

# Case Report

## Recurrent Peripheral Calcifying Cystic Odontogenic Tumor (Calcifying Odontogenic Cyst)

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### ABSTRACT

Peripheral calcifying cystic odontogenic tumor (PCCOT) is a rare odontogenic lesion, which represents about 1% of jaw cysts. Here we report clinicopathologic, radiographic and CT scan images of a new case of recurrent large PCCOT located in mandibular gingiva in a 77-year-old male with history of a nodular mass in the same area, which had been completely excised 3 years ago. In reviewing the literature, we found 55 previous reported cases with a mean age of 48.8 yr at the time of diagnosis. A majority of cases appear as a circumscribed nodule less than 1.5 cm in greatest diameter, without bony involvement. No case larger than 4cm in diameter has been reported. Only one case of recurrent PCCOT was seen in reported cases.

**Key words:** Odontogenic Tumors, Recurrence, Calcifying Odontogenic Cyst

### Introduction

Calcifying odontogenic cyst (COC) or Gorlin cyst was described first by Gorlin (1) and Pretorius (2) as a definite pathologic entity. A rare developmental odontogenic lesion (1% of jaw cysts) is described as having features of both cystic and solid neoplasm (3-5). The latest WHO classification of odontogenic tumors (done in 2005, Table 1) renamed the cystic type of this entity as calcifying cystic odontogenic tumor (CCOT) with four subtypes: 1) simple cystic 2) odontoma associated 3) ameloblastomatous proliferating and 4) CCOT

associated with benign odontogenic tumors other than odontoma (6). The term dentinogenic ghost cell tumor (DGCT) was retained for neoplastic type (6). Most cases of CCOT arise centrally within bone, but occasionally they may occur peripherally (7, 8). Peripheral calcifying cystic odontogenic tumor (PCCOT), also known as the "extra osseous CCOT" is similar to CCOT in histological characteristics but occurs in the soft tissue overlying the tooth-bearing areas of the maxilla and mandible, most often as a slowly enlarging asymptomatic mass with a mean size of 1.5cm (7-9). Recurrence is very rare and only one case of recurrent PCCOT has been reported (6).

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**Table 1:** 2005 WHO calcification of CCOT and DGCT

<p>Calcifying cystic odontogenic tumor(CCOT) includes peripheral and central cases:</p> <p>CCOT type 1:simple cystic</p> <p>CCOT type 2:odontoma associated</p> <p>CCOT type 3:ameloblastomatous proliferating.</p> <p>CCOT type 4:CCOT associated with benign odontogenic tumors other than odontoma</p> <p>Dentinogenic ghost cell tumor(DGCT):</p> <p>DGCT type 1:central ,solid,aggressive variant</p> <p>DGCT type 2:Peripheral ,less aggressive variant</p>
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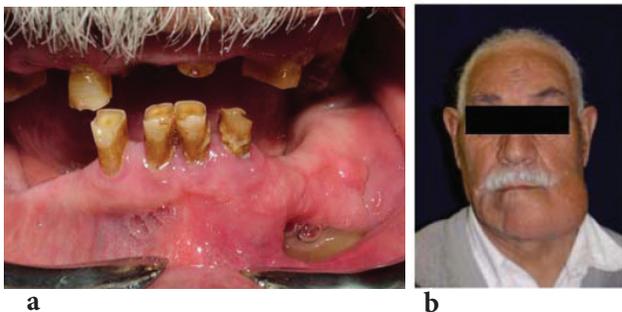
CCOT : Calcifying cystic odontogenic tumor; DGCT: Dentinogenic ghost cell tumor

This article presents a unique case of large recurrent PCCOT with rapidly enlargement that also caused face enlargement in left mandibular area and reviews previously reported cases of PCCOT in literature and Shahid Beheshti Oral and Maxillofacial Pathology department cases during 10 years.

### Case report

A 77-year-old man referred to our Pathology center with a painless mass about 7 cm in diameter in left mandibular buccal gingiva extending from molar teeth to anterior region. The patient had noticed the mass from 8 months ago, but he stated that the lesion increased rapidly in size during past 2 months. The lesion was firm on palpation and the overlying mucosa was normal in color and smooth in texture (Fig. 1a). Other intraoral examination was normal and no problem was detected. The lesion extraorally caused face swelling in left mandibular area (Fig. 1b). History of patient showed similar gingival mass but smaller in size (3x1.5x1cm) involving left mandibular buccal gingiva with pathologic diagnosis of PCCOT, 3 years ago.

The lesion has been completely excised then. No recurrence was present after 1- year follow up.



