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Primary Cardiac Angiosarcoma in a Middle Aged Woman

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Introduction: Primary cardiac angiosarcoma is the most common primary sarcoma in adults between the 3rd and 4th decades of life. Nearly 90% of angiosarcomas occur in the right atrium, which is responsible for the late onset of symptoms.
Case Presentation: We presented a 56-year-old woman admitted to our center with lung emboli symptoms. Transthoracic and transesophageal echocardiography (TTE and TEE) demonstrated very large size (more than 10 cm diameter) multilobulated mass with mobile particles extended from the right atrium to the right ventricle and the right ventricular outflow tract which destructed the right atrium (RA) wall and penetrated to the pericardial space.
Conclusions: Unfortunately the tumor was unresectable and just an incisional biopsy was performed. She received chemotherapy as palliative care.

Keywords: Cardiac Tumors Sarcoma; Angiosarcoma; Echocardiography

1. Introduction

Primary cardiac tumors, with the incidence of 0.001 - 0.03% in autopsies, are benign in approximately 75% of cases. Among remaining 25% malignant tumors, sarcomas are the most common types (1).

Primary cardiac angiosarcoma is the most common primary sarcoma in adults between the 3rd and 4th decades of life. Nearly 90% of angiosarcomas occur in the right atrium, which is responsible for the late onset of symptoms (2, 3). Due to this latency, 66% to 89% of patients present with evidences of metastasis at initial presentation, which is associated with poor prognosis (4). We presented a 56-year-old woman presented to our center with lung emboli symptoms.

2. Case Presentation

A 56-year-old woman presented to our center due to dyspnea at rest (New York heart association [NYHA] functional class III-IV), tachycardia, tachypnea and hemoptysis. Physical examination showed a systolic murmur of grade II/VI at left sternal border, sinus tachycardia and tachypnea.

A 12-lead electrocardiogram showed normal sinus rhythm with normal axis and no Q wave or ST-T changes (Figure 1). Chest X- ray showed marked cardiomegaly (cardiothoracic ratio = 70%) and right costophrenic angle was blunted (Figure 2). In lab data, HCT = 8.5, CRP = + and ESR = 50 were demonstrated.

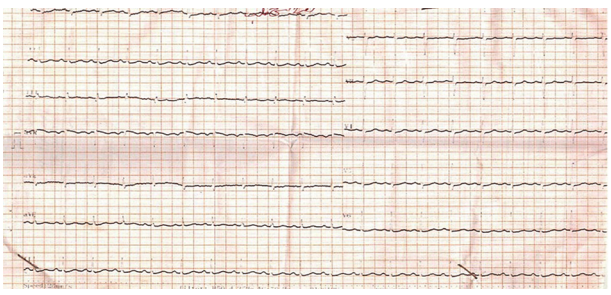

Figure 1. 12-Lead Electrocardiography of the Patient Shows Normal Sinus Rhythm

Her symptoms started six months prior to her presentation to our center. She had fever, night sweating, weight loss and anorexia in the recent weeks. Six months before admission, she had a thoracic back pain in the right paravertebral region. It was a localized intermittent pain with no exacerbation on exertion. She was initially evaluated in another center. First, a chest x ray was performed showing right sided pleural effusion.

Further investigation including diagnostic thoracocentesis, bronchoscopy and Bronchoalveolar lavage (BAL) showed an inflammatory process and could not establish a definite diagnosis.

The patient underwent colonoscopy and abdominal sonography without any characteristic findings. An angiography was performed for her but no specific findings were reported. After ten days, patient was discharged

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with recommendation for outpatient following to define the etiology of this inflammatory process.

Transthoracic and transesophageal echocardiography (TTE and TEE) demonstrated very large size (more than 10 cm diameter) multilobulated mass with mobile particles extended from the right atrium to the right ventricle and the right ventricular outflow tract which destructed the right atrium (RA) wall and penetrated to the pericardial space (highly suspicious of intracardiac malignant tumor). The tumor obstructed inflow of tricuspid valve. Moderate to severe tricuspid regurgitation was seen (Figure 3).

The previous angiography was rechecked and a mass with vascular pattern in the right atrium (tumor blanch view) was reported. Moreover, it was detected that the left circumflex artery origin was in the right coronary cups (Figure 4).

