CASE REPORT

Salivary Gland Choristoma of the Middle Ear: A Case Report and Review of the Literature

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A choristoma is a benign mass of mature tissue that develops at a site in which such tissue is not usually found. These lesions are a result of defective embryonic development. Choristomas of the salivary gland rarely occur in the middle ear. In this case report, we present a patient with such a tumor and review pertinent studies in the literature. An analysis of the 31 cases (including our account) published thus far showed that salivary gland choristomas of the middle ear are usually unilateral (96.8%) and most often develop in the posterosuperior tympanum (75%). The reported cases were usually associated with an ossicular deformity or erosion (81.3%) or an abnormal intratympanic facial nerve (61.2%). A conductive hearing loss that developed during the patient's childhood was the most common presenting symptom (93.6%). A mass behind an intact tympanic membrane was visible (61.3%) or was, discovered by computed tomography or during exploratory tympanotomy. A definitive diagnosis of this lesion can be based only on the results of a pathologic examination. Although choristomas are benign, they should be excised if the facial nerve will not be jeopardized. Hearing improvement was reported in 6 of 7 (85.7%) patients who underwent ossicular reconstruction.

A choristoma is a mass of mature normal tissue that is located in an area in which such tissue is not typically found. It differs from a hamartoma, which is a local overgrowth of normal mature cells that are indigenous to the site of involvement.[1] Reported choristomas include glial (neural) and salivary gland tissues. Salivary gland choristomas (also called ectopic or heterotopic salivary glands) have been reported in multiple locations in the head and neck,[2] but only rarely in the middle ear. In 1961, the first case of a salivary gland choristoma in the middle ear was reported,[3] and as of June 2006, only 30 cases have been published in the literature.[3-31] One report[8] described a postmortem case in which the patient had bilateral lesions, and another[15] described a choristoma of the salivary gland in the middle ear of 2 patients. A new case is presented in this article.

CASE REPORT

A 32-year-old man presented to our department complaining of a hearing loss that had persisted since childhood in his right ear and an intermittent discharge (the history of which was imprecise) from that ear. He had no other otologic complaints. He was born via an uncomplicated delivery after his mother had had a normal pregnancy. However, 2 of his cousins suffered from a hearing loss of unknown origin. Both tympanic membranes were dull, thinned, and slightly retracted, and the right ear drum had bluish shade but without pulsation. His facial movements were bilaterally symmetrical. The remainder of his ear, nose, and throat examination and an evaluation of his overall health were unremarkable. Pure-tone audiometry demonstrated a moderate degree of conductive hearing loss in the right ear (air-bone gap average 45 dB) and a mild degree of high-frequency sensorineural hearing loss in the left ear. The speech discrimination score for both ears was excellent. Multifrequency tympanometry showed a bilateral normal pressure and low compliance (type As). High-resolution computed tomography of the temporal bone revealed no abnormality. Otosclerosis was set as a provisional diagnosis.

After having provided informed consent, the patient received a general anesthetic and underwent an exploratory tympanotomy performed via an endomeatal approach. A small reddish pedunculated granular mass (2 mm) was noted arising from a dehiscent horizontal portion of the facial canal. The facial nerve was hanging over the oval window from the horizontal portion of the fallopian canal. The long process of the incus and the head of the stapes had been eroded by the mass, but the malleus was normal in appearance. A facial nerve neuroma was suspected, but unfortunately, frozen pathologic examination was not available at that time. After hemostasis had been achieved, a clear cleavage plane was attained, and the mass was completely dissected and was removed without jeopardizing the nerve. A clear view of middle-ear anatomy was achieved, but the stapes was removed during the manipulation. Because of the low position of the facial nerve, ossicular chain reconstruction was difficult but was successfully accomplished. We used fat to obliterate the oval window and applied a Teflon total ossicular reconstruction prosthesis to a piece of cartilage under the tympanic membrane. The patient was satisfied with his hearing postoperatively, and audiologic reevaluation after 12 months revealed an improvement in right ear hearing to the level of a mild conductive hearing loss (air-bone gap average 15 dB). The patient experienced partial facial palsy immediately after surgery, which had progressed to complete paralysis by the next day. A full dose of steroids was given, and the facial palsy improved after 2 weeks to a level of grade II. At the follow-up visit 12 months later, the patient's facial movements were bilaterally symmetrical.

