Case Report

Gorlin cyst in maxilla: A clinical and histological rarity

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Abstract

Gorlin cyst which is, also, recognised by the synonyms calcifying odontogenic cyst (COC), calcifying ghost cell odontogenic cyst, calcifying cystic odontogenic tumor (CCOT) and dentogenic ghost cell tumor, is a rare developmental lesion was first reported by Gorlin *et al.* in 1962. It was, later, renamed as calcifying cystic odontogenic tumor (CCOT) in the WHO classification devised in 2005 due to its histological complexity, morphological diversity and aggressive proliferation. Gorlin cyst was, later, re-named several times including Gorlin cyst, calcifying odontogenic cyst (COC), calcifying ghost cell odontogenic cyst, calcifying cystic odontogenic tumor (CCOT) and dentogenic ghost cell tumor. The present case report briefs a case of Gorlin cyst in a 21 year old female which was diagnosed during the diagnostic work-up.

Keywords: Calcifying cystic odontogenic tumor and dentogenic ghost cell tumor, calcifying ghost cell odontogenic cyst, calcifying odontogenic cyst, gorlin cyst

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INTRODUCTION

Gorlin cyst which is, also, recognised by the synonyms calcifying odontogenic cyst (COC), calcifying ghost cell odontogenic cyst, calcifying cystic odontogenic tumor (CCOT) and dentogenic ghost cell tumor, is a rare developmental lesion that arises from the odontogenic epithelium[1] and representing about 2% of all odontogenic pathologies seen in the jaws.[2] Gorlin cyst is clinically characterized as a painless, slow growing lesion which does not have a predilection for either maxilla or, mandible though has a predilection for anterior region of the jaws and usually arises intra-osseously, though, may have an extra-osseous origin, too. It has a peak incidence during the second and third decades of life with a mean age of incidence of 30.3 years with no gender predilection.[3-6] Radiographically, Gorlin cyst may appear as a unilocular or, multilocular radiolucent lesion with either well-circumscribed or, poorly-defined margins and may, also, be observed in association with unerupted teeth. Calcification is an important radiographic sign for the diagnosis of Gorlin cyst and is detected in approximately half of the reported cases.[7-10] The typical histopathological features of Gorlin cyst include a fibrous wall and a lining of odontogenic epithelium composed of cells resembling ameloblasts. Stellate reticulum-like cells overlay the basal cell layer while ghost cells, which are consistently seen, may occasionally, show signs of calcification.[11,12] The treatment of choice for Gorlin cyst is conservative and wide base surgical enucleation usually suffices with the treatment. However, recurrence is frequent seen, especially, in neoplastic cases like dentogenic ghost cell tumors.[13]

CASE REPORT

A 21-year-old female patient reported to the Outpatient Department (OPD) with a chief complaint of swelling in

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upper front tooth region since 6 months with a history of discomfort due to the swelling although no associated pain was reported. On extra-oral examination, gross asymmetry over left middle third of face [Figures 1 and 2] was evident, 3 × 3cms in size, hard in consistency and non-tender on palpation. On intra-oral examination, tooth # 63 was found retained while tooth # 23 was missing. Vestibular obliteration was noted in relation to teeth # 22,23,24,25 region was noted with egg shell crackling on palpation [Figure 3]. FNAC was positive yielding 3.8ml of straw colored fluid [Figure 4]. Intra-oral periapical radiograph (IOPAR) [Figure 5] and maxillary left lateral occlusal radiograph [Figure 6] revealed a well-defined periapical radiolucency in relation to teeth # 22,23,24,25 region with root resorption present in relation to teeth # 63, 24 and 25. Orthopantomograph (OPG) revealed a well-defined radiolucency in the same region with impacted tooth # 23 and root resorption in relation to teeth # 63, 24 and 25 [Figure 7]. Based on the patient's history and clinical and radiographic examination, a working diagnosis of dentigerous cyst in relation to tooth # 23 was given with the list of differential diagnoses including adenomatoid odontogenic tumor (AOT), unicystic ameloblastoma and Gorlin cyst. Complete surgical enucleation of cyst was done and the specimen was submitted for histopathological examination which revealed a cystic epithelium of variable thickness along with the presence of ameloblast-like cells, stellate reticulum-like cells and ghost cells with signs of calcification along with numerous bundles of collagen fibers with sub-epithelial hyalinization confirming to a histopathological diagnosis of Gorlin cyst [Figure 8].

