Cholesteatoma of Maxillary Sinus Simulating Neoplasia: A Rare Case Report

Abstract
The term cholesteatoma refers to “chole”: cholesterol, “steat”: fat and “oma”：“tumor”. This tumor has been reported to be the most common in the middle ear. The occurrence of such a tumor in the maxillary sinus is deemed to be very rare and hardly 4 cases were reported in India and 26 cases described worldwide. This case report intends to discuss the uniqueness and indolent nature of this lesion in terms of histopathology and radiography.

Keywords: Epithelium, keratin, maxillary, sinus, squamous

Introduction
Cholesteatoma is a condition where respiratory mucosa is either partially or totally replaced by hyperkeratotic squamous epithelium, which leads to the formation of a lamellar sheet of keratin.[1] It was first described by Cruveilhier (1829) as a pearly tumor by its highly refractive and nodular surface. The term cholesteatoma was given by Muller (1838) to describe the presence of cholesterol crystal in a cavity lined by squamous epithelium and filled with layers of dense, squamous keratin.[1] Haeggstrom (1916) reported the first case of cholesteatoma in the frontal sinus. The first case of maxillary sinus cholesteatoma was reported by Hutcheon (1941).[2]

Importance
Only four cases of cholesteatoma of maxillary sinus have been reported to date, the last reported case was in 2014. In this case report, we intend to discuss the various differential diagnosis associated with cholesteatoma which could be easily misdiagnosed. Due to its expansile and space-occupying nature, a provisional diagnosis of ameloblastoma of the maxillary sinus was given. The histopathology satisfied Vickers and Gorlin’s criteria in the epithelium, and dense keratin, a tentative diagnosis of keratoameloblastoma, was bestowed. The deeper section exhibited a lack of ameloblastic tumor islands and dense keratin in the connective tissue which is a characteristic of keratoameloblastoma. Therefore, a further evaluation was done, which revealed a gradual and marked a transition from pseudostratified ciliated columnar respiratory epithelium to the stratified squamous epithelium and dense flakes of acellular keratin supraepithelially suggestive of cholesteatoma.

Case Report
A 36-year-old male reported with a complaint of pain and swelling at the upper right back tooth region for 45 days. On extraoral examination, there was an infected tract opening arising from the maxillary sinus [Figure 1a] with a discharge on the right side of the face near the nasolabial fold. The patient gave a history of mild pain and pus discharge for a month. The patient also gave a history of extraction of 17 three months back. On intraoral examination, there was swelling at the right upper back tooth region obliterating the vestibule in relation to 14–17 region. On radiographic examination, computed tomography revealed the presence of an osteolytic lesion which was hypodense, with well-defined borders and deviation of the nasal septum [Figure 1b]. A relevant consent was taken from the patient for any further procedure to be performed on him.

On surgical exposure, both palatal and facial cortices were found to be eroded by a solid, creamish-white mass. On gross examination, an incisional biopsy was taken...
from 14 to 17 regions. Grossly, the specimen was creamish white in color [Figure 1c] with membranous plaques and soft in consistency. A provisional diagnosis of central jaw lesion is given. The tissue was processed and stained with hematoxylin and eosin. Histopathologically, it revealed a marked transition from pseudostratified ciliated columnar respiratory epithelium to stratified squamous epithelium with lamellar sheets of keratin supraepithelially [Figure 1d and e]. It also exhibited alternating sheets and flakes of loose and dense eosinophilic, acellular, and anucleated areas resembling orthokeratin supraepithelially [Figure 1f] and varying degrees of squamous cell metaplasia [Figure 1g]. The connective tissue also exhibits multinucleated giant cell, few cholesterol clefts [Figure 1h and i], areas of hemorrhage, inflammatory cells, and blood capillaries interspersed with fibrocollagenous stroma. Therefore, correlating all the clinical and histopathological features, it was suggestive of “cholesteatoma of maxillary sinus.” The patient was referred to the oral and maxillofacial surgery department for further treatment.

**Discussion**

It is a relatively common lesion in the middle ear and mastoid cavity.² Cholesteatoma of the maxillary sinus is a rare condition with 12 cases reported to date, since 1965. It presents as a painless swelling with nasal obstruction and discharge. The mean age was 37.1 years. These are biologically nonneoplastic³ but can erode bone and expand into adjacent areas.

Pathogenesis: Some fundamental theories were proposed:³⁴⁻⁵

- Remark and Bucy (1854) stated that they arise from misplaced epithelial rests during the embryonic stage
- Wendt (1873) proposed that in response to infection, nonkeratinizing squamous epithelium lining the cavity undergoes metaplastic change and produces keratin
- Habermann (1888) theorized that migration of keratinizing squamous epithelium in an area where it is not usually found
- Lange (1925), Reudi (1978) proposed that papillations, pseudopods, or microcysts filled with keratin formed in the basal cell layer of the pars flaccida
epithelium, invade the subepithelium of Prussack’s space

- Ewing (1928) stated that during previous trauma and after nasal or sinus surgery, it arises secondary to the direct entry of epithelium
- Wittmack (1933) stated poor aeration of epitympanic space and the structures around it are drawn medially by retraction forming a retraction pocket. It causes negative pressure that restricts the normal migratory pattern of the tympanic membrane, thereby losing its ability of self-cleaning and enhancing keratin accumulation
  - The most recent theory proposed by Jackler et al. (2015) described the mucosal migration onto the inner surface of the tympanic membrane and the interaction between the mucociliary movement of middle ear mucosa leads to the formation of the epithelial pouch which leads to form cholesteatoma.

