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CASE REPORT

Acute Aortic Valvulitis as an Initial Presentation of Systemic Lupus Erythematosus

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SUMMARY **Valvular involvement in patients with systemic lupus erythematosus (SLE) is not uncommon but patients rarely present with it. The mitral valve is most commonly involved. We report a 36-year-old man who had an episode of acute fever, arthritis, and acute aortic insufficiency with a small vegetation at the tip of the aortic valve mimicking infective endocarditis, proven later to be due to SLE. SLE should be considered as one of the uncommon causes of acute aortic insufficiency.**

Systemic lupus erythematosus (SLE) is a connective tissue disease, characterized by the presence of autoantibodies directed against cellular antigens, resulting in inflammatory damage to multiple organ systems. Cardiac involvement is one of the common clinical manifestations, contributing to significant morbidity and mortality. The cardiac pathology is pancarditis, which can affect pericardium, myocardium, endocardium and coronary arteries. Libman-Sacks verrucous vegetation is the pathognomonic valvular lesion of SLE, mostly affecting the mitral valve.[1,2] It is characterized by multiple pea-sized, flat or slightly raised granular lesions adhering to the endocardium, occurring on the ventricular surface or at the atrioventricular valves. Aortic valve involvement is uncommon. Valvular involvement rarely presents as an initial manifestation of SLE, but when it does present, it is generally considered to have minimal hemodynamic disturbances.[3,4] We report a patient with SLE who presented with fever and acute aortic insufficiency mimicking acute infective endocarditis.

CASE REPORT

A 36-year-old military officer, who had been healthy, had a history of fever, pain in the small joints of his hands, and increasing dyspnea on exertion, which he had had for 1 month. A few days before admission, he experienced orthopnea and could walk for only a short distance. He had been smoking and drinking for 10 years. There was no history of alopecia, oral ulcers, skin rashes or Raynaud's phenomenon. Family history of coronary heart disease or high blood pressure was negative.

His vital signs showed a temperature of 38.5°C, blood pressure of 160/60 mmHg, pulse rate of 110/minute and regular, and respiratory rate of 24/minute. A few faint maculopapular rashes on his face and discoid lesions on both ears were noted. The neck vein was engorged up to the angle of the mandible. The heart was enlarged with its apex at the 6th intercostal space anterior axillary line. A grade III diastolic blowing murmur was heard at Erb's point. Fine crepitation was noted in both

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basal lung fields. The liver was 2 cm below the right costal margin. A mild pitting edema was noted on both legs. A mild degree of arthritis was noted on wrists and small joints of hands. The water hammer pulse, Deroziez's sign, and the pistol-short sounds were positive.

A chest radiograph demonstrated a moderate degree of cardiomegaly with pulmonary venous congestion and minimal right pleural effusion. An echocardiographic study showed minimal enlargement of the left ventricle with an ejection fraction of 46%. There were severe retraction and thinning of the left aortic cusp. The right aortic cusp showed abnormal folding, and a small vegetation was noted at its tip (Fig. 1). A mild tricuspid regurgitation was also observed. A color Doppler study showed severe aortic regurgitation. An antinuclear antibody (ANA) test showed a speckled pattern. Anti ds-DNA antibody was positive. C3 was 68 μg/ml (normal 550-1200), C4 was 74 μg/ml (normal 200-500) and CH_{50} was 0 U/ml (normal 20-40). A complete blood count, urine analysis, blood and urine cultures, renal and liver functions, serum uric acid, Widal's, Weil-Felix, VDRL, anti-streptolysin-O (ASO), anti-Sm, anti-RNP, anti-Ro/SSA, anti-La/SSB, and anti-phospholipid (APL) antibodies were all normal or negative.

Hydrochlorothiazide 50 mg and enalapril 5 mg were given orally to control heart failure. Intravenous ampicillin 12 gm and gentamicin 240 mg were prescribed for possible infective endocarditis. During these therapies, an acute abdomen developed. A computed tomography of the abdomen showed thickened bowel wall, splenomegaly and ascites. An exploratory laparotomy showed multiple hemorrhagic spots on the small bowel walls without perforation. One week later, when the culture results were all negative

