

Metastatic Sarcoma Affecting the Heart and Presenting as Infective Endocarditis: About a Single Medical Case

Feres Mansour*, Sameh Ben Farhat, Aymen El-Hraiech, Sami Ouanes, Mehdi Slim, Rym Gribaa, Elyes Neffati

Department of Cardiology, Sahloul University Hospital, Sousse, Tunisia

ABSTRACT

Both primary and metastatic heart sarcomas are scarce neoplasms with various clinical features and are known to be great imitators of several clinical representations. This article presents a case of a cardiac metastatic sarcoma complicated by infective endocarditis in a 35-year-old female patient with a history of rheumatic fever, brain surgery and COVID-19 infection. The diagnosis was based on echocardiography, computed tomography and biopsy results. Despite receiving six weeks of antibiotic treatment, the patient's prognosis was bleak due to the lack of effective palliative options. The article underlines the challenges in the management of cardiac sarcomas and the need for further research in this field.

Keywords: Metastatic cardiac sarcoma; Infective endocarditis; Diagnosis; Prognosis

Abbreviations: CD: Cluster of Differentiation; CMR: Contrast-enhanced Cardiac Magnetic Resonance; COVID-19: Coronavirus 2019; CRP: C-Reactive Protein; CT: Computed Tomography; ECG: Electrocardiography; e.g.: For example; ESC: European Society of Cardiology; IE: Infective Endocarditis; MRI: Magnetic Resonance Imaging; PanCK: Pan Cytokeratin; PCR: Polymerase Chain Reaction; PET: Positron Emission Tomography; PET-CT: Positron Emission Tomography-Computed Tomography; STIR: Short-TI Inversion Recovery; TTE: Transthoracic Echography; 18F-FDG: 18-Fluorodeoxyglucose

INTRODUCTION

Primary cardiac tumors are exceedingly rare, with a prevalence ranging from 0.001% to 0.03%, as indicated by post-mortem studies [1,2]. The majority of cardiac tumors are metastatic, outnumbering primary tumors by a factor of 20 to 40 [3]. Malignant primary cardiac tumors constitute approximately 10%, with sarcomas accounting for 95% of these cases [4]. These sarcomas exhibit diverse histologies, with angiosarcomas (37%), undifferentiated sarcomas (24%), malignant fibrous histiocytomas (11%-24%), leiomyosarcomas (8%-9%) and osteosarcomas (3%-9%) being the most prevalent.

Typically, asymptomatic until reaching a significant size that impacts hemodynamic status [4], primary cardiac tumors may present with a variety of symptoms, including the classic triad: Cardiovascular symptoms (for example (e.g.), dyspnea) due to

intracardiac obstruction, functional angina or syncope, signs of systemic embolization (such as stroke) and systemic symptoms indicative of neoplastic disease (e.g., general malaise, asthenia, widespread unexplained pain and anorexia) [5]. Metastatic lesions exhibit rapid proliferation, significantly affecting clinical prognosis (median survival is 5 months with metastasis and 15 months without) [6].

Despite advancements in diagnostic tools like transthoracic echocardiograms, Computed Tomography (CT) scans and cardiac Magnetic Resonance Imaging (MRI), the prognosis for malignant cardiac sarcoma remains bleak due to the rarity of these tumors, hindering the establishment of a standardized treatment approach [1,2]. Consequently, a multidisciplinary approach involving oncologists, cardiologists and radiologists is crucial for the accurate diagnosis and management of cardiac metastasis.

Correspondence to: Feres Mansour, Department of Cardiology, Sahloul University Hospital, Sousse, Tunisia; E-mail: feres121.Mansour@hotmail.com

Received: 26-Jan-2024, Manuscript No. JCEC-24-29353; **Editor assigned:** 29-Jan-2024, PreQC No. JCEC-24-29353 (PQ); **Reviewed:** 12-Feb-2024, QC No. JCEC-24-29353; **Revised:** 18-Feb-2025, Manuscript No. JCEC-24-29353 (R); **Published:** 25-Feb-2025, DOI: 10.35248/2155-9880.25.16.972

Citation: Mansour F, Farhat SB, El-Hraiech A, Ouanes S, Slim M, Gribaa R, et al. (2025) Metastatic Sarcoma Affecting the Heart and Presenting as Infective Endocarditis: About a Single Medical Case. J Clin Exp Cardiol. 16:972.

