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LETTER TO THE EDITOR

Concomitance of benign epilepsy with centrotemporal spikes and childhood absence epilepsy: an unusual case

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Dear Editor:

An 8-year-old girl presented with a right-sided focal motor seizure with secondary generalization during sleep. The focal motor seizure involved her face and mouth with salivation. After the first seizure, her mother noticed frequent brief episodes of staring and unresponsiveness during the daytime. The past and family medical histories were otherwise unremarkable. Neurologic examination and mental function were normal. The EEG simultaneously showed frequent centrotemporal spikes and waves on the left and synchronous 3 Hz generalized spike and wave (GSW) complexes up to 17 s (Fig. 1a, b). And, centrotemporal spikes preceded typical 3 Hz GSW complexes (Fig. 1c). The MRI brain scan was normal. Concomitant BECTS and CAE were diagnosed, and valproic acid was initially prescribed at the dose of 15 mg/kg/day. However, she developed rashes and itching on her skin. Lamotrigine was substituted and increased to 5 mg/kg/day. The patient has been seizure free and without symptoms for 6 months at the time of this report.

The coexistence of two idiopathic epilepsy syndromes remains controversial, whereas the coexistence of different seizure types within a focal epilepsy syndrome is widely accepted. BECTS and CAE are the most common epilepsy syndromes in children. However, the concomitance of BECTS and CAE is extremely rare and generally not accepted [1, 2]. To our knowledge, only three patients have been reported with simultaneous clinical manifestations of BECTS and CAE [1–3]. Datta et al. reported that the coexistence of 3 Hz GSW and Rolandic discharge on EEG is extremely rare in their large cohort over a period of 25 years. Furthermore, incidental Rolandic spikes on EEG in patients with absence epilepsy were the most common finding. Therefore, Rolandic discharges were considered most likely an incidental finding which is not suggestive of BECTS. They concluded that the coexistence of BECTS and CAE is coincidental and distinct pathophysiological mechanisms are likely [4].

Some investigators have studied circuit mechanisms common to GSW discharges and focal discharges. Huntsman et al. demonstrated that the thalamic circuit, integral for 3 Hz GSW, could produce local recurrent activity that is highly organized and repetitive, such as Rolandic discharges [4, 5]. While thalamocortical oscillatory hypersynchrony was intensified in mutant mice with absence seizures, a small fraction of isolated thalamic slices demonstrated focal reverberant activity [5]. Further evidence suggests that a cortical focus initiates GSW complexes and leads to a paroxysmal oscillation within the thalamocortical loop. By speculating on the evolving concepts concerning the pathophysiology of CAE, such as the "cortical focus" theory, focal cortical changes may also influence local recurrent oscillatory activity of the thalamocortical loop, and this could induce the appearance of different epilepsy syndromes in a patient with genetic susceptibility [3].

However, it is difficult to explain the association of the two syndromes in the same patient. Furthermore, Rolandic discharges tend to be enhanced during sleep while 3 Hz GSW in CAE typically occur during the awake state. More experimental studies would contribute to a better understanding and further studies based on clinical and electrophysiologic features are needed in cases of concomitant BECTS and CAE. The clinical expression of both syndromes is defined as multifactorial and resulting from interactions between genetic and environmental factors [1]. Our case indicates the possible association of BECTS and CAE. BECTS and CAE might be pathophysiologically related, especially in patients with underlying genetic susceptibility [3]. Thus, genetic research would also be helpful to evaluate the pathophysiology of these two epilepsy syndromes.

