CLINICAL IMAGES

Progressive multifocal leucoencephalopathy – a case report

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Progressive multifocal leucoencephalopathy (PML) is a demyelinating disease caused by the human neurotropic JC (John Cunningham) virus, a polyomavirus.[1,2]

Following on the worldwide HIV/AIDS pandemic there has been a dramatic increase in the incidence of PML. However, cases of PML, an AIDS-defining illness, have rarely been reported from Africa, an area where HIV-1 clade C infection predominates.[3-5]

Clinical and imaging details

A 27-year-old woman presented to Chris Hani Baragwanath Hospital, Johannesburg, with new-onset seizures. She was heterosexual, did not abuse intravenous drugs, and was retroviral therapy-naïve. On examination in the medical ward she was found to be encephalopathic and pregnant. Further investigations revealed that she was HIV-positive with a very low CD4+ count of 7 cells/μl, and a viral load greater than 750 000 copies/ml. She was referred for imaging, and PML was diagnosed on the basis of computed tomography (CT) and magnetic resonance imaging (MRI) appearances.

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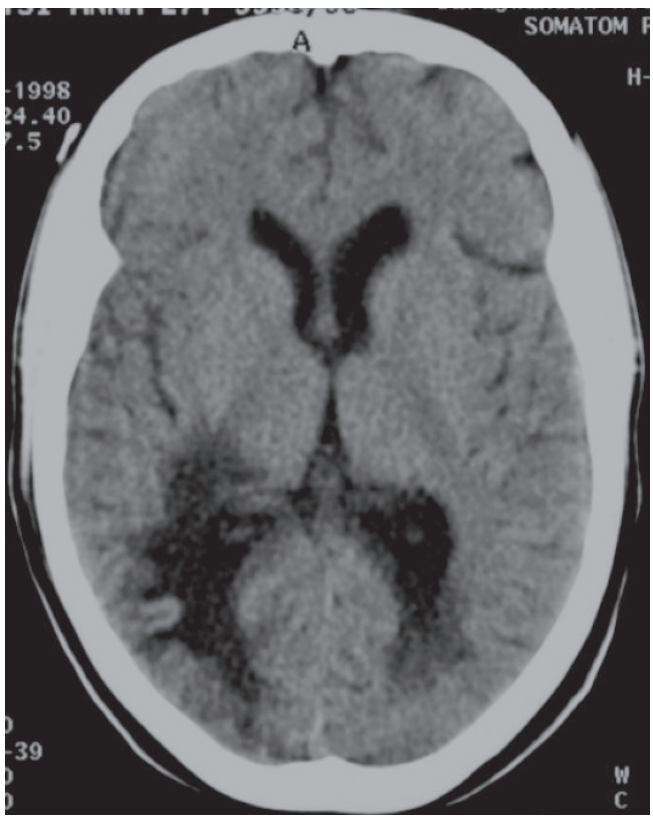
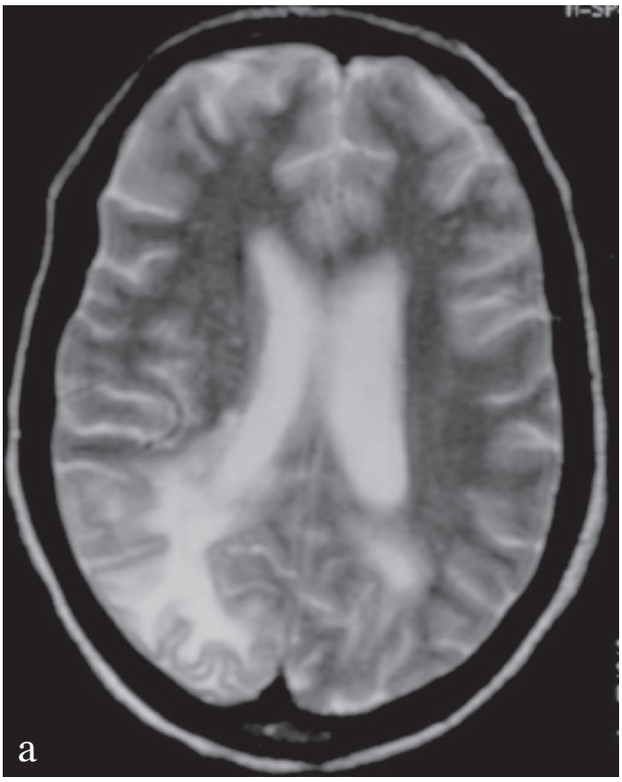


Fig. 1. Axial CT scan of the brain showing deep white-matter hypodensity involving predominantly the right parieto-occipital lobes. There is no mass effect.

The lesions on CT imaging were typical; they were scalloped in appearance, involved the white matter of the frontal, parietal



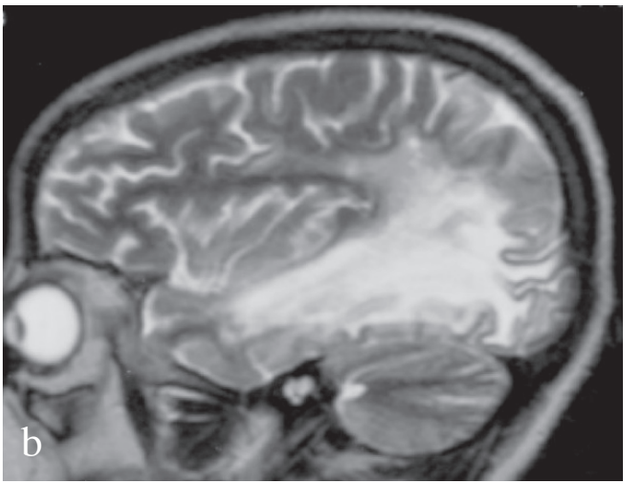


Fig. 2, a and b. T2 axial and sagittal MR images demonstrating extensive white-matter hyperintensities involving the frontal, parieto-occipital, and temporal lobes. There is background cerebral atrophy typical of advanced retroviral disease.

and occipital lobes, were hypodense, had no mass effect, and were non-enhancing (Fig. 1). On MRI the lesions were hypointense on T1 and hyperintense on T2-weighted MRI scans (Fig. 2, a and b).

The very low CD4+ count of 7 contributed to the diagnosis of PML. The diagnosis was confirmed on polymerase chain reaction (PCR) identification of the JC virus.

The patient died in hospital within a few days, and the diagnosis of PML was confirmed at autopsy.

Discussion

This is a rare case of autopsy-proven PML from a clade C HIV-1 infected area.[6] It is pertinent to highlight this demyelinating opportunistic viral infection as access to antiretroviral therapy is currently improving, and the immune reconstitution inflammatory syndrome (IRIS) that occasionally results following the initiation of highly active antiretroviral therapy (HAART) may in rare instances induce a paradoxical clinical deterioration in patients suffering from PML.[7] It has been documented that HAART-induced paradoxical aggravation of AIDS-related PML due to IRIS is reversible in most cases with the use of prolonged steroid therapy.[7,8]

Imaging can be particularly helpful when managing patients with PML and IRIS. PML lesions in patients not on HAART generally do not enhance, while in patients with IRIS contrast enhancement of the lesions may be seen.[7]

It therefore behoves us to be more vigilant when managing our patients with PML on HAART, as the early diagnosis and timely management of IRIS is critical.[8]

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