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Mid-term outcome of wide resection in musculoskeletal fibrosarcoma patients

Resit Sevimli[1], Adem Emeli[2], Semih Eriten[3]

[1]Inonu University, Faculty of Medicine, Department of Orthopaedic and Traumatology, Malatya, Turkey
[2]Usak Resarch Hospital, Clinic of Orthopaedic and Traumatology, Usak, Turkey
[3]Malatya Education and Research Hospital, Department of Emergencey, Malatya, Turkey

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Abstract
The aim in this study, musculoskeletal system fibrosarcomas which are rare malignant soft tissue tumors that originate from fibroblasts, was to evaluate the mid-term outcome of patients that were diagnosed, treated and followed-up at our clinics. Included in the study were 12 patients treated for fibrosaecoma at our clinics between 2014 and 2017. The patients were evaluated in terms of age, location of tumor and time of resection, and were followed-up for mid-term recurrence. A wide resection was performed on all patients. No recurrence was found in all but one patient during follow-up. Musculoskeletal system fibrosarcomas are rare but represent high mortality and morbidity risks since the diagnosis is commonly delayed. The most frequently seen symptom is a painless mass reaching large dimensions. Early diagnosis plays a major role in prognosis, as is the case with other malignant tumors. In conclusion, we suggest that malignancy should be considered in the presence of giant, fixed and painful tumors, and a wide resection should be applied.

Keywords: Soft tumor, fibrosarcoma, wide resection, recurrence, primary organs, diagnose

Introduction

Fibrosarcoma is a malignant neoplasia arising from fibroblasts.1 Incidences increase among those in the 30–55-year age group, and especially at around 45 years, although it can be seen in all age groups.2 The tumor is frequently located in the deep fascia of the distal extremity of the skeletal system. Growth is commonly painless, and so diagnosis may be delayed accordingly.3 The subgroups are adult fibrosarcoma, infantile fibrosarcoma, myxofibrosarcoma, low-grade fibromyxoid sarcoma and sclerosing epithelioid fibrosarcoma.4 Fibrosarcomas are solitary lesions and are generally infiltrative, however they can be misleadingly well demarcated. A histopathology reveals intersecting fibroblast bundles that are sometimes in a herringbone sequence. Nuclear atypia and mitotic activity will be present but may be mild in some cases and grading is important in terms of prognosis in such tumors.5 The invasion of the skeletal structure and superficial ulcerations are seen in advanced cases.6 They are normally encountered as painless masses, provided there has been no invasion into the nerve trunks.7 Recurrences are especially high in cases with an insufficient resection.8 In this study, recurrences identified at mid-term follow-up were evaluated in fibrosarcoma cases in which a wide resection was carried out.

***Coresponding Author:** Resit Sevimli, Inonu University, Faculty of Medicine, Department of Orthopaedic and Traumatology, Malatya, Turkey
E-mail: resitsevimli@hotmail.com

Material and Methods

All patients participating in the study provided written informed consent. After obtaining the required informed consent, 12 patients who were treated in the clinic with a diagnosis of fibrosarcoma between 2014 and 2017 were included in the study. For the evaluation of the patients, after systemic physical examination of all patients, in radiological examinations, lesions reaching gigantic dimensions in direct radiographs were seen, in addition to mass lesions with an expending character; and pathological vascularizations that were heterogeneous in T1 and hyperintense in T2 images, located in the soft tissues, and separated from arterial and nervous structures by a thin capsule and causing obstructions, as seen in MRI images (Figure 1 and 2).

The patients were followed-up for location of the tumor, age of the patients and time of resection, and the mid-term outcomes were evaluated.

Statistical analysis

Statistical comparisons were performed using the paired-samples

t-test and IBM SPSS Statistics, version 21 (IBM Corp., Armonk, NY, USA). Statistical significance was accepted at $p<0.05$.

