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Ameloblastoma Arising from Gorlins Cyst: A Rare Case Report

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Abstract:- Gorlin cysts aka calcifying epithelial odontogenic cyst (CEOC) is a rather diverse clinical entity with a wide range of clinical and histopathological variants, posing a challenge to its accurate diagnosis. Complicating this, is the fact that most cystic and neoplastic variants are associated with benign tumors of the oral cavity, especially, Ameloblastoma. This case report entails that of a 27 year old male patient, who reported with complaints of a painful swelling over the left lower jaw region, and was subsequently diagnosed preoperatively as a case of CEOC but later on specimen in Toto histological evaluation revealed areas of Ameloblastomous changes.

Keywords:- *Ameloblastoma, Calcifying Epithelial Odontogenic Cyst (CEOC) Calcifying Cystic Odontogenic Tumor (CCOT), Enucleation, Gorlins Cyst.*

I. INTRODUCTION

Much like odontogenic keratocyst, the CEOC also has undergone ambiguity regarding its nomenclature and classification. Described first as an odontogenic epithelial cyst in 1962, it has however undergone various reclassification due to its variable histology, clinical behavior and association with dysregulated β-catenin signaling, leading to confusion till date whether or not it is a reactive or developmental or neoplastic entity.[1] These are particularly notorious in their association with other tumors such as Adenomatoid Odontogenic tumor, Odontomas and Ameloblastoma. There has been only few such reported cases, about less than 22 wherein theirs ameloblastomaous changes. Here in this case report we present a rare case of Ameloblastoma arising from a biopsy proven lesion of CEOC.

II. CASE REPORT

A 27 year old male patient presented to the department of Oral and Maxillofacial Surgery, with chief complaint of swelling over the left lower front tooth region since past six months. Patient noted a small asymptomatic swelling over the gum in the left lower front teeth region, which had progressively grown to the present size. The swelling was associated with pain and paresthesia on presentation. On general examination, patient was well nourished and well built. The medical history of the patient was unremarkable.

On local examination, facial asymmetry due to swelling in the left lower jaw region, angle of mouth lifted slightly on the left side.[Fig 1] Skin over the lesion was normal and not breeched. On palpation, swelling had a hard bony consistency, non-compressible, non-fluctuant in nature, not fixed to skin. Tender on palpation, associated with paresthesia. Neck nodes were not palpable.

On Intra Oral Examination, mouth opening was adequate. A Swelling extending from 41 to 37 region, obliterating the buccal vestibule.[Fig 2]

Mucosa not involved. Though it was hard in consistency, there were certain areas of decortication. There were no signs of inflammation or pus discharge. The teeth involved were not mobile or tender on percussion. On pulp testing, certain teeth, namely, 32,33,34,35 were seen to be non-vital. On the first seating aspiration and bone biopsy was done. The aspirate was serosanguinous in nature. Based on these clinical findings, a provisional diagnosis of Ameloblastoma, Odontogenic keratocyst, Central Giant Cell Granuloma and Aneurysmal Bone Cyst were discussed.

- *Investigations: -*

OPG revealed well defined radiolucent multilocular lesion extending from below the apex of 41 to 37, root resorption noted with respect to 36. Lower border continuity noted, no pathological fractures or tooth displacement seen.[Fig 3]

CT scans revealed a lytic expansile lesion with certain areas of decortication with respect to the buccal cortex, thinning of the lingual cortex as well and the inferior alveolar nerve canal was seen to to be compressed to lower border of the mandible.[Fig 4] All other routine investigations were done and medical and anesthetic fitness is obtained. The bone biopsy results revealed a case of Calcifying odontogenic cyst.

- *Treatment Done: -*

Based on the findings, a treatment plan was devised. Cyst enucleation under GA, with chemical cauterization and peripheral osteotomy was planned. On endodontic opinion patient was advised intentional RCT with respect to 31,32,33,34,35,36,37. Pulp extirpation was done pre-operatively. Patient is advised to complete his root canal treatment postoperatively.

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➢ *Surgical Procedure: -*

After induction of GA, nasal intubation was done and patient was painted and draped aseptically, LA infiltration using lignocaine 2% with adrenaline 1:1,00,000 was given at the lower vestibular region. Through an intraoral vestibular approach, with incision extending from 43 to 37 region. The mucoperiosteal flap is reflected, taking care not to injure the mental nerve. The buccal cortex was thin and fragile and had areas of fenestration. The cystic lining was carefully teased out without perforating the walls and the mass was removed in Toto.[Fig 5] The nerve canal was intact and nerve not breeched. About 2-3 mm bone is removed through peripheral osteotomy using a mastoid bur. A cotton pledged with modified Carnoys solution is packed into the bone cavity, left for 3-5 minutes and removed. The bony cavity is irrigated with saline, antiseptic solutions and hemostasis achieved. The cavity is packed with sticky bone (hydroxyapatite blocks and I-prf) and closure done. All the teeth in the affected side is preserved as per endodontic opinion.

The post op specimen's histopathology revealed ―CEOC giving rise to Ameloblastoma‖.

