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Case Report

Hippocampal signal abnormality on the first day of illness in acute encephalopathy with biphasic seizures and late reduced diffusion caused by HHV-6 infection

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Abstract

We report a 13-month-old girl who developed acute encephalopathy with biphasic seizures and late reduced diffusion (AESD) with transient reduced diffusion in the hippocampus and anterior commissure on diffusion-weighted imaging (DWI), which was performed on the first day after febrile status epilepticus (FSE) as the initial neurological symptom of AESD. DWI just after late seizures showed high signal intensity lesions in both left hippocampus and anterior commissure, and left extratemporal and occipital subcortical white matter. HHV-6 DNA was positive in both blood and cerebrospinal fluid samples. DWI at two months after onset showed atrophy in the left mesial temporal lobe and extratemporal and occipital lobe without the signal abnormalities.

Although it has been reported that magnetic resonance images tend to show no acute abnormality during the first two days in typical AESD, transient reduced diffusion could be found on the DWI performed on the first day of AESD.

Keywords: Febrile status epilepticus; Hippocampal injury; Human herpesvirus-6; Acute encephalopathy with biphasic seizures and late reduced diffusion; Diffusion-weighted image

1. Introduction

In typical acute encephalopathy with biphasic seizures and late reduced diffusion (AESD), magnetic resonance imaging (MRI) shows no abnormality during the first two days [1]. However, recent studies have reported that hippocampal abnormalities, such as reduced diffusion or enlargement, appear on diffusion-weighted imaging (DWI) within five days of febrile status epilepticus (FSE) in childhood [2,3]. Although FSE is also characterized as the initial neurological symptom of AESD, hippocampal abnormalities on the first day from the onset of AESD have not been reported.

Herein, we report a patient developing AESD with transient reduced diffusion in the hippocampus and anterior commissure on DWI performed on the first day after FSE as the initial neurological symptom of AESD.

2. Case report

A 13-month-old girl, the first child of healthy Japanese parents, was born at term by vaginal delivery,

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and had no previous health or developmental problems. She could walk without support and speak words from the age of 12 months. There was no family history of neurologic disease. She developed febrile illness, and the next day, prolonged generalized convulsions occurred for 30 min. Her seizures were halted transiently by injecting 0.1 mg/kg of intravenous midazolam (MDZ). However, at 4 h after cessation of the first prolonged seizure, a seizure cluster occurred, characterized by tonic–clonic seizures predominantly in the right arm and leg, and associated with eye deviation to the right. Ictal electroencephalography (EEG) showed high-amplitude spike-wave bursts arising predominantly from the left hemisphere. Seizures were stopped by intravenous continuous injection of 0.15 mg/kg/h of MDZ, and interictal EEG showed spikes on the left anterior temporal region and generalized high-amplitude slow-waves. Routine hematological studies, blood gas analysis, and all biochemical studies and cerebral spinal fluid examinations were normal. HHV-6 DNA was positive in both blood and CSF samples as detected using quantitative polymerase chain reaction. DWI performed on the first day after FSE showed reduced diffusion in the left hippocampus and anterior commissure (Fig. 1a-1 to a-3). ^{99m}Tc-ECD on the third day after FSE showed hyperperfusion at the same area (Fig. 2a). Because neuroradiological features, post-ictal EEG features, and repetitive seizures are atypical of febrile seizure, two courses of methylprednisolone pulse therapy (30 mg/kg/day × 3 days/week) were performed

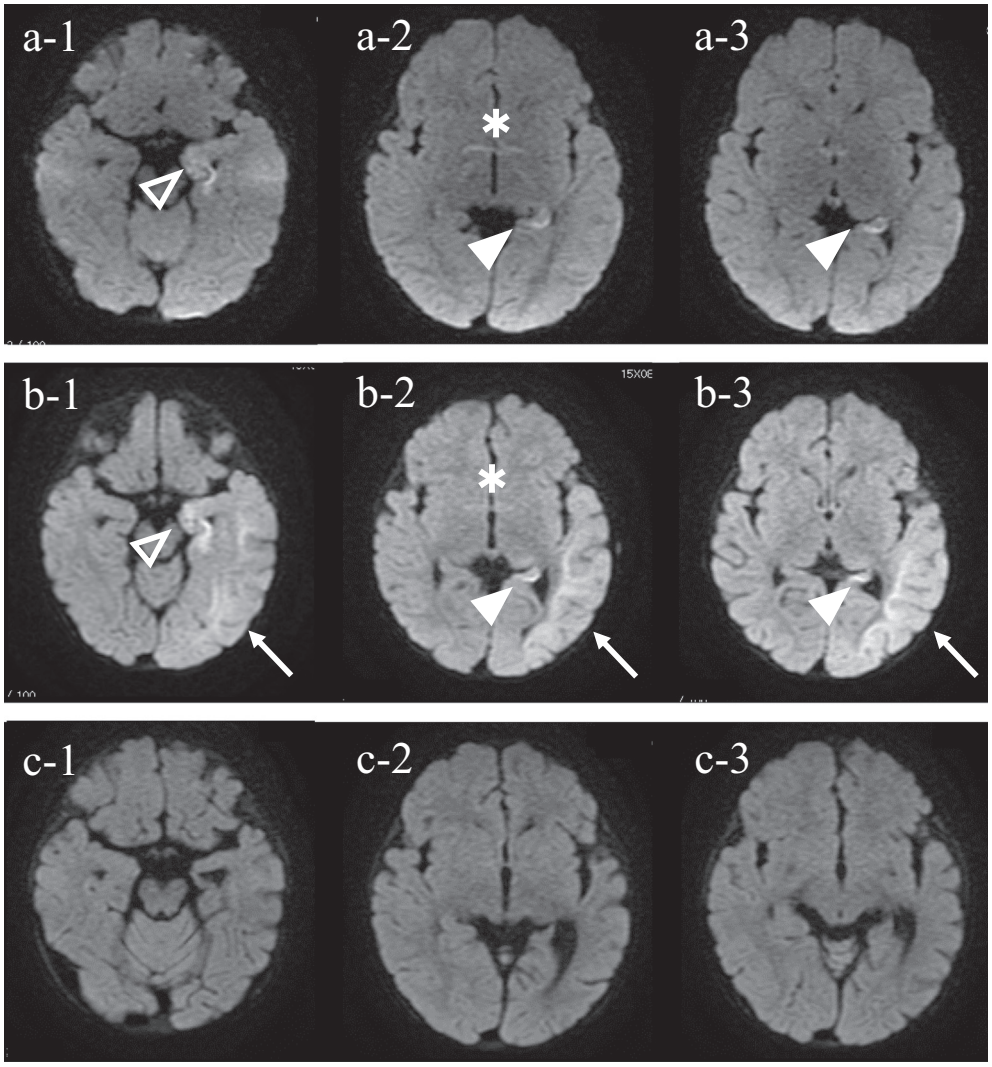

Fig. 1. Brain axial diffusion-weighted imaging. Upper row (a-1, a-2, a-3): on the first day of illness, reduced diffusion in the left amygdaloid body, uncus of parahippocampal gyrus, and hippocampus (a-1; open arrowhead) were found. Reduced diffusion in the left hippocampal tail (a-2 and a-3; closed arrowhead) and anterior commissure also were found (a-2; asterisk). No signal abnormalities were found in the extratemporal or occipital lobe (a-2 and a-3). *Middle row* (b-1, b-2, b-3): on the fifth day of illness, reduced diffusion in both in the left amygdaloid body, uncus of parahippocampal gyrus, and hippocampus (b-1; open arrowhead), anterior commissure and hippocampal tail (b-2 and b-3; asterisk and closed arrowhead), and extratemporal and occipital subcortical white matter (b-2 and b-3; straight arrows) were found. *Lower row*: two months after onset, atrophy in both left hippocampus (c-1) and extratemporal and occipital lobe (c-2 and c-3) appeared without the signal abnormalities.