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Original Article

Clinical Evaluation of Soft Tissue Sarcoma: An Observational Study on 26 Cases in a Tertiary Care Teaching Hospital

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Abstract

Introduction: "Sarkoma" (Greek) means fleshy growth. Tumors that arise from common embryonic ancestry, the primitive mesoderm, are known as sarcomas. The aim of this observational study is to evaluate the demographic distribution, histopathological presentation, clinical presentation, and various modes of management of soft tissue sarcomas (STS).

Materials and Methods: The present study is a descriptive hospital-based study conducted in the Department of General Surgery in a tertiary care hospital from November 2013 to November 2015. Patients of all ages and both genders presenting to the outpatient department of general surgery with malignant soft tissue tumors were included in the study. After clinically diagnosing as a case of STS, each patient was subjected to various necessary clinical and biochemical investigations pertaining to confirmation of the diagnosis and the patient was treated according to the standard management protocol following in this institution.

Observations and Results: Clinical parameters of evaluation such as incidence of STS, gender distribution, time of presentation of tumors, anatomical and histopathological distribution, presence of lymphadenopathy, occurrence of distant metastasis, mode of treatment, recurrence rate, and follow-up were evaluated in each patient and conclusions were drawn. Data were expressed in absolute numbers and percentages and tabulated statistically.

Conclusion: STS is a rare malignancy with poor prognosis. With the advent of immunohistochemistry and combined therapies, survival rate and quality of life can be improved significantly.

Key words: Malignancy, Metastasis, Prognosis, Sarcoma, Soft tissue tumor

INTRODUCTION

Soft tissue can be defined as non-epithelial extraskeletal tissue of the body exclusive of the reticuloendothelial system, glia, and supporting tissue of various parenchymal organs. It is represented by the voluntary muscles, fat, and fibrous tissue along with the vessels serving the tissues. By convention, it also includes peripheral nervous system because tumors arising from nerves also present as soft tissue masses.

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"Sarkoma" (Greek) means fleshy growth. Tumors that arise from common embryonic ancestry, the primitive mesoderm, are known as sarcomas.[1] The term soft tissue sarcomas (STSs) refer to a large variety of malignant tumors arising in the soft tissues that are grouped together due to their similarities in pathogenic appearance, clinical presentation, and behavior.[2]

Malignant tumors arising from connective tissue are termed as sarcomas and from epithelial tissue are called carcinomas. Soft tissue tumors are both benign and malignant. Benign soft tissue tumors outnumber malignant tumors by a margin of about 100:1 in hospital population.[3] STSs, compared with other neoplasms, are relatively rare and constitute <1.5% of all cancers with an annual incidence of 6 per 100,000 persons.[4]

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The endothelium lining of vascular and lymphatic channels and the mesothelium lining of body cavities and visceral organs are two types of epithelium that arises from the mesoderm. Malignant tumors that arise from these structures behave like tumors that develop from connective tissue cells.[5] For this reason, tumors arising from endothelium and mesothelium are included in the category of sarcoma.

Due to similarities in anatomical sites of origin, clinical presentation, and clinical behavior, tumors arising in Schwann cells which arise from neural tube of primitive ectoderm are also included in the category of STSs.[6] Sarcomas account for <1% of all new malignancies detected every year.[7,8]

The aim of this observational study is to study the demographic distribution, histopathological presentation, clinical presentation, and various modes of the management of STS.

MATERIALS AND METHODS

The present study is a descriptive hospital-based study conducted in the Department of General Surgery in a tertiary care government general hospital from November 2013 to November 2015.

Inclusion Criteria

Patients of all ages and both genders presenting to the outpatient department of general surgery in our tertiary care hospital during the period from November 2013 to November 2015 with malignant soft tissue tumors were included in the study.

Exclusion Criteria

Patients who were clinically diagnosed as STS but on investigation found to have other non-malignant conditions were excluded from the study. After obtaining approval from the Institutional Ethics Committee, a written/informed consent was taken from the patients in his/her vernacular language. A thorough clinical history was taken regarding chief complaint, duration, and other relevant history. Each patient was clinically examined to arrive at a clinical diagnosis, and every detail was documented. After clinically diagnosing as a case of STS, each patient was subjected to various necessary clinical and biochemical investigations pertaining to confirmation of the diagnosis, and the patient was treated according to the standard management protocol following in this institution. At the end of the study, the observations and results were tabulated and analyzed statistically. Data were expressed as absolute numbers and percentages.

