

Cardiac Angiosarcoma: A Common Presentation of an Uncommon Malignancy

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Received: July 25, 2020; **Published:** August 11, 2020

Abstract

Cardiac angiosarcoma is the most common primary malignant tumor of the heart, but remains a rare entity. It usually affects young males and is typically associated with constitutional signs and symptoms. This unfortunately results in delayed diagnosis and subsequently poor prognosis. We describe a case of primary cardiac angiosarcoma that presented with abdominal fullness, diarrhea and syncope. We performed a review of the available literature on this rare malignancy.

Keywords: Cardiac Angiosarcoma; Malignancy; Heart

Introduction

Cardiac angiosarcoma is a rare primary malignant tumor of the heart that typically arises in the right atrium causing symptoms of right sided heart failure or cardiac tamponade. It often manifests late with nonspecific constitutional signs and symptoms, resulting in delayed diagnosis and poor prognosis [1-4]. Our case illustrates the role of different diagnostic modalities in management of such a rare disease in addition to the therapeutic challenges in the absence of concrete guidelines.

Case Description

This is a 32 year old gentleman with no known significant past medical history who developed diarrhea and abdominal fullness four days prior to presentation. He complained of dizziness followed by a witnessed syncope as he laid himself to the ground. He was taken to an outside hospital where an abdominal CT scan showed no acute findings. Incidentally noted was a large pericardial effusion for which he was transferred to our institution. On arrival to our hospital, he denied chest pain, shortness of breath, fever, recent viral illness, rashes or tick bites. His vital signs were significant for sinus tachycardia. He had normal blood pressure. Physical examination demonstrated elevated jugular venous pressure, muffled heart sounds and pulsus paradoxus (18 mmHg). He also had mild epigastric tenderness. CRP was elevated at 17.2 mg/L (reference range < 5 mg/L). Infectious laboratory work up, including acute hepatitis panel and HIV serology, was negative. Abdominal ultrasound revealed ascites, but was otherwise unremarkable. Electrocardiogram (ECG) showed sinus tachycardia with electrical alternans.

Transthoracic echocardiogram (TTE) was done on arrival and confirmed the presence of large pericardial effusion. This was associated with right ventricular collapse suggestive of tamponade physiology, which was in line with the concerning signs for tamponade on physical examination. Furthermore, a mass adherent to the roof of the right atrium was also noted on TTE. Pericardiocentesis elicited 800cc of grossly bloody fluid and a pericardial window was performed. Pericardial fluid cell count revealed lymphocytic predominance. Cytology, infectious and rheumatologic workup of the pericardial fluid was negative. He underwent cardiac CT to further characterize the right atrial mass. However, it offered inconclusive assessment of the mass given significant mixing artifact. Subsequent cardiac MRI demonstrated

enhancing soft tissue density concerning for neoplasm throughout the free wall of the right atrium, filling the right atrial appendage and extending into the interatrial septum (Figure 1 and 2). The mass was not seen to extend beyond the right atrium into the superior vena cava. It also spared the inferior vena cava and right atrium junction. Transesophageal echocardiogram (TEE) guided myocardial biopsy was performed and histopathology was consistent with cardiac angiosarcoma (Figure 3-5). It was decided to pursue a neoadjuvant approach rather than initiating treatment by surgical excision. Treatment consisted of five cycles of Gemcitabine and Docetaxel followed by definitive proton beam irradiation. Following completion of the treatment course, CT of the chest, abdomen and pelvis showed no evidence of metastatic disease. Re-staging cardiac MRI did not show evidence of progression of intra-cardiac disease after thirteen months of diagnosis. He is now scheduled to undergo surgical excision.

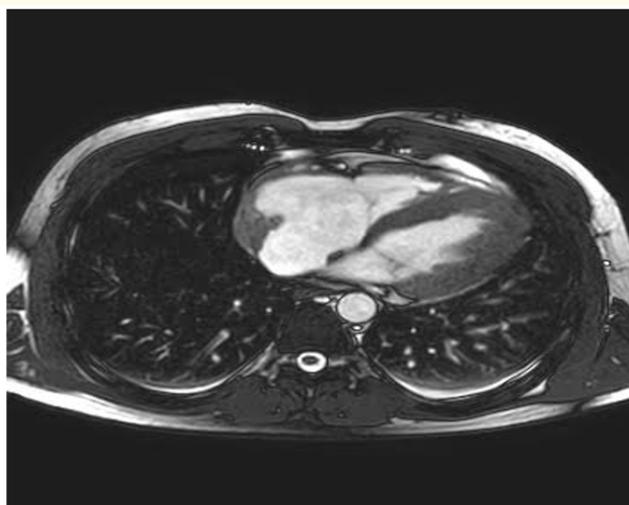


Figure 1: Cardiac MRI showing a mass originating from the right atrial wall.

