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CASE REPORT

Granular Cell Ameloblastoma of Maxilla: Masquerading as Pyogenic Granuloma

[1]Nishat Afroz, [2]Shagufta Qadri, [3]Nida Shamim, [4]Shakeba Qadri

ABSTRACT

Ameloblastoma is a slow growing odontogenic epithelial tumor of the jaw, however maxillary ameloblastoma is a rare histopathological entity. This tumor arises from odontogenic epithelium of embryonal tooth element and is most frequent in the mandibular molar region, with only 20% occurring in the maxilla. Of the various histological patterns of ameloblastoma, the granular cell type is extremely rare, accounting for 4% of all reported cases of ameloblastomas. We report a rare case of multicystic granular cell ameloblastoma of maxilla in a 15 years old male patient who presented clinically as slow growing painful swelling on left side of the face. Histological examination of the excised tumor revealed islands comprising of cells with extensive granular cell transformation, surrounded by peripheral ameloblast-like cells. Immunohistochemistry showed positivity for pancytokeratin while negative staining was seen for S-100 and other neural and muscle markers, thereby confirming the diagnosis of granular cell ameloblastoma.

Keywords: Ameloblastoma, Granular cell, Pyogenic granuloma, Stellate reticulum.

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INTRODUCTION

Ameloblastoma is the second most common odontogenic tumor, accounting for 1% of all tumors and cysts arising in the maxilla and mandible.[1] Most cases are diagnosed between 30 and 60 years of age,[2] without any gender predilection. Ameloblastomas, in general are considered benign neoplasm with the tendency of local invasiveness, however the maxillary ameloblastoma is a more aggressive lesion, presumably because the thin and fragile bone of the maxilla allows a relatively unimpeded spread of the tumor to the adjacent structures including the maxillary sinus, nasal cavity and the orbit. Granular cell ameloblastoma (GCA) is an extremely rare variant of ameloblastoma characterized by having numerous large eosinophilic granular cells,[3] hence showing a close histological resemblance to a granular cell neoplasm.

This case report is being presented here because GCA arising in maxilla is an extremely rare condition having a unique histopathological and immunohistochemical findings and hence it should be differentiated from the other variants of ameloblastoma and also from other granular cell lesions because of its high recurrence rate.

CASE REPORT

A 15-year-old male patient reported to the department of maxillofacial surgery with the complaint of an intraoral swelling arising in his upper gingiva, which was painful. He had first noticed this swelling 3 months back. On extraoral examination, the swelling was tender, measuring 4 × 3 cm seen in the left anterior maxillary region, fixed to the underlying soft tissue and bone. Intraorally, there was a firm, nodular, swelling 3 × 2 cm creamish-grey in color with hemorrhagic areas, extruding in the upper gingival region extending from upper left incisor to the left upper canine (Fig. 1), obliterating the buccal vestibule. The swelling was tender, and bleed with touch. Routine hematological, urine examination and biochemical tests of the patient were normal. The orthopantomograph (OPG) was done as an initial diagnostic approach, which revealed a multilocular radiolucency. FNA was advised and the smears were suggestive of a pyogenic granuloma. Subsequently, incisional biopsy followed by partial maxillectomy was done and postsurgery rehabilitation of maxillary defects was achieved by the free tissue transfers along with the bone grafting.

On gross examination of the resected specimen, the tumor appeared encapsulated gray white nodular mass measuring 4 × 3 × 3 cm. Cut section showed solid and cystic areas, with foci of hemorrhage and necrosis (Fig. 2). Entire tissue was processed with the inclusion of margins. Microscopic examination of the paraffin embedded, hematoxylin and eosin (H&E) stained sections revealed an encapsulated tumor of odontogenic origin. Sections showed large ameloblastic follicles lined by preameloblast like cells, showing peripheral palisading

[1]Associate Professor, [2,3]Senior Resident, [4]Junior Resident

[1-3]Department of Pathology, Jawaharlal Nehru Medical College Aligarh Muslim University, Aligarh, Uttar Pradesh, India

