doi:10.5368/aedj.2011.3.2.3.8

UNI CYSTIC AMELOBLASTOMA: CLINICALLY A MISSED DIAGNOSTIC ENTITY- 3 CASE REPORTS.

[1]Sreedhar Gadiputi [1]*Professor*
[2]Sumalatha.M.N [2]*Senior lecturer*

[1,2] Department of oral pathology and microbiology, Kothiwal Dental College and Research Centre, Kanth Road, Moramustaqueem, Moradabad-244001, Uttar Pradesh, India.

ABSTRACT

Unicystic ameloblastoma a variant of solid or multicystic ameloblastoma mimicking clinically, radiographically, and grossly with jaw cysts, but histopathologically demonstrates a typical ameloblatomatous (odontogenic) epithelial lining with or with out luminal and /or mural growth. Unicystic ameloblastoma tend to occur in younger age group with preferred site of occurrence in the posterior mandible with favorable biological behavior. This report presents 3 cases of unicystic ameloblastoma in different age group with varied clinical and radiographic features which directed to misdiagnose the lesion clinically. But histopathology rooted to confirm the diagnosis.

KEY WORDS:. Intra mural, Intra luminal, Multilocular, Unicystic, Multicystic.

INTRODUCTION

In 1977 Robinson and Martinez identified a subset of ameloblastoma called unicystic ameloblastoma[1]. Prior to 1977 it was termed as Mural, Monocystic, Intracystic, Cystogenic, or Cystic ameloblastoma, and ameloblastoma developing in a radicular cyst[2]. Later Gardner (1980, 81, 83, 84), Leider and Eversole (1984and85) proved that it is different from its conventional or solid counter part by its clinical, radiographic, histopathological, and prognostic profile[3].

It accounts for less than 10-15% of all intraosseous ameloblastomas[4], tends to occur in younger age group (2nd to 3rd decade, mean age of 22years), with male to female ratio **1.3:1** and preferred site of occurrence in posterior mandible associated with or with out impacted tooth (mandible to maxilla= **3to13:1**). UA are usually asymptomatic swellings and are diagnosed during routine radiographic examination. The term UA is derived from the macro and microscopic appearance. The lesion is being well defined often large monocystic cavity with lining focally but rarely entirely composed of odontogenic epithelium[5]. It appears very similar to non-neoplastic odontogenic cysts and frequently clinically misdiagnosed as dentigerous cyst, odontogenic kerato cyst, and solid ameloblastoma and hence histopathologiacl confirmation is mandatory[6]. UA are believed to be less aggressive and possess a much better prognosis after enucleation or curettage than doe's solid or multicystic ameloblastomas[7]. Here we report 3 clinically misdiagnosed, histopathologically confirmed cases of unicystic ameloblastomas.

Case reports

The reported cases in this article was visited with their chief complaint to the department of oral medicine and radiology, kothiwal Dental College and Research Centre, Moradabad, U.P for needful diagnosis and treatment.

Case-1

A 58 year old female reported with a persistent asymptomatic swelling on the lower left side of the face since 11 months with history of extraction past 20 years back i.r.t 37 and38 due to caries **(Fig.1).** Solitary, smooth, diffuse swelling of size 1.5x3cms extending from the body to ramus of the mandible aneroposteriorly, from lower border of the mandible till submandibular region supeorinferiorly **(Fig.-2).** Intra orally swelling was minimal on both the buccal and lingual sides **(Fig.3).** Aspiration from the swelling yielded 2ml of amber colored fluid **(Fig.4)**. With the above findings provisional diagnosis of Odontogenic kerato cyst, Residual cyst, Ameloblastoma was made. Radiographically panoramic view revealed a large well defined unilocular radiolucency measuring approximately 6x4cms extending from 35 to subcondylar region with resorption of roots of 36, lower border and angle of the mandible(**Fig.-5).**Occlusal view showed radiolucency i.r.t 36 with buccal and lingual expansion(**Fig.6).** With the above findings working diagnosis of odontogenic kerato cyst, differential diagnosis of residual cyst, ameloblastoma, was made.