A histopathologic study of the excised mass revealed a punch of connective tissue stroma covered by pseudocolumnar ciliated epithelium. The underlying stroma was occupied by normal salivary gland tissue (Figure 1), a configuration that was consistent with a choristoma of the salivary gland.

Review of the Literature

A salivary gland choristoma of the middle ear is a rare condition. To our knowledge, 31 cases (including that of our patient) have been described in the literature.

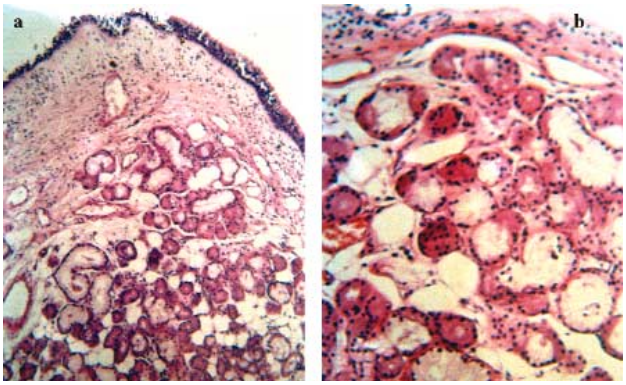

Figure 1: Histopathology of salivary gland choristoma of the middle ear. (a) Low power picture shows a mass composed of mucous and serous glands resembling normal salivary tissue covered with the middle-ear mucosa (H&E; x40). (b) High power picture exhibits detailed glands (H&E; x200).

Tables 1, 2, and 3 summarize the clinical data and the findings reported during middle ear exploration, as well as the outcomes of treatment reported in the literature.

Table 1. Clinical data of 31 patients with a middle ear salivary gland choristoma

Variables	No.	Percent
Sex		
Female	18	58.1
Male	13	41.9
Laterality		
Left	19	61.3
Right	11	35.5
Bilateral	1	3.2
Presentation		
Hearing loss	29	93.6
Tinnitus	9	29.0
Otorrhea 3	9.7	
Facial palsy	1	3.2
Asymptomatic	1	3.2
Type of hearing loss		
Conductive	24	77.4
Mixed	6	19.4
Sensorineural	1	3.2
Degree of conductive hearing loss		
Moderate	12	38.7
Severe	10	32.3
Not reported	9	29.0
Otoscopic examination		
Mass behind tympanic membrane	19	61.3
Dull opaque tympanic membrane	7	22.6
Normal tympanic membrane	4	12.9
Perforated tympanic membrane	1	3.2
Other congenital anomalies		
None	20	64.5
Auricular swelling	3	9.7
Temporal alopecia	2	6.4
Facial asymmetry	2	6.4
Preauricular pits	1	3.2
Conchal bands	1	3.2
Shortened cochlea	1	3.2
Mondini's dysplasia	1	3.2
Contralateral encephalocele	1	3.2
Pharyngeal teratoid polyp	1	3.2
Branchial cyst	1	3.2

Table 2. Findings during 32 middle ear explorations in 31 patients with a salivary gland choristoma of the middle ear*

