Journal of Rawalpindi Medical College (JRMC); 2018;22(1): 58-62

Original Article

Surgical Management of Soft Tissue Sarcoma

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Abstract

Background: To study the management pattern of soft tissue sarcomas in a tertiary care setting

Methods: In this descriptive study patients who were operated for soft tissue masses were included. Intra-operative findings, procedure details and postoperative orders were recorded. Postoperative chemo-radiotherapy records were reviewed and recorded. Depth of the tumour was grouped as deeper or superficial than 5 cm. The follow up records were accessed from the outpatient department and any surgical complications were recorded up to three years.Staging was done using clinical and radiological criteria taking into account the histological grade,tumour size , depth, local lymph node invasion and metastasis.Surgical procedure for removal of STS are wide local excision (WLE), intralesional excision (IE) or tumor debulking, marginal excision (ME) and radical excision (RE).

Results: Sixty eight patients with mean age of 43.0 ± 17.258 SD were diagnosed as cases of soft tissue sarcomas. Male to female ratio was 3.25:1.The most common histopathological variety was malignant fibrous histiocytoma (35.3%) , followed by Rhabdomyosarcoma(30%). Most common involved site was lower limbs (35.3%).Wide local excision was performed in majority (82.4%) . Most common postop complication was wound infection (10.3%)

Conclusion: Clinicians must be agile about the nature of these tumours and their referral to a specialist surgeon for further management. Prompt diagnosis, accurate investigations and early intervention will benefit the patients and help us understand this disease entity.

Key Words: Soft tissue sarcoma, Surgical oncology, Multidisciplinary management

Introduction

Soft tissue sarcomas are rare tumours with a prevalence of only 1% among all the diagnosed malignancies. The low prevalence of these tumours has predictably resulted into less experience of primary healthcare physicians and surgeons about their management with consequent errors in diagnosis, treatment delays and reporting. Advances in molecular biology, oncogenetics, techniques of modern imaging, histopathological techniques, immunochemistry, good surgical care and chemo-radiotherapy have led us to better understand these rare mesenchymal tumours, commonly known as soft tissue sarcomas (STS).[1-3]

Almost 40 years ago, surgical resection was deemed as the only therapeutic intervention for STS which in high grade tumours resulted in poorer prognosis and shorter survival rates. Good investigative techniques, higher clinical vigilance with pre- and post-operative radiation and chemotherapy has helped increase long-term survival and reduced morbidity in patients of these rare tumours.[4-6]

Although STS comprise almost 1% of the total cancer diagnoses, due to its insidious nature and apparently non-alarming initial clinical appearance it causes significant morbidity and mortality, especially in the young age groups.[7] A majority of patients present late due to initial misdiagnosis by the primary care physician or locally available surgeon who happen to least suspect a soft tissue mass as being malignant. The diverse variety of histopathologic nature of these tumours can be reflected by the fact that there are more than 40 different subtypes of adult STS and similar number of STS subtypes in paediatric age groups. Survival estimates still depends upon the individual histopathological diagnosis and ranges from 92% to 19%.[8,9]

A multidisciplinary team approach especially with dedicated STS teams is the best method which can benefit patients in terms of lower morbidity and improved survival.[10-12] Similarly, intraoperative radiotherapy, pre-operative chemo-radiotherapy and the use of modern imaging techniques in order to monitor long term outcome and relapse rate are the tools which could be the key to success.[13-15]

Methods

After the Hospital Ethical Committee's approval, the study was conducted at the General Surgery Unit II of

Pakistan Institute of Medical Sciences Islamabad. In this descriptive study all patients, between the ages of 14 years to 85 years of age, who were operated for soft tissue masses were included. Preoperative investigative studies such as CT, MRI and fine needle aspiration cytology reports were recorded. Intra-operative findings, procedure details and postoperative orders were recorded. Postoperative chemo-radiotherapy records were reviewed and recorded. Size and depth of the tumour and structures invaded or compressed by the tumour was assessed using the radiology. Size of the tumour was grouped as smaller or larger than 5 cm. Depth of the tumour was grouped as deeper or superficial than 5 cm. Patients were consented after full discussion of the pros and cons of the procedure and the need for postoperative radio-oncology and general surgery follow up were preoperatively discussed with them. Those patients who were clearly labelled as having malignant tumours on FNA reports were also referred to the oncologist for receiving preoperative chemo-radiotherapy. The follow up records were accessed from the outpatient department and any surgical complications were recorded up to three years.Staging was done using clinical and radiological criteria taking into account the histological grade (G1: well differentiated, G2: moderately well differentiated and G3 & G4: poorly differentiated, undifferentiated), tumour size (smaller or larger than 5 cm in greatest dimension), depth (deeper more or less than 5 cm), local lymph node invasion (N0: no lymph nodes, N1: lymph nodes positive) and metastasis (M0: no metastasis, M1: metastasis positive).[16]Surgical procedure for removal of STS are wide local excision (WLE), intralesional excision (IE) or tumor debulking, marginal excision (ME) and radical excision (RE). The need for a particular surgical procedure was determined according to clinical findings and radiological evaluation.Paediatric patients, recurrent cases, non-operable cases and tumours with clearly benign clinical and histological findings were excluded from the study.

