Cluster headache – a symptom of different problems or a primary form? A case report

Klasterowy ból głowy – objaw czy choroba samoistna? Opis przypadku

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Abstract

Headache with severe, strictly one-sided unilateral attacks of pain in orbital, supraorbital, temporal localisation lasting 15-180 minutes occurring from once every two days to 8 times daily, typically with one or more autonomic symptoms, is recognized as cluster headache (CH). Headache with normal neurological examination and abnormal neuroimaging studies, mimicking cluster headache, is reported by several authors.

We present an elderly woman with a cluster-like headache probably associated with other comorbidities. We differentiate between primary, but 'atypical' CH and symptomatic cluster headache due to frontal sinusitis, pontine venous angioma or vascular compression of the trigeminal nerve root. This headache is not so rare in the general population and its secondary causes must be ruled out before the diagnosis of a primary headache as cluster headache is made.

Key words: cluster headache, cluster-like headache, symptomatic headache, secondary headache.

Streszczenie

Klasterowy ból głowy jest bardzo silnym, jednostronnym bólem zlokalizowanym w okolicy oka, nadoczodołowej i skroniowej. Napad klasterowego bólu głowy trwa 15–180 minut, a liczba napadów wynosi od 1 do 8 dziennie. Dodatkowo bólowi towarzyszą bardzo charakterystyczne objawy autonomiczne. W każdym przypadku klasterowego bólu głowy badanie neurologiczne oraz badania neuroobrazowe powinny być prawidłowe. Każde odchylenie w badaniu przedmiotowym i laboratoryjnym sugeruje objawowe tło choroby. Przypadki bólu głowy przypominające ból klasterowy są stosunkowo często opisywane w piśmiennictwie.

W pracy przedstawiono przypadek starszej kobiety, u której rozpoznano klasteropodobny ból głowy związany prawdopodobnie z innymi chorobami. W trakcie przeprowadzania diagnostyki różnicowej rozważano atypowy klasterowy ból głowy i objawowy ból głowy spowodowany zapaleniem zatok obocznych nosa, naczyniakiem żylnym lub uciskiem naczynia na korzenie nerwu trójdzielnego.

Ten rodzaj bólu głowy nie jest bardzo rzadki w populacji ogólnej, a w każdym przypadku rozpoznania samoistnego bólu klasterowego, szczególnie o nietypowym przebiegu, należy wykluczyć objawową przyczynę dolegliwości.

Słowa kluczowe: klasterowy ból głowy, klasteropodobny ból głowy, objawowy ból głowy, wtórny ból głowy.

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Introduction

Headache with severe, strictly one-sided unilateral attacks of pain in orbital, supraorbital, or temporal localisation lasting 15-180 minutes and occurring from once every day to 8 times daily, typically with one or more autonomic symptoms (conjunctival injection, lacrimation, nasal congestion, rhinorrhea, facial and forehead sweating, miosis, ptosis, eyelid oedema), is recognized in the second edition of the International Classification of Headache Disorders (ICHD-II) [1] as cluster headache (CH). History, general, otolaryngological, ophthalmological and neurological examinations do not suggest any other disorders, and other causes of similar headache are excluded. Most patients (more than 80%) have the episodic form of cluster headache – the pain occurs in series lasting for weeks or months with periods of remission lasting weeks or (more often) months. A 20-40-year-old man is a typical patient with cluster headache. Any atypical course of headache and any atypical features of headache should suggest secondary cluster-like headache (CLH) recognized in ICHD-II in groups 5-12 [2]. A total of 156 case reports of this CLH have been published thus far [3].

Any pathological or morphological lesion in the central or peripheral nervous system should not be demonstrated in primary CH in neuroradiological investigations. Secondary (symptomatic) cluster headache (CLH) is rare compared to primary cluster headache.

We present a female patient with a history of headache meeting the ICHD-II criteria for ICHD-II who was diagnosed with secondary episodic CH due to vascular compression of the trigeminal nerve root. First, we differentiated between primary, but 'atypical' CH and secondary, symptomatic cluster headache due to frontal sinusitis, pontine venous angioma or vascular compression of the trigeminal nerve root.

