

## Primary angiosarcoma of the right atrium: Case report

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**Key words:** Primary angiosarcoma

### Abstract

Primary tumors of the heart are rare and angiosarcoma is the most common primary cardiac malignant tumor.

We present a 45-year-old lady admitted on November 2015 to the Coronary Care Unit at Hawler Teaching Hospital with shortness of breath for one month with clinical features of right-sided heart failure. Transthoracic echocardiography showed large mass in the right atrium. Computed tomographic pulmonary angiography confirmed a large atrial mass with direct extension to the superior and inferior vena cava up with extensive thrombosis involving the right subclavian vein and internal jugular vein in addition to a thrombus occluding the distal branch of the proximal segment of both right and left pulmonary artery.

Open-heart surgery done at Hawler Cardiac Center after obtaining informed consent from the patient. Surgery revealed a big solid tumor in the right atrium protruding to the right ventricle through tricuspid valve with extension to superior and inferior vena cava. De-bulking of the tumor was done but the evolution was marked by sudden death intra-operatively. The pathological and immune histochemical examination revealed a primary cardiac angiosarcoma.

**Conclusions:** Primary atrial angiosarcoma may present late with clinical features of right-sided heart failure and may be associated with vascular metastasis as well as pulmonary embolism.

### Introduction

Primary cardiac tumors are rare entities (0.0017–0.003%) of routine autopsy studies,<sup>1</sup>. Angiosarcoma is the most frequent primary malignant cardiac tumor (31% of all the malignancies)<sup>2</sup>.

Primary cardiac angiosarcoma (PCA) mostly arise in the right atrium and have a tendency to occur in the third to fifth decade and more commonly in males<sup>3</sup>. They

frequently extend to the pericardium, vena cava, or tricuspid valve, causing tamponade and/or heart inflow obstruction with superior vena cava syndrome.

The authors describe a case of large primary angiosarcoma located in the right atrium in a 45-year old lady with direct vascular metastasis. To our knowledge this association has not previously been described.

### **Case presentation**

A 45-year-old lady with no previous co-morbid illness presented to the Coronary Care Unit (CCU) of Erbil Teaching Hospital with gradual shortness of breath for one month. There was no history of alcohol ingestion, neither was there any history of cigarette smoking.

On examination the patient was conscious oriented to time and place with mild jaundice, not cyanosed, she had a tachycardia of 105 bpm regular, blood pressure of 110/70 mmHg in both arms, respiratory rate of 26/min, a temperature of 36.5°C. and oxygen saturation of 90% on room air. Bilateral pitting leg edema up to the knees. Cardiovascular examination revealed regular heart sounds with no murmur, a slightly raised jugular venous pressure, and pulses of good volume.

Examination of the chest revealed stony dull percussion on the right lower zone and decreased vesicular breath sound on the right lower zone. The abdomen was moderately distended, soft, not tender, no organomegaly and no other remarkable finding on clinical examination.

The resting electrocardiography (ECG) showed sinus tachycardia, right axis deviation, low voltage and no ischemic changes.

The hemoglobin was 13.7gm/dl and the white blood cell count was  $8.5 \times 10^9/L$ , with lymphocyte count of 22.3%, neutrophil count of 71%, and platelet count of  $205 \times 10^9/L$ , normal renal function test. Total and direct bilirubin levels were elevated at 3.8 mg/dl and 2.3mg/dl respectively, Aspartate Transaminase, Alanine Transaminase, Alkaline Phosphatase were all elevated at 51.2 IU/L, 47.9 IU/L, 1067IU/L respectively (Normal being 10-50 IU/L, 10-45IU/L and 40-125IU/L respectively. Total protein was 6gm/dl and albumin level of 3.5gm/dl. Normal level of both prothrombine and partial thromboplastin time. Viral were negative.

Chest x ray showed slight cardiomegaly (cardiothoracic index of 0.58) with obliteration of right costophrenic angle.

Transthoracic echocardiography revealed markedly dilated right atrium containing large fixed mass with irregular vague borders (5cm long x 1.3cm maximum width) attached to the inter-atrial septum, shifting the septum toward the left atrium and partially obstructing the tricuspid valve (Figure-1), normal left ventricular size, wall thickness and global left ventricular systolic function with ejection fraction of 64%.

Abdominal ultrasound revealed mild ascites, no intra or extra hepatic biliary dilatation, no organomegaly or other remarkable findings.