[4]Department of Dentistry, Ziauddin Ahmad Dental College Aligarh Muslim University, Aligarh, Uttar Pradesh, India

Corresponding Author: Shagufta Qadri, Senior Resident Department of Pathology, Jawaharlal Nehru Medical College Aligarh Muslim University, Aligarh 202002, Uttar Pradesh, India Phone: +91-9897532879, e-mail: qadridrshagufta@gmail.com

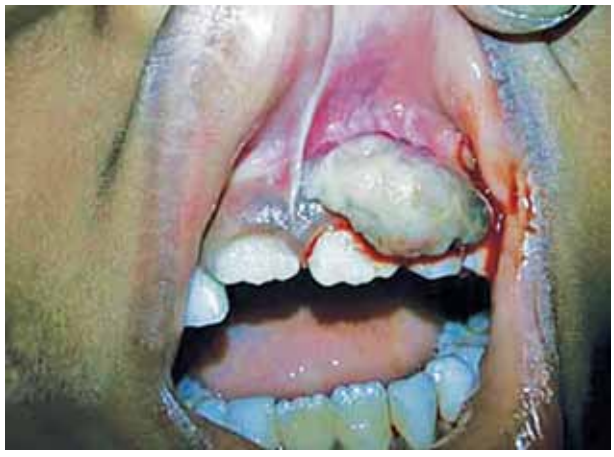

Fig. 1: Firm, nodular, swelling of 3 × 2 cm, creamish-grey in color with hemorrhagic areas, extruding in the upper gingival region

Fig. 2: Cut section of resected tumor showing solid and cystic areas with foci of hemorrhage and necrosis

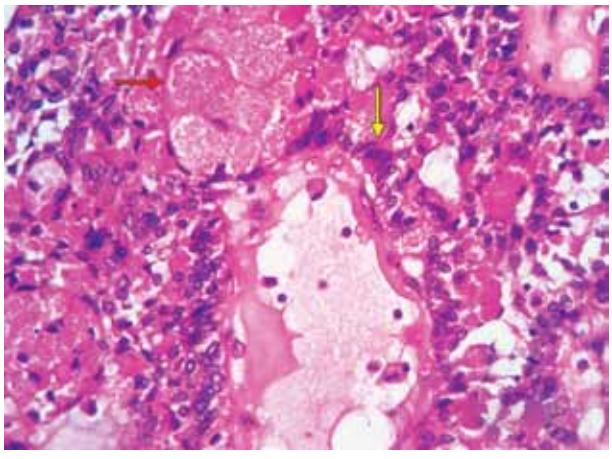

Fig. 3: Follicles lined by pre-ameloblast like cells (yellow arrow), showing peripheral palisading and reversal of nuclear polarity. Follicular center showing cystic degeneration and replacement of stellate reticulum like cells by large granular cells (red arrow) (H&E, 100×)

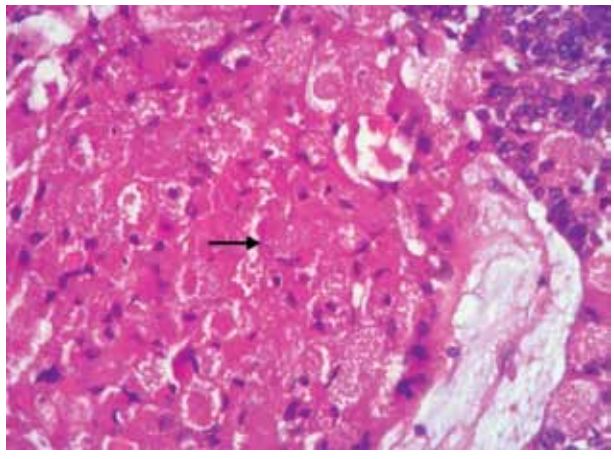

Fig. 4: Island of large granular cells (black arrow) with round to polyhedral outline and pyknotic nuclei displaced to the periphery, cytoplasm distended with coarse eosinophilic granules (H&E, 400×)

