Libman-Sacks Endocarditis in SLE

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Abstract

Systemic lupus erythematosus (SLE) is an autoimmune disease in which organs and cells undergo damage mediated by tissue-binding auto-antibodies and immune complexes. The diagnosis of SLE requires four or more of the American College of Rheumatology (ACR) criteria. In our patient, the diagnosis of SLE was based on the following four criteria: renal involvement, haematological abnormalities, positive ANA, and positive double-stranded DNA.

Key words : SLE. Libman-Sacks Endocarditis

Introduction

The characteristic cardiac valvular manifestation in SLE is Libman-Sacks (verrucous) endocarditis, and its reported incidence varies widely. It was first described in 1924 by Emanuel Libman and Benjamin Sacks, in which they characterized non-bacterial verrucous valvular disease in 4 patients with SLE.

According to the literature, the prevalence of cardiovascular involvement in patients with SLE has been estimated to be more than 50%. Valvular involvement is the most commonly encountered form of heart disease in SLE. Thickening of the valves is encountered more frequently (51~52%) than valve masses/vegetations (34~43%).[1] Functionally, valvular regurgitation has been reported to occur in up to 74% of patients, 7~41% of cases having moderate or severe regurgitation, while valvular stenosis is seen in only 3~4% of patients and usually accompanies regurgitation. Involvement of the mitral valve is most frequently encountered. However, any valve or multi-valvular affection may occur.[2]

Case History

A 15 yr old, adolescent female was admitted with h/o peri-orbital puffiness, oliguria since 5-6 months. Patient also had h/o swelling over both feet since 5 months. There was no h/o breathlessness, joint pain, photosensitivity, seizure, psychosis, and headache. There was no significant past history of any other illness. There was no history of drug consumption.

On examination, patient was afebrile, pulse was 90/min, regular, all peripheral pulsations were well felt and were bilaterally equal. BP was 104/70mm of Hg. She had mild pallor. There was no icterus, cyanosis or lymphadenopathy. There was no joint tenderness or joint swelling. There was no e/o oral or nasopharyngeal ulcerations, any rash or s/o congestive cardiac failure. Respiratory, CVS, CNS, P/A examination was within normal limit. Based upon history and clinical findings, patient was thoroughly investigated.

Investigations

Her Hb was 8.9gm%. Total leucocyte count (TLC) was 4200/mm^3, differential count (DLC) was- Polymorphs=50%, Lymphocytes=40% and platelet count was 81,000/mm^3.Urine-albumin was ++, urine-sugar was nil. Renal function was impaired. Blood urea was 60mg/dl and sr. creatinine was 1.8mg/dl. Liver function tests (LFT) were normal. Fasting lipid profile showed no abnormality apart from hypertriglyceridemia (290mg/dl). ECG was normal. Anti-nuclear antibody (ANA) was positive. Double-stranded DNA (dsDNA) was also strongly positive.(50.75 IU/ml, <20- Negative, >20- Positive) 24 hr proteinuria was 0.656 gm/day. Antiphospholipid and

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Anticardiolipin antibodies were negative. 2D Echocardiography was done which showed e/o endocarditis in the form of vegetation of 5mm on PML on LA side with trivial MR. Blood culture was done as per protocol and all 3 samples were negative.

Based on these findings, diagnosis of SLE with lupus nephritis with Libman-Sacks endocarditis was made.

Treatment Given

Patient was started on T. Prednisolone (1mg/kg/day) along with diuretics (T.Furosemide). Her general condition improved, edema subsided. Patient was started on maintenance dose of T. Prednisolone (5mg/kg/day) and has lost to follow-up

Discussion

In patients with SLE, the clinical expression of the musculoskeletal and muco-cutaneous disease predominates, even in patients who have cardiovascular involvement.[3] However, in our case, there were no musculoskeletal and mucocutaneous presentations clinically. She, in fact, presented with renal and cardiovascular involvement.

Cardiac manifestations may be numerous and can involve many components of the heart, including the pericardium, conduction system, myocardium, heart valves, and coronary arteries.[2]

The mechanism for the development of valvular damage in SLE is not completely understood. Vegetations consist of fibrin and thrombus with minimal inflammatory infiltrate. The presence of antiphospholipid and anticardiolipin antibodies may have a role in the pathogenesis of Libman-Sacks endocarditis through the initiation of nonbacterial thrombotic endocarditis. It is thought that immune complex deposition and complement activation cause acute, chronic or reccurent inflammation of the valve leaflets[4] . These processes can lead to valvular regurgitation, stenosis or both. The use of steroids in the management of SLE nephritis seems to have decreased its prevalence.

Libman-Sacks vegetations are described on TEE as being usually smaller than 1 cm2, with irregular borders, heterogenous echogenecity and no independent movement. In contrast, infective vegetations have a homogenous echogenecity, and present with vibratory or rotatory motion independent of leaflet motions[3]. Echocardiographic findings are nonspecific, and a definitive diagnosis can only be made on pathologic examination of the affected valves.

Treatment of valvular manifestations of SLE depends on the type and severity of involvement. Some investigators have suggested that the introduction of corticosteroids as treatment of SLE may have decreased the frequency. A large autopsy series showed the prevalence of Libman-Sacks endocarditis in SLE patients was 59% before corticosteroids began being used and 35% after their use[5] . However, Bulkley and Roberts stated that corticosteroid therapy may lead to healing of the verrucous lesions, with subsequent scarring and shortening of the posterior mitral valve leaflet and chordae tendinae, increased adherence to the endocardium, and valvular insufficiency[6].

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