

CASE REPORT

Coexistence of congenital cholesteatoma and non-neoplastic neuroectodermal remnants in the temporal bone: Case report

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The most common location of congenital cholesteatoma, diagnosed these days at an increasing rate, is the anterosuperior quadrant of the tympanic cavity. Mastoid origin is rare, and neuroectodermal tissue remnant in the middle ear has not been reported to date. We discuss a patient whose congenital cholesteatoma was confined to the mastoid and coincided with the occurrence of non-neoplastic neuroectodermal remnant in the middle ear. The neuroectodermal remnant was accompanied by a blue tympanic membrane. The congenital cholesteatoma was asymptomatic at the time of initial diagnosis. Radiologic and clinical features of this very rare coincidence are discussed. This coexistence may help to support embryonic-remnant theory in congenital cholesteatoma formation.

Congenital cholesteatoma of the temporal bone develops as a whitish mass behind an intact tympanic membrane. It is believed that it grows from embryogenic epithelial remnants in the temporal bone. In recent years, diagnoses of congenital cholesteatoma has been increasing. Symptoms and signs relate to extension and location of the lesion. Patients are usually asymptomatic but may have hearing loss, vertigo, or facial palsy.^[1] The most common location of congenital cholesteatoma is the anterosuperior quadrant of the tympanic cavity. The mastoid cavity is only rarely affected.

Levenson and colleagues have proposed 6 requirements for the diagnosis of congenital cholesteatoma: 1) a whitish mass, medial to a normal, intact tympanic membrane; 2) normal pars flaccida and pars tensa; 3) no history of otorrhea or perforation; 4) no prior otologic procedures (including parasyntesis); 5) exclusion of canal atresia and intramembranous and giant cholesteatoma; and 6) no exclusion due to prior bouts of otitis media.^[2]

There are many theories that explain the pathogenesis of congenital cholesteatoma. These lesions may be epidermoid formations, the result of middle ear mucosal metaplasia, defective development of the tympanic ring, or viable amniotic cellular contents within the middle ear.^[3]

We report on a patient with non-neoplastic neuroectodermal remnant in the middle ear that coincides with congenital cholesteatoma in the mastoid bone.

CASE REPORT

A 34-year-old man presented with hearing loss in the left ear of 15 years' duration and dizziness after effort lasting a few minutes for the past 5 years. There was no history of ear infection, discharge, trauma, or surgery. Otomicroscopic examination revealed a bluish, dull tympanic membrane in the left ear and a normal one in the right.

Audiometric examination demonstrated a U-shaped moderate conductive hearing loss with a tendency toward very slightly depressed bone conduction (BC) threshold on the left (average 4-tone air conduction [AC] threshold: 42 dB; average 4-tone BC threshold: 27 dB). Tympanometry showed low compliance (0.21 mL)

with normal middle ear pressure. Stapedius reflexes were absent on the left. Hearing acuity and tympanometry were normal on the right.

Computed tomography (CT) and magnetic resonance imaging (MRI) of the temporal bone were performed for further evaluation. CT scans showed soft tissue confined to the mastoid and located mostly lateral and slightly medial to Korner's septum on the left (Figure 1). Lateral semicircular canal fistulization was also detected (Figure 2). Minor destruction of the mastoid antrum was noted.

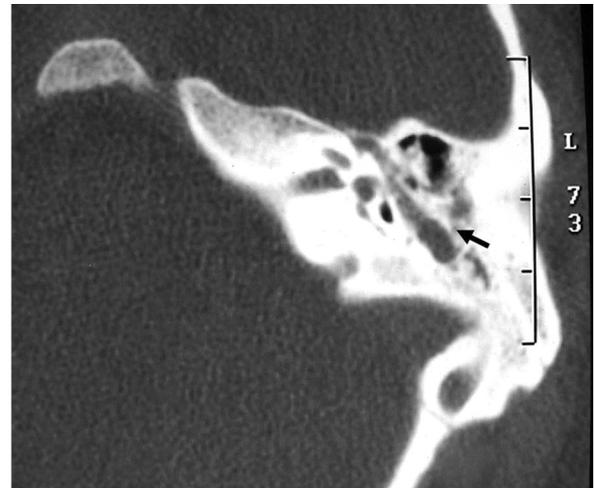


Figure-1: Axial computed tomography scan showing Korner's septum (black arrow) and soft tissue in the mastoid.

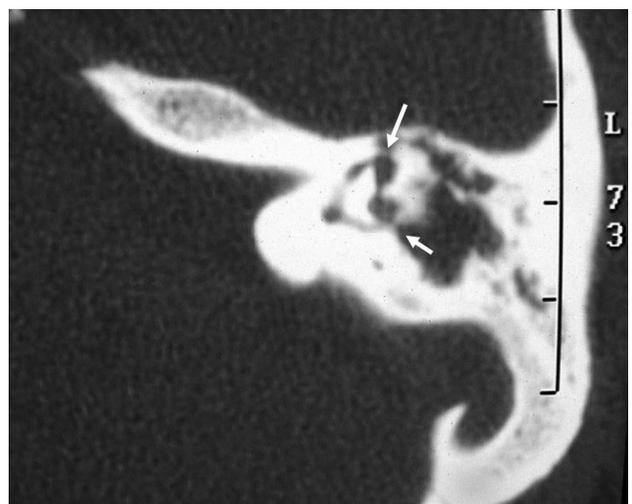


Figure-2: Lateral semicircular canal fistula on axial view of computed tomography scan (white arrows). Also shown is soft tissue erosion of the mastoid bone.

