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Guest Editorial

Revocable attention deficit hyperactivity disorder- An eye-opener

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ABSTRACT

Aim: To highlight the clinical and electroencephalographic features, treatment strategies and outcome in atypical variant of benign childhood epilepsy with centrotemporal spikes (BECTS).
Materials and Methods: Subject underwent video electroencephalograph (EEG), high-resolution magnetic resonance imaging (MRI), neuropsychological evaluation and language assessment and response to medication was followed up. In addition to sodium valproate, treatment with intravenous methylprednisolone (given as monthly bolus doses) was given as the seizures remained refractory to anti-seizure medications (ASM) alone.
Results: Earlier onset, increased frequency and duration of focal seizures compared to prototype Rolandic epilepsy are cardinal features suggestive of atypical variant of BECTS. Head drop and truncal sway due to axial or axiorhizomelic atonia occurring several times per day or week was the key manifestation. When the atypical seizures commenced, one or more of the following problems can occur: hyperactivity, attention deficit, and mild cognitive or language dysfunction. Child became seizure free on a combination of intravenous methylprednisolone, with sodium valproate and his hyperactivity reversed.
Conclusion: BECTS in children with an early age of onset, frequent and prolonged seizures and scholastic decline pari passu with seizure onset are more likely to evolve into atonic-BECTS. Carbamazepine, oxcarbazepine and some benzodiazepines may worsen these seizures. They can become seizure free with immunomodulatory therapy, namely methylprednisolone (given as monthly bolus doses) or intravenous immunoglobulin (IVIG), and have complete resolution of the transient motor and cognitive impairment. Atonic-BECTS needs to be differentiated from Lennox–Gastaut syndrome since it is potentially treatable and children recover with no sequel.

1. Introduction

Benign childhood epilepsy with centrotemporal spikes (BECTS) is the commonest idiopathic, age-specific epilepsy syndrome of childhood with an estimated prevalence of 20–25% in school children with epilepsy.[1,2] The diagnostic criteria for BECTS include: (i) brief, stereotyped , focal, unilateral facial motor seizures with somatosensory symptoms, with a tendency for secondary generalization, commonly occurring in sleep; (ii) onset between the ages of 3 and 13 years; (iii) spontaneous recovery before 16 years; (iv) absence of anatomic central nervous system (CNS) lesions that could cause epilepsy.[3]

Most subjects with BECTS have infrequent seizures which can be managed with one anti-seizure medication (ASM). However, some cases require poly therapy especially those with early onset (< 5 years).[2] Upto 10% of the cases may need multidrug therapy and the existence of bilateral abnormalities and high spike load in electroencephalograph (EEG) is a predictor for multidrug therapies.

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BECTS is a heterogeneous entity with many atypical forms. In one of the atypical forms of BECTS, typical Rolandic seizures interspersed with atonic seizures, focal or generalized, that lead to frequent falls has been described.[3] There are several case series describing children with this isolated phenomenon.[3]However, atonic-BECTS is often misdiagnosed as catastrophic epileptic encephalopathy of childhood namely Lennox-Gastaut syndrome (LGS) because of the drop attacks, polymorphic seizures and scholastic deterioration. In addition to atonic-BECTS, electrical status epilepticus in sleep (ESES) and some ASM can accelerate cognitive decline in children with BECTS leading to a poor outcome.

2. Case Vignette

A five year old boy born of non-consanguineous parentage, with normal birth and early development presented with seizures from three and half years of age. His seizure semiology was characterized by uprolling of eyes followed by ictal cry and sudden jerking of both upper and lower limbs with impaired awareness lasting for few seconds. He immediately regained consciousness after the event, with no post ictal headache or vomiting. The event frequency was high and he often had 4- 5 seizures per day. He had behavioral issues at school and he tended to cause physical harm to peers. His academic performance was poor and he failed to assimilate basic language skills and was unable to keep at par with his classmates. Within a few months he developed severe hyperactivity and had difficulty in attending classes which forced his parents to withdraw him from school. Attempts at home education also proved futile. He later developed a new seizure semiology in the form of slow head drops where he would slump without losing consciousness. The frequency of these episodes gradually escalated to approximately sixty per day.

He was evaluated with a scalp EEG which showed normal background activity with bifrontal spike polyspike and wave discharges in wakefulness with marked activation of bifrontal, centro-parieto-temporal and generalised discharges during sleep with burst attenuation pattern (Figure 1). Magnetic resonance imaging (MRI) of the brain (1.5T) was normal. He was started on sodium valproate which was gradually hiked upto 30 mg/kg/day which abated his generalised seizures and reduced his head drops by approximately 50 %. He continued to have daily head drops of around 30 times per day and further addition of zonisamide was unrewarding. A repeat scalp EEG did not show any remarkable difference from the first. He was initiated on immunotherapy with pulse intravenous methyl prednisolone 20mg/kg five doses every month along with continuation of antiepileptic drugs. After the first dose of pulse intravenous steroid the child was seizure free and after 6 months of monthly 5 day steroid pulse schedule his scalp EEG showed absolute clearance of discharges in wakefulness and sleep with a normal background activity. His cognition and behavioral symptoms also improved as evidenced by an objective change in his Receptive Expressive Emergent Language Scale (REELS) score and intelligence quotient. His pulse intravenous steroids were further reduced to once every 2 months with continuation of ASM. He continues to be seizure free with improved cognition and no behavioral issues.

