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### Treatment Modalities of Unicystic Ameloblastoma - A Case Series

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#### ABSTRACT

Unicystic ameloblastoma is a unique variant of conventional ameloblastoma which has considerably less recurrence rate than its counterpart. Unlike conventional ameloblastoma which mostly requires aggressive management, unicystic ameloblastoma has different treatment modalities which can be conservative or aggressive depending upon the histological nature of the tumor. We report five cases of unicystic ameloblastoma with different treatment modalities with long term follow up and satisfactory results.

**Keywords:** Ameloblastoma; Unicystic Ameloblastoma; Odontogenic Tumor; Enucleation; Resection; Maxillectomy.

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#### INTRODUCTION

One of the most common tumor of odontogenic origin is ameloblastoma which develops from epithelial cell remnant during development. This tumor is considered to be aggressive in nature and warrants aggressive treatment approach. It is reported to have high recurrence rate following conservative treatment. Unicystic ameloblastoma is a distinct variant of conventional ameloblastoma with considerably less recurrence rate. Unicystic ameloblastomas are less aggressive than the multicystic variant.<sup>[1,2]</sup> These lesions show clinical and radiological characteristic of an odontogenic cyst but histologically a typical ameloblastomatous lining part of the cyst cavity with or without luminal or mural proliferation is seen. The

goal of treatment of ameloblastoma is to achieve complete excision and appropriate reconstruction. However, for unicystic ameloblastoma, because of its propensity for less recurrence, conservative treatments like enucleation, curettage etc have been suggested. Aggressive variants demand radical treatment like resection. The choice of treatment depends on the histo-pathological picture as suggested by Ackermann et al<sup>[3]</sup> in 1988 who reclassified UA into three types with prognostic and therapeutic implications. Type I & II are usually managed by conservative treatment like enucleation and curettage, while type III lesion, due to aggressive nature necessitates an aggressive approach like resection. However, following conservative management long term follow up is advocated. We report five cases of unicystic ameloblastoma with different treatment modalities and long term follow up.

#### Case Series

##### Case 1

A 25 year old male patient reported to our department, with a swelling in the left angle region of the mandible. The swelling had gradually increase in size over a period of one year and was not associated with any pain, or discharge. On examination there was a marked distortion over the lower third of the face due to increased size of the swelling, extending from the lower border of the mandible to the ala -tragus line. Intra-oral examination revealed bicortical expansion extending from the lower left retromolar region to the second premolar region. Radiographic examination Figure 1 showed a unilocular expansile lesion with scalloped margins extending from 36 to 38 region. Resorption of roots was noted in relation to 36,

37 and impacted 38. The lower border was intact. Impacted 38 had migrated distally. A blood tinged straw coloured fluid was noted on aspiration. Incisional biopsy confirmed a unicystic ameloblastoma. Following Ackermann's criteria, the lesion was enucleated under general anesthesia along with removal of impacted tooth, followed by electrocauterization of the bony cavity Figure 2. The excised lesion was sent for histopathological examination which showed presence of cystic space lined by a thin epithelium overlying fibrocellular connective tissue stroma. The epithelium was non uniform with focal areas of thickening along with flat epithelial lining. The epithelium was 2-3 cell layer thick, cells of basal layer were columnar in shape with hyperchromatic nuclei, along with nuclear palisading and polarization. Proliferation at few places within the lumen was noted. Histological features were confirmatory of unicystic ameloblastoma. Patient has been followed up till date (31months) with no evidence of recurrence.



