CASO CLÍNICO/CASE REPORT

Cluster Headache Secondary to Trauma Presenting with Unilateral Mydriasis

Cefaleia em Salvas Secundária a Trauma com Midríase Unilateral à Apresentação

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Abstract

Cluster headache is the most common trigeminal autonomic cephalalgia, it can be a primary or secondary headache. Attacks are frequently associated with autonomic symptoms, due to parasympathetic activation and sympathetic defect. An 86-year-old woman, with no previous history of headache, developed a right supraorbital headache, two weeks after a fall, that resulted in mild head trauma to the ipsilateral frontal region. She complained of intense, daily attacks, with a circadian rhythm, lasting approximately 50-60 minutes, without psychomotor agitation. During the episodes, she noted right monocular blurred vision, and occasionally perceived colored shapes. During the attacks, right eye mydriasis was seen, without other autonomic signs. The diagnosis of cluster headache was admitted and started symptomatic treatment with oxygen and prophylaxis with verapamil, with unequivocal improvement. We pretend to highlight this complex and poorly understood relation between trauma and cluster headache, as well as emphasize the atypical autonomic manifestations of this entity.

Resumo

A cefaleia em salvas é a mais comum das cefaleias trigémino-autonómicas, podendo tratar-se de uma cefaleia primária ou secundária. As crises associam-se habitualmente a sintomas autonómicos, resultantes de ativação parassimpática e defeito simpático. Mulher de 86 anos, sem antecedentes de cefaleia, desenvolveu, duas semanas após queda com trauma ligeiro da região frontal direita, cefaleia supraorbitária ipsilateral. Apresentava crises diárias, intensas, com ritmo circadiano, duração de 50-60 minutos e sem agitação psicomotora associada. Durante os episódios, descrevia sensação de visão turva, ocasionalmente perceção de formas coloridas e apresentava midríase do olho direito, sem outros sinais autonómicos acompanhantes. Admitiu-se cefaleia em salvas e foi iniciado tratamento sintomático com oxigénio e profilaxia com verapamil, com melhoria significativa. Pretendemos evidenciar a complexa e ainda pouco esclarecida relação entre o trauma e a cefaleia em salvas, bem como alertar para as possíveis manifestações autonómicas atípicas desta entidade.

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Introduction

Cluster headache is the most common of the trigeminal autonomic cephalalgias and it has a prevalence of 0.1% of the population.[1,2] It is characterized by severe unilateral pain, located in the orbital, supraorbital, or temporal regions with a duration of 15-180 minutes. Cluster headache attacks are frequently associated with autonomic symptoms.[1] Parasympathetic activation results in lacrimation, conjunctival injection, and rhinorrhea while sympathetic defect originates miosis and ptosis.[3] However, the pathogenesis of these manifestations is not completely clear, and there may be atypical symptoms.

We report a case of probable cluster headache with some unusual manifestations, namely regarding autonomic symptoms. There are reported cases of mydriasis that propose activation of the sympathetic nervous system as a possible etiology. With this report, we intend to highlight this complex entity, whose pathophysiology is not yet fully understood.

Case Report

An 86-year-old woman, with multiple vascular risk factors and degenerative osteoarticular pathology, presented with a right supraorbital headache, that started two weeks after a fall with mild trauma to the ipsilateral frontal region. She complained of a daily headache, with circadian rhythm, initially occurring always at 7 pm, but with the progression of the clinical picture, sometimes also at 3 pm. It was difficult to define the character of the headache, but the patient described it as an intense headache lasting 50-60 minutes. During this time, she was not agitated or restless. Concomitantly, she described a right monocular blurred vision sensation, that occasionally described as perception of colored shapes that lasts less than 20 minutes. She denied nausea, photophobia and phonophobia, as well as previous history of headache. She reported myalgias and symptoms compatible with possible mandibular claudication, although with no other complaints suggestive of giant cell arteritis. The neurological examination in the emergency department revealed anisocoria, with the right pupil 1.5 mm larger than the left and motor difficulties caused by the osteoarticular pathology, but she had no other abnormalities. She was examined by an ophthalmologist, who diagnosed bilateral cataracts, excluding glaucoma and retinal pathologies. Because of the atypical signs, the patient was hospitalized for investigation. During hospitalization, she maintained the attacks, during which we observed mydriasis of the right eye, without other autonomic signs and with resolution between the attacks. We carried out an extensive investigation to exclude potentially serious secondary causes. Laboratory tests, including repeated evaluation of erythrocyte sedimentation rate and C-reactive protein; cranial and orbit magnetic resonance imaging (MRI); intracranial and extracranial vessel computed tomography (CT) angiography; and electroencephalogram were unremarkable. We considered the diagnosis of probable cluster headache secondary to trauma, given the features of the headache and after exclusion of other secondary causes. Symptomatic treatment with oxygen and prophylaxis with verapamil were started. The patient showed a significant clinical improvement, so she was discharged after starting this treatment. One month after discharge, the patient had only two attacks, with a good response to oxygen. She was then diagnosed with aortic stenosis, so verapamil was discontinued. She completed a month and a half of treatment with verapamil and considering the clinical improvement, no other prophylactic was initiated. One month after discontinuing verapamil, she continues without recurrence of attacks.

