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Original Research Article

Study of soft tissue sarcomas over a period of 3 years

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ABSTRACT

Background: Soft tissue malignancies constituted a heterogeneous group of rare solid tumors of mesenchymal cell origin with distinct clinical and pathological features. The aim of the study was to know the prevalence of soft tissue sarcoma, sex, age and site distribution, histopathology and various treatment options adopted with follow up.
Methods: A total of 26 cases of soft tissue sarcoma were analyzed for a three-year period. Clinical presentation, age and sex distribution, histological type and treatment modalities adopted were recorded and analyzed.
Results: Out of 26 cases 44% of cases were between 30-50 years and 44% of tumors were situated in lower extremity. The commonest histological type was liposarcomas and fibrosarcoma. Lymph node metastasis was seen in 4% of cases. Distant metastasis was present in 3 cases, 2 with lung metastasis and l with lung and liver metastasis. Surgery was the main modality of the treatment. 12% of the cases presented with recurrent tumor, the duration between surgery and recurrence was 6 months. Only 38% turned for follow up, 2 patients succumbed to death because of multiple pulmonary secondaries and chest infections.
Conclusion: In the present study, all the cases of soft tissue sarcoma presented in late stage of the disease due to illiteracy and lack of health education. Recurrence was seen in 12% of cases. The overall survival rates and quality of life of the patients can be improved by frequent health camps at primary health centers for early detection of the disease, providing adequate health education, diagnostic and management facilities.

Keywords: Histopathology variants, Management, Prevalence, Soft tissue sarcoma

INTRODUCTION

Soft tissue sarcomas (STS) are a rare group of tumors, arising from mesenchymal tissue with heterogeneous differentiation. Soft tissue sarcoma can arise from muscles, fat, fibrous tissue, blood vessels and other supporting tissues of the body. About 1% among adults and 15% among children are affected by soft tissue sarcoma.[1] Little is known about the etiology of STS. Specific risk factors, like radiation, herbicides containing chlorophenol and phenoxy acids have been identified in the causation of soft tissue sarcoma. Increasing incidence rates of STS have been reported by several authors.[2,3]

This trend of increasing rates is probably due to recent shifts in the diagnostic criteria, gender predilection is varying in different nations and studies.[4-6] For stage II and III sarcomas, surgically removing the tumor is still the main treatment modality. Lymph nodes will be removed if they have metastatic carcinoma. If the tumor is large or in a place that would make surgery difficult, the patient may be treated with chemotherapy, radiation, or both before surgery. For large tumors in the arms or legs, giving chemo by isolated limb perfusion is also an option.

The goal of treatment is to shrink the tumor, making it easier to remove. These treatments also lower the chance of recurrence of the tumor. Smaller tumors may be treated with surgery first, then radiation to lower the risk

of recurrence. Sometimes chemotherapy is given as well. The drug most often used is doxorubicin (Adriamycin). This drug may be combined with Ifosfamide (Ifex) and other drugs.

In rare cases, amputation is needed to remove the entire tumor. As with stage I sarcomas, radiation therapy with or without chemotherapy can be used when the tumor's location or size or the patient's health in general makes surgery impossible. There is evidence that chemo after surgery may benefit some people with stage II and III sarcomas.[5]

METHODS

A total of 26 cases of soft tissue sarcoma admitted in the surgery department and operated for soft tissue sarcoma were analyzed for a three-year period from January 2012 to January 2015. Clinical presentation, duration of symptoms, site and size of lesion, age and sex distribution, histological type and treatment modalities adopted were recorded and analyzed.

RESULTS

Out of 26 cases 44% of cases were between 30-50 years, youngest patients are at the age of 8years and oldest is 75 years (Table 1).

Table 1: Age distribution in soft tissue sarcomas-26 cases.

Age in years	No. of cases	Percentage %
0-10	1	1.92
11-20	2	9.62
21-30	4	17.31
31-40	7	25
41-50	5	19.23
51-60	3	13.46
61-70	3	9.62
71-80	1	3.85

Table 2: Clinical features in soft tissue sarcomas-26 cases.

