Seizure 22 (2013) 565–571

Contents lists available at SciVerse ScienceDirect

Seizure

journal homepage: www.elsevier.com/locate/yseiz

Encephalopathy with hemi-status epilepticus during sleep or hemi-continuous spikes and waves during slow sleep syndrome: A study of 21 patients

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ARTICLE INFO

Article history:
Received 13 December 2012
Received in revised form 4 April 2013
Accepted 9 April 2013

Keywords:
Polymicrogyria
ESES/CSWSS syndrome
Symptomatic/structural and non-idiopathic
Children
Treatment

ABSTRACT

Purpose: To retrospectively analyze the electroclinical features, etiology, treatment, and prognosis of 21 patients with encephalopathy with hemi-status epilepticus during sleep (ESES) or hemi-continuous spikes and waves during slow sleep (CSWSS) syndrome.
Methods: Charts of 21 patients with hemi-ESES/CSWSS syndrome followed between 1997 and 2012 were analyzed. Inclusion criteria were: (1) Focal seizures or apparently generalized seizures and focal EEG epileptiform discharges; (2) Further occurrence of atypical absences, and myoclonic, atonic, and/or generalized seizures; (3) Cognitive impairment and/or behavioral disturbances; (4) Hemi-continuous spike-and-wave discharges during slow sleep in more than 85% of non-REM sleep at onset and throughout the ESES/CSWSS period.
Results: Mean follow-up from onset of hemi-ESES/CSWSS was 8 years (range, 2–15 years). Idiopathic cases were not identified. Unilateral polymicrogyria was found in 11, shunted hydrocephalus in four, a porencephalic cyst associated with polymicrogyria in three, and a thalamic lesion in three children. All started with focal seizures with or without secondary generalization. During the hemi-ESES/CSWSS period, all children developed new types of seizure, such as negative and positive myoclonus, absences, motor deterioration, cognitive impairment, and behavioral disturbances. All AED responders returned to baseline cognitive development. Seven patients were refractory to AEDs.
Conclusion: Our study suggests that the hemi-ESES/CSWSS syndrome has electroclinical features compatible with an epileptic encephalopathy. The most commonly used treatments were clobazam, ethosuximide, and sulthiame, alone or in combination. In refractory cases, high-dose corticosteroids were administered. Although the number of patients in this study is too low to draw definite conclusions, we consider that in children with hemi-ESES/CSWSS secondary to a unilateral lesion, surgery should be considered.

1. Introduction

Electrical status epilepticus during sleep was first reported in six children in 1971 by Patry et al.[1] Subsequently, Tassinari and colleagues introduced the term encephalopathy related to electrical status epilepticus during sleep (ESES) for the phenomenon. The term continuous spikes and waves during slow sleep (CSWSS)was used as a synonym.[2] Here we will use the term ESES/CSWSS to describe the EEG pattern and the clinical features of the entity.

The clinical spectrum of and guidelines for the electroencephalography (EEG) of the ESES/CSWSS syndrome have recently been published.[3–5] The syndrome may be diagnosed when ESES/CSWSS occurs in more than 85% of non-REM sleep, however, many other authors have used different cut-off rates[5–6] and the classification of the ILAE does not specify the cut-off value.[7] Several authors have described isolated cases with hemi-ESES/CSWSS based on the distribution of epileptiform activity during sleep.[8–14]

From the clinical point of view, deterioration of one or more cognitive functions with or without motor, behavioral, and/or psychomotor decline has been described in children associated with ESES/CSWSS.[15,16] ESES/CSWSS may be responsible not only

Abbreviations: PMG, polymicrogyria; PC, porencephalic cyst; FMS, focal motor seizures; CFS, complex focal seizures; SGTCS, secondarily generalized tonic-clonic seizures; FSE, focal status epilepticus; AA, atypical absences.

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http://dx.doi.org/10.1016/j.seizure.2013.04.006

for acquired aphasia, but also, and often concomitantly, for other dysfunctions, such as severe behavioral disturbances, apraxia, and negative myoclonus.[15] Idiopathic cases and patients with benign childhood epilepsies associated with the ESES/CSWSS syndrome have been published.[17–19] The syndrome may occur in children with organic brain lesions, such as unilateral polymicrogyria (PMG), hydrocephalus, and thalamic lesions.[13,20–25]

Treatment of the ESES/CSWSS syndrome has frequently been disappointing. Classic and new AEDs have been switched to benzodiazepines and ethosuximide.[17] An association of valproic acid and ethosuximide is still favored by different groups.[17,19,23] Sulthiame and levetiracetam have also been used.[26–29] In refractory cases, therapeutic alternatives such as corticosteroids, gamma-globulins, the ketogenic diet, and surgery have been tried.[30,31]

There have been numerous descriptions of patients with unilateral ESES/CSWSS, as isolated in a larger series or sometimes compared to patients with bilateral ESES/CSWSS.[3,4,6,13,20,27,32–37] However, they were not included in a well-designed methodological study analyzing only patients with unilateral ESES/CSWSS.

The aim of this study was to describe the electroclinical features, etiology, treatment, and prognosis of 21 patients with the hemi-ESES/CSWSS syndrome.

