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A unique case of inferior vena cava thrombosis in a patient with metastatic dorsal myxofibrosarcoma

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Abstract

Myxofibrosarcoma (MFS) is a sarcoma derived from fibroblasts, comprising approximately 5 to 10% of all malignant soft tissue tumors. MFS can occur at various ages but primarily affects the extremities of elderly individuals aged 60 to 80 years [1]. It typically presents in the extremities as a slow-growing, painless mass. MFS can also arise in the trunk and head and neck region [2]. The superficial fascia of the trunk or extremities is the primary site in 80 to 90% of MFS cases [1]. Despite chemotherapy, patients often exhibit a higher rate of distant metastases [3]. Here, we report a unique case of metastatic venous invasion involving the inferior vena cava extended to the right atrium, renal veins, and common iliac veins in a patient with dorsal myxofibrosarcoma.

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Introduction

Myxofibrosarcoma (MFS) is a malignant soft tissue tumor, derived from fibroblasts, comprising approximately 5 to 10% of all malignant soft tissue tumors [4]. It is characterized by the presence of myxoid (gel-like) and fibrous components and typically affects patients after the fifth decade of life [2]. MFS can occur at different ages but primarily affects the extremities of elderly individuals aged 60 to 80 years [1], often in the limbs but also rarely in the trunk, head, and neck. It exhibits a high rate of local recurrence despite repeated surgical resections with wide negative margins [4]. MFS is known for its infiltrative behavior and potential for metastasis [2].

Case

A 44-year-old female patient, followed for dorsal myofibrosarcoma and undergoing ifosfamide-based chemotherapy, was admitted to our department for a follow-up thoraco-abdomino-pelvic CT scan. Clinical examination finds an alteration in the general condition, with diffuse edema of both lower limbs.

Thoraco-abdomino-pelvic CT scan revealed endoluminal material within the Inferior Vena Cava (IVC), extending to the right atrium, renal veins and common iliac veins, showing tissue density and enhancement after injection of iodinated contrast material (Figure 1). A complementary abdominal ultrasound confirmed the tissue nature of the metastatic invasion of the IVC, also visualized on color Doppler (Figure 2). Subsequently, an abdominal MRI was performed, confirming the tumoral nature of the venous invasion with hyperintensity on T2-weighted sequences, restricted diffusion, and enhancement after gadolinium injection (Figure 3).

Discussion

Soft Tissue Sarcomas (STS) encompass a diverse group of malignant mesenchymal tumors, representing only 1% of all adult cancers [5]. Among them, liposarcoma and leiomyosarcoma are the most common, with myxofibrosarcoma (MFS) affecting approximately 20% of sarcoma patients [6]. These tumors are characterized by slow growth but demonstrate a strong tendency for local and distant recurrence [6]. They exhibit significant

