Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Oncology

Soft Tissue Sarcomas: Epidemiological Profile and Evolving Clinical Aspects and Management in the Oncology Radiotherapy Department at Marrakech University Hospital

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DOI: https://doi.org/10.36347/sjmcr.2025.v13i09.047 | **Received:** 08.07.2025 | **Accepted:** 18.09.2025 | **Published:** 23.09.2025

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Abstract | **Case Report**

Soft tissue sarcomas (STS) are rare malignant tumors, characterized by abnormal proliferations arising from connective tissue. They represent a heterogeneous group of tumors, both clinically, histologically, and prognostically. Due to their rarity, evolutionary heterogeneity, and histological diversity, soft tissue sarcomas (STS) pose significant challenges in terms of diagnostic pathology, prognostic evaluation, and the development of therapeutic strategies. Our study is etrospective of 65 cases of soft tissue sarcomas managed by the Oncology- Radiotherapy Department of the CHU Mohammed VI in Marrakech, over a five-year period, from January 2019 to December 2023. The aim of our study is to highlight the epidemiological profile, clinical presentation, diagnostic methods, histopathological profile, therapeutic strategy, evolution, and prognosis of these tumors within our healthcare institution. The mean age of our patients was 40.1 years, with a slight male predominance (52.3%). The average consultation delay was 7 months, with the main reason for consultation being the tumor syndrome. Tumors were predominantly located in the lower limb (60%), compared to 29.3% in the upper limb. MRI was the reference examination performed in 80% of our patients, while histopathological examination was conducted in all patients. Histological evidence was obtained for all patients : 63.1% through surgical biopsy and 36.9% through percutaneous biopsy. Synovial sarcoma (40%) was the predominant histological type, followed by liposarcoma (20%) and leiomyosarcoma (10.8%). The initial treatment was surgical, with conservative surgery performed in 86.1% of patients and radical surgery in 13.9%. Surgical margins were R0 in 58.6% of cases, R1 in 34.5%, and R2 in 6.9%. Adjuvant radiotherapy was associated with surgery in 72.7% of patients, while chemotherapy was administered to 55.6% of patients. Follow-up allowed the detection of 18 cases of local recurrence (40%) and 8 cases of pulmonary metastases (17.8%). Evolution was determined in 38 patients, with clinical remission maintained in 7 patients. Five patients died, and seven patients were lost to follow-up.

Keywords: Epidemiology, Soft Tissue Sarcomas, Prognosis, Treatment, Histopathology.

INTRODUCTION

Soft tissue sarcomas (STS) represent only 2% of adult cancers. They are rare tumors of mesodermal origin, such as muscles. fasciae, tendons, adipose tissue and vessels, and do not concern tumors of osseous origin. Liposarcomas and leiomyosarcomas are the most frequently encountered subtypes. (2)

This type of tumor can develop anywhere in the body, with a wide range of clinical and histological variations, complicating diagnosis and making treatment more difficult. histological heterogeneity makes immunohistochemistry and molecular biology essential. Approximately 27% of soft tissue sarcomas occur in the lower limbs. Localisations in the upper limbs and other sites account for less than 15%. The most frequent sites are the trunk, mediastinum and retroperitoneal region (less than 12%) (3).

Treatment relies mainly on surgery, which is the mainstay of soft-tissue sarcoma therapy: The aim of this study is to report on the epidemiological particularities and evolutionary aspects of MTS in the oncology-radiotherapy department of the Mohammed VI University Hospital in Marrakech.

METHOD

This is a descriptive retrospective study carried out over a five-year period, from 2019 to 2023. It covers 65 cases of soft-tissue sarcoma treated at the

radiotherapy oncology department of the Mohammed VI University Hospital Center (CHU) in Marrakech.

Data were collected from the hospital registers of the radiotherapy oncology department of the Mohammed VI University Hospital, Marrakech, and from the HOSIX computer system.

Inclusion criteria were as follows: all patients with diagnosed soft-tissue sarcomas whose sarcomatous nature had been confirmed by pathological analysis.

Data collection was carried out with respect for patient anonymity and confidentiality and patient confidentiality was respected.

RESULTS

We collected 65 cases of soft-tissue sarcoma, representing 0.9% of the total number. These were treated cancer patients, with a mean age of 40.1 [24-74] years.