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We report a case involving a young woman diagnosed with metastatic myocardial sarcoma complicated by infective endocarditis, emphasizing the pivotal role of cardiac imaging in diagnosis and guiding treatment strategies, whether they involve cardiac surgery or palliative care.

CASE PRESENTATION

This case report outlines the medical history and current condition of a 35-year-old female patient with a longstanding history of rheumatic fever dating back to childhood, coupled with a history of inconsistent follow-up. Approximately one month ago, the patient underwent surgery to address a right frontal tumor and subsequently faced postoperative complications attributed to Coronavirus 2019 (COVID-19) pneumopathy. Following a 12-day treatment course, she displayed a positive clinical trajectory with a negative Polymerase Chain Reaction (PCR) control for COVID-19.

The patient presented to our emergency department with a fever and diffuse pain, without any accompanying signs. Upon clinical examination, the patient exhibited normal blood pressure (100 mmHg for systolic and 65 mmHg for diastolic), regular tachycardia (heart rate=100 beats per minute), a systolic murmur localized at the mitral valve site and fever reaches 38.5°C. Notably, no other abnormalities were detected, including adenopathy, thyroid goiter, lumbar tenderness, signs of heart failure and pericardial friction and the anterior surgical scar remained clean with no signs of pus discharge. Electrocardiography (ECG) revealed regular sinus tachycardia with no conduction or repolarization abnormalities.

Biological tests disclosed normocytic normochromic anaemia (haemoglobin level=7.8 g/dl), moderate thrombocytosis (platelets=517,000/ μ l) and elevated inflammatory markers (C-Reactive Protein (CRP)=144 mg/l, procalcitonin=0.09 ng/ml).

According to ultrasonography conducted in our department, the left ventricle is moderately dilated with good contractile function and a preserved ejection fraction of 68%. A moderately thickened mitral valve is seen, along with thickening of the subvalvular apparatus and retraction of the small valve, resulting in pseudoprolapse of the large mitral valve. Notably, there is severe mitral insufficiency, with 0.4 cm² of regurgitant orifice area and 87 ml of regurgitant volume. A large vegetation, measuring 18 × 18 mm, is identified at the expense of the cordage of the large mitral valve. Additionally, substantial, highly mobile vegetation is observed on the atrial side of the large mitral valve, conferring a high risk of embolic complications. Furthermore, examination reveals that the aortic valve is thin and tricuspid, with no signs of valvulopathy. When the focus shifts to the right ventricle, longitudinal contractile function is preserved and there is no evidence of pulmonary hypertension or dry pericardium (Figures 1-3).

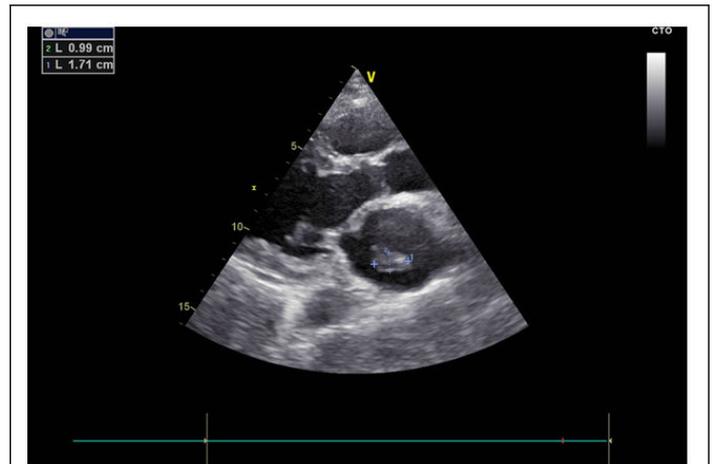


Figure 1: The large vegetation depending on the atrial side of the anterior leaflet of mitral valve viewed on sonographic cardiac examination.

