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Clinical Study of Soft Tissue Sarcoma Cases in A South-Indian Teaching Hospital

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ABSTRACT

Background: The purpose of present study is to know the incidence, clinical presentation and management of soft tissue sarcoma cases and to observe the outcome of the same. Extremity swellings are suspected to be Mesenchymal origin but other areas like Trunk, abdominal wall, Retro peritoneum also may be sites for Soft tissue Sarcomas. The pathological diagnosis by careful tissue study is pivotal. Early recognition, correct histological diagnosis by dedicated Pathologist and early surgical intervention are the most important points regarding the prognosis for the patient which is otherwise very gloomy.

Methods: A prospective study of Soft tissue sarcomas in patients attending Government General Hospital, under Rangaraya Medical College has been undertaken from 2012 to 2014. Forty-six patients with soft-tissue sarcomas have been included in the study. out of which 28 Male and 18 female patients were proved to be having Soft Tissue Sarcomas at various sites. Core-needle biopsy showing Soft-tissue sarcoma of various cell types in both sexes have been included in the study, Exclusion criteria are patients reluctant for follow-up.

Results: 46 patients of various age groups were selected .The peak incidence is between 3rd to 6th decades.The site of Sarcomas have been recorded to be extremities (50%), Trunk (23%), Viscera(13%) and Retroperitoneum(10%), have been in that order the site of Sarcomas.Most common presentation is Asymptomatic swelling followed by recurrent swelling.Wide excision with Post-operative radiotherapy is the most common management protocol that was followed.

Conclusions: Soft tissue sarcomas are a group of lesions that arise from the extra Skeletal connective tissue of the body. They include all the non epithelial extra skeletal tissues with the exception of glia of central nervous system, reticulo endothelial system and the supporting tissue of various parenchyma organs. A high index of suspicion with a Histopathological study is very important. Soft tissue imaging

with MRI is an important pre-operative investigation to determine the extent, integrity and neurovascular involvement. Incision is to be well planned to include the entire swelling. A size more than 5 centimetres, infiltration into surrounding structures, involvement of neurovascular bundle, enlargement of loco-regional lymph nodes are all signs of high risk and poor prognosis. A wide local excision with tumour free margins is very important as local recurrences are to be avoided. Post operative follow up is mandatory and moderate to poorly differentiated sarcomas need Local radiotherapy and chemotherapy.

Key-Words: Core needle Biopsy, MRI study of extremity soft tissue swellings, Recurrent swelling Wide local excision.

DISCUSSION

The soft tissue sarcomas are by definition those which arise from the primitive mesoderm. Soft tissue sarcomas are grouped together because of their similarities in pathologic appearance, clinical presentation and behaviour. Soft tissue sarcomas constitute only about 1% of malignant tumours. There is no definite etiological or predisposing risk factor for the occurrence, most cases of soft tissue sarcoma are thought to be sporadic and their cause is unknown. In rare cases, genetic and environmental factors, prior radiation therapy, viral infections, and immunodeficiency have been associated with the development of sarcoma. In addition, sarcomas have been reported to arise in scar tissue, fracture sites, or sites associated with prior soft tissue trauma. Genetic syndromes such as neurofibromatosis, familial adenomatous polyposis, and the Li-Fraumeni syndrome have all been shown to be associated with the development of soft tissue sarcoma. But, majority of cases are sporadic.

The classification of the soft tissue sarcomas is based on the type of tissue that they contain (Enzinger and Weiss 1983).⁴ and the degree of differentiation. The most commonly found are liposarcoma, leiomyosarcoma, pleomorphic

malignant fibrous histiocytoma (pMFH), GIST, desmoids, myxofibrosarcoma, and synovial sarcoma.

The common subtypes in the extremities are liposarcoma, Malignant fibrous histiocytoma, myxofibrosarcoma, and synovial sarcoma. In retroperitoneal and intra-abdominal sites liposarcoma and leiomyosarcoma are the most common types, whereas in the visceral location, gastrointestinal stromal tumors (GIST), leiomyosarcoma, are found. Desmoid tumors are almost exclusively found in the anterior abdominal wall.

