Case Report

Calcifiying Odontogenic Cyst: Report of a Case

Kalsifiye Odontojenik Kist: Olgu Sunumu

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ÖZET

Kalsifiye odontojenik kist, nadir görülen gelişimsel odontojenik bir kisttir. Bu lezyonlar, gömülü dişlerle, odontoma, ameloblastoma, adenomatoid odontojenik tümör gibi odontojenik tümörlerle veya bazı vakalarda diğer odontojenik kistlerle birliktelik gösterebilir. Her yaşta görülebilse de en sık 2.dekatta görülmektedir. Genellikle lokal eksizyonla tedavi edilmekle birlikte, büyük lezyonlarda dekompresyon veya marsüpyalizasyon tercih edilebilir. Bu kistlerin uzun dönem takibi önerilmektedir. Bu çalışmada, 16 yaşında erkek hastada, gömülü mandibular kanin diş ve persiste süt dişi ile beraber ortaya çıkan; normalde görülenden daha küçük boyutlarda izlenen bir kalsifiye odontojenik kist vakasının sunulması amaçlanmıştır.

Anahtar Kelimeler: Kalsifiye odontojenik kist; gömülü diş; persiste süt dişi

ABSTRACT

Calcifying odontogenic cyst (COC) is a rare odontogenic cyst with a developmental origin. COC can be associated with unerupted teeth, odontomas, ameloblastomas, adenomatoid odontogenic tumors, and in some cases with other odontogenic cysts. It occurs over a wide age range particularly common in 2nd decade of life. Generally, the treatment method for COC is local excision. For extensive lesions, decompression or marsupialisation can be also used. Long term follow-up is recommended in COC cases. Here, we present a clinical case of COC associated with an impacted mandibular canine tooth and a persistent deciduous tooth and of a smaller size than is normally encountered in a 16-years old male patient.

Keywords: Calcifying odontogenic cyst; impacted tooth; persistent tooth

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INTRODUCTION

The calcifying odontogenic cyst (COC) is an uncommon cyst with a developmental origin which was described by Gorlin et al in 1962.[1] The lesion was accepted as the oral analogue of pilomatrixoma of the skin (the cutaneous calcifying epithelioma of Malherbe) which is a cystic lesion lined with ameloblastomatous epithelium containing focal accumulations of ghost cells.[2] Since COC is a member of the family of ghost cell lesions, World Health Organization (WHO) considered it as a benign cystic tumour having odontogenic epithelium with odontogenic ectomesenchyme with/without dental hard tissue formation in 2005.[3] Finally, in 2017, it has been reclassified in the group of odontogenic cysts.[2]

COC is an uncommon lesion and represents approximately 1-7% of all odontogenic lesions.[4,5] Clinically, it occurs over a wide patient age range, and it peaks in the second decade of life.[6] There is no preledictionin gender distribution.[7] It usually occurs in anterior parts of jaws.[7] Radiographically, it shows unicystic or multicystic well-bordered radiolucent appearance and it can also contain irregular radiopaque foci.[2] COC is characterized by cystic proliferation of the odontogenic epithelium, some are solid and present mixed histological features. The epithelial lining has characteristic features with a basal cell layer consisting of palisaded columnar or cuboidal cells.[7] The overlying layer can loosely be arranged and may resemble as stellate reticulum like ameloblastoma. In the fibrous connective tissue wall, there are usually islands of odontogenic epithelium.[7] The cystic variant of COC is characterized by a unicystic lesion associated with or without odontoma and are in majority.[7] In the literature, COC's were reported with ameloblastomas, adenomatoid odontogenic tumours, dentigerous cysts, odontogenic keratocyst and ameloblastic fibroma.[8] In general, lesions' size is between 2-4 cm in diameter, but it has been reported that there are also large lesions reaching 9-12 cm.[6] Generally applied method for COC is conservative treatment with local excision.[9]

Herein, we present a case of small size COC associated with an unerupted deciduous tooth. Although cases with unerupted teeth are usually associated with odontoma, the uniqueness of our case comes from the size of the lesion and with impacted tooth and dental follicle association.

CASE REPORT

A sixteen-year-old male patient without any remarkable medical history was referred by a general dentist to the Department of Maxillofacial Surgery, Faculty of Dentistry, Gazi University with the finding of a radiolucent lesion with an impacted tooth determined in orthopantomography (OPG) (Figure 1).