Variables	No.	Percent
Site of choristoma		
Posterior and/or superior segments	24	75.0
Anterior and/or inferior segments	3	9.4
Entire middle ear	3	9.4
Not reported	2	6.2
Ossicles		
Deformed or eroded	26	81.3
Malleus	8	25.0
Incus	25	78.1
Stapes	22	68.7
Footplate fixation	2	6.2
Normal	4	12.5
Not identified	2	6.2
Intratympanic facial nerve		
Abnormal	20	62.5
Dehiscent	20	62.5
Adherent to the mass	16	50.0
Displaced inferiorly/laterally	7	21.8
Normal	6	18.7
Not identified	6	18.7
Other middle ear anomalies		
Absent cochleariform process	2	6.2
Absent tensor tympani	1	3.1
Absent stapedius tendon	1	3.1
Persistent stapedial artery	1	3.1
Dermoid	1	3.1
Cholesteatoma	2	6.2

* Including 1 postmortem case (a patient with salivary choristoma of both middle ears).

Table 3. Treatment outcomes of 30 patients with a salivary gland choristoma of the middle ear*

Variables	No.	Percent
Approach		
Transmeatal	23	76.7
Postauricular	7	23.3
Mass excision		
Total	16	53.3
Partial	8	26.7
Biopsy	6	20.0
Ossicular reconstruction		
Performed	7	23.3
Hearing improvement	6/7	85.7
No improvement	1/7	14.3
Postoperative facial nerve function		
Not affected	26	86.7
Temporary weakness	3	10.0
Permanent palsy	1	3.3

*Excluding 1 postmortem case.

A review of the literature on patients with a salivary gland choristoma showed that the age at the time of diagnosis ranged from 9 months to 52 years. There was a minor preponderance of women (58.1%) with this type of tumor. Salivary gland choristomas were almost always reported as a unilateral lesion (96.8%), and the left side was most frequently affected (61.3%). Bilateral salivary gland choristomas of the middle ear was found in postmortem evaluation of 1 patient.[8]

Hearing loss (usually a moderate-to-severe conductive or mixed hearing loss) diagnosed during infancy or childhood was the usual presenting symptom (93.6%). One patient was asymptomatic, but a screening audiogram at school had revealed a conductive hearing loss. Tinnitus was a less common symptom (29%). Facial weakness was rare (3.2%), despite high incidence of facial canal dehiscence (61.2%) and adherence to the mass (48.4%).

Other congenital anomalies (temporal alopecia, auricular swelling, conchal bands, facial asymmetry, branchial cyst) were encountered in about one-third (35.5%) of the patients. Two patients had additionally bilateral inner ear abnormality: a shortened cochlea in one[8] and Mondini's dysplasia in the other.[26] It has been suggested that salivary gland choristomas are part of a syndrome[26], but our review does not support that assumption because almost two-thirds of the patients did not demonstrate other ectodermal manifestations.

Otoscopic examination revealed a mass behind an intact tympanic membrane in 61.3% of the patients, and a dull opaque tympanic membrane was found in 22.6%. A mass was accidentally discovered during the insertion of a tympanostomy tube to treat persistent secretory otitis media in 4 patients.

During exploratory tympanotomy, the lesion was found primarily in the posterosuperior tympanum (75%), although salivary gland choristomas have been identified in the anterior and inferior tympanum (9.4%). These masses varied in size and filled the entire middle ear in 9.4% of the patients reported. Our review of the literature also revealed that an ossicular deformity or erosion was identified in the form of an absent or malformed incus (78.1%) or stapes (68.7%); whereas, the malleus was deformed in 25%. The most frequent combination of anomalies noted was a deformed incus and an absent stapes head.

Middle ear surgery was performed via a transmeatal approach (76.7%) or postauricular (23.3%) approach when mastoidectomy was necessary. The mass was removed completely in 53.3% of the patients and was partially excised in 26.7%. A potassium-titanyl-phosphate (KTP) laser was used for uncomplicated mass removal in 1 patient.[27] Ossiculoplasty was performed in 7 patients (23.3%), and 85.7% of those individuals experienced an improvement in hearing. Temporary postoperative facial palsy was noted in 10%, and permanent paralysis occurred in only 1 patient.[9] It has been suggested that intraoperative facial nerve monitoring will improve the outcome of facial nerve function.[20]

Follow-up data (duration, 2-60 months) were available for only 10 patients. Neither regrowth nor malignant transformation was reported.