Case Report-1

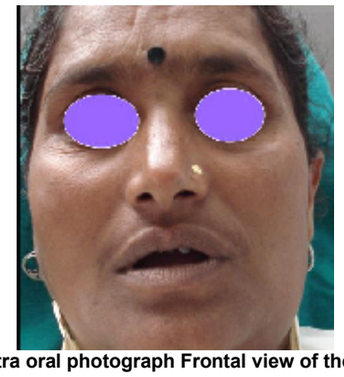

Fig.1. Extra oral photograph Frontal view of the swelling

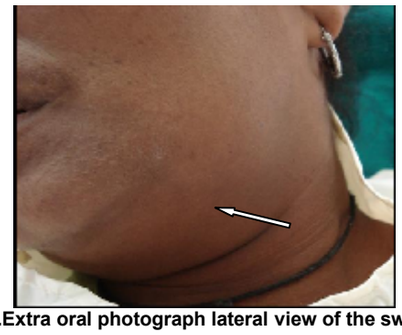

Fig.2 .Extra oral photograph lateral view of the swelling.

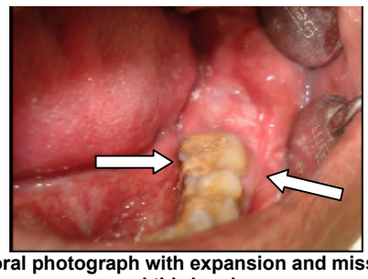

Fig.3 Intra oral photograph with expansion and missing second and third molar.

Fig.4 .Serosanguinous amber colored fluid from the swelling

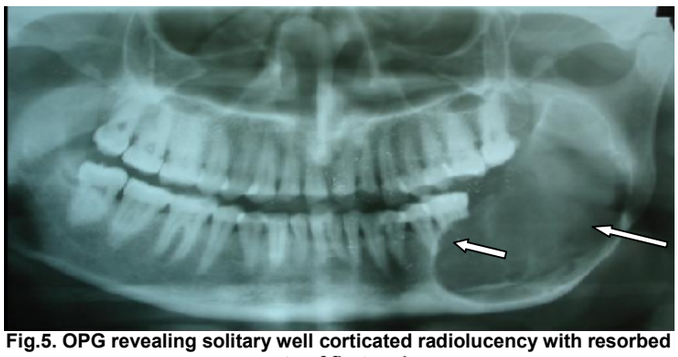

Fig.5. OPG revealing solitary well corticated radiolucency with resorbed roots of first molar

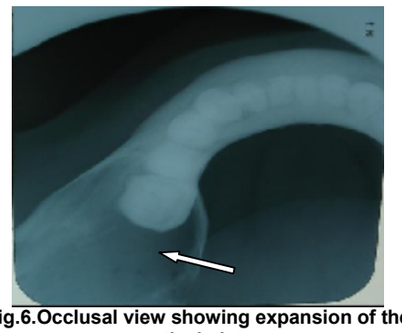

Fig.6.Occlusal view showing expansion of the cortical plates

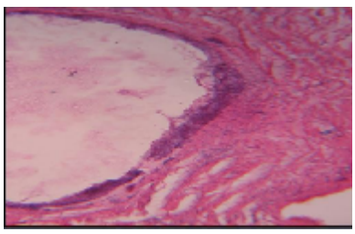

Fig.7. sections showing thin stratified squamous epithelium with fibrocellular connective tissue stroma. (H&E, X4).

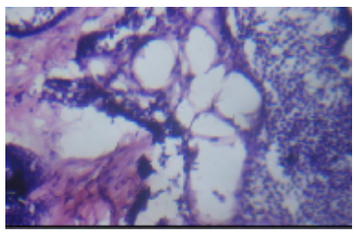

Fig.8.sections showing proliferating odontogenic epithelium in the form of follicle showing areas of cystic degeneration. (H&E, x40)

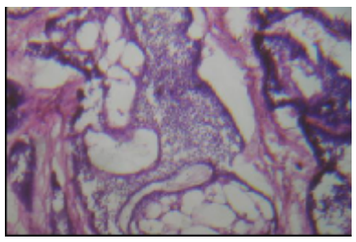

Fig.9.sections showing odontogenic epithelial islands lined by low columnar epithelial cells, polarized hyper chromatic nuclei with central stellate reticulum (H&E, X40).

