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CASE REPORT

ACQUIRED ATRESIA OF EXTERNAL AUDITORY CANAL WITH CANAL CHOLESTEATOMA: A CASE REPORT

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A B S T R A C T

Acquired secondary atresia of external auditory canal (EAC) is a rare entity in routine otolaryngology practice. The atretic canal can desquamate and trap keratin debris leading to formation of dreaded condition called canal cholesteatoma. This is also accompanied with moderate to severe conductive hearing loss. A cholesteatoma is a cystic keratin filled mass lined with stratified squamous epithelium. It is an inflammatory lesion of the middle ear and mastoid that uncommonly involves the external auditory canal (EAC). The canal cholesteatoma has bone eroding properties and might progress unhindered into middle ear and mastoid and thereby producing various complications. EAC cholesteatoma can have numerous differential diagnoses so it poses a diagnostic challenge. In this case report we have illustrated the clinical details of a 20 years old male whole presented with progressive deformity of left pinna, progressive occlusion of left ear canal and progressive decreased hearing in left ear since 4 months. Pure tone audiometry revealed conductive hearing loss of 45 dB in left ear. High resolution computed tomography (HRCT) demonstrated complete occlusion and a soft tissue density lesion filling the left external auditory canal. There was limited erosion of the canal wall with ipsilateral mastoid air cells showing good pneumatisation. The middle ear cavity was uninvolved. Thus a diagnosis of acquired atresia with cholesteatoma of external auditory canal was established and was managed accordingly.

INTRODUCTION

External auditory canal cholesteatoma is a rare condition with an estimated incidence of 1.2 per 1,000 new otological patients (Anthony PF *et al*., 1982). A cholesteatoma, although a misnomer, is a term used to describe a destructive lesion lined by keratinizing stratified squamous epithelium with associated bone erosion and periosteitis (Heilbrun ME *et al*., 2003; Baráth K *et al*., 2011). Although cholesteatoma occurs almost always in the middle ear cavity, there are special types such as mural and external auditory canal cholesteatoma (EACC) (Baráth K *et al*., 2011).

External auditory canal cholesteatoma (EACC) presents itself by an accumulation of epithelial debris in the ear canal, and early reports on such manifestations have been made in 1850 by Toynbee (Toynbee J, 1850) and later in 1893 by Scholefield. While these cases have appeared as EACC, they may also have represented cases of keratosis obturans, which has similar characteristics. In fact the two terms have previously been used interchangeably but since treatment strategies are different, the distinction between the two conditions is important (Piepergerdes MC *et al*., 1980; Naiberg J *et al*., 1984; Persaud RA *et al*., 2004). Present definitions have mainly been based on a review by Piepergerdes *et al*. in 1980 (Piepergerdes MC, 1980), and a histopathological study by Naiberg *et al*., in 1984.

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Secondary EACC is related to a variety of conditions but predominantly they are associated in ear with prior surgical intervention (Spector GJ *et al*., 1979). Other factors like recurrent inflammation as well as post-inflammatory and post-traumatic stenosis or atresia with ear canal obstruction can also lead to EACC (Tos M, 1997). In addition, radiation therapy incorporating the ear canal can also lead to EACC (Adler M *et al*., 1985; Farrior J, 1990; Martin DW *et al*., 1999). Whereas these conditions to some extent can be explained by their causes of origin, the aetiology in primary EACC is unknown (Tos M, 1997).

Case Report

Informed consent in written was taken from the patient regarding documentation and publication of the clinical details, including pre-operative, imaging, surgical photographs and follow up photographs. A 20 year old male belonging to an urban area presented with complaints of progressive deformity of left pinna, progressive occlusion of left ear canal and

progressive decreased hearing in left ear since 4 months. According to the patient he suffered burn injuries to left side of face and left ear while working in factory about 4 months back. Following this injury he developed contracture of the pinna along with progressive meatal stenosis resulting in complete occlusion of the external auditory canal (EAC). He also complained of progressive decrease in hearing in the same ear for the same period. There was no history of ear discharge, ear ache or hearing difficulty prior to this injury. On examination the left pinna was found grossly deformed with sagging and drooping of the helix, contracted hypoplastic tragus and complete obliteration of the external auditory canal covered with skin which felt firm on probing (Fig 1).

