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Diagnosis and management challenges of recurrent lupus pericarditis in pregnancy: A case report

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ABSTRACT

Systemic lupus erythematosus (SLE) is an autoimmune multisystem disease. Pericarditis in SLE can lead to severe effusion and cardiac tamponade, and is associated with significant morbidity and mortality. Therefore, early diagnosis and treatment are essential. A 32-year-old woman at 21 weeks of gestation with a history of lupus pericarditis two years previously presented to the emergency department with shortness of breath, fever, and weakness. Transthoracic echocardiography revealed a massive pericardial effusion, right ventricle failure, pulmonary hypertension, severe tricuspid regurgitation, and mild mitral regurgitation. A chest X-ray suggested pericardial effusion accompanied by pulmonary edema. Due to worsening of the patient's health, a joint decision was made with her and her family to terminate the pregnancy. Most cardiac manifestations of SLE worsen during pregnancy and can lead to life-threatening conditions such as cardiac tamponade or congestive heart failure. This is a rare case of recurrent lupus pericarditis in pregnancy accompanied by massive pericardial effusion, heart failure and pulmonary edema. Management is challenging because the most effective drugs are known to be harmful to the fetus.

1. Introduction

Systemic lupus erythematosus (SLE) is a multisystem chronic inflammatory autoimmune disease that occurs most often in women of reproductive age. The etiology is unknown. However, several genetic, immunological, endocrine, and environmental factors play a role in the etiopathogenesis of SLE [1]. The highest incidence of SLE is in African-Americans, while the lowest is in Caucasians [2].

SLE can affect many organ systems, including bones, joints, lungs, skin, and heart. This case report describes the presentation and management of a 32-year-old woman at 21 weeks of gestation with lupus pericarditis, massive pericardial effusion, pulmonary edema and right heart failure.

2. Case Presentation

A 32-year-old woman, G3P2002, at 21 weeks of gestation, presented to the emergency department with a 5-day history of shortness of breath at rest, fever, and weakness. She had a past medical history of SLE with pericardial effusion and had undergone pericardiocentesis two years previously, and was currently being treated with mycophenolate sodium and methylprednisolone. She had had two successful pregnancies delivered vaginally and the younger child was aged 7 years. The patient was not using contraception and was unaware that she was pregnant due to irregular menstrual cycles. She was underweight (BMI = 15.76 kg/m^2). On examination she was pyrexic (38.7 °C), dyspneic (28×/min), tachycardic (138×/min), and hypotensive (80/44 mmHg). Physical examination revealed bilateral basal crepitations suggestive of pulmonary edema as well as a heart murmur. No other manifestations of systemic lupus were found.

Bedside ultrasound examination found a 21/22 singleton intrauterine pregnancy. Fetal heart rate was 164 beats/min. A transthoracic echocardiogram revealed a massive pericardial effusion, right ventricle failure, pulmonary hypertension, severe tricuspid regurgitation, and mild mitral regurgitation. Chest X-ray showed a pericardial effusion accompanied by pulmonary edema (Fig. 1). Laboratory examination revealed hypoalbuminemia (2.9 g/dL) and elevated brain natriuretic peptide (6633.10 pg/mL), C-reactive protein (CRP) (6.60 mg/dL), and complement C4 (8 mg/dL). Urinalysis revealed an elevated albumin to creatinine ratio (≥ 300 mg/g Cr) and protein creatinine ratio (≥ 50 mg/g). She was diagnosed with lupus pericarditis with massive pericardial effusion, a high probability of pulmonary hypertension, right heart

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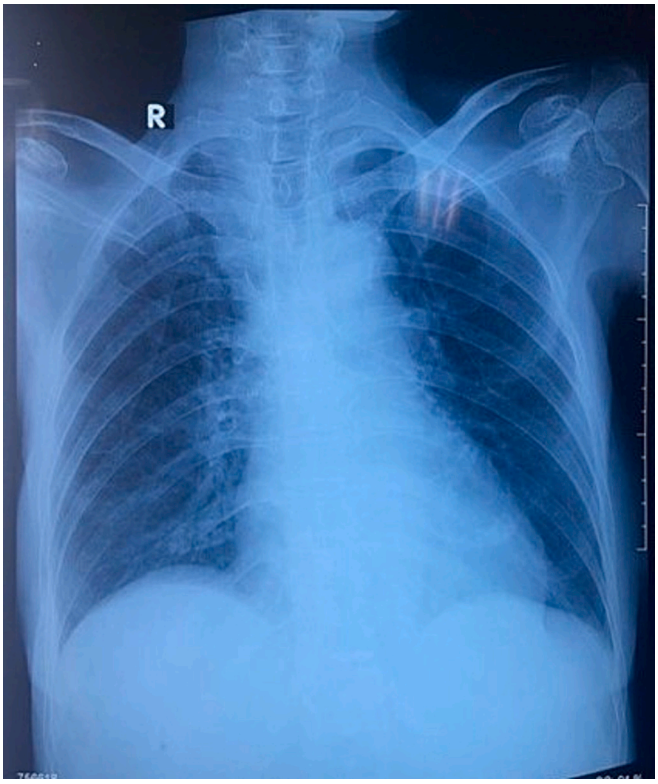

Fig. 1. Chest X-ray showing pericardial effusion accompanied by pulmonary edema.

failure, pulmonary edema and hypoalbuminemia. Pulse dose glucocorticoid 500 mg IV per day for 3 days was planned for disease control, as well as pericardiocentesis.

