Right Atrial Angiosarcoma with Severe Biventricular Dysfunction and Massive Pericardial Effusion

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Abstract- This paper presents the case of a 35 year-old woman with symptoms of heart failure from the last month. A physical examination at admission showed paleness, dyspnea, peripheral edema and fatigue. In a two-dimensional echocardiography and transesophageal echocardiography, normal thickness but severe left and right ventricular dysfunction with severe pericardial effusion and thickened pericardium were found. In the enlarged right atrium, an oval-shaped structure was found with features of continuity with lateral right atrial wall and also a bulging of the structure through the orifice of the tricuspid valve to the right ventricle. In the echocardiography, we did not saw any blocking of the tricuspid valve or the inflow from inferior vena cava (IVC) or superior vena cava (SVC) or coronary sinus. On the basis of the echocardiography examination and clinical presentation, tentative diagnosis of the right atrium myxoma was made. A coronary angiography revealed normal coronary arteries and no feeding of tumor by branch of right coronary artery (RCA). Surgical removal of the tumor was performed without complication. The histopathological examination confirmed the diagnosis of angiosarcoma. In the follow-up echocardiography carried out after three months, severe left ventricular (LV) and right ventricular (RV) dysfunction continued and was demonstrated. Magnetic resonance imaging revealed no lymphadenopathy or re-growth of the tumor in the mediastinum or pericardium.

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Keywords: Angiosarcoma; Heart tumor; Pericardial effusion

Introduction

Primary heart tumors are rare and found in only about 0.02% of autopsies and 25% of those cases are malignant, while one-third of those are angiosarcoma (1). The most common malignant tumors are metastatic tumors, while primary angiosarcomas are very rare. Other malignant tumors include: rhabdomyosarcoma, osteoblastoma, leiomyosarcoma, undifferentiated sarcoma and primary lymphoma. Infiltration of a tumor to the epicardium creates massive pericardial effusion (2). Angiosarcoma is localized in the right atrium in about 74% of patients, while in 14% it occurs in the ventricle and in 12% in the left atrium pulmonary vein and pericardium. The diagnosis work up of an angiosarcoma is based firstly on the transesophageal and transthoracic echocardiography examination (3). These two modes of examination make it possible for cardiologists to establish a tentative diagnosis and define the anatomic relation of the tumor to nearby structures and to localize the pedicle attachment as well as the presence of intravascular necrosis. The definitive diagnosis of the angiosarcoma is possible solely on the basis of a histopathologic examination of the surgically removed mass (4,5).

Case Report

A 35 year-old female patient was referred to our hospital due to severe dyspnea, weakness, fatigue and peripheral edema. These complaints had been becoming more pronounced over the last month. On the physical examination on the day of admission, peripheral edema, anemia, and dyspnea were found.

Her heart rate was 100/min and arterial blood pressure was 100/60 mmHg. She was afebrile with no

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cyanosis on her lips or fingernails. The chest exam revealed fine crypitation on auscultation. Heart sounds were normal and there were no murmurs. The liver was palpable 2 cm below the inferior costal margin. The blood exam revealed anemia (Hemoglobin, 7.5 g/dl) and an elevated erythrocyte sedimentation rate (ESR) of 70. The electrocardiography (ECG) demonstrated right atrial enlargement. Chest x-rays showed severe cardiomegaly due to pericardial effusion. Tran-thoracic echocardiography showed a right atrial mass prolapsing to the right ventricle and the site of origin of the mass could not be ascertained from transthoracic echocardiography, therefore a transesophageal echocardiography was carried out. Angiography revealed a normal coronary artery and tumor that was not vascularized by any right coronary artery branch. An urgent operation was indicated and the patient was taken to the operating room. After a mid-line sternotomy and aortic and bicaval cannulation, the aorta was cross - clamped with cardioplegic arrest, the right atrium was opened carefully, and a lobulated, necrotic mass was noted to have infiltrated the lateral wall of the right atrium to the epicardium (Figure 1).

