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Bilateral Cochlear Nerve Hypoplasia with Vestibular Nerve Agenesis: A Rare Cause of Congenital Sensorineural Hearing Loss

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Abstract | **Case Report**

Bilateral cochlear nerve hypoplasia associated with bilateral vestibular nerve agenesis is an exceptionally rare neuroimaging finding and an uncommon cause of congenital sensorineural hearing loss (SNHL). We present the case of a 50-year-old patient with a long-standing history of bilateral hearing loss since childhood, who underwent MRI of the internal auditory canals (IACs) for reassessment. High-resolution T2-weighted MRI demonstrated bilateral cochlear nerve hypoplasia, more pronounced on the right, and complete agenesis of both superior and inferior vestibular nerves bilaterally. This article emphasizes detailed imaging features, clinical significance, diagnostic pitfalls, and differentials. A comprehensive review of updated imaging literature supports recognition and appropriate classification of cochleovestibular nerve anomalies.

Keywords: Cochlear Nerve Hypoplasia, Vestibular Nerve Agenesis, Congenital Hearing Loss, MRI, Internal Auditory Canal, Cochleovestibular Anomalies.

INTRODUCTION

Congenital sensorineural hearing loss (SNHL) affects 1 to 3 per 1000 live births. While genetic causes are common, approximately 20% of congenital SNHL is due to inner ear malformations or cochleovestibular nerve anomalies. Cochlear nerve hypoplasia is rare, and even more so when associated with vestibular nerve agenesis. MRI is essential for identifying these conditions and guiding auditory rehabilitation strategies.

CASE PRESENTATION

A 50-year-old male with a history of bilateral non-progressive congenital hearing loss was referred for reassessment prior to potential cochlear implantation. Audiometry showed profound SNHL bilaterally. There were no vestibular symptoms. No history of prematurity, meningitis, or familial deafness was present.

Imaging Protocol

MRI was performed on a 1.5 T with a dedicated temporal bone protocol. Sequences included: 3D CISS (constructive interference in steady state), axial and coronal T2-weighted images, pre- and post-contrast T1.

Findings

- Bilateral hypoplasia of the cochlear nerves, defined as nerves with significantly smaller caliber than the adjacent facial nerves, more marked on the right.
- Agenesis of both superior and inferior vestibular nerves bilaterally, with absent delineation on CISS and T2 sequences.
- Internal auditory canals (IACs) of normal diameter (right: 3.2 mm; left: 3.4 mm), suggesting isolated neural hypoplasia rather than bony stenosis.
- Normal cochlear morphology bilaterally (2.5 turns present).
- No enhancement or mass lesion.

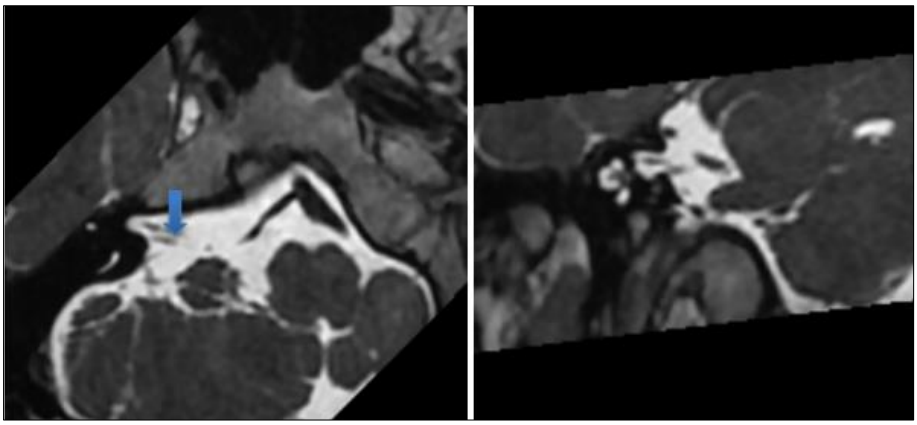

Figure 1: Axial 3D CISS MRI showing right and left cochlear nerve hypoplasia (arrow) compared to facial nerve

