

Granular cell ameloblastoma

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Abstract

A 42-year-female patient presented with a swelling on the left side of the face for the past 10 years. The radiograph shows multilocular radiolucency with evidence of root resorption. Histopathology reveals fibrous connective tissue exhibiting numerous odontogenic epithelial islands with peripheral tall columnar cells showing a reversal of polarity. The center of the island shows stellate reticulum like cells. The connective tissue also shows the presence of extensive coarse granular eosinophilic cells distributed throughout the section.

Keywords: Benign odontogenic tumor, granular cell, granular cell ameloblastoma

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INTRODUCTION

Odontogenic tumors are derived from the epithelial or mesenchymal remnants of the tooth forming apparatus.^[1] Ameloblastoma is well recognized as a locally invasive benign neoplasm thought to arise from the cellular components of the enamel organ. It is an epithelial odontogenic tumor of jaw that exhibits diverse microscopic patterns and occurs either singly or in combination with other patterns.^[2]

It was described by Broca in 1868 and constitutes about 1%–2% of all cyst and tumors of the jaw. It is most often seen in the posterior mandible in the region of the third molar. There is no definite sex predilection and is seen most commonly in fourth and fifth decades. The most common clinical presentations are painless slow-growing swelling, accompanied by facial deformity, malocclusion, tooth loss and pain and paresthesia of the affected region.^[2] Granular

cell ameloblastoma is a less common histological subtype of ameloblastoma.^[3] This article highlights the rare tumor with its unique microscopic feature that distinguishes itself from other jaw tumors.

CASE REPORT

A 42-year-old female patient presented with a swelling in the left side of the face for the past 10 years. A firm large swelling was present in the left side of the entire mandible with sinus opening at the lower border of mandible. Intraoral examination showed swelling with obliteration of sulcus in relation to the left side of the mandible. The swelling was small in size initially which grew progressively to a size of 20 cm × 15 cm in duration of 3 months. Mucosa appeared normal. Specimen radiograph shows bilateral expansion of both the buccal and lingual cortical plates [Figure 2]. Multilocular radiolucency (honeycomb pattern) is also evident. Root resorption in the molar teeth is evident [Figure 3].

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Histopathology of the given section shows fibrous connective tissue exhibiting numerous odontogenic epithelial islands with peripheral tall columnar cells showing a reversal of polarity. The center of the island shows stellate reticulum like cells [Figure 4 inset]. The connective tissue also shows the presence of extensive coarse granular eosinophilic cells distributed throughout the section [Figure 5]. Most of the odontogenic islands also reveals such granular cells within them [Figure 5]. The central stellate cells may be replaced by large eosinophilic rounded or polyhedral granular cells [Figure 6]. The granular cells may take up a complete epithelial island, and then even the basal cells are granular. Histopathological differential diagnosis includes granular cell myoblastoma and granular cell ameloblastic fibromas. However, based on the clinical, radiographic and histopathology, a final diagnosis of granular cell ameloblastoma is arrived.

DISCUSSION

The granular cell ameloblastoma is one of the rarest entities and accounts for about 5% of all ameloblastomas.^[2] Histopathologically, it is characterized by having numerous large eosinophilic granular cells.

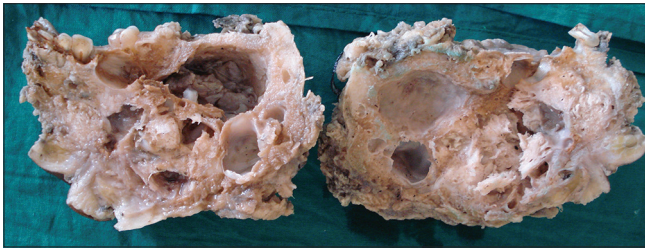


Figure 1: Macroscopic appearance of the specimen revealing the multilocular cyst like appearance of the lesion exhibiting compartmented appearance



Figure 3: Radiographic appearance of the jaw eliciting the multilocular radiolucency with root resorption

These cells usually form the central mass of the epithelial tumor islands and cords. Sometimes, even peripheral cells also exhibit similar features which are usually nongranular cells.^[4]

Granular cells change in classic ameloblastoma is well-recognized phenomenon.^[3] It was first seen by Krompecher in 1918 and was called pseudoxanthomatous cells. Whether granular cell change in ameloblastoma is a degenerative process or a harbinger of more aggressive course is a matter of debate.^[5] Granular cells are transitional or matured phase in the lifecycle of ameloblastoma, initiating its process from normal stellate reticulum like cells to the production of granules and finally leading to degeneration and formation of cystic areas.^[6] The granular cells acquire small pyknotic nuclei and bulky cytoplasm

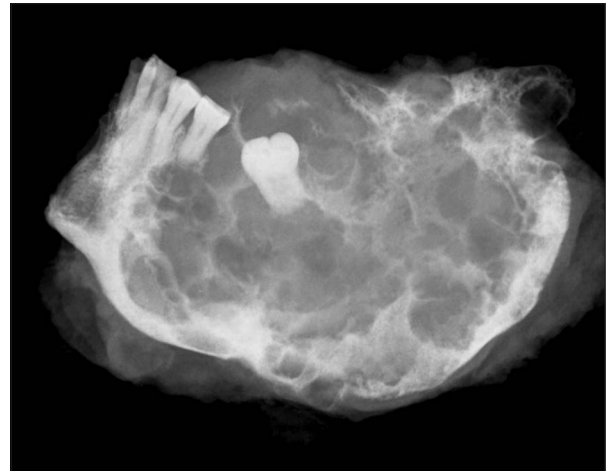


Figure 2: Specimen radiographic appearance of the jaw eliciting the multilocular radiolucency with root resorption postero-anteriorly

