CASE REPORT

Multiple non-syndromic odontogenic keratocysts in three siblings

Amit Nirwan,¹ Sangeeta Panjab Wanjari,² Rashmi Saikhedkar,¹ Vinayak Karun¹

¹Department of Oral & Maxillofacial Surgery, College of Dental Science & Hospital, Indore, Madhya Pradesh, India
²Department of Oral Pathology, Modern Dental College & Research Centre, Indore, Madhya Pradesh, India

Correspondence to
 Sangeeta Panjab Wanjari,
 drsangeetawanjari@gmail.com

SUMMARY

Occurrence of multiple cysts (MC) involving the jaw is rare. When multiple, it is usually associated with a syndrome. Occurrence of MC without syndromic association is extremely rare. Multiple odontogenic cysts mostly could be odontogenic keratocysts or dentigerous cysts. Odontogenic keratocyst shows involvement of mandible over maxilla, with peak incidence in second and third decade and it is exceedingly rare before 10 years of age. However multiple odontogenic keratocysts found in children are often reflective of nevoid basal cell carcinoma syndrome. Here is a case report which documents multiple jaw cysts involving both the jaws, in three siblings of ages 10, 13 and 17 years with negative parental history. All three reported cases were free of any systemic involvement. As odontogenic keratocyst spreads through bone marrow, destruction is more before any clinical manifestation. Therefore, early detection and intervention are essential in preventing extensive destruction.

BACKGROUND

Odontogenic cysts (OCs) are a commonly occurring pathology in the jaws. In general, cyst presents as solitary lesion in the jaws, while multiple cysts (MCs) are a rare presentation. Few of the commonly reported multiple odontogenic jaw cysts (MOJCs) are odontogenic keratocyst (OKC) and dentigerous cyst (DC).

Multiple OCs tends to occur among all cysts with some frequency. It is also reported that cystic lesions found in children are often reflective of multiple OKC as a component of Gorlin and Goltz syndrome (GGS). However, in some reported cases MCs have been found independent of any associated syndrome.¹

It is stated that 5% of the patients with OKC tend to have multiple OKCs.² OKC is known for its high-recurrence rate (HRR). Moreover in patients with multiple OKCs a significantly HRR of 30% has been reported compared to solitary keratocyst of 10%.

We hereby report a case of non-syndromic multiple jaw cysts (NSMJCs) in the three siblings which were treated successfully by early detection and intervention.

CASE PRESENTATION

Case 1

A 10-year-old girl reported with a chief complaint of swelling and pain on the left middle region of face. Swelling started appearing approximately 6 months previously and was slowly increasing in size with moderate pain.

She was a moderately built otherwise healthy girl without any other systemic pathology. All her vitals were within normal limits. Extraoral examination revealed diffuse swelling in left mid-facial region causing obvious facial asymmetry (figure 1). Swelling was firm and tender without any rise in local temperature. Intraoral examination showed vestibular obliteration from canine till molar region. Swelling was fluctuant, tender with stretched overlying mucosa. Mixed dentition was evident with mobility of left maxillary deciduous canine and molars (figure 2). A thorough general examination ruled out for any other systemic pathological involvement.

Case 2

Just after 5 months, one of her elder brothers aged 13 years reported with a chief complaint of pain and swelling in the maxillary right cheek region since 3 months. Swelling was slowly increasing in size with moderate, intermittent pain. No history of trauma was elicited.

He was a moderately built healthy boy without any other systemic pathology, with all vitals within normal limits. Extra oral examination revealed diffuse swelling in right mid-facial region causing obvious facial asymmetry. Swelling was firm and tender without any rise in local temperature. Intra oral examination revealed missing 13, over-retained 53 and vestibular obliteration from 12 to 17. Swelling was soft to firm in consistency, slightly tender with stretched overlying mucosa (figure 3). No mobility of teeth was associated.

With a conjecture of MC involving a familial tendency component, her eldest and youngest brothers, aged 17 and 8 years respectively were also screened.

Figure 1 Extraoral view of case 1 showing swelling on left side of the face.

To cite: Nirwan A, Wanjari SP, Saikhedkar R, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2012-007503

Figure 2 Intraoral view of case 1 showing swelling obliterating left maxillary vestibule.

