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## SUNCT: a new case with some unusual features

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**Abstract** A healthy 22-year-old man complained of primary stabbing headache (PSH) for about two months. The headache recurred after one year and after a month the pain took on the characteristics of short-lasting unilateral neuralgiform headache with conjunctival injection and tearing (SUNCT). This patient shows some unusual features: juvenile onset, miosis during attacks as part of autonomic phenomena and close temporal relationship with stressful events. The

association between PSH and SUNCT may be interpreted as the coexistence of two different headaches or that the PSH is a forerunner to the SUNCT.

**Keywords** Primary stabbing headache • SUNCT

### Introduction

Short-lasting unilateral neuralgiform headache with conjunctival injection and tearing (SUNCT) is a rare headache that was described first by Sjaastad and co-workers in 1978 [1]. In the typical form several attacks of moderate or severe short-lasting orbital, periorbital or temporal stabbing or pulsating pain lasting 5–240 s with lacrimation, conjunctival injection and rhinorrhoea or nasal obstruction occur in the same day [2]. But atypical cases regarding the temporal pattern, localisation of pain, accompanying phenomena or precipitating mechanisms, and secondary cases, have been described [3, 4], indicating that the entire spectrum of the disease is not well-known. So the description of a new case with some unusual characteristics will extend our knowledge of the full spectrum of this disease.

### Case report

A healthy man without significant familial or personal history of headache presented at the age of 22 years with a stabbing pain of moderate intensity in the occipital region sometimes extending to the left temple. The pain occurred daily once or twice without precipitating events, lasting one or two seconds and usually occurring as a single episode but on occasion as volleys of pain and without autonomic symptoms. After a couple of months his physician prescribed clonazepam 0.5 mg t.i.d. and after a few days the pain stopped.

After one year of respite and during a stressful period of family problems the pain recurred with the same characteristics, but clonazepam 1 mg t.i.d. was ineffective and was stopped after two weeks. A month later the features of pain changed: it started abruptly in the left temple spread-

ing to the left orbital and periorbital areas. It was sharp, sometimes mild but usually of moderate intensity and accompanied by ipsilateral conjunctival injection, lacrimation, miosis and rhinorrhoea. The miosis lasted about one minute whereas the other associated symptoms lasted two or three minutes. The duration of the pain was 5–10 s with frequency ranging from 1–2 attacks per day to 7–8 a day; rarely did the pain stop for an entire day. The pain occurred in the daytime without a definite temporal pattern but never occurred at night. Physical and neurological examination, routine blood tests and blink reflex were normal. Cranial magnetic resonance imaging (MRI) was normal except for a small colloid cyst in the left maxillary sinus. The patient refused any therapy and the pain stopped spontaneously after three months, shortly after the resolution of his family problems. At the last follow-up at the age of 28 years he was still headache-free.

## Discussion

In the diagnosis of short-lasting primary headaches, primary stabbing headache (PSH), SUNCT, episodic paroxysmal hemicrania (EPH) and first division trigeminal neuralgia must be taken into account. The length of pain is greater in EPH, ranging from 2 to 30 min [2]. The accompanying autonomic phenomena are pronounced both in SUNCT and EPH whereas in the first division trigeminal neuralgia they are missing and occur in the later stage or in atypical cases [5]. Moreover the blink reflex was normal in our patient whereas it is often abnormal in trigeminal pain [6]. Some characteristic traits of PSH and SUNCT are summarised in Table 1: they differ mainly because PSH is not associated with autonomic phenomena while SUNCT is. The presenting symptoms of this patient were those of PSH [2] but with some unusual features. Indeed this headache is more common in females: female/male ratio is 1.49 in the Vaga study [7]; the age of onset is usually older than in this patient:  $47.1 \pm 14.5$  years

in the Pareja's and co-workers series [8] but about 28 years in the Vaga study [7]; and most patients report the coexistence of another headache, usually migraine or cluster headache [8]. In the PSH pain usually occurs in the distribution of the first division of the trigeminal nerve, but sometimes (as in this case) in parietal or occipital areas [8]. The site of pain is the same as in occipital neuralgia but this diagnosis is ruled out by the absence of tenderness in the occipital zones. The pain usually lasts seconds but cases with longer duration up to two hours have been described [8]; it is possible that a rapid succession of stabbing pains can be perceived as pain lasting for a longer time. Unfortunately a trial with indomethacin was not performed in this patient. The disappearance of the pain after clonazepam 0.5 mg t.i.d. may have been a coincidence; indeed the second trial with clonazepam 1 mg t.i.d. was ineffective. But because the lack of effective therapies both in PSH and SUNCT (with the possible exception of indomethacin in PSH and lamotrigine and topiramate in SUNCT) and the effectiveness of clonazepam in some cases of trigeminal neuralgia [9], other attempts may be justified.