Grossly soft tissue sarcomas grow in an expansive centrifugal fashion leading to compression of peripheral cells into parallel layers and flattening of normal tissues adjacent to the lesion. Blood spread is the most important route of distant spread. This may result from tumor cell emboli originating in reactive neovasculature about the pseudo capsule or from intraoperative manipulation of the neoplasm. Most commonly involved organs are lungs, liver and bones in that order. Virtually metastases are reported in all organs of the body.

STATISTICS

The total number of patients treated at government general hospital, Kakinada for soft tissue sarcoma during 2012 to 2014 is 46. The incidence rate is shown in table1.

Table :1 General Incidence

Total number of cases admitted to govern general hospital, Kakinada.	1,15,222
Total number of patients with malignancies.	2760
Total number of patients with soft tissue sarcoma	46
Percentage of patients of soft tissue sarcoma among total patients	0.039
Percentage of patients of soft tissue sarcoma among the patients with malignancies.	1.66

It is seen that soft tissue sarcoma are slightly more common in males, the ratio being 1.5: 1.

Table: 2 Sex Incidences

Sex	Number of cases	percentage
Male	28	60.8
Female	18	39.2

Figure 1: AGE INCIDENCE

In our study soft tissue sarcoma is predominantly seen in middle and old ages between 21-60 years with peak incidence during 3rd - 5th decades.

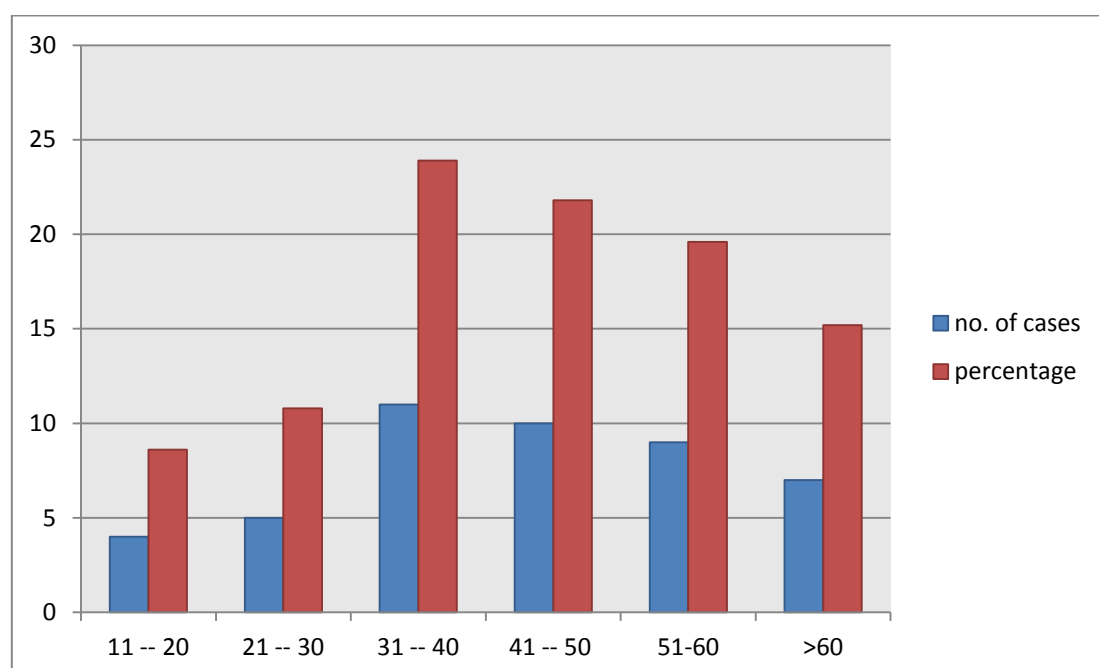
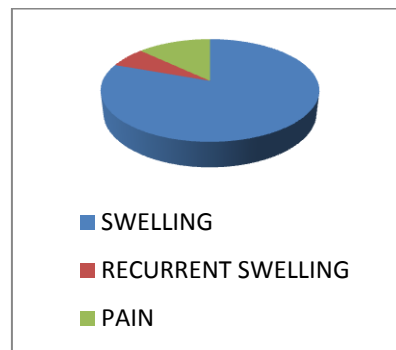


TABLE: 3 SITE OF PRESENTATION

The most common site of soft tissue sarcoma is extremities, followed in decreasing order of frequency trunk, visceral and retro peritoneum.

