

CASE REPORT

Case report of congenital cholesteatoma in the oval window and posterior mesotympanum surrounding a dehiscence fallopian canal in a young adult

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ABSTRACT

Congenital cholesteatoma is defined as cholesteatoma occurring behind an intact tympanic membrane (TM). In the middle ear, it has different stages according to its site and relation with surrounding structures. We report an unusual case which is congenital cholesteatoma in the oval window and posterior mesotympanum surrounding a dehiscence fallopian canal in a young adult without obvious lesion behind intact TM, discussing its management. We have focused light in an unusual case in our practice and concluded that early intervention in such cases is recommended to prevent complications and planning a staged surgery is the treatment of choice.

KEYWORDS: Cholesteatoma, Congenital, Middle ear, Temporal bone

INTRODUCTION

Congenital cholesteatoma is found in many sites of the temporal bone including 5 locations: Tympano-mastoid, petrous apex, cerebellopontine angle, and jugular foramen.^[1] The fifth is between tympanic membrane (TM) layers.^[2,3]

Other very rare sites include infratemporal fossa.^[4,5] In addition, Granato *et al.* described isolated congenital cholesteatoma of the mastoid process.^[6]

By far, congenital cholesteatoma is originally presented as a pearl behind normal TM mostly located in the anterosuperior

quadrant.^[7] Levenson *et al.* in 1989 defined certain criteria for diagnosis of cholesteatoma of the middle ear including: (1) Presence of a whitish mass behind a normal TM, (2) a healthy pars flaccida and pars tensa, (3) no previous history of otorrhea or perforation, (4) No previous ear surgery, and (5) previous attacks of otitis media are not excluded.^[8]

In our case, no white mass appeared behind the TM. Furthermore, the anterosuperior quadrant was free of cholesteatoma.

CASE REPORT

A 20-year-old male patient came to otolaryngology department complaining of right sided hearing loss since a long duration but not exactly determined, progressive in a course with no other otologic symptoms. Clinical local examination revealed right

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conductive hearing loss, otomicroscopic examination revealed mild dullness and retraction of the right ear drum [Figure 1] with no other abnormal findings, and intact TM on both sides.

No history suggestive of trauma, otitis media or previous ear surgery.

Audiogram: Bilateral conductive hearing loss, moderate on the right side, and mild on the left side.

Tympanogram: Right type AD and left type A tympanometry curves.

Computed tomography findings

Right middle ear revealed soft-tissue nondependent opacity in the oval window region surrounding the tympanic portion and second turn of the facial nerve, the long process of incus and stapes are not seen, mastoid is free on both sides and the left ear showed no abnormality [Figures 2 and 3].

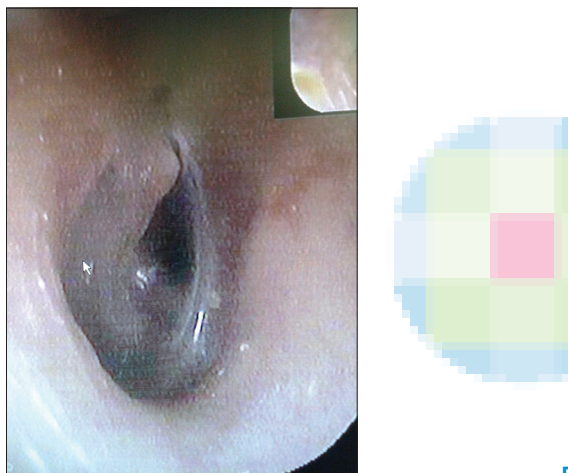


Figure 1: Otomicroscopic examination of the patient showing intact but dull and retracted right tympanic membrane

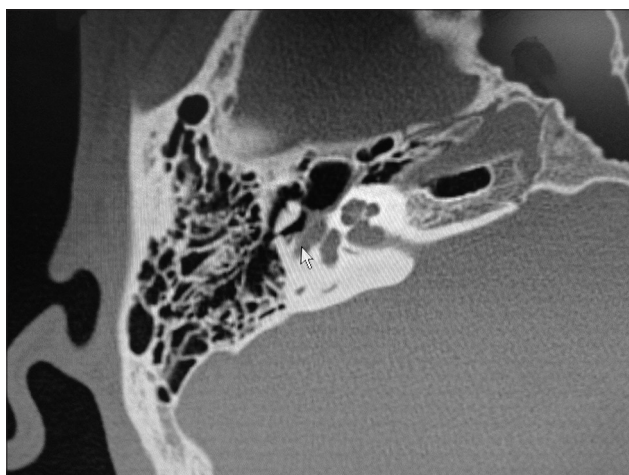


Figure 3: Axial computed tomography temporal bone of the patient showing soft-tissue opacity surrounding the tympanic segment of the facial nerve

Surgical findings

Under general anesthesia, postauricular approach was done with elevation of the tympanomeatal flap and getting access to the middle ear, limited atticotomy from within and drilling the most medial part of post bony meatal wall exposing the posterior wall of the tympanic cavity and ossicular chain. We found cholesteatoma (whitish mass in the region of posterior mesotympanum with absent incus and stapes superstructures). Cholesteatoma was covering the O.W niche and sitting on the facial nerve (dehiscent fallopian canal in a part of the tympanic segment, second turn [Figure 4], and the upper part of the mastoid segment).