OBSERVATIONS AND RESULTS

Frequency of Disease

A total number of patients with malignant tumors admitted in the general surgical ward during the study period was 2441. A number of diagnosed STS cases were only 26. Percentage of STSs was found to be 1.07%. None of the patients had relevant family history and history regarding exposure to radiation or carcinogens or association of family members with malignant tumors. Regarding age distribution, 44% were between 30 and 50 years, the age of the youngest patient was 8 years, and the age of the oldest patient was 75 years which is represented in Table 1 and Figure 1.

Sex

The ratio of male to female was about 1.17:1, as represented in Table 2.

Symptoms

A number of patients presented with painless mass were 21 which were 77% of the total patients, of which 5 patients (20%) were presented with local pain, ulceration, fungation, and swelling of distal part of the limb as represented in Figure 2. Recurrent tumor after excision was noted in six patients which are 23.1% of the total patients.

Table 1: Age distribution

Age (years)	Number of cases (%)
0–10	1 (1.92)
11–20	2 (9.62)
21–30	4 (17.31)
31–40	7 (25.00)
41–50	5 (19.23)
51–60	3 (13.46)
61–70	3 (9.62)
71–80	1 (3.85)

Table 2: Sex incidence

Sex	Number of cases (%)
Male	14 (54)
Female	12 (46)
Total	26 (100)

Table 3: Time of presentation after onset of symptoms

Duration	Number of cases (%)
<6 months	9 (35)
6 months–1 year	3 (11)
1–2 years	12 (46)
2–3 years	2 (8)
Total	26 (100)

Time of Presentation after Onset of Symptoms

About 46% of the patients came for the treatment after 1 year of onset of swelling, whereas 54% of the patients came to the hospital after 2 years of onset of complaints, as represented in Table 3.

Anatomical Distribution

About 44% of the tumors situated in lower extremity, 27% in upper extremity, 6% in thorax, 17% in abdomen, 3% in back, and 3% in head and neck as represented in Table 4.

Pressure Effects

Patients with tumors over extremities presented with edema of limb and weakness.

Regional Lymphadenopathy

Lymph nodal involvement was present in one case accounting to 4%.

Histopathology

Of the 26 cases of STS, 4 cases (15%) were liposarcomas, 3 (12%) were leiomyosarcomas, 3 (12%) were malignant fibrous histiocytomas, 3 (12%) were synovial sarcomas, 3 (12%) were fibrosarcomas, 2 (7%) were dermatofibrosarcoma protuberans, 2 (7%) were pleomorphic sarcoma, 2 (7%) were myxoid liposarcomas, and 1 (4%) each of Ewing's sarcoma, hemangiopericytoma, and rhabdomyosarcoma as represented in Table 5.

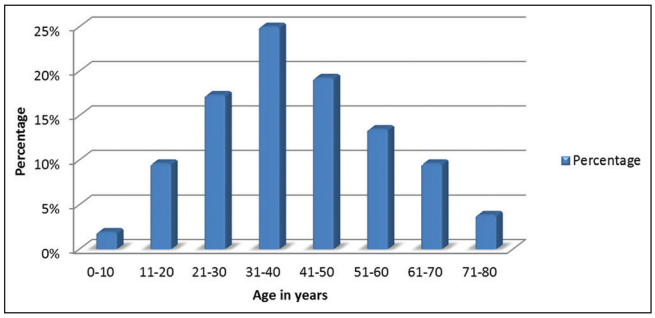

Figure 1: Age distribution

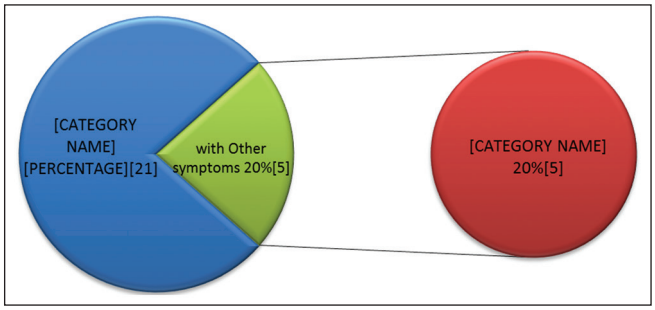

Figure 2: Symptoms

Distant Metastases

Distant metastases were present in 3 cases (10%): 2 with lung metastasis and 1 with lung and liver metastasis.

