Pode um colesteatoma ter uma recidiva tardia?

Caso Clínico

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Artigo recebido a 13 de Julho de 2023.
Aceite para publicação a 17 de Setembro de 2023.

Resumo

Reportamos o caso de um doente de 73 anos, género masculino, com recidiva tardia de um colesteatoma do ouvido médio, diagnosticado 33 anos após a cirurgia primária.
O doente apresentou-se com sintomatologia de otite externa refratária ao tratamento médico, o que levou à suspeita de colesteatoma, tendo em conta a história pregressa.
A tomografia computorizada revelou uma lesão volumosa de densidade de tecidos moles com complicação craniana iminente.
A ressonância magnética confirmou o diagnóstico de colesteatoma. Foi realizada petrosectomia subtotal com obliteração do ouvido médio para remoção completa do colesteatoma.
O principal objetivo deste caso é salientar a importância do seguimento durante toda a vida dos doentes submetidos a cirurgia por técnica canal *wall up* por otite média crónica colesteatomatosa, nos casos em que não foi realizada uma ressonância magnética com difusão ou uma cirurgia *second look*.

Palavras-chave: Colesteatoma; complicações intracranianas; recidiva; recidiva tardia

Introduction

Cholesteatoma is a non-neoplastic, well-demarcated cystic lesion of the temporal bone, resulting from an abnormal growth of keratinizing stratified squamous epithelium[1]. It can be classified, according to their pathogenesis, into congenital or acquired, being the latter more common[2]. The treatment is surgical and numerous attempts have been made to optimize the surgical techniques and to lower the recidivism rate[3]. We describe the case of a patient with late recidivism 33 years after the first surgery, with imminent intracranial complication.

Case Presentation

A 73-year-old male patient presented with a 1-month history of left ear otorrhea and long-term hearing loss in the same ear. He had a history of left ear surgery for cholesteatoma, 33 years ago. After the primary surgery, the patient had regular clinical follow-up visits during 27 years without clinical signs of recidivism, but he did not perform magnetic resonance imaging (MRI) neither was submitted to second-look surgery. The records of this primary surgery were not available. On examination, a whitish discharge and swollen of the left external auditory canal was detected, which was initially interpreted as an external otitis. Audiometrically, the left ear audiogram showed a mixed hearing loss of moderate degree. After several unsuccessfully attempts of treatment with antibiotic and steroid drops, as well as systemic antibiotic, an ear computed tomography (CT) scan was performed. The CT scan revealed a large expansive and erosive lesion with benign characteristics measuring 4.2x2.8x3.1 cm, involving the left mastoid, which determined erosion of the tegmen mastoideum with possible intracranial communication, erosion of the posterior aspect of the otic capsule and lytic destruction of the ossicular chain (fig. 1). Differential diagnosis as cholesteatoma, mucocele and cholesterol granuloma could be considered. To clarify the diagnosis, a magnetic resonance imaging (MRI) was requested, which showed a very large lesion measuring 2.7x4.1x4.2 cm centred to the left mastoid, that invaded the sinodural region and elevated the tegmen, but seems to be contained by the dura. The referred lesion was also associated with lateral sinus exclusion and moderate mass effect action on the left cerebellar hemisphere (fig. 2). MRI confirmed the diagnosis of cholesteatoma. Patient was treated with urgent subtotal petrosectomy with middle ear obliteration, cul-de-sac closure of the external auditory canal, obliteration of the Eustachian tube and complete removal of the cholesteatoma, including a meticulous

Figure 1
Axial CT image soft tissue window: Extensive and erosive soft tissue density lesion measuring 4.2x2.8x3.1cm involving the left mastoid (*); erosion of the posterior fossa cortical with possible intracranial communication (arrow) and extension of the lesion to the skin of the left external ear canal.

