ISSN: 2581-5989
PubMed - National Library of Medicine - ID: 101738774

International Journal of Dental Science and Innovative Research (IJDSIR)

IJDSIR : Dental Publication Service

Available Online at: www.ijdsir.com

Volume – 5, Issue – 1, February - 2022, Page No. : 64 - 70

Unicystic Ameloblastoma of Maxilla misdeemed as a Dentigerous Cyst: A Unique Case Report of a 9year-old child

[1]Dr. Keerthi R, MDS, Professor, Department of Oral and Maxillofacial Surgery, V S Dental College & Hospital, KR Road, VV Puram, Bengaluru – 560

[1]Dr. Ashwin D P, MDS, Professor, Department of Oral and Maxillofacial Surgery, V S Dental College & Hospital, KR Road, VV Puram, Bengaluru – 560

[2]Dr. Monica Roy Chandel, BDS, Post-graduate Student, Department of Oral and Maxillofacial Surgery, V S Dental College & Hospital, KR Road, VV Puram, Bengaluru – 560

[2]Dr. Priyanka Umarani, BDS, Post-graduate Student, Department of Oral and Maxillofacial Surgery, V S Dental College & Hospital, KR Road, VV Puram, Bengaluru – 560.

Corresponding Author: Dr. Monica Roy Chandel BDS, Post-graduate Student, Department of Oral and Maxillofacial Surgery, V S Dental College & Hospital, KR Road, VV Puram, Bengaluru – 560.

Citation of this Article: Dr. Keerthi R, Dr. Ashwin D P, Dr. Monica Roy Chandel, Dr. Priyanka Umarani, "Unicystic Ameloblastoma of Maxilla misdeemed as a Dentigerous Cyst: A Unique Case Report of a 9year-old child", IJDSIR- February - 2022, Vol. – 5, Issue - 1, P. No. 64 – 70.

Type of Publication: Case Report

Conflicts of Interest: Nil

Abstract

Maxillofacial skeleton tumours can be classified as either odontogenic or non-odontogenic and benign or malignant. Ameloblastoma is the most common benign aggressive tumor among other odontogenic tumors, accounting for 1% of tumors and cysts of the jaw and 10% of odontogenic tumors overall. The incidence of ameloblastoma is approximately 80% in the molar and ramus region of the mandible, while 20% occur in the maxillary posterior region.

Unicytic ameloblastoma is among one of the variants of ameloblastoma encasing about 6% of ameloblastomas. They're represented as cystic lesions that reveal clinical and radiographic features of a cyst, but the histopathological features are demonstrated a typical ameloblastomatous epithelium lining the cyst.

This report presents a case of unicystic ameloblastoma in an uncommon location in a 9- year-old child. The lesion was initially thought to be a dentigerous cyst, based on its location and radiographic appearance. In this article, the clinical and radiographic features, histopathology and treatment of unicystic maxillary ameloblastoma are reviewed with an added emphasis on a literature review of ameloblastoma in pediatric patients.

Keywords: dentigerous cyst, maxilla, odontogenic tumour, pediatric, unicystic ameloblastoma.

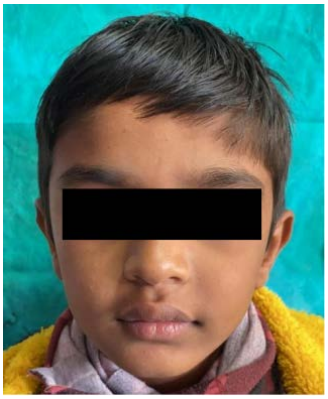
Introduction

Ameloblastoma is the most common tumor of the odontogenic epithelium, representing 1% of all oral odontogenic epithelial tumors and 10% of all odontogenic tumors. Ameloblastomas are persistent, grow slowly, locally invasive and demonstrate benign growth characteristics.