Symptoms	No. of cases	Percentage %
Painless mass	21	80%
Local pain/ ulceration	5	20%
Total	26	100%

Male:Female ratio is 1.17: l. 80% of the patients presented with painless mass and 20% presented with local pain or ulceration (Table 2). 46% of patients attended the clinics after one year of onset of the swelling and 54% of patients after 2 years of the onset of the swelling (Table 3).

Table 3: Duration of symptoms.

Duration	No. of cases	Percentage %
less than 6 months	9	35
6 months-1 year	3	11
1-2 year	12	46
1-3 years	2	8
Total	26	100%

Table 4: Anatomical Distribution of soft tissue sarcomas-26 cases.

Site of tumor	No. of cases	Percentage %
Lower extremity	11	44
Upper extremity	7	27
Thorax	2	6
Abdomen	4	17
Back	1	3
Head and neck	1	3
Total	26	100%

44% of tumors were situated in lower extremity, 27% in upper extremity, 6% in thorax, 17% in abdomen, 2% in back and 3% in head and neck (Table 4). Patients with tumor over extremities presented with edema of limb and weakness. Lymph nodal involvement was present in one case.

Table 5: Histopathological types of soft tissue sarcomas-26 cases.

Type	No. of the cases	Percentage %
Liposarcoma	6	23.07
Leomyosarcoma	3	11.5
Malignant fibro histiocytoma	5	19.25
Synovial sarcoma	3	11.5
Fibro sarcoma	6	23.07
Ewings sarcoma	1	3.84
Angiosarcoma	1	3.84
Rhabdomyosarcoma	1	3.84
Total cases	26	100%

Of the 26 cases of soft tissue sarcoma 23.07% were liposarcomas (Figure 1, 2), 11.5%wereleiomyosarcomas, 19.25% were malignant fibrous histiocytoma (Figures 3, 4), 11.5% synovial sarcoma (Figures 5-8), 23.07% fibro sarcoma (Figure 9), 3.84% each of Ewing's sarcoma, angiosarcoma and rhabdomyosarcoma (Figures 10, 11). (Table 5). Rhabdomyosarcoma occurred in child aged 8 years.

Lymph node metastasis was seen in 4% of cases. Distant metastasis was present in 3 -cases, 2 with lung metastasis and 1 with lung and liver metastasis. Surgery was the

main modality of the treatment. In present study in 80% of cases surgical treatment was given in the form of wide local excision followed by radiotherapy to the tumor bed. Amputation was performed in 2 cases (6%). Radiotherapy as a primary treatment was given for 4 cases (14%).

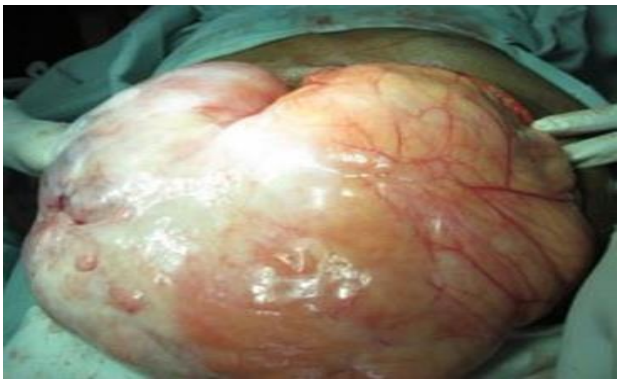

Figure 1: Intraoperative photograph of intraabdominal liposarcoma presenting as a large well circumscribed lobulated yellowish white mass.

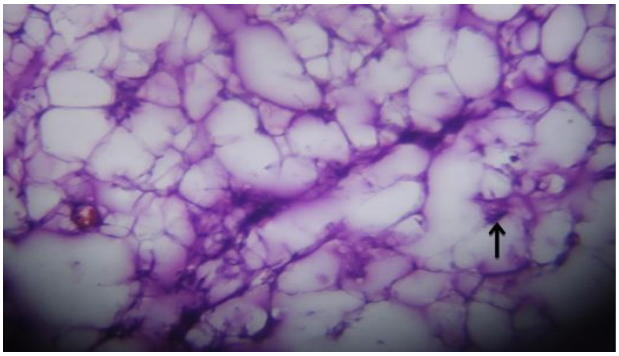

Figure 2: Photomicrograph of Liposarcoma showing variable sized adipocytes with lipoblast (arrow) (H and E,400X).

