

Unicystic ameloblastoma: Mimicking a cyst

Dandekar RC¹, Shankar AA²

¹Professor & Head, ²Post Graduate Student, Department of Oral Pathology & Microbiology, M. A. Rangoonwala Dental College, 2390-B, K. B. Hidayatullah Road, Azam Campus, Camp, Pune

Abstract

A 17 year old patient reported with a swelling in the left posterior mandibular region since 3 months. On clinical examination, the swelling was well-circumscribed and extended from the ala-tragus line to lower border of the mandible on the left side. It was hard and tender on palpation. Radiographic examination revealed a unilocular appearance and was suggestive of a cyst or tumour. An aspirate of the fluid was obtained, and protein analysis revealed 4.3 gm/dl, suggesting a keratinizing cyst/tumour. The entire lesion was excised leaving the condyle in position, upto the mental foramen. An iliac crest graft, 10cm in length, was placed, and the mandible reconstructed. The final histopathological diagnosis was given as Unicystic Ameloblastoma.

Key words: Ameloblastoma, Vicker and Gorlin

Introduction

Ameloblastoma is usually unicentric, nonfunctional, intermittent in growth, anatomically benign and clinically persistent¹. It is a true neoplasm of the enamel organ type tissue, but does not undergo differentiation upto the point of enamel formation². There are various varieties of ameloblastoma, viz. follicular, plexiform, acanthomatous, granular and unicentric. The follicular, plexiform and unicentric variety are the most common varieties, and may co-exist in the same specimen.

The usual presenting age of an ameloblastoma is considered to be in the 3rd and 4th decades with a predilection for Asian population. The unicystic ameloblastoma is a well-defined, often large monocystic cavity with a lining, focally but rarely entirely composed of odontogenic epithelium³. It is histopathologically divided into intracystic, luminal or intraluminal unicystic ameloblastoma. Also, tumours associated with an unerupted tooth are considered to be the dentigerous variant, whereas those lacking an association with an unerupted tooth were considered to be the non-dentigerous variant. The dentigerous variety is known to occur in younger patients.

Case report

A 17 year old male presented with pain and discomfort in the left posterior mandibular region. A swelling present in the lower left region, extending from the left ala-tragus line to the lower border of mandible was noted (Fig 1), and was found to be hard on palpation. Disfigurement and asymmetry of the face were evident. Intraoral examination revealed a missing lower left second molar tooth (Fig 2).

The radiographic findings showed a multilocular radiolucency extending from the molar to the condyle region (Fig 3). The inferior border of the mandible could not be traced well. A computed tomography scan revealed a tooth in association with the lesion (Fig 4).

The entire tumor was removed in toto. A gross examination of the lesion showed a smooth cyst wall, without any perforations (Fig 5). A tooth was noted in association with the lesion, but did not seem to be part of the lesion (Fig 6).

A detailed histopathological examination was carried out. The slides were stained with routine H&E stain. The

Correspondence

Dr. Akhil A. Shankar, Post Graduate Student, Department of Oral Pathology & Microbiology, M. A. Rangoonwala Dental College, 2390-B, K. B. Hidayatullah Road, Azam Campus, Camp, Pune - 411001, E-mail: akhil1904@hotmail.com

histopathology showed a cystic lining with the basal cells displaying a palisading appearance. Hyalinization of the subjacent connective tissue capsule was noted at places (Fig 7). A higher magnification showed hyperchromatic nuclei in cuboidal to columnar basal cells. The superficial epithelium showed prominent intracellular spacing, creating a stellate reticulum like appearance (Fig 8 & Fig 9). The above features in relation to the radiographic findings were suggestive of a Unicystic Ameloblastoma type 1.2.

Table 1: Histologic Unicystic Ameloblastoma Subgrouping by Ackermann et al

Subgroup	Interpretation
1	Luminal UA
1.2	Luminal and Intraluminal UA
1.2.3	Luminal, Intraluminal, and Intramural UA
1.3	Luminal and Intraluminal UA



Fig 1: Swelling of the mandible on the left side (arrows)



Fig 2: Absence of lower left second molar



Fig 3: OPG image showing a multilocular radiolucency

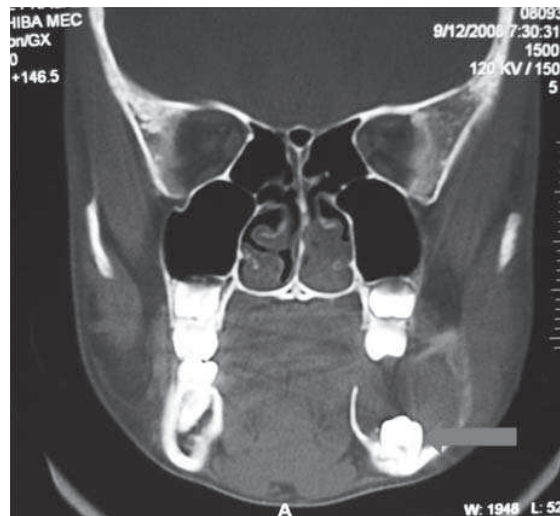


Fig 4: CT shows tooth in association with lesion (arrow)