Cystic fluid assessed for micro protein estimation using semiautomatic analyser was 8.2gm% ruled out the possibility of OKC.

Tissue specimen received from incisional biopsy fixed under 10% neutral buffered formalin processed for routine Haematoxylin and Eosin staining and stained sections revealed the presence of 3-4 cell layers thick epithelium overlying the fibrocellular connective tissue stroma with proliferating epithelial islands **(Fig.-7).** The basal cells of epithelial lining were columnar with hyperchromatic, palisaded, polarized nuclei. The connective tissue stroma revealed proliferating odontogenic epithelial islands arranged in the form of follicles showing cystic degeneration at places, the follicles are lined by columnar epithelial cells having hyperchromatic nuclei with central stellate reticulum cells **(Fig.8 and Fig.9).** Thus the diagnosis simple unicystic ameloblastoma intramural follicular variant (Sub Type1-3) was given.

The histopathology was reconfirmed after surgical resection and the patient is under strict follow up with no signs of recurrence.

Case-2

A 25 year old female reported with complaint of pain and swelling in the lower right back tooth region since 6 months **(Fig.-10).** On examination a Solitary, smooth, diffuse swelling measuring approximately 8x4 cms extending anteroposteriorly from premolar to anterior part of tragus of the ear, superoinferiorly from zygomatic buttress till submandibular region(Fig.-11). Intra orally swelling was marked on lingual aspect with missing third molar **(Fig.-12).** On palpation swelling was painful soft to firm in consistency with expansion on both the sides. Aspiration yielded 3-4ml of straw colored fluid from cystic lesion **(Fig.-13).** With the above findings provisional diagnosis of dentigerous cyst, differential diagnosis of Odontogenic kerato cyst, Ameloblastoma, Gorlins cyst was made.

Panoramic view revealed two well defined radiolucencies larger one measuring approximately 8x3cms extending from distal part of 47 to subcondylar region of the mandible, smaller radiolucency measuring approximately 1x1cms located below the apices of 46, impacted third molar below the roots of 47 and is pushed towards the lower border of mandible**(Fig.14).** Occlusal view showed radiolucency i.r.t 46 and 47 with buccal and lingual cortical plate expansion **(Fig.-15).** With the above findings working diagnosis of dentigerous cyst and differential diagnosis of odontogenic kerarocyst, ameloblastoma, Gorlins cyst was made.

Cystic fluid assessed for micro protein estimation using semiautomatic analyser was in the range of 4.9gm% was in favor of dentigerous cyst.

Tissue specimen received from incisional biopsy fixed under 10% neutral buffered formalin processed for routine Haematoxylin and Eosin staining and stained sections revealed the presence of thin stratified squamous epithelium overlying the fibrocellular connective tissue stroma with proliferating epithelial islands **(Fig.16).** The basal cells of epithelial lining was low columnar with hyperchromatic, palisaded, polarized nuclei. The connective tissue stroma revealed proliferating odontogenic epithelial islands arranged in the form of follicles lined by columnar epithelial cells having hyperchromatic nuclei with central stellate reticulum cells **(Fig.17 and Fig.18).** Thus the diagnosis of simple unicystic ameloblastoma intramural follicular variant (sub type 1-3) was given.

The histopathology was reconfirmed after surgical resection of the mandible and the patient is under strict follow up and free of recurrence has been reported so far.

