A RARE CASE OF UNICYSTIC ADAMANTINOMA WITH PROMINENT MUCINUS METAPLASIA

Srikumar Chakravarthi[1*] and Barani Karikalan[2]

[1]Faculty of Medicine, Biosciences and Nursing, MAHSA University, Selangor, Malaysia
[2]Faculty of Medicine, RCSI-Perdana University, Kuala Lumpur, Malaysia

***Corresponding Author, Email:** activedoctor@gmail.com*

ABSTRACT

This is a case report of a unicystic adamantinoma with mucinous cell differentiation in the anterior maxilla who suffered from painful swelling for 2 months. A radiograph showed an irregular radiolucent lesion between the roots of 22 and 23. Microscopic examination revealed the cystic lesion was lined with ameloblastic epithelium and goblet cells in the epithelium. The mucous cells reacted positively to the PAS stain. The possible pathogenic mechanism of this case reflects the pluripotential character of the odontogenic epithelium. The prognosis is probably that expected for conventional unicystic ameloblastoma. The significance of identifying such a case of mucous metaplasia in unicystic ameloblastoma is that it should be differentiated from mucous cell We report a case of unicystic ameloblastoma with mucous cell differentiation in the anterior maxilla who suffered from painful swelling for 2 months. A radiograph showed an irregular radiolucent lesion between the roots of 21, 22 and 23. Microscopic examination revealed the cystic lesion was lined with ameloblastic epithelium and goblet cells in the epithelium. The mucous cells reacted positively to PAS stain. The possible pathogenic mechanism of this case reflects the pluripotential character of the odontogenic epithelium. The prognosis is probably that expected for conventional unicystic ameloblastoma. The significance of identifying such cases of mucous metaplasia in unicystic ameloblastoma is that it should be differentiated from mucous cells containing lesions like a glandular odontogenic cyst and mucoepidermoid carcinoma.

Keywords: Unicystic Ameloblastoma, Mucous Metaplasiaoma, Mucous Metaplasiaoma, Prosoplasia, Prosoplasia

Introduction

Ameloblastoma is a benign tumor of odontogenic epithelial origin. It accounts for ~1% of all odontogenic cysts and tumors[1]. There is no significant difference in incidence between males and females. It most frequently involves the posterior region of the lower jaw and has a very wide age range (10-92 years)[1]. Although it is slow growing it can show aggressive behavior. Radiographically, the lesion always presents with multilocular or unilocular radiolucency, commonly accompanied by the resorption of the tooth roots. It has been studied extensively because of its unique clinicopathological features.

According to the recent 2017 WHO classification ameloblastoma can be of four types i.e., conventional, unicystic, extraosseous/peripheral type and metastasizing type. For conventional ameloblastoma two predominant growth patterns are follicular and plexiform. Conventional ameloblastomas also have some less common cell types, including acanthomatous, granular, and basaloid. Additionally, other rare cell differentiations, such as mucous cell differentiation, have been documented in the literature.

Mucous metaplasia is commonly seen in various odontogenic cysts. However, its occurrence in ameloblastoma is an extremely rare phenomenon. Until now only nine such cases have been reported in the literature (Table 1). Out of which only two cases were associated with unicystic ameloblastoma. The present paper reports a third case of unicystic ameloblastoma with mucous metaplasia.

The significance of identifying such cases of mucous metaplasia in unicystic ameloblastoma is that it should be differentiated from mucous cells containing lesions like glandular odontogenic cyst and mucoepidermoid carcinoma which is discussed later in this article.

Comparison of All Cases of Ameloblastoma with Mucous Cell Differentiation

Sr no.	Author	Age/sex	Location	Diagnosis
1.	Hartenian 1976	53/F	Anterior mandible	Solid multicystic ameloblastoma
2.	van Wyk, 1986	21/F	Anterior maxilla	Unicystic ameloblastoma
3.	Raubenheimer,1995	Not available	Anterior mandible	Follicular ameloblastoma
4.	Takata, 1999	51/M	Anterior maxilla	Desmoplastic ameloblastoma
5.	Wilson, 2001	31/M	Anterior mandible	Solid multicystic ameloblastoma
6.	Punnya 2008	17/M	Anterior mandible	Solid multicystic ameloblastoma
7.	Punnya 2008	32/M	Anterior maxilla	Desmoplastic ameloblastoma
8.	Yoon 2009	24/M	Posterior mandible	Unicystic ameloblastoma
9.	Gata 2015	80/M	Posterior mandible	Solid multicystic ameloblastoma
10.	Present case	24/M	Anterior maxilla	Unicystic ameloblastoma

Case Report

A 24-year-old male patient was reported with pain and swelling on the left side of anterior palate region for one and half months, for which he visited a hospital and got a Root canal opening done with 21 22 and 23 but still swelling did not subside.

