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Case Report

Beyond the blink: a case report on Jeavons syndrome

Ali Kumble, Abhishek K. Phadke, Anciline Siriac*, Manju Jacob

Department of Pediatrics, Indiana Hospital and Heart Institute Mangalore, India

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***Correspondence:**
Dr. Anciline Siriac,
E-mail: ancilinesiriac@gmail.com

ABSTRACT

Jeavons syndrome is a distinct syndrome characterized by the triad of eyelid myoclonia with or without absence seizures, eye closure induced generalized paroxysms, and EEG photosensitivity. We present a 5-year-old female child who was admitted for workup of high blood sugars. On evaluation child was diagnosed with type 1 diabetes mellitus (T1DM). At admission child had features of moderate diabetic ketoacidosis, requiring PICU admission for IV fluid correction and insulin infusion. Low C-peptide levels and elevated GAD antibodies confirmed the diagnosis of T1DM. Notably, the child had a 3-year history of eyelid myoclonus, prompting further evaluation. EEG revealed generalized 4-5 Hz spike-wave discharges with fixation-off phenomenon, leading to a diagnosis of Jeavons syndrome, a rare epileptic disorder. Child was started on antiepileptics. Now child is on regular follow up.

Keywords: Epilepsy, Eyelid Myoclonus, EEG photosensitivity

INTRODUCTION

Jeavons syndrome, also known as epilepsy with eyelid myoclonia, is a distinct form of generalized epilepsy characterized by a unique triad of symptoms: eyelid myoclonia (with or without absence seizures), EEG paroxysms triggered by eyelid closure, and photosensitivity. This condition is responsible for up to 13% of all generalized epilepsy cases.[1] It is classified as a generalized epilepsy but may represent an occipital epilepsy with rapid spreading. Children usually present between age 2 years and 14 years. The seizures are brief but occur multiple times per day. In addition to eye closure, bright light, not just flickering light, may precipitate seizures.[2] Between seizures, brief bursts of fast (3-6 Hz) generalized polyspike and wave are typical on the EEG. The cause of this epilepsy syndrome is unknown. A family history of seizures or epilepsy, usually generalized epilepsy, has been reported in up to 80% of children with this seizure type.[3] Seizures associated with Jeavons syndrome often exhibit poor response to antiepileptic drugs, necessitating combination therapy. Commonly used medications include levetiracetam, valproic acid, ethosuximide, lamotrigine, topiramate, and clobazam, among others, which are frequently employed in various combinations to achieve optimal seizure control.[3]

CASE REPORT

A 5-year-old female child was admitted for work up of high blood sugars. On evaluation child was diagnosed with T1DM. At admission child had features of moderate diabetic ketoacidosis, requiring PICU admission for IV fluid correction and insulin infusion. Low C-peptide levels and elevated GAD antibodies confirmed the diagnosis of T1DM. Notably, the child had a 3-year history of eyelid myoclonus, prompting further evaluation. EEG revealed generalized 4-5 Hz spike-wave discharges with fixation-off phenomenon, leading to a diagnosis of Jeavons syndrome, a rare epileptic disorder.

Child was started on levetiracetam 20 mg/kg/day in 2 divided doses. Child was on regular follow up. Last hospital visit was 1 month back, improvement of symptoms was noted. We plan to follow up the child

regularly. Child will be continued on levetiracetam till complete resolution of symptoms.

