Case Report

Ameloblastomatous calcifying cystic odontogenic tumour involving the mandibular anterior region: An unusual case report

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Abstract

Ameloblastomatous calcifying odontogenic cyst (COC) is a very rare histopathological variant of COC, which is an odontogenic cyst of the jaw. The term "calcifying odontogenic cyst" is not covered in the World Health Organisation (WHO) Classification of Tumors 2005 and was re-named as calcifying cystic odontogenic tumour (CCOT). There are only a few reports containing details on CCOT being associated with ameloblastoma. This variant has been classified as ameloblastomatous CCOT (type 3) as per the WHO 2005 classification. In this article, we reported an exceptional case of ameloblastomatous CCOT in a 15-year-old boy involving the mandibular anterior region, which is a rare combination for age and site of the lesion, along with an impacted tooth which is again an uncommon association.

Keywords: Ameloblastoma, ameloblastomatous calcifying odontogenic cyst, CCOT

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INTRODUCTION

In 1971, the World Health Organisation (WHO) had published the first classification of odontogenic cysts and tumours, which set the criteria for designation and a nomenclature, which is internationally accepted, named as "Histological typing of odontogenic tumors and jaw cysts and allied lesions".[1] But with time, new variants were found. Gorlin was the first who described the entity called calcifying odontogenic cyst (COC) in 1962.[2] There is a known association between COC and various other odontogenic tumours, due to which various classifications of COC were proposed.[3,4] The majorities of COCs are cystic, but some are solid and a few are neoplastic in nature. In the 2005 WHO publication on odontogenic tumours, it was classified as a benign odontogenic tumour having odontogenic epithelium with odontogenic ectomesenchyme with or without dental hard tissue formation and was renamed as calcifying cystic odontogenic tumour (CCOT). However, in 2017, it was reclassified in the group of odontogenic cysts.[5,6] When a lesion is associated with ameloblastoma, there is always a question regarding the nature, behaviour and prognosis. Hence, knowledge of this rare entity becomes mandatory for general clinicians in order to aid in early diagnosis. The

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purpose of this article was to add one more rare case of ameloblastomatous CCOT to the existing literature, with a brief discussion on its clinical, radiological and histological features.

CASE REPORT

A 15-year-old male patient reported to the department of oral medicine and radiology with the chief complaint of swelling in his lower left front teeth region of the jaw for the past one year. The swelling was initially smaller and had increased in size over the period of one year to the present size. There was absence of pain or any other associated symptoms. Past medical history was non-contributory. Past dental history revealed that a root canal treatment had been done in the lower front teeth one year back. On extraoral examination, single diffuse swelling was present on the lower one-third of the face, extending antero-posteriorly from the mandibular symphysis region to two centimetres in front of the angle of the mandible, and supero-inferiorly from the level of the lower lip to the lower border of the mandible. Overlying skin appeared normal in colour but stretched [Figure 1]. On palpation, the swelling was afebrile, non-tender and firm-to-hard in consistency. Intraoral examination revealed swelling in the lower left buccal vestibule region extending medio-laterally from the mandibular right permanent central incisor to the left side second premolar teeth (41 to 35) region, and supero-inferiorly from the free gingival margin to the buccal vestibule, thus obliterating the vestibule in that region. Also, the swelling was present on lingual aspect extending medio-laterally from the mandibular left permanent central incisor to the left second premolar teeth (31 to 35) region, and supero-inferiorly from the free gingival margin to the floor of the mouth [Figure 2]. The buccal and lingual cortical plates were expanded on the left side. The swelling was non-tender and soft in consistency. There was a presence of over-retained deciduous left mandibular canine (73) which was mobile, and there was clinically missing mandibular left permanent canine (33). Also, the mandibular left first premolar (34) was lingually tilted. On the basis of history and clinical examination, a provisional diagnosis of dentigerous cyst with 33 was given. The patient was then subjected to panoramic radiography. The panoramic radiograph showed a well-defined, multilocular radiolucency extending from the mesial aspect of the root of the mandibular right permanent canine (43) to the distal aspect of the root of the mandibular left second premolar (35), thus crossing the midline, with loss of cortication at some places along with horizontally impacted mandibular left permanent canine (33), which was pushed to the inferior border of the mandible. Few radiopaque septa were visible near the root apices of the left mandibular premolars (54, 35). Also, well-defined, linear radiopacities were seen occupying the root canal spaces of all of the permanent mandibular incisors (31, 32, 41, 42), right permanent canine (43), and left first and second premolars (34,35), suggestive of root canal treatment done. Over-obturation was seen with the left second premolar (35), and distal displacement of roots was seen with 41, 42, 31, 32, 34, 35, which indicated the space-occupying nature of the lesion with its epicentre between 32 and 34. Also, root resorption was seen with 35 [Figure 3]. Thus, a working diagnosis of infected dentigerous cyst with mandibular left permanent canine (33) was given, and the patient was subjected to

