Case Report

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Non-syndromic Multiple Odontogenic Keratocysts: A Rare Case Report and Current Review of Literature

Harsha Gorrela[1*] Sunnam Madhavi[2] Aditya Mohan Alwala[3] Ramesh K[4] Rathod Prakash[5] Anusha Maganti[6]

[1]Professor and Head, Department of Oral and Maxillofacial Surgery, MNR Dental College and Hospital, Hyderabad, Telangana, India.
[2]PG Student, Department of Oral and Maxillofacial Surgery, MNR Dental College and Hospital, Hyderabad, Telangana, India.
[3]Associate Professor, Department of Oral and Maxillofacial Surgery, MNR Dental College and Hospital, Hyderabad, Telangana, India.
[4]Associate Professor, Department of Oral and Maxillofacial Surgery, MNR Dental College and Hospital, Hyderabad, Telangana, India.
[5]Assistant Professor, Department of Oral and Maxillofacial Surgery, MNR Dental College and Hospital, Hyderabad, Telangana, India.

ABSTRACT

Background: Odontogenic Keratocyst (OKC) is a developmental odontogenic cyst affecting the maxillofacial region. Multiple OKCs are usually seen in association with nevoid basal cell carcinoma syndrome (NBCCS), but approximately 5% of patients with OKC have multiple cysts without concomitant syndromic presentation. We report a rare case of multiple OKCs in a non-syndromic female patient, with emphasis on its diagnosis, radiographic features, treatment and current review of literature.

Keywords: Multiple odontogenic keratocysts; Gorlin goltz syndrome; Non-syndromic.

INTRODUCTION

The maxillofacial region is more prone to cystic lesions than any other part of the body. Keratocystic odontogenic tumors are the most common form of cystic lesions effecting the maxillofacial region [1,2]. They are clinically aggressive lesions which are thought to arise from the dental lamina or its remnants [1,3,4] and are usually seen during the second to fourth decades of life with a slight male predilection. Multiple OKCs are usually seen with cutaneous, skeletal, ocular and neurologic abnormalities as a component of nevoid basal cell carcinoma syndrome (NBCCS). The features of this syndrome were first described by Gorlin and Goltz in 1960, so it is also recognized as Gorlin-Goltz syndrome [5,6]. This article reports a case of multiple OKCs without any syndromic manifestations.

CASE REPORT

A 24 year old female patient was referred to the department of oral and maxillofacial surgery, MNR dental college and hospital with a chief complaint of pain and swelling in the lower right anterior region of the mandible since 1 month.

Extra oral examination revealed no gross facial asymmetry, overlying skin was normal (Fig -1). On Intraoral examination a diffused swelling was noted in the right mandibular buccal vestibule with obliteration seen extending from lower 1st premolar to 2nd molar and pus discharge was seen irt 41,43, and missing tooth in relation to 42 (Fig 2).

OPG, revealed multiple radiolucent lesions in the maxilla in relation to 18, 28 impacted teeth and in mandible in relation to 42, 48 impacted teeth (Fig 3). CBCT revealed multilocular radiolucent lesion surrounded by radiopaque border in right ramus, angle, and body of mandible extending up to coronoid process of size 5.3 cm x 3.0 cm in relation to impacted 48 tooth. CBCT also revealed one more multilocular radiolucent lesion surrounded by radiopaque border in right body and symphysis

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*Correspondence Dr. Harsha Gorrela.
Department of Oral and Maxillofacial Surgery, MNR Dental College and Hospital, Hyderabad, Telangana, India.
Email: Not Disclosed

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region of mandible of size 5.6 cm x 2.3 cm crossing midline in relation to impacted 42 tooth. In Maxilla, CBCT scan revealed radiolucent lesions surrounded by radiopaque border measuring approximately 2.2 cmx2.3 cm in relation to impacted 18and 28 tooth were noted (Fig 4,5,6).

