

# CONGENITAL CHOLESTEATOMA: A CASE REPORT

## KONJENİTAL KOLESTEATOM: VAKA SUNUMU

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**SUMMARY:** Congenital cholesteatoma is a rare entity characterized by a white mass behind an intact tympanic membrane. It arises from ectopic epidermoid remnants in the middle ear. In this article, a case of congenital cholesteatoma is discussed with regard to the presentation, localization and treatment of the disease.

**Key Words:** Congenital Cholesteatoma, Middle Ear, Cholesteatoma.

### INTRODUCTION

Congenital cholesteatoma is a rare entity characterized by a whitish globular mass behind an intact tympanic membrane. Congenital or primary cholesteatoma is far less common than its acquired counterpart. It arises from ectopic epidermoid remnants in the middle ear (1,2). Two typical sites of origin are described: the anterior epitympanum, in the proximity of the Eustachian tube opening, and the posterior mesotympanum, close to the incudostapedial joint. Progressive conductive hearing loss may be the patient's only complaint. In almost all patients, there is no history of chronic otitis media. This is in contrast to acquired cholesteatoma of the middle ear, which is characterized by a history of chronic infection and aural discharge. In congenital lesions, the eardrum is preserved, and mastoid pneumatization is normal (1).

In this article, a case of congenital

**ÖZET:** Konjenital kolesteatom sağlam kulak zarı arkasında beyaz kitle ile karakterize nadir görülen bir kulak hastalığıdır. Orta kulakta yer alan epidermoid kalıntılardan kaynaklandıkları düşünülmektedir. Bu yazıda konjenital kolesteatomlu bir vaka sunularak hastalığın prezentasyonu, lokalizasyonu ve tedavisi tartışılmıştır.

**Anahtar Kelimeler:** Konjenital Kolesteatoma, Orta Kulak.

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### CASE REPORT

A 24-year-old female patient presented with a one-year history of hearing impairment on the right side. An otoscopic examination evidenced a whitish mass behind an intact tympanic membrane in the postero-superior quadrant. The light reflex and the malleus handle were not visible (Fig. 1). There was no history of previous ear surgery, trauma, or otitis media. Pure tone audiometry revealed an air-bone gap of about 43 dB on the right side (Fig. 2). Speech discrimination scores were normal on both sides. A computed tomography scan of the temporal bone showed a mass in the mesotympanum behind the eardrum, extending to the mastoid antrum. The ossicles could not be identified and the temporal bone was pneumatized but not as

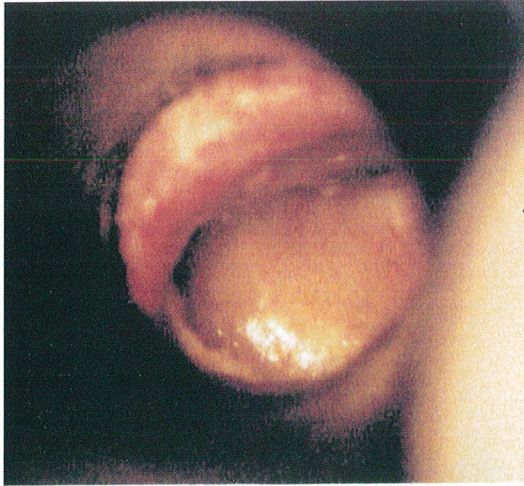


Fig. 1: A whitish mass behind an intact tympanic membrane in the postero-superior quadrant (otomicroscopic appearance).

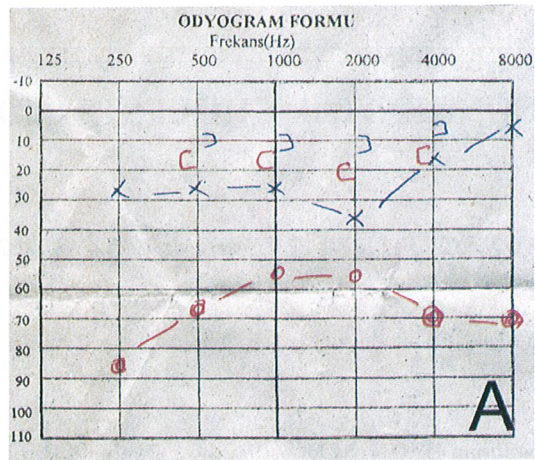


Fig. 2: The air-bone gap of about 43 dB on the right side in preoperative pure tone audiometry.

