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CASE REPORT

Severe Mitral Valve Regurgitation Secondary to Fulminant Myocarditis in the Setting of a Lupus Flare

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Abstract

Systemic Lupus Erythematosus represents a chronic autoimmune disorder characterized by multiorgan involvement. Lupus myocarditis is a rare presentation of one of the cardiac complications of lupus with an incidence of 3–9%. It usually presents with non-specific symptoms such as dyspnea, orthopnea, chest pain, pedal edema, fever, diaphoresis, paroxysmal nocturnal dyspnea, nausea, vomiting, or palpitations. Even though endomyocardial biopsy is considered the gold standard diagnostic approach, other non-invasive diagnostic alternatives including cardiac magnetic resonance (CMR) have been studied.

Therapeutic interventions may range from high-dose steroids, and IVIG, to the most advanced strategies such as mechanical circulatory support including VenoArterial Extracorporeal Membrane Oxygenation (VA-ECMO), and Impella, among others.

Keywords: Lupus myocarditis, Cardiogenic shock, Endomyocardial biopsy, Cardiac magnetic resonance, Impella, VA-ECMO

1. Introduction

Systemic Lupus Erythematosus is a chronic autoimmune disorder characterized by the involvement of skin, joints, serous membranes, and kidneys in predominantly young women. Cardiac manifestations may occur in half of all patients, most commonly involving the pericardium but can involve any of the cardiac membranes. Lupus myocarditis is a rare presentation of one of the cardiac complications of lupus reported in only 9% of patients with lupus.[1]

Patients may present with complaints of dyspnea, orthopnea, chest pain, pedal edema, fever, diaphoresis, paroxysmal nocturnal dyspnea, nausea, vomiting, or palpitations.[2] Endomyocardial biopsy and echocardiogram remain to be the definitive disease diagnosis however recent evidence suggests CMR imaging with gadolinium enhancement may be a better modality.[3] Cardiogenic shock remains one of the rarer presentations of this disease with fulminant cardiogenic shock often requiring mechanical support (VenoArterial Extracorporeal Membrane Oxygenation [VA-ECMO], Impella, among others).[4]

We present a unique case of a 55 y/o lady with longstanding well-controlled lupus on steroid therapy who went into cardiogenic shock requiring pressor support and intubation, left heart catheterization showed clean coronaries and was found to have severe mitral regurgitation requiring transfer to a tertiary center for mechanical device placement.

2. Case presentation

The patient is a 55-year-old lady with a past medical history of Systemic Lupus Erythematosus (SLE) on prednisone 20 mg daily who presented to the Emergency Department (ED) with complaints of epigastric abdominal pain associated with non-bloody vomiting, watery diarrhea, generalized weakness, fever,

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and skin rash for 1 week prior to presentation. The patient reported taking prednisone for SLE for many years, however, she states he has not been taking her medication as indicated for the past few weeks and she has not followed up with her rheumatologist for over a year. She denied chest pain, dyspnea, orthopnea, cough, lower extremities (LE) swelling, recent sick contact, or recent travel. Vital signs showed a temperature of 102.9 °F, heart rate of 109 per minute, respiratory rate of 16 per minute, blood pressure of 85/60 mmHg, and 93% O2 saturation in room air.

Physical examination revealed Jugular Venous Distension (JVD) on the neck exam, heart exam was normal with S1−S2, no murmurs, rubs, or gallops, lung exam was normal, and the abdominal exam showed diffuse abdominal tenderness with no guarding or rigidity. LE exam showed warm and well-perfused extremities with no edema. A skin exam showed a petechial rash extending from the upper chest to bilateral LE. An electrocardiogram (ECG) showed sinus rhythm, left axis deviation, and non-specific ST and T wave abnormalities with no ST elevation or depression. Labs showed a white blood count of 11.7 [K/UL] with polymorphonuclears predominance (88.3%), hemoglobin of 10.6 [g/dL], erythrocyte sedimentation rate of 115, c-reactive protein >20 [ng/dL], troponin-I of 58.61 (normal <0.05 ng/mL), brain natriuretic peptide of 4545 [<100 pg/mL], creatinine of 2.2 [0.4−1 mg/dl], blood urea nitrogen of 33, and lactic acid of 2.1 [0.5−2.2 mmol/L]. The total complement level was 50 (31−60 U/ml), complement C3 level was 155 (83−193 mg/dl), and complement C4 level was 20 (15−57 mg/dl). Serology for coxsackie virus, parvovirus, and HIV was negative. Urinalysis showed microscopic hematuria and proteinuria. Chest X-ray showed a small right pleural effusion.

CT scan of the chest without contrast showed minimal bilateral pleural effusion, small pericardial effusion, and mild pulmonary vascular congestion. CT scan of the abdomen/pelvis without contrast showed no acute pathology. Transthoracic Echocardiogram (TTE) was done urgently and showed a left ventricular ejection fraction (LVEF) of 40−45%, global hypokinesis, moderate-severe mitral regurgitation, small pericardial effusion, moderately elevated pulmonary artery systolic pressure.

