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Case Report

A Case of McCune–Albright Syndrome with External Auditory Canal Stenosis Treated with Image-Guided Surgery System-Assisted Temporal Bone Surgery

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McCune–Albright syndrome is a disorder of fibrous bone dysplasia complicated by skin pigmentation and endocrine abnormalities. Although temporal bone lesions are rare, surgical treatment is required when external auditory canal (EAC) stenosis develops. However, no consensus has been reached regarding surgical approaches. To safely perform surgery for temporal bone malformations, knowing the exact location of the critical organs in the temporal bone in relation to the bony lesion is important. Otherwise, intraoperative orientation may be lost owing to differences from the normal anatomy. Although image-guided surgery systems (IGSSs) would be useful in surgery for temporal bone malformations, few studies have reported on the use of IGSS in temporal bone surgery. In this report, we describe a case of McCune–Albright syndrome with EAC stenosis that was safely treated using IGSS-assisted temporal bone surgery.

KEYWORDS: Ear surgery, external auditory canal stenosis, fibrous dysplasia, hearing loss, image-guided surgery system, McCune–Albright syndrome, temporal bone

INTRODUCTION

McCune–Albright syndrome is characterized by unilateral multiple fibrous dysplasia (FD) with skin pigmentation and endocrine abnormalities.[1,2] Fibrous dysplasia in the temporal bone region is rare and may require surgical intervention because of the slow progression of external auditory canal (EAC) stenosis and the formation of secondary cholesteatoma.[3] Surgical treatment for temporal bone FD requires careful consideration because it is a benign osteo-proliferative disease in which the lesion extends into vital organs in the temporal bone, such as the facial nerve, semicircular canal, and ossicles. Most importantly, there is a high risk of restenosis even after canalplasty.

Temporal bone surgery involves the outer, middle, and inner ears, and the adjacent skull base, indicating that bony structures occupy most of the target area. Important blood vessels and nerves that must be preserved are intricately located in the temporal bone, and knowing the exact location of these vital organs in relation to the bony lesion is important to safely perform the surgery. However, notably, in cases of temporal bone malformations, which differ from the normal anatomy, even careful preoperative preparation can result in a completely different surgical view and loss of orientation as the surgical operation progresses.[4]

Recent advances in imaging technology have enabled detailed preoperative and intraoperative imaging evaluations of the surgical areas. Computed tomography (CT) is the most useful imaging modality for evaluating temporal bone surgery. Although an image-guided surgery system (IGSS) is useful in cases of malformations that differ from normal anatomy, achieving the accuracy (error of

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less than 1 mm) required for temporal bone surgery is challenging if a conventional IGSS, which places reference markers on the skin or a noninvasive patient tracker, is used. To improve accuracy, it has been reported that CT images are taken with a reference marker fixed to the bone in the surgical area, and registration of the image information enables a highly accurate IGSS.[5,6]

In this paper, we report a case in which canalplasty was safely performed, assisted by an accurate IGSS, for temporal bone FD in a patient with McCune–Albright syndrome. Based on our experience with this case, we believe that IGSS-assisted temporal bone surgery is a safe and useful surgical approach for the treatment of temporal bone FD.

CASE PRESENTATION

A 35-year-old female presented at National Defense Medical College hospital with left-sided hearing loss. She was diagnosed with McCune–Albright syndrome at 1 year of age because of precocious puberty (breast growth) and hyperpigmentation (café-au-lait spots) and was regularly followed up in our pediatric clinic.The patient provided consent for the publication of this case report. She experienced left-sided hearing loss 1 month prior, and the symptoms did not improve despite treatment at a nearby otolaryngology clinic. Therefore, the patient visited department of otolaryngology at National Defense Medical College hospital. She underwent osteosynthesis treatment at the age of 16 years for a fracture of the right femoral neck and bone graft surgery at the age of 21 years for a refracture of the right femoral head. The patient had no relevant family history. Upon otological examination during the first visit to our department, the right EAC and tympanic membrane (TM) were normal. The left EAC was severely narrowed in the bony region, epithelial debris accumulated deeper than the narrowed lesion, and the TM was not observed (Figure 1A). Pure-tone audiometry revealed a mean hearing threshold of 18.8 and 72.5 dB in the right and left ears, respectively, with mixed hearing loss (Figure 1B). A CT scan of the temporal bone (0.25 mm slice) showed circumferential proliferation of the bony part of the left EAC, resulting in EAC stenosis. The deep part of the EAC was filled with soft shadows in contact with the TM. No pathology was noted in the middle ear cavity, and no abnormal findings were observed in the shape of the ossicles or the course of the facial nerve (Figure 1C and Supplementary Figure 1). The internal auditory canal (IAC) length was longer on the diseased side than on the healthy side (12.07 mm > 9.50 mm, Supplementary Figure 2).

