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Case Report

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Peripartum cardiomyopathy with preeclampsia in a parturient: A case report with literature review

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Abstract:

Peripartum cardiomyopathy (PPCM) is a rare disease of unknown cause that affects women of childbearing age. A high index of suspicion should be maintained in the pregnant and peripartum woman who presents with sudden cardiac decompensation without any prior history of cardiac disease. The diagnosis can be confirmed with echocardiographic evidence of global left ventricular dysfunction. Timely diagnosis and institution of therapy for heart failure can avoid adverse outcomes in a parturient with PPCM. In this case report, we describe the management of primigravida presenting to the hospital's emergency department with acute cardiac failure and respiratory distress due to PPCM. The case also highlights that though preeclampsia and PPCM are two separate entities, these can coexist in the same parturient due to the common pathophysiological mechanism. In the review, the recommended medical management of heart failure in PPCM with the "BOARD" (Bromocriptine, Oral heart failure drugs, Anticoagulants, Vasorelaxing agents, and Diuretics) scheme is discussed.

Keywords:

Echocardiography, parturient, peripartum cardiomyopathy, preeclampsia, respiratory distress

Introduction

Peripartum cardiomyopathy (PPCM) is rare and life-threatening cardiomyopathy of unknown etiology that affects women in the last month of pregnancy or within the first 5 months of the postpartum period. The incidence of PPCM varies widely from 1:100 to 1:10,000 live births depending on the ethnic, racial, and regional background of women.[1] Multiparity, black race, advanced maternal age, multiple gestation, selenium deficiency, and preeclampsia are the risk factors predisposing to PPCM.[2] The diagnosis of PPCM is based on the guidelines by the National Heart, Lung, and Blood Institute which includes (1) new-onset cardiac failure in the last month of pregnancy or within 5 months of delivery, (2) absence of an identifiable cause for the cardiac failure, (3) absence of preexisting cardiac disease, and (4) left ventricular systolic dysfunction with decreased ejection fraction (EF) of <45%.[3]

Despite growing literature, mortality due to PPCM remains high (30%–60%) and survivors have a high risk (50%–80%) of developing cardiac failure during future pregnancies. Patients of PPCM present with symptoms such as fatigue, edema, and dyspnea which can even be seen in late normal pregnancy, other pregnancy-related comorbidities such as pulmonary embolism, and other causes of heart failures such as preexisting heart disease, toxic cardiomyopathy, and

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Takotsubo syndrome.[4] Therefore, the diagnosis of PPCM is often delayed, and this disorder is commonly unrecognized resulting in unfavorable prognosis for both the mother and fetus.

Case Report

A 22-year-old primigravida with 35-week period of gestation presented in the emergency department (ED) with complaints of breathlessness for 1 day and progressive orthopnea for 2 days. At the time of admission, her pulse rate was 140 beats/min, blood pressure was 170/110 mmHg, respiratory rate was 32/min, and Sp02 (oxygen saturation) of 66% with oxygen on a face mask. On random midstream urine dipstick test, her urine protein was 3+ (suggestive of proteinuria). On general examination, the parturient had dyspnea and tachypnea, neck veins were distended, and bilateral Grade 2 pedal edema was present. On auscultation, bilateral rales with a gallop heart sounds were heard and fetal heart sounds were absent. Blood gas analysis showed severe hypoxemia with metabolic acidosis (pH – 7.25, paO_2 – 35.3, $PaCO_2$ – 46.4, HCO_3 – 15, and SaO2 – 75%). Her medical records showed no previous history of heart disease, and her antenatal visits had been unremarkable. The patient's presenting complaints and examination indicated the presence of a cardiovascular system problem with severe preeclampsia.

In the ED, immediate medical management was started with injection furosemide 20 mg IV (to decrease preload and pulmonary congestion), nitroglycerin infusion (to control the raised blood pressure and reducing afterload), and noninvasive ventilatory (NIV) support. Her chest roentgenogram showed enlarged cardiac shadow with pulmonary edema. Thereafter, she was shifted to the intensive care unit (ICU) due to clinical deterioration, as there was little improvement in the blood gas parameters with NIV support, she was intubated and put on invasive ventilation with low tidal volume (350 ml), high positive end-expiratory pressure (12 cm H_20), and high FiO_2 (0.8–1.0) ventilator settings. Bedside echocardiography was done immediately on arrival at the ICU and showed global dilatation of all heart chambers with severe hypokinesia and reduced EF of 25%, lung ultrasound showed lung rockets indicating pulmonary edema, and abdominal ultrasound showed absent fetal cardiac activity. With the above findings, she was diagnosed as a case of PPCM with severe preeclampsia with intrauterine fetal demise.

