CASE 5

Submitted by: Dr. Jose M. Bonnin, Department of Pathology, Methodist Hospital, Indianapolis, Indiana and Dr. Julio H. Garcia, Department of Pathology, University of Alabama at Birmingham, Birmingham, Alabama

Reference # 16912

A 33 year old black man with a history of paranoid schizophrenia was transferred from a state hospital for evaluation of a slowly progressive neurological deterioration. Two years earlier, he was found to have ataxia with mild proximal weakness, diffuse hyperreflexia and normal plantar response, stocking-glove sensory deficit for pain and a positive Romberg sign. There were no alterations in mental status or cranial nerve functions. Muscle tone was normal. There was no clinical evidence of a spinal cord lesion and, nerve conduction studies and a muscle biopsy were normal. A CT scan of the head showed minimal bifrontal atrophy and mild dilatation of the lateral ventricles.

Thirteen months earlier his condition suddenly deteriorated and became sleepy, confused and disoriented as well as hypotensive. This was thought to be a side effect of the antipsychotic medication which was adjusted. Over the following weeks his ataxia worsened and had several near-syncopal episodes. EEGs and a CT scan of the head as well as an extensive laboratory work-up did not provide any clues as to the cause of his condition. Over the ensuing months, he became increasingly irritable, confused and disoriented with slurring of speech, hypersomnia, increased use of profanity and inability to follow simple commands.

Five months earlier, a lumbar puncture yielded a fluid with a total protein of 167, glucose 40, IgG 30.4, IgG synthesis ratio 179 and Ig beta:gamma ratio of 0.2. VDRL and oligoclonal bands were both negative. An MRI scan disclosed small lesions in the left internal capsule and right basal ganglia. Visual and brain evoked potentials were abnormal. Multiple sclerosis was considered and a two-week ACTH trial was given with considerable improvement of his mental status, speech and gait. However, he soon returned to the previous status after cessation of therapy. One month prior to being admitted to the University of Alabama Hospital, he became comatose.

On admission, he was unresponsive with right preferential gaze and head turned to the right. Corneal and gag reflexes were present. He withdrew all extremities to noxious stimuli and had dicorticate posturing of the upper extremities. Tone was increased in upper extremities without cog wheeling or plastic component. Soon after admission the patient suffered a respiratory arrest. At the intensive care unit no electrical activity could be detected on EEG and there were no brain stem functions, clinically. After discussing with the family, all life support measures were discontinued.

The general autopsy revealed no gross abnormalities. The brain was of normal weight and the leptomeninges had a slight whitish discoloration. There were no focal lesions. The prosector did not remove the spinal cord.

Material submitted: One H&E stained and one unstained sections of the cerebrum.

Point for discussion: Diagnosis