

2015 Diagnostic Slide Session

Case 3

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Disclosures

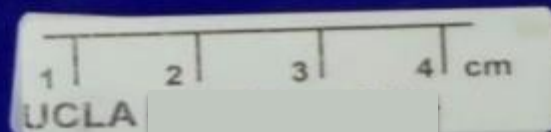
No financial disclosures or
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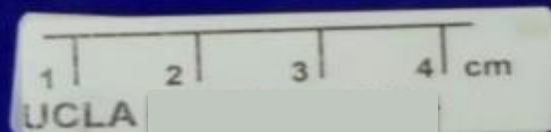
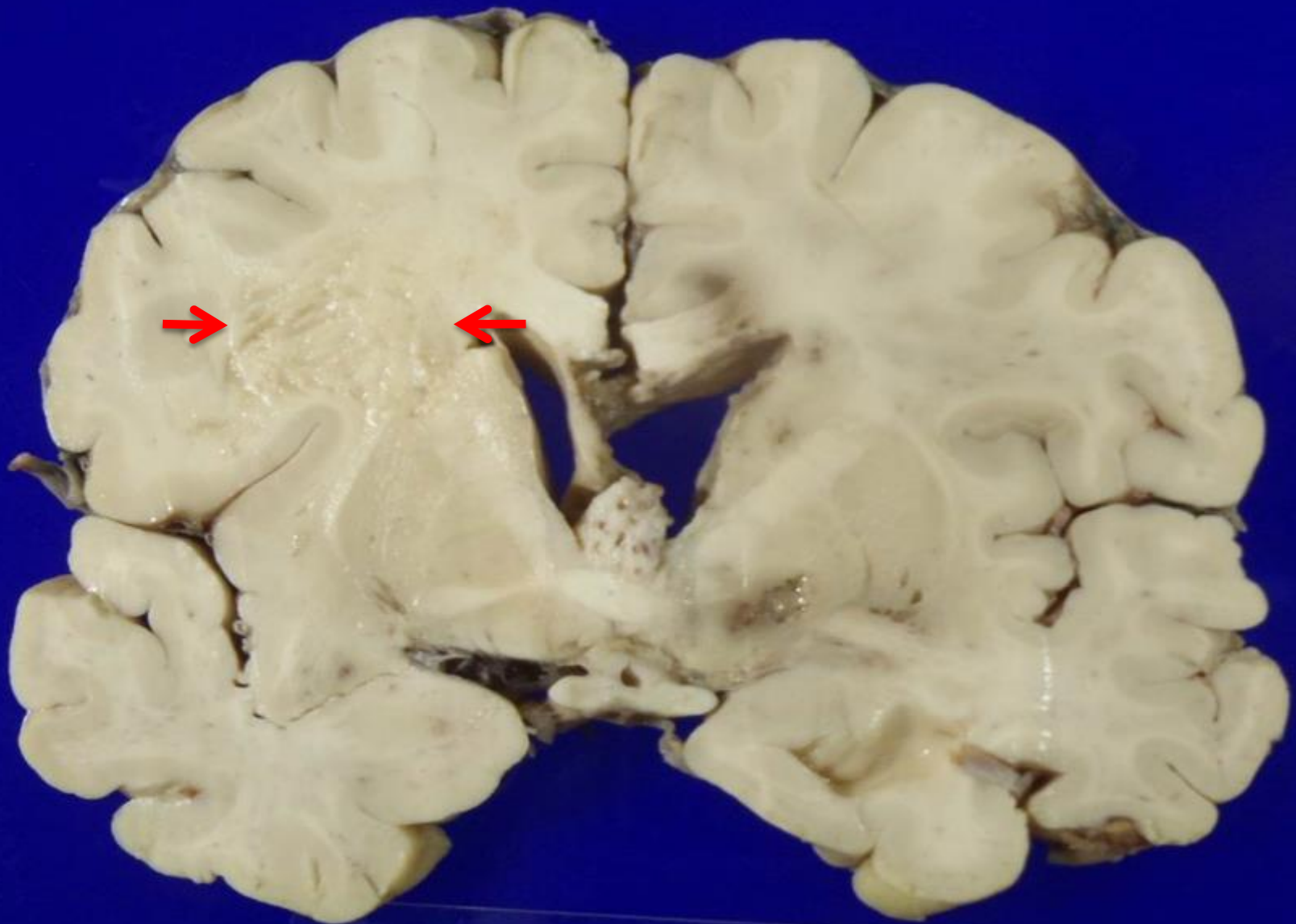
Clinical History

