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Case report

Multiple Dentigerous Cysts: two case reports of nonsyndromic twins

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ABSTRACT

Dentigerous cysts are odontogenic cysts associated with the crowns of permanent unerupted teeth. Multiple dentigerous cysts are very rare and mostly found in patients with syndromes. This paper reports two homozygous twins with dentigerous cysts in both unerupted lower second molars. In one patient, a third dentigerous cyst was associated with an unerupted maxillary canine. Enucleation of smaller lesions and marsupialization of the larger lesions were performed to preserve the permanent teeth, with good final positioning. (Rev Port Estomatol Med Dent Cir Maxilofac. 2019;60(1):32-36)

Múltiplos Quistos Dentígeros: relato de caso em gémeos não sindrómicos

Palavras-chave:
Quisto dentígero
Dente não erupcionados
Gémeos

RESUMO

Quistos dentígeros são quistos odontogénicos associados à coroa de dentes permanentes não erupcionados. Múltiplos quistos dentígeros são considerados raros e encontrados principalmente em pacientes com síndromes. Este trabalho apresenta um caso de gémeos homozigotos com quistos dentígeros em ambos os segundos molares inferiores não erupcionados. Num dos pacientes, um terceiro quisto dentígero foi associado ao canino maxilar não erupcionado. Para o tratamento, foi realizada enucleação das lesões menores e marsupialização das lesões maiores para preservar os dentes permanentes, com bom posicionamento final. (Rev Port Estomatol Med Dent Cir Maxilofac. 2019;60(1):32-36)

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Introduction

The dentigerous cyst (DC) is the most common odontogenic cyst of developmental origin.[1] DCs are thought to result from the proliferation of the reduced enamel epithelium (REE) shortly after enamel formation during dental tissue development.[2,3] The DC may partially or completely involve the crown of an unerupted tooth, and be attached to the tooth at the cement-enamel junction.[2,3] Degeneration of the stellate reticulum, accumulation of fluid between the layers of REE, diminished adherence of REE, and monoclonal proliferation in the REE through altered protein patched homolog (PTCH) signaling in the primary cilium have been suggested to explain the pathogenesis of DC.[4]

It is estimated that 1.4 DCs are found for every 100 unerupted teeth. This cyst is most common in males, has a peak incidence in the second decade of life, and affects mostly mandibular third molars.[5] They are usually slow growing asymptomatic lesions that are not discovered until they affect surrounding organs, during orthodontic treatment planning or from the delayed eruption of a permanent tooth.[5,6] On radiographic examination, a DC commonly appears as a well-delimited unilocular radiolucency, encircling the crown of an unerupted tooth.[6]

Over 95% of DCs are single lesions.[3,5] Multiple occurrences of DC have been described in mucopolysaccharidosis (Maroteaux-Lamy and Hunter's syndrome).[7,8] Moreover, the association of multiple DCs with other syndromes, such as cleidocranial dysplasia and basal cell nevus syndrome, has been poorly documented in the literature.[5] However, strong evidence taken from a large series of cases has indicated that the multiplicity of DCs is not a syndromic event.[5] Although some authors believe that multiple and bilateral DCs are rare in the absence of developmental syndromes,[9] many cases of multiple DCs in nonsyndromic patients have previously been reported.[10-12] We present the case of nonsyndromic twins with multiple DCs, a condition that, to our knowledge, has not been previously reported.

Case report

Two 14-year-old male twins were referred by their orthodontist to the Oral and Maxillofacial Surgery Service of the Federal University of Uberlândia, Brazil, for the evaluation of asymptomatic cystic lesions. Their past medical history showed no evidence of any significant alteration, and their routine laboratory tests were within normal limits. Extraoral examination revealed a symmetric appearance of the face, without swelling or tenderness.

Patient 1: The intraoral examination revealed a discrete swelling in the maxillary anterior labial vestibule extending from the left central incisor to the left lateral incisor. Teeth 23, 17, 27, 37 and 47 were absent of the oral cavity (Figure 1). The panoramic radiograph showed well-circumscribed, corticated and unilocular radiolucent lesions associated with the unerupted teeth 23, 37 and 47 (Figure 2). Based on clinical and radiographic findings, a working diagnosis of dentigerous cyst was made. The odontogenic keratocystic tumor was also considered in the differential diagnosis. Marsupialization of the larger lesion (tooth 47) and surgical enucleation of the smaller lesions (teeth 23 and 37) were performed under local anesthesia. Care was taken to preserve the teeth involved. No complementary surgical procedures were necessary for these areas. In addition, an ulectomy was also performed in the region of teeth 17 and 27 to assist eruption. The postoperative period was uneventful.

