Canadian Association of Neuropathologists
L'Association Canadienne des Neuropathologistes

CASE No 6

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Clinical Summary

An 84-year-old man had symptoms of subtle cognitive decline for approximately one year, followed by rapidly progressive confusion and hallucinations and incoordination with falls over the two to four weeks prior to hospital admission. He had a history of coronary artery disease, rheumatoid arthritis treated with methotrexate, spontaneous deep vein thrombosis with pulmonary embolus, and remote polio.

On admission he was cachectic, afebrile, alert, and oriented to time and person but not place. Neurological examination revealed dysdiadochokinesis, striking bilateral dysmetria in the arms and legs (more on the right) and an ataxic gait with marked truncal instability. He demonstrated myoclonus inconsistently to stimulation. Reflexes were preserved and plantar responses were both down-going.

Laboratory investigations; negative for ANNA-1, ANNA-2, and Purkinje cell antibodies. B12, TSH , and WBC count were normal. Hemoglobin was 111. Serum was negative for Lyme disease and HIV. CSF was negative for malignant cells and JC/BK virus, but was positive for 14-3-3 protein.

CSF glucose, protein, WBC count were within normal limits.

Brain CT scan showed diffuse brain atrophy from chronic ischemic changes, particularly in the right parietal-occipital white matter.

MRI revealed the old right occipital infarct and hyperintensities in the white matter of left cerebellar hemisphere. Twelve days later MRI showed worsening of the "vasogenic edema" in the left cerebellum. Both studies did not demonstrate gadolinium enhancement. A CT of his chest, abdomen and pelvis were noncontributory.

He continued to deteriorate and died at home with the diagnosis of possible Jacob-Creutzfeld disease.

Material Submitted: H&E section of cortex
Question: Diagnosis?