

Recurrent Cholesteatoma Invading the Internal Auditory Canal and Cerebellar Pontine Angle

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Abstract

Cholesteatoma is a fairly otologic common problem. However, cholesteatoma invading the internal auditory canal (IAC) is rare and typically results in profound hearing loss and facial paralysis. This is a case of a 46-year-old female with a history of prior right complex cholesteatoma that had undergone multiple procedures. She had multiple complications including right cerebral spinal fluid (CSF) leak, meningitis, recurrent mastoid bowl infections and right facial paralysis which resulted in multiple facial plastics procedures and over-closure of the right ear. Over the last three years, she has noticed an increase in right sided otalgia, facial pressure, facial numbness and headaches. An MRI temporal bone with diffusion weighted imaging (DWI) showed a DW positive soft tissue mass filling the mastoid bowl as well as extending into the IAC and cerebellar pontine angle (CPA) cistern with contact of the middle cerebellar peduncle and trigeminal nerve. A translabyrinthine approach to the IAC found the mastoid bowl to be filled with cholesteatoma and an osseous defect from the mastoid bowl along the labyrinthine facial nerve tracking cholesteatoma into the IAC/CPA. This case highlights the complex and aggressive nature a cholesteatoma can take and the need for diligent surveillance in any ear that had prior cholesteatoma. The utilizations of MRI temporal bone with diffusion weighted imaging allow for surveillance in an over closed ear canal that is vital to the care of cholesteatoma patients who have a similar history.

Keywords

Translabyrinthine Approach, Recurrent Cholesteatoma, Cranial Base, Otology/Neurotology

1. Introduction

Cholesteatoma, or more specifically acquired cholesteatoma, is a well-known

cystic lesion of the ear caused by either perforation or eustachian tube dysfunction that seeds epithelium into the middle ear and mastoid [1]. Bony erosion is usually confined to the ossicular chain and scutum at least initially but over time can cause erosion of the otic capsule, fallopian canal and tegmen. Tegmen erosion often leads to cephalocele or cerebral spinal fluid (CSF) leak. Extensive cholesteatomas are more likely associated with invasion of sigmoid and jugular bulb and intracranial abscesses. The decision for blind sac closure is also raised in these large cholesteatomas [1]. This can lead to increased meningitis risk causing morbidity and mortality. In the setting of chronic infections, the erosive osteolytic effect of cholesteatoma can be accelerated leading to more bony erosion than expected [2]. When cholesteatoma involves erosion of the bony labyrinth and IAC, there is a threat to all functions housed in the ear. IAC invasion of cholesteatoma is rare and initiates from either acquired giant cholesteatomas or from congenital cholesteatoma that develop close to the IAC [3]. There is a known classification system in the temporal bone based on location of cholesteatoma. It is as follows: supralabyrinthine cholesteatoma, infralabyrinthine cholesteatoma, retrolabyrinthine cholesteatoma, diffuse massive cholesteatoma and apical cholesteatoma [4]. Sana *et al.* has reported that there are three possible routes for IAC invasion of cholesteatoma. Above the labyrinth or supralabyrinth is the first, or through the supralabyrinthine mastoid cells, geniculate ganglion and cochlea, invading the superior anterior IAC. Lastly, it could also course below the labyrinth, the infralabyrinthine route where it extends via the retrofacial cells and vestibule into the posterior IAC [5]. Others have detailed a simpler version of this as either supra- or infra-labyrinthine. Often patients with cholesteatoma involving the IAC have invasion through the supralabyrinthine route. In these cases, retrospective analysis has shown a high incidence of facial nerve paralysis and profound deafness with acquired cholesteatomas being the most common etiology [3]. This is a case of a patient with an aggressive supralabyrinthine spreading cholesteatoma requiring multiple revisions procedures that led to a CSF leak causing the need for over closure of ear canal making surveillance more difficult. This highlights the importance of close follow-up on recurrent aggressive cholesteatoma and the need for close MRI with DWI follow-up in cholesteatoma.

