Case Report

A Lady with Systemic Lupus Erythematosus and Mitral Stenosis

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Abstract:

Mitral stenosisis a valvular heart disease caused by a number of diseases. Chronic rheumatic fever is the most important cause. Among rare causes, some rheumatoid diseases like SLE may involve cardiovascular system causing libman- sacks endocarditis,pericardial diseases and other valvular lesions mostly associated with positive antiphospholipid and anticardiolipin antibody.Here, we presented a case of rheumatic mitral valvular heart disease having systemic lupus erythromatosus but negative antiphospholipid and anticardiolipin antibody.

Keywords: Rheumatic fever, Systemic lupus erythematosis, Mitral stesnosis

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Introduction:

Rheumatic fever(ARF) is the most important cause of valvular heart diseases,[1]but there are some rheumatoid diseases where heart valves are also involved.[2-7]Systemic lupus erythematosus (SLE) is one of the chronic systemic autoimmune disease, associated with valvular heart diseases, Libman-Sacks lesions, serositis, pericardial disease, venous and arterial thrombosis. All these manifestations are mostly associated with antiphospholipid antibodies.[3]In a study conducted on echocardiography of SLE patients, variable valvular diseases such as mitral stenosis,mitral valve thickening or vegetation, mitral valve prolapsed, mitral, aortic, and tricuspid regurgitation; were reported.[8]Here we reported a known case of rheumatic MS with incidental findings of SLE with negative antiphospholipid and anticardiolipin antibody.

History: A young lady of 34 years, housewife, normotensive, nondiabetic, admitted into CCU of Mitford Hospital on 9th November 2015 with history of shortness of breath for 6 years, palpitation for 2 years, multiple joint pain& oral ulcer, skin rashes on face, arm &legs for 2 months(Fig:1). She gave history of marked weight loss but there was no history of fever, spontaeneous abortion, bleeding,venous thrombosis or convulsion. She had history suggestive of rheumatic fever 22 years back and repeated hospital admission without any improvement.

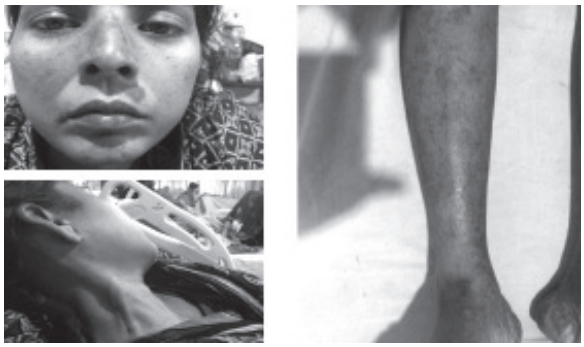

Fig-1: *Characteristic rashes of SLE.*

Clinical examination revealed tachycardia, dysponoea, anaemia, oedema, skin rashes over face, legs, arms, raised JVP, apex beat in left 5th intercoastal space tapping in nature, palpable P_2, epigastric pulsation with left parasternal lift were present.There was loud first heart sound in mitral area with mid diastolic murmur and apansystolic murmur over left lower sternal edge. Liver was palpable and shifting dullness was present. Her both knees, ankles and small joints of both hands were swollen and tender.Investigations showed: Hb%-9.8gm/dl,anti-

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nuclear Ab(ANA): positive, anti-ds DNA: positive, APL Ab: negetive, anti-cardiolipin-Ab: negetive, CCr-30.21ml/min, ECG :sinus Tachycardia (Fig:2), pericardial effusion on X-Ray chest P/A view(Fig:3). Echocardiography revealed severe mitral stenosis (MVA-0.59cm2) with thicking and calcification of both AML and PML with diastolic doming of AMLwith fused both Commisures, moderate subvalvualar changes,moderate pericardial effusion (23mm) with moderate pulmonary hypertension(PASP-51mmHg) without any thrombus (Fig:4).