Recent investigations on bony erosions highlighted the role of cytokines like TNFα. They act directly on the bone and indirectly by stimulating the release of proteolytic enzymes. Overexpression of epidermal growth factor receptor and transforming growth factor-α has also been detected, indicating that the dysregulation of these genes is associated with the initiation and progression of cholesteatomas.

Compilation of various cases of Cholesteatoma of Maxillary sinus was in detail in Figure 2. Theories of etiology of cholesteatoma till now were explained in Figure 3.

Radiologically, it is circumscribed with smooth marginal sclerosis. On panoramic imaging, it is large, hypodense, non-enhancing, expansive, homogeneous lesion.

Histopathologically, it is partly lined by respiratory epithelium and partly by keratinized stratified squamous

| S.No | AUTHOR | COUNTRY | AGE/GENDER | SITE | RADIOGRAPHIC FEATURES | TREATMENT |
|------|--------|---------|------------|------|------------------------|----------|
| 1.   | Pogrel et al(1985) | United states | 46years/Male | Left Maxillary sinus | Bone defect, diffuse haziness of left orbit | Myringoplasty |
| 2.   | S.K.Das (1977) | Ludhiana, India | 55years/Female | Right Maxillary sinus | Opacity of right antrum, widening of cavity and thinning of wall | Caldwell-Luc, Approach, Scopelit |
| 3.   | Sadeghi et al.(1999) | United states | 55years/Female | Left Maxillary sinus | Expansion of the left antrum, with thinning and erosion of maxillary sinus and bony septum | Excision |
| 4.   | Steeper J.S.et al.(1992) | United states | 32years/Male | Left Maxillary sinus | Enlarged mass, causing destruction of inferior, medial, and lateral walls. Mass extends into nasal vestibule | Caldwell-Luc antrostomy |
| 5.   | Vishwanath et al.(2007) | Bangalore, Karnataka, India | 45years/Female | Left Maxillary sinus | Large, hypodense, expansive lesion, usual fossa obturata for nasal septum. | Wide inferior nasal antrostomy |
| 6.   | Malhar H Chavhan,et al(2011) | Chandigarh, India | 47years/Female | Right Maxillary sinus | Well circumscribed, expansive bony destruction | Endoscopic excision |
| 7.   | Pethurajath JG, et al(2014) | Bangalore, Karnataka, India | 25years/Female | Left Maxillary sinus | Destruction of the medial and lateral wall of the sinus and destruction of medial wall of the left maxillary sinus | Excision |
| 8.   | Srawsky J.et al.(2015) | United states | 72years/Male | Right maxillary sinus | Erosion of the right medial maxillary wall | Endoscopic sinus surgery |
| 9.   | J. M. Lee et al(2015) | Korea | 18years/Female | Left Maxillary sinus | Radiopaque lesion, no expansion, except bone defect | Endoscopic excision |
| 10.  | Mii et al(2016) | Korea | 44years/Female | Left Maxillary sinus | Cystic lesion with thin sclerotic rim, homogenous, expansive lesion | Excision with Caldwell-Luc approach |
| 11.  | Baveja S.W.et al(2017) | Thailand | 46years/Female | Right Maxillary sinus | Novel cavity destroying the nasal septum, bony destruction | Maxillectomy |
| 12.  | Bo-han Lim et al(2018) | Korea | 45years/male | Left maxillary sinus | Non-homogeneous soft tissue density in the left maxillary sinus without bony remodeling | Endoscopic excision |
epithelium with supraepithelial acellular keratin flakes. The fibrous stroma exhibits cholesterol clefts with some surrounding foamy histiocytes and multinucleated giant cells with chronic inflammation were also observed.[1]

Lesions that should be differentiated from this are nonneoplastic lesions such as mucocele, mucus retention cyst, and pseudocyst. Benign neoplastic lesions include papilloma, mucin impaction tumor, meningioma, schwannoma, hemangioma, juvenile nasal angiofibroma, and malignant lesions like squamous cell carcinoma of the maxillary sinus, ameloblastoma.[7]

As per literature, in India, the first case in maxillary sinus was reported by S.K.Das et.al. (1971) followed by Sadoff RS et.al.(1989) in United states.

The appropriate treatment is Caldwell-Luc surgery. The wall of the cholesteatoma should be removed entirely to stop further erosion of the surrounding structures.[3,10] The presence of residual epithelium is the usual cause of recurrence.

**Conclusion**

Based on our case report, cholesteatoma should be also considered for any slow-growing, expansile lesion in the maxillary sinus. While it often appears as a high-grade malignancy on radiographic imaging, it can be very easily misdiagnosed as a malignant lesion. Hence, a proper histopathological evaluation and screening of all the sections are required and suggested.

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**Conflicts of interest**

There are no conflicts of interest.

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