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*Methods:* This is a retrospective descriptive study that has been reviewing a 55 patient with soft tissue sarcoma presented to the hospital during the period from January 2010 to August 2012.

Results: the mean age is 51 years. The peak age range was 40–60 years, with the most common site affected by soft tissue sarcoma is the lower limbs (43.6%) and common size of presentation more than 15 cm in the greatest dimension (47.3%). (36.4%) of patients presented with past history of excision. MRI and U/S stand as the most common imaging requested (29.1%) and (30.6%) respectively. Excisional biopsy done for the majority of patients (58.2%) in which most of them has clear margin (56.4%).

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# Soft Tissue Sarcoma (STS) in Gezira Sudan Presentation, Management and Outcome

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Conclusion: The late presentation of the tumors as reflected by huge size at first presentation making the management a real challenge. Soft tissue sarcoma found to be common in our community and causing big problem in its treatment because of its high rate recurrence and delayed presentation.

## I. Introduction and Literature Review

oft tissue sarcoma refers to a sarcoma that arises from the non-bony tissues of the extremity, pelvis or trunk. These tumors can arise directly from the muscles or from the other soft tissues between the muscles. They can also arise from the skin and subcutaneous tissues and fascia. The fascia is a thick sheet of tissue that underlies the skin and acts like an envelope around all the muscles of an extremity. It

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separates the skin and underlying fat from the muscles, blood vessels, nerves and bones of the extremities, pelvis and trunk.<sup>(1)</sup>

Soft tissue sarcomas (STS) are a rare group of cancers compromising ~ 1% of all malignancies. There has been a slight increase in incidence, and half of STS patients are over the age of 65 years. (2) the pathogenesis of most STS remains unknown. Genetic, environmental and immunological factors have been identified as risk factors. It is likely that the cause of STS is multifactorial. STS may arise in any part of the body, predominantly in the extremities (45% lower limb 29% upper limb (16%), trunk (25%), head and neck (13%), and retroperitoneum (9%) (2) The etiology of most benign and malignant soft tissue tumors is unknown. In rare cases, genetic and environmental factors, irradiation, viral infections and immune deficiency have been found associated with the development of malignant soft tissue tumours. There are also isolated reports of soft tissue sarcomas arising in scar tissue, at fracture sites and close to surgical implants<sup>(3)</sup> however, the large majority of soft tissue sarcomas seem to arise de novo, without an apparent causative factor. Several studies, many of them from Sweden, have reported an increased incidence of soft tissue sarcoma after exposure to phenoxyacetic herbicides, chlorophenols, and their contaminants (dioxin) in agricultural or forestry work(4). Other studies have not found this association. One explanation for different findings may be the use of herbicides with different dioxin contaminations (5) The reported incidence of post-irradiation sarcoma ranges from some few per thousand to nearly one percent. Most incidence estimates are based on breast cancer patients treated with radiation as adjuvant therapy<sup>(6)</sup>. Human herpes virus 8 plays a key role in the development of Kaposi sarcoma and the clinical course is dependent on the immune status of the patient (7). Epstein-Barr virus is associated with smooth muscle tumors in patients with immunodeficiency<sup>(6)</sup>.

## II. CLINICAL PRESENTATION AND DIAGNOSIS

Most soft tissue sarcomas of the extremities and trunk present as painless, accidentally observed tumors, which do not influence function or general health despite the often large tumor volume. The seemingly innocent presentation and the rarity of soft

tissue sarcomas often lead to misinterpretation as benign conditions. Epidemiological data regarding size and depth distribution for benign and malignant soft tissue tumors' in Sweden have been used to formulate simple guidelines for the suspicion of a sarcoma: superficial soft tissue lesions that are larger than 5 cm and all deepseated (irrespective of size) have such a high risk (around 10 percent) of being a sarcoma(8-9) that such patients should ideally be referred to a specialized tumor centre before surgery for optimal treatment (10-12).

