—Case Reports—

Microtia with Coexistent Large External Auditory Canal Cholesteatoma: A Case Report

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Microtia is a congenital disorder characterized by an anomaly in the auricle. It is often associated with atresia, stenosis, or obstruction of the external auditory canal. In cases of microtia, the incidence of a normal external auditory canal, congenital external auditory canal stenosis, and congenital complete closure of the external auditory canal (aural atresia) is reported to be 8%, 8%, and 84%, respectively. Congenital stenosis of the external auditory canal can sometimes be accompanied by intractable otorrhea and otalgia, raising the possibility of complications of external auditory canal cholesteatoma. Here, we report a case of an adult with microtia and external auditory canal stenosis who presented to our clinic with complaints of otalgia. A large external auditory canal cholesteatoma was found in the patient's left ear. Although cholesteatoma is common in cases of canal stenosis, its extensive spread within the temporal bone is quite rare. A temporal bone-targeted computed tomography scan revealed a soft tissue shadow in the left external auditory canal with distensible expansion and bony destruction in the upper, anterior, and posterior walls of the external auditory canal. In patients with microtia who experience severe aural pain, the possibility of latent extended cholesteatoma should be considered.
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Key words: external auditory canal, microtia, cholesteatoma

Introduction

Microtia is often associated with congenital aural atresia (CAA) or external auditory canal (EAC) stenosis. In children with CAA or EAC stenosis, particularly those with a narrow EAC, cholesteatoma formation is a recognized risk. In previous retrospective studies of patients with CAA and congenital EAC stenosis, 19-48% of patients developed cholesteatoma during follow-up[1].

Any child with EAC stenosis that cannot be properly examined should be considered at risk of developing acquired EAC cholesteatoma, particularly if the tympanic membrane is not visible. Careful consideration is required in these cases. Some children have a stenotic EAC that can be cleaned. Computed tomography (CT) of the temporal bone is crucial for diagnosing cholesteatoma in children with stenotic EAC, although it involves a small dose of radiation and requires sedation. If this type of detailed examination is missed during childhood, undiagnosed EAC cholesteatoma could result in serious complications, including destruction of the temporal bone, severe infection, facial nerve disturbance, loss of middle or inner ear function, and intracranial extension[2].

Cases of acquired EAC cholesteatoma with extensive temporal bone involvement are rare. Here, we report a case of a large acquired EAS cholesteatoma in an adult that had grown and extended widely in the temporal bone over a long period.

Case Presentation

This case report complied with the Helsinki Declaration

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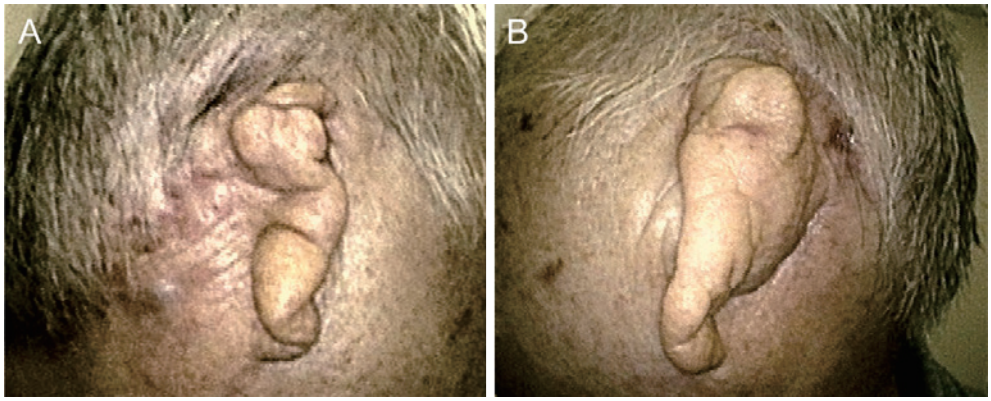

Fig. 1 Appearance of the auricles
Bilateral microtia are shown (A: right ear, B: left ear).

