Less is more: The conservative management of Pediatric Ameloblastoma - A case report

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Συντηρητική Αντιμετώπιση Παιδιατρικού Αδαμαντινοβλαστώματος. Αναφορά περίπτωσης

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Case Report
Αναφορά περιστατικού

SUMMARY: Ameloblastoma is a benign tumor of the jaws of odontogenic origin, known for its aggressive behavior involving adjacent vital structures. Though the tumor predominantly involves the adolescents in their second and adults in third decades of life, it is very rare below 10 years of age. The treatment of ameloblastoma is controversial as the surgeon is left to choose between a conservative, which results in recurrence or an aggressive approach, which causes the resection of vital structures in close proximity. Surgical treatment of ameloblastoma in children should follow the principles of craniofacial growth due to the incomplete growth of the jaws and the pathological aspects of the tumor. In this paper, we discuss a rare case of plexiform ameloblastoma in a 9 year old child treated conservatively using total enucleation, chemical cauterization followed by extraction of involved tooth, with a good follow up prognosis of the case.

KEY WORDS: *Ameloblastoma, Conservative management, enucleation, pediatric, recurrence*

ΠΕΡΙΛΗΨΗ: Το αδαμαντίνοβλάστωμα είναι ένας καλοήθης όγκος των γνάθων οδοντογενούς προέλευσης, γνωστός για την επιθετική του συμπεριφορά που περιλαμβάνει παρακείμενες ζωτικές δομές. Αν και ο όγκος αφορά κυρίως τους εφήβους στη δεύτερη και τους ενήλικες στην τρίτη δεκαετία της ζωής τους, είναι πολύ σπάνιος κάτω των 10 ετών. Η θεραπεία του ααδαμαντίνοβλαστώματος είναι αμφιλεγόμενη καθώς ο χειρουργός μπορεί να επιλέξει μεταξύ μιας συντηρητικής, η οποία οδηγεί σε υποτροπή ή μιας επιθετικής προσέγγισης, η οποία προκαλεί την εκτομή ζωτικών εγγύς ανατομικών δομών. Η χειρουργική θεραπεία του αδαμαντίνοβλαστώματος στα παιδιά θα πρέπει να ακολουθεί τις αρχές της κρανιοπροσωπικής ανάπτυξης λόγω της ατελούς ανάπτυξης των γνάθων και των παθολογικών πλευρών του όγκου. Σε αυτή την εργασία, συζητάμε μια σπάνια περίπτωση πλεγματοειδούς αδαμαντίνοβλαστώματος σε παιδί 9 ετών που υποβλήθηκε σε συντηρητική θεραπεία με ολική εκπυρήνιση, χημική καυτηρίαση ακολουθούμενη από εξαγωγή εμπλεκόμενου δοντιού, με καλή πρόγνωση του περιστατικού στη διάρκεια της παρακολούθησης.

ΛΕΞΕΙΣ ΚΛΕΙΔΙΑ: *Αδαμαντίνοβλάστωμα, Συντηρητική αντιμετώπιση, εκπυρήνιση, παιδιατρική, υποτροπή*

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INTRODUCTION

Odontogenic tumors accounts for 1% of tumors occurring in the oral cavity, with 13%–58% of them being of odontogenic origin (1). Ameloblastoma, a benign tumor of odontogenic origin, is notorious for its highly invasive, infiltrative and destructive growth pattern that usually extends beyond the clinical margins, often yielding a high recurrence rate. They originate from the epithelial rests/ remnants of Hertwig's epithelial root sheath and/ or remnants of the dental lamina.

It is predominantly a tumor of the 3rd to 4th decades of life, the incidence of ameloblastoma is very rare especially under the age of 10, with only less than 10% of the cases diagnosed (2,3). The surgical management of this tumor depends on age of the patient, tumor location, size or histologic variant (4, 5). The decision is split between employing a radical approach or a conservative method. Though radical approach prevents any recurrence6 it can cause surgical deformity during reconstruction which profoundly impacts the craniofacial growth and exerts a heavy toll on the psychological state of the child. Here, we present a rare case of ameloblastoma in a 9 year old patient treated conservatively with one year follow up and emphasize on the importance of considering conservative management when treating pediatric ameloblastoma in patients younger than 10 years of age.

