

Non-syndromic bilateral dentigerous cysts of mandibular premolars: a rare case and review of literature

Sumita Mahajan *, MDS

Vineet Raj *, BDS

Karen Boaz *, MDS

Thomas George *, MDS

ABSTRACT Dentigerous cysts are developmental cysts of odontogenic origin, which surround the crown of unerupted teeth, odontomas, or supernumerary teeth. Usually single in occurrence, their bilateral presentation is rare, especially in the absence of syndromes like Maroteaux-Lamy, basal cell nevus or cleidocranial dysplasia. Non-syndromic bilateral cysts show a predilection for the mandible, especially the molar area. Presentation in any other location is extremely rare. We describe a case of a 13-year-old child presenting with bilateral radiolucencies in relation to the crowns of impacted mandibular premolars that were diagnosed histopathologically as dentigerous cysts which is a unique presentation in the absence of any syndrome. We also review the 16 previously reported cases of such rare non-syndromic bilateral dentigerous cysts.

Introduction

Dentigerous cyst is a developmental anomaly of odontogenic origin, which surrounds the crown of an impacted tooth, an odontoma, or a supernumerary tooth. It is formed by the accumulation of fluid between the reduced enamel epithelium and the crown, with consequent expansion of the tooth follicle, and is characteristically attached to the cervical area of the tooth. Although it may be encountered in any location of the jaw, most frequently it is seen in relation to the mandibular molars followed by maxillary canines. In many instances the cyst may be asymptomatic till it attains a large size. It usually presents as a slowly enlarging, sometimes painful swelling, particularly if infected. Radiographically it appears as a well-defined radiolucency, usually with

sclerotic borders, associated with the crown of an unerupted tooth ¹.

Although dentigerous cysts are by far the most common non-inflammatory cysts in the oral region, their bilateral occurrence is extremely rare especially in the absence of any underlying systemic disease or syndromes ². We present a rare case of non-syndromic bilateral dentigerous cysts in relation to mandibular premolars.

Case report

A 13-year-old male reported to the Department of Orthodontics complaining of malaligned teeth. Intraoral examination revealed diffuse hard swellings affecting the body of the mandible on both sides with obliteration of the buccal sulcus. Teeth numbers 33, 35, 43, 44, and 45 were clinically absent. Teeth numbers 73, 84, and 85 were over-retained. The panoramic radiograph revealed a unilocular well-defined radiolucency associated with an impacted mandibular left second premolar. There was also a multilocular radiolucency associated with impacted teeth numbers 43, 44, and 45 involving the mandibular body and extending up to the symphysis anteriorly and mesial to the unerupted third molar

* Department of Oral Pathology and Microbiology, Manipal College of Dental Sciences, Mangalore, India

Correspondence to:

Dr. Vineet Raj

Department of Oral Pathology and Microbiology, Manipal College of Dental Sciences, Light House Hill Road, Mangalore 575001, Karnataka, India

Tel : (91-824) 2428 716

Fax : (91-824) 2422 653

e-mail : vineetraj@yahoo.com



Figure 1 Orthopantomograph showing bilateral radiolucencies (arrows) in relation to impacted mandibular left premolar and mandibular right canine, first and second premolars

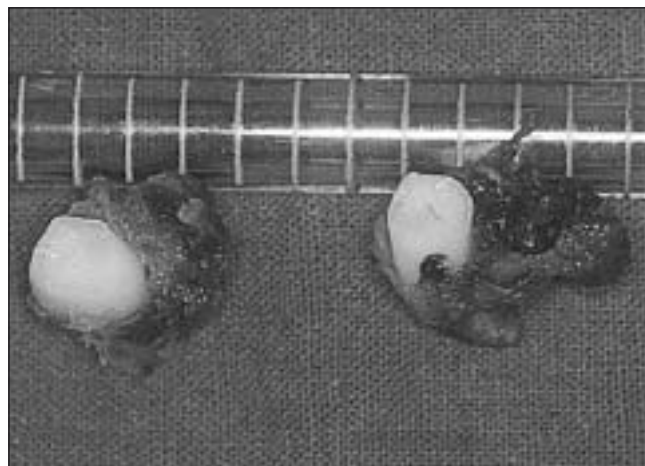


Figure 2 Gross picture of the specimen showing cystic sacs attached to mandibular left second and mandibular right first premolars

