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CASE REPORT

Journal Section

Clinical, Radiological features of Calcifying Epithelial Odontogenic Tumor: Report of two rare cases

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Abstract

The calcifying epithelial odontogenic tumor (CEOT) is a rare benign odontogenic tumor constitutes around 1% of all odontogenic tumours involving the jaw. The intraosseous and extraosseous variant constitutes about 95% and 5% of CEOT respectively. We have reported two CEOT cases. One is associated with an impacted left third molar in the maxilla of a 39-year-old female patient, and the other with a sessile soft tissue gingival mass in the left molar region of a 30-year-old female patient. It also emphasizes the importance of advanced imaging and peculiar findings of computed tomography in diagnosing this rare tumour. The visualization of the internal structure of the lesion and the involvement of the neighbouring structures were considered very helpful for diagnosis and treatment planning. Early detection and treatment planning for such rare cases is required to prevent further complications.

KEYWORDS

CEOT; Computed Tomography; Calcifications; Diagnosis; Pindborg tumor

1 | INTRODUCTION

The calcifying epithelial odontogenic tumor (CEOT) also known as Pindborg tumor is a relatively uncommon lesion that is classified as a benign epithelial odontogenic tumor. CEOT was first described by Pindborg in 1955.[1] The tumor is a locally aggressive that constitute for less than 1% of all odontogenic tumors.[1] Majority of the cases of CEOT (52%) are associated with impacted teeth. It is two times more common in the mandible than the maxilla.[2] CEOT have been reported in all age groups, with a mean age of 43.5 years. It is found to affect both genders equally. CEOT basically classified into two type as intraosseous (central [95%]) or extraosseous (peripheral [5%]). The central CEOT on radiographical examination presents as a pericoronal lesion. Multiple small radiopaque flecks around the crown of the impacted tooth within the lesion is a hallmark sign of CEOT.[3] The main histopathological features of CEOT include polyhedral, epithelial tumor cells arranged in sheets, cords or islands with intercellular bridges. They usually have a well

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defined cell border. Treatment modalities of CEOT depends on tumor size, location, extent and it ranges from enucleation or curettage to radical surgical resection followed by reconstruction. According to various literature, recurrence rate of 14% is reported after the conservative management.[2] Two cases of CEOTs manifested as central and peripheral type are reported in the present article.

2 | CASE 1

A 39-year-old female reported with a complaint of pain, swelling in upper left posterior teeth region in the past six months. On extraoral examination, there was facial asymmetry noted on the left side of face because of an ovoid diffuse swelling in the maxillary sinus region. Overlying skin appeared normal and on palpation the swelling was bony hard and tender [Figure 1 (a)]. Intraoral examination revealed missing maxillary left third molar. Grade 1 mobility was present with the maxillary left second premolar, first and second molar. The overlying mucosa was normal in colour and consistency. Buccal cortical plate expansion was seen from maxillary left second premolar to second molar region obliterating the buccal vestibule. The area was mildly tender to palpation. The swelling was well defined and oval shaped bony hard. [Figure 1 (b)]. Thus, based on the clinical features, the provisional diagnosis of dentigerous cyst or a benign tumor in the left maxilla was suspected.

Intraoral periapical view shows a radiolucent lesion extending from 26 region till the maxillary tuberosity region. Few radiopaque specks were noted within the lesion suggesting calcifications. The floor of left maxillary sinus is not clearly traceable. Root resorption was noted in 26 and 27 [Figure 2 (a)]. Panoramic radiograph showed the complete extent of the lesion. The margins were well-defined and corticated. The lesion extended anteroposteriorly from the region of the 25 to the distal margin of the maxillary tuberosity, and infero-superiorly from the alveolar crest of the upper left molars to the just approximately 1 cm to 1.5 cm below orbital floor. The lesion was unilocular with superiorly displaced impacted maxillary left third molar within it. Multiple small radiopaque specks were seen within the lesion, located close to the crown of the impacted tooth [Figure 2 (b)]. Computed tomography images showed a large, expansile, well defined, partially corticated heterogeneously enhancing lesion of approximate size involving left maxilla, inferiorly extending up to alveolar ridge and superiorly extending into the maxillary sinus with an unerupted tooth projecting into the lesion with minute irregular hyperdense areas that is calcifications around the crown of the impacted tooth [Figure 2 (c),(d),(e)]. The radiographic diagnosis of dentigerous cyst with dystrophic calcification or a CEOT was suspected, with differential diagnoses of adenomatoid odontogenic tumor and ameloblastoma. Excisional biopsy of the lesion was done, with extraction of 26 and 27. The impacted third molar within the lesion was removed along with the lesion. On gross examination, the specimen was ovoid in shape, approximate size of 4 cm x 3.5 cm, greyish white colour, smooth surface and firm consistency [Figure 3].

