Short Case Report

Calcifying odontogenic cyst: a report of two clinical cases

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Abstract – Introduction: Cystic maxillary lesions are common. In 1962, Gorlin described a rare cystic form termed the calcifying odontogenic cyst (COC) or Gorlin's cyst. Two cases of this form were treated at Bordeaux University Hospital. **Observation:** The first case was a 17-year-old patient with mandibular odontoma, which had developed over the previous 6 months. Excision was performed under local anesthesia, and the diagnosis of COC was made following pathological analysis. A 6-month follow-up was planned. The second case was a 62-year-old patient with a post-extraction mandibular lesion, which had been evolving for 1 year. Enucleation under local anesthesia led to the diagnosis of COC. No recurrence was observed after 5 years of follow-up. **Discussion:** COCs are rare lesions affecting mainly the anterior aspect of the mandible. COCs are usually discovered in unforeseen circumstances, and they can be observed as a clinically painless and well-defined oral deformation. Radiological examination often reveals radiolucent and uniloculated lesions, sometimes associated with radiopaque lesions. Pathological analyses are required for final diagnosis. Management is based on complete excision, more or less associated with marsupialization, and requires an annual clinical radiographic monitoring over the next 5 years. **Conclusion:** COC are rare lesions, usually asymptomatic, whose treatment is based on complete excision. Clinical and radiological follow-up is necessary until complete reossification is achieved.

Introduction

Cystic maxillary bone lesions are common. The most common cystic lesions are dentigerous cysts and keratocysts. Calcifying odontogenic cysts (COCs), or Gorlin's cysts, are rare lesions that account for less than 1% of all odontogenic cysts [1]. This article details two cases of COC, as observed at Bordeaux University Hospital.

Observation

Case #1

A 17-year-old patient, 8 weeks pregnant with amenorrhea, consulted for the management of a mandibular odontoma. Clinically, there was a vestibular arch on teeth 41–43 (Fig. 1A), which had been present for 6 months. Panoramic radiograph showed a mixed image of 41–45 and an impacted tooth at the mandibular symphysis. Cone beam computed tomography of the mandible (Fig. 1B) showed a hypodense image of 31–45 in which regions of dental density were identified. Thinning of the vestibular cortex as well as root resorptions of the teeth 31, 32, and 41 were identifiable. Surgical management comprised

enucleation under local anesthesia, which allowed for the complete excision of the lesion as well as odontomas (Fig. 1C). The surgical procedures showed no complications. Clinical follow-ups 15 and 30 days after the procedure showed no pain; moreover, the mandibular incisors were not mobile, and the vitality tests were positive. Histological examination (Fig. 1D and E) led to COC diagnosis. In fact, a cystic wall covered with an epithelium with ameloblastic differentiation and ghost cells, some of which were calcified, was noted. A 6-month postoperative consultation with a clinical radiographic follow-up was planned.

Case #2

A 62-year-old patient consulted for a mandibular lesion observed 1 year after the avulsion of tooth 36. Clinical examination revealed lack of healing with regard to the alveolus of the extracted tooth, with gingival swelling (Fig. 2A), without pain or sensitivity disorder. Radiological examination (Fig. 2B) revealed a mixed, uniloculated, and well-circumscribed osteolytic lesion. The excision of the lesion was performed under local anesthesia. It was a lesion with a cystic appearance, with partial invasion of the bone tissue and extension to the inferior alveolar nerve, which was preserved (Fig. 2C). Histological analysis

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Fig. 1. (A) Intra-oral photography highlighting a vestibular curve from 41 to 43. (B) Cone beam mandibular frontal section found a hypodense lesion dotted with hyperdense lesions. Note the presence of tooth 33 included. (C) Intraoperative photograph after elevation of a vestibular flap and before removal of the lesion. (D) and (E) Pathological anatomical section ($\times 10$) HES staining with odontogenic-dentinoid formation (orange) and mummification (arrow) by keratinization giving ghost cells.

Fig. 2. (A) Preoperative view with gingival swelling. (B) Intraoperative view of the intervention after exeresis of the Lesion and preservation of the mental nerve. (C) Panoramic tooth finding an osteolytic lesion of mixed, uniloculated and well circumscribed appearance. (D) Panoramic tooth at 5 years finding no recurrence.

revealed COC. Following anatomoclinical consultation, a simple clinical and radiological surveillance every 3 months was planned. Five years after the resection, no clinical or radiological recurrence was observed (Fig. 2D).

