



## Dentinogenic Ghost Cell Tumour- A Review

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### Abstract

The calcifying odontogenic cyst (COC) was first described by Gorlin and his colleagues (1962), as a separate entity of odontogenic origin. The cystic lesions are termed as "calcifying cystic odontogenic tumors" (CCOT) and the neoplastic entity as a "Dentinogenic ghost cell tumor" (DGCT). It usually occurs in elderly persons with a slight male predilection. It shows a tendency to occur in the anterior segment of the jaws. Radiographically, DGCT shows a well-defined, mostly unilocular radiolucent to mixed radiolucent/radiopaque appearance depending on the amount of calcification. A meticulous clinical and radiographic examination can be forestalled under-diagnosis of this specific entity. In time referral of the patient for biopsy and histologic analysis, surgical intervention and a long-term follow-up are crucial for the successful treatment of the patient.

**Keywords:** Calcifying Odontogenic Cyst; Dentinogenic Ghost Cell Tumor; Calcifying Cystic Odontogenic Tumors; Granular Cell Odontogenic Tumor; Solid Dentinogenic Ghost Cell Tumor; Peripheral Dentinogenic Ghost Cell Tumor

### Abbreviations

COC: Calcifying Odontogenic Cyst; CCOT: Calcifying Cystic Odontogenic Tumors; DGCT: Dentinogenic Ghost Cell Tumor; GCOT: Granular Cell Odontogenic Tumor.

### Introduction

The calcifying odontogenic cyst (COC) was first described by Gorlin and his colleagues (1962), as a separate entity of odontogenic origin [1]. There are still arguments as to whether COC is a cyst or a tumor as entire lesions are not cystic, and the biological behavior is often not compatible with a cyst [2]. Based on this dualistic concept, some authors consider that COC comprised of two entities: a cystic and a neoplastic variety [3]. World Health Organization (WHO) has classified all COC's as neoplasms [4]. The WHO defined this tumor as "A locally invasive neoplasm characterized by ameloblastoma-like islands of epithelial cells in a mature connective tissue stroma. Aberrant keratinization may be found in the form of ghost cells in association with varying amounts of dysplastic dentin the cystic lesions are termed as "calcifying cystic odontogenic tumors" (CCOT) and the neoplastic entity as a "Dentinogenic ghost cell tumor" (DGCT) [1].

Dentinogenic ghost cell tumor is a rare tumor constituting only 11.5% of all COCs posing a significant challenge in diagnosis along with therapy [2]. It usually occurs in elderly persons with a slight male predilection. It shows a tendency to occur in the anterior segment of the jaws [2]. Microscopically, it consists of ameloblastomatous epithelial islands, with areas of ghost cell formation and a varying amount of dentinoid material [1]. DGCTs are more aggressive and much more likely to recur. Both local and metastatic cases have been reported so far. Granular cell odontogenic tumor (GCOT)'s are frankly malignant lesions often occurring as a result of the malignant transformation in a CCOT or DGCT.

The reported cases of DGCTs can develop peripherally or centrally. Peripheral tumors arise in extraosseous, gingival or alveolar mucosa. Depending upon the aggressive behavior two versions of the DGCT has been determined: the solid (aggressive) variant and a peripheral (non-aggressive) variant. The most common site of occurrence was found to be the mandible followed by maxilla.

Radiographically, DGCT shows a well-defined, mostly unilocular radiolucent to mixed radiolucent/radiopaque appearance depend-

ing on the amount of calcification [5]. Multilocular appearance also reported in the review of literature of 5 cases. Resorption of adjacent teeth and associated impacted teeth have been described [5].

Clinical differential diagnosis included an ameloblastoma, central giant cell granuloma, adenomatoid odontogenic tumor, and pindborg tumor. The presence of large numbers of ghost cells and dysplastic dentin in DGCT distinguish it from ameloblastoma. DGCT may be difficult to distinguish from a multicystic calcifying cystic odontogenic tumor (CCOT) [5]. Lack of cystic spaces rules out the diagnosis of the multicystic variant. Another relevant differential diagnosis for DGCT is the malignant counterpart: the ghost cell odontogenic carcinoma where numerous mitoses are seen [6-8]. The absence of mitosis eliminates the possibility of malignancy.

### Management

Surgical excision with wide histological margins (at least 0.5 cm) is advocated as the primary mode of management, with post-operative radiotherapy in selected cases [9,10], and multidisciplinary discussion prior to and following treatment.

Initially, enucleation was the primary treatment for central DGCT, but local recurrences were recognized. Hence a more radical approach is hired at the present scenario, which comprised of a segmental resection or an en bloc excision depending on the site and extent of the lesion [11]. Local recurrences can be present in COCs in general and DGCTs in particular. Central DGCTs have been found to have a high rate of recurrences after resection [12]. Recurrent cases have occurred over 5-8 years following initial treatment [13]. Distant metastasis also reported in some cases that occur in the maxilla. Consequently, the management of these cases can be nearly as same as that of some other malignant tumors of head and neck origin with baseline postoperative magnetic resonance imaging, interval imaging and long-term surveillance.

When there is a necessity for bone grafting, the tumor removal should take place first and then the graft bone should be harvested to prevent the hematogenous spreading of tumor seeds and contamination of the initial donor site [14]. DGCTs are extremely rare and only a few published cases exist to date [15,16]. So there is no specific clinical guidelines [15,17], and, the definitive diagnosis principally depends upon the histopathological analysis of the lesion due to the fact that the DGCT can resemble other odontogenic tumors both clinically and/or radiographically.

### Conclusion

A meticulous clinical and radiographic examination can be forestalled under-diagnosis of this specific entity. In time referral of the patient for biopsy and histologic analysis, surgical intervention and a long-term follow-up are crucial for the successful treatment of the patient.

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### Conflict of Interest

NIL

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