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Multi-Disciplinary Approach in a Case of Neurotrophic Keratitis with Vestibular Schwannoma and Seventh Cranial Nerve Palsy

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Abstract
Multidisciplinary approach is essential in some cases of keratitis as here we required multidisciplinary approach for a 54 yrs female reported to OPD with diminished vision, lagophthalmos & neurotrophic keratitis of Right eye along with other features of seventh cranial nerve palsy. On detailed clinical & radiological investigation patient was provisionally dignosed having vestibular schwannoma. Patient had undergone right retromastoid-suboccipital craniotomy and subtotal excision of the mass confirmed histopathologically to be vestibular schwannoma. Lateral tarsorrhaphy was done as well as medical treatment was also given to protect vision. 6 week follow up revealed neurotrophic keratitis healed, residual corneal opacity remained. Vision improved to 6/36. Exposure keratitis as well as neurotrophic keratitis can also occur due to various intracranial causes, we need to find & treat the cause, multidisciplinary approach can save life as well as improve quality of life.
Keywords: *vestibular schwanoma, seventh nerve palsy, neurotrophic keratitis.*

Introduction

Vestibular schwannoma is a benign slow growing tumor due to overproduction of Schwann cells arising from the eighth cranial nerve and due to inactivation of the NF2 gene and its product merlin[1]. Early detection of the tumor is difficult as it remains asymptomatic at early stages of disease. Incidence is 1 out of every 100,000 individual per year.[1] The tumor consists of approximately 8% of all tumors inside the skull. Mostly unilateral cases are known to occur sporadically[2]. Vestibular schwannoma can affect eyes in various forms as depicted by Rogers NK & Brand CS in a retrospective study of 138 patients, 61 patients (44%) required lid surgery, 18 (13%) developed minor superficial exposure keratopathy, 13 (9%) developed corneal opacification or clouding, two had recurrent infective abscesses and four developed optic atrophy. Corneal complications mostly arising out of preoperative fifth nerve involvement[3].

Presentation of the case proper

- A 54 yrs female reported to OPD with chief complaint of Dimness of vision and inability to close her right eye for past 2 years.
- Chronic Foreign body sensation, excessive watering, photophobia in right eye for past 2 years.

On detailed history taking it revealed that patient also experienced Right sided hearing loss for 3yrs, tinnius, vertigo for same duration along with episodic Vomiting & headache for 2.5 yrs. For last 6 months patient is experiencing gait unsteadiness, headache and drooling of saliva from right side of mouth.

Her past history remained not significant without any history of trauma or recent infection. Ocular examination revealed

	RIGHT EYE	**LEFT EYE**
BCVA	HM + , PR ACCURATE	6/36
COLOUR VISION	Patient can read all plates of Ishihara chart	Patient can read all plates of Ishihara chart
BROW AND FOREHEAD	**REDUCED WRINKLING OF FOREHEAD**	NORMAL
CORNEAL SENSATION	REDUCED IN ALL FOUR QUARDRANTS	NORMAL
LIDS AND ADNEXA BELLS' PHENOMENON	INABILITY TO CLOSE UPPER EYE LID. PRESENT	NORMAL PRESENT
GOLDMAN APPLANATION TONOMETRY	14 mm of Hg (11:25 A.M)	14 mm of Hg (11;25 A.M)
GONIOSCOPY	Details of structures not visible due to corneal opacities.	
DIRECT OPHTHALMOSCOPY	Fundus details not visible	Disc- Normal Vessels-Normal Macula- Healthy
DEGREE OF LAGOPTHALMOS	**6mm of lagopthalmos on gentle closure of lids and 5mm lagopthalmos on forceful closure**	No lagopthalmos
TEAR BREAK UP TIME	6 sec	10 sec
OCULAR MOTILITY AND STRENTH OF ORBICULARIS OCULI MUSCLE	**Ocular motility normal but strength of orbicularis oculi muscle decreased.**	Normal
BLINK RATE	INFREQUENT, INCOMPLETE.	NORMAL
SCHIRMER TEST	24mm	17mm

Fig. 1-patient at primary gaze & having lagophthalmos in right eye

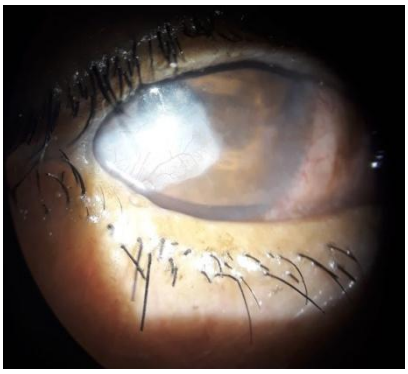

Fig: 2- slit lamp examination of trophic ulcer, a 4 X 3mm epithelial defect

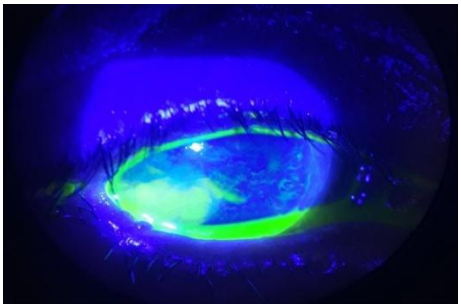

Fig 3- trophic corneal ulcer under fluorescent staining.