Results

Out of sixty eight patients 76.5% were male and 23.5% were female. Mean age was 43 years ± 17.358 SD (range = 14-85). (Table 1). The most common diagnosis was of malignant fibrous histiocytoma (MFH) (35.3%), was followed by rhabdomyosarcoma (17.6%) and liposarcoma (3.2%). (Table 2).Lower extremity(35.35), especially gluteal and thigh are, was the most frequent site involved (Table 3).Forty nine (72.1%) cases were larger than 5 cm while 27.9% cases were smaller tumours (size < 5 cm). Thirty six (52.9%) tumours were classified as superficial (tumour depth < 5 cm). Forty (58.8%) of tumours had radiological evidence of invasion of the local structures while 8 (11.8%) cases were encountered with metastasis to other regions of the body (most commonly lungs and liver). Twenty (29.4%) cases were well differentiated tumours (G1) (Table 4). Preoperative oncological treatment was given only in 5 (7.4%) cases while 35 (51.5%) patients received postoperative oncology treatment. Resection margins were positive in 14 (20.6%) specimens after histopathological review.Wide local excision was performed in majority (82.4%) (Table 5). Postoperative complications occurred in 39.7% . (Table 6)

Table 1: Age groups distribution

Age Group	No	Percentage
1 (14-25 years)	14	20.6
2 (26-40 years)	17	25.0
3 (41-55 years)	21	30.9
4 (56-70 years)	13	19.1
5 (71-85 years)	3	4.4
Mean Age: 43 ± 17.358 SD		

Table 2: Tumour subtypes and their frequencies

Tumour Subtype	Number	Percentage
MalignantFibrousHistiocytoma	24	35.3
Rhabdomyosarcoma	12	17.6
Liposarcoma	9	13.2
Fibrosarcoma	8	11.8
Synovial Sarcoma	4	5.9
Ewing's Sarcoma	3	4.4
Leiyomyosarcoma	3	4.4
Schwannoma	2	2.9
Angiosarcoma	1	1.5
Dermatofibroma	1	1.5
Neurofibroma	1	1.5

Table 3: Body sites and tumour frequencies

Body Site	Number	Percentage
Lower Extremity	24	35.3
Upper Extremity	14	20.6
Intra-abdominal	13	19.1
Trunk	10	14.7
Retroperitoneal	4	5.9
Head and Neck	2	2.9
Intrathoracic	1	1.5

Table 4: Tumour staging and their occurrence

Tumour Stage	Frequency	Percentage
Stage 1a	14	20.6
Stage 1b	19	27.9
Stage 2a	7	10.3
Stage 2b	7	10.3
Stage 2c	12	17.6
Stage 3	6	8.8
Stage 4	3	4.4

Table 5 : Surgical procedures in the study

Surgical procedure	No(%)
Wide local excision	56 (82.4)
Radical excision	7(10.3)
Marginal resection	3 (4.4)
Intralesional resection	2 (2.9)

Table 6: Postoperative complications

Complications	Frequency	Percentage
Wound infection	7	10.3%
Subcutaneous Seroma	6	8.8%
Bleed / Haematoma	3	4.4%
Wound Dehiscence	3	4.4%
Flap necrosis	3	4.4%
Local Muscle Paralysis	2	2.9%
Tumour Bed Abscess	1	1.5%
Pleural Effusion	1	1.5%
Postop Pneumothorax	1	1.5%

Discussion

Soft tissue sarcomas are heterogeneous tumours of mesenchymal multipotent stem cells origin with insidious onset and no specific set of symptoms or signs. Most of the tumours present with localised swellings, pressure effects or symptoms due to invasion of body organs or structures. In most cases before the patient reaches the tertiary care clinics, opinion from locally available primary physicians has already been sought. In such situations, diagnostic delay, misdiagnosis and consequent mistreatment always lead to delayed presentation or presenting with complications of the disease.[17-19]

