Trigeminal trophic syndrome

The trigeminal trophic syndrome is an extremely rare cause of facial ulceration. It occurs as a consequence of trigeminal nerve damage or impaired central sensory connections.

The triad of unilateral crescentic nasal alar ulceration with anesthesia and paresthesia of the trigeminal dermatome (may present with severe pruritus and self-induced skin lesions from scratching). A result of trigeminal nerve injury [1)]

Trigeminal trophic syndrome (TTS) is a rare condition in which there is the involvement of the skin innervated by branches of the trigeminal nerve. Because of an alteration in the sensory function of the trigeminal nerve, an exaggerated manipulation of the skin by the patient occurs, with secondary ulcers in the affected areas. They are usually unilateral and located mainly at the beginning of the nose wing. There are very few publications in the current literature, so it is in the interest of doctors to know this rare pathology. [2)].

Treatment

Treatment of trigeminal trophic syndrome is challenging and often unsatisfactory, particularly in patients with cognitive impairment. Preston et al. reported the novel use of a thermoplastic dressing in two patients with trigeminal trophic syndrome. Use of the dressing resulted in successful healing of ulceration in both patients, which has been maintained in the short term, representing a simple and economical therapeutic option in the management of this difficult condition [3)].

The treatment has included: carbamazepine, diazepam, amitriptyline, chlorpromazine, clonazepam, or pimozide.

Jowett and Pineda summarize contemporary medical and emerging surgical approaches for the therapeutic management of this rare and devastating disease state [4)].

Trigeminal Trophic Syndrome Secondary to Refractory Trigeminal Neuralgia Treated with CyberKnife® Radiosurgery [5)].

Case reports

Case of trigeminal trophic syndrome secondary to endovascular treatment for internal carotid artery aneurysm [6)].

The case of a child who, shortly after undergoing suboccipital craniotomy for resection of a medullary cavernoma, developed corneal and conjunctival epithelial breakdown of the right eye with ipsilateral facial hypoesthesia as well as erosions and crusting of the eyelids, nostril, and lips on the right side. This combination of findings likely results from acute injury to the ipsilateral trigeminal ganglion, leading to acute neurotrophic keratitis and trigeminal trophic syndrome [7)].

Cervical trophic syndrome - a rare clinical variant of neural trophic syndrome [8)].

Trigeminal Trophic Syndrome Secondary to Refractory Trigeminal Neuralgia Treated with CyberKnife® Radiosurgery [9)].

A patient with multiple intraoral ulcerations of the right buccal and alveolar mucosa that had previously been treated with nerve ablation therapy for trigeminal neuralgia. Positive immunohistochemistry staining of a biopsy specimen for cytomegalovirus suggested a viral etiology; however, lesions persisted despite antiviral therapy, and immunohistochemistry was negative on follow-up biopsy. Diagnosis of TTS is one of exclusion as it can mimic many other conditions, and should be considered in patients with unilateral painless ulcerations with a history of trigeminal nerve damage [10)].

Acase of trigeminal trophic syndrome (TTS) that occurred as a complication of a neurosurgical procedure. Three years after a second surgical treatment for a meningioma of the cerebellopontine angle, this 32-year-old woman developed TTS with progressive skin ulcers on the left ala nasi and the left side of the forehead and chin. To the authors' knowledge, this is the first report of lesions in the dermatomes of all three branches of the nerve after a neurosurgical procedure. Early recognition of this disorder is important, as treatment is difficult and often unsatisfactory. Many clinicians are not aware of this disease, thus, it may be more common than previously thought. The importance of recognizing and diagnosing TTS, as well as its treatment, are discussed. [11)].

The skin area affected by neurotrophic ulceration is successfully treated with autologous cultivated epidermal cells. This form of tissue engineering is already a clinically established procedure for treating burns and chronic wounds. The results show for the first time that transplantation of in vitro cultivated epidermal cells can induce tissue regeneration and may be an effective tool in the treatment of neurotrophic ulcerations in the facial region. [12)].

Macrodactyly. Trigeminal trophic syndrome. Extramammary Paget's disease [13)].

