

SUNCT syndrome responding absolutely to steroids in two cases with different etiologies

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Abstract SUNCT is a syndrome of short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing. We are presenting now two cases absolutely responders to steroid therapy, one of them a SUNCT-like secondary to a prolactinoma and the other primary.

Keywords SUNCT · Steroids · Prolactinoma

Introduction

SUNCT is a syndrome of short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing. It was first described in 1978 and fully characterized in 1989 [1, 2].

Suggested criteria for its diagnosis include attacks of unilateral, moderately severe orbital or temporal stabbing or throbbing pain lasting 5–240 s, with an attack frequency of 3–200 per day and pain is associated with important

ipsilateral autonomic manifestations [3]. It is among the rarest headache syndromes.

Several patients with secondary SUNCT syndromes have been reported since 1991. They were associated with cerebellopontine angle MAV, parapontine cavernous angioma, dorsolateral brainstem infarction, HIV infection, basilar impression, craniosynostosis, and hypophyseal tumors [4].

Treatments—including methysergide, lithium, propranolol, amitriptyline, carbamazepine and prednisone—usually have poor or negative results. Recently, some partial improvements have been described with the use of lamotrigine, gabapentin and topiramate [5–8].

We are presenting now two cases absolutely responders to steroid therapy, one of them a SUNCT-like secondary to a prolactinoma and the other primary.

Case 1

A 50-year-old male patient had his first pain attack 4 years before admission. He described a daily severe pulsating pain over the left supraorbital ridge and orbit, extended to the left temple, associated with prominence of autonomic symptoms such as lacrimation and conjunctival injection. Each individual attack lasted 15–120 s and occurred three to six times per day. Neck and head movements triggered pain attacks. He was prescribed amitriptyline, indomethacin, topiramate, and carbamazepine, which did not ameliorate his pain.

In between, he developed an ipsilateral shoulder pain and was locally infiltrated with steroids. After this procedure he remained free of head pain for a month.

He had had a left IV nerve palsy 3 years before which improved spontaneously. At this point neuroimaging was not done because the patient did not accept it.

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Fig. 1 Macroprolactinoma invading left cavernous sinus at the moment of diagnosis

On admission, clinical examination was unremarkable. Verapamil was prescribed and the pain worsened both in frequency and intensity as described [9]. The following therapeutic choice was prednisone at a dose of 40 mg per day with remission of pain in 24 h. Each time we tried to stop the steroid treatment, recurrence of pain was observed. Finally, a minimal dose 4 mg/day was sustained with optimal results. The patient was recommended again to

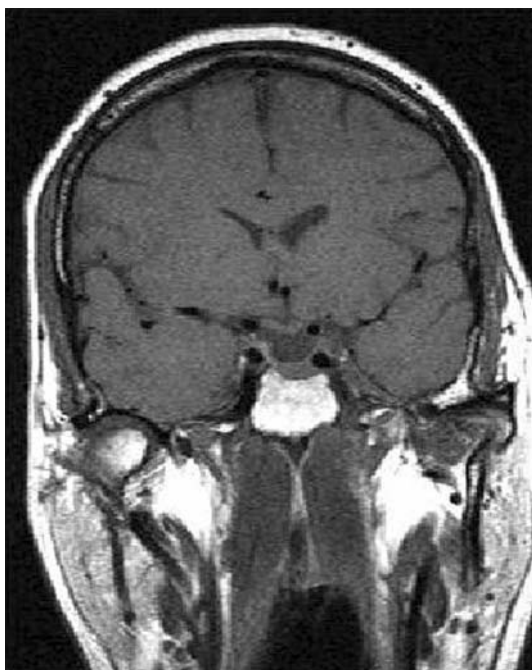


Fig. 2 Reduction of the tumoral mass at 7 months of cabergoline treatment

have a neuroimaging study done and this time he accepted. An MRI scan of the brain with and without gadolinium was performed. The study revealed a pituitary adenoma also extended to the left cavernous sinus wall (Fig. 1). No signs or symptoms of the tumor were present. Visual field testing was normal. Serum prolactin levels were markedly elevated (1,853 ng/mL, normal range 0–20 ng/mL), but testosterone, oestrogen, luteinizing hormone, follicle stimulating hormone, thyroid stimulating hormone, free thyroxin and growth hormone levels were normal. A diagnosis of macroprolactinoma was made and treatment with 2 mg/week of cabergoline was started. A month later prednisone was stopped with no recurrence of headache. Three months later prolactin serum levels were within normal range (5 ng/mL) and a control MRI after a 6-month follow-up showed a marked reduction of the neoplasm size (Fig. 2). The patient has remained asymptomatic with 1 mg/week of cabergoline (prolactin 2.4 ng/mL) since prolactinoma was diagnosed 7 months before.

