Olgu Sunumu / Case Report
Aegean J Med Sci 2024;1:53-56

A Rare Cause of Trigeminal Throphic Syndrome: Ischemic Cranial Neuropathy

Trigeminal Trofik Sendromun Nadir Bir Nedeni: İskemik Kraniyal Nöropati

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ÖZET

Trigeminal trofik sendrom, trigeminal sinirin farklı nedenlere ortaya çıkan hasarı sonucu trigeminal anestezi, fasiyal parestezi ve yüzde ülsere lezyonlar ile karakterize nadir görülen bir hastalıktır. Ülserasyon en sık burun kanadında ve tek lezyon şeklinde görülmekte olup trigeminal sinir dermatomuna uyan bölgelerde çoklu ülsere lezyonlarla nadiren karşılaşılmaktadır. Burada iskemik kraniyal nöropati sonrası çoklu fasiyal ülserasyonlarla başvuran bir Trigeminal Trofik sendrom olgusu sunulmaktadır.

Anahtar Kelimeler: fasiyal parestezi, fasiyal ülser, trigeminal trofik sendrom

ABSTRACT

Trigeminal trophic syndrome is a rare disease characterized by trigeminal anesthesia, facial paresthesia, and ulcerated lesions on the face as a result of damage to the trigeminal nerve due to different causes. Ulceration is most commonly seen as a single lesion on the nasal wing. Multiple ulcerated lesions are rarely encountered in the regions at trigeminal nerve dermatome. Here, a case of Trigeminal Trophic syndrome presenting with multiple facial ulcerations after ischemic cranial neuropathy is reported.

Keywords: facial paresthesias, facial ulcer, trigeminal trophic syndrome

INTRODUCTION

Trigeminal trophic syndrome (TTS) is characterized by the classical clinical triad which consists of trigeminal anesthesia, facial paresthesia and ulcers on trigeminal nerve dermatome (1). It is twice as common in women (2).

Trigeminal trophic syndrome is a rare disease, firstly described in 1901 by Wallenberg and later it was defined as a skin ulcer located in a trigeminal dermatome by Loveman in 1933 (3). Since its description, there have been limited number of case reports in literature.

Although it was initially believed that the lesions were the sequelae of degraded fibers causing loss of neuronal trophic factors, it became clear that the ulceration was caused by self-manipulation for relief from distressing dysesthesias (4). The diagnosis of TTS is based on the presence of a condition that may lead to trigeminal nerve injury and the exclusion of other differential diagnostic possibilities (5).

Here, a case is reported with multiple facial ulcerations diagnosed with ischemic cranial neuropathy, a rare etiology of TTS.

CASE

A 85-year-old man with diabetes mellitus, hypertension and Bell's palsy presented with a 2-month history of an expanding multiple ulcers located on the left side of nasal wing, nasolabial sulcus and lateral side of periorbital skin (Figure 1). The patient had Bell's palsy and partial ptosis on the left side for two years and had an episode of diplopia due to ischemic abducens nerve cranial neuropathy two months ago. After that episode, he began to experience altered sensations on the left side of his face, characterized by numbness, tingling and persistent itching and started rubbing his skin resulting in multiple ulcers. Neurologic examination revealed decreased sensation of pain, light touch and temperature on the left side of the face, more prominent around the ulcers .

Hematological and biochemical laboratory tests were within normal limits. Neuroimaging revealed age-related cortical atrophy and diffuse leukoaraiosis related to hypertension and diabetes mellitus (Figure 2). Systemic examination revealed no abnormality. An underlying malignant, vasculitic or infectious cause for the ulcer was excluded by a skin biopsy at another advanced center. He was diagnosed as trigeminal trophic syndrome caused by

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Received/Geliş Tarihi: 29 08 2023 || **Accepted/Kabul Tarihi:** 27 12 2023

simultaneous trigeminal nerve ischemia with abducens nerve paralysis.

Figure 1. *Ulcers on the nasal wing, nasolabial sulcus and lateral periorbital skin, periorbital edema and sequelae of ptosis due to self-mutilation are shown.*

The patient and relatives were informed about the self-induced nature of the ulceration and the importance of using soft cotton gloves. Treatment plan was to add low dose gabapentin such as 300 mg daily for painful dysesthesias and to increase the doses gradually with the use of clopidogrel for secondary prophylaxis on ischemic cranial neuropathy. Unfortunately, the patient died of bilateral massive subdural hematoma while on gabapentin 600 mg daily.

