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IP Journal of Otorhinolaryngology and Allied Science

Journal homepage: https://www.joas.co.in/

Case Report

Keratosis obturans masquerading as cholesteatoma- A case report

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ARTICLE INFO

Article history:
Received 07-01-2022
Accepted 28-01-2022
Available online 16-04-2022

Keywords:
Ear Canal
Keratosis Obturans
Cholesteatoma
Facial Palsy
Ballooned-Out Ear Cavity

ABSTRACT

Keratosis obturans is a condition of the external auditory canal, characterised by the stratified accumulation of keratin plugs within the external auditory canal. The progressive accumulation of keratin debris can lead to the gentle erosion and widening of the canal eventually. Whereas, Cholesteatoma also known as active squamosal chronic suppurative otitis media (CSOM) is a benign keratinizing epithelialized cystic structure found in the middle ear and mastoid. It can cause destruction of the local structures – ossicular chain and otic capsule, thereby leading to complications such as hearing loss, vestibular dysfunction, facial paralysis and intracranial disease or infection. Both these entities may present with severe otalgia and conductive hearing loss with radiological evidence of bony erosions. The similarity in clinical presentation can cause confusion between these two conditions. We present a case of keratosis obturans that was misdiagnosed as active squamosal CSOM. The case was diagnosed and managed by microscope guided examination and exploration under general anaesthesia.

1. Introduction

Keratosis obturans (KO) is a rare condition of the external auditory meatus defined by the accumulation of keratinaceous material in a lamellar arrangement that leads to dilatation and blockage of the ear canal.[1] This condition was first described by Tonynbee in 1850, the term KO was first used by Wreden in 1874 who distinguished the condition from impacted wax.[2,3] The annual incidence of cholesteatoma was found to be approximately 3 per 100000 in children and 9.2 per 100000 in an adult Caucasian population in Northern Europe.[4] Keratosis obturans appears to be an obscure and relatively uncommon entity, even in literature search of journals and reference texts, so much so that there is not even any prevalence or incidence statistics available. However H.J Lesser et al. had reported the estimated incidence of KO as four to five patients per 1000 new otological cases.[5]

The clinical features includes severe otalgia and hearing loss due to the pressure exerted by the desquamated keratin debris in the ear canal.[6] Whereas in cholesteatoma, there is almost invariably associated mucosal disease that can be seen and produces the secretions, perhaps along with squamous epithelial debris. These secretions, are mostly mucopus, which can dry and be mistaken for wax.[7] It can cause destruction of the local structures — ossicular chain and otic capsule, thereby leading to complications such as hearing loss, vestibular dysfunction, facial paralysis and intracranial disease or infection.[5]

As the clinical presentations of both these entities are comparable and since the incidence of cholesteatoma is higher than keratosis obturans, KO can often be confused for a case of cholesteatoma. Herein we present a case of

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https://doi.org/10.18231/j.ijoas.2022.005

a patient who was initially diagnosed as having squamosal CSOM with polyps in the external auditory canal and with CT findings suggesting the same. She was diagnosed as having KO intra-operatively along with serous otitis media which was later confirmed on histopathologic examination.

2. Case Report

A 20 year old female presented to us in the ear, nose, throat (ENT) OPD with 6-8 months of right sided severe otalgia and fullness of right ear accompanied by ipsilateral hearing loss. She had undergone serial ear toileting and suctioning for the wax debris in multiple clinics with no relief of her symptoms. 1-2 months later she developed right sided mucopurulent ear discharge which was occasionally blood stained. It was temporarily relieved with oral antibiotic and local antibiotic drops. She presented to us with right sided facial weakness since 1 week and was referred to our OPD.

On examination, patient was afebrile, vitals stable. Otoscopic examination revealed, polypoidal mass filing the external auditory canal reaching upto the conchal bowl with external surface appearing brownish probably due to oxidisation, thickened with crusting. The tympanic membrane could not be visualised due to the polyp filling up the entire external canal. On facial nerve examination revealed there was loss of right nasolabial fold and deviation of angle of mouth to the left on attempt to smile thus having House-Brackmann Grade II right lower motor neuron palsy. The left ear demonstrated mild ear wax, with unremarkable tympanic membrane findings. There were tender right level II lymph nodes. Rest of the examination appears to be normal.

The pure tone audiogram (Figure 1) showed minimal conductive loss on the left side, while right ear had moderate conductive hearing loss with air bone gap of 20-30 dB. HRCT scan of the temporal bone (Figure 2) showed right sided otomastoiditis with soft tissue shadow seen in the middle ear space encasing the ossicles, with ossicles grossly intact; extending into the external auditory canal. Erosion of the tegmen and sigmoid plates where noticed. The right inner ear was intact with normal left ear structures. There was no evidence of facial canal dehiscence or erosion; however signs of inflammation over the tympanic segment of facial canal were noted.

