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Case Reference No.: A191.98

Clinical History:

This 34 year old right handed male initially presented in the summer of 1991 with dragging of his left leg. By the summer of 1993 he had to use a cane to walk and was using a walker by July of 1994. In March of 1994 his speech was slurred, he had trouble with language formulation, as well as difficulty with math and calculations. His wife had noticed a change in his personality. In July of 1994 he suffered a seizure, lasting 60 seconds, during which he turned his head to the right side and his eyes deviated to the right. A brain biopsy was performed during this hospitalization which was interpreted as "consistent with leucodystrophy." In November of 1994 he had bilateral release signs, a right extensor plantar response, apraxia, and a slow shuffling gait. He also required burrhole drainage for a chronic subdural hematoma.

His family history is interesting. His father suffered from a similar neurological degenerative disease which was felt to be cortical basal ganglionic degeneration. No autopsy was performed. His mother suffers from depression.

Physical examination was performed on March 24,1998. He could comprehend simple commands but had inappropriate laughter possibly due to pseudobulbar palsy. He was dysarthric. Visual fields were intact and he had full horizontal pursuit movements of his eyes but inadequate vertical upward gaze. His eyes could be made to go upwards with turning of his head down and having him fixate on a pen. The fast phase of opticokinetic nystagmus was absent to the right and impaired on the left. There was a positive glabellar tap. The jaw jerk was brisk and there was spontaneous palatal elevation. Voluntary tongue movements were present but slowed especially on the horizontal plane. There was left hemiplegia with spasticity. A flexion contracture of the left upper limb was present . There was spasticity of the lower extremities with clonus of both ankles. There was antigravity power in the right lower limb to the hip. He could lift the right upper limb. He had lost individual fine finger movement. His tone was increased and he had dystonic posturing of his trunk.

He passed away on November 03, 1998. Autopsy was limited to brain only.

Necropsy Findings: Fresh brain weight, 1360 grams.
Material submitted: H&E section from frontal lobe
Points for discussion: Diagnosis