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Case Report

# Flocculus Herniation into the Internal Acoustic Canal in Incomplete Partition Type I: A Case Report

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In this study, we present the first case with cerebellar herniation into the internal acoustic canal in incomplete partition type I anomaly. Cerebellar herniation into the internal acoustic canal is very rare with only a few cases reported in the literature. Although it is a rare clinical situation, cerebellar herniation into the internal acoustic canal may be seen in patients with incomplete partition type I. We presented magnetic resonance imaging findings of a 3-year-old girl with a history of meningitis, middle ear effusion, and bilateral congenital sensorineural hearing loss. Magnetic resonance imaging showed bilateral incomplete partition type I malformation and an additional flocculus herniation into the right internal acoustic canal. In the presented case, predisposition to cerebrospinal fluid leak in incomplete partition type I anomaly may be the reason for cerebellar herniation into internal acoustic canal. Also, possible increased intracranial pressure due to meningitis may be a contributing factor.

**KEYWORDS:** Cochlea, sensorineural hearing loss, cerebellar herniation, internal acoustic canal

## INTRODUCTION

Incomplete partition type I anomaly (IP-I), one form of cochlear partition abnormalities, occurs due to an earlier developmental arrest in the otic placode (in the fifth week) and it is characterized by sensorineural hearing loss (SNHL).[1] The absence of internal architecture within the cochlea, including absent modiolus and cribriform plate with a lack of interscalar septum (ISS) leads to a cystic appearance.[2] Dilated vestibule usually accompanies IP-I, which is also known as cystic cochleovestibular malformation. The vestibular aqueduct can be normal or enlarged.[3] Internal acoustic canal (IAC) can be normal, enlarged, or atretic.[4] Semicircular canals (SCC) can also be dysplastic.[1,3] Patients with IP-I may have normal, hypoplastic, or aplastic cochleovestibular nerves, and cochlear implantation is a treatment method with a high risk of cerebrospinal fluid (CSF) gusher in cases with intact cochlear nerves.[3,5]

Herein we report flocculus herniation into the IAC in a patient with IP-I anomaly, which may affect patient management and treatment outcome by increasing complications. Although there are a few cases with IAC herniation caused by mainly increased intracranial pressure (ICP), to the best of our knowledge this is the first case with cerebellar herniation into the IAC in IP-I anomaly.

## CASE PRESENTATION

A 3-year-old girl with a history of post-natal meningitis and middle ear effusion was admitted to our hospital. The patient also had a history of congenital SNHL detected on post-natal screening. A routine temporal bone magnetic resonance imaging (MRI) was performed using a standard head coil on a 1.5 T MRI scanner. The temporal bone protocol included axial and coronal T1 weighted imaging (WI) (TR/TE: 350/8.1 ms, slice thickness: 2 mm) and axial T2WI (TR/TE: 3760/150 ms, slice thickness: 2 mm), and axial and sagittal oblique 3-dimensional (3D) constructive interference in steady-state (CISS) sequences (FOV: 150 × 170, TR/TE: 13/6.5 ms, slice thickness: 1 mm).

Magnetic resonance imaging revealed a bilateral IP-I anomaly characterized by bilateral cystic cochleae, dilated vestibules, and dilatation of proximal parts of SCCs. Modioli and ISSs could not be visualized and the cochlear apertures were enlarged. The endolymphatic duct was enlarged on the left with a measurement of 1.7 mm with the right counterpart being at the upper normal limit (0.9 mm).

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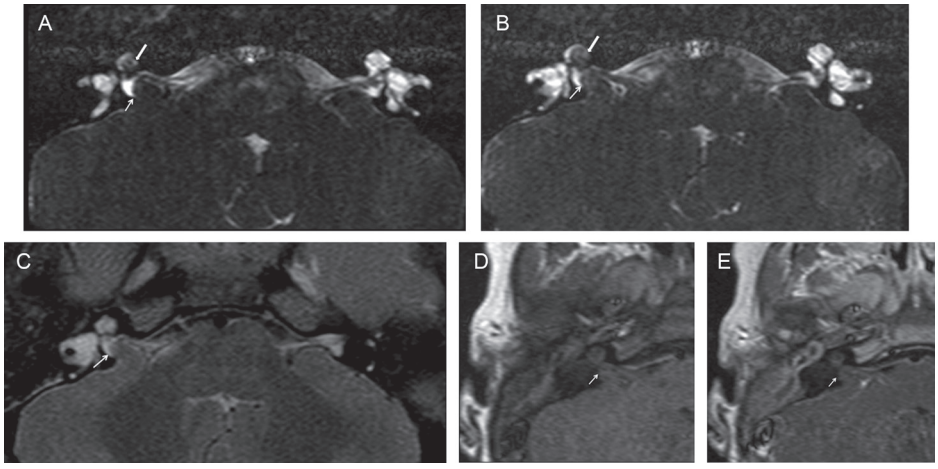


**Figure 1** . Axial heavily T2WI (CISS) (A and B) show bilateral IP-I malformation. Bilateral modioli and interscalar septum could not be visualized. On the right side, there was herniation of the cerebellar flocculus into the internal acoustic canal (thin arrows). Herniated floccular tissue is isointense compared to the cerebellum on axial CISS (A and B), T2WI (C), pre-contrast T1WI (D), and postcontrast T1WI (E). Coexisting right intracochlear hypointensity on CISS was considered likely to represent fibrous tissue due to previous meningitis (A and B, thick arrows) and showed no contrast enhancement on postcontrast T1WI (e). An enlarged left endolymphatic duct is also seen (b; VA). CISS, constructive interference in steady-state; IP-I, incomplete partition type I; T1WI, T1 weighted imaging; T2WI, T2 weighted imaging.

