



Calcifying cystic odontogenic tumour associated with odontoma involving mixed dentition

Tumor odontogênico cístico calcificante associado a odontoma envolvendo dentição mista

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ABSTRACT

Calcifying cystic odontogenic tumor (CCOT) is a benign tumor relatively uncommon that may be associated with other odontogenic lesions. This manuscript describes the case of a 9-year-old girl affected by CCOT associated with odontoma, presenting as a slight swelling along with prolonged retention of a maxillary primary central incisor. Radiographic examination showed a unilocular radiolucent lesion containing radiopaque material associated with an unerupted tooth. Taking into consideration the clinical diagnostic of odontoma, an excisional biopsy was carried out. Microscopic examination established the diagnosis of a CCOT associated with odontoma. The association of these lesions is rare in individuals in the first decade of life, particularly involving mixed dentition, as outlined in this case. The patient remains under care with no clinical signs of recurrence.

KEYWORDS

Odontogenic tumors; Calcifying cystic odontogenic tumor; Odontoma; Gorlin cyst.

RESUMO

O tumor odontogênico cístico calcificante (TOCC) é um tumor odontogênico cístico benigno relativamente raro e pode estar associado a outras lesões odontogênicas. Este manuscrito descreve um caso de CCOT associado a odontoma em uma menina de 9 anos de idade, que se apresentou com um ligeiro aumento de volume associado a retenção prolongada de um incisivo central superior. O exame radiográfico revelou uma lesão radiolúcida unilocular contendo material radiopaco, associado a um dente incluso. Tomando em consideração o diagnóstico clínico de odontoma, uma biópsia foi realizada. O exame microscópico estabeleceu o diagnóstico de um TOCC associado a odontoma. A associação destas lesões é rara em indivíduos na primeira década de vida, particularmente envolvendo dentição mista, conforme descrito no presente caso. O paciente permanece sob cuidados sem sinais clínicos de recorrência.

PALAVRAS-CHAVE

Tumores odontogênicos; Tumor odontogênico cístico calcificante; Odontoma; Cisto de Gorlin

INTRODUCTION

Calcifying cystic odontogenic tumor (CCOT), a painless slowly growing mass, is a benign odontogenic neoplasm of slow growth. CCOT mainly affects the anterior region of the jaws

without predilection for maxilla or mandible [1,2]. There is no preference for gender and it is most commonly reported in young adults in the third and fourth decades of life, rarely affecting pediatric patients [1,3,4]. CCOT can present some unusual associations with other

odontogenic tumors, such as adenomatoid odontogenic tumor, ameloblastic fibroma, ameloblastic fibro-odontoma and, most frequently, odontoma [5,6,7].

This report aims at a case of CCOT associated with odontoma in a child, highlighting the importance of the pediatric dentist for the screening and diagnosis of oral pathologies when there is delay in the eruption of permanent teeth.

CASE REPORT

A 9-year-old Caucasian girl presented to our department with a chief complaint of prolonged retention of the tooth 61. There was no history of trauma or inflammation in the region, and the medical history was noncontributory. No alterations were observed on extraoral examination. Intraorally, there was an asymptomatic slight swelling on the cortical bone, surrounding tooth 61, firm on palpation and covered by healthy pink mucosa. Radiographic examination revealed a unilocular radiolucent area affecting the left maxillary area in buccal side, based on 61 tooth. The lesion was well circumscribed and demarcated by a

radiopaque halo in association with the impacted unerupted tooth 21 (Figure1). Radiopaque material similar to denticles was evident within the radiolucency (Figure1 and Figure2). Based on the clinical and radiographic features, the diagnostic hypothesis was odontoma. Complete surgical removal of the lesion and associated teeth was performed. The surgical specimen was fixed in 10% formalin and sent for microscopic examination. Histological sections stained with Hematoxylin & Eosin revealed an odontogenic cystic lesion, partially lined by stratified squamous epithelium with clusters of ghost cells, sometimes calcified, in the superficial layer. Sheets of ghost cells were observed in proximity to tubular and immature dentin, pulpal tissue, and in less extent enamel matrix and cementum, recapitulating the organization of a normal tooth (Figure3). A diagnosis of CCOT in association with a compound odontoma was made. Periodical clinical examination, periapical and panoramic radiographies have been performed every six months since diagnosis. No clinical or radiographic signs of recurrence have been observed during a follow-up period of 18 months.



Figure 1 - Panoramic radiograph showing the well-defined radiolucent area containing a radiopaque mass, resulting in impaction of tooth 21.



Figure 2 - Periapical radiographs showing details of the lesion. Note the close relationship of the lesion with unerupted tooth 21.

