



UNICYSTIC AMELOBLASTOMA- A CASE REPORT

¹Dr. S.Fatima Khan, ²Dr. Deeplaxmi, ³Dr. Ekta Sawriya, ⁴Dr. N Priyadarshee, ⁵Dr. Ankit Sharma

¹Professor and HOD ²Professor ³Senior lecturer ⁴Senior lecturer ⁵PG Student

¹Department of Oral Medicine, Diagnosis and Radiology

¹Rungta College of Dental Sciences and Research, Bhilai, Chhattisgarh

ABSTRACT:

Background: Unicystic Ameloblastoma is a variant of ameloblastoma characterized by its cystic presentation, primarily affecting younger patients. This report presents a case of a 20-year-old female diagnosed with unicystic ameloblastoma. **Case Presentation:** The patient presented with a swelling in the right mandibular posterior region, which had gradually increased in size in a span of 9 months. Radiographical examination revealed a well-defined radiolucency.. Histopathological analysis confirmed the diagnosis of unicystic ameloblastoma, showing a cystic cavity lined by a stratified squamous epithelium with focal areas of ameloblast-like cells. **Discussion:** Unicystic ameloblastoma is often associated with a favorable prognosis compared to its solid counterpart. The treatment involved segmental resection of the mandible with load-bearing reconstruction plates. This case highlights the importance of early diagnosis and appropriate management in young patients to minimize complications. **Conclusion:** Unicystic ameloblastoma should be considered in differential diagnoses for cystic jaw lesions in young adults. A thorough clinical and histological evaluation is crucial for effective management and positive outcomes. Further studies are needed to better understand the biological behavior and recurrence rates of this variant.

Keywords: Unicystic ameloblastoma, cystic jaw lesions, diagnosis, radiographical examination, histopathology, treatment

INTRODUCTION:

Ameloblastomas are benign tumors whose importance lies in its potential to grow into enormous size with resulting bone deformity. They are typically classified as Unicystic, multicystic, peripheral and malignant subtypes. ^[1]

Unicystic Ameloblastoma (UA) is a less encountered variant of the Ameloblastoma, referring to those cystic lesions that show clinical and radiographic characteristics of an odontogenic cyst but in histological examination shows a typical ameloblastomatous epithelium lining part of the cyst cavity, with or without luminal and mural tumor proliferation. ^[2]

Ameloblastoma is a slow-growing locally aggressive, benign odontogenic epithelium with a greater tendency for recurrence and involves aggressive bone loss and exfoliation of teeth. It is believed to be derived from cell rests of the enamel organ, either remnants of dental lamina or Hertwig's sheath, the epithelial rest of Malassez. ^[3-5]

In this case report, a large Unicystic Ameloblastoma in a 20-year-old female patient with typical radiographic and histopathologic features followed by management of the same diagnosis is discussed.

CASE REPORT:

A 20-year-old female reported to the Department of Oral Medicine and Radiology at Rungta College of Dental Science and Research, Bhilai, Chhattisgarh, India, with a chief complaint of slowly growing swelling over the right side of face since 9 months with associated pain while swallowing and difficulty in mastication. Pain and swelling did not subside on taking medication. Patient medical and family histories were essentially unremarkable. On General Physical Examination, no abnormality was detected, except for asymmetrical face due to swelling.

ON EXTRAORAL EXAMINATION



(Fig 1) showing swelling on the lower right posterior region of the face

A solitary diffuse swelling was seen on the right side of the face (Fig 1) involving the middle and lower 1/3rd extending: Superior-inferiorly from Ala-tragus line till approximately 3cm below the inferior border of the mandible and Anterior-posteriorly Para symphysis till the angle of the mandible measuring approximately 6x6 cm in its greatest dimension with ill-defined edges and stretched overlying skin.

On Palpation: All the inspectory findings confirmed, swelling was hard, non-reducible, non-fluctuant & not compressible with no local rise in temperature.

