Cardiac angiosarcoma: case report and review of the literature

Introduction

Neoplastic involvement of the heart at autopsy has been reported to occur in 2% to 20% of all patients dying of malignancy [1]. In contrast, primary tumors of the heart are exceedingly rare. Angiosarcoma is the most common primary malignant tumor of the heart in adults with a uniformly dismal prognosis [2]. Herein, we report a case of an extensive angiosarcoma involving the right heart and review the literature on this type of tumor with emphasis on symptoms, diagnosis and therapeutic options.

Case report

A 63-year-old woman with a 1-month history of malaise, weakness and dyspnea on exertion was re-
ferred to our institution for further evaluation. She reported two episodes of non-exertional syncope without premonitory symptoms, but denied having palpitations, chest pain, abdominal pain, fever, or weight loss. She had undergone total abdominal hysterectomy 15 years earlier for benign leiomyomas. Otherwise, her medical and surgical history was unremarkable. She was taking no medications and had no drug allergies. Physical examination revealed a normal-appearing, well nourished woman in no acute distress with a regular pulse rate of 80/min and a blood pressure of 120/80 mm Hg. Her jugular venous pressure and contour were normal. Of note, neither the Kussmaul sign nor a paradoxical pulse was present. The apical impulse was difficult to palpate and the heart sounds were muffled. There was no click or rub. Systole and diastole were free of murmurs. The lungs were clear, and findings on the rest of her physical examination were unremarkable. Electrocardiography showed normal sinus rhythm, low voltage, and nonspecific ST and T-wave abnormalities. Initial laboratory results (reference ranges shown parenthetically) were remarkable for a hemoglobin concentration of 11.4 g/dL (12–16), platelet count of 510×10^9/L (150–400), and lactate dehydrogenase level of 244 U/L (135–214). Routine trans-thoracic echocardiography disclosed a substantial pericardial effusion and a large intracardiac mass adjacent to the right heart (Fig. 1). Continuous-wave Doppler echocardiography of the right atrium and right ventricle from the apex with the Doppler beam directed across the tricuspid valve did not find any degree of obstruction to blood flow in the right heart. Pulsed-wave Doppler interrogation of mitral and tricuspid inflow did not reveal any significant respiratory changes suggestive of increased intrapericardial pressure. The right and left ventricular function appeared normal. Multiplane transesophageal echocardiography (Fig. 2) and computed tomography (Fig. 3) were performed to better define the location and extension of the mass. The patient underwent echocardiographically guided pericardiocentesis with removal of 850 mL of hemorrhagic fluid. Cytologic examination identified abundant erythrocytes, several granulocytes and mesothelial cells, but failed to show any malignant cells. Results of examinations for tuberculosis and other bacterial and fungal agents were negative. Coronary angiography

![Fig. 1](image1.png)

**Fig. 1** Transthoracic parasternal longitudinal view of the left ventricle (LV) showing a circumferential pericardial effusion (PE) and mural mass (M) with a polycyclic surface (arrow) infiltrating the epicardium of the right ventricular (RV) free wall

![Fig. 2](image2.png)

**Fig. 2** Transesophageal longitudinal view of the interatrial septum demonstrating a large (7×9 cm) echogenic mass (M) in the anterior and superior mediastinum. The tumor appears to involve and replace the entire thickness of the right atrial wall and protrudes into the right atrial (RA) cavity and superior vena cava (SVC)

![Fig. 3](image3.png)

**Fig. 3** Multislice computed tomography in a four-chamber orientation (multiplanar reconstruction). After administration of contrast agent, the mass (M) is delineated from the right atrium (RA) and ventricle (RV). Arrowheads denote pericardial effusion. LA left atrium; LV left ventricle