

Trigeminal Trophic Syndrome

Ryan Owen DO

Lehigh Valley Health Network, Ryan.Owen@lvhn.org

Joshua M. Levin MD

Lehigh Valley Health Network, Joshua_M.Levin@lvhn.org

Stephen M. Purcell DO

Lehigh Valley Health Network, Stephen.Purcell@lvhn.org

Follow this and additional works at: <http://scholarlyworks.lvhn.org/medicine>



Part of the [Dermatology Commons](#), and the [Medical Sciences Commons](#)

Published In/Presented At

Owen, R., Levin, J., & Purcell, S. (2013, September 19-21). *Trigeminal Trophic Syndrome*. Poster presented at: The LVHN Resident's Research Day, Allentown, PA.

This Poster is brought to you for free and open access by LVHN Scholarly Works. It has been accepted for inclusion in LVHN Scholarly Works by an authorized administrator. For more information, please contact LibraryServices@lvhn.org.

Trigeminal Trophic Syndrome

Ryan Owen, D.O., Josh Levin, M.D., Stephen Purcell, D.O.

Lehigh Valley Health Network, Allentown, Pennsylvania and Philadelphia College of Osteopathic Medicine, Philadelphia, Pennsylvania

Case Presentation:

History of Present Illness: A 50 year old Caucasian male presented with an approximate 4 month history of left sided facial pruritus and paresthesias that started 2 weeks following an ischemic stroke with subsequent unilateral facial ulcers. He reported regularly picking and scratching the affected areas. According to MRI results at the time, the left cerebellum and medulla were affected in the region of the left vertebral basilar artery. Patient reported mild motor dysfunction and paresthesias on the right side of his body.

Medical History/Surgical History: Hypertension, hyperlipidemia, chronic back/neck pain, finger surgery

Family History: Father hypertension

Medications: Aspirin, metoprolol, pravastatin, lorazepam, tramadol

Current Treatment: Mupirocin ointment, petrolatum, benzoyl peroxide wash

Physical Examination: Well demarcated angulated crusted ulcers located on the left ala, cutaneous upper lip, medial cheek, forehead, and scalp. There is scarring alopecia of the left frontal scalp with milia. Diminished tactile sensation of the left side of face in the V1 and V2 distribution of the trigeminal nerve is appreciated.

Laboratory Data: Gram stain, AFB, and fungal stains all negative

Biopsy: *Advanced Dermatology Associates, Ltd. (AD13-00946, 1/22/2013)* Left nasolabial fold: "Ulceration with fibrosis and inflammation."

Discussion:

Trigeminal trophic syndrome (TTS) is a rare syndrome characterized by the triad of trigeminal anesthesia, facial paresthesia, and ulceration of the lateral nasal ala "ulceration en arc." This condition was originally described by Wallenberg in 1901. Peripheral or central injury to the trigeminal sensory nuclei or spinal trigeminal tract, ganglion, or peripheral nerve branches is thought to be the underlying pathogenesis. The most common cause is from sequelae following treatment for trigeminal neuralgia including alcohol injection, rhizotomy of the sensory component of the trigeminal nerve, or destruction of the Gasserian ganglion. An important central cause is cerebrovascular accident representing approximately 33% of cases.

Patients typically present with unilateral facial ulceration of the nasal ala with less common involvement of the lip, cheek, jaw, temple, forehead and scalp. Interestingly, the nasal tip is spared as its innervation is via the anterior ethmoidal nerve branch of the ophthalmic division of the trigeminal nerve. The ulcerations are self-induced and prompted by ongoing paresthesia and anesthesia. The ulcers have been reported to develop from 2 weeks up to 30 years following the inciting injury.

Histopathological findings of the ulcerations are non-specific and consist of an ulcer with lymphohistiocytic infiltrate without vasculitis, neoplasia or infection. Thus, the diagnosis of TTS is clinical in nature. Differential diagnoses for a non-healing facial ulcer include basal cell carcinoma, squamous cell carcinoma, syphilis, leprosy, yaws, mycobacterial, fungal or parasitic infections, T-cell lymphoma, Wegener's granulomatosis or pyoderma gangrenosum.

Treatment for trigeminal trophic syndrome is multifaceted and involves treatment of underlying paresthesias, behavior modification for prevention of self induced injury and if severe enough surgical intervention. Educating the patient on their primary role in the pathogenesis of this disorder is paramount. Self-mutilation may be discouraged through the use of protective gloves and protective dressings such as hydrocolloid or alginate preparations with adjunctive topical lidocaine. Another topical preparation that has shown success is trolamine/sodium alginate containing emulsion. Wound infection may be prevented with topical antibiotics. Medical treatments used with various success include carbamazepine, amitriptyline, pregabalin, gabapentin and pimezide. Less commonly, surgical reconstruction and transcutaneous electrical nerve stimulation may be used as treatment.

