CASE REPORT

Exploding head syndrome – the early steps

Síndrome da cabeça explodindo – os primeiros passos

Elcio Juliato Piovesan[1,2], Pedro André Kowacs[1], Helder Granhold Campos[3], Lucas Pires Augusto[3], Lucas Coluni[3], Lineu Cesar Werneck[1,2]

[1]Neurology Service, Internal Medicine Department, Hospital de Clínicas da Universidade Federal do Paraná (UFPR), PR, Brazil
[2]Experimental Laboratory, Health Sciences Sector (LESCS), Universidade Federal do Paraná (UFPR), PR, Brazil
[3]Faculdade de Medicina, Universidade Federal do Paraná, PR, Brazil. *Sponsored by a grant from Conselho Nacional de Desenvolvimento Científico e Tecnológico (CNPq)*

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ABSTRACT

Exploding head syndrome is a rare entity associated with migraine that occurs during sleep onset. A male migraine with aura patient presented with episodes of abrupt awakening following perceptions of sounds resembling a speeding up motorcycle engine interspersed with bursts of exhaust explosions like noises, accompanied by an exploding sensation in the head. The patient presented in the evolution of self-limited period of headache chronicity. This syndrome has been associated with an atypical form of acoustic aura that often leads to migraine chronification.

Keywords: Acoustic aura; Exploding head syndrome; Migraine.

RESUMO

A síndrome da cabeça explodindo é uma entidade rara associada com a migrânea que ocorre durante o início do sono. Um paciente do sexo masculino com migrânea com aura apresenta episódios de despertar súbito após perceber um som como uma motocicleta acelerando intercalada com estouros de um escapamento. O paciente evoluiu com período autolimitado de cronificação da cefaleia. Esta síndrome tem sido relacionada a uma forma atípica de aura acústica e aparenta intima relação com cronificação da migrânea.

Palavras-chave: Aura acústica; Migrânea; Síndrome da cabeça explodindo.

INTRODUCTION

Exploding head syndrome (EHS) is a rare phenomenon characterized by a painless loud noise at the onset of sleep.[1] Armstrong-Jones described it for the first time in 1920 and referred to it as "snapping of the brain".[2] Pearce coined the name "exploding head syndrome in 1989.[3] This is a rare benign sleep-wake transition disorder of unknown aetiology.[4] The attacks are characterized by a sudden "bomb-like explosion" or "shotgun" noise felt in the head and in 10-20% of patients there is also a sensation of "flashing lights".[4] The attacks are not painful but are unpleasant.[4,5] This phenomenon occurs in relaxed wakefulness or at the transition from wakefulness to sleep.[5] The sensation lasts for a few seconds only and disappears completely when awake, although it may recur on further attempts to fall asleep.[6] The onset is usually over the age of 50 years old and there is a slight female preponderance.[3] The attacks occur with variable frequency (from seven in one night to one in a few weeks or months).[6] Symptoms such as nausea and vomiting did not occur.[6] The vast majority of patients with EHS are migraine with aura patients. Reports of patients with EHS preceding the onset of migraine attacks suggest that EHS can be considered as a form of migraine aura.[7]

Here we present a new EHS case and compare it characteristics to those of the cases described in the literature.

CASE REPORT

A 45-year-old man with a five years history of episodic migraine with fortification spectra aura described a peculiar sensation in the head, occurring once a year, similar to the noise of an exploding bomb only at night while going off to sleep. The "explosion" would wake him up and disappear completely at the moment he woke up. This would make him wake up extremely scared and tachycardic. Regarding the last episode, the patient described a sound like the one of a motorcycle being accelerated followed by exhausting pipe bursts. Three of these sequences of sounds were perceived until the patient was awake (Figure). The patient observed a close relationship to anxiety. After EHS episodes, the patient reported migraine exacerbation, lasting 45 days. The headache has been well described as migraine: alternating unilateral, throbbing, disabling and associated to nausea, phonophobia and photophobia, besides important and persistent visual phenomena. General physical examination was normal, as it was the neurological examination, including mental status, cranial nerves, muscle strength, muscle tone, stretch and superficial reflexes, cerebellar function, gait and sensory testing. Impedance and audiometry tests were normal, as well as magnetic resonance imaging and magnetic resonance angiography of the brain.

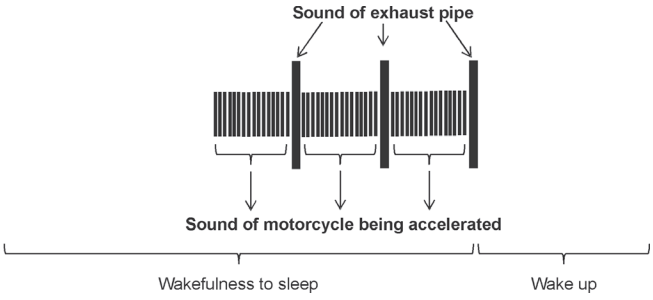

Figure – Sounds described by the patient

DISCUSSION

In this syndrome, the sudden start of the symptoms resembles thunderclap headache. As the patient is not yet dreaming, these sounds occur in a context totally unknown to the patient. Maybe this is why patients wake up very scared, looking for the source of the noise. This is a rare disorder and our experience is limited to one case. Even after an exhaustive literature review, only a few cases seem to have been shared in almost 100 years of its initial description.

The population affected by this syndrome is usually also stricken by migraine with aura. From the standpoint of pathophysiology, this syndrome cannot be confused with nocturnal epilepsy since tests such EEG and polysomnography (PSG) during EHS attacks do not suggest this etiology.[8,9] On video PSG and multiple sleep latencies test (MSLT), EHS attacks showed at the transition from wakefulness to sleep (non-rapid eye movement (NREM) sleep stage 1, NREM1) and from NREM2.[4] EHS occurs at any age but usually occurs after age 50. A gradual increase of stage 1 sleep occurs with brain aging.[6,12] The basis for EHS is thought to be a delay in the reduction of activity in selected areas of the brainstem reticular formation as the patient passes from wakefulness to sleep.[6,13]

Many speculations had been done, especially after the use of drugs that were able to control symptoms in isolated cases. The existence of a transient calcium channel dysfunction was hypothesized as a cause, since the nifedipine,[10] flunarazine,[6] and topiramate (P type calcium channel)[1] produced improvement of the symptoms. Other drugs have shown satisfactory results, as clonazepam([11]) and clomipramine.[9-13]

The EHS attacks occur in relaxed wakefulness or at the transition from wakefulness to sleep.[4,5] A very interesting way patients, such as our, have reported that the onset the EHS is directly associated with a worsening of migraine taking some clinical aspects of chronicity.[4] Recent work has suggested that EHS is considered an atypical acoustic aura.[4,7]

Two hypotheses suggest a momentary disinhibition of the cochlea or its central connexions in the temporal lobes,[3] sudden involuntary movement of the tympanum or the tensor tympani,[3] rupture of the labyrinthine membrane or a springing open of the Eustachian tubes.[3,14] Our case suggests a central origin since the sounds are not only more elaborate single explosion.

In summary the exploding head syndrome is extremely rare, occurs in patients with migraine, seems to be associated with a clinical worsening of migraine and is considered a form of acoustic migraine aura.

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Correspondence
Elcio Juliato Piovesan, MD
Rua General Carneiro, 1103/102
80060-150 – Curitiba, PR, Brasil
piovesan1@hotmail.com