2015 Diagnostic Slide Session

Case 3

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Disclosures

No financial disclosures or conflicts of interest

Clinical History

- 42-year-old male with complicated history of rapidly progressive neurologic deterioration
- Treated for presumptive diagnosis of tumefactive multiple sclerosis with minimal improvement
  - Solumedrol, Cyclophosphamide, plasmapharesis
- Initial biopsy:
  - Features consistent with a “macrophage-rich lesion”

Autopsy Findings

- Brain (gross examination):
  - Mild bilateral cerebral edema
  - Bi-hemispheric disease: poorly defined, concentric foci of “coarsening” of white matter in centrum semiovale
  - Poorly delineated, focally hemorrhagic lesion (~1.5 cm maximal dimension) in the right cingulate gyrus
  - Hemorrhagic cystic cavity in the right putamen


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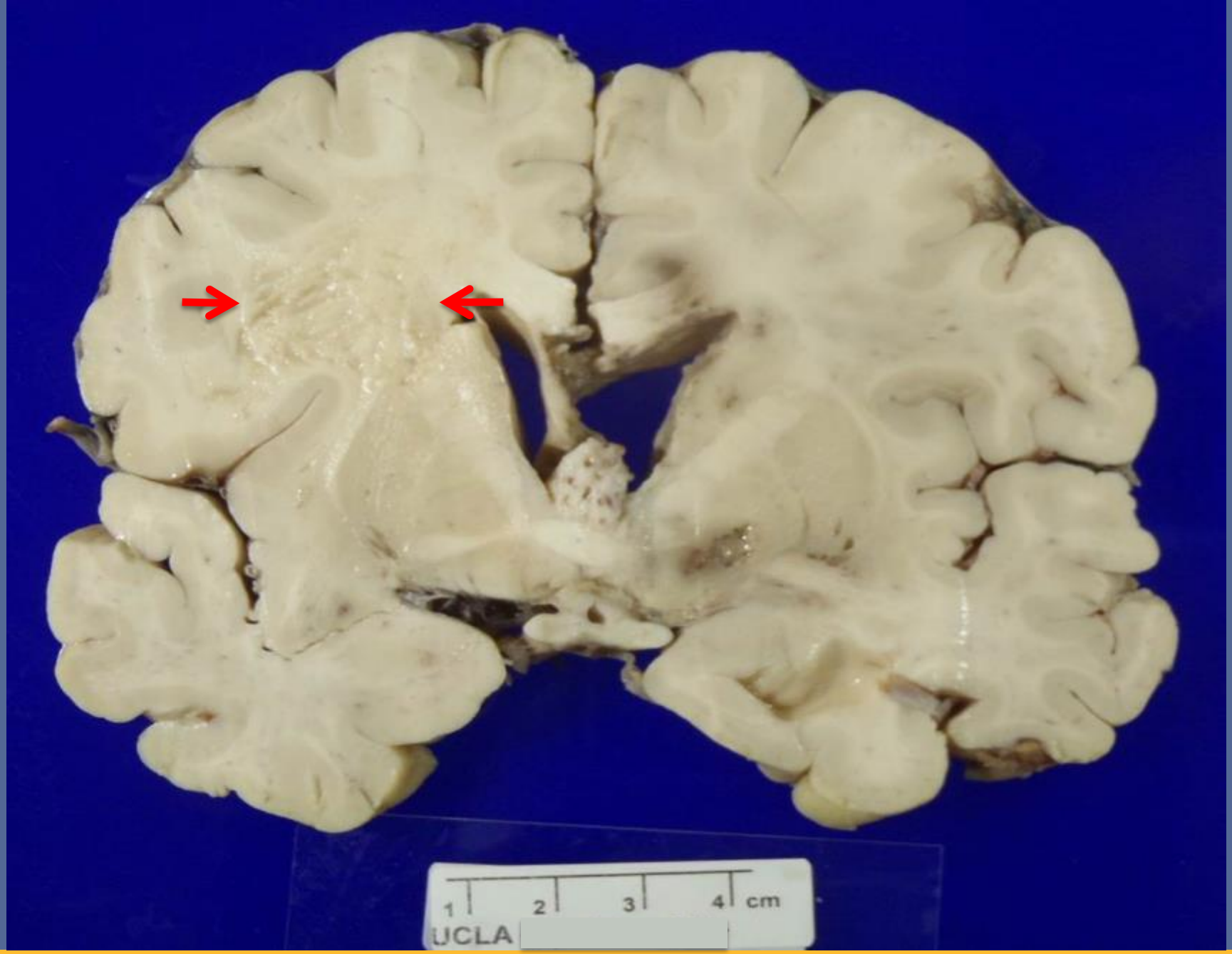
cm
