Trends in Pediatric Ameloblastoma and its Management: A 15 year Indian Experience

Neelam N. Andrade · Shweta P. Shetye ·
Tejas S. Mhatre

Received: 5 February 2012 / Accepted: 30 April 2012 / Published online: 28 June 2012
© Association of Oral and Maxillofacial Surgeons of India 2012

Abstract

Purpose The purpose of this study is to put forth our 15 year experience with pediatric ameloblastoma in the Indian population.

Materials and Method This retrospective study was carried out in the pediatric group of 18 years and below, presenting with and diagnosed for ameloblastoma, at our institution over the past 15 years. The required data was collected by reviewing patient's case notes, relevant radiographs, histopathological reports, and treatment charts. The incidence of pediatric ameloblastomas with respect to age, sex, site of occurrence, histopathologic type, the type of treatment instituted, and recurrence rate was noted.

Results Of the total 165 pediatric tumors, ameloblastoma was the most common, 29 cases (17.5%). It occurred commonly in the age group of 12–18 years (mean age 14 years) with a marked male predilection. The most common site of occurrence was the posterior mandible. Majority were of the unicystic type. Two ameloblastomas which appeared as unilocular radiolucency were diagnosed as solid type. Twenty-eight were benign and one was a rare malignancy (Ameloblastic carcinosarcoma). Only two recurrences were noted over 15 years, of which one tumor interestingly recurred as a peripheral variety which

responded well to local excision. Ameloblastomas were either enucleated with mechanical curettage or resected followed by primary reconstruction with either a reconstruction plate or free fibula flap.

Conclusion We conclude that ameloblastomas are not uncommon in Indian pediatric population. Unilocular, unicystic ameloblastomas in the pediatric age group can be treated conservatively owing to their growth potential. Emphasis must be given to a long-term regular follow-up, conserving a more radical approach in case of a recurrence. However, Unilocular appearing ameloblastomas may be of the solid type which needs to be borne in mind as it not only alters the treatment modality but also emphasizes the importance of pre-operative incisional biopsy. Solid and unicystic aggressive ameloblastomas must be treated radically. Primary reconstruction with the free fibula flap is a viable option.

Keywords Ameloblastoma · Pediatric · India · 15 years

Introduction

Ameloblastoma is an aggressive, benign, epithelial odontogenic tumor that comprises of 10 % of the odontogenic tumors [1, 2]. Its occurrence is considered a rarity in the younger age group and accounts for approximately 10–15 % of all reported cases of ameloblastoma [3, 4]. The treatment for ameloblastoma is still controversial especially in children and adolescents. The growth potential of the jaws in the younger age group needs to be given due consideration, while determining the appropriate treatment modality.

Although, a lot has been written about this tumor, there still exists paucity of information regarding the behavior

N. N. Andrade (✉) · S. P. Shetye · T. S. Mhatre
Department of Oral and Maxillofacial Surgery, Nair Hospital
and Dental College, 107/1st Floor, A. L. Nair Road,
Mumbai Central, Mumbai 400 008, India
e-mail: drnnandrade@yahoo.co.in

S. P. Shetye
e-mail: shetye_shweta@rediffmail.com

T. S. Mhatre
e-mail: mhatretejas@gmail.com

and management of this tumor among the Indian population in the pediatric age group. Hence, this study aims at reviewing the incidence, behavior and management of pediatric ameloblastoma from the west of India. We hope this study will contribute to further meta-analysis, increase the cognition of the tumor and help develop a more rational treatment protocol in children and adolescents.

Material and Method

A retrospective study was carried out in Indian patients of age 18 years and below, who presented with and were histopathologically diagnosed for an ameloblastoma at our institution in Mumbai, India over the past 15 years (i.e., from January 1997 to July 2011). This study was carried out following the approval of the Ethics Committee of our institution. The required data was collected by reviewing the patients' case notes, relevant radiographs, histopathology reports, and treatment charts. The incidence of the tumor with respect to the age, sex, site of occurrence, histopathologic type, type of treatment instituted, and the recurrence rate was recorded. All the patients had undergone incisional biopsy prior to surgery. The surgical techniques included:

1. Enucleation with mechanical curettage removing approximately 2-mm of bone.
2. Resection followed by primary reconstruction with either a titanium reconstruction plate or a free fibula flap.

