

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

**Sushma Vakalapudi,
Sumit Majumdar,
Divya Uppala**

Department of Oral
Pathology and Microbiology,
Gitam Dental College and
Hospital, Visakhapatnam,
Andhra Pradesh, India

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

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Address for correspondence:

Dr. Sushma Vakalapudi,
Department of Oral Pathology
and Microbiology, Gitam
Dental College and Hospital,
Visakhapatnam - 530 045,
Andhra Pradesh, India.
E-mail: sushmav05@gmail.com

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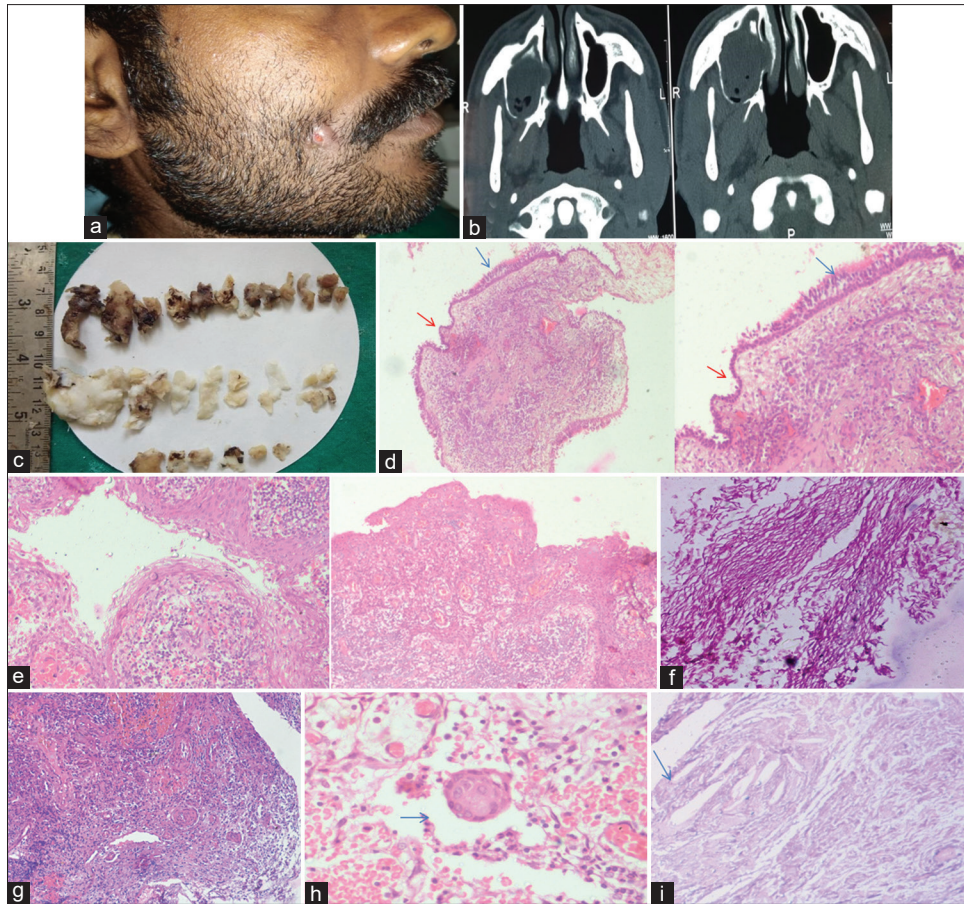


Figure 1: (a) Infected maxillary sinus tract opening and discharge on right side of the face. (b) Coronal view: computed tomography revealed a hypodense osteolytic space-occupying lesion with a deviation of the nasal septum. (c) Grossly, it exhibits brownish and creamish white in color. (d) In $\times 10$ view (left), $\times 20$ view (right), epithelium exhibits a marked transition from pseudostratified ciliated columnar respiratory epithelium (blue arrow) to stratified squamous epithelium (red arrow) (e): $\times 20$ view (left) revealed a stratified squamous epithelium with lamellar sheets of keratin supraepithelially. $\times 10$ view (right) exhibits stratified squamous epithelium with inflammatory cells and blood capillaries. (f) $\times 10$ view exhibits loose sheets and flakes of dense eosinophilic, acellular, and anucleated areas resembling orthokeratin supraepithelially (g) $\times 10$ view shows squamous metaplasia and areas of hemorrhage in the fibrocollagenous connective tissue stroma (h) $\times 20$ view exhibits multinucleated giant cell in the connective tissue (i) $\times 10$ view revealed cholesterol clefts in the connective tissue stroma

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 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 nonneoplastic^[3] but can erode bone and expand into adjacent areas.

