CASE REPORT

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Non-syndromic multiple odontogenic keratocyst finding with Cone-beam Computed Tomography (CBCT): A rare case report

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ABSTRACT

Objectives: The aim of this case report is to describe the radiograph pattern of non-syndromic multiple odontogenic keratocyst (OKC). The oral and maxillofacial region was frequently affected by the developmental odontogenic cyst known as OKC, which develops from the dental lamina or its remnants. The nevoid basal cell carcinoma syndrome (NBCCS) or the Gorlin-Goltz syndrome was typically associated with multiple OKC; however, in about 5% of patients, there were many cysts without a concurrent syndromic presentation.

Case Report: A 38- year-old female came to the dental radiology installation of the Dental and Oral Hospital, Universitas Padjadjaran, who had been referred for a CBCT examination. She had a history with a dentist about 1 month ago, complaining of dislodged fillings on anterior mandible teeth and complaints of missing several posterior mandible teeth. The patient had a previous panoramic examination; multiple radiolucent lesions were found incidentally. She has no pain, and clinical features showed no evidence of swelling; then, the dentist recommended a CBCT examination with a suspect dental cyst on a posterior mandible dextra.

Conclusion: Based on the examination results, it was concluded that the cone beam computed tomography examination showed the radiolucent lesion, well-defined with a scallop border; this case was radiodiagnosis as suspected non-syndromic multiple odontogenic keratocyst.

Keywords: *Odontogenic keratocyst, primordial cyst, CBCT 3D*
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INTRODUCTION

Odontogenic Keratocyst (OKC) is a developmental cyst of odontogenic origin that accounts for 10–15% of all jaw cysts and is characterised by aggressive clinical behaviour and a high recurrence rate. It develops from the dental lamina's cell rests. OKC was called a keratocystic odontogenic tumour (KCOT) by the World Health Organization (WHO) in 2005 due to its aggressiveness, infiltrative nature, histology, and genetic behaviour. The WHO described OKC as having a distinctive lining of para-keratinized stratified squamous epithelium with a tendency for aggressive, infiltrative activity.[1,2]

OKC can manifest in sporadic or numerous ways in cases connected with syndromes and non-syndromes. Nevoid Basal Cell Carcinoma Syndrome (NBCCS) or the Gorlin-Goltz syndrome has been linked to multiple OKC, but they have also been associated with the Orofacial Digital Syndrome, Ehler-Danlos syndrome, Noonan syndrome, Simpson-Golabi-Behmel syndrome. Multiple odontogenic keratocysts are normally related to concomitant cutaneous, skeletal, ocular, and neurological anomalies as part of nevoid basal cell carcinoma syndrome or the Gorlin-Goltz syndrome. Non-syndromic multiple odontogenic keratocyst is uncommon; only about 5% of patients report this type. Compared to non-syndromic OKC, OKC-related NBCCS exhibit more aggressive behaviour and greater recurrence rates.[3,4]

Multiple basal nevi, multiple OKC, and skeletal abnormalities are the symptoms characterising Gorlin-Goltz syndrome. If not, the multifocal nature of OKC may be to blame for these numerous lesions. They could also be the initial signs of Gorlin -Goltz syndrome.[2] various developmental problems are associated with this syndrome, which is known to have a high degree of penetrance and varying expressivity. There is 65-75% OKC in patients with Gorlin-Goltz syndrome. These typically appear around the lower third molar and maxillary canine, but they can be bilateral, affecting both jaws and sometimes bilateral. At least ten years before cases of isolated OKCs, Gorlin-Goltz syndrome-related OKCs start to appear.[5]

OKC originate from the remaining dental lamina and often affects people of all ages, although they disproportionately affect men. The ramus and posterior mandible body is where odontogenic keratocyst most frequently occurs. Odontogenic keratocysts may become invasive neoplasms such as ameloblastoma and squamous cell carcinoma.

