Right Atrial Myxoma Presenting as Nonresolving Pulmonary Emboli: Case Report

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A case of right atrial myxoma causing extensive pulmonary embolization is described. Five years elapsed between the initial consultation and the correct diagnosis. The patient has been free of symptoms for 3 years following surgical removal of the tumor. The clinical manifestations of right atrial myxoma, the differential diagnosis, and the atypical lung-scan features are discussed.

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Primary tumors of the heart are rare, with a reported incidence of 17 per million autopsies. Half of all cardiac tumors are myxoma, of which 25% are located in the right atrium (*1,2*). While cardiac myxoma is considered a benign tumor, death may result either from primary obstruction to blood flow or from embolization of tumor fragments (*3*). In patients with right atrial myxomas, pulmonary embolization of tumor fragments, usually misdiagnosed as thromboemboli, is not uncommon (*4*).

CASE REPORT

A 61-year-old black woman presented with a 5-month history of malaise, fatigue, and anorexia. Pertinent past medical history included a clinic visit 3 years previously for shortness of breath, ankle swelling, and headaches of 3 weeks' duration. A presystolic murmur at the lower left sternal border and ankle edema were noted at that time, and a diagnosis of mild early right-sided heart failure was made. The patient was treated with Diuril (500 mg twice a day) and placed on a low-salt diet.

Four months before this admission, the patient was admitted for removal of a vaginal polyp. No heart murmurs were noted, although an S3 gallop was heard along the left sternal border. The patient had no edema. Interim medical problems included adult-onset diabetes, with blood sugar levels of 220–294 mg%, a persistently elevated erythrocyte sedimentation rate, and an iron-deficiency anemia. Barium studies of the entire gastrointestinal tract were normal. An intravenous urogram showed a renal cyst. An occult malignancy was suspected but not found.

Physical examination at this last admission was unremarkable, except for a grade 2/6 presystolic murmur heard along the lower left sternal border. The murmur did not radiate. The lungs were clear to percussion and auscultation. Mild ankle edema was seen. The following laboratory values were obtained: hemoglobin 9.9 gm; hematocrit 31, iron 45 mg%, total iron-binding capacity 260 mg%, elevated gamma-2-globulins, erythrocyte sedimentation rate 117, and elevated haptoglobins. The stool guaiac was positive. Latex fixation and lupus erythematosus preparations were normal. A chest x-ray revealed mild cardiomegaly unchanged from previous studies.

Because of the patient's persistent dyspnea, the diagnosis of pulmonary emboli was suggested, and a lung scan was obtained (Fig. 1A). The left lung field was well perfused, but no perfusion was visualized on the right. A pulmonary angiogram showed complete occlusion of the right pulmonary artery by a smooth ovoid defect, considered to be a large embolus (Fig. 1B). Anticoagulant therapy was in-

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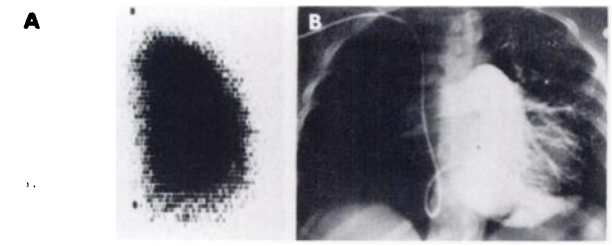

FIG. 1. (A) Lung scan shows absent perfusion of right lung, with normal perfusion of left lung. (B) Pulmonary angiogram reveals complete occlusion of right pulmonary artery.

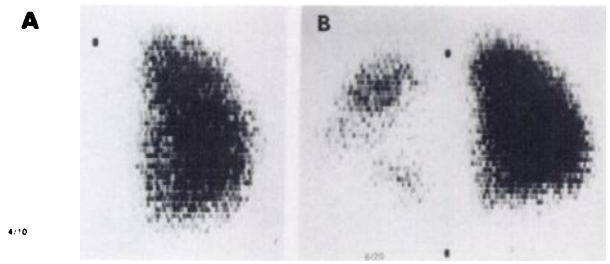

FIG. 2. (A) Followup lung scan at 2 weeks shows no improvement despite vigorous anticoagulation therapy. (B) Followup lung scan at 3 months shows large residual perfusion defects on right. This pattern persisted for 18 months.

stituted, and the patient was subsequently discharged. Six followup lung scans over the next 18 months revealed some improvement in the right pulmonary perfusion. However, continued marked underperfusion of the right lung was noted on all of the studies (Fig. 2).

The patient's dyspnea, ankle edema, and intermittent murmurs indicated further cardiac evaluation. Before cardiac angiography, a new diastolic rumble and a 3/6 presystolic murmur were heard at the lower left sternal border. A right atrial angiogram disclosed a dumbbell-shaped mass originating in the right atrium and extending into the right ventricle. Marked resolution of the previously described obstruction in the right pulmonary artery was noted. On the other hand, multiple obstructions and irregularities were found in the right pulmonary vasculature with an overall diminished perfusion of the right lung (Fig. 3).

At operation a right atrial myxoma, measuring 7.5 × 4.5 cm, was found. It was fixed only at the fossa ovalis and extended through the tricuspid valve into the right ventricle. The patient had a relatively uneventful postoperative course and was discharged on the 15th day after surgery.

