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Jeavons syndrome in China

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ABSTRACT

Objectives: Jeavons syndrome (JS) is one of the underreported epileptic syndromes and is characterized by eyelid myoclonia (EM), eye closure-induced seizures or electroencephalography (EEG) paroxysms, and photosensitivity. In the Western populations, it has been reported to be characterized by focal posterior, occipital predominant epileptiform discharges (OPEDs) or frontal predominant epileptiform discharges (FPEDs) followed by generalized EDs in both interictal and ictal EEG recordings. However, it is not clear if there are different clinical manifestations between OPEDs and FPEDs. The clinical and electrographic presentations in the Chinese population are largely unknown. Here, we report the clinical and electroencephalographic features of 50 Chinese patients with JS and evaluate for the presence of different clinical features between patients with OPEDs and patients with FPEDs.
Methods: We identified 50 cases who met the Jeavons syndrome criteria from 4230 patients with epilepsy at Xijing Hospital, Xi'an, China from the period of January 2010 to November 2011. These patients underwent long-term 24-hour video-EEG recording. Brain imaging was performed using magnetic resonance imaging (MRI) or computerized tomography (CT). Webster IQ testing was performed to determine intellectual development. We reviewed and described the interictal abnormalities, ictal EEG pattern, and demographic, clinical, and neuroimaging findings of these 50 Chinese patients in Xi'an. We divided the 50 patients into two groups according to the predominance of EDs and analyzed their clinical features.
Results: Twenty-five of these 50 patients were male. Twenty-two out of 32 patients in the group with FPEDs were male, and 3/18 patients in the group with OPEDs were male. The median age of EMA–EM onset in FPEDs was 8 years and that in OPEDs was 5.8 years. Eyelid myoclonia occurred in all the 50 patients. Twenty-one out of 32 patients in the group with FPEDs had EM with absences, and 14/32 of them had EM with eyeball rolling up. Two out of 18 patients in the group with OPEDs had EM with absences, and only 1 of 18 had EM with eyeball rolling up.
Conclusion: Eyelid myoclonia with or without absences or JS diagnosis is easily missed and underreported in China. As an IGE, either the frontal or the occipital lobe may initiate generalized spike-and-wave discharges (GSWDs) and generalized seizures (GSs). There may be two subtypes of JS with distinctive clinical and electroencephalogrphic features: a predominantly male group with frontal predominant epileptiform discharges, eyelid myoclonia, and eyes rolling up and a predominantly female group with occipital predominant epileptiform discharges with eyelid myoclonia alone.

1. Introduction

Eyelid myoclonia with or without absences (EMA-EM) has been recognized as a seizure type by the International League Against Epilepsy (ILAE) in their 'proposed diagnostic scheme for people with epileptic seizures and epilepsy'. This unique manifestation, confirmed as a separate entity of idiopathic generalized epilepsies (IGEs), was named 'Jeavons syndrome' (JS) [1–6]. However, the view of JS being an individual IGE has been challenged by some other authors who have argued that EMA-EM may occur in several epileptic conditions, such as idiopathic, cryptogenic, and symptomatic epilepsies; thus, its occurrence alone may not be sufficient to characterize a definite epilepsy syndrome [2]. Nevertheless, EMA-EM has been widely regarded as the typical seizure type of JS. In some Western countries, idiopathic EMA-EM (or JS) has been characterized as a long-lasting condition mainly developing in childhood and having a twofold higher incidence in girls than in boys [7,8]. The EEG presentation of JS has been described as focal occipital or frontal predominant epileptiform discharges in both interictal and ictal stages which may lead to generalized EDs [2,9]. To date,

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there have been few JS cases reported in China, and its features in a Chinese population are not clear. It is not certain if there are different clinical features between the predominance of EDs in JS. The objective of this study was to review the clinical and EEG features of Chinese patients with JS and to observe whether there were differences between the group with FPEDs and the group with OPEDs.

2. Methods

2.1. Standard protocol approvals, registrations, and patient consent

Ethical permission for scanning the patients was obtained from the Xijing Hospital Research Ethics Committee and individually from the patients' parents.

2.2. Patients

We identified 50 patients who met the diagnostic criteria of JS including EMA/EM, eye closure-induced seizures/EEG paroxysms, and photosensitivity out of 4230 patients with epilepsy between January 2010 and November 2011 at the EEG Department, Xijing Hospital. All these patients in the study underwent follow-up examination after 6 months to 2 years.

