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CASE REPORT

# Neurotrophic keratitis caused by trigeminal nerve palsy secondary to a cerebellopontine angle epidermoid cyst: A rare case presentation.

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**ABSTRACT…** Neurotrophic keratitis (NK) is a rare corneal disorder characterized by decreased corneal sensitivity and impaired corneal healing due to damage or dysfunction of the trigeminal nerve. We present a case of NK caused by trigeminal nerve palsy secondary to a cerebellopontine angle (CPA) epidermoid cyst. The patient exhibited corneal epithelial defects, persistent corneal ulceration, and decreased corneal sensation. Prompt diagnosis and management of the underlying brain lesion were crucial for successful treatment of the disease.

**Key words:** Corneal Sensation, Cerebellopontine Angle, Epidermoid Cyst, Neurotrophic Keratitis, Neurosurgery, Trigeminal Nerve Palsy.

## INTRODUCTION

Neurotrophic keratitis also known keratopathy is a rare optical disorder that causes degeneration of corneal epithelium as a result of impairment of trigeminal innervation. NK characterizes as impaired sensitivity and healing of cornea, stromal ulceration and perforation.[1,2] Trigeminal nerve is responsible for connecting cornea to brain and supplies the cornea with trophic factors that maintain ocular function and anatomical integrity. When corneal nerve is impaired, it leads to dysfunction of morphological and metabolic epithelium cells resulting in frequent defects. The trigeminal nerve can be impaired as result of congenital or iatrogenic disorders including herpes keratitis, corneal surgery, chemical injury, ablative procedures, surgery for jaw fractures and prolonged use of contact lenses. Some possible but rare causes of NK also include aneurysms, schwannoma and meningioma which compress the corneal nerve and reduce sensation. Diabetes, multiple sclerosis and leprosy are systemic diseases that can disrupt trigeminal nerve function.[2] In this case report, we presented a case of NK secondary to trigeminal nerve palsy caused by a CPA epidermoid cyst.

### Case Presentation

A 24-year-old female presented to our ophthalmology department with a history of painless blurring of vision in right eye associated with redness and watery discharge, progressive right-sided facial numbness, and difficulty chewing food for the last 3 months. Her medical history was unremarkable, with no prior ocular disease, trauma or surgery.

On examination, there was a noticeable atrophy of muscles on the right temporal region. A hemifacial sensory loss along with weakness of muscles of mastication were noted on the right side. Ophthalmic examination revealed a visual acuity of counting fingers in the right eye and 20/20 in the left eye. Slit-lamp examination of the right eye showed a corneal epithelial defect measuring 3x1.5 mm with surrounding infiltrates and conjunctival injection. In addition, corneal sensitivity was found to be significantly reduced in the right eye compared to the left eye. The anterior chamber and fundus examination were

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unremarkable. All of these unilateral findings suggested trigeminal nerve palsy of the right-side (Figure-1).

A Magnetic Resonance Imaging (MRI) of brain was ordered (Figure-2). A non-enhancing cystic signal intensity lesion was found at the right CPA represented by blue arrows in Figure A (axial T1-weighted), Figure B (axial T1-weighted contrast-enhanced) Figure C (coronal T1-weighted contrast-enhanced), Figure D (axial T2-weighted), Figure E (coronal T2-weighted), Figure F (axial FLAIR), Figure G (DWI), Figure H (ADC). This lesion showed diffusion restriction, extending into the ambient cistern causing mass effect over the brainstem, right cerebellar peduncle, right cerebellar hemisphere and fourth ventricle without any retrograde dilation of the ventricular chain.

Based upon these clinical and radiologic findings, a diagnosis of right-sided epidermoid cyst was made causing trigeminal nerve palsy on the right side presenting in the form of neurotrophic keratitis, facial numbness and weakness of muscles of mastication on the same side. The patient was prescribed lubricants, artificial tears and prophylactic antibiotics for her keratitis and was advised on the importance of eye protection and avoidance of trauma to the affected eye. This was followed by a total tarsorrhaphy of the affected eye. A neurosurgical consultation was obtained and the patient was referred to the neurosurgical department for surgical resection of the epidermoid cyst. A right retromastoid suboccipital craniotomy was done and the space-occupying lesion was excised.