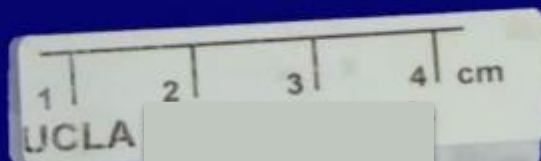
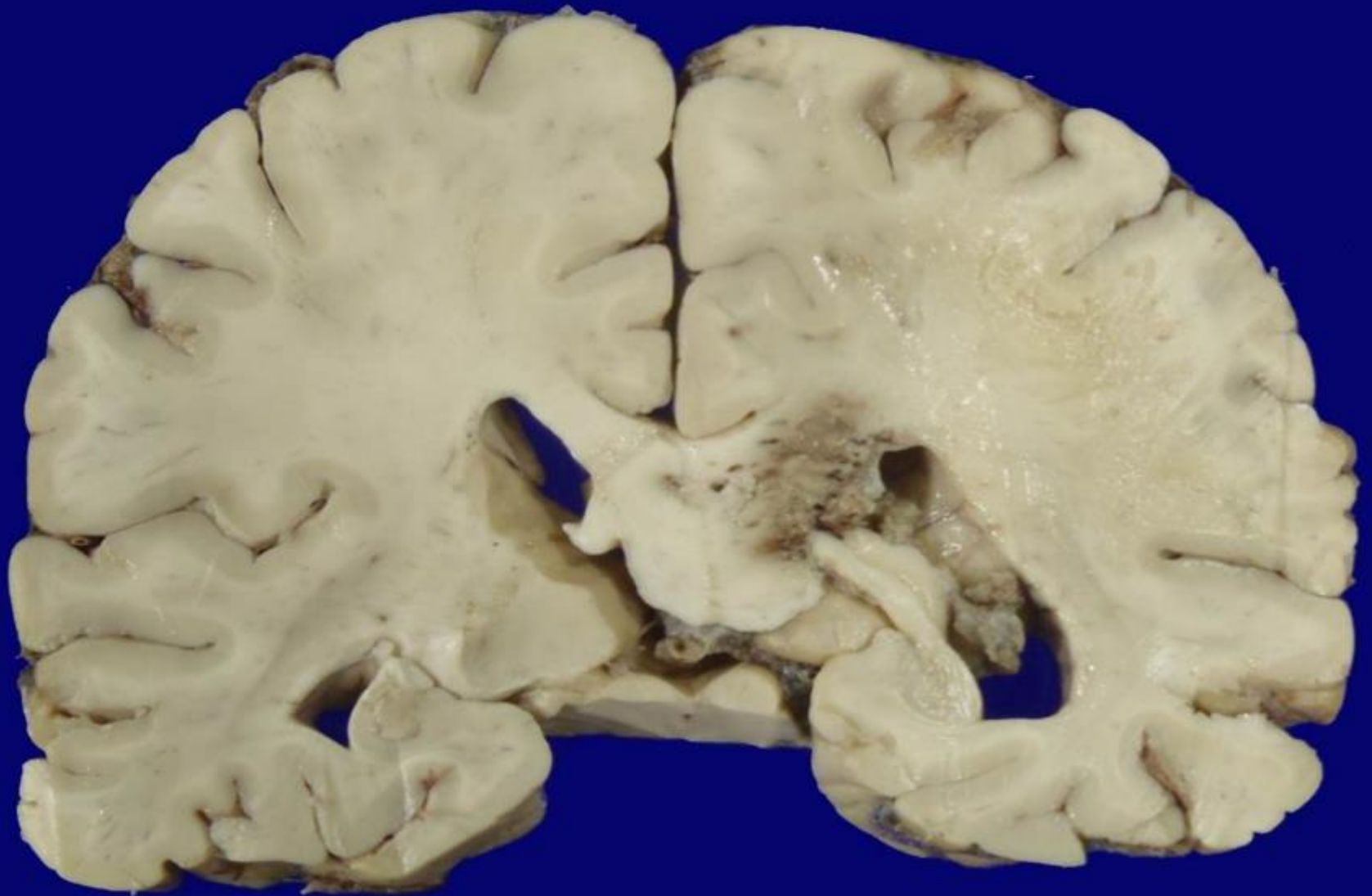
- 42-year-old male with complicated history of rapidly progressive neurologic deterioration
- Treated for presumptive diagnosis of tumefactive multiple sclerosis with minimal improvement
 - Solumedrol, Cyclophosphamide, plasmapheresis
- Initial biopsy:
 - Features consistent with a “macrophage-rich lesion”