Imaging of soft tissue sarcomas is often helpful diagnostic(13). many cases may be Roentgenograms are useful when the lesion is large or causes injury or damage to a bone. A computed tomography (CT) scan is frequently valuable in clearly displaying the alteration in the shape and size of a part and the character of the soft tissue material<sup>(14)</sup>. The magnetic resonance image (MRI) provides the most useful information about soft tissue sarcomas and not only demonstrates the shape and size of the lesion often with remarkable clarity, but also provides some clues as to the nature of the lesion. (13, 14-16) Positron emission tomography (PET) scans are for the most part active over the soft tissue sarcoma, but more importantly, they are useful if seeking metastases, additional lesions, or particularly lymph node extensions<sup>(17-18)</sup>. Biopsy is a key step in the diagnosis of soft-tissue tumors. The position of the biopsy site within the lesion has a major significance because soft-tissue tumors may have regional morphologic variations (19-21). As a result of that heterogeneity, multiple samples are required to establish a diagnosis. Carcinomas, by contrast, are commonly homogeneous and a single tissue core or aspirate is sufficient for diagnosis. The biopsy incision or the needle puncture hole, and the tract to the tumor, must be made with the planned surgical incision site so that they will be included within the surgical specimen. Open incisional biopsy is a reliable diagnostic method because it allows the pathologist to evaluate cellular morphologic features and tissue architecture from different sites of the lesion. Furthermore, it provides material for performing ancillary studies such as immune-histochemistry, cytogenetics, molecular genetics, flow cytometry and electron microscopy.

#### III. Mangement of Soft Tissue Sarcoma

Althouah surgery remains the therapeutic modality in soft tissue sarcoma, the extent of surgery required, along with the optimum combination radiotherapy and chemotherapy, controversial. For high grade sarcomas, greater than 5 cm, there are several possible approaches to treatment that are based on not only achieving good local control but also reducing the risk of developing subsequent metastasis. The value of systemic chemotherapy depends on the specific histological

subset of the sarcoma. Chemotherapy is usually indicated as primary "neoadjuvant" therapy in the treatment of Ewing sarcoma and rhabdomyosarcoma. Adjuvant chemotherapy is indicated for these specific tumour types, even if the primary site has been resected, because of the very high risk of metastasis. For other histological types of soft tissue sarcoma the value of systemic chemotherapy remains controversial. The histological type and location of disease are important predictors of sensitivity to chemotherapy and thus may help in decisions on the potential benefit of chemotherapy. The majority of the randomized chemotherapy trials have shown no significant impact on overall survival; however they have found that chemotherapy does improve disease free survival, with loco-regional improved local and Radiation is often helpful in decreasing the likelihood of local recurrence and possibly metastasis. It may be given preoperatively, intra-operatively and sometimes postoperatively. (22)

Brachy therapy, which consists of inserting a catheter and implanting a radioactive source for usually a 3-day period, appears to decrease the likelihood of local recurrence. (23)

The objectives of this work are to evaluate the management of soft tissue sarcoma (STS) In fields of the clinical presentation of STS regarding the age, sex, site, size, the surgical procedure that done and its complications and the effect of radiation and chemotherapy in soft tissue sarcoma.

#### IV. Patients and Methods

This is a retrospective descriptive study that has been investigating the patients of soft tissue sarcoma presented to Medani teaching hospital & oncology hospitalduring the period from January 2010 to August 2012. It include a 55 case of STS, patients who presented with tumor not amenable to surgical intervention either not fit for surgery or with distal metastasis; also those patients whom were less than 15 years of age and are treated in the Pediatric Center were excluded.

#### V. Results

During the period from January 2010 through August 2012 (32 month), out of a total of 55 soft tissue sarcoma 29 of patients were female 26 were male with ratio of 1.1: 1, 43 of them live in rural area (78.2%) and 12 live in urban (21.8%). The ages ranged from 17 to 85 years, with a mean age 51 years. The peak age range was 40–60 years 21 patients (38.2%) (Table 1)

We found that the lower limb is the most common site affected by soft tissue sarcoma 24 (43.6%) then the retroperitoneal region 13 (23.6%) then trunk 9 (16.4%) then upper limb 6 (10.9%) then head and neck 2 (3.6%) others 1 (1.8%) (Fig-1). also the

common size of presentation more than 15 cm in the greatest dimension 26 of patients (47.3%) and few of them present with tumour less than 5 cm in its greatest dimension 5 patients (9.1%). At presentation 20 cases of our studied patients (36.4%) had past history of surgery for the presenting mass, most of them in a form of excision. The imaging done for our studied patients 16 of them (29.1%) imaged by doing MRI, U/S done for 17 (30.6%), X rays done for 13 (23.6%) and CT done for 9 (16.4%) of them. The biopsy taken for diagnosing our studied patients are excision done for 32 patients (58.2%), Tru-cut biopsy for 16 (29.1%) and incision for 7 patients (12.7%).38 patient (69.1%) excised their tumour, 14 of them (25.5%) just debulking done for them, and amputation of a limb done for 3 patients (5.5%).

