Short-Lasting Unilateral Neuralgiform Headache Attacks with Cranial Autonomic Symptoms (SUNA)

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Abstract

Short-lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms (SUNA) is a rare primary headache syndrome. The diagnostic criteria include attacks of unilateral orbital, supraorbital or temporal stabbing pain accompanied by one of the following: conjunctival injection and/or tearing, nasal congestion and/or rhinorrhea, and eyelid edema. The duration of pain is 2 seconds to 10 minutes, and the frequency of attacks is described as once a day or more. The etiology and pathology of SUNA has yet to be documented. We report an 18-year-old man with SUNA. Lomerizine hydrochloride which is used as a preventive medicine for migraine, improved his headaches.

Key words: short-lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms (SUNA), short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT), lomerizine hydrochloride, migraine

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Introduction

The Second Edition of the *International Classification of Headache Disorders* (ICHD-II) published in 2004 began to use the term “trigeminal autonomic cephalalgias (TACs)”. TACs comprehensively refers to unilateral headache of short duration and headache that simultaneously exhibits cranial autonomic symptoms such as lacrimation and rhinorrhea, which had previously been reported as cluster headache, paroxysmal hemicrania, short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT). Furthermore, the possibility that SUNCT is a subtype of a broader disease concept, i.e., short-lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms (SUNA), was presented in the appendix of the ICHD-II. We herein report a case that was considered to correspond to SUNA.

Case Report

The patient was an 18-year-old man with pain from a

deep area of the left eye to the temporal region as the primary symptom. In March 2007, attacks of pulsating and stabbing pain in the left orbital to temporal region with a sudden onset and duration of about 5 minutes began to occur 4-5 times almost daily. Since the pain became worse with head motion, the patient would let it pass by resting his head on the desk if it occurred during classes. It did not occur more frequently during any particular period of the day. Photophobia and phonophobia were noted during attacks, but there was no nausea, vomiting, or lacrimation: nasal congestion was the only autonomic symptom. Also, the superficial temporal artery in the left temple became dilated and was tender during attacks. The patient consulted a local neurosurgeon in December of the same year, and was found to have a congenital brain anomaly by brain magnetic resonance imaging (MRI), but no abnormality was noted on electroencephalography (EEG). Subsequently, the patient was prescribed antiepileptics such as sodium valproate (VPA) and carbamazepine (CBZ) and analgesics (loxoprofen sodium, diclofenac sodium), but there was no change in the symptoms. The patient also consulted ophthalmologists and otorhinologists, but no particular abnormality including si-

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Figure 1. Brain MRI shows cortical dysplasia (arrows). The signal level of this lesion is equal to that of the surrounding cortex on T1- and T2-weighted images but it is higher than that in part of the cortex on fluid-attenuated inversion recovery (FLAIR) images.

nusitis was found. The patient visited the outpatient clinic of our department in late January 2008. He had no history of headache or convulsion from infancy and no familial history of migraine.

The patient was 170 cm tall and weighed 70 kg. The examination was performed in an attack-free period, no swelling or reddening of the scalp arteries including the superficial temporal artery was noted, and no herpetic rash was observed on his face or head. No particular neurological abnormality was noted. Blood tests showed no abnormality in peripheral blood or general biochemical analyses, no sign of inflammatory reaction, and no abnormality related to collagen disease or in hypothalamohypophyseal hormones such as prolactin or thyroid hormones. On EEG, there was no particular finding including epileptic abnormal waves. Brain MRI (Fig. 1) revealed cortical dysplasia with intrusion of the cortex deep into the precentral sulcus nearly to the lateral ventricle, and the signal level of this lesion was equal to that of the surrounding cortex on T1- and T2-weighted images but it was higher than that in part of the cortex on fluid-attenuated inversion recovery (FLAIR) images. No sinusitis, pituitary tumor, nor lesion of the posterior fossa was detected, and MR angiograms were normal.

CBZ was administered at doses of up to 400 mg/day, but no marked change was noted. Indomethacin farnesil was administered at dose of 400 mg/day (a maximum dose of 75 mg/day as indomethacin) with chronic paroxysmal hemicra-

nia (CPH) in mind, but there was no response. Thereafter, the patient did not visit our department for about six months until September 2008, when he consulted us due to the persistence of symptoms. The administration of lomerizine hydrochloride was started at a routine dose of 10 mg/day. From about 10 days after the beginning of treatment, the frequency of headache attacks gradually decreased to 1-2 times a week, their duration was shortened to 1-2 minutes each time, and pain was alleviated to about half of the previous level. About 6 months after that, the symptoms had almost disappeared.

Discussion

Diseases that cause short-lasting paroxysmal headache in the unilateral orbital-temporal region, as in the present patient, can be differentiated. First, symptomatically, there have been reports of patients who developed headache attacks in the trigeminal nerve region due to sinusitis, pituitary tumor, tumor of the posterior fossa, and vascular lesions (2-4). In some patients, such headache attacks completely fulfill the diagnostic criteria of primary headache such as cluster headache, and caution not to overlook such organic disorders has been stressed. In the present patient, while no such lesion was detected, cortical dysplasia was noted. This lesion may have been a focus causing headache as in epileptic attacks, but no abnormality was observed on

Table 1. A Summary of Characteristics of TAC, Trigeminal Neuralgia and the Present Patient

	SUNCT (SUNA)	CH	CPH	TN	present case
Gender	man>woman	man>woman	man<woman	man<woman	man
Type of pain	stabbing, pulsating	boring pulsating	stabbing boring	stabbing	stabbing and pulsating
Duration of attacks	5~240sec (2sec~10min)	15~180min	2~30min	~2min	5min
Frequency of attacks	3~200/day (>1/day)	1/2days~8/day	>5/day	few~many	4~5/day
Autonomic symptoms	+	+	+	—	nasal congestion

CH = cluster headache ; CPH = chronic paroxysmal hemicrania ; TN = trigeminal neuralgia

EEG. In temporal arteritis, the superficial temporal artery is palpated and tender, as in our patient, but the disease was excluded from differential diagnoses for our patient, because it is frequently observed in elderly people, headache is often sustained and blunt, and inflammatory reaction is always positive on blood tests.

