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Calcifying Odontogenic Cyst: Report of Two Cases Report in a Different Stage of Evolution

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Abstract **Case Report**

Odontogenic tumors with calcified components contribute substantially to the diversity in the field of oral surgery. Among these, the Calcifying Odontogenic Cyst (COC), also known as Gorlin's cyst, stands out. It is a rare odontogenic lesion, first described by Gorlin in 1962. It has a vast variety of radiological, clinical, biological behaviors and histopathological features. In this article, two cases of Calcifying Odontogenic Cyst (COC) treated successfully with surgical interventions at oral Medicine and Oral Surgery department at the academic dental Clinic of Monastir, in order to underline clinical, radiological and histological features of this cyst and highlights the interest of detailed complementary investigations particularly anatomopathological ones to precisely define the diagnosis.

Keywords: Gorlin's Cyst, Ghost cell, Odontogenic cyst, Enucleation, Benign.

INTRODUCTION

Calcifying odontogenic cyst (COC) or Gorlin cyst is a benign developmental cyst. It is a rare lesion first recognized as a distinct clinicopathological entity by Gorlin in 1962.

The classification of this lesion is still subject to debate. In 1992 and 2005, it was classified by the World Health Organization (WHO) as odontogenic tumors. In 2017, COC was classified among the developmental cysts [2, 3]. In its latest classification in 2022 [5], the 5th edition, COC has continued in the cyst classification with an important change in the definition that also affects the diagnostic criteria [3].

In the definition of the 2017 classification, 'ameloblastoma-like epithelium' was excluded and COC is now defined as "a developmental odontogenic cyst characterized histologically by ghost cells, which often calcify." While most COCs still have ameloblastoma-like epithelium, that feature was moved from an essential feature to a desired one [5].

It is estimated that COC accounts for 1-2% of all odontogenic cysts of the jaw with a predominance at the anterior maxilla in patients of the second-third decade [1-3, 6].

The instability of classification and the rarity of this cyst can make a diagnostic challenge especially in standard radiographs or at advanced stage where calcifications, which are pathognomonic features making diagnosis easier, can be visualized only at histological examination. Hence the aim of this work, which was to report two cases of calcifying odontogenic cyst where calcifications were revealed only in the 3 D x-rays for the first case and in the histological examination for the second one, in order to highlights it's clinical, radiological and histological features and detail the therapeutic approach.

CASE PRESENTATION

Case 1:

A 22-year-old male patient was referred to the Department of Oral Medicine and Oral Surgery for a fistula in the right anterior maxillary sector. His familial and past medical history were noncontributory.

The extraoral examination was normal. There was no history of paresthesia, pain or lymphadenopathy.

The intraoral examination showed a slight swelling in the area extending from the tooth11 to the tooth13, with a fistula opposite 12. The swelling is covered by a normal mucosa and was fluctuating and slightly tender on palpation (Fig 1, 2). The vitality test carried out at the tooth12 was negative.

The panoramic radiograph revealed a mixed well-defined radiolucent-radiopaque image in the region of the fractured apex of the tooth12 and the apex of the tooth13 (Fig 3).

To precisely defined the lesion limits, a cone beam computed tomography was required and it showed a blowout of the outer wall with fenestration opposite the apex of the tooth13 and no invasion of the nasal cavity (Fig 4).

Based on the clinical and radiographic examinations, provisional diagnoses were suggested: Calcifying odontogenic cyst, Calcifying epithelial odontogenic tumor and an inflammatory periapical cyst with osseous reaction in relation with the tooth12.

Patient underwent first an endodontic treatment of the tooth 12 (Fig 5), then the lesion was enucleated under local anesthesia (Fig 6 -10).

Histological examination confirmed the diagnosis of calcifying odontogenic cyst (Fig 11). The patient was recalled periodically for regular follow up which was uneventful (Fig 12).

