Case Report and Series

A Case of Frequent Blinking and Abnormal Eyeball Movement Associated with Generalized Epilepsy

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Abstract

Eyelid myoclonus is an idiopathic generalized epileptic syndrome that can occur with or without absence seizures. The features include frequent blinking, an upward roll of the eyeballs, and slight backward movement of the head. It can be spontaneous or stimulated by light. Light and eyelid closure are triggers to the seizures. A 13-year-old young male student presented with a four months history of frequent blinking and abnormal eye movements. There was a positive history of leg tapping while asleep which started a year prior to presentation, but there was no loss of consciousness.

On presentation, the patient was a young healthy looking myope who frequently blinks with sudden upward and left jerky movements of the eyeballs. Visual acuity was CF at 2 meters in both eyes improving with -4.50DS to 6/6. Intraocular pressures were 12 mmHg bilaterally. Anterior and posterior segment findings were normal. Brain MRI was normal, but EEG was abnormal with features suggestive of generalized epilepsy. He was co-managed with the neurologist and placed initially on Tabs Sodium valproate 250mg for 3 months. This was modified to Tabs Sodium Valproate (controlled release) 500mg at night since there was little change. This resulted in an appreciable reduction in blinking and abnormal eye movement. The leg tapping stopped on this dose. Eyelid myoclonus (EM) is a rare form of epilepsy. It is of utmost importance to create awareness of the disease among physicians. Early diagnosis and treatment are important prognostic factors of the disease.

Keywords: Frequent Blinking; Abnormal Eye Movement; Jerky Leg Movement; Generalised Epilepsy; Sodium Valproate; Eyelid Myoclonus.

Key Messages: There is a need to create awareness of Eyelid Myoclonus among physicians as this can easily be missed or misdiagnosed.

Introduction

Eyelid myoclonus is an idiopathic generalized epileptic syndrome that can occur with or without absence seizures. Eyelid Myoclonus with Absence (EMA) is also known as Jeavon's syndrome [1]. The features include frequent blinking, an upward roll of the eyeballs, and slight backward movement of the head. It can be spontaneous or stimulated by light. Light and eyelid closure are triggers to the seizures [1].

The case is being reported because of its very rare occurrence as we did not come across any case reported in Nigeria or Africa. Also, to describe the clinical features seen, to create awareness and share our experience with our management of this case. Consent to report this case was verbally gotten from the parents of the child and the child and

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How to cite this article: How to Cite: Okudo AC, BabalolaOE, Ani-OshekuI.A Case of Frequent Blinking and Abnormal Eyeball Movement Associated with Generalized Epilepsy. Niger Med J 2022;63(6):480 – 484

documented in the case note and Ethical Approval was gotten from the Institution's review board.

Case History

A 13-year-old male student presented on 24/03/2021 with a four-month history of frequent blinking and abnormal eye movements. Initially, the issue of concern was the frequent blinking as then the abnormal eye movement was sparse. Four months later the abnormal eye movement became very frequent and that caused the mother to present in the clinic. There was no history of loss of awareness of self or his environment, nil reported aura, sphincteric incontinence, or postictal phenomenon. There was a positive history of abnormal jerky movements of the legs when sleeping which started a year prior and continued till presentation. The mother noticed the leg movements occurs at about 4 am and occurs about 6 times while he is sleeping, each episode lasting for about 1 minute.

He had no family history of seizure disorder or a previous history of febrile seizures, jaundice, accident, or trauma to his head. His antenatal, birth, delivery, and postnatal history were uneventful. His development was normal and he performs brilliantly in school. He is also a known spectacle wearer for the past 3 years. He had presented in two eye hospitals before ours and his mother was told he was malingering.

On examination, we saw, a young boy, apparently healthy-looking. On ocular examination, he was blinking frequently and had abnormal eye movements. Both eyes suddenly turn left and upwards and return to normal position (video 1). His visual acuity was CF at 2 meters corrected to 6/6 with -4.50DS. The anterior and posterior examination was normal.

A brain Magnetic Resonance Imaging (MRI) was normal. Awake Electroencephalogram (EEG) showed an admix of bilateral and synchronous high amplitude mixed frequency sharp and polymorphic spike and wave complexes with bi-frontotemporal dominance. (Figure 1) Hyperventilation and photic protocols revealed paroxysms of generalized spike-wave epileptic discharges with a conclusion of generalized epilepsy. (Figure 2).

He was co-managed with the Neurologist and initially placed on controlled release Sodium Valproate 250mg with a sub-optimal response. This was later increased to 500mg nocte which resulted in an appreciable reduction in blinking and abnormal eye movement. The abnormal jerky movement of the legs stopped while on this dose.

Repeat EEG done on 10/05/2022 while he was on Sodium Valproate 500mg showed background 15 – 18 Hz beta activity with some 11- 14 Hz alpha waves; these are bilaterally synchronous but response to eye opening was not ascertained. Left sided sharp and slow wave discharges, lasting about 1.5 seconds at P3-O1 and C3-P3 channels. (Figure 3).

