Bilateral dentigerous cyst associated with polymorphism in chromosome 1qh+

Puneet Batra* / Ajoy Roychoudhury** / Prahlad Balakrishan*** / Hari Parkash****

Non-syndromal bilateral dentigerous cysts are rare. It is conceivable that they are either under-recognized or under-reported. In this article we report the unusual occurrence of non-syndromic bilateral dentigerous cysts associated with mandibular third molars with polymorphism in chromosome 1qh+. It is suggested that in non syndromal cases of bilateral dentigerous cysts karyotyping should be done to confirm the association with chromosomal 1 anomaly.
J Clin Pediatr Dent 28(2): 177-182, 2004

INTRODUCTION

Odontogenic cysts arise from an aberration at some stage of odontogenesis.[1] Dentigerous cysts are the second most common odontogenic cysts after radicular cysts, accounting for approximately 24% of all true cysts in the jaws.[2] A dentigerous cyst arises from the enamel organ as a result of an alteration in the reduced enamel epithelium. Though usually associated with the crowns of teeth, it has also been observed in association with other anomalies like odontomas and mostly seen in the second and third decades of life. The teeth most commonly involved are the mandibular third molars and the maxillary cuspids. The incidence is slightly higher in males than in females.[3]

Dentigerous cysts are frequently discovered when radiographs are taken to investigate a failure of tooth eruption, a missing tooth or mal-alignment. There is usually no pain or discomfort associated with the cyst unless it becomes secondarily infected. Radiographs show a unilocular, radiolucent lesion characterized by well-defined sclerotic margins and associated with the crown of an unerupted tooth. While a normal follicular space is 3 to 4mm, a dentigerous cyst can be suspected when the space is more than 5mm.

Most dentigerous cysts are solitary. Bilateral and multiple cysts are usually found in association with a number of syndromes including cleidocranial dysplasia and Maroteaux-Lamy syndrome.[5] In the absence of these syndromes, bilateral dentigerous cysts associated with the mandibular third molars are rare. There have been only few reported cases in the literature (Table I).[6-12] Although this finding may reflect the true rarity of the condition, it is conceivable that bilateral dentigerous cysts are either under-recognized or under-reported. Chromosomal abnormalities are very common, but few reports in literature have reported pathogenic effects associated with dental and skeletal abnormalities. Here, we report the unusual occurrence of nonsyndromic bilateral dentigerous cysts associated with mandibular third molars and left second premolar with polymorphism in chromosome 1qh+.

Case Report

A 15-year-old female patient reported to our unit with a complaint of swelling in the lower left second premolar region. The medical history was non-significant except for a surgery to enucleate a sebaceous cyst from the planter surface of the left-hand three ago.

Intra-oral examination revealed a cystic swelling in relation to a retained deciduous second molar and clinically absent second premolar. The aspirate from the swelling was sent for FNAC, which revealed a large number of anucleate squamous cells with acute inflammatory exudate. Protein estimation of the aspirate was 8.7gm/dl.

The panoramic radiograph showed bilateral, unilocular well-defined corticated radiolucencies surrounding the unerupted second left premolar as well as around the crowns of both mandibular third molars. The anterior border of both radiolucencies around the third molar crowns appeared to involve the distal root

* Puneet Batra MDS, Senior Resident, Department of Dental Surgery, All India Institute of Medical Sciences, New Delhi.
** Ajoy Roychoudhury MDS, Assistant Professor, Department of Dental Surgery, All India Institute of Medical Sciences, New Delhi.
*** Prahlad Balakrishan MSc, Genetic unit, Department of Pediatrics, All India Institute of Medical Sciences, New Delhi.
**** Hari Parkash MDS, FIMFT, FICD, MNAMS, FACD, Professor and Head, Department of Dental Surgery, All India Institute of Medical Sciences, New Delhi.

Send all correspondence to Dr. Ajoy Roychoudhury, Assistant Professor, Department of Dental Surgery, All India Institute of Medical Sciences, New Delhi 110029, India.

