NEW RESEARCH

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CASE REPORTS

Topiramate Responsive Exploding Head Syndrome

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Exploding head syndrome is a rare phenomenon but can be a significant disruption to quality of life. We describe a 39-year-old female with symptoms of a loud bang and buzz at sleep onset for 3 years. EEG monitoring confirmed these events occurred in transition from stage 1 sleep. This patient reported improvement in intensity of events with topiramate medication. Based on these results, topiramate may be an alternative method to reduce the intensity of events in exploding head syndrome.

Keywords: Topiramate, exploding head syndrome, auditory sleep starts

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Exploding head syndrome (EHS) is a rare phenomenon characterized by a painless loud noise at the onset of sleep. The phenomenon was reported initially in 1920 by Armstrong-Jones when he referred to it as "snapping of the brain." The term *exploding head syndrome* was coined by Pearce in 1989 in a paper in which he described 40 patients with EHS.[2] Given the rarity of the disorder, no treatment studies have been done. A thorough review of the literature revealed a few case reports and opinion papers. Events described in the literature start spontaneously and are not associated with hypnic jerks. Although treatments have been reported, no trials have been performed. In these cases and case series, there was no mention of topiramate being prescribed for these events.

REPORT OF CASE

A 39-year-old female reported symptoms of a loud bang and buzzing noise at sleep onset for 3 years. She said that, if the sound was external, her "husband should be able to hear it downstairs when she was up in her bedroom." Associated with this noise, she experienced brief jerking movement of her head, leg, or arms at sleep onset on a daily basis. She noted these symptoms for years; because of the increase in intensity and frequency, she saw a neurologist. The patient had become anxious about these events, fearing that they were a hallmark of more serious medical issues. Her neurological exam, laboratory test results, and neuroimaging were normal. Because of the stereotypic nature of the events and the level of disturbance to the patient, she was admitted for continuous video EEG monitoring for 4 days. Coincidently, the patient's neurologist prescribed topiramate 50 mg twice a day for migraine prophylaxis.

The topiramate was tapered off during the first 2 days of monitoring. There was video confirmation of her hearing a loud bang sound correlating with the transition from NREM stage 1 sleep to wake (**Figure 1**). She had normal sleep architecture, and a total of 11 typical episodes were recorded, most associated a leg jerk. No evidence of seizure activity was noted during monitoring. The patient had been placed on topiramate for migraine headaches, and this was increased to 150 mg daily. Two months after admission, she reported improvement in the intensity of the noise. At a daily dose of topiramate 200 mg, the patient reported the bang had significantly improved, and now sounded like a low buzzing noise. The frequency of the events was unchanged, but the intensity of the events decreased to the point of being mildly noticeable. She had marked improvement in subjective ability to fall asleep and felt these events were no longer disruptive.

