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CASE REPORT

BEGINNER

CLINICAL CASE

De-Differentiated Myxofibrosarcoma Metastasis in the Right Atrium

An Invasive But Diagnostic Approach

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ABSTRACT

Solitary cardiac metastasis remains an uncommon diagnosis. Herein, the authors report describes a rare case of a 53-year-old woman with cardiac metastasis of a peripheral de-differentiated myxofibrosarcoma. This case demonstrates the complexity of pairing multimodality imaging and invasive techniques to achieve tissue characterization and diagnosis. (**Level of Difficulty: Beginner.**) (J Am Coll Cardiol Case Rep 2019;1:151–5) © 2019 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

Myxofibrosarcoma was first described in 1977 as the most common soft tissue sarcoma arising in the limbs of older adult patients, with a slight male predominance (1). It is a connective tissue neoplasm of fibroblastic origin in a myxoid matrix with a high recurrence rate (2,3). Description of this tumor in the literature reported that the most common sites for metastases were the lymph nodes and lungs (4). However, no case reports to date have described solitary metastasis to the heart from peripheral myxofibrosarcoma lesions.

We present the first case report of metastasis of peripheral myxofibrosarcoma to the heart of a 53-year-old woman with equivocal noninvasive and invasive diagnostic tests, in whom the final diagnosis was made through exploratory surgery.

LEARNING OBJECTIVES

- Cardiac metastases are underdiagnosed and associated with diagnostic challenges that require multimodality investigations. The index of suspicion should be raised for intracavity myocardial lesions in the context of a history of malignancy.
- Despite surgical resection being the gold standard diagnostic and therapeutic approach to solitary cardiac masses, complete resolution of metastatic disease remains challenging despite advances in therapeutic options.
- In the absence of further case reports, we conclude that the long-term prognosis and recurrence rates of solitary metastatic cardiac myxofibrosarcoma remains unknown.

HISTORY OF PRESENTATION AND MEDICAL HISTORY

A 53-year-old woman presented to our emergency unit with dyspnea (New York Heart Association functional Class III) and severe stabbing retrosternal pain. Her medical history included excision of a myxofibrosarcoma in her right lower leg 3 years prior; after surgery she had radiotherapy immediately not during follow-up. It was part of the initial management of her leg lesion. Therefore, radiotherapy was initiated

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ABBREVIATIONS AND ACRONYMS

CMR = cardiac magnetic resonance imaging

FDG = fluorodeoxyglucose

MDT = multidisciplinary team

NYHA = New York Heart Association

PET-CT = positron emission tomography–computed tomography scan

SVC = superior vena cava

TEE = transesophageal echocardiography

TTE = transthoracic echocardiography

immediately after surgical excision. And during follow-up no recurrence of malignancy was seen.