The result of histopathology showed that 31 patients (56.4%) excised with clear margins, and 24 patients (43.6%) with involved margins.

46 patients (83.6%) their surgical procedures end without complication, 6 patients (10.9%) have skin loss treated by skin graft , one patient end with ulnar

nerve injury, one end with radial artery injury which was ligated and one them had knee stiffness treated with physiotherapy.

All the patients under study received adjuvant therapy 29 of them treated by radiotherapy alone, 17 of them by chemotherapy alone, and 9 of them treated by both.

The most common type of soft tissue sarcoma is fibrosarcoma 19 (34.5%) then liposarcoma 11 (20%) then rhabdomyosarcoma 9 (16.4%) then MFH 8 (14.5%) then leiomyosarcoma 6 (10.9%) then synovial and chondrosarcoma sarcomas 1 (1.8%). (Fig-2)

There was high rate of recurrence in our studied sample 39 patient (70.9%) within the 1<sup>st</sup> year after surgery, 15 patients (27.3%) present with distal metastasis 10 of them to the lungs, 3 to the liver, and 2 to the bone.

Also we found that recurrence of 14 out 19 fibrosarcoma, 8 out of 9 rhabdomyosarcoma, 7 out of 11 liposarcoma, 6 out of 8 MFH, 3 out of 6 leiomyosarcoma, and 1 out of 1 in chondrosarcoma.

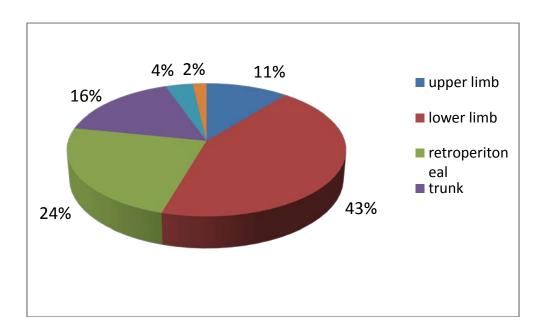


Fig. 1: Site of STS of patients treated Madani hospital 2010-2012

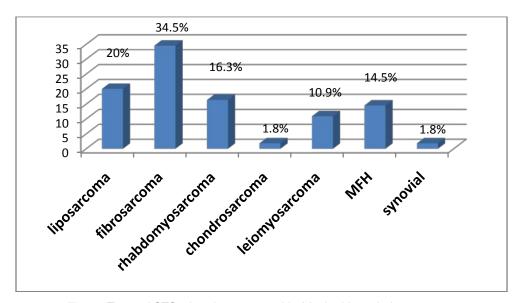


Fig. 2: Type of STS of patients treated in Madani hospital 2010-2012

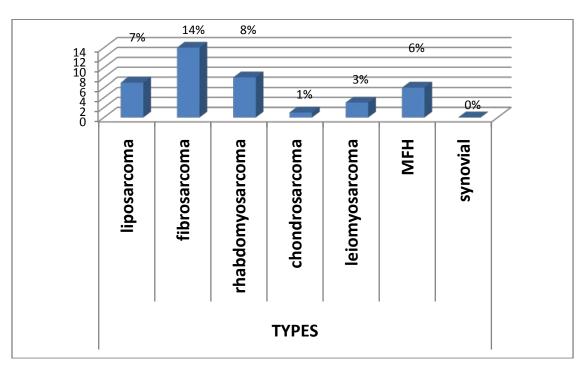


Fig. 3: Types VS Recurrence Crosstabulation of patients with STS in Madani hospital 2010-2012

Table 1: Frequency of the affected age group of patients with STS in Madani hospital 2010-2012

