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Management of Non Syndromic Multiple Odontogenic Keratocyst- A case report with literature review

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Abstract

Odontogenic keratocyst is a common developmental odontogenic cyst affecting the maxillofacial region. Multiple Odontogenic keratocysts are usually seen in association with nevoid basal cell carcinoma syndrome (NBCCS) but approximately 5% of patients with odontogenic keratocyst have multiple cysts without concomitant Syndromic presentation. These multiple odontogenic keratocysts warrant aggressive treatment at the earliest because of the damage and possible complications associated with them. Recurrence in these lesions is the most characteristic feature that has to be taken in consideration while explaining the prognosis to the patient.

After histopathological examination Surgery was performed under general anesthesia. After reflection of full thickness mucoperiosteal flap, extraction of associated teeth in all the four quadrants was done. Enucleation with peripheral osteotomy was performed followed by application of freshly prepared Carnoy's solution for 4 minutes in all the four quadrants. Closure was done with absorbable sutures.

An aggressive management is usually required to address the cases of odontogenic keratocyst which does not end after the surgery. The management continues in the form of keen follow up due to high recurrence of odontogenic keratocyst. The follow up of cases with multiple odontogenic keratocyst is even more demanding as the

recurrence as well as development of any feature of NBCCS has to be checked for.

Keywords: Carnoy's Solution,Nevoid basal cell carcinoma syndrome, Odontogenic Kerato cyts,peripheral osteotomy

Introduction

Odontogenic keratocyst is an entity that has jumped under various headings of classification of Odontogenic and maxillofacial bone lesions proposed by WHO. Its journey has been through both cysts and tumor. The latest classification of WHO (2017) restores odontogenic keratocyst to odontogenic cysts and discards it from the category of tumors.[1]

Occurrence of multiple odontogenic keratocysts (odontogenic keratocysts) though quite rare but whenever encountered demands a higher vigilance in terms of presence of-Associated syndromes (Gorlin goltz-Goltz,Noonans,Orofacial digital syndrome, Ehlors danlos syndrome & Simpson golabi behmal syndrome) and a higher rate of recurrence of 30% compared to that of solitary lesions being only 10%.[2,3]

This article intent to present a case of non Syndromic multiple odontogenic keratocyst and its management with review of literature.

Case Report

A 32 year female with the complaint of pain in lower right back region of jaw reported to the department of oral and maxillofacial surgery.

History of presenting illness revealed that the pain was dull aching type that gets relieved on taking medications. There was a history of discharge 1 month back from the same region which got resolved on its own. There was no history of trauma.

There was no significant past medical or family history.

Intra oral examination revealed-

- Grossly Carious 15, 16
- Mesially tilted 37, 47, 48
- Mild tenderness on palpation over mandibular right buccal vestibule with associated discharge in relation to 37

On Radiographic examination Panaromic radiograh (Figure 1) revealed,

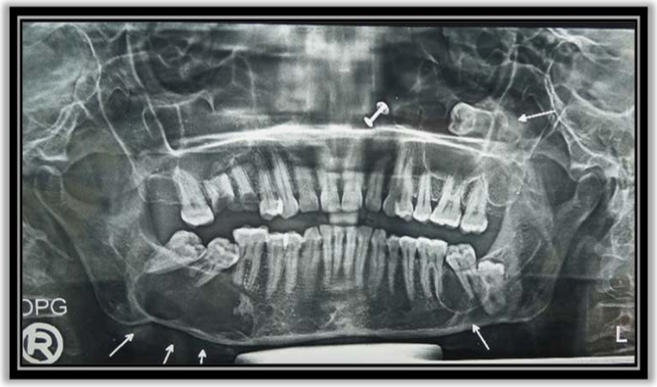

Figure 1: Panormic Radiograph

- Maxillary right quadrant, there was a unilocular radiolucency presenting a well-defined peripheral rim involving the 1st and 2nd molars. The 1st molar and 2nd premolar were found to be carious.
- Maxillary left quadrant, there was a unilocular radiolucency with a sclerotic border involving the second premolar, 1st and 2nd molars. The third molar was seen to be displaced into the maxillary antrum.
- Mandibular left quadrant, unilocular radioluceny with sclerotic border was seen inolving the distal aspect of 1st molar to distal aspect of second molar. 3rd molar was impacted.
- Mandibular right quadrant, unilocular radiolucency with well defined peripheral rim from mesial of 1st molar to distal of 3rd molar extending upto lower mandibular border. 2nd and 3rd molars were mesialy tilted.

