CASE REPORT

Granular cell type of a unicystic ameloblastoma: An unusual case and review of the literature

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ABSTRACT

Ameloblastomas are locally invasive tumors of odontogenic epithelial origin. They have been categorized broadly into three biologic variants: solid, cystic (unicystic) and peripheral, of which only 13% of all cases are unicystic. Although rare in unicystic or cystic ameloblastoma, granular cell change in ameloblastoma is a recognized phenomenon. Here we present a distinctive case of cystic ameloblastoma with an unusual histopathological pattern along with an added emphasis on its review.

Key words: Ameloblastoma, granular cell type, unicystic ameloblastoma

INTRODUCTION

Ameloblastomas are benign tumors whose importance lies in its potential to grow to enormous size resulting in bone deformity. This slow-growing, persistent, locally invasive odontogenic tumor has been classified into unicystic, multicystic, peripheral and malignant subtypes.[1] Histopathologically, ameloblastomas have a wide range of presentations. In case of solid ameloblastoma, the follicular and plexiform patterns are the most common. Less common histopathologic patterns include the acanthomatous, desmoplastic, basal cell type and granular cell type.[2]

Unicystic ameloblastomas account for 6-15% of all intra-osseous ameloblastomas.[3]It occurs in a younger age group, with slightly more than 50% of cases occurring in patients in the second decade of life.[4] In 1977, Robinson and Martinez first contributed the term "unicystic ameloblastoma".[5] They indicated that the cystic variant is less aggressive type of ameloblastoma that shows a better response to enucleation or curettage than the solid type.

There are various subtypes of unicystic ameloblastoma depending on the character and extent of ameloblastic proliferation within the cyst wall.[2] Luminal cystic ameloblastoma (as in our case) is confined to the luminal surface of the cyst. The lesion includes a fibrous cyst wall with a lining that consists of ameloblastic epithelium showing basal palisading. The overlying epithelial cells are loosely cohesive and resemble stellate reticulum.

The purpose of the present article is to report another case of unicystic ameloblastoma with granular cell change that was reported rarely in the previous literature.

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CASE REPORT

A 57-year-old woman was referred to the Oral and Maxillofacial Pathology Department with swelling over the buccal aspect of her right mandible. The swelling had been present for the last one year and had increased gradually in size over that period. The patient denied any trauma or pain but reported an unusual feeling in that area. Her medical history was unremarkable; she was not taking any medications and had no history of known drug allergy.

Extra-oral examination did not reveal any obvious facial swelling or asymmetry and there was no cervical lymphadenopathy. Intraoral examination demonstrated a round, painless swelling of about 2 cm in diameter in the buccal cortex of right mandible at the apical region of the lower right second premolar. This bony hard swelling showed no history of parasthesia or surface changes. The overlying mucosa was pink and appeared normal. Radiographic examination [Figure 1] revealed a round radiolucency with a well-defined margin at the previous extraction site of first premolar, close to the apex of the lower right second premolar. The first premolar had been extracted more than 10 years ago and the patient could not remember any specific reasons for its extraction. The second premolar showed a normal response to vitality test and there was no root resorption.

Considering the size and clinico-radigraphic features of the lesion, cyst enucleation was done under local anesthesia and the specimen was submitted for histopathological examination. Its gross examination revealed a spherical, yellow cyst measuring 1 cm in diameter containing a milky, semi-translucent viscous fluid. The thickness of the cyst wall was about 1 mm.

Microscopically, hematoxylin and eosin stained sections showed a cyst lined with stratified ameloblastic epithelium consisting of relatively palisaded basal cell layer with plump epithelial cells, granular cytoplasm, small nuclei and distinct borders. These granular cells were closely packed cells distributed in groups throughout the epithelium. The lumen of the cyst was filled with granular cells and connective tissue wall of the cyst was well developed in most areas and not prominently vascular; small areas of hemorrhage and inflammatory cells were also present [Figures 2 and 3]. The granular cells showed variation in both size and shape. Most of them were round; but, angular, oval and polyhedral forms were also common. The nuclei of the granular cells were small, pyknotic and often darkly stained [Figures 4 and 5].

Post-operative follow up of 12 months was uneventful and the patient was continuing to receive routine followup at the time of this study.