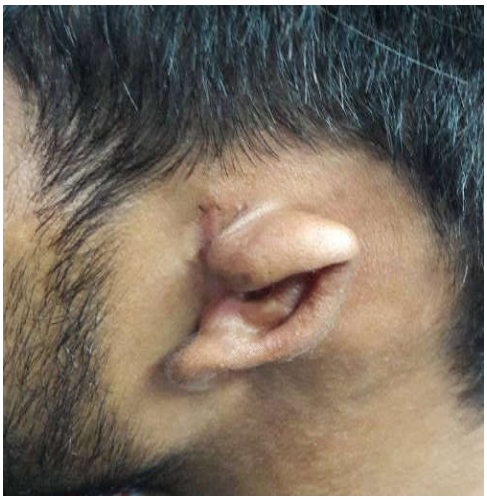

Fig 1 Pre- operative photograph showing meatal atresia and sagging of pinna with contracture marks

There were contracture marks at the crus of helix. Otoscopy could not be performed. Rinnie's test was negative for left ear and Weber's test was lateralized to the left ear suggestive of conductive hearing loss on the left side. Pure tone audiometry revealed conductive hearing loss of 45 dB in left ear (Fig. 2).

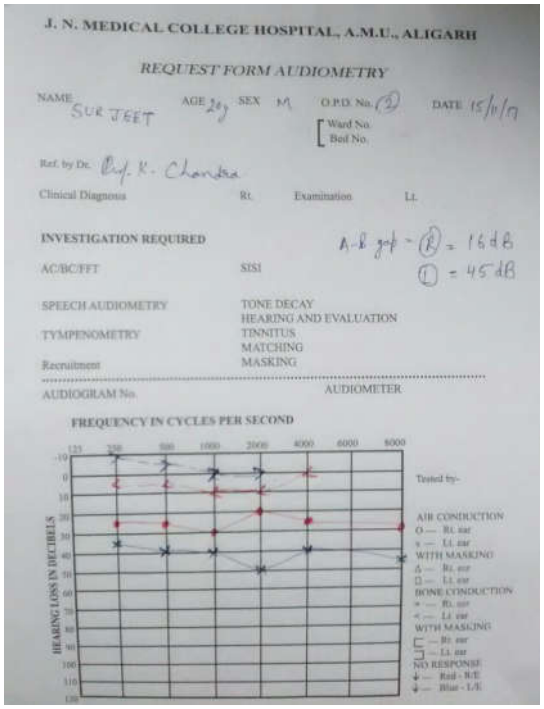

Fig. 2 Pre-operative pure tone audiogram showing 45 dB hearing loss in left ear

The findings of right ear was however normal. High-resolution computed tomography (HRCT) scan of bilateral temporal bone demonstrated complete occlusion and a soft tissue density lesion filling the left external auditory canal with intact tympanic membrane. There was mild widening or erosion of the canal wall with ipsilateral mastoid air cells showing good pneumatisation. The middle ear cavity was uninvolved (Fig. 3).

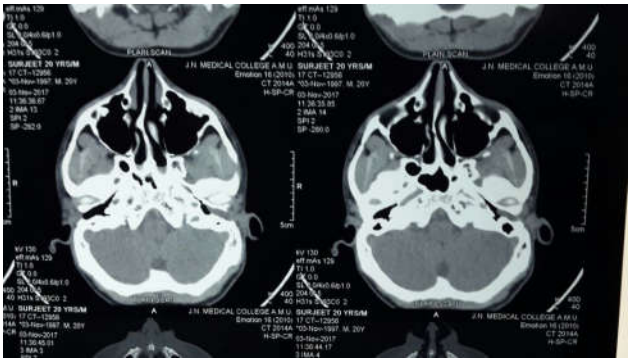

Fig 3 HRCT bilateral Temporal Bone s/o obliteration of external auditory meatus with hypodense soft tissue filling left EAC, limited medially by tympanic membrane. Mild widening with limited erosion of EAC seen.

The clinical and imaging details of the patient suggested burn contracture of pinna with acquired EAC atresia and resultant cholesteatoma limited to EAC not involving the middle ear cleft. The patient was prepared for surgery under general anaesthesia, and his routine pre-anaesthetic work-up was normal. For surgery post auricular approach was adapted (Fig. 4 and Fig. 5).

