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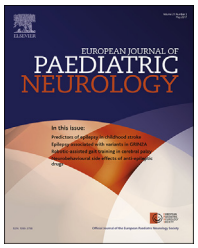

Original article

Coexistence of childhood absence epilepsy and benign epilepsy with centrotemporal spikes: A case series

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ABSTRACT

Aim: Childhood absence epilepsy (CAE) and benign childhood epilepsy with centrotemporal spikes (BECTS) are the most common forms of childhood epilepsy. Recent studies in animal models suggest that the two phenotypes may represent a neurobiological continuum. Although the coexistence of CAE and BECTS has been reported, this issue remains controversial. The purpose of this study was to analyse the electro-clinical characteristics of a group of children with contemporary or subsequent features of absence seizures and focal seizures consistent with BECTS.

Material and methods: A systematic record review from 8 epilepsy centres was used to identify 11 subjects, 5 females and 6 males, with electro-clinical documented consecutive or contemporary coexistence of CAE and BECTS.

Results: Patient's age ranged between 7.8 and 17.3 years. Four out of 11 patients presented concomitant features of both syndromes, whereas the remaining 7 experienced the two syndromes at different times.

Conclusions: Although CAE and BECTS are clearly defined syndromes and considered very different in terms of their pathophysiology, they share some features (such as similar age

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of onset, overall good prognosis), and can occur in the same patient. The long term prognosis of these patients seems to be good with an excellent response to anticonvulsant therapy.

1. Introduction

Childhood absence epilepsy (CAE) is one of the main age-dependent (age of onset between 4 and 10 years, with peak age between 5 and 7 years) forms of idiopathic generalized epilepsy, accounting from 2% to 10% of all childhood epilepsies; the electro-clinical characteristics include recurrent typical absence seizures (AS) accompanied by bilateral, symmetrical, and synchronous discharges of 3-Hz generalized spike and waves on the electroencephalogram (EEG).[1–4]

Recent studies have shown a possible coexistence of AS with other forms of epilepsy such as West syndrome, Panayiotopoulous syndrome, Gastaut-type idiopathic occipital epilepsy, Myoclonic epilepsy of infancy, Juvenile myoclonic epilepsy and Benign epilepsy with eentrotemporal Spikes (BECTS).[5–11]

BECTS accounts for 8–20% of childhood epilepsies and it is the most common focal childhood epileptic syndrome. Generally, seizures start between 3 and 13 years of age, with a peak at 8–9 years. This syndrome is characterized by oropharingolaryngeal and hemifacial sensorimotor seizures with hypersalivation, often during sleep, and typical EEG pattern of central and temporal focal epileptiform discharges. BECTS usually has a self-limited course with recovery before or during puberty.[1,4,12]

Although strongly different in terms of their electro-clinical characteristics and pathophysiology, BECTS and CAE have some common features such as a marked genetic predisposition, a similar age of onset, a normal neurodevelopmental profile, a normal background activity at EEG, normal neuroimaging findings and an overall good prognosis. Based on the observation of cases with concomitant clinical and EEG characteristics consistent with both rolandic and AS, some authors have speculated about a possible clinical crossover in these childhood epilepsies.[13–17]

Previous studies described the occurrence of occasional generalized epileptiform discharges in BECTS patients[18] but the coexistence of AS and rolandic seizures in the same patient has been quite rarely reported.[5] Although in the majority of published cases electro-clinical BECTS features precede the occurrence of AS, the presence of seizures with a generalized onset and a focal electro-clinical evolution over time, or the presence of contemporary features of both syndromes has also been reported, but the long term prognosis is not clearly established.[4,13–16]

The present study aims to describe the electro-clinical characteristics of a group of paediatric patients with a history of contemporary or subsequent diagnosis of CAE and BECTS with particular reference to the long term prognosis.

2. *Material and methods*

The medical records of all paediatric patients referred to 8 epilepsy centres in Italy and one in the USA between January 2000 and December 2015, were retrospectively evaluated. A systematic review of all clinical records was conducted and 11 subjects were identified. All records were reviewed by expert neuropaediatricians who also accessed the research databases for archived information.

All patients included in the study met the following inclusion criteria: 1) both concomitant or consecutive BECTS and CAE diagnosed according to the ILAE Classification[1]; 2) data concerning follow-up >2 years from the first observation available for each patient.

For each selected patient the following data were recorded and examined: age, gender, family history of febrile seizures and epilepsy, interictal and ictal (only for AS) EEG features, contemporary or consecutive clinical semiology, history of AEDs medications, therapeutic response and outcome.

Prolonged digital Video-EEG was recorded according to the International 10–20 scalp-electrode scalp placement system. Clinical and EEG details (interictal EEG recordings including sleep and recorded AS), were reviewed and anonymously agreed upon by all authors.

According to each Epilepsy Centre diagnostic protocol, brain MRI scans were obtained in nine patients and resulted normal.

3. Results

We identified 11 subjects, 5 females and 6 males, with a consecutive or contemporary coexistence of CAE and BECTS. Patient's age ranged between 7.8 and 17.3 years (mean age: 11.6 ± 3.5). Age at seizure onset ranged between 3.6 and 13.2 years. The patients' general characteristics are summarised in Table 1.

Seven (64%) patients had personal history for febrile seizures and five (45%) patients had a positive family history for febrile seizures, whereas only two patients (18%) had a positive family history for epilepsy (neither BECTS nor CAE). In one patient, concomitant migraine was reported. Four out of 11 patients presented concomitant features of both syndromes whereas the remaining 7 patients experienced the two syndromes at different times. In particular, AS was the first epileptic manifestation in 5 patients, whereas rolandic features were reported at onset in the remaining 2 subjects.

In all patients anticonvulsant therapy, reported in detail in Table 1, was started exclusively after the appearance of electro-clinical features consistent with CAE; those patients who presented BECTS features before CAE were not treated until the diagnosis of CAE.