

Case Report

A unique case of clear cell variant of calcifying epithelial odontogenic tumor involving the maxilla

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ABSTRACT

Calcifying epithelial odontogenic tumor (CEOT) is a rare, benign, odontogenic tumor arising from the odontogenic epithelium and accounts for approximately 1% of all odontogenic tumors. Clear cell variant of CEOT is a distinct entity and has more aggressive biological behavior and higher chances of recurrence. Here, we present a unique case of clear cell variant of CEOT involving the left side of the maxillary alveolus in a 73-year-old female patient with thorough clinical, radiological, and histological details.

Key Words: Amyloid, calcifying epithelial odontogenic tumor, Congo red, odontogenic tumor

INTRODUCTION

Calcifying epithelial odontogenic tumor (CEOT), also known as Pindborg tumor, is a rare, benign, odontogenic neoplasm of the jaws, which comprises <1% of all odontogenic tumors and was introduced in scientific literature almost 50 years ago by Dr. J. J. Pindborg.^[1-3] The origin of the neoplasm is controversial, though it is believed to be derived from the oral epithelium, reduced enamel epithelium, stratum intermedium, or remnants of dental lamina.^[4,5]

In general, two varieties of CEOT are recognized – extraosseous and intraosseous with an incidence of 6% and 94%, respectively. Intraosseous tumors commonly involve mandible than maxilla with a ratio of 2:1 in the molar and premolar region.^[1,4] It is most frequently found in patients between 30 and 50 years of age without sex predilection.^[5] Clinically, CEOT is usually presented as asymptomatic, slowly

enlarging swelling associated with expansion of cortical plates. Radiographically, it is characterized as unilocular or multilocular radiolucent lesions that often exhibit a mixed pattern because of areas of scattered flakes of calcifications and produces a typical “driven snow” appearance. Sometimes, the lesion may be associated with an impacted tooth.^[1,3-5]

The histopathology of CEOT, in its classic pattern, comprises sheets of polyhedral neoplastic epithelial cells with well-defined cell borders and distinct intercellular bridges. These neoplastic cells may demonstrate pleomorphism but only rarely typical mitoses. Other characteristic findings are the presence of amyloid-like substances and calcified concentric Liesegang rings. Amyloid-like substances stained positively with Congo red and produced apple-green birefringence under polarized light. Tumor may show variations in the classic histologic

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appearance such as noncalcifying CEOT with Langerhans cells, CEOT displaying cementum and bone-like material, and clear cell CEOT, of which clear cell CEOT shows more aggressiveness with a high recurrence rate (22%).^[1,6] Yamaguchi *et al.* concluded that the clear cells represent a feature of cytodifferentiation rather than a simple degenerative phenomenon.^[7]

Treatment options may range from simple enucleation and curettage to radical and extensive resection. The prognosis of CEOT is good, but long-term follow-up is mandatory.^[1]

Based on the above-mentioned clinicopathological, radiological, and histopathological features, a case of clear cell variant of CEOT involving the left maxilla was diagnosed and has been discussed herewith.

CASE REPORT

A 73-year-old female patient reported to the Department of Oral and Maxillofacial Pathology of Guru Nanak Institute of Dental Sciences and Research, Panihati, Kolkata, India, with a chief complaint of swelling involving the left side of the face for the past 8–9 months. The patient gave a history of extraction of her maxillary left first, second, and third molar teeth 8–9 months back and followed by the development of a swelling, involving the molar region, which had gradually increased in size accompanied by localized intermittent pain.

Extraorally, there was a diffuse swelling involving the left middle third of her face. Overlying skin appeared normal without any regional lymphadenopathy.

Intraoral examination revealed the presence of a diffuse, large, moderately tender, noncompressible, firm to hard swelling involving the partially edentulous maxillary left molar region associated with marked expansion of buccal and palatal cortical plates. Orthopantomogram and paranasal air sinus view revealed the presence of a large lesion having a mixed radiographic appearance characterized by the presence of radiolucencies interspersed with multiple areas of radiopacities produces a typical “driven snow” type of appearance, without encroaching the left maxillary antrum [Figure 1]. Routine hematological investigations were within the normal limits. Based on the above clinical and radiological findings, the provisional diagnosis of fibro-osseous lesions and odontogenic neoplasms was made.

