Introduction

Trigeminal autonomic cephalalgias (TACs) such as cluster headache, chronic paroxysmal hemicrania (CPH), episodic paroxysmal hemicrania, short-lasting unilateral neuralgiform headache with conjunctival injection and tearing (SUNCT) are a group of unilateral painful syndromes all of which are characterized by pain attacks in the facial area innervated by the first branch of the trigeminal nerve and autonomic signs on the same side of the head. Overall, these headaches have a common pathogenetic mechanism, consisting in the activation of the trigeminal and autonomic systems by means of a trigeminal-autonomic reflex. The activation of the autonomic system is not dependent on the intensity of pain.

Levels of calcitonin gene-related peptide (CGRP) and vasoactive intestinal peptide (VIP) in cluster headache and in CPH are significantly higher during the painful phase compared to migraine with and without aura. These forms, therefore, have in common a brief period of pain (of varying length in the different forms) and the presence of local autonomic signs, which are the elements upon which specific diagnostic criteria are based.

The 1988 classification of the International Headache Society [1] identifies in this group two nosographic entities represented by cluster headache (in the episodic and chronic forms) and CPH, currently classified in group 3. Regarding this, the subcommittee suggested that the definition of the idiopathic headache group be changed from cluster headache and CPH to cluster headache and CPH to unilateral headache with autonomic signs, so that other forms may also be included within the same group. Even though the diagnostic criteria remain unchanged with respect to those of the international classification, some modifications to the comment on CPH have been proposed by the subcommittee.

As far as SUNCT is concerned, this form should be included in the group of unilateral headaches with autonomic signs, considering the large number of papers available in the literature on this topic and the clinical characteristics that are sufficiently homogeneous to permit precise diagnostic criteria to be defined.

The inclusion of episodic paroxysmal hemicrania and hemicrania continua is still debatable. An increase in the number of cases and more extensive critical reviews of the latter are necessary. The presence of analogous clinical pictures which are, however, secondary to organic diseases, makes it mandatory that adequate instrumental investigations be carried out for all forms diagnosed as TACs.

We propose the following modifications, hereupon applicable for cluster headache, chronic paroxysmal hemicrania and SUNCT to point 3 of the IHS classification [1]:

3. Unilateral headache with autonomic signs

3.1 Cluster headaches (no changes)

3.2 Chronic paroxysmal migraine

A. At least 50 attacks fulfilling criteria B-E.
B. Pain of severe intensity with unilateral, orbital, supraorbital and/or temporal location, always on the same side.
C. Frequency, >5 attacks per day for more than half the time.
D. Pain associated with at least one of the following signs on the side of the pain:
   – Conjunctival injection
   – Lacrimation
   – Nasal congestion
   – Rhinorrhea
   – Ptosis
   – Eyelid edema
E. Absolute response to indomethacin
F. Exclusion of organic causes through diagnostic instrumental examinations.

3.3 SUNCTs

A. At least 50 attacks fulfilling criteria B-E.
B. Attacks of unilateral pain in the territory of distribution of the first branch of the trigeminal nerve.
C. Attacks lasting from 5 seconds to 3 minutes.
D. Attacks having at least 3 of the following 4 characteristics:
   1. Stabbing and/or burning and/or neuralgic (like an electric shock) pain.
   2. Moderate to very severe pain intensity.
   3. Frequency varying from less than 1 per day to more than 30 per hour.
   4. The crises are triggered from trigeminal or extra-trigeminal trigger zones.
E. Pain is accompanied by lacrimation and conjunctival injection on the symptomatic side.
F. Exclusion of organic causes through diagnostic instrumental examinations.

Few case reports without autonomic signs have been reported. These need to be further investigated and the number of cases should be increased. Few cases of no response to indomethacin have been described, which need further confirmation. Some cases of the episodic form reported in the literature may represent pre-chronic forms of the same illness. Data are at the moment insufficient to describe the episodic form as a separate nosographic entity.

