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CASE REPORT

Cholesteatoma of Maxillary Sinus: Mimicking as Sinus Tumor

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ABSTRACT

Paranasal sinuses are normally lined by respiratory mucosa which is pseudostratified ciliated columnar epithelium. Cholesteatoma of paranasal sinus is a condition where respiratory mucosa is either partially or totally replaced by hyperkeratotic squamous epithelium which lead to formation of lamellar sheet of keratin and this condition is known as cholesteatoma. We report one such rare occurrence of maxillary sinus cholesteatoma managed endoscopically.

Keywords: Cholesteatoma, Paranasal sinuses, Tumor, Excision.

INTRODUCTION

Paranasal sinuses are normally lined by respiratory mucosa which is pseudostratified ciliated columnar epithelium. Cholesteatoma of paranasal sinus is a condition where respiratory mucosa is either partially or totally replaced by hyperkeratotic squamous epithelium which lead to formation of lamellar sheet of keratin and this condition is known as cholesteatoma. It is also known as primary epidermoid tumor, keratoma, epidermoid cyst, dysembryodysplasia and keratocyst.[1-3] Hartman and Stankiewicz (1991) in their review of literature of cases of paranasal sinus cholesteatoma found only 20 cases[4] and since then, only one case has been reported by Borlingegowda Viswanatha et al (2007).[5]

CASE REPORT

A 47-year-old female patient presented with right nasal obstruction with discharge and painless cheek swelling on the right side for 6 months. The swelling was insidious in onset and gradually progressive. There was no history of trauma, headache, sinus surgery, blurring of vision, diplopia or any other visual symptoms.

On external examination, a 2 × 3 cm size, smooth, rounded, hard swelling was present in right cheek area obliterating nasomaxillary groove and was nontender on palpation.

On anterior rhinoscopic examination, a pinkish smooth mucosa covered swelling was present pushing lateral wall of nose medially, obstructing the right nasal cavity and pushing nasal septum to left side. The posterior rhinoscopic examination was grossly normal.

The computed tomography showed well-circumscribed non-enhancing mass lesion of right maxillary sinus with expansile bony destruction of medial and anterolateral walls.

Intraoperative findings: Endoscopic excision with 0° and 45° Hopkins telescopes was done under general anesthesia. There was a smooth cystic lesion which was opened on the anterolateral aspect with sickle knife. It yielded whitish grey pultaceous cheesy material which was scooped out (Fig. 1). The cyst wall could be separated all around and was dissected from underlying tissues with

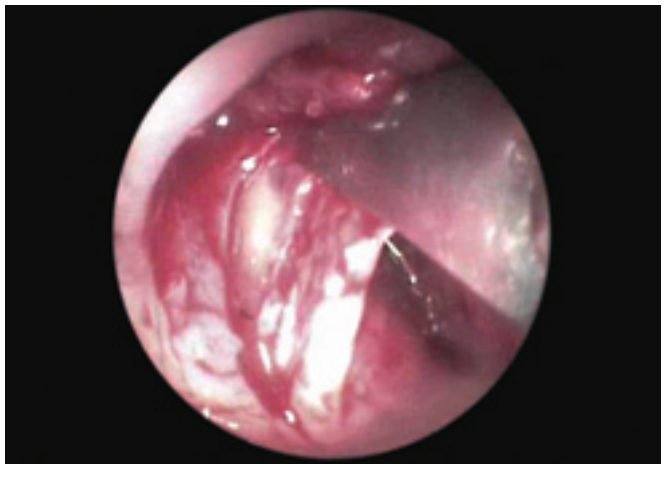

Fig. 1: Intraoperative endoscopic photograph showing pultaceous material filling the cystic lesion of right maxillary sinus

Freere's elevator and both the wall and scooped material sent for histopathological examination, an unerupted tooth was found which was present in cyst wall in posterolateral part was also included. The histomorphology showed irregular multiple tissue pieces partly lined by respiratory epithelium and partly by keratinized stratified squamous epithelium with acellular keratin flakes (Fig. 2). The stroma showed many cholesterol clefts with surrounding foamy histiocytes and multinucleated giant cells (Fig. 3), confirming a diagnosis of cholesteatoma. In addition, bony fragments with chronic inflammation were also seen.

DISCUSSION

Cholesteatoma was first described by Cruveilhier in 1829, as pearly tumor by virtue of its highly refractive and nodular surface.[6] The term cholesteatoma was given by Muller in 1838 to describe the presence of cholesterol crystal in a cavity lined by squamous epithelium and filled with layers of dense, squamous keratin. In 1854,[7] Virchow said that cholesterol is not necessary component of this entity.[8] Epidermoid term was coined by Critchley and Ferguson.[9]

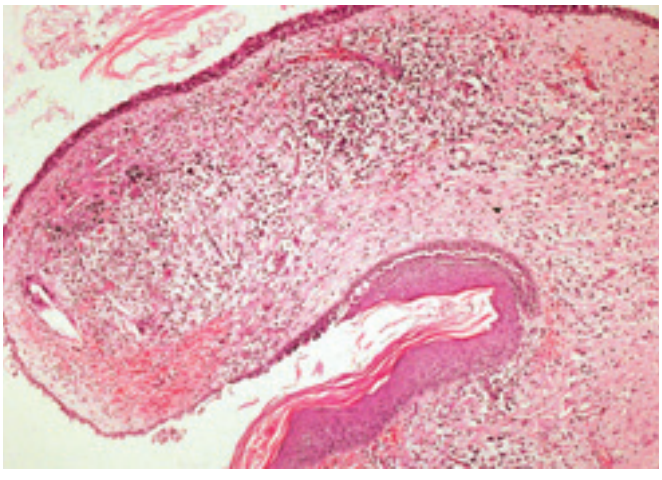

Fig. 2: Microphotograph showing keratinized squamous lining with keratin flakes and partly respiratory lining. The stroma shows inflammation and giant cells (H&E × 240)

