



UNICYSTIC AMELOBLASTOMA- A CASE REPORT AND A SHORT REVIEW OF LITERATURE

Maxillofacial Surgery

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ABSTRACT

Ameloblastoma has been described by Robinson as a tumor which is “usually unicentric, nonfunctional, intermittent in growth, anatomically benign and clinically persistent”¹. It mostly occurs in second and third decades of life but it can be seen in children. It is usually asymptomatic for a long period, slow growing, aggressive, locally invasive tumor with multiple histological variants and high recurrence rates⁴. The term Unicystic Ameloblastoma (UA) refers to those cystic lesions that show clinical and radiological characteristics of an odontogenic cyst but on histological examination show a typical ameloblastomatous epithelium, lining part of the cyst cavity, with or without luminal and/or mural tumor growth⁶. Unicystic ameloblastoma (UA) is an uncommon variant of ameloblastoma and behaves totally different from the solid multicystic variant of ameloblastoma (SMA)⁷. Here, we report a case of unicystic ameloblastoma which was misdiagnosed radiographically as dentigerous cyst in a 29-year-old female with a review of the literature.

KEYWORDS

Unicystic ameloblastoma, Impacted tooth, Enucleation, Mural type, Conservative management

INTRODUCTION

In the latest WHO classification of head and neck tumors, ameloblastomas are subcategorized into solid multicystic, unicystic, peripheral and metastasizing types.^{2,7}

Unicystic ameloblastoma was first described in 1977, and has been considered a special type. The young age at which it occurs, its unilocular radiographic appearance, macroscopic cystic appearance and most importantly the better response to conservative treatment, makes it a distinguishable entity.^{5,9}

Unicystic ameloblastoma is considered to be a less aggressive form compared with solid or multicystic type and accounts for 5 to 15% of all ameloblastomas.^{10,11,12}

We report a case of a 29-year-old female with unicystic ameloblastoma (involving the crown of unerupted mandibular second molar). This case report stresses on the fact that there is always a potentiality of a discrepancy in clinical, radiological and histological diagnosis.

Case Report

A 29 year old female was referred to our department, with a slow growing mass in the right side of the lower jaw, noticed since a year. She was otherwise healthy, no significant medical history. Patient was asymptomatic with discreet facial asymmetry and no cervical lymphadenopathy. Extraorally, the mass was hard in consistency, nontender, not mobile. Intraorally, there was expansion of buccal and lingual cortices of the right body of the mandible. The size of the lesion was approximately 3x3 cm with clinically missing lower right second molar. Grade I mobility of right lower first molar. Patient had paresthesia in the distribution of right V3. A panorama Xray and cone beam CT was done. OPG revealed a unilocular radiolucency with an impacted lower right 2nd molar (Fig 1); and resorption of the distal root of lower right 1st molar. Cone beam CT revealed a unilocular radiolucency in the right body of the mandible with an impacted molar measuring around 5x4 cm, with cortical expansion and thinning (Fig 2-3a and 3b). The right lower 1st molar vitality was questionable even after the vitality test. A differential diagnosis of dentigerous cyst was made and surgery planned accordingly.

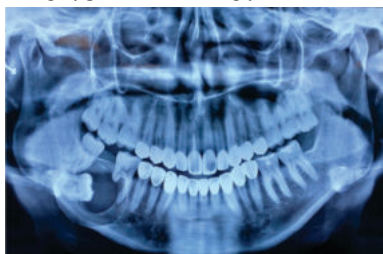


Fig. 1 Panorama Xray- Unilocular Radiolucency

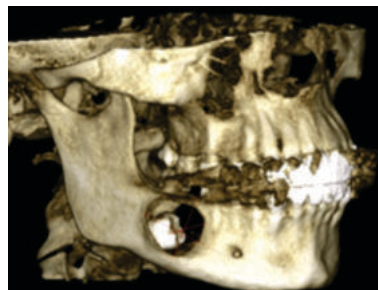


Fig 2 Cone Beam CT 3d



Fig.3a Cone Beam CT – Axial View, Expanded Buccal And Lingual Cortices



Fig.3b Cone Beam CT – Coronal View, Showing Expanded Buccal And Labial Cortices

The patient was prepared for enucleation of the cystic lesion along with a reconstruction with iliac crest graft, in case of a pathological fracture of the mandible. Patient underwent enucleation of the cyst along with the impacted lower right 2nd molar and extraction of lower right 1st molar under general anesthesia. The specimen- cystic lining was sent

for histopathological investigation (Fig 4-6). The lower border of the mandible was intact so bone grafting was deferred.

The histopathology of the specimen was reported to be a mural and luminal unicystic ameloblastoma.



Fig4. After The Mucoperiosteal Flap Was Raised, Buccal Cortex Was Thinned Out, Bulging Cyst Can Be Appreciated



Fig.5 Cyst Was Enucleated, Impacted Tooth Within Was Extracted.



