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CASE REPORT

Unicystic Ameloblastoma in 6-Year-Old Child and Its Significance

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ABSTRACT

Ameloblastoma is a true neoplasm of odontogenic epithelial origin. Unicystic ameloblastoma refers to those cystic lesions that show clinical, radiographic or gross features of a mandibular 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. It accounts for 5 to 15% of all intraosseous ameloblastomas. They believed to be less aggressive and respond more favorably to conservative excision than the solid or multicystic ameloblastomas. We report a case of unicystic ameloblastoma of the anterior part of the mandible that was treated by enucleation and use of Cornoy's solution under suspicion of an odontogenic cyst. The nature of the lesion became evident only when the enucleated material was available for histologic examination. With this report, we illustrate the importance and complexity of a differential diagnosis of lesions with a cystic aspect in the anterior region of the mandible, among them are dentigerous cyst, odontogenic keratocysts, adenomatoid odontogenic and unicystic ameloblastoma.

Keywords: Unicystic ameloblastoma, Enucleation, Ameloblastoma, Unicystic, Cornoy's solution.

INTRODUCTION

Ameloblastomas are benign tumors whose importance lies in their potential to grow to enormous size with resulting bone deformity. They originate from the epithelium involved with the formation of teeth: Enamel organ, odontogenic rests of Malassez, reduced enamel epithelium and odontogenic cyst lining. It is a slow-growing, persistent and locally aggressive neoplasm of epithelial origin. Its peak incidence is in the 3rd to 4th decades of life. It is often associated with an unerupted third molar.[1]

There are three forms of ameloblastomas, namely multicystic, peripheral and unicystic tumors.[2] Multicystic ameloblastoma is the most common variety and represents 86% of cases. Peripheral tumors are odontogenic tumors with the histological characteristics of intraosseous ameloblastoma that occurs solely in the soft tissues covering the tooth-bearing parts of the jaws. Unicystic tumors include those that have been variously referred to as mural ameloblastomas, luminal ameloblastomas and ameloblastomas arising in dentigerous cysts.[3] The goal of treatment ameloblastoma is to achieve complete excision.

The literature indicates that the cystic variant is biologically less aggressive and has a better response to enucleation or curettage than the solid ameloblastoma.[4] The overall recurrence rate of unicystic ameloblastoma was 15% with some evidence to suggest that the mural histological subtype had a greater recurrence rate than the others.[5]

We present a case of a unicystic mandibular ameloblastoma in a 6-year-old kid who was treated for under the impression of odontogenic keratocyst, but on histopathological examination, showed it to be different.

CASE REPORT

A 6-year-old female child was referred to the Department of Oral and Maxillofacial Surgery, RV Dental College and Hospital, Bengaluru, Karnataka, on 2nd August, 2008, because of a slowly growing swelling on the lower front region of the face since 1 month (Fig. 1). There was no associated pain, tenderness, difficulty in opening the mouth, chewing and no associated discharge. On clinical examination, there was a hard nontender mass, measuring 3 by 3 cm arising from anterior region of the mandible, involving the left parasymphyseal area from midline till angle of the mouth. Intraorally (Fig. 2), swelling extending from free gingiva till sulcus both buccal and lingually involving deciduous central, lateral, canine region on left side which altered the vestibular depth. The overlying mucosa was normal. Swelling was nontender, firm, hard in consistency on palpation and centrally fluctuant with obvious expansion of the buccal and lingual cortex. Personal history and medical history were unremarkable. She was taking no medication and had no history of known drug allergy.

An orthopantomogram (OPG) was done, which showed large cystic lesion in the right side of anterior part of the mandible in which a well-defined radiolucency extending from mesial side

