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Imaging Findings of Bilateral Mondini Malformation Associated with Enlarged Vestibular Aqueduct: A Case Report

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Abstract | **Case Report**

Mondini malformation, also known as incomplete partition type II (IP-II), is one of the most common congenital inner ear malformations associated with sensorineural hearing loss. It results from arrested embryological development of the cochlea during the seventh week of gestation, leading to incomplete cochlear differentiation. Imaging plays a key role in diagnosis, classification, and preoperative evaluation before cochlear implantation. We report the case of a 26-year-old female presenting with bilateral progressive post-lingual sensorineural hearing loss referred for cochlear implant evaluation. High-resolution computed tomography (CT) of the temporal bones demonstrated bilateral incomplete cochlear partition characterized by reduction of the cochlea to one and a half turns, absence of the modiolus, cystic dilatation of the apical turn, enlarged vestibule and enlarged vestibular aqueduct. Magnetic resonance imaging (MRI) confirmed the presence of intact cochlear nerves without associated intracranial abnormalities. This case highlights the essential role of CT and MRI in the diagnosis of Mondini malformation and in guiding surgical planning before cochlear implantation.

Keywords: Mondini malformation; Incomplete partition type II; Enlarged vestibular aqueduct; Cochlear implant; CT; MRI.

INTRODUCTION

Congenital inner ear malformations are an important cause of sensorineural hearing loss and account for nearly 20% of congenital sensorineural hearing loss cases with radiologically detectable abnormalities. Mondini malformation results from arrested cochlear development during the seventh week of gestation and corresponds to incomplete partition type II according to the Sennaroglu classification. It is characterized by a 1.5-turn cochlea, absent or hypoplastic modiolus, and frequently associated enlarged vestibular aqueduct. Patients usually present with congenital or progressive sensorineural hearing loss, sometimes associated with vestibular symptoms. High-resolution CT and MRI are essential for diagnosis, classification, and preoperative evaluation before cochlear implantation.

CASE REPORT

A 26-year-old female patient presented with progressive bilateral post-lingual sensorineural hearing loss and was referred to our department for radiological evaluation prior to cochlear implantation. There was no history of meningitis, otologic surgery, head trauma, or congenital syndromic disorder. Clinical examination showed no external or middle ear abnormality.

High-resolution computed tomography (CT) of the temporal bones demonstrated bilateral incomplete cochlear partition with reduction of the cochlea to one and a half turns, absence of the modiolus, cystic dilatation of the apical turn, enlarged vestibule and enlarged vestibular aqueduct, consistent with Mondini malformation.

Magnetic resonance imaging (MRI) of the cerebellopontine angles confirmed the presence of intact cochlear nerves without associated intracranial abnormalities.

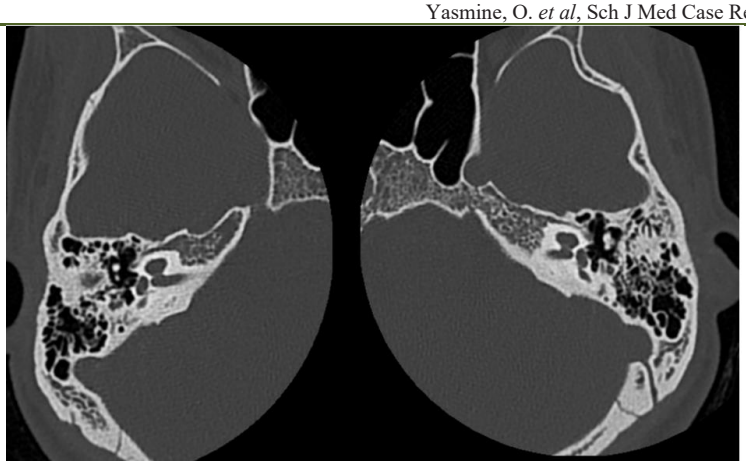

Figure 1: Axial HRCT image demonstrating incomplete partition type II (Mondini malformation) characterized by a preserved basal turn, cystic dilatation of the apical cochlea, reduction of the cochlea to 1.5 turns, absence of the modiolus, and associated enlarged vestibular aqueduct.

