

# Diagnostic Slide Session

## Case 2016-9

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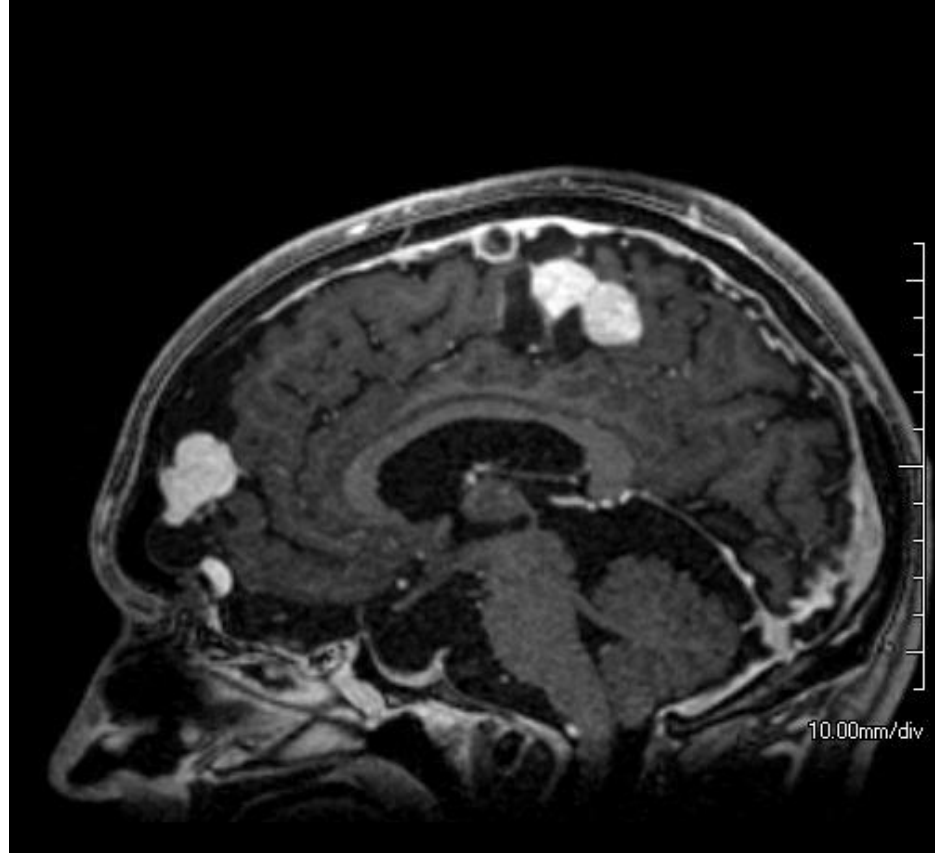
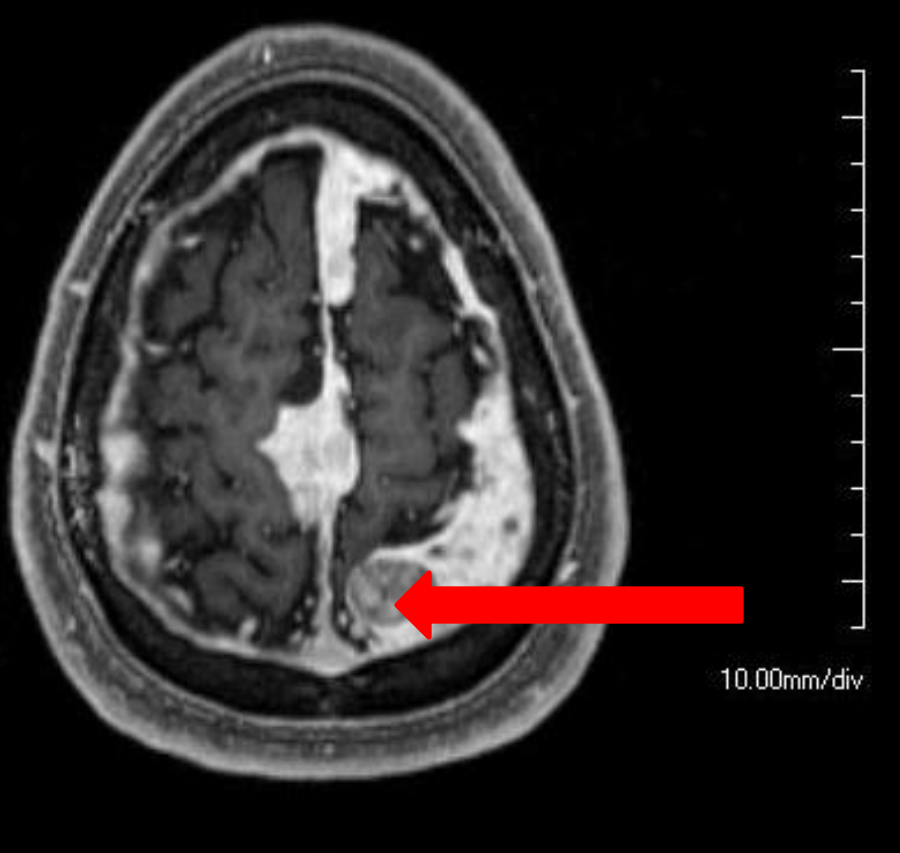
# Disclosures

I have no disclosures.

