

Original Research Article

Clinicopathological pattern of soft tissue sarcoma in a tertiary health institution in North Western Nigeria

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ABSTRACT

Background: Soft tissue sarcomas are a rare group of heterogeneous tumours that consist of several neoplasia which differentiate into different cell lines. They could arise from the mesodermal or ectodermal embryonic germ layers. Rhabdomyosarcoma is the most common soft tissue tumour in the paediatric age group in most studies, while in adults, malignant fibrous histiocytoma, liposarcoma and fibrosarcoma are seen to be more predominant. The study aims to highlight the histological patterns, clinical presentation, associated risk factors and the patients' performance status at presentation.

Methods: This is a 5-year retrospective between the period of 2010 to 2015 of all cancer patients diagnosed with soft tissue sarcoma, seen in the department of radiotherapy and oncology, a tertiary health centre in Sokoto Nigeria. Data collected included socio-demographic, histological type, clinical features, and performance status.

Results: A total of 123 patients were reviewed during the study period of 2010 to 2015: males were 81 (65.9%) and females were 42 (34.1%), with a male to female ratio of 1.93:1. The commonest histological type in both the adult and paediatric age group was rhabdomyosarcoma. Most patients presented with a swelling as seen in 62 (50.4%) cases. The extremities were the commonest site of involvement, with the lower limbs consisting 41 (33%) of the total patients reviewed.

Conclusions: Rhabdomyosarcoma was the most predominant histological type of soft tissue sarcoma seen over the review period and was seen in both the paediatric and adult age groups as the commonest type.

Keywords: Sarcoma, Soft tissue, Tumour

INTRODUCTION

Soft tissue sarcomas are a heterogeneous group of mesenchymal tumours which consists of 1% of all adult malignancies and about 12% of pediatric cancers.^{1,2} It can arise from both the mesodermal and ectodermal layers and has the capacity to mature into several adult cell lines/ tissues which include striated and smooth muscles, adipose tissues, fibrous tissues, among others. There are

also those whose line of differentiation are not clearly defined.^{3,4}

The classification of soft tissue tumours includes adipocytic tumours, fibroblastic/myofibroblastic tumours, so called fibrohistiocytic tumours, smooth muscle tumours, pericytic tumours, gastrointestinal stromal tumours, nerve sheath tumours, tumours of uncertain differentiation and undifferentiated sarcomas.³

Malignant fibrous histiocytoma is the most common soft tissue sarcoma globally, though there are other studies where liposarcoma was seen as the commonest histological type.^{5,6}

A pathological grouping of soft tissue sarcomas in a study showed malignant fibrous histiocytomas to be 34.2% of the histological type seen, followed by synovial sarcoma (17.1%), liposarcoma (16.3%) and rhabdomyosarcoma (12.6%). Fibrosarcoma was however the commonest soft tissue sarcoma in another study followed by malignant fibrous histiocytoma.⁷ Rhabdomyosarcoma is the commonest paediatric soft tissue sarcoma seen but was also the commonest type in a study in Niger delta region of Nigeria in both the adult and paediatric age group.^{8,9} A review of soft tissue sarcoma over a 20 year period showed the peak incidence of age occurred in the third and sixth decades of life, while another study showed the mean age of presentation of soft tissue sarcoma to be 37.4±12.6 years and the age range from 18 to 85 years among adult population but with inclusion of paediatric age group in another study, the youngest patient seen was 3 years and the oldest was 73 in another study.⁸

There is a slight gender predilection seen in soft tissue sarcoma in males compared to females. This is also seen in a number of studies in Nigeria that showed soft tissue sarcoma to be commoner among the males than females in all age groups.¹⁰⁻¹²

Risk factors of soft tissue sarcoma include environmental exposure to carcinogens which include vinyl chloride, herbicides and pesticides. It also includes genetic susceptibility, chronic lymphoedema, previous radiation exposure and interaction between these factors. Immunosuppression is also a risk factors for soft tissue sarcomas.^{13,14} HIV associated malignancy have increased risk for patients having soft tissue sarcoma. Kaposi sarcoma is a soft tissue sarcoma seen in patients with AIDS.^{15,16} However, most cases of soft tissue sarcoma do not have an identifiable cause.¹³

The most affected site of the body of presentation of soft tissue sarcoma are the extremities with the lower limbs constituting most of the cases seen, followed by the lower limb while the least affected site was the retroperitoneum.¹⁷ There are varied predilection site based on the different classes of soft tissue sarcoma.⁸ A similar finding was seen in another study where the extremities was the commonest site which was closely followed by the trunk but the retroperitoneum constituted 15%, while the head and neck region was 9%.^{18,19} The performance status of cancer patients at presentation affects the prognosis of the disease as patients with poor performance status have worse survival, due to poor tolerance to treatment.^{20,21}

There is an increasing number of soft tissue sarcoma in our environment, though breast and cervical cancer

comprises the greatest proportion of cancer patients, soft tissue sarcoma consists of a significant number of patients seen in Nigeria. Due to the wide heterogeneity of soft tissue sarcoma, characterization of this group of malignancy to know the pattern, identify the risk factors and highlight the presenting symptoms of patients which could be easily identified for prompt referral, diagnosis and treatment.

