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An Aggressive Calcifying Epithelial Odontogenic Tumor: A Rare Case Report

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Authors' contributions

This work was carried out in collaboration among all authors. Authors SS and AP designed the study, performed the statistical analysis, wrote the protocol and wrote the first draft of the manuscript. Authors PR and AS managed the analyses of the study. Author AJ managed the literature searches. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Calcifying epithelial odontogenic tumor is an epithelial origin locally aggressive benign odontogenic tumor. It is an extremely rare neoplasm comprise of <1 % of all odontogenic tumors. It manifests clinically as asymptomatic, slow-growing, and locally aggressive lesion which causes expansion of the affected bone. About 400 cases of CEOT (calcifying epithelial odontogenic tumor) are reported in the literature. This article reported a case 18 years old female with a locally aggressive calcifying epithelial odontogenic tumor involving the mandible, without much disfigurement of the face.
Aim: The aim of this article to put a drop of water in the ocean of literature. Report a case with aggressive nature.

Keywords: Calcifying epithelial odontogenic tumor.

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1. INTRODUCTION

Odontogenic tumors represent a spectrum of lesions ranging from malignant and benign neoplasms to dental hamartoma. The odontogenic tumors are unique to jaw because it is arising from odontogenic residues either odontogenic epithelium, ectomesenchyme or from both components [1]. WHO classified Odontogenic Tumors based on which component involved in the development of a tumor. They classify into three categories epithelial origin, mesenchymal origin, and mixed origin [2]. The calcifying epithelial odontogenic tumor is an epithelial origin locally aggressive benign odontogenic tumor. It is also commonly known by the abbreviation CEOT. It is an extremely rare neoplasm comprise of <1 % of all odontogenic tumors [3]. It is presented in two forms central and peripheral, central (intraosseous) form is more common than peripheral (extraosseous) form comprises about 95% and 5% respectively [4]. It manifests clinically as asymptomatic, slow-growing, and locally aggressive lesion which causes expansion of the affected bone. The posterior mandible is the most common site for its occurrence and approximately 50% of cases are associated with erupted or unerupted tooth [5]. It is commonly seen in the 4th to 6th decade of life with no gender predilection [6]. Heinz described CEOT in a German Dental journal more than 2 decades earlier than a first English language paper on CEOT published by Pindborg in 1958. The well-known text book of Oral pathology by Shafers first include the term Pindborg tumor for CEOT in its second edition published in 1963 [7]. Till now 400 cases of CEOT are reported in the literature [8]. This article reported a case 18 years old female with a locally aggressive calcifying epithelial odontogenic tumor involving the right premolar region of mandible and crossing midline involving left mandible also, without much disfigurement of the face. The lesion was locally aggressive shows cortical destruction and expansion and displacement of adjacent teeth. The aim of this article to put a drop of water in the ocean of literature.