CASE REPORT

A 9 year old male patient reported to our centre with a chief complaint of swelling involving the left side of his lower jaw. The patient had noticed a mild swelling involving the left side of his lower jaw, which gradually increased in size. The swelling along with pain had started a week ago for which the patient had sought treatment elsewhere, where he was advised to undergo resection and reconstruction of the mandible, but the patient was reluctant to accept the surgical option. There was no other contributory history. On extra oral examination, Mouth opening was found to be adequate. Facial asymmetry was evident corresponding to the left side of the lower jaw from the body of mandible extending towards the ear lobule. There was a solitary ill-defined diffuse swelling over the lower third of the face measuring about 5×8 cm extending inferiorly from the left pretragal region to the lower border of the mandible. The surface appeared to be smooth, and was of normal colour. Patient had no signs of paresthesia. It was non-tender and hard on palpation (Figure 1). Intraorally, there was expansion of the buccal and lingual cortex starting at the first molar region and extending along the body of the mandible. There was no pathologic fracture and no perforation of mucosa. Grade I mobility of 36 was seen. Panoramic radiograph revealed a radiolucent lesion approximately 3.5 to 4cm antero-posteriorly in size involving the distal root of 36 extending till the ramus of the mandible. The tooth bud of second permanent molar was seen to be contained within the radiolucency. There was another tooth bud pushed to the coronoid process .To analyse the extent of the lesion, CBCT Scan (Cone Beam Computed Tomography) was taken. The axial section of the jaw revealed a large well-defined radiolucent lesion in the left body and ramus of the mandible with multilocular appearance causing expansion of the body and ramus (Figure 2, 3, 4). Considering the history, clinical features and radiological findings, a provisional diagnosis of aggressive benign cyst of the jaw was made. Taking into account, the young age of the patient, total enucleation of the cyst, fixation with Carnoy's solution with or without reconstruction plate fixation was the treatment planned. After preoperative evaluation and fitness certificate obtained from a general physician patient was taken up for surgery.

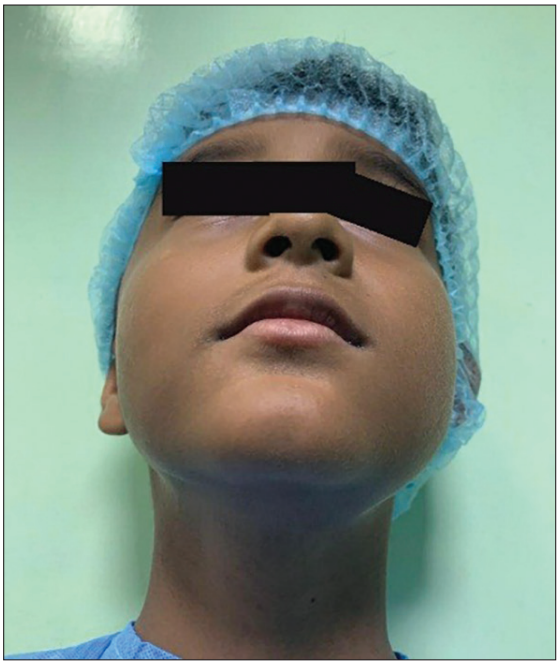

Fig. 1: Pre operative clinical photograph.

Under nasotracheal intubation, general anesthesia (GA) was administered. Care was taken by the anesthetist while using laryngoscope not to apply excess force on the mandible which may cause pathologic fracture. Incision was made from 74 with anterior release along the ascending ramus extending till the coronoid process. Periosteal elevation of the flap was done meticulously to reveal the buccal and lingual cortex of the mandible. The buccal and lingual cortex was found to be extremely thin and highly expanded. Mild perforation was noted in the ramus region. Aspiration was done with a wide bore needle following which blood aspirate was obtained.The

