

Acute Mitral Regurgitation: The Dreaded Masquerader



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INTRODUCTION

Unilateral pulmonary edema in patients with hypoxemic respiratory failure is usually secondary to infectious etiology. However, cardiogenic unilateral pulmonary edema is a rare yet important entity that is often misdiagnosed initially, leading to significant delays in treatment. Cardiogenic unilateral pulmonary edema is often secondary to acute mitral regurgitation (MR), a life-threatening hemodynamic abnormality that requires urgent medical attention and usually surgical intervention. Acute posterior leaflet flail has been associated with right-sided unilateral pulmonary edema. However, anterior leaflet flail leading to right upper lobe opacity is very rare.

CASE PRESENTATION

A 73-year-old woman presented to the emergency department with progressively worsening shortness of breath over 5 days, associated with nonproductive cough, fever, and chills. Her medical history was significant for poorly controlled hypertension, diabetes, nonobstructive coronary artery disease, and complete heart block with wide QRS complex escape rhythm, for which she declined cardiology referral. The patient collapsed on her way from the hospital's parking lot to the emergency department, where she was found to be in severe respiratory distress. Initial vital signs were a respiratory rate of 28 breaths/min, a heart rate of 52 beats/min, blood pressure of 70/50 mm Hg, and temperature of 98.1°F. Her oxygen saturation was 79% on room air. Physical examination revealed bilateral crackles and normal heart sounds with no appreciable murmur or gallop. Chest radiography demonstrated a large right upper lobe opacity and bilateral interstitial infiltrates (Figure 1).

Because of worsening respiratory failure, the patient was intubated and initiated on mechanical ventilation. Initial investigations demonstrated leukocytosis (15,300/ μ L), elevated lactate (13.5 mmol/L), mildly elevated cardiac troponin I (0.188 ng/dL), and markedly elevated B-type natriuretic peptide (2,002 pg/mL) prior to intubation. The patient remained afebrile despite subjective sensation of fever and chills. Blood, sputum, and urine cultures were obtained on admission and did not demonstrate any growth. Electrocardiography revealed sinus bradycardia

with complete heart block and wide QRS complex escape rhythm. In the medical intensive care unit, the patient received antibiotic therapy and intravenous fluids for presumed diagnosis of multilobar pneumonia. After an initial hemodynamic improvement, the patient's course rapidly deteriorated, with development of hypotension requiring initiation of norepinephrine and transcutaneous pacing.

Subsequently, transthoracic echocardiography demonstrated normal left ventricular ejection fraction, estimated at 55%–65%, with evidence of restrictive filling pattern (Video 1). Left ventricular end-diastolic diameter and end-systolic diameter were measured at 50 and 35 mm, respectively. The mitral valve leaflets appeared thickened. There was severe flail motion of the anterior mitral leaflet, with evidence of ruptured chordae tendineae (Figure 2, Videos 2 and 3). There was a severe eccentric MR directed toward the posterior wall of the left atrium (Figures 3 and 4, Video 4). The left atrium was markedly dilated (Video 1). The right ventricle was dilated, with reduced systolic function (Video 1). Pulmonary artery systolic pressure was estimated in the range of 60–65 mm Hg (Figure 5). Transesophageal echocardiography was performed to better differentiate between infective endocarditis and myxomatous degeneration, and to exclude the possibility of acute ischemic MR considering the recent diagnosis of complete heart block. Transesophageal echocardiography confirmed anterior mitral leaflet flail with evidence of ruptured chordae tendineae and intact papillary muscles (Figure 6, Videos 5 and 6). The valve morphology was consistent with myxomatous proliferation. The MR jet was eccentric, directed toward the posterior wall of the left atrium and the right upper pulmonary vein, with systolic reversal of flow in the pulmonary vein (Figures 7 and 8, Video 7). Applying the volumetric flow equation method at the mitral valve annulus and left ventricular outflow tract, the regurgitant volume and regurgitant fraction were calculated to be 66 mL and 62%, respectively.¹ The effective regurgitant orifice area by the continuity method was calculated to be 0.6 cm².¹ There was no evidence of valvular vegetation. Subsequently, the working diagnosis was changed from pneumonia to cardiogenic shock secondary to acute MR caused by ruptured chordae tendineae. The etiology of mitral valve disease was presumed to be myxomatous degeneration.

