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Large middle ear schwannoma of the Jacobson's nerve with intracranial extension

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ABSTRACT

The patient is a 64-year-old woman who developed a sensation of right ear fullness and hearing loss in early November 2010. Physical examination revealed a painless reddish granular lesion filling in the right external auditory canal. Her right ear was deaf, and no facial palsy was noted. Computed tomography, magnetic resonance imaging and positron emission tomography revealed a middle ear mass extending to the external auditory canal with intracranial invasion causing temporal lobe retraction and inferiorly extending just anterior to the jugular bulb as well. A combination of transmastoid and middle cranial fossa approach along with anterior rerouting of the facial nerve was employed for a near-total removal of the tumor. Based upon the operative findings, it was deemed that the tumor could have arisen from the Jacobson's nerve.

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1. Introduction

Schwannomas are common benign tumors of the temporal bone, arising mostly from the vestibulocochlear nerve in the cerebellopontine angle or the internal auditory canal in the head and neck region. These tumors arise from the Schwann cells which surround the myelin sheath of all peripheral nerves. They can arise from any peripheral nerve but schwannomas of the middle ear are relatively rare and the most commonly reported cases are facial nerve neuromas. It is extremely rare for it to occur from the tympanic branch of the glossopharyngeal nerve (Jacobson's nerve).

Clinical management of the tumors in the tympanic cavity should be considered in accordance with its pathology, size, location along with facial function, hearing acuity and equilibrium function, since many functionally important structures exist. When it has a benign nature, functional preservation must be the first priority to consider. Thus 'wait and scan' policy could be the first to be considered in cases such as facial nerve neuroma, especially when not accompanied by facial nerve palsy. But when it is large enough to cause additional symptoms, as in our case, surgical manipulation should be undertaken considering possible functional preservation. One such case we encountered was a giant intratympanic schwannoma extending into the intracranial cavity and external auditory canal with deaf ear but normal facial function. In this case, the treatment goal was total removal of the tumor with preserving facial function. Surgical procedure was undertaken for this case and suspected tumor origin was discussed based upon the findings of the surgery along with a pertinent paper review.

2. Case report

A 64-year-old woman complained of a sensation of right ear fullness and hearing loss in early November 2010. She had visited a nearby hospital and had been diagnosed as having right ear cerumen impaction; however, the symptoms persisted. At the end of the same month, she came to our hospital and her physical examination revealed a painless reddish granular lesion in the right external auditory canal (Fig. 1A), and the left ear was normal. She had no dizziness nor facial nerve paralysis. However, she had a history of temporary right facial weakness in 2005. A biopsy from the external auditory canal mass was performed for histopathological examination which revealed interlacing bundles of elongated spindle-shaped cells with palisading of the nuclei in a collagenous matrix (Antoni type A) and areas of sparsely arranged spindle cells with occasional cyst formation (Antoni type B)

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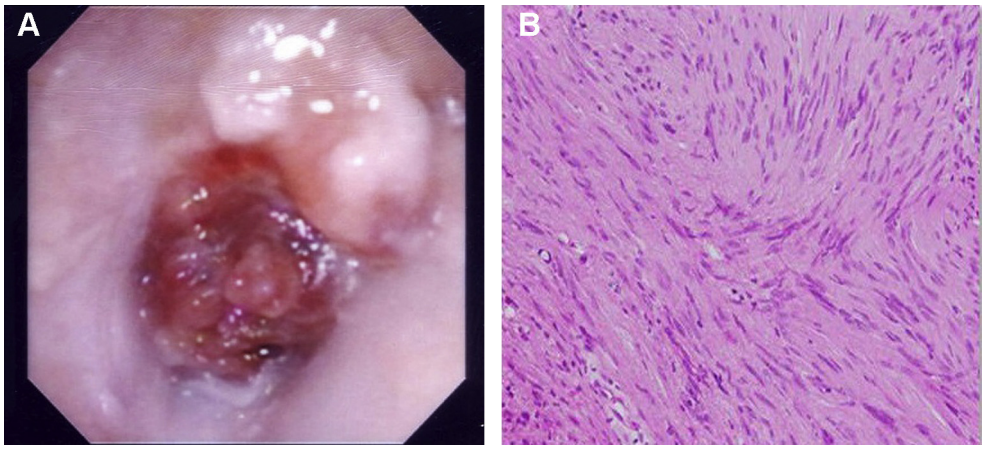

Fig. 1. Otoscopic view of the right ear and histopathological examination. (A) A reddish granular mass had perforated through the posterior meatal wall. (B) Interlacing bundles of elongated spindle-shaped cells with palisading of the nuclei in a collagenous matrix (Antoni type A) and areas of sparsely arranged spindle cells with occasional cyst formation (Antoni type B).

