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Case Report

A Case of Vestibular Schwannoma with Deafness Showing Remarkable Hearing Recovery Following Hearing-Preserving Surgery

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Surgery for vestibular schwannoma can be divided into hearing-preserving and nonhearing-preserving surgeries. Hearing-preserving surgery is usually not considered in patients with deafness due to vestibular schwannoma, because hearing is unlikely to improve, and surgery aims to maximize the tumor resection at the expense of hearing. We report an extremely rare case of a 46-year-old man with unilateral profound hearing loss due to a vestibular schwannoma with marked cystic degeneration in the left cistern, which significantly recovered to near-normal hearing levels after hearing-preserving surgery. Hearing loss gradually worsened, and preoperative pure-tone evaluation showed complete hearing loss in the left ear. However, the response to the distortion product otoacoustic emission was preserved, and hearing loss was considered to be retrocochlear. Tumor resection was performed using the retrolabyrinthine approach with continuous monitoring using dorsal cochlear nucleus action potential, auditory brainstem response, and facial nerve function muscle action potential. The cistern portion of the tumor was almost completely resected along with the wall. Postoperatively, the pure-tone threshold on the left side markedly improved. The present case clearly demonstrates the possibility of hearing recovery in patients with retrocochlear hearing loss. We should consider expanding the indications for hearing-preserving surgery.

KEYWORDS: Vestibular schwannoma, retrocochlear hearing loss, hearing preservation surgery, deafness, auditory brainstem response, dorsal cochlear nucleus action potential

INTRODUCTION

Vestibular schwannomas account for approximately 80% of cerebellopontine-angle tumors. In a recent study, the prevalence of vestibular schwannoma was 42.0 per 100 000 people.[1] Most vestibular schwannomas originate from the inferior vestibular nerve and are often discovered during magnetic resonance imaging (MRI) for a thorough evaluation of asymmetric sensorineural hearing loss.

The early symptoms include deafness, tinnitus, and dizziness. The risk of loss of useful hearing in vestibular schwannomas is reported to be 43% using the 70/30 rule, which is defined as a speech discrimination score ≥70% combined with a pure-tone average ≤30 dB.[2] Sensorineural hearing loss significantly affects patients' quality of life[3]; however, the treatment has not been established. Most cases of sudden hearing loss due to vestibular schwannoma are treated symptomatically with steroids,[4] with no established surgical treatment. Progression of vestibular schwannomas can cause symptoms such as sensory disturbances in the trigeminal nerve distribution area, cerebellar ataxia due to compression of the brainstem, and sensorineural hearing loss.

Currently, the 3 major treatment options for patients with vestibular schwannoma are surgery, radiation therapy, and observation.[5,6] Management decisions are based on the tumor size, signs and symptoms, patient age, comorbidities, and patient preferences.[7]

We recently encountered an extremely rare case in which unilateral profound hearing loss due to vestibular schwannoma significantly recovered to a near-normal hearing level after hearing-preserving surgery. Herein, we report the course of this case in detail.

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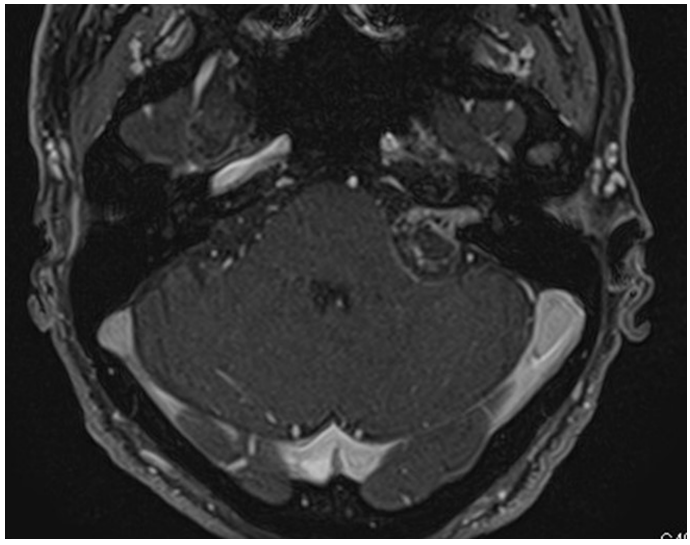

Figure 1. Preoperative magnetic resonance image. A 16-mm-sized vestibular schwannoma with marked cystic degeneration is observed in the left cistern. The tumor obscures the internal ear canal and contacts and presses the brainstem.

