Annals of African Medicine
Vol. 4, No. 1; 2005: 23 – 30

MAXILLOFACIAL SARCOMAS IN NIGERIA

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Key words
Jaws, sarcoma, osteosarcoma, surgery

Abstract
Background: Maxillofacial sarcomas are rare constituting between 4-8% of all malignancies in the region. A few case reports of individual tumours are available while reviews of significant series is lacking. This report presents 80 cases of sarcoma collected over 23 years at a tertiary oral care centre in Kaduna, Nigeria.
Method: Retrospective study of all maxillofacial sarcomas in northern Nigeria.
***Results*:** There were 406 maxillofacial malignancies of which 80 (20%) were sarcomas. Fourteen histopathologic types were found of which osteosarcoma (28%), chondrosarcoma (17%), rhabdomyosarcoma (12%) and fibrosarcoma (12%) were predominant. The male to female ratio was roughly equal (1.3:1). Patients with sarcoma were between 24 days and 90 years old (mean age 31±15 years) with most patients (26%) in the third decade of life. Cases presented with symptoms such as swelling (100%), pain (54%) and tissue ulceration (26%). Surgery was performed for 46% of cases treated while radiotherapy was used for 26%. Some cases refused hospital treatment because of poverty and ignorance while poor medical infrastructure limited treatment options in several cases regarded as advanced lesions.
Conclusion: In Northern Nigeria, sarcomas account for 20% of all maxillofacial malignancies with the osteosarcoma as the predominant type. Most affected were people in the third decade of life. Surgery was the main modality used for treatment while some patients had no treatment due to self-discharge and late presentation. The need for improved medical awareness and upgrading of infrastructure was stressed.

Mots clé:
Les mâchoires, le sarcome, osteosarcome, la chirurgie

Résumé
Fond: les sarcomes de Maxillofacial sont rares constituant entre 4-8% de toutes malignités dans la région. Les rapports de quelques cas de tumeurs individuelles sont disponibles pendant que les revues de feuilleton significatif manquent. Ce rapport présente 80 cas de sarcome recueilli par-dessus 23 années à un centre de soin oral tertiaire à Kaduna, Nigéria.
Méthode*:* l'étude rétrospective de tous les sarcomes de maxillofacial au nord du Nigéria.
Résultats: il y avait 406 malignités de maxillofacial dont 80 (20%) étaient des sarcomes. Ils ont été trouvés quatorze types histopathologiques dont osteosarcome (28%), chondrosarcome (17%), rhabdomyosarcome (12%) et fibrosarcome (12%) étaient prédominant. La proportion de mâle à femelle était à peu près égal (1.3:1). Les malades avec le sarcome étaient âgés d'entre 24 jours et 90 ans (âge moyen 31±15 ans) avec la plupart des malades (26%) dans la troisième décennie de vie. Les cas présentés avec les symptômes tels qu'accroissement (100%), la douleur (54%) et l'ulcération de tissu (26%). La chirurgie a été exécutée pour 46% de cas traités pendant que radiothérapie a été utilisée pour 26%. Quelques-uns ont refusé le traitement d'hôpital à cause de la pauvreté et l'ignorance pendant que le manque d'infrastructure médicale a limité les options de traitement dans plusieurs cas considérés

comme avancés.
Conclusion: Au nord du Nigéria, les sarcomes représentent 20% de toutes malignités de maxillofacial avec l'osteosarcome comme le type prédominant. Les plus affectés étaient des gens dans la troisième décennie de vie. La chirurgie était la modalité principale utilisée pour le traitement pendant que quelques malades n'ont pas eu de traitement en raison de fuite et la présentation tardive. Le besoin pour une forte conscience médicale et l'amélioration d'infrastructure a été accentué.

Introduction

In the oral and maxillofacial region, sarcomas like other malignancies are uncommon. However, their occurrence results in considerable morbidity and mortality. In adults, neoplasms are often epithelial in origin and malignant in character, [1,2] while children usually have more of mesenchymal tumours with greater tendency to benignity. [3,4,5]

Compared to carcinomas, sarcomas are rare. Budhy et al, [6] examined 994 histopathological specimens of malignancies of the mouth and jaws from East Java, Indonesia. They found only 42 (4%) sarcomas while squamous cell carcinomas made up 70%. Using similar criteria in Zaria, Nigeria, 8% of malignancies were sarcomas with 75% being squamous cell carcinomas. [7,8]

The predominant oral and maxillofacial sarcoma is controversial. In the young, rhabdomyosarcoma is the commonest sarcoma according to Soule et al, [9] while osteogenic sarcoma (osteosarcoma) predominates after puberty. [8] All body parts and any age group can develop sarcomas but while the so-called soft tissue sarcomas (STS) commonly arise in the soft tissues, osteosarcoma and chondrosarcoma usually arise from bone and cartilage respectively.

