An Unusual Cause of Unilateral Profound Hearing Loss and Disabling Tinnitus - A Case Report

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ABSTRACT

Introduction: The aneurysm or loops of the anterior inferior cerebellar artery (AICA) in the internal auditory canal are exceeding rare and its clinical manifestations vary. The vascular loop of the AICA in the internal auditory canal may result in compression of the eighth or vestibulocochlear nerve. Microvascular compression of the eighth cranial nerve often causes tinnitus, hearing loss and vertigo. Several works of literature are reporting the microvascular compression of the vestibulocochlear nerve at the cerebellopontine angle (CPA) and not intrameatal area.

Aim: To report an unusual cause of unilateral profound hearing loss and disabling tinnitus.

Case Report: A 42-year-old man was presenting with hearing loss and tinnitus in the left ear for 2 months. MRI confirmed the intrameatal compression of the vestibulocochlear nerve by the vascular loop of the AICA.

Discussion: The neurologic examinations of the patient were within normal limits. The caloric test was normal on both sides. Auditory brainstem response (ABR) showed latency of wave I and III brainstem potentials. MRI showed the vascular loop of the AICA compressing the vestibulocochlear nerve.

Conclusion: Vascular loop by AICA in the internal auditory canal can compress the cochlear nerve which results in cochlear symptoms like tinnitus and sensorineural hearing loss. This vascular malformation can be diagnosed with help of clinical symptoms and magnetic resonance imaging (MRI).

Key Words: Vascular loop, Anterior inferior cerebellar artery, Internal auditory canal, Tinnitus, Cerebellopontine angle, MRI

INTRODUCTION

Hearing loss and tinnitus are very distressing symptoms that affect many people in the world.¹ Hearing loss and tinnitus can affect people of any age group. Unilateral sensorineural hearing loss (SNHL) or tinnitus often gives temptations for clinicians to rule out any intracranial lesions. The unilateral SNHL may be acute and/or progressive and ranges from mild to profound. The majority of the cases with unilateral SNHL have unknown aetiology.² The known cause of the unilateral SNHL are tumours at the cerebellopontine angle, demyelination, stroke and autoimmune disease.¹³ The clinical manifestations of unilateral hearing loss which evolves gradually, tinnitus and unsteadiness make suspicion of the acoustic neuroma or cerebellopontine angle tumour. The cerebellopontine angle (CPA) tumour is usually diagnosed by magnetic resonance imaging (MRI) with gadolinium contrast. However, the vascular loop of an anterior inferior cerebellar artery (AICA) compressing the cochlear nerve with symptoms of unilateral hearing loss and tinnitus may misdiagnose or delay the diagnosis and treatment because of its rarity and ignorance of this clinical entity to the clinicians. The compression of cranial nerves by the vascular loop is proposed as an etiological factor for several clinical manifestations. The details of the internal auditory canal other than the seventh and eighth cranial nerves namely the vascular loop of AICA can be easily identified by MRI. The clinical importance of this vascular loop is still controversial. Auditory symptoms like deafness and tinnitus are two important clinical manifestations that may be associated with the rarely seen vascular loop of AICA in the internal auditory canal. Here we are presenting a case of the vascular loop of AICA in the internal auditory canal leading to unilateral profound hearing loss and disabling tinnitus.
CASE REPORT

A 42-year-old man attended the outpatient department of Otorhinolaryngology with complaints of hearing impairment and tinnitus in the left ear for two months. He had taken medications from local doctors but showed no improvement. The tinnitus was non-pulsatile and persistent. He had no history of vertigo or unsteadiness and aural fullness in the ear. Otoscopic examinations showed normal external auditory canal and tympanic membrane on both sides. Pure tone audiometry showed left side profound hearing loss. A balanced examination of the patient did not reveal any imbalance tendencies to the right or left side. As the patient had unilateral hearing loss and tinnitus, so MRI was advised to rule out CPA lesions. MRI showed no abnormality in the brain but revealed a left intrameatal AICA loop (Fig. 1) as a possible aetiology for these disabling cochlear symptoms. The vascular loop of AICA in the internal auditory canal compressing the vestibulocochlear nerve. His neurologic examinations were within normal limits. The caloric test was normal on both sides. Auditory brainstem response (ABR) showed latency of wave I and III brainstem potentials. After confirmation of the diagnosis of the vascular loop of AICA in the internal auditory canal, the patient was referred to the neurosurgical department for further treatment where the patient underwent left retro-sigmoid craniotomy and microsurgical exploration of the vestibulocochlear nerve after drilling the posterior wall of the internal auditory canal. The neurovascular decompression was performed at the internal auditory canal. The disabling tinnitus was relieved after surgery. There was no change in hearing improvement after surgery.

