CASE REPORT

Interesting presentation of Systemic lupus erythematosus in a postpartum lady

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SUMMARY

Systemic lupus erythematosus (SLE) is a multi-systemic autoimmune disease and is commonly affecting women of childbearing age. It can present for the first-time during pregnancy, causing diagnostic and treatment challenges and resulting in poor maternal and foetal outcomes. Cardiac tamponade as the manifestation in SLE is rare. We reported a case of cardiac tamponade as the presentation of SLE in a post-partum patient, where both the patient and the new-born had a good outcome and discussed its management.

INTRODUCTION

SLE is a systemic autoimmune disease that affects multiple organs and hence causing various clinical presentations. Cardiac involvement such as pericardial effusion is well recognized in SLE. However, it is usually small and hemodynamically insignificant.[1] The incidence of cardiac tamponade in SLE is usually less than 1%.[2] The risk factors of cardiac tamponade in SLE are female, presence of anaemia, renal disease, pleuritis, higher ESR or lower C4 levels.[1] Urgent pericardiocentesis, high doses of glucocorticoids and hydroxychloroquine are usually the mainstay of treatment. We presented a rare case of cardiac tamponade as the presentation of SLE in a post-partum patient, and described its predisposing factors, management and its successful outcome.

CASE PRESENTATION

Mrs A, a 27-year-old para 2 female presented to us with heart failure symptoms for one month after delivery. Antenatally, she was under another hospital follow up for persistent proteinuria, which was detected at 36 weeks of gestation. 24 hours urine protein was 1.24g/dL. She was normotensive throughout the pregnancy. However, toward the end of her pregnancy she started to have mild pedal oedema, otherwise no other symptom. Clinically she had no features of connective tissue disease. She delivered a baby boy via normal vaginal delivery in that particular hospital uneventfully. She developed failure symptoms after delivery and it was progressively worsening. Upon arrival she was tachypnoea and tachycardia. Her blood pressure was 98/67 mmHg, heart rate was 138 beat/minute and respiratory rate was 28 breath/minute. Cardiovascular examinations showed jugular venous pressure (JVP) elevated with muffled heart sound. Her condition worsened rapidly and was intubated for impending respiratory distress. Electrocardiography (ECG) revealed sinus tachycardia with decreased voltage (Figure 1). Her chest X-ray showed cardiomegaly. Bedside echocardiogram showed large circumferential pericardial effusion with evidence of cardiac tamponade (Figure 2). Left ventricular (LV) systolic function was severely impaired with the ejection fraction of 25-30%. She was initially diagnosed as cardiac tamponade secondary to acute heart failure with peripartum cardiomyopathy. An urgent pericardiocentesis was performed under echocardiographic guidance after consulted cardiology team of the referral centre. 1.5 litre of yellowish pericardial fluid was drained. Haemodynamic monitoring was immediately improved after pericardiocentesis. She was admitted to intensive care unit for close monitoring and further care.

Laboratory investigations revealed anaemia (Hb 8.1g/dL) with normal white cell and platelet counts. Iron studies confirmed iron deficiency anaemia. Pro BNP was elevated to 792pg. Further investigations were performed to determine the underlying cause. Immunological workup showed a strongly positive antinuclear antibodies (ANA) with the titre of 1:1280, homogenous and speckled pattern, positive anti-double stranded DNA (DsDNA) and low C3 complement. 24 hours urinary protein was 2g/day. Otherwise, troponin I, thyroid function test, sputum tuberculosis tests and renal ultrasound were normal (Table I). With the positive ANA, clinical and immunological findings, she was diagnosed as SLE after rheumatology consultation. She was started with intravenous (IV) methylprednisolone and hydroxychloroquine. Renal biopsy was performed subsequently by nephrology team and the result showed class III lupus nephritis. Cyclophosphamide was added. She was given IV Cyclophosphamide 0.75g/BSA for total of 6 cycles. She responded well to the treatment given. Currently she is on maintenance therapy with oral Azathioprine. Both the mother and the baby were subsequently discharged well. The baby does not have any lupus rash or bradycardia. She was seen in clinic during follow up, clinically she was well. Repeated echocardiography showed an improvement of LV ejection fraction to 48% with minimal pericardial effusion without recurrence (Figure 3).

DISCUSSION

Our patient presented with cardiac tamponade, which is a medical emergency that requires urgent intervention. The clinical features include chest discomfort, dizziness, shortness of breath, tachycardia and tachypnoea. Beck's triad with

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Table I

INVESTIGATIONS	RESULTS
C3	0.19 g/L (0.9 - 1.8)
C4	0.11 g/L (0.1 - 0.4)
ANA	Positive, 1:1280, homogenous & speckled
ENA	Ro52, Ro60, SSB positive
DsDNA	Positive 517IU/ml
Lupus anticoagulant	Not detected
Anti-cardiolipin antibody	Normal
Anti-beta 2 glycoprotein 1	Normal
Pericardial fluid FEME	Yellowish
pH: 8	
No cell count	
No organism	
Pericardial fluid biochemistry	Glucose: 7.36
Protein: 41.9g/L (serum 69)	
LDH: 219U/L (serum 530)	
Pericardial fluid culture	No growth
Pericardial fluid cytology	Atypical cells seen. Likely reactive mesothelial cells
Pericardial fluid MTB C&S	No MTB isolated

muffled hearts sounds, hypotension and jugular venous distension might be demonstrated in physical examinations. The diagnosis is usually supported by the specific ECG changes and chest X-ray. The classical yet uncommon ECG change is electrical alternans, and the more common change is sinus tachycardia or decreased voltage. The diagnosis is usually confirmed by echocardiography, which can determine the size of pericardial effusion, look for collapsed ventricles and assess the diastolic function.