In Nigeria, few case reports of specific sarcoma have been published, [10 - 13] but detailed studies are lacking.

Patients and Methods

Patient data were obtained from a retrospective search of medical records at the Maxillofacial Unit, Ahmadu Bello University Teaching Hospital, Kaduna, Nigeria seen from January 1977 - December 1999. From records of cases of histopathologically diagnosed malignancy of the oral and maxillofacial region, sarcoma was selected and studied. Lymphomas and lymphosarcomas were excluded.

Results

Relative incidence and histopathologic types
There were 406 cases of malignant neoplasms of the oral and maxillofacial region within the study period of which 80(20%) were sarcomas as compared to 191(47%) cases of squamous cell carcinoma. Fourteen types of sarcoma were diagnosed with no record of secondary neoplasia. Two cases of chondrosarcoma were recurrent, haven been previously managed elsewhere. Table 1 shows the relative distribution of the lesions and their sex incidence. Osteosarcoma was the most common lesion (n=22, 28%) followed by chondrosarcoma (n=14, 17%), rhabdomyosarcoma (12%) and fibrosarcoma (12%).

Sex and age distribution
There were only slightly more male than female patients in these series of 80 cases, a ratio of 1.3:1 respectively (Table 1). Age range of patients was from 24days to 90 years (mean 31±15 years, median age 29years) from Table 2. The age distribution of the patient population showed that most (26%) were in the third decade of life while those in the first and sixth decades were 10% each.

Tumour site distribution
Sarcoma of the maxillofacial region occurred more in the hard tissues of the jaws (72%) than in the soft tissues (11%) from Table 3. On the basis of tumour type, twice as many cases of osteosarcoma occurred in the maxilla (n=7) as in the mandible (n=14). STS such as rhabdomyosarcoma favoured the cheeks (n=4, 40%) over either jawbone (n=2, 20% each). There was a case of Ewing's sarcoma, which developed tumour secondaries on the forearm while on cytotoxic chemotherapy in the hospital

Clinical features and duration of symptoms
In Table 4, all cases (100%) presented with swelling of the affected site. No case of asymptomatic lesion was recorded. Apart from swelling, other prominent features were pain (54%) and ulceration (26%). Patients presented within 0.5-192 months of onset of symptoms. Almost all cases of osteosarcoma (n=18,90%) came to hospital while complaints were less than a year old. A case of chondrosarcoma in Table 4 presented with proptosis and loss of vision due to retrobulbal involvement by tumour.

Treatment and follow-up
Among 80 cases of sarcomas seen in the oral and maxillofacial region of this northern Nigerian population, the mainstay of treatment was surgery (46%) followed by chemotherapy (26%) using cyclophosphamide, adriamycin, vincristine and methotrexate (Table 5). Specific drugs used, dosages and number of courses were determined by a consultant radio-oncologist. Patients for radiotherapy were referred to the University College Hospital, Ibadan, Nigeria. Follow-up was available for only two treated cases.

Table 1: Types and sex distribution of 80 maxillofacial sarcomas

Neoplasm	Sex		No. (%)
	M	F	22 (27)
Osteosarcoma	9	14	14 (17)
Chondrosarcoma	8	6	10 (13)
Rhabdomyosarcoma	6	4	10 (13)
Fibrosarcoma	5	5	5 (6)
Malignant fibrous histiocytoma	5	0	5 (6)
Liposarcoma	3	2	2 (3)
Malignant schwannoma	2	0	1 (1)
Synovial sarcoma	1	0	1 (1)
Retinoblastoma	0	1	1 (1)
Kaposi's sarcoma	0	1	1 (1)
Haemangiopericytoma	1	0	1 (1)
Malignant giant cell tumour	0	1	1 (1)
Ewing's sarcoma	1	0	1 (1)
Unclassified sarcoma	4	2	6 (8)
Total	45	35	80 (100)

