

Peripartum Cardiomyopathy Presenting with Pulmonary Edema: A Rare Case Report

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Abstract: *Peripartum cardiomyopathy (PPCM) is a dilated cardiomyopathy defined as systolic cardiac heart failure in the last month of pregnancy or within five months of delivery. Risk factors include multiparity, advanced maternal age, multiple pregnancies, pre-eclampsia, chronic hypertension, smoking, alcoholism, malnutrition, and long-term tocolysis. Its diagnosis is often delayed because its symptoms closely resemble those within the normal spectrum of pregnancy and the postpartum period. When PPCM is misdiagnosed or its diagnosis is delayed, the consequences are deadly with high mortality rate. Such a case is reported where a 6th gravida presented with pulmonary edema in ER requiring invasive mechanical ventilation undergone lower segment caesarean section (LSCS) later diagnosed as PPCM.*

Keywords: Peripartum cardiomyopathy; Echocardiography; Heart failure, pulmonary edema, mechanical ventilation

1. Introduction

Peripartum Cardiomyopathy (PPCM) refers to an idiopathic Heart Failure (HF) of unknown causes, characterized by marked impairment of left ventricular systolic dysfunction that presents in the last month of pregnancy or lasting for 5 months after delivery. ^[1]In 1997, the National Heart, Lung and Blood Institute and the Office of rare diseases commenced the workshop that established the following diagnostic criteria for peripartum cardiomyopathy:

- Development of Cardiac failure in the last month of pregnancy or within 5 months after delivery.
- Absence of an identifiable causes for the cardiac failure.
- Absence of recognizable heart disease prior to the last month of pregnancy.
- Left ventricular systolic dysfunction demonstrated by classic echo graphic criteria such as depressed shortening fraction or ejection fraction or global LV hypokinesia. ^[2, 3]

Risk factors include advanced maternal age, multiparity, multiple gestation, obesity, gestational hypertension, preeclampsia, and black race. ^[5, 6]The etiology of peripartum cardiomyopathy remains unknown, and many potential causes—including viral myocarditis, abnormal immune response to pregnancy, abnormal response to the increased hemodynamic burden of pregnancy, hormonal interactions, malnutrition, inflammation, and apoptosis—have been proposed but not proven. Another theory suggests that oxidative stress during late pregnancy leads to the proteolytic cleavage of prolactin. The resulting 16 - kDa prolactin fragment has been found to be cardiotoxic and can impair the metabolism and contractility of cardiomyocytes. Based on this proposed mechanism, bromocriptine therapy has been suggested because it inhibits prolactin secretion. Indeed, there has been at least one preliminary study in which bromocriptine improved recovery of affected women. ^[1]Another intriguing mechanism is to link peripartum cardiomyopathy to preeclampsia syndrome. This is biologically plausible given that hypertensive disorders frequently coexist with peripartum cardiomyopathy. This

shows that antiangiogenic factors—already known to be associated with preeclampsia—can induce peripartum cardiomyopathy in susceptible mice. Thus, they posit peripartum cardiomyopathy to be a vascular disease precipitated by antiangiogenic factors that act in a host made susceptible because of insufficient proangiogenic factors.

Symptoms of PPCM, which include fatigue, edema, and dyspnea, are similar to those for the normal spectrum of peripartum states and pregnancy comorbidities such as pulmonary emboli and eclampsia. Therefore, diagnosis is often delayed and the disorder is unrecognized, with devastating consequences. Mortality is as high as 20% to 50% ^[4].

2. Case Report

A 36 - year - old G6 P5 L5 A0 patient at 37 weeks period of gestation brought to our casualty with acute dyspnea. She had Five full - term normal vaginal deliveries and all her children are alive and healthy without any antenatal and postnatal complication. she was known case of hypertension and hypothyroidism on regular medication. She had no family history of preeclampsia or any cardiac illness. At the time of admission, she was severely breathless (NYHA grade 4).

On examination, her vitals were - PR of 130/min, BP 160/96 mmHg, RR 32cpm, SpO₂ 60% on room air. On auscultation bilateral extensive crepitations were heard with S3 gallop. Obstetric examination revealed uterus of 37 weeks size, firm, with cephalic presentation, FHS 106/min, regular. Internal examination showed closed os and cervix uneffaced. Hb- 9.1 gm/dl, WBC - 8800/c mm, platelet count - 414000/c mm, normal coagulation profile, raised CPK - MB and positive Trop T. ABG showed reduced PaO₂. all other investigations were within normal limits. Patient was intubated and mechanically ventilated. Pink froth was coming from endotracheal tube.

Emergency lower segment caesarean section (LSCS) was performed in view of fetal distress. A male baby of 1.9 kg was delivered. Baby was shifted to NICU for further management.

