

# Peripartum Cardiomyopathy and Systemic Lupus Erythematosus Carditis, a diagnostic dilemma.

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## Abstract

A young woman at 8 months postpartum presented with dyspnoea, orthopnoea and swelling of lower limbs in which physical examination, chest radiography and echocardiogram were suggestive of acute congestive heart failure with left ventricle dilatation and dysfunction. A suspicion of peripartum cardiomyopathy was made and treated with conventional drug therapy but the patient continued to develop multiple episodes of heart failure. Over time she developed fever and polyarthritis following which autoantibodies, complement level and 24-hour urinary protein were done which helped us to make the diagnosis of Systemic Lupus Erythematosus (SLE) nephritis. The patient was started on high dose corticosteroids. However, after a week, patient developed cardiogenic shock following which intravenous pulse Cyclophosphamide was started and the patient improved clinically and biochemically.

**Keywords:** Lupus myocarditis; Peripartum cardiomyopathy; Systemic Lupus Erythematosus.

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## Introduction

Peripartum cardiomyopathy (PPCM) is idiopathic cardiomyopathy presenting with heart failure secondary to left ventricular systolic dysfunction toward the end of pregnancy or in months following delivery where no other cause of heart failure is found.<sup>1</sup> Partial or complete recovery is common in PPCM.<sup>2</sup> We present a case of 25 years female who presented symptoms of cardiac failure 8 months following delivery. Diagnosed and managed as PPCM, she went on to develop refractory heart failure more than 4 times in a year. Only when she developed fever and arthralgia, she was diagnosed with Systemic Lupus Erythematosus (SLE) carditis. Since PPCM is a diagnosis of exclusion this case justifies the importance of proper evaluation especially when heart failure doesn't respond to the usual treatment.

## Case Presentation

A 25-year-old female presented at a cardiac centre with

dyspnoea, orthopnoea and swelling of the body for 15 days at 8-month postpartum. She had delivered a male child at term and the child had no health issues. Clinical examination revealed bilateral pretibial pitting oedema and bi-basal inspiratory pulmonary crackles. Chest X-ray showed cardiomegaly and interstitial pulmonary oedema. Electrocardiogram (ECG) revealed sinus tachycardia. Echocardiogram showed severe left ventricular systolic dysfunction (LVEF 30%), moderate mitral regurgitation (MR) and mildly dilated left ventricle end-diastolic dimension/end-systolic dimension (LVEDD/LVESD; 5.7/4.7cm). A provisional diagnosis of PPCM was made and she was managed with salt restriction, high dose diuretics and angiotensin-converting enzyme inhibitors. Her initial symptoms were controlled for 3 months with medications. However, she developed multiple episodes of heart failure after 3, 5, 6 and 12 months of initial episode and was labelled and treated as refractory heart failure.

Few months after the last episode, she developed intermittent

fever associated with polyarthritis for which she visited our centre. On examination, heart murmur with parasternal heave was noted, other examinations were normal. On evaluation, haematological parameters showed pancytopenia with haemoglobin 8.9 gm/dl, total count 3260 per cumm, and platelet 113000 per cumm. Urine routine showed mild proteinuria (1+albumin) with 2-3 RBCs / hpf. Twenty-four-hour urinary protein was 1.12 gm/day (Reference range <0.15 gm/day) and LDH was raised to 1346.0 U/L. Abdominal ultrasound scan was performed which demonstrated normal-sized kidneys however there was increased echotexture. Other investigations for infective pathology of fever were negative.

Evaluation of immunological markers was made. Abnormal findings were; positive Anti Nuclear Antibody (ANA) (not quantified), positive anti-dsDNA (66 IU; Reference positive >50 IU), positive anti-cardiolipin (22.4 GPLU/ml; Reference positive >14 GPL/ml) and low complement C3 (0.6; Reference 0.9-1.8). On the background of pancytopenia, positive ANA, ds DNA, significant 24-hour urinary protein and low C3 level she was diagnosed with SLE nephritis (SLICC Criteria). Treatment of active lupus nephritis was commenced with loop diuretics, hydroxychloroquine, spironolactone and prednisolone. Renal biopsy was planned but not performed.

One week after starting the treatment, the patient developed shortness of breath and orthopnea NYHA class IV. On examination, mild confusion with blood pressure 80/60 mmHg, heart rate of 155bpm, increased Jugular Venous Pressure (JVP), bi-basal crackles and a grade 3/6 pansystolic murmur was present. Inotropic support was started with Dobutamine and Noradrenaline which gradually settled the signs of heart failure but there was persistent tachycardia. A clinical diagnosis of acute lupus myocarditis with cardiogenic shock was made. Loop diuretics, ACE inhibitors, hydroxychloroquine, spironolactone and prednisolone were continued and the patient was started on IV Cyclophosphamide. Beta-blocker was added following which the patient showed clinical and biochemical improvement. The level of anti ds DNA fell to 22.9 U/ml (<30: negative 30-50 borderline and > 50 positive) and CRP level to 5563 ng/ml (reference: 68-8200 ng/ml) showing decreased disease activity. The patient was discharged on oral steroid and azathioprine.