The parturient blood pressure dropped to 90/47 mmHg, and for initial stabilization of blood pressure, vasopressor support with norepinephrine infusion was started. Invasive arterial line and central venous line were inserted to guide fluid therapy. The patient's initial laboratory investigations were normal except for raised leukocyte count of 18,000/mm^3, highly elevated pro-brain natriuretic peptide (BNP) 27,395 pg/ml (normal range <133 pg/ml), and elevated 24 h urine protein.

On day 2, norepinephrine infusion was slowly tapered off and furosemide infusion was started to decongest the lungs; she had a spontaneous vaginal delivery. On day 3, there was an improvement in blood gas parameters (pH 7.4, pO2 – 78, PCO2 – 28, HCO3 – 20, and SaO2 – 95%), and the parturient was weaned off the ventilator and extubated. She was given salt-restricted diet, tablet carvedilol 6.25 mg twice daily, tablet ivabradine 5 mg twice daily, tablet furosemide 20 mg twice daily, tablet bromocriptine 2.5 mg twice daily, oral potassium chloride, and injection dalteparin 5000 units subcutaneous once daily with intermittent NIV ventilatory support. Echocardiography was repeated after 1 week and showed a persistent low EF of 30% with global hypokinesia. The parturient was transferred to the ward for the continuation of medical treatment; 1 week later, she was discharged to home with advice to follow-up in the cardiac outpatient department. Follow-up echocardiography at 6 months showed recovery of EF to 45%.

Discussion

PPCM is a rare disorder of unclear etiology. The postulated mechanism is a two-hit theory which has host susceptibility (genetic predisposition, hypertension, and selenium deficiency) superimposed by systemic angiogenic imbalance with high levels of prolactin and soluble tyrosine kinase-1.[5,6] The host predisposing factors include multiparity, elderly gravida, family history, African–Asian ethnicity, smoking, diabetes, hypertensive disorder of pregnancy, malnutrition, prolonged use of tocolytics, genetic predisposition, and viral infections. Oxidative stress during pregnancy leads to proteolytic cleavage of prolactin into its 16 kDa fragment which is cardiotoxic and impairs cardiomyocyte contractibility. It has also been found that hypertensive disorders of pregnancy frequently coexist with PPCM; elevated levels of antiangiogenic factors such as soluble tyrosine kinase 1 related to the pathogenesis of preeclampsia are also found to be elevated in patients with PPCM, thus suggesting a common etiological pathway for the two conditions.[7] PPCM with concomitant preeclampsia is associated with increased morbidity and mortality.

The differential diagnosis of PPCM includes heart failure from other causes such as dilated cardiomyopathy, adult congenital heart disease, valvular heart disease, toxic cardiomyopathy (viral, chemotherapy), Takotsubo

syndrome, hypertensive heart disease, and severe preeclampsia. In Pregnant patients presenting with acute cardiac decompensation; other causes of cardiogenic shock including severe preeclampsia, pulmonary embolism, amniotic fluid embolism, and myocardial infarction should be ruled out.[1] Although preeclampsia and PPCM often coexist, the clinical differentiating features of heart failure due to preeclampsia include the presence of echocardiographic finding of diastolic dysfunction and left ventricular hypertrophy with the previous history of hypertension or preeclampsia.[8]

An electrocardiogram (ECG) should be performed in all parturients with suspected PPCM, although it is not specific; it helps to distinguish PPCM from other causes of heart failure. Electrocardiographic findings can include sinus tachycardia, nonspecific ST and T wave abnormalities, and voltage abnormalities. Prolonged QTc interval and tachycardia are associated with poor outcomes in PPCM.[9] The diagnosis of PPCM is confirmed by echocardiography findings of left ventricular EF <45% or M-mode fractional shortening <30% (or both) and end-diastolic dimension >2.7 cm/m^2. Cardiac protein assays of BNP or N-terminal proBNP (NT-proBNP) are consistently elevated in PPCM, the cutoff values being 100 pg/ml and 300 pg/ml, respectively, for BNP and NT-pro BNP for acute heart failure.[10] Chest radiographs may show signs of pulmonary congestion, cardiac enlargement, and pleural effusion.