Table 2: Age of 79 patients with maxillofacial sarcoma

Neoplasm	Age (years)							
	0 - 9	10 - 19	20 - 29	30 - 39	40 - 49	50 - 59	60+	Mean
Osteosarcoma	-	4	5	4	4	3	2	37
Chondrosarcoma	-	-	7	4	2	1	-	32
Rhabdomyosarcoma	5	4	1	-	-	-	-	16
Fibrosarcoma*	1	5	1	1	1	-	-	26
Malignant fibrous histiocytoma	-	-	1	1	1	2	-	39
Liposarcoma	-	1	-	1	1	2	-	39
Malignant schwannoma	-	-	-	-	2	-	-	47
Synovial sarcoma	-	-	1	-	-	-	-	28
Retinoblastoma	1	-	-	-	-	-	-	4.5
Kaposi's sarcoma	-	-	-	-	-	-	1	60
Haemangiopericytoma	-	-	-	1	-	-	-	35
Malignant giant cell tumour	-	-	-	1	-	-	-	35
Ewing's sarcoma	-	1	-	-	-	-	-	15
Unclassified sarcoma	-	1	4	1	-	-	-	27
Total	7	16	20	14	11	8	3	

*Age of one patient not known

Table 3: site distribution of 80 maxillofacial sarcomas

Neoplasm	Site (%)								
	Maxilla	Mandible	Palate	Cheek	Floor of mouth	Eye	Nose	Nasoph-arynx	Lip
Osteosarcoma	7	13	2	1	-	-	1	-	-
Chondrosarcoma	7	8	-	-	-	-	-	-	-
Rhabdomyosarcoma	2	2	1	4	1	-	-	-	-
Fibrosarcoma	1	6	2	-	1	-	-	-	-
Malignant fibrous histiocytoma	1	3	-	-	-	-	-	-	1
Liposarcoma	1	-	1	3	-	-	-	-	-
Malignant schwannoma	-	-	1	-	-	-	1	-	-
Synovial sarcoma	-	-	-	-	-	-	-	1	-
Retinoblastoma	-	-	-	-	-	1	-	-	-
Kaposi's sarcoma	-	-	-	1	-	-	-	-	-
Haemangio-pericytoma	-	1	-	-	-	-	-	-	-
Malignant giant cell tumour	-	-	1	-	-	-	-	-	-
Ewing's sarcoma	-	1	-	-	-	-	-	-	-
Unclassified sarcoma	2	3	-	-	-	-	-	1	-
Total	21 (26)	37 (46)	7 (9)	9 (11)	2 (3)	1 (2)	2 (3)	2 (3)	1 (2)

Site was not available for 2 osteosarcoma
More than one site may be involved
The Ewing's sarcoma metastasized to the forearm

Table 4: Clinical features in 80 patients with maxillofacial sarcoma

Neoplasm	Feature (% of cases)								Duration (months)
	Swelling	Pain	Ulceration	Tooth-ache	Loose tooth	Pus discharge	LMOP	Others	
Osteosarcoma	100	68	36	-	9	9	-	50	1.5-192
Chondrosarcoma	100	71	36	14	-	-	21	43	2-13
Rhabdomyo-sarcoma	100	60	2-	-	-	-	10	30	0.5-4
Fibrosarcoma	100	30	10	30	10	-	30	20	2-72
Malignant fibrous histiocytoma	100	20	40	10	-	-	-	40	3-60
Liposarcoma	100	20	20	-	-	-	-	-	1.8-13
Malignant schwannoma	100	100	100	100	-	-	-	-	1-6
Synovial sarcoma	100	-	-	-	-	-	-	-	5
Retinoblastoma	100	100	-	-	-	-	-	100	NA
Kaposi's sarcoma	100	100	100	-	-	-	100	100	4
Haemang-iopericytoma	100	-	-	-	-	-	100	-	3
Malignant giant cell tumour	100	100	-	100	-	-	-	-	1
Ewing's sarcoma	100	100	-	100	-	-	-	-	3
Unclassified sarcoma	100	33	-	17	-	-	17	17	2-11
Total	100	54	26	13	4	3	13	34	0.5-192

LMOP: limitation of mouth opening
Others: bleeding, tooth displacement, proptosis, metastatic features
NA: not available