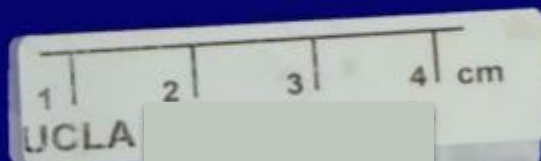
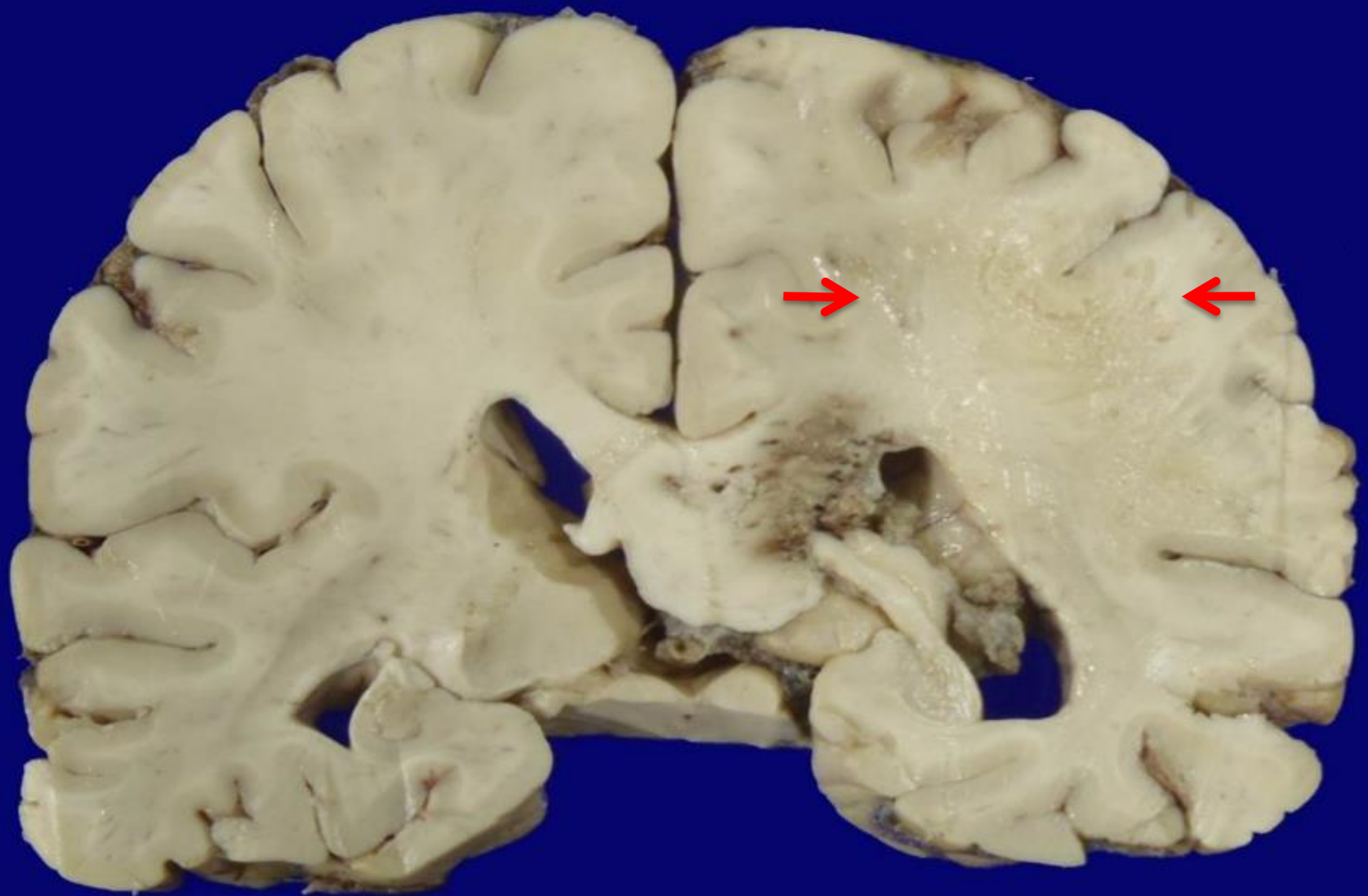
Autopsy Findings

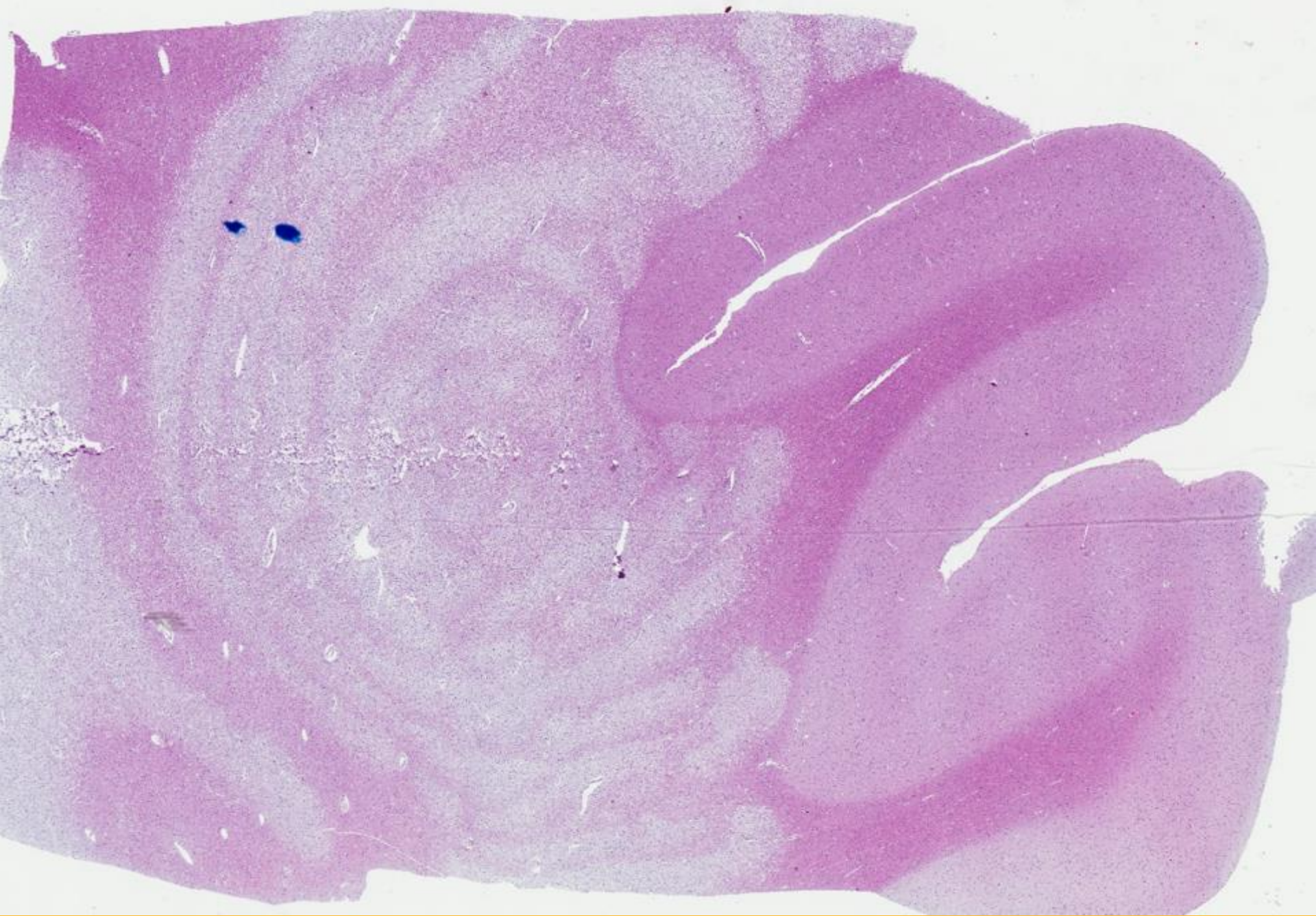
- Brain (gross examination):
 - Mild bilateral cerebral edema
 - Bi-hemispheric disease: poorly defined, concentric foci of “coarsening” of white matter in centrum semiovale
 - Poorly delineated, focally hemorrhagic lesion (~1.5 cm maximal dimension) in the right cingulate gyrus
 - Hemorrhagic cystic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



