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CASE REPORT

A singular association of migraine with brainstem aura and Alice in Wonderland syndrome

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Abstract

Background In this work, we describe an association of brainstem headache with aura (BHA) and Alice in Wonderland syndrome (AIWS) in a 17-year-old male, suffering from crises of vertigo, weakness, dysarthria, and diplopia, in half-hour duration, followed by diffuse or occipital headache, lasting several hours.

Methods The frequency of the attacks was monthly, and once there was short loss of consciousness. The last episodes were accompanied by symptoms such as deformation of figures and objects, small or large in shape.

Results Diagnostic examinations were performed, mainly neuroimaging tests such as brain MRI and brain angio-MRI, all resulting normal; and treatment with flunarizine was followed by improvement of both BHA and AIWS symptoms.

Conclusions There would be a correlation between BHA and AIWS, presumably represented by dysfunction of temporo-parieto-occipital carrefour.

Keywords Brainstem headache · Alice in Wonderland syndrome · Carrefour region

Introduction

The International Classification of Headache Disorders, 3rd edition (ICHD.3) (2018) inserted brainstem headache with aura (BHA) (1.2.2) in the chapter of migraine with aura (1.2). Previously used terms were basilar artery migraine, basilar migraine, or basilar-type migraine. Symptoms clearly originating from the brainstem, without motor weakness [1]. They include dysarthria, vertigo, tinnitus hypacusis, diplopia, ataxia not attributable to sensory deficit, and decreased level of consciousness. The majority of the patients include young people and children with female predomination. The onset of the disease usually occurs before the age of 25. Differential diagnosis should exclude pathology of posterior fossa, cerebellar diseases, complex epileptic seizures, as well as CADASIL and MELAS syndromes. In the prophylaxis sodium valproate and calcium-antagonists and, especially in vertigo, betahistine chloride is used [2]. Alice in Wonderland syndrome (AIWS) named for Lewis Carroll's novel, is a disorder characterized by transient episodes of visual hallucinations and perceptual distortions, during which objects or body parts are perceived as altered in various ways (metamorphopsia), including enlargement (macropsia) or reduction (micropsia) in the perceived size of a form [3]. Additionally, patients with Alice in Wonderland syndrome can experience auditory hallucinations and changes in their perception of time [4]. Currently, there is no known specific cause of AlWS. Some theories point to infections such as the Epstein-Barr virus, medications such as topiramate, and migraines. Neuroimaging has revealed brain regions involved such as temporo-parieto-occipital junction or carrefour region [5]. There are no current specific treatments for AlWS.

Case description

A 17-year-old male reported episodes of vertigo, weakness, dysarthria, and diplopia, in half-hour duration, followed by diffuse or occipital headache lasting several hours. The

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frequency of the attacks was monthly and once there was short loss of consciousness. The last episodes were accompanied by symptoms such as deformation of figures, and objects in smaller or larger shaped. The body was altered by sensations such as loss of sensory stimuli along the limbs and lack of spatial perception. Brain MRI with gadolinium, Angio-MRI, laboratory analysis, echocardiogram, audiometric examination and vestibular tests, basal electroencephalogram, and with activation tests such as sleep deprivation and sleep, ultrasound doppler of supra-aortic, and transcranic vessels were normal. He did not refer to familiarity with epilepsy or migraine. The patient was treated with calcium-antagonists, anti-depressant, anti-platelet drugs, without bringing any benefit. We administered flunarizine at dose of 10 mg daily; noting in the last 2 months, an improvement of the symptoms of the brainstem aura, in particular diplopia, while the metamorfopsias persisted, though of minor duration.

Discussion

An association of two disorders characterized by the presence of double aura is a singular event. But what are the mechanisms that determine such a condition in the same individual? BHA was initially considered as a vascular pathology caused by spasms of the basilar artery. The vascular hypothesis has not been proved, and is now believed to be a type of migraine with aura. BHA location of symptoms is brainstem or both occipital hemispheres, while, in a migraine with typical aura, it mainly involves unilateral hemisphere [6]. BHA is often more disabling than a migraine without aura and migraine with typical aura, because of increased severity and longer duration of symptoms. An important study was conducted by Kirchmann who recruited 105 families comprising 362 patients with migraine aura (MA). Among these patients, 38 patients from 29 families had BHA. In 12 of the BHA families with an apparently dominant inheritance, the authors found in chromosome 19 and in chromosome 1 genes responsible for most cases of the autosomal dominantly inherited familial hemiplegic migraine, resulting a linkage between two auras. BHA occurred in 10% (38/362) of patients with MA. BHA had a median duration of 60 min and comprised vertigo 61%, dysarthria 53%, tinnitus 45%, diplopia 45%, bilateral visual symptoms 40%, bilateral paresthesias 24%, decreased level of consciousness 21%, hypacusia 21%, and ataxia 5%. The relative frequency of the individual BHA symptoms was not different from patients with hemiplegic migraine from a previous study [7]. The attacks of MA were identical in families with or without BHA. BHA may occur at times in any patient with MA [8].

Ilik found a combination of visual and somatosensory auras occurring before the headache phase. Aura visual symptoms included the perception of flashing lights, while somatosensory aura comprised numbness and tingling in the lips or fingers, involving a profound alteration of the perception of space and time [9].

There is a clinical co-occurrence of depressive episodes and AIWS. Altered perception of body image and sense of time and space can be similar to depression. Brain alterations may be observed as metabolic abnormalities in visual and parieto-temporal association cortices on PET images, About pathophysiology of both AIWS and BHA, in the mechanisms underlying AIWS, there would be the involvement of the occipital, temporal, and parietal lobes [10]. Occipito-temporal lobes are considered responsible for visual disorders such as phosphenes scotoma, teichopsias, while alterations of the parietal lobe cause the lack of spatial location and appearance of visual perceptions, such as dysmetropsias. Temporo-parieto-occipital junction or carrefour region is considered starting point of AIWS [11, 12]. The case described suggests some considerations: BHA is a form of migraine autonomic aura, without involvement of the cortical spreading depression, which is a cortical phenomenon. Therefore, the link between the two auras might be the region of carrefour, common site of somatosensory and visual auras. Then, the efficacy of flunarizine, a drug used in migraine syndromes, both in migraines and in AIWS, would not explain the effect on the BHA, which is a brainstem headache.

Conclusions

An association of brainstem headache and Alice in Wonderland syndrome is present in a young male. Brainstem headache is a rare form of migraine with aura. Aura consists of autonomic signs, while temporo-parieto-occipital carrefour would be responsible for visual and somatosensory aura. The case is singular and adds new hypotheses about mechanisms of headache auras.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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