



DENTIGEROUS CYST OR UNICYSTIC AMELOBLASTOMA: A DIAGNOSTIC DILEMMA

Dr. Kaushal Jha¹, Dr. Satyajith Naik², Dr. Swati Tripathi^{3*} and Dr. Srinandan Pradhan⁴

¹Third Year Post Graduate Student Department of Pedodontics Institute of Dental Sciences.

²Professor, Department of Pedodontics, Institute of Dental Sciences.

³Reader, Department of Pedodontics, Institute of Dental Sciences.

⁴1st year Post Graduate Student, Department of Pedodontics.

***Corresponding Author: Dr. Swati Tripathi**

Reader, Department of Pedodontics, Institute of Dental Sciences.

Article Received on 29/02/2016

Article Revised on 20/03/2016

Article Accepted on 10/04/2016

ABSTRACT

This is a case of an ameloblastoma which arose in the wall of a dentigerous cyst. The clinical presentation radiographic and histological findings were similar to those of dentigerous cysts, which were again affirmative while doing an incisional biopsy. Enucleation was done intraorally under local anaesthesia. Post-operative histopathological examination of cystic lining confirmed unicystic ameloblastoma.

KEY WORDS: ameloblastoma, cyst, enucleation, unicyclic.

INTRODUCTION

Ameloblastoma is a benign odontogenic neoplasm which frequently affects the mandible. The term ameloblastoma includes several clinicoradiological and histological types. Apart from the most commonly encountered clinicopathologic models, there are few variants, whose biological profile is unknown or not elicited. Among these types, unicystic ameloblastoma is the least encountered variant of the ameloblastoma. Unicystic ameloblastoma 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 tumor proliferation. Unicystic ameloblastoma is a less encountered variant of the ameloblastoma and is believed to be less aggressive. As this tumor shows considerable similarities with dentigerous cysts, both clinically and radiographically, so histopathological investigations of such pathology is of immense importance to come on conclusive diagnosis. The term 'Unicystic Ameloblastoma' was adopted in the second edition of the international histologic classification of odontogenic tumors.

Unicystic ameloblastoma was first described by Robinson and Martinez in 1977.^[1] It accounts for 10-15% of all intraosseous ameloblastomas.^[2] The term 'unicystic ameloblastoma' is derived from the macro- and microscopic appearance, the lesion being a well defined, often large monocystic cavity with a lining, focally but rarely entirely composed of odontogenic (ameloblastomatous epithelium). It is often accompanied

by an innocuous epithelium of varying histologic appearance that may mimic the lining of a dentigerous or radicular cyst.^[3] This case reports describes an unusual representation of unicystic ameloblastoma in young female patient in the maxillary anterior region.

CASE REPORT

A 10 year-old female patient presented with a progressively increasing swelling in the left maxillary midface region for last six month. Because of lack of relief after consultation with a dentist, she was referred to Dental College. The patient was apparently healthy and her past medical history was noncontributory. Extraoral examination revealed a single diffuse solitary, oval swelling on left side of maxilla extending laterally till the ala of nose. Intra oral examination revealed the diffuse swelling involving both labial and palatal sides of 22, 63, 24, with distal tipping of 22 and impacted 23 and cortical plate expansion in relation to 22 to 24 (Fig.I). Grade II mobility in relation to 63 was also observed. There was no history of pain, toothache, pus discharge or paresthesia. Cystic fluctuancy was present on palatal aspect of swelling. A provisional diagnosis of dentigerous cyst was made on clinical findings.

Diagnostic orthopantomograph (Fig.II) showed a well defined unilocular radiolucent lesion associated with 22, 63 and 23 roughly ovoid in shape and surrounded by cortical border and mesially inclined 22, 23 was seen impacted and enclosed within the lesion. The size of the lesion was roughly 3.0x2.cm.

Routine laboratory parameters were normal. On aspiration a yellow straw- colored 2 ml fluid was obtained and provisional diagnosis of dentigerous cyst in relation to impacted 23 was made. Under local anaesthesia cystic lesion was enucleated along with this 63 was removed. Grossly, the creamy white soft tissue specimen was around 4.0 x1.5 cm in dimension, and soft in consistency.

The tissue lining was subjected for histopathological examination. The final diagnosis after histopathological examination was made of unicystic ameloblastoma arising from dentigerous cyst. Post treatment the patient is under strict follow up.



Figure 1.Extraoral view of swelling on the left side middle third of face



Figure 2. Intraoral view-Swelling extending from 22 to 24 with normal overlying mucosa.