The patient was urgently taken to the cardiac catheterization laboratory for further hemodynamic evaluation and stabilization. An intra-aortic balloon pump and temporary transvenous pacemaker were placed for hemodynamic support. Right-heart catheterization revealed a mean pulmonary artery pressure of 44 mm Hg and pulmonary capillary wedge pressure of 34 mm Hg. Using the assumed Fick method, the cardiac index was calculated at 2.1 L/min/m².² Following a discussion with the cardiovascular surgery team, coronary angiography was not performed, as only emergent mitral valve replacement was contemplated in this critically ill patient. Intraoperatively, the patient was found to have ruptured chords of the middle segment of the anterior mitral valve leaflet (A2). The mitral valve was replaced with a 27-mm Mosaic bioprosthetic porcine pericardial valve prosthesis (Medtronic, Minneapolis, MN). Pathologic examination of the mitral

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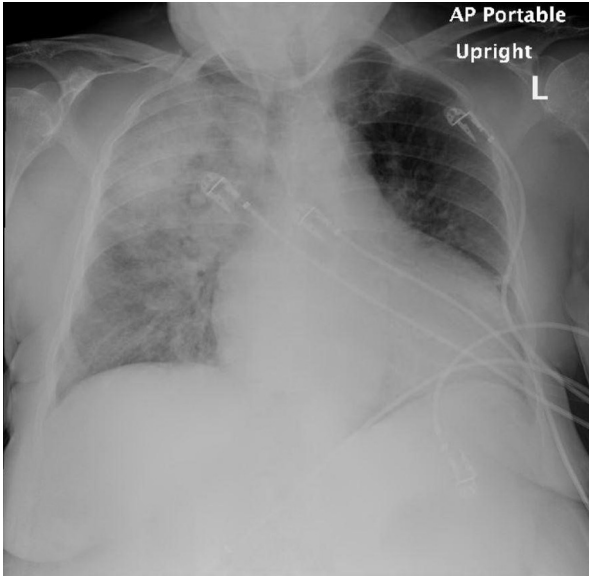


Figure 1 Chest radiograph showing right upper lobe opacity with bilateral interstitial infiltrates.

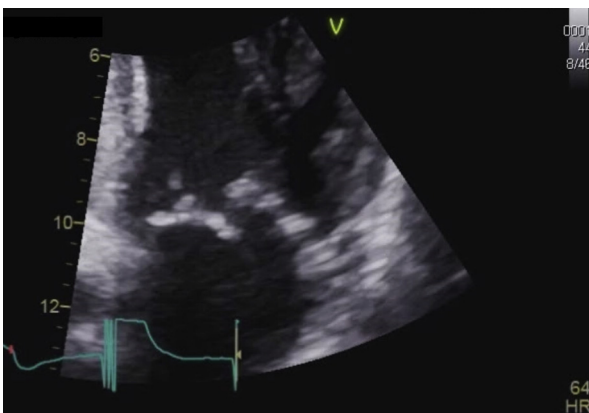


Figure 2 Transthoracic echocardiography, apical four-chamber view, demonstrating flail motion of the anterior leaflet of the mitral valve.

valve specimen revealed fibrocalcific valve tissue, myxoid changes, and no evidence of infection, thus confirming myxomatous degeneration as the etiology of mitral valve disease.

The patient was extubated and weaned off vasopressors on the fourth postoperative day. Postoperative transthoracic echocardiography demonstrated a normally functioning bioprosthesis in the mitral position and normal left ventricular systolic function (Video 8). The patient also underwent dual-chamber permanent pacemaker placement and was discharged home in a stable condition and continues to receive outpatient clinic follow-up.

