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Case report

A case of atypical benign partial epilepsy with action myoclonus

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ABSTRACT

We describe a boy, 3 years and 6 months old, who experienced a rolandic seizure accompanied by a cluster of atypical absence seizures, the EEGs for which corresponded to those of atypical benign partial epilepsy (ABPE). Of note, this patient suffered from developmental delay beginning in infancy and exhibited giant middle-latency somatosensory evoked potentials with action myoclonus. With the exceptions of ethosuximide, acetazolamide, and adrenocorticotropic hormone, which have been reported to be effective in ABPE, the atypical absence seizures were intractable despite extensive treatment with various anticonvulsants. The drugs that were effective led to a remarkable reduction in seizure frequency and EEG improvement, but the efficacy was temporary. The patient demonstrated moderate mental retardation without regression and could not walk with support or speak any meaningful words at the age of 3 years and 6 months. Based on thorough differential diagnosis, although further studies will be necessary, we propose that this boy may present a new phenotype of ABPE: ABPE with action myoclonus.

1. Introduction

Atypical benign partial epilepsy (ABPE) of childhood was described by Aicardi and Chevrie.[1] The onset of seizures occurs between 2 and 7 years of age. A period of typical rolandic seizures is followed by frequent atonic seizures, partial or generalized, that lead to frequent falls. These seizures were later termed "negative myoclonus of the lower limbs".[2] In addition to seizures characterized by falls, other types of seizures such as partial and generalized motor seizures as well as atypical absence seizures also occur.[1,2] The electroencephalogram (EEG) shows typical bilateral rolandic spikes and spike–wave activity during sleep. Based on these characteristics, ABPE has been considered to be a "classic" atypical variant of benign childhood epilepsy with centrotemporal spikes (BECT). Seizure prognosis in ABPE may be resistant to treatment. In the previous study of 43 patients with ABPE, 7 patients (16%) were not seizure-free at the end of the study.[6] Their ages ranged from 5 to 13 years. Furthermore, cognitive problems and mental retardation have frequently been noted.[6]

Giant middle-latency somatosensory evoked potentials (giant SEPs) have been reported in subjects with BECT. The giant SEPs in those patients have been interpreted as a consequence of an excitable sensorimotor cortex.[3,7] However, action myoclonus with giant SEPs has not been previously reported in cases of ABPE or other types of benign childhood epilepsy.

We describe a boy with rolandic seizures accompanied by clusters of atypical absence seizures; the EEG of the latter corresponded to those associated with ABPE. Of note, he suffered from developmental delay beginning in infancy and experienced giant SEPs with action myoclonus.

2. Case report

This boy was born uneventfully after 38 weeks of pregnancy. At the age of 10 months, tremor was observed in the extremities during wakefulness. Zonisamide and clonazepam were started because epileptic discharges were observed in bilateral frontal areas; this treatment did not affect the tremor. A GC–MS analysis of the urine showed no abnormalities.

At the age of 1 year and 4 months, he was referred to our hospital for rehabilitation due to developmental delay. On examination, he had normal facial appearance and mild hypotonia. His tremor increased at voluntary movements, which was considered to be action myoclonus because a surface electromyogram (EMG) showed