Patient was further advised for computed tomography (CT Scan) Figure 2 which revealed,

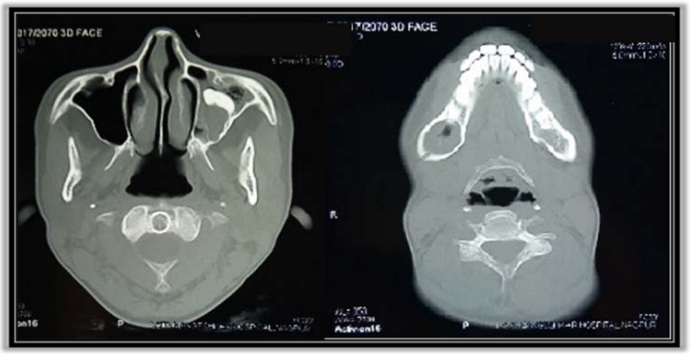

Figure 2: Axial view of CT scan

- Two soft tissue attenuation lesions noted bilaterally in the body of mandible measuring 2.2cmX1.5cm on right side and 1.3X1.4cm on left side. Expansion of mandibular cortex with thinning was distinguished with the breech in the lingual cortex of mandible on its superior margin on right side.
- Tooth with associated cyst was noted in the left maxillary antrum.

A clinico-radiographical diagnosis was made to be multiple odontogenic keratocyst where as dentigerous cyst and unicyctic ameloblastoma were considered for differential diagnosis. An incisional biopsy from right mandibular posterior region was sent for histopathological correlation which was found to be odontogenic keratocyst.

In accordance with clinical, radiological and histopathological examination, final diagnosis of odontogenic keratocyst in mandibular right posterior region was made and the findings of other three quadrants were provisionally assumed to be **odontogenic keratocyst/cycstic lesion** and further investigations were done to rule out any associated syndrome.

- Dermatological examination revealed no abnormal finding like palmer or Planter keratosis and nevi (Figure 3)
- Chest and skull radiographs were normal without evidence of bifid ribs and bridging of sella tursica (Figure 4)
- USG lower abdomen showed no abnormality like Ovarian fibroma.

Figure 3: No signs of Palmer or planter Keratosis

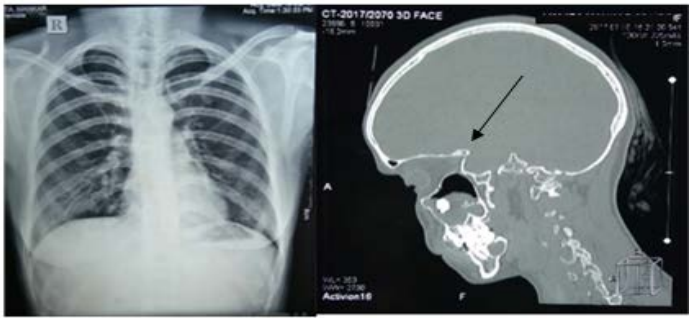

Figure 4 : Chest radiograph and sagital skull view show no sign of abnormality. Black arrow shows Normal Sella Turcica

Patient was posted for surgery under general anesthesia after pre anesthetic fitness.

After giving incision and elevation of full thickness mucoperiosteal flap, extraction of associated teeth in all the four quadrants were done (Figure.5). Enucleation of all the cystic lining was performed with peripheral osteotomy followed by application of freshly prepared Carnoy's solution for 4 minutes in all the quadrants (Figure.6,7). In mandibular right posterior lesion, lingual cortex perforation was seen and after peripheral osteotomy lower border of mandible was still intact. Closure was done using vicryl 3-0 absorbable suture after thorough irrigation of all the four quadrants with 10% Povidone Iodine

solution. The cystic lining from all the four quadrants were sent for histopathological examination.

