

An Unusual Presentation of Unicystic Ameloblastoma in the Anterior Maxilla

Himanshu Lakhani¹, A.G. Annaji², M. Manjunath³, Khalida Shaik Begum⁴, Anjana Ramanathan⁵, Abhinetra. M.S⁶

1,4,5-Post graduate, Dept Of Oral Medicine And Radiology, Vokkaligara Sangha dental College And Hospital, Bengaluru, Karnataka. 2,3,6-M.D.S, Oral medicine and Radiology, Vokkaligara Sangha Dental College And Hospital, Bengaluru, Karnataka.

Correspondence to:
Dr. Lakhani Himanshu, Post graduate, Dept Of Oral
Medicine And Radiology
Contact Us: www.ijohmr.com

ABSTRACT

Ameloblastoma is the most common odontogenic tumor. Unicystic ameloblastoma is considered to be a variant of the solid or multicystic type. It is a less aggressive tumor with variable recurrence rate. Unicystic lesions are the ones that show clinical, radiographic and gross features of a cyst but on histologic examination shows a typical ameloblastomatous epithelium which lines a part of the cyst cavity, with or without luminal and/or mural tumor growth. We report a case of unicystic mural ameloblastoma in 21 year old male with swelling in the right upper front region of the face since 2 months. Fine needle aspiration yielded no fluid. Radiographs revealed a multilocular radiolucency in the right anterior maxillary region with buccal cortical plate expansion.

KEYWORDS: Ameloblastoma, Unicystic, Mural, Maxilla

INTRODUCTION

Robinson defined Ameloblastoma as “Unicystic, nonfunctional, intermittent in growth, anatomically benign and clinically persistent.” They are tumors of odontogenic epithelial origin, they may arise from basal cells of the oral mucosa, an odontogenic cyst lining or a developing enamel organ.¹

About 85% of the conventional Ameloblastomas occur in the mandible, most often in the molar-ascending ramus area whereas 15% occur in the maxilla, usually in the posterior region. The tumor is often asymptomatic and smaller lesions are usually detected only during a radiographic examination. A painless swelling or expansion of the jaw is the usual clinical presentation.¹

They are usually discovered around fourth and fifth decades of life with the exception of the Unicystic variant which is more common in second and third decade of life.²

Radiographically, Ameloblastomas can either present as a unilocular or a multilocular radiolucency. The lesion is often described as having a “soap-bubble” appearance when the radiolucent loculations are large and as being “honey-combed” when the loculations are small. Buccal and lingual cortical expansion are frequently present. Resorption of roots of teeth adjacent to the tumor is also seen.

Histologically, Ameloblastomas are classified into multicystic/solid, unicystic, and peripheral types. The Unicystic variant is less aggressive and has a low recurrence rate except for those lesions which demonstrate mural invasion. Such lesions should be treated aggressively.

Here we report a case of Unicystic intramural

Ameloblastoma of the maxilla in a 21 year old male.

CASE REPORT

A 21 year old male presented with a swelling in the right upper front tooth region since 2 months. He gave a history of swelling that gradually increased in size over a period of 2 months and was not associated with pain, tenderness or discharge.

On extraoral examination, a diffuse swelling was noted on the right side of the face measuring about 3cm x 4cm in size which was extending from ala-tragus line superiorly till 3cm inferiorly and from the philtrum of the upper lip mesially till right corner of the mouth distally (Figure 1). The skin overlying the swelling appeared to be normal. The swelling was hard in consistency and non tender.



Figure 1- Extra oral photo of the patient

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On Intraoral examination, a diffuse swelling was noted on the labial vestibule extending from 11 to 14, (Figure 2) and palatally extending from 11 anteriorly upto 16 posteriorly and medially extending upto the mid palatine raphae (Figure 3 & 4). There was palatal rotation seen i.r.t 12. Slight extrusion noted i.r.t 11. On palpation, the swelling was bony hard on the labial aspect and soft on the palatal side. It was non tender.



Figure 2-Extent of the lesion on the buccal aspect



Figure 3-Palatal extent of the lesion



Figure 4- Palatal extent of the lesion

IOPAR and Occlusal radiographs revealed a well defined multilocular radiolucency measuring about 5cm × 7cm with radiopaque margins seen extending from 11 to distal

of 16 (Figure 6 & 7) . There is slight extrusion seen i.r.t 11. Buccal cortical plate expansion is seen in the right



Figure 5- Intraoral Periapical radiograph showing distal displacement of root of 13 and mesial displacement of root of 12



Figure 6-Intraoral Periapical radiograph showing extrusion of 11.