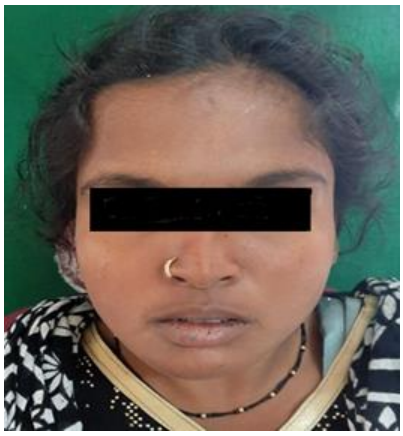

Fig 1: Extraoral

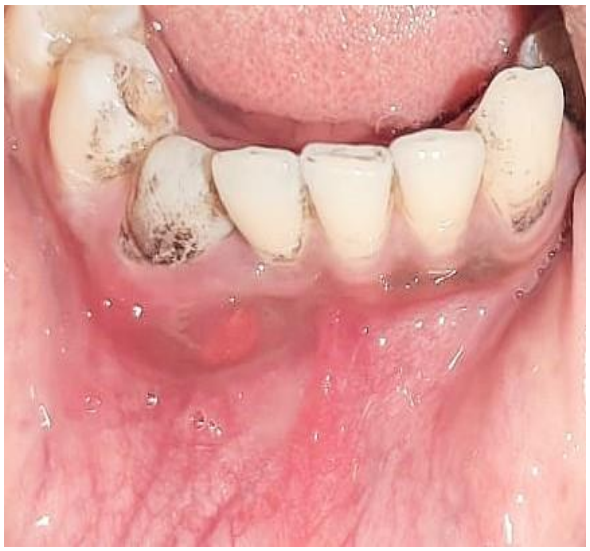

Fig 2: Intraoral photograph.

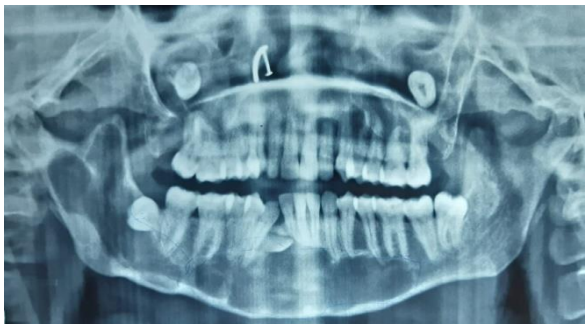

Fig 3: OPG – multiple radiolucent lesions in relation to 48, 42, 18, 28 teeth

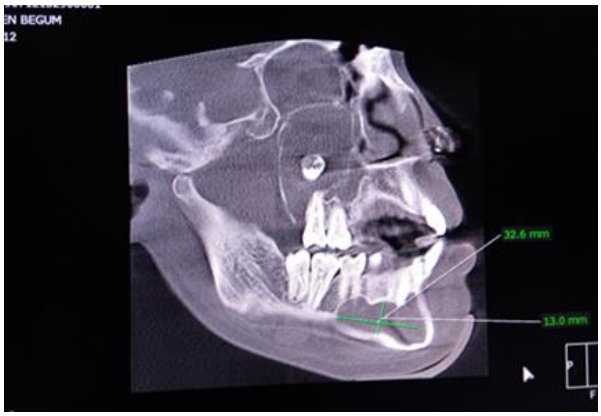

Fig 4 : CBCT- Expansile radiolucent lesion in maxilla in r.to 18 tooth.

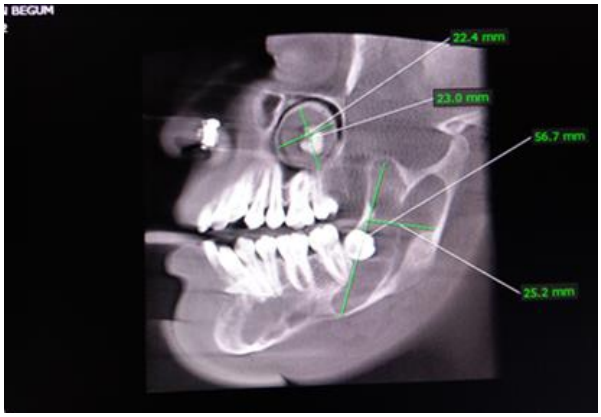

Fig 5: CBCT scan revealing cystic lesion crossing midline and extending up until.

