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CASE REPORT

Non-syndromic bilateral dentigerous cysts with significant root resorption: a case report

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Abstract

Dentigerous cysts are the most common developmental odontogenic cysts and are associated with impacted teeth, usually third molars. Bilateral dentigerous cysts are rare and usually occur in patients with a known syndrome, such as cleidocranial dysplasia. In this study, we report a case of bilateral dentigerous cysts with associated significant root resorption in an otherwise healthy 38-year-old female patient. The patient presented with left-sided non-tender mandibular swelling and mobility of the left mandibular first and second molar teeth. Radiological investigations revealed two large, unilocular, well-circumscribed, radiolucent lesions involving the impacted lower wisdom teeth. There was extensive root resorption of the lower molars. The patient underwent enucleation of both cysts and extraction of the involved teeth. Histological features were consistent with dentigerous cysts, with both lesions consisting of a fibrous wall lined by non-keratinizing stratified squamous epithelium. Non-syndromic bilateral dentigerous cysts are extremely uncommon. This case, with associated significant root resorption, reflects aggressive biological behaviour and shows that commonly encountered lesions can present as a diagnostic challenge, emphasizing the importance of histopathological analysis in the definitive diagnosis and the overall management.

Clinical relevance

This report describes the case of a 38-year-old healthy female patient who presented with two synchronous mandibular lytic lesions. These displayed aggressive biology with significant root resorption of her lower molars. Histopathological analysis revealed that both lesions were dentigerous cysts.

To date only 22 cases of bilateral non-syndromic dentigerous cysts have been reported, but none with evidence of root resorption.

This case, although involving a very common pathological entity, displays certain features that could generate a substantial diagnostic dilemma. With the opportunity of this case, we describe the relevant diagnostic algorithm and highlight the importance of the histopathological analysis.

Introduction

Dentigerous cysts are the most common odontogenic developmental cysts associated with unerupted teeth. They account for 24% of cysts in the jaw[1]. They are generally asymptomatic and usually present as incidental radiographic findings.

Radiologically, a dentigerous cyst presents as a well-defined, unilocular, radiolucent area surrounding the crown of an unerupted tooth, most commonly a third molar, continuous with its cemento-enamel junction.

Dentigerous cysts are occasionally associated with root resorption of the responsible or neighbouring teeth. Bilateral or multiple dentigerous cysts are generally associated with syndromes such as cleidocranial dysplasia or conditions like mucopolysaccharidosis[1,2].

Here we report a case of non-syndromic bilateral dentigerous cysts associated with significant root resorption.

Case report

A fit and well 38-year-old woman presented with a left-sided non-tender mandibular swelling. The patient noticed intra-oral bone swelling and mobility of the lower left molar teeth several weeks prior to her presentation. There was no history of trauma, infection or pain.

On examination, there was no visible extra-oral swelling or asymmetry. On palpation extra-orally, the mandibular contour felt regular and the lower border of the mandible was intact. There was no paresthesia of the lower lip.

Intra-oral examination revealed a non-tender left-buccal sulcus swelling which was firm and smooth on palpation, consistent with buccal bone expansion. No soft tissue expansion or dehiscence was noted. There was no discoloration of any teeth, and the wisdom teeth were unerupted, with the LL7 demonstrating grade II and the LL6 grade I mobility. The pulp vitality test was positive.

The orthopantomogram (Fig. 1) revealed two large, unilocular, well-circumscribed radiolucent lesions involving the unerupted LL8 and the contralateral LR8. The larger lesion extended from the left body of mandible into the left ramus and up to the sigmoid notch. The lower border of the mandible appeared intact, and the inferior alveolar canal was displaced to the lower border. There was extensive root resorption of LL6, LL7 and LL8. A similar smaller lesion was seen in relation to the impacted LR8, which also demonstrated features of early root resorption.

A cone beam computed tomography scan was undertaken, which confirmed the aforementioned findings and demonstrated bilateral intra-osseous cystic lesions with aggressive biological behaviour causing significant root resorption (Fig. 2).