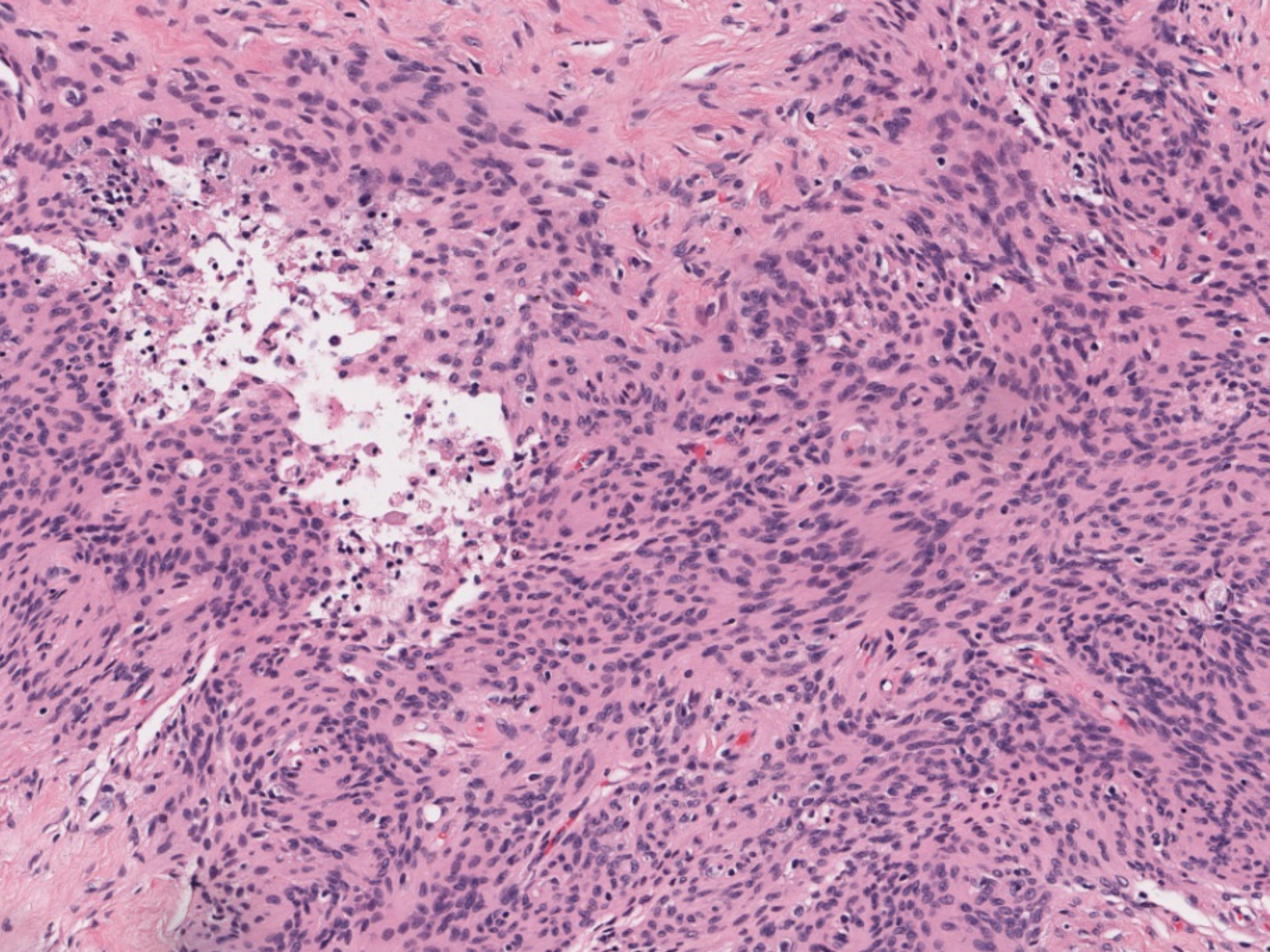
# Clinical history

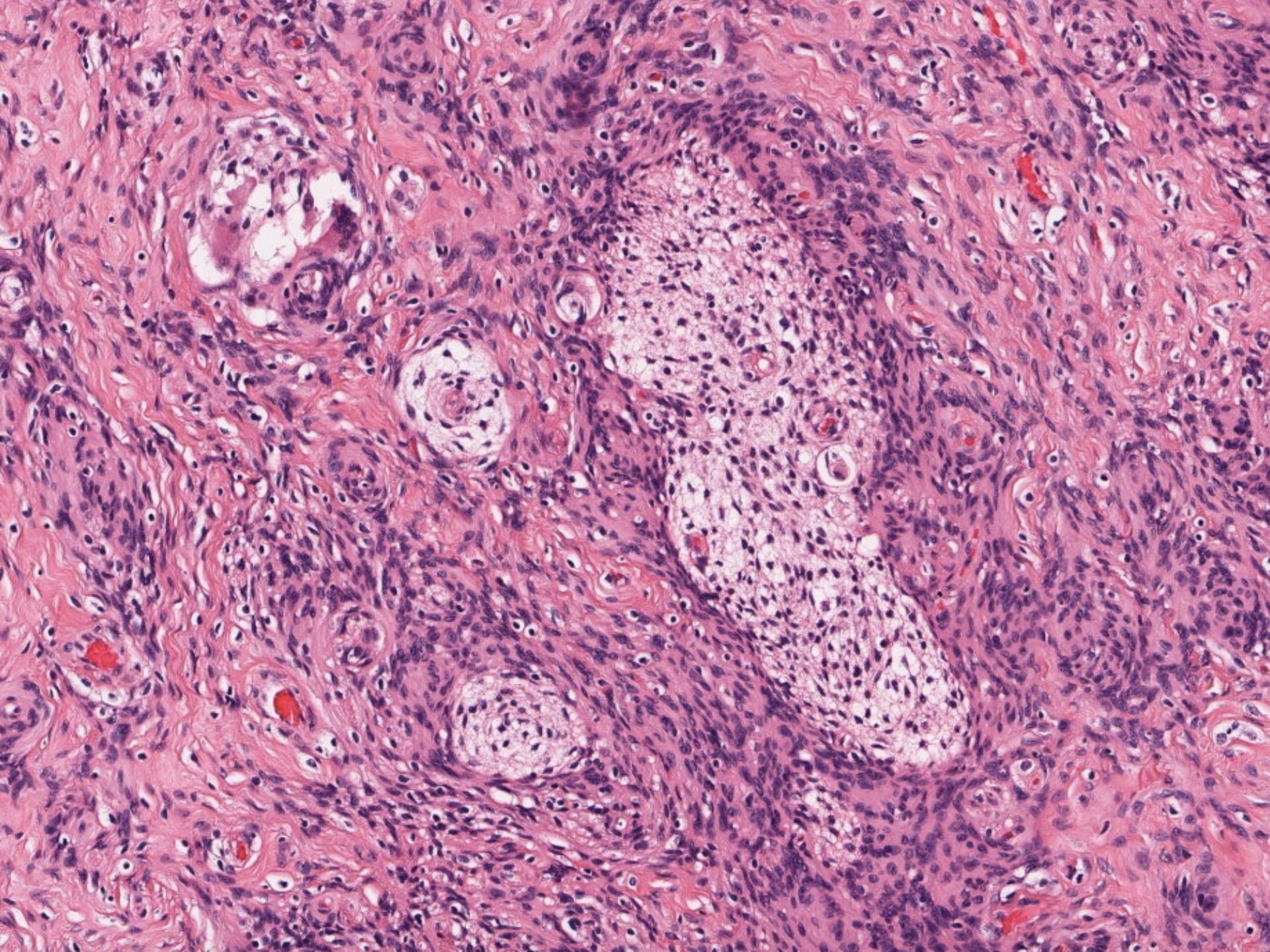
- 19-year-old male college student with a history of narcolepsy but healthy and active
- Woke up one Saturday morning with:
  - Headache
  - Clumsiness of RUE
  - Forced, dysarthric speech w/ perseverative stereotyped phrases (e.g. "What's going on, I don't know") and limited fluency
  - Lip smacking and chewing automatisms
  - Holding both hands in front of him "as if he'd never seen them before"

