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A CASE OF UNICYSTIC AMELOBLASTOMA

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Abstract
Ameloblastomas are amongst most commonly occurring, locally aggressive, benign tumors of odontogenic origin worldwide. Unicystic ameloblastomas (UA) are the deceptive lesions of the jaw that mimics odontogenic cyst. Unicystic Ameloblastoma considerably differs from the conventional Ameloblastoma in mean age of occurrence, clinical behaviour, prognosis and treatment outcome. The following report discussed below is a case of a 32 years old male patient who was a diagnosed case of Unicystic Ameloblastoma and was conservatively managed upon first incidence in private clinic which later recurred after a period of 1 year and then was treated at our institute

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INTRODUCTION

Ameloblastomas are the most common locally aggressive benign tumor of odontogenic origin that accounts for 1% of all jaw tumours[1]. It was classified by WHO into 3 variants namely- Solid Multicystic Ameloblastoma (SMA), Unicystic Ameloblastoma (UA) and Peripheral Ameloblastoma. Amongst its three variants, UA accounts for about 10%-15% [2].UA was first described as a distinct entity by Robinson and Martinez in the year 1977[3,4]. It has been called by various names like- *mural ameloblastoma, intracystic ameloblastoma, cystogenic ameloblastoma, cystic ameloblastoma, and plexiform UA*[5].

Definitions have been attempted by various authors to precisely describe about the nature and behaviour of ameloblastomas. Finally it was in the year 1992 when WHO defined Ameloblastoma as "a benign, locally invasive, polymorphic neoplasm consisting of proliferating odontogenic epithelium which usually has a follicular or plexiform pattern lying in a fibrous stroma."

Ameloblastoma's are more common in mandible as compared to maxilla; ratio varying from 3:1 to 13:1 [1,6].In mandible, the frequent sites involved are ascending ramus- molar region followed by premolar and anterior region, percentage of occurrence being 70%, 20% and 10% respectively[7,8].The mean age of occurrence of SMA is 4^{th}-5^{th} decade of life whereas in Unicystic Ameloblastoma its 2^{nd} -3^{rd} decade.UA are monocystic lesion lined by odontogenic epithelium with/ without intraluminal and/or intramural growth.

Conventionally, SMA are multilocular lesions showing soap bubble or honeycomb appearance radiographically. However, 24-47 % of SMA demonstrates unilocular configuration[7,9].UA most often exhibits unilocularity on radiographic examination, however multilocularity can also be encountered especially in non-dentigerous variant of UA.

CASE REPORT-

A 32 year old male patient reported to the OPD at Sharad Pawar Dental College and Hospital, Wardha with the chief complaint of swelling over left side of the face since 5 years. Patient was apparently alright 5 years back when he experienced swelling which was gradual in its onset, associated with pain which was dull, intermittent and non-radiating in its nature. Upon asking past dental history, patient informed about previous extraction of 38 due to deep, unrestorable carious lesion 1 year ago. Patient also gave history of paresthesia on the affected site since 6 months. Radiographic evaluation followed by incisional biopsy was advised to the patient. Upon careful histopathological evaluation, the patient was diagnosed as UA. The patient visited to private practitioner for further needful management. After 6 months, patient again reported at our institute with the complaint of swelling in the same region. Previous treatment records were retrieved from the patient, wherein the patient had underwent conservative management alongwith chemical cauterization using Carnoy's solution. Upon general examination, patient was found to be of average built and height with normal vital signs. On extraoral examination, the size of the lesion was found to be 8x5 cm approximately extending supero-inferiorly from 2cm below the outer canthus of eye upto the lower border of mandible and anteroposteriorly from 2cm behind the corner of mouth to the posterior border of ramus on left side of face. **(Fig.1)**