2. CASE REPORT

18 years old female reported in the Department of Oral Medicine and Radiology (OMR) with the chief complaint of pain in mandibular front teeth for 2-3 months and an intraoral swelling in the mandibular anterior region which was present for more than 1 year. As the swelling was asymptomatic and not cause much disfigurement of the face, the patient did not consult to the clinician. With due time swelling size increases and associated with intermittent pain. She consulted a clinician in her hometown, where some investigation was advised. As per the given history, that spontaneous exfoliation of a tooth from the mandibular anterior region preceded the appearance of swelling which gradually increased in size with the asymptomatic course. Her past medical and family history was noncontributory. On examination her face was symmetrical, no well-marked extraoral swelling was seen (Fig. 1). Intraorally a well-defined expansile swelling seen in the mandibular anterior region extended from 33 to 44 region, lesion entirely positioned lingual to incisors which were firm, tender, and bleed on palpation. The buccal expansion also was seen adjacent to 43 and 44 which was nontender, and bony hard in consistency (Fig. 2). The provisional diagnosis was made central giant cell granuloma based on clinical presentation. OPG(Orthopantomography) revealed a multilocular mixed radiolucent and radiopaque lesion involving mandible, extending from 34 to 46 region. A well corticated scalloped margin causing thinning of the lower border of the mandible. An eccentrical presence of radio-opaque mass near the lower border of the mandible in a right Para symphyseal region seen suggestive of possible calcification (Fig. 3). The radiographical deferential diagnosis was made CEOT, Dentigerous cyst associated with odontoma, Calcifying Odontogenic Cyst, Odontogenic myxoma, and Central ossifying fibroma. Aspiration was done on the same day, which shows a dry tap which excludes the cystic lesions from the differential diagnosis. An incisional biopsy was planned for the next appointment. On her first visit to the OMR, she did not bring previous reports that brought on the further visit. CT Scan and CBCT (Cone-beam computed tomography)were done in a previous hospital. CBCT shows the involvement of the inferior cortex of the mandible in the region of 44, 45 (Fig. 4e). The lesion shows areas of irregular hyperdensity in the region of 43, 44 (Fig. 4a). There is a displacement of roots 46 buccally and slightly superiorly, displacement of the roots of teeth 43, 44, and 45 (Fig. 4 a-c). No evidence of root resorption, there are areas of buccal and lingual cortical destruction seen (Fig. 4 a-d). CT shows the lesion has caused the expansion and thinning of both buccal and lingual cortical plates, with areas of decortication in the right posterior mandible and areas of expansion and thinning of the lingual cortex in regions of 41, 31, 32, 33, 34.

The incisional biopsy was performed under local anesthesia. Microscopic examination shows connective tissue stroma consisting of sheets and islands of odontogenic cells that have fine borders with granular cytoplasm and intercellular bridges. The cells show giant nuclei with pleomorphism. Deeper areas also show calcifications and eosinophilic globular masses (Fig. 5) The overall histopathological features are suggestive of Calcifying Epithelial Odontogenic Tumor. Conservative excision of the lesion with preservation of inferior border of mandible was done. The excised tissue sends for histopathological evaluation which again confirms CEOT in an excisional biopsy report. The patient put on a follow up quarterly basis. There is no sign of recurrences and complications up to her recent visit (>1.5 years of follow up).

Fig. 1. Extraoral picture of the patient showing a symmetrical face without well-marked deformity

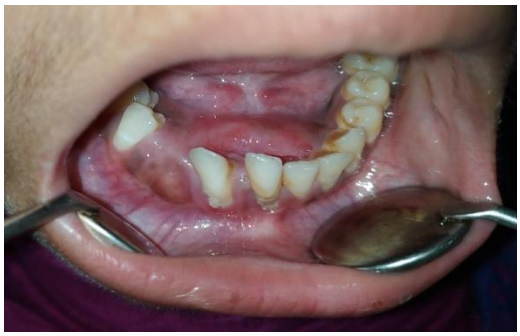


Fig. 2. Intraoral picture shows clinical lesion entirely present in the anterior region of the mandible with overlying mucosa red and edematous in the lingual aspect of the lesion

3. DISCUSSION

The calcifying epithelial odontogenic tumor is classified as an uncommon, benign, odontogenic neoplasm that exclusively epithelial in origin, the etiology of which remain enigmatic. The source of epithelial cells initially suggested by Pindborg was reduced enamel epithelium but today, most investigators believe the cells of origin are stratum intermedium [1]. To explain the pathogenesis Peacock et al conducted a study on seven cases of CEOT. They conclude that the sonic hedgehog pathway (SHH) to be involved in the development of CEOT and they also noticed mutation in PTCH1 [9]. PTCH1 is a tumor suppressor gene within the Sonic hedgehog pathway. The sonic hedgehog pathway regulates the development of multiple organ systems, including odontogenesis by controlling cell to cell interaction and cell proliferation in tissue. PTCH1 mutation first detected in nevoid basal cell carcinoma syndrome [9]and has also been found in both CEOT and keratocystic odontogenic tumors [10]. However, the clinical significance of these mutations is unknown [10].CEOT affected a wide range of ages, most commonly occurs between 20 and 60 years of age, with a peak incidence in the 5th decade [10] and equal distribution between both gender [5]. However, some cases reported before 20 years of age [4,10], present case also reported CEOT affected an 18-year-old female. It commonly presented as asymptomatic, slow-growing, and locally aggressive lesion [5]. Initial clinical signs/symptoms of the lesion are a local expansion of bone with the migration of the teeth of the affected region. Later the overlying mucosa becomes so inflamed that even a slight trauma can lead to bleeding from that side [4]. The present case reported as long-standing expansile swelling in the anterior mandible with dull, intermittent pain and inflamed overlying mucosa with a bleeding tendency. The premolar and molar region of the mandible is the most prevalent site for its occurrence. Although the case also reported in maxilla but less frequent as compared to mandible [10]. The further central intraosseous form is the most common, larger, and more invasive [11] and about 50% of cases show signs of cortical plate perforation [12]. A study conducted by Bruno R. et al according to results the distribution of 247 cases of CEOT among different ethnic groups/races is as follows white-90, Asian-55, Indian -54, blacks -29, Hispanics- 8, Persian/Iranian -6, and 5 in Turkish [12].

