Unicystic Mural Ameloblastoma : A Report of Two Cases

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Abstract

Many benign lesions cause mandibular swellings, and these can be divided into odontogenic and non-odontogenic origin. The most common tumor of odontogenic origin is ameloblastoma which develops from epithelial cellular elements and dental tissues in their various phases of development. Ameloblastomas are well-publicized, ectodermal, odontogenic tumors that are reported to constitute about 1 to 3 percent of tumors and cysts of the jaw. Ameloblastoma is a noncancerous (benign) tumor that develops most often in the premolar-molar region of jaw. Ameloblastoma begins in the cells that form the protective enamel lining of teeth. The term ameloblastoma includes several clinico-radiological and histological types. Among these types, unicystic ameloblastoma is the least encountered variant of the ameloblastoma. As this tumor shows considerable similarities with dentigerous cysts, both clinically and radiographically, the biologic behaviour of this tumor group was reviewed. We report a series of cases of unicystic mural ameloblastoma with female predominance with review of literature.

Keywords: Odotogenic Tumour, Unicystic Ameloblastoma, Dentigerous cyst.

Introduction

Many benign lesions cause mandibular swellings, and these can be divided into odontogenic and non-odontogenic origin. 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 accounts for 1% of all cysts/tumors of jaws and 18% of all odontogenic neoplasms and potential to grow to enormous size which results in bone deformity. They are typically classified as unicystic, multicystic, peripheral and malignant subtypes. [1]

In 1977 Robinson and Martinez[2] identified a subtype of ameloblastoma called unicystic ameloblastoma (UA). Prior to 1977 it was termed as mural, monocyclic, intracystic, cryptogenic or cystic ameloblastoma and ameloblastoma developing in a radicular cyst.[3] Unicystic ameloblastoma, which encounters about less than 10-15% of all intra-osseous ameloblastomas, with a prevalence of younger age group (2nd and 3rd decade)[4], referring to those cystic lesions that show clinical and radiographic 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.[2] UA is believed to be less aggressive and possess a

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much better prognosis after enucleation or curettage than does solid or multicystic ameloblastoma.[5] The gender distribution shows a slight male predilection with a male to female ratio of 1.6:1.[4] This paper illustrates case report of patients of unicystic ameloblastoma of the mandible.

Case report 1

An 18-year-old female patient reported to the outpatient department with a chief complaint of pain and swelling in the lower right back tooth region since 15 days. Patient was apparently normal before that and gave history of swelling which was gradually increasing in size. Pain was dull in nature and intermittent and aggravated during mastication and relived upon rest. No history of pus discharge and bleeding from the swelling. On extra-oral examination, a diffuse swelling was seen on the right side of the angle of the mandible which was measuring approximately 2x2 cm in size. Overlying skin was normal, no visible pulsations and no discharge were seen. On palpation, the swelling was firm in consistency and tender and no local rise of temperature was felt. It was non-reducible, non-compressible and non-fluctuant. A single right submandibular lymph node of size measuring about 0.5×0.5 cm in size was palpable, which was firm in consistency, mobile, and tender. On intraoral examination, an oval shaped swelling measuring approximately 5x4 cm in size was seen extending from mesial aspect of right mandibular 2nd molar till the retromolar area obliterating buccal and lingual vestibule with missing 1st and 3rd mandibular molars. On palpation the swelling was tender to touch with firm in consistency on buccal side and soft in consistency on lingual side with crepitus felt on the buccal aspect of 2nd molar region (Fig. 1). 2nd molar was found to be Grade-I mobile. On needle aspiration, straw colored fluid was observed (Fig. 2). Based on the patient's history and clinical findings, the case was provisionally diagnosed as Dentigerous Cyst. Clinical differential diagnosis of Ameloblastoma, Odontogenic Keratocyst (OKC) and Odontogenic Myxoma were considered.

