

CASE REPORT



Combined transmastoid/middle fossa approach for a petrous bone cholesteatoma: A case report and literature review

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ABSTRACT

We report the case of a 14-year-old girl with petrous bone cholesteatoma resected through a combined approach from the middle cranial fossa and mastoid process. A favorable outcome can be achieved when applying the transmastoid approach to a cholesteatoma by incorporating the middle fossa approach to preserve inner ear function. We emphasize the utility of integrated three-dimensional image reconstruction as a surgical strategy to determine beneficial routes for facial and inner ear function and accomplishment of radical resection. Visualization by an integrated image of irregular tumor infiltration within the petrous bone facilitates information sharing for better cooperation between surgeons in otolaryngeal and neurosurgical fields.

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Petrous bone cholesteatoma; Middle fossa/transmastoid combined approach; Hearing preservation; Integrated 3D image; Simulation

Background

A petrous bone cholesteatoma is defined as a cholesteatoma that is congenital in nature and is detached from the tympanic membrane or external auditory meatus. It is a rare pathologic entity with a reported incidence of 4%–9% of all petrous pyramid lesions [1]. Such lesions span from the upper internal auditory meatus to the geniculum of the facial nerve and the petrous bone of the superior semicircular canal. Various approaches can be considered, including the translabyrinthine approach or the middle cranial fossa. Tumors are located in such complex anatomical region that in any surgical approach, radical resection of the tumor with the preservation of VII and VIII nerve function is challenging. To solve this problem, we performed a preoperative three-dimensional (3D) simulation to grasp safe surgical corridors connecting the otological and cranial routes.

Here, we report a case of petrous bone cholesteatoma with a favorable outcome achieved through a combination of two approaches intended to preserve

the inner ear function and emphasize the utility of integrated 3D image reconstruction as a surgical strategy to determine beneficial routes for facial and inner ear function and accomplish radical resection.

Case description

A 14-year-old girl experienced frequent episodes of otitis media for more than six years since childhood. In July 2011, the patient was examined at a clinic with the chief complaint of hearing loss in the right ear. Pure tone audiometry confirmed conductive hearing loss in the right ear with scores of 55.0 and 10.0 dB in the right and left ears, respectively. Her VII, IX, X, and XI cranial nerve functions were intact. She was diagnosed with right congenital cholesteatoma and was referred to our hospital's otolaryngology department to which she was admitted for treatment associated with a neurosurgical procedure in October. Preoperatively, the patient was alert and oriented without any neurological abnormalities. Blood sampling revealed no abnormalities in the

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CT was performed with a 64-multidetector row CT scan (Aquilion ONE, Toshiba Medical System, Corp., Tokyo, Japan) at 0.5-mm interval. Cranial CT was performed with a 64 multidetector row CT scan (VCT, General Electric Healthcare, Japan) with a 5.0-mm interval. Running the workstation of a 3D image analysis system (Synapse Vincent, Fuji Film Co., Tokyo, Japan), any structure can be extracted from the acquired images [2]. The tumor, semicircular canals, cochlea, and facial canal were tracked and visualized using petrous bone CT. These visualized structures were integrated with the cranial CT. The positional relationship of these structures from any viewing direction was revealed by rotating and making the skull transparent. Surgical views of the transmastoid and middle fossa approach were created, and the information was provided to otolaryngologists and neurosurgeons. Fast imaging employing steady-state acquisition showed progression of the lesion into the upper region of the anterior semicircular canal and vicinity of the arcuate eminence. There were no abnormalities in the facial nerve, vestibular nerve, cochlea, or semicircular canal (Figure 1(c–f)).

Course in the hospital

Simulated by integrated 3D images (Figure 1(d,e,f,g)), we applied a combined approach intended to preserve the inner ear function wherein we resected the lesion in the geniculum of the facial nerve *via* the transmastoid approach and lesion in the anterior semicircular canal and petrous region *via* the middle fossa approach.

