Case Report

# Tricuspid Valve Libman Sack Endocarditis with Pericardial Effusion in an Antiphospholipid Syndrome Negative, Systemic Lupus Erythematosus Patient : An Interesting Case Report

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**Non bacterial thromboembolism (NBTE) and pericardial effusion represent some of the unusual manifestation of systemic lupus erythematosus. Right sided endocarditis in SLE is uncommon and can lead to pulmonary embolism. Pericardial effusion in SLE patient can be due to disease activity as well as infection like tuberculosis. Routine screening can reduce morbidity from these conditions. NBTE can develop in anti Phospholipid syndrome serologically negative patients.**

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***Key words*** **: Libman-Sacks endocarditis, SLE, Antiphospholipid syndrome, tricuspid valve, pericardial effusion.**

Libman-Sacks endocarditis is a non infective verrucous thrombotic endocarditis and cardiac manifestation of systemic lupus erythematosus (SLE) and antiphospholipid syndrome (APS)[1]. Libman-Sacks lesions are associated with lupus duration, disease activity, anticardiolipin antibodies, and antiphospholipid syndrome manifestations[2].

## Case Report

Our patient is a 28 year old, non diabetic, non hypertensive, non hypothyroid female patient who presented to us with fever, joint pain and pedal swelling for five months duration. The joint pain was inflammatory in nature involving large and small joints. The fever was high grade in nature with no typical localizing symptoms. The patient had a gradually progressive shortness of breath which was predominantly exertional but there were two bouts of shortness of breath for which she required admission to local hospital prior to presenting to us. She also complained of mild vague chest pain in the left axillary region on deep expiration or occasional coughing for the last 1 month. But there is no history of cough or expectoration. Patient had normal urine output. She had no significant weakness associated with joint pain. On asking, she gave history of 3 abortions, all were spontaneous and in first trimester. She has two living children currently. Her menstrual and obstetric history is otherwise non significant. After admission on examination she was found to have moderate pallor and pedal swelling. Her JVP was not engorged. She had tachypnea and tachycardia; Respiratory rate was 28/min,pulse rate was 110/min. She had multiple firm, non matted palpable cervical lymph nodes of size around 2.5cm which were non tender.Her cardiovascular and chest examination revealed muffled heart sounds and bilateral vesicular breath sound respectively. She had palpable hepatomegaly and traube's space percussion was dull. Immediate echocardiography was done which revealed a pericardial effusion measuring 20mm anteriorly with normal ejection fraction. Pericardiocentesis was done and the fluid was sent for investigation. Her routine blood report showed a normocytic normochrmic anemia with normal WBC count and deranged urea creatinine. Her serum LDH was raised and serum potassium was on the higher side of normal. A blood for Direct Coomb's test was sent which came back positive. Serum Anti nuclear antibody was positive with low complement and high dsDNAlevels.Her APS profile including anti Cardiolipin, anti beta2 GPI and Lupus anticoagulant were negative. Urine routine examination showed active sediments with hematuria and proteinuria which was quantified and came back to 1.46gm/24 hours. Her kidney size was normal. During hospital stay, her blood creatinine values normalized gradually without any intervention. Kidney biopsy was done which revealed Class III C proliferative lupus nephritis. Her APS profile was negative (Repeated). Pericardial effusion study had increase fluid protein 6.1gm and raised ADA level 48.8U/L but fluid CBNAAT was

***Editor's Comment :***

- In SLE, dyspnoea may be due to valvular involvement, pericardial effusion or pulmonary involvement.
- In diffuse alveolar disease of SLE, alveolar hemorrhage must be ruled out as it is rapidly life-threatening.
- The possibility of seronegative APLA must be thought of in SLE with suitable clinical features like recurrent foetal loss.

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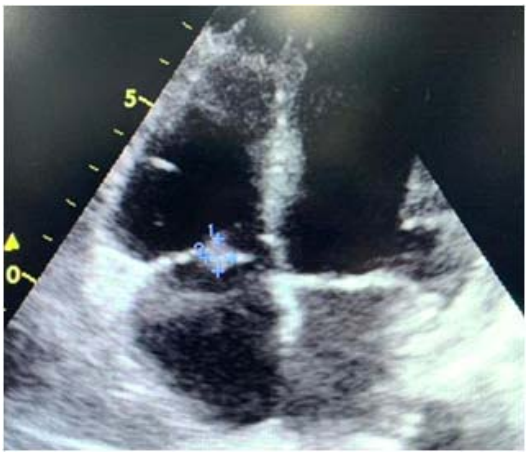

Fig 1 — Figure showing the echocardiographic imaging of the vegetation on tricuspid valve

