Bilateral Dentigerous Cyst in a Nonsyndromic Patient: Case Report and Literature Review

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ABSTRACT

Dentigerous cysts are the most common developmental cysts of the jaws and are associated with impacted teeth. Bilateral dentigerous cysts are rare and typically occur in association with a developmental syndrome. The occurrence of bilateral dentigerous cysts in the absence of a developmental syndrome is very rare. The purpose of this paper was to report on a nonsyndromic, 5-year-old boy who presented mandibular bilateral dentigerous cysts.
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A dentigerous cyst (**DC**) is the second most common odontogenic cyst, after radicular cyst. It encloses the crown of an unerupted tooth at the cementoenamel junction; the mandibular third molars are the most commonly affected. This cyst accounts for approximately 24% of all true cysts of the jaws, and their frequency in the general population has been estimated at 1.44 cysts for every 100 unerupted teeth. There is usually no pain or discomfort associated with the cyst unless it becomes secondarily infected.[1,2]

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Radiographically, it is observed as an expansive, well-circumscribed, radiolucent lesion with a sclerotic reactive line associated with an unerupted tooth. Microscopically, it reveals a fluid-filled cyst lined by a thin, nondistinctive, nonkeratinized epithelium. The epithelial lining consists of 2 to 6 layers of cuboidal epithelial cells, and the epithelium-connective tissue interface is flat. The inflammatory DC exhibits mononuclear cells in the cystic wall, with a possibility of squamous metaplasia of the epithelium.[3,4]

Most DCs are solitary. Bilateral or multiple cysts are usually associated with developmental syndromes, such as mucopolysaccharidosis, basal cell nevus syndrome, and cleidocranial dysplasia.[5-8] The occurrence of bilateral dentigerous cysts in the absence of a developmental syndrome is rare.[2,3,9]

CASE REPORT

A 5-year-old boy attended the Stomatology Service of João Barros Barreto University Hospital, Federal University of Pará, Belém, Pará, Brazil, with bilateral facial swelling. The patient's medical history was noncontributory and, except for