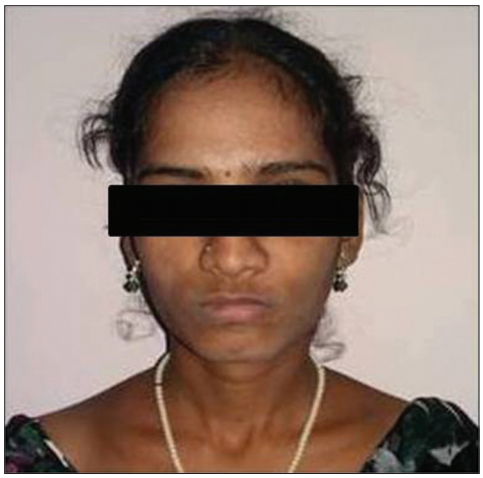

Figure 1: Frontal profile of the patient revealing gross asymmetry over left middle third of face

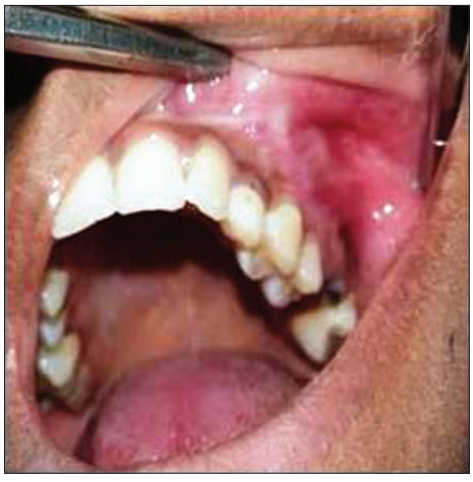

Figure 3: Vestibular obliteration in relation to teeth # 22, 23, 24, 25 region

DISCUSSION

Gorlin cyst which is, also, recognised by the synonyms calcifying odontogenic cyst (COC), calcifying cystic odontogenic tumor (CCOT), calcifying ghost cell odontogenic cyst and dentogenic ghost cell tumor, was first reported by Gorlin *et al.* in 1962.[2] At that time, it was classified as a cyst-related to the odontogenic apparatus. The lesion has some features which resemble a cyst while some features which are characteristic of a solid neoplasm making the lesion a little different from the rest of the lesions which more or, less have a clear classification pattern. Although it has commonly been recognized as a benign odontogenic cyst since its original description by Gorlin *et al.* in 1962,[2] this pathologic

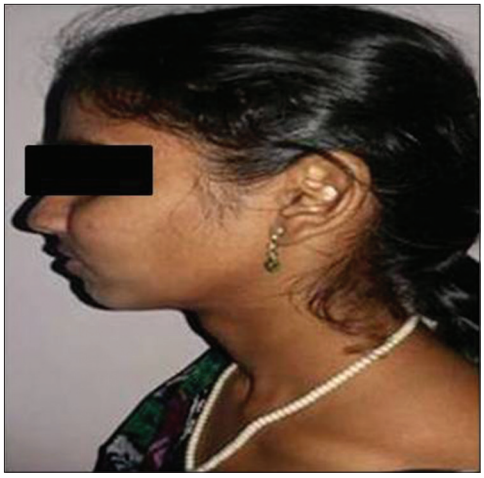

Figure 2: Lateral profile of patient revealing gross asymmetry over left middle third of face

Figure 4: A positive fine needle aspiration cytology with 3.8ml of straw colored fluid aspirated

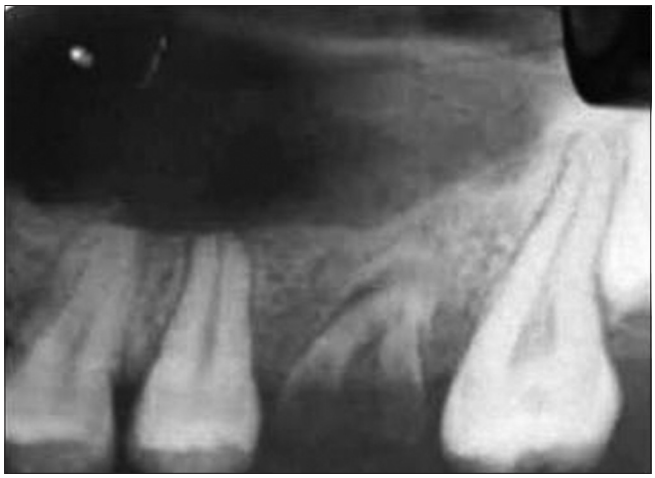

Figure 5: Intra-oral periapical radiograph revealing a well-defined periapical radiolucency with root resorption in relation to teeth # 63, 24 and 25