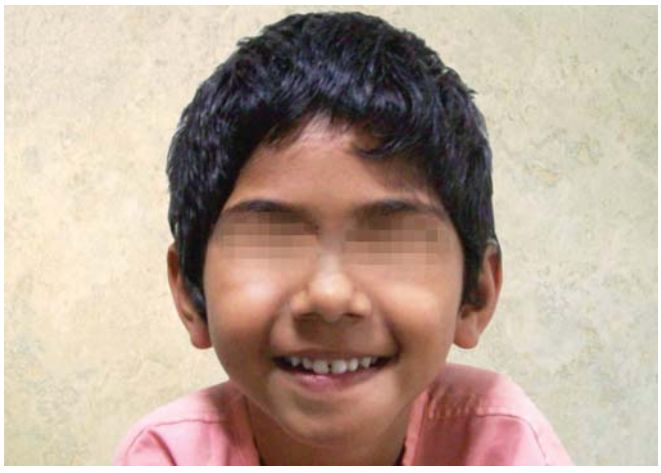

Fig. 1: Preoperative extraoral photograph

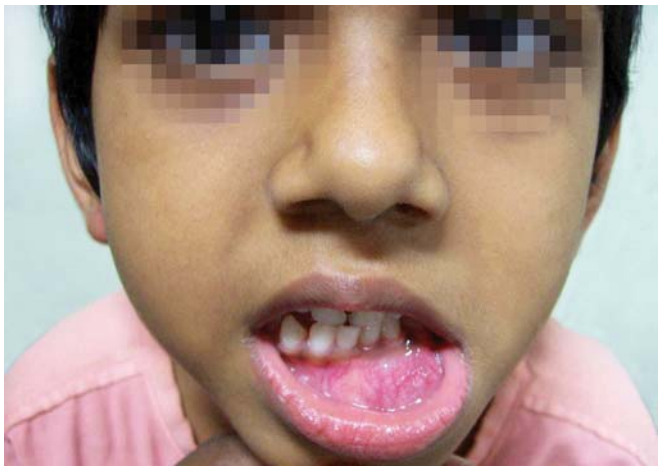

Fig. 2: Preoperative intraoral photograph

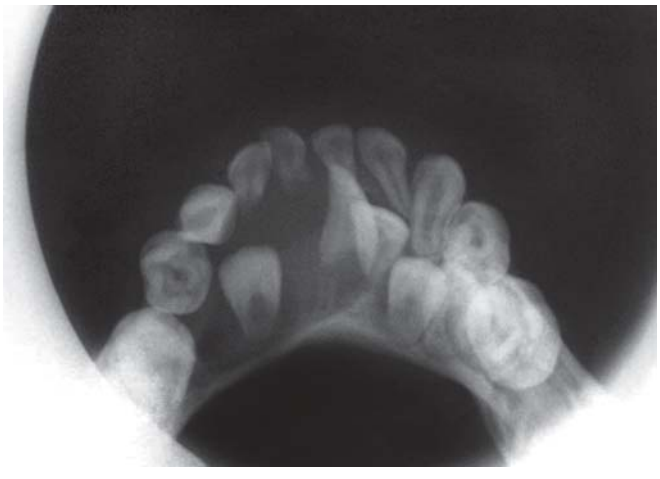

Fig. 3: Occlusal radiograph

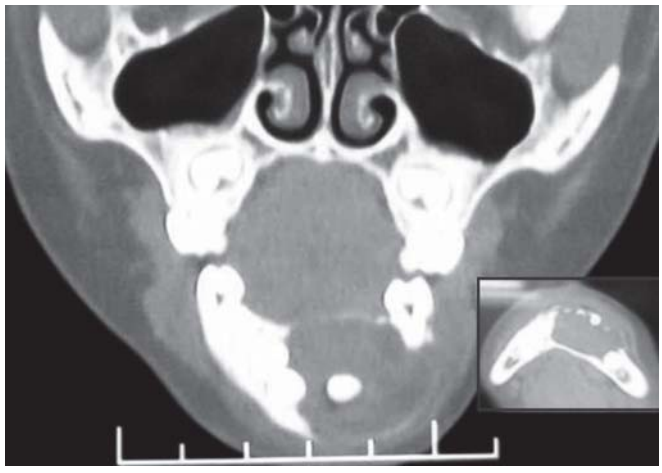

Fig. 4: CT scan (coronal view)

