Libman-Sacks Endocarditis “A Wolf in Sheep’s Skin”

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Libman-Sacks Endocarditis “A Wolf in Sheep’s Skin”

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Introduction

Libman-Sacks endocarditis (LSE) is a form of non-bacterial endocarditis seen in patients with systemic lupus erythematosus (SLE). It was first described by Emanual Libman and Benjamin Sacks at Mount Sinai Hospital in New York City in 1924. Libman-Sacks valvular lesions are sterile fibrinousvegetations that favor the left-sided heart valves and usually form on the ventricular surface of the mitral valve. The pathogenesis is thought to involve the formation of fibrin-platelet thrombi that organize and leads to Libsnaks and scarring with subsequent valve dysfunction. An association has been made between LSE and antiphospholipid antibodies (APAs), showing an increased risk of thrombotic events, such as stroke. Here we report an atomic case where the patient was found for the first time positive for lupus anticoagulant and antiphospholipid syndrome (APS) that likely presented with an acute peripheral arterial thromboembolic event accompanied by clinical signs and symptoms of congestive heart failure.

Case Report

History of Present Illness: A 28-year-old African American female with past medical history of hypertension, iron deficiency anemia, tobacco abuse and depression presented to the emergency department with shortness of breath at rest, and worsening over the past 3 weeks. Over this time period, she noticed swelling in both of her lower extremities as well as darkening of her fingertips, which had increased in severity within the same time span. She denied fevers, chills, nausea, vomiting or diarrhea. There was no history of cough, abdominal pain, bloody bowel movements or melena. She did, however, admit to occasional retrosternal chest discomfort, described as a dull ache, rated 3 out of 10, without any radiation to any particular site.

Hospital Course: Patient was admitted to the medical/surgical floor. She had an initial hemoglobin of 6.4, WBC of 8.0 and platelets 162,000. On physical exam, she was afebrile, heart rate was 57 per minute and systolic blood pressure was 155/100 mmHg and oxygen saturation of 100% on room air. Patient had mildly elevated JVD as well as a loud S2. There was a 4 out of 6 systolic murmur heard at the left sternal border, which increased with inspiration and a 3 out of 6 systolic murmur at the mitral area, radiating to her axilla. There was trace bilateral pitting edema of lower extremities, halfway up her tibial surface with normal pedal pulses. There were ischemic changes noted on her right fifth extremity, with normal pedal pulses. There were ischemic changes noted on her right fifth extremity, with normal pedal pulses. There were ischemic changes noted on her right fifth extremity, with normal pedal pulses.

Discussion

This case report describes a patient with SLE (established according to the criteria of the American College of Rheumatology), 1 who presented with signs and symptoms of congestive heart failure as well as multiple areas of suspected ischemia possibly secondary to either LSE or APS. In fact, the patient had saccular vegetations on the mitral valve with associated positive lupus anticoagulant antibodies characterizing a diagnosis of APS.

More than half of the patients with SLE, when assessed with transesophageal echocardiogram have clinically silent valvular alterations. One complication of SLE seen in approximately 9% of patients is a thromboembolic phenomenon with the brain being most affected. In most cases, embolic episodes are subclinical, but sometimes may manifest as signs and symptoms of ischemia. Approximately 33% of SLE patients are found to have positive APA antibodies. An increased risk of thromboembolic events is observed in LSE, which might be in part due to the presence of the APAs. 1,2 Patients with moderate to high titers of antiphospholipid IgG antibodies have a higher incidence of valve lesions, whereas there are patients with valve disease in whom lupus anticoagulant are the only APAs detected. 1,3

It should be considered that infectious endocarditis is not uncommonly seen in SLE patients with LSE as patients already have valvular disease. Thus a broad differential diagnosis is mandatory. In this aspect, three laboratory data are important: leukocytoc count, CRP levels, and APA levels. Leukocytoc tend to decrease during lupus activity and the opposite occurs in infectious endocarditis. As for the APAs, it is usually helpful for the clinician to reevaluate the presence of the APAs while considering the presence of SLE. Very high CRP levels usually indicate an infectious cause, as lupus patients are less capable of presenting a large response of this protein; however, for a definitive differential diagnosis, blood cultures are imperative. 1 In the present case, a diagnosis of LSE was attained, as the leukocytoc count was normal, the CRP was not very elevated, APA was positive and 4 pairs of blood cultures had no growth. There is scarce information in the literature regarding treatment of LSE. It is known that the use of corticosteroids and immunosuppressive drugs seem to have no effect on valve lesions; however, anticoagulation therapy must be used for treatment of patients with thromboembolic events. 1,2

In the present case, the patient initially received Lovenox and ultimately Warfarin therapy to maintain an INR of 2.5-3.5. It is also should be considered that prospective, controlled, and randomized studies have shown that moderate (INR 2.0-3.0), as well as a more intensive (INR 3.0-4.0) anticoagulation with warfarin was similarly effective in preventing new thromboembolic events in patients with APS after the first thromboembolic event. 4,5

Hydroxychloroquine and prednisone (60 mg/day) were also added to their treatment regime as she had signs of disease activity (pericardial effusion, increased ESR and decreased CRP). The potential usefulness of hydroxychloroquine in preventing events related to the APS remains in the case of SLE patients, particularly those with APAs, as there is evidence of fewer thrombotic events with its use. 6,7

References:


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