Maxillary Ameloblastoma: An Enigma for the Surgeon.

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SUMMARY

Background: Ameloblastoma is a benign but locally aggressive odontogenic tumour. Worldwide, maxillary ameloblastoma is rare but its late detection renders adequate treatment difficult. Majority occur in the mandible with about 5-20% occurring in the maxillary bone.

Objective: The purpose of this study was to analyze 21 cases of maxillary ameloblastoma seen and managed at the Oral and Maxillofacial Unit of Ahmadu Bello University Teaching Hospital, Zaria, Nigeria and Alba Clinic and Medical Centre, Kaduna, Nigeria.

Study Design: A retrospective study of cases of maxillary ameloblastoma from all cases of ameloblastoma seen from January 1993 to August 2008. Data with respect to patient's sex, age, tumour location, clinical presentation, radiologic features, biological and histopathologic type, surgical treatment and recurrences were analyzed.

Results: Out of 350 cases of ameloblastoma seen within the period, 21(6%) Patients were with maxillary ameloblastoma. Of the 21 cases, there were 13 males and 8 females, a male female ratio of 1.6 to 1, with an age range of 17-55 years (mean = 38.14), peaking at the 4th and 5th decades of life (61.9%). Tumour duration was from 3 months to 14 years. There were 18 unilateral and 3 bilateral swellings. Clinically, maxillary ameloblastoma presented with grotesque swellings, with antral involvement in 19 cases, teeth mobility/exfoliation. Radiologically, there were 20 multilocular and 1 unilocular radiolucent lesions. The most common histopathologic type was follicular (11, 52.4%).there were 22 procedures done on 21 patients; 21 maxillectomies and 1 enucleation. Follow up period of 18 patients was between 3 months and 10 years from which 3(16.7%) recurrences were observed.

Conclusion: Ameloblastuma is uncommon in the maxilla. While maxillary ameloblastoma is indistinguishable histologically from its mandibular counterpart, it is very lethal. An excellent result achieved in this study was due to the radical mode of treatment of the multilocular variety. Rehabilitation postoperatively remains a challenge. Periodic life-long follow-up is recommended.

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INTRODUCTION

Ameloblastoma is an invasive, potentially malignant neoplasm that consists of proliferating odontogenic epithelium supported by fibrous stroma[1]. It occurs in the mandible in a ratio of 5 to1[2]. About 5 to 20% occur in the maxillary bone with majority in the molar region[3,4]. While ameloblastoma occur in all age groups[4,5] the maxillary ameloblastoma occur 12 years later than that of its mandibular counterpart[2]. There is no sex predilection[2], although some authours have recorded more males than females[6,7] while some have documented more females than males[8].

Anatomically, the maxilla is associated with the presence of the paranasal sinuses, orbital and cranial cavities. Also, it's more cancellous nature make maxillary ameloblastoma to be more aggressive than their mandibular counterparts. Owing to the locally aggressive and infiltrative nature, a radical approach is usually advocated[7,8,9,10] to reduce the recurrence rate which could be as high as 100%[8]. A long time follow-up has been advocated because recurrence 30 yeas post-operatively have been reported[11]. Because of its uniqueness an analysis of 21 cases of maxillary ameloblastoma seen and managed at the Oral and Maxillofacial Unit of Ahmadu Bello University Teaching Hospital, Zaria, Nigeria and Alba Clinic and Medical centre, Kaduna, Nigeria is presented.

MATERIALS AND METHOD

The materials for this study consisted of a retrospective study of maxillary ameloblastoma seen and managed at the Maxillofacial Unit, Ahmadu Bello University Teaching Hospital, Kaduna and Zaria, Kaduna State, Nigeria and Alba Clinic and Medical Centre, Kaduna, Kaduna State, Nigeria. From the entire cases of 350 ameloblastoma seen between January 1993 and August 2008, medical records of patients with maxillary ameloblastoma were retrieved for analysis of patient's sex, age, tumour location, clinical presentation, radiologic features, biological and histopathologic types, surgical treatment and

recurrences. Data collected was analyzed using Microsoft Excel.

RESULTS

There were 21 cases of maxillary ameloblastoma representing 6 % of total ameloblastoma seen in both hospitals during the study period under review. They were 13 males and 8 females, a male –female ratio of 1.6 to 1 (Table 1). The age range was between 17 and 55 years old with a mean of 38.14, majority 13 (61.9%) were in the 4th and 5th decades of life (Table 1). Duration of lesion ranged between three months and 14 years. There were 18 (85.7%) unilateral and 3(14.3%) bilateral cases of maxillary ameloblastoma (Fig. 1). While 20 (95.2%) were posterior tumours and one (4.8%) anterior tumour. Three of the posterior tumours had crossed the midline (Fig. 1).