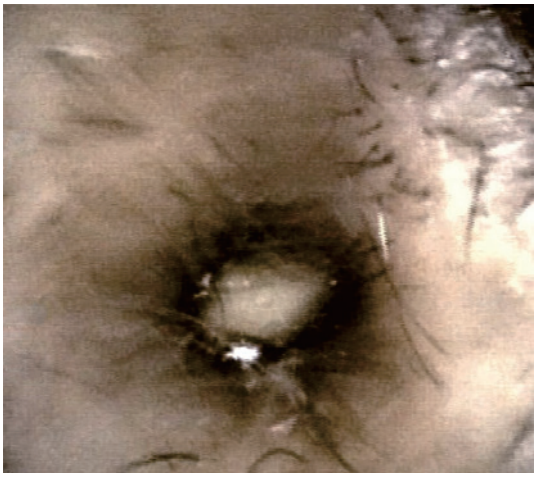

Fig. 2 Appearance of the left external auditory canal
The external auditory canal was highly constricted a few millimeters from the entrance.

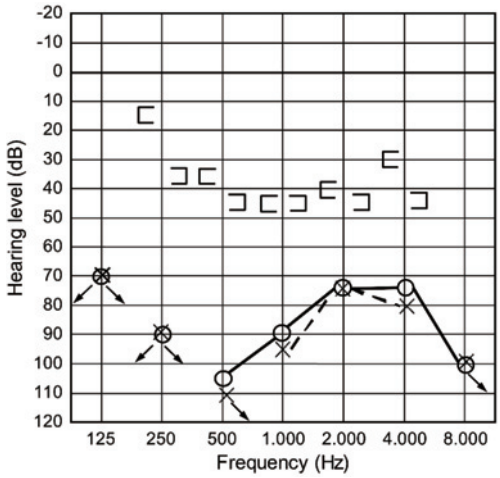

Fig. 3 Audiogram
Pure tone audiometry shows bilateral conductive hearing loss.

standards, and we obtained written informed consent from the patient. We examined a patient with a large, acquired EAC cholesteatoma that had grown and spread extensively in the temporal bone over a long period.

A 73-year-old man was referred to our department owing to severe left-sided otalgia with a significantly narrowed EAC, requiring investigation and treatment. At birth, he had been noted to have deformities of bilateral auricles, atresia of the right external auditory canal, and stenosis of the left external auditory canal. Since childhood, he has experienced bilateral conductive hearing loss and has worn bilateral bone conduction hearing aids. Because his last visit to the ear-nose-throat clinic was more than 50 years ago, he had no relevant medical records. Therefore, his early otological details are unknown.

There was no notable family or medical history. The chief complaints at the time of presentation were pain in the left ear and occasional spasms of the left side of his face. He had visited the otolaryngologist and neurologist at a local hospital, where no significant lesion was identified in the central nervous system at the department of neurology. CT revealed a large soft tissue mass in the left external auditory canal, leading to his referral to our hospital for further examination and treatment.

Current Medical History

Bilateral microtia (**Fig. 1A, 1B**), complete closure of the right EAC, and severe stenosis of the left EAC were noted. The EAC was highly constricted about 5 mm from the entrance (**Fig. 2**). Pure tone audiometry revealed bilateral conductive hearing loss (**Fig. 3**), and the patient wore a bone conduction hearing aid in the form of a hairband on the left side.

CT Findings

Temporal bone-targeted CT revealed a soft tissue shadow in the left EAC with distensible expansion and

