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CASE REPORT

Non-Epileptic paroxysmal eyelid movements associated with generalized epilepsies: a challenge in assessing seizure burden

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ABSTRACT

Background: Abnormal ocular movements may also mimic epileptic seizures, such as in paroxysmal tonic upward gaze, blepharospasm, ocular tics, and paroxysmal eyelid movements (PEM). From a diagnostic, management and prognostic perspective, it is important to undertake an accurate assessment of whether abnormal ocular movements are epileptic in origin or not.

Case presentation: We present a child from a tertiary pediatric epilepsy center in the United Kingdom with PEM associated with photoparoxysmal responses, which was a source of diagnostic confusion. EEG recordings captured the PEM episodes to confirm the diagnosis.

Conclusion: The presentation of children with epilepsy with ocular abnormal movements, particularly in the presence of photoparoxysmal responses, should raise suspicion of possible PEM. Confirmation is essential by capturing these events on EEG.

Keywords: Epilepsy, myoclonia, non-epileptic, EEG, case report.

Background

Abnormal movements of eyes and eyelids, such as in typical absences and absences with eyelid myoclonia, are clinical features of different types of epileptic seizures. Absences with eyelid myoclonia are considered a type of generalized epilepsy, under the International League Against Epilepsy (ILAE) ILAE Operational Classification of Seizure Types proposed in 2017 [1]. Abnormal ocular movements may also mimic epileptic seizures, such as in paroxysmal tonic upward gaze, blepharospasm, ocular tics, and paroxysmal eyelid movements (PEM). It is, therefore, important from a diagnostic, management and prognostic perspective to make an accurate assessment of whether abnormal ocular movements are epileptic in origin or not.

PEM are non-epileptic in origin and are characterized by eyelid closure, upturning of the eyes, and rapid eyelid flutter. Awareness is fully retained with children able to continue to speak during PEM without any hesitation and comprehend with complex questions. PEM increase in frequency during periods of stress, excitement and tend to decrease with periods of relaxation. These cannot be induced by hyperventilation test clinically which is helpful to differentiate them from epileptic absences and eyelid myoclonia. Reports in the literature suggest a link between PEM and abnormal EEG response to visual stimuli, known as the photoparoxysmal response [1,2]. A photoparoxysmal response is defined as an abnormal electrographic cortical response to intermittent photic stimulation. It may be restricted to posterior visual areas on EEG with occipital spikes or maybe more generalized. Absences with eyelid myoclonia or Jeavons syndrome is an idiopathic generalized epileptic syndrome characterized by eyelid myoclonia with or without absences with generalized epileptiform activity triggered by eye closure with an onset in childhood.

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We present a child from a tertiary pediatric epilepsy center in the United Kingdom with paroxysmal eyelid movements (PEM) associated with photoparoxysmal responses, which was a source of diagnostic confusion.

Case presentation

A previously well 10-year-old girl presented with episodes of upward rolling of eyes several times a day lasting for a few seconds, associated with unresponsiveness.

Interictal EEG showed frequent generalized spikes and polyspikes and slow waves more dominant in the frontal regions sometimes seen with a right sided emphasis. A photoparoxysmal response was elicited and the EEG was interpreted as supporting the diagnosis of primary generalized epilepsy. These discharges were not related to eye closure (Figure 1).

Lamotrigine was ineffective but Levetiracetam reduced the frequency of these episodes.

At the age of 12 years, the girl reported several episodes of eye blinking in the day different from the original episodes, with fully preserved awareness and responsiveness. The family reported that these were triggered with stress and excitement with increased frequency during school days and reduction in frequency over weekends. Video footage of these episodes revealed eyelid fluttering, spontaneous eye opening and eye deviation upwards and to the left on eye closure.

A clinical diagnosis of eyelid myoclonia was made. A photoparoxysmal response was present as photic stimulation continued to provoke generalized spike and polyspike and wave activity (Figure 2). However, an EEG was performed which did not show epileptiform discharges correlating to these eye blinking episodes (Figure 3).

The ocular movements were hence identified as PEM, this was discussed with the child and family and no further change was made to anti-epileptic medication regime.