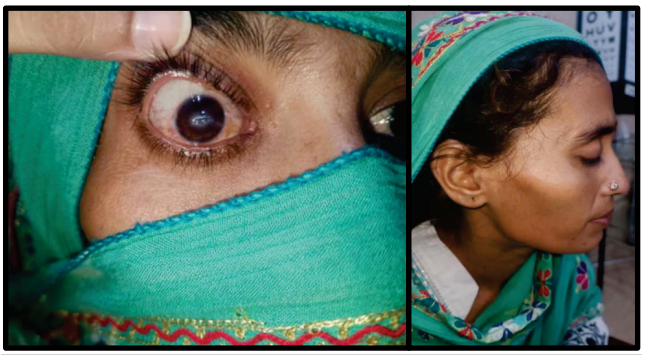

**Figure-1. Patient examination**

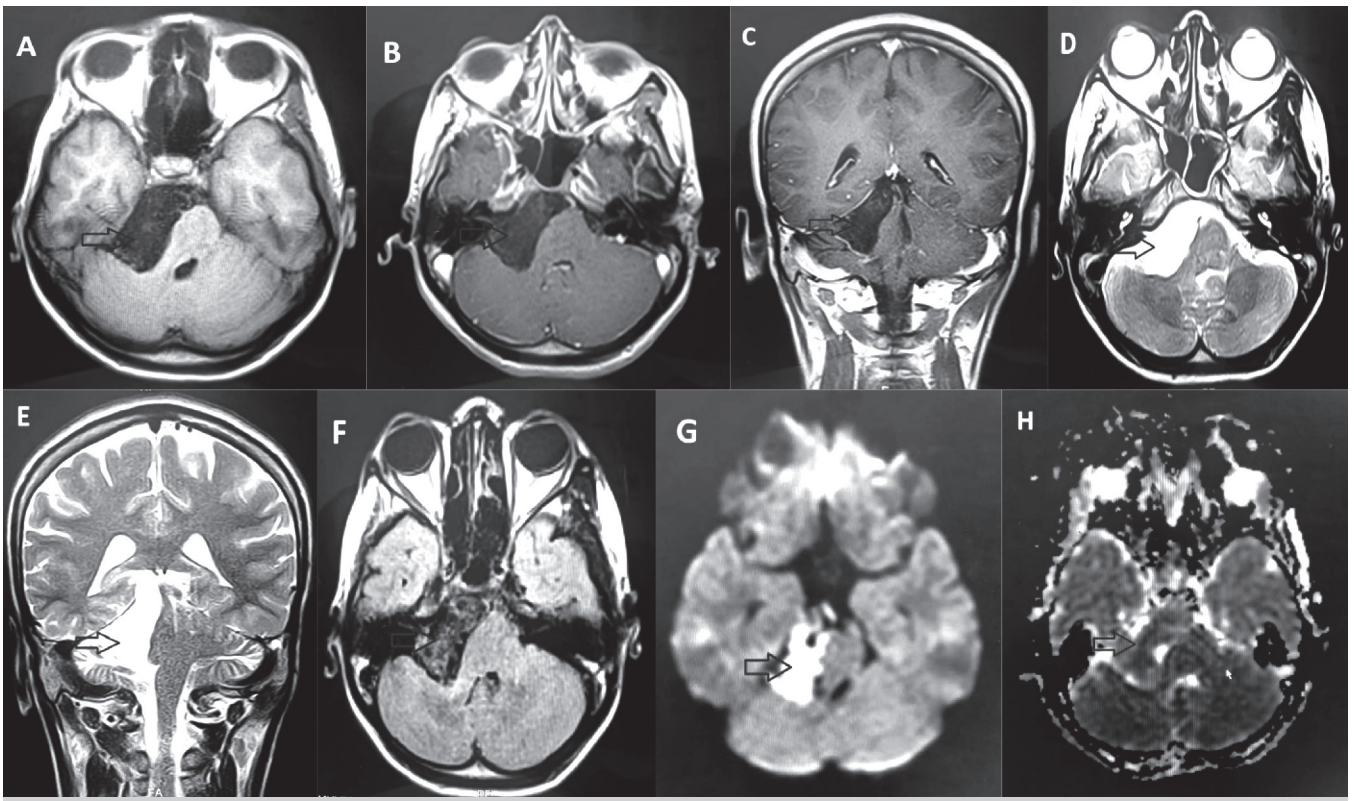


**Figure-2. MRI examination**

Histopathological assessment of the resected tissue confirmed the diagnosis of epidermoid cyst. The patient tolerated the procedure well and recovered without any complications or postoperative neurological deficits. Follow-up visits showed a resolution of trigeminal nerve palsy and a gradual improvement in corneal defect.

## DISCUSSION

Neurotrophic keratitis presents with optical dryness, impaired vision, photophobia, less blinking and difficulty in prolonged reading as a result of epithelial defects. Symptoms can worsen in the morning and aggravating factors such as travelling in an airplane, sitting in air-conditioned space, hot air from car heating and using blue light devices for long.[5] Prompt diagnosis, treatment based upon severity, and regular monitoring and follow-up speeds up healing and prevents disease progression. Affected cornea can be preserved by using artificial tears with 2-4 hours difference and an eye ointment before sleep.[2]

Epidermoid cysts are also known as primary cholesteatomas or pearly tumors.[3] They are the third most common tumors in the CPA region, representing 1% of all primary intracranial tumors.[3] They may also occur in the 4$^{th}$ ventricle and suprasellar region.[3] They usually surround important structures such as cranial nerves, brainstem, and vascular structures.[3] Despite being present congenitally, they grow slowly and then compress neurovascular tissue with only minimal clinical symptoms in the early or middle adulthood.[3,4] MRI is the preferred imaging modality to visualize the intracranial and extracranial course of the trigeminal nerve.[6] The gold standard treatment for CPA epidermoid cysts is retromastoid craniectomy, although subtotal removal is also justified if the tumor adheres densely to vital neurovascular structures.[4] Adjuvant radiotherapy may be applied only for recurrent epidermoid cyst or epidermoid cyst with malignant transformation.[4]

