J Surg Med. 2024;8(12):207-210.
https://doi.org/10.28982/josam.7873

JOSAM

Journal of Surgery and Medicine

e-ISSN: 2602-2079

Case report
https://jsurgmed.com/

Recurrent malignant fibrous histiocytoma of the forearm: A rare soft tissue sarcoma

Kağan Gökçe [1], Demet Doğan [2], Merve Yazıcı [3], Ayşe Nimet Karadayı [4], Ahmet Midi [5]

[1] Okan University, Faculty of Medicine, Department of Surgery, Surgical Oncology Unit, İstanbul, Turkey
[2] Okan University, Faculty of Medicine, Department of Radiology, İstanbul, Turkey
[3] Okan University, Faculty of Medicine, 3rd Grade Student, İstanbul, Turkey
[4] Okan University, Faculty of Medicine, Department of Pathology, İstanbul, Turkey
[5] Bahcesehir University, Faculty of Medicine, Department of Pathology, İstanbul, Turkey

ORCID of the author(s)

KG: https://orcid.org/0000-0003-4712-0512
DD: https://orcid.org/0000-0003-0792-9042
MY: https://orcid.org/0009-0002-0935-0114
NK: https://orcid.org/0000-0001-8659-2155
AM: https://orcid.org/0000-0002-6197-7654

Abstract

Malignant fibrous histiocytoma (MFH), while rare, is the most prevalent soft tissue sarcoma (STS) found in extremities. Its prognosis hinges on factors such as tumor size, invasion depth, and differentiation degree. For MFH and all STSs, wide surgical resection is the gold standard treatment method. In this study, a 48-year-old woman, initially diagnosed with lipoma and with a mass excised from her right forearm, later exhibited recurring MFH masses in the same location. Superficial tissue ultrasonography identified malignant soft tissue tumors in the lesion area. Positron emission tomography was used to stage the tumor preoperatively, revealing fluorodeoxyglucose uptake in the repeating lesions. Right forearm magnetic resonance imaging confirmed the malignant lesions. Post imaging, the patient underwent wide surgical excision, eliminating the fascia. Regular follow-up revealed no recurrence or metastasis at 6 months and 1 year. Excisions frequently occur without radiological imaging, based only on a preliminary lipoma or sebaceous cyst diagnosis. However, excisions should follow radiological imaging, particularly when soft tissue tumors beneath the skin appear benign.

Keywords: malignant fibrous histiocytoma, soft tissue sarcoma, magnetic resonance

Corresponding Author
Kağan Gökçe
Okan University, Faculty of Medicine, Department of General Surgery, Surgical Oncology, İstanbul, Turkey
İçmeler Mahallesi, Aydınlı Yolu Caddesi No:2
34947 İçmeler-Tuzla / İstanbul, Turkey
E-mail: kagangokce2023@gmail.com

Informed Consent
The authors stated that the written consent was obtained from the patient presented with images in the study.

Conflict of Interest
No conflict of interest was declared by the authors.

Financial Disclosure
The authors declared that this study has received no financial support.

Published
2024 December 13

Introduction

Soft tissue carcinomas are significantly diverse and comprise various subtypes [1]. One rare subtype within this group is malignant fibrous histiocytoma (MFH). Although the specific cellular origin of MFH remains unclear, it shows a strong correlation with tumor cells, particularly fibroblasts or myofibroblasts. While most common in middle-aged and elderly adults, MFH can present at any age. It tends to grow in subcutaneous tissues, bones, muscles, and tendons. Though rare, MFH can also develop in internal organs [2]. Geographically, it predominantly occurs in the limbs (mainly the lower ones), trunk, or the head and neck region [3]. The cause of MFH remains unknown, but potential risk factors could include age, radiation exposure, genetic preconditions, and exposure to chemicals like arsenic and vinyl chloride [4]. MFH is identified by irregularly shaped spindle cells that deeply penetrate the dermis, often accompanied by hemorrhage, necrosis, and infiltration of lymphoid tissue cells. These tumor cells – usually large and atypical – exhibit irregular mitotic images [5].