Table 5: Treatment modalities for 80 malixillofacial sarcomas

Neoplasm	Treatment (% of cases)				
	Surgical excision	Resection± reconstruction	Chemotherapy	Radiotherapy	None
Osteosarcoma	5	32	27	5	50
Chondrosarcoma	14	36	21	-	36
Rhabdomyosarcoma	30	10	30	10	40
Fibrosarcoma	30	30	20	-	20
Malignant fibrous histiocytoma-	40	40	40	-	20
Liposarcoma100	20	20	20	20	20
Malignant -schwannoma	-	100	-	-	-
Synovial sarcoma	-	-	-	-	100
Retinoblastoma	100	-	-	-	-
Kaposi's sarcoma	-	-	-	-	100
Haemangiopericytoma	-	-	-	-	100
Malignant giant cell tumour	-	100	-	-	-
Ewing's sarcoma	-	-	100	-	-
Unclassified sarcoma	-	-	100	-	-
Total	16	30	25	4	39

Some patients had more than one form of treatment

Discussion

The incidence of maxillofacial sarcomas is not known. Budhy et al, [6] examined 994 histopathological specimens of maxillofacial malignancies from East Java, Indonesia. They found 42 (4%) sarcomas in their sample. In a similar review of specimens in Zaria, Nigeria, Rafindadi and Ayuba, [7,8] found 20 sarcomas (8%) out of 259 malignancies of the oral mucosa and jaws. Out of 406 maxillofacial malignancies seen at our center within 23 years, 80 (20%) were sarcomas. While this is higher than previous records, it may reflect harvesting of cases in the northern part of Nigeria (estimated population 70 million in 1998) by our center. It is also the contention of Smith, [14] that statistics from histopathological specimens as reported in the references from East Java, Indonesia and Zaria, Nigeria is biased in several respects. We believe that a yearly sarcoma incidence of 4 cases in the estimated population served by our center shows rarity of this lesion in the population.

The most common sarcoma is controversial. It depends on age group, site and possibly racial factors. According to Soule et al, [9] rhabdomyosarcoma is the commonest oral and maxillofacial sarcoma of childhood while in adults; osteogenic sarcoma (osteosarcoma) is predominant. [8] While the neoplasms called soft tissue sarcomas (STS) occur mostly in maxillofacial soft tissues, osteosarcoma and

chondrosarcoma are more from the jawbones.[7, 8,14] It is the belief of Miller and Dalager [15] that rhabdomyosarcomas are commoner among Caucasians than Negroes speculating a genetic factor in the Caucasoid stock.

In Ibadan, Nigeria, osteosarcoma accounts for 37% of sarcomas over a 15years period from the report of Daramola et al.[10] This is similar to the 30% incidence from Pretoria, South Africa. [16] The frequency of osteosarcoma (50%) from Zaria, Nigeria is much higher others in the African literature.[7, 8] However, the Zaria report had fewer variety of sarcomas. As with other reports, the most common sarcoma in our series was osteosarcoma (27%) from Table 1. Among 22 cases of osteosarcoma found in our series, there were fewer males than females, ratio 1:1.5. This is a reversal of the male to female ratio of 1.3:1 found from 7 cases from Ibadan, Nigeria.[10] No sexual predilection for osteosarcoma was observed in American and British studies.[14, 17] In view of the marginal differences in sexual predilection from these Nigerian reports, it is unlikely that osteosarcoma has any real preference for either sex in the Nigerian population.

Unlike in the rest of the human body where osteosarcoma occurs mostly in the 2nd decade, that of the maxillofacial region occurs in older persons (mean age 38years) from the study of Harrison and Lund. [14] A lower mean age for occurrence of osteosarcoma (25years) probably reflecting the age expectancy of that period was found at Ibadan, Nigeria by Daramola et al.[10] In view of the rise in life expectancy of the Nigerian population to 57years in 2000, the mean age of 38 years for osteosarcoma in the present series (Table 2) is understandable. Thirteen patients (59%) in our series with osteosarcoma were between 24-50 years old showing its bias for occurrence in adults.

Sarcomas can originate in any part of the body but certain types show a predilection for some parts of the maxillofacial region. Considering specific neoplasms, osteosarcoma occurs more in the maxilla (n=4) than the mandible (n=3) from the work of Daramola et al.[10] Though soft tissue involvement by the tumour can occur it is rare. Table 3 showed that among 22 cases of osteogenic sarcoma more occurred in the mandible (59%) than maxilla (32%) with few in the palate (9%) and cheeks (4%).