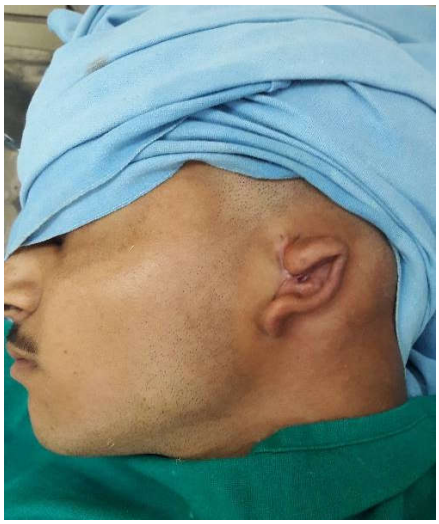

Fig 4 Pre-Operative photograph

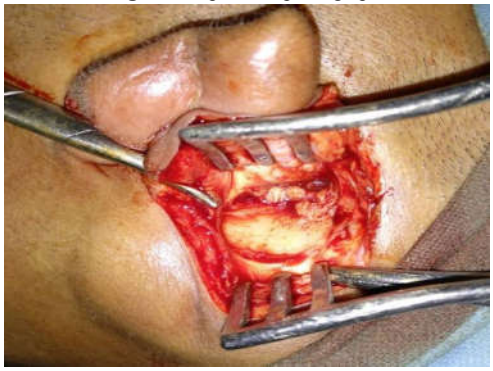

Fig. 5 Intra-Operative photograph showing post auricular approach with cholesteatoma in external auditory canal

Cholesteatoma debris along with sac was removed from the EAC (Fig. 5). Tympanic membrane was found to be intact. The bony canal was widened and polished with a burr as subsequent osteoblastic activity may lead to re-stenosis. The reconstruction of the atretic portion of EAC was done by removing the fibrous plate and covering the bare area of the canal with redundant skin obtained from the fibrous partition. This was further augmented with wide meatoplasty.

Following which reconstruction of the deformed pinna was carried out. Post auricular incision was extended till the tragus along the crus of helix removing the contractures. Undermining of the skin was performed all along the conchal cartilage posteriorly to helix superiorly then to the tragus anteriorly. Excess skin from the posterior conchal and superior helix region was used as skin flap. It was rotated and pedicled at the cus of helix and was attached on to the defect at the tragas and antitragas region created as a result of removal of contractures and wide meatoplasty. Post auricular incision was closed using horizontal mattress sutures. Meatoplasty was packed with antibiotic soaked merocel pieces. Skin graft and pinna was covered with bactigrass dressing and tight mastoid bandage was applied. Stiches were removed on 10th post-operative day (Fig. 6). During the follow up, for subsequent 1 month (Fig. 7), regular dressing of the wound was done using hydrogen peroxide, oxum spray, povidone iodine and the ear canal antibiotic merocel packs were periodically replaced to prevent re-stenosis. Presently the patients has benefitted from cosmetically acceptable pinna architecture with wide EAC meatus and good hearing.

