Sensorineural hearing loss caused by a high jugular bulb

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Abstract

A 15-year-old female presented with sensorineural hearing loss related to a high jugular bulb. She noticed temporary worsening of her right hearing with mild dizziness when her neck was compressed whereas she noticed no change in her left hearing. An audiogram showed temporary worsening of low tone bone-conduction on such occasions. A large jugular bulb covering the right round window was observed through the perforation. Computed tomography (CT) revealed a large jugular bulb obliterating the round window niche. Deterioration of bone conduction during neck compression was thought to be caused by the protrusion of the jugular bulb into the inner ear via the round window.

This case suggested the possibility of sensorineural hearing loss due to the jugular bulb. As in the present case, jugular bulb or vein-related ear disorders should be examined by neck compression.

Key words: Hearing loss, Sensorineural; Jugular Veins

Case report

A 15-year-old female with anaemia and thrombocytopenia of unknown cause was referred with left otorrhoea. She had noticed slight right hearing loss on lying down several years before consultation.

Both of the tympanic membranes were moderately perforated. A white, slightly bluish mass protruded into the inferior-posterior part in the right tympanic cavity, almost hiding the round window (Figure 1). The mass was covered with bone and a small dehiscence was seen. From this dehiscence, the venous wall could be observed and it bulged with pulsation when her neck was compressed on either side. A part of the venous wall seemed to appear in the gap between the promontory and mass during compression. The venous walls also bulged during the Valsalva manoeuvre, when lying down or straining. However, this mass and venous walls did not make contact with either the ossicles or the membrane.

During the neck compression, etc., she complained of a mild hearing loss and ear fullness only on the right side. She also noticed a temporary slight dizziness on such occasions. The audiogram showed conductive hearing loss with bilateral sensorineural hearing impairment at high frequency. Sensorineural hearing loss was also experienced at low frequencies on the right (Figure 2). Both air and bone conduction in the right ear deteriorated during neck compression (Figure 3). Békésy audiometry also showed

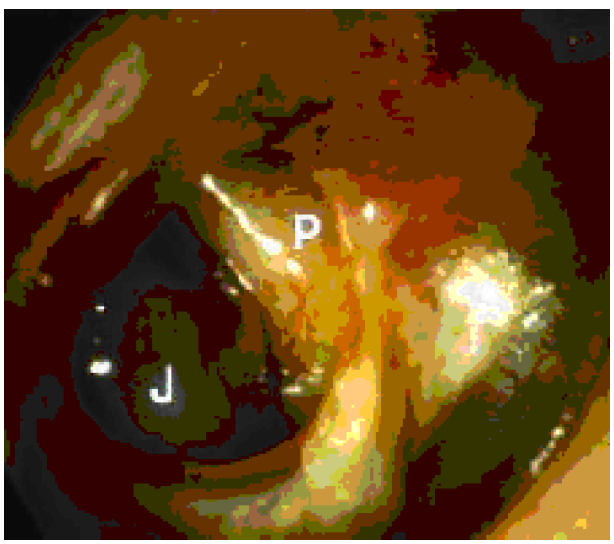


Fig. 1
Right ear of patient. J: Jugular bulb, P: Promontory.

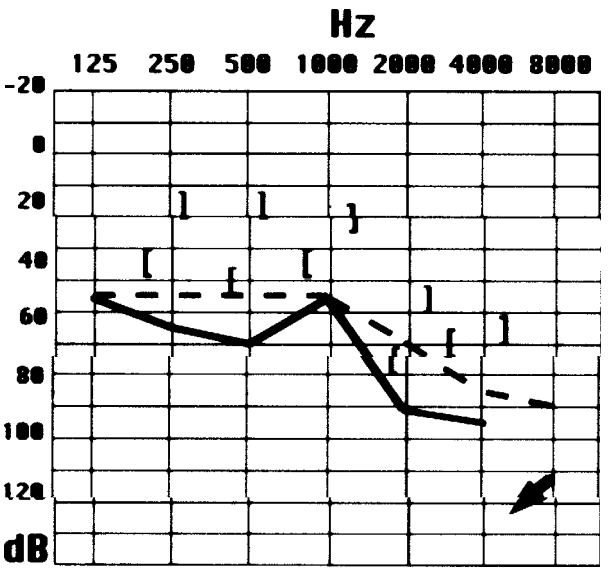


Fig. 2
Audiogram of patient. Dotted line indicates left air-conduction.

