

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

CASE No 9

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A 50 year old female without previous pertinent medical or surgical history, presented with a clinical subacute progressive neurologic syndrome evolving over a 4-5 year period. She initially presented with tremor, gait difficulty and disequilibrium along with motor deficits characterized by hyperreflexia, clonus and rigidity. Over the next few years she developed a frontal dementia with psychotic elements. Weeks before her death, she presented episodes of syncope and falls followed by seizures, culminating in a status epilepticus.

Laboratory investigation including CT-Scan and MRI were normal. A positron emission tomographic brain scan showed a slight bilateral frontal deficit. CSF analysis, revealed a slight increase in proteins, but was otherwise normal including search for oligoclonal bands, infectious work-up as well as 14-3-3 protein analysis. EEG showed a slight cortical dysfunction. Complete metabolic work-up was negative as well as heavy metal screening. Family history was negative for neurologic disease. Postulated clinical diagnosis was Creutzfeldt-Jacob disease.

Materials submitted: one representative H&E

Questions: Diagnosis?
 Postulated etiology?