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HEMICRANIA CONTINUA

A report of ten new cases

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ABSTRACT - Hemicrania continua (HC) is an uncommon primary headache first described as a syndrome in 1984. Being quite unusual, its clinical characterization still demands better description. The aim of this study is to present the main clinical characteristics of 10 patients with the diagnosis of HC seen in a tertiary center, critically discussing their main features. All subjects had strictly unilateral headache without side shift and absolute response to indomethacin. Seven patients (70%) presented autonomic features during pain exacerbations. Four (40%) had migrainous symptoms during the exacerbations and one presented partial relief with dihydroergotamine. One patient had pain excruciatingly severe during the exacerbations. Although the cardinal features of HC – continuous, unilateral, indomethacin responsive, remain strongly reliable, a refinement on the clinical characterization is needful and desired.

KEY WORDS: hemicrania continua, clinical characterization, classification.

Hemicrania continua: relato de dez novos casos

RESUMO - Hemicrania continua (HC) é uma cefaléia primária relativamente incomum primeiramente descrita enquanto sindrome em 1984. Por ser relativamente rara, sua apresentação clínica ainda carece de melhor descrição. O objetivo do presente estudo é apresentar e criticamente discutir as principais características clínicas de 10 pacientes com HC consecutivamente vistos em um centro de atendimento especializado. Todos os pacientes apresentavam cefaléia estritamente unilateral, sem alternância de lado e com absoluta resposta a indometacina. Sete pacientes (70%) apresentavam sinais autonômicos durante a exacerbação da dor. Quatro (40%) apresentavam sintomas migranosos durante as exacerbações e um apresentava alívio parcial com diidroergotamina. Um paciente apresentava exacerbações excruciantemente severas. Embora as principais características da HC – dor continua, unilateral, responsiva à indometacina – permaneçam extremamente associados à sindrome, um refinamento na caracterização clínica é necessário.

PALAVRAS-CHAVE: hemicrania continua, características clínicas, classificação.

Hemicrania continua (HC) is an uncommon primary headache first described as a syndrome in 1984 by Sjaastad and Spierings[1]. They described a unilateral headache absolutely responsive to indomethacin, thus being one of the indomethacin-responsive headache syndromes[2]. During the next five years of its description, the number of HC cases reported increased to 18[3]. This relatively small number of cases reported in the literature at that time may have been one reason for not including HC in the International Headache Society (IHS) classification system[4,5], this disorder being expected to be addressed in its new revision[6]. A clinical description of HC was included in the International Association for the Study of Pain classification[7].

Several aspects concerning its clinical characterization and consequently classification remain controversial[8]. According Spierings[2] and Pareja et al.[4], HC is a unilateral headache syndrome with fixed lateralization, that is, the headaches always occur in the same side of the head. On the other hand, HC alternating sides, although rare, was already described[3,9]. Similarly, some authors consider the presence of autonomic features as a diagnostic criterion for HC[10], while others consider their presence as a negative criterion[4].

The aim of this study is to present the main clinical characteristics of 10 patients with the diagnosis of HC seen in a tertiary center, critically discussing their main features.

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METHOD

Clinical records and the headache diaries of 10 patients with HC seen between 1990 and 2002 at the New England Center for Headache, a tertiary referral center, were included after being randomly selected and reviewed. All patients have been followed for at least two consecutive years. During the entire time period reviewed, the clinic had utilized a uniform clinical intake form and headache calendars. The clinical intake form contains information regarding: 1 – Intensity of pain: pain is graded on a 3 point scale as severe, moderate or mild. The following parameters are evaluated for all intensities of pain the patients might present; 2 – Frequency of pain; 3 – Location of pain; 4 – Quality of pain; 5– Duration of pain; 6 – Aura and associated symptoms: 7 – Behavior during attacks.

After analysis of records, spreadsheets and headache calendars, relevant information were transferred to a standardized form that included the clinical description, response to indomethacin, treatment and outcome.

RESULTS

Our sample consisted of 10 patients, being 7 (70.0%) females, with age ranging from 34 to 61 years (mean= 45.4 years). The mean time of following was 31.3 months.

Table 1 displays the clinical characterization, response to indomethacin, response to treatment and outcome.

All subjects had strictly unilateral headache without side shift and absolute response to indomethacin. Patients 5 and 6, despite also showing an excellent response to indomethacin, developed gastric side-effects. Patient 5 did not show good outcome to other treatments, including non-steroidal anti-inflammatory, dihydroergotamine, corticosteroids and gabapentin. Patient 6 presented a satisfactory therapeutic response to rofecoxib. The 8 patients that did not have intolerance to indomethacin had complete and prolonged response.

Table 1. Clinical characterization of ten patients with hemicrania continua.