Case 3

The girl's 17-year-old eldest brother too complained of slowly growing, painless swelling in the mandibular left posterior area, since 1 month associated with mild continuous pain, since 1 week.

Extra oral examination revealed diffuse swelling in left mandibular molar-ramus region causing obvious facial asymmetry. Swelling was tender, firm, without rise in local temperature. Intra oral examination revealed slight expansion of buccal cortical plate in 35–38 regions, which was firm in consistency. Overlying mucosa appeared stretched. All the teeth except third molars were present without mobility. Thorough screening has ruled out the systemic involvement.

Case 4

However, youngest brother aged 8 years was free of any symptoms. He too was screened thoroughly to eliminate any other systemic pathology.

INVESTIGATIONS

Radiographic examination of *case 1* revealed single well-defined radiolucency in left maxillary region involving antrum. Superior displacement of 23, 24 and 25 was noted with almost 180° rotated 23 near the orbital floor. A thorough radiographic examination revealed two well-defined radiolucencies—one in

Figure 3 Intraoral view of case-2 showing swelling involving buccal maxillary alveolar ridge on right side.

Figure 4 Orthopantomogram of case-1 having multiple cysts.

33 region and second in the left ramus area distal to developing 37 with scalloped borders (figure 4).

Her postero anterior chest radiograph showed bifid third and fourth ribs bilaterally, which were asymptomatic. She was diagnosed as NSMJC (figure 5).

Radiographic examination of *case 2* revealed multiple well-defined radiolucencies—two in maxilla bilaterally and three in mandible. In maxilla, one was in 13 region displacing the tooth superiorly while 53 was over-retained and other in 24–25 regions (figure 6). Moreover in mandible two radiolucencies appeared bilateral distal to 37 and 47 and third between 43 and 44 displacing the teeth. The radiolucency distal to 37 showed scalloped margins. As he was without any associated systemic pathology, he was diagnosed as a case of NSMJC.

Radiographic examination of *case 3* too revealed multiple radiolucencies in both the jaws of which two radiolucencies were in the mandible on left side, one in distal to 37 with scalloped margins, and horizontally impacted 38 and second between 33 and 34 displacing the teeth. Single small well-defined radiolucency was observed between 46 and 47. Two more bilateral radiolucencies were evident in maxilla, one in 13 region displacing it superiorly, while other was in 25 region (figure 7). As he was also without any associated systemic disease, the pathology was diagnosed as NSMJC.

DIFFERENTIAL DIAGNOSIS

Differential diagnosis included dentigerous cyst.

TREATMENT

Surgery was planned under general anaesthesia for case 1. The left maxillary lesion was completely enucleated.

Figure 5 Posteroanterior chest radiograph of case 1 showing bifid third and fourth ribs bilaterally.

Figure 6 Orthopantomogram of case 2 showing multiple radiolucencies involving maxilla and mandible.

While in case 2 right-sided maxillary symptomatic lesion and in case 3 left mandibular body lesion was surgically enucleated under general anaesthesia.

Rest of all the asymptomatic cystic lesions in all the three siblings were enucleated in the next few weeks. Histopathologically all the lesions were diagnosed as OKC.

OUTCOME AND FOLLOW-UP

Histopathological evaluation of all lesions showed thin uniform cystic lining of stratified squamous epithelium with picket fence appearance of basal cells and corrugated parakeratin at the surface. Epithelial-connective tissue interphase appeared almost flat. Underlying connective capsule was thin and dense fibrous with interspersed cellularity. Features were contributing for the diagnosis of OKC (figure 8).

All the four children were routinely reviewed every month for 3 months. Periodic follow-up of them now for 2 years at the interval of 3 months revealed prognosis to be excellent with no recurrence.

DISCUSSION

Cyst is defined as a pathological cavity containing fluid or semi-fluid as its content and may or may not be lined by an epithelium.³ Cysts generally arise from different developmental structures and so vary in type, clinical, radiographic and histological features.

Cysts usually present as a single pathology; however, occasionally MCs could be associated. OKC and DC tend to form MCs in the jaws. MCs can be either associated with a syndrome or may be non-syndromic; however, NSMJCs are rare as compared to syndromic MCs.⁴

Whether syndromic or not, MCs remain as a rare entity found in the jaws. Ivy⁵ was the first to report an occurrence of multiple DC in siblings and Beyrent in 1980 reported a case of MJCs in four members of a family. Mencer and Brown⁶ reported a case of multiple DC with pathological fracture.