# Imaging



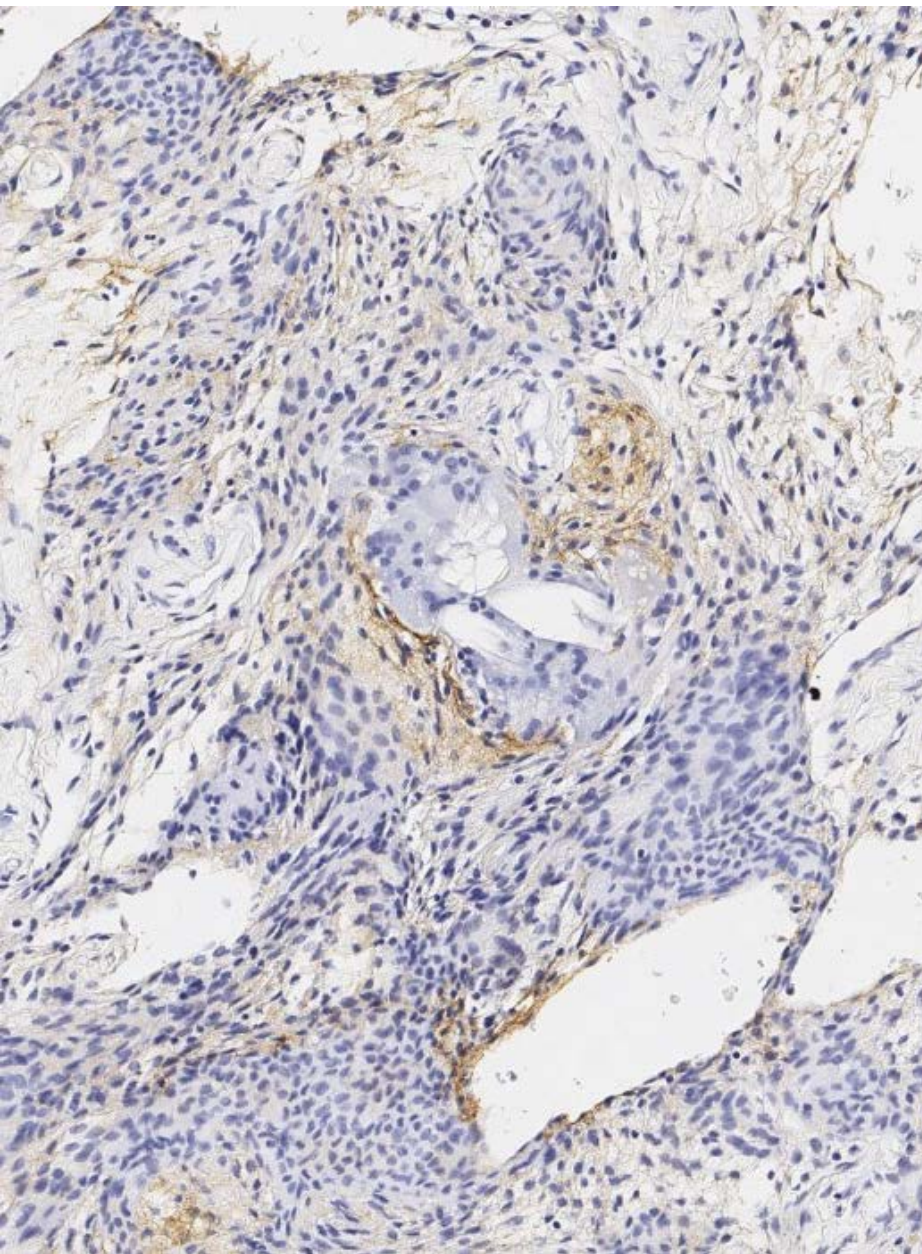
multiple heterogeneously enhancing dural-based masses and dural nodularity