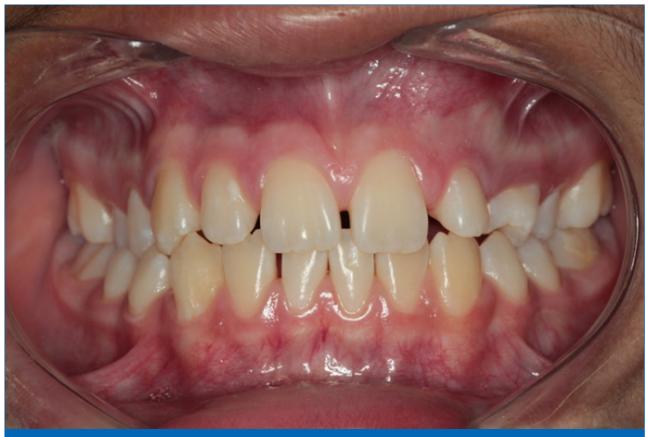

Figure 1. Patient 1 – Intraoral aspect.

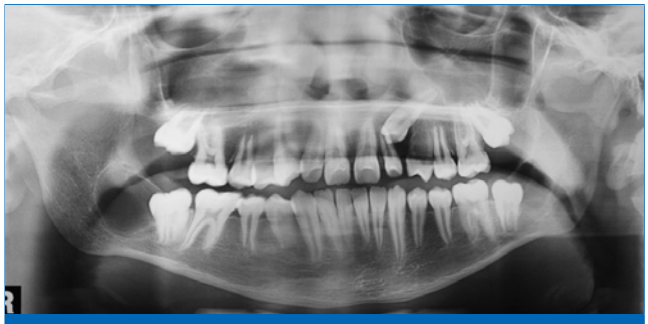

Figure 2. Patient 1 – Panoramic radiograph evidencing the lesions associated with teeth 23, 37 and 47.

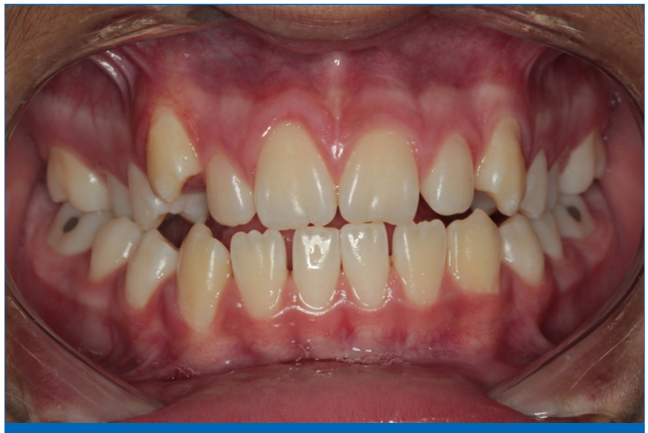

Figure 3. Patient 2 – Intraoral aspect.

Patient 2: The intraoral examination revealed the absence of teeth 17, 27, 37 and 47 (Figure 3). The panoramic radiograph

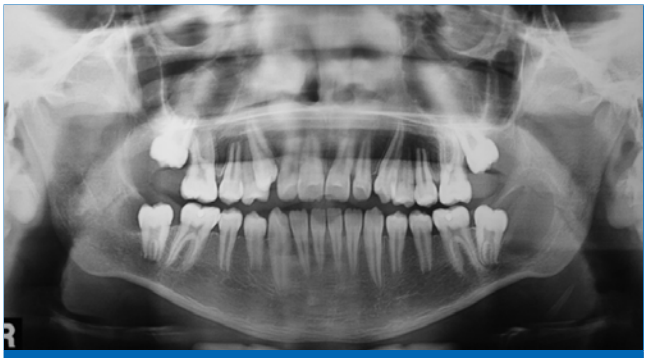

Figure 4. Patient 2 – Panoramic radiograph evidencing the lesions associated with teeth 37 and 47.

