



# A Case of Resistant SUNCT That Responded to Steroid Treatment

## Steroid Tedavisine Yanıt Veren Dirençli SUNCT Olgusu

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### Summary

Conjunctival injection and tearing with unilateral short lasting neuralgiform headache syndrome is called as SUNCT. Even though these headaches are reported seldomly, their prevalence is possibly higher than what is expected. Detection of this rather uncommon symptom is of importance since its management differs from common primary headaches. Reports to date show that it has been treated with different types of drugs. Here we reported a 30 years old male patient with normal neurological examination, blood examination and neuroimaging. Our patient gave no response to indomethacin, gabapentine and carbamazepine treatments. This case is an example of SUNCT treated with steroid. (*Turkish Journal of Neurology* 2014; 20:23-25)

**Key Words:** Headache, SUNCT, steroid, treatment

### Özet

Konjunktival kızarıklık ve göz yaşarmasının eşlik ettiği unilateral kısa süreli nevaljiform ağrılar sendromu (Short-lasting Unilateral Neuralgiform headache attacks with Conjunctival injection and Tearing) SUNCT olarak tanımlanmıştır. Seyrek olarak bildirilmesine rağmen, sıklığı tahmin edilenden daha yüksektir. Tedavisi de diğer yaygın baş ağrısı sendromlarından farklı olduğundan tanınması önem taşımaktadır. Şimdiye kadar bildirilen vakalar birçok farklı ilaç ile tedavi edilmeye çalışılmıştır. Biz burada 30 yaşındaki nörolojik muayenesi, rutin kan tetkikleri ve görüntülemesi normal olan bir erkek hastayı sunduk. Hastamız indometazin, gabapentin ve karbamazepin tedavilerine cevap vermemiştir. Bu vaka steroid ile tedavi edilen SUNCT hastalarına bir örnektir. (*Türk Nöroloji Dergisi* 2014; 20:23-25)

**Anahtar Kelimeler:** Baş ağrısı, SUNCT, steroid, tedavi

### Introduction

SUNCT ("Short-lasting Unilateral Neuralgiform headache attacks with Conjunctival injection and Tearing"), is characterized by stabbing or throbbing pains with sudden onset and offset, lasting 5-240 seconds and targeting unilateral orbital, supraorbital or temporal regions. The attack frequency can vary between once a day and 30 in one hour. The pain can be accompanied either by conjunctival redness or tears in addition to the possible co-occurrences of cranial autonomic symptoms such as nasal congestion, rhinorrhea or eyelid edema. The ipsilateral conjunctival redness and tearing starts 1-2 seconds after the pain. The pain is maximal at the trigeminal nerve's first branching, unilateral, at medium or high intensity and can be described as stabbing, burning or electrical in character (1).

SUNCT is a treatment-resistant syndrome. Here we present a SUNCT case which responded to steroid treatment and discuss it in the context of findings from the literature.

### Case

Thirty year old male patient described stabbing and burning pain located on the right temple and the upper part of the eyes, started 3 months ago with 5-6 attacks per day and increasing up to 20-25 attacks in the last month. He reported that these attacks lasted 5-10 seconds initially but they got longer recently, reaching 15-20 seconds. The patient expressed that the pain struck at a random time of the day and night attacks had begun recently.

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The patient, who did not report a remarkable finding in the medical history and anamnesis was seen to be normal in the neurological and systemic examination, and showed normal results in the complete blood count, routine biochemistry and hormonal profile tests. His cranial magnetic resonance imaging (MRI) and MR angiography examinations were also normal. Trigeminal neuralgia, paroxysmal hemicranias and primary stabbing headache were ruled out for the patient who did not respond to indomethacin 150 mg/day, gabapentin 1800 mg/day and carbamazepine 800 mg/day. After seeing a lack of response to the additional two antiepileptic drugs, lamotrigine was not used.

The patient was started on a 48 mg/day oral methylprednisolone treatment with the SUNCT diagnosis. In the second day of the treatment, there was a marked improvement in his complaints. The dosage was gradually reduced in the following 8 weeks. During his 3 month follow-up, his autonomic symptoms as well as the frequency and the intensity of his attacks were remarkably reduced.