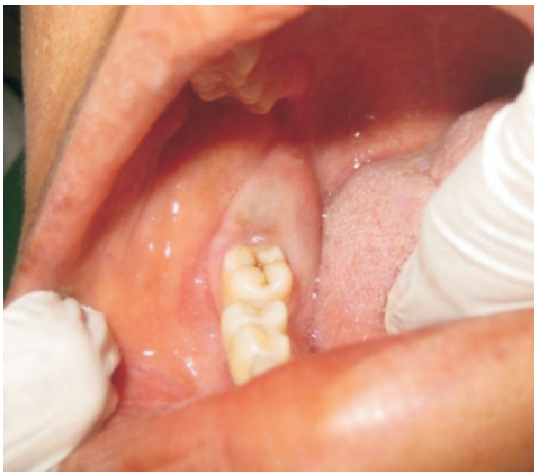

Fig. 1:Intra-oral photograph showing the swelling in relation to 47 extending till the retromolar region

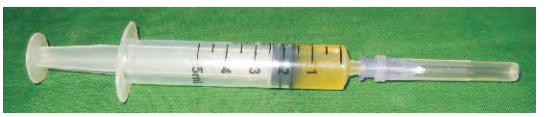

Fig. 2: Aspiration of Cystic fluid

Panoramic Radiograph revealed a well defined unilocular radiolucency with impacted 3rd mandibular molar and missing 1st molar with mesially tilted 2nd molar (Fig. 3). Lateral oblique view of ramus of mandible shows a well defined unilocular radiolucency (Fig. 4). CT scan revealed expansion and perforation of the cortical plates and the extent of lesion on the right side of the ramus of mandible (Fig. 5&6). Incisional biopsy was done and submitted for histopathological examination. The biopsy showed the cystic lining of 3-4 layered thickness of proliferation of ameloblastomatous epithelium with

nuclear palisading along the margins and loose stellate reticulum, containing moderate to abundant pale acidophilic vacuolated cytoplasm and round to oval vesicular nucleus. The underlying connective tissue showed islands of odontogenic epithelium with peripheral ameloblast like cells and central stellate reticulum type of tissue. Based upon the radiological and histopathological report, the case was diagnosed as Unicystic Mural Ameloblastoma.

Case report 2

A 22-years old female patient visited to our department with a chief complaint of pain and swelling in the lower left back tooth region since 1 week. Patient gave history of pain associated with swelling in the same region about 1 month back with pus discharge intra-orally subsequent to which patient consulted local physician and had taken medication. On extra oral examination, a diffuse swelling measuring approximately 1 x 2 cm over the angle of the mandible with no visible pulsations and no discharge were seen and overlying skin was normal. On palpation, swelling was painful, firm in consistency with no local rise of temperature. It was non-pulsatile, non-compressible and non-fluctuant. Intra-orally, a diffuse swelling was seen in the lower left buccal and lingual vestibule with respect to mandibular 1st and 2nd molar and partially erupted 3rd molar, measuring about 2 x 2 cm in size. It extended anterio-posteriorly from distal aspect of 1st molar to retromolar area and superio-inferiorly from the gingival margin of 2nd and 3rd molar to the depth of vestibule (obliterating it). On palpation the swelling was tender to touch with bony hard consistency, and crepitus felt over the 2nd and 3rd molar region, both buccally and lingually. It was non-pulsatile, non-compressible, non-fluctuant and no discharge was noticed and no pulsation was felt (Fig. 7). Aspiration revealed 0.5 ml of the thick yellow colored pus from cystic lesion. Based on the above findings, provisional diagnosis was given as Infected Dentigerous Cyst. Clinical differential diagnosis of Ameloblastoma, Odontogenic Keratocyst (OKC) and Odontogenic Myxoma / Calcifying Epithelial Odontogenic Tumour were considered.

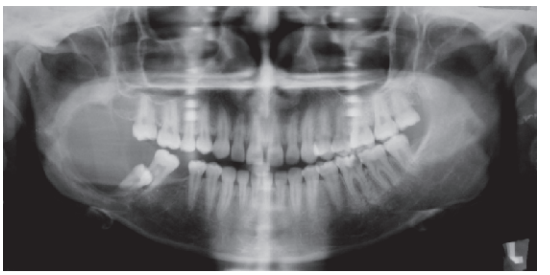

Fig. 3: Panoromic Radiograph shows an extensive well defind unilocular radiolucency on the right side of mandible extending from mesial of 47 to the ascending ramus. Inferiorly displaced impacted 48 is also seen

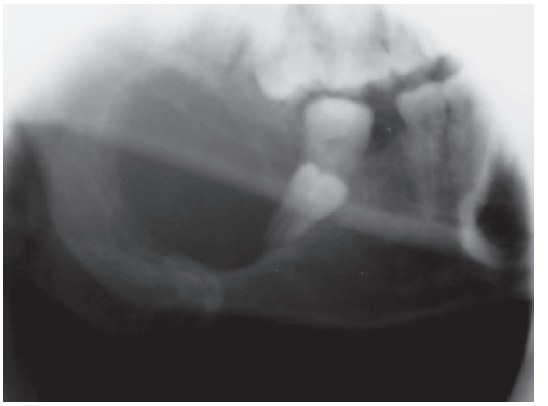

Fig. 4: Lateral oblique view of mandible shows findings similar to the panoramic view

