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RESEARCH ARTICLE

UNICYSTIC AMELOBLASTOMA: A DIAGNOSTIC DILEMMA

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ABSTRACT

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 the cyst cavity with or without luminal and/or mural tumour proliferation. (Nagalaxmi *et al*., 2013) A number of morphological variants of ameloblastoma have been documented in the literature and at times, may pose a diagnostic challenge to the pathologist. (Shaikhi *et al*., 2012) The dilemma in the diagnosis of UA exists on a radiograph, when it is associated with impacted third molar where it exclusively shows similarity to the dentigerous cyst. Thus, the diagnosis of UA becomes evident only when the entire specimen is evaluated histopathologically. (Laxmidevi *et al*., 2015) Till date, lot of controversies exist among oral surgeons and oral pathologists regarding this entity. An attempt is being made here to discuss all the diagnostic dilemmas associated with UA. (Arora, 2015) Here, we report one case of luminal, intramural variant of UA in a young male patient.

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INTRODUCTION

Many benign lesions cause mandibular swellings, such as ameloblastoma, radicular cyst, dentigerous cyst, keratocystic odontogenic tumour, central giant cell granuloma, fibro osseous lesions and osteoma. They can be divided into lesions of odontogenic or nonodontogenic origin (Pallagatti *et al*., 2013). Ameloblastoma is one of the most common benign odontogenic tumour accounting for approximately 1% of all tumours and 19% of all odontogenic tumours (Arora *et al*., 2015). Ameloblastoma which develops from epithelial cellular elements and dental tissues in their various phases of development. More than 80% of all ameloblastomas are solid or multicystic variants, with unicystic ameloblastoma being an important clinicopathologic form of ameloblastoma and occupying the remaining 20% of the cases along with peripheral ameloblastoma (Arora *et al*., 2015). UA, a variant of ameloblastoma, was first described by Ackermann *et al*. in 1988. (Nagalaxmi *et al*., 2013) UA represents 5 - 15% of all reported cases (Arora *et al*., 2015). Unlike the solid (muticystic) variant of ameloblastoma, UA differs in its presentation showing more similarity with dentigerous and other cyst.

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Case Report

A 16 year old male patient presented with a chief complaint of swelling over lower left back tooth region since 2 months. The small bulge on lower left posterior region of mandible gradually increased to present size over the period of 2 months. His medical, past dental and family history were non significant. Physical examination revealed no abnormality other than those related to chief complaint. Extra oral examination revealed slight facial asymmetry due to diffuse swelling of the left side of face mainly localized to lower third. The skin over swelling was smooth with no surface changes. Lymph nodes were non palpable and non tender. On palpation, swelling was bony hard, non tender with no localized rise in temperature. Intra orally, a swelling was about 2.5 cm × 2 cm in size, oval in shape extending from distal of 35 to the retromolar trigone posteriorly was observed. Obliteration of buccal vestibule was noted with the expansion of both buccal and lingual cortex. The swelling was tender on palpation, firm in consistency. 36 was missing and 37 was seen erupting. (Figure 1) Considering the history, nature of lesion, location and age group of patient, a provisional diagnosis of the dentigerous cyst with respect to impacted 37 and 38 was made. Other lesions included in the differential diagnosis were ameloblastoma of the left side of mandible and keratocystic odontogenic tumour.

Investigation

Orthopantomogram revealed a well defined solitary unilocular radiolucent lesion involving the left ramus and posterior body of the mandible. The radiolucency extended from the distal surface of 35 extending posteriorly. There was expansion of angle of mandible. There was mild scalloping of the borders. 38 was seen near the posterior inferior region of lesion approximating the inferior alveolar canal. External root resorption of 37 along with periapical radiolucency. (Figure 2)

Diagnosis

A incisional biopsy was taken where a diagnosis of UA subgroup1.3 was given which was followed by a excisional biopsy which confirmed the same diagnosis. The slides were stained with routine H&E stain. The histopathology showed cystic areas lined by ameloblastic epithelium with tall columnar basal cells displaying a palisading appearance. (Figure 3) A higher magnification showed hyperchromatic nuclei in tall columnar basal cells along with reverse polarity of the nuclei. The superficial epithelium showed prominent intracellular spacing, creating a stellate reticulum like appearance. Infiltration of ameloblastic epithelium into the connective tissue of the cyst wall as separate islands was also evident. The above histological features in relation to the radiographic findings were suggestive of a UA, subgroup 1.3 – luminal and intramural (Figure 4).

