Cluster headache (CH) is characterised by attacks of severe unilateral pain in the orbital, supraorbital and/or temporal areas that last from 15 to 180 min, with recurrence up to 8 times daily and accompanied by ipsilateral autonomic symptoms. Although effective acute treatments are available for CH attacks (e.g. subcutaneous sumatriptan injections), most patients also require preventive therapy. Several drugs, such as verapamil, methysergide and lithium carbonate, have proved to effectively prevent CH attacks and shorten bouts. Oral steroids are considered to provide the most effective transitional preventive treatment, though they may provide limited relief in some cases; moreover, some patients become steroid-dependent and develop serious steroid-related adverse effects within months. CH is marked by its circadian rhythm. Episodic cluster periods start at the same time each year, occur at the same time each day and the duration of each CH is almost the same for every attack. These clinical features, along with the hormonal alterations documented in CH patients, suggest that the hypothalamus plays a role in the genesis of CH. PET studies by May et al., revealed hypothalamic activation during CH attacks, supporting the hypothesis of hypothalamic involvement. The concept of the hypothalamus acting as a CH generator has also been entertained. However, not all CH patients present the same symptoms, nor do all respond to the same medications, which suggests that atypical or even non-hypothalamic forms of CH may exist. Although the cervico-occipital onset of CH is not contemplated by the International Headache Diagnostic Criteria II-version (IHDC - II), it is not uncommon to find patients with this painful symptomatology in clinical practice. Anatomical and clinical data suggest that the greater occipital nerve (GON) may trigger pain that has the typical cluster characteristics and is associated with the autonomic symptoms noted in CH. Sensory neurons in the trigemino-cervical complex receive ipsilateral and contralateral input from the GON.

We describe an atypical cluster headache with trigeminal symptoms that improve after the blocked of the greater occipital nerve in one patient with occipital neuroma.

Case

A 37-year-old woman came to the Headache Unit in our Pain Center in December 2005. She had a positive past medical history for migraine without aura. In March 2005, two weeks after a neck soft tissue trauma, she started experiencing daily headache attacks without periodicity. Each attack lasted from 30 to 120 min and occurred in the afternoons or evenings, with an average frequency of four attacks per day. The pain, which was always unilateral (right-sided), invariably started in the right occipital region and subsequently spread to the right eye and frontal region (Figure). The pain was severe and squeezing in nature, and was associated with right-eye ptosis, unilateral right-sided lacrimation and rhinorrhea (Figure). The pain was not associated with nausea, vomiting or photophobia. During the attacks the patient was restless, rocking her head and body while standing or sitting.

Oxygen and sumatriptan (subcutaneous 6 mg) were inconsistently effective, while NSAIDs did not provide any pain relief. The patient reported that the most effective pain control mechanism prior to referral to our Headache Unit had been the application of digital pressure to the right occipital region of the neck during the attacks. Preventive monotherapies (verapamil, valproate and steroids) previously prescribed by other physicians had been ineffective.

Both the general and neurological examinations were normal, apart from the presence of a subcutaneous nodular lesion in the right occipital region of the neck in the area in which the patient applied digital pressure to relieve pain. During the clinical examination, digital pressure in this area evoked a shock-like sensation in the right occipital and parietal region. Neither MRI of the brain and the cervical spine nor an extensive study of the

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**Neuroma Occipital Desencaadeando Cefaléia em Salvas Responsiva ao Bloqueio do Nervo Occipital Maior**

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Greater occipital nerve blockade

Di Stani et al.

vascular and craniomandibular systems (MR-angiography, temporomandibular joint X-rays) revealed any abnormalities. Ultrasonography of the neck demonstrated a round echogenic mass (approximately 5 mm in diameter) where the presumed neuroma was believed to be located.

After informed consent, we performed a GON blockade by injecting lidocaine 2% (5 ml) and betamethasone (4 mg) in the right occipital region (ipsilaterally to the CH), over the neuroma site; the GON blockade was defined as the appearance of hypo-anaesthesia in the GON area after the procedure. The clinical response was good, the patient becoming pain-free within minutes. Preventive therapy with gabapentin was thus started, at a dose that was gradually raised to 1800 mg daily.

The patient did not display any side effects after the injection. At the one-month follow-up visit, she had not any attacks since the GON blockade, and at the six-month follow-up visit the patient was still pain-free.

DISCUSSION

Our patient satisfied some of IHDC-II criteria for episodic cluster headache. However, the lack of rhythmicity of the headaches, the occipital onset of pain during the attacks, and the ability to sit still during a headache are all qualities that are uncharacteristic of “hypothalamic-influenced CH”. Our patient describe did have a CH, but with somewhat uncommon features.

The fact that the CHs in this patient were relieved by digital pressure on the presumed right-occipital neuroma raises the question of whether the GON can play a role in the pathogenesis of CH. We recently showed that digital pressure over the greater occipital nerve improve the pain of the migraine patient, probably by the diffuse nociceptive inhibitory control mechanisms.