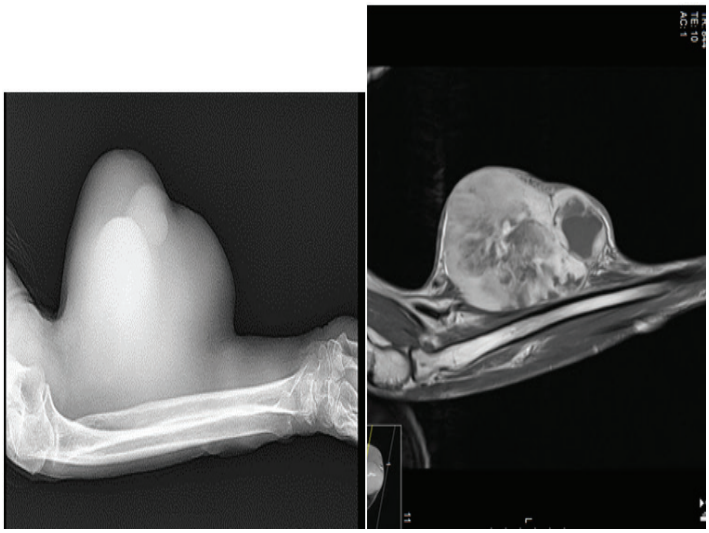

Figure 1. Direct graphy and MRI images of a 56-year old patient who presented with a forearm fibrosarcoma

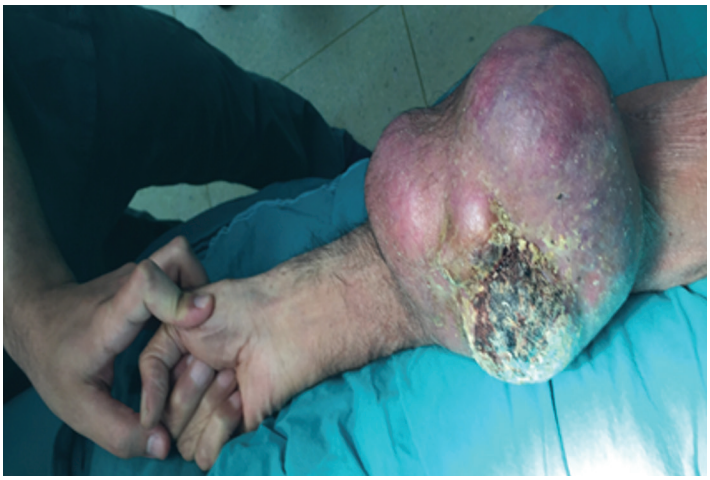

Figure 2. Fibrosarcoma reaching gigantic dimensions and skin necrosis seen in the same patient

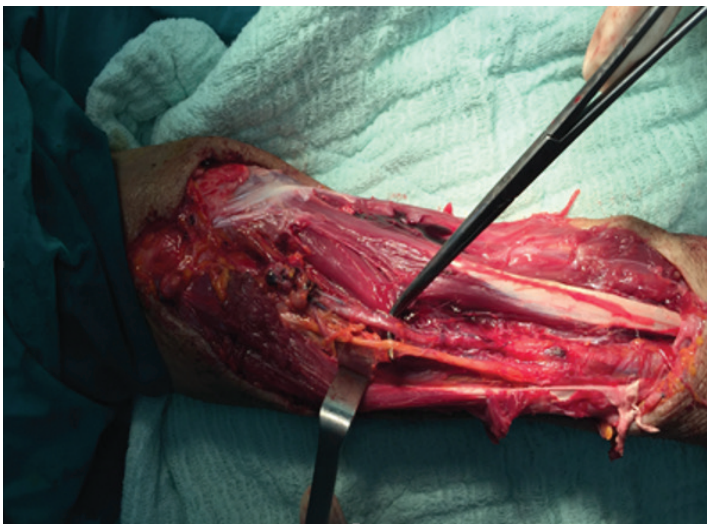

Figure 3. Neurovascular bundle seen intact after a wide excision

Results

The age range of the patients was 15–70 years (mean 46) (Table 1). The tumors were localized in the upper and lower extremities in seven (60%) and five (40%) patients, respectively (Table 2). The most common localization in the upper extremity was at the level of the forearm/elbow. According to Eneking's soft tissue tumor classification, three patients were at stage 1, five were at stage 2 and four were at stage 3 (Table 3). All tumoral structures were resected, reaching the mass by way of an incision around the tumor, including the skin, under general anesthesia. Skin defects were closed by plastic surgeons using autografts (Figure 3), and the patients were referred to the appropriate departments for chemotherapy and radiotherapy.