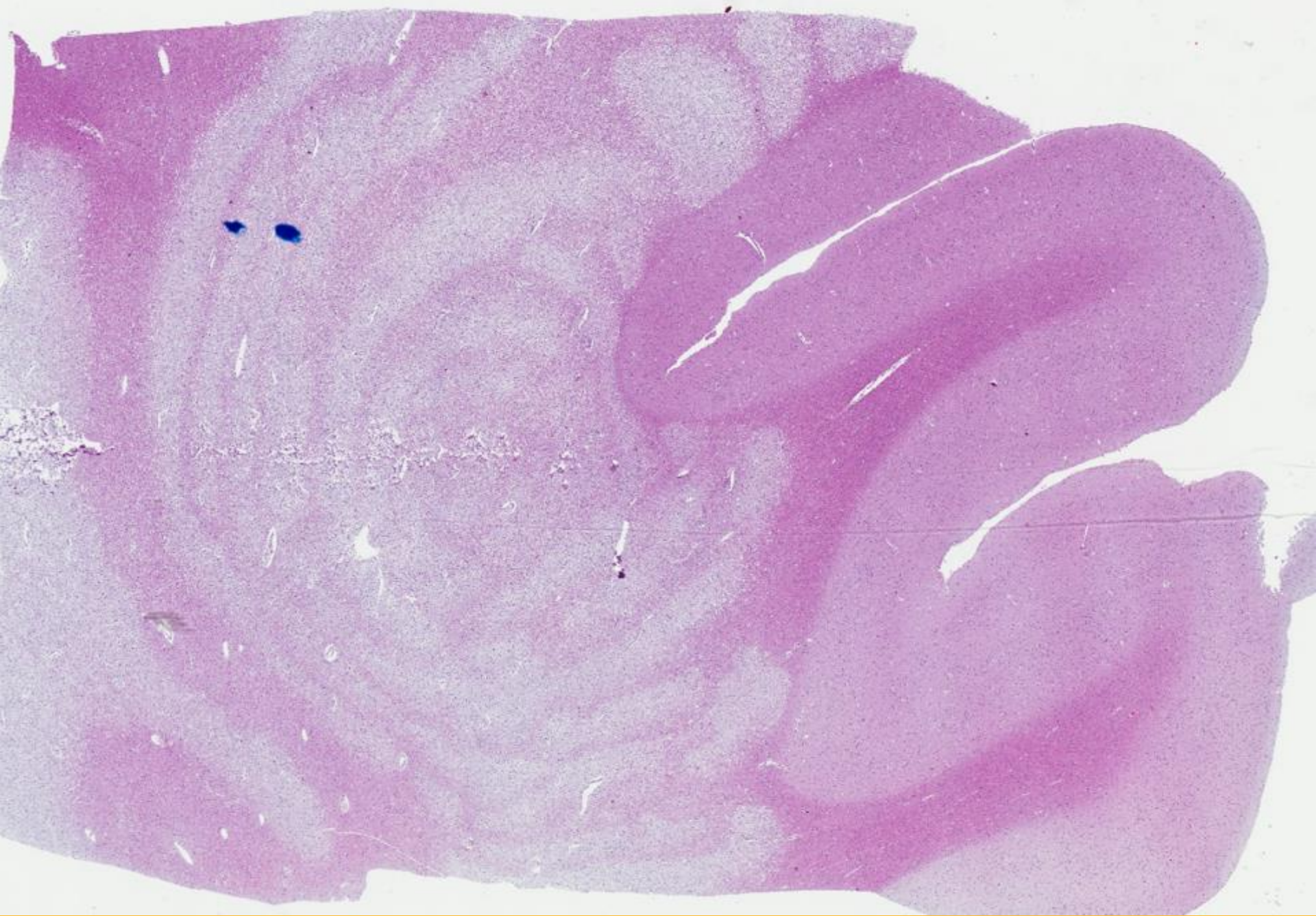