Intravenous antibiotics (Inj. Ceftriaxone 1gm twice daily and Inj. Metronidazole 500mg thrice daily) and analgesics were given for 5 days.

Before the discharge of patient maxillomandibular fixation was done to avoid any chance of pathological fracture in 4th quadrant.

Patient was kept on follow up to rule out recurrence. A follow up after six months (Figure 8) and two and half years (Figure 9) showed no signs of recurrence and development of Nevoid basal cell carcinoma Syndrome.

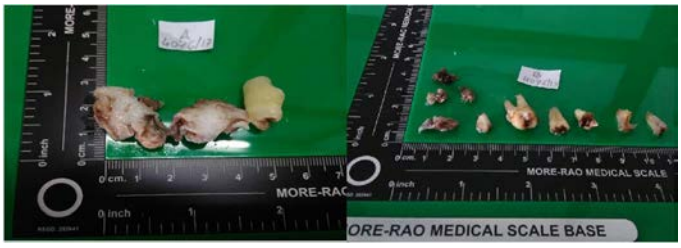

Figure 5: Extracted teeth specimen

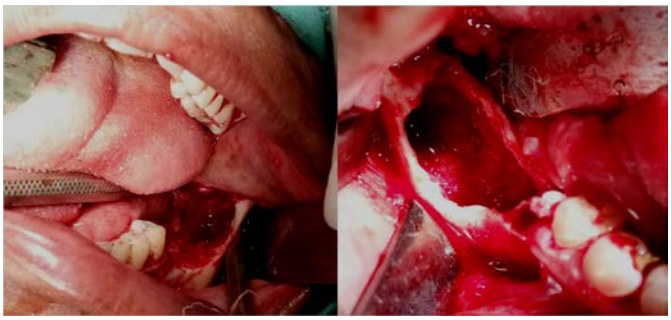

Figure 6: Cystic cavities after enucleation and peripheral osteotomy

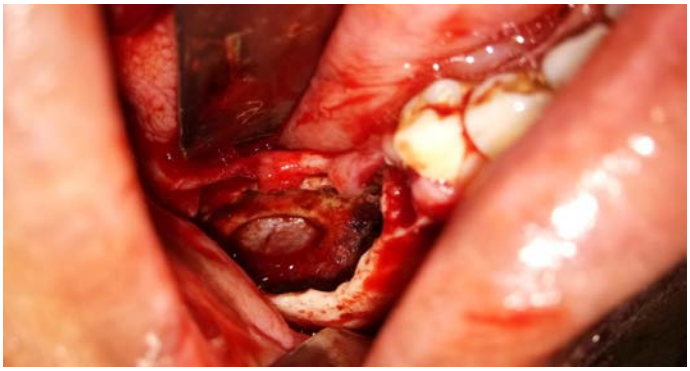

Figure7: Cystic Cavity after application Of Carnoy's Solution

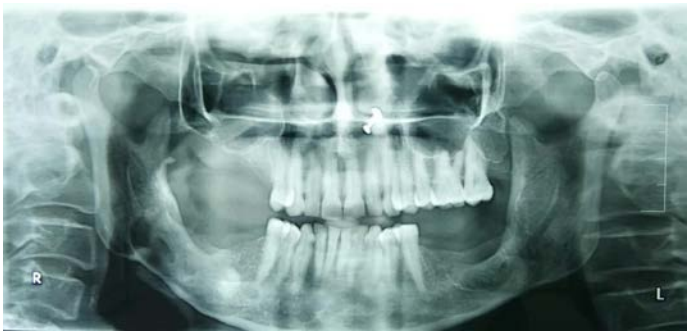

Figure 8: Six month follow up radiograph shows no sign of recurrence and with bone formation