Case report

A 69-year-old healthy woman, without any concomitant diseases, reported a very intense, severe headache located in the right fronto-temporal region with eyelid oedema, lacrimation, miosis and ptosis. The pain with additional symptoms lasted 2 hours in the early morning every two days during a week. She experienced a similar pain with similar symptoms for the first time about 5 years ago when she was 64, with similar duration and frequency of attacks. During the attacks she was restless and agitated. The family history was negative. General (including blood pressure and heart rate), neurological, otolaryngological and ophthalmological examinations were normal. The patient had no important medical history, but probably problems of sinusitis with rhinorrhea were recognised in the past. The brain magnetic resonance imaging (MRI) detected a subarachnoid cyst in the posterior fossa, bilateral white matter vascular lesions and pontine venous angioma. The MRI with T2-weighted 3D high resolution sequence and 3D TOF angiography revealed right-sided neurovascular conflict caused by the superior cerebellar artery compressing the right trigeminal nerve at its root entry zone (Fig. 1).

To date, a few studies have analysed single fibre electromyography (SFEMG) in cluster headache; mild abnormalities in jitter results revealed in CH patients could suggest subclinical impairment of neuromuscular transmission [4,5].

In our patient, SFEMG of the voluntarily activated left extensor digitorum communis muscle was conducted using Keypoint electromyography. Twenty pairs of potentials were recorded, and the mean consecutive difference (MCD) in jitter value was calculated for each pair. The results were compared with EMG laboratory reference values and considered abnormal when the mean MCD value exceeded 32 μs and two or more pairs had MCD values above 55 μs. The SFEMG did not reflect any abnormalities in neuromuscular transmission.

The conduction velocity of facial nerves, performed mandatorily before blink reflex examination, was examined and it was normal.

The blink reflex was elicited from electrical stimulation of the left and right supraorbital nerve with an early, ipsilateral R1 and a late, bilateral R2 response. Electrical stimulation of supraorbital nerves was performed with a peripheral nerve stimulator and silver/silver chloride disc surface electrodes. The latencies and amplitudes of all R1 and R2 responses were normal.

The X-ray examination of the sinuses showed frontal sinusitis. She was treated with antibiotics and sinus puncture with aspiration. After some weeks of sinusitis treatment, cluster-like headache disappeared completely for 1 year.

She was first diagnosed with secondary cluster headache as a manifestation of frontal sinusitis. One year later, however, the same pain in the same region with the same symptoms appeared and she complained of similar regularity but not so often. Attacks were less frequent (1-2 times a week) and less intense (she could continue her activity), accompanied by similar autonomic symp-

toms. This time, her X-ray of sinuses was completely normal. The final diagnosis was probable CLH because of neurovascular conflict.

Neurosurgery was considered, but because the first diagnosis was CLH due to frontal sinusitis, neurosurgery microvascular decompression of the nerve root was decided against and then, after final diagnosis, pharmacological treatment with verapamil (which is the first-line choice for prophylactic treatment of CH) in a daily dose of 240 mg was successful. She never used 100% oxygen inhalation or sumatriptan injection.

Discussion

The onset of CH usually takes place between the third and fifth decade of life but earlier [6] or very late onset of CH is reported [7]. Elderly onset is rare and generally symptomatic. Additional examinations performed in elderly patients suggest other disorders. The need for special caution for older groups is confirmed by reappraisal of symptomatic cases since the average age of onset in CLH patients turns out to be 42.7 years [3].

The reported symptomatic CH lacked two typical features: age and sex. The remission of the attacks with verapamil should indicate a primary form, but according to ICHD-II diagnostic criteria for CH, the primary form may be recognized if history and physical and neurological examination do not suggest any of the other disorders listed in ICHD-II: headache attributed to cranial vascular disorder (one-sided neurovascular conflict, pontine venous angioma), headache attributed to disorder of sinuses or other cranial structures (subarachnoid cyst in the posterior cranial fossa). Although the disappearance of headache after removal of the supposed cause is fundamental for the diagnosis of a secondary form, this is not applicable in our patient. No surgical treatment for neurovascular conflict was performed. The disappearance of pain after sinusitis treatment could well be spontaneous, representing just the end of a cluster period, as the pain reappeared one year later.