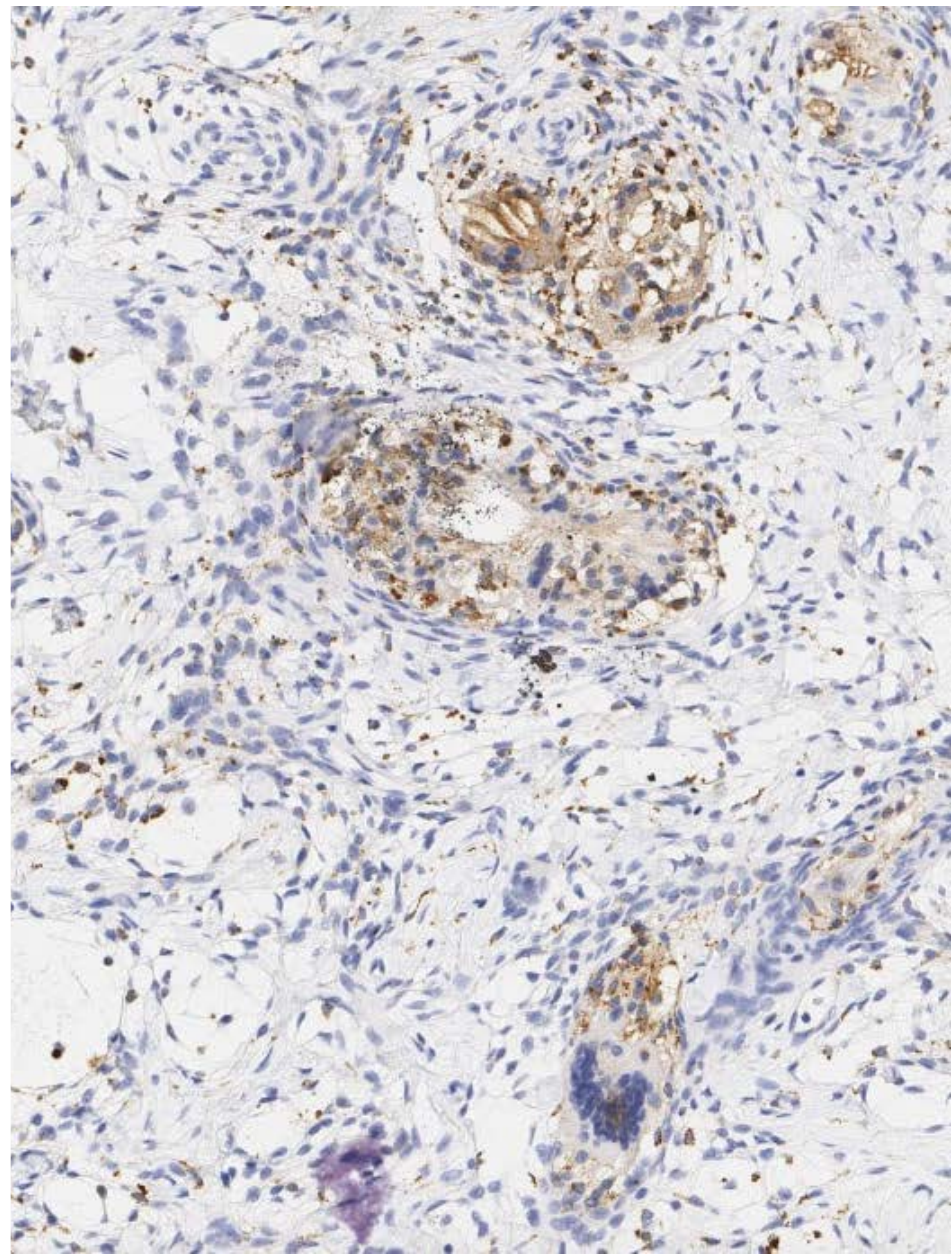


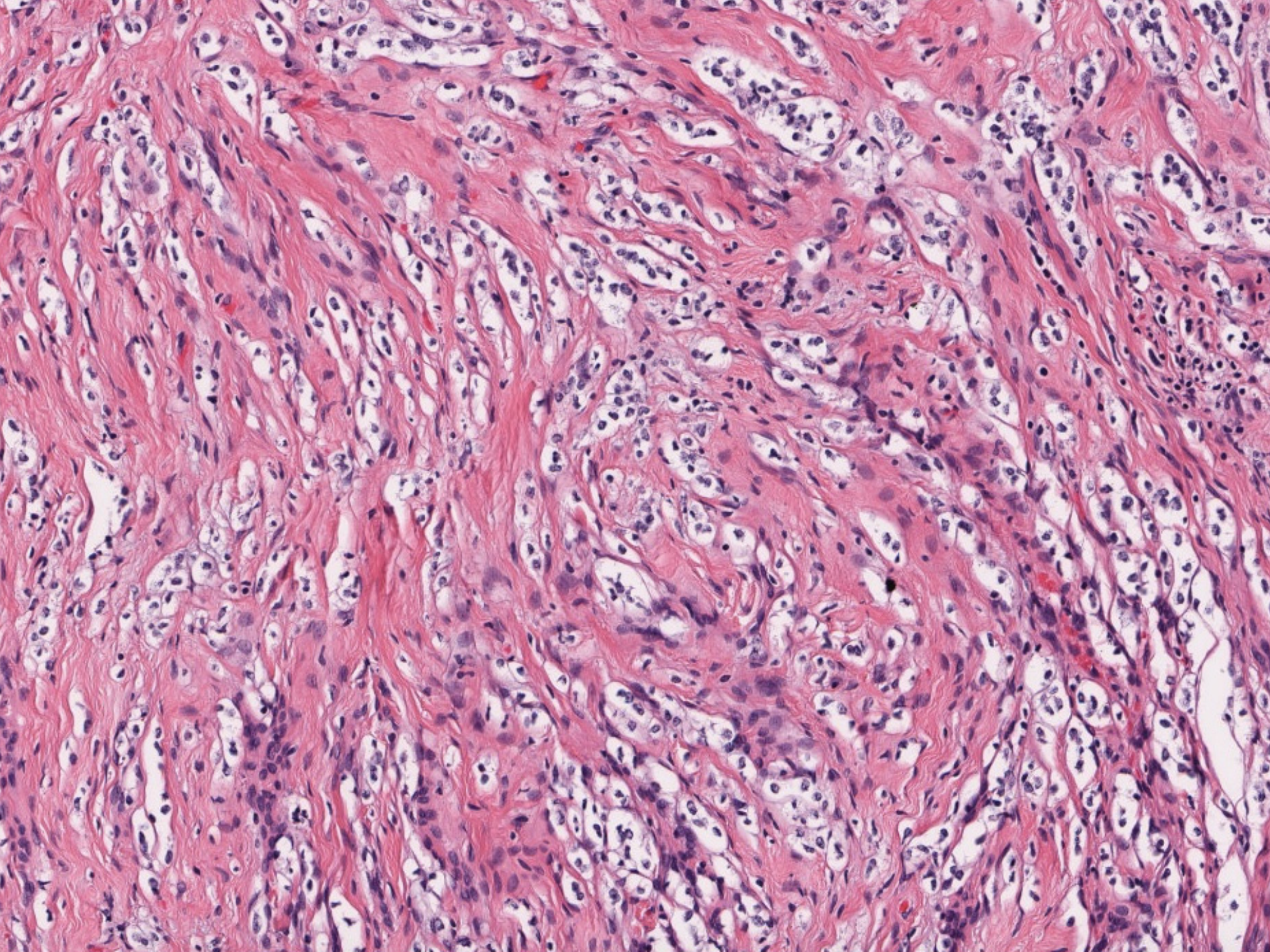
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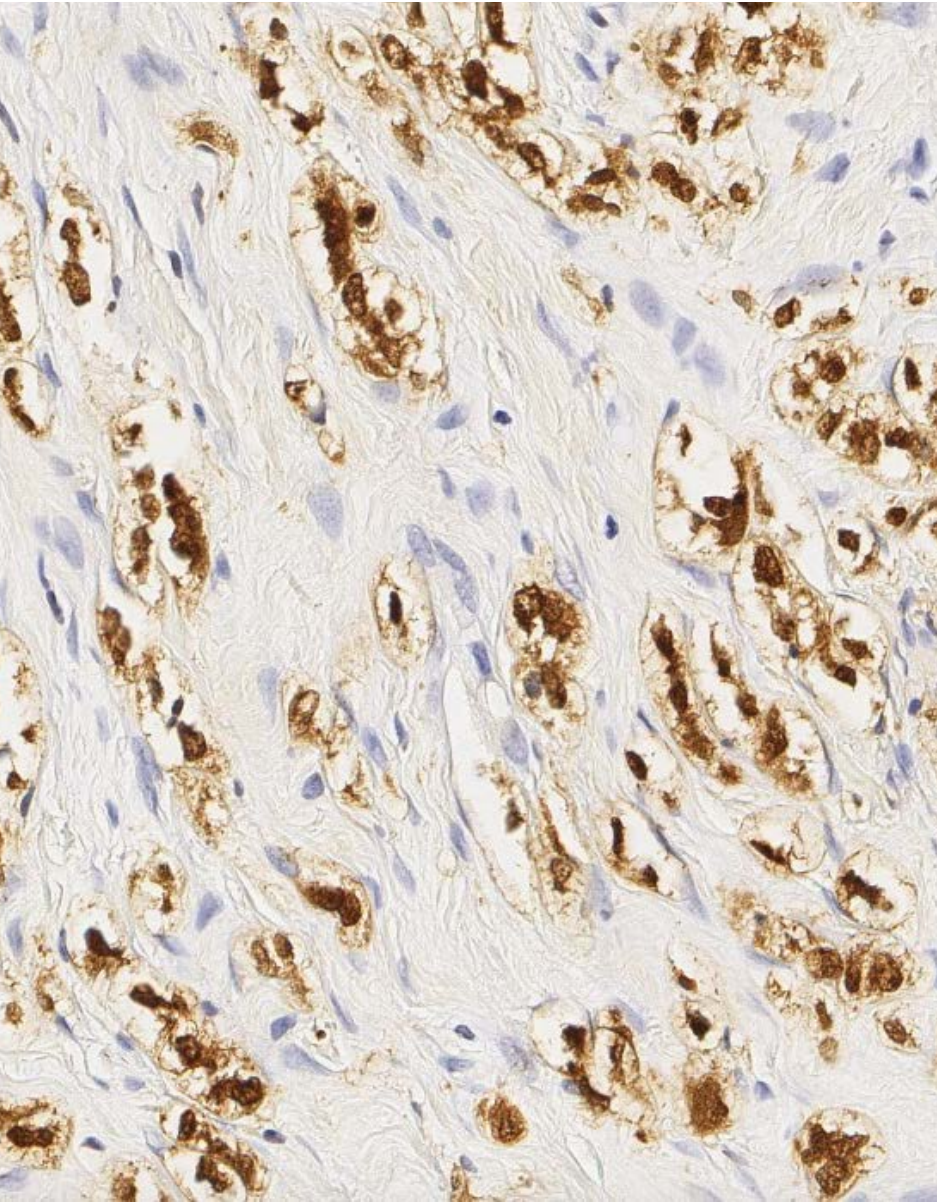


