CASE REPORT

LUPUS MYOCARDITIS WITH PERICARDIAL EFFUSION: A RARE PRESENTATION OF SYSTEMIC LUPUS ERYTHEMATOSUS

Saba Khalid[1], Kashif Ali Hashmi[2], Ammar Akhtar[2]
[1]Nishtar Medical University, Multan, Pakistan, [2]Chaudhary Pervaiz Elahi Institute of Cardiology, Multan, Pakistan

Systemic lupus erythematosus (SLE) can manifest with diverse cardiac presentations, with pericarditis being the most common. However, myocarditis, albeit rare, poses significant risks. Here, we present the case of a 20-year-old woman initially diagnosed with tuberculosis but later found to have lupus myocarditis, a rare complication of SLE. She presented with features of left ventricular failure and pericardial effusion, demonstrating the importance of considering alternative diagnoses in such cases. Prompt management with supportive care and oral steroids led to eventual improvement.

Keywords: lupus myocarditis; serositis; SLE; pericardial effusion

Citation: Khalid S, Hashmi KA, Akhtar A. Lupus Myocarditis with Pericardial Effusion: a Rare Presentation of Systemic Lupus Erythematosus. Pak Heart J. 2024;57(02):159-161. DOI: https://doi.org/10.47144/phj.v57i2.2743

INTRODUCTION

Cardiac involvement in systemic lupus erythematosus (SLE) is prevalent, with over 50% of patients affected. While pericarditis is the typical cardiac manifestation, myocarditis is less common, occurring in only 9% of cases.[1] A study in Central Punjab, Pakistan, reported cardiovascular system involvement in 55% of the local population, with 18.46% presenting with pericardial effusion.[2] Myocarditis in SLE can range from asymptomatic to life-threatening, necessitating early diagnosis and intervention to prevent complications such as heart failure and conduction defects. The pathophysiology involves immunological injury and ischemia. Steroids and immunosuppressants constitute the mainstay of treatment for SLE-induced cardiovascular manifestations.[3] Here, we present a case of a young woman with features of left ventricular failure and pericardial effusion, underscoring the importance of investigating the underlying cause of heart failure to optimize patient outcomes.

CASE REPORT

Patient Information: A 20-year-old unmarried female presented to the emergency department with shortness of breath and chest pain, which developed shortly after receiving a blood transfusion. She had a history of low-grade fever, lymphadenitis, hair loss, and discoloration and swelling of digits since childhood. She was previously treated with anti-tuberculous medication but developed rashes and oral ulcers after the first dose. On examination, she displayed signs of systemic lupus erythematosus and cardiac involvement, including pericardial effusion and left ventricular failure.

Clinical Findings: Upon examination, the patient was afebrile but hypotensive with elevated jugular venous pressure and muffled heart sounds. Bilateral crackles and decreased air entry were noted on lung examination, consistent with acute pulmonary edema. Electrocardiogram showed sinus tachycardia with low QRS voltages. Transthoracic echocardiogram revealed severe global left ventricular hypokinesia, severe right ventricular dysfunction, and moderate pericardial effusion.

Timeline: The timeline of events for the patient unfolds as follows: approximately 15 months prior to presentation, she experienced low-grade fever and lymphadenitis. Subsequently, she underwent anti-tuberculous treatment, which unfortunately resulted in the development of rash and oral ulcers. Her condition escalated when she presented to the emergency department following a blood transfusion, complaining of shortness of breath and chest pain. Consequently, she was admitted to the critical care unit, where clinical examination revealed hypotension, elevated jugular venous pressure, and muffled heart sounds. Further evaluation confirmed severe biventricular dysfunction and pericardial effusion. Treatment commenced with intravenous loop diuretics and standard heart failure therapy. Pericardial fluid analysis indicated inflammation consistent with lupus myocarditis and pericarditis, prompting the initiation of oral prednisolone alongside continued heart failure management. Over the course of one week, the patient exhibited improvement in symptoms and cardiac

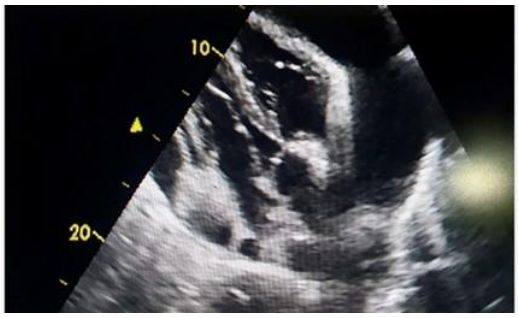
function. Subsequent follow-up in the outpatient department facilitated ongoing management and monitoring of her condition.

Diagnostic Assessment: Diagnostic assessments included transthoracic echocardiogram, electrocardiogram, chest X-ray, pericardial fluid analysis, and laboratory investigations for autoimmune markers. These tests confirmed severe biventricular dysfunction, pericardial effusion, and positive autoimmune markers indicative of lupus myocarditis and pericarditis.

Therapeutic Intervention: The patient was initially treated with intravenous loop diuretics and standard heart failure therapy. Upon confirmation of lupus myocarditis and pericarditis, oral prednisolone was initiated alongside continued heart failure treatment. Improvement was observed in symptoms and cardiac function following steroid therapy.

