

CASE REPORT

Clear cell calcifying epithelial odontogenic (Pindborg) tumor involving the maxillary sinus: A case report and review of literature

Priya Sahni, Meghanand T Nayak, Abhishek Singhvi, Jayanti Sharma

Department of Oral and Maxillofacial Pathology, Vyas Dental College and Hospital, Jodhpur, Rajasthan, India

Address for correspondence:

Dr. Priya Sahni,
Department Of Oral and Maxillofacial Pathology,
Vyas Dental College and Hospital, Kudi
Houd, Pali Road, Jodhpur, Rajasthan, India.
E-mail: priyajodhpur@yahoo.com

ABSTRACT

Calcifying epithelial odontogenic tumor (CEOT) is a rare benign odontogenic neoplasm of the jaws, accounting for less than 3% of all odontogenic tumors. It rarely extends into the maxillary sinus. Till date, six cases involving maxillary sinus have been reported. In this paper, we report the seventh case of a 52-year-old male with CEOT in maxilla extending from distal surface of the right maxillary canine to retromolar area and involving maxillary sinus with no association with impacted teeth. The diagnosis was confirmed by aspiration cytology and histologically, the tumor was composed of sheets of epithelial cells, with areas of clear cell changes. The presence of clear cells in the histological sections, accounts for the aggressive nature of the tumor simulating the clinical appearance. Prevention of recurrence can be achieved by radical resection.

Key words: Calcifying epithelial odontogenic tumor, clear cells, fine needle aspiration cytology, impacted tooth, maxillary sinus

INTRODUCTION

Calcifying epithelial odontogenic tumor (CEOT), a rare odontogenic neoplasm arising from odontogenic epithelium was first identified by Thoma and Goldman.^[1] It was not until 1955, however, that CEOT was recognized as a separate entity by Pindborg.^[2] Fifty-two percent of CEOT are associated with a tooth impacted and/or displaced by the tumor.^[3] The tumor can be divided into two clinico-topographic variants: Intraosseous (central) and extraosseous (peripheral) with an incidence of 94% and 6%, respectively.^[4] The intraosseous tumors, when present have a predilection for the mandible more than maxilla with a ratio of 2:1 in the premolar and molar region. Extraosseous tumors are often located in the anterior region of the jaws and involve the gingiva.^[5]

The most common clinical manifestation of CEOT, when detectable, is a localized swelling of the involved jaw. Pain or paresthesia may or may not be a feature depending upon the size of the tumor, the growth pattern, or its anatomic location

and proximity to neurovascular structures. Radiographically, CEOT is characterized as a unilocular or multilocular radiolucent lesion that often exhibits a mixed radiopaque-radiolucent pattern.^[6] The mixed pattern is due to areas of scattered flecks of calcification in the central radiolucency. However, calcifications sometimes, may not be evident on radiographs.^[5]

The histological criteria listed by Franklin and Pindborg^[7] for the diagnosis of CEOT are sheets of polyhedral epithelial cells that have well defined border and often show prominent intercellular bridges. Pleomorphism of the epithelial cells is often noticed along with prominent nucleoli. Mitotic figures are rarely seen. A characteristic feature seen within the sheets of epithelial cells are circular areas filled with a homogenous substance resembling amyloid-like material, which stains positively with Congo red. Some of these cells are also filled by calcified material in the form of concentric Leisgang's rings, which are pathognomic of this tumor.^[7] The tumor cells bear close resemblance to the cells of stratum intermedium of enamel organ and dental lamina remnants, based on its anatomic distribution in the jaw.^[8] CEOT may show variation in this classic histological appearance. Three such variants reported in the literature are the noncalcifying CEOT with Langerhans' cells, the CEOT displaying cementum-like and bone-like material, and the clear cell CEOT.^[9-14] The clear cell CEOT variant is more aggressive with a higher recurrence rate (22%) and is often associated with cortical perforation. Some authors consider this form to be a low-grade odontogenic

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carcinoma.^[8,15,16] When a CEOT affects maxilla, it rarely extends into the maxillary sinus. There are six reported cases in the literature so far.^[17-22] This report describes the seventh case of CEOT in this anatomically uncommon location.