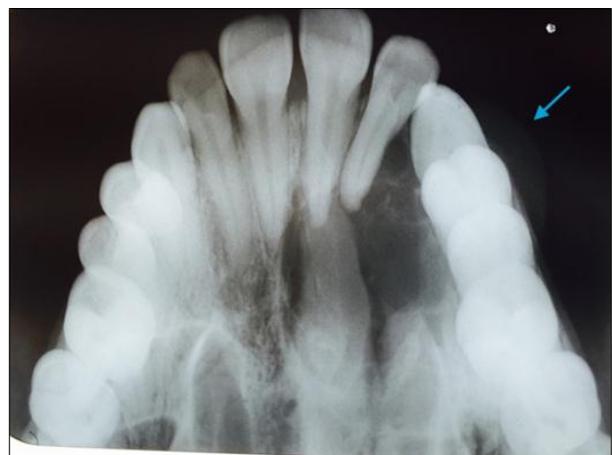


Figure 7-Maxillary cross sectional occlusal radiograph showing a multilocular lesion and its extent. Buccal cortical plate expansion (blue arrow) is seen.

side of the maxilla on a cross sectional occlusal radiograph. Medial displacement of the root of 12 and distal displacement of 13 is seen (Figure 5). CBCT revealed the expansion of the cortical plate and extension of the lesion (Figure 8). Thus going by the clinical and radiographic findings, a provisional diagnosis of Adenomatoid Odontogenic Tumor was considered. Aspiration of the lesion was non productive. An incisional biopsy was performed under local anesthesia from the palatal aspect of 13 and 14 and submitted for histopathological examination. The microscopic examination of H&E stained section showed cystic lining consisting of tall columnar cells in the basal layer with hyperchromatic nuclei, which resembled ameloblasts. The overlying epithelial cells were loosely cohesive and resembled stellate reticulum. Follicles were seen within the connective tissue stroma (Figure 9). Thus, the overall histopathological features were suggestive of Unicystic Ameloblastoma- Type III(Mural type) . Later the lesion was surgically excised (Figure 10) and subjected to histopathological analysis which confirmed the diagnosis of Unicystic Ameloblastoma with mural proliferation. Post operative follow-up of the patient had been satisfactory.

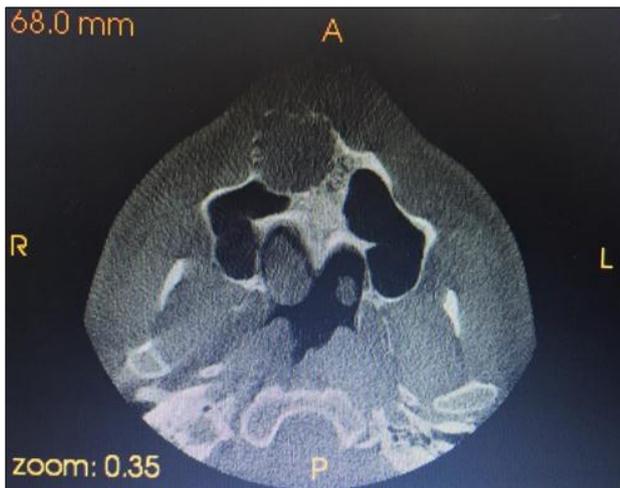


Figure 8 -Axial section of Cone Beam CT showing the extent of the lesion

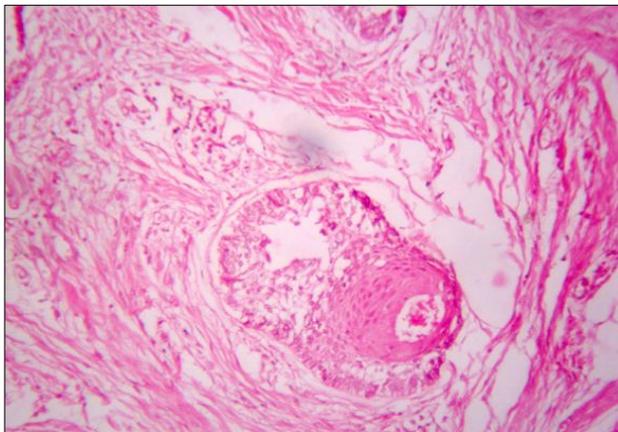


Figure 9- Histopathological picture showing cystic lining consisting of basal layer of tall columnar cells with hyperchromatic nuclei, resembling ameloblasts.



