

# Ghost Cell Odontogenic Carcinoma-a review of the literature

Shermil Sayd<sup>1\*</sup> and Suresh Vyloppilli<sup>2</sup>

<sup>1</sup>Department of Oral and Maxillofacial Surgery, Kannur Dental College, Anjarakandy, India

<sup>2</sup>Malankara Orthodox Syrian Church Hospital and Medical College, Kolenchery, India

Received: July 14, 2017; Published: August 01, 2017

**\*Corresponding author:** Shermil Sayd, Department of Oral and Maxillofacial Surgery Kannur Dental College, Anjarakandy, India-670712, Tel: +919446230425; Email: [shermil12@gmail.com](mailto:shermil12@gmail.com)

## Abstract

Ghost Cell Odontogenic Carcinoma is an extremely rare odontogenic tumor with unclear etiology. Although known by many names, the basic histologic identifying feature includes the identification of the ghost cells and the malignant changes associated with the cells. These tumors show a spectrum of growth from a slow-growing locally invasive tumor to a highly aggressive, rapidly growing, infiltrative tumor. Therefore, it is prudent to perform a wide local excision with histologically clear margins. Is seen associated with squamous cell carcinoma, radiotherapy also should be included in the treatment plan. Long term follow-up is highly recommended.

**Keywords:** Calcifying Odontogenic Cyst; Odontogenic Ghost Cell Carcinoma; Ghost Cell Tumor

**Abbreviations:** COC: Calcifying Odontogenic Cyst; WHO: Health Organization; OGCC: Odontogenic Ghost Cell Carcinoma; EOGCT: Epithelial Odontogenic Ghost Cell Tumor

## Introduction

The calcifying odontogenic cyst (COC), classified as an odontogenic tumor by the World Health Organization (WHO), was first identified as a distinct entity by Gorlin et al. in 1962 [1]. In 1972, Fejerskov and Krogh [2] used the label calcifying ghost cell odontogenic tumor. The Odontogenic Ghost Cell Carcinoma (OGCC) belongs to the fourth of the four categories of WHO classification of odontogenic carcinoma. In 1981, Praetorius et al. [3] recognized four different histological patterns of COC and classified them as type 1A (simple unicystic), type 1B (odontome-producing), type 1C (ameloblastomatous proliferating) and type 2 (dentinogenic ghost cell tumor). They opined that although type 2 shows histological characteristics of cystic variants, its solid growth pattern suggests that its classification as a neoplasm is more appropriate. Ellis and Shmookler [4] used the term Epithelial Odontogenic Ghost Cell Tumor (EOGCT) for the neoplastic variant of COC. Although inconsistent, the ghost cells depicted by the terminology are the most distinctive histological feature of this tumor.

OGCC is exceedingly rare, with only 18 cases reported in the English literature to date. A photograph published in 1971 WHO monograph with no clinical information got touted as the first reported case in literature [5]. Ikemura et al. [6] reported the first well-documented case of an OGCC to appear in the English literature. Although described by a variety of terms, including malignant COC [6-8] odontogenic ghost cell carcinoma [9-11] aggressive epithelial

ghost cell odontogenic tumor, [4,12] dentinogenic ghost cell ameloblastoma [13] and malignant calcifying ghost cell odontogenic tumor [14], they all demonstrated malignant histological features such as infiltration, cellular pleomorphism, numerous mitoses and necrosis.

An epithelial lining with well defined basal columnar cells along with a stellate reticulum mimicking cells and masses of ghost cells which may or may not be calcified forms the histologic diagnostic criteria. Commonly seen accompanying the above includes atypical epithelial cell foci exhibiting mitosis, keratin pearls, necrosis and other malignant features [5]. Unlike its benign counterpart, OGCC has a slight male predilection with a ratio of 3.4:1 [8,14]. It usually appears as a painful, hard swelling in the maxilla or mandible with osseous destruction, and may or may not exhibit paresthesia. It may cause expansion of the mandible or maxilla. Radio graphically; OGCCs description depicts it as a purely radiolucent or mixed lesion. Conventional radiographs show a large, poorly defined osteolytic lesion of the jaw with several foci of increased radiopacity within the pathological site. Radiographs are insufficient to make a standalone diagnosis of OGCC. Their characteristics mimic malignant tumors, and there is no pathognomonic feature for the diagnosis. Other possible conditions to which the OGCC radio graphically relates to comprises of osteosarcoma and malignant ameloblastoma. Histological examination is the only method to attain a definitive diagnosis.

