Original Article

Orofacial Soft Tissue Sarcoma: A Review of 36 Cases in Enugu, Nigeria.

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ABSTRACT

Orofacial soft tissue sarcomas (STSs) are malignant mesenchymal tumours in the oral and maxillofacial region. There has been only one primary study in Nigeria, and the epidemiology in Enugu, southeast Nigeria is unknown. The objective is to study the prevalence, distribution and features of orofacial soft tissue sarcomas at a tertiary hospital in Enugu, Southeast Nigeria. This is a 10-year (2012 to 2021) retrospective study of consecutive patients with orofacial soft tissue sarcomas presenting at a tertiary hospital in Enugu. Thirty-six (36) orofacial soft tissue sarcomas (STSs) were diagnosed out of 897 orofacial lesions. The prevalence among all orofacial biopsies, orofacial malignancies and orofacial sarcomas were 4.0%, 13.6%, and 76.6% respectively. Fibrosarcoma 36.1% (n=13) and rhabdomyosarcoma 19.4% (n=7) were most prevalent. Orofacial STSs presented a male to female ratio of 1.1 to 1, a mean age of 39.3 ± 19.7years, and were mostly diagnosed at the second and fifth decades, 25.2% (n=7). The mean age and duration among male patients were 33.8 ±19.9years, and 7.7 ± 6.7 months respectively, while for the female group: 44.7±18.1years and 11.1±6.9months respectively. The orofacial STSs were distributed as follows: mandible 36.1% (n=13), maxilla 33.3% (n=12), palate and cheek each had 8.3% (n=3). Pain 69.4% (n=25), bleeding 27.8% (n=10), and recurrence 19.4% (n=7) were often observed. In conclusion, orofacial STSs were infrequent and mostly involved the fibrous and skeletal tissues. The mean age of occurrence, especially among women is higher in Enugu.

Keywords: Fibrosarcoma, Head and Neck, Orofacial, Rhabdomyosarcoma, Soft tissue sarcoma

INTRODUCTION

Soft tissue sarcomas (STSs) develop from soft tissue components of connective and subcutaneous tissues, which include adipose, muscular, neural, fibrous and vascular tissues.[1] Sarcomas constitute 1–2% of total body malignances,[2] out of which approximately 80% are soft tissue sarcomas.[3] The aetiology of sarcomas is largely unknown.[4] Soft tissue, bone (osteosarcoma, chondrosarcoma and Ewing's sarcoma), and visceral sarcomas are the three classes of sarcomas.[1] HIV-related Kaposi's sarcoma contributed to the increased prevalence of sarcomas reported in some studies following the HIV pandemic.[5,6]

Studies of head and neck sarcomas,[7] and those on orofacial malignant neoplasms in Nigerian literature did not highlight the specific epidemiologic features of orofacial soft tissue sarcomas.[9,10,11] Most epidemiologic studies on orofacial sarcomas

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evaluated both soft and bone sarcomas as one group without distinction, thereby making them unspecific as a reference material.[12,13] An earlier study in Enugu jointly analyzed soft and bone sarcomas of the head and neck region with emphasis on the management challenges.[11] To the best of the authors knowledge, only one Northern Nigeria study, in Zaria, primarily focused on orofacial STSs as a distinct group.[14] This epidemiological study in a tertiary hospital in Enugu would add to the Nigerian literature on the subject and provide a reference on the burden of the disease in Southeast region.

MATERIALS AND METHODS

This is a 10-year (April 2012 to March 2022) retrospective observational study of patients at a tertiary hospital in Enugu, with orofacial soft tissue sarcomas (STS) based on histologic diagnoses. Their biodata and clinicopathologic information were obtained from the biopsy forms, histopathology reports and case files archived in the Records department of the hospital. The data included: age at presentation, gender, site of tumour, duration, complaint of pain, lymph node involvement, ulceration, bleeding, histologic type, and recurrence. These were used to determine the prevalence, mean age, gender ratio and distributions, histological groups and anatomic location. Other primary sarcomas of head and neck region, of bone and other anatomical regions outside the orofacial tissues, and metastatic sarcomas were excluded.

Some cases diagnosed as sarcomas but without a conclusive histologically satisfactory elucidation of the histiogenicity were left as unclassified soft tissue sarcomas, pending a confirmatory diagnosis by immunohistochemistry (IHC) marker panels. This study received the institutional approval of the Research Ethics Committee of the College of Medicine of the University of Nigeria, with the Protocol Number**:** 0147/04/2022

Statistical Product for Service and Solution (IBM SPSS version 23) was used in the statistical analysis. The data was analyzed using descriptive statistics while the test for a statistical association between the variables at a 95% confidence interval was carried out using the ChiSquare test.

