Radiology Case Reports 18 (2023) 1666–1670

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journal homepage: www.elsevier.com/locate/radcr

Case Report

A rare case of primary adult cardiac rhabdomyosarcoma with lower extremity metastasis ☆

Roozbeh Tarighati Rasekhi, MD[a], *Nastaran Hosseini*[b], *Savalan Babapoor-Farrokhran, MD*[c], *Ali Babapour Farrokhran, MD*[d], *Barzin Behzad, MD*[e,*]

[a] *Department of Neurology, SUNY Upstate, Syracuse, NY 13210 USA*
[b] *Isfahan University of Medical Sciences, Isfahan, Iran*
[c] *Leon H. Charney Division of Cardiology, Cardiac Electrophysiology, NYU Langone Health, New York University School of Medicine, NY, USA*
[d] *Urmia University of Medical Sciences, Urmia, Iran*
[e] *Department of Radiology, Division of Musculoskeletal, University of Pennsylvania, 3737 Market St, Mialbox #4, Philadelphia, PA 19104*

ARTICLE INFO

Article history:
Received 25 December 2022
Accepted 26 January 2023

Keywords:
Cardiac rhabdomyosarcoma
Cardiac sarcomas
Primary cardiac tumors

ABSTRACT

Primary cardiac tumors are uncommon. Rhabdomyosarcomas are among the rarest type of cardiac sarcomas. Echocardiography, cardiac MRI, and computed tomography scan can help the diagnosis and presurgical management. In this article, we report a rare case of primary cardiac rhabdomyosarcoma originating from the mitral valve with left femoral metastasis in a patient in her 60s. The diagnosis was made using transesophageal echocardiography and cardiac MRI. A metastatic lesion was found in an extended PET scan in one of her clinical follow-ups due to her leg pain. Based on this report, we suggest that extending PET scan to the lower extremities could be helpful in the early diagnosis and treatment of remote metastases of cardiac rhabdomyosarcomas.

Introduction

Primary cardiac tumors are uncommon, with an average prevalence of 0.002%-0.03% [1,2]. About 75% of initial cardiac tumors are benign, mostly myxomas. The remaining 25% are malignant, with sarcomas being the most common. Rhabdomyosarcomas account for about 5% of all malignant cardiac neoplasms in adults [3].

Transthoracic echocardiography is usually the first line to detect cardiac tumors. However, cardiac magnetic resonance imaging (CMR) is the preferred imaging study for the diagnosis. Cardiac computed tomography (CT) and CMR can also be used for preoperative planning, but surgical specimen and

☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

* Corresponding author.
E-mail address: barzin.behzad@pennmedicine.upenn.edu (B. Behzad).