DISCUSSION

We describe the case of an elderly woman who presented with right upper lobe opacity, constitutional symptoms, and hypotension, masquerading as right upper lobe pneumonia. Transthoracic and transesophageal echocardiography were crucial to arriving at the correct diagnosis of myxomatous degeneration and flail anterior mitral

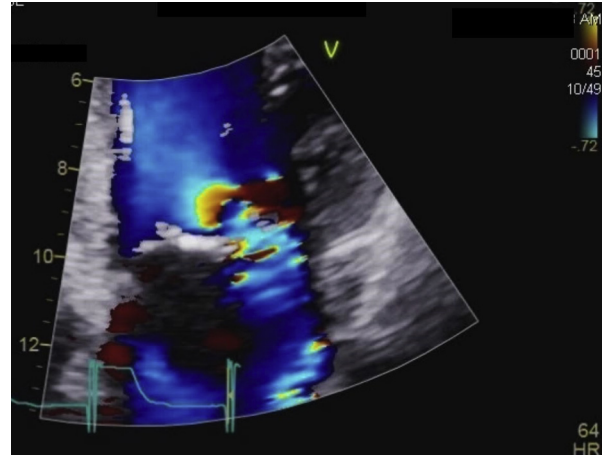


Figure 3 Transthoracic echocardiography, apical four-chamber view, with color Doppler demonstrating an eccentric, posteriorly directed MR jet.

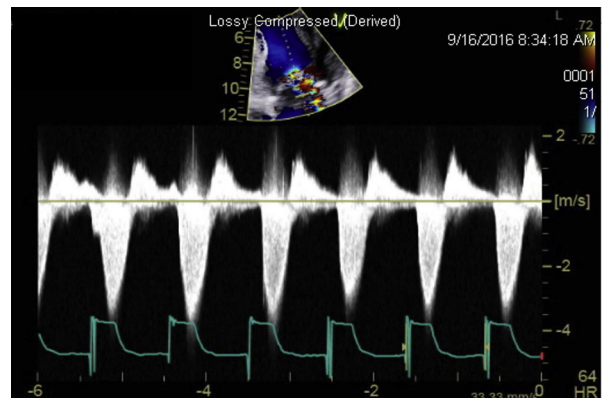


Figure 4 Transthoracic echocardiography, apical four-chamber view with continuous-wave and color Doppler across the mitral valve, demonstrating a dense, triangular, early peaking MR jet.

valve leaflet leading to severe acute MR, localized pulmonary edema, and cardiogenic shock.

In our patient, the diagnosis of acute MR was delayed by >36 hours from the initial presentation. This case highlights the importance of considering cardiogenic pulmonary edema as a potential etiology of unilateral pulmonary opacity.^{3,4} In a study of 131 patients with severe MR, Schnyder *et al.*⁵ reported that the prevalence of right upper lobe opacity was 9.2% (12 patients). Moreover, in a study of 869 consecutive patients admitted with cardiogenic pulmonary edema, Attias *et al.*⁴ found that 18 patients (2.1%) had unilateral pulmonary edema on chest radiography, of whom 16 (89%) had right-sided unilateral pulmonary edema, with the right upper segment being the most commonly affected. Alternatively, left-sided unilateral pulmonary edema was seen only in two patients (11%). In this study, all 18 patients with unilateral pulmonary edema had severe MR. Among these patients, 16 (89%) had posterior leaflet prolapse, and only two (11%) had anterior leaflet prolapse. There were 10 patients with organic MR, among whom eight had right-sided pulmonary edema on chest radiography from posterior leaflet prolapse, one patient had right-sided pulmonary edema from anterior leaflet prolapse, and one patient had left-sided pulmonary edema from anterior leaflet prolapse. Among the eight patients with functional MR, seven had right-sided

