

BLACK, WHITE AND SHADES OF GREY

SUNCT or short-lasting chronic paroxysmal hemicrania?

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ABSTRACT - *Aim of the study:* To report a case of unilateral headache with two possibilities of diagnosis. *Method:* Case report. *Results:* Patient with unilateral, intense, stabbing periocular headache with conjunctival injection and tearing. Although the duration of attacks was typical of SUNCT, there was complete remission of the pain with indomethacin, suggesting that this was a case of chronic paroxysmal hemicrania with unusually short attack duration. *Conclusion:* Therapeutic trials of indomethacin on younger patients presenting clinical diagnosis of SUNCT could be tried on a more regular basis.

KEY WORDS: SUNCT, chronic paroxysmal hemicrania, headache.

Preto, branco e tons de cinza: SUNCT ou hemicrania paroxística crônica de curta duração?

RESUMO - *Motivo do estudo:* Relatar um caso de cefaléia unilateral com duas possibilidades diagnósticas. *Métodos:* Relato de caso. *Resultados:* Paciente com cefaléia unilateral, intensa, em pontadas em região periocular, com congestão conjuntival e lacrimejamento. Embora a duração das crises fosse típica de SUNCT, houve remissão completa da dor com indometacina, sugerindo que se trate de um caso de hemicrania paroxística crônica com crises de duração atipicamente muito curta. *Conclusão:* Testes terapêuticos com indometacina em pacientes mais jovens com diagnóstico clínico de SUNCT poderiam ser tentados de forma mais regular.

PALAVRAS-CHAVE: SUNCT, hemicrania paroxística crônica, cefaléia.

The classification of headaches by the International Headache Society (IHS)¹ classifies a special group of headaches involving the trigeminal-autonomic system as belonging to "group 3 of primary headaches". These are typically recurrent, unilateral headaches of moderate to severe intensity, and relative short duration (from a few seconds to a few hours). They are accompanied by conjunctival injection, tearing, rhinorrhea, sweating and pupillary alterations ipsilateral to the pain, denoting the involvement of the autonomic system. The most typical representative headache of this group is the cluster headache, described in 1952². Other entities were described, including chronic paroxysmal hemicrania³ and "Short-lasting Unilateral Neuralgiform headache attacks with Conjunctival injection and Tearing" (SUNCT)⁴ but, as in cluster headache⁵, all these headaches can be in summary described as intense, accompanied mainly by lacrimation and conjunctival injection. The most typical difference among these three headaches (and the neuralgia of the first division of the trigeminal

nerve as well 6) is the duration of the attacks¹, although there are other features to be considered, such as age of appearance of the headache, gender and trigger points⁷. The latter ones are not part of the diagnostic criteria. Table summarizes the main findings in these headaches from the group three of the international classification.

We report the case of a patient whose attacks of pain led to the discussion of the criteria "duration of the attack" as a setback for the classification of her headache.

CASE REPORT

Female caucasian patient, aged 48, presenting a history of headache for the last two years. The headache was strictly unilateral (left side) and, as described by the patient, it started all over the temporal area and within a couple of seconds it is "concentrated" on the left eye, which becomes red and "full of tears". The pain was very intense, like a "burning heavy pressing stabbing on my eye" (9 out of 10 in the numeric pain intensity scale). The patient also referred that, during the attack, a sensation of heat devel-

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Table. Differential diagnosis of cluster headache, chronic paroxysmal hemicrania and SUNCT.

Characteristics	Cluster headache	Chronic paroxysmal hemicrania	SUNCT
Prevalence	0.1 to 0.4%	rare	very rare
Gender ratio (M:F)	9:1	1:3	8:1
Localization	orbital/temporal	orbital/temporal	orbital/temporal
Intensity	unbearable	very intense	very intense
Pain	excruciating	transfixing stabbing	transfixing stabbing
Attack duration	15 to 180 min	2 to 30 min	5 to 240 sec
Ipsilateral autonomic signs	present	present	present
Indomethacin efficacy	usually no	yes	no

oped in the left side of the forehead, followed by sweating. She described her appearance during the attack as following: "People who see me during an attack cannot believe how much I change in such a short time".

