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Review Article

Sunflower Syndrome: A Summary and an Approach to Evaluation and Management

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Abstract

Sunflower syndrome is a rare form of reflex and self-induced epilepsy. Symptoms include a characteristic affinity for the sun known as heliotropism and hand waving followed by absence seizures. Patients may also have eyelid myoclonia, generalized tonic-clonic, or even myoclonic seizures [2,3]. In this article, we discuss the characteristic features of sunflower syndrome to understand better the etiology, background, and management of this condition.

Keywords: *Sunflower Syndrome; Heliotropism; Hand Waving*

Introduction

A predisposition to recurrent unprovoked seizures typically characterizes epilepsy. The International League Against Epilepsy [ILAE] defines epilepsy as (1) At least two unprovoked (or reflex) seizures occurring > 24h apart; (2) one unprovoked (or reflex) seizure and the probability of further seizures similar to the general recurrence risk (at least 60%) after two unprovoked seizures, occurring over the next ten years; (3) diagnosis of an epilepsy syndrome [1]. Reflex epilepsy is a group of seizures that can be either provoked or precipitated by specific stimuli. Sunflower syndrome is considered to be a reflex epilepsy and may be self-induced. This article aims to briefly summarize this rare form of epilepsy to recognize it in clinical practice better.

Background

Sunflower syndrome is a rare form of reflex and self-induced epilepsy. Typical features of this condition include eyelid myoclonia. These seizures may or may not be accompanied by concurrent absence seizures. However, generalized tonic-clonic seizures or myoclonic may also be seen [2,3]. Hand-waving motions may self-induce seizures as the patient faces sunlight or other bright light sources. Patients may feel a desire/compulsion to seek out these light sources themselves. The hand-waving or sometimes eye blinking, rocking motion, rubbing the forehead, and rarely looking at checkered or striped patterns [2] simulate flickering light, likely similar to photic stimulation, given their rhythmic character. These behaviors may trigger seizures. There has been speculation as to the cause of this self-induction of seizures. There is literature to suggest a compulsion to induce these events. Others speculate that the seizures themselves may be pleasurable and reduce anxiety versus the possibility of a secondary gain (for example, school avoidance versus avoidance of an unpleasant situation) [2]. Livingston and Torres, however, suggested that the hand-waving was a manifestation of the seizure itself, given that, in their experi-

ence, it occurred at the same time that the epileptiform discharges appeared on the EEG. Moreover, their patient did not recollect doing the hand-waving movements, making them suspect that these movements were likely involuntary [4].

Prevalence

Sunflower syndrome is a rare form of epilepsy and has a higher prevalence in females than males. The typical age of onset is between 2 and 8 years of age [3]. However, some patients may not be as forthcoming with this information as they may be embarrassed and may have developed a more unconscious behavior; hence, the exact prevalence may be unknown or difficult to determine [2].

Etiology

Given the complexity of the nature of these seizures, the cingulate cortex has been proposed as the likely origin for seizures associated with sunflower syndrome [5].

CHD2 gene variants have also been proposed as a genetic etiology for photosensitive epilepsies [6]. However, no clear etiology may be found in some patients [3].

Clinical features

Typically seen in the pediatric age group as described above.

Patients demonstrate an unusual affinity for sunlight or bright lights, termed heliotropism.

Parents describe unusual behaviors like trying to face the sunlight of bright lights with hand waving, eye blinking, or rocking movements to induce seizures. Eye blinking can be distinguished from eyelid myoclonia as the child does this voluntarily to evoke a seizure, though distinguishing the two is difficult at times.

Seizures may be described as eyelid myoclonia, absences, myoclonic jerks, or generalized tonic-clonic seizures [2,3,10]. These events may be reproducible on request [7].

Sunflower syndrome has been described in children with cognitive impairment. However, this can also be seen in children without any delays [3].

Although behaviors associated with sunflower syndrome may mimic stereotypies or tics, tics are typically not self-induced and do not alter awareness. Moreover, patients with tics or stereotypies do not typically demonstrate heliotropism.

Evaluation

Evaluation is typically based on history and clinical findings.

Sometimes, older children can describe a compulsion to induce these events [4].

EEG is usually helpful and can show a normal background with epileptiform abnormalities (3 - 4 Hz spike and wave +/- polyspike discharges). Sometimes, a photo paroxysmal response may also be seen, and the hand-waving motions may induce seizures. Neuro-imaging is typically unremarkable.

Treatment

Sunflower syndrome is typically refractory to treatment, and patients may require more than one antiepileptic medication. Although valproic acid has been considered one of the most effective medications for this particular condition [3], other helpful medications include levetiracetam, ethosuximide, zonisamide, and lamotrigine. Fenfluramine has been tried in patients with benefit. It should be noted that

fenfluramine's mechanism of action is to reduce the compulsion to induce seizures [8]. It also modulates sigma-1 and serotonergic receptors [9]. Various forms of psychotherapy have also been explored to treat this condition.

Zeis 1 blue lenses are beneficial in reducing seizures and have been shown to diminish the photo paroxysmal response on EEG [7,11,12].

Points for Clinical Practice/Conclusion

Sunflower syndrome is a rare form of reflex epilepsy, with seizures that may be self-induced. Although it bears many similarities to stereotypies and tics, it may lead to seizures as described above. Prompt recognition and referral to neurology are required to manage and treat this condition.

Ethical Consideration

This article has not been published elsewhere and is not under consideration for publication in any other journal.

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