

## Original Research Article

# Clinicopathological study of soft tissue sarcoma-retrospective study of tertiary cancer institute in eastern India

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## ABSTRACT

**Background:** Soft tissue sarcomas are a rare and heterogeneous group of malignant tumors of mesenchymal origin that comprise less than 1 percent of all adult malignancies. Although they occur anywhere in the body, they involve most commonly in extremities, trunk, retroperitoneum and head and neck. The aim of the study was to analyze clinical and histopathological features of various soft tissue sarcomas.

**Methods:** This was a retrospective study, conducted in tertiary cancer centre in Odisha during the period 2015 to 2018. We collected clinical parameters like age, sex, site of swelling, any associated pain and biopsy reports and these variables were correlated with final histopathology reports.

**Results:** A total of 107 patients were included in the study, with male to female ratio of 2:1(71 and 36) and average age of 43.45 years. All of them presented with a swelling. The lower extremities were the most common sites i.e. 44.62%. Pleomorphic sarcoma was the most frequent histologic variety comprising 43% and less frequent variety were angiosarcoma, and myxoid sarcoma.

**Conclusions:** Soft tissue sarcoma are predominant in males and middle aged population are frequently affected. Most common affected site is lower extremity and pleomorphic sarcoma is the prominent histologic type.

**Keywords:** Soft tissue sarcoma

## INTRODUCTION

Soft tissue sarcomas are a rare and heterogeneous group of malignant tumors of mesenchymal origin that comprise less than 1 percent of all adult malignancies.<sup>1</sup> Although they occur anywhere in the body, they involve most commonly in extremities, trunk, retroperitoneum and head and neck.<sup>2</sup> The patient usually presents with painless lump and seek consultation when it attains large size. Few patient present with swelling with fungation or ulceration or with neuro vascular symptoms like numbness, paresthesia, weakness etc. Predisposing factors of soft tissue sarcomas comprise environmental exposure to

carcinogenic agents like vinyl chloride, herbicides and pesticides etc. Other important predisposing factors are chronic lymphoedema and previous radiation exposure. Radiation induced sarcomas have poorer prognosis compared to sporadic soft tissue sarcomas.<sup>3</sup> Genetic syndromes commonly associated with sarcomas are neurofibromatosis, bilateral retinoblastoma and Li-Fraumeni syndrome.<sup>4</sup> Familial sarcomas present in early decades of life. HIV associated malignancy have increased risk for developing soft tissue sarcoma like Kaposi sarcoma.<sup>5</sup> However, most cases of soft tissue sarcoma do not have a known risk factor. Diagnosis of soft tissue tumors are done by MRI or CT scan and core-cut biopsy

of the swelling. MRI scan is the preferred investigation for extremity sarcomas as it gives better soft tissue resolution. Core-cut biopsy provides histologic type and grade of the tumor in most of the time. It helps to triage treatment option i.e. upfront surgery or preoperative chemo or radiotherapy since some sarcomas are chemo sensitive. Soft tissue sarcomas should be managed in high volume centers with multidisciplinary team approach because many times unplanned surgery result in early recurrence and distant metastasis. Surgery comprise limb sparing, function preserving, resection with adequate margin whenever possible. Adjuvant radiotherapy can be given either by intraoperative brachytherapy catheter placement or external beam radiotherapy based on final histopathology report. Adjuvant radiotherapy reduces the chance of local recurrence.<sup>6</sup> Various histologic types are diagnosed by either conventional methods like Light microscopy, special stains, or immunohistochemistry or chromosomal studies.

Local recurrence is main concern after the treatment and it mainly depends on margin status of the resected specimen and grade of the tumor. Prognosis of soft tissue tumors mainly depend on tumor size, tumor grade, location, margins, clinical staging, DNA ploidy and chromosomal alterations.<sup>7</sup> The adverse prognostic factors for soft tissue sarcomas' are: size, increasing age, high grade, metastasis at diagnosis, local recurrence at diagnosis (following unplanned excision), positive surgical margin, deep to muscular fascia, high levels of tumor necrosis.<sup>7-9</sup>

Although soft tissue tumors are frequently encountered in clinical practice, they are seldom reported, especially in India and therefore there is less published literature from our sub-continent. This retrospective study aimed to analyze clinical and histopathological features of various soft tissue sarcomas in eastern part of India.