(Fig. 1B). From this histopathological findings, schwannoma was diagnosed. An audiogram showed normal hearing in the left and deafness in the right ear (Fig. 2). The radiological evaluation, including computed tomography scanning (CT), magnetic resonance imaging (MRI) and positron emission tomography (PET) scanning, was performed. The computed tomography demonstrated a well-circumscribed soft tissue mass filling the middle ear space and extending to the external auditory canal with intracranial extension through the tegmen tympani causing temporal lobe retraction and extending also toward the jugular foramen (Fig. 3A). There was also erosion of the ossicular chain except for malleus head and short crus of incus, floor of the middle ear, horizontal facial canal, basal turn of the cochlea and vestibule with widening of the jugular foramen (Fig. 3B). Magnetic resonance imaging showed hypointense dumbbell-shaped mass reaching the vicinity of the jugular foramen and pushing the temporal lobe with heterogenous gadolinium enhancement (Fig. 3C). Magnetic resonance angiography did not show any remarkable changes. PET scanning showed high uptake at the region of the jugular foramen (SUV = 7.9) while the upper part of the lesion showed low uptake (SUV = 3.7)(Fig. 4 A and B).

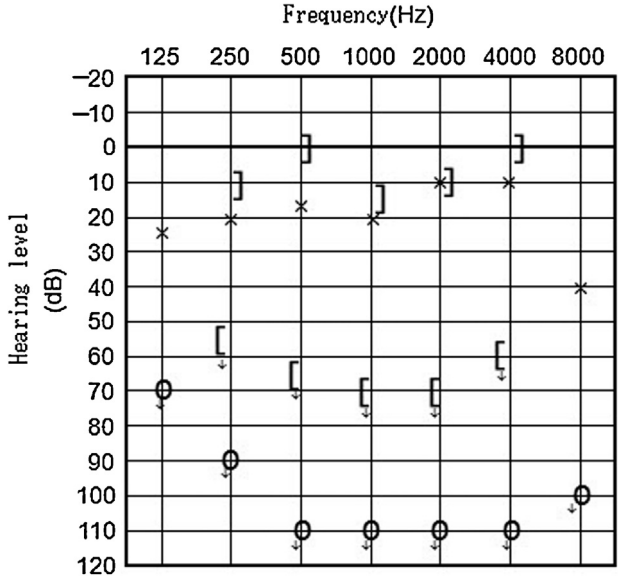

Fig. 2. An audiogram showing normal hearing in the left ear and the deaf right ear.

Schirmer's test and electrogustometry were normal [right −2 dB, left 4 dB]. Based upon these findings, a combined approach of middle cranial fossa and trans-mastoid with anterior rerouting of the facial nerve was employed. At first, mastoidectomy was performed to expose the tumor and to confirm the facial canal. The tumor occupied from aditus ad antrum to around the facial genu, which was removed and re-confirmed as schwannoma by frozen section. When carefully elevating the tumor from the facial canal, it became clear that the tumor had no communication with the facial nerve. To give access to the antero-inferior portion of the tumor, anterior rerouting of the facial nerve was performed by drilling out the surrounding bony tissue along the vertical portion of the nerve (Fig. 4C) and the tumor was removed piece by piece. Near the jugular bulb, some parts of the tumor were strongly adhered to the vein; so a small part of the tumor was left behind as shown by postoperative gadolinium enhanced MRI (Fig. 3D). Then tumor removal was performed from middle cranial fossa. Most of the tumor capsule was elevated from the dura mater; however at the most anteromedial part of the tumor, severe adhesions were noted, and because of that, a small perforation of the dura was caused. This was closed by suturing the dura and sealing with Neovail sheet (Gunze limited medical division, Kyoto, Japan) and fibrin glue, then covering by fat. The auditory tube was closed by a piece of muscle; the large defect was filled with abdominal fat, and finally, the wound was closed. As for the nerve origin, it was considered Jacobson's nerve because the facial nerve was easily separable from the tumor and the route of the tumor progression. The patient developed slight facial weakness just after the surgery which improved gradually and eventually reached House–Brackmann grade I (Fig. 4D). Three years have passed since the initial surgery. It has recently been found that the residual tumor at the jugular foramen has an increasing tendency to slightly extend into the cerebellopontine angle. Accordingly, radiotherapy (X-knife) is now being scheduled.

3. Discussion

A mass behind an intact tympanic membrane could be epithelial or mesenchymal in origin, benign or malignant. Tumors include cholesteatoma, neuroma or schwannoma, paraganglioma, carcinoma (squamous cell), osteoma or osteosarcoma, adenoma or adenocarcinoma, lymphoma, chondroma or chondrosarcoma, and rhabdomyosarcoma [1].

Middle ear schwannomas may originate from the nerves of the tympanic cavity or by the extension from outside. Schwannomas

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