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Case Report

A case of acute encephalopathy with biphasic seizures and late reduced diffusion: Utility of arterial spin labeling sequence

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Abstract

A 1-year-old boy was admitted because of febrile status epilepticus (FSE). A secondary cluster of seizures was seen on day 5 after onset, and the patient eventually displayed developmental delay. Conventional magnetic resonance imaging (MRI) showed no abnormal findings on day 1 after onset, but showed reduced diffusion in the subcortical regions of bilateral frontal lobes on day 5 after onset. Acute encephalopathy with biphasic seizures and late reduced diffusion (AESD) was diagnosed. Arterial spin labeling (ASL) revealed reduced cerebral blood flow (CBF) in bilateral frontal lobes on day 1 after onset and showed increased CBF in the corresponding region in the subacute phase. Outcomes after prolonged febrile seizures are usually good, but mental deficit and/or epilepsy often remain in AESD. Discriminating between these syndromes is difficult, because no useful biomarkers have been identified. Reduced CBF in bilateral frontal lobes was observed on ASL on day 1 of FSE in the present case, and this finding may be predictive of developing AESD.

Keywords: Febrile status epilepticus; Magnetic resonance imaging; Arterial spin labeling sequence; Acute encephalopathy with biphasic seizures and late reduced diffusion

1. Introduction

Acute encephalopathy affects 400–700 children each year in Japan, and acute encephalopathy with biphasic seizures and late reduced diffusion (AESD) is the most common subtype, accounting for about 30% of all cases [1]. AESD was reported as a new type of acute encephalopathy in 2006 [2,3]. In patients with AESD, febrile status epilepticus (FSE) is observed at onset, followed by transient recovery of consciousness. A secondary cluster of seizures occurs on day 3–6, with magnetic resonance imaging (MRI) showing reduced diffusion in subcortical regions. Although the mortality rate is low (<5%), the morbidity rate is high. Mild cases show recovery of higher cortical functions during subsequent weeks or month, but severe cases are left with more severe mental deficit and/or epilepsy [4]. Discriminating between AESD and complex febrile seizure in the early phase has been difficult, because no useful biomarkers have been established [5]. For example, MRI findings prior to the secondary phase have been reported as normal [4].

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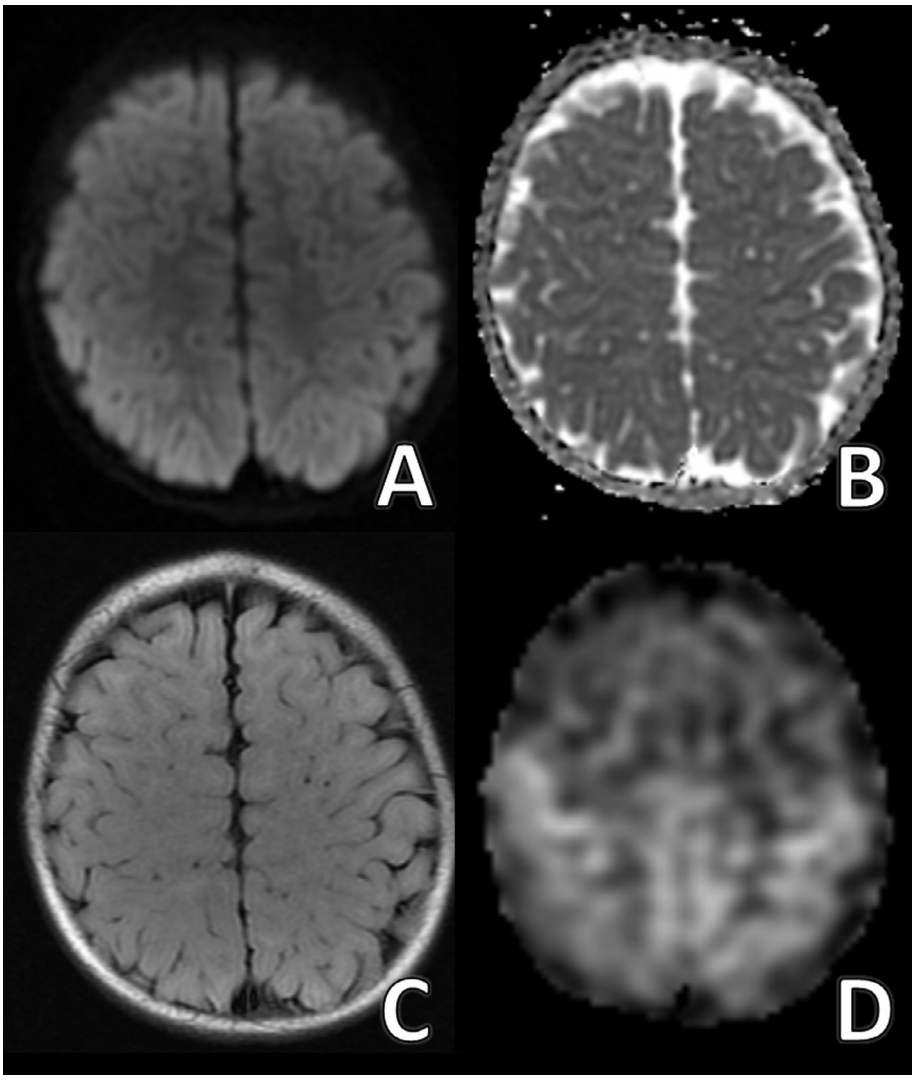

Fig. 1. MRI on the day of admission. No abnormal findings are observed on conventional sequences such as DWI (A), ADC mapping (B), and FLAIR (C). In contrast, markedly reduced CBF is observed in bilateral frontal lobes on ASL (D).

Arterial spin labeling (ASL) is a noninvasive MRI sequence that uses magnetically labeled blood water as a flow tracer, allowing imaging of cerebral blood flow (CBF). Previous studies have confirmed the utility of CBF measurements from ASL in evaluating seizures, particularly in the localization of seizure foci [6]. However, no studies have clarified ASL findings in AESD. In this case report, we performed ASL for a patient with AESD on day 1 of FSE and discuss the utility of this sequence in predicting AESD.

2. Case study

A previously healthy, 16-month-old boy was referred to our institution with FSE. He had a 2-day history of febrile illness. The seizure lasted 50 min and was controlled by administration of midazolam. On admission, he was comatose and electroencephalography showed bilateral high slow waves, predominantly in the frontal area. On brain MRI in the postictal state; 21 h after the first seizure, no abnormalities were evident on T1-weighted imaging, T2-weighted imaging, fluid attenuation inversion recovery (FLAIR) images, diffusion-weighted imaging (DWI) or apparent diffusion coefficient (ADC) mapping (Fig. 1A–C). However, CBF was markedly reduced in bilateral frontal lobes on ASL (repetition time, 4618 ms; echo time, 10.704 ms; post-labeling delay, 1525 ms; acquisition time, 90 s; Fig. 1D). After admission, he showed gradual recovery. On hospital day 4, rash developed on the trunk of the body, and exanthema subitum was therefore diagnosed. On day 5, brain MRI was performed after the patient became uncharacteristically grumpy. DWI and ADC mapping showed reduced diffusion in bilateral frontal lobes, predominantly in the