Chiari Malformation Type I Presenting as Cluster-like Headache

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Abstract

Purpose: Although different types of headache have been described in Chiari malformation type I, reports of cluster-like headaches are rare.

Case Report: We report a 26-year-old man who presented with a two-week history of excruciating headache in the right temporal region after coughing, which was accompanied by autonomic features including right-sided nasal congestion and tearing from his right eye. Sensory deficits in the first branch of the right trigeminal nerve and along C3-5 dermatomes were noted, and brain magnetic resonance imaging was compatible with a diagnosis of Chiari malformation type I.

Conclusion: A diagnosis of secondary cluster headache was made due to a lack of typical periodicity, and the presence of anhidrosis and sensory abnormalities and cough headache.

Key Words: Chiari malformation, cluster headache, trigeminal autonomic cephalalgia

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INTRODUCTION

Chiari malformation type I (CMI) is characterized by caudal displacement of the cerebellar tonsils through the foramen magnum. It is usually associated with syringomyelia, obstructive hydrocephalus, and bony anomalies. Of the various symptoms of CMI, headache is the major presenting symptom in 26-50% of patients[1]. The distinctive headache in CMI is precipitated by coughing, sneezing, or abdominal straining, and occurs in the occipital-suboccipital region. However, many other types of headache patterns have been reported in CMI, including migraine, tension-type headache, headache resembling spontaneous intracranial hypotension, and even laugh headache[2-4]. Cluster headache, a strictly unilateral and extremely severe headache with autonomic features and circadian periodicity, rarely occurs in patients with CMI.

CASE REPORT

A 26-year-old man suffered from sudden excruciating pain in his right forehead after coughing two weeks before admission. The pain was so severe that he was unable to fall asleep or work, even after taking painkillers. The headache gradually shifted to the right temporal area over the following two days and sometimes involved the right suboccipital region. The pain became intense and was

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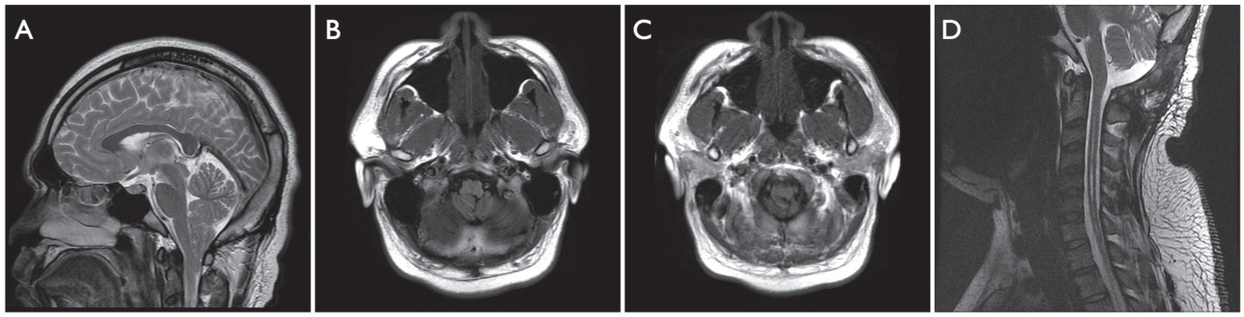

Figure 1. Magnetic resonance imaging of the brain revealed a 25-mm caudal descent of the cerebellar tonsils (A, T2-weighted image, sagittal view) with compression mainly to the right posterolateral aspect of the lower brain stem and upper cervical spinal cord (B, C, T1-weighted images, axial view). Postoperative magnetic resonance imaging showed release of the compression (D, T2-weighted image, sagittal view).

easily precipitated by coughing and physical exertion. Each attack of the headache lasted for about two to three hours and it occurred approximately three to four times a day without regular periodicity. The pain was always accompanied by congestion in the right side of his nose and tearing from his right eye. Persistent anhidrosis of his right face and trunk developed one week later. Moreover, persistent numbness in his right forehead, postauricular area, right arm and chest ensued. The headache was not related to changing posture or turning his head.

On neurological examination, the pinprick test demonstrated sensory deficits in the first branch of the right trigeminal nerve and along C3-5 dermatomes. No eyelid edema, miosis, ptosis, involvement of other cranial nerves, or cerebellar signs were noted. His neck was supple and there were no obvious tender points. Computed tomography angiography did not show an intracranial aneurysm. Magnetic resonance imaging of the brain showed a 25-mm caudal descent of the cerebellar tonsils with compression to the right posterolateral aspect of the cervical spinal cord (Fig. 1A-C) and syringomyelia (Fig. 1D). One day before the scheduled operation to decompress the craniocervical junction, his right limb movements became clumsy. His headache completely resolved a few days after the decompression surgery, however, his motor, sensory, and autonomic symptoms remained.