Five months after the first visit to our department of National Defense Medical College hospital, the patient was scheduled to undergo IGSS-assisted canalplasty under general anesthesia. The Medtronic Stealth Station ENT Navigation System was used for IGSS. Four titanium self-drilling screws with a diameter of 1.5 mm and a length of 3 mm were used as reference markers, and a skull-mounted patient tracker was used as the patient tracker. Four reference markers were fixed to the temporal bone under local anesthesia 2 days before the surgery, and a temporal bone CT (with reference markers) examination was performed a day before the surgery. These images were imported into the IGSS for registration. On the day of surgery, an S-shaped incision was made in the retro auricular region, and when the temporal bone was exposed, the 4 reference markers fixed 2 days earlier were identified. Additionally, a patient tracker different from the reference markers was fixed to the temporal bone >2 cm from the reference markers, and the IGSS was activated by registering each marker and tracker on the instrument. After confirming that the 4 reference markers were accurately registered (error of less than a few millimeters) (Figure 2A), they were removed because they interfered with mastoidectomy. When mastoidectomy was performed, the temporal bone was soft and clay-like compared to the normal bone and was easily hemorrhagic. The posterior wall of the EAC was removed while checking the distance to the facial nerve and TM using the IGSS. As the mastoidectomy proceeded, the short crus of the incus was identified, which also confirmed that the IGSS error value was 0.8 mm (less than 1 mm, Figure 2B and C, Supplementary Figure 3, and Supplementary Figure 4A). We confirmed the response of the facial nerve to electrical stimulation in the absence of exposure. Subsequently, an incision was made in the EAC skin, and epithelial debris was extracted from the EAC. Since no abnormal findings were found in the TM (Supplementary Figure 4B) or middle ear, manipulation of the middle ear was not performed. Overhang of the anterior wall of the EAC was observed. However, the anterior wall was not treated because it was fully opened by drilling the posterior wall of the EAC. Finally, meatoplasty was performed, the temporal fascia was applied to the ablated bony surface, and the surgery was completed.

Postoperative pathological examination of the temporal bone revealed that it was an irregularly bifurcated and kyphotic woven bone with an increased number of fibrous connective tissues and spindle-shaped cells with poorly defined round-to-spindle-shaped nuclei (Figure 3A).

No postoperative complications, such as facial paralysis or dizziness, were observed, and the patient was discharged 5 days postoperatively. Three weeks after the surgery, the packing in the ear

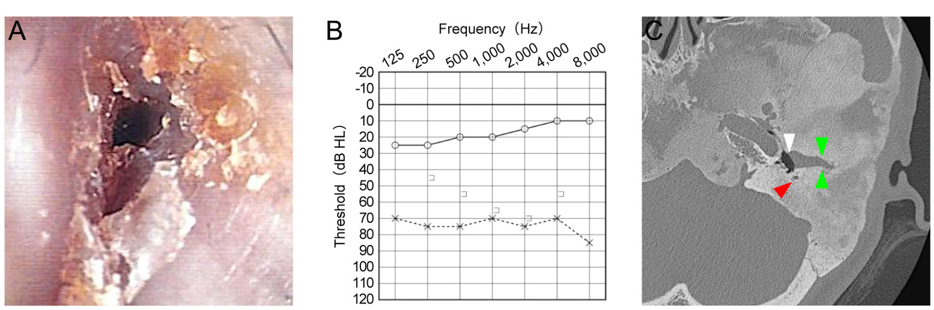

Figure 1. Preoperative findings. (A) Left EAC stenosis was observed. (B) Pure-tone audiometry showing left-sided mixed hearing loss. (C) CT examination showed bone stenosis of the EAC. A soft-tissue shadow was observed in the deep part of the EAC attached to the TM. No facial nerve abnormalities were observed. Green arrowhead: EAC; red arrowhead: facial nerve; white arrowhead: TM. CT, computed tomography; EAC, external auditory canal; TM, tympanic membrane.

