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## CASE REPORT

# An Unusal Case Of Unicystic Ameloblastoma Involving The Anterior Of Maxilla

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### ABSTRACT:

Unicystic ameloblastoma is believed to be less aggressive lesion and responds well to a conservative surgery than a solid ameloblastoma or multicystic ameloblastoma. Unicystic ameloblastoma predominantly occur in the mandibular 3<sup>rd</sup> molar region

Here in our case we report Unicystic ameloblastoma in the anterior of maxilla in female patient who was treated by conservative surgery under the suspicion of radicular cyst .But the neoplastic behavior of the lesion was seen histopathologically

**Key words:** ameloblastoma, cyst, maxilla, Unicystic, radicular cyst

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### Introduction:

The ameloblastoma is the most common neoplasm arising from the primary odontogenic or tooth forming, tissue. In contrast to squamous cell carcinomas of the oral cavity, which are relatively common, these tumors are rare.

Ameloblastomas are benign tumors whose importance lies in their potential to grow to enormous sizes, with resulting bone deformity. These tumours characteristically expand within the jaw and displace the bone, the teeth and their roots. Occasionally, infiltrating tumours may erode through the bone and extend into the soft tissue. They originate from the epithelium which is involved in the formation of the tooth or the teeth: enamel organ, odontogenic rests of malassez, reduced enamel epithelium and odontogenic cyst lining or from the basal cells of the oral mucosa. Ameloblastomas are usually

asymptomatic and are found on routine dental radiographs. However, they may be present with the expansion of the jaws.

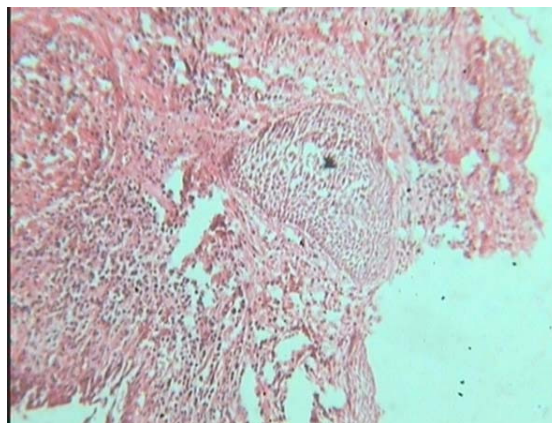
Radiographically, ameloblastomas may either have unilocular or multilocular radiolucencies with scalloped or sclerotic margins. The teeth adjacent to the tumor may show root displacement or resorption. Histologically, ameloblastomas are differentiated into unicystic, intraosseous, multicystic, solid intra osseous or peripheral. Here, the term Unicystic ameloblastoma refers to those cystic lesions that clinically, radiographically or grossly resemble a jaw cyst, while its histological examination shows a typical ameloblastomatous epithelium lining the cystic cavity, with or without luminal and/or mural tumor growth. The unicystic type of ameloblastoma is one of the least encountered variant of the ameloblastoma. This type appears more frequently in the 2<sup>nd</sup> or 3<sup>rd</sup> age group, with

no sexual or racial predilection. However, it is frequently encountered asymptotically in the posterior of the mandible. The treatment ranges from simple enucleation to *en bloc* resection or excision. Here, we report a rare case of Unicystic ameloblastoma in the anterior of the maxilla in a 28 year old female.

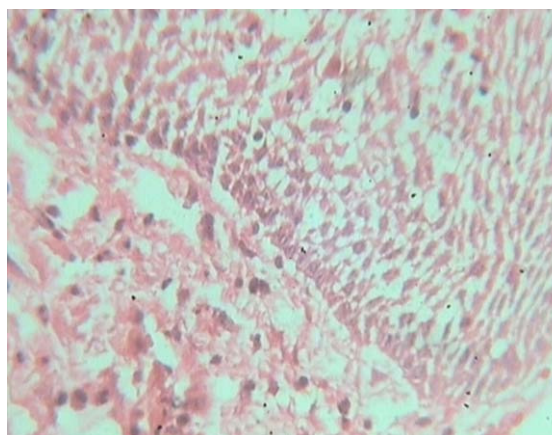
### Case report

A 28 year old female patient reported to the OPD of Gian Sagar Dental College, Banur, with a swelling on the front right side of the upper jaw since 3 months. The patient's history revealed that the swelling was slowly growing with a foul discharge. However, her middle third of the face did not show obvious distortion due to the swelling. The upper right lateral incisor, upon intra oral examination, was found to be nonvital. The submandibular lymph nodes on the right side were enlarged and non tender. Her past medical history was unremarkable. She was taking no medication and had no drug allergy and her physical examination revealed no abnormality other than those related to her chief complaint. The IOPA revealed a radiolucent lesion with sclerotic border in the right anterior of the maxilla, involving 13, 12, 11 and 21. A provisional diagnosis of infected periapical cyst was made. **The enucleation** of the lesion was performed completely and the lesion was sent for histopathological investigations.