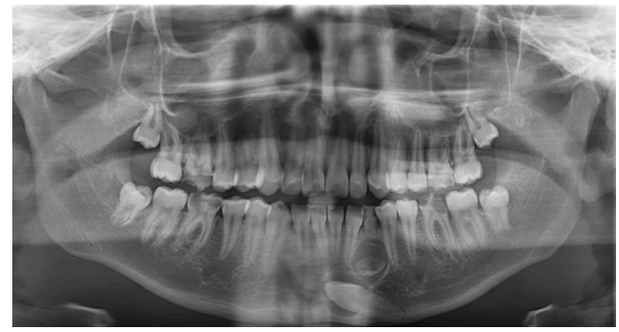

Figure 1. Preoperative orthopantomograph.

The intraoral and extraoral examinations were within normal limits but the minor decay on the persistent tooth. The patient had no clinical symptoms such as pain or paresthesia.

The orthopantomography revealed a unilocular, well-defined radiolucency between the persistent mandibular canine tooth and mandibular left first premolar tooth in connection with cervical part of the impacted left mandibular canine tooth. Root resorption was present with the association of the lesion. There was also a well-defined radiolucency around the crown of the persistent mandibular canine tooth (Figure 1). In 3D radiographic examination (Figure 2) an expansive, well-bordered, 10x10x16 mm in dimensions, a radiolucent lesion which lays between roots of the mandibular left central tooth and left first premolar tooth was observed. After having a consultation with the Department of Orthodontics, at the same institution, it was decided to extract the impacted tooth due to the position of the tooth, and surgical removal of the lesion.

The lesion was enucleated and the impacted tooth and persistent deciduous tooth were extracted under the general anaesthesia. A full-thickness mucoperiosteal flap was elevated with sulcular incision in association with vertical incisions and bone was removed with a

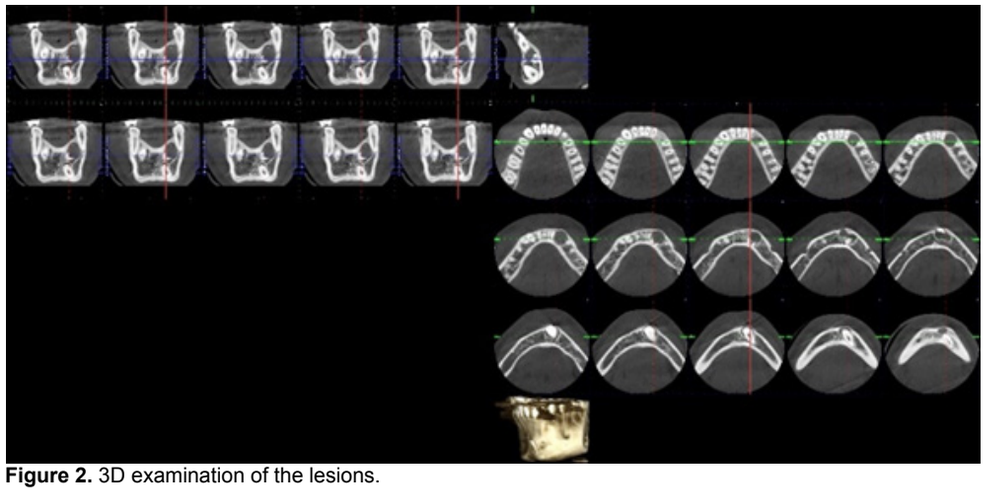

Figure 2. 3D examination of the lesions.

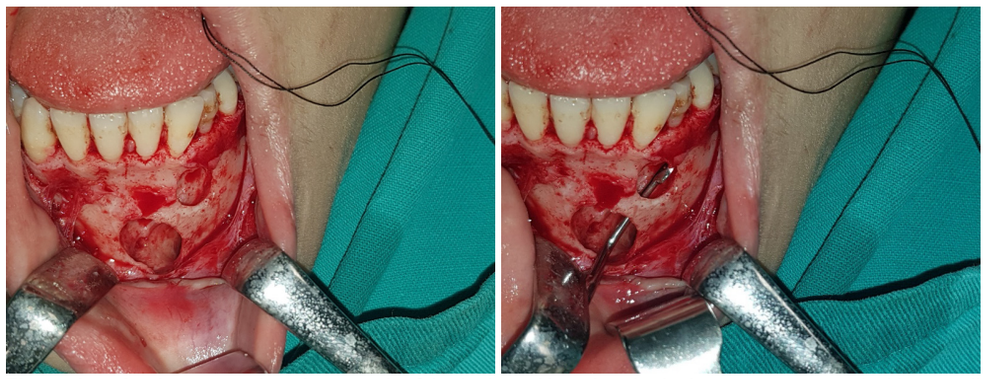

Figure 3 and 4. Intraoperative photograph of the lesions. Note that the lesions are connected with a small perforation.

bur. The lesion was divided into two sections with a wall and these sections were connected with a small perforation (Figures 3 and 4). Excised lesions and extracted teeth were fixed in formalin (10% buffered) and sent to the Department of Oral Pathology, at the same institution, for histopathological examination.