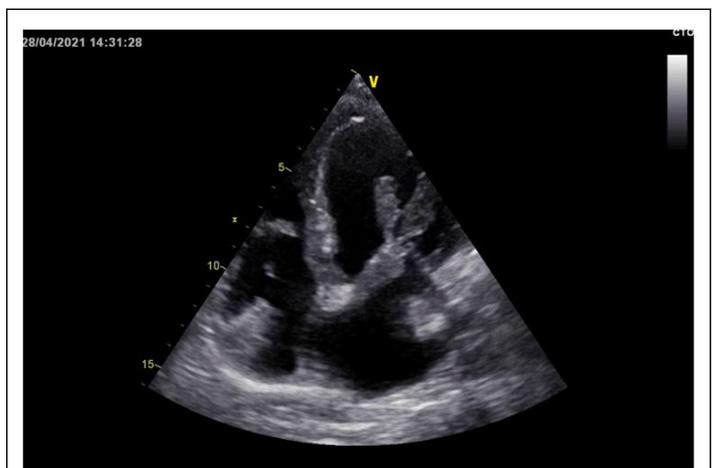


Figure 2: A large vegetation depending on the chordae tendineae attached to the anterior leaflet of the mitral valve shown on cardiac ultrasound.

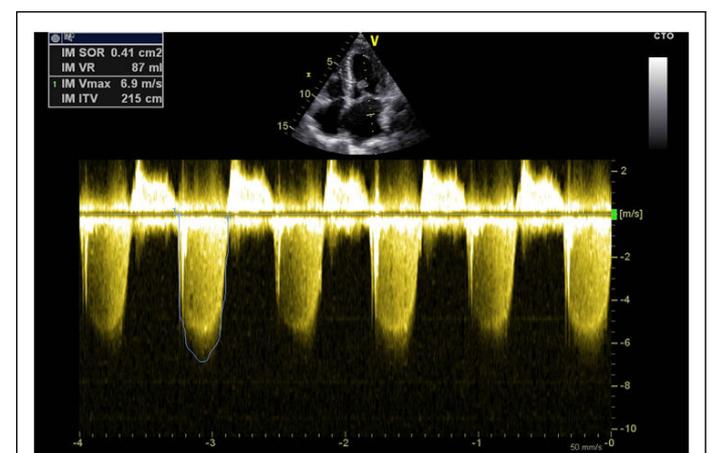


Figure 3: Severe mitral insufficiency shown on cardiac ultrasound with a calculated regurgitant volume at 87 ml and a regurgitant orifice area estimated at 40 cm².

A CT scan of the thoraco-abdomino-pelvic region conducted at a private institution identified secondary bone lesions and a suspicious lesion on the upper pole of the left kidney. It is noteworthy that scars suggestive of a COVID-19 pneumopathy

were identified. Hypodense images were also noticed in the left ventricle, on the side of the mitral valve and in the left atrium, raising the suspicion of possible secondary cardiac lesions (Figure 4).

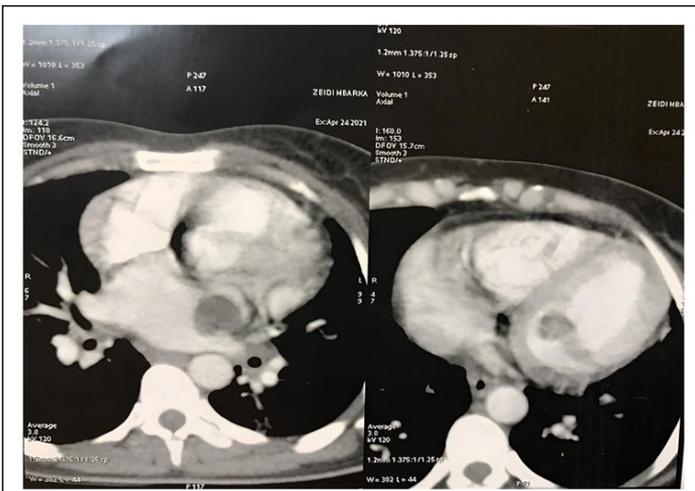


Figure 4: Hypodense images in the left ventricle near the mitral valve and in the left atrium shown on thoracic CT scan.