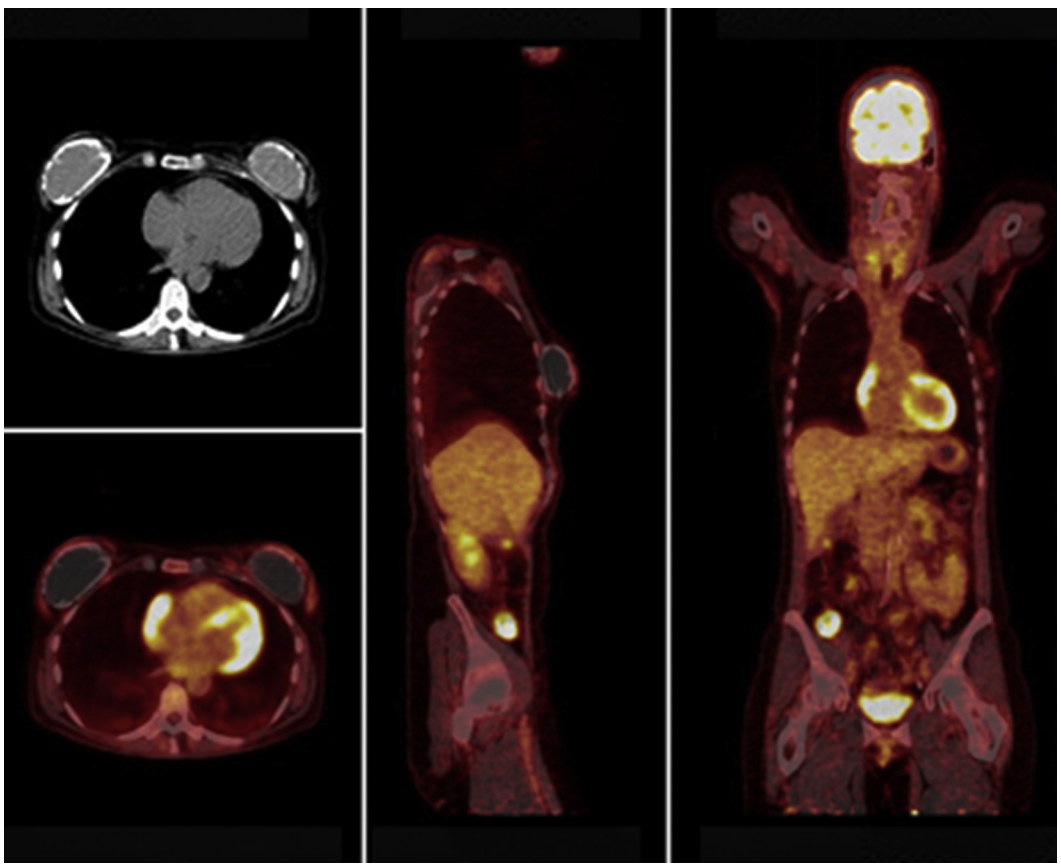
INVESTIGATIONS

Urgent workup in the emergency unit was carried out. Transthoracic echocardiography (TTE) revealed cardiac tamponade, which prompted emergency percutaneous pericardiocentesis. This resulted in aspiration of 340 ml of hemorrhagic pericardial fluid. Molecular analysis of this pericardial fluid marked a presence of T-lymphocytes (positive CD3, BCL2, and CD5) with a nonsignificant amount of B-lymphocytes (positive CD20), which concluded that the collection had evidence of an inflammatory rather than a malignant process.

Nevertheless, because of the history of malignancy, we proceeded to a positron emission tomography/computed tomography (PET-CT) scan to investigate metastatic spread. This outlined areas of hypermetabolic uptake within the hepatic flexure of the colon, cecum, and a prominent uptake against the right atrium (Figure 1). This was a new finding compared with previous PET-CT scans. The lesions in the colon and cecum were resected and diagnosed as tubulovillous adenomas with high-grade dysplasia, which meant that the nature of the cardiac lesion remained unknown.

Next, we proceeded to cardiac magnetic resonance, which revealed a 5.5 cm by 6.0 cm nonhomogenous mass spreading from the right atrial appendage to the superior vena cava (SVC). The mass had an intermediate nonhomogenous signal on T1-weighted images and a low signal intensity on T2-weighted turbo spin-echo images with abnormal gadolinium uptake (Figure 2). In light of this, we pursued a transvenous endomyocardial biopsy of the tumor under transesophageal echocardiography (TEE) guidance. This

FIGURE 1 Initial PET-CT Scan: Patient Injected with 7-mCi FDG Intravenously



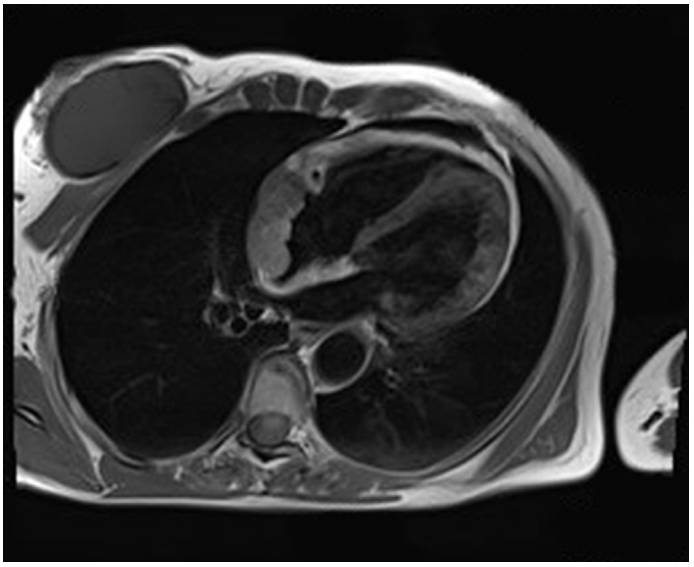
Transverse, sagittal, and coronal views revealed suspicious hypermetabolic uptake within the hepatic flexure of the colon, cecum, and a prominent uptake against the right atrium. FDG = fluorodeoxyglucose; PET-CT = positron emission tomography/computed tomography.

was nondiagnostic, with microscopic analysis showing an increase of fibrous strands between cardiomyocytes with an increased size and irregularities in the nucleus, but no specific diagnostic features.

