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Seizure in benign epilepsy with centro-temporal spikes

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ABSTRACT – Rolandic epilepsy is frequent in children but seizures are rarely seen by the physician or captured by video-EEG monitoring. In most children the attacks are few and sporadic and generally occur at night. Also, a high percentage of children with benign epilepsy with centro-temporal spikes (BECTS) have characteristic centro-temporal spikes based on routine EEG monitoring and therefore do not require further tests such as telemetry or sleep studies. We report a video-EEG recording of a seizure in a patient with rolandic epilepsy which may be useful for educational purposes. *[Published with video sequences]*

Key words: benign rolandic epilepsy, benign epilepsy with central temporal spikes

Case study

A 13-year-old boy presented with a history of nocturnal seizures from the age of seven years. The frequency was approximately two per week. His mother had described twitching of the eyes and mouth, salivation and jerking of arms. On awakening, he felt unusual, dribbled and was unable to speak properly following the seizures. He was otherwise a healthy child. Apart from a paternal uncle who had fits as a child (the exact details not known to the parents), there was no other significant family history. There was nothing of note on examination and no neurological abnormality was detected.

At the age of 10 years, he experienced daytime seizures with an aura in which he felt something was wrong. He felt numbness in his tongue, was jerking and dribbling and was aware of having a seizure but unable to control his actions. There was no loss of consciousness.

The increased frequency of nocturnal seizures along with daytime seizures resulted in day-time somnolence. Treatment was commenced with carbamazepine as the drug of choice to control partial seizures. Unfortunately, he developed a rash and fever within two weeks of starting the medication which was then changed to sodium valproate.

The frequency of seizures increased with sodium valproate to about four per week. With increasing dose, seizure control improved, however, he developed pancytopenia along with excessive tiredness which was reflected in an increase in ammonia. Therefore, a decision was made to stop sodium valpraote and start levetiracetam treatment. His seizures and tiredness responded well to the treatment change. Due to atypical clinical features (duration of epilepsy and especially daytime seizures) brain MRI was performed and even repeated. The repeated MRI brain scan showed mild asymmetry of the temporal horns, with the right smaller than the left. There was no volume loss/signal change and no evidence of mesial sclerosis.

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Video-EEG

Inter-ictal

Background activity was within normal limits for the child's age. During the EEG-intermittent spikes or spike-and-wave complexes upon awakening, phase reversing occurred in the right centro-temporal region with amplitudes of up to 100 µv. The complexes appeared in runs and clusters at 1-5/second, becoming florid and more persistent with increasing drowsiness and sleep.

Ictal

During stage 2 NREM sleep (after 36 minutes into recording) there was a continuous 14-second run of 2-3 cycles per second-delta and spike-and-wave activity phase reversing in the right temporal region. The patient became alert from sleep (which was reflected in the EEG) and after two seconds opened his eyes. Three seconds later, he then turned his head to the left (from lying on his right side) and the left side of his mouth was drawn upwards. At this point, he was asked if he was OK but was unable to reply (afterwards he confirmed that he had heard and understood but was unable to speak). The EEG at this time showed a build-up of continuous rhythmic spikes/sharp waves in the right mid and post temporal regions and the right frontal region. The left hand then came up to his mouth and there was a rapid rhythmic twitching of the left mouth and around the eyes (with rapid rhythmic eyelid flickering), on the left more than the right. The head turned further to the left and there was rhythmic jerking of the head and shoulder with pharyngeal/laryngeal sounds and hypersalivation (the seizure activity appeared more widespread at this time, however was masked by movement and myogenic artefact).

Post-ictal

The seizure lasted 50 seconds and ceased abruptly at which point the patient relaxed and appeared briefly dazed and tired with mild coughing. At the offset there was a general reduction in overall amplitude together with some post-ictal delta activity over the right hemisphere (many electrodes on the left side had fallen off). There were no discernable spikes for 80 seconds following the seizure, after which they gradually returned as before. After 30 seconds, he was able to nod positively in response to a question, and was able to speak after 40 seconds. Consciousness was retained throughout the seizure.

The clinical history, inter-ictal and ictal EEG features and the presence of a focal motor seizure arising from stage 11 sleep, together, are consistent with the diagnosis of benign epilepsy with centro-temporal spikes (BECTS) or rolandic epilepsy *(see video sequence)*.

Discussion

BECTS is the most common type of partial motor epilepsy in children and constitutes up to 15-20% of all childhood epilepsies, with onset between 2-14 years and peak at 8-9 years.

Central temporal spike waves occur in 2-3% of normal school-aged children of whom less than 10% develop rolandic epilepsy (Panayiotopoulos, 2005). Centro-temporal sharp waves are found in 1-3% of healthy children (Panayiotopoulos, 2005) and are seen in up to 30% of siblings of children with BECTS (Doose *et al.*, 1997). It is estimated that only 8.8% of children with rolandic spikes have seizures (Lüders *et al.*, 1987).

BECTS is characterized by: onset of seizures between two and 14 years of age (usually between three and 10 years), simple partial motor seizures (occurring most often during sleep) as an exclusive or dominant type of seizure in the vast majority of cases, characteristic EEG foci occurring on a normal background tracing in the lower rolandic (sylvian or "midtemporal") area and absence of neurological or intellectual abnormalities before and during seizure activity (Arzimanoglou *et al.*, 2004). Seizures occur during sleep or upon awakening in about 65-70% of cases, exclusively when awake in 10-20% cases and during sleep and the awake state in the remaining cases (Dalla Bernardina *et al.*, 2002).

In BECTS, brain MRI is usually normal. For 15% of patients with BECTS, patients may have abnormal findings because of static or other brain diseases unrelated to the pathophysiology of BECTS (Gelisse *et al.*, 2003). Hippocampal abnormality has been detected in some children with BECTS by MRI and proton magnetic resonance spectroscopy (Lundberg *et al.*, 2003).

Inter-ictal EEG, as described above, is usually seen in a routine recording. Topography of the paroxysms may change and bilateral and unilateral foci may be found on different tracings in the same patient. The spikes in BECTS are unilateral in about 60% of cases and bilateral in 40% of cases (Lerman, 1998).

Very few reports of ictal EEG have been described for patients with BECTS in the literature (Panayiotopoulos, 2005). Ictal video-EEG is not often captured because of the following reasons:

– in most children the attacks are few and sporadic;

– attacks generally occur at night and not during the day when most routine EEGs are recorded;

– a high percentage of children with BECTS have characteristic centro-temporal spikes during routine EEG monitoring and therefore do not require further tests such as telemetry or sleep studies which would increase the possibility of capturing ictal events;

– the benign characteristic of BECTS means that further investigations such as sleep deprived EEG are not generally undertaken.

The lack of opportunity to observe the clinical events or semiology in patients with BECTS, therefore, makes case studies, such as the one reported here, of interest to the clinician. □

Legend for video sequence

Example of typical epileptic seizure in benign epilepsy with central temporal spikes.

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