Patients	1	2	3	4	5	6	7	8	9	10
Unilaterality without side shift	X	X	X	X	X	X	X	X	X	X
Absolute and protracted indomethacin effect	X	X	X	X	X	X	X	X	X	X
Continuous but fluctuating severity	X	X	X	X	X	X	X	X	X	X
Intensity of pain during exacerbations										
Mild (no exacerbations)						X	X			
Moderate	X	X		X				X		X
Severe					X				X	
Excruciatingly severe			X							
Autonomic features with severe pain exacerbations										
a. None	X			X		X				
b. Conjunctival injection		X	X						X	X
c. Lacrimation		X			X				X	X
d. Nasal congestion		X			X					X
e. Rhinorrhea										X
f. Ptosis										X
g. Eyelid edema							X	X		
h. Idiopatic stabbing headache									X	X
Migraine symptoms during exacerbation										
a. None	X	X	X	X		X	X	X		
b. Throbbing pain					X				X	X
c. Nausea										
d. Photophobia									X	X
e. Phonophobia									X	X
Response to antimigrainous drugs									X	
Prolonged treatment with indomethacin	X	X	X	X			X	X	X	X
Persistence of pain					X					

Seven patients (70%) presented autonomic features during pain exacerbations, as shown in Table 1. Four (40%) had migrainous symptoms during the exacerbations and one presented partial relief with dihydroergotamine. One patient had pain excruciatingly severe during the exacerbations.

DISCUSSION

HC is an unusual, despite not rare[8], primary headache. Although criteria for HC were not incorporated in the International Headache Society (IHS) Criteria, established in 1988[5], operational criteria have been proposed[4,10]. Even the nosologic status of HC is not well established, with a group proposing that HC should be included in the group III of the IHS criteria[4] and other group defending that HC must be included in the group of the chronic daily headaches[10]. It seems reasonable, however, that the unique and overlapping clinical features with other trigeminal autonomic cephalalgias should assume priority over simply the number of headaches within a 30-day period[11].

One first mandatory feature of HC, according the authors that first described the syndrome[1,2] and one proposed diagnostic criteria[4], is that the headache must be unilateral with fixed lateralization. Our 10 patients showed strictly and fixed unilateral headache. Although this seems to be by far more common, attacks that remain strictly unilateral but switch from side to side have been described in patients with HC[12].

Patients with HC often respond in a dramatic manner to indomethacin. The complete response to indomethacin as a diagnostic test, however, remains a contentious issue, some authors arguing that this requirement is problematic[11]. Spierings defend that "hemicrania continua that is resistant to indomethacin, as suggested by Kuritzky[13] does not exist"[2]. The IHS classification does require therapeutic response criteria to make the diagnosis of some headaches, Tolosa-Hunt syndrome being one well-know example[5,14]. The response to indomethacin being so dramatic, it is reasonable to consider it as a strong indicative of HC. But it seems somewhat exaggerate to consider that HC not responsive to indomethacin does not exist. Since this criterion does not consider the responsiveness to medications other than indomethacin, the main issue appears to relate the possibility of response to other agents. A variety of medications other than indomethacin have been reported to be effective in patients with HC, including dihydroergotamine[15], methysergide[15], corticosteroids[16], lamotrigine[17], gabapentin[18] and rofecoxib[19]. Our 10 patients had total and absolute response to indomethacin. With the ongoing treatment, two developed gastric side-effects. One presented good, besides not complete, response to rofecoxib.

The diagnostic criteria proposed by Pareja et al.[4] require an intensity of pain from mild to severe, but not excruciatingly severe. This seems to be quite subjective. In our series, 2 (20%) patients had mild pain without exacerbations, 5 (50%) presented exacerbations of moderate intensity, 2 (20%) of severe intensity, but one of our patients had exacerbations excruciatingly severe, therefore do not fulfilling this criterion. In a series of 18 patients with HC, two attempted suicide because of the severity of the headache[3]. A patient with a classic picture of HC, as presented by patient number 3, should not be excluded if her subjective experience of pain was reported as very severe[6].

Although the intensity of cranial autonomic features associated with HC is not as intense as those seen with chronic paroxysmal hemicrania or cluster headache, and in spite of the fact that some authors consider their presence as a negative proviso[4], their presence in a significant number of patients has been demonstrated[8]. Our data support this concept. Just 3 (30%) patients did not present autonomic features during episodes of pain exacerbations.

Similarly regarding the migrainous features, also considered a negative proviso by some authors. Migraine-related symptoms are not so uncommon in HC[6]. Peres et al.[8] reported the presence of nausea in 46% of their patients, vomiting in 15%, phonophobia in 46% and photophobia in 54%. The same group reported visual aura in one patient with HC[20]. We found migrainous features in 3 (30%) patients, one presenting partial relief with dihydroergotamine.

The relative rarity of HC has made it difficult to figure out a comprehensive clinical picture. We aimed to present the main clinical features of this syndrome in a sample of consecutive patients seen in the tertiary care center, briefly discussing some relevant concepts. Although the cardinal features of HC – continuous, unilateral, indomethacin responsive, remain strongly reliable, a refinement on the clinical characterization is needful and desired.

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