UCLA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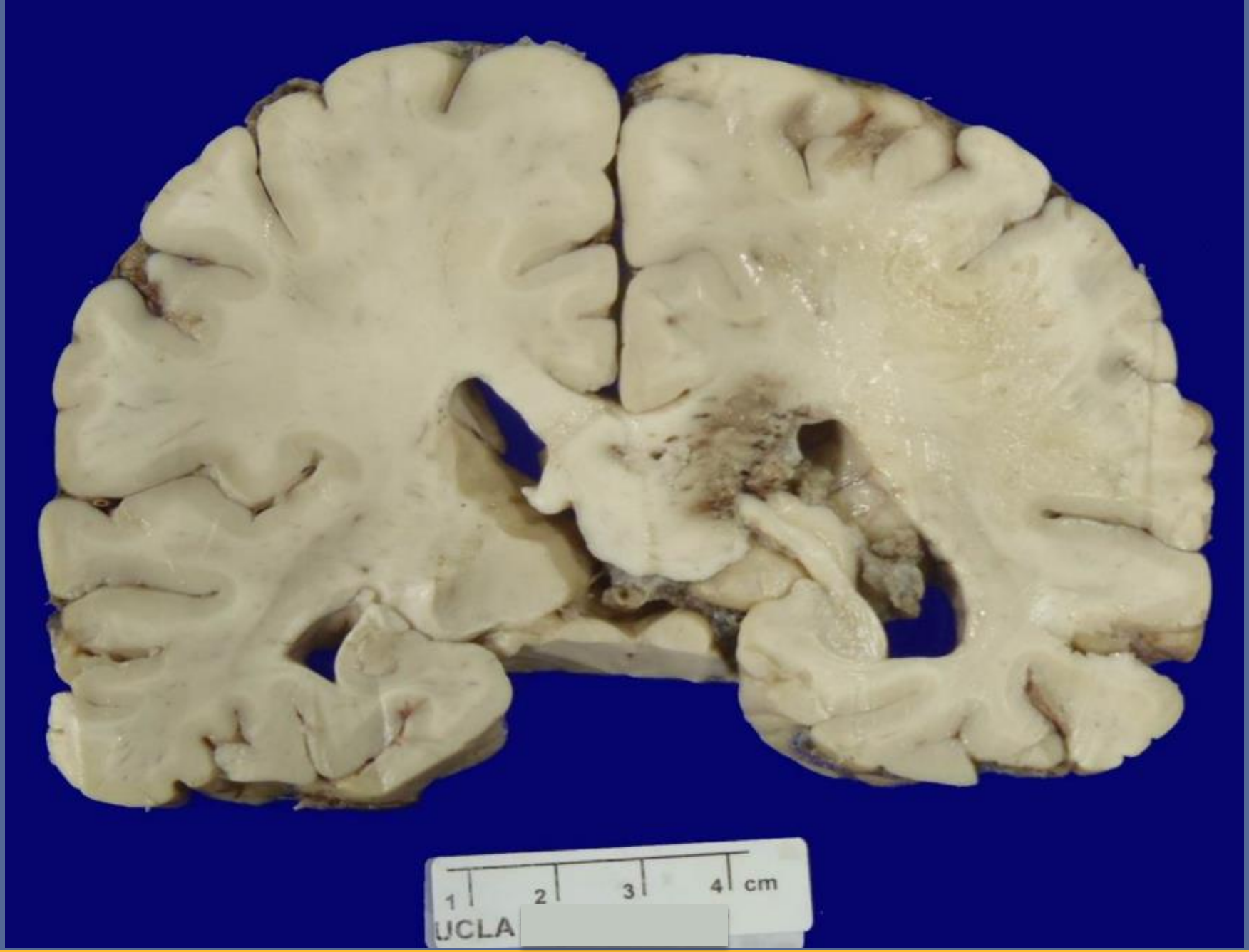
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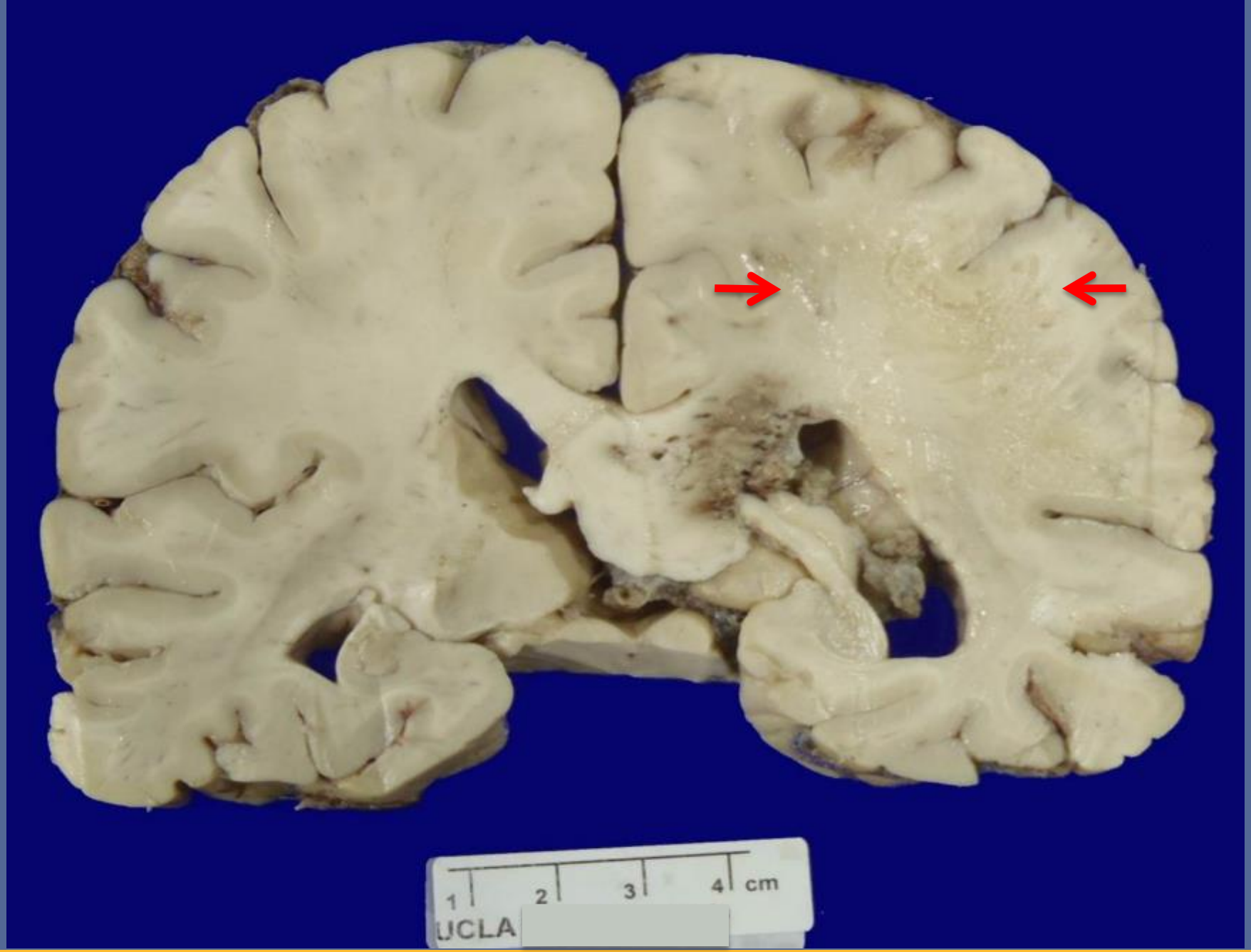
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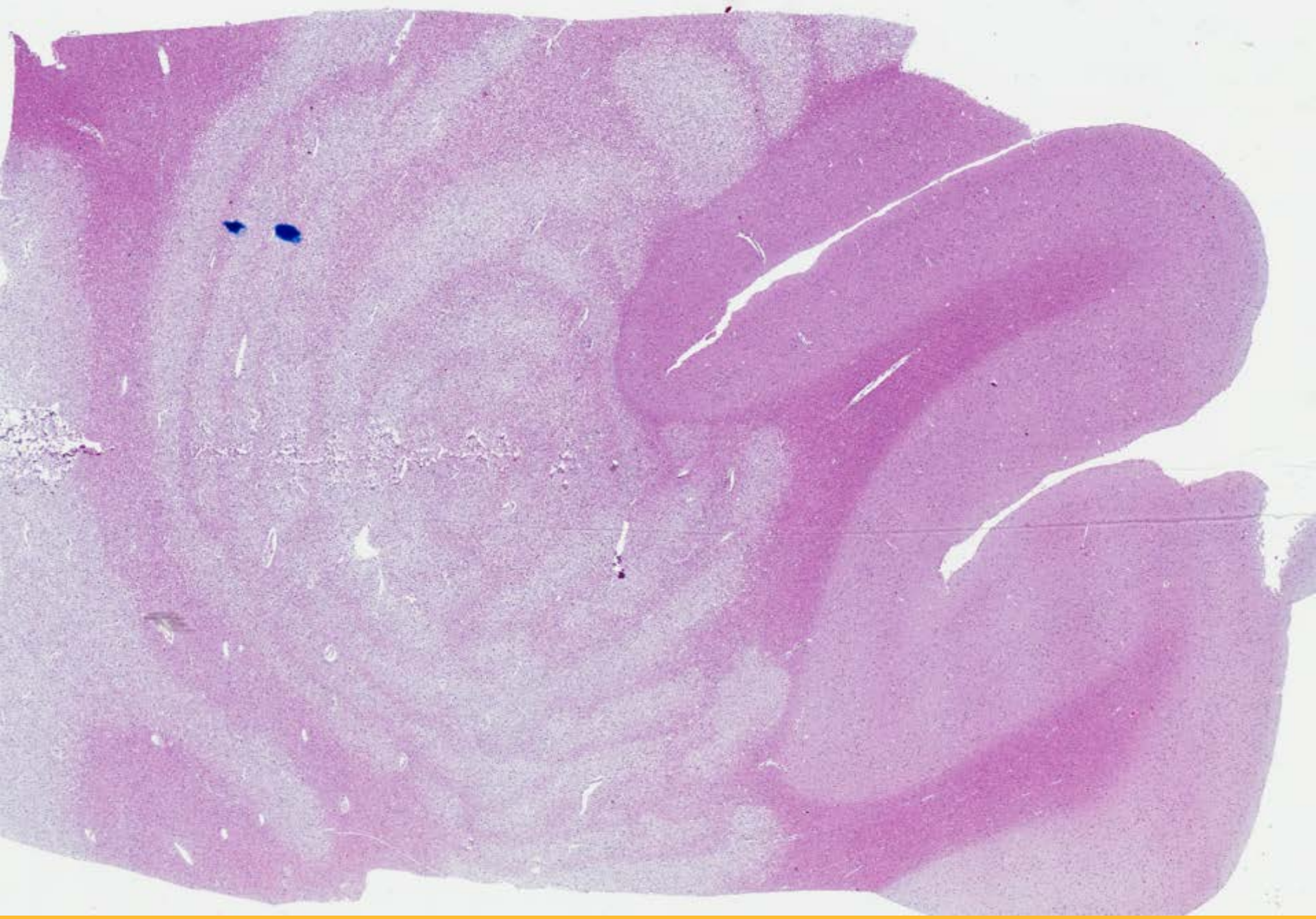
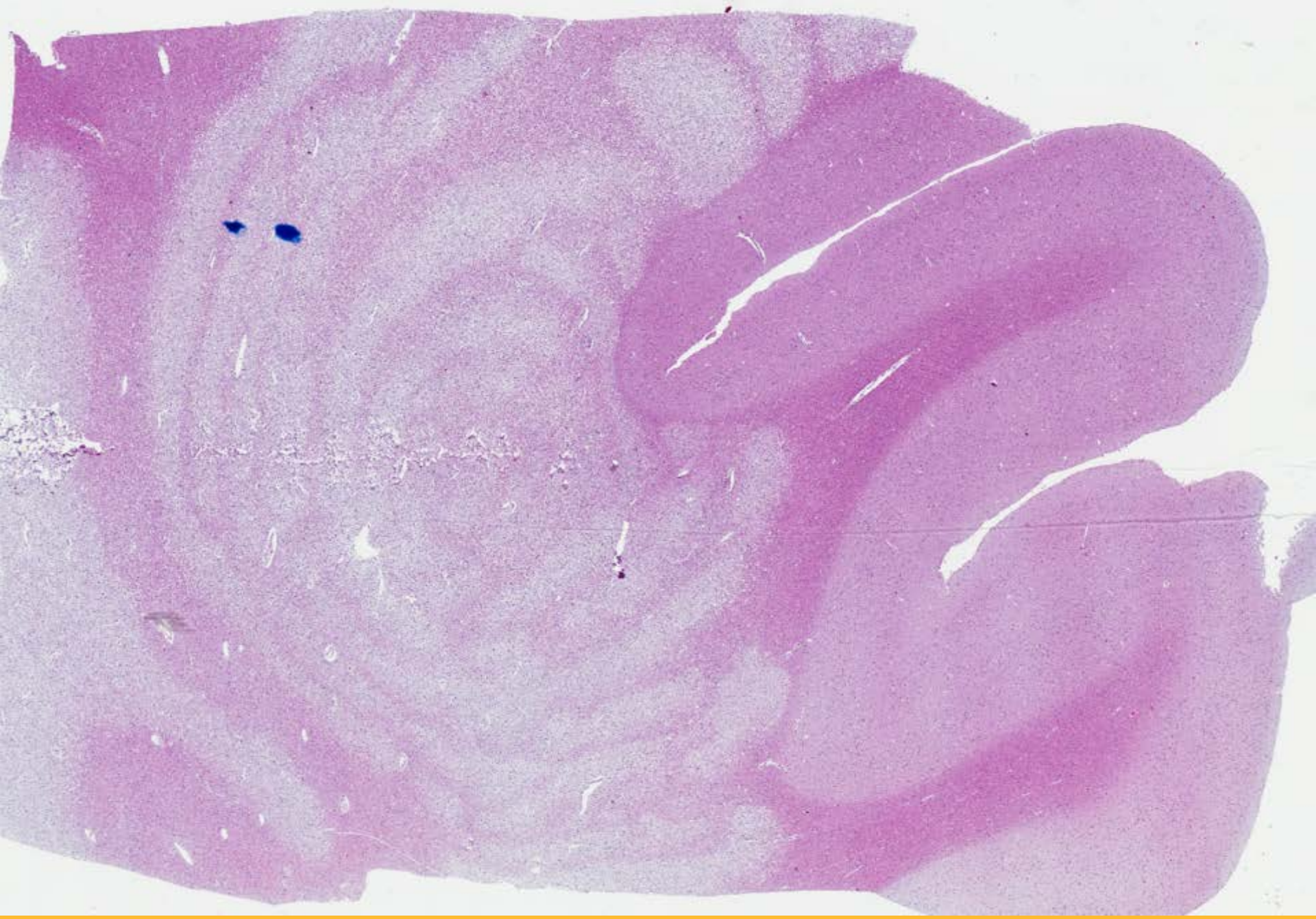
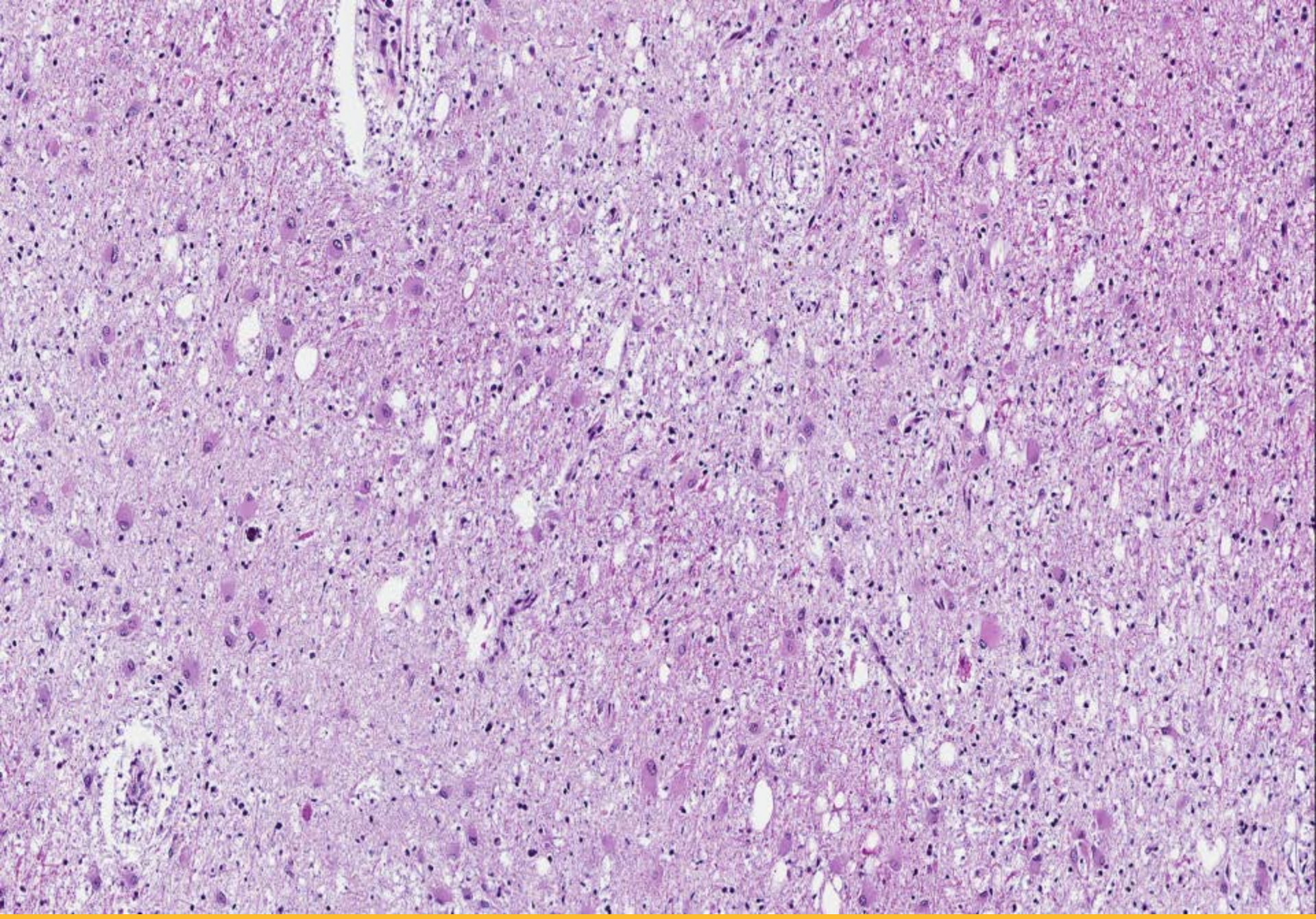