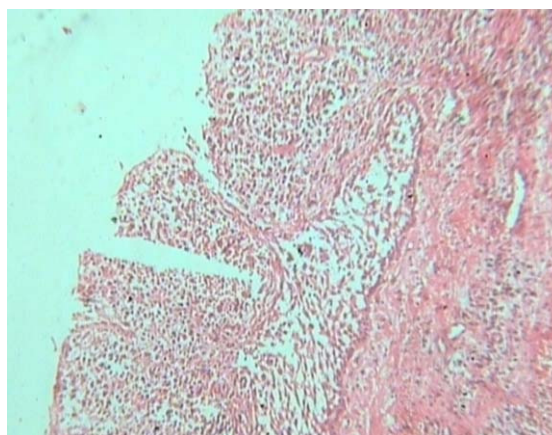
**Histopathological features:** The H and E stained tissue section revealed a fibrous cyst wall with islands consisting of ameloblastic epithelium comprising of a basal layer of columnar to cuboidal cells, with hyper chromatic nuclei and loosely cohesive overlying epithelial cells, resembling stellate reticulum [Table/Fig 1] and [Table/Fig 2]. The ameloblastic epithelium was seen to infiltrate the underlying connective tissue wall at many places [Table/Fig 3]. The connective tissue also showed islands of ameloblastic epithelium. Chronic inflammatory cells were also seen. The above mentioned features were suggestive of Unicystic ameloblastoma - mural type.



[Table/Fig 1]



[Table/Fig 2]



[Table/Fig 3]

### Discussion

Unicystic ameloblastoma, a variant of ameloblastoma, which was first described by Robinson and Martinez [1] in 1977, refers to the cystic lesion that shows the clinical and radiological characteristics of an odontogenic

cyst, but when examined histologically, this lesion shows an ameloblastic epithelium lining the cystic cavity with or without luminal or mural proliferation. This variant of ameloblastoma is less aggressive and tends to affect the younger population with a predilection for occurrence in the mandibular 3<sup>rd</sup> molar region. Depending on the extend of tumor cell proliferation within the cyst wall, several histological sub types of Unicystic ameloblastoma are recognized, which include the luminal type and the intra luminal type / the mural type[2][6].

Unicystic ameloblastomas account for 10-15% of all extra osseous ameloblastomas. The occurrence of this lesion may be de novo and whether it is a result of neoplastic transformation of a non – cystic epithelium or not, has been long debated. More than 90% of such lesions occur in the posterior of the mandible and are asymptomatic. A large lesion may cause painless swelling of the jaws [3],[4].

The radiographical features of UA are typically unilocular and there is a round area of radiolucency. Therefore, this lesion is often misdiagnosed as an odontogenic keratocyst or a dentigerous cyst. One of the efficient diagnostic tools which can be used to detect UA is Contrast enhanced (CE)-MRI. It is done to diagnose the cases of unilocular, round radiolucent lesions which can be visualized by panoramic radiography and/or CT. In the cases of UA, low signal intensity (SI) is observed on the T1-weighted images (WIs), a markedly high SI is observed on the T2WIs; and a relatively thick rim-enhancement with/without small intraluminal nodules is observed on the CE-T1WIs. CE-MRI is considered to be useful in the diagnosis of UA[5].

Apart from CE -MRI, another important diagnostic tool for detecting UA is immunohistochemistry. By this, one can differentiate UA from other types of Ameloblastomas. The expression of proliferating cell nuclear antigen (PCNA) is markedly observed in the tumors cells of other types of ameloblastomas, whereas there is no expression of PCNA in the cells of any variants

of UA. Moreover,  $\beta$ -catenin was characterized by a more positive marked expression in the UA than in other types of ameloblastoma and the cells that expressed this substance were not PCNA positive cells. This distinguishes UA from other ameloblastomas [5].

