

# Pindborg tumor of maxilla: Unusual presentation with review

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## ABSTRACT

The calcifying epithelial odontogenic tumor (CEOT), also known as the Pindborg tumor, is a rare benign odontogenic neoplasm, accounts for 0.4-3% of all odontogenic tumors. The CEOT is a benign, locally invasive, slow growing neoplasm occurring as intraosseous and extraosseous forms. The extraosseous variant is diagnosed slightly earlier (mean age 34.4 years) than the intraosseous type (mean age 38.9 years), with equal gender predilection. The most frequent location is the mandibular premolar and molar area; less frequently the lesion is found in the maxilla. This case report is a peculiar case of Pindborg tumor characterized by a rare localization of the lesion extending from anterior to the posterior of the maxilla in a 76-year-old female.

**Key words:** Calcifying epithelial odontogenic tumor, female, intraosseous, maxilla, pindborg tumor

## INTRODUCTION

Calcifying epithelial odontogenic tumor (CEOT) is a rare benign odontogenic epithelial neoplasm, forming 0.4-3.0% of all intraosseous tumours. CEOT was first described by Dutch pathologist Dr. Jens Jorgen Pindborg in 1955.<sup>[1]</sup> The origin of this neoplasm is uncertain, although it is generally accepted to be derived from oral epithelium, reduced enamel epithelium, stratum intermedium or dental lamina remnants.<sup>[2]</sup> The tumor can be divided into two clinical variants: Intraosseous (central) and extraosseous (peripheral) with incidence of 94% and 6%, respectively.<sup>[3]</sup> The intraosseous tumors, when present have a predilection for the mandible more than maxilla (3:1), in the premolar and molar region. Extraosseous tumors are located in the anterior region of the jaws and involve the gingiva. The most common clinical manifestation of CEOT is a localized swelling of the involved jaw. The differential diagnosis includes adenomatoid odontogenic tumor, calcifying odontogenic cyst, dentigerous cyst, ameloblastic fibro-odontoma and odontoma.<sup>[4]</sup>

Here, in this article, a rare case report of intraosseous CEOT in an elderly female patient on the right side of the maxilla is presented with emphasis on the clinical and imaging features and its management.

## CASE REPORT

A 76-year-old female patient presented with a painless swelling over the right cheek region since 1 year. History of presenting illness revealed that swelling was insidious in onset, initially small and now increased to attain the present size. There was no history of trauma, fever or similar swelling elsewhere in the body. Swelling was progressively increasing in size and was painful from the last 15 to 20 days. There was no history of discharge. Patient's medical history was relevant for hypertension since 10 years. Patient's dental history was significant for extraction of 16, 17 and 18, 5-years back due to loosening of teeth. All the vitals were within the normal limits. Extra-oral examination revealed a solitary, large diffuse swelling on the right side of the cheek region, causing facial asymmetry. The overlying skin was intact and normal in color. The swelling extended from nasolabial fold to 2 cm in front of the right ear in the horizontal direction and from the zygomatic process of maxilla to 2 cm above the inferior border of mandible in the vertical direction on the right side, approximately measuring 6 cm × 5 cm in size. On palpation, there was no localized increased in temperature, overlying skin was pinchable and the swelling was non-tender. Regional lymph nodes were not palpable.

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10.4103/2278-0513.125822

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On Intraoral examination, a solitary well defined swelling measuring 5 cm × 4 cm × 1.5 cm extending from the distal surface of right maxillary second premolar to tuberosity area in the antero-posterior direction and from the midline of the hard palate to right maxillary buccal vestibule in mediolateral direction. The overlying mucosa was normal and intact except in the center of the swelling, which was ulcerated and covered with necrotic slough due to trauma from mandibular posterior teeth. On palpation, the swelling was firm in consistency mildly tender near the center.

On the radiographic examination, the intra-oral periapical and maxillary occlusal radiograph showed bone loss and diffuse soft-tissue shadow distal to 15. Panoramic view showed radiolucent lesion involving the right alveolus with erosion of underlying bone. Computed tomography (CT) scan showed large, oval shaped expansile, osteolytic lesion involving right alveolus. The lesion extended superiorly to the floor of maxillary sinus with erosion [Figure 1].

Aspiration was tried, but it was non-productive. Based on history, clinical and imaging findings a provisional diagnosis of benign locally aggressive odontogenic neoplasm was considered. Patient was referred to the department of oral and maxillofacial surgery, where the incisional biopsy was performed under local anesthesia.

