



## A Case of Acute Mitral Regurgitation in Systemic Lupus Erythematosus

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**Abstract :** Systemic lupus erythematosus (SLE) may have multiple cardiac manifestations. These involve pericarditis, myocarditis and endocarditis. Acute myocardial infarction has also been observed. Valvular abnormalities are relatively uncommon. Acute mitral regurgitation in SLE is a rare form of valvulitis than can lead to fatal pulmonary edema. We present an unusual case of male SLE with acute onset of severe mitral regurgitation leading to worsening congestive cardiac failure and death.

**Keyword :** Keywords Systemic lupus erythematosus, acute mitral regurgitation, pancarditis, cardiac failure

### Introduction

Many patients with SLE develop cardiac manifestations. Pericarditis is the most common among these, with a prevalence reported to be around 60%<sup>1</sup>. Myocardial involvement is uncommon. Valvular involvement can be demonstrated in up to 77 % of SLE patients. This remains mostly asymptomatic, however, symptoms can occur when there is valvular incompetence leading to pulmonary edema or cardiac failure. Mitral or aortic valves are commonly involved. High-dose steroid therapy can lead to development or worsening of the regurgitant lesions.

### Case

A 30-year old male presented to our Nephrology outpatient department with anasarca for 2 months. He was diagnosed to have SLE based on the presence of Antinuclear antibody (ANA) and double-stranded DNA (dsDNA) positivity, autoimmune hemolytic (direct Coomb's test positive) anemia and nephrotic range proteinuria. At diagnosis, his ANA was 4+, dsDNA titres were 605, and DCT was 2+. 24-hour urine protein was 4.1 g. He was diagnosed to have probable lupus nephritis based on presence of significant proteinuria and active urine sediment. Renal biopsy could not be performed in view of severe anemia, hemoglobin being 6.1 g%. He was initiated on steroids, Prednisolone at 1 mg/kg (50 mg once daily). Renal biopsy was planned after improvement of hemoglobin. 10 days after initiation of steroid therapy, he presented with cough with mild hemoptysis and worsening dyspnea over 5 days, along with high grade fever for 2 days. At presentation, his pulse rate was 150 per minute, blood pressure was 120/60 mm Hg, respiratory rate was 40 per minute and saturation on room air was 86%. He was febrile. Jugular venous pulse was not elevated. He had bilateral pitting pedal edema.

Respiratory examination revealed bronchial breath sounds in bilateral inter and infrascapular areas, with prominent findings on the right side. There were no murmurs on examination of the cardiovascular system. Other systemic examination was also normal. Chest X ray revealed a right mid and lower-zone consolidation.

He required ICU care where he was initiated on Piperacillin-Tazobactam with injection azithromycin for severe community acquired pneumonia. He required non-invasive ventilation for 3 days intermittently. He clinically improved with these measures. However, on the 3<sup>rd</sup> day, he developed worsening dyspnea with moderate hemoptysis with worsening oxygen requirements. In view of this deterioration, diffuse alveolar hemorrhage was considered highly likely, and he was initiated on methyl prednisolone pulse, 1 g once daily given for 3 days, with which he improved. He was also initiated on diuresis as he had developed fluid overload state during ICU admission. ECHO done in ICU at this time was normal. CT thorax could not be done as he was not clinically stable for the same. He was shifted to the general ward after stabilization. Cyclophosphamide pulse was given, along with continuation of oral steroids at 1 mg/kg. In the ward, he developed worsening orthopnea again, with elevation of JVP along with left ventricular third heart sound and a loud S2 along with mitral systolic murmur. He had extensive crepitations over both lung fields. Urgent Echocardiography showed severe mitral regurgitation with pulmonary artery hypertension. He developed worsening cardiac failure and required mechanical ventilation. He continued to deteriorate in ICU. The family decided on withdrawal of life-sustaining measures. These were withdrawn and he succumbed to his illness subsequently.

### Discussion

Cardiac involvement is common in SLE. This can involve the endocardium, myocardium or pericardium. Pericardial involvement in the form of pericarditis and pericardial effusion as part of serositis is the commonest cardiac manifestation. Myocarditis is rare but potentially fatal. Endocardium involvement can be in the form of valvulitis and valvular incompetence or endocarditis (Libbman-Sacks endocarditis). Valvulitis is mostly asymptomatic and may be demonstrable in upto 77 % of SLE patients<sup>1</sup>. Valvular incompetence leads to pulmonary edema and cardiac failure. Valves commonly involved are mitral and aortic valves. Valvular lesions are associated with IgG anticardiolipin antibodies and disease duration.

Chronic disease can be associated with valvular incompetence. However, acute valvular involvement is also described. Acute mitral regurgitation can be the presenting manifestation of SLE flare occurring with increase in disease activity<sup>2</sup>. A case report has also described the development of recurrent valvular incompetence in association with disease flare<sup>3</sup>. These patients present in acute pulmonary edema and features of cardiac failure. Acute mitral regurgitation has also been described to develop following administration of high dose corticosteroid therapy<sup>4</sup>. Adverse effects of steroid therapy on valvular abnormalities, leading to shrinkage and scarring of valves is well known, which may be the pathogenetic basis for this manifestation. Management involves repair of the mitral valve in these patients to avoid pulmonary edema<sup>5</sup>. Case reports describe management of acute mitral regurgitation with emergent mitral valve replacement.

#### **Conclusions**

Acute mitral regurgitation is a rare cardiac manifestation of SLE. It can be a presenting feature of an acute flare of SLE. It can also follow administration of high-dose steroid therapy. Management involves repair of the mitral valve. Conflicts of Interest: None

#### **References**

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