CASE REPORT

Non-calcifying variant of calcifying epithelial odontogenic tumor with clear cells–first case report of an extraosseous (Peripheral) presentation

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ABSTRACT

Calcifying epithelial odontogenic tumor or Pindborg tumor is a rare benign odontogenic tumor that was first described by a Danish pathologist Jens Jorgen Pindborg in 1955. It is thought to arise from the epithelial element of the enamel origin and is histologically characterized by the presence of polygonal epithelial cells, amyloid globules, and Liesegang ring calcifications. A few cases have been shown to demonstrate clear cells (Langerhans cells) in the tumor. Non-calcifying epithelial odontogenic tumor (NCEOT) s very rare and only four cases have been documented in the English Language Literature to date. All these cases were present intraosseously (central). We present the first case of an extraosseous (peripheral) NCEOT with clear cells (Langerhans cells) and briefly discuss as well as review the histogenesis along with the differences in its clinicopathological and prognostic profile with respect to other variants.

Key words
Calcifying epithelial odontogenic tumor, clear cells, extraosseous, langerhans cells, non-calcifying, peripheral

INTRODUCTION

Calcifying epithelial odontogenic tumor (CEOT) was documented originally by Thonay Goldman.[1] It was known as Pindborg tumor due to its description in 1955 by Pindborg; and its description in full detail was accomplished in 1958.[1] It is a rare and important tumor for its similarity to a carcinoma, and they cannot be clearly differentiated.[1] Pindborg tumor represents about 1% of the odontogenic tumors.[1] The most common mode of presentation is a slow growing intraosseous mass in the mandible, in the fourth to fifth decade of a person's life. Histopathology is the gold standard for diagnosis of this tumor.[2] Non-calcifying morphology, presence of clear cells, and extraosseous or peripheral location are extremely rare features. We present here the first case of its kind, where all of the above-mentioned rare elements are present, together with a young age of the patient at diagnosis; we also highlight the atypical microscopic features of this tumor.

Access this article online	
Quick Response Code:	Website: www.ejgd.org
	DOI: 10.4103/2278-9626.106825

CASE REPORT

A 20-year-old woman presented with a slow-growing hard mass in the right upper oral cavity since the past 1 year. The mass was progressively increasing in size and it was painless. Local examination revealed a hard nodule measuring 1 cm in diameter, located just above the right lateral incisors, submucosally. It was not attached to the underlying bone. Radiography revealed a non-ossifying soft tissue mass [Figure 1]. There was no abnormality in the maxilla or teeth. The patient's medical history and physical examination were non-contributory. Probing into the mass was done under local anesthesia, but no fluid or any other material could be aspirated.

An excisional biopsy with adequate margin was performed. The histopathological examination revealed an epithelial odontogenic neoplasm composed of scattered small islands of polygonal cells in an abundant fibromyxoid connective tissue stroma. The cells had centrally placed vesicular nuclei with inconspicuous nucleoli and eosinophilic cytoplasm. Amidst the epithelial nests, there were many islands and spherical bodies of amorphous, acellular, eosinophilic amyloid-like material. Few to occasionally several clear cells were also observed in many nests. No areas of calcification were present [Figure 2]. On immunohistochemical evaluation, the epithelial cells showed positivity for cytokeratin (CK

Cocktail, Clone AE1+AE3, Biogenex CA, USA) [Figure 3] and the clear cells showed S-100 positivity (Clone 15E2E2, Biogenex CA, USA), suggesting them to be Langerhans cells [Figure 4].

Based on the histological and immunohistochemical findings, a diagnosis of extraosseous, non-calcifying variant of CEOT harboring Langerhans cells was suggested.

The tumor was completely excised and the patient continues to be on regular follow-up; the tumor has not recurred 6 months after excision.

DISCUSSION

The odontogenic tumors are a complex group of lesions with several clinical behaviors and histological variations. CEOT, also known as Pindborg tumor, is a rare lesion that represents about 1% of the odontogenic tumors.[1] Despite its odontogenic origin, its histogenesis is uncertain. The tumoral cells of the interosseous variant present morphology similar to the cells of stratum intermedium of enamel, whereas the extraosseous type is derived from dental lamina epithelial or the basal cells of gingival epithelium.[3]

Classically, CEOT presents as a painless slow-growing mass in the mandible. The mean age of presentation is 40 years, with equal incidence in men and women.[4] Clinically intraosseous (central) tumors are more common (94%) than extraosseous (peripheral) tumors (6%).[5] The central type of CEOT occurs in individuals in age group of 20-60 years. Two-third of the lesions occur in the jaw, more commonly the molar area, with a tendency to occur in the premolar areas.[5] In the maxilla it can cause proptosis, epistaxis, and nasal airway obstruction. The peripheral type is commonly found in the anterior region of the maxilla and occurs as a soft tissue swelling.[5]