Lung and mediastinum (computed tomography) CT was performed for patient and demonstrated a large size tumoral lesion (approximately 15 × 8 cm) that its origin was in posterior wall of the right heart cavities. Multiple metastatic lesions in subpleural space of both lungs, multiple lymph nodes (approximately size = 10 cm) in mediastinum and right axillary and one lymph node in aortocava region (approximately = 18 cm) were seen. Those findings were highly suspicious of malignant cardiac tumor (high probable diagnosis was angiosarcoma and low probable diagnosis was rhabdomyosarcomas) (Figure 5).

The patient was in a poor condition, she had symptoms of metastatic emboli, so she underwent surgery. Surgeon reported one polypoid mass that exited from pericardium with severe adhesion to pericardium and cardiac wall and extended from RV to hilum of the right lung. In appearance, tumor had vascular nature. Unfortunately the tumor was unresectable and just an incisional biopsy was performed.

Pathology reported a malignant mesenchymal neoplasm composed of poorly differentiated atypical cells with vascular channels and slit-like spaces between them containing numerous RBCs. Tumor cells had spindle or polygonal vesicular nuclei with frequent mitosis. Large areas of necrosis and hemorrhage were also reported. To confirm the diagnosis, immunohistochemistry (IHC) was recommended. At the end, primary cardiac angiosarcoma was proved by IHC (Figure 6). She received a course of palliative chemotherapy at the hospital and set to continue her chemotherapy course with her oncologist.

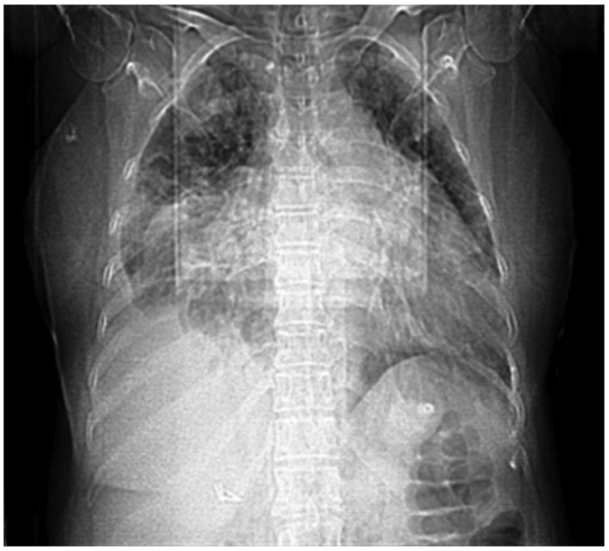

Figure 2. Chest X-Ray Demonstrating Increase of Cardio Thoracic Ratio and Costophrenic Angle Blunting

