A Rare Cause of Pulmonary Hypertension and Right Ventricular Failure: Left Atrial Myxoma

Arielle Schwartz, MD, Evan Blank, MD, and Marc Thames, MD, *Atlanta, Georgia*

INTRODUCTION

Primary cardiac tumors are rare, with an estimated incidence of 0.1%. Atrial myxomas make up about 50% of these tumors. Patients may present with a variety of symptoms, which are related to obstruction of circulation, embolization, interference with cardiac valves, and/or direct invasion of the myocardium or adjacent lung tissue.[1] The first recorded attempt at diagnosing a left atrial tumor in a living person occurred in 1951 using an unsuccessful surgical technique following identification of an enlarged left atrium on angiography.[2] It wasn't until the early 1970s that the first two-dimensional echocardiogram was used clinically, allowing for visualization and characterization of myxomas.[3] As technology has evolved, diagnostic accuracy and safety of excising such tumors have improved. Here we describe a rare case of a left atrial myxoma with obstruction of mitral flow and the integral role that transthoracic echocardiography (TTE) played in identifying the mass and quantifying the secondary pulmonary hypertension and right ventricular (RV) failure that resulted from its obstructive physiology.

CASE PRESENTATION

A 58-year-old female with no significant past medical history presented to her primary care physician with progressive fatigue and shortness of breath over several months. Her symptoms progressed to orthopnea and paroxysmal nocturnal dyspnea with palpitations. Her primary care physician noted bilateral lower extremity edema, decreased breath sounds bilaterally, and an audible S3 on exam. The patient was subsequently admitted and found to have a B-type natriuretic peptide of 2,473 pg/mL. Transthoracic echocardiography (Figure 1) revealed a 5.1 × 3.4 cm left atrial mass (A) attached to the interatrial septum at the level of the foramen ovale obstructing the mitral valve inflow (Figure 1A and B). Left ventricular ejection fraction was 60%–65% with a severely dilated RV and moderately to severely reduced RV systolic function. The estimated pulmonary arterial pressure was 90–100 mm Hg (Figure 1C), with the RV systolic pressure severely elevated at 93.1 mm Hg, tricuspid regurgitant velocity of 4.42 m/sec, and an estimated right atrial pressure of 15 mm Hg. Both atria were severely dilated. The Doppler inflow signal across the mitral valve showed almost total obstruction of the mitral orifice by the myxoma with no transmitral flow detectable in the second half of diastole (Figure 1D). Coronary angiography revealed patent vasculature and a left ventricular end-diastolic pressure of 20 mm Hg. The decision was made to urgently excise the tumor. The procedure was complicated by RV failure, requiring high-dose vasopressor and inotropic support, intra-aortic balloon pump, and inhaled epoprostenol. The patient was initially transferred to the intensive care unit with an open chest. She was subsequently stable enough to return to the operating room for thoracic closure. She recovered well, with resolution of her symptoms. The hematoxylin and eosin stain of the 5.5 × 5.0 × 3.3 cm tan surgical specimen revealed an atrial myxoma.

VIDEO HIGHLIGHTS

Video 1: Parasternal long-axis view of left atrial myxoma across the mitral valve.

Video 2: Parasternal short-axis view of left atrial myxoma across the mitral valve.

Video 3: Apical four-chamber view of left atrial myxoma.

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DISCUSSION

This case details a patient found to have pulmonary hypertension and reduced RV function secondary to a left atrial myxoma, which obstructed transmitral blood flow. The patient's elevated pulmonary arterial pressure and RV dysfunction indicate the chronic nature of this tumor and resulting cardiac manifestations from its hemodynamic effects. Pulmonary manifestations are quite common in patients with left atrial myxoma perhaps secondary to obstruction of mitral flow mimicking severe mitral stenosis. However, pulmonary hypertension, severe RV systolic dysfunction, and tricuspid regurgitation are less commonly recognized sequelae.[4-7]

In a study at the All India Institute of Medical Sciences, a tertiary referral center for patients with cardiovascular disease throughout Northern India, 70 cases of cardiac myxomas were identified in the 93,500 echocardiograms performed over an 11-year period.[7] Only five of the 70 patients had moderately to severely elevated RV pressures with tricuspid regurgitation, and only three patients had associated pulmonary hypertension, as seen in our patient.

Interestingly, signs and symptoms of RV failure were not reported in the largest retrospective reviews of atrial myxoma cases. Over 40 years, 112 patients with atrial myxoma in a French hospital were identified. Dyspnea and orthopnea were the most common presenting symptoms, occurring in about half of these patients with associated pulmonary edema.[6] What is not mentioned in this review is the incidence of signs and symptoms of right-sided failure, such as abdominal bloating and peripheral edema. Additionally, RV pressures were not reported. This information may have been helpful in understanding the chronicity of the tumors in these patients and their resulting

From the J. Willis Hurst Internal Medicine Residency Program (A.S.) and Emory University Division of Cardiology (E.B., M.T.), Atlanta, Georgia.

