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Case Report:

Luminal unicystic ameloblastoma of maxilla in 53-year-old female patient - a case analysis

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Abstract

The term unicystic ameloblastoma (UA) refers to those cystic lesions that show clinical, radiographic, or gross features of a jaw cyst, but on histologic examination show a typical ameloblastomatous epithelium lining part of the cyst cavity, with or without luminal and/or mural tumor growth. Here, we present a case report on unicystic variant of ameloblastoma (Luminal Type) in the maxilla. An attempt has been made to emphasize that it can involve the maxillary jaw, which is rarely affected. A literature review on the topic has been added along with the case analysis.

Keywords: Maxilla, Unicystic Ameloblastoma

INTRODUCTION:

Ameloblastoma as described by Robinson, is a benign tumor that is “usually unicentric, non-functional, intermittent in growth, anatomically benign and clinically persistent.” Ameloblastoma is a local invasive tumor which originates from remnants of the dental lamina and odontogenic epithelium and it accounts for only 1 % of all oral tumors.¹ Radiographically Ameloblastoma appears either multilocular and unilocular. In unilocular ameloblastoma single large locule with smooth or scalloped periphery. The cortical plates may be thin, expanded and may be perforated if the lesion is in advanced stage. In multilocular ameloblastoma Honeycomb or soup bubble appearance is seen with thinning & expansion of cortex with perforation if lesion is in advanced stage, scalloped periphery with knife edge root resorption.²

Based on the World Health Organization (WHO) classification of head and neck tumours, there are four forms of ameloblastomas: multicystic, peripheral, desmoplastic and unicystic ameloblastomas.² Unicystic ameloblastoma, a variant of ameloblastoma, was first described by Robinson and Martinez in 1977. UA is a rare subtype of ameloblastoma, accounting for about 6% of intra osseous ameloblastoma. Its manifestation involves younger age group, with almost 50% of the cases in the second decade of life and more than 90% are located in the mandible.⁸ No sexual or racial predilection for UA. The involvement of mandible to maxilla is in a ratio of 13:1.¹

The pathogenesis of cystic ameloblastoma remains obscure. There have been many debates regarding whether unicystic ameloblastoma develops de novo or arises in an existing cyst. Leider et al proposed three pathogenic mechanisms for the evaluation of unicystic ameloblastoma <1> reduced enamel epithelium, <2> from dentigerous cyst and <3> due to cystic degeneration of solid ameloblastoma. The neoplasm originates within the mandible or maxilla from epithelial that is involved in the formation of the teeth. Potential epithelial sources include enamel organ, odontogenic rests (cell of malassez, cell rest of Serre) reduced enamel epithelium and lining of epithelial cyst especially dentigerous cyst.³ Between 50 and 80% of cases are associated with tooth impaction, the mandibular third molar being most often involved. Patients most commonly present with swelling and facial asymmetry, pain being an occasional presenting symptom. Mucosal ulceration is rare, but may be caused by continued growth of the tumor. Small lesions are sometimes discovered more on routine radiographic screening examinations or as a result of local effects (like tooth mobility, occlusal alterations and failure of eruption of teeth) produced by the tumor⁴

Radiographically Unicystic ameloblastoma appears as unilocular lesion with well defined sclerotic borders with impacted tooth with thinning and expansion but can be seen also in interradicular, periapical and edentulous region as well.²Clinically Unicystic ameloblastoma is usually found in association with the crowns of mandibular 3rd molar teeth, but can be seen in maxillary region. It is presented as a painless swelling, facial asymmetry, tooth impaction, tooth displacement, mobility or tooth resorption.^{2,3}

UA has less aggressive biologic behavior and lower recurrence rate than the classic solid or multicystic ameloblastoma. Although the unicystic ameloblastoma is a “cystic” appearing lesion on gross examination, microscopic examination shows the presence of ameloblastoma within the cyst wall.⁵ It is histopathologically divided into intracystic, luminal or intraluminal unicystic ameloblastoma. Also, tumours associated with an unerupted tooth are considered to be the dentigerous variant, whereas those lacking an association with an unerupted tooth were considered to be the nondentigerous variant. The dentigerous variety is known to occur in younger patients⁶

Here we report a rare case of unicystic ameloblastoma (Luminal Type) of in 53-year-old female in maxillary left canine region which is again rare site of occurrence.

Case Report

A 53-year-old female patient visited to Oral Medicine and Radiology department with chief complain of swelling on middle 3rd of left side of face since 6 years which gradually increases to attain the present size. Patient has not consulted any doctor for the same as it was painless and smaller in size before 1-month surgical marsupialisation had been done & clinically artificial sinus tract was visible in Maxillary left canine region. Past H/O Myocardial infarction before 5 years and patient is taking medication for the same. H/O Masala chewing over a longer period of time 40years 3 to 4 times per day was very significant.

