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# Unicystic Ameloblastoma of Mandible- Imaging Features: A Case Report and Literature Review


## Abstract

Ameloblastoma are benign tumors whose importance lies in its potential to grow into enormous size with resulting bone deformity. They are typically classified as unicystic, multicystic, peripheral, and malignant subtypes. Unicystic ameloblastoma (UA) refers to those cystic lesions that show clinical, radiographic, or gross features of an odontogenic cyst but on histological examination show a typical ameloblastomatous epithelium lining, with or without luminal and/or mural tumor growth. We present a very rare case of unicystic ameloblastoma in a girl child with an age of 10 years; clinical and radiographic features of UCA, its differential diagnosis, histopathology, and current concepts of management have also been discussed in the present paper.


**Keywords:** *Ameloblastoma, computed tomography, imaging of ameloblastoma, unicystic ameloblastoma*


**Archana Pokala[1], Avinash Tejasvi M.L.[1], Geetha Paramkusam[2], Revath Vyas[3], Harsha Bhayya[4], Pavani Donempudi[5]**

*[1]Department of Oral Medicine and Radiology, Kamineni Institute Dental Sciences, Narketpally, Hyderabad, Telangana, India, [2]Department of Oral Medicine and Radiology, Sai Venkateshwara Dental Clinic, Hyderabad, Telangana, India, [3]Department of Oral Medicine and Radiology, Primary Health Care Centre, Nampally, Hyderabad, Telangana, India, [4]Department of Oral Medicine and Radiology, HKDET Dental College, Hospital and Research Institute, Humnabad, Karnataka, India, [5]Department of Oral Medicine and Radiology, Y.C.M.M and R.D.F'S Dental College, Ahmednagar, Maharashtra, India*


## Introduction

Ameloblastoma is the most common benign odontogenic tumor accounting for approximately 1% of tumors and cysts of the jaw and 10% of all the odontogenic tumors.[1] It is a slow-growing, persistent, and locally aggressive neoplasm that may originate from the epithelium involved with the formation of teeth such as enamel organ, odontogenic rests of Malassez, reduced enamel epithelium, and odontogenic cyst lining.[2]

Ameloblastoma may occur centrally within the bone or peripherally, without an intraosseous component in the soft tissues overlying the alveolar ridge. Intraosseous lesions are of two types solid/conventional/multicystic and unicystic.[3] Unicystic ameloblastoma (UA), a variant of ameloblastoma first described by Robinson and Martinez[4] in 1977, refers to those cystic lesions that show clinical and radiologic characteristics of an odontogenic cyst but in histologic examination show a typical ameloblastomatous epithelium lining part of the cyst cavity with or without luminal and/or mural tumor proliferation. Before the report by Robinson and Martinez, this variant had been referred to as a mural or intraluminal ameloblastoma. Recognition of this growth pattern is very important because of its unicystic radiographic appearance, histologic findings, association with an unerupted tooth, occurrence in the mandible of younger patients, and a recurrence rate after conservative surgical treatment lower than that of its conventional counterpart.[5]

We present a case of a unicystic ameloblastoma in a 10-year-old child patient who reported with a complaint of swelling of her right lower jaw.

## Case Report

A 10-year-old female child came with a complaint of swelling on the right lower side of her jaw for 1 month which was insidious in onset and got gradually progressed to the present size. It was associated with severe, intermittent, and dragging type of pain which radiates to the right ear. Pain relieved with medication. It was also associated with extraoral swelling for 15 days. On extraoral examination, facial asymmetry was seen in the right lower third of the face. On intraoral examination, a solitary diffuse swelling was seen in the buccal vestibular region of 85 and 46, extending anteroposteriorly from middle third of 85 to distal surface of 46. Superiorly the swelling of the lesion is extending from attached gingiva of 85 and 46 to inferiorly into the buccal vestibule.

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***Address for correspondence:***
*Dr. Archana Pokala, Department of Oral Medicine and Radiology, Kamineni Institute Dental Sciences, Narketpally, Telangana, India. E-mail: pokala.archana9@gmail.com*

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Sinus opening with pus discharge was seen on the attached gingiva of 85 (buccal or lingual). On palpation, swelling was tender, hard in consistency, noncompressible, and nonreducible. Expansion of buccal cortical plate was felt in relation to 85 and 46.

