58th Annual Diagnostic Slide Session

Case 1

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- No disclosures or conflicts of interest

Clinical History

HPI:

- 51 y/o female
- New onset seizures and progressive confusion x two months
- Frequent headache and blurry vision
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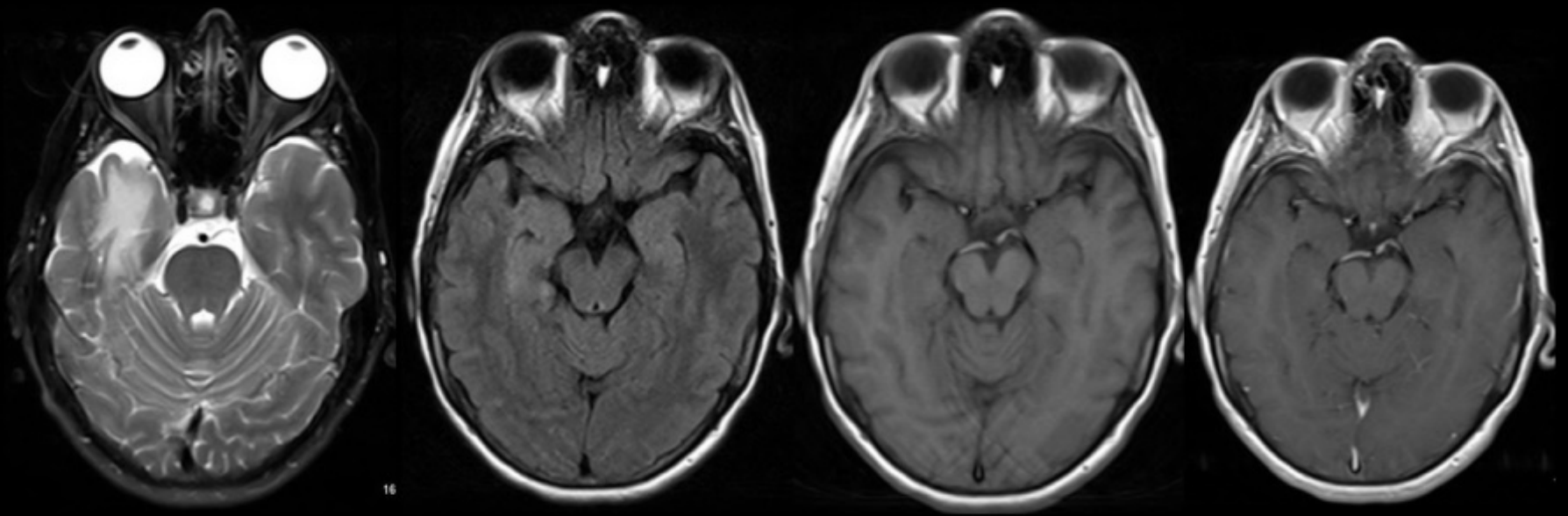
PMH:

- Restless leg syndrome
- Depression
- Anxiety
- CAD, HTN and hyperlipidemia

Neurological exam:

- No focal deficits

Pre-operative MRI



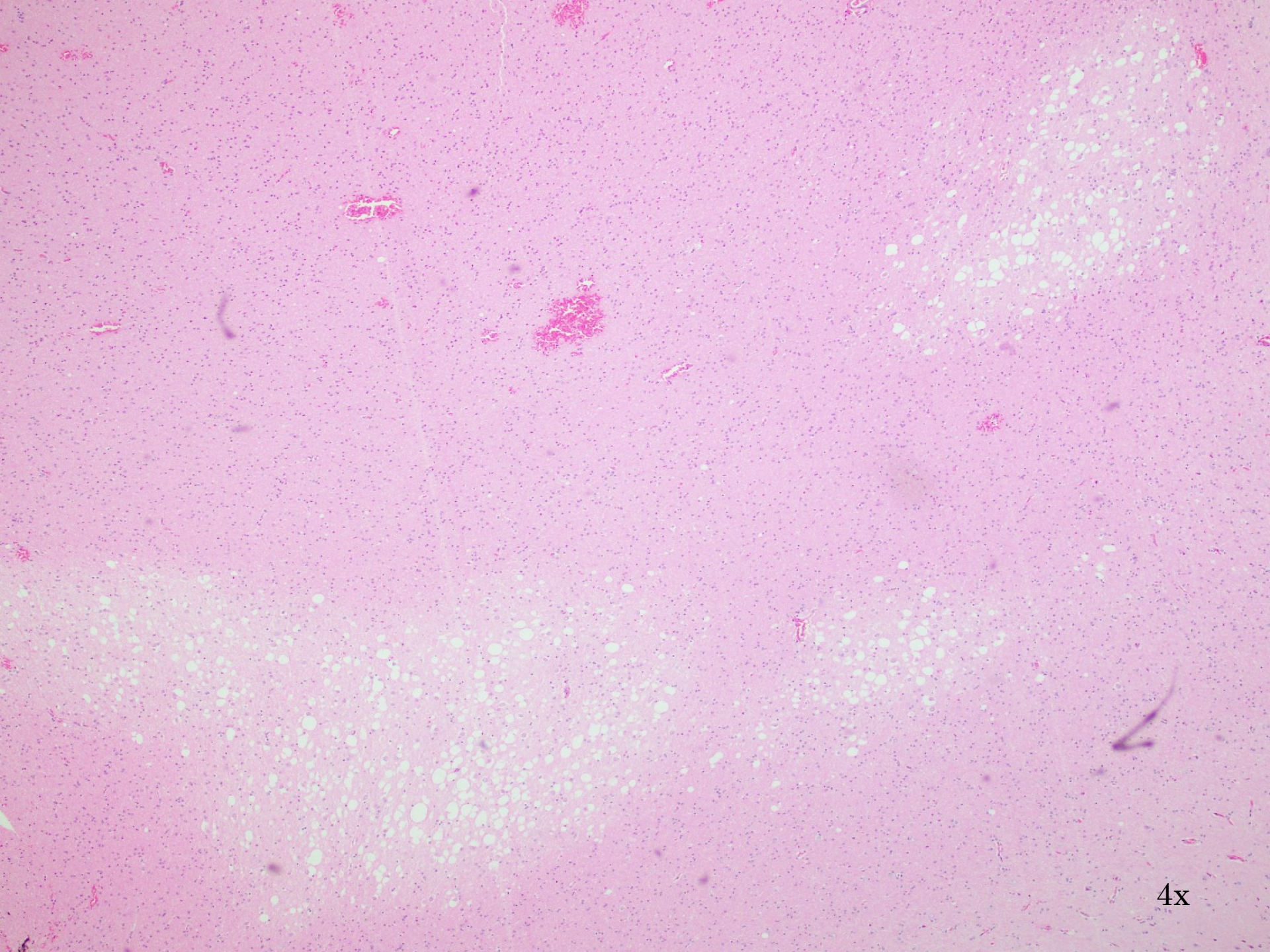
T2 FLAIR T1 T1 GAD

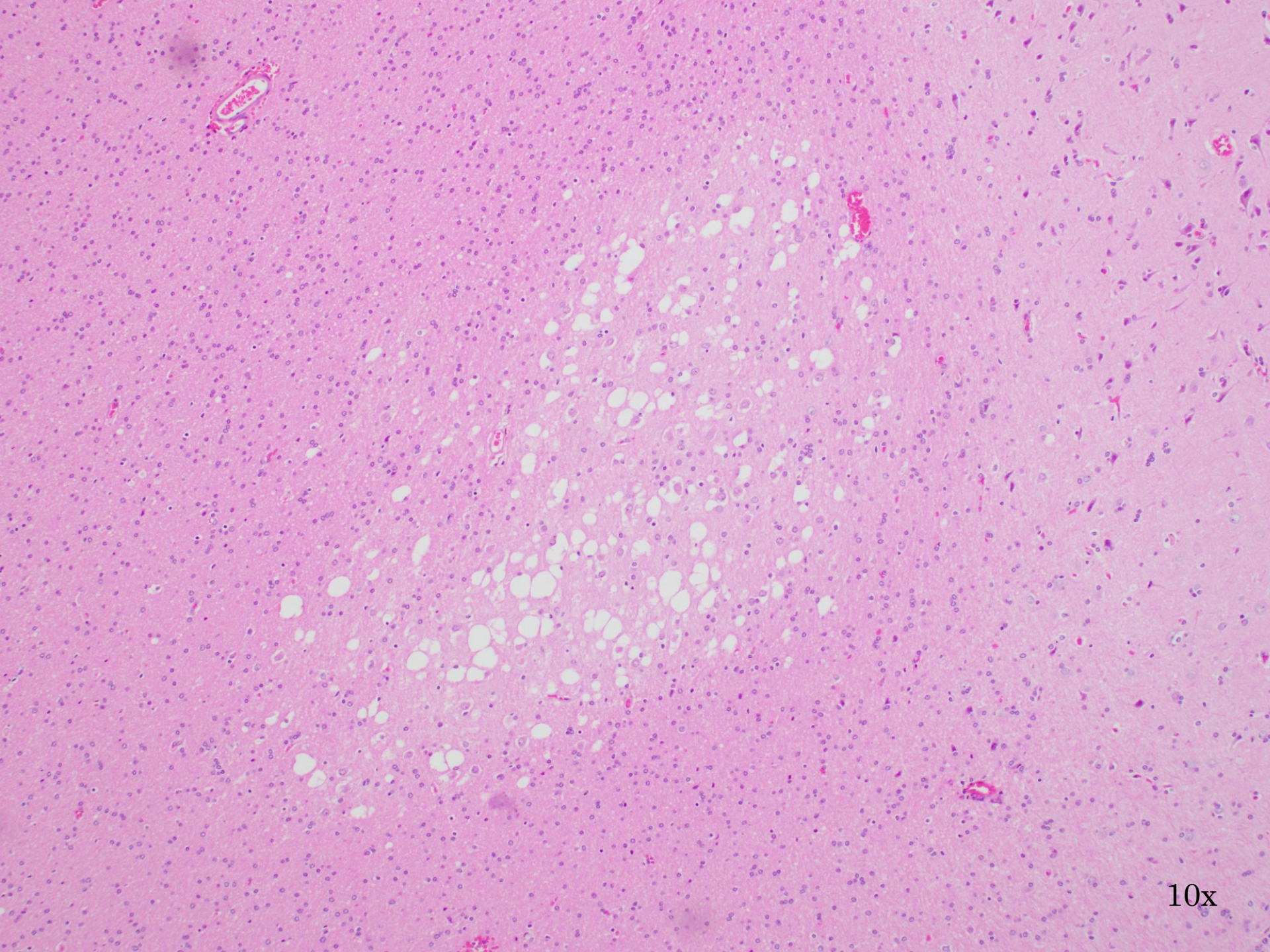
Hyperintense infilatrating lesion involving the right mesial temporal lobe. No gadolinium enhancement.