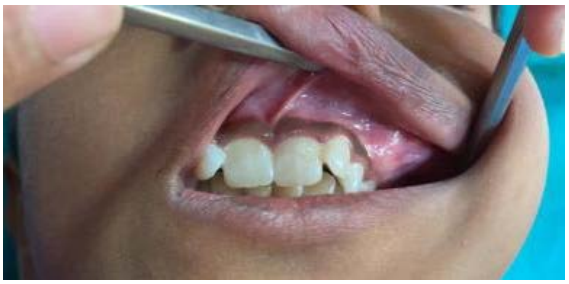
According to 2005 World Health Organization histological classification, Ameloblastomas are classified as: conventional, solid or multicystic, unicystic, peripheral (extraosseous) and desmoplastic.[1] Unicystic ameloblastoma is a distinguishable entity of ameloblastomas, characterized by slow growth and being relatively locally aggressive.[2] They have unilocular or multilocular radiolucencies radio graphically with a soap-bubble or honeycombed appearance. Rarely they resemble a dentigerous cyst, appearing as a circumscribed radiolucency surrounding the crown of an unerupted tooth. Resorption of the adjacent tooth is not uncommon finding. Hence, diagnosis can be confirmed through the radiographic appearance of the lesion, clinical behavior and most definitively, biopsy of the lesion.[1]

Case Report

A 9-year-old boy presented with the chief complaint of swelling in the upper posterior tooth region since 15 days, followed by subsequent development of pain. Clinical examination revealed swelling in the left cheek region that was intraorally extending from the primary canine up to the primary second molar area, being more prominent in the buccal sulcus and alveolar process (Fig. 1).



(A)



(B)

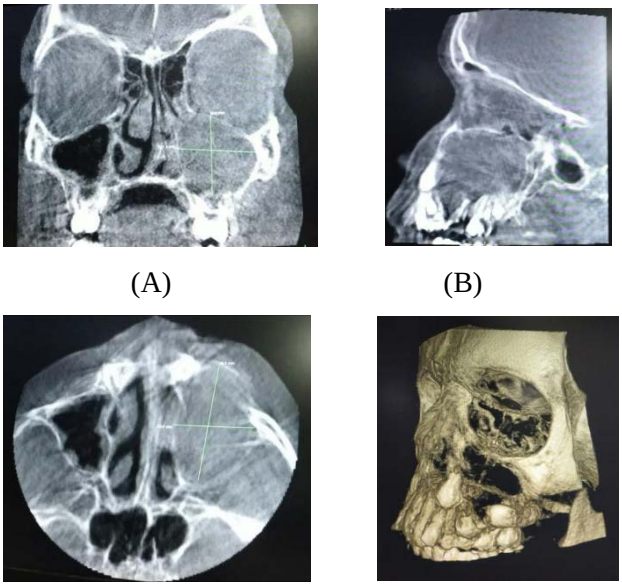
Fig.1: Preoperative clinical images (A. Extra-oral and B. Intra-oral).

Radiographic Examination

In the CBCT sections, coronal section revealed ballooning expansion measuring 27.3 X 26.9 mm, involving superiorly : the floor of the orbit, medially : lateral wall of the nose, laterally : lateral wall of sinus and inferiorly : floor of the maxillary sinus. Thinning of left floor of orbit and left lateral wall of the nose was seen.

Obliteration of middle concha present along with complete haziness in the left maxillary sinus (Fig. 2A). Saggital section revealed a tooth like structure placed vertically at the level of floor of the nose above the apices of 65. Dome shaped expansion of labial cortial plate was present extending anteroposteriorly from labial cortex to posterior maxilla and superoinferiorly from floor of the orbit to the alveolar process of maxilla (Fig. 2B).

Axial section revealed labial and buccal cortical plate expansion along with thinning of the floor of the orbit and lateral wall of the nose. Medial displacement of lateral wall of the nose was seen (Fig. 2C). Solid reconstruction image of maxilla revealed a labially placed 23 at the floor of the nose. Slight breach in the orbital floor was present. Also labially rotated teeth i.e 25 was seen (Fig. 2D).



(A) (B)

(C) (D)

Fig.2: Preoperative CBCT (A. Coronal Section, B. Saggital Section, C. Axial Section, D. Solid Reconstruction).

OPG revealed mixed dentition, vertically erupting 23 showing loss of dental follicle surrounding the tooth. Distal aspect of 23 revealed an ill-defined radiolucency arising laterally from the CEJ extending till the root apices of 62,63 and 64. Sinus floor cannot be appreciated on the left side. Slight upward displacement of sinus floor was seen. Nasal floor obliteration and transposition of tooth wrt 25 observed. (Fig. 3).

