59th ANNUAL DIAGNOSTIC SLIDE SESSION 2018.

CASE 2018-4a

Submitted by:

Anne Shepler, MD and Julia Kofler, MD
University of Pittsburgh Medical Center Presbyterian Hospital
Division of Neuropathology
200 Lothrop Street, Room S701 Scaife Hall
Pittsburgh, PA 15213

Clinical History:

The patient was a 37-year-old lawyer with no family history of dementia, who at age 33 began to have performance issues at work with inattentiveness to detail and a lack of concern for deadlines. He became progressively abulic and socially withdrawn with a loss of interest in his hobbies and personal appearance. By age 34, he began to choke frequently on food and was noted by his wife to have developed a nasal voice and bilateral ptosis. While he could remember his two young children's names, he was unable to care for them. He also developed reduplicative paramnesia wherein he insisted that there were two "2B" apartments in his apartment building where another woman (with his wife's name) lived with two children. While he did not get agitated if corrected, he returned to this delusion repeatedly.

On neurological examination, he showed no lateralizing sensory or motor signs and muscle strength was normal. No fasciculations were present. His reflexes were brisk and symmetric with no sustained clonus or Babinski sign. He was fluent with intact comprehension, repetition, naming, reading, and writing. He often answered questions too quickly with impulsive errors as he would not wait of the conclusion of the question; he was undisturbed when he made a wrong answer. Other than ptosis and a mild weakness of the orbicularis oculi, the cranial nerve examination was unremarkable. Initial MRI and CT scans were normal. A PET scan revealed bilateral frontal diminution of glucose utilization, worse on the right than the left, with extension to the right caudate. An EMG was negative.

Autopsy findings:

The brain weight was 1160 grams (fresh). Neuropathologic examination was notable for moderate atrophy of the frontal lobe and caudate and mild atrophy of the temporal and parietal lobes.

Material submitted:

One H&E slide of frontal cortex

Points for discussion:

1. Differential diagnosis
2. Useful immunohistochemical stains