Canadian Association of Neuropathologists
L'Association Canadienne des Neuropathologistes

CASE No 12

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This 35 year old male presented with a progressive 15 month history of unsteadiness and slurred speech. He was previously healthy but his parents did note that he was a slow learner. He left school in grade 8 and worked for the past 17 years in Prince Edward Island in the lobster fishery. His family history was significant as his sister had complained of similar symptoms. She died at the age of 34 from a ruptured appendix before any diagnostic work-up of her neurological symptoms was undertaken. She had bilateral cataracts. His youngest brother who is age 26 is congenitally blind as a result of cataracts. There is an elder sister who is age 38 and is healthy. On physical examination he had marked dysmetria, past pointing and dysdiadochokinesis. He walked with a wide gait. He had brisk reflexes and sustained ankle clonus on the left more than the right. CT and MRI scan revealed bilateral symmetrical lesions of the cerebellar hemisphere related to the dentate nuclei. There were signal characteristics of the lesions that suggested evidence of calcification as well as hemorrhage. There were also white matter changes in the cerebrum and bilateral calcification in the globus pallidus. He was investigated for a primary progressive cerebellar disorder and because of his progressive symptoms a brain biopsy was suggested and performed by neurosurgery.

Materials Submitted:	One representative H & E slide from the cerebellar biopsy.
Questions:	Possible diagnosis? Investigations needed to establish this diagnosis?