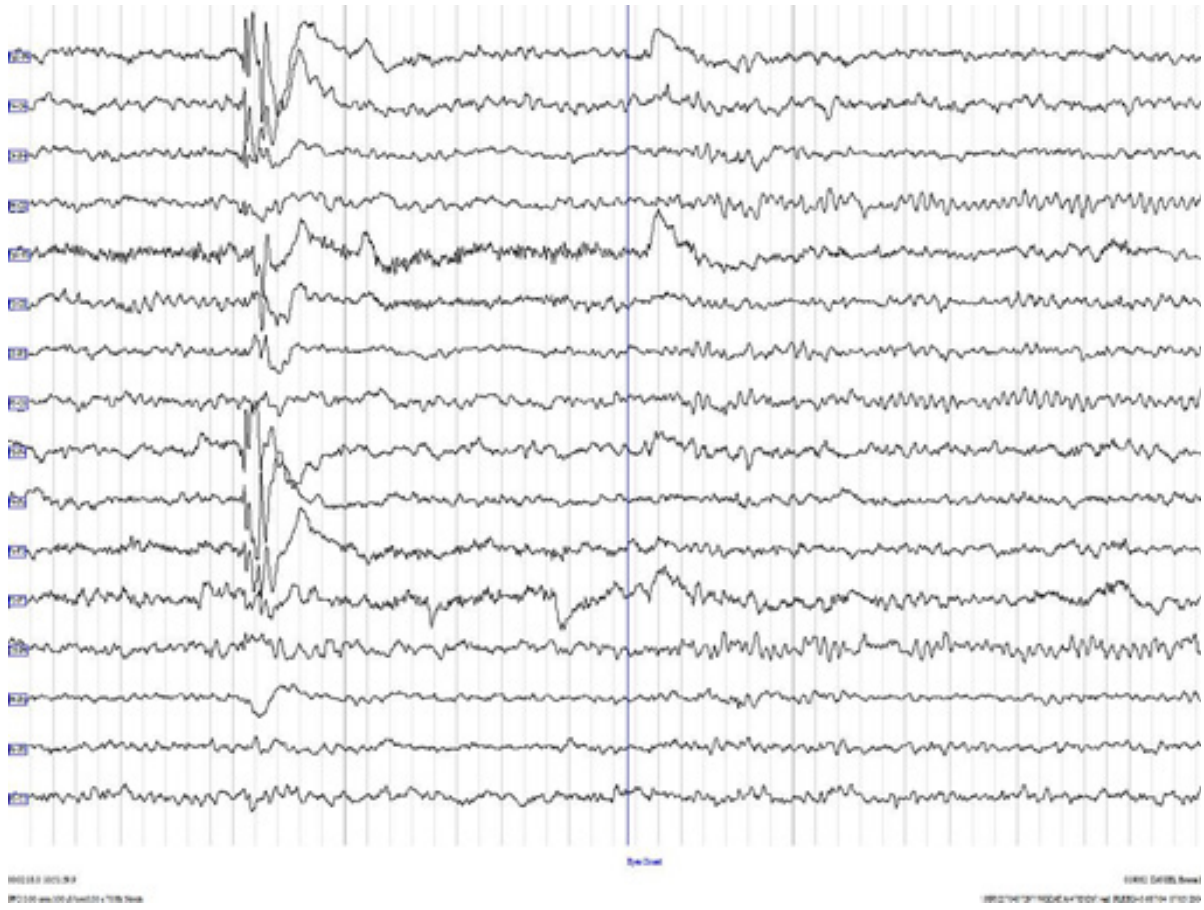

Figure 1: *ictal EEG showing frontally predominant spike/polyspike and wave activity with a right sided emphasis.*