CASE PRESENTATION

A 46-year-old man with sudden left-sided hearing loss and tinnitus ear received steroid pulse therapy. Although his deafness improved, tinnitus persisted. Five years later, tinnitus worsened and dizziness occurred.

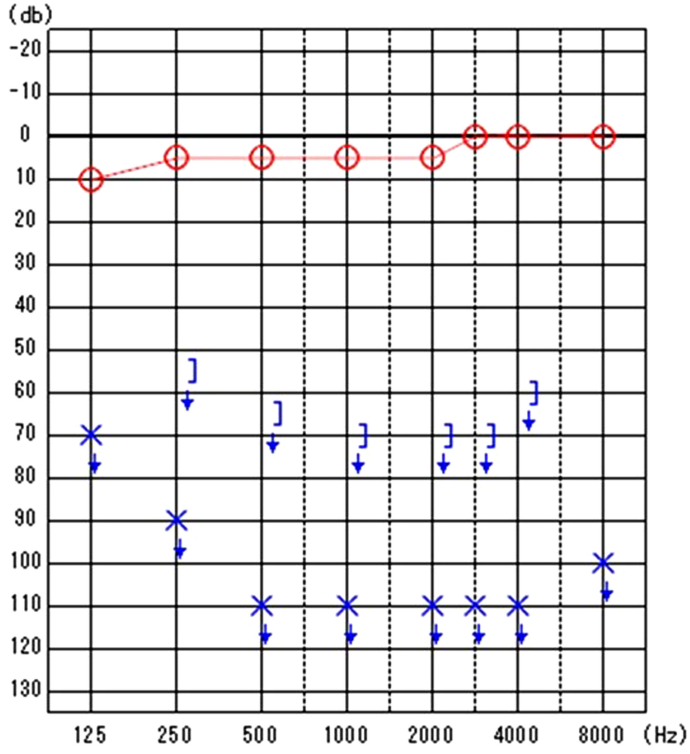

Figure 3. Pure-tone audiogram 5 days before the intervention. Pure-tone audiometry 5 days before surgery reveals deafness on the left side.

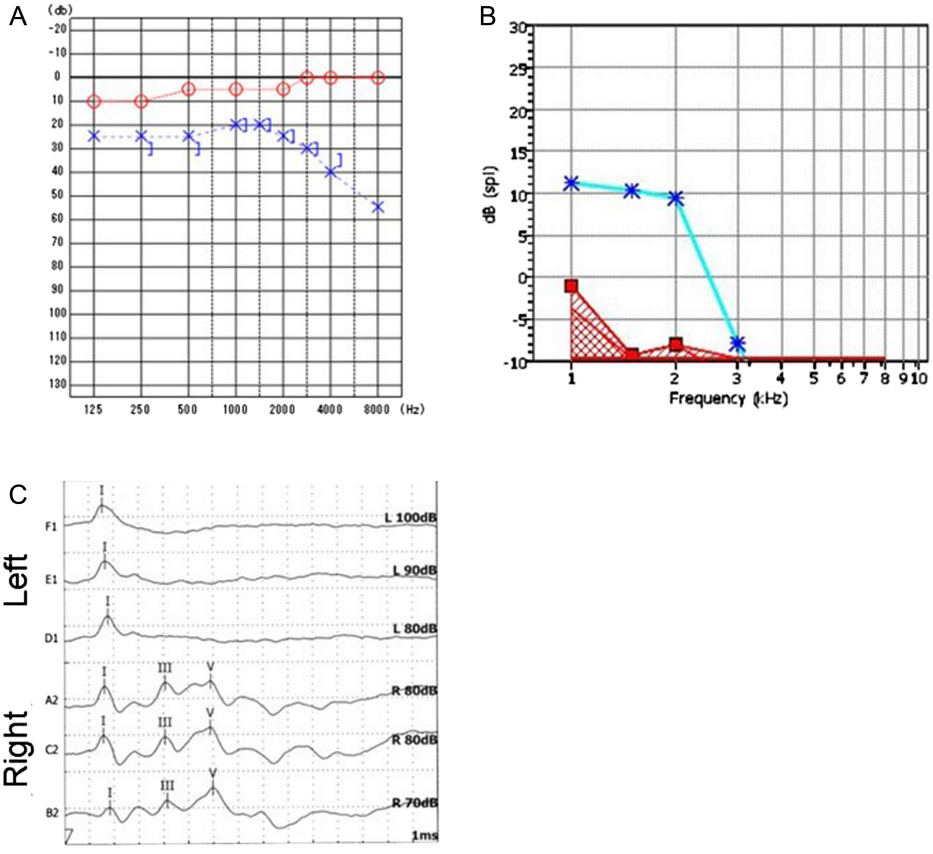

Figure 2. Preoperative audiological evaluation. (A) Pure-tone audiometry: 5.0 dBHL on the right side and 22.5 dBHL on the left side 3 months before surgery. (B) The distortion product otoacoustic emission (DPOAE) shows a relatively high response, with a signal-to-noise ratio of ≥20. (C) A clear wave I is observed in the auditory brainstem response (ABR); however, the other waves are not observed.