Based on EEG result, his medications were reviewed and increased to 750mg Sodium Valporate at this dose the clinical features sparingly occur.

There has been a stepwise increase in his medications and he is presently on 1000mg.

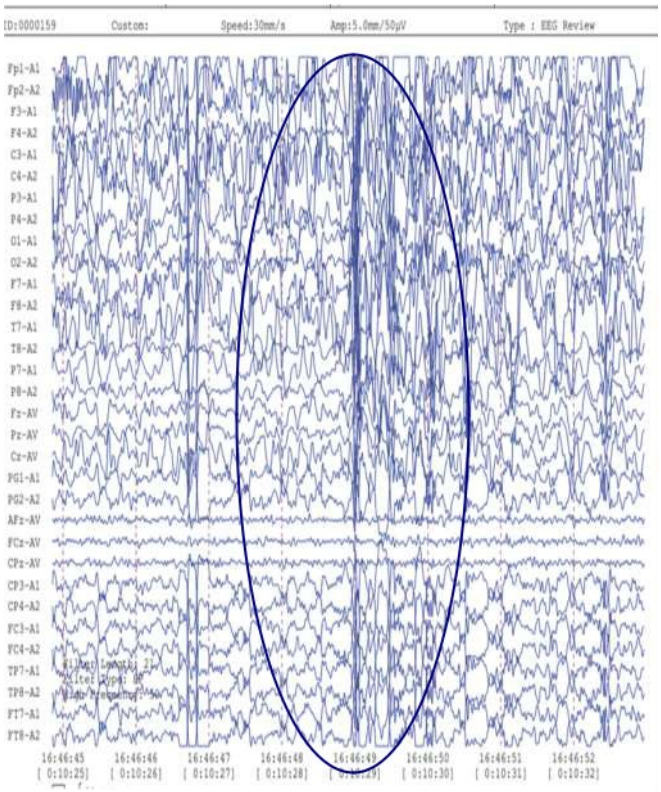

Figure 1: Awake Electroencephalogram (EEG) showed an admix of bilateral and synchronous high voltage brief (< 1 second) poly spike and sharp wave complex with bi-frontotemporal dominance.

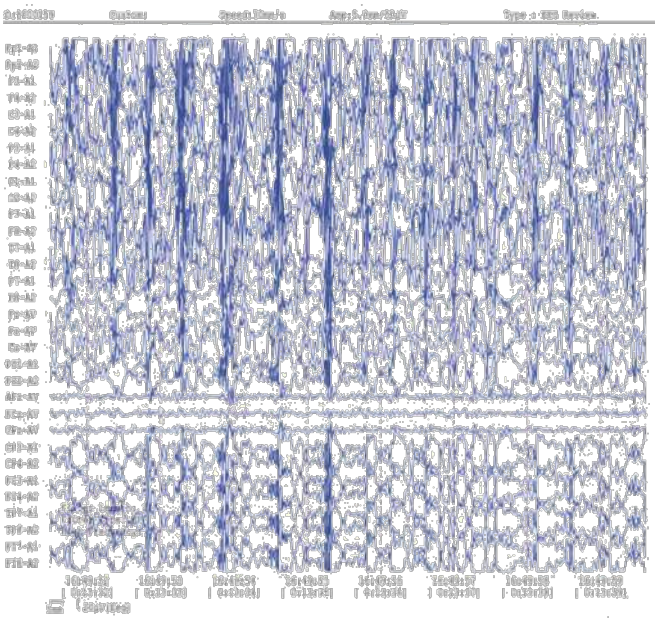

Figure 2: Hyperventilation and photic protocols revealed paroxysms of generalized spike-wave epileptic discharges with a conclusion of generalized epilepsy.

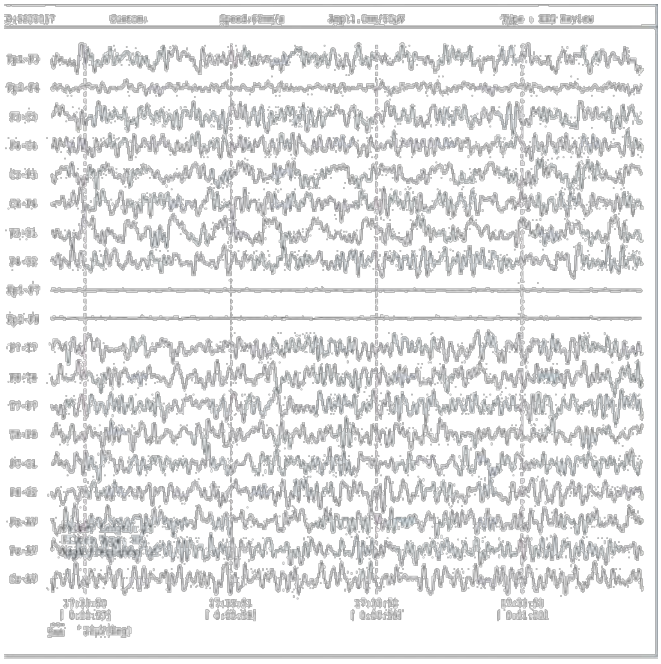

Figure 3: Repeat EEG done on 10/05/2022 while he was on Sodium Valproate 500mg showed background 15 - 18 Hz beta activity with some 11-14 Hz alpha waves; these are bilaterally synchronous but the response to eye-opening is not ascertained. Left-sided sharp and slow wave discharges, lasting about 1.5 seconds at P3-O1 and C3-P3 channels.