Because it was extensively attached to the lateral wall of the right atrium, the tumor was completely excised together with the lateral atrial wall that was reconstructed with an autologus pericardial patch (Figures 2 and 5). The tumor measured 5.5 x 3.5 x 5 cm and its microscopic appearance was consistent with an angiosarcoma (Figures 3 and 4). Microscopically tumor differentiation was reflected by formation of irregular anastomosing vascular spaces. Myxomas are composed of myxoma cells, which have an ovoid nucleus with inconspicuous or large nucleoli, abundant eosinophilic cytoplasm, and indistinct cell borders. Mitotic figures and areas of hemorrhage and necrosis are always present. Myxoma cells form complex structures including rings, syncytia, and cords that are typically infiltrated by lymphocytes and macrophages. A myxoid background is present in areas without fibrosis. Hemosiderin is present within histiocytes and some myxoma cells. The diagnosis of cardiac angiosarcoma can be confirmed by immunohistochemical staining for endothelial maker, such as CD31 and others specific coagulation factors (6-9).

The post-operative course was uneventful, and she remained asymptomatic for three months after surgery with no evidence of tumor re-growth.

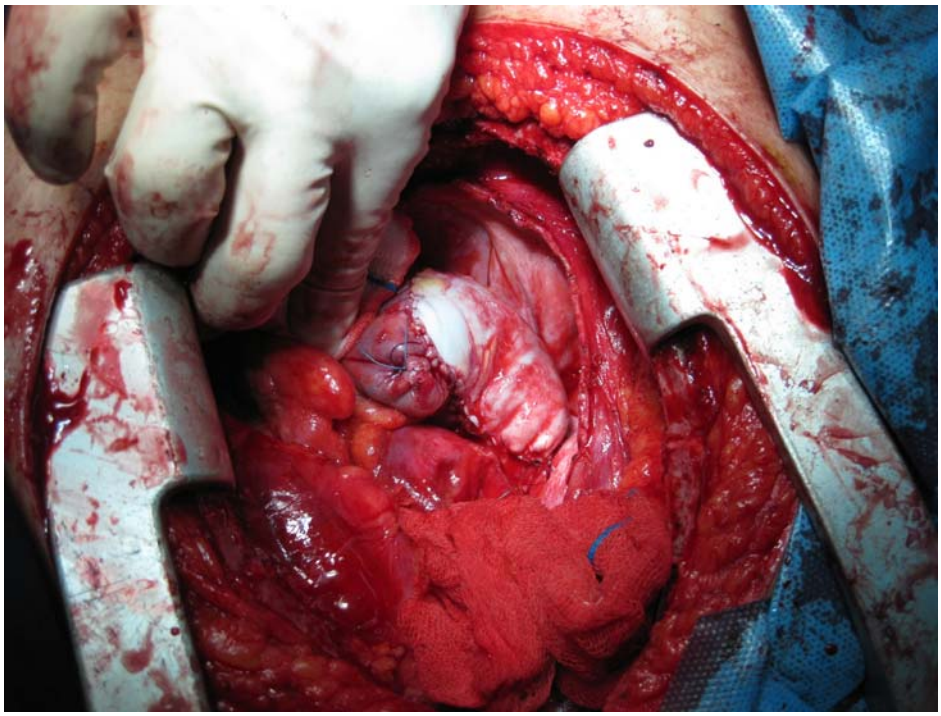

Figure 1. The tumor was completely excised together with the lateral atrial wall that was reconstructed with an autologous pericardial patch

