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Case report

MANAGEMENT OF BILATERAL IDIOPATHIC CHOLESTEATOMA OF THE EXTERNAL AUDITORY CANAL: A CASE REPORT

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ABSTRACT

Cholesteatoma of the external auditory canal is an extremely uncommon pathology that has an incidence of 0.1% in the world population. Generally, cholesteatoma presents in a unilateral form with accumulation of epidermal deposits. We described a case of a young Caucasian woman, with history of bilateral hearing loss and ear fullness. The external auditory canals (EAC) presented desquamated epithelia on the floor and bony erosion of the anterior portion, without involving tympanic membrane and middle ear. Patient performed bilateral canaloplasty, meatoplasty, and cholesteatomas were drilled out. Cholesteatoma is not a pathology exclusive to the middle ear but can also be found, rarely, in the EAC. It is important to perform a differential diagnosis between necrotic otitis, epidermal cap, neoplasms of EAC and epidermal cysts. The biopsy of the lesion, a radiological study, and the surgical treatment with total remotion of the cholesteatoma are the gold standard for best management of the pathology.

1. Introduction

Cholesteatoma of the external auditory canal (EAC) is a unique clinical entity defined as a conglomeration of epidermis combined with bone corrosion osteitis, being mainly idiopathic or occurring after surgery or damage to the auditory canal [1]. It is worth to note that cholesteatoma should be distinguished by keratosis obturans, which is instead defined as conglomeration of desquamated keratin in the ear canal [2-3].
Since its initial description by Toynbee [4] in the 19th century, otolaryngologists have speculated about the pathophysiology of the disease, trying to attribute the cause to a defective epithelial migration [1]. However, the exact mechanism of EAC cholesteatoma (EACC) formation remains ambiguous. Association with other diagnoses of EAC abnormalities can provide insight into the disease process. The most frequent pathology that may affect the morphology of the ear canal are Beckwith-Wiedemann syndrome, Turner syndrome, Treacher Collins syndrome, Down syndrome, focal dermal hypoplasia, and cleft palate [5].

External auditory canal cholesteatoma has an incident percentage of 0.1% worldwide, equally distributed between female and male gender [6-7].
Mono-lateral cholesteatoma is more frequent, since this disorder is demolitive and can spread to the middle ear causing auditory injury. During clinical diagnosis EACC is often traded with middle ear cholesteatoma [8-9].
EACC should be classified in:
- idiopathic;
-secondary: post-stenotic, post-operative, post-traumatic, post-radiogenic and post-inflammatory [10].
Physical examination and computer tomography (CT) of the temporal bone play a major role in the diagnosis. In particular, high-resolution CT offers images with elevated resolution and high anatomical detail, thus improving diagnosis, surgical procedure, and prognostic assessment [7].
If EACC is not correctly treated, it can cause different complications. Its development can invade the anterior tympanic box and inferior auditory canal.

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In cases of high jugular bulb, the growth of EACC can lead to jugular bulb exposure and invading it can cause severe bleeding to occur. The development of the EACC in the middle ear could lead to the same complications of the middle ear cholesteatoma, for example facial nerve canal invasion and facial paralysis, labyrinth bone erosion and deafness or labyrinthitis, sigmoid sinus and skull base invasion with thrombophlebitis, meningitis, encephalitis, and cerebral abscess [7].

In this manuscript we described a rare case of bilateral congenital cholesteatoma of the external ear that was diagnosed in a young adult without any previous trauma or any history of previous ear surgery. The aim of the article is to describe the best management and treatment in the case of this rare pathology.

2. Case report

We report the case of a 17 -year-old woman treated in a tertiary referral center in August 2018 with bilateral auricular fullness. She reported a longstanding history of auricular fullness with frequent removal of cerumen plugs. She has an history of chronic myeloid leukemia BCR-ABL+ (gene), treated with Glivec e Ara-C with good control of the pathology.

