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Calcifying epithelial odontogenic (Pindborg) tumor involving a 16-year-old girl with no prominent clinical manifestation: A case report

Alireza Halaj-Mofrad DDS, MSc[1], Moones Rajabi DDS, MSc[2], Shahriar Dabiri MD, FIAC[3]

Case Report

Abstract

BACKGROUND AND AIM: The calcifying epithelial odontogenic tumor (CEOT) is a rare entity described by Pindborg and represents < 1% of all odontogenic tumors. Nearly 200 cases of this neoplasm have been reported to date. It mostly occurs in the posterior mandible associated with an impacted tooth, most often a mandibular molar. Patients are usually between 30 and 50 years of age, with no sex predilection.

CASE REPORT: Presented here is a case of an intraosseous CEOT, which occurred in a 16-year-old female in the body of the mandible extending from tooth nos. 32 to 47 with no specific clinical manifestations.

CONCLUSION: CEOT accounts for < 1% of all odontogenic tumors. Approximately 200 cases have been reported today. This case report describes the clinical, radiographic and microscopic features of a large CEOT in a 16-year-old girl, which although was big in size, had no specific clinical manifestations. The patient reported a relatively rapid evolution of the tumor in the mandibular region, which is a unique manifestation.

KEYWORDS: Calcifying Epithelial Odontogenic Tumor, Intraosseous, Odontogenic Tumors

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The calcifying epithelial odontogenic tumor (CEOT), known as Pindborg tumor, a rare benign odontogenic neoplasm of the jaws, was first described by Thoma and Goldman as a separate clinicopathological entity.[1] Later, Pindborg described four cases of this unusual lesion;[2] subsequently Shafer et al. used the term Pindborg tumor.[3] Nearly 200 cases of this neoplasm have been reported to date.[4] This neoplasm may have derived from the oral epithelium, reduced enamel epithelium, or stratum inter medium.[5] It most often occurs in the posterior mandible associated with an impacted tooth, most often a mandibular molar.[6] Patients are usually between 30 and 50 years of age, with no sex predilection. The most common sign is a painless, slow-growing mass.[4]

This paper describes the clinical, radiographic and microscopic features of a large CEOT in a 16-year-old girl, which although it was big in size, had no specific clinical manifestations.

Case Report

A 16-year-old female reported to the department of oral and maxillofacial pathology, School of dentistry in Kerman, Iran, with an incidental finding of a lesion during radiographic picture taken for orthodontic purpose. The lesion measured

1- Assistant Professor, Kerman Oral and Dental Disease Research Center AND Department of Oral and Maxillofacial Surgery, School of Dentistry, Kerman Social Determinants on Oral Health Research Center, Kerman University of Medical Sciences, Kerman, Iran
2- Assistant Professor, Kerman Oral and Dental Disease Research Center AND Department of Oral and Maxillofacial Pathology, School of Dentistry, Kerman Social Determinants on Oral Health Research Center, Kerman University of Medical Sciences, Kerman, Iran
3- Professor, Department of Pathology, Afzalipour School of Medicine, Kerman University of Medical Sciences, Kerman, Iran
Correspondence to: Moones Rajabi DDS, MSc
Email: moones.rajabi@gmail.com

33.7 × 12.4 mm and extended from tooth nos. 32 to 47. The patient had no complaint of the lesion but a very slight swelling in the buccal table of alveolar bone (Figure 1).

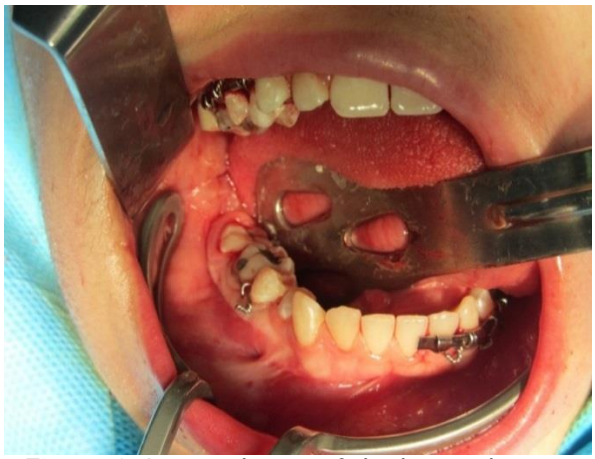


Figure 1. Intraoral view of the lesion showing slight expansion of the buccal cortex