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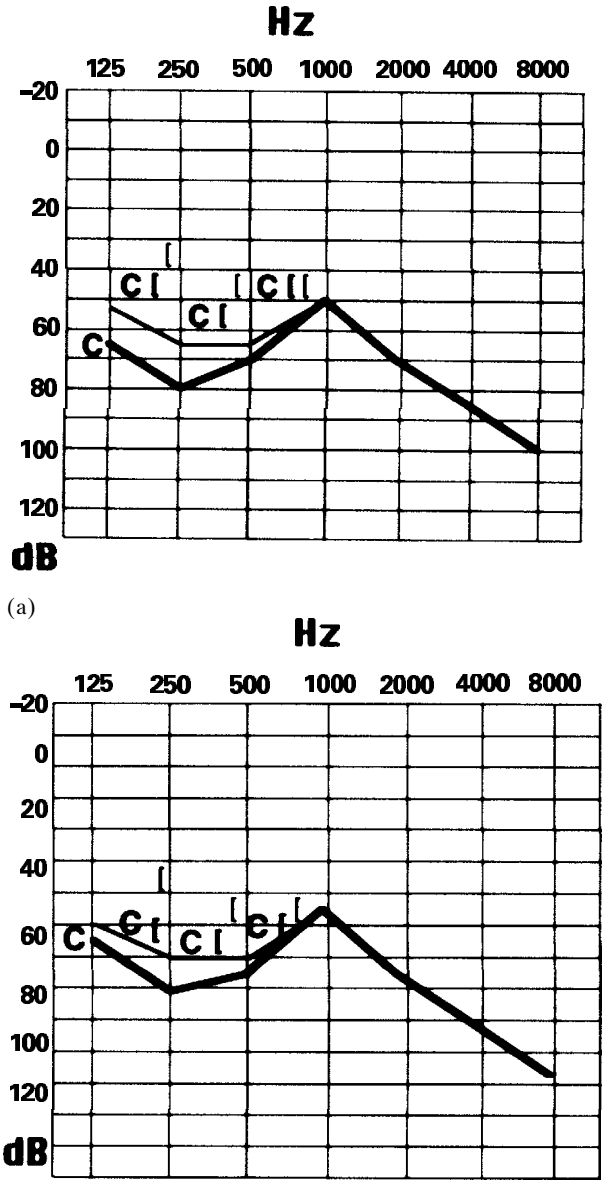


FIG. 3

Audiogram of the right ear during right neck (a) and left neck (b) compression. Neck compression (C) affected not only air conduction but also bone conduction.

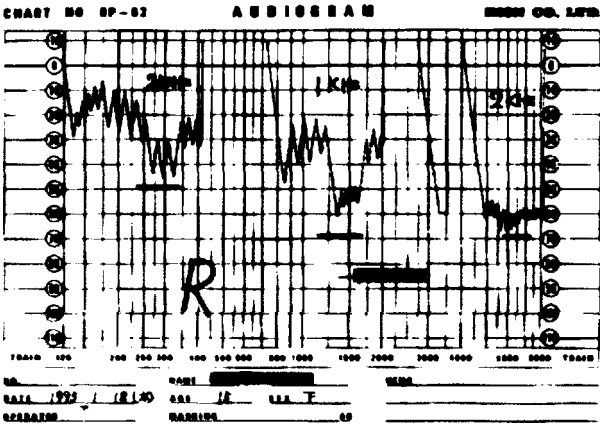


FIG. 4

Békésy's audiogram of bone-conduction of 250, 1 K and 2 KHz. Bars indicate neck compression.

worsening of hearing during neck compression, however, the recruitment phenomenon was not obvious at these frequencies (Figure 4). The worsening of hearing immediately recovered when the neck compression was stopped. The left hearing did not change. There were no remarkable changes in auditory brainstem responses and electrocochleograms with or without compression, however, thorough assessment was impossible due to the difficulty of examination. No obvious eye movement nor nystagmus were seen either on Frenzel goggle nor electronystagmogram during neck compression.

CT scan revealed a large jugular bulb in the right ear. The superior part of the bulb was divided by labyrinthine bone and the lateral part of the bulb obliterated the round window niche. A bony septum between the bulb and round window was not apparent (Figure 5).

Re-examination of this patient over the past five years have resulted in a continuation of these findings and symptoms.

