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Angiosarcoma of the Right Ventricle: A Rare Encounter

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ABSTRACT

We report a case of angiosarcoma involving the right ventricle. The patient was seen in our Cardiology Department and subsequently referred to our unit for surgery. He gave a 1-week history of lethargy, chest pain, breathlessness on exertion, fevers, and night sweats. Echocardiography and computed tomography of the chest showed a large pericardial effusion with multiple densities, raising suspicions of a hemorrhagic effusion. Surgical exploration showed an epicardial mass. Histopathology revealed angiosarcoma.

INTRODUCTION

Sarcomas are malignant tumors of mesenchymal origin. They may present as fibrosarcoma, angiosarcoma, rhabdomyosarcoma, or lymphosarcoma, rarely as primary tumors of the heart. Of all primary cardiac tumors, 25% are malignant, mainly as sarcomas, the most common being angiosarcoma (33%). Primary angiosarcomas are aggressive, carrying a high mortality rate due to relapse and metastasis. They may arise within the heart or epicardium. Antemortem diagnosis is increasing with advances of diagnostic methods.

CASE REPORT

A 26-year-old man was admitted to a regional medical unit in Queensland, Australia, with a 1-week history of increasing lethargy, left chest and shoulder pain, cough with yellowish sputum, breathlessness, and episodes of sweats and fever. He had no history of trauma or medical problems but had been an abuser of intravenous drugs 4 years previously and was a heavy smoker. Physical examination revealed a temperature of 38.1°C, a regular pulse rate of 120/minute, BP 140/80mmHg. The jugular venous pulse was not elevated; there was no evidence of cyanosis or splinter hemorrhages and no peripheral edema. Heart auscultation revealed a pericardial rub and no murmurs. Electrocardiogram showed sinus tachycardia. A chest x-ray showed a large cardiac silhouette. Transthoracic echocardiography showed a 3 to 4 cm pericardial effusion with multiple densities, suggesting possible hemorrhagic clots and a mass over the right ventricle (Figure 1 ◎). There was neither tamponade nor vegetation. Heart valves were normal and ejection fraction was 45%. Computed tomography confirmed the effusion and showed a mass over the right ventricle (Figure 2 ◎). The lungs and mediastinum were normal. Blood tests showed a hemoglobin of 100g/L; white cell count was 25.2×10^9/L (neutrophils, 20.87×10^9/L). Serology for Q fever, mycoplasma pneumoniae, antinuclear antigen, and extractable nuclear antigens produced negative results.

Surgical drainage was performed through a left anterior thoracotomy. This procedure revealed extensive, well-formed, hemorrhagic, and fibrinous pericardial clots, which were extremely difficult to evacuate, and so a median sternotomy was performed. Opening of the pericardium revealed a 3 × 2 cm firm mass on the right ventricle with extensive pericardial adhesions (Figure 3 ◎). The mass was dissected and sent for histopathological study (Figure 4 ◎). A pericardiectomy was performed. Samples of tissue and clots were sent for culture, virology, and acid-fast bacilli test, which had negative results. The patient was transferred to the intensive care unit on inotropic support and ventilation.

Two days later, the patient suffered a drop in blood pressure, a raised jugular venous pulse, and decreased urine output consistent with tamponade. Echocardiography showed reaccumulation of clots and restricted filling of both ventricles. The chest was reopened and evacuation of clots was performed. He made a good subsequent recovery in intensive care and the ward and was discharged home after 17 days. Histopathology showed interconnected ectatic vascular spaces lined by highly atypical cells with large pleomorphic nuclei and mitotic figures. The cells stained positive for CD34, CD31, and factor VIII R antigen and negative for CAM 5.2, EMA, S-100, and SMA. Findings were consistent with angiosarcoma. The pericardium was also involved. A CT of the chest 1 month after discharge revealed a lesion on the left lung lingula. The patient underwent radiation therapy.

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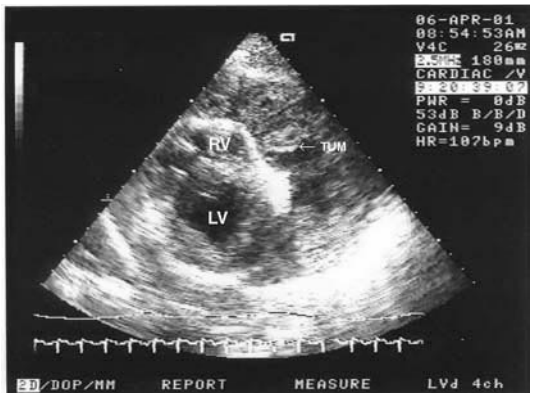

Figure 1. Echocardiography picture showing a mass over the right ventricle. LV indicates left ventricle; RV, right ventricle; TUM, tumor.