Voice: +91-11-26594453
E-mail: ajoy@aiims.ac.in

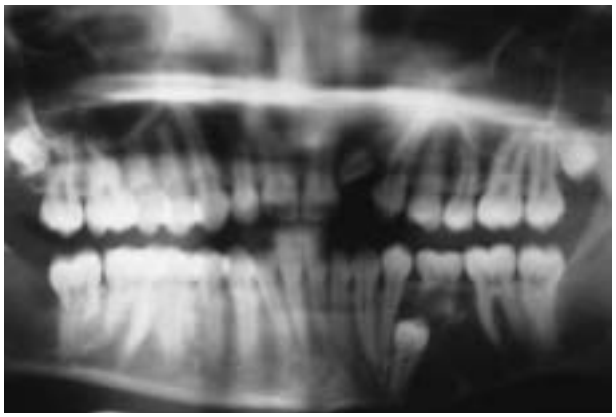

Figure 1. OPG showing the bilateral dentigerous cysts in relation to the mandibular third molars.

of the second molar (Figure 1)**.** Both third molars crowns were grossly displaced to the posterior border of the mandible. Occlusal radiograph of the maxilla did not reveal evidence of clefting (Figure 2).

In view of the multiple dentigerous cysts it was decided to evaluate the patient for any syndromic association. The skeletal survey was done to rule out cysts in any other bones of the skeleton. The chest radiograph revealed an abnormal bony cage with supernumerary ribs in the right lower zone and multiple expanded anterior ends of the ribs on the left side (Figure 3.). The radiograph of the right hand showed a radiolucency in the first metacarpal (Figure 4). Barium meal and enema did not show any intestinal polyposis. No syndromic association could be made.

Peripheral blood lymphoctos were used for the chromosomal analysis Micro culture method of Morehead, *etal*[29] was used and Giemsa-trypsin banding of metaphase chromosome was performed using standard methodology.[30] Cytogenetic studies did not reveal any structural or numerical abnormalities. However, the chromosome 1 was found to have an elongated long arm resulting in 1qh+. (Figure 5)

The cysts were enucleated together with the associated second premolar, retained deciduous second molar as well as third molar teeth bilaterally. Healing was uneventful, and six months after the surgery, the surgical sites showed good healing (Figure 6).

The submitted specimen consisted of three sacs of soft tissue, the largest measuring 32 x 8 x 4mm. Microscopic sections of both specimens were similar, showing cystic walls composed of fibrocollagenous tissue and lined by stratified squamous, non-keratinized epithelium (Figure 7)**.**

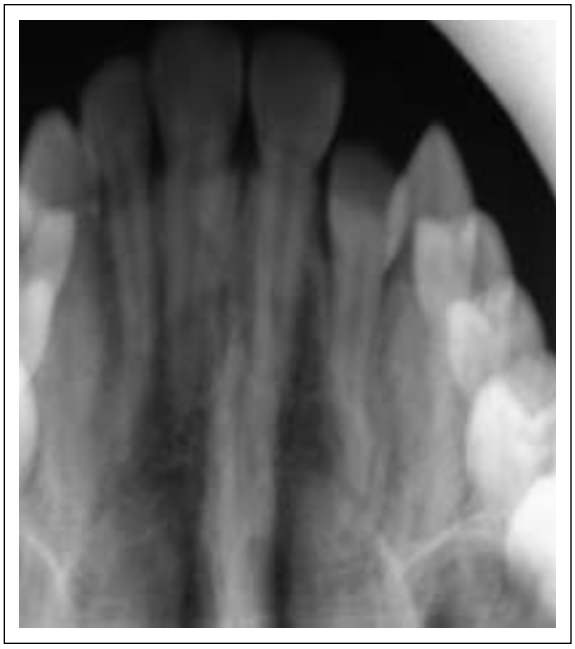

Figure 2. Occlusal radiograph maxilla showing no evidence of clefting.