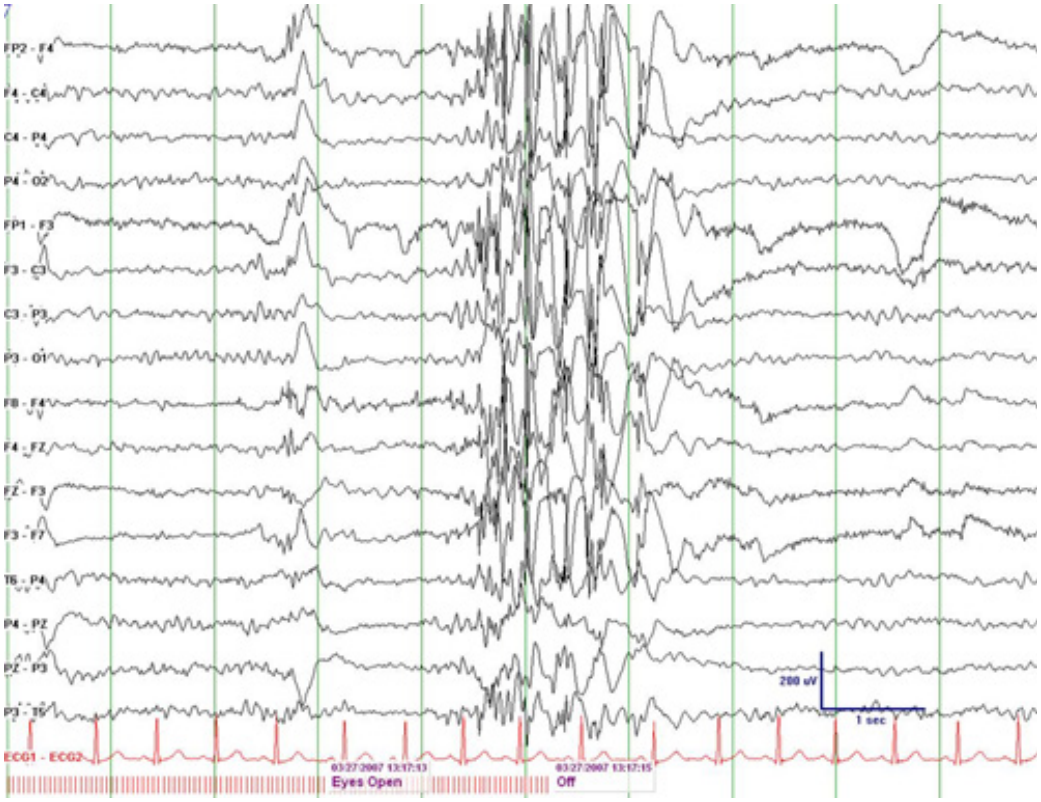

Figure 2: *Photoparoxysmal EEG response to 20Hz.*

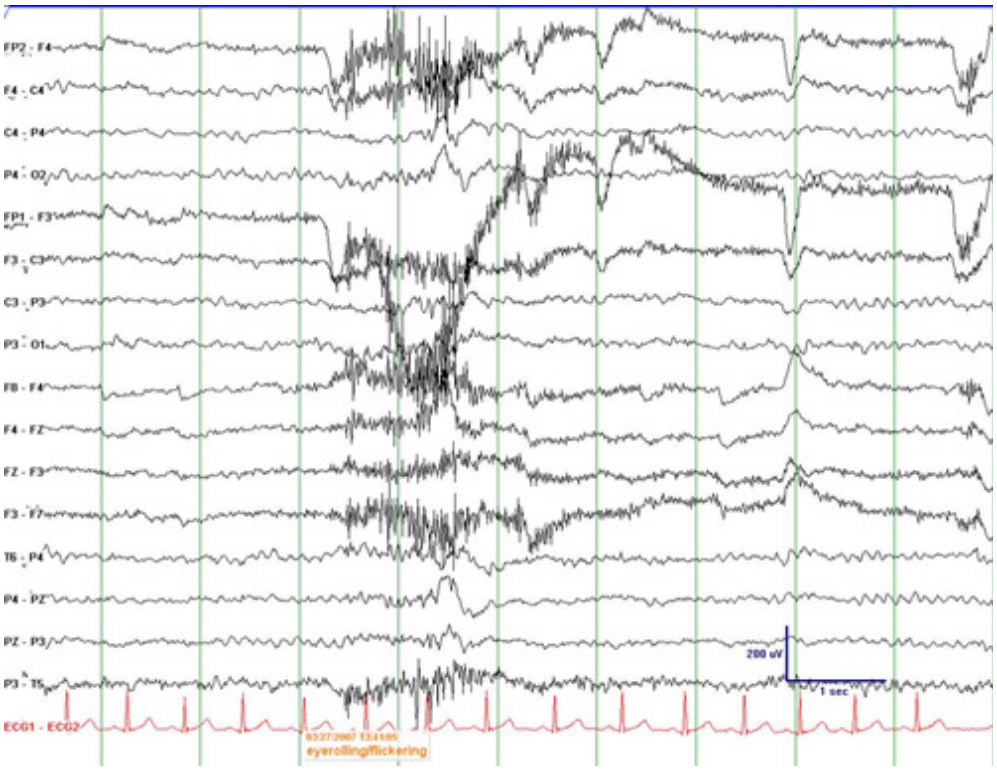

Figure 3: *Interictal EEG showing no epileptiform features associated with paroxysmal eyelid movement.*

