Case Report

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Pediatric Odontogenic Tumor of the Jaw – A Case Report

Dentistry Section

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ABSTRACT

Central jaw tumors (intra osseous) in children occur infrequently and few oral pathologists have had the opportunity or experience in diagnosing these lesions and predicting their biological behavior. Some children are not diagnosed correctly at the initial stages as having a neoplasm and are wrongly treated for infections by antibiotic administration. Subsequent to an unresponsive antibiotic therapy radiographs are taken to reveal a radiolucent or radio dense lesion in the jaws. Finally a tissue diagnosis becomes necessary in order to diagnose and initiate proper therapy. One among the central jaw tumors that occur infrequently in children is Ameloblastoma. It is often aggressive and destructive, with the capacity to attain great size, erode bone and invade adjacent structures. Ameloblastoma not only accounts for 1% of all tumors of maxilla and mandible but also 11% of all odontogenic tumors. It has a high percentage of local recurrence rate and possible malignant development when treated inadequately. Here we present a central jaw tumor in an 8-year-old child which was a case of unusually large plexiform ameloblastoma involving entire ramus up to the condyle, and part of body of the mandible.

Keywords: Plexiform ameloblastoma, Odontogenic tumor, Mandible

INTRODUCTION

Pediatric central jaw tumors are broadly classified into odontogenic and non-odontogenic groups among which odontogenic tumors are considered to be common in children. Tumors and tumor-like growths arising from odontogenic tissues constitute a heterogeneous group of interesting lesions, as they display the various inductive interactions that normally occur among the embryologic components of the developing tooth germ [1]. One among the central jaw odontogenic tumors that occur infrequently in children is Ameloblastoma. It is often aggressive and destructive, with the capacity to attain great size, erode bone and invade adjacent structures. It has a high percentage of local recurrence rate and possible malignant development when treated inadequately [2].

Ameloblastoma is a true neoplasm of enamel organ type tissue which does not undergo differentiation to the point of enamel formation [3]. It was described by Robinson (1937) as a benign tumor that is "usually unicentric, non-functional, intermittent in growth, anatomically benign and clinically persistent". The World Health Organization (1991) defined ameloblastoma as a benign but locally aggressive tumor with a high tendency to recur, consisting of proliferating odontogenic epithelium lying in a fibrous stroma [4]. The mean age of patients at the time of diagnosis for Ameloblatoma was 35.9 years with a range of 4 to 92 years [5]. Most ameloblastomas develop in the molar-ramus region of the mandible with 70% of these arising in the molar-ramus area [6]. It is located centrally or intraosseously in both jaws, and there are few or no clinical signs in the early stages. Later there is gradually increasing facial deformity, teeth in the area may become loose, and spontaneous fracture may occur in cases where only a rim of normal bone forms the base of the mandible. The affected part of the jaw is bony hard and bulky. Pain occurs with varying, often quite low, frequency. It is well-known that ameloblastoma can be radiologically unilocular or multilocular radiolucency with a honeycomb or soap bubble appearance [7].

Ameloblastoma histological appearance is similar to that of the early cap-stage ameloblastic elements of developing without complete differentiation to stage of enamel formation [8]. Six histological subtypes of ameloblastoma have been identified-follicular, plexiform, acanthomatous, granular, basal cell and desmoplastic type. Even it frequently recurs after inadequate surgical treatment, ameloblastomas infrequently metastasize [9]. Here we present a case of plexiform ameloblastoma in an eight year old child along with clinical, radiographical and histopathological features discussed.

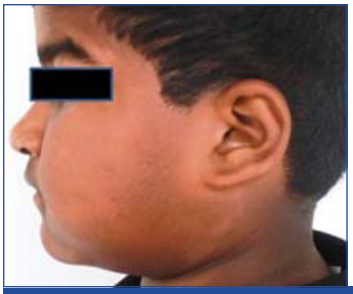
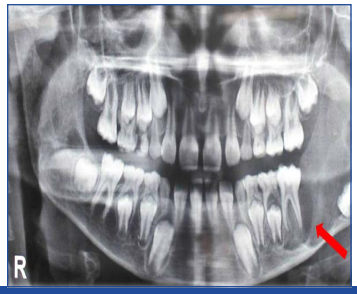
CASE REPORT

