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IMMUNOCOMPETENT PML—AN IMMUNOLOGICAL SCOTOMA?

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We describe two cases of progressive multifocal leucoencephalopathy (PML) presenting as stroke in the immunocompetent.

A 73-year-old man with a background of treated prostate carcinoma presented in October 2013 with a right hemiplegia; a diagnosis of lacunar infarction was made following CT. However, he progressed with increasing pyramidal weakness, ataxia, dysphasia and dysarthria. MRI in December revealed confluent T2 and FLAIR hyperintense white matter change with normal DWI. Extensive infective, vasculitic and malignancy screen were unremarkable; PCR in serum and CSF demonstrated JC virus DNA. Unfortunately the patient relentlessly deteriorated and died.

A 74-year-old lady presented via stroke services with a cerebellar syndrome. This progressed, and she developed emotional lability, seizures and myoclonus. MRI brain showed T2 and FLAIR hyperintensities without restricted diffusion, predominantly in the brainstem and posterior fossa. Extensive malignancy and infective screens were negative. Biopsy of the cerebellar lesion showed changes consistent with JC virus-induced demyelination. She improved spontaneously post-biopsy.

PML rarely occurs in immunocompetent patients; only a minority remit spontaneously. Patients may present with a focal deficit, followed by multi-focal progression. Thorough investigation for underlying immunosuppression is mandatory. There is a high risk of relapse, presumably due to an undetectable deficit in the cellular immune system.