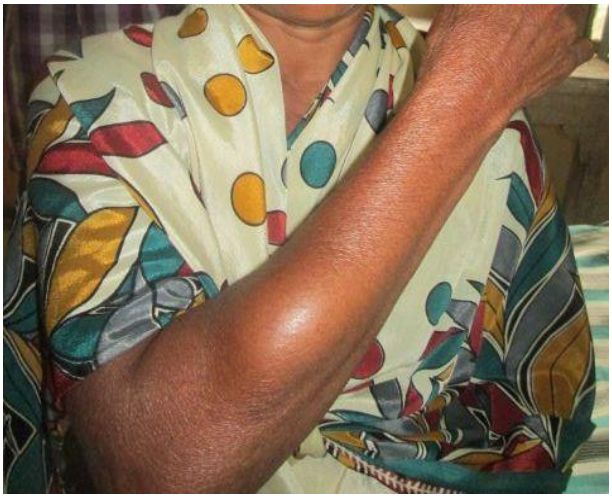

Figure 3: Clinical photograph of malignant fibrous histiocytoma presenting as a subcutaneous swelling on the forearm.

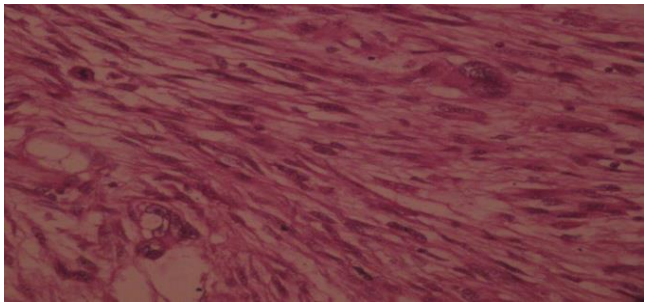

Figure 4: Photomicrograph of malignant fibrous histiocytoma: cells showing marked nuclear pleiomorphism, (H and E; 400X).

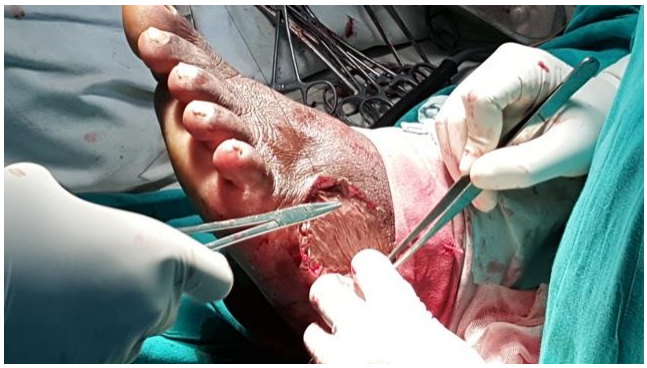

Figure 5: Intraoperative photograph of synovial sarcoma left foot.

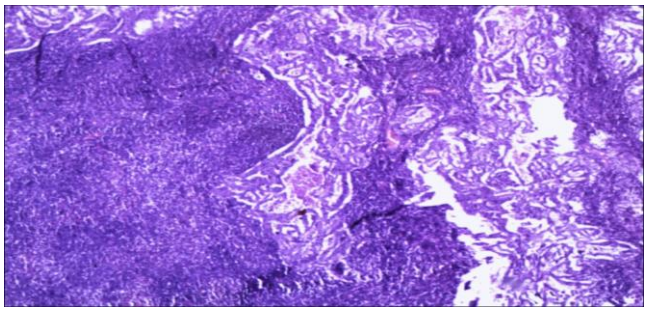

Figure 6: Photomicrograph of biphasic synovial sarcoma showing both stromal and epithelial component with glandular morphology. (H and E;40X).