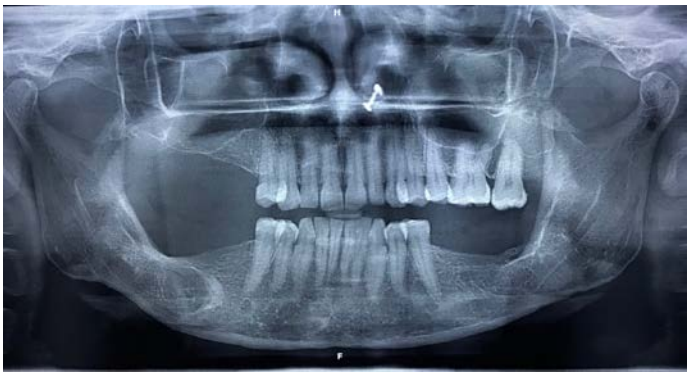

Figure 9: Two and half years follow up radiograph shows no sign of recurrence

Discussion

The overall incidence rate of multiple odontogenic keratocyst (Syndromic and Non syndromic) is found to be less as compared to solitary lesion. [4]

Brannon et al (1976) reported a clinicopathologic study of 312 lesions in 283 patients where in 20 patients out of 283 had multiple odontogenic keratocyst in which 10 were Syndromic and 10 non Syndromic. [5]

Myoung et al (2001) in his review of 256 cases for recurrence and clinicopathologic parameters of odontogenic keratocyst reported that multiple odontogenic keratocyst had developed in 39 of the 256 patients. In 11 cases (4.3%), the odontogenic keratocysts were confirmed to be associated with nevoid basal cell carcinoma syndrome. [6]

Habibi et al (2007) in his study of 83 cases of odontogenic keratocysts in 74 patients in an Iranian population found 6

cases of multiple odontogenic keratocyst where in all were associated with NBCCS. [7]

Year	Author	No. of patients	No. of cases with multiple odontogenic keratocysts	No. of Syndromic multiple odontogenic keratocyst
1976	Brannon et al	283	20	10
2001	Myoung et al	256	39	11
2007	Habibi et al	74	6	6

And many other such studies confirmed the association of multiple odontogenic keratocyst with syndromes such as Gorlin goltz-[8] Noonans[9] Orofacial Digital Syndrome[10] Ehlors Danlos[11] Simpson Golabi Behmal[12].

Gorlin goltz syndrome in our case was ruled out by Kimoni's criteria. Table 1& 2. [13]

Table 1: Kimoni's Major criteria to rule out Gorlin goltz syndrome

Major criteria	Our case
More than 2 basal cell carcinomas (BCCs) or 1 BCC in a patient < 20 years of age	Absent
Odontogenic keratocysts of the jaws (proven by histopathologic analysis)	Present
3 or more palmar or plantar pits	Absent
Bilamellar calcification of the falx cerebri	Absent
Bifid, fused or markedly splayed ribs	Absent
A first-degree relative with NBCCS	Absent

Table 2: Kimoni's Minor criteria to rule out Gorlin goltz syndrome

Minor criteria	Our case
Macrocephaly	Absent
Congenital malformations (e.g., cleft lip or palate, frontal bossing, coarse facies and moderate or severe hypertelorism)	Absent
Other skeletal abnormalities (e.g., Sprengel deformity, marked pectus deformity and marked syndactyly of the digits)	Absent
Radiologic abnormalities (e.g., bridging of the sella turcica, vertebral anomalies, modelling defects of the hands and feet, flame-shaped lucencies of the hands and the feet)	Absent
Ovarian fibroma or medulloblastoma (not applicable if patient is male)	Absent

Apart from NBCCS other syndromes were also ruled out after meticulous clinical examination.

The management of odontogenic keratocyst is a much debatable topic due its rate of recurrence as high as 62.5%**.** [14] The fragile lining of odontogenic keratocyst and presence of daughter cysts are the main causes of recurrence with almost 100% of recurrent cases manifesting the above mentioned. [15,16,17]

So a meticulous management should be directed towards eliminating the root cause of recurrence whereby only enucleation does not suffice. Enucleation with adjunctive procedures is usually required to overcome the problem of recurrence. [5,6,18,19]

Carnoy's solution as the only adjunctive to enucleation is a debatable solution to fragile lining and daughter cyst removal.

