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Case Series on Peripartum Cardiomyopathy

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Absract

Introduction: Peripartum cardiomyopathy is an idiopathic and reversible form of dilated cardiomyopathy. It is defined as systolic heart failure in the last month of pregnancy or within 5 months of delivery. With an unknown etiology it can be fatal in young women but with prompt diagnosis and intensive supportive measures, it can be treated well with good prognosis. Although the prognosis of PPCM is favorable, misdiagnosis or delay in diagnosis leads to deadly consequences to the patient and thus high maternal morbidity and mortality. We present 5 cases of peripartum cardiomyopathy which presented to our hospital in a span of 6 months who were promptly diagnosed, bettter managed with good prognosis in all the patients.Hence high index of suspicion and vigilance required by the treating physician to facilitate timely recognition, intervention, and management of peripartum complications.

Keywords: Peripartum Cardiomyopathy (PPCM); Cardiac Disease in Pregnancy; Dilated Cardiomyopathy; Maternal Mortality.

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Introduction

Peripartum cardiomyopathy is defined as an "idiopathic cardiomyopathy manifested as heart failure due to left ventricular systolic dysfunction towards the end of pregnancy or in the months after delivery when no other cause of heart failure is found" according to European Society of Cardiology Working Group on Peripartum Cardiomyopathy 2010 [1]. It was first described in the year 1800. Its incidence varies from 1 in1300 to 1 in 15000 pregnancies worldwide, highest incidence being in African American women [2].

The etiology of PPCM remains unclear contributing to poor outcome. The risk factors for PPCM appears to be African American ethnicity, older maternal age, pregnancy induced hypertension/preeclampsia, multiparity, multiple gestation, obesity, chronic hypertension and prolonged use of tocolytics [3]. Classically they present with dyspnea, edema, and fatigue; symptoms like in normal pregnancy. Thus, the diagnosis usually is missed or delayed. Also, the symptoms mimic conditions like pulmonary embolism or heart failure due to preeclampsia/eclampsia thus posing a diagnostic dilemma.

The following case reports illustrate both typical and atypical presentation of PPCM. The article also reviews the possible etiologies, symptomatology, management, and prognosis for PPCM to provide patients with an efficient and proper care.

Case Series

Case 1

A 28-year-old G3P2L2 was referred at 35 weeks of gestation with preeclampsia with intrauterine fetal demise with placental abruption. On examination she was found to be tachycardic with blood pressure readings of 150/100mmHg with presence of

pallor. Artificial rupture of membranes was done when she went into active labor with oxytocin infusion. Blood transfusion was started. Patient progressed and delivered one hour after admission a fresh still born male baby with retroplacental clots. Post delivery she had atonic PPH which was medically managed. She was shifted to SICU in view of hypovolemic shock and started on inotropes. Echocardiography was done which was suggestive of features of peripartum cardiomyopathy with mild left ventricular dysfunction with ejection fraction of 45%. She was then started on digoxin and potassium sparing diuretics. Patient was symptomatically better by 24 hours and shifted out from SICU after 72 hours of admission.

Case 2

A 19-year-old lady was referred in view of acute episode of breathlessness following spinal anaesthesia for Emergency LSCS. She had developed abnormal movements with clenching of teeth and profuse frothing from mouth. On examination in our casualty, she was unconscious, not responding to painful stimuli, with feeble pulse rate of 100bpm, blood pressure of 80mmHg systolic, saturation of 40% at room air. There was bilateral coarse crepetitions, with tachypnoea, tachycardia with S_3 gallop. She was immediately intubated to SIMV mode of ventilation and shifted to SICU with 100% PiO_2.

Bedside Echocardiography reported as dilated cardiomyopathy with ejection fraction of 20%. She was started with inotropes and diuretics and stabilized. A diagnosis of cardiac failure secondary to peripartum cardiomyopathy was made and managed accordingly.

Case 3

A 20-year-old female presented to us with history of 9 months of amenorrhea, with labor pains. She also gave history of cough for 2 days. Patient was diagnosed to be preeclamptic and was started on anti-hypertensives 2 days prior to admission. On examination, she was tachycardic with blood pressure readings of 170/130mmHg with 94% saturation and fine crepitations on respiratory examination. Provisional diagnosis of pneumonia was made. She was taken for emergency LSCS for late decelerations. Intra-op she continued to have cough and crepitations and treated as Acute respiratory distress syndrome. Post-op she underwent Echocardiography which revealed peripartum cardiomyopathy with left ventricle and left atrium dilated with diastolic dysfunction and ejection fraction of 35-40%. She was started with ACE inhibitors and diuretics TID. She was symptomatically better and was discharged on POD [14].

Case 4

A 25-year-old patient with 9 months of amenorrhea, presented with acute onset of breathlessness 6 hours prior to admission. She was a primi-gravida with no history suggestive of preeclampsia. There was no palpitation, or chest pain or syncopal attacks. On admission, she was tachycardic, with tachypnoea and high blood pressure readings of 170/100mmHg. Patient was gasping with bilateral coarse repetitions. She was started with inhalation of oxygen, injection of Lasix and deriphylline with hydrocortisone. In view of worsening crepitations, patient was intubated with SIMV mode of ventilator.

Provisional diagnosis of accelerated hypertension with dilated cardiomyopathy with left ventricular failure was made and was started on inotropes and Lasix. Echocardiography showed dilated cardiomyopathy with ejection fraction of 20%. She was induced and delivered by forceps assisted vaginal delivery 12 hours after induction. Patient continued to be unconscious on ventilator and inotropes.

On PND_2 patient was conscious, was changed from SIMV to CPAP and slowly inotropes were tapered. She was started on ACE inhibitors and diuretics with digoxin in view of peripartum cardiomyopathy. Patient was extubated the next day and was continued with same treatment. Patient recovered well and was continued with Furosemide and asked to review after 3 months.

