

CASE REPORT

A Case of Intralabyrinthine Schwannoma with Extension into the Tympanic Cavity

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Objective: To report a rare case of intralabyrinthine schwannoma extending to the tympanic cavity and undergoing surgical management.

Case report: A 56-year-old man presented with left deafness, who had been treated since 1990 for left otitis media and hearing loss with tympanic perforations. In 2005, he was suspected of neurogenic tumor in a histopathologic examination of granulomatous tissues on the tympanic perforations and referred to us for diagnosis and management. On presentation, the patient was deaf in the left ear and had left tympanic perforation with yellowish-white tumor lesion in the left perforated area. The tumor had an apparent origin in the cochlear to vestibule, which was separate from the facial nerve, destroyed the promontory bone, and protruded into the tympanic cavity. The patient underwent a partial resection of the tumor which extended to the tympanic cavity and closure of tympanic perforation. Histopathologic examination of the tumor was consistent with schwannoma. A subsequent follow-up has not shown increase of the tumor.

Conclusions: Reports of intralabyrinthine schwannoma have increased in recent years. The disease is uncommon, and the case reported herein is extremely rare with extension of the tumor to the tympanic cavity. Indication for the surgical treatment of intralabyrinthine schwannoma should be determined in terms of postoperative complications.

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Introduction

Intralabyrinthine schwannomas are rare tumors. Refinement of MRI imaging technique^[1] has provided precise imaging of the inner ear for diagnosis of labyrinthine lesions, and reports of intralabyrinthine schwannoma have increased in recent years^[2-8]. We report an unusual case of intralabyrinthine schwannoma which extended from the inner ear to the middle ear cavity and discuss management of intralabyrinthine schwannoma.

Case Report

A 56-year-old man presented with complaints of hearing loss and otorrhea of the left ear. Since 1990, he had received treatment by a local otorhinolaryngologist for chronic otitis media and tinnitus of the left ear and was referred to us in June 2005 because of suspicious neurogenic tumor in histopathologic examination of biopsied granulomatous tissue in pyramidal perforation. He had long been deaf in the left ear, but did not suffer from facial palsy.

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Computed tomography did not reveal osseous defect but soft tissue lesions in the labyrinth (Fig. 1a, 1b). Gadolinium-enhanced T1-weighted images in MRI showed enhancing pathologies not in the internal auditory canal but in the cochlea, vestibule, and middle inferior tympanic cavity (Fig. 2a). Gadolinium-enhanced T2-weighted images did not reveal pathology in the inner auditory canal but depict defects in the cochlea and vestibule and high signal areas in the middle and inferior tympanic cavity (Fig. 2b). These findings led to suspicious neurogenic tumor extending from the inner ear to the middle ear cavity. We performed a surgical intervention of the lesions on August 2005 to perform histopathologic examination of tissues, to elucidate the relation of the tumors with the facial nerve, and to treat the tympanic perforation.

Surgical findings. The tympanic cavity was released by retroauricular incisions and the tumor lesions were removed. The tumors appeared dark red, soft, and smooth in surface, and filled the middle tympanic cavity. A possibly large portion of the tumors, and granulation tissues in the anterior to inferior tympanic cavity were removed. The incus and stapes were resected. There was evidently no continuity between

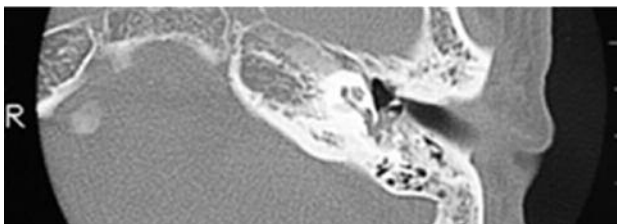


Figure 1a. Axial CT, illustrating not bony destruction but soft tissue pathologies in the posterior portion of the middle labyrinth.

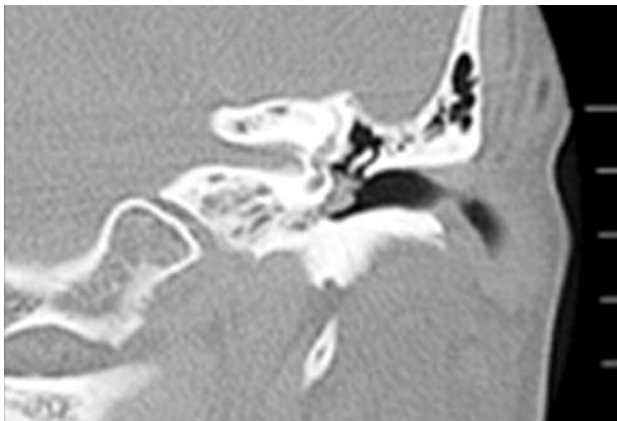


Figure 1b. Coronal CT, illustrating soft tissue lesions without osseous destruction in the posterior portion of the middle ear.

