CASE REPORT

Not just a usual headache: Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing

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SUMMARY

An elderly lady was diagnosed with short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) in a general practice outpatient setting and treated successfully with lamotrigine. The diagnosis of SUNCT is made clinically but the treatment for SUNCT is very specific and it does not respond to treatment for usual headaches. SUNCT is severe and disabling, and we hope to raise the awareness of the non-neurologist practitioner to recognize the condition to provide the best care to patients. Although SUNCT is rare, it deserves attention.

INTRODUCTION

Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) is a rare but disabling primary headache which is grouped under the trigeminal autonomic cephalalgias (TAC). The diagnosis is entirely clinical but frequently misdiagnosed and mismanaged due to a lack of familiarity. We are reporting a patient with SUNCT who was treated successfully with oral lamotrigine in a primary care outpatient setting. We aim to raise the practitioner's awareness of the disease and share our outpatient treatment experience of a patient with SUNCT syndrome during the SARS-CoV-2 pandemic period.

CASE REPORT

A 68-year-old Chinese lady with a background history of controlled hypertension, type 2 diabetes mellitus, atrial fibrillation, hyperlipidemia, and non-debilitating cerebrovascular accident suffered from recurrent episodes of severe right-sided headache for 3 weeks before being seen at our General Practice (GP) clinic. Her medications included rivaroxaban 15mg once daily, linagliptin 5mg once daily, gliclazide MR 60 mg once daily, bisoprolol 5mg once daily, and atorvastatin 20 mg on nocte.

She described the pain as right orbital discomfort which rapidly escalated to maximum intensity within a few seconds to become sharp and severe lancinating pain over the right orbital and supraorbital regions. The pain was scored 10/10 on the visual analogue scale. Each episode was brief, lasted less than 2 minutes, went away and recur shortly after without treatment. The number of piercing and stabbing pain attacks was numerous (more than 20 times per day) and persisted daily without fail. Attacks occurred both during the day as well as at night and she was awake from sleep with pain. Each painful episode was associated with tearing of the right eye and right eyelid swelling. Rhinorrhea occurred on few occasions when the symptoms were severe. The attacks were triggered whenever she yawned. There was no photophobia, phonophobia, nausea, or vomiting. She denied any history suggestive of rash indicative of herpes zoster over the head or facial region. She denied any history of migraine or previous significant headaches. She was a housewife and denied any stress. The pain was debilitating and stressful which affected her activities of daily living.

The clinical examinations revealed a tearing lady with oedematous right upper eyelid and right-sided partial ptosis (Figure 1a). Other relevant negative clinical findings included normal visual acuity, no scars suggestive of recent shingles, and no other neurological deficit.

She had in-patient as well as out-patient care by an ophthalmologist, medical specialist, and even an acupuncturist. She underwent investigations where blood panels, plain CT brain, MRI brain, and MRA all did not reveal any causative reasons for her headache. Before she visited us, she had tried etoricoxib 90mg once daily, amitriptyline 25mg thrice daily, and pregabalin 75mg on nocte without any pain relief.

After careful history taking, physical examination and review of her previous investigations, a clinical diagnosis of SUNCT was made. We offered a referral to a neurologist for inpatient intravenous (IV) lidocaine; however, the patient refused, given our country was at the peak of SARS-CoV-2 endemic at that time, and she needed to travel interstate to go to the nearest specialist centre where IV lidocaine is available. Oral lamotrigine 25 mg once daily was started and the dosage was titrated upwards weekly to once daily. All other analgesics were discontinued. The patient was made aware of the possible adverse effects of lamotrigine, including Stevens Johnson's syndrome. Lamotrigine was effective. She noticed significant improvement and when the total daily dosage of lamotrigine reached 75 mg, the attacks were suppressed. At three weeks, her eyelid swelling had almost totally resolved (Figure 1b). The dosage was then maintained at 50 mg twice daily. Three months after the initial attack, she was pain-free except for a little dysesthesia over the right

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Table I: Diagnostic criteria for SUNCT/SUNA based on ICHD-3

Diagnostic criteria* for SUNCT/SUNA:
A. At least 20 attacks fulfilling criteria B–D
B. Moderate or severe unilateral head pain, with orbital, supraorbital, temporal and/or other trigeminal distribution, lasting for 1–600 seconds and occurring as single stabs, series of stabs or in a saw-tooth pattern
C. At least one of the following five cranial autonomic symptoms or signs, ipsilateral to the pain: 1. conjunctival injection and/or lacrimation 2. nasal congestion and/or rhinorrhoea 3. eyelid oedema 4. forehead and facial sweating 5. miosis and/or ptosis
D. Occurring with a frequency of at least one a day
E. Not better accounted for by another ICHD-3 diagnosis.
SUNCT: Above diagnostic criteria* plus both of the following, ipsilateral to the pain: 1. conjunctival injection 2. lacrimation (tearing).
SUNA: Above diagnostic criteria* plus not more than one of the following, ipsilateral to the pain: 1. conjunctival injection 2. lacrimation (tearing).