Our study shows slight male predominance, with 47.7% women for 52.3% men, with sex ratio of 1.04. In our series, 73.8% of patients had AMO (compulsory health insurance) coverage.

Four patients had a history of trauma, and 1 had been exposed to ionizing radiation. The average consultation time was 7 months, with extremes ranging from 3 to 18 months.

The appearance of a mass was the main symptom revealing the disease, observed in 75.4% of patients, followed by the presence of pain. This latter was reported in 58.4% of cases, and deterioration in general condition in 13.9% of cases.

The location of the sarcoma was as follows: lower limb (60%), upper limb (29%), other (12%). Clinical examination revealed swellings of varying sizes, ranging from 3 to 24 cm. Tumour size was greater than 5 cm in 92.3% of patients. cm in 92.3% of cases. Inguinal lymph node involvement occurred in 3.07% of cases. Axillary adenopathy accounted for 1.54%. 16% of soft tissue sarcomas were metastatic to the lung and liver. MRI was performed in 80% of our patients, and in all cases observed,

The lesion showed enhancement on imaging, as indicated by the lesions appearing in hyposignal T1 in 78.9% of cases and hypersignal T1 in 21.1% of cases. Tumour necrosis was common in 9.24% of cases. Lesions were T2 hypersignal in 86.6% of cases and T2 hypposignal in 15.4% of cases.

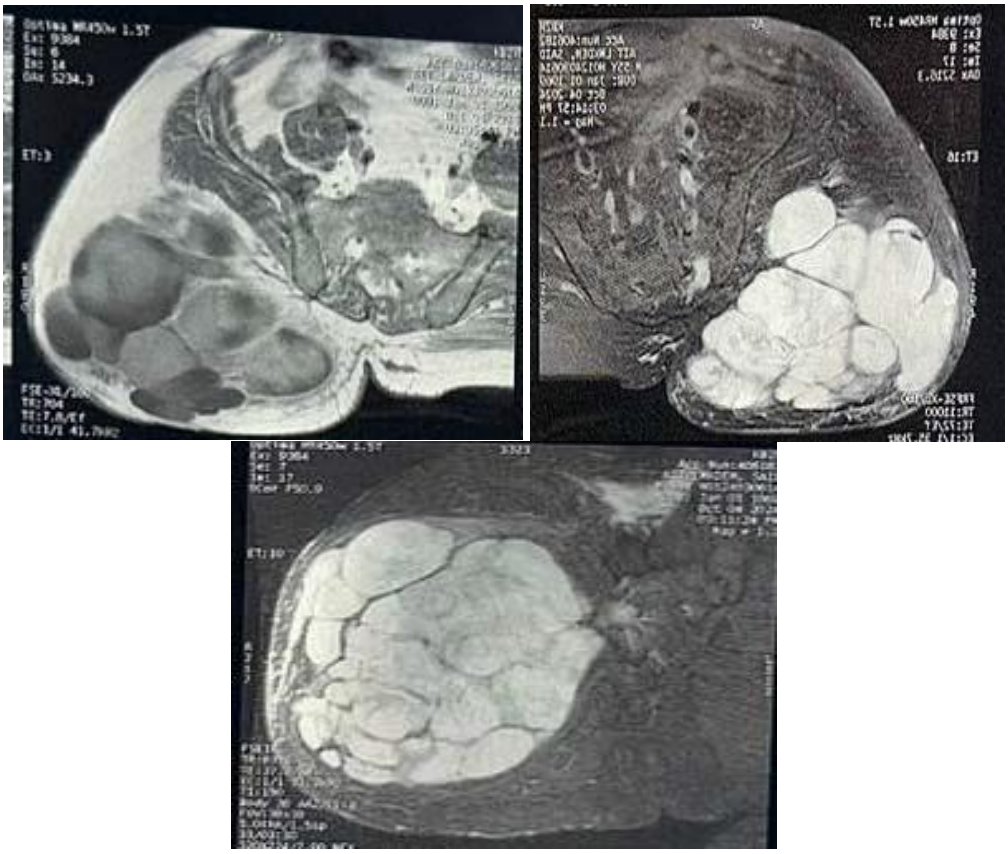

Figure 1: Presence of a large, well-demarcated, poly-lobed mass occupying the right gluteus medius and rectus maximus muscles. The lesion shows a T1 hyposignal in relation to the muscles, with areas of T1 hypersignal and T1 fat sat, as well as an area of T2 heterogeneous hypersignal. The mass measures 20 x 20 cm and extends to a height of 21 cm

Surgical biopsy was performed in 63.1% of these cases. Percutaneous biopsy was performed. Percutaneous biopsy was performed in 36.9% of cases. Immunohistochemistry was performed on all patients.