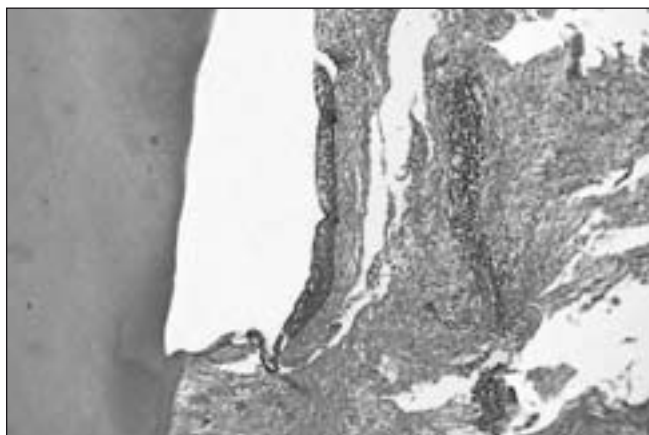


Figure 3 Photomicrograph showing 3-5 layers of epithelium resembling reduced enamel epithelium lining the cystic capsule (H&E, x4)

posteriorly. The permanent mandibular left canine was missing in the radiograph (Figure 1). The medical history was non-contributory. No other abnormality was detected on complete systemic examination. Chest radiograph, serum calcium and phosphate levels, and other laboratory findings were normal.

Fine-needle aspiration of the cysts yielded a straw-colored fluid from the right side and a blood-tinged fluid from the left side. Microscopic examination of the fluid showed polymorphonuclear leukocytes, eosinophils, histiocytes, foam cells, and a few epithelial cells. Incisional biopsy from the right side showed 2-6 layers of non-keratinized stratified squamous epithelium with ex-

cytosis and intercellular edema overlying a collagenous connective tissue and a dense infiltrate of acute and chronic inflammatory cells. A diagnosis of an infected cyst was made.

The patient underwent enucleation of both the cysts under general anesthesia along with removal of associated impacted teeth, and the specimens were submitted for histopathological examination.

Grossly they consisted of two cystic sacs attached to the cervical area of teeth numbers 35 and 44 (Figure 2). The specimens were sectioned so as to facilitate subsequent microscopic examination of the relationship of the cystic sacs to the teeth. Later they were fixed for 24 hours and then decalcified. On histopathological examination the cystic lumina were lined by 3-5 layers of non-keratinized epithelium, resembling reduced enamel epithelium (Figure 3), with focal areas of proliferation. The connective tissue was myxomatous to fibrous with a dense infiltrate of acute and chronic inflammatory cells in some areas. A few islands of odontogenic epithelium were also seen within the connective tissue (Figure 4). Decalcified sections of teeth showed epithelium, similar to that lining the cystic lumen, attached at the cemento-enamel junction (Figure 5). Based on these findings a diagnosis of bilateral dentigerous cysts in relation to teeth numbers 35 and 44 was made.

Discussion

After radicular cysts, the second most common are

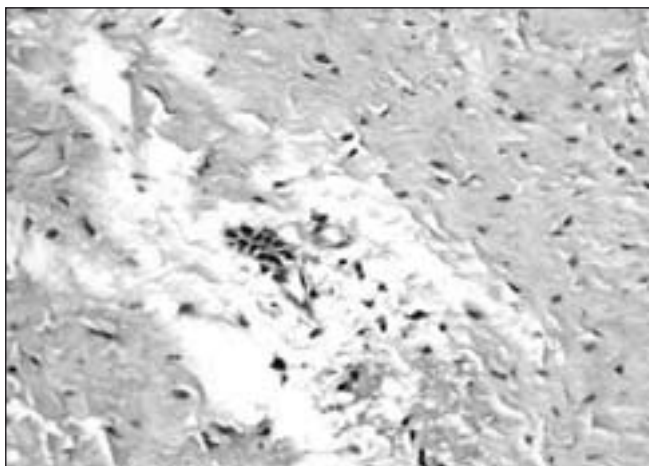


Figure 4 Photomicrograph showing an odontogenic island within the connective tissue wall of the cyst (H&E, x10)