ON INTRA ORAL EXAMINATION: A solitary diffuse swelling present on the Mandibular right posterior region extending to the buccal vestibule from the distal aspect of 44 to distal aspect of 48 measuring approximately 3.5x2 cm in its longest dimension with normal surrounding mucosa. Occlusal pit caries with respect to 46,47 (Fig 2)



(Fig 2) showing intraoral swelling on the lower right posterior region of the jaw w.r.t. 46,47

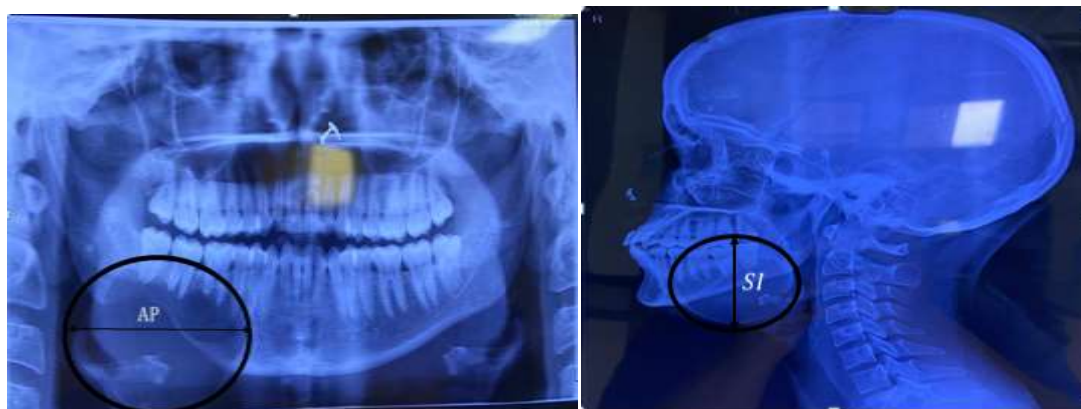
On Palpation: All the inspectory findings confirmed. Swelling was hard in consistency, fixed & non tender with no secondary changes.

On the basis of clinical examination and history, a provisional diagnosis of Ameloblastoma involving right mandibular posterior region was given. Under differential diagnosis, Dentigerous Cyst, Odontogenic Keratocyst, Aneurysmal Bone Cyst and Giant Cell Granuloma were considered. Patient was advised for radiological investigation and biopsy.

RADIOGRAPHIC INVESTIGATIONS:

Well-defined oval radiolucency is seen involving the right mandibular posterior region extending anteroposterior from 44 beyond the angle of the mandible & Superiorinferiorly from the crest of the alveolar bone till 2 cm inferior to the lower border of the mandible.(Fig 3) Resorption of the cortical plate leaves a thin 'eggshell' of bone. Root resorption with 46,47 and based on OPG findings radiological diagnosis :-

Unicystic Ameloblastoma was made with Differential diagnosis Odontogenic Keratocyst, Aneurysmal Bone Cyst and Giant Cell Tumor.



(a)

(b)

(Fig 3) showing Well-defined oval radiolucency in the right mandibular posterior region both in (a) Anteroposterior view in a OPG and (b) Supero-inferiorly in a lateral cephalogram

MANAGEMENT:

Preoperative diagnosis of the lesion was made as Unicystic Ameloblastoma based on the clinical & radiological findings.

The tumor was surgically managed in Department of Oral And Maxillofacial Surgery with segmental resection of the mandible with load-bearing reconstruction plates. The entire specimen was then submitted for the histopathologic examination.



(a) (b)

(c)



(d)

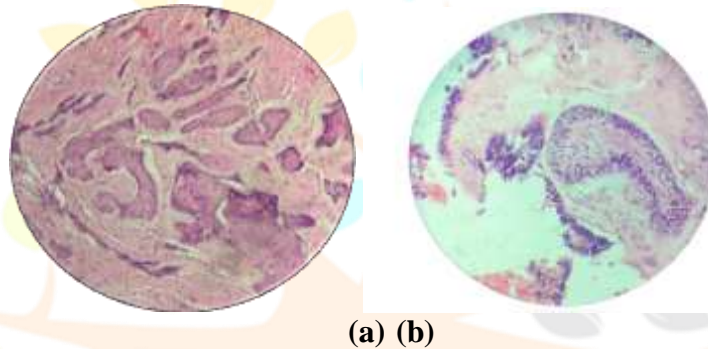
(e)

Fig 4: (a) Removal of the flap (b) Tumor exposed (c) Specimen (d) Surgical defect (e) Fixation

Post Surgery:

Fig 5: showing post operative image

The specimen was fixed and processed. Histopathological examination revealed follicles lined by Ameloblast-like cells in the periphery and centrally by stellate reticulum-like cells. Extravasated RBCs were also seen in a few areas. (Fig 4)



