Journal of Infection (2007) 54, e175—e178

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CASE REPORT

Fatal cerebral lymphomatoid granulomatosis in an HIV-1-infected patient

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Accepted 1 November 2006
Available online 12 December 2006

KEYWORDS
LYG;
Lymphomatoid granulomatosis;
HIV-infection

Summary Lymphomatoid granulomatosis (LYG) is a rare systematic lymphoproliferative disorder of uncertain etiology. We present a rare case of isolated cerebral LYG developing in an HIV-1-infected patient after initiation of highly active antiretroviral therapy (HAART).
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Introduction

Lymphomatoid granulomatosis (LYG), first described in 1972 by Liebow et al.[1] is an angiocentric and angiodestructive lymphoreticular proliferation. Although the lungs are the most common site of involvement, other organs such as the skin, kidneys, liver and the central nervous system (CNS) can be affected. The pathogenesis of LYG is unknown; however, recent studies have demonstrated that immunologic dysfunction and Epstein—Barr-virus (EBV)-associated B-cell transformation seem to play an important pathogenetic role. Currently LYG is considered as a B-cell lymphoma associated with a pronounced, benign, T-cell reaction.[2,3] Histopathological studies have demonstrated the presence of Epstein—Barr virus RNA within malignant B-cells in most cases and EBV and/or HIV have been discussed as an etiological factor for LYG.[4,5] Although LYG predominantly occurs in immunocompetent patients, there are numerous reports of LYG in patients with various forms of immune dysfunction, including HIV-infection.[6—8] The natural course of LYG may be variable and spontaneous remissions have been reported.[9] However, in most cases, the disease is progressive, with a 5-year mortality rate of 60—90%.[9] Although a variety of chemotherapeutic regimens have been utilized, there is no standard treatment for LYG. Experimental treatment approaches include interferon alfa-2b, ganciclovir or anti-CD20 immunotherapy.[10]

Here we report a rare case of isolated cerebral LYG associated with HIV-1-infection. To our knowledge this is

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doi:10.1016/j.jinf.2006.11.003

the first case of cerebral LYG presenting in the context of immune reconstitution after initiation of an effective HAART.

Case report

A 37-year-old intravenous drug user presented with a pneumocystis pneumonia in 1996. At that time, a severe immunodeficiency (0 CD4 cells/μl) and co-infection with HIV-1 and hepatitis C virus (HCV) was diagnosed. Mainly due to adherence problems, various antiretroviral drug combinations failed to achieve a sustained virological and immunological response. Consequently, the patient developed several AIDS-defining illnesses during the following years, including *Mycobacterium avium* complex infection, pulmonary tuberculosis and recurrent esophageal candidiasis.

In February 2002, a new HAART regimen containing boosted lopinavir, indinavir, lamivudine and tenofovir was started. During the following weeks, the absolute CD4 cell count increased from 30 (relative 4%, CD4/CD8 ratio 0.1) to 110 cells/μl (11%, ratio 0.2) and the HIV plasma viremia decreased to below 50 copies/ml for the first time for 2 years. HCV-RNA PCR was 1,800,000 IU/l. In April 2002, the patient complained of severe recurring headache, nausea, dizziness and increasing weakness. On admission to our hospital, the general physical and neurological examination was normal. Computed tomography and T1 and T2 weighted magnetic resonance imaging (MRI) scans of the brain revealed distinct symmetric lesions in the basal ganglia with diffuse contrast medium enhancement and perifocal edema (Fig. 1A and B). Cerebro-spinal fluid (CSF) showed mild pleocytosis (11 WBC cu/mm, normal range 0—10), an increase of protein of 2.3 g/l (0.2—0.4 g/l) and lactate of 3.1 (<2.2 mmol/l). There was no evidence of malignant cells or of infectious agents in the CSF, including EBV, cytomegalovirus (CMV), herpes simplex virus, variella zoster virus, JC virus, enterovirus-, HIV-1, *Mycobacterium tuberculosis*, *Cryptococcus neoformans* and *Toxoplasma gondii*). Routine peripheral blood tests exhibited mild eosinophilia of 8%, but no other abnormalities were present. Biochemical and serological peripheral blood tests were consistently normal (negative serology for *Treponema pallidum*, *Toxoplasma gondii*, *Bartonella Henselae*, CMV, *Cryptoccocus neoformans* and *Histoplasma capsulatum*), as were CT scans of the thorax and abdomen. Because neurological symptoms progressed, a stereotactic brain biopsy of the distinct lesions in the right basal ganglia was performed. Histological examination revealed diffuse mixed-cellular leukocytic infiltrations dissociating vessel wall structures without any evidence of a causative organism, atypic lymphoid cells or LYG (Fig. 2A). Despite the administration of numerous antibiotic regimens including therapy for cerebral toxoplasmosis the patient's condition continued to deteriorate. Six months after onset of symptoms, the patient died of epileptic status.

