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Ophthalomology

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CASE REPORT: REFRACTORY NEUROTROPHIC KERATOCONJUNCTIVITIS AND ESODEVIATION SECONDARY TO MULTI-CRANIAL NERVE PALSIES FOLLOWING TRIGEMINAL SCHWANNOMA RESECTION

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ABSTRACT

Background: Neurotrophic keratopathy is a rare disorder caused by the loss of corneal sensation. It is characterized by persistent epithelial defects, corneal ulceration, and, ultimately, corneal perforation if not managed in a timely manner. The management includes aggressive lubrication, prophylactic topical antibiotics, therapeutic contact lenses, tarsorrhaphy, and amniotic membrane transplantation. Topical insulin therapy is one of the new therapeutic possibilities. **Case Presentation:** A 59-year-old female presented with severe vision loss following the surgical resection of a large left cerebellopontine (CP) angle trigeminal schwannoma. The patient had a history of obstructive hydrocephalus managed via a right ventriculoperitoneal (VP) shunt. Postoperatively, she developed concurrent left-sided cranial nerve V (trigeminal), VI (abducens), and VII (facial) nerve palsies. Ophthalmic examination revealed profound left lagophthalmos, severe esodeviation with abduction restriction, and Stage II/III Neurotrophic Keratoconjunctivitis characterized by a lusterless cornea and a central persistent epithelial defect (PED) complicated by filamentary keratitis. Despite systemic and neurological stability, her left eye demonstrated severe tear deficiency (Schirmer's test: 6 mm in 5 minutes) and immediate tear film breakup. Management required an aggressive, preservative-free multi-modal ocular surface regimen, topical immunomodulators, and a structured steroid-antibiotic taper. **Conclusions:** Concomitant paralytic lagophthalmos and esodeviation drastically accelerate the progression of neurotrophic corneal breakdown.[2] Preserving visual acuity requires rapid staging and immediate transition to intensive preservative-free therapies to prevent descemetocele formation and corneal melting.[3]

INTRODUCTION

The corneal epithelium relies heavily on sensory innervation from the ophthalmic division (V_1) of the trigeminal nerve. Trige-minal axons release essential neurotrophic factors including nerve growth factor (NGF), substance P, and calcitonin gene-related peptide (CGRP)—which maintain epithelial integrity, stimulate cellular proliferation, and drive wound healing.[1] Damage to these pathways leads to neurotrophic keratitis (NK), an insidious condition characterized by corneal anesthesia, epithelial breakdown, and poor healing responses.[4]

While trigeminal schwannomas represent less than <0.2% of all intracranial lesions, their location in the petroclival and cerebellopontine angle puts them adjacent to multiple skull-base structures. Surgical excision frequently risks damage to neighboring cranial nerves.[5]When a patient suffers a trineuropathy involving CN V (loss of sensation), CN VII (loss of orbicularis oculi motor function leading to lagophthalmos), and CN VI (loss of lateral rectus function leading to esodeviation), the cornea is deprived of chemical trophic support, physical mechanical wetting, and normal anatomical protection.[2,3] This report details the clinical challenges and structured management of this rare, triad-nerve ocular surface crisis.

Case Presentation

History and Examination

A 59-year-old female was referred for a comprehensive neuro-ophthalmic evaluation presenting with persistent irritation, severe foreign body sensation, neuropathic pain, and a progressive decline in vision in her left eye. Nine months prior, she had undergone subtotal surgical resection of a left CP angle trigeminal schwannoma at a tertiary neuro-institution. Her neurosurgical history was also notable for a right ventriculoperitoneal (VP) shunt placed to resolve obstructive hydrocephalus. While her systemic, cognitive, and lower-limb motor functions had recovered postoperatively, she was left with profound left-sided cranial nerve deficits.

Clinical and Ophthalmic Findings

On presentation, the patient's best-corrected visual acuity (BCVA) was 6/24 in the right eye (OD) and significantly compromised at 2/60 in the left eye (OS). Intraocular pressure (IOP) in the right eye was normal at 14 mmHg via non-contact tonometry; tonometry was deferred or limited in the left eye due to advanced corneal disease.

Extraocular Motility & Strabismus Examination

- **Right Eye:** Full, unrestricted ranges of motion in all cardinal directions.
- **Left Eye:** Demonstrated a prominent esodeviation in the primary gaze accompanied by severe abduction restriction, confirming a persistent, isolated postoperative left sixth (abducens) nerve palsy.

