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A congenital tympanic membrane cholesteatoma (CTMC) in the adult: a case report

M. CASALE*, A. DE FRANCO*, M. NEGRI**, F. PIAZZA°, A. INCAMMISA*, F. SALVINELLI*, C. ZINI*

*Area of Otolaryngology, Interdisciplinary Center for Biomedical Research (CIR), University Campus Bio-Medico, Rome (Italy)
**Otorhinolaryngology Operative Unit, Ramazzini Hospital, Carpi (Italy)
°Otorhinolaryngology Operative Unit, Carlo Poma Hospital, Mantova (Italy)

Abstract. – *Objective:* **We present an additional very rare case of a congenital tympanic membrane cholesteatoma (CTMC) in the adult.**

Method: **Case report and literature review of CTMC.**

Case Report: **A 54-year old man was referred to us by his primary care physician who noted a white mass on the right tympanic membrane without prior history of otorrhea, tympanic perforations or previous otologic procedures. The pearl was about 5 mm diameter, centered on the umbo of a normal tympanic membrane (TM). The audiogram and the tympanogram was absolutely normal. CT confirmed a soft round shape tissue mass, located in the centre of the TM near umbo. The mass protruded both in the auditory canal and in the middle ear space, touching the malleus extremity, without any relationship with medial wall of the cavum tympani. A surgical excision was performed using a "minimal" retroauricolar transcanalar approach: the CTMC was located into the thickness of the TM, between epidermic and mucous layers. The ossicular chain was preserved intact. A partial myringoplasty (underlay technique) using a temporalis fascia graft was necessary. Histopathology confirmed a cystic cholesteatoma. After two months and one year follow-up, otoendoscopy showed a well-healed TM with a preserved normal audiogram and tympanogram.**

Discussion: **This exceptional (probably the first reported) case showed the possible localization of the CC in the TM, also in the adult. Criteria for classification of a TM cholesteatoma as congenital and possible pathogenetic mechanisms are discussed.**

Key Words:

Congenital cholesteatoma, Tympanic membrane, Tympanoplasty.

Introduction

The literature has many reports on the congenital middle ear cholesteatomas[1], but very few reports exist on the congenital tympanic membrane cholesteatoma (CTMC). All cases were observed in childhood[2-5]. We report a very exceptional case of CTMC observed in an adult man.

Case Report

A 54-year old man was referred to our Out-Patient clinic in October 2003 by his primary care physician who noted a white mass on the right tympanic membrane. No prior history of otorrhea, tympanic perforation, previous trauma or otologic procedures.

The pearl was about 5 mm diameter, centered close the umbo of a normal tympanic membrane (Figure 1). Audiogram and tympanogram were normal.

Coronal and axial CT scan showing soft mass protruded both in the auditory canal and in the middle ear space, touching the malleus extremity, without any relationship with medial wall of the cavum tympani (Figure 2A, B).

Surgical excision was performed in November 2003 using a "minimal retroauricolar transcanal approach". The cyst was extracted "en bloc". It was located between the epidermic and the mucous layers. No cholesteatoma was found in the cavum tympani, except a minimal free epidermoid material in the hypotympanum, leaking from a little laceration of the cyst medial wall. The ossicular chain was intact and the mucosa of the middle ear was absolutely normal. Hystopatholo1gy confirmed a cystic cholesteatoma, located between the epidermic and the mucous layers.

At two months and two years follow-up otoendoscopy showed a well-healed tympanic membrane with a preserved normal audiogram and tympanogram.

