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Case report

Salivary gland choristoma of the middle ear with alopecia

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ABSTRACT

Middle ear salivary choristoma is a rare entity and only 40 cases have been detected since the first report in 1961. Choristoma is a benign tumor, but can cause conductive hearing loss. Therefore, appropriate treatment is important for patients. Here, we have presented one case of salivary gland choristoma in the middle ear with alopecia and have included a review of the literature.

1. Introduction

Choristoma is a benign tumor defined as a mass of tissue that is histologically normal but located in an abnormal site. Salivary choristoma in the middle ear is extremely rare, and there have been 40 cases reported since the first description in 1961. Here, we have presented another case of salivary gland choristoma located in the middle ear with associated alopecia.

2. Case report

A 10-year-old girl was referred to Kanazawa University hospital for the evaluation of unilateral hearing loss in the left ear, which was first identified at the age of 6 years. She had no other otologic disorders. She had no family history of deafness, and she was born after a normal pregnancy and delivery. Otoscopic examination detected a pinkish mass at the posterosuperior quadrant of the left tympanic membrane with no associated perforation. Physical examination revealed alopecia only in her left ear (Fig. 1). A pure-tone audiometry test revealed a 77.5 dB mixed hearing loss in the left ear (Fig. 2). The right ear was normal. The stapedial reflex was absent in the left ear. Computed tomography (CT) of the temporal bone confirmed the presence of a soft mass at the level of the hypotympanum (Fig. 3). It appeared to be adjacent to a horizontal portion of the facial nerve. We suspected congenital cholesteatoma or facial nerve neurinoma as a preoperative diagnosis.

An exploratory tympanotomy was performed through a transmeatal approach (Fig. 4). The superstructure of the stapes was defective. The other ossicles and the facial nerve showed no abnormalities. The mass was smooth and attached to a horizontal portion of the facial nerve. It was shaped like a snowman, and the bottom part was reactive to nerve integrity monitor (NIM) but the head part was nonreactive. We resected the head part and performed intraoperative consultation. The frozen section showed glandular tissue, and there was no malignant tissue. The bottom part was left in place to avoid the occurrence of any postoperative facial palsy. Interposition with an incus was performed to reconstruct the ossicular chain. Histopathologic examination revealed glandular tissue, which consisted of serous and mucinous glands (Fig. 5). It was also partially covered with pseudostratified epithelium.

One month after the surgery, a pure tone audiometry of the left ear was 16.3 dB (Fig. 6). During the follow-up 1 year later, there was no change in her facial nerve function and no evidence of tumor growth.

3. Discussion

Choristoma is a mass of tissue that is histologically normal but is located in an abnormal location. A salivary gland choristoma is a normal salivary gland tissue in an ectopic location; however, its

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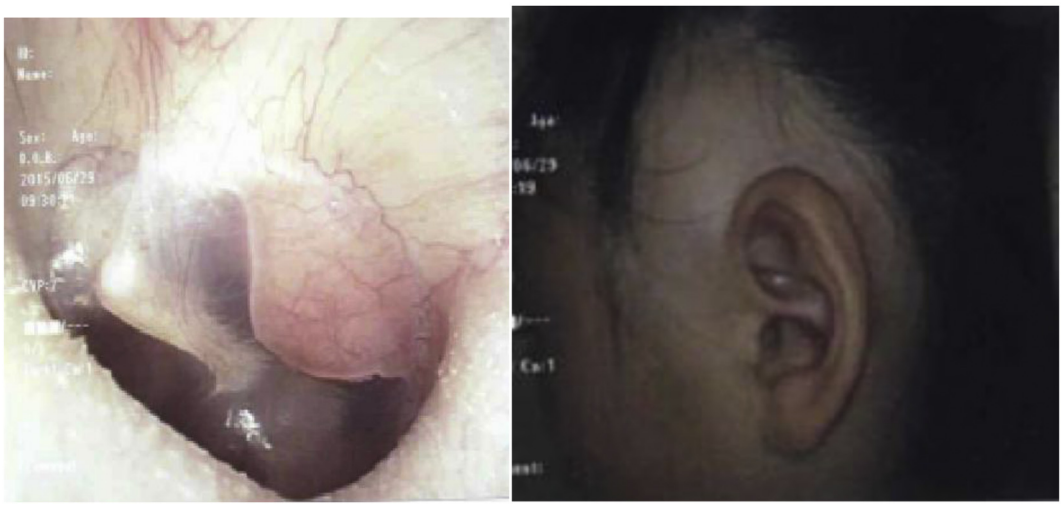

Fig. 1. Pinkish mass in the left middle ear in PSQ without perforation of the tympanic membrane (*left*). The alopecia existed around the left auricle (*right*).

presence in the middle ear is extremely rare.