DISCUSSION

Vickers and Gorlin, first described the features of the early ameloblastic change that occurs within the wall of a cyst.[6]

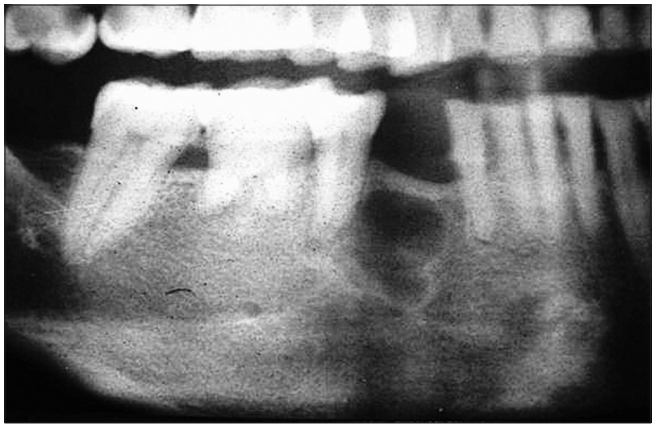

Figure 1: Radiographic examination showing a round unilocular radiolucency with a well-defined margin at the previous site of first premolar extraction near the apex of the lower right second premolar. No root resorption was detected

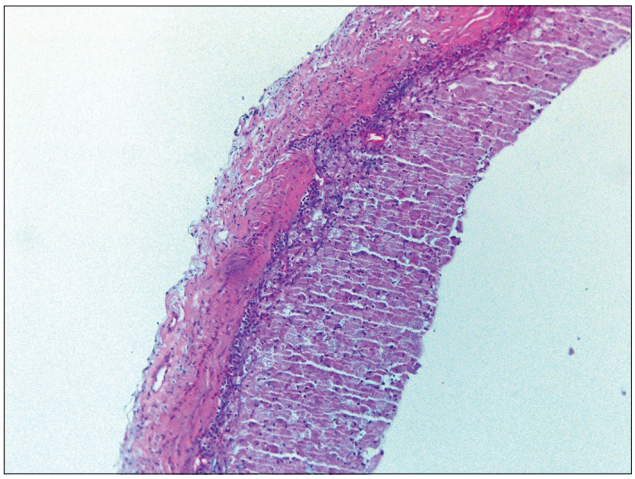

Figure 3: Photomicrograph showing a cyst lined with stratified ameloblastic epithelium consisting of relatively palisaded basal layer with plump granular epithelial cells. The connective tissue wall of the cyst was well developed in most areas and was not prominently vascular. Small areas of hemorrhage and inflammatory cells were also present. The lining of the cyst with abundant granular cells is also evident. (H&E stain, x100)

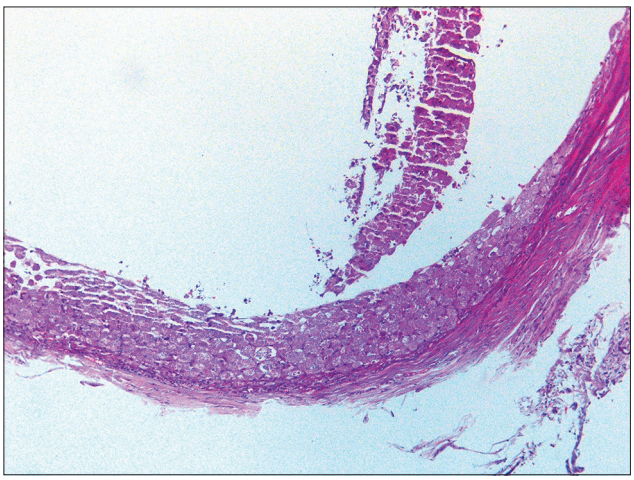

Figure 2: Photomicrograph of the sections of the surgical specimen showed a cyst lined with stratified ameloblastic epithelium consisting of plump epithelial cells with granular cytoplasm and small nuclei. The lumen of the cyst was filled with granular cells as well. (H&E stain, ×40)

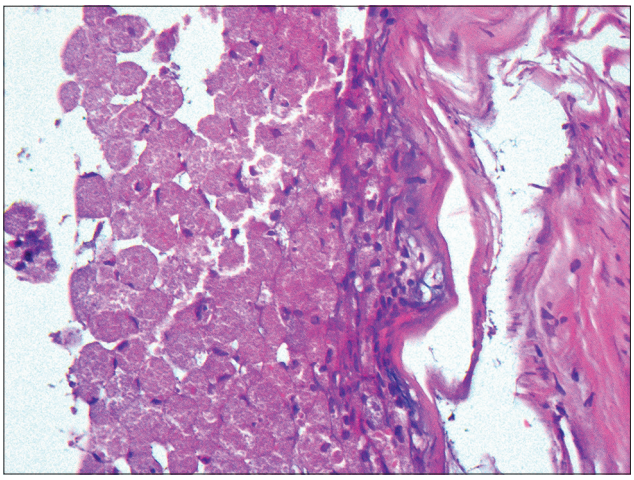

Figure 4: Photomicrograph showing variation in granular cells in both size and shape. Most of them were round; but angular, oval and polyhedral forms were also common. The nuclei of the granular cells were small, pyknotic and often darkly stained. (H&E stain, x400)