External & Anterior Segment Evaluation

- **Right Eye:** Mild Meibomian Gland Dysfunction (MGD) and early nuclear cataract formation. The cornea was clear with preserved sensation.
- **Left Eye: Lids/Adnexa:** Severe left facial palsy (CN VII denervation) resulting in complete lagophthalmos. The patient was completely unable to close the left eye voluntarily or reflexively, creating extensive exposure of the inferior and central globe.
    - **Conjunctiva:** Diffuse, deep ciliary and conjunctival congestion.
    - **Cornea:** The cornea was completely anesthetized to cotton-wisp testing, establishing complete loss of Trigeminal (V_1) sensory function. The surface was macroscopically lusterless and irregular. Slit-lamp biomicroscopy with fluorescein staining revealed a large, central persistent epithelial defect (PED) with rolled, thickened edges, characteristic of Stage II/III Neurotrophic Keratoconjunctivitis. Adherent mucinous filaments (filamentary keratitis) were present across the margins of the defect, anchored to degenerating epithelial cells.

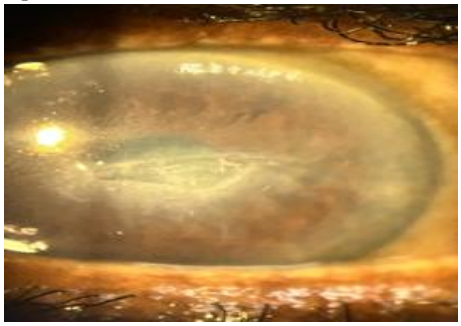


Figure 1: Anterior segment of left eye

Posterior Segment Examination

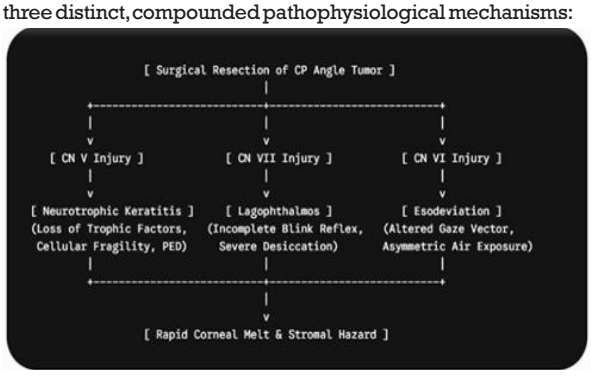
Fundoscopic evaluation of the right eye was normal. Examination of the left fundus yielded a highly hazy view due to the lusterless and irregular cornea; however, the optic disc and primary arcades were grossly seen and appeared stable, with no evidence of acute papilledema or optic atrophy.

COMPARATIVE OCULAR SURFACE METRICS

Metric	Right Eye (OD)	Left Eye (OS)
Schirmer's Test I (Tear Volume)	29 mm in 5 minutes (Normal)	6 mm in 5 minutes (Severe Aqueous Deficiency)
Tear Break-Up Time (TBUT) (Film Stability)	6 seconds (Marginal)	Immediate (Severe Evaporative Loss)
Corneal Sensation	Intact	Completely Absent
Eyelid Closure	Complete	Lagophthalmos

Pathophysiology of the Ocular Surface Crisis

The rapid degeneration of this patient's left cornea is driven by three distinct, compounded pathophysiological mechanisms:



1. **Loss of Sensory Neurotrophic Trophism (CN V):** Without neural-derived substance P and NGF, the corneal epithelial cells lose their hemidesmosomal anchoring complexes to Bowman's layer, leading to mechanical sloughing, impaired mitosis, and a persistent epithelial defect.[1]
2. **Paralytic Exposure (CN VII):** The loss of the blink reflex entirely halts the physical distribution of the tear film. Constant ambient air exposure evaporates what little tear volume is produced, turning a neurotrophic cornea into an arid, desiccated environment.[2]
3. **Static Esodeviation (CN VI):** Because the eye is structurally deviated inward due to lateral rectus paralysis, standard vertical eyelid movements fail to distribute moisture symmetrically across the exposed corneal curvature. This locks the central-lateral cornea into a vector of maximized environmental stress.