2. Case Report

This is a case of a 46-year-old female with a history of prior right complex cholesteatoma that had undergone multiple procedures for removal of cholesteatoma first diagnosed in 2001. Informed consent was obtained from the patient for this case. She was diagnosed with a right sided cholesteatoma and had an initial canal wall up tympanomastoidectomy in 2001. About 9 months later a second look procedure was planned and there was recurrent cholesteatoma found and canal wall down tympanomastoidectomy was completed with ossiculoplasty in 2002 with mild improvement in hearing. Over time she noticed worsening hearing and right facial weakness that was slow and progressive. In 2014 more cholesteatoma was

detected on MRI temporal bone DWI and a petrosectomy was planned. After this third surgery she was a House-Brackmann 6/6 on the right with no facial function and she had no serviceable hearing on the right ear. In 2015 she had a bone anchored hearing aid (BAHA) connect implanted on the right. On 12/11/2017 she was taken to operating room for intermittent CSF otorrhea and recurrent mastoid bowl infections for subtotal petrosectomy revision, abdominal fat graft for obliteration of mastoid cavity and overclosure of ear canal. This decision which was made by her prior Neurotologist was perhaps made due to the CSF leak and meningitis history she had. Carefully weighing the benefits and disadvantages an over closed ear canal has in a patient with a cholesteatoma history is one decision that should not be made lightly. The closure adds the disadvantage of inability for direct surveillance and cleaning but lowers the risk of CSF leak and meningitis dramatically. She underwent gold weight and cross facial reanimation and gracilius muscle flap in the facial plastics department from 2019 to 2020. She recently noticed increased right sided otalgia, headaches, and facial numbness over the last three years. An MRI Brian with and without showed increase signal in the mastoid cavity and in the IAC/CPA. When her Neurotologist did not think this was concerning, she wanted a second opinion.

On presentation to our clinic in March 2024 she had a well healed right BAHA, an over closed right ear canal and good right eye closure with her gold weight and good facial tone in the lower branches of her face. She described 7-8/10 otalgia on right side of head and her numbness on all branches of the right trigeminal nerve. An MRI temporal bone with diffusion weighted imaging (DWI) was ordered to give more detailed view of the area involved. A soft tissue mass that was DW positive was found filling the entire mastoid bowl as well as the IAC extending into the cerebellar pontine angle (CPA) cistern with contact of the middle cerebellar peduncle and trigeminal nerve (**Figure 1(A)** & **Figure 1(B)**). On CT non-contrast temporal bone opacification of the entire mastoid bowl with an obvious linear osseous defect connecting the middle ear and mastoid to IAC widening it from the mastoid bowl along the geniculate and labyrinthine segment of facial nerve (**Figure 2**). This is a common finding in cholesteatoma spreading into the IAC along the supralabyrinthine route [3]. There was also obvious multifocal dehiscence of the tympanic facial nerve in the tympanic and mastoid segments.

A combination translabyrinthine approach with Neurotology and Neurosurgery was planned in August 2024. The mastoid bowl from prior canal wall down approach was filled with cholesteatoma and an osseous defect from the mastoid bowl along the tympanic facial nerve which showed keratin tracking along its course and was found to have cholesteatoma tracking onto the labyrinthine facial nerve into the IAC/CPA. This was a case of supralabyrinthine spread to the IAC. Complete removal was accomplished by careful dissection of keratin using an array of sizes of round knives and utilizing a 4.0 mm, through 2.0 mm coarse and fine diamond burs to drill through the labyrinth to the roof of the IAC to find keratin filling the IAC (**Figure 3**). The Neurosurgery team then utilized soft tissue

removal with preservation of nerves within the IAC and CPA. The cochlear vestibular nerve had no visible keratin invasion and was intact at the end of the procedure. Once complete removal of keratin from the IAC and CPA was accomplished DuraGen © was used to repair the dura. All air cells were waxed off and middle ear and eustachian tube was obliterated with temporalis muscle and Surgicel © and an abdominal fat graft was completed to fill the bony defect. The patient had an uncomplicated post-operative course with a three-night hospital stay with preservation of her facial function as expected and transient vertigo that was brief and only one episode and no signs of CSF leak. She is scheduled for follow up visit for removal of sutures post op day 10 and repeat MRI DWI in 6 months.