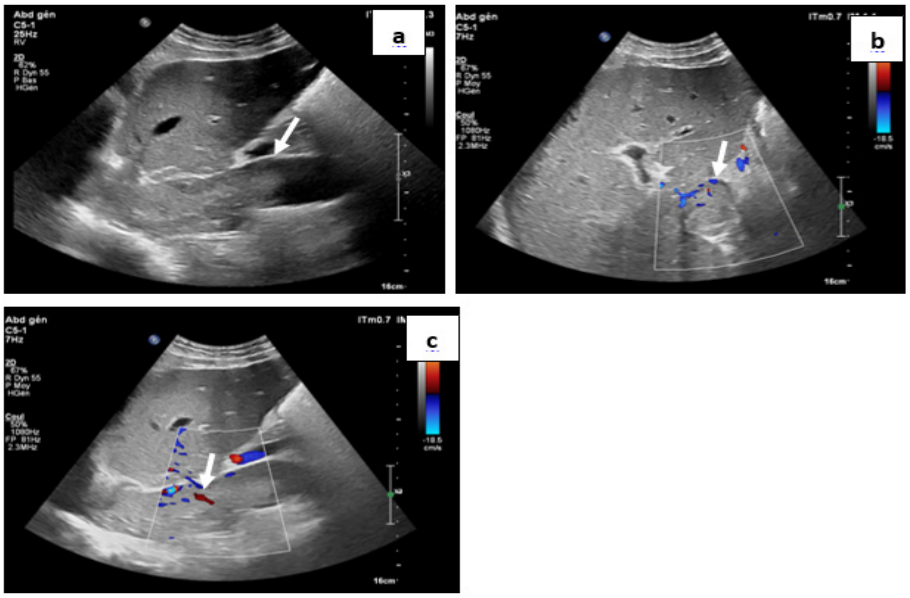

Figure 1: Abdominal ultrasound revealing extensive tumor thrombosis of the inferior vena cava **(a)** with color Doppler flow **(b,c)**.

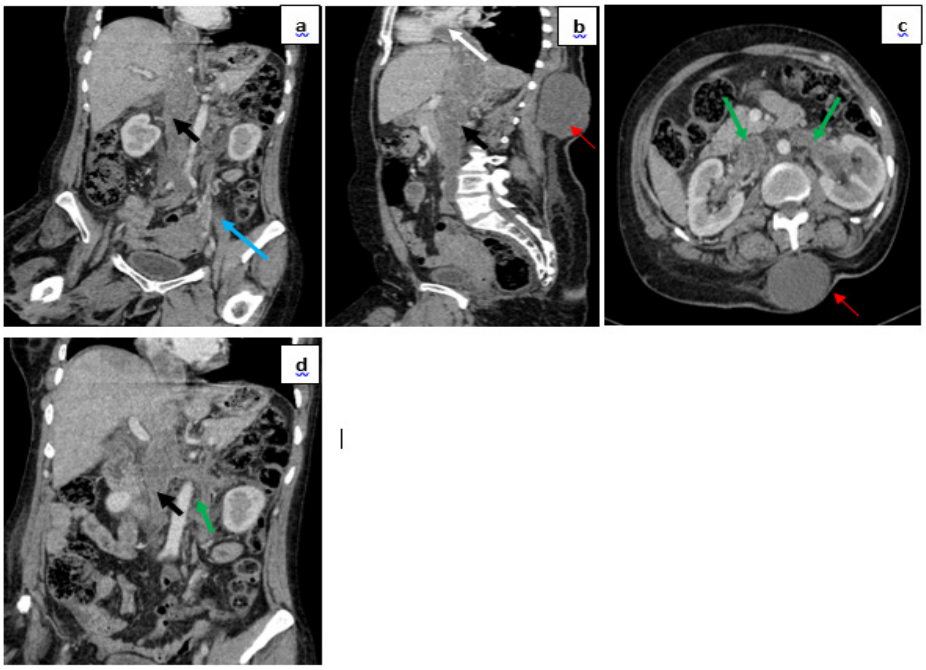

Figure 2: Contrast-enhanced thoraco-abdomino-pelvic CT scan revealing intraluminal material within the Inferior Vena Cava (IVC) (Black arrow) **(a,b,d)**, extended to the right atrium (White arrow) (b), renal veins (Green arrow) **(c,d)**, and common iliac veins (a) (Blue arrow).

Note: Subcutaneous tissue mass at the level of D12, oval-shaped, well-defined, measuring 96x56 mm, consistent with dorsal myxofibrosarcoma (Red Arrow).

histological diversity, with over 100 subtypes and a wide range of clinical presentations [6].

Previously considered a myxoid variant of Malignant Fibrous Histiocytoma (MFH), MFS was reclassified as a distinct entity by the WHO in 2002. It is characterized by a myxoid stroma, cellular pleomorphism, curvilinear vessels, and a high rate of local recurrence [4]. While these tumors predominantly arise in the lower limbs of elderly individuals, they can also occur in other parts of the body such as the upper limbs, trunk, head, neck, hands, and feet, albeit less commonly [5].