METHODS

Study area

The study was conducted in the department of Radiotherapy and Oncology, Usmanu Danfodiyo university teaching hospital, Sokoto state, Nigeria. The hospital is located in the North western part of Nigeria and receives referrals to the Department of Radiotherapy from health institutions of neighbouring states. These include Zamfara, Kebbi, Kano, Katsina as well as from other parts of the country. These referrals are mainly due to paucity of oncology facilities in those regions.

Study design

This is a five-year retrospective study of all patients with soft tissue sarcoma seen in the department of radiotherapy of the Usmanu Danfodiyo university teaching hospital between the periods (1st January 2010 to 31st December 2015).

Data was collected from cases notes, histology reports and treatment cards. Data collected included; socio-demography, presenting symptoms, site of soft tissue sarcomas, and performance status of the patients at presentation

Inclusion criteria

All patients seen with soft tissue sarcoma.

Exclusion criteria

- All patients with bone sarcoma.
- All patients without a histological confirmation of soft tissue sarcoma.

Data analysis

Data was analyzed using SPSS version 21.0. All continuous variables were assessed for normality and presented as means ±standard deviations (SD). Qualitative variables will be summarized as tables and charts.

RESULTS

Table 1 shows the socio-demographic data of the patients. There was a predominance of the male gender which was seen in 81 (65.9%), while females were 42

(34.1%) of the total 123 patients seen during the study period. The majority of the patients were between the age group 21-30 years seen in 26 (21.1%) patients, followed by 51-60 years with 23 (18.7%) patients, with 1 patient seen was over 70 years. The mean age of the patients was 36.1 ± 18.0 years.

Table 1: Socio-demographic characteristics of the patients.

Characteristics	Frequency (%)
Gender	
Male	81 (65.9)
Female	42 (34.1)
Age group (years)	
1-10	12 (9.8)
11-20	16 (13.0)
21-30	26 (21.1)
31-40	19 (15.4)
41-50	20 (16.3)
51-60	23 (18.7)
61-70	6 (4.9)
>70	1 (0.8)
Occupation	
civil servant	27 (22.0)
Trader	37 (30.1)
Housewife	22 (17.9)
Others	37 (30.1)
Level of education	
No level of Education	47 (38.2)
Primary	32 (26.0)
Secondary	18 (14.6)
Tertiary	26 (21.1)
Total	123 (100.0)

Mean age is 36.1 ± 18.0 years, Male to female ratio 1.93:1

Table 2: Risk factors for soft tissue sarcoma.

Risk factors	Frequency
Chemical agents	
Yes	3 (2.4)
No	120 (97.6)
Family history	
Yes	15 (12.2)
No	108 (87.8)
Total	123 (100.0)

Most of the patients in table 1 were petty traders which comprises of 37 (30.1%) persons while significant percentage of 17.9% (22) were housewives. Others include professionals and semiskilled professionals/artisans like carpenters, plumbers, tailors, and farmers. Forty-seven (38.2%) patients had no formal level of education, while 32 (26.0%) and 18 (14.6%) patients had primary and secondary level of education

respectively. Only 26 (21.1%) had attained tertiary level of education as seen in Table 1.

The risk factors as shown in Table 2 identified chemical carcinogens in 3 (2.4%) patients, while 15 (12.2%) patients had a first or second-degree family history of cancer but not necessarily soft tissue sarcoma.

Table 3: Histological types of soft tissue sarcoma.

Histology	Frequency (%)
Angiosarcoma	2 (1.6)
Chondrosarcoma	3 (2.4)
DFSP	15 (12.2)
Esthesioneuroblastoma	2 (1.6)
Fibrosarcoma	9 (7.3)
Kaposi sarcoma	6 (4.9)
Leiomyosarcoma	5 (4.1)
Liposarcoma	7 (5.7)
MFH	4 (3.3)
MPNST	19 (15.4)
Rhabdomyosarcoma	48 (39.0)
Synovial sarcoma	3 (2.4)
Total	123 (100.0)

Table 4: Histological types of patients with soft tissue sarcoma with gender distribution.

