Nonpainful Ulcerations on the Nose and Forehead

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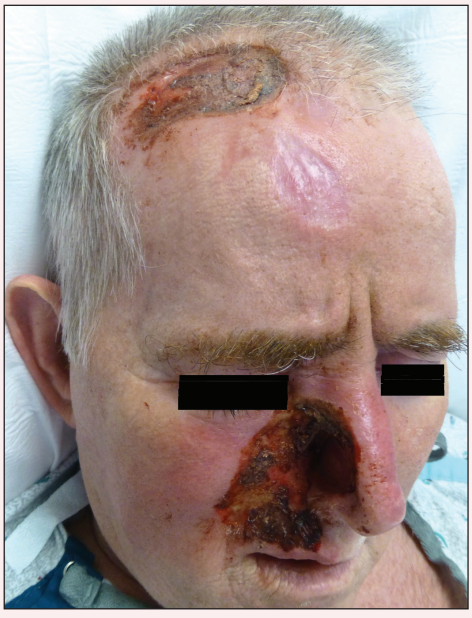

A 57-year-old man with a history of multiple cerebrovascular accidents was transferred from an outside hospital to our inpatient rheumatology service with nonpainful erosions of the forehead and nasal ala of 6 months' duration. The patient reported that he initially developed a sore on the nose months prior to presentation with worsening sensations of itching and tingling on the forehead and nose. He also noted a headache and gradual loss of vision in the right eye. The patient was immunocompetent and denied arthralgia or any other skin lesions.

What's the diagnosis?

a. basal cell carcinoma
b. histoplasmosis
c. leishmaniasis
d. pyoderma gangrenosum
e. trigeminal trophic syndrome

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The Diagnosis: Trigeminal Trophic Syndrome

Trigeminal trophic syndrome (TTS) is a rare condition occurring after injury to the sensory roots of the trigeminal nerve. Trigeminal trophic syndrome was first described in 1933 by Loveman[1] as a complication of ablative treatment of trigeminal neuralgia; however, it has been observed with lesions to the central or peripheral nervous system that damage sensory components of the trigeminal nerve. In our patient, the cause was likely an infarction of the posterior inferior cerebellar artery, which supplies both the cerebellum and the trigeminal nuclei in the brain stem.

Other possible causes of TTS include injury from herpes zoster ophthalmicus; vertebrobasilar insufficiency; trauma; *Mycobacterium leprae* neuritis; spinal cord degeneration; syringobulbia; or tumors such as astrocytomas, meningiomas, and acoustic neuromas.[2] The most typical presenting manifestation is a unilateral, crescent-shaped ulceration of the nasal ala, specifically where cartilage is lacking.[3] Less frequently, it affects the upper lip, cheeks, forehead, scalp, and ear. It characteristically spares the nasal tip, which derives sensory innervation from the medial branch of the anterior ethmoidal nerve.[4] The affected dermatomal distribution can show anesthesia or paresthesia, promoting the urge to touch, pick, or rub the area.

Histology of the TTS lesion is nondiagnostic, usually showing a mixed dermal inflammation without evidence of infection, vasculitis, or malignancy. In our patient, Gram stain, Grocott-Gomori methenamine-silver stain, and immunohistochemical stains to rule out leukemia or lymphoma were all negative. In addition, flow cytometry of the forehead tissue also was normal. Tests for human immunodeficiency virus, herpes simplex virus types 1 and 2, hepatitis, syphilis, histoplasmosis, blastomycosis, coccidioides, and tuberculosis were negative. During the biopsy, the patient was noted to have anesthesia of the skin. He also admitted to facial manipulation and was noted to have blood and tissue under the fingernails on mornings when the lesion was left uncovered overnight.

Treatment of TTS often can be difficult, especially if patients do not admit to wound manipulation or if manipulation occurs during sleep. Prevention of further ulceration can sometimes be achieved by occlusive dressing alone with mupirocin. Physical barriers can be supplemented with medications such as amitriptyline, carbamazepine, diazepam, chlorpromazine, and gabapentin to attempt to reduce paresthesia.[2] Psychiatric consultation can sometimes be necessary and helpful.[3] Additionally, negative pressure wound therapy has been used with success in the pediatric setting when digital manipulation is difficult to control.[5] More aggressive treatments include transcutaneous electrical stimulation, stellate ganglionectomy, radiotherapy, and innervated rotation flaps.[6,7]

In summary, recognition of this relatively rare cause of unilateral facial erosions is critical, both to prevent counterproductive treatments such as steroids and to initiate measures to prevent further trauma to the lesion.

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