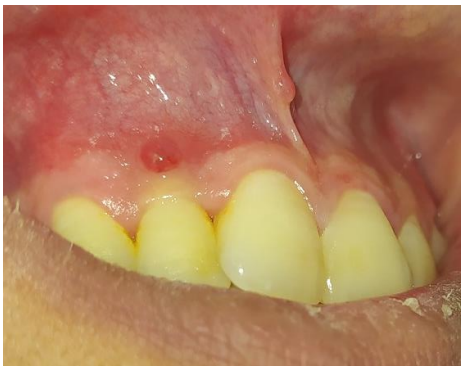

Figure 1: Slight vestibular swelling in the anterior right region + fistula adjacent to tooth 12

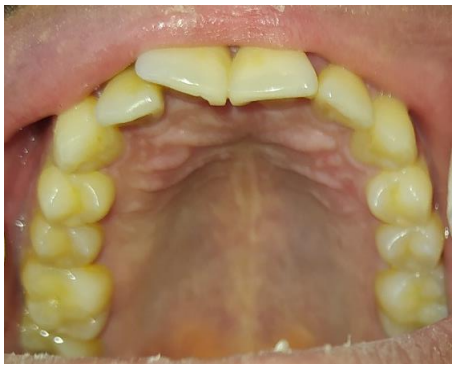

Figure 2: Occlusal view: Absence of palatal swelling

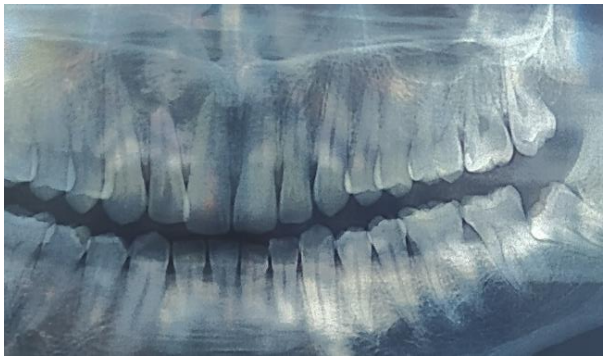

Figure 3: Panoramic radiographic showing a well-defined mixed radiolucent/radiopaque image in the maxillary anterolateral area

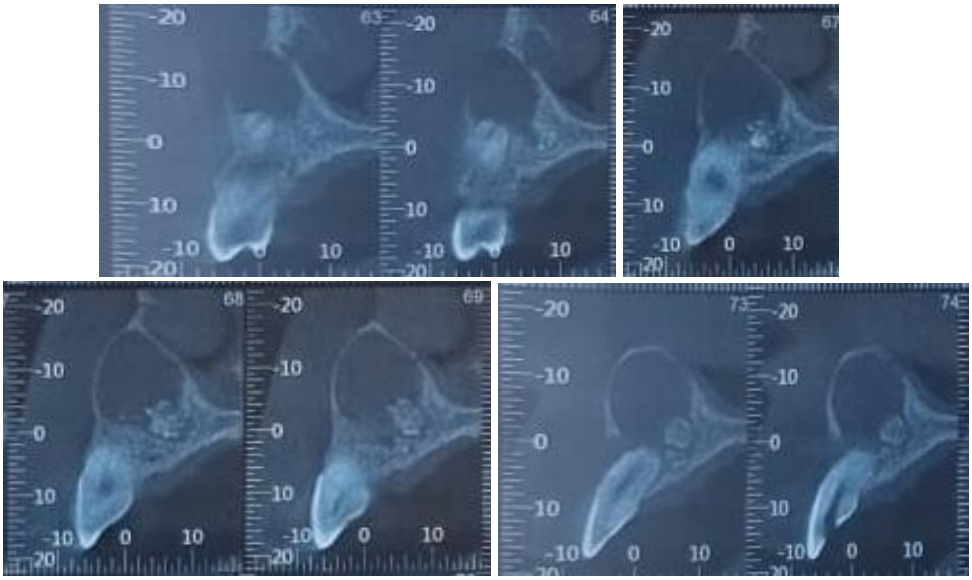

Figure 4: CT scan showing an image of a lytic lesion pushing back the buccal wall with the presence of central hyperdensities

