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Case Report

# A Rare Case of Hearing Impairment due to Cerebello–Pontine Angle Lesion: Trigeminal Schwannoma

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Schwannoma of the trigeminal nerve is a rare condition. Even rarer is hearing loss occurring as a result of this lesion. The aim of this study is to highlight this rare cause of hearing impairment. Here we report the clinical features and findings of the imaging and audiological investigations of a case of trigeminal schwannoma diagnosed at our institution. Our patient presented with headache, giddiness, tinnitus, left-sided facial weakness, left-sided hearing loss, right-sided hemiplegia, and unintelligible speech. Radiological studies revealed a large well-defined mass lesion in the left cerebellopontine angle with a significant mass effect on posterior fossa structures, suggestive of trigeminal nerve tumor. Audio-vestibular assessment was done with pure tone audiometry, impedance audiometry, otoacoustic emission, brainstem-evoked response audiometry, and electronystagmography, which pointed toward a retrocochlear pathology for hearing loss and imbalance.

**KEYWORDS:** Cerebellopontine angle tumor, vestibulocochlear nerve bundle, trigeminal schwannoma, electronystagmography

## INTRODUCTION

Schwannomas, which usually exhibit benign behavior, are tumors arising from Schwann cells in the axon myelin sheaths [1]. Schwannoma of the trigeminal nerve comprises only 0.2% to 0.4% of all intracranial tumors and primarily arises in the Gasserian ganglion [2]. They are relatively rare and less common than vestibular schwannoma [3]. They account for 0.07%–0.36% of all intracranial tumors and 0.8%–8% of intracranial schwannomas [4, 5]. It is a rare tumor in the cerebellopontine angle (CPA). Those located in the posterior fossa account for 20% of all cases. It is a rare cause of hearing impairment. The pathophysiology of trigeminal schwannoma is less known, and a schwannous proliferation can appear in CPA, the cavernous sinus, infratemporal fossa, or in one of the three branches of the trigeminal nerve beyond the Gasserian ganglion [6]. Owing to the rarity of its occurrence, most audiologists are unaware of this as a cause for hearing impairment; hence, it is important to create awareness among them (Audiologist) to look for the not-so-obvious causes of hearing impairment.

## CASE PRESENTATION

A 37-year-old male with no significant past medical history presented to the otorhinolaryngology department with symptoms of headache, giddiness, tinnitus, left-sided facial weakness, and hearing loss in the left ear since the last 6 months. He initially had a constant sense of imbalance for 3 months, after which he developed weakness of right upper and lower limbs, followed by unintelligible speech for 1 month before presentation. After informed consent was obtained from the patient, a thorough clinical examination was performed, which revealed right-sided hemiplegia with left-sided facial palsy. The examination of other cranial nerves revealed trigeminal nerve palsy with an absent corneal reflex and decreased sensation on the left side of the face. His otoscopic examination showed a normal tympanic membrane in both ears. Tuning fork tests showed much reduced absolute bone conduction in the left ear. Cerebellar signs were positive.

Computed tomography (CT, 64-slice, Siemens, Germany) of the brain showed a large well-defined lesion in left CPA that enhanced with intravenous contrast. Magnetic resonance imaging (MRI, 1.5 Tesla, Siemens, Germany) of the brain showed a large well-defined extra-axial mass lesion in the left CPA and parasellar region with heterogeneous enhancement and a significant mass effect on posterior fossa structures, suggestive of a trigeminal nerve tumor (Figure 1).

The patient underwent a complete audiological work-up comprising pure tone audiometry, speech audiometry, immittance audiometry, otoacoustic emission (OAE), brainstem-evoked response audiometry (BERA), and electronystagmography (ENG). The pure tone audiometry (Arphi 500, India) showed normal hearing in the right ear and profound hearing loss in the left ear (Figure 2).

This study was presented at the 44th Indian Speech and Hearing Association Conference held at Hyderabad, India, 2012.