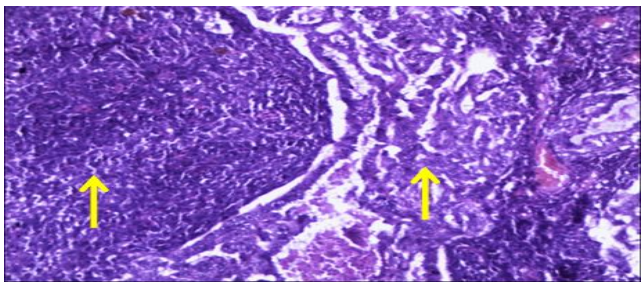

Figure 7: Photomicrograph of biphasic synovial sarcoma showing spindle shaped tumours cells (left arrow) and epithelial component with glandular morphology (right arrow) (H and E; 100X).

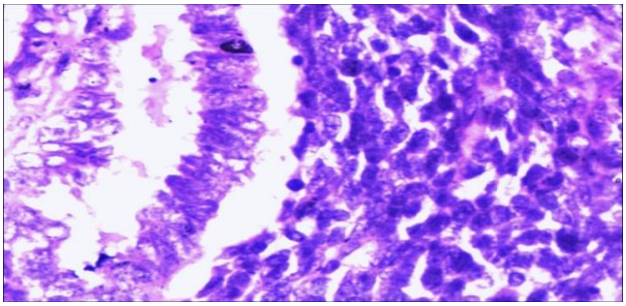

Figure 8: Photomicrograph of biphasic synovial sarcoma showing epithelial component as glands lined by columnar cells (H and E, 400X).

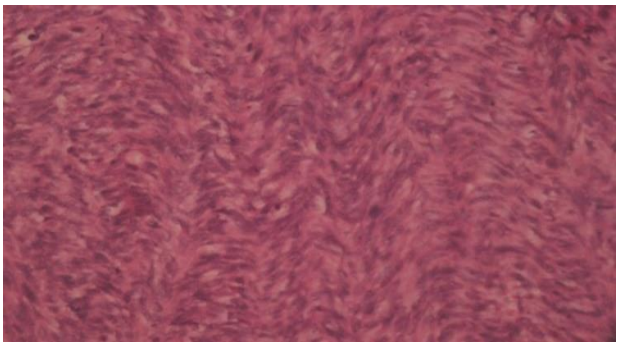

Figure 9: Photomicrograph of fibrosarcoma with spindle shaped cells showing herring bone pattern (H and E; 100X).

Most of these tumors were situated near trunk, upper thigh and axilla. In advanced cases and retroperitoneal sarcomas chemotherapy was adopted, besides debulking surgery and radiotherapy to the tumor bed. 12% of the cases presented with recurrent tumor diagnosed as malignant fibrous histiocytoma and fibrosarcoma on histopathology. The duration between surgery and recurrence was 6 months. Only 10 patients (38%) turned for follow up, 2 patients succumbed to death because of multiple pulmonary secondaries and chest infections.

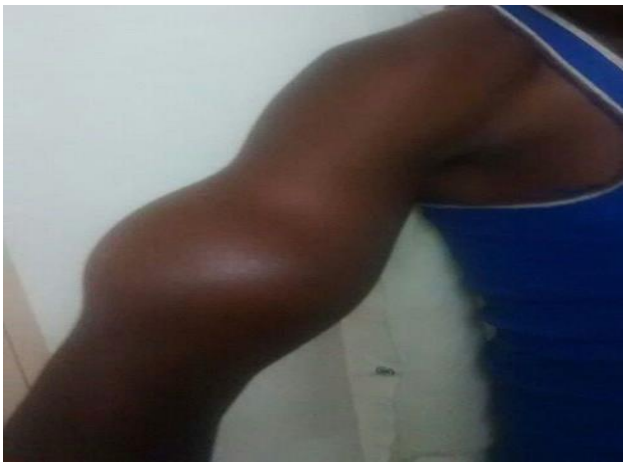

Figure 10: Clinical photograph of rhabdomyosarcoma right arm.