Computed tomographic (CT) pulmonary angiography confirmed the large atrial mass with direct extension to the superior and IVC up to the level of renal veins as well as extensive thrombosis involving the right subclavian vein and internal jugular vein ((Figure-2). The CT pulmonary angiography also showed a thrombus occluding the distal branch of the proximal segment of both right and left pulmonary artery. (Figure-3). Mild pleural effusion was seen on the right side of the chest.

The differential diagnosis was primary tumor of the right atrium, mostly expected to be malignant with vascular metastasis into the superior and IVC with sub-massive pulmonary embolism.

The patient received oxygen therapy and low-molecular-weight heparin (Enoxoparin, given subcutaneously, 8000 IU twice daily).

The patient referred next day to Hawler Cardiac Center and a team including cardiologist, cardiovascular surgeon and anesthesiologist was built. After obtaining the patient informed consent, open-heart surgery was done.

Cardiopulmonary bypass was instituted via cannulation of left common femoral vein and innominate vein as the venous line and the ascending aorta as the arterial line, there was severe adhesion between the heart and the pericardium, adhesiolysis was done.

There was a big solid tumor in the right atrium, which has a wide base arising from infero-lateral wall of right atrium and almost filling the lumen of right atrium, invading the lumen of SVC, protruding to right ventricle through tricuspid valve and heavily

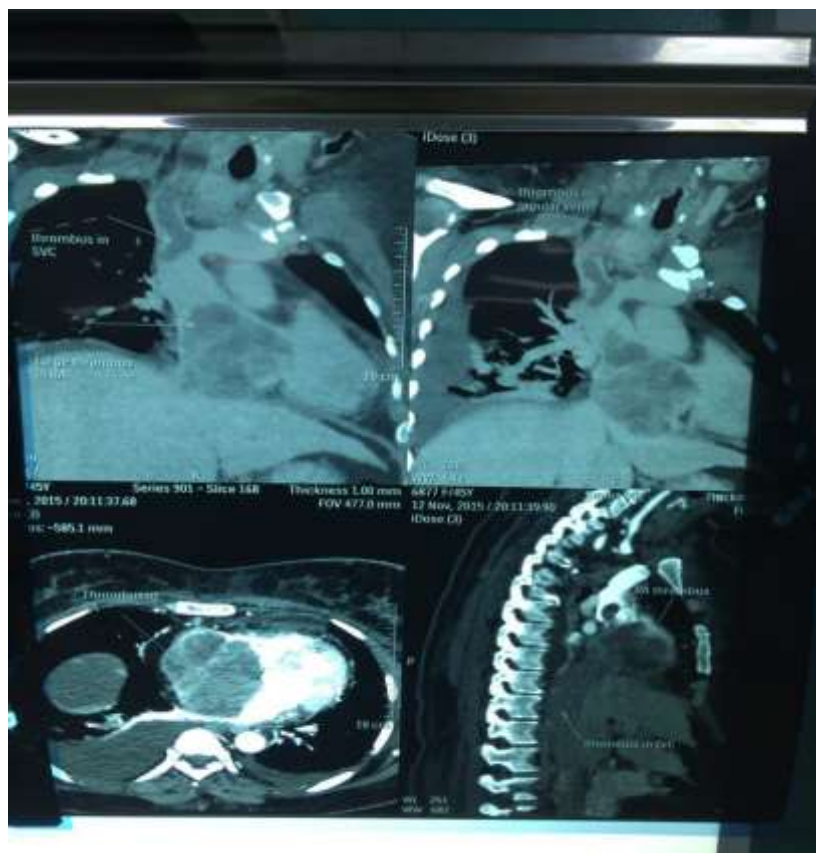
infiltrating the wall of IVC and sub totally occluding the lumen of IVC.

De-bulking of the tumor was done (Figure-4), there was restoration of blood flow through SVC, IVC, and tricuspid valve. Closure of Right atrium was done .the patient was gradually weaned from bypass machine however; the evolution was marked by the sudden death of the patient intra-operatively.

