

# **Case 10**

## **Diagnostic Slide Session, 2015**

### **American Association of Neuropathologists**

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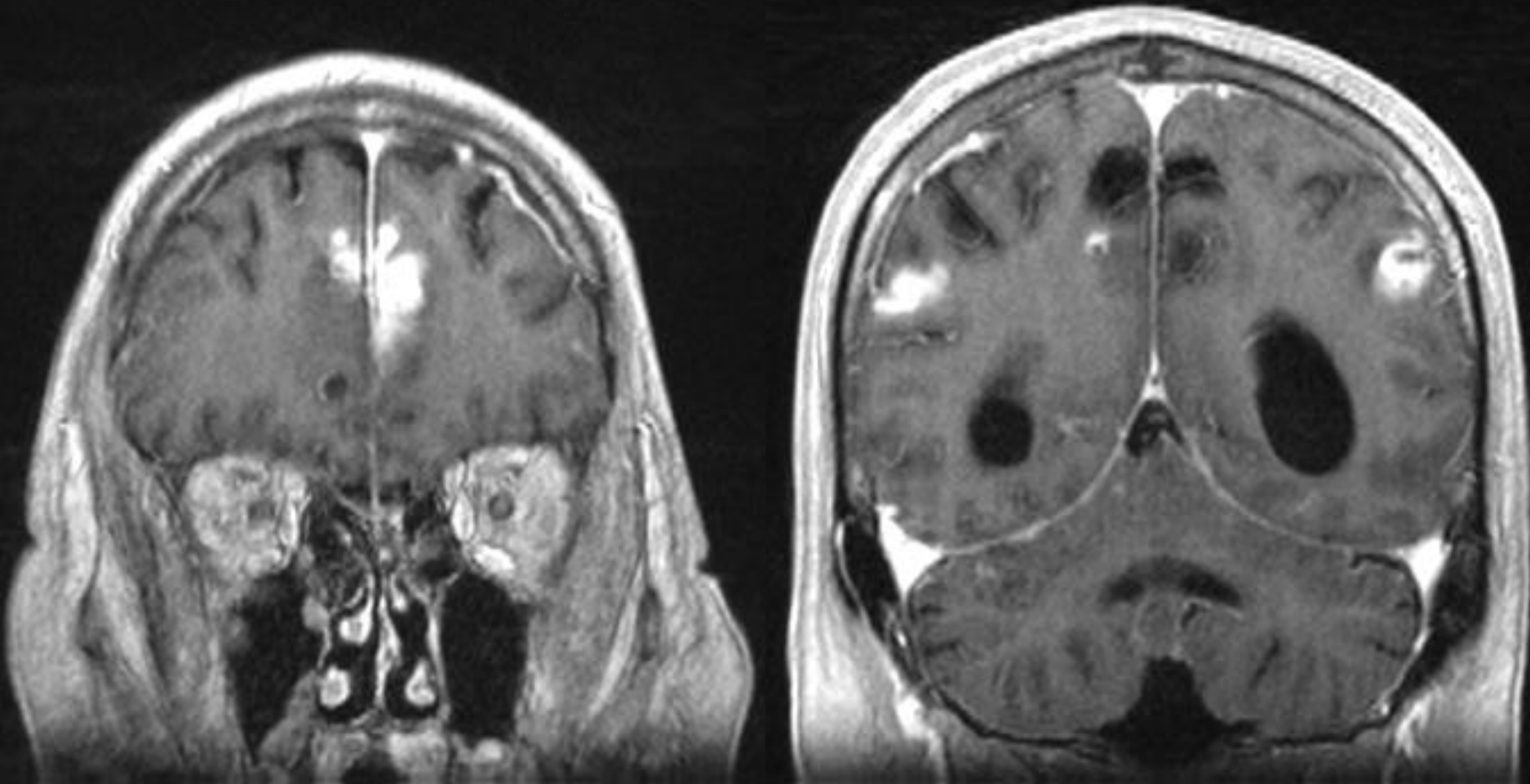


**DEPARTMENT OF PATHOLOGY**  
UNIVERSITY OF MICHIGAN  
HEALTH SYSTEM

# Clinical History

- 62-year-old man with CAD and HTN presented with 2 week history of decreased appetite, neck/back pain, nausea, vomiting
- Admitted to an outside hospital 1/8/2014
- Suffered tonic-clonic seizure, intubated

**MRI, Outside Hospital**  
**T1-weighted, post-contrast**



# Laboratory Work-up

- **Lumbar puncture**

Protein: 539 mg/dL

normal = 15–45 mg/dL

Gram stain/culture:

Negative

- **Urine Histoplasma Ag**

- Positive

- Negative on repeat

- **ACE, CSF**

6.4 U/L

normal = 0.0 – 2.5 U/L

- **Negative tests**

Blastomyces Ag (CSF)

Brucella Ab

CMV, EBV, JCV, VZV PCR (CSF)

Enterovirus PCR (CSF)

Fungal serology (CSF)

HIV-1 PCR

Leptospira Ab

Lyme (CSF serology)

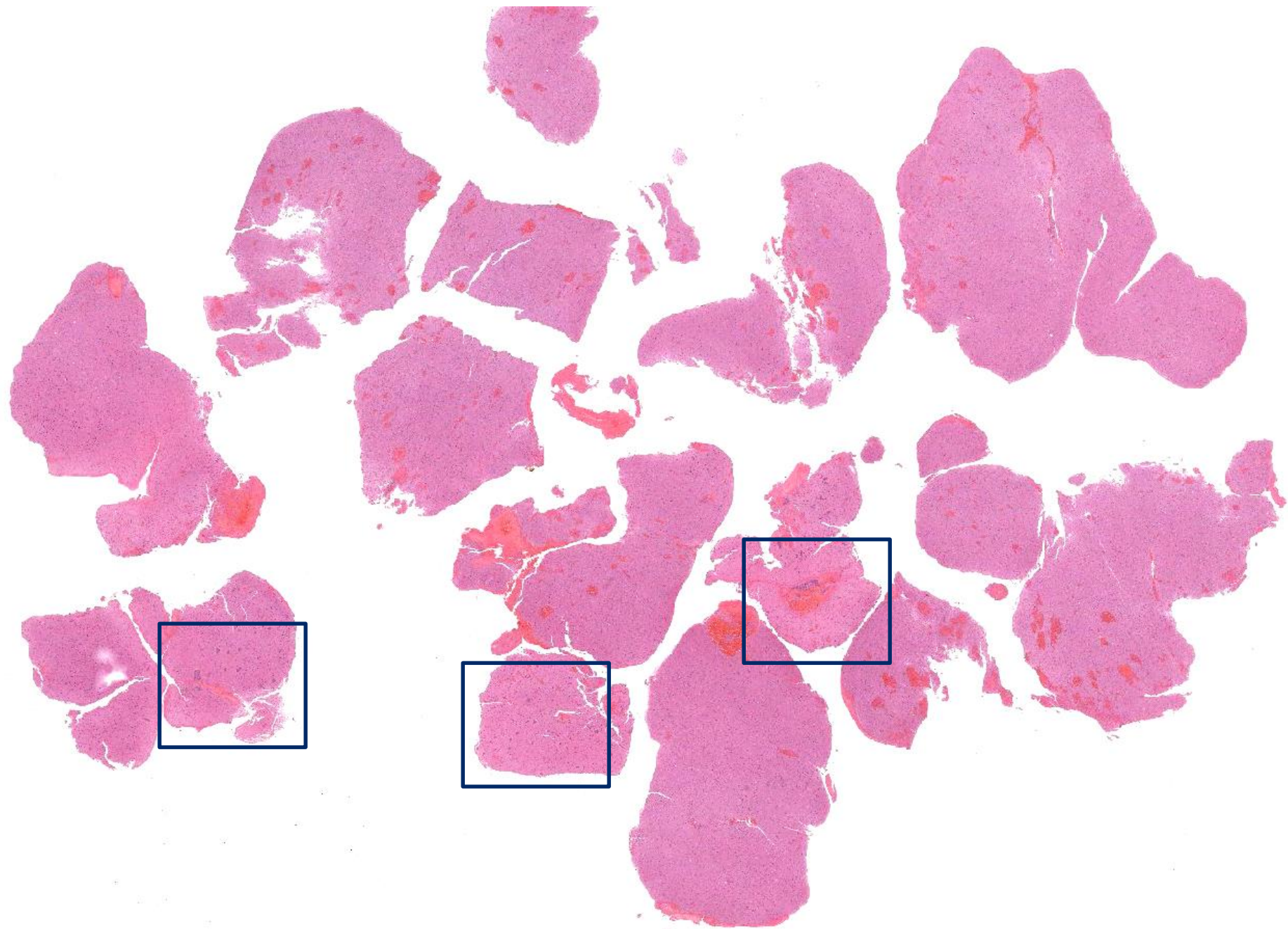
Q Fever

Mycobacterium TB PCR (CSF)

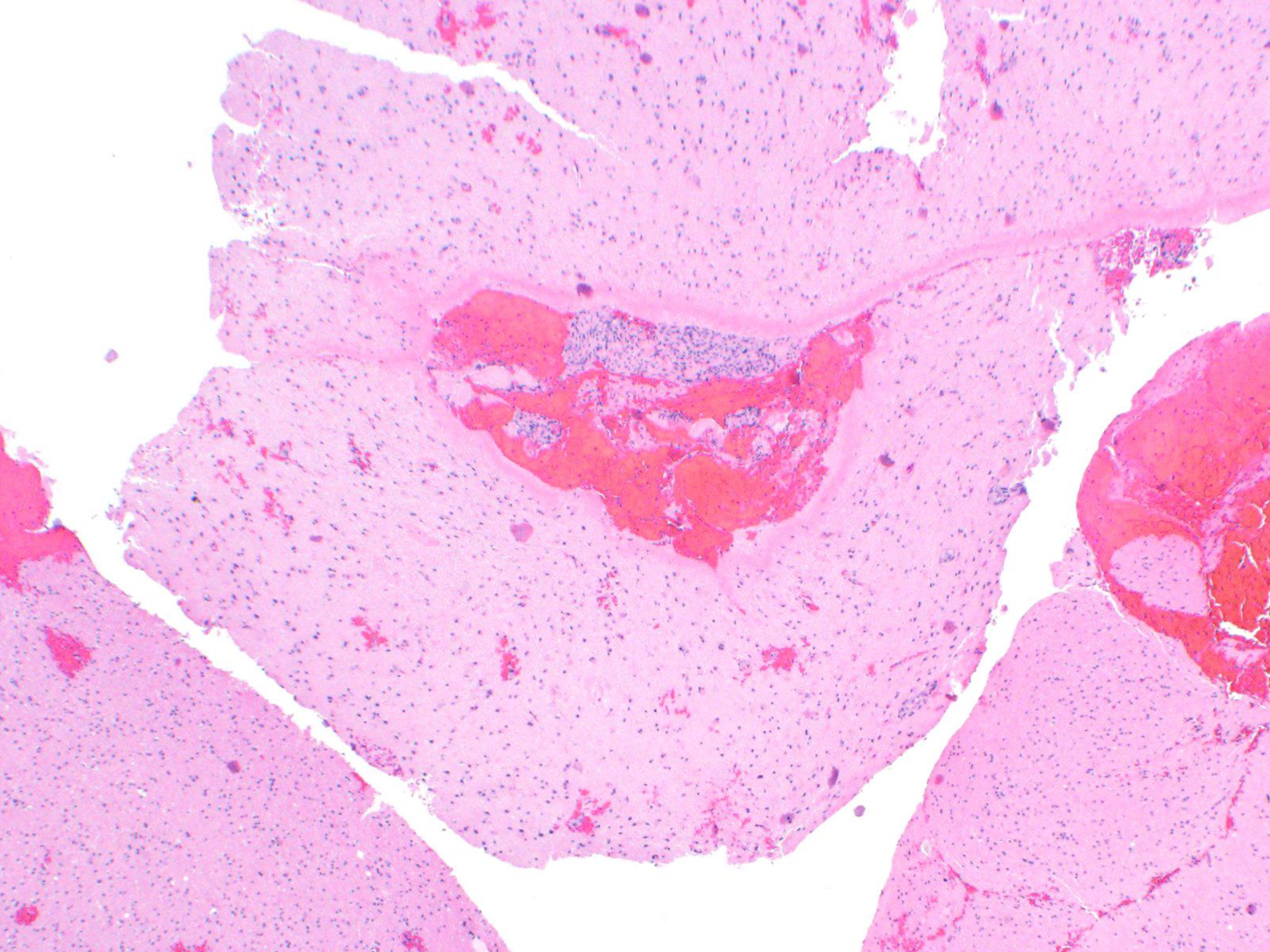
VDRL (CSF)

Paraneoplastic autoAb (CSF)

