

CODE: TX200 M5078

ID NOS: A16926

DIAGNOSIS: BRAIN- PROGRESSIVE MULTIFOCAL LEUKOENCEPHALOPATHY

CLINICAL HISTORY: F/55. The patient presented with a 2 week history of neck stiffness and truncal rigidity, and was found to have a flaccid right hemiparesis and a resting tremor of both feet. There were no sensory changes detected. She had a 20 year history of schizophrenia and 2 year history of chronic lymphocytic leukemia. The patient became comatose, developed convulsions and died 6 weeks after admission.

MACROSCOPIC: Pale lesions up to 2mm diameter are present in the deep cortex (arrow), at the junction of grey and white matter and in the outer layers of the white matter. There were also lesions in the thalamus, corpus callosum and brain stem.

MICROSCOPIC: The sections show pale focal areas of demyelination, and there is a perivascular leukemic lymphocytic infiltrate (upper right image). The demyelinated foci contain pale macrophages with infected oligodendrocytes containing nuclear inclusions (left lower image, arrow). Electron microscopy of an oligodendrocyte shows round particles and filamentous forms of JC polyoma virus in the nucleus (bottom right image).

COMMENT: PML is a demyelinating disorder which usually occurs in immune-compromised patients, eg lymphoproliferative disease as in this case, in acquired immunodeficiency syndrome, and patients who have had immune-suppressive therapy. It is due to re-activation of latent JC neurotropic virus.

