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Original Article

Oral and Maxillo-facial soft tissue sarcomas in an Africa population

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Abstract
Background: Soft tissue sarcomas (STS) are not common and account for less than 1% of all malignancies in the maxillofacial region. These tumours are under reported in Africa South of the Sahara in current literature. This report is a review of our experience with STS over a 23 year period at a busy Tertiary Oral care centre in Kaduna, Northern Nigeria.
Methods**:** Patients presenting to the Maxillofacial Unit, of the Ahmadu Bello University Teaching Hospital, Kaduna, with oral and maxillofacial malignancies between the years 1977 and 1999 were retrospectively studied. Soft tissue sarcomas were studied. Demographic features, clinical presentation, treatment options , outcome and histologic variants were studied.
Results: There were 38(<1%) soft tissue sarcomas out of 415 oral and maxillofacial malignancies recorded within the study period. Male to female ratio was 1.6:1. Age range was 24 days to 60 years (median 28 years). There were more adults (79%) than children (21%). There were eleven histologic types but the more frequent ones were rhabdomyosarcoma (10/38,(), fibrosarcoma (10/38 (26%), liposarcoma (5/38 (13%) and malignant fibrous histiocytoma (5/38(13%) . Site of primary occurrence was mainly the mandible (32%), palate (21%) and cheek (18%). Surgery was the mainstay of treatment 28/38 (74%) while 10/38(26%) patients had no active treatment.
Conclusion: Most patients present late with difficult deforming tumours. We can blame patients knowledge, Socio-economic
Key words: soft tissue sarcoma, oral, maxillofacial, poverty, ignorance, treatment, African, management, tumours

Introduction
Soft tissue sarcomas (STS) are solid malignancies mostly of mesodermal origin whose rarity and variety present formidable challenges in characterization and treatment. According to Pisters and Brennan, STS make up less than 1% of all oral malignancies[1]. Like other tumours, they can occur at any anatomical site and at any age. The extremities and trunk are favoured sites for occurrence and only about 5-10% of STS occur in the head and neck region [1, 2.] Some STS are of ectodermal origin but all STS are considered as a group because of the similarities in their clinical features, natural history, treatment and disease outcome. Rhabdomyosarcoma is the most common histologic type of STS in the oral and maxillofacial region, followed by the malignant fibrous histiocytoma (MFH) and the fibrosarcoma 2 . Adults and children are affected and in most reports 80-90% are adults and 10-20% are children 3. Reports from Africa on the clinical behaviour of oral and maxillofacial STS are scanty and are mostly short reports. This study presents oral and maxillofacial soft tissue sarcomas in a Native African population Northern Nigeria in the Savanna belt south of the Sahara desert.

Methods
Medical records of patients with histopathologically diagnosed malignant oral and maxillofacial tumours between January 1977 and December 1999 were retrospectively reviewed at the Maxillofacial Unit, Ahmadu Bello University Teaching Hospital, Kaduna, Nigeria. The Unit is the leading, best staffed and equipped center for the management of oro-facial diseases in Northern Nigeria (estimated population 60 million in 1998). The catchment area of this center is the 19 states in Northern Nigeria. Patient records examined included case filesoperative records and histopathology results. Cases of STS were selected from other malignancies and analysed for age at
presentation, sex, tumour site, clinical features, histopathological type, treatment and follow up

reports. Tumour staging and grading was not done routinely within the study period.

Results From January 1977 to December 1999, there were 406 cases of histopathologically diagnosed oral and maxillofacial malignancies of which 38(<1%) were STS. Mean age of cases of STS was 28.4 ± 15.6years, median age 28.5 years (range 24days to 60years). 79% of patients were above 18 years and (21%) were, < 18 years old. Male to female ratio was 1.6:1. Common oral and maxillofacial sites of STS were mandible (n=12, 32%), palate (n=8, 21%), cheek (n=7, 18%) and the maxilla (n=6, 16%). There were 11 histologic types of STS studied (Table 1). More common were rhabdomyosarcoma (n=10, 26%), fibrosarcoma (n=10, 26%), liposarcoma (n=5, 13%) and MFH (n=5, 13%). The most frequent treatment option of STS was surgery (n=20, 74%). Some patients had chemotherapy (n=6, 22%). Six patients defaulted after confirmatory diagnosis while four tumours were deemed inoperable and patients palliated with analgesics and terminal care.

Table 1 Histologyand sex distribution of oral and maxillo-facial STS

Histologic Type	Sex (% of No)		
	M	F	
Rhabdomyosarcoma	5 (50)	5 (50)	10 (26.3)
Fibrosarcoma	6(60)	4 (40)	10(26.3)
Liposarcoma	3(60)	2(40)	5(13.2)
MFH	5 (100)	-	5(13.2)
Malignant schwan	2(100)	-	2(5.3)
Kaposi's sarco	-	1(100)	1(2.6)
Retinoblastom	-	1(100)	1(2.6)
Malignant giant cell	-	1(100)	1(2.6)
Synovial sarcom	1(100)	-	1(2.6)
Haemangiopericyt	1(100)	-	1(2.6)
Ewing's sarcoma	1(100)	-	1(2.6)
Total	24	14	38

