GRANULAR CELL AMELOBLASTOMA- A CASE REPORT

Sonal Saigal [1] Ankur Bhargava [1] Monali Chalishazar [2]

[1] *Senior lecturer, Department Of Oral Pathology, Government Dental College, Raipur (Chhattisgarh), India.*
[2] *Professor and Head, Department of Oral Pathology, Ahamdabad Dental College and Hospital, Ahamdabad, Gujarat*

Corresponding Author : Ankur Bhargava , 291- A Block, Chitrakut Nagar, Bhuwana Extension, Udaipur (Rajasthan), Ph: 08959242326, E-mail: sonal_ankur@rediffmail.com

Abstract
Ameloblastoma is an epithelial odontogenic tumour of the jaw and exhibits diverse microscopic patterns which occurs either singly or in combination with other patterns. Granular cells can occur in various odontogenic and non-odontogenic tumours. Granular cell ameloblastoma is a rare variant of ameloblastoma as making up 1 to 5% of all ameloblastomas that histopathologically has numerous large granular cells. The "granular change" is thought to be due to a dysfunctional status of neoplastic cells, and the pathogenesis of this tumour seems to be age-related. This article describes a case of granular cell ameloblastoma affecting a 60-year-old woman. The lesion was located in the right side of the mandible. The tumour was excised and no recurrence has been observed after one year of surgery.

Keywords : Granular cell ameloblastoma (GCA), Granular, Neoplastic cells.

Introduction

Odontogenic tumors are lesions originating from epithelial and/or mesenchymal components of the tooth-forming apparatus. They are unique to the jaws and if left untreated, often lead to extensive tissue destruction and deformity. Ameloblastoma is the most frequent and enigmatic odontogenic tumor [1,2]. The terms "ameloblastoma" has derived from (Old French, amel, enamel, plus Greek, blastos, germ) and "adamantinoma" (Greek, adamos, hard substance) are both misnomers for this tumor because ameloblasts that elaborate enamel need not be present, and since enamel is not produced, the tumor is not hard this tumor may arise from one of many dental sources: the epithelial lining of a dentigerous cyst; the remnants of the dental lamina or of the enamel organ [3].

Ameloblastomas consist of epithelial strands or islands. The former pattern is called plexiform, the latter follicular. Peripheral cells are columnar, while cells lying more centrally are fusiform to polyhedral and are loosely connected to each other. In the follicular type, in particular, an increase in intercellular oedema may cause cysts that coalesce to form large cavities. In the plexiform type, cavity formation arises through stromal degeneration. The tumour infiltrates the adjacent cancellous bone, whereas cortical bone and periosteum usually expand but will not be perforated. Spread into soft tissues is highly unusual. Acanthomatous, granular cell and basal cell (basaloid) ameloblastoma are variants with squamous metaplasia, granular cells and basaloid cells, respectively [4]. The most common odontogenic tumour with a granular cell component is the GCA, making up 1 to 5% of all ameloblastomas [5].

Histopathologically GCA has numerous large eosinophilic granular cells. These cells usually form the central mass of the epithelial tumor islands and cords. The periphery of the islands consists of non-granular tall columnar cells. The GCA is rich in lysosomal granules. In which there is marked transformation of the cytoplasm, usually of stellate reticulum like cells, so that it takes a very coarse, granular, eosinophilic appearance. Granular cell variety of ameloblastoma appears to be an aggressive lesion with a marked

proclivity for recurrence. In addition several cases of this type have been reported, as metastasizing [6].

Case Report

A 60-year-old female reported to our hospital with a history of swelling over right side of face for past 5–6 months. History of slowly progressing intraoral swelling of right buccal space with no apparent distress. She had moderate right facial swelling but no facial paralysis, paresthesia, or anesthesia and no palpable regional lymphadenopathy.

Extra oral examination revealed

Facial asymmetry with well defined swelling on the right side of face. The swelling extended from corner of mouth to the angle of mandible. Colour of skin over the swelling was normal, with ovoid shape, measuring about 2x4 cm. No pulsation or impulse on coughing. On extraoral palpation, a firm, freely movable mass could be felt, measuring approximately 3.0 cm in diameter.