was removed, and the patient was followed up at our clinic. Two months postoperatively, the reconstructed EAC was dry, and a fully opened EAC and normal TM were observed (Supplementary Figure 5A). Five months postoperatively, CT showed a fully opened EAC (Supplementary Figure 5B), and audiometry showed marked improvement in left-sided hearing (Figure 3B). No evidence of restenosis of the EAC was observed 1 year postoperatively. Consent for publication was obtained from the patient.

DISCUSSION

Fibrous dysplasia is a benign osteo-proliferative disease of unknown etiology characterized primarily by the replacement of normal bone and marrow bone with fibrous tissue.[1,2] McCune–Albright syndrome, described by McCune, Bruch, and Albright in 1937, is a rare disease characterized by multiple FD, skin pigmentation, and precocious puberty.[7] Among FD in the head and neck region, the temporal bone lesion is extremely rare because the maxilla, mandible, ethmoid, frontal, and sphenoid bones are more common.[8] In addition, FD is more common in young women, whereas temporal bone FD is slightly more common in men. The 3 main symptoms in temporal bone FD are painless swelling of the temporal region, stenosis of the EAC, and conductive hearing loss.[9]

Although surgery is the only treatment for FD-induced EAC stenosis, it is a benign disease, and its progression often stops after puberty. Therefore, patients are generally followed up unless the clinical symptoms are severe. However, temporal bone FD is frequently complicated by EAC stenosis and causes secondary cholesteatoma formation, and surgery is required if these complications occur once.[10] However, there is no consensus on the timing and surgical procedure because of the possibility of restenosis of the EAC, even after surgery. Performing surgery after adulthood is preferred when bone growth ceases rather than during childhood when bone proliferation is more active.[11,12] However, since some cases occur after puberty and disease progression is observed, surgery after puberty does not guarantee that the disease will not recur. Furthermore, there have been reports of secondary cholesteatoma caused by the accumulation of epithelial debris within a few months. Therefore, performing surgical treatment as early as possible in cases of complete stenosis of the EAC is important. Since this is a benign disease with a good prognosis and a very low frequency of malignant transformation, surgery should be limited to minimally invasive treatments for symptomatic improvement.[13] Partial resection, rather than total removal of the temporal bone lesion, is considered appropriate for improving hearing function and preventing complications. The present case is rare, in which

