RECENT TRENDS IN THE MANAGEMENT OF SOFT TISSUE SARCOMAS

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ABSTRACT

BACKGROUND

Soft tissue sarcomas are a heterogeneous group of tumours arising from primitive multipotent mesenchymal cells undergoing differentiation into one or more cell lines. They constitute 1% of human cancers.^[1] Most cases of soft tissue sarcoma are sporadic and the cause is unknown. Soft tissue sarcomas are one of the most common radiation associated tumours^[2] and they develop from any site and have more than 50 histological types and subtypes.

The aim of this study is to provide an overview of its distribution and to emphasise the recent trends in the management of soft tissue sarcoma with considerable change towards limb and organ saving attitude.^[3]

MATERIALS AND METHODS

This study was conducted in KAPV Government Medical College and Mahatma Gandhi Memorial Government Hospital, Trichy between January 2014 and December 2016 over a period of 2 years. This study included 50 cases of soft tissue sarcoma, who were all inpatients in KAPV Government Medical College and Mahatma Gandhi Memorial Hospital, Trichy. Patients were descriptively studied for the distribution of soft tissue sarcoma and the mode of management adopted towards them.

RESULTS

The study shows the gender distribution of soft tissue sarcoma among 50 cases with 33 males (66%) and 17 females (34%). Among the 50 cases the incidence is higher in 51 - 60 years of age 10 cases (20%), incidence is equal in 21 to 30 years and 31 - 40 years' age group, both having 9 cases with 18% incidence. The lowest incidence is in above 70 years' age group, 1 case only (2%). Lower limb is the site mostly affected with 20 cases (40%). Upper limb and abdomen show equal numbers with 11 cases each (22%) and lowest incidence is in gluteal site soft tissue sarcoma 1 case (2%). Among the 14 types of soft tissue sarcoma studied, fibrosarcoma shows higher incidence with 17 cases (34%) followed by malignant fibrous histiocytoma (MFH) 8 cases (16%), chondrosarcoma, malignant haemangioendothelioma, GIST and spindle cell sarcoma have 1 case each (2%). 27 cases were found to have stage 3 disease (54%) and stage 4 disease among 3 cases (6%). Regarding the treatment given, surgery alone was done in 3 cases (6%). Surgery with chemotherapy and radiotherapy were done on 39 cases (78%). Among the surgical procedures, wide local excision was done in 34 cases (68%) and radical hysterectomy was done in 1 case (2%). The recent trend in management of soft tissue sarcoma is towards organ saving attitude with the advent of multimodality approach. Surgery is the primary modality. Patients mostly underwent wide local excision and compartmental excision followed by postoperative radiotherapy and chemotherapy. Postoperative radiotherapy is given with external beam radiotherapy. Adjuvant chemotherapy was given with Vincristine, Adriamycin and Cyclophosphamide (VAC regimen). Out of 50 cases, 3 cases had recurrence in 2 years follow-up period.

CONCLUSION

Soft tissue sarcomas are rare tumours of mesenchymal origin. Multimodality approach in the management is the recent trend aimed at organ salvage. Surgery is the mainstay of management. Wide local excision and compartmental excision with postoperative adjuvant radiotherapy and chemotherapy gives good survival rates. Out of 50 cases only 3 cases had recurrence, thus showing the effectiveness of multimodality approach with surgery, radiotherapy and chemotherapy.

KEYWORDS

Soft Tissue Sarcoma, Wide Local Excision, Compartmental Excision, Amputation, Radiotherapy, Chemotherapy.

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BACKGROUND

Soft tissue sarcoma are defined as heterogeneous group of tumours arising from primitive multipotent mesenchymal

cells undergoing differentiation into one or more lines. There are no clear predisposing factors. There are a wide variety of histological types according to the cell line of maturation with varying biological behaviour. Generally, these tumours do not cross anatomical barriers like bone, skin and fascia. Commonly spread by blood stream. They clinically present as painless lump. Diagnosis is established by tissue biopsy.^[4] These tumours were approached aggressively with radical procedures like amputation twenty years back. The management of these tumours have undergone a radical change in the approach in the recent past due to better understanding of the tumour biology, advances in various

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fields of surgery like microvascular technique for flap coverage, availability of excellent chemotherapeutic agents and advances in radiotherapy.^[5] With the availability of multimodality treatment, patients are benefitted with good quality of life with useful functioning limb or organ without compromise on the survival.^[5] The aim of the study is to give an overview of the distribution of soft tissue sarcomas and to emphasise the recent trend in the management of soft tissue sarcoma with considerable change towards limb and organ saving attitude.

MATERIALS AND METHODS

Study Place

KAPV Government Medical College and Mahatma Gandhi Memorial Hospital, Trichy.

Duration of Study

January 2014 to December 2016.

Study Design

Descriptive Study.

Ethical Committee

Institutional Ethical Committee, KAPV Government Medical College, approved in January 2014.

Inclusion Criteria

Patients aged above 12 years. All sex group patients who were confirmed with soft tissue sarcoma.

Exclusion Criteria

Patients aged less than 12 years. Patients not confirmed with soft tissue sarcoma through Tru-cut biopsy.

Data Collection and Methods

Patients confirmed with soft tissue sarcoma in Mahatma Gandhi Memorial Government Hospital, Trichy, were included in the study. Details of patients with soft tissue sarcoma and their histological types were identified. Among the many types of soft tissue sarcoma, fourteen histological types that were available were studied descriptively. I have selected 50 cases of soft tissue sarcoma with 14 histological types who were admitted in Mahatma Gandhi Memorial Government Hospital, Trichy, between January 2014 and December 2016. They were managed with- 1. Surgery alone, 2. Surgery with radiotherapy and 3. Surgery with radiotherapy and chemotherapy. Surgical methods employed were Wide Local Excision, Compartmental Excision, Amputation, Resection anastomosis, Radical hysterectomy and Lymph node dissection. Postoperative histopathological reports were obtained and adjuvant treatment with radiotherapy or with chemotherapy and radiotherapy were advised. Patients were followed up for a period of two years. In the follow-up period of the 50 cases, 3 patients had recurrence. Among the three cases, one case did not turn up for further follow-up.