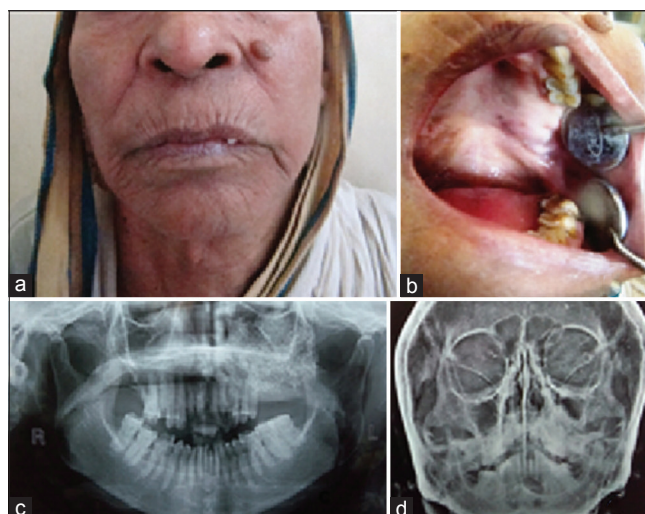


Figure 1: (a) Extraoral clinical photograph of the patient showing a diffuse swelling involving the left middle third of face, (b) intraoral photograph showing the presence of a large, diffuse swelling involving the partially edentulous maxillary left molar region with marked expansion of palatal cortical plate, (c and d) panoramic orthopantomogram and paranasal air sinus view revealed the presence of “driven snow” type of radiological appearance without involvement of the left maxillary antrum.

Ethical clearance to this study was received. Following this, incisional biopsy was performed from the representative site of the lesion after obtaining informed consent from the patient. The light microscopic features revealed the presence of sheets of polyhedral, neoplastic, odontogenic epithelial cells with prominent cellular outlines and intercellular bridges along with the presence of homogeneous, eosinophilic, amorphous materials. Calcified areas were also noted within the sheets of epithelial cells [Figure 2]. One of the most interesting features was the presence of clear cells having vacuolated cytoplasm within the odontogenic epithelial islands which stains positively with periodic acid–Schiff (PAS). Our provisional diagnosis was clear cell variant of odontogenic tumor, and to confirm this diagnosis, we had gone for immunohistochemical (IHC) evaluation. IHC markers such as cytokeratin 8 confirmed the presence of odontogenic epithelium within the neoplasm. Congo red-stained, amyloid-like material produces a typical apple-green birefringence viewed under a confocal microscope with polarized light [Figure 3].

After considering the microscopic features, the diagnosis of clear cell variant of CEOT was made, and the patient was referred for surgical treatment and management. However, she had refused to undergo

any surgical treatment because of her old age. Hence, any follow-up treatment could not be possible.

DISCUSSION

CEOT is rare, benign, but locally aggressive neoplasm of epithelial origin that accounts for <1% of all odontogenic tumors and characteristically contains calcifying masses and homogeneous, eosinophilic hyaline-like material within the tumor epithelium as well as connective tissue stroma. It was first introduced into scientific literature almost 50 years ago

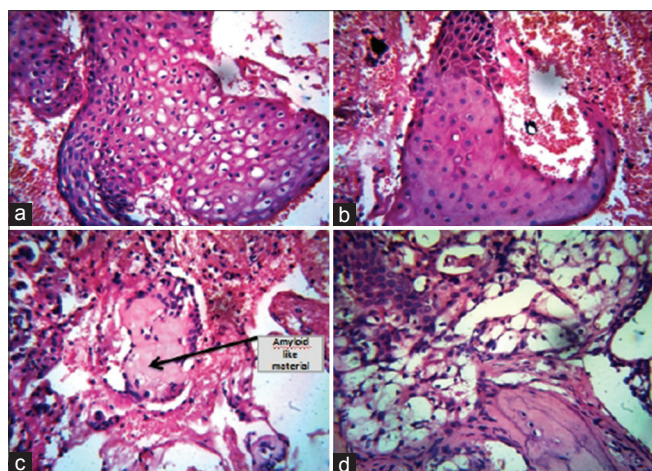


Figure 2: (a and b) High-power photomicrographs showing the presence of sheets of polyhedral neoplastic odontogenic epithelial cells with distinct intercellular bridges along with the presence of clear cells with foamy cytoplasm (H and E, $\times 40$), (c and d) high-power photomicrographs showing the presence of amyloid-like material within the connective tissue stroma and areas of calcification (H and E, $\times 40$).