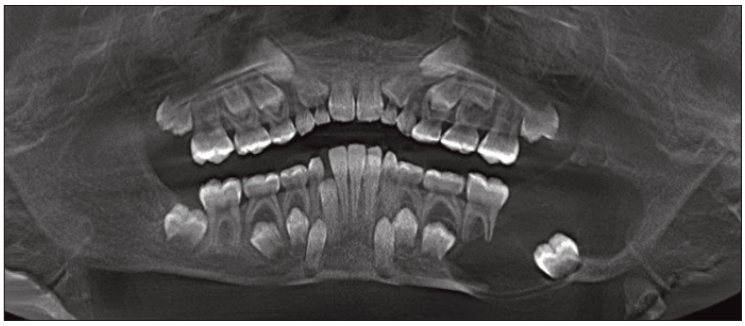

Fig. 2: Panoramic radiograph revealed a radiolucent lesion approximately 3.5 to 4cm in size involving the distal root of 36 extending till the ramus of the mandible.

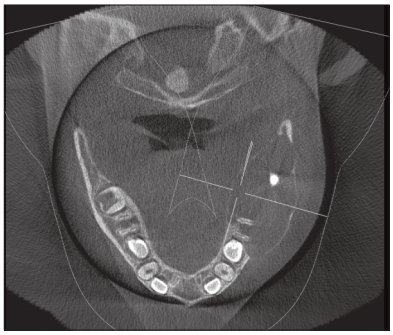

Fig. 3: The axial CT of the jaw revealed a large well-defined radiolucent lesion in the left body and ramus of the mandible with multilocular appearance causing expansion of both the cortex.

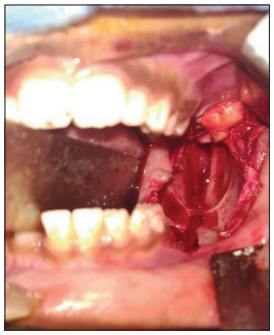

Fig. 4: Enucleated surgical field.

expanded buccal plate was slowly removed using low speed thin fissure bur. Care was taken not to apply any pressure to the body of the mandible during the entire surgery. The tumor was visualized and enucleated with cotton pushers, 36 and 74 were extracted along with the unerupted second molar contained within the lesion and the unerupted third molar tooth bud which was pushed to the coronoid process which also was removed (Figure 5).

The entire cystic content was removed, curetted without any remnants and submitted for histopathological analysis (Figure 6). Inferior alveolar nerve seen at the base of the cystic cavity was left undisturbed and preserved. The tumor bed was chemically cauterized with Carnoy's solution following which it was packed with BIPP (Bismuth iodine paraffin pastepack) with a small exit at the alveolar region. Closure was done with 3-0 vicryl suture. The mandible was checked for continuity and occlusion. Anesthesia was reversed the patient was shifted to ICU for postoperative recovery. Histopathological analysis of the enucleated specimen revealed a fibro vascular connective tissue exhibiting interlaced strands of odontogenic epithelium with central stellate reticulum like cells admixed with inflammatory cell infiltrate and red blood cells, which was suggestive of plexiform variant of ameloblastoma (Figure 7).

The BIPP pack was removed after a week in increments. Patient was asked to stay strictly on soft diet for a month and advised not to have any untoward injury to the left side jaw. Intra oral irrigation was taught to the patient care takers and patient was checked every 3 months. On a follow-up examination after a year, intra oral surgical site was completely healed with healthy mucosal covering and patient did not report any problems. Panoramic radiograph and CBCT taken revealed intense bone opacity indicating sufficient and significant bone formation from the margins to the center of the defect, reducing the remaining osseous cavity when compared to the preoperative radiograph (Figure 8,9,10,11)

DISCUSSION

Incidence of solid ameloblastoma under the age of 10 is a rare occurrence. They are uncommon in children, with 2.2% cases under 10 years and 8.7% between the ages of 10 and 19years (7). The first report of ameloblastoma in children was in 1962, where 7 cases were reported in children under the age of 9 years old (8, 9). Chaudhary et al analyzed 356 cases of ameloblastoma and observed that 91.86% of the cases were between the age group of 11 and 20 years and 8.14% were below the age of 10 (10). Ord et al. reported a higher percentage of unicystic ameloblastoma in children with only 2.2% under 10 years old although Asian and African regions showed a higher percentage from 14.6 to 25 % with no specific gender predilection (11-13).

