

Radiological Perspectives in Non Syndromic Multiple Odontogenic Keratocysts:

Report of Two Cases and Review of literature

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Abstract

Odontogenic keratocyst (OKC) is a common developmental odontogenic cyst affecting the maxillofacial region. Occurrence of multiple OKCs in the jaws is usually associated with nevoid basal cell carcinoma syndrome (NBCCS), but non syndromic association is extremely rare. This report presents the occurrence of multiple OKCs in two non-syndromic patients.

Keywords: Multiple odontogenic keratocysts, Keratocystic odontogenic tumour, Nevoid basal cell carcinoma syndrome

Introduction

The term “odontogenic keratocyst” was first coined by Philipsen in 1956, when he segregated seven jaw cysts from cholesteatomas that occurred in other cranial areas [1]. Keratocystic odontogenic tumor (KCOT) was introduced by WHO in 2005 and defined as ‘a benign unicystic or multicystic, intraosseous tumor of odontogenic origin, with a characteristic lining of parakeratinized stratified squamous epithelium and potential for aggressive, infiltrative behaviour [2]. These cysts are generally derived from epithelial remnants of the tooth germ and another origin is from basal cells of the overlying oral epithelium [3]. Nevoid basal cell carcinoma syndrome (NBCCS) is characterised by multiple KCOTs with associated cutaneous, skeletal, ocular and neurologic abnormalities. This syndrome was first described by Gorlin and Goltz in 1960, so it is also known as Gorlin-Goltz syndrome. Typically, multiple KCOTs usually occur in association with NBCCS, but rarely occur without syndromic association which has been reported previously in literature [4]. This article presents two cases of non syndromic multiple OKCs highlighting the imaging features.

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Received: 27.06.19

Accepted: 14.08.19

Case Report - 1

A 19 year old male patient reported to our department with a chief complaint of pain in the right lower back tooth region for the past 4 months. On extraoral examination, there was no facial asymmetry and no obvious swelling. Intraoral examination revealed retained deciduous tooth 84. Panoramic radiograph revealed three distinct cystic lesions in the mandible.

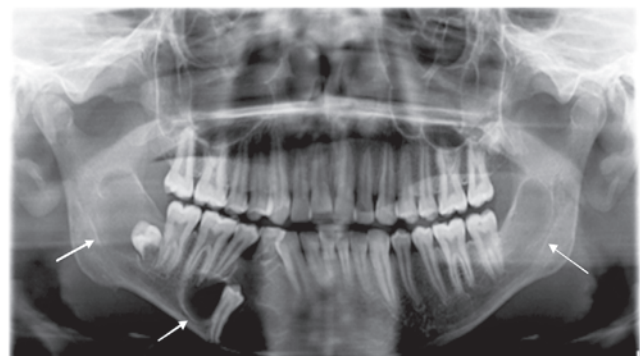


Figure 1 : Panoramic radiograph showing three cystic lesions

Cone beam computed tomography revealed evidence of well defined hypodense lesions with sclerotic borders present in the right ramus of mandible with impacted 48 and in the right body of mandible with impacted 44 and perforation of buccal cortical plate. The third well defined hypodense lesion was present in the left ascending ramus with thinning of lingual cortical plate.

With the above findings the case was provisionally diagnosed as multiple odontogenic keratocysts. There

Table 1: Description of panoramic radiographic findings in Case 1.

Region	Radiographic Appearance	Approximate size (cm)	Effect on adjacent structures
Right ramus of mandible involving impacted 48	Well defined unilocular radiolucency with sclerotic borders	3.6 x 2.3 cm	none
Right body of mandible with impacted 44	Well defined multilocular radiolucency with sclerotic borders	4.1 x 3 cm	mandibular canal displaced inferiorly
Left ascending ramus	Well defined multilocular radiolucency with sclerotic borders	3.5 x 2.4 cm	none



Figure 2



Figure 3

Figure 2 : 3D reconstructed CBCT showing Figure 3 : Axial CBCT showing lesion in the buccal cortical perforation mandibular body with incomplete septae

were no cutaneous abnormalities, the haematologic tests were within normal limits and chest and skull radiography revealed no abnormalities. All the three lesions were enucleated with removal of impacted teeth. Enucleation was done in order to reduce the morbidity associated with resection and to preserve the integrity of the mandible, considering the young age of the patient. Histopathological report revealed cystic lesion characterized by an epithelial lining with 5-7 layers thickness with basal cells showing hyperchromatism and palisading pattern with surface parakeratinization and corrugation. The underlying connective tissue was fibrous and showed inflammation.