Since the beginning of this headache the attacks have always had these same characteristics, lasting one minute on average, and only once the pain lasted for nearly four minutes. The patient was very sure about the duration of the attacks and often timed it. The frequency of attacks was between 6 and 10 per day, and the patient did not know any way to force them to appear. She had been woken up by the attacks, though this was extremely rare to occur. Previous to these two years, the patient had not experienced headaches. The present pain made the patient seek medical help with clinicians and, during the investigation, she received a variety of diagnosis she was not aware of. At the time of her consultation with us, she was undergoing treatment for high blood pressure, taking captopril 50 mg/day and hydrochlorothiazide 25 mg/day. She had diabetes mellitus type 2, well controlled with diet, metoformin 850 mg/day and glibenclamide 15 mg/day. Hypertriglyceridemia was under control with ciprofibrate 100 mg/day. All these medications had been in use for less than a year and did not have temporal relation to the headache. There was no family history of headaches. Her father had died three years ago due to pneumonia, her mother was alive and well, with controlled diabetes mellitus type 2. Her clinical and neurological examinations were normal, except for exudates on the retina, classified as type I hypertensive retinopathy.

Although the headache was classified as SUNCT, we decided for a therapeutic trial with indomethacin considering it was worth excluding the diagnosis of chronic paroxysmal hemicrania. The patient returned one month later with significant improvement of the headache taking indomethacin at the dose of 50 mg/day. The attacks were now infrequent, about one day per week, and the intensity of the pain was reduced to "moderate, tolerable" (7 out of 10 in the visual analogical scale of pain). Indomethacin was suspended and the headache returned with the same intensity and frequency after one day. Upon restarting indome-

thacin, the headache was again much less frequent and less intense. The patient preferred to keep the dose of 50 mg/day rather than increasing it, claiming that the remaining few attacks of headache were "perfectly tolerable". She remains with this dose and is very satisfied with the result.

The patient gave the informed consent for this case report.

DISCUSSION

SUNCT is one of the most rare and most difficult to treat types of headache. Despite descriptions of improvement with the use of antiepileptic drugs (more recently known as membrane-stabilizing drugs), there is no definite treatment for SUNCT⁸. On the other hand, chronic paroxysmal hemicrania is completely responsive to treatment with daily doses of indomethacin³. SUNCT is more frequent in older males⁷ while chronic paroxysmal hemicrania predominates in younger females⁹. There have been cases of SUNCT described in younger women¹⁰ and children¹¹, but these are most infrequent.

The patient here described had a headache fulfilling all the criteria for diagnosis of SUNCT. However, being a female on her 4th decade, we considered it worth a trial with indomethacin, since SUNCT is a diagnosis that would invariably lead to a frustrating sequence of trial treatments. Indeed, if we had started her on other drugs than indomethacin, it is most likely that she would not have responded at all.

Having a successful remission of her headache with indomethacin, the case was then re-classified as "probable chronic paroxysmal hemicrania", since it did not fulfill item B of the classification of the IHS¹, that is, the attacks did not last between two and 30 minutes. We considered this re-classification better than considering it to be a SUNCT case which full-

filed all the criteria of the IHS¹, but responded to indomethacin.

The difficulties regarding classification of headaches in some patients led Young et al. to consider a modular theory for headaches¹². These authors propose that groups of neurones called modules become activated to produce each symptom of a primary headache disorder, and that each module is linked to other modules, producing the headache of a particular individual. A group of Brazilian authors has recently published a case with overlapping characteristics of episodic paroxysmal hemicrania and cluster headache¹³.

In the IHS criteria there is an overlap of duration of attacks between SUNCT (five to 240 seconds) and chronic paroxysmal hemicrania (two to 30 minutes) making it difficult to classify attacks which last between 120 and 240 seconds. For some cases, it has been suggested that a trial of indomethacin could be performed in order to confirm or exclude the diagnosis of chronic paroxysmal hemicrania¹⁴. We suggest that such trial should be performed even if the attacks last for less than two minutes.

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