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CASE REPORT

Non Calcifying Type of Calcifying Epithelial Odontogenic Tumor: An Unusual Case Report with Special Emphasis on Histogenesis of Calcifications

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Abstract Calcifying epithelial odontogenic tumour also known as Pindborg tumour, is a rare benign odontogenic neoplasm of locally aggressive behavior. It is thought to arise from the epithelial element of the enamel origin which are reminiscent of the cells in the stratum intermedium layer of enamel organ in tooth development. The tumour is characterized histologically by the presence of polygonal epithelial cells, calcifications and eosinophilic deposits resembling amyloid. Non-calcifying epithelial odontogenic tumours are very rare and unusual. Only five cases have been reported in the English literature till date. Here, we present an additional case of non calcifying type along with a review of previously reported cases. It has a much lower recurrence and malignant transformation rate.

Keywords CEOT · Stratum intermedium · Odontogenic tumour

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Introduction

Calcifying Epithelial Odontogenic Tumour (CEOT) is a benign odontogenic tumour of epithelial origin recognized by Thonay Goldman [1] and described as separate entity by Dutch pathologist Jens Jorgen Pindborg in 1955 which was accomplished in 1958 [2]. Shafer et al. in 1963 gave the eponym Pindborg tumour [3]. It is thought to arise from remnants of cells in stratum intermedium layer which is the epithelial element of the enamel [1]. It accounts for less than 1 % of all odontogenic neoplasms. Typical histopathological features comprise polygonal epithelial cells, calcifications and eosinophilic deposits close to amyloid [4]. An extensive research has revealed only 5 well documented cases of non-calcifying type of CEOT. Here, a sixth case is being reported along with review of previously reported cases and a brief discussion on histogenesis of this variant has been described.

Case Report

A 27 year old female patient came with a chief complaint of swelling in lower left back tooth region. The swelling was progressively increased in size over a period of 6 months and it was painless. Extraorally the patient presented with a diffuse swelling on the left side of the face measuring approximately 2.5 × 2 cm in size (Fig. 1). Single, left submandibular lymph node was palpable. The patient's medical history and general physical examination were non contributory.

Intraorally, obliteration of buccal vestibule from 34 to 38 was noticed (Fig. 2). Absence of 37, grade I mobility in relation to 38 was observed. The swelling was hard in consistency and non-tender. Investigations were performed.