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Multiple OKCs are rare and occur either in syndromic or non-syndromic groups. The aggressive behaviour of OKC might be expected to correlate with the proliferation of cyst epithelium. The syndromic and non-syndromic types of multiple keratocyst are more invasive than the solitary types. Presentation with Multiple OKCs should be thoroughly scrutinized for categorizing into syndromic and non-syndromic varieties.[2,6,7]

Multiple KCOTs are mostly seen with Gorlin Goltz syndrome. However, a few cases of non-syndromal multiple keratocystic odontogenic tumours have rarely been reported in the literature. Multiple OKC is very rare and may be the first symptom of the syndromic condition. Since the non -syndromic type is less common.[7,8] this study was conducted to report a non-syndromic multiple OKC involving the mandible in a 38-year-old female patient.

This article aims to report a rare case of multiple odontogenic keratocyst (OKC) in a non-syndrome patient and highlights a comparison with syndromic multiple OKC and also as a guideline for the dentist to know the importance of diagnosing the disease with the help of CBCT 3D and enforcing a strict long-term follow-up whenever such a case like this is found. This case report explains the radiographic appearance of a non-syndromic multiple odontogenic keratocyst found on a cone beam computed tomography (CBCT) exam.

CASE REPORT

A 38-year-old female came to the dental radiology installation of the Dental and Oral Hospital, Universitas Padjadjaran, who had been referred for a CBCT examination. She had a history with a dentist about 1 month ago, complaining of dislodged fillings on anterior mandible teeth and complaints of missing several posterior mandible teeth. The patient had a previous panoramic examination (Figure 1), and multiple radiolucent lesions were found incidentally. She had no pain, and clinical features showed no swelling (Figure 2); then, the dentist recommended a CBCT examination with a suspect dental cyst on a posterior mandible dextra.

The radiograph revealed multiple extended cysts in the posterior and anterior mandible dextra with no swelling. The CBCT image showed multiple radiolucencies (Figure 3). The coronal view appears to be a radiolucent lesion on the periapical tooth 46 which extends to the region edentulous tooth 47, unilocular shape, well-define, scallop border, with a size of ± 14.6x 15.6mm (mesial root area) and ± 15.1 x 16.2 mm (distal root area) with density averages 7-32.9 HU and involves the mandibular canal. From the axial view, there appears to be a radiolucent lesion on the periapical tooth 46 which extends to the region edentulous tooth 47, unilocular shape, well-define, scallop border, with size ± 17.7 x 23.7 mm with a mean density of -52.5 HU and involves the mandibular canal. Figure 4 shows that a radiolucent lesion appears on the mesial crown of tooth 46, reaching the depth of the dentin. A radiolucent lesion was found in periapical with well-defined and scallop borders, ± 23.6 x 16.8 mm sizes and density 101.9 grayscale, involving the mandibular canal dextra. Then, from a sagittal view, a radiolucent was found in a 42 43 44 region with well-defined and scallop borders, 13.8 x 16.5 mm sizes and a density of 143 grayscale. Three-dimensional (3D) reconstruction of CBCT of buccal view is shown in Figure 5.

In our cases, the patient has confirmed that there are no complaints in these teeth, painless, no swelling and revealed no systemic diseases or symptoms in the skin and the involved areas. OKC can be unilocular or multilocular, showing a well-defined radiolucent region with smooth and corticated edges on radiological examination. An

