CLINICAL VIGNETTE

Scleroderma and Intracardiac Myxoma: A Case Report

Pamela Law and Lisa Skinner, MD

Abstract

A 54-year-old female with a history of scleroderma, interstitial lung disease, and pulmonary hypertension was found to have an atrial myxoma during an echocardiogram.

Case Report

A 54-year-old African-American woman with known scleroderma presented with acute worsening shortness of breath in the setting of interstitial lung disease and pulmonary arterial hypertension. Her past medical history began in the 1980's when she presented with fatigue, an elevated erythrocyte sedimentation rate, and uveitis. She was treated with prednisone at that time and her symptoms remained stable until 1990, when she began having acid reflux and difficulty swallowing. She was found to have a positive ANA, lymphadenopathy, and mild sclerodactyly and was subsequently diagnosed with limited scleroderma.

In 2000, the patient began to experience progressively worsening shortness of breath. She did not have a diagnostic workup until 2005, when she underwent pulmonary function testing. Her forced vital capacity at that time was found to be 56%. She was started on Bosentan therapy for pulmonary arterial hypertension as well as 24-hour supplemental oxygen. Her oxygen need progressively increased to approximately 4 to 6 liters daily. For her scleroderma, she received IV Cyclophosphamide.

Her symptoms remained relatively well managed until 4 days prior to admission, when she developed shortness of breath at rest despite her usual 4 to 6 liters of supplemental oxygen. She noted some increased swelling in her lower extremities and fleeting chest pain and was occasionally awakening at night with extreme dyspnea. She was admitted to the hospital for further workup and treatment, including possible initiation of a second agent for her pulmonary hypertension.

In the Emergency Department, the patient was noted to be tachycardic and hypoxic. She was given supplemental oxygen and diuresed for presumed exacerbation of right heart failure and pulmonary hypertension. Her laboratory studies revealed BNP elevated to 1490 pg/mL. Chest X-ray showed a large cardiac silhouette with bilateral pleural effusions and cardiogenic pulmonary edema. An echocardiogram demonstrated a large right ventricle with decreased function, right atrial enlargement, moderate tricuspid and pulmonic regurgitation, and severe pulmonary hypertension. Incidentally, a mobile, pedunculated mass of the interatrial septum was noted within the left atrium. Cardiac MRI confirmed a polypoid, 1cm x 1cm left atrial mass consistent with a cardiac myxoma.

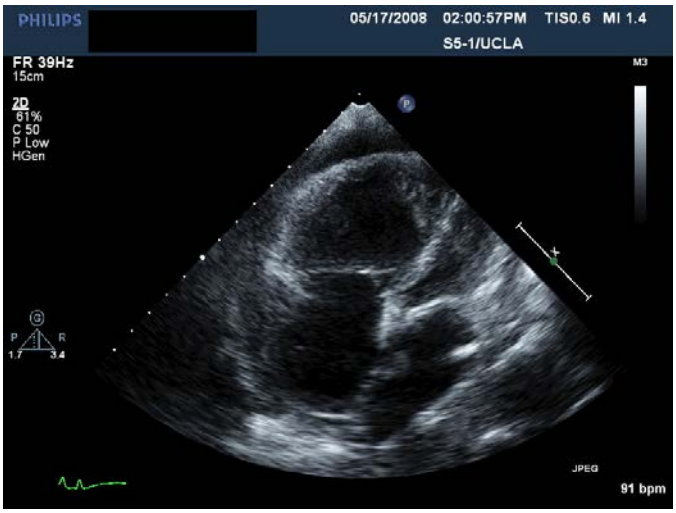

Figure 1. The 2-D echocardiogram shows a left-atrial myxoma of inter-atrial septum. Notably, the patient's right heart is extremely enlarged secondary to chronic pulmonary arterial hypertension of scleroderma.

