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Internal auditory canal duplication with facial and cochlear nerve dysfunction: A case report

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Abstract

Internal auditory canal duplication is a rare anomaly of the temporal bone. The condition is diagnosed on performing High-Resolution Computed Tomography of the temporal bone and magnetic resonance imaging for sensorineural hearing loss. A bony septum divides the internal auditory canal into dual compartments. Duplication may be either unilateral or bilateral and vestibulocochlear nerve may be aplastic or hypoplastic. Rarely, patient may present with facial nerve palsy. A 26-year-old female presented with right grade IV lower motor neuron facial nerve palsy for 12 years and right-sided hearing loss for 9 years. Pure tone audiogram revealed 45 dB of moderate degree sensorineural hearing loss on the right ear. On High-Resolution Computed Tomography of the temporal bone, an incomplete bony septum was visualized in the right internal auditory canal, dividing it into two compartments. Internal auditory canal on the left side was normal. Other inner and middle ear structures were normal. On magnetic resonance imaging, both the vestibulocochlear and facial nerves were well visualized with normal calibers.

Keywords

Auditory canal, duplication, facial nerve, vestibulocochlear nerve

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Introduction

Internal Auditory Canal (IAC) duplication is a rare congenital malformation of the temporal bone.[1] Usually these cases are unilateral, however, few bilateral cases and a case of triplicated IAC have been reported.[2,3] Duplication of IAC is detected on High-Resolution Computed Tomography (HRCT) of the temporal bone, which could either be a partial or complete bony septum.[4] On magnetic resonance imaging (MRI), the vestibulocochlear nerve can be aplastic or hypoplastic.[4]

Duplication of IAC may be associated with other inner, middle, or external ear anomalies. Similarly, it may form a part of syndromic presentation such as Klippel–Feil syndrome.[1] The common symptom is congenital sensorineural hearing loss.[4] However, we report an unusual case of IAC duplication with facial palsy and sensorineural hearing loss. Only two cases of IAC duplication with facial palsy have been reported till date to the best of our knowledge.[4,5]

Case report

A 26-year-old female presented to outpatient department of Otorhinolaryngology with right-sided facial nerve palsy for 12 years and hearing loss on the same side for 9 years. She denied other aural and vestibular symptoms. There was no family history of sensorineural hearing loss. Her perinatal history was unremarkable, with no history of meningitis, otitis media, trauma, or ototoxicity.

On examination, normal tympanic membranes were noted bilaterally. Tuning fork test showed air conduction to be

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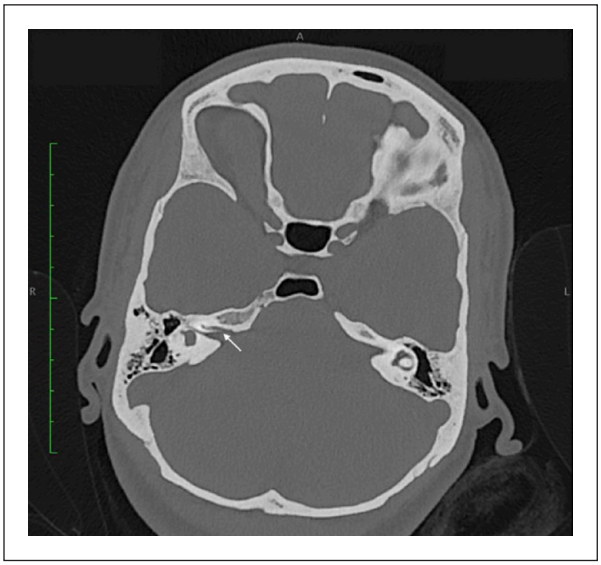

Figure 1. Bony septum in the IAC extending from fundus to two-third of its length of the canal medially (white arrow).

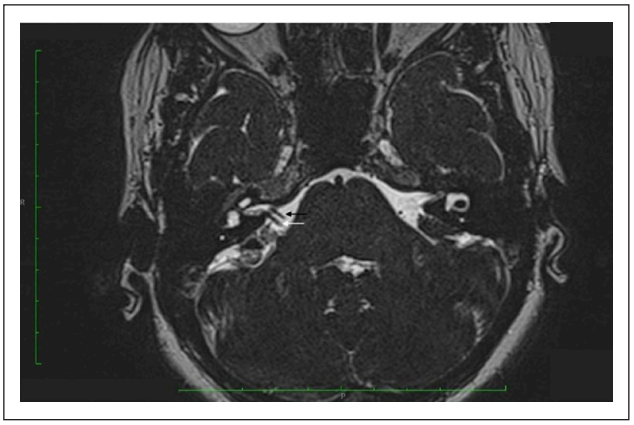

Figure 2. Axial T2 weighted MRI showing facial nerve (black arrow) and vestibulocochlear nerve (white arrow) in the right IAC.

greater than bone conduction in both the ears while Weber lateralized to the left ear. She had right House–Brackmann grade IV lower motor neuron facial nerve palsy.