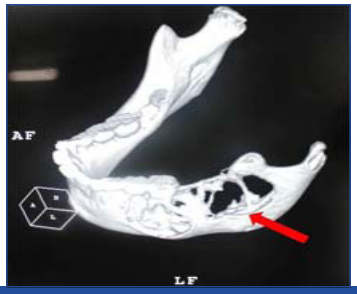
An eight-year-old male patient presented with a chief complaint of swelling on the left side of the face since one month. Extra oral examination of the patient was notable for facial asymmetry and a firm swelling was present in the area of the left mandibular body region extending up to the base of the mandible. Clinical examination revealed diffuse, smooth surfaced, hard swelling measuring about 5x6 cm in size on left side of the face. It extended from the zygomatic region to the inferior border of mandible superio-inferiorly, and from the corner of mouth to the angle of mandible anterio-posteriorly [Table/Fig-1]. Left submandibular lymph nodes were palpable. Intra orally obliteration of left vestibule in the deciduous second molar and permanent first molar region was present and the region was slightly tender and the involved teeth had grade I mobility.

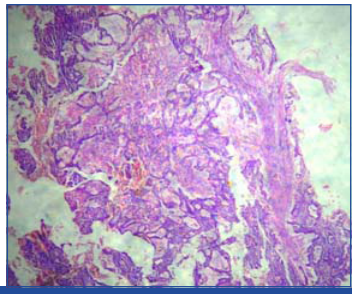
Panoramic radiography (OPG) showed a large multilocular radiolucent area extending from the left deciduous second molar (75) region to the neck of condylar process and the coronoid process including the ascending ramus area on the left side [Table/Fig-2]. The patient was advised CT scan where it revealed an extensive osteolytic lesion involving both bucco-lingual cortical plates with complete perforation of the cortical bone and infiltration of the tumor mass into the surrounding soft tissue [Table/Fig-3]. Based on the radiographic investigations a provisional diagnosis of ameloblastoma/carcinoma of the mandible were made. An incisional biopsy was made and was sent for histopathological examination.

The histopathological processing of the tumor revealed odontogenic epithelium arranged as a tangled network of anastomosing strands [Table/Fig-4]. The cords or sheets of epithelium are bounded by columnar or cuboidal ameloblast like cells surrounding more loosely arranged stellate reticulum like cells in the centre [Table/Fig-5]. The supporting stroma is loosely arranged and vascular with few areas showing stromal degeneration [Table/Fig-6]. Based on the histopathological findings the pathological report confirmed the diagnosis of a plexiform ameloblastoma and the patient was scheduled for surgical resection of the involved mandible.

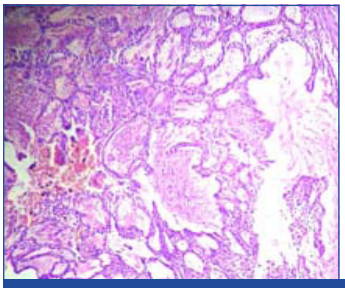
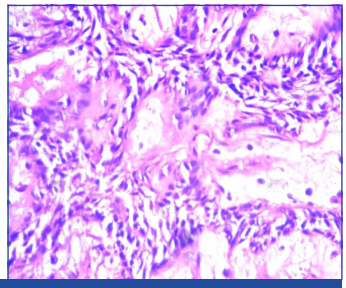
Surgical resection of the tumor was carried out through an extra oral submandibular approach. Mandibulectomy was performed by maintaining a safe margin of 1.5 cm of uninvolved bone and temporary maxilla-mandibular fixation was done. Contra-lateral fifth and sixth ribs were harvested as a costochondral graft through inframammary incision and secured into place by means of