At the onset of the disease, symptoms are generally mild or non-existent. However, MFH often manifests as a swiftly expanding mass or lump, potentially leading to distressing symptoms like pain, swelling, or tenderness. Growth of the tumor can result in the creation of a soft tissue mass, along with deteriorating strength or function, particularly if it impacts muscles and bones. It could also result in pathological fractures [6]. The definitive diagnosis of MFH is made via a pathological examination of a biopsy from the tumor. Such an examination not only confirms the diagnosis but also ascertains the level of malignancy.

How to cite: Gökçe K, Doğan D, Yazıcı M, Karadayı AN, Midi A. Recurrent malignant fibrous histiocytoma of the forearm: A rare soft tissue sarcoma. J Surg Med. 2024;8(12):207-210.

Treatment selection for MFH is influenced by factors such as the size of the tumor, its extent of spread, and the patient's overall health condition. The therapeutic options for MFH include surgery, radiation therapy, chemotherapy, and targeted therapies. Similar to other soft tissue carcinoma treatments, a wide surgical resection is the gold standard approach for MFH [3,7]. Complementary radiotherapy is particularly crucial for cases involving larger lesions, highly aggressive tumors, and positive or narrow surgical margins. Soft tissue sarcomas (STSs) like MFH may be responsive to radiotherapy either in combination with surgery or to target remaining tumors post-surgery [8]. The effectiveness of chemotherapy in MFH remains uncertain, but it can be used as an adjuvant therapy [7]. Conventional chemotherapy is usually reserved for cases with widespread disease, as substantial trials have not shown much benefit [3]. A significant meta-analysis concluded that additional use of doxorubicin did not enhance overall survival rates. Nonetheless, many experts suggest that pre-operative and/or supplementary chemotherapy can lower the risk of unseen local or metastatic disease [9].

The prognosis of MFH hinges on factors like tumor size, degree of spread, condition of regional lymph nodes, and the patient's overall health. Early diagnosis and suitable treatment can lead to successful outcomes in some instances. However, due to the disease's aggressive nature and potential to spread, the prognosis is frequently unfavorable [10].

Case presentation

A 48-year-old woman came to the clinic with three lesions on her right forearm that she noticed by touch. She had no other medical conditions. Her medical history showed a previous surgery for a lipoma in the same area where the lump was found. A physical examination showed a surgical scar on the extensor area of her right forearm, beneath which three separated lesions could be felt (Figure 1).

The patient, previously diagnosed with lipoma, had undergone surgery. However, despite a pathology report confirming MFH, a second operation was not performed. A superficial tissue ultrasound of the lesion area unveiled soft tissue tumors with potential malignant features. These included mildly lobulated contours, a solid appearance, and minimal blood flow, as seen through Doppler ultrasound (Figure 2). Prior to surgery, a positron emission tomography/computed tomography (PET/CT) scan was performed for staging. The scan revealed increased fluorodeoxyglucose (FDG) uptake in the recurring lesions (Figure 3).

Figure 1: Right forearm extensor area with pre-operative recurrence.

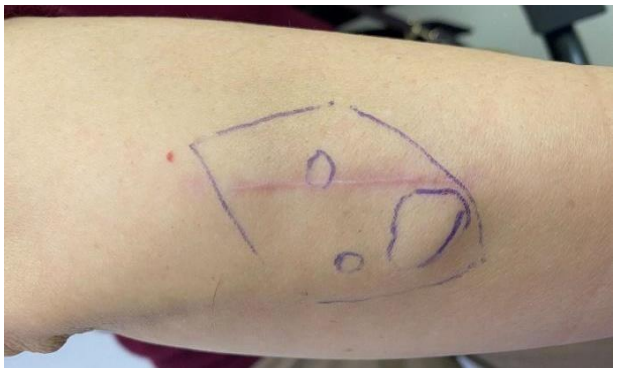


Figure 2: On US examination, a hypoechoic solid appeared as a lesion with smooth oval contours.

