J Headache Pain (2007) 8:60–62
DOI 10.1007/s10194-007-0355-6

BRIEF REPORT

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Atypical primary headache responding to finger pressure: possible involvement of the vagus nerve?

Received: 15 September 2006
Accepted in revised form: 3 October 2006
Published online: 15 January 2007

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Abstract We describe a case of atypical primary headache strongly responsive to prolonged pressure in the anterior aspect of the neck. We hypothesize that, at least in this case, the trigemino-cervical system and its connections with the vagus nerve are involved.

Keywords Atypical primary headache • Trigeminocervical complex • Topiramate • Vagus nerve

Introduction

Cluster headache and chronic paroxysmal hemicrania (CPH) are characterised by multiple attacks of severe, strictly unilateral periorbital pain [1]. Nevertheless, patients who have features typical of one primary headache disorder and an additional feature typical of a second disorder have been identified. Examples of this include bilateral cluster headache, cluster headache with aura and cyclic migraine, cluster-migraine and cluster-tic disorder.

Headache patients may fail to exhibit a diagnostically important feature of a primary headache disorder yet in all other ways fulfil criteria for that disorder, for example migrainous disorder and aura without headache.

Finally, one disorder may be completely subsumed within another: idiopathic stabbing headache may occur independently but it is considered as a secondary manifestation of migraine when it is associated with that disorder [2].

The whole spectrum of atypical primary headaches was first described by Young [2].

Case report

GB, a 55-year-old female, came to our observation at the Headache Unit of our Pain Center, in March 2005. She had been suffering from headache at the age of 43. For ten years she experienced daily headache attacks in periods of four consecutive months, approximately once per year. In the last year the headache had lost its yearly periodicity, and at the time of observation GB had been complaining of daily headaches for 15 months. Each attack lasted 60–120 min and occurred in the afternoon or evening, with an average frequency of four attacks per day.

The pain was severe and squeezing in nature, associated with a right-eye ptosis, bilateral lacrimation and rhinorrhoea. It was initially located in the right mandibular region, subsequently spreading to right eye, right anterior region of her neck and the right nuchal region. Later on the headache became bilateral and symmetric. The pain was associated with nausea, vomiting and phonophotophobia. During the attacks, the patient was restless, rocking her head and body while standing or sitting in a dark, quiet room.

The headache was triggered by lack of sleep and alcohol consumption.

Sumatriptan (subcutaneous and tablets 100 mg) did not provide relief; NSAIDs were somewhat effective in relieving her pain. GB reported that the most effective pain control mechanism prior to referral was to apply digital pressure in the right anterior region of the neck. The finger pressure is continuously and strongly applied during all the attacks. This became a habit, which eventually resulted in a cutaneous lesion in the stimulated area (Fig. 1). The picture shows an indurate inflammatory plaque (approximate diameter 30 mm), with a central reactive hyperkeratosis and irregular borders.

General and neurological examinations were normal. MR imaging of the brain as well as extensive study of vascular and craniomandibular systems and soft tissues of the head and neck (MR angiography, TMJ X-rays and echography) did not show any abnormalities.

A trial of indomethacin at a dose of 75 mg was followed by a reduction in pain intensity from 7 to 5 on the Numerical Rating Scale 0–10 (NRS; 0=no pain, 10=the worst pain imaginable) and a reduction of frequency of the attacks from 30 days/month to 20 days/month. Because of the failure of previous treatments (verapamil, valproate, atenolol, amitriptyline), after one month of clinical observation we started topiramate monotherapy 75 mg/day. In the following three months GB experienced a further strong reduction in pain intensity (daily NRS: 1–2/10) with the frequency of the headache attacks dropping to 1 per month. The attacks were still responsive to digital pressure of the neck. At 1-year follow-up visit GB is clinically stable.

Fig. 1 Cutaneous lesion of the stimulated area in the anterior aspect of the neck due to prolonged finger pressure

Discussion

This patient satisfied some of the International Headache Society criteria [1] for chronic migraine and chronic cluster headache. The striking temporal profile of episodic short-duration headaches with the active phase lasting four months, separated by approximately one year, the presence of autonomic symptoms as well as the headache frequency and circadian periodicity, are typical of cluster headache. The presence of positive familial history, phonophotophobia, nausea and vomiting is more typical for the migraine diagnosis.

In the pathophysiology of all primary headaches the brainstem trigeminocervical complex (TCC) seems to play an important role. The TCC is a nociceptive structure that exerts fundamental control over inputs from cervical and trigeminal nociceptors [3].

Continuous vagal nerve stimulation (VNS) for 24 h in rats produced significant antinociceptive effects in a model of trigeminal pain [4]. The same efficacy was observed in humans affected by chronic refractory migraine and cluster headache [5]. VNS appears to inhibit activation of the TCC pain signalling sensory neurons and decrease pain-related behaviour in rats. Vagal afferent

stimulation predominantly inhibits sensory processing in the TNC [6] and in the ventral posteromedial thalamic nucleus [7].

In humans, on the cervical level the majority of sensory vagus-nerve fibres are unmyelinated C-fibres that convey sensory inputs from larynx, pharynx and visceral organs. Evidence has recently been provided that stimulation of these vagal fibres is able to decrease the activity of peripheral and spinal nociceptive pathways via central and peripheral mechanisms [8]. We hypothesise that the continuous pulsed finger pressure self-applied to the anterior neck region could have provoked a mechanical stimulation of the parasympathetic visceral fibres (e.g., vagus nerve), with a comparable analgesic effect to that reported in the above-mentioned observations and experimental animal and human models of trigeminal pain [3, 5, 8].

Akerman and Goadsby demonstrate that topiramate is capable of inhibiting neurogenic dural vasodilatation in an animal model [9]. The results complement an electrophysiological study that demonstrates inhibition of trigeminocervical neurons activated by a nociceptive stimulus at comparable doses. It is feasible that trigeminovascular inhibition plays a role in the antimigraine effect of topiramate, although it is unlikely to be the full explanation for its effects. The effect of topiramate on TCC could play a major role in its antimigrainous activity [10].

In summary we described an atypical primary headache (cluster-migraine?) [2] responsive to anterior finger pressure and to topiramate. A possible link between these therapeutic approaches may be represented by the TCC and its role in the primary headache pathogenesis.

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