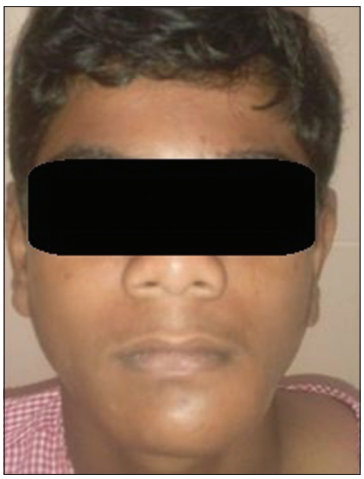

Figure 1: Extraoral clinical picture showing diffuse swelling in mandibular symphysis and left parasymphysis region

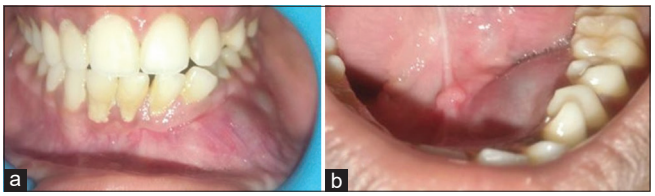

Figure 2: Intraoral clinical picture showing evident swelling in a) lower labial vestibule extending laterally towards buccal vestibule region; and b) lingual sulcus region on left side of mandible

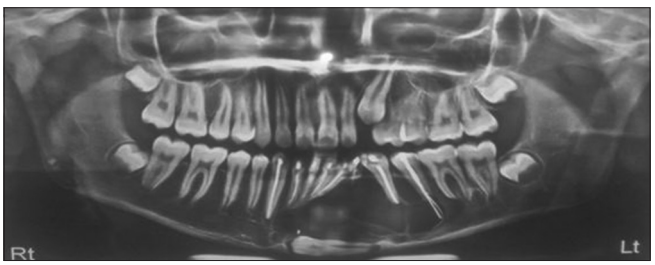

Figure 3: Orthopantomogram showing a well-defined, multilocular radiolucency extending from 43 to 35 region along with horizontally impacted 33

incisional biopsy. The specimen thus collected was sent for histopathological examination.

On haematoxylin and eosin (H&E) staining of the section, cystic lining composed of thin, stratified squamous epithelium showing characteristic odontogenic features of prominent cuboidal basal cell layer and presence of columnar cells at places with palisaded nuclei were seen. At places, the epithelium showed nuclear hyperchromatism. The epithelium also showed groups of ghost cells. The basal layer showed budding of strands and follicles of ameloblastomatous epithelium [Figure 4a]. Also, a thicker area of epithelium showing groups of ghost cells were seen [Figure 4b and c]. Based on these features, the final diagnosis of ameloblastomatous CCOT was confirmed.

DISCUSSION

CCOT is a rare pathology which accounts for only 1%–2% of jaw cysts, as reported in literature.[7] The classification by the WHO in 1971 described COC as a "non-neoplastic cystic lesion".[1] In the 1992 edition of World Health Organization International Classification of Tumors, the authors replaced this phrase with "most lesions appear to be non-neoplastic". The 1992 WHO classification cited the terms "dentinogenic ghost cell tumour" and "odontogenic ghost cell tumour," especially for the solid lesion.[8] Although, in 2005, WHO had classified COC in the group of odontogenic tumours, it was reclassified in the group of odontogenic cysts in 2017.[5] The lesion is often associated with tumours of odontogenic origin such as odontoma, ameloblastoma, ameloblastic fibroma, ameloblastic fibro-odontoma and adenomatoid odontogenic tumour.[6] According to Hong *et al*.,[9] it has the following four variants: type 1, which includes simple cysts named as calcifying odontogenic cysts; type 2, which consists of cysts associated with odontogenic hamartomas or benign neoplasms; type 3, which consists of ameloblastomatous COC characterised by ameloblastoma-like cystic lining epithelium with ghost cells or calcification; and type 4, which includes COC associated with odontoma. The histological findings in the present case were in accordance with type 3 of the above-mentioned classification.

