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Adult Soft Tissue Sarcoma: A Prospective Observational Real-World Data

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Abstract

Introduction There is a lacuna of prospective studies on soft tissue sarcoma (STS) from the Indian subcontinent in published literature.

Objectives We conducted this study to describe the clinical profile and outcomes of STS in North India.

Materials and Methods This is a single-center, prospective, observational study conducted from October 2017 to September 2019. All consecutive patients aged ≥18 years with histopathological diagnosis of STS were enrolled. The study end points included overall response rate, progression-free survival (PFS), and overall survival (OS).

Results A total of 140 patients were included with a median duration of follow-up of 14 months (range: 1–25 months). The median age of patients was 45 years. The median duration of symptoms before diagnosis was 5 months (range: 1–18 months). The most common histopathologic subtype was undifferentiated pleomorphic STS (22%). Of 105 localized patients, 21 received neoadjuvant therapy with external beam radiotherapy and/or doxorubicin-based chemotherapy and reported partial response in 38% (8/21) of the patients; the remaining 62% (13/21) of the patients had stable disease. Neoadjuvant therapy resulted in nonsignificantly higher complete resection rates with relative risk of 2.37 (p = 0.19). Of the remaining 35 metastatic STS patients, 31 received chemotherapy and reported partial response in 39.1% (n = 9/23), stable disease in 30.4% (n = 7/23), and disease progression in 30.4% (n = 7/23) of the patients. For localized STS patients, 1-year disease-free survival (DFS) and OS rates were 87.6 ±3.5 and 95.3 ± 2.3%, respectively. The median OS for metastatic STS patients was 23.90 months (95% confidence interval [CI]: 7.43–40.36). Among metastatic STS, median OS was not reached for those who underwent curative resection versus 12.66 months (95% CI: 9.28–16.04) for those who received systemic therapy alone.

Conclusion Median age of 45 years is a decade earlier than seen in the Western population. Neoadjuvant therapy improved complete resection rates, though it was statistically nonsignificant. Curative resection among metastatic STS patients improves survival.

Keywords
- survival outcomes
- developing countries
- epidemiology
- India
- real-world data
- soft tissue sarcoma
- STS

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Introduction

Soft tissue sarcomas (STSs) constitute around 1% of all cancer types and include a heterogeneous group of more than 50 histopathological subtypes.[1,2] STS can arise from the mesenchymal tissue of any anatomical site. The clinical presentation of STS patients may have various symptom complexes depending on the location of origin, the aggressiveness of the disease, and the extent of spread. The management of resectable STS primarily involves surgery. The addition of perioperative radiotherapy and/or chemotherapy improves disease-free survival (DFS) and overall survival (OS).[3,4] The multimodality approach depends on patient characteristics, anatomical site, histopathological subtype, stage, and available resources.[5] Despite these advances, the overall 5-year survival probability is around 50%.[6]

Similar to other cancers, even among sarcomas, there appear to be significant ethnic and racial influences on clinical profile (such as age, stage at presentation, and tumor grade) and outcomes.[7,8] In addition, in developing countries, due to lack of health awareness, limited access to health and insufficient information among primary care providers may result in delayed diagnosis. This delay in diagnosis may result in advanced disease status and poor outcomes. There is a paucity of prospective studies in STS from the Indian subcontinent and low-to-middle-income countries, as most of the previous studies in the published literature are retrospective case series.[9-12] The present study was done prospectively to gain insight into the disease patterns and outcomes of STS in the Indian setup. Understanding STS patients' profile is essential in improving care for sarcoma patients and also for further research, to emphasize the significance of multidisciplinary management, and to raise awareness about the importance of STS collaborative network group in India.

Materials and Methods

This prospective, observational study was conducted from October 2017 to September 2019 at a tertiary care center in India. In this study, we had planned to include total 100 patients of STS, considering 5% level of significance (α), 7% margin of error (L), taking 1-year mortality of 13%,[7] and assuming 10% drop-outs/attrition/lost to follow-up. Patients aged 18 years and above, with histologically proven STS, were included in the study. Patients with recurrent disease, Ewing's sarcoma (EWS), gastrointestinal stromal tumors, and sarcomas arising from the bone were excluded from the study.

