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Cystic ameloblastoma in young girl of 18 years old: a non-dentigerous variant

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Abstract

Ameloblastoma traditionally occurs in the 3rd and 4th decade, usually in the body of mandible (molar region) but in recent literature their occurrence in young individuals have been reported. It is a benign locally invasive epithelial odontogenic tumour comprising 1% of all tumours and cysts arising in the jaw. In this report, we present a case of unicystic ameloblastoma (non dentigerous variant) in a young girl of 18 years. A detailed history, clinical, radiographic features and histopathology and treatment of the case is discussed here. It highlights the variations in presenting features especially the young age of the Indian girl.

Keywords: Unicystic, Ameloblastoma, Benign tumor, Young age.

Introduction

Ameloblastoma though infrequently seen, is a benign locally invasive epithelial odontogenic tumour comprising 1% of all tumours and cysts arising in the jaw.(1) It is the most common type of odontogenic tumor accounting for 1% of all tumours in head and neck region and approximately 11% of odontogenic tumors.(2) It is a persistent and locally invasive tumor that has aggressive but benign growth characteristics. There are different clinico-radiographic types: the conventional solid/multicystic ameloblastoma, the unicystic ameloblastoma and the peripheral ameloblastoma.(3) Histopathologically ameloblastoma can be classified as follicular, acanthomatous, granular, basal, desmoplastic and plexiform.(3)

Unicystic ameloblastomas generally resemble dentigerous cyst (dentigerous variant) clinically and radiographically, few are not associated with unerupted teeth (non-dentigerous variant).(3) They typically occur more often in younger patients.(3) Radiographically they appear as unilocular or multilocular.(4) Both forms have been shown to recur, particularly following inadequate surgical treatment. The periphery may be smooth or scalloped. The present case is a non-dentigerous variant of unicystic ameloblastoma seen in ramus area of mandible with large area of extension and morbidity.

Case Report

A 18 year old girl reported with a complaint of painless swelling in the lower right back jaw since 1.5 years which was insidious in onset and gradually progressed to the present size along with mild pain since 2 months.

On extraoral examination, a diffuse swelling of 4cm was seen in the pre-auricular region, on the right side of the face extending upto the angle of mandible (Fig. 1). On palpation, the swelling was soft to firm in consistency, tender, with slight elevated temperature. No discharge fluctuation or crepitation was seen and the right submandibular lymph node was palpable and movable. Mouth opening was normal.

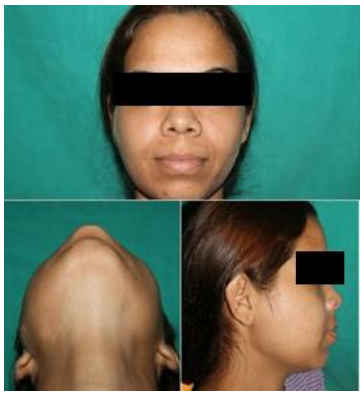

Fig. 1: Extra-oral view of the patient showing the right posterior region of mandible with a diffuse swelling

Intra-oral examination revealed a diffuse swelling in relation to 46, 47and 48. The surface appeared lobulated, slightly erythematous with ill-defined margins and extended into the retromolar area(Fig. 2). On palpation the swelling was tender, soft in consistency, with expansion of the buccal cortical bone and the ascending ramus. 47 and 48 showed slight mobility. Crepitation was felt on palpation.

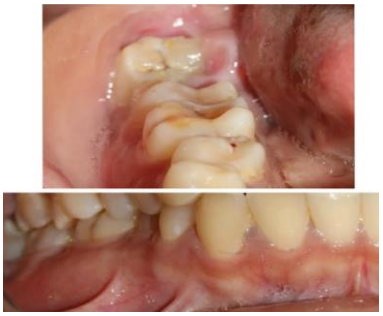

Fig. 2: Diffuse swelling seen intra-orally extending from 46 to 48

Considering the clinical findings, it was thought to be either a benign odontogenic tumour or a dentigerous cyst.

The patient was subjected to radiographic and routine haematological examination. Hematological findings were not significant.