Site	Number of cases	Percentage
Extremities	24	52.17
Trunk	11	23.92
Retro peritoneum	5	10.86
visceral	6	13.05
Total	46	100

Figure 2: The most common presentation of soft tissue sarcomas is asymptomatic swelling. Pain is the least common symptom.

**Table: 4** Duration of Symptoms:

In our study the duration is between 6 -18 months. Due to the asymptomatic nature of the disease, patients present late to the hospital.

S NO	DURATION	NUMBER OF CASES	PERCENTAGE
1	BELOW 6 MONTHS	10	21.8
2	6-12 MONTHS	16	34.8
3	13-18	8	17.4
4	19 – 24	8	17.4
5	ABOVE 24 MOTHs	4	8.9
	TOTAL	46	100

Figure 3: Metastatic Presentations:

In our study the Soft tissue sarcoma most commonly metastasized to lung followed by soft tissue and liver.

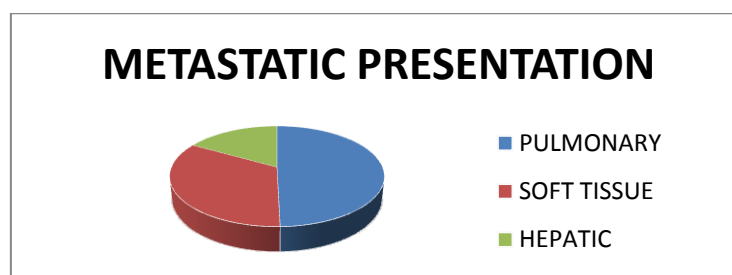


Table:5 Histological Type

The most common histological type is malignant fibrous histocytoma, followed by liposarcoma, leiomyosarcoma, fibro sarcoma, synovial sarcoma.

S NO	TYPE	No of cases	percentage
1	Fibro sarcoma	4	8.69%
2	Liposarcoma	9	19.56%
3	Malignant fibrous histocytoma	12	26%
4	Dermatofibrosarcoma protuberence	1	2.17%
5	Rabdomyosarcoma	2	4.34%
6	Leiomyosarcoma	5	10.86%
7	Neurofibro sarcoma	1	2.17%
8	Synovial sarcoma	3	6.52%
9	GIST	3	6.52%
10	Desmoids tumor	3	6.52%
11	Malignant peripheral nerve sheath tumor	3	6.52%

The common treatment modality is wide excision followed by radiotherapy.

ANALYSIS AND DISCUSSION

This study was conducted in a tertiary care, government medical college from June-2012 to April-2014 after approval from the ethics committee. It is a randomized observational prospective study comprised of 46 patients of soft tissue sarcoma.

Cases were randomized by assessing the patients as they were admitted to surgical wards, government general hospital Kakinada.

The study was conducted with an aim To know the incidence of soft tissue sarcomas, their clinical presentation, management and their outcome. Age and sex incidences are also studied.

INCIDENCE OF SOFT TISSUE SARCOMA

The total incidence in our study is 0.039% of all admissions during 2012-2014 and 1.66% of malignancies during the same period. A study by

SYAM SUNDARA et al 2011³¹ has showed an incidence of 1.7% of all malignancies attending a tertiary care center in India which is truly correlating with the present study.

SEX INCIDENCE

Soft tissue sarcoma occurs in either sex. Most of sarcoma shows no sexual predilection. According to Vaara S et al, Kellers J et al 1998³² 50.94% were males where as 49.06% were females (n= 316). According to study by Shyama Sundara the incidence rate is 59% in males and 41% in females. In our study 60.8% were males where as 39.2% were females. The difference in our study might be due to small sample size.

AGE INCIDENCE

Soft Tissue Sarcoma is more common with increasing age. It occurs in any age group with peak incidence occurring in fifth decade of life.

According to Vaara S et al, Kellers J et al 1998³², the peak incidence is in between 5th and 6th decades. According to study by Shyama Sundara the peak incidence is 4th-5th decade. In our study the peak incidence is 4th-5th decade which is well coinciding with the above study.