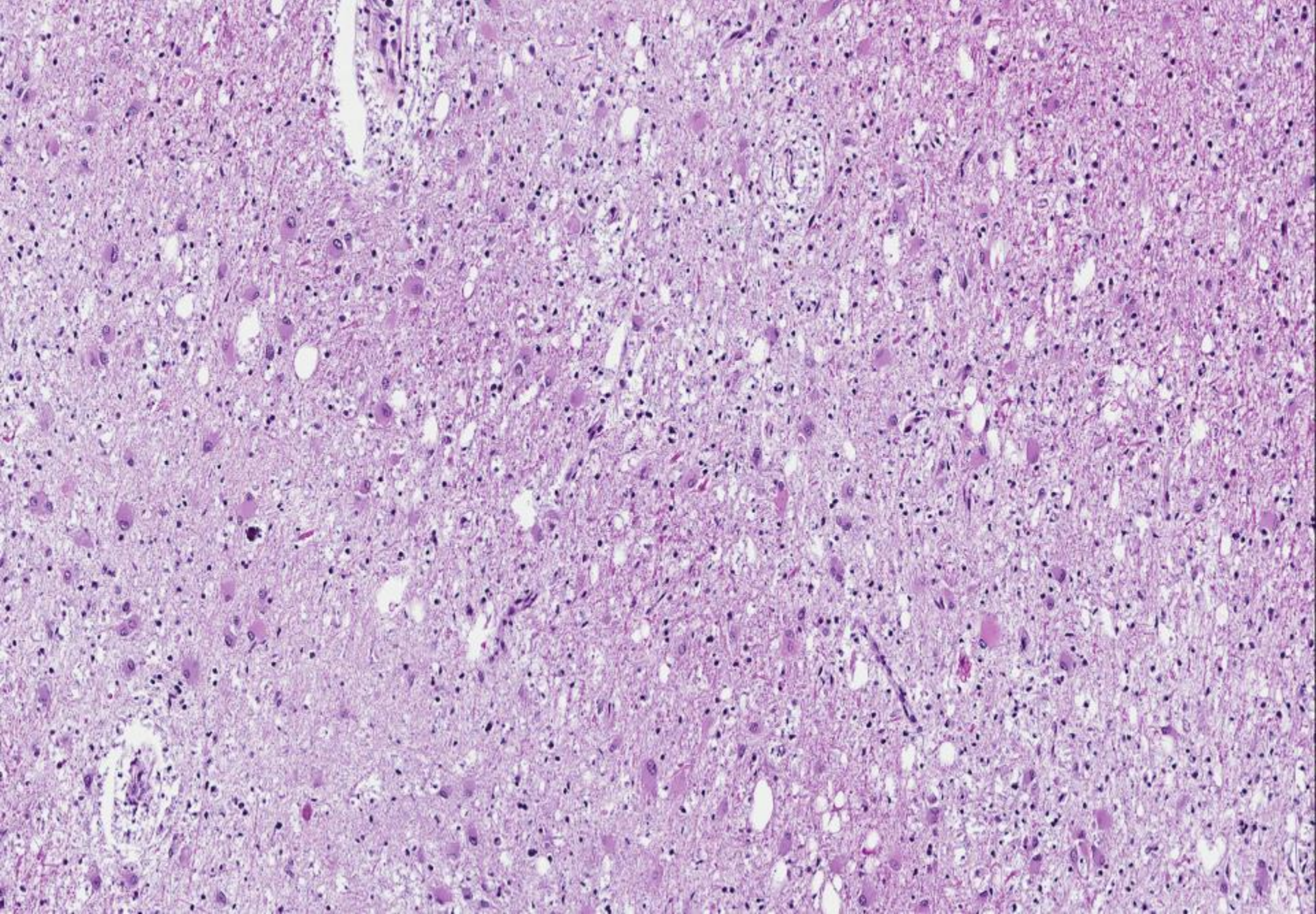


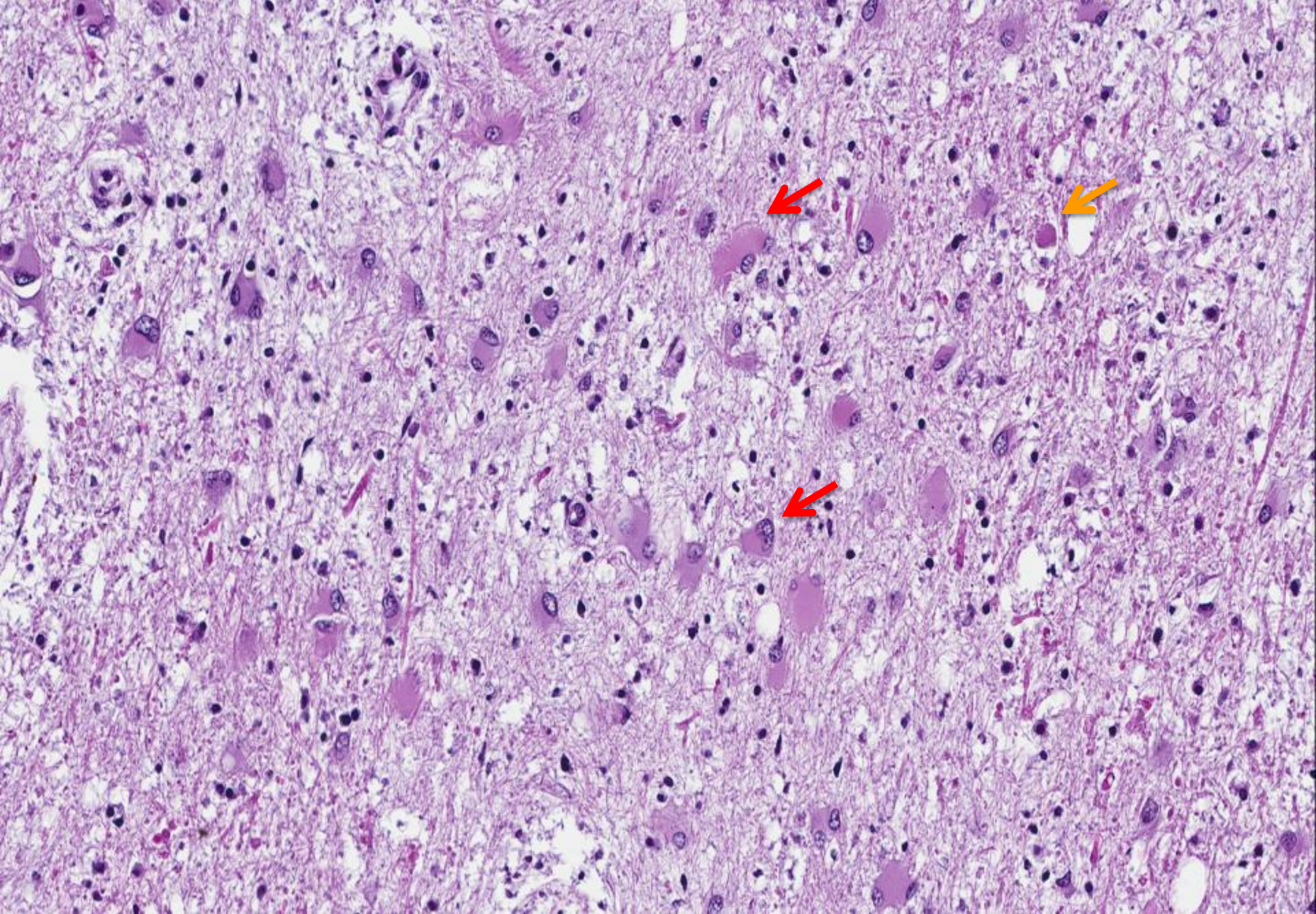


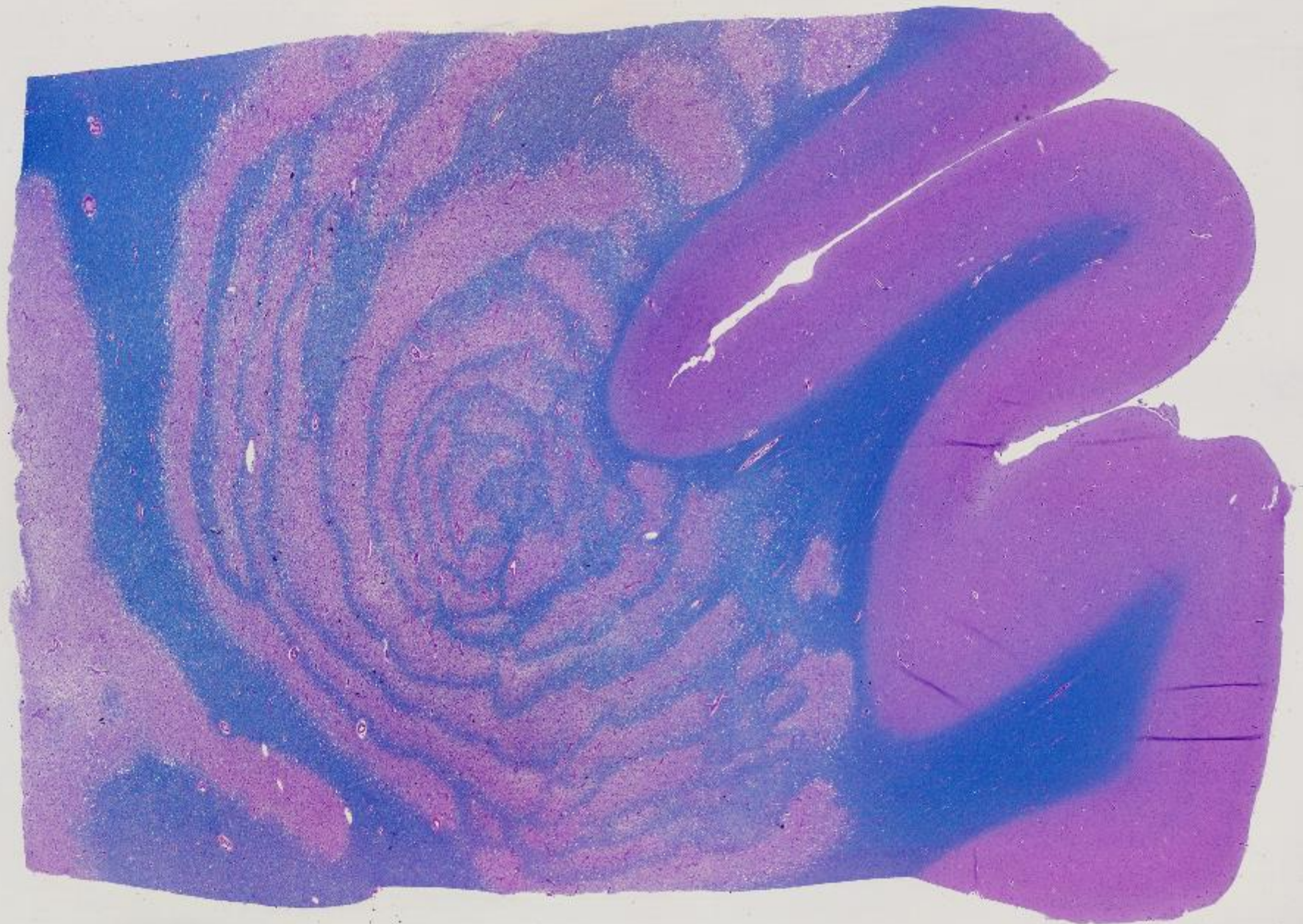


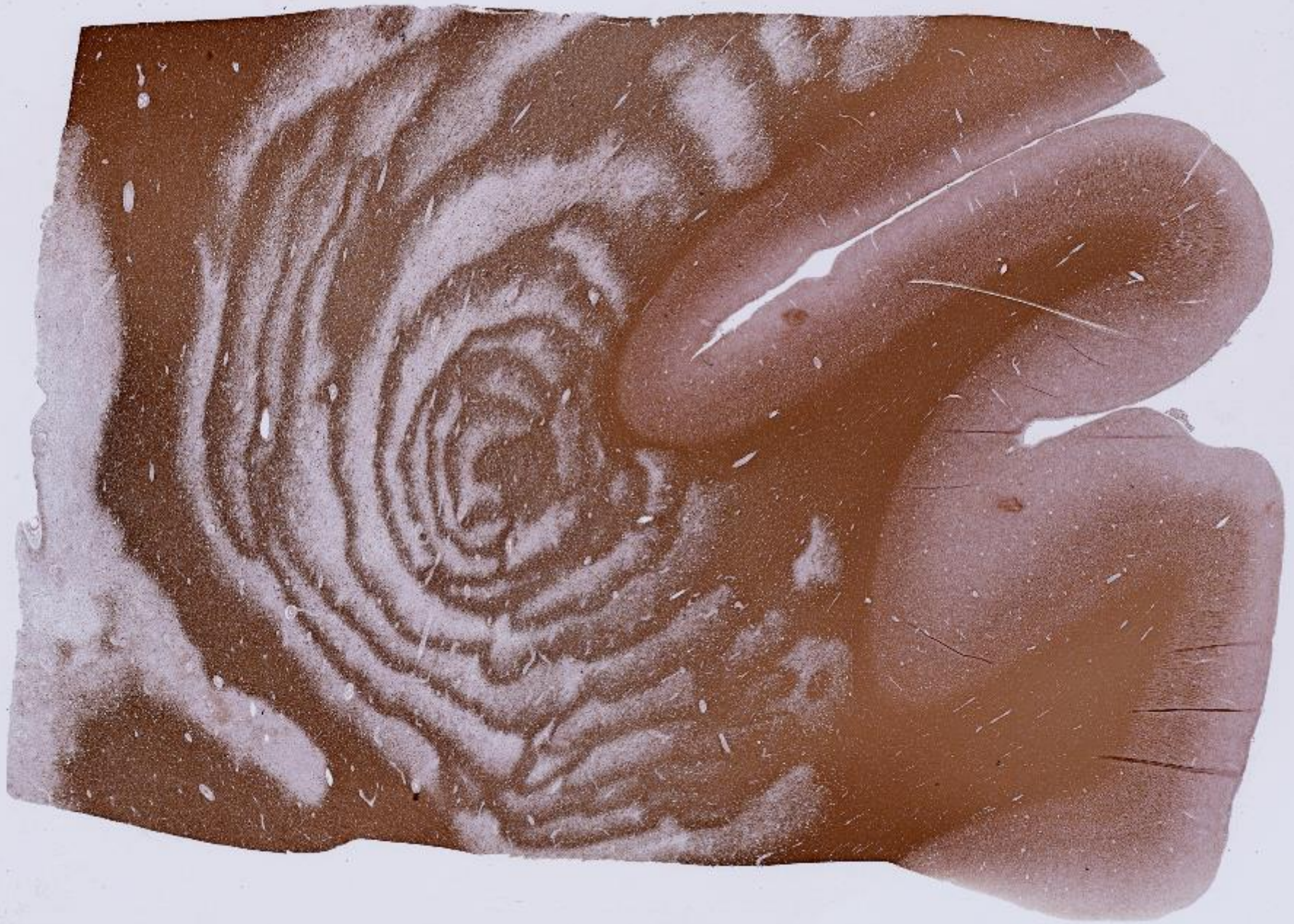
Diagnosis?