CASE REPORT

A 52-year-old male patient presented with a swelling over the right cheek area since 18 months. On clinical examination, a solitary well defined swelling measuring 3×2×1.5 cm extending from the distal surface of right maxillary canine to retromolar area of the hard palate was seen. Grade II mobility of all the involved molars were observed. The swelling was firm in consistency and the overlying mucosa was normal [Figure 1].

On the radiographic examination, the occlusal radiograph showed complete obliteration of the sinus. Panoramic view showed radiolucent and radiopaque lesion involving the right alveolus and the maxillary sinus with erosion of lateral wall and floor of the sinus. Multiple foci of radiopacity simulating flecks of calcifications were seen giving it a “snow-driven appearance”. Computed tomography (CT) scan showed the right maxillary sinus involvement and demonstrated mixed radiolucent and radiopaque areas. The lesion extended toward the lateral wall of the nose [Figures 2a and b]. The swelling was aspirated and a cytological smear was prepared. It showed clusters of epithelial cells with uniform basophilic centrally placed nuclei intermixed with RBCs and inflammatory cells. Epithelial cells were polyhedral in shape with well-defined borders having pale eosinophilic cytoplasm. Few cells showed nuclear and cellular pleomorphism [Figures 3 and 4]. A provisional diagnosis of CEOT was made and for further confirmation an incisional biopsy was performed.

The histopathological sections revealed varying sizes of epithelial islands surrounded by hemorrhagic areas in a fibrous connective tissue stroma. Some areas of circular amorphous eosinophilic hyalinized amyloid-like material were seen. [Figure 5] Areas of clear cells having vacuolated cytoplasm were identified within the epithelial islands [Figure 6]. Few cells with pleomorphism and mild degree of variability in nuclear size were seen. As clear cell variant of CEOT was diagnosed, which being aggressive in nature, a hemimaxillectomy using Le Fort I surgical approach was performed. The excised tissue was further histo-pathologically diagnosed as CEOT with free margins. The patient was followed up for 3 years postsurgically. There was no evidence of recurrence during this period.

DISCUSSION

CEOT has been reported under a variety of different terms such as “Adamantoblastoma,^[1] “unusual Ameloblastoma,”^[23] and “Cystic complex odontoma.”^[24] CEOT or Pindborg tumor accounts for less than 3% of all the odontogenic tumors.

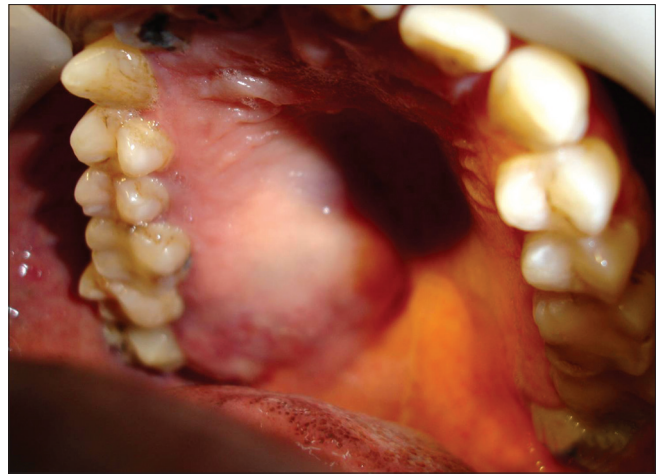


Figure 1: An expansile well-defined swelling involving the right hard palate and extending toward the anterior border of the soft palate

The tumor frequently affects adults with equal distribution between men and women in an age range of 20–60 years, with a peak of incidence between 40 and 60 years. CEOT is generally, considered as a benign tumor of odontogenic origin. In few cases, the tumor has been described as being locally aggressive, invading the surrounding soft tissues and bone marrow.^[18] Intraosseous CEOT affects the mandible twice as often as does the maxilla and it predominates in the premolar/molar region of the mandible, although other sites may also be involved.^[21,25,26]

