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Case Report

Dentistry Section

Granular Cell Ameloblastoma: A Rare Case Report and Literature Review

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ABSTRACT

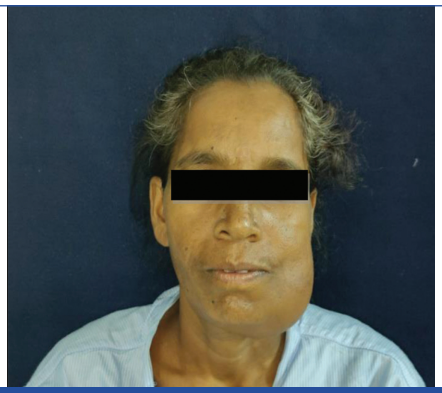
Ameloblastomas are rare, benign, aggressive neoplasms of odontogenic epithelial origin. Among the various types of ameloblastomas, granular cell ameloblastoma is rare and accounts for less than 5% of the total. Granular cells are epithelial in origin. The distinct histopathological characteristics of granular cells are associated with high lysosomal content in tumour cells cytoplasm. A 43-year-old female patient presented with a painful swelling on the left-side of the jaw. The diagnosis was based on clinical, radiological and histopathological examination. A complete surgical resection was done. Histopathology showed features of granular cell ameloblastoma. The operative procedure was uneventful. Granular cell ameloblastoma has a high tendency for recurrence. Around 33.3% recurrence is reported, according to various studies. This rate is much higher than other subtypes of ameloblastoma, which are more common. Hence, it is essential to provide appropriate surgical treatment on time to prevent tumour recurrence and metastasis.This case report aims to highlight the unique features of granular cell ameloblastoma, distinguishing it from other subtypes and discussing the pathogenesis and treatment modalities.

Keywords: Local, Lysosomes, Neoplasm recurrence

CASE REPORT

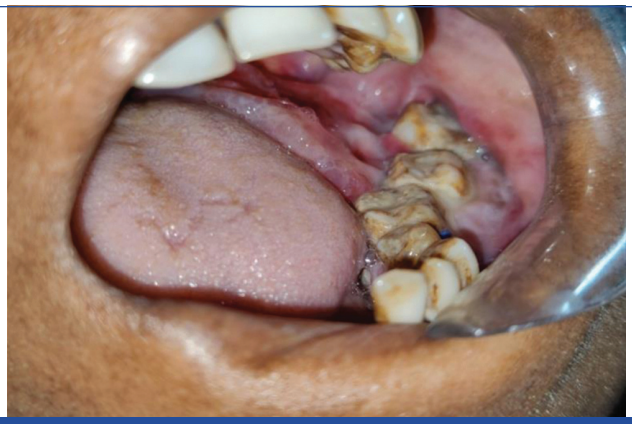
A 43-year-old female patient reported with a painful swelling over the left lower back region of the jaw of 3.5 years duration. The associated pain was gradual in onset, dull aching, continuous, and radiating to left-side of forehead region. Pain aggravated on mastication and relieved on its own over time. The swelling was initially small in size and increased to its present size of 10×9 cm. There was history of decreased salivation, change in consistency of saliva from thin to thick and ropy since three days. Patient gave history of difficulty in mastication since two months. There was no significant past medical or dental history.

On extraoral examination, the face appeared grossly asymmetrical due to swelling on the left-side extending anteroposteriorly from the left corner of the mouth to the left angle of the mandible and supero-inferiorly from 3 cm below the left lateral canthus of the eye to 2.5 cm below the inferior border of the mandible. On palpation, tenderness was present, and the swelling was firm and fixed to underlying structures [Table/Fig-1].



[Table/Fig-1]: Clinical photograph of the patient showing swelling over left side of the jaw.

