

## CARDIAC ANGIOSARCOMA PRESENTING AS RIGHT HEART FAILURE SECONDARY TO PULMONARY VASCULAR CARCINOMATOSIS

Ahmed Bahammam, MD, MRCP

Angiosarcoma is a rare tumor, representing less than 2% of all soft tissue sarcomas.<sup>1</sup> Information about its clinical features is limited. It usually affects adults and can develop from the endothelium of lymphatics or blood vessels.

### Case Report

A 29-year-old male nonsmoker presented initially to his family physician with a febrile illness and vague retrosternal pain, which was treated with a course of oral antibiotics as a possible acute bronchitis. The patient's condition did not improve and he developed anorexia, progressive weight loss, progressive dyspnea, dry cough, lower limb swelling, and right-sided abdominal pain. He was referred to our hospital for further management six weeks after his initial presentation.

On admission, the patient was afebrile with severe respiratory distress. Respiratory rate was 50/min., heart rate 120/min. regular, and blood pressure was 100/60. He was cyanosed with lower limb edema and mild jaundice. There was no clubbing or palpable lymph nodes, but jugular venous pressure (JVP) was elevated. Breath sounds were diminished, with coarse inspiratory crackles in the mid and lower zones bilaterally. Right-sided S<sub>3</sub> gallop was audible, but there were no murmurs or pericardial rub. The patient had some tenderness in the right hypochondrium, but no palpable organomegaly. He had no clinical signs of superior or inferior vena caval obstruction, and the rest of the examination was normal.

Chest x-ray showed confluent nodular infiltrates throughout the lung fields, with a globular bulge at the right heart border (Figure 1). Electrocardiogram showed sinus tachycardia, with nonspecific T-wave changes. Arterial blood gases on 6-liter nasal cannula showed: pH 7.52; PO<sub>2</sub> 53; PCO<sub>2</sub> 24; and HCO<sub>3</sub> 20.5. Blood tests showed mild impairment of liver function, but were otherwise nonspecific. Transthoracic echocardiogram was done but it was suboptimal and technically difficult, as the patient was unable to stay supine. The study showed a

small amount of pericardial fluid with normal cardiac size and normal left ventricular systolic function. The right heart function was difficult to assess, but there was no evidence of outflow obstruction. No cardiac or juxtacardiac masses were visualized. Central venous pressure was 15-19 mm Hg. Due to respiratory failure, the patient was intubated and mechanically ventilated. Pulmonary artery catheter was not inserted. Fibro-optic bronchoscopy showed some blood in the bronchi bilaterally, but no endobronchial lesion. Bronchoalveolar lavage showed no microorganisms or malignant cells. Open lung biopsy was performed and the diagnosis of angiosarcoma was made. The patient died two days after admission.

At autopsy, the heart weighed 1450 g (normal average weight 325 g). Globoid enlargement of the right atrial wall was seen but it did not obliterate the cavity (Figure 2). The tumor replaced the atrial walls except the interatrial septum. The tumor penetrated the pericardial cavity, which was obliterated by a diffuse and organizing pericarditis, encasing the right and left ventricles. There were no valvular lesions and no intracardiac shunt. The histological features of the tumor were variable, with areas showing virtually solid growth of pleomorphic spindled and epithelioid cells to geographic aggregates of tightly clustered tumor cells bordering blood-filled spaces. The main pulmonary arteries were not involved, nor were the segmental branches. The gross pathology of the lungs showed multiple metastases of less than 1 cm in diameter. Microscopy revealed multiple bilateral pulmonary metastases, with numerous occlusive tumor emboli plugging the pulmonary vasculature (Figure 3). There was no evidence of active pulmonary parenchymal infection. The liver had passive congestion but no metastasis.

### Discussion

Although cardiac angiosarcoma is the most common malignant tumor of the heart,<sup>2</sup> it still occurs only rarely. It usually affects the right side of the heart, especially the atrium,<sup>1,3,4</sup> and presents with chest pain, cough, syncope or dyspnea in young and middle-aged patients. The diagnosis is usually made postmortem,<sup>3</sup> probably due to the rarity of this tumor and the nonspecificity of the presenting symptoms. In the present case, the patient presented with a progressive respiratory failure and right-sided heart

From the Department of Medicine, College of Medicine, King Khalid University Hospital, King Saud University, Riyadh, Saudi Arabia.

Address reprint requests and correspondence to Dr. Bahammam: Department of Medicine, King Khalid University Hospital, P.O. Box 7805, Riyadh 11472, Saudi Arabia.

Accepted for publication 8 November 1998. Received 4 August 1998.

failure. Patients with simple lung metastasis in the absence of lymphagitic carcinomatosis or tumor emboli do not usually have such a dramatic presentation. Moreover, our patient had no evidence of cardiac tamponade or gross right-sided outflow obstruction to account for it. We think that the subacute respiratory failure and the right heart failure resulted at least partially from the embolic tumor plugs of the pulmonary vasculature, as was documented by the autopsy findings. Tumor emboli typically present subacutely with progressive dyspnea over days to weeks, and are sometimes accompanied by hemoptysis.<sup>5</sup> This may partly explain the right-sided heart failure in this patient, as tumor emboli increase pulmonary arterial pressure and result in right ventricular failure. Even in patients with known cancer, the correct diagnosis of tumor emboli is made antemortem in only 6% of cases.<sup>6</sup> The most common sources of embolic carcinomatosis are lung, breast, prostate, kidneys and colon.

