Case report

EEG and seizure exacerbation induced by carbamazepine in Panayiotopoulos syndrome

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ABSTRACT – We report on a 4-year 8-month-old boy with Panayiotopoulos syndrome who showed atypical evolution with newly developed absence seizures and EEG exacerbation induced by carbamazepine. Soon after the introduction of carbamazepine, EEGs began to worsen, and finally absence seizures and myoclonic seizures appeared. Immediately after we discontinued carbamazepine, the seizures disappeared and the EEG improved. Carbamazepine may induce unusual electroclinical features, electrophysiologically explained by bilateral synchrony. This case provides more evidence of the close links between Panayiotopoulos syndrome and benign childhood epilepsy with centrotemporal spikes.

Key words: Panayiotopoulos syndrome, carbamazepine, absence, benign partial epilepsy, seizure exacerbation

Panayiotopoulos syndrome (PS) is a type of benign partial epilepsy of childhood (Panayiotopoulos 2002a, Panayiotopoulos 2002b, Caraballo et al. 2000). Recently, the relationship between benign childhood epilepsy with centrotemporal spike (BECTS), another representative type of benign partial epilepsy of childhood, and PS has been investigated from various standpoints, and attention has been drawn to the pathophysiological similarity of these two syndromes (Caraballo et al. 2000, Yoshinaga et al. 2005). One of the common characteristics of these two syndromes is that some patients demonstrate similar, atypical evolutions in which absence seizures and myoclonic seizures appear concomitantly with a worsening of the EEG (Caraballo et al. 2001, Ferrie et al. 2002).

CBZ is the drug of choice for benign partial epilepsy. However, CBZ has been reported to cause atypical evolutions associated with a deteriorating EEG in BECTS (Perucca et al. 1998, Nanba and Maegaki 1999). To the best of our knowledge, a similar exacerbation with CBZ has not been previously reported in a patient with PS. This is the first report of a boy with PS who showed EEG and seizure exacerbation induced by CBZ.

Case report

The patient was born uneventfully to non-consanguineous parents, and his development was normal. His younger sister had experienced febrile convulsions. His first seizure occurred at age 4 years and 3 months during...
of brief spells disappeared. The VPA dose was valproate (VPA). After complete replacement, the episodes of brief spells disappeared. The VPA dose was 300 mg/day, and the VPA blood level was 53.6 µg/mL. His EEG improved dramatically, with infrequent spike waves involving the left temporal-occipital area (figure 1F). The frequency and amplitude of spike discharges decreased, and the generalization of the spike-wave discharges disappeared. After the replacement of CBZ by VPA, he became alert as regards movements and expression, and his appetite also improved.

Discussion

Because of the typical clinical symptoms and the characteristic EEG findings, we concluded that our patient was suffering from PS, a type of benign partial epilepsy of childhood. PS occurs in normally developed children, with a peak age-at-onset of 4 or 5 years with no abnormal findings on neuroimaging examinations. The most characteristic seizure manifestations of PS are the existence of autonomic symptoms, mainly emetic, often accompanied by a deviation of the eyes and/or unresponsiveness. The seizure frequency is usually low, but prolonged seizures and complex partial status epilepticus, as in our case, are not unusual. Nevertheless, PS has a good neurological outcome. Intertical EEG predominantly shows occipital spikes. Although PS was first described as an early-onset childhood epilepsy with occipital paroxysms (CEOP), in contrast to a late-onset CEOP described by Gastaut, it has been recently thought that PS has closer links with BCECS than with the Gastaut type of CEOP. Several patients with PS have been reported whose seizure manifestations changed with age from typical PS seizures to sylvian seizures. Taking all these findings together, Panayiotopoulos and his colleagues proposed a unified concept for benign childhood partial epilepsy, that is, benign childhood seizures susceptibility syndrome, including PS as the earliest type and BCECS as another type.

It is well known that 19% of patients with Panayiotopoulos syndrome show diffuse epileptic discharges in their EEG evolution. However, absence seizures are rarely observed in PS (Carballo et al., 2004). Although we could not record the ictal EEG of our patient’s brief spells, diffuse slow spike and wave bursts lasting several seconds in his interictal EEG strongly indicate that these episodes were absence seizures. Atypical evolutions of BCECS with absence seizures are well known as atypical benign partial epilepsy. Several cases of PS patients reportedly showed similar, atypical evolutions in their clinical courses (Carballo et al., 2001, Ferrie et al. 2002). In 2001, Carballo et al. reported two patients with PS who began to have absence seizures and myoclonic seizures associated with mental regressions. The EEGs of both patients changed to CSWS (Carballo et al. 2001). Although our patient had absence seizures and epileptic myoclonus along with the appearance of diffuse slow spike and waves, his EEGs did not display typical CSWS. In 2002, Ferrie et al. reported a...
Figure 1. A) An EEG after the first complex partial status epilepticus at age 4 years and 3 months. It shows left occipital spikes during sleep. B) An EEG during sleep, after the introduction of CBZ at age 4 years and 8 months. It shows so-called cloned-like repetitive complexes in the left posterior head area associated with slight generalization. C) An EEG during sleep, after the regular intake of CBZ at age 5 years. Cloned-like, repetitive, multifocal spike-wave complexes increased and showed a strong tendency toward generalization. D) An EEG during wakefulness at age 5 years. It shows diffuse slow spike and wave bursts that continued for about three seconds, suggesting the existence of absence seizures. However, no clinical manifestations were noticed at this EEG recording. E) An EEG-EMG polygraphic recording performed at the same age as figures 1C and D. Note that the spike component of the generalized spike and wave discharges is time-locked with the myoclonic jerks (indicated by the arrow) recorded from the deltoids. F) An EEG during wakefulness at age 5 years and 1 month, when the patient took only VPA after discontinuing CBZ.
A girl with PS who began to have absence seizures and absence status. She initially had clinical features typical of PS, followed by sylvian seizures two months later. She then began to have absence seizures (Ferrie et al. 2002). As in our patient, she took CBZ at the time of the appearance of absence seizures. However, her absence seizures did not disappear with the discontinuation of CBZ. In contrast, our case showed a dramatic improvement on the EEG and a disappearance of the absence seizures immediately after CBZ was stopped. In conclusion, although our patient shared similar clinical and EEG evolution with those reported by other authors (Caraballo et al. 2001, Ferrie et al. 2002), he was characterized by the closer relationship between his evolutional change and CBZ treatment.

Seizure exacerbations induced by CBZ were initially reported in cases that had generalized seizures, such as absence seizures and generalized tonic-clonic seizures (Shields and Saslow 1983). However, Talwar et al. found eight cases of symptomatic, localization-related epilepsy among a total of 26 cases that displayed EEG exacerbation at an age of less than 6 years (Talwar et al. 1994). Regarding benign partial epilepsy and seizure exacerbations by CBZ, Lerman (1986) first reported a patient with BCECS that began to have drop spells after the initiation of CBZ. Nanba and Maegaki also reported a patient with BCECS who began to have epileptic negative myoclonus and appearances of diffuse slow spike and waves in the EEG that were induced by CBZ (Nanba and Maegaki 1999). Similar to our patient, all these cases showed EEG improvement and seizure cessation immediately after the discontinuation of CBZ.

To the best of our knowledge, this is the first report of a boy with PS who showed EEG and seizure exacerbation induced by CBZ. We believe this case provides more evidence of the close links between PS and BCECS. Although CBZ is a drug of choice for PS, we should consider the possibility of exacerbation and observe carefully the clinical changes and EEG evolutions during CBZ therapy.

References