Discussion

Many STS are of mesodermal tissue origin such as rhabdomyosarcoma, fibrosarcoma and liposarcoma while a few are of neuroectodermal origin such as malignant schwannoma and Ewing's sarcoma. STS of the head and neck are quite rare with only a few reports in English literature [4,5,6]. Some case reports from Africa are on specific tumours. Sts make up less than 1 % of all malignancies in the head and neck region and between 5% and 10% of all body sarcomas [1, 2.] In this 23 years survey at our tertiary oral care center in Nigeria, STS was less than 1% (38 cases) of all malignancies of the oral and maxillofacial region confirming its frequently reported rarity. Though no age group is exempt from sarcomas, maxillofacial STS generally affect the younger age group less commonly. According to Eeles et al., [7] STS occurs at a median age of 32 years with 53% of patients being males. Between 80-90% of STS occur in adults while 10-20% of patients are children[3]. The median age of STS occurrence of the 38 cases seen in this study was 28years with fewer less females than males. Male predilection was also observed by other authors 5,6. The most common type of STS differs in different reports. Greagar and colleagues, [8] observed that fibrosarcoma is commonest (27%) followed by malignant schwannoma (15%),while Weber et al., 2 noted that rhabdomyosarcoma (18%) was more common followed by followed by MFH and fibrosarcoma. In his report of 164 adult STS Tran et al.,9 angiosarcoma (16%) was the commonest followed by fibrosarcoma (12%) and unclassified sarcomas (11%). Quite recently, Pandey et al., [10] reviewing 22 adult patients from India found unclassified spindle-cell sarcomas (32%) and MFH and rhabdomyosarcoma (14% each). In our series involving 38 cases, the foremost STS were rhabdomyosarcoma and fibrosarcoma (26% each) and MFH and liposarcoma (13% each) Rhabdomyosarcoma is the neoplastic analogue of the embryogenesis of skeletal muscle. The rhabdomyoblast recapitulates skeletal muscle embryogenesis in a highly disorganised manner 11. The tumour appears to be commoner among Caucasians than Negroes causing speculation of a genetic predilection 12. The paucity of reports on head and neck rhabdomyosarcoma fromAfrica makes these conclusions tenuous. Ten cases of rhabdomyosarcoma seen in this study had a male to female ratio of 1.5:1. Among the 10 cases in this six had histological subtyping available; five were embryonal and one alveolar. The cheek was the commonest site overall (40%) followed by the mandible (20%). Four histologic varieties; pleomorphic, alveolar, embryonal and botryoid types have been described[13]. Among these forms, the embryonal is reportedly commoner in the maxillofacial region most commonly affecting the hard palate[7, 13]. Rhabdomyosarcoma is the most common soft tissue malignancy of the head and neck in children, 14.In the younger adults the tumour is more frequently found affecting the extremities and trunk. In 110 embryonal rhabdomyosarcomas of the body, Enzinger and Shiraki [15] found the lesion occurring between 10 and 20 years of age (mean age 15years). A similar age range was recorded in three cases whose subtype was not disclosed by Pandey et al., [6]. Patients in this study were between 24 days and 40years old (mean 14 years). From the five cases of embryonal subtype in this series, all but one was below 10years of age. Presenting features of oral and maxillofacial rhabdomyosarcoma was frequently as swelling (90%), pain (60%) while ulceration of skin or mucosa was uncommon (20%). These have reported duration was 1.4 months from. According to Maurer et al., [16] a combination of surgery, radiation treatment and multichemotherapy could give dramatic survival rates in patients with rhabdomyosarcoma. This form of treatment has dramatically improved the outlook for these patients. Surgery is the mainstay

