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CASE REPORT

Unicystic Ameloblastoma as a Differential Diagnosis for Odontogenic Cysts

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ABSTRACT

Unicystic ameloblastoma (UA) is a variant of the solid or multicystic ameloblastoma. Radiographically, though the unilocular appearance is common, it can produce multilocular lesions also. Histologically, the minimum criterion for diagnosing a lesion as UA is the demonstration of a single cystic sac lined by ameloblastomatous epithelium often only in focal areas with or without areas of intraluminal or intramural proliferation or both. The clinical and radiologic presentation of UA can give a confusing picture of odontogenic cysts especially when it is seen in the inter-radicular or periapical area. Also, tooth associated UA may show features similar to dentigerous cyst. Hence histopathologic examination is essential to diagnose such cases. Here we present such a lesion which was initially diagnosed as an odontogenic cyst.

Keywords: Ameloblastomatous epithelium, Dentigerous cyst, Unilocular.

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INTRODUCTION

Unicystic ameloblastoma (UA) is a variant of the multicystic or solid ameloblastoma. UA was first described as a distinct entity by Robinson and Martinez in 1977. Clinically, it may be of 'dentigerous' or 'nondentigerous' variety. Though the lesion is a single cystic cavity, it can present as a unilocular or a multilocular radiolucency. Histologically, the minimum criterion for diagnosing a lesion as UA is demonstration of a single cystic sac lined by ameloblastomatous epithelium often seen only in focal areas.[1] The subtypes include simple, simple with intraluminal proliferations, simple with both intraluminal and intramural proliferations and simple with intramural proliferations only.[1] The clinical and radiologic presentation of the nondentigerous variant can give a confusing picture of odontogenic cysts especially when it is seen in the inter-radicular or periapical area. Also, dentigerous variety may show features similar to dentigerous cyst. Hence, histopathologic examination is essential to diagnose such cases. Here we present such a lesion which was initially misdiagnosed as an odontogenic cyst.

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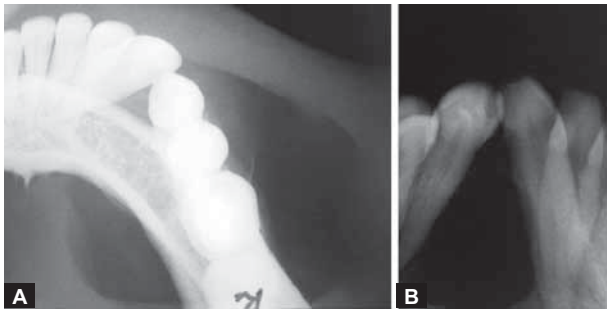
CASE REPORT

A 25-year-old female patient presented with a swelling in the lower jaw noticed since 3 months. There was no other symptom like pain or paresthesia. On examination, a swelling of size 3 × 3 cm was seen extending from 42 to 45 (Fig. 1). The crown of 43 was found to be distally tipped and that of 44 mesially tipped. Both the buccal and lingual cortical plates were found to be expanded. Radiographs were taken and revealed the presence of a unilocular radiolucency between the roots of 43 and44 (Fig. 2B). Buccal and lingual cortical plates were expanded (Fig. 2A) and the roots of 43 and 44 were seen to be displaced. On aspiration, straw colored fluid was obtained and the cytology was nonconclusive. A provisional diagnosis of odontogenic keratocyst/lateral periodontal cyst was given. Enucleation of the cyst was done along with 43 and 44.

The cystic specimen was of size 2 × 2 × 1.5 cm with a smooth surface and brownish in color. The cyst was found attached to the buccal surface of 43 and 44 and the inner surface was seen to be smooth (Fig. 3). The histopathologic examination revealed the presence of a lining epithelium showing varying thickness. At many areas, basal cells were tall columnar with polarized nucleus and superficial cells

Fig. 1: Intraoral photograph showing the swelling in relation to 43 and 44



Figs 2A and B: (A) Occlusal radiograph showing an expansile lesion, (B) IOPA showing ovoid radiolucency tilting the roots of 43 and 44

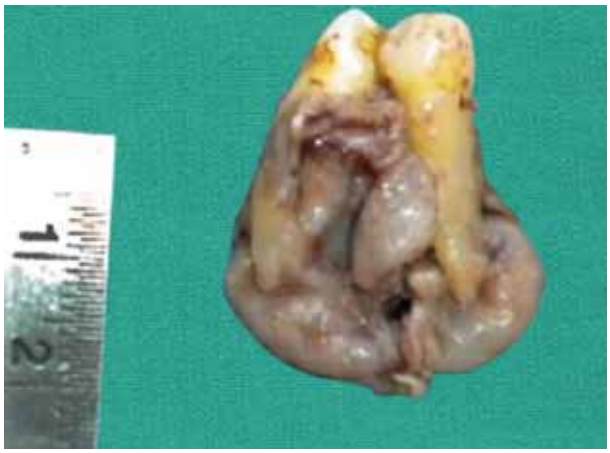


Fig. 3: Gross cystic specimen attached to the labial surface of roots of 43 and 44

resembled the stellate reticulum (Fig. 4A). Invading islands of ameloblastoma were found within the connective tissue capsule (Figs 4B and D). Plexiform type of proliferation of the lining epithelium was also noticed in some areas (Fig. 4C). Based on these findings a report of unicystic ameloblastoma with mural proliferation was given.