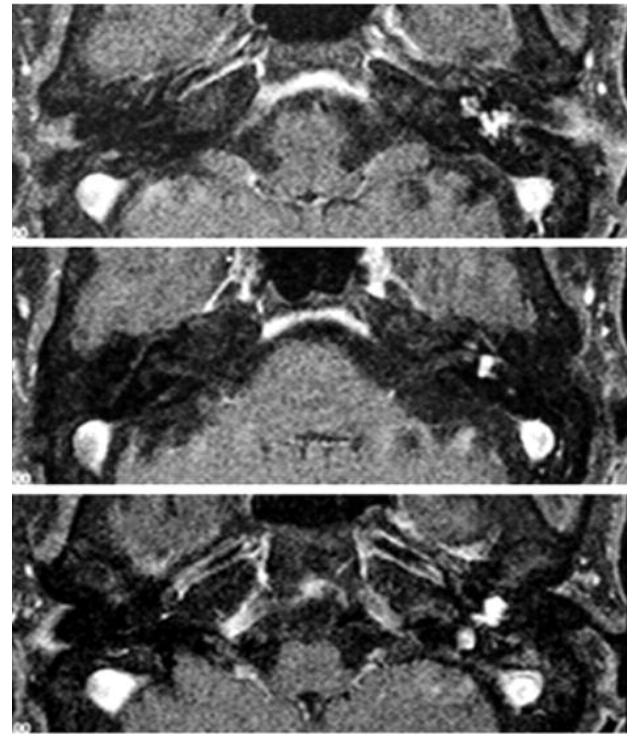


Figure 2a. Preoperative, Gadolinium-enhanced T1-weighted MRI (Axial) imaging, showing enhancing pathologies in the left cochlea, vestibule, and middle and inferior tympanic cavity.

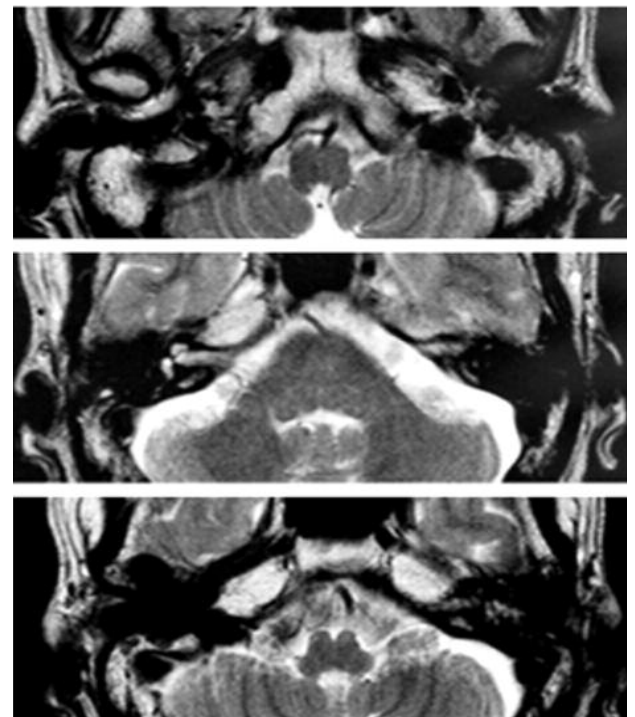


Figure 2b. Preoperative, Gadolinium-enhanced T2-weighted MRI (axial) imaging, showing absence of high intensity area in the left cochlea and vestibule, and preserved high intensity area in the middle and inferior tympanic cavity.

