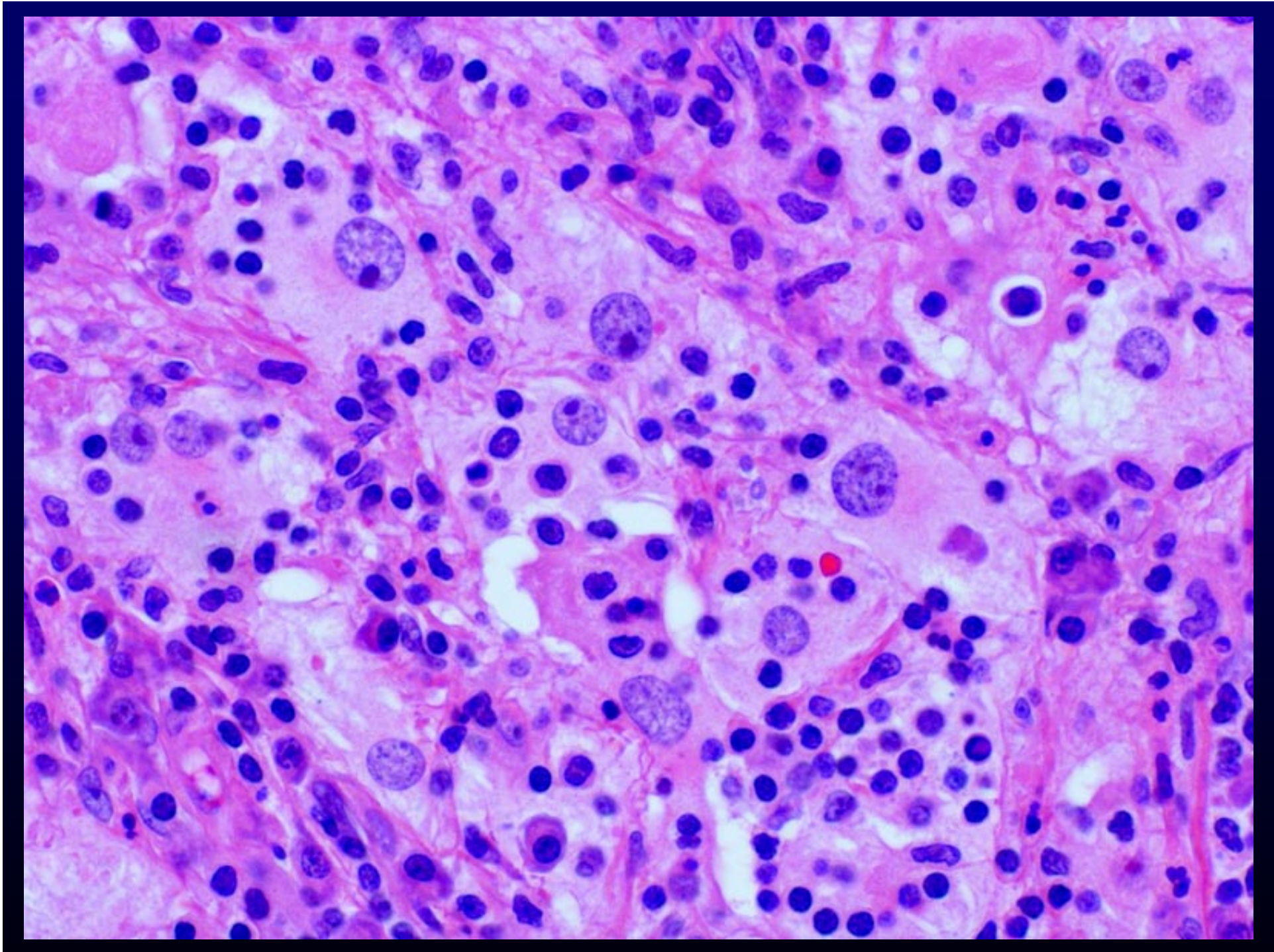
- **35-year-old man presented with new onset tonic clonic seizures**
- **Imaging revealed bilateral, contrast-enhancing masses that were consistent with meningiomas**
- **Past medical history was unremarkable.**
- **The patient underwent a craniotomies with excision of the lesions in two separate operations**