Figure 7 Orthopantomogram of case 3 showing multiple radiolucencies involving mandible and maxilla.

Figure 8 Microphotograph showing corrugated parakeratinised thin stratified squamous epithelial lining with dense collagenous capsule of odontogenic keratocyst (H&E stain, ×150).

Moreover, OKC is the most common form of MCs. Further Brannon too in his study of 312 cases reported that most common MCs occurring together are the OKCs and are generally seen in very young age group. Further, he had reported that 5.8% of the cysts were from patients with MCs but were non-syndromic.⁷

Clinical and radiographic features and characteristics of solitary or multiple OKC are not very different, and show more common occurrence in molar-ramus area in mandible. There may be extensive involvement of bone with little or no bony expansion making them generally asymptomatic unless secondarily infected. However, at times it can expand and perforate both buccal and lingual cortical plates and involve adjacent soft tissue.⁸

Histopathologically, two different variants might be seen even in the same jaw. Such a case of multiple OKC with diverse histological features showing orthokeratinised and parakeratinised types were reported by Rudagi and Kiyi.⁹ As in regard recurrence rate is concerned, it is said non-syndromic OKCs are less aggressive and have a less recurrence rate as compared to syndrome-associated OKC.¹⁰

Other cyst type known to form MCs is the dentigerous variety which generally shows solitary lesion although occasional cases of multiple DCs have also been reported. DCs characteristically show fluid accumulation between crown and reduced enamel epithelium of a tooth with most common occurrence in third molar followed by maxillary canine. Expansion is the most important feature of these lesions. Radiographic presentation would be as well-defined radiolucency with hyperostotic borders unless secondarily infected.⁸ Tournas *et al*¹¹ reported a case of multiple unilateral maxillary DCs in non-syndromic patient. They have further stated that presence of NSMJC is extremely rare and occur exclusively in the mandible. MCs also have a propensity to occur in mandible than maxilla. Ustuner *et al*¹² differs in this aspect and reported that occurrence of MCs have the maxilla : mandible ratio of 2 : 1 with mandibular angle and body region being the most common site. In the present series multiple OKCs were noticed involving both the jaws with equal propensity.

A case of bilateral dentigerous non-syndromic cysts was reported by Freitas and Tempest,¹³ which was associated with maxillary third molar and mandibular second molar. They claimed such a case of multiple DC in maxilla and mandible has not been reported before.

No specific aetiology for the formation of MCs has been defined as such but some reports claim it to be associated with chronic use of medicines. Formation of MCs has been associated with long-term use of drugs such as cyclosporine and calcium

channel blockers. One such case was reported by DeBiase, Ottolenghi *et al.*¹⁴

Occurrence of NSMJC is rare as compared to multiple syndromic cysts. Few cases of syndrome associated with MCs have been reported in the literature. Few syndromes associated with MCs are Nevoid basal cell carcinoma syndrome (NBCCS) also known as GGS, orofacial digital syndrome, Gardners syndrome, etc. Among the cysts, OKCs are commonly found associated with GGS, while multiple DCs are found associated with other syndromes.

Multiple OKCs associated with GGS tend to occur in the age group of less than 10 years while those not associated with any syndrome tend to occur at a slightly higher age group.¹⁵ However, Lorenzolo *et al.*¹⁶ reported that presence of MCs are often the first sign of underlying GGS in 78% of cases and can be detected in age group less than 10 years. In the present article too all the patients were of higher age which was ranging between 10 and 17 years.

Multiple DCs also found associated with few syndromes as Mucopolysaccharidosis 6, Gardners syndrome and Cleido-cranial dysplasia. Sands, Tacchio¹⁷ reported a case of syndromic multiple DCs but also claimed syndromic DCs remain a rare entity.

Multiple radiolucencies in the jaws can also be formed by various other pathologies and should be differentiated from multiple cystic lesions. Few lesions forming multiple jaw radiolucencies include multiple myeloma, metastatic carcinoma, langerhans disease, etc.¹⁸

In our case report three of the four siblings suffered from multiple non-syndromic OKC. This clearly indicates the role of familial tendency in the formation of MC. Many a times dentists might be the first one to diagnose such cases of MC. Such cases should be then carefully evaluated clinically, radiologically and histologically and managed accordingly.

