

CASE REPORT

Self Induction of Photosensitive Epilepsy - Role of Secondary Gains and Psychological Management: Twin Case Report and Literature Review

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Abstract

Two cases of photosensitive epilepsy, the process of self-induction and relationship with likely secondary gain are detailed in the reports. The discussion highlights the role of behavioral intervention along with anti epileptic drugs (German J Psychiatry 2007; 10: 96-99).

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Introduction

Reflex epilepsy is not a syndrome but a collective of different seizure types and their specific provoking stimuli. Seizures or epileptic syndromes triggered by a stimulus are designated as "reflex" (Shashi and Lionel, 2005).

The International League Against Epilepsy (ILAE) included reflex epilepsies as 'epilepsies characterized by specific modes of seizure precipitation' among the special syndromes and divided them into simple and complex forms. In simple forms, seizures are precipitated by simple sensory stimuli (eg. light flashes). The intensity of the stimuli is decisive, the latency of the response short (seconds or less) and mental anticipation of stimulus without effect. In complex forms, the triggering mechanisms are elaborate (eg. looking at one's own hands, listening to specific music etc). The specific pattern of the stimulus not the intensity is the decisive factor. Latency of response is longer and mental anticipation of stimulus, even in dreams may be effective. The various types

of reflex epilepsies include those with visual triggers (i.e. photosensitive seizures), those induced by complex activities like thinking, reading eating, music or language, movement induced seizures, hot water epilepsy and miscellaneous other types eg. those evoked by touch, vestibular stimuli etc (Singhi and Bansal, 2004). In noogenic or thinking epilepsy seizures are precipitated by an elaborate mental activity implicating decision complexity, sequential factors, and possibly related stress (Koutroumanidi et al., 1998)

Seizures evoked by visual stimuli (photosensitive epilepsy) though a rare phenomenon still stands out as the most common form of reflex epilepsy (Senanayake, 1994; Parra et al., 2005). Photosensitive epilepsy has been reported to affect 1 in 3000 people and comprises 2% of all newly presenting cases with epilepsy (Quirk et al., 1995). In 80 % of the photosensitive epilepsy patients, attacks are precipitated by environmental visual stimuli (Parra et al., 2005).

Photosensitivity is defined as an abnormal electroencephalographic or clinical response to light. (Shashi and Lionel, 2005). The International League Against Epilepsy (ILAE)

has proposed substituting the word "Photosensitive" by the term "Visual Sensitive" (Engel, 2001).

Photosensitive epilepsy (PE) is the most frequent form of reflex epilepsy. The first form is PE with environmental factors of the process' provocation, which is characterised by genuine character of disease, pre- and pubertal period of its beginning, considerable prevalence of males among the patients, rare generalized tonic and clonic fits and favorable prognosis. The second form of PE is significantly rarer PE form -with self-induction of the fits. It is characterized by the beginning of the disease in preschool period, even sex distribution, quite frequent evaluated organic damages of brain, non-convulsive fits and non-efficiency of antiepileptic drugs (Karlov et al., 1997).

Furthermore, photosensitivity is generally believed to be genetically determined, benign type of epilepsy in childhood and adolescence but when associated with self-inducing behaviour is interpreted as a sign of mental subnormality (Kasteleijn- Nilst Tr., 1989). Whether or not these conceptions are valid and whether photosensitivity is or is not a special subtype of epilepsy remains unsolved.

Photo-convulsive response (PCR) during intermittent photic stimulation is reported to account for around 5% of the epileptic population in general (Oomman and Madhusudan, 2001; Kasteleijn., 1989). A study done in North India reported a rather low prevalence of photosensitivity (0.6%) with only 0.3% having a history of seizures provoked by visual stimuli (Saleem et al., 1994).

A rarer phenomenon reported in 0.62% of patients with photosensitive epilepsy is self-induction of seizures (Senanayake, 1994). The majority of patients who induced their seizures were photosensitive and they induced seizures by gazing at the sun and waving a hand in front of their eyes. The segment of population that is more prone to this kind of problem is children and adolescent. Often there is clear social advantage such as avoidance of school or gaining attention associated with the phenomenon of self induction (Binnie, 1988). The self induced photogenic seizures are rare and therefore often misdiagnosed as some sort of "psychogenic problem" and are poorly controlled by antiepileptic drugs (Singhi and Bansal, 2004).