2. Methods

We conducted a retrospective, descriptive study of 21 consecutive patients with hemi-ESES/CSWSS syndrome followed between 1997 and 2012. All of them met the following inclusion criteria: (1) Onset with focal or apparently generalized seizures and focal EEG discharges; (2) Further appearance of atypical absences, and myoclonic, atonic (with or without epileptic falls), and/or generalized seizures; (3) Cognitive impairment and/or behavioral disturbances related to the hemi-ESES/CSWSS period; (4) Hemi-ESES/CSWSS occurring in more than 85% of non-REM sleep, at onset and during the ESES/CSWSS period.[2,5] Patients who started with diffuse ESES/CSWSS and then evolved into hemi-ESES/CSWSS were excluded.

Patients with other epileptic encephalopathies (e.g. Landau–Kleffner syndrome, myoclonic astatic epilepsy with cognitive deterioration, and Lennox–Gastaut syndrome) and focal epilepsies with secondary bilateral synchronies not fulfilling the criteria for the hemi-ESES/CSWSS syndrome were also excluded.

In this study all patients underwent at least three prolonged sleep EEGs (more than one hour), and 12 of these 21 patients underwent all-night video-EEG in addition to the routine EEG recordings. The spike-wave index (SWI) on the non-REM sleep EEG during the hemi-ESES/CSWSS period was visually calculated based on the total number of spike-waves per unit of time or based on the information provided by the epileptologist in the clinical charts. Nevertheless, the fact that different epileptologists may have used different methods for calculation of the SWI based on different reports in the literature[2,5] may have caused a certain heterogeneity in the results. The term ESES/CSWSS used here includes both the EEG pattern and the clinical features.

Brain magnetic resonance imaging (MRI) was obtained in all patients. Prolonged sleep EEGs were repeated two or more times per year. Data on school achievements and neuropsychological evaluations (Terman-Merrill or WISC III or IV) were repeatedly obtained during the follow-up of 2 to 15 years. In the absence of formal neuropsychological tests, based on the obtained information the degree of cognitive changes was evaluated according to clinical judgment in seven patients.

The onset and the resolution of the hemi-ESES/CSWSS phase were defined as the time of onset of cognitive impairment and behavioral abnormalities and the time of significant functional improvement which coincided in all cases with the time of the onset and offset of the pattern on the EEG.[38,39]

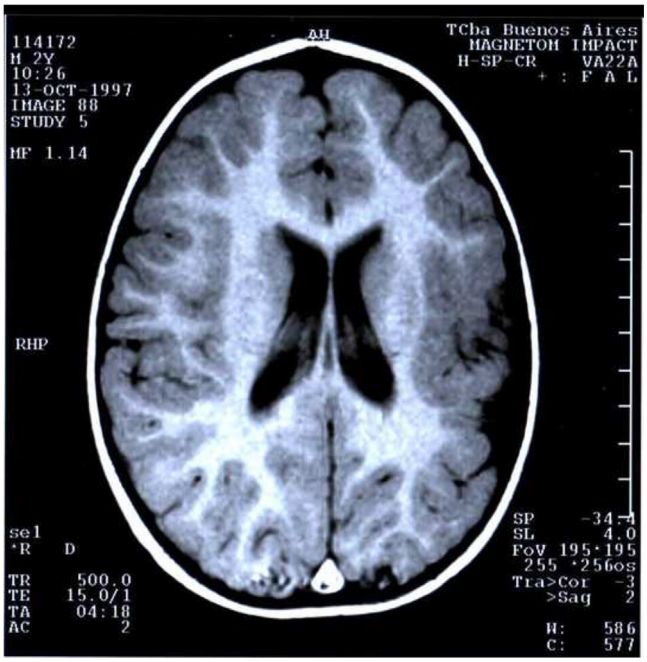

Fig. 1. Axial T1-weighted brain MRI showing extensive left PMG with atrophy of the ipsilateral hemisphere in a 10-year-old boy.

Biochemical controls, urine analysis, and plasma levels of classic AEDs were studied. The AEDs before and during the hemi-ESES/CSWSS phase were analyzed, as well as other treatment options such as corticosteroids, gamma-globulins, the ketogenic diet, and surgical treatments during the hemi-ESES/CSWSS period.

Determination of efficacy of the AED treatment was based on electroclinical criteria. Response of EEG abnormalities was graded as a normal recording, a more than 75% improvement of the SWI, a more than 50% improvement of the SWI, a more than 30% improvement of the SWI, focalization of the continuous generalized paroxysms, and no response. The percentages of improvement of SWI were related to the baseline SWI, e.g., a 50% improvement meant a decrease from 80% to 30%. Clinical response was considered as the decrease of cognitive and behavioral disturbances. The clinical response was graded as complete disappearance of clinical manifestations observed during the hemi-ESES/CSWSS phase, and clinical improvement of more than 75%, of more than 50%, or of more than 30%. It was a rough estimation of severity based on the clinical data (Figs. 1–3)

3. Results

3.1. General characteristics

A total of 21 children (14 boys and 7 girls) with the hemi-ESES/CSWSS syndrome were identified between March 1997 and April 2010 at the Garrahan Hospital of Buenos Aires.

Regarding probable etiology, 11 children had PMG, and four presented with shunted hydrocephalus, three had porencephalic lesions associated with PMG, and three had a thalamic lesion. PMG was localized in the frontotemporal lobes in seven patients, in the parietooccipital lobes in two, and in the whole hemisphere in two other patients. The brain magnetic resonance imaging showed unilateral involvement in all cases except in three patients with thalamic lesions and one with hydrocephalus. From a syndromic point of view, all patients had cerebral palsy, 17 of which presented