PEER REVIEWED LETTER

Chronic Cluster-Like Headache Secondary to an Epidermoid Clival Lesion

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Cluster headache is recognized by the International Headache Society as a primary headache disorder, consisting of seasonal and daily attacks of severe unilateral periorbital pain accompanied by autonomic symptoms.[1] As with other primary headache disorders, intracranial lesions can rarely mimic cluster headaches, in particular if it presents in an atypical fashion. We report a patient with chronic cluster-like headache secondary to a clival based epidermoid cyst. This is the first such association reported in the literature.

A 39-year-old man with no significant past medical history and no current medications was referred with a three-year history of strictly right-sided headaches. Two to three times daily, he suffered attacks of excruciating pressure-like pain in the periorbital and frontal area on the right. These lasted between 10 and 90 minutes and occasionally woke him from sleep. They were always accompanied by numbness of the upper part of his face, increased lacrimation, rhinorrhea and ptosis on that side. Occasionally, he had nausea and vomiting as well. Since the onset of symptoms, attacks occurred almost daily and never remitted for more than four weeks. He tried methysergide, gabapentin, carbamazepine, lithium carbonate and corticosteroids without success. Later, his attacks have occasionally involved the left side.

Physical and neurological examinations were unremarkable except for clearly decreased sensation to pinprick over the first and second divisions of the right trigeminal nerve. Although he had preserved corneal reflexes, his right nasal tickle reflex was absent.

Computer tomography and magnetic resonance imaging showed a clival based lesion, which appeared spontaneously hyperintense on T1- and on T2-weighted sequences and extended slightly posteriorly and to the right (Figure 1). It was causing mild mass effect and narrowing of the right internal carotid artery in the cavernous sinus and seemed to be lifting the trigeminal nerve in Meckel's cave (Figure 2). Compared to imaging a year earlier, no significant change was noted. Differential diagnosis included chordoma, chondroid chordoma or chondrosarcoma.

He had a suboccipital craniotomy with a far lateral extension to decompress and resect this cystic lesion completely. There were no complications. Histopathological analysis established a diagnosis of epidermoid cyst. One month after his surgery, he reported complete resolution of his chronic cluster headaches.

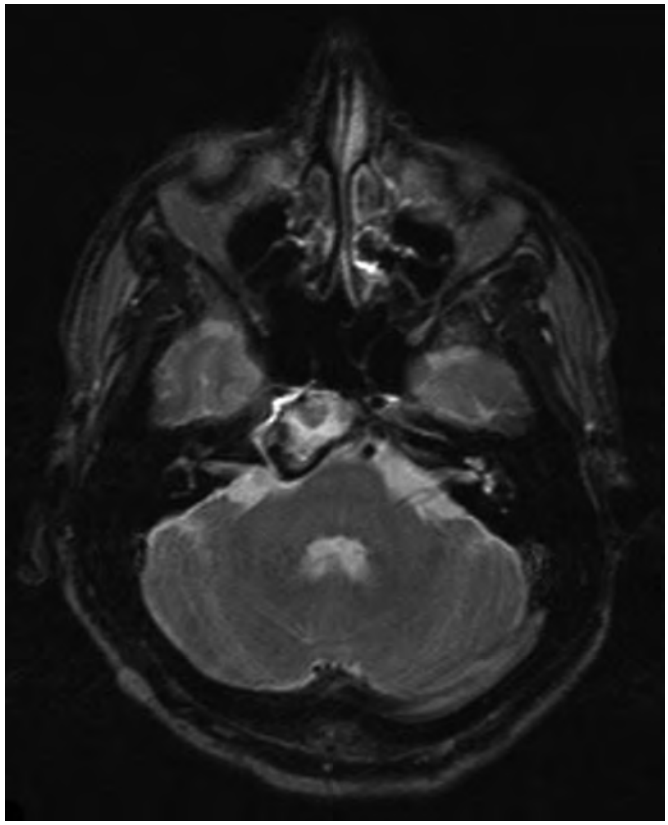

Figure 1: *Axial T2-weighted magnetic resonance imaging demonstrating the right clival-based lesion causing mild compression of the pons.*

