



Case Report

Calcifying epithelial odontogenic tumour at an unusual site - A case report

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ABSTRACT

Calcifying epithelial odontogenic tumour (CEOT), also known as Pindborg tumour, is a rare benign odontogenic tumour of locally aggressive behaviour forming 0.4–3.0% of all intraosseous tumours and 1% of all odontogenic tumours. CEOT is traditionally described as a rare, benign, locally-invasive, and slow-growing odontogenic neoplasm of exclusively epithelial tissue of origin. The most frequent location is the mandibular premolar and molar area, less frequently the lesion is found in the maxilla in the ratio 3:1, typically in the fourth to fifth decades of life with no gender predilection. It usually starts as a painless swelling and is often concurrent with an impacted tooth. The present case highlight the occurrence of CEOT in a younger individual on the unusual site i.e. in the left maxillary canine and premolar region as a painless swelling. This article also attempts to briefly emphasize the concepts of CEOT with the review of literature.

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1. Introduction

Calcifying epithelial odontogenic tumour (CEOT) commonly known as Pindborg tumour was first introduced into scientific literature almost 50 years ago by late Dr. Jens J. Pindborg. CEOT is a rare benign odontogenic neoplasm, forming 0.4–3.0% of all intraosseous tumours.¹ It is an uncommon, benign, odontogenic neoplasm that is exclusively derived from odontogenic epithelium. It usually presents as a hard, painless mass, generally affecting the mandible. Few sporadic cases affecting the maxilla have also been reported. Although its biological behaviour is indolent, the maxillary tumour tends to grow rapidly and are locally invasive. The tumour shows a variable radiographic view based on its development; mixed radiolucent–radiopaque feature is the most prevalent, seen in 65% of cases. The tumour is similar in behaviour to ameloblastoma but has a slightly lesser recurrence rate after surgery. 14% recurrence rate is seen on longstanding cases. The prognosis is considered to be very favourable in most of the cases.²

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2. Case Report

A 22-year old male patient presented with the painless swelling in the left upper anterior teeth region since 1 month. Patient had noticed swelling 1 month back, which was insidious in onset gradually progressing and has slightly increased in size from the time he first noticed, not associated with any other signs and symptoms, no history of trauma and has no history of similar complaint in the past.

On extra oral examination, a solitary diffuse swelling was evident on left middle third of face extending anteroposteriorly from philtrum region to 3cm laterally on left side and superoinferiorly from ala tragal line to the line joining corner of the mouth to lobule of the ear, roughly oval in shape measuring about 3x2cm in size, skin overlying the swelling was normal, surface was smooth and margins were diffused. On palpation all the inspeactory findings were confirmed, swelling was bony hard in consistency, non fluctuant, non mobile and non tender.(Figure 1)

On intraoral examination, a solitary well-defined swelling was evident in the left premaxillary region extending anteroposteriorly from the mesial aspect of 22 to mesial aspect 25 and superoinferiorly from the marginal

gingiva wrt 23,24,25 to the base of labial and buccal vestibule, roughly oval in shape, measuring about 3x2cm in size, margins were diffused, surface appears smooth and overlying mucosa was normal. On palpation all the inspeactory findings were confirmed swelling was bony hard in consistency in the anterior aspect and firm in the posterior aspect, non fluctuant, non mobile and non tender, causing labial and buccal vestibular obliteration in the region of 22, 23, 24, 25 with labial and buccal cortical plate expansion without noticeable palatal expansion. Grade I mobility wrt tooth 23, spacing between the teeth 23 and 24 and distopalatally rotated 24 was evident.(Figure 2)

Based on history and clinical findings a provisional diagnosis of benign odontogenic tumour involving left premaxillary region was given and clinical differential diagnosis of Adenomatoid odontogenic tumour and Calcifying epithelial odontogenic tumour were considered.

Intraoral periapical radiograph of 23,24 region showed, a solitary well defined unilocular mixed radiolucent radiopaque lesion interdentially between the roots of 23 and 24, extending anteroposteriorly from the distal aspect of root of 23 to mesial aspect of root of 24 and superoinferiorly from floor of maxillary sinus to about 1.5cm inferior to it, circular in shape measuring about 1.5cm in diameter, margins are well-defined and non corticated, internal structure appears mixed radiolucent and radiopaque with multiple small radiopaque foci of calcification giving granular pattern and lateral displacement of roots of 23 and 24 was evident.(Figure 3)

Orthopantomograph (OPG) showed presence of unilocular mixed radiolucent radiopaque lesion between the roots of 23 and 24, extending superoinferiorly from floor of maxillary sinus to 1cm above the alveolar crest, measuring about 2x2cm in size with corticated margins and mixed radiolucent radiopaque internal structure casing superior shift in the floor of maxillary sinus in the region of 23 and 24 was seen.(Figure 4)

Cone beam computed tomography scan was taken and the following 3D radiographic features were analysed.