Case 5

A 30-year-old P_3L_3 postnatal immediate presented with postpartum hemorrhage and hypovolemic shock. On examination, patient was breathless, with tachycardic, tachypneic, hypotensive with blood pressure being 90mmHg systolic with saturation being 84%. She was started on oxygen inhalation, PPH medically managed, blood transfusion started and shifted to SICU. There she was started on inotropes.

PPH was controlled. Patient was symptomatically better, but she continued to be hypotensive. She was evaluated further, and Echocardiography report suggested peripartum cardiomyopathy with ejection fraction of 40% with diastolic dysfunction. She was then started with diuretics, digoxin, and ACE inhibitors. Slowly inotropes were tapered. Patient improved, shifted out of SICU, and was discharged on PND [13].

Discussion

Peripartum cardiomyopathy is associated with 1 in 3000-4000 live births. It is a rare form of dilated cardiomyopathy with unknown etiology. The relationship between pregnancy with dilated cardiomyopathy was recognized in 1870's and was further classified as a separate entity in 1930 [4]. There have been proposed mechanism of the disease which include viral etiology, autoimmune, increased prolactin, cardiovascular stress of pregnancy, inflammatory response in pregnancy- elevation of TNFα and IL-6 and nutritional deficiency of selenium [5,6].

Diagnosis of PPCM must fulfil all 4 criteria. All 4 of the following [5]

❖ **Classic**

- Development of cardiac failure in the last month of pregnancy or within the first 5 months of postpartum.
- Absence of any determinable etiology for the cardiac failure.
- Absence of any known prior heart disease.

❖ **Additional**

- Left ventricular systolic dysfunction with depressed ejection fraction.
- Ejection fraction ≤ 45%
- Fractional shortening(FS) <30%
- End-diastolic dimension EOD >2.7cm/m^2

❖ **Diagnostic Testing of PPCM:**

- Complete family history to identify familial association.
- Serum tests.
- Complete blood cell count with differential counts.
- Creatinine and urea levels
- Electrolyte levels, including Ca and Mg.
- Cardiac enzymes, including troponin.
- Level of BNP and N terminal pro B type natritic protein.
- LFT, TSH levels.
- Chest radiograph.
- ECG.
- Transthoracic echocardiogram
- Cardiac MRI and or endomyocardial biopsy.

The initial symptoms of PPCM are like that of heart failure i.e. fatigue, shortness of breath, cough, chest pain, palpitations, tachycardia, increased blood pressure readings, fluid retention, pedal edema. As there is overlap of symptoms with those of pregnancy especially in the last trimester of pregnancy or after delivery and thus the diagnosis is either delayed or missed [7]. In our case, all patients had variale symptoms and clinical scenario. But 3 out of 5 patients had preeclampsia. So, preeclampsia may be one of the commonest predisposing factor.

Further testing is required to establish the presence of cardiac failure. Chest X-ray demonstrates cardiomegaly and pulmonary edema. Echocardiography confirms ventricular failure with increased left ventricular end diastolic dimensions, decreased ejection fraction with diastolic dysfunction [8]. Invasive evaluation like cardiac catheterization, endomyocardial biopsy is often not needed for confirmation. Inflammation with myocarditis is present in up to 50% of specimens.

Coordinated management with specialists (obstetrician, physician, cardiologist, pediatrician) is required with fetal heart monitoring. ACE inhibitors and ARBs are main treatment for postpartum women with heart failure but are contraindicated in pregnancy due to their teratogenic effects. Digoxin, beta blockers, loop diuretics and drugs reducing after-load such as hydralazine and nitrates are proven to be safe and are mainstay drugs in the medical management [8]. Beta blockers are recommended for peripartum cardiomyopathy as they improve symptoms, ejection fraction and survival. Anti-coagulation treatment is recommended for those with severe left ventricular dysfunction or documented cardiac thrombosis [8,9]. Newer ventures into this field has brought up pentoxifylline drug to be helpful to improve outcome, left ventricular functions in small prospective studies [8,9]. Bromocriptine therapy has been used as a modality those of which had shown increased prolactin delivery.

Vaginal delivery is preferred mode as it is associated with lower rate of complications unless the mother is decompensating. If patient is non-responsive to medical management and must be delivered, then induction of labor can be done to achieve vaginal delivery.

Caesarean delivery is associated with post-operative third spacing of fluid which leads to increased intravascular volume and decompensation [9].

Cardiac transplantation and left ventricular assist devices for patients with progressive left ventricular dysfunction have been tried [9].

Prognosis depends on recovery of left ventricular function. Recovery usually occurs between 3-6 months postpartum. Mortality ranges from 7-50%. Cause of death are progressive heart failure, arrhythmia, thromboembolism. If the abnormal ejection fraction continues, the life span of the patient is shortened, and future pregnancy is not recommended. According to survey, 70% of women with recovered left ventricular function have a normal outcome [9]. In all our cases timely diagnosis and proper management helped to have good prognosis with no mortality.

Conclusion

Peripartum cardiomyopathy though a rare disease (0.1% of pregnancies) can lead to devastating consequences with high morbidity and mortality if not diagnosed and treated early [5,6]. The cases presented here demonstrate the variability of clinical presentation of PPCM and the early diagnosis and management respectively of each case. As preeclampsia is a common condition seen, it can be taken as a predisposing factor. Treatment includes ACE inhibitors, ARBs, Beta blockers digoxin and Diuretics [3]. Careful assessment of risk factors, causative agents helps in early diagnosis of PPCM. Hence there is requirement of high index of suspicion when managing a dyspneic patients to determine possible early interventions and have better pregnancy and postpartum outcome [2].

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