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Unicystic Ameloblastoma in 1 year Old Child – A Rare Case Report

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Abstract:
Ameloblastoma is a benign odontogenic neoplasm affecting the jaw bones. There are several variants of ameloblastoma based on the clinical,radiological and histopathological presentation. Unicystic ameloblastoma being the least encountered variant with good prognosis and less recurrence rate. The term 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 part of the cyst cavity, with or without luminal and/or mural tumour growth. It accounts for 10-15% of all intraosseous ameloblastomas. It occurs in 2nd and 3rd decades of life but only about 10% of cases are reported in children and less than one third of those occur in children below 10 years. Here we are presenting an extremely rare case of unicystic ameloblastoma in a one year old male patient.

Key Words*: unicystic ameloblastoma; benign tumour of oral cavity; odontogenic tumour*

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I. Introduction

Ameloblastomas are benign tumours having potential to grow into enormous size resulting in bone deformity. They can be classified in to unicystic, multicystic, peripheral and malignant subtypes[1]

Unicystic ameloblastoma , a variant of ameloblastoma was first described by Robinson and Martinez in 1977 and it refers to those cystic lesions that show clinical and radiologic characteristics of an odontogenic cyst but in histological examination show a typical ameloblastomatous epithelium lining part of the cyst cavity, with or without luminal and/or mural tumour proliferation [2]

The term 'Unicystic Ameloblastoma' was adopted in the second edition of the international histologic classification of odontogenic tumours. It constitutes 10-15% of all intraosseous ameloblastomas [3].This lesion occurs in a younger age group, with slightly more than 50% cases occurring in patients in their second decade of life[4].Unicystic ameloblastoma have a slight male predilection and frequently originate from the posterior region of mandible. Mandible is more affected than maxilla with a ratio of 13:1. Unicystic ameloblastomas are slow growing and relatively locally aggressive cystic lesions [5]. Radiographically, the lesions commonly present as expansive unilocular radiolucencies with a well demarcated border. Around 50-80% of cases are associated with an impacted or unerupted tooth [6]. Therefore, the clinical and radiographic presentations of unicystic ameloblastoma are at times indistinguishable from those of dentigerous cysts.[3,7,8,9]. Three histological types are identified according to the degree of ameloblastomatous epithelial extension, namely luminal, intraluminal and mural types [3].

Here we are describing about a mural type unicystic ameloblastoma in a 1 year old child.

II. Case Report

A 1 year old male patient reported to the Department of Oral and maxillofacial surgery, Government Dental College, Calicut,Kerala in January 2021, with complaint of swelling of lower left side of face. Parents noticed the swelling 2 weeks back and consulted a nearby private hospital. CT scan of the mandible was taken

from there and referred to our centre for further management. There was no history of fever, pus discharge or redness. Patient was active and playful but not co-operative .

On general examination, patient was moderately built and nourished. Patient had no relevant medical history and no known drug allergy. Physical examination revealed no abnormalities other than those pertaining to the present complaint. On extra oral examination, a diffuse swelling of size 3cm x 3 cm was seen on the left lower third of face, extending antero posteriorly from the mid-body of mandible till the angle of mandible and supero inferiorly from the zygomatic arch till the lower border of mandible. Overlying skin was normal in colour. There was no sinus discharge or fistula formation on the affected side. On palpation the swelling was non tender,firm in consistency and non compressible .There was no local rise in temperature and no evidence of lymphadenopathy.

Intra oral examination was performed on table during surgery as the patient was extremely un cooperative. On inspection, buccal cortical expansion noticed in relation to left mandibular posterior region,distal to erupted deciduous lower second molar with normal overlying mucosa. No mucosal ulceration was seen. On palpation there was palpable buccal and lingual cortical expansion extending from distal of deciduous lower second molar to ramus area on the left side. Egg shell crackling of buccal cortical wall noticed. There was no mobility or displacement of teeth in the vicinity.