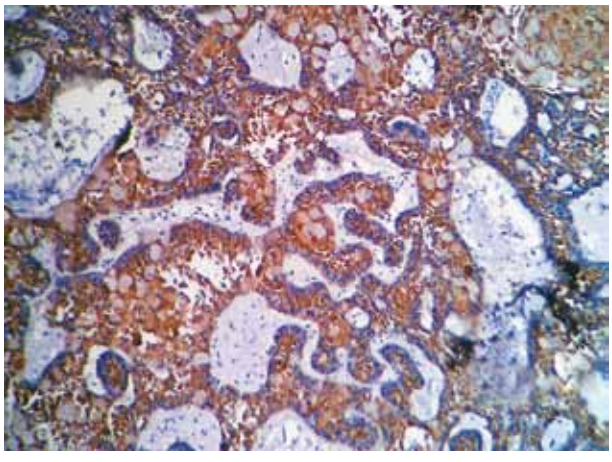

Fig. 5: Granular cells showing membranous and nongranular cells showing diffuse cytoplasmic positivity with pancytokeratin (IHC Pancytokeratin, 100×)

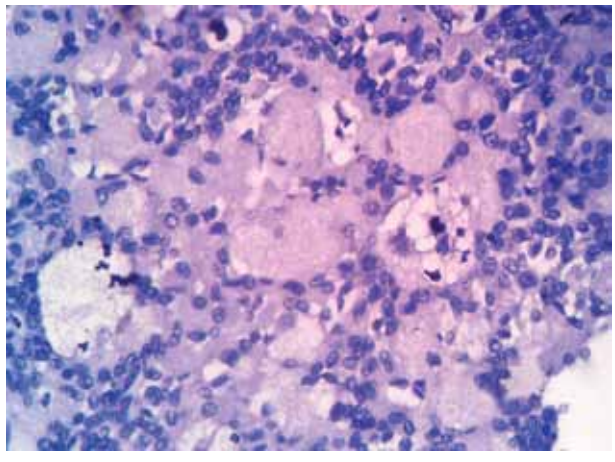

Fig. 6: Ameloblastic and granular cells showing negative immunostaining for S-100 (IHC S-100, 400×)

and reversal of nuclear polarity (nuclei oriented away from the basement membrane). The center of the follicles exhibited cystic degeneration as well as replacement of stellate reticulum like cells by large granular cells (Fig. 3). The granular cells were large with round to polyhedral outline and pyknotic nuclei displaced to the periphery of the cells; prominent coarse eosinophilic granules were seen distending the cytoplasm (Fig. 4). Foci of calcification, dense inflammatory infiltrates and congested newly formed capillaries were seen in the fibrous connective

stroma. Diagnosis of granular cell ameloblastoma of maxilla was rendered on histopathology.

Immunohistochemistry (IHC) for pancytokeratin revealed membranous positivity of granular cells while nongranular cells showed diffuse cytoplasmic positivity (Fig. 5). Negative staining was seen for S-100 (Fig. 6), vimentin and other neural and muscle markers in both the type of cells, thereby confirming the diagnosis of granular cell ameloblastoma.

The postoperative period was uneventful. Patient is doing well and is kept on a prolong follow-up because of high recurrence rate of granular cell ameloblastoma of maxilla.