(a) (b)

Fig 6: shows (a) Histological image showing fibro cellular connective tissue stroma with multiple odontogenic islands (b) Follicles are lined by ameloblast like cells in the periphery and centrally by stellate reticulum like cells. Extravasated RBCs are also seen in few areas

DISCUSSION:

Unicystic Ameloblastoma is a unique variant of Ameloblastoma first identified by Robinson and Martinez in 1977. This condition pertains to cystic lesions that exhibit clinical and radiographic features resembling an Odontogenic cyst, yet are characterized histologically by an epithelial lining with typical ameloblastomatous features, which may include luminal and/or mural tumor proliferation.^[6]

The diagnosis of Ameloblastoma requires imaging as well as a biopsy. Although Ameloblastoma originating within bone is often detected on dental X-rays (OPG) or plain film.^[7,8] Radiographically, it may appear unilocular or multilocular with soap bubble or honeycomb appearance; buccal and lingual expansion of the cortex invariably accompanies Ameloblastoma. Thinned and intact cortex shows egg shell appearance.^[9]

In present case, OPG showed well defined oval radiolucency and resorption of the cortical plates along with root resorption.

Routine panoramic radiography is essential for diagnostic investigations in pathology screening. However, it cannot definitively exclude other conditions with radiological features that may resemble those of Ameloblastomas.^[10] These include conditions such as Odontogenic Keratocyst, Giant Cell Tumor, Fibromyxoma, Fibrosarcoma, Aneurysmal Bone Cyst. Additional limitations involve insufficient visualization of the tumor's bony margins and an unclear interface between the tumor and surrounding soft tissue. Moreover, Ameloblastomas often have a tendency to perforate the cortex, which is a crucial factor for differential diagnosis. Unfortunately, this characteristic may not always be detectable through conventional radiography.^[11]

CT scans typically reveal a well-defined, uni- or multilocular radiolucent expansile lesion and are effective for assessing cortical destruction and soft tissue extension.^[7] CECT scan shows an enhancement effect in the solid components.^[12] In Ameloblastoma, there has been a significant correlation between microvessel density (which depicts the vascularization of tumour tissue) and contrast enhancement characteristics in CT scans.^[13] As a result, contrast enhancement is one of the most essential characteristics of benign Odontogenic tumours.

However, the extent of soft tissue or bone invasion is often not accurately assessed. On radiographs, Unicystic Ameloblastoma typically appears as a lytic lesion with scalloped margins, often accompanied by impacted molars and root resorption. The more common multilocular Ameloblastoma is characterized by the classic "soap bubble" appearance on Radiographs. For Ameloblastomas originating in the maxilla, MRI is preferable to CT, as it provides better delineation of any extensions to the skull base, orbit, or paranasal sinuses. Additionally, MRI is the preferred imaging modality for Desmoplastic Ameloblastoma due to its ill-defined soft tissue borders, which can be mistaken for a fibro-osseous lesion.^[7,8]

Unicystic Ameloblastoma is typically diagnosed only after a histopathological examination, as it closely resembles an Odontogenic cyst both clinically and radiologically.^[12]

Ackermann et al.^[10] classified Unicystic Ameloblastoma into three types with prognostic and therapeutic implications such as:

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 (invasive islands of ameloblastomatous epithelium in the connective tissue wall not involving the entire epithelium)

CBCT, CT and MRI are much more efficient in defining differential diagnoses.^[12] Another disadvantage of conventional radiography is the inability to assess the internal contents of the lesion. Contrast-enhanced CT and MRI are modalities, which are helpful in this regard.^[14] Although advanced imagery is required in most cases, it is not feasible due to the lack of availability and the high cost.