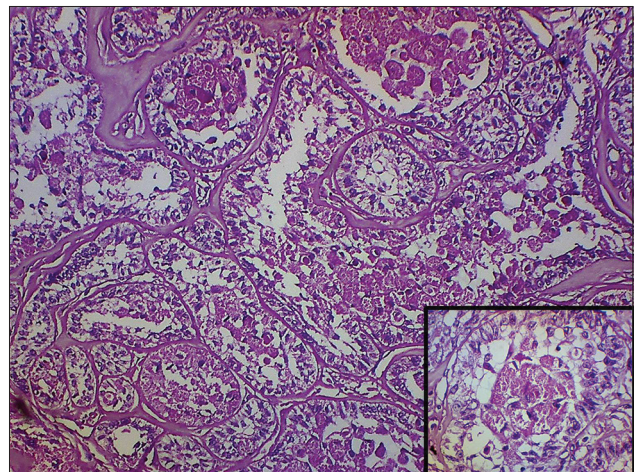


Figure 4: The microscopic appearance exhibiting fibrous connective tissue exhibiting numerous odontogenic epithelial islands with peripheral tall columnar cells showing reversal of polarity. The center of the island shows stellate reticulum like cells which is replaced by granular cells (inset) (under $\times 10$ magnification)

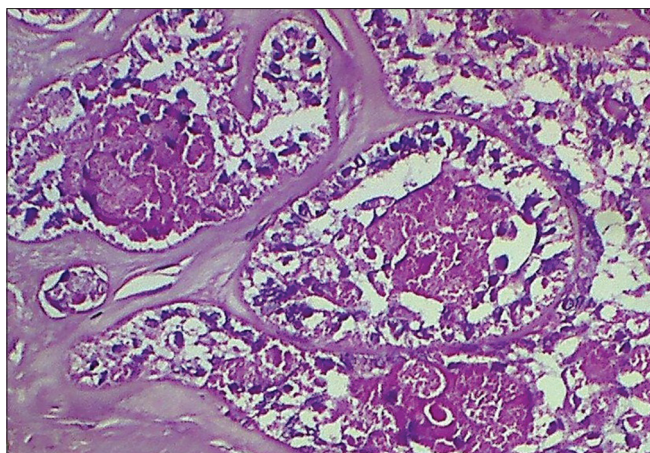


Figure 5: The microscopic appearance shows central stellate cells replaced by large eosinophilic rounded or polyhedral granular cells (under $\times 10$ magnification)

filled with coarse eosinophilic granules indicating there is an apoptotic process taking place.^[7]

Nature of granular cells in ameloblastoma has been explained by various theories. Granular cells are epithelial in origin and several ultrastructural and histochemical studies have described them as lysosomes.^[6] Lysosomal aggregation within the cytoplasm is caused by dysfunction of either a lysosomal enzyme or lysosomal-associated protein involved in the enzyme activation, enzyme targeting, or lysosomal biogenesis.^[8]

Granular appearance of the tumor cells is seen in various oral tumors, such as granular cell myoblastoma, congenital epulis and granular cell ameloblastic fibromas. The morphology of granular cells is similar, but their origins are different. According to histogenesis, granular cell ameloblastoma is epithelial, while others appear to be of mesenchymal origin.^[9]

Granular cell ameloblastoma is locally aggressive and has a relatively high chance of recurrence. The recurrence rate for a granular cell ameloblastoma was reported to be 33.3% which is higher as compared to that of follicular, plexiform and acanthomatous subtypes.^[3]

CONCLUSION

Granular cell ameloblastoma is an aggressive lesion with a marked proclivity for recurrence. Appropriate surgical

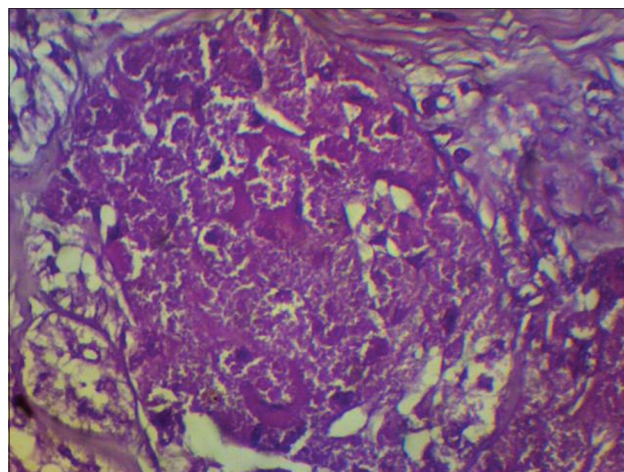


Figure 6: The microscopic appearance of granular cell (under $\times 40$ magnification)

measures have to be taken at the first instance since it has a tendency for metastasis.

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Conflicts of interest

There are no conflicts of interest.

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