Figure 1: Preoperative OPG of Case 1

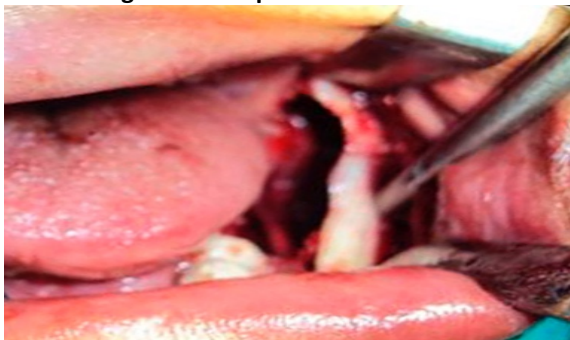


Figure 2: Intraoperative view of Case 1

#### Case 2

A 29 year old female patient reported to our department with a swelling in the lower anterior mandibular region of 1 week duration, associated with numbness of the lower lip on the same side. On examination, mild asymmetry of the chin was noted and intraorally a firm swelling extending from 32-37 region was noted with grade II mobility of 32-34. Radiographic examination Figure 3 showed unilocular radiolucency with sclerotic border, extending from 37 to 46 region, with associated root resorption of involved teeth on left side. A pale straw-coloured fluid was aspirated. Based on the clinical and radiographic findings provisional diagnosis of dentigerous cyst

was arrived at. Histopathological examination revealed intraluminal mass arising from the cyst wall. This major cystic space was lined by thin epithelium showing basal palisading and spongiotic changes, characteristics of unicystic ameloblastoma. The lesion was enucleated under general anaesthesia, and the cavity was filled with iodoform gauze packing to facilitate healing with secondary intention. Histopathological evaluation Figure 4 of the excisional biopsy specimen confirmed features of unicystic ameloblastoma. The patient has been followed till date (26 months) without any evidence of recurrence.

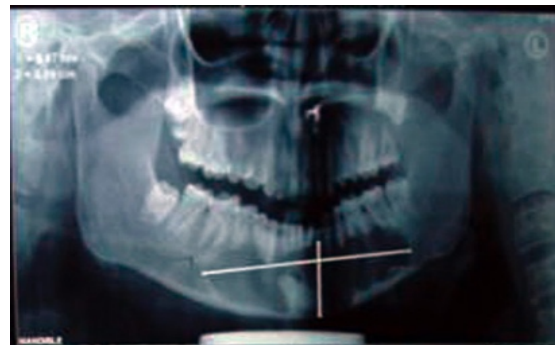


Figure 3: Preoperative OPG of Case 2

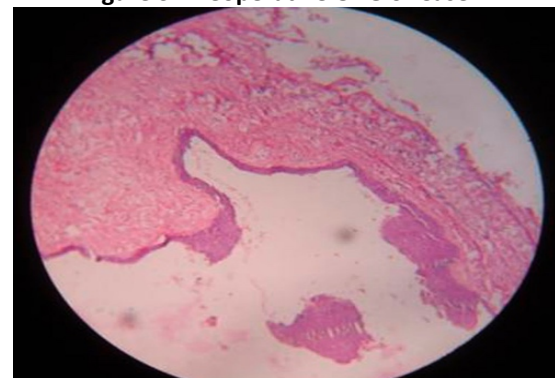


Figure 4: Histopathological Picture (10 X) of Case 2

#### Case no. 3

A 40 years old male patient reported to us with painful swelling of 11 month duration, in the left lower mandibular region extending from ear lobe to the angle of mandible. On examination there was gross asymmetry of the face, and intraorally grade III mobility of 36 was noted. Intraorally bicortical expansion was noted, extending from 35 to 37 region. Blood tinged pus like fluid was noted on aspiration.



Figure 5: Preoperative OPG of Case 3

Radiographic examination Figure 5 showed a large, well-defined unilocular radiolucency with scalloped margin extending from the 35 region to the level of sigmoid notch. Resorption of the roots of 35, 36 was noted. Incisional biopsy confirmed characteristic features of unicystic ameloblastoma. Considering the size and histopathological nature of the lesion based on Ackermann's criteria, marsupialisation was carried out Figure 6 under general anaesthesia. After marsupialisation, a pre-fabricated obturator was used to keep the window of the lesion open. Patient has been regularly reviewed till date (6 months) and there have been no signs of recurrence. Patient is due to have enucleation of the lesion in the near future.



Figure 6: Intraoperative view of Case 3

**Case no. 4**

A 30 year old female was referred to our department with a chief complaint of pain and swelling in the lower left mandibular region for 6 months. Extraorally, a swelling in the left mandibular angle region extending from the corner of mouth to the pre auricular region was noted. Intraorally a firm ulceroproliferative swelling extending from the left mandibular first molar area to the ascending ramus was noted.