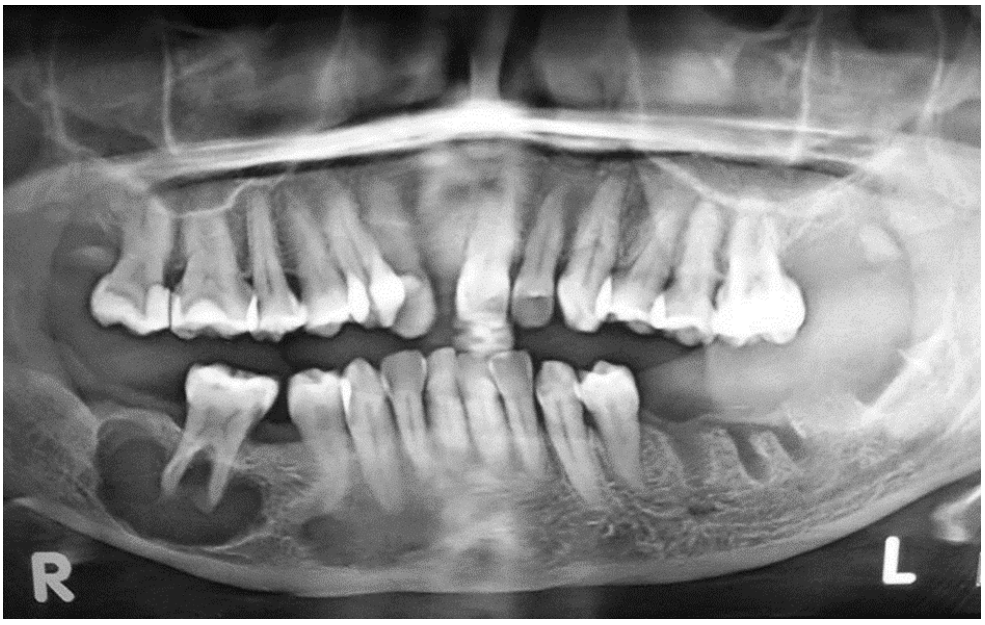

Figure 1. Panoramic radiograph with multiple radiolucent lesions

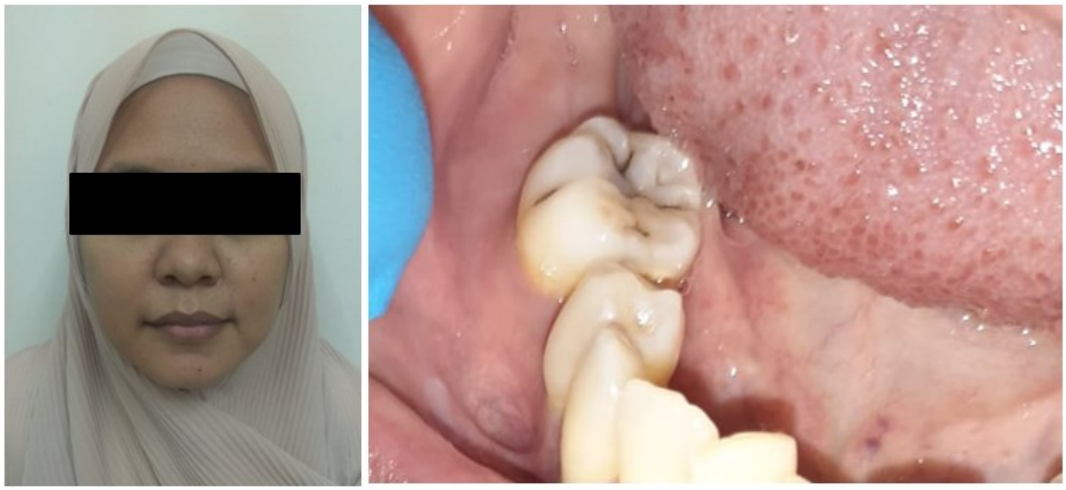

Figure 2. Clinical examination of the patient patient has no evidence of swelling

