Case Report

Lupus myocarditis-A rare case

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Abstract

Myocarditis in patients of systemic lupus erythematosus is extremely rare and is potentially life threatening. This may be the first presentation of the disease. Here, we report a patient who presented with features of heart failure and was later diagnosed to have SLE.

Keywords: Cardiogenic shock, myocarditis, systemic lupus erythematosous

Introduction

Lupus myocarditis is a potentially life-threatening condition, clinically found in 3–9% of SLE patients.[1] However, the incidence is as high as 57% (post mortem analyses), suggesting a high prevalence of subclinical disease.[2] Patients may present with complains of dyspnea, orthopnea, chest pain, pedal edema, fever, diaphoresis, paroxysmal nocturnal dyspnea, nausea, vomiting, or palpitations. Myocarditis may progress to arrhythmias, conduction disturbances, dilated cardiomyopathy, and heart failure which need to be identified and treated early.[3] The pathophysiology of myocardial dysfunction in SLE is usually multifactorial. The prime factors involved are immunological injury and ischemia. Here, we are reporting a case who presented with features of heart failure and was initially thought to be due to diabetes and hypertension per se but was later diagnosed to have lupus myocarditis. She improved with steroids, immunosuppressive agents, and other supportive care.

Case Summary

A 40-year-old female was admitted with complaints of shortness of breath at rest for three days. She was diagnosed with type-2 diabetes mellitus, hypertension, and hypothyroidism and was on regular medication. She also had history of pain involving large joints along with low-grade intermittent fever.

Upon examination she was febrile and dyspnoeic. Pulse was 110/min, regular, and blood pressure was 90/50 mmHg. The jugular venous pressure was raised. There was mild pallor and bilateral pedal edema. There was no cyanosis, clubbing, icterus, or lymphadenopathy. Examination of chest revealed bilateral basal creptitions. Cardiovascular system examination revealed normal heart sounds. There was no pericardial rub, murmur, or gallop. Liver was enlarged and tender. Examination of the musculosleletal system showed Z-shaped thumbs, swan neck deformities of the distal interphalangeal joints, and reduced flexion at the metacarpophalangeal (MCP) joints [Figure 1]. X-ray of the hands confirmed nonerosive disease suggestive of Jaccoud arthropathy [Figure 2]. She also had oral ulcers.

Upon investigation, there was normocytic normochromic anemia with a high ESR of 60 mm. The CRP was high (17.38) with slightly raised creatinine and urine ACR was 486.71 mg/g. While in hospital, she deteriorated rapidly developing significant dyspnea and hypoxemia, hypotension, azotemia, and decreased urine output. She was shifted to the intensive care unit and started on noninvasive ventilation and needed double inotropic support.

ECG showed sinus tachycardia [Figure 3], chest X-ray revealed cardiomegaly, and bilateral pleural effusion [Figure 4]. Echocardiogram showed generalized wall hypokinesia with an

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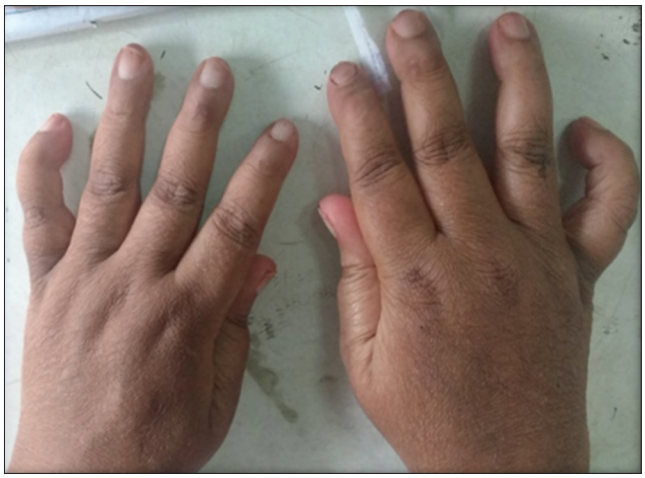

