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Identification of Interictal Paroxysmal Diffuse Sharp Activity with Eye Closure in Patients with Generalized Epilepsy

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ABSTRACT

Interictal EEG recordings of patients with generalized epilepsy have known interictal abnormalities such as generalized spike and wave activity during photic stimulation and hyperventilation, interictal spike and wave or diffuse sharp activity [1]. We report three patients with confirmed generalized epilepsy who's interictal recordings showed paroxysmal diffuse sharp 10 Hz activity in all leads with eye closure following eye blinking. This pattern was not associated with interictal generalized spike and wave activity, clinical change in the patient or did not follow seizure activity. Abnormal eye movement with generalized spike and wave activity has been described in Jeavon's syndrome, eyelid myotonia and Sunflower syndrome. However, our patients did not meet criteria for any of these diagnoses. Therefore, we feel that our finding of paroxysmal diffuse sharp alpha activity is a novel finding in these patients with primary generalized epilepsy and may be a newly reported marker for patients with primary generalized epilepsy. Recognition of PDSA activity and further study of this pattern is encouraged.

Keywords: Interictal Paroxysmal Diffuse Sharp Activity; Interictal EEG

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Introduction

Interictal EEG recordings of patients with generalized epilepsy have known interictal abnormalities such as generalized spike and wave activity during photic stimulation and hyperventilation, interictal spike and wave or diffuse sharp activity [1]. We report three patients with confirmed generalized epilepsy whose recordings showed generalized sharp 10 Hz activity in all leads with eye closure following eye blinking.

Case Report

Patient 1 was diagnosed with epilepsy at age 3. She initially presented with daily staring spells and convulsions triggered by photic stimulation. EEG showed 3-4 Hz generalized spike and wave activity during photic stimulation, hyperventilation, wakefulness and sleep. MRI brain was normal. She was treated with anti-epileptic medications. However, as a young child she had a tendency to self-induce seizures by opening and closing blinds which induced absence seizures with eye fluttering. Convulsive seizures occurred rarely with anti-seizure medication treatment. Breakthrough seizures usually occurred when she was febrile or sleep-deprived. Over the years, she was treated with valproic acid, levetiracetam, zonisamide, clobazam, and topiramate. Medications at time of EEG (see figure 1A) were valproic acid and levetiracetam.

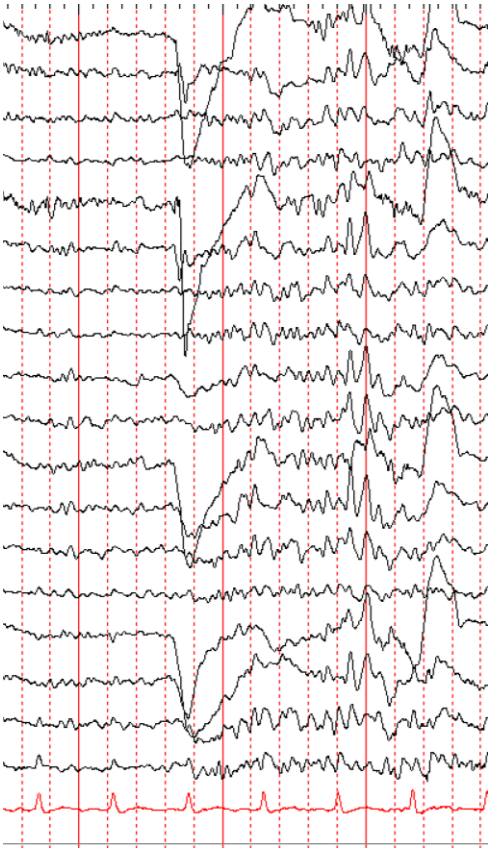

Figure 1 A
Legend: Bipolar montage. HF 30 Hz. Sens 7 mV.

Her seizure frequency improved as she grew older, however, interictal epileptiform discharges remained frequent on EEG testing. Currently at the age of 21, she is treated with a homeopathic type of CBD oil and levetiracetam. She has not had a convulsive seizure in 5 years. Absence seizures are rare and only witnessed by the family on a monthly basis, usually catamenial.

