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CASE REPORT

INTRAOSSEOUS AND EXTRAOSSEOUS VARIANTS OF DENTINOGENIC GHOST CELL TUMOR: TWO CASE REPORTS

Kemik içi ve Periferik Dentinogenik Gölge Hücreli Tümör: İki olgu sunumu

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ABSTRACT

This paper aims to present both intraosseous and extraosseous variant of dentinogenic ghost cell tumor as well as a review of the literature. An 11-year old female patient presented a swelling and pain in the molar area of the mandible and a 15-year-old female patient reported a complaint of swelling in the right vestibular region of teeth 12 and 13(FDI 2-digit classification system). Microscopic examinations showed similar features which characterized by ameloblastoma-like islands of epithelial cells, containing numerous ghost cells. The patients have been disease-free for one year. This paper aims to describe this rare tumor and to increase the number of cases in the literature to better understand its biologic behavior and treatment options.

Keywords: *Dentinogenic ghost cell, intraosseous, extraosseous, odontogenic tumors*

ÖZ

Bu yazıda dentinogenik gölge hücreli tümörün kemik içi ve kemik dışı olmak üzere iki tipini sunmaktayız. Mandibula molar bölgede ağrı ve şişlik şikayeti olan 11 yaşında bir kız çocuğu ve maksilla 12- 13 numaralı dişlerin vestibül bölgesinde şişlik şikayeti olan 15 yaşında kız çocuğu kliniğe başvurmuştur. Mikroskopik değerlendirmede benzer görüntüler izlenmiştir. Epitelyal hücrelerin oluşturduğu yoğun gölge hücreleri içeren ameloblastik adacıklar görülmektedir. Bir yıllık kontrollerinde bir yineleme yoktur. Bu yazı ile ender görülen bu olguların literatüre kazandırılması ve biyolojik davranış ve tedavi seçeneklerinin daha net anlaşılması için amaçlanmıştır.

Anahtar kelimeler: *Dentinogenik gölge hücreli, kemik içi, kemik dışı, odontojenik tümörler*

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Introduction

The calcifying odontogenic cysts was defined as a cystic lesion in the 1992 classification which the epithelial lining shows a well defined basal layer of columnar cells, an overlying layer that may resemble satellite reticulum, and masses of ghost epithelial cells that may be in the epithelial lining or in the fibrous capsule. The ghost cell may become calcified. Dysplastic dentin may be laid down adjacent to the basal layer of the epithelium (1).

In 2005, WHO classification of odontogenic tumors, calcifying odontogenic cysts were renamed and divided into 2 subtypes. The first one is the calcifying cystic odontogenic tumor; a benign cystic tumor of odontogenic origin characterized by an ameloblastoma-like epithelial component with ghost cells that may calcify. Dysplastic dentin may be seen in the adjacent connective tissue. The other one is the dentinogenic ghost cell tumor (DGCT); 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 associate with varying amounts of dysplastic dentin (2).

DGCT represents in intraosseous and less commonly in extraosseous variant. The intraosseous type has an aggressive behavior and more aggressive local resection is recommended to avoid recurrence. Extraosseous variant is less aggressive and can be controlled by local excision. No recurrences have been reported (2-4). On rare occasions, the lesion transforms into carcinoma (5, 6).

In this report, the nature, radiographic, and histopathological features of DGCT are discussed along with a review of literatures. This paper will also provide to increase the number of cases of this rare tumor in the literature.

Case 1

An 11-year old female patient presented a swelling and pain in right molar side of the mandible. The swelling extended from first premolar to the retromolar trigon involving the anterior the anterior side of the ramus with expansion of the buccal and lingual cortical bone.

The patient had a history of incisional biopsy with a tentative diagnosis of odontogenic tumor. Clinically, the provisional diagnosis of odontogenic myxoma was made. Radiographically, the lesion showed unilocular, well-defined radiolucent lesion associated with two teeth germs (Figure 1). The lesion was treated by conservative curettage and sent for histopathological examination.

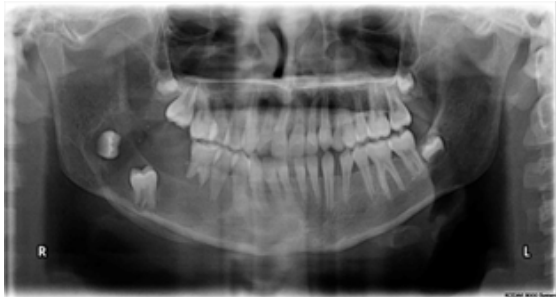

Figure 1. *Panoramic radiograph showing well-defined, unilocular radiolusent lesion with tooth germs.*

Histopathological examination revealed the ameloblastoma-like epithelial islands which were formed by peripheral palisading of columnar cells and centrally simulating the stellate reticulum (Figure 2).