Based on the clinical examination and CT Scan findings the patient was case was primarily thought as a case of cholesteatoma with grade II facial palsy. The patient was admitted and was started on intravenous Amoxicillin and clavulanic acid (45/25mg/kg/day) in two divided doses along with steroids- methylprednisolone(1gm/kg /day) for a week with the aim of reducing the size of the polyp and the surrounding inflammation. The patient was posted for right ear exploration under microscopic guidance.

During ear exploration surgery on local ear toileting, the right external auditory canal was seen to be filled up with what we thought as wax debris. During attempted removal of the wax debris, we observed that the external auditory cabal(EAC) was dilated with bone remodelling around the impacted wax. The ballooned up cavity also lead to the erosion of the dural and sigmoid sinus plate posteriorly. The debris collected was sent for histopathologic examination. The tympanic membrane was grossly intact but dull with an intact annulus. The ballooned up cavity lead to the mechanical obstruction of the aditus leading to the reactionary mucosal oedema and subsequent serous discharge filling up the middle ear cavity.

After clearing the serous discharge, the ossicles were observed to be intact and in continuity.

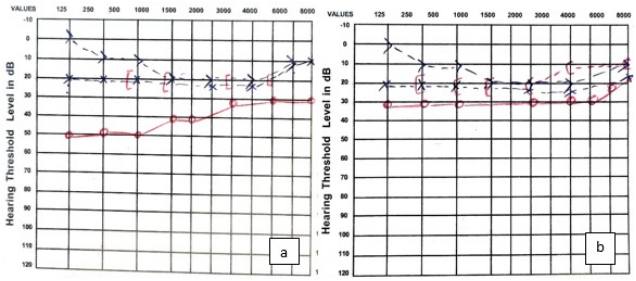

Fig. 1: a) Shows pre-operative PTA showing moderate conductive hearing loss with an air-bone gap of 20-30 db. b) Shows the post-operative PTA showing closing of the air- bone gap.

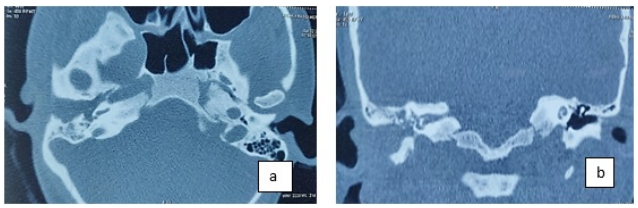

Fig. 2: Figure 2: Shows the HRCT temporal bone images a)axial cuts b) Coronal cuts. Showing soft tissue shadow occupying the middle earspace and encasing the ossicles along with thinning and erosion of the dural andsigmoid sinus.

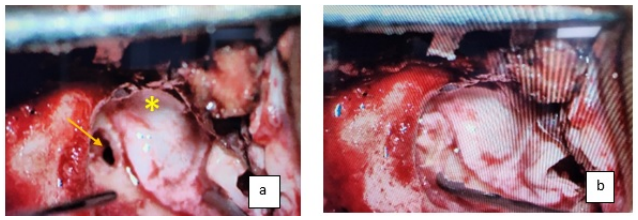

Fig. 3: a) Shows the intraoperative findings, shows a ballooned out external cavity with thinning of the dural (yellow (*) asterix) and sigmoid plates creating an auto-mastoid cavity, antrostomy (yellow arrow) done. b) Showing the defect and the external cavity lined by fascial grafts.

Attico-antral patency was checked and confirmed by creating a small control hole (Figure 3) the tympanic membrane was reposited after giving steroid washes in the middle ear (Figure 3). The eroded sinus and dural plates secured and reinforced by fascial grafts supported by medicated gelfoams. The right ear was duly packed with medicated gelfoams, patient extubated and shifted to the ward with no evidence of postoperative complications. Facial asymmetry improved with only slight weakness noted upon close inspection with no other accompanying complications.

She was given intravenous amoxicillin +clavulanic acid, with tapering dose of prednisolone for 3 days postoperatively. Complete resolution of facial asymmetry was seen from postoperative day 3. The patient was discharged on oral amoxicillin+ clavulanic acid and anti-inflammatory for 1 week. Suture removal was done after 1 week. During follow up 1 week later, ear pack removed postoperative cavity healed and healthy. Post-operative histopathology of the removed plug revealed acellular lamellated keratin flakes and keratinous material associated with chronic inflammation thereby confirming our diagnosis of keratosis obturans. Postoperative audiometry performed 2 months later showed complete closure of the air-bone gap (Figure 1) and hearing was within normal limits.