MFS primarily affects patients aged 60 to 70 years, with a slight male predominance [4]. Clinically, it typically presents as a painless, slow-growing mass [3]. High-grade forms are associated with an increased risk of metastasis, with 15% to 38% of local recurrences progressing to higher histological grades and increased metastatic potential [5].

MFS is known for its infiltrative behavior and high metastatic potential [2]. To our knowledge, the case we report here is the first to document direct invasion of the Inferior Vena Cava (IVC) with extension into its branches. While MFS rarely involves large vessels, cases of vascular metastases have been reported in the literature [4]. Song J et al. reported a case of superior vena cava vascular invasion in a patient with myxofibrosarcoma [4].

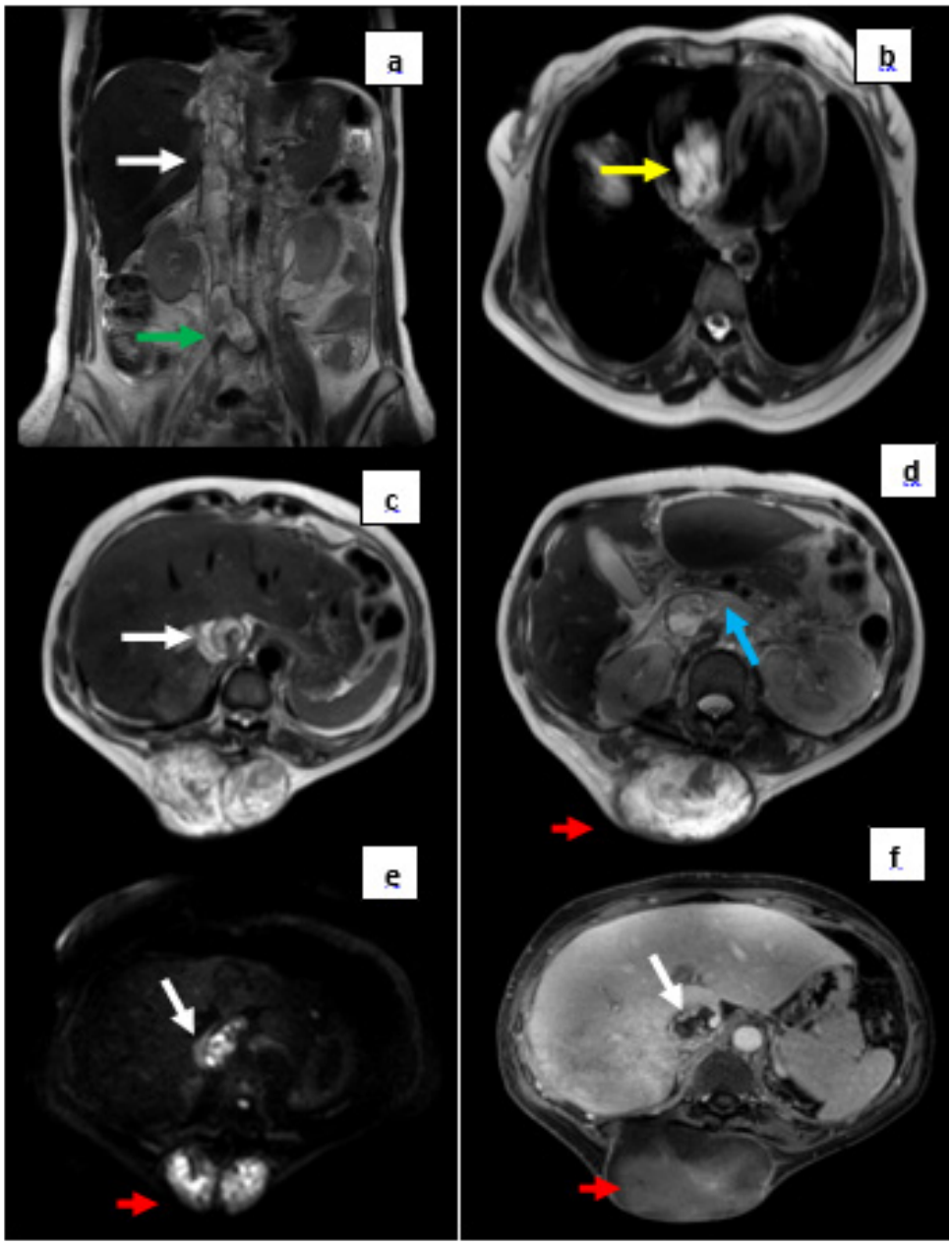

Figure 3: Abdominal MRI revealed tumoral venous invasion of the Inferior Vena Cava (IVC) (white arrows) **(a)** extending to the right atrium (Yellow arrow) **(b)**, renal veins (Blue arrow) **(d)**, and common iliac veins (Green arrow) **(a)**, with hypersignal on T2-weighted imaging **(a,b,c,d)**, restricted diffusion **(e)**, and moderate enhancement after Gadolinium injection **(f)**. The dorsal myxofibrosarcoma appeared as hypersignal on T2-weighted imaging, restricted diffusion, and showed moderate enhancement after Gadolinium injection (Red arrow).

The role of chemotherapy in managing metastatic MFS remains controversial, with mixed results despite the use of anthracycline-based regimens as first-line treatment. Second-line chemotherapy, including ifosfamide, pazopanib, gemcitabine, and docetaxel, shows modest response rates around 10% [2]. It is also noteworthy that some patients undergoing chemotherapy have an increased risk of distant metastases despite treatment [3].