MRI showed 2 lesions with slightly different characteristics. T1-weighted scans revealed an isointense cholesteatoma and a slightly hyperintense neuroectodermal remnant that intersected by Korner's septum (Figure 3). T2-weighted images showed a difference in hyperintensity. The cholesteatoma had homogeneous hyperintensity; the neuroectodermal remnant was more heterogeneous, especially in the posterior portion (Figure 4). There was subtle peripheral enhancement around the cholesteatoma in gadolinium-enhanced scans.

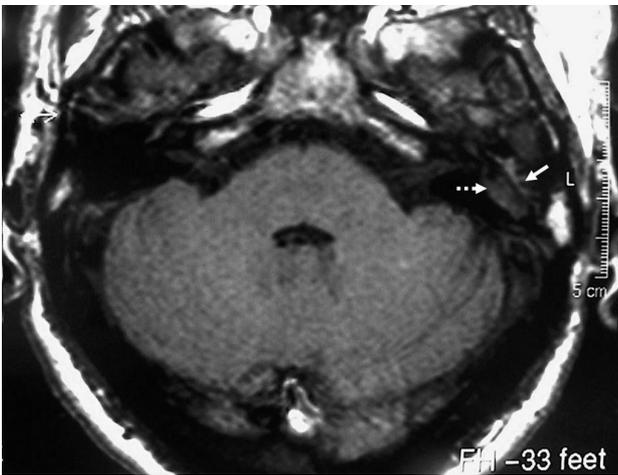


Figure-3: T1-weighted unenhanced axial image shows a soft tissue mass (cholesteatoma) isointense to the brain (dashed white arrow) at the medial side of Korner's septum and another lesion (neuroectodermal remnant) with a signal intensity slightly hyperintense to the brain (white arrow) on the lateral side of the septum.

An exploration of the mastoid and middle ear was planned. Korner's septum was reached during initial drilling, and the cholesteatoma was located mostly posterior to Korner's septum. Superior extension toward the middle fossa dura and slightly anterior extension to the antrum were also detected. Posterior extension of the cholesteatoma had eroded the lateral semicircular canal, and a fistula was found. Small, brownish, soft tissue with granulation was seen anterior to Korner's septum. After removal of this lesion, we noted that this nonbulky lesion scattered superficially throughout the middle ear mucosa. The cholesteatoma and granulated tissue were completely removed and sent for histopathologic examination. The fistula was sealed with a temporal

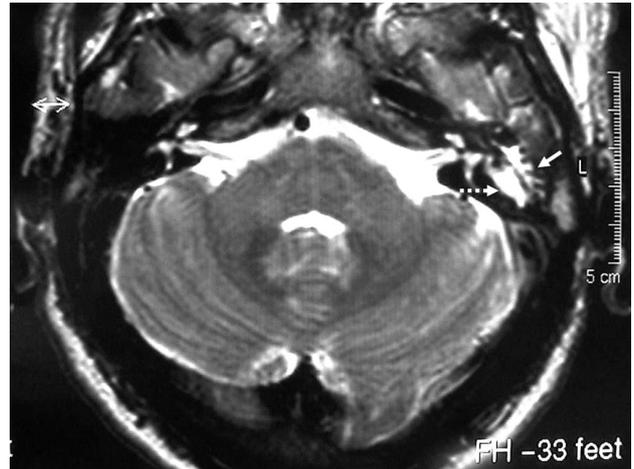


Figure-4: T2-weighted axial magnetic resonance image. The mass at the medial side of Korner's septum (dashed white arrow) is hyperintense (cholesteatoma), whereas the mass at the lateral side is slightly heterogeneously hyperintense (neuroectodermal remnant).

fascia. The brownish soft tissue was reported as connective tissue within clusters of non-neoplastic neuroectodermal cells. Diagnosis of cholesteatoma was also confirmed histopathologically (Figure 5).

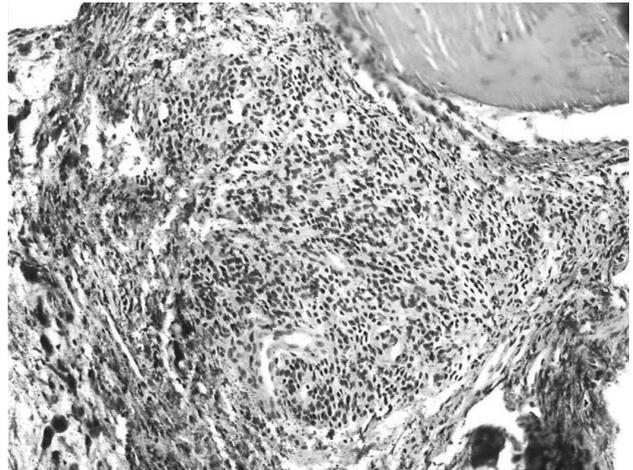


Figure-5: Neuroectodermal cell remnants were seen in a connective tissue fragment.