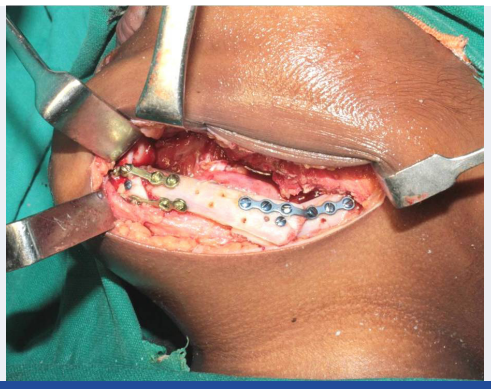




[Table/Fig-1]: Facial asymmetry and a firm swelling in the left mandibular ramus region
[Table/Fig-2]: OPG showing multilocular radiolucency extending fromthe deciduous second molar to the neck of condylar process and the coronoid process
[Table/Fig-3]: CT scan showing extensive osteolytic lesion involving both bucco-lingual cortical plates with complete perforation of the cortical bone
[Table/Fig-4]: H & E stained section shows odontogenic epithelium arranged as a tangled network of anastomosing strands (4X)

[Table/Fig-5]: H & E stained section shows cords or sheets of odontogenic epithelium bounded by columnar or cuboidal ameloblast like cells surrounding more loosely arranged stellate reticulum like cells in the centre (10X)
[Table/Fig-6]: H & E stained section showing odontogenic epithelium arranged in the form of strands along with stromal degeneration in few areas (40X)



[Table/Fig-7]: Surgical resection of left mandible and harvested by fifth and sixth costochondral graft in continuation with the distal end of resected mandible

[Table/Fig-8]: OPG to ensure the stability of the graft

reconstruction plate [Table/Fig-7,8]. The patient was recalled after 15 days of surgery for post operative follow up and OPG was taken to ensure the stability of the graft [Table/Fig-8].

DISCUSSION

Ameloblastomas are aggressive benign tumors of epithelial origin that may arise from the enamel organ, remnants of dental lamina, the lining of an odontogenic (dentigerous) cyst, or possibly from the basal epithelial cells of the oral mucosa [10]. They are usually common in adults and infrequent in children. In a review of 1,036 ameloblastomas of jaw by White SC and Pharoah MJ, the average patient age was 38.9 years, with only 2.2% (19 of 858) were under 10 years and 8.7% (75 of 858) were between 10 and19 years [7]. Typical ameloblastoma starts insidiously as a central bony lesion which is slowly destructive; however tends to expand the bone instead of punching a hole through it [11]. Ameloblastoma appears equal frequency between sexes although a higher frequency in males than in females has been described [8].

Eighty percent of ameloblastomas occur in the mandible almost exclusively in the molar ramus region of mandible and are often associated with an unerupted tooth. The remaining 20% occur in the maxilla with maxillary tuberosity being the most common site. [5]. Three types of intraosseous ameloblastoma are currently agreed upon: the solid-multicystic or conventional variant, the unicystic variant, and the desmoplastic variant. The solid-multicystic variant is the most common, comprising 86% of ameloblastoma and it has a tendency to be more aggressive than the other types and has a higher incidence of recurrence. Sometimes ameloblastomas continue to enlarge and may cause the surrounding bone to become so thin that crepitating or eggshell crackling may be elicited [8].

Clinically, ameloblastoma frequently manifests as a painless swelling, which can be accompanied by facial deformity, malocclusion, and loss of dental pieces, ulceration and periodontal disease. Sometimes pain occurs with varying intensities, but often quite low. It is not known whether the cause of the pain is pressure from the tumor on peripheral nerves or secondary infection [8].

In most cases ameloblastoma present a characteristic but not diagnostic radiographic appearance. It may present in three different patterns where the most common form is the multilocular with various cysts that are in groups or separated by osseous reinforced septa (soap bubble appearance). Another image is a beehive pattern, this being the second most common type. A third radiographic manifestation, which is very important in terms of a differential diagnosis, is the unilocular form [10].

Ameloblastoma is a polymorphic neoplasm consisting of proliferating odontogenic epithelium, which usually has a follicular (32.5%) or plexiform pattern (28%), lying in a fibrous strorma. Other histological types are acanthomatous, papilliferous-keratotic type, desmoplastic, granular cell type, clear cell type, vascular type, basal cell type and mucous cell differentiation type [8].