Cardiac tamponade occurs when accumulation of pericardial fluid is causing increased intrapericardial pressure to the critical point where ventricular filling is restricted and subsequently reduced cardiac output and haemodynamic instability. Pericardial effusion can be caused by increased production of pericardial fluid or by accumulation of pericardial fluid due to increase in systemic venous pressure resulting in reduced reabsorption, which is commonly seen in heart failure or pulmonary hypertension.[3] The development of cardiac tamponade also depends on the rate of pericardial fluid accumulation. In the case of rapid accumulation, such as trauma, small amounts of pericardial fluid may lead to cardiac tamponade.[3] If the pericardial fluid increases slowly the pericardial sac can expand to accommodate more fluid before the development of cardiac tamponade. The common causes of cardiac tamponade are pericarditis, tuberculosis, malignancy and trauma. Other uncommon causes include pneumopericardium, bacterial infection, aortic dissection, uraemia, post-myocardial infarction, radiation induced and collagen vascular diseases, as seen in our patient.[3] It is important not to miss SLE as the underlying cause of cardiac tamponade as SLE can affects both the mother and the foetus. Pregnant mother is at risk of developing pre-eclampsia, gestational diabetes, and lupus nephritis.[4] Foetal is at risk of congenital heart block, miscarriage, intrauterine growth restriction, intrauterine foetal death or preterm labour.[5]

SLE is a multi-systemic chronic autoimmune disease and commonly affects women in child bearing age. It is commonly diagnosed from 15-44 years old with females to males in 12:1 ratio.[6] Studies showed that in pregnant patient there is a 2-3-fold increase in SLE activity.[7] The common presentations of SLE include constitutional symptoms, occurred in 50-90% of patients, musculoskeletal up to 90% of patients and cutaneous involvement seen in 50% of patient.[7] Most cases of SLE involve cutaneous or musculoskeletal lesion, however, it was not seen in our patient. Other organs involvements are renal, pulmonary involvement and cardiac manifestation. The most common cardiac manifestation in SLE patients is pericarditis, with or without pericardial effusion, occur in up to 25% of patients.[7] Other cardiac manifestations include myocarditis and coronary artery disease. It is commonly associated with high ANA levels. Other positive lupus serologies include Anti-DsDNA, anti-Smith and Anti-Histone Antibodies [6]. it is also important to check Ro/SSA and La/SSB as they are associated with congenital heart block. However, there are no complication during our patient's pregnancy and baby is born without notable rash and has normal heart rate.

In a retrospective study of 409 SLE patients conducted in India, 104 patients (25%) was diagnosed with pericarditis and 24 patients (5.9%) diagnosed with cardiac tamponade. Out of the 24 patients with cardiac tamponade, 12 (2.9%) had cardiac tamponade as the presenting feature.[8] In another series of 395 SLE patients, it shows that 75 patients (19%) had pericarditis. 10 of them (2.5%) developed tamponade and 4 patients (1%) presented as the initial presentation.[1] Therefore, it is rare for cardiac tamponade to occur in SLE, either throughout the disease course and or as the initial presentation.

The treatment of SLE involves aiming for disease remission, preventing flares and reducing their severity. Steroids should be initiated especially in severe cases such as cardiac tamponade or lupus nephritis. Hydroxychloroquine is a safe medicine in pregnancy. It is also effective in preventing disease flare, pre-eclampsia and neonatal heart block.[9] Aspirin is a useful and safe medicine in pregnancy and

