CASE 1990-3

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Clinical Abstract:

The patient was a 59-year-old white man, a NASA engineer, with a 3 month history of progressive neurologic deterioration. His initial symptoms were fatigue, balance problems, muscle cramping, myoclonus and spasticity of the lower extremities. He then developed increasing disorientation and lethargy along with declining cognitive function. His condition worsened despite some early mild improvement with steroid therapy.

He was transferred to Hermann Hospital in Houston, Texas on October 25 for further evaluation. Physical examination at this time revealed that he was oriented to person only and able to follow one-step, but not two-step, commands. Dysnomia, unclear speech, and poor memory were evident. The patient had several myoclonic jerks during the examination. His cranial nerves and motor strength were intact. His tendon reflexes were brisk and symmetrical with bilateral upgoing toes and no clonus.

Initial laboratory results were as follows: Hgb 16.4 g/dl, Hct 49%, WBC 15.7 cumm (88% polys, 9% lymphocytes, and 3% monocytes), platelets 251,000, LDH 302 U/L and electrolytes within normal range. Cultures of blood and CSF revealed no growth. Spinal fluid analysis revealed 10 WBCs, 110 RBCs, protein 107 mg/dl and glucose 77 mg/dl. No oligoclonal bands were present in the CSF. During his hospitalization the patient had a fluctuating mental status without notable improvement. A magnetic resonance imaging scan demonstrated multiple extensive abnormalities which were mostly present in the white matter and principally involved the frontoparietal area. A brain biopsy was performed on November 1.

Material submitted: One H & E and 1 unstained slides from the brain biopsy.

Points for Discussion: 1. Diagnosis
                2. Pathogenesis