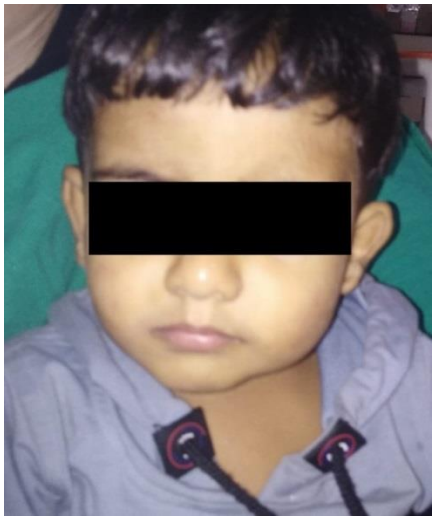

Figure 1: Extra-oral view

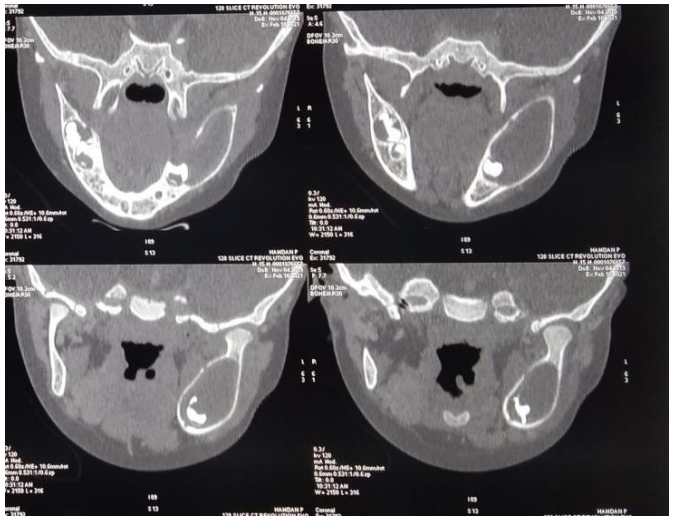

Figure 2: CT Coronal and axial cuts

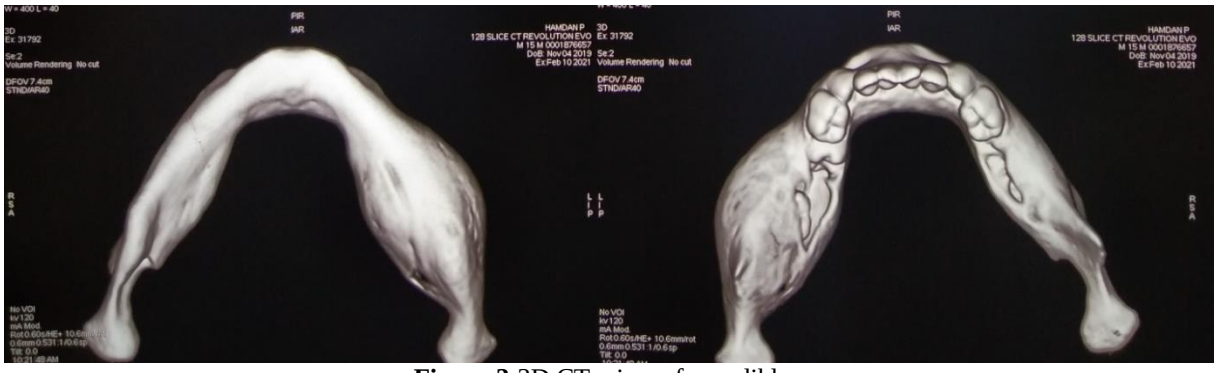

Figure 3:3D CT view of mandible

CT scan of the mandible shows an expansile unilocular lytic lesion of size 40mm x 24 mm at the region of ramus and posterior hemi mandible on the left side. Unerupted dysmorphic tooth bud seen within the lesion. A provisional diagnosis of odontogenic keratocyst was made. Differential diagnosis considered were dentigerous cyst and unicystic ameloblastoma.

Routine blood investigations were carried out. Surgical excision of the lesion along with chemical cauterization was planned under GA. Vestibular incision placed from canine region till anterior border of ramus and the muco-periosteal flap reflected. Buccal and lingual cortical expansion noticed on the left mandibular posterior body region. Buccal cortical plate was thinned out. A bony window created and enucleation of the entire lesion along with the embedded tooth bud was done, followed by peripheral ostectomy and chemical cauterization using Carnoy's solution. The excised specimen was sent for histopathological examination.

Surgical site was closed with poly-glactin sutures. Patient was treated with IV antibiotics and analgesics for 5 days.

