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Case Report

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# An Infant With Paroxysms of Screaming and Unilateral Lacrimation and Rhinorrhea

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## Abstract

We describe the case of a 5-month-old girl who presented to the emergency department with left-sided periorbital edema and erythema. During the assessment, she was observed to suffer repeated hyper-acute episodes of screaming associated with left-sided hemifacial flushing, lacrimation and rhinorrhea. This child was diagnosed with probable short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT), one of the trigeminal autonomic cephalalgias. We believe that this is the youngest probable case of SUNCT for which there is photographic and video evidence.

**Keywords:** Headache; Neuralgiform; Trigeminal; Autonomic; Cephalalgia

## Introduction

Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) is one of the trigeminal autonomic cephalalgias (TACs). TACs are a group of primary headache disorders characterized by a unilateral nature, often with ipsilateral parasympathetic autonomic features [1]. The most recognizable TAC is the cluster headache. SUNCT are overall less common, and seem to be rare in children [2].

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## Case Report

### Investigations

A previously well 5-month-old girl was brought to the emergency department with acute onset periorbital swelling.

The parents described that she had been happily sitting in a highchair when she began screaming uncontrollably without identifiable cause. She was initially able to be settled; however, the screaming returned in paroxysms every 3 - 10 min. The screaming was described as being quite unlike her normal cry in its intensity. Her parents subsequently noticed periorbital swelling and redness of the left eye.

There was no history of head trauma, fever, cough or coryza. She was not receiving any medications and had no past medical history. Antenatal and perinatal histories were normal, she was born at 40 weeks' gestation with a birth weight of 3.575 kg (78th centile), and head circumference was 34 cm (50th centile), with Apgar scores 9 and 9. Developmentally she was appropriate: she rolled both ways, sat with support, reached for objects with a good voluntary grasp, brought her hands to the midline, and was babbling. Her immunizations were complete for age, and she was breast fed. There was a history of migraine in the maternal grandmother, but no other family history of neurological disorders. She lived with her parents.

On admission, the heart rate was 128 beats/min, blood pressure was 76/40 mm Hg and temperature was 36.2 °C. Her growth parameters were: head circumference 40.0 cm (5th centile), length 62.5 cm (9th centile) and weight 5.4 kg (0.5th centile). There was mild left-sided periorbital edema with conjunctival injection with no other skin markings and her face was otherwise symmetrical. She was happy; limb tone and power were normal.

During the initial assessment, four episodes of unusual semiology were observed. Their onset was hyper-acute: she appeared startled; this was rapidly followed by intense screaming and a facial grimace giving the impression of intense pain. There was associated well-demarcated erythema of the left hemiface with ipsilateral conjunctival injection, lacrimation and rhinorrhea. Each episode lasted about 30 s. She was miserable between episodes, during which time her facial erythema, lacrimation, and rhinorrhea were bilateral. Clinical photographs (Fig. 1a and b) and videos were taken of the events and uploaded to the electronic medical record with parental consent.

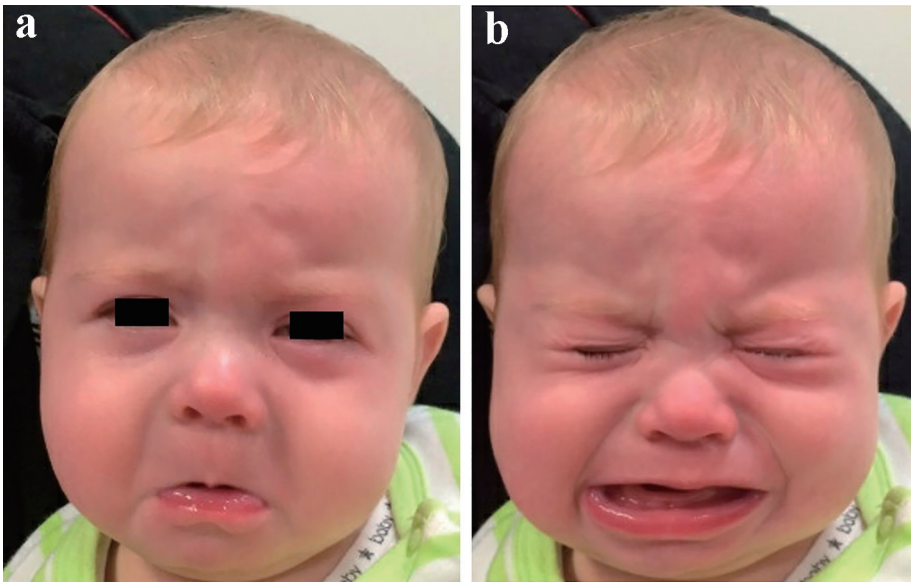


**Figure 1.** (a) Demarcated erythema of left hemiface. (b) Facial grimacing with impression of intense pain.

Following admission to the general pediatric ward, her general and neurological examinations were repeated. Cardiorespiratory and abdominal examinations were normal, as was examination of her tympanic membranes and pharynx. She was settled, the facial skin changes had resolved and her face remained symmetrical. Pupils were equal and reactive to light and she fixed visually. There was minimal head lag and normal truncal tone. When prone she raised herself on straight arms. Limb tone and reflexes were normal with antigravity movements noted.

Over the next 2 days as an inpatient, no further events were noted. No laboratory testing was performed as she remained clinically well.

### Diagnosis

Magnetic resonance imaging (MRI) with a 3T magnet of the head under sedation with a single dose of chloral hydrate 50 mg/kg the following day was normal.

The diagnostic challenges were to clarify what form of paroxysmal event this could be, given that trigeminal neuralgia causes stabbing pain in the distribution of the trigeminal nerve [3], but TACs are rare in childhood and in young children in particular [2, 4], and to exclude an intracranial lesion.

