

## Granular Cell Ameloblastoma

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### Abstract

Ameloblastoma is an epithelial odontogenic tumour of the jaw and exhibits diverse histopathologic subtypes like follicular, plexiform, acanthomatous and desmoplastic variants which occurs either singly or in combination. Granular cell ameloblastoma is a rare histological subtype of ameloblastoma accounting for less than 5% of the total. The rare granular cell variant is seen in combination usually with follicular or plexiform subtypes. The aim of this paper is to describe clinical and microscopic features of Ameloblastoma of a pure granular cell histopathological subtype occurring in a seventeen year old adolescent female patient in the lower right region of the mandible.

**Key Words:** Odontogenic tumors, Ameloblastoma, granular cell ameloblastoma

### Introduction

Ameloblastoma is a true neoplasm of odontogenic epithelial origin characterized by a local invasiveness and high frequency of recurrence. It includes several clinicoradiographic and histologic subtypes like follicular, plexiform, acanthomatous, basaloid and granular cell types. Follicular and plexiform are the commonly encountered variants accounting to 32.5% and 28.2% respectively, followed by the acanthomatous subtype with 12.1% while desmoplastic is extremely uncommon with incidence rates ranging from 4-13%. Less common histopathologic subtypes include the granular cell and basal cell ameloblastoma.<sup>1</sup> Although the treatment and prognosis are virtually the same (with the possible exception of more aggressive desmoplastic variant), knowledge of various histopathologic subtypes is a prerequisite for accurate diagnosis and management.<sup>2</sup> The granular cell Ameloblastoma is a relatively rare histologic subtype (less than 5%)<sup>3</sup>, and in most instances, it is found as an admixture with other histologic patterns particularly the follicular subtype.<sup>4</sup> The granular cell subtype of ameloblastoma is characterized by the groups of round, cuboidal or columnar granular cells,

which have abundant cytoplasm filled with eosinophilic granules. The granules have been identified as lysosomal aggregates, both ultrastructurally and histochemically. The acquisition of granular cell phenotype has been attributed to an aging or degenerative change in long-standing lesions; however, it may also affect young patients.<sup>1</sup> When this granular cell change is extensive in an ameloblastoma, the designation of granular cell ameloblastoma is appropriate.<sup>5</sup> The purpose of this paper is to present an unusual case of granular cell ameloblastoma to highlight its unique microscopic features that allow its distinction from other jaw tumours with a granular cell constituency.

### Case report

A 17 year old female was referred to Department of Oral Pathology with the chief complaint of pain and swelling in relation to lower right jaw region mandibular since 1 year. Past medical, dental & family history of the patient was noncontributory. There was no history of trauma, sinus opening or pus discharge. Extraoral examination revealed facial asymmetry due to swelling on lower right side of the face extending from parasymphysal region to 2cm away from the corner of mouth anteroposteriorly. Superiorly, the swelling extended from middle third of the cheek till inferior border of mandible (Fig. 1). Intra-oral examination revealed obliteration of buccal sulcus in the region of 43,44,45,47 with both buccal and lingual cortical expansion (Fig. 2). On palpation the swelling was bony hard in consistency with no associated lymphadenopathy. On Radiological examination, the OPG revealed well defined multilocular radiolucent lesion extending 33 to 37 periapical region with root resorption of 36

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Fig. 1: Photograph showing extraoral swelling on the right side of the lower jaw.



Fig. 2: Photograph showing intraoral swelling on the right mandible with expansion of buccal and lingual cortical plates.

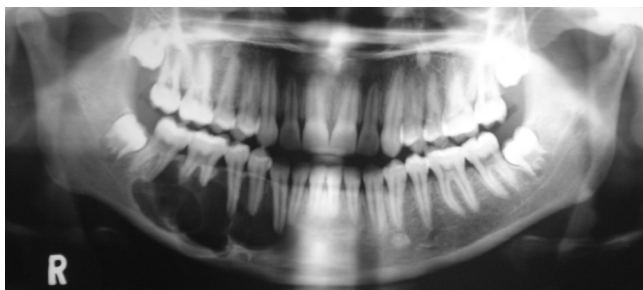


Fig. 3: Photograph showing OPG radiograph multilocular radiolucency from 43 to 47

(Fig.3). A provisional diagnosis of Ameloblastoma and Odontogenic Keratocyst was considered and an incisional biopsy was taken. Macroscopically, the specimen received was 3.0 x 2 cm in size, grayish brown in color with firm consistency. The cut section H & E stained sections showed the presence of areas of follicles with peripheral palisading preameloblast with central granular cells replacing the stellate reticulum like cells. The connective tissue stroma is loose fibrous. The center of the follicles showed cystic degeneration. (Fig. 4, 5)