Figure 1. Mydriasis of the right eye during an attack.

Discussion

The features of pain, its circadian rhythm and the response to therapy are some of the clues that suggest a probable cluster headache diagnosis. However, the patient's age, the onset after trauma, the behavior during the attack, the visual manifestations suggestive of aura, and the mydriasis, are some of the atypical symptoms in this case. In this context, these unusual manifestations led us to exclude other important causes of secondary headache.

Initially, it was considered the possibility of a vascular cause. CT-angiography excluded arteriovenous malformations and arterial dissections. Inflammatory disorders may also produce symptoms resembling cluster headache and considering the patient's age and the systemic symptoms initially reported, it was also important to exclude the hypothesis of giant cell arteritis. Repeated evaluation of erythrocyte sedimentation rate and C-reactive protein were normal, making this diagnosis less likely. There were no other symptoms or imaging abnormalities that suggested another inflammatory or infectious disease. Neoplasms, like pituitary tumors and metastases, can also mimic primary cluster headache, and brain MRI also excluded these possible etiologies. Furthermore, orbit MRI allowed us to exclude eye, ocular muscles, or optic nerve pathology.

The differential diagnosis for cluster headache also includes other primary headache disorders, however, the features of our patient's headache did not meet the criteria for any other primary headache. The visual symptoms described by our patient could lead us to consider the hypothesis of migraine, however, migraine-like aura symptoms have been reported in up to 23% of cluster headache patients.[4] Admitting the diagnosis of probable cluster headache, there was still an atypical symptom that raised concern and prompted further investigation. Typically, this type of headache is accompanied by miosis resulting from sympathetic defect, but during attacks our patient presented mydriasis. For this reason, we completed the investigation with cervical magnetic resonance imaging, which did not demonstrate any structural lesion of the descending sympathetic pathway.

After exclusion of other secondary causes, we considered the diagnosis of probable cluster headache secondary to trauma. Head trauma is a condition that has been associated with cluster headache and some reviews of the literature point up to 16.5% of patients with cluster headache and a history of head injury.[5,6] An important review published in 2006 by Manzoni *et al*, clearly demonstrated the significant relation between head trauma and cluster headache.[7] The temporal relationship between headache onset and head injury can be variable, ranging from immediate onset to 30 years after the trauma.[5] Turkewitz *et al*, reported one case of post-traumatic cluster headache, with clinical symptoms emerging 6 days after the trauma, so with close proximity between head injury and the onset of cluster headache.[6] There are also reports of symptoms initiated a few weeks after trauma, similar to our case.[7] However, the nature of this association is still unclear. Two hypotheses have been proposed, head trauma may damage extra or intracranial peripheral or central nervous structures, leading to the development of cluster headache or head injuries may be more frequent in this population because of their lifestyle, which leaves them more exposed to risk.[7] We hypothesize that this unusual and less studied etiology could also explain some of the uncommon manifestations in our patient, since the pathophysiology of the disease may not be the same as the primary cluster headache.

Cluster headache attacks are frequently associated with autonomic symptoms.[1] It has been extensively demonstrated that parasympathetic activation results in lacrimation, conjunctival injection, and rhinorrhea while sympathetic defect originates miosis and ptosis.[3,8,9] However, there have been reports of cluster headache associated with mydriasis, as well as other autonomic symptoms, like eyelid retraction and hemifacial hyperhidrosis, the known Pourfour du Petit syndrome.[10] Jeremy Nadal *et al*, recently reported a case of a 48-year-old woman with cluster headache and Pourfour du Petit syndrome.[10] There are also similar reports in other trigeminal autonomic cephalalgias, like one case in short-lasting unilateral neuralgiform headache.[11] It was proposed that these atypical autonomic symptoms were caused by hyperactivity of the ipsilateral sympathetic pathway.[10] We also found in the literature another report of persistent isolated mydriasis as an early sign of internal carotid artery dissection, in which the possibility of irritative mechanisms of the sympathetic nervous system causing this hyperactivity is reinforced.[12] However, the exact trigger and which factors determine the hyperactivity or hypofunction of the sympathetic pathways in headache is not well established.

This was a challenging case, not only for the rarity of the complete clinical picture but also for its many unusual manifestations. Although our case does not meet the international headache criteria for cluster headache or post-traumatic headache, we consider cluster headache secondary to trauma to be the most likely diagnosis. We reported this case, for two main reasons: the onset of cluster headache after trauma (in an elderly woman with no previous history of headache) and the occurrence of mydriasis instead of miosis as expected. We intend to emphasize that cluster headache is a complex entity, and when there is an unusual manifestation or atypical finding, we should keep in mind possible secondary etiologies. There seems to be a relation between cluster headache and trauma, making it important to explore the previous history of head injuries in our patients. The pathophysiology of the autonomic symptoms in trigeminal autonomic cephalalgias is not yet well established. Cluster headache can rarely present with mydriasis due to sympathetic pathways hyperactivity, resembling Pourfour du Petit syndrome. ■

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