FIGURE 10 -Enucleation of the lesion was done along with teeth 11,12,13 and 14

DISCUSSION

Ameloblastoma is a benign , locally aggressive odontogenic neoplasm with variable clinical expression and accounts for 1% of all cysts and tumors of jaws and 18% of all odontogenic neoplasms.³

Unicystic ameloblastoma , a variant of ameloblastoma was first described by Robinson and Martinez in 1977.⁴

It accounts for about 6 % of ameloblastomas and usually occurs in a younger age group of 16-20 years with about 50% of the cases occurring in the second decade of life.^{5,6}

It occurs more commonly in males than females with a male to female ratio of 1.6:1. However when the tumor is not associated with an unerupted tooth, the male : female ratio is 1:1.8.³

The location of this lesion in the anterior maxilla is considered to be rare and atypical with more than 90% of unicystic ameloblastomas occurring in the mandibular posterior region , followed by the parasymphiseal region , the anterior maxilla and the posterior maxilla. The ratio of mandibular to maxillary unicystic ameloblastoma has been reported to be 13:1.⁵

Small lesions are usually asymptomatic but a larger lesion may result in a painless swelling of the jaws. Continuous growth of a tumor may cause mucosal ulceration which is not seen commonly.⁷

The clinical and radiographic findings in most cases of unicystic ameloblastoma suggest that the lesion is an odontogenic cyst, particularly dentigerous cyst. However, some of them are not associated with an impacted tooth and are therefore called as non-dentigerous variant.⁷ Most of the unicystic ameloblastomas are associated with an impacted tooth, mandibular third molar being most commonly involved. But in our case, there was no associated impacted tooth, so it is a non-dentigerous variant.

The radiographic appearance of unicystic ameloblastoma has been divided into 2 main patterns: unilocular and multilocular. The dentigerous variant shows unilocular

pattern predominantly with a unilocular to a multilocular ratio of 4.3:1 whereas the non dentigerous type shows a ratio of 1.1:4.^{8,9}

The differential diagnosis for lesions in this location includes dentigerous cyst, radicular cyst, odontogenic myxoma, ameloblastoma and ameloblastic fibroma. The clinical and radiographic features are not sufficient to come to a definitive diagnosis. A histopathological confirmation is always required for diagnosis.¹⁰

Ackermann classified unicystic ameloblastoma into the following histological groups :

- Group I: Luminal unicystic ameloblastoma (tumor confined to the luminal surface of the cyst.)
- Group II: Intraluminal/ plexiform unicystic ameloblastoma (nodular proliferation into the lumen without infiltration of tumor cells into the connective tissue wall.)
- Group III: Mural unicystic ameloblastoma (invasive islands of ameloblastomatous epithelium in the connective tissue wall not involving the entire epithelium.)

Recently Philipsen and Reichart modified the classification of Ackermann et al. :

- Subgroup 1: Luminal Unicystic Ameloblastoma
- Subgroup 2: Luminal and Intraluminal
- Subgroup 1.2.3: Luminal, Intraluminal, and Intramural
- Subgroup 1.3: Luminal and Intramural.

According to the author, this classification can help the surgeon in deciding the treatment plan. They indicated that the tumor in subgroup 1 and 1.2 can be treated with careful enucleation, whereas subgroups 1.2.3 and 1.3 require radical resection as for a solid or multicystic ameloblastoma. Following enucleation, curettage of the bone should be avoided as it may implant foci of ameloblastoma deeper into the bone. Chemical cauterization with Carnoy's solution is also advocated for subgroups 1 and 1.2. Subgroups 1.2.3 and 1.3 have a high risk for recurrence, requiring more aggressive surgical procedures.

A definitive diagnosis of unicystic ameloblastoma cannot be made on the basis of clinical and radiographic grounds alone. A histopathological examination of the biopsy is not representative of the entire lesion and may result in an incorrect classification.³

The epithelial lining of unicystic ameloblastoma is not always uniformly characteristic and is often lined partly

by a non specific thin epithelium that mimics the dentigerous cyst lining. Thus, true nature of the lesion becomes evident only after enucleation when the entire specimen is available for microscopy.⁷

CONCLUSION

The pathogenesis of cystic ameloblastoma remains obscure. Whether unicystic ameloblastoma originates as a neoplasm from the beginning or whether it is a result of neoplastic transformation of non-neoplastic cyst epithelium is still not clearly understood. Some investigators believe that it arises from pre-existing odontogenic cysts, particularly, a dentigerous cyst, while others think that it arises de novo.

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