Tumour type	Sex		
	Male	Female	Total
Angiosarcoma	1	1	2
Chondrosarcoma	3	0	3
DFSP	11	4	15
Esthesioneuroblastoma	1	1	2
Fibrosarcoma	2	7	9
Kaposi	6	0	6
Leiomyosarcoma	3	2	5
Liposarcoma	5	2	7
MFH	3	1	4
MPNST	16	3	19
Rhabdomyosarcoma	27	21	48
Synovial sarcoma	3	0	3
Total	81	42	123

Rhabdomyosarcoma was the commonest soft tissue sarcoma seen in the total patients reviewed, 48 (39.0%); of which 27 (56.3%) were males and 21 (43.7%) were females.

This was followed by malignant peripheral nerve sheath tumour in 19(15.4%) patients, of which males comprised 15 (78.9%), and 3 (21.1%) patients were females. Fifteen (21%) of the patients had dermatofibrosarcoma protuberance for which males were 11 (73.3%) and females were 4 (26.7%). Other histological types seen were fibrosarcoma seen in 9 (7.3%) patients, liposarcoma 7 (5.7%), Kaposi sarcoma 6 (4.9%), leiomyosarcoma 5

(4.1%), malignant fibrous histiocytoma 4 (3.3%), extraskeletal chondrosarcoma 3 (2.4%), synovial sarcoma 3 (2.4%), while angiosarcoma and esthesioneuroblastoma were only seen 2(1.6%) patients respectively as seen in Table 3 and 4.

Rhabdomyosarcoma was the most common histological type seen in both the paediatric and adult age groups and comprised 20 (41.7%) and 28 (58.3%) respectively. See Table 5.

Table 5: Histological types of soft tissue sarcoma with age group distribution.

Histology type	Age group								Total
	1 - 10	11 - 20	21-30	31-40	41-50	51-60	61-70	71 - 80	
Angiosarcoma	0	1	1	0	0	0	0	0	2
Chondrosarcoma	0	1	1	1	0	0	0	0	3
DFSP	0	0	2	4	8	0	1	0	15
Esthesioneuroblastoma	0	0	2	0	0	0	0	0	2
Fibrosarcoma	0	1	2	1	2	3	0	0	9
Kaposi	0	0	2	3	0	0	1	0	6
Leiomyosarcoma	0	0	0	0	2	3	0	0	5
Liposarcoma	0	1	2	1	0	2	1	0	7
MFH	0	0	1	2	1	0	0	0	4
MPNST	1	2	3	1	4	5	2	1	19
Rhabdomyosarcoma	11	9	10	5	3	9	1	0	48
Synovial sarcoma	0	1	0	1	0	1	0	0	3
	12	16	26	19	20	23	6	1	123

Most patients presented with a swelling which as seen in 62% (76) of the patients, while 40% (49) had pain, 9% (11) ulcerative lesion and 4% (5) presented as an incidental finding as depicted in Figure 1.

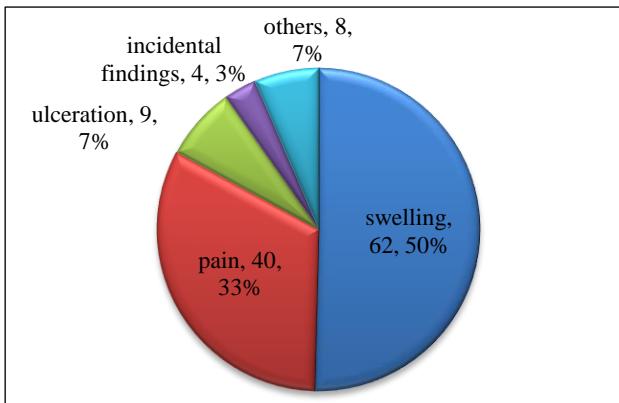


Figure 1: Symptoms experienced by the patients at first presentation.

The extremities were the commonest primary site of soft tissue sarcoma which was found in 53 (43%) of the patients, of which 41 (33%) had lesions in the lower extremities while an additional 12 (10%) had upper limb involvement as seen in Figure 2. The head and neck region in Figure 2 was the second most common site of involvement and was noted in 25 (20%) patients, then followed by the abdominal region in 17 (14%) and thoracic region in 12 (10%) patients.

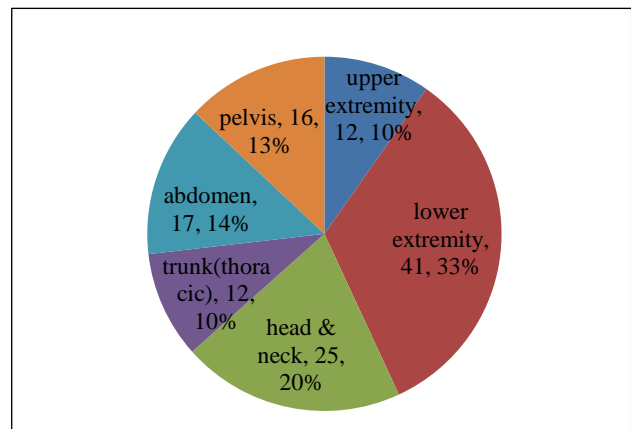


Figure 2: Tumour site of involvement of soft tissue sarcoma.

