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Case Report

Human herpesvirus-6 infection-associated acute encephalopathy without skin rash

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Abstract

Background: Human herpesvirus-6 (HHV-6) is the etiological agent of exanthema subitum-associated encephalopathy, which usually occurs in children younger than 3 years. Brain imaging shows various abnormalities.

Patient: A previously healthy 4-year-old girl developed acute encephalopathy with clinical features consisting of fever, repetitive seizures, and a disturbance of consciousness. The patient did not show skin rash suggestive of exanthema subitum during the course of her illness. The primary HHV-6 infection was diagnosed based on the absence of IgG against HHV-6 and identification of the virus DNA in the acute phase serum and a significant increase of the anti-HHV-6 IgG titers in the convalescent phase sera. Diffusion-weighted images showed transient high signal intensity in the bilateral periventricular white matter and splenium of the corpus callosum and in the gray matter structures such as the bilateral basal ganglia and thalami. Upon therapy with steroid and γ-globulin, the patient recovered without any neurological deficits.

Conclusion: Primary HHV-6 infection can cause acute encephalopathy without exanthema subitum. The etiological diagnosis is possible only by examining the blood and cerebrospinal fluid, when the patient shows no skin rash. This condition should be included in the differential diagnosis of acute encephalopathy even in patients older than 3 years.
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Keywords: Diffusion-weighted imaging; Encephalitis; Encephalopathy; Exanthema subitum; Human herpesvirus-6; Magnetic resonance imaging; Periventricular white matter

1. Introduction

Human herpesvirus-6 (HHV-6) is the etiologic agent of exanthema subitum, which is clinically characterized by a transient rash that develops following a fever for about 3 days [1]. Although the disease is usually self-limiting, several severe manifestations, particularly in the central nervous system (CNS), can occur [2]. The prognosis of exanthema subitum-associated encephalopathy is poor and nearly half the patients manifest neurologic sequelae [2]. A nationwide survey of acute encephalopathy in Japan revealed that the pathogens remained unidentified in approximately 40% of the cases [3]. We report HHV-6 identification by a multivirus real-time PCR [4] using the acute phase serum of a patient with acute encephalopathy, who did not present with exanthema subitum. Our case suggests that HHV-6

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may be associated with etiologically unidentified acute encephalopathy in patients without skin involvement.

2. Case report

A 4-year-old Japanese girl was admitted to a local hospital with fever, seizures, and a disturbance of consciousness (Fig. 1). She had been well until 3 days before admission when high fever developed (day 0). After admission, she developed repetitive generalized tonic seizures despite treatment with diazepam and midazolam. On day 4, she was transferred to our hospital. She was comatose, with a body temperature of 39.6 °C and Glasgow Coma Scale of E1V1M3. The extremities were flaccid without spontaneous movements. She had a positive Babinski reflex. Her deep tendon reflexes were normal. Hepatosplenomegaly, lymphadenopathy, and skin rash were not present. The initial hematological and chemical work-up was normal. Analysis of the cerebrospinal fluid (CSF) revealed 1 mononuclear cell/mm^3; protein level of 21 mg/dL; glucose concentration of 78 mg/dL; and no elevation of myelin basic protein (MBP) level (<40 mg/dL). The first magnetic resonance imaging (MRI), performed on day 4, revealed abnormally high signal intensity in the bilateral periventricular white matter and splenium of the corpus callosum on T2-weighted (T2WI) and fluid-attenuated inversion-recovery (FLAIR) images. These lesions exhibited an increased signal on the diffusion-weighted image (DWI) with a marked decrease in the apparent diffusion coefficient (ADC) (Fig. 2). Electroencephalogram (EEG) showed diffuse high-voltage slow waves but no paroxysmal waves. Based on these results, the patient was diagnosed with acute encephalopathy.

The patient was administered a treatment of γ-globulin (1 g/kg for 1 day) and high-dosage corticosteroid (methylprednisolone 30 mg/kg/day for 3 consecutive days). Acyclovir was started until cultures and PCR results for herpes simplex virus were negative. She partially responded to the therapy: she opened her eyes but her voluntary movements and speech remained to be impaired. After reinitiating the 3-day regimen of intravenous methylpredonisolone on day 11, she showed a marked improvement in alertness and speech with no further seizures. On being discharged from our hospital nearly 1 month after the onset of the illness, she could walk unaided and speak two-word sentence. Cognitive assessment using the Kyoto Scale of Psychological Development 2001 revealed the borderline developmental quotient (DQ) with cognitive–adaptive DQ 79 and language-social DQ 85.

Serial MRI examinations were performed on day 10, 18, and 38 after the first imaging on day 4 (Fig. 1). On day 10 and 18, DWI demonstrated high signal intensity in the bilateral periventricular white matter and splenium of the corpus callosum and in the gray matter structures such as the bilateral basal ganglia and thalami with sparing of the cerebral cortex and the adjacent subcortical white matter. No abnormal enhancement was observed in the affected regions on contrast-enhanced T1WI. On day 38, the abnormal high signal intensity on DWI had resolved but diffuse cerebral atrophy became pronounced with the cystic lesions in the periventricular white matter.

To elucidate the cause of encephalopathy, we performed multivirus real-time PCR, which can detect 163 human viruses simultaneously [4]. HHV-6 variant B DNA was detected in the acute phase serum, but

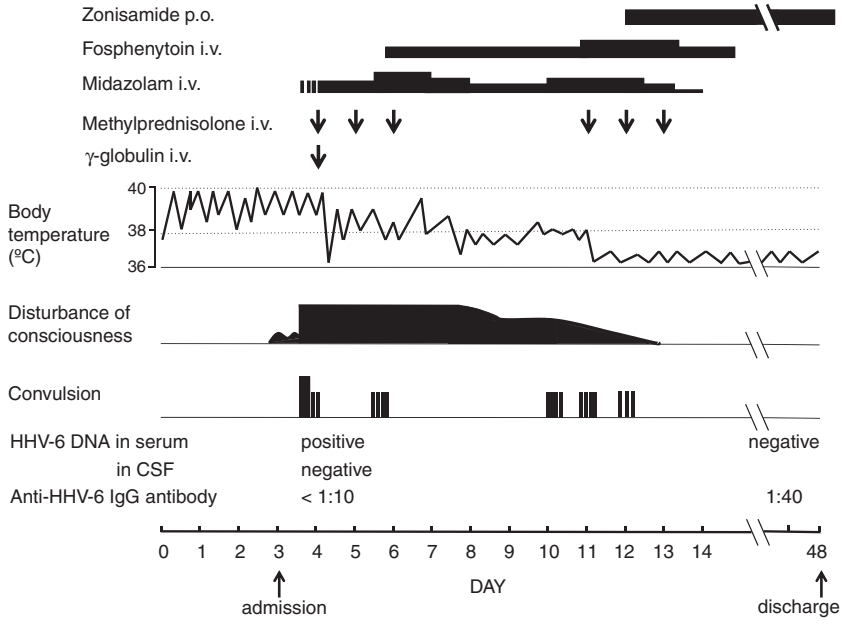

Fig. 1. Clinical course of the patient with HHV-6-associated encephalopathy. p.o., per os; iv, intravenous injection; CSF, cerebrospinal fluid.

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