Central granular cell odontogenic tumor: Report of an unusual case

Mani Madan, Shaleen Chandra, Vineet Raj, Rohit Madan

ABSTRACT

Central granular cell odontogenic tumor (CGCOT) is an unusual benign odontogenic neoplasm characterized by the presence of granular cells associated with apparently inactive odontogenic epithelium. These tumors tend to occur in the posterior mandible and usually present as well-defined unilocular or multilocular radiolucent lesions. So far, only <40 cases of CGCOT have been described in the literature under various terminologies. Though these tumors were not considered as distinct entity in the recent WHO classification of odontogenic tumors, long-term follow-up is recommended as malignant counterpart of CGCOT has already been reported. The main aim of this article is to report an additional case of CGCOT to the literature, occurring in a 73-year-old male.

Key words: Central granular cell odontogenic tumor, granular cell, odontogenic tumor

CASE REPORT

A 73-year-old male patient presented with a hard swelling in lower front region of the jaw. Extraorally, it extended from left corner of the mouth to the mid part of the body of the mandible on the right side. Intraorally extended from 33 to 46 regions on both buccal and lingual aspect. Swelling gradually increased from past 3 years to attain the present size. On palpation, it was firm to bony hard in consistency and was not associated with any pain or paresthesia. The lesional area was partially edentulous, and the overlying skin was erythematous. Slight facial asymmetry with right submandibular lymphadenopathy was also observed. Radiographic examination revealed an expansile multilocular radiolucency extending from 33 to 46 regions with scalloped borders. Based on above clinical findings, a provisional diagnosis of central giant cell granuloma or ameloblastoma was considered.

An incisional biopsy was performed, and specimen was sent for histopathological examination. Formalin fixed, paraffin embedded, hematoxylin, and eosin stained sections of the specimen revealed the presence of granular cells associated with thin cords, discreet islands, and follicles of odontogenic epithelium. Many granular cell ameloblastoma may indeed represent CGCOT.

Central granular cell odontogenic tumor (CGCOT) is an exceedingly rare benign odontogenic neoplasm; the first description of this tumor was reported by Werthemann in 1950 under the term spongiocytic adamantinoma. From then, many authors have suggested various terminologies for this tumor which includes, granular cell ameloblastic fibroma, central granular cell tumor of the jaw, central granular cell odontogenic fibroma, and central odontogenic fibroma granular cell variant. Still, a great deal of controversy exists in the nomenclature of CGCOT as it was not considered as a distinct entity in the recent WHO classification of odontogenic tumors, but few recently published literature suggest the term CGCOTs for tumors characterized by varying amount of large eosinophilic granular cells with eccentrically placed nuclei associated with apparently inactive odontogenic epithelium. Many granular cell ameloblastoma may indeed represent CGCOT.

Till today, to the best of author’s knowledge only 38 cases of CGCOT have been reported in the literature. Considering the rarity of the tumor and discuss the nature of these tumors we are reporting a case of CGCOT affecting the anterior mandible and crossing the midline in a 73-year-old male.
stoma [Figures 3 and 4]. These granular cells were oval to polygonal with finely granular eosinophilic cytoplasm and eccentrically placed nuclei. Majority of the granular cells were found in association with odontogenic islands lined by ameloblasts like cells [Figures 5 and 6]. These findings favored the final diagnosis of granular cell odontogenic tumor. Further complete excision of the lesion along with segmental resection of the mandible was performed and no evidence of recurrence after 9 months follow-up.

