Left Atrial Angiosarcoma: A Rare Cardiac Tumor at an Uncommon Site

Amitabh Poonia, Rekha Mishra, Priya Giridhara, Yogendra Kumar Arora
Department of Cardiology, National Heart Institute, 1Department of Cardiology, VMMC and Safdarjung Hospital, New Delhi, India

Abstract

Cardiac angiosarcoma is the most common among primary malignant cardiac tumors in adults. Malignant cardiac tumors commonly arise in the right-sided cardiac chambers unlike benign tumors that commonly arise in the left-sided chambers. Cardiac tumors on the left side need to be carefully assessed for malignant features for deciding treatment strategy and prognostication. We present the case of a 62-year-old female with a large left atrial mass infiltrating the interatrial septum and adjacent myocardial wall. Histology was suggestive of angiosarcoma. Although a radical excision was done, the tumor recurred within 6 months of the postoperative period and she died shortly after the recurrence.

Keywords: Angiosarcoma, malignant tumor, primary cardiac tumor

Introduction

Angiosarcoma is the most common primary malignant cardiac tumor and is notorious for its poor prognosis and high-recurrence rate after resection. In contrast to the benign tumors which commonly arise in the left atrium, malignant tumors arise mostly in the right-sided chambers, especially in the right atrium. However, careful echocardiographic assessment is important to differentiate angiosarcoma from a myxoma, especially when the tumor originates from the left-sided cardiac chambers.

Case Report

A 62-year-old female presented with progressive exertional dyspnea of 6 months duration. At admission, she was afebrile, tachypneic with a heart rate of 120/min, blood pressure of 110/60 mm Hg, and elevated mean jugular venous pressure. On auscultation, mid-diastolic murmur was audible at the apex. Transthoracic echocardiogram revealed an echogenic mass measuring 48 mm × 14 mm arising from the left atrial roof and protruding into the left atrial cavity. On transesophageal echocardiogram, a 65 mm × 28 mm large, wide-based, multilobed, irregular-shaped, heterogeneous mass with multiple hypoechoic areas arising from the posterolateral aspect of the interatrial septum was seen. The mass infiltrated into the septum, surrounding myocardium, and also protruded into the left atrial and left ventricular cavities causing moderate functional mitral stenosis. Further assessment with cardiac magnetic resonance imaging and positron emission tomography (PET) scan was advised for further tumor characterization, but declined by the patient and got discharged from the hospital. Three weeks later, she presented to the emergency department with worsening symptoms and pulmonary edema. She was in atrial fibrillation. She underwent surgical excision of the left atrial mass successfully following initial medical stabilization. The excised mass was firm, cherry red colored, somewhat elongated, and multilobed. Histopathology revealed both hypocellular and hypercellular areas. The hypocellular areas showed myxoid degeneration with thin-walled staghorn blood vessel proliferation, whereas the hypercellular areas revealed sheets of neoplastic cells with moderate nuclear atypia, pleomorphism, and atypical mitosis.

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Histopathology findings were consistent with an angiosarcoma. Subsequent postoperative echocardiogram revealed no residual mass, and the patient was discharged. No cardiac or metastatic tumor activity was detected at a 1-month follow-up on PET scan or echocardiogram. However, 6 months later, she came back with the complaints of palpitations and progressive exertional dyspnea of 2 weeks duration. Transthoracic echocardiogram revealed a 29 mm × 22 mm heterogeneous mass arising from the same site and protruding into the left atrial cavity. Both the mitral leaflets were thickened with markedly restricted movement and causing mitral stenosis which suggested infiltration of the tumor [Figure 3]. Unfortunately, the patient had sudden cardiac arrest and expired on the day of admission.

**DISCUSSION**

Only 10%–15% of primary cardiac tumors are malignant, with angiosarcoma being the most common of them.[3] It is notorious for its poor prognosis and high recurrence rate after excision.[3] The presenting signs and symptoms are generally nonspecific and depend on the size and location of the tumor. Malignant tumors may invade the myocardium and pericardium, resulting in arrhythmias, heart failure, and pericardial effusion. Sudden death is infrequent but can be the first presentation of the tumor.[4] Sarcomas, in general, grow rapidly, extensively, and metastasize early. The diagnosis is often delayed due to the nonspecific nature of signs and symptoms. Histopathology is the gold standard for the diagnosis of cardiac tumors. Two-thirds of angiosarcomas are moderately differentiated displaying irregular, anastomosing vascular networks that are surrounded by pleomorphic, atypical cells with frequent mitoses, whereas the rest one-third are poorly differentiated, consisting of anaplastic spindle cells in a hyaline stroma and pools of extravasated red blood cells focally. Strong positivity for factor VIII, CD31, and CD34 on immunohistochemistry suggests the endothelial origin of the tumor cells.[3]

Although histopathology is the gold standard, echocardiogram is the most important initial imaging modality for the diagnosis. The location, size, shape, extent, and mobility of the tumor can be assessed by transthoracic echocardiography. Transesophageal echocardiography provides better delineation of the tumor, with a sensitivity of 97% for detecting cardiac masses.[6]

In contrast to the benign tumors which are commonly located in the left atrium, malignant tumors mostly arise in the right-sided chambers, especially in the right atrium.[1,2] The most common primary cardiac tumor arising from the left atrium is the myxoma. Careful echocardiographic assessment is necessary to differentiate an angiosarcoma from myxoma, especially when it originates from the left-sided cardiac chambers. Compared to a myxoma, the angiosarcoma has a broad base or has no stalk, lobulated, distinctly heterogeneous mass with areas of necrosis, and/or hemorrhage. Furthermore, angiosarcoma infiltrates the underlying tissue and is commonly associated with pericardial effusion.

At the time of diagnosis, 89% of the cases reveal metastasis, and still, radical excision is the most effective treatment for primary cardiac malignancies. The prognosis is very poor in spite of adjuvant treatments such as chemotherapy or radiotherapy.[7] They do not improve survival rates.[7]

**CONCLUSION**

Any broad-based, multilobed, heterogeneous cardiac mass with or without infiltration of underlying myocardium or pericardium, irrespective of the type of cardiac chamber from where they arise should be assessed by multiple noninvasive modalities to rule out malignancy. Transesophageal echocardiogram helps to delineate and characterize the cardiac masses better than a transthoracic echocardiogram.
and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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Figure 3: Transthoracic echocardiogram revealing a 29 mm × 22 mm heterogeneous mass (arrows) attached to the interatrial septum and protruding into the left atrial cavity. Both the mitral leaflets are thickened. LA = Left atrium, LV = Left ventricle, RA = Right atrium, RV = Right ventricle

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published