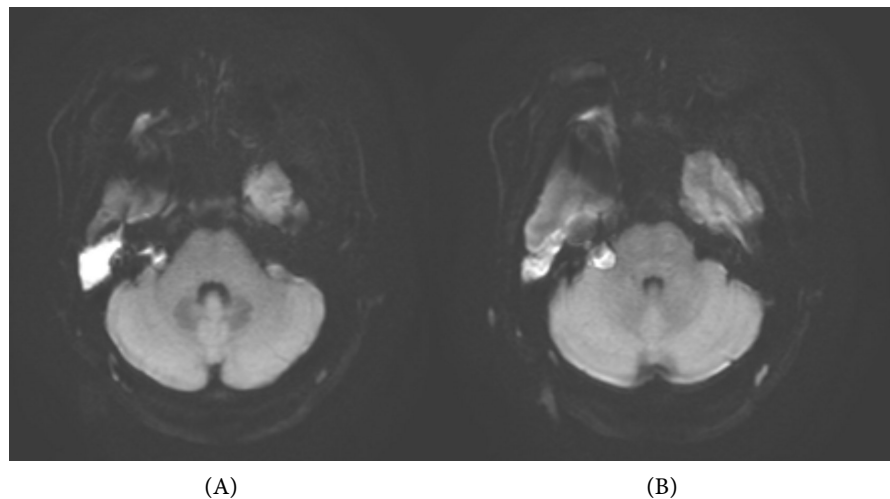


Figure 1. Axial Image of MRI Temporal Bone with diffusion weight imaging. A mass appears to fill the right mastoidectomy bowl and residual middle ear. The lesion demonstrates avid restricted diffusion as well as peripheral enhancement. The lesion measures approximately $17 \times 25 \times 21$ mm. **A. (Left)** Avid restricted diffusion in right mastoid bowl into the IAC. **B. (Right)** Restricted diffusion into the CPA abutting the cerebellar peduncle.

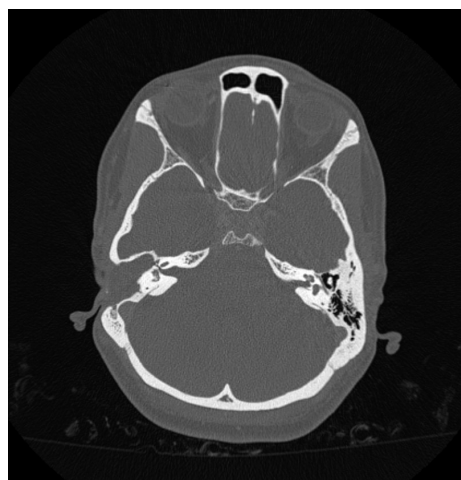


Figure 2. CT temporal bone axial view with opacification of right mastoid bowl with possible bony erosion and path along tympanic facial segment and geniculate ganglion with slight widening of labyrinthine facial and into the right IAC.