After one year of respite the same symptoms reappeared but after a few weeks the pain took on the characteristics of probable SUNCT (point 3.4.3 of ICHD-II). However, some unusual features are present. The age at onset usually is older than in this patient:  $50.7 \pm 14.8$  years in the Pareja and Sjaastad review [4], but juvenile and even childhood onset have been described [10, 11]. In the IHS Classification [2] the frequency of attacks is from 3 to 200 daily, but in their review Pareja and Sjaastad [4] report a greater variability: from less than 1 attack daily to more than 30 attacks per hour and the usual frequency ranged from 1–2 to 20–80 attacks per day. Among the autonomic phenomena, miosis is noteworthy: in Pareja and Sjaastad's review [4] it was found only in one patient and recently another case with permanent miosis has been described [12]. In our patient miosis was first observed by his wife and after by himself and despite the fact that the miosis was never observed by a physician, the observation

**Table 1** Clinical characteristics of typical PSH and SUNCT. The diagnostic criteria are summarised in items 1–5 (2); the other features are drawn from the literature

	PSH	SUNCT
Site	Mainly first division V nerve	Orbital, supraorbital, temporal
Type	Stabbing	Stabbing or pulsating
Duration	Up to a few seconds	5–240 s
Frequency	From 1 per day to many	3–200 daily
Autonomic features	Absent	Intense
Age	28–47 years	23–77 years
Sex predominance	Female	Male
Response to indomethacin	Possibly present	Absent

seems reliable. An MRA was not performed in order to rule out a dissection or an aneurysm, but this hypothesis is unlikely because of the intermittence of miosis and the following disappearance. So, in agreement with other cases described, miosis must be accepted as a rare autonomic phenomenon of SUNCT. Unfortunately it was impossible to establish if miosis and other autonomic symptoms started at the same time as the pain or if they were delayed; surely the miosis was never observed outside the attacks.

The close relationship with stressful events is another unusual feature: PSH usually seems to be spontaneous and rarely are precipitating mechanisms, including emotional stress, recognised [13]. In the Pareja and Sjaastad review only in three patients was there a close temporal relationship between emotional stress and SUNCT [4]. Atypical features are quite common in the few symptomatic cases

[14]; MRI in this patient demonstrated only a colloid cyst of the maxillary sinus, presumably by chance and thus this is not a symptomatic case.

A case of SUNCT from transformation of trigeminal neuralgia has been described [15] and in another patient SUNCT disappeared after decompression of the trigeminal nerve [16] and PSH is often associated with migraine and cluster headache [2] and not with other trigeminal autonomic cephalalgias (TACs). To the best of my knowledge the association between PSH and SUNCT has not been described.

In conclusion, this patient suffered from SUNCT with some unusual features: juvenile onset, appearance of miosis during attacks and close temporal relationship with stressful events. It is possible that SUNCT was preceded by PSH; another possible explanation is that PSH was an incomplete form of SUNCT.

## References

1. Sjaastad O, Russel D, Horven I, Bunaes U (1978) Multiple neuralgiform headache attacks associated with conjunctival injection and appearing in clusters. *Proc Scand Migraine Soc* 31
2. Headache Classification Subcommittee of the International Headache Society (2004) The International Classification of Headache Disorders, 2nd edn. *Cephalalgia* 24[Suppl 1]:9–160
3. Pareja JA, Joubert J, Sjaastad O (1996) SUNCT syndrome. Atypical temporal patterns. *Headache* 36:108–110
4. Pareja JA, Sjaastad O (1997) SUNCT syndrome. A clinical review. *Headache* 37:195–202
5. Sjaastad O, Pareja JA, Zukerman E, Jansen J, Kruszewski P (1997) Trigeminal neuralgia. Clinical manifestation of first division involvement. *Headache* 37:346–357
6. Cruccu G, Leandri M, Feliciani M, Manfredi M (1990) Idiopathic and symptomatic trigeminal pain. *J Neurol Neurosurg Psychiatry* 53:1034–1042
7. Sjaastad O, Pettersen H, Bakketeig LS (2002) The Vaga study; epidemiology of headache: the prevalence of ultra-short paroxysms. *Cephalalgia* 21:207–215
8. Pareja JA, Ruiz J, de Isla C, Al-Sabbah H, Espejo J (1996) Idiopathic stabbing headache (jabs and jolts syndrome). *Cephalalgia* 16:93–96
9. Sidebottom A, Maxwell S (1995) The medical and surgical management of trigeminal neuralgia. *J Clin Pharm Ther* 20:31–35
10. Bouhassira D, Attal N, Esteve M, Chauvin M (1994) SUNCT syndrome. A case of transformation from trigeminal neuralgia? *Cephalalgia* 14:168–170
11. Sprenger T, Valet M, Platzer S, Pfaffenrath V, Steude U, Tolle TR (2005) SUNCT: bilateral hypothalamic activation during headache attacks and resolving of symptoms after trigeminal decompression. *Pain* 113:422–426
12. D'Andrea G, Granella F (2001) SUNCT syndrome: the first case in childhood. *Cephalalgia* 21:701–702
13. Blatter T, Capone Mori A, Boltshauser E, Bassetti C (2003) Symptomatic SUNCT in an eleven-year-old girl. *Neurology* 60:2012–2013
14. Prakash KM, Lo YL (2004) SUNCT syndrome in association with persistent Horner Syndrome in a Chinese patient. *Headache* 44:256–258
15. Pareja JA, Kruszewski P, Caminero AB (1999) SUNCT syndrome versus idiopathic stabbing headache (jabs and jolts syndrome). *Cephalalgia* [Suppl 25]:46–48
16. Trucco M, Mainardi F, Maggioni F, Badino R, Zanchin G (2004) Chronic paroxysmal hemicrania, hemicrania continua and SUNCT syndrome in association with other pathologies: a review. *Cephalalgia* 24:173–184