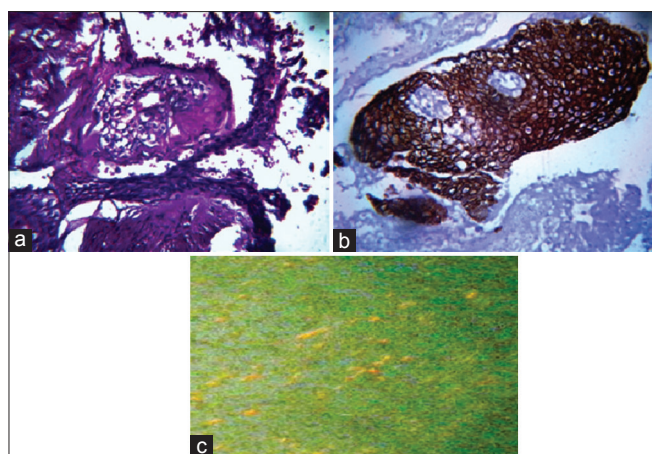


Figure 3: (a and b) High- and low-power photomicrographs ($\times 40$) showing periodic acid-Schiff positive clear cells and cytokeratin positive odontogenic epithelial cells, (c) apple-green birefringence of amyloid-like materials (Congo red stain $\times 10$).

by Dr. J. J. Pinborg.^[1,2] The clear cell variant of CEOT is a distinct entity and has more aggressive biological behavior and higher chances of recurrence (22%).^[1,6] Histogenesis of this neoplasm is uncertain, mostly believed to arise from stratum intermedium of dental lamina because of the morphological resemblance of tumor cells to that stratum intermedium and a high activity of alkaline phosphatase and adenosine triphosphate in both these cells.^[4,5,8-10]

A painless, slow-growing swelling is the most common presenting sign according to the authors of various studies.^[1-5,7-11] The patient under discussion was a 73-year-old female, having moderately tender, noncompressible swelling involving partially edentulous maxillary left molar region associated with marked expansion of cortical plates. The site of involvement and the age of occurrence of CEOT in our case are in accordance with the cases published in the previous reports [Table 1].^[1,11-15] Of them, only two cases^[1,13] were histologically proved to be clear cell variant of CEOT.

Radiographically, according to various studies,^[1-5,11-14] CEOT exhibits a unilocular or multilocular radiolucencies with radiopaque flakes of calcifications within the radiolucent area producing a typical “driven snow” type of appearance. Our case also revealed similar radiological features but was not associated with an impacted tooth.

Histological sections of CEOT showed islands of polyhedral epithelial cells, having nuclei of varying form and size along with the extracellular, eosinophilic, amyloid-like material and areas of concentric calcifications in the form of Liesegang rings within the connective tissue stroma. The clear cell variant of CEOT is very rare and consists of clear cells with foamy cytoplasm which stains positively with “PAS” stain because of the presence of glycogen.^[1,2,10] Amyloid-like material showed positive, apple-green birefringence under polarized light. Odontogenic epithelial cells stain

Table 1: Age and sex distribution of calcifying epithelial odontogenic tumor involving maxilla as described by different authors for 6 years

Authors	Age/sex	Year of study
Sedghizadeh <i>et al.</i> ^[15]	51/male	2007
Nascimento <i>et al.</i> ^[14]	35/male	2009
Angadi and Rekha ^[11]	30/male	2011
Sahni <i>et al.</i> ^[1]	52/male	2012
Kamath and Abraham ^[12]	35/male	2012
Badrashetty <i>et al.</i> ^[13]	36/female	2013

positively with cytokeratin 8 (Type II).^[1,6] The nature of arrangements of tumor tissue and staining characteristics of cells in the present case was strongly mimicking the features of clear cell variant of CEOT though Liesegang rings were absent. Histopathologically, clear cell variant of CEOT contains amyloid-like material which showed apple-green birefringence after staining with Congo red under polarized light. These features help differentiate it from clear cell odontogenic carcinoma, central mucoepidermoid carcinoma, central acinic cell carcinoma, and metastatic renal cell carcinoma.

The method of treatment will depend on multiple factors such as size, location of the tumor, general condition of the patient, histopathological findings, and operator skills. Small intrabony lesions are treated by simple enucleation and curettage whereas large tumors require aggressive approach, either by hemimandibulectomy or hemimaxillectomy. The recurrence rate was reported to be 10%–20%; hence, periodic follow-up is essential to prevent further recurrence.^[8,9]

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Conflicts of interest

The authors of this manuscript declared that they have no conflicts of interest, real or perceived, financial or nonfinancial in this article.

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