Intraorally swelling was 3×2 cm in size extending from gingiva of 21, 22, 23 and 24 to mid-palatine raphe with roughly oval shape (fig.1 a&b) Margins were ill-defined. Radiograph shows ill-defined radiolucency involving roots of 21, 22 and 23 (Fig.2 a&b). Enucleation of cystic lesion with apicectomy of 21, 22 & 23 was done.

H &E stained section of cystic lesion showed cystic cavity focally lined by non-keratinised stratified squamous epithelium of variable thickness. At places lining epithelium exhibited basal cuboidal to columnar cells with hyperchromatic nuclei and loosely arranged stellate reticulum-like cells suggestive of odontogenic epithelium. Focally superficial cell layers of lining epithelium also exhibited mucous metaplasia. Fibrous connective tissue wall exhibited epithelial islands in form of follicles. These follicles showed peripheral tall columnar cells with hyperchromatic nuclei and central stellate reticulum like cells.

Connective tissue capsule shows parallelly arranged collagen bundles peripherally with mild inflammatory infiltrate, moderate degree vascularity and areas of hemorrhage (fig 3 a & b)

Therefore, the final diagnosis was given as unicystic ameloblastoma – mural variant with mucous

Discussion

As stated by Robinson, Ameloblastoma is usually unicentric nonfunctional, intermittent in growth, anatomically benign and clinically persistent. Historically, Ameloblastoma has been recognized for over a century and a half. Its frequency, persistent local growth, and ability to produce marked deformity before leading to serious debilitation probably account for its early recognition. Recurrence, especially after conservative treatment, has also contributed to the awareness of this lesion [15].

Histologically conventional ameloblastoma shows various types like follicular, plexiform, acanthomatous, Granular, desmoplastic and basal cells. In a granular cell, ameloblastoma granularity is due to the marked transformation of the cytoplasm of stellate reticulum-like cells into a coarse, granular eosinophilic appearance previously it was considered to be aggressive in nature, with a marked propensity for recurrence and metastasis. But Recently it is considered that all types of histological variants of conventional ameloblastoma exhibit similar aggressive behavior. So treatment modality is similar in all variants[15].

Ameloblastomas have been broadly divided into three biological variants, of which the unicystic type is least aggressive. Unicystic ameloblastoma shows an incidence of around 5–22%, but with a lower recurrence (6.7–35.7%) generally affecting a younger population[16]. It presents three histopathologic subtypes. The luminal variant is a cystic lesion with a flat ameloblastic cystic lining. The intraluminal subtype is characterized by tumor growth into the cyst lumen, while the mural subtype presents infiltrating growth into the wall of the cyst and possibly beyond into the surrounding bone[16].

Leider et al. proposed three pathogenic mechanisms for the evolution of UA: the reduced enamel epithelium associated with a developing tooth undergoes ameloblastic transformation with subsequent cystic development; ameloblastomas arise in dentigerous or other types of odontogenic cysts in which the neoplastic ameloblastic epithelium is preceded temporarily by a non-neoplastic stratified squamous epithelial lining; and a solid ameloblastoma under goes cystic degeneration of ameloblastic islands with subsequent fusion of multiple microcysts and develops into a unicystic lesion[17].

Ameloblastomatous epithelium of UA can show various histologic patterns most commonly plexiform patterns. Some cases are reported with granular, acanthomatous and basaloid pattern inlining epithelium[16]. Mucous cell differentiation is also seen in some cases. The present article reports a tumor that exhibits a characteristic histopathological appearance of Unicystic ameloblastoma. Along with this most interesting aspect of this tumor is the presence of mucous cell differentiation[8].

The finding of clear or pale cells such as mucous cells—in ameloblastomas is a very rare, but not unreported, phenomenon. But its presence in odontogenic cysts like radicular cyst is common. The diagnostic implications of the presence of clear or pale cells in ameloblastomas are several and relate mainly to the question of histopathologic differential diagnosis. because of the presence of mucous cells, there are possibilities of other histopathological diagnoses like mucoepidermoid carcinoma, clear cell odontogenic tumor or clear cell odontogenic carcinoma.