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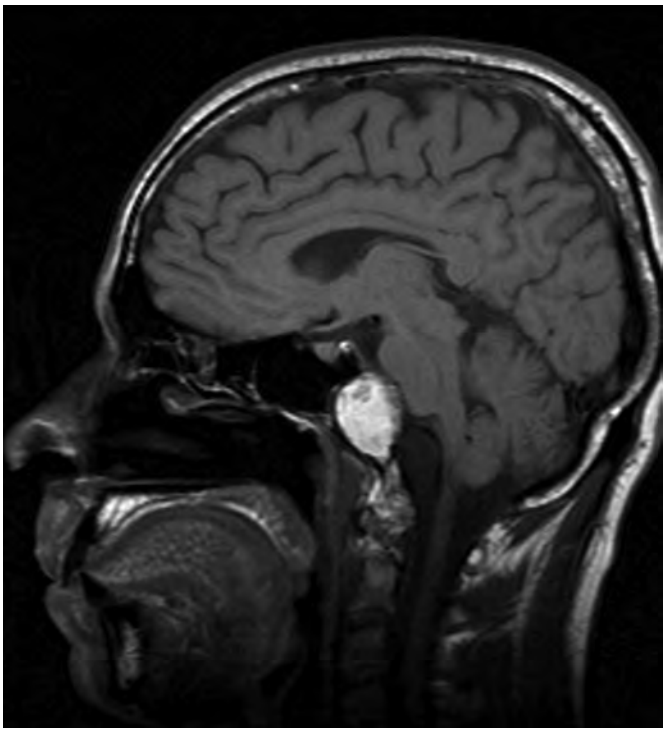

Figure 2: Sagittal T1-weighted magnetic resonance imaging demonstrating the spontaneously hyperintense clival-based lesion.

Discussion

In the last ten years, advances have been made in the understanding of the pathophysiology of primary cluster headaches. Two systems seem to be involved.

It had long been postulated that a local inflammatory reaction at the level of the cavernous sinus was the likely cause of the headaches,[2] based on vascular changes ipsilateral to the pain. Studies have subsequently shown that these changes are likely only secondary and not causative. Given the pattern of pain in the trigeminal distribution and the occurence of autonomic ipsilateral symptoms, it is now believed that the vascular changes are secondary to activation of the trigeminovascular system and the trigeminoautonomic (parasympathetic) reflex[3] through the superior salivatory nucleus.

A second important aspect in this disorder seems to involve the inferior posterior hypothalamus. This is further supported by the neuroendocrine changes noted in cluster headache patients as well as the surprising chronobiological rythmicity of the disorder, implying involvement of the biological circadian clock.[4] Positron emission tomographic scans showed marked activation of the ipsilateral ventral hypothalamic grey matter in patients with cluster headaches and other autonomic cephalalgias but not in patients with migraine headaches.[5] More impressively, a patient's symptoms improved after stimulation of this hypothalamic area.[6]

However, even though connections exist between these two systems, it is still unclear how the two interact to give rise to the cluster headache pain.

The precise pathophysiology of secondary cluster headaches is even less clear. The first descriptions of symptomatic cluster headache date back to 1949 when eight patients with whiplash injuries subsequently developed such attacks.[7] Several reports of different lesions have followed including inflammatory, vascular and neoplastic causes.[8] Most lesions involve the trigeminal nerve or its subdivisions at any point during their course but no lesions of the hypothalamus itself have been described. Some of the vascular lesions have been considered to exert their effect by compressing the rich sympathetic nerve fibers coursing along them.

In this patient, the lesion clearly seemed to be impinging on the trigeminal ganglion itself, in Meckel's cave. One postulates that the local pressure effect of the mass caused activation of the trigeminovascular pathway leading to the cluster headache symptoms. An unusual feature is the location of the lesion, arising not from the Sella Turcica or the cavernous sinus as in most cases described so far, but more inferiorly and posteriorly at the clivus. An epidermoid cyst has never been described to give rise to cluster headaches.

Finally, it is important to stress that our patient had several atypical features compared to those with idiopathic cluster headache. First, on history, the patient had associated nausea and vomiting on occasion, which are usually not seen with cluster headache. Some of his headaches lasted less than 15 minutes and, in the first years, they were side-locked to the right. The lack of responsiveness to currently available medications over a three-year period should also prompt one to look for other causes for his headache. On physical examination, the decreased sensation to pinprick over the first and second divisions of the right trigeminal nerve and the absent nasal tickle on the right only, are not consistent with a primary headache disorder but rather point to a lesion. Atypical features on history or physical examination should lead the clinician to be wary of diagnosing a primary headache disorder and to consider further imaging the patient.

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