DISCUSSION

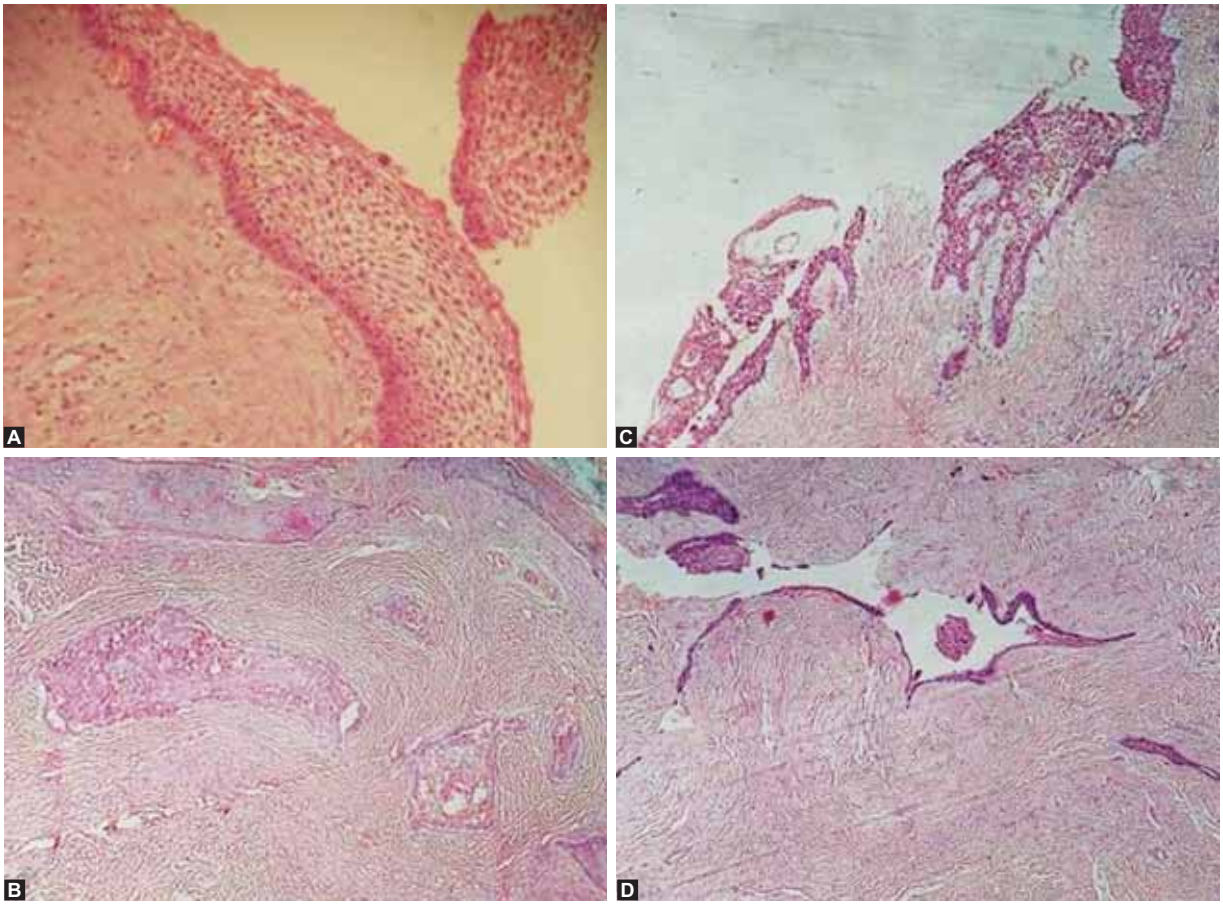
UA comprises 5 to 22% of all ameloblastomas. It is less aggressive with a recurrence rate less than 25%.[2] The term 'unicystic' implies that the lesion is either unilocular or arise from a grossly unilocular cystic cavity.[3] The mandibular molar and ascending ramus region are the most commonly affected sites and it is usually asymptomatic. It has an almost identical sex distribution. UA is usually found in the 2nd and 3rd decade.[4] The nondentigerous variety is found to occur at a later age than the dentigerous variety.[1] The various radiographic patterns documented in UA include pericoronal unilocular, extensive pericoronal unilocular, pericoronal scalloped, periapical unilocular, inter-radicular and multilocular.[5]

