Exploding Head Syndrome as Aura of Migraine with Brainstem Aura: A Case Report

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This article reports a case of exploding head syndrome (EHS) as an aura of migraine with brainstem aura (MBA). A middle-aged man presented with intermittent episodes of a brief sensation of explosion in the head, visual flashing, vertigo, hearing loss, tinnitus, confusion, ataxia, dysarthria, and bilateral visual impairment followed by migraine headache. The condition was diagnosed as MBA. Explosive head sensation, sensory phenomena, and headaches improved over time with nortriptyline. This case shows that EHS can present as a primary aura symptom in patients with MBA. *J Oral Facial Pain Headache 2018;32:e34–e36. doi: 10.11607/ofph.1950*

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Migraine with brainstem aura (MBA), previously called basilar migraine, is a rare form of migraine with aura characterized by recurrent and reversible attacks of brainstem symptoms such as vertigo, dysarthria, tinnitus, and diplopia, followed by migraine.[1,2] Exploding head syndrome (EHS) is also a rare disorder characterized by brief episodes of sudden imaginary noise or the feeling of an explosion, shotgun, or thunder, occasionally accompanied by visual flashes.[3,4] The etiologies of MBA and EHS remain elusive, but brainstem dysfunction is suspected in both conditions. MBA and EHS are considered unrelated and seldom described in the same individuals. EHS has been reported twice as aura in migraines[5,6] and once in association with stabbing headaches.[7] However, EHS has never been reported as aura in a case of MBA; thus, the following appears to be the first report of such an association.

Case Report

A 49-year-old African-American male presented with an 18-month history of episodic headaches lasting 5 to 20 minutes that were intense to very intense, throbbing, located mainly in the right occipital-parietal region, and at times bitemporal. Headaches were associated with nausea, photophobia, and phonophobia and occurred randomly throughout the year with a frequency of one to three episodes per month. These headaches were all preceded by a snapping sensation on the head with the feeling of an explosion, visual flashing, hearing loss, vertigo, tinnitus, confusion, ataxia, dysarthria, and bilateral visual impairment. Hearing loss was subjective (based on the patient's report) and limited to the headache episodes only; hearing was normal between attacks. Audiometry was not performed. The explosion and flashing lasted 1 to 2 seconds, and the rest of the sensory phenomena lasted up to 5 seconds. The patient could not actually hear the explosion, but felt it inside his head as an explosion. Twice during these exploding episodes he experienced a brief loss of consciousness for a few seconds where he fell on the ground. All these events occurred during the daytime while the patient was fully awake and alert. The patient denied ptosis, eyelid edema, conjunctival injection, lacrimation, rhinorrhea, facial

sweating, or psychomotor agitation. He suffered from tension-type headaches for many years from mild to moderate headache with a vice-like distribution, a duration of more than 4 hours, and without photophobia, phonophobia, or nausea. These headaches were very different from this new, short-lasting severe headache.

The patient was diagnosed with MBA. Medical history was pertinent for hypertension, hypercholesterolemia, long-standing type 2 diabetes mellitus, tobacco dependence, and remote history of alcohol abuse. Family history was negative for migraines, EHS, and psychiatric illness. General and neurologic examinations were unremarkable. Findings from brain magnetic resonance imaging (MRI) and electroencephalography (EEG) were also unremarkable. Polysomnography showed primary snoring, but no sleep apnea. Genetic testing for familial hemiplegic migraines did not identify any genetic mutations of ATP1A2 (ATPaseNa+/K+ transporting subunit alpha 2), CACNA1A (calcium voltage-gated channel alpha subunit 1A), or SCN1A (sodium voltage-gated channel alpha subunit 1). EHS, sensory aura, and migraine headaches were chronically reduced to four per year after the patient started nortriptyline 50 mg every night. No side effects were reported. Attempts to discontinue nortriptyline were associated with recurrence of all symptoms.

Discussion

According to the International Classification of Headache Disorders Third Edition beta version (ICHD-3 beta), the diagnosis of MBA is based on at least two reversible attacks of one or more typical aura symptoms (eg, visual, sensory, or speech/language) and two or more brainstem aura symptoms (eg, vertigo, dysarthria, tinnitus, diplopia, ataxia, hypoacusis, and/or decreased levels of consciousness).[2,8] Aura may last between 5 and 60 minutes, simultaneously or in succession, and followed or accompanied by migraine.[1,2] Among all patients with MBA, 31% report two, 45% report three, 8% report four, 8% report five, and 8% report six aura symptoms.[1] Headache is usually bi-occipital[1] and is otherwise not different from other forms of migraine with or without aura. Motor symptoms are not present.[1,2,8] This patient fulfilled the criteria for diagnosis of MBA, though his aura and headaches were shorter in duration than the typical MBA (where aura lasts at least 5 minutes and migraine at least 4 hours).