Discussion

This very exceptional (probably the first reported) case of tympanic membrane cholesteatoma

Corresponding Author: Manuele Casale, MD, PhD; e-mail: m.casale@unicampus.it

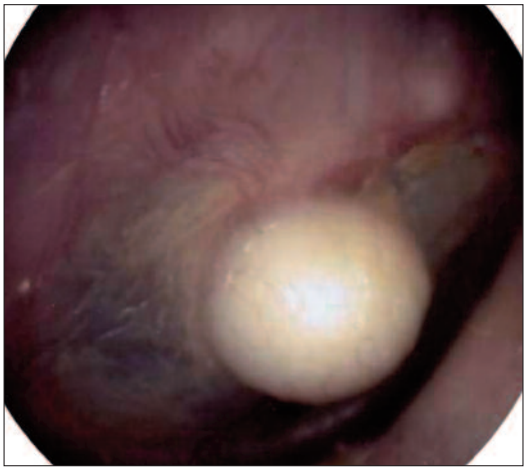

Figure 1. Pre-operative otoendoscopy showed a 5 mm diameter pearl centered close the umbo of a normal tympanic membrane.

(TMC) in adult patient inserted into absolutely normal pars tensa between the epidermic and the mucous layers, without any pathology of the middle ear mucosa, classified as congenital.

The criteria for classification of a tympanic membrane cholesteatoma as congenital were:

- White mass inserted into a normal pars tensa, without connexion with pars flaccida and/or promontorium
- Normal pars flaccida
- No prior history of otorrhea or tympanic membrane perforation
- No previous otologic procedures or trauma

This CTMC could derived from the development of ectodermic cells remained included during embriogenesis, according with the Sanna-Zini's hypothesis[6]. His location, close to the ectodermic and mesodermic tissue borderline, seems to confirm also the Aimi's pathogenetic hypothesis that congenital cholesteatoma is the result of a development mistake caused by the failure of the natural stop signal by mesodermic tympanic ring to ectodermal tissue migrating from external auditory canal[7]. For the surgical technique, we performed a "minimal retroauricolar transcanal approach", instead of a simple endocanalar approach, in order to perform a more safe complete excision of the cyst and a better middle ear exploration.

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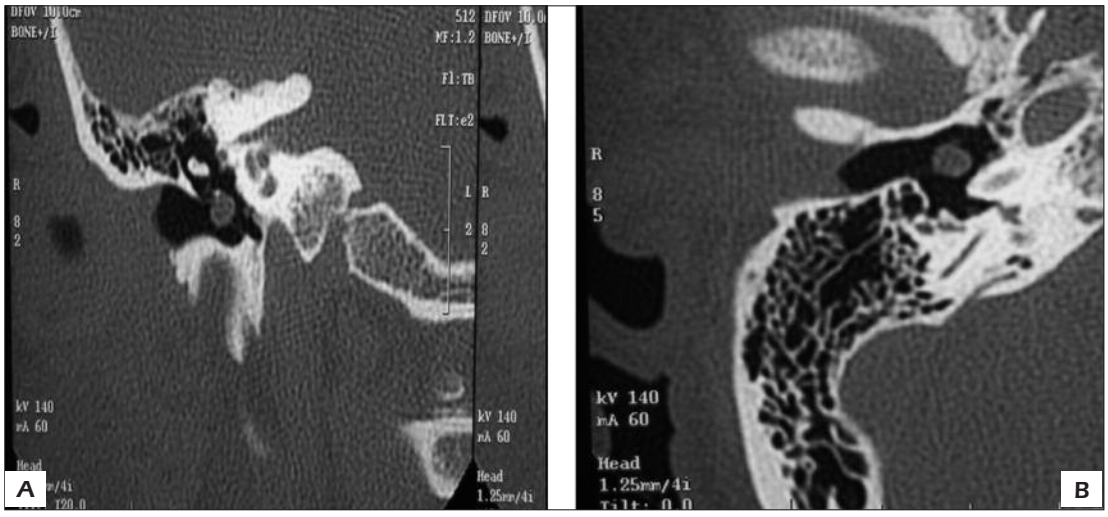

Figure 2. Coronal *(A)* and axial *(B)* CT scan showing soft mass protruded both in the auditory canal and in the middle ear space.