The patient was managed on the lines of pulmonary edema with I/V furosemide, ventilatory support. Fluid was restricted to maintain a CVP of 8 cm of water and I/V furosemide and mechanical ventilation was continued. On post - op day 2 there was significant resolution of pulmonary edema and the patient was weaned off from the ventilator.

A 2D - Echo done on the day of admission showed reduced LV systolic function with global left ventricular hypokinesia with ejection fraction of 18% with severe MR. Based on clinical presentation and above investigations diagnosis of PPCM was made. She was discharged on day 10 with diuretics, digoxin, and ACE inhibitors. 2D - Echo after 4 weeks showed an improved ejection fraction of 30% and it became 45% after 2 months. She was followed up to 6 months and all parameters remained normal.

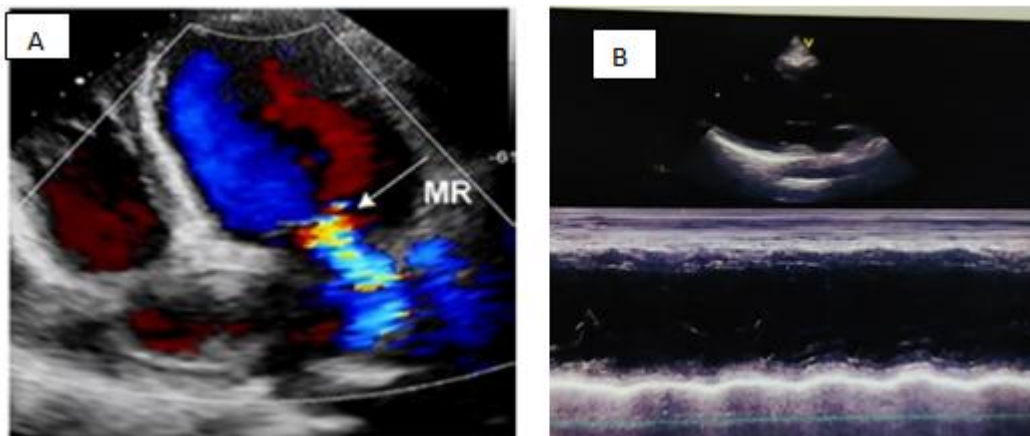


Figure: [A] Echocardiography showing: Echocardiography showing regurgitation of mitral valve (MR) (white arrow).
[B] Echocardiography showing dilated LV cavity (LVID: 65/60 mm)

3. Discussion

PPCM is a diagnosis of exclusion.^[1] Although the left ventricle may not be dilated, the ejection fraction is nearly always reduced below 45%. The incidence is quoted to be varying from 1 in 4000 to 1 in 15000 deliveries, 75% present within the first month and 45% in the first week postpartum.^[2, 5] Up to 7% may present in the last trimester of pregnancy. The etiology is uncertain; viral, autoimmune, and idiopathic have been considered.^[5, 7] The prognosis of PPCM is related to its presentation as well as to recovery of ventricular dysfunction.^[4, 8] ECG is generally within normal limits. However, sinus tachycardia or atrial fibrillation may be there if cardiomyopathy is severe. Outcome is dependent on the ejection fraction and left ventricular end - diastolic volume at diagnosis, response to medical therapy, and normalization of left ventricular function within 6 months of pregnancy.

Therapy regimens include diuretics to reduce volume overload (preload), after load reduction with angiotensin - converting - enzyme inhibitors (postpartum only) and beta - blockers after signs and symptoms of pulmonary congestion have improved. ACE inhibitors should be considered as mainstay of treatment for PPCM after delivery to prevent cardiac remodeling.^[9] Digitalis, an inotropic agent, is also safe in low dose (0.125mg) during pregnancy and may help to maximize contractility and rate control, but serum levels must be closely monitored since excessive digoxin concentrations in serum have been associated with worse outcomes in women.^[10, 11]

The route of delivery should be preferably vaginal, reserving caesarean section for obstetric indication only. Mortality rate varies from 7% to 50%.^[4] Most common causes for

mortality are progressive heart failure, arrhythmia, and thromboembolism.

The prospects of future pregnancy should be discussed with relatives. 78% of women with fully recovered left ventricular function have a normal outcome.^[12] Persistent ventricular dysfunction serves as a relative contra - indication to plan a future pregnancy. However, before planning a subsequent pregnancy, evaluation by cardiologist is recommended. Areas for future research include immune system dysfunction, the role of viruses, nonconventional treatments such as immunosuppressant, immunoabsorption, apheresis and antiviral treatment.^[1] Even though PPCM is rare, the aim of reporting this case is to create awareness of this entity, since survival depends upon accuracy of diagnosis and prompt and vigorous treatment of heart failure.

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