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SeLECTS—Self-Limiting Epilepsy with Centrotemporal Spikes: Presence of ESES Correlates with Neuropsychological Regression

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Int J Ep

Abstract

We present a developmentally normal 8-year-old male child with infrequent nocturnal focal seizures since 6 years of age. Since the onset of seizures, he has been having poor academic performance with difficulty in learning and preserved language. Electroencephalogram (EEG) showed a normal background and frequent bisynchronous biphasic centro-parieto-temporal large amplitude spikes with maximum negativity over the left temporal region. His sleep record showed continuous runs of large amplitude generalized spikes with maximum negativity over the centrotemporal region and spike-wave index > 90. There is a clear correlation between the presence of electrical status epilepticus during sleep (ESES) and neuropsychological deficits. EEG is resistant to antiseizure medications, and these findings disappear before mid to late adolescence, and deficits recede with the remission of ESES.

Keywords
- BECTS
- SeLECTS
- CSWS
- ESES
- benign epilepsy with centrotemporal spikes

We present the case of an 8-year-old male, presented with complaints of infrequent episodes (three witnessed episodes over 1 year) of nocturnal awakening and behavioral arrest with sustained deviation of head and eyes to the right side with subtle focal clonic jerks of the right upper and lower limbs, with onset at 6 years of age. He was born nonconsanguineously, with no abnormal antenatal, perinatal, or developmental history with prior good academic performance. Since the onset of the seizures he had difficulty in academic performance with reduced attention and memory in school. Intelligence quotient and neuropsychological assessment was incomplete due to poor cooperation of the child. Magnetic resonance imaging was normal and awake electroencephalogram (EEG) had a normal background and frequent bisynchronous biphasic centro-parieto-temporal large amplitude spikes with maximum negativity over the left temporal region (►**Figs. 1** and **2**). His sleep record showed continuous runs of large amplitude generalized spikes with maximum negativity over the centrotemporal region and spike-wave index (SWI) > 90 (►**Fig. 3**). He was treated with valproate, brivaracetam, and clobazam currently. Oxcarbazepine previously administered was discontinued in view of rashes. In view of the scholastic regression despite antiseizure medications, a course of intravenous immunoglobulin (IVIG) was provided. Our patient reported a significant subjective increase in scholastic performance with improvement in attention and memory after the administration of IVIG; however, there was no discernible change in the EEG SWI done after 1, 3, or 6 months.

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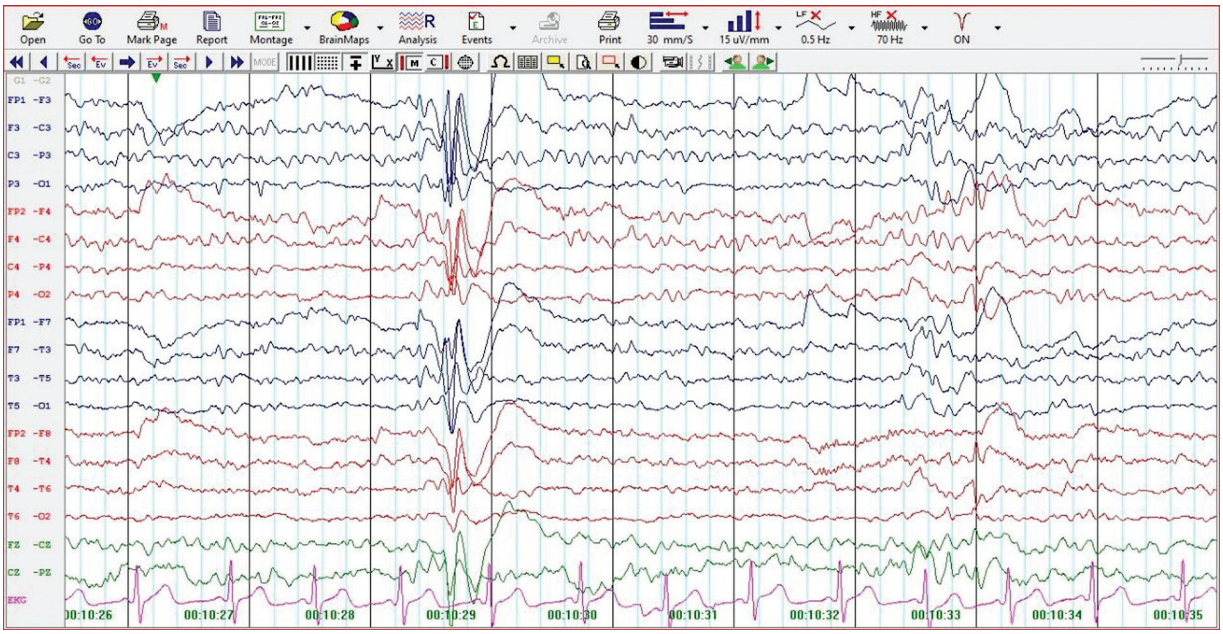