Follow-up and Outcomes: The patient was followed up in the outpatient department, where repeat echocardiography showed improvement in left ventricular systolic function and resolution of symptoms. This underscores the importance of early diagnosis and appropriate treatment in improving outcomes for SLE patients with cardiac involvement.


(A)



(B)

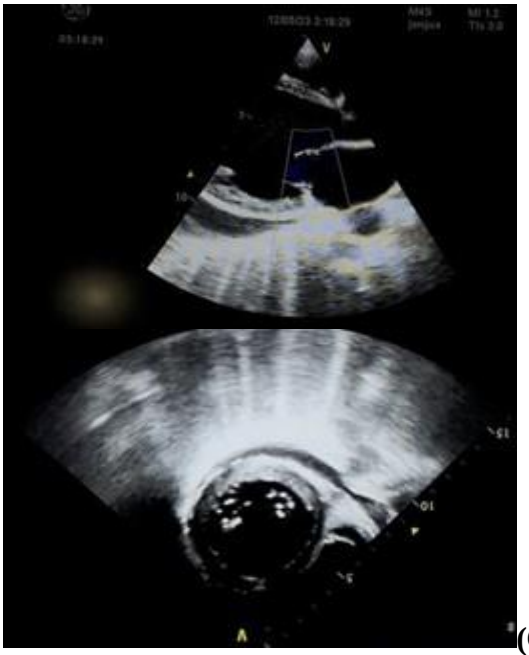

(C)

Figure 1: Patient with Lupus myocarditis with pericarditis A) Oral ulcers and facial rash B) Transthoracic echocardiography showing pericardial effusion C) Echocardiography after one week of initiating steroid therapy showing resolution of pericardial effusion

DISCUSSION

Systemic lupus erythematosus (SLE) manifests in various cardiovascular complications, encompassing pericarditis, cardiomyopathy, valvular dysfunction, coronary artery disease, conduction defects, thromboembolism, and myocarditis. While myocarditis is relatively rare in SLE, with a clinical prevalence of approximately 3-9%, its implications are severe, demanding vigilant clinical management due to the potential for lethal complications[4]. There exist reports of heart failure and acute myocarditis as the initial presentations of SLE, posing diagnostic challenges. For instance, Shamma Ahmed et al. documented cases where patients initially presented

with fever and lymphadenopathy, resembling our case.[5]

Symptoms of lupus myocarditis mirror those of myocarditis from other etiologies. Tailored treatment plans hinge upon symptomatology and the activity of concurrent lupus. High-dose corticosteroids, in conjunction with standard cardiac management, constitute the cornerstone of treatment.[6] In cases of steroid resistance, immunosuppressive therapy might be warranted. Our patient exhibited underlying left ventricular dysfunction attributed to lupus myocarditis, exacerbated by blood transfusion, but demonstrated a favorable response to steroid therapy.

CONCLUSION

This report underscores the importance of maintaining a high index of suspicion for lupus myocarditis in SLE patients presenting with typical rash alongside signs of heart failure. Early recognition and initiation of corticosteroid therapy are pivotal in improving outcomes in such scenarios.

DISCLAIMER

Informed consent for the publication of this case report, along with the patient's image, was obtained from both the patient herself and her brother

AUTHORS' CONTRIBUTION

SK, KAH, and AA: Concept and design, data acquisition, interpretation, drafting, final approval, and agree to be accountable for all aspects of the work. SK, KAH, and AA: Data acquisition, interpretation, drafting, final approval and agree to be accountable for all aspects of the work.

Disclaimer: None.

Conflict of interest: Authors declared no conflict of interest.

Source of funding: None.

Double blinded peer review history:
Submission complete: March 18, 2024
Review began: March 27, 2024
Revision received: June 11, 2024
Revision accepted: June 11, 2024

REFERENCES

1. Ward NK, Linares-Koloffon C, Posligua A, Gandrabur L, Kim WY, Sperber K, et al. Cardiac manifestations of systemic lupus erythematous: an overview of the incidence, risk factors, diagnostic criteria, pathophysiology and treatment options. Cardiol Review. 2022;30(1):38-43.
2. Kikuchi N, Watanabe E, Nagao M, Yoshizawa S, Kobashigawa T, Hagiwara N. Acute myocarditis complicating systemic lupus erythematosus: detection and evolution of transmural spiral late gadolinium enhancement on cardiac magnetic resonance imaging. Circulation: Cardiovascular Imaging. 2021;14(2):e011319.
3. Raza MA, Khan MIH. Systemic lupus erythematosus: disease manifestations in patients from Central Punjab (Pakistan). Int J Clin Rheumatol. 2012;7(6):607-14
4. Mohanty B, Sunder A. Lupus myocarditis-A rare case. J Fam Med Prim Care. 2020;9(8):4441.
5. Al-Nokhatha SA, Khogali HI, Al Shehhi MA, Jassim IT. Myocarditis as a lupus challenge: two case reports. J Med Case Rep. 2019;13(1):1-4.
6. Thomas G, Aubart FC, Chiche L, Haroche J, Hié M, Hervier B, et al. Lupus myocarditis: initial presentation and longterm outcomes in a multicentric series of 29 patients. J Rheumatol. 2017;44(1):24-32.

Address for Correspondence:

Prof. Kashif Ali Hashmi, Department of Cardiology, Nishtar Medical University, Multan, Pakistan.
Email: drkhashmi72@gmail.com