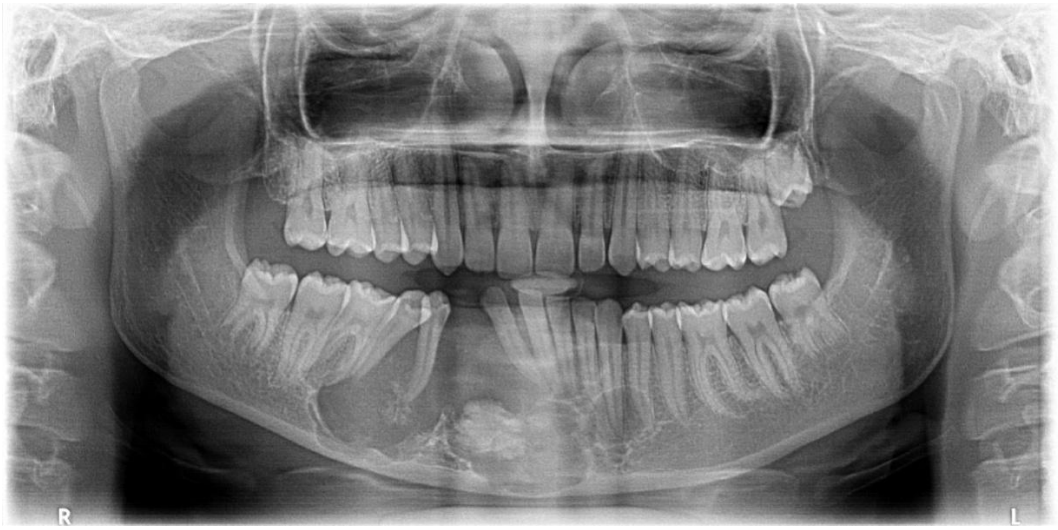


Fig. 3. Orthopantomogram showing a multilocular radiolucent lesion with well-defined scalloped margins and soap bubble lesion in relation to32, 33, and 34; A radiopaque mass present near the lower border of the lesion and another focus of calcification seen apical to 44