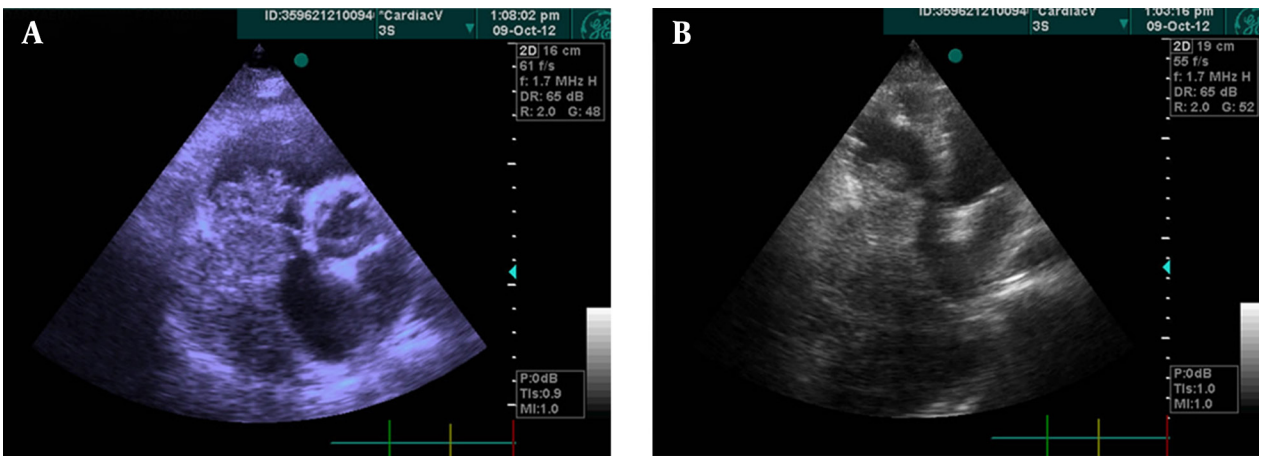

Figure 3. A, Transthoracic Short-Axis View Shows Large and Multilobulated Mass Extend From RA to RV and Destroyed Tricuspid Valve; B, Transthoracic Long Axis View Shows Pericardial Effusion and Thick Pericardium With Mass

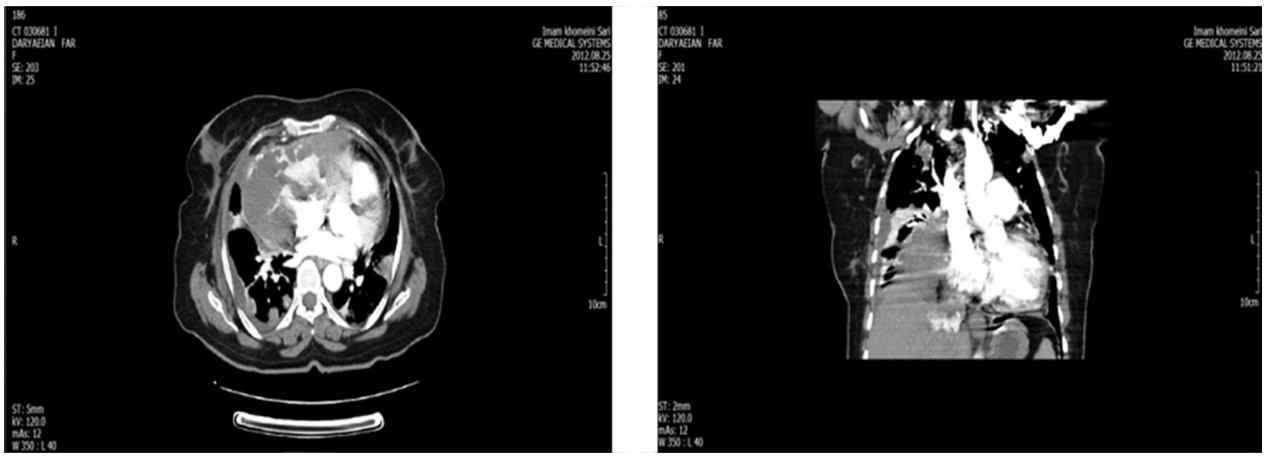

Figure 4. Coronary Angiography Shows Abnormal Origin of the Left Circumflex Artery From the Right Coronary Cups and a Tumor With Vascular Pattern in the Right Atrium

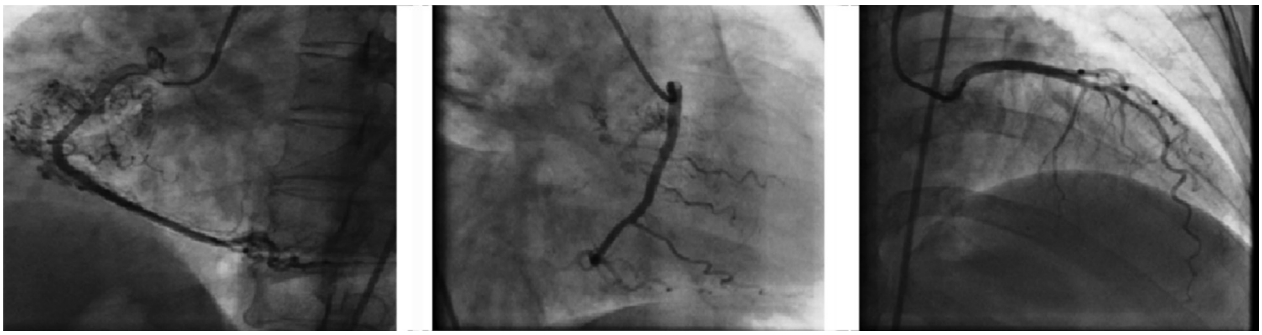

Figure 5. Lung and Mediastinum CT Shows Tumoral Mass in the Right Side Heart and Metastatic Lesions in Pleural Space

