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Hemicrania Continua

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Hemicrania continua (HC) is a rare, indomethacin-responsive headache disorder characterized by a continuous, moderate to severe unilateral headache that varies in intensity, waxing and waning without disappearing completely.[1] It is frequently associated with jabs and jolts (idiopathic stabbing headache). Exacerbation of pain is often associated with autonomic disturbances such as ptosis, miosis, tearing, and sweating. Although HC is not triggered by neck movements, tender spots in the neck may be present. It may be accompanied by photophobia, phonophobia, and nausea. This disorder almost invariably has a prompt and enduring response to indomethacin. Because some cases do not respond to indomethacin but meet the phenotype, an alternate means of diagnosis has been suggested.[2]

EARLIEST DESCRIPTIONS

Until recently, the diagnosis of HC was based solely on the clinical response to indomethacin; however, unilateral or one-sided headaches have been described for centuries. The earliest recognition of a headache syndrome involving one side of the head is attributed to Aretaeus of Cappadocia (second century AD), but Egyptian descriptions[3] appear in papyri dating from 1500 BC. The god Horus invokes the goddesses Isis and Nyphthys, begging them to let down a spare head from the sky because he cannot stand his one-sided headache any longer. Although the word for this headache, *gestep,* has been translated as "migraine," it could refer to the unilateral headache of HC.

Galen (AD 131-201) introduced the term *hemicrania* for unilateral headache. It was later transformed into the Old English *megrim* and French *migraine.* Today we accept the term *migraine,* but the condition is indistinguishable from hemicrania continua.

Medina and Diamond[4] were probably the first authors to describe HC (including a response to indomethacin). This was in 1981, in a subset of their 54 patients with variants of cluster headache. Thirty-nine of their patients had strictly (28) or predominantly (11) unilateral headache. The pain was usually mild to moderate, and throbbing or pressurelike. Forty-six patients had atypical cluster headache (exacerbation or localized pain without headache-free periods), 28 had background vascular headache (a chronic, continuous unilateral headache of variable severity), and 20 had multiple jabs. All patients who had atypical cluster headache plus multiple jabs or background headache (n=24) responded to indomethacin. In 17 patients the response was excellent, in 6 it was good, and in 1 fair. Many of these patients had strictly unilateral headaches that were responsive to indomethacin.

The term HC was coined by Sjaastad and Spierings[5] in 1984. They described 2 patients, a woman aged 63 years and a man aged 53 years, who developed strictly unilateral headaches that were continuous from onset and absolutely responsive to indomethacin. The man had associated autonomic features such as redness, lacrimation, and sensitivity to light, whereas the woman had superimposed jabs and jolts. These cases were presented in September 1983 at the first International Headache Congress in Munich, Germany. The same year, Boghen and Desaulniers[6] described a patient with a similar headache that they called "background vascular headache

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responsive to indomethacin." The patient was a 49-year-old man who had a 20-year history of left-sided headache that rarely radiated to the right side. The headache had a sustained, pressurelike quality and was associated with intermittent jabs. Treatment with indomethacin, 50 mg/d completely relieved the pain.

Approximately 100 cases of HC have been reported, but there is still uncertainty about its clinical features. Atypical features including bilaterality, alternating-side headaches, and unresponsiveness to indomethacin have been described. Two secondary cases, one a patient with a mesenchymal tumor[7] and another with human immunodeficiency virus,[8] have also been reported.

CLINICAL FEATURES

Hemicrania continua exists in both continuous and remitting forms. The continuous variety can be subclassified into (1) an evolutive, unremitting form that arises from the remitting form, and (2) an unremitting form characterized by continuous headache from the onset.[9] Although one of the essential features of HC is unilateral headache, some bilateral or alternating-side cases have been reported.[10,11] The female-male ratio of typical cases is 2.8:1.

ASSOCIATED SYMPTOMS

Associated symptoms can be divided into 3 main categories: autonomic symptoms, jabs and jolts, and migrainous features. Autonomic symptoms consist of conjunctival hyperemia, tearing, rhinorrhea, nasal stuffiness, eyelid edema, and forehead sweating. These symptoms are not as prominent in HC as they are in cluster headache and chronic paroxysmal hemicrania. Symptoms of ocular discomfort, at times premonitory, have been described in patients with HC.[12] Some patients report a feeling of sand in the eye, which may be specific for HC.

Jabs and jolts syndrome, described by Sjaastad[13] as sharp, knifelike pains less than 1 minute in duration, occurs in patients with tension-type, migraine, and cluster headaches and in headache-free individuals. Its prevalence in the general population is 30%.[14] We found 12 descriptions (26%) of jabs and jolts in the 46 cases in which the information could be ascertained.

Migrainous features are common in HC; 23 patients (50%) had at least 1 symptom of migraine (nausea, vomiting, photophobia, or phonophobia). It was not possible to apply International Headache Society diagnostic criteria for migraine in the previously reported cases because of a lack of information. Nausea, vomiting, photophobia, and phonophobia also occur in cluster headache and chronic paroxysmal hemicrania.[9]

Associated neurologic signs and symptoms include attacks of hemiparesis with a familial history of hemiplegic migraines[15] and unilateral paresthesias.[11] Antonaci and Sjaastad[7] reported visual phenomena described as dark spots in a case of HC secondary to a mesenchymal tumor.

TREATMENT AND PROGNOSIS

Hemicrania continua has been defined in the past by its response to indomethacin, with the dose ranging from 50 to 300 mg/d. Goadsby and Lipton[2] stated,

> It is likely that the indomethacin response that is shared between the paroxysmal hemicranias and HC will have some clear linked pharmacological basis and further suggests that a convenient classification of these headaches together may eventually have some biological basis.

Many patients discontinue the medication because of adverse effects. Other successful treatments include ibuprofen at 800 mg/d,[16] piroxicam β-cyclodextrin at 20 to 40 mg/d,[17] and rofecoxib at 25 mg/d.[18] Sumatriptan[19] and many other analgesics are not effective.

Espada et al[20] described 5 men and 4 women who were diagnosed as having HC using Goadsby and Lipton's preliminary diagnostic criteria[2] (8 cases were continuous and 1 was remitting, with a mean age at onset of 53.3 years [range, 29-69 years]). All 9 patients had initial relief with indomethacin (mean daily dose, 94.4 mg; range, 50-150 mg). Follow-up was possible in 8 patients. In 3 patients, indomethacin therapy was discontinued after 3, 7, and 15 months, respectively, and the patients remained pain free. Three patients discontinued treatment because of adverse effects and experienced headache recurrence; however, 2 of these had relief with aspirin. Two other patients continued to take indomethacin with partial relief.

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