Extension of the tumor into the maxillary sinus is a rarity with only six cases been reported in the literature [Table 1].^[17-22] The present report speaks about first case in an Indian patient. The age range in all reported cases was between second and third decade, whereas the present case was in the fifth decade. There was no sex predilection^[20] as all the reported cases have equal sex distribution. Some of these cases showed an association with unerupted teeth^[17-21] while other cases,^[20,22] including the present one, showed no association with an impacted teeth. The CEOT typically presents as a painless asymptomatic expansile mass, along with an extra oral swelling of the cheek. Four of the reported cases showed clinical signs of nasal obstruction.^[17-20] and two cases also reported epistaxis and proptosis.^[21,22] The current case presented with an expansile swelling of the palate and extraoral swelling on the cheek. The sizes of all the reported cases were variable.

Radiographically, CEOT exhibits a unilocular or multilocular radiolucency with diffuse radiopacities within.^[21] The CT and magnetic resonance imaging (MRI) features are more specific, with CT showing a well-defined area of soft tissue alteration with scattered high-density foci. The expansile nature of the lesion produces cortical thinning, no bone destruction and no soft tissue invasion. MRI image are reported to be low signal intensity on T1-weighted and high signal intensity on T2-weighted image.^[27] The report of the current case showed complete obliteration of the right maxillary sinus with erosion

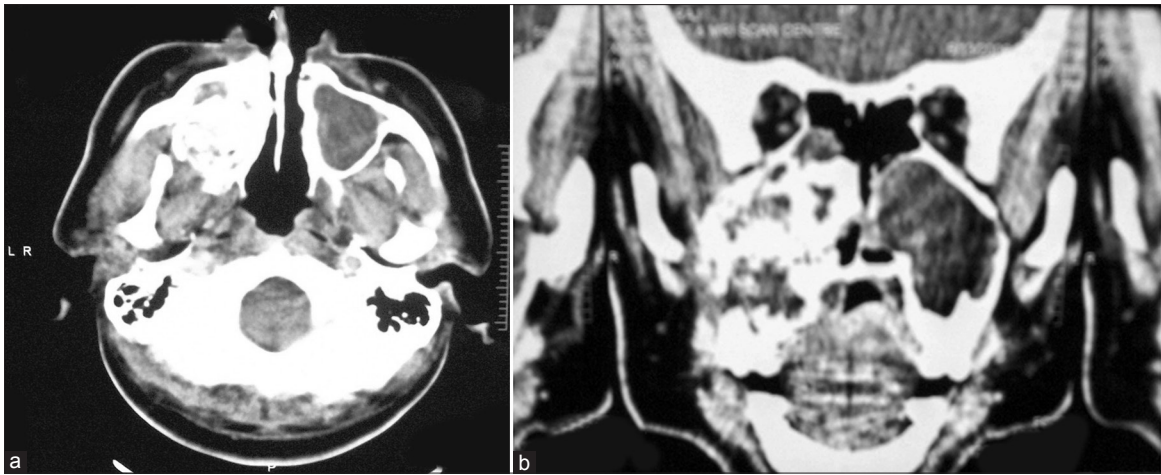


Figure 2: (a) Computed tomography (CT) scan showing a large tumor involving the right maxillary sinus, demonstrating mixed radiolucent and radiopaque areas encroaching the orbital floor (b) Computed tomography (CT) scan demonstrating a massive radiopacities within the maxillary sinus causing erosion of the lateral wall of the nose

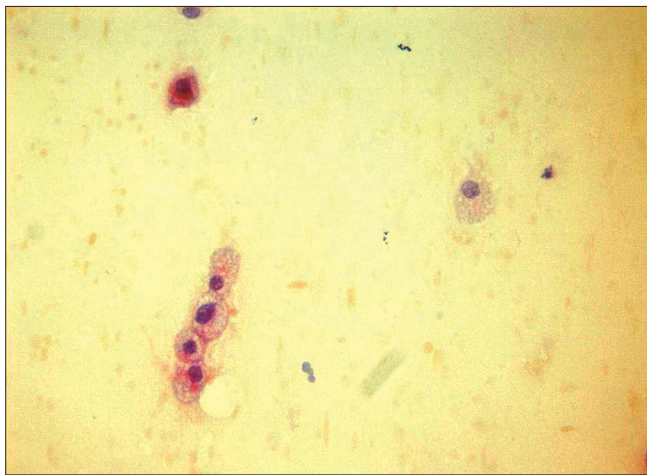


Figure 3: FNAC showing epithelial cells in clusters with uniform basophilic central nuclei. (Magnification 40x; Hematoxylin and Eosin stain)