Ameloblastoma presents with painless swelling with expansion of the buccal and lingual cortices of bone, and a possibility of bone perforation and unerupted tooth in the range of 70% to 83% (14,15). This was very similar

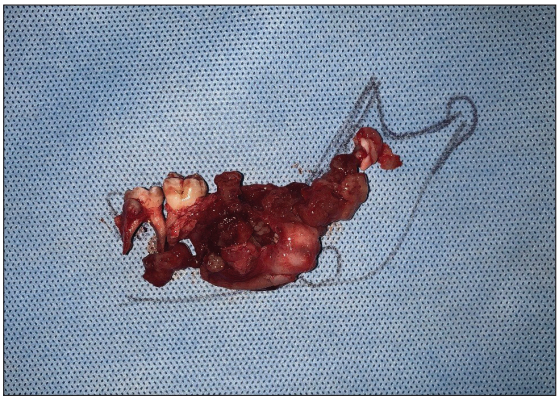

Fig. 5: Gross Enucleated specimen with mandible drawn for reference.

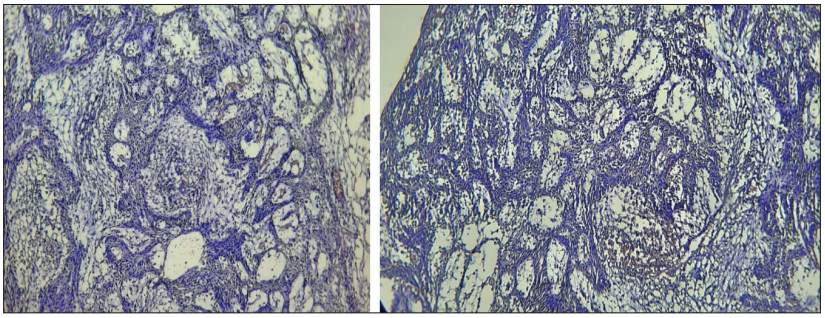

Fig. 6: Histopathological section revleaing the plexiform strands of ameloblastomatous epithelium associated with a fibrous connective tissue (H & E, 10x).

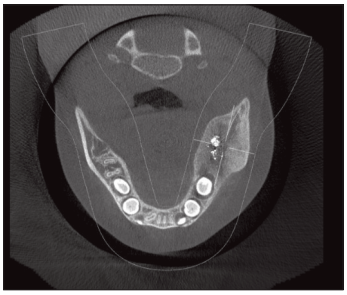

Fig. 7: Post operative Axial section of CBCT.

with our case where the unerupted second permanent molar tooth bud was contained within the tumor. The aggressive expansion of the tumor in relatively short span of time is due to the greater amount of cancellous bone in young patients, which contributes to the rapid proliferation of tumor cells (16).

The regimen for treating ameloblastoma falls into two categories 1) conservative approach using enucleation, curettage, marsupialization and 2) radical approach. Since this guarantee a very low recurrence rate while preserving the vital structures in the maxillofacial region (17). Though current literature supports the viewpoint of using wide surgical resection in recurrent cases and to use conservative approach in younger population as conservative approach preserves and maintains the natural growth of the mandible and craniofacial region (18). Aggressive approach in pediatric cases impacts the growth dynamics of the craniofacial complex, it could result in deformity and dysfunction of the jaw thereby affecting the physical and psychological development of the child. Several authors have found a recurrence rate of 55 to 90% for all ameloblastoma treated conservatively. However, the incidence of recurrence following even when radical resection is 5 to 15% (19). Unless resection is deemed necessary the most appropriate treatment would be treating the tumor conservatively with enucleation, chemical cauterization with the Carnoy's solution. Nakamura, et al. compared the long-term results between different approaches in treating 78 patients with ameloblastoma and found that conservative treatments including marsupialization and enucleation followed by sufficient bone curettage were useful and reduced the need for jaw resection (20). In our case, we resorted to enucleation and curettage followed by chemical cauterization, this had not only preserved the bone structures in good condition, but also saved the patient from psychological trauma resulting from aggressive surgery.