Macroscopically, the surgical specimen has consisted of two excised soft tissue materials. The first one was an opened cystic sac attached to the apex of a deciduous tooth with 0.6X0.6X0.5 cm in size, dark brown in colour. The other material was a piece of irregular soft tissue, brown in colour, 1.3X1.1X0.5 cm in size, sent with attached to the crown of the mandibular canine tooth.

Histologic examination of the hematoxylin-eosin stained sections demonstrated a defined cystic lesion with a fibrous capsule and a lining of odontogenic epithelium. The basal cells of the lining epithelium were mainly columnar or cubic cells and the overlying layers were loosely arranged, like as stellate reticulum cells (Figure 5A).

In the parts of cystic epithelial lining towards to lumen, there were pink, hyaline deposits without a nucleus (ghost cells) and containing an amorphous material with eosinophilic foci of calcification (Figure 5B). Plenty of odontogenic epithelial rests were observed in the connective tissue wall of the cyst. Alcian Blue PAS (pH:2,5) and Van Gieson histochemical stainings highlighted and differentiated the ghost cells (Figures 5C and 5D). The final diagnosis established as calcifying odontogenic cyst.

The other specimen was a soft tissue with fibromyxoid structure without an overlying epithelium. Mild mononuclear inflammatory cell infiltration and odontogenic epithelial rests were observed in the connective tissue ground. Since no cystic epithelial lining was found in the material examined in serial sections, the final diagnosis was established as dental follicle.

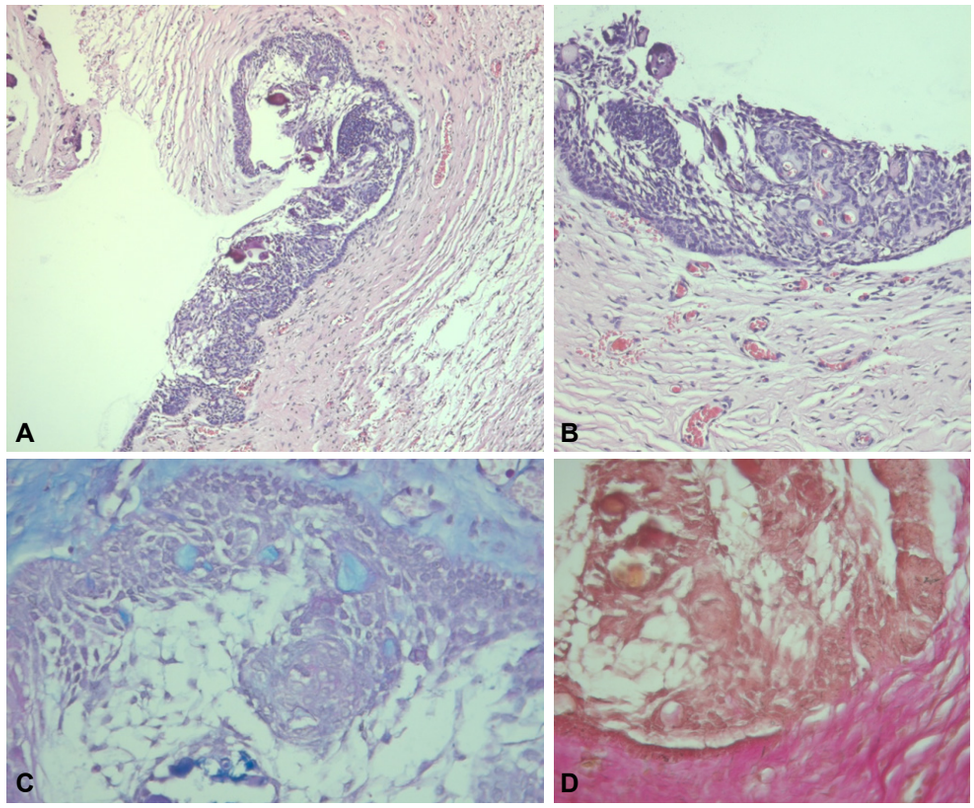

Figure 5.
A. The epithelial lining of the cyst (X10, Hematoxylin&eosin)
B. The cystic lesion consists of epithelial cells undergo ghost cell change in suprabasilar epithelium and calcifications (ghost cells) (X20, Hematoxylin&eosin)
C. Ghost cells in the ameloblastomatous epithelium which are positive with Alcain Blue PAS staining (X40, AB-PAS (pH:2,5))
D. Van Gieson staining reveals the presence of the ghost cells (X40, Van Gieson)