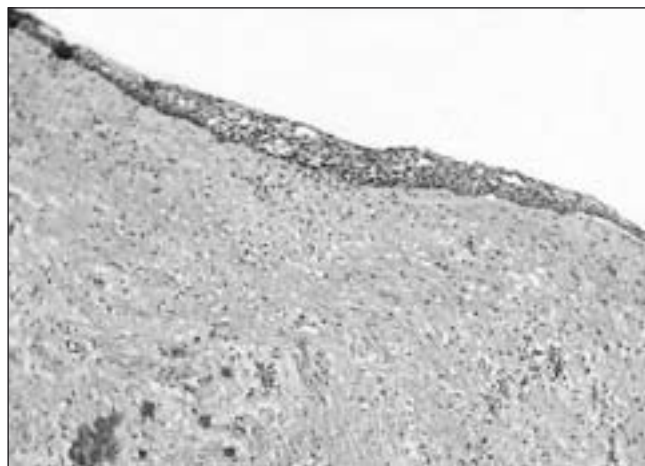


Figure 5 Photomicrograph showing cystic lining attached to the cemento-enamel junction (H&E, x4)

dentigerous cysts of odontogenic origin and account for about 16.6% of all such jaw lesions. They occur more frequently in males (male to female ratio about 1.6:1)¹, are usually asymptomatic but can become extremely large and cause cortical expansion and erosion.

Radiographically, the dentigerous cyst appears as a unilocular radiolucency of variable size with well-defined sclerotic borders, associated with the crown of an unerupted tooth. In an infected cyst the borders may be ill-defined. There may be difficulty distinguishing a small cyst from a normal tooth follicle. It has been suggested that any follicular space of >4 mm should be suspected to be a dentigerous cyst². The radiographic appearance of such a cyst, though quite typical, is not diagnostic. Other lesions may mimic their radiographic appearance including: odontogenic keratocyst, radicular cyst, and some odontogenic tumors like ameloblastoma, Pindborg's tumor, adenomatoid odontogenic tumor, calcifying odontogenic cyst, and ameloblastic fibroma. In most instances, microscopic evaluation is therefore necessary to reach a definitive diagnosis.

Histologically, the dentigerous cyst displays a thin fibrous cyst wall with a myxomatous appearance. The epithelial lining consists of 2-4 layers of flat or cuboidal cells, which in fact is the reduced enamel epithelium and is characteristically non-keratinized. Nests, islands or strands of odontogenic epithelium are often seen in the fibrous capsule. Localized proliferation of epithelial lining may occur in response to inflammation. Hyaline (Rushton) bodies may be found in the epithelium, especially in cysts exhibiting inflammation. Sometimes mucous se-

creting cells and rarely ciliated cells form a part of the epithelial lining, and occasionally sebaceous cells and lymphoid follicles with germinal centers are seen in the connective tissue^{1,3}.

The treatment of a dentigerous cyst is usually dictated by the size of the lesion. If small, it can be enucleated, but marsupialization may be needed for the complete removal of a large cyst. Every effort should be made to allow the involved tooth to erupt, provided the path of eruption is favorable. Recurrence of a dentigerous cyst is rare and could be due to residual fragments of cyst lining.

Although dentigerous cysts are commonly encountered, their bilateral occurrence has rarely been reported, especially in the absence of associated systemic disease or syndromes⁴. Bilateral or multiple dentigerous cysts are usually encountered with Maroteaux-Lamy syndrome⁵, and cleidocranial dysplasia or after prolonged concurrent use of cyclosporine, and calcium channel blockers⁶. In our patient, these associations were ruled out by the absence of any skeletal anomalies, abnormal laboratory findings and clinical history.

Bilateral dentigerous cysts in the absence of any syndrome were first reported by Myers⁷. Thereafter, only 16 cases have been individually reported in the English literature by different authors (Table)^{4,6-20}, which re-emphasizes the rarity of the condition.

