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Original article

Atonic variant of benign childhood epilepsy with centrotemporal spikes (atonic-BECTS): A distinct electro-clinical syndrome

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Abstract

Purpose: To describe the clinical and electroencephalographic features, treatment strategies and outcome in a series of children with the atonic variant of benign childhood epilepsy with centrotemporal spikes (atonic-BECTS).

Material and methods: Out of the 148 patients with BECTS reviewed from January 2005 to June 2010 in our Institute, there were seven (5%) with atonic-BECTS. All underwent video EEG, high-resolution magnetic resonance imaging (MRI), neuropsychological evaluation and language assessment. Their progress was followed. In addition to sodium valproate, three were treated with steroids, followed by intravenous immunoglobulin (IVIG) when the seizures relapsed while tapering or after stopping the steroids.

Results: All of the children had earlier onset (mean = 2.4 years), increased frequency and increased duration of focal seizures compared to typical BECTS. Head drop and truncal sway due to axial or axiorhizomelic atonia occurring several times per day or week was the key manifestation. The atonic seizures worsened with carbamazepine in three, clonazepam in two and clobazam in one. When the atypical seizures commenced, some children developed one or more of the following problems: hyperactivity, attention deficit, clumsy gait, and mild cognitive or language dysfunction. Three children became seizure free, one on steroids and the other two on IVIG.

Conclusions: BECTS in children with an early age of onset and frequent and prolonged seizures is more likely to evolve into atonic-BECTS. Carbamazepine and some benzodiazepines may worsen these seizures. Three children became seizure free with immunomodulatory therapy, one on steroids and the other two on IVIG, and had 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. Although all the children in this series continued to be on treatment with sodium valproate it is currently undetermined whether they would have required to do so if followed up for an extended period of time.

Keywords: Atypical benign partial epilepsy; Pseudo-Lennox syndrome; Rolandic epilepsy; Immunomodulatory therapy

1. Introduction

Benign childhood epilepsy with centrotemporal spikes (BECTS) is the most common 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, stereotypical, simple partial, unilateral facial motor

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seizures associated with somatosensory symptoms, and a tendency for secondary generalization, commonly occurring in sleep; (ii) onset of seizures between the ages of 3 and 13 years; (iii) spontaneous recovery before the age of 16 years; (iv) absence of anatomic central nervous system (CNS) lesions that could be related to the epilepsy [3].

BECTS is a heterogeneous entity with many atypical forms. Consensus definition on atypical subgroups of BECTS is still lacking. In one of the atypical forms of BECTS, typical Rolandic seizures interspersed with frequent atonic seizures, partial or generalized, that lead to frequent falls has been described [4,5]. There are several small case series describing children with this isolated phenomenon [5–8]. However, atonic-BECTS is not generally recognized as an established syndrome and it is often misdiagnosed as catastrophic epilepsy because of the atonic drop attacks. It is currently not included in the international classification of epilepsies and epileptic syndromes.

Against this background we undertook a study on children with atonic-BECTS with the following objectives:

1. To describe the clinical, imaging, electroencephalographic, language and neuropsychological characteristics of a series of children with atonic-BECTS.
2. To assess the response to antiepileptic drugs and/or immunomodulatory therapy.
3. To assess the seizure and cognitive outcome.

2. Materials and methods

2.1. Subjects

The study was undertaken in the Department of Neurology, Sree Chitra Tirunal Institute for Medical Sciences and Technology, which is a tertiary referral center for neurological diseases in South India. From the prospectively maintained database, the electroencephalograms (EEG) of all children between January 2005 and June 2010 were reviewed. Those cases with centrotemporal discharges in the EEG were reviewed by one of the epileptologists involved in the study (AR) to determine if the neurophysiologic features were typical of BECTS as defined by the International League Against Epilepsy [9]. The latter describes "blunt high-voltage centrotemporal spikes, often followed by slow waves that are activated by sleep and tend to shift or spread from side to side." After systematically screening all the medical records of those satisfying the above criteria, we identified all cases of BECTS, who satisfied the following inclusion criteria:

1. Onset between the ages of 1 and 14 years.
2. A unilateral or bilateral spike focus located in the centrotemporal (Rolandic) area on the interictal EEG with seizures associated with sleep and one or more of the following ictal phenomena:
   a. Oro-pharyngo-laryngeal manifestations.
   b. Speech arrest.
   c. Somatosensory symptoms around the face.

Details of seizure semiology, including time of occurrence, duration, and localizing features, medications, patient and family history, and clinical and neuropsychological outcome were recorded.

We excluded patients having the following.

1. Exclusive occipital or frontal foci in EEG.
2. Abnormal magnetic resonance Imaging (MRI) suggestive of a symptomatic cause.
3. Presence of cognitive and language impairment before the onset of seizures.

From this cohort, we identified children with atonic-BECTS characterized by initial focal Rolandic seizures followed by seizures characterized by loss of tone of trunk and/or head drops and/or negative epileptic myoclonus of extremities with or without falls. During the study period, 148 children satisfied neurophysiologic and clinical criteria for BECTS. Of these, seven children (5%) fulfilled the criteria for atonic-BECTS (Table 1).

2.2. Electroencephalography (EEG)

All recordings were carried out on a 16-channel digital EEG acquisition system (NicVue, Nicolet-Viking, USA); with the scalp electrodes placed according to the International 10–20 system. The scalp-EEG (SEEG) was recorded for 40 min (20 min awake and 20 min sleep record) and included 3 min of hyperventilation and photic stimulation in wakefulness. A partial sleep deprivation protocol was used [10]. SEEG was performed at baseline and every 6 months on follow-up. All patients with atonic-BECTS also underwent a 24 h video-EEG (VEEG) at presentation and at each relapse. The distribution of interictal epileptiform discharges (IEDs) during prolonged video-EEG monitoring was assessed by visual analysis of interictal EEG samples for 15 s every 15 min.

The EEG analysis included assessment of the background activity for focal slowing, defined as the presence of localized slow waves not present in the other homotopic region. IEDs were categorized as either a diphasic/triphasic sharp-wave/spike or a spike-wave complex pattern with or without a tangential dipole. The location and maximum negativity of the discharges were categorized as central (C3/4), mid-temporal (T3/4), or parietal (P3/4). Extra-Rolandic epileptic foci were also noted. The laterality of the discharges was characterized as unilateral (right or left-sided) or bilateral. The latter were described as either bilateral dependent (when they occurred within 30 ms) or bilateral independent (with more than 30 ms dispersion). The spike frequency, presence of trains, and location of