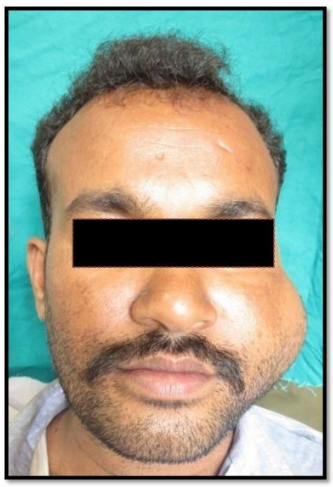


Figure 1- Extraoral photograph

Upon palpation the lesion was tender, firm to bony hard in consistency, non-compressible with no elevation of local temperature and surface changes with single palpable, mobile and tender ipsilateral submandibular lymph node. The pertinent intraoral findings being obliteration of left buccal vestibule. Orthopantomogram showed a large lytic lesion involving the entire ramus of the mandible of left side. **(Fig. 2)**

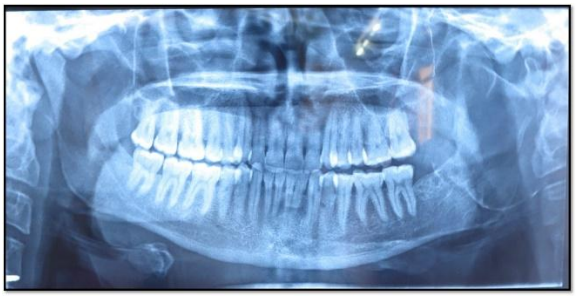
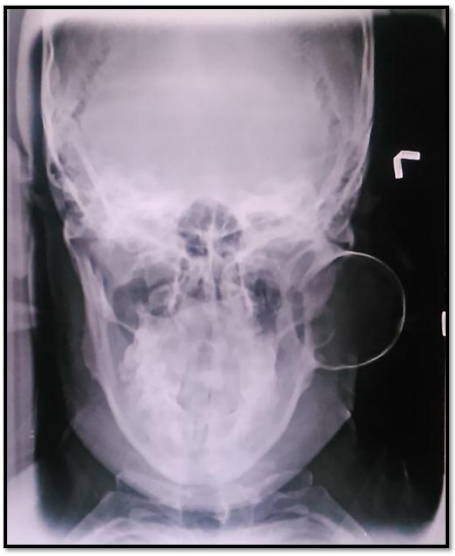


Figure 2- Large unilocular lytic lesion in left ramus of the mandible

The PA mandible showed cortical plate expansion.



(Fig.3)

Figure 3- Buccal cortical plate expansion on left side mandible

Taking into account previous history of UA, incisional biopsy was skipped this time. The tumour was resected under general anaesthesia with all aseptic precaution and was then further sent for histopathological evaluation.

Histopathologic examination:

Under scanner view (4x), H&E stained tissue section showed cystic cavity lined by epithelium and underlying connective tissue stroma. **(Fig.4)**

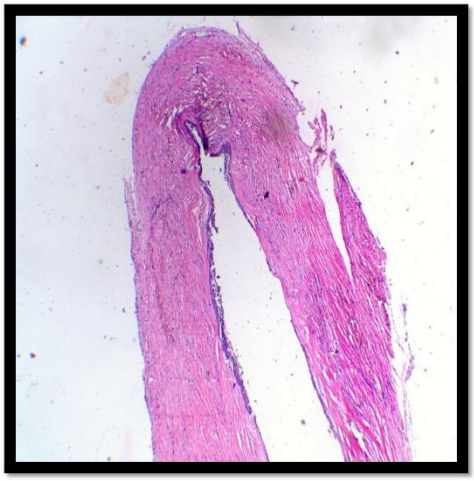


Figure 4 – Scanner view (H & E stain)

Under low power view (10X), H & E stained tissue section showed cystic cavity lined by odontogenic epithelium. The odontogenic epithelium was proliferating towards the cystic lumen in a plexiform pattern.
(Fig.5)

