Case Report

A Young Lady Presenting with Fever, Polyarthralgia and Breathlessness - A Case Report

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Abstract:

Heart valve abnormalities are the most frequent cardiac manifestations in patients with antiphospholipid syndrome (APS) with or without systemic lupus erythrematosus (SLE) though clinically significant valvulopathy occurs rarely. Here, we present a case of a 20-year-old young lady presenting with intermittent fever and polyarthralgia for 2 weeks, breathlessness for 1 week and bilateral leg swelling for 3 days. On examination, moderate anaemia, high blood pressure, generalized lymphadenopathy and evidence of mitral regurgitation was found. She had Hb% 8.5 gm/dl with high ESR. In Doppler echocardiography, there was MVP with mitral regurgitation (MR) grade III. Transeosophageal echocardiography revealed features of type A Libman-Sacks (LS) endocarditis along with MVP and MR. Unfortunately; she had strongly positive ANA, positive anti ds-DNA, positive direct coombs' test & positive antiphospholipid antibody (aPL) IgG with low C3 level & proteinuria. On the basis of clinical presentation and laboratory reports, a diagnosis of SLE with secondary antiphospholipid syndrome (APS) with LS endocarditis with secondary MVP with coomb's positive haemolytic anaemia was made. For treatment purpose, she was transferred to the department of Rheumatology. After 1 month during her follow-up visit, she was improved clinically & echocardiogracally.

Keywords: Systemic Lupus Erythrematosus, Antiphospholipid Syndrome, Libman-Sacks Endocarditis, Mitral Valve Prolapse, Mitral Regurgitation

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Introduction:

Valvular heart disease is the most important cardiac manifestation of systemic lupus erythematosus (SLE). The presence of antiphospholipid (aPL) antibody is significantly associated with an increased risk of valvular lesions as aPL are associated with hypercoaguable state and thrombosis is the possible mechanism of valvular lesion. Libman-Sacks (LS) endocarditis is frequent in antiphospholipid syndrome (APS), especially with SLE. The importance of identifying such lesions is linked not only to assess valvular dysfunction but also to reduce the risk of arterial thromboembolism. Here, we described a case of a young lady presented with intermittent fever, polyarthralgia, breathlessness and leg swelling and examination and investigation findings suggestive of SLE with secondary APS with LS endocarditis with secondary mitral valve prolapse with coomb's positive haemolytic anaemia.

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Case Report:

A 20-year-old lady presented with fever, multiple joint pain for 2 weeks, exertional breathlessness for 1 week and leg swelling for 3 days. Fever was intermittent in nature, not associated with rigor, maximum temperature 104^0 F, and subsided after taking anti-pyretic drug with profuse sweating. One day later, pain started in different small joints of both hands initially and afterwards in large joints, mainly knee and ankle joints, was of dull aching in nature & constant, aggravated during movement and had no specific relieving factors. On query, there was no stiffness, redness of overlying skin, deformity, history of tuberculosis or syphilis. Last one week, she felt short of breath, which

was progressive and more on exertion. She had no history of cough, wheeze, pleuritic chest pain, angina, palpitation, childhood wheeze or atopic allergy, any symptoms of psychogenic hyperventilation, morning headache, and light headedness. 4 days before admission, she noticed bilateral leg swelling progressing day by day without any history of decreased frequency or volume of urination, facial swelling on waking and abdominal swelling. She had no previous history of psychosis, seizure, malar rash, abortion. She used oral contraceptive pill for birth control. On examination, she was moderately anaemic, having high blood pressure (BP- 150/100 mm Hg), ++ bilateral leg oedema. There was tender generalized lymphadenopathy, largest one around 2.5 x 2.5 cm^2 in posterior cervical region. Most of them were firm with few soft in consistency and not matted, not fixed with underlying structure or overlying skin and no discharging sinus. She had features of mitral valve prolapse – apex beat in normal position, loud 1^{st} heart sound and an ejection systolic murmur in mitral area. Examination of all joints revealed mild tenderness with restricted movement associated with slightly impaired functional activity. There was pansystolic murmur at the apex. Initial investigation report revealed Hb% 8.5 gm/dl with high ESR (117 mm in 1^{st} hour), 6,000/mm^3 white cell with normal percentage of neutrophil and lymphocyte, 2,00,000/mm^3 platelet counts. Peripheral blood film showed hypochromic microcytic anaemia. Serum creatinine, serum electrolytes, random blood sugar was normal. In urine microscopic examination, ++ protein, 2-4 pus cell/ HPF and occasional RBC were found. CRP was 16.2 mg/dl. Blood culture and urine culture revealed no growth. The report of febrile antigens was insignificant. Colour Doppler echocardiography revealed mitral valve prolapsed with MR, grade – III (Figure 01).