Figure 1: Hand deformity

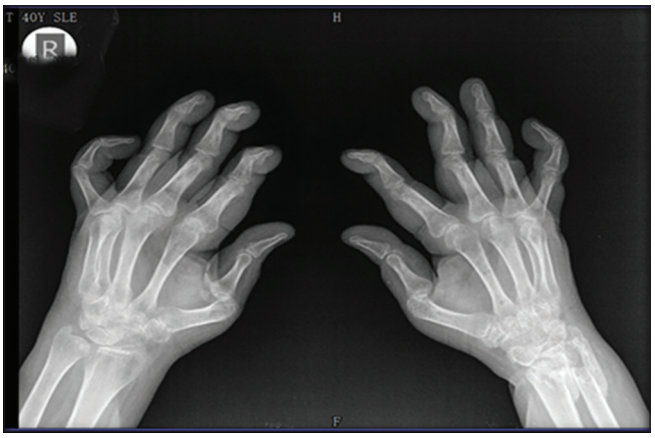

Figure 2: X-ray of hands showing jaccouds arthropathy

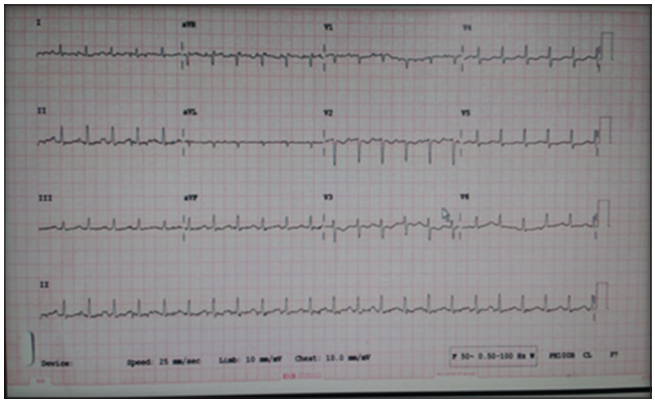

Figure 3: ECG-sinus tachycardia

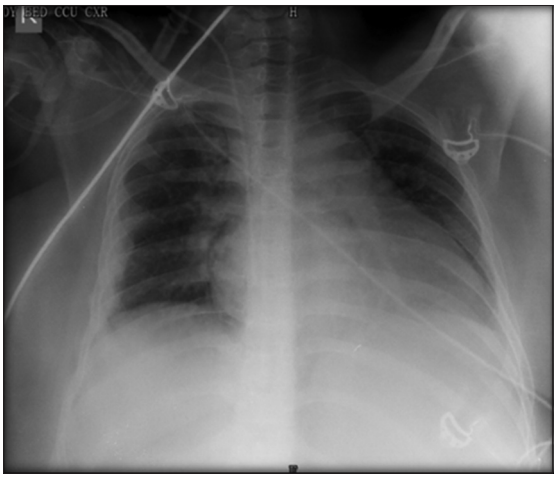

Figure 4: Chest X-Ray- cardiomegaly

ejection fraction of 36%, mild mitral and tricuspid regurgitation along with minimal pericardial effusion.

On further workup, ANA & DsDNA were positive. She was diagnosed with systemic lupus erythematosous. She was treated with intravenous methylprednisolone followed by oral prednisolone, hydroxychloroquine and mycophenolate mofetil. She improved and was shifted out of intensive care unit. Finally, she was discharged on mycophenolate mofetil, hydroxychloroquine, oral diabetic agents and deflazacort. She is on regular follow up and a repeat echo after three months showed normal ejection fraction (70%).

Discussion

Systemic lupus erythematosous patients can present with features of myocarditis, pericarditis, endocarditis, valvular lesions, and conduction abnormalities. Cardiovascular involvement in SLE might be associated with disease severity and activity.[4] Cardiac involvement as an initial manifestation of SLE is rare but when present it is associated with significant morbidity and mortality in 50% of cases.[5]

Endomyocardial biopsy is one of the modality for diagnosis of myocarditis but is less sensitive due to the focal nature of the disease. The histopathology report usually shows areas of myocardial fibrosis with sparse interstitial mononuclear cell infiltrates. There may be areas of occasional myocyte necrosis with immune complex deposition. The diagnosis is usually based on clinical and imaging studies. In our case, the diagnosis of SLE was made based on the basis of SLICC criteria. Our patient had features of serositis (pleural and pericardial effusions), positive ANA and anti-dsDNA. Jaccoud arthropathy (JA) with the involvement of the metacarpophalangeal (MCP), proximal interphalangeal joints and wrists was a clue in our case.

Primary lupus myocarditis is uncommon. There are few reports of heart failure and acute myocarditis as the initial manifestation of SLE.[5-11] Borenstein *et al.* found only five cases of myocarditis in 140 patients with SLE.[12] Badui *et al.* have reported 14% incidence in a prospective study.[13] Cheng *et al.* reported one such case for the first time who presented with severe LV dysfunction without other manifestations of SLE.[14] Tanwani *et al.* reported a series of patients of myocarditis in 2018.[15] Immune-complex deposition with complement activation is the primary pathogenetic mechanism in myocardial lesions.

Treatment with high-dose corticosteroids has been shown to improve left ventricular function in lupus myocarditis.[16,17] Other drugs used in treatment are azathioprine, cyclophosphamide, rituximab and intravenous immunoglobulins. Sandrasegaran *et al.* reported a patient of SLE presenting with myocarditis who responded dramatically to steroid therapy.[18] Two case reports were published by Shamma Ahmed *et al.* where it was a challenge to diagnose lupus myocarditis.[19]

There were challenges and difficulties in the diagnosis of lupus myocarditis mainly due to the presence of other factors potentially responsible for myocardial damage, such as anemia, hypertension, diabetes, and hypothyroidism. Initially, she had a slow response to therapy oriented with diuretics, intravenous vasodilators, and noninvasive mechanical ventilation but later on showed good response after the addition of intravenous methylprednisolone. Therefore, it is essential for the family physicians to identify and treat these cases early.

We followed up the case and with steroids and mycophenolate mofetil. Her left ventricular function improved dramatically to normal. Though myocarditis is a rare initial presentation of SLE, its timely diagnosis is of paramount importance to prevent morbidity and mortality. The mainstays of treatment are corticosteroids, immunosuppressive agents, and other supportive care.

Conclusion

The purpose of reporting this case is to emphasize the importance of systemic lupus as a differential diagnosis in cases who present with features of myocarditis along with comorbid conditions. A high degree of clinical suspicion, early diagnosis, and supportive care in combination with immunosuppressants may lead to favorable outcomes.

Institutional ethical permission

Taken

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patients have given their consent for their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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