2.1. Coronal sections

Showed presence of a solitary well defined mixed radiolucent radiopaque lesion in the maxillary left anterior region, roughly circular in shape, measuring about 23.1x23mm, extending anteroposteriorly from 23 to 25 regions and superoinferiorly from floor of maxillary sinus to 7mm above the alveolar crest, margins are well defined and corticated, internal structure shows multiple radiopaque foci of varying size dispersed all through out the lesion, the lesion is in close proximity to the left antrum region and causing lateral displacement of the 24 and 45. Loss of lamina dura on distal aspect of root of 24 and mesial aspect of root of 25.(Figure 5 C)

2.2. Sagittal section

Showed thinning and expansion of labial cortical plate with perforation of labial cortex in few areas.(Figure 5 B)

2.3. Axial sections

Showed labial cortical plate expansion and thinning with thinning and perforation of left lateral nasal wall. Few radiopaque foci of calcification with positive HU values (+447) is seen.(Figure 5 D)

Based on the above mentioned radiographic findings, radiographic differential diagnosis of Extrafollicular Adenomatoid Odontogenic tumour and Calcifying Epithelial Odontogenic Cyst were considered.

FNAC was done from the posterior aspect of the lesion resulted in the aspiration of 0.5ml of blood tinched fluid, cytosmear showed presence of few inflammatory cells and RBCs.

Incisional biopsy of the lesion showed features consistent with CEOT. Excision of the lesion was done under local anaesthesia. Histopathology showed infiltrating lesion composed of fibrous stroma with islands and sheets of polyhedral epithelial cells with eosinophilic to clear cytoplasm and sharply defined cell borders. Mild nuclear pleomorphism is seen and few eosinophilic hyaline material and numerous calcified deposits, few with concentric lamellation and wide areas of calcification is seen. All the findings confirmed the diagnosis of Calcifying Epithelial Odontogenic Tumour and the patient is under follow up.



Fig. 1: Diffuse extraoral swelling on left side of face



Fig. 2: Well defined swelling on left premaxillary region with buccal cortical plate expansion

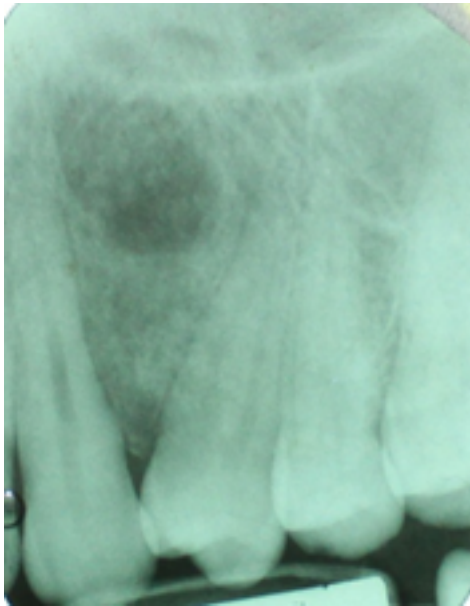


Fig. 3: Intra oral periapical radiograph wrt 23, 24



Fig. 4: Orthopantomograph showing mixed radiolucent radiopaque lesion wrt 23 and 24

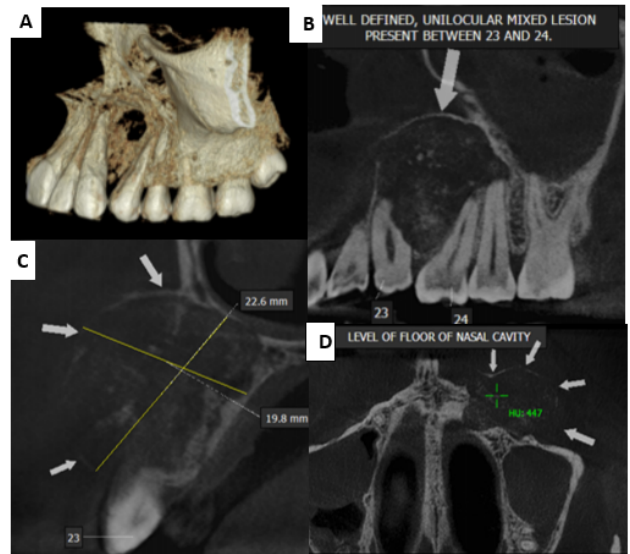


Fig. 5: CBCT images **A:** 3D reconstructed image. **B:** Coronal section of CBCT showing mixed radiolucent radiopaque lesion between the roots of 23 and 24. **C:** CBCT sagittal section – showing mixed radiolucent radiopaque lesion causing buccal cortical plate expansion, **D:** Axial section at the level of mid of right and left antrum- close proximity of the lesion to left antrum

3. Discussion

Calcifying epithelial odontogenic (CEOT) is a rare benign odontogenic tumour comprising less than 1% of all odontogenic neoplasms. CEOT was previously described in the literature as adenoid adamantoblastoma, ameloblastoma of unusual type with calcification.³ Thoma and Goldman described the tumour as a neoplasm arising from the odontogenic epithelium; subsequently, the German pathologist Jorgen Pindborg recognised it as a separate entity in 1955 later in his honour, this lesion was termed as the Pindborg tumour. In 1967, The term ‘CEOT’ has been accepted by the WHO in the first edition of ‘Histological Typing of Odontogenic Tumours, Jaw Cysts and Allied

