CASE 2004-8

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Clinical History: 50 year-old previously healthy Caucasian male who initially presented in the ER with difficulty mentating and focal facial twitching. Lumbar puncture showed CSF protein of 48 mg/dL, CSF glucose of 49 mg/dL (serum glucose 85 mg/dL), with 39 WBC/mm3, 94% lymphocytes. EEG was normal on two occasions. MRI of the brain was initially noncontributory; a second MRI showed progressive changes involving both the cortex and the white matter of the right temporal and frontal lobes. He also had right eye pain for > 1 year, which was diagnosed as episcleritis. He was treated with Tegretol for single partial seizures, and given a full course of Acyclovir for presumed viral encephalitis. After discharge, his family reported that his personality remained the same, he always felt fatigued, and although he improved slightly, he never returned to baseline. He presented for his second admission one month later with fever to 101º F, headache, right eye pain, and constant sleepiness. PE: Both eyes showed red sclerae, and no rashes or lesions were noted. He appeared lethargic and indifferent, and was oriented to name only. He followed simple commands, would perseverate, answering "JULY" to everything, and was unable to repeat. Pupils were equal and reactive to light, and extraocular movements were intact. There was bilateral papilledema, but no optic neuritis. Neurological exam: normal finger-to-nose and heel-to-shin, normal gait, negative Romberg's, normal pinprick and vibration. SH: Worked as a copier salesman, coached soccer in his community, Married with 2 children. Traveled to Mexico 1 year ago on a cruise ship, but never got off the ship. Lab: HIV negative.

Hospital Course: Acyclovir was re-started, despite two previous negative tests for HSV by PCR. TB tests were negative. EEG was abnormal, though non-focal. PET scan showed bilateral hippocampal enhancement. Chest and abdominal CT scans were negative. A brain biopsy of the left frontal lobe showed multiple lesions within brain parenchyma consisting of clusters of perivascular neutrophils and mature lymphocytes, macrophages, giant cells, and reactive astrocytes. Special stains showed no bacteria, fungi, mycobacteria, or parasites; electron microscopy was non-contributory. Immunohistochemical stains characterized the inflammatory cells as mature T-cells. The patient became more awake and alert after the institution of steroids. Several days following the biopsy, the patient was found unresponsive and failed efforts of resuscitation.
Necropsy findings: Massive pulmonary embolism. Gross examination of the brain showed features of cerebral edema, and no focal lesions. The histopathological process was diffuse, though most severe in the temporal lobes.

Material submitted: H&E section of cerebrum and one unstained slide.

Points for discussion: 1. Diagnosis
2. Pathogenesis