**Fig. 1:** a- Large nodular mass in left side of mandibular gingiva. b- Face enlargement caused by intra-oral lesion

Based on previous diagnosis, aspiration of the lesion was done and approximately 15 cc yellow fluid was extracted. Panoramic X-ray of the present lesion showed superficial erosion of underlying bone in some area (Fig. 2a).

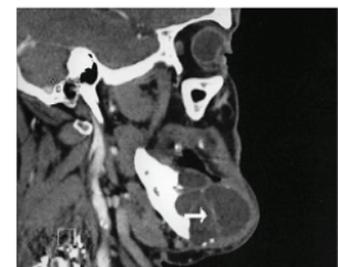
CT scan imaging showed well-defined homogenous soft tissue mass with internal Septa, extending from left ramus area to anterior area of mandible crossing the midline and extended to right side (Fig. 2b,2c).



a



b



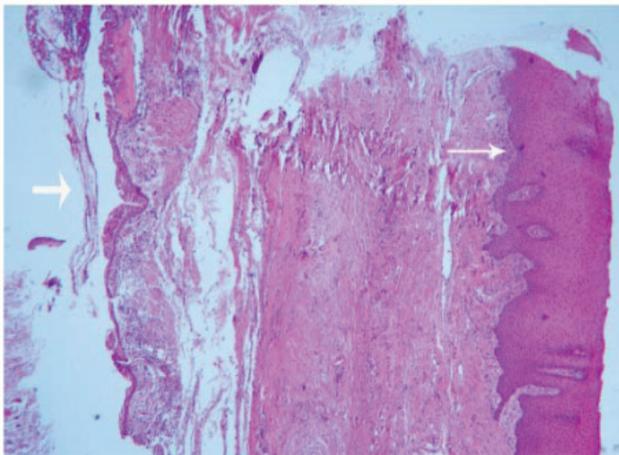
c

**Fig. 2:**a- panoramic radiograph showing superficial erosion in left mandibular area.(arrows)b-Axial view of CT shows extension of lesion from left side to right side.c- Sagittal view of CT shows soft tissue mass with internal septa

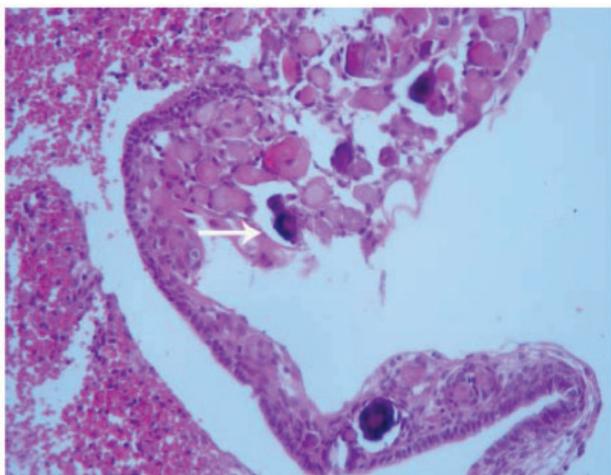
The lesion was completely excised under general anesthesia. Grossly the mass was creamy brown in color, covered with mucosa in some area, measuring 7x4.5x1.3cm. The specimen was submitted to oral and maxillofacial

pathology service and prepared for histological examination with hematoxylin and eosin staining.

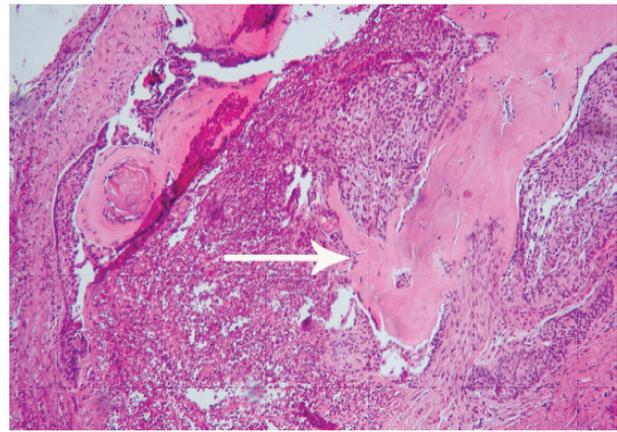
Microscopically, sections revealed oral mucosa with parakeratinized stratified squamous epithelium and underlying fibrous connective tissue. Within the connective tissue, a well circumscribed cystic lesion lined by a characteristic layer of cuboidal or columnar cells was seen (Fig. 3). The overlying layer of epithelium lining was loose and myxomatous resembles the stellate reticulum. Several foci of eosinophilic ghost cells which some of those undergone calcification was also present adjacent to epithelial component (Fig. 4). Irregular foci of tissue resembling dentin or osteodentin were also observed along the periphery of the lesion (Fig. 5). Based on histological findings, a diagnosis of peripheral calcifying cystic odontogenic tumor, type 1 (simple cystic) was made.



