

**USCAP 2008 Specialty Conference:**

**NEUROPATHOLOGY: Case #1**



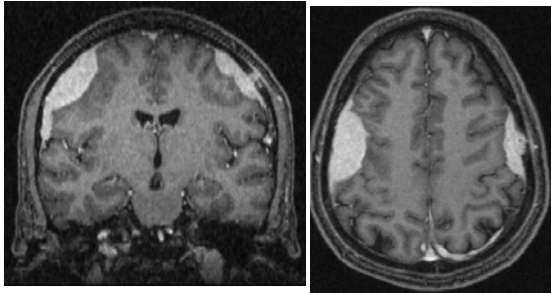
**Anthony T. Yachnis, MD**

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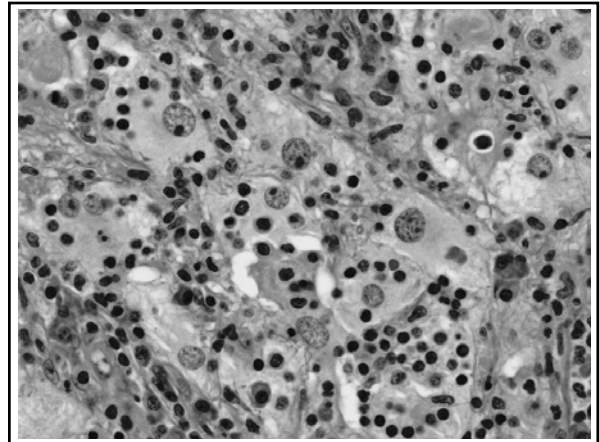
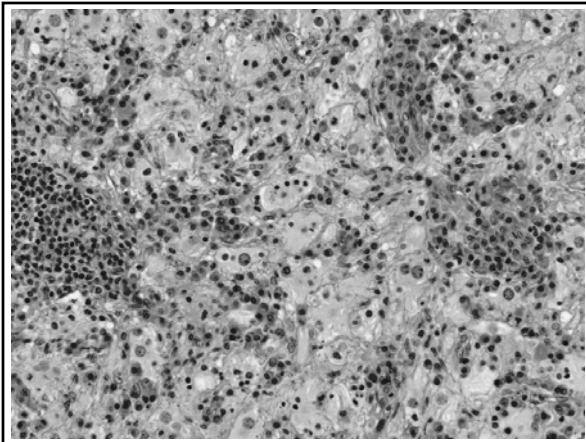
Department of Pathology, Immunology  
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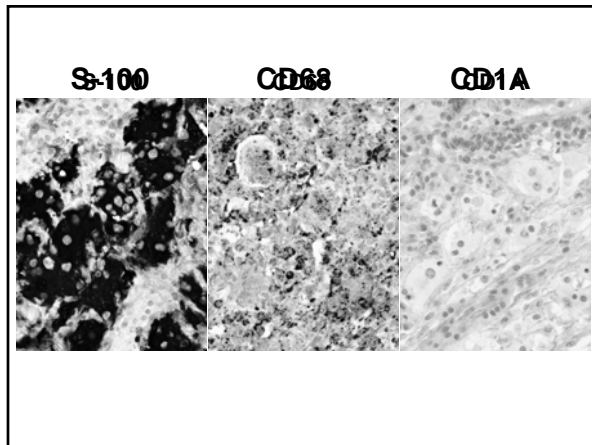
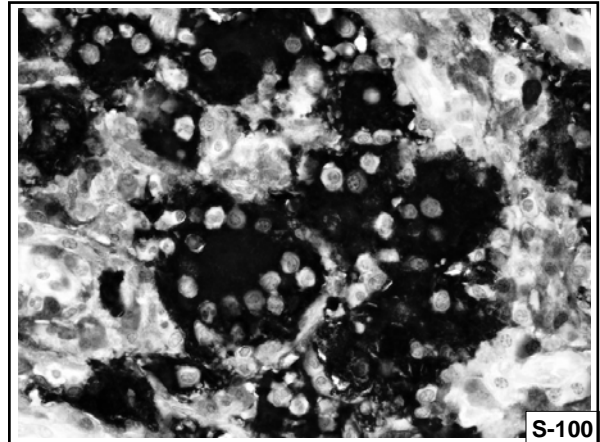
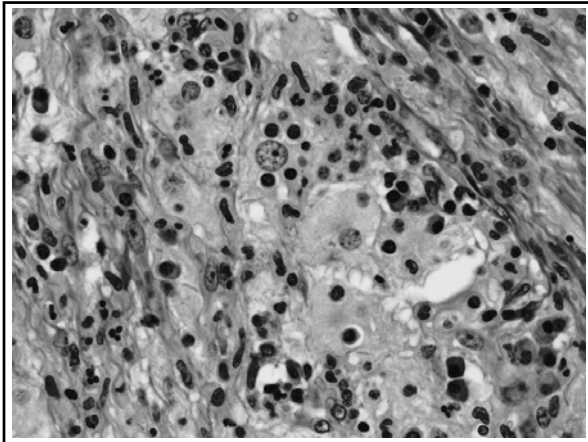
**Clinical History**

- 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 craniotomies with excision of the lesions in two separate operations



**T1-weighted post-contrast MRI**





**Diagnosis:**

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

**Rosai-Dorfman Disease**

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• **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:***

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- 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