In the case of Metastasizing Ameloblastoma, a (PET) scan is generally preferred to detect distant metastasis. Management of Ameloblastoma can be done in three ways (1) Conservative which includes Enucleation and Curettage, as well as the use of adjuvant therapies such as Carnoy's solution and Cryotherapy, (2) Marsupialization, and (3) Radical surgery, which includes marginal or block resection (1-1.5 cm margins result in the highest chance of local control) and immediate bone reconstruction. Iliac crest grafts or microvascular fibular flaps may be required for facial reconstruction procedures.^[14]

Recurrence rates for Unicystic Ameloblastoma after conservative surgical treatment (curettage or enucleation) are generally reported to be <25%. For intraluminal and plexiform type of Unicystic Ameloblastoma, recurrence rate was found to be as low as 10.7%. Recurrence rates for solid Multicystic Ameloblastoma was found to be about 50%–90%.^[15]

CONCLUSION:

In conclusion, this case report highlights the significance of diagnosis and appropriate management of Unicystic Ameloblastoma. Despite its often less aggressive nature compared to other forms of Ameloblastoma, careful monitoring and surgical intervention are crucial to prevent potential recurrence. The radiological findings underscore the importance of considering Unicystic Ameloblastoma in differential diagnosis for Odontogenic cysts and tumors, as timely and accurate identification can lead to better patient outcomes. Ongoing follow-up is essential to ensure long-term success and detect any signs of recurrence.

REFERENCES:

1. Burket's Textbook of Oral Medicine, Greenburg & Glickman; 12th edition
2. Differential diagnosis of Oral lesions; Wood & Goaz, 11th edition.
3. Textbook of Oral Medicine & Radiology, Anil Ghom; 3rd edition.
4. Textbook of oral radiology, White & Pharoah; 1st edition.
5. Textbook of oral radiology Freny Karjodkar; 1st edition.
6. Dolanmaz D, Etoz OA, Pampu A, Kalayci A, Gunhan O. Marsupialization of unicystic ameloblastoma: a conservative approach for aggressive odontogenic tumors. *Indian J Dent*

Res. 2011 Sep-Oct;22(5):709–12. doi: 10.4103/0970-9290.93461. [[PubMed](#)] [[CrossRef](#)] [[Google Scholar](#)]

7. . Ameloblastoma: a clinical review and trends in management. McClary AC, West RB, McClary AC, et al. *Eur Arch Otorhinolaryngol*. 2016;273:1649–1661. [[PubMed](#)] [[Google Scholar](#)]
8. Diagnostic value of MRI for odontogenic tumours. Fujita M, Matsuzaki H, Yanagi Y, et al. *Dentomaxillofac Radiol*. 2013;42:20120265. [[PMC free article](#)] [[PubMed](#)] [[Google Scholar](#)]
9. Eversole LR, Leider AS, Strub D. Radiographic characteristics of cystogenic ameloblastoma. *Oral Surg Oral Med Oral Pathol*. 1984;57(5):572-7.
10. Ackermann GL, Altini M, Shear M. The unicystic ameloblastoma: A clinicopathological study of 57 cases. *J Oral Pathol* 1988;17:541-6.
11. Kitisubkanchana J, Reduwan NH, Poomsawat S, PornprasertsukDamrongsri S, Wongchuensoontorn C. Odontogenic keratocyst and ameloblastoma: radiographic evaluation [published online ahead of print, 2020 Feb 6]. *Oral Radiol*. 2020; 10.1007/s11282-020-00425-2.
12. More C, Tailor M, Patel HJ, Asrani M, Thakkar K, Adalja C. Radiographic analysis of ameloblastoma: a retrospective study. *Indian J Dent Res*. 2012 Sep-Oct;23(5):698. doi: 10.4103/0970-9290.107436.
13. Hayashi K, Tozaki M, Sugisaki M, Yoshida N, Fukuda K, Tanabe H. Dynamic multislice helical CT of ameloblastoma and odontogenic keratocyst: correlation between contrast enhancement and angiogenesis. *J Comput Assist Tomogr*. 2002 Nov-Dec;26(6):922–6. doi: 10.1097/00004728-200211000-00011.
14. Hertog D, van der Waal I. Ameloblastoma of the jaws: a critical reappraisal based on a 40-years single institution experience. *Oral Oncol*. 2010 Jan;46(1):61–4. doi: 10.1016/j.oraloncology.2009.11.002
15. Gardner DG, Corio RL. Plexiform unicystic ameloblastoma. A variant of ameloblastoma with a low-recurrence rate after enucleation. *Cancer* 1984;53:1730-5)(Archana Pokala1 , Avinash Tejasvi M.L.1 , Geetha Paramkusam2 , Revath Vyas3 , Harsha Bhayya4 , Pavani Donempudi5 *Indian Journal of Medical and Paediatric Oncology* | Volume 41 | Issue 3 | May-June 2020)

