

Nonsyndromic Bilateral Keratocystic Odontogenic Tumor: A Rare Case

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ABSTRACT

Here, we report a case of keratocystic odontogenic tumor occurring bilaterally in a male patient in fourth decade of life. This case is different from bilateral lesions of keratocystic odontogenic tumor which are most commonly parakeratinized and are associated with syndromes like nevoid basal cell carcinoma syndrome. In our case, we could not find out any syndromic association and furthermore histopathology was in line of orthokeratinization.

Keywords: Keratocystic odontogenic tumor, Nonsyndromic, Orthokeratinized odontogenic keratocyst, Parakeratinized odontogenic keratocyst, Nevoid basal cell carcinoma syndrome, Multiple odontogenic keratocysts.

INTRODUCTION

Odontogenic keratocysts are clinically aggressive lesions which are thought to arise from dental lamina and its remnants.¹ Odontogenic keratocysts constitute up to 21% of all odontogenic cysts.^{2,3} Odontogenic keratocysts occur over a wide range from first decade to ninth decade, many studies have demonstrated bimodal age distribution with peak frequency in second decade and the second peak in the fifth decade.¹ Odontogenic keratocysts are generally found more frequently in male patients than females.^{1,4-8} Majority of lesions occur in the mandible, mainly in the posterior body and ascending ramus^{3,5,9} the angle⁹ and symphyseal area is also a frequent locus for this lesion.¹⁰ Odontogenic keratocysts can be aggressive lesion with high recurrence rate and tendency to invade adjacent tissues.⁵ Treatment options vary from marsupialization and enucleation, combined with adjuvant cryotherapy or chemical cautery or Carnoy's solution to marginal or radical resection.^{6,11-15} The recurrence rate has been reported to vary from 2.5 to 62.5%.^{3,6,9,12,16-18} Malignant transformation of odontogenic keratocysts has also been reported.¹⁹ The high recurrence rate and aggressive behavior of the odontogenic keratocyst have caused WHO to regard it as a benign neoplasm rather than a cyst and WHO has renamed it as keratocystic odontogenic tumor.^{3,20,21}

CASE REPORT

A male patient aged 32 years reported to our hospital with the chief complaint of pain and swelling on the left side of the face for the past 3 months. Further history revealed trauma in

the left posterior mandibular area 3 months back. The swelling was small initially which gradually progressed to the present dimension. The swelling extended from the anterior corner of the mouth to the posterior border of mandible and superiorly from zygoma to inferiorly 2 cm below the lower border of mandible. The swelling measured about 5 × 7 cm with a defined shape and hard in consistency (Fig. 1).

Intraoral examination of lower left region revealed obliteration of the buccal vestibule which extended from the molar region up to the ramus (Fig. 2). Medical history was not significant.