OPG shows a well-defined large radiolucency extending from mesial of 47, involving the ramus and extending till the coronoid process. The radiolucency is well-corticated with no internal septae and few scalloped margins. Diffuse area of altered trabeculae is seen in the body of mandible mesial to the radiolucency. Inferior alveolar canal is displaced inferiorly and external root resorption is seen in 43, 44, 45, 46, 47, 48. (Fig. 3)

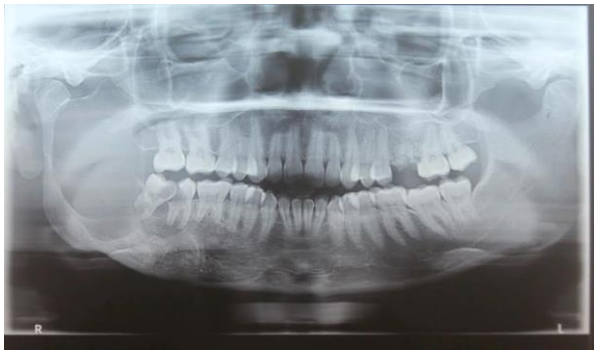

Fig. 3: Orthopantomogram

Axial CT sections show an expansile, hypodense, osteolytic lesion involving the right premolar region, extending to the ramus .Buccal cortical perforation seen in the body of mandible and lower part of ramus shows large perforations buccally and lingually. Masseter muscle however appears normal (Fig. 4). Coronal CT sections show an osteolytic lesion causing medio-lateral enlargement of ramus along with multiple sites of perforation sparing only the condylar head.(Fig. 5)

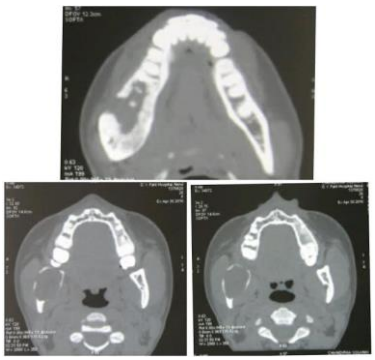

Fig. 4: Axial CT sections

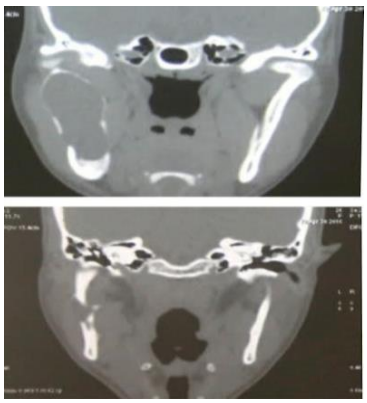

Fig. 5: Coronal CT sections

Based on the radiographic features of cortical expansion, perforation, large area of involvement and root resorption and lack of septae, a diagnosis of unilocular/unicystic ameloblastoma was made.

After incisional biopsy, hemimandibulectomy was carried out with reconstruction using free fibula graft (Fig. 6).

The histopathologic sections showed cystic odontogenic epithelial lining consisting of basal layer of cuboidal to columnar ameloblast like cells with hyperchromatic nuclei, reverse polarity and basillar cytoplasmic vacuolization (Vicker's Gorlin Criteria).

Fibrous walls of cyst is infiltrated by follicles of odontogenic epithelium suggestive of unicystic ameloblastoma with mural proliferation (Subtype 1.3)(Fig. 6).

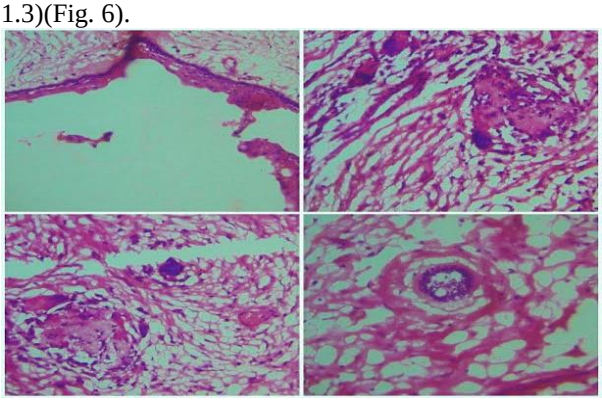

Fig. 6: 10X and 40X magnification microscopy showing luminal and mural proliferation