Case-3

A 16 year old girl reported with persistent painless swelling on the left lower jaw since 3 months **(Fig.-19).** On clinical examination extraorally a Solitary, smooth, diffuse swelling measuring approximately 3x4 cms extending from corner of the mouth till the body of the mandible anteroposteriorly, from Occlusal plane to lower border of the mandible superoinferiorly **(Fig.-20).** Intra orally swelling was extending from 34 to 36 with spacing between 34 and 35 **(Fig.-21).** On palpation swelling was painful, firm in consistency with expansion on buccal side. Aspiration yielded 3ml of straw colored fluid **(Fig.-22)** helped to carve the provisional diagnosis towards Odontogenic kerato cyst, Ameloblastoma and primordial cyst.

Panoramic view revealed well defined solitary radiolucency measuring approximately 6x2cms extending from 31-36 region of the left mandible showing root resorption, separation i.r.t 34 and 35 **(Fig.-23).**Occlusal view revealed well defined solitary radiolucency i.r.t 34 and 36 with buccal and lingual cortical plate expansion **(Fig.-24).** With the above findings working diagnosis of odontogenic kerarocyst and differential diagnosis of ameloblastoma and primordial cyst was made.

Cystic fluid assessed for micro protein estimation using semiautomatic analyser was in the range of 3gm%.

Incisional biopsy specimen fixed under 10% neutral buffered formalin processed for routine Haematoxylin and Eosin staining and stained sections revealed the presence of thin stratified squamous epithelium overlying the fibrocellular connective tissue stroma **(Fig.-25).** The basal cells of epithelial lining was low columnar with hyperchromatic, nuclei, supra basal cells revealed reduced number of intercellular contacts appear to be

Case report-2

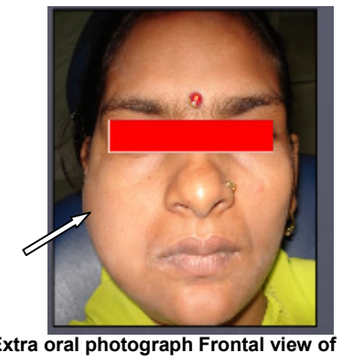

Fig.10. Extra oral photograph Frontal view of the swelling.

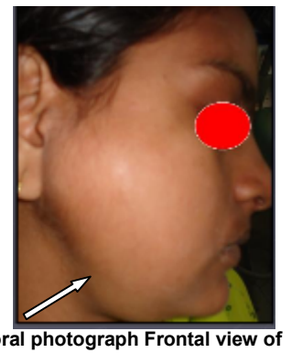

Fig.11. Extra oral photograph Frontal view of the swelling.

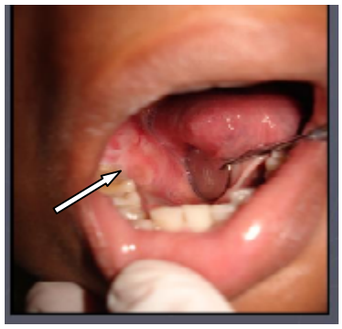

Fig.12. Intra oral photograph with expansion and missing third molar.

Fig.13.Clear straw colored fluid from the swelling.

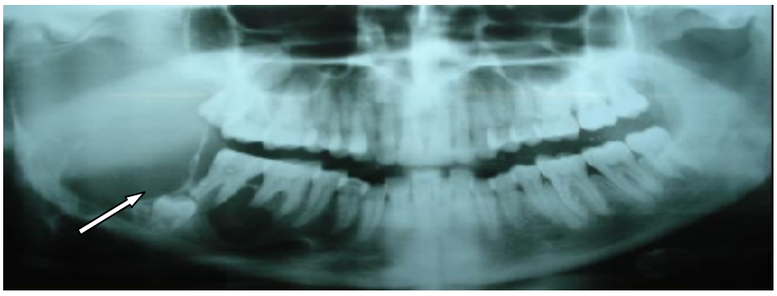

Fig.14 .OPG revealing well corticated large and small radiolucencies with impacted third molar located in the lower border of the mandible.

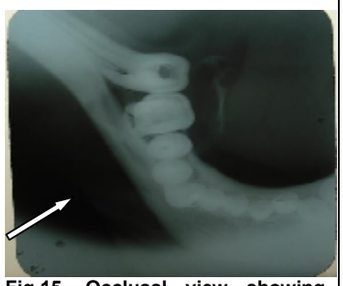

Fig.15. Occlusal view showing expansion of the cortical plates.