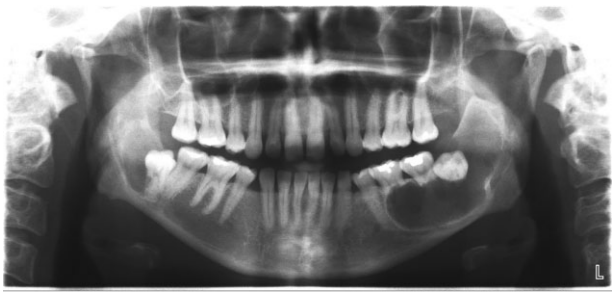

Figure 1 Orthopantomogram demonstrating synchronous bilateral mandibular radiolucent lesions associated with impacted lower wisdom teeth and significant root resorption of the neighbouring molars.

The haematological investigations, including alkaline phosphatase test, were normal, and the myeloma screen was negative. There was no history of cutaneous lesions such as multiple naevi, basal cell carcinomas, metabolic disorders or other signs that would suggest an underlying syndrome or other systemic involvement.

On the basis of the similar bilateral pathology and the aggressive biological features of the lesions, the differential diagnosis included bilateral odontogenic keratocysts, ameloblastomas, dentigerous cysts or histiocytosis X. As both lesions were over 1 cm in diameter, a biopsy was recommended.

The patient underwent enucleation of both cysts and extraction of the involved teeth under GA, followed by histological examination of the lesions.

Twenty-four months post-operatively, the patient remains free of any recurrence.

Histology

Both cysts consisted of variable thickness of fibrous wall lined by thin non-keratinizing stratified squamous epithelium. There were occasional mural nodules composed of cholesterol clefts and associated giant cells, and the fibrous wall contained diffuse lymphoplasmacytic infiltrates. There was no evidence of ameloblastoma or keratinisation (Fig. 3).

Discussion

Dentigerous cysts are the second most commonly occurring odontogenic cysts following radicular cysts. They are developmental cysts that enclose the crown of an unerupted tooth, most commonly third molars. A dentigerous cyst develops from the follicular tissues of the unerupted tooth by accumulation of fluid between the reduced enamel epithelium and the associated crown via a mechanism that still remains unclear.

Dentigerous cysts show a male predilection and occur in a wide age range from childhood to middle age. Most dentigerous cysts are solitary, slowly progressive lesions. They are generally asymptomatic and therefore present as incidental radiographic findings, unless they become infected.

Histologically, the lining of a dentigerous cyst appears as a thin, regular layer of non-keratinized stratified squamous epithelium, resembling the reduced enamel epithelium. It is supported by a fibrous connective tissue capsule; occasionally, cholesterol clefts are observed.

