Autoimmune Disease with Cardiac Valves Involvement: Libman-Sacks Endocarditis

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ABSTRAK

Laporan kasus ini bertujuan untuk mengevaluasi respons terapi steroid pada endokarditis autoimun. Insidens penyakit jantung katup mengalami peningkatan dengan berbagai macam penyebab mulai dari kongenital sampai yang didapat, tetapi yang disebabkan oleh autoimun sangat jarang. Laporan kasus ini menyajikan kasus wanita, 34 tahun, dengan gejala dan tanda mirip lupus eritematosus sistemik (LES). Pada pemeriksaan penunjang didapatkan data yang mendukung adanya penyebab autoimun dan mengenai organ jantung. Pasien didiagnosis Libman-Sacks Endocarditis dan diberikan terapi steroid. Pasien mengalami perbaikan klinis dan struktural jantung setelah mendapatkan terapi selama 6 bulan.

Kata kunci: *lupus eritematosus sistemik (LES), steroid, Libman-sacks endocarditis.*

ABSTRACT

This case study aim to evaluate the response of steroid treatment for autoimmune endocarditis. Valvular heart disease is relatively rising in both congenital and acquired cases, but the autoimmune endocarditis remains rare. In this case, a 34 year old woman with clinical manifestation resembling systemic lupus erythematosus (SLE) is diagnosed with Libman-sacks Endocarditis. After six months of steroid treatment, her clinical manifestations and heart structure improved.

Keywords: *systemic lupus erythematosus (SLE), Steroid, Libman-sacks endocarditis.*

INTRODUCTION

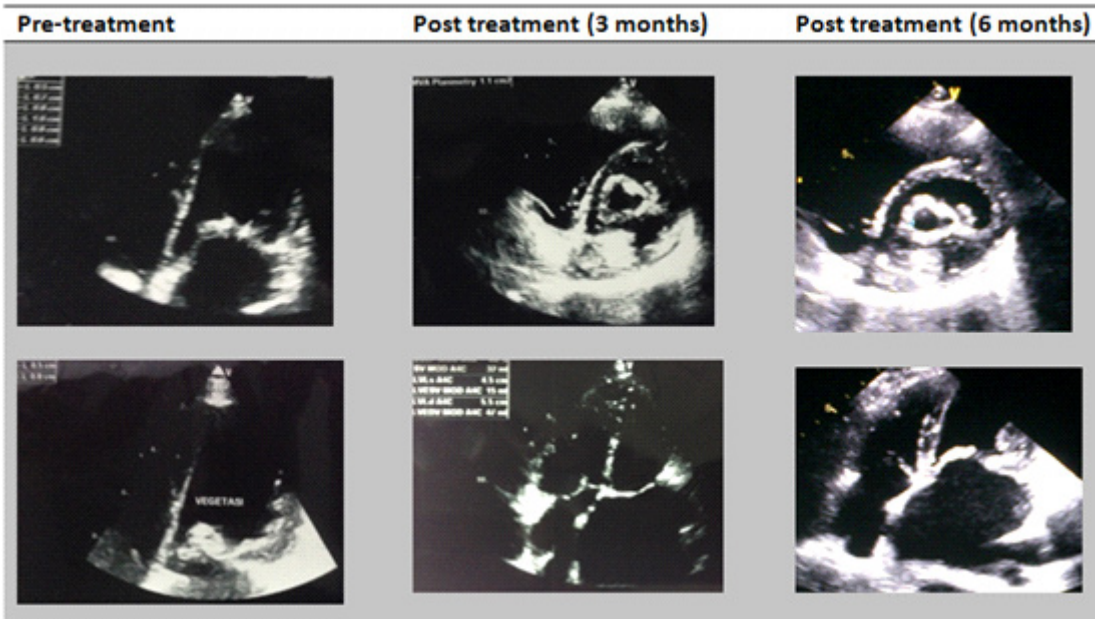
Libman-Sacks endocarditis was first described in 1924 in four patients with atypical sterile verrucae lesions of the valvular and mural endocardium.[1,2] These lesions were pathologically distinct from other etiologic endocarditis, were believed to be characteristic of systemic lupus erythematosus (SLE).[2] This vegetation were found in 35%-65% in early autopsy studies but routinely were clinically silent and of minor hemodynamic importance.[2,3] Echocardiography diagnostic showed frequent occurrence of thickness and functionally impaired cardiac valves in SLE.[1] There are two morphological types of valve lesion represent different stages of the same pathology process.[1,2] The shift in valve pathology has been ascribed to steroid therapy and generally long survival patient with SLE that allows more frequent emergence of fibroses, malfunctioning valve as the end-stage or healed from Libman-Sacks endocarditis.[1,2]

