



Case report

Atypical presentation of congenital cholesteatoma in an adult case with good hearing result[☆]Fevzi Solmaz, Davut Akduman^{*}, Mehmet Haksever¹, Ercan Gündoğdu¹, Atilla Mescioğlu¹

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ABSTRACT

Introduction: Congenital cholesteatoma is thought to be caused by inadequate folding of the epidermoid formation inside the middle ear cleft. During development of the middle ear mucosa, stratified squamous epithelium accumulates in the embryonic life. Its typical appearance is a “pearl” beneath the anterosuperior quadrant of the tympanic membrane.

Presentation of case: We report 28 years-old case with congenital cholesteatoma in the posterosuperior quadrant of middle ear cavity. The main complaint was the hearing loss which had developed slowly over several years.

Discussion: The case was surgically treated. Postoperative hearing result was satisfactory.

Conclusion: Congenital cholesteatoma may occur in atypical locations and ages. Many authors prefer canal wall down tympanomastoidectomy. But it can also be treated successfully by intact canal wall tympanomastoidectomy with good hearing results.

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1. Introduction

Congenital cholesteatoma (CC) is thought to be caused by inadequate folding of the epidermoid formation inside the middle ear cleft. During development of the middle ear mucosa, stratified squamous epithelium accumulates through the 3rd and 5th weeks of embryonic life. Squamous cells remained in the middle ear cavity from this period grows slowly over time, gives rise to cholesteatoma formation mostly in childhood [1]. Its typical appearance is a “pearl” beneath the anterosuperior quadrant of the tympanic membrane. Conductive hearing loss caused by congenital cholesteatoma is known to have poor prognosis [2].

Although there is mostly intradural involvement, 20% of cases present with extradural involvement. The main sites of extradural involvement are middle ear cavity, external meatus, mastoid, and squamous and petrous portions of the temporal bone [1,3]. It is either asymptomatic or presents with conductive hearing loss. Otalgia, vertigo and facial paralysis may be seen in advanced cases. There may be disturbance of taste due to chorda tympani

involvement [1]. Other criterias to diagnose CC are; the case should not have the history of otorrhea, trauma, or ear surgery; and the tympanic membrane must be intact. On computerized tomography (CT), it has a non-isolated, hypodense appearance indicative of an invasive lesion. Treatment is surgical excision.

Potsic et al. classified congenital cholesteatoma of the middle ear in 4 stages. Stage I denotes disease limited to only one quadrant. Stage II means disease involving multiple quadrants without involvement of the ossicles or mastoid. Stage III represents ossicular involvement with no disease in the mastoid. Disease occupying the mastoid is stage IV [4].

2. Case report

A 28 years old male patient presented with hearing loss and tinnitus, which had advanced over a few years with acceleration within one month. His history revealed no otorrhea or ear surgery. On otoscopy, appearance of the tympanic membrane was normal. The facial nerve functions were intact. On pure-tone audiometry (PTA), air/bone mean conduction thresholds were 18/18 dB and 57/19 dB for the right and left ears respectively; and Weber was lateralized to the left. Both ears had type-B tympanograms. Compliance of the left ear was lower than that of the right ear (0.05 mL and 0.63 mL respectively).

On temporal CT, the left ear cavity had a soft tissue density neighboring the tympanic segment of the facial canal inferior to the

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epitympanum, which was compatible with cholesteatoma (Fig. 1). Additionally, an appearance of the preoperative tympanic membrane is shown in Fig. 1. On exploration of the tympanic cavity with general anesthesia, a pearl-colored, fragile cholesteatoma was detected just medial to the scutum, involving the lenticular process of the incus (Fig. 2). The cholesteatoma was found to extend to the facial recess and sinus tympani, and erode the long process of the incus and suprastructure of the stapes. The cholesteatoma was totally removed, and body of the incus was reshaped and placed over the oval window base for columellar effect (Fig. 2). An underlay temporalis fascia graft was then replaced.

No intra- or postoperative complication was seen in the patient. The diagnosis of cholesteatoma was confirmed histologically. The pure-tone average thresholds obtained at the 6th week of the operation were 20/12 dB (air/bone thresholds respectively).