Sarcomas could be detected during routine examination for other conditions or due to non-specific symptoms. Since cure of orofacial malignancies is enhanced by early detection and initiation of adequate treatment, the dental surgeon or medical practitioner has a vital role to play in early detection particularly at the asymptomatic stage through opportunistic screening.[16, 18] The presenting features of sarcomas are non-specific and depend on tumour location, size, rate of growth, duration and the level of cancer awareness of the individual.[14, 18] Cases of osteosarcoma seen at Ibadan, Nigeria all presented as huge swellings of the jaws while a few had pain, tooth loosening, derangement of teeth and ulceration.[10] By Nigerian standards, the reported duration of these lesions were quite short 0.5-18months (mean 4.8). Tumour duration in benign oral and maxillofacial lesions in our environment is in years rather than months.[19, 20,21] In our 22 patients main features encountered were swellings (100%), pain (68%) and ulceration (36%). Ninety percent presented within one year of onset of disease. It is possible that the short duration reflects the rapidity of growth, pain due to nerve involvement and infection of large lesions.

Following adequate investigations and appropriate workup, treatment is individualized for each patient with sarcoma. Surgery, chemotherapy and radiotherapy are treatment strategies for malignancies. Regarding osteosarcoma, surgery is the gold standard for its treatment in the maxillofacial region.[22] while the results of radiation therapy have been unacceptable.[23] Resection with clear margins is the most important prognostic factor in treatment.[24] Smeele et al, [25] reported more favorable outcome in patients without residual disease after surgery. Daramola et al, [10] reported on seven osteosarcomas treated with radical excision (4), surgical excision (1) and inoperable (2). In the maxillofacial region, clear margins are often difficult to obtain due to possibility of such procedures resulting in intolerable morbidity hence suggested margins are 2 to 3cm in bone and soft tissue respectively.[22]

Patients and surgeons need to dialogue to arrive at the acceptable treatment with tolerable side effects. In the light of our current knowledge, only about 32% of the cases treated at our center during the study period received adequate curative surgery i.e. those who had "radical resection ", 50% had no treatment at all due to self-discharge and inoperable cases. During follow-up of the 3 cases radically excised by Daramola et al, [10] all recurred within five years with one having metastases to the lungs, skull and femur despite adjuvant chemotherapy. There was no follow-up record for the cases treated at our center. While there is controversy on the benefit of adjuvant chemotherapy in the treatment of maxillofacial osteosarcoma [14] some insist there is survival advantage in such patients [23,25], though the question of the timing of the administration of drugs remain. [22]

Chondrosarcoma is less common than osteosarcoma from reports. Rafindadi and Ayuba, [7,8] had 12% incidence of chondrosarcoma. This is similar to our result (17%) in Table 1. The lesion occurs more in males than females, ratio 2:1 according to Arlen et al, [26], which is higher but similar to our value of 1.3:1. There was no sexual bias in other works. [27,28]

In the mouth and jaws, the lesion afflicts younger persons than in other parts of the body. [12,14] From five cases reported earlier from our center, Adekeye, [12] found patients` age range was 26-30years (mean 26years). Table 2 showed that cases of chondrosarcoma recorded were between 22-50years (mean 32years). Of our 14 cases, 11(79%) were in the 3rd and 4th decades of life. Primary chondrosarcoma affects the maxilla more than the mandible.[12, 26] The ratio of maxillary to mandibular cases given in an earlier report from our center was 4:1. [12] Almost equal incidences in either jaw (1.1:1) were found in the

cases recorded in this study. It is noteworthy that the earlier series from our center only had five cases, [12] while this series had 10. Larger series are needed to ascertain the actual site predisposition for chondrosarcoma among Nigerians.

