Original Article ISSN (O):2395-2822; ISSN (P):2395-2814

A Study of Clinico-Histopathological and Management of Soft Tissue Sarcoma.

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Received: December 2017
Accepted: December 2017

ABSTRACT

Background: Soft tissue tumors are a diverse and heterogeneous group of tumors. The incidence of benign soft tissue tumors are high relative to malignant tumors. The natural course of soft tissue tumors is un predictable and very aggressive if not diagnosed early. Aim: To study the clinicopathological presentation and modes of treatment of soft tissue sarcomas and to analyse the morbidity and mortality after treatment. **Methods:** In this retrospective 31 histologically confirmed cases of Soft tissue sarcomas cases were included. A detailed gross examination of the soft tissue specimen was performed to record the tumor size, shape, colour, consistency and distance from the deep resected margins. **Results:** The maximum incidence of soft tissue sarcomas was in the 4th, 5th, 6th, & 7th decades and together they made up about 67.74% of cases studied. Malignant fibrous histiocytoma and dermatofibrosarcoma protuberans had the maximum occurrence followed by rhabdomyosarcoma. The commonest site of the primary tumour was the lower extremity followed by the trunk &retroperitoneum and together they account for 58.06% of the primary tumours. Surgery is the main treatment modality and radical clearance of the primary with minimum functional disability should be the aim. **Conclusion:** Even though soft tissue sarcomas are rare, they are life threatening posing a significant diagnostic and therapeutic challenge and must be diagnosed early for better management.

Keywords: Soft tissue sarcoma, pathology, management.

INTRODUCTION

Soft tissues refer to the extra skeletal connective tissues of the body that connect, support and surround other discrete anatomic structures. The soft somatic tissues are ubiquitous and comprise more than 50% of body weight. This portion of the body mass lying between the epidermis and parenchymal organs includes fat, fibrous tissue, smooth and skeletal muscles, tendons, blood vessels, lymphatics and synovial tissue.[1] Soft tissue sarcomas refer to malignant growths that arise from the soft tissues, and they are grouped together because of similarities in pathologic appearance, clinical presentation and behavior. The Greek word Sarcoma means a fleshy growth and virtually all tumors included in the soft tissue sarcomas arise from a common embryonic ancestry, the primitive mesoderm.[2,3] These include tumors of the peripheral nerves, the components of which are derived from neuroectoderm. Although bone and soft tissue sarcomas are uncommon, exposure to a number of different agents has been associated with an increased risk. Moreover advances in cytogenetics and molecular biology have resulted in a significant increase in information related to this disease. Overall, the age-adjusted annual incidence of soft tissue sarcomas ranges from 15 to 35 per 1 million populations. The incidence increases steadily with age and is slightly higher in men than in women. Malignant soft tissue tumors constitute less than 1% of all cancers and occur twice as often as primary bone sarcomas.[4,5] Soft tissue sarcomas are the fourth most common malignancy in children, after hematopoietic neoplasm, neural tumor and Wilms tumor. It accounts for 15% of all childhood cancers.[6] Soft tissue sarcomas may occur anywhere but three fourths are located in the extremities (most common in thigh) and 10% each in the trunk wall and retroperitoneum.[7] The different types of soft tissue tumors have distinct age distributions. Rhabdomyosarcoma is seen more frequently in children and young adults. Synovial sarcoma arises in young adults. Malignant fibrous histiocytoma and liposarcoma generally occur in older adults.[5]

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Aim

To study the clinicopathological presentation and modes of treatment of soft tissue sarcomas and to analyse the morbidity and mortality after treatment.

MATERIALS AND METHODS

A retrospective 31 histologically confirmed cases of Soft tissue sarcomas admitted to the Tirunelveli Medical College Hospital, Tirunelveli were included in the study. Clinical details of all the cases were collected in pretested proforma meeting the objectives of the study. A detailed gross examination of the soft tissue specimen was performed to record the tumor size, shape, colour, consistency and distance from the deep resected margins. Only those cases with tissue diagnosis of soft tissue sarcoma confirmed by a pathologist are included in this study. Sarcomas of the viscera and other organs like bone are not included in this study.

RESULTS

The youngest patient in this series was only 24 years old and the oldest one was 75 years old. The average age of the patients studied was 45 years. The maximum number of cases came under the 60-70 years age group. [Table 1]

Table 1: Age incidence of soft tissue sarcomas

Age	Number of cases	Percentage
<10	3	9.6%
10-20	2	6.45%
20-30	4	12.90%
30-40	5	16.12%
40-50	5	16.12%
50-60	5	16.12%
60-70	6	19.35%
70-80	1	3.22%

Males were more affected than females. Of the 31 cases studied 23 were males i.e. 74.19% and 8 cases were females i.e. 25.81%. Thus the male: female ratio was 3:1.

Table 2 Anatomic Location of the Primary.

Site	Number of cases	Percentage
Head & Neck	6	19.35%
Upper extremity	7	22.58%
Lower extremity	10	32.26%
Trunk & Retroperitoneum	8	25.81%

The lower extremity was the commonest anatomic location of the primary tumour 32.25%, followed by the Trunk &Retroperitoneum 25.81%. [Table 2] The ratio of upper extremity involvement to lower extremity involvement was 3:5 compared to other studies where the ratio is 1:3.

Measurement of the primary tumour at the time of presentation was documented in 23 out of 31 cases. The smallest tumour was 4 cms and the largest 25 cms. The average size was 12 cms. The presenting symptoms included painless enlarging mass, abdominal discomfort, pain and pressure affects like numbness, paraesthesia, muscle weakness, pedal edema etc. This was the most common presenting symptom and it was seen in 28 cases – 90.31%. Of the 8 cases of truncal/retroperitoneal sarcomas 4 cases complained of ill-defined abdominal discomfort aggravated by prolonged standing. Thus 50% of the truncal sarcomas had this symptom and this group forms about 12.9% of total cases. None of these patients had any bowel or bladder complaints. This includes paraesthesia, numbness, muscle weakness due to involvement of nerves, venous engagement, dependant edema due to pressure on the veins and also dyspnea, dysphagia etc. due to compression of the trachea or esophagus. The commonest complaint was paraesthesia followed by pedal edema. Of the cases studied 3 had such complaints – 9.67%.

Table 3: Distribution of symptoms

Symptom	Number of cases	Percentage
Mass	28	90.32%
Pain	23	74.19%
Abdominal discomfort	23	4.19%
Pressure effect	3	9.67%

The histology of the entire specimen was studied in all those cases where the tumour was excised. In some cases, only incision biopsy specimens were available. In a very few cases, only a fine needle aspiration biopsy was performed and the patient was lost to follow up. Of the 31 cases in the present study, the commonest sarcoma was malignant fibrous histiocytoma.

Table 4: Distribution of Histologic subtype.

Histologic subtype	No. of cases	Percentage
MFH	5	16.2%
Liposarcoma	2	6.45%
Fibrosarcoma	3	9.67%
Neurofibrosarcoma	3	9.67%
RMS	4	12.90%
Dermatofibrosarcoma protuberans	5	16.12%
Angiosarcoma	1	3.22%
Others & unclassified	8	25.8%

This includes an USS of the region affected by the primary tumour as well as an USS of the abdomen, to assess the involvement of the liver or other intra – abdominal organs and the presence of ascites. In this study 14 patients underwent USS of the primary tumour. In all these cases, the imageologist could pick up a soft tissue mass but could not definitely say whether the tumour was malignant or not. Most of the time, the tumour had density similar to fat or muscle and delineation was difficult. Hence a CT scan was advised in almost all cases but CT scan was done in only 7 cases. However the imageologist could suggest the possibility of soft tissue sarcoma in 5 out of 7 cases. CT scan of the primary was available in only 7 cases out of the total 31. The other patients could not afford it. CT scan could

provide excellent definition of the extent of the tumour and relationship of muscles. The relationship to blood vessels could be demonstrated by contrast enhancement. Identification of recurrent tumours was difficult due to postoperative scarring. Radiological feature suggestive of malignancy could be obtained in 5 cases. MRI scan was not done in any of the cases.

