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Generalized Tonic-Clonic Seizure in a Pediatric Patient with Sunflower Syndrome: A Case Report

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Case Report

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Abstract

Introduction: Sunflower syndrome (SFS) is a rare epileptic disorder characterized by a stereotypical hand- waving motion in front of the eyes when exposed to a light source. SFS is still poorly understood, but there are reports of patients who have concurrent seizure disorders as well, such as generalized tonic-clonic (GTC) and absence seizures.

Case: We present a 10-year-old boy with a past medical history of sunflower syndrome, non-compliant with pharmacotherapy, who experienced his first GTC seizure. After carefully reviewing the patient's history, a decision was made to start the patient on valproic acid.

Discussion: The question remains as to whether sunflower syndrome increases the risk of progressing to generalized seizures or if they are just more likely to suffer from both seizure disorders at the same time. It is possible that continuous electrical signals seen in sunflower syndrome could reach the threshold needed to progress to a GTC seizure; however, more research will need to be conducted on this topic.

Conclusion: Although there is little research that indicates sunflower syndrome can progress to GTC seizures, our case highlights the importance of careful seizure management and the risks of untreated epileptic disorders.

Keywords: Seizure; Epilepsy; Pediatric; Neurology; Partial Seizure

Abbreviations

SFS: Sunflower Syndrome; GTC: Generalized Tonic Clonic; CHM: Children's Hospital of Michigan; SUDEP: Sudden Unexpected Death in Epilepsy.

Introduction

Sunflower syndrome (SFS) is a rare reflex epileptic seizure that is triggered by exposure to light and involves a stereotypical hand-waving motion in front of the eyes [1,2]. It is estimated there are only 1,300 diagnoses of SFS worldwide, predominately affecting females in the first decade of life [3,4]. SFS often coincides with other seizure forms, most commonly generalized tonic-clonic (GTC) seizures [1].

The stereotypical presentation of SFS includes looking toward a light source and waving a hand with abducted fingers in front of the eyes [2]. It is most commonly the same hand every episode, with around 60% using their dominant

hand [2,5]. An ongoing debate in literature is whether the stereotypical hand waving is done to self-induce the seizures or if it is an ictal response. Previously, the leading theory was the action is purposefully caused by the patient with underlying motivation including the avoidance of unpleasant situations, seeking pleasure, addiction, compulsion, or relieving tension [6]. However, recent data using video EEG to determine the relationship of the handwaving to the EEG epileptiform has shown that handwaving episodes start less than 1 second after the start of the epileptiform, indicating that the hand motion is an ictal response [7].

Unlike other photosensitive diseases, SFS is very drug-resistant, requiring the frequent use of polytherapy [1]. Studies have found valproic acid to be the most effective pharmacologic agent [2,3]. If patients cannot tolerate valproic acid, they must consider high-dose lamotrigine or polypharmacy. Current investigations also show promising results for fenfluramine for future use [1,2,3]. Non-pharmacologic therapy, including Z1 blue lenses, focusing attention (playing sports or driving a car), occupying both hands, avoiding stimuli, and staying indoors have all been shown to decrease or prevent episodes [1,2,5,6].

We report on a 10-year-old male with a previous diagnosis of SFS, currently not on any pharmacologic therapies, who experienced his first tonic-clinic seizure.

Case Description

The patient is a white male born in 2013. At age 6, he was seen at the Children's Hospital of Michigan (CHM) Stilson Center for an autism spectrum disorder assessment. Concerning characteristics included below-average academic performance, behavioral issues, and motor tics. The patient was reported to stare into space and could not retain information repeated to him multiple times. He knew only 25 words by recognition. He was reported as aggressive, at times hitting and growling at others. He was also observed to have eye-rolling and nose-rubbing episodes. In the initial workup, EEG showed 3 Hz spike-wave discharges with unusual behavior of bringing left hand to face. He was started on ethosuximide 250 mg BID at that time.