At bilateral otoscopic evaluation of the EACs, the patient presented with: desquamated epithelia on the floor, bony erosion of the tier and anterior portion on EAC evident after the removal of the keratin tissue, the exposed bone bleeding on touch, normal tympanic membrane without any pathology in the middle ear and no otorrhea. The patient reported no pain, no history of previous trauma except cerumen plugs removal and no history of previous ear surgery.

The patient initially was treated medically with removal of the excessive accumulation of desquamated keratin, with instillation of topic lotion with antibiotics + corticoids (Ciprofloxacin + Dexamethasone for 2 weeks) and cleaning lotion with low response to medical treatment with continuous bone exposure and erosion.

Audiometric evaluation was performed after the bilateral microscopic cleaning of the EACs and it showed no bilateral hearing loss.

CT of the temporal bone showed bilateral concentric phlogistic thickening of the EACs and erosion of the floor without any involvement of the ear drum or the ossicular chain and a normal pneumatization of the mastoid cells without any pathology (Figs. 1-2).

After the failure of the conservative treatment with otomicroscopic cleaning and medical procedures such as eardrop instillation Ciprofloxacin + Dexamethasone, the patient was submitted to bilateral surgical procedure.

Surgery consists in a retro-auricular approach, with microscope auxilium. Bilateral canaloplasty was performed, drilling out the cholesteatomas. Meatoplasty was executed to control recurrence during medication. The material, removed bilaterally, was examined histopathologically with response of "lamellar horny material comprising superficial squamous epithelium commingled with anucleate scales, granulocytes and flaps of squamous epithelia with Parakeratosis. No atypia - Finding consistent with Cholesteatoma - comprising lamellar horny material."

During follow-up in the last 3 years, there was no sign of recurrence of pathology with otoscopic examination and CT scan (Figure 3).

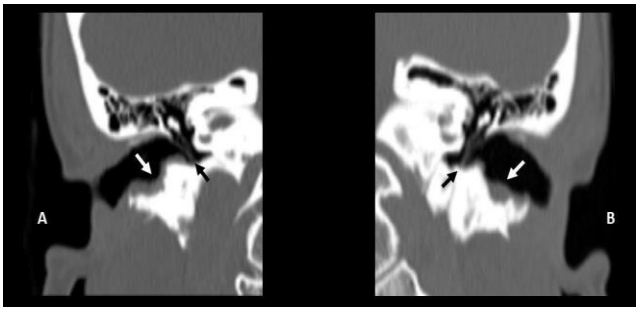

Figure 1. Coronal section of CT scan. Fig. 1A right side, Fig. 1B left side: white arrow: Erosion of the inferior wall of the EAC with phlogistic thickening (accumulation of desquamated keratin); black arrow: no pathology on the tympanic membrane and normal appearance of the middle ear cavity with no involvement of the ear drum.

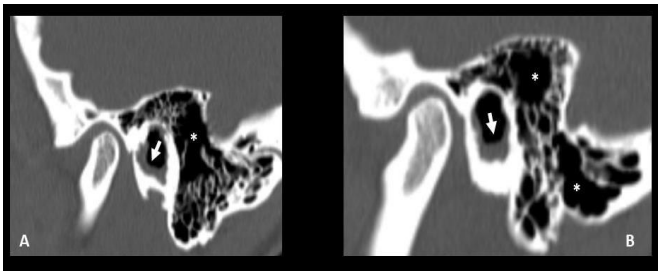

Figure 2. Sagittal section of CT scan. Fig 2A right side, Fig 2B: left side; white arrow: erosion of the inferior wall of the EAC with phlogistic thickening (accumulation of desquamated keratin); white asterisk: mastoid cells with regular pneumatization and not affected by pathology.