Results

Of the 165 pediatric tumors, 65 were of odontogenic origin and 100 of non-odontogenic origin. Among the 65 odontogenic tumors, ameloblastoma was the most common tumor comprising of 29 (44.6 %) cases and accounted for 17.5 % of the total pediatric tumors.

Age and Sex

The patients were divided in the age groups of 1–5 years, 6–11 years and 12–18 years. Most common age group of tumor occurrence was 12–18 years (mean age of 14 years) (Table 1). Of the 29 patients, 19 were males and 10 females, with the male to female ratio of 2.1:1.

In all the cases, the tumor was found in the mandible. In 21 of 29 cases (72.4 %), ameloblastoma occurred in the posterior mandible, 3 (10.3 %) in the anterior mandible and 5 (17.3 %) had the tumor crossing the midline. Hence, the most common site of occurrence was the posterior mandible.

Table 1 Age distribution of ameloblastoma in a pediatric age group

Age group (years)	No. of patients (%)
0–5	1 (3.4)
6–11	5 (17.3)
12–18	23 (79.3)

Fig. 1 Orthopantomogram showing unilocular radiolucent lesion in right hemimandible (yellow arrow). (Color figure online)

Histopathologic Typing

28 were benign and one was malignant—ameloblastic carcinosarcoma, which is one of the rarest reported tumors in the literature [5]. On the basis of established histological criteria, it was observed that 20 (69 %) of 29 exhibited the ‘unicystic’ type and nine (31 %) were of the ‘solid’ type. Apart from the standard histopathologic typing, immuno-histochemical study of tumor suppressor protein p53 is ideally recommended to delineate the more aggressive ameloblastomas. However, considering the high cost of this study and the low socio-economic status of the patients referred to our institute from a tertiary institute, it was not possible to perform this specific test.

Radiographic Appearance

Of the total 29 Ameloblastoma, seven cases that presented as multilocular radiolucency were diagnosed as ‘solid’ variety of the tumor, clinically (i.e., during surgery) as well as histopathologically. Twenty patients with unilocular radiolucency were diagnosed as the ‘unicystic’ variety as expected. Interestingly, two patients presenting with unilocular radiolucency were diagnosed as the solid variety of the tumor, as was seen on the cross section of the gross specimen and histopathologic slide (Figs. 1, 2, 3).

Surgical Management

After an incisional biopsy, a total of 29 cases were diagnosed with ameloblastoma. However, 17 of the 29 patients

Fig. 2 Coronal CT view showing expansile, unicystic nature of the ameloblastoma in the right mandible

Fig. 3 Cross section of gross specimen of unicystic appearing ameloblastoma showing its solid nature

underwent surgical management as the remaining patients were lost to follow up after the initial biopsy.

Ten cases which were histopathologically diagnosed as the ‘unicystic’ variant were treated by enucleation followed by mechanical curettage with rotary bur removing approximately 2-mm of bone (Figs. 4, 5, 6, 7, 8, 9). Four patients who were diagnosed of the ‘solid’ variety and three patients who were considered to be unicystic but showed signs of an aggressive tumor (i.e., root resorption of teeth, severe cortical perforation, marked thinning of the inferior border of the mandible) underwent segmental resection with a 1 cm margin of healthy bone. This was followed by primary reconstruction with a reconstruction

Fig. 4 A 13-year-old female with swelling of right mandibular angle (preoperative view)

Fig. 5 Preoperative orthopantomogram showing unicystic ameloblastoma with impacted and displaced third molar in the right mandibular molar ramus region

plate in five patients and a microvascular free fibula flap in two.

In those cases where significant cortical perforation was noted, but the overlying periosteum was intact, a supra-periosteal dissection was carried out, and an en block resection with the next intact anatomic barrier (ie., periosteum) was performed. Similarly, in cases where involvement of the periosteum or the overlying soft tissue was noted, the adjacent muscle and/or mucosa was excised along with the specimen.

