CASE REPORT

DIAGNOSIS & MANAGEMENT OF UNICYSTIC AMELOBLASTOMOUS LESION OF THE MANDIBLE – A CASE REPORT

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DOI: 10.5455/ijmsph.2013.120620132 **Received Date: 07.05.2012** **Accepted Date: 12.06.2013**

ABSTRACT

Ameloblastomas are an enigmatic group of tumours. Their name implies a resemblance to cells of enamel forming organ (dental hard tissues). Ameloblastoma is an odontogenic tumour which is an archetype of a neoplasm where the neoplastic component is epithelial only without contribution from the ectomesenchyme. Most common odontogenic neoplasm in Indian population. Ameloblastoma is locally aggressive, but often asymptomatic, showing a slow growth which is manifested as a facial swelling or radiographic incidental finding. Unicystic ameloblastoma refers to those cystic lesions that show clinical, radiographic, or gross features of a mandibular cyst, but on histological examination show a typical ameloblastomatous epithelium lining with or without luminal tumour growth. We present an unusual case of unicystic ameloblastoma appearing as an interdental radiolucent lesion of the mandible in a 26 years old male patient. In addition, an insight on the various pathologies such as radicular cyst, lateral periodontal cyst, collateral keratocystic odontogenic tumour, Squamous odontogenic tumour, central giant cell granuloma, extra follicular adenomatoid odontogenic tumour, central odontogenic myxoma, glandular odontogenic cyst, calcifying epithelial odontogenic tumour, with similar presentations were critically evaluated & discussed in differential diagnosis of such entities.

Key-Words: Ameloblastoma; Mandible; Unicystic Plexiform Variety

Introduction

Many benign lesions cause mandibular swellings, & these can be divided into those of odontogenic & non odontogenic origin. Lesions include lateral periodontal cyst, collateral KCOT, CGCG, fibro osseous lesions, ameloblastoma, osteomas, etc.[1-6] The term ameloblastoma was coined by Churchill in 1933. Robinson described it as 'Usually unicentric, nonfunctional, intermittent in growth, anatomically benign & clinically persistent'.[1]

Ameloblastoma in the mandible can progress to great size & cause[6]:

- Facial asymmetry
- Displacement of teeth
- Teeth mobility
- Resorption of roots of teeth
- Pathologic fractures.

In 1977, Robinson & Martinez first contributed the term 'unicystic ameloblastoma'. Unicystic ameloblastoma is a rare type of ameloblastoma accounting for about 6% of ameloblastomas.[7] Small lesions are sometimes discovered more on routine radiographic screening examinations or as result of local effects like tooth mobility, facial asymmetry, occlusal alterations) produced by the developing tumour. The unicystic ameloblastoma is considered a variant of the solid ameloblastoma, accounting for 6-15% of all intraosseous ameloblastomas, more than 90% of the cases the unicystic ameloblastoma is located in the mandible.[8]

Case Report

A 26 years old male patient reported to the department of oral & maxillofacial surgery, Tatyasaheb kore dental college & research centre, with an asymptomatic hard swelling on the right side of lower jaw since 6 months. Medical & social histories were unremarkable & non-contributory. His physical examination revealed no abnormality other than those related to the chief complaint. No remarkable finding seen on extra oral head & neck examination. There was no evident lymphadenopathy. Clinical examination revealed bony hard, non-tender mass over right side of the lower jaw in relation to 44, 45 region which was covered by normal pinkish red, intact & immobile mucosa (figure 1).

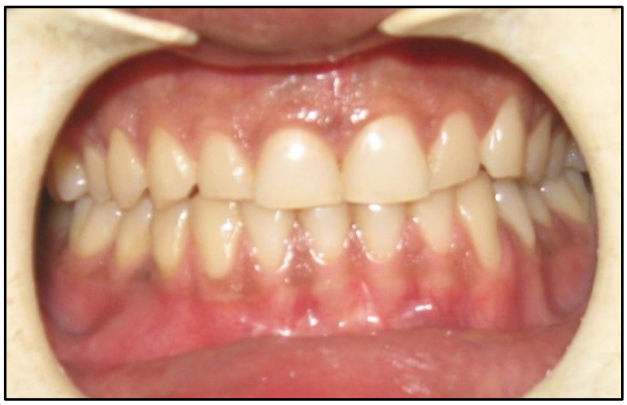

Figure-1: Preoperative Intra Oral View of the Lesion Showing Vestibular Obliteration