During her course of admission, she was given IV fluid resuscitation and treated with piperacillin-tazobactam 3.375 g. She remained hypotensive despite IV fluids and was started on Norepinephrine. Emergent Left and Right Heart Catheterization (LHC/RHC) was done. RHC hemodynamic pulmonary capillary wedge pressure was 28 mmHg in the left upper trunk and 30 mmHg in the left pulmonary trunk. Pulmonary artery (PA) pressure was 55/20 mmHg with a mean PA pressure of 26 mmHg. Right ventricular pressure was 55/10 mmHg. Right atrial pressure was 12 mmHg. Left ventricular (LV) cavity: no ventriculography was performed due to acute kidney injury. LV end-diastolic pressure (LVEDP) was 30 mmHg. The pre-A wave pressure was 20 mmHg with an extremely steep upslope after atrial contraction and the post-A wave pressure was 30−32 mmHg. A coronary angiogram showed normal coronaries with no obstructive lesions.

Post Catheterization, the patient was diagnosed with acute myopericarditis with severely elevated LVEDP with severe mitral regurgitation due to lupus flare. She remained hypotensive despite vasopressor support and received IV Methyl prednisone 250 mg. She was developing a picture of cardiogenic shock and cardiorenal syndrome. She was transferred to a tertiary center where an Impella device was placed along with VA ECMO. She received IV immunoglobulin (IVIG), and high-dose steroids, and needed to start hemodialysis. Renal biopsy was not pursued since her condition continued to deteriorate despite aggressive medical management and the patient's family opted for comfort care measures (See Fig. 1).

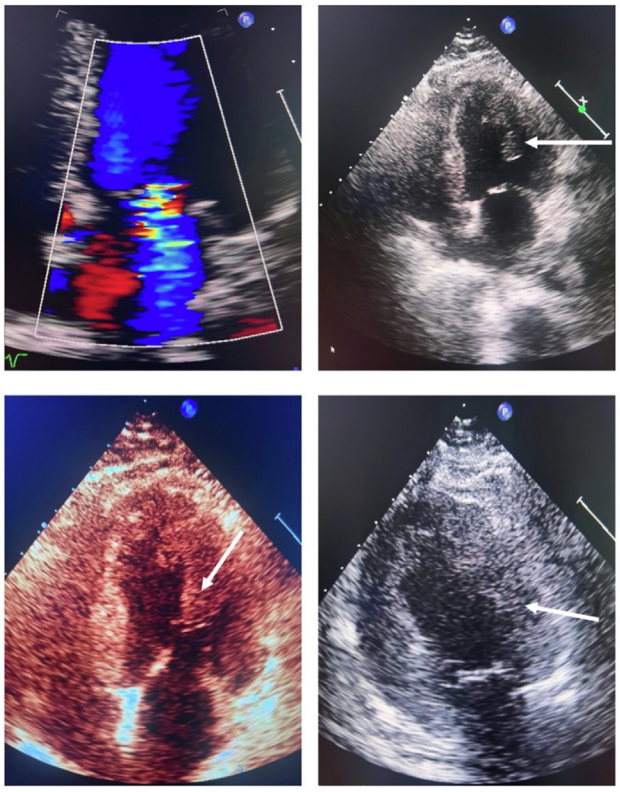
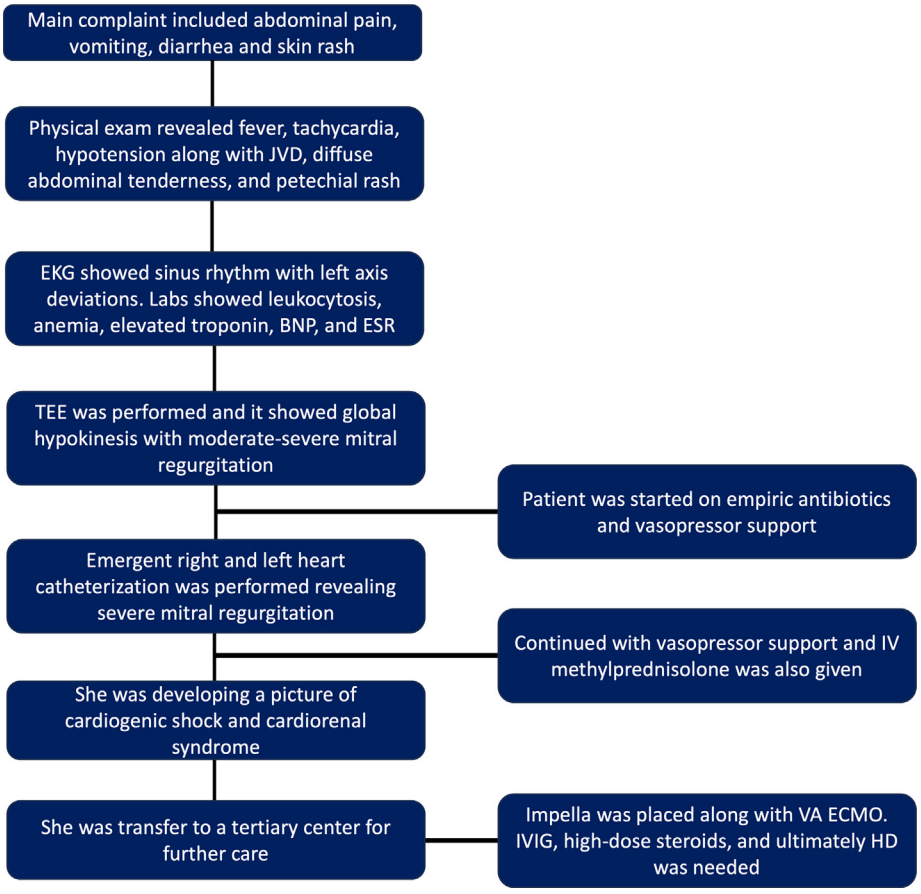