Patient 2 had her first convulsive seizure at age 12 when she was sleep-deprived. Routine EEG the following week showed 3 Hz generalized spike and wave discharges in sleep. MRI brain was normal. She was begun on anti-epileptic

medication several months later when staring spells that the family had witnessed in the evenings were confirmed to be seizure activity based on EEG findings. She was treated with levetiracetam, then topiramate. At the time of the EEG (see Figure 1B) she was only taking topiramate. Currently at the age of 16, her convulsive seizures are well controlled on lamotrigine and only occur about once a year in the setting of sleep-deprivation. Evening absence seizures are seen only on days when she has been very active.

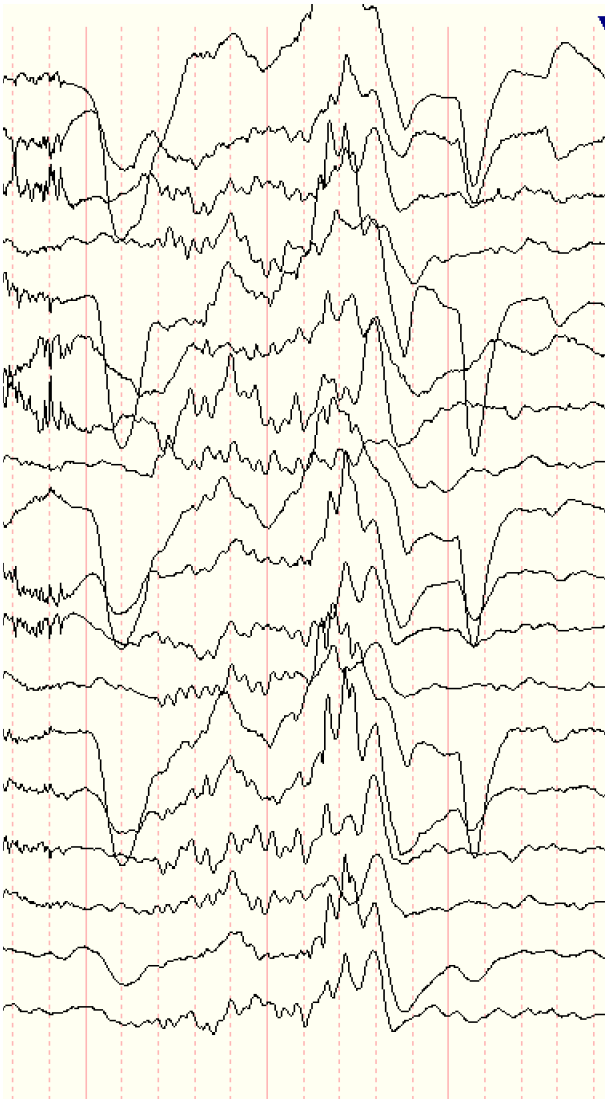

Figure 1 B

Patient 3 was diagnosed at age 8 with generalized epilepsy following a convulsive seizure and electroencephalogram showing 3 Hz generalized spike and wave discharges. A Vagus Nerve Stimulator has controlled his convulsive seizures and he only has rare absence-type seizures.

These patients had an identified unusual paroxysm of diffuse, sharp alpha activity with eye closure that followed eye blinking. See figures 1A, B and C. All patients were awake at the time of the paroxysm and eye blink artifact can be seen prior. Video recording confirmed presence of eye blink movement. Then, the eyes closed and presence of this paroxysm occurred. Staring, eye fluttering or eyelid myotonia was not identified in the video recording at these times. In patient 1 (figure 1A), the frequency of the sharp activity was 10 Hz and was present in all leads. Amplitude was low and steady at 40 microvolts which actually matched her background amplitude. In patient 2 (figure 1B), eye blink artifact was seen, then she closed her eyes and presence of sharp 10 Hz waveforms

was in all leads. Amplitude varied from 30 microvolts to 100 microvolts. Patient 3's activity (figure 1C) also consisted of alpha activity with an amplitude of 60 microvolts. Muscle artifact can be seen in frontal leads; however, diffuse alpha is also noted. Review of EEG data following these paroxysms did not reveal any seizures or presence of interictal generalized spike and wave activity.