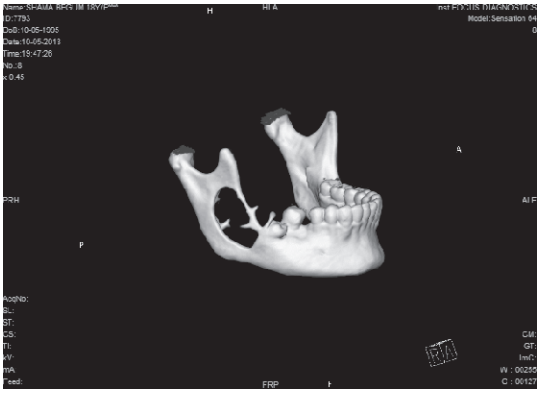

Fig. 5: Computed Tomography of mandible with 3D reconstruction showing the osteolytic lesion in the region of 47 and 48 extending on the ramus

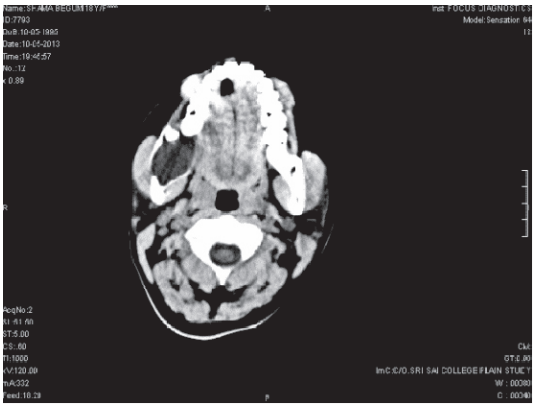

Fig. 6: Axial view of CT Scan showing the osteolytic lesion in the right side of mandible with expansion of buccal & lingual cortical plates

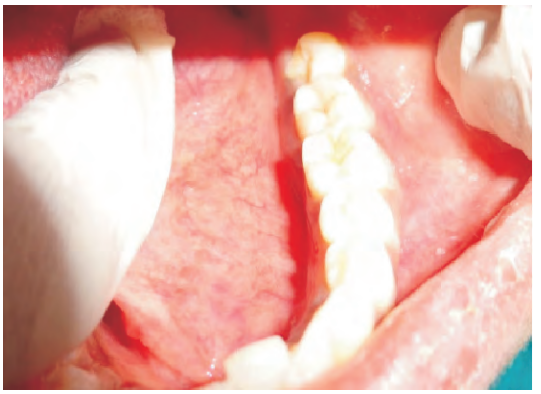

Fig. 7: Intra-oral photograph showing obliteration of buccal vestibule and partially erupted 38

Mandibular lateral occlusal view showed expansion of lingual cortical plate with perforation (Fig. 8). Panoramic Radiograph revealed a well defined radiolucency with corticated borders and partially erupted 3rd molar with resorption of root in relation to 2nd and 3rd molar (Fig. 9). CT scan revealed expansion and perforation of the cortical plates and the extent of lesion in relation to 2nd and 3rd molar (Fig. 10). Incisional biopsy was done and submitted for histopathological examination. The histopathological report showed 1-2 layers of cystic epithelial linings with transformation to cuboidal or columnar basal cells with hyperchromatic nuclei and nuclear palisading. The proliferation of ameloblastomatous tissue resembling plexiform pattern with intra-murally ameloblastomatous tissues are seen. (Fig. 11). Based upon the radiological and histopathological report, the case was diagnosed as Unicystic Mural Ameloblastoma.