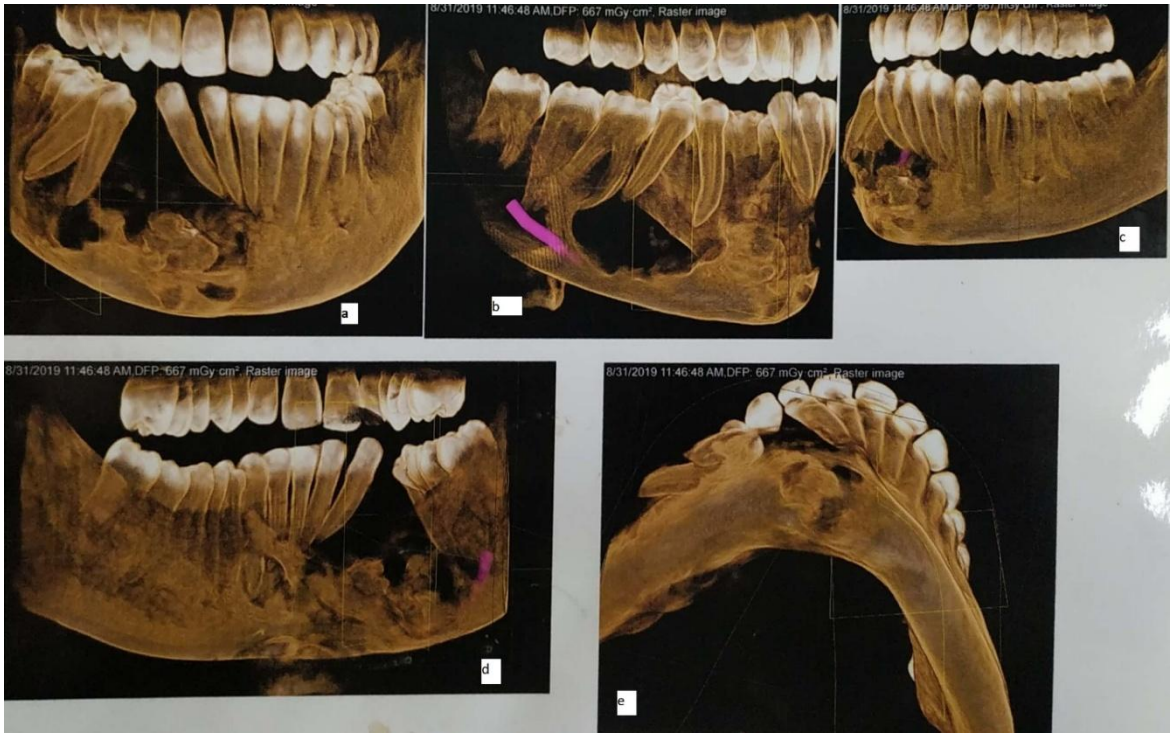
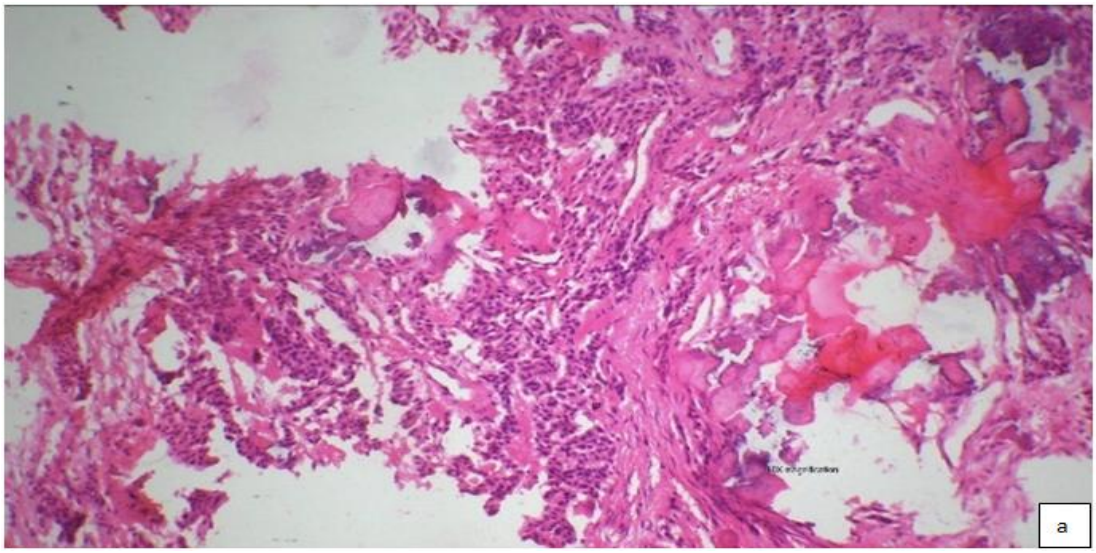