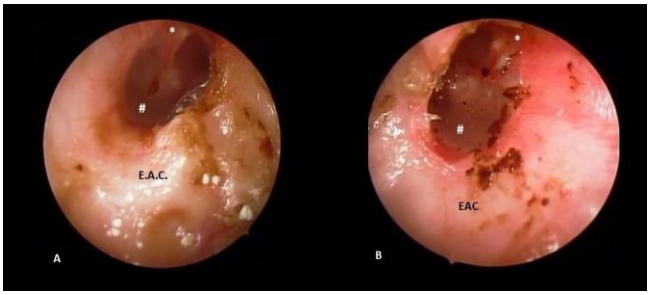

Figure 3. Endoscopic evaluation of the left EACs 3 years after surgery: (A) right side; (B) left side. (#) integrity of ear drum; (*) integrity of pars flaccida; EAC with no presence of pathology after 3 years.

3. Discussion

Cholesteatoma is an abnormal, heterotopic, noncancerous skin growth. Generally, it presents similar to a cystic structure plated by layered keratinizing squamous epithelium accompanying periostitis and bone erosion [11-12].

Cholesteatoma are found in most cases in the middle ear and mastoid and they can be divided as acquired and congenital, with the acquired one representing 98% of cases [13].

An estimated percentage of 0.1-0.5% cholesteatoma can be found in EAC and it can be idiopathic or appear after a trauma or surgery; cholesteatoma does not represent a pathology exclusive of the middle ear, as it can also be found, though rarely, in the paranasal sinuses and neighboring anatomic areas, like the pterygopalatine space and petrous apex of the temporal bone [14].

Congenital middle ear cholesteatoma and cholesteatoma of paranasal sinus are attributed to aberrant embedding of ectodermal epithelial cells during the fusion of epidermal surfaces [15].

However, there is still an anamnestic mystery about the causes leading to idiopathic EACC formation. Until now, no pathognomonic clinical symptoms or objective examination are available; diagnosis is performed by medical examination accompanied by CT of the temporal bone and biopsy [16].

Regularly, the tympanic membrane is untouched, excluding cases of wide cholesteatoma. In this case, it's difficult to define the origin, i.e., middle ear (most common) with invasion of the EAC (just 0.1-0.5%) [15].

Middle ear congenital cholesteatoma originates from residual embryonic epithelium that remains trapped in the tympanic cavity during embryogenesis. Congenital cholesteatoma presents as a pale mass that may be visible through an intact and transparent tympanic membrane, and patients have a negative history of otitis media. If there is perforation of the tympanic membrane and the patient had a history of chronic otitis media, the diagnosis is that of otitis media due to acquired cholesteatoma [16].

In our case no previous history of otologic pathology and surgeries, the age of the patient, the bilateral presentation with no pain or otorrhea, the previous history of CML BCR-ABL+ or the bleeding on touch of the exposed bone induced the medical team to differential diagnosis including neoplasms of EAC, epidermal cap, epidermal cysts, and external necrotic otitis [17].

The etiopathogenetic hypothesis we propose for underlying bilateral EACC as reported herein is:

i. incorrect maneuver of removing ear plugs
ii. secondary neoformation given by chemotherapy previously performed by the patient according to her medical history of CML BCR-ABL+, treated with Glivec e Ara-C, that can cause alteration and dryness of the skin, therefore also in the EAC.

If on the one hand CT is not mandatory, it represents a fundamental tool to plan adequate surgery and to avoid complication due to anatomical variants [18].

The diagnosis of EACC at an early stage is done with an EAC biopsy. After that, a CT scan is useful to understand the lesion extension. However, if the lesion invades the middle ear, the objective radiological exam is similar to that of a middle ear cholesteatoma.

It is important to understand the different characteristics between EACC and middle ear cholesteatoma.

