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Original article

Manifestation of both emetic seizures and sylvian seizures in the same patients with benign partial epilepsy

Harumi Yoshinaga *, Katsuhiro Kobayashi, Takashi Shibata, Takushi Inoue, Makio Oka, Tomoyuki Akiyama

Department of Child Neurology, Okayama University Graduate School of Medicine, Dentistry and Pharmaceutical Sciences, Japan

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Abstract

Purpose: Benign childhood epilepsy with centro-temporal spikes (BECTS) and Panayiotopoulos syndrome (PS) have different pathophysiologies and show different types of seizures, yet they overlap in some important respects. In an attempt to understand the ways in which they differ from each other and overlap each other, we performed a detailed investigation on patients who had both characteristic types of seizure manifestations, namely, sylvian seizures and emetic seizures. *Subjects and methods:* We recruited consecutive subjects from the EEG database of outpatients who had visited our hospital between 2008 and 2010 and who had been diagnosed with BECTS or PS. As a result, 45 patients with BECTS and 50 patients with PS were selected from the database. Viewing the clinical records of these 95 patients, five patients were selected who had experienced both sylvian seizures and emetic seizures. Next, the clinical features and EEG findings of these five patients were retrospectively observed at the date of investigation: October 1, 2011. *Results:* We found that all the patients showed rolandic spikes when they had sylvian seizures, and occipital spikes or multifocal spikes when they had emetic seizures. We also report in detail on one patient who showed two different types of ictal EEG patterns: one of which started in the occipital area and the other of which was located in the rolandic area. *Conclusion:* Based on these findings, we conclude that widespread cortical hyperexcitability that includes the occipital area is necessary to produce the autonomic seizure manifestations seen in PS.

Keywords: Panayiotopoulos syndrome; Benign childhood epilepsy with centro-temporal spikes; Rolandic spikes; Occipital spikes; Emetic seizures; Sylvian seizures

1. Introduction

Benign childhood epilepsy with centro-temporal spikes (BECTS) [1] and Panayiotopoulos syndrome (PS) [2] are both types of benign childhood partial epilepsy. Both are observed in normal children without any abnormal brain structures and both have a good prognosis. The differences between two syndromes are onset age, seizure manifestations and typical electroencephalogram (EEG) findings. The characteristic seizure manifestation of BECTS is the so-called sylvian seizure [3], while the typical seizure manifestation of PS is characterized mainly by autonomic symptoms, particularly vomiting [2,4–6]. Centro-temporal spikes, namely rolandic spikes, are the hallmark of BECTS, whereas interictal EEGs of PS show greater variability, with a predominance of occipital spikes and occasional rolandic spikes [4].

The overlap of these two syndromes has been widely investigated by several authors including groups of

* Corresponding author. Address: Department of Child Neurology, Okayama University Medical School, 2-5-1 Shikatacho, Okayama 700-8558, Japan. Tel.: +81 86 235 7372; fax: +81 86 235 7377.
E-mail address: magenta@md.okayama-u.ac.jp (H. Yoshinaga).

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Panayiotopoulos as a main stream. Sibling cases of PS and BECTS have been reported [4] and it has also been reported that 3.5–12% of patients with PS developed sylvian seizures later in their course [2,4,5].

Therefore, in this study, we performed an investigation on patients who manifested both sylvian seizures and emetic seizures.

2. Subjects and methods

For this study, we recruited consecutive patients from the 9840-EEG database of patients who visited our hospital between 2008 and 2010 and who had been diagnosed with BECTS or PS. The diagnostic criteria for BECTS were unilateral sensory and/or motor seizures involving the face and rolandic spikes, and those for PS were autonomic seizures, especially emetic ones. As a result, 45 patients with BECTS and 50 patients with PS were selected from the original database of 1386 epileptic patients under 16 years of age. Viewing the clinical records of these 95 patients, five patients were found who had exhibited both clinical seizure types, namely, sylvian seizures and emetic seizures. Next, the clinical features and EEG findings of these five patients were retrospectively observed at the date of investigation: October 1, 2011.

The epileptic focus of each EEG was determined using referential and bipolar recordings to find the maximum voltage or the phase reversal of each spike. Rolandic (R) spikes were defined as spikes discretely localized on electrodes C3, C4, T3, T4 or spikes with phase reversal on these electrodes. Occipital (O) spikes were defined as spikes discretely localized on electrodes P3, P4, O1, O2, Pz or spikes with phase reversal on electrodes P3, P4, Pz. Multi-focal spikes were defined as rolandic spikes and occipital spikes concurrently observed with or without frontal spikes.

This study was approved by the Okayama University Ethics Committee.

3. Results

We found one male and four females who had both types of seizures. The clinical courses with the features of seizures and spike foci are shown in Fig. 1.

Age at onset of afebrile seizures ranged from 3 years and 1 month to 6 years and 1 month. The timing of occurrence of the two types of seizures was as follows: All the patients experienced one or several seizures in which both types of seizure semiology were mixed and observed concurrently. Vomiting seizures appeared before sylvian seizures in one patient. In two patients, sylvian seizures occurred before the mixed type of seizures. As shown in Fig. 1, sylvian seizures did not necessarily occur after emetic seizures or the mixed type of seizures. All of the patients studied had prolonged seizures, and autonomic epileptic status was observed in four patients. In patient 5, the onset seizure type was status epilepticus with the mixed semiology, and no other seizures occurred after this first single seizure in spite of the fact that this patient received no anti-epileptic treatment.

The foci of the interictal spikes varied by case. The most severe case, patient 3, once showed continuous spike-waves during slow wave sleep (CSWS). In contrast, patient 5 showed EEG improvement without treatment within just 1 year of the first seizure manifestation. Occipital spikes or multifocal spikes including the occipital area were commonly observed when the patients experienced the mixed type of seizures.

In what follows, we will discuss patient 2 in detail because this case is of particular relevance to the present study. This patient was a girl in whom two patterns of ictal EEGs were recorded. She was referred to us because two febrile seizures with prolonged generalized atonia and facial pallor occurred at 2 years and 2 months of age. She had had a normal development after an uneventful gestation and delivery. Her EEG first showed occipital spikes at the first visit.

At the age of 3 years and 9 months, she developed an afebrile seizure during sleep with hemifacial twitching. Since then her EEG began to show rolandic spikes interictally.

At the age of 5 years and 6 months of age, she experienced a prolonged left hemiconvulsion with vomiting that lasted for 45 min. Just after this seizure, we performed an EEG and fortuitously caught an ictal EEG of a left hemiconvulsion with vomiting. Although her interictal EEG showed multifocal spikes including rolandic spikes, her ictal EEG showed predominance in the occipital area and occipital focal slow waves remained after seizure cessation (Fig. 2).

After this seizure, she started to have intractable sylvian seizures with hemifacial twitching without any autonomic signs including vomiting. Her EEG began again to show rolandic spikes without occipital spikes.

In this type of seizure, as shown in Fig. 3, her ictal EEG started with repetitive small spikes in the rolandic area followed by alpha activity in the same area, while the occipital area was never involved. This ictal pattern is different from that shown in Fig. 2. After being started on lamotrigine, her seizures ceased. Both her neurological and mental states were normal, and her MRI was also normal.

4. Discussion

Caraballo et al. reported in their prospective study of 192 PS patients that 24 children (12.5%) also had seizures with speech arrest and hemifacial motor symptoms that are characteristic of BECTS, concomitantly with otherwise typical PS seizures [5]. Sylvian seizures in