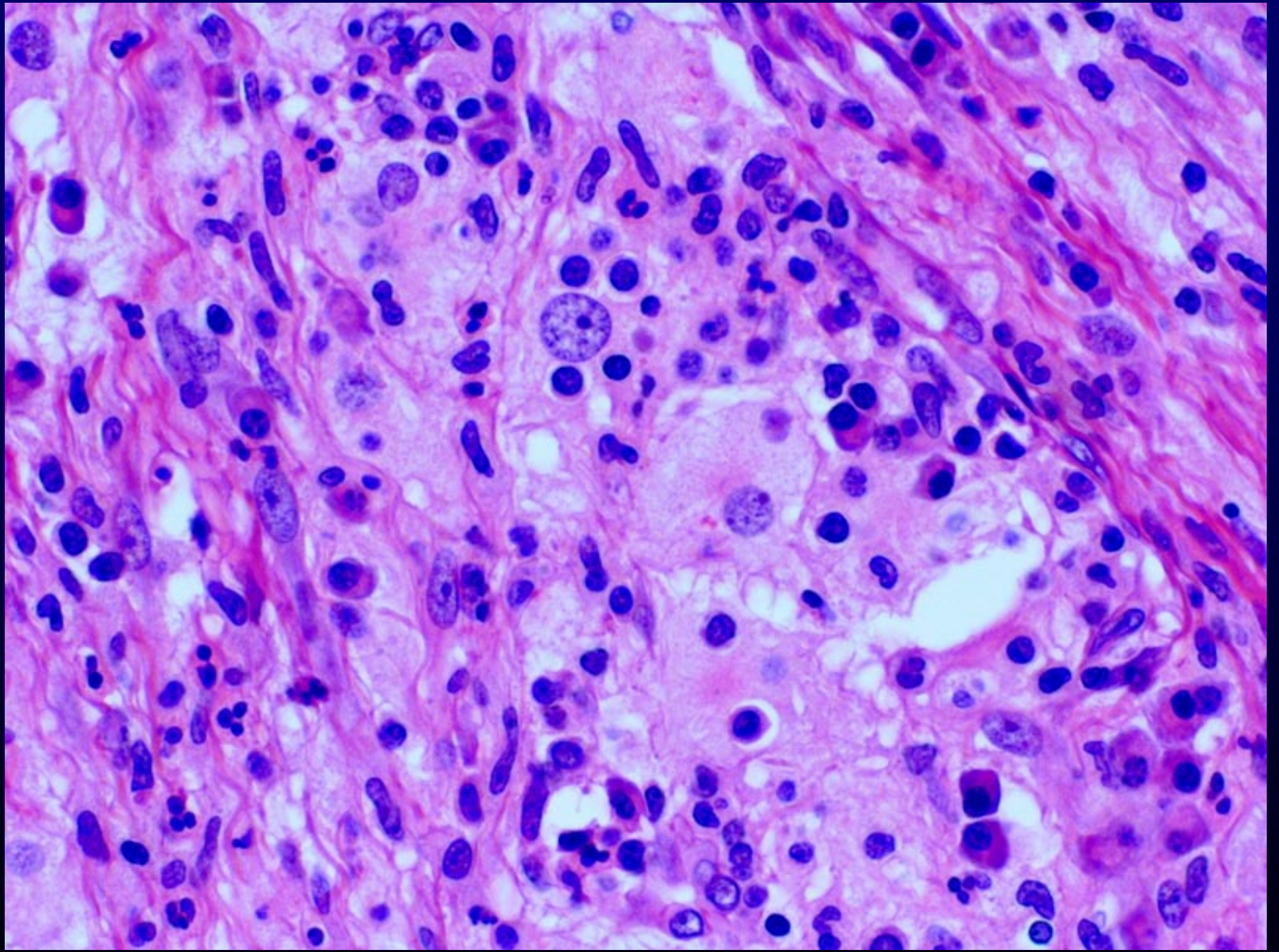


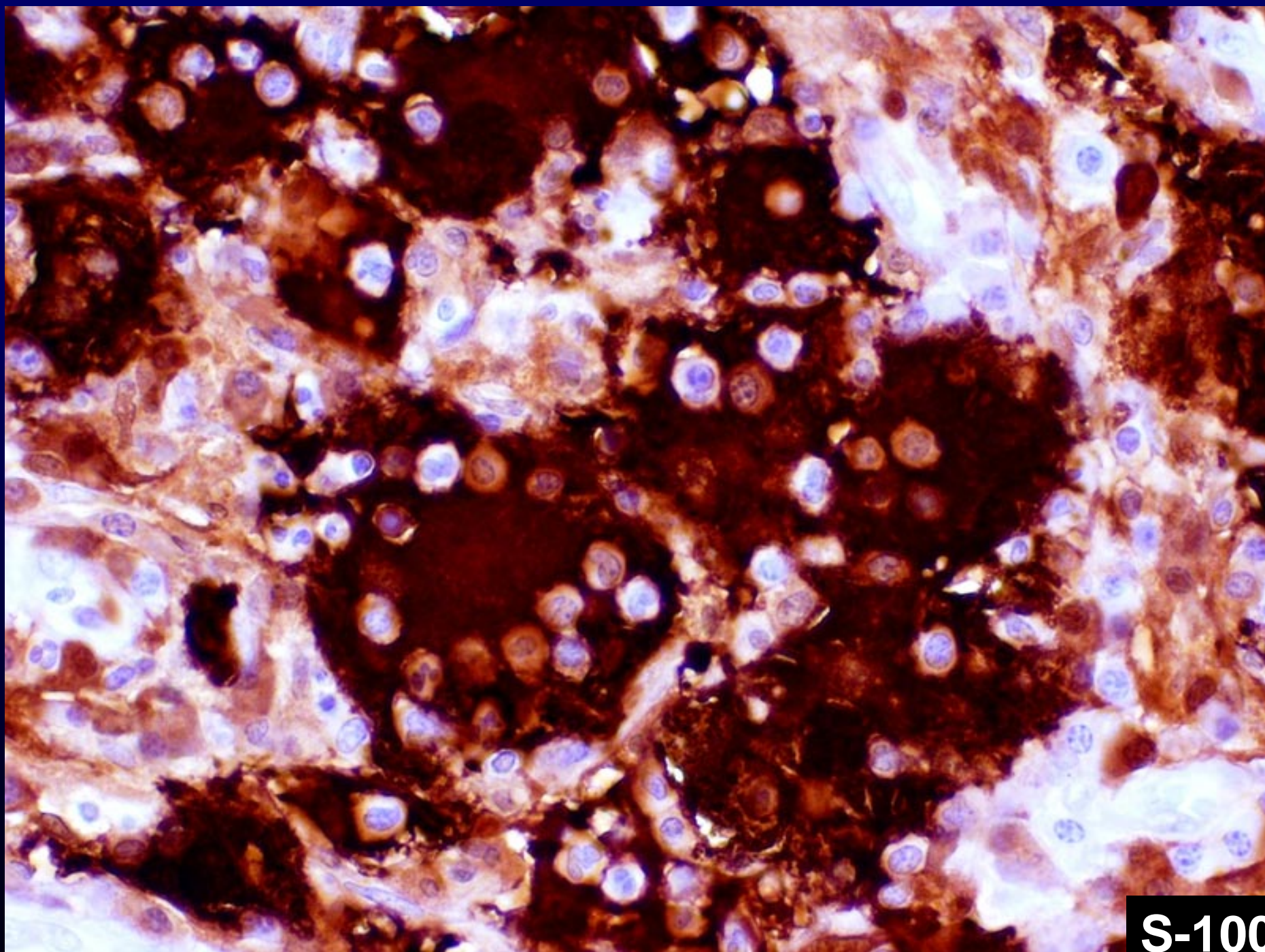
T1-weighted post-contrast MRI





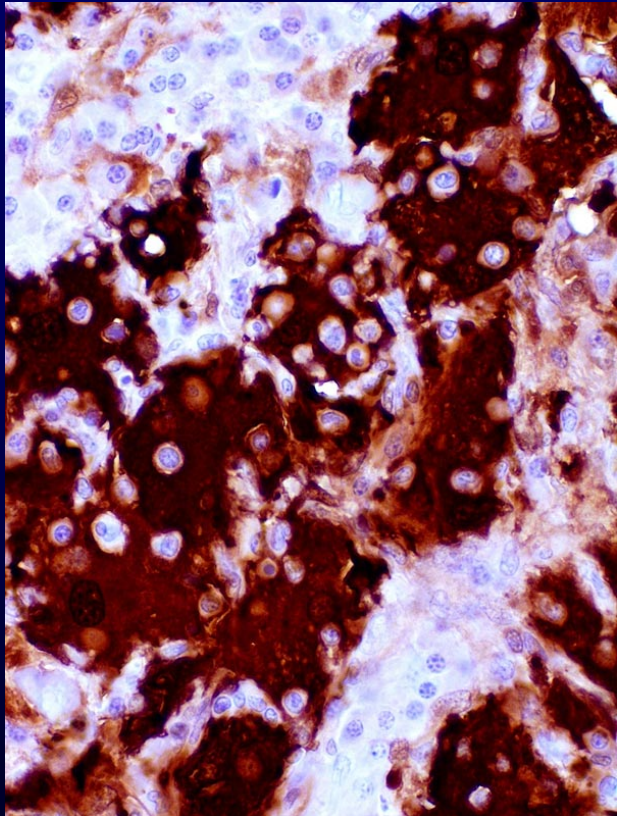