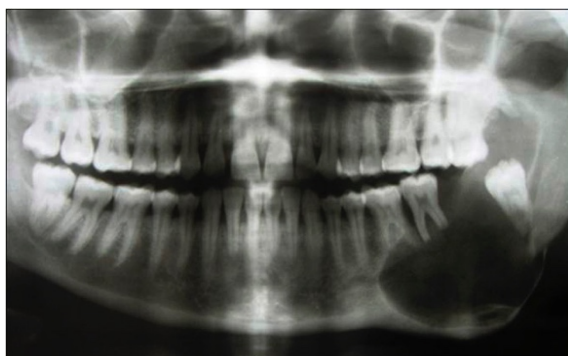


Figure 7: Preoperative OPG of Case 4

Radiographic examination Figure 7 revealed a well defined, radiolucent lesion extending from lower left canine to ramus of the mandible. Aspiration showed a blood tinged pus like material. Histopathological evaluation of the incisional biopsy specimen showed characteristic infiltration of fibrous wall of the lesion by suggestive of mural unicystic ameloblastoma (Ackermann's Type III). Under general anaesthesia surgical resection of the tumor mass with a 2cm rim of uninvolved bone was carried out, sparing the condyle (Figure 8). Histopathological evaluation of the

resected tumor mass confirmed mural variant of unicystic ameloblastoma. The follow up of the patient for past 44 months has been uneventful.



Figure 8: Postoperative OPG of Case 4

**Case no. 5**

A 17year old female patient reported to our department with swelling over the right side of the face for one and a half year, extending from right corner of the mouth to the angle region. Extra oral examination showed moderate asymmetry of the right side of the mandible Figure 9. Intra orally the swelling was extending from 43 to 48 region. Radiographic examination showed a typical osteolytic unilocular radiolucency extending to 43 to 48 region. A straw coloured fluid was aspirated. Histopathological evaluation of the incisional biopsy specimen revealed presence of invasive islands of ameloblastomatous epithelium in the connective tissue of the cyst, suggestive of Ackermann's Type III. Wide reaction was carried out under general anesthesia. A transport distractor was placed to achieve reconstruction of the resected segment Figure 10. But unfortunately in this patient following consolidation phase of distraction osteogenesis there was collapse of the alveolar arch on the resected side compromising the aesthetics. Osteotomy was planned but the patient declined further treatment. There has been no evidence of recurrence in a follow up period of 45 months.