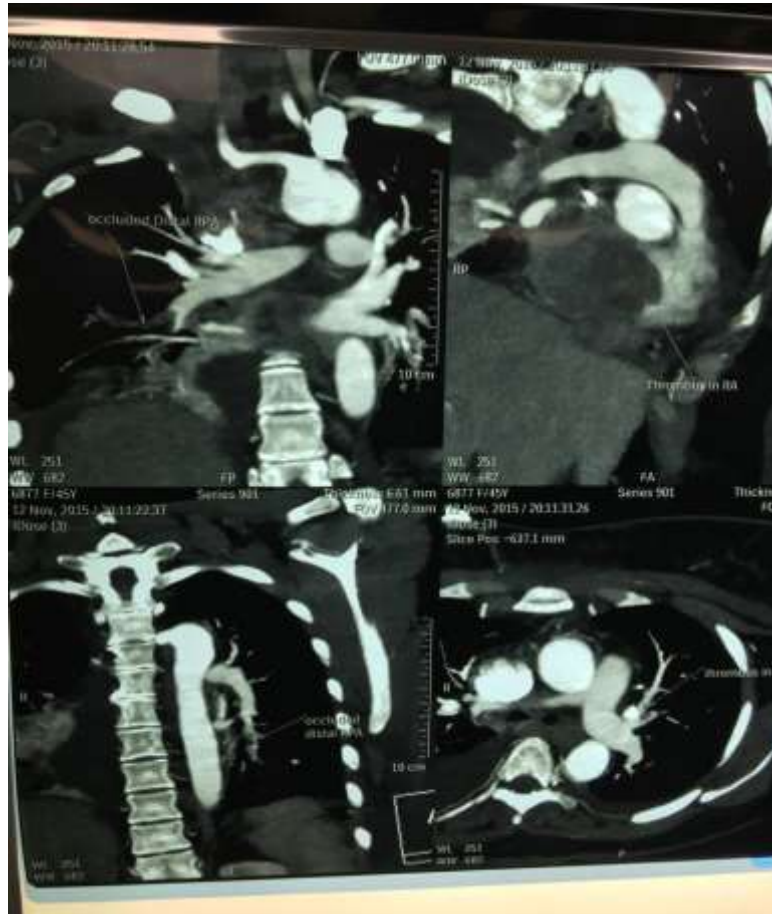
The gross pathology of the specimen showed two pieces of tissue measured 5 cm in aggregate, gray brown in color. The histopathological examination of the biopsy material obtained revealed primary angiosarcoma of the right atrium. The microscopic finding stained by Hematoxylin & Eosin (H&E) revealed a complex anastomosing vascular channels ,blood filled spaces lined by atypical endothelial cells with enlarged hyperchromatic nuclei with many mitotic figures and wide area of necrosis, the tumor is suggestive of primary cardiac angiosarcoma grade III (Figure-5,A) and confirmed by immune-histochemical staining which demonstrate their endothelial origin and aid in the diagnosis as a panel of Antibodies that include CD31 and CD34 were positive (Figure-5,B), vimentin positive (Figure-5,C).



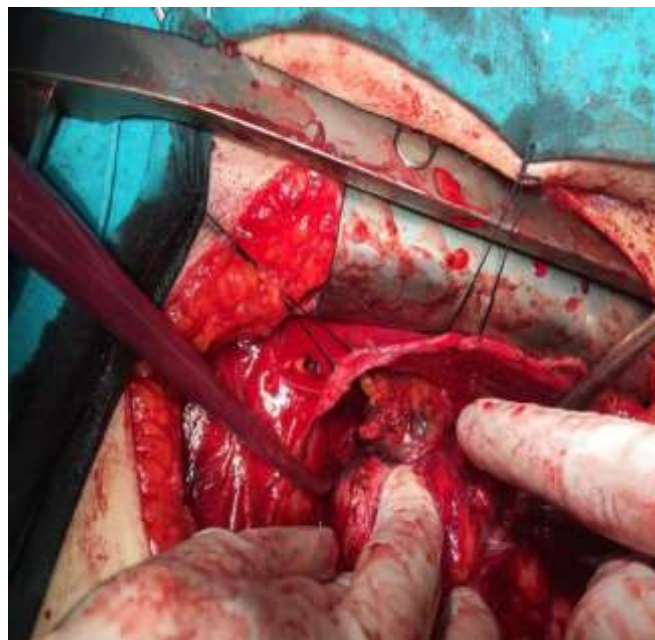
**Figure 1: Transthoracic echocardiography showing large right atrial mass protruded through the tricuspid valve toward the right ventricle**