The Hematoxylin and eosin stained sections observed under low power microscope, revealed presence of sheets of polyhedral odontogenic epithelial cells with prominent hyperchromatic nuclei, cellular outline and intercellular bridges. Clear cells with vacuolated cytoplasm seen in the epithelium, 2-3 areas show amyloid like material. Multiple areas of calcification were also seen in connective tissue stroma [Figure 4 (a) and (b)]. Depending on the histopathological features, radiological and clinical findings, the final diagnosis of CEOT was made.

3 | CASE 2

A 30-year-old female patient came to our department with a growth in the upper left quadrant since the past one year. A sessile gingival mass was seen in the maxillary left first, second and third molar region with a smooth surface. The maxillary left second and third molar were displaced distally and palatally. Provisional diagnosis was made as an epulis or Benign tumor[Figure 5].

Intraoral periapical radiograph showed the distal displacement of 27 and 28 and floor of maxillary sinus was not traceable clearly [Figure 6 (a)]. Panoramic radiograph showed an area of osteolysis in 27 and 28 regions with multiple small radiopaque specks which were seen within the soft tissue mass. The tooth 28 was displaced distally [Figure 6 (b)]. Contrast enhanced computed tomography (CECT) images shows an expansile osteolytic lesion with ill-defined margins in the left maxillary alveolus. The soft

tissue mass was isodense and showed homogenous enhancement after injecting the contrast medium. The mass was also seen to be projecting into the maxillary sinus within the soft tissue, there were specks of hyperdense mass suggestive of calcifications [Figures 6 (c) and (d)].

Overall features are suggestive of a destructive lesion in the left maxilla suggested a calcified epulis or a peripheral CEOT. Incisional biopsy was done under local anesthesia. The lesion was completely excised with extraction of maxillary left second and third molar. Histological report confirmed the diagnosis of a peripheral CEOT.

4 | DISCUSSION

The recent classification of WHO (2022) is based on the biological behaviour of the tumor and from which tissues (epithelium or mesenchyme) they are derived. According to this, CEOTs are classified as benign epithelial odontogenic tumors.[4] The tumor is supposed to arise from dental lamina remnants, from the reduced epithelium or intermediate layer of the enamel organ, or even from the oral epithelium, however, the etiopathogenesis of CEOT is still doubtful.

According to the current statistics, CEOT has no sex predilection. Most patients of central and peripheral type CEOT are above 40 and 35 years of age, respectively. The present cases reported were also 39 years (central type) and 30 (peripheral type) of age. The central variety was associated with an impacted tooth. CEOTs are most commonly associated with impacted tooth (52%), frequently affecting mandibular second molar.[4,5] The central type of CEOT is usually asymptomatic for longer periods and a bony hard swelling is evident only in the advanced stages of the tumor. Clinically they present as a slowly growing asymptomatic hard swelling with an impacted tooth causing bone expansion. In our case of central CEOT patient has swelling for the last six months. The common site of occurrence of peripheral CEOTs is gingiva, especially of the maxillary anterior region.[5] Our case also had similar presentation of peripheral CEOT on the gingiva in the maxillary molar region. Kaplan et al. reported that root resorption occurred in only 4% of 67 cases. Whereas, in solid ameloblastoma root resorption is a more common feature, this might help in differentiating it from CEOT. [6] The present case also showed only mild root resorption of the apical third of the teeth involved in the lesion. On the radiograph, central CEOTs appear as unilocular or multilocular, well to ill-defined, corticated or partly corticated radiolucent lesions.[7] Lesions of the maxilla are majorly unilocular. In the mandible, the tumor is frequently associated with the impacted mandibular molars, in which mandibular second molar is more common as compared to the first and third molars.[8] In the present case, the lesion was unilocular with well-defined borders and was associated with impacted third molar. The most classical features of CEOTs are the pericoronal flecks of calcification of different shapes and sizes around the associated impacted tooth.[6] Displacement of teeth within the tumour is a common finding in central and peripheral CEOTs. In the present case of central type within the tumor the impacted tooth was displaced superiorly and in the peripheral type the teeth were displaced palatally and distally.

