Surgical Outcome of Fungating Sarcomas.

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ABSTRACT

Objective: To determine the surgical outcomes of fungating sarcomas.
Methods: This descriptive study was conducted in Orthopaedics and Spine Unit Hayatabad Medical Complex Peshawar and Rehman Medical Institute Hayatabad Peshawar from 2nd January 2016 to 2nd December 2020.Patients of both gender and all ages with fungating sarcoma of bony or soft tissue origin meeting the inclusion criteria were included in this study and operated. Analysis of patient's characteristics, tumour type, surgery, complications, recurrence and mortality was noted in each case.
Results: We operated 16 patients of fungating sarcomas. The mean age was 30.5±17 years. All patients were male. Soft tissue sarcoma was present in 8(50%) and bony in 8(50%). Upper limb sarcoma was noted in 9(56.25%),lower limb in 6(37.5%) and neck in 1(6.25%) patient. Resection of soft tissue sarcomas followed by reconstruction was performed in 4(8.69%) patients. Forequarter amputation was performed in 6(37.5%) cases, 3(18.75%) patients underwent hip disarticulation and 3(18.75%) patients had transfemoral amputations. Local complication was noted in 4(25%) patients with 3(18.75%) had wound dehiscence and 1(6.25%) had local recurrence at three months. Post surgery the mean survival time was 8 months and at 2 years follow up only 4(25%) patients were alive.
Conclusion: Poor post operative prognosis and high mortality was noted for fungating sarcoma in our series. We however advise surgery for fungating sarcomas to alleviate symptoms and misery of these patients and improve their quality of life.
Keywords: Fungating, Sarcomas, Tumour.

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INTRODUCTION

Sarcomas are rare cancers comprising of 0.5 to 1% of all cancers.[1] Prognosis of sarcomas is multifactorial and depends on age of the patient, grade, size, extent and type of tumour.[2] Sarcomas can present as lump, pain, loss of function and pathological fractures. Ulcerating sarcoma is due to the cutaneous infiltration of a malignant lesion from the subcutaneous or deeper tissues that leads to the formation of a fungating lesion which can cause significant pain, bleeding, and bad smell leading to social segregation and poor quality of life.[3] Various terms are used for ulcerated malignant wounds and the term "fungating" is equivalent to "malignant cutaneous lesions" and denotes the appearance of raised cauliflower like nodules at the edges of the wounds.[4] Lack of large scale studies on fungating sarcomas hindes effective treatment guidelines. Most of the evidence comes from smaller case series and case reports including sarcomas originating from chronic osteomyelitis.[5-7] Earlier diagnosis and prompt referral to sarcoma centres can lead to better outcomes. However, this requires significant patient and clinician education, awareness campaigns and development of dedicated centres for treatment of these rare tumours in a multidisciplinary setting. This situation becomes even more complex in resource constrained countries and health care systems posing a significant challenge to the treating surgeon.[8]

We aimed to review the surgical outcomes of fungating sarcomas presenting to our dedicated sarcoma treatment centre.

METHODS

We conducted this descriptive study in Orthopaedics and Spine Unit Hayatabad Medical Complex Peshawar and Rehman Medical Institute Hayatabad Peshawar from 2nd January 2016 to 2nd December 2020. All patients of both gender and all ages with a bleeding, ulcerating or fungating wound secondary to an underlying sarcoma (Bone or soft tissue) were included in the study. For the purpose of this study any breach in the continuity of the epidermis/dermis due to an underlying sarcoma was considered as an inclusion criterion. Besides incisional biopsy patients who were surgically treated in other hospitals were excluded. The study was approved by the Ethical Committee of both hospitals. Informed written consent was taken from all the participants of our study. In the included subjects complete history, physical examination and relevant investigations were undertaken.

A multi disciplinary approach involving Orthopaedic surgeon, oncologist, radiologist, physiotherapist and psychologist was adopted for each and every case. Soft tissue sarcomas were managed as per protocols of Grimer.[9] Bony sarcomas were managed as per Gerrand protocols.[10] Post operative follow up was done as per European Society of Medical Oncology(ESMO) recommendation.[11]

We analysed our data with SPSS version 24. Important quantitative variables were represented as mean and standard deviation while qualitative variables as frequency and percentages. Data was presented in table where necessary.

RESULTS

In this study 16 patients of fungating sarcomas were included. The mean age was 30.5±17 years. All patients were male. Soft tissue sarcoma was present in 8(50%) and bony in 8(50%). Upper limb sarcoma was noted in 9(56.25%),lower limb in 6(37.5%) and neck in 1(6.25%) patient. Osteosarcoma was diagnosed in 7(43.75%), Spindle sarcoma in 2(12.5%), Liposarcoma in 2(12.5%) synovial sarcoma in 2(12.5%) patients. Dermatofibrosarcoma protuberans, Ewing sarcoma and Epitheloid sarcoma was present 1(6.25)% patient each.(Table I).