Pure tone audiogram revealed 45 dB of moderate degree sensorineural hearing loss on the right ear. Hearing was normal on the left ear. On HRCT of temporal bone, an incomplete bony septum was visualized in the right IAC, dividing it into two compartments (Figure 1). The partition extended from the fundus of IAC till two-third of its length medially. IAC on the left side was normal. Other inner and middle ear structures were normal. On MRI, both the vestibulocochlear and facial nerves were well visualized with normal caliber (Figure 2). Mild deviation from usual course of the nerves were seen with small angulation at the level of internal auditory meatus. No altered signal intensity was seen in these nerves.

The patient was given an option to use hearing aid in her right ear, which she refused. She was advised for facial reanimation surgery and facial physiotherapy for right facial palsy. However, the patient lost to follow-up. Written informed consent was taken from the patient for the publication of this case report.

Discussion

The diameter of IAC ranges from 2 to 8 mm, with average of 4 mm.[6] The malformation of IAC such as atresia, stenosis, hypoplasia, and aplasia accounts for 12% of temporal bone anomalies.[5,7] The frequency of IAC duplication among patients with sensorineural hearing loss is 0.019%.[4]

IAC is labelled to be stenotic when IAC diameter is less than 2 mm.[8] IAC stenosis and IAC duplication are often confused. However, its vital to differentiate between these two conditions because patients with IAC stenosis have dysplastic vestibulocochlear (VIII) nerve and they could benefit with cochlear implantation (CI) while patients with IAC duplication have aplastic or hypoplastic VIII nerve, hence, CI is contraindicated.[4]

IAC duplication is usually unilateral and presents with congenital sensorineural hearing loss.[7,9] Cross-sectional image shows a bony septum dividing IAC into two separate canals. The VIII nerve is either hypoplastic or aplastic.[10] Facial (VII) nerve is normal in most of the reported cases.[5]

In our case, the patient was asymptomatic till 14 years of age when she had sudden onset of right facial palsy and gradual right-sided hearing loss from 17 years of age. HRCT of the temporal bone depicted a bony septum in the IAC while MRI delineated the normal course of facial and vestibulocochlear nerves. The cases reported in literature till date had sensorineural hearing loss and facial palsy since childhood. The late clinical presentation and absence of vestibular symptoms in our case may be due to normal size and signal intensity of both VII and VIII nerves. Facial palsy may occur in cases of IAC anomalies due to inflammation, compression or ischemia of the nerve.[11,12]

In 2011, Kew and Abdullah[5] reported the first case of vestibulocochlear nerve aplasia and facial nerve hypoplasia in a case of duplicated IAC. Wang et al.[4] reviewed HRCT and MRI of the temporal bones of patients with sensorineural hearing loss and they reported a case of facial nerve hypoplasia with IAC duplication in 2018 (Table 1).

Dual canal on HRCT temporal bone is the characteristic finding of duplicated IAC.[4] HRCT of temporal bone and MRI with multiplanar reconstruction and volume rendering must be done to identify IAC duplication.[10] High-resolution gradient echo MRI such as three-dimensional constructive interference in steady state best depicts the neural integrity.[5]

The patient in this study had moderate degree of unilateral hearing loss, therefore, use of hearing aid would have benefited her. However, those patients with profound degree

Table 1. Reported cases of IAC duplication with facial palsy.

Study	Age (years)	Sex	Side	Age of onset	Presentation	Hearing loss	Facial palsy	Radiological findings
Kew and Abdullah[5]	34	Male	Right	Since childhood	Facial palsy, vestibular dysfunction	Profound	Grade III	Aplastic VIII nerve and hypoplastic VII nerve
Wang et al.[4]	2	Female	Left	Since birth	Microtia, atresia of ear canal, facial palsy	Severe	Grade unknown	Hypoplastic VII nerve
Present study	26	Female	Right	14 years (facial palsy), 17 years (hearing loss)	Facial palsy, hearing loss	Moderate	Grade IV	Normal caliber of VII and VIII nerves

of hearing loss may require CI. The role of CI in cases of hypoplastic cochlear nerve is debatable. Cochlear implant failures have been reported in cases of IAC anomalies.[13] To the contrary, successful implantation have been achieved in few cases of patients with hypoplastic cochlear nerve.[1,14]

Facial nerve paralysis of long-standing duration could be managed by reconstructive surgeons using regional or free muscle transfer. Gracilis muscle is widely used for free muscle transfer during facial reanimation. Static techniques of facial reanimation such as lateral tarsorrhaphy, eyelid weight placement, medial canthopexy, and nasolabial fold modification may be necessary depending upon patient's clinical presentation.[15]

This case report would have been better if we had followed up the patient and mentioned about her progress after treatment. IAC duplication should be suspected when a patient presents with sensorineural hearing loss and facial palsy.

Conclusion

Duplication of IAC is a rare congenital anomaly of the temporal bone. The commonest presentation is sensorineural hearing loss. However, some patient may present with facial nerve palsy. HRCT of the temporal bone and three-dimensional high-resolution gradient echo MRI aid in its diagnosis.

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Author contributions

Conceptualization: Roshan Ghising and Kripa Dongol. Writing—original draft: Kripa Dongol and Roshan Ghising. Writing—review and editing: Kripa Dongol and Sundar Suwal. Approval of final article: all authors.

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Ethical approval

Our institution does not require ethical approval for reporting individual cases or case series.

Informed consent

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