Circulation: Heart Failure

IMAGES AND CASE REPORTS IN HEART FAILURE

Cardiac Shock Revealing Systemic Lupus Erythematosus

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Pericarditis is the most commonly recognized cardiac complication in systemic lupus erythematosus (SLE). However, myocarditis with cardiac shock as the initial manifestation of SLE is uncommon. We describe a case of a 28-year-old man who presented cardiogenic shock revealing SLE.

CASE REPORT

A 28-year-old man, white, with a history of smoking, presented to the emergency department in December 2016 with dyspnea. Blood pressure was 165/75 mm Hg, pulse rate 113 beats per minute, body temperature 36°C, and oxygen saturation 100%. Cardiovascular examination was remarkable for signs of congestion with edemas of lower limbs. Skin examination revealed livedo on both feet (Figure 1). Initial laboratory data on admission showed the following: Troponin I 0.053 µg/L (N<0.034 µg/L), brain natriuretic peptide 3342 pg/mL (N<100 pg/mL), aspartate transaminases 2334 IU/L (N<34 IU/L) and alanine transaminases 1120 IU/L (N<55 IU/L), white blood cell count 3700/mm^3, hemoglobin 10.4 g/dL, platelet count 206 000/mm^3, serum creatinine 84.9 µmol/L. Urine dipstick was negative for protein and blood. The ECG showed sinus tachycardia, negative T wave in V_4 through V_6 leads, and incomplete right bundle branch block. The chest radiograph revealed cardiomegaly. Echocardiography demonstrated a dilated cardiomyopathy and a biventricular edema, with a left ventricle ejection fraction evaluated at 15% to 20% without pericardial effusion and no significant valvulopathy. Two days later, physical examination showed a rapid decrease in blood pressure (85/70 mm Hg). Heart rate was 133 per minute and diuresis 500 mL/d. Blood lactate level was 3.5 mmol/L. These features were suggestive with a cardiogenic shock and dobutamine infusion was introduced. Contrast-enhanced cardiac magnetic resonance imaging showed multiple linear late intramyocardial contrasting of both ventricles suggestive of myocarditis (Figure 2). Coronary angiography was not performed. Serum

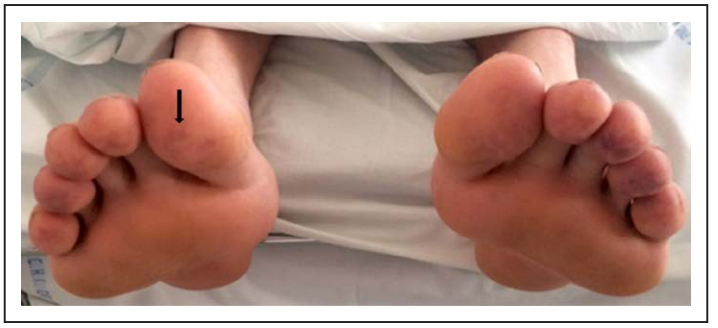

Figure 1. Livedo racemosa and cyanosis involving the toes. The black arrow shows the site of the biopsy.

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Key Words: glucocorticoids ■ heart failure ■ lupus erythematosus, systemic ■ magnetic resonance imaging ■ myocarditis

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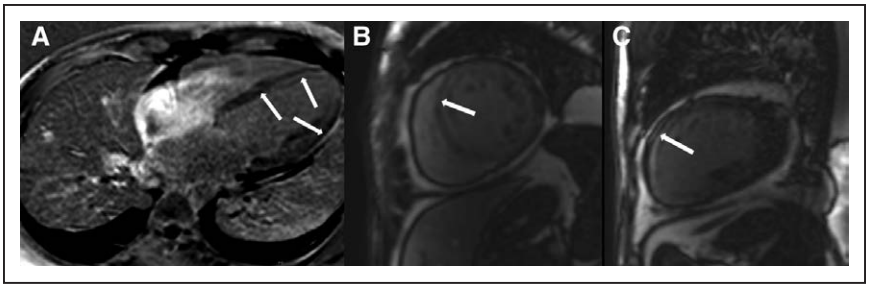

Figure 2. Cardiac magnetic resonance imaging in short-axis (A), 2-chamber (B), and 4-chamber views (C) (gadolinium-enhanced T1 weighted), showing delayed hyperenhancement in the lateral wall and in the septum with a patchy pattern (full white arrows).