Figure 9: Preoperative Extraoral View of Case 5

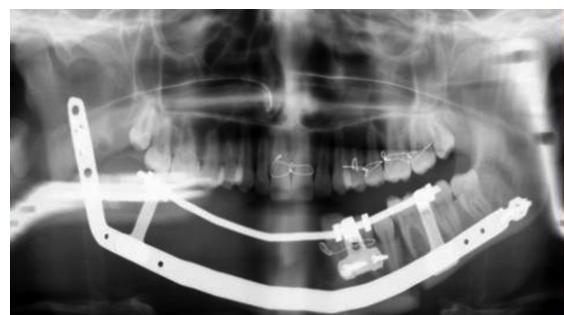


Figure 10: Postoperative OPG of Case 5

## DISCUSSION

WHO in 1992 defined ameloblastoma as benign but locally invasive polymorphic neoplasm consisting of proliferating odontogenic epithelium, which usually has a follicular or plexiform pattern lying in a fibrous stroma. Robinson defined ameloblastoma as Unicentric non functional intermittent in growth anatomically benign and clinically persistent. Robinson and Martinez<sup>[1]</sup> in 1977 described a distinct entity called unicystic ameloblastoma, known for less aggressive behavior as compared to solid ameloblastoma. It has been termed variously as cystic ameloblastoma<sup>4</sup>, intracystic ameloblastoma<sup>[5]</sup>, cystogenic ameloblastoma<sup>6</sup> and mural ameloblastoma<sup>[7]</sup>. WHO in 2017 classified ameloblastoma into 4 types: Conventional (solid/multicystic) type ameloblastoma, Unicystic ameloblastoma, Peripheral/extraosseous ameloblastoma, and Metastasising (malignant) ameloblastoma. In this new classification, a uniform terminology of unicystic ameloblastoma was accepted<sup>[8]</sup>. Macroscopically unicystic ameloblastoma is cystic in nature but on histological examination, shows a typical ameloblastomatous epithelium lining part of cyst cavity with or without luminal and or mural tumor growth and most important is its less aggressive biologic behavior and low recurrence rate than the classic solid or multicystic ameloblastoma. It accounts for 10 to 15% of all intraosseous ameloblastoma in various studies and more than 90 percent of these lesions are found in mandible usually posterior region which are often asymptomatic, although large lesions may cause a painful swelling of the jaw. Most of the ameloblastomas are chance finding as it initially causes mild facial deformity which may go unnoticed. The mean age range of the patients at the time of diagnosis is 25.5 years according to Li et al. 46% of the cases occur in the second decade with an age range between 6-79 years<sup>[9]</sup>. However in our case series 4 out of 5 cases were below 30 years with 3 patients falling in the third decade. This is suggestive that unicystic ameloblastoma occur considerably earlier than the solid multicystic ameloblastoma. Literature suggests 95.4% cases affected the mandible with a distinct predilection for the posterior mandible. This study is similar to our case series where in 4 out of 5 patients had posterior mandible involvement.

According to Li et al<sup>[9]</sup> 55% of the cases were males. However in our case series there was a female predilection with a ratio of 4:1. Classical clinical presentation of unicystic ameloblastoma is swelling of the affected jaw. Pain may be present at a later stage or when secondarily infected. In our case series 3 patients had initial complaint of only swelling however 2 patients complained of pain associated with swelling in which swelling was noticed prior to onset of pain.

Radiographically unicystic ameloblastoma presents typical unilocular radiographic appearance of circumscribed radiolucency with peripheral sclerotic

border that frequently surrounds the crown of an unerupted mandibular molar. Rarely the borders can be scalloped indicating the aggressive nature of the lesion. Radiographic appearance of all unicystic ameloblastoma has been divided into two main patterns, unilocular and multilocular, with a clear predominance of the unilocular configuration in all studies where features was evaluated.<sup>[6]</sup> In all our cases also we found a well defined typical unilocular expansile radiolucency having a scalloped margin causing root resorption of the involved tooth and displacement of the involved crown by the cystic tumor.

The histopathological features of unicystic ameloblastomas have been established by several authors, all of whom recognize various subtypes determined by the pattern and extent of ameloblastomatous proliferation in relation to the cyst wall.<sup>[7]</sup> Features include thin, nonkeratinized epithelium, basal palisading, spongiosis, epithelial invaginations and subepithelial hyalinization.

Histopathologically in 1988, Ackermann et al<sup>[3]</sup> reclassified UA into three types with prognostic and therapeutic implications.

In Type 1, the tumor is confined to luminal surface of cyst with a lining of ameloblastomatous epithelium.

Type 2 is characterized by epithelial nodules arising from the cystic lining and projecting into the cyst lumen. There is no evidence of infiltration of the fibrous cystic wall in either type of lesion.

In Type 3, the fibrous wall of cyst is infiltrated by a trabecular pattern that resembles the plexiform pattern seen in conventional ameloblastoma. In addition, it is characterized by a basal layer of columnar cells with hyperchromatic nuclei. These cells are loosely cohesive and resemble stellate reticulum epithelium.

Treatment of unicystic ameloblastoma depends on the histological picture of the lesion. In cases of Ackermann type I & II, conservative treatment modalities like enucleation and curettage etc are usually advocated, where as for Ackermann type III lesions, considering its aggressive nature literature suggests aggressive approach with resection followed by appropriate reconstruction. In the literature, recurrence after conservative treatment of conventional ameloblastoma ranges from 50% to 90%<sup>[10]</sup>. However for unicystic ameloblastoma it is reported to be between 10% & 20%; whereas conservative management of a lesion with mural invasion has recurrence rate upto 43%.