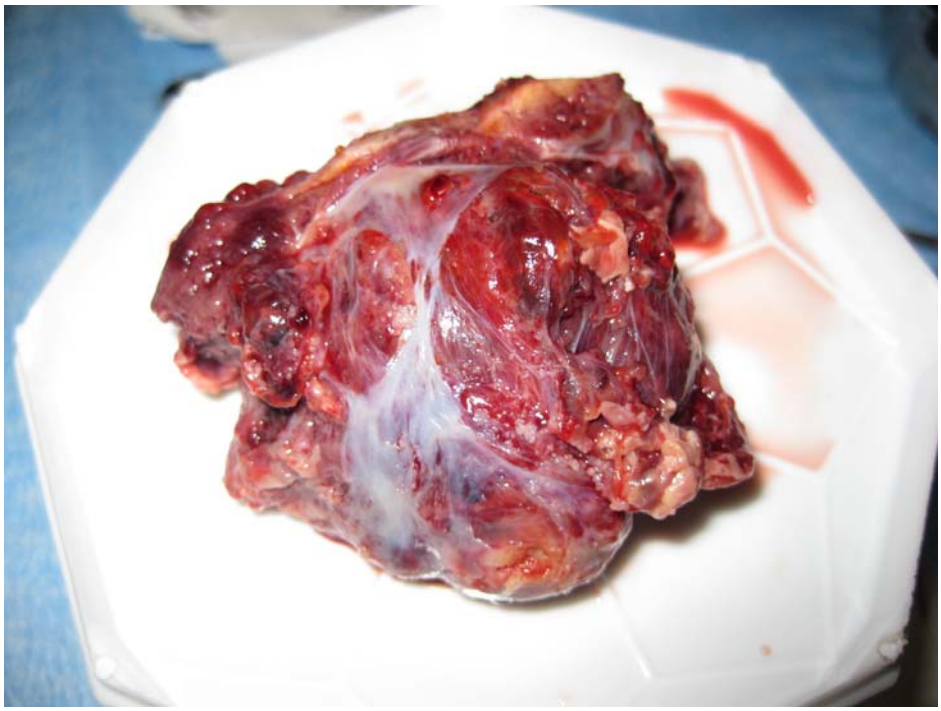

Figure 2. Macroscopic appearance of tumor.

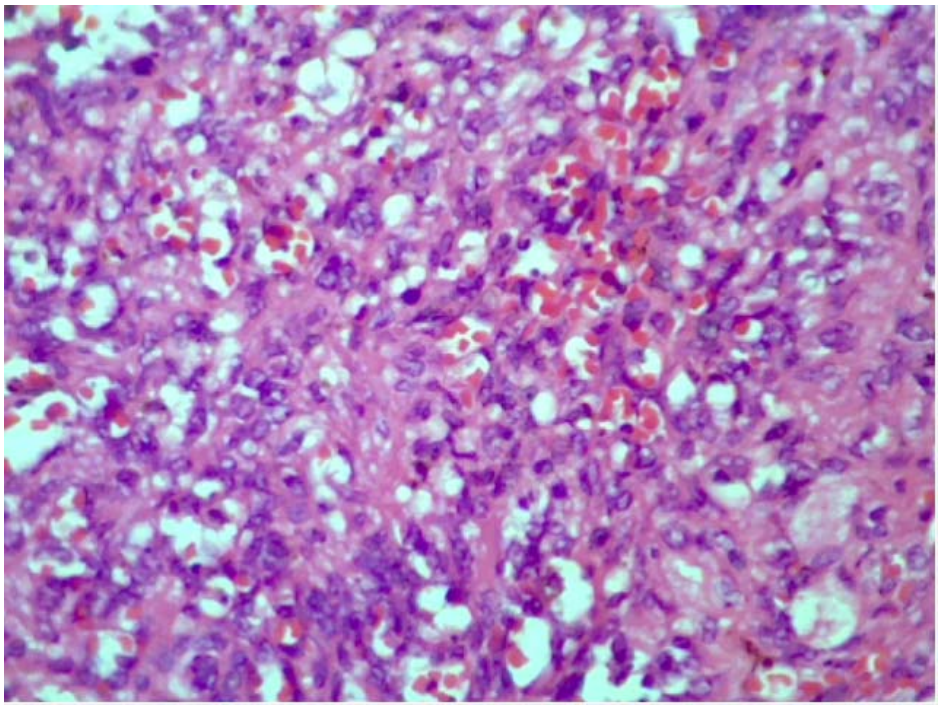

Figure 3. Shows microscopic view of malignant angiosarcoma cells.

