

Cardiac Angiosarcoma: A Case with Bilateral Pleural and Pericardial Effusion

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ABSTRACT

Cardiac sarcomas are rare malignant tumors. Angiosarcoma is the most common cardiac sarcoma and is present in up to 33% of cases. Angiosarcomas have a poor prognosis, with a short survival expectancy. We report a case of a right atrial angiosarcoma treated by partial tumor resection followed by chemotherapy.

INTRODUCTION

Primary tumors of the heart are uncommon but not rare. The incidence of primary cardiac neoplasm ranges between 0.017% and 0.19% in various autopsy series [Strauss 1945; Lam 1993; Kurian 2006]. Cardiac tumors have a wide range, from nonneoplastic lesions to high-grade malignancies. Cardiac angiosarcomas are malignant tumors of mesenchymal origin with unclear etiology and are present in 0.0017%-0.033% of autopsy cases [Kurian 2006]. These tumors mostly occur in the third to fifth decade of life, with male predominance [Reardon 2006]. Over 80% arise in the right atrium. Cardiac sarcomas are often asymptomatic until the advanced stages; even then, they cause a variety of nonspecific symptoms and may mimic other pathologies, delaying diagnosis.

Because angiosarcoma is essentially not responsive to current regimens of chemotherapy and irradiation, early complete resection is recommended as the treatment choice [Look Hong 2012]. However, complete resection is not always possible because of the limited amount of myocardium and expansion of the tumor at the time of diagnosis [Look Hong 2012]. We report a case of right atrial angiosarcoma referred with repeated pleural and pericardial effusion treated by tumor resection and chemotherapy.

CASE REPORT

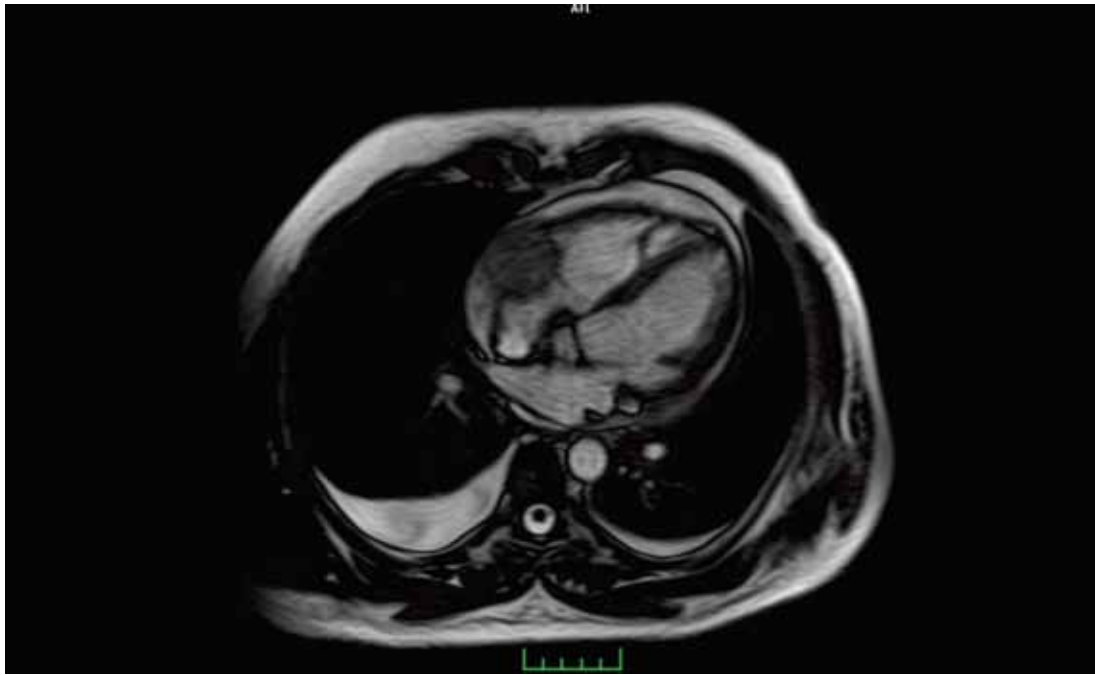
A 44-year-old man was admitted in our university hospital emergency department due to dizziness and unconsciousness. He was otherwise healthy. On physical examination, his vital signs were stable, other than a mild tachycardia (heart rate 104 bpm). There were no palpable lymph nodes. Jugular venous pressure, carotid upstroke, and heart sounds were unremarkable. There were no pathological murmurs or rubs. On respiratory exam, the chest wall was dull on percussion at the right and left base and he had decreased air entry bilaterally, but no crepitations were heard. His abdominal exam result was normal. The chest x-ray showed bilateral pleural effusions. On surface electrocardiogram he had widespread ST elevations. Transthoracic echocardiography (TTE) revealed moderate circumferential pericardial effusion. The patient was referred to the cardiology department and was hospitalized with the initial diagnosis of pericarditis.