Microscopically, three variants of Unicystic ameloblastoma have been described [3],[7]:

**Luminal Unicystic ameloblastoma:** Is confined to the luminal surface of the cyst. The lesion consists of a fibrous cyst wall with a lining that consists totally or partially of ameloblastic epithelium, showing cuboidal or columnar epithelium, with hyperchromatic nuclei showing reverse polarity and basilar vacuolization. The overlying cells are loosely cohesive and resemble stellate reticulum.

**Intra luminal Unicystic ameloblastoma:** Here, the lesion projects into the cystic lumen from the cystic lining; the nodules may be small or large enough to fill the cystic lumen. The nodules which project into the cystic lumen demonstrate a pattern resembling the plexiform pattern of ameloblastoma and are thus referred to as plexiform Unicystic ameloblastoma.

**Mural Unicystic ameloblastoma:** Here, the cystic wall is infiltrated by follicular or plexiform ameloblastoma. The extent and depth of the ameloblastic infiltration may vary considerably. With any presumed unicystic ameloblastoma, multiple sections through many levels of specimens are necessary to rule out the possibility of the mural invasion of the tumor cells.

Another histological sub grouping by Philipsen and Reichart [8] has also been described:

**Subgroup 1:** Luminal Unicystic ameloblastoma (UA)

**Subgroup 1.2:** Luminal and intraluminal

**Subgroup 1.2.3:** Luminal, intraluminal and intramural

**Subgroup 1.3:** Luminal and intramural

The UAs which are diagnosed as subgroups 1 and 1.2 may be treated conservatively (careful enucleation), whereas the subgroups 1.2.3 and 1.3, showing intramural growths, must be treated radically, i.e., as a solid or multicystic ameloblastoma.<sup>11</sup> Vigorous curettage of the bone is discouraged, since it may implant the foci of ameloblastoma more deeply into the bone. Chemical cauterization with Carnoy's solution is also advocated for subgroups 1 and 1.2. Subgroups 1.2.3 and 1.3 in which the cystic wall is involved with islands of ameloblastoma tumor cells and in which there is possible penetration into the surrounding cancellous bone, are thought to be associated with a high risk for recurrence, thus requiring more aggressive surgical procedures. [9]

Late recurrence following treatment is commonly seen, the average interval for recurrence being 7 years. Recurrence is also related to the histological subtypes of UA, with those invading the fibrous wall having a rate of 35.7%, but others having a rate of only 6.7%. Recurrence rates are also related to the type of initial treatment. The recurrence rates are 3.6% for resection, 30.5% for enucleation alone, 16% for enucleation followed by Carnoy's solution application and 18% by marsupialization followed by enucleation (where the lesion reduced in size)[4],[7],[10],[14]

The age of the patient is another influencing factor which is related to the choice of treatment. As unicystic ameloblastoma tends to affect young adolescent patients, the concern to minimize surgical trauma and permit jaw function should be one of the important aspects in tumor management. In our patient, in order to obviate the problem of deformity, a simple enucleation was performed to remove the whole lesion.

While conservative surgery seems to have been justified with preference over mutilating radical surgery for this young patient, the choice of treatment has to be considered in conjunction with other clinical and pathological factors such as the size, location and growth pattern of the tumor. Whatever surgical approach the surgeon decides to take, long-term follow-up is

mandatory, as the recurrence of unicystic ameloblastoma may be long delayed [11],[12],[13].

However, the clinical and the radiological findings in most of the cases suggest that the lesion is an odontogenic cyst. These lesions are treated by enucleation and the diagnosis of Unicystic ameloblastoma is made only after the microscopic examination of the presumed cyst. If the ameloblastic elements are confined to the lumen of the cyst, with or without intraluminal tumor extension, then cyst enucleation is the preferred treatment. The patient should however be kept under long-term follow-up.

In our case, the Unicystic ameloblastoma was found to be in the anterior of the maxilla, which is contrary to the documented site. As this tumor itself was very rare, it was treated by enucleation.

## Conclusion

In conclusion, we can say that the diagnosis of Unicystic ameloblastoma in the present case report was made only after the histopathological evaluation of the specimen, as the lesion was present at an atypical location that is, at the anterior of the maxilla, involving a non vital tooth.

## Acknowledgement

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