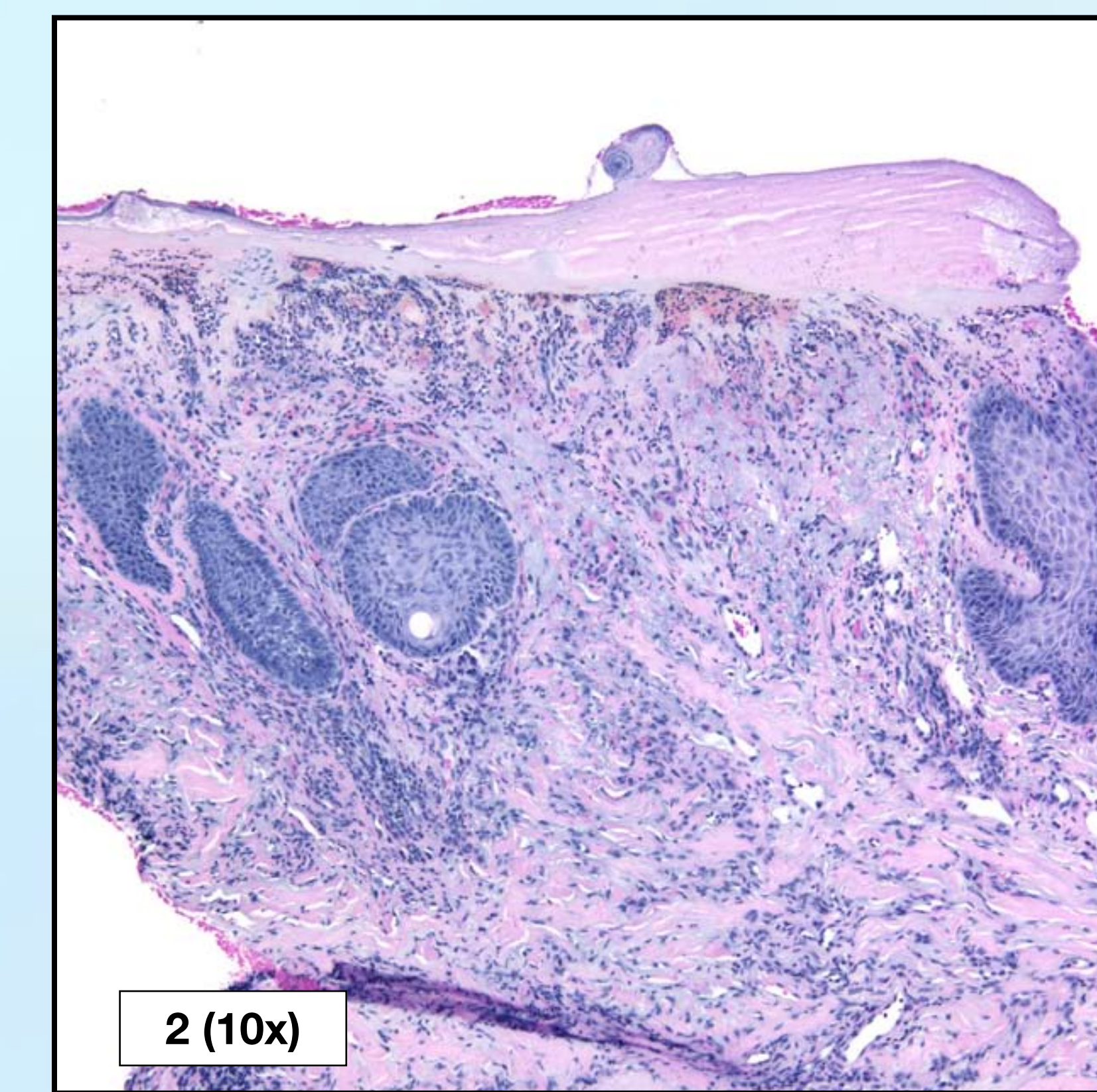


Figure 1a: Multiple ulcers on the scalp with evidence of milia and scarring alopecia

Figure 1b: Irregular angulated ulcers of the nasal ala, cutaneous upper lip and medial cheek. Note the classic sparing of the nasal tip

Figure 2: Histopathology showing ulceration with fibrosis and inflammation

References:

- 1 Curtis A, Oaklander A, Johnson A, Yosipovitch G. Trigeminal trophic syndrome from stroke: an under-recognized central neuropathic itch syndrome. *Am J Clin Dermatol* 2012;13(2):125-128.
- 2 Mishra S, Nayak C, Deshpande D, Pereira R. Trigeminal trophic syndrome: a rare entity. *Indian J Dermatol Venereol Leprol* 2011;77:729.
- 3 Fruhauf J, Schaidler H, Massone C et. al. Carbamazepine as the only effective treatment in a 52-year old man with trigeminal trophic syndrome. *Mayo Clin Proc* 2008;83(4):502-504.

© 2013 Lehigh Valley Health Network

A PASSION FOR BETTER MEDICINE.™

 **Lehigh Valley
Health Network**

610-402-CARE LVHN.org