DISCUSSION

Ameloblastoma is a true benign neoplasm arising from enamel organ forming tissue that fails to undergo differentiation to the point of enamel formation. Tumor cells usually originate from one of the following sources: (i) epithelial lining of odontogenic cyst, (ii) dental lamina or enamel organ, (iii) disturbances of developing enamel organ, (iv) basal cells of surface epithelium, or (v) heterotopic epithelium of other parts of the body.[4] Broca was first to report this distinct neoplasm in the scientific literature in 1868.[5] Ameloblastoma is the most common odontogenic tumor, accounting for 1% of all tumors in the head and neck region and approximately 11% of all odontogenic tumors.[1] Radiologically, ameloblastomas present as unilocular or multilocular translucencies. Microscopically various patterns have been described (follicular, plexiform, acanthomatous, papilliferous-keratotic, granular cell type, desmoplastic, vascular, and dentinoameloblastoma). Out of these follicular and plexiform variants are more commonly encountered accounting for 32.5 and 28.2% respectively while acanthomatous subtype follow them by 12.1% and desmoplastic type by only 4 to 13%. Less common histopathological variants include granular cell 3 to 5% and basal cell accounting for 2%.[6] It is generally speculated that only 20% of ameloblastomas occur in the maxilla,[7] although some reports indicate an incidence as low as 1% in the maxilla.

The granular cell ameloblastoma is an unusual variant of ameloblastoma showing marked transformation in the cytoplasm of the tumor cells, which are usually stellate reticulum-like cells. They can be in pure form or as an admixture with other histologic patterns. This transformation was first noticed by Krompecher in 1918 and was called as pseudoxanthomatous cells.[4] The transformed cells possess very coarse, granular, eosinophilic cytoplasm.[8] The defining characteristic of granular cell ameloblastoma is the presence of granular cells in the central portion of the epithelial islands, strands and cords. Occasionally, granular cell change also affects the peripheral columnar cells.[9] Ultrastructural and immunohistochemical studies of cytoplasmic granules of tumor cells have confirmed that these are rather lysosomal aggregates.[6] Lysosomal aggregation within the cytoplasm is due to dysfunction of either a lysosomal enzyme or lysosome-associated protein that are responsible for the enzyme activation, enzyme targeting or involved in the lysosomal biogenesis. These defects lead to the accumulation of substrate that normally undergoes degradation in the endosome-lysosome system.[10] On immunohistochemistry granular cells exhibit a membranous positivity with cytokeratins while the nongranular cells of the same tumor shows a diffuse cytoplasmic reactivity. Moreover, granular cells are also seen to show marked cytoplasmic positivity with CD68 antibody while a consistent negative staining is seen for human mitochondria and S-100 protein.[11]

The differential diagnosis of granular cell ameloblastoma (GCA) includes other oral lesions with similar histomorphology, such as (i) granular cell odontogenic tumor, (ii) granular cell tumor (iii) congenital epulis,[11] (iv) granular cell myoblastoma and (v) granular cell ameloblastic fibromas. The granular cells morphology is similar in all these tumors, but they differ in their histogenesis, as GCA is of epithelial, while the others appear to be of mesenchymal origin.[1] Immunohistochemical analysis is a helpful tool in distinguishing GCA from these tumors.

Granular cell ameloblastoma is locally aggressive tumor with a relatively high recurrence rate being 33.3%, which is higher as compared to the more common follicular, plexiform and acanthomatous subtypes.[6] It has also been postulated that GCA has a closer relationship with malignant transformation.[12] Hertog et al proposed annual follow-up of patients with maxillary ameloblastoma up to a period of at least 10 years after radical surgery or en block resection, while only 5 years for other solid ameloblastomas.[1]

CONCLUSION

- Maxillary ameloblastomas are predominantly painless and slow growing tumors, having tendency to invade the adjacent structures because they lack a thick cortical plate, have plentiful cancellous bone and the proximity to the nasal cavity, nasopharynx, paranasal sinuses, orbits and skull base.
- Delay in the recognition of the maxillary ameloblastoma extending into these structures may prove fatal in some cases, which can be avoided only by early stage diagnosis and management.
- Granular cell ameloblastoma (GCA) occurring in maxilla is an extremely rare entity. Granular cell transformation in ameloblastomas probably occurs

as a consequence of extensive molecular changes occurring in central stellate reticulum-like cells.

- Close follow-up of patients with GCA is mandatory due to their aggressive behavior, high recurrence rate and anticipated malignant transformation and metastasis.

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