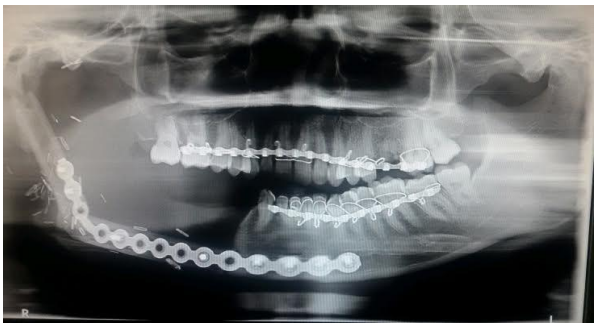

Fig. 7: OPG showing reconstruction plates along with arch bar wiring

Discussion

Conventional ameloblastomas are usually seen between the age of 20-50 years. Studies have shown that unicystic variant may occur in young patients between 20-30 years. We have encountered this variety of ameloblastoma in a girl of 18 years who had a history of swelling since 1.5 years which highlights that this condition is not limited to 3-4th decade as was seen earlier. Rekha K et al, reported that ameloblastoma in young patients under 19 years was seen in 21.9% of their study group.[2] Reichart et al has reported that ameloblastomas tend to occur at a young age in developing countries and attributed this to the accelerated aging process due to poor nutrition and health care .

Unicystic ameloblastoma is a rare type of ameloblastoma, accounting for 6% of all ameloblastomas.[5] It usually occurs in the younger age group, with about 50% cases occurring in second decade of life.[5,6] More than 90% are located in the mandible.[5,6,7,8] Patients commonly present with swelling and facial asymmetry, pain being present occasionally.[5] Mucosal ulceration is rare, but may be caused by continued growth of tumor.[5] Small lesions are sometimes discovered more on routine examinations or as result of local effects, like tooth mobility, occlusal alterations or failure of eruption.[5]

Mandibular 3rd molar is mostly associated with impacted tooth in unicystic ameloblastoma called dentigerous variant[9] but a few cases are not associated with impacted teeth which are called non-dentigerous variant.[10] According to Konouchi H et al, mean age of non-impacted tooth related cystic ameloblastoma was 35 years in comparison to 16.5 years for the impacted tooth related variant.[11] However, here in this case the non-dentigerous variant was seen at the age of 18 years. According to Rekha et al,[2] unilocular ameloblastoma is seen in younger patients along with cortical expansion of bone. Both these features are predominant in females. Ameloblastoma can be diagnosed radiographically based on their locularity, root resorption and cortical expansion.[12]

Unicystic ameloblastoma is believed to be less aggressive and responds favourably to conservative treatment than the solid or multicystic ameloblastoma.[13,6,14,15]

Ackermann classified unicystic ameloblastoma into three histologic groups:

Luminal unicystic ameloblastoma(tumor confined to the luminal surface of cyst)

- Intra-luminal unicystic ameloblastoma (nodular proliferation into lumen without infilteration of tumor cells into connective tissue wall)
- Mural unicysticameloblastoma (Invasive islands of ameloblastomatous epithelium in the connective tissue wall not involving entire epithelium)
- Another histologic subgrouping is by Philipsen and Reichartet al:[7]
- Subgroup 1: Luminal unicystic ameloblastoma
- Subgroup 1.2: Luminal and intraluminal
- Subgroup 1.2.3: Luminal, intraluminal & intramural
- Subgroup 1.3: Luminal and intramural

According to Philipsen and Reichart:

Unicystic ameloblastoma diagnosed as subgroups 1 and 1.2 can be treated conservatively whereas 1.2.3 and 1.3 showing intramural growth requires radical resection.[7] Recurrence rates vary in each type. The average interval of recurrence is 7years.[16] The unicystic ameloblastoma invading fibrous wall have a recurrence rate of 35.7%, others only have 6.7%.[16] The recurrence rate for resection is 3.6%, for enucleation alone is 30.5%, 16% for enucleation followed by carnoy's solution application and 18% by marsuplisation.[17]

Since the present case was diagnosed as subtype 1.3, and taking into consideration the amount of morbidity, hemimandibulectomy was carried out with free fibula graft placement (Fig. 7).

Patient was recalled after 6months and did not show any signs of recurrence. Patient is kept under 6month follow up for the next 5years.

Conclusion

Unicystic ameloblastoma in younger age group, is located predominantly in posterior mandible along with wide age difference between dentigerous and non-dentigerous type and predominance of unilocular ameloblastoma over multilocular ameloblastoma has been reported in literature. But in this case the non dentigerous variant occurred in a female of 18 years who complained of swelling since 1.5 years which indicates that probably the lesion should have initiated at a tender age of 16 years. This definitely is an alarming situation in developing countries like India.

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