Magnetic resonance imaging revealed a 16-mm vestibular schwannoma with marked cystic degeneration in the left cistern (Figure 1). The patient was referred to our hospital for further treatment. On the first visit, pure-tone audiometry scores on the right and left sides were 5.0 dBHL and 22.5 dBHL, respectively. (Figure 2A) Accordingly, the patient was diagnosed with left-sided sensorineural hearing loss.

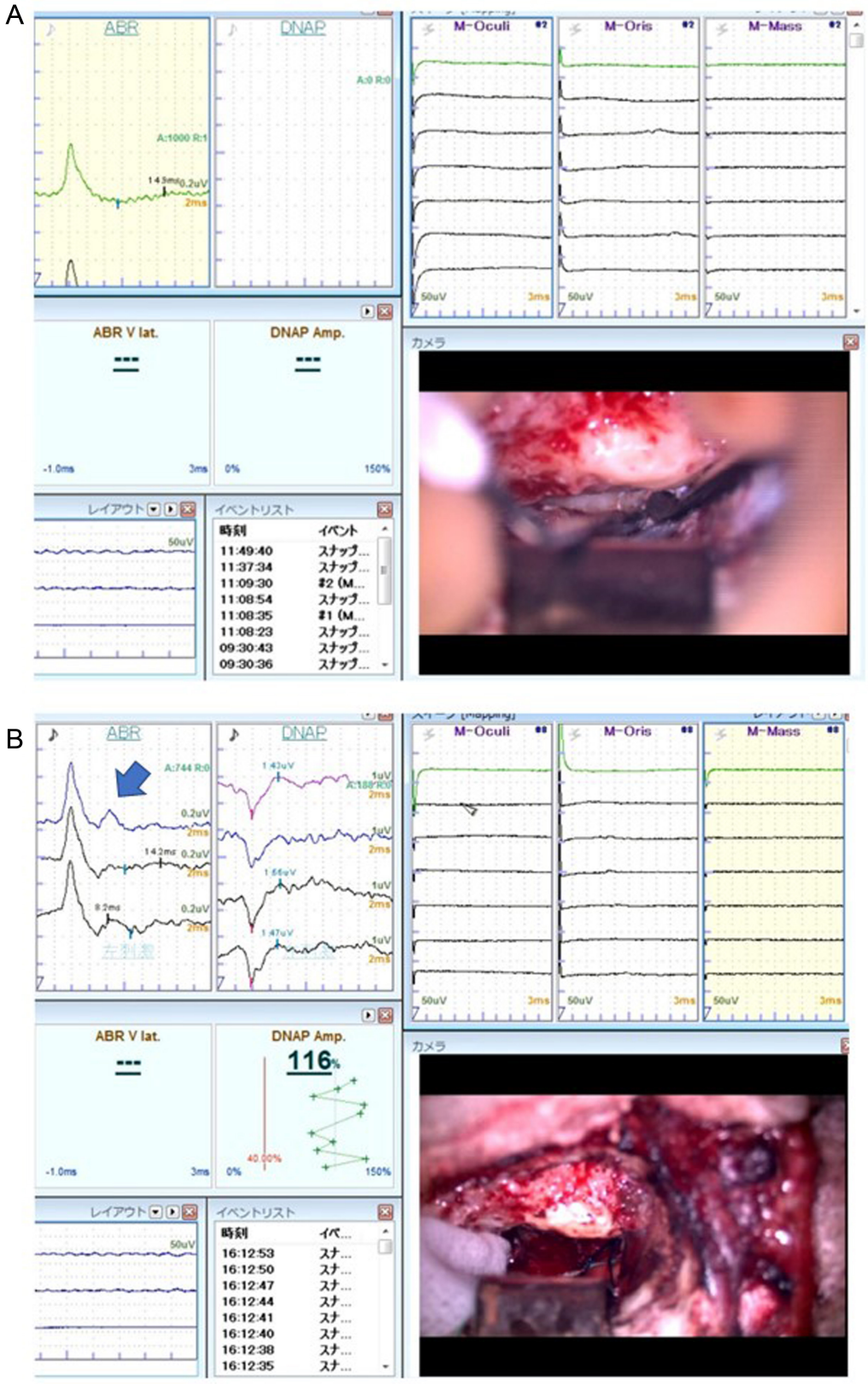

Figure 4. Intraoperative monitoring findings. (A, B) Only wave I is observed in the ABR before tumor resection. During the removal of the tumor in the cistern, wave III is clearly observed (arrow in B), and at the end of the surgery, a slight wave V is also observed (B).

The signal-to-noise ratio on distortion product otoacoustic emission (DPOAE) was ≥20, indicating good inner-ear function. Although a clear wave I was observed in the auditory brainstem response (ABR), waves II–V disappeared (Figure 2B and C). Hearing-preserving surgery was planned 3 months later; however, the hearing gradually deteriorated and the patient became deaf 5 days before the operation (Figure 3). As no change was observed in DPOAE, deafness was considered to be due to retrocochlear hearing loss. Therefore, we decided to perform a hearing-preserving surgery via the retrolabyrinthine approach with the hope of hearing improvement. Surgery was performed with continuous monitoring using the dorsal cochlear nucleus action potential (DNAP), ABR, and facial nerve function muscle action potential.[8]

The tumor was located in contact with the brainstem. Before removing the tumor, only wave I was observed in the ABR (Figure 4A), whereas wave III was clearly observed during the removal of the cystic part of the tumor in the cistern. We decided not to remove the tumor from the internal auditory canal to maximize the possibility of preserving inner-ear function. At the end of the surgery, a slight wave V was observed (Figure 4B). Dorsal cochlear nucleus action potential monitoring also showed a more apparent wave compared to that before tumor removal. The amplitude was maintained at almost 100%, even after tumor removal.