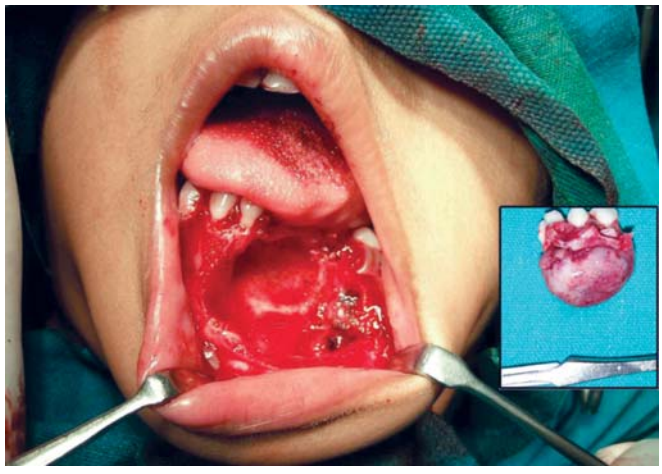

Fig. 5: Intraoperative enucleation

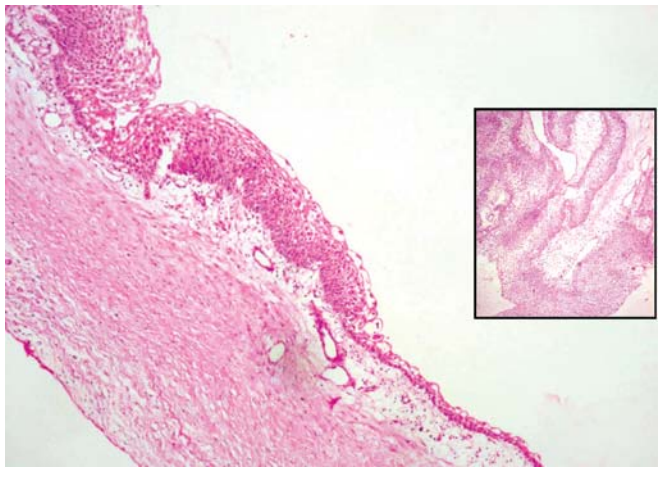

Fig. 6: Histopathology

of right deciduous lateral incisor to distal side of left deciduous first molar. Occlusal view (Fig. 3) radiograph showed expansion of buccal cortical plate. CT scan (Fig. 4) showed that the cystic lesion was confined to the anterior part of the mandible extending from right deciduous lateral incisor to left deciduous first molar with a thinned-out cortex (Fig. 3). Fine needle aspiration cytology (FNAC) was performed. The cytosmear showed numerous neutrophils, macrophages and few desquamated epithelial cells in an eosinophilic background, and hence, no conclusive evidence could be drawn from the same.

In consideration of the prospect of the underlying tooth germs of the permanent teeth, enucleation with extraction of the overlying deciduous teeth (B, C, D and E) was planned on 28 August, 2008. The patient was taken up for surgery under general anesthesia. Routine preparations undertaken, lignocaine with adrenalin injected for hemostasis and crevicular incision

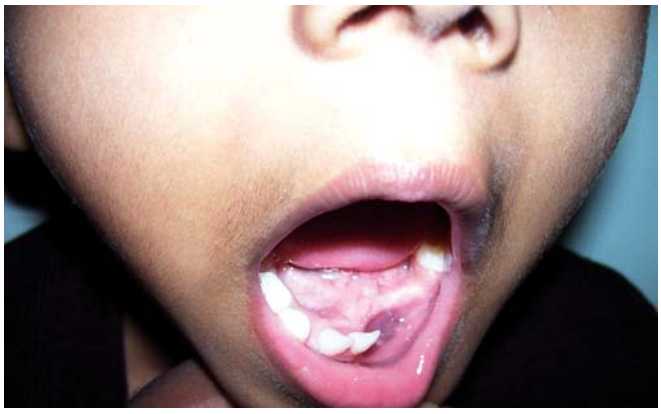

Fig. 7: Postoperative photograph

throughout the mandible to expose the whole of the buccal cortex. Thinned-out cortex was drilled and rounded out. The cystic lining was separated from the inferior border of the mandible taking care not to injure the inferior alveolar nerve and mental nerve. The deciduous teeth was delivered out with its cystic lining (Fig. 5) and ensured complete enucleation. Cornoy's solution used.