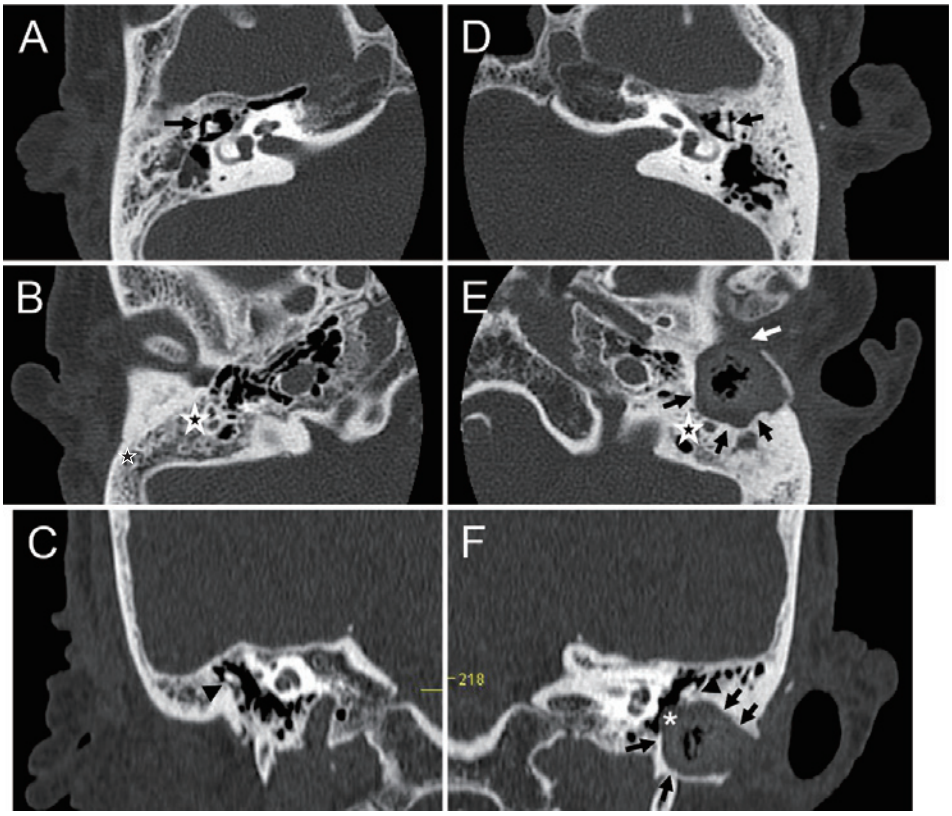

Fig. 4 CT imaging of the present case
A large soft tissue shadow is widely expanded, and bony destruction in the upper, anterior, and posterior walls of the EAC is observed (**E, F**: black arrows). The mass is in contact with the left temporomandibular joint capsule (**E**: white arrow). The mass protruded into the tympanic cavity along with the soft tissue (**F**: asterisk). Although the malleus and incus were observed on both sides, their shapes were deformed. The Malleus head and incus body were bony attached (**A, D**: black arrows). Both malleus heads are adhered to the scutum (**C, F**: black arrowheads). The facial nerve passes more anteriorly than usual through the mastoid process of the temporal bone on both sides (**B, E**: black stars).

bony destruction in the anterior, posterior, and upper walls of the EAC. The mass was in contact with the capsule of the left temporomandibular joint (**Fig. 4E, 4F**).

The right EAC was completely absent and closed. The mass protruded into the tympanic cavity with soft tissue (**Fig. 4F**). However, both tympanic cavities and the mastoid antrum were small but air-containing. The malleus and incus were observed on both sides, but were deformed. The malleus head and incus body were adherent to the bone (**Fig. 4A, 4D**), and the malleus heads were adherent to the scutum on both sides (**Fig. 4C, 4F**). The superstructure of the stapes was barely visible on both sides. The mastoid air cells were poorly developed. There was no obvious malformation on either side of the inner ear. The facial nerve passed more anteriorly than usual through the mastoid process of the temporal bone on both sides (**Fig. 4B, 4E**).

A white, pearl-like mass was visible through the pinhole-shaped left EAC foramen. A biopsy was performed, and histopathological examination revealed keratinized stratified squamous epithelium, which is typical of cholesteatoma.

Intravenous infusion of Ceftriaxone Sodium Hydrate, 2 grams per day, was administered against the infection of cholesteatoma and its surrounding tissues. The patient underwent surgical treatment of the left EAC stenosis to remove the cholesteatoma and reconstruct a wide EAC.

A postauricular incision was made, and dissection was carried out to expose the cortex of the temporal bone. Inflamed tissue under the skin was debrided. The cortical bone was removed from the area where the EAC was presumed to be, revealing an enlarged canal filled with the debris of the cholesteatoma (**Fig. 5**).

There was partial bony destruction of the upper and posterior EAC walls, but the cholesteatoma had not penetrated the mastoid cavity from the lateral side of the

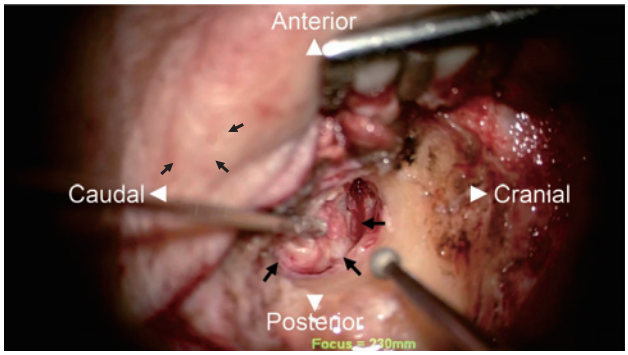

Fig. 5 Intraoperative view
The cholesteatoma was removed by careful drilling of the temporal bone on the left side. Cholesteatoma is indicated by black arrows.