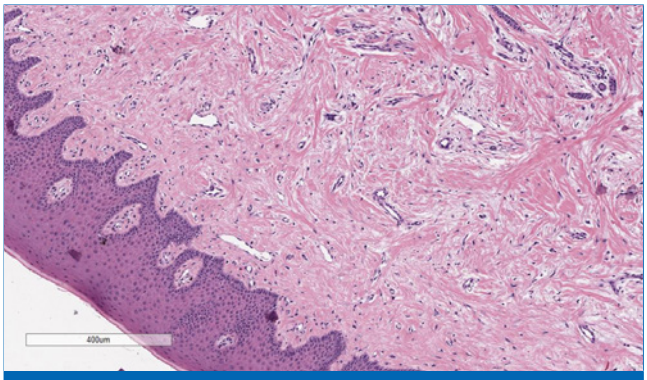

Figure 5. Histopathological findings showing both left cystic lesions coated by non-keratinized stratified squamous epithelium of variable thickness and the presence of hyperplastic epithelium and areas of intense chronic inflammation on the cyst wall. Hematoxylin & Eosin.

Figure 6. Patient 1 – Post-operative intraoral aspect.

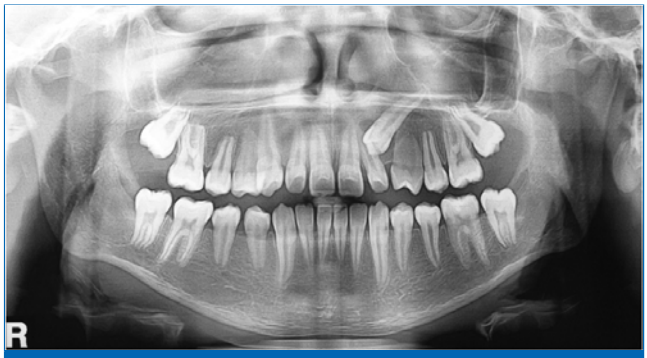

Figure 7. Patient 1 – Post-operative panoramic radiograph evidencing bone repair and better position of teeth 23, 37 and 47 after 12 months of follow-up.

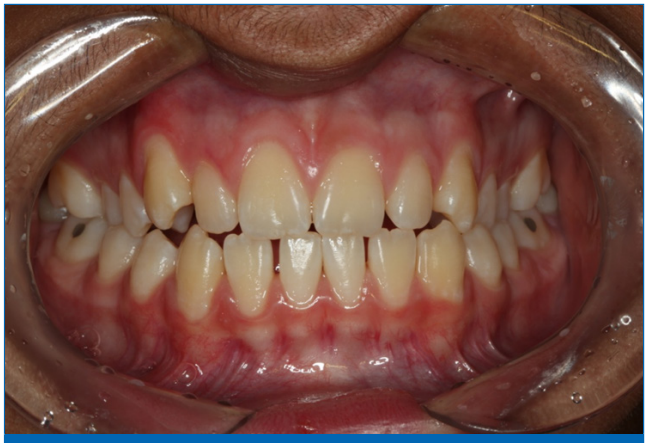

Figure 8. Patient 2 – Post-operative intraoral aspect.

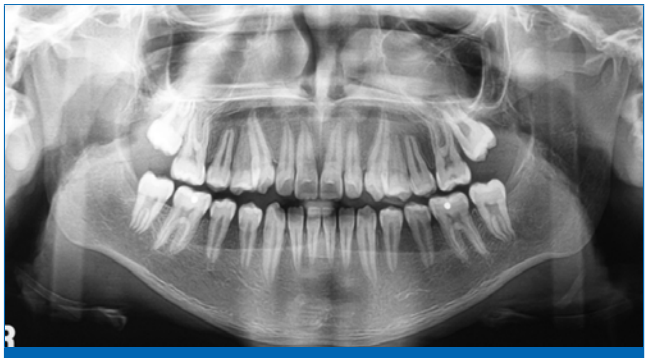

Figure 9. Patient 2 – Post-operative panoramic radiograph evidencing bone repair and better position of teeth 37 and 47 after 12 months of follow-up.

showed well-circumscribed, corticated and unilocular radiolucent lesions associated with teeth 37 and 47 (Figure 4). A working diagnosis of dentigerous cyst was made, and the odontogenic keratocyst was considered in the differential diagnosis. Marsupialization of the larger lesion (tooth 37) and surgical enucleation of the smaller lesion (tooth 47) were performed under local anesthesia. Care was taken to preserve the tooth involved. No complementary surgical procedures were necessary for these areas. In addition, an ulectomy was performed in the region of teeth 17 and 27 to assist eruption. The postoperative period was uneventful.