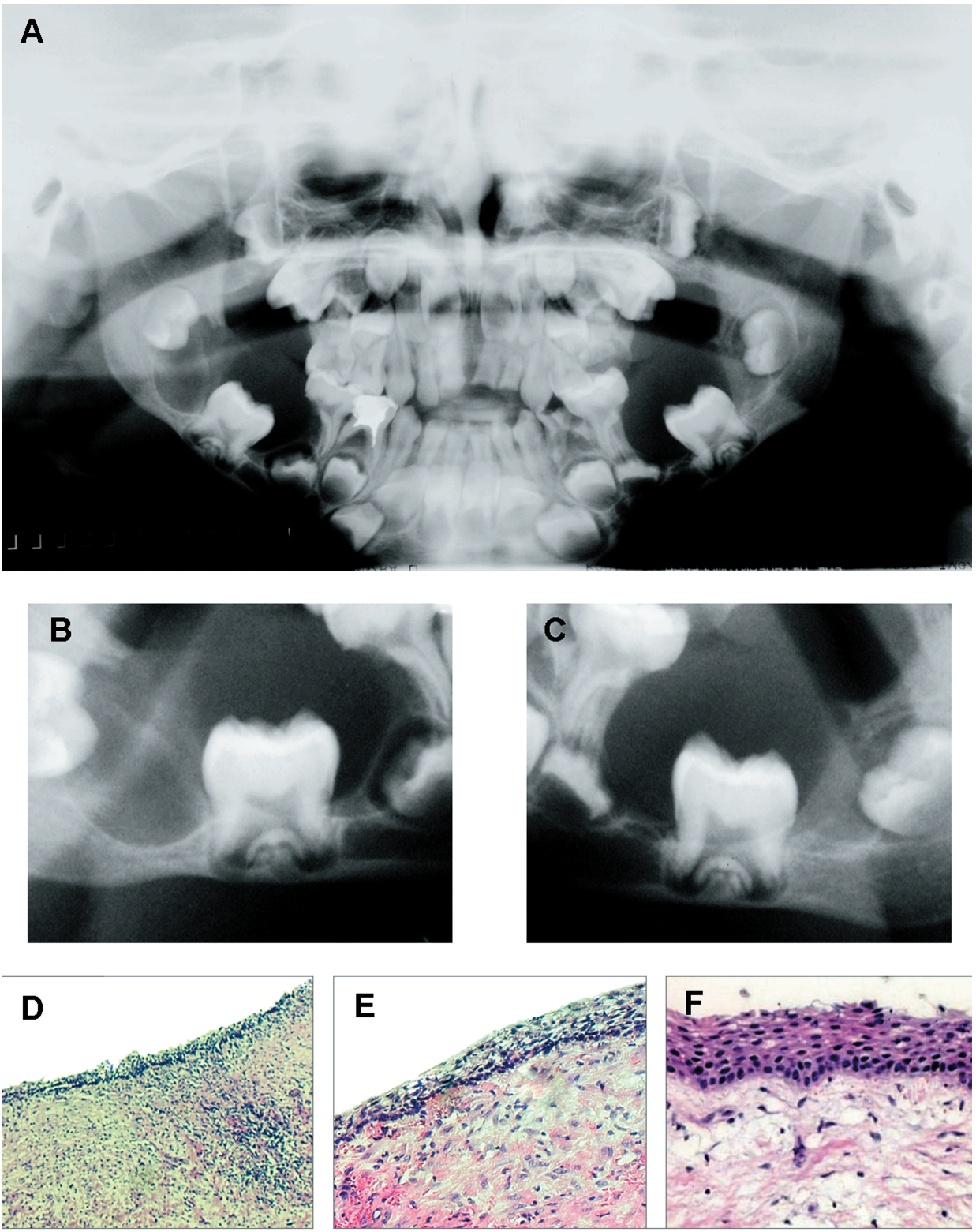

Figure 1. Unilocular radiolucent lesions associated with the crowns of unerupted permanent mandibular first molars—panoramic radiograph (A). In detail, well-defined sclerotic margins and root resorption of the deciduous second molar—periapical radiograph of the right side (B) and left side (C). Cystic wall composed of fibrous tissue with focal mononuclear inflammatory cells lined by a thin, stratified, squamous, nonkeratinized epithelium—microscopy low magnification X100 (D). Microscopy high magnification X400 (E) and X1,000 (F).

his facial asymmetry, physical examination was unremarkable. Intraoral examination revealed buccal swelling on both sides of the mandible. The mucosa was unaffected. Other disturbances, such as condylar defects and gingival hyperplasia, were not detected.

Panoramic and periapical radiographs were obtained, which revealed 2 unilocular radiolucent lesions characterized by well-defined sclerotic margins associated with the crowns of unerupted permanent mandibular first molars. Root resorption of the deciduous second molar was also observed (Figures 1a, b, and c).

The patient was referred to a pediatric geneticist, who did not find any signs of developmental syndromes such as clavicle aplasia, excessive enlargement of the cranium diameter, kidney or spleen expansion, abnormal bone formation, abnormal auditory and respiratory function, visual and cardiovascular alterations, and palmar and plantar pits. Also, dermatological findings resembling basal cell carcinoma or epidermal cyst were not observed. Additionally, the parents presented no signs of developmental syndromes.

The cysts were enucleated by intraoral approach under general anesthesia. The teeth surrounded by the lesion

