CASE REPORT

Trigeminal Neurotrophic Ulceration

Ahmed El-Daly, M.D., and Carl H. Snyderman, M.D.

ABSTRACT—A 74 year-old female developed a trigeminal neurotrophic ulcer (TNU) 20 years following surgical ablation of the trigeminal nerve. The diagnosis of this unusual disorder is suggested when an ulcerative lesion develops in the ala nasi in a patient with trigeminal sensory loss. A history of self-induced trauma to that area and some form of mental impairment further support the diagnosis. Exclusion of basal cell carcinoma and other causes of facial ulceration is mandatory before the diagnosis of TNU is established.

INTRODUCTION

Trigeminal neurotrophic ulceration (TNU) is an uncommon but highly recognizable complication of trigeminal sensory loss of the face, also known in the medical literature as trigeminal trophic syndrome,¹ ulceration en arc,² and trigeminal neuropathy with nasal ulceration.³ The most common etiological factor (75% of cases) is trigeminal surgical ablation by rhizotomy or alcohol injection in the gasserian ganglion for the treatment of trigeminal neuralgia. Less common etiological factors include vertebrobasilar insufficiency,⁵ syringobulbia, acoustic neuroma,⁶ and postencephalitic parkinsonism.⁷

CASE REPORT

A 74-year-old white female presented in June 1993 with repeated epistaxis, a crescentic ulcerative lesion of the right ala nasi, and a superficial erosive lesion of the frontal scalp with sensory loss in the distribution of the right trigeminal nerve. Past medical history was remarkable for hypertension and organic brain syndrome with dementia. She also had surgical repair of an aortic aneurysm and multiple procedures for the management of trigeminal neuralgia. Her husband noted that she consistently picked her nose on the right side.

A biopsy of the sinonasal mucosa showed ulcerated respiratory mucosa with granulation tissue and focal squamous metaplasia (Fig. 1). Important negative findings included an absence of granulomas, vasculitis, ab-

normal lymphoid cells, infectious organisms, and birefringent material.

Healing of the scalp lesion was achieved with local wound care, but the patient continued picking her nose and was admitted in December 1993 with periorbital cellulitis and dacryocystitis for which she was managed medically. In February 1995 she presented again with more destruction of the ala nasi and the upper lip with loss of the medial orbital tissue and marked visual deterioration (Fig. 2).

Nasal endoscopy revealed destruction of the inferior and middle turbinates, anterior and posterior ethmoid sinuses, anterior sphenoid wall, and medial orbital wall. These findings were confirmed by computed tomographic (CT) scan. There was no evidence of acute inflammation or granulation surrounding the lesion (Fig. 3).

It was thought that the defect must be protected to avoid further trauma. External gauze dressings were ineffective in preventing self-induced trauma. An extraorally fixed appliance secured with a multiplane external skeletal fixation device was used. Nine months later the patient was tolerating her osseointegrated prosthesis, and the edges of the lesion were well-healed with no evidence of further trauma (Figs. 4 and 5).

DISCUSSION

Trigeminal neuralgia is a relatively common disorder in the elderly. Permanent relief of the spasmodic pain that

Figure 1. (A) Ulcerated respiratory mucosa with focal squamous metaplasia. Hematoxylin and eosin, $\times 200$. (B) Mature granulation tissue deep to ulcerated surface. Hematoxylin and eosin, $\times 200$.

usually accompanies this disorder can be achieved by cutting the sensory root of the trigeminal nerve or alcohol injection of the gasserian ganglion. Relief of pain, however, leaves the patient with complete loss of temperature and light touch sensation and frequently paraesthesia in the distribution of the ipsilateral trigeminal nerve. The development of facial ulceration following sensory denervation of the face was first described as a clinical entity in 1933 independently by Loveman⁸ and McKenzie.⁹