In a review of Recurrence probability for keratocystic odontogenic tumor of 6427 cases by Chrcanovic et al enucleation with application of Carnoy's solution showed 5.3% recurrence. [20]

In a systematic review and meta-analysis of what surgical treatment has the lowest recurrence rate following the management of keratocystic odontogenic tumor by Morraissi et al found 11.5% recurrence in enucleation with application of Carnoy's solution.[21]

Many others such as Zhao et al reported 6.70% recurrence, Apajalahti et al reported 39% recurrence, Sanchez-Burgos et al reported 100% recurrence when enucleation with application of Carnoy's solution was used.[22,23,24]

Though the enucleation with Carnoy's solution has been used majorly till date but a complete assurance of no recurrence cannot be deciphered from above mentioned studies and hence, addition of one more adjunctive procedure should be considered.

In further search of better treatment modality below mentioned studies can be coated for a more comprehensive treatment of odontogenic keratocyst.

A retrospective review of treatment of the odontogenic keratocyst by Morgan et al showed no recurrence in cases treated with enucleation followed by peripheral ostectomy with application of Carnoy's solution. Similar results were found in a study conducted by Najwa et al on 29 subjects with a follow up of 7 years. [25, 26]

Similar plan of treatment was performed in our case which was- enucleation followed by peripheral ostectomy with application of Carnoy's solution.

A follow up of two and half years shows no evidence of recurrence and development of any feature of NBCCS.

Conclusion

An aggressive management is usually required to address the cases of odontogenic keratocyst which does not end after the surgery. The management continues in the form of keen follow up due to high recurrence of odontogenic keratocyst . The follow up of cases with multiple odontogenic keratocysts is even more demanding as the recurrence as well as development of any feature of Nevoid basal cell carcinoma syndrome has to be checked for.

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References

1. Speight PM, Takata T. New tumour entities in the 4th edition of the World Health Organization Classification of Head and Neck tumours: odontogenic and maxillofacial bone tumours. Virchows Archiv. 2018 Mar 1;472(3):331-9.[doi:10.1007/s00428-017-2182-3]
2. Auluck A, Suhas S, Pai KM. Multiple odontogenic keratocysts: report of a case. Journal of the Canadian Dental Association. 2006 Sep 1;72(7).
3. Vandana S, Jahan AJ, Kavya L, Palla S. Non-syndromic multiple odontogenic keratocysts-a case report. International Journal of Medical and Dental Case Reports. 2017;4(1):1-4.
4. Sohanian S, Seifi S, Dezfouli MK, Kiani M, Khakbaz O. Non-syndromic Multiple Keratocystic Odontogenic Tumor: A Case Report. Journal of Otolaryngology Advances. 2017 Aug 19;2(1):7. [doi:10.14302/issn.2379-8572.joa-17-1668]
5. Brannon RB. The odontogenic keratocyst: A clinicopathologic study of 312 cases. Part I. Clinical features. Oral Surgery, Oral Medicine, Oral Pathology. 1976 Jul 1;42(1):54-72.
6. Myoung H, Hong SP, Hong SD, Lee JI, Lim CY, Choung PH, Lee JH, Choi JY, Seo BM, Kim MJ. Odontogenic keratocyst: review of 256 cases for recurrence and clinicopathologic parameters. Oral Surgery, Oral Medicine, Oral Pathology, Oral

Radiology, and Endodontology. 2001 Mar 1;91(3):328-33. [doi:10.1067/moe.2001.113109]