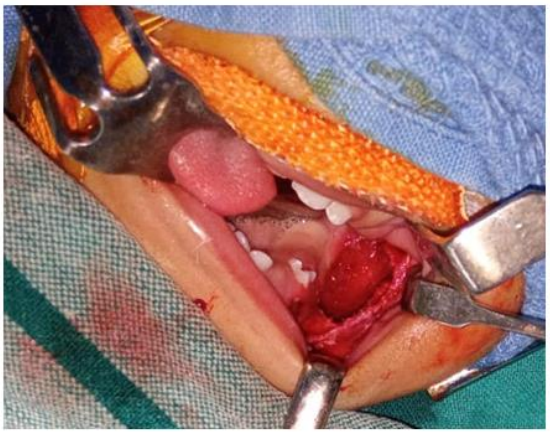

Figure 4: Intra-operative view

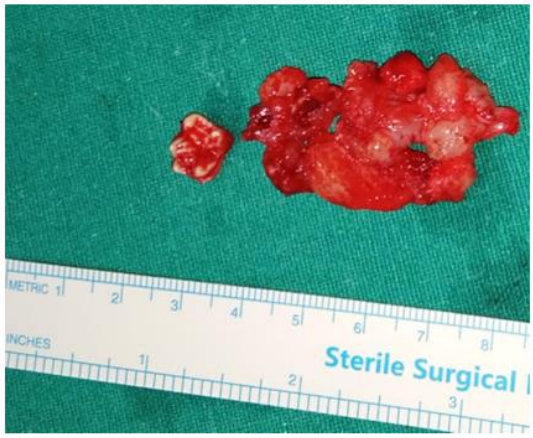

Figure 5: Excised specimen

Histopathological report suggested unicystic ameloblastoma with mural proliferation. Submitted bit of tissue is composed of cystic cavity lined by ameloblastomatous epithelium. Epithelium is proliferating in to the underlying connective tissue. Active odontogenic islands are also seen in the primitive mesenchyme like connective tissue. Diffuse inflammatory cell infiltrate intermingled with endothelium lined vascular spaces filled with RBCs and areas of hemorrhage also seen.

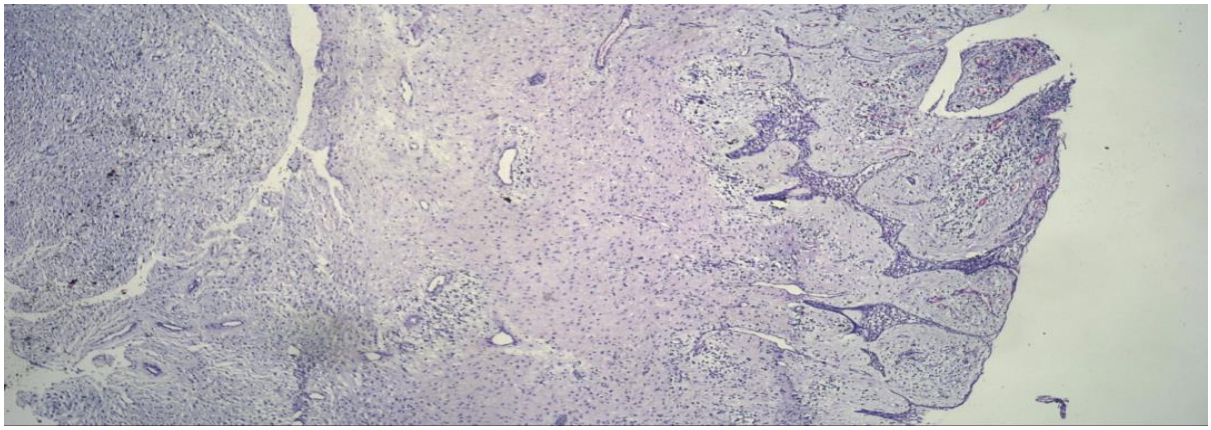

Figure 6: Histopathology showing cystic cavity lined by ameloblastomatous epithelium proliferating in to the underlying connective tissue

III. Discussion

Ameloblastoma is a benign, locally aggressive and infiltrative odontogenic neoplasm with rare metastatic capacity which constitutes only 1.3% of all jaw cysts and tumours and the 2nd most common odontogenic neoplasm comprising 10 % of neoplasm of odontogenic origin.[10]

As per the WHO system of 2003, ameloblastoma is classified based on the differences in biologic behavior, treatment plan and recurrence rate as: (1) classic solid/multicystic ameloblastoma, (2) unicystic ameloblastoma, (3) peripheral ameloblastoma (4) desmoplastic ameloblastoma, including the so-called hybrid lesions.[4]