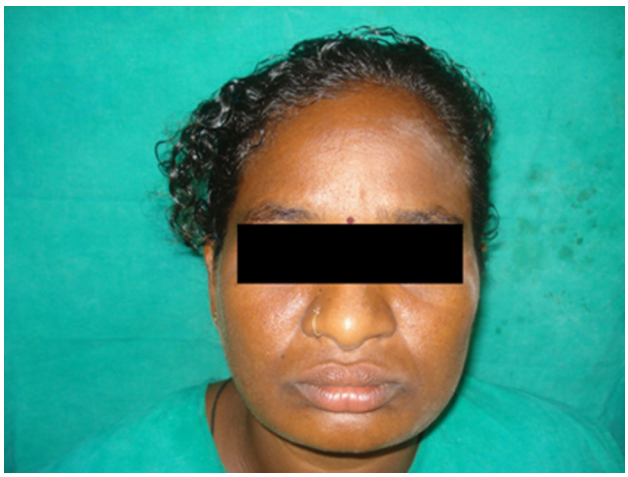

Fig. 1 Extraorally diffuse swelling seen on left side of face

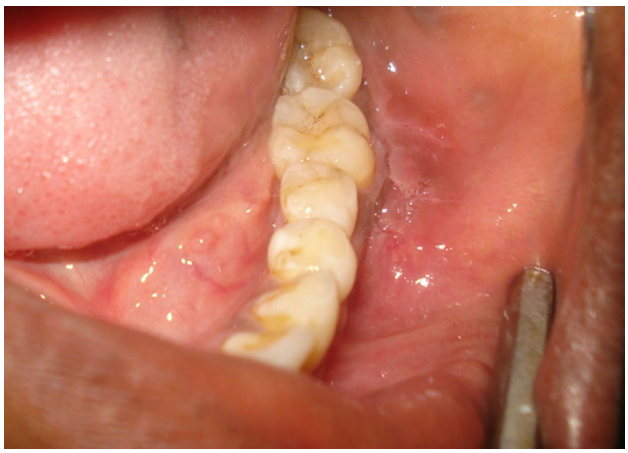

Fig. 2 Intraorally obliteration of buccal vestibule from 34 to 38

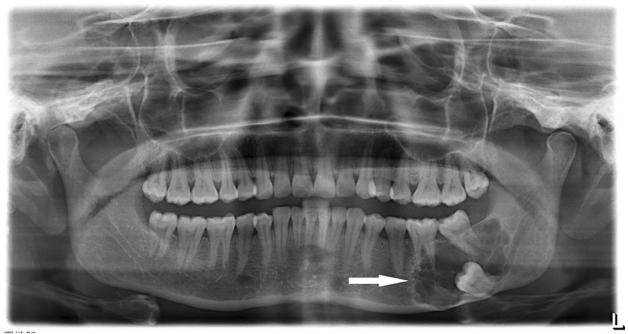

Fig. 3 Orthopantamograph revealing multilocular radiolucency with areas of radiopacity in the region of 36 and unerupted 37

On panoramic radiograph, multilocular radiolucency extending from 36 to 38 region posteriorly with irregular borders was seen. Unerupted 37 and mesial migration of 38 have been noticed (Fig. 3). On aspiration no fluid or any other material was obtained. Based on these findings a provisional diagnosis of Ameloblastoma of mandible was given.

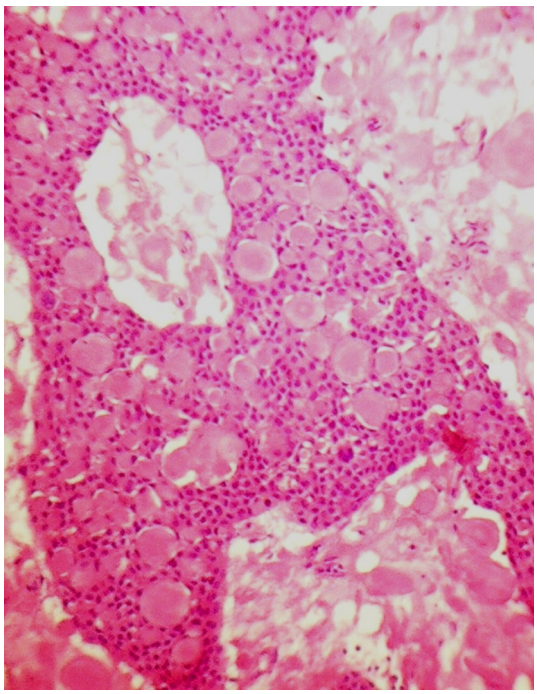

Fig. 4 Photomicrograph showing sheets of polyhedral tumour cells with prominent eosinophilic cytoplasm and pools of amorphous eosinophilic amyloid

An incisional biopsy was carried out. Histopathology showed strands and sheets of polyhedral epithelial cells with some cells showing nuclear pleomorphism, extensive areas of amorphous eosinophilic amyloid like material was enclosed between epithelial cells (Fig. 4). A diagnosis of non-calcifying type of CEOT was made. The tumour area was surgically exposed and a wide surgical excision was done (Fig. 5a). The surgical margin was clear of any tumour tissue and reconstruction was done by placing iliac crest graft (Fig. 5b) and reconstruction plates (Fig. 5c). The excised specimen was sent for histopathological examination (Fig. 5d). Post-operative followup was done (Figs. 6, 7). The histopathological features were correlated with that of incisional biopsy and a final diagnosis of non-calcifying type of CEOT was confirmed.