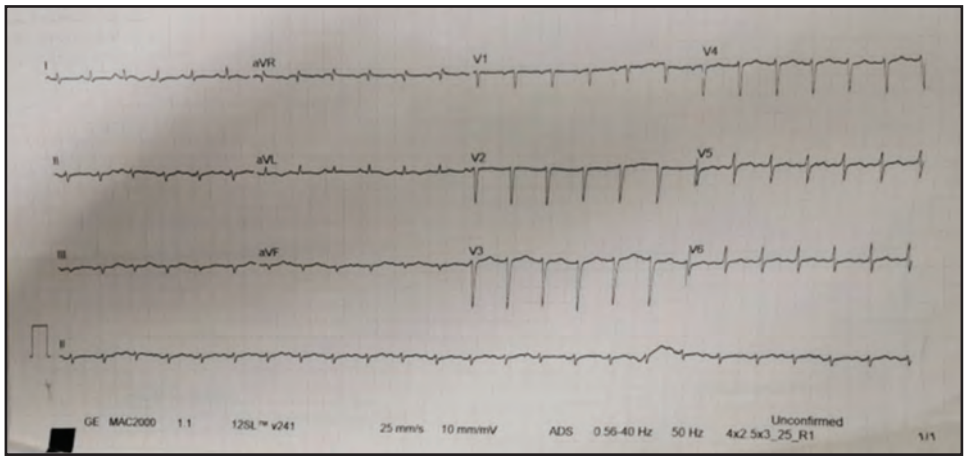

Fig. 1: ECG

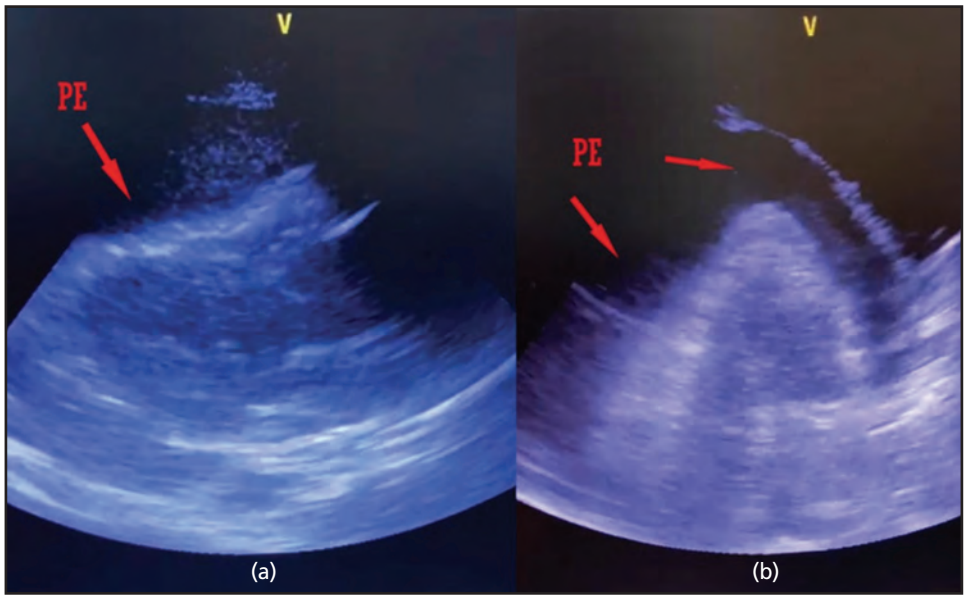

Fig. 2: a) Subcostal view: arrow showed large pericardial effusion compressing right atrium and right ventricle. b) Apical 4 chambers view: arrows showed global pericardial effusion.

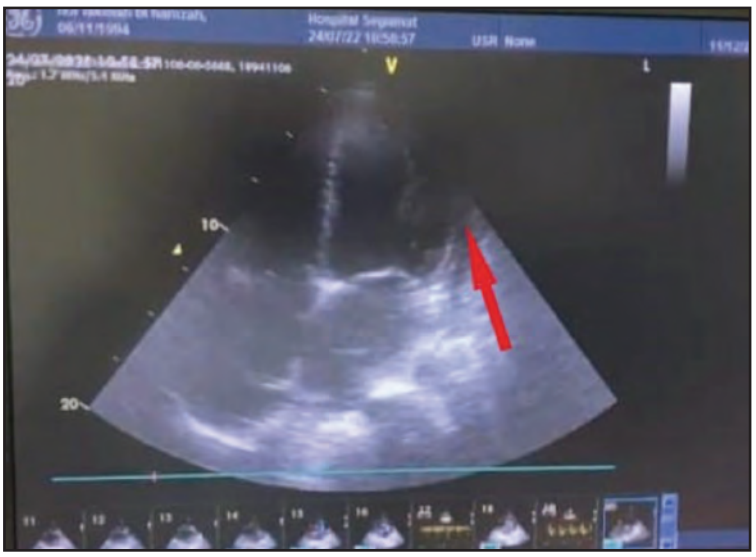

Fig. 3: Arrow showed minimal pericardial effusion

should be started at 12 weeks' gestation.[9] Immunosuppressants such as mycophenolate and cyclophosphamide are contraindicated in pregnancy.

CONCLUSION

Cardiac tamponade is a life-threatening medical emergency that could be fatal. This case highlights the interesting atypical presentation of SLE in a post-partum lady and the diagnostic and treatment challenges especially in a district hospital. Our intention of reporting this case is to emphasize the importance of early detection, high index of suspicion, timely intervention and collaborative multi-disciplinary team approach to ensure the optimal outcomes for the patient and the new-born.

TAKE HOME MASSAGE

SLE is diagnosed by the European League Against Rheumatism (EULAR) and American College of Rheumatology (ACR) classification criteria, which consists of different organs. Patients may present with variety of features and manifestation, and may be overlooked and lead to delay in making the diagnosis. Hence, in patients present with symptoms involving more than one organ, they should be follow up closely and a suspicion for SLE should be high.

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