

## CASE REPORT

# Trigeminal trophic syndrome associated with neuralgia after ischemic stroke—a case report

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## Abstract

Trigeminal trophic syndrome (TTS) is a rare condition resulting from self-manipulation of the skin after an injury to the trigeminal nerve. We describe the case of a 70-year-old woman who was hospitalized because of erythema, ulceration, and crusts on the right side of her face, accompanied by itching, burning, and irritation forcing self-manipulation of the skin. She had been previously repeatedly treated with antibiotics, acyclovir, and topical medications without any improvement. Her medical history revealed an episode of ischemic stroke in 2016, after which a deformation and partial atrophy of the right ala nasi appeared. Trigeminal neuropathy was confirmed by a neurological consultation. Diagnosis and treatment of TTS requires a multidisciplinary approach. Awareness of this disease should be raised to enable proper diagnosis and management of difficult-to-treat facial ulcers that cannot be explained by common causes.

## KEYWORDS

facial ulcers, ischemic stroke, trigeminal trophic syndrome (TTS), trophic disorders

## 1 | INTRODUCTION

Trigeminal trophic syndrome (TTS) is a rare condition resulting from self-manipulation of the skin after a peripheral or central injury to the sensory root of the trigeminal nerve. The syndrome consists of a classic triad of symptoms like trigeminal anesthesia, facial paraesthesia, and a crescentic lateral ulceration of the ala nasi.<sup>1</sup>

Loveman first described TTS in English literature in 1933 as a “carcinomatous – ulcer” on the forehead after ablation of the Gasserian ganglion.<sup>2</sup> However, it was first mentioned by Wallenberg in 1901 as a cutaneous ulceration in the trigeminal dermatome, when he delineated Wallenberg's syndrome leading to excoriation on the right

ala nasi.<sup>3</sup> So far, a little more than 100 cases have been described.

Many names have been used for TTS such as “trigeminal neurotrophic ulceration,” “trigeminal neuropathy with nasal ulceration,” and “trophic ulceration of the ala nasi.”<sup>4</sup> In the following article, we report a case of TTS following an ischemic stroke in a 70-year-old woman.

## 2 | CASE REPORT

A 70-year-old female patient was admitted to the Department of Dermatology, Sexually Transmitted Diseases and Clinical Immunology in Olsztyn in October

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2021 due to erythematous-inflammatory lesions of the right side of the face with signs of a secondary infection. In 2016, she had an ischemic stroke of the right cerebellar hemisphere and medulla oblongata. During hospitalization in the neurology department, a nasogastric tube was inserted and remained in place for about 6 weeks. After that, ulceration and destruction of the right nasal wing appeared. The first skin lesions, on the cheek and above the upper lip on the right side, appeared in 2017 in the form of small erythematous lesions and erosions covered with scabs. The patient has admitted to frequently touching and “picking at” the lesions due to the sensation of a sudden cold wave or numbness of the affected skin area. The affected skin area gradually expanded to the entire right side of the face, the auricle, and the adjacent scalp. The patient was repeatedly treated for this condition as shingles, bacterial skin infection or allergic eczema with acyclovir, antibiotics and topical anti-bacterial, anti-inflammatory drugs, and emollients without any effect. Allergic prick tests were negative. Her personal and family history denied skin and allergic diseases. The patient's chronic diseases included post-stroke right-sided cerebellar syndrome and Worster—Drought syndrome, hypertension, sick sinus syndrome, supraventricular arrhythmias, and frontal sinus osteoma. On admission to the clinic during clinical examination, right-sided involvement of the entire skin of the face, auricle and adjacent 3–4 cm of the scalp in the form of well-demarcated erythematous lesions with numerous erosions covered with scabs and accompanying swelling of the involved skin was visible (Figure 1A). In addition, destruction (partial loss) of the right nasal wing with superficial ulceration covered with scabs was present (Figure 1B). The conjunctiva of the right eye was congested; eyelids were swollen with the presence of abundant purulent discharge in the conjunctival sac (Figure 1A).

