

**UNILOCULAR RADIOLUCENCY OF MANDIBULAR ANGLE  
REGION –A clinicopathological Conference**

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**ABSTRACT**

Calcifying Cystic Odontogenic Tumor (CCOT) constitutes 1% of all jaw cysts and considered as one of the rarest odontogenic lesions. It's a group of odontogenic entities which exhibits variety of clinical and biological behaviors. Even after many classifications and sub classifications COC remains a tough nut to crack for clinicians and pathologists. According to recent WHO classification of 2005 it has been reclassified as a tumor due to its diverse nature. Ameloblastomatous Proliferating type is very rare variant of CCOT which is not widely described in literature. This Clinicopathological conference presents one such case in young female patient and also discuss in detail about the differential diagnosis and management of such cases.

**Keywords:** Ameloblastomatous CCOT, Clinicopathological Conference, Mandibular Angle Region, Unilocular Radiolucency

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**INTRODUCTION**

A 16 year old female came to the Dept. of Oral & Maxillofacial Surgery with complain of swelling on left side of the face since last 3months (Fig. 1). Swelling was small peanut in size when patient first noticed, it gradually increased to attain the present size. Patient also had history of dull, intermittent type of pain since 1month. Facial disfigurement was also present because of the swelling. Swelling was not associated with any discharge. Patient did not have

any significant past medical or dental history. Clinical examination revealed asymmetry of the face due to swelling of lower 1/3<sup>rd</sup> of the face on left side. Extraoral swelling was hard in consistency without any associated fever or pus discharge. No history of paresthesia was present. Intraoral examination revealed presence of all teeth except 37. Swelling was noticed in lower left mandible region behind 36 tooth. Lesion seemed to be intraosseous causing expansion of the buccal cortical plate. Swelling was non tender on palpation & overlying mucosa appeared normal.

A panoramic radiograph revealed unilocular radiolucency extending from left mandibular 1<sup>st</sup> molar to left subcondylar and coronoid region (Fig.2). It also revealed the presence of crown of 37 tooth which was displaced to the lower border of the mandible. Lesion also caused thinning and scalloping of the lower border of the mandible. Lesion was extending upto 36 tooth but it did not cause any displacement or resorption of that tooth. Lateral oblique view also helped to further confirm the findings of panoramic view (Fig. 3).

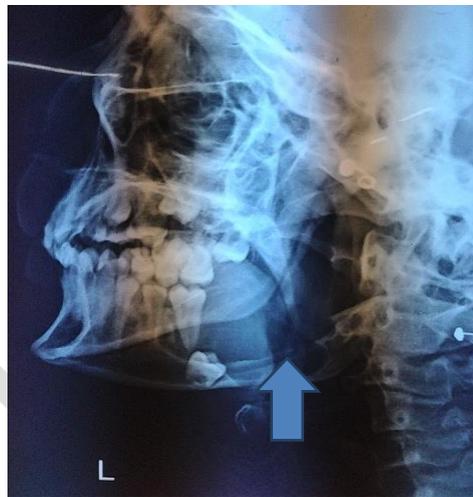
Cone Beam Computed Tomography (CBCT) revealed presence of unilocular radiolucency extending from 1<sup>st</sup> molar to posterior border of ramus anteroposteriorly and sigmoid notch to inferior border of mandible superoinferiorly. Dimensions of the lesion were 30×48×18. Axial & Coronal sections also showed marked expansion of buccal and lingual cortical plates. (Fig.4.) Nonconclusive radiographic picture prompted incisional biopsy.



*Fig. 1. Frontal profile picture of patient. Note swelling of lower 1/3<sup>rd</sup> of left side face*



*Fig. 2. Massive, unilocular, radiolucency in left mandibular angle region involving displaced 37 tooth*



*Fig. 3. Lateral oblique radiograph further help to locate position of displaced 37 crown*