Macroscopic examination at autopsy showed a massively swollen edematous brain (1750 g) with signs of herniation of the cerebellar tonsils and fatal compression of the brain stem into the foramen magnum. On coronal sections bilateral symmetric dense masses localized in the basal ganglia were discovered. Microscopic examination of the brain demonstrated leukocytes within small vessels infiltrating the adjacent brain parenchyma and fibrinoid necrosis of vessels, consistent with the diagnosis of an angiocentric and angiodestructive LYG, in the present case located bilaterally in the basal ganglia (Fig. 2B and C). Furthermore a polymorphous leucocytic, angiocentric, and angiodestructive cellular infiltrate with small foci of necrosis was found (Fig. 2C). Immunohistochemistry revealed predominantly CD45+ leucocytes (Fig. 2D), consisting of predominantly CD3+ T cells, few CD20 B-cells with a blastic morphology, and some CD138+ plasma cells (Fig. 2E). EBV RNA was

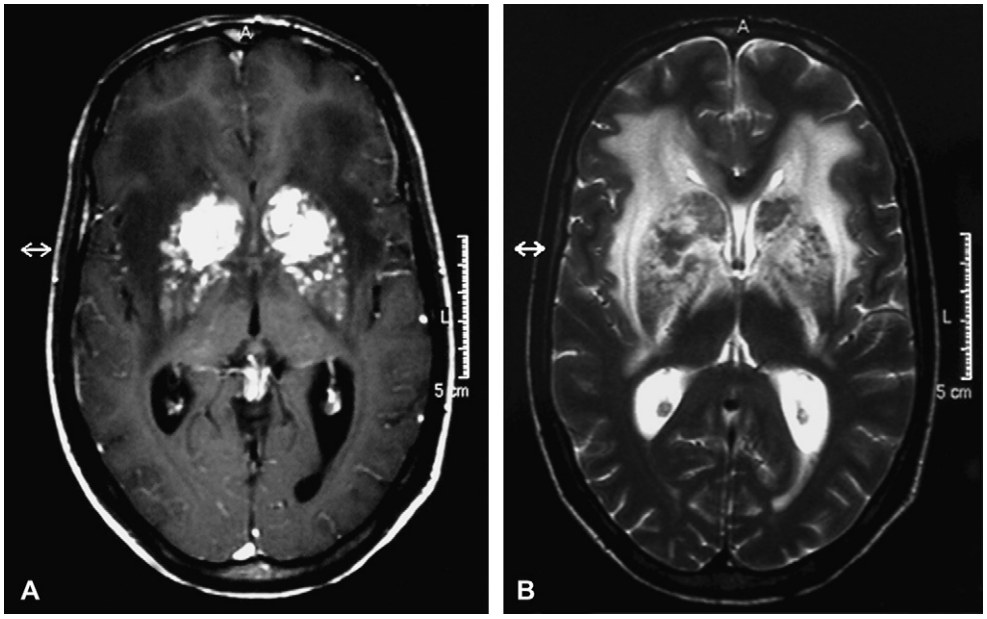

Figure 1 MRI scans. (A) Distinct symmetric hyperintense lesions in the basal ganglia on the T1 weighted MRI scan. (B) Diffuse contrast medium enhancement in the basal ganglia and perifocal edema on the T2 weighted MRI scan.

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