	Frequency	Percent
<20years	4	7.3%
20-40 y	15	27.3%
40-60 y	21	38.2%
>60 y	15	27.3%

Type Yes No Liposarcoma 6 13 Fibrosarcoma 8 Rhabdomyosarcoma Chondrosarcoma 1 0 Leiomyosarcoma 1 5 MFH 1 7 Synovial 0 1

Table 2: TYPES VS METS Crosstabulation of patients with STS in Madani

#### VI. Discussion

This study done over a periods of 32 months in 55 case of soft tissue sarcoma, male: female ratio 1:1.1 which is slightly high in female, comparing it with study done by Yosif A.A, in Omdurman Teaching Hospital 2008<sup>(24)</sup> of 38 cases from sep 2005 to march 2008 which revealed 1:1 ratio also study done in Nigeria<sup>(25)</sup> done in 232 cases from Jan 1985 to Dec 2006 another study done in USA in Memorial Sloan-Kettering Cancer Center (MSKCC)<sup>(26)</sup> over 17 years from July 1982 to June 1999, a total of 1092 patients treated from STS affecting extremities 54% of them were male 46% female.

The age of patients in this study was ranging from 17 years to 85 years, with mean age 51 years. It is revealed that there is no pediatric case treated with STS in the centre at that period of time when this study was conducted.

Yosif A.A found a similar result, with age ranging 16 years to 81 years with slightly lower mean age 44 years. And also there was no pediatric case.

In this study fibrosarcoma found to be the most common type (n-19) 34.5% the same as Yosif A.A (n-11) 28.9%. That differ from the other study in Nigeria and internationally where they found MFH as most common type encounter as found in the study done in MSKCC 28% of patients were MFH.

The geographical distribution of patients in this study revealed that the rural people suffer more; they constitute 78.2% of patient. This reflected their problem of poor health care, because they live far away from medical centers. The geographical distribution not included in the others studies.

The common site that are affected in this study is the lower limb account 43.6% of the studied patients, in the lower limb the thigh is affected more than the other part, this may be due to the bulk of the muscle found in this site, which is in agreement with locoregional and international data. (25, 26)

Most patient present late in the course of their disease and that is reflected by large tumour size at presentation, 47.3% of our studied patients present with

size more than 15cm in greatest dimension which is the same finding in Yosif AA study, but in the study which was done in MSKCC most of patients present with mass less than 5cm in diameter constituting 38% of patients.

The history of past excision found in 36.4% of patients that affects the recent surgery in achieving clear margins (56.4% only excised with clear margins) without injury to the nearby structure and wound complication, which was also found by Yosif AA, and in the study of MSKCC 37% of patients had their tumours resected elsewhere then referred to the centre.

When Yosif AA study was conducted he recommended to use the advance tool of imaging to be used specially CT&MRI, in this study we found that there is partial improving in this setting, the MRI used in 29.1% and CT in 16.4% in studied patient.

The surgery performed for patients in this study, 69.1% of them their tumour excised with clinical clear margins, 25.5% of them just debulking done for them because of large size of the tumor or incasing vital structures, and amputation done for 5.5% of the patients little lower than Yosif AA where it done for 8.4% of patients.

Because our patients have seen in a combined clinic including surgeons, oncologists, radiologists, and histopathologists they have full chance of receiving adjuvant therapy. 52.7% of them received radiotherapy, 30.9% received chemotherapy and 16.4% of patients treated by radio chemotherapy. Where in Yosif AA study radiotherapy done for 47.4% and chemotherapy done for 18.4% of patients. There were 4 patients in Yosif AA study treated by BCG which is not carried in this study.

Follow up of patients in this study done when they came in the combined clinic and others by phoning them. We found that there is high rate of recurrence of their tumors which was happened in 70.9% most of whom debunking was done for them, MSKCC study better outcome regarding recurrence which was occurred 15% of their patients.

Metastasis happens in 27.3% of patients of our studied patients, near the result found in MSKCC study which constituting 25% of their patients.

#### VII. Conclusion

The late presentation of the tumors as reflected by huge size at first presentation making the management a real challenge. Soft tissue sarcoma found to be common in our community and causing big problem in its treatment because of its high rate recurrence and delayed presentation.

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