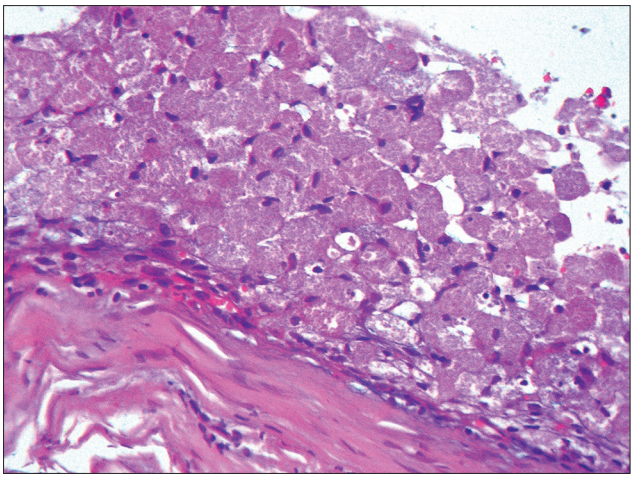

Figure 5: Photomicrograph revealing granular cells that show variation in both size and shape. Most of the cells are round; but angular, oval and polyhedral forms were also common. The nuclei of the granular cells were small, pyknotic and often darkly stained. (H&E stain, x400)

These include hyperchromatism of the nuclei in the basal cell layer of the epithelial lining; palisading and polarization of the basal cell nuclei away from the basement membrane and cytoplasmic vacuolization of the basal cells. The reason why some ameloblastomas become completely cystic may be related to epithelial discohesiveness or more likely to the intrinsic production of proteinases (e.g., metalloproteinases and serine proteinases); enzymes that normally degrade the central zone of the enamel organ after tooth development.[2]

Although this case may not follow all the criteria described by Gorlin and Vicker, the most striking feature of this case was the transformation of epithelial cells to eosinophilic cells containing granules. These granular cells appear to have developed from the basal layer or from the epithelial cells above. Granular cell type of a unicystic ameloblastoma was first described as a granular cell odontogenic cyst in 1970 by Gold and Christ.[7] In 1973, another case was reported by Bouchner.[8] However, Abaza in 1989 concluded that granular cell odontogenic cysts described earlier probably do not exist and they were actually unicystic granular cell ameloblastomas.[9] Another case of cystic ameloblastoma with granular cell change showing ball-like or nodular thickening was also reported in 2010.[10] This cystic ameloblastoma also did not fulfill all Gorlin and Vicker's criteria for an ameloblastic changes.[10]Granular cells have been described in odontogenic tumors as well, such as granular cell ameloblastoma and granular cell ameloblastic fibroma. Similar cells were found in other oral lesions, such as congenital epulis and granular cell tumor.[8]

During normal amelogenesis, an increase in the autophagic lysosomes has been observed in the ameloblasts between secretory and absorptive stages and from reduced ameloblasts to squamous epithelium. Accumulation of granular cells is also noted in enamel organ of deciduous tooth.[11] This may be due to lysosomal insufficiency or overproduction of unused material in the odontogenic epithelium. Thus, odontogenic epithelium seems to be capable of undergoing granular change under certain conditions.[12] There is good evidence documented by electron microscopy and histochemistry[12,13] that cytoplasmic granularity is caused by lysosomal overload. Lysosomal aggregation within the cytoplasm is caused by dysfunction of either a lysosomal enzyme or lysosome-associated protein involved in enzyme activation, enzyme targeting or lysosomal biogenesis.[14] Many authors concluded that numerous lysosomes represent increased cellular actions of the tumor ameloblasts to digest unwanted components.[12] Tsukada *et al*.[15] in 1965 suggested that the granular cells represented an aging or degenerative process. On the other hand, Gold and Christ[7] believed that the granules within the cytoplasm represent a metabolic phenomenon rather than a degenerative process. Abaza *et al*.,[9] concluded that the presence of granular cells in unicystic ameloblastoma is not a permanent feature and may be of a little value as a prognostic indicator of aggressiveness. Kumamoto and Ooya[16]also suggested that cytoplasmic granularity might be caused by increased apoptotic cell death in neoplastic cells followed by phagocytosis of these cells by neighboring neoplastic cells. Therefore Fas/FasL-induced apoptotic cell death may function in the disposal of terminally differentiated (squamous metaplasia) or degenerative tumor cells (granular transformation and cystic changes) in ameloblastomas.[17] Whether the unicystic ameloblastoma originates de novo as a neoplasm or whether it is the result of neoplastic transformation of non-neoplastic cystic epithelium has been long debated. It could be hypothesized that this granular pattern of lining cells could be pointing to a degenerative process occurring in a solid tumor, which ends in such a cyst formation.

CONCLUSION

Although this case may not follow all the criteria described by Gorlin and Vickers clearly, we believe that among all odontogenic lesions, granular cell type of a unicystic ameloblastoma is the only diagnosis that could be matched with both clinical and histopathologic features of this case and should be treated as such.

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