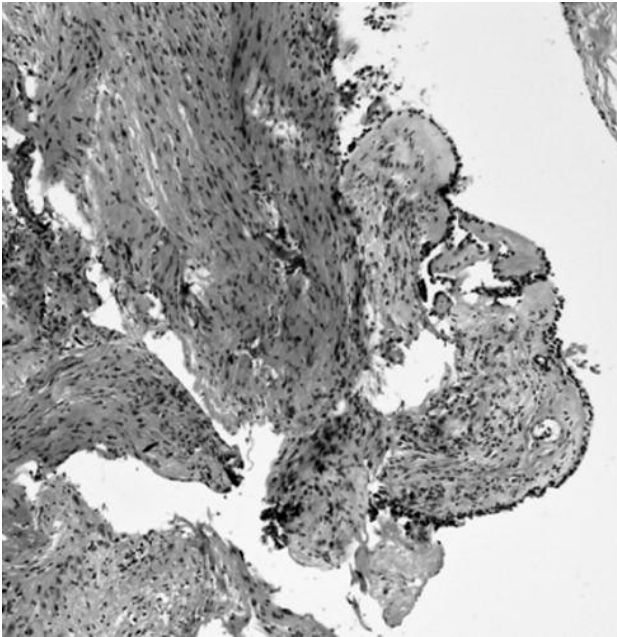


Figure 3a. Histopathologic examination with hematoxylin and eosin staining(100), illustrating proliferations of palisaded spindle cells beneath middle ear membranous tissues.

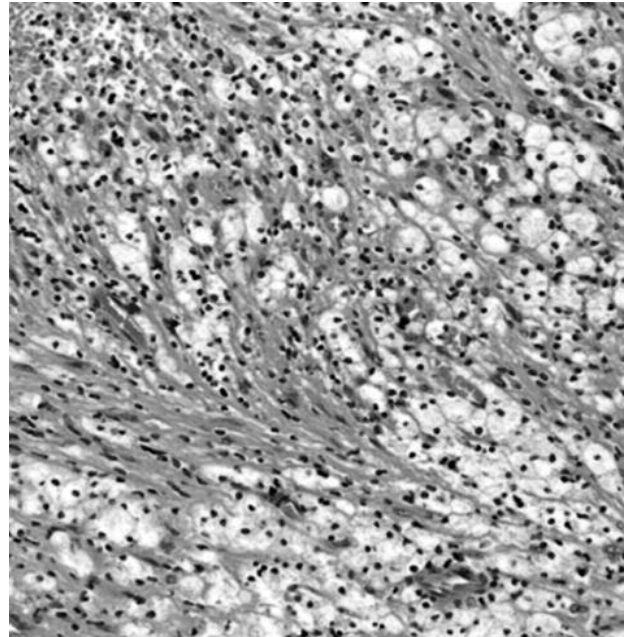


Figure 3b. Histopathologic examination with hematoxylin and eosin staining(200), illustrating infiltrations of foamy histiocytic cells in a cellular tissues.

the tumor and facial nerve. Osseous defects were present in the tympanic sinus and inferior tympanic cavity, and the tumor lesions in the inner ear evidently extended to the middle ear cavity.

Histopathologic findings. In areas beneath membranous tissues in the middle ear there were proliferations of palisaded spindle cells (Fig. 3a) and there were acellular areas infiltrated by foamy histiocytic cells. The

findings defined a histopathologic diagnosis of schwannoma.

Postoperative course. The tympanic membrane perforations were successfully closed, and the otorrhea was ceased. There were no complications including facial palsy. In a five-year follow-up study, there is no evidence for proliferation or extension of tumor mass into the internal auditory canal as evaluated by MRI images (Fig. 4a, 4b).

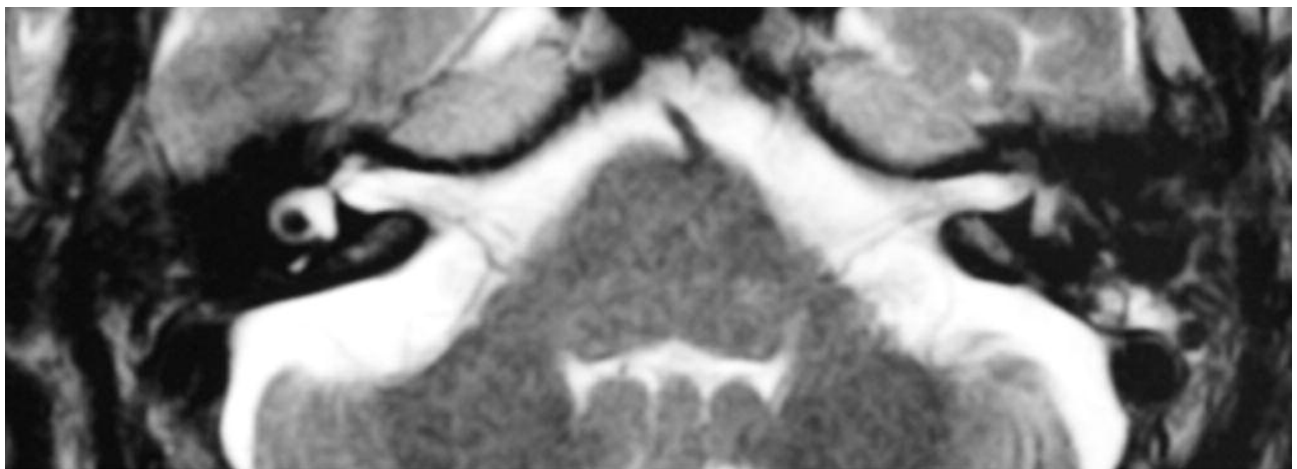


Figure 4a. Gadolinium-enhanced T2-weighted MRI imaging, 4 months after surgery, showing defect of high intensity area in the cochlea and presence of relative high intensity area in the vestibule.