Fig. 6 Intra operative view showing the residual bony defect after enucleation and mechanical curettage filled with demineralised freeze dried bone powder

Fig. 7 Orthopantomogram taken 2 weeks post enucleation of unicystic ameloblastoma. Note the radiolucent bony defect in the right posterior mandible

Fig. 8 1 year postoperative OPG showing good bone fill and no evidence of recurrence in right posterior mandible

The only case of malignancy in this study i.e., ameloblastic carcinosarcoma, was referred to an exclusive and well reputed oncology centre in the west of India. This

Fig. 9 1 year postoperative extra oral view

patient underwent two cycles of chemotherapy after which she was taken up for a total mandibulectomy and selective supraomohyoid neck dissection followed by primary reconstruction with a free fibula flap.

Recurrence

Only two recurrences were noted over 15 years. In one of these cases, the original tumor was a unicystic ameloblastoma, which had then shown a significant cortical expansion without a cortical perforation and was treated by an enucleation procedure. However, at recurrence, the patient was taken up for segmental resection followed by primary reconstruction with a free fibula flap. This patient was finally rehabilitated with transitional dental implants after a period of 2 months following the hardware removal. The primary tumor of the other case had not shown any cortical perforation, but periosteal involvement at the alveolar crest. Interestingly, in this case the tumor recurred as a ‘peripheral’ variety at the same site as the previous intra osseous lesion, which responded well to a local excision, and has not shown any signs of recurrence since then.

Discussion

Ameloblastoma is uncommon in the pediatric population, with only 8.5–15 % of all ameloblastomas in the Western countries [3, 6, 7]. The Asian and the African reports show

a higher percentage, ranging from 14.6 to 25 % [8, 9]. In our study, ameloblastoma accounted for 17.5 % of the tumors in the pediatric group which is comparable to the other reports [1, 3, 6–12].

Among the 65 odontogenic tumors in the pediatric group, ameloblastoma was the most common tumor comprising of 29 (44.6 %) cases. This differs from other reports in which odontomes were the most common odontogenic tumor [13, 14].

In our study, the incidence of ameloblastoma was highest in the age group of 12–18 years (79.3 %) with the mean age of occurrence being 14 years [15] which is consistent with a study by Arotiba et al. [16].

The male to female ratio of 2.1:1, indicates a marked male preponderance which is in unison with some reports [1, 10, 12]. However, some authors have reported equal gender distribution [7, 9, 11].

All the cases occurred in the mandible which reiterates the fact that this tumor has a marked predilection for occurrence in the mandible. The molar-ramus region was the most common site of occurrence as was noted by Zhang et al. [15].

The patients included in this study belonged to poor socio-economic strata where regular dental checkups and awareness regarding the same are lacking. Hence, these patients sought treatment only after a significant facial deformity due to the tumor or a noticeable mass intra orally, which led to the relatively late diagnosis by the clinician. Correlating with the history of all the patients over a period of many years, it is our opinion that the time interval between the onset and presentation of the disease could be around 12–15 months.

Ameloblastomas are currently classified into three types:

1. Solid or multicystic—which radiologically is usually characterized by multilocular radiolucency.
2. Unicystic type—which is radiologically characterized by unilocular radiolucency
3. Peripheral or extra osseous type.

We found the unicystic variety to be the most common type (69 %) which is a common finding in some studies [6, 17, 18]. However, another study has found the solid variant to be the predominant one [15].

By correlating the radiographic appearance and the histologic type, 20 cases which showed unilocular radiolucency were diagnosed of ‘unicystic’ variant, and all the cases which presented as multilocular radiolucencies, were of the ‘solid’ type as expected. However, surprisingly, two cases which presented as unilocular radiolucency were diagnosed as ‘solid’ type, which also has been reported in some studies [15].

Therefore, when a suspected ameloblastoma appears as a unilocular radiolucent lesion, the diagnosis of ‘solid’ type

also should be considered as this would alter the treatment because the solid varieties need to be treated more radically. This would also avoid the high recurrence rate associated with the solid ameloblastomas treated conservatively.