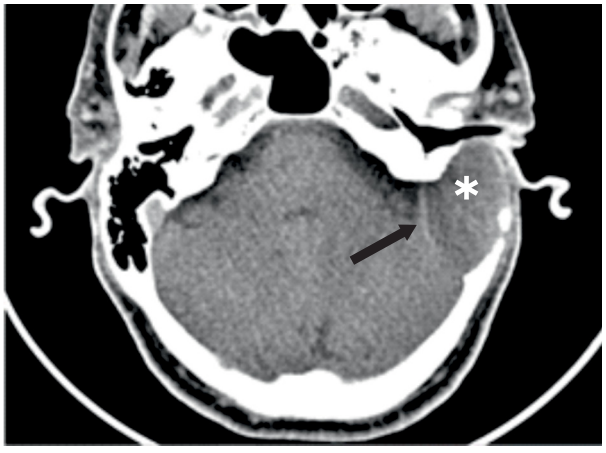
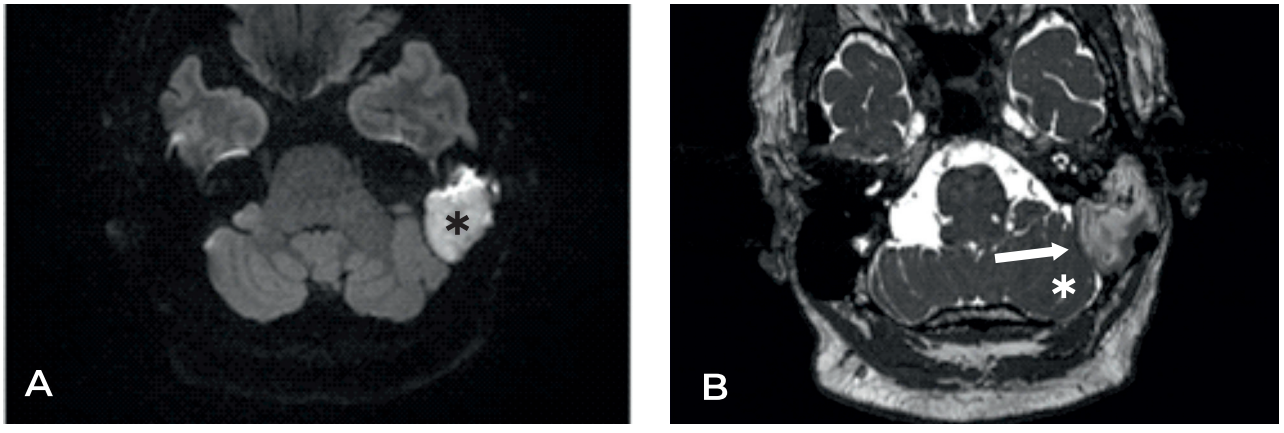


Figure 2
Cholesteatoma on MRI: (A) (axial diffusion weighted non-echo planar sequences): large soft tissue lesion measuring 2.7x4.1x4.2 cm centred to the left mastoid with hyperintense signal (*); (B) (axial CISS): The lesion shapes the cerebellum (*), but the dura is intact (arrow).



resection of the entire cholesteatoma matrix (CM) that was adherent to the dura mater (DM). In a posterior retrolabyrinthine location, the matrix was already adherent to the arachnoid, so, to remove the matrix, it was created a discontinuity of the arachnoid with subsequent exit of liquor (fig. 3-7). Histopathologic analysis showed abundant keratin lamellae with tissue flap covered by stratified squamous epithelium, consistent with cholesteatoma. Patient was discharged 5 days after surgery. He was followed once a week until 1 month, then at 3 months and 6 months. One year after surgery he will perform a CT scan as well as an MRI with diffusion protocol.

Discussion

Despite its benign nature, once established, cholesteatoma is a destructive lesion that grows gradually and causes erosion of adjacent structures. This can lead to several intra and extracranial complications[4-6]. Many patients with cholesteatoma complain of intermittent or constant fetid otorrhea. Hearing loss, due to involvement of auditory system, is usually conductive, but can also be sensorineural or mixed. Vertigo and facial nerve palsy occur when there is involvement of the vestibular system and facial nerve, respectively. Less common manifestations are tinnitus, otalgia and headache[4,7]. Nowadays, due to the diffuse use of antibiotics, secondary infections of cholesteatomas have decreased substantially. However, clinicians should be

Figure 3
(A) Cul-de-sac definitive closure of the external ear canal (*); (B) The mastoid cortical has been eroded (*) by the cholesteatoma anteriorly and posteriorly.