The cholesteatoma extends deep to the facial nerve into the tympanic sinus (difficult and unpleasant site), careful peeling of cholesteatoma matrix was done, malleus was removed.

Silicon sheet was introduced in the tympanic cavity covered by temporalis fascia and a piece of auricular cartilage in the attic

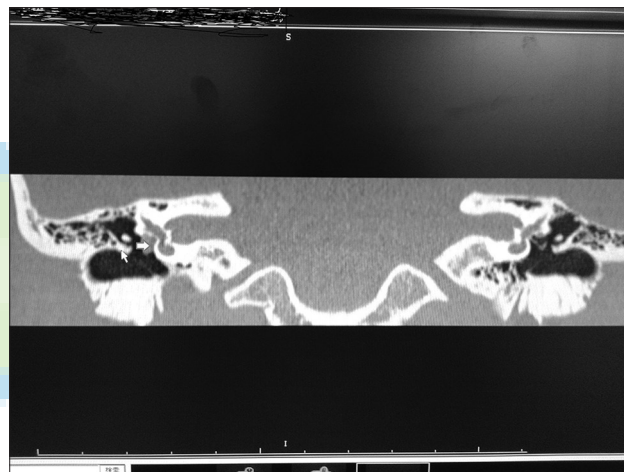


Figure 2: Coronal computed tomography temporal bone of the patient shows nondependent soft-tissue opacity in the oval window region and surrounding the tympanic segment of the facial nerve with free mastoid on both sides, normal left ear

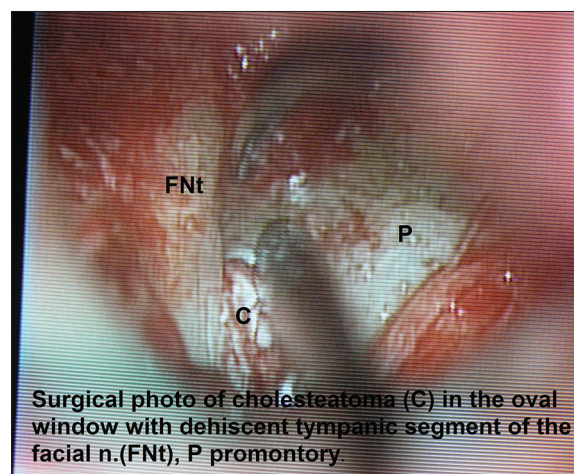


Figure 4: surgical photo showing cholesteatoma mass in the oval window, and dehiscent tympanic portion of the facial nerve

for second look surgery and reconstruction of the middle ear to restore hearing. No intra- or post-operative complications were recorded.

Histopathological report

Cholesteatoma (ingrown squamous epithelium in chronic inflammatory fibrous tissue).

DISCUSSION

The first otologist who described congenital cholesteatoma behind intact ear drum was House in 1953.^[9]

A staging system for congenital cholesteatoma was suggested by Postic *et al.* in 2002 that depends upon quadrants affected by cholesteatoma with or without ossicular or mastoid involvement.^[7]

- Stage 1: Cholesteatoma involving one quadrant
- Stage 2: More than one quadrant without ossicle involvement
- Stage 3: Ossicular involvement by cholesteatoma or surgically removed to clear cholesteatoma
- Stage 4: Mastoid involvement.

According to this staging system, our case is stage 3 as stapes suprastructure and incus are destroyed by cholesteatoma.

Congenital cholesteatoma is originally represented as a pearl behind an intact TM mostly in its anterosuperior quadrant.^[7] However, in our case no white mass was revealed behind the ear drum but some dullness of the posterior part of the ear drum was observed. Furthermore, the anterosuperior quadrant was free of cholesteatoma.

Despite that the most common location of congenital cholesteatoma of the middle ear is the anterosuperior quadrant, a systemic review and meta-analysis were performed by Hidaka *et al.* in 2013 and revealed that congenital cholesteatoma is found more in the posterosuperior quadrant in the Asian population than western countries.^[10]

In this case, a large area of the fallopian canal was dehiscence, but the patient had no facial nerve paralysis. The facial nerve sheath was intact and still not involved or compressed by cholesteatoma, may be if the surgery was delayed, facial palsy would be the next sequel.

We think that staged surgery is the best choice for this patient, as he is still young, and the expected lifespan is long, so it is desired that eradication of all cholesteatoma debris would be confirmed in second stage operation and then reconstruction of middle ear to restore hearing is recommended.

CONCLUSION

We have focused light in an unusual case in our practice, discussed its management, and concluded that early intervention in such cases is recommended to prevent complications and planned surgery is the treatment of choice.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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