Modern imaging techniques, histopathological advancements and novel techniques of the immunohistochemistry and oncogenetics have enabled the modern oncological research to identify more than 50 different types of adult STS.[20-22]In Pakistan however, the newly available diagnostic techniques, histopathology services and more importantly expertise in the field of soft tissue neoplasm management is a scarcity.[23] Most reports have cited the data available from tertiary care centres and epidemiological studies.[24] The need of the moment is to enhance the surgical research on planned techniques of excision, repair and postoperative care of this subset of patients.[25]

As emphasized by Umar HM et al planned excision is the only approach which appear feasible for controlling local recurrence and disease spread.[24] Although several authors have shown that local recurrence is not significantly associated with unplanned excision,[26] others have demonstrated the efficacy of planned excision of a STS in order to reduce the risk of local recurrence.[18,24,27] Though we did not include data about local recurrence of the primary tumours in our study, surgical outcome studies have included this aspect as viable to study overall survival as well as morbidity free interval and is the standard of care in all soft tissue sarcoma care centres.[24,27,28]

Another important aspect of proper referral for sarcoma patients is that surgical excision increasingly becomes difficult, intraoperative complications, wound complications and the risk of tumour seeding in neighbouring tissues become high.[24,27] Obtaining a negative surgical margin is another factor which becomes difficult in recurrent disease and in those cases with very large tumours, both related to delayed diagnosis or improper treatment. Liu C et al has described a surgical margin of at least 10 mm for good postoperative outcome and good control of local recurrence.[29] The goal of our surgical planning in these cases was to obtain a negative margin of at least 2 cm. However, the 8 cases of grade 3 & 4 disease and 6 from the stage 2c patients were found to have a positive margin on histopathology review of the specimen. Liposarcoma and synovial sarcoma were the most common tumours which affected the surgical margins clearance.

Opinion, attitude towards new practices and expertise of the sarcoma treating physician, surgeon or oncologist is of paramount importance which determines the efficacy of the pre- or post-operative treatment. Treatment preferences among various specialty physicians regarding neoadjuvant radiotherapy and chemotherapy were highly variable and was influenced by specialty and clinical experience of the physician.[28]

In our study majority of patients were male (76.5%) with mean age of 43.0 years ± 17.358, while malignant fibrous histiocytoma was the predominant subtype of tumour encountered (35.3%). Similarly, the most commonly encountered body site were the extremities (55.9%) followed by intra-abdominal tumours (25.0%). Some of the findings about age distribution are

concurrent with the studies of Bhurgri Y et al and Qadir I et al where average age was 40.5 years and 41.8 years respectively with a majority of male patient.[23,30] In the study by Bhurgri Y et al, Rhabdomyosarcoma accounted for 22.2% tumours and the most abundant one.[30] However, in the study by Qadir I et al, the predominant tumour type was synovial sarcoma and leiyomyosarcoma (36.1%) as compared to our study (MFH: 35.3%; Rhabdomyosarcoma: 17.6%; Synovial sarcoma: 5.9%).[23] Despite the scarcity of resources for widespread multimodality treatment facilities in Pakistan, Qadir I et al have shown that our local recurrence and overall survival rates are comparable to the developed countries.[23] This is a quite encouraging finding for our cancer research community and broadly for our healthcare community.

The most common method of preoperative tissue diagnosis in our cohort of STS patients was the utilisation of fine needle aspiration coupled with contrast enhanced CT/MRI. Specific cytological and radiological markers of malignancy are important in this regard as discussed by Ahmed Z et al in elucidating the importance of histopathology and immune-histochemistry.[31] As is pointed by Chintamani, the most important factor in diagnostic difficulty of STSs is their histologic heterogeneity, which has also hampered the treatment advancement for years.[32]Data regarding follow-up once the patient left for postoperative oncology treatment, recurrence and disease free or overall survival rates are necessary for evaluating the patients suffering from STS and the success or failure of a particular modality of treatment. These limitations can be avoided by designing proper long-term prospective studies with survival and recurrence assessment. More important though, is the formation of multimodality treatment boards in order to treat this subset of patients.

Conclusion

1.In the management of soft tissue sarcomas increased vigilance, regarding preoperative workup, and proper referral are of paramount importance.

2.Preoperative oncological treatment and planned excision needs emphasis. Multimodality surgical and oncological support in tertiary care centres can be translated into increased survival and improved quality of life for these patients.

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