CLINICAL CORRESPONDENCE

Persistent idiopathic facial pain responsive to topiramate

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We present a patient who at initial visit appeared to have hemicrania continua (HC). Due to the absence of response to indomethacin, the traditional treatment for HC, and the atypical pain characteristics that did not meet all the migraine criteria, the patient was diagnosed with atypical idiopathic facial pain. As antiepileptic medications have shown efficacy in neuropathic pain and facial neuralgias (1–4), the patient was put on topiramate, which has been described as an effective treatment for primary headaches (5, 6). After 20 months of follow-up, topiramate continues to demonstrate effectiveness in the treatment of his idiopathic facial pain.

Case report

A 58-year-old white male physical education teacher presented with a rapidly progressive headache that started at age 49 years. The headache was characterized by continuous, unremitting stabbing pain, of moderate to severe intensity, in the left eye, as well as mild to moderate stabbing and pressure pain in left frontal and temporal regions. The pain fluctuated in intensity, without side shift, possibly associated with sensitivity to light and sound early in its course, not worsened by exercise, not aggravated by eye movement or eye or orbital pressure, with tearing in both eyes only early in this headache syndrome. The patient described having occasional, intermittent episodes of left orbital/retro-orbital sharp pain of up to 30 min that were aggravated by noise and stress without any other symptom; he also recalled having had three episodes of severe, throbbing global pain, of 2–4 h duration, associated with dizziness, nausea, sensitivity to light and sound, which interfered with activity and increased with exercise when he was 50 years old. For the last several years before treatment, he had had only the constant left orbital and temple pain (intensity 7 out of 10) without any associated symptoms of migraine or any autonomic symptoms. The neurological and ophthalmological examinations were normal.

With the possible diagnosis of HC, the patient was treated initially with indomethacin 50 mg t.i.d. for 1 month without pain relief and with gastric pain as a side-effect. The patient was next treated with topiramate 25 mg hs which was slowly raised to 100 mg hs, plus imipramine 50 mg each night for chronic insomnia. He had marked improvement of pain from an intensity of 7/10 to 3/10 in a period of 3 months, and to mild pain (1/10) for the next 12 months. The imipramine was tapered off gradually and he did well for 1 month. Over a period of 4 months the pain slowly increased in intensity in the left eye without any autonomic symptoms and the topiramate was adjusted upwards slowly to 125 mg b.i.d. with rapid and continuous benefit from an intensity of 7/10 down to daily, continuous mild pain (1/10) for the last 5 months.

Past medical history included a benign thyroid nodule. He had a normal magnetic resonance imaging of the brain and orbits (2000), and repeat magnetic resonance angiography of the brain (2001), with and without contrast.

Discussion

Daily or near-daily headache is a widespread problem in clinical practice (7). The term chronic daily headache (CDH) encompasses primary headaches presenting at least 15 days per month and lasting more than 4 h per day, according to the Silberstein and Lipton criteria (8, 9). By these criteria, CDH includes transformed migraine (TM), chronic
tension-type headache (CTTH), new daily persistent headache (NDPH) and HC (8, 9). The International Classification of Headache Disorders (ICHD II) also lists chronic migraine (CM) as a CDH form (10). CDH affects a large number of patients according to population-based data from the USA, Europe and Asia (approximately 4–5% of the population) (7, 11). Approximately 2–3% of the population suffer from CTTH, which affects twice as many females as males; approximately 2% suffer from CM, 0.1–0.2% from NDPH and HC is even more rare (7, 11, 12).

This case represents a chronic, side-locked headache, with pain almost exclusively in the eye and slightly in the left temple. The most important diagnosis to rule out is HC, because of the potential for highly successful treatment with indomethacin, although this patient does not fit all the International Headache Society (IHS) criteria for HC, given the absence of clear unilateral autonomic features, lack of idiopathic stabbing pain and the lack of indomethacin responsiveness (10, 13–15). Hemian cranias continua, an uncommon primary headache syndrome first described in 1984 by Sjastaad and Spierings (16), represents up to 1.7% of patients with CDH (12). Interestingly, some aspects of the IHS diagnostic criteria for HC are still controversial; the first is the criterion of fixed lateralization, as there are case reports with side shift (17), and the second is the suggestion of indomethacin-resistant patients (the IHS criterion includes 100% indomethacin response) (18). Other clinical characteristics that have been described in HC are the presence of migrainous features during exacerbation periods (70.6%) (19), jabs and jolts or idiopathic stabbing pain (41–75%) (15, 20), and a foreign body sensation in the eye (20).