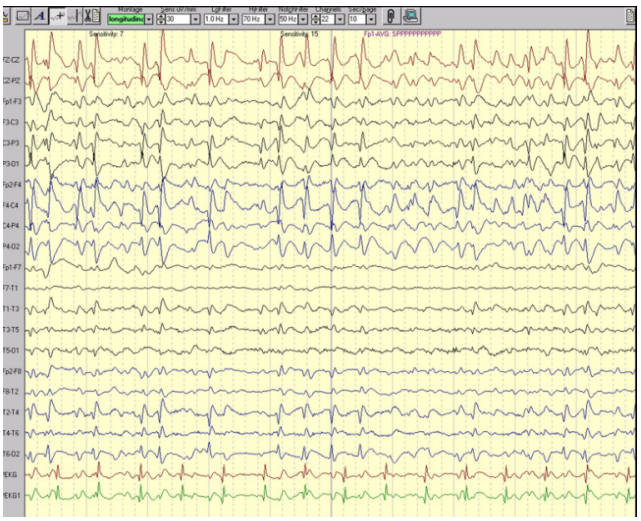

Fig. 1: An increased frequency of centro-parieto-temporal spikes has been reported in children who present with attention deficit hyperactivity disorder where the spike load is ~ 130 /minute (sensitivity 7 microV; HFF=70 Hz ; LFF=1 Hz)

Table 1: Features of atypical BECTS

1.	Earlier age of onset- usually by 3 years of age
2.	Seizure semiology: Focal atonic seizures in trunk and extremities. Focal seizures-more likely to become hemi-generalized or generalized
3.	Seizure frequency/duration: atonic seizures-occurring multiple times daily. Focal seizures-more frequent and prolonged
4.	Cognition and language: Major changes in cognition and language have been associated along with behavioral problems, attention deficit, and cognitive and language dysfunction
5.	Treatment- Broad spectrum AEDs like sodium valproate, ethosuximide, zonisamide preferred. Drugs for focal epilepsy- carbamazepine, phenytoin, oxcarbazepine and benzodiazepines may worsen seizures
6.	Immunomodulation helps in medically refractory cases
7.	Imaging Normal
8.	Tendency to relapse seen

3. Discussion

BECTS is generally associated with a favorable prognosis. However, atypical variants of this syndrome exist and

Table 2: Electroencephalograph (EEG) predictors for anatypical BECTS

1.	Presence of bilateral centro-temporal spike and wave discharges in trains
2.	Presence of fronto-central and centroparietal spike and wave discharges
3.	High spike load with spike load more than 60-80 /minute in slow wave sleep,
4.	Initially positivity of the centro- parieto- temporal spikes
5.	Presence of polyspikes
6.	Rarely generalised paroxysmal fast activity (GPFA) and burst attenuation pattern (mimicking LGS)

if diagnosed wrongly can lead to incorrect treatment decisions. The proportion of children with BECTS that have atypical-BECTS has been reported as ranging from 1% to 7%.[1] The known atypical BECTS subtypes include those with a higher seizur frequency and other co-morbidities such as attention deficit hyperactivity disorder(ADHD), tics and/or learning disability.[2,4] The distinction between BECTS and atonic-BECTS is that, atonic seizures are the signature feature of the latter. The atonic seizures usually appear in clusters. In addition, there are focal and generalized motor seizures as well as atypical absences. Atonic-BECTS can rarely manifest as falls, initially without preceding typical Rolandic seizures.[1] The clinical picture includes slurred speech, atonic nodding of the head, or facial myoclonias.[5] Syndromes with centrotemporal spikes may vary from the benign to the severe. Landau-Kleffner syndrome and ESES are associated with significant cognitive impairments[4] and are at the severe end of the spectrum of these epilepsies. The features of atypically evolving BECTS have been summarized in Table 1 and EEG predictors for an atypical BECTS in Table 2.

4. Conclusion

Atonic-BECTS is a relatively infrequent but distinct electro-clinical variant of BECTS. BECTS in children who have an earlier age of onset than in the classic form, with more frequent and prolonged focal seizures are more likely to evolve into atonic-BECTS. Atonic seizures of the trunk and extremities occurring multiple times per day with unprecedented falls are the predominant seizure subtype in atonic-BECTS. High resolution MR imaging is normal and video-EEG monitoring may be essential to confirm the diagnosis. Carbamazepine and benzodiazepines can worsen these seizures. The onset of the atypical seizures may coincide with behavioral problems, attention deficit and mild cognitive and language dysfunction.[4] They have a favorable response to steroids and/or intravenous immunoglobulin; the transient motor and cognitive impairments noted during the period of seizure worsening are completely reversible.[3]

This case highlights a completely revocable ADHD which needs to be recognized.[6,7] A thorough evaluation with EEG and imaging is warranted before embarking on anti ADHD medication.

5. Conflict of Interest

The authors declare no relevant conflicts of interest.

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None.

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