Fig. 1: Swelling on the left side of the face



Fig. 2: Intraoral examination showing obliteration of buccal vestibule

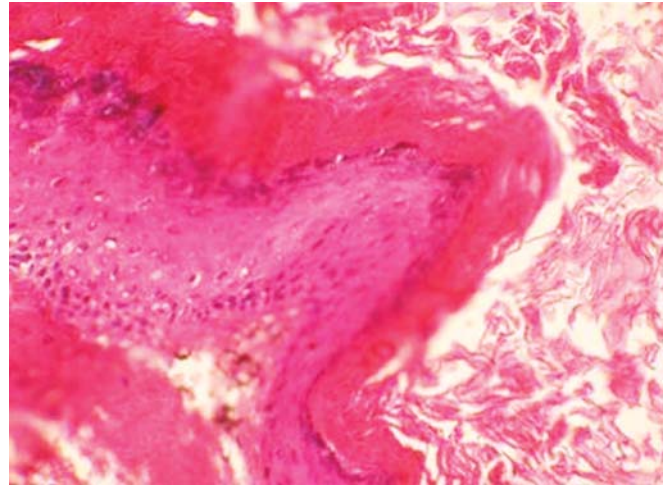


Fig. 5: Lesion of the left side under 40x



Fig. 3: OPG view demonstrating bilateral radiolucency

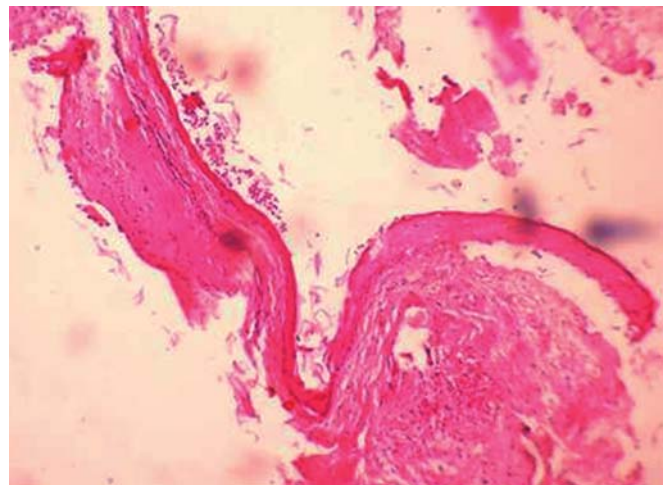


Fig. 6: Lesion of the right side under 10x

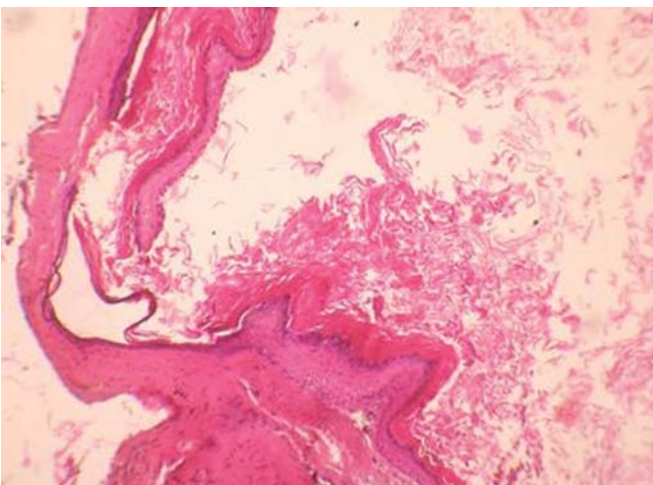


Fig. 4: Lesion of the left side under 10x

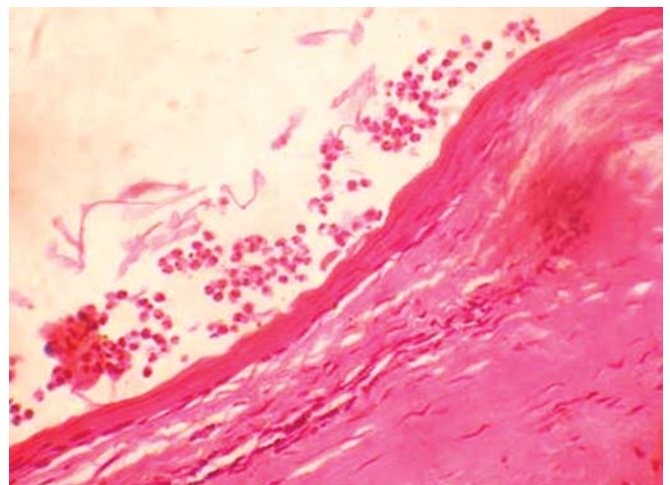


Fig. 7: Lesion of the right side under 40x

The panoramic radiograph revealed a multilocular radiolucency in the left mandibular posterior region. The radiolucency extended from mesial root of the mandibular left first molar till the angle of mandible extending up to the coronoid process involving the ramus and as an incidental finding the

radiograph also demonstrated a well-defined unilocular radiolucency on the right side of the mandible distal to the third molar (Fig. 3). Correlating history, clinical and imaging findings a provisional diagnosis of bilateral keratocystic odontogenic tumors was considered.

Incisional biopsies were done from left lateral border of mandible and right retromolar area. During biopsy white cheesy material was found extruding from cystic lesion.

Microscopy of the lesions revealed orthokeratinized stratified squamous cystic lining epithelium overlying the connective tissue. The epithelium was 4 to 6 layers thick with no rete pegs. The cells of the basal cell layer were relatively flat and hyperchromatic. Cystic lumen showed keratin deposits. The epithelium showed separation from the underlying connective tissue in certain areas (Figs 4 to 7). Diagnosis of keratocystic odontogenic tumor was made.

