

# SUNCT syndrome in a child: a rare cause of paroxysmal headache

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**S**UNCT (short-lasting unilateral neuralgiform headache with conjunctival injection and tearing) syndrome is a rare type of headache, first described in 1978 by Sjaastad.<sup>1</sup> The syndrome is characterized by mild-to-severe burning, stabbing or electrical orbital/periorbital headache accompanied by autonomic signs. Increased intraocular pressure and eyelid edema occur on the symptomatic side in the course of headache attacks. Two to 40 pain attacks, each lasting for 2 to 60 minutes, may occur daily. Most SUNCT cases have been observed in men at a mean age of 50 years.<sup>2</sup> In this report, we describe a rare case of a 10-year-old child who was diagnosed with this syndrome.

## CASE

A 10-year-old male presented with a 4-year history of headache. The patient also complained of shooting pain in the eye bulb accompanied by swelling, rash, and falling of the eyelid on the symptomatic side. Pain was accompanied by photophobia, but was not aggravated by nausea, vomiting or physical activity. Prior to presentation these symptoms had occurred 6 times, lasting for 5 to 10 minutes each time with a frequency of 3 to 4 times per day for about 10 to 15 days. The patient received treatment for sinusitis each of the three times that he had been hospitalized in the past for these complaints. He had been followed up for asthma and his mother had migraine-type headaches. His physical examination was normal including a completely normal neurological examination except that ptosis, rash and lacrimation were observed in the patient's left eye. The pupil reflexes and retinal examination were normal and the eye movements were full without diplopia. MRI of the cranium was completely normal without intracranial or orbital abnormalities. Based on the typical clinical findings and the normal MRI, we diagnosed SUNCT syndrome and prescribed lamotrigine. The patient was given lamotrigine with the dose increased by 25 mg every week until a

total dose of 100 mg/day was reached. The patient was followed under lamotrigine treatment for 6 months and was symptom free during that period.

## DISCUSSION

SUNCT syndrome differs from other causes of primary headache in that it is accompanied by autonomic clinical symptoms. It is thought to be associated with hemicrania continua, paroxysmal hemicrania and cluster headache. SUNCT was classified among the subclasses of trigemino-autonomic headaches by the International Headache Society in 2004.<sup>3</sup> SUNCT is a rare syndrome, particularly during childhood; an extensive literature search yielded three children with SUNCT at the ages of 5, 10, and 11 years.<sup>4</sup> The age of onset typically ranges from 23 to 77 years, and the syndrome is more common in men (male/female ratio 17:2). Individual headache attacks last between 5 and 250 seconds (mean, 49 seconds).<sup>5</sup> Frequency of pain attacks varies from 1 to 2 per month to 30 per day. Headaches generally are located unilaterally in the orbital/periorbital region; they rarely have temporal, nasal, mandibular, frontal, palatal and periauricular localizations.<sup>6</sup> Most headache attacks are mild-to-severe burning, stabbing or electrical in nature. Conjunctival congestion is the most common autonomic symptom, and may be accompanied by lacrimation. Frontal sweating, nasal congestion, or rhinorrhea may be observed. Increased intraocular pressure and eyelid edema occurs in the course of headache attacks. Rarely flushing, tachypnea, photophobia, blepharospasm, ipsilateral myosis, nausea, ptosis, conjunctival injection, polyuria, and dilated vessels on the eyelid may be observed. Irregular attacks are characteristic; nocturnal attacks, have also been reported.<sup>7</sup> Triggering by environmental stimulants (e.g., washing the face, eating food, brushing teeth), as in trigeminal neuralgia, has also been reported for SUNCT syndrome. Mechanical movements of the neck, cold or hot weather, emotional

stress, and instant postural changes may precipitate the headache. SUNCT syndrome may also accompany other types of headaches.<sup>8,9</sup>

The etiology and pathogenesis in most cases are not known (idiopathic cases). SUNCT-like cases having posterior fossa lesions, frequently as vascular lesions/malformations of the cerebellopontin junction and brainstem have been reported. The pathologies found in association with SUNCT like syndrome were three artero-venous malformations (in posterior fossa), a brainstem ischemic lesion, an HIV infection and two congenital bone malformations, mainly in the posterior skull (one with seborrheic dermatitis). Most of the lesions found in association with this syndrome involve the posterior fossa or, in general, the posterior part of the brain.<sup>6</sup> These lesions are the causes of secondary SUNCT syndrome.<sup>10,11</sup> The literature suggests that the most common mimics of SUNCT are lesions in the posterior fossa or involving the pituitary gland.<sup>2</sup> A cranial MRI must be performed for such patients to exclude tumors or other causes of headache.

Cluster headache, chronic paroxysmal hemicrania, trigeminal neuralgia of the first nerve division, idiopathic stabbing headache ('jabs and jolts syndrome'), and other headaches are among the main differential diagnoses for the SUNCT syndrome. Cluster headache is a very severe, unilateral pain, lasting 15 to 180 minutes, and occurring from once every other day to 8 times a day. Most of these patients are also male. Chronic paroxysmal hemicrania is severe, lasting 2 to 30 minutes, 1

to 40 times a day and occur more commonly in females. Idiopathic stabbing headache attacks last 3 seconds or less and recur with irregular frequency ranging from one to many times per day.<sup>3</sup> Trigemino-autonomic reflex (TOR) is thought to underlie the basic physiopathology. TOR consists of the trigeminal nerve, the parasympathetic branch of the facial nerve, and brainstem structures. SPECT (single-photon emission computed tomography) and functional MRI studies reveal changes not only in the pericranial, but also in the intracranial blood flow during SUNCT attacks. The main reasons for the SUNCT diagnosis were attack and frequency and duration. In SUNCT syndrome, the usual duration ranges from 5 to 250 seconds, although reported range of duration is 2 seconds to 20 minutes.<sup>12</sup> Chronic paroxysmal hemicrania (CPH) was excluded because of attack frequency is higher (5-10 attacks/day) in CPH. Cluster headache was excluded since its duration is higher than SUNCT (15-180 minutes).

Carbamazepine, ergotamine, sumatriptan, and prednisolone have been used for treatment of SUNCT syndrome. However, none of these drugs is effective long term.<sup>14</sup> Valproate, gabapentin, topiramate, nifedipine, and verapamil are also used for long-term treatment of SUNCT. Recently, lamotrigine has been reported as beneficial.<sup>9</sup> Thus, we started prophylaxis with lamotrigine. At the last follow-up the patient was taking lamotrigine and was symptom free. This rare syndrome should be considered among the causes of headache in childhood.

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