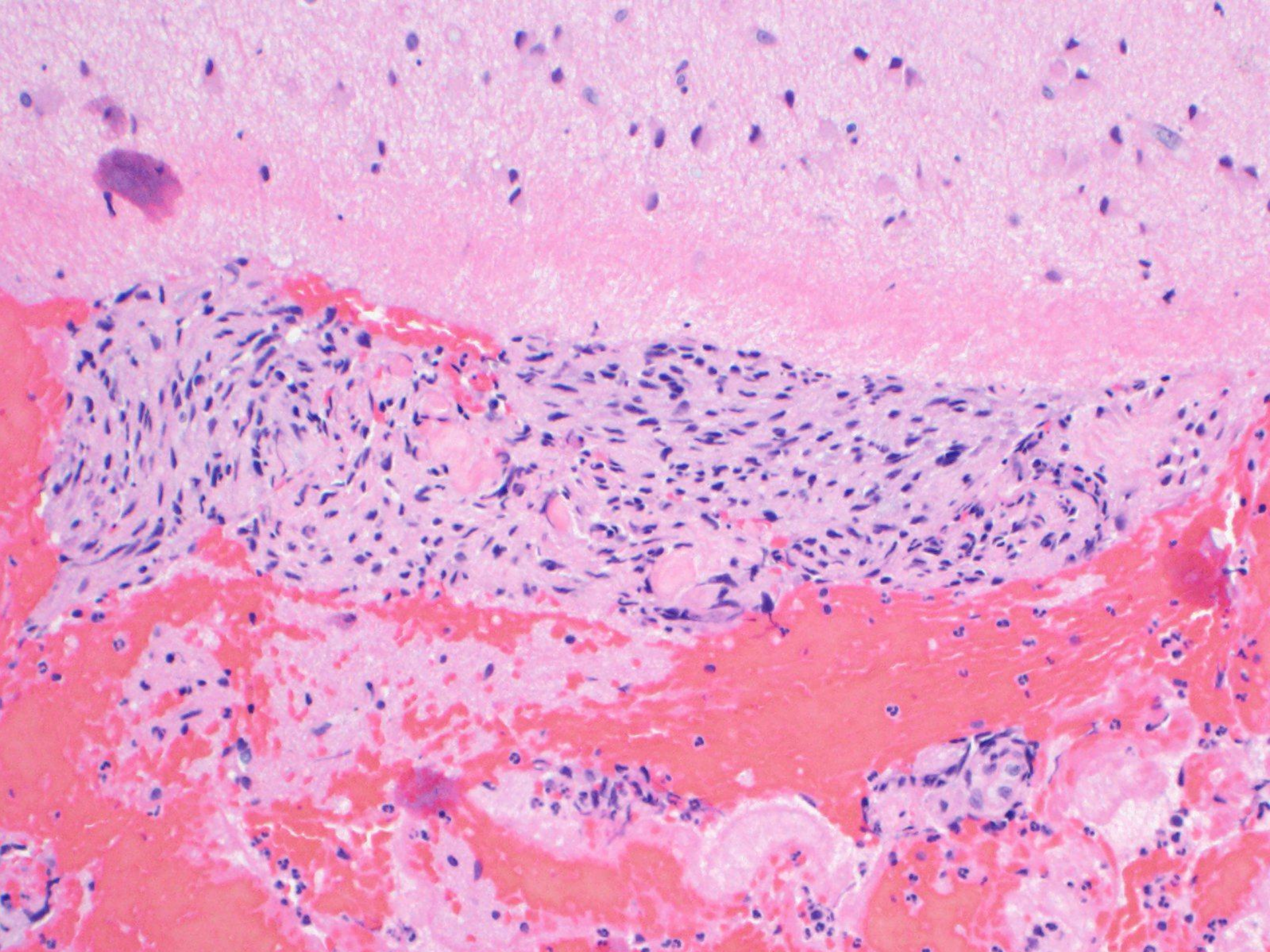
**Biopsy**  
2/11/2014



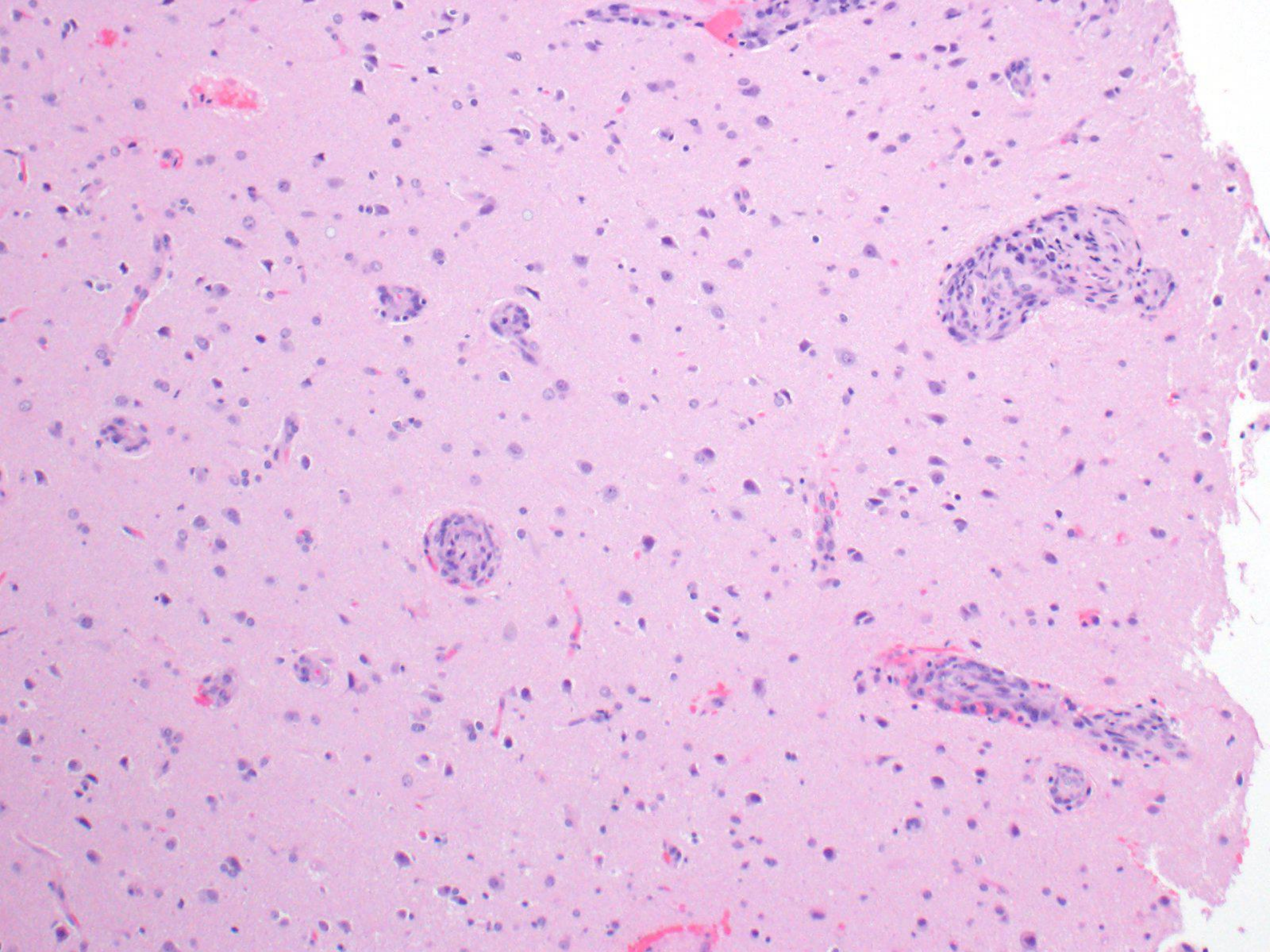




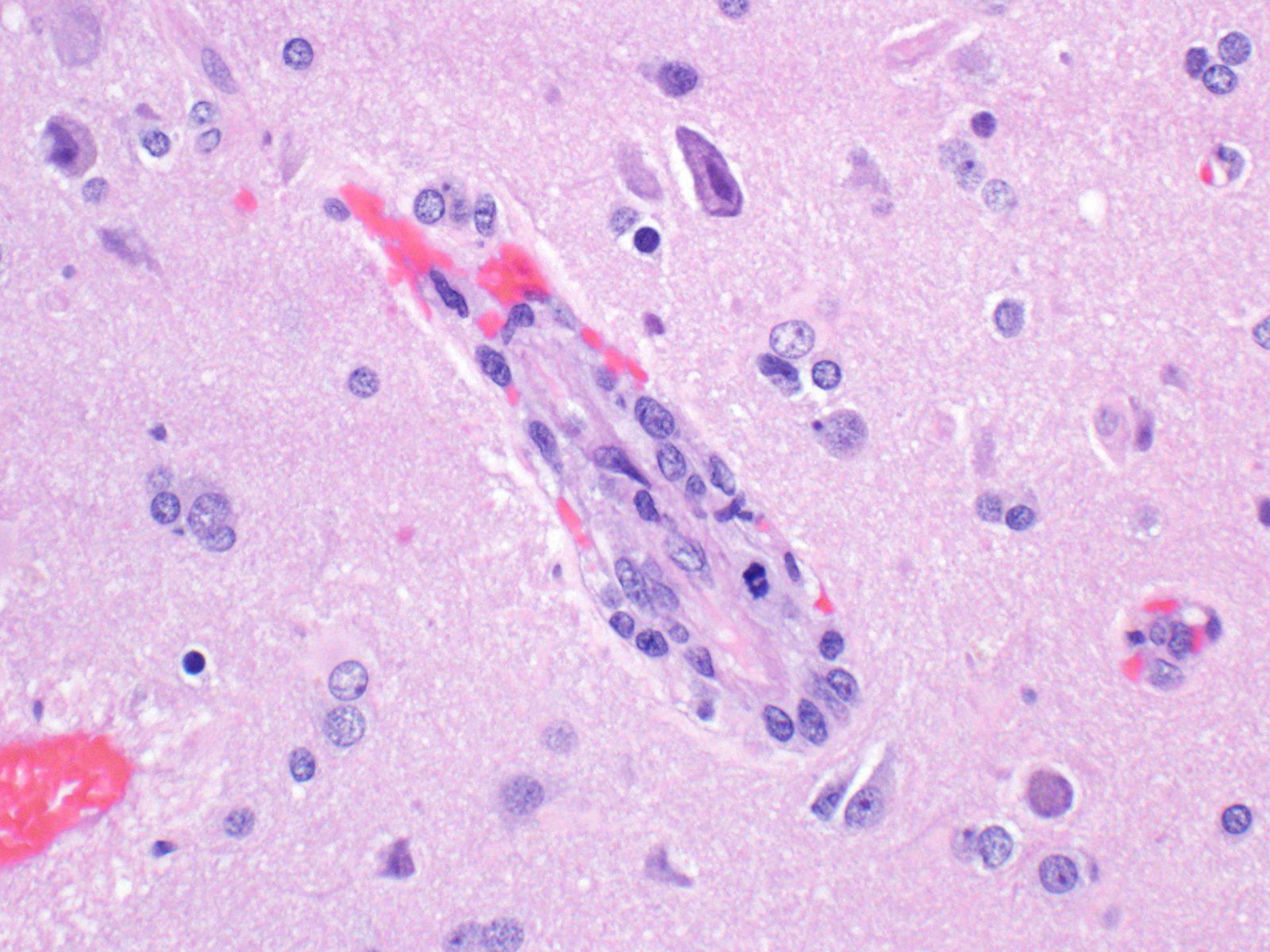




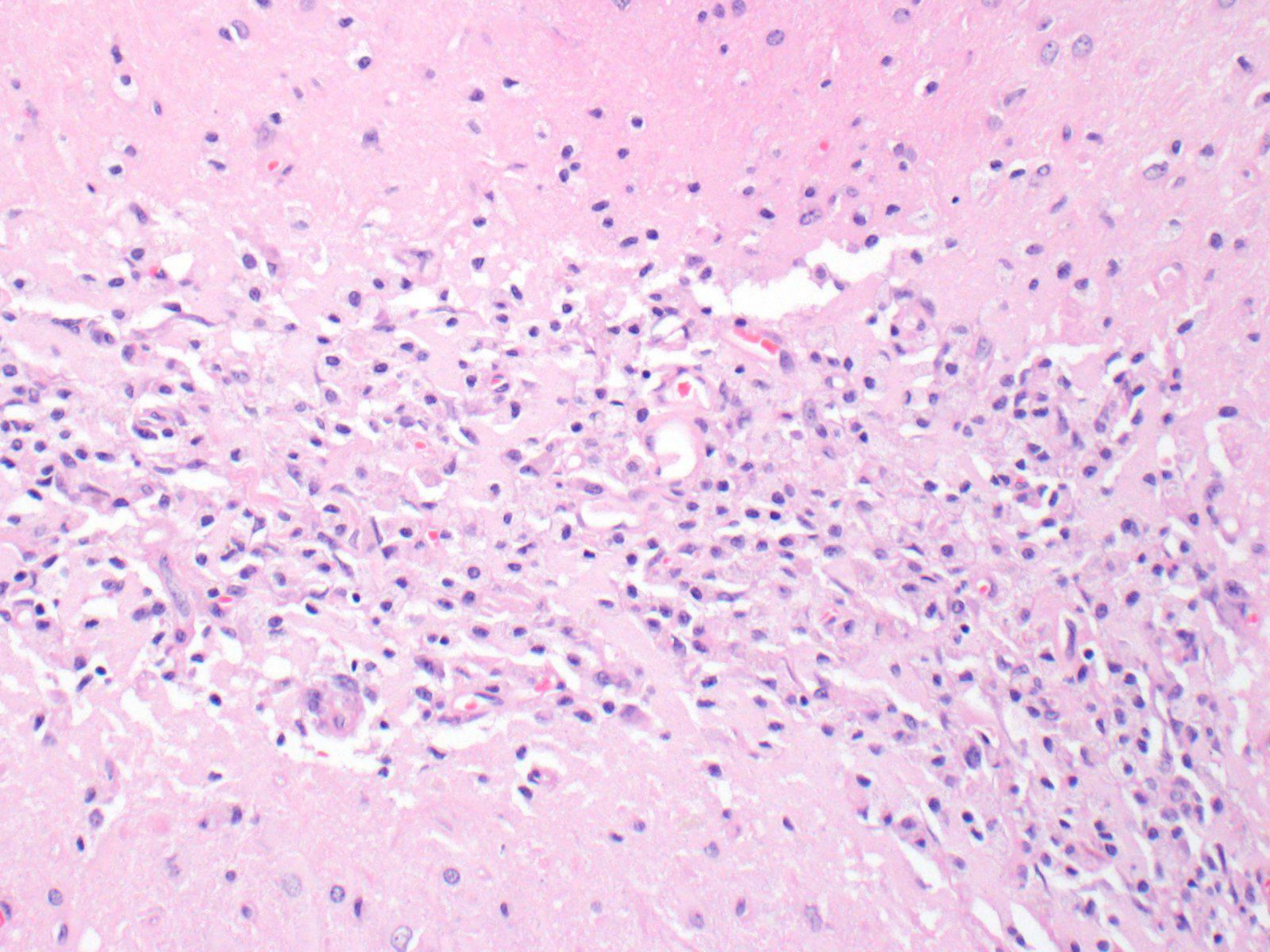














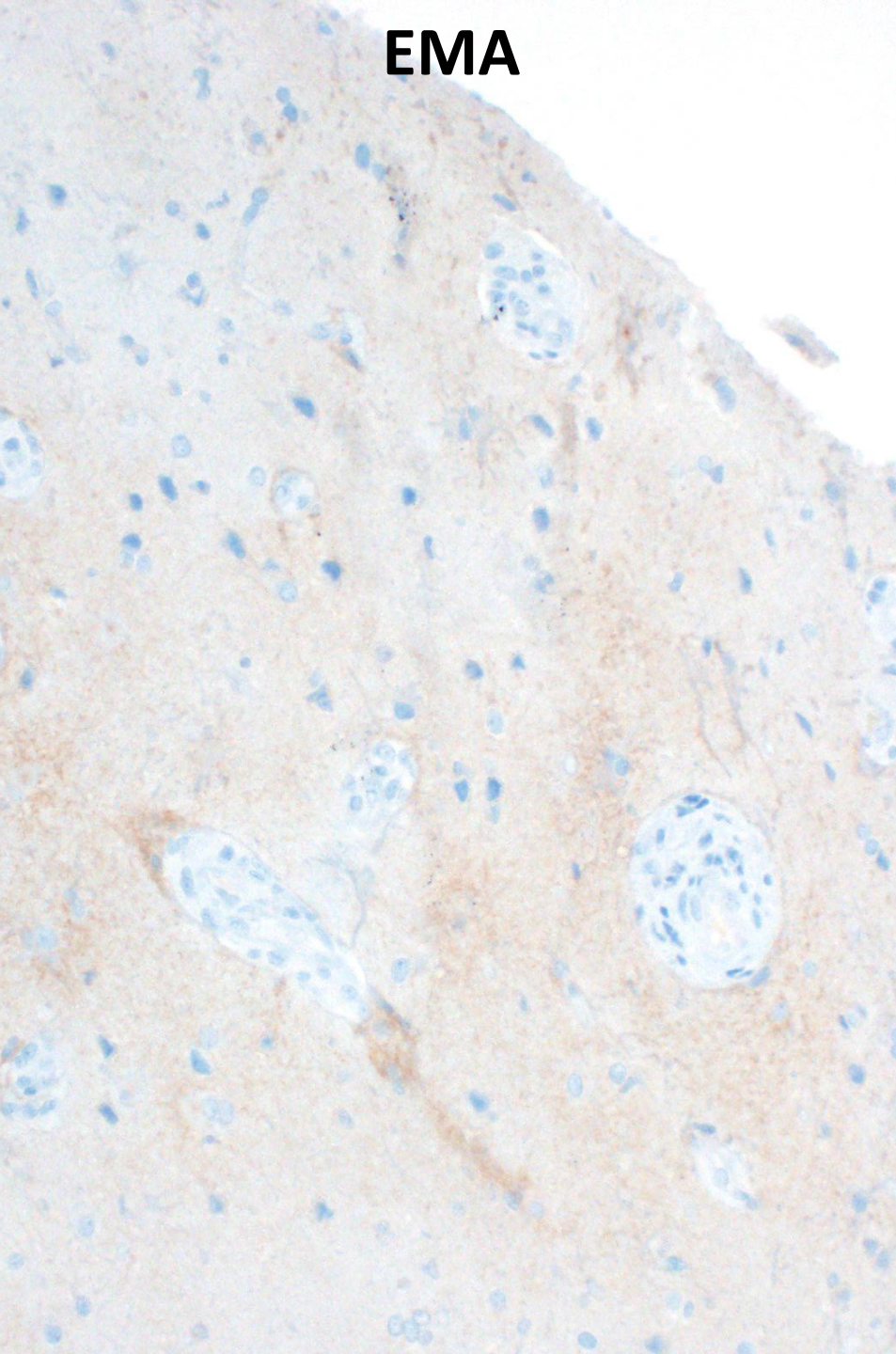