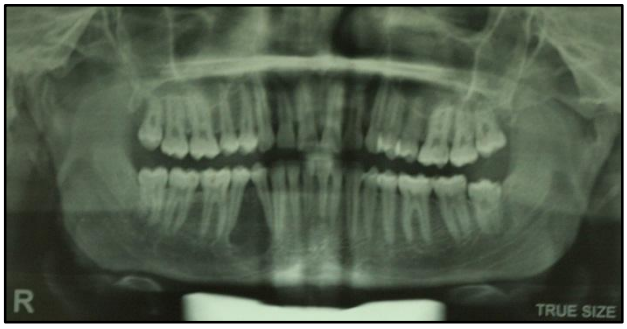

Figure-2: Pre-Operative OPG Showing Well Defined Radiolucent Lesion in Relation to 43, 44, 45 Region (RCT Treated – 44, 45)

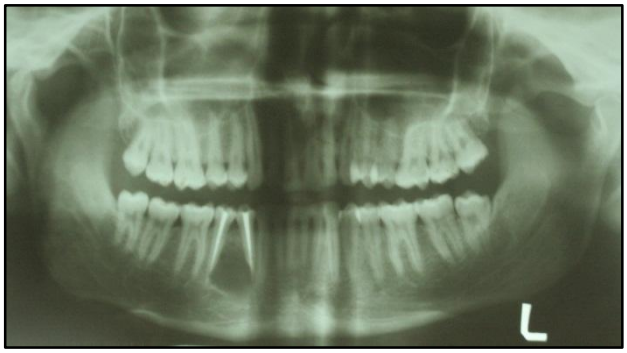

Figure-3: Pre-Operative OPG Showing Well Defined Radiolucent Lesion in Relation to 43, 44, 45 Region (RCT Treated – 44, 45)

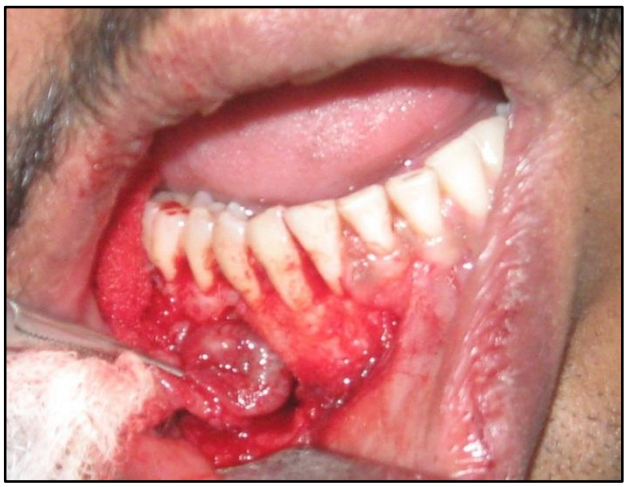

Figure-4: Identification of the Cystic Lining & Relationship with Mental Neurovascular Bundle

