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Case Report

External Auditory Canal Cholesteatoma - A Rare Disease

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Abstract

Cholesteatomas of external auditory canal are a form of invasive keratitis characterised by accumulation of desquamated debris in the wall of canal. External ear canal cholesteatoma is a rare condition with an estimated incidence of 1.2 in 1,000.

63-year female presented to us with complaints of left ear pain and left ear discharge. Otoscopy was normal. Pure tone audiometry showed mild mixed hearing loss in left ear.

Preoperative CT scan was done. It shows the defect in external auditory canal. Canal wall down mastoidectomy with type 3 tympanoplasty was performed. Cholesteatoma sac was seen and was removed completely. Histopathology confirmed diagnosis of cholesteatoma.

Primary auditory canal cholesteatoma is the invasion of squamous tissue from ear into localised area of bony erosion. The aetiology is uncertain, but in some of the series primary auditory canal cholesteatoma is post-traumatic or postsurgical. Diagnosis is confirmed on CT scan. External canal cholesteatoma is a rare condition and it poses challenge in diagnosis as it can be missed easily.

Keywords: Cholesteatoma; Auditory Canal; HRCT; Histopathology

Introduction

External auditory canal cholesteatoma (EACC) is a rare condition. Its incidence is 1.2 per 1000 new otological patients [1]. Compared to the incidence rate of middle ear cholesteatoma which is 9.2 per year per 100,000, the incidence of primary EACC is 0.30 per 100,000 population [2]. EACC was first described by Toynbee in 1850 [3]. Here we will discuss clinical, radiological, preoperative finding and management of such a case with a primary EACC in a female which was managed by modified radical mastoidectomy.

Case Report

A 63-year-old female came with left ear earache, dull aching and left discharge since one year. Ear discharge was insidious in onset, gradually progressive, scanty in amount, occasionally blood tinged and foul smelling. No history of reduced hearing or tinnitus. No history of trauma. Family history was not significant.

On otoscopic examination scanty and foul smelling discharge was present over the floor of left external auditory canal. Tympanic membrane was dull and intact. On tuning fork tests, Rinnes was negative for 256 Hz and positive for 512, 1024 Hz. Webers was lateralised to left and ABC was reduced on left. Facial nerve examination was normal and nose and throat examination was normal.

Initially patient was treated symptomatically with medical line of treatment but as she didn't improved, we did all routine blood investigations which were normal.

Patient was subjected to pure tone audiometry which showed mild mixed hearing loss in left ear while right ear was normal. Otoendoscopy showed bony erosion over the posterior wall of EAC. HRCT temporal bone axial cuts were showing intact TM, bony erosion in the posterior wall of external auditory canal, soft tissue den-

sity mass involving left side mastoid air cells as well as anterior attic area and ossicles were intact and normal (Figure 1 and 2).

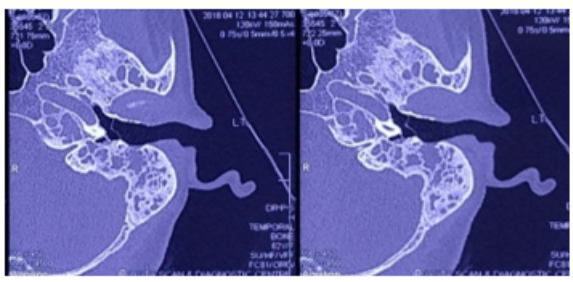

Figure 1

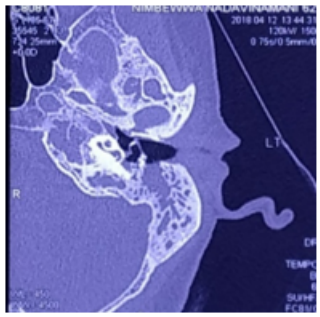

Figure 2

Patient was taken up for surgery. Postauricular Wildes incision was taken and pinna was retracted forwards. Using ball point probe we judged the depth of bony erosion of posterior wall. We entered middle ear as CT showed soft tissue density filling the anterior attic area (Figure 3). All the cholesteatoma was cleared from anterior attic area and mastoid.

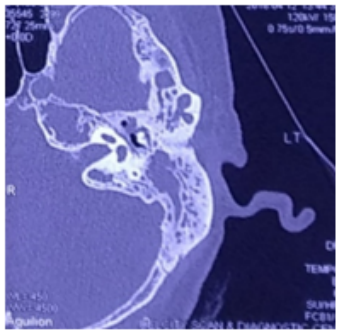

Figure 3

Canal wall down mastoidectomy was performed using inside out technique and cholesteatoma sac was cleared from posterior bony wall of EAC. Incus was displaced and removed to clear the cholesteatoma. Wide clearance of the squamous debris and a cholesteatoma sac was done, and the irregular eroded bone was saucerized by diamond drill until normal healthy bone was found (Figure 4). Myringostapediopexy was performed.