# Biopsy Discussion

- Differential diagnosis?
- Stains?

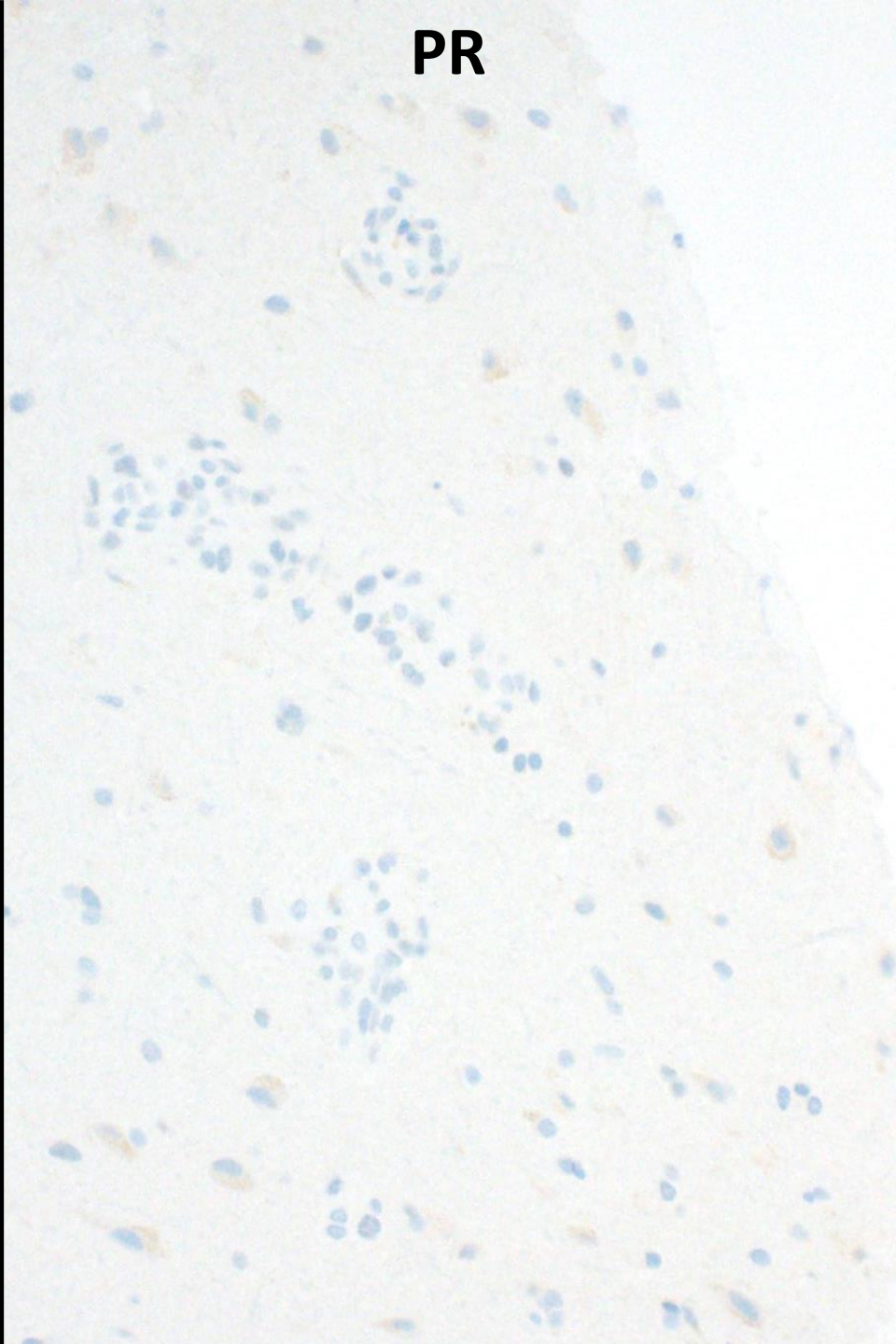
# Differential Diagnosis

- Meningioangiomatosis
- Vasocentric/perivascular process
  - Neoplastic
  - Non-neoplastic