Our review of PubMed revealed only 40 cases, including our case, since Tayler and Martin first reported a salivary gland choristoma in the middle ear in 1961. Alessandra et al. had reviewed 31 cases up to 2004; these authors found that there was a left-side predominance and a higher incidence among females below 20 years of age [1]. In most of the cases, chief complaints were conductive hearing loss. These patients had anomalies in the ossicular chain or facial nerve. Subsequently, we reviewed nine cases after Alessandrais review in 2004 (Table 1).

In the nine cases, the patients were between 10 months and 32 years of age, with a mean age of 11.4 years; eight of these cases (89%) were females. Excluding the case of a patient with mental retardation, all cases were associated with unilateral conductive hearing loss. In this case, the alopecia around the auricle was found only in the left ear, which had been reported in only two of the 40 cases that were previously reported [2,3].

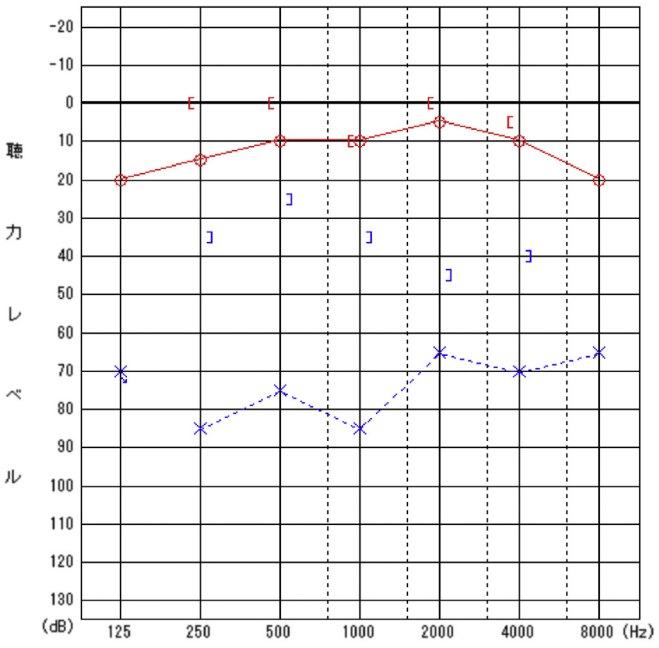

Fig. 2. A pure-tone audiometry revealed 77.5 dB conductive hearing loss.

The occurrence of salivary gland choristoma of the middle ear is unknown but may be associated with the embryological failure of the first or second branchial arch during the 4th week of gestation [4]. Peron and Schukunecht postulated that an aberrant parotid bud remained in the lateral skull bone [5]. Meanwhile, Hinni and Beaty presumed that the extension of the primitive salivary gland into the middle ear could cause salivary gland choristoma [6]. In most cases, the patients had abnormalities of the ossicles or facial nerve, which implied occurrence before the organogenesis.

In cases of middle ear tumors, the differential diagnosis by clinical presentation is difficult. We consider congenital cholesteatoma, facial nerve neurinoma, glomus tumor, teratoma, and lymphoma. As for the diagnosis of choristoma in the middle ear, Buckmiller et al. [7] mentioned the following four criteria: (1) normal salivary gland tissue in the middle ear, (2) lateral conductive hearing loss, (3) abnormalities of the ossicles, and (4) an aberrant facial nerve. In their report, 86% of the cases met more than three of the standards.

Diagnosis using the clinical features at an early stage is needed. Despite the progress of imaging, we can only diagnose by a histologic examination.

When sampling the choristoma, careful management of the facial nerve is required. In some cases, removal of the choristoma caused a postoperative facial palsy [8,9]. When the mass is unconnected to the facial nerve, a complete removal could be performed. In most cases, the choristoma is close to or adjacent to the facial nerve, and any abnormalities in the facial nerve could cause nerve damage. Taking that particular feature of the choristoma into consideration, partial resection is sufficient for diagnosis. Moreover, a nerve integrity monitor (NIM) is useful to avoid facial nerve injury [2]. In this case, we resected the portion that did not react with NIM, which led to a diagnosis without facial palsy.

During a hearing reconstruction, an ossiculoplasty may be needed to improve the hearing level. In cases with stapes deformities, stapes surgery should be considered. When the intraoperative diagnosis is difficult and the risk of recurrence exists, a staged reconstruction procedure appears to be effective [10]. In some cases, the ossiculoplasty may be difficult for the abnormality of ossicles and facial nerve and the hearing results after surgery may be unsatisfactory [2]. Bone conduction implants (BAHA®) could also be considered.

In our case, the mass was involved with the facial nerve. We