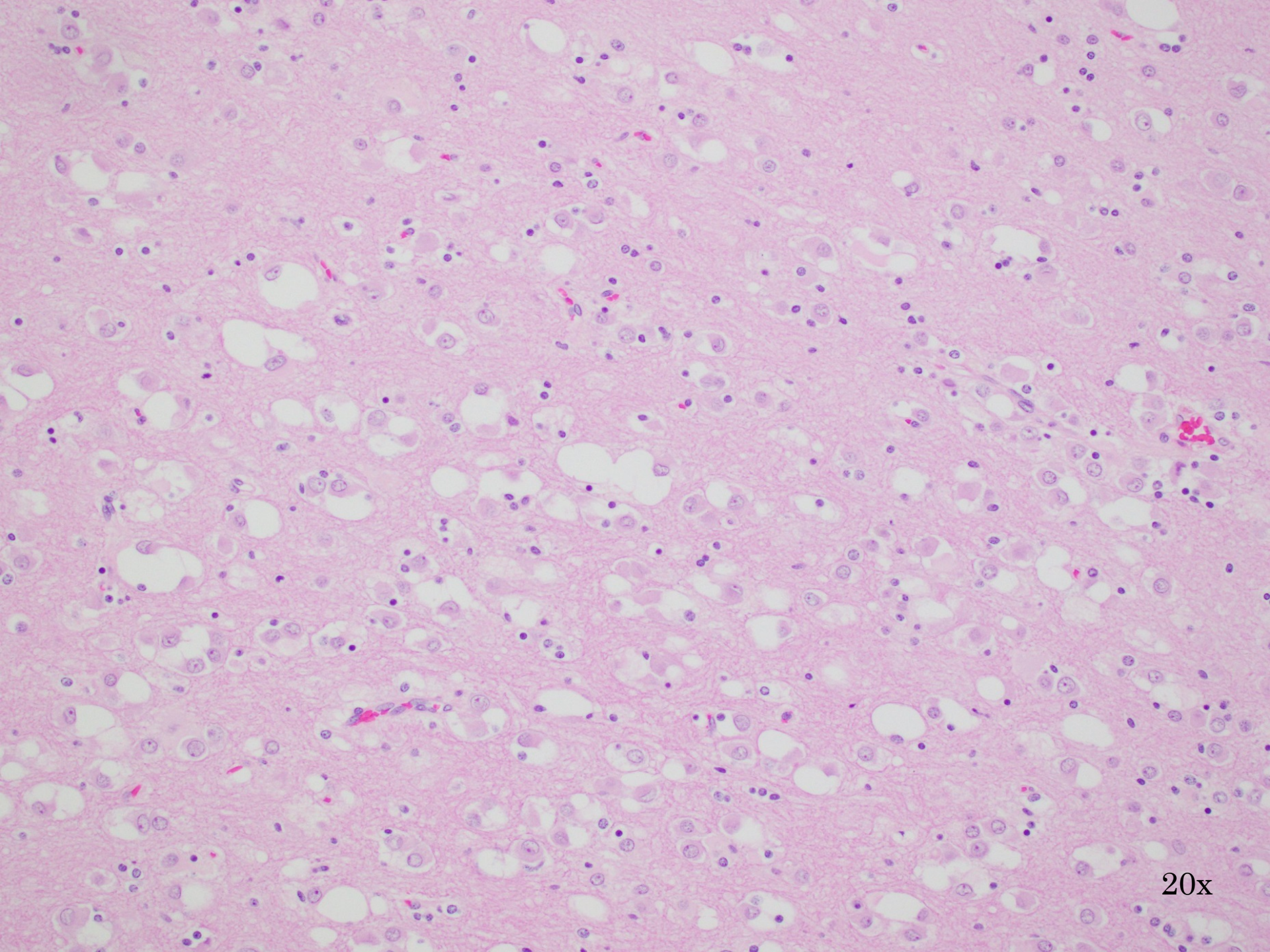
Resection

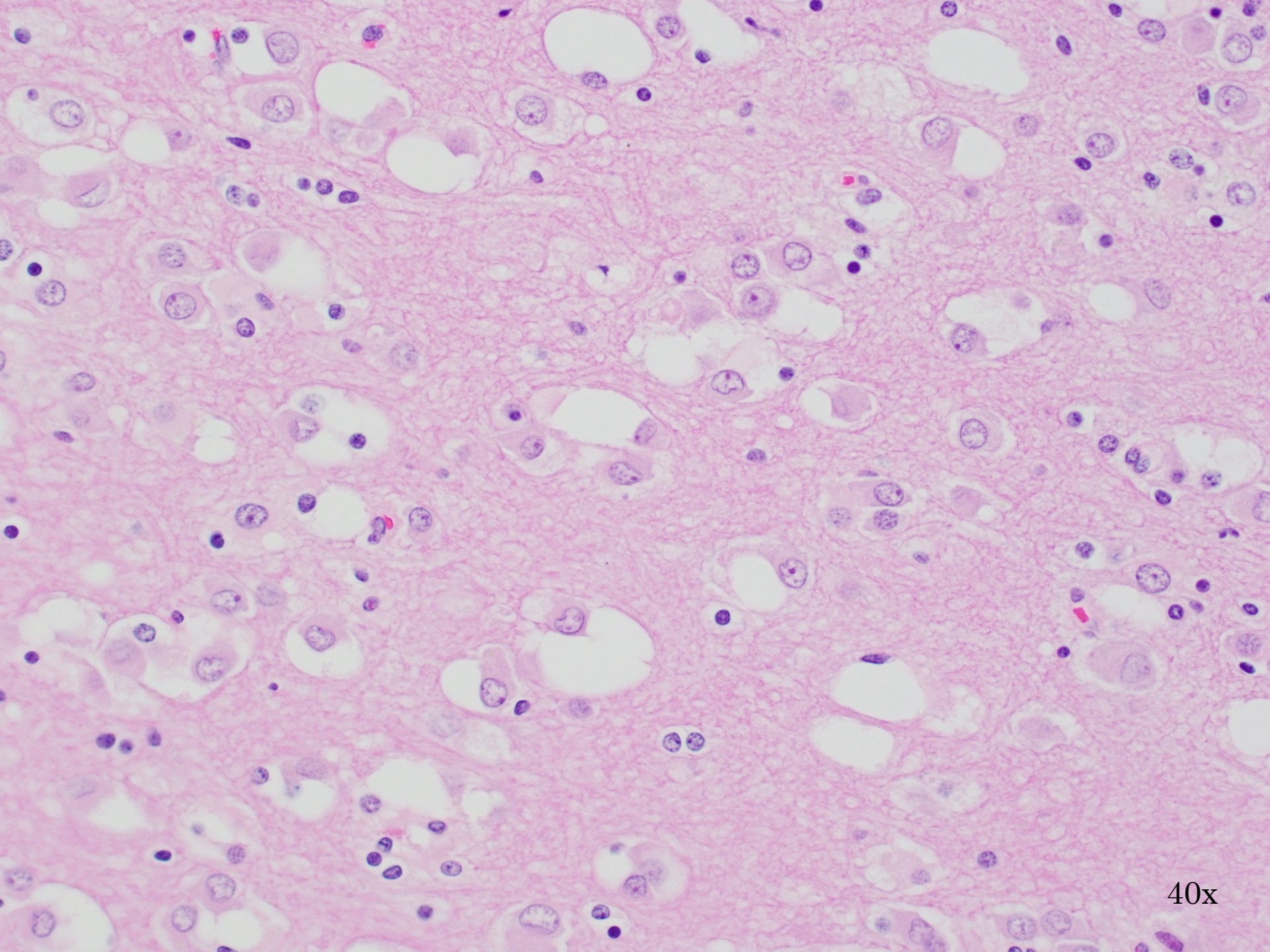
Gross Findings:

- 4.5 x 4.2 x 0.8 cm right lateral temporal lobe fragment
- Slightly yellow/white lesion with visually indistinct border; along the cortical-white matter junction