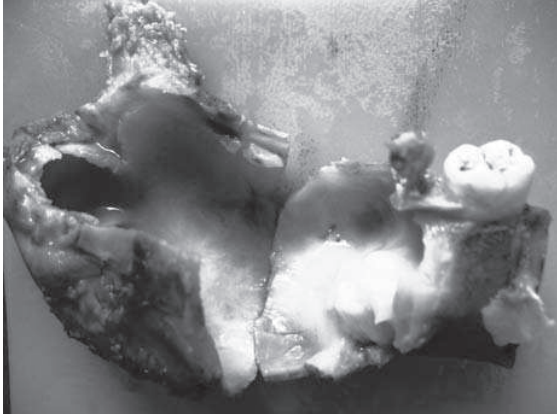


Fig 5: Gross specimen – smooth cystic walls

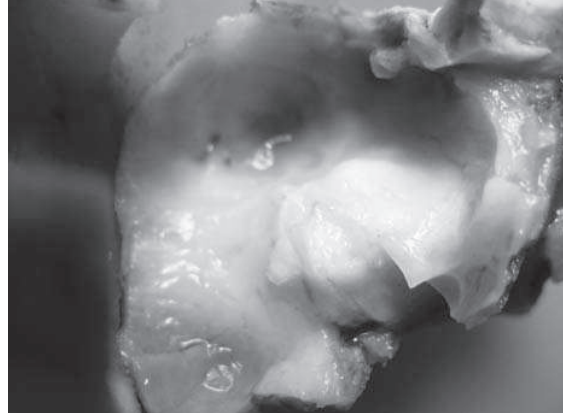


Fig 6: Impacted tooth, outside the lesion

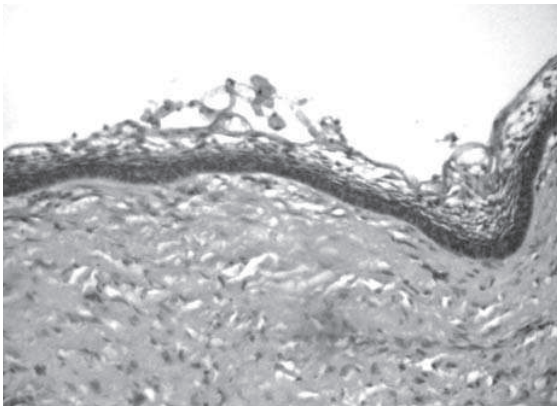


Fig 7: Palisaded basal cells and hyalinization of the subjacent connective tissue

