

Unicystic Ameloblastoma with Varying Histological Patterns Managed Conservatively

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Abstract

Introduction: Unicystic ameloblastoma (UCA) is a variant of ameloblastoma and accounts for 6% of all ameloblastomas. It arises from pre-existing odontogenic cysts or de novo. Rarely, does UCA present with diverse histopathologic patterns.

Case Presentation: We present a case of UCA that was treated conservatively which presented with a mixture of histological patterns.

Management and Prognosis: A 12-year-old male patient complained of swelling in his left mandibular region. Radiographic investigation showed multilocular radiolucency. On histopathologic examination, a diagnosis of unicystic ameloblastoma was made. A conservative treatment option was undertaken for the patient.

Conclusion: Unicystic ameloblastoma of the mural type require resection but given the young age of the patient, a conservative approach was planned and the patient responded well to the treatment.

Keywords: Unicystic ameloblastoma, Pediatric oncology, Cyst enucleation

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Introduction

Swellings in the oral cavity are caused by a range of factors, including infections, local sepsis and most commonly, odontogenic and non-odontogenic benign and malignant lesions. One of the most common aggressive benign tumors of the jaw is Ameloblastoma, which presents as a slow-growing, painless mass and sometimes presents as an incidental finding. While Ameloblastomas can be of several types, Unicystic ameloblastoma (UCA) is a distinct entity and is distinguished by numerous growth patterns but seldom do they exist within a single lesion. The origin of UCA is still debated, but some authors regard it as a lesion arising de novo.^{1,2,3,4} Treatment of pediatric patients with UCA is still not fully understood and aggressive treatments are not advised as they can have adverse effects on their developing bodies and minds. Additionally, the therapy of UCA is still quite debatable if presented with a varying histopathologic pattern, making it difficult to determine the best course of action for this tumor.⁵ This report presents the case of mandibular UCA in a 12-year-old boy who was successfully treated by enucleation.

Case Report

A 12-year-old male patient presented with a chief complaint of swelling in the lower left side of the face for the past one week. On extraoral examination, the patient had a firm swelling in relation to the left lower side of the face which was tender on palpation. On intraoral examination, a 4x3 cm diffuse firm swelling with a smooth surface was noted in relation to the left side of the mandible extending from 33

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teeth region up to the retromolar region and was tender on palpation.

Panoramic radiograph revealed a unilocular radiolucency extending from 36 tooth region to the ascending ramus of the mandible involving the 38-tooth germ (Figure 1A). Advanced imaging modalities, three-dimensional computed tomography revealed an expansile multiloculated lesion in the mandible with expansion and thinning of buccal and lingual plates (Figure 1B). Based on Clinical and Radiographic findings a presumptive diagnosis of Dentigerous cyst and Odontogenic keratocyst was made.

An incisional biopsy was performed which revealed cystic cavity lined by ameloblastic epithelium and stellate

reticulum-like cells and a diagnosis of UCA. After which the patient underwent enucleation and chemical cauterization of unicystic ameloblastoma in relation to the left side of the mandible. Carnoy's solution - dipped cotton pellet was placed for a few minutes; saline irrigation was done and sutures were placed. The impacted 38,37,35 teeth were extracted.

Formalin-fixed Hematoxylin and Eosin-stained multiple sections representing all areas were thoroughly analyzed. Microscopically cystic cavity lined by ameloblast like tall columnar cells having peripheral palisading hyperchromatic nuclei. Intraluminal proliferation of ameloblast-like cells was found arranged in a plexiform pattern and mural proliferation was also seen (Figure: 2A,2B). The underlying fibrous capsule revealed various-sized islands of odontogenic epithelium. (Figure: 2C,2D).

In the present case, the connective tissue stroma and the lumen of the cyst displayed microscopic epithelial growth. A final diagnosis of UCA with mural proliferation was made. Most authors advise having a surgical resection as a result of this diagnosis. According to Marx and Stern, enucleation is one successful treatment for UCA.⁶ Additionally, the patient's age, clinical characteristics, and radiologic features were taken into account; as a result, enucleation was the preferred course of action.

Discussion

Churchill first used the term ameloblastoma in 1934.⁷ Ameloblastomas have the potential to proliferate into larger sizes and have the capability of severely eroding and destroying local bone. Ameloblastoma has been detected in a variety of forms, UCA being one separate entity.^{8,9} On basis of their clinical presentation, they can arise in three forms namely, conventional, UCA and peripheral ameloblastoma. UCA which was first described by Robinson and Martinez

in 1977, is a lesion that can resemble an odontogenic cyst clinically and radiographically but microscopically can have a typical ameloblastic epithelium lining the cyst. Unicystic ameloblastoma (UA) is a rare entity that makes up around 6% of all ameloblastomas.