In addition, MRI also demonstrated a solid structure isointense to the cerebellum in all sequences, in continuity with the flocculus of the cerebellum extending from the cerebellopontine angle cistern into the right IAC (Figure 1). No apparent evidence of raised ICP was observed on the T2WI of the brain. Additionally, CISS images showed intracochlear hypointensity on the right side (Figure 1). This intracochlear hypointensity was not isointense with flocculus on T1WI and T2WI and showed no contrast enhancement on postcontrast T1WI. Additionally, no distortion or signal abnormality in the right cerebellum was detected.

The patient was lost to follow-up and no further examination could be done.

The present case report did not require ethical approval based on the rules of the local ethics committee. Appropriate patient informed consent and permission from the legal guardian of the patient were received for publication of this case report including case details or images of the patient.

## DISCUSSION

Brain stem or cerebellar herniation into IAC which can cause hearing loss is very rare with only a few cases reported in the literature.[6-9] Michiwaki et al[6] reported unilateral flocculus herniation into the IAC and hearing loss in an adult patient due to the mass effect of a large tentorial meningioma and concomitant hydrocephalus. Two pediatric cases with multiple cranial nerve palsy (vision loss, hearing loss, and ocular and facial paralysis) due to the unilateral brainstem herniation into the IAC were previously reported.[7,8] One of these cases had apparent hydrocephalus. In the other one, no apparent finding for increased ICP was present; however, raised ICP was detected with lumbar punction (opening pressure was 50 cm $H_2O$).[8] No accompanying inner ear abnormality was mentioned in these previous reports; however, both cases had a history of myringotomy operations. The main reason for herniation was concluded as increased ICP in both, supported by the regression of herniation into IAC following surgical procedures (tumor resection and ventriculoperitoneal shunt).

In our case, cerebellar herniation was also unilateral without apparent evidence of increased ICP. Previously reported symptoms of herniation into IAC include sudden hearing loss, facial paralysis, and vestibular balance disorder. Our case had already congenital SNHL and no additional symptoms. Additionally, our case had a history of post-natal meningitis with no MRI study at that time. Therefore, the occurrence time of IAC herniation was not clear. Thus, we cannot completely exclude an increase of ICP due to meningitis as an etiologic factor. The right intracochlear hypointensity on CISS was probably compatible with fibrous tissue as a sequel of meningitis. However, there was no follow-up imaging or histopathological confirmation; thus, additional intracochlear flocculus herniation was not completely excluded.

Nozawa et al[9] reported a pediatric case with multiple lytic lesions in the temporal bones diagnosed as Gorham-Stout disease of the skull base who presented with sudden onset right SHNL due to bilateral cerebellar herniation into IACs. The patient had a history of repeated CSF rhinorrhea and bacterial meningitis and left-sided hearing loss. Clinical symptoms and radiological findings of bilateral cerebellar herniation were improved after antiangiogenic therapy.[9] In that case, the bone fragility secondary to the osteolytic lesions of the skull base was accused for cerebellar herniation into IAC.

Development of raised ICP is one of the complications of meningitis due to complex pathophysiologic mechanisms, including obstruction of CSF flow (interstitial edema and hydrocephalus), the effect of toxic factors from pathogen and neutrophils on cellular elements (cytotoxic edema), increased blood-brain permeability (vasogenic edema).[10] In our case, no MRI study was present at the time of meningitis, and the presence of accompanying raised ICP secondary to meningitis is not clear but cannot be completely excluded. In the setting of inner ear abnormality, an accompanying CSF leak secondary to the preformed CSF pathway may be the reason for meningitis and/or cerebellar herniation into IAC in our case. Spontaneous CSF fistula and defect in stapes footplate and CSF leak in IP-I cases were previously reported.[2] In IP-I, high CSF pulsation at the stapes footplate

secondary to open communication between cochlea and IAC, is a risk for the development of CSF fistula. . However, the patient was lost to follow-up, and further interpretation could not be done with confidence.

## CONCLUSION

Herein, we aimed to draw attention to an IP-I case with flocculus herniation into the IAC. In this case with IP-I with a known history of meningitis, a predisposition to CSF leak in this anomaly may be the reason for cerebellar herniation into IAC. Also, a possible increased ICP due to meningitis may be a contributing factor. Awareness of predisposition to CSF leak and risk of meningitis and accompanying increased ICP is crucial for appropriate diagnosis and early treatment to prevent further development of herniation into IAC in IP-I.

**Ethics Committee Approval:** N/A.

**Informed Consent:** Appropriate patient informed consent and permission from the legal guardian of the patient were received for publication of this case report including case details or images of the patient.

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