Based on the history and clinical examination, a provisional diagnosis of ameloblastoma was made. Under differential diagnosis, ameloblastic fibroma and odontogenic keratocyst were considered. Intaoral periapical view (IOPA), occlusal view**,** orthopantomogram (OPG), and computed tomography (CT) scan were taken. IOPA taken in relation to 85 and 46 [Figure 1] showed diffuse radiolucency distal to 46 with the absence of 47 tooth bud. OPG [Figure 2] view showed a solitary, well-defined radiolucency of size 3 cm × 2 cm extending anteroposteriorly from mesial root of the tooth number 46 is 1 cm from the posterior border of the ramus of the mandible, superoinferiorly 1 cm below the sigmoid notch, to the inferior border of the mandible thinning the inferior cortical margin. Internal structure was radiolucent with the displaced tooth bud of 47 in the ramus region.

Panoramic CT section [Figure 3] and sagittal CT section [Figure 4] showed hypodense area of size 2.5 cm × 3 cm surrounding the developing tooth bud of 47. Axial CT section [Figure 5a] showed tooth within the hypodense area, and axial CT section [Figure 5b] showed buccal cortical plate expansion with breakdown of lingual cortical plate.

An incisional biopsy was done and it showed epithelial lining with ameloblast-like cells and adjacent connective tissue stroma. There was no luminal proliferation of epithelium, suggestive of intraluminal ameloblastoma [Figure 6]. Following the diagnosis, the parents were informed about the condition and proposed treatment. Surgical enucleation along with chemical cauterization with Carnoy's solution [Figure 7] was done under general anesthesia along with extraction of 47 [Figure 8] considering age of the patient. The patient is under follow-up, with no functional or esthetic complaints. Six months posttreatment, OPG shows signs of new bone formation [Figure 9].

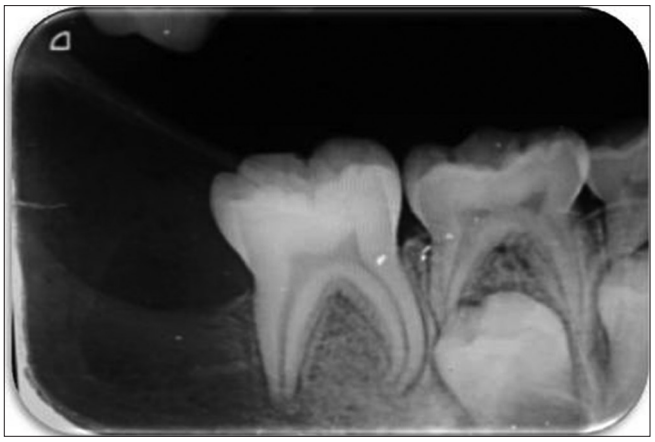

**Figure 1: Intraoral periapical radiograph showing diffuse radiolucency distal to 46 with the absence of 47 tooth bud**

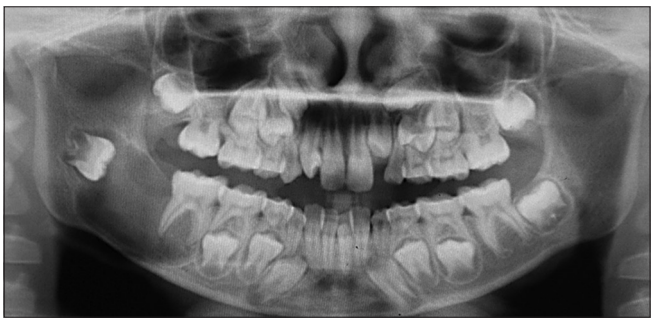

**Figure 2: Preoperative orthopantomogram showing solitary, well-defined radiolucency of size 3 cm × 2 cm surrounding the crown of 47 tooth bud which got displaced into the ramus region with thinning of the inferior border of the mandible**

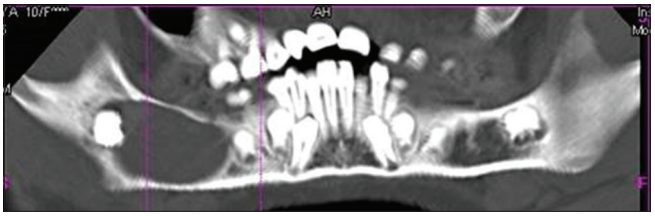

**Figure 3: Panoramic computed tomography section showing complete extension of the lesion**

## Discussion

UA accounts for 6%–15% of all intraosseous ameloblastomas.[1] It is less aggressive and usually occurs in an earlier age group than the solid or multicystic with about 50% of the cases occurring in the second decade of life. As in the present case, >90% of UA are seen affecting the mandibular region,[6] which was also seen in present case. In most cases, UA are associated with impacted tooth, mandibular third molar being the most common.[7]