Since then, many authors described chronic facial ulceration, mainly at the region of the ala nasi following sensory impairment of the trigeminal nerve. Weintraube et al⁴ analyzed 63 cases of TNU and found a mean age at presentation of 75 years and a female to male ratio of 2.2–1. The true incidence of TNU is difficult to esti-

mate.¹⁰ Henderson¹¹ found that ulceration of the face occurred in 18% (12/66) of patients after alcohol injection into the gasserian ganglion, and he noted that ulcers appeared at the alar margin from 1 to 6 months after the surgical procedure. However, TNU has been reported to occur up to 10 years after the trigeminal insult.¹²

Pathogenesis

The development of TNU is always linked to some form of trauma to the face, which is usually self-inflicted. Paraesthesia causes the patient to consistently scratch, rub, or pick the skin of the face. With advanced age and cognitive impairment, as seen in our patient, the

Figure 2. Characteristic trigeminal neurotrophic ulcer: crescentic-shaped defect with loss of ala nasi. Continued self-induced trauma resulted in loss in the area of the medial canthus, periorbital cellulitis, and dacryocystitis.

self-mutilation becomes a kind of habitual or compulsive behavior, which leads to persistence of the facial ulcer. It is postulated that destruction of the gasserian ganglion results not only in facial sensory loss, but autonomic nervous dysfunction as well, as the sympathetic fibers from the carotid plexus that normally link to the trigeminal are also interrupted, leading to sympathetic hypotonia in the region of the face. The resulting vasodilation will result in constant cooling of the skin and slows venous return, creating unfavorable conditions for the metabolic processes related to wound healing.¹³

Clinical Presentation

The nasal ulcers in TNU are characteristically crescentic with loss of the ala nasi corresponding to areas of the nose that lack cartilage ("ulceration en arc").² In more severe cases, scarring and tissue loss in the upper lip and the adjacent area will cause retraction of the mucocutaneous junction, giving a characteristic "sneer."⁴ Only 13% of the cases reported by Weintraube et al⁴ developed ulceration in the forehead, scalp, ear, palate, or jaw.

Bauer¹³ defined the limitation of the area affected by ulceration to the ala of the nose as the border area between the first and second divisions of the trigeminal innervation and stated that the inequality or the different degree of the denervation is what makes the subjective

feeling of itching or paraesthesia to be more intense at this area.

Differential Diagnosis

Although the clinical presentation of TNU is often characteristic, a biopsy is usually indicated to exclude other diseases, especially basal cell carcinoma, which is characterized by nests of proliferating basaloid cells with distinct peripheral palisading. Other less common causes of facial ulceration include Wegener's granulomatosis, polymorphic reticulosis (also known as angiocentric T-cell lymphoma, nasal T-cell lymphoma, and Stewart's type of lethal midline granuloma), granulomatous infections, and cocaine abuse. Wegener's granulomatosis is characterized by a histological triad of vasculitis, necrosis, and necrotizing granulomas. The accompanying mixed inflammatory reaction consists of lymphocytes, histiocytes, and, less commonly, polymorphonuclear leukocytes and eosinophils. Polymorphic reticulosis consists of a polymorphic infiltrate of small lymphocytes, plasma cells, histiocytes, immunoblasts, and, unlike Wegener's, atypical mononuclear cells with a tendency to invade blood vessel walls. Subsequent studies have shown that most cases are T-cell lymphomas, hence the current term angiocentric T-cell lymphoma.^{15,16} Granulomatous infections are recognized by using special bacterial and fungal stains to identify the causative organisms, while cocaine abuse is suggested by the presence of a giant cell reaction to polarizable foreign material and an absence of vasculitis.

Treatment

Various treatment modalities have been used in an attempt to heal these ulcers. Ipsilateral cervical sympathectomy has resulted in healing of ulceration in a few patients.⁹ Loveman⁸ used radiation therapy to successfully treat his patient. Westerhof and Bos¹⁷ treated a patient successfully with transcutaneous electrical stimulation. The value of these treatment modalities cannot be established due to the rarity of the cases and the potential for misdiagnosis. Consequently, prevention of continued trauma and control of secondary bacterial infection continues to be the mainstay of treatment.