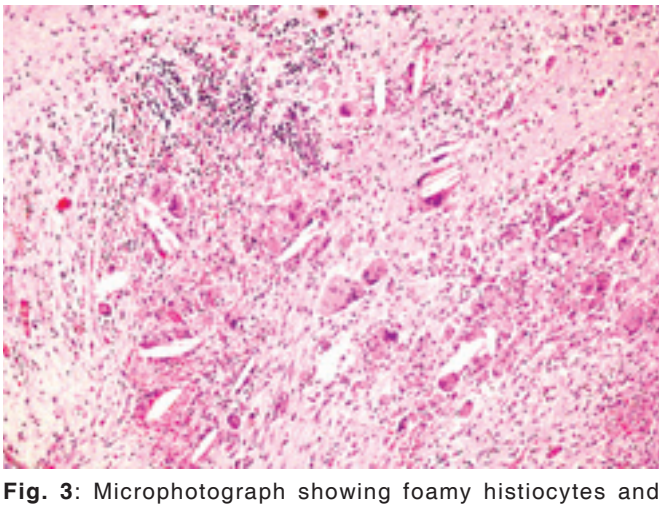

Fig. 3: Microphotograph showing foamy histiocytes and multinucleated giant cell reaction around cholesterol clefts (H&E × 540)

Confusion is there regarding the use of term nasal cholesteatoma and antral cholesteatoma which was made clear by Kotton (1973). He described nasal cholesteatoma as synonym to pseudocholesteatoma or rhinitis caseosa whereas antral cholesteatoma is true cholesteatoma containing squamous epithelium and keratin debris. Duplay used the term rhinitis caseosa (1868) and Eggston and Wolff used the term pseudocholesteatoma. Wolfowitz (1973) described histopathology of nasal cholesteatoma or rhinitis caseosa and showed that it contains no squamous epithelium and filled with necrotic tissue, inflammatory cells, granulation tissue and also foreign body giant cells, cholesterol crystals[10] (Eggston and Wolff, 1947).

Cholesteatoma can occur in skin, kidney, breast, central nervous system and it also involves the cranium, like temporal bone, mandible, orbit and paranasal sinuses. Amongst paranasal sinus, frontal sinus is most commonly involved followed by ethmoid and maxillary sinus.[1] Haeggstrom (1916) reported the first case of cholesteatoma of any paranasal sinus. He reported a case of frontal sinus cholesteatoma presenting with proptosis and diplopia.[11] Hutcheon (1941) reported first case of cholesteatoma of maxillary sinus, in which sign and symptoms resembled that of malignancy of maxillary sinus thus despite the rarity of the lesion antral cholesteatoma should be kept as differential diagnosis.[12]

Histologically, cholesteatoma is consisting of three components: Keratin debris core, squamous epithelium and fibrous stroma, last two components being termed as matrix. This is non-neoplastic expansile lesion with capacity of bone erosion due to proteolytic enzymes. Harris demonstrated two proteolytic enzymes, leucine aminopeptidase and nonspecific esterase in subepithelial layer of cholesteatoma.[13] However, we did not performed any enzymatic study in our patient.

Pathogenesis

Cholesteatoma can be classified into primary or secondary. Primary cholesteatoma occurs without any implantation or migration of squamous epithelium into the site of origin. Four basic theories have been proposed to explain its development:

1. *The theory of congenital epithelial rest (1854):* Remark believe that cholesteatoma arise from misplaced epithelia rests that develop during the embryonic stage.[14]
2. *The metaplasia theory (1873):* According to this theory, proposed by Wendt, the nonkeratinizing squamous epithelium that lines the cholesteatoma is a result of squamous metaplasia.[15]

3. *The immigration theory (1888):* This theory was proposed by Hebermann, where cholesteatoma is caused by the migration of keratinizing squamous epithelium into an area where it is not usually found.[16]
4. *The implantation theory (1928):* Ewing proposed that cholesteatoma arises secondary to direct implantation of epithelium during trauma.[17]

Radiologically cholesteatoma is typically a sharply circumscribed bony defect with smooth marginal sclerosis mimicking mucocele. On CT, cholesteatoma is nonenhancing, expansile, homogeneous lesion. On MRI, it exhibits low intensity on T1-weighted and high intensity on T2-weighted images.[18]

DIFFERENTIAL DIAGNOSIS

Cholesteatoma of maxillary sinus should be differentiated from both non-neoplastic lesions (mucocele, mucus retention cyst, pseudocyst and pyocele) and neoplastic lesions. Neoplastic lesions may be benign or malignant. The benign lesions include papilloma, mucin impaction tumor, meningioma, schwannoma, chondroma, hemangioma, chordoma, fibrous dysplasia, juvenile nasal angiofibroma and squamous cell carcinoma of maxillary sinus among the malignant lesion.

Cholesteatoma of paranasal sinus is unusual diagnosis, as only 21 cases have been reported till date, the last reported case was in 2007 prior to this one case was reported in 1992.

SUMMARY

Cholesteatoma of paranasal sinus is a condition where respiratory mucosa is either partially or totally replaced by hyperkeratotic squamous epithelium which leads to formation of lamellar sheet of keratin and this condition is known as cholesteatoma also known as primary epidermoid tumor, keratoma, epidermoid cyst, dysembryodysplasia and keratocyst. We are presenting a case of 47-year-old female patient presented with right nasal obstruction, nasal discharge and right painless cheek swelling for 6 months, which was insidious in onset and gradually progressive. It is only 22nd time in world literature when cholesteatoma of paranasal sinus is being reported.

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