Discussion

The episodes in our case did not have any clinical features suggestive of blepharospasm or tics. Blepharospasm is a subset of focal dystonia characterized by contraction of orbicularis oculi; clinically there was no suggestion of orbicularis oculi involvement although her case was not excluded by electromyography [3]. Motor tics are repeated but not rhythmic in nature, typically wax and wane and may change over time [4].

PEM was reported by Camfield et al, 2004 to be associated with photosensitive generalized epilepsies [1]. The case series reported the occurrence of PEM in 19 patients with generalized epilepsies: three with childhood absence epilepsy, eight with juvenile myoclonic epilepsy, and eight with absence epilepsy plus generalized tonic-clonic seizures. The episodes were stereotyped, frequent, and seemed to be triggered by emotions such as stress. No EEG abnormalities were associated with PEM, and they were not triggered by hyperventilation or photic stimulation during the EEG.

A study of 26 patients with PEM revealed a correlation with photoparoxysmal responses, pattern sensitivity, increased rate of blinking and absences with eyelid myoclonia, in a cohort of both focal and generalized epilepsies [3].

Therefore, PEM represents a non-epileptic phenomenon that is closely linked with different types of epilepsy, particularly in the presence of photoparoxysmal responses.

This can pose a difficulty in accurately assessing seizure burden in children with epilepsy who present with PEM, and can potentially result in an escalation of anti-epileptic medication regime, with an increased risk of adverse effects and no benefit in managing the PEM.

Conclusion

The presentation of children with epilepsy with ocular abnormal movements, particularly in the presence of photoparoxysmal responses, should raise suspicion of possible PEM. EEG recordings should be sought to capture the episodes to be able to confirm an electroclinical diagnosis of these episodes.

Acknowledgements

The authors thank Neurophysiology Department at Sheffield Children's Hospital for providing EEG recordings.

List of Abbreviations

ILAE	The International League Against Epilepsy
EEG	Electroencephalogram
PEM	Paroxysmal eyelid movements

Conflict of Interests

The authors declare no potential conflict of interest with respect to the research, authorship and/or publication of this case report.

Funding

None

Consent for publication

Informed parental consent was obtained for publication of this case report in a medical journal.

Ethical approval

N/A

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Authors' contributions

SRM was responsible for recognizing the importance of publicizing the lessons from this case to the wider medical community. AF was the primary writer, and SRM provided advice for revision and important intellectual content.

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Summary of the case

Patient (gender, age)	1	12 year old, female
Final Diagnosis	2	Non-epileptic Paroxysmal Eyelid movements in a child with epilepsy
Symptoms	3	New onset eyelid fluttering with eye rolling with no loss of consciousness in child with generalized epilepsy
Medications (Generic)	4	Lamotrigine and Levetiracetam
Clinical Procedure	5	Electroencephalogram (EEG)
Specialty	6	Pediatric epilepsy, Neurology
Objective	7	To report a case of non-epileptic paroxysmal eyelid movements in a child with generalized epilepsy
Background	8	12 year old girl with generalized epilepsy diagnosed at 10 years of age presented with new onset paroxysmal eyelid movements
Case Report	9	A case of paroxysmal eyelid movements in a child with generalized epilepsy which was a source of diagnostic confusion. EEG recordings captured the PEM episodes to confirm the diagnosis.
Conclusions	10	The presentation of children with epilepsy with ocular abnormal movements, particularly in the presence of photoparoxysmal responses, should raise suspicion of possible PEM. Confirmation is essential by capturing these events on EEG.
MeSH Keywords	11	Epilepsy, myoclonia, non-epileptic, EEG, case report