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CLINICAL HISTORY:

Four years before his death a 58-year-old man experienced the insidious onset of memory problems followed by declining language function and mild word finding difficulties with no verbal comprehension impairment. At age 56, he had lost 50% of his ability to solve crossword puzzles and showed socially inappropriate behavior. He treated a traffic red light as a stop sign and did not wait for the green light to proceed. He was able to perform his self-care activities and continued to work as an accountant with no difficulties, as well as running a separate company and managing all accountings. At the time, he was renovating his farmhouse and his skill as a remarkable woodcarver remained undiminished. He was an amateur marathon athlete and continued his daily training runs unimpeded. There was a strong family history of "Alzheimer's disease" affecting his mother, a maternal uncle, a maternal cousin aged 55 years and a 53-year-old brother. His 57-year-old sister had no cognitive complaints. His four children were healthy.

Examination at age 56 showed mild shorter memory deficit and mild fluent aphasia, the rest of the neurological examination of CNS and PNS was unremarkable. From then on the patient deteriorated fast; within 5 months he was mute but understood a lot and had become dependent for self care activities and inappropriate in his behavior. During the last year of his life the patient was confined to a chronic care facility and the nurse's notes indicated that towards the end, he had developed swallowing difficulties. He died of bronchopneumonia. The brain weighed 1360 grams and showed moderate fronto-temporal atrophy, slight ventricular dilatation and pallor of S.nigra.

MATERIAL SUBMITTED:

- 1. H&E stained section of either frontal lobe or temporal lobe
- 2. Unstained paraffin section of hippocampus

POINT FOR DISCUSSION: Diagnosis