The histopathological sections revealed varying sizes of epithelial islands surrounded by hemorrhagic areas in a fibrous connective tissue stroma. Some areas of circular amorphous eosinophilic hyalinized amyloid-like material were seen [Figure 2]. Few cells with pleomorphism and mild degree of variability in nuclear size were seen. These findings were consistent with the diagnosis of CEOT. Being aggressive in nature, a hemimaxillectomy using Le Fort I surgical approach was performed. The excised tissue was

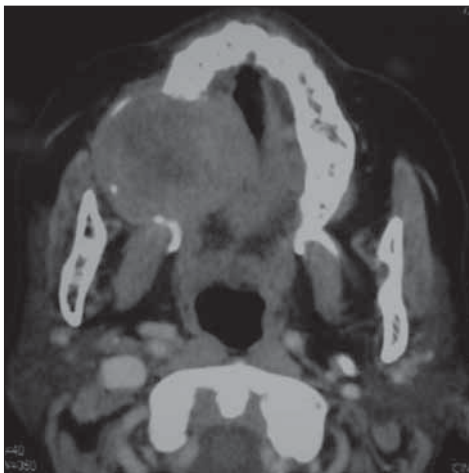
further histopathologically diagnosed as CEOT with free margins. Patient was followed-up for 3 years post-surgically. There was no evidence of recurrence during this period.

## DISCUSSION

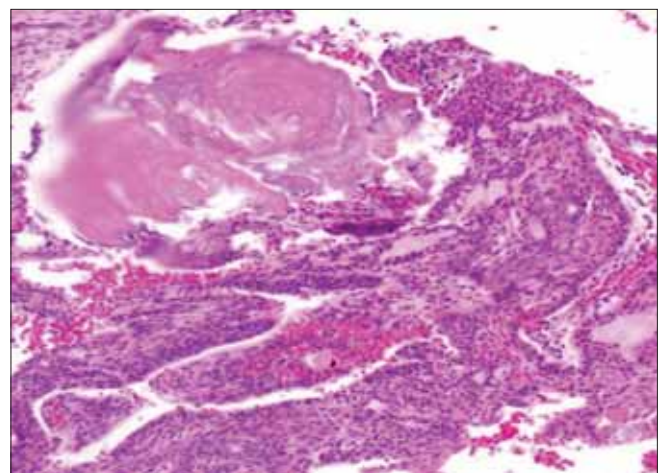
The CEOT is a benign, rare odontogenic and locally invasive tumor with less than 1% of all odontogenic neoplasm. It is most commonly reported in patients during the 4<sup>th</sup>, 5<sup>th</sup> and 6<sup>th</sup> decades of life, more common in whites, without any sex predilection.<sup>[4]</sup> CEOT has been reported under a variety of different names such as “adamantoblastoma,” “unusual ameloblastoma,” and “cystic complex odontoma.” or tumor.<sup>[5]</sup>

The origin of this tumor is uncertain. Some authors speculate that it originates from the remnants of dental lamina based on its anatomic distribution in the jaws and/or basal cells of gingival surface epithelium.<sup>[5]</sup> Others suggest that it bears structural resemblance to the cells of stratum intermedium of the enamel organ along with a high activity of alkaline phosphatase and adenosine triphosphate.<sup>[2]</sup> The extraosseous variant is diagnosed slightly earlier (mean age 34.4 years) than the intraosseous type (mean age 38.9 years).<sup>[5]</sup> Mandible is more commonly affected than the maxilla in the ratio of 3:1. In maxilla, posterior region is the preferred site with 3 times more prevalence in the molar region than in the premolar region.<sup>[4]</sup> It may present as a slow growing, painless, expansile bony hard swelling causing cortical bone to become egg shell thin before perforation and subsequent soft-tissue infiltration. It may cause tooth tipping, migration, rotation and/or mobility secondary to tooth resorption.<sup>[5]</sup> In rare instances of maxillary CEOT, patient may report of pain, nasal stuffiness, headaches, epistaxis or proptosis.<sup>[6]</sup> In the present case, patient did not report of such problem.

