

# Maxillary unicystic ameloblastoma: A review of the literature

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## ABSTRACT

The term unicystic ameloblastoma (UA) refers to those cystic lesions that show clinical, radiographic, or gross features of a jaw cyst, but on histologic examination show a typical ameloblastomatous epithelium lining part of the cyst cavity, with or without luminal and/or mural tumor growth. Although the histology suggests that cystic ameloblastomas follow a biologically low-grade course, recent evidence suggests that they may often behave clinically as aggressive tumors. This is supported by the high incidence of cortical perforation, tooth resorption, increase in lesion size, bony destruction, and a high rate of recurrence after simple enucleation. Here, the authors present a case report on unicystic variant of ameloblastoma in the maxilla. An attempt has been made to emphasize that it can involve the maxillary jaw, which is rarely affected and could be more aggressive than previously thought. A literature review on the topic has been added along with the case report. It is important to remember that a proper and timely diagnosis of the character and extent of a UA (with a thorough histopathologic examination of the entire specimen) can help in the overall long-term well-being of the patient.

**Key words:** Ameloblastoma, maxilla, unicystic

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## INTRODUCTION

Ameloblastoma is one of the most common types of odontogenic tumor, however, it accounts for only 1% of all oral tumors.<sup>[1]</sup> It is a benign tumor whose importance lies in its potential to grow to enormous size resulting in bone deformity. The origin of the tumor is thought to be from sources that include residual epithelium of the tooth-forming apparatus, such as the epithelial cell rests of Malassez; epithelium of odontogenic cysts; basal cells of the surface epithelium; epithelium of the enamel

organ; and heterotopic epithelium from extraoral sites, such as the pituitary gland.

The unicystic type comprises 10%–15% of all ameloblastomas.<sup>[2]</sup> The unicystic ameloblastoma (UA) was first described as a distinct entity by Robinson and Martinez, although there were several references to these lesions in the earlier literature. It has been termed variously as mural ameloblastoma, intracystic ameloblastoma, cytsogenic ameloblastoma, cystic ameloblastoma, and plexiform UA.<sup>[1]</sup>

The term unicystic ameloblastoma refers to those cystic lesions that show clinical, radiographic, or gross features of a jaw cyst, but on histologic examination show a typical ameloblastomatous epithelial lining part of the cyst cavity, with or without luminal and/or mural tumor growth. This variant is believed to be less aggressive, tends to affect patients at a younger age, and its response to enucleation or curettage is

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more favorable than the classic solid or multicystic ameloblastomas.<sup>[3]</sup>

The location within the jaw bones favors greatly the mandible. It is noteworthy that the UA series published by Philipsen and Reichart showed that mandible to maxilla ratio varied from 3 to 13:1 and the posterior mandible is the single most often affected region.<sup>[4]</sup>

The report is a rare case of occurrence of UA in the maxilla of a 25-year-old male patient.

## REVIEW OF THE LITERATURE

UA, a variant of ameloblastoma, was first described by Robinson and Martinez and accounts for 10%–15% of all ameloblastomas. The term “unicystic ameloblastoma” is derived from the macro- and microscopic appearance, the lesion being a well-defined, often large monocystic cavity with a lining, focally but rarely entirely composed of odontogenic (ameloblastomatous epithelium). It is often accompanied by an innocuous epithelium of varying histologic appearance that may mimic the lining of a dentigerous or radicular cyst.<sup>[4]</sup>

Some investigators believe that the UA arises from preexisting odontogenic cysts, in particular dentigerous cyst, while others maintain that it arises *de novo*. Leider *et al* proposed 3 pathogenic mechanisms for the evolution of UA: (1) the reduced enamel epithelium associated with a developing tooth undergoes ameloblastic transformation with subsequent cystic development; (2) ameloblastoma arise in dentigerous or other types of odontogenic cysts in which the neoplastic ameloblastic epithelium is preceded temporarily by a nonneoplastic stratified squamous epithelial lining; and (3) a solid ameloblastoma undergoes cystic degeneration of ameloblastic islands with subsequent fusion of multiple microcysts and develops into a unicystic lesion.<sup>[5]</sup>