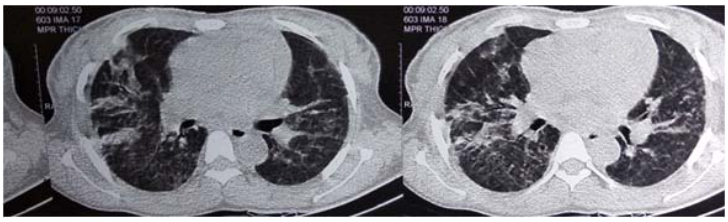

Fig 2 — HRCT thorax showing interstitial lung disease

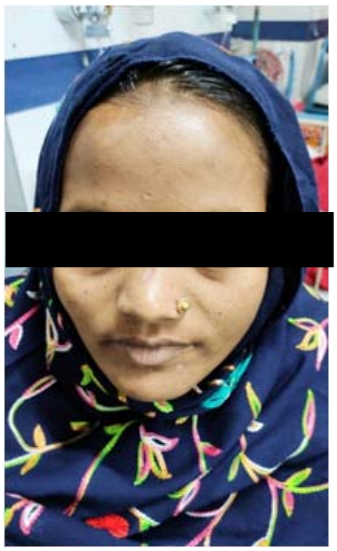

Fig 3 — The patient

negative. The patient was started on 0.5mg/kg steroid and Hydroxychloroquine and was planned for NIH regimen Cyclophosphamide. But the patient then developed sudden onset shortness of breath with severe tachypnea. Repeat echocardiography (Fig 1) revealed vegetation on tricuspid valve of size 7*4 sqmm which was further confirmed by transesophageal echocardiography. But the pericardial effusion was reduced, She had right ventricular strain pattern on ECG during the episode. Blood culture sent by infective endocarditis protocol came back negative. HRCT thorax and CT pulmonary angiography was done. It showed diffuse parenchymal lung disease with alveolar hemorrhage and there were no signs of chronic or acute thromboembolism (Fig 2&3). This presented a therapeutic dilemma about the need for anticoagulation, and considering the alveolar hemorrhage, it was not given. BAL study was done which revealed no bacterial, mycobacterial or fungal growth and RBC were seen in the BAL fluid, thus confirming alveolar hemorrhage. On steroids and supportive management, the patient stabilized and was discharged for follow up of vegetation size, renal parameters and lung manifestation (Table 1).

Table 1 — *Table showing the various blood parameters during hospital stay*

| | D1 | D3 | D5 | D10 | D14 |
|---|---|---|---|---|---|
| | | (steroid from D4) | | | |
| Hb(gm/dl) | 7.0 | 6.0 | 6.8 | 8.1 | 9.1 |
| PCV(%) | 23.6 | 20.4 | 23.1 | 26.4 | 31.4 |
| TLC (per µL) | 10,000 | 9000 | 8900 | 13400 | 11000 |
| Ur/Cr(mg/dl) | 92/5.1 | 110/2.8 | 64/1.8 | 47/1.1 | 55/1.1 |
| Na/K (mEq/l) | 134/5.5 | 137/5.0 | 134/4.2 | 140/3.4 | 138/4.4 |
| ESR (mm) | 130 | | | | |
| CPK(U/L) | 62 | | | | |
| ALP/ALT/AST IU/L | 74/14/18 | | 74/18/10 | | 60/12/16 |
| pH | 7.315 | | 7.335 | | |
| pCO2/HCO3 | 24.7/12.4 | | 25.9/13.5 | | |
| LDH (U/L) | 920 | | | | |
| C3/C4 (mg/dl) | | 18.5/5.15 | | | |
| dsDNA (IU/ml) | | 810 | | | |
| Ferritin(mcg/L) | | 47 | | | |
| PT(control)/ | 16.4(14.2)/ | | | | |
| APTT(control)/ | 32.9(33)/ | | | | |
| INR | 1.15 | | | | |

## Discussion

The most frequently involved valve in SLE with endocarditis is the mitral valve followed by the aortic valve. Tricuspid valve isolated involvement is rare3. Libman-Sacks endocarditis has been assumed to involve the formation of fibrin-platelet thrombi on the altered valve, the organization of which leads to valve fibrosis, distortion, and subsequent dysfunction[1]. They could be the main source of complications, including ischemic strokes, peripheral embolization, severe valvular regurgitation, and need for surgery.Libman sacks endocarditis is usually more commonly present in APS positive individuals, but can be present even in isolated SLE patients. APS profile can be negative in so called seronegative APS patients owing to IgA antibodies, or undetectable antibodies 4.Differentiating pericardial effusion of tuberculosis from SLE disease activity remains a clinical challenge.

## References


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