Myocarditis in a girl as an initial manifestation of systemic lupus erythematosus

Sirs,
Although only one of the 11 criteria for the classification of lupus describes cardiac involvement, it is known that the heart is frequently affected by this immunological disorder (1). However, acute myocarditis with severe cardiac failure is very uncommon. Myocarditis as the initial manifestation of lupus is still rare. We report a case which presented initially with severe myocardial failure without classical clinical stigmata of lupus.

A 16-year-old girl was admitted to our hospital because of palpitations, peripheral edema, and dyspnea of 2 days duration. There was no history of fever, arthralgias or oral ulcers. Physical examination disclosed a dyspneic patient. The pulse rate was 120/min, regular in rhythm, and the arterial pressure was 100/60 mmHg. There was jugular venous distension and a gallop rhythm. No murmur was audible. There was no pericardial rub. The ECG showed sinus tachycardia. The chest X-ray revealed cardiomegaly and pulmonary venous hypertension. Laboratory investigations showed elevated serum creatinine (4.5 mg/dl) and hyperkalemia (5.9 meq/L). The hemoglobin was 6.5 g/dl, and leukocytes 6,800/mm³ with 10% lymphocytes. The urinalysis revealed proteinuria 3+, hematuria, and granular casts. An urgent echocardiogram demonstrated global hypokinesis with an ejection fraction of 28%. A small pericardial effusion was present with no evidence of cardiac compression. The patient responded rapidly to diuretic agents, vasodilator drugs, digitalis, and blood transfusion. A diagnosis of lupus was suspected due to the presence of serositis, lymphopenia, and renal damage. The patient was found to have anti-dsDNA antibodies of 50 IU/ml measured by enzyme immunoassay (normal value 0–3.0). Complement (C3) level was 66 mg/ml by radial immunodiffusion (normal value 70–176 mg/ml). The patient was given intravenous methylprednisolone 500 mg/day for 5 days and then oral prednisone 50 mg/day. The patient was discharged after 10 days to continue diuretics, digitalis, vasodilator drugs and oral prednisone. Her serum creatinine had decreased to 2.2 mg/ml. Four days later she experienced a recurrence of her symptoms due to the fact that she had discontinued diuretic agents and oral prednisone. Chest radiographs showed acute pulmonary edema. Diuretics agents and inotrope infusion were commenced. Because of her respiratory distress intubation and mechanical ventilation were required. Intravenous methylprednisolone (500 mg/day) was added. The cardiac failure resolved and at present she is receiving hemodialysis and oral prednisone.

Lupus myocarditis is a life-threatening complication of lupus (2). Clinically overt myocarditis is uncommon and is not considered in the diagnostic criteria for the classification of lupus (3). The pathogenesis of myocarditis in lupus has been ascribed to autoimmunity, drugs, and coexisting diseases (3). However, myocarditis as the initial manifestation of lupus is still rare. Cheng et al. (4) reported a patient who presented with severe LV dysfunction but no other manifestations of lupus. Sandrasegaran et al. (5) published the case of a 46-year-old woman with subclinical lupus who presented with acute myocarditis. She responded rapidly to high dose steroids. Cases of LV failure secondary to myocarditis complicating lupus in adults and children have been reported (2, 6).

Our patient was not known to have lupus. She presented with severe heart failure, dyspnea, gallop rhythm, renal insufficiency, and tachycardia not due to fever. Although echocardiography did not establish the diagnosis of myocarditis, it provided evidence of global wall motion abnormality indicating myocardial inflammation. Acute myocarditis was accompanied by pericarditis. The possible immunological cause was confirmed by the presence of high anti-dsDNA levels and hypocomplementemia. She improved rapidly with standard therapy which included high-dose corticosteroids and cardiac drugs. Unfortunately, her renal condition could not be resolved.

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References