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

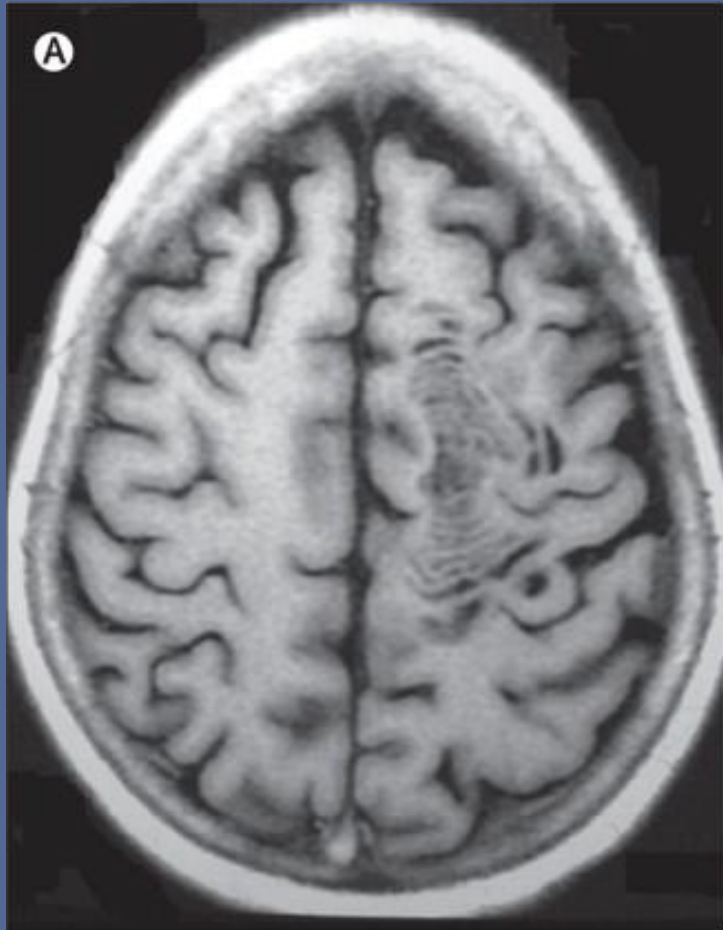
Baló's Concentric Sclerosis

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

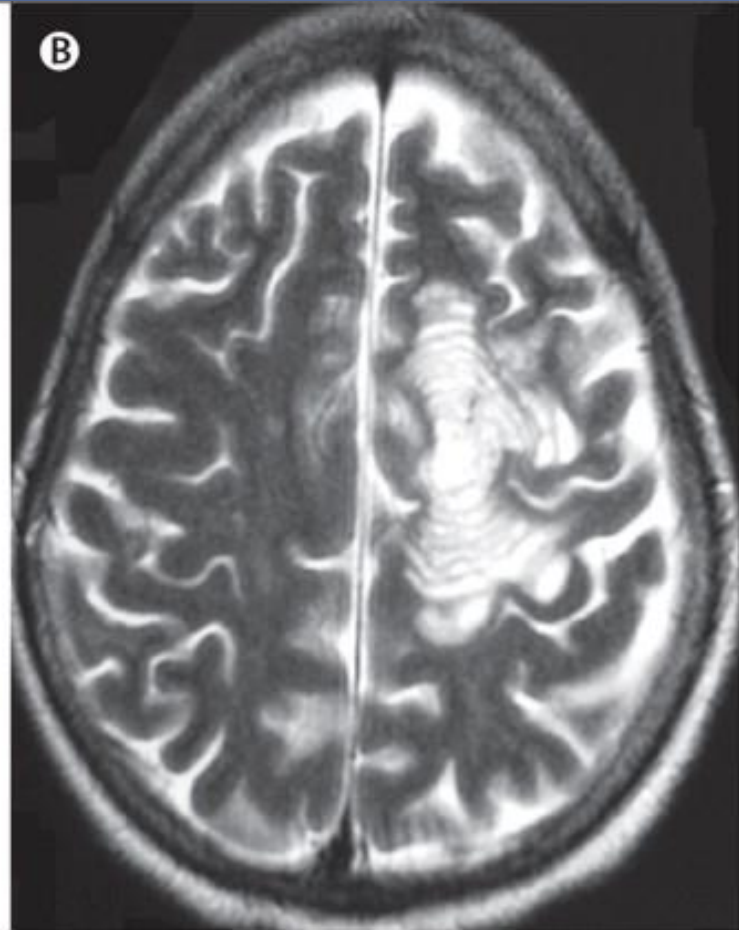
Baló's Concentric Sclerosis

- 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

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!