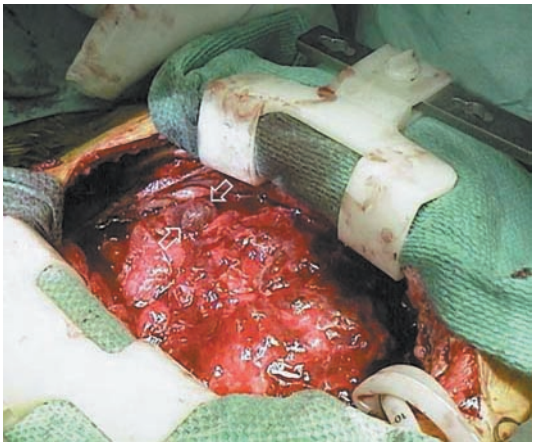

Figure 3. In-theatre picture showing a 3 × 2 cm mass on the right ventricle.

DISCUSSION

Primary cardiac tumors are extremely rare. Approximately 75% to 80% are benign; most are myxomas. Angiosarcomas represent approximately 33% of primary malignant cardiac tumors. They are more common in men, with a ratio of 2-3:1; the reported mean age is 41 years [Burke 1992]. The right side of the heart, primarily the right atrium, is more involved. Patients usually present with congestive heart failure, arrhythmia, lethargy, and left chest pain. Other presentations include fever, sweats and pericardial effusions, usually hemorrhagic.

Clinical onset is rapid, with progressive deterioration due to aggressiveness of the tumor and distant metastases (lungs, liver, or brain) by the time of diagnosis. Metastases have been reported in 66% to 89% of patients, with a mean survival of 3 to 6.6 months [Janigan 1986, McFadden 1997]. Advances in technology have allowed antemortem diagnosis and treatment strategies to be made. Echocardiography and computed tomography are important in showing epicardial or myocardial involvement. Histopathology via a thoracotomy remains the mainstay for a definitive diagnosis. Recently, transvenous endocardial biopsy has been employed.

Management is difficult due to aggressiveness and poor prognosis of the tumor. Extensive resection has been proposed, with pre- and postoperative radiotherapy and/or combination chemotherapy (cyclophosphamide, doxorubicin, vincristine, and mitomycin). Because of poor prognosis and quality of life despite management regimens, many reports have not justified subjecting patients to major surgery [Biniwale 1999].

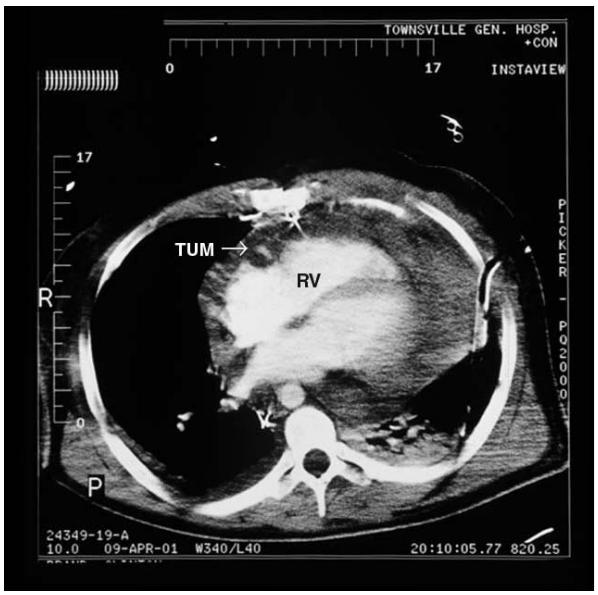

Figure 2. Computed tomography of the chest showing pericardial effusion and a tumor over the right ventricle. RV indicates right ventricle; TUM, tumor.

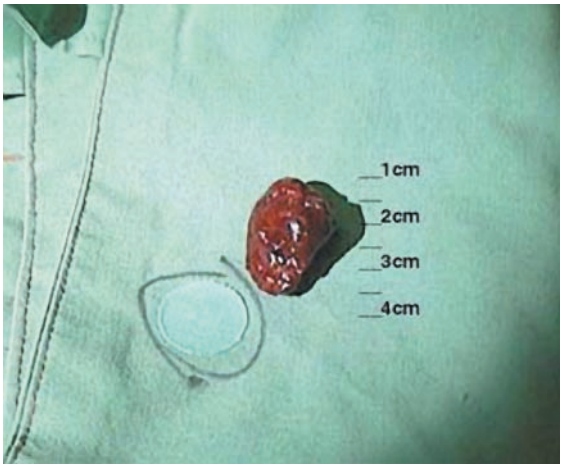

Figure 4. In-theatre picture showing the tumor removed from the right ventricle.

Cardiac transplantation, although technically successful, remains a limited option; the maximum survival has been 8 to 9 months, and malignant cardiac tumors are considered by most to be a contraindication to transplantation [Crespo 1993]. Goldstein et al [1995] published outcomes of 8 patients who underwent orthotopic cardiac transplantation for primary cardiac tumors (4 of which were malignant) with no distant metastasis. We do not believe that our patient was a candidate for transplantation, because he had shown evidence of distant metastasis to the pericardium and later to the lung. The issue of transplantation remains debatable by some authors.

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