Case Report - 2

A 14 year old male patient reported to our department with the chief complaint of swelling in his right lower back tooth region for past days 15 days. Patient gave history of similar swelling in his right maxillary region for which he underwent enucleation 2 years back.

On extraoral examination, a diffuse swelling was seen on right mandible which extends from the parasymphysis

to the body of mandible. The swelling was non tender and soft in consistency. Intraorally, swelling was seen in 45,46 region with obliteration of the buccal vestibule. The swelling was soft, tender, fluctuant and there was an intra oral draining sinus.



Figure 4: Diffuse swelling of the right



Figure 5: Intra-oral sinus in 45,46 region side of face

Table 2: Description of panoramic radiographic findings in Case 2.

Region	Radiographic Appearance	Approximate size (cm)	Effect on adjacent structures
Right angle of mandible	Illdefined radiolucency with sclerotic borders	3.1 x 2.1 cm	Mandibular canal displaced inferiorly
Right body of mandible	Well defined radiolucency	1.3 x 2.1 cm	none
Right maxillary sinus	illdefined radiolucency	1 x 1.5cm	none
Left angle of mandible	Well defined radiolucency with sclerotic border	3.8 x 2.2 cm	none

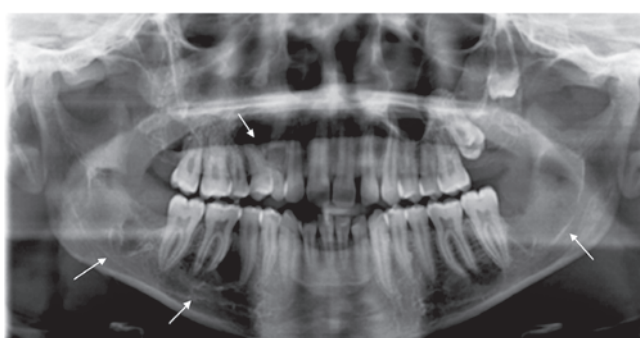


Figure 6: Panoramic radiograph showing multiple cystic lesions

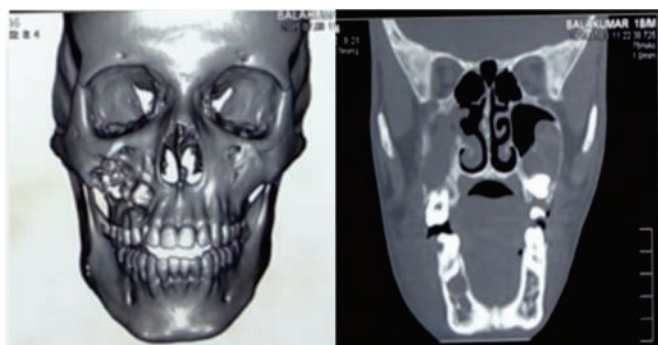


Figure 7-8 : 3D reconstructed & coronal CT showing perforation of the right maxillary sinus

Computed tomography revealed multiple well defined hypodense areas present in the right angle, right and left body of mandible and maxillary sinus with areas of perforation. The case was provisionally diagnosed as recurrent multiple OKCs. There were no other systemic signs and symptoms, haematological tests were within normal limits and chest and skull radiography findings were unremarkable.

