

**Canadian Association of Neuropathologists
L'Association Canadienne des Neuropathologistes**

CASE No 10

A. Easton¹ and J. Moeller²

¹Department of Pathology, Division of Anatomic Pathology, ²Department of Medicine, Division of Neurology, Capital Health and Dalhousie University, Halifax, Nova Scotia, Canada

This 49 year old right handed man presented in December 1995 with a 2.5 year history of progressive imbalance and incoordination. There was a 3 year history of erectile dysfunction, with urinary difficulties and occasional fecal incontinence. His past medical history was non-contributory. He was a social drinker, and did not smoke and was not on any medications. His father had a history of falls and incoordination. He had 10 siblings who were in good health. He had a PhD in oceanography and had immigrated to Canada from Malaysia 10 years previously.

His general medical examination was normal. He had mild dysarthria but no nystagmus. He showed finger-to-nose and heel-shin dysmetria with a broad based ataxic gait. He had mild difficulty with clock drawing and word list generation on the mental status exam. MRI at the time showed atrophy of the cerebellum and brain stem.

Between 2000-2004 he required a walker and then a wheelchair. He developed urinary incontinence in 2002 and by 2004 he was using incontinence pads and found to be choking on his food. He developed an aspiration pneumonia in 2006 and required tracheostomy and insertion of a PEG tube. In 2008 he died following an episode of aspiration and an autopsy was requested.

Materials submitted: H&E/LFB slide of the lentiform nucleus

Question: Diagnosis?