In the follow-up cerebral CT scan conducted during the patient's hospitalization, postoperative changes were observed in the right frontal bone. There was a lytic lesion with well-defined boundaries in the odontoid, along with a punched-out bone defect in the vault.

As part of the exhaustive staging evaluation of her neoplastic disease, we performed additional workups. A cervical ultrasound revealed a normal thyroid. An uro-CT scan, focused on the thoracic and abdomino-pelvic regions, showed diffuse septal thickening, masses in the left atrium (Figure 5) and lesions in both kidneys. The kidney's most prominent lesion, in the left upper pole, was suggestive of infarction, while multiple osteolytic lesions, particularly in the mid-shaft of the right 10th rib, seemed to be secondary to bone involvement. Hence, bone scintigraphy was performed, which confirmed a pattern consistent with axial and peripheral polymetastatic bone lesions (Figure 6).

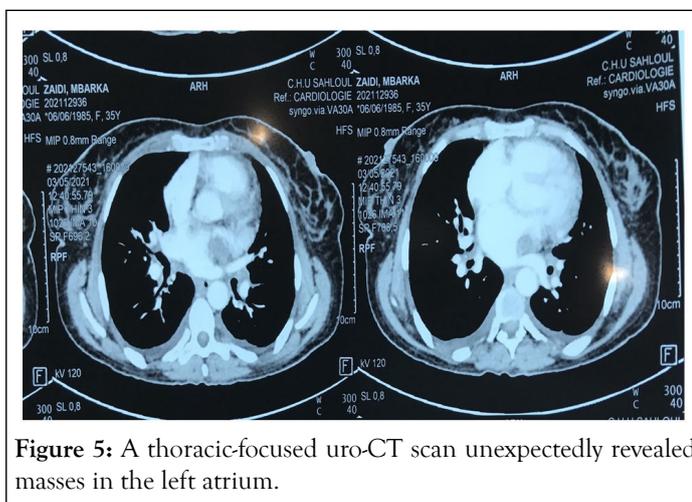


Figure 5: A thoracic-focused uro-CT scan unexpectedly revealed masses in the left atrium.

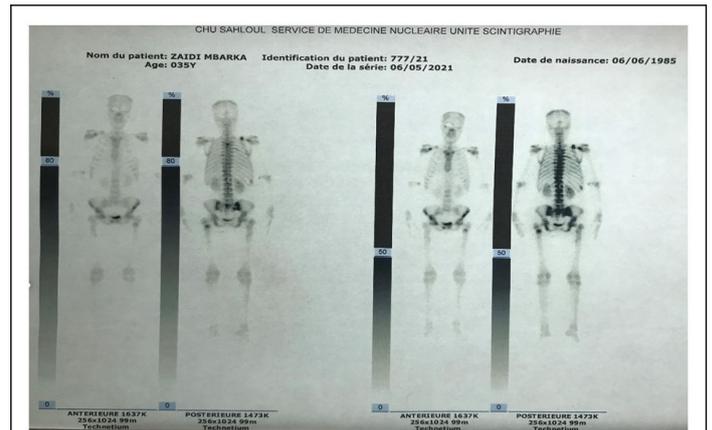


Figure 6: Bone scintigraphy revealing axial and peripheral polymetastatic bone lesions.

Thus, the initial therapeutic approach involved conducting a series of blood cultures, all yielding negative results. Following the guidelines of the European Society of Cardiology (ESC), an empirical triple antibiotic regimen was initiated, encompassing daily gentamicin at 240 mg, ampicillin at 2 g every 6 hours and oxacillin at 2 g every 6 hours. Nevertheless, with the persistence of fever and a subsequent increase in CRP levels to 165 mg/l after 72 hours of antibiotic administration, adjustments were made to the initial antibiotic protocol. The combination of oxacillin and ampicillin was replaced with vancomycin, administered at a dose of 1 g every 8 hours for one month, in conjunction with imipenem at a dose of 1 g every 8 hours for two weeks. The administration of gentamicin was upheld for a specific two-week period.