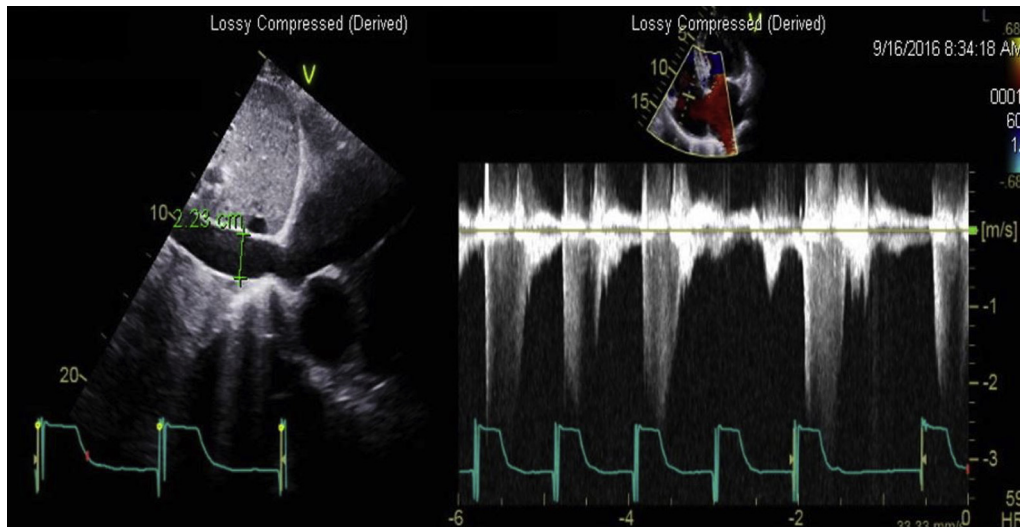


Figure 5 (Left) Transthoracic echocardiography, subcostal view, demonstrating dilated inferior vena cava. (Right) Transthoracic echocardiography, apical four-chamber view with continuous-wave Doppler across the tricuspid valve, demonstrating tricuspid regurgitation jet with estimated pulmonary artery systolic pressure in the range of 60 to 65 mm Hg.

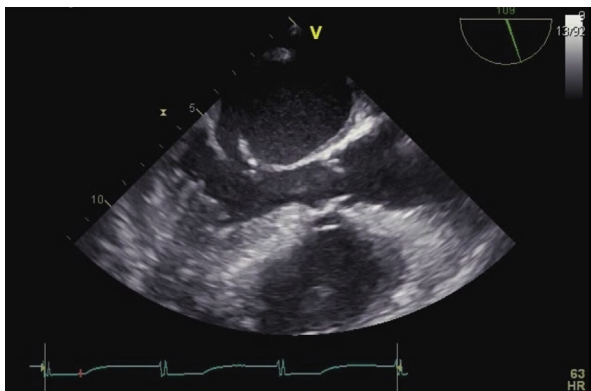


Figure 6 Transesophageal echocardiography, midesophageal long-axis view, demonstrating flail motion of the anterior leaflet of the mitral valve.

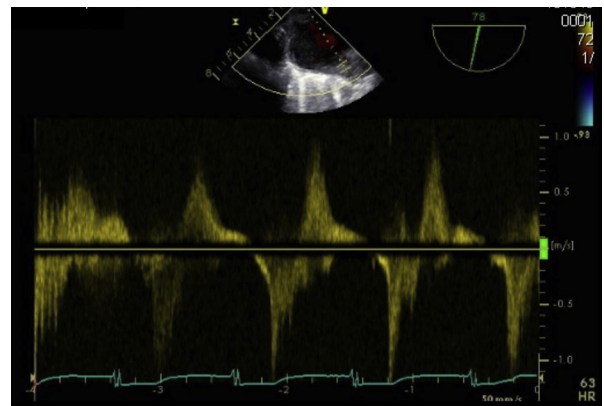


Figure 8 Transesophageal echocardiographic pulsed-wave Doppler of right upper pulmonary vein showing a systolic flow reversal.