To our knowledge, no similar case presentation of CPA epidermoid cyst causing NK has been reported in the past. Therefore, this case report adds to the existing literature an important cause of NK that ophthalmologists should consider during clinical examination and workup.

## CONCLUSION

Neurotrophic keratitis resulting from trigeminal nerve palsy secondary to a CPA epidermoid cyst is a rare but potentially sight-threatening condition. Prompt diagnosis and management of the underlying lesion, along with appropriate ophthalmic care, are crucial for successful treatment and preservation of vision. Collaborative efforts between ophthalmologists and neurosurgeons are essential for optimal patient outcomes. In this case, surgical resection of the CPA epidermoid cyst resulted in the resolution of trigeminal nerve palsy and gradual improvement in corneal healing. Aggressive lubrication, prophylactic antibiotics, and topical autologous serum drops were employed to promote corneal healing and prevent complications. Regular monitoring of corneal sensation and follow-up visits ensured the evaluation of progress and early intervention if necessary.

### Informed Consent

Written informed consent was obtained from the patient for the publication of this case report and accompanying images. The patient understood the purpose and nature of the report and provided consent for the use of relevant clinical information and images while ensuring anonymity.

## CONFLICT OF INTEREST

The authors declare no conflict of interest.

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## AUTHORSHIP AND CONTRIBUTION DECLARATION

| No. | Author(s) Full Name | Contribution to the paper | Author(s) Signature |
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| 2 | Tahmina Sajjad | Presentation and writing. | |
| 3 | Shahroze Ahmed | Interpretation and design. | |