The presentation of the intraosseous and extraosseous types is similar and both have similar histological features. Radiologically, intraosseous CEOT shows radiolucent areas with occasional calcification, whereas the extraosseous type may show bone erosion near the tumor.[6] Intraosseous CEOT is more aggressive, with a reported recurrence rate of 14%.[6]

Histopathologically, CEOT is characterized by the presence of epithelial cells, homogenous eosinophilic amyloid-like material, and calcification. The epithelial cells are arranged in nests and sheets and are polygonal, with clear to eosinophilic cytoplasm and vesicular nuclei having prominent nucleoli. A cribriform and pseudoglandular pattern of epithelial cells is also described. Although moderate pleomorphism can be observed, necrosis and atypical mitosis are uncommon. Rounded, pale, eosinophilic material resembling amyloid is observed interspersed amidst the tumor cells and is characteristic

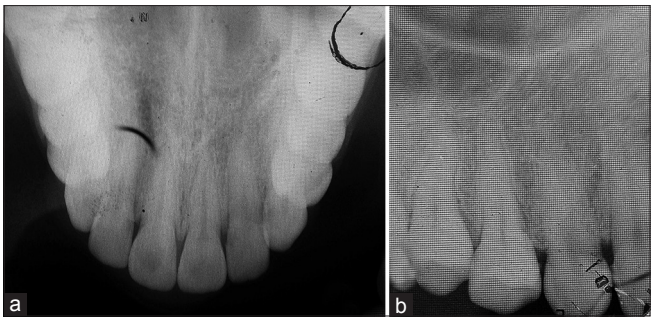

Figure 1: Radiograph of maxilla (front view) showing no bony abnormality or calcification

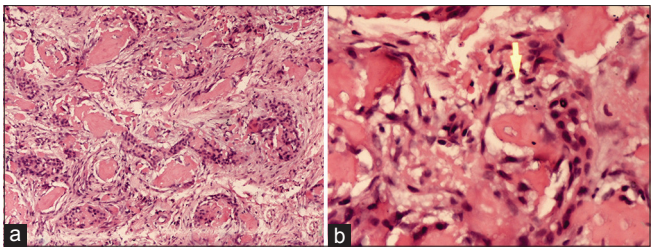

Figure 2: Photomicrograph showing sheets and nests of epithelial cells with eosinophilic amyloid-like material and clear cells (arrow) interspersed between the epithelial cells. Note the absence of calcification (H and E, ×10 and ×40)

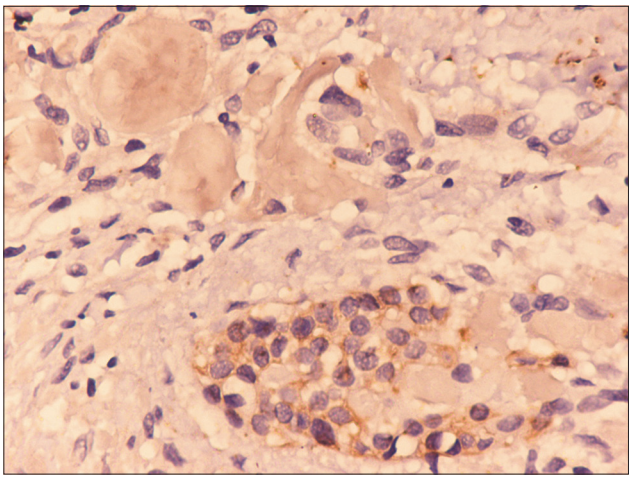

Figure 3: Photomicrograph showing cytoplasmic cytokeratin positivity by epithelial cells. The clear cells are negative (pancytokeratin, ×40)

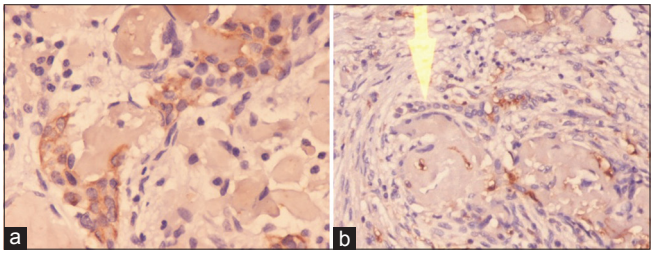

Figure 4: Photomicrograph showing cytoplasmic S-100 positivity by the scattered clear cells. The epithelial nests are negative (arrow) (S100, ×40)

of CEOT. Although the exact origin of this amyloid is not known, it is believed to be derived from filamentous degradation of keratin filaments secreted by tumor epithelial cells. The presence of calcification is another defining feature of Pindborg tumor. The extent and shape of calcification can vary from minimal small, round concretions to Liesegang rings and large aggregates.[2]