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Speech audiometry was done with the use of spondee words in the right ear with normal scores (100%). In the left ear, scores were obtained using a general conversational speech with very poor scores (20%). Immittance audiometry (GSI 33, Version II, USA) showed a normal tympanogram in both ears, with stapedial reflexes absent in the left ear and present in the right ear.

Otoacoustic emission (Neuro Audio, Version 2010, Russia) was absent in the left ear with normal results in the right ear. BERA (Intelligent Hearing System Smart EP, Version 3140, USA) showed no peaks in the left ear at 90 dBnHL (Figure 3), which correlates with the findings of profound hearing loss in the pure tone audiometry. This is indicative of either a cochlear or retrocochlear pathology. BERA of the right ear showed I, III, and V peaks at normal absolute latencies with normal inter-peak latencies at 11.1/S and 91.1/S repetition rates.

In ENG (Micromedical Technologies, USA), the saccadic, pursuit, and optokinetic tests (Figure 4, 5, 6) pointed toward a central vestibular pathology with bithermal caloric tests reinforcing the same inference. Cervical vestibular-evoked myogenic potential could not be conducted on this patient because his muscle tone could not be sustained as a result of a cerebrovascular accident. All the above tests pointed toward a retrocochlear pathology in the left ear causing both hearing and balance problems.

## DISCUSSION

Schwannomas are more common in the fourth and fifth decades, though they may occur at any age [6]. Schwannomas are slow-growing benign tumors arising from Schwann cells in the axon myelin sheath. A trigeminal schwannomas is an extremely rare entity [6], which usually presents with pain or sensory loss in the face as initial symptoms [7]. As the tumor grows, it extends along the course of the trigeminal nerve. The Trigeminal nerve is found superior to the CPA, therefore, a tumor originating from this nerve may present like any other CPA lesion.

CPA is a triangular space located posterior to the pyramid, inferior to the tentorium, lateral to the pons, and ventral to the cerebellum [8] and contains the trigeminal, abducent, facial, and vestibulocochlear nerves and superior cerebellar and anterior inferior cerebellar arteries. The most common CPA tumor is vestibular schwannoma, and it accounts for 80%–94% of them, followed by meningiomas (3%–10%) and epidermoids (2%–4%). Much rarer primary tumors are schwannomas of the trigeminal nerve and facial nerve [9]. CPA lesions usually present with headache, hearing loss, dizziness, tinnitus, and other symptoms of space-occupying lesions in the cranium [10].

In our patient, the presentation of the tumor was at a very advanced stage, at which point, all surrounding nerves were involved, including the surrounding brain tissue, resulting in contralateral hemiplegia and unintelligible speech. In the presence of vestibular symptoms, one must consider the possibility of an irritative or a compressive mechanism on the vestibular nerve in its trajectory in the posterior fossa between the internal auditory meatus and the brainstem. This possibility is frequently neglected in favor of a peripheral origin at the level of the labyrinth or a central origin at the level of the brainstem. From the clinical point of view, one must suspect CPA lesions when vertiginous syndrome is accompanied by signs involving other cranial nerves.

An audiological test battery pointed toward a retrocochlear pathology. Pure tone audiometry revealed unilateral profound hearing loss