2.3. Video-EEG examination with photic stimulation

Twenty-four-hour video-EEG monitoring was performed on all the patients. Video-EEG was performed in an illuminated room by a 32-channel digital video-EEG system (NicVeu; NicoletOne; American and Nihon Kohden; Japan; Bio-logic; American) with scalp electrodes placed according to the international standard lead 10–20 system. The reference was placed at Cz′, a location 1 cm behind Cz. The sampling rate was 500 Hz with a band pass filter (high pass cutoff frequency at 0.3 Hz and low pass cutoff frequency at 70 Hz). Static, hyperventilation, or photic (3–23 Hz) stimulation EEG with passive or active eye closure and opening was monitored and recorded. Electromyography electrodes were placed over both deltoid muscles. Three technicians and one physician reviewed and analyzed the EEG and determined the onset and offset of interictal or ictal EDs. We analyzed focal or generalized interictal EDs during both asleep and awake stages. We also analyzed the ictal EEG features which appeared when patients presented with EM on the video monitor.

We performed intermittent photic stimulation (IPS) with frequencies that ranged from 3 Hz to 23 Hz with increments of 2 Hz. Intermittent photic stimulation was delivered to the patients with eyes open for 5 s, then with eyes closed for another 5 s; the interval was 10 s, then the next frequency was used. If the EDs or seizure was evoked, the IPS test was terminated immediately.

2.4. Neurological examination and brain imaging

Neurological examination and brain MRI or CT scan were performed in our 50 patients.

2.5. Cognitive function test

A Webster IQ test was performed in the Epilepsy Centre of Xijing Hospital in our 50 patients.

2.6. Grouping

We divided the 50 patients into two groups: (1) the group with frontal predominant EDs and (2) the group with occipital predominant EDs according to their ictal and interictal EEG characteristics. The frontal or the occipital ED predominance was determined when the EDs were consistent with one of the following criteria: interictal or ictal focal EDs arose from the frontal or the occipital region only; GSWDs had focal onset either from the frontal or the occipital region.

3. Results

3.1. Clinical characteristics

The first group comprised 32 patients who had frontal predominant EEG discharges (Table 1). Twenty-two of these patients were male. The median age of EMA–EM onset was 8 years (range: 5–17 years). All 32 patients had EM characterized by marked eyelid myoclonia with absences occurring in 21. Fourteen patients had eyes rolling up, and 20 patients had generalized tonic–clonic seizures (GTCSs) before commencing treatment.

The second group of 18 patients had occipital predominant EEG discharges. The gender distribution was different from that in the first group as 15 patients were female and only 3 were male. The median age of EMA–EM onset was 5.8 years (range: 2–13 years). All 18 patients had EM, 2 patients had absences, and only one patient had eyes rolling up. One female patient presented with eyelid myoclonic status epilepticus lasting 1–2 h. Five patients had GTCSs before commencing treatment, and the seizures mainly occurred when they woke up in the morning.

3.2. Interictal video-EEG findings

Aside from the one female patient with EM status epilepticus in the second group whose background EEG rhythm was slower than normal for children of her age, all patients had a normal posterior dominant alpha rhythm reactive to eye opening.

In the first group of 32 patients, interictal EEG showed generalized spike–wave or polyspike–wave discharges predominant in the frontal

Table 1
Clinical features and video-EEG findings of patients with Jeavons syndrome in China.

Patients with EM (n = 50)												
EEG characteristic (distribution of PEDs)	Number	Sex (F/M)	Mean age of EM onset (years)	Seizure type						Photic stimulation provoking EM and or PEDs		Hyperventilation provoking EM and or PEDs
				EM				EMSE	GTCS	With eyes open	On eye closure	
				With absence	Without absence	With eyes rolling up	Without eyes rolling up					
With frontal predominance	32	10/22	8 (range: 5–17 years)	21	11	14	18	0	20	5	32	18
With occipital predominance	18	15/3	5.8 (range: 2–13 years)	2	16	1	17	1	5	2	18	3

Abbreviations: ED: epileptic discharge without EM, EM: eyelid myoclonic, SWs: spike waves, PSWs: polyspike waves, GED: generalized epileptiform discharge, PEDs: paroxysmal epileptiform discharges, EMSE: eyelid myoclonic status epilepsy, GTCS: generalized tonic–clonic seizure.