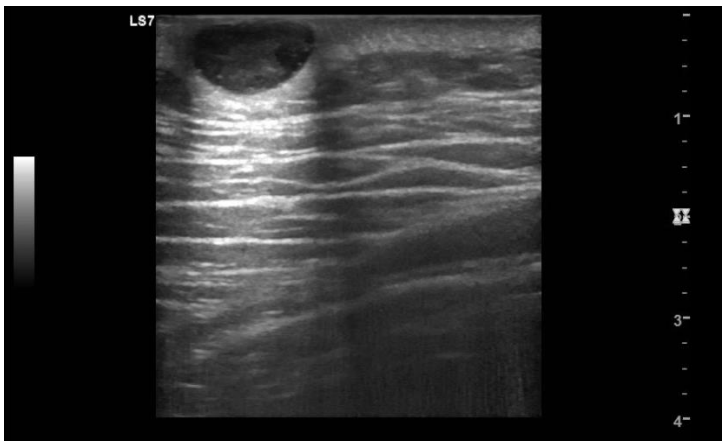
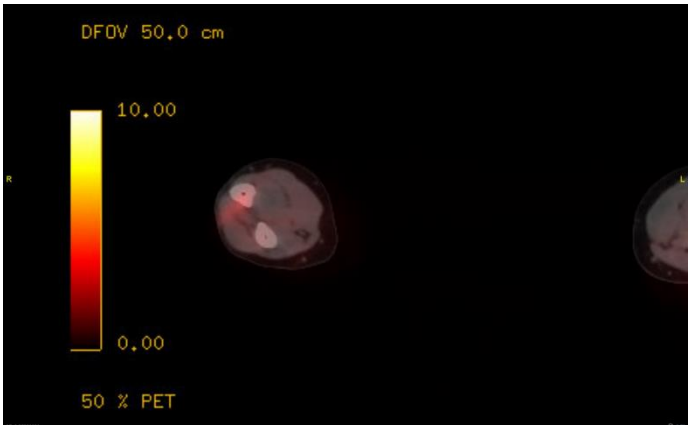
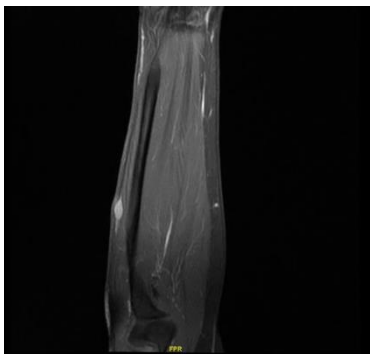


Figure 3: FDG uptake is compatible with malignancy in the axial section in PET-CT examination.



No metastasis was found in any areas beyond the identified lesion. Magnetic resonance imaging (MRI) of the right forearm showed muscle tissue invasion by the lesions (Figure 4).

Figure 4: In contrast-enhanced fat-suppressed coronal section T1-Weighted MR examination, the lesion showed contrast enhancement.



The patient underwent a wide resection surgery where a 2-cm margin of the extensor muscle group's fascia was removed (Figure 5). The resected area was then repaired using radiotherapy mapping and a fasciocutaneous flap. The excised tissue was preserved in a 10% formaldehyde solution and forwarded to the pathology lab.

Upon examination, the tissue specimen presented as a yellow-beige, homogeneous tumor without a capsule, measuring 15×10×8 mm with uneven yet distinct borders. Tumor samples were placed in cassettes for tracking and further embedded in paraffin blocks after routine procedures. These blocks were then sliced into 2-µm-thick sections for microscopic analysis. The stained samples displayed a tumor with misshapen, hyperchromatic nuclei, noticeable nucleoli, and sporadically forming a storiform pattern. There was a noticeable presence of giant tumor cells and mitotic cells (Figures 6, 7, and 8).

Figure 5: Pre-operative image.

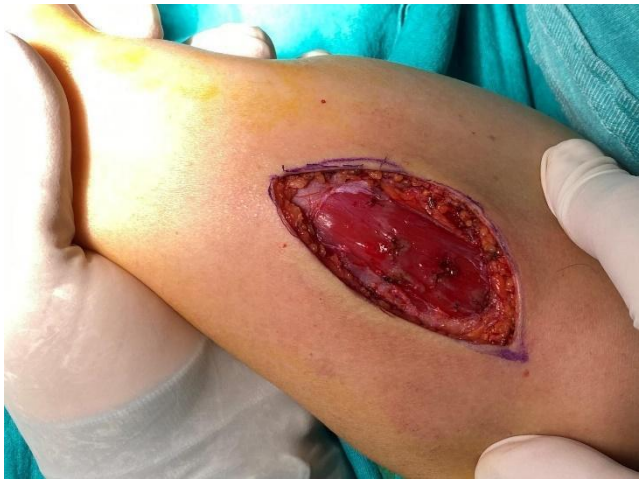


Figure 6: Tumor with distinct borders in subcutaneous tissue. Hematoxylin and eosin ×40.

