Case study

Dual Cholesteatoma in a non-operated ear - A rare presentation

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Abstract

Objective

We report an interesting case where two cholesteatomas were found simultaneously in a non-operated ear. The presentation, management and implications for other patients with cholesteatoma are discussed.

Case Report

A 49-year-old male with a unilateral mixed hearing loss was found to have two cholesteatomas. Only one was suspected clinically, in the anterior attic, the other a large congenital cholesteatoma in the mastoid tip as an incidental finding on non echo-planar diffusion weighted image (Non-EPI DWI) MRI scan. The radiological findings were confirmed at surgery (combined approach tympanoplasty).

Conclusion

Congenital cholesteatoma in the mastoid process is extremely rare and multiple cholesteatomas occurring simultaneously in a non-operated ear is also rare. Non-EPI DWI MRI is increasingly used as a non-invasive technique for detecting recurrent or residual cholesteatomas post-operatively. This case supports extending the role of non-EPI DWI MRI scanning pre-operatively in primary disease to exclude additional cholesteatomas and to assess the suitability of permeatal endoscopic surgery.

Introduction

Cholesteatomas are non-cancerous lesions which can occur in the middle ear , mastoid[1] and the external auditory canal (EAC). Cholesteatomas are classified as being congenital or acquired. Histology confirms them as well-defined necrotic masses of squamous epithelium with considerable keratinisation[2]. Cholesteatoma is an uncommon condition and epidemiological evidence[3] suggests an incidence rate of 9.2 per 100,000 population per year in Northern Europe. Congenital cholesteatoma accounts for 2-5% of all cholesteatomas[4]; the most common site is in the middle ear, the rarest site being in the mastoid process, comprising only 3-4% of congenital cholesteatomas[5].Multiple foci of cholesteatoma are identified in previously operated ears but the presence of multiple cholesteatomas in non-operated ears is rare.

Keywords: Cholesteatoma, DWI Non-EPI MRI Temporal bones, Endoscopic Ear Surgery (ESS)

Case Report

A 49-year-old South-Asian male presented with history of a hearing loss which was discovered during a screening hearing test at his opticians. He had been struggling with conversations with background noise as well as localisation of sounds but had developed coping strategies. There was no history of otalgia, discharge, tinnitus and vertigo or loud noise exposure was recorded. The

patient had Asperger's Syndrome and was on sodium valproate for a Bi-Polar disorder. On examination the right ear was normal but left ear revealed an anterior attic retraction pocket with possible cholesteatoma. No discharge or facial palsy were present.

Audiometry was normal in the right ear but the left ear had a mixed hearing loss with bone conduction of 5-35dB and air conduction of 30-65dB. Tympanometry was normal.