**Paroxysmal hemicrania (chronic and episodic)**

Although CPH [2–137] is also defined by the presence of accompanying autonomic signs, some cases lack these signs [8, 12, 46]. In other cases, the response to indomethacin is lacking (further criterion for classification) or, in the same patient, it was necessary to subsequently increase indomethacin dosages [13, 123]. This observation makes it necessary to reconsider the diagnosis in poorly responsive patients and to exclude organic causes, if this criterion has not been previously respected.

The lack of response to indomethacin may be a discriminating factor in borderline forms of cluster headache. In fact, the existence of CPH forms, in which painful episodes are of low frequency and long duration, for instance 5 episodes lasting 45 minutes, suggests this possibility. Under these conditions, pharmacological diagnostic tests may be useful, such as the induction test with nitroglycerin and the evaluation of the clinical response to subcutaneously administered sumatriptan or to steroid treatment.

Cases of apparently non-chronic paroxysmal hemicrania have been described. Therefore, a separate nosographic entity (like cluster headache) has been hypothesized. The lack of long-term follow-up does not allow the possibility to verify the stability of this episodic form in the same patient or to say if this form already represents a pre-chronic phase of CPH.

**SUNCT**

The headache form defined as SUNCT (currently not included in the IHS classification [1]) has been repeatedly described in the literature [138–176], and the homogeneity of the clinical pictures suggests that it should be included under the classification of unilateral headaches at point 3.3. The brief duration of pain (not more than 180 seconds) is typical in these patients. Few patients have crises which are of a longer duration, making a differential diagnosis difficult with brief episodes of CPH. In this case, the absolute response to indomethacin, to which SUNCT is unresponsive, should be useful to discriminate between the two forms. The lack of a significant response to indomethacin or to other treatments is, in fact, typical of this form, whereas treatment with lamotrigine seems to give promising results.

In the cases described in the literature, lacrimation and conjunctival injection were the more frequent autonomic signs [45]. The exclusion of organic causes (secondary forms have been already described in the literature) is also mandatory for this form (as in the other unilateral headaches) [45].

**Hemicrania continua**

Another clinical entity described in the literature is hemicrania continua [3–5, 6–9, 177–226]. This entity deserves to be included in unilateral headaches due to the location of the pain. Pain is described as continuous and of moderate intensity with “exacerbations”, that are sometimes characterized, as far as symptoms and duration are concerned, as more typical of migraine or cluster headache, with accompanying autonomic signs. In a strict sense, only these exacerbations should be classified as unilateral headaches with autonomic signs. The underlying headache, in fact, is never accompanied by autonomic signs.

Such exacerbations should be better characterized and possibly considered as headache crises superimposed on the underlying pain. It may be possible, in fact, to recognize among them some of the nosographic entities already codified by the IHS (e.g. migraine or cluster headache) or not yet included in this classification (e.g. SUNCT, idiopathic stabbing headache). As such, it is recommended to begin prophylactic treatment of these superimposed forms and to determine the characteristics of the eventual residual headache.

The observation that indomethacin may relieve the underlying pain and the exacerbations leads one to think that the onset of one of the latter forms is facilitated by the chronic underlying pain.
The pain of hemicrania continua may present in a non-periodic, discontinuous way, and in about 20% of the cases described in the literature exacerbations were not present [22, 112].

The classification criteria and the recommendations for hemicrania continua which may be included in group 3 of the IHS classification [1] are the following:

### 3.4 Hemicrania continua

#### 3.4.1 Continuous hemicrania continua.
- **A.** Headache which is present for at least three months.
- **B.** Strictly unilateral location of the pain.
- **C.** Pain with the following characteristics:
  - Continuous but fluctuating.
  - Moderate intensity, for at least half the time.
  - Absence of precipitating factors.
- **D.** Exacerbations of the pain with or without autonomic signs on the side of the pain are frequent.
- **E.** Absolute response to indomethacin.
- **F.** Exclusion of organic causes through diagnostic instrumental examinations.

#### 3.4.2 Discontinuous hemicrania continua.

The pain of hemicrania continua may present in a non-periodic, discontinuous way, and in about 20% of the cases described in the literature exacerbations were not present [22, 112].

The classification criteria and the recommendations for hemicrania continua which may be included in group 3 of the IHS classification [1] are the following:

### References

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