They are more commonly found to affect the young age groups; with an incidence of 50% mainly in the second and third decade of life with a slight male predilection [1.6:1].^{10,11} The peak incidence of UA was observed to occur between the ages of 11 and 20 years, with 89% of cases occurring in the mandible.¹² Impacted teeth are linked in 50-80 % of instances, with the third molars being the most common.³ Radiographically it can present with assorted presentations but frequently is seen as a unilocular radiolucency.^{13,14} It is uncommon to have a multilocular radiography pattern with an impacted third molar, yet this was noted in the current case. More often it can resemble a Dentigerous cyst or an Odontogenic keratocyst radiographically and is considered a differential diagnosis. According to one study, UA may be distinguished from dentigerous cysts and radicular cysts using the expression of the BRAFV600E protein and the detection of the BRAFV600E mutation.¹⁵

The unicystic ameloblastoma was divided into three distinct entities by Ackerman and Shear as luminal, intraluminal/plexiform and mural UCA. Further classifications were given by Philipsen and Reichart into four subclasses, which are 1: luminal UA; 1.2: luminal and intraluminal; 1.2.3: luminal, intraluminal and intramural; and 1.3: luminal and intramural.

The luminal and intraluminal types may be successfully treated by enucleation or curettage; however, it has been suggested that the third group of lesions, the mural type where recurrence after conservative surgery is more likely to occur, should be treated by radical resection.^{3,16} According to Marx

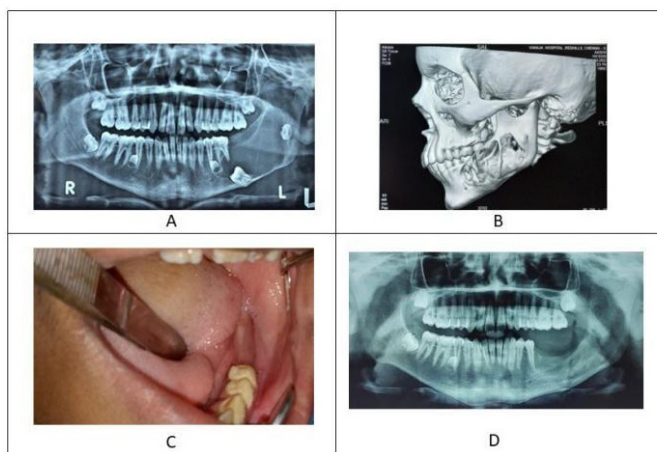


Fig. 1: **A:** Unilocular radiolucency extending from 36 tooth region to the ascending ramus of the mandible involving the 38-tooth germ. **B:** 3D CT reveals a cavity in relation to the lower left back tooth region with bony expansion and perforation. **C & D:** Postoperative radiograph shows favorable prognosis and good bone formation.

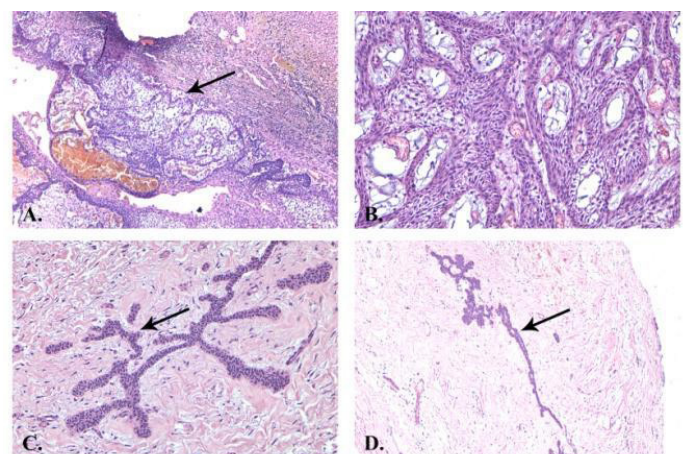


Fig. 2: **A:** Cyst lined by ameloblastic epithelium showing features of an intraluminal plexiform pattern (arrow). Underlying connective tissue shows chronic inflammatory cell infiltration. H & E 10X. **B:** Plexiform type of Ameloblastic epithelium. H & E 10X. **C:** Photomicrograph displaying features of strands of mural islands (Arrow) in the connective stroma. H & E 10X. **D:** Photomicrograph displaying a focal area of mural island (Arrow) in the connective tissue stroma. H & E 4X

and Stern, enucleation should be used to treat ameloblastoma "in situ," which develops in and is restricted to the epithelial lining of a cyst, as well as ameloblastoma "micro-invasive," which arises from the epithelial lining and proliferates into the connective tissue layer. However, ameloblastoma that is "invasive" and develops from the epithelial lining of a cyst and spreads over the entire thickness of the connective tissue layer should be removed.⁶ Several authors agree that vigorous treatment of ameloblastoma is necessary to prevent recurrences. An aggressive surgical approach in juvenile patients may cause a number of issues, including dysfunction and deformity. The patient's age must be always kept in mind while deciding the treatment plan.¹⁷ Our case demonstrated a favourable prognosis with conservative treatment, and good bone formation (Figure 1C, 1D) after 8 months period of follow-up that favoured the avoidance of radical dissection in a young patient.