Intra oral examination revealed

Poor orodental hygiene and lesional side was edentulous. Swelling was firm, non tender and extending anterior right lower region till retromolar area (Fig. 1).

Investigations

Routine biochemical and hematological investigations were within normal limits. The orthopantomographic imaging revealed that there was an expansile multiloculated enhancing lesion with marked thinning, scalloping and lysis of buccal and lingual cortices involving the respective side (Fig. 2).

An incisional biopsy was performed under local anesthesia. The specimen was fixed in formalin, paraffin embedded and stained by hematoxylin-eosin. Microscopic features were composed of nests or organoid clusters of neoplastic cells embedded in a fibrous stroma, few follicles lined with single layer of tall columnar ameloblast like cells (reversed polarity of nucleus) surrounding a central core of loosely arranged angular cells resembling stellate reticulum or enamel organ with few microcyst formation within them (Fig. 3). Numerous epithelial islands consisting of granular cells were evident which were occurring in small groups or large islands, showed cell outlines, usually clear, but varied considerably both in size and shape(Fig. 4). These findings were consistent with the diagnosis of GCA.

On the basis of above diagnosis patient was planned for surgery. General anesthesia was secured with nasal endotracheal tube and all aseptic protocol was followed. Right composite hemimandibulecomy was performed. Reconstruction was carried out with 2.5 mm stainless steel reconstruction plate and advancement local flaps. After one year follow up patient is doing well.

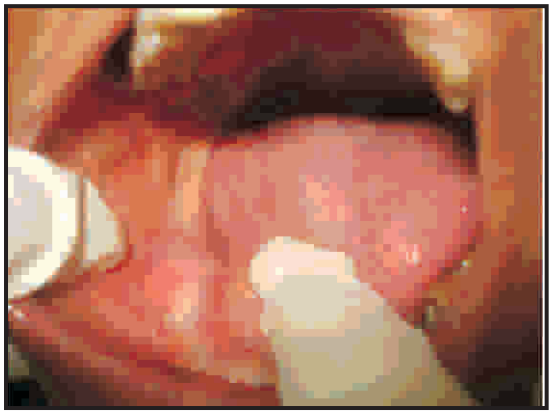

Fig1:Photomicrograph showing intra oral swelling extending from anterior right lower region to retromolar area.

Fig2: Orthopantomographic imaging revealed an expansile multiloculated enhancing lesion with marked thinning, scalloping and lysis of buccal and lingual cortices on right mandibular region.

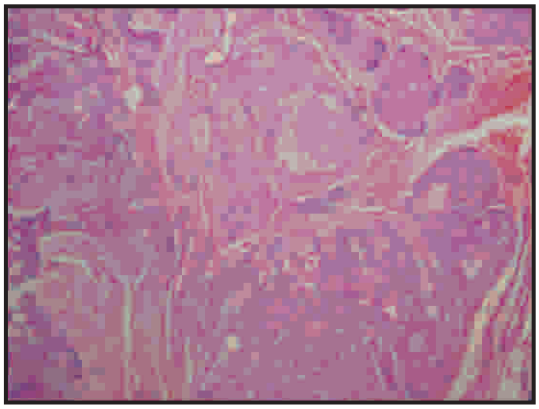

Fig3: Photomicrograph showing numerous follicles lined with single layer of tall columnar ameloblast like cells surrounding a central core of loosely arranged angular cells resembling stellate reticulum (H & E)

Fig4: Photomicrographs showing tumour islands with a peripheral layer of ameloblasts and extensive central areas of granular cells with large, eosinophilic cytoplasmic granules (H & E).

Disscussion

The cases of classical ameloblastoma with only focal presence of granular cells have to be differentiated from the pure variant of GCA, in which the neoplastic granular cells are diffusely present and predominate. In this case, the granular cells occur in all the tumour follicles, replacing completely or in part the stellate reticulum like cells.