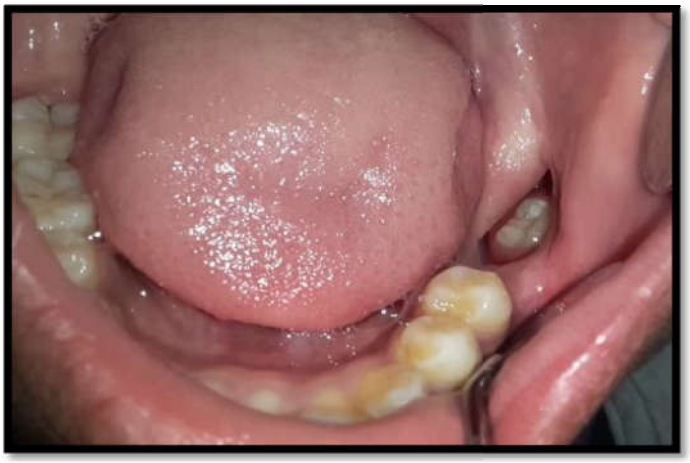

Figure 1. Preoperative intraoral photograph

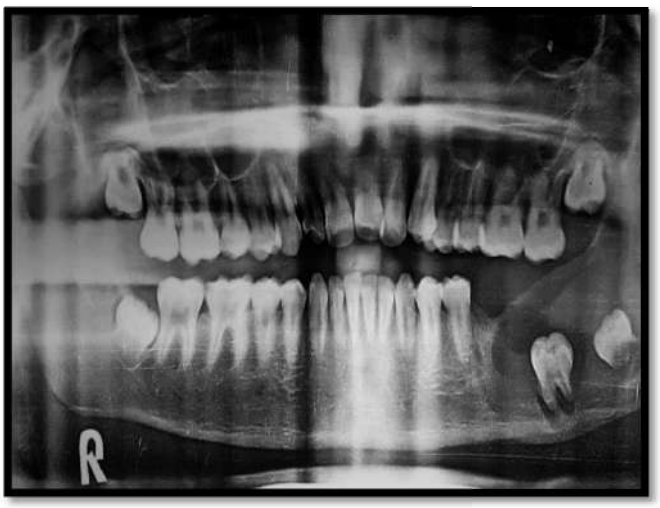

Figure 2. Preoperative orthopantomograph

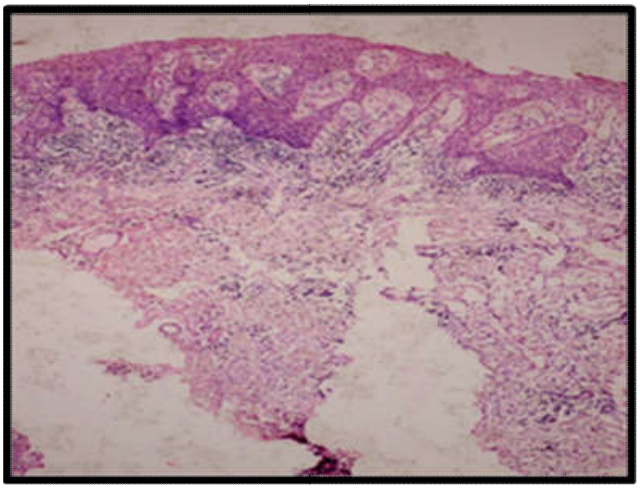

Figure 3. Hand E stained section under 10X magnification

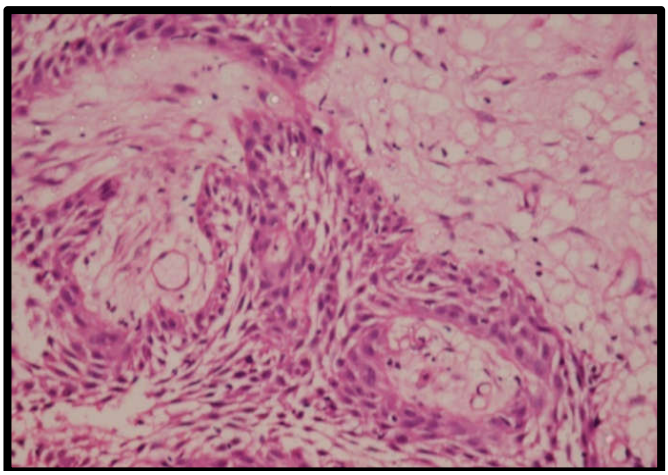

Figure 4. Hand E stained section under 40 X magnification

Treatment

The lesion was treated conservatively with careful enucleation. The patient when recalled for follow up did not report any fresh complaints and showed uneventful healing at the site of excision. The patient was then referred to the department of prosthodontics for construction of feeding plate. After the feeding plate a cast partial denture was given to the patient for restoring the routine functions.

Outcome and follow-up

Postoperatively, the surgical site healed uneventfully with no complications. The patient recovered well. Follow-up of 1, 3, 6 months and 1 year showed no recurrence and the patient remained asymptomatic.

DISCUSSION

The term ameloblastoma was suggested by Churchill in 1934. (Pallagatti *et al*., 2013) Ameloblastoma, a true neoplasm of the enamel organ tissue type that does not undergo differentiation up to the point of enamel formation. (Reddy *et al*., 2012) It has been described by Robinson as being a tumour that is usually unicentric, nonfunctional, intermittent in growth, histologically benign and clinically persistent. (Rai *et al*., 2015) According to the new classification, ameloblastoma is included under "benign neoplasms and tumour-like lesions arising from the

odontogenic apparatus showing odontogenic epithelium with mature fibrous stroma, without ectomesenchyme" and is divided into four types:

- The classic solid/multicystic ameloblastoma,
- The unicystic ameloblastoma,
- The peripheral ameloblastoma and
- The desmoplastic ameloblastoma, including the so called hybrid lesions (Reddy, 2012)