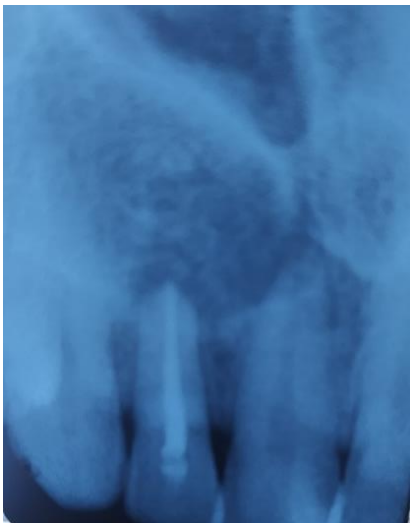

Figure 5: Hermetic canal obturation of the tooth 12

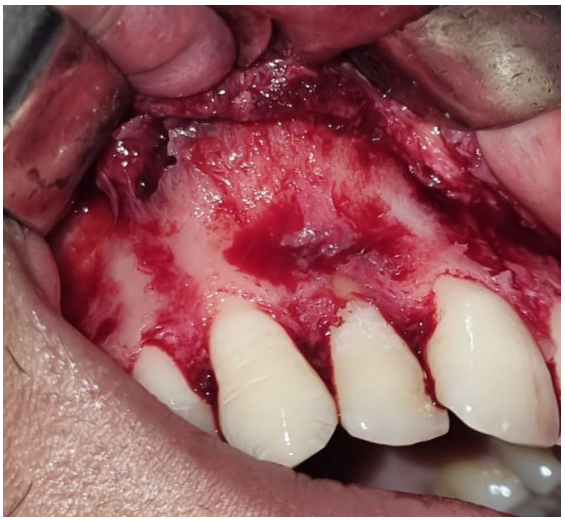

Figure 6: Intraoperative image demonstrating buccal cortical bone obliteration and cyst lining

