Granular cell Ameloblastoma of the Jaw: A Case Report and Review of Literature

Satish Kumar Bansal¹, Gopal Singal², Umesh Chhabra³, Sandeep Kumar Goyal³, Yudhvir Singh⁴, Geetanjali Gupta⁵
¹Associate Professor, ²Professor, ³Assistant Professor, ⁴Senior Resident, Department of General Surgery, ⁵Junior Resident, Department of Dentistry, MAMC, Agroha, Hisar, India

Abstract:
Ameloblastoma is a neoplasm of odontogenic epithelium, especially of enamel organ-type tissue that has not undergone differentiation to the point of hard tissue formation. It accounts for approximately 10% of all tumors originating from gnathic bones. It exhibits diverse microscopic patterns which occurs either singly or in combination with other patterns. Granular cell ameloblastoma is a rare condition, accounting for 3.5% of all ameloblastoma cases that shows marked transformation in the cytoplasm of tumor cells, which are usually stellate reticulum like cells. It is also considered an aggressive variant of ameloblastoma with potential recurrence and malignant transformation. This article describes a case of granular cell variant of ameloblastoma affecting a 50-year old male.

Key Words: Ameloblastoma, Granular Cell, Odontogenic Tumor

Corresponding Author: Dr. Satish Kumar Bansal, Associate Professor, Department of General Surgery, MAMC, Agroha, Hisar, India, E mail: capric2@yahoo.com


INTRODUCTION
Ameloblastoma is a neoplasm of odontogenic epithelium, especially of enamel organ-type tissue that has not undergone differentiation to the point of hard tissue formation.¹ It often presents as a slow growing, painless swelling, causing expansion of the cortical bone, perforation of the lingual and buccal plates and infiltration of soft tissue.² It occurs in three different clinical-radiographic patterns: conventional intraosseous solid and multicystic (86%), unicystic (13%) and peripheral (1%). The common histological patterns include the follicular (32.5%) and plexiform (28.2%) ones, less common being the acanthomatous (12.1%), granular, desmoplastic (8.6-13%) and basal (2%) forms.³ Granular cell ameloblastoma (GCA), a relatively rare histologic subtype, representing 3-5%, is characterised by large round/cuboidal eosinophilic cells with granular cytoplasm and, in most instances, it is blended with a follicular subtype.⁴ Only few cases of granular ameloblastomas are reported in the literature. The purpose of this article is to present a case of unusual variant of ameloblastoma highlighting its unique microscopic features that allow its distinction from other jaw tumors with a granular cell consistency.

CASE REPORT
A 50-year old male reported with a swelling in the lower left back tooth region since 2 years. Patient noticed small swelling which was initially pea-nut in size and progressed to present size. There was no significant past medical history. Extraorally, facial asymmetry was noted on left side of face. The swelling was 4x2cm in size extending antero-posteriorly from parasympysis to angle of mandible and supero-inferiorly 2 cm from angle of mouth to inferior border of mandible on left side (Figure 1). Tender on palpation and was firm in consisteny. Intraoral examination revealed a diffuse swelling in the mandibular posterior region on left side extending along the buccal vestibule extending from 34 to 37 which was irregular in shape, pale pink in color, stony hard and tender. Obliteration of the buccal vestibule was seen in relation to 36. Missing
teeth in relation to 36, 37. Aspiration of the lesion revealed reddish brown fluid. Orthopantamograph revealed an ill-defined radiolucent area extending from 33 to angle of mandible with a discontinuity in lower body of mandible on left side. Lesion exhibited a compartmented appearance with septa of bone extending into the radiolucent tumour mass (Figure 2). A provisional diagnosis of ameloblastoma, calcifying epithelial odontogenic tumor, or a fibro-osseous lesion was made and a biopsy was performed. The selected treatment planning was enucleation and reconstruction with non-vascularised iliac bone graft, and osteosynthesis by plate and screws. HE stained sections revealed ameloblastic epithelium arranged in follicles consisting of stellate reticulum-like cells in the mature fibrous connective tissue stroma. Follicles show palisaded, peripheral tall columnar cells with nuclear hyperchromatism and reverse polarity. Few epithelial islands enclosing eosinophilic granular cells were evident (Figure 3). The final diagnosis of Granular Cell Ameloblastoma was made. The postoperative period went without complications, with good aesthetic and functional final results. At 1.5 years follow up, there are no signs of recurrence.

**DISCUSSION**

GCA represents a rare variant of ameloblastoma only 1 out of 77 ameloblastoma cases was classified as the granular cell subtype. Reichart et al. reviewed all available literature on ameloblastoma of the jaws from 1960 to 1993 and reported that out of a total of 1593 cases with available data on histologic subtypes; there were only 56 (3.5%) cases of the granular cell variant. Granular cells are just a transitional or matured phase in the lifecycle of ameloblastomas, starting with normal stellate reticulum like cells leading to production of granules and finally leading to degeneration and formation of cystic areas.

Numerous theories have been put forward on the origin and nature of these granular cells in ameloblastomas, with an epithelial origin, several ultrastructural and histochemical studies have described them as lysosomes and many authors have reported the immunohistochemical (IHC) findings of GCA, yet the actual contents within the granular cells remains largely undefined. It was speculated that, with age, the unnecessary aged components progressively increase in the cytoplasm of some of the tumor cells, while the ability of lysosomes to eliminate these materials decreases; hence, their cytoplasm becomes packed with lysosomal granules. A recent IHC and ultrastructural study suggests that the cytoplasmic granularity might be attributed to the increased apoptotic cell death of the neoplasic granular cells, and to their subsequent phagocytosis by the adjacent granular cells.

**Figures**: 1): Clinical picture showing facial asymmetry on left side of face; 2): Tumour exhibits a compartmented appearance with septa of bone extending into the radiolucent tumour mass. 3): Low power view shows islands of enamel organ like tissue consisting of stellate reticulum like cells in the mature fibrous connective tissue stroma; 4): High power view shows peripherically placed ameloblast like cells with nuclear hyperchromatism reverse polarity. Eosinophilic granular cells are also evident.
The most recent studies suggested that the synthesis of signalling molecules, such as beta-catenin and Wnt-5a, is upregulated in the granular cells of GCA, but their transportation or secretion is impaired, their accumulation within granular cells, as autophagosomes, thus resulting. Also, the heparinase enzyme, which is associated with invasiveness, fails to be activated, being accumulated within these cells, in its latent form. Of the odontogenic tumors, granular cells have been described in the granular cell ameloblastoma and the granular cell ameloblastic fibroma. Similar cells occur in the congenital epulis, a lesion regarded by some as a product of dental epithelium and more specifically of ameloblastic. The striking morphological and tinctorial resemblance of the granular cells in these three lesions to those of the granular cell myoblastoma have led some authors to conceive congenital epulis as a granular cell myoblastoma and of the GCA as a coincidental mixture of ameloblastoma and granular cell myoblastoma. The origin of the, granular cells has been a matter of great speculation as concerning histogenesis, the GCA's are of epithelial nature, and arise from ameloblasts. Conversely, the granular cells found in other lesions of the oral cavity are of mesenchymal derivation.

CONCLUSION
Early diagnosis and prompt surgical treatment in granular cell ameloblastoma is of prime importance. Noteworthy is that granular cell ameloblastoma ‘s may rarely behave in a malignant fashion giving rise to metastasis. Patients should be kept under periodic observation because of reports of recurrences even up to 8 years after initial treatment.

REFERENCES

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