A Study on Soft Tissue Sarcomas (STS)

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Abstract
Introduction: STS constitute a significant challenge to the surgeon. They are rare tumors accounting for 1% of all malignancies. There are more than 20 histologic varieties. They are aggressive neoplasms and have the propensity for early metastatic spread.
Aim: To study age and sex incidence, clinical presentation, various histology, diagnostic modalities, various treatment modalities, post-operative complications.
Materials and method: Biopsy proven 40 cases of STS.
Results: Rare tumor in this region. Common in extremities. Peak incidence is 50 years and above. 75% of patients had presented in advanced stage of disease. Surgery is the prime modality of treatment.
Conclusion: STS remains an ideal model for investigation of strategies designed to improve long-term outcome. Multiple prognostic factors are being defined and the disease process is varied enough to provide a fertile field for treatment strategies.
Keywords: Histology, Treatment modalities, Follow-up

Introduction
Soft tissue neoplasms are defined as a heterogeneous group of tumors arising from PRIMITIVE MULTIPOTENT MESENCHYMAL cells, which undergo differentiation along one or more lines as the tumor evolves. There are wide variety of histopathological types each with varying biological behavior and a uniform character for hematogenous spread. Soft tissue tumors often pose a diagnostic challenge to the pathologist, since many of the benign tumors are unencapsulated and show infiltrating margins, thus making it difficult to differentiate benign from malignant tumors. Although benign soft tissue neoplasms represent the commonest neoplasm in human beings, their malignant counterparts accounts for only 0.8%-1.0% of all malignant tumor adults, and 6-8% of those in children.
A variety of special procedures such as conventional special stains, immunohistochemistry, solid tumor cytogenetics, electron microscopy, quantitative image analysis, and flow cytometry are now available to improve the diagnostic accuracy but none of them are much helpful to differentiate benign tumors from malignant neoplasms, till date. These tumors constitute a fertile field for clinicopathologists and pose a significant challenge to the treating surgeon.
Aim of Study
During the two years period from 2015-2017, cases of soft tissue tumors were followed and those which were histopathologically proved to be sarcomas were included in this study. Various surgical procedures like Excision & biopsy, Wide excision and Radical procedures were done. Soft tissue sarcomas present as a challenge to the surgeon. Hence the study for the following reasons:

- To study
- The age and sex incidence of soft tissue sarcomas.
- The clinical presentations (signs and symptoms)
- Diagnostic modalities used in evaluation of soft tissue sarcomas
- Various types of treatment offered
- Post-operative complications
- The various histology of soft tissue sarcomas

Materials and Methods
From Dec 2015 to Dec 2017 for a period of over 2 years, 43 cases were admitted in TMCH. All the 43 cases were studied in detail with clinical examination, Histopathological examination and Radiological investigations.

Observation
Sex

<table>
<thead>
<tr>
<th>Sex</th>
<th>No of Patients</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male</td>
<td>31</td>
<td>70%</td>
</tr>
<tr>
<td>Female</td>
<td>12</td>
<td>30%</td>
</tr>
</tbody>
</table>

Age
The peak incidence of these tumors was in the 5th decade of life.
The youngest patient in our study was a 2yr old male child with rhabdomyosarcoma whereas the oldest was a 75yr old male with fibrosarcoma.

<table>
<thead>
<tr>
<th>Age Group</th>
<th>No of Patients</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt; 10</td>
<td>3</td>
<td>7%</td>
</tr>
<tr>
<td>10 - 20</td>
<td>4</td>
<td>10%</td>
</tr>
<tr>
<td>20 - 30</td>
<td>5</td>
<td>12%</td>
</tr>
<tr>
<td>30 - 40</td>
<td>9</td>
<td>20%</td>
</tr>
<tr>
<td>40 - 50</td>
<td>9</td>
<td>20%</td>
</tr>
<tr>
<td>50 And Above</td>
<td>13</td>
<td>30%</td>
</tr>
</tbody>
</table>

Site Distribution
The tumor site was predominantly in the extremities [80%]

<table>
<thead>
<tr>
<th>Anatomical Site</th>
<th>No of Patients</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lower Limb</td>
<td>22</td>
<td>54%</td>
</tr>
<tr>
<td>Upper Limb</td>
<td>12</td>
<td>26%</td>
</tr>
<tr>
<td>Abdomen</td>
<td>8</td>
<td>18%</td>
</tr>
<tr>
<td>Chest Wall</td>
<td>1</td>
<td>2%</td>
</tr>
</tbody>
</table>

Histopathology Distribution
Out of 43 cases of soft tissue sarcomas, the most common were malignant fibrous histiocyto ma and malignant neural tumours.