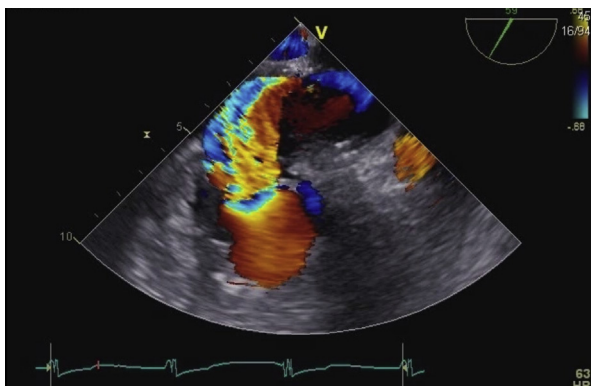


Figure 7 Transesophageal echocardiography, mitral commissural view with color Doppler, demonstrating an eccentric, posteriorly directed MR jet.

pulmonary edema, and one had left-sided pulmonary edema.⁴ In the study by Attias *et al.*, most cases of right-sided pulmonary edema were due to posterior leaflet prolapse. However, our patient had a flail ante-

rior mitral valve leaflet, which is an uncommon cause for right upper lobe opacity.

There are many theories as to why acute MR causes unilateral pulmonary edema.⁴⁻⁶ The most popular theory is that it is related to the direction of the regurgitant jet toward a pulmonary vein, leading to a selective increase in the hydrostatic pressure in one of the pulmonary veins.⁷ As the plane of the mitral valve faces posteriorly, superiorly, and to the right, the regurgitant stream is directed toward the right pulmonary veins and predominantly the right upper pulmonary vein. In studies evaluating the direction of the regurgitant flow, it was observed that anterior leaflet flail was associated with a posteriorly directed MR jet, while posterior leaflet flail was implicated with an anteriorly directed jet.^{6,7} Our patient had an eccentric jet, directed toward the posterior wall of the left atrium.

Our patient presented with subjective fever, chills, leukocytosis, and had no audible systolic murmur, which confounded the initial diagnosis. In the study by Attias *et al.*,⁴ when patients presenting with unilateral pulmonary edema were compared with those presenting with bilateral pulmonary edema, patients with unilateral pulmonary edema were found to have a higher rate of leukocytosis (72%

vs 40%, $P = .02$) and received antibiotics at a much higher rate (61% vs 6%, $P = .008$). In the same study, among patients with acute MR and unilateral pulmonary edema, fever on presentation was an uncommon finding, seen in only 11% of patients, and all the bacteriologic samples were sterile. In addition, among patients with acute MR and unilateral pulmonary edema, 44% had no murmur. Delay in treatment, defined as a >6-hour lapse without optimal medical therapy, occurred at a higher rate in patients with unilateral pulmonary edema (33% vs 4%, $P = .003$). In a retrospective study of 262 patients with severe MR by Zhou *et al.*,⁸ 60% of patients with acute flail mitral valve were initially misdiagnosed, with a maximum delay in diagnosis of 4 days. Among the patients with acute flail mitral valve, fever was the presenting symptom in 20% of the patients. Physical examination alone is not adequate to make a reliable diagnosis.^{4,6,8} In acute MR, the rapid equilibration of ventricular and atrial pressures during systole results in a faint systolic murmur rather than the holosystolic murmur that is usually heard in chronic MR.⁶ In summary, although presence of fever is unusual in patients with MR, the presence of leukocytosis and absence of systolic murmur are not uncommon.

The most common etiologies of acute MR are infective endocarditis and myxomatous degeneration of the valve, while collagen vascular disease, rheumatic fever, and spontaneous rupture of the chordae tendineae are less common.⁹ Myxomatous degeneration is the most common cause of MR requiring surgical repair in the elderly.¹⁰ In the case of our patient, myxomatous degeneration was the evident etiology.

CONCLUSIONS

Unilateral pulmonary edema, especially when involving the right upper lobe, should raise suspicion of acute MR as an etiology. This condition can be associated with fever and leukocytosis, which may confound the clinical picture. A flail anterior mitral valve leaflet is a rare etiology of this presentation. Prompt imaging with echocardiography can establish the diagnosis and guide a timely intervention.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at <https://doi.org/10.1016/j.case.2017.10.002>.

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