Management of Neurotrophic Keratitis by a Lower Eyelid Elevation with Scleral Graft, External Tarsorrhaphy and Multilayer Amniotic Membrane Transplant

Introduction

Neurotrophic keratitis is a degenerative disease characterised by damage to the corneal surface due to a sensory defect.

The sensitivity of the ocular surface depends on the nasociliary branch of the ophthalmic nerve, the first branch of the trigeminal nerve (cranial nerve V). These afferent fibres transmit sensory information from the upper eyelid, conjunctiva and cornea, playing a fundamental role in the detection of thermoalgesic, mechanical and chemical stimuli. On the other hand, the branches of the facial nerve (cranial nerve VII) are responsible for the motor innervation of the facial musculature, controlling blinking, as well as containing sympathetic and parasympathetic branches which control reflex lacrimal secretion through an integrated neuronal circuit between both systems.

Disruption of this sensory pathway and the neural feedback loops between the ocular surface and lacrimal glands can lead to corneal damage, either by defect, hypoesthesia as a cause of dry eye and neurotrophic keratitis, or by excess, hypersensitivity as a cause of neuropathic pain. [1]

Neuronal damage involves a deficit in the blink reflex, the tear secretion and the arrival of neurotrophic factors to the cornea, leading to epithelial damage that can manifest with different degrees of severity from superficial punctate keratitis to severe ulceration or perforation. Due to the lack of sensitivity, the predominant symptom is usually blurred vision and red eye. [1,2]

Corneal hypoesthesia can be caused by different diseases, with the most frequent being viral infections like herpes, extrinsic nerve compression (intracranial expansive lesions), and damage derived from neurosurgical interventions, chemical burns, trauma, ocular surgery, ischemia or metabolic alterations such as diabetes. [2,3]

The management of neurotrophic corneal ulcers is challenging, and the treatment must be tailored to the severity of the damage and the characteristics of the patient.

Initially, the treatment is based on lubrication and preservation of the corneal surface by means of artificial tears and moisturising ointments. Therapeutic contact lenses and eye drops with factors that promote epithelialization, such as autologous serum eye drops, plasma rich in growth factors (PRGF), eye drops with substance P, IGF-1, or Thymosinß-4) or collagenase inhibitors such as N-acetylcysteine, tetracyclines or medroxyprogesterone can also be used. Topical steroids may be useful in cases of inflammation but should be used with caution as they inhibit the stromal healing and may increase the risk of corneal fusion and perforation.

Surgical approach include different techniques such as upper eyelid ptosis induced by neuromodulation (injection of botulinum toxin into the elevator of the eyelid), tarsorrhaphy, autologous conjunctival flap, and amniotic membrane transplantation. In cases of severe corneal damage, keratoplasty techniques can also be considered. Corneal neurotization by the insertion of the contralateral supraorbital and supratrochlear nerves into the corneal limbus has shown promising results improving corneal sensitivity in some studies. [2, 3, 4, 5, 6]

Case presentation

A 46-year-old male with a history of stage IV nasopharyngeal carcinoma with facial and orbital invasion who was referred to the Ophthalmology department due to proptosis. Diagnostic imaging tests (Fig. 1) showed tumour extension to the right cavernous sinus, temporal fossa, orbital and petrous apex, surrounding the internal carotid artery.

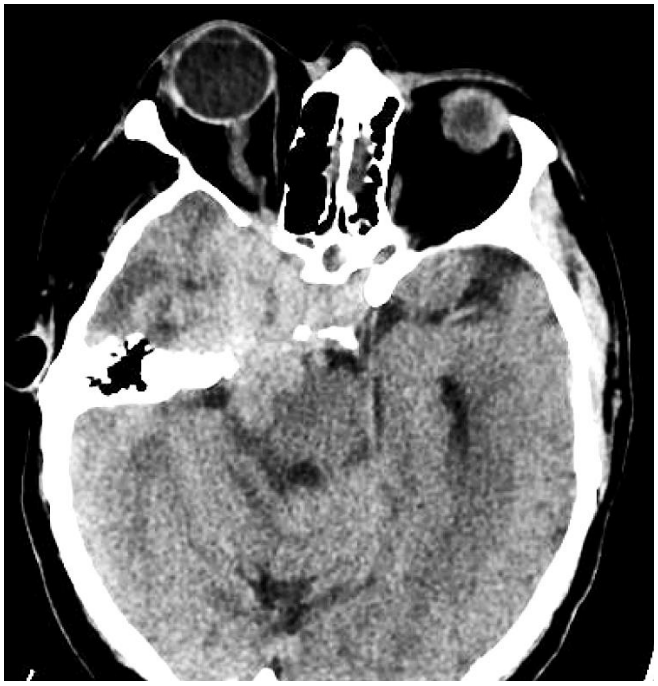
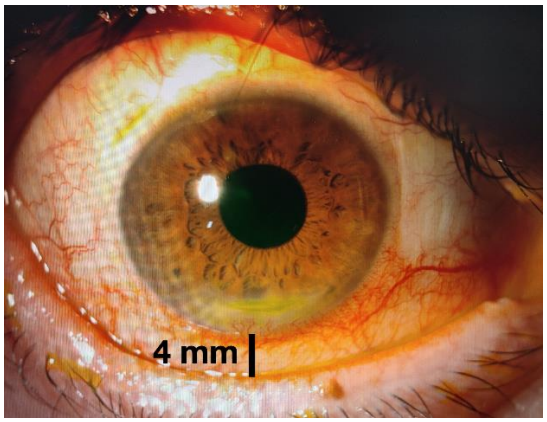


Figure 1: Cranial CT scan in axial view. The right ocular proptosis is evident, as well as the posterior tumour extension.

