# CASE 2000-6

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Clinical History:
This 82-year-old man was in his usual state of health presented with complaints of fatigue, periodic somnolence and loss of short-term memory. He had no other cognitive difficulties. Orientation, speaking, reading and writing were normal. He had no fever, headache or meningismus. He had no history of insect or animal bites. He had no risk factors for stroke and no history of previous stroke. The review of systems was noncontributory, except for nonprogressive chronic peripheral neuropathy. His neuropathy had been evaluated extensively in the past and not found to be due to paraneoplastic or rheumatologic etiologies. EMG showed sensory-motor neuronopathy. His past medical history was remarkable for hypertension, enterovesical fistula that was surgically repaired two years earlier, divcrticulosis, right ankle fracture with ORIF the same year and a remote appendectomy. His family history was negative for neurological disorders. He did not use drugs or alcohol. He had a 40 pack-year history of smoking, but had stopped 10 years earlier. He had no known allergies. On examination his MMSE was 35/38, with points lost for poor drawing of a cube and poor recall. The general medical and neurologic exams were normal except for loss of pinprick, light touch and temperature sense in a stocking and glove distribution.

An MRl showed bilateral, symmetrical temporal lobe hyperintensities. He was treated empirically for Herpes encephalitis, but was subsequently shown to be serologically negative for HSV. PCR was performed on CSF, and it too was negative for HSV 1 and 2. Other negative results included serologic tests for Eastern and Western equine encephalitis, St Louis encephalitis, TBc, mumps, measles, Rocky Mountain spotted fever, Ehrlichia Q fever, Mycoplasma pneumonia, brucella. Toxoplasma, Bartonella, CMV, HIV, EBV and Lyme disease. PCR of CSF was negative for Whipple disease. Serologic tests for anti-neuronal antibodies were negative. CT with contrast of chest and abdomen did not reveal any evidence of tumors. Routine laboratory studies showed no hematologic abnormalities.

He had steady progression of his neurological disorder. A brain biopsy was performed three months after presentation. Afterwards, high dose steroids and Cytoxan were administered, but he showed no signs of improvement. A repeat MRl one month later showed worsening of the temporal lobe lesions and extension of the process into both orbital frontal lobes. He was placed in hospice care two months later and died two months later with wasting, decubitus ulcers and pneumonia. A complete autopsy was performed.

Material submitted: H&E stained section of hippocampus

Points for discussion: Differential diagnosis; etiology