DENTINOGENIC GHOST CELL TUMOR - A CASE REPORT

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INTRODUCTION
Gorlin et. al first described the term “calcifying odontogenic cyst” (COC).[1] Not all of these lesions are cystic and the biological behaviour is often not compatible with a cyst.[2] This dualistic concept evokes a controversy whether COC is a cyst or tumor. Few authors consider COC to have 2 entities: a cyst and a neoplasm. The WHO classified all COCs to be neoplasm. The cystic lesions are termed as “calcifying cystic odontogenic tumors” (CCOT) and the neoplastic entity as a “Dentinogenic ghost cell tumor”(DGCT).[3] DGCT is a rare tumor accounting for only 11.5% of all COCs.[2] Age of incidence is usually in the elderly with a male predilection. It commonly involves the anterior segment of the jaws. [2] Microscopic features comprise of ameloblastomatous epithelial islands, with areas of ghost cell formation and varying amount of dentinoid material. [1]

CASE REPORT
A 25 year old woman reported to the dental OPD with the chief complaint of pain and swelling in the lower right back region of jaw. (Fig 1) She apparently noticed a pea sized
swelling intra orally in the same region about 6 months ago which gradually increased to the present size and further resulted in the extra oral swelling as well. According to the patient, the pain was dull, throbbing in nature occurred spontaneously and subsided on medication. The swelling presented no aggravating or precipitating factors. Patient had undergone an eventful extraction of 46, a decade ago. She had no underlying medical condition or history of prior hospitalization. On lifestyle evaluation, there seemed to be no apparent physical or mental stress. History of exposure was negative. A non-vegetarian by diet, her daily oral care comprised of use of a toothbrush and fluoride containing toothpaste in a horizontal motion. General assessment revealed patient was in a good, conscious state and co-operative in behavior. A single, diffuse swelling extending from the right ala tragus line superiorly to the lower border of the mandible inferiorly and from the right lateral border of the nose to 1 cm behind the outer canthus of the eye antero-posteriorly was observed. The was no change in the colour, texture or temperature of the overlying skin or the surrounding area.(Fig.2A & 2B). Soft to firm consistency with moderate tenderness was noted. Right submandibular and pre auricular lymph nodes were palpable and tender. On TMJ examination, there was no deviation, deflection or clicking sound on opening or closing of mouth. Intra oral examination reveals missing 28,46 and 48. 26 was root stumps and caries was present in relation to 36, 37. Periodontal status showed firm, pale pink gingiva with melanin pigmentation and scalloped contour. No other abnormality was detected in the oral cavity. Intra oral examination revealed a bony hard swelling in the right posterior region of the mandible extending from 46 region to the pterygomandibular region involving both the floor of the mouth lingually and the adjacent buccal vestibule. It appeared to be of the same colour as that of the normal mucosa except near 48 where it occurred erythematous with whitish nodular specks. On palpation, the swelling was found to be bony hard except in the region of 48 where it was soft to firm in consistency. Tenderness was present. Obliteration of both the buccal and lingual cortical plates were also noted. Based on the clinical presentation, a provisional diagnosis of aggressive tumor or cyst was given. Further investigation included radiographic examination through CBCT which revealed presence of a well defines radiolucent mass extending from 45 to 48 region with perforation of both the buccal and lingual cortical plates.(Fig.3A &3B). A biopsy was performed at the concerned site. Histopathologic examination revealed odontogenic islands with hyperchromatic ameloblastoma like cells and central polygonal cells surrounded by mature connective tissue stroma. Multiple masses of eosinophilic dentinoid or osteodentine like substance interspersed
between the odontogenic islands were evident. The overall histopathological features were suggestive of dentinogenic ghost cell tumor. Surgical intervention involving right hemimandibulectomy was conducted and a post op histopathological examination of the site was performed. (Fig. 4) It revealed anastomosing cords, strands as well as follicular areas lined by columnar cells with hyperchromatic nuclei arranged in palisading pattern with reverse polarity. The core was composed of connective tissue resembling stellate reticulum whereas focally hypercellular areas comprising of sheets of spindle cells were seen. These features were suggestive of an ameloblastoma. Hence based on the post op analysis a dinal diagnosis of ameloblastoma in the right posterior mandible was given.

DISCUSSION
In this case both pre-op and post-op biopsy was conclusive of odontogenic tumors such as ameloblastoma, odonto-ameloblastoma, ameloblastic fibro-odontoma, odontoma, adenomatoid odontogenic tumor, and cementoma. The presence of numerous ghost cells, and masses of dentinoid material in the pre op biopsy suggested dentinogenic ghost cell tumor (DGCT). This is one of the characteristic feature of DGCT.[4] These ghost cells are nothing but cells that show resistance to degradation and are characterized by the the loss of nuclei, preservation of basic cellular outlines. These may appear blurred and fused and hence the name “ghost cells”. [5] Here lies a diagnostic dilemma as to whether the lesion should be diagnosed as an ameloblastoma or a DGCT. Clinically, they present with similar characteristics, however histopathologic analysis reveals the distinguishing features that help the diagnostician conclude a final diagnosis.

CONCLUSION
So, this is a case where the provisional diagnosis was contraindicated based on the histopathologic analysis.

LEGENDS:
- FIG. 1 – Extra oral examination.
- FIG 2- Intra oral examination showing right posterior region
- FIG 3- CBCT showing buccal and lingual cortical plate perforation.
- FIG 4- post op surgical specimen after right hemimandibulectomy.

Keywords:
Dentigerous ghost cell tumor, ameloblastoma, diagnostic dilemma, bony swelling, mandible.
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