The age distribution of patients presenting with bilateral dentigerous cysts ranges from first to sixth decade; almost two thirds do so in the first and second

Table Summary of previously reported patients with non-syndromic bilateral dentigerous cysts in English literature ^{4,6-20}

Author	Sex	Age (years)	Maxilla/Mandible	Location	Treatment
Myers (1947) ⁷	F	19	Mandible	Third molars	Enucleation
Stanback (1970) ⁸	M	9	Mandible	First molars	Enucleation
Callaghan (1973) ⁹	M	38	Mandible	Third molars	Enucleation
Burton and Scheffer (1980) ¹⁰	F	57	Mandible	Third molars	Enucleation
Swerdloff <i>et al</i> (1980) ¹¹	F	7	Mandible	First molars	Enucleation
Crinzi (1982) ¹²	F	15	Mandible	Third molars	Enucleation
McDonnell (1988) ¹³	M	15	Mandible	Second premolar and second molar	Enucleation
Eidinger (1989) ¹⁴	M	15	Mandible	First molars	Enucleation
O'Neil <i>et al</i> (1989) ¹⁵	M	5	Mandible	First molars	Enucleation
Toller <i>et al</i> (1995) ¹⁶	M	13	Mandible	Molars and canines	Enucleation following marsupialization
Banderas <i>et al</i> (1996) ¹⁷	M	38	Mandible	Third molars	Enucleation
Sands and Tocchio (1998) ¹⁸	F	3	Mandible	Central incisors and first molars	Enucleation
Ko <i>et al</i> (1999) ⁴	M	42	Mandible	Third molars	Enucleation
Shah <i>et al</i> (2002) ¹⁹	M	39	Mandible	Third molars	No treatment
Ustuner <i>et al</i> (2003) ⁶	M	6	Maxilla	Canines	Enucleation
Batra <i>et al</i> (2004) ²⁰	F	15	Mandible	Third molars and left second premolar	Enucleation

decades. This is in contrast to conventional dentigerous cysts, which typically present between the second and fourth decade ¹. As for the sex predilection in unilateral cysts ¹, they are more common in males in about a 2:1 ratio. However, the number of reported cases of bilateral cysts is too small to form any reliable conclusions about their age and sex distribution.

The most common site of presentation for unilateral dentigerous cysts is the mandibular molar area ¹, and the same trend is apparent for bilateral cysts; occurrence in other locations is extremely rare. Bilateral involvement of the premolar region has not yet been reported. A review of literature of cases of bilateral dentigerous cysts revealed premolar involvement to be unilateral only, with contralateral side showing involvement of molars ^{13,20}. We believe ours is the first report in the English literature of a case of bilateral dentigerous cysts involving only premolars.

Chromosomal polymorphism has been reported in one patient with bilateral dentigerous cysts, where karyotyping revealed a large secondary constriction in chromosome 1qh+ ²⁰. Although chromosome 1 polymorphism is very common, there is only one report of pathogenic effects associated with dental and skeletal abnormalities ²⁰.

The prognosis of non-syndromic bilateral dentigerous cysts does not differ from that of a unilateral cyst. All

of the previous patients had simple enucleation of their cysts and none were reported to have recurred. Shah *et al* ¹⁹ even suspected spontaneous regression of these cysts after 3 years, without any intervention. Arguably though, disappearance of the radiolucencies (as reported by the authors) may have been due to faulty angulation of the follow-up radiograph ²¹. The present case was treated by enucleation and no recurrence was evident during the 1-year follow-up period.

Conclusion

The frequency of dentigerous cysts in children may be higher than is generally acknowledged, as they may be underreported due to their asymptomatic nature. These cysts are usually slow-growing lesions and may attain a considerable size with minimal or no symptoms. Ameloblastoma, mucoepidermoid carcinoma, and squamous cell carcinoma have also been reported to arise from the lining epithelium of dentigerous cysts, indicating the pluripotentiality of their cells ⁶. Early detection and removal of such cysts is therefore important to reduce potential morbidity.

As it is common to find an unerupted tooth as the only initial presenting symptom of a dentigerous cyst, it is important to undertake radiographic examinations of all such teeth that are well past their expected eruption date. Despite the rarity of bilateral occurrence, once a

dentigerous cyst has been identified, attempts must be made to rule out the presence of any co-existent lesions in other parts of the jaws.

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