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CASE REPORT

Short-term comparative results of segmental resection versus radical resection in two patients with myxofibrosarcoma of the limbs - case series

Sergiu-Andrei Iordache*, Bogdan Șerban* **, Andreea Vlad*, Adrian Cursaru* ****
*Orthopedics Department, University Emergency Hospital Bucharest, Romania
**"Carol Davila" University of Medicine and Pharmacy, Bucharest, Romania
***"Grigore Alexandrescu" Children's Clinical Emergency Hospital Bucharest, Romania

Correspondence to: Sergiu-Andrei Iordache, PhD,
Orthopedics Department, University Emergency Hospital, Bucharest,
169 Independenței Street, Code 050098, Bucharest, Romania,
Mobile phone: +40766 315 091, E-mail: sergiu.iordache92@yahoo.com

Abstract

Introduction: Myxofibrosarcoma is a rare subtype of soft tissue sarcoma with a locally infiltrative behavior and ability to determine distant metastases.

Materials and methods: We presented two myxofibrosarcoma cases who benefited from segmental or radical resection.

Management and outcome: In the case of the 80-year-old woman, with grade 3 myxofibrosarcoma, we practiced radical surgery with scapulohumeral disarticulation followed by adjuvant radiotherapy.

The therapeutic option for the 77-year-old man with grade 2 myxofibrosarcoma was segmental resection followed by adjuvant radiotherapy. After three months, the patient was in a good clinical condition with no sign of local recurrence, but with the presence of pulmonary metastases for the patient who benefited of segmental resection.

Discussion: The radical resection had better short-term results, with no local or distant metastases at three months after surgery, although the tumor had a higher grade (G3), compared to segmental resection practiced for a lower grade tumor (G2), in which case the patient developed pulmonary metastasis at three months follow up.

Conclusion: Myxofibrosarcoma represents a challenging situation regarding the management due to its unpredictive clinical course. Our cases raised the following question: should we consider treating it more aggressively in order to obtain good local control and reduce the risk of metastasis?

Keywords: myxofibrosarcoma, segmental resection, radical resection, treatment, case report

Introduction

Myxofibrosarcoma (MFS) is a malignant neoplasm of fibroblastic origin and represents one of the approximately 50 histological subtypes of soft tissue sarcomas. The most common clinical presentation for MFS is a palpable, painless soft tissue mass. It occurs most commonly in the extremities, in elderly patients, in the sixth to eighth decades of life.

Histologically, it arises in connective tissue characterized by a proliferation of spindle cells in a myxoid stroma with high potential for local recurrence and metastasis. At gross pathologic examination, the lesion is multilobulated, with necrosis or hemorrhage on the cut surface. Increased attention should be paid intraoperatively because the border of the tumor may appear well defined at first sight, but a microscopic spread along muscle fibers and fascial planes is often present [**1**].

Because of the insidious clinical presentation, the patient usually postpones presentation to the hospital. Any palpable mass exceeding 5 cm raises the suspicion of a malignant tumor.

The diagnosis algorithm includes performing a magnetic resonance imaging (MRI), angiography and scintigraphy or positron emission tomography (PET-scan), but the positive diagnosis can only be made by incisional biopsy. The European Society for Medical Oncology and European Rare Adult Solid Cancers Network (ESMO-EURACAN) recommends MRI as the main imaging modality in the extremities, pelvis and trunk [**2**].

The treatment for myxofibrosarcoma varies from primary surgical resection to neoadjuvant or adjuvant radiotherapy or cytotoxic chemotherapy. The actual mainstay of treatment consists in a wide surgical resection with a 2 cm soft tissue margin, which may sometimes require complex vascular and plastic surgery reconstruction. Rare cases of spontaneous regression or after incisional biopsy are also described [**3**].

Materials and Methods

We comparatively followed the clinical evolution and the moment of appearance of metastases or signs of local recurrence in two patients diagnosed with myxofibrosarcoma, who presented to the Orthopedics Department of the University Emergency Hospital, Bucharest.