Conclusion

Myxofibrosarcoma (MFS) is a malignant soft tissue tumor characterized by notable histological diversity, high potential for local recurrences and metastases, and significant therapeutic challenges, particularly concerning chemotherapy. Our case is particularly exceptional due to the invasion of the inferior vena cava, underscoring the rarity and complexity of this disease.

Declarations

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References

1. Liu W, Yan Y, Wu X, Hou X, Qu X. Myxofibrosarcoma involving brachial plexus diagnoses by contrast-enhanced ultrasound: A case report. Medicine (Baltimore). 2023; 102(50): e36626. doi: 10.1097/MD.0000000000036626.

2. Lucarelli E, De Vita A, Bellotti C, Frisoni T, Vanni S, et al. Modeling Myxofibrosarcoma: Where Do We Stand and What Is Missing? Cancers (Basel). 2023; 15(21): 5132. doi: 10.3390/can-

cers15215132.

3. H Engellau J, Ghanei I, Vult von Steyern F, Styring E. High Recurrence Rate of Myxofibrosarcoma: The Effect of Radiotherapy Is Not Clear. Sarcoma. 2019; 2019: 8517371. doi: 10.1155/2019/8517371.

4. Song J, McDermott VM, Ryan P, et Bird, BRH. Myxofibrosarcome envahissant directement la veine cave supérieure provoquant une obstruction soulagée avec succès par un stent endovasculaire. Acta Oncologica. 2014; 54(1): 141-143. https://doi.org/10.3109/0284186X.2014.953261

5. Vanni S, De Vita A, Gurrieri L, Fausti V, Miserocchi G, et al. Myxofibrosarcoma landscape: diagnostic pitfalls, clinical management and future perspectives. Ther Adv Med Oncol. 2022; 14: 17588359221093973. doi: 10.1177/17588359221093973.

6. Leonhardt LP, Pervez A, Tang W, Amen M. A Unique Presentation of Pulmonary Vein Thrombosis in a Patient With Rapidly Progressive Metastatic Pelvic Myxofibrosarcoma. Cureus. 2022; 14(9): e28887. doi: 10.7759/cureus.28887.