CLINICAL CORRESPONDENCE

Hemicrania continua evolving from migraine with aura: clinical evidence of a possible correlation between two forms of primary headache

A Palmieri[1], F Mainardi[2], F Maggioni[2], F Dainese[2] & G Zanchin[2]

[1]*Headache Unit, Service of Neurology, San Donà di Piave Hospital and* [2]*Headache Centre, Department of Neurosciences, Padua University, Padua, Italy*

Professor Giorgio Zanchin, Headache Centre, Department of Neurosciences, Padua University, via Giustiniani 5, 35128 Padua, Italy. Tel. +39 49 827 3600, fax +39 49 875 1770, e-mail giorgio.zanchin@unipd.it *Received 3 November 2003, accepted 3 March 2004*

Hemicrania continua (HC) is an uncommon primary headache disorder originally described in 1984 by Sjastaad and Spierings (1) and characterized by a continuous, strictly unilateral headache of fluctuating intensity with exacerbations of more severe pain usually accompanied by autonomic disturbances such as conjunctival injection, lacrimation, nasal congestion, rhinorrhoea, ptosis, or eyelid oedema. Associated stabbing headaches have also been described. HC almost invariably has a prompt and enduring response to indomethacin (2). Remitting and unremitting forms have been identified (3). This entity has been included in the second revision of the International Headache Society (IHS) classification. Even though migrainous features can be part of HC, its relationship with migraine without or with aura has not been clarified.

We report a case of remitting HC with absolute response to indomethacin evolving from migraine with aura.

Case report

A 40-year-old woman was referred for a unilateral daily headache which had started 3 months earlier. She had suffered from migraine with typical aura from the age of 15, with a range of attacks of three to six a year. Her attacks were characterized by throbbing, moderate to severe headache, almost exclusively localized on the left side, and accompanied by nausea, phonophobia and photophobia. Headache was regularly preceded by ipsilateral visual scotoma and sometimes numbness and tingling in the left upper limb. The presentation of aura symptoms was gradual over time and its duration did not exceed 30 min; the algic phase began 15–20 min after the resolution of the aura and lasted about 6–12 h if untreated. Symptoms remained unchanged until 3 months before the first observation, when the patient began to complain of daily, continuous, left-sided headache with superimposed exacerbations of more severe pain. These exacerbations were always preceded by numbness of her left hand rapidly extending to the forearm and the left side of the mouth and tongue, lasting 5–15 min. In a few days paresthesias disappeared, but the symtomatology remained otherwise unchanged. Daily continuous headache localized in the left fronto-temporal region and mild to moderate in intensity was associated with two or three daily exacerbations of severe pain with nausea, eye redness and nasal congestion. Frequent short-lasting ipsilateral jabs and jolts were also reported. Common analgesics and non-steroidal anti-inflammatory drug administration had little or no effect. She never took indomethacin for the attacks or during previous migrainous spells.

The patient's mother had been affected by a migraine headache without aura; anamnesis was otherwise unremarkable. Neurological and general examinations were normal except for trigger point tenderness in her left supraorbital region. Neck symptoms were not present and neck movement was in the range of normal. Brain magnetic resonance imaging was normal, as were blood cell count, erythrocyte sedimentation rate, autoimmunity and coagulative tests.

A diagnosis of possible HC was made and the patient was administered indomethacin 75 mg/day. On the second day of therapy the symptoms gradually improved and on the third day the patient became pain free. Symptomatic drugs (including triptans) were not taken during this period owing to the rapid improvement of pain. After 1 month of treatment, indomethacin was gradually tapered and withdrawn

without recurrence of symptoms. The patient was still asymptomatic at the 6-month follow-up, except for a single spell of usual migraine with aura.

Discussion

The coexistence of HC with another form of primary headache is rarely reported in the literature, perhaps as a consequence of the very recent introduction of this headache in the IHS classification.

Recently our group described a case of HC with contralateral episodic cluster headache (4), in which each form of headache responded to its own appropriate therapy (indomethacin and verapamil, respectively). Our patient provides further evidence for a possible relationship between migraine and HC. She was affected by migraine with typical aura since adolescence, but at the age of 40 she suddenly developed a new form of continuous, unilateral, headache associated with pain exacerbations and autonomic symptoms, meeting the international diagnostic criteria for HC, including the absolute response to indomethacin (Table 1) (5). At the beginning of HC she experienced typical sensitive auras strictly preceding exacerbations of pain, representing an interesting clinical link between the two headache syndromes.

The clinical and pathogenic relationship between HC and migraine is poorly understood and described. The exact prevalence of migraine in patients suffering from HC is still unknown. The two clinical entities share common aspects such as migraine-like exacerbations associated with vegetative symptoms. Strict unilateral manifestation of pain and prompt response to indomethacin make HC closer to trigeminal autonomic cephalalgias. The so-called side-locked migraine shows intermediate characteristics between the two groups of headaches (6). Nevertheless, HC has been included in the fourth group (other primary headaches) in the recent revision of the IHS classification (5), which reflects the difficulty of a pathogenic classification of this syndrome.

Table 1 Diagnostic criteria of hemicrania continua (5)

A	Headache for >3 months fulfilling criteria B–D
B	All of the following characteristics:
	1. Unilateral pain without side-shift
	2. Daily and continuous, without pain-free period
	3. Moderate intensity, but with exacerbations of severe pain
C	At least one of the following autonomic features occurs during exacerbations and ipsilateral to the side of the pain:
	1. Conjunctival injection and/or lacrimation
	2. Nasal congestion and/or rhinorrhoea
	3. Ptosis and/or miosis
D	Complete response to therapeutic doses of indomethacin
E	Not attributed to another disorder

Auras are not an exclusively migraine-dependent phenomenon. They can present alone or in combination with other primary headaches such as cluster headache (7) and paroxysmal hemicrania (8). Recently, Peres et al. described four patients affected by HC with aura (9). In all patients aura was visual and occurred before or during a pain exacerbation phase. A personal or familial history of migraine or auras was absent in all patients. According to the authors, the complete response of auras and headaches to indomethacin suggests a possible pathophysiological relationship between the two phases, aura and headache, in HC. Our patient differs because she was primarily a migraine with aura patient who presented a coexistent remitting HC. Her migraine auras and headache were side-locked (almost exclusively involving the left side). At 6 months of follow-up she was asymptomatic but for a single attack of her previous migraine with aura. Our patient's headache could represent a clinical and pathogenic continuum between migraine, particularly side-locked migraine, and HC.

Further descriptions and more careful analysis of migraine aspects in HC patients are needed for better clarification of the relationship between migraine and HC.

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