Histologically, UA is a cystic cavity lined by varying epithelium showing focal areas of ameloblastomatous epithelium. Sometimes nodules may be seen arising from the lining and projecting in to the lumen comprising of odontogenic epithelium with a plexiform pattern (intraluminal). Also, these cyst wall may contain one or more mural nodules or focal thickenings of the cyst wall which comprise invasive islands or strands of conventional ameloblastoma (intramural).[1] UA only has a good prognosis if it is still confined within the cyst wall.[6] The histological subtypes of UA as modified by Philipsen and Reichart are:

- Simple UA
  - Simple and intraluminal UA
    - Simple, intraluminal and intramural UA
  - Simple and intramural UA

UAs may present with characteristics of an odontogenic cysts as they are less aggressive than the conventional ameloblastoma, tend to occur at an earlier age, and often exhibit a myxoid-like stroma.[7] Hence, most small UAs may be diagnosed clinically as odontogenic cysts, they are enucleated on that basis. It is only when the pathologist examines the entire specimen that the diagnosis of UA becomes apparent. UA may be lined by variable epithelium ranging from typical ameloblastic characteristics to metaplastic epithelium consisting of layers of nonkeratinizing squamous cells, differentiation from other odontogenic cysts may be problematic. Odontogenic cell rests are usually found in odontogenic cysts.[4] Some of them may show palisading of basaloid cells located at the outer circumference while other signs of true ameloblastomatic differentiation is lacking, such as basal cell hyperchromatism, polarization of the basal cell nuclei, or vacuolation of the basal cell cytoplasm.[8] Intraluminal proliferation in UA may resemble hyperplastic odontogenic epithelium. But the absence of inflammation and the presence of delicate, vascular connective tissue of UA is a useful clue.[6] These points should be carefully considered as all areas of the cyst lining may not show the Vickers and Gorlin criteria for ameloblastoma.

Many studies have been done on the various methods which could be used to differentiate UA from odontogenic cysts. The earliest attempts were on the expression of blood cell carbohydrates. Though it was found useful initially, it was later disproved.[9,10] Imaging studies like the use of Contrast enhanced MRI have also been documented. It has been found to be useful as there was thick enhancement in the walls of UA.[11] Histoenzymological studies were done to find the variations in levels of activities of oxidative enzymes, diaphorases, acid phosphatases and naphthol esters. Odontogenic cysts showed nonspecific activity whereas in ameloblastoma there were uniformly low oxidative enzymatic activities in the epithelium and widespread activity of alkaline phosphatase in the stroma.[12] Thus, alkaline phosphatase activity may be useful in distinguishing



Figs 4A to D: (A) Lining epithelium showing typical ameloblastomatous features (H&E, 400×); (B) ameloblastomatous islands in the connective tissue capsule (H&E, 400×); (C) plexiform type of proliferation of the lining epithelium (H&E, 100×); (D) mural ingrowth of epithelial lining seen (H&E, 100×)

the cystic ameloblastomas. Studies done on the binding sites for lectins like Ulex europaeus agglutinin-I(UEA-I) and Bandeiraea simplifolia agglutinin-I(BSA-I)showed that only cysts gave positive reactions.[13] Expressions of cytokeratins in jaw cysts have been investigated and showed that they have distinct differences in their cytokeratin content.[14] Studies done on silver stained nucleolar organizer regions (AgNOR) did not show any significant differences between odontogenic cysts and ameloblastomas.[15,16] Comparison of proliferating cell nuclear antigen (PCNA) and Ki-67 expression of epithelial lining of UA with odontogenic cyst linings have been done.[17] They found that all areas of cystic tumor epithelium contained significantly more PCNA positive cells than dentigerous cysts even in areas similar to dentigerous cyst lining. Also in UA, cell islands invading the cyst wall-exhibited a significantly higher PCNA index than cells in the intraluminal proliferation and in the cystic tumor lining. Similar results were obtained for Ki-67 expression. Calretinin is a calcium binding protein found in normal human tissues and tumors like ameloblastoma. Studies have shown that calretinin is expressed only by UA indicating that it may be a specific marker for ameloblastic tissues.[18] Both the PCNA and calretinin studies give support to the theory that UA are distinct lesions and may arise *de novo*. A definitive diagnosis of UA can be made only after examining the whole specimen. Hence, incision biopsy may not always be correct as the epithelium shows variation. Thus, multiple sections from the whole specimen should be examined for a final diagnosis.

CONCLUSION

Currently histologic examination is the most sensitive tool for differentiating UA from odontogenic cysts.[4] The clinical and radiologic findings contribute to the diagnosis. Careful examination of the whole specimen is essential with multiple sectioning. Thus, lesions which clinically and radiographically appear to be odontogenic cysts may prove to be ameloblastomas. UA is a tumor with a propensity for recurrence, especially when the ameloblastic focus penetrates the adjacent tissue from the wall of the cyst. The ability to predict this potential occurrence prior to surgery would greatly enhance therapeutic strategies for reducing the incidence.

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