The subsequent histopathologic diagnosis was a (Fig. 6) unicystic ameloblastoma. Histologically, the cyst was lined by ameloblastic epithelium with tall columnar basal layer, subnuclear vacuoles, reverse nuclear polarity and thin layer of edematous, degenerate appearing stellate cells.

The immediate postoperative healing was uneventful (Fig. 7). The patient was closely followed for 2 years, during which time there was no sign of recurrence.

DISCUSSION

Unicystic ameloblastoma, a variant of ameloblastoma first described by Robinson and Martinez[6] in 1977, refers to those cystic lesions that show clinical and radiologic characteristics of an odontogenic cyst, but in histologic examination, show a typical ameloblastomatous epithelium lining part of the cyst cavity with or without luminal and/or mural tumor proliferation. Prior to the report by Robinson and Martinez, this variant had been referred to as a mural or intraluminal ameloblastoma.

This variant is believed to be less aggressive, usually occurs in a younger age group, with about 50% of the cases occurring in the second decade of life. More than 90% are located in the mandible.[7] Between 50 and 80% of cases are associated with tooth impaction, the mandibular third molar being most often involved.

There are various subtypes of unicystic ameloblastoma depending on the character and extent of ameloblastic proliferation within the cyst wall. Vickers and Gorlin, in 1970,[8] first described the features of the early ameloblastic change that occurs within the wall of a cyst. These include hyperchromatism of the nuclei in the basal cell layer of the epithelial lining, palisading and polarization of the basal cell nuclei away from the basement membrane and cytoplasmic vacuolization of the basal cells.

In a clinicopathologic study of 57 cases of unicystic ameloblastoma, Ackermann[2] classified this entity into the following three histologic groups:

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

Another histologic subgrouping by Philipsen and Reichart[3] has also been described as follows:

- *Subgroup 1*: Luminal
- *Subgroup 1.2*: Luminal and intraluminal
- *Subgroup 1.2.3*: Luminal, intraluminal and intramural
- *Subgroup 1.3*: Luminal and intramural.

The unicystic ameloblastomas diagnosed as subgroups 1 and 1.2 can be treated conservatively (enucleation), whereas subgroups 1.2.3 and 1.3 showing intramural growths require treated radical resection, as for a solid or multicystic ameloblastoma.

Following enucleation, vigorous curettage of the bone should be avoided as it may implant foci of ameloblastoma more deeply into bone. Chemical cauterization with Carnoy's solution[9] is also advocated for subgroups 1 and 1.2 which will benefit to reduce the rate of recurrence. Subgroups 1.2.3 and 1.3 have a high risk for recurrence, requiring more aggressive surgical procedures. This is because the cystic wall in these cases has islands of ameloblastoma tumor cells and there may be penetration into the surrounding cancellous bone.[10]

If specimens show extension of tumor into fibrous cyst wall for any appreciable distance, subsequent management is more controversial. Some surgeons believe that local resection of area is indicated as a prophylactic measure, while others prefer to keep the patient under radiographic observation and delay further treatment until there is evidence of recurrence. Recurrence rate of 10 to 20% has been reported after enucleation and curettage of unicystic ameloblastoma. This is considerably less than 50 to 90% recurrence rate noted after curettage of conventional solid and multicystic extraosseous ameloblastoma.[11,12] Lau et al[13] reported recurrence rates of 3.6% for resection, 30.5% for enucleation alone, 16% for enucleation followed by Carnoy's solution application and 18% by marsupialization followed by enucleation (where the lesion reduced in size).

Whatever surgical approach the surgeon decides to take, long-term follow-up is mandatory as recurrence of unicystic ameloblastoma may be long delayed.

CONCLUSION

Ameloblastomas in children differ from adults with a higher percentage of unicystic tumors. Unicystic ameloblastoma is a

tumor with a strong propensity for recurrence, especially when the ameloblastic focus penetrates the adjacent tissue from the wall of the cyst. Although enucleation has been claimed to give acceptable recurrence rates in unicystic ameloblastoma, there are no large series with long follow-up in children. The histologic pattern that exhibits mural invasion in unicystic ameloblastoma suggests that more aggressive surgery is necessary. Our study shows employing the application of Carnoy's solution alongwith the enucleation, which suggests a possible benefit against recurrence.

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