Surgical therapy may play a role, and patients might benefit from closing the defect using sensate flaps from the contralateral (unaffected) side.^{18,19} The use of a protective device that covers and prevents manipulation of the affected area may be helpful. A prosthesis, however, could be complicated and tedious for the elderly, especially in the presence of psychotic problems, which are frequently associated with this syn-

Figures 3(A-D). CT scans demonstrate destruction of the medial wall of the maxilla, ethmoids, anterior wall of the sphenoid, and lamina papyracea. Note the presence of an ipsilateral temporal craniectomy from previous surgical procedures to the gasserian ganglion.

Figure 4. Extraoral osseointegrated implant in well-healed lesion.

Figure 5. The patient wearing her osseointegrated prosthesis.

drome.^{13,20} As a result, treatment is difficult and disappointing in most cases.

REFERENCES

1. Finlay AY: Trigeminal trophic syndrome. *Arch Dermatol* 115:1118–1123, 1979
2. Jaeger H: New type of neurotrophic ulcer of the ala nasi following retrogasserian neurectomy. *Dermatologica* 100:201–205, 1950
3. Spillane JD, Urich H: Trigeminal neuropathy with nasal ulceration: Report of two cases and one necropsy. *J Neural Neurosurg Psychiatry* 39:105–113, 1976
4. Weintraube E, Soltani K, Hekmatpanah J, et al: Trigeminal trophic syndrome. *J Am Acad Dermatol* 6:52–57, 1982
5. Haensch R: Neurotrophic ulcer associated with the Wallenberg syndrome. *Acta Dermatol Venereol (Stockh)* 48:622–629, 1968
6. Karnosh LJ, Scherb RF: Trophic lesions in the distribution of the trigeminal nerve. *JAMA* 115:2144–2148, 1940
7. Rosenberg SJ, Solovay J: Trophic ulcer following encephalitis lethargica. *Arch Dermatol Syph* 39:825–829, 1939
8. Loveman A: An unusual dermatosis following section of the fifth cranial nerve. *Arch Dermatol Syph* 28:369–375, 1933
9. McKenzie KG: Observations on the results of the operative treatment of trigeminal neuralgia. *Can Med Assoc J* 29:492–496, 1933
10. Freeman AG: Trigeminal neuralgia: complications of its surgical treatment. *Br Med J* 1:631–632, 1967
11. Henderson WR: Trigeminal neuralgia: The pain and its treatment. *Br Med J* 1:7–15, 1967
12. Walton S, Kecezkas K: Trigeminal neurotrophic ulceration—A report of four patients. *Clin Exp Dermatol* 10:485–490, 1985
13. Bauer F: Trophic ulceration of the ala nasi following trigeminal denervation. *J Laryngol Otol* 80:422–425, 1966
14. Howell JB: Neurotrophic changes in the trigeminal territory. *Arch Dermatol* 86:442–449, 1962
15. Ho FCS, Choy D, Loke SL, et al: Polymorphic reticulosis and conventional lymphomas of the nose and upper aerodigestive tract: A clinicopathologic study of 70 cases, and immunophenotypic study of 16 cases. *Hum Pathol* 21:1041–1050, 1990
16. Medeiros LJ, Peiper SC, Elwood L, et al: Angiocentric immunoproliferative lesions: A molecular analysis of eight cases. *Hum Pathol* 22:1150–1157, 1991
17. Westerhof F, Bos JD: Trigeminal trophic syndrome: A successful treatment with transcutaneous electrical stimulation. *Br J Dermatol* 108:601–604, 1983
18. Abyholm FE, Eskeland G: Defect of the ala nasi following trigeminal denervation. Case report. *Scand J Plast Reconstr Surg* 11:87–90, 1977
19. McLean NR, Watson ACH: Reconstruction of a defect of the ala nasi following trigeminal anaesthesia with an innervated forehead flap. *Br J Plast Surg* 35:201–203, 1982
20. Miglets AW: Factitial dermatitis following trigeminal denervation. *Arch Otolaryngol* 88:196–171, 1968