The patient reported a clinical evolution of only 2 months of the lesion which shows a relatively rapid evolution of the tumor in the mandibular region. No loosening of teeth, resorption of roots or tenderness in percussion was reported. Orthopantomograph (Figure 2) revealed a well-defined mixed lesion in the right area of the mandible, extending from mesial aspect of left lateral incisor to mesial aspect of right second mandibular molar and causing divergence of teeth adjacent to the lesion. The lesion shows radiopaque core. Lingual and buccal mandibular cortical plates show expansion, but no alveolar bone destruction can be traced. The provisional clinical diagnosis of adenomatoid odontogenic tumor, calcifying odontogenic cyst and ameloblastic fibro-odontoma was made. An incisional biopsy was sent for histopathological examination. A neoplasm composed of polyhedral epithelial cells arranged as sheets and anastomosing small and large islands.

These cells were interspersed by homogeneous hyaline acellular material. Areas of concentric lamellated calcifications were seen. The neoplastic cells have abundant eosinophilic cytoplasm and hyperchromatic mildly pleomorphic nuclei, few bizarre nuclei were seen, however, no abnormal mitosis was seen (Figure 3, A and B). The eosinophilic material was confirmed as amyloid upon Congo red staining, confirming the diagnosis of CEOT (Figures 4 and 5).

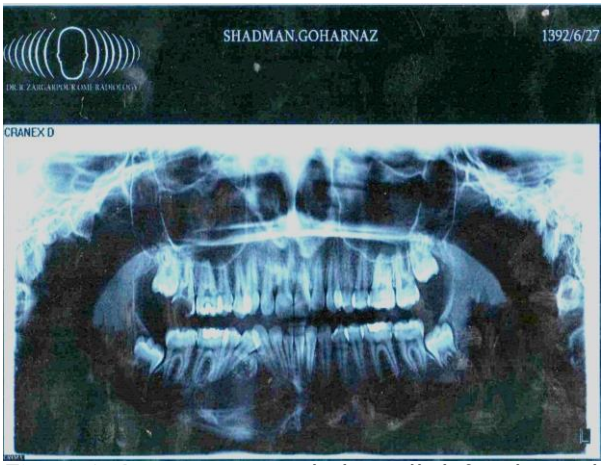
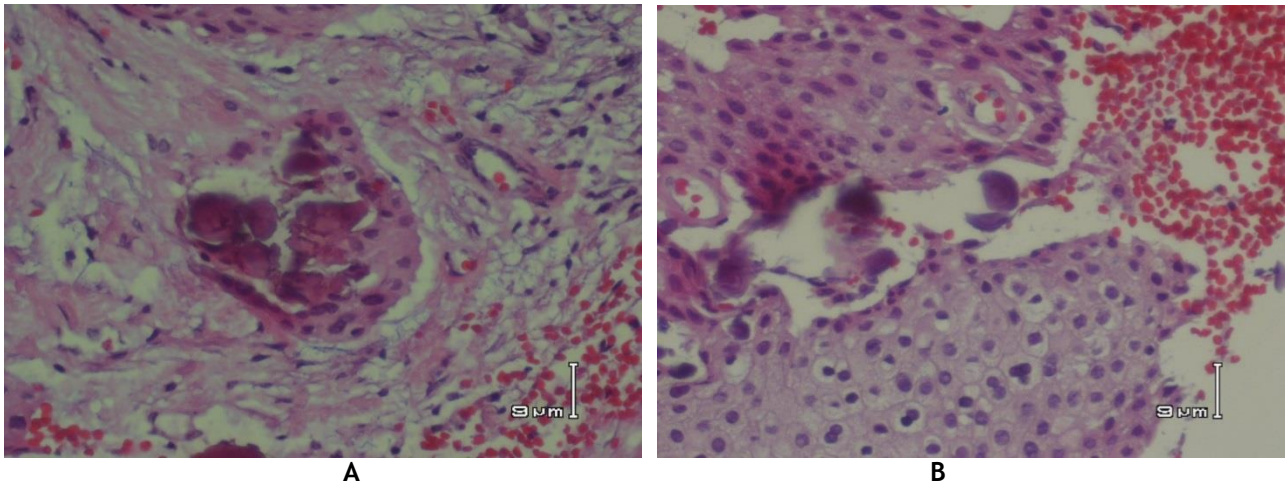