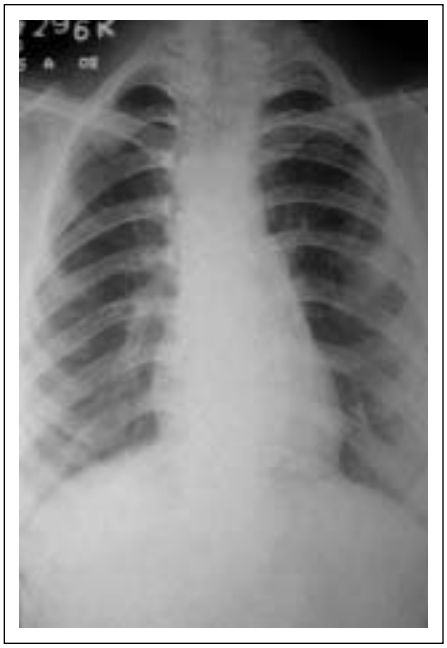

Figure 3. Chest radiograph showing supernumerary ribs in the right lower zone and multiple expanded anterior ends of the ribs on the left side.

DISCUSSION

Anomalies associated with dental and skeletal effects have been reported in literature in association with chromosome 2,4,6,17 and 19.[13-27] These association and linkage studies mostly pertain to non-syndromic cleft lip and palate. Dysplastic teeth with skeletal anomalies in a family have been reported with large secondary constriction in chromosome 1qh+.[28] Karyotyping revealed a large secondary constriction in one of the chromosome 1 pair (1qh+). Chromosome no. 1 polymorphism is very common, but only one report in literature has reported

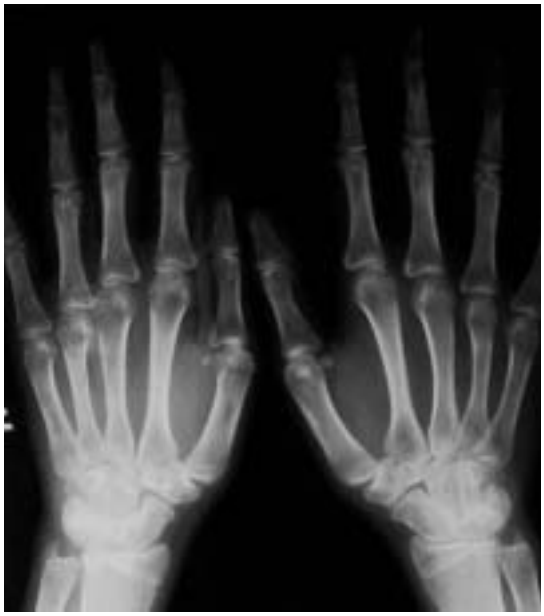

Figure 4 The radiograph of the right hand showed a radiolucency in the first metacarpal.

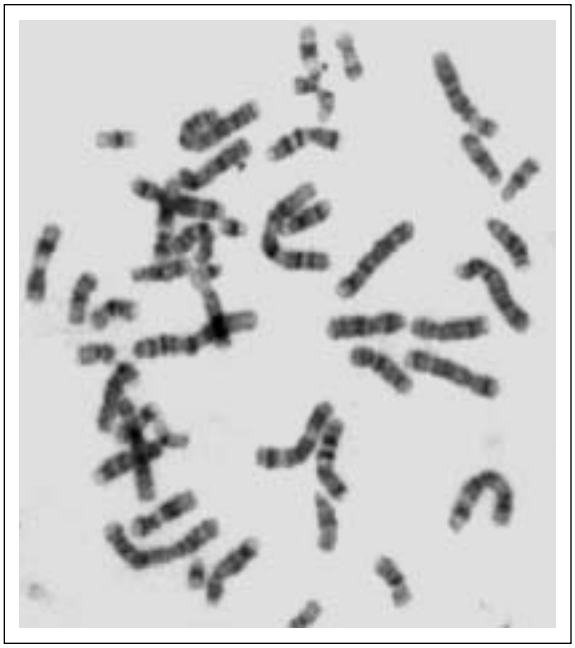

Figure 5. Karyotype revealing a large secondary constriction in one of the chromosome 1 pair (1qh+).