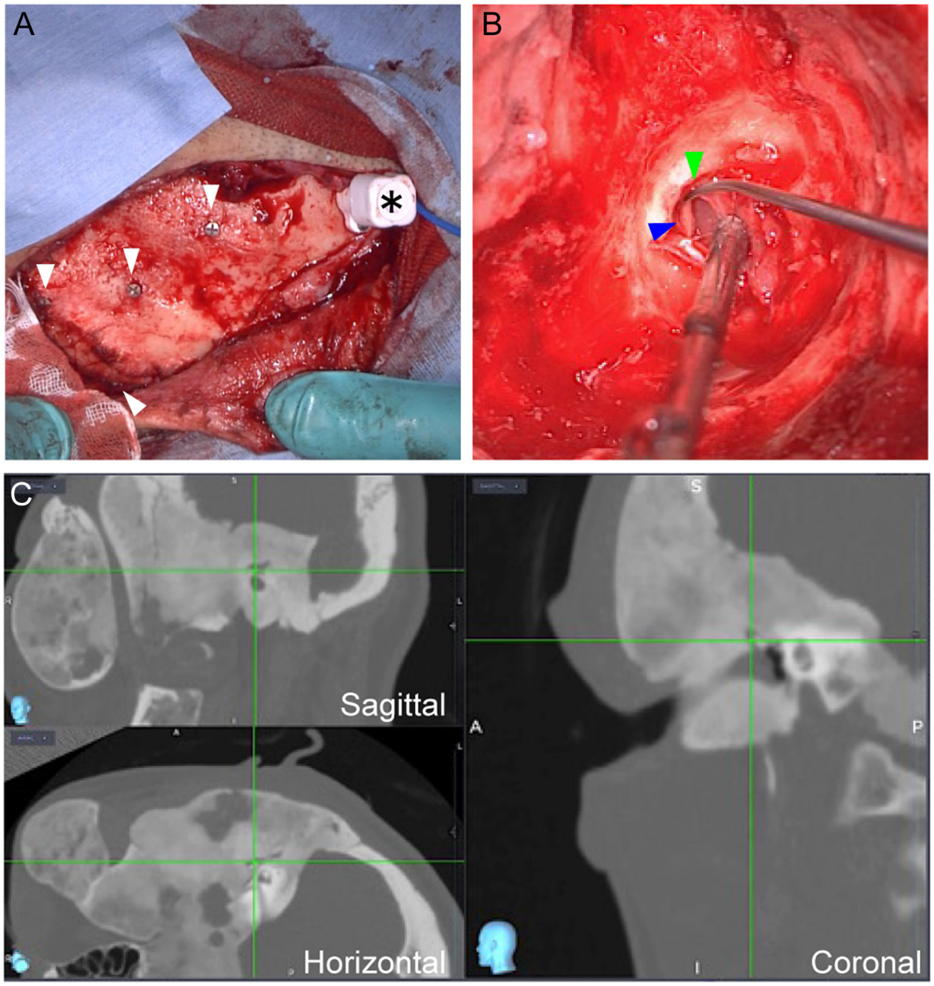

Figure 2. Intraoperative findings. (A) To register the IGSS, the patient tracker (*) was fixed to the temporal bone. Four reference markers are shown (white arrowheads). (B) Surgical field for the identification of the short crus of the incus. Green arrowhead: IGSS probe; blue arrowhead: short crus of the incus. (C) IGSS monitor for confirmation of the incus. IGSS, image-guided surgery system.

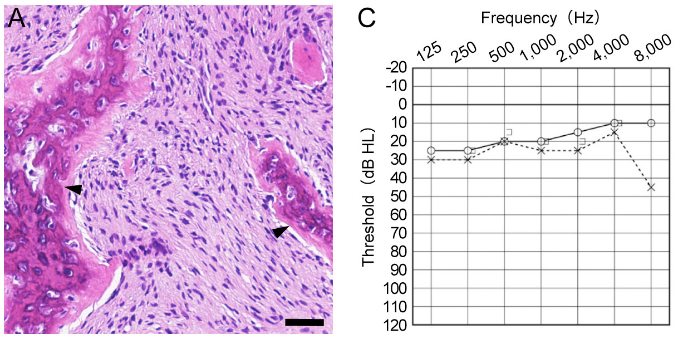

Figure 3. Postoperative findings. (A) Pathological examination shows an increase in spindle-shaped cells with spindle-shaped nuclei and irregularly branched and kyphotic woven bones (black arrowheads). Scale bar is 100 µm. (B) Pure-tone audiometry showing left-sided hearing recovery.

the EAC stenosis slowly progressed to obstruction in adulthood, necessitating surgical treatment, although the stenosis had been noted long before. In addition to the EAC, extensive FD was observed in the left facial bone. However, canalplasty was performed, and the patient's hearing improved.

Although most patients with temporal bone FD showed conductive hearing loss without elevation of bone conduction, this patient had mixed hearing loss, despite proper masking. A previous study demonstrated that hearing loss was identified in 22.4% of ears with temporal bone FD. Conductive hearing loss was the most frequent, affecting 65.9% of the ears. Sensorineural hearing loss affected 29.3% of the ears. Conductive hearing loss, which involves the bony epitympanum, is most commonly associated with FD and leads to ossicular chain crowding and EAC stenosis. Sensorineural hearing loss is associated with IAC elongation.[14] The association between sensorineural hearing loss and IAC length suggests that stretching of its contents may be a potential mechanism for sensorineural hearing loss. Consistent with this, in our case, the IAC length was longer on the diseased side than on the healthy side, as shown in Supplementary Figure 2. However, it has been difficult to improve the bone-conduction threshold after middle ear surgery when there is true bone-conduction threshold elevation, that is, inner ear damage. Previous studies have demonstrated that although the skull bone is the principal contributor to bone-conducted energy transmission, such energy also imposes sound pressure on the EAC and inertial forces on the TM and ossicles.[15-17] This suggests that the EAC pathology in our patient possibly raised the bone conduction threshold and that bone conduction would be improved by ear surgery.

