Libman–Sacks endocarditis with an unusual presentation

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Abstract
Systemic lupus erythematosus is relatively common medical disorder with female predominance. This disorder can affect any organ system. Cardiac involvement is variable which can include pericardium, myocardium and endocardium. The endocardial involvement commonly affects mitral and aortic valves. This report discusses lupus endocarditis in young man with atypical presentation.

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1. Introduction
Systemic lupus erythematosus (SLE) is a multisystem autoimmune disorder in which the inflammatory process involves every organ system. Females are more commonly affected than males. Cardiac involvement is common in this disorder and pericarditis is the most common cardiovascular manifestation.

This report describes a young man who was admitted with fever and pancytopenia and signs of endocarditis which had been confirmed by transeosophageal echocardiography and clinical improvement of the patient condition after starting corticosteroid therapy.

A 32 year, non-smoker male was admitted with three weeks history of persistent fever, night sweat, rigors, headache, anorexia and general fatigue, in-spite of empiric course of ciprofloxacin therapy. Medical history included large left thyroid lobe with normal thyroid function since 10 years. Two days prior to admission, he had dry cough. On admission, he appeared ill but fully oriented, febrile (38°C), blood pressure 120/80, pulse rate 110 BPM, with no jaundice or pallor. There was alopecia (Fig. 1a), submental swelling, mouth ulcer at soft palate and crusted lesions at lower lip, maculopapular and petechial rash over trunk, vasculitis lesions at finger pulps and splinter hemorrhages (Fig. 1b). Patient developed left sided pleuritic chest pain but cardiac exam was normal; there were scattered ronchi on lung auscultation with few bi-basal rales. Fundal examination showed large soft exudates inferolateral to the optic disc on left side (which eventually increased and became bilateral). Overtime the patient lost around 4 kg of weight.

Laboratory investigations showed reduction in platelet count (79000/l) and white cell count (1900/l). Neutrophils (75%), lymphocytes (20%), hemoglobin (11.2 g/dl), MCV (84 fl), MCH (28 pg) and reticulocyte count (1.1%) were in normal range. Urine analysis showed ++RBCs but no casts. While the coagulation profile was normal; Lactate dehydrogenase (589 u/l) was considerably high. Malaria smears were negative. Chest radiograph and other system examinations were normal.

Differential diagnosis for fever and pancytopenia includes infections (salmonella, brucella, tuberculosis, HIV, hepatitis, and endocarditis), connective tissue disease like Systemic Lupus Erythematosus (SLE), and hematological disorders (leukemia, myelodysplasia, myeloproliferative disorders), and febrile conditions (infections, autoimmune, neoplastic). The patient was started on high dose of corticosteroid therapy due to his condition.
Lupus Erythematosus (SLE), neoplasms like lymphoma and leukemia), and miscellaneous such as Behcet’s disease and Inflammatory bowel disease. Transesophageal Echocardiography (TEE) revealed aortic valve (Fig. 2) with three small vegetation projections (two at coronary cusps and the third at the non-coronary cusp) of the aortic valve (arrows).

Treatment was initiated with weight adjusted steroid therapy with significant response. The patient was afebrile and showed marked improvement in health when he returned home after a month; and was on oral steroid therapy to be tapered on out-patient follow-up with discharge white cell count 4500/µl, Hb 10 g/dl and Platelets 109,000/µl. Need for long term aspirin for prevention of atherothrombotic events was discussed with the patient.

2. Discussion

A survey in Birmingham women inferred that substantial number of undiagnosed cases of SLE exists due to its wide range of symptoms, hence further work needs to be done to focus on its diagnosis and treatment (Johnson et al., 1996). Libman–Sacks endocarditis is characterized by sterile verrucous valvular lesion with a predisposition for mitral and aortic valve, a cardiac manifestation of SLE as well as of antiphospholipid syndrome. Its prevalence is usually underestimated because of the mild clinical presentation (Lee et al., 2009). Our case presents an unusual aortic valve involvement in a male patient. Aortic involvement was detected with the help of TEE which is a very useful diagnostic tool for diagnosis of Libman–Sacks endocarditis with a unique characteristic features (Roldan et al., 2008). The patient was treated with corticosteroids, despite numerous side-effects and controversies associated with its use with marked clinical improvement. Corticosteroids prevent SLE relapse (Bootsma et al., 1995) and patient tends to live longer (Menard, 2008).

References