CASE REPORT

Odontogenic Keratocyst of the Posterior Maxilla: Report of an Unusual Case

NR Figueiredo, AD Dinkar, MM Khorate

ABSTRACT

The odontogenic keratocyst is a developmental odontogenic cyst. It can occur anywhere in the jaws, but it is commonly seen in the posterior part of the mandible. Lesions have a propensity to grow along the internal aspect of the jaws, and clinically observable expansion of bone occurs late. These lesions have a different mechanism of growth as compared with other cysts, and may show varying radiographic appearances. This paper reports a distinctive case of an odontogenic keratocyst in a 33-year-old female patient. The cyst has an unusual radiographic presentation of two unilocular radiolucencies overlapping each other in the left maxillary premolar-molar region. The study also presents a literature review.

Keywords: Maxilla, odontogenic keratocyst, unilocular

INTRODUCTION

Cysts of the jaws commonly occur due to the presence of numerous odontogenic, epithelial remnants that remain after tooth formation. The odontogenic keratocyst is a developmental, non-inflammatory, odontogenic cyst, and is thought to account for 10–12% of all developmental odontogenic cysts (1). Odontogenic keratocysts are unique due to their histological features, clinical characteristics and biological behaviour (2). Unlike other cysts, which are thought to grow solely by osmotic pressure, the epithelium in an odontogenic keratocyst appears to have an innate growth potential similar to a benign tumour. This difference in mechanism of growth gives these lesions a different radiographic appearance.

CASE REPORT

A 33-year-old female patient reported to our out-patient department with a chief complaint of swelling and pain over the left side of the face since three months. History revealed that the swelling was initially small in size and had gradually increased to its present size. On extra-oral examination, a diffuse swelling was seen over the left cheek region, extending from the left nasolabial fold region posteriorly to the malar region, and supero-inferiorly from 1 cm below the infra-orbital margin to 1 cm away from the left angle of mouth, measuring around 4 × 3 cm.

The overlying skin was normal with no evidence of any discharge. On palpation, the swelling was firm in consistency, tender, non-pulsatile and non-compressible, with no local rise in temperature. Intra-oral examination revealed a diffuse swelling in the left, maxillary, buccal vestibule, extending from 24 to the 27 region (Fig. 1). The swelling was firm in consistency with a smooth surface and tender on palpation. Examination of the teeth showed a carious root piece in relation to 26, while 27 was missing. No tenderness or mobility of the involved teeth was noted.

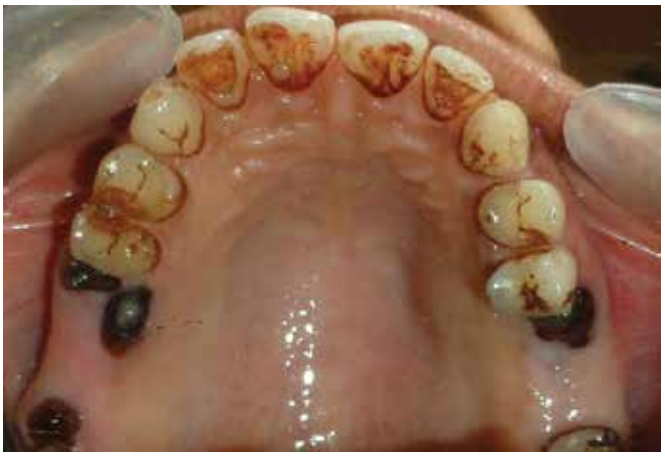

Fig. 1: Preoperative intra-oral view.

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An intra-oral, periapical radiograph showed a large, unilocular radiolucency with a well-defined, thin, corticated border extending from the apex of 24, encircling the periapical region of 25, 26 and 27 and terminating at the alveolar crest in the region of 27. Another small, oval-shaped radiolucency was seen in relation to the apex of 26, resembling a periapical granuloma, and a third unilocular radiolucency with corticated borders in the edentulous 27 area. Root resorption of 25 was noted on the distal aspect in the apical one-third (Fig. 2).

