



## Introduction

- Myocarditis is a rare complication of lupus.
- Presentation varies from asymptomatic to hemodynamic collapse.
- Prior to medical advances, diagnosis was made at autopsy.
- Cardiogenic shock may progress rapidly, with paradoxical complete recovery in patients who survive.
- Low-voltage EKG and elevated natriuretic peptides can be useful prognostically, although non-specific.

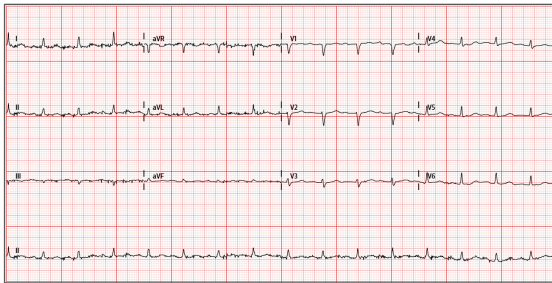


Figure 1. Admission EKG with low voltage in all leads.

- Gold standard = endomyocardial biopsy (EMB); not routinely performed due to low sensitivity and potential complications.
- Treatment consists of high-dose steroids with possible additional immunosuppression and standard cardiac management.
- However, little is known about ideal treatments and long-term recovery given rarity of condition.



Figure 2A & B. Erythematous maculopapular rash over the patient's abdomen, bilateral legs, & bilateral arms (not pictured), likely secondary to her lupus flare.

## Case Presentation

- 66 y/o woman presented with 6 weeks of progressive weakness, joint pain, dyspnea, diarrhea, & widespread rash (Figure 2A & B).
- Initial EKG showed low voltage in all leads (Figure 1).
- Initial echocardiogram showed normal LV ejection fraction (LVEF).
- She received 4L intravenous fluids for acute kidney injury, and high-dose prednisone and hydroxychloroquine for lupus.
- On day 4, the patient developed acute hypoxia, requiring 6L O<sub>2</sub> nasal cannula with chest X-ray showing new pulmonary edema (Figure 3).
- N-terminal pro-brain natriuretic peptide (NT-pro BNP) increased from 884 to 32,205.
- On day 6, the patient developed new atrial fibrillation and O<sub>2</sub> saturations dropped to 80's despite max Ventimask. She was treated with further IV diuresis, beta blockers, & bilevel positive airway pressure with minimal improvement.
- Bedside echocardiogram now showed severely reduced LVEF (20%) with globally weakened contraction.

Pulmonary artery pressure (PA)	Systolic: 23 mmHg	Diastolic: 15 mmHg
Central aortic pressure (AO)	Systolic: 92 mmHg	Diastolic: 81 mmHg
Stroke volume / pulmonary pulse pressure (SV/PP)	9.88 mL/mmHg	
Cardiac output (CO)	6.4 L/min	
Cardiac index (CI)	3.6 L/min	
Cardiac power output (CPO)	1.22 W	

Table 1. Heart catheterization with pressure readings consistent with cardiogenic shock.

- Shortly after, the patient went into cardiac arrest with return of pulse after 6 minutes of CPR.
- With cardiac catheterization pressures consistent with cardiogenic shock (Table 1), the patient was started on venoarterial extracorporeal membrane oxygenation (VA ECMO) and Impella for 7 days with gradual improvement in the cardiac ICU.
- EMB was not obtained because it would not change management.
- The patient was hospitalized for 6 more weeks, requiring intensive PT and respiratory conditioning, as well as medical management of lupus with IV cyclophosphamide and prednisone.

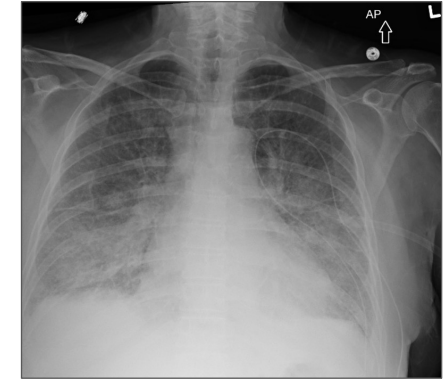


Figure 3. CXR demonstrated new-onset pulmonary edema after IV fluid boluses.

## Conclusion

- This case demonstrates an atypical presentation of lupus myocarditis as acute cardiogenic shock despite initial normal echocardiogram.
- It is unclear what triggered the acute decompensation on hospitalization day 4 that elicited rapid cardiac decline.
- Patient possibly had underlying myocardial inflammation and injury, given elevated NT-pro BNP and pulmonary edema.
- Thrombosis, infection, & cardiac ischemia were ruled-out.
- This case emphasizes the need to consider lupus myocarditis in a lupus flare with rise in NT-pro BNP, low voltage EKG, & poor tolerance to IV fluids, even in absence of initial changes in cardiac function on echocardiogram, to avoid potentially devastating outcomes of fulminant myocarditis.

## References

1. Kociol, Robb D., et al. "Recognition and initial management of fulminant myocarditis: a scientific statement from the American Heart Association." *Circulation* 141.6 (2020): e69-e92.
2. Thomas, Guillemette, et al. "Lupus myocarditis: initial presentation and longterm outcomes in a multicentric series of 29 patients." *The Journal of rheumatology* 44.1 (2017): 24-32.