The clinical presentations varied greatly, all the 21 cases had grotesque swellings (Fig 2). In terms of site majority, 19(90.5%) had antral involvement with two (9.5%) extending into the zygomatic bone, the temporal region and into the orbit with blindness respectively. There were involvement of nasal cavity in five (23.8%) and one (4.8%) case in the palate. Presenting complaints were teeth mobility in 12 (57.1%) cases, exfoliated teeth in 7(33.3%), nasal swelling/obstruction in 5(23.8%), ulceration of lesion in 9 (42.9%), proptosis in 3(14.3%), occlusal furrow in 10(47.6%), bleeding in 4(19.0%) and epiphora in 1(4.8%) case. Biologically, there were 18(85.7%) solid, 1(4.8%) cystic and 2 (9.5%) solid-cystic (Table 2) lesions. The histopathologic types were 11(52.4%) follicular, 3(14.3%) plexiform, 3(14.3%) follicular with squamous metaplasia and 4(19.0 %) acanthomatous (Table 2). Radiography showed multilocular radiolucency and opacification of the maxillary sinus in 20 (95.2%) and one (4.8%) unilocular radiolucency (Fig. 3). Radical treatment was the modality of treatment in 20(95.2%) and one (4.8%) had enucleation (Table 3). There were two (18.2%) recurrences of 11 followed up cases and (9.5%) of total cases.

Table 1: Showing age and sex distribution of 21 patients with maxillary ameloblastoma.

Age range	Sex		No (%)
	Male	Female	
0 - 9			
10 - 19	1	1	2(9.5)
20 - 29	3		3(14.3)
30 - 39	3	2	5(23.8)
40 - 49	7	1	8(38.1)
50 - 59	2	1	3(14.3)
TOTAL	**13**	**8**	**21 (100)**

Table 2: showing the histologic types and the gross appearance.

Histopathologic types	No	%
Follicular	11	52.4
Follicular with squamous metaplasia	3	14.3
Plexiform	3	14.3
Acanthomatous	4	19.0
Total	21	100
Gross appearance		
Solid	18	85.7
Cystic	1	4.8
Solid-cystic	2	9.5
Total	21	100

Table 3: Treatment modalities of 21 patients

Treatment modalities	No
Total maxillectomy **	15
Bilateral maxillectomy	2
Palatoalveolectomy	1
Subtotal maxillectomy **	2
Radical maxillectomy +excision of zygoma	1
Enucleation	1
Total	**22**

** 1 patient had left total maxillectomy with right subtotal
*2 patients had exenteration

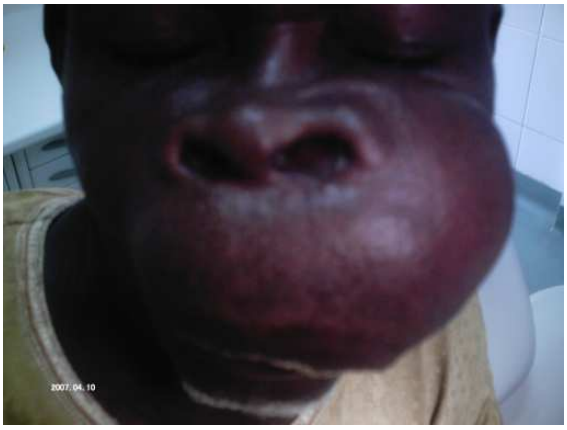

Fig 1. Bilateral maxillary ameloblastoma in a 55 year old male Nigerian.

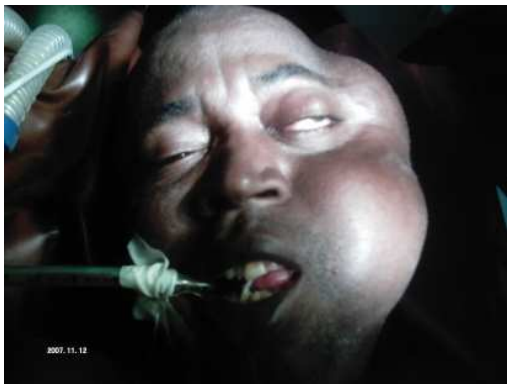

Fig 2. Unilateral maxillary ameloblastoma with temporal, zygomatic bone and arch with facial nerve involvement.