Pathogenesis: Some fundamental theories were proposed:^[3]

- 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)^[4,5] proposed that papillations, pseudopods, or microcysts filled with keratin formed in the basal cell layer of the pars flaccida

Figure 2: Compilation of various cases of Cholesteatoma of Maxillary sinus to date from English literature

S.No	AUTHOR	COUNTRY	AGE/GENDER	SITE	RADIOGRAPHIC FEATURES	TREATMENT
1.	Pogorel s et.al(1965)	United states	36years/Male	Left Maxillary sinus	Bony defect, diffuse haziness of left orbit	Maxillary antrostomy
2.	S.K.Das (1971) [8]	Ludhiana, india	55years/Female	Right Maxillary sinus	Opacity of right antrum, widening of cavity and thinning of wall	Caldwell-Luc Approach, scooped out
3.	Sadoff RS.et.al(1989) [9]	United states	55years/Female	Left Maxillary sinus	Expansion of the left antrum, with thinning and resorption of maxillary alveolar and buttress bone.	Excision
4.	Storper I.S.et.al(1992)	United states	12years/Male	Left Maxillary sinus	Erosive mass, causing destruction of anterior, medial, and lateral walls. Mass extends into nasal vestibule.	Caldwell-Luc antrostomy
5.	Viswanatha et.al(2007) [5]	Bangalore, Karnataka, india	18years/Female	Left Maxillary sinus	Large, hypodense, expansile lesion. nasal fossa abutting the nasal septum.	Wide inferior meatal antrostomy
6.	Mahendra Chouhan, et al(2011) [1]	Chandigarh, India	47years/Female	Right Maxillary sinus	Well circumscribed, expansile bony destruction	Endoscopic excision
7.	Puttamadaiah GM, et al.(2014) [10]	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 orbit .deviation of septum	Excision
8.	Sozansky J et.al(2015) [7]	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 bony defect	Endoscopic excision
10.	Min et al(2016)	Korea.	34years/Female	Left Maxillary sinus	Cystic lesion with thin sclerotic rim, homogenous, expansile lesion	Excision with Caldwell-Luc approach
11.	Bourchom W,et.al(2017) [3]	Thailand	74years/Female	Right Maxillary sinus	Nasal cavity abutting the nasal septum, bony destruction	Maxillectomy
12.	Bo-Mun kim et.al(2019)	Korea	41years/male	Left maxillary sinus	Non-homogeneous soft tissue density in the left maxillary sinus without bony remodeling.	Endoscopic excision

Figure 3: Theories of etiology of cholesteatoma till now

S.No	Theory	Authors
1.	Congenital epithelial rests	Remark and Bucy (1854)
2.	Metaplasia	Von Trolsch (1864) and Wendt (1873)
3.	Invasion (immigration)	Bezold (1890), and Habbermann (1988)
4.	Hyperplasia	Lange (1925) and Reudi (1978)
5.	Implantation	Ewing (1928)
6.	Invagination (retraction pocket) (Most widely accepted until now)	Wittmack (1933)
7.	Wound healing	Albino et al. (1998), Huisman et al. (2008)
8.	Epitympanic dysventilation	Marchioni et al. (2010, 2011, 2013)
9.	Mucosal traction (Most recent theory)	Jackler et al. (2015)

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)^[4,5] 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)^[5] 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 TNF α . 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.^[6]

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, nonenhancing, expansile, homogeneous lesion.^[1]

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

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)^[8] followed by Sadoff RS.*et al.*(1989)^[9] 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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