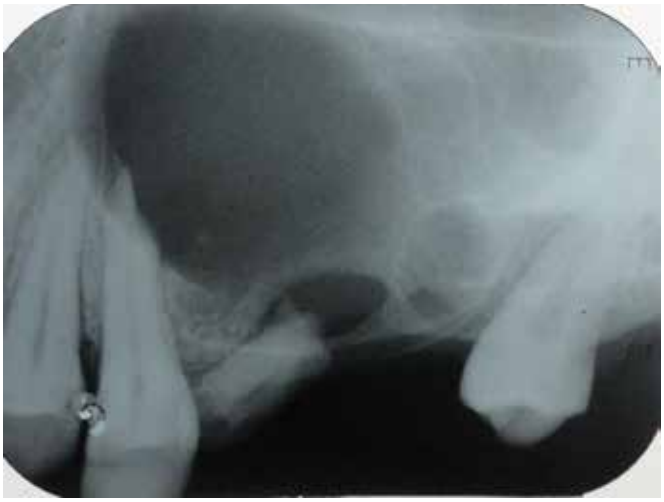


Fig. 2: Intra-oral periapical radiograph showing two separate unilocular radiolucencies overlapping in the 27 region with root resorption of 25.

A maxillary, true, occlusal view showed a large, unilocular radiolucency in the 25, 26, 27 region causing expansion of the buccal cortical plate in the affected area (Fig. 3).

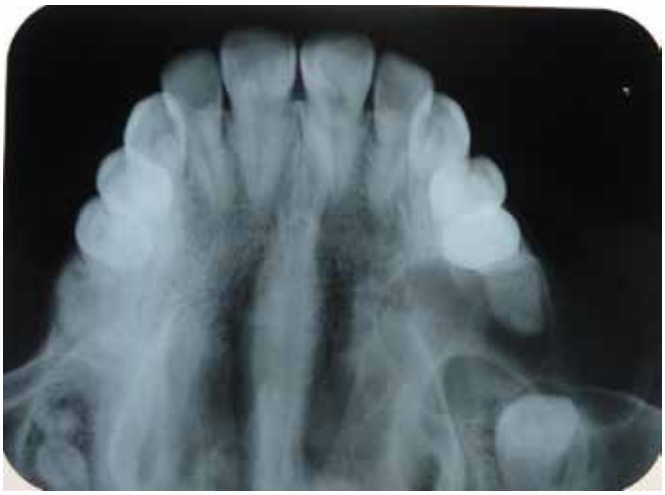


Fig. 3: Maxillary true occlusal radiograph showing expansion of buccal cortical plate in the region of 25, 26 and 27.

Panoramic radiography showed a well-defined, oval-shaped unilocular radiolucency with thin, corticated borders extending from the mesial aspect of 24 up to the 27 region, superiorly involving the maxillary sinus and inferiorly the maxillary alveolar crest. The internal structure was completely radiolucent. Another small radiolucency with a thin, corticated border was seen overlapping the above lesion in the edentulous 27 region (Fig. 4).

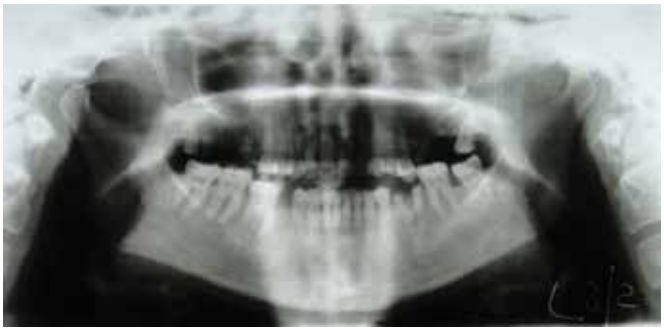
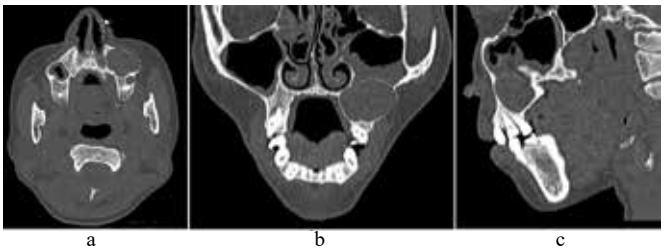


Fig. 4: Orthopantomograph showing two unilocular radiolucencies overlapping in the 27 region.