Baseline assessment included demographical data, clinical history, physical examination, histopathology on core-needle biopsy, or surgical sample. For staging, contrast-enhanced computed tomography (CECT)/magnetic resonance imaging (MRI) for the primary tumor and CECT chest or positron emission tomography (PET)/CT for metastatic workup were done. Histopathological subtyping and grading were done as per the World Health Organization classification and Federation Nationale des Centres de Lutte Contre le Cancer (FNLCC) grading system, respectively.[2,13,14] The staging was done as per the American Joint Committee on Cancer eighth edition tumor–node–metastasis staging.[15]

In the case of localized and resectable oligometastatic STS, patients were treated with surgery. Only lung-limited metastases were considered for resection. The local multidisciplinary team determined the resectability of lung metastases after discussion in the tumor board. Perioperative therapy with radiotherapy ± chemotherapy was considered as per the standard guidelines and/or tumor board decision. Chemotherapy in the adjuvant or neoadjuvant setting included doxorubicin 25 mg/m^2 for 3 days with ifosfamide 3 g/m^2 for 3 days once every 21 days except in rhabdomyosarcoma, in whom vincristine, actinomycin D, and cyclophosphamide were used. In the metastatic setting, depending on the treating physician's discretion, doxorubicin 25 mg/m^2 for 3 days with or without ifosfamide 2 to 3 g/m^2 for 3 days once every 21 days was used based on fitness, age, and tumor burden.

In the neoadjuvant setting, response assessment was done after 4 weeks of completion of radiotherapy or after three cycles of chemotherapy. The response assessment was done using the RECIST version 1.1 response criteria.

At the end of therapy, CT/MRI of the primary site and CT chest were done to document disease status. Patients who have achieved remission or stable disease were followed-up once in every 3 months with clinical evaluation. CT/MRI of the primary site and CT chest were done in every 6 months, in the first 2 years, and then annually in surgically treated patients. Patients treated with palliative chemotherapy and those who achieved at least stable disease were followed-up with repeat imaging in every 3 months or at clinical progression. For analysis of results, patients who belonged to stage IA to Stage IIIB were grouped under localized STS cohort and those with lymph-nodal or distant metastases under stage IV or metastatic disease. DFS was defined as the time from the date of biopsy to the time of disease recurrence or death, whichever occurred first. Progression-free survival (PFS) was defined as the time from the date of biopsy to the time of progression or death, whichever occurred first. OS was defined as the time from the date of biopsy to the time of death due to any cause. Data regarding the recurrence/progression of the disease and survival were noted. The study was conducted after due approval from the Institutional ethics committee, Army Hospital (R & R), Delhi Cantt (Institutional Ethical Committee Rge no: 99/2017, Date: October 24, 2017). The procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional or regional) and with the Helsinki Declaration of 1964, as revised in 2013. All patients provided written informed consent for participating in this study.

Statistical Analysis

Data were analyzed with SPSS software version 25.0, Released 2017, SPSS for Windows, SPSS Inc., Chicago, Illinois, United States. Descriptive statistics were used for defining the study population and baseline disease characteristics. A Chi-square test was used for the analysis of categorical variables in the study. Kaplan–Meier curves were plotted for survival analyses. A log-rank test was used to compare the data in different groups.

Results

A total of 140 patients of histologically proven STS were included in the study. The median age of the patients was 45 years (range: 18–84 years), and 54.3% (*n* = 76) were male. The median duration of symptoms before presentation was 5 months (range: 1–18 months). Extremity sarcoma constituted the most common type constituting 54.3%, followed by trunk sarcoma (17.86%), retroperitoneal sarcoma (14.29%), thoracic and abdominal visceral sarcomas (9.29%), and head-and-neck sarcomas (4.29%). The most common presentation was swelling seen in 70% (*n* = 99) of the patients; the swelling was painless in 58.5% (*n* = 82) and painful in 12.5% (*n* = 17) of the patients. Other complaints at presentation were abdominal pain in 15% of the patients and weight loss, constitutional symptoms, dry cough, breathlessness, radiculopathy, and bleeding per vagina, each in <10% of the patients. Thirty-five patients had lymph node or distant metastases. The most common site of distant metastases was lungs as seen in 65% of the patients (*n* = 23/35), followed by liver in 25% of the patients (*n* = 9/35). Other sites included pleura (8.5%), spine (8.5%), musculoskeletal (5.6%), and brain (2.8%). The most common histopathologic subtypes were undifferentiated pleomorphic STS (22%), followed by leiomyosarcoma (19%) and synovial sarcoma (16%). The baseline demographic and clinical characteristics of the study population are summarized in ►**Table 1**.