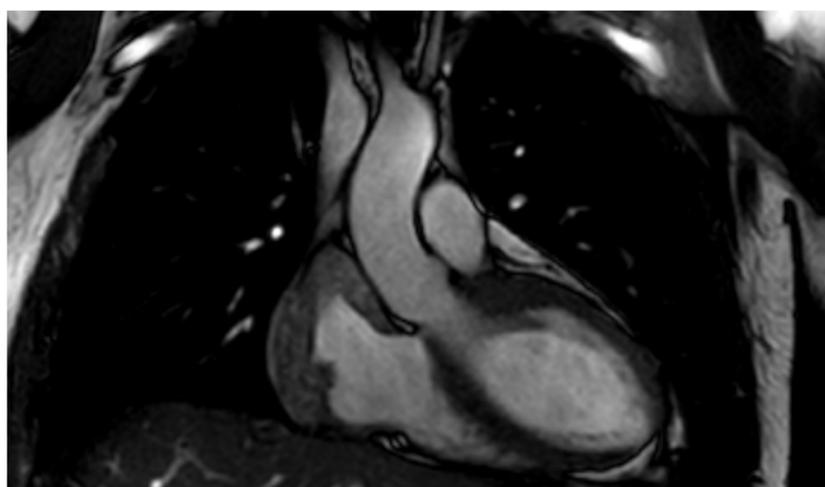
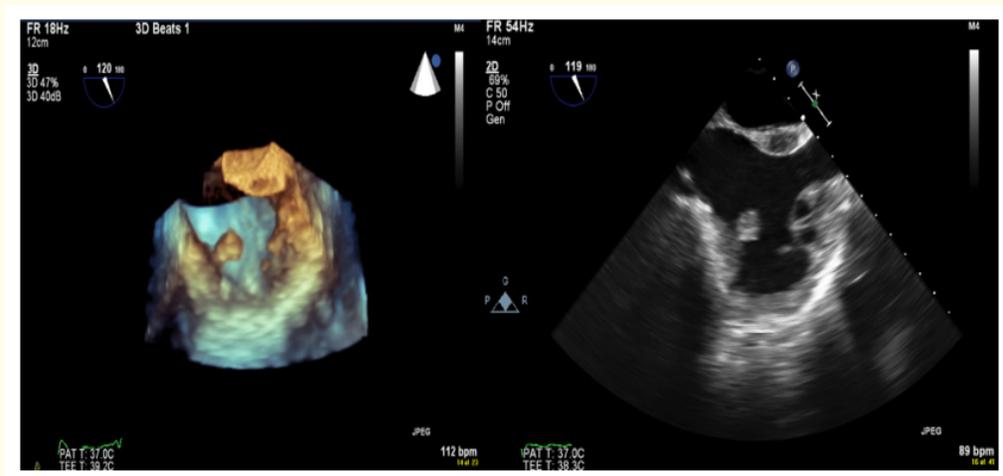
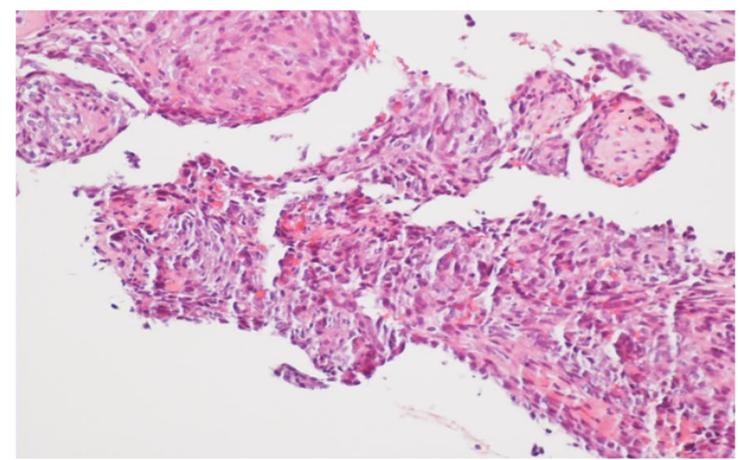


Figure 2: Cardiac MRI demonstrating extension of the right atrial mass throughout the free wall of the right atrium to the interatrial septum.