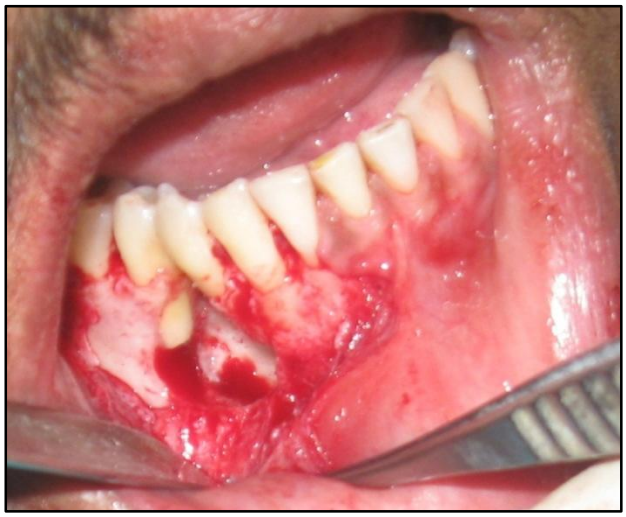

Figure-5: Intoto Retrieval of the Lesion

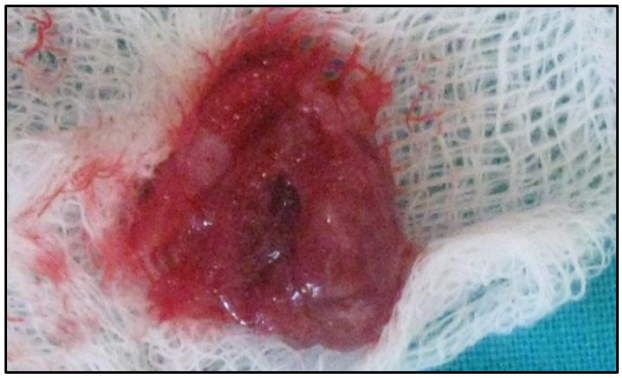

Figure-6: Excised Specimen In Vitro

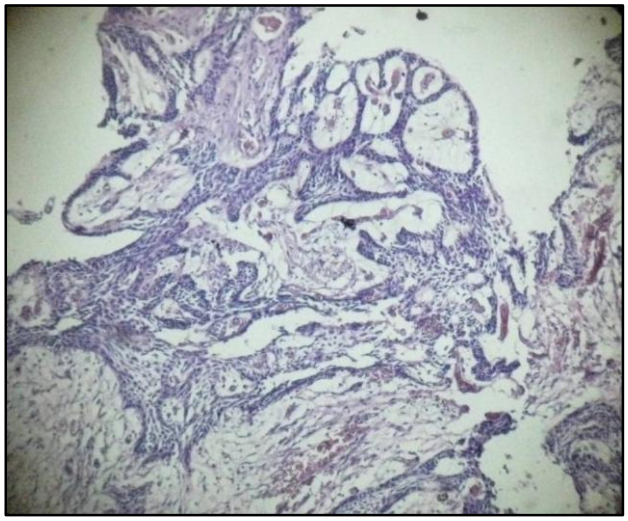

Figure-7: Photomicrograph of the Excised Specimen Suggestive of Plexiform Ameloblastoma

Panoramic radiography revealed a well-defined unilocular radiolucent lesion measuring approximately 0.75 X 1.25cm in diameter evident over the right side of mandible extending from the distal aspect of 43 till the mesial aspect of 46 (root canal treated teeth – 44, 45). The radiolucency was corticated & separated by thick septae with scalloped borders showing typical honey comb appearance. Displacement of 44, 45 evident with significant resorption of roots. Overall

radiographic picture was suggestive of unicystic benign tumor of odontogenic origin (figure 2, 3).

Fine needle aspiration of the fluid was done & sent for biochemical examination which revealed protein content of 6.1gms%. Based on the history & clinical examination a provisional diagnosis of benign odontogenic lesion was given. After obtaining the informed surgical consent, patient was taken up for surgical enucleation with curettage under local anesthesia & conscious sedation of the said lesion via intra oral approach. After raising mucoperiosteal flap (figure 4) the lesion was identified & meticulous dissection was carried out to preserve the right mental neurovascular bundle which was in close vicinity with the lesion. In toto retrieval of the lesion including a margin of normal tissue was done respectively (figure 5). Curettage, apicectomy in relation to 44, 45 carried out under copious irrigation of normal saline to avoid thermal necrosis of the bone. Haemostasis achieved. Excised tissue specimen was stored in 10% formalin solution & subjected for histopathological evaluation (figure 6). Wound closure achieved using black braided silk suture material. Histopathological findings were suggestive of unicystic plexiform ameloblastoma (figure 7). At his first follow-up after 1 month later, he had no significant complaints except for slight discomfort over cheek region.

Differential Diagnosis

Based on the clinical (table 1) & radiographic interpretation (table 2), various possible odontogenic & mesenchymal lesions presenting as interdental radiolucent lesions of the mandible in the given clinical scenario would be one of the following enlisted entities[9]:

- Lateral periodontal cyst
- Collateral keratocystic odontogenic tumour
- Ameloblastoma
- Central giant cell granuloma
- Calcifying epithelial odontogenic tumour
- Odontogenic myxoma
- Squamous odontogenic tumour
- Extra follicular adenomatoid odontogenic tumour
- Glandular odontogenic cyst.

The first in the clinical differential diagnosis was the most common lesion at this site to be considered - **Lateral Periodontal Cyst**, which is rare amongst developmental odontogenic cysts. Presents as asymptomatic gingival swelling, localized in canine & premolar region with significant loss of lamina dura.[10,11]

Table-1: Clinical Differential Diagnosis

Features	Ameloblastoma	Keratocystic Odontogenic Tumour	Central Giant Cell Granuloma	Calcifying Epithelial Odontogenic Tumour
Incidence	11% of odontogenic tumours	7-11% of odontogenic tumours	< 7% of benign lesions of the jaw	0.4-3% of odontogenic tumours
Age	Third, fourth decade	Second, third decade	< 20 yrs	8-92 years
Sex	M > F	M > F	F > M	M > F
Signs & Symptoms	Painless swelling, slow growth	Painless swelling	Aggressive; pain & swelling, rapid growth	Painless swelling & slow growth
Site & Location	Mandibular posterior ramus	Mandibular molar- ramus	Mandible > Maxilla; anterior to first molar, may cross midline	Mandible: Maxilla -2:1 Premolar-molar region