Translocations were assessed in 10% (*n* = 14) of the patients. Of these 11 suspected synovial sarcoma patients, 10 tested positive for X: 18 translocation. In another two probable myxoid liposarcoma patients, one turned out positive for t (12;16)(q13;p11), that is, TLS-CHOP. Furthermore, one round blue cell sarcoma of the abdomen had t (11;22)(p13;q12), that is, EWS–Wilms' tumor 1, conforming to desmoplastic small round cell tumor. Consequently, out of 14 patients, accurate subtyping of STS could be done in 12 patients with the molecular studies. The FNLCC grading was available in 88.6% of the patients (*n* = 124); 35% (*n* = 49) of the patients had grade 3, followed by grade 2 in 34.3% (*n* = 48), and grade 1 in 19.3% (*n* = 27) of the patients. In the remaining 11.4% of the patients (*n* = 16), FNLCC grading could not be evaluated due to various reasons including missing data, insufficient test material, and inapplicability to certain histopathological subtypes such as alveolar STS, angiosarcoma embryonal, and alveolar rhabdomyosarcoma.[15]

Treatment Patterns

Of 105 patients with localized disease, 92 patients had wide local excision, 5 patients required amputation, and, in 8 patients, tumor was not resectable. Among 97 resected patients, 21 patients received neoadjuvant therapy, 58 patients received adjuvant therapy, and 26 patients underwent surgery alone. Of 21 patients receiving neoadjuvant therapy, 10 patients received radiotherapy (preoperative radiotherapy 50 Gy with 2 Gy/# over 25 days) and 8 patients received chemotherapy (doxorubicin 25 mg/m^2 for 3 days + ifosfamide 2–3 g/m^2 for 3 days in a 3-week cycle). Three patients received both radiotherapy and chemotherapy before surgery. Fifty-eight patients received adjuvant therapy in the form of external beam radiotherapy (*n* = 28) or doxorubicin-based chemotherapy (*n* = 16) or both (*n* = 14). Of the 21 patients receiving neoadjuvant therapy, partial response was seen in 38% (*n* = 8/21) of the patients, and the remaining 62% (*n* = 13/21) had stable disease. In the neoadjuvant group, 90.5% of the patients (*n* = 19/21) had complete resection (R0) as compared with 76.3% of the patients (*n* = 58/76) among those who underwent upfront surgery. The relative risk of having incomplete resection in upfront surgery cohort compared with that of postneoadjuvant therapy cohort was 2.37 (95% confidence interval [CI]: 0.59–9.4; *p* = 0.19).

Among 35 metastatic STS patients, 31 patients received chemotherapy; majority (65.7%, *n* = 23/35) received it as palliative therapy and 39.7% (*n* = 8/35) of the patients were treated with curative intent (with surgery + chemotherapy [doxorubicin 25 mg/m^2 for 3 days + ifosfamide 2–3 g/m^2 for 3 days in a 3-week cycle]) in adjuvant setting, following resection of both primary tumor and metastases. Of 23 patients receiving palliative chemotherapy, partial response was seen in 39.1% (*n* = 9/23) and stable disease in 30.4% (*n* = 7/23) of the patients; disease progression was reported in 21.8% (*n* = 7/23) of the patients.

Survival Outcomes

Among localized STS patients, 36 patients had a recurrence or progressive disease. These were 16 local recurrences and 18 distant metastases. And also, six deaths (5.7%, *n* = 6/105) had occurred in localized STS patients. In metastatic STS patients, there were 14 (40%, *n* = 14/35) deaths and 23 (65.7%, *n* = 23/35) patients had disease progression.

After the median follow-up of 14 months (1–25 months), the 1-year PFS and OS rates were 75.8 ± 3.1 and 87.4 ± 3.1%, respectively. For localized STS patients, the median DFS was 20.93 months (95% CI: 18.09–23.77); 1-year DFS and OS rates were 87.6 ± 3.5 and 95.3 ± 2.3%, respectively. For metastatic STS patients, the median PFS and OS were 9.83 months (95% CI: 4.8–14.85) and 23.90 months (95% CI: 7.43–40.36), respectively. One-year PFS and OS rates were 36.53 ± 9.8 and 61.97 ± 9.1%, respectively (►**Fig. 1**).