while radiotherapy and cytotoxic chemotherapy are adjuvants[16] Pelliteri et al., [17] believe that while the influence of the size of head and neck STS on prognosis is controversial; the possibility of successful en-bloc resection of tumour is limited in large tumours due to involvement of critical neurovascular structures. Hence, poor local control and survival would accompany surgical therapy of large lesions in the head and neck. In this report, four cases were treated surgically, three with cytotoxic chemotherapy and one case referred for radiotherapy . Out of the four cases treated surgically, only that treated by resection could be said to have had optimum initial treatment (Cases no 9) though no review reports were available.Also, during this study period, facilities for head and neck radiotherapy were not available within a 700km radius of our center while the cost of radical surgery with need for reconstruction remains unaffordable to many of our patients unlike the Indian cases reported by Pandey et al., [6] It should be noted that there is no functional health insurance scheme for most citizens of sub-Saharan Africa including Nigeria. Despite the gruesome sizes on presentation of many tumour cases in Nigeria [18], cases of default after diagnosis and inoperable cases abound. This is due to ignorance of medical problems among patients and their relatives, poor access to modern health care facilities, native superstitions forbidding surgical treatment and the social problem of poverty. Hence, the combination of ignorance, poverty and inadequate medical infrastructure limit treatment options and survival of STS patients in our environment. The impact of treatment on survival of the cases seen in this report could not be assessed, as follow-up review visits was poor in this study. Stout[19] states that 45% of fibrosarcomas occur between the ages of 20 and 40 years while Dahlin and Ivins [2.0] found 13 of 114 central bone cases of the lesion originating in the mandible. There were 10 cases of fibrosarcoma seen at Kaduna, Nigeria occurred between 8 and 60 years of age with 70% below the age of 40years.Among these cases , the mandible was the commonest site (60%) followed by the palate (20%). Fibrosarcoma grows rapidly or slowly [13]. Duration ranging from 3-72 months was recorded in this study; most cases (80%) however, presented within 6months. The most acceptable form of treatment for fibrosarcoma is radical resection. Any form of limited excision especially due to its atimes pseudo encapsulation inevitably leads to recurrence [13,21.] Eight cases were managed during the period of this study especially by surgery (n=6). One case managed surgically by tumour resection was followed up for 6months with no record of recurrence. It is noted that this follow-up period is too short to assess recurrence. No survivor was recorded 5years after treatment of fibrosarcoma of the jaws in the Netherlands [22..] Even though fat tissue is abundant in the oral and maxillofacial region, liposarcoma in this region is exceptionally rare. Between 1958 and1965, only four cases were seen in southern England [23.] . Hudson et al., [24], reported the first case of liposarcoma from our center in 1978. This tumour is rare below the age of 30 years and shows a slight male preponderance (55%) according to Enzinger and Winslow, [25] . Table 4 shows 80% of the cases in this study where above 30 years old while the male to female ratio was 1.5:1.

Liposarcoma occurred mostly as cheek swellings (60%), the rest were in the palate (20%) and maxilla (20%) in .Several histological sub classifications of liposarcoma have been described. One classification describes the cell types as myxoid, round cell, adult and pleomorphic [26.] . It has been noted that majority (71% of 35 cases) of liposarcomas of the maxillofacial area are of the myxoid cell type, are well differentiated with good prognosis [27.] Among our five cases, 60% were of the myxoid variety . Surgical excision with or without radiotherapy is used for treating liposarcoma with prognosis related to several factors especially tumour cell type [13.] Four cases of liposarcoma were treated, two by surgery and one with chemotherapy. One patient was referred for radiotherapy. No follow-up record of treated cases was available. MFH is regarded as one of the commonest STS of later life[28] where it affects the musculature of the limbs. It is rare in the maxillofacial region. In a large series of 200 cases, Blitzer et al., [29] found only six in the head and neck. According to Pandey et al., [6], all three cases of MFH from a Regional Cancer Centre in India occurred between 13 and 54 years of age, all are males. Table 2 shows that 80% of cases of MFH seen at Kaduna, Nigeria, were above the third decade of life, (mean age 39.4years). All our cases were also males. This raises the possibility of male predilection for occurrence. The tumour is believed to occur mostly in the sixth decade of life [21.] The tumour is rare in the oral soft tissues; most are central lesions of the jawbones[30.], In the report by Pandey et al., [6], 66% of cases of MFH were in the jawbones. Four of the five cases in our series occurred at the jawbones similar to the findings of Pandey et al., [6] Radical local excision is the most useful form of treatment for this lesion [2.1] Malignant schwannoma can occur *de novo* or less frequently from pre-existing lesions of neurofibromatosis. While cases occurring *de novo* present later in life, those arising secondarily are generally seen in adolescents and young adults[13.] . Cases in this report had no history of malignant schwannoma arising secondarily. The two cases seen were males aged 46-48years. Any of the oral soft tissues such as the lip, palate, gingiva or the hard tissues such as the jawbones can be involved in the lesion. Patients managed in this report presented with swelling, pain and ulceration of the palate and nostril.. Case no 6 recurred within two months of resection of tumour. Patient then opted to leave the hospital for home care. Prognosis of malignant schwannoma varies greatly and factors responsible are tumour location, degree of differentiation, duration at presentation, and method of treatment. Tumours secondary to von Recklinghausen`s disease have a worse outcome compared to primary tumours[13]. Recurrence following local excision can be up to 80% with a 5year survival between 15-30% [31]. Adjuvant radiotherapy and

chemotherapy do not improve the efficacy of local control and long-term prognosis,[21.. The management of STS requires close collaboration between an experienced head and neck surgeon, medical oncologist, pathologist, radiation oncologist and diagnostic radiologist in a comprehensive cancer centre[6]. There is a role for the oral/maxillofacial surgeon and prosthodontist in assessment and follow-up care of some of these cases. The guarantee for optimum patient treatment is lacking in much of sub-Saharan Africa. Better patient care would result from the establishment and equipping of regional centres for the management of malignancies as available in most developed world and Indian subcontinent. Only by this can our patients escape the devastation form the relatively uncommon STS

Conclusion The frequency of STS in Nigeria (1%) is comparable to that in other parts of the world. However the quality of life of patients and survival after treatment is not comparable. Better patient care would be possible with the establishment of regional centers for the management of malignancies and to benefit from the skilled manpower in Africa. Patient education should be encouraged to seek treatment early in the course of disease to reduce the problems of delayed management of malignancies. Patients follow up should be meticulous and structured to catch early those who recur post intervention.

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