A case of Hughes syndrome with Libman Sack’s Endocarditis


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32 year old housewife, with history of unexplained consecutive spontaneous abortions 7 times in the past was admitted in the medical ward with history of dyspnea on exertion and easy fatigability. Clinical examination revealed pallor, pulse 90/min regular, BP 110/70 mm of Hg and a Grade 3/6 PSM at the apex. Her laboratory tests revealed anemia with hemoglobin 7 gm%, platelet count 91,000, ESR 145 mm/hr, blood urea 46 mg/dl, serum creatinine 2.6 mg/dl and urine routine showed microscopic hematuria. Three sets of blood cultures failed to yield any microorganisms. Autoantibody profile was negative for ANA and anti-ds DNA. The APLA IgM and IgG were strongly positive. Chest X-ray showed mild cardiomegaly with LV apex and ECG revealed sinus rhythm with normal QRS axis. The patient was referred to us for echocardiography to rule out infective endocarditis.

Echocardiography revealed dilated LA and LV, moderate mitral regurgitation and an echogenic mass of size 14 X 9 mm without intrinsic mobility, attached to the tip of anterior mitral leaflet on the LV side (Figure 1A, B & C). The findings were confirmed by trans-esophageal echocardiography (Figure 1D).

Clinical and echocardiographic finding were suggestive of Hughes syndrome (Anti Phospholipid Syndrome- APS) with Libman Sack’s Endocarditis and APS nephropathy. According to the revised Sapporo criteria (Sydney criteria, 2006), definite APS is considered if at least one of the clinical criteria (vascular thrombosis, pregnancy morbidity) and at least one of the laboratory criteria (IgG and/or IgM anticardiolipin antibodies, Antibodies to beta2-glycoprotein or Lupus anticoagulant activity) are satisfied.

Figure 1: Transthoracic echocardiography in (A) Parasternal long axis and (B) Apical 4 chamber views showing the sessile vegetation on anterior mitral leaflet, (C) Colour Doppler echo showing moderate mitral regurgitation and Transesophageal echocardiography (D) showing the mass on anterior mitral leaflet.

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Case Report

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Discussion

Antiphospholipid syndrome (APS) was described in full in the 1980s. The syndrome is also referred to as "Hughes syndrome", after the rheumatologist Graham R.V. Hughes who played a central role in the description of the condition. Hughes syndrome can be primary or secondary. Primary APS occurs in the absence of any other related disease. Secondary APS occurs with other autoimmune diseases, such as systemic lupus erythematosus.

Libman-Sacks endocarditis was first described by Emanuel Libman and Benjamin Sacks in 1924. The association between Libman-Sacks endocarditis and antiphospholipid syndrome was first noted in 1985. Echocardiographic studies reveal valvular abnormalities in 28-74% of patients, with valvular masses in 4-43% of patients with systemic lupus erythematosus, with higher reported rates in transesophageal imaging and in subjects with antiphospholipid antibodies. One cohort study reported that Libman-Sacks endocarditis was found in 11% of patients with lupus. The prevalence of valvular abnormalities detected during echocardiography in patients with primary antiphospholipid syndrome has been reported at 30-32%.

Valvular abnormalities occur as masses, diffuse leaflet thickening, valvular regurgitation, and, infrequently, stenosis. The lesions typically consist of accumulations of immune complexes and mononuclear cells. The vegetations are small and formed from strands of fibrin, neutrophils, lymphocytes, and histiocytes. The left-sided valves are involved most often. The mitral valve is typically affected, and the vegetations occur on the ventricular and atrial surface of the valve. Libman-Sacks lesions rarely produce significant valve dysfunction and the lesions only rarely embolize.

Echocardiographic features of the vegetations in Libman–Sacks endocarditis include irregular borders, heterogeneous echodensity, and an absence of intrinsic mobility (ie, verrucous vegetations) located on the cardiac valves and endocardium. The masses are usually small and sessile, but they can be occasionally large. The basal portion and mid portion of the mitral and aortic valves are involved most commonly. Diffuse leaflet thickening of the mitral and aortic valves or focal leaflet thickening of the mid-basal leaflet may also be observed.

The pathology is the same as non bacterial thrombotic endocarditis except focal necrosis (hematoxylin bodies) can be found only in Libman-Sacks endocarditis. Postmortem studies describe mulberry like clusters of verrucae on the ventricular surface of the posterior mitral leaflet, often with adherence of the mitral leaflet and chordae to the mural endocardium. The histologic stages of Libman-Sacks endocarditis have been described as active or healed lesions, characterized by clumps of fibrin on the valvular leaflet tissue, which is focally necrotic, with plasma cells and lymphocytes in the active verrucae or dense, vascularized, fibrous tissue in healed lesions.

The patient responded well to treatment with aspirin, packed RBC transfusions, broad spectrum antibiotics and supportive care and was discharged with the advice for regular out-patient follow up.

Reference