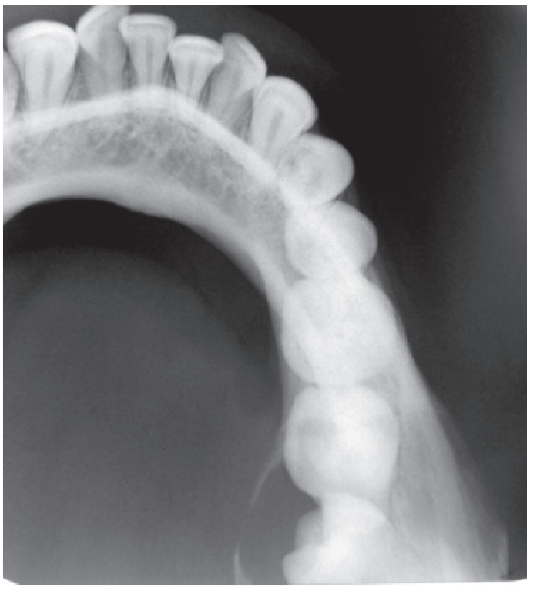

Fig. 8: Lateral occlusal view of mandible showing radiolucent lesion in the region of 37 & 38 with expansion of buccal & lingual cortical plates

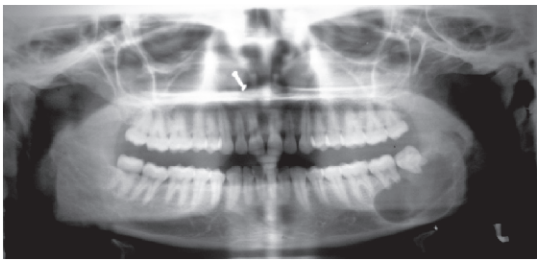

Fig. 9: Panoramic view showing a large well defined unilocular radiolucency at the apices of 36,37&38 causing resorption of the roots

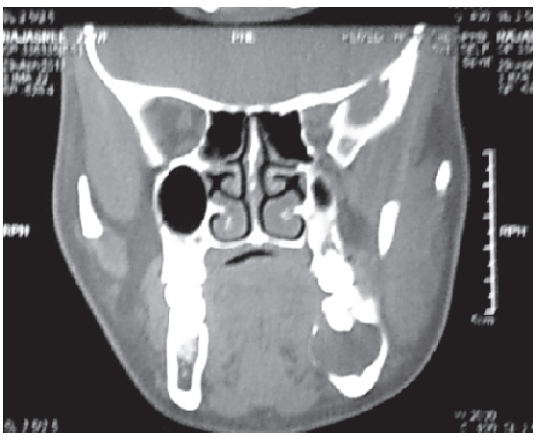

Fig. 10: Coronal view of CT Scan shows an osteolytic lesion causing expansion of buccal & lingual cortical plates in the left of mandible

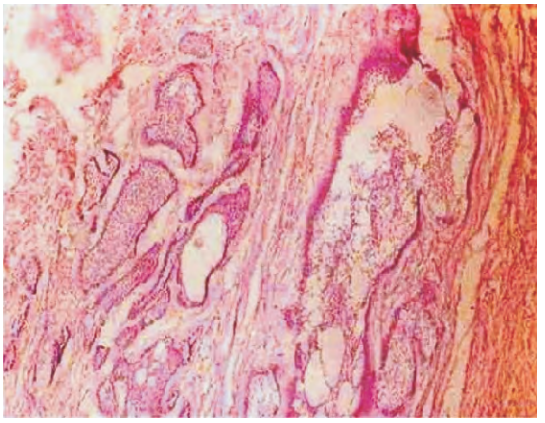

Fig. 11: Histopathological view, showing mural plexiform ameloblastomatous tissue proliferation.

Discussion

Ameloblastoma is a benign, locally aggressive odontogenic neoplasm with variable clinical expression. It is typically slow growing, locally aggressive and rarely metastasizes but has a high rate of recurrence (55–90%) if not removed adequately.

As per the WHO system of 2003, ameloblastoma is classified based on differences in biologic behavior, treatment plan and recurrence rate as follows:

(1) Classic solid/multicystic ameloblastoma,

(2) Unicystic ameloblastoma,

(3) Peripheral ameloblastoma,

(4) Desmoplastic ameloblastoma, including the so-called hybrid lesions.[6]