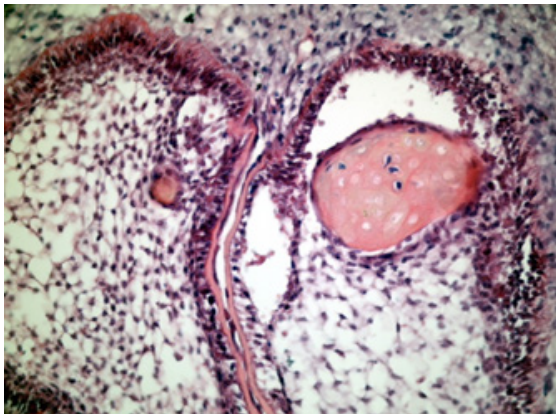

Figure 2. *Characteristic ameloblastic island of odontogenic epithelium with peripheral columnar cells and central stellate reticulum like cells (H&E x40).*

Clusters of ghost cells were seen within the ameloblastomatous epithelial component in a mature connective tissue stroma (Figure 3).

The dentinoid was seen juxtaposed to the odontogenic epithelium, and it was calcified into bone-like tissue (Figure 4).

No atypical mitoses, pleomorphism and necrosis were observed. Conventional keratin staining clearly differentiated the ghost cells from dentinoid which the ghost cells taking a red color and the dentinoid and osteodentin taking blue color (Figure 5).

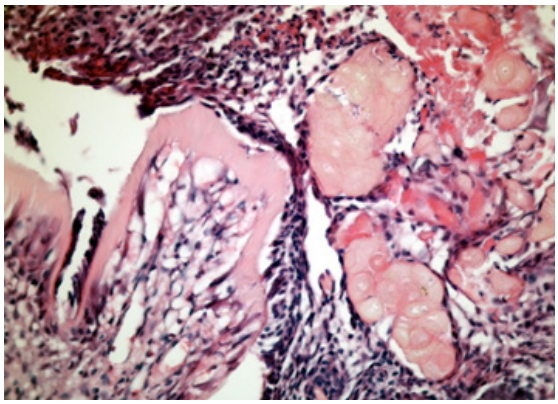

Figure 3. *Clusters of ghost cell were seen within ameloblastic odontogenic epitehelium, central area showing stellate reticulum like cells transforming into ghost cell (H&E x40).*

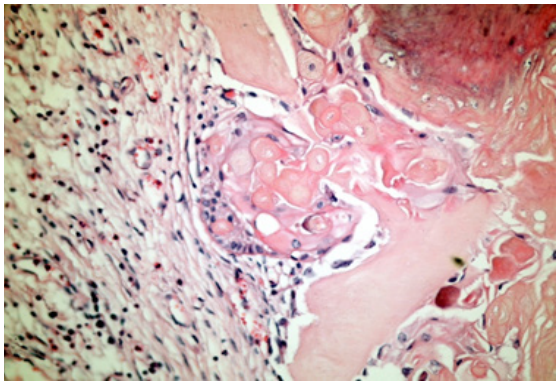

Figure 4. *Areas of dentinoid with aggregates of ghost cells in the connective tissue (H&E x40).*

Also CK 1-3 was performed immunhistochemically and the odontogenic epithelium showed positive staining. Finally, the diagnosis of central variant DGCT was made and six months of follow-up has not shown any signs of recurrence. The patient is under follow-up with panoramic radiograph and clinical examination every 6 months.

Case 2

A 15-year-old female patient reported a complaint of swelling in the right vestibular region of teeth 12 and 13. The swelling was firm and painless, covered by partly ulcerated oral mucosa measuring about 3 mm in diameter in the vestibular gingival between teeth 12 and 13. Clinical diagnosis was irritation fibroma. Radiological examination showed no feature. The lesion was surgically removed under local anesthesia, and the tissue was sent for histopathological examination. The histopathological examination revealed a solid, well-circumscribed and encapsulated soft tissue mass in a fibrous connective tissue covered by ulcerated oral mucosa (Figure 6).

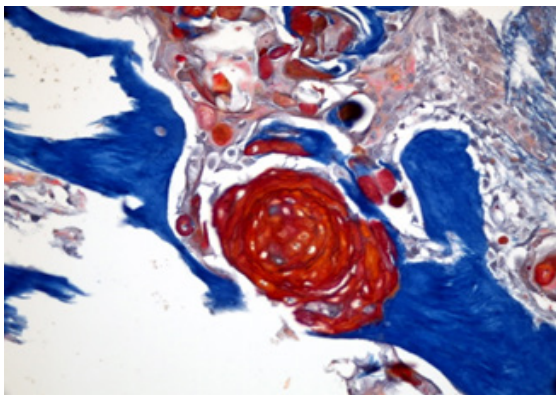

Figure 5. *Keratin stain showing ghost cells staining red and surrounding dentinoid-like material blue (H&E x40).*

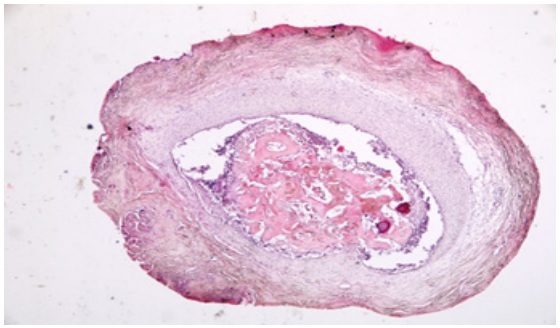

Figure 6. *Solid well-circumscribed mass in the connective tissue showing odontogenic epithelium with ghost cells. (H&E x4).*

The tumor mass closely resembled ameloblastoma-like areas of odontogenic epithelial cells with ghost cells showing keratinization and small calcification (Figure 7).