Scrutinization of the available literature revealed that OGCC has a tendency towards the Asian population, and the same is exhibited by its benign counterpart too [15]. The biologic behavior of OGCC is unpredictable, supported by the fact that some authors reported the case with relatively indolent growth [16] and others by an aggressive and potentially fatal course [17] and this reflects a spectrum of growth patterns from a slowly growing, locally aggressive tumor to a highly aggressive and rapidly growing neoplasm. Distant metastases are uncommon with only one case of pulmonary metastases reported [17]. Identifying the malignant epithelial component in association with a classical benign COC has become the basis of diagnosis for OGCC.

Despite showing similarities in pathological characteristics, COC and OGCC have significant differences which help in predicting their malignancy potential. Furthermore, measurement of the expressed biomarkers, Ki-67 and MMP-9, associated with tumor proliferation and invasion, is useful in evaluating the prognosis of OGCC.

## Management

Literature reviews have shown that the patient reporting with OGCC often has a history of multiple recurrences of COC or long-term persistent swelling followed by the onset of rapid, painful swelling before a definitive diagnosis of OGCC [18]. Clinically, this evidence is sound enough to suspect a malignant degeneration of a long-standing or recurrent benign process into a malignant process. Therefore, it should be suspicious, and considering the biological unpredictability, careful, long-term follow-up is highly recommended after therapy [19].

The recommended treatment for OGCC has been wide surgical excision [20,21]. The controversy revolves around the use of postoperative radiation therapy with or without adjuvant chemotherapy. Goldenberg et al. [22] reported a case of OGCC where he achieved surgically clean margins with no clinical or radiographic evidence of local or distant metastasis, and postoperative radiotherapy was not initiated. Most of the authors who reported a case of OGCC has refrained from giving adjuvant radiotherapy due to the debacle over the treatment protocol. Multiple authors stated that they did not have are currency despite having not given radiotherapy.

Recurrence and metastasis can occur years after the excision of the primary lesion. A 3-year delay was reported by M. Kumar et al. [23] for a case, but up to 11 years may elapse before the full extent of spread is apparent [24]. There have been instances of OGCC showing characteristics of squamous cell carcinoma. In such cases, there commendation is for a combined approach of all the modalities [25].

## Conclusion

OGCCs are rare malignant counterparts of calcifying odontogenic cysts. These tumors show a spectrum of growth from a slow-growing locally invasive tumor to a highly aggressive, rapidly growing, infiltrative tumor. Therefore, it is prudent to perform a wide local excision with histologically clear margins. So

it is recommended that close long-term surveillance of recurrent or long- standing benign COCs and OGCC.