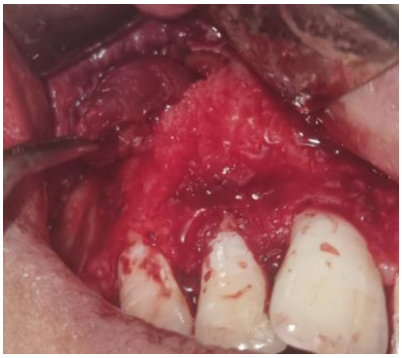

Figure 7: Enucleation of the cystic wall

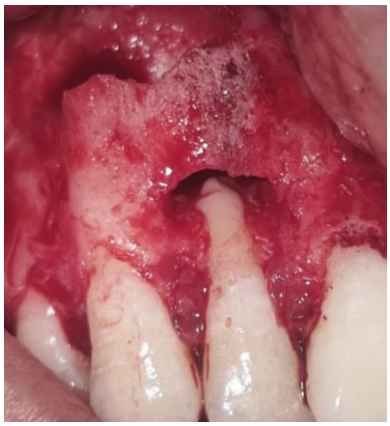

Figure 8: Root resection of the tooth 12

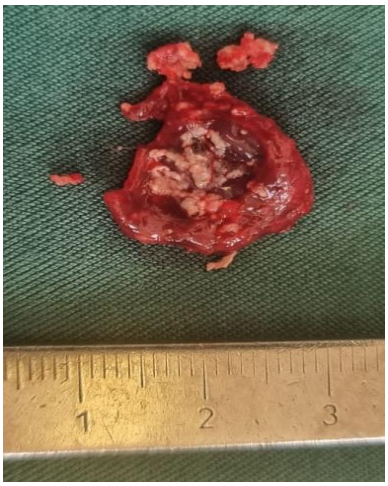

Figure 9: Image of exicisional specimen

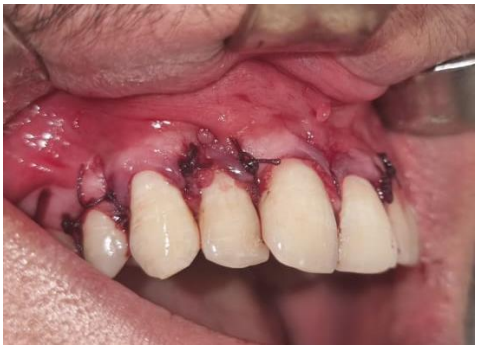

Figure 10: Sutures

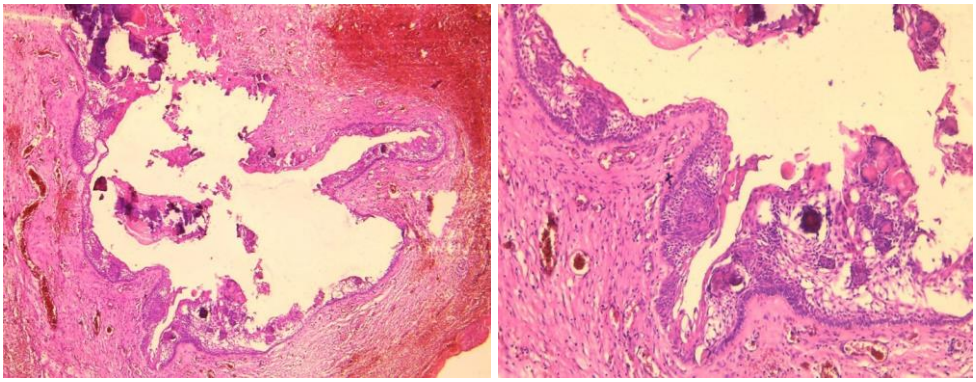

Figure 11: Histopathological view: Ameloblastomatous cyst lining showing gost cells and calcifications. Cyst wall consists of non inflammatory conjonctive tissue (A:HEx40, B:HEx100, C:HEx100)

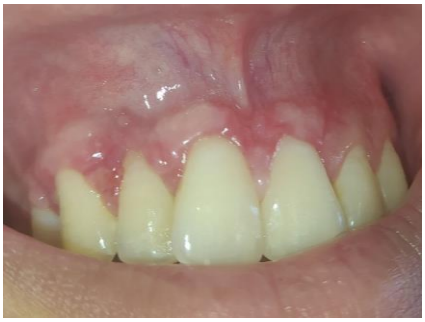

Figure 12: Follow-up, 10 days later

Case 2:

A 33-year-old male patient was referred by his general dentist for a swelling in the anterior mandibular left region of 6 months duration. His medical and past medical history was noncontributory.

The extraoral examination was normal. There was no history of trauma, pain or paresthesia. On intraoral examination, there was a slight swelling in the area extending from the tooth 42 to the tooth 35. The swelling was covered by a normal mucosa, it was hard and painless on palpation. The cold tests carried out at all teeth of the lesion's area were positive.

A panoramic radiograph revealed a large well-defined periapical multilocular radiolucency of the anterior mandibular region extending from #42 to #35 (Fig 13).

A Cone beam computed tomography (CBCT) was required and showed a well-defined multilocular hypodense lesion and destruction of the vestibular cortical (Fig 14).

Based on the history, clinical and radiographic examination, provisional diagnosis were suggested: Solitary bone cyst, odontogenic keratocyst, Ameloblastoma…

Surgical enucleation of the lesion was performed (Fig 15). Histological examination concluded the diagnosis of calcifying odontogenic cyst (Fig 17). Patient was recalled for continued follow up which was favorable.

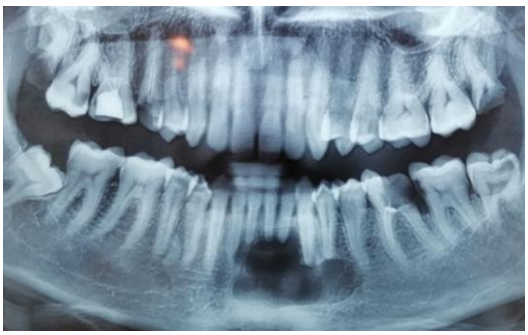

Figure 13: Orthopantomograph revealing a well-defined multilocular radiolucent lesion extending from #42 to #35 region

