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Case Report

Bilateral Trigeminal Trophic Syndrome

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Abstract

Trigeminal trophic syndrome (TTS) is a rare condition that arises secondary to damage to the peripheral or central sensory pathways of the trigeminal nerve. TTS classically presents with a triad of trigeminal anesthesia, facial paresthesias, and self-induced lateral ala nasi ulceration. TTS should be considered in the differential diagnoses of unilateral facial ulcerations due to the unique management that it requires. Patient education to prevent compulsive manipulation of the lesion is a fundamental piece to the treatment of TTS. We describe a 65-year-old woman who presented with a rare case of bilateral trigeminal trophic syndrome and right-sided nasal ulceration.

Keywords: Ala nasi ulceration; Facial ulceration; Paraesthesia; Trigeminal trophic syndrome

Introduction

Trigeminal trophic syndrome is a disease rarely addressed in the medical literature with about 100 cases reported to date [1]. It was first described by Wallenberg in 1901 as a cutaneous ulceration in the trigeminal dermatome and later by Loveman in 1933. TTS is often reported in patients who have sustained damage to peripheral or central components of the trigeminal nerve. Patients present with a classic triad of paraesthesia, anesthesia, and ala nasi ulceration. The combination of paraesthesia and anesthesia in the sensory distribution of the trigeminal nerve leads to recurrent self-manipulation and the eventual painless formation of an ulcerative lesion. The tip of the nose is classically spared due to its innervation by the medial nasal branch of the anterior ethmoidal nerve.

Oftentimes TTS occurs after procedures aimed at the treatment of trigeminal neuralgia such as surgical trigeminal ablation by rhizotomy of the dorsal trigeminal nerve root or alcohol injection that interrupts the Gasserian ganglion [2].

Interestingly however trigeminal trophic syndrome only occurs in a minority of patients with damage to the trigeminal nerve system. TTS occurs in 18% of patient's post-trigeminal ablation and 75% of TTS cases occur after disruption of the Gasserian ganglion [1]. Stroke is another common cause of TTS, especially in the case of Wallenberg lateral medullary syndrome and posterior cerebral artery stroke. The time period between the injury to the trigeminal pathway and onset of TTS can differ from 2 weeks to 30 years with the median interval of 1 year [3].

Early recognition of the disease and prompt treatment is crucial in the management of these patients and therefore TTS needs to be considered in the differential diagnosis of patients along with possible infectious, malignant, and vasculitic pathologies. Here we report the case of a 65-year-old woman with rare bilateral trigeminal trophic syndrome.

Case Report

Our patient is a 65-year-old woman who presented for evaluation of a right nasal and paranasal ulcerative lesion. The patient reported the presence of bilateral facial hypoesthesia affecting all three nerve branches beginning in December of

2019 with accompanying neuralgic pain and paraesthesias predominantly in the right V2 dermatome. These paraesthesias caused continuous itching and irritation of the right nasal and paranasal territory which occurred even unconsciously during her sleep. Beginning in March 2020 an ulcerated lesion began to appear on the right nasal ala progressively extending to affect the totality of the right nostril and adjacent skin surface (Figure 1). Due to the global SARS-CoV-2 health crisis the patient remained in confinement in her home until she was seen by her primary care provider near the end of June 2020 at which point she was referred to the hospital for further evaluation.

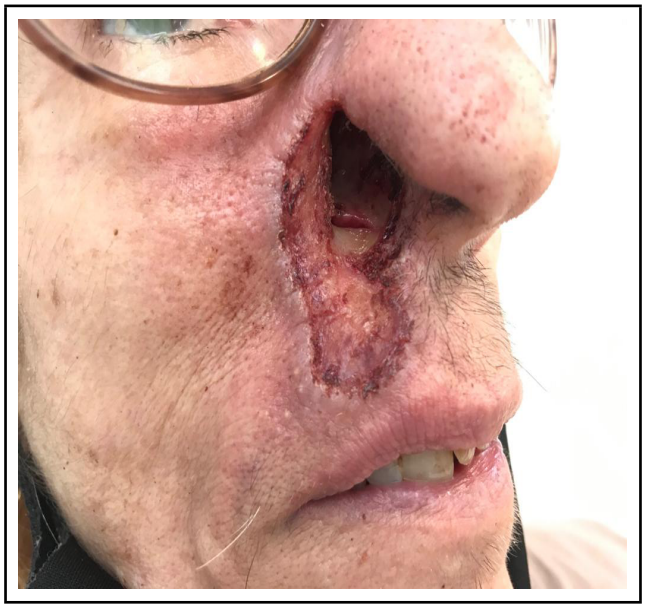

Figure 1: An ulcerated lesion affect the totality of the right nostril and adjacent skin surface.

Upon physical exam the patient presented with bilateral facial hypoesthesia predominating in the right side of the face and the V2 dermatome. She did not present with facial paresis. At this time the patient demonstrated a right nasal ulcer of 6 months evolution which affected the entire nostril, nasal septum, and right lateral wall. Biopsy of the lesion demonstrated inflammatory tissue of the squamous epithelium and pseudoepitheliomatous hyperplasia and was negative for malignancy. Cultures were performed that confirmed the presence of penicillin-resistant Staphylococcus aureus, for which treatment with trimethoprim-sulfamethoxazole was given. All further microbiological studies were negative.

