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Eyelid myoclonia seizures in adults: An alternate look at the syndrome paradox

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ABSTRACT

Objectives: Eyelid myoclonia (EM), without or with absences (EMA), is induced by eye closure (ECL)-associated generalized paroxysms of polyspikes and waves. Although considered as an epileptic syndrome, it has been listed as a type of seizure in the recent epilepsy classifications, perhaps because of its clinical heterogeneity. In this study, we aimed to specifically study the clinical and electroencephalogram (EEG) features and the prognosis of long-term followed-up adult patients with EMs and to determine common points between EMAs, idiopathic generalized epilepsies (IGEs), and symptomatic epilepsies.
Methods: Between 1996 and November 2011, 61 adult patients with EMs with or without absences and bilateral EEG paroxysms were retrospectively enrolled in the study and followed up for 1–34 years (mean: 5.8 years).
Results: According to patient history, seizure semiology, and EEG findings, we classified the patients having EM seizures into three main groups. In group 1 ($n = 31$), all patients had prominent EMs with or without absences associated with upward rolling of eyeballs. The second group included 20 patients with EM seizures associated with generalized tonic–clonic seizures (GTCSs) and/or massive myoclonias. The third group of 7 patients had varying diagnosis of symptomatic epilepsies. In the first group with pure EMA, the diagnosis was more delayed than in the other groups ($p = 0.01$). In the group with pure EMA, EMs continued in adulthood ($p = 0.00$), and only 24% of patients were seizure-free, which was considered poor prognosis. On EEG, occipital ($n = 3$) and frontal ($n = 4$) focal discharges were found in the group with pure EMA. Interestingly, 2 patients with symptomatic epilepsy with frontal lesions also had EM seizures.
Conclusion: The patients with pure EMA have many similarities to patients with IGEs. We also demonstrated that EMs could be seen as a seizure type in symptomatic epilepsies. Eyelid myoclonia with absences meets the criteria for an epileptic syndrome with the early onset and long duration of seizures, special seizure type, specific EEG findings, possibility of cognitive impairment, precipitating modalities, photosensitivity, and presence of family history, suggesting a strong genetic background.

1. Introduction

Eyelid myoclonia without or with absences (EMA) was first documented in 1937 by Radovici and colleagues as a case report [1], and, subsequently, the main features of this condition were described by Jeavons in 1977 [2]. Although EMA is considered well defined with its clinical and electrophysiological features described by several authors [3–11], its place in the epilepsy classifications is still debated. Even in the latest ILAE (International League Against Epilepsy) classifications, EMA has been listed as a type of seizure [12–14].

EMA is characterized by fast and marked jerking of the eyelids with rolling of the eyes immediately after eye closure, accompanied sometimes by subtle loss of awareness. It is associated with a brief 3–6 Hz generalized discharges of spike-and-wave activity in the electroencephalogram (EEG). Eyelid myoclonias (EMs) are provoked by photic stimulation and are completely abolished when the eyes are closed in total darkness [2,15–18].

Although the first description of EMA states that 'EMA is a homogeneous epileptic syndrome and, if it is seen once, will never be forgotten' [16], there have been several patient reports showing different clinical and electrophysiological features. Hence, EMA has been included with other types of idiopathic generalized epilepsies (IGEs) [10,11,19–22].

In this study, we aimed to specifically study the clinical and EEG features and prognosis of our long-term followed-up adult patients with EMs and to determine common points between EMA, IGEs, and

Abbreviations: EM, eyelid myoclonia; EMA, eyelid myoclonia with absences; EEG, electroencephalogram; IGEs, idiopathic generalized epilepsies; ECL, eye closure; GTCSs, generalized tonic–clonic seizures; MM, massive myoclonia; ILAE, International League Against Epilepsy; VEMU, video-EEG monitoring unit; MRI, magnetic resonance imaging; ECLS, eye closure sensitivity; HPV, hyperventilation; IPS, intermittent photic stimulation; VPA, valproic acid; CLN, clonazepam; LTG, lamotrigine; LEV, levetiracetam; JME, juvenile myoclonic epilepsy; PME, progressive myoclonic epilepsy; PPRs, photoparoxysmal responses; PNESs, psychogenic nonepileptic seizures.

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symptomatic epilepsies based on the idea that the patients with EMs are a heterogeneous group.

2. Methods

We retrospectively collected EEG recordings displaying 3–5 Hz spike–wave and/or polyspike–wave discharges associated with EMs from our outpatient and video-EEG monitoring unit (VEMU) between 1996 and 2011. The inclusion criteria for EMs consisted of the following: (a) EMs seen in video-EEG, (b) and/or EMs noted down by the EEG technician or doctor during analog EEG, (c) and/or a history of eye blinking. We examined 61 patients having EMs with generalized EEG abnormalities.

All video-EEG recordings were investigated for interictal and ictal findings. Hyperventilation (HPV), intermittent photic stimulation (IPS), eye opening, eye and closing were used for activation. IPS was performed at least 3–4 min after HPV and in three different conditions of the eyes (eye closure, eyes closed, and eyes open) compatible with recommended methods [23,24]. Semiological findings including EMs and other eyelid movements, head movements, and other findings of generalized seizures were documented. Magnetic resonance imaging (MRI) was obtained for 46 patients and was reviewed retrospectively. Demographic data, neurological examinations, and clinical features were analyzed too.