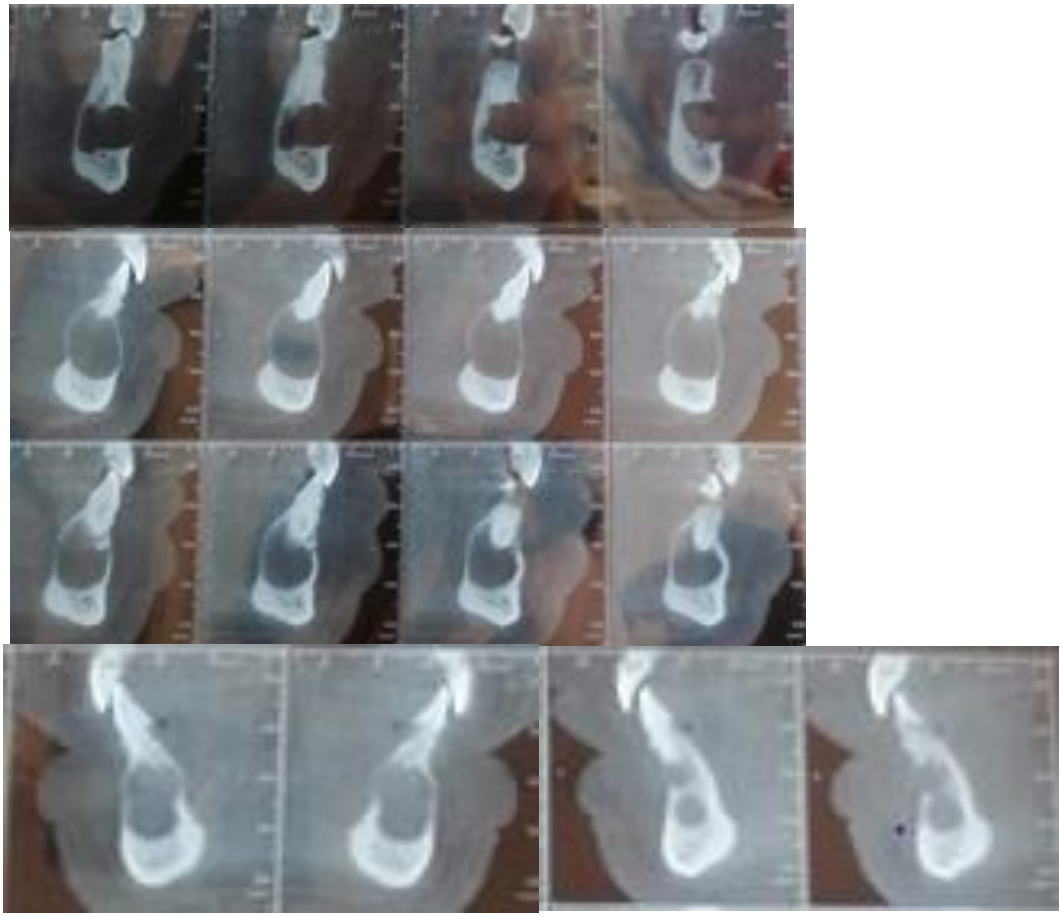

Figure 14: Cone beam computed tomography (CBCT) showed a well-defined multilocular hypodensity and destruction of the vestibular boundary