The presence of anti-phospholipid antibodies (aPLs), determined as Immunoglobulin (Ig) G, Ig M or Ig A anti-Phospholipid cardiolipin antibodies (aCLs) and those identified in coagulation test are labelled as Las, was found to be associated with mitral or aortic verrucae valve thickening, global valve thickening and dysfunction, and mitral and aortic regurgitation.[2] Valve involvement has been shown to be influenced both by SLE disease duration and Ig G aCLs.[2] It can reveal that the duration of SLE also affected myocardial function, whereas the aCLs were related only to the damaged endocardium.[2]

The predominantly abnormal function is regurgitation, whereas stenosis is rarely seen.[2] The mitral is mainly affected, followed by the aortic valve.[2] Additional involvement of the tricuspid or pulmonary valve has seldom identified.[2] The valve lesions, as described in early pathological studies are sterile fibro-fibrous vegetation that may develop anywhere on the cardiac surface of the heart but with propensity for the left valves, particularly the ventricular space of the mitral valve.[2]

CASE ILLUSTRATION

A 34-year-old woman presented with new onset palpitation and exertional dyspnoea. She gave a history suggestive of SLE such as fever and shaking chills, cough, and rash spots in the chest and waist but without migratory arthralgia (only back pain), butterfly rash, and chest pain. On physical examination, heart rates (HR) was 80-100 beats/min, respiratory rates (RR) were 20 breaths/min and blood pressure (BP) 150/70 mmHg, a diastolic murmur, grade III-IV, in the apex while all other organs were unremarkable. Laboratory examination, show anti-nuclear antibody/ANA (-), double stranded Deoxyribonucleic acid (ds DNA), complement/ C3 83, Complement (C4). Other results were within normal limits. Electrocardiography (ECG) examination is normal but chest X-ray showed cardiomegaly with minimal pericardial effusion (PE). Echocardiography revealed dilated heart and global normal kinetic motion, severe mitral stenosis (MS), mild mitral regurgitation (MR) and tricuspid regurgitation (TR). Left ventricle



In the above picture of pre-treatment, severe mitral stenosis (MS) was seen and (below picture of pre-treatment) vegetation in the mitral valve (MV) was seen. In the above picture of three months post treatment, moderate MS was observed and (below picture of post-treatment), no more vegetation was seen in the MV. In the above picture of six months post treatment, moderate MS was noted and in the below picture of six months, no vegetation or thrombus was observed in the MV.

Figure 1. Echocardiography results from pre-treatment to six months post treatment.

(LV) and right ventricle (RV) functions were good, there was vegetation on mitral valve (MV), no thrombus, and ejection fraction (EF) was 53%. According to these findings, the patient was diagnosed with autoimmune endocarditis with severe MS, mild MR and TR caused by SLE and anti-phospholipid syndrome (APS), and congestive heart failure (CHF) functional class I-II.

The patient was given methyl prednisolone 1x 8 mg, then tapered down to 1x 4 mg, and oral anticoagulant (warfarin) 1x3 mg for six months. Subsequently, patient felt better. Echocardiography on follow-up, shows a global normal kinesis motion, moderate MS, LV and RV functions were good, vegetation on MV, thrombus (-), and EF is 64%. After six months of treatment, echocardiographic finding improved.

DISCUSSION

Based on diagnostic criteria of SLE, patient presented with malar rash, fever, reduced weight, and cardiac organ involvement.[4-5] In addition, laboratory examination also support this diagnosis with Anti ds-DNA (+2), C3 (82) and C4 (12) but ANA (-).

According to echocardiography, we found mitral and tricuspid valve involvements that was severe mitral valve stenosis, mild mitral and tricuspid regurgitation. Therefore, the diagnosis for this patient is autoimmune disease caused by SLE and APS with severe mitral valve stenosis, mild mitral and tricuspid valve regurgitation.

According to the guidelines, there is no specific therapy to treat a patient with Libman-Sacks endocarditis, but the management focused on heart failure due to the dysfunction of valves according to usual guidelines.[1] Because of the autoimmune aetiology, we gave corticosteroid for this patient. Corticosteroids are not beneficial and may even facilitate valve damage. Immunosuppressive agents should only be used for the treatment of an underlying condition.[1]

Steroids are used to control the autoimmune inflammation. After treatment for six months, the clinical manifestation disappeared and the cardiac valves improved. But patient's bodyweight increased and appearance of her face became puffy with moon face. We suggest that the physician and cardiologist have to deal with the adverse effect of steroid treatment.[2]

Oral anticoagulants were given with caution to the patient in view of static flow that can be a risk factor for thrombosis and also for the vegetation in the MV wall. INR monitoring was done during oral anticoagulant treatment to prevent bleeding. We hope that early diagnosis can improve the patient's outcome.

CONCLUSION

We conclude that this patient with autoimmune endocarditis has good response to six months steroid treatment and re-evaluation of clinical manifestations and cardiac structure should be performed routinely every 3 months by echocardiography.

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