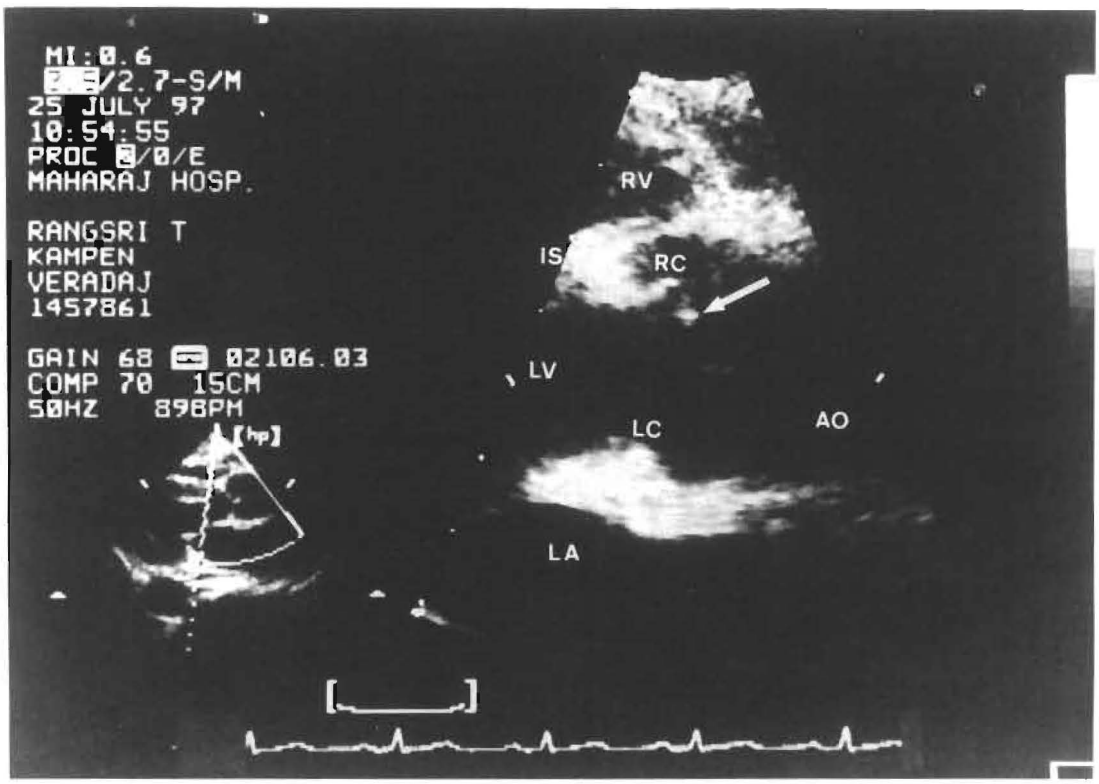

Fig. 1 An end systolic parasternal long-axis two-dimensional echocardiogram showing severe retraction and thinning of the left aortic cusp (LC) and abnormal folding of the right aortic cusp (RC). Note a small nodular vegetation (arrow) at the tip of the right aortic cusp. RV = right ventricle, LV = left ventricle, LA = left atrium, AO = aorta, IS = interventricular septum.

and the autoantibodies results were available, the diagnosis of SLE was made and prednisolone 50 mg orally was prescribed to control lupus carditis. Fever disappeared the next day with gradual improvement of the clinical symptoms. The patient showed progressive improvement of his cardiac function during initial outpatient follow-up. Prednisolone was gradually tapered off. Unfortunately, he was lost to follow-up after 6 months.

DISCUSSION

Cardiac involvement in SLE had been studied by many authors. Doherty *et al.*[2] reviewed autopsy studies of cardiovascular manifestations in SLE and found that pericardium, myocardium and endocardium were involved in 62%, 40% and 43% of cases respectively. The incidence of Libman-Sacks endocarditis declined from 59% in the pre-steroid era to 36% in the steroid era. The mitral valve was most commonly affected followed by the aortic, tricuspid and pulmonary valves. With the prolonged survival of patients with SLE and the improvement in diagnostic techniques, especially two dimensional echocardiography and color Doppler studies, cardiac disease in live SLE patients has become more apparent. The incidence of valvular involvement in live SLE patients has been reported to be 7.3-68%.[3-7] The mitral valve has been most commonly involved; aortic valve involvement has been uncommon. Valvular insufficiency has been the most common finding, but valvular stenosis or the combination of valvular insufficiency and stenosis can occur. The great variation in the incidence of valvular lesions has been related to the criteria for selection of patients and the diagnostic techniques used to evaluare heart disease. Approximately 1-2% of patients with SLE have developed hemodynamically significant valve disease that has required surgery.[3,7]

Galve *et al.*[4] described two major types of echocardiographic findings of valvular lesions in SLE. They found that Libman-Sacks endocarditis, which is pathognomonic for SLE, usually developed early in the course of the disease, while thickened valves usually developed late in the course of the disease. They suggested that the thickened valvular lesions might be considered as the healing stage of Libman-Sacks endocarditis, as evidenced by the older age of the patients, the longer duration of SLE, and the greater amount of corticosteroids received by this group of patients. Moreover, these thickened valve lesions tended to progress and these patients had more significant hemodynamic dysfunction requiring valve replacement. These results differed from those of Roldan *et al.*,[8] who recently found that both valvular thickening and vegetation were common in their SLE patients. These valvular abnormalities could disappear, or persist but change in appearance or size between the initial study and a mean follow up of 6 years. Neither the clinical nor immunological markers of the activity of SLE nor its treatment appeared to be related to the presence of or changes in valvular diseases.

The diagnosis of SLE at the time of presentation in this case was difficult. First, the patient was male, the uncommon sex for SLE. Second, acute aortic insufficiency mimicking infective endocarditis, which was the only major manifestation seen in this case, is an unusual presentation of SLE. Acute aortic insufficiency has been reported as a principal manifestation in 3 patients with SLE, who all died shortly after the development of aortic insufficiency.[9] The pathology showed marked thinning of the valves and perforation of the cusps. Cases of SLE complicated by infective endocarditis have also been described.[10,11] However, the negative blood cultures and ASO test made the diagnosis of infective endocarditis and acute rheumatic fever in this case less likely. The only clues suggesting SLE in this case were the presence of the diffused maculopapular rash on his face and the discoid lesions on his ears. This was supported by the presence of ANA and anti-ds DNA antibodies.

An association between heart valve disease and the presence of APL antibodies has been recently reviewed.[12,13] However, many controlled studies in patients with SLE have conflicting results about this association.[7,12-15] The significance of APL antibodies in the pathogenesis of valvular lesions is not clear. The deposition of immunoglobulin and complement in the valvular lesions suggest that immune complexes may play a role in growth and proliferation of vegetation, and APL antibodies may contribute to further thrombosis of the cardiac valves. These antibodies were not found in our patient.

In summary, we describe a case of SLE who presented with acute aortic valvulitis mimicking infective endocarditis, which was

later proven to be due to SLE. SLE should be considered as one of the unusual causes of acute aortic insufficiency.

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