On examination, her vital signs were significant for worsening dyspnea, hypoxemia, and hypotension. She was transferred to the intensive care unit and started on noninvasive ventilation and inotropic support. A multidisciplinary team consisting of cardiology, rheumatology, and maternal–fetal medicine discussed treatment options. The main concern was the risk of worsening of the disease if the pregnancy progressed. Furthermore, treatments such as mycophenolate sodium or cyclophosphamide could not be used because of their known teratogenicity. Intravenous immunoglobulin, which is regarded as relatively safe during pregnancy, was the only option available besides steroids and azathioprine. However, the effectiveness of this agent for lupus carditis has not been proven. After discussion with the patient and her family about her life-threatening health condition, possible treatments options and the risks of continuing the pregnancy, it was agreed to terminate the pregnancy. Sterilization was also advised and agreed.

Due to the patient's unstable clinical condition, induction of labor and vaginal delivery were deemed unsuitable. Hysterotomy was therefore planned. After administration of pulses of glucocorticoid, the patient underwent pericardiocentesis and pigtail catheter installation, followed by hysterotomy and sterilization. The procedures were well tolerated. She was continued on methylprednisolone 62.5 mg daily, carvedilol 12.5 mg twice a day, furosemide 80 mg daily, sildenafil 20 mg three times a day, and beraprost sodium 20 μg three times a day. After the procedure, the patient was treated for one day in the intensive care unit, and on the second day, treatment continued in the integrated cardiac care room for eight days. Nine days after the procedure, the patient's clinical condition had improved and she was discharged for outpatient treatment.

Table 1
Recommendations for diagnosis of acute pericarditis [8].

Recommendations	Class of recommendation	Level of evidence
Echocardiography is recommended in all patients with suspected acute pericarditis	I	C
Transthoracic echocardiography is recommended in all patients with suspected acute pericarditis.	I	C
Chest X-ray is recommended in all patients with suspected acute pericarditis.	I	C
Assessment of markers of inflammation (i.e., CRP) and myocardial injury (i.e., CK, troponin) is recommended in patients with suspected acute pericarditis.	I	C

3. Discussion

The incidence of lupus flares in pregnancy is still controversial. Some studies show an increase in the number of lupus flares in pregnancy, while others do not [3]. Cardiac manifestations are common complications of SLE in pregnancy, affecting up to 50 % of patients. Cardiac manifestation of SLE can present with features of myocarditis, pericarditis, endocarditis, valvular lesions, and conduction abnormalities [4]. Acute pericarditis at the time of diagnosis of SLE is rare, occurring in only 1 % of individuals [5]. It is usually mild and asymptomatic. However, in rare cases, it can lead to severe effusion and cardiac tamponade [6].

Echocardiography is the standard method to investigate pericardial abnormalities and can demonstrate mild effusions or thickening of pericardial layers. Electrocardiography shows widespread ST elevation or PR depression in the acute phase. Pericardial effusion can be confirmed via chest X-ray in a massive effusion [7]. Laboratory findings supporting acute pericarditis diagnosis include leucocytosis, elevated CRP, and elevated erythrocyte sedimentation rate. Elevated creatine kinase (CK) and troponin are markers of myocardial injury in myocarditis (Table 1) [8].

The management of life-threatening acute pericarditis in pregnant woman can be challenging, and depends on whether maternal safety or preservation of the pregnancy will be the priority. Decisions need to balance risks to maternal and fetal health. Treatment depends on the severity of the disease and the amount of pericardial inflammation. The management of pericarditis with massive pericardial effusion is pericardiocentesis, followed by pericardial fluid analysis to find the cause [8]. In mild pericarditis, nonsteroidal anti-inflammatory drugs and corticosteroids are effective. In more severe cases or tamponade, a higher dose of corticosteroid is necessary, and in patients with recurring pericarditis, chronic immunosuppression with methotrexate, cyclophosphamide, or mycophenolate mofetil is the recommended therapy and has shown a good response. However, those drugs are not recommended for use in pregnancy because their adverse effects on the fetus [9].

4. Conclusion

Recurrent pericarditis as a relapse of SLE is rare. The diagnosis and management are still challenging. A multidisciplinary team approach with cardiologists, rheumatologists, and obstetricians is important in managing patients with complex conditions. The risk of worsening disease by continuing the pregnancy and the risks and benefits of treatment options should be clearly explained to the patient and family. Discussion of future fertility and contraception is essential.

Contributors

Alexander Indra Humala contributed to patient care, the conception of the case report, the acquisition and interpretation of data, the

literature review, and the drafting of the manuscript.

Manggala Pasca Wardhana contributed to the conception of the case report, performed the literature review, and drafted the manuscript.

Both authors approved the final submitted manuscript.

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Patient consent.

The patient consented to publication of the report and all accompanying images.

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Conflict of interest statement

The authors declare that they have no conflict of interest regarding the publication of this case report.

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