Unicystic Ameloblastoma: A Rare Case Report with Literature Review

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ABSTRACT

Ameloblastoma is traditionally considered a benign epithelial neoplasm with virtually no tendency to metastasize. It is the second most common odontogenic neoplasm. Its incidence, combined with its clinical behavior, makes ameloblastoma the most significant odontogenic neoplasm. Most ameloblastomas have been classically described as a multilocular cyst-like lesion of jaw. The unicystic variant is far less frequent and has been reported only 6% of ameloblastomas. Unicystic ameloblastoma refers to those cystic lesions that show clinical, radiographic, or gross features of a cyst, but on histologic examination show a typical ameloblastomatous epithelium lining part of the cystic cavity, with or without luminal and/or mural tumor growth. Here, we report a case of unicystic ameloblastoma in a 28-year-old male patient.

Keywords: Ameloblastoma, Cystic, Luminal, Mural


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INTRODUCTION

Many benign cysts and tumors involve mandible; these can be of odontogenic origin or of nonodontogenic origin. Lesions include ameloblastoma, radicular cyst, dentigerous cyst, keratocystic odontogenic tumor, central giant cell granuloma, fibroosseous lesions and osteomas. The most common tumor of odontogenic origin is ameloblastoma, which develops from epithelial cellular elements and dental tissues in their various phases of development. It is the classical example of a true neoplasm of enamel organ type tissue that lacks the potential to undergo differentiation, and hence has aptly defined as ‘unicentric, nonfunctional, intermittent in growth, anatomically benign and clinically persistent’ by Robinson. The earlier term used to describe this neoplasm ‘adamantinoma’ has been replaced by ameloblastoma as the neoplasm is not associated with hard tissue formation. Its peak incidence is in the 3rd to 4th decades of life and has an equal sex distribution. It is often associated with an unerupted third molar. The vast majority of ameloblastomas arise in the mandible, and the majority of these are found in the angle and ramus region. They are symptomless until the swelling becomes obtrusive. Although maxillary ameloblastomas are less common, they are potentially lethal, especially when the maxillary sinus is involved or tumor cells invade through bone into the soft tissues. There are three variants of ameloblastomas, namely multicystic, peripheral, and unicystic tumors. The term unicystic is derived from the macro 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. Here we present a case of a unicystic mandibular Ameloblastoma in a 28-year-old male.

CASE REPORT

A 28-year-old male patient reported to Department of Oral Medicine and Radiology, Institute of Dental Sciences, Bareilly, Uttar Pradesh, with a chief complaint of bleeding and pus discharge from lower left back tooth region since 15 days. History of present illness revealed that initially pain was present in the same quadrant due to decayed tooth. The patient had undergone extraction of the same tooth 15 days back. Extraoral examination revealed that there was gross facial symmetry with diffuse swelling measuring about 1 × 1.5 cm on the left side of the face extending anteroposteriorly 1 cm away from corner of mouth to angle of mandible and superioinferiorly from tragus of ear to lower border of mandible. Color of overlying skin was pale red in color. On palpation, all inspectory findings were confirmed it is nontender and there was no local rise in temperature.

Intraoral examination revealed 37 and 38 were missing with a unhealed socket w.r.t 37 and vestibular obliteration was present w.r.t 37 and 38. Under soft tissue examination a diffuse swelling was present on left retromolar area roughly measuring around 1.5 cm in diameter, roughly round in shape. Color of the overlying skin was pale red in color (Fig. 1). On palpation, all inspectory findings were...
its hard consistency and was nontender provisional diagnosis of a residual cyst was given w.r.t 37, with a differential diagnosis of ameloblastoma was given. Under routine investigations orthopantomogram and biopsy was considered. Orthopantomogram revealed a well defined radiolucency with sclerotic borders seen in left mandibular molar region, roughly oval in shape, measuring approximately 1.5 × 3 cm in size extending from coronoid process to angle of mandible (Fig. 2). On the basis of radiographic findings, radiographic differential diagnosis of ameloblastoma and odontogenic keratocyst was given. Histopathological examination showed fibrous wall of cyst infiltrated by long anastomosing cords or larger sheets of odontogenic epithelium, cords of sheet bounded by columnar or cuboidal ameloblast—like cells and loosely arranged epithelial cells (Fig. 3). Based on histological findings, final diagnosis of unicystic ameloblastoma in left retromolar region w.r.t 37, 38 was given.