Figure 2. Panoramic revealed a well-defined mixed lesion in the right area of the mandible, causing divergence of teeth adjacent to the lesion

A pre-surgical computed tomography scan (Figure 6) was obtained to ascertain the diagnosis and boundaries of the neoplasm. It revealed an osteolytic lesion with foci of calcifications. For treatment, resection of the involved portion of the mandible and reconstruction with an AO UniLock 2.4 mm reconstruction plate was planned. The mandible was exposed via an extraoral approach and resection from 32 to 47 with safe clinical margins of 1.5 cm was performed, and the defect was bridged by 2.4 mm UniLock reconstruction plate. Furthermore, iliac crest of the patient was used for reconstruction. Resected specimen was dark brown to gray in color surface measuring 7.5 × 5.5 × 4 cm, demonstrating perforation of the buccal crest. Specimen was submitted for histopathological examination where it was confirmed as CEOT. The patient was followed for a year after the surgery and revealed no recurrence.



A B

Figure 3. Photomicrograph showing Liesegang rings and polyhedral epithelial cells with pleomorphic nuclei (H and E × 400) (A and B)

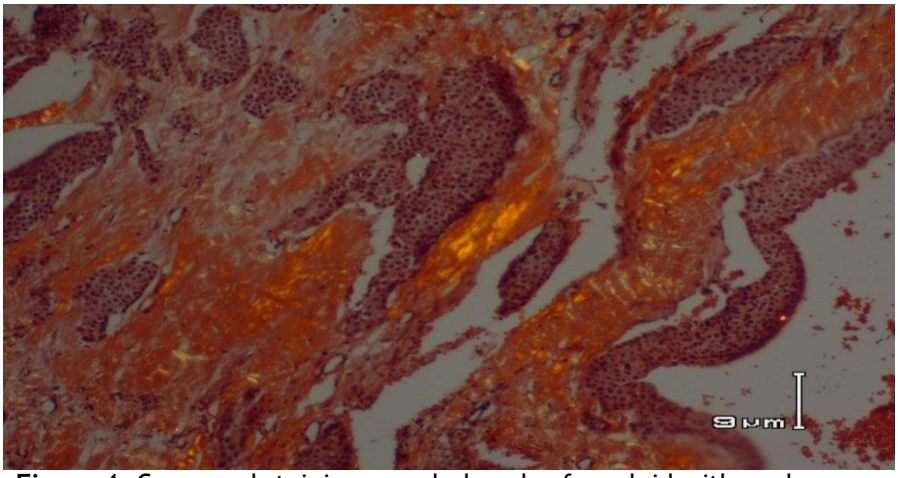


Figure 4. Congo red staining revealed pools of amyloid with apple-green birefringence when viewed with polarized light (× 100)