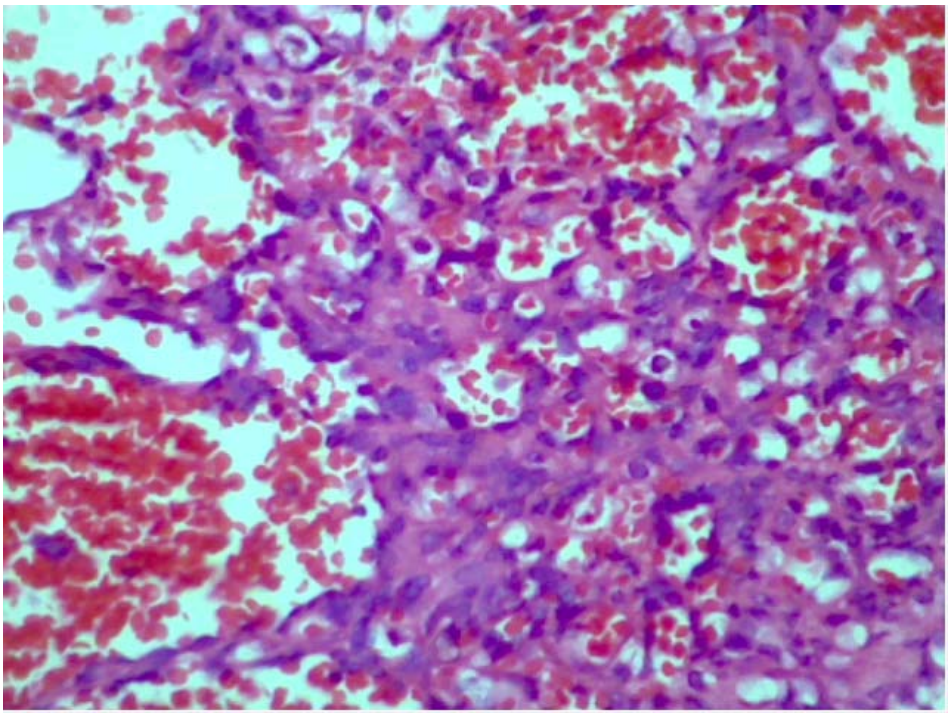

Figure 4. Shows microscopic view of malignant angiosarcoma cells with hyper vascular pattern.