Case report

An 80-year-old woman with a history of 12 months palpable mass in the proximal arm, with a fast growth in the last 6 months, presented to the Orthopedic Department of the University Emergency Hospital, Bucharest (**Fig. 1**). The clinical examination revealed a palpable mass in the proximity of the scapulohumeral joint with limited range of motion and moderate pain. Lab values showed decreased levels of hemoglobin-9.6 g/ dl (N:12.5-16.3 g/ dl) and increased values of inflammatory markers: CRP-11.7 mg/ dl (N:0.0-0.9 mg/ dl), ESR-108 mm/ h (N:5-10 mm/ h), fibrinogen-601 mg/ dl (N:238-498 mg/ dl) and elevated creatine kinase I-351 U/ L (N: 39-308 U/ L). The diagnosis protocol in this case consisted of a distant metastasis assessment using a computerized tomography (CT) scan and the evaluation of local invasion using an MRI and an angiogram (**Fig. 2**). The patient presented no sign of distant metastasis and was proposed for an incisional biopsy. The anatomopathological examination confirmed the diagnosis of grade 3 myxofibrosarcoma.

Fig. 1 Preoperative macroscopic image of the 80-year-old female patient with upper limb MFS (personal archive)

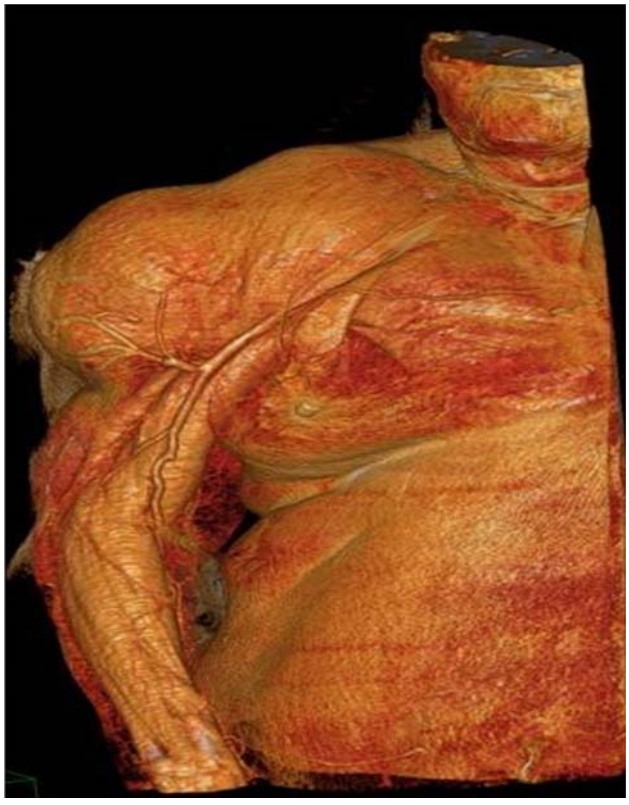

Fig. 2 Preoperative CT-scan image of the 80-year-old female patient with upper limb MFS (University Emergency Hospital, Bucharest)

Because of the proximity of the scapulohumeral joint and the local invasion, the patient was admitted for scapulohumeral disarticulation. After surgery, no significant perioperative complications were observed and the patient was discharged in good clinical condition. Shortly after discharge she was admitted to the Oncology Department for adjuvant radiotherapy.

In the next three months, during the oncological follow up, the patient had no sign of local recurrence or distant metastasis.

Another case was a 77-year-old man admitted to the Orthopedic Department of the University Emergency Hospital, Bucharest with a history of 6 months palpable mass of the lateral 1/ 3 of the thigh. According to the patient, the symptoms appeared 3 months earlier when a slight pain was noted in the right lower limb. The clinical examination revealed a palpable mass on the anterior lateral third of the thigh. The approximate dimension of the tumor was 14/ 6 cm. No other remarkable findings were observed at the physical examination.