Diagnosis?

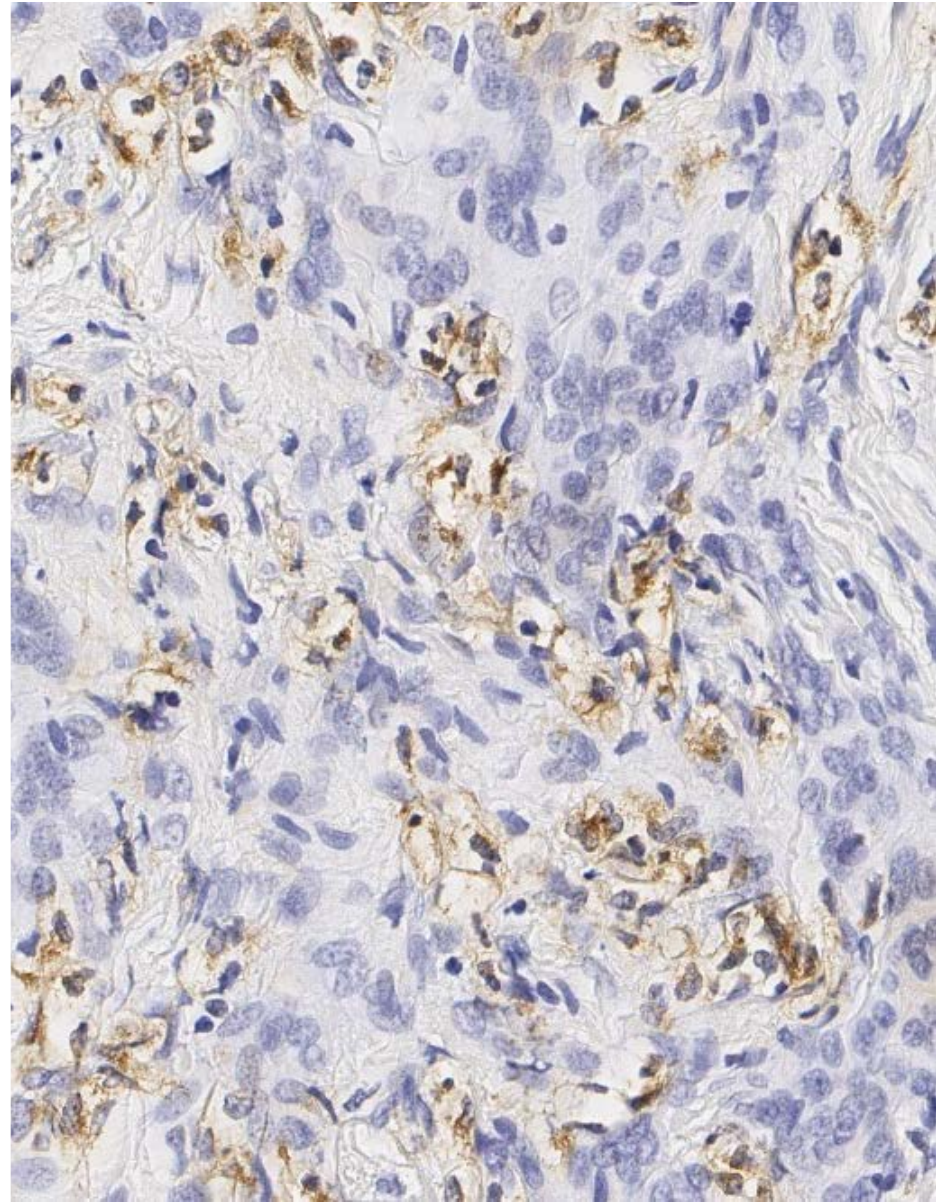
# Differential Diagnosis

- Neoplasm with reactive meningotheelial proliferation and granulomatous changes
  - Germ cell tumor
  - Melanoma
  - Histiocytic
  - Carcinoma
  - Sarcoma
  - Other
- Wegener's granulomatosis-like process
- Fibrotic Rosai-Dorfman disease
- Meningioma (meningiomatosis)
- Infection