DISCUSSION

This young patient's recurrent fixed-side intense headache with prominent autonomic features fulfilled the International Headache Society criteria for cluster headache[3]. However, after we considered his symptoms, the lack of periodic attacks, anhidrosis instead of facial sweating, and sensory deficits led us to make a diagnosis of secondary cluster headache. The differential diagnosis of secondary cluster headache includes posterior fossa tumors, cavernous sinus lesions, internal carotid artery dissection, and sellar region tumors[5]. Furthermore, the coexistence with cough headache also hinted at a diagnosis of secondary headache. A previous study showed that six of 83 patients with cough headache had lesions in the posterior fossa, including two patients with CMI[6].

To the best of our knowledge, this is only the second case of CMI presenting with cluster-like headache in the literature[7]. Unlike our patient, the headache in the other reported case was associated with head posture. The headache also lacked periodicity, and the authors postulated that the symptoms were caused by an extremely large cervical syrinx. The symptoms were relieved after foramen decompression surgery. Our case had a greater descent (25 mm) of the cerebellar tonsils than that usually seen in CMI patients. Aboulezz et al. reported 13 patients with Chiari malformation, in whom the average distance of the tonsillar tips below the foramen magnum was 10.3 ± 4.6 mm[8]. The headache with hemi-anhidrosis and ipsilateral sensory deficits in our patient could be explained by unilateral compression to the trigeminal nucleus caudalis and upper cervical cord by the herniated cerebellar tonsils. Various extra-axial cervical lesions such as trauma and

upper cervical meningiomas have been reported to present as cluster-like headache[9]. The regularity of attacks in cluster headache is usually attributed to dysfunction of the hypothalamus[10], and since the hypothalamus is not involved in CMI, characteristic periodic attacks are absent.

Though the pathophysiology of cough headache in CMI remains unclear, usually it is attributable to be the transient pressure dissociation between the intracranial and the intraspinal compartments[1]. According to Pascual et al's study, the degree of tonisllar descent correlated with the presence of cough headache[11]. Greater extent of tonsiller descent with exquisite cervical cord compression probably also explains why our patient is so unique. Despite the resolution of his headache after the operation, our patient's motor, sensory, and autonomic symptoms remained. This result is consistent with a previous study in which CMI patients with syringomyelia exhibited less symptomatic improvement after decompression surgery[12].

In conclusion, our case offers evidence that CMI can cause cluster-like headache. Craniocervical imaging studies are warranted for patients with atypical symptoms of cluster headache.

REFERENCES

1. Taylor FR, Larkins MV. Headache and Chiari I malformation: clinical presentation, diagnosis, and controversies in management. Curr Pain Headache Rep 2002;6:331-337.
2. Grazzi L, Andrasik F. Headaches and Arnold-Chiari syndrome: when to suspect and how to investigate. Curr Pain Headache Rep 2012;16:350-353.
3. Headache Classification Subcommittee of the International Headache Society (IHS). The international classification of headache disorders, 2nd edn. Cephalalgia 2004;24(suppl 1):1-160.
4. Boga T, Haas L, Hunn M, Hamilton D. Laugh headaches: a rare form of headache associated with type 1 Arnold-Chiari malformation. Intern Med 2011;41:707-708.
5. Cittadini E, Matharu MS. Symptomatic trigeminal autonomic cephalalgias. Neurologist 2009;15:305-312.
6. Chen PK, Fuh JL, Wang SJ. Cough headache: a study of 83 consecutive patients. Cephalalgia 2009;29:1079-1085.
7. Seijo-Martinez M, Castro del Río M, Conde C, Brasa J, Vila O. Cluster-like headache: association with cervical syringomyelia and Arnold-Chiari malformation. Cephalalgia 2004;24:140-142.
8. Aboulezz AO, Sartor K, Geyer CA, Gado MH. Position of cerebellar tonsils in the normal population and in patients with Chiari malformation: a quantitative approach with MR imaging. J Comput Assist Tomogr 1985;9:1033-1036.
9. Kuritztky A. Cluster headache-like pain caused by an upper cervical meningioma. Cephalalgia 1984;4:185-186.
10. Massimo L, Gennaro B. Pathophysiology of trigeminal autonomic cephalalgias. Lancet Neurol 2009;8:755-764.
11. Pascual J, Oterino A, Berciano J. Headache in type I Chiari malformation. Neurology 1992;42:1519–1521.
12. Kalb S, Perez-Orribo L, Mahan M, Theodore N, Nakaji P, Bristol RE. Evaluation of operative procedures for symptomatic outcome after decompression surgery for Chiari type I malformation. J Clin Neurosci 2012;19:1268-1272.