Case Report

Multiple dentigerous cysts in a nonsyndromic minor patient: Report of an unusual case

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ABSTRACT

A dentigerous cyst is an odontogenic cyst, thought to be of developmental origin. It is associated with the crown of an unerupted (or partially erupted) tooth. The cyst cavity is lined by epithelial cells derived from the reduced enamel epithelium of the tooth forming organ. Most dentigerous cysts are solitary. Multiple/bilateral cysts are usually found in association with a number of syndromes including cleidocranial dysplasia, Maroteaux-Lamy syndrome and Gorlin--Goltz syndrome (multiple OKCs). In the absence of these syndromes, the occurrence of multiple dentigerous cysts is rare. Here, we report the unusual occurrence of non-syndromic multiple dentigerous cysts.

Key words: Dentigerous cyst, enucleation, unerupted teeth

Introduction

A dentigerous cyst is an epithelial-lined developmental cavity that encloses the crown of an unerupted tooth at the cementoenamel junction. Dentigerous cysts are the second most common odontogenic cysts after radicular cysts, accounting for approximately 24% of all true cysts in the jaws. Their frequency in the general population has been estimated to be 1.44 cyst for every 100 unerupted teeth. The cyst arises from the separation of the follicle from the crown of an unerupted tooth and although it may involve any tooth, the mandibular third molars are the most commonly affected. Dentigerous cysts are frequently discovered when radiographs are taken to investigate a failure of tooth eruption, a missing tooth or malalignment. 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 4 mm, a dentigerous cyst can be suspected when the space is more than 5 mm.

Most dentigerous cysts are solitary. Multiple/bilateral cysts are usually found in association with a number of syndromes including cleidocranial dysplasia, Maroteaux--Lamy syndrome and Gorlin--Goltz syndrome. In the absence of these syndromes, the occurrence of multiple dentigerous cysts is rare.[1-5]

Here, we report the unusual occurrence of non-syndromic multiple dentigerous cysts [Table 1].

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Materials and Methods

A 22 year male was referred to the Department of Oral and Maxillofacial Surgery from the department of Oral Medicine for the evaluation of an asymptomatic, cystic lesion in all the four quadrants. Intraoral examination revealed many missing teeth in all the quadrants. No extra-oral swellings or tenderness was noted. The patient's medical history was non-significant. There were no associated syndromes present.

Table 1: Reported cases till year 2000

Authors	Year	Sex	Race	Age (years)	Location	Treatment
Sands and Tocchio	1998	F	N/A	3	Md. central incisors and first molars	Enucleation
Banderas and others	1996	M	C	38	Md. third molars	Enucleation
O'Neil and others[6]	1989	M	Bl	5	Md. first molars	Enucleation
Eidinger[7]	1989	M	C	15	Md. first molars	Enucleation
McDonnell[8]	1988	M	N/A	15	Md. second premolar and second molar teeth	Enucleation
Crinzi[9]	1982	F	Bl	15	Md. third molars	Enucleation
Swerdloff and others[10]	1980	F	C	7	Md. first molars	Enucleation
Burton and others[11]	1980	F	Bl	57	Md. third molars	Enucleation
Callaghan[12]	1973	M	C	38	Md. third molars	Enucleation
Stanback[13]	1970	M	N/A	9	Md. first molars	Enucleation
Myers[1]	1943	F	N/A	19	Md. third molars	Enucleation

N/A= not available, M= male, C = Caucasian, F= female, Bl = black; Md. = mandibular, Credit- J Can Dent Assoc 1999; 65:49-51

The investigations done were FNAC, O.P.G, IOPAR and routine blood investigations [Figure 1].

A panoramic radiograph showed missing teeth in all the four quadrants. Unilocular well-defined corticated radiolucencies surrounding all the missing teeth were identified. Missing teeth associated with cyst were 15,12,11,21,22,25,33,35,43,44,45 [Figures 2-5].

The surgical removal of the cysts of the lower quadrant was performed under conscious sedation and the upper quadrants were planned and performed in hospital under general anesthesia and the material was analyzed. Pathological analysis of the superior lesions showed a cyst wall composed of fibrous tissue and lined by non-keratinized stratified squamous epithelium. Pathological analysis of the inferior lesions showed an inflamed cyst with a thicker epithelial lining with rete ridges and fibrous capsule with a diffuse chronic inflammatory infiltrate. The final diagnosis of all the lesions was a dentigerous cyst.