https://doi.org/10.1016/j.radcr.2023.01.089

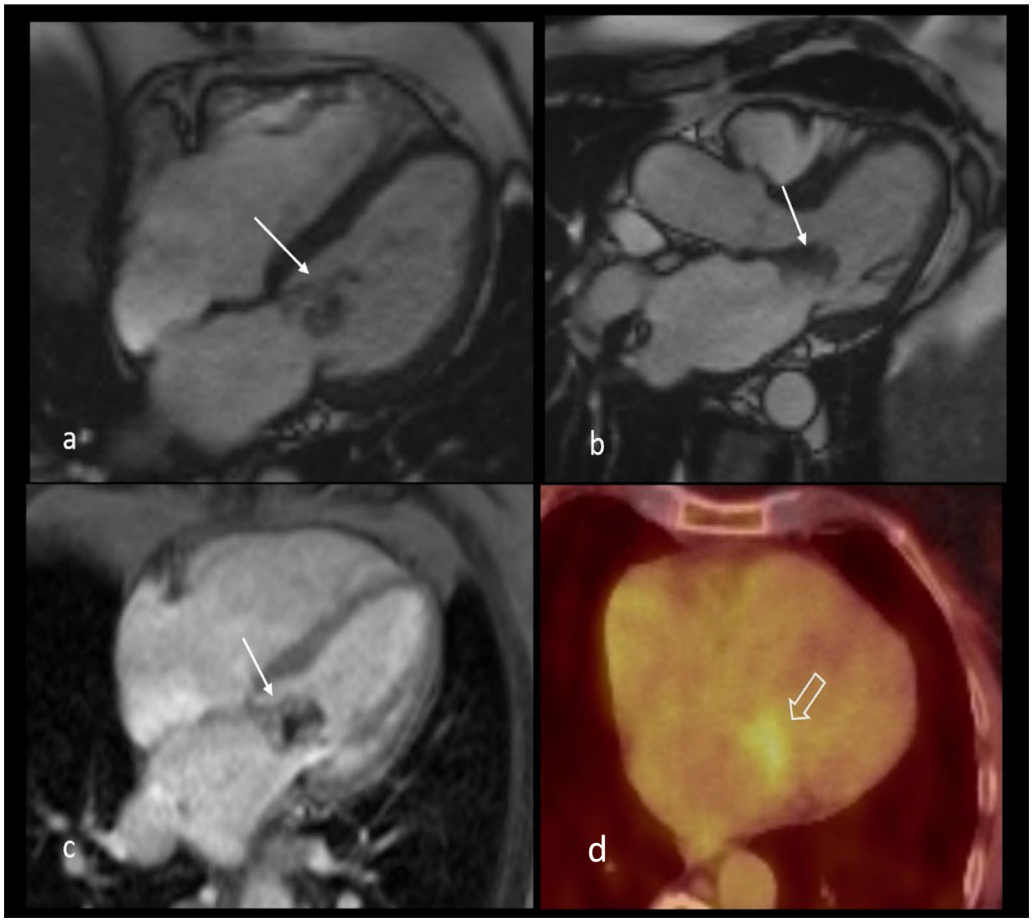

Fig. 1 – Representative images of cardiac MRI and PET scan. (A) Four chambers view, white blood, (B) Three chambers view, white blood, (C) Four chambers view, early enhancement, (D) PET scan. A lobular mass involving the anterior mitral valve leaflet with early enhancement (arrow on panels A, B, and C). The area of focal F-18 fluorodeoxyglucose uptake, more than blood pool, at the area of the mitral valve (open arrow on D).

histopathological analysis confirm the diagnosis of intracardiac malignancies [3–5]. Excision of the primary tumor, which is accompanied by chemotherapy and radiation therapy, is the standard therapy [4,6,7].

Although median survival rates vary between 1 to 4 years for cardiac sarcomas in different studies, depending on the extent of the disease, this number is usually less than 1 year for primary rhabdomyosarcoma [4,8].

In this article, we report an exceedingly rare case of rhabdomyosarcoma involving the mitral valve that was complicated by left femoral metastasis. We demonstrate findings on the imaging studies that helped with the diagnosis and treatment.

Case presentation

A female patient in her 60s presented to the hospital complaining of palpitations and shortness of breath. She had a history of breast cancer treated with radiotherapy and chemotherapy. Her electrocardiography was consistent with atrial fibrillation, and subsequently, she was scheduled for radiofrequency ablation. Transesophageal echocardiogram prior to ablation showed a 2 cm mass on the mitral valve on the atrial side, thus ablation was aborted.

CMR demonstrated a multilobulated mass encasing the entire anterior mitral valve leaflet with a mobile component and some echogenic punctate components prolapsing into the left ventricular chamber on the superior portion of the mass (Fig. 1). The mitral valve tumor was surgically removed. Surgery was complicated by a complete heart block for which a dual-chamber pacemaker was implanted. Histopathologic findings of the resected mass indicated lobular proliferation of round to spindled cells in the myxoid and sclerotic stroma with rhabdomyoblastic differentiation, and sections with botryoid architecture. There was necrosis with nuclear atypia and a high mitotic rate (>20/10 in high-power field). Nuclear atypia and rhabdoid features were more pronounced in the cellular and solid sections with less myxoid stroma. The above findings were consistent with the diagnosis of spindle cell/sclerosing rhabdomyosarcoma. Accordingly, chemotherapy was started for the patient.