DISCUSSION

Ameloblastomas are benign tumors whose importance lies in its potential to grow to enormous size with resulting bone deformity. Unicystic ameloblastoma is the second and far less frequent growth pattern seen among all the intraosseous ameloblastoma. Unicystic ameloblastoma, was ameloblastoma is a rare type of ameloblastoma, accounting for about 5 to 10% of intraosseous ameloblastomas. Unicystic ameloblastoma is a distinct entity and on histologic examination unicystic ameloblastoma shows the mural extension into the cystic wall that is the frequently seen, and the term mural unicystic ameloblastoma is used when the thickened lining (either plexiform or follicular) penetrates the adjacent capsular tissue.

The pathogenesis of cystic ameloblastomas remains obscure. There have been many debates regarding whether unicystic ameloblastoma develops de novo or arises in an existing cyst. Leider et al proposed three pathogenic mechanisms for the evolution of unicystic ameloblastoma: (1) reduced enamel epithelium, (2) from dentigerous cyst and (3) due to cystic degeneration of solid ameloblastoma. The neoplasm originates within the mandible or maxilla from epithelium that is involved in the formation of teeth. Potential epithelial sources include enamel organ, odontogenic rests (cell rest of Malassez, cell rest of Serre) reduced enamel epithelium and epithelial lining of odontogenic cyst especially dentigerous cyst. More than 90% are located in the mandible. Between 50 and 80% of cases are associated with tooth impaction, the mandibular third molar being most often involved.

The radiographic appearance is important in the diagnosis; which determines whether the lesion is unilocular, a necessary criterion for unicystic ameloblastoma. Lesions are usually demarcated and may even corticated. The two main radiographic appearances of unicystic ameloblastoma
has been divided into: unilocular and multilocular and these have clear preponderance for the unilocular pattern.\textsuperscript{11} Eversole et al identified predominant radiographical patterns for UCA: unilocular, scalloped, pericoronal, intradicular or periapical expansile radiolucencies.\textsuperscript{12} Ackermann et al classified this entity into the following three histologic groups:\textsuperscript{2}

- **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 UA (invasive islands of ameloblastomatous epithelium in the connective tissue wall not involving the entire epithelium).

Different types of treatment option has been proposed for the management of ameloblastoma, ranging from curettage to a combination of surgery and radiation therapy.\textsuperscript{13} 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% by marsupialization followed by enucleation (where the lesion reduced in size).\textsuperscript{8,14} Thus, the overall prognosis for unicystic ameloblastoma is considerably better than other variants.\textsuperscript{15} Literature suggested that even unicystic ameloblastoma are associated with 10% recurrences and hence require a systemic follow-up.\textsuperscript{3}

**CONCLUSION**

Ameloblastoma is a true neoplasm of odontogenic epithelial origin. It is the second most common odontogenic neoplasm, and only odontoma out numbers it in reported frequency of occurrence, with strong predilection for the posterior region of the mandible ameloblastoma is a tumor with a strong propensity of recurrence, especially when the ameloblastic focus penetrates the adjacent tissue from the wall of the cyst. Radiographically, most of ameloblastoma show multilocularity, whereas unicystic ameloblastoma show a single large unilocular radiolucency. Several clinico-radiological and histological types are included in the ameloblastoma. Apart from the most commonly encountered clinicopathologic models there are few variants, whose biological profile is unknown or not elicited.

**REFERENCES**