The management of ameloblastoma in the younger age group can be very challenging despite using reconstruction plates as this has a profound effect on the subsequent growth dynamics of the craniofacial skeleton, dentition, and the soft tissues. Conservative management for unicystic ameloblastomas like enucleation with curettage and use of physio-chemical agents has been long debated. Rosensteil et al. [19] have reported a high recurrence rate of 64 % for unicystic ameloblastomas treated conservatively and has concluded that simple enucleation and curettage may be inappropriate treatment. However, there have been reports of unicystic cases treated with enucleation and peripheral ostectomy with no recurrence even after 2–15 years [10]. Published reports now recommend a relatively conservative treatment like enucleation with peripheral ostectomy in the first instance, reserving a more aggressive therapy for any recurrence [6, 11, 15] considering the growth potential and capacity for bone regeneration in children. In our study, 10 cases of unicystic ameloblastomas were treated by enucleation and mechanical curettage with rotary bur removing approximately 2-mm of bone. In our institution patients with unicystic ameloblastomas treated conservatively are encouraged to adhere to a regular long term follow-up in order to detect any recurrences. Till date, only two recurrences have been noted, of which one was a unicystic type which had been treated conservatively. This patient on recurrence underwent segmental resection followed by primary reconstruction with free fibula flap and was then rehabilitated with transitional dental implants after a period of 2 months following the hardware removal. The other case interestingly recurred as a ‘peripheral’ variety of the tumor at the same site as the previous intra osseous lesion, which responded well to local excision, and has not shown any signs of recurrence since then. This recurrence could have been due to seeding of the intra osseous tumor cells through the perforated cortices and the periosteum into the overlying oral mucosa.

Two unicystic cases which showed signs of locally aggressive behavior, underwent segmental resection with 1 cm margin of healthy bone followed by primary reconstruction with a reconstruction plate or free fibula flap (Figs. 10, 11, 12, 13, 14, 15, 16, 17).

The four solid ameloblastomas which underwent treatment, were taken up for segmental resection followed by primary reconstruction owing to its high recurrence rate (75–90 %) following conservative treatment [6, 12].

The only malignancy noted in this study was an ameloblastic carcinosarcoma which is one of the rarest reported

Fig. 10 8-year-old boy with swelling of right mandible

Fig. 11 Preoperative intra oral view showing the lesion in the right mandible

Fig. 12 Preoperative orthopantomogram showing aggressive ameloblastoma of right mandible

Fig. 13 Intra oral residual defect after resection of ameloblastoma

Fig. 14 Plate fixed to the free fibula flap. Note the double barreling of the fibula to achieve adequate bone height

Fig. 15 Immediate postoperative orthopantomogram showing the microvascular free fibula flap in situ. Note the vertical mandibular height achieved with the double barrel technique

malignancies. Until now, only four cases of ameloblastic carcinosarcoma have been reported in the literature [5]. It is a rare mixed malignant odontogenic neoplasm with the

Fig. 16 1 year postoperative frontal view showing good facial contour

Fig. 17 1 year postoperative intra oral view showing good mandibular bone height

ameloblastic features seen in the epithelial as well as the fibroblastic component. In the literature, this malignancy has been seen to arise in pre existing lesions like the ameloblastoma, ameloblastic fibrosarcoma [20, 21], and ameloblastic fibroma [5] or arise de novo [22, 23]. This tumor may metastasize to the lungs and the regional lymph nodes [5], however, our case did not show any such findings despite thorough investigation. In spite of this, the patient underwent selective supraomohyoid neck dissection

in view of the risk of micro metastasis and total mandibulectomy followed by primary reconstruction with free fibula flap. Owing to the scarcity of reported cases, the long-term clinical significance is not known. More case descriptions would help understand the nature of this neoplasm and its clinical implication.

Conclusion

In conclusion, ameloblastomas are not uncommon in the Indian pediatric population. This tumor occurs commonly in the age group of 12–18 years with a marked male predilection. The most common site of occurrence is the posterior mandible. A correlation between the radiographic appearance and histological type must be sought by a pre-operative incisional biopsy, as unilocular radiolucent appearing lesion may be solid, which alters the treatment. Unilocular, unicystic ameloblastomas can be treated conservatively with enucleation and peripheral ostectomy, conserving a more radical approach in case of a recurrence. However, long-term and regular follow-up is a must to rule out recurrence. Solid and unicystic aggressive ameloblastomas must be treated radically, i.e., resection with 1 cm clear margin. Primary reconstruction with free fibula flap is a viable option as it restores form and function at the earliest with minimal donor site morbidity.

Conflict of interest The authors hereby wish to state that this paper does not have any financial and personal relationships with other people or organisations that could inappropriately influence (bias) their work.