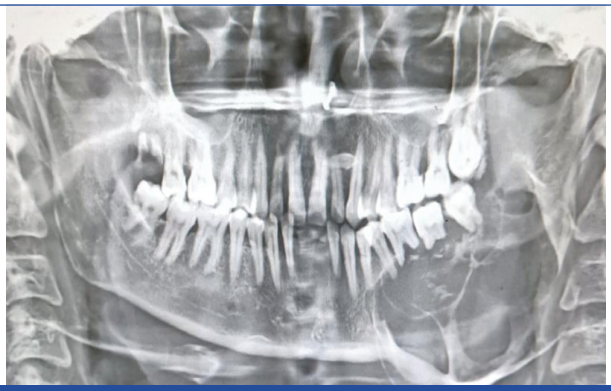
Intraoral examination revealed an ulcerative lesion of 5×4 cm present over the lower left-side of the alveolus extending from 37 anteriorly, and the posterior extent could not be assessed [Table/Fig-2]. Superoinferiorly, the swelling extended from the level of occlusion of upper teeth to the depth of the lower left gingivobuccal sulcus involving the floor of the mouth.



[Table/Fig-2]: Ulcerative lesion on lower left alveolus.

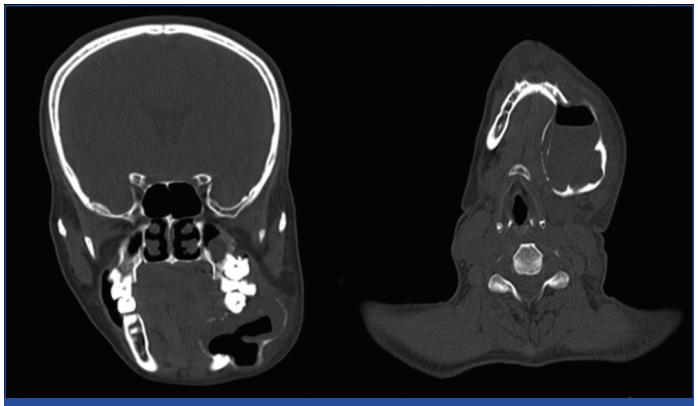
A provisional diagnosis was made of odontogenic tumour of left body of mandible. A differential diagnosis of dentigerous cyst, odontogenic keratocyst, fibrous dysplasia, odontogenic myxoma, osteosarcoma and aneurysmal bone cyst was also considered. The tumour was not aspirated and the patient was advised radiological investigation for further evaluation.

On radiological examination, a multicystic radiolucent lesion was seen in the left mandibular premolar and molar region involving half of the ascending ramus [Table/Fig-3]. Lobulation of the inferior cortex was seen at the angle of the mandible. Computed dental



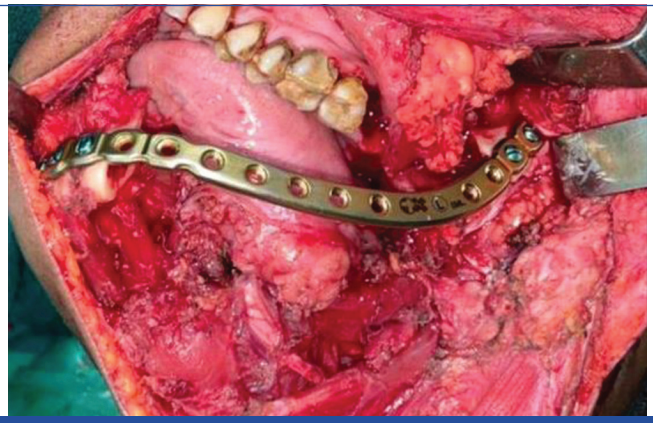
[Table/Fig-3]: Panoramic radiograph of the lesion.

tomography [Table/Fig-4] showed a large multicystic non enhancing expansile lesion with few internal enhancing areas of the solid component in the left lower body and ramus of mandible causing the cortical break.

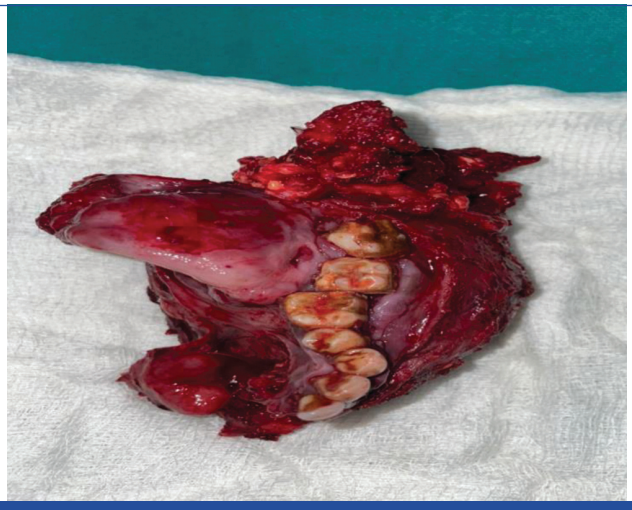


[Table/Fig-4]: Computed Tomography (CT) images.