Eastern cooperative oncology group (ECOG) performance status as presented in Table 6 used to assess the patients showed about half of the patients had a performance score of 1. ECOG performance status of 2 and 3 were seen 31 (25.2%) and 20 (16.3%) of the patients respectively. Five patients (4.1%) had a performance status of 4 at initial presentation.

DISCUSSION

Soft tissue sarcoma epidemiology differs slightly among several regions around the world. There are studies that showed a 2.4% and above prevalence of soft tissue

sarcoma in Nigeria which is higher than global incidences.^{1,13,22} There is a predominance of the male gender in patients with soft tissue sarcoma in most studies and also conforms with this study where males were 65.9%, while females were 34.1%. The median age of presentation of soft tissue sarcoma is about 60 years, which is slightly higher than previous studies conducted.^{12,14} The mean age in a study in Japan was 51 years while that from a study in Kano, Nigeria was 39 years.^{10,12} The majority of the patients were between the age groups 21-30 years (21.1%) and 51-60 years (18.7%), with 1 patient being over 70 years.^{10,14} The mean age of the patients was 36.1±18.0 years. In this study, it was similar to that seen in study in AKTH, Kano, Nigeria.^{12,14}

Table 6: ECOG performance status of the patients at initial presentation.

Performance status (ECOG)	Frequency (%)
0	8(6.5)
1	59(48.0)
2	31(25.2)
3	20(16.3)
4	5(4.1)
Total	123(100.0)

Nigeria being a low income country has most of its inhabitant in the northern part of Nigeria being traders, housewives and farmer as seen in this study. Others include professionals and semiskilled professionals like carpenters, plumbers, tailors, and farmers. The level of literacy is still very low in the northern part of Nigeria as most patients have no formal education.

The risk factors identified were chemical carcinogens in 2.4% of the patients, while 12.2% had a first or second degree family history of cancer. Majority of the patients had no identifiable risk factors. This correlates with a study in France and other studies where most patients prospectively studied did not have any identifiable risk factors.^{6,13}

Rhabdomyosarcoma was the commonest soft tissue sarcoma seen in the total patients reviewed with 39.0% seen which was similar with a study in the south western part of Nigeria where it made up 13.1% of all soft tissue sarcoma but this differs from a study in Kano where Kaposi sarcoma was the predominant soft tissue sarcoma seen but only 4.9% of the patients seen in this study had Kaposi sarcoma.¹² Rhabdomyosarcoma was also the commonest histological type seen in both the paediatric and adult age group.

This is in keeping with other studies conducted in Nigeria and other countries.²³⁻²⁵ Malignant peripheral nerve sheath tumour comprised of 2% of all soft tissue sarcoma but this study showed a much higher value of 15.4%. About 1-6% of all soft tissue sarcoma are

dermatofibrosarcoma protuberance, this is at variance with our study where a much higher frequency of 12.2% was seen with this histological type.^{26,27} Fibrosarcoma and leiomyosarcoma were 7.3% and 4.1% respectively in this study but were among the commonest in the study at Sagamu.²⁸ Also malignant fibrous histiocytoma was 3.3%, however other reports showed malignant fibrous histiocytoma as the commonest soft tissue sarcoma.²⁹

Most patients presented with a swelling which was seen in 62% of patients, while 40% had pain, 9% ulcerative lesion and 4% presented as an incidental finding. This is in keeping with findings in most studies.^{19,30}

The extremities were the commonest primary site of soft tissue sarcoma, 41 (33%), had lesions in the lower extremities while an additional 10% had upper limb involvement. This is also seen in most studies globally and in Nigeria.^{17,22,31} The head and neck region were the second most common site and was noted in 20% of the patients, then followed by the abdominal region and thoracic region which constituted 14% and thoracic region 10% respectively.

Patients usually present late in Nigeria and thus their advanced disease causes a decline in their performance status.³² This is seen in the performance status (ECOG) used to assess the patients showed about half of the patients had a performance score of 1. A WHO performance status of 2 and 3 were seen in 25.2% and 6.3% of the patients respectively. About 4% had a performance status of 4.

Limitation of the study:

- It was a retrospective study.
- The grade of histological diagnosis of patients with soft tissue sarcoma in most histology reports was not documented.
- It was not a population-based study.

CONCLUSION

Soft tissue sarcomas are rare tumours that consist of various histological types. Its epidemiological characteristics differs globally in several literatures. Rhabdomyosarcoma is seen to be the predominant histological type in both paediatric and adult age groups, but this is not the case in other reports in most other studies. The extremities remain the commonest site of involvement as seen in both this study and most reports. Most patients still present late in our environment and thus have a poor performance status at presentation, which has been noted to affect their survival.

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