S100

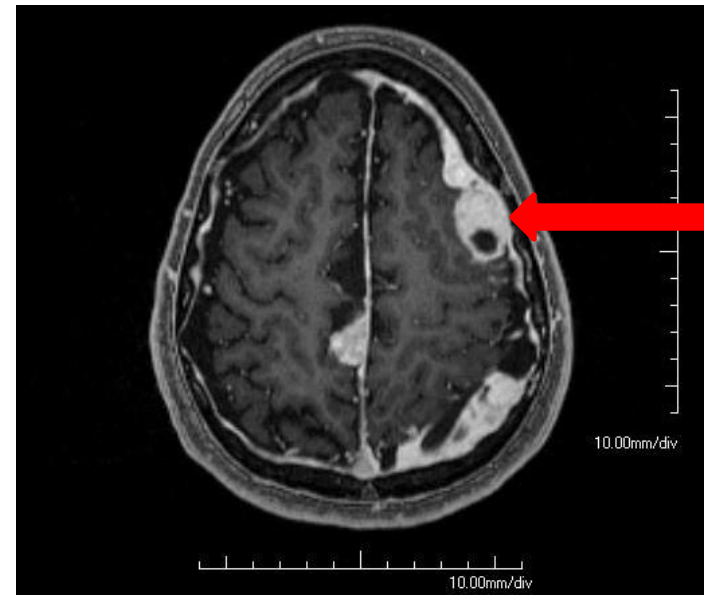


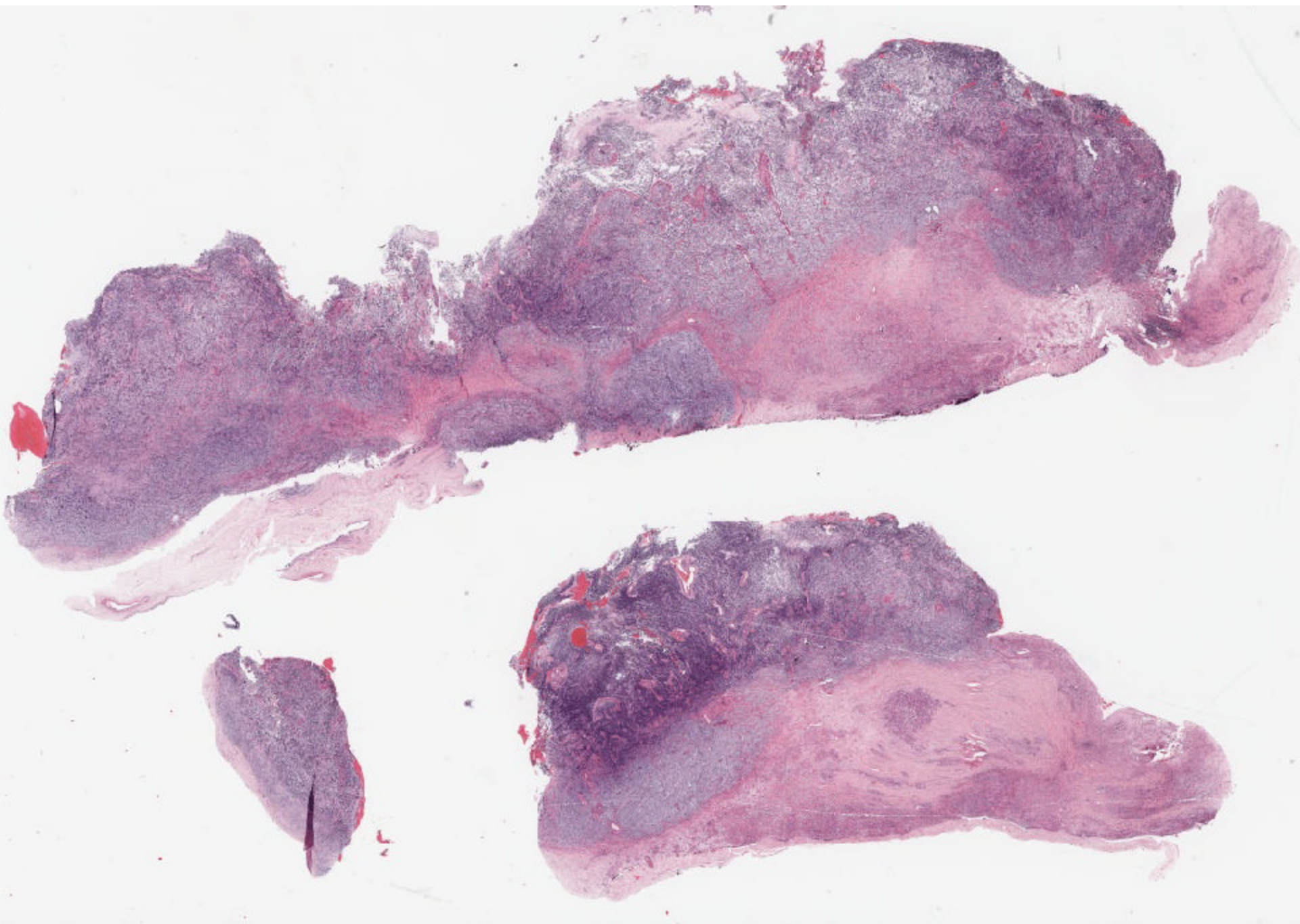
synaptophysin

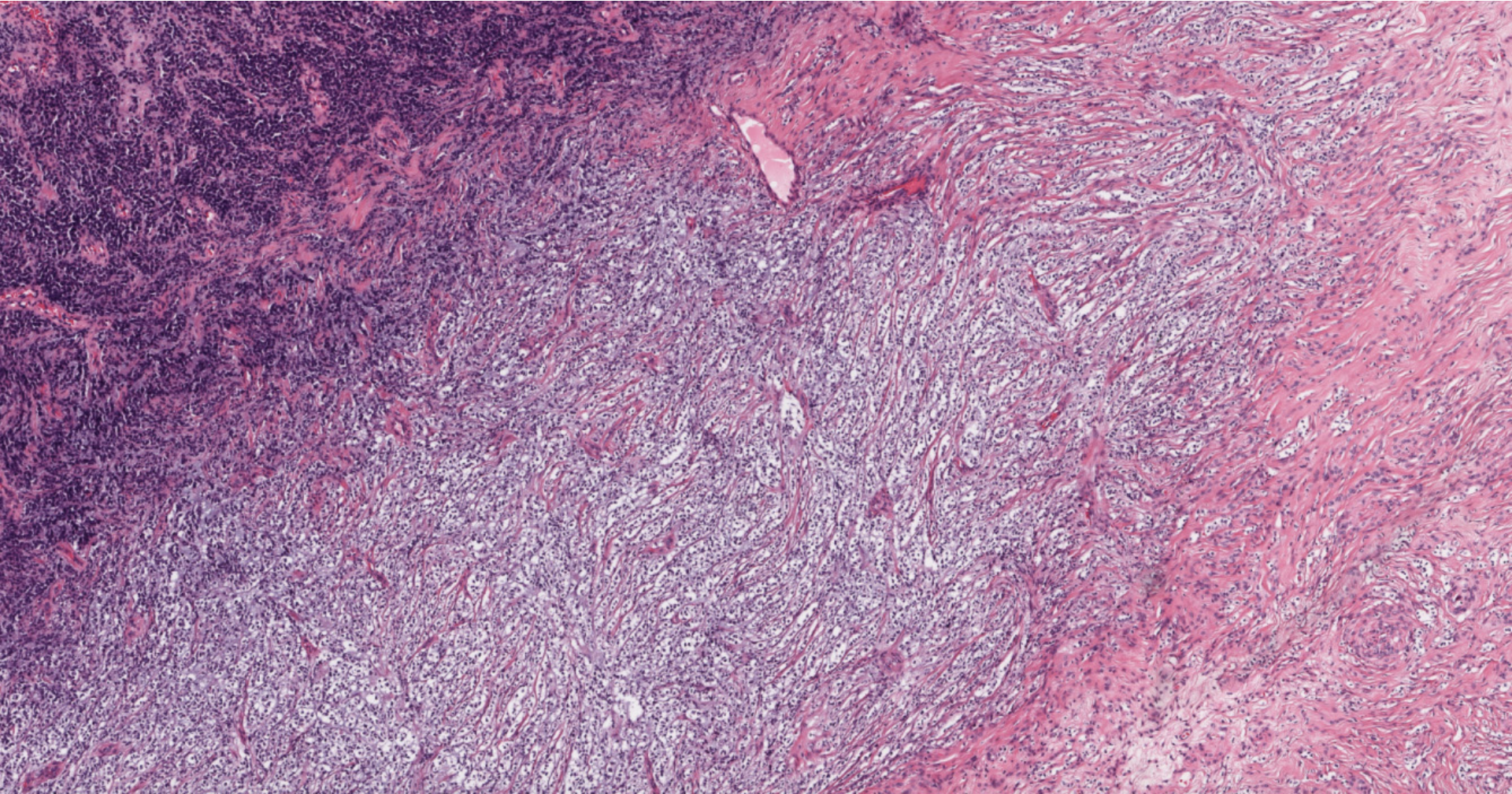


# History continued:

- Patient sent home with follow-up
- Case signed out descriptively
- Patient was readmitted for worsening headaches with light sensitivity, nausea, and vomiting.