At the 18-month follow-up visit, she complained of left thigh pain. The echocardiogram revealed a very large, multilobulated mass in the right atrium (3.8 × 2.2 cm). The exact attachment point was not delineated. Subsequent cardiac

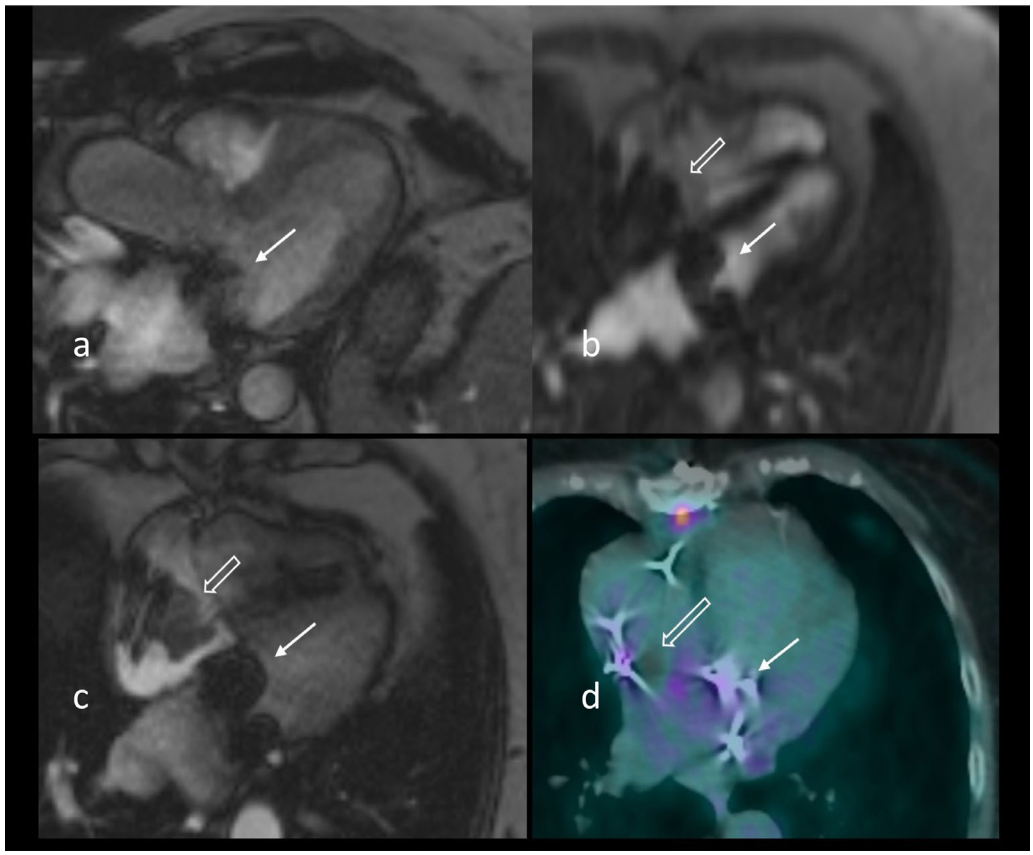

Fig. 2 – Representative images of the second cardiac MRI 18-month post tumor resection and mitral valve replacement and PET/CT. (A) Three chambers perfusion, (B) Four chambers perfusion, (C) Four chambers GRE, (D) PET/CT scan. Mitral valve replacement (arrow on A-D) with no evidence of local recurrence. There is a clot within the right atrium abutting pacemaker leads (open arrow on B, and C). A focal area of photopenia corresponds to a clot visualized on MRI (open arrow on D).