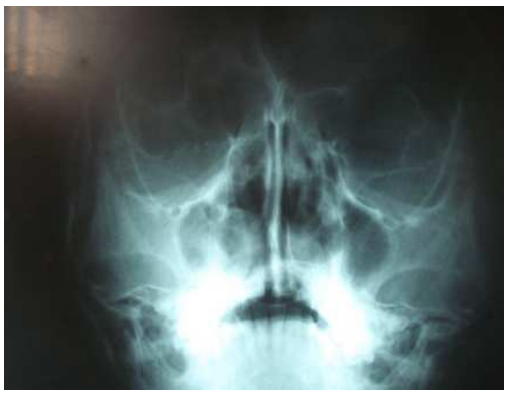

Fig 3. Multilocular radiolucency of the left maxilla with opacification of the left maxillary antrum

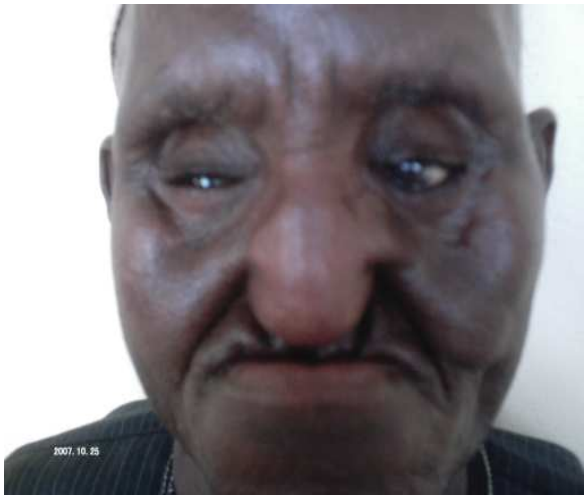

Fig 4. Facial disfigurement following bilateral ameloblastoma

DISCUSSION

Globally, maxillary ameloblastoma is more rare than mandibular lesions with reports in the literature often limited to case reports or case series [3,12-18]. From our centre, the last report on ameloblastoma was by Olaitan et al [6] who recorded 4.7% as maxillary ameloblastoma. This current report of 21 cases of maxillary ameloblastoma collected over 15 years 8 months representing 6% of total ameloblastoma seen is in close agreement with the global ratio 1 to 5[3,4,6] and reports by Tsaknis and Nelson[7] and Zwahlen and Gratz [19] of 21 and 26 cases of maxillary ameloblastoma respectively. Our study shows a male preponderance of 1.6:1 which is similar to other reports [7] but contrasts with the Zurich study by Zwahlen and Gratz [19] who have documented a female: male ratio of 1.5 to 1 and Scaccia and others[17] report of equal sex distribution.

Ameloblastoma occur at all ages[5,710,20], however majority present during the 3rd or 4th decades of life. However, in the maxilla, lesions are seen more in patients over a decade older[7]. In this study, 61.9% were in the 4th and 5th decades. A mean of 38.14 recorded in our study is lower than 45.6 years recorded by Tsaknis and Nelson.[7] All our patients presented with gross maxillary swellings (Fig. 1). Maxillary ameloblastoma may be asymptomatic. With time and growth of the tumour within the richly vascularized cancellous bone of the maxilla, tumour spreads to the adjacent paranasal sinuses, nasal cavity, nasopharynx, orbital adnexials, lacrimal apparatus, skull base and intracranial structures[7,12,14,15,19]. This results in difficulty in mastication, deglutition, loosening of teeth, epistaxis, nasal obstruction, with rhinorrhoea[12,18,21]. Ulceration may occur following trauma, long history or topical application of herbal medication[4,7,10]. While ulceration was the most common presentation in the report of Tsaknis and Nelson[7], in our study there were only nine cases of ulcerated lesions with a history of topical application of herbal medication in six cases.

Majority are located in the posterior region[5,7]. In our series, there were 20 (95.2%) posterior tumours and only one (4.8%) anterior tumour. Posterior maxillary tumours tend to be larger and more destructive with frequent involvement of the maxillary antrum (90.5%), the zygomatic bone (9.5%), the globe (9.5%) and the temple (9.5%) as shown in Figure 2. One of the temporal lesions was found to have eroded the bone with facial nerve deficit (Fig. 2). Tumour site distribution in this report compares favourably with that by Tsaknis and Nelson's [7] who found one anterior lesion with the others involving the orbit, nasal floor and the maxillary sinus among 21 cases of maxillary ameloblastoma. According to Small and Waldron [4], 21 (47%) occur in the molar region, 15(33 %) in the antrum and nasal floor, 4(9%) in the premolar and canine region respectively and 1(2 %) in the palate.

Bray *et al*[12] reported a maxillary ameloblastoma presenting as a nasal polyp. In this series there were five cases with nasal obstruction and one with epiphora following compression and or invasion of the nasolacrimal duct. Ophthalmic complication recorded in two cases were probably due to exposure keratitis following proptosis, involvement of the ophthalmic artery at the postero-supero-medial aspect of the maxillary sinus or by direct invasion of the globe via the intra-ocular muscles. Orbital, fronto-ethmoidal sinus, skull base and intracranial extension have resulted in the death of the patients[9,13-15,17] either from before or after treatment from recurrent lesions. Muller and Slootweg[9] concluded that ameloblastoma invades the spongy bones readily with little tendency of the cortical bone, with the periosteum acting as a great barrier and no definite capsule where the tumour adjoins the oral mucosa.