Mixed-type hearing loss persisted after surgery (AC: 52 dB, BC: 30 dB). Seventy-eight percent caloric weakness was detected on the left. The patient was followed up in 3-month intervals. At 1-year postoperation, even though the otomicroscopic view

seemed normal, temporal bone CT scan and MRI showed that cholesteatoma had recurred. Canal wall down (CWD) mastoidectomy and labyrinthectomy was planned. During the operation, we noticed that the posterior part of the otic capsule had eroded. Complete labyrinthectomy was performed in conjunction with CWD mastoidectomy. Postoperative follow-up was uneventful.

DISCUSSION

Congenital cholesteatoma usually occurs in the anterior-superior quadrant of the tympanic annulus and adjacent to the opening of the eustachian tube.^[1] Potts and colleagues have proposed 4 stages of localization and extension of the congenital cholesteatoma.^[1] Stage I is characterized by single-quadrant involvement, with no ossicular involvement or mastoid extension (40% of patients). Stage II features involvement of multiple quadrants but not of the ossicles and mastoid (14% of patients). Ossicular erosion with no mastoid extension occurs in stage III (23% of patients). Mastoid extension occurs in stage IV (23% of patients). Higher stages always correlate with the probability of residual disease and worsening of hearing postoperatively.

Congenital cholesteatoma with mastoid origin is very rare. We observed a cholesteatoma at Korner's septum, extending posteroinferiorly. A labyrinthine fistula visualized by CT was confirmed intraoperatively. At that time, we thought that the cholesteatoma was completely removed. Additional sensory hearing loss was not observed postoperatively.

Michaels observed epidermoid remnants localized at the anterior-superior position near the tympanic annulus and adjacent to the opening of the eustachian tube in fetuses. The remnants typically disappear after 33 weeks' gestation. However, epidermoid formation is believed to persist in some cases, and this provides a nidus for cholesteatoma formation.^[4] Levine and associates studied 76 fetal specimens 20 to 40 weeks' gestation and 30 full-term neonates and children up to 5 year olds. Ten percent of 44 specimens in the 20-to-33-week gestational age range were found to have

epidermoid formation vs 12.7% of specimens after 33-week gestational age. In all cases, epidermoid formation was located at the anterior-superior position near the tympanic annulus adjacent to the opening of the eustachian tube.^[3] This finding seems to explain the high incidence of anterior-superior location of congenital cholesteatoma.

We made another interesting finding in our patient, in addition to the rare mastoid location of his congenital cholesteatoma. Non-neoplastic neuroectodermal cell clusters in the connective tissue were scattered superficially throughout the mucosa of the middle ear and mastoid antrum. Concurrence of neuroectodermal remnants and congenital cholesteatoma is considered rare even though cholesteatoma originates from the ectoderm. This coexistence seems to support the embryonic-remnant theory in congenital cholesteatoma formation. Passing amniotic cellular content into the middle ear via eustachian tube at parturition has been reported in congenital cholesteatoma formation.^[5-7] However, this likelihood seems to be very low in our patient because of the coexistence of neuroectodermal remnant.

Ossicular chain is normal in about 20% of patients with congenital cholesteatoma.^[8] Our patient had an ossicular chain surrounded by granulated tissue causing blockage of the aditus ad antrum. The congenital cholesteatoma did pass anteriorly beyond Korner's septum. The granulated tissue was removed at the mastoid and middle ear; however, this did not improve the conductive component of hearing loss as much as expected.

Based on otomicroscopic examination showing a bluish tympanic membrane, we initially suspected cholesterol granuloma. MRI characteristics of the neuroectodermal remnants were also similar to those of cholesterol granuloma. Cholesterol granuloma appears hyperintense on T1- and T2-weighted MRIs and may contain heterogeneous areas through septation.^[8] The lateral portion of the lesion was slightly hyperintense on T1-weighted MRI and slightly heterogeneously hyperintense on T2-weighted MRI. The medial portion of the lesion was isointense on T1-weighted MRI and

hyperintense on T2-weighted MRI. Cholesteatomas demonstrate long T1 and T2 characteristics on MRI. Therefore, they appear as hypointense on T1-weighted and hyperintense on T2-weighted magnetic resonance images.¹⁹¹ In our patient, the medial portion was isointense on T1-weighted MRI. We surmise that this may arise from infiltration of cholesteatoma by granulated tissue.

Non-neoplastic neuroectodermal remnant in the temporal bone has not been reported in the literature to the best of our knowledge. This lesion must be considered one of the causes of blue tympanic membrane, or blue ear.

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