Case 5

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Clinical Abstract:

This 25-year-old man with a history of alcohol and IV drug abuse and hepatitis developed peripheral neuropathy involving the right median nerve and the right common peroneal nerve at age 20. The etiology of these neuropathies was never identified. At age 23 he was hospitalized because of pain, tenderness and weakness in the calves and thighs, pretibial pitting edema and erythematous tender skin papules over the lower extremities. A muscle biopsy showed degeneration of muscle fibers with chronic lymphoid infiltrates without evidence of vasculitis or primary myositis. EMG was normal and FANA was negative. 3 months later the patient was hospitalized for ethanol abuse and fever of unknown etiology. 3 months later he was readmitted for status epilepticus and myoclonic jerks of his right arm. Examination at this time showed dementia, dysconjugate gaze, supranuclear ophthalmoplegia, persistent myoclonus of his right arm and obstructive pulmonary disease. Lumbar puncture showed pleocytosis (40-60 white cells, 40-90% lymphocytes). CSF glucose and protein were normal. Stains and cultures were negative. EEG showed diffuse slowing and brain stem auditory evoked responses were abnormal bilaterally. CT scan was normal. Three vessel angiography and brain biopsy were normal. Pulmonary function tests showed moderately severe obstructive disease. During the following months, the patient had periods of sudden obtundation followed several hours later by resumption of normal alertness. By December 1982, his pulmonary status had worsened and he had lost 60 pounds during the previous four months.

He was again admitted to the hospital at age 23 where he was noted to have a mild dementia, myoclonus of the right arm, volitional gaze palsies in all directions and normal doll's eye movements. General physical examination showed only expiratory wheezing. CT scan was normal. Lumbar puncture showed normal opening pressure (129 red cells, 6 white cells, glucose 71 gm.%, protein 41 mg.%). CSF gamma globulin 5.7 and VDRL negative. Cultures and stains for bacteria and fungus were negative. During his hospitalization, the patient had almost continuous myoclonic jerks. A cisternal tap showed no evidence of infection. A brain biopsy showed increased amounts of lipofuscin. No other abnormalities were noted. A two-week trial of antibiotics was given without clinical improvement. An open lung biopsy showed acute and chronic inflammation. Attempts at small bowel biopsy were unsuccessful. 3 months later had a respiratory arrest at the nursing home and had signs of anoxic encephalopathy and died 6 weeks later.

Autopsy Findings: The general autopsy findings showed myocarditis, bilateral bronchopneumonia, pulmonary emboli, emphysema, persistent hepatitis and focal interstitial nephritis. The brain weighed 1220 gms. and showed widespread narrowing of the cerebral cortex particularly in the frontal and temporal lobes. In some areas, the cortex was friable and discolored with slit-like cavities.

Material submitted: One H & E section of hippocampus
One unstained section of frontal lobe
One 2 x 2 Kodachrome

Points for discussion: 1. Diagnosis
2. Pathogenesis