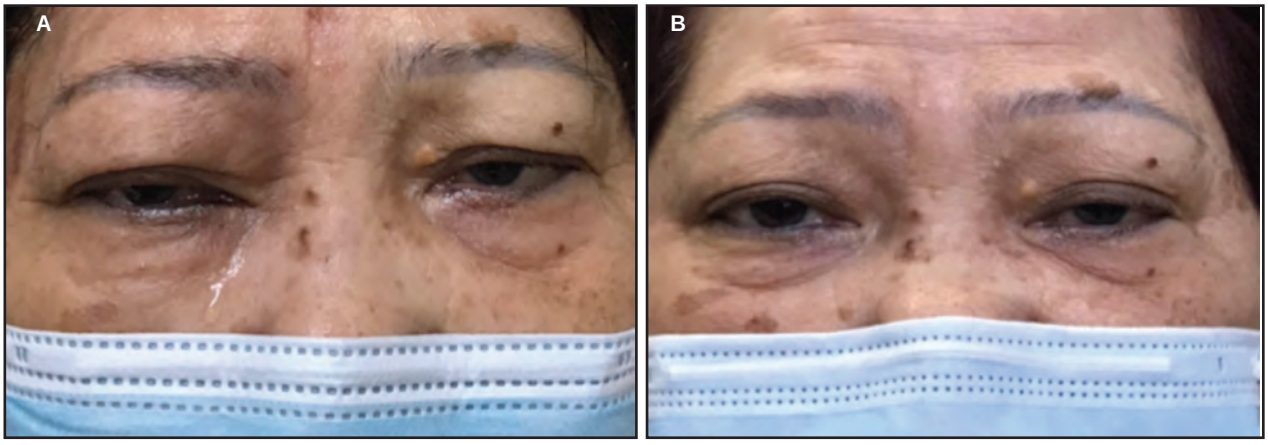

Fig. 1: Patients eyelid lesions. (a) The appearance of the oedematous right upper eyelid, ptosis and tearing of the patient's right eye during the presentation. (b) The appearance after resolution of the eye signs after 3 weeks of treatment.
*Patient wearing mask as the standard operating procedure during COVID-19 endemic.

periorbital region. At the end of the fourth month, she stopped the medication on her own as she had no further attacks. Follow-up reviews for up to 1 year revealed that she remained well.

DISCUSSION

SUNCT is grouped under Trigeminal Autonomic Cephalgia (TAC) based on the International Classification of Headache Disorders 3rd edition (ICHD-3). The other three members are cluster headache, paroxysmal hemicranias, and hemicrania continua.[1]

SUNCT is rare and the awareness of the diagnosis is low. William et al.[2] estimated a prevalence of SUNCT/SUNA at 6.6 per 100,000 and a conservative annual incidence of 1.2 per 100,000. It is characterised by a sudden brief attack of severe unilateral pain in the trigeminal nerve distribution (most commonly 1st division) accompanied by ipsilateral cranial autonomic symptoms. The details of the features are tabulated in Table I based on ICDH-3. Most patients described SUNCT as the most painful condition they had ever experienced.[2,3,4]

The pathophysiology behind SUNCT is not fully understood. However, the principal structures involved in the pathogenesis of SUNCT are the hypothalamus, trigeminovascular system, and cranial autonomic system.[5] Given the increased prevalence of symptomatic causes in patients presenting with symptoms suggestive of SUNCT, a narrative review suggested that all patients undergo a detailed brain MRI with dedicated views of the pituitary, cavernous sinus, and trigeminal nerve.[6] At the moment, the diagnosis of SUNCT is primarily clinical and depends on a good history and clinical examination.[6]

Because of the rarity of the disorder, most of the treatment is based on observational studies, case series, and case reports. Lately, a single-arm meta-analysis was published using the available data and derived a treatment algorithm. This latest evidence suggested intravenous lidocaine as the treatment of choice for transitional therapy and oral lamotrigine as the first-line agent for preventive therapy.[7] Other options are carbamazepine, oxcarbazepine and topiramate.[7] Surgical treatment is reserved for patients who are refractory to medical management.[6] For our patient, with oral Lamotrigine, her pain dramatically improved. It is to our

surprise as it was recommended as preventive therapy rather than abortive therapy. We believe it is an important finding for patients especially those with no access to IV lidocaine.

CONCLUSION

SUNCT is a distinct headache that can be so severe as compared to childbirth, fractures, and kidney stones. However, the diagnosis is often delayed due to poor recognition. The diagnosis is clinical, and investigations are required if secondary causes are suspected. The disorder is eminently treatable but treatment options are highly selective and differ from the common primary headache. We reported a case with successful outpatient treatment using oral lamotrigine. We believe it is an important finding especially when IV lidocaine is not accessible. We hope that non-neurologist practitioners are made aware of this devastating headache that would not respond to typical headache treatment.

ACKNOWLEDGEMENT

None.

INFORMED CONSENT

Informed consent for usage of photo and for publication was obtained from the patient.

CONFLICT OF INTEREST

None to declare.

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