Short-Lasting Unilateral Neuralgiform Headache with Conjunctival Injection and Tearing Secondary to Head and Neck Trauma: Literature Review and Case Report

Marcia V. Rojas-Ramirez, DDS
Resident
Orofacial Pain Clinic
University of Kentucky
College of Dentistry
Lexington, Kentucky, USA

Elizangela Bertoli, DDS, MS
Professor
Orofacial Pain Clinic
University of Kentucky
College of Dentistry
Lexington, Kentucky, USA

Jonathan H. Smith, MD
Assistant Professor
Department of Neurology
University of Kentucky
College of Medicine
Lexington, Kentucky, USA

Correspondence to:
Dr Marcia V. Rojas-Ramirez
University of Kentucky
College of Dentistry
740 S Limestone Street
Lexington, KY 40536-0284, USA
Email: marcia.rojas@uky.edu

This article reports a case of secondary short-lasting unilateral neuralgiform headache with conjunctival injection and tearing following head and neck trauma due to a violent assault. Following the incident, the patient began experiencing 4 to 30 shooting/sharp pain attacks per day in the left anterior temporal and supraorbital areas, with an intensity of 10/10 on a numeric rating scale. Each attack lasted between 10 and 60 seconds. These attacks were accompanied by ipsilateral conjunctival injection, tearing, ptosis of the left eye, blurry vision, and occasional rhinorrhea. Significant improvements in sleep, autonomic symptoms, and pain were observed with a combination of melatonin 10 mg per day, gabapentin 300 mg twice daily, physical therapy, and psychotherapy. This case highlights the relevance of a multidisciplinary approach in the treatment of challenging cases when there is evidence of more than one contributing factor, with the aim of reducing pain and improving the patient's quality of life. *J Oral Facial Pain Headache 2016;30:68–72. doi: 10.11607/ofph.1549*

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Short-lasting unilateral neuralgiform headache with conjunctival injection and tearing (SUNCT) is defined as a short-lasting headache accompanied by autonomic features; it was first described by Sjaastad et al in 1989.[1] It is a rare condition, with an incidence of 1.2 per 100,000 individuals and a reported prevalence of 6.6 per 100,000 individuals.[2,3] It has been reported to be more prevalent in males than females,[4,5] although the male/female ratio varies throughout the literature, ranging from 2:1 to 17:2.[4] SUNCT is currently classified by the International Headache Society (IHS)[6] as a primary headache disorder under the category of trigeminal autonomic cephalalgias (TACs) when idiopathic in nature, and as a secondary headache disorder when the onset of clinical features occurs in parallel with another disorder that can cause headache, such as trauma, pituitary tumor, or whiplash.

The headaches under the classification of TACs are differentiated by the duration and frequency of the pain attacks, as well as by their response to treatment.[7] SUNCT has the shortest duration, with attacks that last between 1 and 600 seconds, while paroxysmal hemicrania attacks last between 2 and 30 minutes, and cluster headache attacks last between 15 and 180 minutes.[6] SUNCT attacks are frequent, occurring anywhere from 3 to 200 times per day, whereas cluster headaches are the least frequent, occurring as little as once every other day. Furthermore, paroxysmal hemicrania is responsive to indomethacin, whereas cluster headache is often responsive to verapamil. However, SUNCT does not respond to either of these treatments, and its management involves pharmacotherapy with anticonvulsants.[8]

The TACs are all associated with ipsilateral autonomic signs, often reflecting activation of the trigeminofacial parasympathetic reflex. Cohen et al[3] reported that among 43 SUNCT patients, the most common autonomic symptoms were ipsilateral rhinorrhea or nasal congestion (53%), ipsilateral ptosis (51%), and nasal blockage (40%). Other

autonomic symptoms included in the International Classification of Headache Disorders (ICHD)[6] are conjunctival injection and/or tearing, ipsilateral eyelid edema, facial flushing or sweating, and a sensation of fullness in the ear.

The pain attacks that characterize SUNCT are intermittent, repetitive, short, unilateral, and stabbing or pulsating in quality. Paroxysms/attacks have an abrupt onset, reaching high intensity within seconds. They are located in the orbitotemporal region, occurring most frequently in the distribution of the ophthalmic (V1) and maxillary (V2) branches of the trigeminal nerve.[6,9] The majority of the patients are pain-free in the interictal periods, with some of them reporting a persistent dull, aching pain.[10–12] There are reports available describing secondary SUNCT involving whiplash,[13] cavernous angioma of the brainstem,[14] pituitary tumors,[15] neurovascular compression,[16] and traumatic head injury.[17] The current report presents a case of SUNCT secondary to head and neck trauma.

