

# SUNCT-Like Syndrome In A Patient With Small Cell Lung Cancer

Küçük Hücreli Akciğer Kanserli Hastada SUNCT Benzeri Sendrom

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Short-lasting unilateral Neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) is a rare form of primary headache disorder. SUNCT-like syndrome or secondary SUNCT has been also described in the literature. We report the first case of SUNCT syndrome in a patient with a previous diagnosis of small cell lung cancer. Brain magnetic resonance imaging of this patient revealed a lesion located in the right temporal muscle. He had a moderate response to the treatment with carbamazepine. SUNCT-like syndrome diagnosis should be evaluated carefully in patients with previous history of lung cancer. Cranial metastases may be a trigger for this headache syndrome, and cranial MRI is needed in investigating secondary SUNCT in these patients.

**Key Words:** *small cell lung cancer, SUNCT syndrome, magnetic resonance imaging*

Kısa süreli tek taraflı nöraljiform baş ağrısı atakları, konjonktival injeksiyon ve gözde sulanma (SUNCT-Short-lasting unilateral Neuralgiform headache attacks with conjunctival injection and tearing) primer baş ağrılarından nadir bir formudur. SUNCT benzeri sendrom veya sekonder gelişimli SUNCT literatürde tanımlanmıştır. Biz SUNCT sendromlu küçük hücreli akciğer kanseri tanısı olan ilk olguyu sunuyoruz. Hastanın kraniyal manyetik görüntülemesinde sağ temporal kas içinde yerleşimli bir lezyon saptandı. Karbamazepin ile tedaviye orta derecede yanıt verdi. SUNCT benzeri sendrom tanısı, akciğer kanseri olan hastalarda dikkatli bir şekilde değerlendirilmelidir. Kraniyal metastazlar bu baş ağrısı sendromunu tetikleyebilir ve hastalarda sekonder SUNCT araştırılırken kraniyal MRG gereklidir.

**Anahtar Kelimeler:** *küçük hücreli akciğer kanseri, SUNCT sendromu, manyetik rezonans görüntüleme*

SUNCT syndrome (Short-lasting unilateral Neuralgiform headache attacks with Conjunctival Injection and Tearing) is a rare form of strictly unilateral headache with prominent autonomic symptoms (1). This syndrome was first described in 1978 by Sjaastad et al (2). This syndrome is one of the Trigeminal Autonomic Cephalgias (TACs), this form of short-lasting headache is also among the rarest of headache syndromes (3). The clinical picture consists of brief attacks of moderate to severe orbital / periorbital pain accompanied by ipsilateral conjunctival injection, lacrimation, and nasal obstruction or rhinorrhea, the accom-

paniments being rather constant and prominent (4). The character of the pain is a burning sensation, stabbing or electric shock-like, lasting from 5 to 250 seconds. Patients may have up to 30 episodes per hour but usually the frequency is 5-6 per hour (4-6). Diagnostic criteria for SUNCT syndrome was defined in the second edition of the International Classification of Headache Disorders (ICHD-II) (7). SUNCT syndrome is a rare condition that predominates slightly in men (8). In the vast majority of patients, etiology and pathogenesis are unknown. A secondary SUNCT or SUNCT-like syndrome has been described in

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some patients with either intra-axial or extra-axial posterior fossa lesions, mostly vascular disturbances / malformations, and pituitary abnormalities. It is remarkable that most of the lesions involve the posterior fossa or the posterior part of the brain (3, 6, 8, 9).

We present a new case of secondary SUNCT syndrome in a 57 year old male patient with a diagnosis of small-cell lung cancer (SCLC).

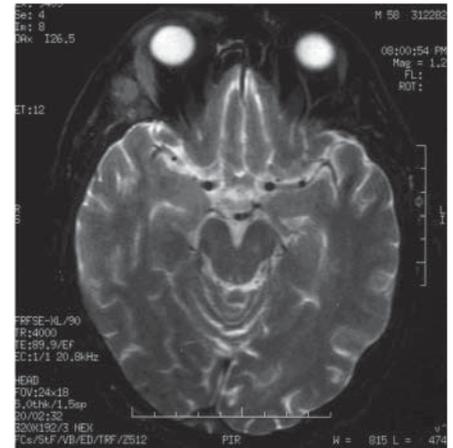
## Case Report

A 57-year-old man with a previous diagnosis of small cell lung cancer, was admitted because of a new history of headache. He was diagnosed with an extensive disease metastatic to the liver and bones six months ago. His cranial computed tomography (CT) scan was normal at baseline. He had an early relapse after six cycles of cisplatin-etoposide chemotherapy with a progression of the primary thoracic tumor. On admission, he described few recent episodes of severe left sided orbital and temporal pain. His pain was strictly unilateral on the left side with a stabbing character. He had an excruciating pain located at the periorbital area with radiation to the fronto-parietal area of the head. This brief paroxysmal pain was accompanied by lacrimation and redness of the ipsilateral eye, and nausea. The usual duration of pain attacks was changing between 60 to 120 seconds. He had 1-2 attack per day. Any well defined triggering movement was described. A cranial CT scan showed any lesions. Thereafter the patient received two cycles of topotecan as a second line therapy. He experienced a mild improvement for this complaint of unilateral headache

with diclofenac sodium, but his pain had worsened over the second line therapy period, and he readmitted with this complaint of headache. He was a 70 pack-years ex-smoker who had quit smoking one year earlier, and he had a history of type 2 diabetes mellitus, controlled with insulin. He had no previous history of migraine or headache, but his mother and his brother had a history of migraine. His neurological and ophthalmologic examinations were normal. A cranial magnetic resonance imaging (MRI) (Figure-1) revealed an increased intensity in the region of the right temporal muscle. A SUNCT-like syndrome was diagnosed, and he was started on carbamazepine 400 mg daily from which there was a moderate benefit, and then the carbamazepine dose was increased to 800 mg daily, nevertheless he had no further improvement. He died due to the progression of his malignancy with hepatic encephalopathy one month after his readmission.