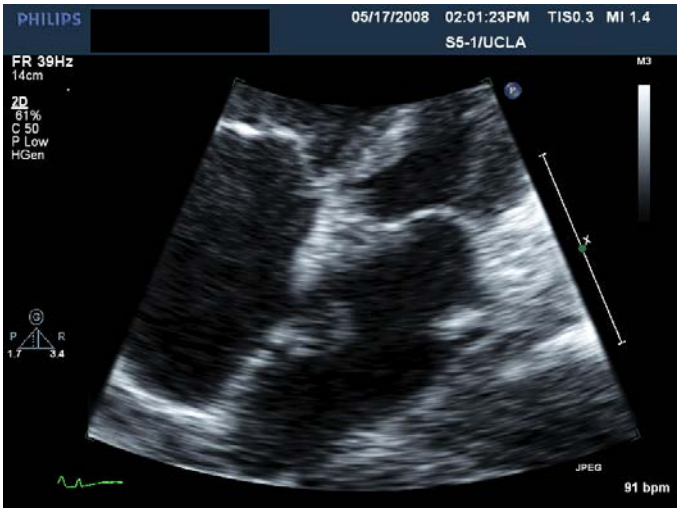

Figure 2. A closer view of the 2-D echocardiogram showing the inter-atrial myxoma. The mass is pedunculated and is located near the fossa ovalis, characteristic findings for cardiac myxoma.

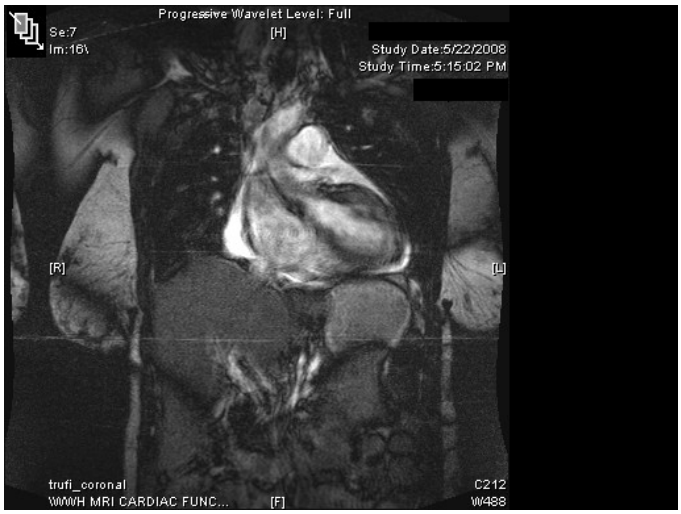

Figure 3. MRI: the myxoma is visible and possibly spans the inter-atrial septum.

The incidental finding of cardiac myxoma during the patient's evaluation for pulmonary hypertension required the team to reprioritize the treatment goals. In addition to treating the patient's congestive heart failure, the team sought to mitigate the risk associated with the myxoma. Anticoagulation therapy was initiated in order to decrease the risk of thromboembolization. The patient's clinical status improved with diuresis and continued Bosentan therapy, her right heart catheterization demonstrated an elevated systemic pulmonary artery pressure of 79 mmHg. The General Medicine team evaluated and prioritized the recommendations from pulmonary and cardiology consultants.

High right atrial pressure indicative of right ventricular failure is a strong predictor of mortality in scleroderma patients. In one retrospective case-control study, 106 patients with scleroderma with symptomatic pulmonary hypertension and right sided heart failure had five year mortality rates of approximately 90 percent versus 20 percent in 106 age, gender, scleroderma subtype, and disease-duration matched controls.[1] In two small retrospective studies with cohorts that included 22 and 19 scleroderma patients with PAH (mean pulmonary artery pressure 48 mmHg), two year mortality approached 60 percent despite therapy.[2] An observational study of 794 patients with scleroderma confirmed this finding with a similar two year mortality rate of 61 percent in patients with mean pulmonary arterial pressures of >45 mmHg.[3]

The team considered that the patient was a very high-risk candidate for surgical removal of the left atrial mass. On the other hand the myxoma, if untreated, conferred a high mortality in itself. Given her pulmonary hypertension and decreased right heart function, the mortality of open-heart surgery was estimated to be as high as 20 to 30%. The team weighed the risks of surgical versus medical therapy in the setting of incurable and progressive lung disease. Multiple discussions explored the goals of care. The primary team was charged with integrating varied points of view regarding the short term risks of surgery versus anticoagulation therapy. It was ultimately decided to defer surgery and continue anticoagulation and medical therapy to optimize pulmonary function.