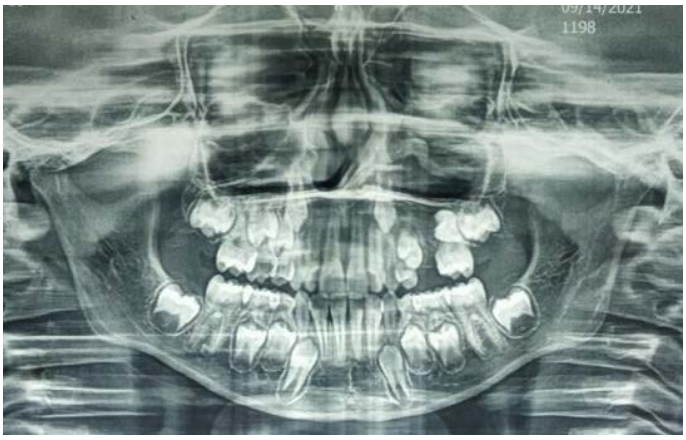
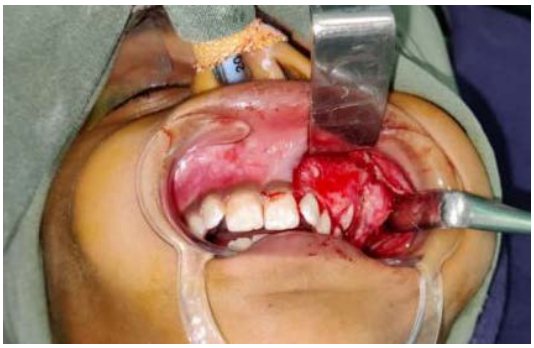


Fig.3: Preoperative OPG.

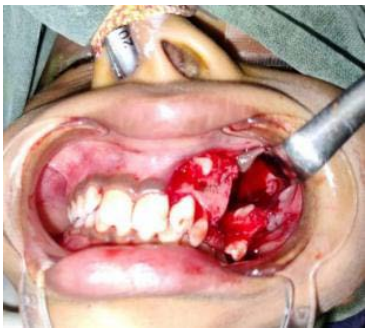
All of this pointed towards the radiographical diagnosis of benign odontogenic cyst, most probably dentigerous cyst.

Surgical Procedure

Under general anaesthesia enucleation was performed in the upper left molar region. The cystic lining was separated from the bone using curette and was removed along with 25 in toto. After this 64 and 24 were extracted followed by primary closure (Fig. 4).



(A)



(B)

(C)

Fig.4: Intra operative pictures (A. Exposure of the surgical site, B. After enucleation, C. excised specimen along with 25.

After histopathological examination of the excised specimen, it was found to be the unicystic ameloblastoma of mural variant (Fig. 5).

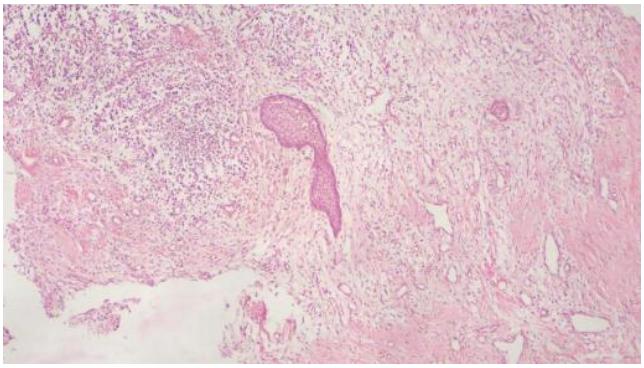


Fig.5: Histopathological slide of Unicystic Ameloblastoma of Mural variant

Postoperatively he was consulted for radiotherapy but was deferred because of his young age and chances of mucositis, radiation caries, myelosuppresion, alopecia etc. The patient was then followed up for one year with no signs of recurrence (Fig. 6 and 7).