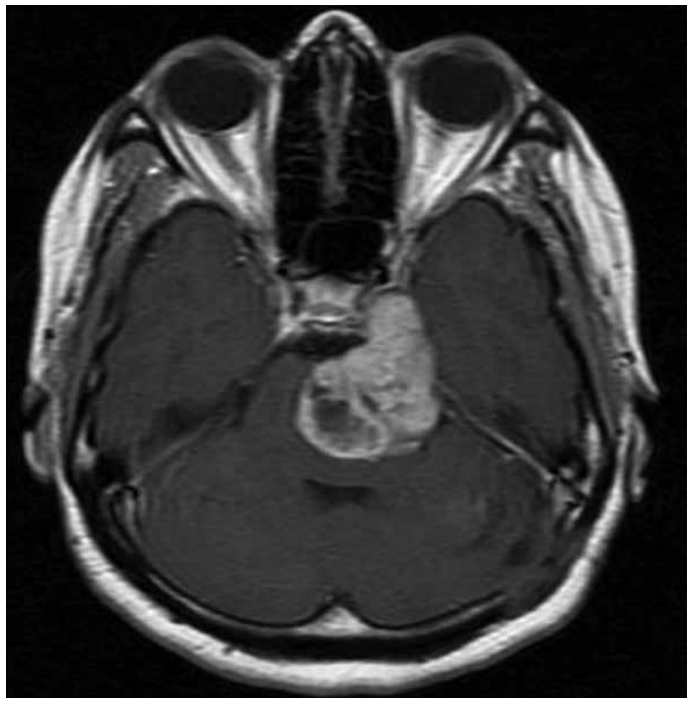

**Figure 1.** T1 with a contrast MRI image of the mass showing a large well-defined extra-axial mass lesion in the left cerebellopontine angle and parasellar region with heterogeneous enhancement and with a significant mass effect on posterior fossa structures

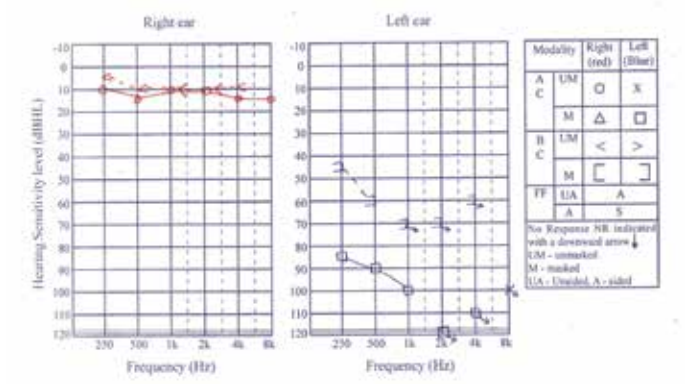


**Figure 2.** Pure tone audiometry showing profound hearing loss in the left ear and normal hearing thresholds in the right ear

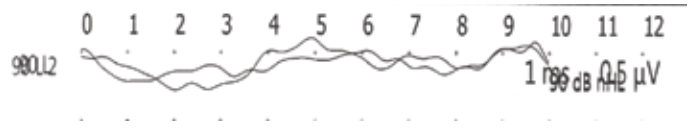


**Figure 3.** Absent BERA waveform in the left ear

with poor speech audiometric scores. Immittance audiometry showed a normal tympanogram with the absence of acoustic reflexes in the left ear. OAE and BERA were found to be absent in the left ear. ENG pointed toward a central vestibular lesion as the cause for imbalance.

After obtaining informed consent from the patient, surgical excision of the schwannoma by the retrosigmoid approach was performed. The diagnosis was confirmed by histopathological exam-