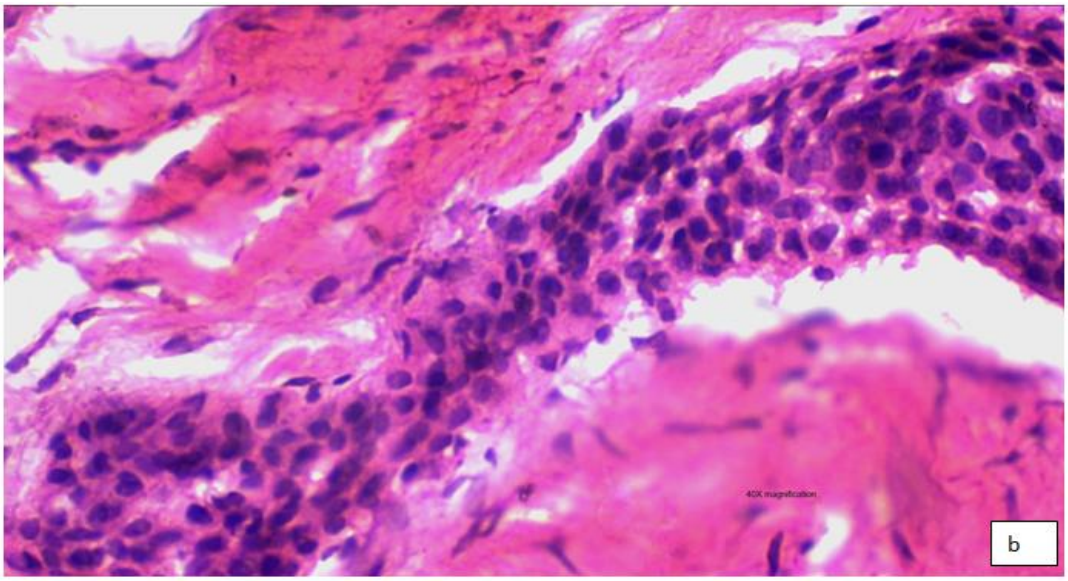
Fig. 4. (a-e): Cone Beam Computed Tomography reveals the involvement of the buccal, lingual and inferior cortex of the mandible and displacement of the teeth in various directions

Radiographically its appearance depends on the different stages of tumor [2,6]. Initially, it is presented as a completely radiolucent lesion with time and progression of the lesion the foci of calcification develops with time, give it mixed and total radiopaque [2,6,10] appearances. The Radiolucent lesion which comprises about 32% of all cases, presented as unilocular or multilocular with a well-defined border or soap bubble/honeycomb appearance in large lesion [6,10]. The foci of radiopaque mass/masses seen in radiolucency which leads to mixed radiopaque radiolucent appearance [10]. This commonly encountered and comprise 65% of all cases [6].

The total radiopaque lesion is seen as snow driven appearance, seen only in 3% of cases [6]. Cortical expansion and tooth displacement are also common findings [3,12]. Only a 13% case shows the resorption of tooth roots [12]. The present case also shows a multilocular mixed radiopaque radiolucent lesion with a well-defined border and soap bubble appearance seen near the left border of the lesion with the displacement of teeth (Fig 3). Although the ordinary radiograph gives the nonspecific finding which may mimic the ameloblastoma, odontogenic myxoma, or other odontogenic cyst or tumors [4,10]. We have also several deferential diagnoses like Central Ossifying fibroma, Odontogenic myxoma, and Calcifying odontogenic cyst (dentinogenic ghost cell tumor). The radiographic diagnosis should be supported by a CT scan (Computed tomography) [4]. In the CT Scan, the expansion of buccal and lingual cortical plates, its thinning, and discontinuation due to breakage is seen clearly [4].CBCT scans may also help in evaluating the true lesion size, the pattern of growth, the presence of calcifications, and the relationship to adjacent structures. It provides a three-dimensional assessment of lesion with higher resolution in relatively low radiation dose [11].

In the present case, CBCT shows clear involvement of the inferior cortex of mandible and displacement of teeth in various directions and CT shows decortication in the right mandible posteriorly.