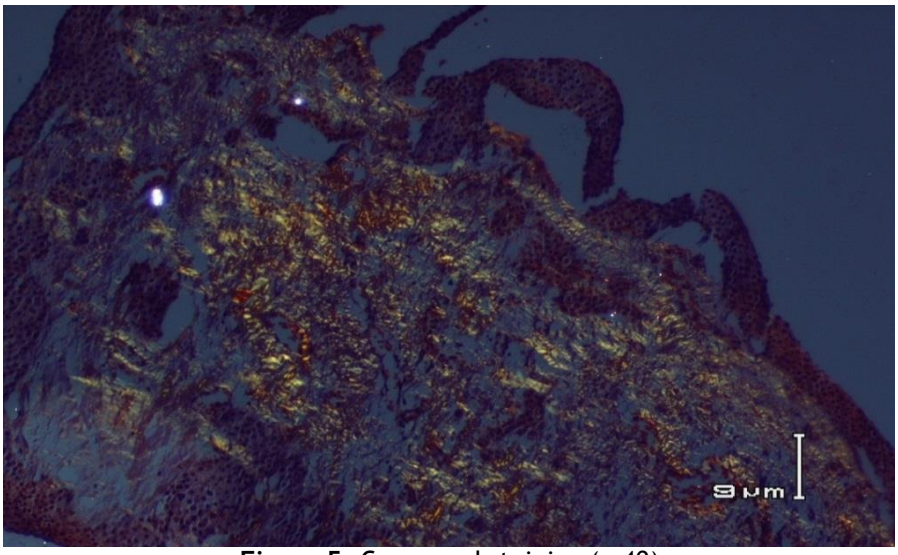


Figure 5. Congo red staining (× 40)

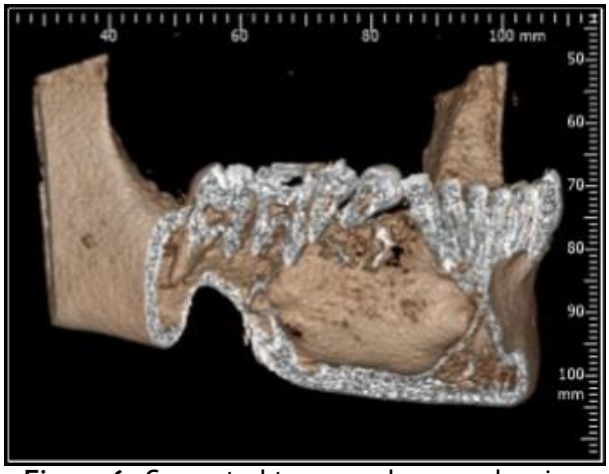


Figure 6. Computed tomography scan showing the boundaries of an osteolytic lesion with foci of calcifications

Discussion

CEOT is a benign epithelial odontogenic tumor, which is termed Pindborg tumor after the name of the founder in 1955.[2] In recent years, some variations have been introduced, such as clear-cell Pindborg tumor[7] or calcifying epithelial tumor with clear Langerhans cells.[8] In Iran, recently Habibi et al. reported a rare case of clear cell variant of extraosseous CEOT, which is a rare case.[9]

CEOT may present as an intraosseous (central) or extraosseous (peripheral) tumor.[10] Intraosseous CEOT is more common than the extraosseous type, accounting for more than 85% of the cases and presenting most commonly at the mandible. The incidence of extraosseous CEOT is reported to be about 6%. It occurs most commonly at the gingiva.[10] Our case reported an intraosseous tumor extending from the mesial aspect of tooth no. 32 to a mesial aspect of tooth no. 47.

Radiographically, the lesion presents as a unilocular radiolucency. Later, as the lesion ages, calcium salts are deposited and it becomes radio-opaque. It simultaneously erodes bone and thus, the lesion is often mixed radiolucent/radio-opaque giving a characteristic "driven snow" appearance on the radiograph located on the crown of the impacted teeth. The lesion may be unilocular or more commonly, multilocular in appearance. The present case revealed a well-defined mixed lesion in the right area of the mandible, showing that it has been a while that the patient is involved with the lesion without being aware of its existence.

About 60% of cases often involve one or more impacted teeth.[6] Kaplan et al. reported 41 cases of impacted teeth (60%) associated with a total of 67 cases of CEOT.[11] Out of these; the most prevalent were the molars (62%) followed by premolars, canines, incisors and the least were the supernumerary or unidentified teeth (4%). Sharma et al. reported a case in which the radiolucency was pericoronal and unilocular with a thin sclerotic border, containing a supernumerary tooth with resemblance to a dentigerous cyst.[12]

In contrast to Kaplan et al.'s research,[11] some studies have found this feature in only 12% of published cases with adequate radiographic documentation. In our case, no impacted tooth were associated with the tumor, which is similar to recent studies. [4]

In most reported cases the age range of CEOT was between 20-60 years of age with mean around 40 years. In 113 cases reviewed by Franklin and Pindborg, the mean age of patients was at 40 years.[13] Cicconetti et al. reported that tumor more frequently affects adults in the age range of 40-60 years with peak incidence in the fifth decade.[14] The present report speaks about a case in a 16 years old patient which is not a normal age range for occurrence of CEOT. Similarly, Sharma et al. reported the tumor in a 13-year-old boy.[12]