Figure 4: FNAC showing polyhedral cells with distinct cell borders, nuclei, and pale eosinophilic cytoplasm. (Magnification 40x; Hematoxylin and Eosin stain)

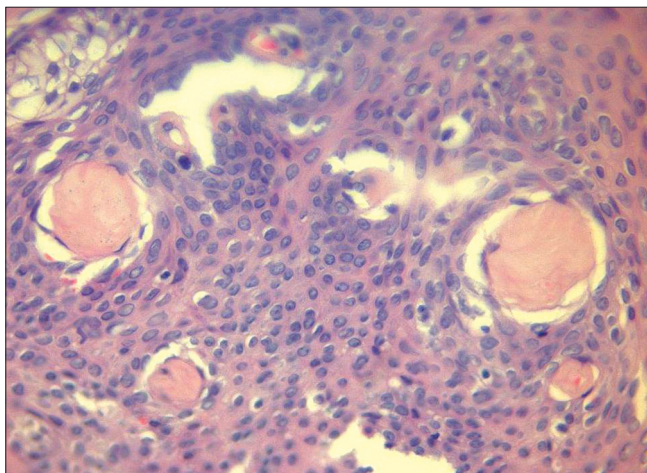


Figure 5: Photomicrograph showing sheets of tumor epithelial cells along with pools of amorphous eosinophilic amyloid. (magnification 40x; Hematoxylin and Eosin stain)

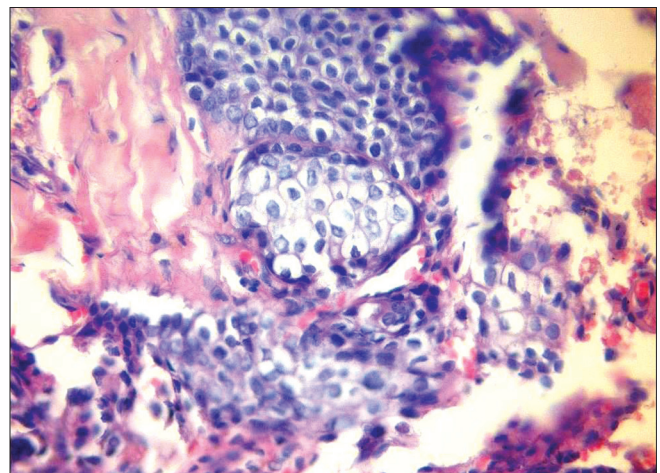


Figure 6: Photomicrograph showing significant areas of clear cells with vacuolated cytoplasm amidst polyhedral epithelial cells. (magnification 40x; Hematoxylin and Eosin stain)