Fig. 1 Awake electroencephalogram (EEG) record showing normal background and bisynchronous centro-parieto-temporal (CPT) large amplitude spikes with maximum negativity over the temporal region (with an amplitude dominance over the left hemisphere).

SeLECTS (self-limited epilepsy with centrotemporal spikes), formerly known as benign epilepsy with centrotemporal spikes (BECTS)benign rolandic epilepsy, typically onsets between 4 and 10 years of age (peak 7 years) and usually remits by mid to late adolescence and has a male preponderance.[1] EEG is characterized by normal background activity and activation of high-amplitude biphasic and triphasic centrotemporal spike and slow wave complexes with a transverse dipole (frontal positivity, temporoparietal negativity) during drowsiness and sleep, which may be unilateral, bilateral, or independent. The presence of continuous spike-wave activation (SWI > 85% is required for diagnosis of electrical status epilepticus during sleep [ESES]) during sleep warrants evaluation for progressive language or cognitive impairment. The spectrum of BECTS, atypical benign focal epilepsy of childhood or pseudo-Lennox syndrome, status epilepticus of benign epilepsy with centrotemporal spikes, acquired epileptic aphasia or Landau–Kleffner syndrome, and epileptic encephalopathy with continuous spike-and-wave during sleep was differentiated based on the degree of language, neuropsychological impairment/regression, and the degree of ESES.[2] Recently retermed, epileptic encephalopathy with spike-wave activation in sleep (EE-SWAS) is made when a developmentally normal child has neuropsychological regression after

