

Case Report

Hemi-ESES associated with agenesis of the corpus callosum and normal cognition

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1. Introduction

Continuous spike wave during sleep (CSWS) is an epileptic encephalopathy mainly defined by cognitive decline, various seizure types, and a specific electroencephalographic pattern, called electrical status epilepticus during sleep (ESES) [1,2]. ESES is characterized by abundant and diffuse spike-and-wave complexes during slow-wave sleep. The percentage of these spike-and-waves during sleep is called the spike-wave index (SWI) [3]. To define ESES, the SWI should be present in at least 85% of NREM sleep; however, the cutoff point has been defined in different studies to be between 25 to 85% of NREM sleep duration [2,4]. CSWS is a rare and age-related epileptic encephalopathy with a prevalence of 0.5% in children with epilepsy [2].

The pathophysiology of this syndrome is unknown; however, recent investigations suggest that continuous spike and wave during sleep may interfere with sleep-related physiologic functions and the restorative process of slow-wave sleep due to the lack of sleep down-scaling (overnight decrease in slow-wave slope), entailing cognitive and memory dysfunction [5,6]. In almost all patients with CSWS, seizures stop and the ESES pattern disappears during or after puberty; in only 10–40% of these children, however, cognition and language becomes normal after the ESES pattern has resolved. Therefore, early detection and treatment have a great impact on the final prognosis [1,6].

ESES is often a generalized pattern, but some studies have reported cases with hemi-ESES in children with polymicrogyria, porencephalic cysts, hydrocephalus, and thalamic lesions [7–9].

The corpus callosum is the most prominent anatomic commissural structure connecting and integrating the activities of both cerebral hemispheres. Moreover, the disruption of callosal fibers prevents seizure propagation and evolution to a generalized seizure [10].

We herein describe a child with normal cognition and nocturnal seizures, after a thorough workup, we found hemi-ESES and absolutely no corpus callosum in the patient.

2. Case report

Our patient was a 6-year-old girl referred for nocturnal seizures. Her parents were first cousins, and she was delivered by cesarean section following an unremarkable pregnancy. Her development was normal, and she was right-handed. At age 3, she developed an event during sleep, later defined as seizure. The seizure began as a left perioral twitch with subsequent left hand clonic movements and left lateral gaze lasting for about 2 min, without any significant postictal confusion. A thorough neurological examination, biochemistry workup, and a standard EEG were performed, but no signs of abnormality were seen. A year following the first seizure, she developed two more seizures, which were similar to the first one. After these seizures, sodium valproate was started, and a brain MRI was performed. Her MRI showed complete absence of the corpus callosum along with colpocephaly (Fig. 1). Her one-hour video EEG showed frequent right centrotemporal spike-and-slow waves. She experienced no seizures over the next two years. Later on, however, she developed another semiology of nocturnal focal seizures manifest as left hand and face clonic jerks. After these seizures reoccurred, she referred to our center for long-term video-EEG monitoring (LTM).

After admission at our center, we discontinued her medication and waited for three days prior to conducting LTM. After four days of monitoring, her EEG showed very few right centrotemporal spike-and-slow waves during the awake state. We observed a prominent accentuation of these spike-and-slow waves during slow sleep. Her SWI showed a hemi-ESES pattern over the right hemisphere with 50–85% spike-and-slow wave complexes during NREM sleep (Fig. 2). During NREM, the architecture of sleep was normal on the left side, yet severely disturbed on the right side. After administering high doses of benzodiazepine (diazepam orally 1 mg/kg stat at night), the hemi-ESES pattern was completely resolved. We conducted the Wechsler IQ test (WPPSI) just before

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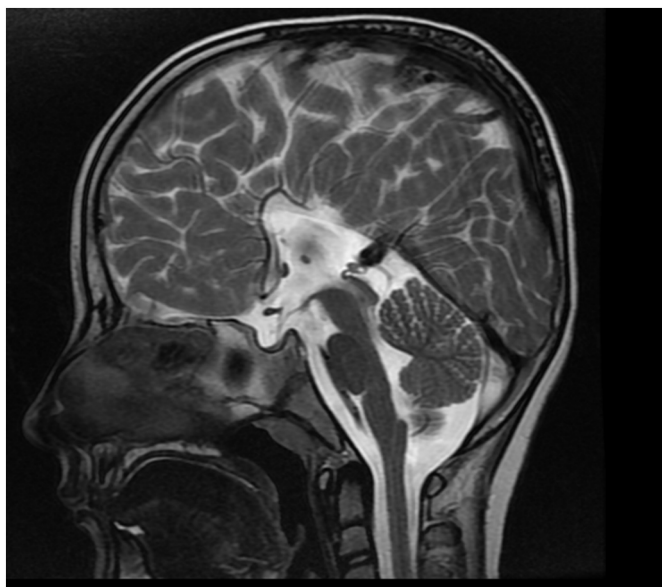


Fig. 1. Sagittal brain MRI of index case. MRI depicts complete agenesis of corpus callosum.

and one day following the administration of benzodiazepine. Her IQ score was surprisingly in a normal range, where the verbal IQ score was 92; average score for verbal tests was 44 and for each subtest: Information = 8, Vocabulary = 10, Similarities = 10, Arithmetic = 10, Comprehension = 6), performance IQ score was 101; average score for performance tests was 51 and for each subtest: Animal house = 10, Picture completion = 10, Mazes = 10, Geometric design = 12, Block design = 9) and total IQ score was 97 with an average total score of 95. As far as intelligence score is concerned, there was no significant difference between her verbal and

performance score, nor was any difference observed before and after taking diazepam.

3. Discussion

The ESES pattern has been reported in patients with focal cerebral lesions including unilateral thalamic lesions [8]. Kelman A. et al. suggested that this finding is elucidated by electrographic bilateral synchrony propagated via corpus callosum on EEG. Other studies have shown the ESES pattern both in patients without a brain lesion, and in those with epilepsy associated with different types of focal cortical lesions. Diffuse cortical spike-and-slow waves could be explained by spreading from a focal onset to the hemispheres bilaterally via the corpus callosum [8].

In our patient, the presence of hemi-ESES is explained by the congenital absence of the corpus callosum inhibiting the propagation of spike and slow waves during sleep from the right side of the brain to the left, thus limiting bilateral synchronous expression of ESES. It is assumed that sparing the left dominant hemisphere from continuous spike-and-slow waves during sleep in our patient accounts for the normal cognitive and language scores in a right-handed patient with agenesis of the corpus callosum. Although sleep has a potent activation role for expressing interictal epileptiform discharges, for example in patients with frontal lobe epilepsy, it differs from ESES with a pattern of centrottemporal spikes with respect to: 1) dipole orientation that has a tangential dipole with a negative pole in the central sulcus and a positive pole on the anterior head region in centrottemporal spikes, and 2) the few seizures involving the hand and face motor area in the typical ESES pattern seen with centrottemporal spikes.

4. Conclusion

Our findings emphasize the focal nature of ESES and role of the corpus callosum in bilateral synchronous expression of hemispheric epileptiform discharges.



Fig. 2. EEG features of index case. EEG shows a hemi-ESES pattern over the right hemisphere with 50–85% spike and slow wave complex during NREM sleep.

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Declarations of interest

None.

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