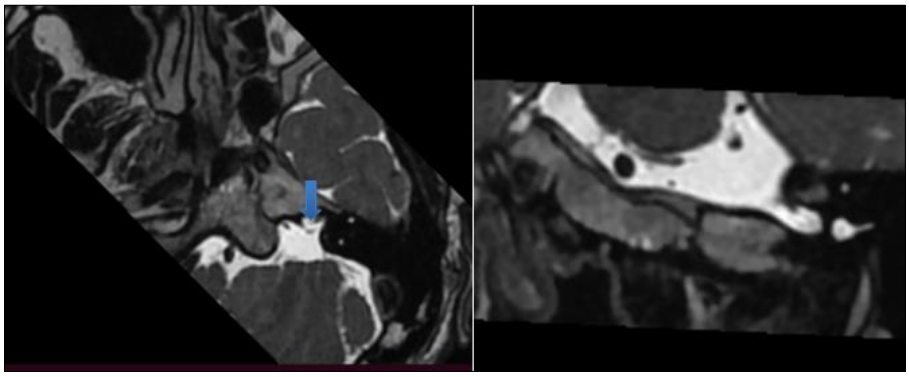

Figure 2: Coronal T2 image with absent superior and inferior vestibular nerves bilaterally

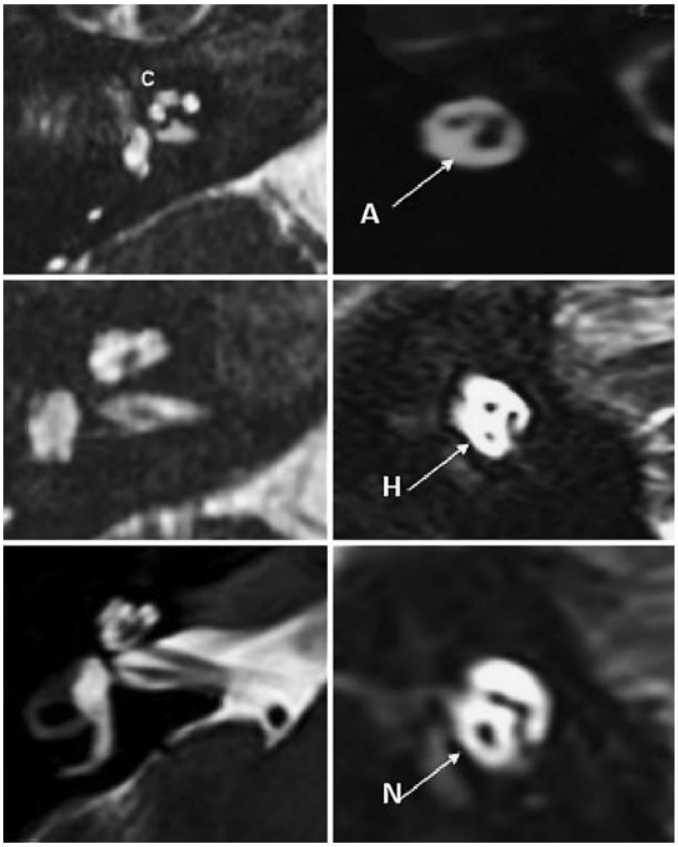

Figure 3: Diagrammatic illustration comparing normal vs hypoplastic/aplastic cochleovestibular nerve anatomy.

DISCUSSION

MRI is the modality of choice for cochleovestibular nerve assessment. High-resolution 3D T2 sequences (CISS/FIESTA) enable direct visualization of cranial nerves within the IAC. In normal anatomy, four nerves are seen: facial, cochlear, superior vestibular, and inferior vestibular. Hypoplasia is diagnosed when the cochlear nerve is significantly smaller than the facial nerve, and agenesis when the nerve is completely absent.

Cochlear nerve hypoplasia and vestibular nerve agenesis affect cochlear implant candidacy. Implantation is typically contraindicated in complete cochlear nerve aplasia but may be considered in hypoplasia with limited outcomes.

Differential Diagnoses

1. Cochlear nerve aplasia – complete absence of the nerve with often associated IAC stenosis.
2. Labyrinthine aplasia (Michel anomaly) – complete absence of inner ear structures.
3. Auditory neuropathy spectrum disorder (ANSD) – preserved nerve but impaired neural conduction, often diagnosed electrophysiologically.
4. Enlarged vestibular aqueduct syndrome (EVAS) – may coexist with cochlear nerve hypoplasia but shows different imaging characteristics.
5. IAC stenosis – diameter <2 mm, often seen in conjunction with nerve hypoplasia or aplasia.

CONCLUSION

Bilateral cochlear nerve hypoplasia with vestibular nerve agenesis is an exceptionally rare condition. MRI is critical for its diagnosis and for planning auditory rehabilitation. High-resolution T2 imaging allows accurate evaluation of the IAC contents and differentiation from other congenital causes of hearing loss.

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