The UA has an almost equal male to female distribution.<sup>[6]</sup> The unicystic variant occurs most commonly in the mandible as an intrabony lesion during the second or third decade of life.<sup>[4,7,8]</sup> Maxillary UA are very rare. The first case of maxillary UA was reported by Gardner and colleagues in 1987, which occurred in a 12-year-old boy in the molar area.<sup>[3,9]</sup> UA in the anterior maxilla is considered to be rare and atypical.<sup>[10]</sup>

The UA occurs in a younger age group, with slightly more than 50% of cases occurring in patients in the second decade of life. Between 50% and 80% of cases are associated with tooth impaction, the

mandibular third molar being most often involved. The “dentigerous” type occurs on average 8 years earlier than the “nondentigerous” variant. The mean age for unilocular, impaction-associated UA is 22 years, whereas the mean age for the multilocular lesion unrelated to an impacted tooth is 33 years. There are no reports of any sexual or racial predilection.<sup>[4]</sup>

Patients most commonly present with a chief complaint of swelling and facial asymmetry. Although the swelling is typically asymptomatic, pain is an occasional presenting sign. Continued growth of the tumor and enlargement of the involved area may eventuate in the ulceration of the overlying mucosa. Small lesions tend to be discovered more often on routine radiographic examination or as a result of local effects produced by the tumor. Such local effects include tooth mobility, occlusal alterations, and failure of eruption of teeth.<sup>[11]</sup>

Radiographic appearance of UA presents with unilocular and multilocular patterns with clear predominance of unilocular configuration. It is often difficult to distinguish UA upon panoramic radiograph or CT scan images. In a clinicopathologic study, Li *et al* showed that 75% of UA are in fact temporarily diagnosed as odontogenic cysts, such as a dentigerous cyst or a keratocyst.<sup>[12]</sup> Contrast-enhanced magnetic resonance imaging was considered useful in the diagnosis of UA.<sup>[13]</sup>

Histologically, the minimum criterion for diagnosing a lesion as UA is the demonstration of a single cystic space lined by odontogenic (ameloblastomatous) epithelium often seen only in focal areas.<sup>[14]</sup>

Histologic subgrouping (modified after Ackermann *et al*) by Philipsen and Reichart:

Subtype 1	Luminal UA
Subtype 1.2	Luminal and Intraluminal UA
Subtype 1.2.3	Luminal, Intraluminal, and Intramural UA
Subtype 1.3	Luminal and Intramural UA

True nature of the lesion becomes evident only when the entire specimen after enucleation is available for histopathologic examination. The specimen should be subjected to multiple and serial sectioning for examination of cells and tissue configuration of an ameloblastomatous nature in intramural nodules.

Plexiform UA, the histologic equivalent of intraluminal UA, refers to a pattern of epithelial proliferation that has been described in dentigerous cysts. It does not exhibit the histologic criteria for ameloblastoma published by Vickers and Gorlin. Plexiform UA is not always associated with unerupted teeth. It exhibits a low rate of recurrence following enucleation and curettage.<sup>[14]</sup>

No particular treatment is advocated for all the variants of UA. Case should be judged on its own merits. Treatment planning depends on the final histopathologic findings. UA diagnosed as subtypes 1 and 1.2 can be treated conservatively [careful surgical enucleation], whereas UAs belonging to 1.2.3 and 1.3 subtypes should be treated aggressively. Aggressive approach can also be utilized if infiltration from the epithelial cyst lining into the cyst wall has been demonstrated.<sup>[14]</sup>

Lau *et al* in their study on 132 patients of UA observed that the recurrence rate was 3.6% for resection, 30.5% for enucleation alone, 16% for enucleation followed by application of Carnoy's solution, and 18% for marsupialization.<sup>[15]</sup>

The surgical treatment of the present case consisted of careful enucleation of the whole lesion, followed by chemical cauterization and extraction of all the involved teeth.



Figure 1: Extraoral swelling on the left side of the face

## CASE REPORT

A 25-year-old male patient, a laborer by profession, reported to the outpatient department of Sardar Patel Post Graduate Institute of Dental and Medical Sciences, Lucknow, with the chief complaint of painless swelling on the left side of the face since one and a half years [Figure 1].