Table-2: Radiological Differential Diagnosis

Features	Ameloblastoma	Keratocystic Odontogenic Tumour	Central Giant Cell Granuloma	Calcifying Epithelial Odontogenic Tumour
Periphery & Shape	Smooth, oval	Smooth, round & oval	Irregular, double boundary	Irregular
Borders/ Margins	Well defined & often curved	Well corticated & scalloped	Well defined, smooth & scalloped	Mandible-well corticated Maxilla- poorly defined
Cortical Expansion	Both lingual & buccal	Minimal expansion, rare	Uneven expansion, both buccal &lingual perforation	Significant expansion
Internal Structure	Multilocular/unilocular, honey comb/soap bubble appearance	Radiolucent	Ill defined	Multilocular, tennis racket, step ladder
Associated with Impacted Tooth	38%	-	-	-
Root Resorption	Extensive	Rare	-	Rare

The first in the clinical differential diagnosis was the most common lesion at this site to be considered - **Lateral Periodontal Cyst**, which is rare amongst developmental odontogenic cysts. Presents as asymptomatic gingival swelling, localized in canine & premolar region with significant loss of lamina dura. [10,11]

Collateral KCOT occurs in the mandibular posterior region which is the next common lesion in this location. Clinically, it manifests as pain, swelling associated with paraesthesia of lip. Tendency to grow in anterior posterior direction with in medullary cavity without causing obvious bone expansion, aspiration may reveal thick, yellow & cheesy material.[1,11]

Central Giant Cell Granuloma is a benign proliferative lesion common seen in young females, typically occurs anterior to the first molar & often crosses the midline. Discovered accidentally, may present with no signs & symptoms, displacement of teeth, evidence of root resorption of associated teeth.[1]

Calcifying Epithelial Odontogenic Tumor has a predilection for the posterior areas of the mandible (premolar-molar area).

Central Odontogenic Myxoma may occur anywhere in the jaws but have predilection for premolar molar region of the mandible & maxilla. Lesions may cross midline. Radio graphically, displacement of teeth, cortical expansion & perforation is common. In the early stage it has an osteoporotic appearance, consisting of multilocular radiolucency with well-developed locules.[1,7]

Squamous Odontogenic Tumor is an intraosseous lesion associated with vital teeth. Mobility of teeth, local pain, and swelling of the gingival, found in all age groups. Mandible more commonly affected than maxilla. Triangular shaped semicircular radiolucency associated with roots of teeth.[4,7,10]

Glandular Odontogenic Cyst is an uncommon jaw cyst, mandibular anterior region being the common site. Asymptomatic condition, but occasionally accompanied by pain.[11,12]

Discussion

Ameloblastomas are an enigmatic group of oral tumors. Their name implies a resemblance to cells of developing dental tissues (ameloblasts-prime cells in the developmental of dental tissues).[2]

Ameloblastoma is a tumor originated from the epithelium involved with the formation of teeth. Representing 1% of all tumors & cysts that involve maxillomandibular area & about 10% of odontogenic tumors.[14]

Ameloblastoma can theoretically arise from remnants of the dental lamina, enamel organ of developing tooth, the epithelial lining of odontogenic cyst or basal cells of the oral mucosa.[14]

Many theories have been put forth discussing possible pathogenic mechanisms, but several causative factors have been proposed such as[15],

- Non-specific irritating factors such as extraction, caries, trauma, infection, inflammation, or tooth eruption;
- Nutritional deficit disorders;
- Viral pathogenesis

Ameloblastomas in the mandible can progress to great size & cause facial asymmetry, displacement of teeth, loose teeth, malocclusion, root resorption, pathologic fractures, etc.[3]

Ameloblastoma can be classified in three types, considering clinical & radiographic features: solid/multicystic, unicystic, peripherical.[13]

Histological subtypes of ameloblastoma include- Plexiform, Follicular, Unicystic, Basal cell Granular cell, Clear cell, Acanthomatous, Vascular, Desmoplastic variants[1].

The unicystic ameloblastoma variant of ameloblastoma, first described by Robinson & Martinez in 1977, is reported to have a less aggressive biologic behavior & lower recurrence rate than the classic solid or multicystic ameloblastoma.