Enbloc resection of the lesion was done along with segmental mandibulectomy and disarticulation of the condyle of the left-side. Reconstruction was done with Recon plate [Table/Fig-5]. The resected specimen was sent for histopathology [Table/Fig-6].

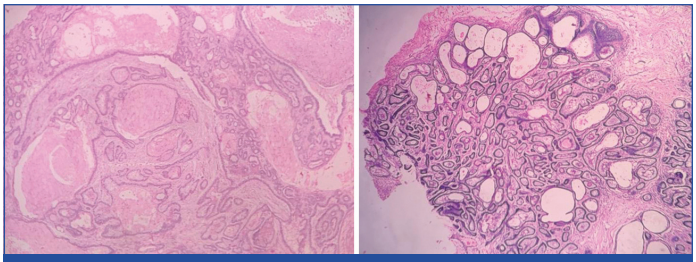


[Table/Fig-5]: Reconstruction done with Recon plate.

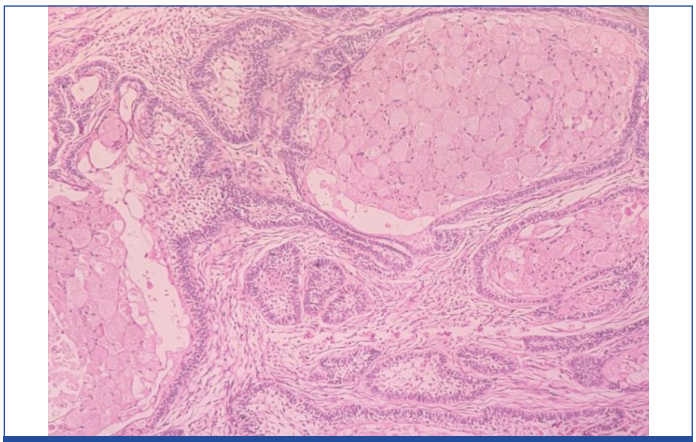


[Table/Fig-6]: Resected specimen (Enbloc resection).

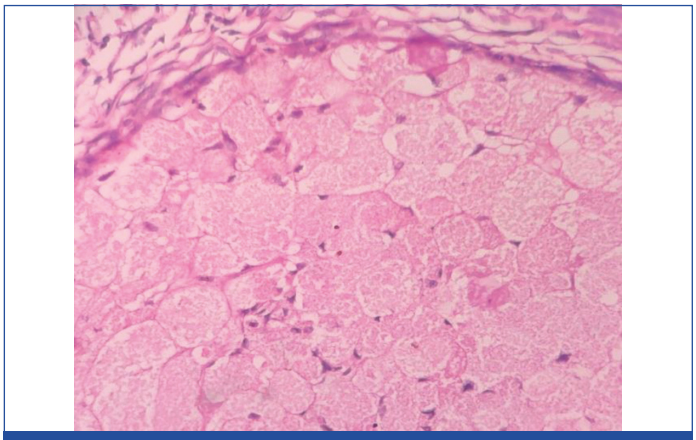
The Haematoxylin & Eosin (H&E) stained tissue section showed odontogenic epithelial islands in interspersed fibrocellular connective tissue stroma [Table/Fig-7,8]. The odontogenic epithelial islands were arranged in follicles lined by tall columnar cells with hyperchromatic nuclei and a palisading pattern [Table/Fig-9]. The suprabasal cells resembled the stellate reticulum. In some odontogenic follicles, spherical-shaped cells filled with eosinophilic granules were seen with eccentric nuclei suggestive of metaplasia [Table/Fig-10]. Based on the histopathological report, it was diagnosed as Granular cell ameloblastoma.



