Ameloblastomatous Calcifying Odontogenic Cyst: A Case Report of a Rare Histologic Variant

Kamran Nosrati DMD*, Maryam Seyedmajidi DMD**

Calcifying odontogenic cyst is an uncommon developmental odontogenic cyst first described by Gorlin in 1962. It is considered as an extremely rare cyst and accounts for only 1% of jaw cysts reported. Because of its diverse histopathology, there has always been confusion about its nature as a cyst, neoplasm, or hamartoma. In this report, we present a rare case of calcifying odontogenic cyst with ameloblastic proliferation—an extremely rare histologic variant in a 22-year-old male in the right mandibular molar region. The lesion was surgically removed. After enucleation no recurrence has been recorded in the ensuing 14 months.

Keywords: Ameloblastic proliferation • calcifying odontogenic cyst • ghost cell • Gorlin cyst

Introduction

Calcifying odontogenic cyst (COC) was first categorized as a distinct entity by Gorlin et al., and was named after him since then. According to Shear, it accounts for 1% of jaw cysts. As the number of reports increased, it was proposed that COC was indeed a heterogeneous group of entities with distinct histopathologic findings. In this report, we present a case of ameloblastomatous COC.

Case Report

A 22-year-old male patient was referred to an oral and maxillofacial surgeon in November 2005 with a painless swelling in the right molar region of the mandible that had been present for approximately 20 days. No unusual features were noted in extraoral examination. Intraorally, there was a firm enlargement in the buccal and lingual right molar region extending from the first to the third molar. The mucosa overlying the lesion was intact. Detailed examination of the involved teeth revealed no mobility or tenderness to palpation. There were also signs of caries at the first molar. There was nothing special in his medical history. Panoramic radiography and computed tomography revealed a well-defined unilocular radiolucent lesion involving the posterior region of the mandible, extending from the first to third molar. The margins of the lesion were scalloped. The first and second right molars exhibited mild root resorption (Figure 1). The differential diagnosis included unicystic ameloblastoma and odontogenic keratocyst. The lesion was surgically removed, and involved teeth were also removed. The first histopathologic diagnosis reported by a general pathologist was ameloblastoma. The patient's

Figure 1. Panoramic radiography of the case revealed a well-defined unilocular radiolucent lesion involving the posterior region of the mandible, extending from the first to the third molars; the margins of the lesion were scalloped.
specimen was referred to an oral and maxillofacial pathologist whose diagnosis of the same biopsy was ameloblastomatous COC.

Microscopic examination revealed ghost cells in the cystic epithelium and juxtaepithelial hyalinization (Figure 2). Acanthomatous ameloblastic islands were seen in the connective tissue lining of the cyst (Figure 3). Basal cell hyperchromatism, vacuolization, and nuclear polarization, which are often seen in ameloblastoma, were absent (Figure 4).

In a postsurgical follow-up of 14 months, the patient did not manifest any recurrences (Figure 5).

Discussion

The COC is an uncommon lesion that demonstrates considerable histologic diversity and presents variable clinical behaviors. Although, it is widely considered to represent a cyst, some investigators prefer to classify it as a neoplasm. Some COCs appear to represent non-neoplastic cysts; other members of this group, variously designated as dentinogenic ghost cell tumors or epithelial odontogenic ghost cell tumors, having no cystic features, may be infiltrative or even malignant, and are regarded as neoplasms.3

In addition, the COC may be associated with other recognized odontogenic tumors, most commonly odontomas. However, adenomatoid odontogenic tumors and ameloblastoma have also been associated with COCs. Treatment and prognosis are likely to be the same as for the associated tumors.3 The WHO classification of odontogenic tumors considers the COC with all its variants as an odontogenic tumor rather than an odontogenic cyst, although it describes that further experience may provide more reliable criteria for classification of the variants.3

Praetorius et al., Hong et al., and Buchner4–6 have attempted to classify the COC based on the dualistic concept in contrast to the earlier monistic
concept. However, it seems the question concerning the nature of COC has been solved more recently by Toida who classified COC into a cyst and a neoplasm. The neoplasm is divided into benign and malignant type. The calcifying ghost cell odontogenic tumor (CGCOT) is used for benign neoplasm and it may appear to be either cystic or solid in architecture. The cystic and solid variant of CGCOT may be named ‘cystic CGCOT’ and ‘solid CGCOT’, respectively. Thus, the lesion showing cystic architecture and an extensive intramural ameloblastoma such as proliferation may be classified as the cystic CGCOT.

Ameloblastomatous COC microscopically resembles unicystic ameloblastoma except for the ghost cells and calcifications within the proliferative epithelium. Ameloblastomatous COC occurs only intraosseously. This subtype of COC is distinct from true ameloblastoma arising in COC. In contrast to ameloblastoma ex COC, the ghost cells and dystrophic calcifications are within the proliferative epithelium, which lacks histopathologic criteria as suggested by Vickers and Gorlin, and is confined to the cyst lumen.

Ameloblastoma ex COC designates an ameloblastoma arising from the cyst lining epithelium of COC (Figure 5). Our review of the literature revealed only four cases. Ameloblastoma ex COC occurs intraosseously, appearing as cyst-like, radiolucent lesions. Whether these tumors are potentially as destructive as typical ameloblastoma and have the same propensity for recurrence is unknown.

Whether ameloblastoma ex COC should be classified as a subtype of ameloblastoma or as a subtype of COC may be open to discussion. Buchner suggested that if the COC was associated with an ameloblastoma, its behavior and prognosis would be of the same as an ameloblastoma, not COC.

The classification advocated by Hong et al. has two categories for COC associated with ameloblastoma: the ameloblastomatous cystic variant and the neoplastic variant associated with ameloblastoma. The former is characterized by a unicystic structure in which the lining epithelium shows unifocal or multifocal intraluminal proliferative activity that resembles ameloblastoma, although it also contains isolated or clustered ghost cells and calcifications. The latter is called ameloblastoma arising from COC (ameloblastoma ex COC). It is characterized histopathologically as comprising few or no ghost cells with calcifications observed in the transformed ameloblastomatous epithelial portion, while the cyst lining of the epithelium contains considerable number of ghost cells and calcifications.

In the present case, although the basal cells showed ameloblastic proliferative activity, they did not completely meet the histopathologic criteria of early ameloblastoma as suggested by Vickers and Gorlin. Hence, the case has been diagnosed as ameloblastomatous COC and it has been fit into the category ‘cystic CGCOT’ as suggested by Toida.

At this present state, it is very difficult to determine whether any individual lesion having a cystic architecture is truly cystic or, in fact, neoplastic in nature. An extensive and systematic analysis of many more cases including immunohistochemical investigations on cell proliferation activity may help resolve this problem.

Ameloblastomatous COC is a rare histologic variant. We found only 13 cases of ameloblastomatous COC in the literature. Our case did not show any evidence of recurrence after treatment, but there is no doubt that careful postoperative observations are necessary for COCs which are associated with an ameloblastoma.

References

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The Portal of National Garden, Tehran/Iran
(photo by Afshin Bakhtiya, Gooya House of Culture & Art)