Cluster-Like Headache Associated with Symptomatic Chiari Type 1 Malformation

Semptomatik Chiari Malformasyonu Tip 1 ile İlişkili Küme Baş Ağrısı

Yüksel KAPLAN, Özden KAMIŞLI

İnönü University Faculty of Medicine, Department of Neurology, Malatya, Turkey

Dear Editor,

Cluster headache (CH) is considered as a primary headache syndrome, thus neuroradiological investigations of the central nervous system should be normal in primary CH patients (1). However, symptomatic cluster-like headaches (CLH) have been reported in association with various disorders such as aneurysms, arteriovenous malformations, cervical trauma, medullary infarcts, inflammatory/infectious diseases, and tumors (2). CLH associated with Chiari type 1 Malformation (CM-1) has been described in only one case report in the relevant literature (3).

Here we describe a case diagnosed with CLH associated with symptomatic CM-1. A 28-year-old female patient had admitted with recurrent headaches since the age of 20 years. She had severe, recurrent, pain in the right frontal and periorbital areas and headache, lasting about 20-90 minutes. She also suffered from ipsilateral tearing, rhinorrhea and ptosis during headaches. These pain attacks occurred irregularly two to four times per day. Although this attack usually appeared spontaneously, it was precipitated by head or neck movements. Additionally, she also suffered from occasional pain in the occipital-suboccipital area, vertigo, blackout in the eyes, palpitation, sweating; all of which disappearing in 5-10 minutes. Neurological examination revealed vertical and down-beat nystagmus, positive Romberg's test, and hyperreflexia. Her cardiologic tests were normal. Cranial and cervical MRIs disclosed CM-1 (Figure 1).

She responded well to methyl-prednisolone 60 mg/day (10 days), which was tapered in a month period.

We believe that CM-1 could be interpreted as symptomatic in this patient. While CM-1 can be asymptomatic, clinical

manifestations, which typically begin in young adulthood, can include headaches, visual disturbances, neuro otological complaints, lower cranial nerve dysfunction, and sleep apnea (4). Among the manifestations of CM-1, headache is one of the most common symptoms, occurring in 15-98% of the patients. The known headache spectrum in CM-1 includes cough headaches, exertional headaches, low cerebrospinal fluid pressure headaches, long-lasting headache attacks, suboccipital headaches, and migraine attacks (4).

While CM-1 and CH seemed as unrelated diseases, determining the actual cause of CH is obviously difficult when these two pathological processes co-occur. It raises the question whether the pathogenesis of these diseases may be linked. With the current precision of central nervous system imaging, CM-1 can be an incidental finding when MRI is conducted for other reasons. In CM-1, variable caudal displacement of cerebellar

Figure 1. Magnetic resonance image of axial T2 sequence of cranial (a) and sagittal T1 sequence of cervical spinal cord (b) showing Chiari type 1 malformation

Correspondence Address/Yazışma Adresi

Yüksel Kaplan MD, İnönü University Faculty of Medicine, Department of Neurology, Malatya, Turkey

Gsm: +90 532 468 71 69 E-mail: yukselkaplan@yahoo.com Received/ Geliş tarihi: 23.7.2013 Accepted/Kabul tarihi: 25.7.2013

© Archives of Neuropsychiatry, published by Galenos Publishing. / © Nöropsikiyatri Arşivi Dergisi, Galenos Yayınevi tarafından basılmıştır.

tonsils occurs into the upper cervical canal. These structural abnormalities in CM-1 may include stretching of cranial nerves or direct compression of brain stem nuclei, compression of the posterolateral part of the medulla and the upper cervical spinal cord, vascular distortion in the territories irrigated by the vertebral and posterior inferior cerebellar arteries, and pressure on the rootlets of C1, C2, and the vagus nerves (4,5). Although the pathophysiology of CH remains undetermined, it has a neuronal component with involvement of the trigeminal nerve (1). The pain of CH may be associated with dysfunction in an area of the brain stem and/or craniospinal pressure dissociation, stimulating pain-sensitive structures in patients with CM-1. Therefore we believe that CM-1 may be a factor associated with symptomatic cluster headache.

References

1. Domitrz I, Gawel M, Maj E. Cluster headache a symptom of different problems or a primary form?. A case report. *Neurol Neurochir Pol* 2013; 47: 184-188.
2. Mainardi F, Trucco M, Maggioni F, Palestini C, Dainese F, Zanchin G. Cluster-like headache. A comprehensive reappraisal. *Cephalalgia* 2010; 30: 399-412.
3. 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.
4. Kaplan Y, Oksuz E. Chronic migraine associated with the Chiari type 1 malformation. *Clin Neurol Neurosurg* 2008; 110: 818-822.
5. Khurana RK. Headache spectrum in Arnold-Chiari malformation. *Headache* 1991; 31:151-155.