## Discussion

SUNCT syndrome is a rare form of trigeminal autonomic cephalgias, that predominates slightly in men, with a mean age of onset around 50 years. It is characterized by strictly unilateral attacks centered on the orbital or periorbital regions, forehead, and temple. Most attacks are moderate to severe in intensity and burning, stabbing or electrical in character. The usual duration ranges from 5 to 240 seconds, although the reported range of duration is 2 seconds to 20 minutes. Ipsilateral conjunctival injection and lacrimation are present in most, but not all patients (8). The new diagnostic criteria according to the ICHD-II for SUNCT is given



**Figure 1.** Cranial magnetic resonance imaging (MRI) showed an increased intensity in the region of the right temporal muscle marked with a thickened arrow.

in table-1 (7), and they have been met for the diagnosis of the reported patient.

The TACs are characterized by short-lasting headaches with autonomic features, since the differential diagnosis includes mainly these trigeminal-autonomic cephalgias. Short-lasting attacks of unilateral pain are much briefer than those seen in any other TAC and very often accompanied by prominent lacrimation and redness of the ipsilateral eye (7). Most cases of SUNCT syndrome described in the medical literature are primary, but several cases of secondary SUNCT or SUNCT like syndrome have been reported, and they are rather related to posterior fossa and pituitary abnormalities. Causes of symptomatic SUNCT syndrome have included posterior fossa lesions (such as cerebellopontine angle AVMs, brainstem cavernous angiomas, posterior fossa lesions in an HIV/AIDS patient, severe basilar impression causing pontomedullary compression in a patient with osteogenesis imperfecta, craniosynostosis resulting in a foreshortened posterior fossa, pontocerebellar astrocytoma, cerebellopontine angle and fron-

**Table 1.** Diagnostic criteria for SUNCT (Short-lasting unilateral Neuralgiform headache attacks with Conjunctival Injection and Tearing)

- A. At least 20 attacks fulfilling criteria B-D
  - B. Attacks of unilateral orbital, supraorbital or temporal stabbing or pulsating pain lasting 5-240 seconds
  - C. Pain is accompanied by ipsilateral conjunctival injection and lacrimation
  - D. Attacks occur with a frequency from 3 to 200 per day
  - E. Not attributed to another disorder <sup>1</sup>
- <sup>1</sup> History and physical and neurological examinations do not suggest any of the disorders listed in groups 5-12, or history and/or physical and/or neurological examinations do suggest such disorder but it is ruled out by appropriate investigations, or such disorder is present but attacks do not occur for the first time in close temporal relation to the disorder

tal lobe meningiomas, anomalous vertebrobasilar vascular development, brainstem and upper cervical lesions secondary to Devic's syndrome and multiple sclerosis, pituitary tumors (such as a non-functioning pituitary adenoma, prolactinomas, acromegaly) and miscellaneous causes such as a cavernous sinus leiomyosarcoma, orbital cyst, intraorbital metastatic bronchial carcinoid, and an HIV patient with no opportunistic infections and normal brain imaging. A case of a patient with SUNCT syndrome who also was HIV-positive, and had no opportunistic infections with a normal brain imaging has been reported. Even the authors raised the possibility of a causal relationship between the two conditions, an alternative explanation is a coincidental occurrence of the two conditions (8, 11). This case is also addressed to a similar possibility, since we report a patient with SUNCT syndrome who also had a

diagnosis of SCLC. Furthermore, brain MRI showed an increased intensity in the region of the right temporal muscle which may be a triggering factor in this patient with extensive disease. The occurrence of SUNCT syndrome in this case of SCLC may also be coincidental, even though our patient's very severe headache fulfilling the SUNCT diagnostic criteria started after the diagnosis of SCLC, it may be argued that a contralateral temporal muscle lesion should be precautiously considered a possible trigger. However, SCLC is a disease which can have relatively frequent occurrence of paraneoplastic syndromes, and if this special headache entity could be considered as one of them, this may be another possible explanation for this headache syndrome presence in that patient.

SUNCT is differentiated by its predominance of autonomic symptoms from trigeminal neuralgias

which is often responsive to anti-epileptics such as carbamazepine. SUNCT syndrome, unlike chronic or episodic paroxysmal hemicranias, is generally considered refractory to the treatment or difficult to treat, although recent open clinical trials suggest that lamotrigine, gabapentin, topiramate, and intravenous lidocaine have produced beneficial therapeutic responses(1,8-10). Our patient had a mild to moderate response to the treatment with a non-steroidal anti-inflammatory drug and carbamazepine.

In summary, we reported the first case of SUNCT syndrome with a diagnosis of SCLC in the literature. SUNCT-like syndrome diagnosis should be evaluated carefully in patients with previous history of lung cancer. Cranial metastases may be a trigger for this headache syndrome, and cranial MRI is needed in investigating secondary SUNCT patients.

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