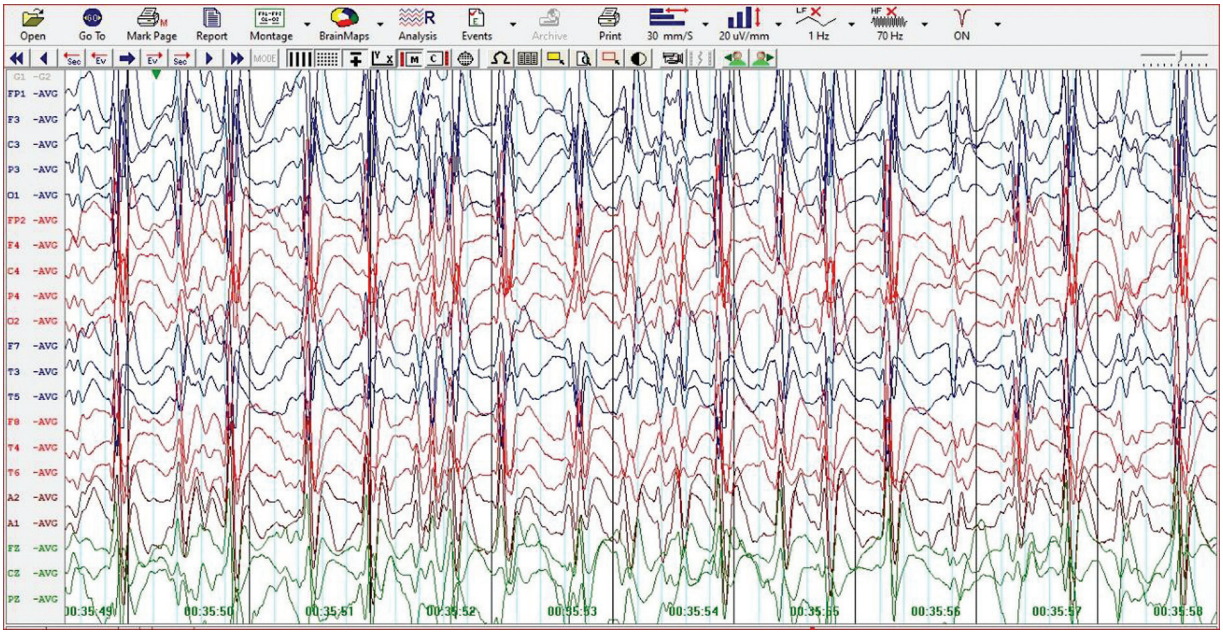

Fig. 2 Activation of abundant bisynchronous biphasic centro-parieto-temporal (CPT) large amplitude spikes with maximum negativity over the left temporal region highlighting frontal positivity and tangential dipole in average montage.

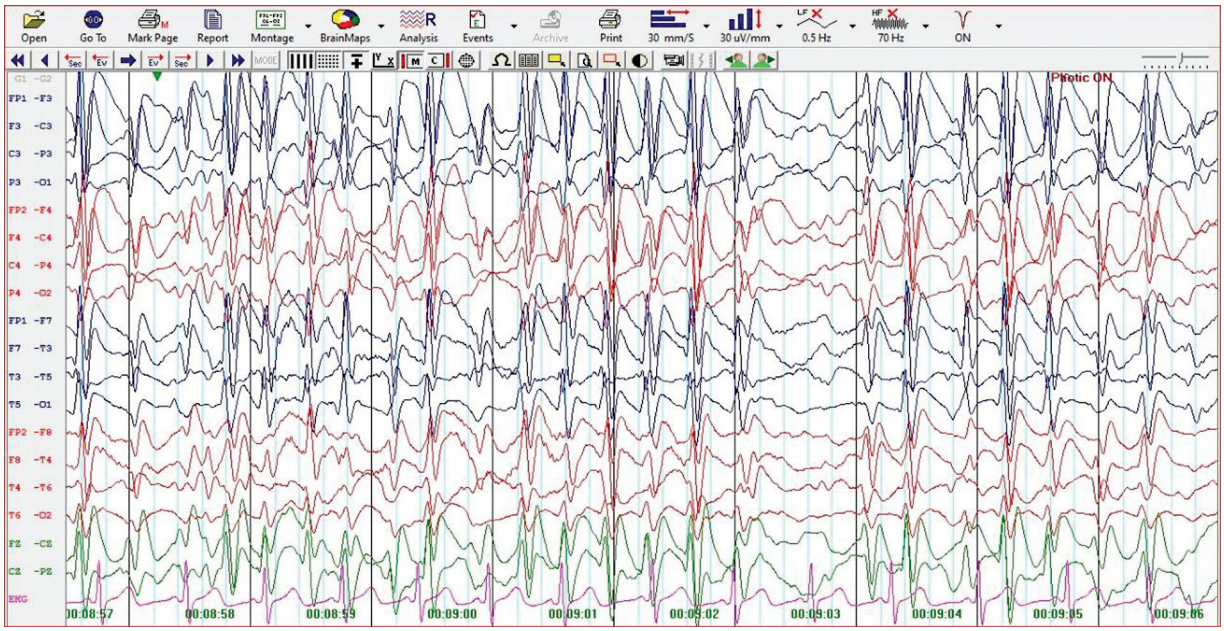

Fig. 3 Activation of large amplitude continuous spike and waves with spike-wave index > 90 during slow wave sleep—electrical status epilepticus during sleep (ESES).

seizure onset.[1] Developmental encephalopathy with spikes-wave activation in sleep (DE-SWAS) occurs in one with pre-existing developmental delay or just language delay. There is a clear correlation with ESES and individuals with neuropsychological deficits when the SWI > 85%.[3] Seizures tend to be resistant to multiple antiseizure medications, and they usually disappear before adolescence and deficits recede with the remission of ESES. Genetic etiologies of heterozygous pathogenic variants in GRIN2A or fragile X syndrome have been reported.[1,2]

Statistical Analysis
None.

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Conflict of Interest
None declared.

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