RESULTS

A total of 36 cases of orofacial soft tissue sarcomas (STSs) were diagnosed out of 897 orofacial biopsied lesions, Tables 1 and 2. The prevalences of orofacial STSs were 4.0% of all orofacial biopsied lesions (36/897), 13.6% of all orofacial malignances (36/264) and 76.6% of all orofacial sarcomas (36/47). Table 3A shows the 2nd and 5th decades with the highest frequency of cases. Fibrosarcoma 36.1% (n=13) was the most frequently diagnosed soft tissue sarcoma followed by rhabdomyosarcoma 19.4% (n=7), Table 3B. Majority of orofacial STSs were derived from the fibrous and muscular tissues and jointly constituted 56.5% (n=20) of the tumours. Some six cases previously diagnosed as angiosarcoma by microscopic study were later disproved by confirmatory immunohistochemistry studies and grouped with three other sarcoma cases with uncertain histogenicity as unclassified soft tissue sarcomas 25.0% (n=9). The orofacial STSs were distributed mostly in the mandible 36.1(n=13) and the maxilla 33.3% (n=12), Table 3C.

Table 4 shows the gender distribution and ratio with a male to female ratio of 1.1 to 1, as well as the overall mean ages. The mean age of male participants at onset of tumour was 33.8 ± 19.9 years, while the mean age of the female patients was 44.7 ± 18.1 years.

Pain was reported in more than half the cases, while lymph node involvement (matted, and unspecified), bleeding, recurrence and two cases of distant metastases are shown in Table 4. There were no statistically significant associations between gender and recurrence P=0.3, sites and ulceration P=0.5, site and recurrence P=0.199, histological diagnosis and recurrence P=0.206.

Table 1: Orofacial soft tissue sarcomas in the upper jaw soft tissues(n,20)

S/N	Sex/ Age (years)	Duration (months)	Site	Pain	Ulceration	Lymph Node Involvement	Bleeding	Histological Diagnosis	Recurrence+metastases
1	F/68	12	parotid /buccal	yes	no	yes (matted)	no	melanoma	yes, + scalp
2	F/62	2	maxilla	yes	yes	no	no	rhabdomyosarcoma	nil
3	F/59	4	maxilla	yes	no	no	no	messenchymal sarcoma	no
4	F/37	7	maxilla	no	no	no	no	unclassified	nil
5	F/35	6	cheek	yes	no	no	no	rhabdomyosarcoma	nil
6	F/17	24	upper cheek	yes	yes	no	no	rhabdomyosarcoma	yes
7	F/46	12	cheek	yes	yes	no	no	pleomorphic fibrosarcoma.	nil
8	F/17	24	mid face	no	no	no	yes	myxofibrosarcoma.	nil
9	F/67	4	maxilla	yes	yes	no	no	rhabdomyosarcoma	nil
10	F/29	10	maxilla	yes	yes	yes	yes	pleomorphic fibrosarcoma.	nil
11	M/40	7	palate	yes	no	yes (matted)	yes	melanoma	Yes + lung
12	M/25	3	maxilla	yes	no	no	yes	myxofibrosarcoma	nil
13	M/57	24	maxilla	no	yes	no	no	myxofibrosarcoma	nil
14	M/49	3	maxilla	yes	no	yes	yes	unclassified	nil
15	M/58	6	maxilla	no	yes	no	no	myxofibrosarcoma	nil
16	M/53	12	maxilla	no	no	no	yes	unclassified	nil
17	M/9	9	maxilla	no	no	no	no	rhabdomyosarcoma	yes
18	M/10	12	maxilla	yes	no	no	no	rhabdomyosarcoma	nil
19	M/58	3	palate	yes	no	no	no	fibrosarcoma	nil
20	M/58	5	palate	no	no	no	yes	malignant melanoma	nil

Table 2: Orofacial soft tissue sarcomas in the lower jaw soft tissues (n,16)

