Calcifying epithelial odontogenic tumour of the maxilla – A rare case report

Annette M. Bhambal, Akhil Trivedi, Priyanka Deshmukh, Shivakumar G.C.
Department of Oral Medicine and Radiology, People’s College of Dental Science and Research Centre, Bhopal, MP, India

Abstract
A calcifying epithelial odontogenic tumour (CEOT) is a rare benign odontogenic tumour, which was first described by J. J. Pindborg in 1955, hence it is also called the Pindborg tumour. It constitutes less than 1% of the entire odontogenic tumours and 0.4–3.0% of all intraosseous tumours. The origin of CEOT is still unclear. It may be derived from the oral epithelium, reduced enamel epithelium, stratum intermedium or remnants of the primitive dental lamina. Surgical management is usually enucleation and the recurrence rate is 15%. In this case report, we have described a case of CEOT that occurred in the left posterior region of the maxilla in a young adult male which clinically appeared as a peripheral lesion but radiological findings showed a central variant.

Keywords: Calcifying epithelial odontogenic tumour, driven snow pattern, Liesegang rings, Pindborg tumour

INTRODUCTION
A calcifying epithelial odontogenic tumour (CEOT) or Pindborg tumour is a rare benign odontogenic tumour that was first described by Jens Jorgen Pindborg in 1955.[1‑5] It constitutes less than 1% of the entire odontogenic tumours and 0.4–3.0% of all intraosseous tumours.[6] CEOT may be derived from the oral epithelium, reduced enamel epithelium, stratum intermedium or from the remnants of the primitive dental lamina.[3] The recurrence rate is 15%.[7]

This case report describes CEOT in the maxillary region of a young adult male which clinically appeared as a peripheral lesion but radiological findings showed a central variant.

CASE HISTORY
A 22-year-old male reported with a chief complaint of painless growth on the left upper posterior teeth region since 6 months. It has started as a small nodule in the left maxillary posterior gums, which gradually increased to the present size. There was no associated pain, pus discharge or loss of sensation. However, he had discomfort while chewing food. The patient was a habitual tobacco chewer for the past 4 years. Family, medical history and general examination were non-contributory.

Extraorally there was a mild facial asymmetry on the left middle one-third of his face due to the lesion, which extended antero-posteriorly from the left ala of the nose to the tragus. Superio-inferiorly, it extended...
from the infraorbital margin to an angle of the mouth. The overlying skin appeared normal. On palpation, the lesion was bony hard and non-tender. A single left submandibular lymph node was palpable, mobile and non-tender [Figure 1].

Intraoral examination showed a well-defined growth present in the left maxillary alveolar region measuring 3 × 5 cm. Antero-posteriorly it extended from 23 to 27. While medio-laterally it extended from the midline of the palate, crossed the interdental space in 24, 26 and obliterated the buccal vestibule. The lesion was hard in consistency, lobulated and non-tender. Twenty-four and 26 were vital with grade 2 mobility. Twenty-five was clinically missing. Fine needle aspiration cytology (FNAC) was negative for fluid [Figure 2].

Based on clinical examination, a provisional diagnosis was given as ameloblastoma. Differentials included were ossifying fibroma, central giant cell granuloma, dentigerous cyst, pleomorphic adenoma of the palate and intraosseous mucoepidermoid carcinoma.

Orthopantamogram (OPG) revealed a well-defined, unilocular, radiolucent lesion in the region of 25, which was present between 22 and 27. The lesion had a distinct sclerotic border superiorly, anteriorly and posteriorly with an indistinct border inferiorly. 25 was displaced apically. Root resorption in 24, 26 and 27 and the displacement of roots in 24 and 26 were observed. Radiopaque flecks were present within the radiolucency [Figure 3]. Radiographic differentials made were dentigerous cyst, adenomatoid odontogenic tumor (AOT), CEOT, calcifying odontogenic cyst, ameloblastic fibro-odontoma, odontogenic fibromyxoma. Enucleation of the lesion was done where the lesion had easily detached from the adjacent bone [Figure 4].

The authors obtained all appropriate patient consent.

Gross microscopic examination showed a single mass of soft, oval-shaped firm tissue, which was greyish in colour and measured 3 × 2 cm in size and associated with an embedded tooth [Figure 5]. Microscopically, the tissue section showed sheets and islands of odontogenic
epithelial cells polyhedral in shape with prominent intercellular bridges. Few clear cell differentiation showing dark basaloïd nuclei, eosinophilic material was observed. ‘Liesegang-ring’ (small irregular foci of concentric calcifications) pattern of basophilic calcifications was also noted [Figures 6 and 7]. The final diagnosis of CEOT was established based on microscopic features.

**DISCUSSION**

CEOT is a rare benign odontogenic tumour, which was first described by Dutch pathologist, Jens Jorgen Pindborg in 1955, hence it is also called a Pindborg tumour. CEOT consists of two varieties - intraosseous or central CEOT and extraosseous or peripheral CEOT; the incidence of central CEOT being 94%. The peripheral variant of CEOT is extremely rare and only 10 cases have been reported (5%). It presents clinically as a nodular mass on gingival mucosa in the anterior tooth location. This tumour occurs between 30 and 50 years of age with no preference for gender. Maxilla is less commonly affected than mandible in the ratio of 1:3. In the maxilla, the lesion is usually located in the posterior molar region. However, CEOT with extension into the maxillary sinus is a rare finding.

In our case, the patient was a 22-year-old male, who had a painless swelling in the left side of the maxillary canine and premolar region with associated bicortical expansion. The age, site and clinical features of the lesion were similar to literature written by Philipsen and Reichart and Shafer et al.

Ideally, CEOT in a radiograph presents itself as a unilocular or multilocular radiolucent lesion together with scattered flecks of calcification internally thereby giving rise to a ‘Driven Snow’ appearance. Only 52% of the reported cases have been associated with impacted/unerupted teeth. Our case revealed similar radiographic features along with the presence of an impacted 25. Resorption of roots of 24, 26 and 27 was also observed which is an uncommon finding according to some authors.

The treatment for CEOT is surgical enucleation, but the treatment plan may vary among patients and may range from simple enucleation to hemi-mandibulectomy/ maxillectomy. Literature recommended performing curettage or maintaining a margin of safety with clinically healthy bone removal. In our case, since our patient was in the third decade of life and the tumour could be easily excised from the bone, conservative treatment was opted.

CEOT was histologically characterized by the presence of three components – polyhedral often pleomorphic epithelial  

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**Figure 5:** Surgical specimen of the lesion with embedded tooth

**Figure 6:** High power photomicrographs (H & E, × 40) showing the presence of islands and sheets of polyhedral cells, areas of extracellular amyloid-like eosinophilic material and small irregular foci of calcification

**Figure 7:** (H & E, × 100) Showing nests of polyhedral cells forming intercellular bridges and nuclear pleomorphism
cells with prominent intercellular bridges, amorphous amyloid-like depositions and calcifications.\textsuperscript{14-16,20} The morphology and size of the nuclei can vary and are associated with ‘Liesegang-rings’. ‘Liesegang-rings’ are extracellular, eosinophilic, amyloid-like material areas of concentric calcifications, which are found in the connective tissue stroma. According to Krolls and Pindborg, the presence of calcification in CEOT may have prognostic implications. The less calcification indicates more chances of recurrence with a poorly differentiated tumour.\textsuperscript{11}

Similar histopathological features were also present in our case with calcification, which indicated a well-differentiated tumour and lesser propensity for recurrence.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

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