**EMA**

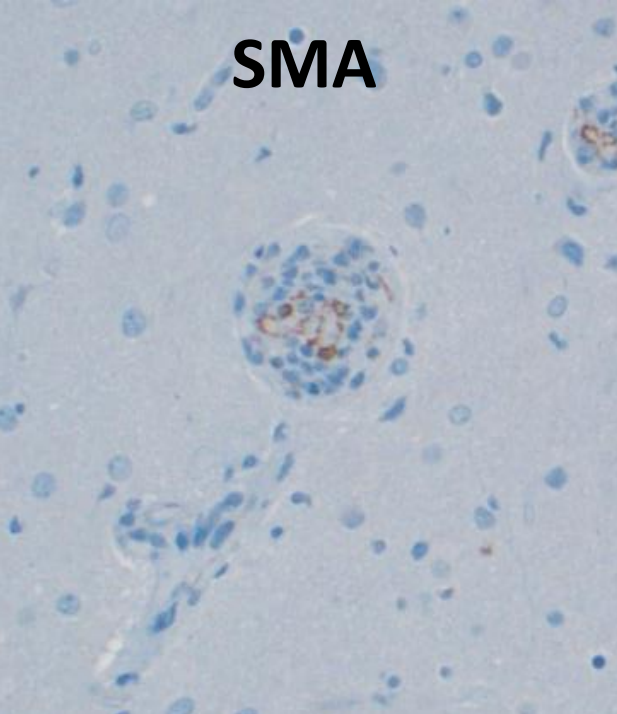


**PR**

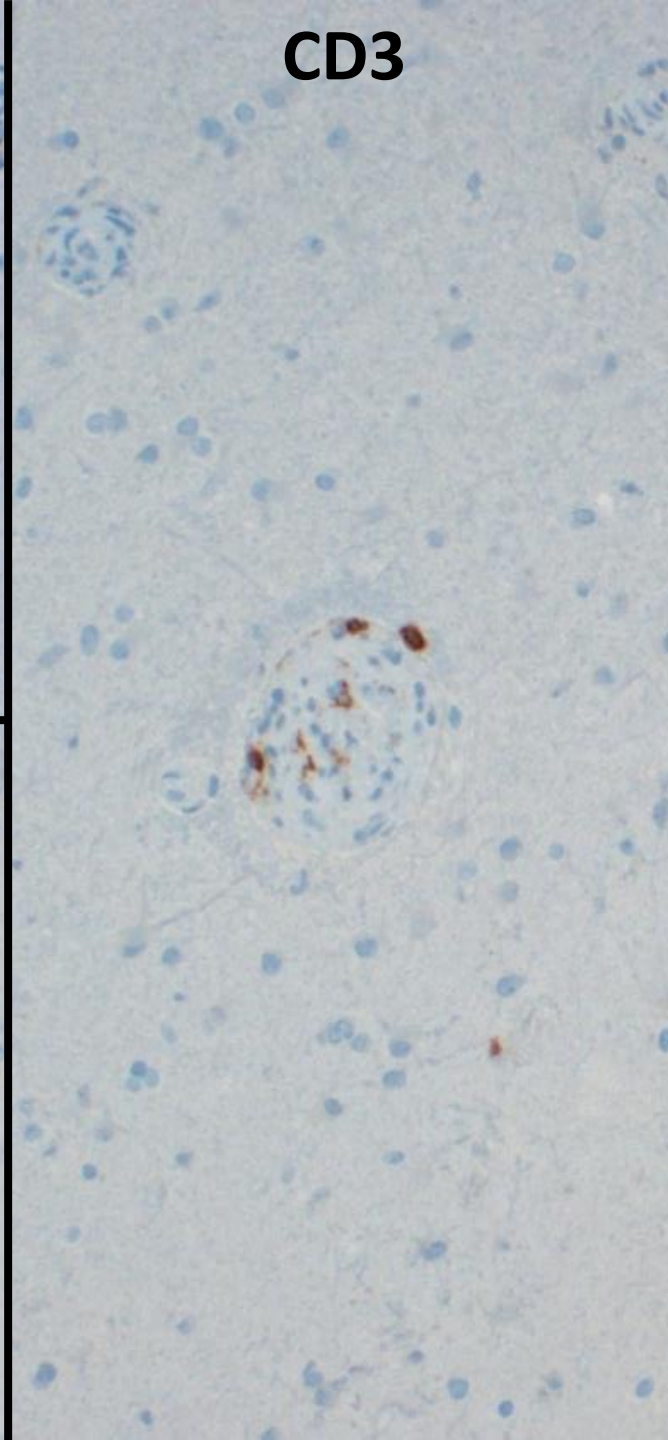




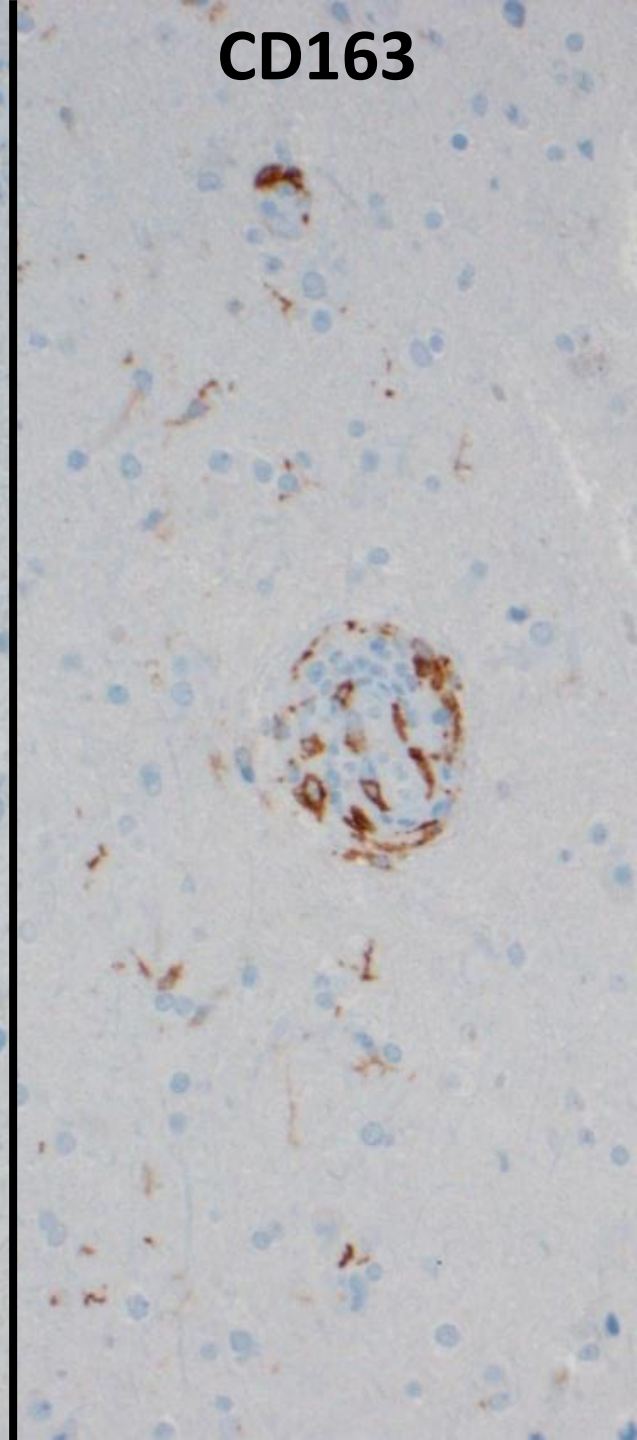
**SMA**



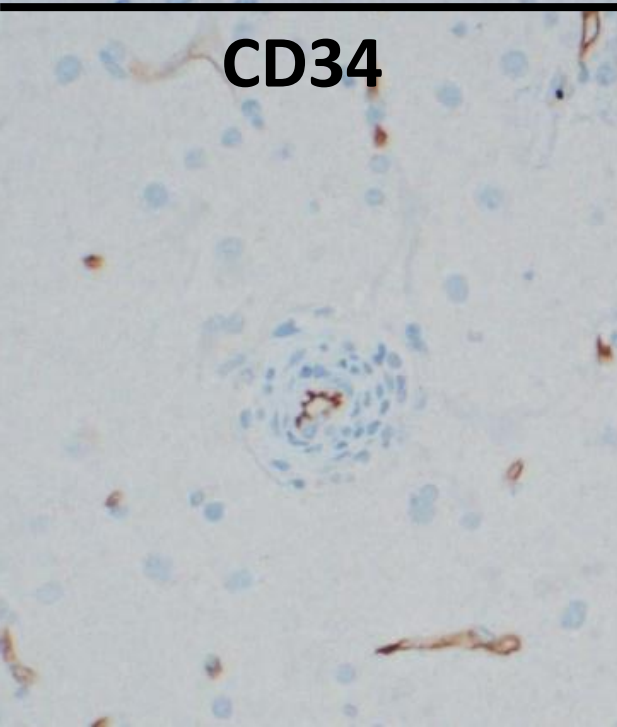
**CD3**



**CD163**

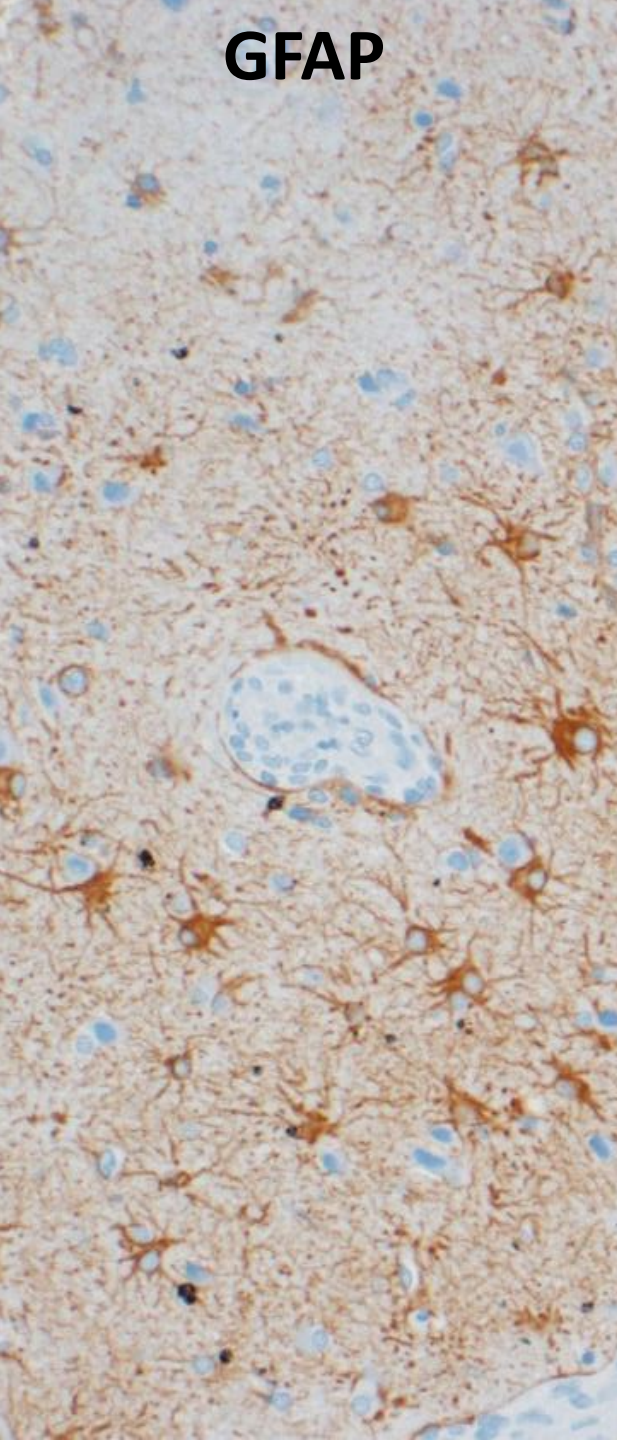


**CD34**

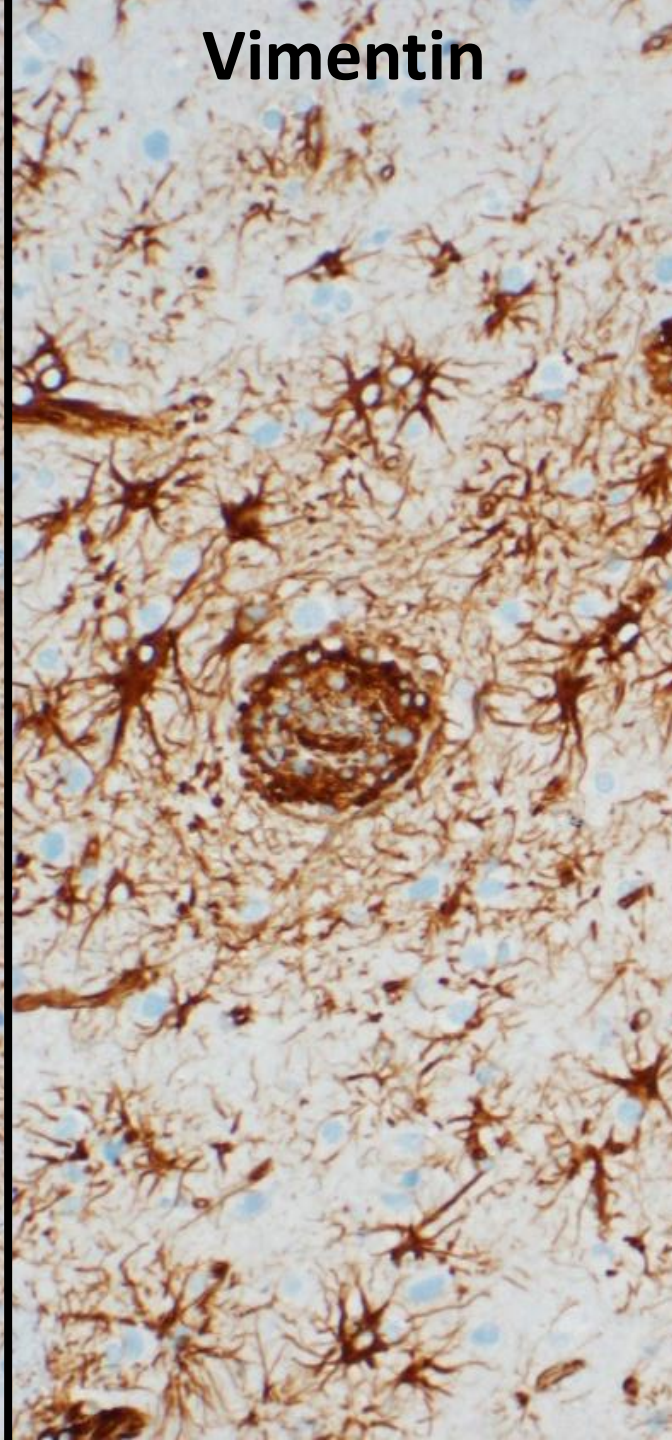




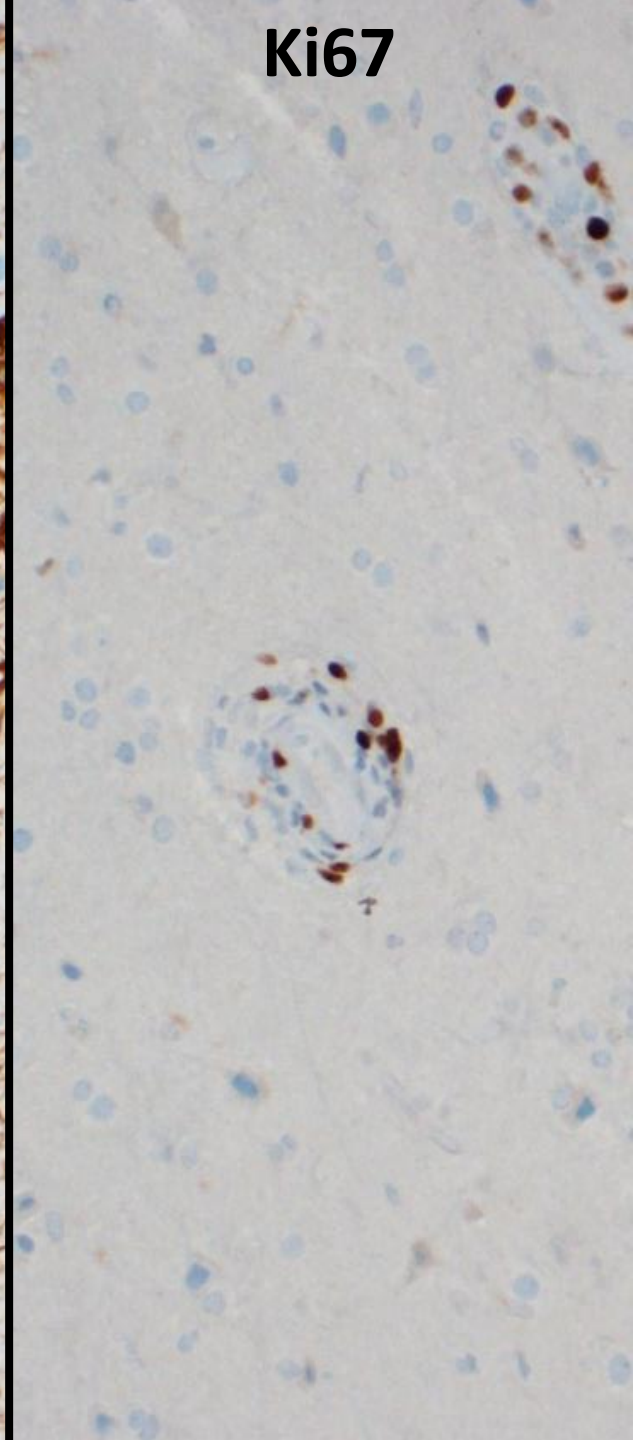