3. Results

We analyzed 61 patients — 45 (74%) females and 16 (26%) males. Patients were followed up for 1–34 years (mean ± SD: 6 ± 7.51). The mean age of the patients was 27.4 ± 5.61 years. The mean age at seizure onset was 11.3 ± 6.13 years.

According to patient history, seizure semiology, and clinical and EEG findings, we classified the patients having eyelid myoclonias into three main groups:

Group 1: all patients had prominent EMs with or without absences associated with upward rolling of eyeballs (n = 31); group 2: patients had features that are similar to those of IGEs, including massive myoclonias on the extremities, absences, and generalized tonic–clonic seizures (GTCSs) (n = 20); and group 3: patients had features of symptomatic epilepsies according to MRI, EEG, and neurologic examination findings (n = 7).

The remaining 3 patients had eyelid myoclonias but were not included in any of the groups because of inadequate information.

3.1. Clinical features

All patients had EMs. All patients in group 1 had prominent EMs characterized with upward position of the cornea. Seven patients had GTCSs, and 6 patients had mild myoclonias limited to the upper body and head or neck in group 1. The patients in group 2 had massive myoclonias, GTCSs, or absence seizures that lasted for a long time. Focal seizures were only seen in group 3.

Seventeen of 31 (54.8%) patients in group 1 had a family history of epilepsy. Four (12.9%) of them had first-degree relatives with history of epilepsy. In group 2, seven (35%) patients had a family history of epilepsy.

Demographic findings, history of febrile convulsion, cognitive and psychiatric status, neuro-radiological findings, and other clinical features are shown in the Table 1.

3.2. EEG findings

The background EEG activity was slow only in 8 patients (6 patients in group 3 and 2 patients in group 1). The ictal and interictal EEG recordings of all patients revealed bilateral polyspike, 2.5–5 Hz spike–wave complex, or sharp wave complex. In the first group, the discharges appeared at a mean duration of 1–3 s and were longer in the second group (3–10 s). Interictal discharges were activated by hyperventilation (HPV) in 20 (64.5%) patients from group 1 and in 14 (70%) patients in group 2.

Eye closure sensitivity (ECLS) was recorded in 18 (58%) patients in the first group (Figs. 1a and b) and in 9 (45%) patients in the second group with a mean latency of 1–3 s. Fourteen (45.2%) patients in the first group and 8 (40%) patients in the second group were sensitive to intermittent photic stimulation (IPS).

In addition to the generalized discharges on EEG, there were patients with focal discharges. Focal EEG findings were seen in 8 patients in group 1, in 4 patients in group 2, and in 3 patients in group 3.

All EEG findings are summarized group-wise in Table 2.

3.3. Treatment and prognosis

The diagnosis of EMA was significantly more delayed (upto 10.3 years) in group 1 than in the other groups (p = 0.01). About half the patients in groups 1 and 2 were on polytherapy. The most commonly used drugs were valproic acid (VPA), clonazepam (CLN), lamotrigine (LTG), levetiracetam (LEV), and several combinations of these drugs. Twenty (69%) patients in group 1 and 9 (45%) patients in group 2 had taken carbamazepine (CBZ) and/or diphenylhydantoin (DPH), which are not recommended for the treatment of myoclonic type seizures.

Despite using appropriate and adequate AEDs, the seizures remained in 22 (76%) patients in group 1 and 16 (84%) patients in group 2. GTCS remained in 11 (38%) patients in group 1 and in 9 (47%) patients in group 2. However, there were significantly more patients (n = 17, 59%) in group 1 who had permanent EM seizures than in the other groups (p = 0.00).

4. Discussion

In our tertiary epilepsy center, we recruited 61 adult patients with EMs between 1996 and 2011.

Three main groups were formed according to seizure semiology and electrophysiological features.

Table 1
Clinical features of patients.

	Group 1 (n = 31)	Group 2 (n = 20)	Group 3 (n = 7)
Family history of epilepsy	17	7	3
Febrile convulsion	7	3	1
Age at onset (±SD)	10 ± 6.2	13 ± 5.7	8 ± 7.5
Delay of diagnosis (±SD)	10 ± 6.8	5 ± 7.5	6 ± 9
Other types of seizures	GTCSs (7), M (6), PNESs (7)	GTCSs (14), MM (12), As (15), PNESs (4)	GTCSs (2), MM (4), FSs (4)
Refractory to AED	GTCSs (11), EMs (17)	GTCSs (9), EMs (2)	GTCSs (4), EMs (1)
Mental impairment	CI (11), MR (4)	CI (9)	MR (4)
Psychiatric problems	13	9	–
MRI abnormalities	–	–	5

GTCSs: generalized tonic–clonic seizures, M: myoclonus, MM: massive myoclonus, As: absences, FSs: focal seizures, CI: cognitive impairment, MR: mental retardation, AED: antiepileptic drug, PNESs: psychogenic nonepileptic seizures, MRI: magnetic resonance imaging.