The results of contrast MRI with T2-weighted 3D high resolution sequence and 3D TOF angiography implies that some changes (compression or cyst) may produce symptoms of cluster headache. Because CH is presumed to activate the trigemino-vascular system, compression of the trigeminal nerve by a vessel, mainly an arterial vessel, may produce symptoms mimicking cluster-like headache. Although MRI studies reveal arterial contact with the sensory trigeminal nerve root in healthy subjects in up to 45%, in patients with symptomatic

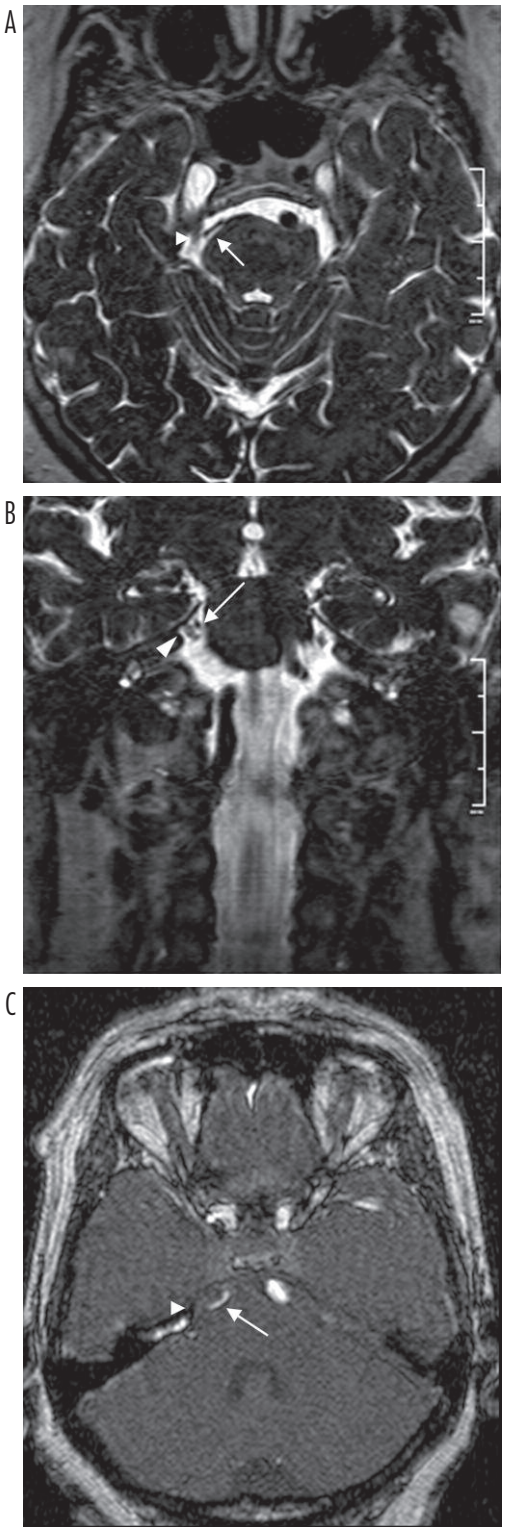

Fig. 1. Neurovascular conflict caused by the superior cerebellar artery. Axial (A), coronal (B) T2-weighted 3D high resolution sequence and axial 3D TOF angiography (C) demonstrate the superior cerebellar artery (white arrow) compressing the root entry zone of the right trigeminal nerve (arrowhead) at the medial site in the prepontine cistern

nerves (trigeminal neuralgia) arterial contact can be found in up to 85% [8]. In that situation, symptoms of every attack are strictly unilateral, but age and sex may be different from the typical course.

