Angiosarcoma of the Heart Presenting as Pulmonary Disease

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A 23-year-old man died from the pulmonary manifestations of cardiac angiosarcoma. The absence of all cardiac signs and symptoms was an unusual feature. The clinical outcome was rapidly fatal. Apparently, the presence of cardiac symptoms in a patient with primary cardiac angiosarcoma is not obligatory.

Angiosarcoma of the heart is a disease rarely diagnosed before death. Although it is the most common malignant primary cardiac tumor, its rare occurrence probably leads to a low index of suspicion. However, clinical symptoms of the disease are reported to be remarkably consistent. In the series described by Strohl, 86 percent of the patients presented with symptoms of pericardial disease or rightsided congestive heart failure caused by obstruction of the vena cava or obstruction of the outflow tract of the right ventricle. The right atrium is by far the most common site of origin of the disease. Metastases can be found in lungs, lymph nodes and liver.

We describe a patient without any cardiac symptoms, in whom angiosarcoma was diagnosed by means of an open lung biopsy. At postmortem, a primary tumor of the right atrium was found. The patient died of intractable respiratory failure.

CASE REPORT

A 23-year-old man was in good health until October, 1986, when he presented with hemoptysis. He had not noted any shortness of breath, chest pain, fatigue, weight loss, cough or fever. The patient had not been involved with IV drug abuse, nor did he have a homosexual history or association with prostitutes. He had not received blood products of any kind. Physical examination revealed a healthy-looking young man. Blood pressure was 150/75 mm Hg, without pulsus paradoxus; rectal temperature was 36.6°C. There was no elevation in jugular venous pressure. The heart rhythm was regular; no murmur or friction sound was heard. Fine crackles were heard over the base of the left lung. The liver and the spleen were not enlarged. No clubbing, cyanosis or peripheral edema was noted. Examination of the skin was unremarkable.

The erythrocyte sedimentation rate was 4 mm/h; hemoglobin was 16.3 g/100 ml. Blood urea nitrogen was 16.6 mg/100 ml; serum creatinine was 1.2 mg/100 ml. The findings from urinalysis were normal. Antinuclear antibodies, antibodies against DNA and antibodies against glomerular basement membrane were not detected.

Arterial blood gas levels on room air were: PaO₂ 89 mm Hg; PaCO₂ 35 mm Hg; oxygen saturation, 96 percent. There was a decrease of arterial PaO₂ to 63 mm Hg during physical exercise. Results of sputum smears were negative; no acid-fast bacilli were seen.

A chest x-ray film (Fig 1) showed bilateral nodular infiltrates. The heart was not enlarged and the electrocardiogram was normal. Bronchoscopic examination showed blood in the basal airways. Biopsy findings were normal except for an increase in the number of hemosiderin-laden macrophages. A tentative diagnosis of idiopathic pulmonary hemosiderosis was made.

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Since the pulmonary bleeding had stopped spontaneously and the patient was in excellent clinical condition, he was sent home. He was readmitted on November 17 because of shortness of breath. Arterial PO$_2$ on room air was 44 mm Hg and arterial PCO$_2$, 20 mm Hg. The hemoglobin level had fallen to 10.8 g/100 ml. The abnormalities on the chest x-ray film had worsened, now forming an alveolar-filling pattern. The patient was treated with supplemental oxygen, high-dose steroids and desferrioxamine. However, his condition deteriorated, and on December 7, artificial ventilation had to be instituted because of severe hypoxemia. Right cardiac catheterization revealed a right atrial pressure of 1 mm Hg, a mean pulmonary artery pressure of 26 mm Hg and a pulmonary capillary wedge pressure of 4 mm Hg. On December 9, right-sided thoracotomy was performed to obtain a histologic diagnosis. The lung was dark red. The pleura was covered with multiple small tumors. A biopsy specimen of the lung was characteristic of angiosarcoma. Subsequently, the patient was started on doxorubicin, but he died December 19 from intractable respiratory failure.

At postmortem examination, cardiac angiosarcoma was found infiltrating the entire thickness of the right atrial wall (Fig 2). There was gross bleeding from multiple metastases in the lungs.

**DISCUSSION**

More than 139 cases of cardiac angiosarcoma have been described in the literature. It is an almost invariably fatal disease with a predilection for male patients and middle age. Many authors have stressed the constancy of symptoms and clinical signs. Since the tumor is almost exclusively right-sided, it usually produces symptoms of right heart failure. Virtually all patients have cardiomegaly. Most patients have ECG changes as well. Fever, weight loss and malaise are present in 10 percent of patients. Precordial pain caused by pericarditis is reported in 75 percent of the cases.

The patient described here had none of these classic symptoms. He presented with hemoptysis, nodular infiltrates in both lungs, and anemia, leading us to make an erroneous diagnosis of idiopathic pulmonary hemosiderosis. Not even right-sided cardiac catheterization was suggestive of mechanical obstruction of the heart. The elevated pressure in the pulmonary artery can be explained by hypoxia and widespread occlusion of the pulmonary vascular tree by tumor emboli. The tumor load in the right atrium turned out to be small, as compared to the larger tumors, described by other authors in similar cases. Diffuse infiltration of both lungs by tumor, in combination with alveolar bleeding, had led to respiratory failure. The clinical picture of this patient was remarkable for its extremely rapid deterioration: death followed only two and a half months after the onset of symptoms.

In their recent review, Janigan et al drew attention to the problem of histologically differentiating angiosarcoma from Kaposi's sarcoma. They define two clinicopathologic forms of cardiac angiosarcomas. In the first, the lesions are in the epicardium or pericardium, are usually small and asymptomatic, and are associated with skin lesions of Kaposi's sarcoma or risk factors of the acquired immunodeficiency syndrome. In the second, the lesions are large and symptomatic and not associated with recognized clinical Kaposi's sarcoma or its risk factors.

From the absence of any such risk factors or cutaneous manifestations of Kaposi's sarcoma in our patient, we must conclude that this was not a case of Kaposi's sarcoma, but of...
an angiosarcoma with unique presentation. In this patient, a relatively small and clinically silent primary tumor in the right side of the heart had given rise to widespread metastatic disease in the lungs, causing pulmonary hemorrhage and respiratory failure. Perhaps, in cases like this, the limited lifespan, caused by the extreme malignancy of metastatic disease, precludes the development of cardiac symptoms. A patient with a history very similar to the one reported here has been described by Yousem. However, although signs of cardiac involvement were notably absent, no autopsy was performed to establish the origin of the tumor. From these findings we conclude that, although angiosarcoma of the heart is a rare tumor, its presence should be suspected in patients with hemoptysis and nodular chest x-ray abnormalities, even in the absence of symptoms of right-sided cardiac disease.

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