On extra oral examination facial asymmetry was appreciated on left middle 3rd of face. the size of the swelling was 2 x 2cm which extended diffusely from zygomatic buttress laterally to ala of nose medially causing slight distortion of face with obliteration of nasolabial fold and infra orbital margin superiorly to angle of mouth inferiorly. The overlying skin had normal colour and temperature, on palpation it was bony hard, Non-compressible, Non-Reducible and Nontender. Sub mandibular lymph nodes on left side were enlarged, measuring 1x1 cm in diameter, tender and mobile in antero-posterior direction (Figure1)

On intra oral examination during inspection, missing canine with a solitary swelling was present in left maxillary antero- posterior region size approx. 3x 3 cm extending antero-posteriorly from distal 11 to distal of 25 and supero-inferiorly 2cm from vestibule with obliteration of vestibular depth with smooth overlying surface and Artificial sinus tract present between 22 and 24. Palatally ill-defined swelling size approx. 2 x 2 cm present which extends antero-posteriorly from mesial of 24 to distal of 26 and medio-laterally from midpalatine raphe to attached gingiva of 24 to 26. Generalised Gingival recession was present. Distal displacement of 22 and mesial displacement of 24 were present. On Intra oral palpation number, site, size margins and extensions were confirmed. Buccal expansion was present. Swelling was soft to firm in consistency, tender, compressible in anterior aspect reducible with clear fluid discharge from artificial sinus tract present between 22 and 24. Grade II mobility of teeth was noted i.r.t 24 25. Associated with this verticle fibrous bands were palpable on right and left buccal mucosa which extends postero-anteriorly from pterygomandibular raphe to commissure of mouth and supero-inferiorly from upper to lower buccal sulcus without any ulceration and erosions (Figure 2) History and above clinical findings, were suggestive of Infected dentigerous cyst of left maxillary canine with oral submucous fibrosis. Differential diagnosis included Infected Lateral periodontal cyst, Infected adenomatoid odontogenic tumor, Infected Dentigerous cyst converted to ameloblastoma.

Intra oral periapical radiograph and Orthopantomograph revealed a single well-defined radiolucent lesion of size approximately 3 x 3cm in relation to 21 to 26 tooth regions with well-defined margins anteriorly, posteriorly and inferiorly along with destruction of alveolar crest superiorly with impacted 23 and calcification foci. Distal displacement of 22 and mesial displacement of 24,25 with root resorption of 24. (Figure 5,7)

A cross Sectional Maxillary Occlusal Radiograph (Figure 6) revealed a single well-defined radiolucent lesion of size approximately 3 x 3cm present on Maxillary left Antero-Posterior teeth region which extends Antero-Posteriorly from 21 to 26 and medio-laterally from Apical 3rd of 11 to 3cm lateral to it with impacted 23 and calcification foci with scalloped periphery. With distal displacement of 22 and mesial displacement of 24. It was followed by computed tomography (Figure 8) that revealed thin walled expansile cystic lesion extending posterior into anterior part of hard palate and projecting superiorly into left maxillary sinus, unerupted tooth seen within it with calcification. Conventional & Computed Tomography report were not conclusive & Radiographic Differential Diagnosis included Adenomatoid odontogenic tumor, Dentigerous cyst with calcification, Dentigerous cyst converted to Ameloblastoma, calcified epithelial odontogenic tumor, and odontogenic keratocyst lesion of left maxillary Canine region.

Then after patient was undergone for surgical enucleation and histopathological evaluation was carried out histopathology examination showed 10x View of H & E Stained section shows 3-4 cell layer thick cystic lining with palisaded basal layer and underlying connective tissue is fibrous and having moderate inflammatory cell inflammation & there is presence of dilated capillaries suggestive of infected unicystic ameloblastoma (Luminal Type) (Figure 10)

The final diagnosis of Infected Unicystic Ameloblastoma (Luminal Type) in Left maxillary antero- Posterior region was thus made. Patient was recalled periodically to check for any recurrence.

Discussion

The presented case of Unicystic Ameloblastoma (Luminal Type) was in 53-year-old female patient & it was a very rare to see at this age group. Usually Unicystic ameloblastoma occurs in younger age group 50% occurrence in in 2nd decade of life. In reported case the lesion was associated with impacted maxillary left canine &

presenting as painless swelling, facial asymmetry with displacement of 22 & 24 with mobility of 24. As such 90% of cases of unicystic ameloblastoma are observed in mandibular in mandibular ramus region so it is rare to see unicystic ameloblastoma in maxilla as in our case.¹