The diagnosis of peripheral variant DGCT was made. The patient has been disease-free for 7 months.

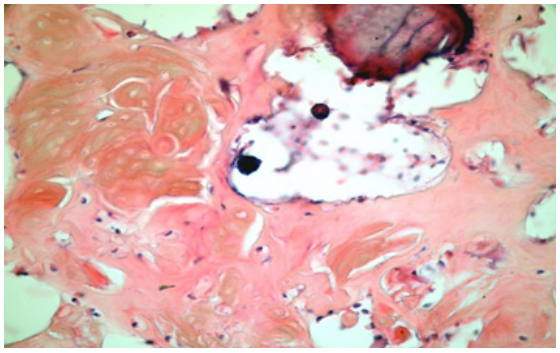

Figure 7. *Abundant ghost cells with dentinoid-like calcification (H&E x40).*

Discussion

DGCT occurs as an intraosseous and less commonly as an extraosseous variant. There is no difference between the histological features of the intraosseous and extraosseous DGCT that the tumor composes of sheets and rounded ameloblastoma-like islands of odontogenic epithelium with clusters of ghost cells and variable quantities of dentinoid material in the surrounding connective tissue and near the odontogenic epithelium (2, 3). Diagnostic histopathological features for DGCT are described clearly and the present cases were all in agreement with previous literatures.

The extraosseous DGCT is apparently rare so it is difficult to typify the age, sex and location distribution. Though according to existing literature knowledge it appears to be slightly more common in the mandible than the maxilla, with a predilection for the elderly age group (7, 8). In our case, the relatively different feature was patient age, 15-year-old. The mean age of the previously reported cases was 59 years old (7).

Generally the presentation of extraosseous type is a nodular swelling on the alveolar mucosa implicating trauma or irritation as an etiologic factor of the tumor. Clinically this feature may mimic the epulis as well as our case (7-9). Radiographically the present case was not shown any saucerization of the underlying bone but this radiological feature has been observed in about 20% of cases (2, 7). The preferred treatment for extraosseous variant is conservative excision and no recurrence has been reported that these findings are compatible with our case.

On reviewing to literatures about extraosseous variant of DGCT, it was observed that the age range is from 12 to 75, with the mean age of 40.72 and it is slightly more common in males than females (10, 11). There is no preference for maxilla and mandible. The present case was 11 years old female patient, which is much younger age than the average age of DGCT and the lesion was seen extending from the right first premolar to the angel of the mandible. These clinical findings were similar with Juneja *et al.* (10). On radiologic examination, DGCT generally shows well-circumscribed, unilocular radiolusent to mixed radiolucent/radiopaque appearance depending on the amount of calcification (2). In this case, the lesion was observed well-demarcated, unilocular radiolucent area without teeth resorption and distinct radiopaque feature. The different finding was to be associated with two teeth germs. The intraosseous type is more aggressive from extraosseous type so local resection is recommended by many authors (4, 5, 12). Sun *et al.* (4) observed that DGCT has a high rate of local recurrence after conservative curettage and enucleation. The present intraosseous case was treated by conservative surgery especially because of the patient age that the jaw is still growing. Therefore the patient was under close observation since six months and no recurrence has been noted up to today. Long-term follow-up has been planned.

Many of extraosseous variant of DGCT can be mistakenly diagnosed as peripheral ameloblastoma as well as intraosseous variant can be confused with ameloblastoma. This special odontogenic tumor is distinguished from ameloblastoma by the presence of large numbers of ghost cells and dysplastic dentin. DGCT may be difficult to distinguish from multicystic calcifying cystic odontogenic tumor (5, 7, 8). In our case the cystic spaces were not observed and that diagnose was ruled out. Another differential diagnose should be made with odontogenic ghost cell carcinoma. Lack of necrosis, atypical mitosis and pleomorphism eliminated the malignant tumor in this case. There are few studies about the genotypic-phenotypic characteristics of DGCT in the literature. One of them was made by Kim *et al.* (13) and they suggested that β-catenin has played an important role in tumorigenesis of DGCT.

We have presented the extraosseous and intraosseous variants of DGCT cases by comparing them with previous literature. This extremely rare odontogenic tumor needs more research with large series to better understand its biologic behavior and treatment options. It would be useful to evaluate the

treatment type based on each case to collect sufficient data in the future.

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Conflict of interest
None declared

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