

Case Report

Pindborg Tumor- Cystic Variant or Entity: A Rare Case Report, Differential Diagnosis and Literature Review

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ARTICLE INFO



Keywords: CEOT, Leisgang Ring, Pathogenesis, Prognosis, AOT, mixed radio-opaque-radiolucency, Pindborg tumor, odontogenictumor, cone beam computed tomography, calcifying odontogenic cyst

ABSTRACT

Pindborg tumor (Calcifying epithelial Odontogenictumor CEOT) are associated with impacted teeth and might be mistaken for a dentigerous cyst on radiographs. CEOT has distinct histologic characteristics, however among its multiple histologic subtypes, the cystic variant is an infrequent and poorly characterised entity. We report a rare case of a young female patient with an anterior maxillary enlargement that turned out to be a CEOT and the current case being a unique cystic variant of CEOT that is clinically resembling Adenomatoid Odontogenic Tumor (AOT), along with an analysis of previous documented cases that can assist clinicians in developing a better treatment strategy and prognosis. Hence, documenting this uncommon case, we suggest to hypothesize that CEOT can be categorized into two types: cystic variant and neoplastic variant where each of these genotypes may have distinct prognosis. Cystic CEOT should thus be considered a histomorphological variant of CEOT.

Introduction

Dr. Jens Pindborg originally reported the Pindborg tumour (CEOT) in 1956, later on in 1992 WHO classified it as a benign odontogenic tumour of epithelial origin. It accounts for less than 1% of all odontogenic tumours. CEOT accounts for around 1% of all odontogenic tumours. [1,2]

CEOT is thought to be arise from oral epithelium, reduced enamel epithelium, stratum intermedium, or dental lamina remnants. [3]

CEOT is distinguished by the presence of amyloid-like material that could also calcify. [4]

Remarkably, despite the rarity of CEOT instances, numerous variants have been discovered, including the presence of clear cells, Langerhans cells, myoepithelial

cells, bone, and cementum-like components, as well as hybrid tumours like CEOT mixed with AOTs or ameloblastoma. [4]

We intend to present a unique cystic variant of CEOT clinically resembling AOT developing in the anterior maxilla, as well as an analysis of previous documented cases (Table 1), which will help clinicians establish an improved treatment approach and prognosis.

Clinical presentation

A young (23-year-old) female patient reported with swelling in the upper left front tooth region since 1 month. She remained asymptomatic for about a month before noticing swelling in the maxillary left anterior

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tooth site, which eventually grew to its current capacity. There were no aggravating or alleviating factors, and the swelling progressed at a gradual rate accompanied by Intermittent, non-radiating pain. Clinical examination revealed a 2-3 cm solitary round to oval swelling extending from 22 to 24 teeth in the maxillary left anterior vestibule (Figure 1A). The lump was tender on palpation and had a soft consistency with well-defined margins. An intraoral periapical radiograph revealed an impacted canine with well-defined radiolucency circumscribing the crown of the canine till

the middle third of its root (Figure 1B). In the pericoronal region of impacted canine (23), cone beam computed tomography revealed a well-defined osteolytic mixed radiolucent radiopaque lesion. There were also a number of small spherical opacities which resembled like calcifications (Snow driven appearance). With mild cortical expansion, there was thinning of the palate cortex and effacement of the buccal cortices (Figure 1C). The features are suggestive of Benign Odontogenic tumor associated with impacted #23.

Table 1: Review of reported cases of cystic variant of CEOT

| Cases Reported | Age (years) | Gender | Site | Clinical Presentation | Radiological Findings | Histopathological findings (H&E) | Special stains |
|------------------------------------|--|--------|---------------|---|---|--|--|
| Gopalakrishnan et al. ⁶ | 15 | M | Left Maxilla | NA | Unilocular radiolucent lesion with calcified deposits with impacted tooth (#27) | Area of transition from thin dentigerous cyst like lining into thicker CEOT cystic epithelium with majority of the lining showed classic features of CEOT. | Congo red stain +ve |
| Channappa et al. ⁷ | 30 | M | Left Maxilla | Diffuse painless swelling | Unilocular mixed-density lesion along with calcifications with impacted tooth (#25) | Cyst lined by odontogenic epithelium of uniform thickness, with classic features of CEOT. | Congo red stain +ve |
| Azevedo et al. ⁸ | The author carried out an immunohistochemical study on CEOT, with 3 cystic variant of CEOT histologically (Separate case details were not mentioned by the author) | | | | | | |
| Barreras et al. ⁹ | 31 | M | Left Mandible | Diffuse rapidly growing Asymptomatic swelling | Unilocular mixed radiopaque/lucent without presence of impacted tooth | Cystic portion featured odontogenic epithelial lining with clear cytoplasm and centrally located round nucleus; and presence of calcified material with characteristics of osteodentin. Diagnosis of clear cell cystic variant of CEOT was made. | Congo red stain +ve and PAS-Diasrase stain for clear cells |
| Kamboj et al. ² | 16 | M | Left Maxilla | Pain, swelling | Homogenous radiolucent lesion involving impacted tooth in centre (#27) | Cystic portion is lined by odontogenic epithelial lining of varied thickness with majority showed classic features of CEOT and areas of eosinophilic amyloid like material and multiple calcifications. | Congo red stain +ve |

