CASE REPORT

Lyric hearing aid: a rare cause of benign necrotising otitis externa/external ear canal cholesteatoma

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SUMMARY

An 80-year-old Caucasian man presented with an incidental and asymptomatic lesion in his right ear thought to be secondary to his use of hearing aids for presbycusis. He used Lyric hearing aids, designed for 24 hours-a-day use for 4 months at a time and had no other previous otological problems. He underwent a bony meatoplasty and vascular flap reconstruction via a retroauricular approach to remove the lesion for histological analysis and regrafting of the area. The lesion was confirmed on histopathology as an ear canal cholesteatoma.

BACKGROUND

In a recent case series, external ear canal cholesteatoma (EECC) (also known as benign necrotising otitis externa (BNOE)) was associated with hearing aid usage in 9% of patients. This rare but often silent condition can be associated with long-standing otorrhoea (65%), dull otalgia (12%), conductive hearing loss >20Db (27%) and also invasion/destruction of associated structures such as the temporomandibular joint (TMJ), mastoid, middle ear and facial nerve.¹ We believe the effect of a hearing aid designed for continuous use for 4 months may lead to malperfusion and bone hypoxia, making EECC an even greater risk.

We report this case with a literature review to raise awareness in audiologists and doctors of a complication of in-the-canal (ITC) hearing aid use, EECC and its differential diagnoses.

CASE PRESENTATION

A fit and well 80-year-old man was referred to ear, nose and throat following routine audiologist review for bilateral Lyric hearing aid renewal. On initial examination, a lesion in the right external auditory canal (EAC) was noted. He had worn bilateral Lyric hearing aids (designed for continuous use for 4 months at a time) for many years for presbycusis without problem (figure 1). He had no other otoneurological symptoms, no risk factors for EECC and was otherwise fit and well.

Otoneurological examination including microscopic assessment revealed a large localised erosion of the anteroinferior aspect of the bony canal (figures 2 and 3). This was difficult to debride fully via microsuction in clinic. There was no facial weakness or cervical lymphadenopathy.

INVESTIGATIONS

Pure tone audiogram demonstrated a bilateral moderate high-frequency sensorineural hearing loss consistent with bilateral symmetrical presbycusis.

DIFFERENTIAL DIAGNOSIS

- ▶ EECC
- ▶ Keratosis obturans
- ▶ Otitis externa
- ▶ Squamous cell carcinoma (SCC) of the EAC

TREATMENT

The findings, differential diagnoses and options for treatment were discussed with the patient. One surgical treatment option was biopsy for diagnosis and simple debridement under local anaesthetic (which carried the risk of recurrence and the need for further management).

The other option was biopsy for diagnosis and bony canal meatoplasty with an inferiorly-based soft tissue flap to cover the defect and reline the EAC. This would be via a retroauricular approach under general anaesthesia and would offer a more definitive solution. After consideration of the risks/benefits associated with each, the patient opted for the latter.

OUTCOME AND FOLLOW-UP

The patient underwent an uncomplicated procedure and recovered well. He was warned to avoid using ITC and completely-in-the-canal hearing aids in his right ear in future.

Figure 1 Diagram illustrating the recommended position of a Lyric hearing aid in situ within the external auditory canal.

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Figure 2 Endoscopic image of the right external auditory canal and tympanic membrane, prior to suctioning/debridement. Note the presence of a lesion in the anteroinferior portion of the canal, obscuring the view of the anterior portion of the tympanic membrane.

The pathology sent from the operation was confirmed as benign, with findings consistent with cholesteatoma.

During his 12 months of follow-up, the canal has remained epithelialised, with no recurrence of cholesteatoma. This is despite use of a conventional air conduction hearing aids during this period.

DISCUSSION

Cholesteatoma is characterised by a cystic mass containing keratinised stratified squamous epithelium and cholesterol deposits, with associated periostitis and bone erosion. The middle ear is most commonly affected, with lesions further defined as either congenital (2%) or acquired (98%).^{2,3}

Figure 3 Endoscopic image of the right external auditory canal and tympanic membrane. The lesion is shown to be undergoing suction and debridement. The presence of keratinous debris within the lesion can be appreciated.

EECC, also known as BNOE, is a rare otological entity affecting the EAC. It accounts for around 0.1% to 0.5% of all new otological presentations. It has been shown to affect both sexes equally and the average age of presentation to be in the sixth decade of life, although it is also commonly seen in younger adults.⁴

It most commonly affects the inferior and posterior aspects of the EAC, and on histology, it has been shown to have features of focal skin disruption, accumulated keratinous debris, associated bone erosion and bone sequestration. If left untreated, in addition to long-standing otorrhoea and otalgia, it can progress to exposure and erosion of the TMJ, mastoid, middle ear and facial nerve.^{1,2}

EECC can be subdivided into primary (spontaneous) or secondary (acquired). Although the aetiology of secondary is well documented when compared with primary EECC, the aetiology of each still remains subject to debate. Recognised causes in secondary EECC include post-surgical epithelial changes, inflammatory processes, postradiation treatment and following acute or repeated trauma. One of the most widely accepted mechanisms is thought to be due to malperfusion and bone hypoxia following sustained pressure exerted on the EAC from foreign objects such as hearing aids or cotton buds.⁵ Traditional views of formation following disruption in the epithelial lining of the EAC leading to impaired epithelial migration and subsequent keratin accumulation have been recently discredited in a case-control paper by Bonding and Ravn in 2006.⁶

In our case, the possible aetiology of malperfusion and bone hypoxia of the EAC secondary to Lyric hearing aid usage is in keeping with cases reported in the literature. A review of 34 cases in a tertiary centre from 1994 to 2006 reported six patients (9%) who developed EECC following hearing aid or in-ear hearing protection use,¹ and a meta-analysis in 2010 further suggested the presence of an in-ear hearing aid as a possible cause of EECC.⁵ We believe that the effect of a hearing aid designed for continuous use for 4 months at a time may well lead to malperfusion of the EAC and bone hypoxia, making EECC even more likely.

The consideration and subsequent exclusion of important differentials is also imperative. Perhaps the most important of these is EAC SCC, which can be diagnosed by biopsy and histological examination. Due to the prognostic implications of SCC, differentiation is important for the planning of further management and follow-up—such as local surgical clearance of the primary tumour, neck dissection and associated oncological

Learning points

- ▶ Hearing aid usage is a recognised cause of external ear canal cholesteatoma (EECC).
- ▶ Hearing aids designed for continuous use for 4 months at a time may cause malperfusion and bony hypoxia possibly making EECC more likely.
- ▶ EECC can present with long-standing otorrhoea, dull otalgia and hearing loss and can progress to exposure of temporomandibular joint, mastoid, middle ear and facial nerve.
- ▶ Differential diagnosis includes keratosis obturans, otitis externa and squamous cell carcinoma, thus referral for biopsy and subsequent management is required.

management. However, biopsy for histology remains the gold standard of initial treatment for *both* SCC and EECC.⁷

Another differential to consider is keratosis obturans, as EECC/BNOE is often misdiagnosed as this. However, EECC can be differentiated by the presence of focal osteonecrosis or sequestration of bone lacking an epithelial covering, infrequent hearing loss and if otalgia is present, it is more likely to be characterised as dull in nature. It is important to differentiate between the two as management may differ depending on diagnosis.^{8 9}

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