Three endocardial masses, roughly spherical in shape with well-defined contours, showed isosignal T1 and hypersignal T2 Short-TI Inversion Recovery (STIR) on cardiac MRI, with no post-gadolinium injection enhancement. These masses appeared immobile relative to valvular movements, suggesting the presence of thrombi within the left atrium and left ventricle. The distribution is as follows: Two within the left atrium, measuring 23 mm and 16 mm in the major axis and one in the left ventricle adjacent to the mitral cordage, measuring 18 mm in the major axis. These findings are concomitant with thickening of the anterior aspect of the mitral valve, likely indicative of a vegetative lesion.

Positron Emission Tomography (PET) with 18-Fluorodeoxyglucose (18F-FDG) was conducted to comprehensively assess lesion dissemination, revealing numerous intensely hypermetabolic foci indicative of malignancy throughout the skeletal structure. Additionally, heightened metabolic activity was observed in the liver's segment VII and the pancreatic tail. The more superficial lesions included the left iliac crest and costal parietal lesions, notably the lateral arch of the 10th right rib and the anterior arch of the 7th left rib. These findings were associated with multifocal hypermetabolism in the cardiac region, consistent with the lesions described in the cardiac MRI (Figure 7).

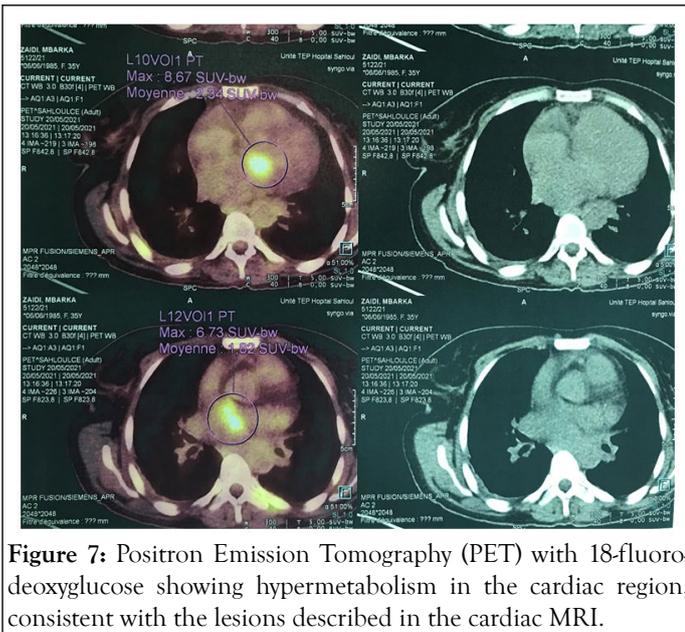


Figure 7: Positron Emission Tomography (PET) with 18-fluorodeoxyglucose showing hypermetabolism in the cardiac region, consistent with the lesions described in the cardiac MRI.

The anatomopathological examination of the ultrasound-guided biopsy from the 10th right rib reveals a prominently necrotic tumor proliferation within the sampled fragment. The tumor cells display characteristics of moderate-sized, ovoid morphology, with eosinophilic cytoplasm and irregular hyperchromatic nuclei. Mitotic figures and mito-necroses are evident. Immunohistochemical analysis demonstrates negative staining for pan Cytokeratin (panCK), Cluster of Differentiation (CD) 3, CD 20, CD 99, myogenin and the presence of desmin in only a few scattered cells. The morphological features strongly suggest a round cell sarcoma.

Therefore, a multidisciplinary consultation involving cardiologists from our department and cardiovascular surgeons determined a palliative approach for the condition, opting against surgical therapeutic interventions due to the advanced stage of the neoplastic disease.

