Clinical Case Report

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Dehiscent high-riding jugular bulb presenting as conductive hearing loss

A case report

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Abstract

Rationale: Jugular bulb anomalies are asymptomatic lesions commonly seen in routine practice. However, some patients with jugular bulb anomalies may present with symptoms such as tinnitus or conductive hearing loss (CHL).

Patient concerns: A 9 year old boy complained right sided hearing disturbance without any vestibular symptoms. Pure tone audiometry (PTA) revealed a mild right sided conductive hearing loss. Otoscopy showed a red-purplish mass like lesion in his right middle ear cavity, which was regarded as hypervascular tumor.

Diagnosis: Based on otoscopic findings, preliminary differential diagnoses included jugular bulb anomaly, hemotympanum, cholesterol granuloma and paraganglioma.

Interventions: We performed contrast enhanced computed tomography of the temporal bone (TBCT).

Outcomes: CT scan showed and enhancing lesion which was bulging from his right jugular foramen to the middle ear with dehiscent jugular bulb. The lesion showed the same degree of contrast enhancement with the venous vasculature. This lesion contacted with the tympanic membrane, incudostapedial joint and round window, which might be attributable to interruption of sound transmission mechanics. Otherwise there was no evidence of mass or trauma related lesions in the temporal bone.

Lessons: Although most of jugular bulb anomalies are asymptomatic, patients may present with conductive hearing loss due to the interference of sound transmission mechanics.

Abbreviations: CHL = conductive hearing loss, HRJB = high riding jugular bulb, PTA = pure tone audiometry, TBCT = temporal bone computed tomography, TM = tympanic membrane.

Keywords: cochlear round window, computed tomography, conductive, ear ossicles, hearing loss, jugular vein

1. Introduction

A jugular bulb results from dilatation of the upper bulbous portion of the jugular vein. Although their size, location, and configuration vary, most jugular bulbs lie below the hypotympanum.[1] Jugular bulb anomalies such as high-riding jugular bulb (HRJB), dehiscence of the jugular bulb, and jugular diverticulum are often encountered in clinical practice, and in most patients, they are asymptomatic. However, in rare cases, patients may present with symptoms such as tinnitus and more rarely, conductive hearing loss (CHL).[1–4]

The possible mechanisms of CHL related to dehiscent HRJB included contact with the tympanic membrane (TM), interfering the ossicles, and involvement of the round window.[1,3,5,6] There were several case reports describing the association between HRJB and CHL, which showed only some part of known mechanisms. Here, we present the case of a child with CHL related to a dehiscent HRJB, presenting all the proposed attributable mechanisms on temporal bone computed tomography (TBCT).

2. Case report

A healthy 9-year-old male visited our otology clinic because of persistent right-sided hearing loss. He had no history of recurrent ear infection or trauma, nor did he complain of other auditory or vestibular symptoms. On otoscopy, a mass-like lesion with red-purplish discoloration was observed behind his right TM; this lesion was suspected to be a hypervascular mass or vascular anomaly (Fig. 1). Pure-tone audiometry revealed 10 to 15 dB right-sided CHL with a 5 dB speech reception threshold.

Contrast-enhanced TBCT was performed to explore the possibility of a vascular anomaly or middle ear tumor. The axial TBCT image showed a dehiscent HRJB protruding into the hypotympanum of the middle ear and contacting both his TM

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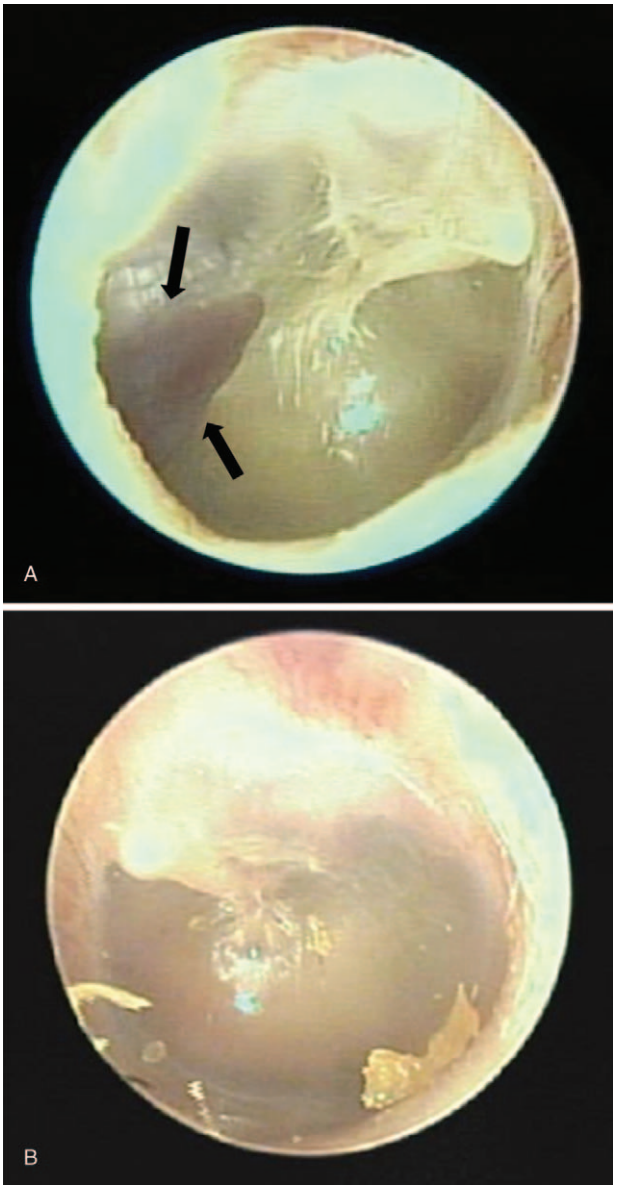