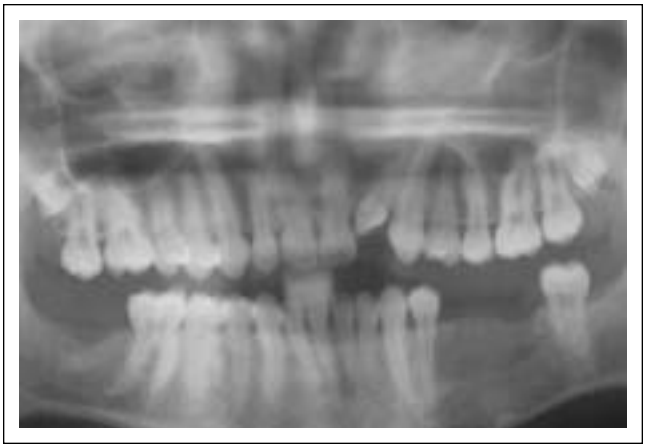

Figure 6. OPG taken six months after the surgical enucleation of the lesion.

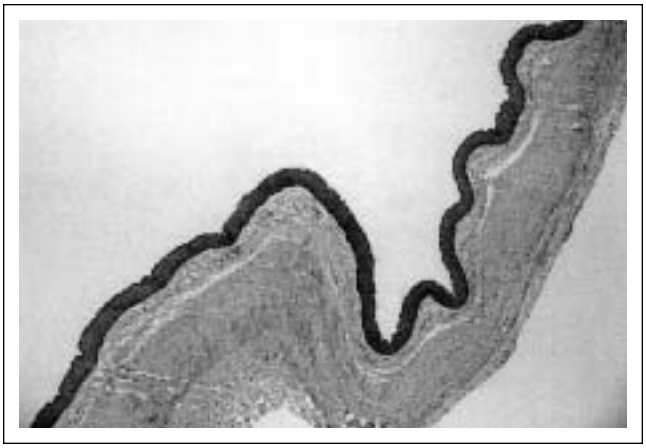

Figure 7. Photomicrograph of dentigerous cyst showing lining of stratified squamous, non-keratinized epithelium. Note the fibromyxoid stroma (H& E stain X 10).

pathogenic effects associated with dental and skeletal abnormalities.[28] It is suggested that in non syndromal cases of bilateral dentigerous cysts karyotyping should be done to confirm the association with chromosomal 1 anomaly or its relation with other chromosomes.

Although dentigerous cysts are common developmental cysts, reported bilateral dentigerous cysts are extremely rare. Bilateral dentigerous cysts are rare in the absence of an underlying syndrome or systemic disease. Bilateral or multiple dentigerous cysts are usually associated with the Maroteaux-Lamy (mucopolysaccharidosis, type VI) syndrome[31] and cleidocranial dysplasia.[32] Both are developmental conditions that are detected in young individuals with stigmata of the syndromes.

Since dentigerous cysts can attain considerable size with minimal or no symptoms, early detection

Table 1. Reported cases of non-syndromal bilateral dentigerous cysts in relation to the mandibular third molars

Authors	Year	Sex	Age (yrs.)	Treatment
Shah N, Thuau H, Beale I[6]	2002	M	39	Spontaneous remission
Ko KS, Dover DG, Jordan RC[7]	1999	M	42	Enucleation
Banderas JA,Gonzalez M, Ramirez F, Arroyo A[8]	1996	M	38	Enucleation
Crinzi[9]	1982	F	15	Enucleation
Burton DJ, Sheffer RB[10]	1980	F	57	Enucleation
Callaghan[11]	1973	M	38	Enucleation
Myers[12]	1943	F	19	Enucleation

and removal of the cysts is important to reduce morbidity.[33] It is therefore important to perform radiographic examination of all unerupted teeth. Bone scans may be useful in detecting multiple lesions throughout the body.[34] Although multiple dentigerous cysts are rare, once a cystic lesion is recognized, the patient must be examined carefully to rule out other possible dentigerous cysts.[35,36] Complete removal of the lesion is essential given that dentigerous cysts or the remnants have been documented to differentiate into ameloblastomas, squamous cell carcinomas, adenomatoid odontogenic tumors, and complex odontomas.[37-39]