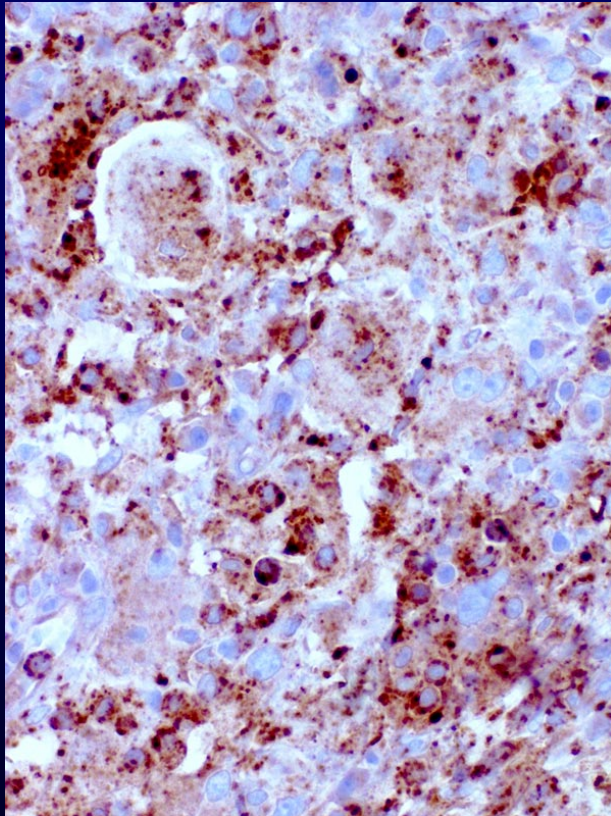


S-100

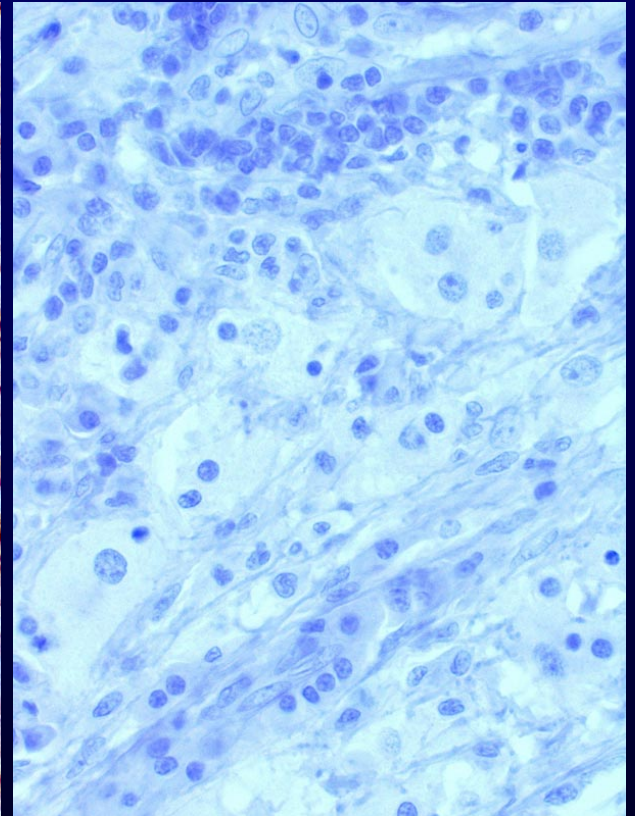
S-100



CD68



CD1A



Diagnosis:

**Extranodal sinus histiocytosis with
massive lymphadenopathy
(Rosai-Dorfman disease)
Bilateral Dural Involvement**

Rosai-Dorfman Disease

- ***Classic form:***

- Young patients with massive, painless cervical lymphadenopathy, fever, weight loss, and increased sedimentation rate
- Lymph node biopsy: inflammatory-like mass with prominent atypical histiocytes that appeared to be engulfing the other cellular elements
- Lesions of similar type found outside the lymphoid organs

Rosai-Dorfman Disease

Intracranial Extranodal Involvement:

- Dural-based mass with meningioma-like imaging findings is most common
- Other nervous system locations:
 - Spinal
 - Suprasellar
 - Cavernous sinus
 - Orbital
 - Intra-axial brain involvement is rare
- Good prognosis overall (recurrent or disseminated forms have occurred)

Rosai-Dorfman Disease

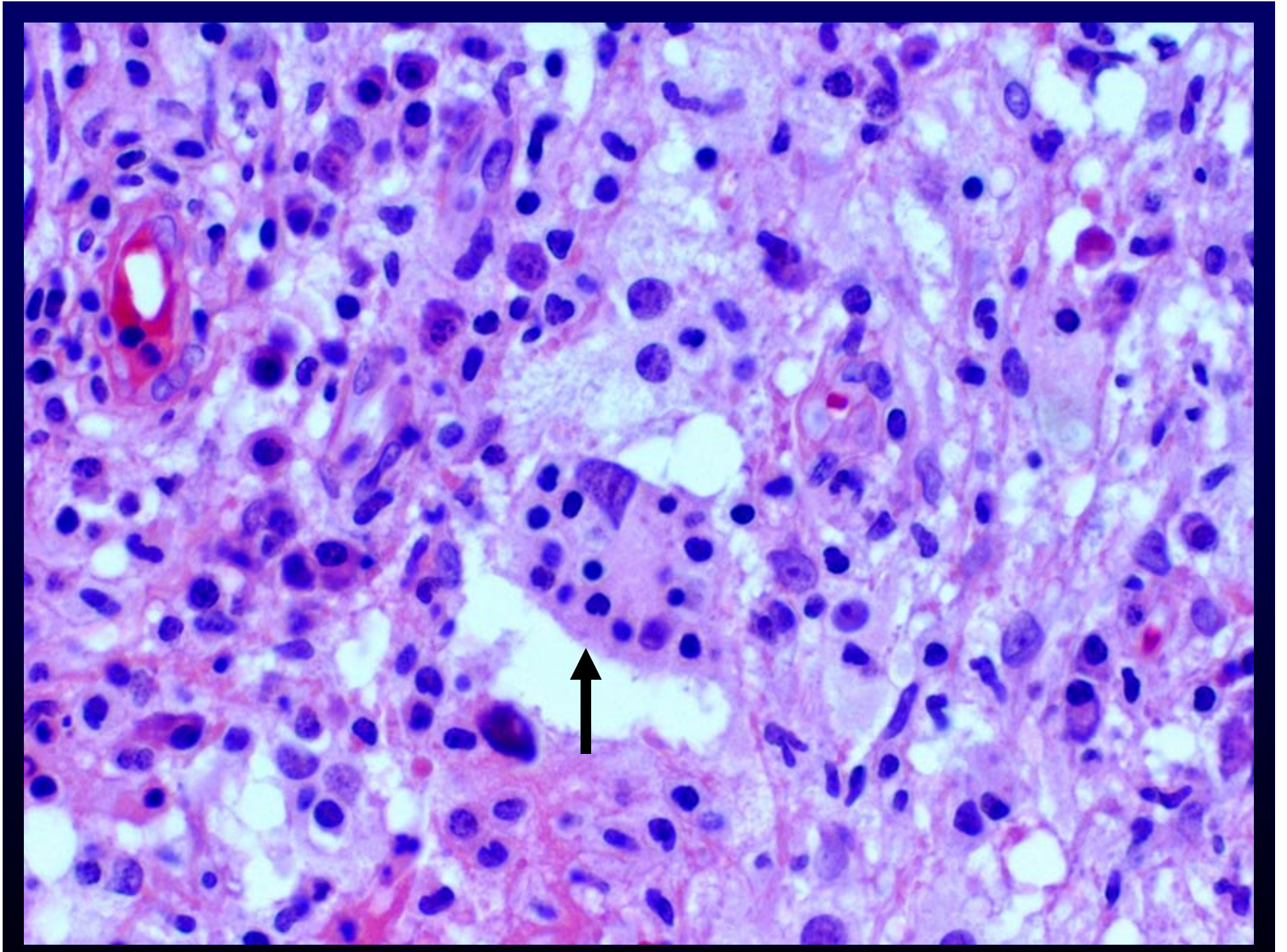
Histopathology:

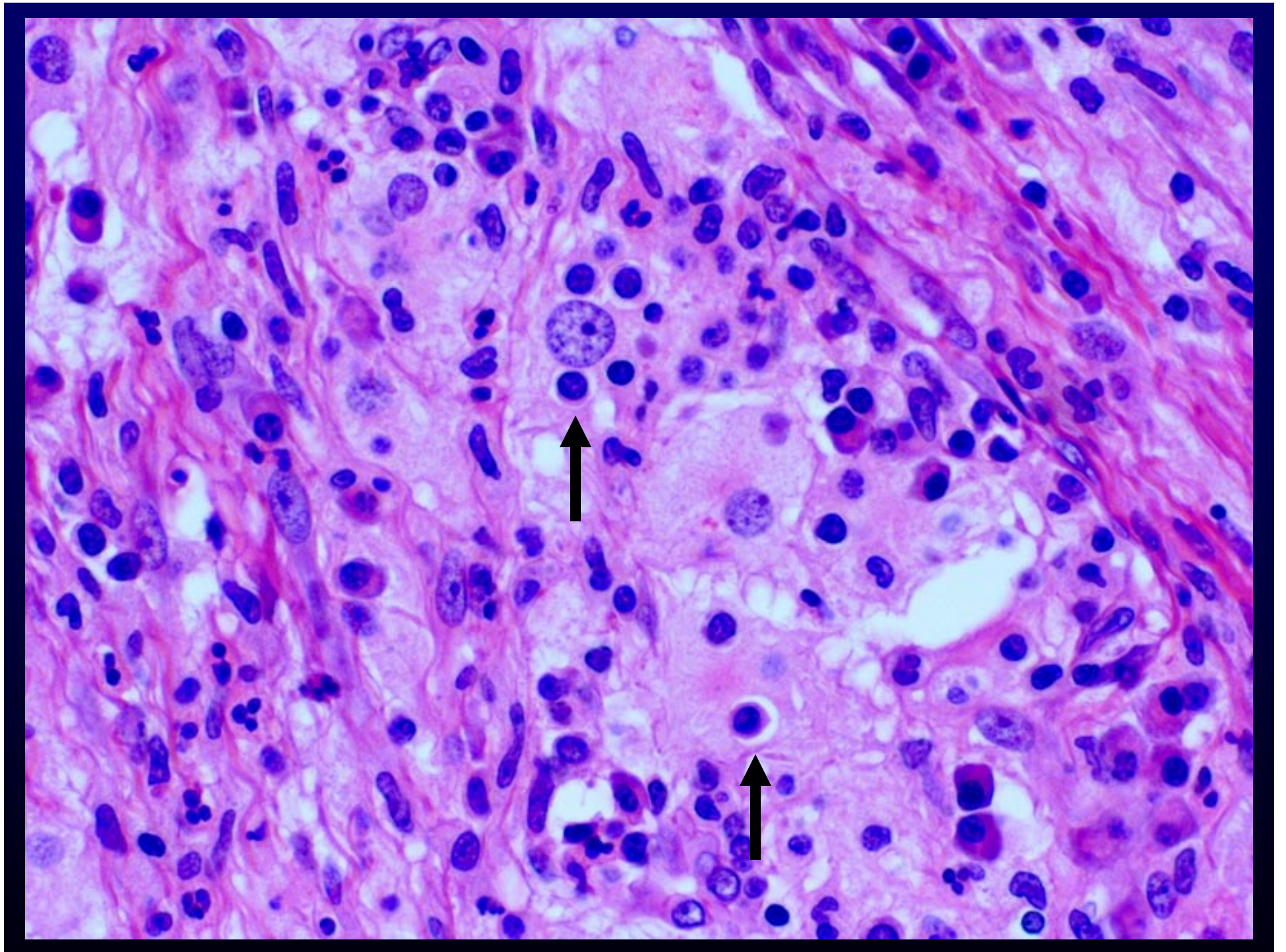
- Mixed inflammatory infiltrate composed of mature lymphocytes, plasma cells, neutrophils
- **Large atypical-appearing histiocytes:**
 - eosinophilic cytoplasm
 - indistinct cell borders
 - round to oval, vesicular nuclei with prominent nucleoli
 - well preserved lymphocytes, plasma cells and red blood cells within cytoplasm (emperipolesis)