The term unicystic is derived from the macroscopic and microscopic appearance, the lesion being essentially a well-defined, often large monocystic cavity with a lining, focally but rarely entirely composed of odontogenic (ameloblastomatous) epithelium.[8]

The pathogenesis of cystic ameloblastomas remains obscure. Some investigators believe that UA arises from preexisting odontogenic cysts, in particular a dentigerous cyst, while others maintain that it arises *de novo*. The reason why some ameloblastomas become completely cystic may be related to epithelial dysadhesion (e.g., defective desmosomes) or, more likely, to the intrinsic production of proteinases enzymes that normally degrade the central zone of the enamel organ after tooth development.[9] (e.g., metalloproteinases and serine proteinases).[9]

Radiographically, the unilocular: multilocular ratio is 13:3 when the lesion is associated with an impacted tooth. For the "nondentigerous" variant, this ratio changes to 8:7. Further, the "dentigerous" type occurs on average 8 years earlier than the "nondentigerous" variant. Finally, the mean

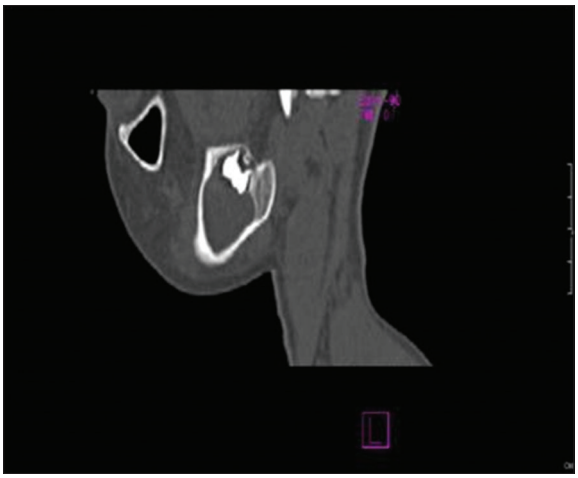

**Figure 4: Sagittal computed tomography section showing hypodense area surrounding the developing tooth bud of 47**

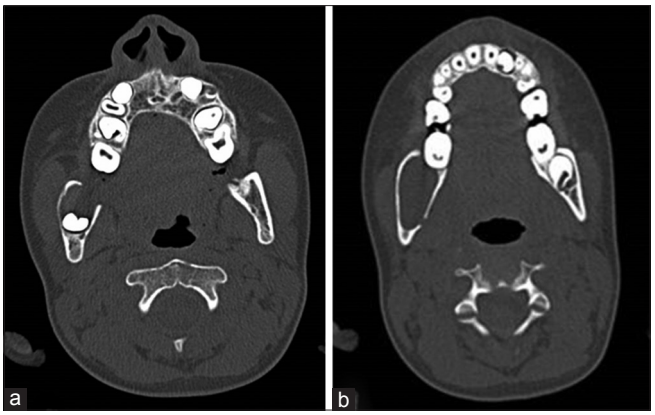


**Figure 5: (a) Axial computed tomography section showing tooth within the hypodense area (b) Axial computed tomography section showing buccal cortical plate expansion with breakdown of lingual cortical plate**