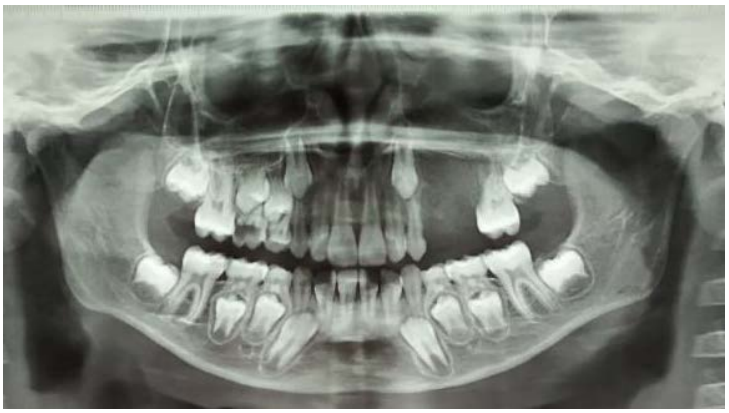
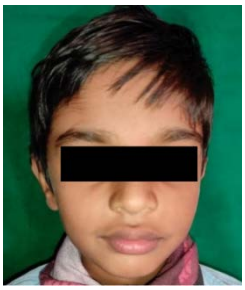
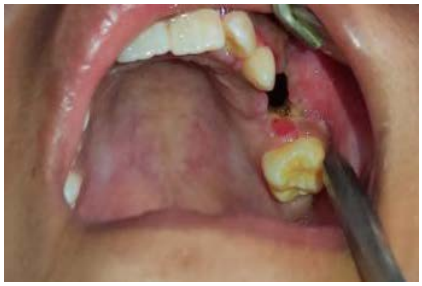


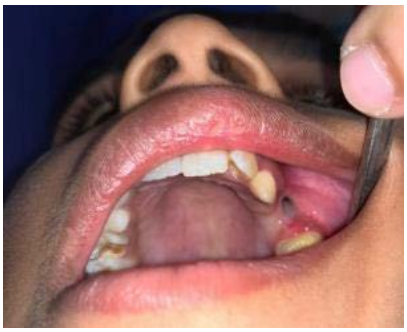
Fig.6: Postoperative OPG.



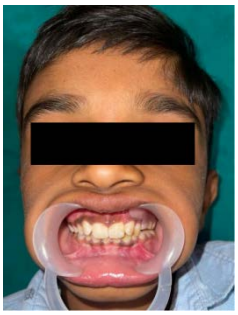


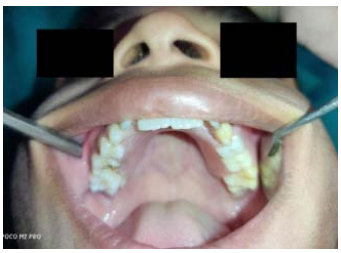
(A) (B)

A. After one week, B. After two weeks.



C. After One Month





(D) (E)

D. And E. After insertion of Removable Partial Denture

Fig.7: Follow up.

Discussion

Ameloblastoma is rarely found in children, accounting for approximately 10-15% of all reported cases of ameloblastoma. Approximately 80%-85% of ameloblastomas occur in the molar and ramus region of the mandible, followed by the mandibular symphyseal area. The remaining 15%-20% of cases occur in the maxilla, usually in the posterior region. Ameloblastoma in the maxilla may extend into the maxillary sinus and nasal floor.

First described by Robinson and Martinez in 1977, unicystic ameloblastoma is a rare variant of ameloblastoma, and is referred to those cystic lesions that show clinical and radiological characteristics of an odontogenic cyst but in histological examination it shows a typical ameloblastomatous epithelial lining part of the cystic cavity, with or without luminal and/or mural tumor proliferation. Several histologic subtypes of unicystic ameloblastoma are recognized, based on the character and extent of tumor cell proliferation within the cystic wall, which include those of simple cystic nature, those with intraluminal proliferation nodules, and those containing infiltrative tumor islands in the cystic walls.[2]

There is commonly a delay in recognition of the maxillary ameloblastoma, because of predominantly painless and slow growth, lack of a thick cortical plate, the plentiful cancellous bone and the proximity of the maxilla to the nasal cavity, nasopharynx, paranasal sinuses, orbits and skull base. The extension of ameloblastoma into these structures itself provides useful diagnostic evidence.

