A case of SUNCT syndrome responsive to verapamil

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SUNCT is a rare headache disorder, first described by Sjaastad et al. in 1989 (1) and recently codified by the International Classification of Headache Disorders as a primary headache (2). The literature, however, discloses some cases of secondary SUNCT syndrome, particularly due to lesions in the posterior fossa or involving the pituitary gland (3–10). The pharmacological treatment of this syndrome is problematic. Some encouraging results have been recently suggested with lamotrigine (11–15), gabapentin (16, 17) and topiramate (18, 19); all previous attempts at treatment with drugs used for other short-lasting headaches were unsuccessful (20). It is noteworthy that two reports (21, 22) even reported a worsening effect of verapamil in patients affected by SUNCT.

We describe a patient with SUNCT syndrome and an ischaemic lesion in the posterior fossa, who was successfully treated with verapamil.

Case report

A 61-year-old man came to our observation 4 years ago because of a persistent headache. At the age of 54 he began to suffer from a stabbing pain, strictly localized in the right supraorbital region, accompanied by ipsilateral conjunctival injection, lacrimation and rhinorrhea; each pain attack lasted for 30–60 s, occurred once or twice a day, and recurred every 1 or 2 months. This headache period ended spontaneously after 7 months; a second attack period began 7 months later and lasted 1 month; on this occasion the attacks occurred twice a day and recurred weekly.

After six painless years, the above short-lasting attacks reappeared and became more severe: they lasted 60–240 s, occurred up to 10 times per day, often at night, and recurred daily for 20 days running; these headache periods alternated with painless periods lasting about 20 days. Four months later our patient experienced a further increase in the frequency of his attacks, occurring up to 20 times per day; he also complained of a duller persistent interictal pain. A 1-month treatment with indomethacin (200 mg/day) had no effect and the patient came to our attention.

Precipitating factors were absent and so were nausea, vomiting, photophobia and phonophobia. His neurological and general examinations were normal. His family history disclosed the presence of cerebrovascular diseases but no headaches, his personal history revealed the presence of high blood level of triglycerides, which the patient never treated. He underwent a magnetic resonance imaging (MRI) and MRI angiography of the brain, which revealed the presence of a previous right cerebellar lacunar infarct.

We started treatment with verapamil (240 mg b.i.d.); 2 days later our patient experienced a complete absence of the attacks and also of the duller interictal pain. After 20 days, because of the recurrence of occasional attacks, we increased the verapamil dosage to 320 mg t.i.d. and the benefit was again complete.

The patient came to our attention again 4 years later. He had been on the treatment with verapamil during the intervening period, at a dosage of 240 mg b.i.d. He had had sporadic attacks (occurring once or twice a day and recurring every 1 or 2 months), lasting about 30 s and consisting of a painful phase alone; he reported no local autonomic manifestations. Whenever the patient tried to reduce the verapamil dose, taking a daily dose of 180 mg/day, the clinical picture worsened markedly. The drug was well tolerated and the patient was satisfied with the results.

Discussion

Our patient’s short-lasting headache meet the diagnostic criteria for SUNCT (2). In fact, this headache sometimes shows some clinical features of
paroxymal hemicrania, but this diagnosis is ruled out by the inefficacy of indomethacin. Sex and age at onset are also consistent with a diagnosis of SUNCT syndrome.

SUNCT is a primary headache; the literature, however, suggests the existence of secondary forms to lesions in the posterior fossa or at level of the pituitary gland (3–10). In our case a right cerebellar lacunar infarct was detected by MRI; it is noteworthy that this lesion is homolateral to the pain side and localized in the posterior fossa. These findings are intriguing, but not sufficient to relate the detected lesion to the clinical picture; data were also lacking about the making of the right cerebellar infarct and the onset of the short-lasting headache. It is therefore difficult to decide if our patient has a primary or secondary SUNCT syndrome.

To our knowledge this is the first SUNCT case responsive to verapamil. The drug, at a dose of 240 mg/day, reduced markedly both the frequency and severity of the attacks and controlled completely the interictal duller pain; whenever the patient tried to reduce the drug dosage, the attacks worsened. This indicates that the results obtained were due to verapamil and that the headache syndrome of our patient is now chronic.

SUNCT syndrome is known to be refractory to pharmacotherapy; most drugs used in the treatment of other short-lasting headaches are not useful in SUNCT (20). Verapamil has even been reported to worsen the attacks in some patients (21, 22). In our case the positive response of SUNCT symptoms to verapamil is, in our view, unquestionable and still unchanged after a 4-year follow-up. This finding suggests that verapamil should not be used as a pharmacological precipitation of SUNCT attacks (22) and should be considered a possible prophylactic treatment for SUNCT, along with lamotrigine (11–15), gabapentin (16, 17) and topiramate (18, 19).

The pathophysiology of SUNCT is still unknown; May et al. (23) recently detected in a SUNCT patient an ipsilateral hypothalamic activation similar to that detected in cluster headache patients; so the positive response of a SUNCT syndrome to an active drug against cluster headache (24) is not surprising.

References

20 Goadsby PJ, Lipton RB. A review of paroxysmal hemicranias, SUNCT syndrome and other short-lasting headaches
with autonomic features, including new cases. Brain 1997; 120:193–209.