Palliative Bypass for a Large Advanced Angiosarcoma of Right Atrium and Right Ventricle with Pericardial Effusion: Case Presentation

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Abstract

We present the case of an advanced and large cardiac angiosarcoma in the right atrium and right ventricle, which occupied the most part of the right heart and created a significant stenosis in the pulmonary circulation. Our patient was in very bad general condition with severe respiratory distress and had very unstable hemodynamic. Also, he experienced two episodes of CPR (cardiopulmonary resuscitation) before he was transferred to the operating room. Given the patient's poor condition and limited life expectancy, we performed a palliative bypass procedure, consisting of cavopulmonary anastomosis***(Iranian Heart Journal 2011; 12 (4):62-65*).**

Keywords: Cardiac tumor■ Angiosarcoma■Pericardial effusion

Primary cardiac tumors occur rarely, with a reported incidence of 0.03% to 0.05%, and the majority of the tumors are benign.[1] Cardiac myxoma is the most common of these primary cardiac tumors. Malignant tumors of the heart are more likely to be metastatic in origin than a primary neoplasm. Sarcomas are the most common type of primary malignant tumors of the heart. Rhabdomyosarcomas are most common in children and angiosarcomas are more common in adults.[2] Primary cardiac angiosarcomasare extremely rare. Most cases of cardiac angiosarcomas have metastasis to multiple organs at the time of diagnosis.[2, 3] Angiosarcomas most commonly arise from the right atrium.[3] Angiosarcomas have a tendency to occur in the third to fifth decade of life and are more common in males. Angiosarcomas are very aggressive, with a high incidence of metastasis at the time of diagnosis.[4]

We herein present an advanced angiosarcoma in the RT atrium and ventricle, which was operated on with palliative bypass to relieve a significant obstruction in the pulmonary circulation.

Case Report

We present the case of a 57-year-old woman with a 3-month history of dyspnea and chest pain. Onphysical examination, she had edematous extremities and probably ascites and bilateral diminish breath sound due to pleural effusion. In her evaluation in CXR, she had wide mediastinum and bilateral pleural effusion. In transthoracic echocardiography, there was massive pericardial effusion and a large (9 × 6 cm) mass in the RA. The mass was attached to the intra-atrial septum and projected into the RV cavity through the tricuspid valve, producing a gradient of 13 mmHg with diastolic RV outflow tract collapse and normal left ventricular function but with shift of Interventricular septum to the LT side(Fig. 1).

Received June. 2, 2011; Accepted for publication Feb. 29, 2012

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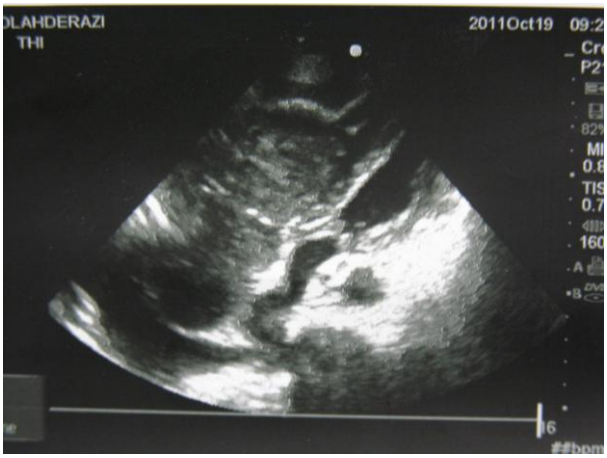

Fig. 1. A large Mass in RT Atrium and ventricle with deviation of septum to left side

In the evaluation period of the patient, she had respiratory distress with O2 saturation about 50%-60% and hemodynamic instability; and unfortunately, even with pericardiocentesis for the release of the pericardial effusion, the patient had sudden cardiac arrest, which responded to immediate CPR. Due to the patient's critical condition, we had no time for other imaging modalities like CT/MRI for a better evaluation of the cardiac mass and local invasion and probably distant metastasis. The patient was transferred to the operating room, where she was subjected to median sternotomy. There was massive pericardial effusion. Next, routine cardiopulmonary bypass was done. A mass (9 × 6 cm) was found to occupy most of the RA with extension to the RV through the tricuspid valve (Fig. 2).

The mass was heterogeneous in nature, with compression effect to the left side. This large mass significantly decreased the contractility of the RA and RV.

No mass was seen in the other cardiac structures.

Intraoperatively, the RA mass was found to have infiltrated the entire wall of the RA, tricuspid valve, and RV and RV outlet tract with near occlusion in the pulmonary circulation.