S/ N	Sex/ Age (years)	Duration (Months)	Site	Pain	Ulceration	Lymph Node Involvement	Bleeding	Histological Diagnosis	Recurrence
1	F/20	22	mandible	yes	no	no	no	mesenchymal sarcoma	nil
2	F/72	5	mandible	yes	yes	yes (fixed)	yes/ pus	myxofibrosarcoma	yes
3	F/47	9	mandible	yes	yes		yes	unclassified	nil
4	F/41	12	mandible	yes	no	no	yes	fibrosarcoma	yes
5	F/48	12	mandible	no	no	yes	no	myxofibrosarcoma	nil
6	F/50	5	mandible	yes	no	no	yes	myxofibrosarcoma	nil
7	F/45	3	tongue	yes	yes	no	no	rhabdomyosarcoma	nil
8	M/16	12	mandible	no	yes	no	yes	rhabdomyosarcoma	nil
9	M/14	7	mandible	yes	no	no	no	fibrosarcoma	nil
10	M/19	2	mandible	yes	yes	yes	no	unclassified	nil
11	M	3	lower lip	yes	no	no	no	haemangiopericytoma	no
12	M/40	6	submandible	yes	no	yes	yes	unclassified	nil
13	M/25	24	mandible	yes	no	no	no	unclassified	yes
14	M/7	2	tongue	yes	no	no	no	unclassified	nil
15	M/20	NA*	mandible	no	no	no	no	fibrosarcoma	nil
16	M/66	1	mandible	yes	yes	no	no	unclassified	nil

NA: Not available

Table 3: Age Group histologic diagnosis and site of orofacial soft tissue sarcomas

A)Age Group	% (n)	B)Histological diagnosis	% (n)	C)Site	% (n)
0-10 years	10.8 (3)	fibrosarcoma	36.1(13)	mandible	36.1(13)
11-20 years	25.2 (7)	unclassified	25.0 (9)	maxilla	33.3 (12)
21-30 years	10.8 (3)	rhabdomyosarcoma	19.4 (7)	cheek	8.3 (3)
31-40 years	14.4 (4)	malignant melanoma	8.3 (3)	palate	8.3 (3)
41-50 years	25.2 (7)	mesenchymal sarcoma	5.0 (2)	mid-face	2.8 (1)
51-60 years	21.6 (6)	haemangioendothelio ma	2.8 (1)	parotid area	2.8 (1)
61-70 years	14.4 (4)	haemangiopericytoma	2.5 (1)	lower lip	2.8 (1)
>70 years	3.6 (1)	**Total**	**100.0 (36)**	submandibular	2.8 (1)
NA*	3.6 (1)			tongue	2.8 (1)
Total	**100.0(36)**			**Total**	**100 (36)**

*NA: Not available

Table 4: Epidemiologic and clinical features of orofacial soft tissue sarcomas

Site	% (n)
Male	52.8% (19)
Female	47.2% (17)
Male:Female Ratio	1.1 : 1
Overall mean age	39.26 ± 19.7 (range: 7 to 72) years
Male Mean age	33.8 ± 19.9 (range 7 to 66) years
Female mean age	44.7 ± 18.1 (range 17 to 72) years
Overall mean duration	9.3 ± 6.9 months
Male mean duration	7.7 ± 6.7 months
Female mean duration	11.1± 6.9months
Pain reported	69.4% (25)
Bleeding	27.8% (10)
Recurrent tumour	19.4% (7)
Distant Metastases	5.6% (2)
Total	**100 (36)**

DISCUSSION

This study observed that the prevalence of orofacial soft tissue sarcomas (STSs) among all orofacial biopsies is 4 %, while fibrosarcoma 36 % and rhabdomyosarcoma 19 % were the most common orofacial soft tissue sarcomas (STSs). The orofacial STSs were distributed mainly in the mandible 36 %, and maxilla 33 %, while pain 69 %, and bleeding 27.8% were the most common features.

This study is among the few on orofacial soft tissue sarcomas (STSs) in Nigeria because previous general reports on sarcomas did not distinguish the epidemiologic features of soft tissue from bone sarcomas.[13,15,16] Similarly, the reports on head and neck sarcomas mostly provided non-specific information on orofacial STSs.[8,11] However, for the purposes of this study, estimated prevalence values and some other clinical features were deduced from some of these publications.[13,15,16] The orofacial STS prevalence of 4.0% in this study falls within the estimated range of 1.8% to 5.5% of all orofacial biopsies in Nigerian literature,[14,15,16,] and the 5.4% reported in Tanzania.[17] The STSs prevalence of 13.6% among orofacial malignances is within the estimated range of 8% to 15.9% from the Nigerian literature,[12,15] and similar to the 13.1% reported in Kenya.[18] The STSs prevalence of 76.6% among orofacial sarcomas is higher than the 42% documented in Kenya,[18] but similar to the 77.3% reported in Tanzania,[17] and within the estimated Nigerian range of 50% to 80.4%.[12,15,16]