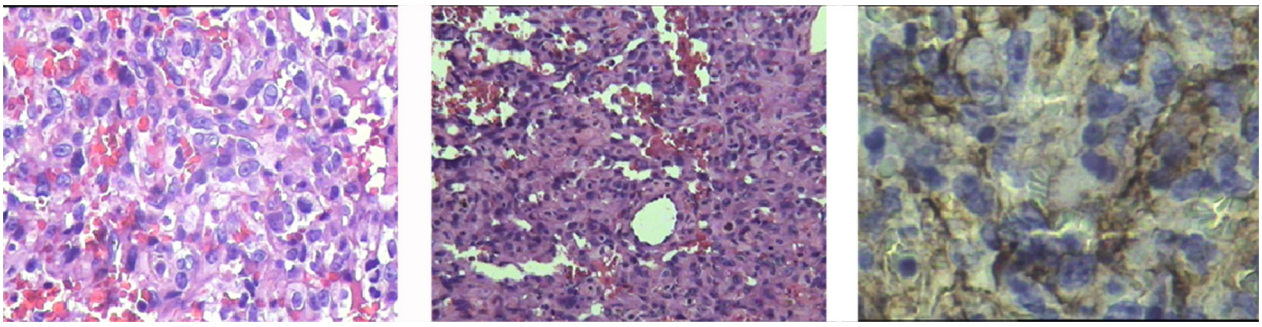

Figure 6. Immunohistochemistry of Tissue Sample

3. Discussion

Cardiac primary tumors are rare and nearly 75% of them are benign (1). Angiosarcoma is the most common type of primary cardiac tumors. Angiosarcoma is more prevalent in male between the 3rd and 5th decades of life (2, 3).

Patients with angiosarcoma usually present with dyspnea in 59 to 88% of cases. Other frequent symptoms include chest pain (mainly right) and heart failure. Pericardial effusion with or without tamponade, vena cava obstruction, pulmonary emboli, hemoptysis, anemia and cardiomegaly are less frequent (3, 4).

Tumor usually occurs in the right atrium (about 90%). Due to the late onset of symptoms and their unspecific patterns, most cases present at their advanced stages (2, 3).

Echocardiography, especially TEE, is the most valuable

tool for early detection of tumor location and its extension. Echocardiography may predict the tumor type on the basis of its appearance (5, 6).

CT scan and MRI are required for better diagnosis of tumor spread and evaluation of the disease systemically. Histopathology is the definite method to diagnosis the tumor. In our case, IHC proved the tumor as primary cardiac angiosarcoma (4, 5).

Our case presented to our center mainly because of a progressive dyspnea, tachycardia, tachypnea and hemoptysis. TTE and TEE showed a huge mass in the right atrium extended to the right ventricle and its outflow tract. The tumor destructed the right atrium wall and penetrated to pericardial space.

The treatment strategies for primary cardiac tumors include surgery, chemotherapy and radiotherapy. For patients with respectable tumor and/or without metastasis, complete surgical removal is the most appropriate method. However, even after a complete resection, the survival period for angiosarcomas is 6 - 12 months after surgery.

Chemotherapy is another treatment, which can be used as neoadjuvant, adjuvant and for metastatic diseases. For patients with unresectable mass and/or systemic metastasis, palliative chemotherapy is the best option (4).

Although our patient had multiple metastatic lesions in her lungs and multiple enlarged lymph nodes in mediastinum and right axilla and aortocava, a surgery was performed because of metastatic emboli symptoms. Unfortunately, the mass was unresectable and just an incisional biopsy was performed. Palliative chemotherapy was performed for her, after discharge she was referred to an oncologist for further treatment.

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Authors' Contributions

Rozita Jalalain: Study concept and design, Analysis and interpretation of data, Critical revision of the manuscript for important intellectual content, Administrative, technical, and material support, Study supervision. Farshad Naghshvar: Analysis and interpretation of data. Valiollah Habibi: Study supervision. Vahid Hakakain: Acquisition of data, Administrative, technical, and material support. Morteza Namazi: Acquisition of data, Administrative, technical, and material support.

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