Treatment

Surgery was the main modality for the treatment of these tumors in the present study; in 20 cases (80%), 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 morality of treatment was given in around 4 of the cases (14%). Most of these tumors are situated near trunk such as upper thigh and axilla. In advanced cases and retroperitoneal sarcomas, chemotherapy was adopted besides debulking surgery and radiotherapy to the tumor.

Follow-up

Only 10 patients (38%) turned up for follow-up. Those patients who underwent only wide excision, local recurrence was noted in 3 cases. The duration between operation and appearance of local recurrence is about 6 months–1 year. Reexcision of the tumor was done in these patients, and post-operative radiotherapy was given [Table 6 and Figure 3].

DISCUSSION

STSs are rare unusual neoplasms that account for about 1% of human adult malignancies and 15% of pediatric

Table 4: Anatomical distribution of STS

Site of tumor	Number of cases (%)
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)

STS: Soft tissue sarcomas

Table 5: Histopathological distribution of the cases

Type	Number of cases (%)
Liposarcoma	4 (15)
Leiomyosarcoma	3 (12)
Malignant fibrous histiocytoma	3 (12)
Synovial sarcoma	3 (12)
Fibrosarcoma	3 (12)
Dermatofibrosarcoma protuberance	2 (7)
Pleomorphic sarcoma	2 (7)
Myxoid liposarcoma	2 (7)
Ewing's sarcoma	1 (4)
Hemangiopericytoma	1 (4)
Rhabdomyosarcoma	1 (4)
Stromal sarcoma	1 (4)
Total cases	26 (100)

malignancies.[9] In the present study, 26 patients with STSs were treated in a period of about 2½ years from November 2013 to November 2015 in a tertiary care hospital. They account to 1.07% of the total number of admissions diagnosed with various soft tissue malignancies during the study period.

According to the study conducted by Cheung *et al.*; on the socio-economic factors that affect the outcome of STS: An analysis of a surveillance, epidemiology and end results (SEER) data, 55% were female patients and 45% were male patients.[10] There is a slight male preponderance in the present study with males being 56% and females 46% when compared to the above study as represented in Table 4.

The mean age of presentation was 53.5 years according to SEER data in the above study, and it was about 40 years in the study conducted. 44% of the tumors occurred between 30 and 50 years of age group in the present study. The duration of symptoms reported by patients in the present study varied widely with a median of 26 weeks for STSs.

About 80% of the patients presented with painless mass as the presenting complaint; among them, 48% of patients presented after 1 year of the onset of symptoms and 52% of patients presented after 2 years of the onset of symptoms.

According to the study of Cheung *et al.*, the majority of primary lesions originate in the lower extremity which accounts to 15% when compared to other sites, and the comparison between anatomical distribution of sites between the present study and above-mentioned study is represented in Table 7. Majority of tumors originated in the lower extremities in both the studies.

More than 50 histological subtypes of STS are recognized, many of which are associated with distinctive clinical, therapeutic, or prognostic features. A study on histological distribution was conducted by Brennan *et al.*, in 10,000 patients with STS treated at Memorial Sloan Kettering Cancer Centre from July 1, 1982, to May 2013.[11]

According to the above study, the most common histologic types of STS in adults (excluding Kaposi sarcoma) are malignant fibrous histiocytoma (24%), leiomyosarcoma (21%), liposarcoma (19%), synovial sarcoma (12%), and malignant peripheral nerve sheath tumors (6%). Histological type was found to be the one of the most important predictors of sarcoma-specific death, with malignant peripheral nerve sheath tumors having the highest risk for mortality.[12,13] The histopathologic distribution of sarcomas in the present study correlated well with the study of Brennan *et al.*, which is represented in Table 8.