Robinson and Martinez were the first persons to describe UA in 1977. It is most commonly seen in individuals who are 16 to 20 years of age. Occasionally, lesions occur in younger patients; rarely, they have been found in patients up to the age of 40. About 90% of the lesions are located in the mandible and between 50 to 80% of these cases are associated with an impacted tooth (Jain *et al*., 2012). It refers to those cystic lesions that show clinical, radiographic or gross features of a mandibular cyst, but on histologic examination shows a typical ameloblastomatous epithelium lining part of the cyst cavity, with or without luminal and/or mural tumour growth hence, UA should be differentiated from odontogenic cysts and also should be recognized for the reason that the former has a higher rate of recurrence than the latter (Jain *et al*., 2012). Also, UA is believed to be less aggressive and responds more favorably to conservative surgery than the solid or multicystic ameloblastoma (Chaudhary *et al*., 2011). The radiographic appearance of UCAs has been divided into 2 main patterns: unilocular and multilocular and these have clear preponderance for the unilocular pattern (Nagalaxmi *et al*., 2013). This predominance is exceptionally marked for the dentigerous variant where the unilocular:multilocular ratio is 4.3:1.2. For the nondentigerous type this ratio is 1.1:1 (Eversole *et al*, 1984). The involved teeth show varying degrees of root resorption. Eversole *et al*. and Paikkatt *et al*. identified predominant radiographical patterns for UCA: unilocular, scalloped macromultilocular, pericoronal, interradicular, or periapical expansile radiolucencies (Nagalaxmi *et al*., 2013).

Leider *et al.* in 1985 proposed three pathogenic mechanisms for the occurrence of UA:

- The reduced enamel epithelium associated with the developing tooth, undergoes ameloblastic change with subsequent cystic transformation.
- Ameloblastoma arise in dentigerous or other types of odontogenic cysts in which the neoplastic ameloblastic epithelium is preceded temporarily by a non-neoplastic stratified squamous lining.
- Solid ameloblastoma undergoing cystic degeneration of ameloblastic islands with subsequent fusion of multiple microcysts and then into a unicystic lesion (Mishra *et al*., 2015 and Anchlia *et al*., 2016).