Specimens were sent for histopathologic analysis, which confirmed the diagnosis of dentigerous cyst (Figure 5). After 12 months of follow-up, the teeth involved moved close to their ideal position (Figures 6, 7, 8 and 9). Both patients were referred to an orthodontist for orthodontic treatment.

Discussion and conclusions

DCs are benign odontogenic cysts surrounding the crowns of permanent teeth.[1] They may involve impacted, unerupted permanent teeth, supernumerary teeth and odontomas, and rarely involve deciduous teeth.[3,6,11] The origin of DCs has been partially attributed to PTCH gene inactivation in a progenitor cell of the enamel organ.[13] Some authors believe that PTCH alterations are necessary and may be the event that triggers the formation and growth of various cysts.[14,15] Other authors have reported a possible association between abnormal chromosome 1 and the occurrence of DCs.[16] These assumptions suggest that genetic factors may be related to the pathogenesis of odontogenic cysts. In the present cases, the occurrence of such similar clinical conditions in homozygous twins was, at least, curious and rare, and genetic involvement cannot be ruled out.

Multiple DCs generally occur associated with syndromes such as mucopolysaccharidosis[7,8] and cleidocranial dysplasia.[5] Both these diseases cause modifications in tooth development and may take part in the occurrence of multiple DCs[17,18] Furthermore, multiple mandibular cysts have previously been reported after prolonged use of cyclosporine A and calcium channel blockers;[10] however, these are uncommon in the absence of a syndrome or systemic disease.[9] The patients analyzed in this case had no clinical or laboratory findings that could point towards syndromes or other systemic changes.

Clinical and radiographic findings are almost conclusive in diagnosing DCs; however, the pathological analysis of the lesion is essential for a definitive diagnosis, especially in cases of multiple lesions, because other changes may share similar radiographic features. The odontogenic keratocyst tumor, for example, most commonly located in the mandibular body or ramus, as in the present case, can appear as multiple lesions in the basal cell nevus syndrome.[17] That is an uncommon disorder characterized by numerous basal cell carcinomas, maxillary keratocysts and musculoskeletal malformations.[17,18] No microscopic and clinical features of this syndrome were found in the patients of the present case report.

The standard treatment for a DC is enucleation and removal of the cyst-associated impacted or unerupted tooth. However, larger lesions can be surgically drained and marsupialized to relieve the pressure within the cyst.[19] In the present cases, enucleation was performed to treat the smaller lesions, and complete removal of the cyst was possible, without damaging the teeth. The larger lesions were marsupialized, which decreased the intracystic pressure, promoting the reduction of cysts and favorable bone formation. This minimally invasive procedure reduced the risk of large bone defects and paresthesia. In addition, it had the advantage of preserving the tooth, allowing its spontaneous eruption. The possible disadvantage would be the presence of a cavity that needed care and irrigation. In the present case, the good oral hygiene and cooperation of the patients contributed to the successful treatment. It is important to remember that, when marsupialization is chosen as the therapeutic option, a second surgical procedure may be necessary to remove remnants of the lesion, which did not happen in this case.

Although DCs are ranked as the second most common odontogenic cysts, their development as multiple lesions is unusual. The involvement of homozygous twins has not been previously reported. Radiographic examination for the investigation of unerupted teeth is essential to evaluate the presence of possible odontogenic cysts and tumors and aim for an early diagnosis of the lesion to preserve anatomical structures. Enucleation and marsupialization can be most successfully used as treatment.

Ethical disclosures

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors have obtained the written informed consent of the patients or subjects mentioned in the article. The corresponding author is in possession of this document.

Conflict of interest

The authors have no conflicts of interest to declare.