Table 1 compares the clinical characteristics of TACs and trigeminal neuralgia and those in our patient. First, in SUNCT, a unilateral stabbing or pulsating pain in the orbital, supraorbital, or temporal region continues for 5-240 seconds, and is accompanied by ipsilateral conjunctival injection and lacrimation. The frequency of attacks is 3-200 times/day, and the occurrence of such headache attacks at least 20 times/day is a diagnostic criterion. SUNA, on the other hand, is diagnosed when one of the following occurs: 1) conjunctival injection and/or tearing, 2) nasal congestion and/or 3) rhinorrhea; and eyelid edema is present among autonomic symptoms. The duration of pain is 2 seconds to 10 minutes, and the frequency of attacks is described as once a day or more on at least half the days during the attack period (Table 2). In the present patient, no conjunctival injection or lacrimation was noted, and only nasal congestion was observed. Therefore, the condition is unlikely to have been SUNCT, but fulfills the diagnostic criteria of SUNA (chronic type) including the duration and frequency of attacks. Concerning other differential diagnoses, in cluster headache, the duration of attacks is 15-180 minutes, which is longer than in our patient. In CPH, the duration of attacks is 2-30 minutes, and their frequency is 5 times/day or higher, as in our patient, but indomethacin, which is very effective in CPH, had no effect in our patient. Trigeminal neuralgia usually causes immediate pain, and a single attack of headache does not persist for as long as 5 minutes, as it did in our patient. Also, no inducing factor or trigger region, which is often present in trigeminal neuralgia, was noted in our patient.

Concerning SUNCT/SUNA, no effective treatment is generally considered to be available, but there have recently been sporadic case reports indicating the effectiveness of antiepileptics such as CBZ and lamotrigine (5, 6). In our patient, CBZ, VPA, and indomethacin were ineffective, and lomerizine hydrochloride was effective. Lomerizine hydrochloride, a calcium channel blocker, is currently used only in Japan as a preventive medicine for migraine headaches. It has been confirmed to control migraine headaches by various actions including inhibition of the contraction of cerebral blood vessels, which is the first step in migraine headaches, suppression of cortical spreading depression, suppression of a neurogenic inflammation due to trigeminal stimulation, and suppression of abnormal platelet functions (7). Regarding its effects on headaches other than migraines, there has been a report that lomerizine hydrochloride shows some effects on cluster headache (7), but the results were insufficient, and there are no reports indicating the effectiveness of lomerizine hydrochloride for the treatment of other types of headaches, possibly because it is marketed only in Japan.

The condition of our patient is considered to exhibit features resembling those of SUNA among primary headaches according to the diagnostic criteria. However, SUNA is a rare disease, so there are few reports, and its pathology has not yet been documented. In a study using functional MRI, Cohen et al reported that uni- or bilateral activation of the hypothalamus was noted during headache attacks in patients with SUNCT but that no activation of the same regions was noted in those with SUNA (6). There has also been a report that there was a history or familial history of migraine in 49% of patients with SUNCT and in 67% of those with SUNA. In addition, it has been reported that agitation was observed in about 60% of the patients with SUNCT or SUNA during headache attacks, but that there was a history of migraine in many patients who showed the exacerbation

Table 2. Diagnostic Criteria of SUNCT and SUNA (ICHD-II)

Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT)

3.3 Diagnostic criteria

- A. At least 20 attacks fulfilling criteria B–E
- B. Attacks of unilateral, orbital, supraorbital or temporal stabbing or pulsating pain last 5–240 s
- 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

Short-lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms (SUNA)

A 3.3 Diagnostic criteria

- A. At least 20 attacks fulfilling criteria B–E
- B. Attacks of unilateral orbital, supraorbital or temporal stabbing pain lasting from 2 s to 10 min
- C. Pain is accompanied by one of
 - 1. conjunctival injection and/or tearing
 - 2. nasal congestion and/or rhinorrhea
 - 3. eyelid edema

D. Attacks occur with a frequency of ≥ 1 per day for more than half the time

E. Not attributed to another disorder

A 3.3.1 Episodic SUNA

Description: SUNA attacks occurring for 7 days to 1 year with pain-free intervals longer than 1 month

A 3.3.2 Chronic SUNA

Description: At least 2 attack periods last 7 days to 1 year separated by remission periods of <1 month (untreated)

of headaches during head movement (8). Dilation of the ipsilateral superficial temporal artery was noted during attacks in our patient. Some patients with migraine or cluster headache are known to show dilation or pulsation of the scalp vessels, but no such report was found in patients with

SUNCT or SUNA. These observations suggest that migraine is involved in the pathology of SUNA in some patients. This is considered to possibly explain the effectiveness of lomerizine hydrochloride, a treatment for migraine, in some patients exhibiting clinical symptoms of SUNA.

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