On clinical examination, lymph nodes of the submandibular region were enlarged. In laboratory tests, except for slightly elevated inflammatory parameters (C-reactive protein = 8.14 mg/dL, norm <5 mg/dL; erythrocyte sedimentation rate = 44 mm/h, norm <10 mm/h), there were no abnormalities. In addition, the blood glucose level was within normal range. A swab of the conjunctival sac and nasal wing ulcer cultured *Staphylococcus aureus* MRSA and topical treatment was initiated according to the antibiogram. The consulting ophthalmologist diagnosed purulent conjunctivitis with corneal erosion of the right eye. The neurologist diagnosed right trigeminal neuropathy recommending distigmine bromide (2 × 5 mg). In addition, topical treatment with detreomycin, ointment with benzocaine and menthol as well as emollients were applied, achieving a reduction of inflammatory skin lesions and healing of most erosions. Ophthalmic treatment (ofloxacin, moisturizing drops) resulted in the resolution of purulent lesions of the right eye; however, conjunctivitis persisted.

The pathomechanism of the skin lesions was explained to the patient, encouraging her to abstain from skin manipulation. The patient checked in at the clinic's dermatology outpatient clinic after 1 month of treatment with significant improvement due to the continuation of the treatment prescribed during hospitalization (Figure 2A,B).

### 3 | DISCUSSION

The exact etiology of TTS is not known. After trigeminal nerve damage, ulceration does not always occur. The causes of trigeminal nerve damage involved in cases of TTS are very diverse. Weintraub et al. after the review of 63 cases found that the majority of ulcers were iatrogenic



**FIGURE 1** (A) Clinical presentation at the initial visit. (B) Destruction of the right nasal wing with superficial ulceration covered with scabs.

**FIGURE 2** (A) An improvement after 1 month of treatment—side-face. (B) An improvement after 1 month of treatment—en-face.



and occurred after the destruction of the gasserian ganglion as therapy for trigeminal neuralgia, 29% after alcohol injection, and 46% after rhizotomy. TTS also appeared in association with other neurological conditions such as post-encephalic parkinsonism, syringobulbia or like in our patient's case—stroke.<sup>5</sup>

The period of latency between trigeminal injury and subsequent progressive ulceration in the affected dermatome ranges from a few weeks to decades.<sup>6</sup> Our patient developed skin lesions after 1 year. The syndrome is more common in women (four women for every three men) and the elderly (the most common in the sixth decade); however, this does not exclude the possibility of the disease even in infants.<sup>5,7</sup>

Although a classical triad of symptoms suggests TTS, other etiologic factors need to be excluded, such as infections, skin neoplasms, vasculitis, and granulomatous disease.<sup>5</sup> Chronic manipulation of the skin enhances bacterial superinfection of the surrounding tissues, which does not exclude the coexistence of TTS, which should not be overlooked in the differential diagnosis.

The right-sided trigeminal neuralgia occurs twice as often as the left-sided. The ulcers are mostly ipsilateral to the sensory changes.<sup>5</sup> Our patient developed lesions unilaterally; they occurred on her right side. The clinical presentation of ulcers is uniform. They are typically located at the ala nasi and less frequently in the adjacent areas of the cheek, upper lip, and forehead or less likely at other atypical locations.<sup>8</sup> The sparing of the tip of the nose has been attributed to its innervation by the medial nasal branch of the anterior ethmoid nerve.<sup>2</sup> Ulcers are usually located on one side; however, there was also a case of bilateral changes.<sup>9</sup> The pathophysiological mechanism of skin changes is central or peripheral