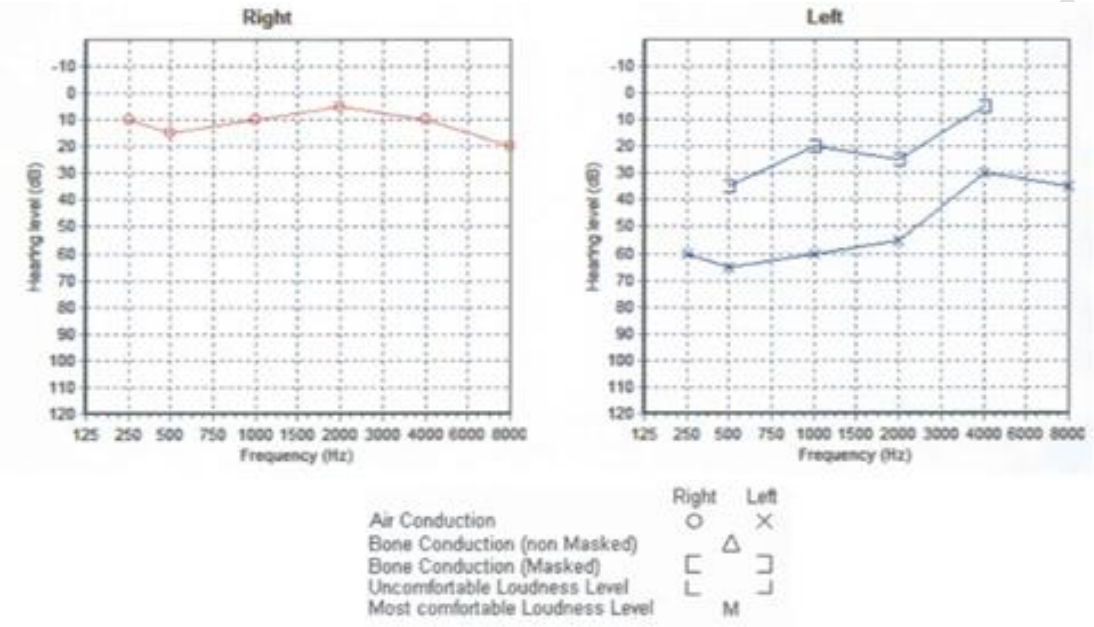

Figure 1- Pre-operative Audiogram

MRI scan of the internal auditory meatus and cerebellopontine angle was arranged to exclude a vestibular schwanomma as a cause of the asymmetrical hearing loss and was normal. In view of the anterior attic abnormality , a non-EPI DWI MRI of the temporal bones **(Fig 2)** was requested which confirmed an anterior attic cholesteatoma. However the scan also demonstrated a large cholesteatoma in the mastoid tip. The subsequent CT scan of the Temporal bones **(Fig 3)** showed areas of soft tissue opacification within the anterior attic and a separate focus in the mastoid tip on the left side corresponding to the areas of restricted diffusion on the MRI scan.

The patient made a good recovery from surgery (Combined Approach Tympanoplasty) where the radiological findings were confirmed reconfirmed. Post op follow up 2 month audiogram demonstrated improvement in the conductive element of the hearing with closure of the air-bone gap.

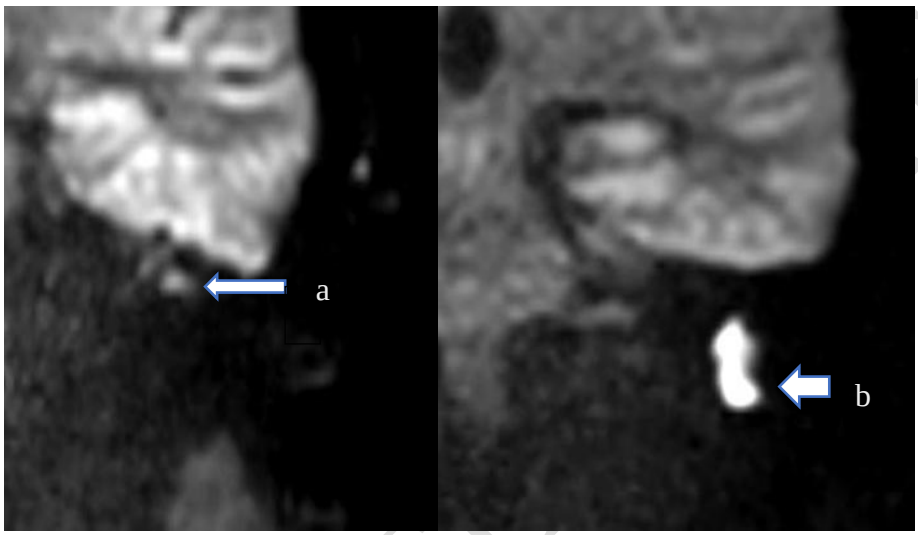

Figure 2- Non-EPI DWI MRI Scan of the temporal bones showing the presence of two cholesteatomas (a-attic and b-mastoid tip) on the left side

