ELSEVIER

Brain & Development 31 (2009) 148–157

BRAIN & DEVELOPMENT
Official Journal of the Japanese Society of Child Neurology

www.elsevier.com/locate/braindev

Original article

Complex partial status epilepticus in children with epilepsy

Kenichi Kikumoto *, Harumi Yoshinaga, Katsuhiro Kobayashi, Makio Oka, Yoko Ohtsuka

Department of Child Neurology, Okayama University Graduate School of Medicine, Dentistry and Pharmaceutical Sciences, 2-5-1 Shikata-cho, Okayama 700-8558, Japan

Received 9 December 2007; received in revised form 28 March 2008; accepted 28 April 2008

Abstract

Purpose: Complex partial status epilepticus (CPSE) is often under-diagnosed, especially in children. The aim of this study was to clarify the characteristics and pathophysiology of CPSE in children with epilepsy. *Subjects and methods:* We retrospectively reviewed the medical records and EEGs of 17 children with epilepsy who were diagnosed as having CPSE by ictal or postictal EEGs to investigate clinical and EEG features. *Results:* The ages at diagnosis of CPSE ranged from 3 months to 17 years. At the time of diagnosis of CPSE, 13 patients had symptomatic localization-related epilepsy, two had epilepsy with continuous spike-waves during slow wave sleep, and each patient had cryptogenic localization-related epilepsy and idiopathic localization-related epilepsy. Only subtle symptoms including autonomic ones associated with disturbance of consciousness were the main clinical features in 12 of 44 CPSE episodes. Another 22 episodes showed minor focal motor elements, and the other 10 had major convulsive phase during or immediately before CPSE. Ictal EEGs of CPSE were divided into three types according to the degree of high-voltage slow waves (HVS) and spike components. Ictal EEGs could show spike-dominant or spike and HVS mixed patterns even if patients showed only subtle symptoms. The epileptogenic areas estimated by the ictal or postictal EEGs showed variability with only two cases of temporal origin. *Conclusion:* The close observation of clinical symptoms such as various subtle symptoms and/or mild convulsive elements and ictal EEGs are absolutely needed for the diagnosis of CPSE in children.

Keywords: Complex partial status epilepticus; Nonconvulsive status epilepticus; Status epilepticus; Ictal EEG; Children

1. Introduction

Status epilepticus (SE) is defined as the state in which clinical and electroencephalographic (EEG) epileptic activity persists for 30 min or more [1–3]. SE is divided into two major categories based on clinical symptoms, namely convulsive SE (CSE) and nonconvulsive SE (NCSE). Both types of SE are associated with significant morbidity and mortality. Therefore immediate diagnosis and medical intervention are absolutely needed. However, NCSE is often under-diagnosed compared with CSE, because its cardinal symptoms are changes of consciousness and behavior, with a lack of major motor phenomena. In particular, it would be conceivably difficult to diagnose NCSE in pediatric patients [4]. Absence status epilepticus and complex partial status epilepticus (CPSE) are two representative types of NCSE [5,6]. In contrast to the relatively well-defined absence status, CPSE has not yet been systematically studied, especially in children. CPSE in adults has been well described by Shorvon and Treiman and Delgado-Escueta [5,7]. A few studies on pediatric patients with CPSE were conducted by McBride et al. and Wakai et al., but both of them included only a limited number of cases [8,9]. Recently, Tay et al. performed a detailed investigation on 19 pediatric cases of NCSE including CPSE [10].

* Corresponding author. Tel.: +81 86 235 7372; fax: +81 86 235 7377.
E-mail address: kkikumoto@mac.com (K. Kikumoto).

0387-7604/$ - see front matter © 2008 Elsevier B.V. All rights reserved.
doi:10.1016/j.braindev.2008.04.009

Their patients had been diagnosed as having CPSE with acute symptomatic causes such as encephalitis.

On the other hand, even in usual outpatient clinics, we have sometimes diagnosed emergency cases of CPSE in children with epilepsy by ictal EEG examinations. CPSE can be deceptively mild, and easily misinterpreted as nonepileptic behavioral changes. Therefore, we believe that the frequency of CPSE might be higher than currently understood. The aim of this study was to clarify the characteristics and pathophysiology of CPSE in children with epilepsy by analyzing clinical features, EEG findings, and prognosis of CPSE.