Conclusion

Histologically, the minimal requirement for classifying a lesion as UCA is the presence of a single cystic space lined with odontogenic (ameloblastomatous) epithelium. The occurrence of UCAs with such diverse histology still remains unclear and requires further research. In the treatment aspect of such cases, the patient's age, clinical characteristics, radiologic features and histopathologic diagnosis should be taken into account. Despite the possibility of recurrence, the conservative surgical approach is the best course of action in children, where structural and functional integrity is preserved.

References

- Ackermann GL, Altini M, Shear M. The unicystic ameloblastoma: a clinicopathological study of 57 cases. *J Oral Pathol* [Internet]. 1988;17(9-10):541-6. Available from: <http://dx.doi.org/10.1111/j.1600-0714.1988.tb01331.x>
- Vickers RA, Gorlin RJ. Ameloblastoma: Delineation of early histopathologic features of neoplasia. *Cancer* [Internet]. 1970;26(3):699-710. Available from: [http://dx.doi.org/10.1002/1097-0142\(197009\)26:3<699::aid-cncr2820260331>3.0.co;2-k](http://dx.doi.org/10.1002/1097-0142(197009)26:3<699::aid-cncr2820260331>3.0.co;2-k)
- Gardner DG. Some current concepts on the pathology of ameloblastomas. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* [Internet]. 1996;82(6):660-9. Available from: [http://dx.doi.org/10.1016/s1079-2104\(96\)80441-0](http://dx.doi.org/10.1016/s1079-2104(96)80441-0)
- Fedhila M, Belkacem Chebil R, Karray S, Sriha B, Oualha L, Douki N. Unicystic ameloblastoma mimicking lateral periodontal cyst. *Case Rep Dent* [Internet]. 2022;2022:8197837. Available from: <http://dx.doi.org/10.1155/2022/8197837>
- Lang M-J, Hwang M-J, Jin Y-T, Chiang C-P. Histopathological study of a mixed intraluminal and mural type of unicystic ameloblastoma. *J Dent Sci* [Internet]. 2022;17(1):573-5. Available from: <http://dx.doi.org/10.1016/j.jds.2021.07.019>
- Siriwardena BSMS, Tennakoon TMPB, Hunter KD, Tilakaratne WM. Unicystic ameloblastoma: Analysis of 370 cases in a single center in Sri Lanka. *J Oral Pathol Med* [Internet]. 2018;47(7):706-9. Available from: <http://dx.doi.org/10.1111/jop.12740>
- Iordanidis S, Makos C, Dimitrakopoulos J, Kariki H. Ameloblastoma of the maxilla. Case report. *Aust Dent J* [Internet]. 1999;44(1):51-5. Available from: <http://dx.doi.org/10.1111/j.1834-7819.1999.tb00536.x>
- James L, Shafath A, Nagaraj T, Biswas A. Unicystic ameloblastoma of mural variant crossing the midline. *Natl J Maxillofac Surg* [Internet]. 2022;13(Suppl 1):S199-202. Available from: http://dx.doi.org/10.4103/njms.NJMS_21_19
- Black CC, Addante RR, Mohila CA. Intraosseous ameloblastoma. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* [Internet]. 2010;110(5):585-92. Available from: <http://dx.doi.org/10.1016/j.tripleo.2010.02.040>
- Ramesh RS, Manjunath S, Ustad TH, Pais S, Shivakumar K. Unicystic Ameloblastoma of the mandible - an unusual case report and review of Literature - *Head & Neck Oncology*. BioMed Central. 2010;2.
- Reichart PA, Philipsen HP. *Odontogenic tumors and allied lesions*. Hanover Park: Quintessence Publishing Inc; 2004.
- Marx RE, Stern D. *Oral and Maxillofacial Pathology: A Rationale for Diagnosis and Treatment*. Odontogenic tumors: hamartomas and neoplasms. 2012;671-8.
- Nagalaxmi V, Sangmesh M, Maloth KN, Kodangal S, Chappidi V, Goyal S. Unicystic mural ameloblastoma: an unusual case report. *Case Rep Dent*. 2013.
- Agani Z, Hamiti-Krasniqi V, Recica J, Loxha MP, Kurshumliu F, Rexhepi A. Maxillary unicystic ameloblastoma: a case report. *BMC Res Notes* [Internet]. 2016;9(1):469. Available from: <http://dx.doi.org/10.1186/s13104-016-2260-7>
- Pereira NB, Pereira KMA, Coura BP, Diniz MG, de Castro WH, Gomes CC, et al. BRAFV600E mutation in the diagnosis of unicystic ameloblastoma. *J Oral Pathol Med* [Internet]. 2016;45(10):780-5. Available from: <http://dx.doi.org/10.1111/jop.12443>
- Garcia NG, Oliveira DT, Rodrigues MT. Unicystic Ameloblastoma with Mural Proliferation Managed by Conservative Treatment. *Case Rep Pathol*. 2016;3089540.
- Guo T, Zhang C, Zhou J. Unicystic ameloblastoma in a 9-year-old child treated with a combination of conservative surgery and orthodontic treatment: A case report. *Clin Case Rep* [Internet]. 2022;10(1):e05241. Available from: <http://dx.doi.org/10.1002/ccr3.5241>