RESULTS AND DISCUSSION

The manuscript "Metastatic sarcoma affecting the heart and presenting as infective endocarditis: A case report" presents a captivating and albeit perplexing clinical scenario. This case report not only highlights the intricate diagnostic masquerade orchestrated by certain malignancies but also reiterates the crucial role of clinical suspicion, comprehensive investigations and a multidisciplinary approach in unmasking such enigmas. Cardiac tumors represent a rare and diverse category of neoplasms, capable of affecting any cardiac structure [5]. Among them, sarcomas constitute a predominant group characterized by a diverse range of histologies [2]. The typical age at presentation is roughly 40 [7]. Despite their clinical significance, the available literature on cardiac sarcomas is limited, primarily consisting of isolated case reports detailing single-patient experiences. The intricate nature of these tumors, coupled with their rarity, contributes to the scarcity of comprehensive studies. Notably, primary and metastatic cardiac osteosarcomas are exceptionally rare and the collective documented instances of antemortem diagnoses number fewer than 100 cases [8]. The limited

prevalence and diverse characteristics of cardiac sarcomas underscore the need for continued research and collaborative efforts to enhance our understanding of these unique malignancies.

The initial presentation of fever seamlessly aligning with infective endocarditis is a testament to the uncanny ability of metastatic sarcomas to masquerade as infectious processes. This phenomenon, albeit documented, underscores the importance of considering alternative etiologies even in the face of seemingly classic presentations [4]. The clinical presentation of metastatic sarcoma in the heart exhibits significant heterogeneity, reflecting the tumor's location and extent [5]. Four main mechanisms contribute to the diverse symptomatology [5,9]:

Valve dysfunction: Disruption of cardiac valve function manifests as dyspnea (60%), fatigue and congestive heart failure (28%).

Local invasion: Tumor invasion can lead to arrhythmias and pericardial effusion, presenting with chest pain (24%), palpitations (24%) and pericardial discomfort.

Systemic thromboembolic events: Embolic complications cause stroke, vision loss, limb pain (5%) and potentially other ischemic phenomena.

Systemic effects: Fever (14%), weight loss, malaise and myalgia (10%) represent the systemic impact of the tumor burden.

Dyspnea emerges as the most frequent symptom, followed by chest pain, palpitations and congestive heart failure. Importantly, constitutional symptoms like weakness and anemia, though less prevalent, should not be overlooked.

Accurate identification of abnormal features, including lack of positive blood cultures and failure to respond to antibiotic therapy, pointed to an underlying neoplastic disease [8-12]. The multimodal approach, integrating transthoracic and transesophageal echocardiography and advanced imaging techniques such as Positron Emission Tomography-Computed Tomography (PET-CT), was instrumental in the accurate identification of the primary cause: Metastatic osteosarcoma with cardiac involvement. To effectively navigate the diagnostic landscape of cardiac malignancies, the importance of combining clinical expertise, careful interpretation of imaging results and appropriate use of investigative tools is highlighted [9].

Transthoracic Echography (TTE) served as the initial diagnostic step, offering a readily available and non-invasive view of the cardiac structures. In this case, TTE likely revealed the presence of an intracardiac mass, prompting further investigation. However, differentiating a malignant mass from Infective Endocarditis (IE) vegetations using TTE alone can be challenging due to their overlapping features [3]. It precisely measures size, shape and mobility, aiding in distinguishing benign from malignant growths [1]. Its comprehensive view scans for hidden tumors in other chambers, ensuring a complete diagnosis. Importantly, this non-invasive approach enables repeated monitoring of the tumor and its response to treatment, making it a cornerstone of accurate diagnosis and effective management in the intricate realm of cardiac oncology.

Distinguishing cardiac sarcoma from benign myxoma involves consideration of various clinical and imaging aspects [5]. Sarcomas commonly originate non-septally, in contrast to the atrial septal wall preference seen in myxomas. Notably, extension into the pulmonary vein signals a potential malignancy. The presence of multiple intracardiac masses, unlike the usual solitary nature of myxomas, raises suspicion for sarcoma. Additionally, the mode of attachment, with sarcomas showing a broad connection to the left atrial wall compared to the narrow pedicle seen in myxomas, serves as a distinguishing feature. Moreover, the consistency of the mass, particularly its semisolid and firm nature on imaging, provides further differentiation, as myxomas typically exhibit a softer, gelatinous texture.