Figures 3 and 4: Transesophageal echo (TEE) 3D and 2D pictures demonstrating the right atrial mass throughout the free wall of the right atrium in the bicaval view.



Figures 5: Histopathology showing an atypical infiltrative spindle cell vascular neoplasm consistent with cardiac angiosarcoma.

Discussion

Primary cardiac tumors are rare and are 20 - 40 times less prevalent than metastatic tumors of the heart [2,3]. They account for about 25% of all primary cardiac tumors, with angiosarcoma being the most common, constituting 33% of cases. Primary cardiac angiosarcomas (PCAs) typically affect males in the 3rd - 5th decade [4-6] and often present with nonspecific constitutional signs and symptoms including fever, nausea, anorexia, diarrhea, dyspnea and atypical chest pain. Given the tendency of PCAs to involve the right side of the heart, right-sided congestive heart failure, pericardial effusion and cardiac tamponade are common presentations [1,3,4]. Other signs include valvular and caval obstruction leading to stenosis, systemic or pulmonary emboli, hemoptysis and supraventricular arrhythmias

[4,5]. PCAs are also known to be aggressive tumors with high incidence of metastatic spread, up to 89% on presentation, resulting in an unfavorable prognosis [7,8].

Diagnosis of PCAs remains challenging due to the low index of suspicion secondary to the rarity of the tumor and the nonspecific symptoms on presentation. Imaging is key to diagnosis with TTE as the initial diagnostic modality. TEE is used to better define the exact location and size of the tumor with 97% sensitivity in detecting cardiac masses [4,6]. Cardiac magnetic resonance (MR) and CT scan are superior to echocardiography in better defining the tumor and evaluating locoregional extension to the lungs and mediastinum [1,2,6,7]. Owing to its high resolution, cardiac MR provides better characterization of soft tissue than CT scan, with the ability to distinguish between different tumor types as well as differentiating cardiac tumors from thrombi [1,6,9]. CT scan is used to assess for the presence of distant metastases. Moreover, in contrast to cardiac MR, CT scan can demonstrate tumor calcifications [2,6]. Previous data concluded that pericardial fluid cytology is positive in 75 - 87% of patients [10]. However, different literature suggested that cytology is rarely positive as malignant cells are seldom found in bloody fluid [6,7,11]. TEE-guided myocardial biopsy increases the diagnostic yield, as was used with our patient [12].

Traditionally, wide surgical excision has been the treatment of choice but is marred by tumor recurrence due to technical limitations and micrometastases. A case series in 2009 reported a median survival time of 27 months for patients who underwent surgical excision of right sided cardiac sarcomas, 15 of which were PCAs [7]. The role of chemotherapy and radiotherapy is controversial. Several case reports described improved outcome with adjuvant chemotherapy when part of a combined modality approach [13-15]. Other data showed that post-operative chemotherapy and radiotherapy provided no mortality benefit [16,17]. A review of literature in 2010 suggested benefit from multimodality therapy compared to one treatment alone [2]. Cardiac transplantation has been used as a therapeutic option for PCAs, but has not been shown to improve outcome [18]. In our patient, it was believed that the hemorrhagic nature of the tumor indicated contamination of the entire pericardial space, making it unlikely to encompass all the disease with surgical resection alone. We favored neoadjuvant chemotherapy with Gemcitabine and Docetaxel followed by proton beam irradiation. Thirteen months after diagnosis, he has had no progression of disease on surveillance imaging and is scheduled to undergo surgical excision.

Conclusion

We report a case of primary cardiac angiosarcoma identified with advanced cardiac imaging and myocardial biopsy treated with a novel multi-disciplinary approach to include neoadjuvant chemotherapy and radiation without progression of disease on surveillance imaging, scheduled for surgical excision.

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Volume 7 Issue 9 September 2020

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