The pathogenesis of mucous cells within ameloblastomas is not clear. It has been suggested that the odontogenic epithelial component might have multipotential differentiation ability (Yoon et al., 2009) and mucous cell differentiation of the epithelial cells might be a response to an altered environment, such as inflammation or necrosis in salivary gland tumors (Taxy, 1992). Another hypothesis is that the differentiated cells undergo the process of prosoplasia, which is known as a forward differentiation either to a higher or intricate function or to a more complex level of organization. The most classical example is the differentiation of the squamous epithelial cells to mucous cell (Sarode, Maniyar, Sarode, Rao, & Patil, 2017). However, those hypotheses need to be further explored in more cases of ameloblastoma with mucous cell differentiation in the future.

Whether a unicystic ameloblastoma in association with mucous cells represents a collision growth of two distinct components or a metaplastic phenomenon within a unicystic ameloblastoma remains speculative. It was previously suggested that the epithelial lining of the odontogenic cysts can undergo metaplasia from a stratified squamous to a more highly differentiated ciliated columnar or glandular type. The stimulus for such metaplasia is unclear, although increased hydrostatic pressure has been suggested. The possible pathogenic mechanism of this case would appear to be a reflection of the pluripotential character of the odontogenic epithelium. This case indicates that multipotential odontogenic epithelial tissue has the ability to develop diverse differentiation. The prognosis, in this case, will probably be as expected for conventional unicystic ameloblastoma.

The histopathologic differential diagnosis due to the presence of mucous cells

includes salivary gland tumor, especially a mucoepidermoid carcinoma, which is known to occur commonly in the jaws, and glandular odontogenic cyst. The diagnosis of mucoepidermoid carcinoma is based on the identification of a mixture of mucous, intermediate, and epidermoid cells, while glandular odontogenic cyst (GOC) is predominantly cystic lined by a typically thin stratified squamous epithelium with a characteristic superficial layer of cuboidal/columnar cells which occasionally exhibit cilia[14]. Additionally, the epithelium in GOC may show swirling spherical aggregates, papillary proliferation, and pools of mucin[1].

Conclusion

The existence of mucous cells in ameloblastoma thus illustrates the multipotentiality of odontogenic epithelium and highlights the need for careful histopathologic assessment of ameloblastoma for documentation of such extraordinary heterogeneities in its histopathologic spectrum and prognostic significance, if any, of this rarity.

REFERENCES

1. Punnya A, Angadi V.Rekha K. "Ameloblastoma with mucous cells": Review of literature and presentation of 2 cases. *Oral Surgery, Oral Medicine, Oral Pathology, Oral Radiology, and Endodontology*, 2008, 106.6: e20-e26.
2. J.-H. Yoon, S.-G. Ahn, S.-G. Kim: Mucous cell differentiation in a unicystic ameloblastoma. Int. J. Oral Maxillofac. Surg. 2009; 38: 91–97.
3. WILSON, David, et al. Ameloblastoma with mucous cell differentiation. Oral Surgery, Oral Medicine, Oral Pathology, Oral Radiology, and Endodontology, 2001, 91.5: 576-578.
4. XIA, Rong-Hui, et al. Ameloblastoma with mucous cells: A clinicopathological, BRAF mutation, and MAML2 rearrangement study. *Oral Diseases*, 2020, 26.4: 805-814
5. Sarode S, Sarode G et al. Mucous cell prosoplasia in oral pathologies: a brief review. *Journal of clinical and diagnostic research: JCDR*, 2017, 11.4: ZE08.
6. Gataa IS, Garib BT, Rashid NH (2015) New Features in Mucous-Ameloblastoma. A Case Report of rare Entity. Int J Oral Craniofac Sci 1(1): 101.
7. Tamgadge S, Tamgadge A, Bhalera S, Periera T. Mucous Cell Differentiation in Desmoplastic Ameloblastoma: Unique Presentation in Posterior Mandible. International Journal of Oral and Maxillofacial Pathology; 2012:3(2):00-00.
8. A Kudva , A. Kamath , N Rao , J Rajan: Rare case of giant unicystic ameloblastoma: Luminal variant, CRANIO (2017)
9. Hartenian KM, Kalfayan B. Ameloblastoma containing mucus glands. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 1976;41:508-15.