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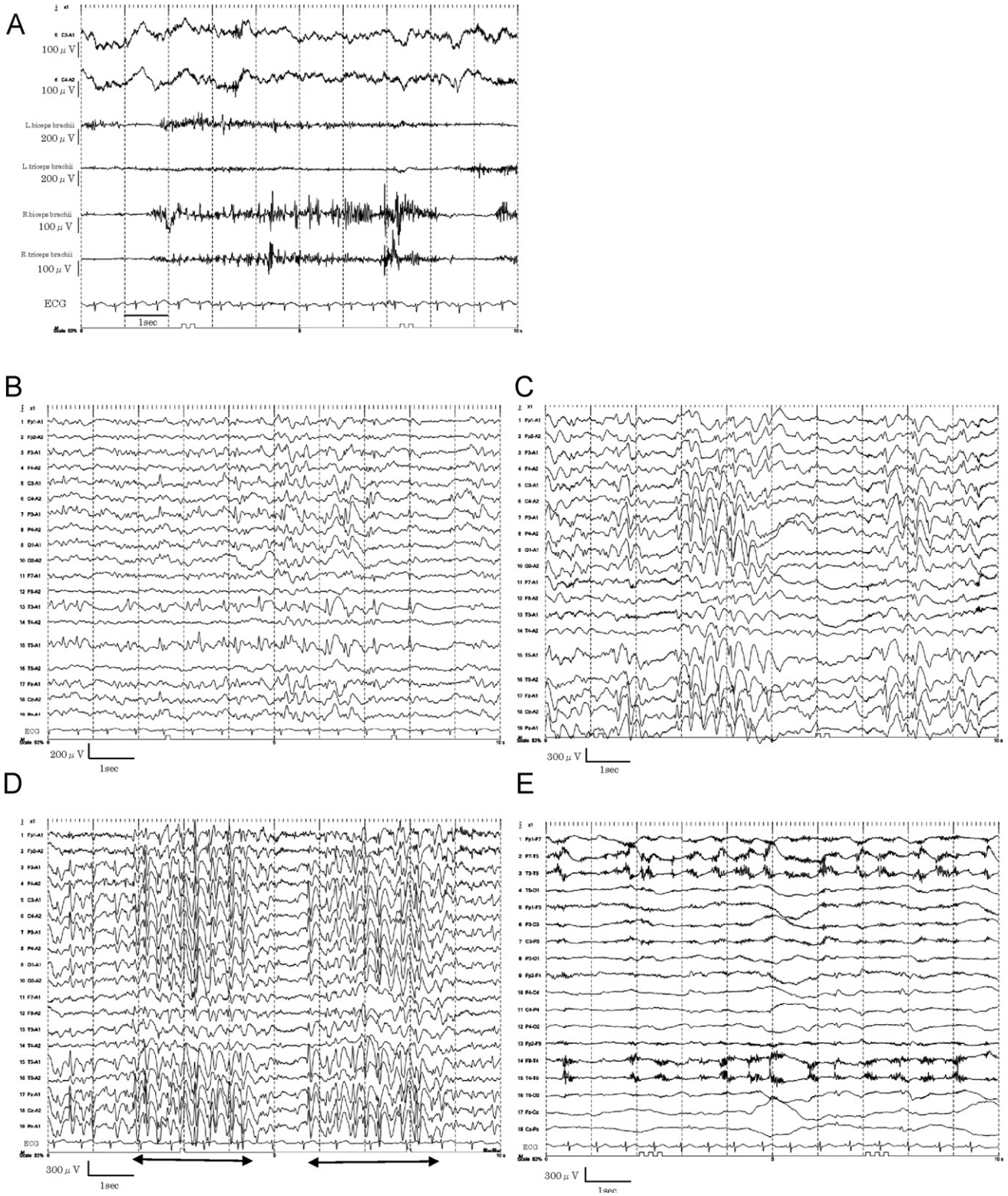

Fig. 1. EEG and surface EMG analysis of the patient. (A) Simultaneous recording of EEG and surface EMG during voluntary movement. Rhythmic co-contraction of the right biceps and triceps muscles, which was not accompanied by epileptic discharges, were observed at a rate of 5 Hz. (B) At the age of 1 year and 8 months, frequent bilateral centrotemporal sharp waves or spikes and waves were recorded and exhibited left-side predominance (sleep record). (C) At the age of 2 years and 6 months, diffuse spike-and-wave complexes were recorded, with maximum accentuation over both centrotemporal regions (awake record). (D) At the age of 2 years and 9 months, motionless arrests for a few seconds (arrows) were frequently observed; these corresponded to almost continuous diffuse spike-and-wave complexes with left-side predominance, which were considered to be clusters of atypical absence seizures (awake record). (E) Diffuse spike-and-wave complexes vanished 1 week after starting ACTH. The artifacts of teeth grinding were seen in the left temporal area (awake record).