Discussion

Cardiac myxomas are the most common benign cardiac tumor in adults. They account for 40-50% of primary cardiac tumors.[4] Seventy-five percent of sporadic myxomas occur in females. Ninety percent are solitary and pedunculated, and 75-85% occur in the left atrial cavity.[4] The most common site of attachment is at the border of the fossa ovalis in the left atrium.[4] Cells that give rise to the tumor are mulitpotent mesenchymal calls that persist after septation of the heart and would normally differentiate into the cells of the myocardium and endocardium. Myxomas vary widely in size, ranging from 1-15 cm in diameter. The rate of growth is not exactly known, although they are generally known to grow fairly rapidly. Myxomas can be polypoid or villous. Polypoid myxomas are compact and show little tendency toward spontaneous fragmentation. The less common villous or papillary myxoma has a surface of fine villous extensions, often gelatinous and fragile and has a tendency to break off pieces.

Symptoms are produced by mechanical interference with cardiac function or embolization. Myxomas account for most cases of tumor embolism due to their intravascular location and friable nature. Embolization occurs in about 30-40% of cases.[4] Symptoms are produced by tumor embolism, heart failure, mechanical valvular obstruction, and various constitutional symptoms. In about 20% of cases, myxoma may be asymptomatic and discovered as an incidental finding.

Though cardiac myxomas are the most common benign cardiac tumor in adults, other causes of cardiac mass must be included in the differential diagnosis, including other primary heart tumors, such as angiosarcoma or rhabdomyosaroma, metastatic tumor, valvular vegetations and cardiac thrombi.[4] Whether metastatic or primary, cardiac tumors increase risk for infection and embolization. Myxomas are difficult to distinguish from thrombi on echocardiogram and other imaging. In fact, because myxomas develop a thrombotic surface, they are often difficult to distinguish from thrombi even upon direct visualization.

The treatment for myxomas is surgical removal. Prompt surgery reduces risk of embolism and sudden death and is usually curative. Sudden death may occur in 15% of atrial myxoma patients.[5] Death is typically caused by coronary or systemic embolization or by obstruction of blood flow at the mitral or tricuspid valve. Surgical resection of atrial myxomas involves cardiopulmonary bypass. The entire base of the pedicle must be removed, including the inter-atrial septum, followed by repair of the atrial septal defect. This is necessary due to the multipotent nature of mesenchymal myxoma cells and the potential for local recurrence after inadequate resection.[4,5]

Table 1: Complications and Associated Symptoms of Cardiac Myxoma (adapted from [5])

Complication	Associated Symptoms
Embolization to pulmonary, cerebral, and retinal arteries	Dyspnea and chest pain Transient or permanent visual loss Stroke, focal neurological symptoms
Heart failure	Dyspnea due to recurrent pulmonary edema Symptoms of right heart failure
Mechanical intracardiac obstruction	Acute heart failure mitral or tricuspid insufficiency Syncope Sudden death Hemolytic anemia
Constitutional symptoms	Fatigue Fever Erythematous rash Arthralgia Myalgia Weight loss

Conclusion

The coincidence of cardiac myxoma in a patient with cardiac and pulmonary complications of scleroderma presented unique challenges in strategizing treatment. The situation necessitated multidisciplinary discussion to weigh the risks and benefits of treatment in a patient with poor baseline prognosis. It was important to weigh the risk open-heart surgery in the background of severe lung disease against the thromboembolic risk of a cardiac myxoma. Ultimately, it was decided that surgical removal of the myxoma would be clinically inappropriate due to the patient's high risk for open-heart surgery and underlying progressive disease.

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