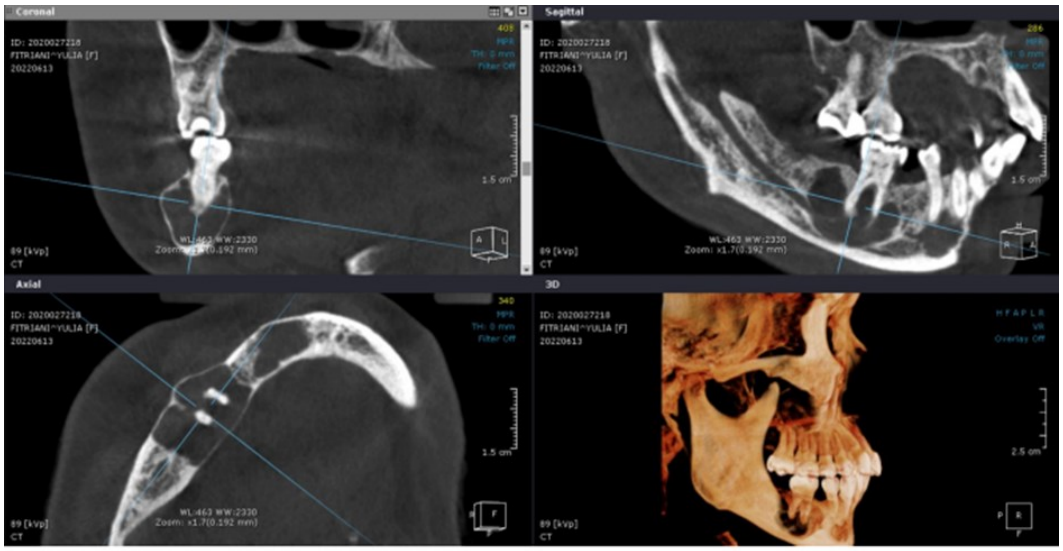

Figure 3. Multiplanar view of the lesion on the right mandible

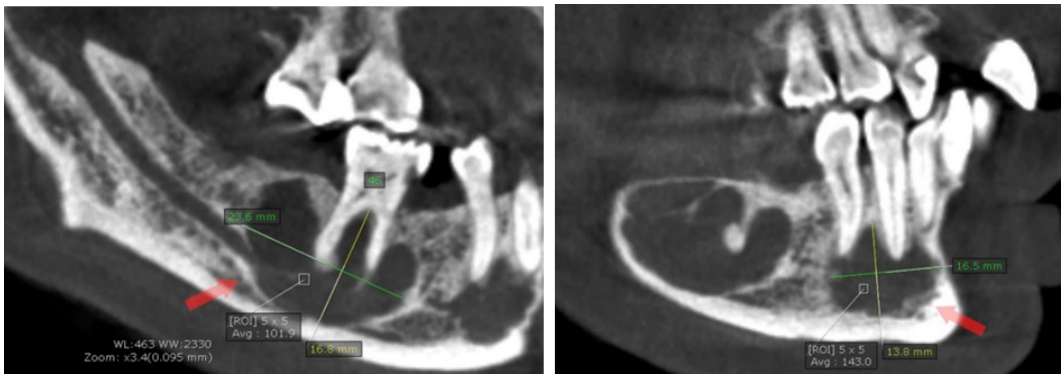

Figure 4. CBCT scan revealing multiple radiolucency on a mandible dextra

unerupted tooth is present in conjunction with the lesion in 25%–40% of patients. Characteristic features under a microscope include a corrugated parakeratinized stratified Squamous epithelial lining with palisaded basal cell layer without rete ridge formation. In this instance, histopathologic slides showed distinctive KCOT characteristics. In this case, we revealed characteristic features of OKC in histopathologic slides. Finally, the diagnosis of OKC was established for all two cystic lesions. Based on the pattern and radiographic characteristics of the radiolucent lesion well-defined with a scallop border, this case was radiodiagnosis as suspected non-syndromic multiple odontogenic keratocysts.

The patient had an odontogenic keratocyst, according to the interpretation of the radiological findings. The patient's clinical evaluation indicated no systemic disease or symptoms in the skin and the affected areas. The histopathologic examination of enucleated tissue was done and confirmed the

diagnosis of odontogenic keratocysts. With a definitive diagnosis of OKC in multiple radiolucent lesions and the absence of any evidence of Gorlin-Goltz syndrome in clinical examinations, these cases were diagnosed as Non-Syndromic Multiple Odontogenic Keratocysts.