FNAC was done in all the 31 cases under study. Malignant cells were obtained in 4 cases. However histological subtyping of these tumors was difficult. USS / CT Guided FNAC was done in 15 cases and in 3 malignant cells were obtained (20%). Using a Tru-cut needle, a core of tissue was retrieved in 14 cases, but the diagnosis could be made only in 9 cases (64.3%). This was employed for bigger size lesions and small wedge of tissue was removed 5 cases were subjected to incision biopsy and the diagnosis was got in 4 cases (80%). About 11 cases were diagnosed after excision only. According to other studies FNAC has yielded more than 50% results whereas tru-cut biopsy has been positive in up to 90 of cases. The low value of positivity yielded by this study may be due to problems in sampling and interpretation. Also, the tumour may be less aggressive yielding less of malignant cells. Again, the number of cases included in this study is small. It should be noted that all the cases were not subjected to each of these biopsy techniques but rather only those case which did not yield positive results were further investigated. All these factors could have caused the gross variation in accuracy compared to other studies.

Most of the patients were treated with local wide excision followed by radiation therapy. For soft tissue sarcomas of the extremely, wide excision is always attempted first followed by radiotherapy. If there is local recurrence, then amputation is performed. Of the 17 cases of extremely sarcomas (10 – lower, 7 – upper), included in this study, wide excision was done in 10 cases whereas compartmental resection was done in only 3 cases. Amputation had to be done in 2 cases. Among the truncal sarcomas, wide excision was attempted but was possible only in few cases. In 5 out of 8 cases, surgical excision of most part of the tumour was done while 3 cases were inoperable.

DISCUSSION

Soft tissue is a non epithelial extra skeletal tissue of the body exclusive of reticuloendothelial system, glia and supporting tissue of the various parenchymal organs. It is represented by the voluntary muscles, adipose tissue and fibrous tissue and vessels serving these tissues. The main purpose of this study was to assess the benign and malignant soft tissue tumors with respect to age & gender, site distribution and to compare this with other similar studies. Recognized causes include various chemical and physical factors, exposure to ionizing radiation, inherited or acquired immunological defects. Evaluation of the exact cause is difficult because of the long latent period. Benign soft tissue tumors were more common than malignant tumors in our study, similar to the studies by Enzinger F.M.S.W8. Myhre Jenson et al., Angerwall et al.[9,10]

Studies have reported the maximum age incidence in the forth to sixth decades of life , both males and females were equally affected and many other studies have reported similar views. The present study includes a very meagre representative sample and shows male preponderance.[11-13]

Table 5: Comparison of results with other studies

Studies	Potter et al14	ACS Survey15	Present study
Total cases	307	4550	
Head & Neck	12 (3.9%)	406 (8.9%)	31
Trunk	84	1440	6 (19.35%)
&Retroperitoneum	(27.3%)	(31.6%)	8 (25.8%)
Upper limb	59	594	7 (22.58%)
Lower limb	(19.2%)	(13.1%)	10
	152	2110	(32.25%)
	(49.5%)	(46.3%)	

Malignant fibrous histiocytoma and dermatofibrosarcoma protuberans had the maximum occurrence followed by rhabdomyosarcoma. Of the imageologic investigations, a CT scan proved to be the most useful one. The value of MRI scan could not be assessed because none of the patients could afford one. Biopsy was absolutely indicated to know whether the mass was malignant or not. FNAC done twice by two different persons at two different sites may yield a positive result in few cases. With USS / CT guidance, the positively rate is little more. Trucut needle core biopsy was positive in only 64.3% cases whereas an incisional biopsy yielded 80% positive results. Proper orientation of the biopsy incision is very important.[16,17]

Surgery is the main treatment modality and radical clearance of the primary with minimum functional disability should be the aim. Surgical excision followed by post-operative radiotherapy is the standard management of high grade limb sarcomas although occasionally amputation remains the only option. Pre-operative treatment with chemotherapy or radiotherapy should be considered for patients with borderline resectable tumours. Isolated limb perfusion may permit limb salvage in some cases where amputation is the only other option. Adjuvant chemotherapy is not routinely recommended but may be considered in certain specific situations. Regular follow up is recommended to assess local control and the development of metastatic disease.[18]

CONCLUSION

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How to cite this article: Sahayam JS, Sulaiman J, Senthurpandian, Anandan H. A Study of Clinico-Histopathological and Management of Soft Tissue Sarcoma. Ann. Int. Med. Den. Res. 2018; 4(1):SG47-SG50.

Source of Support: Nil, **Conflict of Interest:** None declared