

Case Report

A case of Peripartum Cardiomyopathy with associated Post-Cesarean section and Grand mal seizure

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Abstract:

Peripartum cardiomyopathy (PPCM) is a rare but life-threatening disease in later stages of pregnancy. PPCM presents in the last month of pregnancy or within five months of puerperium in the absence of pre-existing heart disease. It remains a poorly understood entity and its management remains a topic of debate. Although majority recovers, long-term morbidity is quite common. Peripartum discomforts often mask and delay the diagnosis of PPCM. Unavailability of previous cardiac imaging is rather common and this makes the diagnosis even more complicated. Early and aggressive management towards reducing preload show evidence of better outcome. Here we report a case of PPCM presenting to us diagnosed after Caesarean Section followed by Grand mal seizure.

Keywords: Heart Failure, Hypertension, Peripartum Cardiomyopathy, Pregnancy.

Introduction:

Peripartum cardiomyopathy (PPCM) is rare but a well-known form of acute heart failure. PPCM affects otherwise healthy women from the last trimester of pregnancy up to 5 months postpartum. It was not up to 1930s, PPCM was recognized as a distinct clinical entity. Definition of PPCM is broad to prevent underdiagnosis. The definition developed by 2010 European Society of Cardiology (ESC) is widely accepted. For PPCM all three following conditions must meet ¹

1. Development of HF in the last month of pregnancy (or towards the end of pregnancy) or within 5 months following delivery
2. Absence of another identifiable cause of heart failure
3. Left ventricular (LV) systolic dysfunction with an LV ejection fraction (LVEF) of less than 45%, with or without LV dilatation

Even in the absence of any pre-existing cardiovascular disease, gestational hypertension and its more severe forms like preeclampsia, the HELLP syndrome and PPCM may induce cardiovascular disease²⁻⁴. Latent viral infection can

flare up in pregnancy and lead to myocarditis⁵. PPCM is suspected when left ventricular systolic dysfunction and symptoms of heart failure occur between the last month of pregnancy and the first five months postpartum^{6,7}. Some patients with higher LVEF (45-50%) can be diagnosed as PPCM if they have typical features and all other possible diagnoses have been excluded¹.

There is no single unanimous identified cause of PPCM but several contributing factors have been identified. Recent studies suggest enhanced oxidative stress, cleavage of prolactin and impaired vascular endothelial growth factor (VEGF) signaling are all related with several mechanisms of pathogenesis of PPCM. Oxidative stress acts as a trigger to activate Cathepsin D. Activated Cathepsin D then cleaves prolactin into 2 parts. One part is an angiostatic prolactin fragment while another part is a pro-apoptotic fragment. These fragments play a detrimental role in the pathogenesis of PPCM. In an observational study, development of PPCM was prevented by a Dopamine (D2)receptor agonist bromocriptine. Bromocriptine is a suppressor of prolactin. This further backs the involvement of prolactin in the pathogenesis of PPCM⁴.

Clinical presentation of PPCM is variable and similar to other forms of systolic heart failure. It has a rapid course of development of symptoms. Delayed diagnosis, advanced New York Heart Association (NYHA) classification at diagnosis, left ventricular thrombus, preexisting cardiac diseases are poor prognostic factors⁹⁻¹⁰. Mortality ranges from 6-10% in developed countries³.

Treatment should be started as early as possible and follows the line of treatment of acute decompensated heart failure. Following is a case report of a patient admitted in our ICU with PPCM with a plethora of symptoms.

Case Report

A 26 years old primigravida underwent lower uterine cesarean section (LUCS) in an out side hospital approximately 24 hours

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before presenting to our emergency room complaining of severe restlessness for few hours and low blood pressure. She also had history of two episodes of generalized tonic clonic seizure (GTCS) just 4 hours after LUCS in that hospital. She had no history of any co morbidities like diabetes mellitus or hypertension. Her LUCS was uneventful. Four hours after her LUCS, in the post operative room patient developed GTCS which lasted for around 2 minutes. Another episode of convulsion occurred shortly. She was conscious following her two events of GTCS. Patient became hemodynamically unstable and she was placed on inotrope there for hypotension after insertion of central venous line. She was brought to our emergency room with restlessness and low blood pressure. Soon her blood pressure was found to be non-recordable. Arterial blood gas (ABG) showed type- I respiratory failure.

