Canadian Association of Neuropathologists
L'Association Canadienne des Neuropathologistes

CASE No 4

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The following history and clinical information is submitted for a woman who died at 57 years of age.

- 15-year history of arm myoclonus (left > right)
- 5-year history of progressive global cognitive decline with markedly impaired spatial/constructional abilities, word-finding difficulties, apraxia, and retrieval problems; MMSE score of 9/30 one year before death (18/30 the year prior); behavioral changes (decline in self-care and social withdrawal)
- 2-year history of gait problems with parkinsonian features (stooped, shuffling gait, postural instability, truncal rigidity) as well as lower extremity spasticity and hyperreflexia with extensor plantar responses bilaterally.

Family history was positive for dementia and gait problems affecting the patient's mother, maternal uncle and two maternal aunts.

Multiple investigations, including TSH, B12, VDRL, lactate, CSF studies, abdominal ultrasound, genetic testing for Huntington's disease, and SPECT scan were all nondiagnostic.

MRI scan was nondiagnostic but suggestive of small vessel ischemic changes, despite no risk factors, and generalized atrophy.

Material submitted: H&E stained section from frontal lobe.

Question: Diagnosis or Differential Diagnosis?