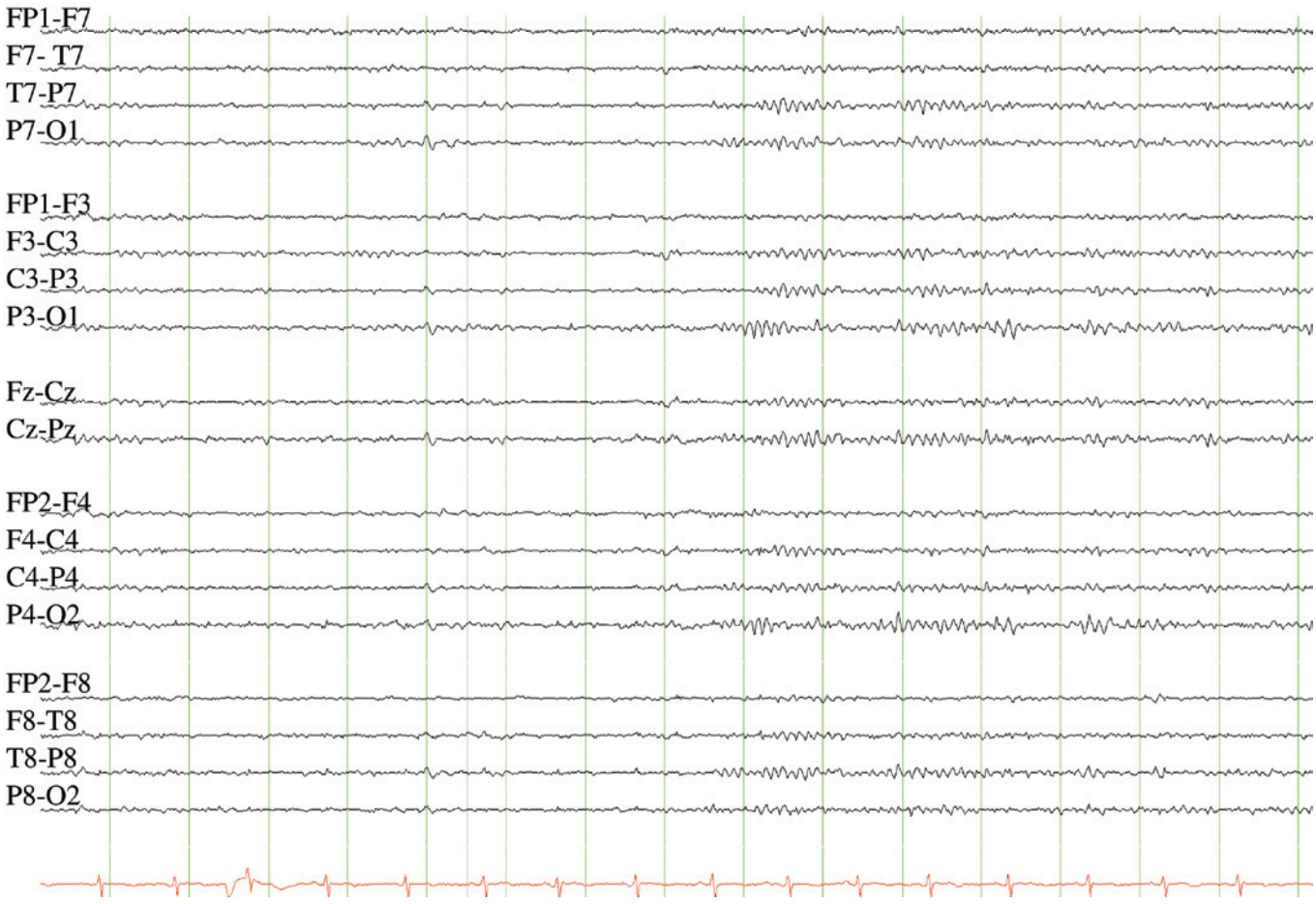
DISCUSSION

We describe a case of EHS in which topiramate diminished the intensity but not the frequency of the events. Our case has classical features of EHS and some interesting observations. For our patient, some of her events were associated with a hypnic jerk. This was not described by Pearce in his original description. Our patient also had history of migraines. Migraines or headaches were also mentioned in three of seven reports; otherwise, no linkage has been made between EHS and headaches.

Our patient also noted a family history of these events. The patient's mother and daughter have similar symptomatology, rasing the possibility that this may be hereditary. There is no literature on the inheritance of this phenomenon. Jacome postulated that EHS might arise from transient calcium channel dysfunction, such as seen in familial hemiplegic migraines resulting from mutation in CACNA1A gene located in chromosome 19. The gene encodes for the alpha 1A subunit of the neuronal P/Q type voltage-gated calcium channel.[6] Mechanism of topiramate is on P type calcium channel.[1] However, to date, no mechanism has been linked to EHS.

Various therapies have been tried to improve the EHS events. Case reports have implicated that clonezapam,[3] clomipramnine,[4,5] and nifedipine[6] have been helpful. There are two case reports by Chakravarty that show benefit with flunarizine.[5] Treatments with doxepin, citalopram, trimipramine, and amitriptyline were unsuccessful.[3] Jacome found no benefit from valproic acid, amitriptyline, propranolol oxycodone, or gabapentin.[3,6] Our case had improvement in intensity of events with the use of topiramate.

Figure 1—Electroencephalographic recording during an event during which the patient heard the loud noise.



The patient aroused from stage 1 sleep.

Exploding head syndrome is a rare phenomenon but can be disruptive. This case report demonstrates and alternative and safe method to treat these events with topiramate medication.

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