[Table/Fig-7,8]: H&E stained section (4X) showing odontogenic epithelial islands/ follicles in connective tissue. (Images from left to right)



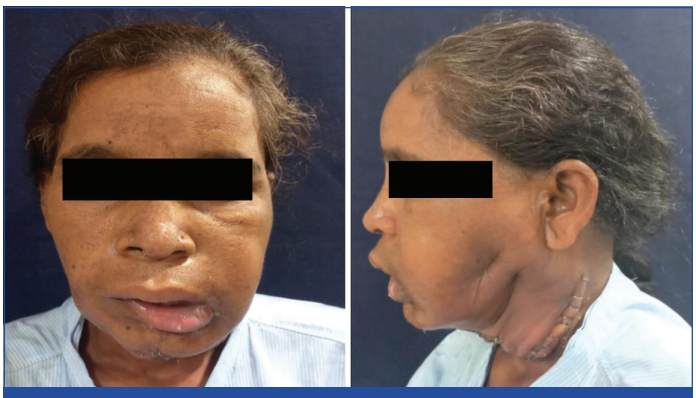
[Table/Fig-9]: H&E stained section (10X) showing follicles lined by tall columnar cells with polarised hyperchromatic nuclei suggestive of odontogenic origin. Metaplasia of central stellate reticulum like cells into granular cells.



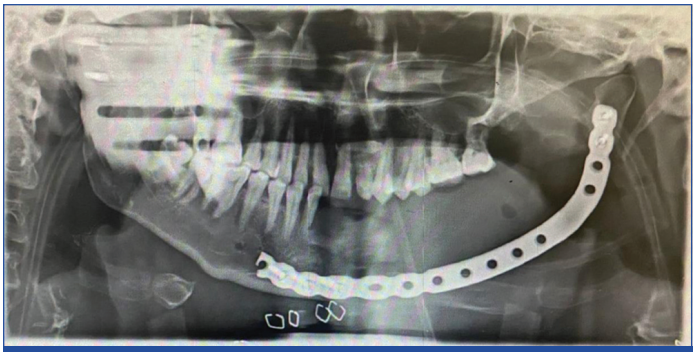
[Table/Fig-10]: H&E stained section (40X) showing large spherical cells filled with eosinophilic granules with eccentric nuclei in odontogenic follicles.

The final diagnosis was made on the basis of routine histopathological examination done by H&E staining procedure. Based on histopathological examination, it was a clear case of granular cell ameloblastoma, so no immunohistochemistry studies were performed on the specimen.

The post-surgical course was uneventful and no recurrence was seen up to 6 months of follow-up period [Table/Figure 11-13].



[Table/Fig-11,12]: Postoperative pictures of the patient (front and side profile). (Images from left to right)



[Table/Fig-13]: Panoramic radiograph showing reconstruction plate after.

DISCUSSION

Ameloblastoma is a benign neoplasm of the jaw of odontogenic epithelial origin. It is a slow-growing, locally invasive tumour that closely resembles enamel organ epithelium [1]. Ameloblastoma occurs more commonly in 3rd to 5th decade of life with no gender predilection [2]. The global prevalence of ameloblastoma is 0.5 cases per million persons per year according to a study done by Brown NA and Betz BL [3]. In Maharashtra (India), ameloblastoma showed an incidence rate of 35.43% in a review of 127 cases of odontogenic tumours [4].

It is most commonly seen near the angle of the mandible [5]. Ameloblastoma is clinically identified as a painless, slow-growing swelling in the jaw. Facial deformity, malocclusion, tooth loss, pain and paresthesia may be seen on the affected side [6]. Ameloblastoma presents as a significant hard tissue swelling in the jaw bone. Sometimes, it may even be present as soft tissue swelling in the maxilla or mandible with no involvement of the underlying bone [7]. This type is known as peripheral ameloblastoma, and it can often be confused with intraosseous ameloblastoma. However, intraosseous ameloblastoma spreads from inside the jaw bone into the gingiva. Past studies have also described these lesions as odontogenic gingival epithelial hamartoma [8]. Solid or multicystic ameloblastoma typically shows a multilocular radiolucency with well-defined borders on radiographs. Conventional ameloblastoma may also indicate a unilocular radiolucency commonly seen in unicystic ameloblastoma. Cortical bone's perforation, expansion, and root resorption/divergence in adjacent teeth are widely seen [5].