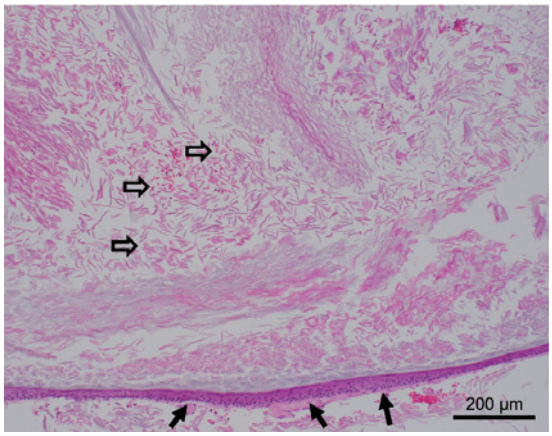

Fig. 6 Histopathological findings
Keratinized stratified squamous epithelium (black arrows) with keratinized material (white arrows) was observed (×100, Scale bar = 200 μm).

EAC. The temporomandibular joint capsule remained intact anterior to the canal. The deep part of the EAC was closed and the tympanic membrane could not be clearly identified. Because the patient had moderate conductive hearing loss, we suggested that tympanoplasty should be performed simultaneously. However, he preferred to continue wearing the bilateral bone conduction hearing aids. Moreover, because he did not consent to manipulation within the tympanic cavity, we could not confirm the condition of the ossicles.

The EAC was widened by enlarging the canal entrance and smoothing the bony canal walls through drilling. The entrance to the cartilaginous EAC was severely constricted, which was presumed to be the cause of cholesteatoma development. This part of the cartilage was removed to widen the opening of the EAC. The skin defect on the wall of the EAC was covered with a pedicle flap, and the operation was completed. Specimens were submitted for histopathological examination, and keratinized stratified squamous epithelium with keratinized material was observed, consistent with cholesteatoma (**Fig. 6**).

The postoperative course was favorable, with the left EAC becoming epithelialized and dry within about two months. The auditory ear canal remained wide without signs of restenosis (**Fig. 7**).

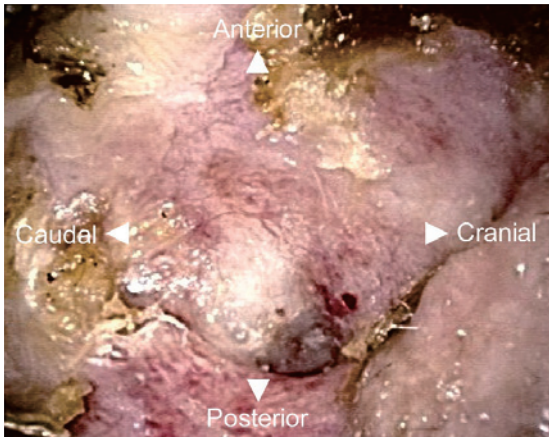

Fig. 7 Postoperative view
The left auditory ear canal was wide without any tendency for restenosis.

Discussion

Deformity of the external and middle ear is a common anomaly in otology clinics, with an incidence rate ranging from 0.05% to 0.1%[3]. The archetype includes microtia, varying degrees of EAC anomalies ranging from complete closure (atresia) to mild stenosis, and anomaly of the middle ear. Furthermore, 11% to 47% of patients also have inner ear anomalies[4]. The diagnosis of congenital anomalies of the external, middle, and inner ear is based on functional examinations and radiologic imaging, such as CT and magnetic resonance imaging[5].

Cholesteatoma is a keratinized epidermal inclusion cyst that typically develops in the middle ear or EAC. It is characteristically observed as a round, pearly mass or as an accumulation of keratinized debris, often accompanied by bony erosion. Although most cholesteatomas develop in the middle ear, they can also form in the EAC and involve the bony EAC lateral to the tympanic membrane[6]. If the keratinized epithelium in the EAC cannot be expelled normally owing to an anomaly, it may continue to accumulate, and EAC cholesteatoma could develop. It has been suggested that EAC diameter is an important factor in the formation of cholesteatoma[7].

There are only a few reported cases of external microtia associated with external auditory canal cholesteatoma[1]. As mentioned in the introduction, if a child loses the opportunity for early examination, such as CT, there is a risk that the EAC cholesteatoma may grow and extensively invade the temporal bone. However, such cases are extremely rare.