10. van Wyk CW, Thompson IOC, Wyma GA. Unicystic ameloblastoma mimicking a "globulomaxillary" cyst: a case report. Br J Oral Maxillofac Surg 1986; 24 :422-5.
11. Raubenheimer EJ, van Heerden WFP, Noffke CE. Infrequent clinicopathologic findings in 108 ameloblastomas. J Oral Pathol Med 1995; 24:227-32.
12. Takata T, Miyauchi M, Ito H, Ogawa I, Kudo Y, Zhao M, et al.
13. Clinical and histopathological analyses of desmoplastic ameloblastoma. Pathol Res Pract 1999; 195:669-75.
14. Shah AA, Sangle A, Bussari S, Koshy AV. Glandular odontogenic cyst: A diagnostic dilemma. Indian journal of dentistry. 2016 Jan;7(1):38.
15. Masthan KM, Anitha N, Krupaa J, Manikkam S. Ameloblastoma. Journal of pharmacy & bioallied sciences. 2015 Apr;7(Suppl 1): S167.
16. Ponce JB, Lima HG, Rodrigues MT, Souza FG, Lara VS. Unusual histological patterns and hyaline ring granulomas in a unicystic ameloblastoma. Hippokratia. 2014 Jan;18(1):83.

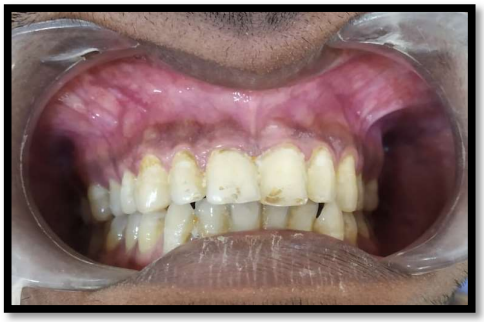

Figure 1-a

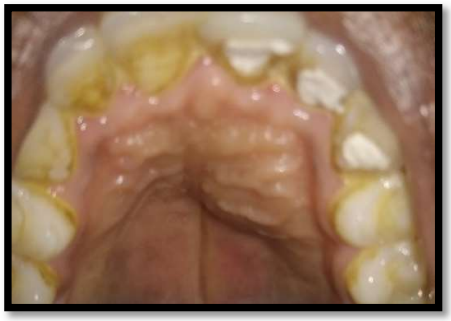

Figure 1-b

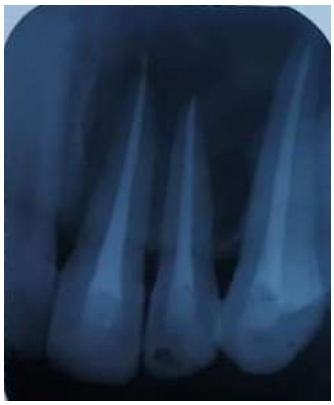

Figure 2-a

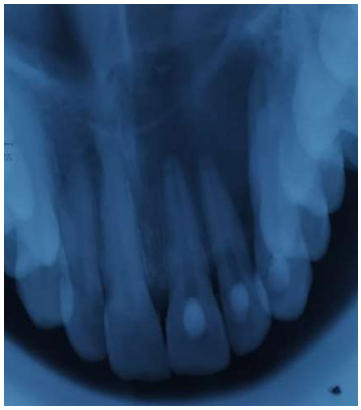

Figure 2-b

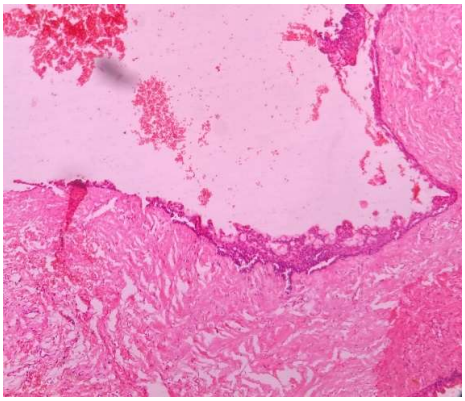

Figure 3-a

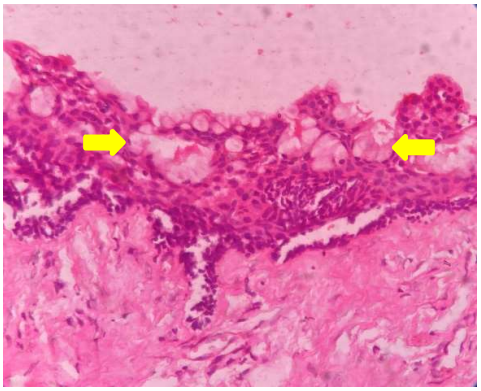

Figure 3-b