In stage-IV STS patients (*n* = 35) and those who had curative-intent/definitive therapy (*n* = 8), the median PFS was 20.43 months (95% CI: 5.83–35.02) as compared with 6.83 months (95% CI: 4.92–8.74) in those in palliative therapy cohort (*n* = 27). The median OS was not reached in the curative intent therapy cohort, whereas it was 12.66 months (95% CI: 9.28–16.04) in the palliative therapy cohort, as shown in ►**Figs. 2A** and **B** .

Discussion

STSs are a rare and heterogeneous group of malignant tumors arising from the mesenchymal tissue. As per cancer statistics in the United States, the incidence of STSs is approximately 0.72%,[1] whereas incidence data on STSs in India are unknown as studies in published literature are limited to the retrospective institutional case series.[9-12]

Table 1 Baseline demographic and clinical characteristics of the study population

Parameter	Value
Age in years, median (range)	45 (18–64)
Male, *n* (%)	76 (54.3)
Female, *n* (%)	64 (45.7)
Duration of symptoms in months, median (range)	5 (1–18)
Type of sarcoma, *n* (%)	
Extremity sarcoma	76 (54.3)
Trunk sarcoma	25 (17.86)
Retroperitoneal sarcoma	20 (14.29)
Thoracic and abdominal visceral sarcoma	13 (9.29)
Head and neck sarcoma	6 (4.29)
Histopathologic subtypes	
Undifferentiated pleomorphic STS	31 (22.1)
Leiomyosarcoma	26 (18.6)
Synovial sarcoma	23 (16.4)
Liposarcoma	21 (15)
MPNST	12 (8.6)
Fibrosarcoma	8 (5.7)
Rhabdomyosarcoma	5 (3.6)
Angiosarcoma	4 (2.9)
Others	10 (14)
Tumor characteristics, *n* (%)	
T stage	
T1	20 (14.29)
T2	54 (38.57)
T3	35 (25)
T4	31 (22.14)
N stage	
N0	132 (94.29)
N1	8 (5.71)
M stage	
M0	109 (77.80)
M1	31 (22.20)
FNLCC grade, *n* (%)	
Grade 1	27 (19.30)
Grade 2	48 (34.30)
Grade 3	49 (35.00)
Grade unknown (Gx)	16 (11.40)
Cancer stage, *n* (%)	
IA	6 (4.29)
IB	18 (12.86)
II	8 (5.71)
IIIA	46 (32.86)
IIIB	27 (19.29)
IV	35 (25)

Abbreviations: FNLCC, Federation Nationale des Centres de Lutte Contre le Cancer; MPNST, malignant peripheral nerve sheath tumor; STS, soft tissue sarcoma.

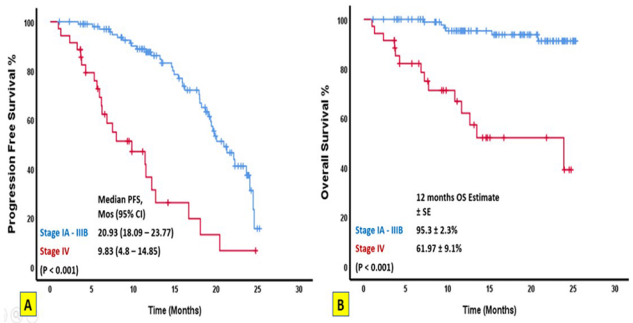

Fig. 1 Kaplan–Meier estimate of progression-free survival (**A**) and overall survival (**B**) between localized soft tissue sarcoma (stage I–stage IIIB) and metastatic soft tissue sarcoma (stage IV). CI, confidence interval; PFS, progression-free survival; OS, overall survival; SE, standard error.

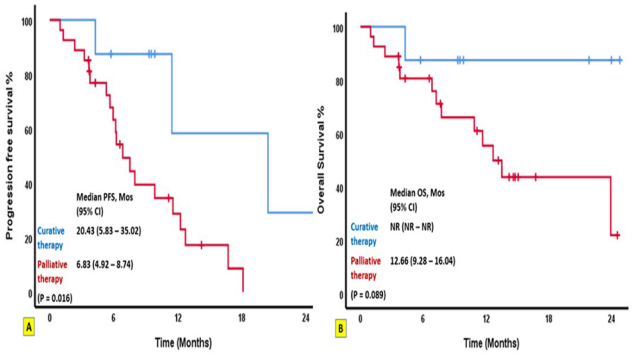

Fig. 2 Kaplan–Meier estimate of progression-free survival (**A**) and overall survival (**B**) among metastatic soft tissue sarcoma patients by type of therapy. CI, confidence interval; NP, not reported; PFS, progression-free survival; OS, overall survival, Mo, months.