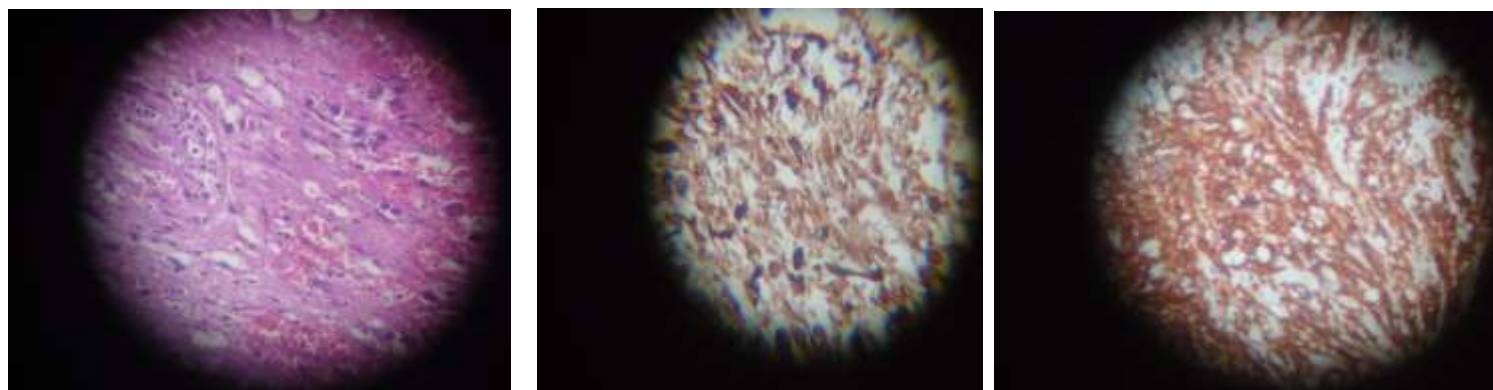
**Figure-2:** Computed tomographic pulmonary angiography showing large right atrial mass with extension to the internal jugular vein, superior and inferior vena cava.



**Figure-3:** Computed tomographic pulmonary angiography showing distal pulmonary occlusion



**Figure-4:** Surgical removal of right atrial mass



A

B

C

**Figure-5:**

A: (H&E) stain showing anastomosing vascular channels lined by malignant endothelial cells.(X400).

B: CD 31 positive (immunohistochemichemical staining).(X 400).

C: Vimentin positive (Immunohistochemical staining).(X 200).

## Discussion

Primary malignant neoplasm of the heart are exceptionally rare, of these primary cardiac angiosarcoma is the most common<sup>4</sup>.

Our patient presented with clinical features of right-sided heart failure manifested as elevated JVP, bilateral leg oedema, hepatomegaly and mild ascites. 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 tumour, the presentation may include edema hepatomegaly, and pulmonary embolization ,other clinical symptoms are fatigue, malaise, low-grade fever and weight loss<sup>5,6</sup>.

The differential diagnosis of a right atrial mass includes benign entities such as myxoma and thrombus and malignant causes such as metastatic involvement of the heart, primary cardiac angiosarcoma and other sarcomas, pericardial mesothelioma, and primary cardiac lymphoma<sup>7</sup>.

The absence of any other primary or metastatic lesions on the initial work-up suggests

that the tumor was a primary cardiac tumor of the right atrium with a rapid metastatic progression, seventy-five percent of PCA occur in the right heart especially in the right atrium<sup>8</sup>

Our patients presented with vascular metastasis, which explain the aggressive behavior of angiosarcoma and the high incidence of metastasis at the time of diagnosis with bad prognosis.

Echocardiography confirms the diagnosis of a cardiac mass. Computed tomography scanning has been a valuable adjunct to echocardiography. Magnetic resonance imaging is rapidly becoming the imaging modality of choice for evaluating pericardial and cardiac tumors due to its accurate evaluation of the mass and extension into neighboring structures.

Treatment options are limited. Results of surgical resection alone have been discouraging because most patients have advanced disease with a mean survival of nine months, depending on whether adjuvant treatment with radiation and/or chemotherapy was given<sup>9</sup>. Angiosarcoma responds poorly to chemotherapy<sup>6</sup>.

## **Conclusions**

Primary atrial angiosarcoma may present with clinical features of right-sided heart failure and may be associated with vascular metastasis. Multi-imaging modalities like CT angiography, MRI, transthoracic echocardiography and trans-esophageal echocardiography are mandatory for early diagnosis.

## **Conflicts of interest**

The authors report no conflicts of interest.

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