Intraoperatively, the elastic, soft, pearly colored lesion was found to have progressed from the geniculum of the facial nerve to the petrous region (upper fundus of the internal acoustic meatus to the petrous apex of the superior semicircular canal) and was accompanied by bone destruction with fistulization. Irregularly shaped tumors invading the bone were removed using an ultrasonic aspirator with a power output of 100% (Sonopet, Stryker Japan, Tokyo), by which tumor resection proceeded with peritumoral petrous bone tissue, ensuring all are resected. The direct facial nerve stimulation was monitored by evoked electromyography (EMG) of the right orbicularis oculi and oris muscles throughout the procedure. Total removal was achieved without cranial nerve damage. Open-type congenital cholesteatoma also existed in the epitympanic to the middle tympanic cavity and the lateral wall of the horizontal semicircular canal with the fragile matrix. The long process of the incus

and the superstructure of the stapes disappeared. After preserving the chorda tympani, the thin matrix was removed from the facial recess and the horizontal portion of the facial nerve. There was no apparent fistula at the horizontal semicircular canal, although the bony canal was thin due to cholesteatoma. The ossicular reconstruction was not performed during the first surgery. The ossicular reconstruction in the middle ear was performed 1 year after the operation using the cortical bone obtained from the temporal bone (tympanoplasty type IVc). In the cranial approach, we detected a greater superficial petrosal nerve, geniculate ganglion, and facial nerve by 1 mA stimulator. In the transmastoid approach, tumor removal around the geniculate ganglion was performed under facial nerve monitoring. During removal of the tumor near the facial nerve around the internal meatus, the amplitude of the EMG in both muscles was reduced to 65% before the removal and recovered to basal level after finishing the resection.

Follow-up

Postoperatively, there were no notable complications, and the patient was discharged. Five years after ossiculoplasty, pure-tone audiometry demonstrated that hearing had been preserved (Figure 2).

Pathological findings

Pathology showed squamous epithelial cells consisting primarily of keratin and cholesterol and were diagnosed as epidermoid cysts. These findings led to the diagnosis of cholesteatomatous otitis media (Figure 3).

Discussion

This case progressed favorably as a result of the resection using the combined transmastoid/middle fossa approach to treat right petrous bone cholesteatoma. The combined transmastoid/middle fossa approach is used to treat petrous bone cholesteatoma and other cerebellopontine angle tumors, such as acoustic schwannoma [3–5]. The choice depends on various factors, including tumor progression, tumor size, degree of hearing loss, and the extent of increased intracranial pressure [3]. With the translabyrinthine approach alone, surgery can be performed without applying pressure to the brainstem. However, this method causes deafness and is, therefore, only appropriate in cases in which hearing preservation is

