

CASE 10

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There is no record of the early history of this 28 year old white man, except that he was not able to sit up or to walk at the usual age. During his early years he was kept in various boarding homes. At the age of 7 he was admitted to a public school for children where he was found to be cognitively delayed and spastic ataxic, particularly in his legs. He learned to differentiate colors and to count a little, but could not learn to read. He showed good rote memory, but no judgment. He loved to sing and remembered the words of the songs. His attention span was short. At 10 years of age he was admitted to a state institution where the diagnosis idiopathic mental deficiency and spasticity was made. For about 14 years he was able to move about although with increasing difficulty. For the last 4 years of his life he was bedridden with frequent bouts of constipation, gastroenteritis and dehydration. Six months prior to death he became cyanotic and pulseless 30 minutes after a fecal impaction was removed, but soon recovered and continued to eat and sleep well until a few days prior to death when he developed a fatal bilateral bronchopneumonia.

Autops findings: Bilateral confluent bronchopneumonia and aspiration of food. All other internal organs were grossly and histologically normal, except for beginning post mortem changes. Brain weight: 1075 grams. Thickening of leptomeninges over the anterior portion of the cerebral convexity. Moderate degree of diffuse cerebral atrophy. Small brain stem and cerebellum. Normal optic nerves. No arteriosclerosis. On coronal sections, the main finding consisted of atrophy of entire cerebral white matter associated with gray scars radiating from the ventricle walls into the white matter of individual convolutions. There was atrophy of the corpus callosum, anterior commissure, and sclerosis of both thalami. No gross changes in cerebral cortex, including Ammon's horn, striate bodies, pallida and subthalamic nuclei, except for indistinct outlines of natural landmarks of pallida. The midbrain was small with shortening of ventro-dorsal axis and possibly sclerosis of peri-aqueductal gray. Small base of pons because of atrophy of fiber tracts. Diffuse scarring in white matter of cerebellum. Atrophy of pyramidal tracts in medulla oblongata.

Gross diagnosis: Atrophy of cerebral and cerebellar white matter and sclerosis of thalami, probably due to an episode of asphyxia at birth or sometime thereafter.

Hjstologic examination: Extensive glial scarring of cerebral and cerebellar white matter, and severe loss of neurons in both thalami. In addition, there were alterations of neurons and glia and interstitial accumulations of globular material as can be seen in the H & E slide submitted. These alterations were most severe in basal ganglia and thalami, and involved also cerebral and cerebellar cortex, midbrain and, less severely, pons and medulla oblongata.

Points for discussion: Diagnostic suggestions