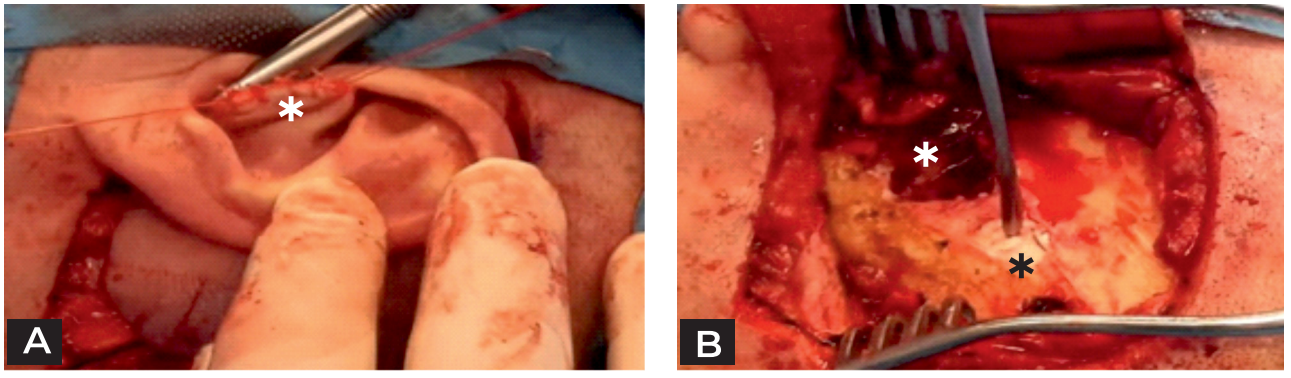


Figure 4
(A) Removal of the mastoid cortical (MC) surrounding the lesion; (B) Debulking of the cholesteatoma (Ch), which superiorly eroded partially the mastoid tegmen with exposure of corresponding dura mater; posteriorly, it extended a few centimetres behind the sigmoid sinus; medially, in the most posterior region, it totally eroded the posterior fossa cortical and compressed the corresponding intracranial structures.