A successful treatment with transcutaneous Electrostimulation [14].

1)
Nagel MA, Gilden D. The trigeminal trophic syndrome. Neurology. 2011 Oct 11;77(15):1499. doi: 10.1212/WNL.0b013e318232ac46. PMID: 21987642.
2) , 5) , 9)
Sallabanda K, Sallabanda M, Barrientos HD, Santaolalla I, Garcia R. Trigeminal Trophic Syndrome Secondary to Refractory Trigeminal Neuralgia Treated with CyberKnife® Radiosurgery. Cureus. 2020 Apr 14;12(4):e7670. doi: 10.7759/cureus.7670. PMID: 32419997; PMCID: PMC7226666.
3)
Preston PW, Orpin SD, Tucker WF, Zaki I. Successful use of a thermoplastic dressing in two cases of the trigeminal trophic syndrome. Clin Exp Dermatol. 2006 Jul;31(4):525-7. doi: 10.1111/j.1365-2230.2006.02125.x. PMID: 16716154.
4)
Jowett N, Pineda R 2nd. Corneal and Facial Sensory Neurotization in Trigeminal Anesthesia. Facial Plast Surg Clin North Am. 2021 Aug;29(3):459-470. doi: 10.1016/j.fsc.2021.03.011. PMID: 34217450.
6)
Minai L, Ogawa Y, Kinoshita M, Ito H, Kume M, Mitsui H, Shimada S, Kawamura T. Case of trigeminal trophic syndrome secondary to endovascular treatment for internal carotid artery aneurysm. J Dermatol. 2022 Feb 24. doi: 10.1111/1346-8138.16336. Epub ahead of print. PMID: 35212008.
7)
Lam BC, Saboo US, Kheirkhah A. Acute neurotrophic keratitis with trigeminal trophic syndrome after craniotomy. J AAPOS. 2020 Dec;24(6):376-379. doi: 10.1016/j.jaapos.2020.07.011. Epub 2020 Oct 22. PMID: 33228954.
8)
Koushk-Jalali B, Kuntz T, Tigges C, Gharevi NT, Wieland U, Kreuter A. Cervical trophic syndrome - a rare clinical variant of neural trophic syndrome. J Dtsch Dermatol Ges. 2020 Dec;18(12):1486-1487. doi: 10.1111/ddg.14171. Epub 2020 Jul 14. PMID: 32666622.
10)
Farahani RM, Marsee DK, Baden LR, Woo SB, Treister NS. Trigeminal trophic syndrome with features of oral CMV disease. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 2008 Sep;106(3):e15-8. doi: 10.1016/j.tripleo.2008.05.002. Epub 2008 Jul 7. PMID: 18602303.
11)
Luksić I, Luksić I, Sestan-Crnek S, Virag M, Macan D. Trigeminal trophic syndrome of all three nerve branches: an underrecognized complication after brain surgery. J Neurosurg. 2008 Jan;108(1):170-3. doi: 10.3171/JNS/2008/108/01/0170. PMID: 18173328.
12)
Schwerdtner O, Damaskos T, Kage A, Weitzel-Kage D, Klein M. Autologous epidermal cells can induce wound closure of neurotrophic ulceration caused by trigeminal trophic syndrome. Int J Oral Maxillofac Surg. 2005 Jun;34(4):443-5. doi: 10.1016/j.ijom.2004.08.012. PMID: 16053858.
13)
Vidimos AT, Lucas A, Warner M, Narurkar VA, Papay F, Bailin PL, Ghorbani AJ, Helm TM. Macrodactyly. Trigeminal trophic syndrome. Extramammary Paget's disease. Self-assessment examination of the American Academy of Dermatology. J Am Acad Dermatol. 1995 Jul;33(1):154-8; quizz 148-53. doi: 10.1016/0190-9622(95)90055-1. PMID: 7601940.
14)
Westerhof W, Bos JD. Trigeminal trophic syndrome: a successful treatment with transcutaneous Electrostimulation. Br J Dermatol. 1983 May;108(5):601-4. doi: 10.1111/j.1365-2133.1983.tb01063.x. PMID: 6601961.