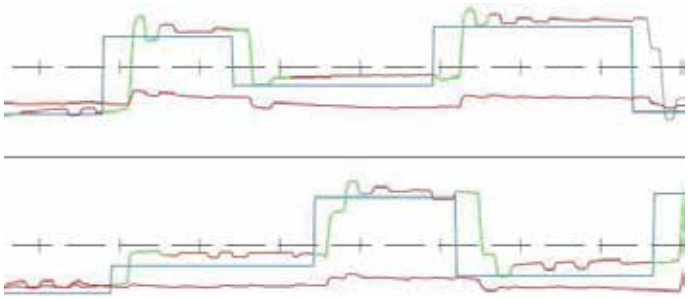

**Figure 4.** Abnormal saccadic test

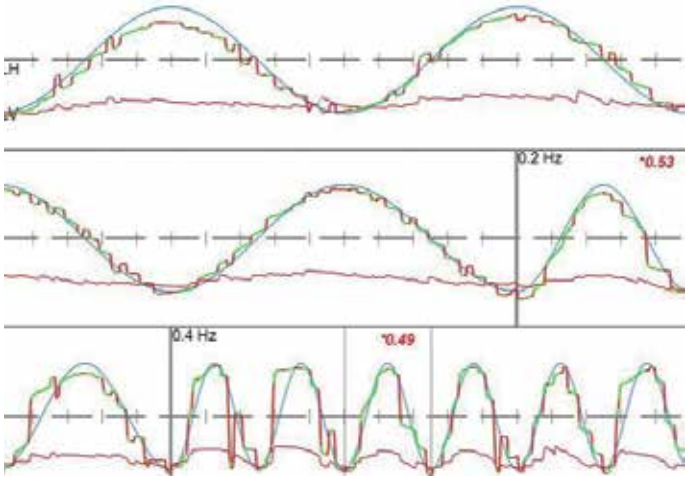


**Figure 5.** Abnormal pursuit test

**Figure 6.** Abnormal optokinetic test

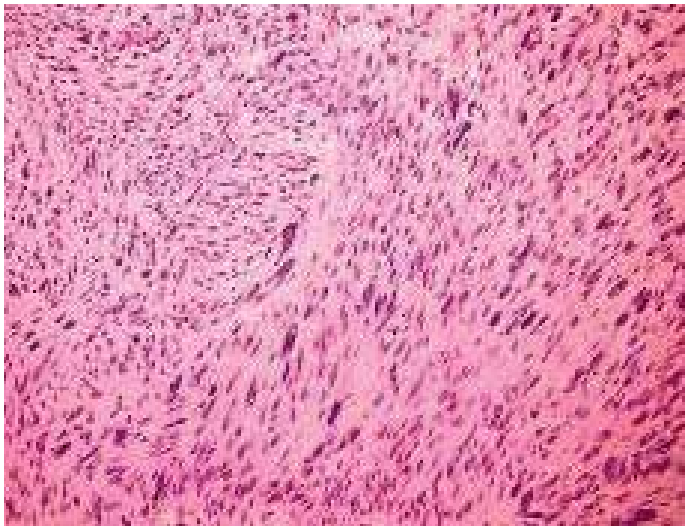

**Figure 7.** Microscopic examination of the excised specimen (using eosin–hematoxylin) showing Antoni A and Antoni B cell clusters

ination of the excised specimen (Figure 7). Following the surgery, the patient underwent speech therapy and physiotherapy for residual neurological deficits. No major complications were noted following surgery.

## CONCLUSION

Vestibular schwannomas are by far the most common cerebellopontine neoplasms and present as a combined intracanalicular/CPA lesion. Large vestibular schwannomas are likely to cause trigeminal nerve symptoms. The internal auditory meatus is rarely enlarged by trigeminal schwannomas, which distinguishes these tumors from vestibular schwannomas on imaging. Hearing impairment in trigeminal schwannoma is caused as a result of compression of the tumor on the vestibulocochlear nerve. The audiological profiling of these patients show features pertaining to those of any CPA mass. Because trigeminal schwannomas are usually picked up at later stages, the profiling would consist of severe to profound hearing loss with corresponding features in OAE and BERA. The aim of this study is to bring to the notice of audiologists that there are rarer causes in CPA other than vestibular schwannoma in patients presenting with hearing impairment with or without vestibular symptoms.

**Informed Consent:** Written informed consent was obtained from the patient who participated in this study.

**Peer-review:** Externally peer-reviewed.

**Author Contributions:** Concept - P.G.; Design - P.G., P.P.K., P.S.; Supervision - P.P.K.; Materials - P.G., P.S.; Data Collection and/or Processing - P.G., P.P.K., P.S.; Analysis and/or Interpretation - P.G., P.S., P.P.K.; Literature Review - P.G., P.S.; Writing - P.G., P.S.; Critical Review - P.P.K.

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