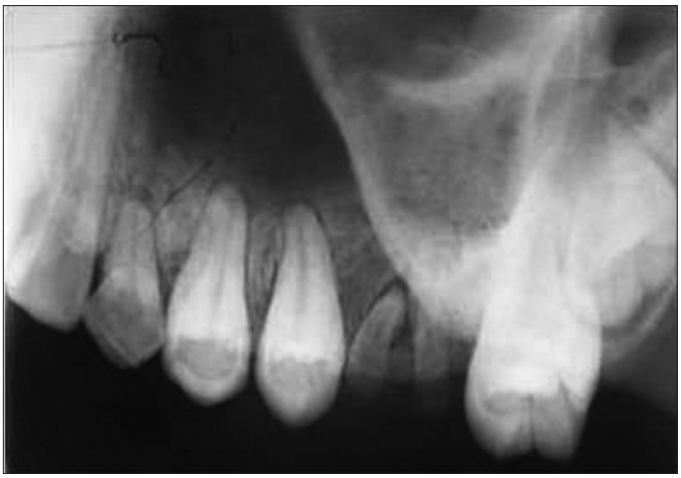

Figure 6: Maxillary left lateral occlusal radiograph revealing a well-defined periapical radiolucency extending from teeth # 22 to 26 region with root resorption in relation to teeth # 63, 24 and 25

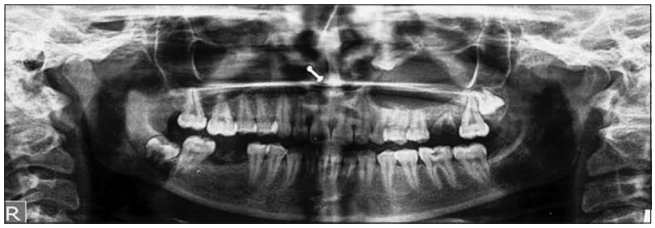

Figure 7: Orthopantomograph revealing a well-defined radiolucency extending from teeth # 22 to 26 region with impacted tooth # 23 and root resorption in relation to teeth # 63, 24 and 25

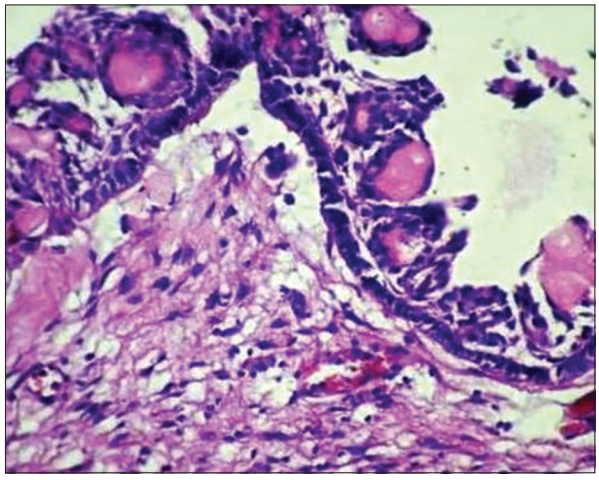

Figure 8: Histopathological examination revealing cystic epithelium of variable thickness along with the presence of ameloblast-like cells, stellate reticulum-like cells and ghost cells with signs of calcification along with numerous bundles of collagen fibers with sub-epithelial hyalinization

entity encompasses a spectrum of clinical behaviours and histopathological features ranging from cystic to neoplastic to characteristically aggressive variants. This concept, called "monistic" by Toida has led some researchers to substitute the term Gorlin cyst with calcifying odontogenic cyst (COC), calcifying ghost cell odontogenic cyst, calcifying cystic odontogenic tumor (CCOT) and dentogenic ghost cell tumor in the later revisions. In addition, a "dualistic" approach has been suggested that Gorlin cyst encompasses two distinct entities as a cyst, calcifying odontogenic cyst (COC), calcifying ghost cell odontogenic cyst as well as a tumor or, neoplasm which can be benign or, malignant and/or, a combined lesion with each of the categories described above being associated with odontoma, ameloblastoma and/or, other odontogenic lesions.[14] As a result of this diversity, different classification schemes and nomenclatures for the lesion have been suggested. Gorlin cyst was renamed as calcifying cystic odontogenic tumor (CCOT) in the WHO classification for Head and Neck Tumors devised in 2005 due to its morphological diversity, histological complexity and an aggressive clinical behavior.[15] The lesion manifests either as a central or, a peripheral variant with the central variant being more common in clinical expression.