<table>
<thead>
<tr>
<th>Histopathological Type</th>
<th>No of Patients</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>MfH</td>
<td>11</td>
<td>26%</td>
</tr>
<tr>
<td>Liposarcoma</td>
<td>7</td>
<td>16%</td>
</tr>
<tr>
<td>Ewing's Sarcoma</td>
<td>6</td>
<td>13%</td>
</tr>
<tr>
<td>Nerve Sheath Tumors</td>
<td>3</td>
<td>7%</td>
</tr>
<tr>
<td>Rhabdomyosarcoma</td>
<td>5</td>
<td>12%</td>
</tr>
<tr>
<td>Fibrosarcoma</td>
<td>9</td>
<td>20%</td>
</tr>
<tr>
<td>Leiomyosarcoma</td>
<td>1</td>
<td>3%</td>
</tr>
<tr>
<td>Synovial Sarcoma</td>
<td>1</td>
<td>3%</td>
</tr>
</tbody>
</table>

The increased incidence of malignant fibrous histiocytoma with a relative fall in fibrosarcoma is probably due to the fact that were previously thought to be fibrosarcomas have now been diagnosed as malignant fibrous histiocytoma.

Clinical Presentation
The symptoms of prime importance for which the patients reported were painless swelling in 80% patients and painful swelling in 20% patients.

Treatment

<table>
<thead>
<tr>
<th>Nature of Treatment</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Refused treatment</td>
<td>24%</td>
</tr>
<tr>
<td>Biopsy followed by RT &amp; CT</td>
<td>30%</td>
</tr>
<tr>
<td>Debulking</td>
<td>9%</td>
</tr>
<tr>
<td>Wide excision with skin grafting</td>
<td>11%</td>
</tr>
<tr>
<td>Wide excision and CT &amp; RT</td>
<td>26%</td>
</tr>
</tbody>
</table>

Complications
Out of 8 abdominal cases, we did excision for 4 cases, debulking for 2 cases. Of these 3 patients came with recurrence and reported with symptoms of abdominal pain, mass and lower limb edema.1 patient presented with features of subacute intestinal obstruction.1 patient with nerve sheath
tumor of popliteal region was operated and after 6 months he presented with paraplegia due to cord compression from metastasis. In extremity sarcomas operated, 4 cases presented with recurrences.

The observation derived from this study are:

- It is a rare tumor in this region with a marginal increase in incidence compared to the international literature.
- Extremity tumors account for the majority of sarcomas
- Peak incidence is between 50 years and above
- Male to female ratio is 2.3:1
- No predisposing factor is correlated with.
- Possesses no problem for physical examination and diagnosis, yet they are frequently diagnosed very late.
- 75% of patients had presented in advanced stage of disease. A third of patients had developed distant metastases.
- Malignant fibrous histiocytoma and fibrosarcoma accounted for the highest number of histologic varieties.
- FNAC is an effective procedure in the diagnosis of soft tissue tumors.
- Surgery is the prime modality of treatment.
- Post excisional defects were reconstructed with myocutaneous flaps, thereby achieving good cosmetic and functional results.
- Multimodality approach with preoperative radiotherapy and post-operative chemotherapy is the standard in the management of sarcomas.

Discussion
For many years, the treatment of soft tissue sarcomas has continued to pose significant challenges. It is due to relative rarity of these tumors, contributed to by lack of proper understanding of their biological behavior and a significant delay in the diagnosis. The past decade has seen a dramatic increase in the adoption of multiple modality therapy for these tumors with a significant reduction in the number of amputations performed for extremity lesions. An effort to decrease local recurrence of extremity sarcomas has been successful. Multimodality approach offers the potential for improved local control with less radical operations.

It is evident from this prospective study that most patients presented very late in the course of illness. Significant delay in diagnosis only contributes to the aggressive behavior of these tumors. About 30-40% of tumors were treated in the form of incision and drainage, excision and like procedures. Out of 43 patients with soft tissue sarcoma 11 patients were diagnosed by biopsy to have malignant fibrous histiocytoma.

Conclusion
Soft tissue sarcomas constitute a significant challenge to the surgeon. They are rare tumours accounting for 1% of all malignancies, and there are more than 20 histologic varieties each with varying biologic behavior and a propensity for early metastatic spread. They are aggressive neoplasms. They are very innocent looking at the initial stages of the disease and relatively symptomless. This probably makes the patients very complacent towards the existence of the tumour and by the time they present, it is already in an advanced stage. To compound to the problem of late diagnosis, the problem of indifferent treatment in the peripheral set up. Multimodality approach is the current standard for the treatment of sarcomas. With majority of tumours presenting in the extremities, the aim of surgery in most situations is limb salvage. Radiotherapy has been convincingly proved to be an effective tool in the treatment armamentarium for soft tissue sarcomas. Chemotherapy has been reserved for metastatic disease although a few selected trials have established the usefulness of pre-operative chemotherapy in multimodal approach. Doxorubicin and Ifosfamide are the two drugs with promise in the field of managing advanced sarcomas.

Molecular biology has promise for the future. The focus on genetic alterations that predispose to
tumour development and progression, has thrown more light on the etiology of these tumours. The relevance of such genetic abnormalities in diagnosis is obvious, but they have yet to be exploited in terms of clinical management. Soft tissue sarcoma remains an ideal model for the investigation of strategies designed to prove long term outcome. Advances in knowledge of etiological predisposition continue to be de. Multiple prognostic factors are being defined and the disease process is varied enough to vide a fertile field for treatment strategies.

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