In general, very few cases of CEOT were reported in teenagers. When encountering a teenager who has shown histopathologic features of CEOT, the pathologist must be careful not to misdiagnose CEOT and dental follicle (DF) showing CEOT-like areas (DF-CEOT). Odontogenic tumors may develop from odontogenic epithelial remnants, including those present in DFs, and CEOT-like areas can be found within DFs.[15]

Although, the CEOT tumor in the maxilla may cause pain, nasal obstruction, epistaxis, headache, and proptosis, it typically presents as a painless, slow-growing, asymptomatic mass.[15] Our patient gave no history of any sign and symptoms before the lesion extends in the body of the mandible. In fact, the patient reported a clinical evolution of only 2 months of the lesion. Hence, supporting the literature, the present study reports the case of a painless tumor, located centrally in the right posterior region of the mandible. However, some of the characteristics found in this case are uncommon when compared with the literature, such as the patient's age, which was below the age of 20 and a relatively rapid evolution in the mandibular region.

The diagnosis of CEOT is also based on histopathological examination revealing Islands, strands or sheets of polyhedral epithelial cells, which have eosinophilic finely granular cytoplasm with sometimes considerable nuclear pleomorphism and giant nuclei. However, this view is not an evident for malignancy in this tumor. Moreover, prominent intercellular bridge may be noted. An extracellular eosinophilic homogenous amyloid-like material is characteristic of this tumor with concentric calcific deposits called Liesegang ring.[4] The case we described revealed all of the information above. For precise diagnosis, we stained the specimen with Congo red and the amyloid aggregations, exhibited apple-green birefringence when viewed with polarized light. (Figure 4 and 5).

There are three variants reported in the English literature, the non-calcifying CEOT with Langerhans cells, the CEOT displaying cementum-like and bone-like material, and the clear-cell CEOT.[16] The former variant of CEOT is devoid of calcifications, and this variant's clinical behavior may be less aggressive than the peripheral lesion.[16] It has been proposed that CEOTs with more amyloid and calcifications could behave less aggressively.[17] CEOTs with large amounts of bone-like or cementum-like material probably indicate a higher level of differentiation and this may be a proof for their more self-limiting behavior.[18] On the contrary, the clear-cell CEOT variant is more aggressive with a higher recurrence rate (22%), so must be treated more aggressively.[19]

Basu et al. reported a malignant CEOT with evidence of local tissue invasion and regional lymph node metastasis.[5] Moreover, Demian et al. reported a malignant case of CEOT.[20] According to their study, the aggressive nature of this tumor is related to its poor histologic differentiation, the presence of necrosis, and a high proliferation rate assessed by Ki-67 labeling index.[20] When comparing the histopathological manifestations of our case with the mentioned variants; it seems that the type of histopathology presentations is in accordance with clinical behavior of the tumor. In other words, the more amyloid and calcifications produced, the less aggression is evident in tumor's behavior.

Table 1 provides brief characteristic of 10 most exciting case reports of CEOT in 2012-2014.[5,7,8,13,17,21-26] As shown in table 1, all cases were under the age of 20 and most of them revealed obvious swellings in the alveolar bone. The present case was different from the literature in these two aspects.

Table 1. Characteristics of 10 most exciting case reports of CEOT in 2012-2014

Characteristics		n
Age (year)	< 20	0
	> 20	10
Location	Maxilla	4
	Mandible	6
Clinical manifestations	With swelling	9
	Without swelling	1
Histopathologic variant	Langerhans cell	1
	Clear cell	3
	Cement and bone formation	6
Presence of impacted tooth	Positive	4
	Negative	6
Central or peripheral	Central	9
	Peripheral	1

CEOT: Calcifying epithelial odontogenic tumor

Since CEOT typically has no or minimal signs and symptoms, the tumor may extent large in size before being diagnosed. As long as many odontogenic tumors and dental lesions may have this characteristic, it is recommended that clinician emphasizes on regular dental follow-up and routine radiography to his patients.

Conflict of Interests

Authors have no conflict of interest.

Acknowledgments

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