H&E, Whole Mount

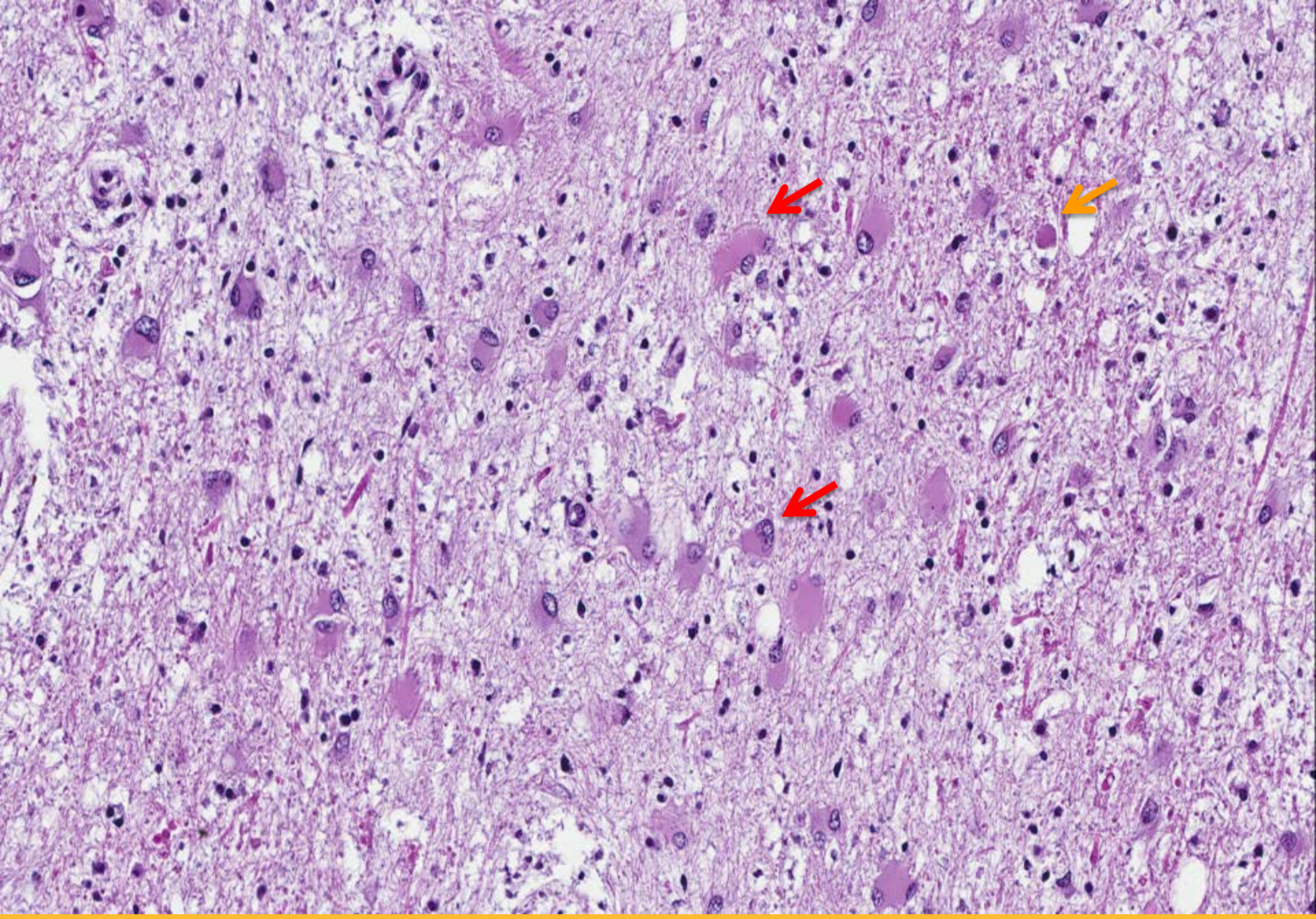
Diagnosis?