The classic histological feature of CEOT constitute of odontogenic epithelium exhibiting polyhedral cells with eosinophilic cytoplasm, prominent round to oval nuclei, and distinct inter-cellular bridges often arranged in sheets, islands or nests amidst the fibrous connective tissue stroma. Sometimes, the stroma exhibits eosinophilic deposits suggestive of amyloid-like material or enamel protein. It is common to find clear cells within the area of sheets of epithelial cells, particularly in the maxillary lesions. The histopathology specimen of the present case of central type CEOT also showed these typical features. The occurrence of Liesegang rings which eventually fuse to form masses of calcification are a common feature of CEOTs.[9] The peripheral and central type of CEOTs show similar histopathological features. However, Liesegang rings are not found in the present case, variants showing the multiple discrete area of calcification were noted. Recent literature shows, three different histopathological subtypes of CEOT which are the clear cell type, cystic or microcystic type and non-calcified or Langerhans cell rich type.[4] The present case 1 also can be classified as the clear cell type of CEOT. The common odontogenic benign tumors or cysts are occurred with CEOT are adenomatoid odontogenic tumor and dentigerous cyst.[10] Bouckaert et al. reported some aggressive cases of CEOTs affecting the

maxillary bone region which are encroaching the maxillary sinus, and ethmoid sinus and may shows invasion into the cranium, which are prone to infections in the brain.[10] In the case of intraosseous CEOT, left maxillary sinus is involved by the tumor. A recurrence rate of CEOTs are 14% for central type as per literature reviewed by Franklin and Pindborg and peripheral CEOTs are not showing recurrences, therefore considered as good prognosis.[10] The comparison between central and peripheral CEOTs are enumerated (Table 1). Treatment of CEOTs is mostly conservative and depends on the clinical extent and location, radiographic features, and histologic variations among the tumor.[11]

The treatment modalities of CEOTs ranges from the simple curettage or enucleation to the radical resection of the jaws. In our case of central CEOT, complete enucleation of the tumor was done and in the peripheral type, excision of lesion was done. Maxillary CEOTs required to treat more carefully because they are not within the confine and the close relation with anatomical structures surrounding maxilla.[1] The patient with central CEOT underwent complete enucleation of the tumor and no recurrence was reported in 5 months of follow up. There is very low incidence of transformation of CEOTs into malignant tumors and few such cases have been reported in the literature and may rarely show metastasis.[12-15] To prevent the recurrence of the treated cases of CEOT periodic follow up is necessary. Five years minimum follow-up period is necessary for treated patients.[16] According the currently available literature, cases of noncalcifying CEOT may also occur, but they tend to have a different presentation.[17,18] Recent literature also showed the evidence of mutations in tumour suppressor genes (PTEN, CDKN2A, PTCH1) and oncogenes (JAK3, MET) in the CEOT.[4] Hence, patients of CEOTs are kept under follow up after every three months. The patients are also under follow up and screened regularly for the recurrence.

5 | CONCLUSION

Two cases of this rare lesion have been presented here with clinical, radiographic and histological findings. A good history, careful clinical evaluation and appropriate and judicious use of advanced imaging methods like CECT, cone beam computed tomography etc., along with histological evaluation aids in early diagnosis and successful management of this rare tumor. A multidisciplinary approach with a team of oral radiologist, oral surgeons and oral pathologist is recommended for successful management of CEOT.

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Nil

Conflict of interest

The authors have no conflicts of interest to declare.

Supporting Information

Additional supporting information may be found at the journal's website.

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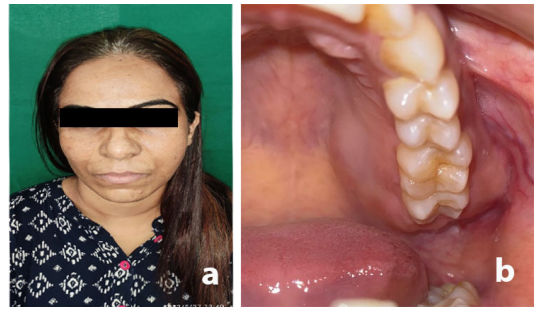

FIGURE 1 a) Extraoral picture showing swelling over left side face. b) Intraoral picture of the lesion showing bi-cortical expansile area in the left maxilla covered with normal-appearing oral mucosa.

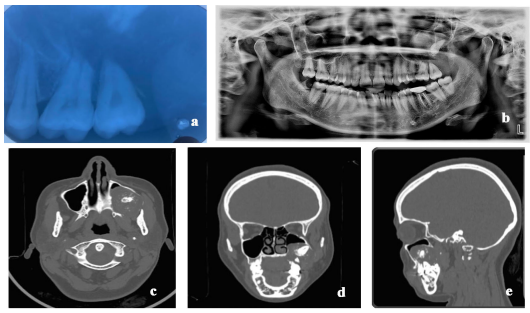

FIGURE 2 a) Radiographs showing a radiolucent lesion extending from 26 region till the maxillary tuberosity region with 26 and 27 root reposprtion and bicortical expansion. b,c,d,and e) Flecks of calcification in the interior of lesion near the crown of impacted maxillary left third molar.