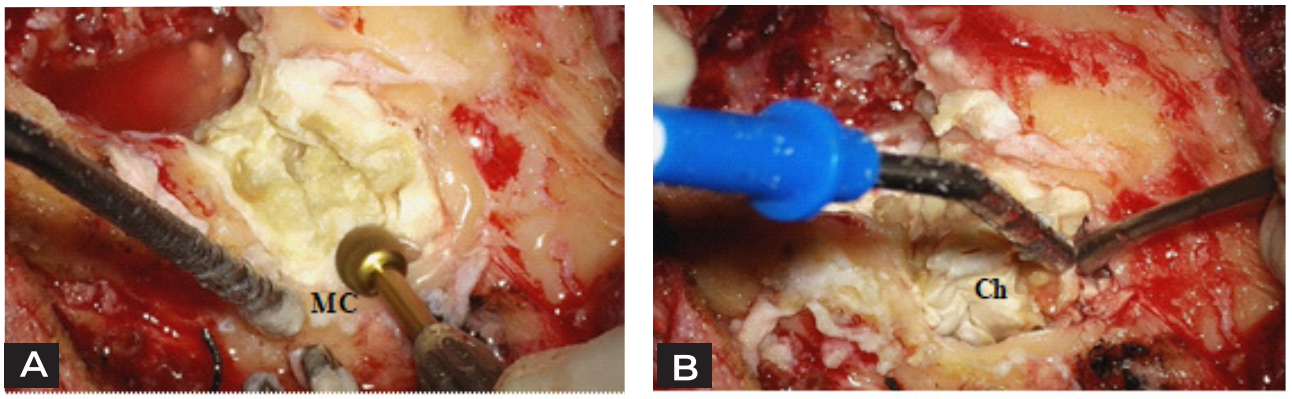


Figure 5
(A) Meticulous resection of the entire cholesteatoma matrix (CM) that was adherent to the dura mater (DM); (B) In a posterior retrolabyrinthine location the matrix was already adherent to the arachnoid, so, to remove the matrix, it was created a discontinuity of the arachnoid (*) with subsequent exit of liquor.

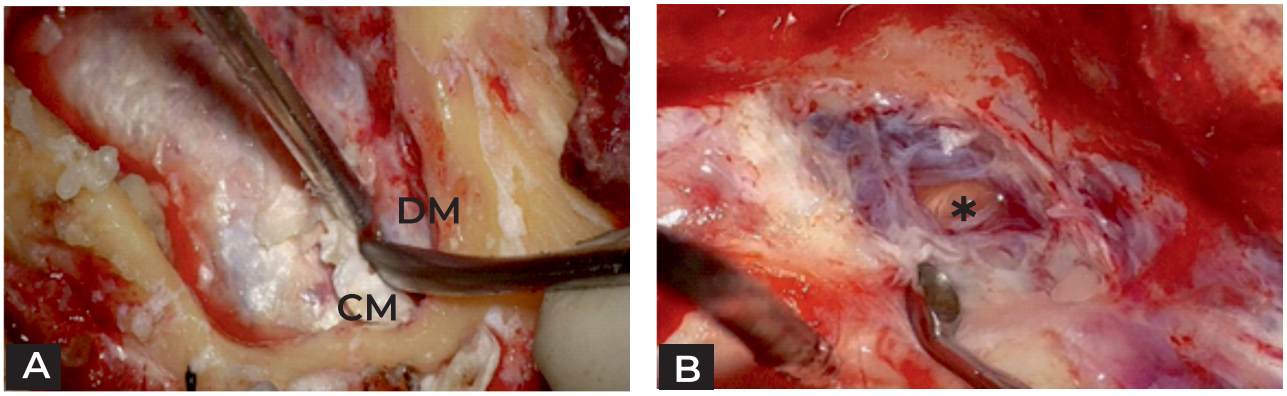


Figure 6
(A) The discontinuity created in the arachnoid has been obliterated by perichondrium (*); (B) The entire dura mater and arachnoid of the posterior fossa was covered with synthetic dura (SD), then fascia lata (FL), then fibrin glue.

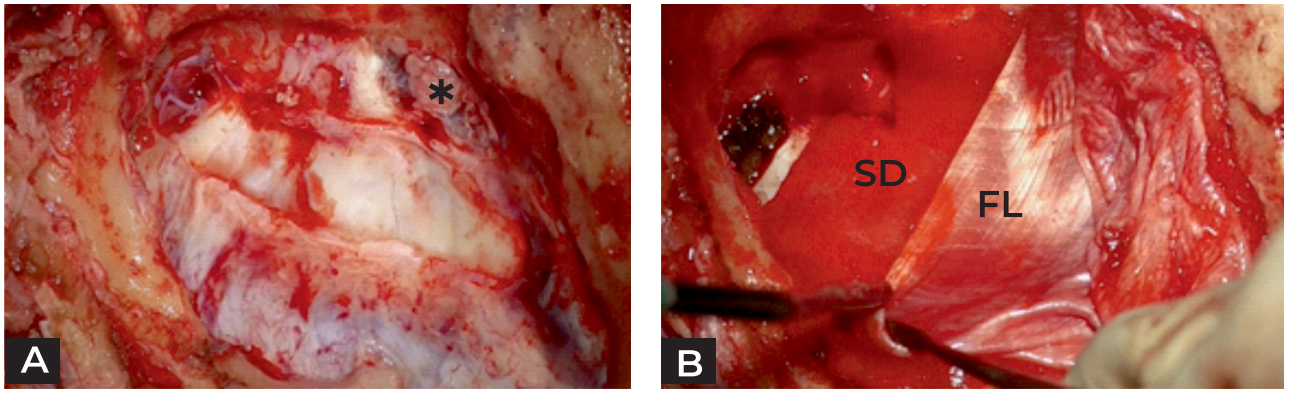
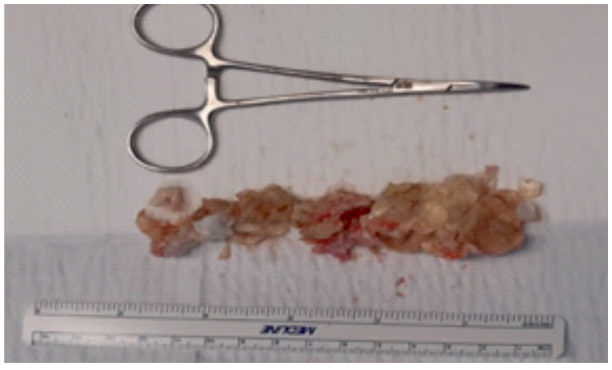


