Dear Sir,

The term SUNHA as a diagnostic category was introduced according to the revised criteria of the International Classification of Headache Disorders (ICHD-3 beta) 2013, and it comprises the headache subtypes SUNCT and SUNA. Both are rare disorders and are associated with cranial autonomic features. The diagnostic features of both are overlapping, with both conjunctival ingestion and tearing being necessary for SUNCT while only one of the two autonomic features suffices for SUNA. The reports of ipsilateral autonomic features in trigeminal neuralgia (TN) date back to 1914. There is a considerable diagnostic overlap between SUNHA and TN occurring in the first division of trigeminal nerve (V1) when the clinical differentiation becomes particularly difficult.

Here we describe an interesting case that presented to us with features suggestive of both TN and SUNCT and improved with symptomatic treatment. A 24-year-old gentleman came to our outpatient department (OPD) with a one-month history of left-sided acute onset periorbital and facial pain headache. It was a lancinating pain triggered by touching the left-side of the forehead while applying soap or wiping sweat, or even chewing food associated with tonic spasm of the left-side of the face. The frequency of such attacks was 15–20 times per day, lasting for 6–10 seconds. Along with this, he also complained of episodes of a second type of pain over the similar site and side, occurring spontaneously, each episode lasting for 2–60 seconds with intense redness of eyes, and severe lacrimation, which remitted on its own. There was no history of any hearing impairment, facial numbness, rash over face, weakness or ataxia of limbs, drooping of eyelids, diplopia, no difficulty in speech or deglutition, impaired consciousness, and features of raised intracranial pressure. There was no past history suggestive of tuberculosis, diabetes, any trauma, any systemic illness, or any such episodes in the past. On examination during the interictal period, his mental functions were within normal limits. All the cranial nerves were found normal on examination, the corneal reflex was normal. Motor, sensory, cerebellar, and gait were normal. The patient had multiple attacks during examination in the outpatient department. During a few attacks, which were spontaneous, he had tonic spasms in the succession of the left side of the face with reddening of the left eye and tearing at the end of the episode and patient complaining of severe pain. He also had a few episodes triggered by touching his left forehead but without any autonomic features. His pupillary examination and extraocular movements were normal during attacks.

He was given a trial of oxcarbazepine in escalating doses with mild improvement. In view of his young age and short history, he was advised neuroimaging. MRI revealed an ill-defined lobulated broad-based extraxial cystic lesion in the left cerebellopontine (CP) angle. It was heterogeneously hypointense on T1W [Figure 1], heterogeneously hyperintense on T2W [Figure 2]. MRI feature was suggestive of epidermoid.

Video Electroencephalogram (VEEG) was done to rule out the focal motor seizure. It was done in the inter-attack periods, and then attacks were induced by touch during the electroencephalogram (EEG). No epileptiform discharges could be seen, and EEG was normal.

He was operated on for the cerebellopontine angle tumor, and the excision was done via retromastoid suboccipital craniectomy approach. The tumor was extra-axial, fleshy white, avascular extending up to midbrain surrounding 5th, 6th, 7th, 8th, and other lower CN. Gross total excision was achieved. On histopathological examinations, a cyst lined by flattened squamous epithelium with extensive keratinized material suggestive of epidermoid was found. He has been in remission after surgical intervention without any drugs. Thus our patient was probably suffering from attacks of trigeminal neuralgia with superimposed attacks of SUNCT in view of associated intense autonomic features. In a clinical setting, it may be extremely difficult to accurately differentiate between SUNAH and TN especially if the latter occurs in the first division of the trigeminal nerve. Autonomic features have been reported in approximately 30%–60% of patients of TN in various series, thus making a clear-cut distinction between the two entities.

Because of a considerable phenotypic overlap, some authors consider them as a single disease entity. The presence of no or minimal cranial autonomic features, associated with the presence of a clear-cut refractory period to the presence of triggers, could be taken as a surrogate clinical marker for TN since 95% of SUNCT cases lack a refractory period.

Figure 1: It was heterogeneously hypointense on T1W
We suspected a secondary cause in our patient because of the younger age of onset and shorter duration of symptoms. Epidermoid presenting as trigeminal neuralgia is a rare presentation and has been described previously in single case reports or case series.\(^6\) The probable cause of pain in TN in the case of epidermoid is attributed to mechanical compression and/or inflammatory response to the tumor’s contents since their surgical excision has excellent relief of pain.

The proposed pathophysiology for pain in SUNCT is the activation of the posterior hypothalamus by the trigeminohypothalamic tract (THT), which in turn modulates the activity of the trigeminal nucleus caudalis, which mediates autonomic dysfunction.\(^8\) Secondary SUNCT is known to occur due to various neoplastic causes, vascular disease, trauma, infection, inflammation, or congenital malformation.\(^9\) However, our case was unusual in that it had TN as an isolated feature which has been described in only two cases previously,\(^10\) and had overlapping features of both TN and SNUCT, which, to the best of our knowledge, has not been reported previously. He underwent surgical resection with complete resolution of pain post-surgery and is now doing fine, nine months post surgery without drugs. Thus in patients with TAC, a detailed clinical history will help us in categorizing patients into the headache subtype. The possibility of two headache subtypes should be considered based on definite history and clinical diagnostic criteria. Neuroimaging should be done in all atypical cases to rule out secondary causes. Symptomatic treatment is rewarding in most cases.

**Abbreviations**

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

SUNA = Short-lasting unilateral neuralgiform headache attacks

TN = Trigeminal Neuralgia

CP angle = Cerebellopontine angle V1: first division of the trigeminal nerve

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**References**

1. Headache Classification Committee of the International Headache Society (IHS). The international classification of headache disorders, 3rd edition (beta version). Cephalalgia 2013;33:629-808.

2. Patrick HT. The symptomatology of trifacial neuralgia. JAMA 1914;62:1519-25.

3. Simms HN, Honey CR. The importance of autonomic symptoms in trigeminal neuralgia. J Neurosurg 2001;115:210-6.

4. Leone M, Mea E, Genco S, Bussone G. Coexistence of TACS and trigeminal neuralgia: Pathophysiological conjectures. Headache 2006;46:1565-70.

5. Cohen AS, Matharu MS, Goadsby PJ. Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) or cranial autonomic features (SUNA)—A prospective clinical study of SUNCT and SUNA. Brain 2006;129:2746-6.

6. Mohanty A, Venkatrama SK, Rao BR, Chandramouli BA, Jayakumar PN, Das BS, et al. Experience with cerebellopontine angle epidermoids. Neurosurgery 1997;40:24-9.

7. Meng L, Yuguang L, Feng L, Wandong S, Shugan Z, Chengyuan W, et al. Cerebellopontine angle epidermoids presenting with trigeminal neuralgia. J Clin Neurosci 2005;12:784-6.

8. Malick A, Strassman AM, Burstein R. Trigeminohypothalamic and reticulohypothalamic tract neurons in the upper cervical spinal cord and caudal medulla of the rat. J Neurophysiol 2000;84:2078-112.

9. Cao Y, Yang F, Dong Z, Huang X, Cao B, Yu S. Secondary short-lasting unilateral neuralgiform headache with conjunctival injection and tearing: A new case and a literature review. J Clin Neurol 2018;14:433-43.

10. Hung LC, Wu CS, Lin CK, Fang WK, Hsa YC. Epidermoid Cyst presenting as isolated trigeminal neuralgia-Two case. Acta Neurol Taiwan 2013;22:133-7.

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