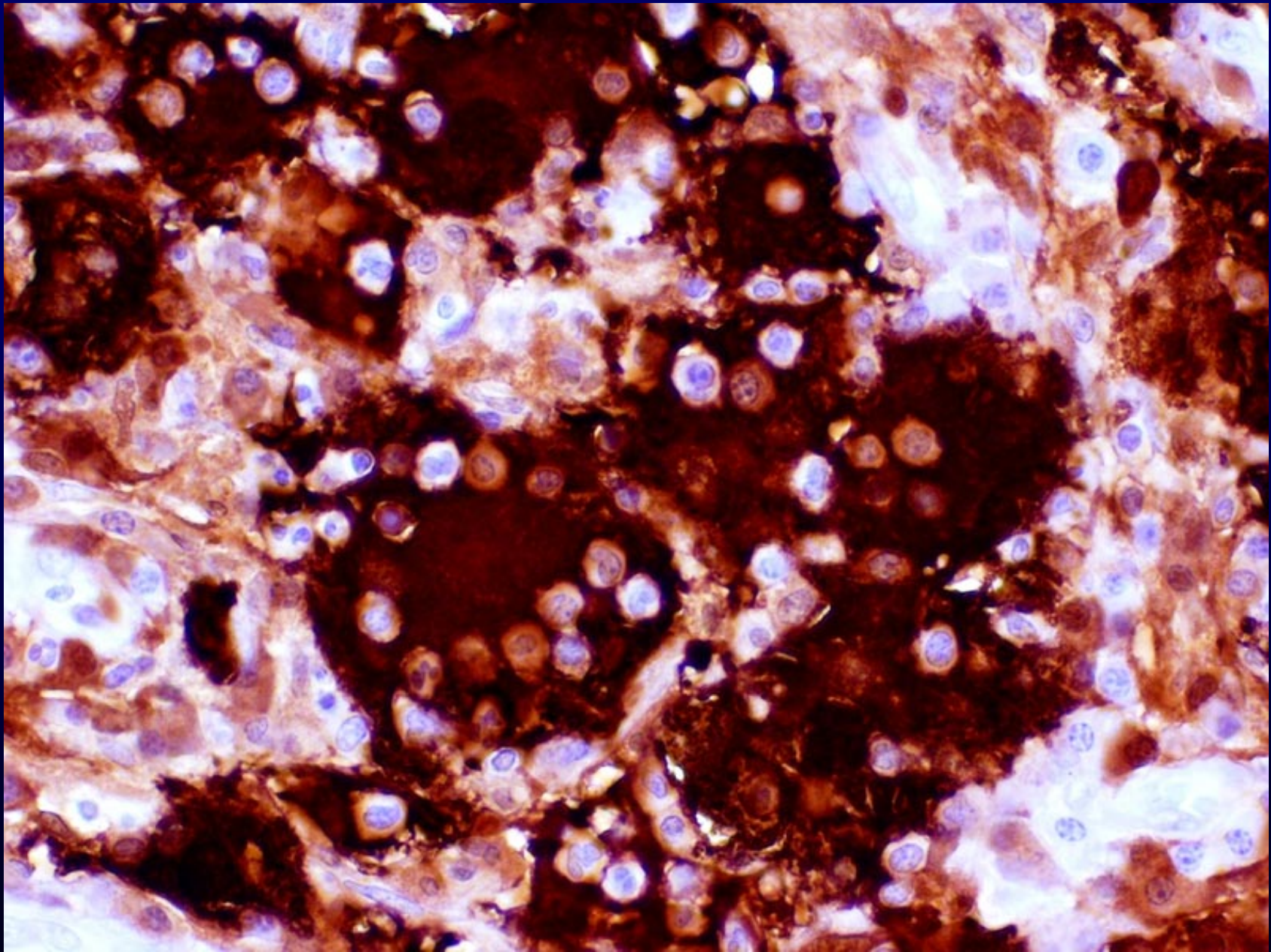
Emperipolesis: (*em* inside + *peri* around
+ *poleomai* to wander about)

“Active penetration by one cell into and
through a larger cell.”

Webster's New World/Stedman's Concise Medical Dictionary







Emperipolesis is often quite striking on immunohistochemistry for S-100.

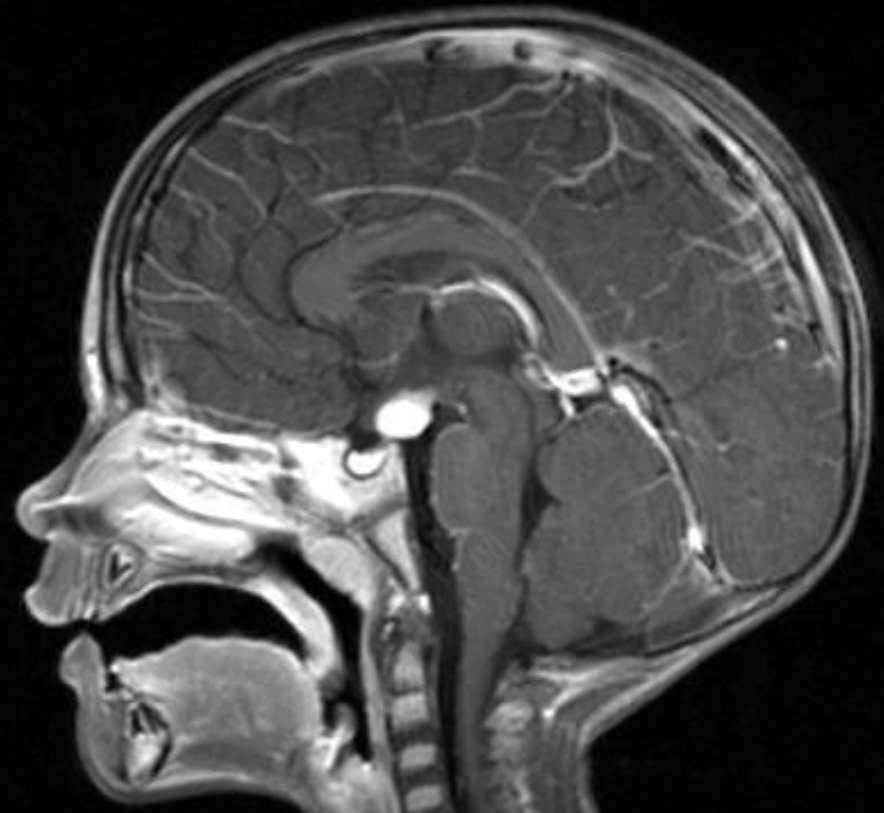
Histiocytoses affecting the CNS

| | CD68 | S-100 | CD1a |
|---|----------|----------------------|----------|
| Langerhans cell histiocytosis: | + | + | + |
| Rosai-Dorfman disease: (sinus histiocytosis with massive lymphadenopathy) | + | + | - |
| | | emperipolesis | |
| Histiocytic sarcoma | + | - | - |
| Erdheim-Chester Disease | + | +/- | - |
| Hemophagocytic Syndrome | + | variable | |
| Juvenile xanthogranuloma | + | - | - |

