Self-induced Nasal Ulceration From Trigeminal Trophic Syndrome

Joshua E. Lane, MD; Steven Deliduka, MD

Trigeminal trophic syndrome is a rare entity characterized by the presence of ala nasi ulceration, trigeminal anesthesia, and paresthesia. It arises secondary to trigeminal nerve injury. Patients with this diagnosis often undergo repeated skin biopsies to rule out suspected nonmelanoma skin cancer due to the clinical appearance. Recognition of this entity is paramount to avoid unnecessary surgical intervention and to attempt therapy.

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rigeminal trophic syndrome is a rare condition characterized by a triad of ala nasi ulceration, trigeminal anesthesia, and paresthesia. ¹⁻³ It occurs following damage to the trigeminal nerve and may be a challenge to diagnose because of suspected nonmelanoma skin cancer due to the clinical appearance. Accurate diagnosis is imperative to avoid unnecessary surgical intervention.

Case Report

An 83-year-old woman presented to our outpatient dermatology clinic for evaluation of a progressive unilateral ulceration of the right ala nasi with secondary involvement of the upper lip of 2 years' duration (Figure). She reported that the ulceration was pruritic and she denied any pain. A home care provider and the patient's daughter reported observing the patient self-manipulate the ulceration. Her medical history was notable for hypertension,

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Dr. Lane is from the Division of Dermatology, Departments of Internal Medicine and Surgery, Mercer University School of Medicine, Macon, Georgia, and the Department of Dermatology, Emory University School of Medicine, Atlanta, Georgia. Drs. Lane and Deliduka are from the Division of Dermatology, Department of Medicine, Medical College of Georgia, Augusta.

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Correspondence: Joshua E. Lane, MD, 2301 Brookstone Centre Pkwy, Suite 200, Columbus, GA 31904 (jlanemd@gmail.com).

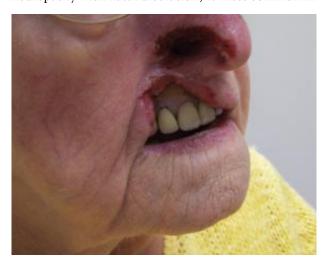
cardiovascular disease, and dementia secondary to transient ischemic attacks.

Physical examination revealed unilateral full-thickness ulceration of the right ala nasi as well as the upper lip and vermilion border. The ulceration approximated the innervation boundaries of the zygomaticofacial and infraorbital nerves. A characteristic "sneer" and sparing of the nasal tip were noted. A skin biopsy was performed and histology demonstrated focal epithelial hyperplasia secondary to external trauma and confirmed the absence of a neoplastic process. Tissue cultures for bacteria, fungus, and atypical mycobacteria were negative.

The patient and her family were educated regarding the diagnosis. Systemic therapy was recommended but declined due to the patient's other medical and behavioral issues. A 3-month trial of tacrolimus ointment 0.1% twice daily failed to provide a remarkable symptomatic or clinical response.

Comment

Trigeminal trophic syndrome (previously reported as trigeminal neurotrophic ulceration and trigeminal neuropathy with nasal ulceration) is most common in



Ulceration of right ala nasi with secondary involvement of the upper lip.

women and the elderly. Most commonly, it involves the ala nasi but also can affect other trigeminal innervation regions.^{1,4} Trigeminal nerve injury typically is either iatrogenic (eg, trigeminal nerve ablation for trigeminal neuralgia) or secondary to a cerebrovascular accident.³ The ulceration occurs from self-induced trauma caused by sensory alteration. The time frame from trigeminal nerve injury to ulceration ranges from weeks to decades.

Diagnosis of this entity is clinical. Cutaneous biopsies often are performed to confirm the absence of a cutaneous neoplasm such as basal or squamous cell carcinoma. Ulceration occurs in noncartilaginous areas, often producing a characteristic crescentic shape, allowing for differentiation from other neoplastic causes. The nasal tip is spared because of its innervation by the anterior ethmoidal branch of the ophthalmic division of the trigeminal nerve. Involvement beyond the ala nasi to the upper lip and/or cheek may result in a characteristic sneer, as seen in our patient. This tologic examination reveals chronic ulceration with minimal inflammation.

Treatment of trigeminal trophic syndrome is challenging. Patient education is critical, especially in those patients lacking the insight of self-manipulation. Protective prostheses may be utilized, both to conceal the ulcer and to shield it from further manipulation. Similarly, meticulous fingernail hygiene can help prevent secondary infection. Systemic and topical antibiotic treatments may be utilized as necessary. A multitude of

treatment modalities have been used with variable success, including amitriptyline, carbamazepine, chlorpromazine, citalopram, clonazepam, diazepam, gabapentin, and pimozide.³ In our experience, tacrolimus ointment 0.1% has been beneficial in some patients with other neurodermatoses but provided little benefit for our patient. Other interventions that have been used include transcutaneous electrical neural stimulation, iontophoresis and nerve blockade, radiation, ipsilateral cervical sympathectomy, stellate ganglionectomy, and surgical reconstruction of the ulceration.

Trigeminal trophic syndrome is a rare cutaneous manifestation of underlying trigeminal nerve injury. Treatment requires a multidisciplinary approach.

REFERENCES

- 1. Racette AJ, Moore A, Brown S, et al. Recognizing trigeminal trophic syndrome [letter]. *J Am Acad Dermatol*. 2006;55:359-361.
- 2. Tollefson TT, Kriet JD, Wang TD, et al. Self-induced nasal ulceration. *Arch Facial Plast Surg.* 2004;6:162-166.
- Monrad SU, Terrell JE, Aronoff DM. The trigeminal trophic syndrome: an unusual cause of nasal ulceration. J Am Acad Dermatol. 2004;50:949-952.
- 4. Sadeghi P, Papay FA, Vidimos AT. Trigeminal trophic syndrome—report of four cases and review of the literature. *Dermatol Surg.* 2004;30:807-812.
- 5. Weintraub E, Soltani K, Hekmatpanah J, et al. Trigeminal trophic syndrome: a case and review. *J Am Acad Dermatol*. 1982;6:52-57.