A GIANT CYSTIC GRANULAR AMELOBLASTOMA OF THE MANDIBLE

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Abstract

Granular cell ameloblastoma is a rare odontogenic tumor, accounting for 3.5% of all ameloblastoma cases. In most instances, it occurs as an admixture with other histologic patterns, particularly the follicular subtype. It is also considered an aggressive variant of the ameloblastoma with potential recurrence and malignant transformation. A 45 year-old male patient reported with a gigantic swelling in the right region of the face since 7 years. Histopathologically, diagnosis of granular cell ameloblastoma showing cystic changes was established.

Keywords: granular cells, ameloblastoma, aggressive evolution

INTRODUCTION

Ameloblastoma (AB) is a benign, locally aggressive odontogenic tumor arising from odontogenic epithelium with mature fibrous stroma without odontogenic ectomesenchyme [1]. It occurs in three different clinical-radiographic – i.e. conventional intraosseous solid/multicystic (86%), unicystic (13%) and peripheral (1%) – patterns.

The common histological patterns include the follicular (32.5%) and plexiform (28.2%) ones, less common being the acanthomatous (12.1%), granular, desmoplastic (8.6-13%) and basal (2%) forms [2].

Granular cell ameloblastoma (GCA), a relatively rare histologic subtype, representing 3-5%, is characterised by large round/cuboidal eosionophilic cells with granular cytoplasm and, in most instances, it is blended with a follicular subtype [3].

Only few cases of cystic granular ameloblastomas are reported in the literature. The purpose of the present article is to report a case of an extremely large cystic granular ameloblastoma.

CASE PRESENTATION

A 45 year-old male with a non-contributing previous clinical history reported to outpatient department with swelling on the right side of the face since 7 years. History revealed that swelling was initially small, progressively increasing up to the present size. On clinical examination, a large well-defined non-tender swelling, extending from the middle part of the zygomatic process to the lower border of mandible, involving angle and body of the mandible, was observed. Inferiorly, it was extending approximately 3 cm below the lower border of the mandible. The overlying skin was stretched, without any sinus or discharge, while consistency was firm in nature (Figs. 1A-B). Intra-orally, it was extending from 43 to the retro molar region with obliteration of the buccal vestibule and obvious expansion.

The CT scan showed a cystic lesion confined to the right side of mandible. There was expansion and thinning of the lower border of mandible, along with perforated bone cortices on both the buccal and lingual aspects of the lesion (Fig 2). Based on clinical and imaging findings, a provisional diagnosis of ameloblastoma was considered. Incisional biopsy punch was done and the microscopic sections showed solid and cystic areas with interspersed follicles which contained tall columnar cells forming the lining, with well-defined darkly stained nucleus and...
aggregates of polygonal cells with granular eosinophilic cytoplasm. A diagnosis of granular cell ameloblastoma with cystic changes was made. (Fig. 3)

Fig. 1A & 1B: Extraoral photo showing a large well-defined swelling on the right side of the face

Fig. 2: 3D CT scan showing tumor mass with multiple radiolucencies

Fig. 3: Photomicrograph showing ameloblastic follicle with central granular cells (marked with arrow) (H and E 40X)

Under general anaesthesia, resection of the lesion with 1.5 cm margin of normal bone, from midline with disarticulation, was done. The resultant defect was stabilised with a 2.7 mm reconstruction plate with condylar head, while
intraoral lining was achieved by pectoralis major musculocutaneous flap. The excised specimen, measuring 13 X 12 X 10 cm, showed on section cystic cavities filled with green colored viscous fluid, separated by thin soft tissue septa. One of the cavities showed soft tissue, with a sand-like consistency. (Fig 4)

Histopathology evidenced follicles of ameloblasts with central round cells showing granular cytoplasm. Cystic areas showed epithelial lining composed of columnar cells resembling ameloblasts with thick layers of granular cells projected into the lumen (Fig. 5). At some areas, a prominent zone of juxta epithelial hyalinization was also noticed, which confirmed the earlier GCA diagnosis with cystic changes.

**DISCUSSION**

AB, one of the most enigmatic odontogenic neoplasms of the oral cavity, has been first described by Broca in 1868 [4]. The term “giant” or “extreme” AB is applied to truly expansive tumors, causing gross asymmetry and impaired function. Patients with large Abs, usually coming from countryside areas, delay the treatment due to ignorance, fear of surgery or because limited or nil access to dental surgeons [5]. In our case, AB grew to an enormous dimension, over 7 years, causing gross facial deformity. Till date, there have been ten reported cases of extreme AB, the maximum size measuring 17 × 15 × 13 cm [5] whereas, in our case, it measured 13 X 12 X 10 cm. All reports referred to large tumors involving half of the mandible, in these ten cases histological diagnosis being either follicular or of AB plexiform type [5]. To our knowledge, this is the second largest reported case of an extreme sized GCA [6]

GCA is one of the rarest entities of all reported ABs [1]. A literature review on the AB of jaws, including the period from 1960 to 1993 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 [2]. A review on 20 cases of the GCA from the files of the Armed Forces Institute of Pathology showed that the average age of reporting was 40.7 years (age interval: 21-65 years). All tumours occurred in the mandible, the vast majority of them (19 out of 20) affecting the posterior regions [7]. In our case too, the patient was 45 year-old, and the posterior mandible was involved.

It is known as locally aggressive among the variants of ABs, being important, because of a higher incidence of malignancy and metastasis [8], to separate GCA from other ABs. A 33.3% recurrence rate for GCA was reported, quite high, comparatively with the more common
follicular, plexiform and acanthomatous subtypes [2]. In another study, 11 of 15 patients (73%) developed recurrent lesions [7].

Numerous theories have been put forward on the origin and nature of these granular cells in ABs, with an epithelial origin; several ultrastructural and histochemical studies have described them as lysosomes and many authors have reported the immunohistochemical (IHC) findings of GCA, yet the actual contents within the granular cells remains largely undefined.

It was speculated that, with age, the unnecessary aged components progressively increase in the cytoplasm of some of the tumor cells, while the ability of lysosomes to eliminate these materials decreases; hence, their cytoplasm becomes packed with lysosomal granules [9]. It was also considered that the lysosomes might have resulted from some genetic alteration of granular cells [10]. A recent IHC and ultrastructural study suggests that the cytoplasmic granularity might be attributed to the increased apoptotic cell death of the neoplasic granular cells, and to their subsequent phagocytosis by the adjacent granular cells [8]. More recent studies suggest that the synthesis of signaling molecules, such as beta-catenin and Wnt-5a, is upregulated in the cytoplasm of some of the tumor cells, while the ability of lysosomes to eliminate these materials is impaired, their accumulation within granular cells remains largely undefined.

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The granular cells of GCA are clearly epithelial in origin, staining exclusively for cytokeratin. It will remain debatable whether the granular cells are a degenerative/ ageing phenomenon or an aggressive presentation of AB [13].

CONCLUSIONS

To the best of our knowledge, the present case is an interesting, most rare occurring variant of AB; a pubmed literature search revealed it as the second case of cystic granular ameloblastoma. Its potential recurrence and metastasis is a matter of concern and such patients need to be evaluated on regular basis for recurrence and for any associated systemic problems, as well. The unique clinical, radiological and histopathological presentation makes this destructive tumor a mystifying proposition.

References

