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Journal of Ear, Nose and Throat Disorders

Case Report

Intratympanic Membrane Cholesteotoma: An Unusual Rare Case

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Submitted: 11 July 2016

Accepted: 23 July 2016

Published: 01 August 2016

Keywords

- Intratympanic membrane
- Cholesteatoma
- Acquired

Abstract

Intraympanic membrane cholesteotoma without a preceding ear trauma or surgery is a rare entity. Only few cases have been documented in the literature. The exact etiology of this lesion is still unclear. We report an unusual case of an intratympaniccholesteatoma in an adult patient. He is a 45-year-old male who presented with ear fullness and hearing loss six months following an acute otitis media without otorrhea or a tympanic membrane perforation. Physical exam showed a central whitish area on the tympanic membrane. CT scan demonstrated thickening of the tympanic membrane. On the audiogram, he had a 20-dB air-bone gap; tympanogram showed a B-curve. We surgically explored his middle ear and encountered a central tympanic membrane 5X3 mm cholesteatoma with intact middle ear cavity. Accordingly we performed a tympanoplasty. The gold standard for diagnosis of tympanic membrane cholesteotoma is oto-microscopy. This early recognition of the cholesteatoma prevented the patient from suffering the consequences of the middle ear cholesteatoma. We recommend a high index of suspicion to diagnosis and treat this entity early, in order to avoid long term complications.

INTRODUCTION

Intratympanic membrane cholesteatoma is a very rare finding especially when it occurs in the absence of otologic trauma or surgery [1]. Almost only 20 cases have been reported in literature [2]. In general, most of them are labeled as congenital in nature although some have beenassociated with otitis media [2]. Congenital cholesteatomas are also rare, comprising 2%-5% of all cholesteatomas [2]; though this number is on the rise, still intratympaniccholesteatomas are even less common [3]. When identifying a whitish tympanic membrane lesion, cholesteataoma should be always considered on the differential diagnosis list. We present a 45-year-old male with a right ear intratympanic membrane cholesteatoma treated with surgical resection and tympanoplasty.

CASE REPORT

A 45-year-old male presented to our clinic with right ear fullness and hearing loss over a six months' period following an acute otitis media. The patient reported no otorrhea at any point in time. He denied previous ear surgery or trauma. Physical exam showed a central whitish area on the tympanic membranemeasuring 5X3 mmwith intact tympanic membrane which was located near the umbo (Figure 1) as a comparison to the healthy looking left tympanic membrane (Figure 2). Audiogram showed a 20 dB air-bone gap and speech recognition test (SRT) of 35-dB; tympanogram showed a B-curve with absent reflexes. CT scan showed thickening of the tympanic membrane without extension into the middle ear or any ossicular deformity along with fluid in the bilateral mastoid cavities (Figure 3). A CT scan was ordered to evaluate any middle ear pathology and ossicular involvement; after the images revealed that the confinement

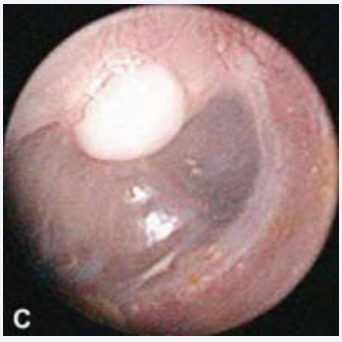

Figure 1 White lesion found near the umbo of the right tympanic membrane.

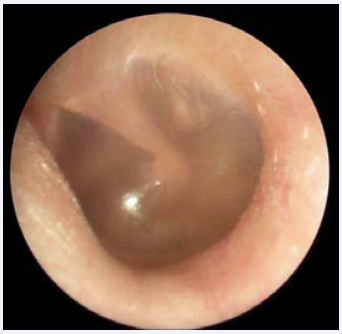

Figure 2 Healthy looking left tympanic membrane.