Case 2

A 45-year-old male patient had experienced severe left orbital side pulsating pain associated with tearing of the ipsilateral eye and conjunctival injection for 2 months. Potential triggers were strong smells.

Pain attacks lasted 30–90 s and occurred 20–50 times a day. He had no past history of headache and did not recall any trauma to his head. Clinical examination and brain MRI were unremarkable.

SUNCT syndrome was diagnosed and therapeutic options started. The response to indomethacin, lithium, carbamazepine, amitriptyline, and ergotamine was frustrating.

Finally 40 mg/day prednisone was prescribed, and a complete remission of pain was reached after 48 h.

Prednisone dose was decreased slowly until reaching 4 mg/day in 6 weeks. At a dose of 2 mg/day, the pain recurred. For this reason, steroid has been maintained in 4 mg/day. The patient has remained asymptomatic since the steroid drug was installed 15 months ago.

Discussion

Although SUNCT is a form of primary headache, symptomatic cases have been reported [4]. The treatment of this head pain has been very difficult and frustrating. Partial response to several drugs has been published [5–8].

We are now reporting another two cases with similar headache characteristics, and therapeutic response but with different etiology. Table 1 shows a comparison of clinical characteristics between both patients and IHS criteria.

Table 1 Comparative clinical findings

IHS criteria SUNCT	Case 1: secondary SUNCT	Case 2: primary SUNCT
Unilateral orbital, supraorbital or temporal stabbing or pulsating pain lasting 5–240 s	Left supraorbital ridge, orbit and temple pain lasting 15–120 s	Left orbital side pain lasting 30–90 s
Pain is accompanied by ipsilateral conjunctival injection and lacrimation	Ipsilateral lacrimation and conjunctival injection	Ipsilateral lacrimation and conjunctival injection
Attacks occur with a frequency from 3 to 200 per day	Three to six times a day	20–50 times a day
Not attributed to another disorder	Hypophyseal tumor	Normal neuroimaging

In the first case (symptomatic), the transient left 4th nerve palsy observed 3 years before, was not studied due to the patient's decision. Probably, the hypophyseal tumor was already present.

When the headache started, different therapeutic drugs were used with disappointing results. When a left shoulder pain was present and a dexamethasone injection was prescribed, the patient was free of head pain for several weeks.

Steroids (first used because of the patient's omalgia and then because of his headache) resolved the pain immediately and recurrence was observed each time they were stopped.

As described previously, verapamil increased pain level. Finally, the patient accepted to be studied and, through laboratory tests and neuroimaging, a macroprolactinoma was diagnosed. Specific treatment with cabergoline was installed with remission of pain in accordance with the reduction of tumor size and serum prolactin levels. Cabergoline therapy allowed to suppress prednisone with no recurrence of SUNCT symptoms.

We can justify the causal relationship considering:

1. Temporal relationship between the prolactinoma and the onset of pain.
2. Side concordance: left cavernous sinus wall infiltration and left side pain.
3. Prompt pain remission with specific therapy for the tumor.
4. A 7-month follow-up.

If we considered that the suprasellar and left extension of the tumor into the cavernous sinus might be responsible for trigeminal activation leading to a homolateral SUNCT syndrome, steroids benefits may be related to an anti-inflammatory effect on both the tumor and the cavernous sinus edema. The reduction of the pituitary mass by

specific drugs made the anti-inflammatory effect of steroids unnecessary.

Considering that the pathophysiological mechanism of primary SUNCT syndrome remains unknown, in the second case presented, it is more difficult to find a rational explanation to the mechanism of prednisone action. In spite of this, some patients may benefit from prednisone treatment.

Conflict of interest None.

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