DISCUSSION

The concept of vascular compression for explaining the hemifacial spasm and trigeminal neuralgia are established in clinical practice. The neurovascular compression of the seventh and fifth cranial nerves is the aetiology for hemifacial spasm and trigeminal neuralgia respectively. However the relationship of neurovascular compression with otologic manifestations such as hearing loss, tinnitus and dizziness are not yet clear. In daily life, otologic symptoms are commonly presented by the patients, so the rare etiological factor as vascular compression by AICA may be missed or result in delayed diagnosis. Although the vascular loop of AICA can result in compression of the eighth cranial nerve at the CPA, the neurovascular compression in the internal auditory canal has been reported in only two cases. The vascular cross compression of the vestibulocochlear nerve by the AICA results in several audiovestibular symptoms such as recurrent vertigo and tinnitus.

The AICA originate from the basilar artery. In 52% of cases, AICA arises from the lower third of the basilar artery whereas from the middle third in 46% and 2% in the upper third of the basilar artery. There are four types of anatomical arrangement of the main trunk of AICA. In the first type, AICA runs to the CPA ventrally to the abducent in approximately 79% cases, dorsally in approximately 16% cases and via the duplicated abducent nerve in approximately 5% of the specimens. In the second type, AICA runs between the pons and medulla to CPA in approximately 14% of cases. In the third type, the combination of the first and second types are seen(26%). In the fourth type (6%), there is a large anastomosis between AICA and the posterior inferior cerebellar artery(PICA). In this fourth type, AICA loops are seen at the exit point and entry zones of the facial nerve and vestibulocochlear nerves near to the brainstem. In our case, the type II arrangement of AICA was found. The pulsatile vascular compression of the eighth cranial nerve by AICA in the internal auditory canal results in demyelination at the level of central glial and peripheral non-glial junction. This also causes arachnoids adhesion, which manifests adhesion of the vessel and nerve. Neurovascular compression leads to impaired blood flow in the vascular loop result in impaired blood flow to the cochlea and vestibule, leading to inner ear dysfunction.

The diagnosis of vestibulocochlear compression is based on the clinical features and radiological findings. There are significant controversies for the neurotologic diagnosis of the neurovascular compression of the vestibulocochlear nerve. Abnormal ABR, brief episodes of vertigo, one side sensorineural hearing loss, one side tinnitus and abnormal vestibular features are reported in the neurovascular compression of the vestibulocochlear nerve. These clinical manifestations may be due to complex interaction between
the compressing artery and nerve which compromise the inner ear circulation. Stimulation of the nerve fibres by pulsation of the artery causes audiovestibular symptoms. As per a cadaver study, the AICA loops are seen inside the internal auditory canal in 12.3% of the human temporal bones. MRI is an important investigation for visualizing the vascular and neural components at the CPA and internal auditory canal. It also helps to evaluate the anatomical relationship between the cranial nerves and vascular structures around it.

The treatment is usually done by microsurgical decompression. The surgery of this condition should be performed by an expert neurosurgeon and lateral skull base surgeon. Microvascular decompression of the vestibulocochlear nerve is considered as the treatment with a high rate of success up to 80% for different vestibulocochlear compression syndrome. based on the clinical manifestations by loops of AICA, different skull base approaches are done such as retro-sigmoid, far-lateral, middle fossa, trans-labyrinthine, trans-cochlear, orbitozygomatic and combination of supratentorial-infratentorial pre-sigmoid approaches. In clinical practice, certain common surgical methods are used for AICA loops or aneurysms are retro-sigmoid sub-occipital approach with or without petrosectomy (posterolateral), sub-temporal middle fossa approach (lateral), orbitozygomatic approach and rarely transclival approach. The clipping of the AICA may pose difficulty because of its anatomical location and adjacent cranial nerves.