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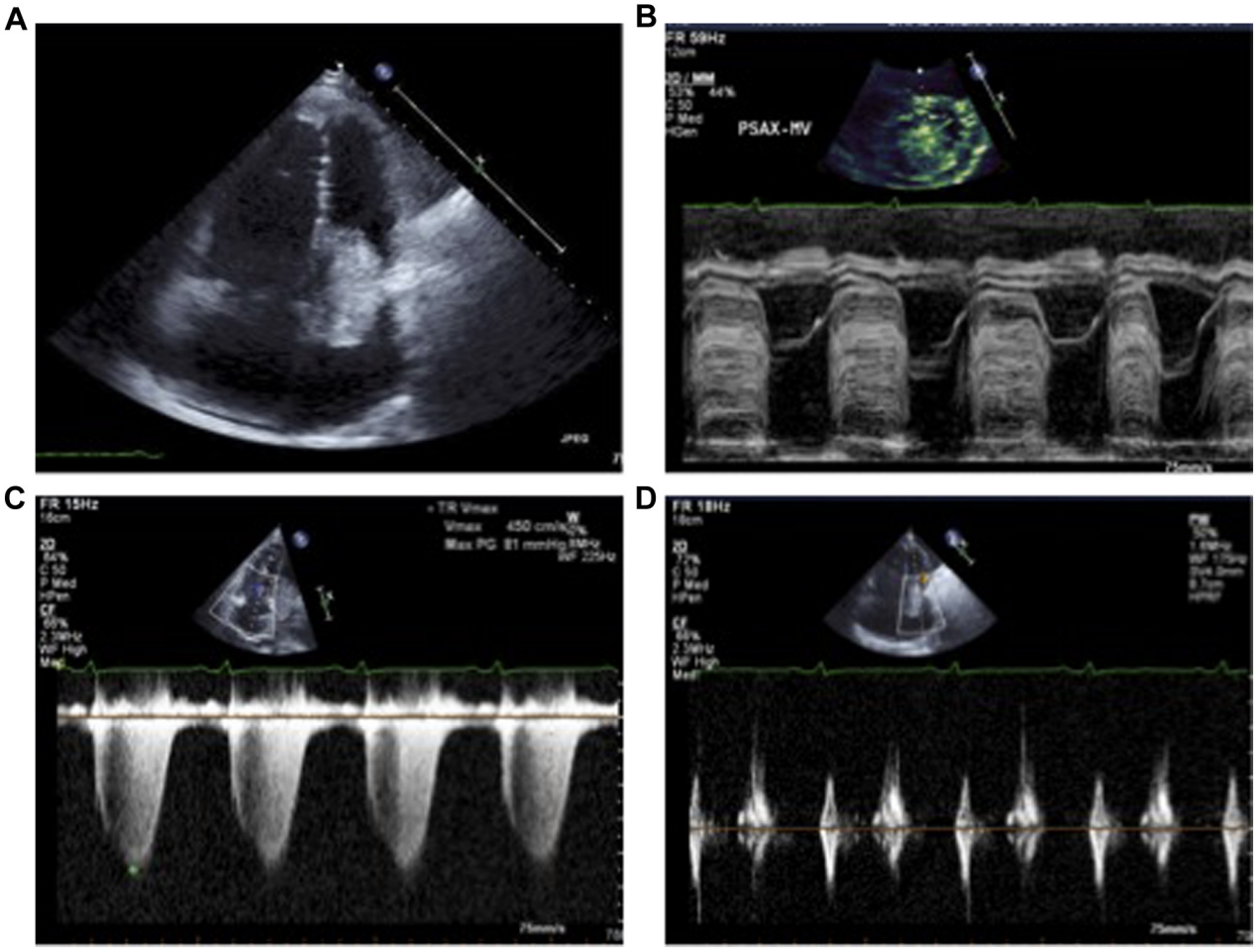

Figure 1 **(A)** Apical four-chamber view of left atrial mass; **(B)** M mode of left atrial mass; **(C)** mitral valve inflow obstruction due to left atrial mass; and **(D)** severe pulmonary hypertension due to left atrial mass.

hemodynamic and structural effects. While the incidence of patients with atrial myxomas who develop RV failure is not known, other cases exist in the literature, suggesting it may be an underrecognized consequence of left atrial myxomas.[4,5,7]

The use of TTE is important in the diagnosis of atrial myxomas since this diagnosis is only suspected clinically in about 5% of myxoma patients.[6] Additionally, as in the case of our patient, TTE allows for observation of hemodynamic compromise and late-stage myocardial changes that ensue from obstructive left atrial tumors. Transthoracic echocardiography should be obtained early in any person with signs or symptoms of heart failure. In the event that a left atrial myxoma is identified, prompt removal is indicated to prevent late-stage complications.

CONCLUSION

We present a rare case of pulmonary hypertension and severe RV systolic dysfunction secondary to left atrial myxoma obstructing mitral valve inflow. Transthoracic echocardiography allows for the identification and quantification of this physiology. Early identification and removal of myxomas is important, especially when obstruction of the mitral valve is involved, to reduce cardiovascular sequelae including pulmonary hypertension.

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SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2020.11.005.

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