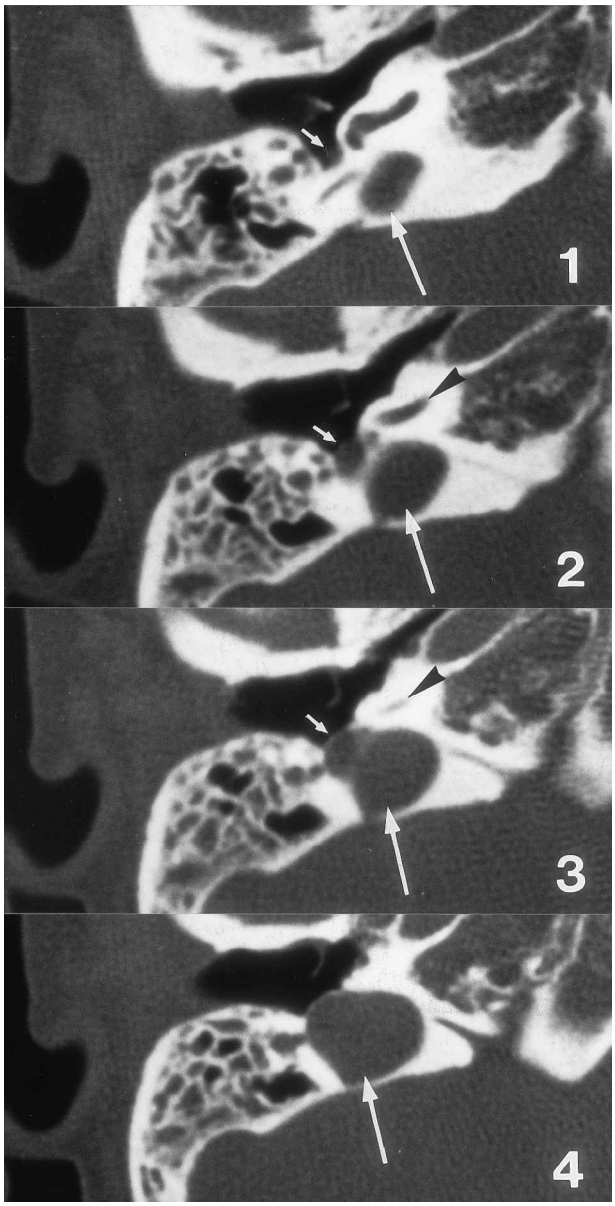


FIG. 5

CT scan. Each slice is at an interval of 2 mm. From above (1) to below (4). Arrow = jugular bulb; wedge = basal turn of the cochlea.

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Discussion

It is well known that the jugular bulb can cause conductive hearing loss and tinnitus.[1–5] In this case, however, the right tympanic membrane had a perforation and the jugular bulb touched neither the tympanic membrane nor the ossicles. In addition, whilst the neck was compressed, the patient complained of a hearing loss with temporary dizziness. Deterioration of bone conduction at low frequencies was also detected on audiograms. CT scan revealed that the jugular bulb obliterated the round window niche and the bony septum was not apparent. These findings suggested a relationship between the jugular bulb and inner ear dysfunction.

Neck compression, Valsalva manoeuvre, straining or a supine position increased venous pressure in the jugular bulb and caused enlargement of the bulb.[2,6,7] Complete closure of the round window without any air-trapping reduced not only air conduction but also bone conduction responses.[8] In addition, low pressure added to the inner ear may interfere with the travelling wave and cause sensorineural hearing loss.[9] From animal experiments, the low pressure added to the perilymphatic space made reversible changes in otoacoustic emissions, summating potentials or cochlear microphonic potentials.[10,11]

Therefore, in this case, it seemed that the jugular bulb obliterated the round window niche and might press upon the window during the neck compression. These situations might influence the inner ear and result in the right sensorineural hearing loss and temporary dizziness.

Several authors reported the jugular bulb-related sensorineural hearing loss or dizziness.[12–17] In their reports, these symptoms were ascribed to encroachment on the internal auditory canal, obliteration of the round window or endolymphatic duct obstruction from the jugular bulb. Nevertheless, in this case, the relationship between symptoms and jugular bulb seemed more reliable, because the hearing loss and dizziness accompanied the neck compression. However, during her course, she had never experienced tinnitus, and the reason for this was also unclear. In addition, over pressure in the perilymphatic sac also inhibits mechanical energy and limits movement of the footplate of stapes, so jugular bulb protrusion in the present case might also affect air conduction to some degree.[5,8,18]

The present case was recently diagnosed as Epstein's syndrome which is characterized by hereditary thrombocytopenia, nephritis and bilateral high tone deafness.[19,20] In this patient, bilateral hearing loss in high frequencies were thought to be due to Epstein's syndrome. Some reports mention a relationship between a systemic bony or cartilaginous disorder and formation of a high jugular bulb.[12,21] However, Epstein's syndrome itself would not have influenced a formation of the high jugular bulb in this case.

Conclusion

From this experience, the high jugular bulb could be the possible cause of the sensorineural hearing loss or dizzy disorder. Neck compression is significant and is a necessary test for estimating jugular vein and jugular bulb-related ear disorders.[2–6]

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