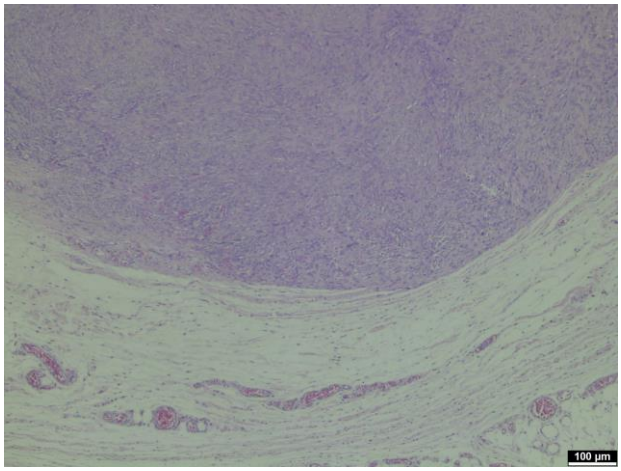


Figure 7: Pleomorphic tumor spindle and mitotic cells. Hematoxylin and eosin ×400.

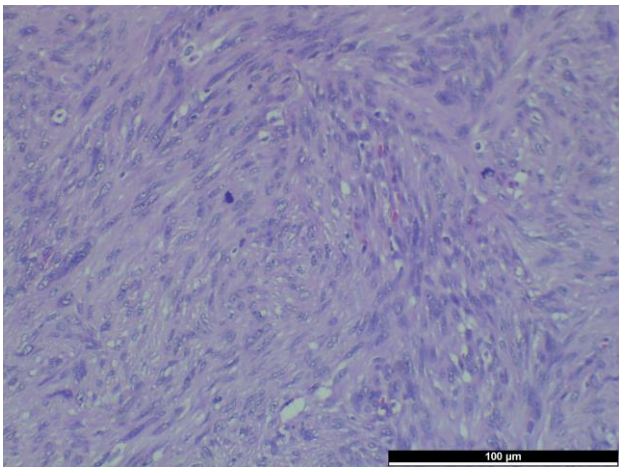
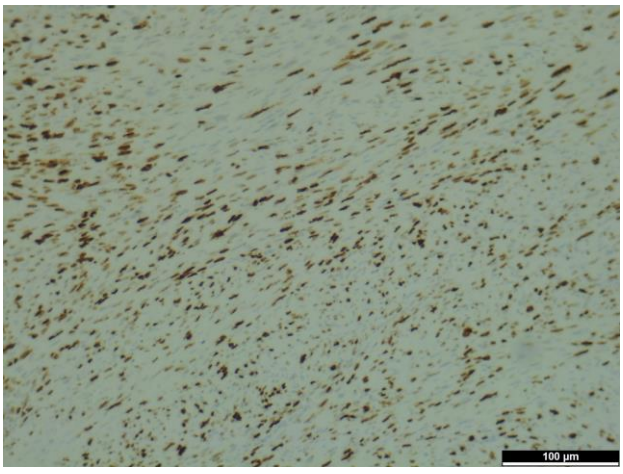


Figure 8: Positive nuclear staining in 35% of tumor cells with Ki-67 immunohistochemical staining.



The patient was diagnosed with MFH based on the findings. A pathological report showed a surgical margin of 5 mm to the deep fascia. Post-surgery, the patient received radiotherapy. After 6 months, a thorax CT scan took place. Ultrasonography (USG) was employed to check the surgical area, and the results showed no recurring lesions. A year later, a thorax CT scan and superficial tissue USG were performed with no signs of metastasis or recurring lesions. Currently, the patient is under regular observation.

