

Endolymphatic Sac Tumor Followed as Meniere's Disease

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Case Description

A 11-year-old girl presented to hospital and she complained recurrent vertigo with left hearing loss and tinnitus. Vertigo continued from 30 minutes to a few hours. Pure tone audiogram revealed elevation of left low tone threshold (Figure 1A) and head magnetic resonance imaging revealed no abnormal findings. Then I diagnosed her left Meniere's disease and prescribed isosorbide and betahistine mesilate. Pure tone audiogram recovered to within normal limit and she did not have vertigo. Two years later, she presented to hospital again, because she complained left facial palsy and left hearing loss. Her left tympanic membrane was bluish. Pure tone audiogram revealed left moderate conductive hearing loss that ranged to all frequencies (Figure 1B). Computed Tomography (CT) of tem-

poral bone revealed bone destroying mass shadow that ranged from left jugular foramen to left mastoid cells and tympanic cavity (Figure 2A, 2B). Then she was referred to another University Hospital. Although she did not have family history and was not detected von Hippel-Lindau gene, she had pheochromocytoma at adrenal and adrenalectomy was performed. Joint surgery with neurosurgeons and otolaryngologists was performed 1 month later. Pathological diagnosis was papilloma. In 1984, Hassard et al. reported the first case of adenoma of the endolymphatic sac [1]. According to Butman et al. [2], findings in 35 ELST patients were the following symptoms: sensorineural hearing loss (SNHL) (89%), tinnitus (71%), and vertigo (69%). All patients developed audiovestibular symptoms. Nineteen patients (54%) had a triad of the most common symp-

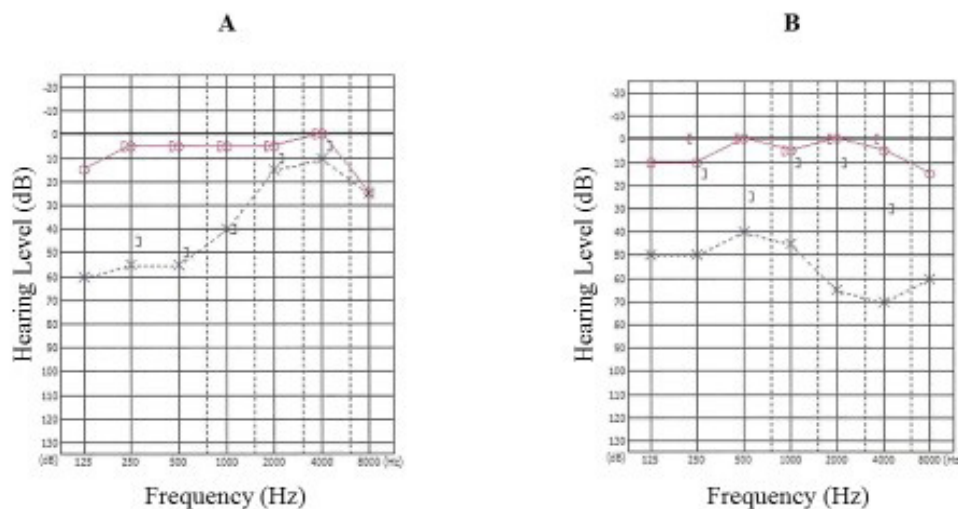


Figure 1

Figure 1: A, Pure tone audiogram performed during the initial visit revealed elevation of left low tone threshold. B, Pure tone audiogram revealed left moderate conductive hearing loss that ranged to all frequencies.

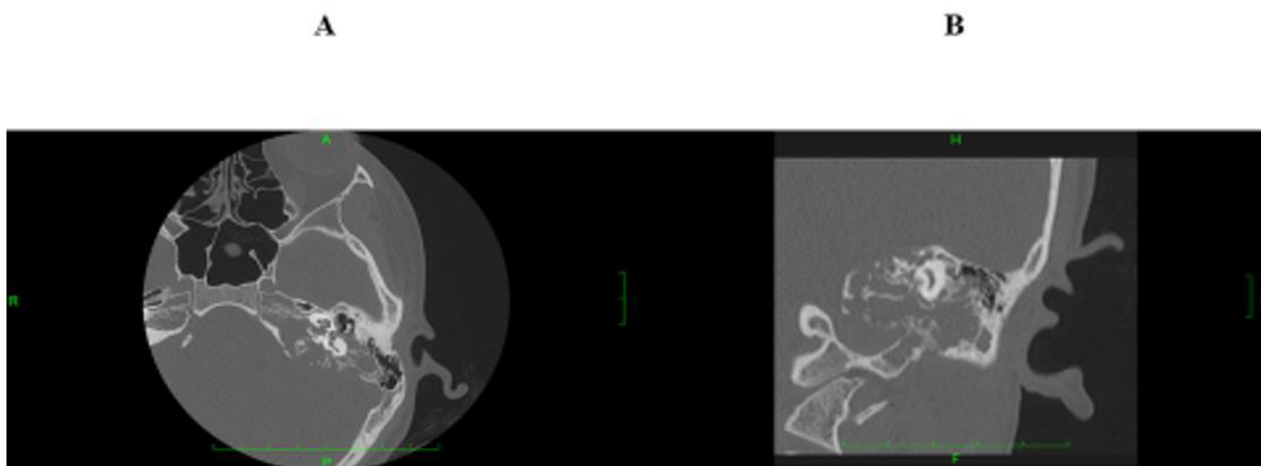


Figure 2: Horizontal (A) and axial (B) CT of temporal bone revealed bone destroying mass shadow that ranged from left jugular foramen to left mastoid cells and tympanic cavity.

toms that included SNHL, tinnitus, and vertigo. As the characteristics of ELST, there are 1) female is dominant; 2) Although chief complaint is unilateral HL, sometimes involve facial nerve palsy; 3) Although invasion of temporal bone is strong, there is no distant metastasis; 4) histologically diagnosed as papilloma. This case satisfied all above characteristics.

Conflicts of Interest and Source of Funding

There was no financial and material support for this study. The author discloses no conflicts of interest.

References

1. Hassard AD, Boudreau SF, Cron CC. Adenoma of the endolymphatic sac. *J Otolaryngol*, 1984; 13: 213-216.
2. Butman JA, Kim HJ, Baggenstos M, et al. Mechanisms of morbid hearing loss associated with tumors of the endolymphatic sac in von Hippel-Lindau disease. *JAMA*, 2007; 298: 41-48.