Discussion

Eyelid myoclonus was recently included as a separate entity in the International league classification of Epilepsy in 2017 as a form of a non-motor generalized seizures. This can either be with or without absence seizures [2]. When it occurs with absence seizures, it is referred to as Jeavon's Syndrome. Our patient has the Eyelid Myoclonus EM without absence variant because there was no loss of consciousness and he did not have EEG features suggestive of absence seizures.

EM is an idiopathic generalized epileptic syndrome that occurs in childhood [1]. It is a genetic disorder whereby there is mutation or deletion of genes such as RORB gene, KIA 2022 gene, GLUT 1 gene or NAA1O gene [1].

The seizure complexes originate from the occipital lobe and pass through connections to the thalamus and frontal lobe. The occipital lobe origin is what causes the frequent blinking and abnormal eye movement in response to a photic stimulus [3,4]. The impulses spread to the frontal lobe and that causes the jerky movement of the leg. The generalized seizures are more pronounced in the nocturnal and early hours in the morning, this could explain why the leg jerky movements occur at about 4 am which has similarly been reported to occur mainly between 5am and 6 am [5].

EM is very rare and we did not come across any reported case in Nigeria and Africa at large. This could also be the reason why his mother was told he was malingering in the two eye hospitals she initially presented in. The age of onset is said to be 6 – 8 years [1], although our patient's symptoms were first noticed when he was 11 years, there is a possibility he might have had features that were not picked up early. EM has also been described as an electrical clinical manifestation that ceases between 15 and 18 years [6]. We would have to monitor the patient closely and observe if these features would cease in the near future. The prognostic indicators of EM include photosensitivity, Eye closure sensitivity (ECS) and family history [1].

There have been documented clinical presentation variations by sex as males have predominant epileptic discharges in the frontal lobe and present

with frequent blinking and eyes rolling up movement which was similar to our case while females have predominant epileptic discharges in the occipital lobe with clinical features of frequent blinking alone but when their elliptical discharges spread to involve the frontal lobe the frequent blinking is then associated with eyes rolling up movement or absence[7].

He also had the classical presentation of rapid abnormal eye blinking, upward rolling of the eyes in which case he looks up and left and a slight movement of head backwards. Blinking and light triggers the seizures, so the frequent blinking is both a manifestation and triggering factor in EM [1].

The drug of choice for EM is Sodium Valporate 20-30mg /kg [8]. His weight was 40kg so the required dose is 1200mg daily. This is why there was a suboptimal response at 250mg. There was a stepwise increase in his medications, at 500 mg the eyelid myoclonus reduced significantly and the leg tapping stopped, but there were still significant EEG findings. His medications were further increased to 750mg at which his clinical features sparingly occur. He is presently on a 1000mg.

EM is usually misdiagnosed or missed early in the disease and usually picked up when they develop a generalized tonic-clonic seizures which occurs usually at the age of 12 years [1]. When misdiagnosed and the individual is given a wrong choice of medications such as carbamazepine or not treated early this tends to worsen the disease and can develop generalized tonic-clonic seizures [9].

One take home message is we need to create awareness of the disease, its rare nature makes the diagnosis easily missed and this delay could have put him at risk of developing generalized tonic-clonic seizures [10].

Possible differentials include sunflower syndrome [1], juvenile myoclonic epilepsy [1], idiopathic photosensitive occipital lobe epilepsy [1] and opsoclonus myoclonus syndrome [8].

Sunflower syndrome is similar to Eyelid myoclonus with features of eyelid myoclonia with or without absence seizures that is triggered when the patient waves their hand in sunlight [1]. Juvenile myoclonic epilepsy involves myoclonic convulsions especially the upper extremities. It can be associated with tonic clonic seizures and absence seizures and begins in puberty [1]. Idiopathic photosensitive occipital lobe epilepsy is a group of idiopathic focal epilepsies characterized by vision changes, hallucinations, complex hallucinations or blindness. It differs from Eyelid Myoclonus in that, occipital paroxysms on EEG occur when the eyes are closed, and disappear or are reduced when eyes are opened which is the reverse with Eyelid myoclonus[1].While Opsoclonus Myoclonus Syndrome is a disease that presents with disturbance in gait, muscle spasms, and irregular eye movements [10].

In conclusion, Eyelid myoclonus is a rare form of epilepsy. It is of utmost importance to create awareness of the disease among physicians. Early diagnosis and treatment are important prognostic factors of the disease.

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