Management Protocol

To halt progression toward corneal perforation or melting, an intensive, multi-modal medical regimen was structured, focusing on eliminating preservatives, minimizing inflammation, and substituting the complex components of the tear film.[1,3]

Slit-Lamp Guided Therapeutics

- **Inflammation & Infection Control:** Lotepred T E/D (Loteprednol etabonate 0.5% + Tobramycin 0.3%) was initiated at 4 times daily for the left eye, scheduled for a strict weekly downward taper (3x daily, 2x daily, then 1x daily) under close monitoring. Loteprednol provides site-specific anti-inflammatory effects with a lower risk of elevating intraocular pressure, while tobramycin provides coverage against opportunistic bacterial pathogens.
- **Preservative-Free Aqueous Supplementation:** Lubrex DS E/D (Carboxymethyl Cellulose Sodium) was prescribed at a high frequency of 8 times daily to the left eye to constantly replenish missing tear volume.
- **Visco-adaptive Healing:** Soha Liquigel E/D (Sodium Hyaluronate) was added at 8 times daily to the left eye. The high viscoelastic property of sodium hyaluronate mimics natural mucins, remaining on the irregular neurotrophic surface to shield newly forming epithelial cells.
- **Nighttime Ointment Barrier:** Lacryl-PF E/O (Hydroxypropyl Methylcellulose ointment) was prescribed 4 times daily to the left eye. This creates a dense, hydrophobic barrier that prevents desiccation during sleep and periods of prolonged lagophthalmos.
- **Trophic and Lacrimal Restoration:** Cyclomune 0.05% E/D (Cyclosporine) was administered 3 times daily to the left eye to downregulate neurogenic surface inflammation, break the dry-eye metabolic cycle, and maximize whatever endogenous lacrimal gland output remained.

Procedural Actions

- **Discontinuation of Prior Toxins:** Prior therapy with Ocupol E/O was immediately stopped due to concern for cumulative medication-induced corneal epithelial toxicity, which frequently worsens a non-healing PED. Discontinuing toxic or preserved medications is essential to protect the compromised corneal stem cells.[3]
- **Lid Hygiene:** Strict instructions for gentle hot fomentation and lid hygiene were instituted to manage co-existing meibomian gland stagnation.

DISCUSSION & FUTURE INTERVENTIONS

Neurotrophic keratitis is traditionally classified into three clinical stages based on severity:[4]

- **Stage I:** Epithelial irregularity, superficial punctate keratitis, and stromal haze.[4]
- **Stage II:** Persistent epithelial defect (PED) with smooth, rolled edges, often involving minimal stromal loss.[4]
- **Stage III:** Corneal ulceration accompanied by stromal melting, descemetocele formation, or frank perforation.[4]

This patient transitions between Stage II and Stage III, demonstrating a long-standing, non-healing PED that has begun forming mucinous filaments due to localized cellular death and mucous stasis.[4] The treatment of neurotrophic keratitis combined with lagophthalmos requires a careful balance between protecting the eye and actively promoting healing.[2] Standard lubricants only address friction; they do not replace the cellular signaling pathways lost to neurotrophic damage.[1]

The patient and her caregivers were extensively counseled that current therapies are protective and stabilizing rather than immediately curative. If the central epithelial defect does not begin to close during the initial 1-week follow-up, the management plan will scale up to incorporate next-tier interventions:

1. **Recombinant Human Nerve Growth Factor (Cenegermin):** Topical application of cenegermin provides exogenous human NGF to directly restore the missing neural signaling pathways, triggering epithelial growth, differentiation, and supporting corneal reinnervation.[1,6]
2. **Amniotic Membrane Transplantation (AMT):** Placing an amniotic membrane over the defect provides a biologic scaffold rich in matrix proteins and fetal growth factors, which suppresses inflammation and promotes rapid re-epithelialization without risking further toxic exposure.[1,3]
3. **Autologous Serum Eye Drops (20% - 50%):** Formulated from the patient's own blood, these drops deliver crucial components missing from a neurotrophic eye, including epidermal growth factor (EGF), transforming growth factor-beta (TGF-beta), fibronectin, and vitamins. This formulation has proven highly effective in closure rates for neurotrophic defects combined with lagophthalmos.[2]
4. **Surgical Tarsorrhaphy:** If medical management fails, a temporary lateral or central tarsorrhaphy remains a vital option to physically narrow the palpebral fissure, reducing exposure and preserving the cornea until extraocular muscle or facial nerve function shows signs of recovery.[3]

Patient Consent

Informed consent was obtained from the patient for the publication of this case details and associated clinical findings.

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