Discussion

COCs, dentinogenic ghost cell tumors, and odontogenic ghost cell carcinomas form a group of odontogenic “ghost cell” tumors. The term “ghost cells,” introduced in 1946 by Thomas and Goldman, highlights the origin and nature of these lesions and describes their microscopic characteristics [1]. COC was first described in 1932 in the work of Rywkind. Then, Gorlin, in 1962, defined COC as a distinct lesion of the calcifying odontogenic tumor [1]. In 2005, the World Health Organization classified COC as a benign tumor. This classification was modified in 2017 in which COC was reclassified among cystic lesions [1].

COC is most often asymptomatic and is often only discovered during a routine radiological examination. There are no clinical signs or pathognomonic radiological signs of this lesion; only histological examination allows for its differentiation from other cystic diseases of the maxillae. There are cases of COC discovered during the avulsion of mandibular wisdom teeth after histological analysis of the pseudofollicular sac [2]. COC can be found in patients of all ages, equally in young children, adolescents, and adults [2,3].

Here, we observed one case in a teenager and another in an adult.

Clinically, patients present for consultation with a painless, well-defined maxillary or mandibular swelling that has evolved over a few months [3]. In case #1, the patient presented with these exact symptoms, while in case #2, the patient presented with atypical clinical signs, with a healing delay.

The anterior mandible is the most frequently affected site by COC, with a prevalence of approximately 65% [4]. The lesion is often unilateral, although bilateral involvement of the mandible has been rarely reported [3]. In case #1, the patient presented the most frequent COC localization, whereas in case #2, the patient presented the posterior mandibular localization, which is more unusual than the descriptions in the literature.

Radiologically, COC is most often in the form of a unilocular and well-defined radiolucent lesion, as observed in both cases presented. Within these radio-clear images, radiopaque areas have been described [3]. COC is frequently associated with odontomas. Root resorptions or cortical bone erosions can be observed on radiography. In case #1, an odontoma and multiple root resorptions were observed.

Histologically, the COC wall is lined with an odontogenic epithelium comprising ameloblastic differentiation cells. Ghost cells corresponding to mummified epithelial cells are found through large-scale keratinization [3,4]. These ghost cells can calcify, and calcifications are constant but vary in number.

COC treatment is based on surgical enucleation [3,4]. Initial decompression by marsupialization, secondarily followed by enucleation is also possible. The risk of recurrence is low (3%–11%) but requires a radio-clinical follow-up of 6 months after the procedure and then annually for 5 years to check for complete reossification. There is a low risk of transformation into a ghost cell malignant tumor; eight such cases have been described between 2003 and 2013 [5].

Conclusion

COCs are rare tumors, which are most often asymptomatic. Treatment is based on complete excision. A clinical follow-up is necessary to ensure complete reossification. Pathological examination is essential because the diagnosis is based on the radio-clinical and histological signs.

Conflicts of interests: The authors declare that they have no conflicts of interest in relation to this article

References

1. Arruda JAA, Monteiro JLGC, Abreu LG, de Oliveira Silva LV, Schuch LF, de Noronha MS, *et al.* Calcifying odontogenic cyst, dentinogenic ghost cell tumor, and ghost cell odontogenic carcinoma: A systematic review. *J Oral Pathol Med.* 2018;47:721–730.
2. Sarode GS, Sarode SC, Prajapati G, Maralingannavar M, Patil S. Calcifying cystic odontogenic tumor in radiologically normal dental follicular space of mandibular third molars: Report of two cases. *Clin Pract* 2017;7:933.
3. Gadipelly S, Reddy VB, Sudheer M, Kumar NV, Harsha G. Bilateral calcifying odontogenic cyst: A rare entity. *J Maxillofac Oral Surg* 2015;14:826–831.
4. Santos HB de P, de Moraes EF, Moreira DGL, Neto LF de A, Gomes PP, Freitas R de A. Calcifying odontogenic cyst with extensive areas of dentinoid: Uncommon case report and update of main findings. *Case Rep Pathol* 2018;2018:1–4.
5. Tarakji B, Ashok N, Alzoghaibi I, Altamimi MA, Azzeghaiby SN, Baroudi K, *et al.* Malignant transformation of calcifying cystic odontogenic tumour — a review of literature. *Contemp Oncol* 2015;19:184–186.