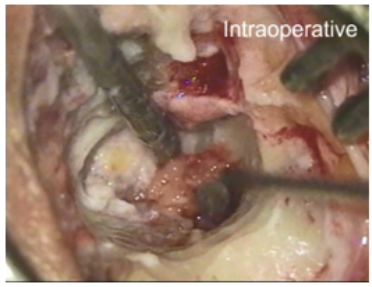

Figure 4

A temporalis fascia graft was used to cover the exposed bone before repositioning the meatal skin. The ear canal was packed with gel foam soaked with antibiotics.

Histopathology confirmed the diagnosis of cholesteatoma (Figure 5).

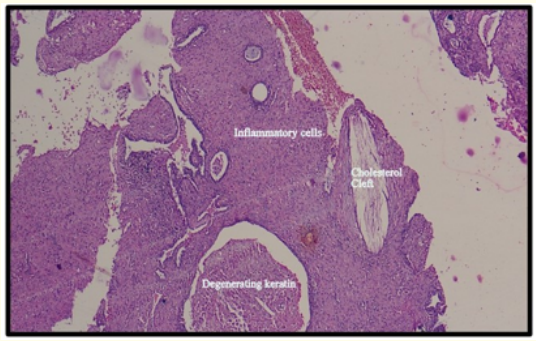

Figure 5: Pathologic findings of hematoxylin and eosin stain showing a squamous epithelium-lined cyst that is filled with fully differentiated, anucleated, laminated masses of keratinous material and cholesterol cleft as well.

Postoperatively patient is on regular follow-up and no recurrence is seen yet.

Figure 6 showing postoperative follow-up of 2 year with no recurrence.

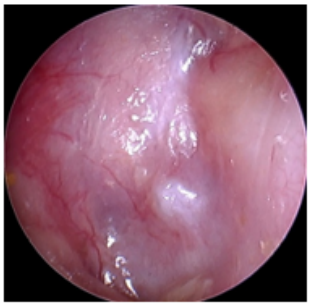

Figure 6

Discussion

Cases of the EACC and keratosis obturans have been reported as early as 1850 by Toynbee [3] and in 1893 by Scholefield [4]. The term keratosis obturans was coined by Werden in 1874 [5]. EACC had been confused with keratosis obturans, and it was distinguished by Piepergerdes., *et al*. in 1980 [6,7]. EACC occurrence is rare. This may be a reason why it is traditionally considered together with keratosis obturans.

Keratosis and EACC are two different clinical and pathological conditions. In EACC, the presenting symptoms are intermittent or persistent otorrhea and a dull, chronic otalgia secondary to an invasion of squamous tissue into a localized area of periostitis in the canal wall. Severe otalgia and hearing loss is seen in keratosis secondary to the accumulation of large plugs of desquamated keratin in the ear canal. In EACC cases , tympanic membrane is normal so hearing loss doesn't occur and the cholesteatoma erodes into the canal wall, not into the middle ear or attic [7]. Rarely does it leads to hearing loss which was seen in our case.

The classification based on the histology and clinical symptoms of the patients was first reported by Naim., *et al*. in 2005 [8]. He classified EACC into four stages: stage I with hyperplasia of the canal epithelium, stage II including periostitis, stage III including a defective bony canal, and stage IV showing an erosion of adjacent anatomic structures. According to which our case can be classified as a stage IV EACC.

A CT classification system put forward by Shin., *et al*. [9] has been found to improve management which states that stage 1 is cholesteatoma of the EAC only, stage 2 has invasion of the tympanic membrane, the middle ear and EAC involvement , Stage 3: EAC defect and involvement of the mastoid air cells and Stage 4: Lesions beyond the temporal bone. Our case was stage 3.

Tos has classified EACC based on pathogenetic theories into primary EACC, secondary EACC, and cholesteatoma associated with congenital atresia of the ear canal [10]. The aetiology of primary EACC is unknown [10]. Smoking and mechanical factors may be predisposing factors [2]. Secondary EACC is related to a variety of different causes, such as postoperative, post-inflammatory, post-irradiatory, posttraumatic, or post-inflammatory stenosis or atresia of the external auditory canal [2,10-12].

Small lesions can be treated conservatively or by minimal procedures under local anaesthesia. Larger lesions need proper surgery. Principle of treatment for EACC is preservation of the normal EAC skin and surgical removal of the cholesteatoma and necrotic bone with preservation of ossicles or reconstruction of ossicular chain if those are removed [7]. Exact detail surgical procedure depends on extent of cholesteatoma and surgeons judgement and choice [13-15].

Conclusion

External canal cholesteatoma is a rare condition and it poses challenge in diagnosis as it can be missed easily on clinical examination. It should be differentiated from keratosis obturans by radiological investigations.

Conflict of Interest

The authors declare that there are no conflicts of interest regarding the publication of this article.

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