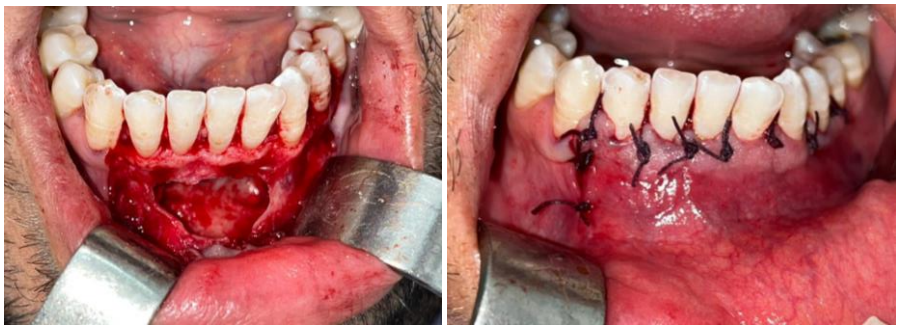

Figure 15: Surgical enucleation of the lesion

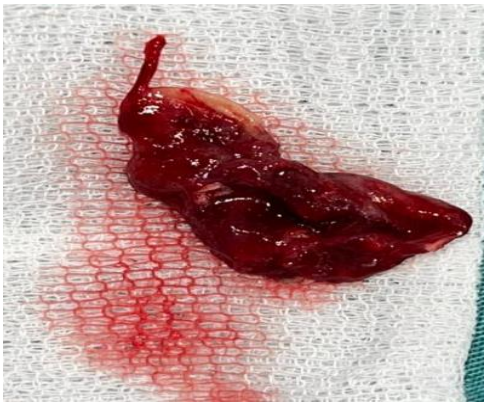

Figure 16: Specimen

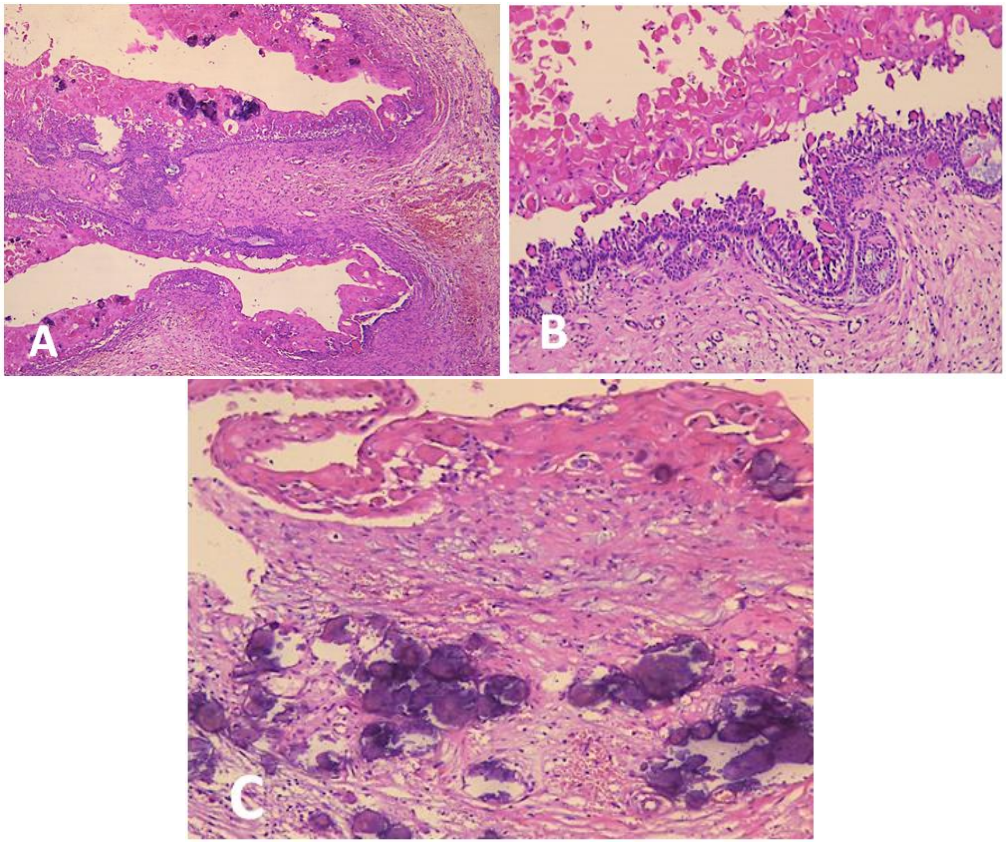

Figure 17: A: Histopathology photomicrograph showing cyst wall consists of fibrous tissue and ameloblastoma-like cyst lining with ghost cells and calcifications (H and E stain, ×40). B: Photomicrograph demonstrationg the features of the superficial ghost cells with loss of the nucleus, and the features of the palisaded ameloblastoma-like cells and adjacent stellate reticulum-like area (H and E stain, ×100). C: Photomicrograph showing dystrophic calcifications within cyst wall (H and E stain, ×100)

DISCUSSION

In 1962, COC was firstly described by Gorlin. It's an uncommon lesion in the oral cavity. It is defined as a cystic cavity lined by an ameloblastoma-like epithelium containing focal ghost cells and calcifications [2, 3]. Focal ghost cells are also present in dentinogenic tumors. This diversity led to a confusion in the classification and the terminology of this lesion.