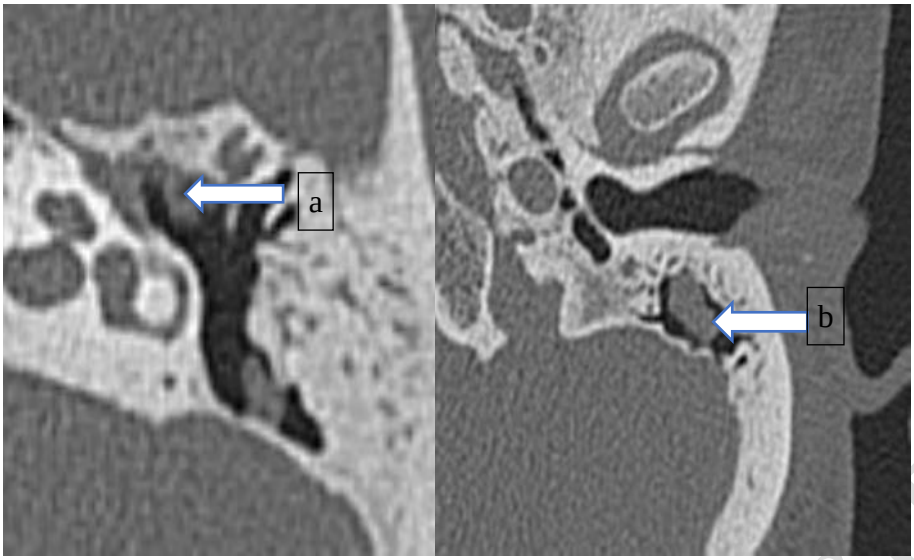

Figure 3- CT Scan with left sided areas of opacification (a-attic and b-mastoid tip)

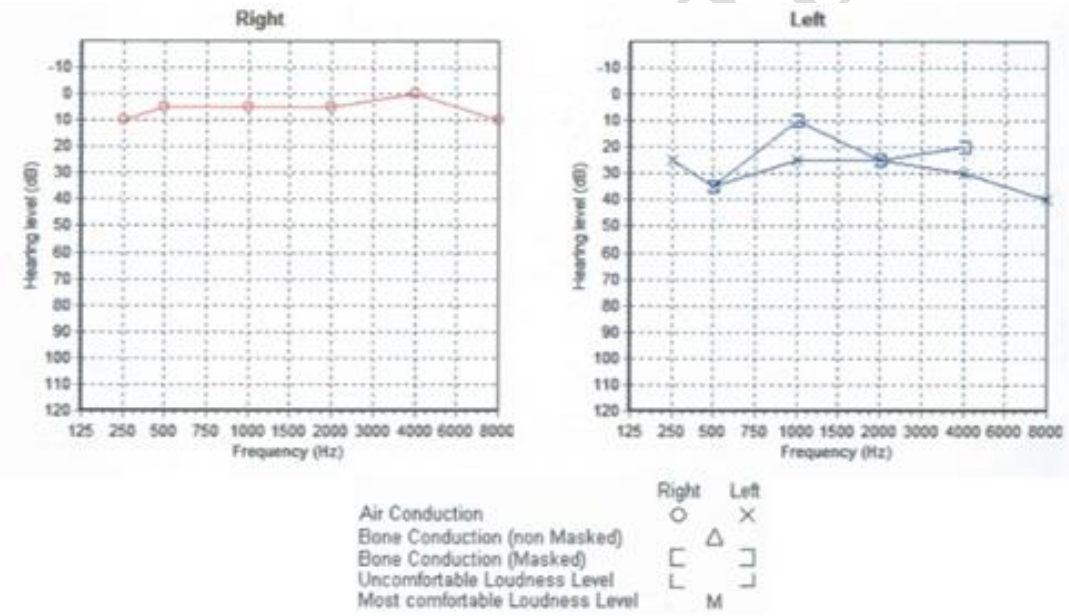

Figure 4- Post-operative Audiogram

Discussion:

Congenital cholesteatoma of mastoid origin has been described in a few case reports in the literature with some patients having symptoms of otalgia, retroauricular swelling, dizziness and narrowing of the ear canal. Warren et al[6] described 9 cases of congenital cholesteatoma in the

mastoid temporal bone and the most common presentation was, as in our case, an incidental finding on radiological examination.