SITE SPECIFIC PRESENTATION

Soft tissue sarcoma can occur in any anatomical site. According to Samuel Singer et al³³ 45% of cases presented over extremities, 20% visceral, 15% retroperitoneal, 10% truncal, and 10% at other locations. According to Vaara S et al, Kellers J et al 1998³², 52% of cases presented over extremities,, 19% retroperitoneal, 28% truncal, and 1% at other locations. In our study, 52.17% of cases presented over extremities, 13.05% visceral, 10.86% retroperitoneal, 23.92% truncal, and 4.4% at other locations. All the other studies support the findings in our study that the most common anatomical site affected is extremities.

CLINICAL PRESENTATION

Most of soft tissue sarcomas present as painless swelling in the extremities and truncal region. According to Shyama Sundara et al³¹ commonest mode of presentation is painless swelling which is coinciding our study.

DURATION OF SYMPTOMS

Majority of soft tissue sarcomas present late due to asymptomatic nature of the disease. Most of them are presenting between 6 months to one and half years. According to Shyama Sundara et al³¹ most common period of presentation is 6 months

to one and half years. In our study also the common period of presentation is again 6 months to one and half years.

METASTATIC PRESENTATION

Sarcomas typically spread heamatogenously, rarely through the lymphatics (rhabdomyosarcoma, angiosarcoma, clear cell sarcoma, synovial sarcoma, epithiloid sarcoma). Metastasis occur to the lungs most commonly, hepatic metastasis is seen in primary gastrointestinal and genitourinary sarcomas. According to Shyama Sundara et al³¹, metastasis occurred in 13.1% of cases of which 7.9% occurred in lungs, 2.6% to the liver, 2.6% to soft tissues.

In our study metastasis occurred in 13.1% of cases of which 6.5% occurred in lungs, 2.2% to the liver, 4.4% to soft tissues. Hence commonest place of metastasis in soft tissue sarcoma is to lungs.

HISTOLOGICAL TYPE

There were 50 histological types and subtypes of soft tissue sarcomas. In our study only 11 histological types are diagnosed. It may be due to small sample size. Commonest histological type in our study is malignant fibrous histocytoma, followed in the order of frequency are liposarcoma, leiomyosarcoma, fibrosarcoma, synovial sarcoma, GIST, malignant peripheral nerve sheath tumors and desmiod tumors. The findings are more or less correlate with other studies by Russel et al, Lindburg et al, Jose Costa e al, and Shyama Sundara et al³¹.

(Other histological types diagnosed in different studies are not mentioned).

Our follow up rate may not be long enough. Among all 46 patients, 26 patients have reported, among them 20 patients were diseases free for a period of 18-24 months. 4 patients presented with distant metastasis, 2 patients presented with local recurrence. On the whole the treatment that is tried is broadly along the lines indicated by other authors. On the whole the results correspond to other series. But better patient education for early presentation and better compliance and development of standardized multi modality therapeutic facilities are required.

CONCLUSIONS

Soft tissue sarcoma though rare is interesting tumours. They frequently occur in young and middle age individuals and this together with the challenges they threw to the pathologist in diagnosing them and to the clinician in treating them, generate fascination in them. Clinically often occur in extremities. They usually manifest as a asymptomatic subcutaneous masses. Precise diagnosis can be made only after histopathological examination, during which the pathologist may have to resort two special stains, electron microscopic studies, immune histochemical studies etc. though many can be identified by routine examination. The clinician now has a wide range of investigations like angiography, ultra sonography, CT, MRI etc. with whose help he can precisely map out the extent of the disease pre operatively. Better understanding of the pathological behavior of these tumors,

development of standardized resection, availability of multi modality of the treatment regimens had drastically cut not only mortality rate of these tumors but also the amputation rate of the extremity sarcoma. Availability of the mega voltage radio therapy for local control and effective chemotherapy of disseminated diseases, including 'micro metastases' further improved the once gloomy prognosis. In this study the soft tissue sarcoma occurring in this region is analyzed and their management outlined. These are found to be in line with other series. However our patients usually present late with advanced disease where conservative measures are not useful. Further the follow up rate is poor. So, full standardization of treatment regimens is wanted.

Recommendations are

- Any swelling not confirmed by the fine needle aspiration cytology, true-cut biopsy must be done. So, differentiation of benign and malignant tumors can be done and grade of lesions low or high grade can be evaluated.
 - So, early diagnosis of swelling and early diagnosis by correct methods lead to better prognosis as size of swelling and grade of the tumors for prognostic factors in soft tissue sarcoma.
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