Anatomical and clinical data suggest that the GON can produce pain that has the typical cluster characteristics and is associated with the autonomic symptoms noted in CH. The sensory neurons in the trigeminocervical complex receive ipsilateral and contralateral input from the GON. Electrophysiological studies on humans suggest that there is a convergence of dural and cervical afferents in the GON, which then converge on the trigeminocervical complex. GON stimulation has been shown to cause frontal head pain in humans.

Piovesan et al. described a patient who, following GON stimulation, developed not only pain in the head area innervated by the ophthalmic division of the trigeminal nerve, but also ipsilateral conjunctival injection and lacrimation. Short-lasting GON stimulation thus appears to be able to produce a cluster-like headache. How the presumed right-occipital neuroma stimulated or activated the GON in the patient we describe can merely be hypothesised.

Neuromas are a significant source of spontaneous ectopic activity in injured primary afferents. These ectopic discharges are characterized by irregular firing patterns and repetitive spikes. The proportion of spontaneously active nerve fibers usually peaks within the first 3 weeks and decreases substantially thereafter. Discharges originating from Aβ and Aδ-fibers are prominent during the first 2 weeks after injury and then subside, whereas spontaneous activity from C-fiber endings lasts longer. Since the hypothalamus in this patient was activated secondarily via the trigeminocervical-hypothalamic pathway following GON stimulation, we hypothesize that a connection between the GON and the hypothalamus could exist.

As previously described by Ambrosini et al., we observed that the GON blockade alone effectively terminated the cluster symptomatology, which in our patient consisted of several attacks per day. We cannot exclude the possibility that the end of the CH attacks merely coincided with the time of the GON-blocking injection, though it is highly unlikely as the frequency of the attacks was increasing and the CH symptoms ended within minutes of the injection. As reported by Roze, the blockade procedure itself (needle insertion) may provoke CH.
symptomatology; in the patient we describe, the GON blockade was instead extremely effective in relieving the CH pain as described in previous studies.\textsuperscript{16,17} Peres et al. after greater occipital nerve blocked in cluster patients reported 28.5% good response, 35.7% a moderate and 35.7% of the cases no response over the pain control.\textsuperscript{17} The presence of both excitatory and inhibitory trigeminocervical convergence mechanisms may provide a clinical explanation in our patient.\textsuperscript{11}

The characteristics of this headache occurs for the first time in close temporal relation to another disorder that is a known cause of headache, it is coded according to the causative disorder as a secondary atypical cluster headache. The resolution of the headache after GON blockade it is very good evidence that the neuroma can cause the CH, suggesting a secondary cause.\textsuperscript{1}

Another possible diagnostic was cervicogenic headache. As we know the major criteria of cervicogenic headache including: (A) pain, referred from a source in the neck and perceived in one or more regions of the head and/or face, fulfilling criteria C and D. Our case related this clinical distribution. (B) clinical, laboratory and/or imaging evidence of a disorder or lesion within the cervical spine or soft tissues of the neck known to be, or generally accepted as, a valid cause of headache. This relationship occurred in our case, neuroma induced the pain symptoms. (C) evidence that the pain can be attributed to the neck disorder or lesion based on at least one of the following (C1, demonstration of clinical signs that implicate a source of pain in the neck), (C2, abolition of headache following diagnostic blockade of a cervical stricture or its nerve supply using placebo-or other adequate controls). (D) Pain resolves within three months after successful treatment of the causative disorder or lesion. Our case fulfill the IHDC-II criteria for cervicogenic headache.

However if we utilized the Sjaastad criteria some differences occurs: (A) precipitation of head pain, similar to the usually occurring after neck movement and/or sustained awkward head positioning, and/or after external pressure over the upper cervical or occipital region on the symptomatic side. In our case the symptoms improve after digital compression over the occipital region, and the movement or sustained head position did not produce headache; (B) restriction of the range of motion in the neck. In our case the patient did not showed it; (C) ipsilateral neck, shoulder, or arm pain of a rather vague nonradicular nature or, occasionally, arm pain of a radicular nature.\textsuperscript{18} Our case showed the ipsilateral pain, although the pain was limited to the head; (D) confirmatory evidence by diagnostic anesthetic blockades; (E) unilaterality of the head pain, without sideshift (D and E criteria occur in our patient).\textsuperscript{10} The patient describe did not fulfill the major criteria (Sjaastad criteria) for cervicogenic headache.

In conclusion, we describe an atypical CH triggered by a presumed occipital neuroma that responded to a GON blockade. We strongly recommend the use of a GON blockade as transitional therapy for CH, particularly in cases in which steroid therapy fails. The patient also had criteria for cervicogenic headache (IHDC-II) but did not major cervicogenic criteria (Sjaastad criteria).

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