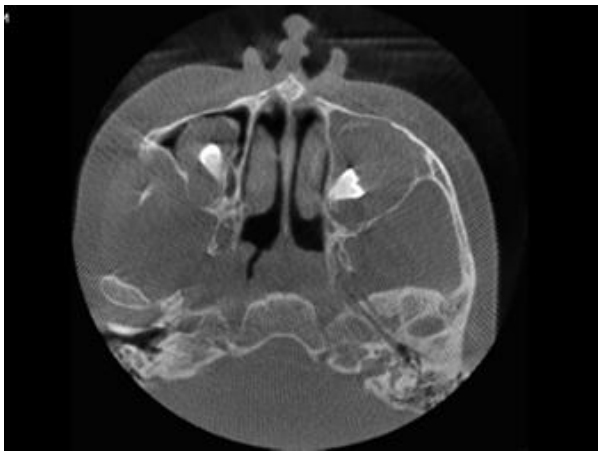

Fig 6: CBCT – radiolucent lesions in relation 18,28 teeth

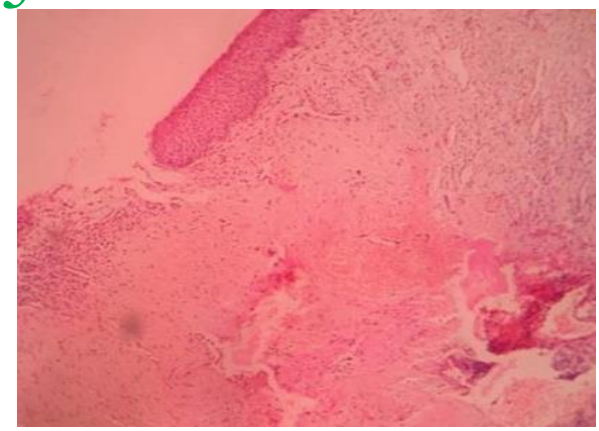

Fig 7: HPE - palisaded pattern suggestive of odontogenic keratocyst.

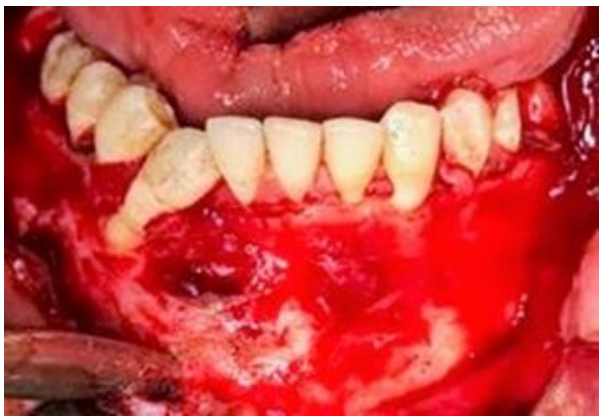

Fig 8: Intra op picture of patient Mandible.

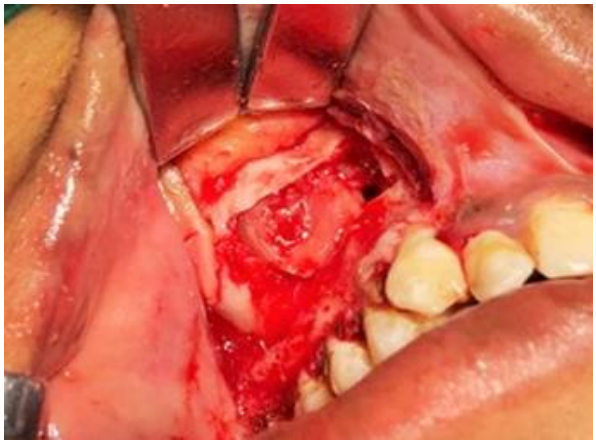

Fig 9: Intra op picture of patient- Maxilla.

Skull and chest radiographs did not reveal any radiographic abnormalities. FNAC and incisional biopsy were done. FNAC revealed cheesy cream colored fluid aspirate. The histopathology examination revealed cystic epithelium and supporting connective tissue wall. The cystic epithelium is comprised of 6-8 layers of parakeratinized epithelium. There is also evidence of basal cell palisading in few areas. The connective tissue wall comprised of moderate chronic inflammatory cell infiltrate features which were suggestive of odontogenic keratocyst (Fig- 7).

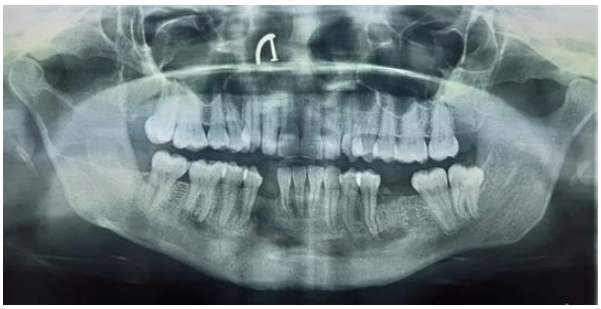

Fig 10: Follow up OPG of the patient.

Enucleation of multiple odontogenic keratocysts and chemical cauterization with carnoy's solution was performed under general anesthesia (Fig- 8,9). Entire lesion with the cystic lining was sent for histopathological examination, which confirmed the final diagnosis of odontogenic keratocyst. Patient was followed up for 2 years with no evidence of recurrence (Fig- 10).