MANAGEMENT

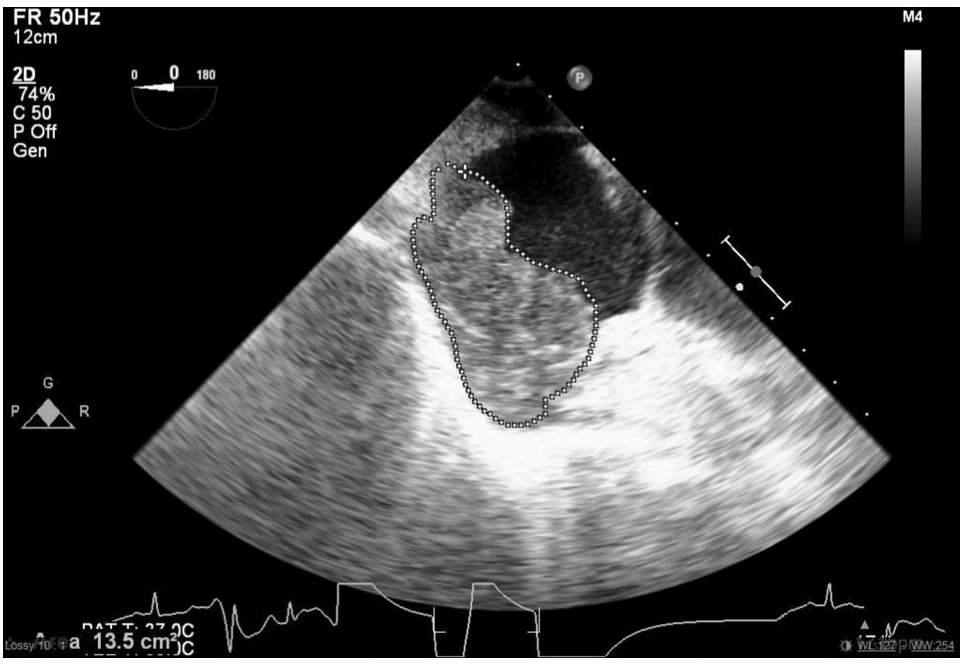
The case was reviewed during the multidisciplinary team (MDT) meeting. Because of the tumor extension, proximity to local vascular structures and the associated hemorrhagic pericardial effusion, malignancy remained a high clinical probability. Therefore, the MDT consensus recommended an invasive resection biopsy, which the patient agreed to. This was performed via traditional open surgery guided by TEE (Figure 3). Surgery revealed a large (5.5 cm diameter) lobulated mass invading into the right atrial free wall. Intraoperative frozen section was carried out and confirmed the malignant nature of the mass. Consequently, an incision from the right atrium to the base of the SVC was performed to excise the mass. Although no tumor infiltration occurred in the nearby valve, a tricuspid valve annuloplasty was performed to establish macroscopically tumor-free margins. Post-operative TTE revealed no tumor remnant.

FIGURE 2 Cardiac Magnetic Resonance Imaging



Nonhomogenous mass (size 5.5 X 6.0 cm) extending from the right atrial appendage to the superior vena cava. The mass shows an intermediate nonhomogenous signal strength on T1-weighted TSE images, low signal intensity on T2-weighted turbo spin-echo images and post-IV gadolinium becomes nonhomogenous.

FIGURE 3 Intraoperative TEE of the Right Atrial Tumor



A modified bi-caval view revealed a large (5.5 cm diameter) lobulated mass invading into the right atrial free wall, with no flow obstruction or tricuspid valve invasion. TEE = transesophageal echocardiography.