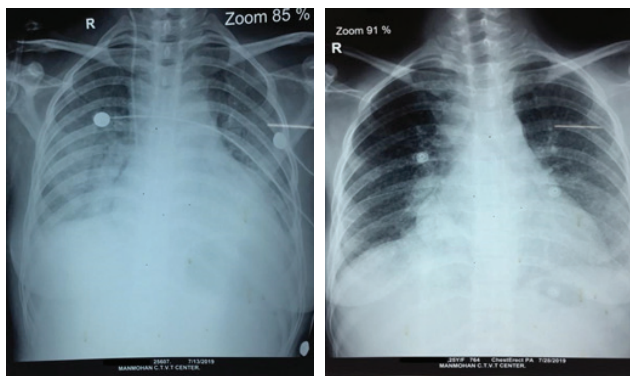


Figure 1: chest radiograph in time sequence: First figure showing interstitial pulmonary oedema that didn't resolve on the treatment of heart failure. The second figure shows the rapid improvement just after 1 week of cyclophosphamide treatment.

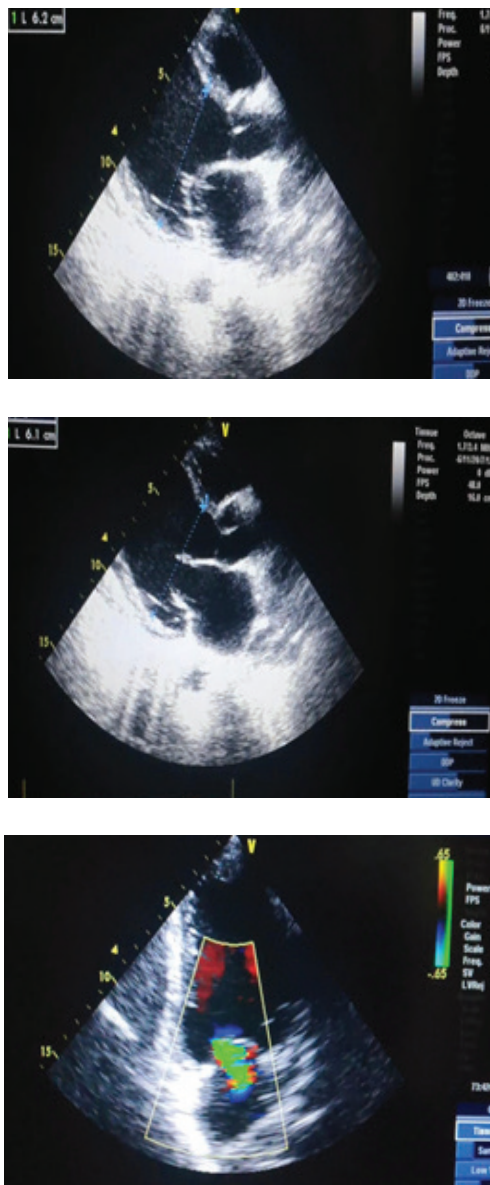


Figure 2: These echocardiographic images were taken when she did not respond to treatment and we planned to start her on IV cyclophosphamide: First figure showing Left Ventricular End Systolic Dimension (LVESD: 6.2 cm). The second figure shows the Left Ventricular End Diastolic Dimension (LVEDD: 6.1 cm). The third figure shows mild to moderate Mitral Regurgitation (MR). The mitral regurgitation was trivial and unlikely to be the cause of cardiac failure rather a manifestation of myocarditis. There was no pericardial effusion.

## Discussion

Pericarditis is the most common cardiac manifestation in SLE patient but lesion of myocardium, valve, coronary vessel, as well as conduction system, can occur.<sup>3</sup> In clinical studies, myocarditis has been found in 9% of patient with SLE.<sup>3,4</sup> The gold standard for diagnosing myocarditis is endomyocardial biopsy<sup>5</sup> but it is invasive and its diagnostic yield is low at 25-58%.<sup>5,6</sup> On the other hand, peripartum cardiomyopathy is a diagnosis of exclusion. It is idiopathic cardiomyopathy presenting with heart failure secondary to LV systolic dysfunction toward the end of pregnancy or in months following delivery where no other cause of heart failure is found.<sup>1</sup> Its incidence is higher in African countries and highest in Nigeria (1 in 102) and lowest (1 in 15533) in Japan.<sup>2</sup> Partial or complete recovery of LV function is common among patient with PPCM<sup>2</sup> and nearly in half of them, recovery of LV function occurred in 6 months.<sup>7,8</sup>

Our patient was in 8<sup>th</sup> month postpartum when she developed signs of heart failure and was suspected as a case of postpartum cardiomyopathy. Many cases of reported peripartum cardiomyopathy may actually be due to myocarditis with autoimmune aetiology.<sup>7</sup> There are case reports where a patient with SLE presenting with acute heart failure post-partum in which incorrect diagnosis of peripartum cardiomyopathy led to detrimental outcome.<sup>4</sup>