The 12 patients included in the study underwent wide excisional surgery following diagnosis and neoadjuvant therapies. No recurrences were found in all but one patient in the Stage 3 group during routine follow-up visits performed in every six months. The case with the recurrence, which was detected in the 8th month of follow-up, underwent a radical resection again, excising the compartment involved.

Table 1. Age distribution of patients

AGE	Number of Patients
15–20	1
20–30	2
30–40	1
40–50	3
50–60	3
60–70	2
AGE RANGE: 15–70	
MEAN: 46	

Table 2. Distribution of patients by anatomic localization

Anatomic Distribution	
Localization	**Number of Patients**
Upper Extremity (60%)	
Shoulder and Arm	2
Elbow and Forearm	4
Hand and Wrist	1
Lower Extremity (40%)	
Hip and Thigh	2
Knee and Leg	2
Foot and Ankle	1

Table 3. Stage and rate of recurrence in 12 patients with fibrosarcoma

Stage and Rate of Recurrence		
Stage	**Number of Patients**	**Rate of Recurrence**
1	**3**	**0**
2	5	0
3	4	1
4	0	0

Discussion

Fibrosarcomas account for 3 percent of all adult sarcomas among all soft tissue tumors and are generally seen more often in those of advanced age.9 No etiologic cause can be detected in most cases, although genetic factors, radiation, trauma and chemical agents are most frequently blamed for the development of fibrosarcoma. Fibrosarcomas originate from the deep tissues of the extremities, head and neck. They may originate in areas of the body subjected to radiotherapy, or areas with foreign material applications.10

Fibrosarcomas are formed from fibroblasts and include varying amounts of collagen. The lesions are hard and single lobuled with regular contours.

The histories of patients reveal a painless mass with a slow growth, and this frequently causes tumors to reach large dimensions. Patients may complain of palpable masses that can lead to compressive symptoms. Computed tomography and magnetic resonance imaging are the commonly used methods in diagnosis, while fine needle aspiration or tru-cut biopsies can be helpful in making a histopathological diagnosis. Microscopic views of fibrosarcomas may be low grade or high grade.11

The most frequent site of metastasis is the lungs via the hematogeneous route. Low rates of local recurrence and five-year survival levels of up to 80 percent can be achieved through appropriate surgical margins and adjuvant radiotherapy. The use of chemotherapy has been suggested to decrease local recurrence rates, although the roles of chemotherapy and radiotherapy are still controversial.12

The treatment of choice in low grade tumors is surgery, as is the case with other soft tissue sarcomas. The tumor should be widely excised with the surrounding normal tissue (wide excision), and radiotherapy and chemotherapy can be applied preoperatively and/or postoperatively, although it is not a rule in high-grade tumors. The rate of recurrence is around 5 percent in soft tissue sarcomas resected with an appropriate surgical margin, and the rate of success is even higher in childhood fibrosarcomas.13

Cases series of fibrosarcoma are rarely reported in literature, although case reports more frequent.

Shvarts et al.14 reported bladder invasion in a case of fibrosarcoma with distant organ metastasis, while Bahrami et al. 15 reported a 5 percent rate of recurrence in a large series, compared to Scott et al.16 who reported a 6 percent recurrence rate.

In this present case report, a recurrence was found in only one case, which is compatible with the findings in literature. No recurrence was found in the rest of our cases during mid-term follow-ups following a wide resection.

Conclusion

In conclusion, musculoskeletal tumors are rare, but are associated with high mortality and morbidity when they occur. That said, patient survival can be increased when the tumors are excised with the appropriate margins with surgical techniques.

Competing interests
The authors declare that they have no competing interests.

Financial Disclosure
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Ethical approval
Before the study, permissions were obtained from the local ethical committee.

Reşit Sevimli ORCID: 0000-0002-5394-9100
Adem Emeli ORCID: 0000-0002-1640-4276
Semih Eriten ORCID: 0000-0001-8516-372X

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