Case Report

History of Complaint

A 57-year-old woman presented to the clinic with the complaint of "head pain and left eye flutters." She reported being victim of a violent assault 2 years earlier, which resulted in head and neck injury. Following this assault, she began experiencing episodic shooting/sharp pain, located in the left anterior temporal and supraorbital areas, with an intensity of 10/10 on a numeric rating scale (NRS; where 0 is "no pain at all" and 10 is "the worst pain imaginable"). The patient reported a frequency of 4 to 30 pain attacks per day, each attack lasting between 10 and 60 seconds. These attacks were accompanied by ipsilateral conjunctival injection, tearing, ptosis of the left eye, blurry vision, and occasional rhinorrhea. Periodically, she would experience left periorbital spasms and nausea after the attacks. No nocturnal attacks were reported. The patient had not identified precipitating factors for these attacks, although she reported that bright lights, changes in weather, and stress aggravated the symptoms. Shortly after the trauma, she underwent neuropsychometric evaluation, which demonstrated changes consistent with mild traumatic brain injury.

Clinical Examination

Cranial nerve examination revealed mild ptosis of the left eye, with no deficit in visual acuity, pupillary function, or extraocular motility. Funduscopic examination was normal.

Examination of cervical range of motion revealed restriction during extension and rotation (less than 70 degrees for both). This examination was based on a general quantitative approach using the reference values proposed in recently published guidelines.[9] No lymphadenopathy was noted. Palpation revealed tenderness in the cervical (bilateral occipitalis and trapezius, and left sternocleidomastoid) and masticatory (bilateral masseter) muscles. Left trapezius muscle palpation reproduced the patient's familiar pain attacks, although it did not trigger the autonomic features. Mandibular function and provocation tests were within normal limits. Range of mandibular motion was 60 mm on maximum assisted opening and 13 mm on lateral excursions. Temporomandibular joint (TMJ) examination revealed the presence of bilateral intermittent asymptomatic subluxation. No other sounds were identified in the TMJs. Intraoral examination was within normal limits. Additional diagnostic tests such as INDOtest or trigger point injections were not performed.

Imaging

A brain magnetic resonance imaging (MRI) scan with gadolinium enhancement was obtained and no abnormal findings were observed.

Psychological Evaluation

During her initial appointment, the patient completed self-report questionnaires assessing psychological well-being. The Symptom Checklist-90 R[18,19] was used to measure psychological distress. No elevations were seen in the Global Severity Index. The Multidimensional Pain Inventory[20] was used to measure psychosocial adaptation. All values were within normal limits. The Satisfaction with Life Scale[21,22] was used to measure the patient's adaptation and degree of happiness. This scale revealed the patient's dissatisfaction with life, which she claimed to be due to the dysfunction and pain following the assault. She also underwent a psychological interview by behavioral medicine professionals who recommended pursuing psychotherapy for anger management as a consequence of the assault.

Treatment Approach

The possible presence of temporomandibular disorders (TMD) was assessed to determine if TMD was a causal or contributing factor in the patient's pain complaints. During the clinical examination there was evidence of masticatory masseter muscle pain, which in view of the patient's history was likely secondary and not the cause of the pain attacks.

The cervical muscles were also painful, especially the left trapezius muscle, which triggered the patient's familiar pain attacks. The first treatment approach included a referral for physical therapy (PT) to address the presence of trigger points, improve cervical strength/motion, and reduce pain. The patient

underwent 8-week sessions of PT treatment. She reported that during the first PT sessions the number of attacks per day increased, but as therapy continued, the frequency slowly decreased. Improvements in the cervical range of motion were also noted.

At a later follow-up, a pharmacologic approach was introduced and the patient was started on lamotrigine (25 mg a day), but she was unable to tolerate this low dose because of side effects. After the 8 weeks the PT treatment concluded, although she continued practicing with a home exercise program that was provided. She also reported a decrease in the frequency of pain attacks to two to four per day, with intermittent pain-free days. She continued having ptosis of the left eye and tearing, especially when exposed to fluorescent light. However, a few months later she reported an increase in attack frequency (two to seven pain attacks per day) and was referred to a neurologist. At that time, she also reported symptoms of depression, frustration, and anxiety. She was evaluated by behavioral medicine specialists, who provided her with a referral for psychotherapy to a clinical psychologist close to her home area.

The neurologist who evaluated the patient recommended a trial of melatonin (up to 12 mg at bedtime) and gabapentin (300 mg slowly titrated up to 900 mg/day). Two weeks later the patient reported taking 10 mg of melatonin per day and 300 mg of gabapentin twice daily. This approach resulted in significant improvements in sleep and reduction of the autonomic symptoms. Additionally, although she continued to have pain attacks, they decreased in frequency and number (one to two attacks per day, with intermittent pain-free days).

Currently, she continues with psychotherapy and a combination of melatonin (10 mg per day) and gabapentin (300 mg twice daily). The improvements noted above continued to be maintained up to the last follow-up, which was performed almost 2 years after the initial appointment.