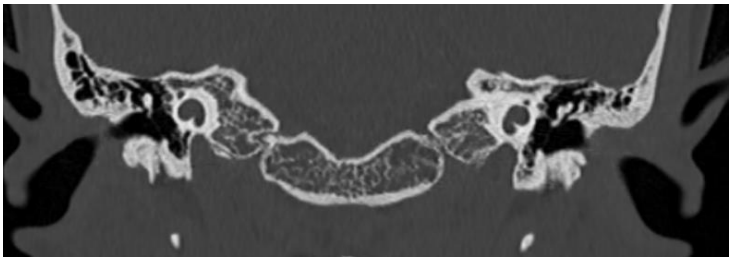

Figure 2: Coronal HRCT image showing a 1.5-turn cochlea with cystic dilatation of the apical turn, consistent with Mondini malformation

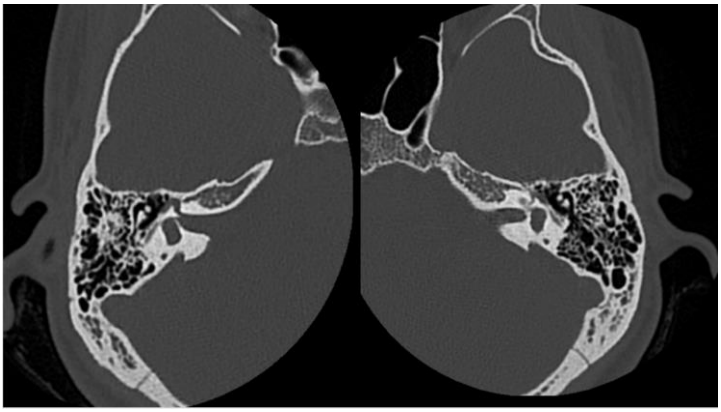

Figure 3: Axial HRCT image demonstrating enlarged vestibule associated with cochleovestibular malformation

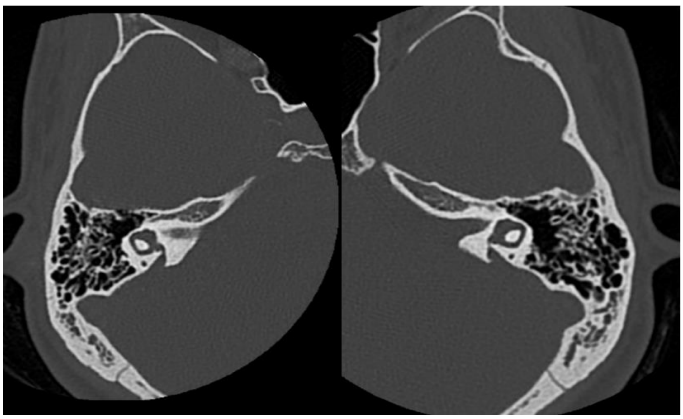

Figure 4: Axial HRCT image showing enlarged vestibular aqueduct associated with incomplete partition type II

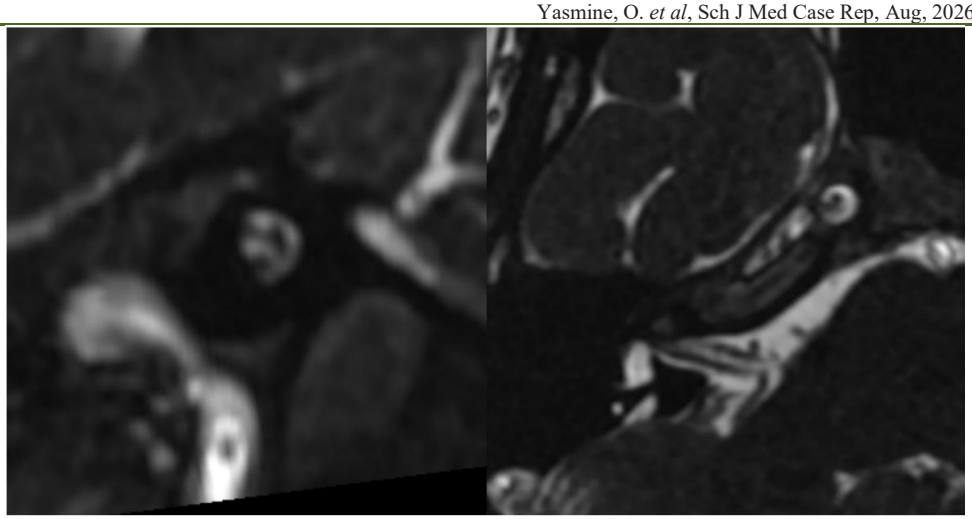

Figure 5: High-resolution T2-weighted MRI demonstrating preservation of the right cochlear nerve