The reason why some ameloblastomas become completely cystic may be related to epithelial dysadhesion (e.g. defective desmosomes) or more likely, to the intrinsic production of proteinases (e.g. metalloproteinases, serine proteinases), enzymes that normally degrade the central zone of the enamel organ after tooth development (Rosenstein *et al*., 2001). UA are lesions with epithelial lining characteristically showing basal cell layer composed of columnar cells with hyperchromatic, palisaded nuclei. Reversed polarity of nuclei is present and a subnuclear vacuole is present between the basement membrane and nucleus, (Arora *et al*., 2015) which is according to Vickers and Gorlin criteria' given in 1970 for diagnosing ameloblasts (Anchlia *et al*., 2014). Overlying layer is composed of stellate reticulum like cells. Sometimes parakeratin layer is also noted and when keratinisation is present, an abrupt transition from the stellate reticulum like layer is observed. However, variability in epithelial lining of UA is also quite common, wherein it appears completely non-descriptive consisting of several layers of non keratinizing squamous cells mimicking radicular and infected dentigerous cyst and it becomes really difficult to diagnose such cases. Arora *et al*. added that diagnostic dilemma can be encountered especially in those cases where hyperplastic epithelium was growing into the underlying connective tissue and was associated with chronic inflammatory cell reaction. Such cases became difficult to differentiate from radicular cyst. Arcading proliferation of UA in particular is reminiscent of the arcading pattern seen in radicular cysts in response to inflammation. Further overlapping clinical and radiographic features add to diagnostic difficulty (Arora *et al*., 2015).

Ackermann classified this entity into the following three histologic groups:

Group I: luminal unicystic ameloblastoma (tumour confined to the luminal surface of the cyst);
Group II: Intraluminal/plexiform unicystic ameloblastoma (nodular proliferation into the lumen without infiltration of tumour cells into the connective tissue wall);
Group III: Mural unicystic ameloblastoma (invasive islands of ameloblastomatous epithelium in the connective tissue wall not involving the entire epithelium) (Nagalaxmi *et al*., 2013 and Hsu *et al*., 2014).

Histologic subgrouping by Philipsen and Reichart has also been described

Subgroup 1: luminal unicystic ameloblastoma;
Subgroup 1.2: luminal and intraluminal;
Subgroup 1.2.3: luminal, intraluminal and intramural;
Subgroup 1.3: luminal and intramural (Nagalaxmi, 2013).

Several attempts have been made in the past to distinguish the lining of the UCAs from that of odontogenic cysts. However, immunohistochemical markers like lectins (Ulex europaeus agglutinin I and Bandeiraea simplicifolia agglutinin I) and proliferating cells (proliferating cell nuclear antigen (PCNA) and Ki-67) may assist in their differential diagnosis (Nagalaxmi, 2013). However, Eversole *et al*. contend that currently unaided histologic assessment for UCA remains the gold standard for diagnosis, because of a variable response of UCA to tissue markers (Kumar *et al*., 2012). Histologically, the minimum criteria for diagnosing a lesion as UCA are the demonstration of a single cystic sac lined by odontogenic (ameloblastomatous) epithelium often seen only in focal areas (Kumar *et al*., 2013 and Kumar *et al*., 2012). UA compare favorably with their solid counterparts in terms of clinical behavior and response to treatment. Treatment planning depends on the patient's age, tumour size, location, radiographic appearance (unilocular or multilocular), final histopathological diagnosis and whether it is an initial presentation or a recurrence (Arora *et al*., 2015). The treatment regimen for UA can be divided into three modalities: Conservative (enucleation and curettage), marsupialization and radical surgery (resection with or without continuing defect).

In case of solid multicystic ameloblastoma, the treatment of choice is general resection with 1.5–2 cm margin beyond the radiological limit. In case of UA, some authors recommend a treatment modality of marsupialization followed by enucleation (Rai *et al*., 2015). The recurrence rate for UA after conservative surgical treatment (curettage or enucleation) is generally reported to be 10–20% and is on average < 25%. This is considerably <50–90% recurrence rate which are noted after the curettage of conventional solid or multicystic ameloblastomas (Rai *et al*., 2015). Recurrence of UA may be long delayed and a long term post operative follow up is essential for proper management of such patients (Gupta *et al*., 2011).