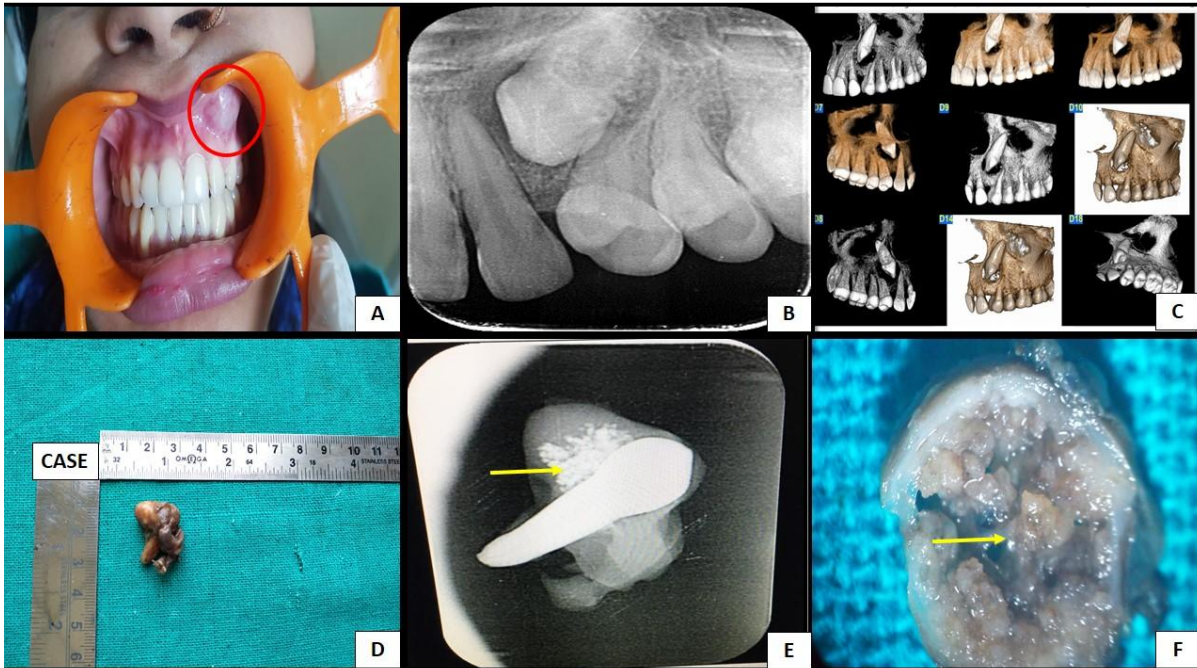


Figure 1: A) Well defined ovoid to round swelling with well-defined margins wrt 23-24 teeth region
 B) Intraoral periapical radiograph shows impacted canine with well- defined radiolucency circumscribing the crown of canine till the middle third of its root
 C) CBCT scan of the lesion depicting the cortical expansion
 D,F) Macroscopic examination of the lesion depicting intraluminal proliferations of the lining epithelium
 E) Specimen radiography reveals flecks of calcification associated with the lesion