H&E, Whole Mount

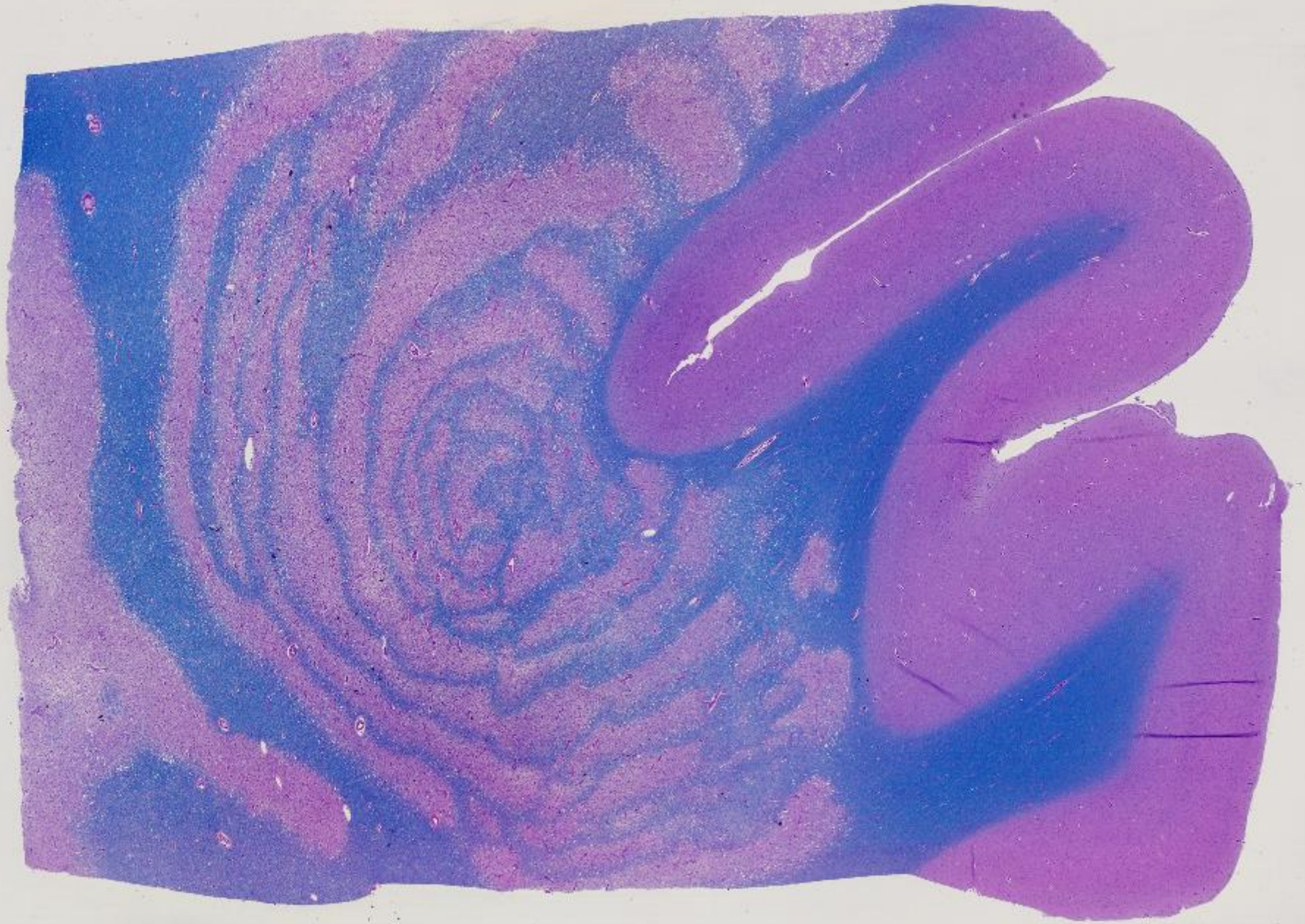


H&E, 10x

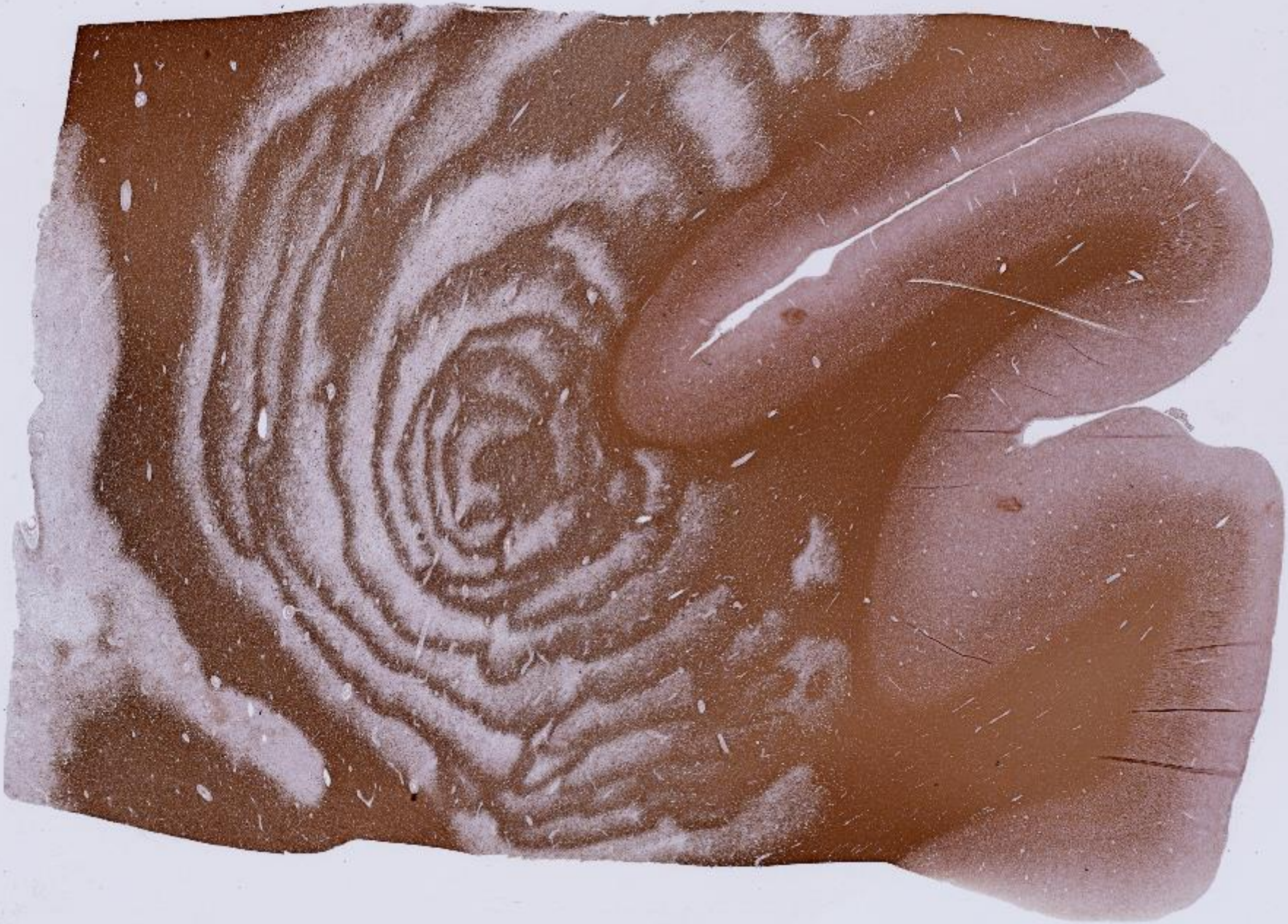


H&E, 20x

UCLA Health System



Klüver-Barrera, Whole Mount



Myelin Basic Protein, Whole Mount

UCLA Health System

Microscopic Findings

- Pathologic findings restricted to white matter in a multifocal, bi-hemispheric distribution
  - Concentric rings and bands of demyelination alternating with relatively spared (myelinated or partially demyelinated) areas
  - Abundant bizarre gemistocytic astrocytes, including many binucleated and multinucleated forms
  - Scattered neuroaxonal spheroids