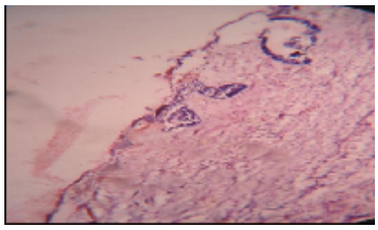

Fig.16 sections showing thin stratified squamous epithelium with proliferating islands in the fibrocellular connective tissue stroma. (H&E, X4).

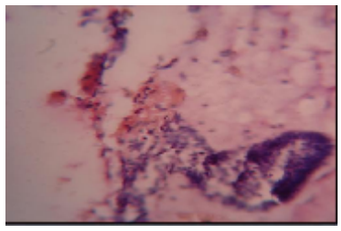

Fig.17. sections showing proliferating odontogenic epithelium in the form of follicle with central degenerated stellate reticulum. (H&E, x40)

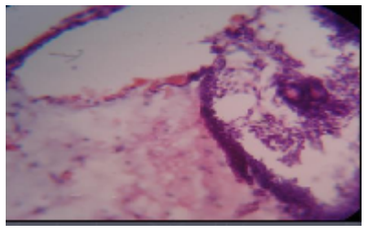

Fig.18. sections showing odontogenic epithelial islands with cystic degeneration. (H&E, X40).

stellate shaped. The fibrocellular connective tissue stroma is loose with odontogenic epithelial islands. Thus the diagnosis of simple unicystic ameloblastoma luminal variant (sub type-1) was given.
The histopathology was reconfirmed after surgical enucleation and the patient is under strict follow up and free of recurrence has been reported so far.

Discussion

Ameloblastomas are enigmatic group of oral tumors, they are true and 2nd most common of all odontogenic tumors. According to WHO Blue book volume "pathology and genetics of tumors of the head and neck'' 2003 they are considered as benign neoplasm's of odontogenic apparatus and divided into four types among which UA is the second type[8].

Ackerman et.al. Reported median age of presentation is 22 years probably attributed to the fact that authors reporting on UAs have focused their studies on children and adolescents. Case no 2 and3 are in favor of above said findings and case-1 is corroborating with El-Adbin andRupercht, Ackerman et.al. Suggesting that UAs are not exclusively tumors of children and adolescents[5].
There are very few reports on sexual and racial predilection for UA but the cases reported here favors the findings of El-Adbin andRupercht, with female predilection male to female= 0.75:1[5].

More than 90% cases of UAs are located in the mandible, with 77% in the molar, ramus region[8].The 3 reported cases are favoring the findings of Khan M.A and Kessler-A by occurring in molar, ramus, body and angle of the mandible. Between 50 and 80% of cases associated with tooth impaction, the mandibular third molar being the most often involved[8].

The impacted tooth associated UAs tend to occur on average of 8 years earlier than lesions independent of impacted teeth. The mean age for unilocular impaction associated UA is 22years[2]. Our case-2 favored above findings with impacted third molar and by occurring at the age 25 years.

Patients with UA often complain of swelling with facial asymmetry. The swelling is asymptomatic, pain is an occasional presenting sign. Painless asymptomatic swelling often indicates a lesion of long duration and significant size[8]. Case 1and3 favors the above findings but case-2 was associated with pain may be due to presence of impacted 3rd molar beneath the apices of 2nd molar.

Radiographically UAs are of two types unilocular and multilocular with predominate unilocular variant. Eversole et.al reported the ratio of unilocular and multilocular=13:3 for cases associated with impaction 8:7 for cases not associated with impaction[2].Unilocular pattern is often misdiagnosed as an odontogenic karato cyst or dentigerous cyst when associated with impaction[9].

All off the reported cases elicited radiographically unilocular radiolucency and case-2 is unilocular with impacted third molar made us to misdiagnose the lesion as other pathology.