Figure 7
Gross appearance of the cholesteatoma



aware to infectious complications, since a failure to control them, can lead to potentially fatal complications, such as brain abscesses, epidural abscesses, meningitis, cavernous sinus septic thrombosis and subperiosteal abscess[2]. This case presented only with left ear discharge and hearing loss in a patient submitted to a cholesteatoma surgery in the referred ear 33 years before. According to Peter Dannatt *et al*, clinicians should be alert that the otoscopy findings of cholesteatoma and otitis externa can be similar. Therefore, in case of persistence of otitis externa despite adequate treatment, a cholesteatoma should be suspected[8]. This patient´s presentation of left otorrhea combined with otoscopic findings of a whitish discharge and swollen of the left external auditory canal, was initially thought to be otitis externa. However, lack of response to treatment, allied to a previous left ear surgery for cholesteatoma, raised the suspicion of a cholesteatoma. High resolution Computed Tomography (HRCT) is considered the gold standard in the diagnostic imaging

of cholesteatoma. In addition, it plays an essential role for surgical planning. This imaging modality has a high sensitivity with a high negative predictive value in excluding cholesteatoma when it shows a free, aerated mastoid and middle ear. However, its specificity is low, with a restricted ability to distinguish cholesteatoma from other lesions with a similar density such as effusion, granulation tissue and cholesterol granuloma. A soft-tissue expansive lesion, erosion of the ossicles, erosion of the tympanic tegmen and scutum blunting are typical findings related to cholesteatoma in CT scans. Magnetic resonance imaging (MRI) is a complementary tool that is considered better in evaluating changes in soft tissues. Cholesteatoma is commonly hypointense/ isointense on T1WI and hyperintense on T2WI. However, these signal-intensity characteristics are not specific for cholesteatoma. The diffusion weighted (DW) imaging sequences are considered more accurate to the diagnosis of primary and relapsing cholesteatomas. Echo-planar images (EPI) and non-echo-planar images (non-EPI) are two different DW-MRI sequences that can be used when evaluating cholesteatoma. There is evidence suggesting that non-EPI sequences have higher ability to identify recurrent or residual cholesteatoma and stronger reliability in the detection of small cholesteatomas. Non-EPI sequences are therefore useful to detect recidivism and used to complement preoperative CT scans[2,4]. The CT scan of this patient revealed a large erosive soft-tissue lesion with lytic destruction of the ossicles and imminent intracranial complication, since the lesion totally eroded the posterior fossa cortical and compressed the corresponding intracranial structures. MRI confirmed the diagnosis of cholesteatoma.

