Olgu Sunumu / Case Report

CASE OF SHORT-LASTİNG UNİLATERAL NEURALGİFORM HEADACHE ATTACKS WİTH CONJUNCTİVAL INJECTİON AND TEARİNG HEADACHE MİMMİCKİNG TRİGEMİNAL NEURALGİA

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ABSTRACT

Short-lasting, Unilateral, Neuralgiform headache Atack with Conjunctival injection and Tearing (SUNCT) headache is a rare, primary headache disorder and a member of Trigeminal autonomic cephalgias (TACs) that is characterized by brief, intense episodes of pain in the head and face accompanied with paroxysmal facial autonomic symptoms . In some cases, the symptoms of SUNCT headache can resemble those of trigeminal neuralgia, a condition that is also characterized by intense facial pain. Both conditions can cause similar symptoms such as sudden, sharp, and severe pain in the face, but there are differences in their clinical presentation and medical treatment. Early and accurate diagnosis is essential because of possible complications and long-term negative impact on the patient's quality of life Trigeminal neuralgia is usually treated with anticonvulsant medications, while SUNCT headache is often resistant to treatment with anticonvulsant medication; and calcium channel blockers, anesthetics like lidocaine and/or nerve blocks being amoung the main treatment options.

Keywords: Trigeminal cephalgia, neuralgiform headache, SUNCT headache

INTRODUCTION

SUNCT (Short-Lasting Unilateral Neuralgiform headache Attacks with Conjunctival Injection and Tearing) and trigeminal neuralgia (TN) are two distinct types of facial pain conditions that share some common features: Facial Pain: Both SUNCT and TN cause intense, shooting pain (in the face), typically on one particular side of the face (unilateral).Trigeminal Nerve Involvement: Both conditions affect the trigeminal nerve, which is responsible for facial sensation and motor functions. Short Duration: SUNCT headaches typically last from a few seconds up to a minute, while TN pain can last from several seconds to several minutes.Triggering Factors: Both SUNCT and TN can be triggered by certain activities, such as eating, speaking, or exposure to the wind.Similar Treatment Options: Both conditions can be treated with medications, such as anticonvulsants and anti-inflammatory drugs, and other treatments like nerve blocks and neurosurgery.It is important to note that while these conditions share some similarities, they are distinct entities, and a proper diagnosis is essential for effective management and treatment of the disorder. The exact pathophysiology of SUNCT headaches is not fully understood, but it is thought to involve hypothalamic dysfunction and trigeminovascular mechanisms have been implicated (Goadsby,2002) .The pain in SUNCT is believed to result from the activation of pain-sensitive nerve fibers within the trigeminal nerve.

It is suggested that a malfunction in the trigeminal nerve's pain-processing pathway may cause the intense, brief headaches that are characteristic of SUNCT. This malfunction may result from various factors, such as injury, inflammation, or an underlying neurological condition(Cephalalgia. 2018, Goadsby ,1999).It is important to note that the pathophysiology of SUNCT headaches is complex and not fully understood, and more research is needed to fully comprehend the mechanisms that underlie this condition.

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CASE PRESENTATION

A 42-year-old female presenting with SUNCT (Short-Lasting Unilateral Neuralgiform headache Attacks with Conjunctival Injection and Tearing) headaches and AICA (Anterior Inferior Cerebellar Artery) vascular compression of the trigeminal nerve was evaluated. In the initial phase, the patient experienced brief episodes of left sided periorbital pain of moderate intensity. Due to lack of conjunctival injection, rhinorrhea, lacrimation, facial sweating, and hypersalivation , we first considered the possibility of trigeminal neuralgia and started pregabalin treatment with slow titration. Pregabalin treatment did not show significant results

In the meantime, the patient had repeated brain MRI imaging, EEG examination, and further investigation for other possible autoimmune diseases at different centers. As a result of this investigation, the patient was found to be completely normal in all examined tests except for the high level of Anti-TPO and Anti-TG. During this period, the patient gave almost no response to high-dose NSAID, opioid, methylprednisolone, and diazepam combined treatments. On repeated brain MRI examination, the presence of AICA vascular compression suggested that the trigeminal nerve may be irritated or injured by the compression of the blood vessel, leading to the development of SUNCT headaches, as indicated in Figure 1.

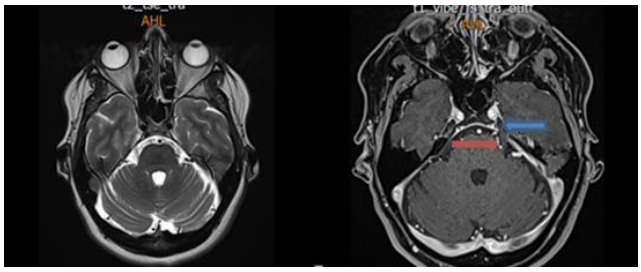

Figure 1. Compression of the left trigeminal nerve by an anterior inferior cerebellar artery in a prepontine cistern on axial MR image, Red arrow: Left anterior inferior cerebellar artery, Blue arrow: Left trigeminal nerve

In the second week, our patient was started on carbamazepine 200mg/day, the patient's simultaneous pregabalin treatment was gradually reduced, and the frequency and severity of the headache attacks began to decrease by 70-75% in the patient on carbamazepine treatment. In the 4-month follow-up of the patient, constant and chronic headaches persisted, but we observed that SUNCT headache episodes continued in the left V1 branch every 2 or 3 weeks. Currently, 600 mg/day carbamazepine treatment is ongoing. However, all NSAID options, methylprednisolone, and tramadol intravenous treatments are completely ineffective in attack treatment. The patient did not give consent for Intravenous lidocaine treatment to be performed under the surveillance of an anesthesiologist.