Statistically, synovialosarcoma is the most common type of sarcoma, accounting for sarcoma, accounting for 40% of cases (undifferentiated synovialosarcoma: 12 cases, or 18.5%; biphasic synovialosarcoma: 8 cases, or 12.3%; and monophasic synovialosarcoma: 6 cases, or 9.2 %). Liposarcomas also play a significant role, accounting for 20% of cases.

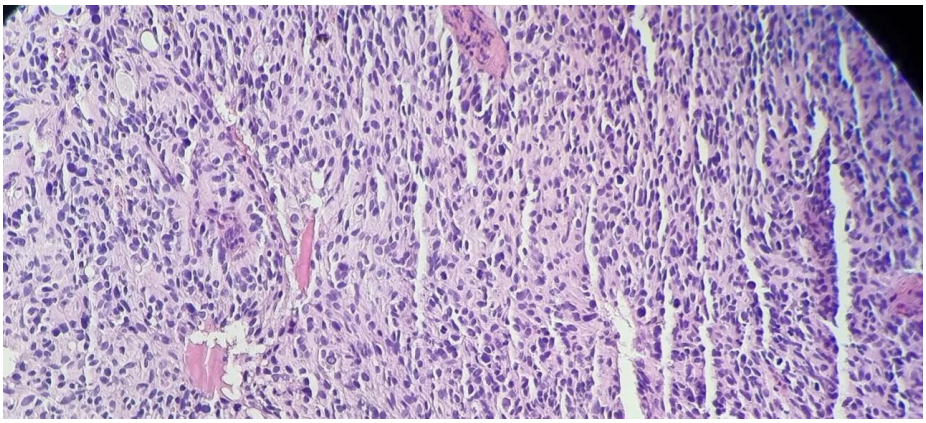

Figure 2: HEx40 - Monophasic mesenchymal proliferation with vascularisation (Synovialosarcoma)

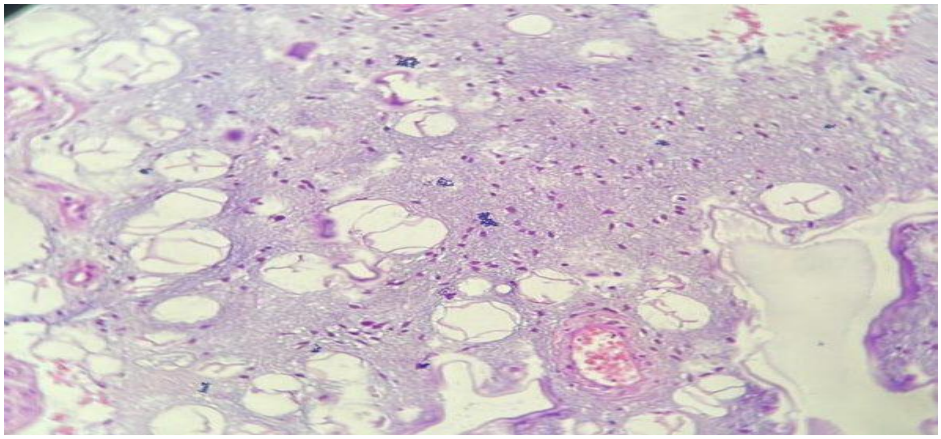

Figure 3: HEx40 - Malignant lipomatous proliferation within a myxoid stroma stroma (myxoid liposarcoma)