A large, unilocular radiolucency of mandibular angle region with displaced tooth suggested an aggressive intraosseous lesion. But it's well demarcated border, confined unilocular nature, cortical expansion with its perforation, no associated paresthesia ruled out any malignant lesion and suggested the presence of benign odontogenic or nonodontogenic cyst or tumor. Considering the clinical and radiographical findings our provisional diagnosis was dentigerous cyst. Dentigerous cysts are the most common jaw cysts after radicular cysts and most commonly associated with an unerupted or impacted tooth. They form around 20% of all jaw cysts and 2.6% of unerupted teeth are associated with dentigerous cysts. The fact that this cyst can lead to expansion of bone with subsequent facial asymmetry, extreme displacement of the associated unerupted tooth to lower border of the mandible and 'hollowing-out' of ramus made us to think this lesion as dentigerous cyst.<sup>1</sup>

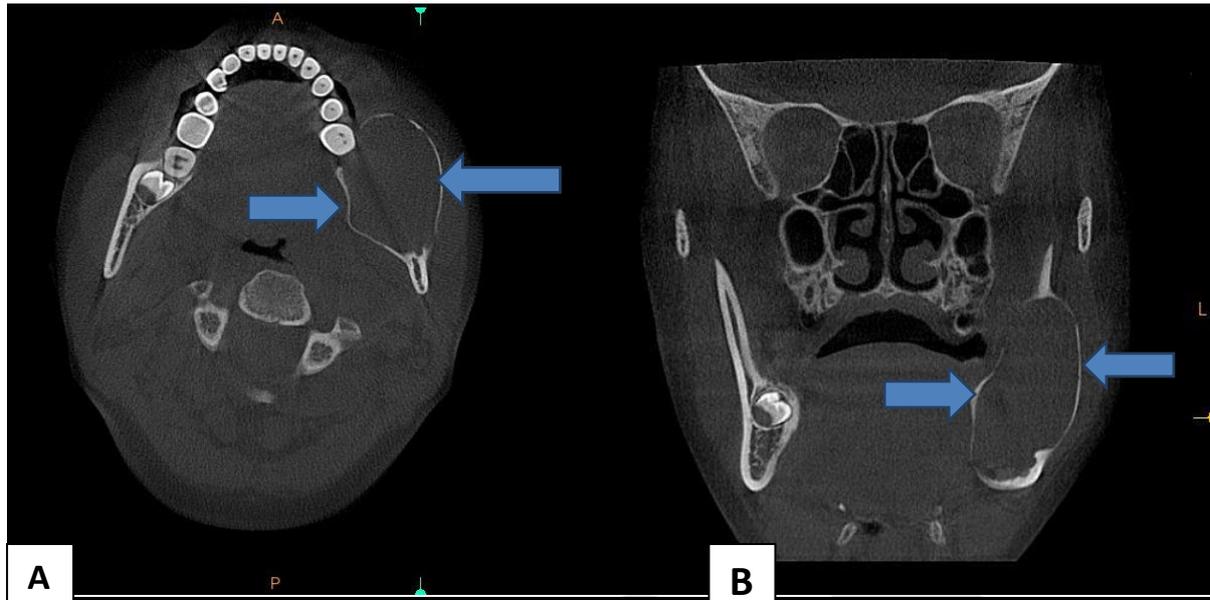
The other lesion that may appear in this region is odontogenic keratocyst known as keratocystic odontogenic tumor as per recent WHO classification. KCOT most commonly occur in 2<sup>nd</sup> and 3<sup>rd</sup> decades of life with 65 to 78% of cases occurring in posterior

mandible.<sup>1,2</sup> Radiographically most KCOTs are unilocular, presenting with well-defined peripheral rim. But KCOTs are less likely to be associated with expansion or perforation of the cortical plates because their propensity to grow along the internal aspect of the jaws. Apart from that KCOTs are less likely to cause displacement of teeth making it a less likely cause in the present case. The odontogenic tumors that can present with such manifestations include unicystic Ameloblastoma, calcifying epithelial odontogenic tumor, adenomatoid odontogenic tumor, calcifying cystic odontogenic tumor. Tumors like odontoma, odontoameloblastoma or ameloblastic fibrodontoma were easily excluded because of the presence of radiopacities in these lesions.