There have been case reports where acute myocarditis was the initial manifestation of SLE.<sup>4,9</sup> This is the diagnostic dilemma for clinician. ECG and chest radiograph are largely unhelpful. Therefore, the diagnosis of myocarditis in SLE largely depends on clinical suspicion and echocardiography finding. Our patient, though was diagnosed and treated for PPCM, developed refractory heart failure multiple times. After she was diagnosed with SLE nephritis she was started on IV methylprednisolone. High dose corticosteroid is the most common therapy used for lupus myocarditis.<sup>10,11</sup> But she still had an episode of heart failure with pulmonary oedema and persistent tachycardia after initiation of medication. So, she was started on IV pulse cyclophosphamide and was symptomatically better. Her improvement was also visualized with a decrease in ds DNA and CRP level. Though anticardiolipin is not a marker of disease activity, ds DNA and CRP can be used as a marker of disease activity in low resource settings.<sup>12,13</sup>

This case illustrated the difficulty in differentiating between peripartum cardiomyopathy and lupus myocarditis. Though cardiac MRI and endomyocardial biopsy are helpful they are rarely available in low resources setting. The usual outcome of peripartum cardiomyopathy is a partial or complete recovery that is why refractory heart failure should not be labeled as PPCM but a thorough a systematic search for secondary causes of carditis is necessary such as in this case.

## References

1. Regitz-Zagrosek V, Roos-Hesselink JW et al. 2018 ESC Guidelines for the management of cardiovascular diseases during pregnancy: The Task Force for the Management of Cardiovascular Diseases during Pregnancy of the European Society of Cardiology (ESC). *European Heart Journal* 2018;39(34): 3165-3241. <https://doi.org/10.1093/eurheartj/ehy340>
2. Isogai T, Kamiya CA. Worldwide Incidence of Peripartum Cardiomyopathy and Overall Maternal Mortality. *Int heart J.* 2019;60(3): 503-511. <https://doi.org/10.1536/ihj.18-729>
3. Gomez-Leon MA, Amezcua-Guerra LM. Cardiovascular manifestation of systemic lupus erythematosus. *Arch Cardiol Mex* 2008;78(4): 421-30.
4. Woo, S. I., Hwang, G. S., Kang, S. J. et al. Lupus myocarditis presenting as Acute Congestive Heart failure: A case report. *J Korean Med sci* 2009;24(1):176-8. <http://dx.doi.org/10.3346/jkms.2009.24.1.176>
5. Ramaekers J, Janssens J. Indication and diagnostic yield of endomyocardial biopsies for unexplained cardiomyopathy, a single center experience. *Acta cardiol* 2020; 75(2):1-9. <https://doi.org/10.1080/00015385.2018.1561597>
6. Bennett MK, Gilotra NA, Harrington C. et al. Evaluation of the role of endomyocardial biopsy in 851 patients with unexplained heart failure from 2000-2009. *Circulation: heart failure.* 2013;6:676-684. <https://doi.org/10.1161/CIRCHEARTFAILURE.112.000087>
7. Cooper, L. T., Mather, P. J., Alexis, J. D. et al. Myocardial recovery in peipartum cardiomyopathy: prospective comparison with recent onset cardiomyopathy in men and non peripartum women. *J card fail* 2012;18(1):28-33. <https://doi.org/10.1016/j.cardfail.2011.09.009>
8. Felker, G. M., Jaeger, C. J., Klodas, E. et al. Myocarditis and long term survival in peripartum cardiomyopathy. *Amercian heart journal* 2000;140(5):785-791. <https://doi.org/10.1067/mhj.2000.110091>
9. Cheng SM, Chang DM. Acute Myocarditis as an initial manifestation of systemic lupus erythematosus: a case report *Zhonghua Yi Xue za Zhi* 1996;58(3):205-8.
10. Chan YK et al. Intravenous cyclophosphamide improves cardiac dysfunction in lupus myocarditis. *Rheumatol* 2003;39(4):306-8. <https://doi.org/10.1080/03009740310003956>
11. Bhakta, P., Biswas BK., Banerjee, B. Peripartum cardiomyopathy: Review of literature *Yonsei Medical journal* 2007;48(5):731-47. <http://dx.doi.org/10.3349/ymj.2007.48.5.731>
12. Spronk PE, Limburg PC, Kallenberg CG. Review : Serological markers of disease activity in systemic lupus erythematosus. *Lupus* 1995;4(2):86-94. <https://doi.org/10.1177/096120339500400202>
13. Zohri MH, Tohammy MA, Obiedallah AA, et al. High-Sensitivity C-Reactive Protein in Systemic Lupus Erythematosus Patients without Cardiac Involvement: Relation to Disease Activity and Intima-Media Thickness. *Jmscr* 2017;5(3):18758-18768. <https://dx.doi.org/10.18535/jmscr/v5i3.78>