Abstract
Ameloblastoma is considered as a true neoplasm of odontogenic epithelial origin which includes several clinico-radiological and histological types. Solid/Multicystic ameloblastoma is the most common subtype while its variant unicystic ameloblastoma (UA) is relatively uncommon. UA refers to cystic lesions showing clinical, radiographic or gross features of a cyst, but on histological examination shows a typical ameloblastomatous epithelium lining with or without luminal and/or mural tumor growth. In this article, we report a case of Unicystic Ameloblastoma in a 14 year old boy which was provisionally diagnosed as dentigerous cyst based on clinico-radiographic features and treated conservatively. Detailed microscopic examination revealed features of Unicystic ameloblastoma which requires long term follow-up to check for recurrence.

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Keywords: Dentigerous cyst, Mandible, Impacted third molar, Unicystic Ameloblastoma, Mural subtype.

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INTRODUCTION
Ameloblastoma is the most common tumor that arises from odontogenic epithelium.[1] Based on the tumor behavior, prognosis, clinical, radiographic and histopathological examination, Leon Barnes has categorized ameloblastoma into four types: Solid/multicystic ameloblastoma (SMA), Unicystic ameloblastoma (UA), peripheral ameloblastoma (PA) and desmoplastic ameloblastoma (DA).[2,3] Still confusion exists with regard to the terminology used for unicystic ameloblastoma. Before Robinson & Martinez (1977) introduced the concept of UA, many terminologies exists for this lesion: cystic (intracystic) ameloblastoma, ameloblastoma associated with dentigerous cyst, cystogenic ameloblastoma, extensive dentigerous cyst with intracystic ameloblastic papilloma, mural ameloblastoma, dentigerous cyst with ameloblastomatous proliferation, and ameloblastoma developing in a radicular cyst.[4,5] 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.[6] The dilemma in the diagnosis of unicystic ameloblastoma exists on a radiograph, especially when it is associated with impacted third molar showing similarity to dentigerous cyst and most of them were treated on that basis. Also, few cases have been reported where UA presents as periapical lesion resembling either a periapical cyst or granuloma.[7] The diagnosis of UA becomes evident only when the entire specimen is evaluated histopathologically.[8,9] Here, we report a case of unicystic ameloblastoma presenting in a 14-year-old boy, which mimics dentigerous cyst associated with an impacted third molar and treated conservatively.

CASE REPORT
A 14-year-old male patient reported to the outpatient department of Best dental science college, Madurai with a chief complaint of swelling in right cheek region noticed from past 6 months. The swelling had gradually increased in size over a period to attain the present size and was asymptomatic. His past medical and dental history was insignificant. Extraoral examination showed facial asymmetry with diffuse swelling measuring about 3x4 cm in the right mandibular posterior region (Figure 1). The overlying skin was apparently normal. On palpation, the swelling was hard, non-tender with no local rise in temperature. Right sub-mandibular lymph nodes were palpable, mobile, firm in
consistency and tender. Intraoral examination showed a solitary diffuse swelling measuring approximately 4 X 3 cm on the buccal aspect of 46, 47 extending into retro molar region with obliteration of buccal sulcus.

Panoramic radiograph revealed a well-defined unilocular radiolucency on right mandibular posterior region measuring approximately 6x4 cm, roughly elliptical in shape with hyperostotic border extending anteriorly 2 mm short of mental foramen, posteriorly 1 cm short of sigmoid notch, superiorly along the external oblique ridge and inferiorly 2 mm above the inferior border of mandible. The radiolucency was associated with developing third molar which was displaced inferiorly close to the lower border of mandible. There was absence of root resorption in adjacent teeth (Figure 3).

The mucosa over the swelling was relatively normal. The adjacent tooth 46 was grossly decayed without any symptoms of sensitivity /pain (Figure 2). On palpation, the swelling appears to be firm to hard in consistency, non-tender and not associated with any kind of discharge.

Aspiration yielded a straw-colored fluid (Figure 4). A preoperative diagnosis of dentigerous cyst was arrived based on the age
of the patient, location of the swelling, associated impacted third molar and the nature of aspirated fluid. However, a differential diagnosis of Odontogenic keratocyst, Unicystic Ameloblastoma, early stage of Ossifying fibroma was also taken into consideration.

Enucleation of the cystic lesion was done along with removal of impacted third molar. Macroscopically, the specimen was described as solitary, thick cystic lining attached to the cervical region of the impacted third molar. The entire specimen was subjected to histopathological examination. (Figure 5)

Histopathologic examination of initial tissue specimen showed a cystic lesion lined by non-specific epithelium and connective tissue wall resembling features of Dentigerous cyst. Successive processing of multiple tissue sections and serial sections revealed different features. Many areas showed odontogenic lining epithelium of varying thickness with basal hyperchromatic palisaded layer of cuboidal/columnar cells resembling ameloblasts and suprabasal cells showing resemblance to stellate reticulum.

The lining epithelium also shows plexiform type of proliferation into the adjacent connective tissue wall. (Figure 6 A,B&C) Based on histopathological report in correlation with clinical and radiological features, a final diagnosis of Unicystic ameloblastoma (mural type) was given. Follow-up after 1 week showed satisfactory healing, at the same time the patient was informed about the diagnosis and advised regular follow up periodically at regular intervals since the recurrence of UA is long delayed. Post-operative reviews after a period of 5 months shows no signs of recurrence and the lesion shows satisfactory bony healing (Figure 7).
Figure 6 A, B & C: H & E stained section showing odontogenic epithelial lining with focal areas of mural ameloblastomatous proliferation into the connective tissue wall (100 X;100X; 400X).

