Case 7

Submitted by: Drs. Elizabeth Sengupta, Chinnamma Thomas and Roohi Shamsai.
Department of Pathology (Neuropathology)
Michael Reese Hospital and Medical Center
Chicago, Illinois 60616

Reference No.: A-63-85. West Suburban Hospital Medical Center, Oak Park, IL.

Clinical Abstract:

This 78 year old white female with no significant past medical history was admitted to the hospital for an approximate two month history of progressive neurological deterioration characterized by loss of memory, incontinence of urine, and gait disturbance. These problems were accompanied by generalized weakness associated with weight loss while maintaining a good appetite. She had an episode of falling with mild head trauma prior to the onset of the more recent problems. A previous neurologic work-up for clinically suspected Parkinson's disease revealed an abnormal EEG showing mild to moderate, generalized slowing with bihemispheric dysfunction and an essentially unremarkable CT scan with and without contrast. On admission the patient was afebrile and disoriented.

On physical examination, the patient was alert but disoriented to time and place. She had bilateral cataracts with mild right conjunctivitis. The mouth was free of periodontal disease. The neck was supple without lymphadenopathy. The lungs were clear. A Grade I systolic ejection murmur was heard at the left sternal border and apex. The abdomen revealed no organomegaly, masses, or focal tenderness. The extremities were within normal limits. The neurologic examination gave the impression of progressive dementia.

Pertinent laboratory data included a CBC revealing 6,700 WBC with 69% neutrophils and 4% eosinophils and a hemoglobin of 10.7 grams. The chemistry profile revealed no abnormal findings. Glucose was 113. Urinalysis was unremarkable, Sedimentation rate was 40. Rheumatoid factor, ANA, and RPR were negative. Folic acid and B-12 levels and thyroid functions were within normal limits. CT scan of the brain showed a focal, low attenuated area in the white matter of the left frontal lobe, possibly an old infarct. The chest x-ray and CT of the abdomen were unremarkable. A spinal tap showed a white cell count of 268 and a red cell count of 18. The white cell differential was 42% neutrophils, 47% lymphocytes, 1% eosinophils, and 10% mononuclear cells. The spinal fluid protein and glucose were 151 and 84, respectively.

The hospital course lasted about three weeks during which period the confusion and weight loss progressively increased. A temporal artery biopsy showed normal blood vessel. The patient was put on steroids. She died within two days of being transferred to a nursing home. The clinical diagnosis was Jakob-Creutzfeldt Disease.

Significant autopsy findings were restricted to the central nervous system. On external examination the brain showed no atrophy, but there was diffuse subarachnoid hemorrhage. The main cerebral and cerebellar arteries grossly had a beaded appearance with aneurysmal dilatations extending to the peripheral portions. The anterior portion of the Circle of Willis was adherent to the optic chiasm and the pituitary stalk and hypothalamus were partly necrotic. The meninges were slightly cloudy. Coronal sectioning revealed intraventricular hemorrhage and several areas of small, old infarction.

Material submitted: One H & E cross section of an intracranial artery.

Points for discussion:
1. Diagnosis.
2. Etiology of progressive dementia.