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Original article

Electro-clinical-etiological associations of epilepsia partialis continua in 57 Chinese children

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Abstract

Objective: Epilepsia partialis continua (EPC) was one type of focal status epilepticus. The aim of this study was to analyze the clinical and electroencephalography (EEG) characteristics, and outcome of 57 child-onset patients with EPC according to different etiologies, and further explore the electro-clinical-etiological associations.

Methods: We retrospectively reviewed 57 children diagnosed with EPC in our department over last ten years. Etiology, clinical and EEG data, and outcome were categorized and analyzed.

Results: For the 57 child-onset patients, EPC was caused by different etiologies, including immune-related disease (43.9%), focal lesions (17.5%), inborn errors of metabolism (24.6%), and unknown (14.0%). EEG background abnormalities showed generalized slowing in 45 patients (78.9%) and focal slowing in two patients (3.5%). Nineteen patients (33.3%) presented clear correlation of ictal EEG/EMG and the remaining 38 patients (66.7%) showed no clear correlation of ictal EEG/EMG. Both EEG background activity and ictal EEG/EMG correspondence among different etiologies had statistical significance ($P < 0.05$). The ictal patterns without clear EEG/EMG correspondence in immune-related disease and the ictal patterns with clear EEG/EMG correspondence in focal lesions were more prominent ($P < 0.05$).

Conclusion: This is the first study of child-onset EPC with a large series in a pediatric epilepsy center in China. The most common cause for EPC was immune-related disease. The EEG background activity and the EEG/EMG correspondence might be influenced by the etiologies of EPC to some degree. These findings might guide the direction of EPC diagnosis in conjunction with other examinations.

Keywords: Epilepsia partialis continua; Children; Etiology; Clinical; Electroencephalography

1. Introduction

Epilepsia partialis continua (EPC), a seizure type with focal motor symptoms, was classified as a type of focal status epilepticus by the International League Against Epilepsy (ILAE) in 2001 [1]. It was defined as continuous regular or irregular muscle jerks of cortical origin occasionally aggravated by action or sensory stimuli confined to a part of the body. These occurred for a minimum of one hour and could reoccur in intervals of 10 s or less. The seizure often lasted for several hours, days, weeks, or even much longer [2,3]. EPC could develop at any age. Rasmussen syndrome (RS) and mitochondrial disease were the most common etiologies of child-onset EPC, while adult-onset EPC

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were common in brain tumor and cerebrovascular disease [4]. The clinical manifestation, treatment and prognosis of EPC varied widely depending on the underlying etiology.

Previous studies suggested that EPC was originated in the primary motor cortex (Brodmann's areas four and six), although the subcortical structures had been considered as another possible generator but still lack of strong evidence [2,5]. In some patients, however, the myoclonic jerks of EPC were not always accompanied by correlated discharges on surface electroencephalography (EEG) [6–8]. This phenomenon led to the diagnostic difficulties in the absence of further examination. In 2006, the ILAE reported that the clinical and EEG features of EPC might point to different types of etiology [9]. Herein, we analyzed the clinical and EEG features, and outcome of 57 child-onset patients with EPC according to different etiologies, and particularly focused on the electro-clinical-etiological associations, thus finding possible indicators to guide the etiological diagnosis of EPC.

2. Materials and methods

We retrospectively reviewed 57 children diagnosed with EPC from June 2005 to December 2015 in the Pediatrics Department, Peking University First Hospital. This institution is a major pediatric epilepsy center for the diagnosis and treatment of patients with epilepsy from all over the country, particularly children with intractable epilepsy. This study was approved by the Ethics Committee of Peking University First Hospital. Informed consent was obtained from patients and their parents. The following inclusion criteria were included: the standard of EPC was combined the definition of Thomas et al. [2] and Obeso et al. [3]; each of patient must presented EPC event and be recorded EPC at least once by video-EEG (VEEG) with electromyography (EMG) in our department; the age of EPC onset was less than 15 years old.

2.1. Scalp VEEG recording

EEG monitoring was performed using a Nihon Kohden digital 1100 K VEEG instrument. Electrodes were placed according to the 10–20 international system for electrode placement. Tests for waking and sleep status about four hours were included in the EEG examination. To examine the stimuli sensitive, the patients were instructed to keep speaking, maintain a certain posture for a while, and receive auditory, tactile and visual stimuli etc. during the EEG monitoring. EMG signals were simultaneously recorded by placing a pair of disc electrodes about three cm apart on the skin overlying each muscle. Two to six muscles were recorded in each of the patients. In a small muscle like the adductor pollicis brevis, only one electrode could be placed over the muscle while another electrode was placed on the tendon.

2.2. Etiology related examinations

All of these patients were done brain magnetic resonance imaging (MRI). Blood samples were all performed for complete blood count, biochemical studies including lactate, pyruvate, ammonia and homocysteine etc., plasma amino acids and acylcarnitine spectrum analysis, and urine amino acids and organic acids analysis. Characteristic antibodies, oligoclonal bands and IgG index of cerebrospinal fluid (CSF) were tested in those patients with suspected immune-related encephalitis. Viral antibody titers of blood and CSF were studied in those with suspected viral encephalitis. Genetic testing including point mutation in mitochondrial genome (A3243G, A8344G, T8993G/C) and gene mutation in *PLOG1* was performed in those with suspected mitochondrial encephalomyopathy with lactic acidosis and stroke like episodes (MELAS) and Alpers syndrome respectively. Pathology studies of skin, muscle, peripheral nerve and brain biopsy, and somatosensory evoked potential (SSEP) were conducted in selected patients.

2.3. Data acquisition and etiology diagnostic criteria

All of these patients' data were obtained from medical records and computerized EEG databases, and supplementary information and recent data were collected by means of outpatient or telephone follow-up. At least two pediatric neurologists performed the etiology diagnosis. RS was defined according to the diagnostic criteria put forward by Bien et al. [10]. The diagnostic criteria of MELAS consulted the report by Lorenzoni et al. [11]. The diagnosis of Alpers syndrome referred to the report by Bao et al. [12]. The EEG data were analyzed independently by two EEG evaluators with more than 10-year experience who were blinded to the clinical data, and the results were in agreement. The myoclonic jerks of cortical origin were defined as an extremely short duration of the EMG correlates, usually less than 50 ms [13]. We defined that a clear ictal EEG/EMG correspondence was at least 80% of EMG bursts representing myoclonic jerks corresponded to EEG discharges, otherwise, defined as without a clear correspondence.

2.4. Classification of etiology and outcome

For the purpose of this study, the etiology of EPC was divided into four categories. Category one: immune-related disease (evidence of immune-mediated central nervous system inflammation), category two: focal lesions (a distinct and relatively static structural lesion in the cerebral cortex), category three: inborn errors of metabolism (various conditions with genetic