DISCUSSION

The clinical manifestations of right atrial myxoma are protean. The most frequent syndrome is one of progressive dyspnea, fatigue, recent development of a cardiac murmur, and signs of right heart failure (*5*). Cardiac auscultation usually reveals a variable presystolic murmur, the presence or absence of which depends on tumor position at the time of atrial systole (*6*). The murmur is best heard at the lower left sternal border. Often it is increased in intensity with inspiration, like the murmur of rheumatic tricuspid stenosis. Wide splitting of the first sound may be heard (*7*).

Variable episodes of syncope can be attributed either to temporary obstruction of blood flow to the right ventricle or to ball-valve movement of the tumor at the tricuspid valve. Associated arrhythmias may rarely cause episodic syncope (*7*).

Constitutional symptoms, when present, include fever, dyspnea, and fatigue (*2,3*). Laboratory findings usually include anemia, thrombocytopenia, an elevated erythrocyte sedimentation rate, and hyperglobulinemia (*1,8,9*). Occasionally a right-to-left cardiac shunt is present. This presumably occurs because the myxoma obstructs blood flow, elevates right atrial pressure, and facilitates flow through a persisting foramen ovale. Such a shunt can cause arterial hypoxia and an elevated hematocrit (*9*).

Emboli from tumor fragments are a potentially fatal complication of cardiac myxoma. Emboli aris-

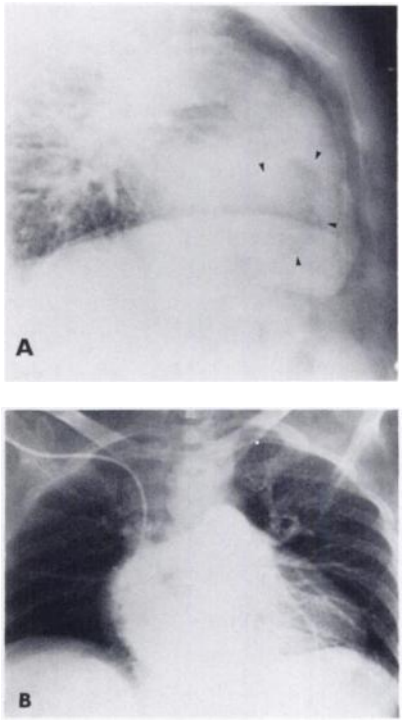

FIG. 3. (A) Right atrial angiogram shows dumbbell-shaped mass (arrows) originating in right atrium and extending into right ventricle. (B) Marked resolution of obstructing lesion in right pulmonary artery is seen. However, overall perfusion of right lung is decreased.

ing from a right-sided myxoma can cause irreversible pulmonary hypertension with cor pulmonale. The resultant pulmonary perfusion deficits are usually extensive on the initial lung scan (*4*), presumably because of sporadic "silent" emboli over a long interval and because of the extremely slow resolution of such emboli. As seen in our patient, perfusion deficits caused by myxoma emboli can persist for long periods of time despite treatment that normally would hasten the resolution of thromboemboli. The static appearance of these perfusion defects may be misinterpreted as evidence for underlying chronic obstructive pulmonary disease. Certainly evaluation of pulmonary ventilation with radioxenon would aid in the differential diagnosis.

Another feature seen in our patient was absent perfusion of the involved lung with entirely normal perfusion of the opposite lung. In our experience, total unilateral absence of pulmonary perfusion due to venous thromboembolism is always accompanied by one or more foci of contralateral ischemia, due to the fact that venous thromboemboli nearly always undergo fragmentation and lead to multiple occlusions. The "saddle embolus" is a conspicuous exception.

Both static imaging of the cardiac blood pool (*10*) and radionuclide angiography (*11,12*) have been used to show intracardiac filling defects due to myxoma. Abnormalities noted on chest films have been relatively nonspecific. In one series of 35 patients (*1*), there were 22 with cardiomegaly, 18 with a prominent right heart border, 4 with nonvalvular intracardiac calcification, 4 with pleural effusion, and 5 with a normal chest. None of the patients showed evidence of pulmonary vascular congestion.

Of major concern in this condition is the long duration of illness before correct diagnosis is made. The average duration appears to be about 3 years, with the shortest reported duration 5 days and the longest 36 years (*1*). Although myxoma is a rare tumor, nevertheless, the diagnosis should be considered in patients with isolated right heart failure without pulmonary congestion, pulmonary disease, or pulmonary stenosis. The variable murmur of tricuspid stenosis in the absence of other valvular auscultatory findings should further strengthen the consideration of right atrial myxoma, since it is much rarer to have isolated tricuspid involvement in rheumatic heart disease. Finally, atypical lung-scan findings, as described above, should alert the nuclear medicine physician to the possibility of myxoma emboli. Once the diagnosis is considered, radionuclide angiography may prove to be an effective screening procedure. Currently, contrast angiography of the right atrium is the diagnostic procedure of choice. Some investigators advocate injection of the superior vena cava to avoid dislodgement of tumor emboli (*2*). Although invasive, contrast angiography provides anatomic detail not available by other diagnostic procedures; however, perhaps whole-body computed tomography will later become competitive. Surgical removal of the tumor is feasible and may result in a complete cure; excision of the area of attachment is controversial (*3*).

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