Discussion

It is an unusual odontogenic neoplasm with unpredictable biologic behavior ranging from very mild to moderate invasiveness [5]. Adenoid ameloblastomas, atypical ameloblastomas or cystic odontoma are the substitutional terms for CEOT [6]. CEOT may exist as an

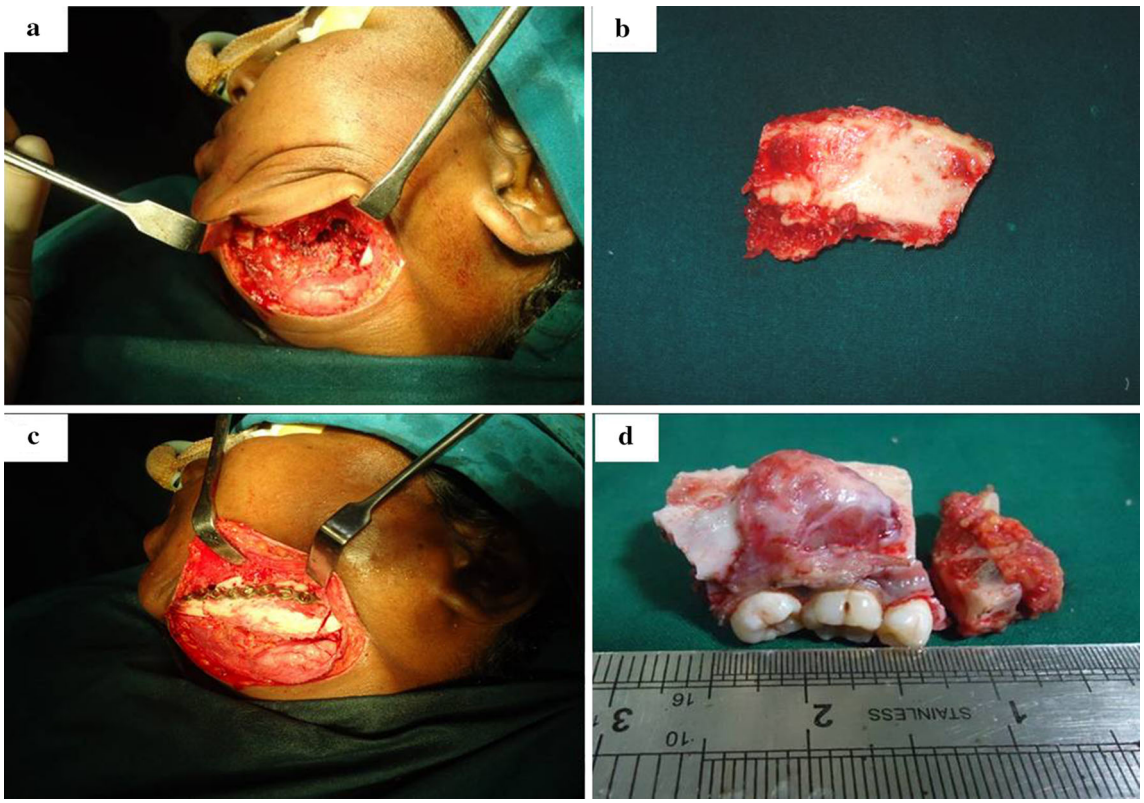

Fig. 5 **a** Wide surgical excision of tumor, **b** iliac crest graft, **c** reconstruction was done by placing iliac crest graft and reconstruction plates, **d** surgically excised specimen