4x

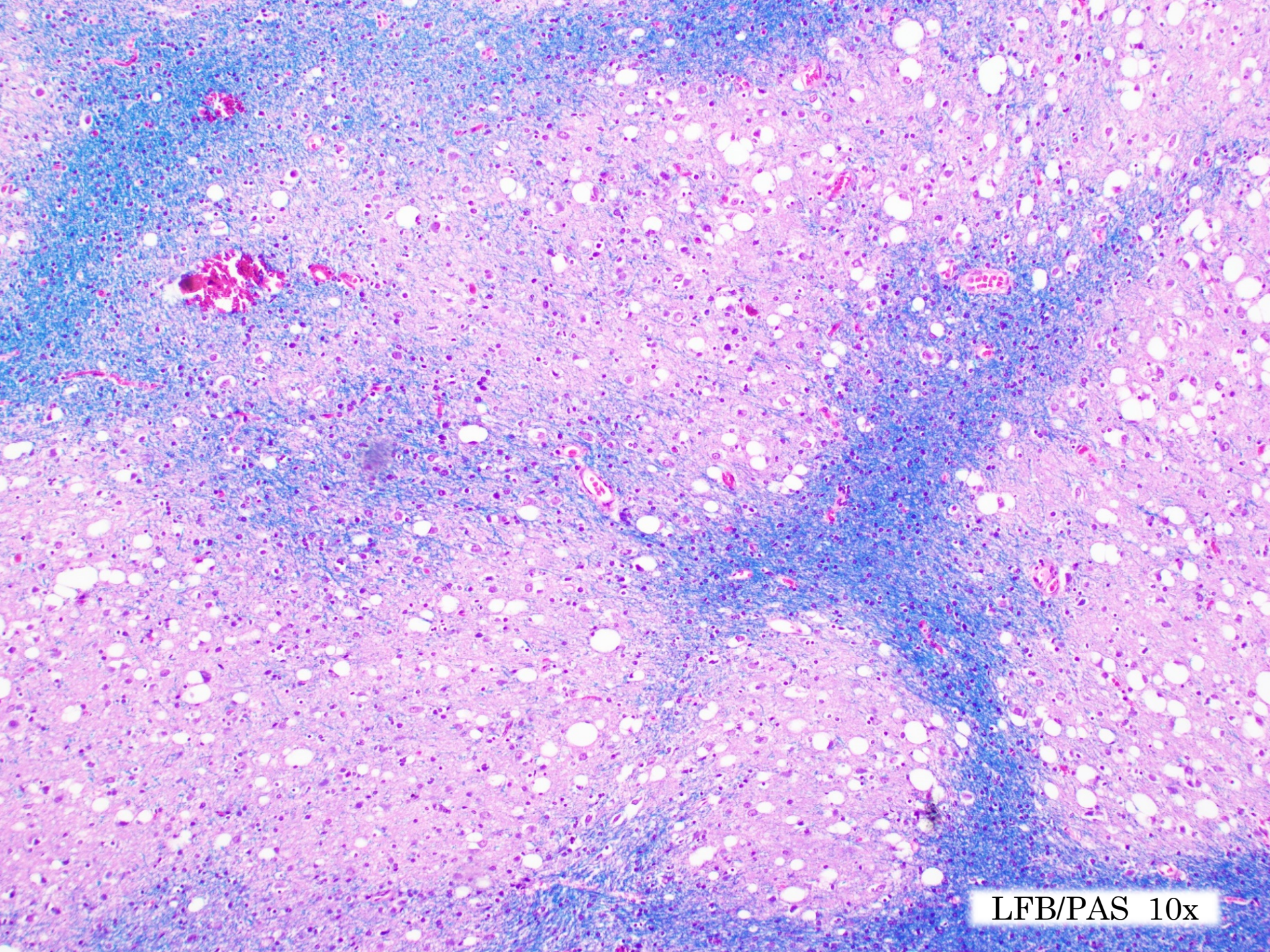

10x

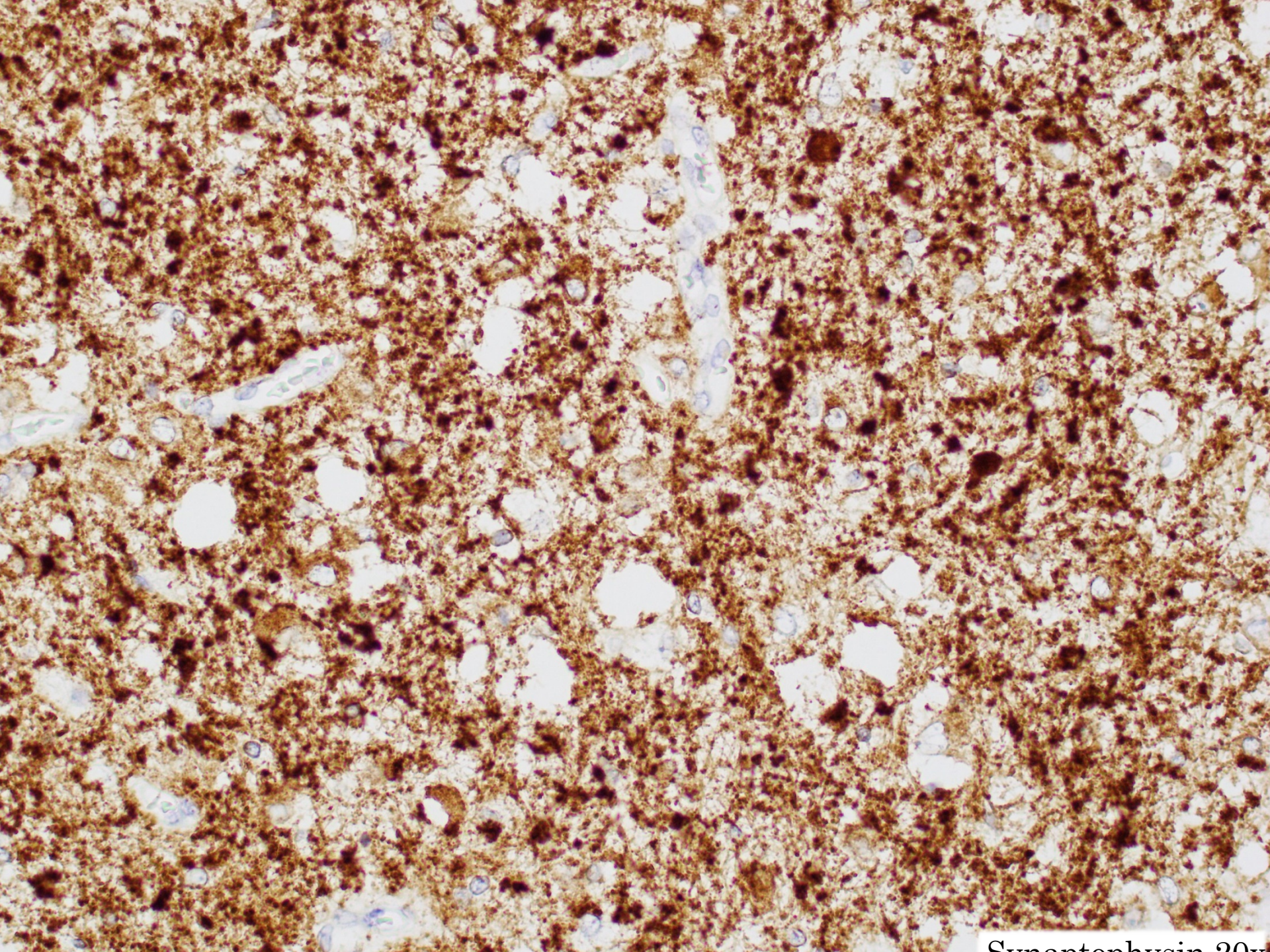

20x

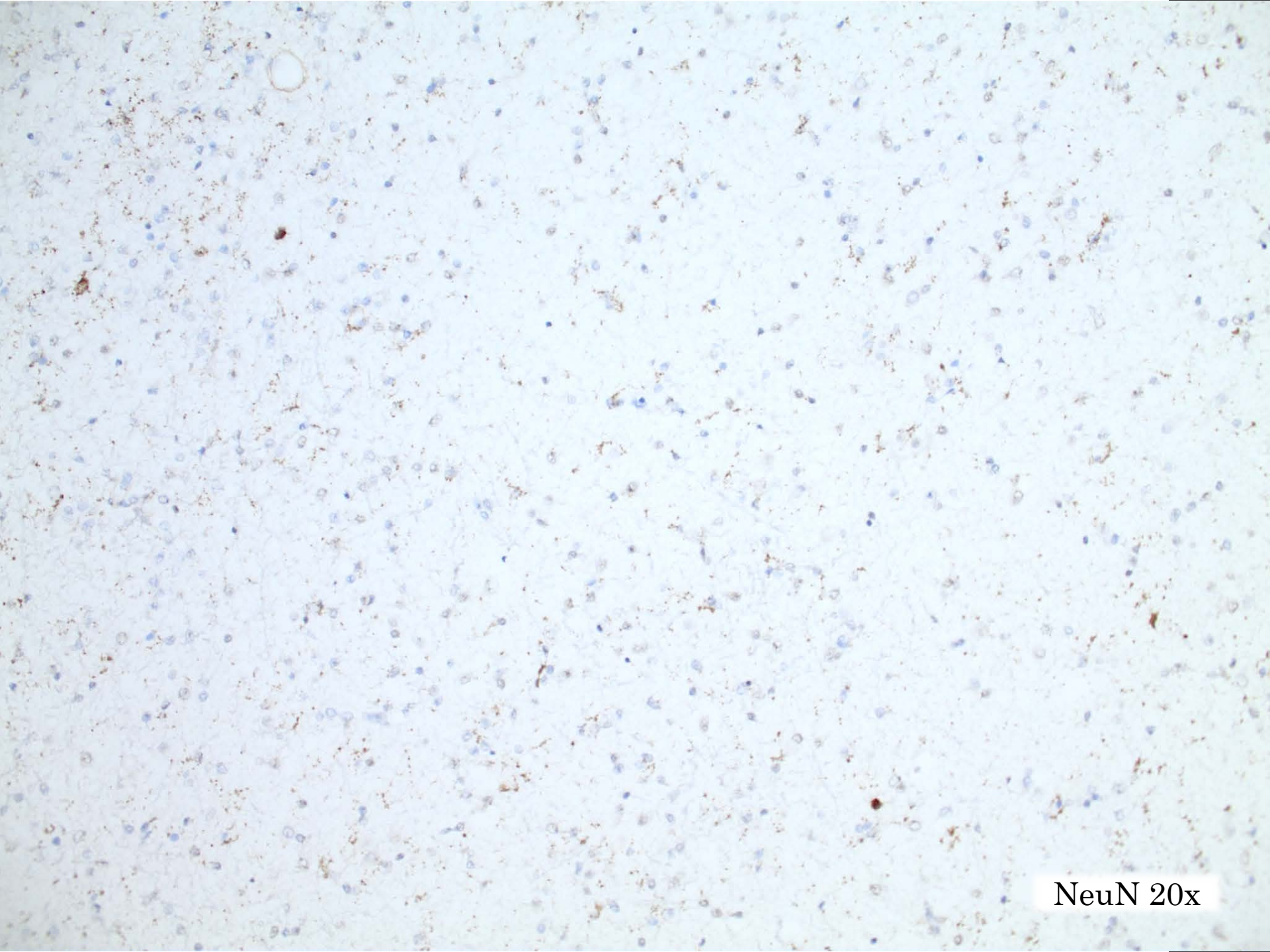

40x

Differential Diagnosis?

Additional studies


LFB/PAS 10x


Synaptophysin 20x


NeuN 20x

IHC stain summary for our case:

Stain	Result
Synaptophysin	Positive in tumor cells
Neu-N	Negative in tumor cells
GFAP	Negative in tumor cells
IDH-1	Negative in tumor cells
ATRX	Intact
p53	Negative in tumor cells

Diagnosis

Multinodular and Vacuolating Neuronal Tumor of Cerebrum

(MVNT)

MVNT

- Provisional entity of the 2016 WHO classification
- Well-differentiated, slow growing neuroepithelial neoplasm
- Exceedingly rare tumor

Clinical and Imaging

- Mostly adults, median age 39.5 years
- Predominantly in temporal lobe (9 cases, 64%)
- Seizures are most common

- MRI:
  - Subtle nodularity at the cortical-white matter junction
  - Hyperintensity on T2 weighted and FLAIR MRI
  - “Bubbly” appearance
  - Absence of enhancement

Histologic Findings

- Multiple well demarcated nodules in the deep half of cortex and subcortical white matter
- Neuroepithelial cells with large nuclei and distinctive nucleoli
- Intracellular and stromal vacuolation
- No atypia, multinucleation, mitosis or necrosis

Reported IHC and Ancillary Findings

HuC/HuD	Positive in tumor cells
OLIG2	Positive in tumor cells
Synaptophysin	Positive in tumor cells
Chromogranin	Negative in tumor cells
Neu-N	Negative in tumor cells
GFAP	Negative in tumor cells
Vimentin	Positive in nodules stroma
CD34	Positive in the neighboring cerebral cortex
IDH1/IDH2	No mutation
BRAF V600E	No mutation
1p19q	No co-deletion

Differential Diagnosis

- Glioneuronal and mixed glioneuronal lesions
  - Gangliogliomа/gangliocytoma
  - Dysembryoplastic neuroepithelial tumor
  - Focal cortical dysplasia

Prognosis

- Overall benign tumor (similar to WHO grade I)
- No recurrence with total or subtotal resection

References

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2. Huse JT, Edgar M, Halliday J, Mikolaenko I, Lavi E Rosenblum MK. Multinodular and vacuolating neuronal tumor of the cerebrum. Brain Pathol 2013; 23: 515-524.

3. Bodi I, Curran O, Selway R et al. Two cases of Multinodular and vacuolating tumor. Acta Neuropathol Commun 2014; 2: 7.

4. Nagaishi M, Hideaki Y, Sumihito N. Localized overexpression of alpha-internexin within nodules in multinodular and vacuolating neuronal tumors. Neuropathology 2015;35,561-568.

Thank you!