MRI showed a 35 × 23 × 41 mm focus adherent to the pacemaker lead in the right atrium, which was homogenously hypointense on first-pass perfusion and late enhancement sequences (Fig. 2). Area of photopenia on subsequent PET scan was consistent with intracardiac thrombus (Fig. 2). Since the patient had complained of discomfort in her left thigh, the PET scan was extended to her lower extremities, revealing a mass in her left femur. According to the imaging evaluation, there was a T1 hypointense marrow signal and enhancement of the distal left femur with significant periosteal reaction, depicting a sunburst pattern on the postcontrast sequence and F-18 fluorodeoxyglucose avid uptake (Fig. 3). An ultrasound-guided biopsy of the extraosseous soft tissue component of the left femoral lesion performed. Pathology exhibited spindle cell rhabdomyoblastic sarcoma of at least intermediate grade, consistent with previously identified cardiac rhabdomyosarcoma.

Based on the above findings, the mitral valve lesion was assumed to be the primary source of the thigh lesion.

Discussion

Primary malignant cardiac tumors are extremely rare. Patients are usually in their fifth decades of their lives, and it equally affects males and females [7]. Although sarcomas are the most prevalent primary cardiac malignancies, rhabdomyosarcomas are among the less common histological variants [7,9–12]. Clinical manifestations differ based on the tumor size and location. Patients may have different cardiac symptoms, including chest pain, palpitations, and syncope. Left-side tumors usually present with symptoms of heart failure, including shortness of breath and fluid congestion [1,7,9]. Imaging studies usually initiate with transthoracic echocardiography. It is a widely accessible imaging modality to evaluate the size and location of the mass. In echocardiography, rhabdomyosarcoma is usually a homogenously hyperechoic mass, while areas of hypo echogenicity are suggestive of intralesion necrosis. Using color Doppler echocardiography help to differentiate thrombus and evaluate the vascularity of the tumor [3,13]. Transesophageal echocardiography could be a more accurate tool for the diagnosis [13].

CMR is a sensitive mode of imaging for initial evaluation and surgical planning. The T1- and T2-weighted acquisitions help with the classification of cardiac tumors based on the imaging characteristics and the chemical content within the tumor [1,4]. On CMR, isointense T1-weighted and hyperintense T2-weighted images are characteristic of rhabdomyosarcomas. Moreover, T2 hyperintense foci can be seen due to central necrosis [13]. Cardiac CT of rhabdomyosarcoma