DISCUSSION

COC is a rare odontogenic lesion which is a cystic lesion lined with ameloblastomatous epithelium containing focal accumulations of ghost cells.[2] This lesion is first described as a distinct entity by Gorlin *et al.*[1] in 1962. From that time, there have been many changes in its name and classifications regarding the origin of the COC. This much change in terminology and classification is due to the unclear points in the pathogenesis of COC. Histologically, the most important feature of COC is that ghost cells can be observed throughout the epithelial lining.[14] Ghost cells are pink anucleated cells with homogeneous eosinophilic cytoplasm and are considered to be abnormally keratinized cells, and they are characteristically seen in COC, pilomatrixomas and craniopharyngiomas. They can be seen in other odontogenic lesions, dentinogenic ghost cell tumours and ghost cell odontogenic carcinomas.[14] These cells are also thought to be transformed odontogenic epithelial cells.[15,16] In a study conducted on COC[17], it was shown that the immunohistochemical staining pattern of COCs was more similar to odontogenic lesions than calcifying epitheliomas of Malherbe, and it was emphasized that ghost cells were aberrantly keratinized odontogenic cells.

Although this lesion is reclassified as a cyst, recent studies have revealed that the formation of COC is caused by genetic alterations.[18,19] One recent study suggested that COC is not cysts, but neoplastic lesions genetically similar to pilomatrixoma and craniopharyngiomas, due to a common driver mutation in the CTNNB1 gene in all these lesions that cause ghost cells to formation.[16] For all these reasons, there are still discussions on whether COC is a cyst or a tumour. In the histologic examination, if the lesion has a cystic structure and the connective tissue wall does not contain ameloblastoma-like

proliferation, it is classified as COC. Dentinogenic ghost cell tumour should be considered in the cases with a neoplastic growth pattern.[6]

COC usually occurs in association with odontoma. In approximately one of the third cases of COC with associated with unerupted tooth[10] and it is reported to be associated with odontoma in 24% of cases.[11] It can be seen that in COC cases associated with odontoma that cause a delay in the eruption of deciduous teeth.[12,13] In the present case, the lesion was associated with persistent tooth not with an odontoma and connected to the follicle of the impacted canine with a perforation. Although the association of impacted tooth with COC is very common, both impacted tooth and the persistent tooth apices relation with the COC has not been widely reported.

Most of calcifying odontogenic cysts are between 20 and 40 mm in diameter. De Arruda *et al.*[4] found that lesions appeared with an average of 32 mm in 20 cases they evaluated in their 26 years retrospective study. In our case dimensions were 10X10X16 mm, smaller in size than usual. This may be because the lesion was detected in early-stage; showing the importance of routine dental examination not only for dental caries or periodontal problems but also to detect benign or malign lesions in early stage in small diameters for better prognosis.

Treatment of COC is generally simple enucleation. Early detection of lesions in small dimensions can cause better morbidity with less effection of adjacent anatomical structures. For extensive lesions, decompression or marsupialisation can be used for treatment.[20] Emam *et al.*[20] applied decompression therapy in their case and concluded that the treatment was successful after 9 months of follow-up. De Arruda *et al.* stated in their review that these lesions are usually treated with enucleation (69.9%); that 2 lesions (0.7%) were treated with marsupialisation.[6] In this case, the lesion was surgically enucleated and the related tooth was extracted under general anaesthesia owing to fracture risk of the mandible and patient comfort. As a result of the consultation with the Department of Orthodontics, it was evaluated that the relevant tooth could not be erupted. Therefore, it is decided to excise the lesion during the tooth extraction procedure instead of marsupialisation treatment. Long-term follow-up is suggested for every COC case, but the recurrence rates of COC vary, as there are treatment methods and follow-up procedures that are not fully standardized.[3-9]

To summarize, although COC is a rare lesion, it is a lesion that should be kept in mind, especially in lesions that have a radiolucent appearance in the presence of persistent deciduous teeth and impacted teeth. Additional case reports and more detailed studies may contribute to resolving unclear points about the nomenclature and the pathogenesis of this cyst.

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