The patient was apparently asymptomatic one and a half years back. Then he noticed a swelling on the left side of the face, swelling was painless in nature from its inception, and initially of a small dimension, which gradually increased in size up to the present size. The patient had consulted at another medical college for the same complaint 6 months earlier and was operated under local anesthesia. An intraoral incisional biopsy was performed after the extraction of first premolar from the left maxillary quadrant. Histopathologic report of the specimen was indicative of ameloblastoma. Past history and medical history were not relevant. He was taking no medication and had no history of known drug allergy. His physical examination revealed no abnormality other than those related to the chief complaint.

On extraoral examination, detectable facial asymmetry was present on the left side. Well-circumscribed, nontender, nonfluctuant, smooth surfaced swelling of hard consistency, spherical in shape and approximately 5 × 4 cm in size was present in the left maxillary region extending from midline to 5 cm anterior to the tragus. Superiorly it extended up to the infraorbital fold. Obliteration of nasolabial fold was present along with slight elevation of alar base on the left side. On superficial examination of the nostrils, the nasal floor was found to be elevated in the left nostril.

On intraoral examination, bicortical expansion was present in the left maxillary quadrant vestibular



Figure 2: Intraoral swelling showing bicortical expansion and vestibular obliteration

area extending from midline to third molar, causing complete obliteration of vestibular on the buccal side and also extending into the palatal region from the left maxillary central incisor region to the greater palatine foramen region and up to the midpalatal region. Egg shell crackling was present buccally as well as palatally [Figure 2].

Vitality test revealed 11, 21, 22, 23 to be vital and 25, 26, 27 to be nonvital. Needle aspiration performed through area of fluctuance revealed yellow straw colored fluid with plenty of cholesterol crystals.

Detailed radiographic examination (including 3-D CT scan) revealed the presence of multilocular lesion extending from left maxillary central incisor to the third molar region. Multilocularity is of honey comb appearance in the anterior part of the maxilla up to premolar, whereas posterior molar part showed soap bubble appearance [Figure 3]. Complete obliteration and haziness of left maxillary sinus along with involvement of lateral wall of piriform aperture



Figure 3: Multilocular radiolucency

was observed [Figure 4]. Interestingly, anterior teeth up to the canine showed mild displacement, which is suggestive of less aggressive nature, whereas premolar and molar showed extensive root resorption, which indicates more aggressive nature of the lesion. Provisionally, a diagnosis of odontogenic cyst was made.

The patient was operated under general anesthesia and with the help of intraoral approach and osteotomies, the lesion was thoroughly removed leaving sufficient tissue margin. Extraction of all involved teeth was done. Surgical bed was treated with freshly prepared Carnoy's solution to achieve chemical cauterization. Excised specimen was sent for histopathologic examination [Figure 5].

Histologic analysis of the surgical specimen revealed UA [Figure 6]. The ameloblastoma was completely surrounded by a dense fibrous capsule and lined with ameloblastic epithelium, with tall columnar basal layer, subnuclear vacuoles, reverse nuclear polarity, and central layer of edematous, stellate cells. The immediate postoperative healing was uneventful.

Postoperative recall checkup was performed to observe recurrence for a period of 1 year in which noticeable improvement was seen clinically extraorally and intraorally [Figures 7 and 8]. The Paranasal Sinus Radiograph showed bone healing and reduction in size of radiolucency [Figure 9]. During the recall checkup and subsequent follow-up period, there was no evidence of recurrence and the patient showed a drastic improvement in his condition.