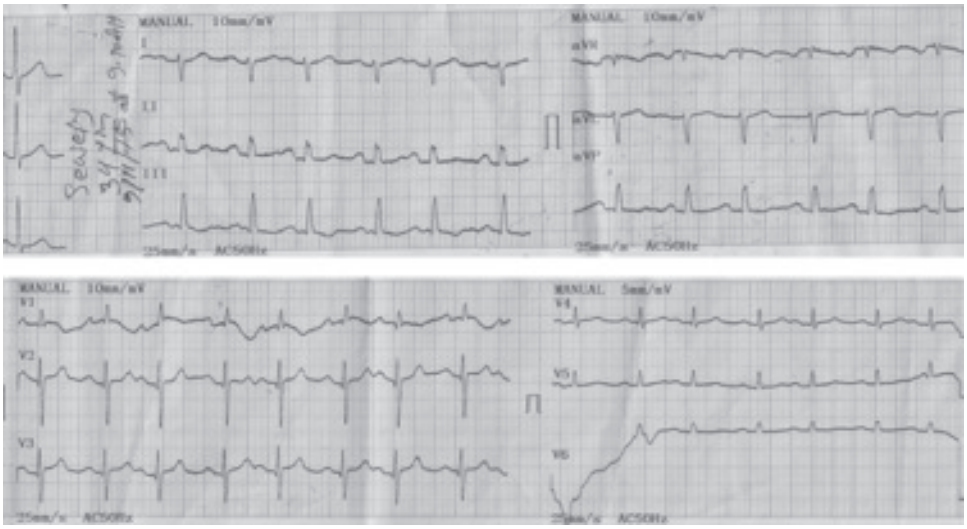

Fig.-2: *ECG: Sinus tachycardia*

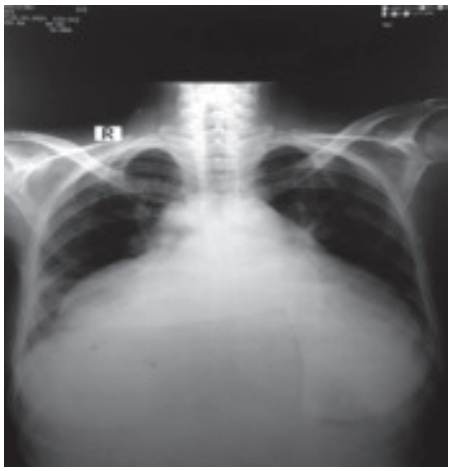

Fig-3: *X-Ray chest P/A view: Features of pericardial effusion.*

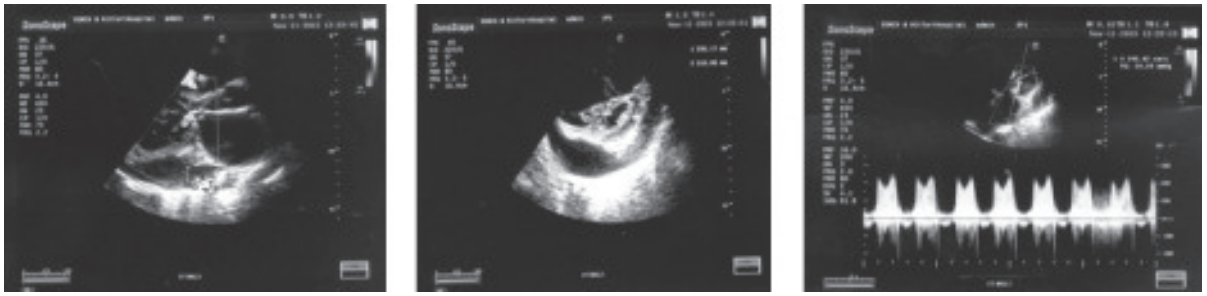

Fig-4: *Echocardiography (2D,M-Mode and Doppler): Severe MS(MVA-0.59cm^2), subvalvular changes and pericardial effusion .*

After diagnosis of SLE,hydroxychloroquine was added, subsequently patient developed visual impairment, fundoscopic examination found macular pigmentations.

Discussion:

Our patient was a known case of rheumatic valvular heart disease for last 22 years,as there was maculopapular skin rashes,investigations regarding SLE was done. SLE is an autoantibody and immune complex disorder, with immunoglobulin and complement deposition in involved organs, including the heart. The serologic findings may be detectable years before clinical disease manifests.[9]In this patient valvular manifestations are mostly due to rheumatic fever because echocardiography showed mitral valve was severely stenosed (MVA-0.6cm2),both AML and PML were thickened and cacified with diastolic doming of AML, both commisures were fused, moderate subvalvualar changes without any thrombus,all these findings are in favour of chronic rheumatic heart disease,whereas in SLE echocradiographic findings include diffuse thickening of valve leaflets with minimal subvalvular changes.

Transthoracic and transesophageal echocardiography had shown that valvular involvement is greater in SLE patients who have positive antiphospholipid antibodies.[10]A number of other manifestations may occur in SLE,most of which are associated with antiphospholipid and anticardiolipin antibodies, such as venous and arterial thrombosis,[11] recurrent fetal loss,[12] pulmonary hypertension,[13]endocardial disease seizures, and migraine.[14] But in our patient antiphospholipid and anticardiolipin antibody were negative.

Studies of patients with SLE have reported a 25% incidence of clinically evident pericarditis, a 50% incidence of pericardial effusion (detected by echocardiography), and 80% incidence of pericardial abnormalities at autopsy.[15,16]Here, in our patient,we also found moderate pericardial effusion(23mm) may be due pericarditis .

Over 95% of patients with SLE have a positive ANA; however, even high titers of ANA are not diagnostic of SLE. Anti–double-stranded DNA is more specific for SLE but is present in 50% to 70% of patients with idiopathic SLE, often in those with glomerulonephritis. Our case had strongly positive ANA and anti ds-DNA with impaired creatinine clearance(30.21ml/min).

There are reports of mitral and aortic valve replacement in patients with SLE.[17]Valve repair has also been described.[18] Our further planning for management of this patient is mitral valve repair &/or replacement.

We also found moderate pulmonary hypertension in this case, which is common in SLE[19]but sig-nificant pulmonary hypertension is less common.

According to ACR/SLICC revised criteria for diagnosis of SLE,our patient had 9 points out of 16(malar rash-2 points, oral ulcer-1 point,pericarditis-1 point,anemia-1 point, high titre ANA-2 points,positive anti ds-DNA-2 points[20] and diagnosed a case of definite SLE.

The scope of investigations like renal biopsy,SPECT or endomyocardial biopsy were limited in this case, to find out further involvement of myocardium and kidney.

Our patient developed macular pigmentation on starting of hydroxychloroquine that need further evalution.

Conclusion: Anuncommon case of SLE without positive antiphospholipid and anticardiolipin antibody along with rheumatic origin of mitral stenosis is presented here. It is recommendeted that two different entity of mitral valvular disease can co-exist.

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