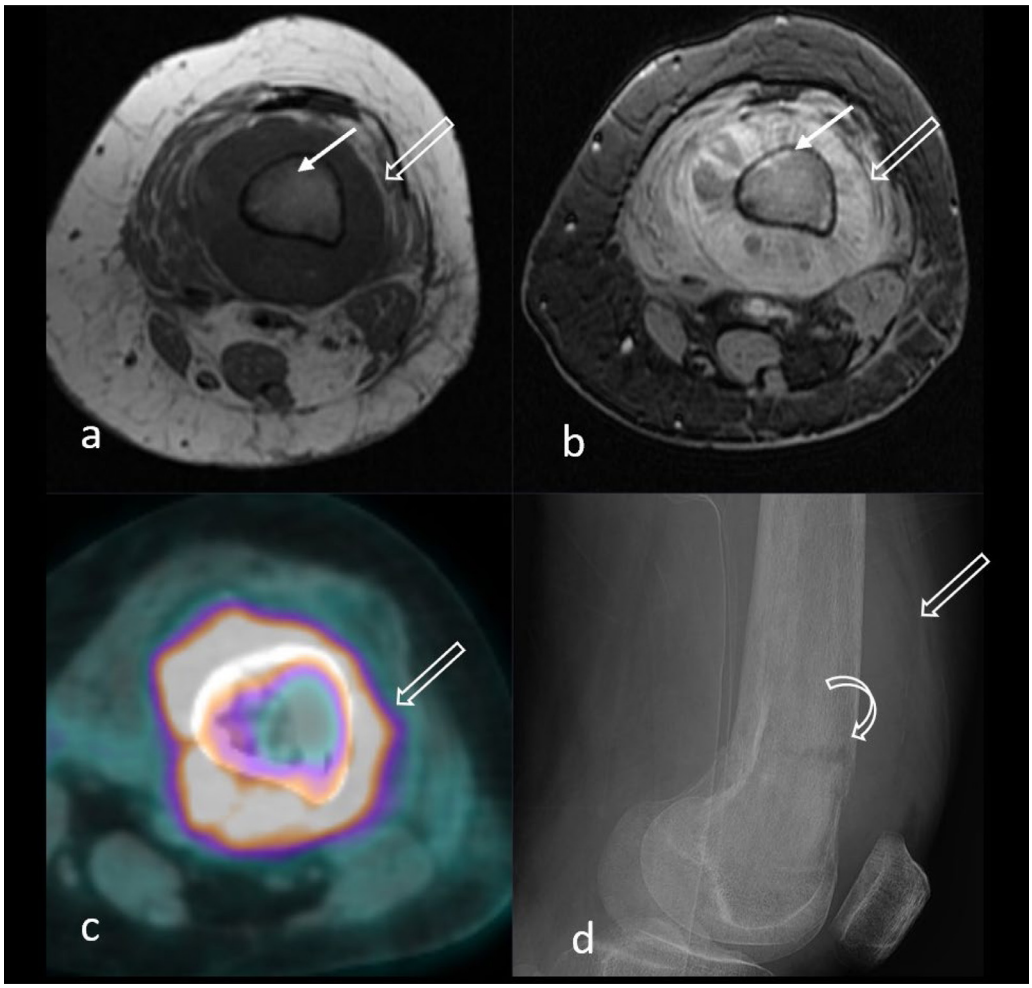

Fig. 3 – Representative images of Axial T1, Axial fat-saturated postcontrast, PET-CT scan, and Lateral radiograph of the distal left femur. (A) Axial T1, (B) Axial fat-saturated postcontrast, (C) PET-CT scan, (D) Lateral radiograph of the distal left femur. There is a T1 hypointense signal and enhancement of the distal left femur (arrow on panels A and B) with extraosseous soft tissue component (open arrow on panels A-D), which demonstrates avid contrast enhancement and F-18 fluorodeoxyglucose uptake. Mildly displaced transverse pathologic fracture of the distal femur (curved arrow on panel D).

may reveal a smooth or irregular low-attenuation mass. Additionally, CT and CMR can be valuable tools to evaluate the extent of the disease and local invasion into nearby structures such as pulmonary arteries, descending aorta, and pulmonary valve involvement [4,14].

While the most common target organs for metastasis of primary cardiac sarcomas are the brain, lungs, liver, and bone, our patient presented with left lower extremity metastasis [6,9]. Given a high risk of brain metastasis, some experts recommend a brain MRI for all patients with primary cardiac tumors at the time of diagnosis [7]. Our patient's femoral metastasis was found in one of her follow-up visits on an extended PET scan study owing to her leg pain. We believe that the aggressive nature of the tumor is serious enough to warrant an extended PET scan of the extremities for all patients with primary cardiac sarcomas at the time of diagnosis.

Furthermore, as evidenced by this case, employing the appropriate imaging modality in these rare cases is of paramount importance. Transthoracic echocardiography is the most available and cost-effective initial test, followed by CT, CMR, and PET scan for further characterization of the mass and surgical planning.

Conclusion

As mentioned previously, primary cardiac rhabdomyosarcoma is a rare tumor. It can present with metastatic soft tissue lesions. However, it could metastasize to a remote tissue that would not be imaged in a conventional PET scan. As reported in this case, the metastatic lesion was in the patient's lower extremity and would have been missed if PET scan was not extended to that region. Therefore, we suggest extending PET scan to the suspicious areas to better assess for any lesions that are suspicious for metastases.

Patient consent

A written informed consent for publication of this case was obtained from the patient.

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