Discussion

Several steps are important to consider when evaluating orofacial pain patients with a headache complaint. The first step is to determine if the condition is a primary or secondary disorder based on the ICHD.[6] In the case presented here, the onset of the pain attacks was in close association with the violent assault involving trauma to the head and neck region, and therefore it was classified as a secondary disorder. Secondary SUNCT has been extensively reported throughout the literature.[13–15,17,23–40] Distinct causes have been linked to this condition, and although not common, indirect injury to the neck and cervical area has been included as an etiologic factor.[13,17] The underlying pathophysiology is not well understood, and theories vary depending on the supposed etiology. In this case, pain attacks were reproduced by palpation of the left trapezius muscle. The literature has reported a connection between the face and neck through a referral mechanism, which can be explained by convergence of trigeminal and cervical spine nociceptive afferents at the level of the trigeminal subnucleus caudalis.[13]

A second step in the evaluation process is to build a differential diagnosis taking into account demographics, pain history, and clinical features. Descriptions involving the presence of unilateral, short-lasting attacks, high pain intensity, and ipsilateral autonomic features should clue the examiner toward the existence of a TAC. As in the present case, differentiation among the TACs is guided by careful documentation of attack frequency and duration.

Another condition that could confuse the examiner and should be included in the differential diagnosis is trigeminal neuralgia. Trigeminal neuralgia is a condition characterized by unilateral, short-lasting, electric shock–like pain that develops in one or more branches of the distribution of the trigeminal nerve.[6] These attacks tend to be elicited by nonnoxious stimuli of particular areas called trigger zones. In the case presented here, the attacks could be reproduced by palpation of the left trapezius muscle. Muscle palpation requires the application of a force that usually varies between 2 and 4 kg,[9] which is not consistent with the type of stimuli that tends to elicit a trigeminal neuralgia attack, such as light touch. Autonomic symptoms are not required to meet the diagnostic criteria for trigeminal neuralgia, although there have been some case reports in the literature of trigeminal neuralgia associated with mild autonomic symptoms.[41,42] On the other hand, autonomic symptoms are required to meet the diagnostic criteria for SUNCT. Regarding the most common sites of pain, trigeminal neuralgia has a higher prevalence in the maxillary (35%) and mandibular (30%) divisions of the trigeminal nerve and a much lower prevalence in the ophthalmic division, whereas SUNCT has a higher prevalence in the ophthalmic division (67%), followed by the maxillary division (33%).[43] Therefore, when the triggering mechanism, the presence of autonomic features, and the branch distribution of the trigeminal nerve are considered, SUNCT is a more probable diagnosis than trigeminal neuralgia.

A third step in the evaluation process is to build a treatment plan addressing the clinical presentation. Management of SUNCT can be challenging due to the lack of a standardized and/or evidence-based treatment strategy. Because of the short duration of the pain attacks, an abortive therapy is not

feasible. Therefore, the use of prophylactic medication may reduce the number of pain attacks experienced. The preventive medications that have shown beneficial outcomes include the anticonvulsant family: lamotrigine,[44–46] topiramate,[47] gabapentin,[48,49] and zonisamide,[50] among others.[2] For the present case, lamotrigine was not suitable due to the emergence of side effects in the patient. Gabapentin has a better side-effect profile and tends to be better tolerated by patients. The exact mechanism of action of gabapentin remains unknown, although it is considered to be an alpha-2 delta ligand of voltage-gated calcium channels.[51,52]

It is also interesting to highlight the partial therapeutic response to melatonin in this patient.[53–55] Melatonin is a structural analog of indomethacin, which has been noted to be helpful in other autonomic cephalalgias, such as hemicrania continua and paroxysmal hemicrania.[53,54] SUNCT is not considered a condition that is thought to be indomethacin-responsive. Adverse events limiting the utility of indomethacin include gastrointestinal ulceration and, not uncommonly, drug-induced headache. The patient in the present case reported a history of gastric ulcers, mandating avoidance of indomethacin.

When a cause or a contributing factor is identified, such as cervical dysfunction, approaches such as PT can positively impact the treatment outcome. In the present case, PT significantly reduced the frequency of the pain attacks and improved the patient's cervical range of motion.

Management of patients with SUNCT continues to be a challenge for clinicians and involves a trial of different approaches. The present case highlights the importance of a multidisciplinary approach for the management of SUNCT, where valuable input from distinct areas of expertise are provided with the goal of managing the pain and improving the patient's quality of life.

Conclusions

Cervical and head traumas are known conditions to refer pain to the orofacial region. In the case presented here, a multidisciplinary approach including PT, psychotherapy, and pharmacologic treatment with melatonin and gabapentin resulted in symptomatic improvements in a secondary case of SUNCT.

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