On a CT scan, an expansile, cystic lesion was seen in the left maxilla, with extension of the lesion into the lower half of the maxillary sinus and expansion and thinning of buccal cortical plate (Figs 5 and 6).



a b c

Fig 5: CT scan of maxilla: (A) axial section, (B) coronal section, (C) sagittal section.

Fig. 6: CT scan of maxilla (3D reconstruction).

Based on the history, clinical and radiographic findings, a provisional diagnosis of a residual cyst was made, with a differential diagnosis of ameloblastoma, odontogenic keratocyst, odontogenic myxoma and calcifying, epithelial, odontogenic tumour. The lesion was treated conservatively with careful enucleation and curettage. Histopathological examination showed a stratified, squamous, parakeratinized epithelium with a prominent palisaded, basal layer of cells with a picket-fence appearance. The connective tissue showed presence of collagen fibres, cholesterol clefts and giant cells, with moderate amounts of dense, chronic, inflammatory cell infiltrate (Fig. 7). Thus, based on the histopathology of the enucleated tissue, a final diagnosis of an infected odontogenic keratocyst was made.

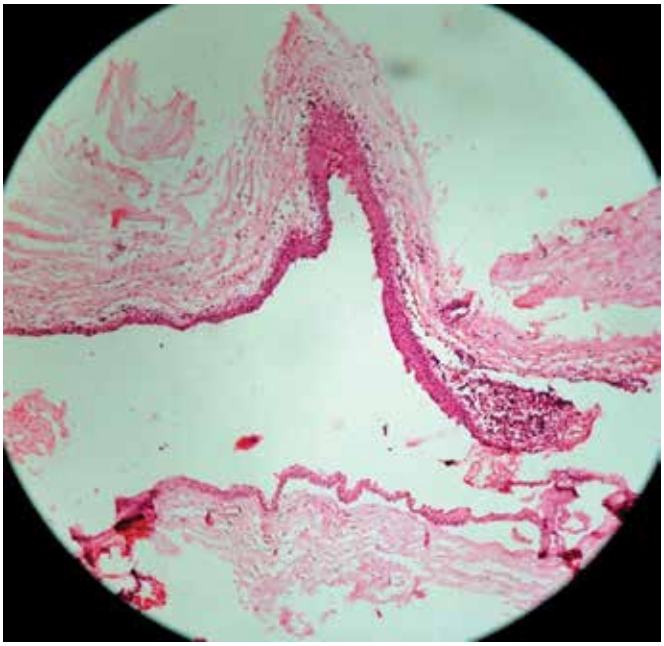


Fig. 7: Histopathology showing a stratified, squamous, parakeratinized epithelium with palisaded, basal layer of cells with a picket-fence appearance, suggestive of odontogenic keratocyst (H&E stain at ×40 magnification).

Post-operative healing was uneventful. The patient has been under regular follow-up for the past two years, during which time no recurrence has been noted.

DISCUSSION

The term odontogenic keratocyst was first used by Philipsen in 1956 (1). Owing to its aggressive nature, this lesion was renamed as keratocystic, odontogenic tumour and reclassified as an odontogenic neoplasm in the histological classification of odontogenic tumours by the World Health Organization in 2005. In this classification, the keratocystic, odontogenic tumour has been defined as 'a benign uni- or multicystic intra-osseous tumour of odontogenic origin, with a characteristic lining of parakeratinized, stratified, squamous epithelium and potential for aggressive, infiltrative behaviour' (3). However, this reclassification has not yet been universally accepted (4).

The odontogenic keratocyst arises from the odontogenic epithelium, which has two main sources—the dental lamina or its remnants, and extensions of basal cells from the overlying, oral epithelium (5). Keratocysts do not enlarge through an increase in osmotic pressure in the lumen like other cysts (6). The epithelium in odontogenic keratocysts has an increased mitotic index compared with other cysts, but similar to the ameloblastoma. Epithelial proliferation, in addition to osmolality, is a significant factor for their enlargement (7).