DISCUSSION

Usually, odontogenic keratocysts occur as solitary lesions.¹ Multiple odontogenic keratocysts commonly occur in nevoid basal cell carcinoma syndrome, Gorlin-Goltz syndrome,²² orofacial digital syndrome, Ehler-Danlos syndrome, Noonan syndrome and Golabi-Behmel syndrome.²³

Odontogenic keratocysts have a biologic behavior similar to a benign neoplasm^{3,20} hence WHO has classified odontogenic keratocysts (OKCs) under odontogenic tumors.²¹

NBCCS is characterized by multiple odontogenic keratocysts, nevoid basal cell carcinomas of the skin, bifid ribs, calcification of the falx cerebri and other features.²⁴ Odontogenic keratocysts, when associated with NBCCS, present together with skeletal, cutaneous, neurologic, ophthalmic and sexual abnormalities.²⁵ However, these features were not present in our case. Patients with odontogenic keratocyst complain of swelling and pain or both.^{1,26,27} In many instances, patients are remarkably free of symptoms until the cysts have reached a large size, this occurs because the odontogenic keratocyst tends to extend in the medullary cavity and clinically observable expansion of the bone occurs late.¹ However, in our case cortical plate expansion on the left side was observed.

Radiographically, OKCs present as a well-defined radiolucent lesions with smooth, usually corticated margins and maybe either multilocular or unilocular. There is involvement of an unerupted tooth in 25 to 40% of cases.²⁸ However, in our case, the left side lesion was associated with a multilocular radiolucency and the right side lesion was associated with a unilocular radiolucency distal to third molar having well-corticated margins. Both the lesions were not associated with impacted teeth.

Histologically, parakeratinized type of epithelium is seen in about 83% of the odontogenic keratocysts and orthokeratinized type is seen only in about 10% of cases.²⁹ The odontogenic keratocyst epithelium is rather thin around 6 to 10 layers unless there has been superimposed inflammation. The lining epithelium shows a parakeratinized surface which typically corrugated, rippled or wrinkled^{26,29} rarely, small foci of orthokeratinization are also found.³⁰ A prominent pallisaded, polarized well-defined basal cell layer composed of columnar or cuboidal cells is seen, often the cells of the basal cell layer have been described as having a 'picket fence' or 'tombstone' appearance.^{1,27,29,31,32} Keratin may also be appreciated in the

lumen.³² Separation of the epithelium from the supporting connective tissue of the cyst is common and is caused by metalloproteinase-mediated degradation of collagen in juxtaepithelial regions.^{33,34}

In case of orthokeratinization, the basal cells are flatter and are composed of cuboidal or flattened squamous cells.³⁵ Histopathological studies have suggested parakeratinization, intramural epithelial remnants and satellite cysts are more frequent among odontogenic keratocysts associated with NBCCS.^{36,37} However, our patient was nonsyndromic and the microscopic findings were consistent with the literature regarding orthokeratinization in nonsyndromic cases, which makes our case unique. Microscopic examination of the lesions revealed orthokeratinized stratified squamous cystic epithelium overlying the connective tissue. The epithelium was 4 to 6 layers thick with no rete pegs. The cells of the basal cell layer were relatively flat and hyperchromatic. Cystic lumen showed keratin deposits. The epithelium showed separation from the underlying connective tissue in certain areas.

Orthokeratinized odontogenic keratocyst represents a distinct biologic entity from the parakeratinized ones with a more limited growth potential and lower recurrence rate, recent immunohistochemical studies that compared orthokeratinized odontogenic keratocyst with parakeratinized odontogenic keratocyst have shown tenascin, a protein that is often overexpressed in epithelial malignancies was expressed in parakeratinized odontogenic keratocyst.³⁸ Therefore, it is said that orthokeratinized odontogenic keratocyst is a distinct entity apart from the parakeratinized odontogenic keratocyst and is expected to behave in a more indolent manner with fewer tendencies for recurrence.²⁷

Treatment modalities include marsupialization and enucleation, combined with adjuvant cryotherapy or chemical cauterly or Carnoy's solution and marginal or radical resection.¹⁴

CONCLUSION

In any patient with multiple odontogenic keratocysts, the possibility of NBCCS must be considered. A complete clinical examination and histopathologic analysis must be performed to detect any features associated with this syndrome.

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