All the lesions were enucleated keeping in mind the need for preserving vital structures and reducing deformities associated with more aggressive treatment modalities as our patient was of adolescent age. Histopathology report revealed cystic lesion with an orthokeratinised stratified squamous epithelial lining of uniform thickness (4 – 6 layers) with surface corrugation. The basal cells were cuboidal with hyperchromatic nuclei. The connective tissue wall was fibrous.

Discussion

OKCs of the jaws are one of the most common developmental odontogenic cysts and comprises of 10-15% of all jaw cysts [5]. Syndromes associated with multiple OKCs include NBCCS or Gorlin-Goltz syndrome, orofacial digital syndrome, Ehler-Danlos syndrome, Noonan syndrome, Simpson-Golabi-Behmle

syndrome [6]. Rarely, multiple KCOTs are seen with non syndromic manifestations. Brannon reported that 5.8 percent of 312 cases of KCOTs, had multiple cysts with non syndromic manifestations [7]. Our paper presents two such rare cases of non syndromic association of KCOTs.

It arises from cell rests of the dental lamina and occurs in a wide age group with peak incidence in second and third decade of life [8]. Molecular basis of the behaviour associated with KCOT results in mutations in the tumor suppressor gene PTCH and are identified as the underlying genetic events in NBCCS. It has been proposed that the development of an OKC would follow the “2- hit” hypothesis. According to this hypothesis, OKCs present in NBCC arise from precursor cells that contain a hereditary “first hit” and the allelic loss represents loss of normal allele while sporadic OKC might arise from susceptible cell in which two somatic “ hits” have occurred. Shear suggested that the two cysts from one syndrome patient that occurred on opposite sides of the mandible had the same pattern of allelic loss, suggesting that this genetic mutation occurred at a very early stage of embryogenesis [9].

Clinically, they present as a swelling, with or without pain. The cyst classically grows within the medullary

spaces of the bone in an anteroposterior direction, causing minimal expansion. Buccal expansion is seen approximately 30% maxillary and 50% mandibular lesions [10]. They occur more commonly in the mandible than maxilla in the ratio 2:1 and has a predilection for the angle and ascending ramus [11]. In this paper, case1 presented with lesions in mandible, whereas case2 presented with lesions in both maxilla and mandible.

Radiologically, KCOTs are described as a well-defined radiolucent area with smooth and often corticated margins and may be unilocular or multilocular. In 25 to 40% of cases, an unerupted tooth is seen associated with the lesion. Rarely root resorption is seen. White and Pharaoh stated that KCOTs exhibit a “propensity to grow along the internal aspect of the jaws, causing minimal expansion” which is used to differentiate from other cysts [12]. In our first case two of the lesions in mandible were associated with impacted tooth.

Histologically, KCOT has been divided into parakeratotic and orthokeratotic. These types refer to the histologic characteristics of the lining and the type of keratin produced. The orthokeratotic subtype produces keratin more closely resembling the normal keratin produced by the skin, with a keratohyaline granular layer immediately adjacent to the layers of keratin, which do not contain nuclei. The parakeratotic subtype has more disordered production of keratin; no keratohyaline granules are present, and cells slough into the keratin contains nuclei and is referred to as parakeratin. The parakeratotic type is the most frequent (80%) and has a more aggressive clinical presentation than the orthokeratotic variants [15].

Treatment modalities of KCOT includes enucleation, and marsupialization or resection. Treatment plan depends on the age of the patient, localization and size of the lesion. Enucleation is the most performed treatment and [14] was done in both our cases. It is reported that 5% of the OKC patients will tend to have multiple OKCs. OKC is known to have high-recurrence rate (HRR). Moreover, patients with multiple OKCs has 30% HRR compared to solitary keratocyst of 10% which has been reported [15]. Hence there is a need for diligent and long term follow up of these patients. Both our patients are under follow up for the past four years and no complications were reported so far.

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Conflict of interest:	All authors declare no COI
Ethics:	There is no ethical violation as it is based on voluntary anonymous interviews
Funding:	No external funding
Guarantor:	Dr. Vidya Jayaram will act as guarantor of this article on behalf of all co-authors.