Osteotomy in temporal bone surgery requires a precise understanding of the exact location of the critical organs in the temporal bone in relation to the bony lesion. However, intraoperative changes in the surgical view cause a loss of orientation in cases of malformations that differ from the normal anatomy.[4] When using IGSS, registering the spatial information of the actual surgical field with the positional information on the images is necessary. The accuracy of IGSS in the field of temporal bone surgery, which requires an accuracy of less than a few millimeters, is currently not met by conventional IGSS systems, hindering its widespread use in temporal bone surgery. During registration, multiple reference markers are used to indicate the location of the real space. During surgery, only the patient tracker could be used to display the position of the probe in the body of the image. In nasal sinus surgery, reference markers and patient trackers are applied to the skin, and the position information is registered via electromagnetic tracking. However, both of these methods are based on movable skin and do not provide the accuracy required in the fields of otology and temporal bone surgery. Another reason why IGSS does not work well in temporal bone surgery is that the surface to be registered in temporal bone surgery is not as bumpy as the surface to be registered in sinus surgery. In addition, as surgery progresses, the surgical field becomes progressively deeper and further away from the reference marker, and the position and angle of the head change during surgery. Recently, in the field of skull base surgery, IGSS has been performed with high accuracy by fixing a reference marker or patient tracker to the facial bone. However, CT imaging is required after the reference marker is fixed to the temporal bone.[5] This is not a problem if the hospital has a hybrid operating room where intraoperative CT examinations can be performed. However, careful preparation is necessary for facilities such as ours where intraoperative CT examinations cannot be performed. In this study, we fixed a reference marker to the temporal bone under local anesthesia 2 days before surgery and performed a temporal bone CT scan a day before surgery. Finally, on the day of surgery, the patient tracker was fixed, and each reference marker was registered to allow the intraoperative use of the IGSS. Image-guided surgery systems registered in this manner have been previously reported to have an anatomical measurement error of <1 mm.[18] In the present case, IGSS was possible with very high accuracy, and the IGSS error value was 0.8 mm, at the level of the short crus of the incus and the facial nerve. Although our IGSS is magnetic, it can be applied to the temporal bone region with high accuracy and to the deep surgical field by fixing the reference marker and preparing temporal bone CT image information in advance. In this report, we applied IGSS to EAC plasty for temporal bone FD, which is expected to be useful for other procedures, such as endolymphatic sac release, cochlear implant surgery with inner ear malformations, and EAC cancer, and will contribute to the development of otology and temporal bone surgery.

To fix the reference marker to the temporal bone preoperatively, as in the present case, minor preoperative surgeries and follow-up CT examinations are required. The use of mobile combi-beam CT allows for highly accurate IGSS during an intraoperative procedure in the operating room by fixing a reference marker to the temporal bone and performing intraoperative CT imaging and registration.[6,19] Although IGSS-assisted temporal bone surgery is safe, further development of IGSS is necessary as an established standard procedure for temporal bone surgery.

Availability of Data and Materials: The data that support the findings of this study are available on request from the corresponding author T.K.

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

Peer-review: Externally peer-reviewed.

Author Contributions: Concept – T.K., K.M.; Design – T.K., K.,M.; Supervision – K.M.; Resources – T.K., T.T., S.Y.; Materials – T.K., T.T., S.Y.; Data Collection and/or Processing – T.K., T.T., S.Y., Y.I., M.S., K.M.; Analysis and/or Interpretation – T.K., T.T., S.Y., Y.I., M.S., K.M.; Literature Search – T.K., K.M.; Writing – T.K.; Critical Review – K.M.

Declaration of Interests: The authors have no conflicts of interest to declare.