Ameloblastoma is the second most common odontogenic tumor after odontoma forming around 11% of all odontogenic tumors. 3/4<sup>th</sup> of the cases occur in posterior mandible region. Ameloblastomas also cause displacement of the tooth. But this tumor is more common in 20 to 50 years age group and has male predilection making it a less possible cause for the present case. Apart from that such a huge ameloblastic lesions may be associated with paresthesia of the nerve which is not present in this case. Ameloblastomas are most commonly multilocular entities only a 6% of the cases have complete unicystic, unilocular appearance.<sup>1</sup> Calcifying epithelial odontogenic tumor (CEOT) also known as Pindborg tumor is another possible diagnosis of this case. CEOTs are also most common in posterior mandibular area and occur with no gender predilections. This tumor is also associated with severe displacement of the teeth and cortical expansion. The tumor can be unilocular though multilocular cases also having been reported. It has been reported to occur between 8-92 years with the mean age of occurrence is 40 years.<sup>1,2</sup>

Calcifying cystic odontogenic tumor (CCOT) is rather a rare entity comprising of 2% of all odontogenic tumors.<sup>5</sup> These tumors are most common in second decade of life with same number of cases reported for maxilla and mandible.<sup>6</sup> The tumor is more common in canine-premolar region. But it's Ameloblastoma proliferating type variant (Type-3) which is very rare is known to occur in mandibular molar region.<sup>13</sup> CCOT are generally a unilocular entity with only 5-13% of cases are multilocular.<sup>1</sup> Clinically it is most commonly associated with a tooth which is displaced because of the tumor. Adenomatoid odontogenic tumor (AOT) is considered a rare entity. It has strong female predilection with 64% of reported cases occurring in females with 74% of cases occurring in patients below 20 years of age making it a candidate for the potential differential diagnosis of the present case. But 65% of the reported cases occurred in maxilla and 76% of the cases occurred in anterior part of the jaws making it a less likely diagnosis for this case.<sup>1</sup>

Other rare lesions which can come in the differential diagnosis of the present case include ameloblastic fibroma, central odontogenic fibroma and odontogenic myxoma. Among these ameloblastic fibroma is usually a unilocular lesion occurs in 2<sup>nd</sup> decade of life with mandible predilection in molar areas. Odontogenic myxomas and central odontogenic fibromas also occur in 2<sup>nd</sup> and 3<sup>rd</sup> decades of life with female predilection but they are mixed radiolucent-radiopaque entities when presented as a huge lesion.



*Fig.4. Axial (4-A) and coronal (4-B) section images of cone beam computed tomography showing expansion of the buccal & lingual cortical plates of the right posterior mandible. Unilocular radiolucency extending from 1<sup>st</sup> molar to posterior border of mandible anteroposteriorly and sigmoid notch to inferior border of mandible superoinferiorly.*

To differentiate between cystic or solid lesion first aspiration cytology was carried out. 3ml of clear, straw colored fluid was aspirated & sent for cytology, report of which showed the presence of abundant cholesterol crystals with cellular debris just pointing out the presence of an inflammatory cyst. This confirmed the presence of an odontogenic cyst. To get the final diagnosis incisional biopsy was carried out. A window was made posterior to 36 tooth region & part of the lesion was excised & sent for the histopathological examination. Hematoxylline & Eosin staining of incised specimen showed cystic lining comprising of stratified epithelium with cuboidal basal cells and stellate reticulum like superficial cells. Squamous metaplasia was seen in few areas. Many areas showed presence of ghost cells & some showed presence of juxta epithelial dentinoid like material formation. Stroma showed thick collagen with hyalinization in few areas. Odontogenic follicles with cystic degeneration were also seen (Fig. 6). Final diagnosis of calcifying cystic odontogenic tumor of Ameloblastoma proliferating type (type-3) was made on the basis of histopathological picture.

Following the diagnosis complete enucleation of the lesion under general anesthesia was done. At the time of surgery total perforation of buccal bone was noticed. Only a thin bony rim was present over the angle region. Lesion was removed in toto preserving the inferior alveolar nerve canal and closure was done in single layer. (Fig. 5). Regular monthly follow-up was done. No paresthesia was present post-operatively. After 18 months follow-up there were no signs of recurrence.