Among the many known histopathologic subtypes of ameloblastomas, the most common patterns are follicular and plexiform [9]. The less common subtypes include the granular cell ameloblastoma, acanthomatous, basal cell, and desmoplastic ameloblastoma.

Granular cell ameloblastoma is a distinct subtype. Granular cell ameloblastoma represents 1-5% of all ameloblastomas [10]. It is identified by groups of granular cells with profuse cytoplasm. These granular cells resemble lysosomes and create a solid mass in the centre comprising of epithelial cords and islands [11]. Granular cell ameloblastoma is aggressive in nature, with an increased rate of recurrence and can even progress into metastasis [12]. Granular cell ameloblastoma does not differ from other subtypes when it comes to radiological or biological behaviour. Histopathology is the only way of differentiating granular cell ameloblastoma from other common subtypes.

Granular cell ameloblastoma is a rare form of ameloblastoma. The treatment and prognosis of granular cell ameloblastoma does not vary when compared to the common subtypes. Although the rate of recurrence of granular cell ameloblastoma is quite high as compared to the other subtypes [5]. Kameyama Y et al., in their research, classified 1 out of a total of 77 cases as granular cell ameloblastoma [13]. Reichart PA et al., reported and discussed the literature on ameloblastoma (jaws) from the year 1960 to 1993 [10]. The author stated that only 56 (3.5%) cases out of 1593 cases were of granular subtype and also suggested that the age group for the granular cell subtype is most common to the other subtypes, for which an average age of 35-year-old is reported, ranging from 4 to 92 years [10]. In another study, Hartman KS did a review of twenty cases of granular cell ameloblastoma from the files of Armed Forces Institute of Pathology (AFIP) [14]. The author noted a mean age of 40.7 years (ranging from twenty-one to sixty-five years). Forty percent of the cases were seen in non Caucasians with no specific gender preference [14]. Most tumours (19 out of 20) affected the posterior region of the mandible [14]. A strong preference for mandible was confirmed by subsequent reviews [15]. The most common symptoms reported were jaw swelling and pain. No distinct radiographic findings have been reported in comparison to other subtypes.

Immunohistochemistry studies were done by Kumamoto H and Ooya K on six cases of granular cell ameloblastoma. They revealed that increased apoptotic cell death and associated phagocytosis might cause granularity in ameloblastoma [16]. Granular cell ameloblastoma showed a similar biological behaviour as other histological subtypes. It is aggressive locally and shows a high recurrence rate [17]. A 33.3% recurrence rate for granular cell ameloblastoma was reported by Reichart PA et al., which was higher as compared to other common subtypes [10]. In the study conducted by Hartman KS, 73% (11 of 15 patients) of recurrent lesions were reported [14]. The prognosis of the disease depends on the type of treatment/surgery. In granular cell ameloblastomas, enucleation or curettage is the choice of treatment which generally results in recurrence. The main reason behind this is the presence of the tumour border within bone (cancellous), which is present beyond the evident macroscopic surface and the radiographic borders of the lesion. Therefore radical surgery is often suggested for treatment [17].

Interestingly, granular cell ameloblastomas seldom show a malignant pattern leading to metastasis [15]. The main histological features of granular cell ameloblastoma are granular cells, typically present in the centre of the tumour, which gradually replaces the stellate reticulum cells. Initially, they were believed to portray ageing or degeneration [18,19]. Still, recent Immunohistochemical (IHC) studies have suggested that it is related to increased apoptotic cell death of the lesional cells and phagocytosis of neighbouring neoplastic cells [16]. The differential diagnosis of granular cell ameloblastomas includes granular cell odontogenic tumours, granular cell tumours and congenital epulis. These lesions show biologically distinct behaviours and should be distinguished from granular cell ameloblastomas [20].

CONCLUSION(S)

Granular cell ameloblastoma is an infrequent variant of ameloblastoma showing distinct histologic and immunohistochemical features. The prognosis and treatment are similar to other common subtypes of solid or multicystic ameloblastoma. Granular cell ameloblastoma should be distinguished from other lesions with granular cells mainly due to its high risk of recurrence. A better knowledge of the molecular pathogenesis of ameloblastoma and its various subtypes may provide diagnostic and therapeutic benefits.

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