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Case report

Segmental cavernous carotid ectasia in a patient with cluster-like headache

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ABSTRACT

Introduction: Cluster headache (CH) is a primary headache with severe, unilateral periorbital or temporal pain lasting 15–180 min, accompanied with various cranial autonomic features. A diagnosis of cluster-like headache can be made whenever underlying cause of CLH is present.

Methods and results: We report a case where an ectatic cavernous segment of the internal carotid artery triggered CHL, most probably due to compression of the ophthalmic nerve within cavernous sinus. The pathological substrate of a vessel ectasia is degeneration of the *tunica intima* as a consequence of atherosclerosis and hypertension. On the other hand, cavernous sinus is unique space where parasympathetic, sympathetic and nociceptive fibers are in intimate relationship which is of great importance for understanding of CH pathophysiology.

Conclusion: Magnetic resonance imaging and MR angiography are mandatory imaging tools used for precise localization of pathological changes in the cavernous sinus, especially in the group of secondary headaches attributed to vascular disorders.

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1. Introduction

According to the criteria given in the third edition of the International Classification of Headache Disorders (beta version) (ICHD-3 beta), cluster headache (CH) is a primary headache disorder characterized by attacks of severe, strictly unilateral orbital, supraorbital or temporal pain lasting 15–180 min and occurring from once every other day to eight times a day [1]. The attacks are associated with at least one of the following cranial autonomic features, all of which are ipsilateral to the side of pain: conjunctival injection and/or lacrimation, nasal congestion and/or rhinorrhea, forehead or facial sweating and flushing, miosis, ptosis, eyelid edema, and sensation of fullness in the ear. The sense of restlessness or even agitation is often present during the attacks. In case that underlying causative conditions are present, a diagnosis of secondary cluster-like headache should be made (CLH).

An extended review of 156 symptomatic, cluster-like headache cases showed that the more frequent pathologies mimicking primary cluster headache were vascular disorders, followed by tumors and inflammatory infectious diseases [2]. Various vascular pathology involving trigeminovascular system can trigger CLH: pseudoaneurysm within cavernous sinus [3], dural fistula [4], carotid artery dissection [5–8], arteriovenous malformation [9] and subclavian steal syndrome [10].

The term ectasia refers to distension of the arterial lumen due to deterioration of the *tunica intima* and weakening of the vessel wall. The main etiologic factors of dolichoectasia are atherosclerosis, defects or destruction of the internal elastic, and hypertension [11,12].

We present a previously unreported case with an offending vascular cause of CLH in the form of an ectatic cavernous segment of the carotid artery compressing the ophthalmic nerve.

2. Case report

Magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA) were ordered in a 49-year-old, non-smoker man with unilateral pain affecting left ophthalmic nerve distribution. The pain was sharp, lasting around 30 min, mainly involving the left eye, spreading to the temporal region, associated with sensation of fullness of the ipsilateral ear. There were no other prominent autonomic features. The pain severity was graded as 9 or 10 on a ten-grade scale. It occurred once a day or every other day in a period of 30 days. Between the attacks, the patient experienced mild numbness and hyperalgesia in the affected region, but the pain could not be triggered by innocuous stimuli. A similar episode of recurrent pain occurred three years earlier, occurring on 45 days, but the symptoms regressed completely during the basic diagnostic survey. Medical history and physical and neurological examinations revealed no abnormalities. Otorhinolaryngologic and ophthalmologic findings were normal. The patient had no family history of headache.

Compressed MRA showed a moderate dilatation of the cavernous segment of the left internal carotid artery (ICA) (Fig. 1a, arrow), measuring 6 mm in diameter. T2 weighted (Fig. 1b) and contrast enhanced T1 weighted coronal images (Fig. 1c) revealed the ectatic segment producing a marked mass effect on the ophthalmic branch of the trigeminal nerve (Fig. 1b, c arrow). No other abnormalities were evident on brain MRI and MRA.