Facial examination

Fifth Cranial Nerve: Temporalis and masseter weakness in right sensory system. Decreased sensation of V1, V2, V3. Right sided corneal reflex absent.

Seventh cranial nerve: Minimal deviation of angle of mouth to the left, reduced wrinkling of forehead in the right side.

Eight cranial nerve: Rinnes AC>BC and weber lateralized to the left.

Other Cranial Nerves: Normal.

Motor system: Tone –normal, Power 5/5, DTRs 2+.

Cerebellar signs: Tandem walking impaired, right side positive.

Routine blood examination: normal

Radiological examination:

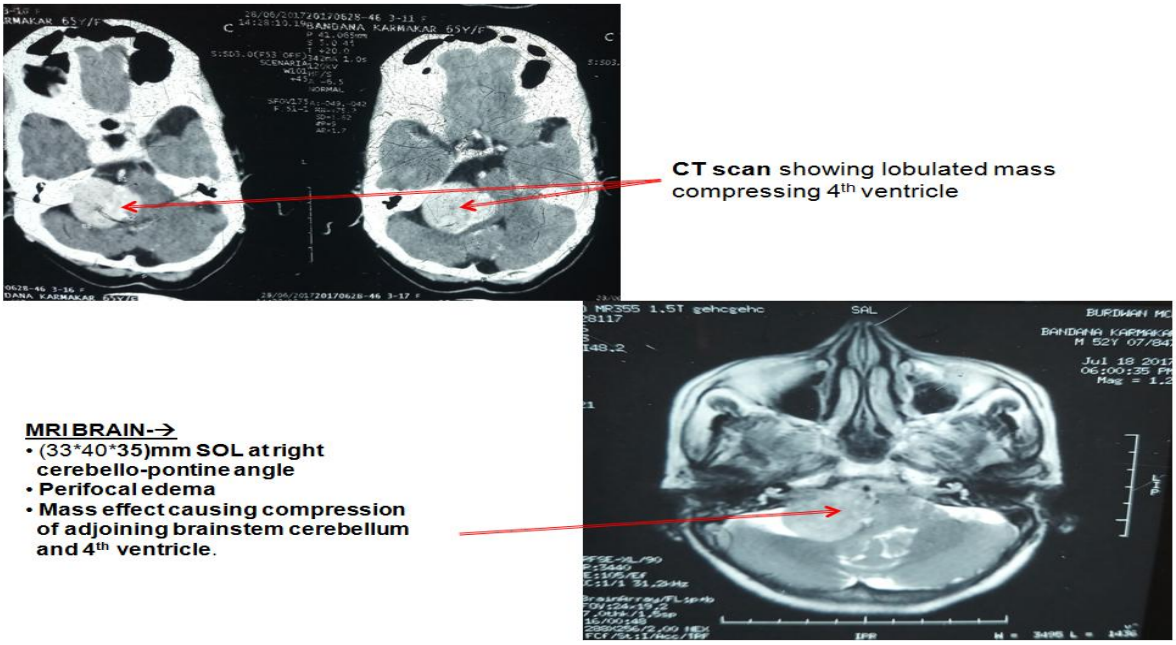

Fig. 4-CT scan & MRI brain

CT scan shows typical ice cream cone pattern lobulated mass in cerebello-pontine angle compressing 4th ventricle which also corroborative with MRI brain findings.

After consultation with neurosurgeons & ENT specialists diagnosis of vestibular schwannoma with VIIth nerve palsy was made. Patient had undergone right suboccipital retrosigmoid approach craniotomy and subtotal excision of the mass. Histopathology revealed vestibular schwannoma.

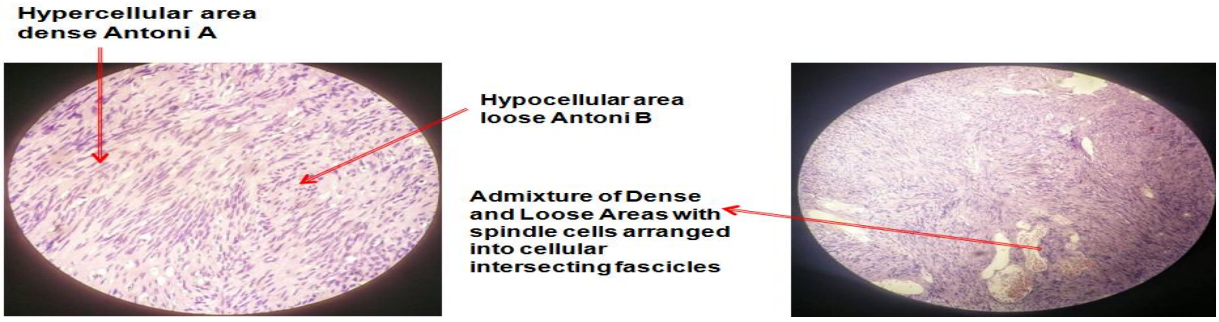

Fig 5- HPE slide of vestibular schwannoma

Lateral tarssoraphy of the right side was done to protect the cornea & hence restore vision.