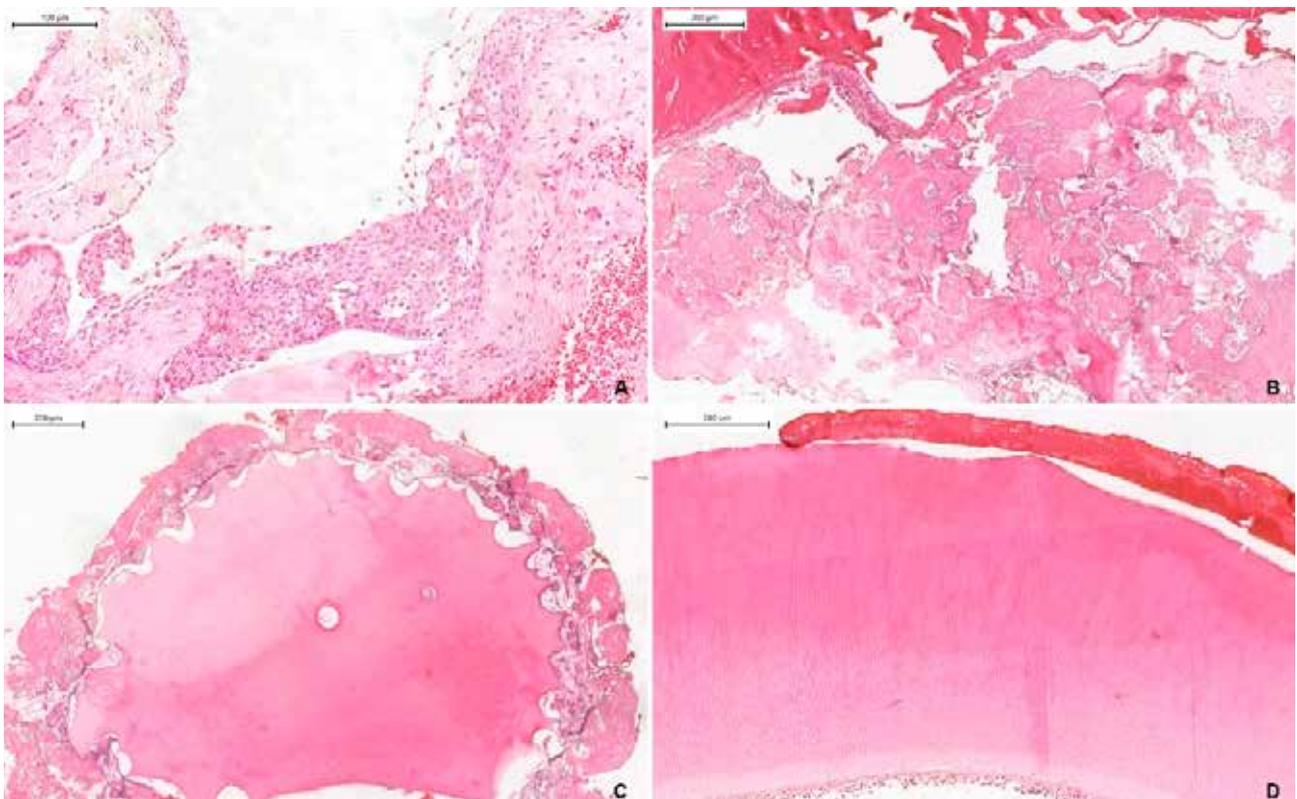


Figure 3 - Cystic cavity lined by a stratified squamous epithelium with cuboidal basal cells and overlying loosely arranged cells, similar to stellate reticulum (A). Sheet of ghost cells in the fibrous wall adjacent to calcified tissue (B). Dentoid tissue involving hollow spaces of odontoma in close proximity to numerous ghost cells (C). Tubular dentin adjacent to odontoblast-like cells (D) (H&E; Bars indicate: 100 μ for A and 200 μ for B, C and D).

DISCUSSION

In 1962, Gorlin et al. first described CCOT, formerly known as calcifying odontogenic cyst (COC) [8]. Since then, the literature continues to discuss its proper nomenclature and classification. In 2005, the World Health Organization (WHO) adopted the term CCOT and reclassified this lesion as a benign cystic neoplasm [9].

CCOT is an uncommon lesion, accounting for about 2% of all odontogenic tumors [10]. Cases involving pediatric and mixed dentition, as in the present report, are rare. To our knowledge, only three similar cases were reported in the literature [3,4,10].

Clinically, CCOT is usually presented as a painless and slow-growing mass, affecting equally the mandible and maxilla with predilection for the anterior region [9]. Radiographic examination of CCOT usually shows a radiolucent unilocular lesion with discrete margins delimitation and displays radiopaque material of irregular sizes in its interior [2]. This tumor may be associated with impacted teeth or other odontogenic lesion such as adenomatoid odontogenic tumor, ameloblastoma and, more often, odontoma [6]. In this case, the lesion affected the anterior maxillary area and the radiographic image revealed a well-defined radiolucent area containing various amounts of irregular radiopaque material, corroborating relevant literature.