Unilocular ameloblastoma is a rare type of ameloblastoma, accounting for about 6% of ameloblastomas. It has been reported previously that unilocular ameloblastomas tend to occur in the younger age groups.[16] Our case report confirmed this tendency.

Small lesions are sometimes discovered more on routine radiographic screening examinations or as a result of local effects produced by the tumour.

Pathogenesis (Hypothesis)

The pathogenesis of cystic ameloblastomas remains obscure. The reason why some ameloblastomas become completely cystic may be related to epithelial dysadhesion (defective desmosomes) or, more likely, to the intrinsic production of proteinases (metalloproteins, serine proteins); enzymes that normally degrade the central zone of enamel organ after tooth development.[17]

Classification of Unicystic ameloblastomas

According to Ackermann GL[8] based on clinicopathologic study of 57 cases of unicystic ameloblastomas classified this entity into following 3 histologic groups,

- Group I: Luminal unicystic ameloblastoma (42%)
- Group II: Intraluminal/Plexiform unicystic ameloblastoma (9%)
- Group III: Mural unicystic ameloblastoma (49%)

According to Philipsen & Reichart[16] histological sub grouping of unicystic ameloblastoma is done into 4 types namely,

- Subgroup 1: Luminal unicystic ameloblastoma
- Subgroup 1.2: Luminal & Intraluminal
- Subgroup 1.2.3: Luminal, Intraluminal & Intramural
- Subgroup 1.3: Luminal & Intramural.

In the present case, the patient was male & was in his third decade of his life. The lesion presented as an dilemma to the clinician, because of its rare & unusual location, hence, the authors critically evaluated the case based on the clinical & radiographic presentation & with the help of biochemical picture following fine needle aspiration of the fluid, & taking into account a set of differential diagnosis that mimic as interdental radiolucent lesions which made it possible to arrive to a provisional & clinical diagnosis & tailor make the treatment plan accordingly.

Following enucleation & curettage of the lesion the specimen was subjected for histopathology & based on the combination of clinical, radiographic, microscopic findings a final diagnosis of plexiform ameloblastoma of the mandible was arrived.

Ameloblastomas are treated by curettage only, enucleation & curettage or radical surgery. However, ameloblastomas that appear as unilocular lesions radio logically may be treated conservatively (i.e., with enucleation or curettage or both) whenever all areas of the cystic lumen can be controlled intraoperatively.[18,19] In the given case surgical enucleation with curettage was selected as the treatment modality which justifies to the statement that 'compared to its multicystic counterpart, the unicystic ameloblastomas tend to be less aggressive & has low recurrence rate'.[6]

As in the reported literature, ameloblastomas are known for its high recurrence rate if excision was incomplete. Since variants of ameloblastomas differ in biologic behavior, appropriate treatment plan has to be formulated as per the given clinical scenario to avoid recurrence.

According to LAU et al[20] recurrence of ameloblastoma is related to,

- Inadequate surgical treatment
- Incomplete removal of tumor or when the treatment consists of enucleation & curettage.

Recurrence rates are related to the type of initial treatment. They are as follows:

a) Resection – 3.6%
b) Enucleation alone – 30.5%
c) Enucleation & carnoy's application – 16%
d) Marsupialization followed by enucleation – 18%.

Post-operative follow up is important in the management of ameloblastoma because more than 50% of all recurrences occur within 5 years

of surgery. Recurrence is also related to histologic subtypes of unicystic ameloblastoma, with those invading the fibrous wall having a rate of 35.7% but others only 6.7%.[18]

Conclusion

Unicystic ameloblastoma is a tumor with a strong propensity for recurrence especially when the ameloblastic focus penetrates the adjacent tissue from the wall of the cyst. On the contrary, if the tumor is confined to the luminal surface of the cyst, than it can be safely enucleated, same evidenced in the present case. This case report emphasizes on the importance of differential diagnosis of various interdental radiolucent lesions of the mandible which is of significance to the clinician for effective treatment planning as well as to prevent recurrence.

ACKNOWLEDGEMENT

The authors would like to thank the patient for giving us the consent & permission for publication of this case report & accompanying images.

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Cite this article as: Kulkarni GH, Khaji SIH, Kulkarni H, Shende S. Diagnosis & management of unicystic ameloblastomous lesion of the mandible – A case report. Int J Med Sci Public Health 2013; 2:1101-1106.
Source of Support: Nil
Conflict of interest: None declared