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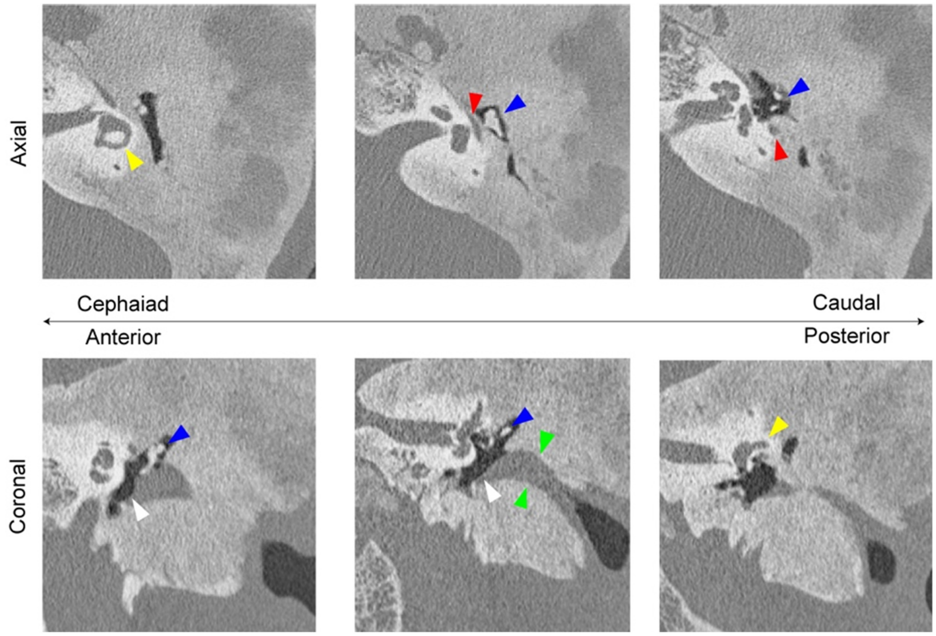
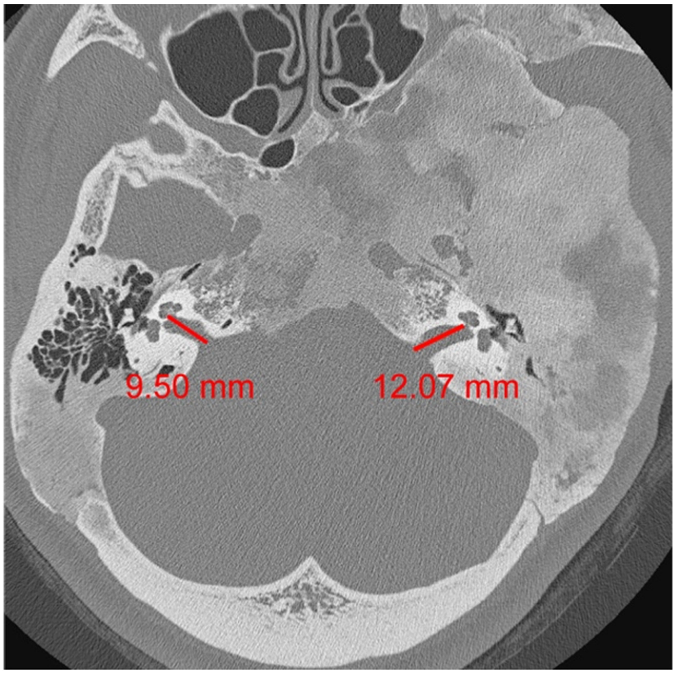
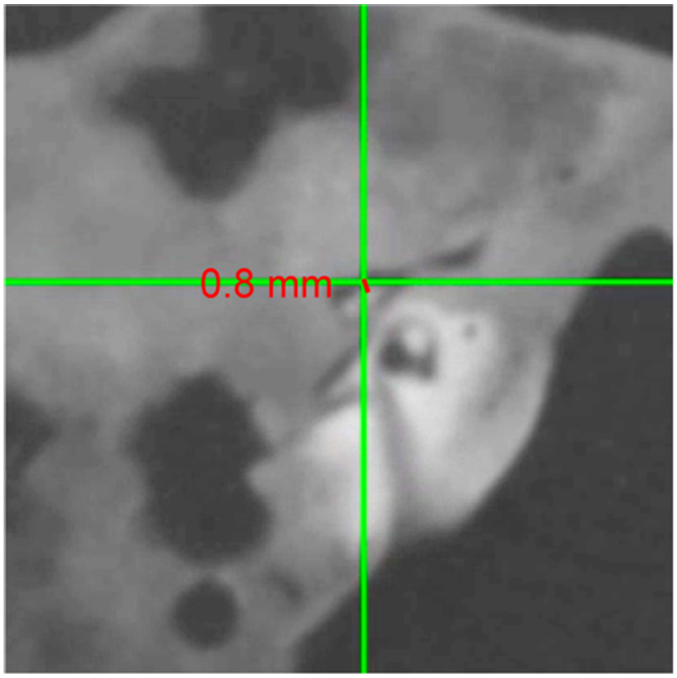