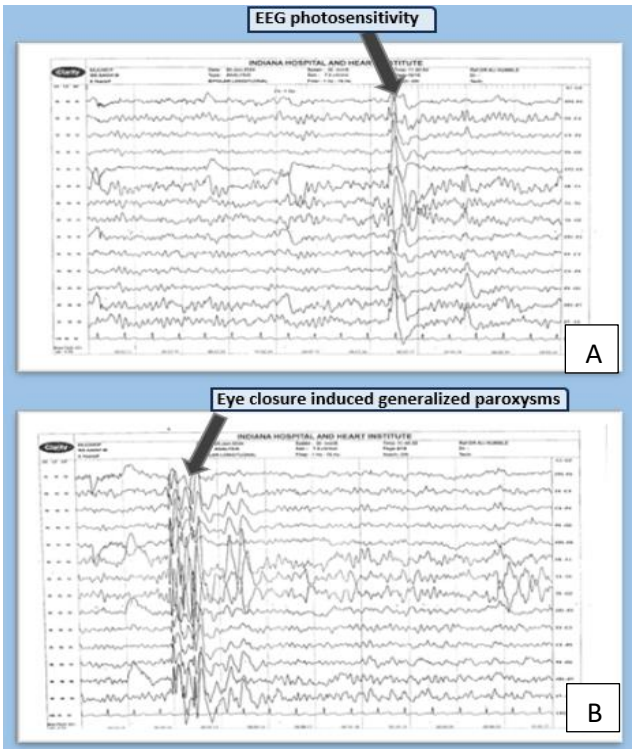

Figure 1 (A and B): EEG

DISCUSSION:

We describe a case of Jeavons syndrome in a female child.

Smith et al through his study revealed that epilepsy with eyelid myoclonus (EEM) is a childhood-onset epilepsy syndrome, more common in females, marked by eyelid myoclonia and often accompanied by other seizure types. Genetic factors are thought to play a role, with some patients having a family history of epilepsy and specific gene mutations identified in a minority. The syndrome typically emerges between 6-8 years, but onset can range from 1-16 years.[4] Similarly, our case was a female child diagnosed at 5 years of age, with family history of eyelid myoclonus.

Research by Zawar et al revealed that most patients with EEM experience a diverse range of seizure types.[5] In their study of 70 patients, the breakdown was: 80% had absence seizures, 61.4% had GTCS, 48.6% had myoclonic seizures and 5.7% had other generalized seizures (tonic and atonic) whereas in our case study, child had no clinical seizures other than eyelid myoclonia. The characteristic eyelid myoclonus can be frequently mistaken for a behavioural issue, resulting in underdiagnosis and inadequate care.

According to a study by Smith et al the diagnosis of Jeavons syndrome is frequently delayed, and certain seizure types may indicate a higher risk of drug-resistant epilepsy. Specifically, the presence of generalized tonic-clonic seizures and seizure types other than absence seizures may predict a lower response to medication, highlighting the need for early identification and tailored treatment strategies.[6] We were able to identify the case early and child was started on effective treatment immediately, and the child responded to treatment well. Patient is on regular follow up for complications.

Yang et al through his study suggested that diagnosis of Jeavons syndrome can be achieved through a combination of clinical evaluation, video-electroencephalography (VEEG) monitoring, and specific tests such as eye closure and intermittent photic stimulation. Treatment options, including valproate and newer antiepileptic medications, have shown effectiveness in managing the condition. However, Jeavons syndrome is a lifelong disorder, and while seizures can be controlled in some cases, treatment-resistant seizures may lead to cognitive and intellectual impairment.[7] We started on levetiracetam to which the child responded well, and the child shows good scholastic performance.

According to Striano et al levetiracetam has been shown to be an effective and well-tolerated treatment option for EEM.[8]

Paibool et al through his research revealed that the modified Atkins diet is a valuable treatment for pediatric EEM patients, yielding both seizure control and notable cognitive enhancements.[9] The diet was well-tolerated, with any adverse effects being mild and easily addressed, highlighting its potential as a safe and effective therapeutic strategy for EEM. In view of associated diabetes mellitus condition in our case, diet was adjusted accordingly.

A study by Cerulli et al suggests that the age of epilepsy onset may be a significant factor in determining the outcome for individuals with Epilepsy with EEM. The study found that those who developed epilepsy at a younger age were more likely to experience intellectual disability, psychiatric comorbidities, and drug-resistant epilepsy, highlighting the importance of early diagnosis and intervention.[10]

CONCLUSION

Jeavons syndrome's characteristic eyelid myoclonus is frequently mistaken for a behavioural issue, resulting in underdiagnosis and inadequate care, highlighting the importance of precise identification and education among the medical practitioners. This case highlights the need to improve diagnostic accuracy and to reduce underdiagnosis. It also enhances importance of timely and appropriate management of Jeavons syndrome and related conditions.

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