Up to this stage, we couldn't able to reach a specific diagnosis. So, we go further. Later, Transoesophageal echocardiography revealed mildly thickened and calcified MV leaflet tips with mild subvalvular apparatus change suggestive of Libman-Sacks endocarditis and mitral valve prolapse with MR grade III (Figure 2). At the same time, strongly positive ANA with homogenous pattern in indirect immunofluorescent test and positive anti ds- DNA (134.5 U/ml) came to our hand. There was low C3 (0.226 gm/l), normal C4 level, positive antiphospholipid ab IgG (25.0 u/ml), positive direct coomb's test and 1540 mg/dl urinary total protein. From above clinical presentation and laboratory reports, it was apparent that she has been suffering from SLE with secondary antiphospholipid syndrome with Libman-Sacks endocarditis with secondary mitral valve prolapse with coomb's positive haemolytic anaemia. For proper treatment, we consulted and transferred her to the department of rheumatology. After 1 month during follow-up, she was improved symptomatically & echocardiographically.

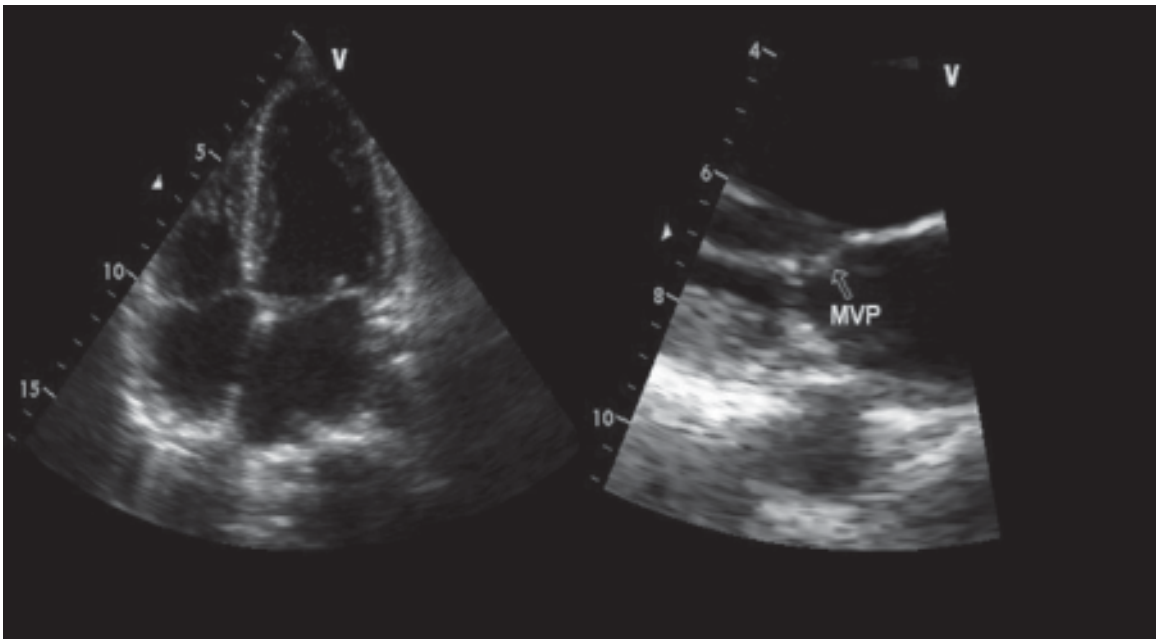

Fig.-1: *Transthoracic echocardiography revealed mitral valve prolapse. MVP, Mitral Valve Prolapse*