**GFAP**



**Vimentin**

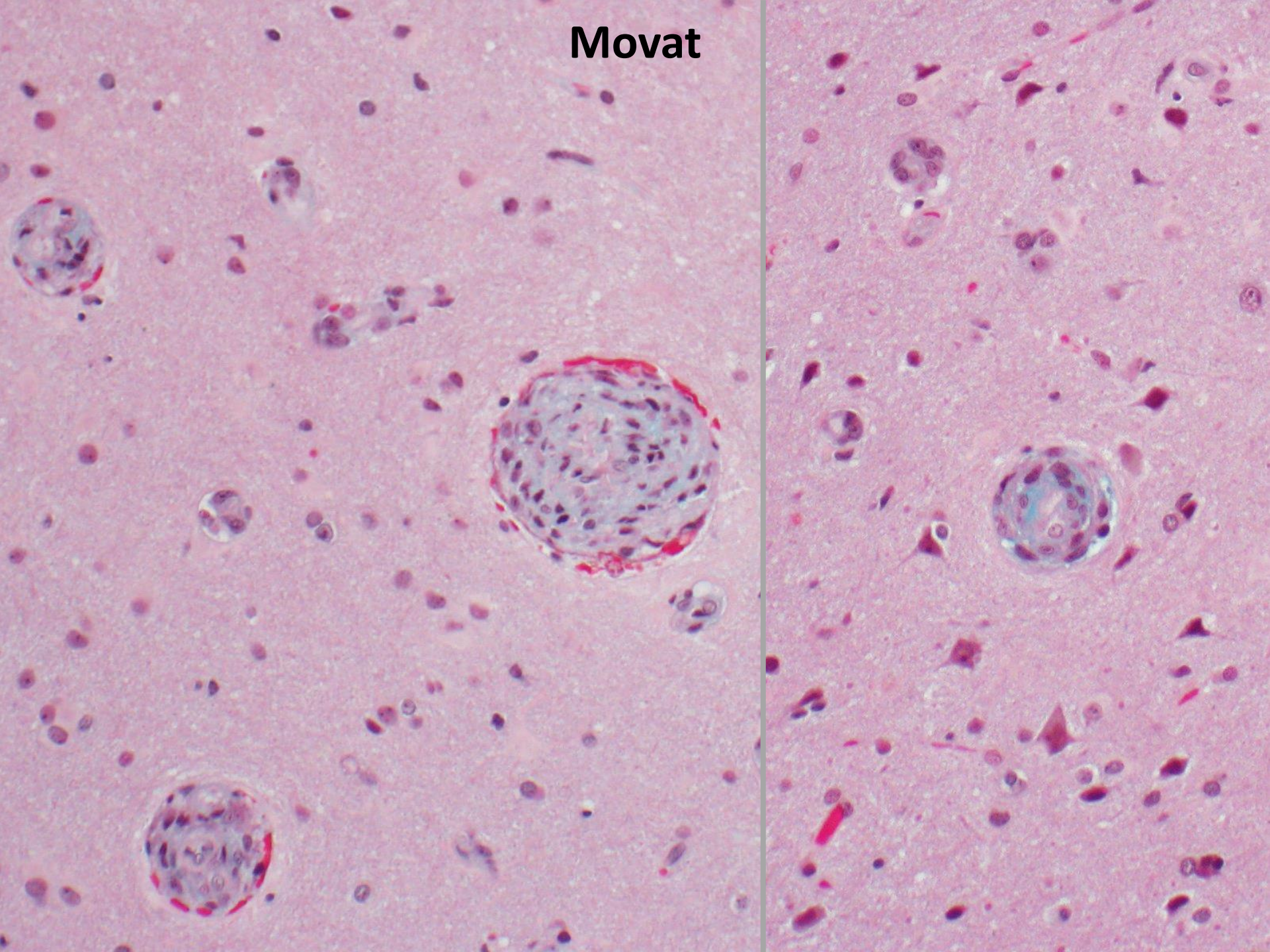


**Ki67**



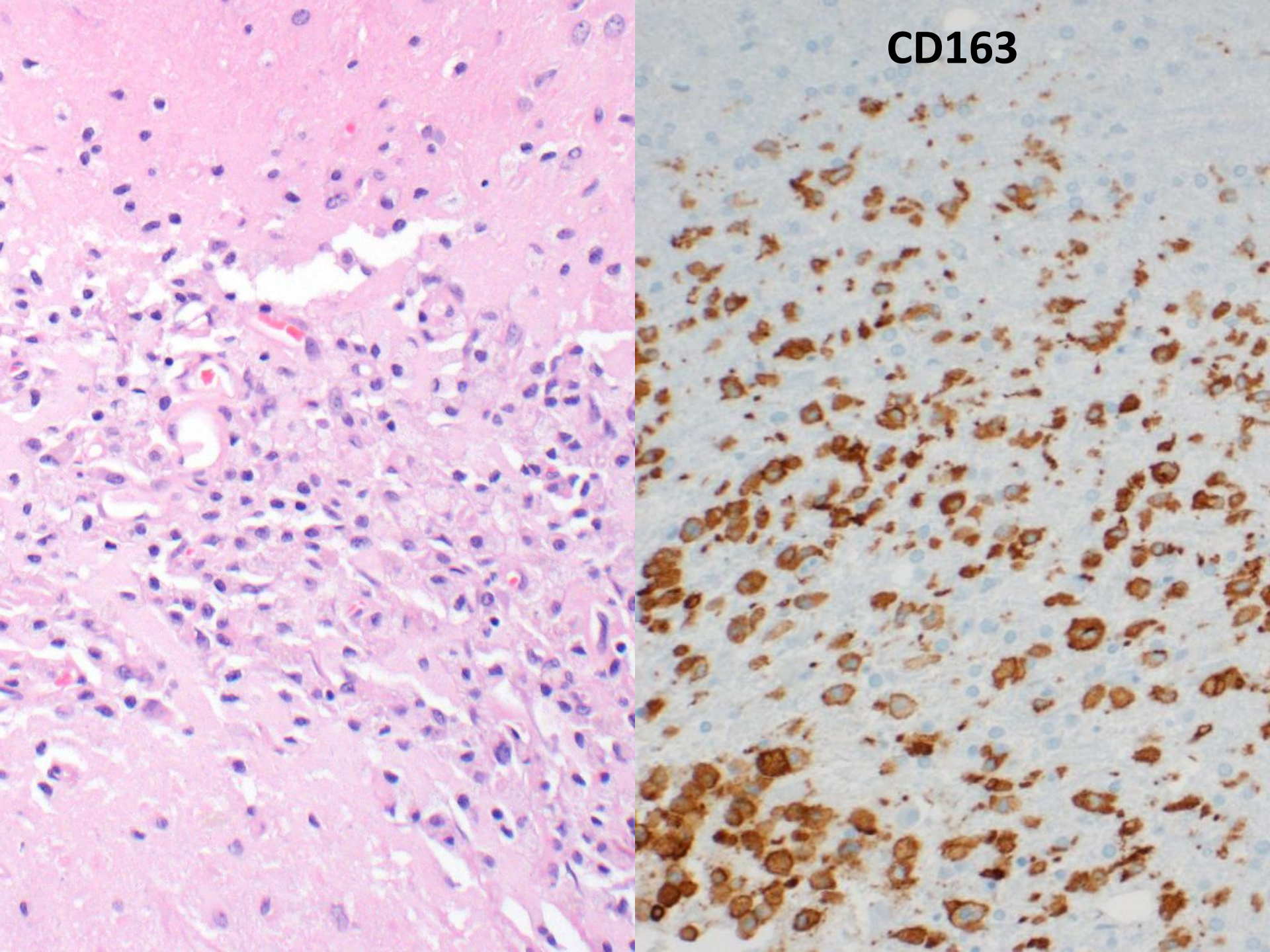


**Movat**





**CD163**



# Other Stains

- **Negative**

CD20

CD1a

S100

Iron

Tissue Gram stain

GMS

Fite

Spirochete

# Biopsy Diagnosis

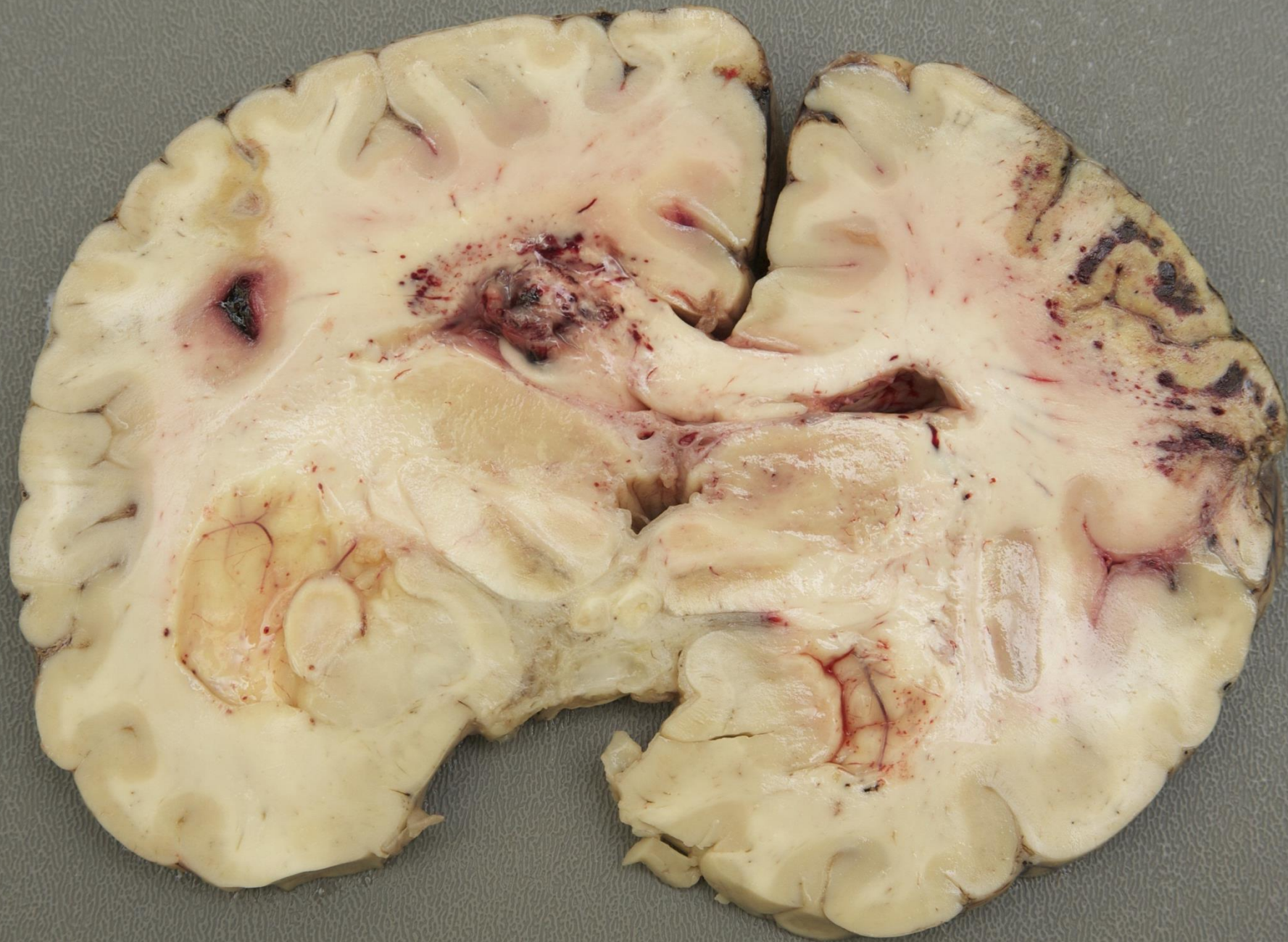
Obliterative microvascular disease with small necrotic foci within brain parenchyma.