In our case series, 2 patients who had a histopathological picture similar to type III had undergone resection and reconstruction with reconstruction plate (case 4) and distraction osteogenesis (case 5). Following transport distraction we encountered complication in the form of arch collapse especially on the resected side thereby compromising the esthetics of the patient. In other patient following reconstruction

**Table 1: Summary of the cases**

Case	Important Observations	Histopathological Diagnosis	Treatment Done	Follow Up
1	3 <sup>rd</sup> Decade Of Life Gradually increasing swelling in the posterior mandible showing bicortical expansion, straw coloured aspirate and unilocular radiolucency	Unicystic Ameloblastoma	Curettage + Electrocauterization + Primary Closure	31 months without any signs and symptoms of recurrence
2	3 <sup>rd</sup> Decade Of Life Gradually increasing swelling in the anterior mandible showing bicortical expansion, straw coloured aspirate and unilocular radiolucency	Unicystic Ameloblastoma	Enucleation + Iodoform Gauge Packing	26 months without any signs and symptoms of recurrence
3	4 <sup>th</sup> Decade Of Life Gradually increasing swelling in the posterior mandible showing bicortical expansion, blood tinged pus as aspirate and unilocular radiolucency	Unicystic Ameloblastoma	Marsupialisation + Obturator	6 months with significant reduction in the size of the tumor
4	3 <sup>rd</sup> Decade Of Life Gradually increasing swelling in the posterior mandible showing blood tinged pus as aspirate and unilocular radiolucency	Unicystic Ameloblastoma	Resection + Reconstruction with Recon Plate	44 months without any signs and symptoms of recurrence
5	2 <sup>nd</sup> Decade Of Life Gradually increasing swelling in the posterior mandible showing straw coloured aspirate and unilocular radiolucency	Unicystic Ameloblastoma	Resection + Reconstruction with Recon Plate + Distraction Osteogenesis	45 months without any signs and symptoms of recurrence

with reconstruction plate denture was fabricated with satisfactory results. These two patients who had Ackermann type III unicystic ameloblastoma, did not show any signs of recurrence even after a mean follow up period of 44 months.

According to Norifumi Nakamura et al<sup>11</sup>, marsupialisation was effective in young patients and varied from 4 months to 9 years and 6 months. In our case series 1 patient had undergone marsupialisation as the lesion was considerably big and was involving the major anatomical structures. In this patient, radiologically there was scalloped margin with an Ackermann type II lesion. Considering the tumor extent, we had opted for marsupialisation.

In 1961 Seldin<sup>[12]</sup> reported the usefulness of marsupialisation as a treatment for unicystic ameloblastoma. This procedure aids in minimizing the tumor volume and limits the extent of surgery to an extent. It simplifies the second surgery or at times eradicates the tumor in toto. In our case following 6 months the lesion has reduced in size, but has not completely resolved. Marsupialisation has potential for new bone formation which is mainly influenced by the age of

the patient and hence it seems to be more useful in young patients.

Unicystic ameloblastoma is a tumor which is more often treated as a cyst especially Ackermann type I & II because of less chances of recurrence and hence conservative management especially enucleation has been widely advocated for its management. When enucleation can be achieved in toto, chances of recurrence are considerably lesser. However Marx et al<sup>13</sup> had demonstrated that ameloblastic tumor cells can extend 2.3 to 8 mm beyond the radiographic margins of the tumor and thus the enucleation followed by the application of Carnoy's solution or electrocautery would help in minimizing the recurrence of the tumor.

Carnoy's solution (chloroform 3 ml, absolute alcohol 6 ml, glacial acetic acid 1 ml, ferric chloride 1 gm) is a powerful fixative, first described by Culter and Zollinger<sup>[14]</sup>, can penetrate bone upto 1.54mm<sup>[15]</sup>

In our case series, in case number 1 we opted for enucleation followed by thermal coagulation of all the raw bony margins which gave us satisfactory results

with a follow up period of 31 months without any recurrence. In case number 2 following enucleation we applied carnoy's solution to the bony margins with cotton applicators for three minutes. Following surgery packing of the bony cavity was done with iodoform gauze to aid in healing with secondary intention<sup>[16]</sup>. This patient is been followed up for past 26 months without any sign of recurrence.

There is no universally accepted treatment modality for ameloblastoma. Our approach to management of these lesions is not significantly different to that advocated in literature. In our follow up series, cases where resection was done have been followed up for a maximum time period without any sign of recurrence Table 1: Summary of the cases. The treatment plan for unicystic ameloblastoma should be tailored to individual patients depending upon their age, signs and symptoms and histopathological picture.