Table 1. Literature Review

Authors	Year	Gender*	Age (ys)	Location	Treatment
Myers[9]	1943	F	19	Permanent mandibular right and left third molars	Enucleation
Henefer[10]	1964	F	52	Permanent maxillary right and left third molars	Enucleation
Stamback[11]	1970	M	9	Permanent mandibular right and left third molars	Enucleation
Callaghan[12]	1973	M	38	Permanent mandibular right and left third molars	Enucleation
Burton and Scheffer[13]	1980	F	57	Permanent mandibular right and left third molars	Enucleation
Swerdloff and Cols[14]	1980	F	7	Permanent mandibular right and left third molars	Enucleation
Crinzi[15]	1982	F	15	Permanent mandibular right and left third molars	Enucleation
McDonnell[16]	1988	M	15	Permanent mandibular second premolar/ second molar (side not informed)	Enucleation
Eidinger[17]	1989	M	15	Permanent mandibular right and left third molars	Enucleation
O'Neil, Mosby, and Lowe[18]	1989	M	5	Permanent mandibular right and left third molars	Enucleation
Banderas and Cols[19]	1996	M	38	Permanent mandibular right and left third molars	Enucleation
Sands and Tocchio[20]	1998	F	3	Primary mandibular right and left central incisors/ Primary mandibular right and left first molars	Enucleation
Ko, Dover, and Jordan[2]	1999	M	42	Permanent mandibular right and left third molars	Enucleation
De Biase and Cols[21]	2001	M	8	Permanent mandibular right and left third molars	Enucleation
Shah, Thuau, and Beale[22]	2002	M	39	Permanent mandibular right and left third molars	No treatment
Ustuner and Cols[8]	2003	M	6	Permanent maxillary right and left canines	Enucleation
Batra and Cols[23]	2004	F	15	Permanent mandibular right and left third molars/second premolar (side not informed)	Enucleation
Freitas and Cols[5]	2006	M	14	Permanent mandibular right and left second and third molars	Enucleation
Cury and Cols*	2008	M	5	Permanent mandibular right and left first molars	Enucleation

**** M=male; F=female***

were maintained, and the deciduous second molars were extracted. Histological analysis of the removed tissue stained with hematoxylin-eosin revealed a cystic wall composed of fibrous tissue with focal mononuclear inflammatory cells lined by a thin stratified squamous nonkeratinized epithelium with 5 to 6 layers of cuboidal cells. Both specimens were diagnosed as DCs (Figures 1d, e, and f).

Close follow-up was planned, and after 12 months the patient showed no evidence of enlargement of the follicular space, indicating lack of recurrence.

LITERATURE REVIEW

To our knowledge, only 18 cases of bilateral DC in nonsyndromic patients have been described in the English dental literature so far (Table 1). Age and gender data were available—the average age was 22 years ranging from 3 to 57 years, but the majority of cases occurred in children or young adults (N=12). There was slight male predilection, with a male:female ratio of 1.57:1. The anatomical location was also available and revealed that most cases (N=14) involved the third molars region, especially in the mandible (N=15). All treatments reported comprised enucleation. This review agrees with findings on a solitary DC.

DISCUSSION

Dentigerous cysts are common developmental cysts and are frequently discovered on radiographs taken to investigate a failure of tooth eruption during orthodontic or general dental examination. There is usually no pain or discomfort associated with the cyst, unless it becomes secondarily infected. Considering a normal follicular space of 3 to 4 mm, a dentigerous cyst should be suspected when the space is more than 5 mm.[7,25]

Differential diagnosis should include radicular cysts, odontogenic keratocysts, ameloblastoma, odontogenic fibromyxoma, and odontomas. Radicular cysts are odontogenic cysts that develop from a periapical granuloma in teeth with carious lesions associated with pulp necrosis. Odontogenic keratocysts are often multilocular and most commonly located in the body or ramus of the mandible. These cysts are lined by stratified squamous keratinized epithelium. Ameloblastoma is the most common radiolucent benign odontogenic tumor that may be unilocular or multilocular. It may cause expansion and destruction of the maxilla and mandible, and histological findings show proliferating epithelial structures resembling "enamel organs." Odontomas are lytic lesions most often accompanied by amorphous calcification. Odontogenic fibromyxoma usually has multiple radiolucent areas of varying size and bony septa, but unilocular lesions have also been described.[9,26-34]

Microscopic evaluation is often necessary to allow definitive diagnosis of these cystic lesions.[4,35]

Reports of bilateral or multiple DCs are extremely rare and are usually associated with developmental syndromes such as mucopolysaccharidosis, basal cell nevus syndrome, and cleidocranial dysplasia.[1,5,6,9]These developmental conditions are usually detected in young individuals.[7]

Mucopolysaccharidoses are a group of diseases resulting from a genetic defect in the degradation of specific mucopolysaccharides. In this syndrome, there is deficiency of N-acetyl-4-sulphatase—resulting in impaired degradation of dermatan sulphate, which accumulates in tissues and is excreted in the urine. Dental features comprise unerupted teeth, dentigerous cysts, malocclusions, condylar defects, and gingival hyperplasia. Other signs and symptoms include kidney and spleen expansion, abnormal bone formation, abnormal auditory and respiratory function, and visual and cardiovascular alterations.[36,37]