**Fig. 3:** Oral mucosa epithelium (thin arrow) and cyst lining in oral mucosa connective tissue (thick arrow). (H&E staining x100)



**Fig. 4:** Epithelial lining of cyst with ghost cells which show calcification in some area. (H&E staining x200)



**Fig. 5:** Dentin like material (H&E staining x200)

## Discussion

PCCOT is an extremely rare odontogenic lesion which is the soft tissue counterpart of CCOT (4,5). Our review of literature showed 55 definite reported cases of PCCOT (9-14). On the other hand, reviewing the Shahid Beheshti Oral and Maxillofacial Pathology Department in a 10-year period also indicating rarity of the lesion. Among 4624 biopsies in this period, there were 543 odontogenic lesions. Out of 543 odontogenic lesions, only two cases were qualified as PCCOT. According to these data, its incidence ranges from 0.2% to 0.6% of odontogenic lesions (6,9-14), and it represents 8% of peripheral odontogenic tumors (15).

PCCOT most often presents as a slowly enlarging asymptomatic mass with smooth surface in the gingiva or alveolar mucosa. The mean age at initial presentation is 48.8 yr (ranged 9 to 92 yr) but the highest incidence is in the 6<sup>th</sup> to 8<sup>th</sup> decades of life. Men and women are affected almost equally. Mandible was the most common site of PCCOT especially in incisor cuspid region and premolar area (9-11,13,16). As most reported cases, our case occurred in 8<sup>th</sup> decade of life in mandibular gingiva. The size of the lesion ranged from 0.5 to 4 cm in greatest diameter and the majority of cases were less than 1.5 cm. However, present case was 7x4.5x1.3 cm in size which is unique and a mass greater than 4 cm hasn't been reported (6,9). Recurrence is also very rare and only one case in a 60-year female located in gingival tissue, with histopathologically diagnosed as type 1 PCCOT similar to our case has been reported (6).

Clinically, a variety of diagnosis are made by clinicians, the most common are fibroma or fibrous hyperplasia. Other differential diagnoses are gingival cyst of adult and peripheral ameloblastoma (9). Its

interesting to note that the mean age of PCCOT (48.8 yr) is similar to that of patients with gingival cyst of adult (mean age 48.2 yr) and peripheral ameloblastoma (mean age 52.3 years), in contrast to CCOT which occurs frequently in second decade of life (13). However, currently the origin of PCCOT is not known but regarding to histogenesis of PCCOT two major sources of origin may be considered. The lesion which are located entirely within connective tissue of gingiva and separated from the surface epithelium by a band of connective tissue (just like present case) probably arise from remnants of dental lamina whereas other lesions appear arising from surface epithelium. A similar histogenesis has been suggested for peripheral ameloblastoma and gingival cyst of adult. More data are needed to draw conclusion relative to any differences in their nature and behavior (9, 15).

Radiographically, PCCOT shows no bony involvement and in few cases, superficial erosion, like our case was seen (4-6, 9, 10, 12).

Microscopically four variants of PCCOT like CCOT were distinguished: type 1 (simple cystic), type 2 (odontoma associated), type 3 (ameloblastomatous proliferating), type 4 (PCCOT associated with benign odontogenic tumors other than Odontoma) (6).

Type 1 is the most common pattern characterized by cystic epithelial lining in which a well-defined basal layer of columnar cells and overlying layers of resemble stellate reticulum of enamel organ and masses of ghost cells which may be located within the epithelial lining or fibrous capsule (16). Sometimes variable quantities of dentinoid material are laid down adjacent to epithelial lining. In some instances, melanin or clear cells could be identified (6). The microscopic differential diagnosis especially type 1 & 3 must be made with peripheral Ameloblastoma. Both lesions share the presence of prominent elongated basal cells and Stellate reticulum zone, but the presence of ghost cells is rare in ameloblastoma (12).

Furthermore, we should notify that some of peripheral reported lesion (30%) are solid type (DGCT) and diagnosis is very important (6, 12) because neoplastic type is more aggressive than cystic type and needs more consideration.

Conclusion: As a conclusion, we reported a rare peripheral odontogenic lesion (PCCOT) in mandibular gingiva which was large and recurrent with rapidly enlargement in 2 months. So it is the first case of PCCOT regarding its size and second recurrent case report (6).

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