Figure 1. Otoscopic images of the right ear show a purplish discoloration in the posteroinferior quadrant of the right tympanic membrane (arrows). The appearance of the left tympanic membrane is normal.

and incudostapedial joint. The ectatic vascular structure was superimposed on the round window niche, which was visualized on the axial images (Fig. 2). Given the nature of his middle ear lesion and the mild hearing loss, clinical follow-up of the patient was discussed with his parents. He is on regular follow-up without any deterioration of hearing function for 2 years.

3. Discussion

The jugular bulb is usually covered with a bony plate and located on the floor of the hypotympanum.[1] Its size and location vary among individuals, and anomalies are frequently seen on otoscopy, during surgery, or in imaging studies.[7] Several definitions of HRJB have been proposed,[1,3–6] with the incidence ranging from 6% to 34%, and that of a dehiscent jugular bulb reported as 3.9%.[1,2,4,6,8–11] Because the jugular vein and dural sinuses are larger on the right, HRJB has a right-sided predominance.[4,6,11] On physical examination, it can be confused with vascular and nonvascular masses, and when covered by bone, its whitish appearance causes it to resemble a cholesteatoma. HJRB may also appear as a red, blue, or purplish discoloration in the posteroinferior quadrant of the TM, in which case, it will resemble a hypervascular tumor such as glomus tympanicum, a hemorrhage, or a cholesterol granuloma.[3,5,7]

The clinical significance of HRJB is that it can cause fatal hemorrhage because of inadvertent puncture in patients undergoing myringotomy or tympanoplasty.[7,8–10] Although in most cases, HRJB is incidentally detected, a small number of patients may suffer hearing loss, tinnitus, and vertigo.[1–4,6] Tinnitus is the most common symptom (50.4%), whereas hearing disturbance is rare (1.9%).[9] In those patients with sensorineural hearing loss, the mechanism is thought to involve and compress the vestibular aqueduct.[3,9,12] Several reports in the literature have described CHL related to jugular bulb anomalies. In these cases, 3 mechanisms have been proposed: contact between TM and the jugular bulb, ossicular chain interference, and round window niche obstruction.[1–3,5,6] All 3 mechanisms occurred in our patient and likely explain the hearing loss.

Round-window niche obstruction is considered the most likely mechanism of CHL resulting from a HRJB owing to the proximity of the round window niche to the floor of the middle ear cavity.[2,3] Previous surgical and imaging studies showed a high incidence of round window obstruction in patients with HRJB. Our patient demonstrated mild CHL in the low frequencies, in agreement with a previous report.[3] The low frequencies are primarily affected because the jugular bulb increases the stiffness of the conducting structures by the 3 mechanisms described above. Our patient shows all 3 of proposed mechanisms on TBCT, when patients included in the previous literature demonstrated only a single or 2 mechanisms on imaging.

Surgical correction of HRJB is not recommended because it could disrupt venous flow and increase the risk of intracranial hypertension. Removing the protruded jugular bulb from the affected middle ear was shown to not improve hearing function.[3,7] Moreover, clinicians and parents should be aware of the fact that myringotomy to correct the CHL may be life-threatening for the patient and should be avoided.