Vertigo is the most common symptom in MBA, and so MBA may be misdiagnosed as vestibular migraine (VM), which is a form of migraine with moderate to severe vestibular symptoms (according to ICHD-3 beta).[1,2,8] A recent study differentiated MBA from VM based on the high prevalence of vertigo with each MBA attack, neurologic symptoms other than vertigo, and saccadic dysmetria during vestibular function tests. In the present case, the presence of vertigo with each attack and neurologic symptoms other than vertigo supported the diagnosis of MBA. No vestibular function tests were conducted in this patient.[9]

A genetic association between Familial Hemiplegic Migraine (FHM) and MBA has been reported in three first-degree relatives with MBA and the ATP A2 gene mutation seen in FHM type 2.[10] MBA has also been genetically associated with episodic ataxia type 2 (EA2) in adolescent males with MBA, EA2 attacks, and a novel nonsense mutation of the CACNA1A gene seen in FHM type 1 (FHM1).[11] The present patient tested negative for all mutations of FHM, including ATP A2, CACNA1A, and SCN1A DNA sequencing. Posterior circulation transitory ischemic attacks were considered unlikely based on multiple recurrent attacks of fully reversible symptoms not associated with any residual clinical deficits or multimodal MRI abnormalities.

EHS is an uncommon disorder presenting with sudden imagined loud noise, such as an explosion, shotgun, thunder, or a sense of explosion within the head. Light flashes are present in 10% of patients. EHS episodes last seconds and occur during the transition from wakefulness to sleep or vice versa. EHS mainly occurs in individuals older than 50 years of age, with a female predominance.[3,4,12] EHS is a benign syndrome without neurologic sequelae and diagnosed by history alone.[3,4] In this case, the clinical description conformed to the definition of EHS, although it was unusual that episodes solely occurred during the daytime while the patient was fully awake.[13]

EHS episodes are considered painless, however, EHS has been associated with primary headaches. A 71-year-old female reported complaints of EHS in association with episodes of generalized muscle twitching and right parietal stabbing pain in the process of falling asleep at night. Headaches, myoclonic jerks, and EHS resolved with slow-release Nifedipine.[7] Also, EHS has been reported as an aura in two migraine cases. The first case was a middle-aged man with exacerbation of chronic migraines with each attack of EHS.[5] The second case was a young Hispanic woman with episodes of EHS that lasted for a few seconds, followed by sleep paralysis for 6 seconds, and then followed by bitemporal migraines lasting for hours.[6] In all cases, EHS lasted seconds, which is unusually brief for migraine aura.

The pathophysiologies of MBA and EHS remain elusive. In MBA, dysfunction of brainstem regions involved in pain modulation is supported by positron

emission tomography scans taken during migraine attacks and showing increased activity at the locus coeruleus, dorsal raphe nuclei, periaqueductal gray, and midbrain reticular formation.[14,15] EHS could result from sudden movements of the middle ear structures or Eustachian tube or from rupture of the labyrinthine membrane.[3,12] Transient voltage-gated calcium channel dysfunction, as seen in FHM type 1 CACNA1A gene mutation, has also been suggested.[7] Recently, brainstem dysfunction with delay in the inactivation of selected brainstem areas during the transition from wakefulness to sleep has been proposed in the pathogenesis.[13,16] However, this hypothesis cannot explain EHS being exclusively present during the daytime while the patient was fully awake, as occurred in the present case. Emotional stress can be a trigger for EHS as well as for migraine; however, this association has not been clearly proven in EHS yet, since no formal neuropsychologic studies were ever conducted in patients with EHS, although there are self-reported psychological complaints.[12]

Conclusions

This article reports a case of EHS as an aura of MBA. The duration of migraine and aura was shorter than in typical migraines with aura; however, the consistency of the presentation of EHS, brainstem symptoms, and migraines in each single attack and the parallel improvement of all symptoms with nortriptyline and worsening with its discontinuation suggest a common pathophysiology. To the authors' knowledge, this is the first case of EHS as aura of MBA. It is suggested that MBA and EHS are probably caused by brainstem dysfunction that results in activation of the trigeminovascular system. This report has the limitation of being a single case, and further studies are required to clarify this association.

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