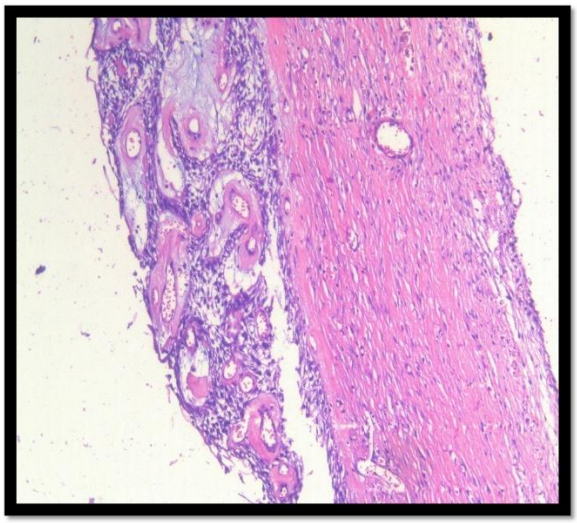


Figure 5- Low power view (H&E stain) demonstrating intraluminal proliferation of odontogenic epithelium in plexiform pattern

Under high power view (40X), islands of tall columnar cells with hyperchromatic nuclei suggesting pre-ameloblast like cells were appreciated in the connective tissue stroma.
(Fig.6)

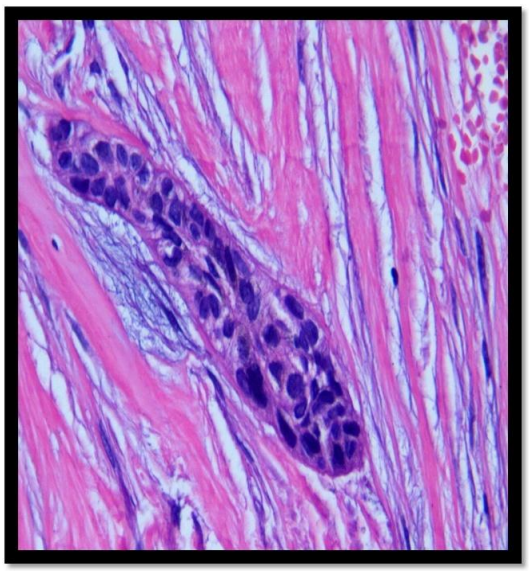


Figure 6- High power view (H&E stain) demonstrating odontogenic island in the connective tissue.

With these histopathologic findings, final diagnosis of Unicystic Ameloblastoma subtype 1.2.3 was made.

DISCUSSION:

UA accounts for 6% of ameloblastomas that are encountered in young age, 50% of cases at the time of diagnosis being in the 2nd decade of life[10]. Intraosseous Ameloblastoma's can be classified into conventional SMA and UA [7].
UA is an odontogenic tumour of less aggressive biologic behaviour with lower recurrence rate than the classic SMA.UA, depending upon presence of impacted tooth within the tumour clinico-radiographically further, is sub-classified into dentigerous and non-dentigerous variant, dentigerous being more common in younger age group.The 'dentigerous' type occurs 8 years earlier on average than the 'non-dentigerous' variant [11].In contrast to conventional SMA-, UA affects at early age, age range being 19-27 years.**(Table 1)** .

Variants of UA	**Mean age of occurrence**
Dentigerous	16.5 years
Non-dentigerous	35.2 years

Table 1- Variants of Unicystic Ameloblastoma with their mean age of occurrence

In our case, the age of the patient was 32 years with no history of impacted tooth in the affected region. So it comes under the non-dentigerous variant of UA.

Amongst the jaw bones, mandible is three to thirteen times more affected in contrast to maxilla which also holds true in this case. Also, the posterior mandible is most frequently involved site followed by parasymphysis region, anterior maxillary segment and posterior maxilla [12]. Our case showed similar involvement of mandibular site which is in consensus with the literature.