The radiographic appearance is important in the diagnosis; which determines whether the lesion is unilocular, a necessary criterion for unicystic ameloblastoma. Lesions are usually demarcated and may even corticated.⁹ Eversole et al identified predominant radiographical patterns for UCA: unilocular, scalloped, pericoronal, interradicular or periapical expansile radiolucency⁴. Our case had Unilocular with well-defined scalloped expansile radiolucency. In Computed tomography unilocular hypodensity with scalloping and thinning and expansion of cortex with tooth impaction was seen which was similar to literature.^{10,11}

In differential Diagnosis infected lateral periodontal cyst was considered due to clinical finding of distal displacement of 22 and mesial displacement of 24 with buccal expansion with artificial pus discharging sinus tract. Infected dentigerous cyst or dentigerous cyst converted to ameloblastoma was also suspected as there was missing canine and displacement of teeth and expansion of buccal cortex and foci of calcification within radiolucency. Adenomatoid odontogenic tumor and Calcified epithelial odontogenic cyst were also considered in differential diagnosis as due to most common site and Impacted canine with calcification with thinning, expansion of buccal cortex with displacement of teeth.

There are different classifications of unicystic ameloblastoma. Based on the clinicopathologic study of 57 cases of unicystic ameloblastoma, Ackerman's classification into three histologic groups is as follows:

- I. Luminal UA (tumor confined to the luminal surface of the cyst);
- II. Intraluminal/plexiform UA (nodular proliferation into lumen without infiltration of tumor cells into connective tissue wall); and
- III. Mural UA (invasive islands of ameloblastomatous epithelium in the connective tissue wall not involving the entire epithelium).

According to this classification, our case study belongs to Group I e.g luminal variety and as per literature it is very rare to occur.²

UA is considered to be a less aggressive form of ameloblastomas that can be successfully removed by simple enucleation or other less aggressive surgery.²

In our case Surgical Enucleation was done.

Literature suggested that even unicystic ameloblastoma are associated with 10% recurrence and hence require a long term follow up.⁴ In our case Patient was recalled periodically to check for any recurrence.

Conclusion

The diagnosis of unicystic ameloblastoma (Luminal Type) is based on clinical, radiographical and histopathological features. In the present case, only microscopic examination of the surgical specimen allowed the establishment of the final diagnosis of unicystic ameloblastoma, illustrating the complexity of the diagnosis process of bone pathologies, especially when the lesions present on non-classical aspects and atypical locations. Long-term follow up is mandatory because of the recurrence risk of unicystic ameloblastoma, which may occur after a long time.



Figure 1. Diffused swelling on left middle 3rd of face



Figure 2. Well defined swelling irt 11 to 25 with obliteration of depth of vestibule and buccal expansion with pus discharging sinus tract



Figure 3 & 4. Reduced mouth opening with blanching of rt and lt buccal mucosa with palpable fibrous bands



Figure 5. IOPA of 11 to 26 region shows a single well-defined radiolucency size approx. 4×6cm present irt 21 to mesial aspect of 25 with Impacted 23 and calcification within radiolucency and root resorption and mesial displacement irt24. thinning and superior displacement of floor of maxi sinus



Figure 6. Maxillary cross-sectional occlusal radiograph shows a single well-defined radiolucency size approx. 4×5 cm present irt 11 to mesial aspect of 25 with impacted 23, with scalloping of border and calcification within radiolucency and root resorption and mesial displacement irt24 and distal displacement of 22



Figure 7. OPG shows a single well-defined radiolucency size approx. 4×6cm present irt 21 to mesial aspect of 25 and superior- inferiorly from alveolus to 3cm superior to it with impacted 23 and calcification within radiolucency and root resorption and mesial displacement irt24. Superior displacement of floor of maxi sinus with thinning.



Figure 8. CT shows a single well defined hypodense are size approx. 5×4cm present irt maxillary left antero-post teeth region within it impacted canine seen. With thinning and expansion with superior displacement of maxillary sinus floor and calcification is seen within lesion.



Figure 9. Enucleated Specimen

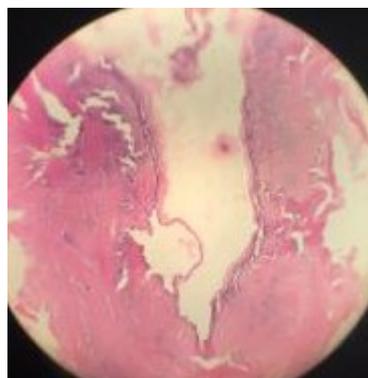


Figure 10. 10x View of H & E Stained section shows 3-4 cell layer thick cystic lining with palisaded basal layer and underlying connective tissue is fibrous and having moderate inflammatory cell inflammation & there is presence of dilated capillaries.



Figure 11. After Surgical enucleation



Figure 12. Follow up after 6 months Complete healing of surgical site



Figure 13. Follow up after 6 months opg shows No any recurrence of ameloblastoma

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