- He also noted bilateral leg weakness.
- MRI performed and was unchanged.
- A second biopsy was performed.







## Positive

- S100
- Chromogranin
- Synaptophysin

## Ruled Out:

- Melanoma
- Carcinoma
- Sarcoma
- Meningiomatosis
- Histiocytic neoplasm

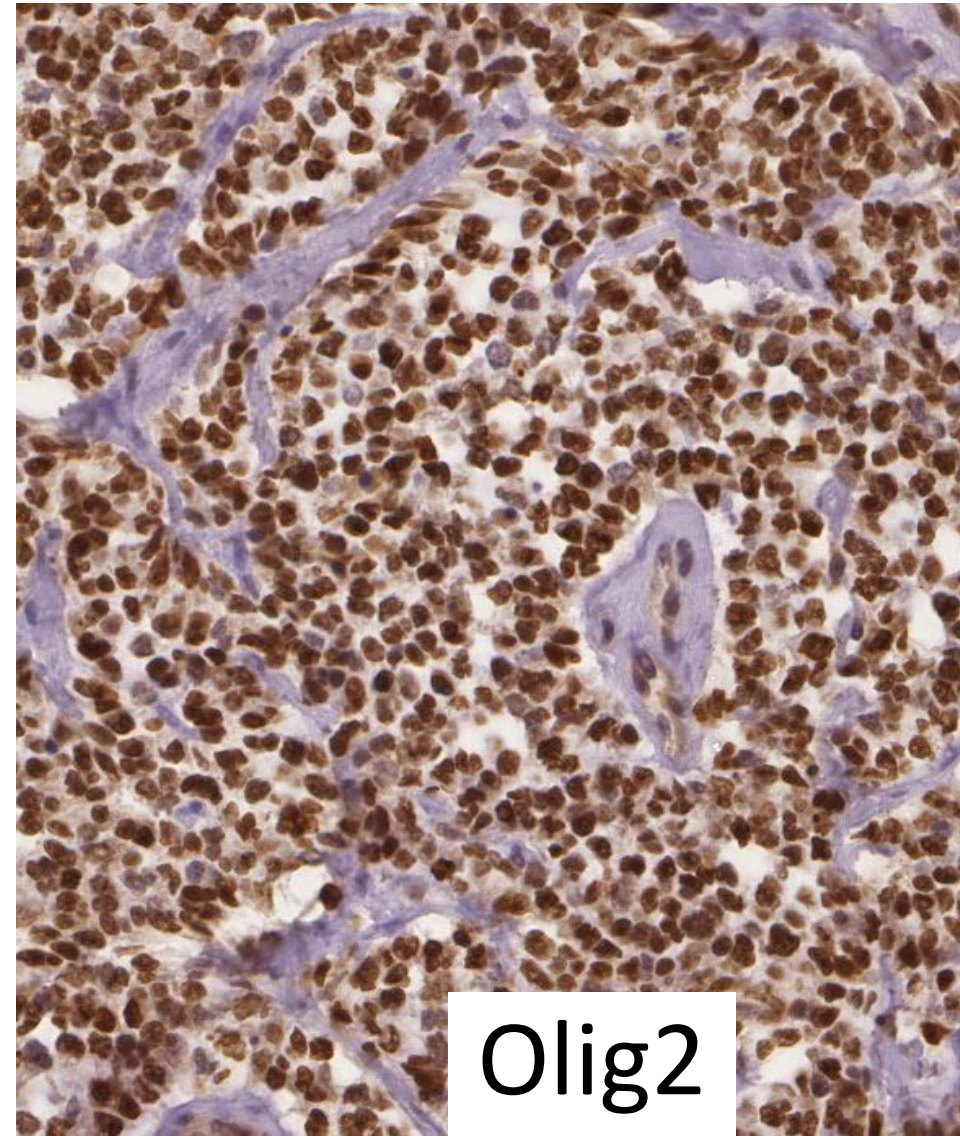
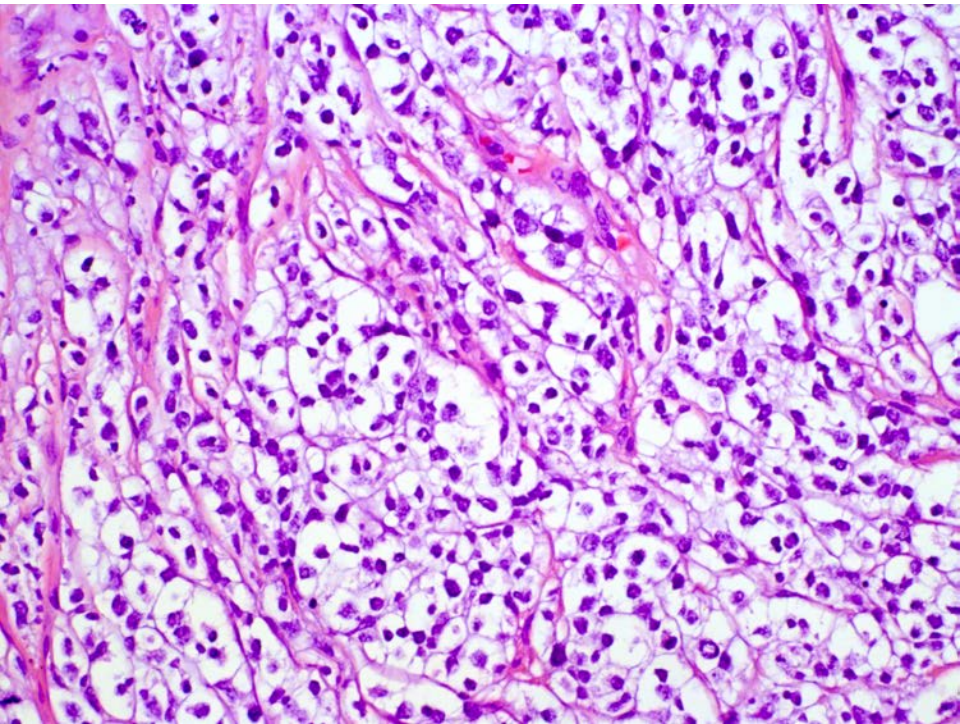
## Negative

