Trigeminal Trophic Syndrome. Presentation of a Case in its Early Stages

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INTRODUCTION

The Trigeminal Trophic Syndrome is a rare condition characterized by anesthesia, paresthesia and self provoked nasal ulceration. The syndrome arises after damage, induced or not, to the sensory part of the trigeminal root or nerve. After the nerve is damaged the patient experiences hemifacial anesthesia and paresthesias, mainly itching and burning that causes continuous touching, rubbing and picking to relieve the perceived paresthesias. The repetitive trauma leads to an ulcer. If untreated or misdiagnosed the ulcer may grow and cause disfigurement.

CASE PRESENTATION

A 43 year old female presented to the office with a nasal ulcer in her left nasal vestibule. (fig 1) She had a history of left trigeminal neuralgia and 6 months prior to her visit she underwent thermocoagulation of the left V2 and V3. 2 weeks after surgery she started experiencing an intense itching and a burning sensation inside and around her left nostril. She has been treated with mupirocin ointment without any results. After ruling out malignancy, vasculitis and infection we confirmed the diagnosis of Trigeminal Trophic Syndrome. Treatment included resection of the ulcer and an island flap to cover the defect (fig 2,3), carbamazepine to reduce the paresthesias and most important education of the patient to make her realize that the ulcer was self inflicted. Histologically the report was of a chronic ulcer with minimal inflammatory infiltrate. One year after repair there is no recurrence (fig 4). The patient is still taking carbamazepine on and off. The paresthesias are still there although less intense. She reminds herself everyday not to touch rub or pick her face.

DISCUSSION

The Trigeminal Trophic Syndrome is a rare condition characterized by the triad of facial anesthesia, facial paresthesia and ulcerations of the nose adopting a crescentic shape.

It was first described by Wallenberg in 1901 (1) and subsequently by Loveman (2,4) and Mckenzie (3,4) The clinical presentation is that of a patient that presents with unilateral facial anesthesia, unilateral facial paresthesias and unilateral facial ulcers most commonly after surgical treatment for trigeminal neuralgia(4) or suffered a stroke (5), neoplasia, trauma or viral infection (1). The patient refers a burning and itching sensation that leads to repetitive touching rubbing and picking of the affected area that provokes tissue loss and formation of an ulcer. Most commonly the patient is unaware that the ulcers are self inflicted.

The differential diagnosis for a non healing facial ulcer includes basal cell carcinoma, squamous cell carcinoma, syphilis, leprosy, fungal, mycobacterial or parasitic infections, Wegener’s granulomatosis and T-cell lymphoma.

Once the diagnosis is made the treatment is multidisciplinary and encompasses patient education making the patient aware that the lesions are self inflicted, surgical treatment for repair of the defects and medical treatment such as carbamazepine (7,8) to relieve the paresthesias. Other medical treatments have been used with mixed results such as Vitamin B, Diazepam, amitriptyline, chlorpromazine, acyclovir and injected triamcinolone (4).

CONCLUSIONS

The Trigeminal Trophic Syndrome is a rare entity that can be encountered by otoaryngologists. The unfamiliarity with the condition leads to misdiagnosis and delay of treatment that can result in disfigurement. Treatment encompasses surgical reconstruction, medical therapy and education of the patient.

REFERENCES

2. Loveman AB an unusual dermatosis following section of the fifth cranial nerve. Arch Dermatol Syph. 1933;28:369-375