Clinical Importance

When a cyst of the jaw is associated with impacted tooth the most common provisional diagnosis is dentigerous cyst, at the same time UA (dentigerous variant) need to be considered as one of the differential diagnosis along with other cysts of the jaws. Then, it becomes important for the radiologist to carefully examine radiograph to assess the true dentigerous cyst impacted tooth relationship to narrow down the diagnosis. On removal of such cyst either in toto or as a cyst wall curettage, it is important for the surgeon and the pathologist to examine both the inner and outer wall of the cyst sac. The presence of several polypoid/exophytic/nodular growths in the inner and/ outer surface of the cyst wall may favor the initial diagnosis of UA rather than dentigerous cyst even though lack of these finding does not contradict the diagnosis of UA. When treatment of such cyst done based on radiographic diagnosis, entire tissue of the cyst after enucleation must be evaluated histopathologically by the pathologist to eliminate possibility of UA and when diagnosed histopathologically as UA, serial sectioning of the entire tissue is mandatory for the pathologist to arrive at the diagnosis of proper subtype of UA, as the recurrence rate changes accordingly. Thus, the collective opinion by the clinician, radiologist, surgeon and pathologist plays a very important role in the effective management of UAs.

Conclusion

Unicystic ameloblastoma is a variant of ameloblastoma, occurs more commonly in second decade of life, presents clinically as a painless swelling in mandible and unilocular radiolucency in most of the cases. Conservative treatment is preferable for younger age group; though recurrence is more commonly observed with this approach. The diagnosis of unicystic ameloblastoma should be strictly based on a combination of surgical, radiological and histopathological correlation.

REFRENCES

Gabhane M, Kulkarni M, Mahajan A. Unicystic Ameloblastoma of Mandible: A Case Report. *Indian Journal of Stomatology*, 2011 Dec 1;2(4).

Hertog D, Bloemena E, Aartman IH, van-der-Waal I. Histopathology of ameloblastoma of the jaws; some critical observations based on a 40 years single institution experience. *Med Oral Patol Oral Cir Bucal*., 2012 Jan 1;17(1):e76-82.

Hsu MH, Chiang ML, Chen JK. Unicystic ameloblastoma. *Journal of Dental Sciences*, 2014 Dec 31;9(4):407-11.

Jain A, Dandagi S, Sangle A, Ahmed V, Khan A. Unicystic ameloblastoma: a diagnostic dilemma and its management using free fibula graft: an unusual case report. *Indian Journal of Dental Advancements*, 2012 Apr 1;4(2):843-7.

Laxmidevi BL, Mohammed M, Kumar KA. Unicystic ameloblastoma with impacted third molar mimicking dentigerous cyst. *Journal of Interdisciplinary Dentistry*, 2015 Jan 1;5(1):37.

Lee SK, Kim YS. Current concepts and occurrence of epithelial odontogenic tumors: I. Ameloblastoma and adenomatoid odontogenic tumor. *The Korean Journal of Pathology*, 2013 Jun 25;47(3):191-202.

Nagalaxmi V, Sangmesh M, Maloth KN, Kodangal S, Chappidi V, Goyal S. Unicystic mural ameloblastoma: An unusual case report. *Case reports in dentistry*, 2013 Apr 23;2013.

Pallagatti S, Sheikh S, Aggarwal A, Singh R, Gupta D, Gupta R, Singla I, Gupta P. Unicystic ameloblastoma of the mandible with all variants. *J Pak Med Stud*., 2013 Apr 1;3(2):106-9.

Pallagatti S, Sheikh S, Aggarwal A, Singh R, Gupta D, Gupta R, Unicystic ameloblastoma of the mandible with all varian ts. *J Pak Med Stud*., 2013; 3(2): 106109

Philipsen HP, Reichart PA: Classification of odontogenic tumors and allied lesions. Odontogenic tumors and allied lesions Quintessence Pub. Co. Ltd 2004.

Rai A, Arora M, Naikmasur VG, Malhotra V, Kumar A. Unicystic Plexiform Aameloblastoma. *Universal Research Journal of Dentistry*, 2015 Sep 1;5(3):211.

Rajendran, R. and B Sivapathasundharam. Shafer's Textbook of Oral Pathology. 5th Edn. *Elsevier*. New Delhi. 2007.

Reichart PA, Philipsen HP, Sonner S. Ameloblastoma: biological profile of 3677 cases. *Eur J Cancer B Oral Oncol.,* 1995.

Thankappan *et al*. Unicystic Ameloblastoma: 3 case reports and review of literature. *Journal of Indian Academy of Oral Med and Radiology*, 2008; 20: 65-70.

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