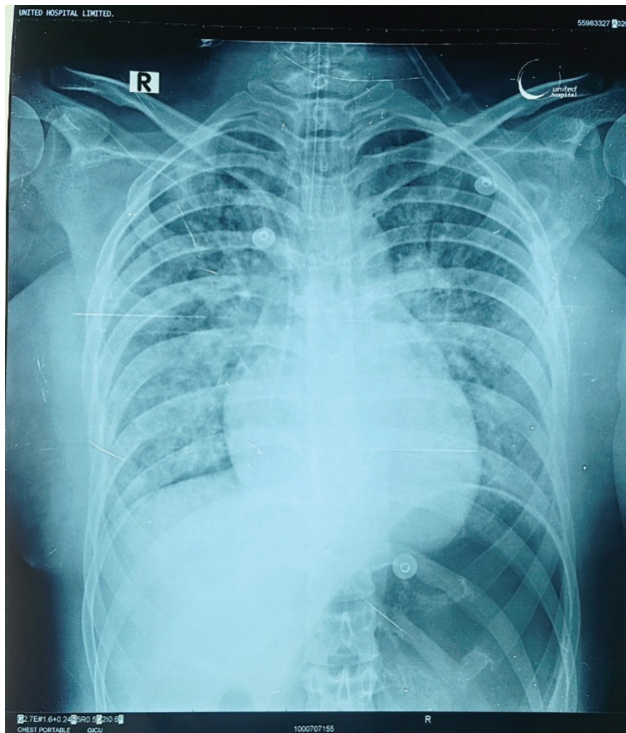


Figure 1: Chest Xray P/A view showing features of pulmonary edema and bilateral pneumonia

Patient was intubated and placed on mechanical ventilator in assist control mode with 100% FiO₂. Chest X-ray was suggestive of bilateral pneumonia and pulmonary edema (Figure-1). ECG showed sinus tachycardia. Non contrast CT scan of head suggested features of mild cerebral edema (Figure-2). Patient was admitted to ICU under department of Critical Care Medicine. Echocardiography was done on the same day (Figure-3). It showed global hypokinesia of LV, severe LV systolic dysfunction with LVEF: 30-35%. Bedside optic nerve sheath diameter was measured with USG, which was slightly raised (Figure-4). Other lab investigations revealed white blood cell count 33.68 X 10³ cmm (77.4% neutrophil), NT-proBNP 10908 pg/ml, HS-Troponin-I 49752 pg/ml, procalcitonin 12.19 ng/ml and slightly altered liver function tests. Serum creatinine and Prothrombin time were within normal limit. Cerebrospinal fluid (CSF) analysis was

done and CSF findings were normal. MRI and MRV of brain was done as well. Reports were unremarkable. Broad spectrum antibiotics were started. Fluid restriction, IV furosemide, I/V mannitol and other supportive managements were applied. She was on intravenous Noradrenaline, and Dobutamine support. Respiratory viral panel, ICT for Dengue and RT PCR for COVID-19 were reported as negative. Mechanical ventilation was continued with appropriate sedation. Patient's condition gradually improved over next 4 days. Patient became hemodynamically stable with minimum inotrope support. Her lab reports gradually became normal. Culture reports (blood, urine, and tracheal aspirate) showed no growth. Patient was extubated after 4 days of ICU admission. Her consciousness level was also improved. Repeat echocardiography after seven days showed global hypokinesia with ejection fraction of 45%. On day eight of ICU admission, patient was shifted to HDU. She subsequently was stepped down to cabin and later discharged from hospital.