III. DISCUSSION

The Gorlins cyst accounts for only about 1- 2% of all odontogenic cysts. Further less, is the incidence of a mixed variant seen in association with another odontogenic entity. The lesion was considered the bony counterpart of the cutaneous CEOC, the *Cyst of Malherbe*.

It's been classified broadly as solid and cystic variant, but this alone fails to include all the different clinic pathological behavior of the lesion. And hence, several other classification system exists over the past years. In 1991, Hong et all classified it as broadly the cystic and the neoplastic variants each having 3 subvariants.

Some authors have described COC as calcifying cystic odontogenic tumor (CCOT) as they have more heterogeneous components than envisaged. Between 2005 and 2017, WHO changed and then reverted back the status of the lesion from being a neoplasm to that of a cystic nature due to its non-neoplastic clinical behavior.[2]

Ameloblastomatous CEOT shares common features with unicystic ameloblastoma except for the ghost cells and calcifications. It fulfills the *Vickers and Gorlins Criteria* as the cystic lining has the capacity to transform into a true ameloblastoma.[3]

The recurrence is low **(3%–11%)** but requires a radio-clinical follow-up of 6 months after the procedure and then annually for 5 years to check for complete reossification. One can never rule out the low risk of transformation into a ghost cell malignant tumor as well.[4,10]

Malignant transformation can occur in CEOT rapidly or after a long time. Arashiyama et al. reported a case of calcifying odontogenic cyst that transformed to malignancy after eighteen years. Therefore, long- term careful follow up of the patients is also recommended.[5]

Tamanna et al. reported a case of a 21-year-old male patient with a painful swelling in the mandibular left back tooth region. Marsupialization was done and the histopathologic features were suggestive of COC. The patient came with a recurrence a year later. This time microscopic examination revealed well-defined cystic lining with ghost cells and calcifications and diagnosis of CEOC with ameloblastomatous proliferation was made. The lesion was surgically removed and thereafter, no recurrence was noted in a year of follow-up[6].

In another prospective study by Patel H et al, a dredging method was done for large cystic lesion exceeding a minimum of 2 cms or in close proximity to vital structures and tooth buds. The method included 3 steps, namely decompression, followed by repeated scraping of the lesion, and finally, peripheral ostectomy providing margin clearance but without significant morbidity. It was concluded that the dredging method may be considered as a modality in between conservative and radical treatment options.[7]

Despite the obvious risk of recurrence, conservative treatmentwith enucleation and curettage seemed to be justified in preference to mutilating radical surgery. In general, lesion of maxilla are more often associated with positive or involved margin post resection and subsequently higher rate of recurrence than that of mandible. In this case, the easily accessible site of the lesion and well encapsulated nature allowed complete enucleation. It was adjuncted with chemical and mechanical cauterization. The patient was counselled and explained about the chances of recurrence and of malignant transformation, the peculiarity of his lesion as noted by specimen histopathology and the possible need for aggressive treatment if it is to recur. The patient is being followed up closely.

IV. CONCLUSION

A primary lesion of CEOC can be treated conservatively with enucleation and curettage, but one must keep in mind that even though the recurrence rate of the lesion is low, more often than not, its seen to give rise to odontogenic benign tumors from its cystic lining, which in turn have the potential to undergo malignant transformation.[8] And hence, when available and not economically constrained, a thorough Immunohistochemically assay revealing certain predictable factors, such as atypical histologic features, necrosis, prominent mitoses, infiltrative growth pattern, aggressive behavior, and high expression of Ki67 and p53 should be undertaken for all CEOC lesion.[9]

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Fig 1 Swelling in the Left Lower Jaw Region with Lifted Angle of Mouth

Fig 2 Swelling Extending from 41 to 37 Region, Obliterating the Buccal Vestibule

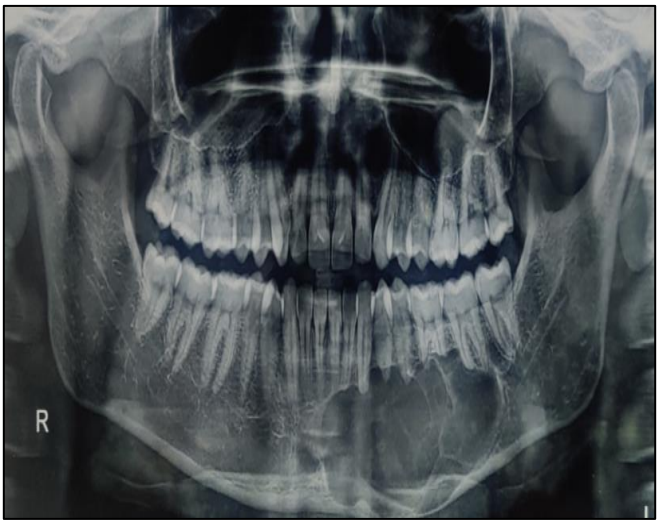

Fig 3 OPG Revealed well Defined Radiolucent Multilocular Lesion

Fig 4 CT Scans Revealed a Lytic Expansile Lesion with Certain Areas of Decortication

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Fig 5 The Cystic Lining was Carefully Teased Out

Fig 6 Post Operative OPG