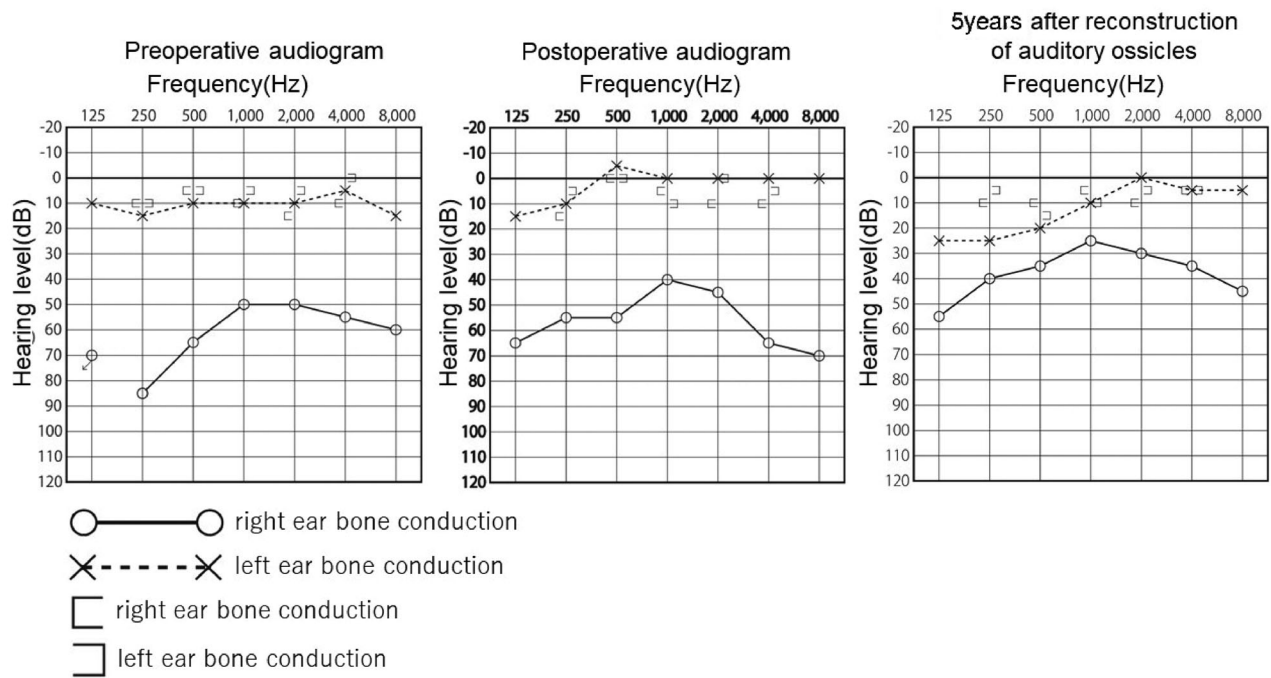


Figure 2. Preoperative and postoperative audiograms. Note the bone conduction hearing threshold was preserved after surgery, and the hearing ability of the right ear improved from 55.0 to 46.7 dB by air conduction at a frequency of 1,000–2,000 Hz.

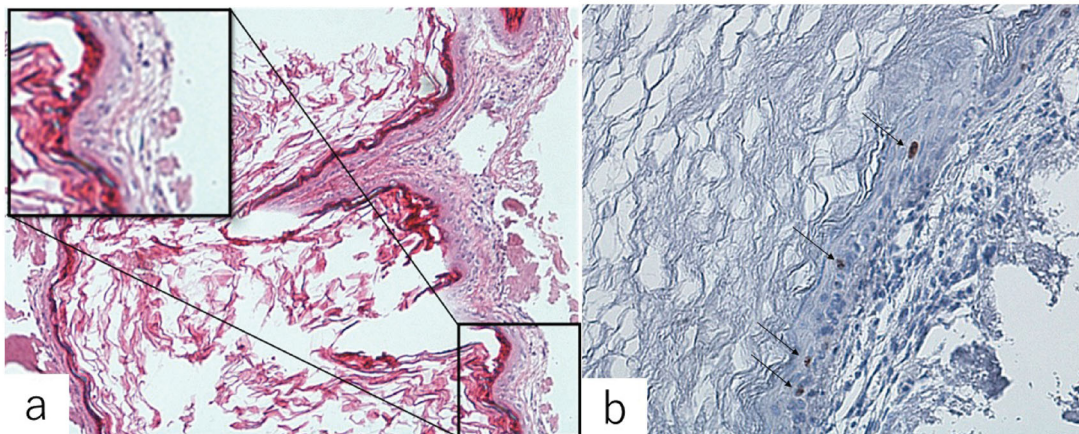


Figure 3. Neuropathological findings. Tissue is primarily composed of keratin and cholesterol. Stratified squamous epithelial cells are observed (black box) (a). Immunostaining shows the MIB-1 stain scattered throughout the stratified squamous epithelium (black arrow). The positive rate is 2.9%. Magnification, x20 in a and b, magnified inset in a, x35.

unnecessary. In combination with the middle fossa approach, the internal auditory meatus can be operated without passing through the labyrinthine [6].

In this report, we suggest the following two points: First, for the selection of surgical strategy, an integrated 3D image is useful for detecting irregularly infiltrated tumors in the petrous bone, which hinders total resection and causes tumor recurrence. Sophisticated high-quality 3D computer graphics data can be acquired from patient-specific data using commercially available software [7]. Selecting the operative corridor and visualizing the microsurgical

anatomy fully depend on the creator. Feedback from real operative information further stimulates highly realistic simulations. Second, an ultrasonic aspirator is a powerful tool to accomplish the total removal of the tumor, because it can easily resect hard bone tissue where the tumor invaded. Highly proliferative tumor membranes that adhere to hard bone tissue are generally difficult to dissect. In this case, the tumor invaded the arcuate eminence and superior semicircular canal. The outer diameter of the Sonopet tip is 1.7 mm; therefore, it is safe and useful for cutting the bone into small fragments [8]. We believe that to

Table 1. Cases of combined transmastoid and middle fossa approaches.