was negative for antineutrophil cytoplasmic antibodies, but positive for antinuclear antibodies, 1 of 640 with native DNA antibodies at 93 IU/mL (N<10). CH50, C3, and C4 were, respectively, decreased at <11 IU/mL, 248 mg/L (N>660), and 18 mg/L (N>93). Serum protein electrophoresis showed polyclonal hypergammaglobulinemia. Anticardiolipin, anti-beta 2 glycoprotein I antibodies, and lupus anticoagulant were negative. Direct antiglobulin test was positive for IgG. Foot skin lesion biopsy showed a leukocytoclastic vasculitis and a positive direct immunofluorescence for IgM, IgA, IgG, and C3/C1q at the dermo-epidermal junction. Treatment was initiated with methylprednisolone 240 mg/d for 5 days, followed by oral glucocorticoids (1 mg kg^{-1} d^{-1}) and 6 intravenous pulses of cyclophosphamide 500 mg every 15 days for 3 months. A specific treatment of heart failure was also initiated with dobutamine and intravenous furosemide. Then, angiotensin-converting enzyme inhibitor, β-blocker, and mineralocorticoid receptor antagonist were introduced. At 1 month, the left ventricle ejection fraction was 50% to 55%, and the skin lesions were healed up. At 6 months, no relapse was noted and echocardiography showed the disappearance of the left ventricular edema and a significant improvement of the left ventricle ejection fraction and the longitudinal strain.

DISCUSSION

Cardiac involvement is the second most frequent manifestation behind kidney involvement, in SLE, and being diagnosed in nearly 50% of patients. Myocarditis occurred in 3% to 15% of cases and was the first manifestation of SLE in more than half of described cases.[1] However, acute myocarditis complicated with cardiogenic shock, and revealing an SLE is rare.

Unexplained fever, dyspnea, palpitations, tachycardia not caused by fever, ventricular gallop rhythm, cardiomegaly, conduction disturbances, new murmurs, an abnormal ECG, or cardiac failure in the setting of SLE should suggest the possibility of myocarditis.

Circulating autoantibodies, including anti-Ro (SS-A), anti-ribonucleoprotein, and antiphospholipid antibodies may be involved in lupus myocarditis. Some studies reported an association between anti-Ro antibodies and myocarditis in patient with SLE. However, these data were controversial. Finally, specificity of circulating antibodies is not sufficient to suggest myocarditis in patient with SLE.

There are nonspecific findings on ECG. Moreover, echocardiographic studies cannot definitely diagnose myocarditis.

Cardiac magnetic resonance imaging is the most sensitive noninvasive technique to investigate myocarditis. However, magnetic resonance imaging alone cannot differentiate viral myocarditis from other causes of acute dilated cardiomyopathy. The gold standard for confirmation of myocarditis is endomyocardial biopsy. This invasive procedure had limitations, such as the focal nature of myocarditis and the risk of complications including cardiac perforation and stroke. Recently, a new technique with robotic-assisted myocardial biopsy was reported, especially for involved area that may be difficult to reach with standard biopsy.[2] Further studies are needed to explore this new procedure in patients with myocarditis.

The differential diagnosis of livedoid lesion with myocarditis includes systemic vasculitis (eosinophilic granulomatosis with polyangiitis, microscopic polyangiitis, granulomatosis with polyangiitis, polyarteritis nodosa, and cryoglobulin vasculitis), infectious diseases (parvovirus B19), or drug hypersensitivity. Skin lesion, histology, and immunologic testing led us to the final diagnosis of SLE. Skin involvement in lupus is frequent, and various types of lesions have been described. The presence of livedoid lesions in SLE is either secondary of a lupus-induced vasculitis (leukocytoclastic vasculitis) or of a thrombosis associated with antiphospholipid syndrome.

Current treatment strategies are based on clinical experience rather than randomized trials. Treatment of lupus myocarditis should include corticosteroids. Few case report of severe form of lupus myocarditis showed promising results for the use of intravenous pulse corticosteroid (eg, methyl prednisolone of 1.0 g/d for 5–7 days) followed by high oral doses (eg, prednisolone; 1 mg kg^{-1} d^{-1}). Immunosuppressive agents such as azathioprine or cyclophosphamide and intravenous immunoglobulin have shown beneficial effect. Rituximab has been successfully used in only 1 pediatric case of SLE myocarditis. Finally, treatment regimens were not standardized. Because cyclophosphamide may cause sev-

eral serious side effects, in particular ovarian toxicity, it should not be a standard of care in the setting of lupus myocarditis and should be reserved in severe patients with altered left ventricular ejection fraction.[3]

The outcome of myocarditis is variable. Comorbidities, such as hypertension, may unfavorably influence the prognosis and black ethnicity and high disease activity at diagnosis were associated with the occurrence of myocarditis.[4]

CONCLUSIONS

SLE has to be considered in cases of myocarditis with cutaneous lesion, as livedoid. Early diagnosis and prompt treatment with high-dose steroid in addition to cardiac treatment may result in good outcomes.

ARTICLE INFORMATION

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Disclosures

None.

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