STS incidence increases with age, with the reported median age in Western literature falling in the sixth decade.[16–18] In our study, the median age was 45 years, which was comparable to that of other studies from different parts of India, as shown in ►**Table 2**. The median age in the Indian subcontinent is a decade earlier than that in the developed countries. This early occurrence may be due to differing age spectrums of the population in developed countries and India, as a significant proportion of the Indian population is young. A slight male preponderance with a male-to-female ratio of 1.4:1 is reported similar to other studies.[22]

Fifty percent of the patients had symptoms for more than 5 months before diagnosis. Painless nature of swelling compounded with a lack of health awareness and limited access to health care results in delayed diagnosis of these tumors.[23] Besides, lack of health expertise among primary health care providers to accurately differentiate sarcomas from more common benign soft-tissue tumors may result in an inadvertent excision and further delay in referral. This delay in diagnosis may hamper the management of the disease and may affect limb salvage and patient survival.[10] Clinical presentation and management also depend on the anatomical site of origin. In our study, extremity was the most common site, followed by trunk (chest and abdominal wall) and retroperitoneum. The study by Shukla and Deo also reported extremity as the most common followed by chest and trunk. On the other hand, Rastogi et al reported extremity followed by retroperitoneum as the most frequent sites.[12]

Being a rare tumor with more than 50 subtypes makes histopathological subtyping difficult. STSs are often labeled high-grade sarcomas or undifferentiated pleomorphic sarcomas when specific lines of differentiation are not identified.[2] In our study, undifferentiated pleomorphic sarcoma was the most common histopathological subtype, followed by leiomyosarcoma and synovial sarcoma. On the other hand, most Indian studies reported synovial sarcoma as the most common histopathological type, as shown in ►**Table 2**.[9,10,12] However, with standardization of diagnostic criteria and extended immunohistochemistry, some of the undifferentiated pleomorphic sarcomas can be reclassified into specific subtypes.[24]

Furthermore, translocation studies may sort out a few discrepancies, as shown in the GENSARC study.[25] In our study, at least in 12 cases (i.e., ~9%), translocation studies were reclassified to a specific subtype. As discussed above, histopathological subtyping is complex and has poor reproducibility. Several simple histopathological grading systems have been proposed which also provide a better prognostic model. Most commonly used and validated in various studies is the three-tier FNLCC grading system.[14,26]

Management of localized STSs involves a multidisciplinary approach, with surgery being the important modality of therapy. Radiotherapy or chemotherapy in a perioperative setting improves outcomes in tumors of size ≥5 cm, deep seated, or high grade.[27] Preoperative therapy is associated with increased ease of resection, decreased local recurrence, reduced late toxicity, and a trend toward improved survival outcomes.[3,28,29] Our study was not randomized, and neoadjuvant therapies were considered in relatively advanced tumors on a case-to-case basis after tumor board discussions. We observed nonsignificantly higher complete resection rates in the neoadjuvant cohort.

In our study, 1-year PFS and OS rates were 75.8 and 87.4%, respectively, for the overall cohort. In a large retrospective study in Germany based on 24,753 patients' survival data, the reported median survival was 5.83 years and a 1-year survival probability was 77%.[30] For localized STS patients, the median DFS estimate was 20.93 months, with 1-year DFS and OS rates being 87.6 and 95.3%, respectively. Bajpai et al reported that 3-year PFS and OS were 48 and 64%, respectively. The outcomes in other Indian studies are shown in ►**Table 2**. The variation in survival outcomes reflects the study population's heterogeneity, such as stage, histological subtypes, grades, and use of different treatment options.

Among metastatic STS patients, the majority received systemic chemotherapy therapy. Still, 25% of the patients also underwent curative resection surgery of both locoregional disease and metastases followed by systemic therapy. In the METASARC observational study, 48.6% of the patients received definitive therapy for locoregional disease and metastases.[31] In our study, the majority of the patients had unresectable metastases.