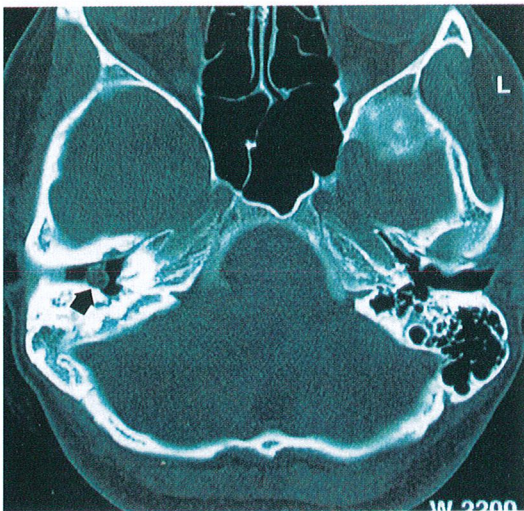


Fig. 3: Computed tomography scan of the temporal bone shows a mass in the mesotympanum behind the eardrum (black arrow). The ossicles cannot be identified and the temporal bone is pneumatized but not as well as the contralateral normal side.

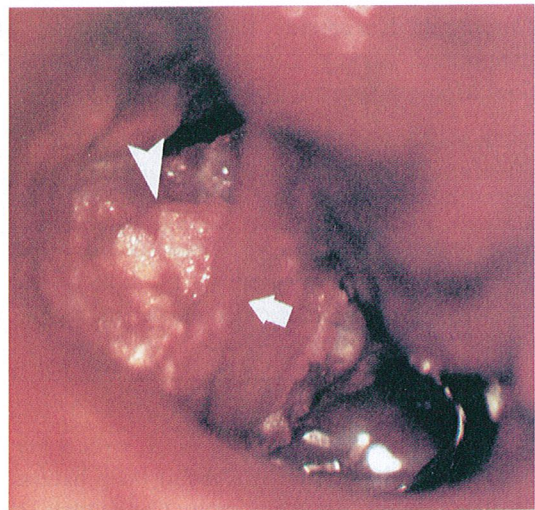


Fig. 4: The appearance of the white keratinized mass in the mesotympanum (white arrowhead), which is typical for cholesteatoma. White arrow: Chorda tympani.

well as the contralateral normal side (Fig. 3). A presumptive diagnosis of congenital cholesteatoma on the right side was made and a retroauricular exploratory tympanotomy was planned. The appearance of the white keratinized mass in the mesotympanum was typical for cholesteatoma (Fig. 4). The mass had eroded the malleus, incus and stapes suprastructure and extended from the posterior mesotympanum to the attic, filling the facial recess and the sinus tympani. An antrotomy was performed to assess the mastoid involvement of the cholesteatoma mass and it was found to extend up to the posterior border of the lateral semicircular canal

(Fig. 5). In order to ensure complete removal from the anterior attic and facial recess regions a canal wall down procedure including a wide meatoplasty was carried out. We performed the ossicular reconstruction using a bone columella. The columella was sculptured from the mastoid cortical bone and was transposed between the tympanic membrane and the stapes base. The microscopic examination of the biopsy material from the mass confirmed the preoperative diagnosis.

After 9 months of follow up the patient did not have otorrhea or other cavity-related

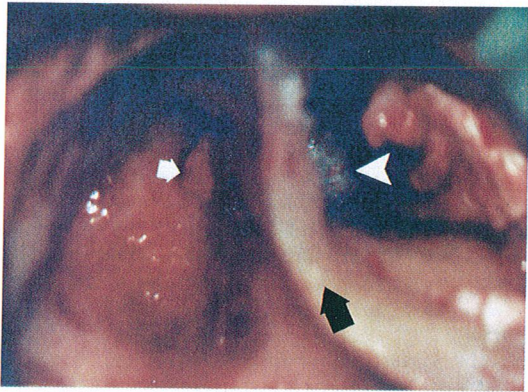


Fig. 5: The mass in the antrum and middle ear. White arrow: Cholesteatoma sac in the antrum. Black arrow: Posterior ear canal wall. White arrowhead: Cholesteatoma in the mesotympanum.

problems. The postoperative pure tone audiometry showed that the air-bone gap had decreased to 20 dB (Fig. 6).