## References

1. Gorlin RJ, Pindborg JJ, Clausen FP, Vickers RA (1962) The calcifying odontogenic cyst-a possible analogue of the cutaneous calcifying epithelioma of Malherbe. An analysis of fifteen cases. *Oral Surg Oral Med Oral Pathol* 15: 1235-1243.
2. Fejerskov O, Krogh J (1972) The calcifying ghost cell odontogenic tumor or the calcifying odontogenic cyst. *J Oral Pathol* 1(6): 237-287.
3. Praetorius F, Hjorting-Hansen HE, Gorlin RJ, Vickers RA (1981) Calcifying odontogenic cyst. Range, variations and neoplastic potential. *Acta Odontol Scand* 39(4): 227-240.
4. Ellis GL, Shmookler BM (1986) Aggressive (malignant?) epithelial odontogenic ghost cell tumor. *Oral Surg Oral Med Oral Pathol* 61(5): 471-478.
5. Pindborg JJ, Kramer IRH, Torloni H (1971) Calcifying odontogenic cyst. In: Pindborg JJ, Kramer IRH, Torloni H (eds) *Histological typing of odontogenic tumours, jaw cysts and allied lesions*. WHO International histological classification of tumours. World Health Organization, Geneva, Switzerland, p. 18.
6. Ikemura K, Horie A, Tashiro H, Nandate M (1985) Simultaneous occurrence of a calcifying odontogenic cyst and its malignant transformation. *Cancer* 56(12): 2861-2864.
7. Dubiel Bigaj M, Olszewski E, Stachura J (1993) The malignant form of calcifying odontogenic cyst. A case report. *Patologia Polska* 44(1): 39-41.
8. Buchner A (1991) The central (intraosseous) calcifying odontogenic cyst: an analysis of 215 cases. *J Oral Maxillofac Surg* 49(4): 330-339.
9. Grodjesk JE, Dolinsky HB, Schneider LC, Dolinsky EH, Doyle JL (1987) Odontogenic ghost cell carcinoma. *Oral Surg Oral Med Oral Pathol* 63: 576-581.
10. Alcalde RE, Sasaki A, Misaki M, Matsumura T (1996) Odontogenic ghost cell carcinoma: report of a case and review of the literature. *J Oral Maxillofac Surg* 54: 108-111.
11. Folpe AL, Tsue T, Rogerson L, Weymuller E, Oda D, et al. (1998) Odontogenic ghost cell carcinoma: a case report with immunohistochemical and ultrastructural characterization. *J Oral Pathol Med* 27(4): 185-189.
12. Siar CH, Ng KH (1994) Aggressive (malignant?) epithelial odontogenic ghost cell tumour of maxilla. *J Laryngol Otol* 108(3): 269-271.
13. Scott J, Wood GD (1989) Aggressive calcifying odontogenic cyst—a possible variant of ameloblastoma. *Br J Oral Maxillofac Surg* 27(1): 53-59.
14. Toida M (1998) So-called calcifying odontogenic cyst: review and discussion on the terminology and classification. *J Oral Pathol Med* 27(2): 49-52.
15. Buchner A (1991) The central (intraosseous) calcifying odontogenic cyst: An analysis of 215 cases. *J Oral Maxillofac Surg* 49(4): 330-339.
16. McCoy BP, O'Carroll MK, Hall JM (1992) Carcinoma arising in a dentinogenic ghost cell tumor. *Oral Surg Oral Med Oral Pathol* 74(3): 371-378.
17. Grodjesk JE, Dolinsky HB, Schneider LC, Dolinsky EH, Doyle JL (1987) Odontogenic ghost cell carcinoma. *Oral Surg Oral Med Oral Pathol* 63: 576-581.
18. Lu Y, Mock D, Takata T, Richard CK (1999) Odontogenic ghost cell carcinoma: Report of four new cases and review of the literature. *J Oral Pathol Med* 28(7): 323-329.
19. Kim HJ, Choi SK, Lee CJ, Suh CK (2001) Aggressive epithelial odontogenic ghost cell tumor in the mandible: CT and MR imaging findings. *AJNR Am J Neuroradiol* 22(1): 175-179.

20. Kamijo R, Miyaoka K, Tachikawa T (1999) Odontogenic ghost cell carcinoma: Report of a case. J Oral Maxillofac Surg 57: 1266-1270.
21. Alcalde RE, Sasaki A, Misaki M (1996) Odontogenic ghost cell carcinoma: Report of a case and review of the literature. J Oral Maxillofac Surg 54: 108-111.
22. Goldenberg D, Sciubba J, Tufano RP (2004) Odontogenic ghost cell carcinoma. Head & neck 26(4): 378-381.
23. Kumar M, Fasanmade A, Barrett AW, Mack G, Newman L, et al. (2003) Metastasizing clear cell odontogenic carcinoma: a case report and review of the literature. Oral oncology 39(2): 190-194.
24. Fan J, Kubota E, Imamura H (1992) Clear cell odontogenic carcinoma: a case report with massive invasion of neighboring organs and lymph node metastasis. Oral Surg Oral Med Oral Pathol 74: 768-775.
25. Mosqueda Taylor A, Meneses Garcia A, Ruiz Godoyrivera LM, SuarezRoa ML, Luna Ortiz K (2003) Malignant odontogenic tumors. A retrospective and collaborative study of seven cases. Med Oral 8(2): 110-121.



### Assets of Publishing with us

- Global archiving of articles
- Immediate, unrestricted online access
- Rigorous Peer Review Process
- Authors Retain Copyrights
- Unique DOI for all articles

<http://biomedres.us/>