The histological features of UA have been established by several authors of whom various subtypes are determined by the pattern and extent of ameloblastomatous proliferation in relation to the cyst wall[9].

The minimum criterion for diagnosing a lesion as UA is the demonstration of a single cystic sac lined by odontogenic epithelium often seen only in focal areas[8].Unicystic ameloblastoma should be differentiated from odontogenic cysts because former has a high rate of recurrence than the later[9].

The 3 reported cases are diagnosed microscopically by using VandG criteria and histological sub grouping of UA modified by H.P.Philpsen and P.A Reichart after Ackerman et.al[2].

Case 1and2 as sub group -1-3 i.e. simple intramural variety, case-3 as sub group-1 simple unicystic ameloblastoma. UA associated with impacted and non impacted tooth showed invasive ameloblastoma tissue in the wall of the cyst (subtype1-2.3, 1-3) with the strongest tendency for this histological pattern to occur in the no impaction category[2].Case-1and3 is in favor of above findings, but case-2 even though associated with impacted tooth showed features of sub type1.3 differed from above findings. Waldron andEl-mofty reported that the follicular pattern was the most prevalent (45%)[10]. our cases 1and2 are corroborated with the above findings.

The unicystic ameloblastoma diagnosed as subgroups 1and1.2 may be treated conservatively (enucleation), where as subgroups 1-2.3 and1-3 must be treated radically i.e. as a solid or multicystic ameloblastoma[2]. Sub groups 1-2.3 and1-3 in which the cystic wall is involved with islands of ameloblastoma tumor cells and there is possible penetration into the surrounding cancellous bone are thought to be associated with a high risk of recurrence requiring more aggressive surgical procedures[8].

In the reported case case1and2 are treated with resection followed by grafting, case-3 treated with simple surgical enucleation.

Average rate of recurrence period is 7 to 10 years with recurrence rate related to histological subtypes with those invading the fibrous wall having 35.7% rate but subtype 1and1-2 6.7%[9].

To assess the recurrence rate in the reported cases regular follow up is done till date but there is no signs of recurrence.

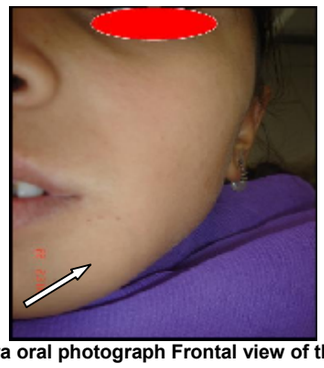

Fig.19. Extra oral photograph Frontal view of the swelling.

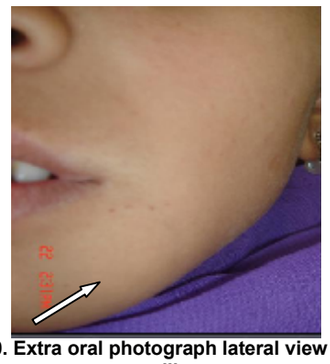

Fig.20. Extra oral photograph lateral view of the swelling.

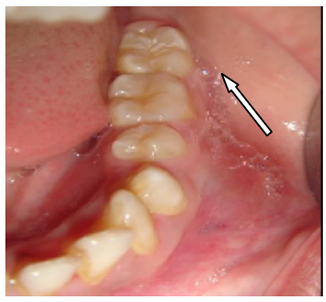

Fig.21. Intra oral photograph with expansion and missing third molar

Fig.22. Clear straw colored fluid from the swelling.

Fig.23. OPG revealing well corticated large and small radiolucencies with impacted third molar located in the lower border of the mandible.

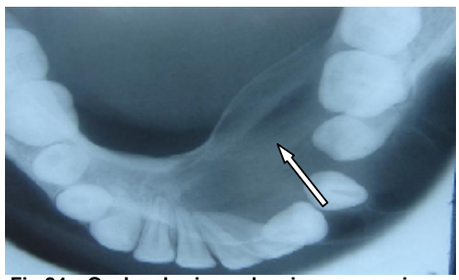

Fig.24. Occlusal view showing expansion of the cortical plates.