DISCUSSION

OKC is a developmental odontogenic cyst originating from the dental lamina's epithelial rest or basal cell extensions from the overlying oral epithelium. OKC is a common developmental odontogenic cyst with biological behaviour similar to a benign neoplasm. Therefore, the latest WHO classification of odontogenic tumors in 2005 has been given the term keratocystic odontogenic tumor. OKC in the mandible-to-maxilla ratio is 2:1, with mandibular and ramus being the most common site. Small OKC is usually asymptomatic, but larger ones may show clinical manifestations like pain, swelling or drainage.[7,8]

Radiographs of OKC commonly present as unilocular cystic lesions with a well-defined border with or without scalloped margins but may also be multilocular. Frequently, they are not easily distinguishable from regular odontogenic cysts. The imaging finding more effective for making a provisional diagnosis of OKC are a well-defined unilocular radiolucent lesion in the posterior jaw and a large radiolucent mandibular lesion with scallop margin and minimal buccolingual expansion. Figure 6 shows the schematic drawing of some OKC possibilities in the mandible and maxilla.[9–11]

Computed tomography is a valuable imaging modality for detecting all dimensions of the OKC with a better definition of anatomical structures such as the maxillary sinus, nasal cavity, inferior alveolar nerve, buccolingual expansion, etc. CBCT can accurately identify cortical perforation (mainly if it is in the angle/ramus of the jaw or the maxillary tuberosity). The overlying oral mucosa is most likely linked to the OKC at the cortical perforation.[9,12]

The most common differential diagnosis of OKC is a dentigerous cyst, ameloblastoma and radicular cyst. OKC associated with an impacted tooth may mimic a dentigerous cyst; when OKC is multilocular and located in the posterior or ramus mandible, it may mimic ameloblastoma; then, when OKC has found in the periapical area, it may be mistaken as a

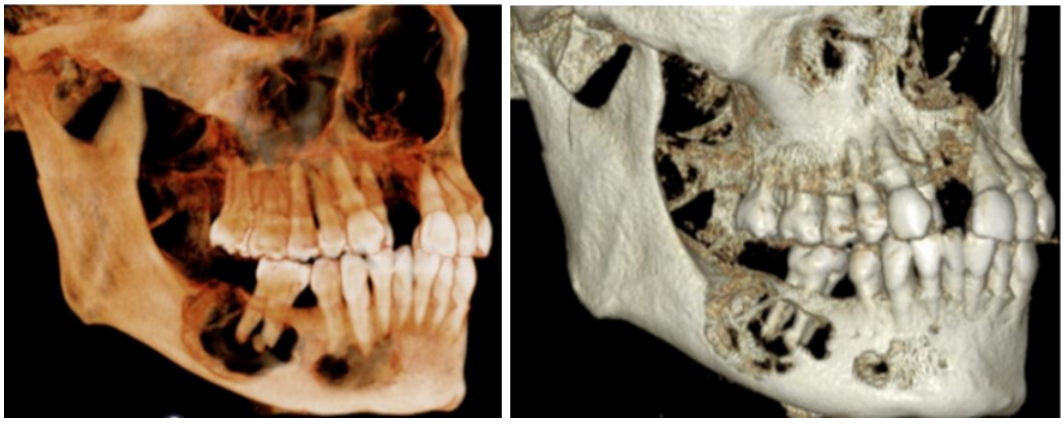

Figure 5. Three-dimensional (3D) reconstruction of CBCT, buccal teeth view (left) and bone view (right)

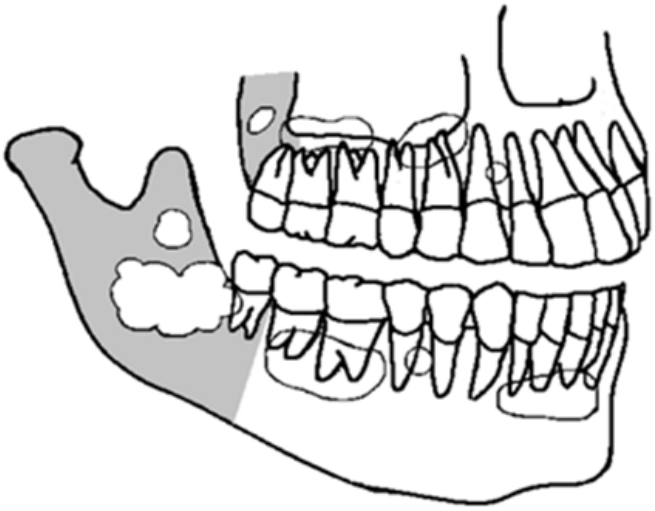

Figure 6. Schematic drawing of some OKC possibilities presentation in the mandible and maxilla. The grey area is the posterior region of the jaw bones[9]