In the oral and maxillofacial region, chondrosarcoma presents in a similar manner to other sarcomas. Spread of neoplasm to the skull base can sadly result in blindness, [26,29] that was seen in one of our patients. Unlike other authors, [12,26] pain was prominent (71%) in the chondrosarcoma cases we reviewed. The neoplasm can exhibit rapid growth following tooth extraction in the affected jaw or inadequate tumour excision. [26,30]

Adequate radical surgery remains the primary treatment, [12,26]with tumour size and histological grade as important prognostic factors. Adequate surgery according to Fu and Perzin, [31] is surgical excision beyond the lesion. The use of chemotherapy and radiation treatment either alone or with surgery may cause tumour regression especially when given for recurrent disease. [32] Chondrosarcoma appears to do better than osteosarcoma in the facial region, [33] only 36% could be said to have had adequate treatment of their lesions. Only cases deemed to be inoperable where given chemotherapy. One of such died from malignant cachexia while on chemotherapy. No record of patient follow-up was available in our records. Cohen and Smith, [30], found that 7 of 9 treated patients died from local recurrence and/or remote metastasis while two cases were free of disease for 3months to 3years respectively. Cure is difficult to define in these patients, as recurrence after 13years after treatment has been reported [26] hence life-long follow-ups is necessary.

According to Breddel et al, [16] rhabdomyosarcoma (25%) are next to osteosarcoma in incidence among South Africans. This is twice our result (13%) Rafindadi and Ayuba, [7,8] had no report of the tumour. Slightly more males than females (ratio 1.4:1) reportedly have rhabdomyosarcoma in the maxillofacial region from Young and Miller, [34] and Green and Jaffe.[35] Our 10 cases showed male to female ratio of 1.5:1. There was preponderance of males over females, ratio 5:1 among Indian cases. [36,37]

Rhabdomyosarcomas can occur at any age but the lesion is commonest in the first decade of life making it the commonest maxillofacial sarcoma of childhood. [9] From an analysis of 166 head and neck rhabdomyosarcomas, 77% of cases were 12 years old or younger; half were below 6years old. [38a] Five cases were recorded in India by Pandey et al, [36,37] whose mean age was 16 years (range 4-33years) with 80% in the 1st and 2nd decades. As with other reports, the mean age of the 10 cases we recorded was 16years with 50% of cases in the 1st decade.

Rhabdomyosarcoma in the maxillofacial region is most frequent around the orbit from where spread to the maxilla can occur.[14] Rarely does the lesion occur primarily in the jawbones or other bones in the skull. Two of three cases seen by Pandey et al, [37] were in the soft tissues of the scalp and lip with the last in the alveolus. Most (40%) of the cases seen in this review occurred in the cheeks with the rest in the jawbones of maxilla (20%) and mandible (20%).

It presents as swelling with epistaxis and proptosis due to nasal and orbital involvement in addition to other features. [14] Regarding this lesion, rhabdomyosarcoma presented as ulceration (2), limitation of mouth opening (1) in addition to swelling found in all cases. Pinkel and Pickren, [39] first reported on the benefit of triple modality therapy i.e. combining surgery, radiotherapy and multiple agent chemotherapy in the management of rhabdomyosarcoma. The previously bleak outlook of this condition has been favourably transformed since the triple modality was introduced. [40,41,42] From the Indian subcontinent, three cases of rhabdomyosarcoma benefited from the triple regimen according to Pandey et al. [37] During follow-up for 13-20 months, two succumbed to metastatic disease while another had local recurrence that was successfully managed by salvage chemotherapy. Among six patients, only two had surgical excision and chemotherapy and radiotherapy. Since no follow-up record is available, we cannot speculate on the survival of our treated cases.

Among three forms of sarcoma reported by Rafindadi and Ayuba, [7,8] fibrosarcoma accounted for 38%. This is much higher than our result. There was no sexual bias in 10 fibrosarcomas of the maxillofacial region found in our series which contrasts with the slight female predominance by a ratio of 1.3:1 in the Dutch report by Slootweg and Müller et al. [43] Fibrosarcomas occurs within the ages of 11 - 59 years (mean 32years) from a Dutch study.[43] It shows a definite peak in the first decade of life.[14]

Fibrosarcomas occur in the soft tissues of the maxillofacial region followed by the maxillary sinus, other paranasal sinuses and the nasopharynx. [38b] Slootweg and Müller, 43 considered the lesion in the jaws of a Dutch population and found more in mandible (n=5) than maxilla (n=2). Our 10 cases were distributed between the mandible 60% and palate 20% with few lesions in the other parts. Harrison and Lund, [14] noted that difficulties occur in distinguishing maxillary lesions on the basis of origin from soft tissues such as periosteum or is intraosseuos. They large sizes of tumours seen in this series emphasize this problem. Slootweg and Müller, [43] found that out of seven cases of fibrosarcoma, painful swelling occurred in 3 persons with tooth loosening, pathological fracture, trismus and paraesthesia of the lower lip in one case each. Tumour duration was from a few weeks to 6months. In our patients, median duration of symptoms was 3months with 80% presenting within 6months of onset.

