AIDS-Related Primary Central Nervous System Lymphoma: Prolonged Remission Associated with Highly Active Antiretroviral Therapy

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Abstract

A 36-year-old HIV-seropositive man developed progressive confusion and unilateral tremor of the hand. His medical history included cryptococcal meningitis and CMV colitis. CT scan revealed a single hyperdense mass with minimal peripheral enhancement at the region of the cerebral peduncle and pons, causing obstructive hydrocephalus. He was treated with ventriculo-peritoneal shunt and cranial radiotherapy. He also received treatment with highly active antiretroviral therapy (HAART). A CD4+ cell count was increased from 2 to 345 cells/mm^3. He returned to normal function for about 32 months after treatment.

Key word : AIDS, Primary Central Nervous System Lymphoma, HAART

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Primary central nervous system lymphoma (PCNSL) is a non-Hodgkin's lymphoma restricted to the nervous system. It is recognized as one of the criteria of AIDS and usually occurs with advanced HIV infection. It is a neoplasm with a very poor prognosis. The mean survival time is 42 (range, 8-127) days without therapy and up to 134 (range, 33-380) days with radiation therapy(1). The outcome of using highly active antiretroviral therapy (HAART) has demonstrated reduction in HIV-related disease progression and mortality. To our knowledge, there has only been one report on an AIDS-patient with long-term remission of PCNSL after treatment with HAART(2). The authors, herein reported the second patient with this clinical setting.

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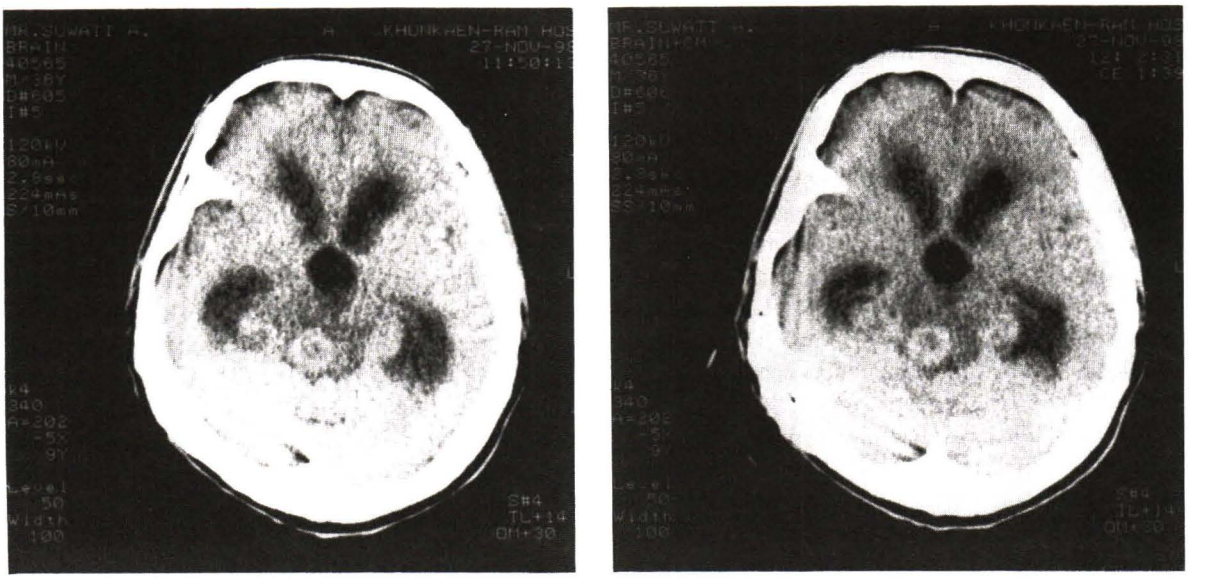

Plain CT

With contrast enhancement

Fig. 1. Computerized tomopraphy scans of the head with contrast enhancement revealed a 1.5 cm ring-like hyperdense lesion and surrounding edema with minimal peripheral enhancement.

CASE REPORT

A 36-year-old HIV-seropositive man was admitted on 30 October 1998 for treatment of a large bed sore. His medical history included cryptococcal meningitis in 1995 and cytomegalovirus colitis in August 1998. He had a CD4+ cell count of 2 cells/mm^3. The antiretroviral regimen (stavudine, lamivudine and indinavir) was started on 1 September 1998. On 24 November 1998, he developed progressive confusion and tremor of the right hand. Computed tomography (CT) scan of the head demonstrated a single 1.5 cm ring-like hyperdense lesion with minimal peripheral enhancement at the region of the right cerebral peduncle and pons, causing obstructive hydrocephalus (Fig. 1). Toxoplasmic serology was negative for immunoglobulin G antibody. Ventriculo-peritoneal shunt was performed. CSF analysis was within normal limits and cytological study of the CSF was negative for malignancy cell. A CD4+ cell count was 85 cells/mm^3. PCNSL was presumably diagnosed. The patient was treated with 4,040 rad of whole-brain radiation. He did not receive chemotherapy or corticosteroid treatment. His condition markedly improved. A follow-up CT scan on 28 January 1999 revealed a decrease in size of the lesion (Fig. 2). He returned to work. On 26