Patients with extremity sarcoma usually present with painless mass, and by that time, they report that the tumor may grow up to 10–15 cm in diameter.[14] Retroperitoneal sarcomas typically do not produce symptoms until they grow large enough to compress or invade contiguous structures.[15]

Most of the times, fine-needle aspiration cytology is inconclusive. Ideally, the initial diagnostic procedure is performed at the centre where the patient is operated. Tru-cut biopsy is the diagnostic procedure of choice. In case of inadequate tissue, an open, linearly placed, incisional biopsy is indicated and this should come into future resectable margin.

During the past 25 years, multimodality treatment approach has been successfully applied to patients with

Table 6: Comparison between SEER data and observed data

Study	Analysis of SEER data (%)	Present study (%)
Male	45	56
Female	55	46

Data were expressed as percentage, SEER: Surveillance, Epidemiology, and End Results

Table 7: Comparison between the anatomical distributions of STS

Anatomical site	Chueng *et al*. study (%)	Present study (%)
Upper extremity	10	27
Lower extremity	15	44
Abdomen	10	17
Head and neck	10	3
Thorax	10	6
Pelvis	10	–
Autonomic nervous system	25	–
Back	10	3

STS: Soft tissue sarcoma

Table 8: Comparison of histological distribution

Histological type	Brennan *et al*. study* (%)	Present study (%)
Liposarcoma	20	15
Leiomyosarcoma	14	12
Undifferentiated pleomorphic sarcoma	14	7
Malignant fibrous histiocytoma	–	12
Synovial sarcoma	5	12
Myxofibrosarcoma	24	7
Fibrosarcoma	14	12
Malignant peripheral nerve sheath tumor	2	–
Others	28	23
Total	100	100

*Memorial Sloan Kettering Cancer center study

extremity sarcomas, and this has led to improvements in both survival and quality of life.[16] Wide local excision with 2 cm margin has not been demonstrated to adversely affect local recurrence or outcome compared to radical procedures such as amputation.

Patients with abdominal sarcomas continue to have high rates of recurrence and poor overall survival.[17]

Neoadjuvant radiotherapy, post-operative radiotherapy, and chemotherapy; the overall 5-year survival rate for patients with all stages of STS is 50–60%.[18,19] Of the patients who die of sarcoma, most will succumb to metastatic disease, in which 80% of the time occurs within 2–3 years of the initial diagnosis.

Clinical diagnosis was established primarily by accurate history and physical examination. Tissue diagnosis was established by Tru-cut biopsy and incisional biopsy, and treatment was planned accordingly.

Distant metastases were present in 3 cases (10%): 2 cases with lung metastasis and 1 with lung and liver metastasis.

Surgery was the main modality of the treatment of these tumors in the present study; in 20 cases (80%), 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 morality of treatment was given in around 4 of the cases (14%), and the modes of treatment in the present study are represented in Table 9 and Figure 4. Most of these tumors are situated near trunk such as upper thigh and axilla. In advanced cases and retroperitoneal sarcomas, chemotherapy was adopted besides debulking surgery and radiotherapy to the tumor bed.

About 12% of the cases (3 cases) presented with recurrent tumor and diagnosed with pleomorphic sarcoma, fibrosarcoma, and dermatofibrosarcoma protuberans. Recurrence rate cannot be estimated accurately as only few percentages of patients came for follow-up and failure on the part of the patients to undergo radiotherapy postoperatively. In the present study, the recurrence rate was reported to be 12% (3 cases) as represented in Figure 5.