Fig. 3 MRI COR-T1 image (reproduced with the consent of the patient from his personal archive)

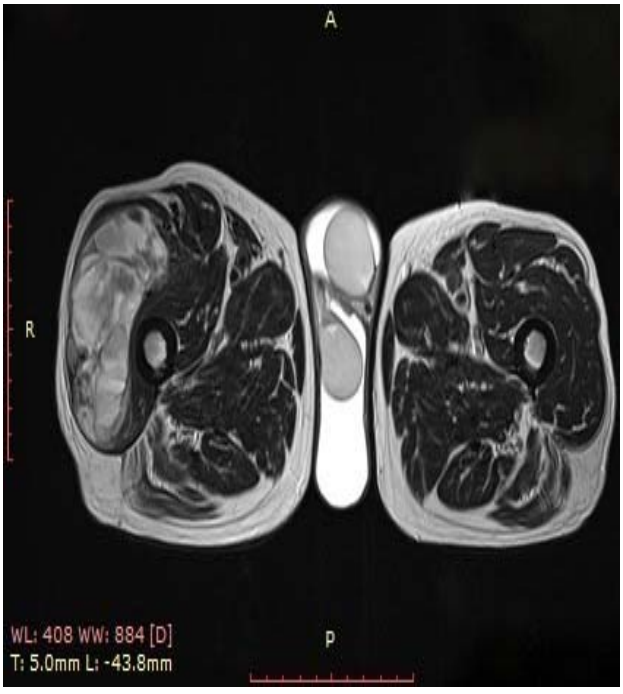

Fig. 4 MRI TRA-T2 image (reproduced with the consent of the patient from his personal archive)

The biological examination revealed elevated inflammatory markers: ESR-54 mm/ h (N:5-10 mm/ h); CRP-3.7 mg/ dl (N:0.0-0.9 mg/ dl) and a decreased level of hemoglobin - 11.6 g/ dl (N:12.5-16.3 g/ dl). The diagnosis process started with an MRI of the thigh, which described an infiltrative tumor (**Fig. 3,4**). For further investigation, a CT-scan and a PET-CT were performed (**Fig. 5,6**).