Praetorius *et al.* suggested bimodal age distribution for the lesion[15] with new cases expressed over a wide age range from 1 year to 82 years of life while second decade of life being the most common for the occurrence of newly reported lesions. There is negligible difference in gender predilection with no characteristic racial predilection reported either. Furthermore, both the jaws are involved with almost an equal frequency.[7-10] Li and Y reported maxilla to be more frequently affected than mandible while the most common site of occurrence for the lesion was canine-premolar region of maxilla. Also, in mandible, several cases cross midline, a finding, less frequently seen in maxilla.[16] The demographic data recorded in the present case was in accordance with the reported literature as it affected anterior maxilla in a female patient who was in

her second decade. Most of the peripheral lesions have been reported in the maxillary or, mandibular gingiva and/or, alveolar mucosa anterior to the first molar[6] while the central lesions are seen in the form of asymptomatic swellings as was seen in the present case producing a hard bony expansion, however, fairly extensive palatal/lingual expansions might, also, sometimes, be observed in a few of the cases.[3-6] Occasionally, Gorlin cyst may perforate the cortical plates as was noted in the present case and extend into the peripheral soft tissues.[16] Sometimes, the cystic lesion may, also, displace the adjoining teeth and/or, lead to extensive root resorption, another peculiar feature noted in the present case, with extensive root resorption present in relation to teeth # 63, 24 and 25 as was revealed on radiographic investigation. Occasionally, few cysts might be seen as completely asymptomatic lesions discovered co-incidentally during the routine radiographic examinations.[16] Extra-osseous lesions tend to be pink to red in appearance depending on their location within the tissues, well or, poorly-circumscribed elevated masses measuring up to 4cms in diameter and are considered as one of the important differentials for gingival lesions.[6] Because these lesions arise in tooth-bearing regions of the jaws, radiographically, they may be seen as destructive unilocular or, multilocular radiolucencies with calcifications of variable density reported in approximately one third to one half of the cases reported. Root resorption and divergence of the adjacent teeth are the common radiological findings while association with an impacted tooth is seen in approximately one third of the cases reported[1] as was seen in the present case. However, the present case did not show evidence of calcification within the radiolucent lesion with the conventional radiography done. As defined in the WHO classification for Head and Neck Tumors devised in 2005, Gorlin cyst is a cystic lesion in which the epithelial lining shows a well-defined basal cell layer of cuboidal or, columnar cells with stellate reticulum-like cells overlaying the basal cell layer while ghost cells seen occasionally in the cystic lining or, within the fibrous capsule of the cyst. The ghost cells might, also, become calcified occasionally while a layer of dysplastic dentin may be laid down adjacent to the basal cell layer of the epithelium. The epithelial lining of Gorlin cyst appears to have the ability to induce formation of dental hard tissues in the adjacent connective tissue wall.[15] The present case, also, revealed a cystic epithelium of variable thickness along with the presence of ameloblast-like cells, stellate reticulum-like cells and ghost cells with signs of calcification along with numerous bundles of collagen fibers with sub-epithelial hyalinization confirming to a histopathological diagnosis of Gorlin cyst. The ghost cells seen in the cystic lining or, within the fibrous capsule of the cyst represent an abnormal type of keratinization and have an affinity for calcification as was evident in the present case where the histopathological examination revealed ghost cells with signs of calcification along with numerous bundles of collagen fibers with characteristic sub-epithelial hyalinization.[11,12] The epithelium may be regular 6-8 cell-layers thick over part of its length and be continuous with parts that may be very thin and others that are considerably thickened. Budding from the basal cell layer into the adjacent connective tissue and epithelial proliferations into the lumen are, also, frequently reported. Melanin deposits are, also, sometimes present in the epithelial lining.[13,17] Enucleation is the choice of treatment for most intra-osseous Gorlin cysts similar to the treatment provided in the present case, however, few recurrences have been reported.[18,19]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

CONCLUSION

A thorough clinical, radiological and histopathological evaluation is mandatory for the diagnosis of this rare but significant jaw lesion of intra-osseous origin. An extensive and meticulous follow-up protocol is, also, equally significant to rule-out any evidence of recurrences as well as chances of malignant transformation.

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Conflicts of interest

There are no conflicts of interest.

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