REFERENCES

1. Shafer WG, Hine KM, Levy BM. A textbook of oral pathology, 4th edition, Philadelphia, WB Saunders Co, pg 260-65, 1983.
2. Mourshed F. A roentgenographic study of dentigerous cysts. I. Incidence in a population sample. Oral Surg Oral Med Oral Pathol 18: 47-53, 1964.
3. Bhaskar SN. Synopsis of Oral Pathology, 3rd edition, St Louis, The C.V. Mosby Company, pg. 206, 1969.
4. Goaz PW, Stuart CW. Cysts of the jaws. In: Oral radiology, principles and interpretation. 3rd ed. St. Louis, The Mosby Company, 1994.
5. Gorlin RJ. Cysts of the jaws, oral floor and neck. In: Gorlin RJ, Goodman HW, editors. Thoma's oral pathology. 6th ed. St. Louis, Mosby Company, Vol. 1, 1970
6. Shah N, Thuau H, Beale I. Spontaneous regression of bilateral dentigerous cysts associated with impacted mandibular third molars. Br Dent J 192: 75-6, 2002.
7. Ko KS, Dover DG, Jordan RC. Bilateral dentigerous cysts—report of an unusual case and review of the literature. J Can Dent Assoc 65: 49-51, 1999.
8. Banderas JA, Gonzalez MA, Ramirez F, Arroyo A. Bilateral mucous cell containing dentigerous cysts of mandibular third molars: Report of an unusual case. Arch Med Res 27: 327-9, 1996.
9. Crinzi RA. Bilateral dentigerous cysts of the mandible. Oral Surg Oral Med Oral Pathol 54: 367, 1982.
10. Burton DJ, Sheffer RB. Serratia infection in a patient with bilateral subcondylar impacted third molars and associated dentigerous cysts: Report of a case. J Oral Surg 38: 135-8, 1980.
11. Callaghan JH. Bilateral impaction of lower third molars in association with bilateral dentigerous cyst formation. A case report. Glasgow Dent J 4: 36-8, 1973.
12. Myers PB. Bilateral dentigerous cysts of the mandible. Brit Dent J 74: 67-8, 1947.
13. Holder SE, Vintiner GM, Farren B, Malcolm S and Winter RM. Confirmation of an association between RFLP's at the transforming growth factor alpha locus and non-syndromic cleft lip and palate. J Med Genetics 29: 390-92, 1992.
14. Feng H, Sassani R, Bartlett SP, Lee A, Hecht JT, Malcolm S, Winter RM, Vintiner GM, Buetow KH, Gasser DL. Evidence from family studies, for linkage disequilibrium between TGFA and a gene for non syndromic cleft lip with or without palate. Am J Hum Genetics 55: 932-936, 1994.
15. Mitchell LE. Transforming growth factor alpha locus and non syndromic cleft lip with or without cleft palate; a reappraisal. Genet Epidemiol 14: 231-240, 1997.
16. Beiraghi S, Foround T, Dlouty S, Bixler D, Delohter- Blanchet D, Conneally PM, Hodes ME. Possible localization of a major gene for cleft lip and palate to 4q. Clin Genet 46: 255-256, 1994.
17. Mitchell LE, Healey SC, Chenevix- Trench G. Evidence for an association between non-syndromic cleft lip with or with out cleft palate and a gene located on the long arm of the chromosome 4. Am J Hum Genetics 57: 1130-1136, 1995.
18. Eiberg H, Bixler D, Nielsen LS, Conneally PM, Mohr J. Suggestion of linkage of a major locus for non syndromic orofacial cleft with F13A and tentative assignment to chromosome. Clin Genet 32: 129-32, 1987.
19. Korman-Bortolotto MH, Farah LMS, Soares D, Corbani M, Muller R, Adell ACA. Terminal deletion 6p23; a case report. Am J Med Genetics 37: 475-477, 1990.
20. Donnai D, Heather LJ, Sinclair P, Thakker Y, Scambler P, Dixon MJ. Association of autosomal dominant cleft lip palate and translocation 6p23;9q22.3. Clin Dysmorph 1: 89-97, 1992.
21. Davies AF, Stephens RJ, Olavesen MG, Heather L, Dixon MJ, Magee A, Flinter F and Ragoussis J. Evidence of a locus for orofacial clefting on human chromosome 6p24 and STS content map of the region. Hum Mol Genetics 4: 121-128, 1995.
22. Carinci F, Pezzeti F, Scapoli L, Padula E, Baciliero U, Curioni C, Tognon M. Non syndromic cleft lip and palate: evidence of linkage to microsatellite markers on 6p23. Am J Hum Genetics 56: 337-339, 1995.
23. Scapoli L, Pezzetti F, Carinci F, Martinelli M, Carinci P, Tognon M. Genetic heterogeneity and evidence of linkage to 6p23 in non syndromal cleft lip with or without cleft palate. Genomics 43: 216-220. 1997.
24. Stein J, Mulliken JB, Stal S, Gasser DL, Malcolm S, Winter R, Blanton SH, Amos C, Seemanova E, Hecht JT. Non syndromic cleft lip with or without cleft palate: evidence of linkage to BCL3 in 17 multigenerational families. Am J Hum Genetics 57:257-272, 1995.
25. Amos C, Gasser D, Hecht JT. Non syndromic cleft lip with or without cleft palate: new BCL3 information. Am J Hum Genetics 59: 743-744, 1996.
26. Wyszynski DF, Maestri N, McIntosh I, Smith EA, Lewanda AF, Garcia-Delgado C, Vinageras-Guarneros E, Wulfsberg E, Beaty TH. Evidence for an association between markers on chromosome 19q and non syndromal cleft lip with or without cleft palate in two groups of multiplex families. Hum Genetics ;9: 22-26, 1997.
27. Martinelli M, Scapoli L, Pezzetti F, Carinci F, Carinci P, Baciliero U, Padula E, Tognon M. Suggestive linkage between markers on chromosome 19q13.2 and non syndromic orofacial cleft malformation. Genomics 51: 177-181, 1998.