DISCUSSION

Odontogenic keratocyst was first identified by Philipsen in 1956 and was defined by the World Health Organization (WHO) as a cyst characterized by a capsule and a lining of keratinized stratified squamous epithelium of usually about five to eight cells in thickness and generally without rete pegs.[1,9] The OKC, now designated by WHO as a KCOT, is defined as "a benign, uni or multicystic, intraosseous tumor of odontogenic origin, with a characteristic lining of parakeratinized stratified squamous epithelium and potential for aggressive, infiltrative behavior." WHO recommends the term "keratocyst odontogenic tumor "as it reflects its neoplastic nature [1,10].

Multiple OKCs usually occur as a component of syndromes such as Nevoid basal cell carcinoma (NBCCS) OR Gorlin-Goltz syndrome, orofacial digital syndrome, Noonan syndrome, Ehler-danlos syndrome and simpson-golabi-behmel syndrome [7,8,11,12].The biologic behavior of OKCs associated with NBCCS is more aggressive and these cysts have higher recurrence rate (82%) compared with solitary keratocyst (61%).[8,13].

The occurrence of single or multiple OKCs in the absence of other features of the syndrome represents the least complete form of NBCCS [8,14,15]. The absence of family history and other features of the syndrome can be due to variation in penetration and expression of different mutations with in the same gene or the effect of modifier genes and environmental factors [8,16].

NBCCS were recognized by multiple KCOTs, nevoid basal carcinomas of the skin, bifid ribs, calcification of the falx cerebri and other features [3,5]. However our patient does not have all these features except for multiple OKCs and was healthy.

Therapeutic interventions of OKC include marsupialization and enucleation, combined with adjuvant chemical cauterization with carnoy's solution, and marginal or radical resection.

In the treatment of recurrent OKCs associated with NBCCS, overlying surface epithelium should be excised along with the cystic lining to prevent recurrences from residual epithelial islands and microcysts [8,19,20]. In addition, use of carnoy's solution following cyst enucleation (applied only over areas where the cyst is attached to the mucosa) and cryosurgery (because of the unique ability of liquid nitrogen to devitalize bone in situ while leaving the inorganic framework untouched) is advocated to kill epithelial remnants and dental lamina within the osseous margin and, thus, prevent recurrences.[8,21,22].

Similar cases to the current case have been reported in a few published articles.

Alok A et al. presented a 24 year old male patient with a multiple non-syndromic KCOT with well-defined radiolucency with corticated margins were present with no other systemic manifestations [1].

Sholapurkur et al. presented a 24 year old case with a multiple non-syndromic KCOT in both jaws with a chief complaint of a slow growing swelling since 3 years and drainage since 15 days. The swelling was associated with pain with gradual onset radiating head on same side. Lesions were cyst like radiolucency associated with impacted teeth on panaromic radiograph[4].

Kargahi N et al. presented a 11year old boy with multiple KCOTs without any notable features of syndromes. But there is no details any follow up of the case were reported [5].

Bartake et al. reported a 20 year old case, shows multiple OKC with frequent recurrences without any other notable features. which are indicative of Gorlin Goltz Syndrome[7].

Guruprasad and Chauhan discussed a 16-year-old patient with multiple KCOTs and a complaint of slow progressing swelling in both jaws without any other features of syndrome [9]. Shabnam et al. reported a case 37 year old male with multiple cystic lesions of KCOTs in the upper and lower jaws . CT scan and panoramic radiographs showed multiple radiolucencies with well-demarcated borders, cortical margins and different sizes.. The clinical examination of the patient revealed no systemic diseases or symptoms in the skin and the involved areas. The chest and skull radiographs also showed no symptoms [17]. Nirwan A et al. reported a case in which three of the four siblings suffered from multiple non-syndromic OKC. This clearly indicates the role of familial tendency in the formation of multiple cysts [18].

CONCLUSION

Multiple KCOT's could occur in patient without an associated syndrome due to its multifocal nature. If any patient reporting with the multiple OKCs should be evaluated thoroughly for the possibility of NBCCS as OKCs may be the first and only manifestation of this syndrome. Also for the fact that OKCs associated with this syndrome have higher rate of recurrence than the isolated OKCs, a very strict follow up has to be followed for a long period of time. The possibility of other features of NBCCS has to be explained to the patient as well as his relatives, so as to allow appropriate genetic counseling and serial screening for the development of malignancies and other complications besides OKCs.

CONFLICTS OF INTEREST

The authors declare they have no potential conflict of interests regarding this article.

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