Trigeminal trophic syndrome as a late complication of sub-occipital vestibular schwannoma surgery: A case report and review of the literature

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Abstract

Introduction
Trigeminal trophic syndrome is a rare disease resulting from injury to the trigeminal nerve. It is characterised by par- and anaesthesia leading to self-manipulation and ulcerative lesions in the trigeminal area. Differential diagnoses are malignancies as well as infectious and autoimmune diseases. Our case presents a severe form of trigeminal trophic syndrome as a late complication of sub-occipital vestibular schwannoma extirpation requiring a multimodal interdisciplinary therapeutic approach.

Case report
An 84-year-old female patient presented to our clinic complaining of progressive loss of vision on the right eye. On physical examination, an extensive right-sided facial defect could be seen affecting the upper lip, cheek, ala nasi, the eyeball as well as the right forehead. Furthermore, after ophthalmologic consultation, right-sided amaurosis was diagnosed.

Conclusion
The patient’s lack of disease comprehension and absence of disease improvement despite our multimodal interdisciplinary treatment with antibiotics, carbamazepine and sertraline, warranted surgical intervention with wound debridement and simultaneous closure of the defect using an anterolateral thigh flap.

Introduction
The trigeminal trophic syndrome (TTS) was first described by the German neurologist Adolf Wallenberg in 1901 as a cutaneous trophic ulceration within the trigeminal dermatomes1. The first description in English was in 1933 by Loveman and McKenzie2. To date, over 100 case reports have been published in the scientific literature on TTS.

The TTS can arise after injury to the sensory trigeminal nuclei, the spinal trigeminal tract, the trigeminal ganglion or peripheral nerve branches2. Possible causes are nerve ablation for treatment of trigeminal neuralgia, cerebellar or brainstem infarcts, craniotomies or trauma as well as meningiomas, vestibular schwannomas or astrocytomas with or without surgery and herpes zoster infections among others. The damage results in ana- or dysesthesia and/or pruritus leading to repetitive self-manipulative behaviour2. Complaints may be aggravated by concomitant diabetes or alcoholism2,3.

The overall incidence of TTS is not known. TTS appears to be more common in the elderly with an average age of 57 years and a female to male ratio of 2.2:1.4-6. Time lag between trigeminal injury and disease onset may vary from weeks to decades5-7.11. Although the ala nasi appears to be the typical location, lesions may also develop in other areas supplied by the trigeminal nerve such as the scalp, forehead, temple, the ear, palate, the jaw and the upper lip1,2,7. Being solely innervated by the medial ethmoidal nerve, the tip of the nose is usually spared, resulting in a so-called ‘ulceration en arc’ with damage only to the non-cartilaginous alar region2.

The differential diagnosis of TTS is broad: malignant diseases such as squamous cell carcinomas, basal cell carcinomas, T-cell lymphomas as well as infectious diseases such as tuberculosis, syphilis, herpes simplex or varicella zoster virus have to be considered1,3,4. Furthermore, autoimmune diseases such as Wegener’s granulomatosis may mimic TTS and must therefore be serologically and historically excluded.

The following case report describes a patient with TTS to a pronounced extent.

Case Report
An 84-year-old female patient presented to our clinic complaining of progressive loss of vision on the right eye. Over 20 years ago, she had had a right-sided vestibular schwannoma extirpated via a sub-occipital approach with simultaneous ventriculoperitoneal shunt placement. Several years later, the patient started to complain of par- and hypoesthesia resulting in a habit of repetitive self-manipulation within the trigeminal area. The self-manipulating behaviour had been aggravated over the past years due to progress of a dementia syndrome.

On physical examination, an extensive right-sided facial defect could be seen affecting the upper lip, cheek, ala nasi, the eyeball as well as the right forehead. Furthermore, after ophthalmologic consultation, right-sided amaurosis was diagnosed.

All authors contributed to conception and design, manuscript preparation, read and approved the final manuscript.

