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Original article

Five pediatric cases of ictal fear with variable outcomes

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Abstract

Purpose: Ictal fear is an uncommon condition in which fear manifests as the main feature of epileptic seizures. The literature has suggested that ictal fear is generally associated with poor seizure outcomes. We wanted to clarify the variability in seizure outcome of children with ictal fear. *Subjects and methods:* We identified five pediatric patients with ictal fear who were followed up on at Okayama University Hospital between January 2003 and December 2012. We retrospectively reviewed their clinical records and EEG findings. *Results:* The onset age of epilepsy ranged from 8 months to 9 years and 10 months. The common ictal symptoms were sudden fright, clinging to someone nearby, and subsequent impairment of consciousness, which were often accompanied by complex visual hallucinations and psychosis-like complaints. Ictal fear, in four patients, was perceived as a nonepileptic disorder by their parents. Ictal electroencephalograms (EEG) of ictal fear were obtained in all patients. Three showed frontal onset, while the other two showed centrotemporal or occipital onsets. Two patients were seizure free at last follow-up, while seizures persisted in the other three. A patient with seizure onset during infancy had a favorable outcome, which was considered to be compatible with benign partial epilepsy with affective symptoms. *Conclusion:* Ictal fear is not always associated with a symptomatic cause or a poor seizure outcome. It is quite important to make a correct diagnosis of ictal fear as early as possible to optimize treatment.

Keywords: Ictal fear; Childhood; Benign partial epilepsy with affective symptoms; Frontal lobe epilepsy

1. Introduction

Ictal fear is an uncommon condition in which fear manifests as the main feature of epileptic seizures. Partial seizures presenting with fear are generally considered to originate in the temporal or frontal lobe [1,2]. Ictal fear is 'unprovoked fear without object of dread' which spontaneously appears associated with epileptic discharges which involve neural networks that include the limbic structures responsible for emotions [3]. It has been empirically shown that not only 'pure' ictal fear but also ictal fear with fearful hallucinations exist [4].

Children with ictal fear are frequently misdiagnosed as having sleep or psychiatric disorders, such as pavor nocturnus, panic attacks and psychogenic reactions [5,6]. Cases reported in the literature show that for most patients with ictal fear, their epilepsy was symptomatic and intractable. We report on five children with ictal fear confirmed by ictal EEGs whose outcomes were more variable than expected.

2. Subjects and methods

We identified five pediatric patients with ictal fear who were followed up on at Okayama University Hospital between January 2003 and December 2012.

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The details of patient #4 were reported elsewhere [7]. The inclusion criteria for ictal fear in this study were epileptic seizures which present fear as a prominent symptom and in which fear is expressed with facial expressions, behavioral changes (i.e. running away, intense crying, clinging to someone nearby), and/or verbal descriptions.

All patients underwent routine EEGs at Okayama University Hospital. Four had ictal EEGs recorded at Okayama University Hospital and one at a different hospital. EEGs were recorded during waking and sleep for more than 40 min using Neurofax (Nihon Kohden, Tokyo, Japan) according to the International 10–20 Electrode Placement System. The follow-up periods were more than 5 years in all patients. We retrospectively reviewed their clinical records and EEG findings.

3. Results

3.1. Case report

Patient #1 is a girl with normal psychomotor development at seizure onset. Her gestation and delivery were uneventful. There was no family history of seizure disorders. At the age of 8 months, she started having seizures consisting of generalized atonia and unresponsiveness with cyanosis and eye fixation, lasting approximately 3 min. She was referred to Okayama University Hospital after having several seizures. Physical and neurological examinations were unremarkable. Her head CT scan and interictal EEG were normal. She was clinically diagnosed as having epilepsy and treated with phenobarbital. Two months later, she experienced three afebrile generalized tonic–clonic seizures in a day. An increase in phenobarbital dosage suppressed her seizures. At the age of 1 year and 5 months, she started having events characterized by sudden intense crying for her mother without any provocation, clinging to someone nearby with a frightened expression, looking around uneasily and a subsequent mild cloudiness of consciousness. These events lasted 1–5 min. Although she was able to respond and walk at approximately 2 min after onset, once or twice she showed the unusual behavior of seeking her mother even when her mother was beside her. She fully recovered within 5 min of onset.

At the age of 2 years and 1 month, while suffering from a cold, she had three similar events within a few hours. On her arrival at Okayama University Hospital, she was alert and her neurological examination was normal. She had another event during an EEG study on the same day. The ictal EEG demonstrated irregular 2–3 Hz delta activity originating from the bilateral occipital head regions, which appeared approximately 10 s prior to the clinical symptoms and built up and spread to the bilateral central head regions (Fig. 1). As the events were confirmed to be partial seizures, carbamazepine was started and her seizures have been completely suppressed since then. Later on, her interictal EEG showed sporadic spike–wave complexes with bilateral occipital dominance (Fig. 2). Carbamazepine was discontinued at the age of six and she remained seizure-free at last follow-up.

3.2. Other results

The clinical characteristics of the five patients are summarized in Table 1. The onset age of epilepsy ranged from 8 months to 9 years and 10 months.

Patient #5 had a febrile status epilepticus with right hemiconvulsion at the age of 5 years and was diagnosed as having acute encephalopathy. She developed moderate mental retardation and autistic behavior as a result of acute encephalopathy. The causes of epilepsy in the other four patients were unknown, and three of them continued to show normal intelligence. Patient #4 developed mild mental retardation, attention deficit/hyperactivity disorder, and severe conduct disorder after the onset of epilepsy.

Four patients had ictal fear at the onset of epilepsy. Patient #1, presented in the above case report, initially had ictal syncope. Patient #3 experienced secondarily generalized seizures concurrently with ictal fear. Patient #5 developed ictal fear followed by secondarily generalized seizures at 9 months after acute encephalopathy.

The common feature of ictal fear was sudden fright and clinging to someone nearby, followed by impairment of consciousness. Patient #1 showed unusual behavior that resembled transient blindness or postictal confusion following fear expressions. Patient #2 complained of dizziness and intense fear at the same time. Two patients (patients #3 and #4) had fear sensation, complex visual hallucinations and psychosis-like complaints simultaneously. In patient #5, fear expression was the sole manifestation. All patients experienced clusters of seizures. Two of them (patients #4 and #5) had status epilepticus many times and required hospitalization.

Although patient #1 showed no epileptic abnormalities on the her first EEG, subsequent EEGs showed spike–wave complexes with bilateral occipital dominance. The other four patients showed frontal spikes. Two of them (patients #2 and #5) had few spikes throughout their courses. Although the remaining two (patients #3 and #4) showed frequent frontal spike-waves intermingled with frontal background slowing, especially during the periods of frequent seizures, they did not have frequent spikes in their baseline states.

In patient #1, an ictal EEG of a seizure presenting with fear showed occipital seizure onset. Three patients had frontal seizure onset, while the remaining patient had centrotemporal seizure onset. In three patients (patients #2, #3 and #4) with frontal seizure onset, ictal