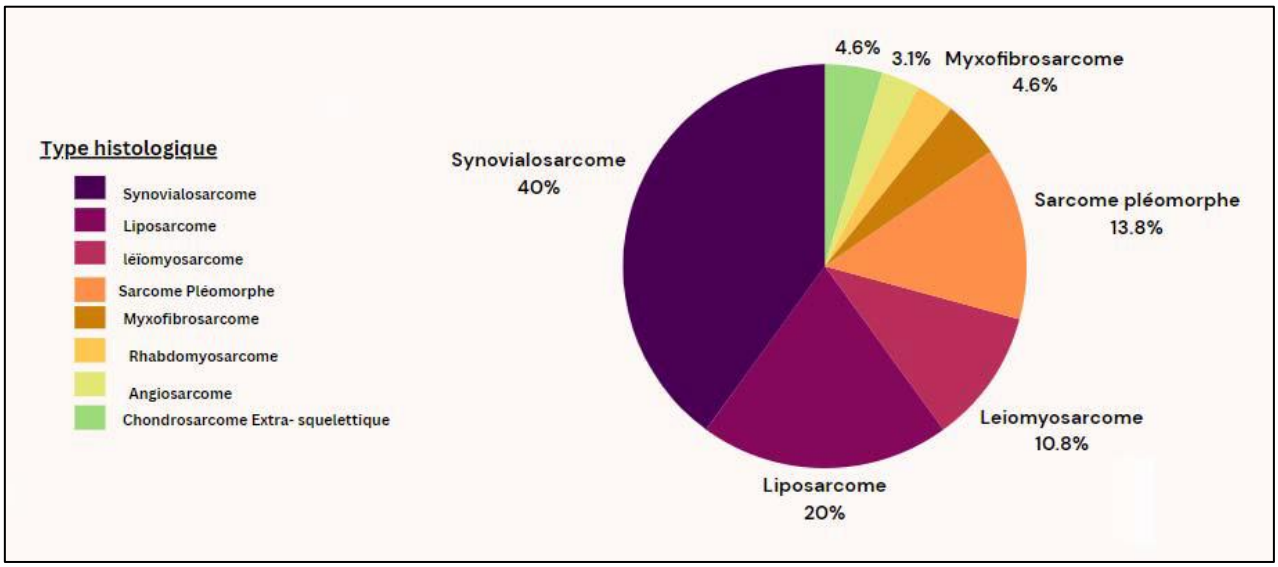

Figure 4: Distribution of VTS by histological type

The histological grade was specified in 90% of patients, of whom 21% had presented with grade 1 STM, 47% with grade 2 STM and 32% with grade 3 STM.

Surgery was performed in 80% of cases. It was conservative in 86.1 and radical in 13.9% of cases. Of these patients who underwent surgery 58.6% had R0 margins, 34.5% had R1 margins, and 6.9% had R2 margins.

In our study, 73.3% of patients received radiotherapy. curative in 87.9% of patients and palliative in 12.1%. All patients benefited from 3D-CR.

In contrast, chemotherapy was administered in 55.6% of patients, as neoadjuvant therapy in 33.3% and palliative in 66.7%. used were ifosfamide and doxorubicin. None of the patients included in our study received targeted therapy.

We observed a local recurrence rate of 40%. approximately 17.8% of patients developed distant metastases during their treatment. treatment. Clinical remission was achieved in 15.5% of patients. patients. In addition, 15.5% of patients were lost to follow-up. Finally, the mortality rate was 11.2%.

DISCUSSION

Soft tissue sarcomas (STS) are rare tumours, accounting for less than 1% of all malignant neoplasms [4,5]. In the United States soft tissue cancer accounts for 0.7% of all new cancer cases [6] According to the Enzinger and Weiss study: they are less frequent than benign tumours The peak incidence of soft tissue sarcoma (STS) is generally between the ages of 40 and 60 [7].

However, the age of onset of sarcoma varies according to the type of histological type. Embryonal rhabdomyosarcoma occurs almost exclusively in children, synovialosarcoma most often affects young adults, whereas undifferentiated pleomorphic cell sarcoma, well-differentiated liposarcoma, leiomyosarcoma and myxofibrosarcoma predominate in older subjects [8]. In our series, the average age of patients was 40.1 years.

Some studies describe a high incidence in males, with a progressive increase over the last three decades, observed only in this population [15]. In our series, we observed a slight male predominance, accounting for 52.3% of cases, with a sex ratio of 1.04. Although the majority of sarcomas occur sporadically, several genetic disorders genetic disorders increase the risk of developing this type of cancer: [10-12] such as peripheral neurofibromatosis (NF1): Also known as

Von Recklinghausen neurofibromatosis, this is an autosomal dominant linked to malignant tumours of the peripheral nerve sheath and other sarcomas. other sarcomas. Central neurofibromatosis (NF2): Associated with bilateral acoustic neuromas neuromas, this condition is linked to the loss of the tumour suppressor gene

NF2 tumour suppressor gene located at 22q11.2q12. Some nerve sheath tumours also show a loss of 17p, affecting the p53 gene in particular.