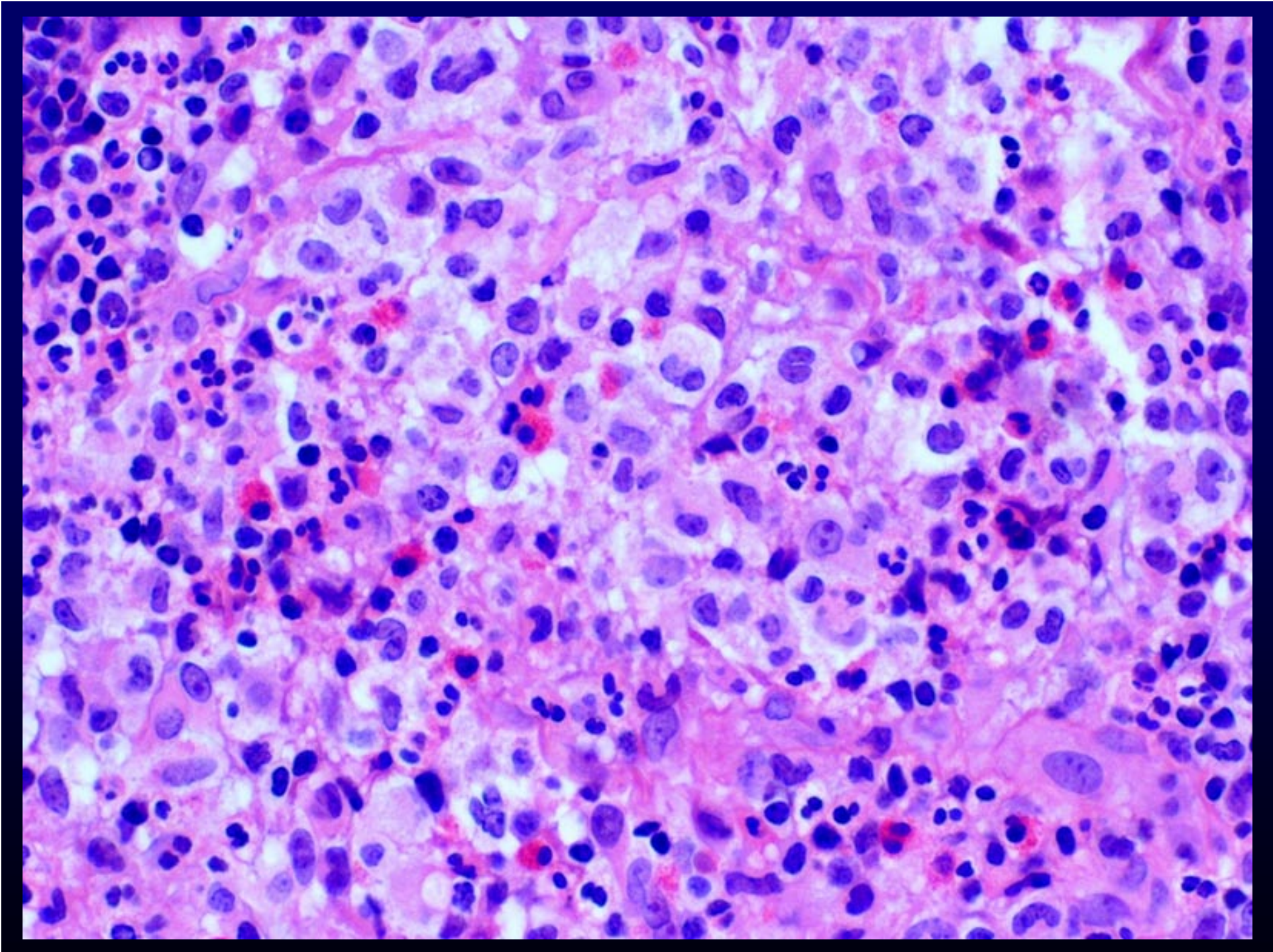
Differential diagnosis of non-meningiomatous dural based lesions:

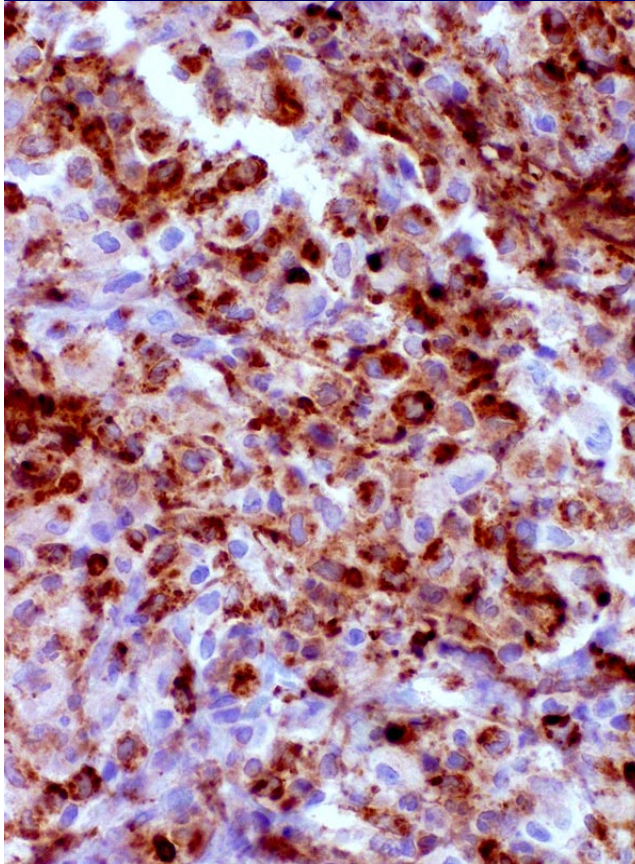
- **Hemangiopericytoma/solitary fibrous tumor**
- **Other sarcomas**
- **Hematologic tumors**
 - **Extramedullary myeloid tumor**
 - **MALT-like lymphomas**
- **Nerve sheath tumors**
- **Granulomatous processes**

Johnson MD, Powell SZ, Boyer PJ, Weil RJ, Moots PL. Dural lesions mimicking meningiomas. *Hum Pathol* 2002;33:1211-26.

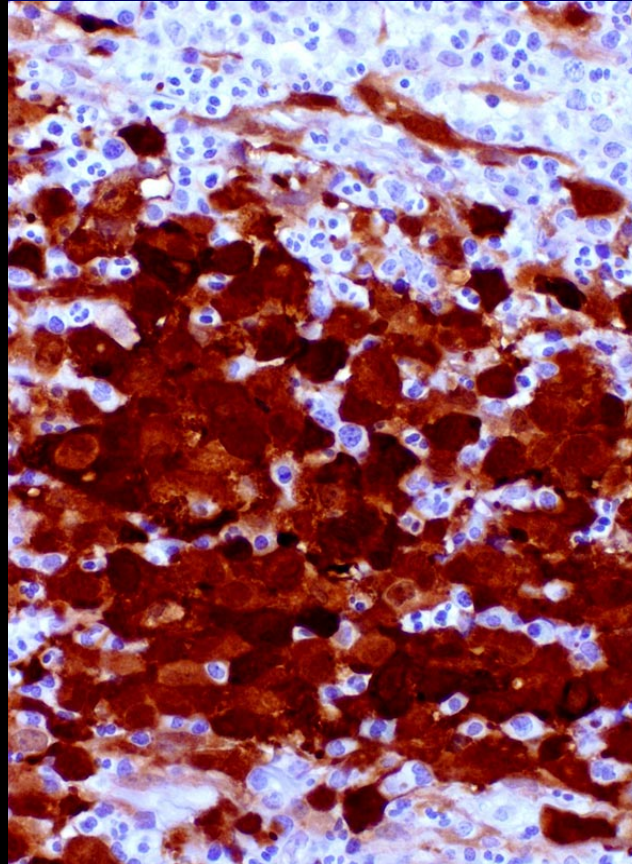


Child with diabetes insipidus and hypothalamic mass.

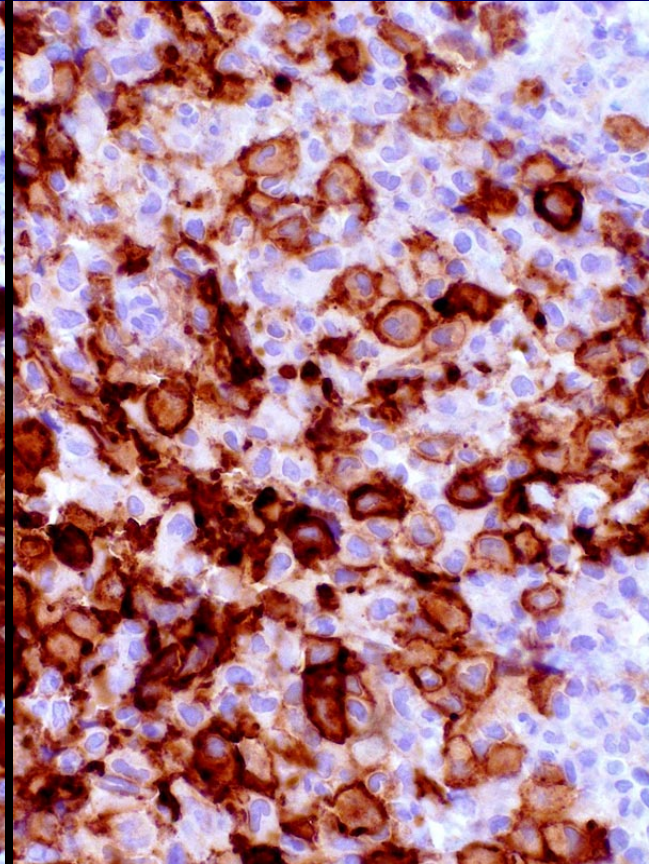




CD68



S-100



CD1A

(Also CD 207/Langerin +)

Langerhans Cell Histiocytoses (Formerly histiocytosis X)

Focal

Eosinophilic granuloma

Multifocal

Hand-Schüller-Christian

Diabetes insipidus, proptosis, and lytic bone lesions

Disseminated

Letterer-Siwe disease

Congenital self healing (Hashimoto-Pritzker)



**Birbeck Granules
Langerhans Histiocytes**