28. Garan A, Nurchi AM, Melis P, Frau G, Costa G. Syndactylia associated with multiple malformation syndromes. Observation of a new symptomatological complex in three brothers. Pediatr Med Chir 10: 227-32, 1988.
29. Moorehead PS, Nowell PC, Mellman WJ, Battips DM, Hungerford DA. Chromosome preparations of leukocytes cultured from human perepheral blood. Exp Cell Res 202: 613-16, 1960.
30. Searbright M. A rapid banding technique for human chromosome. Lancet 2: 971-2, 1971.
31. 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 58: 169-75, 1984.
32. Trimble LD, West RA, McNeill RW. Cleidocranial dysplasia. Comprehensive treatment of the dentofacial abnormalities. J Am Dent Assoc 105: 661-6, 1982.
33. Swerdloff M, Alexander SA, Ceen, RF, Ferguson FS. Bilateral mandibular dentigerous cysts in a seven-year-old child. J Pedod 5: 77-84, 1980.
34. Josell SD, Reiskin AB, Gross BD. Dentigerous cyst with mural ameloblastoma. JADA 99: 634-636, 1979.
35. Carr MM, Anderson RD, Clarke KD. Multiple dentigerous cysts in childhood. J Otolaryngol 25: 267-270, 1996.
36. Toller MO, Sipahier M, Acikgoz A. CT display of multiple dentigerous cysts of the mandible: A case report. J Clin Pediatr Dent 19: 135-137, 1995.
37. Gardner AF. A survey of odontogenic cysts and their relationship to squamous cell carcinoma. J Can Dent Assoc 41: 161-167, 1975.
38. Lee KW, Loke SJ. Squamous cell carcinoma arising in a dentigerous cyst. Cancer 20: 2241-2244, 1967.
39. Valderrama LS. Dentigerous cyst with intracystic adenomatoid odontogenic tumor and complex odontoma. J Philipp Dent Assoc 41: 35-41, 1989.