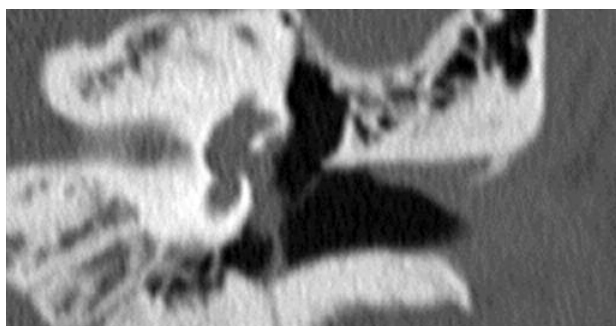


Figure 4b. Coronal CT image, 4.9 months after surgery, illustrating presence of soft tissue pathologies in the middle tympanic cavity.

Discussion

Schwannoma is benign tumor that has an origin in the myelin sheath of peripheral nerve. Intralabyrinthine schwannoma is schwannoma in the labyrinth originating from peripheral sensory nerve in the vestibule or cochlear nerve^[2, 9, 10].

Mayer reported in 1917 the first case of histopathologically proven schwannoma^[11]. In recent years, development of sophisticated MRI technology allowed to examine details of the inner ear lesions^[1], which facilitated detection of the tumor and increased reports of intralabyrinthine schwannomas^[2-7]. Meniere disease-like symptom is most frequent presenting symptom in cases of intralabyrinthine schwannoma^[2, 12-14].

Kennedy and associates^[4] outlined the diagnosis and management of intralabyrinthine schwannomas and proposed a new classification system in terms of the site of lesions. According to the classification system, the case described herein is classified into the tympanolabyrinthine class. To the best of our knowledge, only a few cases of tympanolabyrinthine class schwannoma were reported, all more than 20 years previously^[14, 15].

The case presented above developed a slow growth of intralabyrinthine schwannoma that did not confined to the bony labyrinth but extended into the middle ear cavity. The schwannoma of this case was found on presentation to fill the vestibule and cochlea, and since it already extended widely to the middle ear cavity, the original site of the tumor in the inner ear remains unknown.

Surgical intervention for intralabyrinthine schwannomas has been indicated in cases with untreatable severe hearing loss or tumor extension of the tumor mass into the cerebellopontine angle^[4-6]. This case was deaf at the time of presentation and had no vertigo or facial palsy. Preoperative biopsies were consistent with neurogenic tumor, but the information was insufficient and definite histopathologic diagnosis required more detailed studies. The anatomic relation of the tumor with the facial nerve was also necessary to determine the site of tumor origin. Furthermore, the patient was irritated by frequent annoying otorrhea due to tympanic perforations associated chronic otitis media. To overcome these difficulties surgical management was selected as a choice of treatment, in which a partial rather than total removal of tumors was performed to reduce the risk of inadvertent facial nerve damage. Postoperatively, the patient's otorrhea was resolved, and the primary site of the tumor was verified and its histopathologic diagnosis was established. Fortunately, extension of the tumor to the internal auditory canal has not occurred up to the present probably because the surgical intervention reduced the tumor mass.

If a total removal of the present tumor was aimed for the surgical intervention, the patient would have been informed of, and given consent to, the postoperative risk of facial palsy, cerebrospinal fluid rhinorrhea, and meningitis. Keeping these disadvantages in mind, a partial rather than total removal of schwannoma to reduce tumor size in the middle ear cavity may be an appropriate management of choice for the tympanolabyrinthine class of intralabyrinthine schwannomas. This may provide a better quality of life in patients with intralabyrinthine schwannomas. It is of course mandatory to perform a regular and long follow-up imaging assessment of the residual tumor lesion.

Conclusion

We reported a case of intralabyrinthine schwannomas with extension to the middle ear cavity, and discussed the management of choice for intralabyrinthine schwannomas in an individual patient's basis.

Acknowledgements

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