Cranial magnetic resonance imaging (MRI) was performed and showed significant thickening of the left trigeminal nerve (Figure 2 and 3), from its emergence through the prepontine cisternal tract to at least Meckel's cave, of undetermined etiology, possibly inflammatory, although a schwannoma-type tumor could not be ruled out. The right trigeminal nerve showed slight atrophy but was difficult to assess due to the asymmetry of the two trigeminal nerves. The cranial MRI study also demonstrated multiple cerebral vascular lesions of small vessels of old appearance, together with a recent subacute right lacunar pontine lesion.

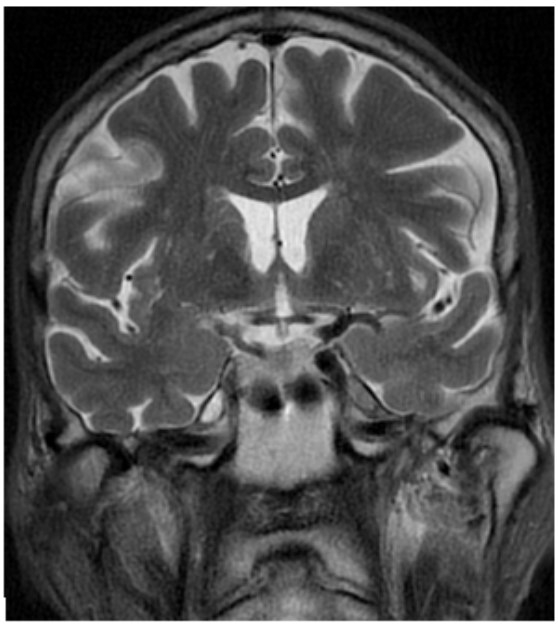

Figure 2: Coronal T2 sequence showing absence of CSF signal in Meckel's cave, which is obliterated by the lesion of the left trigeminal nerve.

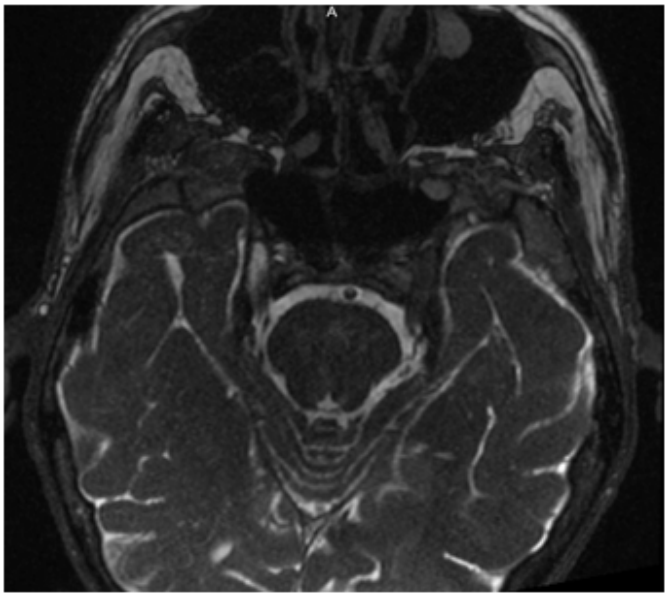

Figure 3: High resolution axial T2 sequence (FIESTA). Thickening of the left trigeminal nerve is observed at the level of the prepontine cistern and Meckel's cave.

The patient was diagnosed with trigeminal trophic syndrome with ulceration of the complete right nare, nasal septum, and lateral wall. Daily Bactroban cures were initiated as well as protective dressing of the ulcer. Additional diagnoses included a right pontine lacunar infarction and several small vessel cerebral infarcts. The patient was educated as to the nature of her condition and instructed of the importance of strict avoidance of all manipulation of the lesion. After discussing the case with plastic surgery, future nasal reconstruction was recommended [4-15].

Discussion

To the best of our knowledge there are very few cases of bilateral trigeminal trophic syndrome reported in the medical literature at this time. The patient's presentation is consistent with classic TTS including the presence of paraesthesia, anesthesia, and persistent self-mutilation of the lesion within the V2 dermatome with sparing of the tip of the nose.

This characteristic presentation of TTS allowed for correct diagnosis through physical examination, patient history, and the appropriate complementary studies. Our patient sustained bilateral trigeminal damage without any history of treatment of trigeminal neuralgia but rather secondary to previous cerebral infarction.

Trigeminal trophic syndrome is a diagnosis of exclusion. When considering TTS a biopsy of the facial lesion should be obtained in order to rule out other diagnoses. Other causes of nasal ulceration include factitial dermatitis, granulomatous conditions, infectious diseases, malignancy, and pyoderma gangrenosum. Most of these causes can be ruled out by microbiological studies and histological analysis. Like TTS, factitial dermatitis is also caused by self-manipulation of the ulcer but unlike TTS these patients do not demonstrate underlying neurological damage and psychiatric symptoms are instead predominant.

Treatment of TTS requires behavior modifical and physical protection of the nasal ulcer from self-manipulation. Pharmacological treatment such as amitriptyline, carbamazepine, chlorpromazine, diazepam, pimozide, and clonazepam should be considered to decrease paresthesias and compulsive behavior. Early identification of this syndrome is vital in order to rule out needless surgical intervention and to provide proper therapy. It is probable that our patient could have received treatment for TTS earlier on in its course if not for the global health crisis caused by SARS-CoV-2.

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