It is generally agreed that wide radical excision is the treatment of choice for fibrosarcoma as no form of limited excision would avoid recurrence [14] and radiotherapy should be reserved for palliation. [44] Three fibrosarcomas in the study by Slootweg and Müller, [43] had resection alone, three others had resection and irradiation ± chemotherapy respectively while one was managed by radiotherapy alone. All died within 6 months – 5.5 years of primary

treatment with local recurrence/metastatic disease. There does not appear to be improved disease-free interval for fibrosarcoma treated using multimodality treatment than those treated by a single modality from the Dutch study possibly due to poor tumour grading. [43] Following from the general acceptance of resection in treating this sarcoma, 30% of our cases had curative treatment though we doubt if any survived without disease for long considering the Dutch experience. One was followed –up for 6months and was disease-free. Life long review is necessary as recurrence after 22years has been recorded. [45]

Tumor rarity makes incidence of malignant fibrous histiocytoma among other oral and maxillofacial sarcomas difficult to find. It accounted for 6% of sarcomas in this study. There are slightly more males than females with malignant fibrous histiocytoma in the maxillofacial region according to Blitzer et al., [46] but we had only males among our patients with this lesion. Harrison and Lund, [14] stated that the lesion occurs mostly in the 6th decade. Cases of malignant fibrous histiocytoma were between 13 and 54 years old (mean 34) according to Pandey et al. [37] In our series, 80% were between 30-50years old (median 40) demonstrating its predilection for older people unlike rhabdomyosarcoma.

Malignant fibrous histiocytoma was found more in the hard tissues such as bone than in soft tissues according to Pandey et al. [37] Out of three lesions they reported, two were in the jaws while one occurred in the scalp. Our five cases were in the mandible (n=3), maxilla (n=1) and lip (n=1) from Table 3. In this series, the lesion presented with bleeding (2) and derangement of teeth (2) while pain was not prominent. These are not dissimilar to features of malignant fibrous histiocytoma in another report. [14] Two cases of malignant fibrous histiocytoma were managed by combination of surgical excision, adjuvant radiotherapy and chemotherapy with survival for 1-65 months after treatment [37], while one was lost to follow-up. Surgical excision is the main treatment for malignant fibrous histiocytoma hence 4 of our cases were adequately managed though no follow-up record was available. Local recurrence after 9years has been reported, [47] hence prolonged follow-up is essential after treatment of malignant fibrous histiocytoma.

Batsakis, [38b] stated that liposarcomas was rarely seen before the age of 30years of age with peak incidence in the 6th decade which is similar to our findings in this report. Like other STS, liposarcomas is essentially a soft tissue malignancy though involvement of craniofacial bones has been recorded. [14] There were five cases of liposarcoma in this review with most in the cheeks (60%) and one in the maxilla (20%). Hudson et al., [11], reported a non-tender mobile mass of the cheek as the first case of liposarcoma at our center in 1978. In this report, lesions seen were swellings (100%), pain and ulceration were rare (20%). Saunders et al., [48] believe liposarcomas are best surgically excised with radiotherapy used for palliation. Only the myxoid form of liposarcoma is believed to respond well to irradiation. [49] Two of the cases we reviewed had wide surgical excision, one each had chemotherapy and radiotherapy respectively. The degree of differentiation of liposarcoma may be directly related to survival. [11]

Especially in developing countries such as Nigeria, poverty, ignorance about medical problems and poorly developed medical infrastructure contribute to morbidity and mortality from malignant conditions such as sarcomas and carcinomas. Within our study period, facilities for external beam megavoltage radiation treatment to the head and neck was not available within 700km radius hence cost and access was real problems in adequate management of sarcomas in Nigeria. While patient management is improved with adequate diagnostic and treatment facilities, health care must be accessible for the population to benefit. To improve the patient survival and freedom from recurrence, there is need for increased cancer awareness and funding for the health sector in Nigeria. Also, regional cancer treatment centers are necessary to cope with the prevalence of malignancies in our environment.

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