Table 1: Reported cases of CEOT occurring in maxillary sinus

Author	Year	Geographic distribution	Age/ Sex	Clinical features	Site	Size (mm)	Impacted/ Non-impacted
Gon ^[17]	1965	African (South Africa)	35/F	Nasal stuffiness, nose bleeds, headache for many years. Later proptosis was seen.	Swelling over right maxillary antrum	N.A	CEOT associated with impacted right first maxillary molar in the in the floor of the antrum
Stimson <i>et al.</i> ^[18]	1968	North American (United States of America)	35/M	Difficulty in breathing through left side of the nose, enlargement left side of the nose and adjacent maxillary bone	Left nasal fossa and maxillary fossa	45×34	CEOT associated with impacted maxillary left third molar
Lee <i>et al.</i> ^[19]	1992	North American (United States of America)	27/F	Difficulty in breathing through left side of the nose, nasal obstruction, facial asymmetry, progressive enlargement of the left malar region associated with infra-orbital dysesthesia	Extension of the left maxillary sinus	25×35×4	CEOT located in the posterior superior aspect of the maxillary sinus impacted maxillary left second premolar
Mohtasham <i>et al.</i> ^[20]	2008	Middle East Asia(Iran)	18/M	Nasal obstruction, enlargement of the left maxillary area	Extension in the left maxillary sinus	35×20×5	Not associated with an impacted tooth
Bridle <i>et al.</i> ^[21]	2006	European (United Kingdom)	30/F	Superior displacement of the globe of the right eye	Extension in the right maxillary sinus	N.A	The displaced maxillary right second premolar was located with in the obliterated right nasal cavity and maxillary right first molar was located in the region of the right pterygopalatine fossa.
da Rosa <i>et al.</i> ^[22]	2011	South American (Brazil)	33/F	Extensive facial Growth of the left side.	Invasion of soft tissues, masseter, orbicularis oris, medial pterygoid, left maxillary sinus and lateral all of the nasal cavity.	40×30	Not associated with an impacted tooth
The Present case	2012	Asian (India)	52/M	Swelling over the right cheek area	Extension into the right maxillary sinus	30×20×15	Not associated with an impacted tooth

CEOT: Calcifying epithelial odontogenic tumor

of lateral wall and floor of the sinus. Multiple foci of radiopacity were seen in the lesion, giving a “snow-driven appearance”.

Fine needle aspiration cytology (FNAC), as a diagnostic tool, has been sparingly used and reported in the literature of CEOT so far. The report of cytological smears are characterized by clusters, sheets and rare isolated pleomorphic cells of squamoid type; blocks of amorphous material encircled by fibroblast and occasional calcifications.^[28] The cytological smear of the present case revealed clusters of epithelial cells with basophilic

nuclei and well defined borders. Few cells showed nuclear and cellular pleomorphism. The histopathological findings classically seen in a CEOT comprises of polyhedral cells arranged in sheets having prominent intercellular bridges along with calcified and noncalcified amyloid substance.^[7] Philipsen and Reichart^[29] reported variations in the typical histo- and cytopathologic CEOT appearance. Sheets of classic polyhedral epithelium with abundant eosinophilic cytoplasm may alternate with zones of epithelium characterized by large cells with clear, foamy cytoplasm, and distinct cell borders. Yamaguchi *et al.*^[30]

believed that the clear tumor cells represent a feature of cyto-differentiation rather than a simple degenerative phenomenon. The present case revealed clear cell changes in the epithelial tumor cells characterized by vacuolated cytoplasm. Few areas of cellular pleomorphism and mild degree of variability in nuclear size were seen. Other variants of CEOT with findings of cementum-like and bone-like material, Langerhan's cells have also been identified in the stroma.^[9-12]

The treatment of CEOT in the past has been varied, ranging from simple enucleation or curettage to hemimandibulectomy or hemimaxillectomy. The method of treatments will depend on size, anatomic location of the tumor, histopathological findings, patient age, health status, and consideration of reconstruction methods following surgical procedure.^[6,21] The Le Fort I osteotomy and its modifications, is a well reported surgical approach for access to the nasopharynx, base of the skull and upper cervical spine.^[31,32] It is also described as a method for surgical access to the mid-face and maxillary antrum.^[33] The tumor in the current case extended from alveolar process to the right maxillary sinus and was treated by hemimaxillectomy using Le Fort I surgical approach. The prognosis of CEOT, postsurgically is good. Franklin and Pindborg^[7] reported recurrence rate of 14%, which was mostly attributed to inadequate treatment. The current case was followed up for 3 year with no evidence of recurrence.

CONCLUSION

The lesions in the maxilla are usually aggressive and require extensive surgeries as they grow rapidly when compared to the mandibular cases and invasion into surrounding vital structures also affects the morbidity of these patients. This case report describes the seventh known case of CEOT invading the maxillary sinus, which complicates the treatment planning further. Resection with tumor-free accurate margins is the preferred treatment choice. Since CEOT with clear cell changes are known to recur more frequently, a life-long follow-up of these patients is mandatory.

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