The mainstay of cholesteatoma treatment is surgery. The main goal of surgery is to eradicate the disease, that is, to create a dry, safe and recurrence-free ear[9]. To compare the different surgical techniques, it is first necessary to distinguish between residual and recurrent cholesteatoma. While residual cholesteatoma occurs due to the persistence of original disease, recurrent cholesteatoma is considered a new cholesteatoma that develops due to a mechanism similar to the initial disease, that is, from a retraction pocket. The term recidivism encompasses both, residual and recurrent cholesteatoma[10]. Our case reports a residual disease, since the tympanic membrane did not show any retraction pocket or perforation.

There are two main types of surgical techniques: canal wall up (CWU) and canal wall down (CWD) mastoidectomies. Some surgeons defend the CWU technique for its simpler post-operative care and maintenance. In this procedure, the preservation of original anatomy of the middle ear allows the cavity to get wet, reducing the impact in patient's quality of life, which is particularly important in younger patients. However, the meta-analysis performed by Julia Tomlin et al, showed that patients undergoing CWU surgery have nearly 3 times greater likelihood to develop recidivistic disease than those submitted to CWD. In fact, in this meta-analysis, the recidivism rate of CWU techniques ranged from 9-70% and for CWD varied only from 5-17%. Owing to the higher probably of recidivism after a CWU procedure, many authors recommend a staged or second look procedure 6-12 months after the first surgery, which can cause great suffering to patients, or a diffusion weighted magnetic resonance imaging. Because of the aforementioned disadvantages, other surgeons prefer the CWD procedure, which implies the removal of the posterior canal wall. This procedure is considered to be easier, requires less surgical experience and has lower recidivism rate than the CWU procedure. The disadvantages of creating a cavity in the CWD procedure include a lifelong necessity to clean the cavity, precautions to not get water in the ear, slower recovery and difficulties in fitting a hearing-aid[2,7,9]. This patient was submitted to a CWU procedure 33 years ago, which is associated with a higher recidivism rate than CWD procedure. The clinical follow-up performed for 27 years did not detect signs of recidivism. However, it was not performed

a second look surgery neither a magnetic resonance imaging in the follow-up period, which is currently recommended to decrease the recidivism rate of the CWU procedure.

Despite surgical treatment, there is a high rate of cholesteatoma recidivism and there is no consensus regarding the minimum duration for follow-up following cholesteatoma surgery.

It was verified that about 90% of recidivism occurred in the first 5 years of follow-up[11]. Nevertheless, there are several long-term studies that showed late postoperative occurrence of recidivism, up to 24 years following the surgery[2]. This case represents a late recidivism diagnosed 33 years after a CWU procedure, underlining the need of standardization in the length and methodology of the follow-up. We emphasize the importance of lifetime follow-up for patients submitted to CWU surgery due to chronic otitis media with cholesteatoma, for which it was not performed imaging with diffusion weighted magnetic resonance imaging or second look procedure.

Conclusion

Cholesteatoma's early recidivism after surgery is common. However, they can grow silently for years until they give rise to an intra or extracranial complication. Once diagnosed, the treatment is surgical. This case illustrates the importance of prolonged follow-up after CWU surgery for chronic otitis media with cholesteatoma, in cases where it was not performed a magnetic resonance imaging or a second look surgery.

Conflito de Interesses

Os autores declaram que não têm qualquer conflito de interesse relativo a este artigo.

Confidencialidade dos dados

Os autores declaram que seguiram os protocolos do seu trabalho na publicação dos dados de pacientes.

Proteção de pessoas e animais

Os autores declaram que os procedimentos seguidos estão de acordo com os regulamentos estabelecidos pelos diretores da Comissão para Investigação Clínica e Ética e de acordo com a Declaração de Helsínquia da Associação Médica Mundial.

Política de privacidade, consentimento informado e Autorização do Comité de Ética

Os autores declaram que têm o consentimento por escrito para o uso de fotografias dos pacientes neste artigo.

Financiamento

Este trabalho não recebeu qualquer contribuição, financiamento ou bolsa de estudos.

Disponibilidade dos Dados científicos

Não existem conjuntos de dados disponíveis publicamente relacionados com este trabalho.

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