Self inductive behaviour may inadvertently trigger a generalized tonic clonic seizure (GTCS). After the occurrence of GTCS frequency of self induction diminishes dramatically for several hours. (Kasteleijn- Nilst Tr., 1989)

Visual stimuli that evoke visual sensitive seizures are classified into four broad categories (a) Simple Flicker (b) Patterns (c) Self – induction (d) Television (TV) and electronic screen video games. The primary modes to self-induce seizures are (1) moving their hands in-front of their eyes. (2) Eye closure. (3) Staring at patterns (Shashi and Lionel, 2005).

With the above background we present two case reports of patients who self-induced seizures by exposing themselves to sunlight. We intend to discuss the characteristics associated with the phenomenon, and find out the reason for this behavior, role of secondary gains and its possible remediation using behavioral therapy and medical treatment.

Case Reports

Case A A 20 year old, male student from upper socio-economic status, presented with a 6 year history of academic decline, more for science and maths, feeling drowsy during classes along with inability to concentrate and recall topics covered. He had a history of three episodes of Generalized Tonic Clonic Seizures starting at 15 years of age, with last episode occurring 5 years back. All three episodes were during early morning hours, and associated with teeth grinding, clenching of jaw, facial grimacing, frothing and tongue bite along with post-ictal confusion and sedation. For the last 5 years patient was maintaining well on 600mg of carbamazepine.

The family members reported history of voluntary hand movements i.e. waving his hand in front of the eyes while gazing at the sun. This was reportedly more common during the morning hours with spontaneous termination within two to three minutes. At the start of the episodes he would be non responsive to verbal commands, stand erect, and be aroused only by tactile stimuli i.e. touching / shaking him. Patient reported amnesia for the episode and reported the entire episode "pleasurable" and beyond his control initially.

His mental status examination revealed abstraction at semi-abstract level and formal intelligence testing revealed borderline intelligence. On Strubb and Black neuro-cognitive assessment scale deficit were found in abstraction, calculation and verbal recall. His physical examination revealed abnormal hairline, low set ears, high arched palate, and micrognathia. Patient's EEG showed generalized sharp-slow wave discharges worsened by photic stimulation.

Case B An 8 year old female, from low socio economic status educated till 4th standard with no significant past or family history, presented with 2 years history of night blindness and episodes of loss of awareness. These episodes were precipitated by patient standing and gazing at the sun and waving the hands in-front of her face. This would generally continue for 20-30 minutes followed by fall and loss of consciousness and subsequently patient would have tonic contractions of all 4 limbs, lasting for 15-30 minutes, associated with frothing, up-rolling of eyeballs, clenching of teeth, followed by 4-5 hours of sleep. The frequency of episodes reported was two per week, except during rainy season and school vacations wherein there were no episodes of self induction or seizure reported. Mother reported irritability and significant interpersonal problems with the patient ever since patient's father deserted them three years back. Physical examination of the patient revealed no significant findings except for bitots spots (secondary to Vitamin A deficiency) and patient was started on supplements for the same. EEG showed generalized sharp-slow wave discharges worsened by photic stimulation. As most of the episodes were self induced a combination of pharmacological and psychological intervention was planned. She was started on T. Carbamazepine 600 mg. Intellectual assessment revealed dull normal intelligence and avoidance of school was found to be the secondary gain. Contingency management was initiated for self induction of seizures, which was met with initial

resistance. However, later patient became more compliant to treatment. Behavioural counseling was done with the mother and reinforcement principles was explained. Patient had only one episode of seizure during her stay of 17 days in the hospital and was maintaining well on follow ups.