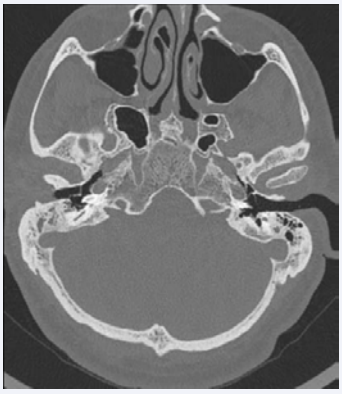

Figure 3 Right thickened tympanic membrane with no middle ear cholesteatoma extension.

of the lesion to the tympanic membrane we decided against performing diffusion weighted MRI. We surgically explored his middle ear through a transcanal approach and encountered a central tympanic membrane 5X3 mm cholesteatoma. The middle ear cavity was intact and ossicles were mobile and intact. Accordingly we removed the affected tympanic membrane area and performed a tympanoplasty. Histopathology confirmed the cholesteatoma diagnosis. At the sixth-month follow-up the patient had a healthy looking tympanic membrane without signs of recurrence; audiogram showed an SRT of 10-dB and a 5-decibel air-bone gap. This audiogram confirmed that the mass effect of the cholesteatoma on the tympanic membrane is the main cause for the hearing loss. An informed consent was obtained from the patient before presenting him in the case report.

DISCUSSION

Hinton in 1863 was the first to describe intratympanic membrane cholesteatoma [4]. More cases were reported consecutively, where Teed reported five cases of congenital intratympaniccholesteatomas in 1936 followed by Smith and Moran in 1977 and Sobol et al., in 1980 [5-7]. In addition, in 1998 Jaisinghani described 5 intratympanic cholesteatomas in 243 temporal bones with chronic otitis media [4]. In the study, two patients had retraction pockets, one had a tympanic membrane perforation, and the other two were not described. Usually, intratympanic cholesteatomas occur as a sequela of otologic surgeries, mainly lateraltympanoplasty [1]. It is very rare to find this entity in patients without previous ear surgery or trauma [5]. Such patients will be labeled as having a congenital cholesteatoma. However, few cases of otitis media do not exclude the possibility of a congenital cholesteatoma [8].

Broadly speaking, cholesteatoma is divided into: congenital and acquired [9]. Although not perfectly clear, several hypotheses have been postulated regarding the cholesteatoma development. Congenital cholesteatomas have been defined as remnant embryonic epithelial tissue in patients with no history of otologic surgery and no otoscopic findings of a tympanic membrane perforation [9]. As for the acquired cholesteatoma, also several theories have existed regarding its formation. These include “squamous immigration/invasion” theory, “squamous obstruction-vacuum retraction/retraction pocket” theory, “squamous metaplasia theory” by Sade, “squamous basal hyperplasia/papillary ingrowth” theory, and the most recent being “mucosal migration and traction exertion on the tympanic membrane” theory [10].

Our patient is a 45-year-old male with bilateral mastoid fluid and an intact tympanic membrane. This excludes the possibility of a congenital cholesteatoma; therefore, he has an acquired cholesteatoma. It is supported by the “squamous metaplasia” theory which states that inflammation in the middle ear leads to metaplasia of the middle ear mucosal lining. However, very few cases reported intratympanic membrane cholesteatoma without middle ear extension due to chronic otitis media or otomastoiditis [4]. It is common in patients who report a history of otolgic trauma or surgery mainly lateral tympanoplasty [1].

On the differential diagnosis of a whitish tympanic lesion, mainly one could think of tympanosclerosis or cholesteatoma. It is important to differentiate intratympanic cholesteatoma from tympanosclerosis, sincemissing a cholesteatoma might lead to severe complications including ossicular involvement and hearing loss. Once the intratympanic membrane cholesteatoma is suspected clinically by oto-microscopy, computer tomography scan is necessary to assess the tympanic membrane lesion, the ossicular chain, middle ear, and mastoid cavity involvement. The standard treatment is surgical resection of the cholesteotomaendaurally. If possible, peeling the cholesteatoma off the tympanic membrane andleaving an intact fibrous layer is the optimal treatment. However, when more extensive, tymplanoplasty is the required alternative [6].

CONCLUSION

When a patient presents with a white lesion on the tympanic membrane, tympanosclerosis and tympanic membrane cholesteatoma arise on the differential diagnosis; here the gold standard for differentiation of a suspicious lesion is oto-microscopy [2,3]. The early recognition of the intra-tympanic membrane cholesteatoma prevents the patient from suffering the consequences of the middle ear cholesteatoma. We recommend a high index of suspicion to diagnosis and treat this entity early, in order to avoid long term complications.

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Cite this article

Adib H, El Natout T, El Natout MA (2016) Intratympanic Membrane Cholesteotoma: An Unusual Rare Case. J Ear Nose Throat Disord 1(1): 1007.