The present case presented the typical CCOT histopathologic findings described in the literature: cystic cavity lined by odontogenic epithelium of varying thickness with basal cells of columnar or cuboid aspect, similar to ameloblasts. The overlying epithelial layer was loosely organized, resembling the stellate reticulum of the enamel organ. The most prominent microscopic feature is the presence of numerous anucleate epithelial cells with eosinophilic cytoplasm, known as “ghost cells”, mostly arranged in cellular sheets or nests

[2,9,4]. In this case, adjacent to CCOT, there were tubular dentin, enamel, cementum and dental pulp-like tissue which corroborated with the final diagnosis of CCOT associated with compound odontoma.

Hirshberg et al. analyzed 52 cases of odontoma-associated CCOT and proposed that this association should be considered a separate entity which could lead to a better understanding of its pathogenesis, suggesting the term “Odontocalcifying Odontogenic Cyst”. These authors reported a higher frequency in women, and in the anterior maxilla, similar to the features described in the present case [2].

The elective treatment for CCOT is surgical removal by enucleation or curettage [11]. When there is association with other odontogenic tumors, the treatment must be conducted according to the recommendation for the lesion with the most aggressive clinical procedure [6,10]. In this case, the surgical removal was chosen for diagnostic establishment with no need for complementary therapy. Follow-up of the patient, including periodic clinical and radiographic examination, should be performed as shown in the case due to the possibility of recurrence, despite the rarity of this event [1,5].

In conclusion, CCOT is an odontogenic lesion uncommon in pediatric patients, showing varied behavior from the clinical, radiographic and histological point of view. The detailed clinical examination and complementary exams are essential for a careful diagnosis, in order to offer the patient the appropriate treatment seeking for favorable prognosis.

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Nil.

CONFLICT OF INTEREST

None declared.

REFERENCES

1. Buchner A. The central (In traosseous) calcifying odontogenic cyst: an analysis of 215 cases. *J Oral Maxillofac Surg.*1991;49:330-9.
2. Hirshberg A, Kaplan I, Buchner A. Calcifying odontogenic cyst associated With odontoma: a possible separate entity (Odontocalcifying Odontogenic Cyst). *J Oral Maxillofac Surg.* 1994; 52: 555-8.
3. Fernandes AM, Duarte EC, Pimenta FJ, Souza LN, Santos VR, Mesquita RA, et al. Odontogenic tumors: a study of 340 cases in a Brazilian population. *J Oral Pathol Med.* 2005 Nov;34(10):583-7.
4. Reyes D, Villanueva J, Espinosa S, Cornejo M. Odontogenic calcific cystic tumor: A report of two clinical cases. *Med Oral Patol Oral Cir Bucal.* 2007 Mar 1;12(2):E126-9.
5. Ledesma-Montes C, Gorlin RJ, Shear M, Prae Torius F, Mosqueda-Taylor A, Altini M, et al. International collaborative study on ghost cell odontogenic tumours: calcifying cystic odontogenic tumour, dentinogenic ghost cell tumour and ghost cell odontogenic carcinoma. *J Oral Pathol Med.* 2008 May;37(5):302-8. doi: 10.1111/j.1600-0714.2007.00623.x.
6. Sonone A, Sabane VS, Desai R. Calcifying ghost cell odontogenic cyst: report of a case and review of literature. *Case Rep Dent.* 2011; 328743. doi: 10.1155/2011/328743.
7. Gorlin RJ, Pindborg JJ, Clausen FP, Vickers RA. The calcifying odontogenic cyst: a possible analogue of the cutaneous epithelioma of Malherbe: an analysis of fifteen cases. *Oral Surg Oral Med Oral Pathol.* 1962 Oct;15:1235-43.
8. Praetorius F, Ledesma-Montes C. Calcifying cystic odontogenic tumor. In: Barnes EL, Everson JW, Reichart P, Sidransky D. *Pathology and Genetics of Head and Neck Tumours. World Health Organization Classification of Tumours.* Lyon France: IARC Press; p. 313, 2005.
9. Poleti ML, Duarte BG, Lara VS, Freitas-Faria P, Rubira-Bullen IR, Gonçalves ES. Odontoma associated with calcifying cystic odontogenic tumor in deciduous dentition: case report. *Oral Maxillofac Surg.* 2013 Mar;17(1):77-80. doi: 10.1007/s10006-012-0333-x.
10. Han PP, Nagatsuka H, Siar CH, Tsujigiwa H, Gunduz M, Tamamura R, et al. A pigmented calcifying cystic odontogenic tumor associated with compound odontoma: a case report and review of literature. *Head Face Med.* 2007;3(35), 2007. 6p. doi: 10.1186/1746-160X-3-35

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