DISCUSSION

Otologists may identify a mass in the middle ear. The differential diagnosis of such tumors includes congenital cholesteatoma, dermoid cyst, teratoma, glomus tympanicum, granuloma, neuroma, glioma, or choristoma. A salivary choristoma is a benign, slowly growing mass that usually causes a progressive unilateral conductive hearing loss that begins during childhood. Otoscopic examination may reveal a mass behind an intact tympanic membrane. A high-resolution computed tomographic scan may be helpful in revealing the nature of the mass in the middle ear and in detecting associated inner ear anomalies. The lesion may accompany other branchial arch anomalies.

A choristoma can be diagnosed only from the results of a histologic examination performed during exploration of the middle ear. Macroscopically, these tumors appear lobulated, white, grey, or reddish-brown. They are firm and not vascular. Each mass is attached to the middle ear via a broad base or a fine stalk. Microscopically, the lesion consists of a mixture of

well-formed serous and mucous acini arranged randomly or in a lobular formation and interspersed with fibroadipose tissue. Salivary ducts may or may not be present. The covering epithelium is usually a pseudostratified columnar ciliated epithelium or (rarely) a squamous epithelium.

The pathogenesis of a salivary gland choristoma of the middle ear is believed to involve an aberration in embryologic development.[26] It is well known that the malleus and the incus, except for the long process, are derived from the mesenchyme of the first branchial arch (Meckel's cartilage), but both the long process of the incus and the stapes are derived from the mesenchyme of the second branchial arch (Reichert's cartilage). The horizontal portion of the facial canal is also considered to be closely related to the development of the second branchial arch, which suggests that an error in the development of the second branchial arch results in an abnormality of the ossicular chain and facial nerve.[24] The parotid gland, which becomes evident during the eighth week of intrauterine life, grows toward the ear and then sends out a primordial cord of cells to form the parotid gland. A salivary gland choristoma in the middle ear may develop as a result of the extension of an epithelial remnant of the parotid cords into the middle ear or from the aberrant extension of the pharyngeal endoderm inside the middle ear. Choristomal tissue becomes trapped during the fusion of portions of the temporal bone. It may disturb the development of the second branchial arch, which in turn causes an ossicular chain and facial nerve abnormality.[16] A choristoma may also disturb the development of the auricular tubercles and second branchial arch at the same time, and this hypothesis may explain the development of several kinds of facial nerve or ectodermal anomalies.[7]

The incidence of facial canal dehiscence in the literature varies according to diagnostic criteria and the inclusion of fetal specimens. However, a recent study reported fallopian canal dehiscence in 19.7% of 300 temporal bones, but intraoperative dehiscence was noted in only 6.4% of 357 otologic procedures.[32] These data contrast to the higher incidence of dehiscence found in patients with choristoma as it is evident in our review.

Treatment is determined by the size and location of the tumor. Conservative management and serial examinations have been recommended by most authors, because there is usually no potential for further growth or malignant change. However, salivary tumors (benign mixed salivary tumor,[33] adenoma,[34] or adenocarcinoma[35]) have been reported to occur in the middle ear. These tumors possibly originated in a choristomatous salivary tissue. Small choristomas or those attached only by a thin stalk can be readily excised. Large or broad-based tumors must be removed cautiously, depending on their relation to the intratympanic facial nerve, the availability of intraoperative nerve monitors, and the competence of the surgeon. Laser excision, if available, may be helpful in dissection. Ossicular reconstruction can be accomplished and usually improves hearing.

CONCLUSIONS

Although a salivary gland choristoma of the middle ear is a rare lesion, it should be included in the differential diagnosis of middle ear masses and unilateral conductive hearing loss. Surgical excision followed by ossicular chain reconstruction may be feasible without causing permanent damage to the facial nerve.

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