Figure 3. Occlusal radiograph

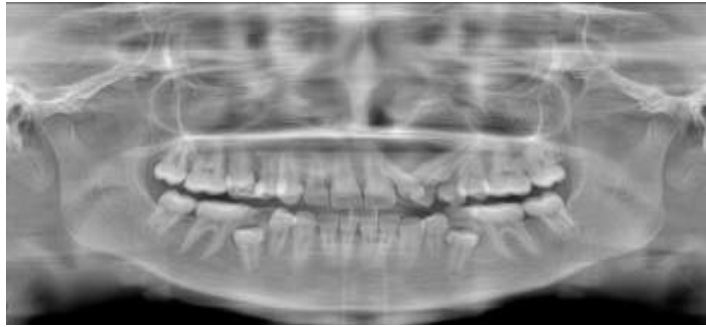


Figure 4. OPG showing well defined unilocular radiolucency on left maxilla



Figure 5. Extracted left primary canine



Figure 6. Operated specimen



Figure 7. Cystic cavity after enucleation

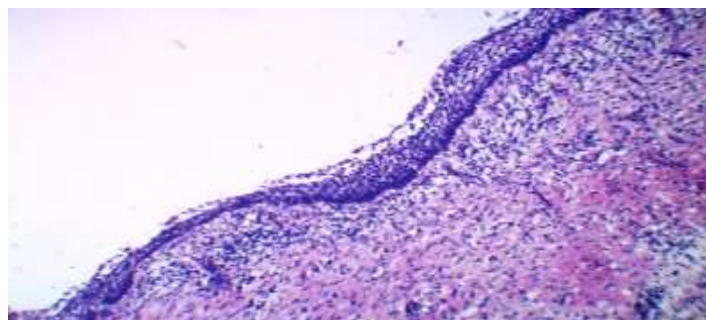


Figure 8. Low power photomicrograph shows ameloblastomatous cystic epithelial lining (H&E stain, 4 x)

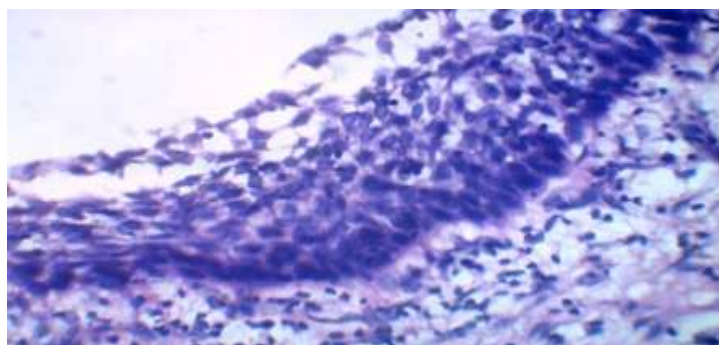


Figure 9. High power photomicrograph shows ameloblastomatous cystic epithelial lining (H&E stain, 40 x)



Figure 10. Six months follow up OPG

DISCUSSION

In their day to day practice Pediatric dentist frequently encounter swelling in and around orofacial structure. Sometime it's convenient to come on diagnosis of the lesion, but at times such swellings are baffling.

Swelling in the jaw can be due to various etiological factors like periapical abscess, cyst, tumour etc. The literature review shows that these cystic lesions if left untreated often shows tumour like pattern of growth. Various variants of ameloblastomas are found in young children but among it unicystic ameloblastomas are a rare variant, which refers to those cystic lesions that shows clinical, radiographic or gross features of a jaw cyst but on histological examination shows a typical ameloblastomatous epithelium lining in the cystic cavity with or without luminal and/or mural tumor growth.^[5] Even though the lesion is not as aggressive as the solid ameloblastomas, an accurate histopathologic diagnosis is essential for the treatment and prognosis. It's most commonly found in mandible than maxilla in ratio of 13:1.^[6] It appears more frequently in the second or third decade of life with gender distribution shows a slight male predilection with a male to female ratio of 1.6 : 1. However, when the tumor is not associated with an unerupted tooth, the gender ratio is reversed to a male to female ratio of 1 : 1.8.^[4]

Basically, ameloblastomas are benign tumors whose importance lies in its potential to grown into enormous size with resulting bone deformity. Ackerman et al classified unicystic ameloblastoma in three histologic subtypes: luminal (type 1), intraluminal (type 2) in which

tumour is confined to the epithelium of the cyst and may be treated conservatively by enucleation; mural pattern (type 3) in which tumour is present in the connective tissue wall of the cyst and should be treated aggressively in exactly the same manner as multicystic ameloblastoma.^[3]

Maxillary unicystic ameloblastoma are very rare finding. The first case reported by Gardner and colleagues in 1987 occurred in a 12 year old boy in the molar area.^[7] There was no bone infiltration.^[4] Unicystic ameloblastoma in the anterior maxilla is considered to be a rare and atypical.^[8] These findings are similar to our case in which the lesion was present in anterior portion of maxilla without any bony infiltration. The lesion was asymptomatic, although a large lesion may cause painless swelling of the jaws as seen in our case.^[9] The mean age of nonimpacted tooth-related cystic ameloblastoma is 5 years in comparison to 16.5 years for the impacted tooth-related variant.^[10] Most of the unicystic ameloblastomas are associated with an impacted tooth, the mandibular third molar being involved most often. But in our case it was associated with impacted maxillary canine, and it was of dentigerous variant.