Author	Sex	Age	Clinical features	Diagnosis	Bone finding	Complication	Healing ability	Resection extent
Liu et al. 2014 ^[3]	M	35	VIIN dysfunction	Cholesterol	details not showing	none	Sensorineural hearing loss (unchanged)	Partial
Tabuchi et al. 2012 ^[5]	F	10	Dizziness	granulomas	TM defect	none	good	Complete
	F	56	unknown	Cholesteatoma	TM/PW defect	none	good	Complete
	M	67	unknown	Cholesteatoma	TT/TM/PW defect	Cerebrospinal fluid leakage	moderate	Complete
	M	45	unknown	Cholesteatoma	TT/TM defect	none	good	Complete
	M	64	unknown	Cholesteatoma	TT/TM/PW defect	none	moderate	Complete
Steward et al. 2000 ^[6]	M	10	unknown	Cholesteatoma	PW defect	none	moderate	Complete
	M	51	unknown	Cholesteatoma	TT/TM defect	none	good	Complete
	M	67	unknown	Cholesteatoma	TT/TM defect	none	moderate	Complete
	7Female	17–63y	Healing loss	Cholesteatoma	3 cases fistulae	none	unchanged	Complete
	4Male		Otorrhea	Cholesteatoma	7 cases erosion	none		Complete
Miyazato&Hokama present case	F	14	Right hearing disturbance	Cholesteatoma	bone destruction and fistulisation	none	good	Complete
			Right otitis media					
TT: tegmen tympani, TM: tegmen mastoideum, PW: posterior wall of the mastoid.								

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specifically prevent recurrence, it is important to totally dissect infiltrating tumors into the petrous bone without any nerve damage or CSF leakage.

A literature review revealed 20 cases of combined transmastoid/middle fossa approach. Patients included both men ($n = 11$) and women ($n = 9$), ranging in age from 10 to 67 years, diagnosed with petrous bone cholesteatoma or cholesterol granuloma (Table 1). Some patients presented with hearing loss, but none of the patients had a postoperative onset or worsening of hearing loss. Cerebrospinal fluid leakage was confirmed in one patient. In the present case, we preserved the anterior semicircular canal with the transmastoid approach for the lesion in the geniculum of the facial nerve and the middle fossa approach for the lesion in the anterior semicircular canal and petrous region. Complications of brain compression or cerebrospinal fluid leakage have been reported [5]. To minimize compression of the brain parenchyma due to the middle fossa approach in this case, we established spinal drainage in advance, drained approximately 20–40 mL of cerebrospinal fluid preoperatively, and administered an osmotic diuretic to minimize damage. Although cerebrospinal fluid leakage occurred intraoperatively in this case, outflow from the internal auditory meatus was confirmed during the resection of the cholesteatoma; therefore, the area was treated with calcium phosphate bone cement (BIOPEX®, HOYA Technosurgical Co. JAPAN TOKYO) and fascia grafts. This is a useful surgical technique for resecting a tumor going beyond the inner ear into the petrosa with the aim of preserving hearing and the facial nerve.

Conclusion

We report the case of a 14-year-old girl with right petrous bone cholesteatoma who underwent surgery using the combined transmastoid/middle fossa approach. A favorable outcome for facial and inner ear function and radical resection was accomplished. Preoperative visualization by 3D integrated images of irregular tumor infiltration within the petrous bone is useful for the selection of a surgical approach and facilitates information sharing between specialists in otolaryngeal and neurosurgical fields.

Disclosure statement

No potential conflict of interest was reported by the author(s).

Informed consent statement

The written consent regarding the use of clinical findings was obtained from the patient.

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