The tumour extension and the inflammation affected cranial nerves II, III, IV, V and VI, causing hypoesthesia in the territory of the first and second trigeminal branches (V1 and V2), right ocular proptosis with incomplete ptosis, non-reactive mydriasis, and hypotropic eye, with absence of extrinsic ocular motility (damage in cranial nerves III, IV and VI). The retinoscopy showed papillary pallor with intense excavation, findings that were corroborated by OCT images suggesting optic neuritis. The retina and macula were normal.

Surface examination showed obvious proptosis (Hertel: 24 mm), incomplete palpebral closure with negative Bell's phenomenon, 4 mm inferior scleral show (Fig. 2), and a 2 mm fluorescein-positive ulcer in inferior hemi-cornea with rolled edges and neovascularization towards the inferior epithelial defect (Fig. 3).



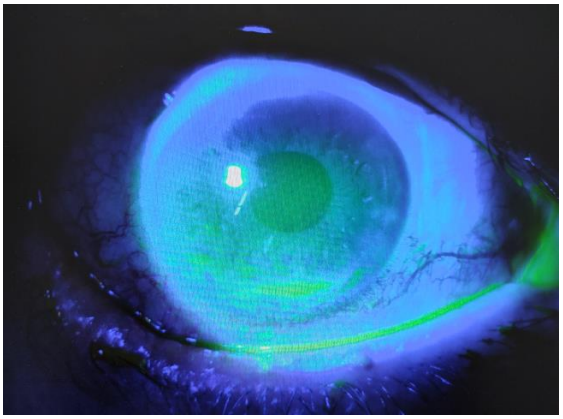


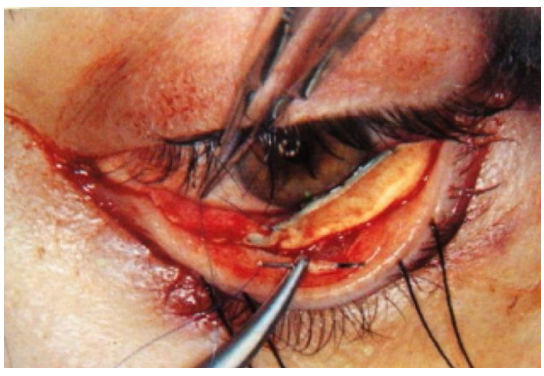
Figure 2: Slit-lamp view of the right ocular surface. Inferior scleral show of 4 mm. Figure 3: Fluorescein highlight in the inferior corneal epithelial defect.

During follow-up at the Cornea Unit, corneal scrapings were collected for smears and culture, and both were negative.

The patient was treated for months with different treatment regimens that included the use of topical antibiotics, oral tetracyclines, topical corticosteroids, medroxyprogesterone and PRGF eye drops, therapeutic contact lenses and even ptosis of the eyelid induced by infiltration of botulinum toxin in the elevator muscle of the upper eyelid.

The ulcer improved partially, but it recurred on several occasions, and for that reason, we decided to undergo surgical correction. It was performed by a lower tarsal elevation with scleral graft (Fig. 4), external tarsorrhaphy and corneal lining with bilayer amniotic membrane (Fig. 5).

The size of the scleral graft was 8 mm vertically (double of the scleral show) and 12 mm horizontally (2 mm greater than the white-to-white distance). The possibility of using the palate for the palpebral elevation was considered, but the scleral graft was chosen because it was considered to be more comfortable for the patient.