The abundant blood supply of the maxilla provides another possible mode of spread. There have been incidences of invasive maxillary ameloblastomas with extension into the orbit, frontal sinus, skull base, middle cranial fossa and petrous apex resulting in the death of the patient. A painless swelling of the involved part of the jaw is the most common clinical symptom of the maxillary ameloblastoma. Ameloblastoma is an osteolytic lesion in which production of mineralized components is a rarity. When the maxillary sinus and surrounding structures are involved, opacification of the sinus and expansion of its walls with or without bone destruction makes it impossible to distinguish ameloblastoma from other malignant and invasive tumours like craniopharyngiomas.[3]

Clinical diagnosis of UA is a challenge. It is frequently misdiagnosed considering their similarity in the clinical and radiographic features to that of odontogenic cysts, most commonly a dentigerous cyst. Hence, histopathologic confirmation is mandatory to arrive at a final diagnosis .

Ackerman in 1988 classified UA on the basis of histopathologic features as.

Group I: Luminal UA where the tumor is confined to the luminal surface of the cyst.

Group II: Intraluminal UA when there is nodular proliferation into the lumen without infiltration of tumor cells in the connective tissue wall.

Group III: Mural UA where there is presence of invasive islands of tumor cells in the connective tissue wall of the cyst.

This classification has a direct bearing on their biological behavior, treatment, and prognosis.[5] Our case falls under Group III as mural variant of UA.

The behaviour of mural variant is quiet similar to that of conventional solid ameloblastoma with a propensity for recurrence after enucleation because of the presence of tumor cells in the fibrous tissue capsule. However, there is no clarification on whether the mural invasion can extend beyond the capsule to the adjacent bone.

The treatment of UA is a controversy considering whether it can be conservative or radical. Conservative treatment comprises of enucleation with or without curettage and marsupialization followed by enucleation. Adjunctive therapy including thermal or chemical cauterization, cryotherapy and radiotherapy can be employed following primary treatment. On the other hand, radical treatment involves segmental or marginal resection of the lesion followed by placement of reconstruction plates whenever required.[5]

The treatment of unicystic ameloblastoma is very controversial and is because of the misapplication of the term "mural". The term "mural" describes the extent to which amelobastomatous changes penetrate the connective tissue layer of a cyst. A mural ameloblastoma does not penetrate the epithelial lining of a cyst similar to mural painting that covers only the surface of a wall.[2]

The recurrence rate post treatment determines the effectiveness of a particular treatment modality. The rate of recurrence is lesser with radical treatment when compared to a more conservative approach.

The recurrence rate after treatment of UA ranges from 10%- 25%. Lau and Samman studied the recurrence rate in UA following various treatment modalities and observed the recurrence rate was 30.5% (highest) following enucleation alone, 18% for marsupialization, 16% for enucleation followed by application of Carnoy's solution and 3.6% (lowest) for resection.[5]

A relatively conservative treatment like enucleation with peripheral ostectomy is recommended in the first instance, according to the published reports, reserving a more aggressive therapy for any recurrence considering the growth potential and capacity for bone regeneration in children.[6]

Even though low recurrence rate is reported following resection, radical treatment is avoided in children for the following reasons:

(1) Continuing facial growth in children and presence of a highly reactive periosteum

(2) Presence of Unerupted permanent teeth

(3) May cause disfigurement and masticatory issues which can be psychologically disturbing to the child. A conservative line of treatment is the best option for pediatric and adolescent patients since it is associated with faster bone fill and restoration of normal bony architecture.[6]

The treatment for ameloblastoma in children includes considerations such as: age, site, size, clinical type, subdivision of the cystic type, the patient's wishes, compliance and understanding, projected recurrent condition and rate, physical and psychological impacts, and also development of new materials and surgical techniques. The risk-benefit ratio of the surgeries should be explained, so the patient and parents have a clear understanding to make the decision. Recurrence is probably not the most important consideration for children, and should not be considered as equivalent to failure.[4]

In general, ameloblastoma is considered to be a radioresistant tumour, although recurrence has been reported after irradiation and can be performed in cases when surgery is not an option. Megavoltage irradiation alone or in combination with surgery has given satisfactory results, especially in large maxillary cases, in order to diminish the volume of the tumour.[4] Contrarily, Atkinson et al in 1984 cited that properly applied megavoltage radiation techniques have a useful role in the management of ameloblastoma, especially where a full surgical excision would be technically difficult, the reason being it's bulk and local invasion or

where other medical factors, including age, that would make radical surgery inappropriate. Though there were some variations of radiation technique in terms of dose and time, a total dose of 4500 rad in 4 weeks would seem appropriate. The megavoltage technique chosen should be implemented according to the particular site of involvement.