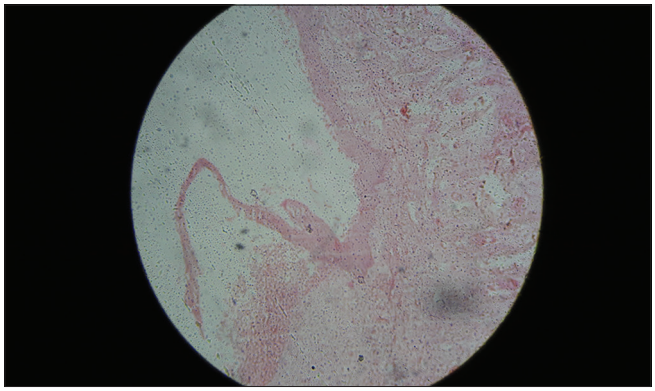

**Figure 6: Low-power histopathological picture of unicystic ameloblastoma showing intraluminal proliferation**

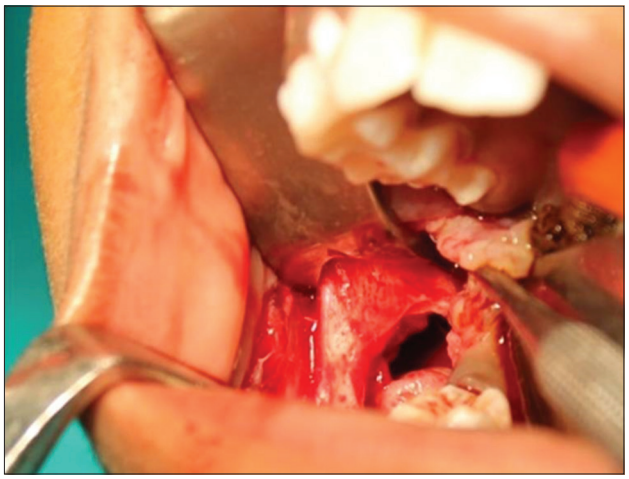

**Figure 7: Surgical site of enucleation**

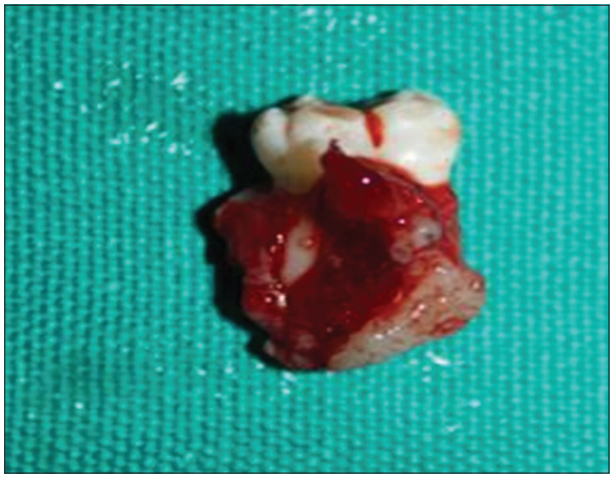

**Figure 8: Surgical specimen with extracted 47**

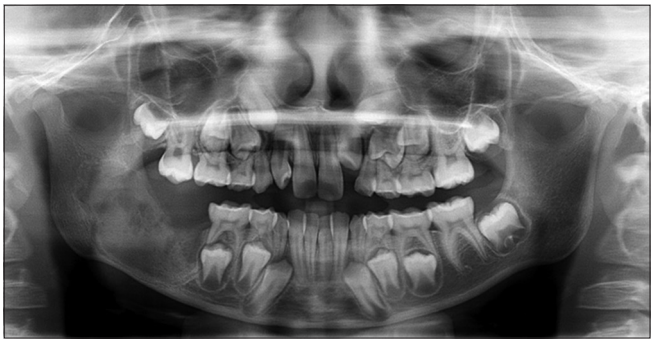

**Figure 9: Postoperative orthopantomogram showing signs of new bone formation**

age for unilocular, impaction-associated UAs is 22 years, whereas the mean age for the multilocular lesion unrelated to an impacted tooth is 33 years.[7]

Ackermann *et al*.[10] classified unicystic ameloblastoma into three types with prognostic and therapeutic implications such as:

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

**Table 1: English literature review till date**

| Year | Author | Age | Sex | Location | Clinical features | Histological features | Radiological features | Treatment |
|---|---|---|---|---|---|---|---|---|
| 1998 | Li *et al*.[16] | 10 | Female | Mandible | Mild fullness over the cheek | Unicystic ameloblastoma | UL | Enucleation |
| 2000 | Li *et al*.[17] | 5 | Male | Maxilla (premolar to second molar) | Cystic lesion | Mural type | INA | Enucleation |
| 2003 | Al-Khateeb and Ababneh[18] | 9 | Female | Mandible | Painless swelling | Unicystic ameloblastoma | UL | Enucleation plus peripheral ostectomy |
| 2007 | Huang *et al*.[19] | 9 | Male | Body-angle of the mandible | INA | INA | UL | Enucleation and peripheral ostectomy |
| 2008 | Qureshi *et al*.[20] | 10 | Female | Mandible | Mild fullness over the cheek | Unicystic ameloblastoma | UL | Enucleation, curettage |
| 2008 | Gulten *et al*.[21] | 8 | Male | Right mandible | Painless hard swelling | Unicystic ameloblastoma | UL | Enucleation and extraction of related teeth |
| 2011 | Chacko and Kuriakose[22] | 9 | Male | Mandible | Pain and swelling in relation to the right side of the lower jaw | Plexiform unicystic ameloblastoma | UL | Enucleation, curettage |
| 2011 | Kalaskar *et al*.[23] | 9 | Male | Right maxilla | Painless swelling | Unicystic ameloblastoma with intraluminal proliferations | UL | Enucleation + Carnoy's solution and extraction of related teeth |
| 2011 | Ponniah[24] | 8 | Female | Left ramus of the mandible | Painless swelling on the left side of the mandible | Unicystic ameloblastoma | UL | Enucleation then segmental resection |
| 2011 | Sudhakara Reddy *et al*.[25] | 6 | Female | Anterior mandible | Slow growing painless swelling | Unicystic ameloblastoma | UL | Enucleation and extraction of related teeth followed by application of Carnoy's solution |
| 2012 | Scariot *et al*.[26] | 9 | Female | Right mandibular body | Painless swelling | Plexiform unicystic ameloblastoma | UL | Curettage with extraction of two adjacent teeth |
| 2013 | Bhutia *et al*.[27] | 5 | Male | Right mandible | painless hard swelling | Type 1 unicystic ameloblastoma | UL | Enucleation of the cyst with extraction of the involved teeth followed by application of Carnoy's solution |
| 2013 | Arora *et al*.[28] | 3 | Female | Left maxilla | Bony hard swelling | Unicystic ameloblastoma (Type 1.2) | UL | Enucleation of the cyst with extraction of the involved teeth |
| 2014 | Present case | 10 | Female | Right mandible | Swelling with mild pain | Unicystic ameloblastoma | UL | Enucleation with chemical cauterization |

INA – Information not available; UL – Unilocular ameloblastoma

The microscopic pattern that exhibits mural invasion in UA suggests a more aggressive potential.[11]

Another histologic subgrouping by Philipsen and Reichart[12] has also been described as follows:

- Subgroup 1: Luminal
- Subgroup 1.2: Luminal and intraluminal
- Subgroup 1.2.3: Luminal, intraluminal, and intramural
- Subgroup 1.3: Luminal and intramural.

The unicystic ameloblastomas diagnosed as subgroups 1 and 1.2 can be treated conservatively (enucleation), whereas subgroups 1.2.3 and 1.3 showing intramural growths require radical resection, as for a solid or multicystic ameloblastoma. Following enucleation, vigorous curettage of the bone should be avoided as it may implant foci of ameloblastoma deeper into bone. Chemical cauterization with Carnoy's solution[13] 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.[14]

Recurrence rates for unicystic ameloblastoma after conservative surgical treatment (curettage or enucleation) are generally reported to be <25%. For intraluminal and plexiform type of unicystic ameloblastoma, recurrence rate was found to be as low as 10.7%.[15] Recurrence rates for solid multicystic ameloblastoma was found to be about 50%–90%.

The present analysis included only publications in English. All well-documented publications during the last

20 years were collected, and several clinicopathological features of each case were studied. The following data were recorded: age (≤10 years), sex, location, clinical features/symptoms, histological type, radiographic appearance, and treatment. Only reports of unicystic ameloblastoma in children <10 years confirmed by histological analysis with all the data required for tabulation were included and the articles not having enough information were excluded [Table 1].

## Conclusion

Ameloblastomas in children differ from adults with a higher percentage of unicystic tumors. Unicystic ameloblastoma is a tumor with a strong propensity for recurrence, especially when the ameloblastic focus penetrates the adjacent tissue from the wall of the cyst. Although enucleation has been claimed to give acceptable recurrence rates in unicystic ameloblastoma, there are no large series with long follow-up in children. The histologic pattern that exhibits mural invasion in unicystic ameloblastoma suggests that more aggressive surgery is necessary. The present case was treated with Carnoy's solution along with the enucleation, which suggests a possible benefit against recurrence.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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### Conflicts of interest

There are no conflicts of interest.

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