Figure 1: Intraoral swelling extending from 33 to 46 with cortical expansion

Figure 3: Photomicrograph showing thin cords of odontogenic islands (H and E, original magnification x100)

Figure 5: Granular cells within the odontogenic Island lined by ameloblast like cells (H and E, original magnification x400)

Figure 2: Orthopantomography demonstrating multilocular radiolucency with scalloped borders

Figure 4: Photomicrograph showing proliferation of discreet follicles of odontogenic epithelial cells in the background of mature stroma (H and E, original magnification x100)

Figure 6: Photomicrograph showing collection of polygonal granular cells exhibiting finely granular eosinophilic cytoplasm with eccentrically located nuclei associated with odontogenic islands (H and E, original magnification x400)
DISCUSSION

CGCOT usually represents as asymptomatic, slow growing expansile lesions of the jaw bone preferring mandibular molar area followed by posterior maxilla, and anterior mandible. Majority of the reported cases are seen in fifth to the seventh decade of life with definite female predilection (3.1:1). Radiographic findings of CGCOT's are mostly nonpathognomonic as these tumors can show varied features ranging from unilocular to multilocular radiolucency with sclerotic borders and sometimes with mixed radioopacities.⁸

Few odontogenic tumors generally exhibit the presence of granular cells which includes granular cell ameloblastic fibroma, central granular cell odontogenic fibroma, and granular cell ameloblastoma. The origin of the granular cells in these tumors has been a matter of dispute. In granular cell ameloblastoma, it has been speculated that the ability of the lysosomes to digest the aged components in the cytoplasm of the tumor cells decreases with age and this phenomena may lead to the formation of lysosomal granules within the cytoplasm. Positivity for the markers such as cytokeratin, CD 68, lysozyme, and alpha-1-antichymotrypsin indicates the epithelial origin of the granules.⁷ Whereas several studies suggested, granular cells of CGCOT shows positivity for vimentin and negativity for cytokeratin and these findings favors the mesodermal origin of granular cells, and also positivity for other macrophagic markers such as HLADR, CD1a, α-1-antitrypsin, and α-1-antichymotrypsin suggest macrophagic differentiation of granular cells.⁹,¹⁰

Histochemically, these cells stain positively for Sudan black B and periodic acid-Schiff diastase resistant and negative for mucicarmine stain.¹¹ Ultrastructural findings suggest that these cells are composed of numerous lysosome-like granules, with little cytoplasmic organelles and prominent indented nuclei. The lysosome-like granules of granular cells show the following appearances: Electron-dense and amorphous granules, aggregates of fine granular particles, vesicular granules, granules with myelin-like structure, and granules with mixed appearance of them.¹²

It is interesting to note that present case was examined in a 73-year-old male, affecting anterior mandible, with cortical expansion, facial disfigurement, and midline crossing. Among the reported cases, CGCOT commonly tends to occur in posterior mandibular region with high female predilection and rarely, the lesions behaved aggressively and showed cortical expansion, facial swelling. However, malignant counterpart of CGCOT was reported by Piattelli et al., under the term Granular cell odontogenic sarcoma or Malignant CGCOT. In their case, tumor cells revealed nuclear pleomorphism, prominent nuclei, and mitotic figures, with extensive invasion of the oral and respiratory mucosa and the adjacent soft tissues.¹³ Granular cell variant of ameloblastomas also appears to be an aggressive lesion.¹⁴ Present case showed two predominant histologic patterns. Most areas revealed the presence of eosinophilic granular cells in mature fibrous stroma associated with odontogenic epithelial islands, and few areas exhibit a proliferation of ameloblasts like cells in pleomorphic pattern admixed with large, round to polygonal cells containing abundant eosinophilic coarse granules. Morphologically the granular cells were of variable size, round to polygonal, with finely granular, eosinophilic cytoplasm and eccentrically located, ovoid to round nuclei. Similar microscopic findings were observed in the cases reported by Altini et al. and they preferred the term plexiform granular cell odontogenic tumor (PGCOT) and combined granular cell ameloblastoma with PGCOT respectively.¹⁵,¹⁶

The treatment modalities of CGCOT include conservative surgical enucleation with reconstruction.¹⁶ Long-term follow-up is recommended because the malignant counterpart of CGCOT have already been reported.

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Conflicts of interest
There are no conflicts of interest.

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