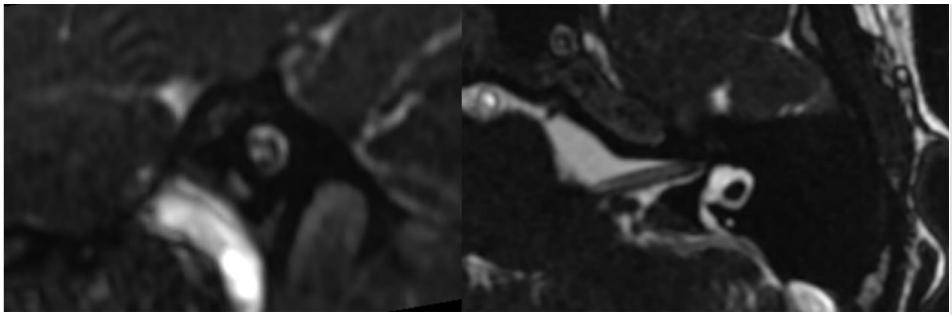

Figure 6: High-resolution T2-weighted MRI demonstrating preservation of the left cochlear nerve

DISCUSSION

High-resolution computed tomography (CT) remains the gold standard for evaluating osseous labyrinthine abnormalities. In Mondini malformation, CT typically demonstrates incomplete partition type II characterized by a cochlea reduced to one and a half turns, absence or hypoplasia of the modiolus, cystic dilatation of the apical turn, and frequently associated enlarged vestibular aqueduct [1]. These characteristic findings were clearly identified in our patient.

MRI plays a complementary role by evaluating the membranous labyrinth and confirming the presence of the cochlear nerve, which is essential before cochlear implantation [2]. Accurate radiological classification is important to differentiate Mondini malformation from other congenital inner ear anomalies such as incomplete partition type I, common cavity deformity, and cochlear hypoplasia, as prognosis and surgical management differ between these entities [3].

The association between Mondini malformation and enlarged vestibular aqueduct is clinically significant because it may contribute to progressive or fluctuating sensorineural hearing loss [4]. The major surgical concern in these patients is the occurrence of perilymphatic or cerebrospinal fluid gusher during cochleostomy due to abnormal communication between the inner ear and subarachnoid space [5]. Therefore, preoperative imaging evaluation is essential for surgical planning and risk assessment.

Despite these surgical challenges, cochlear implantation generally provides satisfactory auditory outcomes when the cochlear nerve is preserved [6].

CONCLUSION

Mondini malformation is an important congenital inner ear anomaly associated with sensorineural hearing loss. High-resolution CT and MRI are complementary imaging modalities that allow accurate diagnosis, anatomical classification, and evaluation of associated abnormalities such as enlarged vestibular aqueduct. In addition to confirming the integrity of the cochlear nerve, preoperative imaging plays a crucial role in surgical planning and in anticipating intraoperative complications, particularly perilymphatic gusher during cochlear implantation. Early and precise radiological assessment is therefore essential to optimize therapeutic management and postoperative auditory outcomes.

REFERENCES

1. Sennaroglu L, Saatci I. A new classification for cochleovestibular malformations. Laryngoscope. 2002 ;112 :2230–2241.

2. Casselman JW, Offeciers FE, Govaerts PJ, *et al.,*MR imaging of congenital malformations of the inner ear. Radiology. 1997 ;202 :773–781.
3. Jackler RK, Luxford WM, House WF. Congenital malformations of the inner ear : a classification based on embryogenesis. Laryngoscope. 1987 ;97 :2–14.
4. Sampaio AL, Cureoglu S, Schachern PA, *et al.,* Massive endolymphatic sac and vestibular aqueduct in Mondini dysplasia. Arch Otolaryngol Head Neck Surg. 2004 ;130 :678–680.
5. Prasad SC, La Melia C, Medina M, *et al.,*Perilymphatic gusher in cochlear implantation. Otol Neurotol. 2021 ;42: e425–e432.
6. Papsin BC. Cochlear implantation in children with anomalous cochleovestibular anatomy. Laryngoscope. 2005 ;115 :1–26.