HC should be differentiated from the trigeminal autonomic cephalgias (TACs) (21–23), a group of primary headache disorders characterized by unilateral trigeminal distribution pain that occurs in association with ipsilateral cranial autonomic features. Although HC shares with paroxysmal hemicrania (PH) the presence of unilateral pain and the response to indomethacin, they differ in the duration and degree of pain (HC is constant and PH lasts for 5–15 min) and the extent of autonomic involvement (10, 13, 17). HC has been differentiated from cluster by the presence of indomethacin response, the male preponderance, the temporal pattern (10), the lack of effect of cluster medications in HC and the abnormal sweating pattern demonstrated in the central part of the forehead on the symptomatic side in some cases of cluster headache (17).

Another disorder that should be ruled out is side-locked migraine (SLM) (24, 25), although our patient does not fit all the migraine criteria (10). Primary trochlear headache was ruled out in our patient based on the absence of increasing pain during ocular movement or during palpation, and the pain improvement without steroid injection in the ipsilateral trochlear region (26).

In this patient, headache caused by jaw, ear or cranial disorders was ruled out by the absence of the suggested IHS criteria for each, the normal neurological examinations and the normal imaging studies (10). Pain around the eye can be caused by local ophthalmic disorders or by disease of other structures sharing trigeminal nerve sensory innervation (27).

There may be a secondary eye pain sometimes from structures not concerned with vision, such as contents of the orbit, superior orbital fissure or cavernous sinus, intracranial infiltrative, neoplastic or inflammatory disease processes such as temporal arteritis, also called giant cell arteritis. This vasculitis primarily affects adults over age 50 typically presenting with severe headaches, fatigue, polymyalgia-like symptoms, ischaemic complaints such as jaw claudication or visual loss in up to 50% (28, 29).

In general, most ocular causes of eye pain also cause the eye to be red, although some disorders do not. Headache can be associated with ocular signs and symptoms (27); in patients with headache seen for ocular examination, females (60%) complained more frequently of headache (P < 0.05), with migraine as the most important primary headache (3.9%). When described, headache is mostly fronto-occipital, frequently associated with decreased vision, eye pain, epiphora, foreign body sensation, itching and photophobia (30). On the other hand, in 64% of migraine patients several abnormalities on eye examination can be found (30). It is also important in our patient to consider the presence of facial migraine not coded in the ICHD II and rarely described, that can represent up to 12% of patients with facial pain. This migraine has been described as unilateral, in the forehead/eye/cheek or isolated to the second division of the trigeminal nerve (31).

Indomethacin has been the gold standard treatment for HC. However, there have been reports of a variety of medications effective in patients refractory to or with contraindication to the drug; these case reports include response to dihydroergotamine (32), methysergide (32), corticosteroids (33), lamotrigine (34), gabapentin (35) and rofecoxib (36, 37). It is not clear at this time if the indomethacin unresponsiveness is a feature strong enough to be considered an exclusion diagnostic criterion for HC (25).
It is our opinion that this patient, although having vague features consistent with HC and CM, has constant, left eye and temple pain without consistent autonomic or migraine features, and therefore fits best the IHS category of persistent idiopathic facial pain (13.18.4) (10). This was termed atypical facial pain according to 1988 IHS criteria. Other potential diagnoses include unilateral CTTHA or probable HC without indomethacin responsiveness (10).

Topiramate has been reported to be effective in migraine prevention (5, 6) and this case report suggests that topiramate may be effective in the preventative treatment of patients with chronic, side-locked unilateral headache in one eye, suggestive of persistent idiopathic facial pain or some of the above unusual diagnoses.

References