CASE 1993-7

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Clinical History: The patient was a 33 year old female who was diagnosed with AIDS after developing thrush and esophageal candidiasis. Her T-helper cells numbered 10 and she was found to be HIV positive. Three months later she presented with difficulty walking. She also complained of fatigue and shortness of breath. On exam she was found to have slightly slowed repetitive movements in her upper extremities. Her lower extremities displayed a mild spastic catch, and her gait was markedly ataxic. Laboratory evaluation revealed an acellular CSF with a protein of 20 and glucose of 41 (serum=87). Her CMV titer was positive and liver function tests elevated. An MRI of the head, cervical and thoracic spine were normal. A culture of bronchial biopsy material was positive for histoplasmosis. She was treated with amphotericin and responded well.

Her lower extremity ataxia progressed and truncal ataxia developed. By five months after presentation she was confined to a wheelchair. Attempts to move her upper extremities produced movements which were almost ballistic. Titubation was present as was mild dysarthria. Her eye movements and cognitive abilities continued to be normal. Anti-neuronal antibodies were not found. An MRI performed a month later revealed cerebellar atrophy and T2 prolongation in the middle cerebellar peduncles. By the next month, her speech was virtually uninterpretable. She developed saccadic intrusions and all movements became ataxic. Her cognitive ability appeared to remain intact for the remainder of her course; she was able to communicate with eye-blinks until her death. She died of aspiration pneumonia, 10 months after the development of her neurologic symptoms.

Necropsy findings: Autopsy was limited to the brain only. The fixed brain weighed 1240 grams. The cerebrum appeared normal; cerebellar atrophy was present. Histologically, the centrum semiovale displayed white matter vacuolation. Occasional microglial nodules and multinucleate giant cells were present, sometimes associated with each other. There was an extensive loss of neurons in the pontine nuclei associated with a reactive astrocytosis. Other features seen in the pons may be found in the submitted slide of cerebellum.

Material submitted:
1. H&E stained section of the cerebellum.
2. Kodachrome of cerebellar ultrastructure.

Points for discussion:
1. Diagnosis
2. Etiology of the cerebellar granular layer cell depletion.