Discussion

Soft tissue sarcomas (STS) are a heterogeneous category of malignant tumors encompassing various subgroups [1]. Gustafson's study featuring 508 patients in 1994 showed that the most prevalent subgroup was MFH [11]. However, a larger study indicated leiomyosarcoma as the most common STS, comprising 23.9% of cases, followed by MFH at 17.1%. In an epidemiological investigation focusing specifically on extremity-located soft tissue carcinomas, MFH proved to be the most dominant [1].

MFH symptoms can differ based on the tumor's size and location. Usually, a visible mass or lump appears in the tumor's area. There might be pain, tenderness, or discomfort in the affected zone. Some instances might show skin discoloration, ulcers, or surface bumps.

USG is pivotal in identifying soft tissue abnormalities and distinguishing between solid and fluid-filled masses. Doppler USG is valuable for evaluating the vascular nature of a lesion, differentiating potential vascular abnormalities. MFH often manifests as non-specific radiological features, forming masses of varied sizes. Although MRI also displays non-specific findings for MFH, MRI is favored for its high-definition resolution in diagnosing and conducting differential diagnosis. MRIs typically indicate low to medium intensity on T1-weighted images and high intensity on T2-weighted images. Enhanced imaging generally presents contrast enhancements. In terms of differential diagnosis, liposarcoma, rhabdomyosarcoma, and synovial sarcoma should be taken into account.

The prognosis in STS depends on factors such as tumor size, degree of invasion, and the level of differentiation. In this specific case, the tumor appeared solid and displayed minimal vascularization, noted through Doppler ultrasound.

MRI was instrumental in offering insights into the characteristics of the soft tissue lesion. Additionally, it enabled the differentiation of tumor-like processes, including benign lipomatous lesions, intramuscular hemangiomas, and hematomas. However, for evaluating calcifications, MRI is not sufficient.

In MRI , MFH generally appears as a hypointense lesion on T1-weighted images becomes hyperintense on T2-weighted images, and displays as a contrast-enhancing lesion in post-contrast images. Unfortunately, MRI lacks specific characteristic features that could suggest MFH.

MRI aids in understanding the spread of the tumor, both internally and externally, from the bone, and it also shows any involvement of nearby soft tissue, blood vessels, and nerves. Consistent with existing literature, the MRI findings in this case revealed no invasion into neighboring soft tissue, blood vessels or nerves.

MFH frequently manifests with vague radiological findings and can lead to variously sized masses. Adjacent bone often shows cortical erosion, a significant feature in radiography. Additionally, it can trigger periosteal responses and pathological fractures. Peripheral calcifications and ossifications are rarely detected in radiography. In our situation, the patient, who had previously had a lipoma removed from the forearm, presented with a reoccurrence of swelling. Consequently, direct radiography was not deemed necessary, and a preliminary superficial ultrasound was carried out instead.

The diagnosis and monitoring of patients often utilize fluor-18-labeled FDG PET/CT imaging. It is instrumental in staging, assessing treatment responses and prognosis, spotting relapse, and differentiating possible relapse from post-treatment alterations [13]. In our case, the PET/CT revealed FDG uptake in the recurrent malignant soft tissue mass, pointing to its malignant nature.

The primary treatment for MFH is surgery, particularly wide excision with a 1–2 cm margin of normal tissue [8]. It has been shown that wide surgical resection, including regional nodal dissection if needed, is the preferred method of treating MFH [14]. Optimal treatment involves surgical resection with negative margins, yet the median survival without distant metastasis is 8.5 months [2]. Recurrence can occur even with treatment [15]. In the case presented, the patient underwent wide surgical resection after recurrence, and no further occurrences were reported in the 12-month follow-up period after the surgery.

Neoadjuvant or adjuvant radiotherapy might be useful, as suggested by one study [16]. Another study indicates that radiation can be used for tumors that cannot be operated on, are larger than 5 cm in the extremities, or are surgically removed but with positive histological margins [14]. Due to recurrence in our patient, postoperative radiotherapy was administered.

Approximately 10% of cases may show metastases from the extremity, typically to the lung, or from the retroperitoneal region to the liver, at diagnosis time. Diagnostic methods such as CT, MR, PET-CT, and USG are necessary. While MRI adequately stages regional soft tissue tumors, PET-CT, bone scintigraphy, and a high-resolution CT scan of the lungs offer the best detection of metastatic spread [13]. There was no evidence of distant metastasis in our case study.