Fig.6- Cystic Lining, Was Sent For Histopathology

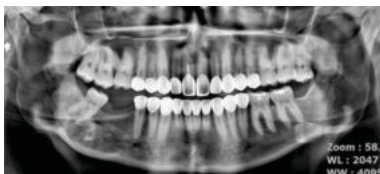


Fig.7- OPG At End Of 1 Year- No Recurrence

DISCUSSION

Unicystic ameloblastoma, as the name suggests is the kind of ameloblastoma that is cystic clinically, radiographically and grossly. It was first described by Robinson and Martinez in 1977.^{11,13}

The etiopathogenesis has been proposed by Leider et al, postulating a three way mechanism:

1) Ameloblastic transformation of reduced enamel epithelium in association with developing tooth, along with subsequent cystic development. 2) Development from neoplastic transition of non-neoplastic ameloblastic epithelium of dentigerous or other odontogenic cysts. 3) Cystic degeneration of ameloblastic islands in a solid ameloblastoma followed by fusion of multiple microcysts resulting in a unicystic lesion.¹⁴

In a clinicopathologic study of 57 cases of unicystic ameloblastoma, Ackermann classified this entity into the following three histologic groups:

Group I: Luminal UA (tumor confined to the luminal surface of the cyst)

Group II: Intraluminal/plexiform UA (nodular proliferation into the lumen without infiltration of tumor cells into the connective tissue

wall), and

Group III: Mural UA (invasive islands of ameloblastomatous epithelium in the connective tissue wall not involving the entire epithelium).⁵

A more recent Histopathological classification by Philipsen and Reichart^{3,15} is:

subgroup 1-luminal UA;

subgroup 1.2-luminal and intraluminal;

subgroup 1.2.3-luminal, intraluminal and intramural; and

subgroup 1.3-luminal and intramural

The different treatment modalities being practiced over the years, which we found during our literature review was very varied and vast.

There are multiple surgical treatment modalities advocated in the literature- including enucleation and curettage with or without the use of Carnoy's solution, marsupialization and resection¹⁸.

The treatment modality depends on the histological type, according to the above classifications. The UAs diagnosed as subgroups 1 and 1.2 can be treated conservatively (careful enucleation), whereas subgroups 1.2.3 and 1.3 showing intramural growths require treated radical resection, as for a solid or multicystic ameloblastoma.³

According to Marx and Stern, invasive ameloblastoma arising from the epithelial lining and proliferating through the complete thickness of the connective tissue layer of a cyst should be treated with resection, and ameloblastoma "in situ" developing in, which is limited to the epithelial lining of a cyst, microinvasive ameloblastoma arising from the epithelial lining and proliferating into the connective tissue layer of the cyst, enucleation can be done.¹⁵

Gardner in his paper has described the treatment based on anatomical and pathological considerations, and has recommended curettage.¹⁶

Lau et al in their review noted that usually larger unicystic ameloblastomas are treated by enucleation and chemical cauterization with Carnoy's solution.^{18,9}

Ogunsalu has described a very novel surgical modality of re-entry surgery in large unicystic ameloblastoma with liquid nitrogen application which might possibly lower the recurrence rate.¹⁹

Khare et al employed a modified pedicled osteoperiosteal flap in large mandibular unicystic ameloblastomas. They have claimed the technique to facilitate healing, resists infection with a flap that has potential for early bony regeneration thus preventing collapsing of the facial soft tissues.²⁰

Girardi et al conclude from their retrospective analysis that unicystic luminal and intra luminal ameloblastomas should undergo enucleation with peripheral osteotomy followed by application of Carnoy's solution.⁸

In the cases series by Meshram et al, they have reported that the conservative surgical treatment of mandibular ameloblastoma in young patients have yielded excellent postoperative function and esthetics with no recurrence.⁹

The ameloblastomas in general, are known to show recurrence. The potential for recurrence has three main contributing factors- macroscopic appearance of the cyst¹⁶, treatment modality^{12,18} and the histopathology.^{18,23}

Cystic variant of ameloblastoma is biologically less aggressive and has a better response to enucleation or curettage than does the solid ameloblastoma with reference to recurrence.^{16,17,22}

In spite of all the studies supporting less recurrence with enucleation as the treatment modality for UA, due to its aggressive nature, a long term follow up is recommended and stressed by many in the literature.^{9,12,13,21}

The relatively high recurrence rate of this benign tumor makes it mandatory to have a regular and long follow with clinical and radiological examinations.^{9,12,21}

In the present case, the cystic tumor with an impacted tooth within, a clinical diagnosis of dentigerous cyst was made. A rational decision

needs to be taken to decide on the surgical modality based on the patients' age, size of the tumor, location, duration and patient's availability for follow up.^{8,16} Since the patient was young, and the size of the cyst small, enucleation was planned. As the histopathology revealed to be mural and luminal unicystic ameloblastoma, she is planned for a long term follow up. At 1 year post operative follow up there was no clinical or radiological signs of recurrence Fig.7.

CONCLUSION

The UAs mimic odontogenic cysts clinically, radiographically and macroscopically; the only difference being histopathology. Due to this similarity the surgical treatment usually will be enucleation in both the scenarios.

Therefore, we conclude that UA is a histopathological diagnosis. This also brings us to another important fact that all surgical specimen should be sent for histopathology. As, it might change the follow up plan of the patient viz., long term follow up for ameloblastomas to catch the recurrences early, in case any- which will further reduce the morbidity associated with the second surgery.

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