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nasi, the eyeball as well as the right forehead (Figure 1). Furthermore, after ophthalmologic consultation, right-sided amaurosis was diagnosed. Computed tomography scans of the cranium and sinuses were performed, showing the trepanation defect after sub-occipital vestibular schwannoma extirpation as well as the ventriculo-peritoneal shunt and a reduction of brain matter. Furthermore, the extensive facial defect with concomitant osseous destruction of the nasal skeleton, the anterior wall of the maxillary sinus and the anterior orbital floor became evident (Figure 2A). The magnetic resonance imaging of the cerebellopontine angle, which was performed outside, displayed an extensive gliosis of the right cerebellar hemisphere could be seen (Figure 2B). Therapeutic approach was interdisciplinary and multimodal: After a gerontopsychiatric evaluation, the patient was initialised on medical therapy with sertraline and carbamazepine.

Simultaneously, local wound cleaning and an intravenous antibiotic therapy were performed for infection prophylaxis. Possibly aggravated by the dementia syndrome, self-manipulation continued despite conservative measures, leading to progress of the facial defect with complete destruction of the eyeball. For that reason and due to the high risk of infection with potential intracranial spread, it was decided that a surgical debridement with exenteratio orbitae be performed. For that reason and due to the high risk of infection with potential intracranial spread, it was decided that a surgical debridement with exenteratio orbitae be performed. Closure of the large surgical defect required a large tissue bulk. Therefore, a free vascular flap transfer was performed in collaboration with the department for plastic and reconstructive surgery using an anterolateral thigh ALT flap with vascular anastomosis to the ipsilateral facial artery (Figure 3). Due to the patients' age as well as the psychiatric co-morbidities resulting in lack of disease comprehension, we refrained from further plastic reconstructive measures. Pathological examination of the removed tissue showed chronic ulcerations with affected bulbus oculi without signs of malignancy. To protect the ALT flap, a facial epithesis was adjusted.

**Figure 1:** Extensive facial defect due to repetitive self-manipulation over years. Arrow (white) showing the ‘ulceration en arc’ with spared tip of nose.

**Figure 2:** (A) Computed tomography scan, coronal section, showing the bony defects. (B) Magnetic resonance imaging scan, T2-weighted, showing the cerebellar gliosis after vestibular schwannoma surgery via a sub-occipital approach.
Case report

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Figure 3: Patient after surgical wound debridement and defect closure using an anterolateral thigh flap.

Discussion

While bleeding, infection, facial paralysis, cerebrospinal fluid leaks, hearing loss, vertigo or paralysis of the caudal cranial nerves are very well known as complications of vestibular schwannoma surgery, TTS is a rarity. In 1982, Weintraub et al. investigated 63 cases of TTS, identifying several causes: 46% of patients had undergone trigeminal rhizotomy, 29% occurred after alcohol injections into the Gasserian ganglion, 9% were due to vertebrobasilar insufficiency, 5% due to postencephalitic Parkinson's disease, 2% due to syringobulbia and 5% due to surgery for vestibular schwannoma. To date, the exact incidence of TTS is unknown.

Treatment of TTS poses a great challenge because the exact underlying disease mechanisms are not fully understood. Several therapeutic options have been published in the literature but no therapeutic gold standard exists. Stopping the self-manipulating behaviour is paramount to ensure wound healing. To reduce manipulation, cotton gloves can be worn. For local wound treatment, hydrocolloid- or thermoplastic dressings can be applied. Medical treatment with amitriptyline, diazepam, chlorpromazine, clonazepam, carbamazepine and vitamin B supplements to reduce paraesthesia and self-manipulation have been described as well as transcutaneous electrical nerve stimulation to improve blood supply and wound healing. Furthermore, antibiotic treatment, as described in our case, should be performed to prevent superinfection. Concerning surgical interventions, free tissue transfers and local tissue flaps from non-affected areas for closure of small defects are being discussed. However, a tendency for recurrent ulcers can be seen.

Conclusion

In our case, the patient’s lack of disease comprehension and absence of disease improvement despite our multimodal interdisciplinary treatment with antibiotics, carbamazepine and sertraline warranted surgical intervention with wound debridement and simultaneous closure of the defect using an ALT flap.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Abbreviations

ALT, anterolateral thigh; TTS, trigeminal trophic syndrome.

References

