

Case Report

Cardiogenic shock as the initial presentation of systemic lupus erythematosus

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ABSTRACT

Childhood onset systemic lupus erythematosus (SLE) is a rare entity and comprise less than 20% of all cases of lupus. Prepubertal onset is even rarer as most cases are reported in adolescent age group. SLE is an immune-complex mediated disease with multiorgan involvement. We report a case of a 12 year old female child with cardiogenic shock, myocarditis and pyrexia.

Keywords: SLE, Cardiogenic shock, Myocarditis

INTRODUCTION

Systemic lupus erythematosus (SLE) is an autoimmune disease that can involve any organ system, exhibiting great diversity in presentation. The onset can be variable and the symptoms can occur for many years. A review of recent studies shows that common initial presentations of childhood onset SLE (cSLE) include constitutional symptoms, renal disease, musculoskeletal and cutaneous involvement. In presenting this case and reviewing the literature, we emphasize the importance of cSLE as a differential diagnosis when presented with myocarditis and cardiac failure.

The case

A 12 year old female child presented with fever and shortness of breath for 8 months. Fever was continuous, high grade and child was regularly receiving antipyretics. Dyspnea was of grade IV with orthopnea. She had decreased appetite and documented weight loss. Over the last few days she was becoming increasingly fatigued and was bed ridden. No history of hair loss, oral ulcers, arthralgia or arthritis. The child was diagnosed with tubercular pericarditis and was started on ATT by a local

physician, on last month of treatment at the time of presentation.

On examination, child was in severe respiratory distress, febrile, hypotensive and had tachycardia. General examination revealed severe pallor and bilateral pedal edema with no significant lymphadenopathy. Non-blanchable hyper pigmented rash with scaling was seen over medial side of upper arm on both sides. Respiratory examination revealed decreased breath sounds at lung base on both sides which were also dull on percussion suggestive of bilateral pleural effusion. Cardiovascular examination revealed raised JVP, apex was hyperdynamic shifted outwards and downwards and visible pulsations noted all over precordium. Abdominal examination revealed moderate ascites and mild hepatosplenomegaly. Neurological examination was normal.

Initial supportive management included inotropic support with dobutamine and dopamine, broad-spectrum antibiotics, and blood transfusion.

Laboratory investigations revealed a normocytic normochromic anemia (5 gm/dl) with a decreased white cell count (200 cells/mm³) and normal differential. Bone

marrow study was normal with no features suggestive of malignancy. Erythrocyte sedimentation rate (ESR) was 80 mm/1st hour. Renal and liver function tests were all normal. Urinalysis was within normal limits and showed no proteinuria. HIV and Mantoux test were negative.

An admission electrocardiogram showed sinus tachycardia with nonspecific T-wave abnormalities and prolonged QT. Troponin-I values peaked at 0.65 ng/mL (normal <0.04ng/ml). Echocardiogram showed systolic dysfunction with left ventricular ejection fraction 30% to 35%; mild to moderate tricuspid, pulmonary regurgitation and small pericardial effusion without tamponade. CT scan of thorax and abdomen revealed no evidence of malignancy.

The patient's serum tested strongly positive (1:1280) for antinuclear antibodies (ANA) with homogenous pattern, anti-smith and anti-double stranded DNA antibodies were also strongly positive.

The diagnosis of SLE was established based on the positive clinical and immunologic findings. The patient satisfied 4 of the 17 Systemic Lupus International Collaborating Clinics (SLICC) criteria for classifying SLE, namely serositis, anemia with leucopenia and, thrombocytopenia, positive serum ANA, anti-sm, anti-dsDNA antibodies.

DISCUSSION

Acute myocarditis and heart failure are uncommon but potentially fatal initial manifestations of SLE. There are no specific signs and symptoms for lupus associated myocarditis and heart failure, but overall clinical manifestations are reflective of the degree of myocardial dysfunction.¹ The diagnosis is challenging due to the higher prevalence of viral myocarditis and idiopathic dilated cardiomyopathy. Manifestations of cardiovascular disease from systemic lupus erythematosus (SLE) range from subclinical findings to life-threatening conditions and may involve all cardiac tissue. In order of prevalence, findings may include pericardial disease, myocardial dysfunction, valvular heart disease, conduction system abnormalities, and atheromatous disease.¹⁻³ There are few reports of heart failure and acute myocarditis as the initial manifestation of SLE.⁴⁻¹⁰ These are not included within the standard diagnostic criteria for SLE developed by the American College of Rheumatology. Therefore, high suspicion is necessary, since timely therapy may help achieve full recovery.

Our patient presented with features of heart failure and shock with long duration of symptoms. She was initially treated for a suspected infectious etiology. Subsequent evaluation demonstrated lupus myocarditis leading to dilated cardiomyopathy. Treatment strategies in patients with lupus myocarditis and heart failure continue to be

supportive and empirical immunosuppression. Since this is an infrequent manifestation of lupus, there are no randomized controlled trials or standard guidelines available for the treatment of this condition. Most studies consist of case reports or series of cases that have demonstrated a favorable response to pulse corticosteroids alone, combined with cyclophosphamide or other cytotoxics such as azathioprine, along with supportive management.⁴⁻⁷

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