Fibrosarcoma and rhabdomyosarcoma continue to be the two most frequently reported orofacial STS in Nigeria.[13,14,15,16] Fibrous derived orofacial STSs were most prevalent with 32.5% in this study, similar to another Nigerian study in Benin that reported a high frequency of 53.5%.[15] In contrast, other studies noted more sarcomas of muscle tissue origin in the form of rhabdomyosarcoma.[12,13,14,16] The reason could be that both fibrous and muscular tissues are probably among the most abundant soft tissues in the orofacial region, with abundant supporting fibrous tissue framework in the extensive fascia surrounding various tissues and organs. These two groups of tissues are probably the most exposed to masticatory motion, facial expressions, and thermal changes, with the fibrous components also giving rise to various forms of fibromas and fibroosseous lesions.[19,20] The authors are unaware of any correlation between the high prevalence of benign fibrous and fibroosseous lesions to the high frequency of fibrous sarcomas in the orofacial soft tissue region. In contrast, there is a high frequency of rhabdomyosarcomas despite the rarity of rhabdomyomas in the orofacial region, unlike in the cardiac muscles.[21,22] Orofacial sarcomas need not be preceeded by benign lesions of the same tissue origin but could also be associated with infections as

reported in countries where the HIV pandemic resulted in the predominance of a vascular malignancy, orofacial kaposi sarcoma (KS).[6,8,17,18] In countries with high prevalence of KS, orofacial fibrous and muscular sarcomas appeared remarkably low in prevalence, especially with rhabdomyosarcoma reported as 2.6% in Uganda,[6] 12.6% in Tanzania,[17] and 14.5% in Kenya.[18]

The slight higher male ratio of 1.1 to 1 in this series is in concord with reports in Northern Nigeria where 1.3:1 and 1.7:1 were reported in Kano,[14] and Kaduna,[12] respectively. This contrasts with reports of higher female ratios of 1:1.1 in Uganda and 1:1.4 in Tanzania,[6,17] all probably attributed to HIV-associated kaposi sarcoma most frequent in women in both countries. The overall mean age at clinical presentation of 39 years in this study is higher than the 29 years reported in Kano Nigeria,[14] 30 years in Uganda,[6] and 33 years in Tanzania.[17] The lower mean age in Kano Northern Nigeria may be attributed to early presentations of cases aged seven months.[14] The higher mean age in Enugu may be due to observed late onset of orofacial STSs in most female patients (44.7 years). The long mean duration of orofacial STSs of 9.3 months before clinical presentation may have been due to poor awareness, and limited access to the few orofacial specialist care centres in Southeast Nigeria,[19] and the observed longer mean duration (11.1 months) of STSs in the women. All these reasons may be partly responsible for the advanced stage of tumours at clinical presentation and poor management outcome.[11]

The 2nd and 5th decades of life had the most cases, 25.2% in this series and the 2nd decadewas similarly reported by Fomete et al.[14] The higher distribution of STSs in the soft tissues of the mandible at 36.1(13) in this series, concurs with two reports from Northern Nigeria (Kano and Kaduna) where the mandible was the most prevalent site at 45%,[14] and 37.2% respectively.[12] The equal distribution of STSs in the palate and cheek 8.3% as the third most common sites, differs from reports that gave higher prevalence to both sites in other studies and countries. The palate was the most common site in countries with orofacial KS such as Kenya, Uganda and Tanzania with high prevalence of HIV/AIDS.[6,17,18] while the cheek was the second most common site at 25% and 18.6% in Kaduna and Zaria, Nigeria respectively.[12,14]

Complaints of pain, bleeding, lymph node involvement, ulceration, and recurrence are among the signs and symptoms frequently reported in orofacial STSs including this study.[14] The observed cases of regional (matted nodes) and distant metastases were from two oral melanoma tumours. Distant metastases from rhabdomyosarcoma especially the alveolar type are known in literature though none was observed in this study.[23] The accompanying orofacial deformity, morbidity and complications make their management challenging. Orofacial STSs require combination therapy and multidisciplinary management that often include surgery and radiotherapy.[14] KS cases are mostly managed by highly active antiretroviral therapy (HAART), and chemotherapy.[6] Nevertheless, most of the orofacial STS patients in Nigeria are often lost to follow-up and death.[11]

CONCLUSION

Orofacial STSs were infrequent and mostly involved the fibrous and skeletal tissues. The mean age of occurrence, especially among women is higher in Enugu. More regional studies are needed to highlight the epidemiology, probable aetiology and management of this group of tumours. The immunohistologic diagnosis and management of these tumours are a challenge in our environment.

LIMITATION

Immunohistochemical investigations to confirm or type the tumours were unfortunately dependent on the financial resources of the patients and this greatly limited the aid which immunohistochemistry (IHC) marker panels would have provided. Though only some six cases had IHC done which excluded some histologic diagnoses, the rest of the lesions did not undergo immunohistochemical confirmatory testing due to financial constraints on the patients. Tumours diagnosed as unclassified soft tissue sarcomas were similar to the reported 21% undifferentiated sarcomas among the STSs in the head and neck region.[7]

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Nil

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