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Progressive multifocal leukoencephalopathy presenting as a solitary gray matter lesion

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Sirs: Progressive multifocal leukoencephalopathy (PML) is a disease characterized by subacute progressive demyelination in the brain and is usually caused by an infection with the JC virus (JCV) [3, 10]. PML has mainly been found today in patients with the acquired immune deficiency syndrome (AIDS) [4, 5], and their average life expectancy is only 4 months [4]. PML may be the initial manifestation of AIDS or may develop during its course. The onset is insidious with focal neurological signs, intellectual impairment or epileptic seizures. Brain magnetic resonance imaging (MRI) and, to a lesser extent, computed tomography show white matter lesions and occasionally gray matter involvement [4, 7, 8, 12]. In cerebrospinal fluid (CSF) JCV DNA can be detected by the polymerase chain reaction (PCR) [2, 11]. There is no effective treatment; however, a few successful treatments with combined antiretroviral therapy [6, 9], cytarabine or zidovudine (AZT) have been reported [1]. Long-term prognosis nevertheless remains poor.

In PML, lesions most often involve the subcortical white matter. In addition, gray matter involvement may occur, but presentation of the disease with a solitary gray matter lesion is rare and not widely recognised. We report such a patient.

Fig. 1 Transverse T2-weighted MRI showing a solitary hyperintense lesion in the left thalamus

A 46-year-old man with AIDS was admitted because of clumsiness of his right arm and leg. He had a right-sided hemiparesis and sensory loss, brisk tendon reflexes, and an extensor plantar response. CT of the brain was normal. Two days later, brain MRI showed a solitary lesion in the left thalamus, which was hypointense on T1- and hyperintense on T2-weighted images but without mass effect or contrast enhancement (Fig. 1). Serological tests in blood and CSF for opportunistic infections and PCR for JCV were negative. After 5 weeks a stereotactic brain biopsy of the left thalamic lesion was performed. Histopathological examination of the biopsy tissue was consistent with PML. Despite treatment with intravenous cytarabine the patient died 3 months after onset of symptoms. Postmortem examination confirmed the diagnosis of PML in the left thalamus; lesions were also found in the border of gray and white matter of the cortex, in the brainstem and in the cerebellum.

The present patient to our knowledge is the first to be reported with

brain biopsy and autopsy confirmed PML, who presented with a solitary gray matter lesion without evident white matter involvement on brain MRI at the time of diagnosis. The lesion was located in the thalamus, a gray matter area, but the thalamus also contains myelinated fibres. PML is considered to be a disease that involves primarily the white matter of the brain, but there are occasional reports of patients with both white and gray matter involvement.

Recent reports of PCR of JCV DNA in CSF show promising results, with a high sensitivity and specificity [2, 11]. The finding of viral DNA in CSF may obviate the need for brain biopsy. However, this case illustrates that CSF findings can be negative, while PCR for JCV in brain biopsy tissue was positive. With respect to therapy there remains a need for an effective therapy for PML [4].

This case illustrates that PML should be considered in AIDS patients with focal neurological deficits and a solitary gray matter lesion on MRI who do not respond to an appropriate trial of empirical antitoxoplasma therapy. When PCR amplifications in CSF for JC viral DNA are negative, brain biopsy is indicated for definite diagnosis.

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