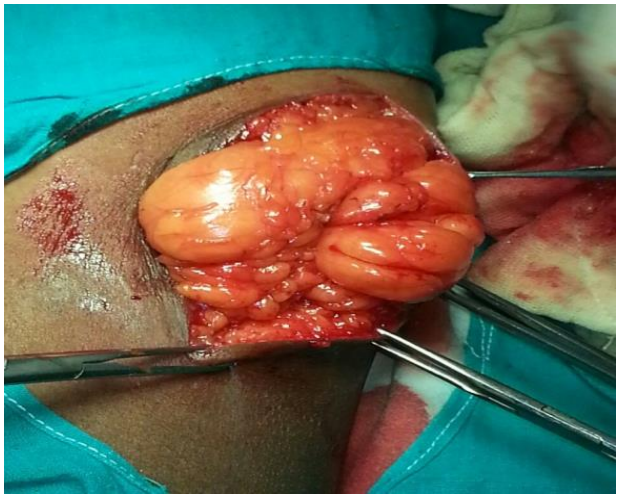

Figure 11: Intraoperative photograph of rhabdomyosarcoma right arm showing fleshy nodular tumour.

is important to consider malignancy in differential diagnoses. Because most soft tissue sarcomas present as a painless mass, clinicians must watch for signs suggestive of malignancy, including large size, rapid growth, and site deep into the deep fascia. The purpose of this study was to determine the relative prevalence, age at presentation, sex distribution, distribution of malignant soft-tissue tumors and to ascertain the relative frequency of these tumors in specific anatomic locations and age groups among a population of patients in a large pathologic consultation service.

Gibson TN et al[7] in their study in adults, the lower limb was the commonest location, followed by trunk and/or upper limb for malignant fibrous histiocytoma, fibrosarcoma and liposarcoma, head and neck for malignant peripheral nerve sheath tumor. In children, head and neck was the commonest site for rhabdomyosarcoma, head and neck and upper limb for malignant fibrous histiocytoma, retroperitoneum for neuroblastoma and trunk for fibrosarcoma. Mandong BM et al in their study the commonest sites affected were leg/foot 26.7%, head/neck 25.6% and thigh 19% with male: female ratio of 2: l.[8] Seleye-Fubara D et al observed that the tumors were more frequent under 20 years of age (22.7%) and least in 70 years and above (7.6%).[9] The lower limb was most affected (36.4%) while the least was the retroperitoneum (6.1%). The commonest predilection sites varied with different classes of malignancies. Adeniji KA et al observed male preponderance of the tumours with a male to female ratio of 1.3: 1.[10] Abudu EK et al reported with a male to female ratio of 1.9:1 and 72.3% cases occurring in the patients above 20 years.[11] Most cases of soft tissue malignancies were seen in the trunk as well in the lower limbs; constituting 63.0% but rhabdomyosarcoma was most common in the lower limbs (76.9%).

In the present study, out of 26 cases 44% of cases were

between 30-50 years, youngest patient was 8 years old. Male: Female ratio was 1.17:1.44% of tumors were situated in lower extremity, 27% in upper extremity, 6% in thorax, 17% in abdomen, 2% in back and 3% in head and neck. The prevalence of soft tissue sarcoma in the study by Reshadi H et al was malignant fibrous histiocytoma (23%), liposarcoma (22%), rhabdomyosarcoma (9%), leiomyosarcoma (8%), malignant schwannoma (5%), dermatofibrosarcoma protuberans (5%), synovial sarcoma (10%), fibrosarcoma (13%), extra skeletal chondrosarcoma (1%), and extra skeletal Ewing sarcoma (4%).[12]

The percentage of tumors in study by Kransdorf MJ et a1 was malignant fibrous histiocytoma (24%), liposarcoma (14%), leiomyosarcoma (8%), malignant schwannoma (6%), dermatofibrosarcoma protuberans (6%), synovial sarcoma (5%), fibrosarcoma (5%), and sarcoma, not classified further (12%).[13]

In Gibson TN et al study the commonest soft tissue sarcoma in adults were "sarcoma, not otherwise specified [NOS]" (20.1%) malignant fibrous histiocytoma (17.9%), fibrosarcoma (12.4%), liposarcoma (10.7%) and malignant peripheral nerve sheath tumour (10.2%).[7] In children, they were neuroblastoma (38.8%), rhabdomyosarcoma (23.9%), "sarcoma, NOS" (9%), fibrosarcoma (6%) and malignant fibrous histiocytoma (6%). Mandong BM et al analyzed 266 cases of soft tissue sarcomas, the histological types being; kaposi sarcoma (KS), rhabdomyosarcoma (RMS), malignant fibrous histiocytoma (MPH), and Fibro sarcoma (FS) accounting for 69 (25.9%), 63 (23.7%), 56 (21.1%) and 49 (18.4%) respectively.[8] Rhabdomyosarcoma occurring more in paediatric and adolescent age groups, while the rest of the subtypes were more prevalent in adults.