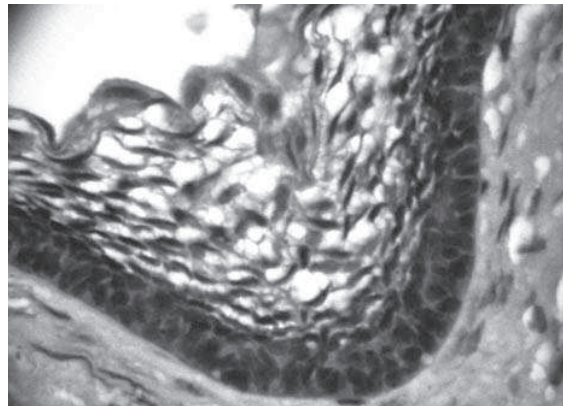


Fig 8: Hyperchromatic nuclei in basal columnar cells

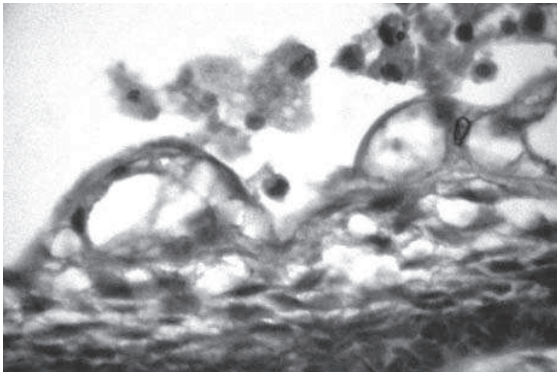


Fig 9: Stellate reticulum like appearance of superficial cells

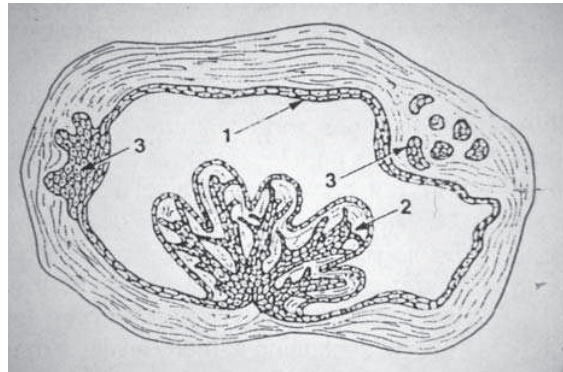


Fig 10: Types of growth in Unicystic Ameloblastoma
1. Luminal, 2. Intraluminal, 3. Intramural

Discussion

The ameloblastoma is the second most commonly occurring odontogenic tumor after the odontoma², and 80 per cent of all cases occur in the molar-ramus region. The term Ameloblastoma was suggested by Churchill in 1934. It is known to occur equally in both sexes and afflicts patients mainly in the 3rd to 5th decade. There are almost upto fifteen different types of this tumour

recorded till date³. The most commonly occurring varieties of this tumour histologically are follicular, plexiform, granular, desmoplastic, basal cell, unicystic and the lesser occurring peripheral variant. The other variants of the tumour are acanthomatous, clear cell, keratoameloblastoma, mucous cell differentiation and hemangiomatous. The surgical management of these

tumours essentially remains the same and primarily differs for the Unicystic variety.

The unicystic ameloblastoma was first defined as Robinson and Martinez in 1977⁴. The frequency of these tumours is reported to be between 5% and 22% of all types of ameloblastomas⁵. An age old debate for the pathogenesis of this lesion still prevails. Three mechanisms have been proposed for the development of the unicystic ameloblastoma⁶:

1. The reduced enamel epithelium associated with a developing tooth undergoes ameloblastic transformation with subsequent cystic development
2. 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
3. A solid ameloblastoma undergoes cystic degeneration of ameloblastic islands with subsequent fusion of multiple microcysts and develops into a cystic lesion.

The fact that the PCNA-positive cells are significantly higher as compared to that of the dentigerous cyst linings suggests that this tumor arises de novo.

One complication known to arise from dentigerous cysts is its transformation to ameloblastomas. Although a highly argued topic, yet as given by Vicker and Gorlin⁷, the following points are used to distinguish UA from any other cystic lesion:

1. Epithelial lining – parts may show transformation to cuboidal or columnar basal cells with hyperchromatic nuclei
2. Nuclear palisading with polarization
3. Cytoplasmic vacuolation with intercellular spacing
4. Subepithelial hyalinization

The above mentioned findings confirmed with those seen in our case.

Ackermann et al⁸ have provided a histological subgrouping of the Unicystic Ameloblastoma as shown in Table 1 and a diagrammatic representation of the same shown in Fig. 10.

Based on the above classification, our case was given a subgroup of 1.2. From a surgical aspect, the tumour under the subgroup class 1 can be enucleated and the jawbone salvaged, provided it is salvageable. A higher subgroup requires the surgery as always performed for any other ameloblastoma, which might vary from segmental resection to hemimandibulectomy, depending on the size of the lesion, and also the amount of bone remaining. A hemimandibulectomy with reconstruction was carried out in this case.

References

1. Robinson HBG. Proceedings of the Fifth Annual Meeting of the American Academy of Oral Pathology. *Oral Surg* 1952;5:177-8.
2. R Rajendran, B Sivapathasundharam. *Shafer's Textbook of Oral Pathology*. 5th Edn. Elsevier. New Delhi. 2007. pp 381-390.
3. Reichart PA, Philipsen HP. *Odontogenic Tumours and Allied Lesions*. 1st Edn. Quintessence Publishing. New Delhi. 2004. pp 77-86.
4. Robinson L, Martinez MG. Unicystic Ameloblastoma. A prognostically distinct entity. *Cancer* 1977;40:2278-2285.
5. Reichart PA, Philipsen HP, Sonner S. Ameloblastoma: Biological profile of 3677 cases. *Eur J Cancer B Oral Oncol* 31B:86-99. 1995.
6. Leider AS, Eversole LR, Barkin ME. Cystic Ameloblastoma. *Oral Surg Oral Med Oral Pathol* 1985;60:624-630.
7. Vickers RA, Gorlin RJ. Ameloblastoma: Delineation of early histopathological features of neoplasia. *Cancer* 1970;26:699-710.
8. Ackermann GL, Altini M, Shear M. The unicystic ameloblastoma: A clinicopathologic study of 57 cases. *J Oral Pathol* 1988;17:541-546.