Basal cell nevus syndrome is an autosomal dominant disorder characterized by specific developmental malformations associated with predisposition to neoplasia. The major associations include odontogenic keratocysts of the mandible, palmar and plantar pits, characteristic lamellar calcification of the falx cerebri, and skeletal developmental anomalies, particularly of the vertebrae and ribs. Predisposition to neoplasia is present, particularly multiple basal cell carcinomas at a young age.[38]

Cleidocranial dysplasia is a rare syndrome usually caused by an autosomal dominant gene, although 40% of cases appear spontaneously with no apparent genetic cause. This condition is characterized by several cranial malformations and underdevelopment, absence of the clavicles, and multiple supernumerary and impacted permanent teeth. Diagnosis of this condition is usually based on the presence of the main features (supernumerary teeth, partial or total absence of one or both clavicles, and bone malformations) and on clinical and familial evidence.[39]

In our case, a series of examinations was carried out by the pediatric geneticist and no signs of any developmental syndromes were found, indicating that this boy is a nonsyndromic patient. We hope that this paper contributes to the limited clinical knowledge on bilateral DC in nonsyndromic patients.

REFERENCES

1. Weber AL. Imaging of the cysts and odontogenic tumors of the jaw. Definition and classification. Radiol Clin North Am 1993;31;1:101-20.
2. Daley TD, Pringle GA. Relative incidence of odontogenic tumors and oral and jaw cysts in a Canadian population. Oral Sur Oral Med Oral Pathol 1994;77:276-80.
3. Mourshed F. A roentgenographic study of dentigerous cysts. I. Incidence in a population sample. Oral Surg Oral Med Oral Pathol 1964;18:47-53.
4. Motamedi NHK, Talesh KT. Management of extensive dentigerous cysts. Br Dent J 2005;198:203-6.
5. Ko KS-C, Dover DG, Jordan RCK. Bilateral dentigerous cysts: Report of an unusual case and review of the literature. J Can Dent Assoc 1999;65:49-51.

6. Freitas DQ, Tempest LM, Sicoli E, Lopes-Neto FC. Bilateral dentigerous cysts: Review of the literature and report of an unusual case. Dentomaxillofac Radiol 2006;35:464-8.
7. Goaz PW, Stuart CW. Cysts of the jaws. In: *Oral Radiology: Principles and Interpretation.* 3rd ed. St. Louis, Mo: Mosby; 1994:400.
8. Norris L, Piccoli P, Papageorge MB. Multiple dentigerous cysts of the maxilla and the mandible: Report of a case. J Oral Maxillofac Surg 1987;45:694-7.
9. Ustuner E, Fitoz S, Atasoy C, Erden I, Akyar S. Bilateral maxillary dentigerous cysts: A case report. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2003;95:632-5.
10. Myers PB. Bilateral dentigerous cysts of the mandible. Br Dent J 1943;74:67-8.
11. Henefer EP. Bilateral dentigerous cysts of the maxilla: Report of a case. Oral Surg Oral Med Oral Pathol 1964;17:296-8.
12. Stanback JS. The management of bilateral cysts of the mandible. Oral Surg Oral Med Oral Pathol 1970;30:587-91.
13. Callaghan JH. Bilateral impaction of lower third molars in association with bilateral dentigerous cyst formation. A case report. Glasg Dent J 1973;4:36-8.
14. Burton DJ, Scheffer RB. Serratia infection in a patient with bilateral subcondylar impacted third molars and associated dentigerous cysts: Report of case. J Oral Surg 1980;38:135-8.
15. Swerdloff M, Alexander SA, Ceen RF, Ferguson FS. Bilateral mandibular dentigerous cysts in a 7-year-old child. J Pedod 1980;5:77-84.
16. Crinzi RA. Bilateral dentigerous cysts of the mandible. Oral Surg Oral Med Oral Pathol 1982;54:367.
17. McDonnell DG. Bilateral dentigerous cysts. A case history. J Ir Dent Assoc 1988;34:63.
18. Eidinger GB. Bilateral dentigerous cysts in the child patient: Report of a case and review of the literature. Univ Tor Dent J 1989;2:20-3.
19. O'Neil DW, Mosby EL, Lowe JW. Bilateral mandibular dentigerous cysts in a five-year-old child: Report of case. J Dent Child 1989;56:382-4.
20. Banderas JA, Gonzalez M, Ramirez F, Arroyo A. Bilateral mucous cell containing dentigerous cysts of mandibular third molars: Report of an unusual case. Arch Med Res 1996;27:327-9.
21. Sands T, Tocchio C. Multiple dentigerous cysts in a child. Oral Health 1998;88:27-9.
22. De Biase A, Ottolenghi L, Polimeni A, Benvenuto A, Lubrano R, Magliocca FM. Bilateral mandibular cysts associated with cyclosporine use: A case report. Pediatr Nephrol 2001;16:993-5.
23. Shah N, Thuau H, Beale I. Spontaneous regression of bilateral dentigerous cysts associated with impacted mandibular third molars. Br Dent J 2002;192:75-6.
24. Batra P, Roychoudhury A, Balakrishan P, Parkash H. Bilateral dentigerous cyst associated with polymorphism in chromosome 1qh þ. J Clin Pediatr Dent 2004;28:177-81.
25. Farah CS, Savage NW. Pericoronal radiolucencies and the significance of early detection. Aust Dent J 2002;47:262-5.
26. Ricucci D, Mannocci F, Ford TRP. A study of periapical lesions correlating the presence of a radiopaque lamina with histological findings. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2006;101:389-94.
27. Ungari C, Poladas G, Giovannetti F, Carnevale C, Iannetti G. Pindborg tumor in children. J Craniofac Surg 2006;17:365-9.
28. Vencio EF, Mota A, de Melo Pinho C, Dias Filho AA. Odontogenic keratocyst in maxillary sinus with invasive behavior. J Oral Pathol Med 2006;35:249-51.
29. Fregnani ER, da Cruz Perez DE, Soares FA, Alves FA. Synchronous ameloblastoma and orthokeratinized odontogenic cyst of the mandible. J Oral Pathol Med 2006;35:573-5.
30. Gomez Triana M, Luengas Monroy JP. Odontogenic fibromyxoma. Ann Pediatr 2005;62:370-2.
31. Tam D, Francis L, Perry C, Thirwall A. Compound odontoma causing airway obstruction of the newborn: A case report. J Laryngol Otol 2006;120:879-81.
32. Rea G, Di Nuzzo L, Di Pietto F, Cangiano G, Ciccarelli R. Gigantiform cementoma of the jaw. A case report. Radiol Med 2002;104:377-81.
33. Al-Khateeb TH, MScD, Bataineh AB. Pathology associated with impacted mandibular third molars in a group of Jordanians. J Oral Maxillofac Surg 2006;64:1598-602.
34. Taylor SF, Cook AE, Leatherbarrow, B. Review of patients with basal cell nevus syndrome. Ophthal Plast Reconstr Surg 2006;22:259-65.
35. Litonjua LA, Aguirre A, Estrada MC. Radiolucency in the mandible. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2004;98:511-5.
36. Roberts MW, Barton NW, Constantopoulos G, Butler DP, Donahue AH. Occurrence of multiple dentigerous cysts in a patient with the Maroteaux-Lamy syndrome (Mucopolysaccharidosis, type VI). Oral Surg Oral Med Oral Pathol 1984;58:169-75.
37. Dorfman A, Matalon R. The mucopolysaccharidoses (a review). Proc Natl Acad Sci USA 1976;73:630-3.
38. Taylor SF, Cook AE, Leatherbarrow, B. Review of patients with basal cell nevus syndrome. Ophthal Plast Reconstr Surg 2006;22:259-65.
39. Tanaka JLO, Ono E, Médici-Filho E, Castilho JCM, Moraes LC, Moraes MEL, Cleidocranial dysplasia: Importance of radiographic images in diagnosis of the condition. J Oral Sci 2006;48:161-6.