Ameloblastomatous Calcifying Cystic Odontogenic Tumour: A Rare Variant

ABSTRACT
Calcifying Cystic Odontogenic Tumor (CCOT) was previously described by Gorlin et al., in 1962 as Calcifying odontogenic cyst. CCOT is a rare lesion which accounts for 2% of all odontogenic pathological changes in the jaws. One of the variants, Ameloblastomatous proliferating type of CCOT is even more rare and very few cases have been reported in the light of literature review. This case report is an effort to bring forth a case of ameloblastomatous proliferating type of CCOT in a 65 year male, who presented with a painful swelling in the right jaw crossing midline causing facial asymmetry and confirmed by histopathological evaluation.

CASE REPORT
A 65-year-old male presented with a swelling in the right lower jaw since four months. History revealed a slow progressive, painful swelling associated with watery discharge on chewing hard foods. Extra orally, gross asymmetry was detected. Intra oral examination revealed well defined, tender, soft, fluctuant swelling extending from 32-47, obliterating right buccal, lingual and labial vestibules. Grade II & III mobility noticed with 31, 32 and 41-46 respectively. Provisional diagnosis of Ameloblastoma was given. Differential diagnosis of CCOT, central giant cell granuloma and calcifying epithelial odontogenic tumour were considered. OPG revealed well defined radiolucency from 32-47 region with mixed internal appearance CT, 3D CT revealed hypodense area extending from 32-47 region. Microscopically, cystic lumen lined by stratified squamous epithelium with basal ameloblast like cells and superficial stellate reticulum-like tissue intermixed with few ghost cells. At focal areas epithelium also exhibits intramural proliferations. Within the connective tissue, few ameloblastomatous islands with peripheral flattened to tall columnar cells having hyperchromatic nuclei placed away from basement membrane and central stellate reticulum like tissue and focal areas of squamous metaplasia are also evident. Connective tissue also exhibits focal areas of hyalinization. Final diagnosis of, “Calcifying cystic odontogenic tumour type 3” was given. Patient was chalked out for surgical enucleation under general anesthesia but the patient showed reluctance towards the treatment.

DISCUSSION
Early literature states that, Gorlin et al., considered CCOT as a separate histopathological entity, owing to the presence of “ghost cells” [1]. Since its first description controversy and confusion have prevailed regarding its nature which propelled various taxonomical classifications and nomenclatures [2]. After many break throughs in the presentation and behaviour of Calcifying odontogenic cyst, finally it secured a place in the group of Odontogenic tumours. In 2005, WHO proposed a Histological classification of Odontogenic tumours in which Calcifying odontogenic cyst was renamed as “Calcifying cystic odontogenic tumor” (CCOT) by Ledesma-Montes [3] due to its neoplastic behavior [Table/Fig-2].

Hong et al., reported 92 cases of CCOT, of which only 11 were ameloblastomatous type [4]. To the best of our knowledge, only 30 ameloblastomatous CCOT cases are reported in the literature.

Keywords: Ameloblastomatous proliferations, Calcifying odontogenic cyst, Ghost cells, Odontogenic tumours

Table/Fig-1: A-B. Extra oral photographs showing swelling causing facial asymmetry in the right mandible; C. Intra oral photograph showing indentations of the opposing teeth over the swelling with supra eruption of 44 and 45. D. Radiolucency with displacement and divergence of roots with 44-45. E-F. Reconstructed 3D views showing a large, expansile lesion with lysis of right buccal & lingual cortices. G. H & E section (10 x) showing ghost cell with in the odontogenic epithelial component (blue arrow). H. H & E section (10 x) showing ameloblastomatous island (blue arrow) with central areas of squamous metaplasia (green arrow) surrounded by delicate to dense connective tissue

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Ameloblastomatous CCOT can be differentiated from ameloblastoma ex CCOT by lack of histopathologic criteria suggested by Vickers and Gorlin. According to Yoshida et al., immunohistochemical studies suggested greater mean Ki-67-LI in ameloblastomatous CCOTs than in those without these histological features, representing a high proliferation potential associated with ameloblastomatous proliferation. Out of seven cases of ameloblastomatous CCOT, six cases (86%) showed expression of Bcl-2 protein [9]. Conservative surgical enucleation is the treatment of choice [5] as prognosis is good with no recurrence, indicating non neoplastic nature of the lesion [2]. Of all the reported cases only one case reported by Yuwanati et al., showed recurrence after six yrs [5].

CONCLUSION

The present case represents the classic features of ameloblastomatous CCOT, which comes under the category of CCOT type 3 (according to Ledesma Montes et al.), which is a very rare odontogenic lesion. Such cases highlight the varied presentation of CCOT and throws light towards its behaviour.

REFERENCES