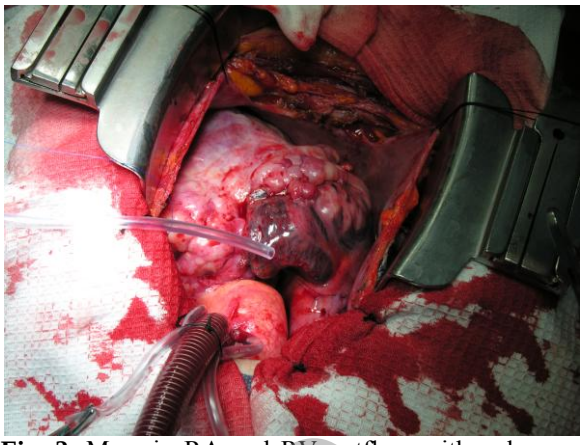

Fig. 2. Mass in RA and RV outflow with pulmonary circulation obstruction

Only the biopsy of the mass was possible; and then to relieve the pulmonary obstruction, palliative bypass anastomosis with superior vena cava (SVC) and RT pulmonary artery (RPA) with a Gortex shunt was done (Fig.3).

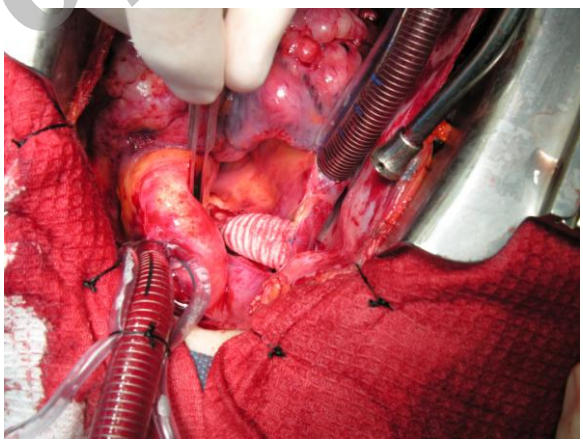

Fig. 3. Palliative bypass with RPA and SVC with a shunt

The patient was weaned successfully from cardiopulmonary bypass with inotrope support and acceptable O2 saturation (85%). She was discharged from the hospital one week later and in follow-up echocardiography, the shunt was patent. Pathologic examination confirmed the diagnosis of primary cardiac angiosarcoma.

Discussion

Cardiac primary tumors are rare in occurrence and approximately 95% of all primary malignant cardiac tumors are sarcomas. Angiosarcomas are the most common primary cardiac malignant tumor and are extremely rare, rapidly spreading vascular tumors. Most cases of angiosarcomas have metastasis to multiple organs at the time of diagnosis. Angiosarcomas are the most common malignant tumors of the heart and are characterized by rapid growth, local invasion, and distant metastasis. They are seen more commonly in males than in females, usually presenting between the third and fifth decade of life.[5,6] Malignant tumors are located mainly on the right side of the heart. Ninety percent of angiosarcomas are located in the RA, and there is a high incidence of pericardial involvement with pericardial effusion.[7] Angiosarcomas of the heart involve the right atrium almost exclusively, but they have been reported in the other cardiac chambers.[8] Angiosarcomas of the heart grow rapidly, usually within the myocardial wall, and they are characterized by a tendency to bleed and are often associated with pericardial effusion and cardiac tamponade. [9] Dyspnea is the most common presenting symptom; additional symptoms include atypical chest pain, hemoptysis, orthopnea, and nonspecific symptoms such as nausea, emesis, fever, and anorexia Transthoracic echocardiography is the screening modality of choice for the initial evaluation of patients with suspected cardiac neoplasms. Echocardiography can define the size, location, and mobility of the neoplasm in real time. Characteristics that show a malignant cardiac tumor on echocardiography include lack of a pedicle, broad base of attachment, and invasive growth pattern. [10] Although echocardiography is the simplest way to show the tumor, an additional imaging modality is CT/MRI.[11]

The treatment of cardiac angiosarcomas is very challenging and includes chemotherapy, radiation, surgical resections, and even transplantation.[12] Regardless of the treatment strategy, the overall survival for patients with cardiac angiosarcomas remains poor; the median survival with complete surgical excision is approximately 17 months and approximately 6 months in those in whom surgical resection cannot be achieved. [13] Angiosarcomas are traditionally associated with a poor prognosis. Surgical resection with or without adjuvant radiation or chemotherapy is the main treatment modality. Surgical treatment can palliate intracardiac obstruction. Chemotherapy after surgical resection has offered no increased survival benefit when compared to surgery alone.[14] Due to the rarity of this disease, there are no accepted treatment guidelines. A multidisciplinary approach, involving surgery, irradiation, adjuvant chemotherapy, and immunotherapy using interleukin-2, may offer hope for increased survival in selected patients.[15]

Conclusion

Primary cardiac angiosarcomas are the most common primary tumor of the heart. The common symptoms are dyspnea and chest pain. Diagnosis is with echocardiography and other imaging modalities like CT/MRI. Tumors have a tendency to rapid local invasion and distal metastasis at the time of presentation. Treatment, if possible, is a combination of surgery and chemotherapy and radiation but with poor prognosis.

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