The presence of a second site cholesteatoma in a non-operated ear is rare .Non-EPI DWI MRI has revolutionised the post-operative follow-up[7] and is now the imaging modality of choice in the detection and localisation of recurrent and residual cholesteatomas by providing a non-invasive alternative to surgery. The diffusion-weighted technique is very specific due to the characteristically high keratin content of cholesteatomas[8]. These show up as areas of high signal intensity whereas other structures generate lower signals. The non echo-planar sequences increase sensitivity by generating imaging slices as thin as 2mm and therefore, they can detect cholesteatomas just as small[12]. Subsequently, when used in conjunction they form a powerful tool for assessing disease post-operatively. It is favored over CT scanning for its specificity and ability to avoid excessive radiation exposure. High resolution CT scanning can provide valuable information on the anatomy of the temporal bone but it is poor at differentiating a cholesteatoma from inflamed and granulomatous tissue, depicting both as non-specific areas of opacification[9].

Patients having a large modified radical mastoidectomy (MRM) or combined approach tympanoplasty (CAT) where the mastoid cavity is fully explored are less likely to have a second cholesteatoma left undetected. In recent years the gaining popularity of endoscopic ear surgery (EES) to remove cholesteotoma limited to the middle ear and mastoid (anterior to the posterior limit of lateral semicircular canal) may necessitate the use of Non EPI DWI MRI scanning prior to primary surgery.

There is however, less literature available on the use of non-EPI DWI MRI scanning pre-operatively to accurately diagnose the primary disease. In our case, it was fortuitous that this sequence was requested at the same time as the MRI scan to investigate the asymmetric hearing loss, otherwise

the cholesteatoma in the mastoid tip would have been missed if a limited endoscopic removal of the attic cholesteatoma was performed.

A meta-analysis published in 2013[10] reviewed 10 studies (total of 342 patients) and concluded that non-EPI DWI MRI is an extremely accurate technique to correctly identify the presence of cholesteatoma with sensitivity of 0.94 (confidence intervals of 0.8-0.98) and specificity of 0.94 (confidence intervals of 0.85-0.98). This shows that the use of this technique in primary pre-operative cases is justified when a diagnosis of cholesteatoma is in question.

Many other studies show a strong correlation between the presence, extent and location on pre-operative scanning (using non-EPI DW imaging) and the findings at surgery[9]. This confirms that it can accurately predict the disease in primary cases and help to plan an endoscopic approach for surgery. Prior to surgery non-EPI DWI Scans positively identified cholesteatoma in 27 patients with the primary disease and in 23 patients with residual or recurrent disease. The results were 98% precise in portraying the localisation and extension of disease (49/50 cases) with the sole exception being an overestimation of disease number in a patient who had previous surgery for cholesteatoma. These were deemed crucial factors in influencing what surgical approach to use. It was found that an endoscopic transcanal technique was better suited for lesions smaller than 8mm and just located in the middle ear or its extensions. An endoscope-assisted retroauricular mastoidectomy was the technique of choice for bigger lesions.

Conclusion

Multiple site cholesteatomas occurring simultaneously in a non-operated ear has rarely been reported in the literature. Non-EPI DWI MRI scans are established as a means to detect cholesteatoma post operatively but with the evolving field of Endoscopic Ear Surgery (EES) there

has been a paradigm shift towards using Non-EPI DWI MRI scans in addition to CT scans of the temporal bones in the preoperative management of cholesteatoma.

References

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