Metastatic disease at presentation was documented in 6(37.5%). All the patients had grade 3 tumours except one patient with Dermatofibrosarcoma protuberans (DFSP). Resection of soft tissue sarcomas followed by reconstruction with either split skin graft alone or local flaps and split skin grafts for coverage (Fig.I, II) was performed in 4(8.69%) patients. Forequarter amputation was performed in 6(37.5%) cases while 3(18.75%)patients underwent hip disarticulation and 3(18.75%) patients had transfemoral amputations. Local complications were noted in 4(25%) patients,3(18.75%) had wound dehiscence requiring wound debridement. One patient had local recurrence at three months following surgery and required above elbow amputation. Post surgery the mean survival time was 8 months and at 2 years follow up only 4(25%) patients were alive. The mean overall survivorship of patients with bone sarcomas was less than soft tissue sarcomas (6.8 Vs 9 months) and was independent of metastasis at presentation.

DISCUSSION

Sarcomas are rare type of tumours and despite significant advances in its management and awareness over the last few decades, the average size at presentation is still around 10 cm.[12,13] Tumour size, grade, extent and type are the main prognostic factors that determines the post treatment survivorship.[2] Fungating sarcoma is an extreme presentation and can lead to significant anxiety in the patient and pose a significant challenge for the operating surgeon. All those factors which are associated with poor prognosis for soft tissue sarcomas are usually present in fungating sarcomas hence leading to poor outcomes.[14] When compared to non-fungating sarcomas the overall results including survivorship are very poor.[15-17] Fungating sarcomas have usually larger size making limb salvage surgery challenging. In our study we achieved limb salvage in only 25 % of the cases.This is similar to other studies.[1] The decision for surgical intervention becomes more difficult in a palliative setting unless the condition causes severe non-responsive pain, a pathological fracture or a fungating lesion leading to a significant deterioration in quality of life.[18-20]

The average age at presentation for these lesions in literature is higher than ours.[5] This can be explained by the fact that most studies with fungating tumours have only reported on soft tissue sarcomas, which tend to present in slightly older patients, whereas our study addressed bone and soft tissue sarcomas with bony sarcomas usually presenting in the younger age group.

In our study the mean duration of symptoms before presentation to our unit was 8.7 months. This unacceptable delay is multifactorial and possible

reasons were lack of awareness, lack of access to proper health care and dedicated sarcoma facilities and low socioeconomic status. Similarly in a study of 18 musculoskeletal oncology cases, the authors found that the reasons for delayed presentation were lack of awareness, lack of health facilities and poor socioeconomic status of the patients.[8] However, in another series the main reason for delayed presentation of soft tissue sarcomas in elderly patients were neglect on part of the patients.[1]

In a study by Parsons *et al* **[21]** 40 major surgeries including forequarter and hindquarter amputations for advanced cancer cases were performed and they found that 14(35%) patients had fungating tumours at presentation. This shows that even in developed countries the rate of fungating musculoskeletal tumours is still alarming. It is interesting to note that in their study the survival rates varied from 3 to 13 months depending upon the intent of surgery which is not much different from our results. This confirms the fact that once a tumour fungates even in the best centres the results are quite dismal. These authors also noted that all patients who attained negative margins at surgery, 79% still had recurrence of disease either locally or at distant sites. We documented similar results in our study as six of our patients had metastasis at presentation and only one patient was alive with no evidence of disease at two years follow up. Merimsky**[22]** reported an incidence of 38% of fungating tumours in their series of major amputations. Potter**[5]** also concluded that a fungating lesion was an independent poor prognostic factor but the author noted better survivorship of his patients than ours. This may be due to the fact that our series was heterogenous and all bony sarcomas were high grade which qualified for poor prognosis as an independent factor. These poor results should come as no surprise indicating that the key to improving outcomes in these cases is education and awareness which can lead early presentation of these tumours to health care facilities.[23]

We accept that the limitations of our study are its descriptive design, smaller sample size and short follow up period. As we do not have a national tumour registry it is difficult to estimate the true incidence of sarcomas let alone the fungating ones. We believe that because of the associated morbidity and stigma with these fungating lesions, lack of facilities and poor socioeconomic background, some patients may never present to any clinician and that the actual figures for these tumours maybe much higher.