## Discussion

First described by Sjaastad in 1980, SUNCT was classified under trigeminal autonomic headaches according to the International Headache Association Classification of Headache Disorders (1,2). There has been over 50 reported cases in the literature. The number of attacks is between 3 and 200 with the duration of each attack changing between 5 and 240 seconds, generally lasting under a minute. The pain is confined in orbital, supraorbital and temporal regions. In most of the attacks, the pain is of pulsatile, throbbing character, presenting as medium or intense burning, stabbing or electrifying pain. The pain is perceived as mild-medium intensity. The pain is accompanied by ipsilateral abnormal vasomotor reactivity and different autonomic disorders: conjunctival redness, tearing and intraocular pressure increase; nasal discharge and labored breathing; eyelid edema with affecting primarily the upper parts. Sometimes subclinical sweating is added to this profile. The attacks can generally be triggered by the stimulation of the innervation areas of the trigeminal nerve and rarely the extratrigeminal region. The attacks are more frequent during the day than during nighttime. They also do not respond to conventional treatment, especially indomethacin. The syndrome is rare and more prevalent among men. Williams et al. reported 24, Cohen et al. reported 43 cases (3,4).

Even though the diagnostic criteria include unilaterality, Kuhn et al. recently reported a case with bilateral attacks (5). The clinical symptoms were thought to developed in response to the trigeminal autonomic reflexes and that the ipsilateral posterior hypothalamic

activity is responsible for the pathogenesis of the syndrome. Among the studies investigating the hypothalamic pathogenesis, cerebral imaging studies, the clinical effect of deep brain stimulation on the posterior hypothalamus in one case, the hormonal and clinical response attained after clomiphene administration in a case with low testosterone levels and the permanent response to verapamil in another case can be considered (6,7,8,9). Even though the hypothalamic involvement seems to be important in idiopathic SUNCT, in the studies published by Zidverc-Trajkovic et al. and Köseoğlu et al. another remarkable phenomenon was that the pressure applied on the trigeminal nerve evoked the symptoms in their cases, by means of activating the trigemino-autonomic reflex arch (10-11). In another case reported by Sprenger et al., vascular compression was observed in the MRI examination. After the surgical decompression treatment, a permanent response was observed during a 7-month follow-up period (12).

May et al argued that lamotrigine is the most effective agent in the treatment of SUNCT; topiramate and gabapentin can also be used and intravenous lidocaine can be effective in the acute treatment (13).

In a bilateral SUNCT case, Kuhn et al. observed positive response to topiramate treatment (5). Similarly Rossi et al. and Matharu et al. also confirmed this positive response in their cases (14,15). Ikawa et al. reported positive results to zonisamide in their own case (16).

Marziniak et al, on the other hand, achieved good outcome with oxcarbazepine and gabapentin (17). Gupta et al. also reported favorably about lamotrigine in a SUNCT case (18).

Trauninger et al. obtained remission in 3 idiopathic and episodic SUNCT cases by using methylprednisolone and suggested that the efficacy of methylprednisolone is possibly due to its anti-inflammatory effect (19). Maihöfner et al. achieved good results in a case with steroid treatment (20) as well as Figuerola et al. in 2 cases (21). Raimondi et al. reported good outcome in 2 SUNCT cases: one with carbamazepine alone and carbamazepine with steroid in the other (22).

Sporadically seen primary headaches constitute a heterogeneous group of headaches with still vaguely defined mechanisms, clinical profiles and treatment strategies. SUNCT is another rare manifestation of such headaches. For this reason, it is of crucial importance to investigate the mechanisms responsible for these disorders by reporting their properties and disseminating the knowledge on treatment methods.

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Table 1. SUNCT Diagnostic Criteria

<b>A.</b>	At least 5 attacks satisfying B and D criteria
<b>B.</b>	Unilateral orbital, supratentorial or temporal throbbing or pulsatile pain lasting 5-240 seconds
<b>C.</b>	Pain accompanied by unilateral conjunctival redness and tearing
<b>D.</b>	Pain repeating 3-200 times a day
<b>E.</b>	Pain unexplained by any other causes

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