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[7]. Of the odontogenic tumors, granular cells have been described in the GCA and the granular cell ameloblastic fibroma[8]. 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 coincidential mixture of ameloblastoma and granular cell myoblastoma[9]. 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.

During normal amelogenesis, ameloblasts show an increase in autophagic lysosomes between the secretory and absorptive stages and from reduced ameloblasts to squamous epithelium[10]. Thus, the odontogenic epithelium seems to undergo granular changes under certain conditions[11]. It is currently thought that the granular change probably occurs as a consequence of an altered function of tumour cells, a hypothesis supported further by the finding that this tumour is age-related[12]. The granular cells may be cuboidal, columnar, or round, and the cytoplasm is filled with acidophilic granules, which have been identified ultrastructurally as lysosomal aggregates. The granularity might be caused by increased apoptotic cell death and associated phagocytosis by neighbouring neoplastic cells[13].

The biological behaviour of GCA does not seem to differ from the other histologic subtypes of ameloblastoma; it can be locally aggressive and has a relatively high chance of recurrence[14]. However the prognosis is similar to that of the classical ameloblastomas. Only one case has been described with an aggressive biological behaviour, with high recurrence rate[15]. In our case, a 1-year follow-up without clinical recurrences confirmed the reported good prognosis of the completely excised classical granular cell type of ameloblastoma.

Conclusion

Early diagnosis and prompt surgical treatment in GCA is of prime importance. Noteworthy is that GCA'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

1) *Daiana P, Stolf ACK, Abhijit GB. Genetic aspects of ameloblastoma: a brief review. Biotechnol Molecular Biol Rev 2007;2:116-22.*

2) *Kim S, Jang H, Ju K. Ameloblastoma: A clinical, radiographic, and histopathologic analysis of 71 cases. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2001;91:649-53.*

3) *Robert G, John C, Joseph EM. Ameloblastoma of the jaw- A clinical study. N Engl J Med 1963;269:126-9.*

4) *Pieter JS. Odontogenic tumour- An update. Curr Diagnos Pathol 2006;12:54–65.*

5) *Georg HR, Emmanuel AB. Granular cells in odontogenic and non-odontogenic tumours. Virchows Archiv A Pathol Anat 1989;415:403-9.*

6) *Anisha M, Yogesh S. Granular cell – Peripheral ameloblastoma: a rare variant. J Maxillofac Oral Surg 2008;3:294–7.*

7) *Reichart PA, Philipsen HP, Sonner S. Ameloblastoma: biological profile of 3677 cases. Eur J Cancer B Oral Oncol 1995;31:86-99.*

8) *Couch DR, Morris EE, Vellios, F. Granular cell ameloblastic fibroma. Report of 2 cases in adults, with observations on its similarity to congenital epulis. Amer J Clin Path 1996;37:398-404.*

9) *Navarrette AR, Smith M. Ultrastructure of granular cell ameloblastoma. Cancer 1971;27:948- 55.*

10) *Reith EJ. The stages of amelogenesis as observed in molar teeth and young rats. J Ultrastruct Res 1970;30:111-51.*

11) *Nasu M, Takagi M, Yamamoto H. Ultrastructural and histochemical studies of granular-cell ameloblastoma. J Oral Pathol 1984;13:448-56.*

12) *Muzio LL, Mignogna SS, Rosa GD. Granular Cell Ameloblastoma: a case report with histochemical findings. Orai Oncol, EurJ Cnncer 1996;32:210-2.*

13) *Peter RA, Philpsen HP. Odontogenic Tumors and Allied Lesions, 1st ed. United Kingdom:Quintessence Publication:2004.*

14) *Ghandhi D, Ayoub AF, Pogrel MA, MacDonald G, Brocklebank LM, Moos KF. Ameloblastoma: a surgeon's dilemma. J Oral Maxillofac Surg 2006;64:1010-4.*

15) *Regezi JA, Sciubba JJ. Oral Pathology: clinical pathologic correlations, 5th ed. Philadelphia:W.B. Saunders;2008.*

Source of Support - Nil

Conflict of Interest - None declared