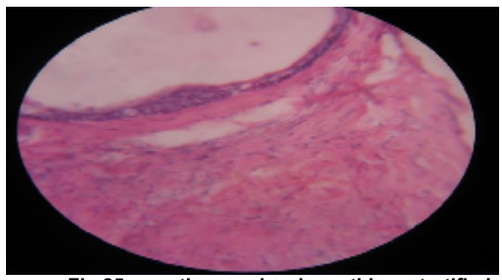

Fig.25 sections showing thin stratified squamous epithelium with proliferating islands in the fibrocellular connective tissue stroma. (H&E, X4).

Despite the fact that unicystic ameloblastoma a variant of solid or multicystic ameloblastoma but mimics various jaw cysts clinically and radiographically paving towards misdiagnosis recommending the microscopic confirmation.

But preoperative incisional biopsy can only be representative fro the entire lesion in extremely few instances and will probably result in misdiagnosis and incorrect classification.

True nature of the lesion may then only become evident when the entire specimen is available for microscopy. Excision or operation specimens should be subject to multiple or preferably serial sectioning to search in for particular for cell and tissue conFig.uration in cyst wall nodules.

Therefore when the diagnosis of unicystic ameloblastoma in younger people remains in doubt after clinical and radiographic examination a biopsy is necessary followed by study of complete enucleated surgical specimen and long term follow up at regular intervals after surgery.

References:

1. Robinson L., Martinez.MG: Unicystic Ameloblastoma: a prognostically distinct entity. Cancer 40:2278; 1977. doi:10.1002/1097-0142(197711)40:5<2278::AID-CNCR2820400539>3.0.CO;2-L

2. Philpsen HP, Reichert PA: Unicystic Ameloblastoma. A review of 193 cases from the literature. Oral Oncol.34: 317.1998.

3. Richard H.Haug, Christopher A., Auer, Brian Smith A. Reviewing the Unicystic ameloblastoma: report of two cases. JADA, 1990, Dec, 703-705.

4. Ackerman GL. Altini M, Shear M. Unicystic ameloblastoma: A clinicopathological study of 57 cases. J Oral Pathol, 1988; 17: 541-516.
doi:10.1111/j.1600-0714.1988.tb01331.x

5. Ademola Abdomen Laitance, Oladepo adekeye; Unicystic ameloblastoma of the mandible: A Long Term Follow –Up, J oral maxillofac surg.55; 345-348, 1997
doi:10.1016/S0278-2391(97)90122-1

6. S.L Salu, N.Samman: Recurrence related to treatment modalities of unicystic ameloblastoma: a systematic review. Int. J Oral Maxilofac Surg. 2006: 35:681-690
doi:10.1016/j.ijom.2006.02.016
PMid:16782308

7. Gardner D G; Some Current Concepts on the pathology of ameloblastoma. Oral surg Oral med Oral pathol, 1996; 82: No-6: 660-669.

8. Simi Thankappan, Valsa Thomas, Sharafudeen kandamparabil, sherin Nair: Unicystic ameloblastoma: 3 case reports and review of literature: 2008/ april-june/volume-20/issue2. 65-70.

9. Philpsen HP, Reichert PA. Classification of odontogenic tumors and allied lesions. In: Odontogenic tumors and Allied lesions. Quintessence Pub. Co Ltd; 2004, P309-322.

10. Waldron CA, El-Mofty Sk. A histopathologic study of 116 ameloblastomas with special reference to the desmoplastic variant. Oral surg Oral med Oral pathol 1987; 63:441-451.
doi:10.1016/0030-4220(87)90257-X

Corresponding Author

Dr. Gadiputi Sreedhar MDS
Professor
Department of oral pathology and microbiology
Kothiwal Dental College and Research centre
Kanth Road, Moramustaqueem,
Moradabad-244001
Uttar Pradesh
Mobile no- +91-9927634333
+91-9760877208
Email: drgadiputi@yahoo.com