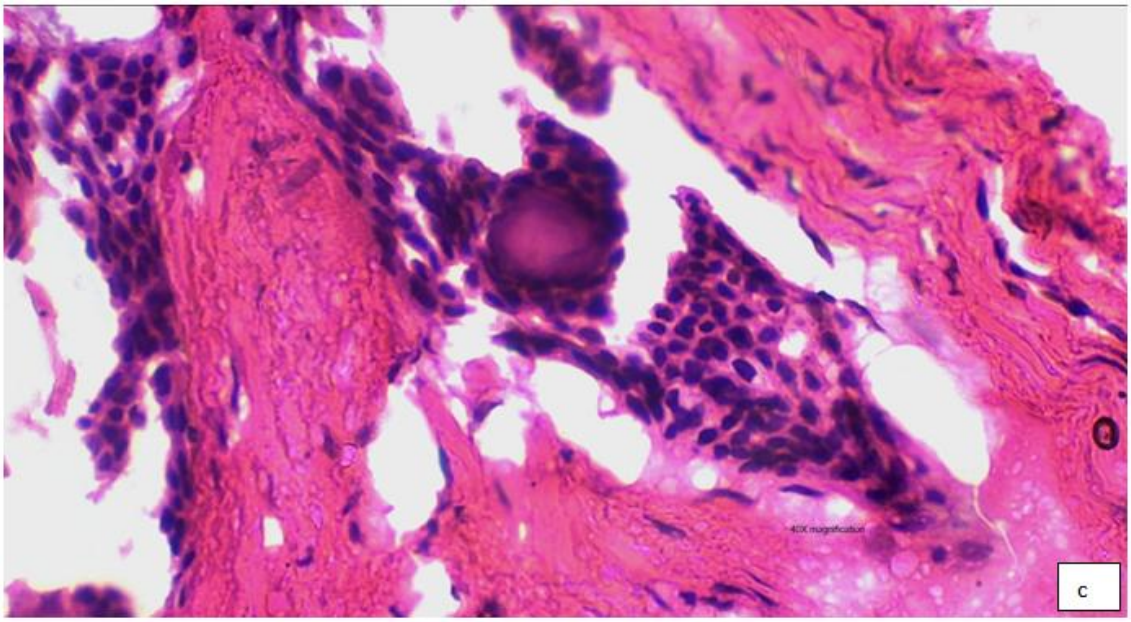


Fig. 5. (a-c): Histopathological slide shows connective tissue stroma consisting of sheets and islands of odontogenic cells, amyloid like material, The cells show giant nuclei with pleomorphism, fine cell out line and intercellular bridge in high power, some areas of calcification surrounded by odontogenic cells also seen

Histological features of the CEOT, represent sheets, cord or nest of the polyhedral cell with eosinophilic cytoplasm, pleomorphic nucleoli, and some degree of mitosis. The most characteristic feature is prominent intercellular bridges. The other commonly seen features are deposition of amyloid-like material and Liesegang ring which are the concentric calcification in amyloid-like material [13]. There is different thought about the origin of amyloid-like material, one thought is, it is derived after degradation of laminadensa while another thought suggests that it is a product of enamel organ protein. This material shows green birefringence under a polarized microscope when stained with congo red stain [10]. There are six histological variants of CEOT reported in the literature in which clear cell variant thought to be the most aggressive one [8]. To date, about 9 cases are reported which was malignant CEOT, in doubtful cases that show any variations from classical histological features IHC should be done [14].

Treatment of the CEOT depends on the site and the size of the lesion. It varies from conservative Surgical management to en-bloc resection. Most of the authors recommend surgical resection along with a 1cm periphery of healthy bone. However, the range of recurrence varies from 15-20% [6].

In the present case, curettage was performed for the preservation of the inferior cortex of mandible. Although the recurrence rate is higher with curettage [12], thereis no sign of recurrence in the present case after 20 months of follow-ups.

4. CONCLUSION

CEOT is a very rare lesion, it first describes more than 6 decades ago, after that around 400 cases are reported in the literature. Its clinical and radiological findings are not so specific and various odontogenic and nonodontogenic lesions come into the differential diagnosis. The present case also radiographically shows scalloped corticated border, calcification and displacement of teeth in various direction, which suggested many differential diagnoses. It also shows cortical perforation that is a sign of its aggressive nature. For effective treatment, correct diagnosis is a must. For making the correct diagnosis a meticulous assessment of clinical, radiological, and histological findings is necessary.

CONSENT AND ETHICAL APPROVAL

As per international standard or university standard guideline participant consent and ethical approval has been collected and preserved by the authors.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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