The pathological diagnosis was a schwannoma. On postoperative day 9, a marked improvement in the pure-tone threshold to 21.3 dBHL was observed on the left side (Figure 5A). A tendency for further improvement was observed at 4 weeks postoperatively. Five months after surgery, wave V was observed on ABR, and the DPOAE response was maintained (Figure 5B and C). This study was approved by Ethics Committee of KEIO University (Approval No: 20200033, Date: April 28, 2020). Informed consent for publication was obtained from the patient.

DISCUSSION

Herein, we described a rare case of vestibular schwannoma with significant improvement in hearing after surgical intervention. Most vestibular schwannomas are slow growing, and treatment is associated with a risk of complications, including facial paralysis, hearing loss, and cerebrospinal fluid leakage. Therefore observation is preferred for relatively minor tumors or tumors in elderly patients.[9]

Surgery or radiation is considered for patients with large vestibular schwannomas that have reached the brainstem and patients with small tumors but progressive hearing loss. Surgery for vestibular schwannomas can be divided into hearing-preserving and nonhearing-preserving surgeries. Hearing-preserving surgery is performed when hearing preservation can be expected, for example, in cases of mild hearing loss. Nonhearing-preserving surgery is the procedure of choice when hearing preservation cannot be expected, for example, in patients with deafness or severe hearing loss, where hearing is unlikely to improve, and surgery aims to maximize the resection

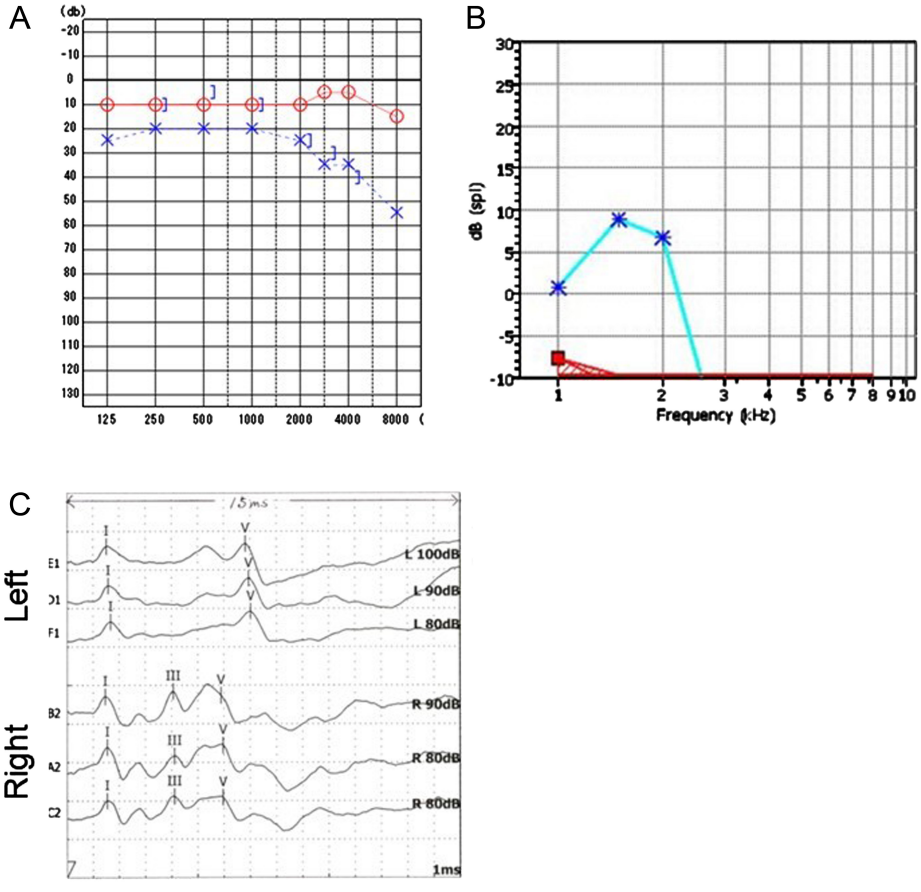

Figure 5. Postoperative audiological evaluation. (A) A marked improvement in the pure-tone threshold to 21.3 dBHL is observed on the left side. (B) A good response to DPOAE is maintained. (C) Waves III and V are observed on the ABR 5 months after surgery.

of the tumor at the expense of residual hearing. Moreover, the best hearing level after surgery for vestibular schwannoma is the preoperative hearing level.[10] The average hearing maintenance rate is approximately 50%-60%, and postoperative hearing rarely recovers to a level more than that before surgery.[11] We performed hearing-preserving surgery on a patient who was deaf in the affected ear because the DPOAE response was well maintained, which suggested that the inner-ear function was well preserved.

Hearing restoration has been reported in cases with good preoperative otoacoustic emission, a history of sudden deafness, surgery within 6 months of worsening deafness, and tumor cysts.[12] In our patient, the prognostic factors for recovery of hearing included surgery within 6 months of worsening hearing loss, presence of wave I on ABR, good otoacoustic emission, and cystic degeneration of the tumor.

We emphasize the importance of advanced auditory monitoring using ABR and DNAP. Reinforced intraoperative hearing monitoring allows for more accurate cochlear nerve preservation.[13,14] In this case, after the tumor was decompressed and electrical signals could be transmitted to the auditory cortex, waves III and V were observed intraoperatively.

The importance of hearing-preserving surgery has been underestimated in patients with "nonserviceable" hearing levels, despite the fact that unilateral hearing loss leads to reduced quality of life, including a loss of sense of direction and difficulty in hearing in noisy conditions. Additionally, tinnitus, which can cause depression, may occur with unilateral deafness.[15] Hearing aids are not expected to be effective in deaf patients, and therapeutic interventions are more complex than in those with residual hearing levels. The translabyrinthine approach may be performed even in patients with residual hearing. The present case shows that high-precision hearing monitoring can improve hearing in patients with severe retrocochlear hearing loss by preserving the cochlear function. Accumulation of more cases of hearing improvement in vestibular schwannomas will help to clarify the specific factors that might maintain hearing.

CONCLUSION

The findings in this case clearly demonstrated the possibility of hearing recovery following hearing-preserving surgery with reinforced monitoring, at least in selected cases, and broaden the indications for hearing-preserving surgery. Based on our findings, hearing-preserving surgeries should be performed even in patients with deafness, provided that deafness is caused by pure posterior labyrinthine hearing loss. The present case highlighted the importance of a hybrid surgical approach, in which surgery was started with the aim of hearing preservation. If needed, surgery could have been changed to non-hearing-preserving surgery. Starting surgery via the retrolabyrinthine approach and converting it into a translabyrinthine surgery if the ABR/DNAP waveform is not observed is a possible approach for such cases.

Ethics Committee Approval: This study was approved by Ethics Committee of KEIO University (Approval No: 20200033, Date: April 28, 2020).

Informed Consent: Written informed consent was obtained from the patient who agreed to take part in the study.

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