Radiographically, UA may either be unilocular or multilocular, unilocular being more common in dentigerous variants in contrast to non-dentigerous variant exhibiting multilocularity. In our case, the lesion being non-dentigerous variant of UA showed unilocular radiographic picture.

Variants	**Unilocularity : Multilocularity ratios**
Dentigerous	4.3:1.2
Non-dentigerous	1.1:1

The minimum criterion for the diagnosis as an UA is the demonstration of a single (often macro-cystic sac, with an odontogenic ameloblastomatous) epithelium, which is usually present only in focal areas [14]

Leider proposed three probable mechanisms underlying its pathogenesis. Firstly, it could originate from the reduced enamel epithelium associated with a developing tooth which subsequently undergoes ameloblastic transformation and cyst formation. Secondly, it may also arise from dentigerous or other types of odontogenic cysts in which the neoplastic ameloblastic epithelium is preceded temporarily by a non-neoplastic stratified squamous epithelial lining and thirdly it could result from cystic degeneration of ameloblastic islands of a solid multicystic Ameloblastoma (SMA).

Recent molecular studies in order to determine the pathogenesis of ameloblastomas has revealed overall low cellular proliferation index, association of higher MIB 1 labelling index and microsatellite alterations, telomerase activity associated with p16 loss, and dysregulation of sonic hedgehog/patched (SHH/ PTCH) pathway. However, its clinical significance still is not clear [16].

Ackerman classified UA as –

Luminal-Tumour confined to luminal surface of the cyst.

Intraluminal/Plexiform UA- Nodular proliferation into the lumen without infiltration of tumour cells in the connective tissue wall.

Mural UA- Invasive islands of ameloblastic epithelium in the connective tissue wall not involving the entire epithelium.[7,15]

Philipsen and Reichart modified the Ackerman classification as- **(Table 2)**.

Subgroup	Interpretation
1	Luminal UA
1.2	Luminal and intraluminal UA
1.2.3	Luminal, intraluminal and intramural UA
1.3	Luminal and intramural UA

Table 2- Modified the Ackerman classification

The significance of this modified Ackerman's classification lies while deciding treatment modality.**(Table 3)**.

Subgroup	Treatment modality
1 & 1.2	Careful enucleation followed by chemical cauterization with Carnoy's solution
1.2.3 & 1.3	Aggressive surgical resection

Table 3- Ackerman's subtype and recommended treatment modality

In the present case, the luminal areas of tumour satisfies Vickers and Gorlin criteria and exhibits intraluminal ameloblastomatous proliferation in plexiform pattern. Also, isolated ameloblast lined follicles are seen in the connective tissue stroma thus appropriately justified by subtype 1.2.3. According to study conducted by Lau et al. recurrence rate is highest after enucleation alone while lowest after surgical resection.[11] **(Table 4).**

Surgical modalities	Associated rate of recurrence
Resection	**3.6%**
Enucleation alone	30.5%
Enucleation followed by Carnoy's solution application	16%
Marsupialization	18%

Table 4 - Estimated recurrence rate associated with various treatment modalities

The recurrence rate of UA after curettage alone varies from 18-25%[17,18]. In our case, the lesion was previously treated with curettage only. Since the lesion had recurred within 6 months after conservative management at private clinic, incisional biopsy was not performed, the rationale being small tissue specimen may not demonstrate modified Ackerman's UA subtypes leading to under diagnosis [3].Also, considering the history of recurrence and large extent of the lesion, surgical resection was opted for the patient. Further, serial sectioning of the resected specimen is recommended to rule out the aggressive histologic subtype 1.2.3 and 1.3, so done in our case. Patient is kept for long term

follow-up and is currently disease free 6 months post-operatively.

CONCLUSION:

To report an Ameloblastoma as unicystic, gross clinical examination, good radiographic interpretation and thorough microscopic evaluation is mandatory. UA subtype 1.2.3 and 1.3 warrants aggressive treatment just like the conventional SMA.

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