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Case Report

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Libman-Sacks Endocarditis and Chordae Tendineae Rupture as Sudden Presentation of Systemic Lupus Erythematous

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Abstract

22-year-old Puerto Rican male was diagnosed with Systemic Lupus Erythematous (SLE) after presenting with gradual onset shortness of breath during the past 24 hours. Transthoracic Echocardiogram revealed moderate-severe mitral regurgitation with anterior leaflet prolapse at A1 level with suspected rupture chordae, which later was confirmed by Transesophageal Echocardiogram (TEE). Laboratory results came back positive for Antinuclear Antibody, Anti-Double stranded DNA and Neuronal Nuclear Antibody, for which Systemic Lupus Erythematous was diagnosed. Broad spectrum antibiotics were provided due to the possibility of Libman-Sacks Endocarditis debuting as SLE also was started on intravenous steroids. Patient underwent Mitral Valve Replacement instead of repair given high risk of native valve recurrent damage or complication due to his newly diagnosed connective tissue disease.

Background

Libman-Sacks endocarditis is a form of non-bacterial endocarditis (NBE) that causes damage to heart valves in the setting of systemic lupus erythematous (SLE). Surgical valve replacement is necessary in patients with large vegetations, valvular insufficiency and recurrent thromboembolic events. We present the case of a 22-year-old male without significant medical history who presented with chordae tendineae rupture as initial presentation of SLE.

Case Presentation

We present a case of a 22 y/o Puerto Rican male without significant past medical history, who had been complaining of sporadic generalized joint pain for the last year who presented to the Emergency Department due to progressive shortness of breath during the last 24 hours associated with left side back pain, fever, chills and bilateral leg edema. Denied chest pain, palpitations, syncope, rash, cough, recent travel or ill contacts. Physical examination showed an average built man, in mild respiratory distress, breathing 24 per minute and with peripheral saturation of 94% at ambient air. He had a grade II-III/V holosystolic murmur that radiated to axilla. Laboratories with leukocytosis of 15.0 x10^9; hemoglobin 11.0g/dl and platelets at 158,000 per mcL. Chemistries were unremarkable. Chest XR revealed bilateral perihilar interstitial opacities with small left pleural effusion. Electrocardiogram showed normal sinus rhythm without ST-T segment changes. Patient was treated with antibiotherapy (Azythromycin 500mg/day and Ceftriaxone 2G/day) due to suspected Community Acquire Pneumonia.

A day after admission he developed chest pain, hypotension and respiratory distress requiring or tracheal intubation and inotropic medication. Laboratories were remarkable for elevated Troponin (0.391 ng/ml). Transthoracic Echocardiogram revealed moderate-severe mitral regurgitation with preserved ejection fraction at >60%. After hemodynamically stabilization, patient underwent Transesophageal Echocardiogram (TEE) which confirmed aforementioned findings with anterior leaflet prolapse at A1 level with suspected rupture chordae regurgitation volume of 47ml. Reference laboratory results came back positive for Antinuclear Antibody (1:640 homogeneous), Anti-Double stranded DNA and Neuronal Nuclear Antibody. Sedimentation rate elevated at 77ml/h. Workup for endocarditis showed no signs of an infective process, as there was no associated fever, constitutional symptoms and physical examination findings and blood cultures were negative. Systemic Lupus Erythematous (SLE) was diagnosed.

Treatment

Initially treated with for CAP and later optimized to broad spectrum antibiotics due to the possibility of Endocarditis debuting as SLE, also was started on pulse steroid therapy due to valve failure as presenting symptom of SLE. Patient underwent surgery and found with A1-A2 chordae of anterior leaflet ruptured, thinned out remaining anterior leaflet and posterior leaflet, dilated left ventricle, nodular inflammation of rest mitral apparatus. Mitral Valve replaced with mechanical one instead of repair given high risk of native valve recurrent damage or complication due to his newly diagnosed connective tissue disease.

Outcome and follow up

Histopathological examination of the leaflets revealed features that are characteristic of NTE, including abundant rich in fibrin

and platelet aggregates but devoid of inflammation or infective organisms. The patient is doing well 6 months after procedure and has had no complications.

Discussion

Valvular involvement is common in SLE, occurring in up to 59% of patients evaluated by Transesophageal echocardiography. Immunoglobulin and complement deposition in the valvular structure will subsequently lead to Libman-Sacks vegetations, valve thickening, and valve regurgitation. Valvular stenosis is rarely seen. The vegetations of Libman-Sacks can affect all four valves and usually accumulate at the valve edge or on both surfaces. These lesions consist of immune complexes, monocytes, fibrin and platelet aggregates and can have varying degrees of fibrosis, granulation tissue, calcification and necrosis. These lesions are typically friable and can fragment and embolize, particularly when they are large. Involvement of the mitral valve is most frequently encountered. Valve disease for most patients is mild and asymptomatic, but patients in whom severe mitral regurgitation develops will present with symptoms of congestive heart failure.

Unlike infective endocarditis where the valve needs to be excised to remove all infected tissue, repair and preservation of the valve is possible in selected patients with NBE and may eliminate the need for anticoagulation, but that is not the case when repair of the affected valve can lead to recurrence. Case reports suggest that native valve repair does not alter the progression of valve thickening and calcification, and that replacement is ultimately necessary. Also, it has been reported that while corticosteroid therapy improves overall survival of patients with SLE, their use may cause shrinking and scarring of heart valves. It has also been noted that porcine bio prosthetic valves can be affected by valvulitis with perforation in the cusps in patients with SLE requiring replacement [1-4].

In our patient, mechanical valve replacement was done due to findings of thin valve and the high probability of recurrence if repaired.

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