# IHC

- MelanA, MITF1, tyrosinase, HMB45
- EMA and PR
- GFAP
- Mutant IDH1 (R132H)
- Cam5.2, panCK, AE1/3
- TFE3
- Desmin, myogenin, SMA
- Mic2, ERG
- INI1 retained
- WT1
- Neurofilaments
- PLAP, c-kit, OCT3/4

# Additional results

Co-Deletion of 1p and 19q,  
with Polysomy Detected in  
greater than 30 percent of  
Tumor Cells (ABNORMAL)



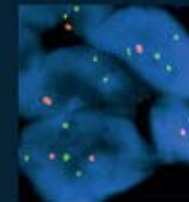
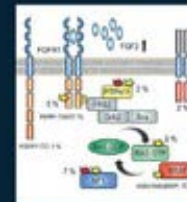
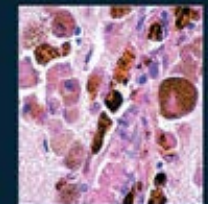
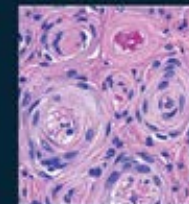


# Diagnosis

- Disseminated leptomeningeal glioneuronal tumor
  - With anaplastic transformation

## WHO Classification of Tumours of the Central Nervous System

David N. Louis, Hiroko Ohgaki, Otmar D. Wiestler, Webster K. Cavenee, David W. Ellison, Dominique Figarella-Branger, Arie Perry, Guido Reifenberger, Andreas von Deimling



WHO

### Original Paper

Acta Neuropathologica

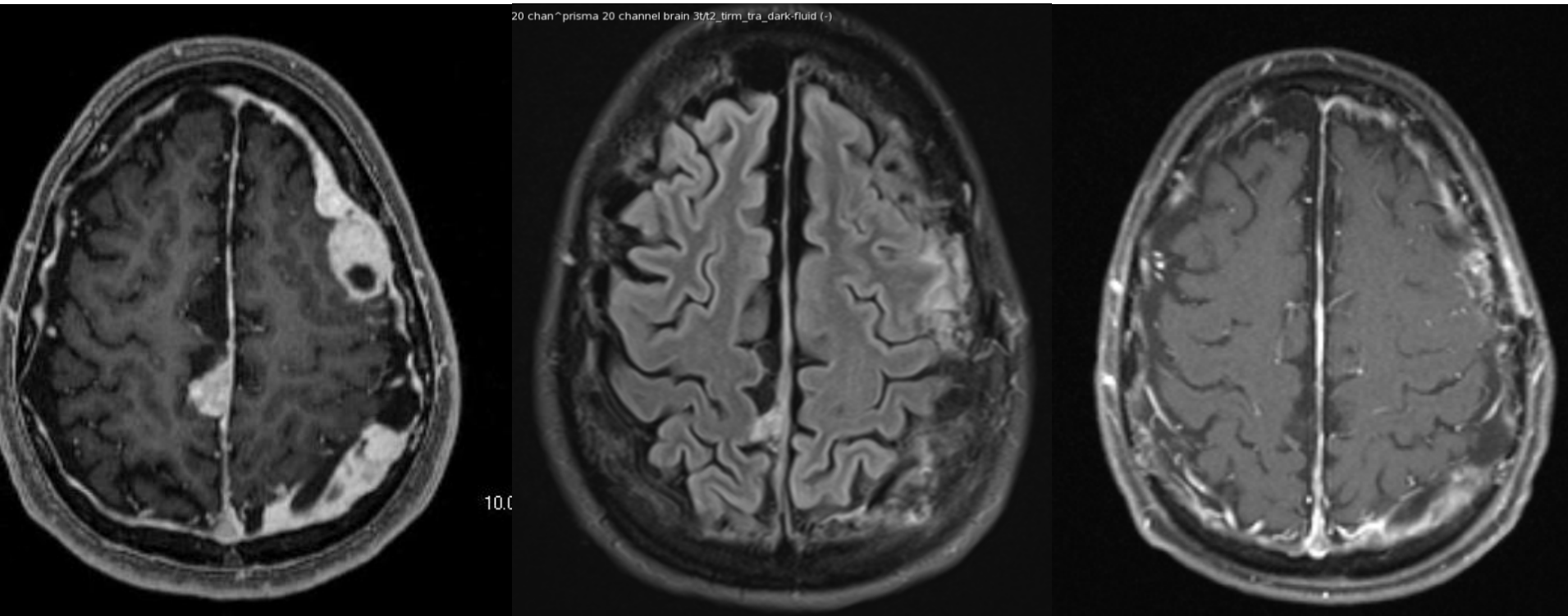
November 2012, Volume 124, Issue 5, pp 627-641

First online: 01 September 2012

# Disseminated oligodendroglial-like leptomeningeal tumor of childhood: a distinctive clinicopathologic entity

Fausto J. Rodriguez , Arie Perry, Marc K. Rosenblum, Sherry Krawitz, Kenneth J. Cohen, Doris Lin, Stacy Mosier, Ming-Tseh Lin, Charles G. Eberhart, Peter C. Burger [Show less](#)

# How is our patient?



Initial presentation

8/10/2015 (post 7 weeks  
of temozolamide)

4/17/2016

# Thanks

- Maria Martinez-Lage
- Mariarita Santi-Vicini
- Ilya Nasrallah