Fig. 5-A



Fig. 5-B

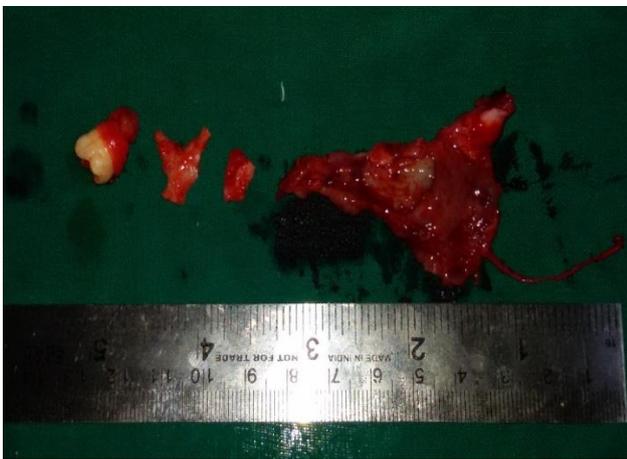


Fig. 5-C

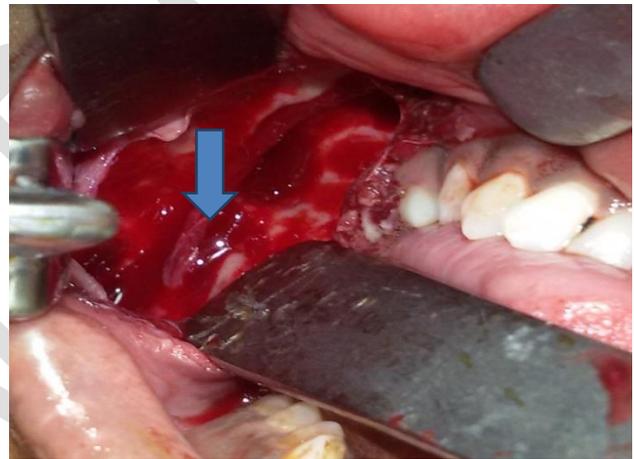


Fig. 5-D

*Fig. 5 A- Exposure of the lesion. B- Tumor has caused complete perforation of buccal bone on mandibular angle region leaving only a thin rim of cortical bone. C- Excised specimen which looked like a cystic lesion in toto along with the associated 37 tooth. D- Locating inferior alveolar canal & preservation of inferior alveolar nerve.*

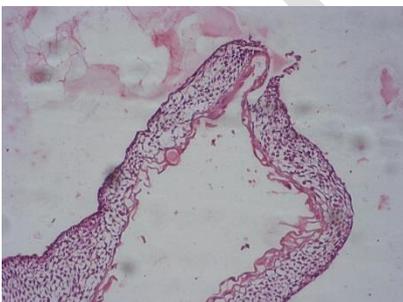


Fig. 6-A

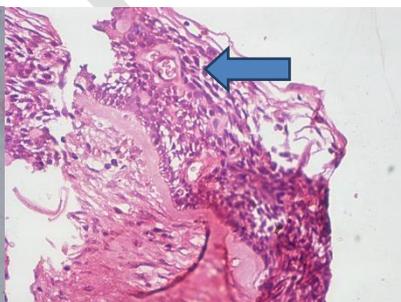


Fig. 6-B

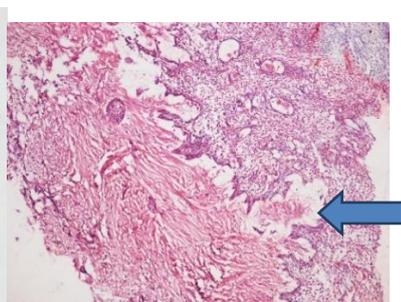


Fig. 6-C

*Fig. 6-A H & E stain showing cystic lining comprising of stratified epithelium with cuboidal basal cells and stellate reticulum like superficial cells. B- Blue arrow shows presence of ghost cells. C- Blue arrow shows Formation of dentinoid like material & ameloblastomatous cells.*