The next day on the repeated TTE, a heterogeneous 3.2 × 3.3-cm cardiac mass was observed on the right atrium. On cardiac magnetic resonance imaging, a 6.0 × 4.5 × 4.0-cm large excentric and inhomogeneous solid mass of the right atrium extending to the atrioventricular groove and right ventricle, surrounding the right coronary artery (RCA) was found (Figure). After this diagnosis the patient was transferred to the cardiac surgery department and surgical intervention was then considered. Histological examination of a frozen section during surgery revealed that the tumor was malignant (most likely a type of sarcoma). As the tumor could not be totally resected because of the anatomical considerations, a palliative approach was adopted, the bulk was partially resected as much as possible without harming the groove and the RCA. In the histopathological findings, there were atypical mitotic cells with nuclear polymorphisms. The tumor cells were vitamin and CD34 positive and were identified as malignant mesenchymal tumor; angiosarcoma. The postoperative TTE was normal, and the patient was discharged home in stable condition 14 days after his surgery, with follow-up appointments with his oncologist. His scintigraphy with 20.mCi Tc-99m MDP revealed increased staining in the 4th-5th lumbar vertebral corpuses. After surgery the patient began receiving chemotherapy.

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Magnetic resonance imaging of cardiac angiosarcoma.

DISCUSSION

Primary tumors of the heart are 20-40 times less common than metastatic lesions, and of these, benign tumors are more common than the malignant ones [Hyde 2005]. Of the malignant tumors, primary cardiac angiosarcoma is the most common cardiac sarcoma and makes up 33% of the cases associated with a poor prognosis. It occurs predominantly in men between the third and fifth decades of life [Kodali 2006; Amonkar 2006]. The right atrium is the most common site of origin, followed by the left atrium, right ventricle, and left ventricle [Kodali 2006; Simpson 2008]. Cardiac sarcomas are often asymptomatic until the advanced stages, and even then, cause a variety of nonspecific symptoms and may mimic other pathologies, delaying diagnosis. Dyspnea on exertion is the most common symptom of primary cardiac sarcomas (79%), followed by chest pain (38%), cough (21%), paroxysmal nocturnal dyspnea (12%), hemoptysis (12%), embolic events (9%), and fever (9%) [Simpson 2008].

According to Shanmugan, cardiac sarcomas manifest via several mechanisms: (1) obstruction to blood flow and interference with valve function; (2) local invasion causing arrhythmias or pericardial effusion with tamponade; (3) embolic phenomena from tumor fragments or peritumoral thrombi; (4) systemic or constitutional symptoms including dyspnea, syncope, chest pain, fever, malaise and weight loss. Left chamber tumors can cause cerebral, coronary, and retinal emboli. Right-sided tumors may be the source of pulmonary emboli that can cause pulmonary hypertension when extensive [Shanmugan 2006].

Although the definitive diagnosis is with biopsy, several diagnostic tools for primary cardiac angiosarcoma are available. Echocardiography is a reliable, noninvasive, and widely available tool for detecting cardiac tumors, the site of tumor attachment, the pattern of tumor movement, and tumor size [Kurian 2006; Meng 2002]. Cardiac computed tomography and magnetic resonance imaging may reveal infiltrative growth and the extra cardiac extent of sarcomas [Shin 1987]. Pericardial fluid cytology is positive in 75% to 87% of patients [Kodali 2006; Amonkar 2006; Simpson 2008]. In all the reported series, the overall prognosis of cardiac angiosarcoma is poor with a median survival ranging from 6 to 13 months [Kodali 2006; Amonkar 2006; Simpson 2008; Look Hong 2012]. Clinicians should therefore be vigilant about the detection, investigation, and referral of patients with cardiac angiosarcoma for multidisciplinary management. The main step for the treatment of cardiac sarcoma is surgical resection. Complete surgical resection is the most important prognostic factor [Percy 1987; Nakamichi 1997]. There are reported cases of patients with angiosarcoma treated with only partial resection followed by chemotherapy and radiotherapy who survived 34 or even 53 months [Percy 1987; Nakamichi 1997; Olsun 2007]. In particular, pre-operative chemotherapy may decrease tumor bulk and eliminate micrometastasis prior to local tumor excision. Multiple studies of patients receiving chemotherapy after surgery demonstrated poor prognosis [Hyde 2005; Kurian 2006]. These findings suggest that there may be a benefit to treating patients with metastatic disease with chemotherapeutic agents prior to surgery. Cardiac transplantation has been attempted, but limited experience with this treatment has resulted in poor outcomes because of local recurrence and metastasis [Talbot 2002].

CONCLUSIONS

Although larger series from multiple centers are necessary to address the efficacy of multiple treatment modalities and long-term outcomes, according to our literature search the overall prognosis is poor, with a short survival expectancy reported. Cardiac angiosarcomas are very aggressive and early metastases are common. We should therefore be vigilant about the detection, investigation, and referral of patients with cardiac angiosarcoma for multidisciplinary management. The onset of new diagnostic and treatment modalities may offer hope to these patients.

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