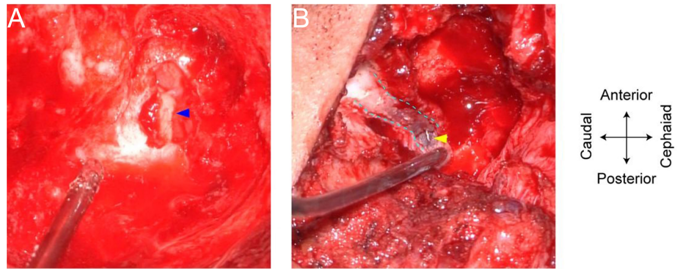
Supplementary Figure 1. Preoperative CT findings: A soft tissue shadow was observed from the deep part of the EAC attached to the TM. Normal ossicles were identified in the middle ear cavity, and no pathologies were observed. No abnormalities were observed in the course of the facial nerve. Green arrowhead, EAC; red arrowhead, facial nerve; white arrowhead, TM; blue arrowhead, ossicles; yellow arrowhead, lateral semicircular canal; CT, computed tomography; EAC, external auditory canal; TM, tympanic membrane.



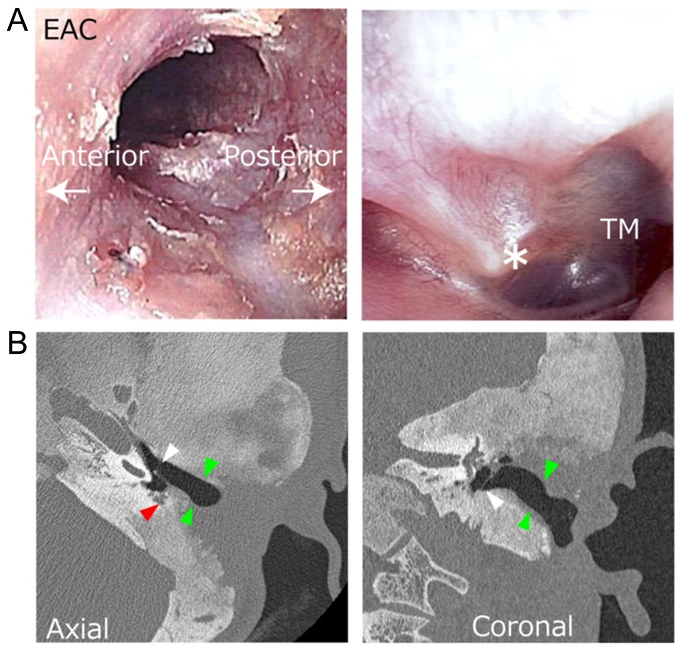
Supplementary Figure 2. IAC length in the preoperative CT image. The IAC length was longer on the diseased side than on the healthy side (12.07 mm > 9.50 mm). IAC, internal auditory canal.



Supplementary Figure 3. Intraoperative IGSS error values. The error value was 0.8 mm. IGSS, image-guided surgery system.



Supplementary Figure 4. Intraoperative findings. (A) Surgical field in the identification of the short crus of the incus. (B) Surgical field in the identification of the TM. Blue arrowhead, short crus of the incus: yellow arrowhead, TM. IGSS: image-guided surgery system, TM: tympanic membrane.



Supplementary Figure 5. Postoperative observation. (A) Two months after surgery, an enlarged EAC was observed. TM appears normal. Asterisk indicates malleus. (B) Five months after surgery, a postoperative CT examination showed a left-sided enlarged EAC. Green arrowhead, EAC; red arrowhead, facial nerve; white arrowhead, TM; CT, computed tomography; EAC, external auditory canal; TM, tympanic membrane.