Table 2 Key adult STS studies from India in published literature

Author, Institution (year of study)	Type of study	Sample size/ type of STS	Median age (years)/M:F	Most common histopathology, (%)	Median follow-up (mo)	Outcomes
Bansal et al, CRI, Dehradun (2018)[19]	Retrospective analysis	43/extremity sarcoma	48/1.8:1	1. Pleomorphic sarcoma, (20.9%) 2. Synovial sarcoma, 16.2%	47	At median follow-up, EFS: 41.49% and OS: 47.64%;
Bajpai et al, TMH, Mumbai (2018)[9]	Retrospective analysis	189/extremity sarcoma	41/1.7:1	1. Synovial sarcoma (31%)	51	3-year EFS: 48%, and OS:64%;
Gupta et al, Wenlock Hospital, Mangalore (2009)[20]	Retrospective study	51/all types	Fourth and Fifth decades/1.36:1	1. Liposarcoma (17.5%) 2. Leiomyosarcoma (15.7%)	NR	NR
Badanale et al, TMH, Mumbai (2009)[11]	Histopathologic review of biopsies	328/trunk and extremity STS	40.5/1.7:1	1. Synovial sarcoma 16% 2. Leiomyosarcoma 11.3% (45/328)	NR	NR
Rastogi et al AIIMS, New Delhi, (2018)[12]	Retrospective analysis of prospective database	156/advanced STS	41 (17–77)/1.73:1	1. Synovial sarcoma (22%) 2. MPNST (16%),	13 months	Median OS: 16 months
Shukla and Deo AIIMS, New Delhi, 2011 [10]	Retrospective analysis of prospective database	300/all types	40.6 (10–85)/2:1	1. Synovial sarcoma (15%) 2. MFH (13.9%).	NR	NR
RGCI, Delhi Tiwari et al, 2017 [21]	Retrospective study	112/extremity and trunk	Approximately 50% were less than 50 years	1. Synovial sarcoma (23.2%) 2. UPS (22.3%)	NR	5-year EFS:42.1%, OS: 73.1%

Abbreviations: AIIMS, all India Institute of Medical science; CRI, Cancer Research Institute; EFS, event free survival; F, female; M, male; MFH, malignant fibrous histiocytoma; MPNST, malignant peripheral nerve sheath tumor; NR, not reported; OS, overall survival; RGCI, Rajiv Gandhi Cancer Institute; STS, soft tissue sarcoma; TMH, Tata Memorial Hospital; UPS, undifferentiated Pleomorphic sarcoma.

The presence of metastases predicts a dismal outcome in STS, mainly if resection of both primary and metastases is not feasible. The median PFS and OS of metastatic STS patients in our study were 9.83 months (95% CI: 4.8–14.85) and 23.90 months (95% CI: 7.43–40.36), respectively. A retrospective analysis of 156 advanced STS patients from North India reported a median OS of 16 months.[12] Another study from the United States of 363 metastatic STS patients reported a median OS of 22 months and 17 months among treated and untreated patients, respectively.[18] The basis of a good outcome in our metastatic patients may be a result of improved outcomes in 25% of these patients who underwent complete resection. Among those who underwent curative resection, median OS was not reached as compared with 12.33 months among those who received palliative therapy alone. In the METASARC study, those who received locoregional therapy/definitive therapy had a better probability of survival.[31] Similarly, the European Organization for Research and Treatment of Cancer Soft Tissue and Bone Sarcoma Group reported a 38% 5-year OS among 255 STS patients who underwent metastasectomy.[32]

Strengths and Limitations

Our study being prospectively conducted helps to understand the spectrum of STS patients managed at a tertiary care center. It highlights the difficulties in accurate histological subtyping in a real-world setting. It acts as a stepping stone for further studies in a neoadjuvant setting and oligometastatic STS. The limitations of this study include the retrospective design, single-center study nature, and small sample size with a short follow-up; randomized trials with long-term follow-up are needed to delineate differences in the outcomes better.

Conclusion

The median age of STS patients is a decade earlier than that in developed countries. Undifferentiated pleomorphic STS was the most frequent histological type. The median OS was not reached for localized STS patients, with 1-year OS rate being 95.3 ± 2.3%. The median OS for metastatic STS patients was 23.90 months (95% CI: 7.43–40.36). Among metastatic STS, the median OS was not reached for those who underwent curative resection versus 12.66 months (95% CI: 9.28–16.04) for those who received systemic therapy alone. STS requires a dedicated multidisciplinary team for appropriate management. To advance sarcoma care and research in India, multiple institutes should cooperate and form the Indian sarcoma network group.

Conflicts of Interest

There are no conflicts of interest to declare.

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