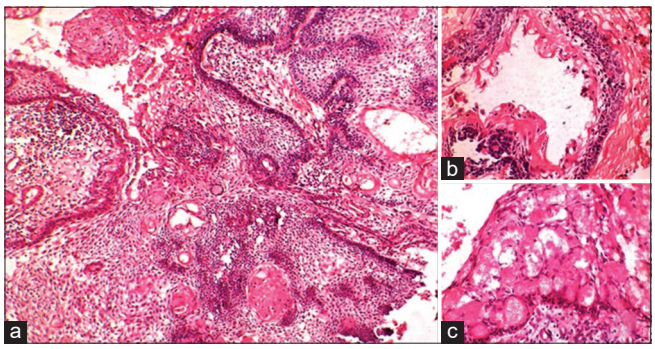

Figure 4: Histopathologic picture showing cystic lining composed of thin stratified squamous epithelium depicting characteristic odontogenic features and the basal layer showing budding of strands and follicles of ameloblastomatous epithelium under 10× (a). Thicker area of epithelium showing groups of ghost cells under 40× (b) and 100× (c) magnifications

As per the literature, only 32 cases of CCOT associated with ameloblastoma have been reported, with very few reports having details about the clinical and radiographic features.[6,7,10] Association of this hybrid lesion with impacted canine in the present case makes it unique.

According to a recent review published in 2018,[6] the age of occurrence ranges from 15 to 65 years, with the mean age being 34.09 years. In the present case, the age of the patient was 15 years. Female dominance has been found in the existing literature, but in the present case, the patient was male. Mandible is affected more frequently than the maxilla, with posterior body and ramus region being the most common site, whereas in the present case, the lesion was present in the mandibular anterior region crossing the midline, which is rarely seen. Clinical presentation varies from being asymptomatic to painful swelling with or without bony expansion. In the present case, the patient had painless swelling with bony expansion of both the buccal and lingual cortical plates. In addition to this, there was absence of permanent tooth in that region due to which the provisional diagnosis of dentigerous cyst was given.

Radiographically, the lesion appears as a well-defined, unilocular or multilocular, often quite large radiolucency that may or may not contain varying amounts of radiopaque material associated with unerupted tooth. Root resorption or root divergence may also be observed.[10] In the present case, a large multilocular radiolucency was seen with interrupted borders associated with horizontally impacted canine at the lower border of the mandible. Also root divergence was seen in all of the teeth associated with the lesion (31, 32, 34, 35, 41, 42). Thus, the radiographic presentation in the present case was suggestive of dentigerous cyst; however, after histopathologic examination, the final diagnosis turned out to be CCOT with ameloblastoma. Fifteen-month follow up showed no recurrence for the lesion in the present case. The presence of calcification, which is seen in about 50% of cases, is the most important characteristic feature for the diagnosis of the COC. The differentiation of this variant of COC from the ameloblastoma arising in COC (Ameloblastoma ex COC) is important since the former requires conservative management while the latter needs aggressive treatment.[7]

Most probable differential diagnosis for this lesion includes dentigerous cyst, ameloblastoma, odontogenic keratocyst, and COC.[6]

Histologically, ameloblastomatous COC resembles unicystic ameloblastoma except for the presence of ghost cells and calcifications within the epithelium, which may be present and the fact that it occurs only intraosseously, as was seen in the present case. In some cases, individual ghost cells may fuse together to form large, amorphous, eosinophilic structures on which calcification may occur and irregular masses of calcified structures suggestive of dysplastic dentin are also present in association with basal cell layer.[6,7,10]

CONCLUSION

CCOT with ameloblastoma is a hybrid odontogenic tumour comprising of two distinct lesions which is extremely rare. Preoperative diagnosis in these cases is usually difficult or sometimes impossible because it has significant clinical and radiographic similarities with odontogenic cysts and tumours. Thus, in such cases, histopathology plays a vital role. Hence the role of histopathology should never be ignored and every cyst and tumour should be sent for histopathologic examination to confirm the diagnosis. CCOT with ameloblastoma should be differentiated from true ameloblastoma arising from COC since the management differs in both cases.

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.

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Conflicts of interest

There are no conflicts of interest.

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