TTE and transesophageal echocardiography are commonly used for the initial visualization of intracardiac masses, whereas chest CT plays a unique and valuable role in the diagnostic landscape of myocardial sarcoma mimicking IE [6,9]. Its key contributions include the differentiation of tumors and vegetation, as CT chest offers superior tissue characterization compared to echocardiography [9]. While cardiac MRI provides detailed views of the heart and surrounding structures, its application may be limited due to contraindications and availability. For patients with implanted metallic devices, including mechanical valve prostheses and intra-cardiac devices, MRI can introduce artifacts that distort the image and hinder accurate diagnosis. In these situations, CT scanning emerges as a powerful alternative, providing clear visualizations of the heart and surrounding structures even in the presence of metallic implants.

In the assessment of the extension, CT of the chest provides a wider view of the mediastinum and chest wall. In contrast to echocardiography, this allows assessment of potential tumor invasion into surrounding structures [6]. This information is important for surgical planning and staging. CT imaging is also excellent at detecting distant metastases, a critical factor in determining prognosis and guiding treatment decisions [8]. In some cases, the tumor can involve the coronary arteries, which requires assessment. CT angiography can potentially have an impact on the feasibility of surgery and guide further management by delineating the involvement of chest CT scans.

CT is a useful diagnostic tool. Nevertheless, it has certain limitations. Challenges such as reduced image quality in the presence of tachycardia and the inability to perform a breath hold need to be considered. There are also restrictions on the use of iodinated contrast media if the patient has advanced renal failure but does not require dialysis and people who are allergic to contrast media need to be premedicated. Furthermore, there are ongoing concerns about radiation exposure and assessment of coronary lesion severity may be limited by the presence of significant calcification, which occurs predominantly in the elderly, those with renal insufficiency or those with a history of radiotherapy. It is worth noting that the technique may be limited to detecting small vegetation.

Contrast-enhanced Cardiac Magnetic Resonance (CMR) stands as the gold standard for cardiac imaging, offering exceptional capabilities for distinguishing between tumors and thrombi. Through the strategic use of a prolonged inversion time, CMR accentuates tissue characteristic differences, rendering thrombi

visually null while enabling the detection of contrast enhancement within the mass, indicative of a tumor. The advantages of CMR over echocardiography in cardiac tumor characterization are manifold [9], including superior spatial resolution, a broader field of view and signal characteristics that aid in histopathological characterization, including hemorrhage, necrosis, vascularity, fatty infiltration and calcification. CMR excels in imaging without attenuation. Nevertheless, its lower temporal resolution diminishes its utility for evaluating valvular vegetation compared to echocardiography. Limitations encompass extended acquisition times (30 minutes to 1 hour), limited availability relative to echocardiography, infeasibility in hemodynamically unstable patients and contraindications such as older generation cardiac devices and claustrophobia.

To enhance the diagnostic precision, PET-CT assumed a pivotal role. Its capacity to visualize the metabolic activity of tissues emerged as an invaluable asset using the radiotracer 18F-FDG [10]. The discernible disparity in metabolic activity between malignant tumors and IE vegetations served as a differentiating factor. This case underscores the indispensable contribution of PET-CT in effectively discriminating cardiac malignancies from other potential mimicking conditions, notably in the context of infective endocarditis.

The challenging prognosis linked to advanced sarcomas adds complexity to the therapeutic scenario. Despite the grim outlook, exploring palliative strategies involving targeted therapy and symptom management provides a ray of hope for prolonging life and improving the quality of life [11]. This case underscores the importance of personalized treatment plans that align with the unique clinical context and disease progression. Additionally, it emphasizes the significance of ongoing research endeavors dedicated to devising innovative therapeutic approaches for these aggressive malignancies [12].

CONCLUSION

In conclusion, this intriguing case report transcends the boundaries of a singular narrative. Cardiac sarcomas are uncommon but aggressive tumors that can mimic other heart illnesses and hence, go undetected for long periods of time. Clinicians must be vigilant and employ improved cardiac imaging tools, particularly echocardiography, to detect and define these malignancies. To improve survival rates, early identification and surgical removal of the tumor are required. The development of new imaging techniques and treatment options for these difficult cancers should be the focus of future research.

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