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CASE REPORT
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A Case of Jeavons Syndrome: Eyelid Myoclonus with Myoclonus Status

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INTRODUCTION

Jeavons initially explained Jeavons syndrome or eyelid myoclonus with or without absence in 1977.[1] It has not yet been acknowledged as a categorical epileptic syndrome.[2] The electroclinical trinity of photosensitivity, eye closure-induced seizures, and electroencephalography (EEG) paroxysms serve this disease's hallmarks.[3] The normal onset occurs in childhood, peaks between the ages of 2 and 14, and is more prevalent in women (F:M= 2-3:1).[4]

Repetitive episodes of eyelid jerking, along with eye upward deviation and head retropulsion, constitute the symptoms.[5] A limited minority of patients may have eyelid myoclonus status; seizures are frequent and brief, occurring hundreds of times each day. It is known as eyelid myoclonus with absence (EMA) when it occurs in conjunction with a loss of consciousness.

The symptoms of eyelid myoclonus include intermittent and recurring seizures, along with slight absence.[6] It might happen naturally or in response to light stimulus. Eye closure and photic stimulation induce the occipital brain to become excitable, which causes eyelid myoclonus. As the photic stimulation intensity rises, generalized seizures may also happen.[7]

Long-term generalized tonic-clonic seizures (GTCS), whether they are spontaneous or light-induced, are possible. When searching in PubMed, there were only 3 reported cases of eyelid myoclonus. We are now going to discuss an unusual instance of tonic-clonic seizures accompanied by eyelid myoclonus, clinical symptoms, and EEG data.

CASE PRESENTATION

A 15 years old male visited at OPD of a tertiary care centre with chief complaints of episodes of frequent eye blinking, upward rolling of eyeballs, backward jerking of head and accompanied by decreased consciousness. These episodes occurred from 10 years of age and were specially triggered by looking at sun. Initially patient had 2–3 episodes per week, subsequently, intensity and frequency of these episodes increased with time and from last 1 year patient started having 10–20 episodes per day and each episode for a prolonged duration. Patient also develops episodes of Generalized tonic and clonic seizure

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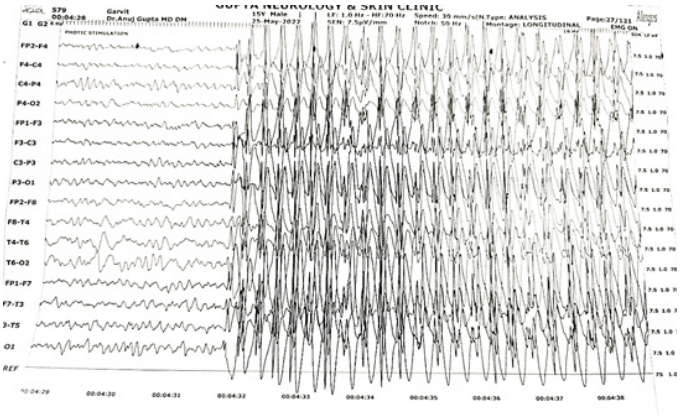

Figure 1: Background rhythm obtained in awake state revealed 9-10 Hz, 50-60 microvolt amplitude, in background of alpha activity bilaterally. During photic stimulation sudden 3-Hz spike wave discharges in generalized pattern was seen.

from last year. There was no significant past family and any medical history of epilepsy. Neurological and biochemical examination were within normal limits and MRI brain was normal. Following eye closure, the EEG displays typical background activity and magnitude 3-Hz discharges of polyspikes and waves amplified during photic stimulation (Figure 1 presenting EEG findings). The diagnosis of eyelid myoclonus with myoclonus status was made on the basis of history and video documentation recorded by family members.

DISCUSSION

The characteristic of jeavons syndrome is eyelid myoclonus, seizure induced by eye closure and photosensitivity. It is idiopathic generalized epilepsy. The eye movements are rhythmic, mostly multiple, like rapid blinking.[8] Eyelid myoclonus may or may not be followed by absences. Seizures are precipitated by eye closure, photosensitivity. There could be multiple episodes per day which may progress into eyelid myoclonus status. When there are multiple repetitive episodes of eyelid myoclonic in alliance with brief absences, then is called as eyelid myoclonus status.[6]

The prevalence of jeavons syndrome is around 3% among people having epilepsy and 13% amongst patients of idiopathic generalized epilepsy.[9] The precipitating factors for this syndrome are sensitivity to bright light, increased screen exposure, sleep deprivation, alcohol abuse, and drug sensitivity and inappropriate antiepileptic use, mainly carbamazepine and phenytoin. The etiology of Jeavons is unknown, but several hypotheses supported by familial and twin studies show genetic contribution (Park *et al* 1996; Striano *et al* 2002; Adachi *et al* 2005).

The occipital visual cortex plays an important role in the pathophysiology jeavons syndrome. The intensity of light alters the volume of occipital cortex, thereby activating the epileptic cortex. Eye closure and intermittent photic stimulation activate the epileptic occipital cortex and excitabilities spread to brainstem to produce eye myoclonus. Electric discharges spread to frontocentral cortex via either thalamocortical pathway to project generalized spike and waves associated eyelid myoclonus with absences.[10]

Physical examination and neurological examinations of these patients are usually normal. MRI brain not of much clinical significance. Video EEG is important for the diagnosis of such conditions. EEG usually shows burst of 3Hz generalized polyspike-wave complexes triggered by photic stimulation

The epileptic condition sharing similar features with jeavons syndrome includes juvenile myoclonic epilepsy, juvenile absence epilepsy, sunflower syndrome, Dravets syndrome etc. Eyelid myoclonia is often misdiagnosed as facial tics and remains undiagnosed for many years.[11]

Antiepileptic medication is usually given, including sodium valproate, benzodiazepines, levetiracetam and ethosuximide. Non-pharmacological treatment can be used for these patients like wearing special glasses or blue Z1 lenses. Sodium valproate showed good response in our patient.

Jeavons syndrome is a lifelong disorder. Absence seizure and photosensitivity disappear in middle age but eyelid myoclonia remains lifelong. Men have a better prognosis than women.

CONCLUSION

In clinical practice, Jeavons syndrome is a very uncommon kind of epilepsy. It is distinguished by myoclonia of the eyelids that develops right after

the eyes close and is linked to generalized EEG paroxysms. It is essential to raise clinicians' knowledge of this condition and increase the rate of discovery. Eyelid myoclonus status occurs less frequently and is rarely reported.

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