The characteristics of the present case can be summarized as follows: 1) accumulation of epithelial debris in the narrowed EAC entrance; 2) the debris extends widely, forming a large cholesteatoma mass in the temporal bone; 3) this results in compression and destruction of the surrounding bone; and 4) infection spreading to surrounding areas.

Microtia is a congenital disorder characterized by tissue defects in the auricle, frequently associated with EAC atresia and stenosis, and varying degrees of ossicular anomalies. Bilateral congenital abnormalities of the external ear are rare, occurring in approximately 1 in 200,000 individuals. Embryologically, the auricle, EAC, and middle ear are derived from the first and second branchial arches, which are closely related and often result in anomalies. In contrast, the inner ear originates from the otocyst dorsal to the branchial arch and is less frequently associated with anomalies because of its different developmental origins[3,4]. In cases of congenital atresia, the therapeutic strategy is divided into bilateral and unilateral conditions. Bilateral cases usually present with moderate conductive hearing loss and may delay language development, requiring the use of bone-conducting hearing aids and speech therapy as soon as possible after birth. However, interventions for hearing are not necessary in cases of unilateral microtia when hearing is intact on the contralateral side, and only auriculoplasty is subsequently required in occasional cases.

Meatoplasty and tympanoplasty are considered for bilateral EAC atresia where the middle ear anomaly is severe[8]. In the present case, the patient had right-ear EAC atresia and left-ear stenosis. He was happy to continue wearing a bone-anchored hearing aid on the left side and did not request auriculoplasty or tympanoplasty for ossicular chain reconstruction to improve his hearing.

Congenital EAC stenosis is associated with a high risk of cholesteatoma owing to a reduced self-cleaning system and accelerated epithelial keratinization associated with cerumen accumulation in the deep skin of the stenosis. Cole and Jahrsdoerfer[9] reported a 59% complication rate of cholesteatoma when the stenosis was less than 2 mm, and this rate increased to 91% when the factor of age greater than 12 years was added. In cases diagnosed after the age of 20 years, cholesteatoma can extend into the tympanic cavity, mastoid cavity, and temporomandibular joint, resulting in a destructive situation. In contrast, Schuknecht[10] reported congenital aural atresia in 51 cases, of which only one had EAC cholesteatoma. In cases of severe congenital aural atresia, such as complete loss of the cartilaginous or bony EAC, the absence of epithelium makes cholesteatoma formation rare. However, in cases of thin membranous atresia, cholesteatoma can occur owing to the presence of an EAC and epithelium at depth. Altmann's classification of stenosis tends to recognize atresia as a more severe condition than stenosis owing to its more severe morphological abnormality[11,12]. However, stenosis requires close attention because the skin is narrowly present in the EAC and the incidence of cholesteatoma increases with age. It is important to carefully examine the extent of atresia, whether it is fibrotic or bony atresia on CT[13].

In the present case, the diameter of the EAC opening was 2.75 mm and the patient felt no pain or otorrhea for an extended period. The patient was satisfied with his bone-conducting hearing aid and did not request auriculoplasty or hearing improvement surgery, resulting in no chance for otological follow-up. This might have allowed the cholesteatoma to develop significantly in the present case. The facial nerve was not in contact with the cholesteatoma on the CT image (**Fig. 4E**), and it was not observed during surgery. However, the occasional spasms of the left side of the face that the patient experienced were likely related to the presence of the cholesteatoma because the spasms disappeared after surgery.

Since no surgical manipulation was performed inside the tympanic cavity, it was not possible to diagnose the presence or absence of fixation or disarticulation of the ossicles in the middle ear.

There are three goals in the treatment of microtia and external auditory canal stenosis: (1) reconstruction of ear morphology, (2) improvement of hearing, and (3) prevention of complications. Since multiple surgeries are required to achieve all three goals and the treatment period is lengthy, patient and family understanding and cooperation are necessary[8]. In contrast, the first two goals are often determined based on the patient's and family's preferences; complications, such as EAC cholesteatoma, are not. Medical staff should carefully explain the possible complications of the disease and intervene in treatment.

Although ear canal stenosis is relatively rare, this case

reminds us that leaving ear canal stenosis untreated for an extended period may be associated with the wide extension of external auditory canal cholesteatoma.

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