Tissue was sent for universal polymerase chain reaction analysis for bacteria, fungi, and mycobacteria (nontuberculous and tuberculosis); all testing was negative.

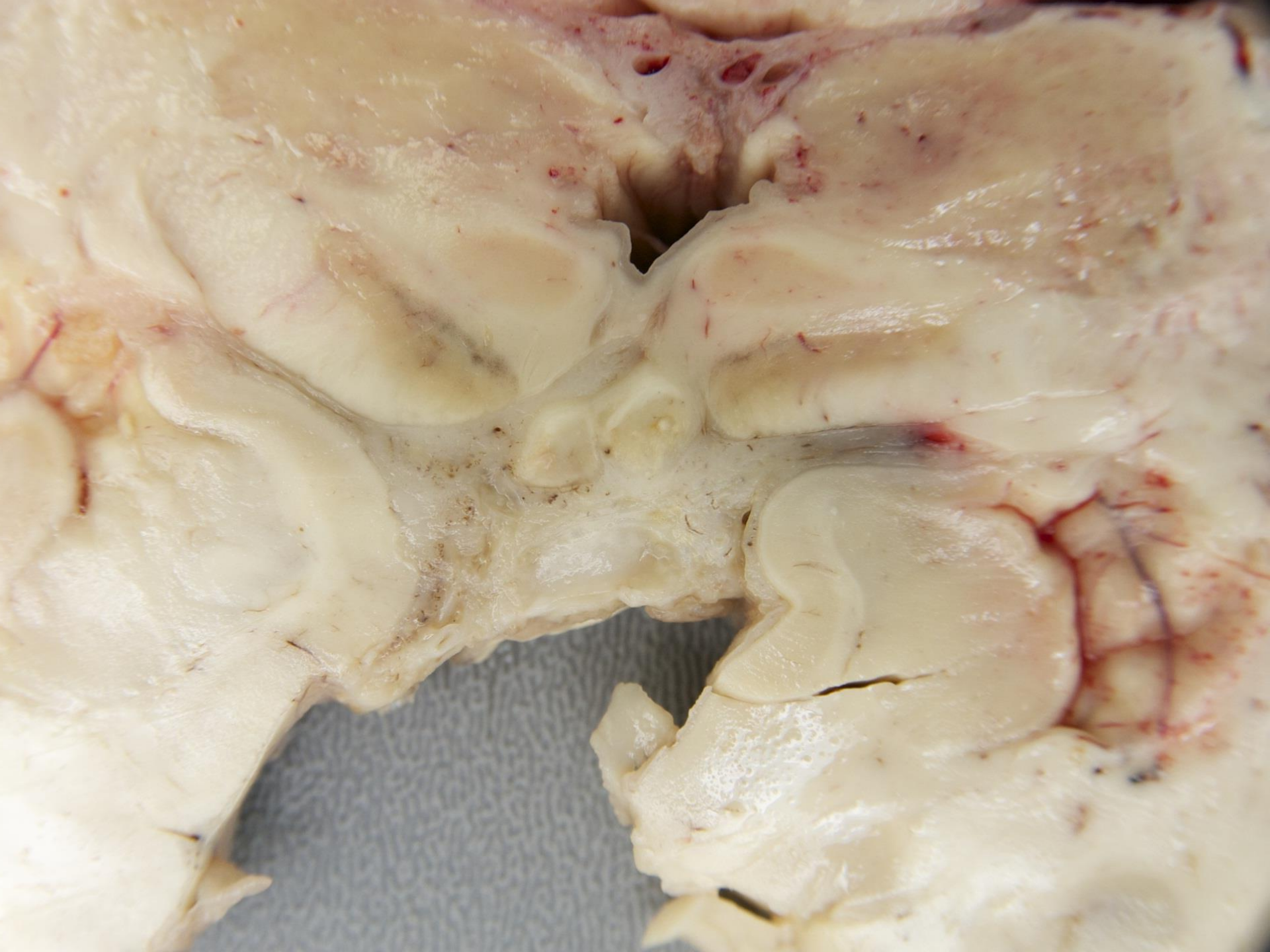


# **Autopsy**

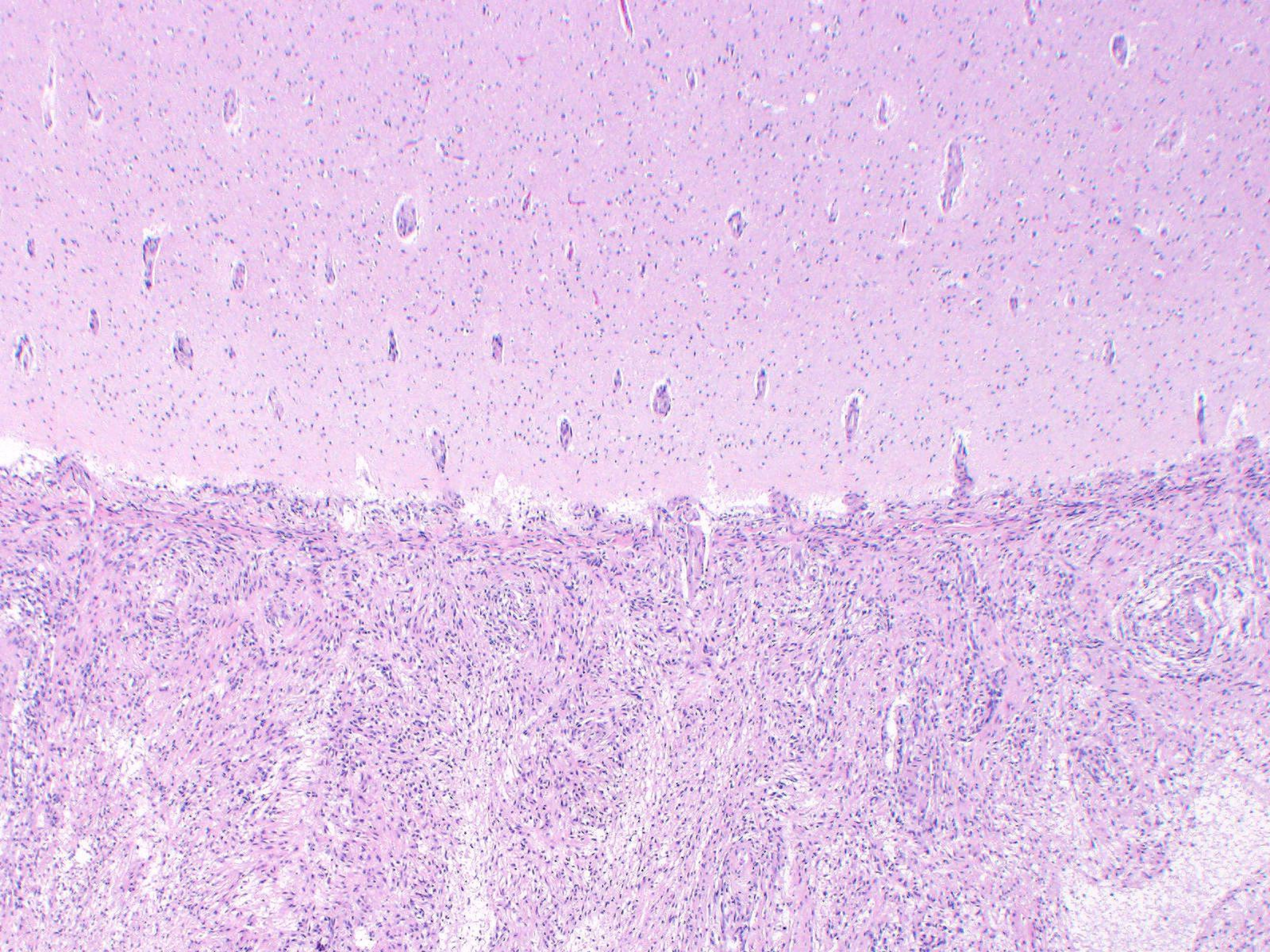
**3/3/2014**



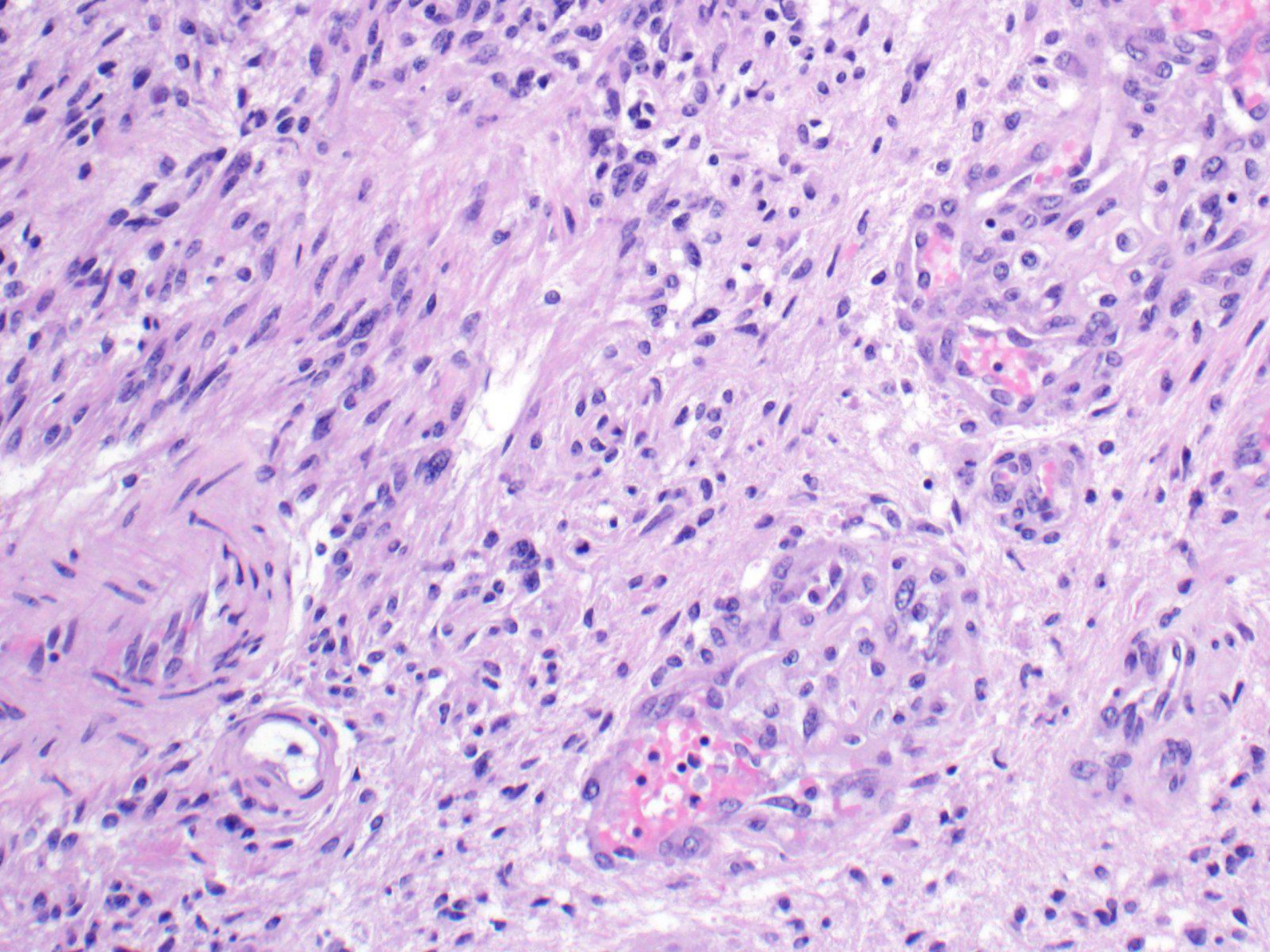










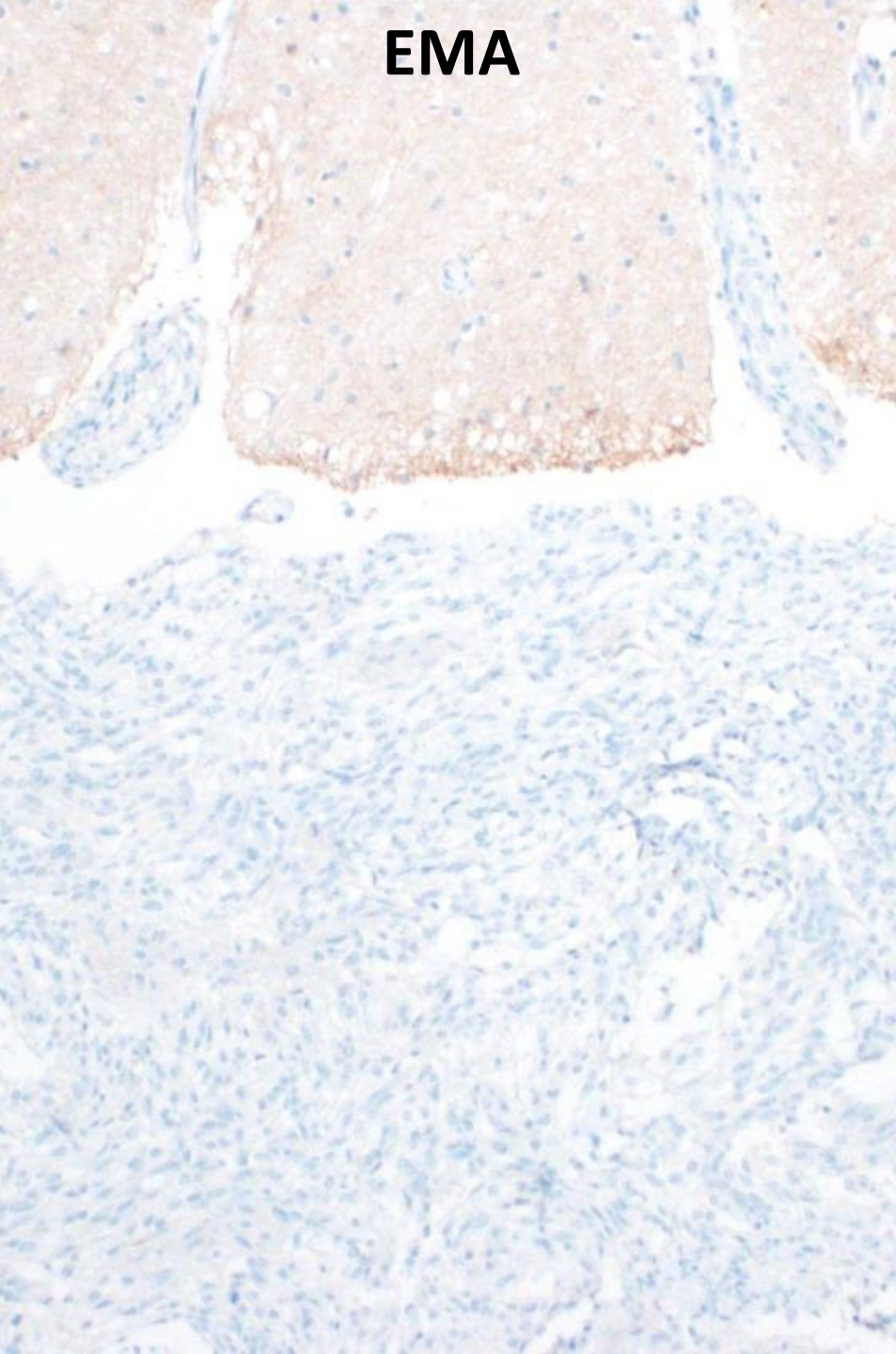




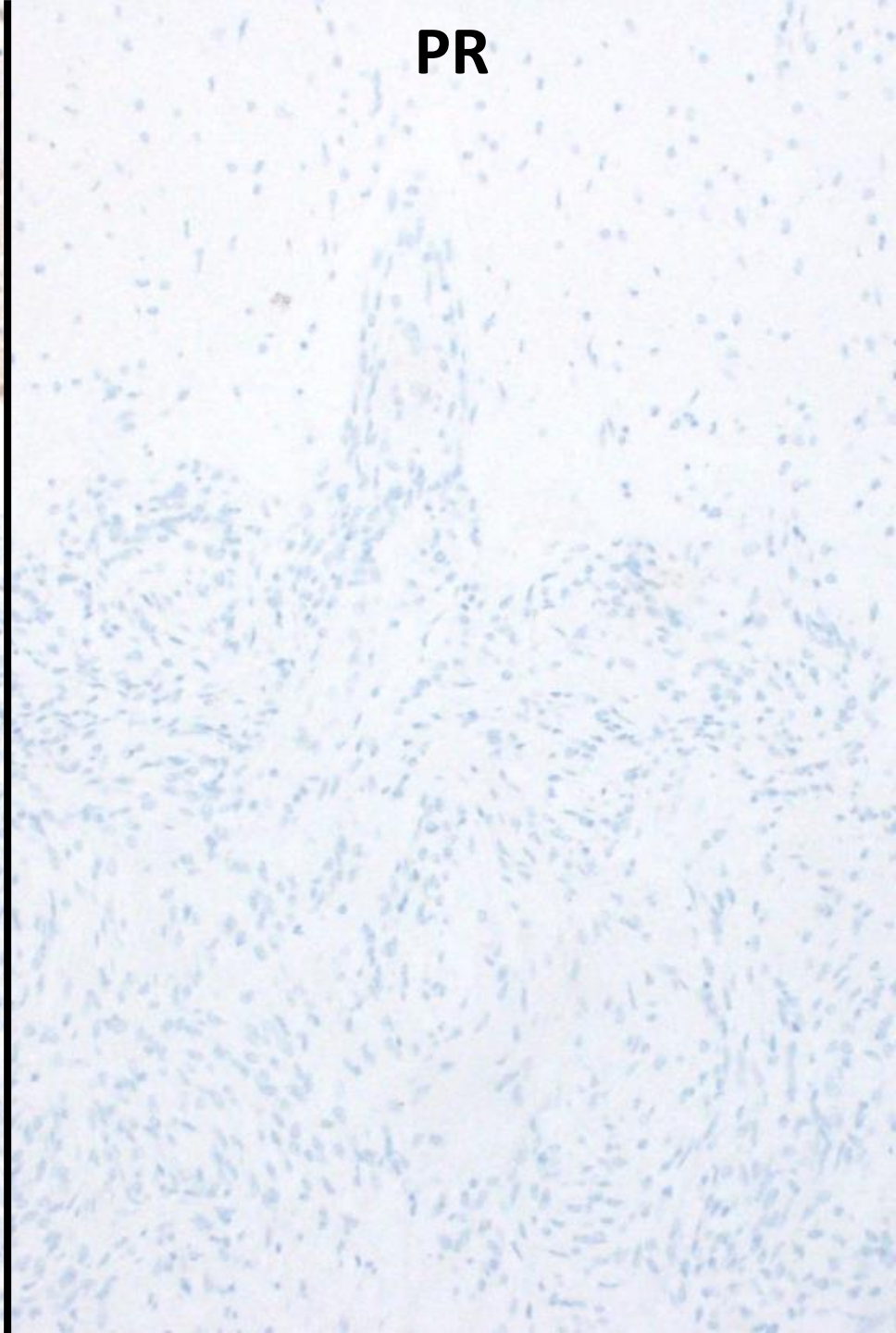
# Our Differential Diagnosis

- Meningioangiomatosis
- Rheumatoid meningitis
- Sarcoidosis
- IgG4-related meningeal disease