Results

The submitted specimen consisted of two sacs of soft tissue, the largest measuring 20 × 4 mm. Microscopic sections of both specimens were similar, showing cyst walls composed of fibrous tissue and lined by stratified squamous, non-keratinized epithelium with Rushton bodies.

These features were suggestive of dentigerous cysts [Figures 6 and 7].

Discussion

Dentigerous cysts are very common developmental cysts and they are generally solitary. Bilateral dentigerous cysts usually occur in association with syndromes like mucopolysaccharidosis (type VI) and cleidocranial dysplasia. Both diseases cause alterations in tooth development or in their eruption. These conditions may participate in the development of multiple dentigerous cysts.

Maroteaux-Lamy syndrome is one of the mucopolysaccharidoses (MPS), a group of diseases resulting from a genetic defect in the degradation of specific mucopolysaccharides. With this syndrome, there is a deficiency of *N*-acetyl-4-sulfatase that results in impaired degradation of dermatan sulfate, which accumulates in tissues and is excreted in the urine. Dental features include unerupted dentition, dentigerous cysts, malocclusions, condylar defects and gingival hyperplasia.[14]

Cleidocranial dysplasia is an autosomal dominantly inherited disorder that results in a partial or complete absence of clavicles, short stature, frontal and parietal bossing, maxillary micrognathia, prolonged retention of the primary dentition, delayed eruption of the permanent dentition and unerupted supernumerary teeth. Multiple dentigerous cyst formation occurs in both conditions and can develop at any site in the upper or lower jaws.[15]

Gorlin-Goltz syndrome is autosomal dominant with a high penetrance and variable expressivity. It is caused by mutations in the patched tumor suppressor gene (PTCH), a human homologue of the Drosophila gene mapped to chromosome 9q21-23. Chromosomal mapping and genetic studies suggest that the underlying basis for this disease is an abnormality in the Hedgehog (Hh) signaling pathway. Gorlin--Goltz syndrome, also known as basal cell nevus syndrome, is an uncommon disorder, which is characterized by numerous basal cell carcinomas (seen in 50–97% of people with the syndrome), maxillary keratocysts (present in about 75% of patients) and musculoskeletal malformations.[16]

Multiple/bilateral dentigerous cysts are extremely rare in the absence of a syndrome or systemic disease. After searching the literature, only 17 cases were identified from 1943 to 2005. The age range for reported cases