Lesions - 1992', where it was recognised as a distinct entity.⁴

3.1. Clinical presentation

Clinically, CEOT presents as a slow growing painless expansile bony hard swelling causing cortical plate expansion, manifests as an intraosseous lesion (central type) in the majority of cases (95%). Extraosseous or peripheral lesions account for less than 5% of cases. It is most commonly reported in patients during the 4th, 5th, and 6th decades of life with only 7% of the reported cases occurring in children.⁵ The mean age range is 33–43 years with no sex predilection. The most common site of occurrence is the mandibular premolar and molar region - 57% and 14% of cases reported in mandibular anterior region and less commonly seen in maxilla with 8% in maxillary anterior region and 21% in maxillary premolar and molar region. When present in the maxilla, the CEOT is preferentially located in the posterior region and are more aggressive.² Most CEOT cases, nearly 52%, are associated with impacted or unerupted teeth or odontomas. CEOT with extension into the maxillary sinus is uncommon.⁶ Less common peripheral variant of CEOT presents as a nodular mass on gingival mucosa in the anterior region.⁷

3.2. Radiographic presentation

Depending on stages of development, CEOT may present variable radiographic appearances. The lesion usually consists of a radiolucent area, which is well-defined. CEOT is often unilocular when small and larger lesions tend to have honeycomb or soap bubble appearance.⁸

CEOT are generally unilocular lesions; although 5–13% of cases have shown multilocular appearance. Variable numbers of radio-opaque bodies are seen in about 50% of CEOT cases. It may have regular outline with well-demarcated margins. Early tumors may appear completely radiolucent. With maturation, they develop calcifications which may show mixed radiolucent-radiopaque appearance. Marx et al. in 2003 discussed three patterns of radiopacity with this tumor; first, salt and pepper pattern of flecks, second, fluffy cloudlike pattern throughout, and third, a crescent-shaped pattern on one side of the radiolucency. In the present case salt and pepper pattern of calcification was seen in CBCT. The mixed radiolucent and radiopaque pattern occurs most often (65%) followed by the completely radiolucent pattern (32%) and least often the totally radiopaque "snow driven" pattern (3%).⁹ When tumour is associated with impacted tooth, it may appear as peri coronal radiolucency with or without small radiopacities.¹⁰

3.3. Histopathology

The histological criteria of the Pindborg tumour are layers of polyhedral epithelial cells with well-defined borders that

oftentimes show prominent intercellular bridges. Figures of mitosis are rarely seen. In the layer of epithelial cells, circles, full of a homogenous amyloid-like substance, were observed. Some of those cells were also filled with a calcifying matter in the form of Liesegang rings, which are pathognomonic of this tumour.¹¹

In addition to the classic histologic appearance of the CEOT, the deposition of amyloid-like substance is another unique feature. There has been controversy over the origin of this homogenous material. El-Labban suggests the amyloid in CEOT is derived from degradation of lamina densa material, secreted by the tumour epithelial cells. Page performed an ultra-structural study of CEOT which showed that the amyloid material is a protein product of the enamel organ completely different from those seen in endocrine-associated amyloid or systemic amyloid. Amyloid-like material in CEOT shows green birefringence by Congo-red stain, which has been suggested as a useful stain for differentiating CEOT from other lesions.¹²

Although CEOT is typically benign, its behaviour varies depending on the histologic features and location. Necrosis, high proliferation index assessed by Ki-67, and nuclear pleomorphism are associated with a more aggressive behaviour. Furthermore, involvement of the maxilla or the maxillary sinus is associated with rapid growth and invasion of the orbits and skull base. Intraosseous involvement is another feature that is associated with higher chance of recurrence as compared to extraosseous tumour. In contrast, the presence of calcification and amyloid-like material indicates more differentiation and a lower likelihood of recurrence.¹²

3.4. Treatment and prognosis

The treatment plan in cases of CEOT depends on multiple factors such as size and location of neoplasm and general condition of patient. If the tumour is small intrabony mandibular lesions with well-defined borders, then simple enucleation or curettage followed by judicious removal of a thin layer of bone adjacent to the tumour is enough. Large tumours have to be approached by segmental resection and reconstruction using grafting or distraction osteogenesis. As these tumours are not encapsulated, it is commonly agreed that the resection should include a safe margin of clinically and radiographically healthy bone.¹³

The prognosis of the CEOT is good with infrequent recurrence. Malignant behaviour is extremely rare. Although it has not been established in the literature, five years should be the absolute minimum follow-up necessary to assess the healing for this type of odontogenic tumours.¹⁴

4. Conclusion

Calcifying epithelial odontogenic tumour is a benign yet locally aggressive neoplasm. It can exhibit unpredictable

clinical behaviour. Because of variations in the clinical and histologic features of CEOT, treatment should be individualized for each lesion. Achieving a correct diagnosis is crucial for obtaining a tumour-free margin. Overall, a range of clinical, histopathologic, and radiographic examinations should be helpful in diagnosing and treating CEOT.

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None.

6. Conflict of Interest

The authors declare that there is no conflict of interest.

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