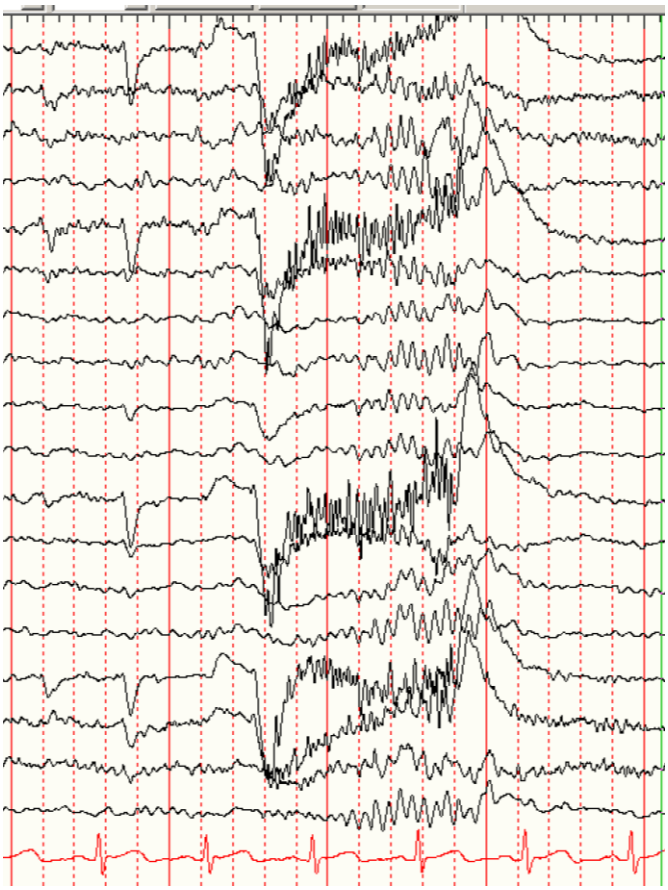

Figure 1 C

Discussion

The above mentioned paroxysmal diffuse sharp alpha activity (PDSA) seems to be a novel interictal marker for patients with primary generalized epilepsy. Although the frequency is in the alpha range, the paroxysm is not a posterior dominant rhythm as it is present in all leads and has a sharp morphology. This is also not an eye blinking induced seizure as the discharges are not spike and wave morphology, as would be seen in Jeavons syndrome. Eyelid myoclonia-type seizures, as seen in Jeavons syndrome, is associated with generalized spike and wave activity [2]. In the sunflower syndrome, it is felt that waving of the hand in front of the eyes can trigger a seizure and then the hand waving movement becomes part of the clinical seizure semiology [3]. Generalized spike and wave activity is the cardinal feature of these epileptic seizures. Waving hand movement did not occur in our patients during the paroxysmal diffuse sharp alpha activity.

Not all EEG recordings capture a seizure so interictal recordings provide useful information when interictal epileptiform abnormalities can be identified. Furthermore, certain abnormalities can suggest a diagnosis of epilepsy and/or a certain syndrome or type of seizure. In patients with primary generalized epilepsy, abnormal interictal findings are generalized spike and wave activity or generalized sharp activity. [1] Sometimes activation procedures can induce seizures, especially photic stimulation in photosensitive epilepsy and hyperventilation with absence epilepsy. However, without seizure induction, interictal abnormalities must often

suffice to make the best diagnosis and treatment plan for the patient.

Conclusion

We have identified a novel paroxysm of diffuse sharp alpha (PDSA) activity with eye closure, following eye blinking. This activity is not itself epileptic and eye blinking in these cases is not inducing a seizure or the result of a seizure. The paroxysm we describe resolves with eye opening and did not progress to an epileptic event. This activity

was also not related to certain medications. All three of our patients have had a favorable outcome with management of seizure treatment. However, all patients had a combination of generalized seizures and absence type seizures.

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