Final Diagnosis

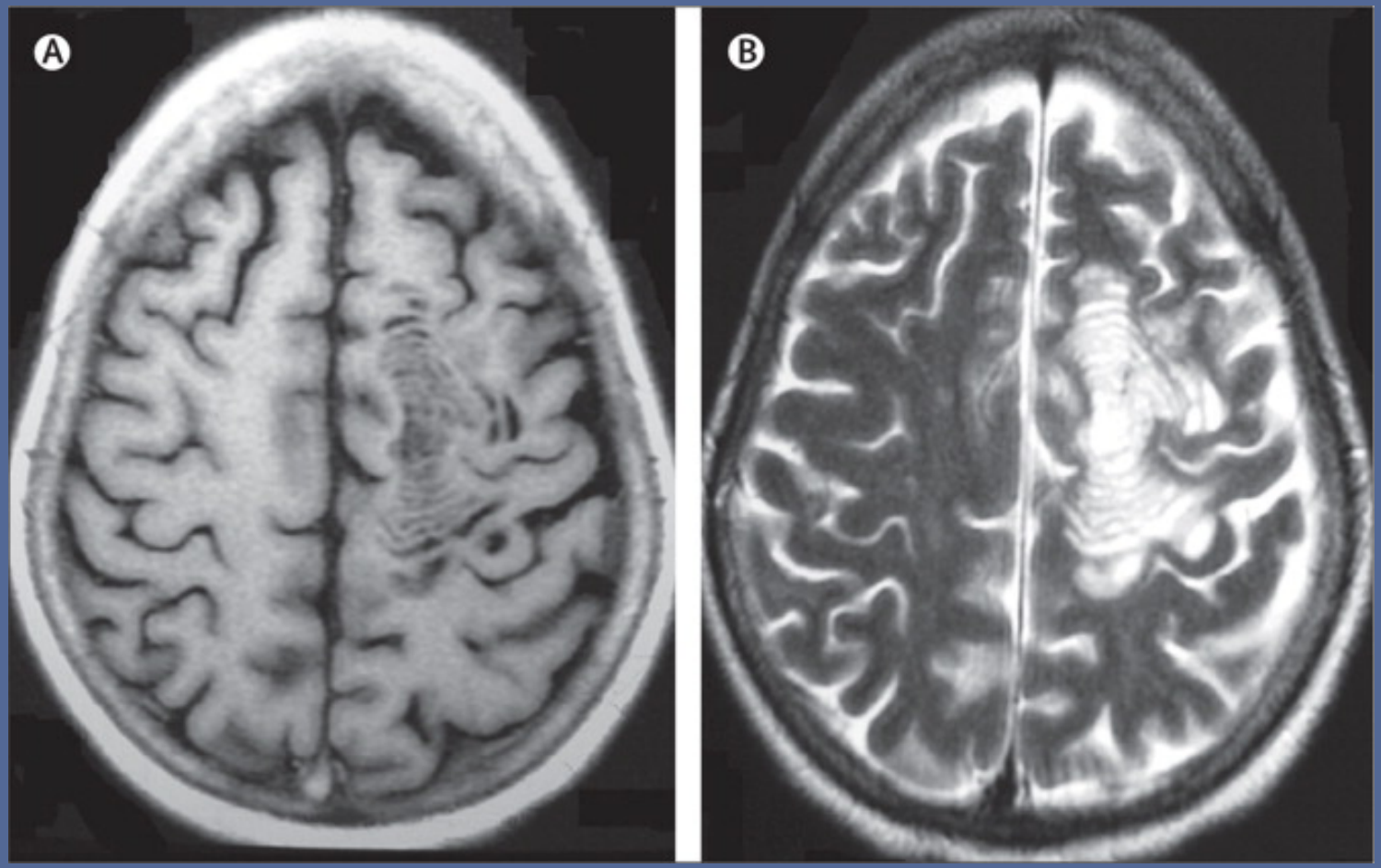
Baló's Concentric Sclerosis

Baló's Concentric Sclerosis

- Rare demyelinating disorder, considered a variant of multiple sclerosis
- Clinical presentation: acute or subacute neurological deterioration, sometimes fulminant and fatal

Baló's Concentric Sclerosis

- Imaging features:
  - Characteristic concentrically-layered lesion ("ringed" or "whorled" appearance) on MRI
  - Layered appearance distinguishes Baló's concentric sclerosis from demyelinating lesions of conventional multiple sclerosis
  - T1-weighted images: alternating isointense and hypointense concentric rings
  - T2-weighted images: alternating hyperintense and iso/hypointense layers surround a T2 hyperintense "storm center"
  - Enhancement may be seen at periphery of the lesion



MRI, T1-weighed

MRI, T2-weighed

Hardy TA, Miller DH. Baló's concentric sclerosis. *Lancet Neurol*. 2014 Jul;13(7):740-6

Baló's Concentric Sclerosis

- Gross features:
  - White matter lesion with "onion bulb" appearance
- Histopathologic findings:
  - Astrocytopathy characterized by hypertrophic/bizarre reactive astrocytes (bi-/multi-nucleation common)
  - Demyelination: rings of relative myelin preservation (early or partial demyelination and/or remyelination) alternating with areas of demyelination with axonal sparing
  - White matter oligodendrocyte loss
  - Cortical gray matter is characteristically spared (unlike conventional MS)

Baló's Concentric Sclerosis

- Pathogenesis:
  - Not entirely clear
  - Unknown stimulus/stimuli
  - Develop around perivenular zone
  - Macrophages and activated microglia produce chemical mediators
  - Successive outward waves of chemical mediator → concentric rings of demyelination (hypoxia-ischemia ?)
  - Disruption of astrocyte/oligodendrocyte interaction ?

References

- Hardy TA, Miller DH. Baló's concentric sclerosis. *Lancet Neurol*. 2014 Jul;13(7):740-6

- Darke M, Bahador FM, Miller DC, Litofsky NS, Ahsan H. Baló's concentric sclerosis: imaging findings and pathologic correlation. *J Radiol Case Rep*. 2013 Jun 1;7(6):1-8.

Thank you!