For example, a maxillary antral invasion will include elaborate techniques to encompass all the bony margins including the floor of the orbit whilst sparing vital structures such as the anterior chamber of the eye. Whereas in the case of mandibular lesions, the whole mandible should be irradiated.[7]

Chemotherapy does not seem to be effective at the present time, when used independently, not withstanding the variety of agents, schedules and routes of administration that have been reported.

A case of an advanced maxillary ameloblastoma is reported to have been successfully treated with the combination of chemotherapy and radiotherapy, perhaps this arena requires more investigation.[4] The determination of the most appropriate treatment can also be determined by the histological type of ameloblastoma present in the maxilla. For example, the unicystic type has a low recurrence rate even after simple enucleation but this type has been rarely found in maxilla. Instead, 50 per cent of the desmoplastic type, despite the very low incidence of this type, as well as maxillary ameloblastomas, has been found predominantly in the anterior and posterior region of the maxilla. This variant has a very low recurrence rate, although differences in behaviour or prognosis among the various types in maxillary ameloblastomas have not been found.[3]

However, radiotherapy in pediatric patients is still a big question. It's side effects such as alopecia, mucositis, dry mouth, radiation caries, osteoradionecrosis, altered growth and development etc. can't be ignored.

Conclusion

To conclude the mural variant of UA has a high potential for recurrence when compared to other types of UA. Radical Surgery can be opted in adult patients with mural UA but the same treatment protocol cannot be applied to the pediatric population since resection may cause an alteration in craniofacial development leading to functional and esthetic damage which can directly affect their quality of life. Hence, in pediatric age group a more conservative approach of enucleating the lesion seems to be a promising treatment modality of UA with a long-term regular follow-up. However, this warrants confirmation by future studies.

Patient Consent The patient's attender was provided written informed consent for the publication and the use of his images.

References

1. Sangeetharaj Sheela, Steven R. Singer, Hani F. Braidy, Albert Alhatem, Adriana G. Creanga. Maxillary ameloblastoma in an 8-year-old child: A case report with a review of the literature. Imaging Science in Dentistry2019;
49:2419https://doi.org/10.5624/isd.2019.49.3.241

2. Natália Galvão Garcia, Denise Tostes Oliveira and Moacyr Tadeu Vicente Rodrigues. Unicystic Ameloblastoma with Mural Proliferation Managed by Conservative Treatment. Volume 2016, Article ID 3089540,pageshttp://dx.doi.org/10.1155/2016/3089540.

3. S. Iordanidis Ch. Makos J. Dimitrakopoulos H. Kariki. Ameloblastoma of the maxilla. Case report. Australian Dental Journal 1999;44:(1):51-55.

4. I-Yueh Huang, Sheng-Tsung Lai, Chung-Ho Chen, Chun-Ming Chen, Chung-Wei Wu, and Yee-Hsiung Shen. Surgical management of ameloblastoma in

children. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2007; 104:478-85.

5. Marimuthu L, Kumar S, Shenoy V, et al. (December 07, 2020) Mural Variant of Unicystic Ameloblastoma in a Pediatric Patient: A Rare Case Report. Cureus 12(12): e11963. DOI 10.7759/cureus.11963.

6. Neelam N. Andrade, Shweta P. Shetye, Tejas S. Mhatre. Trends in Pediatric Ameloblastoma and its Management: A 15-year Indian Experience J. Maxillofac. Oral Surg. (Jan- Mar 2013) 12(1):60–67. DOI 10.1007/s12663-012-0387-1.

7. Chris H. Atkinson, Andrew R. Harwood, Bernard J. Cummings. Ameloblastoma of the Jaw A Reappraisal of the Role of Megavoltage Irradiation. Cancer 53:869-873, 1984