CEOT is less aggressive than ameloblastoma although cases of malignant transformation have been reported.<sup>[7]</sup>



**Figure 1:** Computed tomography scan (axial) view showing large expansile, soft tissue lesion with bone destruction



**Figure 2:** Photomicrograph of the histopathological section showing features of calcifying epithelial odontogenic tumor (H and E, ×40)

The aggressiveness is an important finding in the posterior maxilla. In addition, root resorption is reported as a rare finding in CEOT (4%), which is more in solid ameloblastoma (81%).<sup>[8]</sup>

Depending upon stages of development, CEOT may present with variable radiographic appearances.<sup>[4]</sup> The lesion usually consists of a radiolucent area, which is well-defined. Later, as the lesion matures, it becomes increasingly radio-opaque because of calcium salt deposition. It also simultaneously erodes bone and thus, the lesion is often mixed radiolucent/radio-opaque giving a characteristic “driven snow” appearance on the radiograph. Further, the lesion may be unilocular or more commonly, multilocular in appearance.<sup>[8]</sup> Small lesions are usually unilocular and larger lesions tend to be multilocular. The mixed radiolucent and radiopaque pattern occurred most often (65%) followed by the completely radiolucent pattern (32%) and least often the totally radiopaque “wind driven snow” pattern (3%). When tumor is associated with impacted tooth, it may appear as pericoronal radiolucency with or without small radiopacities. Kaplan *et al.* reported 41 cases of one or more impacted teeth (60%) associated with a total of 67 cases of CEOT. The most prevalent impacted teeth were the molars (62%) followed by premolars, canines, incisors and the least were the supernumerary or unidentified teeth (4%).<sup>[8]</sup> Recently, Sharma *et al.* has reported a case of intraosseous CEOT surrounding a supernumerary tooth in the anterior maxilla (an uncommon site) in a pediatric patient.<sup>[9]</sup>

On CT examination, CEOT shows expansion and thinning of buccal and lingual cortical plates. On magnetic resonance imaging, it reveals predominantly a hypo intense lesion on T1-weighted images and mixed hyper intense lesion on T2-weighted images.<sup>[8]</sup>

According to, the characteristic histological criteria for the diagnosis of CEOT are sheets of large polygonal epithelial cells that have well-defined borders and often show prominent intercellular bridges. There is usually pleomorphism of the epithelial cells. Nuclei are often prominent and show considerable variation in size, shape and number. Cellular abnormalities are frequent, whereas mitotic figures are rarely seen. Cytoplasm is abundant and eosinophilic. Varying amounts of an extracellular amyloid-like material that stains positive with Congo red stain and exhibits an apple-green birefringence under polarized light is also typical of these tumors. It also fluoresces under ultraviolet light with thioflavin T. This amyloid-like material may contain either basement membrane components (type IV collagen) or a mixture of cytokeratins. These tumors may be epithelium predominant with minimal amyloid and vice-versa. Still others may

have abundant clear cells. Concentric calcified masses with a Liesegang’s rings calcification pattern are also pathognomonic for this tumor.<sup>[5]</sup>

Early Surgical intervention is treatment of choice, which depends on the size, location of the neoplasm, the patient’s overall medical condition or tolerance to withstand the surgical procedure and the skill or experience level of the operator.

More aggressive surgery is required in cases of CEOT involving maxilla, since these tumors tend to grow more rapidly and are not circumscribed. Small lesions with well-defined borders may be surgically excised by either thorough intrabony curettage or conservative en-bloc resection sacrificing a thin margin of healthy adjacent bone. For neoplasm greater than 4 cm, en bloc resection is mandatory. Extensive involvement requires partial/hemimaxillectomy requiring post-operative reconstruction and rehabilitation with titanium mesh, grafts and flaps may be required to close the defect. Distraction osteogenesis has shown promising results in such defects. The tumor-free margins are confirmed histopathologically and require radical resection of affected jaw portion and any associated soft tissues with no less than 1 cm in every direction.<sup>[6]</sup> Most studies of tumor report a local recurrence rate between 10% and 20% following conservative treatment. The prognosis is considered as favorable.<sup>[5]</sup> One case of advance disease in patient’s maxilla resulted in local extension to the brain.<sup>[10]</sup> The incidence of malignant transformation of tumor is extremely low and metastasis is rare.<sup>[7]</sup>

## CONCLUSION

The lesions in the maxilla are usually aggressive and require extensive surgeries as they grow rapidly when compared to the mandibular cases and invasion into surrounding vital structures also affects the morbidity of these patients. Resection with tumor-free accurate margins is the preferred treatment choice. This case report is presented in view of rare involvement of maxilla by CEOT. This is to emphasize the need to correlate radiographic, clinical and histopathological findings to arrive at the correct diagnosis and treatment planning. The intraosseous variant of CEOT in the maxilla is in itself a rare entity. In the present case, the patient was totally asymptomatic and hence, the correlation of clinical, radiographic and histopathological examination is stressed upon.

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**Cite this article as:** Sachdeva SK. Pindborg tumor of maxilla: Unusual presentation with review. *Clin Cancer Investig J* 2014;3:126-9.

**Source of Support:** Nil, **Conflict of Interest:** None declared.