Trigeminal Trophic Syndrome: A rare cause of nasal ulceration and successful repair with a paramedian forehead flap

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Abstract
Trigeminal trophic syndrome (TTS) is a less commonly encountered medical condition that will likely present to otolaryngologists and facial plastic surgeons. TTS results in facial ulceration, often in the alar region, after damage to the trigeminal nerve or its central sensory connections. Only a handful of cases have been reported in otolaryngologic literature, but it is important that practicing otolaryngologists be familiar with the classic signs of symptoms of this condition to avoid delays in diagnosis and treatment.

Introduction
Trigeminal trophic syndrome is a relatively rare condition. The disease process involves self-manipulation/destruction of the nasal region following stroke, denervation of the trigeminal nerve, or treatment of trigeminal neuralgia. The clinical features of the disease typically involve one or more painless, crescent-shaped ulcers in the alar region. More severe cases can involve tissue loss and can spread to the cheek and upper lip. Some of the proposed treatments include neurological evaluation, behavioral modification, medical management, and surgical repair. Despite treatment with these multiple modalities, the ulcers frequently recur and pose a dilemma for clinicians.

Case Report
A 49-year-old woman initially presented to the facial plastics clinic on 10/5/2006 with severe right alar retraction and partial destruction of the alar rim. She first noted the retraction in 1999, following a vertebral artery dissection that left her with decreased sensation in the right side of her face. Her nasal symptoms followed an indolent course over the following years with episodes of bleeding and ulceration on the right ala. Eventually, the area expanded to include the medial right cheek and subsequently led to destruction of the right alar rim.

Through clinical consultations, the diagnosis of TTS was reached. She then underwent a three-stage PMFF repair and a fourth stage for repair of vestibular stenosis.

The long-term results have been good. The patient was most recently seen in the office twenty months from her original presentation and six months from her nasal stenosis repair and was maintaining good aesthetic form, contour, and functional airflow through that side of her nose. The ulceration had not recurred.

Presentation
Stage 1
3 weeks later the patient underwent debulking and cartilage graft placement.

Stage 2
After stage 2, the patient underwent a standard division and inset procedure 3 weeks later.

Stage 3 (no photos)
TWO months following stage 3, the patient underwent vestibular stenosis repair with an intravestibular thermoplastic splint and external PTFE tie-over bolster for 2 weeks.

Stage 4 - Stenosis Repair

Discussion
Trigeminal trophic syndrome was first described by Lowman and McKhann in 1933. The lesions are complications of trigeminal anesthesia that occurs following damage to the sensory root of the trigeminal nerve with subsequent self-denervation and progressive ulceration to the affected dermatome. This is most commonly seen after trigeminal ablation treatment for trigeminal neuralgia; however, it can occur with cerebral vascular disease, acoustic neuromas, herpes zoster ophthalmicus, to name a few.

The typical clinical picture of TTS is one or more crescent-shaped ulcers on the nose. The differential diagnosis of TTS is broad and can reach a definitive diagnosis largely involves excluding other causes of facial ulceration. These include basal cell carcinoma, squamous cell carcinoma, herpetic ulceration, syphilis, lepromy, Wegener’s granulomatosis, and other less common causes.

These lesions are persistent and are often a challenge to treat. A multidisciplinary approach should be utilized and may include a neurological evaluation, pain management, psychological management to assist with the reduction of self-manipulation, medical treatment, and surgical repair. A crucial element of treatment involves educating the patient as to the underlying causes of the ulcers and convincing him/her that the lesions are self-induced. Surgical treatment has previously shown good long-term results; however, this has been scarcerly reported in otolaryngologic literature and the successful repair using a PMFF has only been described once, over 25 years ago.

We present a patient with trigeminal trophic syndrome that underwent successful treatment with a paramedian forehead flap. This should serve to increase the awareness of the syndrome among otolaryngologists so that accurate diagnoses can be reached and proper treatment administered in this rare and difficult-to-treat condition.

References