#### DISCUSSION

Cholesteatomas are described as congenital if they present behind an intact tympanic membrane without a previous history of infection or trauma (3). The criterion of primary (congenital) aural cholesteatoma was defined by Derlacki and Clemis (4) and modified by Levenson et al. (5), noting development of cholesteatoma behind an intact tympanic membrane (normal pars flaccida and pars tensa), absence of a previous history of aural infections, no demonstrable connection between the cholesteatoma and the external auditory canal, and no history of prior otologic procedures (6).

The majority of congenital cholesteatomas are asymptomatic and appear to be innocuous keratin pearls. However, they can enlarge and cause complications ranging from ossicular destruction to facial paralysis (3). Two typical sites of origin are described: the anterior epitympanum, in the proximity of the Eustachian tube opening, and the posterior mesotympanum, close to the incudostapedial joint (1). The symptomatology in patients with congenital cholesteatoma is generally related to the direction of its extension. In patients with posterior mesotympanum lesions conductive deafness due to the earlier erosion of the ossicles is anticipated

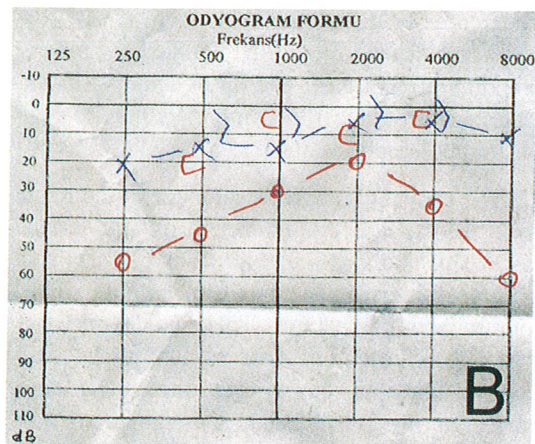


Fig. 6: The postoperative pure tone audiometry shows the air-bone gap has decreased to 20 dB.

(7). In our patient, all of the ossicles were totally eroded by the lesion, which had originated in the posterior mesotympanum. The location of the cholesteatoma in the postero-superior quadrant may have delayed the otoscopic diagnosis, because of the greater opacity of the tympanic membrane in this region, as suggested by Curtis (8). This feature partially explains why the diagnosis was delayed until adulthood in our patient.

In acquired cholesteatoma, the involved mastoid is typically hypopneumatic due to the effects of long-standing chronic otitis media, whereas in congenital disease the mastoid usually possesses normal pneumatization, as in our case (9).

In adults, as in children, the main goal of cholesteatoma surgery remains total removal of the lesion, but restoration of socially acceptable hearing is also important for the quality of the patient's life (10). Three different surgical techniques are generally used for removal of disease limited to the middle ear and mastoid. The first is the closed technique (CT), i.e. the intact canal wall technique, in which the cholesteatoma is removed using a combined transcanal-transmastoid approach. The two others are open techniques (OT), i.e. canal wall down techniques, and shared sacrifice of the posterior wall of the bony canal. One of these is radical mastoidectomy (RM) in which no

rehabilitation of the middle ear cleft is included. In modified radical mastoidectomy (MRM; tympanoplasty in an open technique) the RM is followed by rebuilding of the middle ear, using a temporalis fascia graft and an ossiculoplasty. In some instances, a partial temporalis muscle flap mastoid obliteration can be associated (10).

The choice of surgical technique is the main issue. Most authors prefer CT in appropriate cases. This is in fact a functional choice, adapted to the sociologic and economic background of the country and the patient. This choice must be associated with a very rigorous quality of lesion removal, and with a policy of a systematic second look. On the other hand, the choice of CT necessitates the patient's and the family's collaboration and their adhesion to a fastidious surgical program. We preferred MRM to ensure complete removal of the lesion with control in the facial recess and anterior attic regions. Furthermore, a second look operation was not suitable for our patient with the risk of an unsafe ear in case she was lost to follow up.

If perfectly done and associated with ossicular reconstruction, MRM is not a simpler technique to perform than CT. Instability and inflammation of the cavities secondary to the open technique may necessitate complementary surgical treatments (11). Troublesome infection can be a problem with open cavities and often appears to be related to areas in the cavity in which the lining consists of wet mucosa rather than a dry epithelium (11). In our case we did not encounter any cavity-related problems, with the air-bone gap lowered to 20 dB.

Application of the suitable technique combined with a careful follow-up period should result in the appropriate treatment of this dreadful disease.

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