7. Habibi A, Saghravanian N, Habibi M, Mellati E, Habibi M. Keratocystic odontogenic tumor: a 10-year retrospective study of 83 cases in an Iranian population. Journal of oral science. 2007;49(3):229-35.[Medline:17928730]
8. McGrath CJ, Myall RW. Conservative management of recurrent keratocysts in Basal-cell naevus syndrome. Australian dental journal. 1997 Dec;42(6):399-403.
9. Lindeboom JA, Kroon FH, de Vires J, van den Akker HP. Multiple recurrent and de novo odontogenic keratocysts associated with oral-facial-digital syndrome. Oral Surgery, Oral Medicine, Oral Pathology, Oral Radiology, and Endodontology. 2003 Apr 1;95(4):458-62.
10. Connor JM, Evans DP, Goose DH. Multiple odontogenic keratocysts in a case of the Noonan syndrome. British Journal of Oral Surgery. 1982 Sep 1;20(3):213-6.
11. Carr RJ, Green DM. Multiple odontogenic keratocysts in a patient with type II (mitis) Ehlers-Danlos syndrome. British Journal of Oral and Maxillofacial Surgery. 1988 Jun 1;26(3):205-14.
12. Krimmel M, Reinert S. Multiple odontogenic keratocysts in mental retardation–overgrowth (Simpson–Golabi–Behmel) syndrome. British Journal of Oral and Maxillofacial Surgery. 2000 Jun 1;38(3):221-3.
13. Kimonis VE, Goldstein AM, Pastakia B, Yang ML, Kase R, DiGiovanna JJ, Bale AE, Bale SJ. Clinical manifestations in 105 persons with nevoid basal cell carcinoma syndrome. American journal of medical genetics. 1997 Mar 31;69(3):299-308.
14. Pindborg JJ, Hansen J. Studies on odontogenic cyst epithelium: 2. Clinical and roentgenologic aspects of odontogenic keratocysts. Acta Pathologica Microbiologica Scandinavica. 1963 Sep;58(3):283-94.
15. Stoelinga PJ. Long-term follow-up on keratocysts treated according to a defined protocol. International journal of oral and maxillofacial surgery. 2001 Feb 1;30(1):14-25.
16. Stoelinga PJ. Recurrences and multiplicity of cysts. InTransactions of the International Conference on Oral Surgery 1973 (Vol. 4, pp. 77-80).
17. Stoelinga PJ, Bronkhorst FB. The incidence, multiple presentation and recurrence of aggressive cysts of the jaws. Journal of Cranio-Maxillofacial Surgery. 1988 Jan 1;16:184-95.
18. Abdullah WA. Surgical treatment of keratocystic odontogenic tumour: A review article. The Saudi dental journal. 2011 Apr 1;23(2):61-5.
19. Stoelinga PJ. The treatment of odontogenic keratocysts by excision of the overlying, attached mucosa, enucleation, and treatment of the bony defect with Carnoy solution. Journal of oral and maxillofacial surgery. 2005 Nov 1;63(11):1662-6.
20. Chrcanovic BR, Gomez RS. Recurrence probability for keratocystic odontogenic tumors: An analysis of 6427 cases. Journal of Cranio-Maxillofacial Surgery. 2017 Feb 1;45(2):244-51.
21. Al-Moraissi, E.A., Dahan, A.A., Alwadeai, M.S., Oginni, F.O., Al-Jamali, J.M., Alkhutari, A.S., Al-Tairi, N.H., Almaweri, A.A. and Al-Sanabani, J.S., 2017. What surgical treatment has the lowest recurrence rate following the management of keratocystic odontogenic tumor?: A large systematic review and meta-analysis. Journal of Cranio-Maxillofacial Surgery, 45(1), pp.131-144.

22. Zhao YF, Wei JX, Wang SP. Treatment of odontogenic keratocysts: a follow-up of 255 Chinese patients. Oral Surgery, Oral Medicine, Oral Pathology, Oral Radiology, and Endodontology. 2002 Aug 1;94(2):151-6.
23. Apajalahti S, Hagström J, Lindqvist C, Suomalainen A. Computerized tomography findings and recurrence of keratocystic odontogenic tumor of the mandible and maxillofacial region in a series of 46 patients. Oral Surgery, Oral Medicine, Oral Pathology, Oral Radiology, and Endodontology. 2011 Mar 1;111(3):e29-37.
24. Sanchez-Burgos R, Gonzalez-Martin-Moro J, Perez-Fernandez E, Burgueno-Garcia M. Clinical, radiological and therapeutic features of keratocystic odontogenic tumours: a study over a decade. Journal of clinical and experimental dentistry. 2014 Jul;6(3):e259.
25. Morgan TA, Burton CC, Qian F. A retrospective review of treatment of the odontogenic keratocyst. Journal of oral and maxillofacial surgery. 2005 May 1;63(5):635-9.
26. Alchalabi NJ, Merza AM, Issa SA. Using carnoy's solution in treatment of keratocystic odontogenic tumor. Annals of maxillofacial surgery. 2017 Jan;7(1):51.