Discussion

It has been known that reflex epilepsy generally affects children and adolescent with the female to male ratio being 3:2 (Parra et al., 2005). Both our patients belong to the commonly affected age group of 10-15 years (Rittacio, 1994; Kasteleijn-Nolst Tr., 1989). The stimulus for both patients is sunlight and self-induction by gazing at sun and moving their hand in front of their face. In Case A, a probable stimulus in the form of extra-ocular muscle movements (as a part of REM sleep) cannot be confidently ruled out. Patient's inability to concentrate in the class, feeling sleepy, partial amnesia and academic decline can be explained by sub-clinical seizures induced by thinking and performing arithmetic tasks, also called "seizure induced by thinking" and "noogenic epilepsy". Strong correlation has been shown between mental activities/ cognitive tasks and seizures; there are reports of coexisting self induced noogenic epilepsy in individuals with photosensitive epilepsy. Hence, a possibility of a combination of external eg. visual stimuli, and internal stimuli eg. thinking has to be considered in individuals presenting with photosensitive epilepsy (Koutroumanidi et al., 2006). The co-occurrence of noogenic epilepsy along with photosensitive epilepsy is a clinical possibility in Case A. Photosensitivity is found to be associated with spontaneous seizures in 30-80% of individuals (Rittacio., 1994) and is the case in case A. Based on a classification of epilepsy syndromes precipitated by light (Rittacio., 1994), case A falls in the category of photosensitive epilepsy associated with spontaneous seizures, and case B appears more like pure photosensitive epilepsy.

Self-induction of seizures as seen in both cases, is important from clinical management point as the most extreme of non-cooperation in the treatment of epilepsy is seen in those patients who deliberately induce their own seizures (Binnie., 1988; Singhi and Bansal, 2004)

An overwhelming majority of patients who induce their seizures are photosensitive and make use of visual stimuli to induce either overt seizures or 'sub-clinical' epileptiform EEG discharges (Binnie.,1988). Trigger mechanism for human photosensitivity involves binocularly innervated cells in visual cortex. Thus visual cortex is the seat for the primary epileptogenic process, and photically triggered discharges and seizures may be regarded as primary with secondary generalization (Hennessey and Binnie, 2000).

The best documented mode as seen in both the cases detailed above is self induction (Shashi and Lionel, 2005) wherein a flicker is produced by waving one hand in front of the eyes while gazing at the light source. The other mode that is much more frequently recognized is the use of a maneuver involving slow closure and forced upward deviation

of the eyes. This is exhibited by a quarter of patients with photosensitive epilepsy, many of whom report that the maneuver gives a pleasant sensation or relieves feelings of stress (Binnie, 1988).

EEG signature of pathologic photosensitivity is a photo paroxysmal response to intermittent photic stimulation; a photo-paroxysmal response may be defined as generalized spike/ polyspike and wave discharges in response to photic stimulation but not necessarily accompanied by convulsive movements (Rittacio, 1994). Interictal EEG, mental status examination and neurological examination are typically normal.

The initiation of these seizures may relate to the hyperexcitability of the focal cortical areas, which correspond to those activated by the special EEG activation (Matsuoka, 1990)

The self induction can be due to variety of motives often there is clear social advantage such as avoidance of school or gaining attention. Some patients find that the risk of spontaneous attack is lessened following self induced seizures and can therefore contrive their fits to occur only in situations which are safe and convenient (Binnie., 1988). We were able to establish pleasure in case A as a primary motive. However, in case B gains in the form of avoidance of school was evident and clearly established.

Due to the rarity of the phenomenon of self induced photo-genic seizures, it is often misdiagnosed as some sort of "psychogenic problem" and is poorly controlled by antiepileptic drugs (Singhi and Bansal, 2004). Hence careful look at psychiatric history to look at motives behind induction must be ascertained and handled through appropriate measures. Techniques like education, behavioural management, or medicinal treatment (in case of pleasure due to sense of reward mediated by dopamine, where anti-dopaminergic medications may be of help). As preliminary trials suggest, that in some patient's dopamine antagonists may reduce this behaviour, apparently by making self-induced discharges or seizures non-rewarding (Binnie, 1988)

In contrast to the report that the self-induced seizures are notoriously resistant to antiepileptic therapy even if the patient is compliant (Binnie, 1988); we found that both of our cases had adequate seizure control through with a combination of single anti-epileptic and behavioral intervention and this can be an effective mode of treatment.

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