Unicystic ameloblastoma, a variant of ameloblastoma was first described by Robinson and Martinez [11] in 1977. It refers to those cystic lesions that show clinical and radiological 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 tumour proliferation. This variant is believed to be less aggressive, usually manifests in a younger age group, with about 50% of the cases occurring in the second decade of life. More than 90% of cases are located in the mandible[12]. Ameloblastoma is uncommon in children. The most commonly quoted article regarding ameloblastoma is a review of 1,036 ameloblastomas in which the average patient age is 38.9 years, with only 2.2% (19 of 858) less than 10 years old and 8.7% (75 of 858) between 10 and 19 years

old[13]. But this report, was published in 1955, when adenoameloblastomas and ameloblastic fibromas were included as ameloblastomas and before the histologic characteristics for ameloblastomas in cysts had been delineated[14]. Reports of unicystic ameloblastoma occurring in first decade of life, especially below five years of age are very limited.

Leider et al suggested three pathogenic mechanisms of evolution of Unicystic Ameloblastoma:

1. Reduced enamel epithelium associated with a developing tooth undergoes ameloblastic transformation with subsequent cystic development.
2. Ameloblastomas arise in dentigerous cyst or other types of odontogenic cysts in which the neoplastic ameloblastic epithelium is preceded temporarily by non-neoplastic stratified squamous epithelial lining.
3. Solid ameloblastoma undergoes cystic degeneration of ameloblastic islands with subsequent fusion of multiple microcysts and develops into a unicystic lesion[15]

Unicystic Ameloblastoma is characterized by one or more of the following features of Ackermann criteria.

Ackermann criteria:

Group 1: Luminal type (tumour confined to luminal surface of the cyst)

Group 2: Intra luminal type (nodular proliferation of the lumen without infiltration of tumour cells into the connective tissue wall)

Group 3: Mural type (invasive islands of ameloblastomatous epithelium in the connective tissue wall without involving the entire epithelium)[16]

Our case belonged to the third group - mural type. The treatment of Unicystic Ameloblastoma can be radical or conservative and this topic has always been controversial[17]. Radical treatment includes segmental or marginal resection of the lesion followed by placement of reconstruction plates whenever required. On the other hand, conservative treatment constitutes enucleation with or without curettage and marsupialization followed by enucleation. Lau and Samman studied the recurrence rate of UA following various treatment modalities and observed the recurrence rate was 30.5% (highest) following enucleation alone, 18% for marsupialization, 16% for enucleation followed by application of Carnoy's solution and 3.6% (lowest) for resection[18,19,20]. Though lower recurrence rate is reported following resection, radical treatment is usually avoided in children for the following reasons:

1. Continuing facial growth in children and the presence of a highly reactive periosteum
2. The presence of unerupted permanent teeth
3. May cause disfigurement and masticatory problems which can be psychologically disturbing to the child.

A conservative line of treatment has an excellent role in pediatric and adolescent patients since it is associated with faster bone fill and restoration of normal bony architecture[21].Considering the age of our patient, we planned a conservative management comprising of enucleation,peripheral ostectomy and chemical cauterization.

Unicystic Ameloblastoma is treated conservatively with decompression, enucleation and peripheral ostectomy and periodic long-term follow up. A more aggressive surgical approach may be considered when the lesion recurs more than twice or by the patient's wish. Whatever treatment approach the surgeon decides to take, a long-term follow-up is compulsory as the recurrence of unicystic ameloblastoma may be long delayed[22].

IV. Conclusion

Eventhough ameloblastoma is a benign tumour with high recurrence rate, unicystic variant is considered to have better prognosis and less recurrence rate. But the mural variety of unicystic ameloblastoma has a strong tendency to recur, especially when the ameloblastic focus penetrates the adjacent tissue from the wall of the cyst. The diagnosis of Unicystic Ameloblastoma in children is difficult because most of the lesions radiographically resemble dentigerous cyst as 70 – 80% of cases are presented with unerupted tooth. The treatment plan in pediatric population remains controversial as whether to go for a conservative or radical approach. Literature reviews are very scarce on this topic. Considering the quality of life in pediatric population, a conservative approach is more promising. However regular and strict long term follow ups are needed. Further studies need to be conducted regarding the treatment protocol for pediatric ameloblastomas.

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