65th ANNUAL DIAGNOSTIC SLIDE SESSION 2024

CASE 2024-5

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Clinical History:
A young male child in his first decade, who had been adopted from India approximately three years prior to presentation, was admitted for evaluation of altered mental status and seizures. His development had been normal prior to this admission. His symptoms progressed over weeks to include hypertonia, ataxia, myoclonus, and autonomic storming. Neuroimaging showed progressive T2/FLAIR cortical abnormalities in the bilateral cerebral hemispheres.
The clinical differential diagnosis included infectious encephalitis, autoimmune encephalitis, vasculitis, or mitochondrial disease. CSF and blood cultures were negative. Autoimmune encephalitis and paraneoplastic panels were negative. He received multiple lines of therapy, including multiple rounds of IVIG, plasma exchange, and Rituximab, with no improvement. Given the lack of a definitive etiology and symptomatic progression despite treatment, a brain biopsy was performed. The biopsy was reviewed at the CDC, with negative antigen testing for Measles virus, Eastern equine encephalitis virus, West Nile virus, and LaCrosse encephalitis virus. Tissue from the biopsy specimen was then sent to UPMC.

Material submitted:
H&E section of cerebrum
MR images of brain (coronal T2, axial T2, axial DWI)

Points for discussion:
1. Diagnosis, including differential and workup
2. Pathogenesis