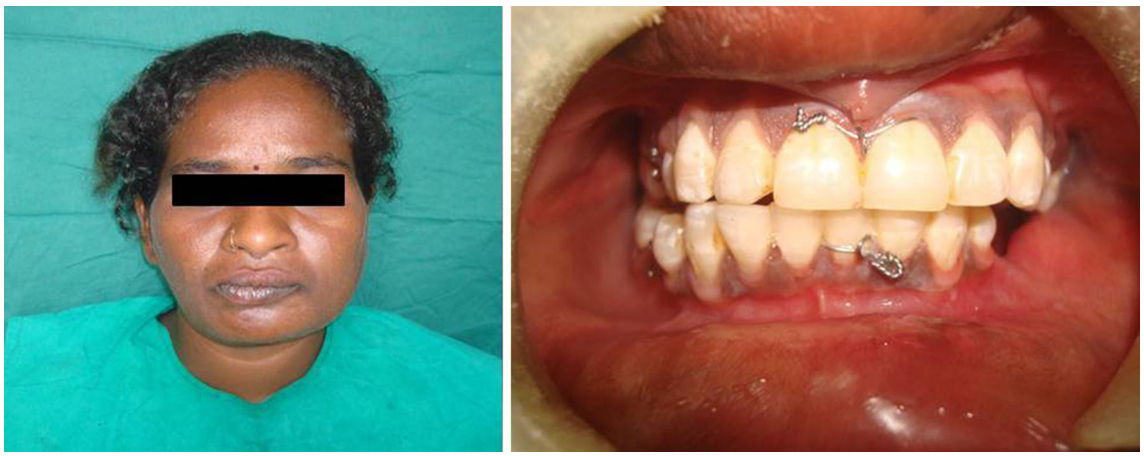

Fig. 6 Post-operative followup photograph (extraoral and intraoral)

intraosseous (central type) or extraosseous (peripheral type) tumour [4]. The histogenesis of this tumour is indecisive and the tumour cells is thought to arise from the stratum intermedium of enamel in case of intraosseous variant, whereas from dental lamina epithelial rests or the basal cells of gingival epithelium in case of the extraosseous variant [4, 6].

Symptomatically, CEOT may be associated with unerupted tooth which presents as painless, calm growing intraosseous mass in the mandible [1]. Seldom patients often complain of associated pain, nasal stuffiness, epistaxis, or headache [7]. Mean age of occurence is 40 years with no sex predominance. Intraosseous CEOT (85 %) accounts for more number of reported cases compared to

that of extraosseous CEOT (6 %) [4]. About two-third cases arise in the molar area which is the most frequent site [1]. In the present case CEOT is associated with unerupted mandibular left second molar.

On radiographs, initially the tumour presents as completely radiolucent. As the lesion progresses it matures and turn into larger pattern which is a mixed radiolucent-radiopaque [5]. Occasionally the lesion may range from unilocular and converts to multilocular with a honey-comb pattern. In few cases multiple radio-opacities are present within the radiolucent area, which creates characteristic "driven snow appearance" [3]. In the present case tumour revealed multilocular radiolucency associated with an impacted 37.

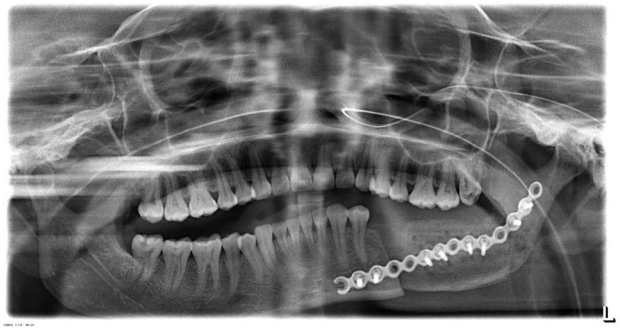

Fig. 7 Post-operative orthopantamograph revealing reconstruction

Histopathology remains gold standard for diagnosis [4]. CEOTs are unencapsulated, infiltrating tumours [5] which are identified by certain features like the existence of epithelial cells, uniform eosinophilic amyloid-like material and calcifications. The epithelial cells are polygonal with eosinophilic cytoplasm, vesicular nuclei and prominent nucleoli. They are arranged in the form of nests and sheets. The precise origin of amyloid is not yet revealed but is thought to arise from the secreted product of tumor epithelial cells by filamentous degradation of keratin filaments [4]. SlootWeg recommended that initiative stimulus for the production of collagen matrix is amyloid like material which is intended to mineralize resembling cementum [8] (Fig. 8).

Krolls and Pindborg stated that the presence or absence of calcification in CEOT has prognostic implications. A lack of calcification indicates less tumour differentiation and hence more chance of a recurrence. Pindborg has also reported recurrence after removal in a CEOT with minimal calcifications [1]. Five cases of non calcifying type of CEOT were reported previously by Aufdermaur [9], Takata et al. [10], Hafian et al. [11], Kaushal et al. [4], Afroz et al. [1]. Here we present an additional case of CEOT with complete absence of calcification (Table 1).