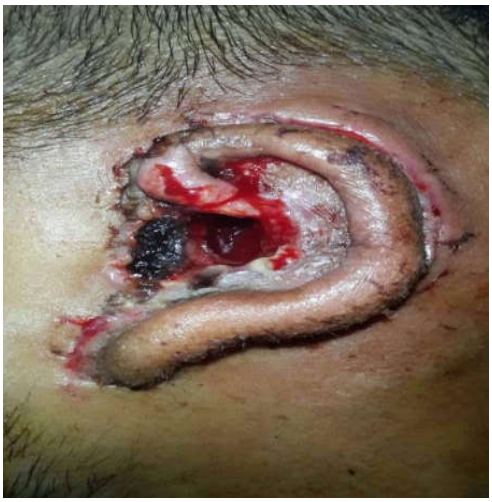

Fig. 6 Follow-up at 10 days

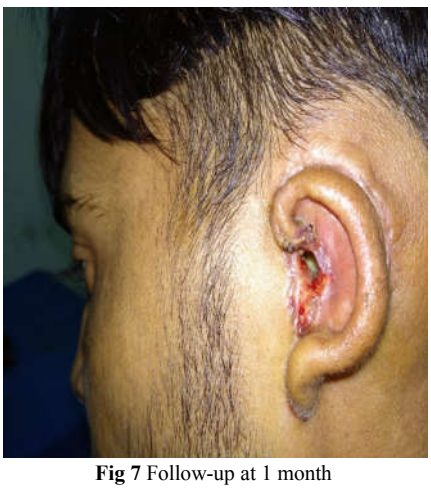

Fig 7 Follow-up at 1 month

DISCUSSION

Acquired atresia of EAC is an rare disease and has an incidence of 0.6 cases/1,00,000 population (Becker BC *et al*., 1998; Namyslowski G *et al*., 2002). Secondary acquired atresia consists of obliteration of a soft tissue plug in the proximal portion of the external auditory canal, medial to the canal skin and lateral to the lateral surface of tympanic membrane. Causative factors are trauma (thermal or chemical burns, radiation, iatrogenic or accidental injuries), chronic otitis externa or neoplasia (Lupin AJ, 1976). Canal stenosis is described as narrowing of the whole external ear canal. Previous otologic surgery accounts for majority of the cases of acquired atresia (Spector GJ *et al*., 1979). Acquired atresia of the external auditory canal may be complete or partial, circular or semilunar. The atretic plate may be made up of fibrous, osseous or cartilaginous tissues or combinations of these. The atretic plate's location is also important. In the great majority of cases, it is located in the cartilaginous portion or at the bony–cartilaginous junction of the ear canal. So the bony canal is involved in only few cases.

Presenting complaints are mostly partial or total occlusion of the EAC which are often covered by normal appearing skin. Prevention of development of acquired atresia is the best treatment. Early treatment of external canal injuries is crucial if stenosis or atresia is to be prevented (McKennan KX *et al*., 1992). Trauma to the ear canal leads to circumferential loss of epithelium. The remanant epithelium between the tympanic membrane and site of circumferential loss continues to desquamate and accumulate keratin debris, which with time build up under pressure and leads to formation secondary canal cholesteatoma (McKennan KX *et al*., 1992).

This can also result as a part of vicious cycle where pre-existing chronic otitis media and/or chronic otitis externa leads to formation of acquired atresia which in turn leads to advancing canal cholesteatoma which in turn propagates chronic otitis condition continuing the cycle. The canal cholesteatoma has the notorious feature of bone erosion and can potentially invade the middle ear cleft and result in serious life-threatening complications.

As acquired atresia of EAC is an uncommon disease, the true incidence of secondary canal cholesteatoma is limited to only a handful of reports. In the study conducted by Magliulo G (Magliulo G, 2009) only 3 out of 41 patients (7.3%) with acquired atresia of EAC developed canal cholesteatoma. Becker BC and Tos M (Becker BC *et al*., 1988) in their retrospective review over 27 years found the incidence to be 9%.

These are in contrast to incidences of cholesteatoma secondary to congenital ear anomalies which are very well documented. In a study of congenital aural atresia involving 94 ears with canal stenosis Casal G *et al*. found 18 ears (19.14%) with acquired EACC (Casale G *et al*., 2014). Mazita A *et al*, found 17 patients with secondary ear canal cholesteatoma in his study group of 41 patients with canal anomalies (Mazita A *et al*., 2011).

HRCT of the temporal bone is the gold standard imaging modality that provides explicit anatomic details of the bony architecture, extent of the cholesteatoma and the possible complications resulting from its erosive property. But as

HRCT has poor soft tissue delineation, it is often difficult to distinguish the extents of true fibrous atresia and the resultant canal cholesteatoma. So, Magnetic Resonance Imaging (MRI) of the temporal bone might be helpful as an adjunct imaging modality to distinguish between these two pathologies.

Presence of soft tissue plug within the EAC without bone erosion is seen in keratosis obturans and post-inflammatory medial canal fibrosis whereas osteolysis is present in EACC, malignant otitis externa, and squamous cell carcinoma (SCC).

Keratosis obturans, the closest differential of EACC (Heilbrun ME *et al*., 2003; Bhagat S *et al*., 2013) presents with smooth widening of EAC. The clinical picture includes acute severe otalgia with conductive hearing loss whereas otorrhea is rare (Heilbrun ME *et al*., 2003). There is a predilection for young age and bilateral involvement. But EACC is typically unilateral; however, occasionally it can involve bilateral EACs (Chawla A *et al*., 2015). Differentiation between the two entities is clinically important because management of keratosis obturans is medical (Heilbrun ME *et al*., 2003).

