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Progressive multifocal leukoencephalopathy

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Abstract

Progressive multifocal leukoencephalopathy (PML) is a fulminating opportunistic infection of the brain that occurs in approximately 4% of AIDS patients. The need to diagnose and treat PML is urgent in patients with HIV as the infections are synergistic [Vazeux R, Cumont M, Girard P, et al. Severe encephalitis resulting from co-infection with HIV and JC virus. Neurology 1990;40:944–8]. Brain biopsy was previously required for definitive diagnosis. With positive JCV PCR CSF results and MR findings characteristic of PML, brain biopsy can be avoided in many AIDS patients [Weber T, Turner RW, Frye B, et al. Specific diagnosis of progressive multifocal leukoencephalopathy by polymerase chain reaction. J Infect Dis 1994;169:1138–41; Moret H, Guichad M, Matherson S, et al. Virological diagnosis of progressive multifocal leukoencephalopathy: detection of JC virus DNA in cerebrospinal fluid and brain tissue of AIDS patients. J Clin Microbiol 1993;10:3313; Post MJD, Yiannoutsos C, Simpson D, et al. Progressive multifocal leukoencephalopathy in AIDS: are there any MR findings useful to patient management and predictive of patient survival? AJNR Am J Neuroradiol 1989;20:1896–906]. A case is presented here and an overview of relevant literature is given.

Keywords: Progressive multifocal leukoencephalopathy; AIDS; Imaging

1. Introduction

Progressive multifocal leukoencephalopathy (PML) is a progressive demyelinating disorder resulting from CNS infection with the JC papova virus. More than 70% of all adults in the United States are infected with the JC virus, usually during early childhood. It usually lies latent in CNS, but in the setting of altered cellular immunity, it targets the oligodendrocytes and causes extensive myelin breakdown and white matter destruction. This includes people undergoing immune-suppressive chemotherapy for cancer, post transplant patients and people with damaged immune systems due to HIV. Prior to the use of combination anti-HIV drug therapy, it was estimated that between 3 and 10% of all people with AIDS developed PML. It usually occurs in people with very low T-cell counts (less than 100), but has been seen in some HIV-positive people with as many as 500 T-cells.

In the early years of the AIDS epidemic, PML was almost always progressive and fatal. Death usually occurred between 1 and 4 months after the first symptoms appeared. While a PML diagnosis today remains potentially rapid in its progression and possibly fatal, improvements in our ability to stabilize the immune system using anti-HIV drugs has helped to improve the prognosis associated with this opportunistic infection.

2. Case report

A 41-year-old Caucasian male presented with progressive expressive aphasia, dysarthria, forgetfulness over 3-week period. He also had right-sided weakness and unsteady gait. He was newly diagnosed with HIV and had a CD 4 count of 176 at presentation. He admitted to being homosexual.

MRI head showed multiple foci of hypointensity on T1 and hyperintensity on T2 and FLAIR throughout the sub-cortical white matter of bilateral frontal, temporal and parietal lobes. Similar lesions were seen in left-pons and right-cerebellar hemisphere. There was no significant mass effect associated

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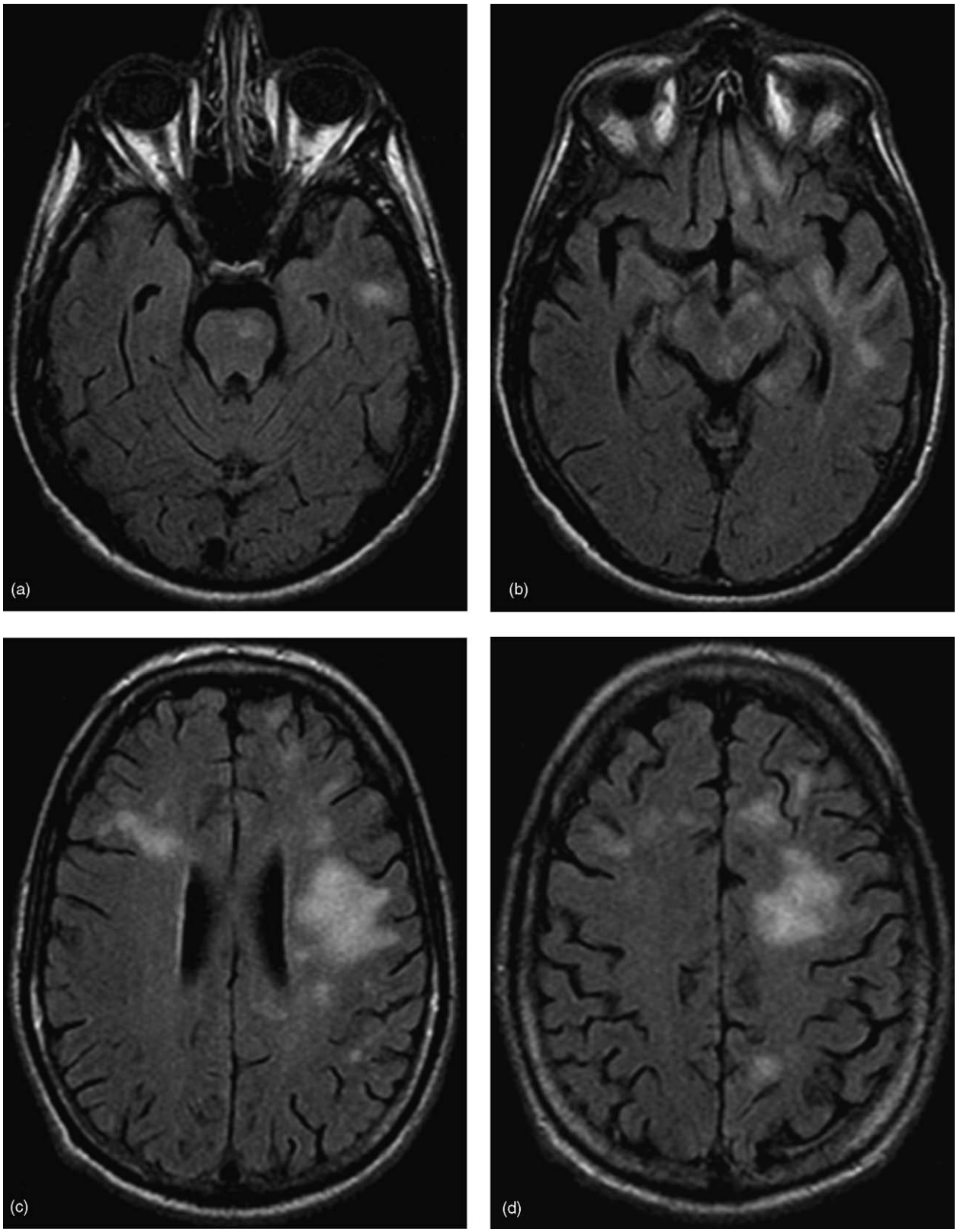

Fig. 1. (a–f) T2 W and FLAIR images—multiple foci of hyperintensity on T2 and FLAIR in the sub-cortical white matter and left pons. No significant mass effect on adjacent sulci. Involvement of sub-cortical U-fibers gives a “scalloped” appearance to the lateral margins of the lesions.