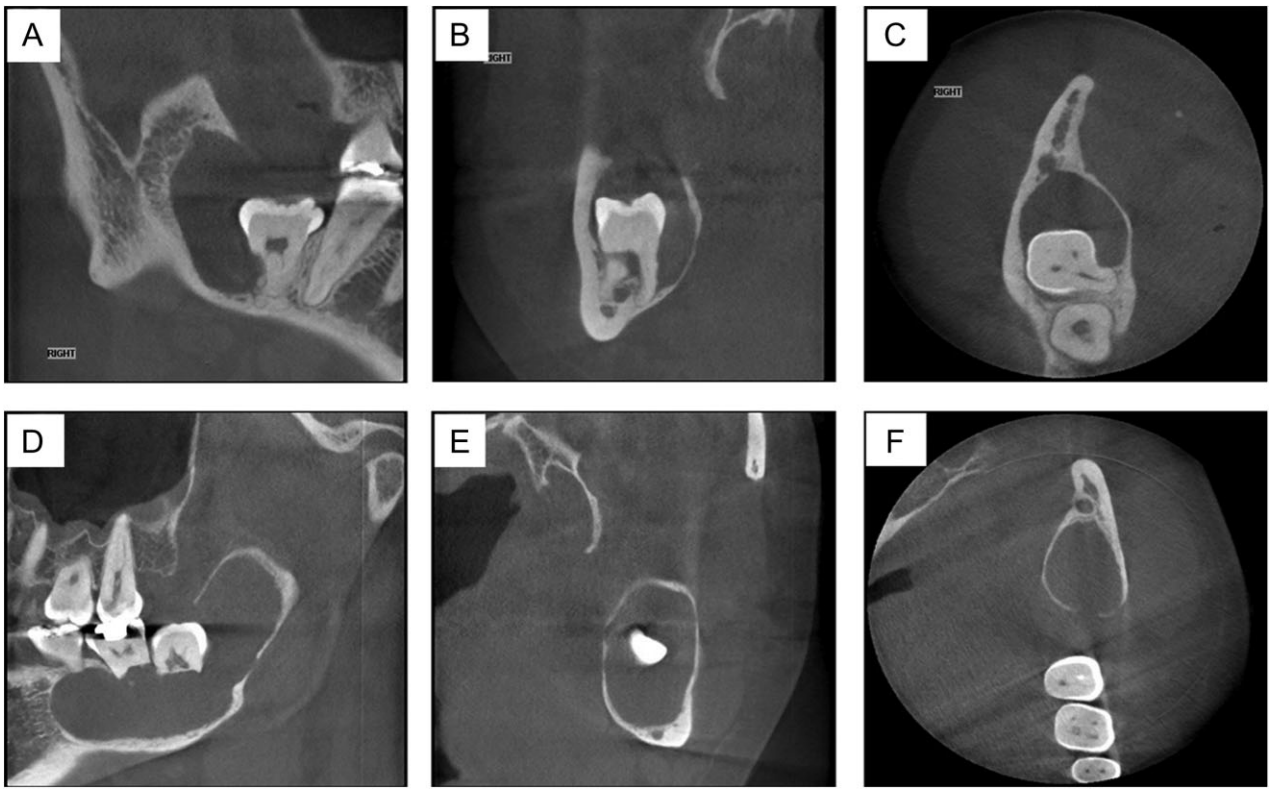

Figure 2 Representative cone beam computed tomographic slides demonstrating bilateral mandibular intra-osseous cystic lesions and associated root resorption. (A) Sagittal view of the right mandibular angle. (B) Coronal cut at the level of the lower right wisdom tooth. (C) Axial cut of the right mandibular angle region. (D) Left mandibular angle sagittal cut. (E) Coronal cut of the left mandibular angle region. (F) Axial cut of the left mandibular body and angle region, showing the cystic lesion in front of the inferior dental canal.

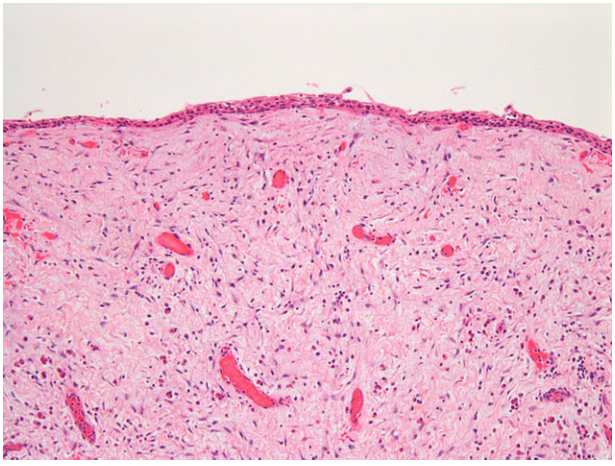

Figure 3 Representative histological picture of the left mandibular cystic lesion, demonstrating fibrous wall, lined by thin non-keratinizing stratified squamous epithelium, with no evidence of ameloblastoma (hematoxylin-and-eosin-stained, 40× original magnification).