damage to the trigeminal nerve, which causes trigeminal dermatome anesthesia, paresthesia, dysesthesia, and post-stroke neuropathic pruritus. Decreased sensation with burning, itching, tickling, or crawling leads to disfigurement from repetitive painless picking and consequent development of persistent skin ulcers in particular facial areas (usually a maxillary branch of the trigeminal nerve).<sup>4</sup> The reason for this unusual predilection is unclear. On one side, there is a possibility that only a limited part of the ganglia is altered or damaged. Therefore, not the entire dermatome is included. Another possibility that some patients develop ulcers after experiencing paresthesia is that some have individual predispositions for self-manipulation and habitual scratching. Therefore, etiology is more likely to be traumatic than trophic origin.<sup>2</sup> Our patient also reported sensations of burning, itching, and tickling, which were provocative of skin manipulation. Sensory abnormalities in anesthesia, dysesthesia and atrophy of the nasal wing appeared immediately after the stroke. Ulcerations appeared throughout the right half of the face with a time of mechanical damage. Trigeminal trophic ulcers are typically described as triangular, crescent, or Y-shaped with bases covered by crusts, dried blood, and granulation tissue. The ala nasi is often destroyed. A fissure is sometimes seen in the floor of the nose.<sup>5</sup> Eyelids are often involved. It typically manifests as an ulceration with corneal exposure.<sup>10</sup>

Treatment of TTS requires a multidisciplinary approach (dermatology, neurology, psychiatry, ophthalmology, surgery). There is no proven standard treatment protocol. It is difficult to claim which approach is the best. Therefore, there is a huge need for further randomized controlled trials.

Therapies aim to target neuropathy through pharmaceutical interventions or surgery. Pharmaceuticals proven to positively reduce anesthesia and paresthesia in the literature include carbamazepine, amitriptyline, pimozide, chlorpromazine, diazepam, and donepezil. Behavior modification and local protectors are particularly important given the traumatic etiology of the disease. It is extremely crucial to educate the patient that they induce ulcer formation. It is also advisable to offer a patient measure to prevent excessive scratching such as keeping nails short, wearing gloves as often as possible, limiting habitual hand movements with a splint or covering the ulcers with a protective device during the night. Surgical interventions can target both the treatment of drug-resistant neuralgia and skin repair if irreversible skin changes occur. Transcutaneous electrical nerve stimulation, reconstruction with innervated grafts, sympathectomy, simple occlusion masks, stellate ganglionectomy, radiotherapy, and iontophoresis have been tried. In TTS therapy, it is extremely important to ensure that ulcers heal properly. To this end, the following were used: antibiotic therapy, vitamin B treatment, hydrocolloid dressings, or negative pressure wound therapy. If ocular lesions appear, local ophthalmic treatment is also essential. A new experimental method for the treatment of ulcers is the transplantation of in vitro-cultured epidermal cells.<sup>4,11</sup>

## 4 | CONCLUSIONS

In conclusion, we present a case of TTS to increase awareness of this uncommon syndrome. The diagnosis should be suspected when there is unilateral facial ulceration, especially involving the ala nasi associated with sensory impairment when other differential diagnoses have been excluded. Therapy should target the patient's education, neuralgia treatment, and wound healing. Increased recognition of TTS will help to promote a more accurate diagnosis and further treatment options. To increase homogeneity in the literature, it would also be good to standardize the name of the disease and use the most common term—TTS.

### AUTHOR CONTRIBUTIONS

**Joanna Rybak-d'Obyrn:** Conceptualization; data curation; formal analysis; investigation; methodology; resources; validation; visualization; writing – original draft. **Lewandowska Julia Alicja:** Conceptualization; data curation; formal analysis; investigation; methodology; resources; validation; visualization; writing – original draft. **Machoń Natalia Joanna:** Conceptualization; data curation; formal analysis; investigation; methodology; resources; validation; visualization; writing – original draft.

**Owczarczyk-Saczonek Agnieszka:** Conceptualization; supervision; writing – review and editing. **Placek Waldemar:** Conceptualization; supervision; writing – review and editing.

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None to declare.

### DATA AVAILABILITY STATEMENT

The authors confirm that the data supporting the findings of this study are available within the article.


### ETHICAL STATEMENT

Approval for a case report by the institutional ethics committee is not required.

### CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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