Occurrence of ameloblastomas in children and adolescents who are below 18 years of age is uncommon. Only 14.6% of cases ameloblastomas were seen in children and adolescents among 206 cases which were evaluated by Lucas.^[11] More than 90% are located in the mandible in the posterior region, followed by the parasymphysis region, the anterior maxilla, and the posterior maxilla. But in our case the lesion was present

in anterior region of maxilla which makes it rare. Unicystic ameloblastoma is usually asymptomatic, although a large tumor may cause painless swelling of the jaws with facial asymmetry which was concurrent to our case.

There have been many debates regarding if unicystic ameloblastoma develops *de novo* or arises in an existing cyst.^[4] Leider *et al.*^[12] proposed three pathogenic mechanisms for the evolution of unicystic ameloblastoma: reduced enamel epithelium, from dentigerous cyst and due to cystic degeneration of solid ameloblastoma. It often involves an impacted tooth and the focal area of the cystic tumor lining is often composed of a non specific, thin epithelium that mimics the dentigerous cyst lining.^[1] In our case the association with an impacted tooth and presence of non specific thin epithelium lining in focal areas of cystic tumor supporting the second hypothesis i.e. arising from preexisting dentigerous cyst. Possibility of misdiagnosing such cases as dentigerous cyst poses a problem where repeat and deeper biopsies are advisable to reveal the underlying tumorous proliferation as seen in the present case.

In view of the reported ameloblastomatous potential of dentigerous cysts, it is thus important to be able to recognize true ameloblastomatous epithelium from ameloblastoma-like epithelium. In most cases of dentigerous cysts, the presence of an ameloblastomatous epithelial lining in inflamed dentigerous cysts is insufficient to diagnose unicystic ameloblastomas, unless other more diagnostic features of unicystic ameloblastomas are evident.^[9]

CONCLUSION

In the present case, making a diagnosis was possible only because histopathological examination of the tissue obtained post surgery. It highlights the importance of enucleation as the first choice of treatment for large, cystic lesions, instead of conservative procedures like decompression and marsupialization which are used for children. Though marsupialization might help in the preservation of vital structures, keeping in view, the potential of more aggressive transformation of the cystic lining, complete removal of the lining by enucleation, with emphasis on possible preservation of vital structures, as in our case, clubbed with a thorough follow-up, is more appropriate.

REFERENCES

1. Li TJ, Wu YT, Yu SF, Yu GY. Unicystic ameloblastoma: A clinicopathological study of 33 Chinese patients. *Am J Surg Pathol* 2000; 24: 1385-92.
2. Langlais RP, Langland OE, Nortje CJ. Pericoronal radiolucencies without opacities. In: *Diagnostic imaging of the jaws*. USA: Williams and Wilkins; 1995; 293-7.
3. Ackermann GL, Altini M, Shear M. The unicystic ameloblastoma: A clinicopathological study of 57 cases. *J Oral Pathol* 1988; 17: 541-6.
4. Philipsen HP, Reichart PA. Unicystic ameloblastoma. In: *Odontogenic tumors and allied lesions*. London: Quintessence Pub. Co. Ltd; 2004; 77-86.
5. Paikkatt VJ, Sreedharan S, Kanna VP. Unicystic ameloblastoma of the maxilla: A case report. *J Indian Soc Pedod Prev Dent*. 2007; 25: 106-10.
6. Robinson L, Martinez M. Unicystic ameloblastoma. *Cancer* 1977; 40: 2278-85.
7. Gardner DG. Plexiform unicystic ameloblastoma. A diagnostic problem in dentigerous cysts. *Cancer* 1981; 47: 1358-63.
8. Navarro CM, Principi SM, Massucato EM, Spoto MR: Maxillary unicystic ameloblastoma. *Dentomaxillofac Radiol* 2004; 33:60-2.
9. Peter A, Reichart PA, Phillipsen HP. Chicago: Quintessence Pub. *Odontogenic tumors and allied lesions*, 2004; 77-86.
10. Eversole LR, Leider AS, Strub D. Radiographic characteristics of cystogenic ameloblastoma. *Oral Surg Oral Med Oral Pathol* 1984; 57: 572-74.
11. Mastan KM, Rajkumari S, Deepasree M. Neoplasms Associated with odontogenic cysts. *J. of Dentistry and Oral Hygiene*. 2011; 3(10): 123-30.
12. Leider AS, Eversole LP, Barkin ME. Cystic ameloblastoma. *Oral Surg Oral Med Oral Pathol*. 1985; 60: 624-30.