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Episodic paroxysmal hemicrania without autonomic symptoms: Are there possible subgroups in PH?

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Paroxysmal hemicrania (PH) is an uncommon primary headache. Between 1974 and 2002, about 120 cases were reported in the English literature (1), and in the last few years single cases have no longer been reported. The prevalence of PH in the population is about 2% of the prevalence of cluster headaches (2), with a female : male ratio of 3 : 1 (3). Chronic paroxysmal hemicrania (CPH) with an unremitting course was the first form described in the group of PH, and the only form present in the first classification of the International Headache Society in 1988 (4). Later, an episodic variety, EPH, was reported and classified in the International Classification of Headache Disorders, 2nd edn (ICHD-II; 2004) (5). Here they are codified at points 3.2.1 and 3.2.2, respectively (5). For this reason, a differentiation in the prevalence between EPH and CPH is actually impossible to determine. The main features of PH are as follows: frequent attacks of strictly unilateral, very severe pain localized in the orbital, supraorbital or temporal regions; attack duration 2–45 min; and frequency usually ranging from two to 40 attacks a day. The pain is associated with autonomic symptoms, such as tearing, conjunctival injection, rhinorrhoea, nasal stuffiness, and ptosis of the omolateral eyelid (5). Among the diagnostic criteria is listed a rapid and complete response to indomethacin (5). CPH does not have a recognizable temporal pattern, whereas EPH, a more rare disorder, is characterized by bouts of frequent, daily attacks with the same clinical features as CPH, but separated by periods of pain remission lasting 1 month or longer (5). CPH can evolve from EPH (6). All cases of EPH in the literature to date have been unilateral and coincided with autonomic symptoms, with only one case with other unusual features other than lacking autonomic features (7). We describe a patient herein with a temporal headache pattern characteristic of EPH and a complete response to indomethacin even without autonomic phenomena.

Case report

A 65-year-old White man presented to our out-patient Headache Centre with a history of a headache of 2 years' duration. The patient's past medical history was negative. The general and neurological examinations were negative. The pain was described as severe, trafictive, periorbital, and always localized to the left side. The attacks had a frequency of 8–10 day without circadian occurrence, were not associated with autonomic features, and were without nausea, vomiting, phono- or photo-phobia, and not aggravated by physical activity or effort. The attacks occurred in series lasting about 15 days consecutively, separated by remission periods generally of 20–40 days, but occasionally longer. The crises, lasting between 10 and 30 min, were unresponsive to acetylsalicylic acid and non-steroidal anti-inflammatory drugs (ketoprofen, ibuprofen, ketorolac, and diclofenac), but completely responsive to indomethacin (25 mg t.i.d.); he had never used triptans. Routine laboratory screening, X-ray chest, cerebral magnetic resonance imaging with gadolinium, and an echo Doppler of the extracranial vessels were all negative. Follow-up evaluation at 1 year demonstrated the recurrence of about five clusters, immediately aborted at the beginning with indomethacin, generally for periods lasting 2–3 weeks.

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Discussion

This case represents some points of interest and topics for discussion. First, this report resembles a headache syndrome sharing the typical symptoms of EPH, with respect to the frequency and duration of attacks and the complete response to indomethacin, so that the ICHD-II diagnostic criteria for EPH are satisfied, with the exception of autonomic symptoms (5). This is the first case of EPH without autonomic features. Based on the literature, there is a case described as EPH without neurovegetative signs, but there are other unusual aspects for the 'classical type' of EPH. Specifically, these aspects include the bilateral localization of pain, photo- and phonophobia, and hormonal influence (7). In our case, the only unusual aspect was the lack of neurovegetative symptoms. Indeed, regarding CPH, two reports have refined the clinical characteristics of this syndrome (1–8), and of the 104 patients considered, the following findings are noteworthy: three patients were diagnosed with CPH without autonomic symptoms; one patient had only a sense of aural fullness; this symptom was present in the other two patients and reported previously in the other patient, and could be associated with other autonomic manifestations (8,9); and a report has been recently published of a trigeminal autonomic cephalalgia (TAC)-like syndrome without autonomic symptoms; however, in this case there was an unusual presentation in the temporal pattern, occurring in single episodes lasting up to 7 h each, only occasionally, for many years before increasing in frequency (10).

Second, we highlight the opportunity to investigate carefully the pain history of every patient. Pain is sometimes considered only a non-specific symptom and not always as thoroughly investigated as necessary. In contrast, we found primary or secondary syndromes in which the diagnosis was based principally on a careful examination of the modality, time of presentation, recurrence, and persistent and triggering factors of the pain, even in the absence of other subjective symptoms or neurological signs. With respect to primary headache, we find the assertion reported above in many forms of cephalalgias included in Chapter 4 of the ICHD-II (i.e. hypnic headache and stabbing headache), and with TACs in particular, the lack of cranial autonomic symptoms should not preclude a trial with indomethacin in a context of typical pain suggestive of PH. In fact it has been reported that in other cranial autonomic cephalalgias such as in cluster headache, a small percentage of patients (about 3%) do not have autonomic symptoms (11–14). It is also possible that even in PH, a small group of patients do not present cranial autonomic symptoms.

Regarding secondary headaches, the pain of claudication mandibularis in the temporal arteritis is a pathognomonic sign, and furthermore very suggestive of the pain in carotid dissection or in subarachnoid haemorrhage.

In the literature, there is some evidence of PH with an atypical presentation, as previously reported in our case of pain without neurovegetative symptoms, but on the other hand, of cases with neurovegetative symptoms, but without pain (15), a case of PH with aura (16), and a case of menstruation-related EPH (17).

In conclusion, we believe that the description of single cases of PH with variations is still useful; it could facilitate a better characterization of this syndrome, and may help to isolate different patterns of autonomic symptoms in PH patients and contribute to a better understanding of the pathogenesis of the symptoms. Clearly, it is necessary that larger PH series can reproduce sporadic findings. If this result is achieved, we propose an appendix in the next classification, the possibility to characterize new subgroups or variances of PH, as follows: without autonomic signs, when the other clinical characteristics are present; without pain, when autonomic signs and response to indomethacin are present; with aura, when preceded or accompanied by aura or menstruation-related with attacks strictly related to menses; and finally, to add to the list of cranial autonomic features, the sense of aural fullness during the attacks, as suggested by Cittadini et al. (8).

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