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Title: Myxofibrosarcoma: A Difficult-To Treat Soft Tissue Sarcoma – Considerations After 85 Patients

Giovanni Beltrami¹, Daniel A. Müller², Guido Scoccianti¹, Rodolfo Capanna¹

¹Orthopaedic Oncology Unit, Careggi University Hospital, Florence, Italy

²Department of Orthopaedics, Balgrist University Hospital, Zurich, Switzerland

Giovanni Beltrami: giovannibeltrami@virgilio.it
Daniel A. Müller: daniel.a.mueller@me.com
Guido Scoccianti: guido.scoccianti@alice.it
Rodolfo Capanna: rodolfo.capanna@gmail.com

BACKGROUND

Myxofibrosarcoma (MFS) is a variant of the group of malignant fibrous histiocytomas and is one of the most common sarcomas in the extremities of elderly patients. MFS represents a particularly difficult to treat type of soft tissue sarcoma due to a high local recurrence rate. Only a few clinical outcome studies dealing with this disease were reported in the literature.

QUESTIONS/PURPOSES

This study provides a large case series with the aim to improve treatment efficacy of MFS. The examined study questions are as follows: What is the local recurrence rate and the metastatic rate of this specific histologic diagnosis? What are the risk factors for a local relapse? Have patients with multiple local recurrences a higher metastatic rate? Do different histologic subtypes of MFS result in diverging outcomes?

PATIENTS AND METHODS

A consecutive case series of 85 patients who underwent a resection of MFS at the extremities and the superficial trunk from 2004 to 2013 was retrospectively reviewed. The mean age was 65.7 years (Range:21-90). The most common localization of the tumor was in the lower extremity (n=57;67.1%), followed by the upper extremity (n=24;28.2%) and the trunk (n=4;4.7%). In 53 patients (74.6%) the lesion was found subfascial, and the diameter of the tumor was in 28 cases (42.9%) smaller than 5cm, in 31 cases (36.5%) between 5 and 10 cm, and in 26 cases (30.6%) bigger than 10 cm. The histopathologic grading was performed using the FNCLCC criteria and revealed 21 (30.9%) grade 1 lesions, 9 (13.2%) grade 2 and 38 (55.9%) grade 3. Two patients (2.3%) showed lung metastases at time of diagnosis. In one of them adjuvant chemotherapy was performed. 49 patients (57.6%) underwent an external radiation therapy, whereas 2 patients were treated pre-operatively and the remaining 47 post-operatively.

RESULTS

In 23 cases (27.1%) the histologic work-up showed a marginal resection with contaminated margins. After a mean follow-up of 58 months (Range 2-132) a total of 16 local recurrences (18.2%) were observed. 5 patients (5.9%) suffered from more than one local relapse. The local recurrence free survival was 78.3% (95%CI:66.2-86.5%) after 5 years and 75.0% (95%CI:61.3-84.5%) after 10 years. No statistically significant difference in local recurrence free survival was found for tumor size and depth. However, high-grade lesions (p=0.016) and inadequate margins (p=0.06) lead to a higher recurrence rate (see figure 1).

In 19 patients (22.3%) distant metastases were found after an average time of 17 months postoperatively. The metastasis free survival was 75.5% (95%CI:63.6-83.9%) after 5 years and 10 years, respectively. Only high-grade lesions were important risk factors for metastases (p=0.003).

An epithelioid subtype of MFS was diagnosed in 7 patients (8.2%). Interestingly, they showed a much worse outcome with a local recurrence rate of 43% and a metastatic rate of 71%.

CONCLUSION

MFS showed a higher recurrence rate compared to other common soft tissue sarcomas. Obtaining wide margins is difficult, due to diffuse microscopic spreading in the surrounding tissue. But, multiple local

recurrences do not result in a higher metastatic rate. Special attention has to be paid to the epitheloid subtype, which seems to have a very poor outcome.

FIGURES

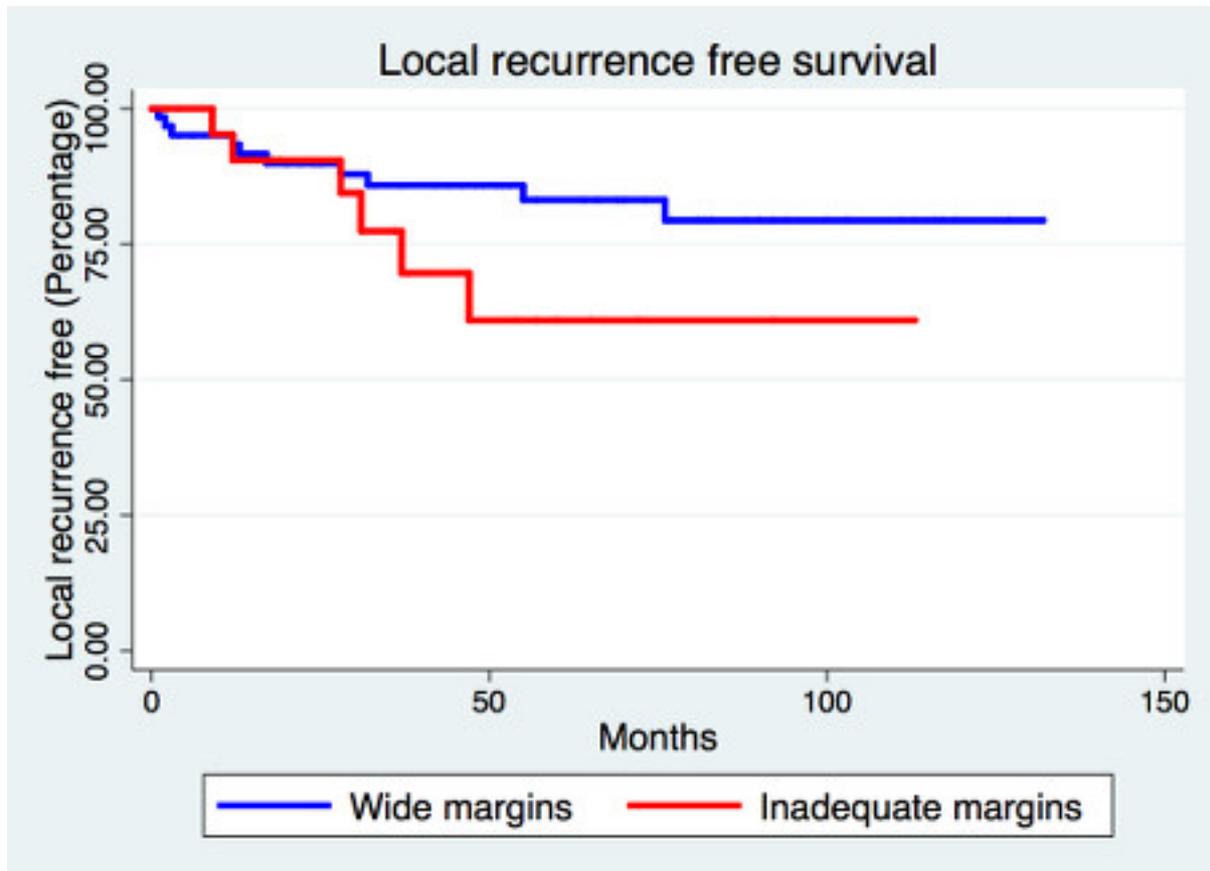


Figure 1: Local recurrence free survival according to achieved margins