3. Discussion

Keratosis obturans is the accumulation of a large plug of desquamated keratin in the external auditory meatus, a geometrically patterned keratin plug within the lumen of an expanded external auditory canal.[7] KO is due to abnormal epithelial migration of the ear canal skin.[5] The movement of the surface epithelium of the pars flaccida was reversed such that it migrated downwards to the pars tensa and then moved inferiorly across the whole drum.The keratin squames are shed from the complete circumference of the deep ear canal forming a lamina (onion skin) arrangement.[6] Owing to the abnormal migratory pattern, the keratin plugs gets accumulated into a lamellar arrangement, leading to pressure necrosis and subsequent osteolysis of the underlying bony structures, leading to the varied array of complications on the long run.

It has been suggested that there may be two types of KO.[7] One is of an inflammatory nature which occurs secondary to an acute problem, such as a viral infection, causing inflammation of the ear canal which temporarily alters the epithelial migration. This is cured by removal. The second or the silent type is a disease that persists and is caused by abnormal separation of the keratin that continues even after the first removal and will need subsequent removals.[5] Our patient may be categorised into the inflammatory type of keratosis obturans.

KO occurs more commonly in younger patients. It classically presents with acute severe otalgia, conductive hearing impairment and widened ear canal.[8] Otorrhea is considered a rare presentation.[9] In our patient, her main complaint was severe otalgia and hearing loss which was followed later by otorrhea and facial palsy. Although rare, few cases of keratosis obturans, presenting with facial weakness have been reported by Glynn F, Keogh IJ[10] and Jeyasakthy Saniasiaya et al.[11] KO can cause extensive bony erosion, sometimes leading to auto-mastoidectomy.[5] This explains why our patient had CT findings of erosion. There are no reports of intracranial complications from this disease. A previous study by C. Saunders, R. Malhotra et all demonstrated that complications of untreated KO include sensorineural hearing loss, dehiscence of the tegmen tympani, and facial palsy.[12]

Computer tomography typically demonstrates soft tissue plug in external ear canal with evidence of ballooning of the osseous part.[5] Facial nerve palsy following keratosis obturans are caused by bony erosion,[5,7] which may be due to the pressure effect exerted by keratin mass in the external canal.[8] In our patient, evidence of bony erosion in the HRCT temporal bone was seen in tegmen tympani, dural and sinus plates with intact facial canal with signs of inflammation over the tympanic segment of the facial canal. The pressure exerted by acute inflammation may have caused the facial nerve palsy which was supported by complete resolution after commencement of antibiotics and surgical removal of the ear mass. A similar case was reported by C. Saunders, R. Malhotra et al.[11] in " Keratosis obturans complicated with facial nerve palsy: a diagnostic dilemma".

On the contrary, cholesteatoma or active squamosal type of CSOM is a mass formed by keratinizing squamous epithelium in the middle ear and/or mastoid, subepithelial connective tissue and by the progressive accumulation of keratin debris with/without surrounding inflammatory reaction.[13] They present with a history of ear discharge often with hearing loss, and on examination a tympanic membrane retraction filled with keratin debris.[5] Discharge from a chronic TM perforation after AOM is more common than cholesteatoma in children, and in both conditions florid granulation tissue and possible aural polyp formation will need treatment before the two can be distinguished.[14]

Other clinical differentials for mass in aural canal with facial nerve palsy includes external auditory canal cholesteatoma, neoplasms of external canal and malignant otitis externa. It is important to distinguish the diagnosis, as management of each of the differential diagnosis is notably different.[11] Hence, thorough and detailed history, physical examination, radiological examination and most importantly histopathological examination is crucial prior to a diagnosis. Histopathological examination of the biopsied or excised mass is the main modality of diagnosis, more so when there is an atypical or rare presentation as in this case.

4. Conclusion

Keratosis obturans, though a rare and benign etiology; if not meticulously diagnosed and treated may result in severe complications. The progressive accumulation of keratin debris if left unattended may result in gradual ballooning of the external ear canal and leads to bony remodelling mimicking bony erosions in cholesteatoma. The clinical and radiological features of keratosis obturans and cholesteatoma may be comparable which can lead to misdiagnosis as reported in this case study. If diagnosed early keratosis obturans can be treated with meticulous ear toileting while cholesteatoma requires surgical management. Thus, high degree of clinical suspicion along with timely intervention may help in prevention of these complications.

5. Compliance with ethical standards

I have abided by the ethical standards of the journal. No human participants or animals were used for the purpose of the research.

6. Declaration of patient consent

The authors assert that we have obtained all appropriate patient consent forms. In the form the patient has given her consent for her image and clinical information to be reported in the journal. The patient understands that their name and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

7. Source of Funding

None.

8. Conflict of Interest

The author declares that there is no conflict of interest.

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Cite this article: Dominic SP, Patial A, Shah R. Keratosis obturans masquerading as cholesteatoma- A case report. *IP J Otorhinolaryngol Allied Sci* 2022;5(1):17-20.