Learning point

- Occurrence of non-syndromic multiple jaw cysts in the siblings is extremely rare. If the lesion is diagnosed in one of the members of the family, other members should be screened although asymptomatic, so that early diagnosis can be made with timely institution of the treatment. This will help in preventing extensive destruction giving best treatment results.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Rajendran R. Cysts & tumors of the odontogenic origin. In: Shafer WG, Hine MK, Levy BM, Rajendran R, Sivapathasundharam B, eds. *Shafer's textbook of oral pathology*. 6th edn. Delhi, India: Elsevier, 2009;258–60.
- 2 Regezi JA. Cysts of the jaws & the neck. In: Regezi JA, Sciubba JJ, Jordan RCK, Penny R, Kimberly A. *Oral pathology: Clinical pathologic correlations*, 4th edn. USA: WB Saunders & Company, 2003:250.
- 3 Shear M. Odontogenic keratocyst. In: Shear M, Speight PM. *Cysts of the oral and maxillofacial regions*. 3rd edn. UK: Blackwell publishing Ltd, 2007:6–11.
- 4 Lester WK, Laskin DM. Cysts of the jaws & oral & facial soft tissues. In: Laskin DM, Warfel D, eds. *Oral & maxillofacial surgery-vol 2*. 5th edn. Delhi, India: C Mosby V & Company, 2003:463–5.
- 5 Ivy RH. Multiple dentigerous cysts with special reference to occurrence in siblings. *Ann Surg* 1939;109:114–25.
- 6 Mincer HM, Brown A. Multiple dentigerous cysts with pathological fractures. *Carolina Dent J* 1963;21:9.
- 7 Brannon RB. The odontogenic keratocyst, a clinicopathological study of 312 cases part 1. *Oral Pathol Oral Surg Oral Med Radiol* 1976;42:54–72.
- 8 Ghom AG. Cysts of jaws. In: Ghom AG. *Textbook of oral medicine*. 1st edn, Delhi, India: Jaypee Brothers Medical Publishers (P) Ltd, 2005:193–9.
- 9 Rudagi BM, Khark VR. Multiple OKC with diverse histological features in non syndromic patients. *Prevera Med Rev* 2010;5:35.
- 10 Dominguez FV, Keszler A. Comparative study of keratocysts, associated and not associated with NBCC syndrome. *J Oral Pathol* 1988;17:39–42.
- 11 Tournas AS, Tewfik MA, Chauvin PJ, *et al.* Multiple unilateral maxillary dental cysts in non syndromic patient. A case report and review of literature. *Int J Paediatr Otolaryngol* 2006;91:100–6.
- 12 Ustuner E, Fitoz S, Atasoy C, *et al.* Bilateral maxillary dental cysts a case report. *Oral Pathol Oral Surg Oral Med* 2003;95:632–5.
- 13 Freitas DQ, Tempest LM, Sicoli E. Bilateral dentigerous cysts, review of literature and report of unusual case. *J Dentomaxillofac Radiol* 2006;35:464–8.
- 14 DeBiase A, Ottolenghi L, Polimeni A, *et al.* Bilateral mandibular cysts associated with cyclosporine use. A case report. *J Paediatr Otolaryngol* 2001;16:993–5.
- 15 Myoung H, Hong SP, Kim MUJ, *et al.* OKC, review of 256 cases for recurrence and clinicopathological parameters. *J Oral Pathol Oral Surg Oral Med* 2001;91:328–33.
- 16 Lorenzolo M, Nocini P, Bucci P, *et al.* Early diagnosis of nevoid basal cell carcinoma syndrome. *J Am Dent Assoc* 1999;130:669–74.
- 17 Sands T, Tacchio C. Multiple dentigerous cysts in a child a case report. *J Oral Health* 1998;88:27–30.
- 18 Wood NK, Goaz PW, Jacobs MC. Multiple separate well defined radiolucencies. In: Wood NK, and Goaz PW, eds. *Differential diagnosis of oral & maxillofacial lesions*. 5th edn, Delhi, India: Mosby & Company, 1997:382–4.

Copyright 2013 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- Submit as many cases as you like
- Enjoy fast sympathetic peer review and rapid publication of accepted articles
- Access all the published articles
- Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow