Severe Pulmonary arterial hypertension PAH is rarely observed as initial manifestation of systemic lupus erythematosus SLE. Lupus myocarditis may rarely be initial presentation as a fatal complication of SLE, which mainly involved with left ventricle. Here we present a case of an incident PAH woman who had a delivery history 4 months previously. During hospitalization, she suffered from extensive myocardial damage, refractory ventricular tachycardia and cardiogenic shock. Ultimately, she was diagnosed with SLE, fulminant lupus myocarditis and PAH associated with connective tissue disease. After a large dose of methylprednisolone and PAH target therapy treatment, she recovered successfully and got a good result. To our knowledge, this is the first case report of pulmonary arterial hypertension and fulminant lupus myocarditis as initial manifestation of SLE in a previously healthy postpartum woman.

**KEY WORDS:** Systemic lupus erythematosus  Myocarditis  Pulmonary arterial hypertension

**INTRODUCTION**

PAH is one of the frequent observed complications of SLE which remodeling of the small pulmonary arteries leads to a progressive increase in pulmonary vascular resistance and right heart failure\(^1\). But severe PAH is rarely observed as initial manifestation of SLE. According to recent data the clinical detection of SLE myocarditis ranges from 3% to 15%, although its frequency in autopsy studies is higher, suggesting subclinical presentation of the disease\(^2\). However, clinical symptoms of heart failure are unusual, occurring in only 5%-7% of patients\(^3\).

Lupus myocarditis may rarely be initial presentation as a fatal complication of SLE. After a careful review of the published literature, we believe that this is the first case of pulmonary arterial hypertension and fulminant lupus myocarditis as initial manifestation of SLE. Furthermore, myocarditis predominantly involved right ventricle rather than left ventricle in this case.

**THE CASE**

A 26-year-old prime previously healthy woman developed exertional dyspnea without dizziness, syncope or chest pain after 4 months of full-term pregnancy. Her pregnancy and puerperium all was normal and her child was healthy. She went to local hospital and echocardiogram revealed a dilated right ventricle with a systolic pulmonary arterial pressure (PASP) of 60 mmHg and mild tricuspid regurgitation. She was referred to Shanghai Pulmonary Hospital for further diagnosis and treatment.

On admission, her temperature was 37.2°C, pulse was 122 beats per minute, blood pressure was 104/70 mm Hg, respiration rate was 20 breaths per minute, and oxygen saturation was 96.2% on room air. Auscultation revealed loud P2, and a grade II systolic murmur at the tricuspid area. Legs was mildly edematous.

Laboratory findings were: NT-pro-brain natriuretic peptide 3473 pg/ml(normal range:0-125 pg/ml), TnI 0.1900 ug/l(normal range:0-0.1 ug/l), ESR 42 mm/h(normal range:0-20mm/h). Arterial-blood-gas analysis showed a pH:7.43, PaO2:84 mmHg, PaCO2: 25.9 mmHg, SpO2:96.2 %. An electrocardiogram revealed sinus tachycardia, right axis
deviation, dilated and hypertrophic right ventricle (Figure 1A). Echocardiogram showed dilated right ventricle with a systolic pulmonary arterial pressure (PASP) 66 mmHg, moderate tricuspid regurgitation, right ventricle systolic dysfunction (TAPSE 1.3 cm), mild pericardial effusion. Left ventricle function was normal (Figure 2A).

We primarily diagnosed her as pulmonary arterial hypertension and right heart failure with WHO-FC III. She was treated with dobutamine and prostanoids (ILOPROST 10ug q4h) inhalation. The patient’s clinical status was slightly improved during treatment.

On the third day of admission, the patient felt palpitation without chest pain, ECG showed sinus tachycardia, ventricular premature beat and ST segment elevation in III, AVF, V1-V4 lead (Figure 1B). Blood test showed high level of cardiac marker with CK-MB 2.8500 ug/L, NT-proBNP increased significantly to 6432 pg/ml. Meanwhile, her serological findings were positive for SSA and SSB. Considering the patient was young woman without coronary artery disease history, ECG showed extensive myocardial damage, and connective tissue disease marker was positive, leading to think as fulminant myocarditis instead of acute myocardial infarction (AMI), highly suspected CTD-PAH. The patient was given methylprednisolone and large dose of vitamin C therapy. Repeated ECG showed persistent ST segment elevation. Six hours later, the patient developed episode of Adams-Stokes syndrome with abruptly decrease in BP and SpO2. ECG showed multifocal PVC and ventricular tachycardia (Figure 1C), echocardiography showed right heart enlargement, severe TR and TAPSE 0.9 cm (Figure 2B). We repeatedly gave her CPR and electric defibrillation 8 times together with lidocaine, large dosage of methylprednisolone (total dosage 240mg), dobutamine, dopamine, adrenaline and norepinephrine. Patient was resuscitated and his consciousness level gradually restored. Since the next morning, ventricular tachycardia no longer occurred. Later, ds-DNA level was reported 37.32 IU/ml (normal range: 0-7.0 IU/ml). ANA titre was positive and antiphospholipid antibody antibodies was negative. The patient was diagnosed with SLE according to the American College of Rheumatology criteria. Methylprednisolone 1-2mg/kg day to maintain the 1-2 week, hydroxychloroquine 0.1 bid po were initiated for lupus myocarditis. Ambrisentan with sildenafil were added to decrease pulmonary vascular resistance as she couldn’t inhale iloprost. ECG showed ST segment gradually declined to baseline (Figure 1D), and cardiac enzyme and NT-proBNP gradually decreased to normal range. After 27 days hospitalization, her RHC showed: RAP 7/2/4 mmHg, PASP 40/19/28 mmHg, PCWP 6/3/4 mmHg, CO 3.17 L/min, CI 5.25 Wood U, negative pulmonary vasoreactive test. Echocardiogram showed mild dilated right heart with a systolic pulmonary arterial pressure (PASP) of 42 mmHg, mild tricuspid regurgitation, TAPSE 2.0 cm. (Figure 2C, Table 1)

**DISCUSSION**

We presented a rare case of SLE who presented with PAH and fulminant myocarditis, right heart failure and life-threatening ventricular arrhythmias (VT/VF). To our knowledge, this is the first case report of PAH and fulminant lupus myocarditis as initial manifestation of SLE in a previously healthy postpartum woman.

PAH is a devastating disease in which remodeling of the small pulmonary arteries leads to a progressive increase in pulmonary vascular resistance (PVR) and right heart failure [1]. In China, the proportion of CTD-PAH is about 19% including SLE 51.1%, pSS 15.6%, SSc 15.5% (unpublished data). But severe PAH is rarely observed as initial manifestation of SLE.

Myocarditis is one of the most possible forms of cardiac involvement in systemic lupus erythematosus. Its clinical presentation ranges from asymptomatic patients with self-limited disease to fulminant heart failure that can lead to death. Most myocarditis in SLE is asymptomatic but may be manifest with fever, dyspnea, palpitation, and nonexertional chest pain [2]. Subclinical cardiac involvement in SLE is seen by the
prevalence of myocarditis from necropsy studies to be 40–70%. However, symptomatic myocarditis had been reported in only 5–10% of cases.[5] The presentation of myocarditis depends on the degree of cardiac dysfunction ranging from mild dyspnea or chest pain, arrhythmias, heart failure, cardiogenic shock, to sudden death.[6] Lupus myocarditis may rarely be initial presentation as a fatal complication of SLE. Lupus myocarditis need urgent clinical attention because of the likely progression to arrhythmias, conduction disturbances and heart block, dilated cardiomyopathy, and heart failure. The prevalence of arrhythmias and conduction system disorders in SLE is currently unknown due to the small number of studies in the literature. Malignant ventricular arrhythmias are rarely reported[7].

In our case, the PAH patient presented with elevated ST segment with cardiac injury and malignant arrhythmias on the second day of admission. As she is a young woman without chest pain, D-dimer nearly normal, abnormal connective tissue disease marker, we considered myocarditis rather than AMI. Subsequent rheumatic and immunology examination indicated as a lupus myocarditis. Symptomatic left ventricular dysfunction is the most common clinical presentation of lupus cardiomyopathy and is potentially life threatening which acute onset of a marked reduction of the left ventricular ejection fraction (LVEF). Echocardiographic findings in lupus myocarditis mainly include decreased ejection fraction, increased chamber size. Interestingly, in our case SLE myocarditis mainly involved right heart as echo showed her right heart size and function was significantly abnormal (TAPSE low and RV dilation) while her left ventricle function was always normal. A systematic MEDLINE/PubMed from 1993 to 2014 only identified one case of myopericarditis with predominantly right ventricle involvement as the initial manifestation of SLE[8].

The goal of treatment for acute lupus myocarditis is to manage heart failure, arrhythmias and SLE activity. In our case, lupus myocarditis predominantly involved right ventricle which mainly presented as PAH and right heart failure, we used large dose of intravenous methylprednisolone, PAH target therapy(sildenafil and ambrisentan), and positive inotropic drugs to help her recover.

This case remind us, clinician should pay more attention on postpartum PAH, especially screening of CTD. Identifying the cause is critical as it dictates therapy. Early diagnosis and a combination treatment for right heart failure, immunosuppression and arrhythmias may result in a favorable outcome.

**Disclosures** None.

**REFERENCES**