Fig. 1. Severe mitral regurgitation with markedly increased thickening of the left ventricular wall and papillary muscle with a bright signal suggestive of inflammatory infiltration.



3. Discussion

We present an unusual case of a 55-year-old lady with longstanding well-controlled lupus on steroid therapy who went into cardiogenic shock requiring mechanical circulatory support.

Lupus myocarditis is considered a life-threatening clinical entity due to its likely progression to dilated cardiomyopathy, cardiac arrhythmias, and heart failure with impending cardiogenic shock with a 10.3% overall mortality. The prevalence of this pathology has been determined to be around 9% of SLE patients. It may present with symptoms consistent with shortness of breath, chest pain, palpitations, and fever; laboratory findings occasionally include troponemia.[5]

Endomyocardial biopsy remains to be the gold standard diagnostic tool for confirmation, which usually reveals myocyte necrotic changes with immune complex deposition, patchy myocardial fibrosis, and interstitial mononuclear infiltrate. Due to its low sensitivity and potential for complications of the myocardial biopsy, other diagnostics modalities including echocardiography and CMR imaging have been proven to be useful in assessing biventricular function. An echocardiogram commonly shows global left ventricular dysfunction; however, regional wall motion abnormalities can also be evidence. Furthermore, CMR imaging can reveal the presence of necrotic or fibrotic changes evidenced by late gadolinium enhancement.[6]

Regarding therapeutic strategies, high-dose corticosteroids are considered a first-line option since has been proven to improve left ventricular function. Rituximab, intravenous immunoglobulins, azathioprine, and cyclophosphamide represent an alternative; however, the last two options carry a risk of significant myelosuppression.[7] Plasmapheresis (PE) is another therapeutic strategy available in cases of severe lupus flare. Notably, PE does not compromise the host's immune response, and adverse events associated with the procedure are infrequent. The primary indications for plasmapheresis include acute life-threatening manifestations and severe, therapy-resistant conditions seen in SLE patients. Examples of such manifestations encompass diffuse alveolar hemorrhage, neurolupus, thrombotic thrombocytopenic purpura,

catastrophic antiphospholipid syndrome, and refractory SLE renal disease. Additionally, plasmapheresis may be considered in pregnancy-related cases. This therapeutic approach aims to address critical and challenging aspects of SLE, offering a targeted intervention for cases that are unresponsive to standard therapies.[8]

Advanced therapy is often required in patients developing cardiogenic shock such as inotropic support (milrinone, or dobutamine) or mechanic circulatory support, including VA ECMO or Impella devices which may provide a bridge to heart transplantation. The Impella device is characterized by a catheter-based ventricular assist device that is placed in the left ventricle and pumps blood from the left ventricle into the ascending aorta keeping a systemic circulation between 2.5 and 5.0 L/min. This results in the unloading of the left ventricle, improving cardiac output and reducing pulmonary congestion and edema.[9] Extracorporeal Membrane Oxygenation (ECMO) tends to elevate afterload on the left ventricle, which, without additional left ventricular venting can lead to dilation and pulmonary edema hence Impella base strategies are usually paired with ECMO support (ECMELLA).[10]

To the best of our knowledge, there is only one systemic review and meta-analysis that compares Impella with VA-ECMO in the treatment of cardiogenic shock secondary to acute myocardial infarction and concluded that both treatment modalities are adequate however Impella was associated with a modest reduction in mortality and complication rates.[11] Further studies need to be performed for a better comprehension of the hemodynamic effects of the mechanical circulatory support systems and their impact on the myocardial preload and afterload for proper use in different clinical settings.

4. Conclusion

Lupus myocarditis remains one of the more challenging complications of this chronic disease given the relative paucity of literature and lack of clarity on treatment. While plasmapheresis, high-dose steroids, and immunosuppression with goal-directed medical therapy for acute systolic heart failure have resulted in improvement, mechanical support with a cardiac assist device remains the gold standard in cases such as these. An aggressive approach with all these modalities results in a good clinical outcome in a large number of patients. However, further studies need to be done to better define appropriate treatment modalities in cases of fulminant cardiac shock such as ours.

Conflict of interest

There is no conflict of interest.

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