The current case depicts successful spontaneous healing of osseous defect after large cyst enucleation without the use of any filling materials, evident by follow up panoramic radiograph taken after a year which revealed intense bone formation in the enucleated site, thus confirming the results from the literature relating to spontaneous healing of large osseous defects without bone grafts in young patients and importance of conservative management in pediatric population (21, 22). Histopathology revealed plexiform strands and cords of odontogenic epithelial elements within the stroma. The plexiform type is more common than the follicular type in children (23). This finding was concurrent with our case, where the histopathological analysis revealed a plexiform variant of ameloblastoma.

In addition to enucleation, we had chemically cauterized the surgical bed with Carnoy solution. Carnoy's solution has been thoroughly studied and published reports indicate chemical cauterization provides a favourable prognosis of the treatment. The solution penetrates the cancellous bone to a depth of 1.54 mm, thereby devitalizing and fix the remaining tumor cells/cell rest (24). In

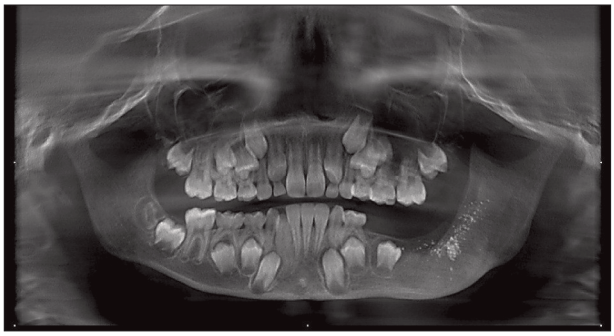

Fig. 8: Panoramic radiograph after one year revealing dense bone growth at the site of enucleation.

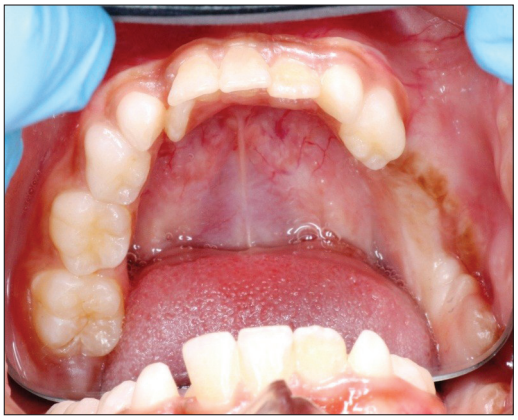

Fig. 10: Intra oral image showing healthy mucosal covering at the surgical site

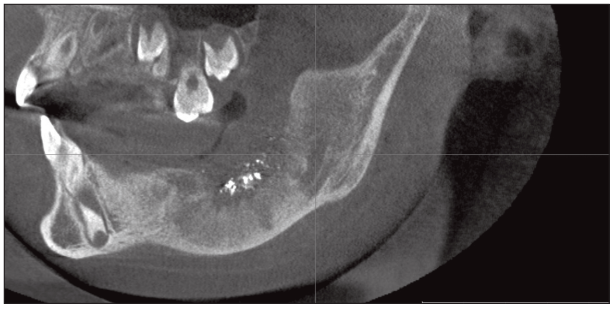

Fig. 9: Sagittal section of CT imaging after one year revealing dense bone growth at the site of enucleation.

our case, considering the age of the patient, we did not opt for a biopsy instead surgical enucleation was performed on the subsequent visit, since this would spare the patient of easily avoiding secondary surgical intervention. We believe that resection and reconstruction of the mandible is more easily performed on patients who are above the age of 15 due to the growth in the mandibular bone density. Fracture of the mandible after enucleation of mandibular cysts are extremely rare in pediatric population owing to the elastic nature of bone when compared to adults, similar to our case. One year follow up review revealed significant bone repair in the enucleated region of the mandible.

Though the recurrence rate is high for conservatively treated cases of ameloblastoma, review visits comprising of clinical and radiographic examination is mandatory due to the potential risk of late recurrence of the tumor in adulthood. Lifetime periodic follow-up is recommended for detection of any recurrence as even a 5 year tumor- free period does not necessarily mean a cure.

CONCLUSION

In pediatric population, it is peremptory that the treatment be performed as early as possible to prevent the possible proliferation of ameloblastoma to the adjacent tissues. Conservative options should considered as a first line of treatment in pediatric population to spare them from psychological trauma resulting from radical surgery.

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