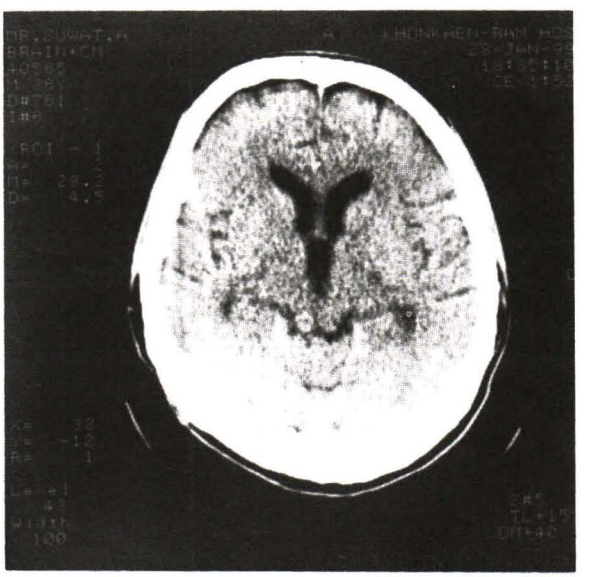

Fig. 2. Post RT: showing a marked resolution of the lesion.

December 1999, a CD4+ cell count was 345 cells/mm^3. The patient remained asymptomatic for 32 months after treatment.

DISCUSSION

PCNSL, which has a strong association with Epstein-Barr virus (EBV), is second in frequency only to toxoplasmosis among CNS mass lesions in AIDS-patients. Patients with PCNSL usually have a history of one or more other AIDS-related disorders before the diagnosis of lymphoma. These include *Pneumocystis carinii* pneumonia, candida esophagitis, CMV retinitis, toxoplasmosis, *Mycobacterium avium-intracellulare* infection, cryptococcal meningitis and histoplasmosis [1,3]. The diagnosis strategies for PCNSL include CT scan, MRI and brain biopsy. CT/MR scan usually demonstrates single or multiple contrast enhancing masses that are radiographically indistinguishable from other processes such as toxoplasmosis. A periventricular lesion, a focal enhancing mass with subependymal spread on CT or MR images and hyperattenuation at nonenhanced CT are the most reliable features in distinguishing between PCNSL and toxoplasmosis[4]. Cranial radiotherapy (RT) is the cornerstone of therapy and produces responses in most patients. However, patients who successfully complete whole-brain RT usually die from systemic opportunistic infection[1]. EBV is present within AIDS-related PCNSL cells. EBV-infected B cells can undergo monoclonal in the presence of immune dysregulation as seen with AIDS and reduction of CD4+ cell count may allow EBV-induced B-cell proliferation. The significant and sustained increase in CD4+ cell count may have restored the ability of the immune system to suppress EBV-immortalized B cells in the CNS, and produce durable remission[2].

In the presented patient, PCNSL could be diagnosed from the clinical presentation, CT scan finding and good response to RT. The report demonstrates the benefit of HAART to produce long-term remission of PCNSL in an AIDS-patient.

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มะเร็งต่อมน้ำเหลืองของระบบประสาทส่วนกลางชนิดปฐมภูมิที่สัมพันธ์กับโรคเอดส์: การสงบของโรคเป็นระยะเวลานานจากการรักษาด้วยยาต้านไวรัสเอ็ชไอวีที่มีประสิทธิภาพสูง

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รายงานผู้ป่วยเอดส์ชายอายุ 36 ปี มีอาการสับสนและมือข้างขวาสั่น ผู้ป่วยมีประวัติเป็นโรคเยื่อหุ้มสมองอักเสบจากเชื้อราคริพโตคอคคัสและลำไส้ใหญ่อักเสบจากเชื้อ CMV การตรวจคอมพิวเตอร์สแกนของสมองพบก้อนมีลักษณะ hyperdense 1 ก้อนที่ทำให้เกิดการอุดตันของทางเดินน้ำไขสันหลัง ผู้ป่วยได้รับการรักษาด้วยการผ่าตัดใส่ท่อระบายน้ำไขสันหลังและการฉายรังสีรักษา และได้รับยาต้านไวรัสเอ็ชไอวีที่มีประสิทธิภาพสูง ผู้ป่วยมีอาการดีขึ้นมาก การติดตามผลการรักษา พบว่าจำนวนเม็ดเลือดขาว CD4 มีจำนวนสูงขึ้นมาก และผู้ป่วยยังมีสุขภาพดีเป็นระยะเวลานานประมาณ 32 เดือนหลังการรักษา

คำสำคัญ : เอดส์, มะเร็งต่อมน้ำเหลืองของระบบประสาทส่วนกลางชนิดปฐมภูมิ, ยาต้านไวรัสเอ็ชไอวีที่มีประสิทธิภาพสูง

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