Hemicrania Continua - 2009

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Hemicrania continua (HC) is a primary headache disorder exquisitely responsive to the drug indomethacin. It was probably initially described by Medina and Diamond in 1981 [1] and officially named in 1984 by Sjaastad and Spierings [2].There are now more than 100 reports of HC in the literature. HC was not described in the first edition of International Classification of Headache Disorders by the International Headache Society in 1988 [3]. However it was included in the second edition in 2004 [4]. The incidence and prevalence of this condition is not known. Initially it was considered a very rare syndrome, but it may have been underdiagnosed [5, 6].The disorder has female predominance with a female to male ratio of approximately 2:1. The syndrome usually begins in adulthood, although the range of onset is from 5 to 67 years of age (mean 28 years) [5].

HC is characterised by a unilateral continuous pain of mild to moderate intensity. The pain is reported to be over mainly the forehead, temporal, orbital and occipital region, yet any part of the head or neck may be affected [7]. Typically patients have persistent background pain which is dull, achy and pressured in quality [5] and most of them experience superimposed exacerbations of more severe pain lasting from 20 minutes to several days [8]. Nocturnal exacerbations occur and can result in the condition's mis-diagnosis as other headache types such as cluster or hypnic headache. The exacerbations may occur in association with cranial autonomic symptoms and migrainous features. Cranial autonomic symptoms (eye dropping, eye-redness, eye-watering and nasal blocking) are often present and occur on the same side of the pain. Migrainous symptoms such as nausea, vomiting, photophobia (sensitivity to light) and phonophobia (sensitivity to sound) may also occur during exacerbations of pain. A typical migrainous visual aura (neurological symptom common in migraine patients) during the exacerbation of the pain has been described in four patients [9].

There are few factors that can precipitate severe pain. Factors typically reported in other headache types such as stress, menstrual period and alcohol generally seem to be less indicated in this condition. Neck movement does not trigger the pain, although occipital tenderness is present in 68 per cent of patients (ipsilateral 44 per cent, bilateral 24 per cent) [5]. Most patients have strictly unilateral pain, without side shift, although five patients with side shift attack [10-14] and three bilateral cases have been described [15,16].

Two temporal patterns are described in HC: an episodic (remitting) form characterised by a headache period separated by pain-free remissions and a chronic form (unremitting) in which the headache is persistent without remissions [8,17,18]. HC is chronic from the onset in 53 per cent of patients; evolves from an episodic pattern in 35 per cent; and is episodic from the onset in 12 per cent [5]. This condition is not characterised by seasonal pattern (whereas it is in cluster headache). However two patients with clear seasonal patterns have been reported [14,20].

HC is frequently misdiagnosed as another primary headache syndrome. The presence of cranial autonomic features during severe pain can lead to the wrong diagnosis of cluster headache (CH) and paroxysmal hemicrania (PH). Similarly the presence of migrainous symptoms during severe pain such as nausea, vomiting, sensitivity to light and sensitivity to sound can lead to misdiagnosis of HC as migraine. HC can be differentiated from cluster headache and migraine by the presence of continuous background pain of mild severity, presence of mild cranial autonomic symptoms during exacerbation of the pain and absence of exacerbation with strictly circadian periodicity. If a person diagnosed with PH experiences pain between attacks (called interictal pain) it may be that

the diagnosis should be HC instead. Useful pointers include: firstly prominent cranial autonomic symptoms in PH, secondly the attacks of PH are more frequent and short-lasting, thirdly it is our general impression that background pain in PH is generally less severe than it is in hemicrania continua [21]. Finally, the differential diagnosis from unilateral chronic migraine can be difficult, and the response to indomethacin is the only current criteria to distinguish these two primary headaches.

The diagnosis of HC is made on the basis of clinical history, neurological examination and positive response to indomethacin [4]. Neuroimaging such as magnetic resonance imaging (MRI) is a sensible screening investigation to rule out secondary underlying causes. It is extremely useful that the patients keep a headache diary before and during the trial with indomethacin in order to provide accurate information to the doctor regarding the headache pattern and also the response to indometacin. The response to indomethacin is usually rapid, with remission of the pain in 1-2 days once the therapeutic dose is achieved. Injectable indometacin 50 to 100 mg intramuscularly ("Indotest")has been proposed as a diagnostic test for HC [22] and complete pain-relief was present within two hours. The indotest is a straightforward test during the diagnostic work-up of this condition.

The treatment of HC is prophylactic and it has rapid and consistent response to indomethacin. The effective dosage is between 25 mg and 300 mg per day [5,8]. Skipping or delaying the dose can lead to a recurrence of the pain. During the indometacin trial a gastric-mucosaprotective agent should be added in order to reduce the possible gastric problem.

There are no other drugs that show consistent response in this condition and drugs that show a partial benefit include naproxen, paracetamol with caffeine, ibuprofen, piroxicam [18],rofecoxib [23],celecoxib [24] and melatonin [25]. Recently topiramate has been used successfully in four patients [13, 26, 27]. This drug may represent a possible option in patients that are unable to tolerate indomethacin. Furthermore, the past few years has seen an increasing interest in neurostimulation of the occipital nerve (ONS) as a treatment for medically intractable primary headache syndromes such chronic migraine, cluster headache and hemicrania continua [28]. Very recently the long term safety and efficacy of ONS in a large group of patients with HC has been reported [29].

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