The treatment varies from simple enucleation or curettage to radical and extensive resection which includes hemimandibulectomy or hemimaxillectomy. Local

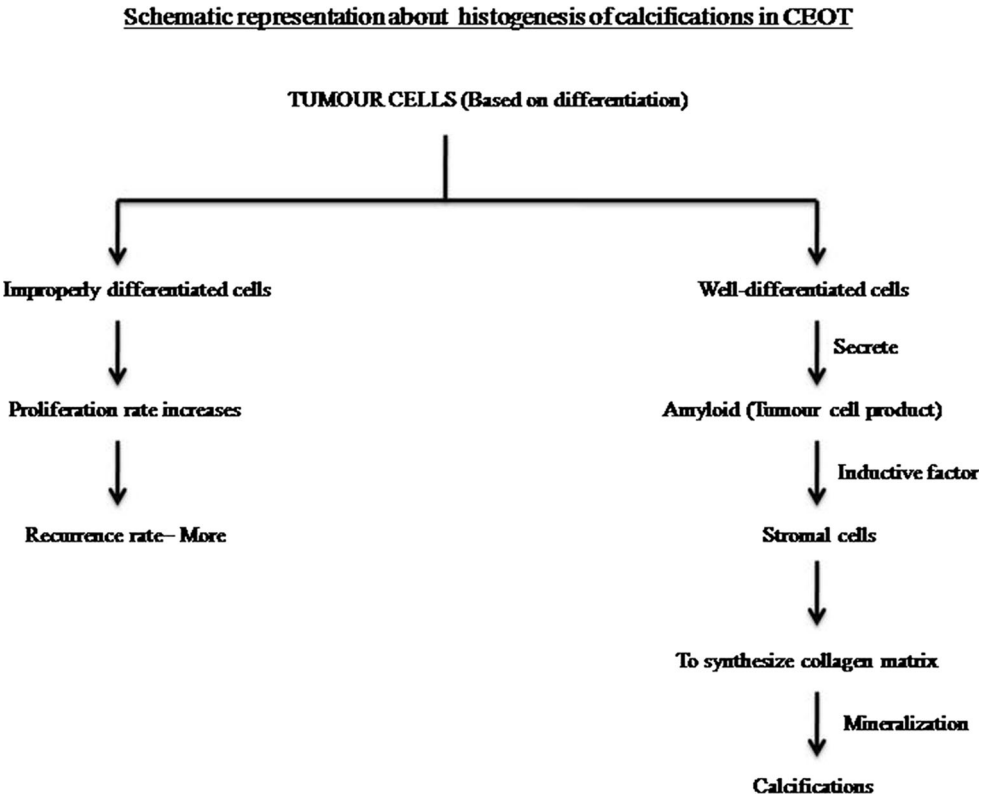

Fig. 8 Schematic representation about histogenesis of calcifications in CEOT

Table 1 Review of non calcifying type of CEOT

Author	Age	Gender	Site	Variant
Aufdermaur [9]	68	Male	Mandible	Intraosseous
Takata et al. [10]	58	Male	Left maxillary canine & premolar region	Intraosseous
Hafian et al. [11]	61	Male	Anterior maxilla	Intraosseous
Kaushal et al. [4]	57	Male	Angle of the mandible on right side	Intraosseous
Afroz et al. [1]	20	Female	Anterior maxilla	Extraosseous
Present case	26	Female	Left side of angle of mandible	Intraosseous

recurrence rates reported is about 10–15 % with rare malignant transformation [2]. In the present case surgical excision was done and the patient is on regular follow-up and the tumour has not shown any evidence of recurrence even after 2 years of surgical excision.

Conclusion

To the best of our knowledge, this is the sixth case of non calcifying type of CEOT reported in the orofacial region which highlights the importance of unusual presentation and microscopic features and also emphasizes on the histogenesis regarding calcifications. It is essential to identify non calcifying type of CEOT as it has prognostic implications. Hence, prompt appreciation can guide surgical treatment which allows the clinician for follow-up.

Compliance with Ethical Standards

Conflict of interest None.

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