Extension of the tumour to the paranasal spaces makes standard radiologic diagnosis difficult[22]. Lesions usually appear as either unilocular or multilocular (soap bubble or honey comb) with well defined scalloped margins. Radiographic records showed 20 multilocular radiolucency with opacity of the maxillary antrum and 1 expansile unilocular lesion (Fig. 3). Incidentally, all the multilocular lesions were in the posterior region. According to Williams[23] multilocular ameloblastornas have a poorer prognosis than their unilocular counterpart. Unilocular lesions may simulate odontogenic cysts while the multilocular lesions simulate odontogenic keratocysts, fibromyxoma, giant cell granuloma and aneurysmal bone cysts. Unilocular lesions occur in younger patients [2,24]. Coincidentally, in our series the unilocular lesion was in the youngest patient. MRI and contrast enhanced CT [25] offer the best imaging methods for visualization of extensive lesions; however in this study none was done because apart from the prohibitive cost, these hi-tech machines only became available in our centre in 2005.

Regarding the biologic gross appearance, ameloblastoma may be cystic, solid or cystic-solid, however nearly all ameloblastomas demonstrate cystic degeneration [4,8,26] as single tumour may occasionally exhibit the three structural characteristics[4]. Small and Waldron[4] believe that cystic degeneration is a function of age. In this study, majority 18(85.7%) were solid, 1 (4.8%) cystic and 2 (9.5%) solid-cystic. Histopathologically, follicular and plexiform patterns are the most predominant[2,4,8,12]. This is similar to our study that has reported more follicular type (Table 2). Sehdev et al.[8] believe the histopathologic types has no clinical and prognostic implications as different sections from same tumour may give mixed histologic patterns. A view contested by some authors[2,5,9]. Surgical approach is the accepted treatment of choice, however, there is

still no consensus regarding conservative versus radical surgery. The maxillary ameloblastoma are more difficult to treat because of the combination of the well vascularized, fragile, cancellous maxillary bones, presence of the paranasal sinuses, nasal and orbital cavities which readily facilitates tumour spread to the zygomatic bone, cranial base and paracranial structures and the pterygomaxillary fissure. Radical surgery as defined by Muller and Slootweg[9] is a procedure in which ameloblastoma is removed with a marginal of normal bone by using segmental or marginal resection. However, most investigators have recommended at least between 1cm and 3cm of surrounding healthy bone[9,27].

Table 3 shows the various surgical approaches used in the treatment of 21 cases of maxillary ameloblastoma. The two cases with orbital involvement had orbital exenteration of the involved globes while lesions involving the temporal region were excised. In one of the temporal lesions where there was erosion of the bone, lesion was teased off the dura mater. Enucleation is adequate for unilocular/unicystic lesions particularly for anterior lesions and where periodic follow-up is available. The most important factor in the treatment of maxillary ameloblastoma is the prevention of local recurrence which varies from 20 to 100%[6-8]. Sehdev *et al*[8] reported 100% recurrence rate following conservative surgery in 11 patients and 22% following radical surgery. In this study, only two (18.2%) of the 11 patients with available follow-up records had recurrence compared to 8(50%) of the 16 cases followed-up by Tsaknis and Nelson[7]. Recurrence rate of maxillary ameloblastoma is usually related to the mode of surgery, extent of the tumour presentation. The low recurrence rate in this study is due to the adoption of radical surgical procedures, which allowed for complete extirpation of tumour with sufficient safety margins in all the four dimensions. Radical surgery in our center involved the removal of tumour with a margin of between 2.5 and 3cm of apparent normal bone because majority of the patients usually default follow-up protocol in our environment.

Rehabilitation of maxillary defects can be achieved by the use of prosthetic obturators[29], pedicled and free tissue transfers with or without bone grafts[28-31]. In this study, 18(85.7%) of our patients had prosthetic obturator rehabilitation with good functional and aesthetic results while the remaining three (14.4%) patients (two patients with bilateral maxillectomy and the patient with total maxillectomy and right subtotal maxillectomies) were not rehabilitated resulting in both functional and psychosocial disability (Fig. 4). Rehabilitation of bilateral maxillectomized patients usually requires the use of implants.

CONCLUSION

Maxillary ameloblastoma is uncommon. The richly vascularized and cancellous maxillary bone facilitating extension into the paranasal sinuses, orbital and cranial cavities make maxillary ameloblastoma very lethal. Radical surgery offers best result. Rehabilitation postoperatively remains a challenge particularly in bilateral maxillectomized patients. A lifelong time follow-up is advocated. Competing interest We hereby declare that there is no competing interest of any type.

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