## **DISCUSSION:**

Rywkind in 1932 first described CCOT as a lesion of the jaw resembling cholesteatoma of ear & hence gave the name cholesteatoma of the jaw. In 1946 CCOT was described by Thoma & Goldman as strange variant of Ameloblastoma.<sup>3</sup> In 1962 Gorlin et al. reported CCOT as distinct histopathological entity due to presence of ghost cells and named it calcifying odontogenic cyst or "Gorlin Cyst".<sup>3,4</sup> Due to its extremely diverse clinical and histopathological biological behavior it was renamed as Calcifying Cystic Odontogenic Tumor by World Health Organization in its recent classification of 2005 of odontogenic tumors.<sup>7,8</sup> CCOT is a rare tumor forming around 2% of all odontogenic tumors.<sup>9</sup> Most of the CCOT are cystic in architecture & some are solid lesions. Some of the solid lesions have the neoplastic nature. CCOT can occur in age groups varying from 1yr to 82 yrs while most commonly seen in 2<sup>nd</sup> decade of life.<sup>3</sup> CCOT can occur intra or extraosseously and it occurs in both maxilla & mandible with same predilections.<sup>10</sup> CCOTs are mostly occurring in canine-premolar region in association with missing cuspids. CCOTs have also been reported to occur in association with other odontogenic tumors such as odontoma, adenomatoid odontogenic tumor, Ameloblastoma, ameloblastic fibroma, Pindborg tumor etc.<sup>11,12</sup>

Four variants of central or peripheral CCOT have been described<sup>4</sup>

CCOT type 1. Simple cystic CCOT. Includes pigmented and clear cell variants

CCOT type 2. Odontoma-associated CCOT

CCOT type 3. Ameloblastomatous proliferating CCOT

CCOT type 4. CCOT associated with benign odontogenic tumours other than odontoma

Of all these Ameloblastoma proliferating variant is extremely uncommon with only 30 cases have been reported. Of the 92 cases of CCOT reported by Hong et al only 11 were Ameloblastoma proliferating type. Out of reported 30 cases most were presented as painless growing swelling, only 3 cases reported by Kamboj and Manish, Abikshyeet et al. and Rama Raju et al. presented as painful swelling as in the present case. This type of CCOT have been reported in age range between 11 to 62 years with slight female predilection. Premolar- Molar region of the mandible is the most common site for this type of CCOT. Cases reported by Kamboj and Manish, Shailesh M. et al. showed the extension of the lesion up to coronoid and condylar process as in our case.<sup>4,16</sup> Radiographically, initially it appears as a unilocular radiolucency and in later stages a mixed radiolucent-radioopaque appearance can be said as "salt and pepper type of pattern".<sup>4</sup>

Hong et al. proposed 2 categories of CCOT associated with Ameloblastoma: The neoplastic variant in association with Ameloblastoma and ameloblastomatous cystic variant. The former is called Ameloblastoma ex CCOT which is characterized histologically as having some or no ghost cells along with calcifications of transformed ameloblastomatous epithelium. The ameloblastomatous cystic variant has unicystic structure while the lining epithelium reveals uni or multifocal proliferations mimicking Ameloblastoma.<sup>3, 13</sup> In our case histologically there was presence of multifocal proliferations along with the ghost cells pointing out towards the presence of ameloblastomatous cystic variant of CCOT. Histologically the present case showed cystic lining comprising of ghost cells some areas of dysplastic dentin. Proliferative odontogenic islands were also seen which led to final diagnosis. Ameloblastomatous CCOT has common histologic

features as unicystic Ameloblastoma apart from presence of ghost cells & calcifications. As the literature suggests treatment of choice for ameloblastomatous CCOT is simple enucleation of the lesion.<sup>14, 15</sup> Among all the reported case only one case reported by Yuwanati et al. showed recurrence.<sup>4</sup> So, the present case was also treated by same treatment modality and 18 months follow-up did not show any recurrence.

### **CONCLUSION:**

This case was not possible to diagnose just by clinical or radiological means alone. Biopsy was mandatory to reach the final diagnosis. It was challenging to reach final diagnosis because of the unusual features which were presented. It further highlights a wide range of clinical and histological presentations which can be shown by odontogenic tumors in general & CCOT in particular. Diverse biological behaviors of these lesions present a challenge to the clinicians to diagnose and treat them.

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