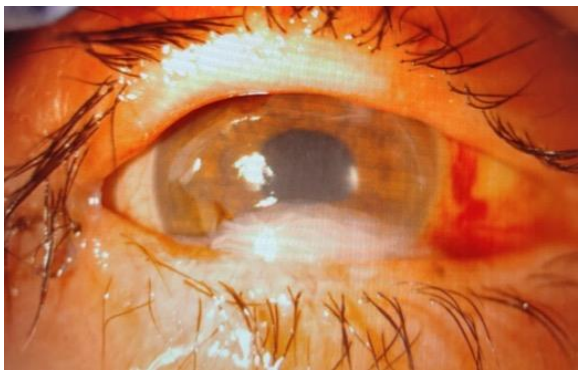
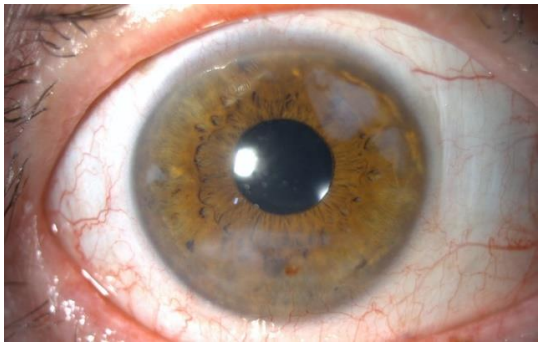
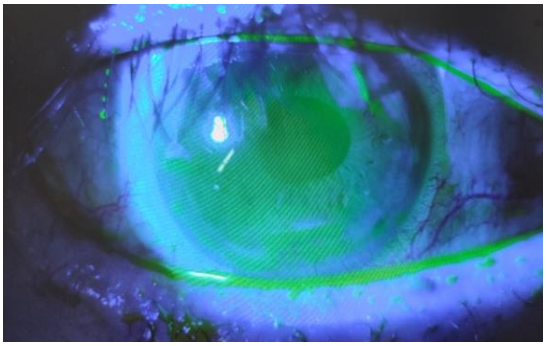
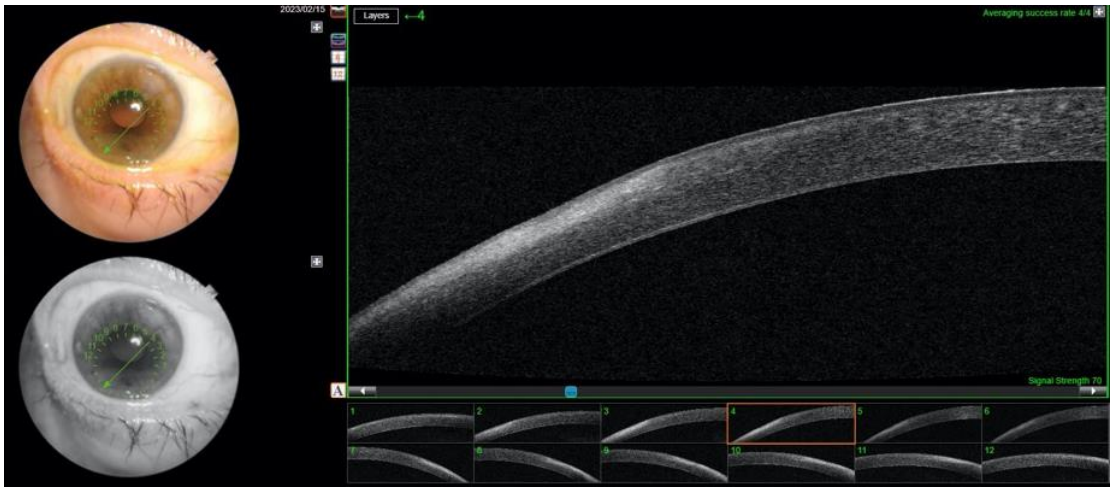


Figure 4: Scleral graft in the inferior tarsus of the right eye. Figure 5: Appearance after placement of multilayer amniotic membrane graft.

Currently, despite the patient continuing with proptosis, hypoesthesia and absence of extrinsic movement, he is subjectively better. He has no pain or discomfort, and the eye looks very healthy. The lower eyelid is well positioned, there is no scleral show, and the cornea is completely epithelialized with subepithelial fibrosis in the lower hemi-cornea (Fig 6, 7 and 8). The patient is only being treated with artificial tears.







State of the ocular surface 2 years after surgical treatment. Figure 6: Slit-lamp view of the ocular surface. Figure 7: Blue light view after fluorescein instillation. Figure 8: Anterior pole OCT image showing mild subepithelial fibrosis.

Discussion

Neurotrophic keratitis is a degenerative disease leading to the progressive deterioration of the corneal surface with a high risk of infection and perforation. The treatment should be based on the severity of the damage, with the main objective being to repair the epithelium and preventing potential complications.

There is currently no specific medical treatment for this pathology beyond hydration and products that promote the epithelialization. Most surgical approaches, with the exception of corneal neurotization, seek to preserve ocular integrity without improving corneal sensitivity, which is essentially the basis of the problem.

When assessing each case and considering the management of these patients, it is essential to carry out a good examination of all the structures involved in the integrity of the corneal surface; not only the cornea, but also including the eyelids and its statics and dynamics, the tear film and the conjunctiva.

In our case, the damage caused by corneal hypoesthesia was compounded by exposure due to proptosis and the absence of motility and no effective blinking due to concomitant paralysis of the III, IV, V and VI cranial nerves. This was the reason why we opted for a surgical correction that would guarantee ocular coverage instead of restoring corneal sensitivity.

Conclusion

The management of neurotrophic keratitis is a challenge for ophthalmologists. Current available medical and surgical treatments are primarily aimed at promoting healing and avoiding corneal perforation as the most serious potential complication. Fortunately, there is increasing research into therapies to restore corneal sensitivity and to treat the problem at its origin.

References

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