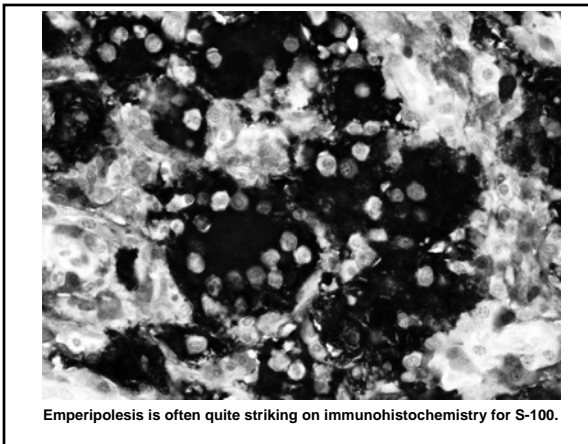
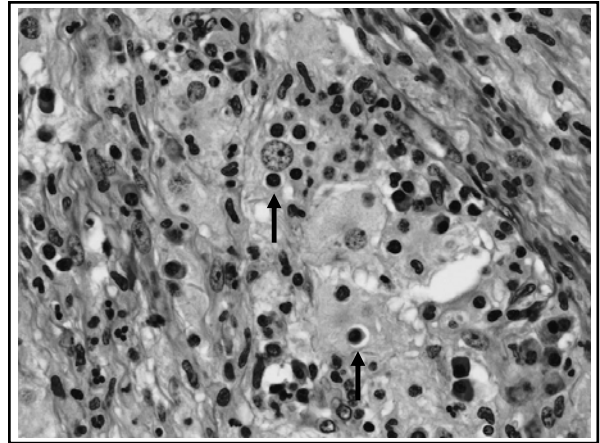
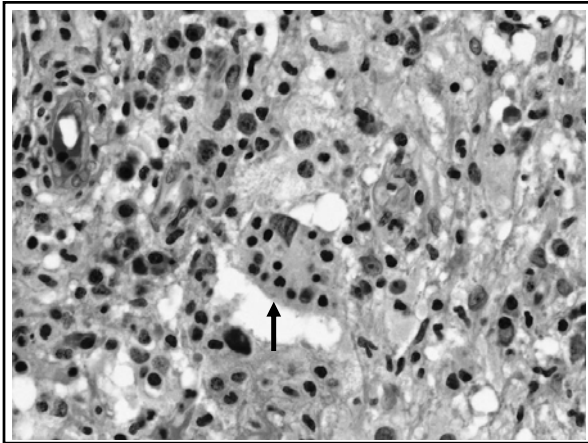
### Histopathology:

- 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.

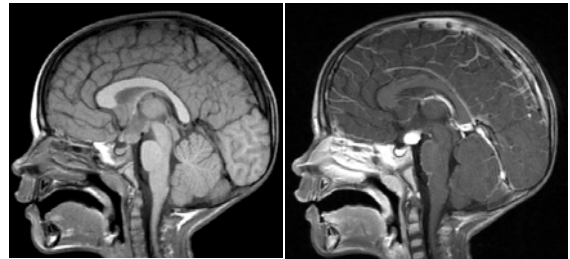
## Histiocytoses affecting the CNS

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

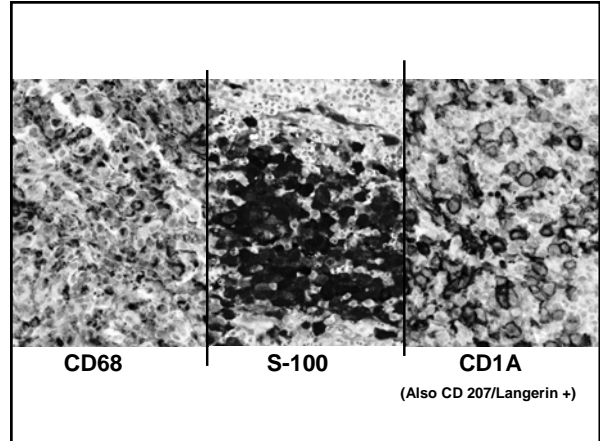
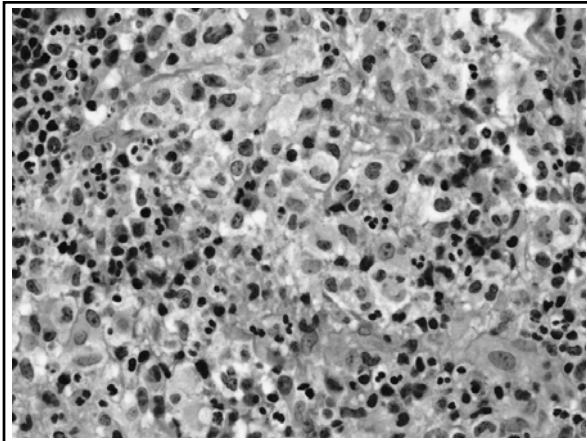
**Differential diagnosis of non-meningiomatous dural based lesions:**

- 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.



**Langerhans Cell Histiocytoses (Formerly histiocytosis X)**

<b>Focal</b>	Eosinophilic granuloma
<b>Multifocal</b>	Hand-Schüller-Christian Diabetes insipidus, proptosis, and lytic bone lesions
<b>Disseminated</b>	Letterer-Siwe disease
<b>Congenital self healing</b>	(Hashimoto-Pritzker)



Birbeck Granules  
Langerhans Histiocytes