Statistical Analysis

Descriptive statistics such as frequency and proportion were calculated.

RESULTS

50 patients with soft tissue tumour appearing malignant were studied.

Gender	No. of Patients	Percentage
Male	33	66%
Female	17	34%
Total	50	100%

Table 1. Gender Distribution

Age	Total	Percentage
13-20	8	16%
21-30	9	18%
31-40	9	18%
41-50	8	16%
51-60	10	20%
61-70	5	10%
Above 70	1	2%
Total	50	100

Table 2. Distribution according to Age

Site	Total	Percentage
Lower Limb		
Thigh	16	32%
Leg	3	6%
Foot	1	2%
Upper Limb		
Hand	4	8%
Forearm	3	6%
Arm	4	8%
Abdomen		
Abdominal wall	4	8%
Intraperitoneal	3	6%
Retroperitoneal	4	8%
Gluteal Region	1	2%
Chest Wall	3	6%
Hand and Neck	4	8%

Table 3. Distribution according to Site

HPE	Male	Female	Total	%
Fibrosarcoma	10	7	17	34%
MFH	5	3	8	16%
Liposarcoma	2	3	5	10%
Dermatofibrosarcoma protuberans	3	-	3	6%
Synovial sarcoma	2	1	3	6%
Leiomyosarcoma	1	1	2	4%
Haemangiopericytoma	-	2	2	4%
Malignant schwannoma	1	1	2	4%
MPSNT	2	-	2	4%
Desmoid tumour	1	1	2	4%
Chondrosarcoma	1	-	1	2%
Malignant haemangioendothelioma	-	1	1	2%
GIST	1	-	1	2%
Spindle cell sarcoma	1	-	1	2%

Table 4. Distribution according to the Histopathology Subtype^{[7],[8]}

Stage	No. of Patients	Percentage
Stage-1	4	8%
Stage-2	16	32%
Stage-3	27	54%
Stage-4	3	6%

Table 5. Distribution according to the Stage of Disease

Modality of Treatment	No. of Patients	Percentage
Surgery alone	3	6%
Surgery + RT	8	16%
Surgery + RT + ChT	39	78%
Total	50	100%

Table 6. Modality of Treatment

Detail of Surgery	No. of Patients	Percentage
Wide Local Excision	34	68%
Compartmental Excision	4	8%
Amputation	6	12%
Resection Anastomosis	20	4%
Radical Hysterectomy	1	2%
Lymph Node Dissection	3	6%

Table 7. Distribution of Patients among various Surgical Procedures

DISCUSSION

As per literature, benign soft tissue tumours outnumber malignant ones which are rare tumours.^[6] Incidence is 1% - 2%. Soft tissue tumours present clinically as painless mass growing over a period of months. Incidence in males is more when compared to females. Extremity sarcomas are common.^[7] Fibrosarcomas and malignant fibrous histiocytomas are more common than other histological variety. Patients usually present in stage II or III disease. Metastatic sarcomas do occur in lesser number. Patients were subjected to core needle biopsy for confirmation of diagnosis and grading of tumour. FNAC is useful in recurrent soft tissue sarcomas. These tumours were treated radically with amputation of limb 20 years back. The recent trend in the management of soft tissue sarcomas is towards limb saving, organ saving attitude with the advent of multimodality approach. Surgery is the primary modality of treatment. Post excisional defects were reconstructed with skin graft and myocutaneous flaps with the availability of microvascular technique with good cosmetic results in functioning limb. Patients usually underwent wide local excision and compartmental excision followed by postoperative radiotherapy to the tumour bed and chemotherapy.^[8] The results are comparable with that of amputation practiced previously. Postoperative radiotherapy was given as external radiotherapy of 50 grey given as 200 cgy per day for 5 days per week for 5 weeks. Adjuvant chemotherapy was given with combination chemotherapy using VAC regimen (Vincristine, Adriamycin and Cyclophosphamide).^[9] Doxorubicin and Ifosfamide was also used in some patients with comparable results. Neoadjuvant chemotherapy or radiotherapy to downstage the disease is not practiced for soft tissue tumours in contrast to bone sarcomas. Isolated limb perfusion technique was not undertaken due to non-availability of technology in our institution. Out of 50 patients only 3 patients had recurrence in the 2-year follow-up period, possibly due to aggressiveness of high-grade tumours. Patients who presented with metastatic tumours were

treated with palliative chemotherapy with palliative resection of primary tumour including amputation. Of the three, one was lost for followup and one died of pulmonary metastasis.

CONCLUSION

Soft tissue sarcomas are rare tumours of mesenchymal origin, presents as painless mass. Extremity sarcomas are common. Diagnosed by core needle or incisional biopsy. Multimodality approach in the management is the recent trend, aiming at limb or organ salvage. Surgery is the mainstay of management. Wide local excision and compartmental excision with postop adjuvant radiotherapy and combination chemotherapy results in good survival rates with adequate limb salvage. Targeted therapy as for GIST with tyrosine kinase inhibitor Imatinib gives promising results. Gene therapy, monoclonal antibodies and angiogenesis inhibitors may play a crucial role in the management of soft tissue sarcoma aiming better survival rate or even cure.

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