Bilateral and multiple cysts are usually found in association with several syndromes, including cleidocranial dysplasia, Maroteaux–Lamy syndrome and mucopolysaccharidosis. Bilateral dentigerous cysts are rare in the absence of these conditions. To date, only 22 cases of non-syndromic multiple dentigerous cysts have been reported, and only one case displayed root resorption of the adjacent teeth[2].

Multiple dentigerous cysts are thought to be extremely rare in non-syndromic patients. Ochsenius *et al.* analysed 2944 cases of odontogenic cysts and identified 546 patients with dentigerous cysts, 61 (11%) of which had synchronous dentigerous cysts; none of these patients had any metabolic or syndromic conditions, but there is no clear evidence of how many of these cysts were bilateral or caused root resorption[3].

The current case presented as a diagnostic challenge for two main reasons: the unusual bilateral simultaneous presentation of clinically and radiologically similar lesions and the aggressive biological behaviour

in terms of significant root resorption. The differential diagnosis would include bilateral odontogenic keratocysts, bilateral unilocular ameloblastomas, odontogenic fibromyxomas or systemic osseolytic conditions such as histiocytosis or multiple myeloma[4].

Odontogenic keratocysts are often multilocular and most commonly located in the body or ramus of the mandible. They can present synchronously in syndromic cases, such as the basal cell naevus syndrome (Gorlin–Goltz syndrome). Although locally aggressive, keratocysts are not usually associated with root resorption[1]. Histologically, the cyst wall is thin, lined by a regular layer of stratified squamous epithelium, with a well-defined basal layer and keratin production. This keratinised epithelium displays mitotic activity and is supported by a relatively thin fibrous capsule, which may give rise to satellite cysts. In this reported case, keratinisation was excluded by microscopic analysis.

Ameloblastoma is the most common radiolucent benign odontogenic tumour that may be unilocular and can cause expansion and destruction of the bone, as well as various degrees of root resorption[1,4]. It is usually encountered in young adult life as an incidental radiological finding. Histologically, it presents as an epithelial lined cyst, comprising a basal layer of columnar cells which displays areas of intraluminal or mural proliferation of ameloblastomatous tissue[4].

Odontogenic fibromyxomas usually have multiple radiolucent areas of varying size and bony septa, often related to unerupted teeth. Unilocular lesions have also been described and can radiologically appear well-defined. Again, microscopy demonstrates specific features, readily distinguishing odontogenic fibromyxomas from other lesions, such as dentigerous cysts. An odontogenic fibromyxoma consists of stellate, fibroblast-like cells within an abundant connective tissue matrix, which contains various amounts of collagen[4].

Histiocytosis X (eosinophilic granuloma) usually occurs in children or young adults and can present as solitary osteolytic lesion in the mandible, exerting local destructive potential and causing loosening of teeth. However, multifocal eosinophilic granuloma (previously termed Hand–Schuller–Christian syndrome) is usually associated with other visceral lesions and multi-organ manifestations such as hepatosplenomegaly, lymphadenopathy and endocrine disorders[4]. The currently reported case was in the fourth decade and lacked any systemic involvement.

On the other hand, multiple myeloma causes multiple osteolytic lesions that can occur in the jaw bones, but it predominantly affects individuals between 50 and 70 years of age. Multiple myeloma is a neoplasm composed of abnormal plasma cells producing large amounts of a single homogeneous type of immunoglobulin that can be traced in blood product electrophoresis[4]. Our patient's age and overall clinical presentation made the possibility of myeloma unlikely. Blood tests and paraprotein electrophoresis proved negative for multiple myeloma.

In summary, the presented case displayed clinical and radiological features that were not typical enough to point confidently towards one single working diagnosis. Non-syndromic bilateral dentigerous cysts are extremely uncommon. This, in conjunction with concomitant significant root resorption, which is reflective of aggressive biological behaviour, shows that commonly encountered lesions can present a diagnostic challenge and emphasizes the importance of histopathological analysis in definitive diagnosis and overall management.

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