**CONCLUSION**

Intrameatal vascular compression of the vestibulocochlear nerve may result in disabling tinnitus and sensorineural hearing loss. The clinical manifestations with unilateral sensorineural hearing loss and tinnitus emphasize the importance for otolaryngologists to be vigilant and maintaining a high index of suspicion for vascular loop of AICA compressing the eighth cranial nerve in the internal auditory canal. So, the clinician or Otolaryngologists should always keep in mind the vascular loop of AICA as a differential diagnosis for unilateral sensorineural hearing loss and tinnitus. The imaging like MRI is very useful to find out such vascular loop in the internal auditory canal and can also be successfully treated by microvascular decompression.

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**REFERENCES**

1. Swain SK, Nayak S, Ravan JR, Sahu MC. Tinnitus and it is current treatment-Still an enigma in medicine. J of Formosan Med Assoc. 2016;115(3):139-144.
2. Voelker CC, Chole RA. Unilateral sensorineural hearing loss in adults: Etiology and management. In Seminars in Hearing. 2010; 32(4):313-325.
3. Swain SK, Sahu MC, Choudhury J. Sudden sensorineural hearing loss in children: Our experiences in a tertiary care teaching hospital of eastern India. Pediatric Polska-Polish J of Paed. 2018;93(2):127-131.
4. Miller LE, Miller VM. Safety and effectiveness of microvascular decompression for the treatment of hemifacial spasm: a systematic review. Br J Neurosurg. 2012; 26(4):438-444.
5. De Ridder D, Ryu H, Muller AR, Nowe V, Van de Heyning P, Verlooy J. Functional anatomy of the human cochlear nerve and its role in microvascular decompressions for tinnitus. Neurosurg. 2004; 54(2):381-390.
6. De Ridder D, De Ridder L, Nowé V, Thierens H, Van de Heyning P, Møller A. Pulsatile tinnitus and the intrameatal vascular loop: why do we not hear our carotids. Neurosurg. 2005; 57(6):1213-1217.
7. Sekhar LN, Schessel DA, Bucur SD, Raso JL, Wright DC. Partial labyrinthectomy petrous apicectomy approach to neoplastic and vascular lesions in petroclival area. Neurosurg. 1999; 44:550-552.
8. Zhang L, Yu Y, Yuan Y, Xu J, Xu X, Zhang J. Microvascular decompression of cochleovestibular nerve in patients with tinnitus and vertigo. Neurology Ind. 2012;60(5):495.
9. McDermott AL, Dutt SN, Irving RM, Pahor AL, Chavda SV. Anterior inferior cerebellar artery syndrome: fact or fiction. Clin Otolaryngol. 2003; 28:75-80.
10. Borghesi-Razavi H, Darvish O, Schick U. Disabling Vertigo and Tinnitus Caused by Intrameatal Compression of the Anterior Inferior Cerebellar Artery on the Vestibulocochlear Nerve: A Case Report, Surgical Considerations, and Review of the Literature. J Neurol Surg Rep. 2014; 75:47–51.
11. Balansard ChF, Meller R, Bruzzo M, Chays A, Girard N, Magnan J. Trigeminal neuralgia: results of microsurgical and endoscopic-assisted vascular decompression. Ann Otolaryngol Chir Cervicofac. 2003; 120:330-337
12. Reisser C, Schuknecht HF. The anterior inferior cerebellar artery in the internal auditory canal. Laryngosc. 1991;101: 761-766.
13. Turcotta EL, Patra DP, Abi-Aad KR, Welz ME, Weisskopf PA, Bendok BR. Microvascular decompression and transposition of the 8th cranial nerve using a fenestrated clip. World Neuros. 2020; 135:233.
14. Brackmann DE, Kesser BW, Day JD. Microvascular decompression of the vestibulocochlear nerve for disabling positional vertigo: the House Ear Clinic experience. Otol Neurotol. 2001; 22(6):882-887.