If cholesteatoma initially involves the EAC, the first stage is the bone destruction in canal with a pneumatic mastoid. So, the most important characteristics of cholesteatoma are the stenosis of EAC and a flask-shape for the internal part. The epitympanic cavity, the Prussak recess and the scutum are involved with a destructive bone erosion from outside towards inside. The anterior part of the mastoid and temporomandibular joint could be involved and injured. The general tendency is that the lesion starts from the external ear canal to the surrounding parts.

In the middle ear cholesteatoma, the mastoid appears sclerotic or with secretions in the cells, the lesion is located in the middle ear and normally there is not an expansion to the external auditory canal. In cases of middle ear cholesteatoma that include epitympanic cavity, the Prussak recess and the scutum, the structures appear with a destructive bone erosion from the inside towards the outside. The middle ear cholesteatoma involves aditus and antrum tympanicum with an obliteration of them. All these characteristics are typical for middle ear cholesteatoma.

Therefore, high-resolution CT (HRCT) can give important details about the extension of EACC. It could be useful to analyze particular conditions that could lead to complications, and to differentiate EACC from middle ear cholesteatoma. Furthermore, HRCT can give an important contribution to analysis of the anatomical parts involved, and to find possible complications. It is important to underline that an accurate diagnosis must be obtained on a detailed physical examination and medical history, follow-up and intraoperative exploration [7].

Udayabhanu introduced the staging criteria for EACC.

Stage I: lesions are limited to the external auditory canal without bone destruction;

Stage II: lesions are characterized by bone destruction with or without middle ear involvement but without the involvement of adjacent structures (including the temporomandibular joint, mastoid process, jugular venous bulb, facial nerve canal, and dura mater)

Stage IIIA: lesions are characterized by bone destruction and involvement of adjacent structures without complications;

Stage IIIB lesions are those with a combination of other complications.

Unlike the classical Holt staging, which includes the area of invasion of the middle and upper tympanic chambers as the areas of predominance for endoscopic surgery, stage III of Holt staging is relatively broad. Compared with Holt's staging criteria, stage II patients have a larger scope (e.g., superior tympanic chamber), and for the current endoscopic management of superior tympanic cholesteatoma, stage II patients of Udayabhanu can be completely resolved, and some stage IIIA patients are also feasible for endoscopic surgery after comprehensive evaluation, but strict indications need to be mastered, and timely replacement of the posterior auricular approach with microscopic mastoid surgery should be prepared. There is preparation of the posterior auricular access microscopic mastoid surgery [19-20].

In this case, the lesion was at Stage II of Udayabhanu classification and surgery was performed with a retro-auricular approach, without involving the middle ear.

As with middle-ear cholesteatoma, the aims of treatment in external auditory canal cholesteatoma cases are the eradication of the disease with preservation of normal structure and function, and the restoration of normal epithelial migration.

Conservative therapy, with frequent debridement of the keratin debris and sequestered bone, is favoured in early stage disease.
If these simple measures are inadequate to control otalgia and otorrhoea, or in the case of a more advanced disease (stages IIb–IV), surgical intervention is necessary. The aim of surgery is to excise the cholesteatoma and to restore a smooth, self-cleaning canal wall epithelium, usually with the aid of cartilage and fascial grafts to protect denuded structures, and fill any canal wall defect. When the mastoid air cells are invaded, a modified radical mastoidectomy may be indicated, with the tympanic membrane and ossicles left intact [21-22].

4. Conclusions

Cholesteatoma does not represent a pathology exclusive of the middle ear as it can also be found, though rarely, in the EAC, as reported herein. The causes and physiopathological mechanisms underlying idiopathic EACC are still unclear and further research are warranted to shed new light on this complex pathology. The lack of pathognomonic clinical symptoms makes its diagnosis challenging, and CT of the temporal bone, with histological and anatomopathological examination are required for definitive diagnosis. In the present case, surgery represented the only solution to remove the pathology, allowing extensive cleaning of the surgical site to avoid recurrence. In addition, the meatoplasty was fundamental to perform a correct medication of the ear canal and to better prevent any recurrence.

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