## CONCLUSIONS

The diagnosis of unicystic ameloblastoma is based on clinical, histopathologic and CT features. Conservative treatment for unicystic ameloblastoma seems to be justified in preference to mutilating radical surgery, especially in cases of Ackermann type 1 and type 2. In cases of recurrence, resection with normal bone margin is advocated. However, for Ackerman type 3 unicystic ameloblastoma or any type of unicystic ameloblastoma in the posterior maxilla (where there is ominous potential for recurrence and infiltration into adjacent structures) resection in form of partial maxillectomy, marginal resection or segmental resection of mandible is necessary. Unicystic ameloblastoma is a tumor with a high rate of recurrence especially when the ameloblastic focus penetrates the adjacent tissues from the wall of the cyst. The ability to predict this potential occurrence prior to performing a particular therapeutic strategy would definitely reduce the recurrence incidence. Irrespective of the treatment modality, long term follow-up is mandatory, as recurrence of unicystic ameloblastoma may be long delayed.

## REFERENCES

1. Robinson L Martinez MG: Unicystic ameloblastoma: A prognostically different identity. *Cancer* 1977; 40: 2278-2285.
2. Gardner DG, Corio RL: Plexiform unicystic ameloblastoma: A variant of ameloblastoma with a low recurrence rate after enucleation. *Cancer* 1984; 53: 1730.
3. Ackermann GL, Altini M and Shear M. The unicystic ameloblastoma: a clinicopathological study of 57 cases. *J Oral Pathol* 1988;17:541-6.
4. Leider AS, Eversole LR and Barkin ME. Cystic ameloblastoma: A clinicopathological analysis. *Oral Surg, Oral Med, Oral Pathol* 1985; 60: 624-30.

5. Rapidis AD, Angelopoulos AP, Skouteris CA, et al. Mural (intracystic) ameloblastoma. *Int J Oral Surg* 1982;11:166-4.
6. Eversole LR, Leider AS and Strub D. Radiographic characteristics of cystogenic ameloblastoma. *Oral Surg, Oral Med, Oral Pathol* 1984; 57: 572-7.
7. Shteyer A, Lustmann J and Lewis-Epstein J. The mural ameloblastoma: a review of the literature. *J Oral Surg* 1978;26:866-72.
8. Soluk-Tekkeşin M, Wright JM (2018). "The World Health Organization Classification of Odontogenic Lesions: A Summary of the Changes of the 2017 (4th) Edition". *Turk Patoloji Dergisi*. 34 (1). [doi:10.5146/tjpath.2017.01410](https://doi.org/10.5146/tjpath.2017.01410). [PMID 28984343](https://pubmed.ncbi.nlm.nih.gov/28984343/).
9. Li T-J and Kitano M. Reviewing the unicystic ameloblastoma: a clinicopathologically distinct entity. *Oral Med Pathol* 1997;2:61-68.
10. Gardner DG. A pathologist's approach to the treatment of ameloblastoma. *J Oral Maxillofac Surg* 1984; 42:161-611.
11. Norifumi Nakamura & Yoshinori Higuchi, et al. Marsupialization of cystic ameloblastoma: A clinical and histopathologic study of the growth characteristics before and after marsupialization. *J Oral Maxillofac Surg* 1995; 53:748-754.
12. Seldin SD: Ameloblastoma in young patients, report of two cases. *J Oral Surg* 19:508, 1961.
13. Marx RE, Smith BH, Smith BR, Fridrich KL. Swelling of the retromolar region and cheek associated with limited opening. *J Oral Maxillofac Surg* 1993; 51: 304-309.
14. Culter E, Zollinger R. Sclerosing solution in the treatment of cysts and fistulae. *Am J Surg* 1933; 19: 411.
15. Voorsmit R. The incredible keratocyst: A new approach to treatment. *Dtsch Zahnarztl Z* 1985; 40: 641.
16. P. K. Lee, N Samman, I.O. Ng: Unicystic ameloblastoma- use of Carnoy's solution after enucleation. *Int J. Oral Maxillofac Surg* 2004; 33: 263-267.