REFERENCES

1. Johnson NR, Gannon OM, Savage NW, Batstone MD. Frequency of odontogenic cysts and tumors: a systematic review. J Investig Clin Dent. 2014;5:9-14.
2. Imada TS, Neto VT, Bernini GF, Silva Santos PS, Rubira-Bullen IR, Bravo-Calderòn D, et al. Unusual bilateral dentigerous cysts in a nonsyndromic patient assessed by cone beam computed tomography. Contemp Clin Dent. 2014;5:240-2.
3. Huang G, Moore L, Logan RM, Gue S. Histological analysis of 41 dentigerous cysts in a paediatric population. J Oral Pathol Med. 2019;48:74-8.
4. Anoop UR, Verma K, Narayanan K. Primary cilia in the pathogenesis of dentigerous cyst: a new hypothesis based on role of primary cilia in autosomal dominant polycystic kidney disease. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 2011;111:608-17.
5. Zhang LL, Yang R, Zhang L, Li W, MacDonald-Jankowski D, Poh CF. Dentigerous cyst: a retrospective clinicopathological analysis of 2082 dentigerous cysts in British Columbia, Canada. Int J Oral Maxillofac Surg. 2010;39:878-82.
6. Aoki N, Ise K, Inoue A, Kosugi Y, Koyama C, Iida M, et al. Multidisciplinary approach for treatment of a dentigerous cyst – marsupialization, orthodontic treatment, and implant placement: a case report. J Med Case Rep. 2018;12:305.
7. Anand R, Bhatia D, Yadav DS. Mucopolysaccharidosis type I Hurler-Scheie syndrome affecting two sisters. Radiol Case Rep. 2015;7:641.
8. de Almeida-Barros RQ, de Medeiros PFV, de Almeida Azevedo MQ, de Oliveira Lira Ortega A, Yamamoto ATA, Dornelas SKL, et al. Evaluation of oral manifestations of patients with

mucopolysaccharidosis IV and VI: clinical and imaging study. Clin Oral Investig. 2018;22:201-8.
9. Dhupar A, Yadav S, Dhupar V, Mittal HC, Malik S, Rana P. Bi-maxillary dentigerous cyst in a non-syndromic child – review of literature with a case presentation. J Stomatol Oral Maxillofac Surg. 2017;118:45-8.
10. AlKhudair B, AlKhatib A, AlAzzeh G, AlMomen A. Bilateral dentigerous cysts and ectopic teeth in the maxillary sinuses: A case report and literature review. Int J Surg Case Rep. 2019;55:117-20.
11. Devi P, Thimmarasa VB, Mehrotra V, Agarwal M. Multiple dentigerous cysts: a case report and review. J Maxillofac Oral Surg. 2015;14(Suppl1):47-51.
12. Jeon JY, Park CJ, Cho SH, Hwang KG. Bilateral dentigerous cysts that involve all four dental quadrants: a case report and literature review. J Korean Assoc Oral Maxillofac Surg. 2016;42:123-6.
13. Gondim JO , Neto JJ , Nogueira RL , Giro EM . Conservative management of a dentigerous cyst secondary to primary tooth trauma. Dent Traumatol. 2008;24:676-9.
14. Levanat S, Pavelić B, Crnić I, Oresković S, Manojlović S. Involvement of PTCH gene in various inflammatory cysts. J Mol Med (Berl). 2000;78:140-6.
15. Pavelić B, Levanat S, Crnić I, Kobler P, Anić I, Manojlović S, et al. PTCH gene altered in dentigerous cysts. J Oral Pathol Med. 2001;30:569-76.
16. Batra P, Roychoudhury A, Balakrishan P, Parkash H. Bilateral dentigerous cyst associated with polymorphism in chromosome 1qh+. J Clin Pediatr Dent. 2004;28:177-81.
17. Deboni MC, Brozoski MA, Traina AA, Acay RR, Naclério-Homem Mda G. Surgical management of dentigerous cyst and keratocystic odontogenic tumor in children: a conservative approach and 7-year follow-up. J Appl Oral Sci, 2012;20:268-71.
18. Saluja JS, Ramakrishnan MJ, Vinit GB, Jaiswara C. Multiple dentigerous cysts in a nonsyndromic minor patient: Report of an unusual case. Natl J Maxillofac Surg. 2010;1:168-72.
19. Khambete N, Kumar R, Risbud M, Kale L, Sodhi S. Dentigerous cyst associated with an impacted mesiodens: report of 2 cases. Imaging Sci Dent. 2012;42:255-60.