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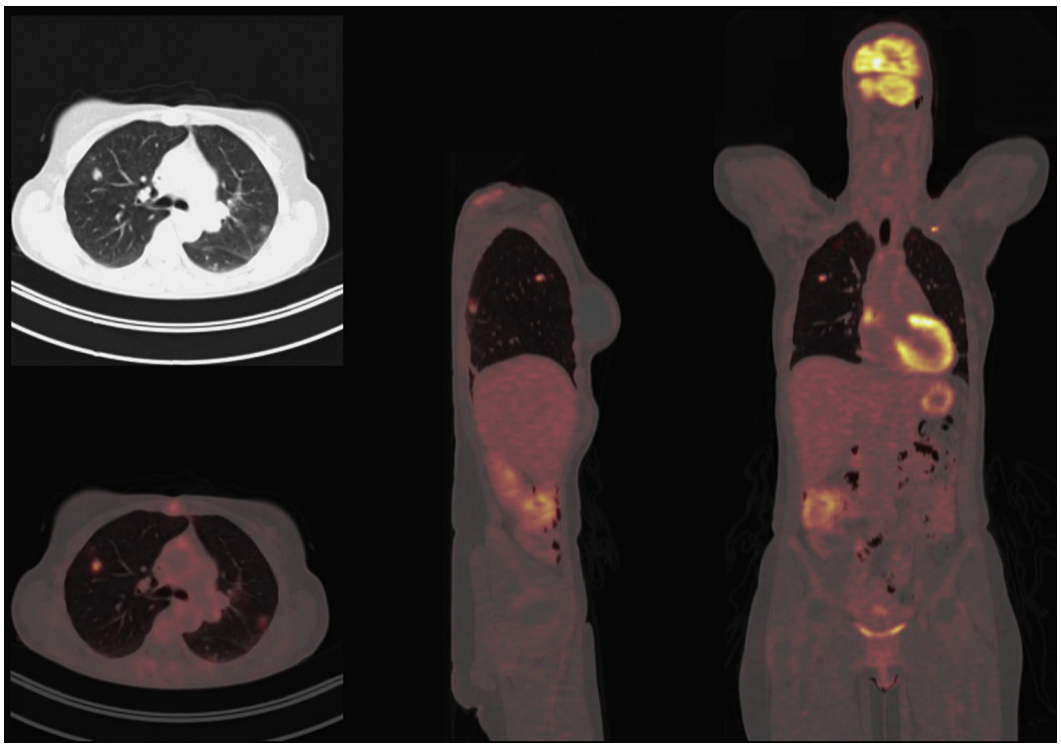
FIGURE 4 Follow-Up PET-CT



Patient was injected with 7-mCi FDG intravenously. Transverse, sagittal, and coronal views revealed new metastatic lesions in the pericardium and lungs, and a decrease in hypermetabolic mass in the right atrium. Abbreviations as in **Figure 1**.

The final anatomopathological diagnosis reported a malignant solitary fibrous tumor in the right atrium extending to the atria-ventricular groove. This tumor was probably linked to the de-differentiated myxofibrosarcoma of the right lower leg.

FOLLOW-UP

Because of the clear macroscopic margins and the solitary nature of the mass, a watch-and-wait approach was pursued. Unfortunately, however, this period was complicated by multiple admissions for dyspnea, splenic rupture, and stroke; a follow-up PET-CT 6 months after the excision confirmed recurrence of the metastatic lesions in the pericardium, lungs, and the right ventricle (**Figure 4**). Unfortunately, the patient died 16 months after explorative cardiac surgery.

DISCUSSION

Cardiac metastasis remains underestimated and discovered in up to 25% post-mortem examinations of patients diagnosed with malignancies (5,6). It is associated with poorer outcomes compared with benign cardiac tumors (7,8). In this case report, attempts at reaching the diagnosis with noninvasive and/or minimally invasive investigations were limited. This could be explained by the development of the de-differentiated myxofibrosarcoma into high-grade myxofibrosarcoma, which sheds its typical cytological characteristics, making it hard to diagnose, as seen in this case (1,6).

The literature on myxofibrosarcoma involving the heart is limited to a few isolated case reports (7), with none describing metastasis of peripheral lesions to the myocardium. Cases of primary cardiac myxofibrosarcoma were described to present with dyspnea, even without a pericardial effusion (7). Studies that described distant metastatic myxofibrosarcoma (to noncardiac tissue) recommended aggressive treatment with chemotherapy (4). However, management of these cases remains challenging due to the unpredictable course of these lesions and the increased recurrence rate and poor prognosis, despite excision with clear margins (9). This mirrored our findings in the case report described.

CONCLUSIONS

We present the first case of solitary metastatic spread of myxofibrosarcoma to the heart. Despite advances in multimodality investigations and increasing treatment options, the case presented a number of diagnostic and management challenges. Further translational studies are warranted to improve the diagnostic yield and choice of therapy.

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