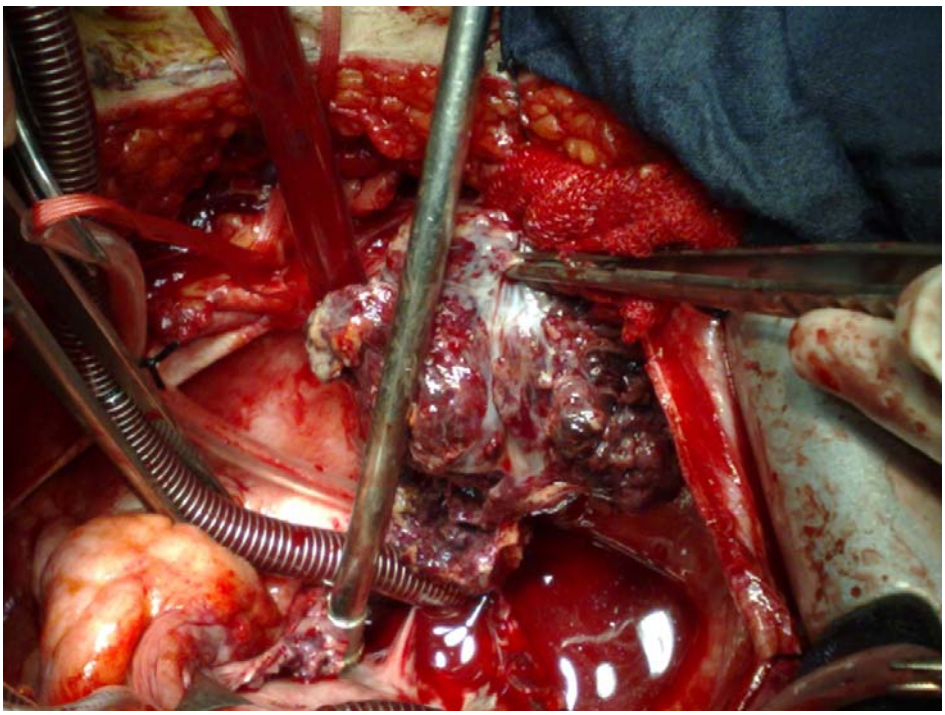

Figure 5. Intraoperative appearance of tumor.

However a special technique had to be used to control embolization which includes: 1) retrograde cannulation of inferior vena cava (IVC) through the femoral vein to avoid any fragmentation of the tumor. 2) institution of cardiopulmonary bypass (CPB) by superior vena cava (SVC) cannulation after opening of right

atrium, under direct vision, and with suction device, in order for IVC to be cannulated (10,11). In this case we had employed the second approach for the IVC cannulation. There was no evidence of tumor embolization during or after the operation. Resection of tumor constitutes the definitive treatment for cardiac angiosarcoma. In this case we resected the right atrial wall and constructed it with a fresh pericardial patch (Figure 1).

Complete resection of the right atrial wall at the site of tumor attachment is necessary to prevent recurrence of the tumor; however there is no evidence in the literature that radical resection of the atrial wall reduces the possibility of recurrences (12).

Discussion

Primary cardiac tumors are uncommon with an incidence of 0.001 and 0.28% in a reported series (13). Cardiac angiosarcoma is the rarest of cardiac tumors comprising approximately 0.0025% of all malignant cardiac tumors. In adults, approximately only 3/4ths are located in the right atrium and 0.25% in other locations (14). An association of right atrial angiosarcoma, pericardial effusion, and low cardiac ejection function is not reported in the medical literature to date. No case of exfoliated right atrial angiosarcoma associated with massive pericardial effusion and severe myocardial dysfunction has been reported in the literature. Most were in a single case report of the three patients we reviewed, and none were complicated by the right atrial wall invasion and tamponade, thus the incidence of the coexistence of these three lesions remains unclear (15,16).

The most common symptom of angiosarcoma is congestive heart failure followed by embolization. These symptoms are usually based on the location of the tumors; vary with the size, shape and physical activity of patients. In this tumor, the presentation may include edema hepatomegaly, and pulmonary embolization also occurs. Other clinical symptoms are fatigue, malaise, low grade fever and weight loss (17,18). Recent progress in diagnostic modalities in cardiology including echocardiography, computed tomography and magnetic resonance imaging which is now allowing diagnosis of primary cardiac tumors without cardiac catheterization or angiography (19). In this case, echocardiography readily showed a right atrial mass lesion. In the previously reported three cases of right atrial angiosarcoma, the echocardiography did not detect a pericardial effusion with global hypokinesia .Excision of right atrial sarcoma using CPB which has been established with generally good clinical results. However, we must use a special technique to control embolization that includes: 1) retrograde cannulation of IVC through femoral vein to avoid any fragmentation of the tumor. 2) Institution of CPB by SVC cannulation and after opening of right atrium, under direct vision, and with suction device, IVC can be cannulated. In this case, we employed the second approach for the IVC cannulation. There was no evidence of tumor embolization during or after the operation. Resection of tumor constitutes the definitive treatment for cardiac angiosarcoma. In this case, we resected right atrial wall and constructed it with fresh pericardial patch (Figure 1).

However complete resection of the right atrial wall at the site of tumor attachment is necessary to prevent recurrence of the tumor but there is no evidence in the literature that radical resection of the atrial wall reduces the possibility of recurrences (20). Angiosarcoma tends to have a broad based attachment to the right atrial wall, as in this case, and the tumor was resected with underlying atrial wall.

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