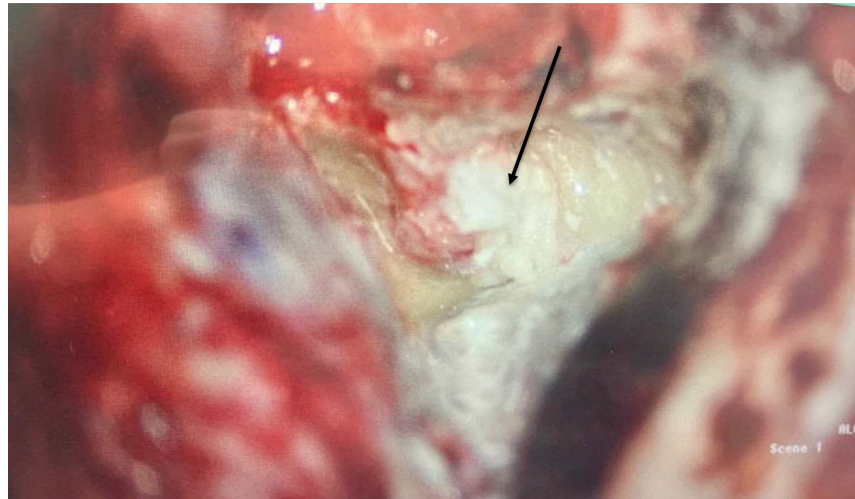


Figure 3. Intra operative microscopic image of keratin invading right IAC and CPA once translabyrinthine approach was completed with all bone removed from middle fossa plate, pre-sigmoid dura, and over the IAC and CA. Left is superior, Right is inferior. Arrow points to keratin in the IAC.

3. Discussion

This case presented an occurrence of extension of cholesteatoma into the IAC/CPA causing otalgia, facial pain and numbness. She already had a profound hearing loss and facial paralysis. One of the more common routes of spread of large cholesteatomas is the supralabyrinthine route along the geniculate and labyrinthine facial nerve as what is seen in this patient. The classification of temporal bone cholesteatomas spreading is described early by Sana *et al.* [5]. Often when the IAC is invaded all functions of the ear have been sacrificed already including hearing, balance and facial nerve function. The patient here had a history of multiple surgical procedures to exteriorize and eradicate the cholesteatoma with subsequent CSF leak after these procedures. Utilizing DWI MRI imaging has shown success in detection of cholesteatomas in ears that have been closed. Cholesteatoma has a very specific signal-intensity on MRI images with very high signal intensity on DWI. In a systemic review of DWI in the assessment of postoperative cholesteatoma a combined sensitivity of 91.4% and positive predictive value of 97.3% was calculated in non-EPI DWI images of MRI. Non-EPI-DWI is a specific sequence of DWI that is known to lack significant image distortion and less artefacts than EPI-DWI [6]-[8]. Meticulous follow up would be necessary. Careful surgical dissection and exposure of all areas where cholesteatoma is necessary during surgical exploration. Further research and larger studies are needed to evaluate best management for patients with extensive cholesteatoma and their complications.

The decision to close the ear canal likely is a difficult one since the ability to surveillance the ear and remove recurrent or recidivistic disease is more difficult. It is hard to say the volume of the CSF leak and if that was indeed necessary in this case. Studies have shown that the majority (80% - 90%) of patient that have had ear canal closure with a history of cholesteatoma have recidivistic disease that

needs further surgical intervention [9] [10]. Approximately 60% of also have extensive disease that was detected with either planned second look surgery or with DWI. Planned second look or closure follow up with MRI DWI is recommended. If possible, we propose that repair of the tegmen defect and CSF leak should be attempted with autologous tissue and the ear canal could have been kept patent or this could have staged if healing and meningitis risk was a concern. This patient had her ear closed for 7 years and a large volume of keratin was found filling her surgical defect. This burden of disease likely leads to the invasion medially into the IAC.

4. Conclusion

This is a case presentation of complex recurrent cholesteatoma that required translabyrinthine surgical approach for removal of cholesteatoma that invaded the IAC/CPA after closure of ear canal. This case highlights the importance of diligent MRI DWI surveillance of over closed ears that have a history of cholesteatoma and the need to avoid permanent over closure of ears with a large burden of cholesteatoma with high risk of recurrence.

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Conflicts of Interest

The authors report no conflicts of interest.

Institutional Review Board

This submission was reviewed by the Wake Forest School of Medicine Institutional Review Board and determined to not involve human subjects research with memorandum attached.

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