Sinus X-ray was performed due to rhinorrhoea and it seemed that the main problem of our patient was frontal sinusitis with CLH symptoms. The argument for the first diagnosis was resolution of attacks after successful treatment with antibiotics and normalization of sinus X-ray. So it was the primary diagnosis of a cause of cluster-like headache. CLH due to sinusitis may be connected with irritation of branches of the ophthalmic division of the trigeminal nerve and recurrent activation of a trigeminal autonomic reaction.

Relapse of CH without sinusitis symptoms after a one-year break is proof of another background for CH, so we diagnosed cluster-like headache as a result of neurovascular conflict because of compression by an artery located near the trigeminal nerve root in the prepontine cistern. We observed regression of symptoms during verapamil treatment (240 mg daily).

Although the pathophysiology of CH remains undetermined, it has a neuronal component with involvement of the trigeminal nerve. Compression of the trigeminal nerve root may be one explanation of CH pathophysiology, although in that situation all CH should be classified as a symptomatic headache.

Symptomatic CH should be suspected when the clinical features of the attack are atypical. Many published CLH cases present atypical manifestation with respect to ICHD-II criteria, but some of them (like our presented case) perfectly mimic CH [9,10]. Many case reports published recently introduced a form of cluster headache that appeared during some diseases [3]. The literature on CLH is dominated by reports associated with a mass lesion in relation to the posterior fossa. The aetiology varies widely from vascular (e.g. internal carotid aneurysms) or inflammatory (e.g. multiple sclerosis) to neoplastic (e.g. pituitary tumour) [3].

Structural diseases of sellar and parasellar structures [10] and internal carotid artery dissection [11] have been reported in the literature as ones that imitate the CH attack. Sinusitis has only rarely been reported as mimicking CH [12]. There is only one report, a very interesting paper by Scorticati *et al*. [13], about CLH associated with a foreign body in the maxillary sinus. More frequent causes of CLH are vascular changes: aneurysm, pseudoaneurysm [14,15], sinus thrombosis or cerebral venous thrombosis [15-17]. Another example of CLH due to pontine demyelination in multiple sclerosis was described by Leandri *et al*. [18]. Although pontine MRI in our patient shows venous angioma, it is probable that she had this malformation from her childhood and her CLH symptoms appeared in old age, when the superior cerebellar artery became more rigid and compressed the root entry zone.

The presented case may be another example of CLH related to trigeminal nerve root compression [19]. Masson *et al*. [20] described a patient with trigeminal neurinoma. We suggest that the diagnosis protocol for patients featuring CH symptoms should include cerebral contrast-enhanced MRI with T2-weighted 3D high-resolution sequence and 3D TOF angiography to identify the possible atypical elements that are part of CLH symptomatology if a secondary cause is suspected.

Cluster headache is related to alterations of the trigeminal autonomic pathways, but its aetiology is still unclear. The pathophysiology of CLH has been hypothesized to be an effect of activation of the trigeminal system giving rise to cluster attacks because of local compression of autonomic nerve fibres [21].

We also applied blink reflex to evaluate the brainstem and the sensory-motor part of the trigeminal-facial circuitry and we did not reveal any abnormalities in the patient. According to the literature, patients with idiopathic trigeminal neuralgia showed normal parameters of blink reflex while patients with symptomatic trigeminal neuralgia showed prolonged latencies of R2, R2' when stimulating the afflicted side [22]. Raudino examined the electrically elicited blink reflex during a symptomatic period in patients with CH. In nearly all cases (11/12) the amplitude of the contralateral R2 response on the symptomatic side was significantly lower [23]. In our patient, blink reflex did not reveal any differences between the symptomatic and asymptomatic side.

The described pain has the same characteristics as the majority of cases published recently and fulfils the criteria proposed for classification.

This type of headache is quite rare, although headaches mimicking CH attributed to other disorders are not rare in the general population. It is worth performing neuroimaging with contrast and angiography to avoid misdiagnosis and to exclude (or confirm) secondary causes of CH. To sum up, we would like to state that the pathogenetic link between CH and other conditions in the presented case is not clear.

Disclosure

Authors report no conflict of interest.

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