3. Discussion

Our case fulfilled the ICHD-3 beta criteria for cluster headache as regards the number, severity, site, duration and frequency of attacks. However, the presence of sensation of fullness in the ear in the absence of other autonomic signs and the absence of sense of restlessness or agitation, makes the case specific when considering the ICHD-3 beta criteria as opposed to the ICHD-2 [13]. Namely, the previous version required the presence of typical autonomic symptoms and the sense of restlessness and agitation in order for criterion C to be fulfilled and the diagnosis of CH to be made. Criterion C for CH in the ICHD-3 beta includes the requirement F – presence of sensation of fullness in the ear, which was fulfilled in our case. Though explicite criteria for CHL does not cite male gender, in our case this fact as well as presence of earlier cluster period and normal physical finding are consisted with

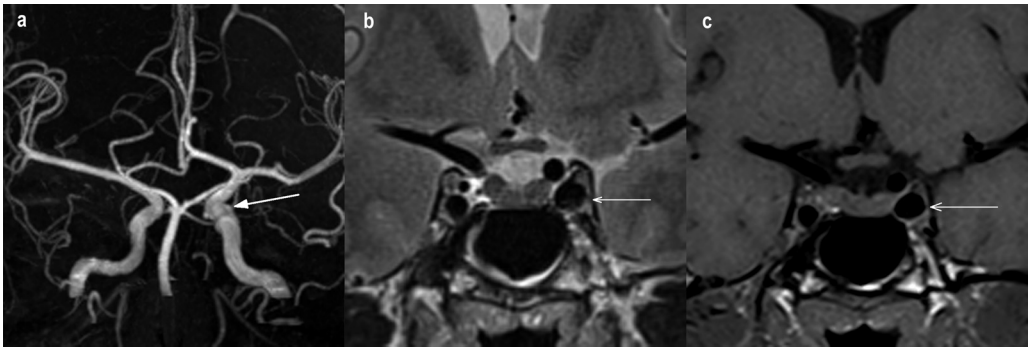

Fig. 1 – Moderate dilatation of the cavernous segment of the left ICA on collapsed MRA (arrow). On high resolution T2-weighted coronal and enhanced T1-weighted coronal images, conflict between left ectatic ICA and ophthalmic nerve is present (b, c, arrows).

diagnosis of CHL. CLH was also suspected due to the older age of our patient compared to typical age for CH (CH cluster periods characteristically start in the third decade of life, and very seldom before the age of 20, but average onset in CLH patients turns out to be at the age of 42.7 years [5]). Furthermore, our patient was a non-smoker, though smoking seems to be the most consistent lifestyle habit in patients with CH across studies [14], the most frequent trigger for CH was absent, and there was no data about periodicity characteristic of biorhythmic disorders. The clinically significant findings suggesting CLH were numbness and hyperalgesia of the affected area. Although allodynia was absent, the presence of hyperalgesia of the forehead may be interpreted as a sign of neuropathic component of pain. Symptoms and signs in the area of branch V1 of the trigeminal nerve indicated a pathology involving the cavernous sinus.

There is a substantial evidence of cavernous sinus role in the pathophysiology of CH [15–18] and a number of pathological processes in the cavernous sinus causing CLH have been described, including pituitary tumors [19–21], dural arteriovenous fistula [22], metastatic tumors [23] and inflammatory changes within cavernous sinus [24].

In our case, MRA and high resolution postcontrast MR study of the cavernous sinus showed conflict of the ectatic cavernous segment of the carotid artery with the V1 branch of the trigeminal nerve. In addition, the pain attributable to stimulation of the ophthalmic nerve by the ectatic blood vessel may be accompanied with parasympathic reflex activation, which in our patient manifested as ear fullness. Trigeminal pain provoked by vertebrobasilar ectasia was reported by Yu [25]. Our patient had a rare type of intracranial carotid ectasia without elongation, excluding the anomaly from the classic group of intracranial dolichoestasias.

This case confirms the significance of precise localization of pathological changes in the cavernous sinus using MRI and MRA according to Sjaastad's recommendations since 1990 [26]. We described carotid ectasia as a possible pathologic substrate that has not been described as a cause of CLH so far or as a case that belongs to broader group of secondary headaches attributed to vascular disorder according to the ICHD-3 beta version.

Conflict of interest

None declared.

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Ethics

The work described in this article has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans; Uniform Requirements for manuscripts submitted to Biomedical journals.

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