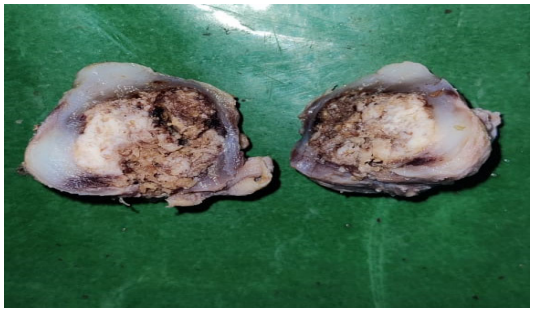

FIGURE 3 Gross specimen was ovoid in shape, approximate size of 4 x 3.5 cm, greyish white colour, smooth surface and firm consistency

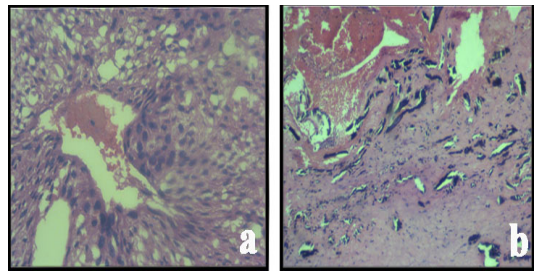

FIGURE 4 (a) H and E (100x magnification) stain section showing odontogenic epithelial cells consists of polyhedral cells with large hyperchromatic nuclei evident. (b) H and E (400x magnification) stained section showing multiple irregular calcified masses are evident.

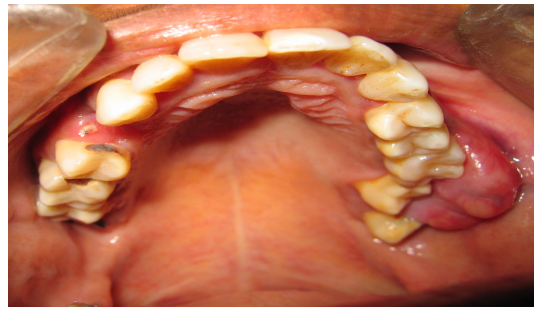

FIGURE 5 Intraoral picture showing sessile soft tissue growth in the left maxillary molar region.

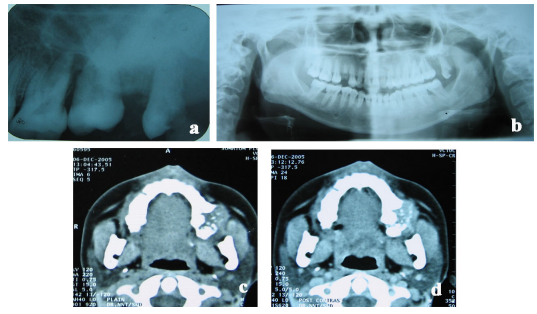

FIGURE 6 Radiograph and CT shows the osteolysis and 27 and 28 displacement along with multiple small radiopaque specks suggestive of calcifications within the soft tissue mass.

TABLE 1 The comparison between central and peripheral CEOTs

Observation	Central CEOT	Peripheral CEOT
Age	Above 40 years	Above 30 years
Sex	No gender predilection	No gender predilection
Location	Mandible >maxilla (2:1)	Maxilla >mandible
Site	Premolar- molars	Maxillary incisors gingival region
Clinical presentation		
Pain	Usually not Present	Absent
Swelling	Bony hard	Soft to firm
Asymmetry	Present	May present or absent
Radiographic presentation		Maxillary anterior teeth
Location	Mandibular and maxillary second molar >first molar >third molar	Osteolytic
Locularity	Unilocular	Ill-defined, non-corticated
Borders	Well to ill-defined and corticated	
Interior	Mixed with multiple foci of calcification (Driven snow appearance)	Mixed with discrete specks of calcification
	Impacted tooth present	
Effects on adjoining structures	Bi-cortical expansion	Not associated with impacted tooth
	Displacement of adjacent teeth	Soft tissue enlargement
	Root resorption reported	Displacement of adjacent teeth
		Root resorption not reported
Differential diagnosis	AOT, Ameloblastoma	Calcified epulis
Histopathological	Sheets of polyhedral epithelial cells.	Sheets of polyhedral epithelial cells
	Amyloid-like substance and calcified concentric deposits (Liesegangs rings)	Amyloid-like substance and calcified concentric deposits. (Liesegangs rings)
	Intercellular bridges	Intercellular bridges
	Apple-green birefringence appearance under polarized light	Apple-green birefringence appearance under polarized light
Management	Enucleation to radical surgical excision	Complete excision
Complications	Low chances of transforming into malignant lesion	Negligible chances of transforming into malignant lesion
Recurrence	14% reported	Not reported