**EMA**

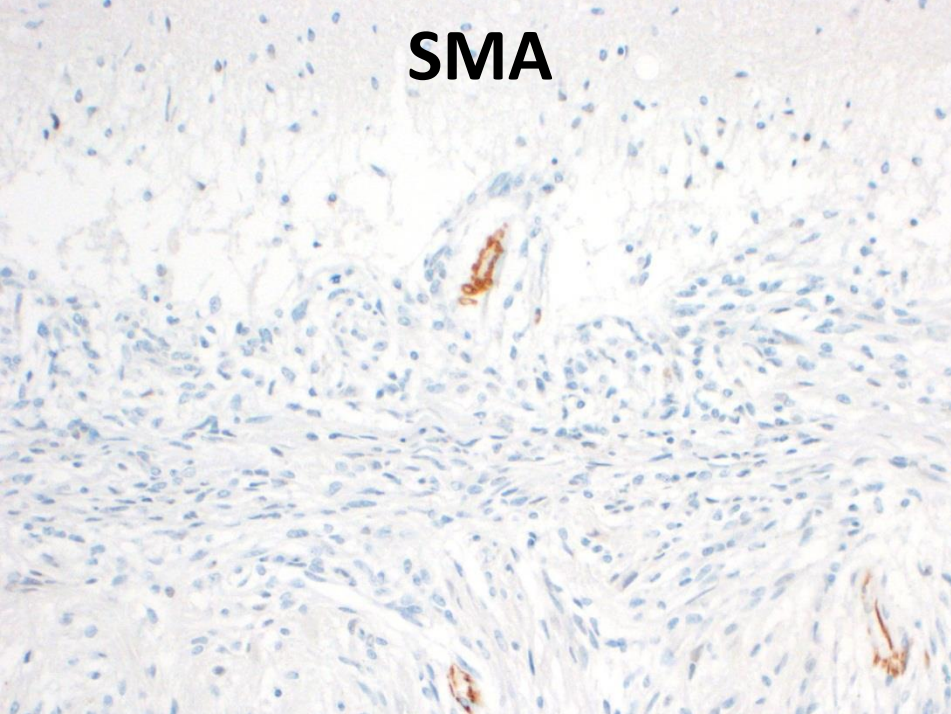


**PR**

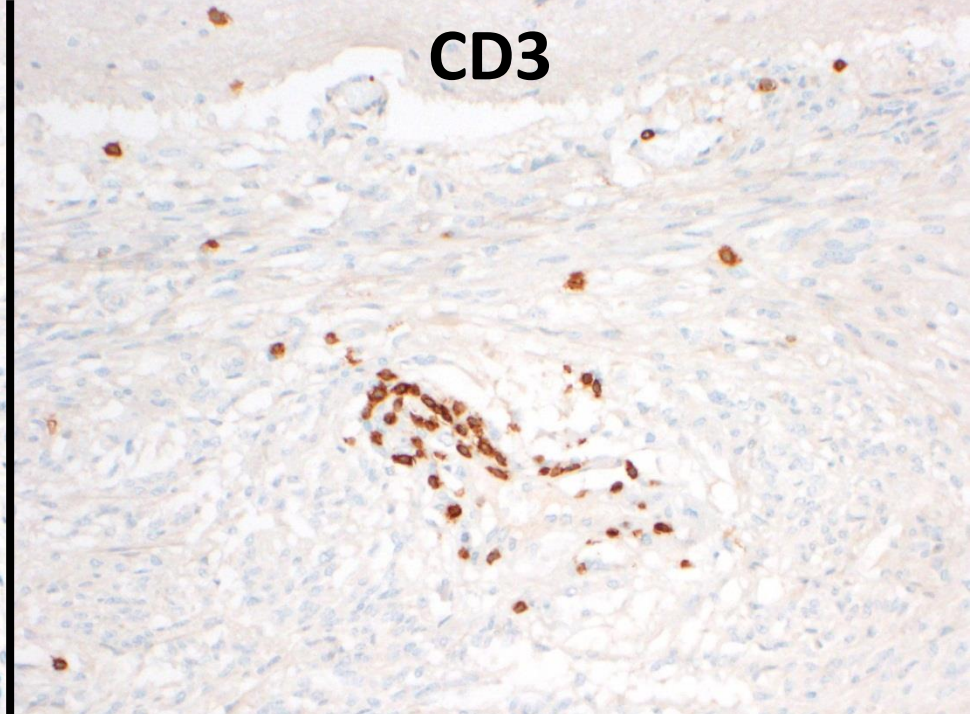




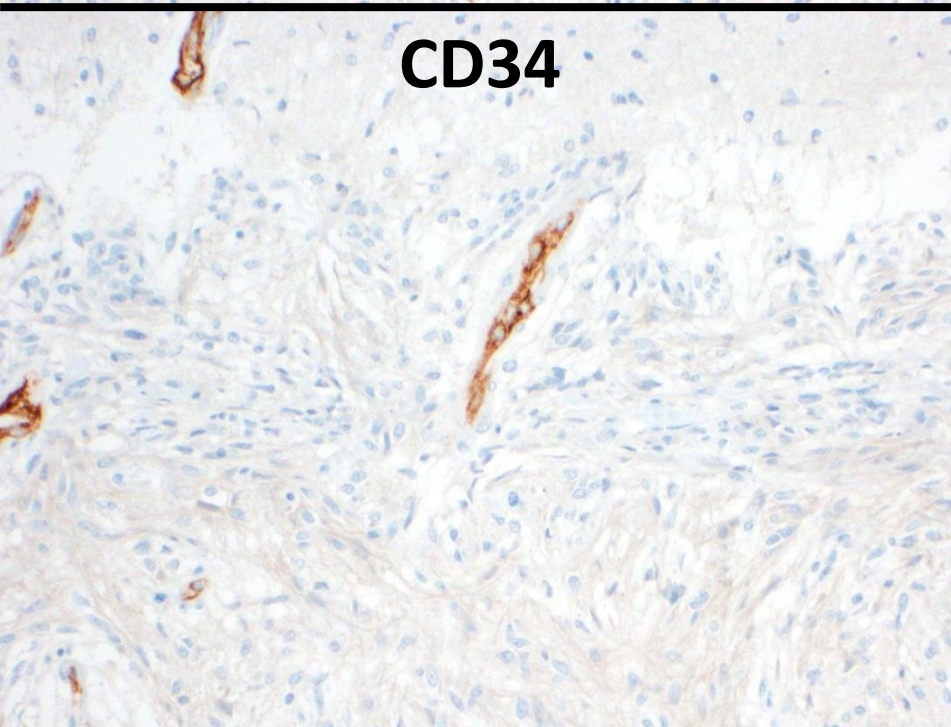
**SMA**



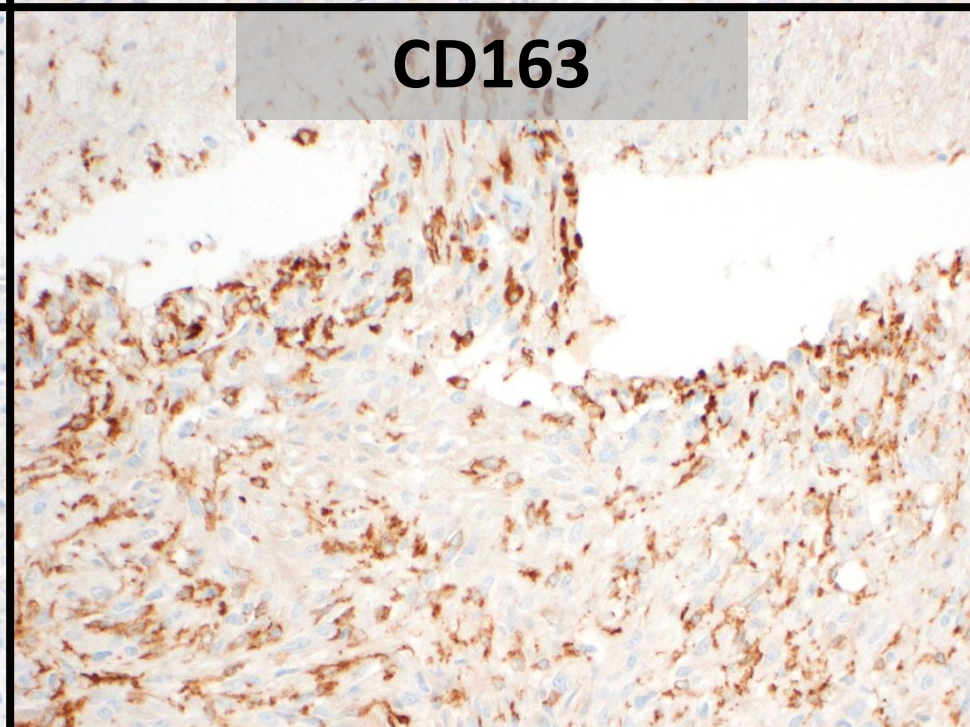
**CD3**



**CD34**

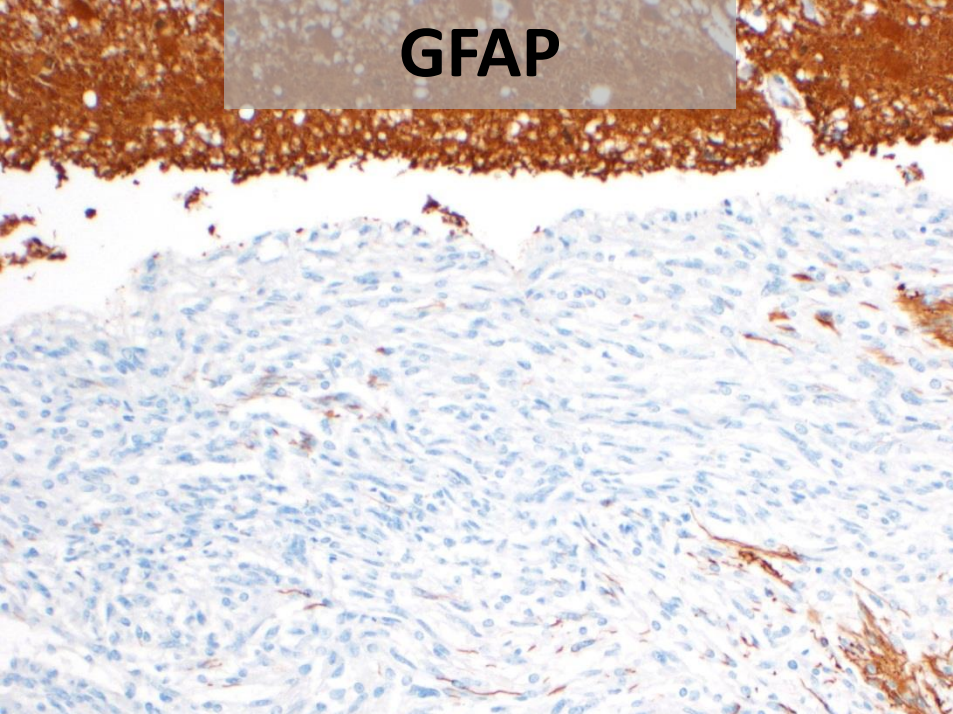


**CD163**

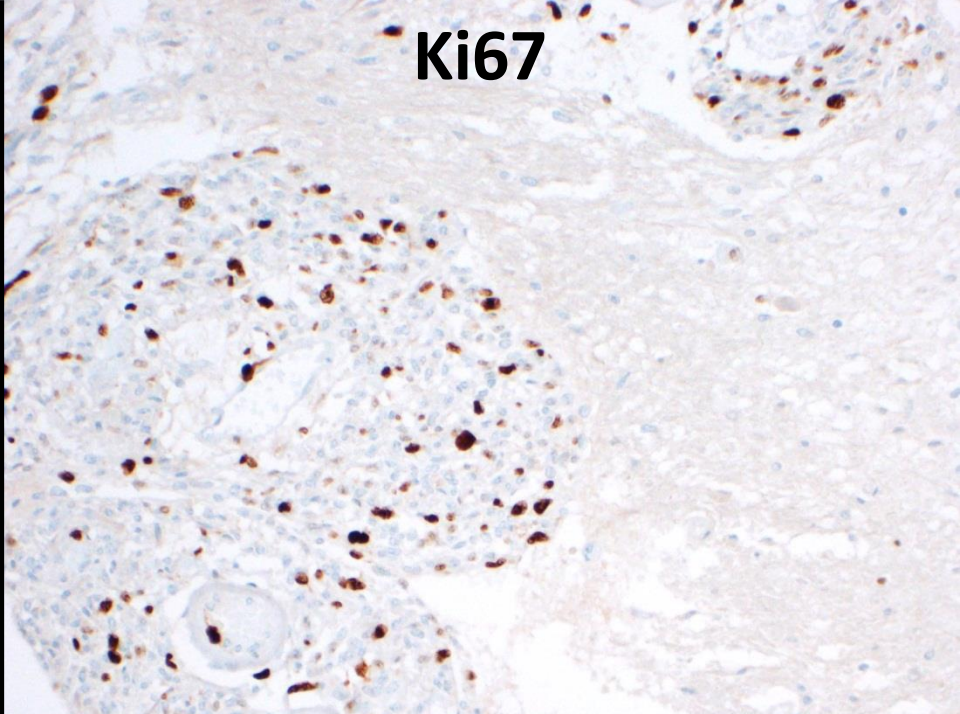




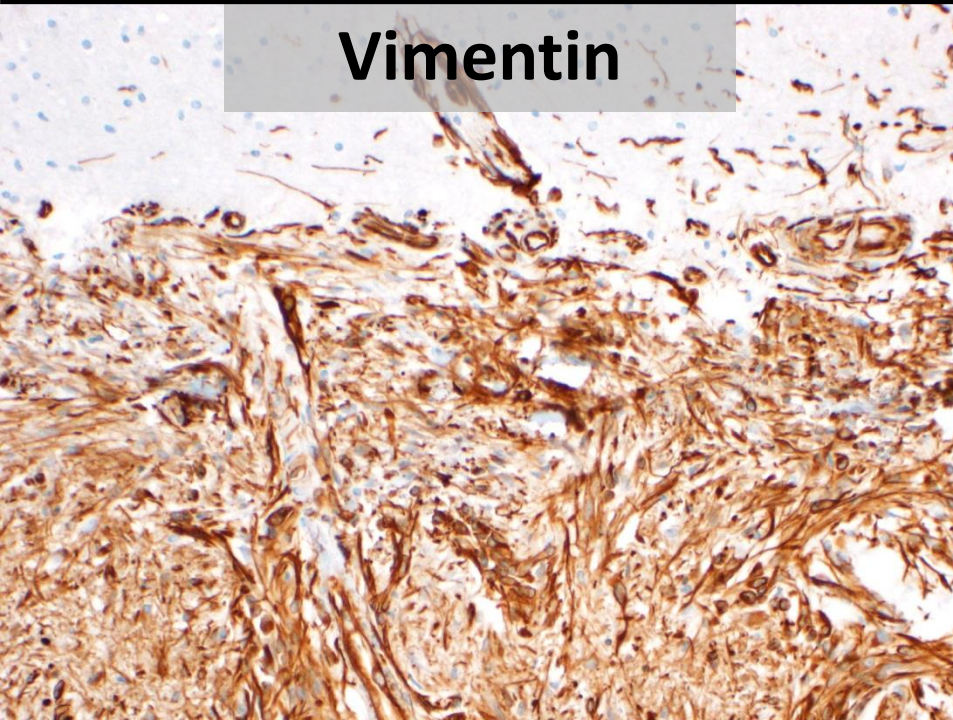
**GFAP**



**Ki67**



**Vimentin**



**Negative:**

CD20

CD1a

IgG, IgG4

S100

Desmin

Tyrosinase

Tissue Gram stain

GMS

Fite

Spirochete

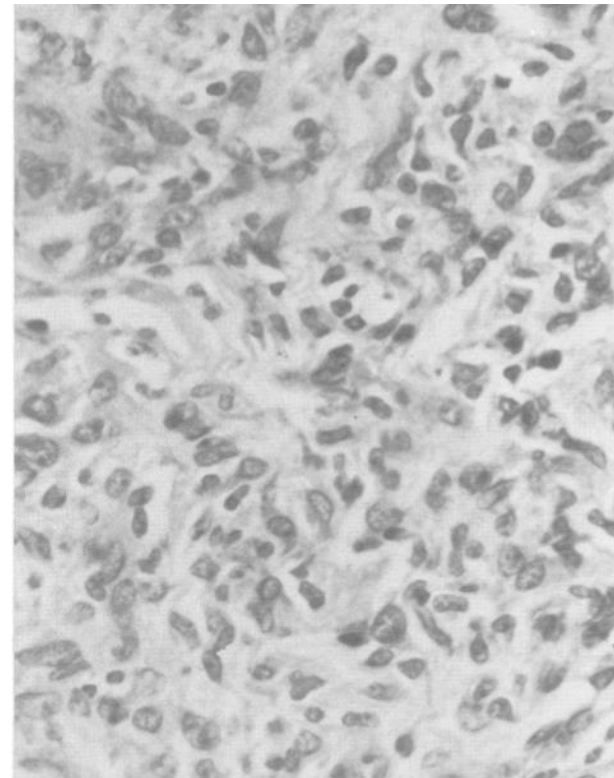
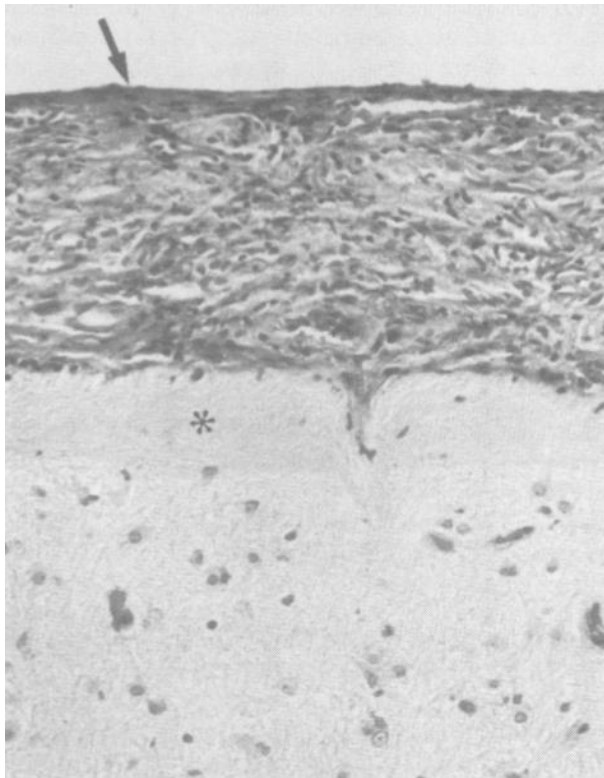


# Our Differential Diagnosis

- Meningioangiomatosis
- Rheumatoid meningitis
- Sarcoidosis
- IgG4-related meningeal disease
- **Progressive leptomeningeal fibrosis**

# Progressive Leptomeningeal Fibrosis

- Case report (Chalif D *et al.*, *JNN Psych*, 1983)  
41-year-old woman with 20 years of facial pain  
Presented with signs/symptoms of increased ICP  
Craniotomy: thick, fibrous leptomeninges





# Autopsy Diagnosis

The gross and histological findings are consistent with a **chronic-active proliferative process**. This process is felt to be **neither infectious nor neoplastic**.

The histologic pattern is **nonspecific**, but is suggestive of **progressive leptomeningeal fibrosis**.

# Summary

- Sampling
- We are open to suggestions!



# References

Chalif D, Duchen LW, Marshall J, Hayward R. Progressive leptomenigeal fibrosis: a clinico-pathological case report. *J Neurol Neurosurg Psychiatry*. 1983 Mar;46(3):261-5.

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