The term plexiform refers to the appearance of anastomosing islands of odontogenic epithelium in contrast to a follicular pattern. [12] Tumor epithelium is arranged as a network which is bound by a layer of cuboidal to columnar cells and includes cells resembling stellate reticulum. Cyst formation occurs but is usually due to stromal degeneration rather than to a cystic change within the epithelium [8]. The common denominator to all ameloblastomas is well-differentiated palisaded cells found around the periphery of nests, strands and networks of epithelium. Nuclei of the palisaded cells are typically polarized away from the basement membrane [10].

GROWTH POTENTIAL AND BEHAVIOR

Ameloblastomas may show various biologic behaviors, ranging from cystic expansion to more aggressive infiltration of adjacent tissue. Unlike carcinomas, ameloblastomas are circumferentially

delineated by a continuous basement membrane, and they tend to spread into tissue spaces by expanding their compartment volumes. The architectural pattern of the ameloblastoma is such that the border of the tumor within cancellous bone lies beyond the apparent macroscopic surface and the radiographic boundaries of the lesion [13]. Hong et al., recently showed that the histopathology of an ameloblastoma is significantly associated with a recurrence. It was shown that the follicular, granular cell and acanthomatous types have a relatively high likelihood of recurrence. In contrast, the plexiform, desmoplastic and unicystic types show a relatively low potential for recurrence [14].

IMMUNOHISTOCHEMISTRY

Proliferative marker Ki 67 positive nuclei in the ameloblastoma are mainly located in peripheral ameloblast-like cells in the follicular as well as in the plexiform areas of the solid ameloblastomas. This staining pattern indicates that the cellular proliferation and consequently the ameloblastoma growth are concentrated in the peripheral areas composed by ameloblast-like cells [15]. Kumamoto et al., demonstrated that telomerase activity, which is linked to cell immortalization, is associated with the proliferative potential of ameloblastoma cells. According to these authors, the telomerase activity detected in ameloblastoma reflects tumor characteristics such as ability of local invasion and high recurrence rates [16]. Apoptotic markers like caspase 3 and Bcl2 are also expressed in ameloblastomas [17]. Ameloblastomas shows higher expression of parathyroid hormone-related protein (PTHrP), osteoclast differentiation factor (ODF). Because of these markers ameloblastomas shows higher invasiveness into surrounding bone via osteoclastic bone destruction, despite its benign nature [18].

TREATMENT

Treatment of ameloblastomas is primarily surgical. There has been some debate regarding the most appropriate method for surgical removal of ameloblastomas. These range from conservative to radical modes of treatment. The conservative modalities include curettage, enucleation and cryosurgery; while the radical modalities are marginal, segmental and composite resections. The recommended treatment for ameloblastoma in children should be radical resection, 0.5 to 1 cm past what appears to be normal bone [10]. Recurrence seems to depend on several factors such as method of treatment of primary lesion, extent of lesion and the site of origin. Recurrence rates also vary for different procedures used to treat primary lesion, several authors have found a recurrence rate 55 to 90% for all ameloblastomas treated with conservatively (enucleation and curettage). However, the incidence of recurrence following radical resection is 5 to 15%. Radiation therapy alone is not warranted, because ameloblastoma is considered to be a radio resistant tumor. Cryotherapy after conservative treatment showed reduced recurrence. There may be less morbidity with an initial radical surgery than multiple repeated conservative therapies with recurrence. In the present case the patient was treated by radical resection of the mandible with 1 to 1.5 cm clear margins and harvested by fifth and sixth costochondral grafts [11].

In the present case, the patient was eight-year-old child with the clinical, radiographical and histopathological findings diagnostic of a plexiform ameloblastoma and the patient was scheduled for surgical resection of the involved mandible with 1.5 cm clear margins and harvested by fifth and sixth costochondral grafts.

CONCLUSION

Though ameloblastoma is a benign tumor it is aggressive, expansile, shows positivity for proliferative markers and osteolytic markers which are responsible for aggressive behavior and higher invasiveness into surrounding bone. This case report illustrates the importance of adequate radical resection to avoid recurrence. All possible efforts must be taken to evaluate the extent of the tumor to adjacent structures before surgery, to avoid recurrence or metastasis in future. Ameloblastoma may recur even after fifteen years or more. Therefore, it is important to keep the patient under long term follow-up.

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