Figure 7: Panoramic radiograph showing bone remodeling 5 months after surgery
DISCUSSION
Unicystic ameloblastoma occurs less commonly, accounting for about 6% of ameloblastomas. It usually occurs in younger age group, around 16–20 years with 50% of the cases occurring in the second decade of life. It often shows male predilection and commonly involves the posterior mandible and ascending ramus. In our case, the lesion occurred in the body of mandible extending into the ramus in a 14-year-old boy which is in agreement with their clinicopathologic nature.

Unicystic ameloblastoma is usually asymptomatic, although a large tumor may cause painless swelling of the jaws with facial asymmetry. The clinical and radiographic findings in most cases of unicystic ameloblastoma suggest it to be an odontogenic cyst, particularly dentigerous cyst associated with impacted tooth and most commonly found in relation to mandibular third molar as seen in our present case. However, few cases are not associated with impacted teeth which are considered as non-dentigerous variant.

Unicystic ameloblastoma present as either unilocular or multilocular radiolucency radiographically, though there is a clear predominance of unilocular configuration in majority of studies. This predominance was exceptionally marked for the dentigerous variant where the unilocular: multilocular ratio was 4.3:1.2. For the non dentigerous type, this ratio was 1.1:1. In our case, well defined unilocular radiolucency was noted associated with an impacted tooth which is of dentigerous variant.

The pathogenesis of unicystic ameloblastoma remains obscure and many debates exists whether it develops de novo or from an existing cyst. Leider et al proposed three pathogenic mechanisms for the evolution of unicystic ameloblastoma: reduced enamel epithelium, from dentigerous cyst or cystic degeneration of solid ameloblastoma. The neoplasm originates within the jaws from epithelium that is involved in odontogenesis. Potential sources of epithelium include enamel organ, odontogenic rests (cell rest of Malassez, cell rest of Serres) reduced enamel epithelium and epithelial lining of odontogenic cyst especially dentigerous cyst. Since the present case is associated with an impacted tooth along with the presence of non-specific thin epithelial lining in focal areas, it seems to arise from preexisting dentigerous cyst. Possibility of misdiagnosing such cases as dentigerous cyst poses a problem where repeat and deeper biopsies are advisable to reveal the underlying
tumorous proliferation. The minimum criteria to diagnose a lesion histopathologically as unicystic ameloblastoma is the demonstration of a single cystic sac lined by odontogenic ameloblastomatous epithelium which is seen only in focal areas.

Ackermann et al classified this entity into three histologic groups: Luminal UA (tumor confined to the luminal surface of the cyst); Intraluminal/plexiform UA (nodular proliferation into the lumen without infiltration of tumor cells into the connective tissue wall); Mural UA (invasive islands of ameloblastomatous epithelium in the connective tissue wall not involving the entire epithelium). The present case belongs to the Mural type which was similar to other reported cases. Another histologic sub grouping by Philipsen and Reichart (2004) has also been described: Luminal UA; Luminal and intraluminal UA; Luminal, intraluminal and intramural UA; Luminal and intramural.

A definitive diagnosis of unicystic ameloblastoma can only be given after thorough histological examination of entire lesion and cannot be predicted preoperatively based on clinical or radiographic findings. As preoperative incisional biopsy is not representative of entire lesion, it may result in misdiagnosis. Furthermore, the epithelial lining of a UA is not always uniformly characteristic and often lined partly by a nonspecific thin epithelium that mimics dentigerous cyst lining. The true nature of the lesion becomes evident only after thorough histopathologic examination of the excised specimen. Multiple sections through many levels of specimen are necessary to rule out the possibility of mural invasion of tumor cells. This also applies to our case where the diagnosis of unicystic ameloblastoma was made after thorough processing and examination of the entire lesion histopathologically.

Treatment of UA remains controversial and greatly differs from that of conventional ameloblastoma. Robinson and Martinez initially recommended conservative treatment for UA because its behavior was thought to be different from solid or multicystic type. The luminal UA, luminal and intraluminal UA can be treated conservatively (careful enucleation), whereas intraluminal and intramural UA, luminal and intramural UA require radical resection similar to solid or multicystic ameloblastoma. Recurrence is related to the type of initial treatment and histologic subtypes of UA, with those invading the fibrous wall having a rate of 35.7%, while others show only 6.7% recurrence. This is because the cystic wall in
these cases has islands of ameloblastoma tumor cells and there may be penetration into the surrounding cancellous bone.\textsuperscript{[12]} Lau et al reported recurrence rates of 3.6\% for resection, 30.5\% for enucleation alone, 16\% for enucleation followed by Carnoy’s solution application, and 18\% for marsupialization followed by enucleation.\textsuperscript{[20]} Thus, the overall prognosis for unicystic ameloblastoma is considerably better than other variants. Literature suggested that even unicystic ameloblastoma are associated with 10\% recurrences and hence require a systematic follow-up.\textsuperscript{[21]} In our case, the lesion was treated conservatively considering the age of the patient and the clinical nature. However, the patient was advised for periodical check up every three months to check for recurrence.

**CONCLUSION**

The diagnosis of unicystic ameloblastoma based on clinical and radiographic features alone may be impossible in many cases due to its similarities with odontogenic cysts and tumors. Histopathologic examination is a sensitive tool for differentiating unicystic ameloblastoma from odontogenic cyst. Moreover, an incisional biopsy may not be able to reflect the true nature of the lesion. Careful postoperative histologic evaluation of the entire lesion is essential with multiple sectioning. Long term follow-up is mandatory since recurrence may occur years after removal. Regular postoperative radiographic examination can play an important role in minimizing recurrences.

**Conflict of Interest Statement**

There is no conflict of interest. Informed consent was taken from the patient.

**REFERENCES:**