A precise diagnosis of MFH typically requires a blend of clinical, radiological, and pathological assessments. Biopsies and histopathological examinations of the tumor tissue, supplemented with immunohistochemistry, can distinguish between various sarcomas and other similar conditions. Consultation with an expert oncologist or pathologist is vital for determining the right diagnosis and suggesting suitable management. MFH was confirmed in our case based on the former diagnosis and histomorphological attributes. Within the surgical practice, it is known that many clinics still perform excisions based on initial diagnoses like lipoma or sebaceous cyst without employing radiological imaging. Conducting an excision after radiological imaging is advisable, particularly due to the potential presence of STS in seemingly harmless soft tissue tumors found beneath the skin.

References

1. Toro JR, Travis LB, Wu HJ, et al. Incidence patterns of soft tissue sarcomas, regardless of primary site, in the surveillance, epidemiology and end results program, 1978-2001: An analysis of 26,758 cases. Int J Cancer. 2006(15);119(12):2922-30.
2. Tong Y, Yu H, Shen B, et al. Primary hepatic malignant fibrous histiocytoma combined with invasion of inferior vena cava: A case report and literature review. Medicine (Baltimore). 2017;96(23):e7110.
3. Henderson MT, Hollmig ST. Malignant fibrous histiocytoma: changing perceptions and management challenges. J Am Acad Dermatol. 2012;67(6):1335-41.
4. Reshadi H, Rouhani A, Mohajerzadeh S, et al. Prevalence of malignant soft tissue tumors in extremities: an epidemiological study in Syria. Arch Bone Jt Surg. 2014;2(2):106-10.
5. Choi SH, Kim SY, Son MK, et al. Malignant fibrous histiocytoma of the maxillary sinus in a spray painter from an automobile repair shop. Ann Occup Environ Med. 2013(1);25(1):30.
6. Lambade PN, Lambade D, Saha TK. Malignant fibrous histiocytoma: an uncommon sarcoma with pathological fracture of mandible. J. Maxillofac. Oral. Surg. 2015;14:283–7.
7. Wu Y, Liu X, Lv Y, Li Z. Malignant fibrous histiocytoma of the floor of mouth: a case report and review of the literature. J Stomatol Oral Maxillofac Surg. 2022;123(3):e106-e111.
8. Pathak S, Soni TP, Gupta AK, Sharma LM. Retrovesical malignant fibrous histiocytoma: a rare tumor. BMJ Case Rep. 2017(2);2017:bcr2017222612.
9. Agha RA, Fowler AJ, Saeta A, et al. The SCARE Statement: Consensus-based surgical case report guidelines. Int J Surg. 2016;34:180-6.
10. Coindre JM, Terrier P, Bui NB, et al. Prognostic factors in adult patients with locally controlled soft tissue sarcoma. A study of 546 patients from the French Federation of Cancer Centers Sarcoma Group. J Clin Oncol. 1996;14(3):869-77.
11. Gustafson P. Soft tissue sarcoma: epidemiology and prognosis in 508 patients. Acta Orthopaedica Scandinavica. 1994;65(sup259):2-31.
12. Teng H, Xinghai Y, Wei H, et al. Malignant fibrous histiocytoma of the spine: a series of 13 clinical case reports and review of 17 published cases. Spine. 2011;36:1453–62.
13. De Schepper AM, De Beuckeleer L, Vandevenne J, Somville J. Magnetic resonance imaging of soft tissue tumors. Eur Radiol. 2000; 10(2):213-23.
14. Rothman AE, Lowitt MH, Pfau RG. Pediatric cutaneous malignant fibrous histiocytoma. J Am Acad Dermatol. 2000;42(2 Pt 2):371-3.
15. Mohan RP, Verma S, Siddhu VK, Agarwal N. Malignant fibrous histiocytoma. BMJ Case Rep. 2013 May 31;2013:bcr2013008875.
16. Seomangal K, Mahmoud N, McGrath JP. Malignant fibrous histiocytoma, now referred to as Undifferentiated Pleomorphic Sarcoma: A Case Report of an unexpected histology of a subcutaneous lesion. Int J Surg Case Rep. 2019;60:299-302.