In our case, a cardiologist reviewed the echocardiographic study retrospectively, and no masses could be visualized. Although the transthoracic echocardiographic study was not optimal, which might have compromised the sensitivity of the test, the limited sensitivity of transthoracic echocardiography in detecting cardiac tumors has been observed in previous reports.<sup>7,8</sup> Transesophageal echocardiography and magnetic resonance imaging (MRI) have been demonstrated to be more sensitive in detecting cardiac tumors.<sup>9-11</sup> As mentioned previously, the diagnosis of tumor emboli is very difficult to make antemortem. Transbronchial lung biopsy can be helpful, but it carries the risk of bleeding in patients with pulmonary hypertension. Another technique for diagnosing pulmonary tumor emboli is pulmonary artery catheter sampling, however, this technique has some technical difficulties and requires an experienced pathologist.

The prognosis of cardiac angiosarcoma is dismal, with a two-year survival of around 17%,<sup>1</sup> because at the time of diagnosis most tumors may have already metastasized. The response to chemotherapy has been disappointing.<sup>12</sup> However, early detection may improve survival.<sup>13,14</sup> Baay et al. recently reported a case of cardiac angiosarcoma treated successfully with cardiac transplantation after preoperative chemotherapy and radiation therapy and postoperative chemotherapy.<sup>14</sup>

From the above, we conclude that despite being rare, cardiac angiosarcoma should be considered in the differential diagnoses of young patients presenting with lung metastasis and right-sided heart failure, or any other cardiac symptoms. Although clinical symptoms caused by tumor emboli alone are usually absent,<sup>15</sup> patients may present with dyspnea and signs of cor pulmonale. It is also important to remember that transthoracic echocardiogram may not detect some of the primary cardiac tumors, and if such a diagnosis is suspected clinically, other tests should tumor with hemorrhagic pericarditis.

## Acknowledgements

I wish to thank Professor Donal McCarthy and Dr. Muhammad Alsaghier of the Health Sciences Centre, University of Manitoba, and Dr. Richard Long of the University of Alberta, for their valuable comments.

## References

1. Naka N, Ohsawa M, Tomita Y, et al. Angiosarcoma in Japan: a review of 99 cases. *Cancer* 1995;75:989-96.
2. Reece IJ, Cooley DA, Frazier OH, et al. Cardiac tumors. Clinical spectrum and prognosis of lesions other than classical benign myxoma in 20 patients. *J Thorac Cardiovasc Surg* 1984;88:439-46.
3. Janigan DT, Husain A, Robinson NA. Cardiac angiosarcomas: a review and a case report. *Cancer* 1986;57:852-9.
4. Afzal N, Alguacil-Garcia A. Primary cardiac angiosarcoma: clinical and pathological diagnostic problems. *Can J Cardiol* 1997;13:293-6.
5. King MB, Harmon KR. Unusual forms of pulmonary embolism. *Clin Chest Med* 1994;15:561-80.
6. Goldhaber S, Dricker E, Buring J, et al. Clinical suspicion of autopsy-proven thrombotic and tumor pulmonary embolism in cancer patients. *Am Heart J* 1987;114:1432-5.
7. Bic JF, Fade-Schneller O, Marie B, et al. Case report: cardiac angiosarcoma revealed by lung metastases. *Eur Respir J* 1994;7:1194-6.
8. Come P, Riley M, Markis J, et al. Limitations of echocardiographic techniques in evaluation of left atrial masses. *Am J Cardiol* 1981;48:947-53.
9. Frohwein SC, Karalis DG, McQuillan JM, et al. Preoperative detection of pericardial angiosarcoma by transesophageal echocardiography. *Am Heart J* 1991;122:874-5.
10. Laissy JP, Bernier P, Patru B, et al. Primary left atrial angiosarcoma: follow-up by magnetic resonance imaging. *Magn Reson Imaging* 1990;8:651-5.
11. Link KM, Lesko NM. MR evaluation of cardiac/juxtacardiac masses. *Top Magn Reson Imaging* 1995;7:232-45.
12. Vergnon JM, Vincent M, Perinetti M, et al. Chemotherapy of metastatic primary cardiac sarcomas. *Am Heart J* 1985;110:682-4.
13. Sorlie D, Myhre ESP, Stalsberg H. Angiosarcoma of the heart. Unusual presentation and survival after treatment. *Br Heart J* 1984;51:94-7.
14. Baay P, Karwande SV, Kushner JP, et al. Successful treatment of a cardiac angiosarcoma with combined modality therapy. *J Heart Lung Transplant* 1994;13:923-5.
15. Neoplastic diseases of the lungs. In: Fraser R, Pare JAP, Fraser R, Pare PD, editors. *Synopsis of Diseases of the Chest*. 2nd edition. Philadelphia: WB Saunders, 1994:525.