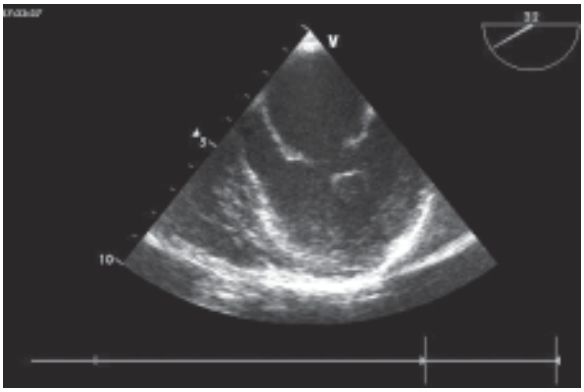

Fig.-2: *Transoesophageal echocardiography revealed Libman- Sacks Endocarditis with mitral regurgitation.*

Discussion:

Fever, polyarthralgia and systolic murmur in mitral area brought out to our mind at least three differential diagnosis- acute rheumatic fever, infective endocarditis, antiphospholipid antibody with or without SLE. Lyme disease is also a rare possibility.

In Bangladesh, a valvular lesion in APS with or without SLE is still unknown. But in developed country, it is frequent in patients with SLE and APS, but clinically significant valvulopathy occur in only a small percentage of patients. Libman-Sacks endocarditis is the end-stage of a progressive process and in typically asymptomatic. In SLE, it was first mentioned in 1924 by Emanuel libman and Benjamin Sacks [1]. After a long time, in 1985 the link between aPL and libman-Sacks endocarditis was first reported by D' Alton el al. [2]. The pathogenesis of this is still unclear; even there are no pathognomic microscopic features. However, there are some characteristic findings suggest the diagnosis in this case, first described by Ménard et al in 2008- low leucocytes count, mildly raised C-reactive protein (CRP) and positive aPL and characteristic echocardiographic findings mentioned in Table -I [3, 4, 5]. Very high level of CRP suggests infective endocarditis.

In echocardiographically, Libman-Sacks vegetations appear as valve masses of varying size and shape, generally more than 2 mm in diameter, irregular border, firmly attached to the valve surface, sessile and exhibit no independent motion [4]. Left sided heart valves are more affected. When mitral valve leaflet is involved, vegetation may extend to the subvalvular apparatus and the adjacent mural endocardium [5]. The end-stage of libman-sacks endocarditis is a fibrous plaque with focal calcification [6]. In this case, mitral valvular apparatus is thickened, mildly calcified tip with minimal elevation with mild changes in subvalvular apparatus. When Libman-Sack endocarditis is found in an early stage, corticosteroid is recommended as it facilitates gradual healing of the lesions by decreasing inflammation [7]. Although, it also promote fibrosis and scarring, resulting in additional valvular damage [5].

Roldan et al. published that valvular thickening was the predominant finding followed by vegetations, valvular regurgitation and stenosis [8]. In this case, along with vegetation, mitral regurgitation was found. Perez-villa et al. stated that mitral valve involvement is more frequent and found in 26% case, aortic regurgitation in 7% and tricuspid regurgitation in 7% cases [9].

Conclusions:

Cardiovascular involvement in SLE and APS is the leading cause of death and it is now essential for clinicians to early recognize Libman-Sack endocarditis and its complication. Not only early recognize, adequate control of the disease process is essential to prevent other sequelae.

Table-I

Helpful marker in distinguishing infective endocarditis from libman-sacks endocarditis

Laboratory parameter	Infective endocarditis	Libman-Sacks endocarditis
WBC	increase	often decrease in SLE flare
Serum CRP	increase	increase (though possibly decrease)
aPL level	can be normal	moderate to high titres
Echocardiography	vegetation located nearer to the leaflet line of the closure, having independent motion and exhibit homogenous echodensity	vegetation located not nearer to leaflet line of closure like infective endocarditis, having no independent motion and exhibit no homogenous echodensity.

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Conflict of Interest:
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