In its latest classification in 2022, COC is defined as "a developmental odontogenic cyst" [5].

The COC can arise at any site in the oral cavity with case occurring most frequently in the anterior maxilla, which corresponds to the reported case where the lesion was located in the maxillary canin region, then it's followed by the posterior mandible [3].

According to the study by Arruda *et al*., COC represents 1.3% of odontogenic cysts, with a female predilection (53%) in women and 47% in men [6]. With a mean age between 20 to 59 years accounted for 47.3%, Children and adolescents (0–19 years) accounted for 35.1% of the sample [2, 7].

The most common clinical sign noted in patients with COC is an asymptomatic swelling in the involved region in both extraosseous and intraosseous locations.

For the lesions with a small size, they are usually painless. Often, an incidental finding revealed on radiographic examination. Sometimes patients may complain of headache, epistaxis and nasal stiffness when it's located in the maxilla [2].

Radiographically, they appear as unilocular or multilocular radiolucency with a well-defined margin and contains calcifications of varying density. The presence of calcification is an important radiographic feature in the interpretation of COC [2, 11].

However, the second case did not show radiopacities as evidence of calcification with the conventional radiography as was suggested by McGowan and Browne [9, 13] who also found that the presence of mineralization was approximately twice as frequently seen on microscopic examination compared to radiographic analysis.

Actually, calcifications may be absent at radiographic examinations at an early stage of the cyst,

hence calcifications are revealed only at histological exploration. This may distort the diagnosis as it is the second case reported.

In the most of time, COC is situated in the periapical or lateral periodontal area of the dentition. Consequently, teeth divergence and root resorption are frequent radiographic findings. According to the study of Uchiyama *et al*., teeth divergence was seen in eight of nine cases and root resorption in seven of nine cases. Association with an impacted tooth is found approximately in six of nine cases. This underscores the critical importance of conducting routine histopathological analysis on all tissues associated with the crowns of impacted teeth, even in the absence of radiographic suspicion of follicular lesions [2, 8].

The CT scan is used to determine the lesion extension and its relationship with anatomical structures.

Also, CBCT offers higher sensitivity in detecting calcifications compared to routine panoramic radiograph [8, 9].

Due to the diverse radiographic presentation of COC, the differential diagnosis of COC is made with all mixed lesions of the maxilla. It includes benign radiolucent lesions, such as calcifying epithelial odontogenic tumor, dentigerous cyst, adenomatoid odontogenic tumor, ameloblastic fibro-odontoma, ossifying fibroma and a partially mineralized odontoma.

Classical histopathological examination findings in COC are an odontogenic epithelium with ghost cells and calcification. The unique feature of COC is ghost cell keratinization which differentiates it from an ameloblastoma. However, the synchronous occurrence of COC with ameloblastomas and other odontogenic pathologies including dentigerous cysts, ameloblastic fibromas and odontomas has been reported [3].

Histological evaluation of the second case revealed calcifications that were not visible on radiographic examination, likely due to their immaturity [10, 12].

The recommended treatment for COC is surgical excision for the extraosseous presentation and enucleation with curettage for the intraosseous presentation, which means enucleation followed by removal of a 1 to 2mm layer of bone around the periphery of the cystic cavity. The reoccurrence rate for both extraosseous and intraosseous COCs is low and the long-term prognosis is good [2, 3].

Initial marsupialization and decompression are conservative treatments indicated for large lesions and young patients. In fact, these techniques are successfully employed as transitory step in order to decrease the lesion volume, inducing bone formation, and reducing the risk of fracture in the second time of enucleation [14].

CONCLUSION

In conclusion, the Calcified Odontogenic Cyst is an uncommon odontogenic lesion that occurs more frequently in the maxilla. The wide variety of clinical and radiological presentations makes clinical diagnosis challenging. Treatment is generally conservative. Despite its rarity, COC should be considered in the differential diagnosis of jaw lesions and treated promptly due its potentially destructive nature and high rate of root resorption. Although the risk of recurrence within 5 years is rare, long term patient follow-up more than 10 years is recommended.

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