DISCUSSION

This case report describes a 42-year-old female with SUNCT headaches and AICA vascular compression of the trigeminal nerve. Initially, the patient was diagnosed with trigeminal neuralgia and treated with pregabalin. However, the patient did not respond to this treatment and was found to have high levels of Anti-TPO and Anti-TG. Repeat brain MRI examination showed AICA vascular compression, which led to the development of SUNCT
headaches. The patient was started on carbamazepine, which reduced the frequency and severity of headache attacks by 70-75%. The patient continues to experience constant and chronic headaches, with SUNCT headache episodes occurring every 2-3 weeks. Lidocaine treatment has been suggested as the most effective treatment for resistant SUNCT headaches. The choice of treatment will depend on the individual case and severity of symptoms. Treatment options for SUNCT headaches with AICA compression may include medications to manage pain and prevent future headache attacks, as well as interventional procedures such as nerve blocks or neurosurgery, to relieve the compression on the trigeminal nerve. The choice of treatment will depend on the individual case and the severity of the symptoms.The most effective treatment for resistant SUNCT headaches is lidocaine treatment (Baraldi 2014)
The patient was diagnosed with SUNCT headache due to severe, unprovoked pain attacks lasting 10-15 seconds on the left side of the face, which persisted for a maximum of 10 minutes. During some periods, the patient experienced up to 100 attacks per day accompanied by symptoms such as conjunctival injection, rhinorrhea, lacrimation, facial sweating, and hypersalivation. Despite medical treatment, the attacks were resistant, and in the second week of treatment, the patient experienced loss of consciousness due to severe headache.Vascular compression of the AICA (Anterior Inferior Cerebellar Artery) has

been proposed as a potential cause of SUNCT (Short-Lasting Unilateral Neuralgiform Headache Attacks with Conjunctival Injection and Tearing) headaches. This hypothesis suggests that the AICA, which supplies blood to the brainstem and cerebellum, can become compressed by nearby structures, leading to irritation or injury of the trigeminal nerve.

In this theory, the compression of the AICA would cause pain in the distribution of the trigeminal nerve, leading to the intense, sudden headaches that are characteristic of SUNCT.Individuals who experience SUNCT headaches are more likely to have AICA compression, in comparison to healthy individuals or those who have other types of headaches. In other words, there may be a correlation between the occurrence of SUNCT headaches and the presence of AICA compression.

It is important to note that the relationship between AICA compression and SUNCT headaches is still the subject of ongoing research and debate, and more studies are needed to fully understand the role of this mechanism in the development of SUNCT headaches. Trigeminal neuralgia may be the expected approach when vascular compression of the trigeminal nerve is observed in brain MRI, but vascular compression should also be considered for SUNCT headache, as it may be the underlying cause. Diagnostic imaging studies, such as magnetic resonance imaging (MRI) or computed tomography (CT) angiography, may be used to confirm the presence of AICA compression and evaluate the extent of any compression-related damage to the trigeminal nerve. Additionally, not all patients with SUNCT headaches present evidence of AICA compression, suggesting that there may be multiple mechanisms involved in the development of this condition. The mechanism of vascular compression in TN is a well-known cause and can be detected between 47-90% in scientific studies (Gardella L 2001), whereas vascular compression for SUNCT is still a controversial issue. In a series of 52 cases of SUNCT and SUNA, only three (7%) vascular compression was documented in brain imaging studies (Cohen AS 2006) while in other studies contact between vascular structures and the trigeminal nerve has been reported in nearly 90% of patients with SUNCT headaches (Williams MH,2008). Since trigeminal nerve vascular compression also causes trigeminal neuralgia (TN), differential diagnosis of trigeminal neuralgia and SUNCT is important. It is extremely important to know the duration of the pain and the frequency of the attacks in differential diagnosis. In a study of trigeminal nerve V1 division neuralgic pain TN patients, a median pain episode duration of 5 seconds (range, 2-32 seconds) was found, compared with 60 seconds (range, 5-250 seconds) for SUNCT syndrome patients, 10 minutes(range, 2-45 minutes) for chronic paroxysmal hemicrania patients (Sjaastad O,1997). Another feature of SUNCT headache is conjunctival injection and lacrimation which is present in 100% of the cases (Weng HY 2018). Another distinctive clinical feature is that 95% of SUNCT syndrome patients do not have a refractory period, which means patients could have another attack immediately after cessation of the preceding one, therefore,it is important to note that the refractory period in SUNCT syndrome is relatively short compared to other types of headaches,for example cluster headaches, in which the refractory period can last for days or weeks. This shorter refractory period in SUNCT syndrome is one of the features that help to distinguish it from other types of headaches (Cohen AS,2006). The involvement of the first branch of the trigeminal nerve (V1), which innervates the periorbital, forehead, and parietal regions and the side of the unilateral nazal prtion, was found to be the most typical location 64.4% in SUNCT patients (Zhang S,2022). During recent follow-up appointments, the patient was assessed by neurosurgeons to determine if decompressive surgery was a suitable option. However, based on their evaluation, the patient was not deemed a suitable candidate for decompressive surgery. Therefore, the patient was advised to be evaluated by algology specialists, who specialize in managing pain and may be able to provide alternative treatment options for the patient's SUNCT headaches.

CONCLUSION

Differentiating SUNCT from trigeminal neuralgia is important for proper diagnosis and treatment. The absence of a refractory period, the presence of conjunctival injection and lacrimation in all cases, and most importantly resistance to classical neuralgia anticonvulsant medical therapy cases, should raise suspicion for SUNCT headaches. Thus, SUNCT and trigeminal neuralgia may

require different medications or surgical procedures , and treatment may invariably be more effective if the correct diagnosis is made.

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