The overall relative 5-year survival rate of people with STS is around 50% according to statistics from the National Cancer Institute.[20,21] The survival rates also cannot be calculated accurately as the follow-up study is short and failure on the part of the patients to undergo radiotherapy postoperatively. A total of 2 patients (7%) succumbed to death due to multiple pulmonary secondaries, chest infections, and malignant cachexia. The reason for the increased recurrence and incomplete excision of the tumor in our study is that most of the patients in the study group are illiterates and come from village background. Due to lack of awareness and ignorance, these patients presented lately after the onset of symptoms, as the mass is painless, with large tumors, which are not resectable

Table 9: Mode of treatment

Mode of treatment	Number of cases (%)
Wide local excision followed by RT	20 (76.9)
Amputation	2 (7.6)
Radiotherapy	4 (15.3)

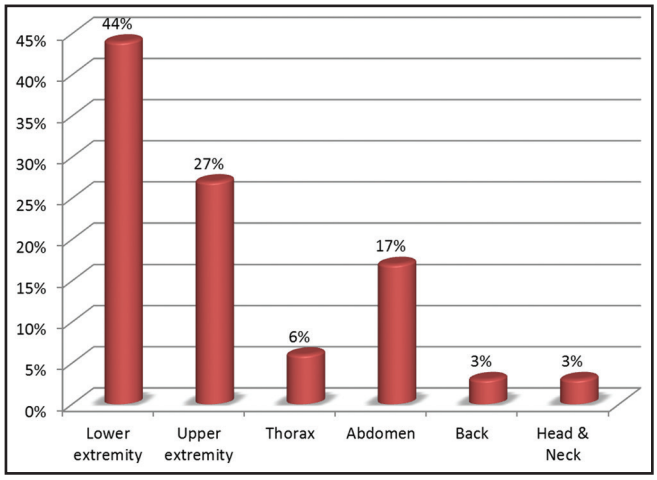

Figure 3: Anatomical distribution of soft tissue sarcoma

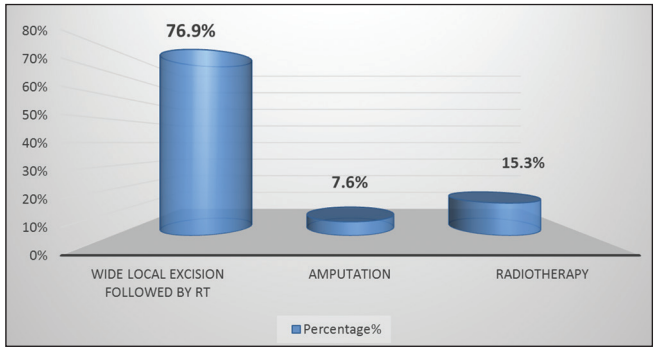

Figure 4: Mode of treatment

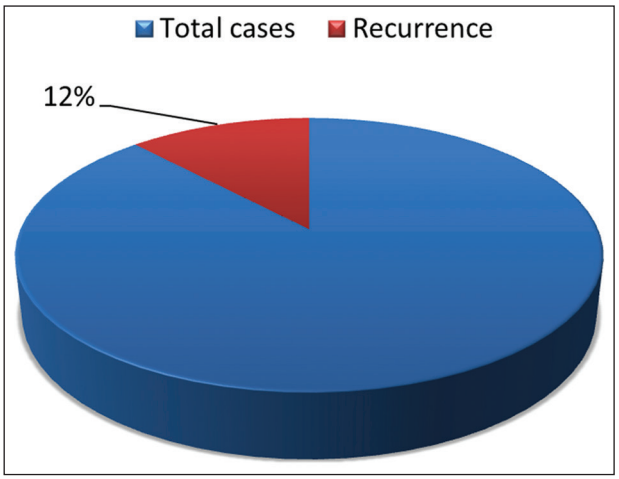

Figure 5: Recurrence rate

completely and many of them are not coming for radiotherapy and missing regular follow-ups. In addition, lack of health education regarding the disease.

CONCLUSION

STS is a rare malignancy with poor prognosis. With the advent of immunohistochemistry and combined therapies, survival rate and quality of life can be improved by

- Frequent medical camps at primary health centers for the early detection of the disease.
- Providing adequate health education to the people so that patient referred to hospital at an early stage, where adequate diagnostic facilities, chemotherapy, and radiotherapy are available.
- Prognosis is likely to be influenced by early reporting by the patients and gross characteristics of tumor itself.
- Thus, management of STS is a teamwork which includes, health personnel, pathologist, surgeon, radiologist, radiotherapist, relatives, and social workers. A coordinated team effort is all which gives much improved results.

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