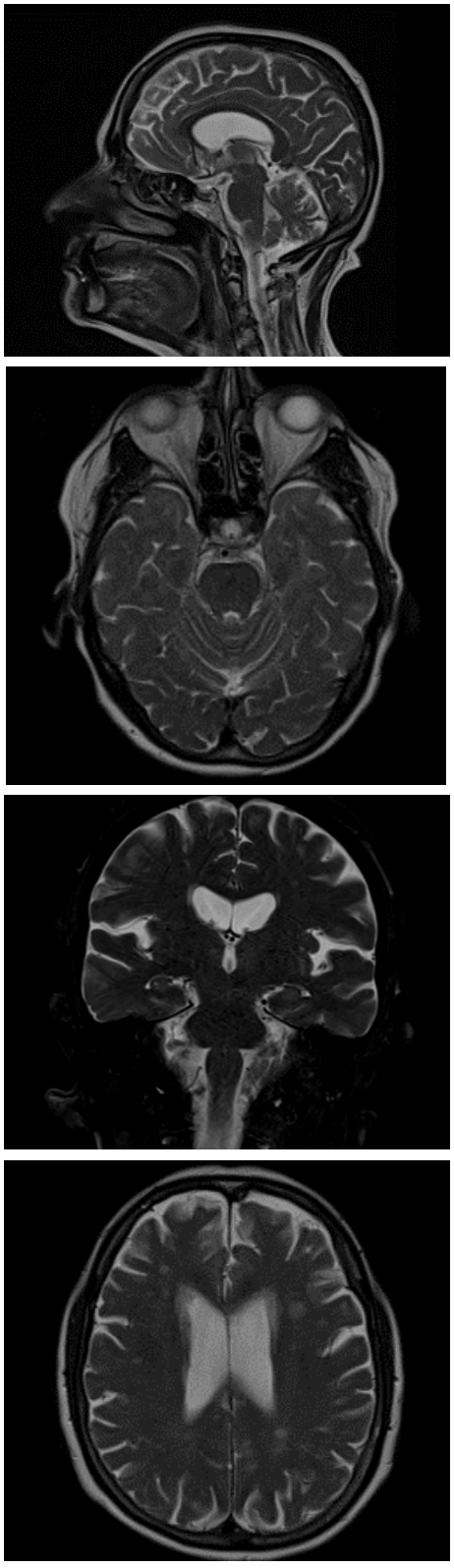
Written informed consent was obtained from the patient for their anonymized information to be published in this article.

DISCUSSION

Trigeminal trophic syndrome is a rare condition characterized by trigeminal anesthesia, paresthesia, and ulceration mostly located at the nasal wing. The nasal wing is characteristically affected the but it can also affect cheeks, upper lip, the frontal region and other areas of trigeminal dermatome (1). The tip of the nose is especially spared due to the different innervation by the medial nasal branch of the anterior ethmoidal nerve (6).

Lesions occur as a result of unintentional repetitive plucking, rubbing and scratching of the affected area due to anesthesia, paresthesia or pain caused by damage to the sensory branch of the trigeminal nerve (2). The fact that the complaints started at the same time as the abducens nerve palsy suggests that TTS occurred as a result of ischemic neuropathy of the trigeminal nerve simultaneously. It is known that the earlier onset of clinical manifestation may be due to involvement of multiple cranial nerves, as in our case. The latency period between the damage and the appearance of the lesions is variable, ranging from weeks to decades (7). In the present case, there was a delay of two months before lesions appeared.

Figure 2. *Cortical atrophy and leukoaraiosis are shown in MRI*



Trigeminal trophic syndrome must be differentiated from other causes of dermatological facial ulcerations; basal cell carcinoma and squamous cell carcinoma, herpes virus infections, granulomatous diseases and pyoderma gangrenosum (8). Although the most common causes are stroke, acoustic neuroma, encephalitis, trauma and herpes zoster, trigeminal trophic syndrome may be triggered by iatrogenic causes, generally following interventional treatment procedures for trigeminal neuralgia (9).

Treatment of TTS is challenging and often requires a multidisciplinary approach. The first step in treatment is the prevention of self-mutilation in patients. Patients should be warned to keep their nails short, to wear a soft bandage on the fingers, and to wear soft gloves (10). If repetitive compulsive behaviors continue, psychiatric consultation should be recommended. Topical and systemic antibiotics should be used to prevent secondary infections for the local wound care (11).

It is clear that recurrence may develop in the future if the trigeminal nerve damage is not repaired. Drugs such as amitriptyline, pregabalin, lamotrigine, phenytoin, baclofen, botulinum toxin, gabapentin, oxcarbazepine and carbamazepine have been used in the treatment of trigeminal neuralgia (12). Recently, gabapentin and carbamazepine have been evaluated as the first choice in the treatment of TTS (13). The most common side effects of carbamazepine are drowsiness, blurred vision, diplopia, ataxia, and balance disorder, which occur in a dose-dependent manner, and it can be life-threatening by causing a fall. Particular attention should be paid to hyponatremia, which is a late complication of treatment with carbamazepine, especially in the elderly (14). Gabapentin was preferred as being a more commonly used neuropathic pain medication and because of the expectation that its side effects would decrease with dose titration.

CONCLUSION

Trigeminal trophic syndrome is a rare condition and can be easily misdiagnosed if it is not well known. The diagnosis of TTS should be considered especially in patients presenting with unilateral neuropathic pain and ipsilateral facial ulceration. Delayed diagnosis and inappropriate management without behavioral modification tend to result in chronic ulcers. This report is important for identifying a rare case of TTS caused by a part of isolated ischemic cranial polyneuropathy.

Etik; Bu yazıda sunulan olgu için sunulan bilgilerin akademik amaçlı kullanımı hakkında detaylı bilgileri de içeren imzalı "Bilgilendirilmiş onam formu" alınmıştır.

Ethics; For the case presented in this article, a signed "informed consent form" was obtained, which includes detailed information about the use of the information presented for academic purposes.

Yazar katkı durumu; Çalışmanın konsepti; Ö,Ö, dizaynı; Ö,Ö, Literatür taraması; Ö,Ö, verilerin toplanması ve işlenmesi; Ö,Ö, yazım aşaması; Ö,Ö,

Author contribution status; The concept of the study; Ö,Ö, design; Ö,Ö, literature review; Ö,Ö, collecting and processing data; Ö,Ö, writing phase; Ö,Ö,

.Yazarlar arasında çıkar çatışması yoktur.

The author declares no conflict of interest.

Finansal Destek: yoktur / Funding : none

doi: https://doi.org/10.33713/egetbd.1351245

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