Table I: Details of patients included in our study

No.	Age (years)	Diagnosis	Metastasis at presentation	Location	Surgery	Complications	Status at last follow up(2 years)
1	60	Dermatofibrosarcoma protuberans (DFSP)	Yes	Leg	Above knee amputation	None	Alive with disease
2	15	Osteosarcoma	No	Shoulder	Forequarter amputation	None	Alive with disease
3	53	Spindle cell sarcoma	Yes	Shoulder	Forequarter amputation	None	Deceased
4	22	Osteosarcoma	No	Knee	Hip Disarticulation	Wound dehiscence	Deceased
5	17	Spindle cell sarcoma	No	Shoulder	Resection+ Local flap	Wound dehiscence	Deceased
6	28	Synovial sarcoma	No	Elbow	Resection+ split skin graft	None	Deceased
7	17	Osteosarcoma	Yes	Hip	Hip Disarticulation	None	Deceased
8	48	Synovial sarcoma	Yes	Thigh	Hip Disarticulation	None	Deceased
9	50	Liposarcoma	No	Shoulder	Forequarter amputation	None	No evidence of disease
10	21	Osteosarcoma	No	Leg	Below knee amputation	None	Alive with disease
11	10	Osteosarcoma	No	Shoulder	Forequarter amputation	None	Deceased
12	25	Osteosarcoma	Yes	Shoulder	Forequarter amputation	None	Deceased
13	17	Osteosarcoma	No	Knee	Above knee amputation	None	Deceased
14	60	Liposarcoma	Yes	Neck [Posterior]	Resection + local flaps.	Wound dehiscence	Deceased
15	13	Ewings sarcoma	No	Shoulder	Forequarter amputation	None	Deceased
16	32	Epithelioid sarcoma	No	Arm	Resection + split skin graft	Local recurrence	Deceased

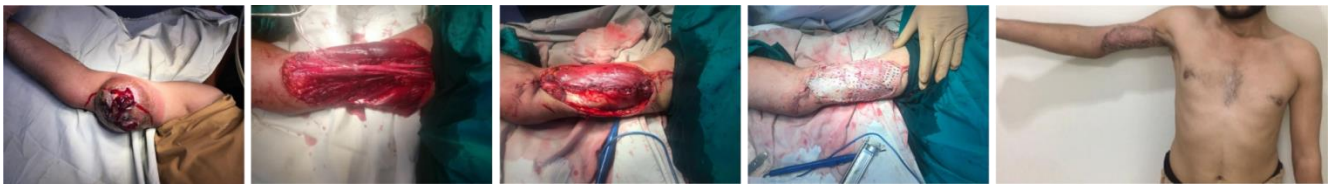

Fig. I Epithelioid sarcoma of the right arm resected with clear margins after preservation of the neurovascular bundle followed by split skin graft for wound closure. Graft uptake can be seen in 6 weeks.

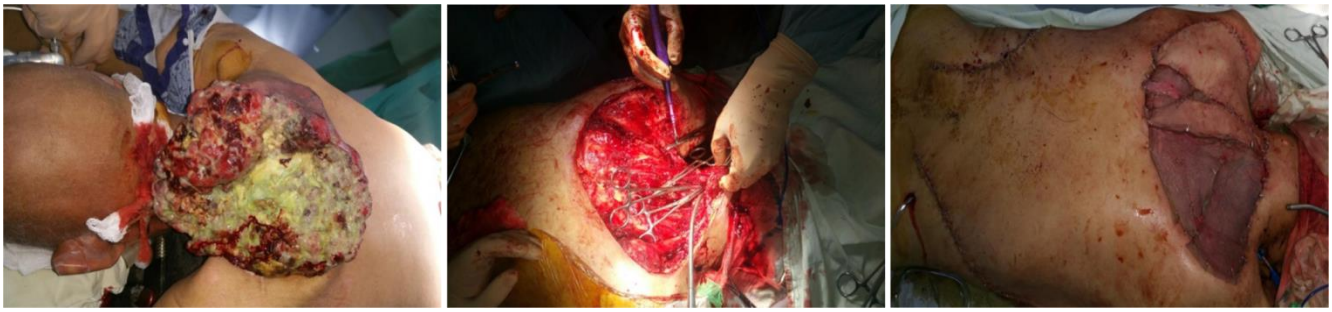

Fig. II. Large fungating liposarcoma of the the neck in a 60 years old patient excised and residual defect was reconstructed with bilateral Latissimus Dorsi flap and skin graft.

CONCLUSION

Poor post operative prognosis and high mortality was noted for fungating sarcoma in our series. We however advise surgery for fungating sarcomas to alleviate symptoms and misery of these patients and improve their quality of life. We must emphasize the importance of awareness among public and medical professionals about these rare tumours and their associated red flag features. Development of dedicated sarcoma units to improve the care of these patients are mandatory.

Conflict of Interest: None

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