

Case 4

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Clinical Abstract: The patient was a 45 year old woman with a 14 year history of progressive intellectual deterioration and ataxia following an episode of "encephalitis." She had been an honor student in art, history and anthropology in college. She had a history of drug abuse (LSD, marijuana and psilocybin) and of minor head trauma. Her father died at age 50 of a progressive dementing illness lasting 12 years that was clinically called "Pick's disease." An autopsy was not done.. A younger sister has also shown signs of dementia in the past three years.

Radiologic studies revealed an arachnoid cyst and diffuse atrophy of cerebrum and cerebellum. There were nonspecific EEG abnormalities and an increase in CSF lymphocytes on one occasion, but normal CSF protein. Brain biopsy 8 years prior to death showed nonspecific changes. All cultures and virological studies were negative.

Her course was characterized by global cognitive impairment, ataxia, seizures, myoclonus and lower extremity weakness. One year before she died her functional disability required skilled nursing care. Terminally, she was bedridden and mute, but reportedly alert and able to follow simple commands. Her lower extremities were flaccid, and her upper extremities were rigid.

Autopsy Findings: General autopsy disclosed bronchopneumonia with abscess formation, mild atherosclerotic vascular disease colloid cyst of the pituitary, and a fluid-filled arachnoid cyst in the left frontal operculum.

Material Submitted: One glass slide of basal ganglia stained with hematoxylin and eosin.

Points for Discussion:

1. What is the diagnosis?
2. What can be said about the familial nature of the condition; its clinical duration, and the localization of the pathological changes that are characteristic of this disease?