## METHODS

This is retrospective observational study, conducted in a tertiary cancer centre in Odisha, India during the period May 2015 to March 2018. The Institutional Ethics Committee approval was obtained and all patients gave an informed consent to be included in this study. We collected clinical parameters like age, sex, site of swelling, any associated pain and final histopathology reports of patients presented with soft tissue sarcoma and those underwent surgery. The data collected from medical record library and final histopathology report collected from oncopathology department. All patients who presented with soft tissue sarcoma of extremity and trunk and got operated were included. Recurrent sarcoma, retroperitoneal sarcoma, bone sarcoma and benign soft tissue tumors were excluded from the study. Immunohistochemical studies and the electron microscopical studies were advised in some of the soft tissue malignant tumors to support the diagnosis. The data was analyzed using software SPSS 21.0 and compiled with help of tables and bar diagrams. Histological subtypes

were classified according to WHO classification of soft tissue tumors.

## RESULTS

A total of 107 patients were included in the study, of them 71 were males and 36 were females with male predominance (Table 1). The mean age at presentation was 43.45 years (Range: 7-78 years; Standard deviation (SD) 16.07. Majority of patients were between the age group 31-50 as shown in the Table 2 and the frequency of disease was 45.8%. Soft tissue sarcoma was infrequent below the age 10 and above the age 70, ie 0.9% and 2.8% respectively. All of them presented with a swelling. Majority of them were painless swelling (82.24%) and few patient had swelling with other symptoms like pain, ulceration, numbness and paraesthesia (17.75%), as shown in Table 3.

**Table 1: Gender distribution.**

Sex	Frequency (N)	Valid percent	Mean age $\pm$ SD (Minimum, maximum in years)
Male	71	66.4	43.45 $\pm$ 16.07 (7, 78)
Female	36	33.6	

**Table 2: Different age groups and incidence of soft tissue sarcomas.**

Age group in years	Frequency (N)	Valid percent
1-10	1	0.9
11-20	10	9.3
21-30	12	11.2
31-40	25	23.4
41-50	24	22.4
51-60	21	19.6
61-70	11	10.3
>70	3	2.8
Total	107	100

**Table 3: Various symptoms of soft tissue sarcomas.**

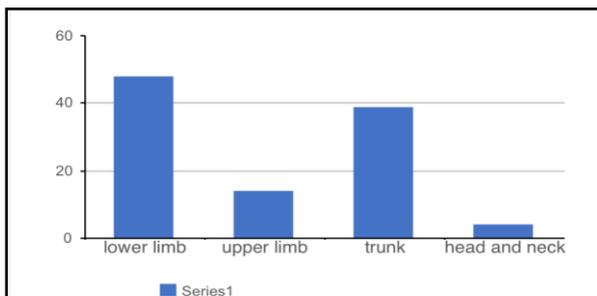
Symptoms	Frequency	Valid percent
Painless lump	88	82.24
Lump with other symptoms (pain, ulcer or neurovascular )	19	17.75
Total	107	100

The lower extremities were the most common sites involved by soft tissue sarcoma, i.e. 44.62% as shown in Figure 1. In lower extremity thigh was the common location of sarcoma (19.62%). Pleomorphic sarcoma was the most frequent histologic variety comprising 43% (Table 4). Other common histologic types were Liposarcoma (6.5%) and synovial sarcoma (6.7%) as depicted in Table 4. Final histopathology was spindle cell

sarcoma in approximately 18.7% cases that necessitated immunohistochemistry study for histologic diagnosis. Less frequent variety were angiosarcoma, and myxoid sarcoma.