Table 1. Diagnostic Criteria of e Gorlin-Goltz Syndrome or NBCCS

Major Criteria	Minor Criteria
• More than two basal cell carcinomas or one in a patient <20 years old age • (Three or more) palmar or plantar pits • Bilamellar calcification of the falx cerebri • OKC of jaws proven histopathologically • First-degree relative with nevoid basal cell carcinoma syndrome. • Bifid, fused, or markedly splayed ribs	• Macrocephaly (adjusted for height). • Congenital malformations, hypertelorism, cleft lip/palate, and frontal bossing • Skeletal deformities, Sprengel deformity, marked pectus deformity, and syndactyly of digits • Radiological abnormalities, bridging of sella turcica, vertebral anomalies, and modelling defects of hands and feet • Ovarian fibroma or medulloblastoma

radicular cyst.[13]

However, to obtain a definitive diagnosis, a histopathological examination is required. Histopathological slides shows the characteristic features like a corrugated parakeratinized stratified squamous epithelial lining with palisaded basal cell layer without rete ridge formation. In this case, we revealed typical features of OKC in histopathologic slides.[14–16] Finally, a diagnosis of OKC was established for this case.

Multiple OKC is considered major criteria of Nevoid Basal Cell Carcinoma Syndrome (NBCCS) or the Gorlin-Goltz syndrome. There are some diagnostic criteria to make a diagnosis of Gorlin-Goltz syndrome have to be taken (Table 1). The presence of two major and one minor or one major and three minor criteria is necessary to establish the diagnosis. The most important criteria for this syndrome are the presence of pigmented basal cell carcinomas, OKC, palmar and plantar pits, and bilamellar calcifications of the falx cerebri.[2,17,18]

Golgire *et al*. reported a case report to evaluate the clinical aspects, histological features, genetics, and syndromic and nonsyndromic instances of recurrences. They presented a case of a 20-year-old female patient with multiple OKC in both jaws simultaneously without any syndromic manifestation. Syndromic cases were relatively more aggressive, which can be linked to a rise in satellite cysts and mitotic figures with higher PTCH gene expression. In syndromic patients, the recurrence rate was 63%, compared to 37% in nonsyndromic cases.[17] Habibi *et al*.; found that only 8.3% of the 83 cases of keratocysts reported in the Iranian population were syndromic.[19]

Based on the existing literature, it is evident that multiple OKC may be the first or initial symptom of NBCCS or any other illness that can emerge 10 years before other symptoms. As most of these patients manifest in the second decade of life, the chance of further NBCCS symptoms or symptoms from other syndromes developing in the future cannot be completely ruled out. Therefore, a comprehensive clinical evaluation, pertinent investigations, the design of a specific treatment plan for that case, and a strict post-treatment follow-up should be used for a line of overall treatment management.[2,20]

However, in this case report, our patient has only the presence of OKC in the radiographs examination; she was healthy and no symptoms in the skin of the patient's hands and legs in his appearance in the clinical examination, then no suggestive features or criteria of these Gorlin-Goltz syndromes. Multiple OKC without other syndromic manifestations in rare cases. Nevertheless, it should be highlighted that the development of multiple OKC should suggest the existence of a syndrome until proven otherwise. A patient with multiple OKCs should be monitored often to evaluate the potential appearance of any other systemic signs or features of the Gorlin-Goltz syndrome.

CONCLUSION

Radiologically, OKC has the character of a well-defined radiolucent area with smooth and often corticated margins and may be unilocular or multilocular with a scallop border. CBCT has an important role in making a suspected radiodiagnosis of this lesion. However, the combination of clinical examination, radiographs and histopathological examination determines the diagnosis, so this case should be continued with histopathological examination. The radiograph finding with CBCT in this case report led to the diagnosis of non-syndromic multiple odontogenic keratocyst.

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FOOTNOTES

All authors have no potential conflict of interest to declare for this article. Informed consent was obtained from the patient for being included in this case report.

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