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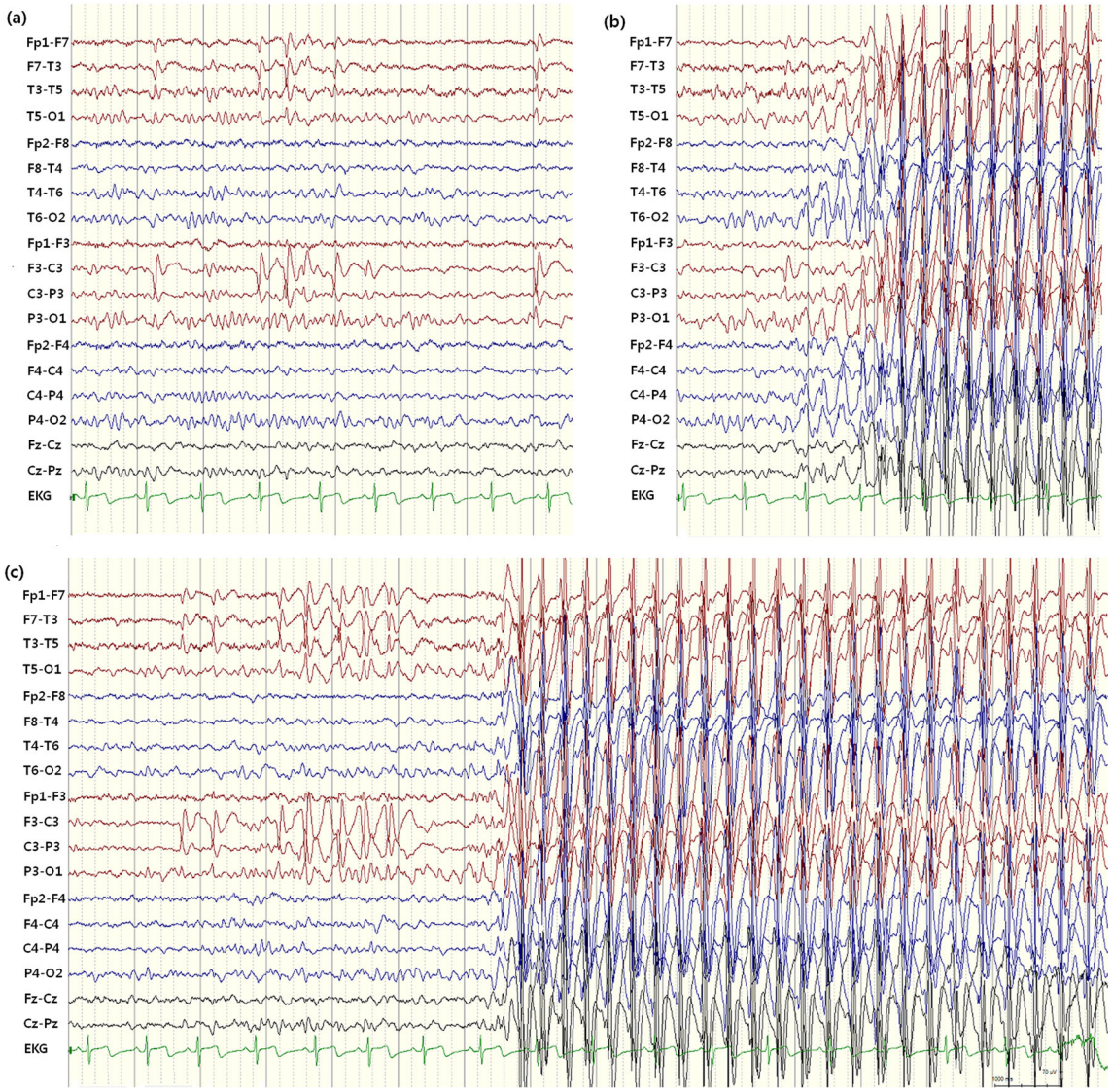

Fig. 1 EEG of the patient. The EEG simultaneously shows frequent centrotemporal spike and waves on the left and synchronous 3 Hz generalized spike and wave complexes (**a**–**c**). Centrotemporal spikes preceded typical generalized 3 Hz spike-wave complexes (**c**)

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflicts of interest.

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