Fig.-6- lateral tarsorraphy of right side

Follow up: 6 week follow up revealed exposure keratopathy healed, residual corneal opacity remained. Vision improved to 6/36

Pure tone Audiometry

Right sided profound perceptive hearing loss. (Inner ear hearing loss).

Left normal hearing.

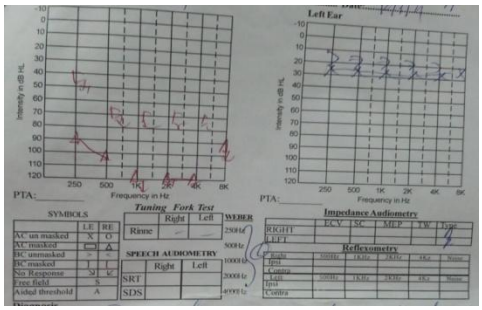

Fig 7- Right side profound sensorineural deafness, left side normal hearing.(Pure Tone audiometry)

Post operative MRI Brain: Showed a thin rim of residual tumor at the cerebello-pontine angle and within Internal Auditory Meatus.

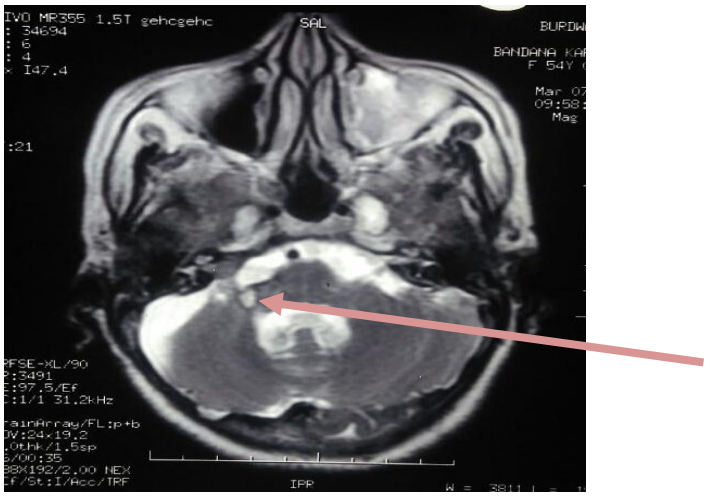

Fig. 8- thin rim of residual tissue at cerebello-pontine angle & within internal auditory meatus

Discussion

The patient had a large vestibular schwannoma (size being 33x40x35 mm as in MRI) according to vestibular schwannoma classification[4]. A study by Huang X, XU J, XU M, et al showed that large vestibular schwannoma can have diverse manifestations in the form of hearing loss (85.8%), facial paresthesia (48.9%), instability of gait (44.6%), tinnitus (40.1%), deafness (26.3%) and facial paralysis (21.1%), absent corneal reflex was the most common sign, observed in 15.2% of the 1,009 patients[5]. As this patient had deafness alongwith facial nerve palsy leading to lagophthalmos & also trigeminal nerve compression. Trigeminal nerve involvement led to absent corneal reflex & mastication problem of the patient. As a result patient also developed neurotrophic keratitis leading to further reduction of visual acuity. As stated in a study by Lambiese et al that trigeminal nerve damage induces long-term corneal functional and morphological changes of all layers of the cornea and that neurotrophic keratitis never completely resolves[6]. Though the patient had subtotal excision of the tumour via retrosigmoid suboccipital approach & it is reported to be one of the best approach to preserve facial nerve function & auditory function[7] but the severe sensorineural deafness as well as ocular symptoms persisted post operatively. Persistence of ocular symptoms corroborating with the findings of Lambiese et al. Finally we performed permanent lateral tarsorraphy to protect cornea & preservative free tear substitutes were added. Cornea healed with improved visual acuity to 6/36 as seen in subsequent follow up visits. As experienced by Kasparova EA et al in their study of 12 patients (13 eyes) with intracranial lesions complicated by neurotrophic keratitis and lagophthalmos receiving permanent partial tarsorrhaphy as a part of combined surgery that included simultaneous keratoplasty, autoconjunctivoplasty, also proposed to be an effective treatment that can help preserve visual function in such eyes[8].

Conclusion

Exposure keratitis as well as neurotrophic keratitis can also occur due to various intracranial causes, we need to find & treat the cause, multidisciplinary approach can save life as well as improve quality of life.

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