## CONCLUSION

Preoperative diagnosis of UA can be difficult or

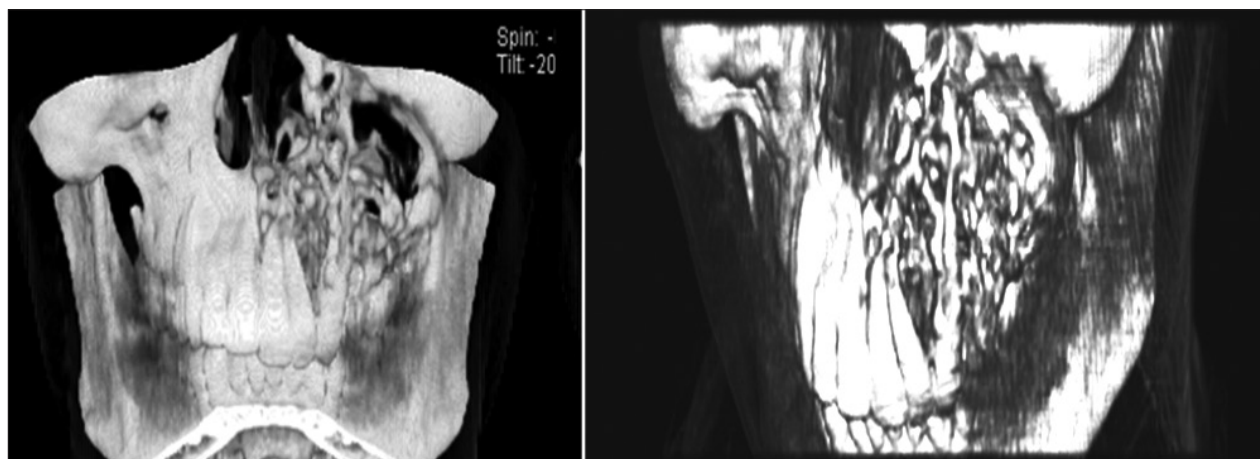


Figure 4: 3-D CT scan reveals obliteration of left maxillary sinus with involvement of the wall of piriform aperture

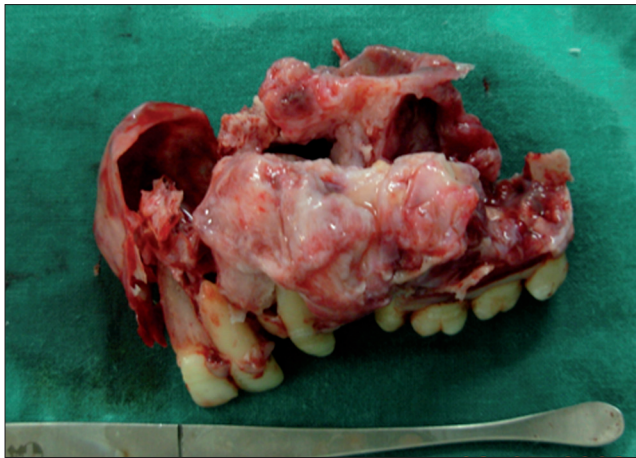


Figure 5: Excised specimen

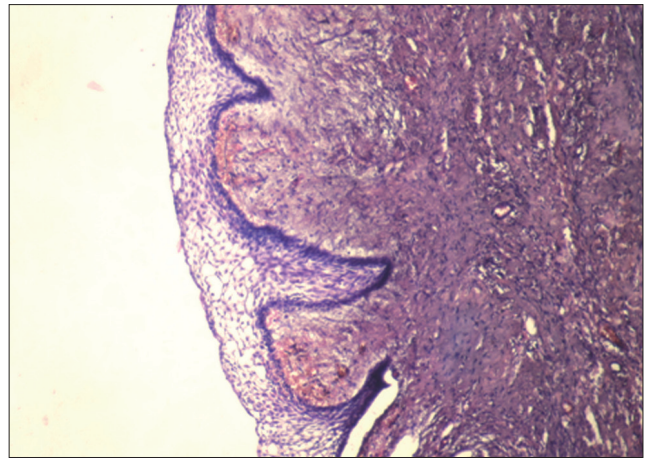


Figure 6: Histologic analysis reveals ameloblastic epithelium and dense connective tissue



Figure 7: Extraoral postoperative photograph

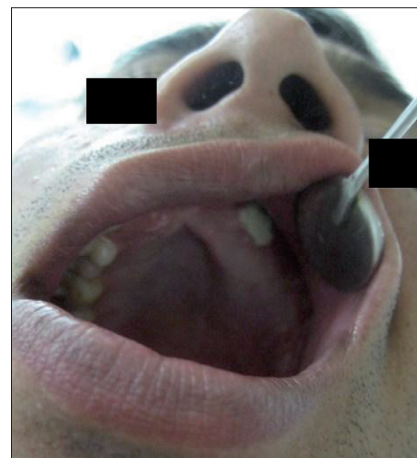


Figure 8: Intraoral postoperative photograph



Figure 9: Paranasal sinus radiograph showing bone healing

sometimes impossible because this variant of ameloblastoma shares significant clinical and radiographic similarities with odontogenic cysts and tumors and because incisional biopsy may not be able to reflect the true nature of the lesion. Long-term

follow-up is mandatory because recurrence may appear years after tumor removal.

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