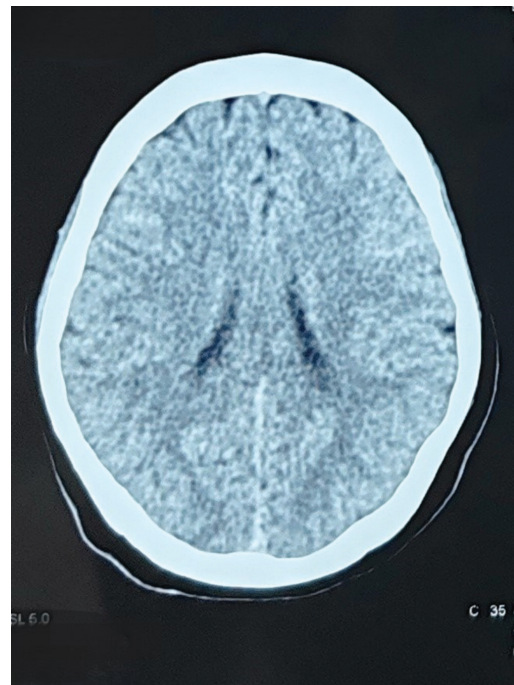


Figure 2: Non-contrast CT scan head (axial view) suggesting mild cerebral edema

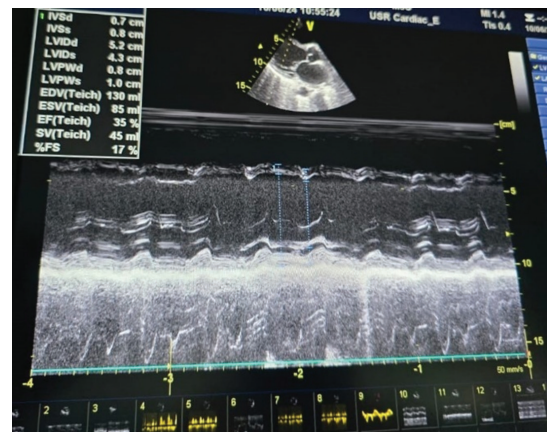


Figure 3 : Bedside echocardiography showing reduced LVEF

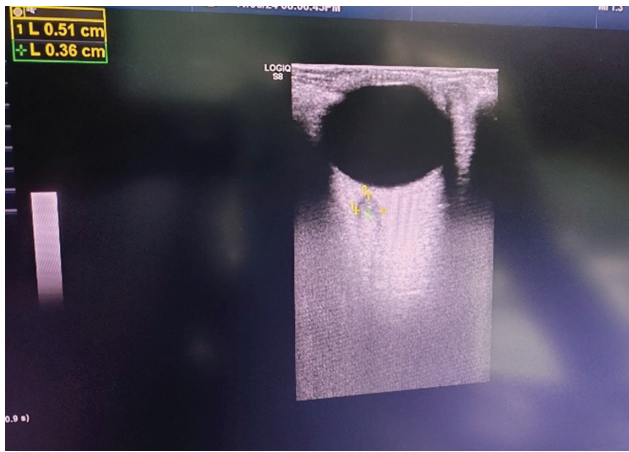


Figure 4: Bedside USG image to measure optic nerve sheath diameter

Discussion

Historically it was thought that PPCM predominantly occurred in older women and black women. But current trend supports that there is gradual increase in incidence of about 24-37% in young primigravida and white women^{3,11}. Table I shows usual presentations of PPCM^{2,6}.

Shortness of breath being a common problem in pregnancy, PPCM is often missed. NTproBNP is often elevated but there is no single diagnostic lab marker for confirmation of PPCM¹¹. However, laboratory studies for exclusion of other diagnoses should also be considered, including serial cardiac enzymes assessment and a pre-eclampsia workup. Chest radiographs can show signs of pulmonary congestion, cardiac enlargement, and even pleural effusions in some cases. Electrocardiographic findings are often normal but can include sinus tachycardia, nonspecific ST- and T-wave abnormalities, and low voltage¹¹.

Table I: Usual presentation of PPCM^{2,6}

Development of Heart Failure	Last month of pregnancy up to 5 months of postpartum
Clinical History	Absence pre-existing heart disease
Risk Factors	Older maternal Age (>30 years) Multiparity (parity>4) Multifetal pregnancy African Descent High blood pressure Prior toxin exposure Use of certain medication to prevent premature labor Maternal cocaine abuse
Causes	Undefined Prior viral illness Abnormal immune response Nutritional deficiencies Coronary artery spasm Small-vessel disease Defective antioxidant defenses Genetic
Arrhythmia	Risk of abnormal heart rhythm
Echocardiographic details	LVED dimension >2.7 cm/m ² M-mode fractional shortening < 30% LVEF <0.45

Note: LVED: left ventricular end diastolic; LVEF: left ventricular ejection fraction

In our case, patient presented with shortness of breath, low blood pressure and two episodes of GTCS. Patient was a young primigravida. Although it is common in older women, trends are now changing. Patient's echocardiography findings, prior negative history of cardiac illness and overall history suggested towards the diagnosis of PPCM. There was associated pneumonia which complicated her clinical condition. Her convulsion and cerebral edema were other significant findings but it resolved soon after treatment and no definite cause could be established. It is hypothesized that

hypotension and hypoxia were predisposing factor for both conditions¹². Patient responded well to symptomatic as well as conservative management.

Treatment of PPCM is similar in line with the treatment of other types of heart failure with LV systolic dysfunction. Although, some cautions are required to ensure safety of unborn/breastfeeding child. Diuretics, beta blockers, hydralazine, digoxin and inotropes are safe during antenatal period. Data regarding use of antithrombotic therapy are inconclusive. So, there is no definite guideline regarding use

of antithrombotic agent now. 2018 European society of cardiology guidelines for the management of cardiovascular diseases during pregnancy suggest that prevention of lactation be considered in patients with severe heart failure because of potential deleterious effect of increased metabolic demand in heart failure¹⁴. Our patient received bromocriptine for lactation suppression later on. She was shifted to cabin after adequate clinical improvement. She was eventually discharged from hospital with advice of follow-up. She and her baby were reported to be in good health.

Conclusion

PPCM is a diagnosis of exclusion. Proper history and suspicion are required to identify such cases. Early initiation of treatment improves the outcome greatly. Nearly 60% patient achieve full recovery of ejection fraction (LVEF >50%)¹³. Weaning of treatment is suggested when patient demonstrate an adequate EF for a consecutive period of six months. All women should receive counselling regarding potential risk of recurrence with future pregnancies. Cardiac transplantation is an option for patients who are resistant to maximal therapy¹⁵.

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