Unicystic ameloblastoma (UCA) is a rare type of ameloblastoma, accounting for about 6% of ameloblastomas. It usually occurs in a younger age group of 16–20 years, with about 50% of the cases occurring in the second decade of life as seen in case report 1.[7] The gender distribution shows a slight male predilection with a male to female ratio of 1.6:1. However, when the tumor is not associated with an unerupted tooth, the gender ratio is reversed to a male to female ratio[8] of 1:1.8 which is in consistent to the case report 1. More than 90% are located in the mandible in the posterior region followed by the parasymphysis region, the anterior maxilla, and the posterior maxilla. UCA is usually asymptomatic, although a large tumor may cause painless swelling of the jaws with facial asymmetry.[7] The clinical and radiographic findings in most cases of unicystic ameloblastoma suggest that the lesion is an odontogenic cyst, particularly dentigerous cyst. However, few are not associated with impacted teeth which are called non-dentigerous variant.[9] The mean age of non impacted tooth-related cystic ameloblastoma was 5 years in comparison to16.5 years for the impacted tooth-related variant.[4] Most of the UCAs are associated with an impacted tooth, the mandibular third molar being involved most often, as in the case report 1. These findings correlate with those reported by Philipsen et al. and Ackermann et al.[3,4]

The radiographic appearance of UCA has been divided into 2 main patterns: unilocular and multilocular, and these have clear preponderance for the unilocular pattern. This preponderance is marked for the dentigerous variant, where the unilocular to multilocular ratio is 4.3:1, and for the non dentigerous type, this ratio is 1.1:1.[10] The involved teeth show varying degrees of root resorption.[6] Eversole et al.[1] and Paikkatt et al.[10] identified predominant radiographical patterns for UCA: unilocular, scalloped macromultilocular, pericoronal, inter-radicular, or periapical expansile radiolucencies. The early ameloblastic changes within the cyst wall were first described by Vickers and Gorlin[11] in 1970, and their histologic criteria for the diagnosis of unicystic ameloblastoma

includes a cyst lined by ameloblastic epithelium with a tall columnar basal layer, sub-nuclear vacuoles, reverse polarity of hyperchromatic nucleus, and a thin layer of edematous, degenerating stellate reticulum-like cells on the surface. The mural extension into the cystic wall is the frequently seen feature, and the term mural UCA is used when the thickened lining (either plexiform or follicular) penetrates the adjacent capsular tissue.[3,7]

In a clinico-pathologic study of 57 cases of unicystic ameloblastoma, Ackermann[4] classified this entity into the following three histological groups:

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)

According to this classification, case reports presented here belong to Group III.

Histologic sub-grouping by Philipsen and Reichart has also been described:[3]

Subgroup 1—luminal UA

Subgroup 1.2—luminal and intraluminal

Subgroup 1.2.3—luminal, intraluminal and intramural

Subgroup 1.3— luminal and intramural.

A definitive diagnosis of unicystic ameloblastoma can only be done by histological examination of the entire lesion and cannot be predicted preoperatively on clinical or radiographic grounds. As incisional biopsy is not representative of the entire lesion, it may result in an incorrect classification. The epithelial lining of a UCA is not always uniformly characteristic and is often lined partly by a non-specific thin epithelium that mimics the dentigerous cyst lining. Thus, true nature of the lesion becomes evident only after enucleation when the entire specimen is available for microscopy.[9]

Ackermann et al.[2] (1988) and Robinson and Martinez[4] (1977) argued that as the epithelium of odontogenic cysts and ameloblastomas have a common ancestry, a transition from a non-neoplastic to a neoplastic one could be possible, even though it occurs infrequently. Leider et al.[12] (1985) proposed three pathogenic mechanisms for the evolution of UA, from reduced enamel epithelium, dentigerous cysts and due to cystic degeneration of solid ameloblastoma. The cases presented here are in support with second hypothesis, arising from preexisting dentigerous cyst as they are in association with an impacted tooth and presence of non specific thin epithelium lining in focal areas of cystic tumor.

However, immunohistochemical markers like lectins and proliferating cells (proliferating cell nuclear antigen PCNA and Ki-67 may assist in their differential diagnosis[13]. However, Eversole et al.[12] contend that currently unaided histologic assessment for UCA remains the gold standard for diagnosis, because of a variable response of UCA to tissue markers.

Histologically, the minimum criteria for diagnosing a lesion as UCA are the demonstration of a single cystic sac lined by odontogenic (ameloblastomatous) epithelium often seen only in focal areas[14]. Treatment planning depends on the histological type of UA. The UA which is diagnosed as subgroups

1 and 1.2 may be treated conservatively (careful enucleation), whereas subgroups 1.2.3 and 1.3 should be treated aggressively[8] and cases presented here were treated conservatively with careful surgical enucleation.

Lau and Samman[15] 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 is reduced in size). Whatever surgical approach the surgeon decides to take, long-term follow-up is mandatory as recurrence of unicystic ameloblastoma may be long delayed.

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