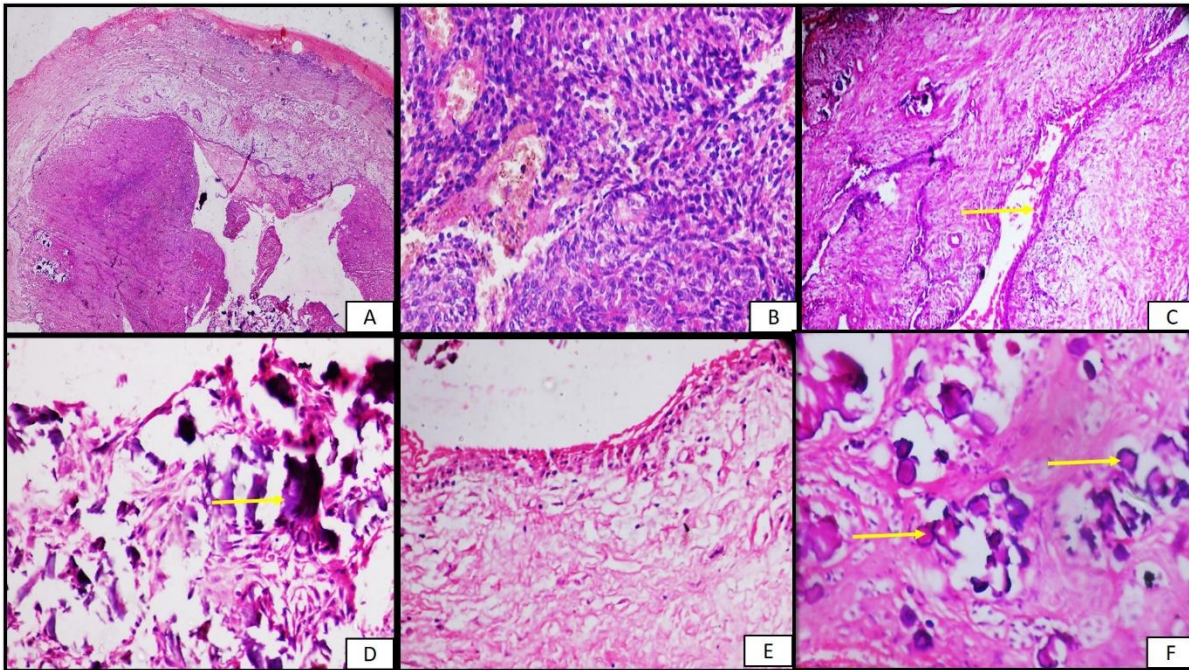


Figure 2: A) Intraluminal proliferation of cystic lining in the form of sheets and nests (H and E, 4x)
 B) Sheets and nests of cells are composed of polygonal cells with prominent junctions suggestive of CEOT like areas (H and E, 40x)
 C) Reduced enamel epithelium like lining in focal areas (H and E, 10x)
 D) Flecks of basophilic amorphous material suggestive of dystrophic calcifications (H and E, 40x)
 E) Higher magnification of the REE like lining (H and E, 40 x)
 F) Amorphous, acellular, globules of eosinophilic material surrounded by a rim of basophilic material suggestive Liesegang ring (H and E, 40x)

DIFFERENTIAL DIAGNOSIS

Dentigerous cyst, adenomatoidodontogenic tumour, calcifying cystic odontogenic tumour, and calcifying epithelial odontogenic tumour were among the differential diagnoses based on the clinical and radiological findings.

Dentigerous cysts are always found on the crown of a tooth that is impacted, embedded, or unerupted. The mandibular and maxillary third molars or maxillary cuspids are the most typically affected teeth, and it

usually occurs in the second and third decades of life, with a male predisposition.

AdenomatoidOdontogenic tumour is a benign odontogenic tumour that mostly affects men in their second decade of life. Radiographically, 2/3rd of the intrabony variants had distinct radiopaque foci with a flocculent pattern. Follicular, extrafollicular, and peripheral AOT are the three types of AOT (maxillary or mandibular canine). The current case is similar to AOT's follicular variant.

The rare odontogenic neoplasm calcifying epithelial odontogenic tumour is thought to have originated in the

stratum intermedium. CEOTs primarily afflict people in their forties and fifties, with an equal gender propensity. It manifests as a painless swelling that affects the mandibular molar area most, followed by premolar region, and ultimately the whole maxilla. Unilocular well-defined radiolucency with scattered flecks of calcification can be noticed radiographically.

Calcifying Odontogenic cyst is a benign Odontogenic tumour that affects both men and women in their second decade of life. Clinically, it manifests as a non-painful swelling in both jaws, but most frequently in the anterior mandibular region. Unilocular radiolucency with scattered flecks to masses associated with impacted tooth can be seen on radiographs.

To establish a final diagnosis, an excisional biopsy was performed. Specimen radiography revealed scattered specks of calcification with a "driven snow" appearance (Figure 1E). Stereomicroscopy revealed that the cystic cavity was filled with several polypoid or papillomatous, pedunculated, exophytic masses (Figure 1F).

A well-encapsulated cystic lesion with intramural proliferation of sheets of epithelial cells was discovered on histopathological examination. Reduced enamel epithelium-like lining was observed in a focal area, along with globules or flecks of amorphous eosinophilic cellular material in the stroma with a basophilic hue suggesting calcification. There was eosinophilic material encircled by a ring of basophilic material that resembled like that of a Liesegang ring (Figure 2).

Discussion

Intraosseous CEOT is the most prevalent of the CEOT variants, accounting for the majority of CEOT cases. This can be further classified into occurrences with or without an unerupted tooth present. [5] The tumour was

found to be associated with an unerupted maxillary canine in the present case.

The presence of varied morphological regions in the CEOT suggests that Odontogenic remnants have pluripotent potential. [6] The dental follicle has also been hypothesised as origin of several cysts, and can exhibit proliferative and metaplastic alterations. This emphasizes the pluripotential character of dental follicle differentiation and so verifies the occurrence of CEOT-like areas inside the dental follicle as demonstrated in this case.

The appearance of reduced enamel epithelium-like lining in the current case was found by histopathological evaluation, implying that the function of reduced enamel epithelium in the origin of CEOT, as established by Pindborg et al. [5,8] It is indeed possible that the current case is the outcome of neoplastic alteration of the affected canine's dental follicular lining.

Financial support and sponsorship: Nil.

Conflicts of interest: There are no conflicts of interest.

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