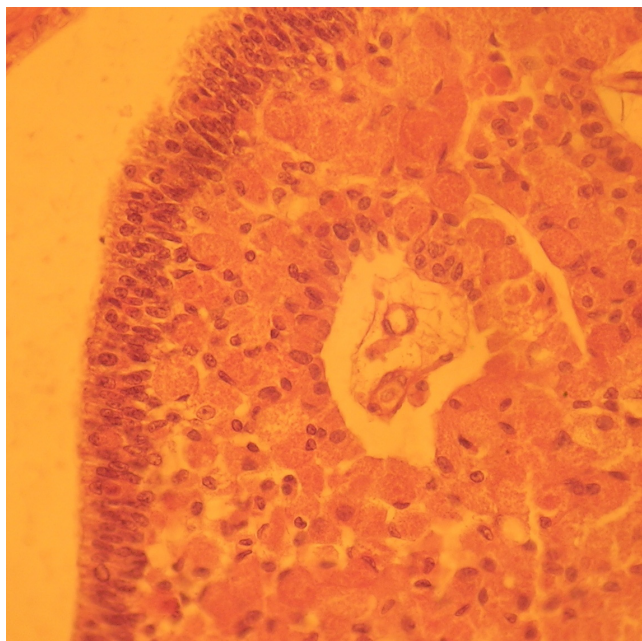


Fig. 4: Photomicrograph showing ameloblastic follicles with peripheral layer of ameloblasts with central granular cells (10X)

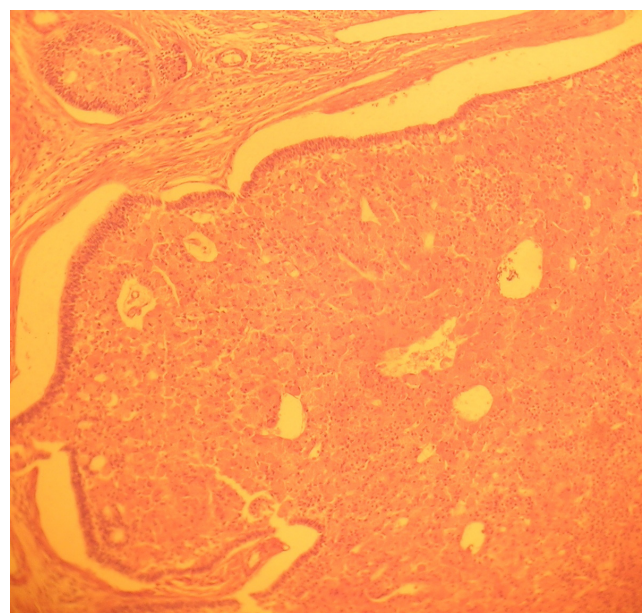


Fig. 5: Photomicrograph showing large cuboidal to oval cells packed with dense eosinophilic granules (40X)

## Discussion

Ameloblastoma chiefly occurs predominantly in 4<sup>th</sup> - 5<sup>th</sup> decade of life and the age range is very broad. The average age of patients with intraosseous ameloblastoma has been reported to be 39 years. The rare lesions occurring in adolescents are usually cystic and appear clinically as odontogenic cysts. In this study, we document the occurrence of granular cell ameloblastoma in a significantly younger patient.<sup>6,2</sup>

Granular cell ameloblastoma is a rare variant of ameloblastoma. According to Reichart et al.<sup>1</sup> 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. Hartman reviewed 20 cases of the granular cell ameloblastoma and reported an average age of 40.7 years (age range: 21 - 65 years) with no distinct gender predilection. The majority of the lesions were reported in mandible with propensity towards the posterior regions of the mandible<sup>5</sup>. Jaw swelling and pain were the most frequent presenting symptoms. There have been no distinguishing radiographic findings for granular cell Ameloblastoma reported. Our case showed similar clinico radiographic parameters except that it was reported in a 17 year old female patient.

The defining characteristic of granular cell ameloblastoma is the presence of granular cells in the central portion of the epithelial islands, strands and cords. The granular cells tend to be large and have an oval to polyhedral outline. The follicles may have a thin rim of stellate reticulum like calls that separates the granular cells from the peripheral columnar layer. The nucleus is displaced to the periphery of the cells. Prominent coarse granules tend to stain eosinophilic and pack and distend the cytoplasm, imparting a distinctive appearance. The granular cells rarely show a distinctive cell borders and the cytoplasm merges imperceptibly. Originally they were considered to represent an aging or degenerative process but recent immunohistochemical studies suggest that this phenomenon is related with increased apoptotic cell death of the lesional cells and the phagocytosis by neighbouring neoplastic cells.<sup>7,8</sup> On Immunohistochemical analysis it is seen that the granular cells show positivity for cytokeratin, CD68, lysozyme and alpha-1-antichymotrypsin, but are negative for vimentin,

desmin, S-100 protein, neuron-specific enolase and CD15, indicating epithelial origin and lysosomal aggregation<sup>8,9</sup> Dina et al. also showed that the granular cells exhibited membranous positivity for cytokeratin and cytoplasmic positivity for CD68.<sup>10</sup>

The differential diagnosis of granular cell ameloblastomas includes other oral lesions with a similar morphology of granular cell accumulation, including granular cell odontogenic tumour, granular cell tumour and congenital epulis. These lesions have different biologic behaviour and should be discriminated from granular cell ameloblastomas.<sup>11</sup>

## Conclusion

The granular cell ameloblastoma is a rare condition with unique histopathologic and immunohistochemical findings; its treatment and prognosis do not significantly differ from those of the other subtypes of the solid/multicystic ameloblastoma.

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