In the study by Seleye-Fubara D et al, Rhabdomyosarcoma was most frequent tumor (39.4%) while the least was leiomyosarcoma (1.5%).[9] Adeniji KA et al analysed soft tissue sarcomas over a period of 18 years (1979-1996).[10] The three commonest tumours were fibrosarcoma, rhabdomyosarcoma and liposarcoma in decreasing order of frequency. Fibrosarcoma and leiomyosarcoma were commonest in middle age whiler rhabdomyosarcoma was commonest in childhood and early adult life. There was a male preponderance of the tumours with a male to female ratio of 1.3: 1. Abudu EK et al soft tissue sarcomas constituted 1.84% of a total number of malignancies diagnosed with a male to female ratio of 1.9:1 and 72.3% cases occur in the patients above 20 years. Rhabdomyosarcoma was the most preponderant soft tissue malignancy, constituting 59.1% cases with the embryonal type predominating (53.8%).[11] Most cases of soft tissue malignancies were seen in the trunk as well in the lower limbs; constituting 63.0%. However, most cases of rhabdomyosarcoma in this study were seen in the lower limbs in 76.9%.

In the study by Wibmeret C et a1, the most common histotypes were sarcoma not otherwise specified (36%), leiomyosarcoma (24%), liposarcoma (12%), malignant fibrous histiocytoma (9%) and fibrosarcoma (5%). Age-adjusted incidence rate was 2.4 per 100 000 per year.[14]

In the present study, out of 26 cases of soft tissue sarcoma 23.07% were liposarcomas, 11.5% were leiomyosarcomas, 19.25% were malignant fibrous histiocytoma, 11.5% synovial sarcoma, 23.07% fibro sarcoma, 3.84% each of Ewing's sarcoma, angiosarcoma and rhabdomyosarcoma. Rhabdomyosarcoma occurred in child aged 8 years.

In general, the main principles of diagnosis and treatment may well apply to all soft tissue sarcomas, including the rarest presentations. Sarcoma treatments can be of various kinds. The common being wide excision of the tumor with associated tissues. The surgery may be either preceded or succeeded by radiotherapy. Another treatment method is chemotherapy, which relies on the action of various drugs on the malignant tissues.

In the present study surgery was the main modality of the treatment followed by radiotherapy to the tumor bed. Amputation was performed in 2 cases (6%). Radiotherapy as a primary treatment was given for 4 cases. Recurrence was seen in 12% of the cases who underwent only wide local excision. The duration between surgery and recurrence was 6 months to one year in cases of fibrosarcoma, malignant fibrous histiocytoma and liposarcoma. Only 10 patients (38%) turned for follow up, 2 patients succumbed to death because of multiple pulmonary secondary's and chest infections.

CONCLUSION

In the present study, soft tissue sarcomas accounted for 1.07% of the total cancers during that period with male preponderance and commonly occurring between 30-50 years of age. Seventy percent of tumors occurred in extremities with liposarcoma and fibrosarcoma being the commonest histological type.

In the present study surgery was the main modality of the treatment followed by radiotherapy to the tumor bed. Radiotherapy as a primary treatment was given for 4 cases. Amputation was performed in 2 cases (6%).

There was increased recurrence and incomplete excision of the tumor in the present study as the patients presented in late stages of the disease. Due to illiteracy and lack of health education the radiotherapy sessions taken were highly irregular with irregular follow ups. Early reporting of patients is very important which needs combined effort by health personnel, pathologist, surgeon, radiologist, relatives and social workers.

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Conflict of interest: None declared
Ethical approval: The study was approved by the Institutional Ethics Committee

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