2. Patients and methods

The subjects were selected from patients less than 18 years of age who visited the Department of Child Neurology Okayama University Hospital between January 1997 and December 2005, and were diagnosed as having CPSE by child neurologists. We retrospectively reviewed the medical records and EEGs of these patients. In this study, we limited the subjects to those with the so-called continuous form of CPSE defined by Gastaut as "status featuring a continuous, long-lasting episode of mental confusion with or without automatic behavior and psychosensorial or psychoaffective phenomena" [2,11].

We excluded the other type of CPSE, namely the discontinuous form from our study because accurate diagnosis is difficult in pediatric patients. All the patients underwent diagnostic EEG during and/or immediately after SE and we included only patients who had ictal patterns or postictal patterns both of which showed definite asymmetrical change. We excluded patients who were diagnosed as having acute symptomatic status epilepticus caused by acute cerebral insults such as encephalitis and encephalopathy. As a result, 17 patients (nine males and eight females) were involved in this study. The EEGs were recorded by using the Nihon Koden Neurofax (EEG-1518, EEG-4542) or the NEC SYNAFIT 1000 with a high-cut filter 120 Hz and a time constant of 0.1 or 0.3. The electrodes were applied according to the international 10–20 system referenced to the ears. Two child neurologists (K.K. and H.Y.) independently evaluated the EEG records. At any disagreement, they were to be advised by a third child neurologist (Y.O.) with no information on clinical background in effort to reach a final decision. We analyzed etiologies, ictal symptoms, EEG findings, and prognosis of CPSE.

3. Results

3.1. Patient demographics and etiologies of epilepsy

Forty-four episodes of CPSE had been observed in 17 patients by the time of survey from their onset of epilepsy. The profiles of the patients are summarized in Table 1. Cerebrovascular disorder was the most common etiology of epilepsy (five of 17). Other various etiologies were observed as shown in Table 1. Two patients were considered to be cryptogenic or idiopathic. No patient had acute provoking events directly related to the occurrence of CPSE.

3.2. Clinical courses

The ages at onset of epilepsy are shown in Table 1. Nine of the patients were less than 1-year-old, with a median age of 8 months at onset. The ages at diagnosis of CPSE ranged from 3 months to 17 years, with a median age of 5 years and 8 months. In one of the 17 patients, CPSE was his initial symptom of epilepsy. Seven patients experienced two or more episodes of CPSE. In six of these seven patients, CPSE occurred accumulatively in certain periods. Six patients experienced CSE as well as CPSE. The classifications of epileptic syndromes at the time of diagnosis of CPSE were as follows: symptomatic localization-related epilepsy in 13, epilepsy with continuous spike-waves during slow sleep (ECSWS) in two, cryptogenic localization-related epilepsy in one, and idiopathic localization-related epilepsy in one. The evolution of epileptic syndromes was seen in four patients (Table 1). In particular, three patients suffered from West syndrome during their clinical courses. CPS was the only habitual seizure type in nine patients. In the other eight patients, the following types of seizure besides CPS were observed. Afebrile generalized tonic–clonic seizures (GTC) were observed in two patients, febrile GTC in one, febrile and afebrile GTC in one, tonic spasms in three, and atypical absence in one. We present a patient as an example who showed a peculiar clinical course.

The patient (case 6) was a 5-year-old girl at the time of diagnosis of CPSE. At 3 months of age, she experienced her initial partial seizure. MRI revealed polymicrogyria in the left frontal lobe. Carbamazepine was initiated, but it was not effective. At 2 years and 1 month of age, her EEG developed into continuous spike-waves during slow wave sleep (CSWS) as shown in Fig. 1a. At 3 years and 11 months of age, she began to have atypical absences (Fig. 1b). She also experienced frequent episodes of atypical absence status epilepticus. After synthetic adrenocorticotropic hormone therapy her atypical absences decreased to less than several times a day. At 5 years of age, she became unresponsive to questions associated with repeated vomiting and deviation of her eyes. When she arrived at our hospital, we first suspected atypical absence status but then reached diagnosis of CPSE based on her ictal EEG finding (Fig. 2a). By an intravenous diazepam (DZP) infusion under EEG monitoring, the ictal pattern on the EEG was sup-