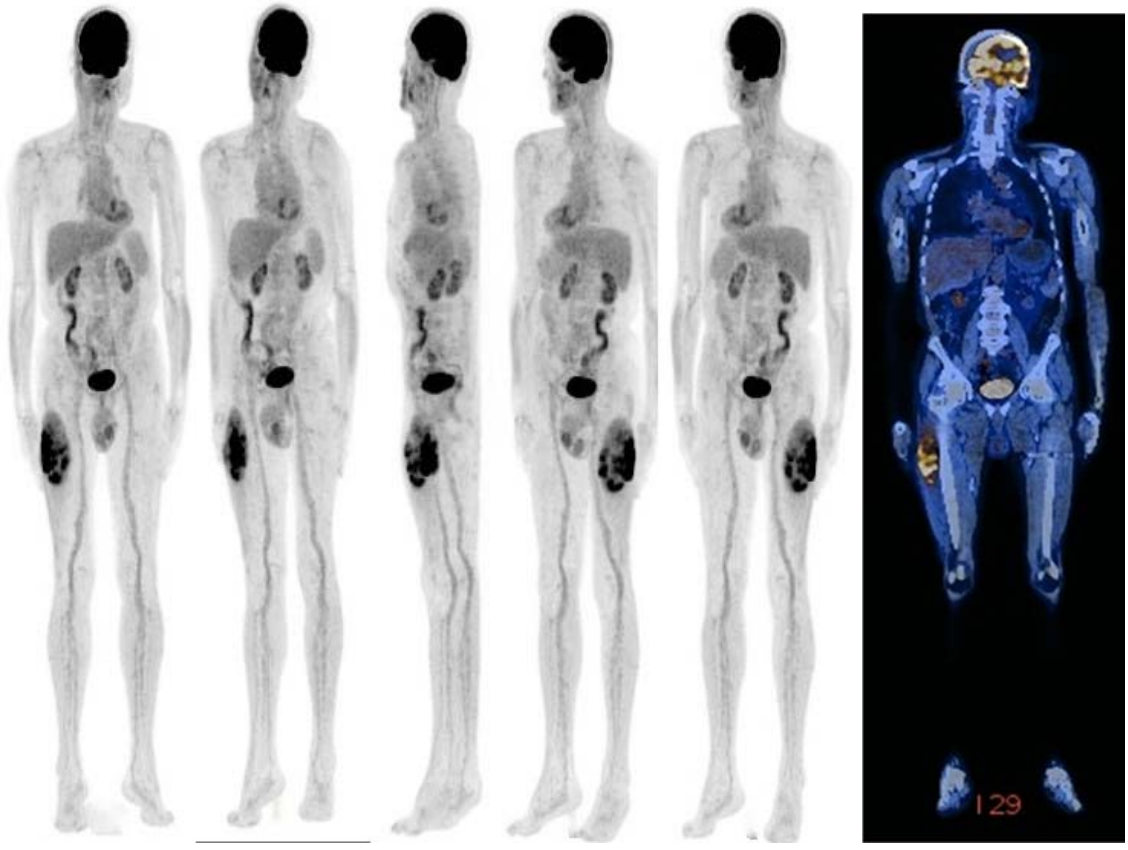

Fig. 5,6 PET-CT images (reproduced with the consent of the patient from his personal archive)

No secondary determinations were reported. After the incision biopsy, the histological examination revealed a grade 2 myxofibrosarcoma. The patient was admitted first to the Oncology Department for neoadjuvant radiotherapy and afterwards he was admitted to the Orthopedics Department for segmental resection of the tumor with wide surgical resection and free oncological limits. After surgery, the patient had no significant complications and he was discharged without any special restrictions. Shortly after, he was admitted to the Oncology Department for adjuvant chemotherapy and radiotherapy.

Three months after segmental resection, the patient presented no sign of local recurrence with good clinical condition, but the CT-scan revealed the presence of pulmonary metastases for which he underwent a new course of chemotherapy.

Discussion

The goal of this comparative case report was to evaluate the presence of local recurrence or distant metastases considering the type of surgical resection (segmental vs. radical resection), the histological degree of the tumor, the period from onset to surgery,

the state of the resection margins and the use of radiotherapy or chemotherapy.

Recent studies have shown that older age and positive/ close margins were significantly associated with a higher risk of local recurrence [**4,5**]. A positive margin is a significant poor prognostic indicator of local recurrence in patients with myxofibrosarcoma. In order to be able to obtain free oncological limits, radical surgeries are often needed [**6**].

The tumor grade and metastasis are independently associated with myxofibrosarcoma specific survival [**7**]. The retrospective analysis of a group of 75 patients revealed that the rate of local recurrence was independent of the histologic grade, but only intermediate and high-grade neoplasms metastasized [**8**]. In our study, the patient with segmental resection had grade 2 MFS, while the other one with radical resection had grade 3 MFS. The radical resection had better short-term results, with no local or distant metastases after three months, although the tumor had a higher grade (G3), compared to segmental resection practiced for a lower grade tumor (G2), in which case the patient developed pulmonary metastasis after three months.

The effect of radiotherapy (RT) on local control is still unclear and there is no correlation between RT and local recurrence [**9,10**]. After surgery, our patients were admitted to the oncology department for adjuvant radiotherapy. The current scientific data in literature are still incomplete and require further studies on significant groups of patients.

Conclusions

Myxofibrosarcoma represents a challenging situation regarding the management due to its unpredictive clinical course. In our case, the patient with segmental resection had a lower grade tumor (G2) and six months between the detection of the mass and surgery, compared with the G3 tumor and the 12 months before surgery in the other patient. Yet, the one with segmental resection developed pulmonary metastases. The therapeutic protocol for the two patients was similar. Both patients were evaluated for distant metastasis and were admitted postoperatively to the Oncology Department for radiotherapy and chemotherapy. It should be mentioned that the patient who benefited from segmental resection also underwent neoadjuvant radiotherapy. At three months postoperatively and after cancer therapy, none of the patients showed signs of local recurrence.

After the segmental resection of the tumor, the patient began to mobilize with no gait restrictions, so the functional result was very good compared to the functional result after amputation.

Radiation therapy has been shown to be effective in ensuring a good local control for the two.

For statistically significant results a larger group of patients and a longer follow-up period is required.

Conflict of Interest statements

Authors state no conflict of interest.

Informed Consent and Human and Animal Rights statements

Informed consent has been obtained from all individuals included in this study.

Authorization for the use of human subjects

Ethical approval: The research related to human use complies with all the relevant national regulations, institutional policies, is in accordance with the tenets of the Helsinki Declaration, and has been approved by the authors' institutional review board or equivalent commitee.

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