### Follow-up and outcomes

No further episodes were noted in hospital. The clinical photographic and video records were provided to the local tertiary pediatric neurology service. Assessment by this service led to the diagnosis of probable SUNCT.

Physiotherapy intervention was subsequently received for mild gross motor delay with otherwise normal development. She was developmentally normal with tracking growth parameters and with no further episodes when discharged from pediatric follow-up at the age of 26 months.

## Discussion

We describe the case of a 5-month-old girl who presented to the emergency department with left-sided periorbital edema and erythema. She was observed to suffer repeated hyper-acute episodes of screaming associated with left-sided hemifacial flushing, lacrimation and rhinorrhea. This child was diagnosed with probable SUNCT. Such a young case has not been previously described.

The rarity of this condition in pediatrics with only case reports [1, 3-8], and its fleeting clinical features, make diagnosis challenging. While clinical photographs and videos aided in our case, the key to diagnosis was a thorough history with careful attention to the nature and timing of the episodes. However, this rarity also means that the entities of SUNCT and short-lasting unilateral neuralgiform attacks with cranial autonomic symptoms (SUNA) are still poorly understood. For example, migraine is more common in children than SUNCT. International guidelines for the diagnosis of migraine differ between children and adults. No such distinction exists for the more common TACs, much less for SUNCT or SUNA [1].

The clinical spectrum of TACs includes cluster headaches, paroxysmal hemicrania, and short-lasting unilateral neuralgiform headache. Key diagnostic features are recurrence and clinical features ipsilateral to the headache (Table 1).

Short-lasting unilateral neuralgiform headache attacks are further subdivided into those patients with: both conjunctival injection and tearing (SUNCT); and not more than one of conjunctival injection and tearing (SUNA).

SUNCT is further classified as episodic SUNCT: episodes occurring over a period of 1 week to 1 year, separated by in-

**Table 1.** Comparison of Cluster Headache, Paroxysmal Hemicrania, and Short-Lasting Unilateral Neuralgiform Headache[a]

| Headache | Cluster headache | Paroxysmal hemicrania | Short-lasting unilateral neuralgiform headache |
|---|---|---|---|
| Site of pain | Unilateral orbital, supraorbital and/or temporal | Orbital, supraorbital and/or temporal | Orbital, supraorbital, temporal and/or trigeminal |
| Severity of pain | Severe or very severe | Severe | Moderate or severe |
| Duration of pain | 15 - 180 min | 2 - 30 min | One to 600 s as single stabs, series of stabs, or saw-tooth pattern |
| Number of attacks | At least five | At least 20 | At least 20 |
| Frequency of pain | Once every other day to eight times per day | At least five times per day | At least once per day |
| Ipsilateral cranial autonomic features | At least one of the following[b]: 1) Conjunctival injection and/or lacrimation; 2) Nasal congestion and/or rhinorrhea; 3) Eyelid edema; 4) Forehead and facial sweating; and 5) Miosis and/or ptosis. | | |

[a]Adapted from Reference [1]. [b]The diagnosis of cluster headache and paroxysmal hemicrania requires at least one of these symptoms and/or a sense of agitation or restlessness. Criteria for the diagnosis of a short-lasting unilateral neuralgiform headache do not require a sense of agitation or restlessness.

tervals of 3 months or more, and chronic SUNCT: episodes occurring for at least a year with remission periods of less than 3 months or no remission period.

Before making a diagnosis of SUNCT, one must exclude a posterior fossa mass. There have also been patients who have an overlap between SUNCT and trigeminal neuralgia or SUNCT and cluster headache [1]. SUNCT has also been associated in adults with pituitary tumors [9]. In our patient, we did not specifically image the pituitary sella, nor did we perform other specific pituitary investigations. Mitigating against the possibility of a pituitary lesion was the very transient nature of her symptoms and her remaining clinically well with tracking growth parameters to the age of 2 years. Repeat neuroimaging had initially been considered but was deferred as she was monitored and remained well and symptom-free. Sonophobia and photophobia have been described with SUNCT and the other TACs in adults, and this may cause diagnostic difficulty with the alternative of migraine [10, 11], although these symptoms are difficult to elicit in the pre-verbal child and would have been impossible to test in our patient given the brief nature of her symptoms.

In our patient, the attacks lasted less than 1 week so she did not meet the diagnostic criteria for SUNCT, although the nature of the attacks was highly suggestive of this diagnosis.

Pathophysiology of the TACs is poorly understood but is postulated to involve the trigeminovascular system, parasympathetic activation, and various areas of the hypothalamus [11, 12].

In adults with SUNCT, lamotrigine or intravenous lignocaine may be effective [11], but the rarity of this condition in children means there are no data from which therapy can be advocated.

### Learning points

Although TACs, and SUNCT in particular, are rarely the cause for headache in the pediatric population, the diagnosis should be considered in a child with unilateral facial or cranial pain with associated autonomic features.

Diagnosis of rare presentations can be challenging. The addition of contemporaneous clinical photographs can be invaluable for documenting fleeting clinical signs.

As a group, the TACs can be difficult to manage. The hyper-acute nature of SUNCT means that analgesia may not be able to be administered in a timely manner.

## Acknowledgments

None to declare.

## Financial Disclosure

None to declare.

## Conflict of Interest

None to declare.

## Informed Consent

Written consent was obtained from the family.

## Author Contributions

Ryan Lucas drafted the initial case report, reviewed and revised the manuscript. Manoj Menezes and Stephen Sze Shing Teo reviewed and revised the manuscript.

## Data Availability

The authors declare that data supporting the findings of this case report are available within the article.

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