

A Case of Posttransplant Acute Limbic Encephalitis Associated with Human Herpesvirus-6

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OBJECTIVES

Human herpesvirus-6 (HHV-6) is a β -herpesvirus isolated especially from patients with hematological malignancies. It may reactivate with immunosuppression and cause central nervous system dysfunctions. Patients undergoing allogeneic hematopoietic stem cell transplantation and bone marrow transplantation are at risk of posttransplant acute limbic encephalitis. A posttransplant acute limbic encephalitis syndrome is often characterized by anterograde amnesia, seizure activity, and enhancement in the uncus, amygdala, or hippocampal areas on magnetic resonance imaging. We aimed to present a case who was diagnosed with standard risk B-cell ALL and underwent bone marrow transplantation and then developed posttransplant acute limbic encephalitis associated with HHV-6.

CASE PRESENTATION

A 7-year-old boy who was followed up with ALL for 3 years and was on the 78th day after bone marrow transplantation, was consulted to our pediatric neurology department with sudden onset headache, urinary incontinence, ataxia, agitation, and confusion. Neurological examination revealed abnormally brisk deep-tendon reflexes, ataxic gait, and clumsiness in cerebellar tests. Etiological workup was found to be normal for underlying infectious, autoimmune and metabolic causes, except positive HHV-6 PCR in cerebrospinal fluid (CSF) samples. Moreover, CSF IgG index was borderline high. Cranial MRI showed bilateral T2 and FLAIR hyperintensities in the frontotemporal lobe and insular cortex. He was treated with various intravenous antibiotic and antiviral therapies, however, no significant clinical improvement was observed. Intravenous immunoglobulin (2 gr/kg, 5 days), therapeutic plasma exchange and intravenous methylprednisolone (30 mg/kg/day, 5 days) were performed. On the 7th day of immunotherapy, his level of consciousness gradually improved.

RESULTS

HHV-6 shows neurotropism and is a rare cause of encephalitis predominantly seen in immunocompromised and occasionally immunocompetent patients. Immunosuppression secondary to solid organ or hematopoietic transplantation may permit reactivation of HHV-6, viral replication and the emergence of a HHV-6 related clinical syndrome. Limbic encephalitis is a potentially treatable immunological condition. The diagnosis of HHV6-associated posttransplant acute limbic encephalitis is based on suggestive clinical and radiological findings along with positive blood/CSF PCR for HHV6. Clinical and laboratory features of this syndrome have not been well characterized. Herein, we determined that the limbic encephalitis that develops after bone marrow transplantation may develop due to HHV-6. One of the possible causes of acute limbic encephalitis may be HHV-6 infection after bone marrow transplantation.

CONCLUSIONS

Patients undergoing bone marrow transplantation are at risk for posttransplant acute limbic encephalitis. Posttransplant acute limbic encephalitis associated with HHV-6 should be kept in mind if radiological findings are compatible in patients presenting with acute onset of alteration of consciousness and seizures after bone marrow transplantation.

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