Progressive Multifocal Leukoencephalopathy in a Patient with Hepatosplenic T Cell Lymphoma

J. González de la Aleja a, E. Giménez-Mesa b, I.J. Posada a, J.F. Gonzalo-Martínez a, R. Bustelo b, L. Escudero c

Servicios de a Neurología, b Hematología, and c Radiología, Hospital Universitario 12 de Octubre, Madrid, Spain

Dear Sir,

Progressive multifocal leukoencephalopathy (PML) is a fatal demyelinating disease of the central nervous system caused by the JC polyomavirus. The diagnosis is made on the basis of clinical and neuroradiological criteria with a positive JC virus DNA polymerase chain reaction (PCR) in the cerebrospinal fluid or by means of histopathological characteristics.

We report the first case, to our knowledge, of PML associated with hepatosplenic T cell lymphoma.

In April 2003, a 56-year-old HIV-negative male was diagnosed with hepatosplenic γδ T cell lymphoma. The patient underwent standard chemotherapy with cyclophosphamide, doxorubicine, vincristine and prednisone (CHOP). He failed multiple conventional therapies aimed at controlling his disease, including pentostatine, etoposide, methylprednisolone, cytarabine and cisplatin (ESHAP) and methotrexate, doxorubicin, cyclophosphamide, vincristine, prednisone, and bleomycin (MACOP-B).

In August 2004, the patient was admitted to the hospital because of a rapidly progressive speech disorder. He was afibrile. Neurological examination showed a normal mental status and occasional word-finding difficulties with semantic and paraphasic errors. Cranial magnetic resonance imaging revealed nonenhancing lesion in the left insula and temporal lobe subcortical white matter, without mass effect on T1-weighted images. The lesion was hyperintense on T2-weighted and on fluid-attenuated inversion recovery magnetic resonance images (fig. 1). Cerebrospinal fluid protein, glucose and cell values were all within the normal ranges. PCR analysis of the cerebrospinal fluid was positive for JC virus DNA. The diagnosis of PML was established. Because of the advanced state of the hematological malignancy, no empirical treatment was applied. The patient died 6 weeks after the onset of his neurological symptoms. The family denied permission for autopsy.

Fig. 1. Hyperintense lesions are seen in both temporal lobes in a fluid-attenuated inversion recovery image. The temporal left lesion is not associated with a mass effect and spares the cortex.

PML was described in 1958 as a rare, fatal complication occurring in the setting of chronic lymphocytic leukemia. With the AIDS epidemic, PML has become one of the deadliest opportunistic infections, strongly correlated with depressed CD4+ lymphocyte counts. However, it is also a rare complication in patients with other immunosuppressive disorders, such as chronic lymphocytic leukemia, follicular non-Hodgkin’s lymphoma, in recipients of renal, heart and lung allografts, and allogenic bone marrow transplants due to the pharmacological immunosuppression state.

T cell lymphoma is a lymphoproliferative disorder that causes a profound immunosuppression. To the best of our knowledge, this is the first published case of PML, confirmed by JC virus PCR, complicating hepatosplenic T cell lymphoma. The reported patient was treated with different chemotherapeutic agents including pentostatine.

When using agents with effects on T-cell-mediated immunity, we must be aware of the risk of viral opportunistic infections without effective treatment or prophylaxis.
PML in a Patient with Hepatosplenic T Cell Lymphoma

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References