At age 8, he was seen again at CHM Stilson Center, where episodes of unresponsive with eye fluttering when staring at bright natural lights were reported. He was continued on ethosuximide. An MRI of the brain without contrast showed no masses but an incidental finding of pineal gland cystic changes and possible gliosis.

At age 9, he was again seen at CHM Stilson Center. At this visit, he was noted to have still episodes of blinking and face rubbing lasting less than 1 minute when exposed to light. Because of these breakthrough seizures, ethosuximide was increased to 375 mg BID, and it was recommended that he switch to valproic acid if symptoms persist.

At age 10, he was seen at Henry Ford Macomb Hospital following a GTC seizure. He was found on the bathroom floor with his head against the wall. All four extremities shook, eyes rolled backward, and he foamed at the mouth. The patient had no loss of bladder or bowel function. The episode lasted less than 1 minute, followed by a postictal state lasting less than 3 minutes. His parents thought the episode was triggered by light coming in from the bathroom window. The patient had been non-compliant with ethosuximide for 9 months due to nausea, vomiting, and repeat breakthrough seizures. A review of the systems was positive for a mild frontal headache and two episodes of vomiting following the episode. A routine EEG showed 3 Hz spikes and slow wave complexes over both hemispheres. As a result, the patient was started on valproic acid sprinkles 125 mg TID. After improved symptoms over 24 hours, the patient was discharged on valproic acid with midazolam as rescue medication but adjusted to rectal diazepam for cost purposes. The patient was to follow up at CHM Stilson Center.

Discussion

The incidence of patients with a multiple-year history of SFS developing a GTC is unknown. Still, it raises the question of whether there is a threshold for SFS to progress into a GTC seizure or if patients are just more likely to suffer from both.

A previous case series reported that 66% of patients with SFS have another seizure type following a prolonged hand-waving episode, such as absence, GTC, or myoclonic [5]. According to this report, the most common association is GTC, which is seen in 50% of all SFS patients [5].

However, the underlying mechanism has not been investigated. As these patients already suffer from a type of photosensitivity epilepsy, one hypothesis is that the GTC seizures are entirely independent of the SFS, as both can be related to flickering light by handwaving. Another hypothesis is that prolonged light or handwaving exposure precipitates a GTC seizure. Previous studies have stated that >70% of patients with focal onset will experience a secondary generalization [8]. Therefore, the network of cortical and subcortical structure activity changes plays a role in motor changes and impaired consciousness in GTC seizures [8].

Using this concept, a prolonged episode of SFS could cause electrical signals to reach a threshold in this network to cause a new GTC seizure. Further research should be conducted to determine the true relationship between SFS and GTC seizure.

Despite the underlying cause of this episode, the development of a first GTC seizure is of concern for patients with Sunflower Syndrome as this is a significant risk factor for sudden unexpected death in epilepsy (SUDEP). GTC seizures, either primary or secondary, are one of the strongest risk factors for SUDEP [9]. Other risk factors for SUDEP include the frequency of GTC seizures, male sex, and young age of epilepsy onset [10]. Research has been conducted to determine which anti-seizure medication are more effective at reducing the risk of SUDEP in those with generalized seizures. The most recent meta-analysis indirectly comparing anti- seizure medications suggests there is no robust difference in effectiveness, indicating that pharmacotherapy should be tailored to fit each patient [11]. Patients with SFS, especially those who develop GTC seizures, need to be carefully monitored for pharmacological and nonpharmacological treatment efficacy with prompt adjustments as needed. Further research should be conducted on the effectiveness and compliance of pharmacologic and non-pharmacologic modalities for patients with SFS in with a focus on decreasing recurrence and the potential progression to SUDEP.

Conclusion

Due to its rarity, SFS remains poorly recognized and understood. Managing SFS involves accurate diagnosis and effective control of symptoms to facilitate daily functioning. While there is limited evidence directly linking SFS to an increased risk of GTC seizures, this case re-affirms the potential and highlights the dangers of improper management, emphasizing the importance of early recognition and appropriate management. Further research should be conducted to the possible mechanism of SFS progression to GTC and to determine the effectiveness and compliance with current SFS therapies.

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