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, 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-weighted

MRI, T2-weighted
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

Thank you!