According to Krolls and Pindborg, the presence or absence of calcification in CEOT has prognostic implications. A lack of calcification indicates less tumor differentiation and hence has more chance of a recurrence. Pindborg has also reported recurrence after removal in a CEOT that had minimal calcifications.[2]

Total absence of calcification in CEOT has been reported in the English Language Literature in only four cases previously.[2,7-9] All these non-calcifying cases presented as intraosseous tumor; however, our case is the first report of an extraosseous presentation. Two of the previously reported cases did not show any evidence of recurrence 1 year after surgical excision;[2,7] follow-up in the other two cases is not documented.

Few authors have reported clear cells as a minor or major component of the tumor, and these cells were found to express S-100 protein, lysozyme, MT1, LN3, and OKT6 by immunohistochemistry but not keratin.[6] Electron-microscopic examination revealed rod-shaped and racket-shaped structures called Birbeck's granules in the cytoplasm of these clear cells,[10] indicating that these are Langerhans cells. The clear-cell variant of CEOT is known to have aggressive behavior.[6]

There is no consensus regarding the treatment of CEOT. It has been observed that conservative treatment (enucleation and curettage) has a recurrence rate of 14%; however, there has been no report of recurrence after aggressive treatment (resection marginal or segmental). It has been suggested that the follow-up of CEOT should be annual and a long period of postoperative attendance is indicated.[1]

In the present case, the tumor was located peripherally, i.e., submucosally, in the anterior maxilla, and showed diffuse amorphous, non-calcifying eosinophilic deposits, along with interspersed epithelial cells and Langerhans cells. Our patient is on regular follow-up and the tumor has not recurred 6 months after surgical excision.

This case highlights the unusual presentation and microscopic features of this rare tumor. Prompt recognition of this variant can guide surgical management and alert the clinician to the need for extended follow-up.

REFERENCES

1. Araujo VS, Chagas JF, Casonato H Jr, Ribeiro AL, Toledo JI Jr. Calcifying epithelial odontogenic tumor: A case report with 3-year follow up. Int J Dent, 2011;10:195-9.
2. Kaushal S, Mathur SR, Vijay M, Rustagi A. Calcifying epithelial odontogenic tumor (Pindborg tumor) without calcification: A rare entity. J Oral Maxillofac Pathol 2012;16:110-2.
3. Aviel-Ronen S, Liokumovich P, Rahima D, Polak-Charcon S, Goldberg I, Horowitz A. The amyloid deposit in calcifying epithelial odontogenic tumor is immunoreactive for cytokeratins. Arch Pathol Lab Med 2000;124:872-6.
4. el-Labban NG. Cementum-like material in a case of Pindborg tumor. J Oral Pathol Med 1990;19:166-9.
5. Philipsen HP, Reichart PA. Calcifying epithelial odontogenic tumour: Biological profile based on 181 cases from the literature. Oral Oncol 2000;36:17-26.
6. Hicks MJ, Flaitz CM, Wong ME, McDaniel RK, Cagle PT. Clear cell variant of calcifying epithelial odontogenic tumor: Case report and review of the literature. Head Neck 1994;16:272-7.
7. Aufdermaur M. Pindborg tumor. J Cancer Res Clin Oncol 1981;101:227-30.
8. Takata T, Ogawa I, Miyauchi M, Ijuhin N, Nikai H, Fujita M. Non-calcifying Pindborg tumor with Langerhans cells. J Oral Pathol Med 1993;22:378-83.
9. Hafian H, Mauprivez C, Furon V, Pluot M, Lefevre B. Pindborg tumor: A poorly differentiated form without calcification. Rev Stomatol Chir Maxillofac 2004;105:227-30.
10. Asano M, Takahashi T, Kusama K, Iwase T, Hori M, Yamanoi H, *et al*. A variant of calcifying epithelial odontogenic tumor with Langerhans cells. J Oral Pathol Med 1990;19:430-4.

How to cite this article: Afroz N, Jain A, Maheshwari V, Ahmad SS. Non-calcifying variant of calcifying epithelial odontogenic tumor with clear cells-first case report of an extraosseous (Peripheral) presentation. Eur J Gen Dent 2013;2:80-2.

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