References

1. Olaitan AA, Adekeye EO (1996) Clinical features and management of ameloblastoma of the mandible in children and adolescents. *Br J Oral Maxillofac Surg* 34:248–251
2. Rapidis AD, Andressakis DD, Stavrianos SD, Faratzis G, Arno-giannaki-Liappi N, Lagogiannis GA et al (2004) Ameloblastomas of the jaws: clinicopathological review of 11 patients. *Eur J Surg Oncol* 30:998–1002
3. Keszler A, Dominguez FV (1986) Ameloblastoma in childhood. *J Oral Maxillofac Surg* 44:609–613
4. Ueno S, Nakamura S, Mushimoto K, Shirasu R (1986) A clinicopathologic study of ameloblastoma. *J Oral Maxillofac Surg* 44:361–365
5. DeLair D, Bejarano PA, Peleg M, El-Mofty SK (2007) Ameloblastic carcinosarcoma of the mandible arising in ameloblastic fibroma: a case report and review of the literature. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 103(4):516–520
6. Ord RA, Blanchaert RH Jr, Nikitakis NG, Sauk JJ (2002) Ameloblastoma in children. *J Oral Maxillofac Surg* 60:762–770
7. Kahn MA (1989) Ameloblastoma in young persons: a clinicopathologic analysis and etiologic investigation. *Oral Surg Oral Med Oral Pathol* 67:706–715

8. Arotiba GT, Ladeinde AL, Arotiba JT, Ajike SO, Ugboko VI, Ajayi O (2005) Ameloblastoma in Nigerian children and adolescents: a review of 79 cases. *J Oral Maxillofac Surg* 63:747–751
9. Chidzonga MM (1996) Ameloblastoma in children. The Zimbabwean experience. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 81:168–170
10. Huang IY, Lai ST, Chen CH, Chen CM, Wu CW, Shen YH (2007) Surgical management of ameloblastoma in children. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 104:478–485
11. Takahashi IK, Miyauchi K, Sato K (1998) Treatment of ameloblastoma in children. *Br J Oral Maxillofac Surg* 36:453–456
12. Daramola JO, Ajagbe HA, Oluwasanmi JO (1975) Ameloblastoma of the jaws in Nigerian children: a review of sixteen cases. *Oral Surg Oral Med Oral Pathol* 40:458–463
13. Chuong R, Kaban LB (1985) Diagnosis and treatment of jaw tumors in children. *J Oral Maxillofac Surg* 43:323–332
14. Sato M, Tanaka N, Sato T, Amagasa T (1997) Oral and maxillofacial tumors in children: a review. *Br J Oral Maxillofac Surg* 1:26–31
15. Zhang Jing, Zexu GU, Jiang Lin, Zhao Jinlong, Tian Meiyu, Zhou Jun, Duan Yinzong (2010) Ameloblastoma in children and adolescents. *Br J Oral Maxillofac Surg* 48:549–554
16. Arotiba GT (1996) A study of orofacial tumours in Nigerian children. *J Oral Maxillofac Surg* 54:34–38
17. Leider AS, Eversole LR, Barkin ME (1985) Cystic ameloblastoma: a clinicopathologic analysis. *Oral Surg Oral Med Oral Pathol* 60:624–630
18. Pogrel MA, Montes DM (2009) Is there a role for enucleation in the management of ameloblastoma? *Int J Oral Maxillofac Surg* 38:807–812
19. Rosenstein T, Pogrel MA, Smith RA, Regezi JA (2001) Cystic ameloblastoma—behaviour and treatment of 21 cases. *J Oral Maxillofac Surg* 59:1311–1316
20. Shinoda T et al (1992) Cytologic appearance of ameloblastic carcinosarcoma (malignant ameloblastoma and fibrosarcoma) of maxilla: a case report. *Acta Cytol* 36:132–136
21. Tanaka T, Ohkubo T, Fujitsuka H (1991) Malignant mixed tumour (malignant ameloblastoma and fibrosarcoma) of the maxilla. *Arch Pathol Lab Med* 115:84–87
22. Kunkal M, Ghalibafiam M, Radner H, Reichert TE, Fischer B, Wagner W (2004) Ameloblastic fibrosarcoma or odontogenic carcinosarcoma: a matter of classification? *Oral Oncol* 40:444–449
23. Slater LJ (1999) Odontogenic sarcoma and carcinosarcoma. *Semin Diagn Pathol* 16:325–332