**Table 4: Final histopathology types and grades and it's frequency.**

Final histopathology	Frequency (N)	Valid percent
Liposarcoma FNCLCC high grade	7	6.5
Spindle cell sarcoma low grade	20	18.7
Pleomorphic sarcoma FNCLCC High grade	34	31.8
Pleomorphic sarcoma FNCLCC Low grade	12	11.2
PNET	6	5.6
Fibro Sarcoma high grade	2	1.9
Fibro Sarcoma low grade	3	2.8
MPNST	1	0.9
DFSP Low grade	5	4.7
Synovial sarcoma low grade	1	0.9
Synovial sarcoma high grade	3	2.8
Pleomorphic rhabdomyo sarcoma	2	1.9
Myxoid Sarcoma	1	0.9
Malignant small round cell tumor	1	0.9
Angiosarcoma	1	0.9
Spindle cell sarcoma consistent with synovial sarcoma	3	2.8
Spinle cell tumour consistent with neurofibroma	1	0.9
Spindle cell sarcoma high grade	2	1.9
Malignant fibroushistiocytoma grade 2	1	0.9
Malignant peripheral nerve sheath tumour grade 3	1	0.9
<b>Total</b>	<b>107</b>	<b>100</b>



**Figure 1: Sites of soft tissue sarcomas.**

**DISCUSSION**

In this study most of the patients presented with painless swelling and few patients had swelling with pain or Neuro vascular symptoms. painless swelling is the most common mode of presentation of soft tissue sarcoma. This is in accordance with many other studies. In this study of 107 patients, males were predominantly diagnosed with sarcoma (66%) than females. Hui et al study showed male preponderance for the incidence, with a ratio of approximately 1.4:1, male-to-female ratio.<sup>10</sup> Similar study by Domajog et al also showed the male predominance of soft tissue sarcomas.<sup>11</sup> But other studies showed equal gender predilection or slight female predominance.<sup>12</sup> This may be due to heterogeneity of disease and difference in geographical distribution.

Similarly in the study of Geer et al, the study of soft tissue sarcomas of the extremities conducted.<sup>13</sup> The age of patients ranged from 16 to 88 years projecting wide range of age distribution and average age of presentation was 37 years. In our study, the age of patients ranged from 7 to 78 years with average age of presentation was 43.45 years.

Soft tissue sarcoma may occur anywhere in the body where mesenchymal tissue is present.

Lower extremities are the most common site of presentation followed by trunk, upper extremities, retroperitoneum and head and neck regions. In our study, approximately 44% of patients presented with sarcoma of lower extremities and thigh was the common site of sarcomas.

All patients underwent local imaging by MRI scan and core-cut biopsy was performed. Surgery was performed with intent of complete R0 resection. In some patients local flap reconstruction performed where primary closure was not possible. Final histopathology report collected from the oncopathology department. Histologic grading performed using three-grade system FNCLCC (French Fédération Nationale des Centres de Lutte Contre le Cancer).<sup>14</sup>

Malignant fibrous histiocytoma has been replaced by pleomorphic sarcoma because immunohistochemical and histopathologic analysis showed that many sarcomas previously classified as pleomorphic malignant fibrous histiocytoma could be reclassified as other histologic types. Undifferentiated pleomorphic sarcoma is now reserved for pleomorphic sarcomas that by current technology show no definable line of differentiation.<sup>15,16</sup>

In our study, we found high grade sarcomas were common than low grade varieties. It implies that most of the patients in our population with localised sarcoma will have poor prognosis and high chance of local recurrence and distant metastasis. So sarcomas should be treated in tertiary cancer centre with multidisciplinary team approach. Similarly, pleomorphic sarcoma was the common

histologic type comprising 43% of all sarcomas. Other similar study by Shukla et al showed synovial sarcoma and malignant fibrous histiocytoma were the predominant histologic types with a frequency of 28.9%.<sup>17</sup> Angiosarcoma and myxoid sarcoma comprised infrequent histologic types in this study.

## CONCLUSION

Soft tissue sarcoma are predominant in males and middle aged population are frequently affected. Most common affected site is lower extremity and pleomorphic sarcoma is the prominent histologic type.

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