Familial adenomatous polyposis (FAP): Approximately 10% of patients with FAP develop a fibromatosis or desmoid tumour. develop a fibromatosis or desmoid tumour, often in the mesentery.

Gardner's syndrome, associated with benign tumours of the soft tissues skin and bone tumours, is also linked to mutations in the APC gene, located in the FAP locus at 5q21-22. Fibromatoses often present with trisomies [4, 5 and 20] or loss of heterozygosity by deletion of 5q.

Li-Fraumeni syndrome: This syndrome exposes patients with p53 germline mutations to an increased risk of soft tissue sarcomas in children and breast Familial retinoblastoma: Patients who survive this rare form of eye cancer develop sarcomas, including osteosarcoma, due to loss of the tumour suppressor gene of the Rb tumour suppressor gene at 13q14. Approximately 10-20% of survivors develop a sarcoma

The risk of developing a sarcoma with a history of radiotherapy (0.5% of patients who have received intensive radiotherapy for a malignant tumour and survived beyond five years develop a soft tissue sarcoma in the irradiated areas). Immunosuppression is a risk factor for MTS, as is HIV infection and transplantation.

Clinically, soft tissue sarcoma typically manifests itself as a triad: presence of a tumour mass, pain and functional discomfort [13]. (Soft tissue sarcomas usually appear as painless masses with progressive growth) [14].

(Those located in the extremities are often detected earlier, whereas pelvic sarcomas are diagnosed late due to their deep location, which prevents palpation in the early stages) [15]. In our study, swelling was the most frequent symptom, reported in 49 patients, representing 75.4% of the sample.

Pain depends mainly on the anatomical location or volume of the tumour [16], but pain is not a sign of malignancy. In our study series, pain was reported in 38 patients, i.e. 58.4% of the sample.

Functional impotence of the affected limb occurs mainly at an advanced stage of the disease in patients with a periarticular location of the tumour, a large tumour volume, or a combination of the two [9].

Lymph node involvement in soft tissue sarcomas is rare but prognostically significant.

prognostic significance, with histological subtype being a key risk factor for metastasis [17,18]. Node metastases associated with soft tissue sarcoma occur in less than 5% of patients [19].

Early detection (within 8 months) may indicate a poor prognosis and is correlated with a reduced overall survival rate [20].

In parallel with the data in the literature, our study revealed macroscopic lymph node involvement in 4.6% of patients. MRI is the paraclinical examination of choice to assess tumour extension and also to look for malignancy criteria.

Positron emission tomography is of vital importance in the management of soft tissue sarcomas, especially as an extension assessment.

Genetic studies based on karyotype analysis and the use of various molecular biology techniques are increasingly important in understanding and identifying sarcomas.

Treatment is essentially based on surgery with a wide range of tumour exeresis and oncological treatments, either radiotherapy or neoadjuvant or adjuvant chemotherapy, and the decision is taken by a multidisciplinary consultation meeting.

Soft tissue sarcomas are potentially aggressive locally and metastatically, especially in the pleura and lung, depending on the following prognostic factors: tumour size, depth of lesion, histological grade, staging and quality of surgical excision.

CONCLUSION

Soft tissue sarcomas represent a rare pathology with a complex and multidisciplinary management.

Morbidity and mortality rates have fallen sharply over the past fifty years, largely thanks to advances in early diagnosis, advanced surgery, and adjuvant and neoadjuvant therapies

Soft tissue sarcomas are mainly found in the lower limb, particularly the thighs. Pain and tumor syndrome are the main functional signs observed in patients.

Magnetic resonance imaging (MRI) is the examination of choice for assessing tumour extension, guiding biopsies and orienting histological diagnosis.

The most frequently encountered types of sarcoma are synovialosarcoma, followed by liposarcoma and leiomyosarcoma. Grade II sarcomas were the most common in this study.

Wide tumor resection remains the most frequently used treatment. radiotherapy is recommended as a complement for patients with incomplete excision or those at high risk of recurrence. Chemotherapy is mainly indicated in metastatic disease.

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