All parturients with cardiogenic shock, respiratory insufficiency, or hemodynamic instability should be managed in an ICU with facility for advanced cardiac support. They should be given supplemental oxygen and should have continuous hemodynamic monitoring with pulse oximetry, invasive blood pressure, and ECG. For those needing intubation and invasive ventilation, rapid sequence intubation can be done using a titrated dose of anesthetic medications. As parturients have a higher oxygen consumption and a significantly lower functional residual capacity, they desaturate more rapidly during a period of apnea. Furthermore, there is a risk of sudden decompensation and cardiac arrest from cardiodepressant effects of anesthetic drugs or sudden rise in systemic vascular resistance during endotracheal intubation. Norepinephrine or levosimendan should be used for hemodynamic support in cardiogenic shock, whereas β-adrenergic agonist drugs are preferably avoided due to increased sensitivity of the heart to its toxic effects. Parturient with severely reduced EF might also need implantation of ventricular assist devices or cardiac transplantation.

Treatment with the "BOARD" scheme (Bromocriptine, Oral heart failure drugs, Anticoagulants, Vasorelaxing agents, and Diuretics) is recommended for PPCM. Bromocriptine is the new disease-specific therapy for PPCM as it inhibits prolactin secretion and release of its cardiotoxic fragments. Bromocriptine therapy has been found to improve cardiac recovery and reduce the mortality in PPCM patients.[11,12]

The medical management of patients with PPCM is similar to that for other forms of heart failure, and treatment aims to reduce afterload, preload, and to increase contractility.

Pharmacological treatment of heart failure includes β-blockers, angiotensin-converting enzyme (ACE) inhibitors, angiotensin receptor blockers (ARBs), mineralocorticoid receptor blockers (MRBs), ivabradine, vasodilators (nitrate and hydralazine), digoxin, and diuretics. Out of these, ACE inhibitor, ARBs, MRBs, and ivabradine have fetal toxicity and thus contraindicated in pregnancy.

Treatment with β-blockers is indicated in PPCM in conjunction with diuretics for pulmonary congestion and vasodilators (hydralazine and nitrates) to decrease systemic afterload.

However, β-blockers should not be initiated in the acute decompensated phase of heart failure. In lactating parturient, cautious use of β-blockers, ACE inhibitors, and MRBs is suggested due to minimal secretion of these drugs in breast milk. Digoxin, an inotropic agent, is safe during pregnancy and may help to maximize contractility and rate control, but serum level monitoring will be needed. It is recommended to continue the treatment for heart failure until complete recovery of cardiac functions and for at least 12–24 months after complete recovery. PPCM is associated with a high incidence of thromboembolism; therefore, prophylaxis with anticoagulant (low-molecular-weight heparin or oral warfarin) is recommended, especially when EF is reduced (<35%).[13] Delivery of the fetus reduces the hemodynamic stress on the heart, and the mode of delivery is based on obstetric indications.

The prognosis of a parturient with PPCM depends on the normalization of left ventricular size and contractility within 6 months after delivery. Despite complete normalization of left ventricular EF (LVEF) in PPCM associated with preeclampsia, these patients are at high risk for recurrent hospitalization or death. About half of the patients of PPCM recover without any complications. Regardless of recovery, a second pregnancy is not recommended in these patients due to the high recurrence rate (30%–50%). The persistence of cardiac dysfunction 6–12 months after the initial diagnosis of PPCM usually indicates an irreversible problem. Implantation of an implantable cardioverter-defibrillator

and cardiac resynchronization therapy is recommended in patients with severe persistent cardiac dysfunction despite optimal medical therapy.[14]

Conclusion

PPCM is a rare disease of unknown cause affecting women of the childbearing age. Diagnosis should be based on clinical suspicion and confirmed with echocardiographic evidence of left ventricular dysfunction. Treatment under the "BOARD" scheme is suggested for patients with PPCM. With timely diagnosis and appropriate treatment, morbidity and mortality may be reduced in PPCM.

Author contributions

Literature search and manuscript preparation was done by NK. Concept, design, manuscript editing, and final review of the manuscript was done by DS.

Divya Sethi takes responsibility for the integrity of the work and acts as a guarantor.

Conflicts of interest

None declared.

Consent to participate

The author certifies that they have obtained all appropriate patient consent forms. The patient has given her consent for clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal the identity.

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