Post-inflammatory medial canal fibrosis also needs to be differentiated from EACC (Heilbrun ME *et al*., 2003; Bhagat S *et al*., 2013). Inflammatory events (chronic otitis externa and or media) can lead to fibrosis of the medial canal of EAC. HRCT shows nonspecific soft tissue in the medial portion of EAC (Chawla A *et al*., 2015). However, EACC have also been found to coexist in association with post-inflammatory medial canal fibrosis (Heilbrun ME *et al*., 2003).

Malignant otits externa, also known as necrotizing external otitis, is another differential of EACC (Heilbrun ME *et al*., 2003; Bhagat S *et al*., 2013). Diagnosis of malignant otitis externa is based on the clinical presentation of an elderly diabetic with rapidly progressive fulminant otits externa diffusely involving the adjacent soft tissues (Heilbrun ME *et al*., 2003) and the skull base (Heilbrun ME *et al*., 2003; Bhagat S *et al*., 2013) presence of severe otalgia and otorrhea, granulation tissue in EAC, pseudomonas aeruginosa isolation as the offending microbe and HRCT imaging suggestive of an enhancing soft tissue EAC lesion with infratemporal extension (Chawla A *et al*., 2015). On the contrary EACC is a slowly progressive, chronic lesion like middle ear/ mastoid cholesteatoma. Bone erosion, however, is a feature of both. While malignant otitis externa is characterized by diffuse osteolysis, bone erosion in EACC typically affects a single wall (Heilbrun ME *et al*., 2003).

SCC of EAC is another rare differential of EACC (Heilbrun ME *et al*., 2003; Bhagat S *et al*., 2013). Frequently, the canal may be secondarily involved from SCC arising in the adjacent regions. It is seen in elderly and presents with irregular erosion. It may be indistinguishable from EACC on imaging alone (Heilbrun ME *et al*., 2003). Further, early stages of neoplastic process may be difficult to distinguish from benign pathologies based on imaging (Heilbrun ME *et al*., 2003) and hence, biopsy must also be done (Chawla A *et al*., 2015).

Treatment of the atretic ear canal with canal cholesteatoma is mainly surgical. It is a challenging procedure. This can be completed in a single sitting as done in this case or in multiple sittings depending upon the extent of disease and amount of reconstruction required. The aim is to eradicate the disease from the EAC and middle ear and to reconstruct the EAC along with pinna deformity and middle ear hearing mechanism if required.

The atretic plate from the EAC is removed. Cholesteatoma debris along with sac is removed from the EAC. The bony canal must be widened and polished with a burr as subsequent osteoblastic activity may lead to restenosis, especially in children. The bare bone is covered by split or full thickness skin graft (El-Sayed, 1988; Cremers WR *et al*., 1993; Bajin MD *et al*., 2015) or pedicled skin flap (Bajin MD *et al*., 2015; Dhooge IJ *et al*., 1999). This prevents formation of granulation tissue and reduces chances of any re-stenosis. In our case the bare bone of the canal covered with redundant skin obtained from the fibrous partition. Furthermore other modalities like Mitomycin C (Battelino S *et al*., 2005) and stetching steel tubes (Tirelli G *et al*., 2015) have been suggested as treatment for atretic EAC. Re-stenosis is the major complication of surgically fashioned EAC. This can be avoided or minimized by periodic re-packing with antibiotic-soaked merocel strips and instilling antibiotic-steroid ear-drops. Revision surgery might be required in cases of re-stenosis. Patient compliance and meticulous follow up care is of utmost importance in order to obtain optimum results.

CONCLUSION

Otorhinolaryngologists must have a high index of suspicion and suspect EACC in cases of long standing atresia of EAC. HRCT scans of bilateral temporal bone is the gold standard investigation and must be performed in all patients to evaluate the real severity of the disease and involvement of adjacent structures (middle ear, mastoid, jugular bulb and cranial nerves). The treatment of choice in majority of cases is surgery, where main objective is to eradicate the disease and if possible, preserve the patient's hearing acuity. Proper knowledge of this rare disease entity is therefore essential to prevent its progression into avoidable complications.

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