On examination of the ear 9 years after the operation, a small perforation was detected on posterosuperior quadrant of the tympanic membrane. The PTAs (air/bone) were 30/8 dB. The decrease in air conduction was thought to have resulted from this perforation on the tympanic membrane. On temporal CT obtained at the same time showed that the left mastoid and tympanic cavity were aerated normally and the ossicular chain was intact. There was no finding indicative of recurrent cholesteatoma (Fig. 3). Also endoscopic examination was made and demonstrated the perforation on the posterosuperior quadrant (Fig. 3). Periodical ENT examination was recommended to the patient, taken into account hearing status and CT result of the patient.

3. Discussion

Congenital cholesteatoma (CC) is derived from congenital epithelial remnants in the temporal bone and can manifest in any age from infancy to adulthood (mean 16.7 years of age) [5]. Its histopathological appearance is similar to that of acquired cholesteatoma; however, presence of some properties in the patient such as an intact tympanic membrane and no history of ear disease or surgery help direct the diagnosis towards CC [6].

CC is typically seen as a pearly white mass behind the tympanic membrane [7], mostly within the anterosuperior quadrant. CC involving the posterior tympanum inevitably results in conductive hearing loss [8]. In the adults and elderly, the disease may involve an unusual localization and conductive hearing loss is the most common symptom in these patients.

If diagnosed late, CC may cause more serious complications such as labyrinthine fistula, facial paralysis, meningitis, cranial abscess, and even death [9].

Congenital cholesteatoma is a disease of mainly the childhood period. In one large retrospective series, 73% of patients operated on during a time period of 24 years were 15 years old or younger [10]. As a general rule, the older the patient is at the time of diagnosis, the more advanced is the disease stage [11]. Our case was 35 years old at the time of diagnosis and surgery.

Evidence suggests that outcomes of cholesteatoma surgery are similar in children and adults [12]. In our case, the early postoperative air-bone gap was as small as 8 dB. This hearing result can be considered as successful. Although an increased air-bone gap of

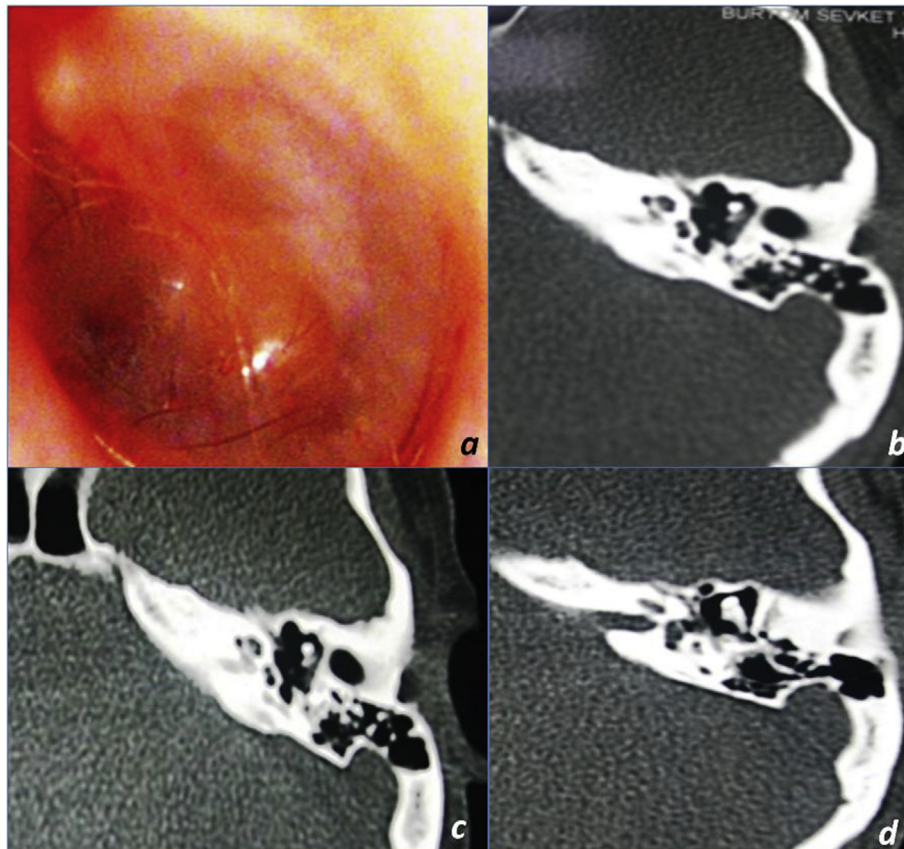


Fig. 1. The Preoperative view of Tympanic Membrane and CT appearance *Red color appearance is Cholesteatoma. **Defeated ossicles are showed the dashed lines in cholesteatoma.

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