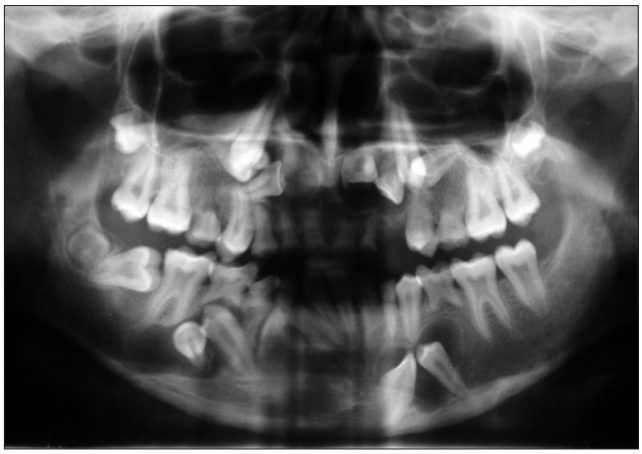

Figure 1: Pre-operative O.P.G

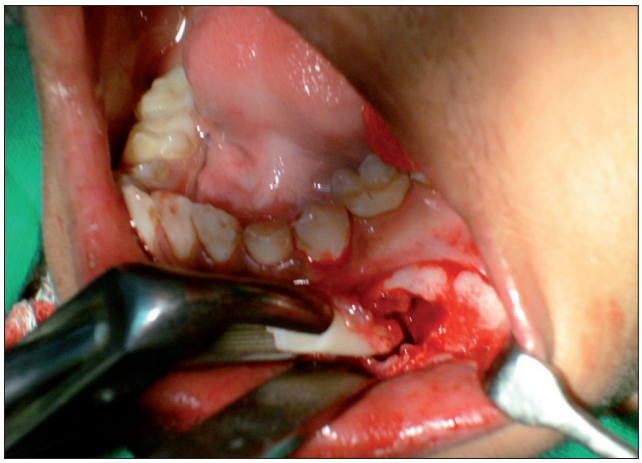

Figure 2: Lower left quadrant

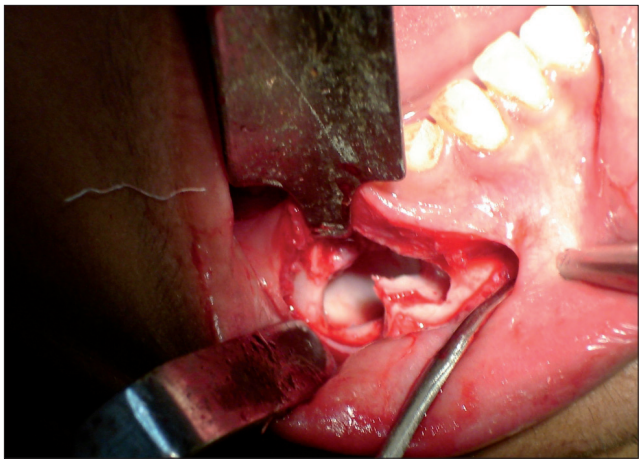

Figure 3: Lower right quadrant

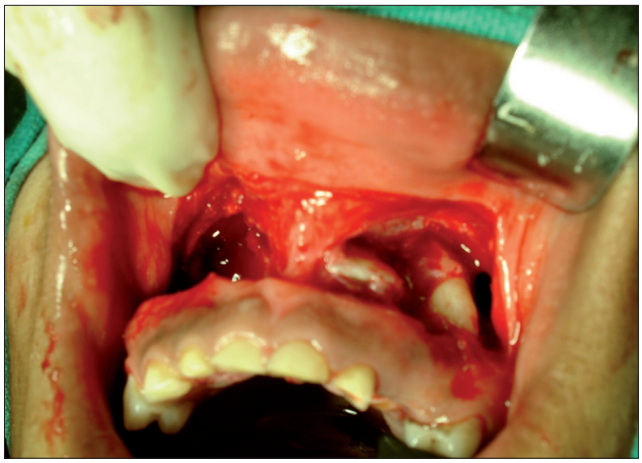

Figure 4: Upper quadrants

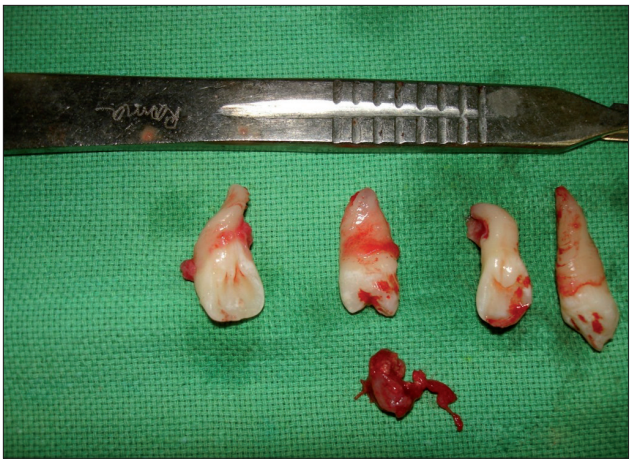

Figure 5: Extracted teeth with a lesion

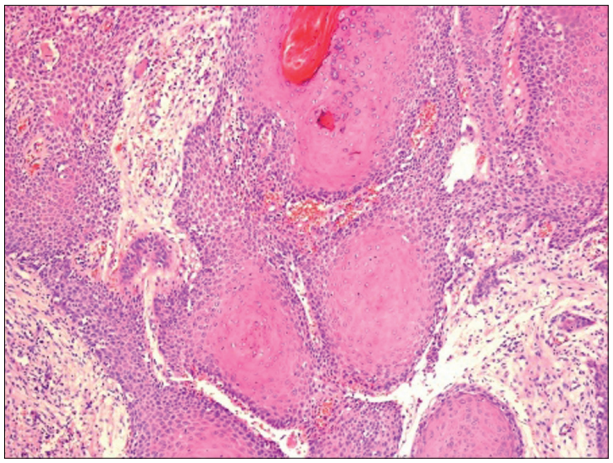

Figure 6: Microscopic section.