In 10% of patients, odontogenic keratocysts occur as one of the signs associated with Nevoid Basal Cell Carcinoma/Gorlin-Goltz syndrome (8). This syndrome comprises a number of abnormalities such as multiple, nevoid, basal cell carcinomas of the skin; skeletal, central nervous system and eye abnormalities; and multiple, odontogenic keratocysts. Hence, early diagnosis and follow-up of a patient with an odontogenic keratocyst is important because of the possibility of such patients developing other features of Gorlin-Goltz syndrome in the future.

Odontogenic keratocysts are thought to have a bimodal age distribution with a peak frequency in the second and third decades of life, and a second peak in the fifth decade, and are thought to have a male predilection (5). Around 70–80% keratocysts are found in the mandible, most commonly in the posterior body and ramus (8, 9). However, these cysts can occur anywhere within the jaws and can resemble other lesions like lateral periodontal, periapical and nasopalatine duct cysts (1). Our case was diagnosed in a 33-year-old female, which is in accordance with the previous literature, but occurred in the maxillary posterior region, which is an uncommon site for these lesions.

The clinical features and radiographic appearance of odontogenic keratocysts are not characteristic. This may lead to misdiagnosis, especially when the lesion is in relation to a non-vital tooth (10). Odontogenic keratocysts tend to grow in an antero-posterior direction within the medullary cavity of the bone, without causing obvious bone expansion. Lesions usually have no symptoms, although patients may complain of pain or swelling. The radiographic appearance of odontogenic keratocysts may range from a small, unilocular radiolucency to a

large, multilocular radiolucency (6). Some unilocular lesions may have scalloped margins, which suggest that unequal growth activity may be taking place in different parts of the cyst lining (5). According to Shear, almost all maxillary lesions tend to be unilocular (5). The present case occurred in the periapical region of the left posterior maxilla and radiographically resembled a residual cyst. Although odontogenic keratocysts are known to present as unilocular radiolucencies, it is very unusual for lesions to show two separate overlapping radiolucencies as was seen in the present case.

The main difference between odontogenic keratocysts and other jaw cysts is their potentially aggressive behaviour. Odontogenic keratocysts recur more often than any other type of jaw cyst. The reasons for this include: (a) growth of a new lesion from small, satellite cysts or odontogenic epithelial rests left behind by the surgical treatment; (b) incomplete removal of the original cyst lining due to its very thin and fragile nature; (c) an attempt to save vital adjacent teeth or nerves, which may lead to incomplete eradication and, hence, recurrence (5).

The histopathologic features of odontogenic keratocysts are pathognomonic. Lesions are characterized by a keratinized, stratified, squamous epithelium, with a prominent, often palisaded, basal layer, composed of either columnar or cuboidal cells, and a connective tissue wall (7). The form of keratinization is parakeratotic in 80–90% cases but may sometimes be orthokeratotic. The parakeratotic layers often have a corrugated surface. The connective tissue may show an inflammatory infiltration in most cases. The present case showed all the typical histologic features of an odontogenic keratocyst.

Other than clinical behaviour that differentiates the odontogenic keratocyst from other cysts, there are additional differences such as protein, immunoglobulin and glycosoaminoglycan content of their fluids and the prostaglandin content of their wall (7).

The treatment of odontogenic keratocysts remains controversial, with some researchers advocating conservative procedures, whereas others suggest aggressive management of lesions. Conservative treatment generally includes simple enucleation, with or without curettage, or marsupialization. The advantage is preservation of anatomical structures and reduced morbidity to the patient. For keratocysts which may resemble other odontogenic cysts radiographically, and which have been treated with enucleation and curettage, immediate further treatment does not appear to be necessary (1). Aggressive treatment generally includes peripheral ostectomy, chemical curettage with Carnoy's solution and resection. It is mostly recommended for large lesions, recurrent cases and syndromic patients (3). However, despite a number of studies on the various treatment modalities, none have shown complete prevention of recurrence of lesions and thus there remains no consensus on the adequate or appropriate treatment of this lesion (11). The present case resembled a residual cyst and was hence treated by enucleation and curettage and the patient has been asked to follow up at regular intervals.

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