4. Conclusion

Our case illustrates a rare finding of a dehiscent HRJB in a pediatric patient presenting with CHL. The CHL associated with HRJB could be attributable to the following 3 mechanisms: contact with the TM, ossicular chain interference, and round-window niche obstruction. Clinicians should be aware that jugular bulb anomalies can cause CHL by interfering with the mechanics of sound transmission. However, surgical correction of jugular bulb anomalies is not recommended.

Author contributions

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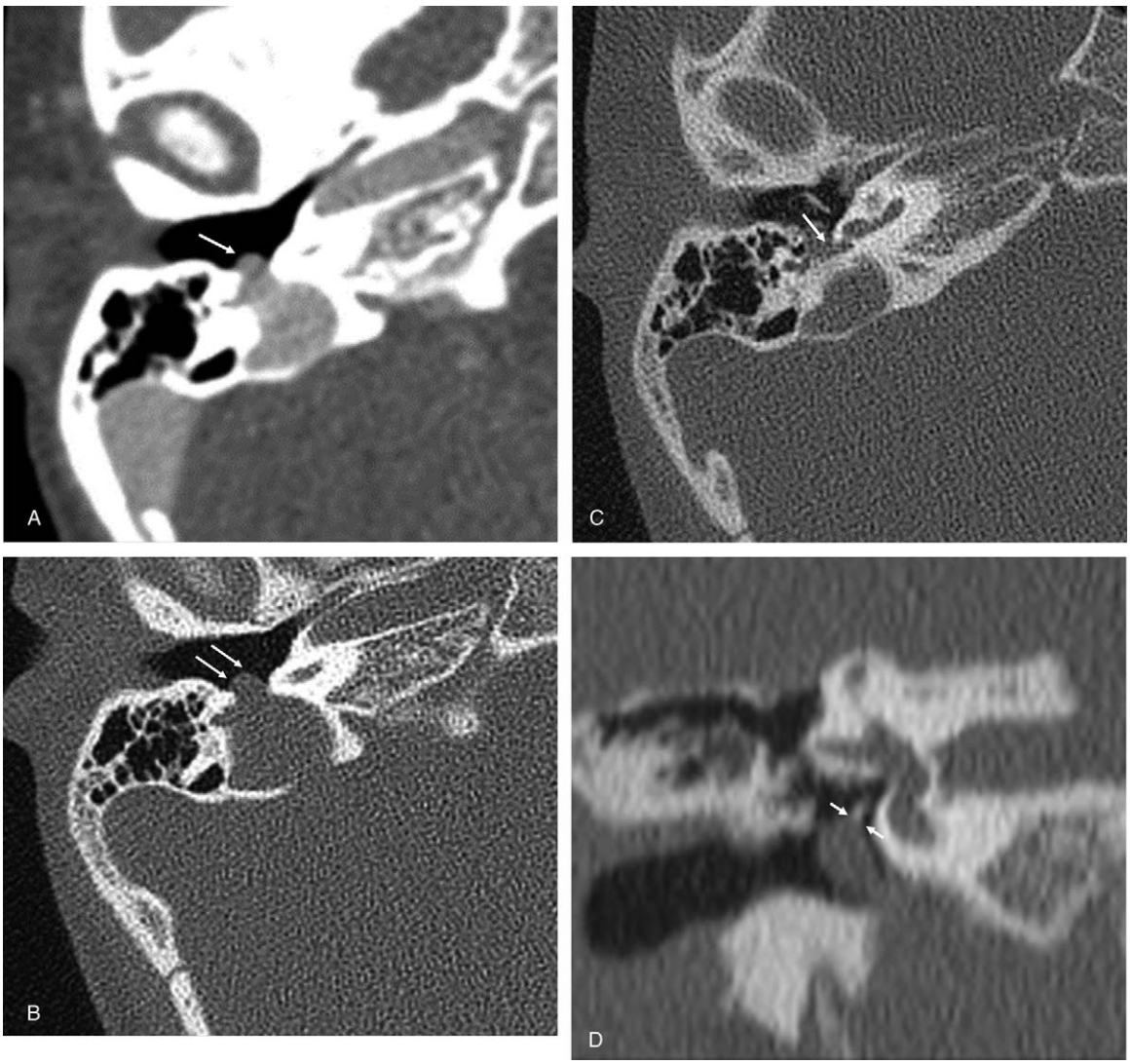

Figure 2. Computed tomography of the right temporal bone. (A) Axial contrast-enhanced images show a high-riding dehiscent jugular bulb protruding into the middle ear cavity. (B) The pre-contrast axial images show contact between the jugular bulb and the tympanic membrane (arrow). (C) The axial image shows the jugular bulb obstructing the round window (arrow). (D) Coronal images reveal contact between the jugular bulb and the incudostapedial joint (arrow).

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