varies widely, from 3 years to 57 years of age. The mean age of the 17 cases was 22.5 years. Ten of them occurred in children under the age of 15 years.[1-3]

Patients frequently present unerupted teeth or asymptomatic slow-growing swellings. As in this case, all but one of the reported cases occurred in asymptomatic individuals. In the present case, in addition to asymptomatic swelling, the patient also presented any unerupted teeth.

In all reported cases, including the present case,

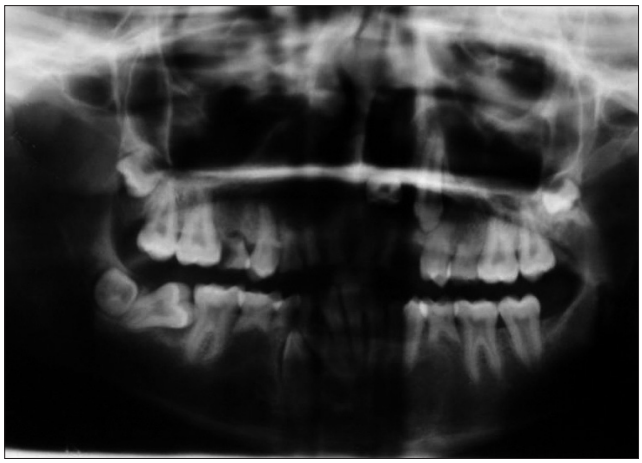

Figure 7: Post-operative O.P.G.

radiographic examination showed a unilocular radiolucent lesion associated with the crown of an unerupted tooth and well-defined sclerotic margins.

It is important to perform radiographic examinations in cases of unerupted teeth. Initially a panoramic radiograph may be used for this examination. However, in cases of extensive lesion, CT imaging becomes necessary. CT imaging gives information about origin, size, content, cortical plates and relationship of the lesion to adjacent anatomical structures. Initially, a panoramic radiograph was evaluated in the present case. Only in a panoramic radiograph was it possible to observe a second lesion associated with the right maxillary third molar, since the lesion had not yet caused swelling. Thus early detection is only possible with radiographic examination of unerupted teeth.

Radiographic examinations provide valuable information. However, pathological analysis of the lesion is essential for the definitive diagnosis. Other lesions may share the same radiological features as dentigerous cysts, such as odontogenic keratocysts and unicystic ameloblastoma. In this case, these lesions were included in the differential diagnosis after the radiographies were observed. Although involvement of the tooth, cortical expansion and radicular reabsorption are characteristics more related to dentigerous cysts, other lesions were not excluded until the results of the pathological analysis were known. Odontogenic keratocysts do not expand the bone to the same degree as dentigerous cysts and are less likely to produce teeth resorption. According to Tsukamoto et al, the mean age of patients with odontogenic keratocyst was less than that of patients with a dentigerous cyst; the mean area of the odontogenic keratocysts was larger than that of dentigerous cysts; and dentigerous cysts are more likely to have smooth periphery and odontogenic keratocysts are more likely to have scalloped periphery. It is not possible to differentiate unicystic ameloblastomas from dentigerous cysts with clinical and radiographic examinations.

All lesions together with the associated tooth were enucleated in hospital under general anesthesia/ conscious sedation when pathological analysis was obtained. Enucleation was the treatment in 16 of 17 reported cases, although larger lesions may be surgically drained and marsupialized to relieve the pressure within the cysts and to prevent damage to the involved permanent teeth. The recurrence of Dentigerous cysts is very rare.[7]

Conclusion

Multiple/bilateral dentigerous cysts are extremely rare in the absence of a syndrome or systemic disease. After searching the literature, only 17 cases were identified from 1943 to 2005.

Appropriate and thorough radiographs, especially in mandibular third molars, are essential for correct diagnosis and management of multiple supernumeraries.

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