Case report

Management of the association of otosclerosis and cholesteatoma: Which pathology to treat first?

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ABSTRACT

Hearing loss can result from a wide range of pathologies affecting patients of all ages. It may be due to abnormalities of the outer, middle or inner ear.

In this article, we present a case of a 50 years old female patient presenting to the ENT and Head & Neck surgery department of 20 August hospital, complaining of bilateral hearing loss, in whom clinical examination found right attic retraction pocket with scales and left normal tympanic membrane. Complementary investigations revealed the association of two distinct conditions, rarely described in literature: otosclerosis, and chronic otitis media with cholesteatoma in right ear and left otosclerosis.

The challenge in this case was to determine the therapeutic strategy: start with left otosclerosis? the right cholesteatoma? simultaneously treat otosclerosis and cholesteatoma right?

1. Introduction

Cholesteatomatous chronic otitis media and otosclerosis are both moderately common pathologies. They may be expected to occur together coincidentally [1]. This association has been rarely point of in literature [2].

Diagnosis is based on clinical examination including bilateral otoscopy, pure tone audiometry and a CT scan of the temporal bone. In fact, once the diagnosis is made; an appropriate treatment in a delayed time must be undertaken. This work has been reported in line with the SCARE 2020 criteria [3].

2. Case presentation

A 50 year old female, with no personal or family history, presented with a long standing bilateral hearing loss that becomes more apparent about 2 years ago. Hearing loss was non-fluctuating and was profound causing social exclusion and depression. In addition to the hearing loss, she had otorrhea on right ear ongoing for years, foul smelling and non-resolved with both oral and oto topical antibiotics. She also reported bilateral non pulsatile tinnitus. Clinical examination showed a normal tympanic membrane in the left ear and an attical cholesteatoma in the right one (Fig. 1). Neither vertigo nor facial palsy was found in the examination. General examination was non-contributory.

A pure tone audiometry was realized showing both right and left sided profound mixed hearing loss around 80 dB, with an average Rinne around 45 (dB) (Fig. 2).

The tympanogram showed the absence of the stapedial reflex on both sides.

For further evaluation, the patient underwent a computed tomography (CT) temporal bones without contrast showing at right ear an homogeneous soft tissue mass with smooth bony expansion of the attic and mastoid antrum, a well ventilated middle ear cavity with partial lysis of the ossicular chain and an eroded scutum, also we noticed a prestapedial hypo density measuring 2.8 mm without cochlear involvement. The left ear was free of any signs of cholesteatoma with a prestapedial hypo density measuring 3.2 mm without cochlear involvement; both classified type 2 according to the Veillon classification. There was no erosion of the canal wall of the facial nerve, nor lateral semicircular canal; both appearing to be grossly intact (Figs. 3-5).

The therapeutic strategy was discussed taking into consideration the depressive state and the social isolation of the patient due to her hearing loss. The decision was to operate first the otospongiosis site of the left side, in order to improve the hearing prognosis as well as the patient’s quality of life; then, the right side starting with the cholesteatoma surgery in the first row only few weeks later. The surgery was performed by

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a surgeon experienced in ear surgery 1 month ago. Under general anesthesia, the first step of surgery was exploration of the ossicular chain and confirmation of the diagnosis of otosclerosis, then we performed a calibrated stapedotomy with a trans-platinum prosthesis.

The second surgery has not been yet performed, it is scheduled in a few weeks in which the right cholesteatoma will be treated, and only after being sure of the total eradication of the cholesteatoma, a surgery of the right otosclerosis will be proposed.

During the postoperative consultations, the patient was satisfied with the improvement in hearing with better social integration. Her post-operative pure tone audiometry showed a gain of 35 dB on the left ear which brings her hearing threshold to 45 dB with a Rinne of 10 dB.

3. Clinical discussion

Otosclerosis is a primitive osteodytrophia of the labyrinthine bone, firstly described in 1893. It is responsible of a progressive hearing loss. Other oto-pathological condition, frequently overlooked, may be concomitant to otosclerosis and their discovery can affect therapeutic decision [4].

The association otosclerosis and other oto-pathological affections have been given little attention in literature, even if these concomitant occurrences can be clinically relevant [5].

Paparella in his study realized in 2007, emphasize the prevalence and types of associated otopathologic conditions. Over 182 cases of otosclerosis studied, 34 % had otitis media, 28 % endolymphatic hydrops without Ménérie's disease symptoms and 15 % serous labyrinthitis. Otitis media included serous otitis, mucoid, as well as chronic otitis with or without cholesteatoma. These combinations are mostly coincidental and not causative except for advanced Ménérie's disease following otosclerosis with obstruction of the vestibular aqueduct [6,7].

After a search on the various scientific search engines Pubmed, DocCISMeF, google scholar, no articles address the prevalence of otosclerosis associated with middle ear cholesteatoma nor of the therapeutic strategy to adopt in this case.

The key to the diagnosis is a targeted questioning looking for personal and family history, and the functional signs, as well as a well-conducted clinical examination of the ENT sphere. Clinical examination is completed by a hearing assessment.

In fact, pure tone audiometry is the mainstay of investigations, confirming the hearing loss, quantifying its severity and precisely its nature whether it is conductive, mixed or sensineural.

Non contrast temporal bone CT is considered the gold standard for otosclerosis imaging [8]. The imaging findings in otosclerosis largely depend on the phase of the disease and its location. It confirms the diagnosis of otosclerosis by showing the thickening of stapes's platine and hypo density images of the labyrinthine bone. It also eliminates other causes of conductive hearing loss and makes it possible to assess its severity by the Veillon classification. It likewise guides the surgeon by seeking surgical risk variants [9].

These findings may influence therapeutic modality, particularly in case of association of cholesteatoma with otosclerosis where the attitude would consist of surgical excision of the cholesteatoma initially given the risk of endocranial complications, and delayed surgery of ostospongiosis after stabilization of otitis.

4. Conclusion

To sum up, the coexistence of otosclerosis with other otopathological conditions such as cholesteatoma is extremely rare and may be coincidental, as is the case in our case and in most patients. In fact, this condition has been rarely described in literature. The management of such a situation requires an experienced surgeon and must take into account the patient’s complaints to combine local therapeutic control and early social integration.

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Personal sources.
Ethical approval

Ethical approval has been exempted by my institution.

Consent

Written informed consent was obtained from the patient's family for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

All the authors worked in coordination to ensure the best management of the disease from diagnosis to the hospitalization and surgery. The authors wrote this article together.

Registration of research studies

This study is not “First in Men”.

Guarantor

Lyoubi Mouna.

Declaration of competing interest

No conflicts of interest.
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