CASE REPORTS

Peripartum Cardiomyopathy: A Case Series

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Abstract:
Peripartum cardiomyopathy (PPCM) is a life threatening entity of peripartum period characterized by left ventricular systolic dysfunction and heart failure in absence of any known cardiac disease. Though its incidence is rising but there is still uncertainty regarding its incidence, pathogenesis, and optimum management protocol. Retrospective analysis of twenty cases of PPCM admitted, within July 2016 to June 2018 in the department of Cardiology, Bangabandhu Sheikh Mujib Medical University (BSMMU), Dhaka is done here. Mean age of study population was 25 (6.3). Majority of the patients were multi-paras 12 (60%). 80% of the patients were diagnosed after delivery. Breathlessness was the most common symptom. Bibasal lung base crepitations was most frequent sign, which was found in 95% cases. Sinus tachycardia was most frequent electrocardiographic change which was found in 80% cases. Mean left ventricular ejection fraction (LVEF) was 35.2(2.8). All the patients had different extent of global hypokinesia of left ventricle (LV) at rest. The patients were treated with bed rest, water and salt restriction, loop diuretic, digitalis, selective Beta-blocker, vitamin B complex and an anticoagulant in relevant cases in antepartum period, Angiotensin converting enzyme inhibitor (ACEI) or Angiotensin receptor blocker (ARB) was added in postpartum period. 70% patients were clinically improved and in 45% the left ventricular functional status returned to normal. 30% developed persistent cardiomyopathy beyond six months of presentation. Maternal mortality was 1 (5%). Among all live births three had intra uterine growth retardation. The pathophysiology, emerging investigations modalities, updated management protocol, and prognosis of PPCM are also discussed in this review.

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Introduction:
Peripartum cardiomyopathy (PPCM) is an idiopathic form of cardiomyopathy where heart failure (HF) occurs due to impairment of left ventricular (LV) systolic function in the absence of any determinable heart disease in the last month of pregnancy or during the first 5 months postpartum. The European Society of Cardiology (ESC) Working Group on PPCM has set the diagnostic criteria. In presence of heart failure, LV ejection fraction should be less than 45% and LV may not be overtly dilated. Though right ventricular dysfunction is not included in the diagnostic criteria but its presence signifies poor prognosis. As it is a cause of significant mortality and morbidity among young woman throughout the world, so he have reported 20 cases of PPCM from our setting.

Case reports
We evaluated the clinical profile, management and analyzed the pregnancy outcomes of PPCM. Follow up was done after treatment to see the prognosis. A total of 20 consecutive patients who got admitted with PPCM from July 20016 to June 2018 in the department of Cardiology, Bangabandhu Sheikh Mujib Medical University (BSMMU), Dhaka, Bangladesh were considered for analysis.

Mean age of study population was 26 (6.3). Majority of the patients were multi-paras 12 (60%). 80% of the patients were diagnosed after delivery and 20% of the patients were diagnosed prior to delivery. Breathlessness was the most common symptom which was present in all of the cases, and palpitation was present in 80% cases. Bibasal lung base crepitations was most frequent sign, which was found in 95% cases. Tachycardia and pedal edema were found in 90% and 60% cases. Anemia was found in 60% cases. 45% of the study population were hypertensive, among them 70% of cases had gestational hypertension.

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Sinus tachycardia was most frequent electrocardiographic change which was found in 80% cases, followed by nonspecific T wave changes in 60% cases. Radiological evidence of pulmonary edema was found in all cases. Detailed echocardiographic evaluations were done to exclude the differentials, and to confirm the diagnosis. Mean LVEF was 35.2(2.8). All the patients had different extent of global hypokinesia of LV at rest. 25% of the patients had evidence of diastolic dysfunction of LV. Only 20% of the patients had right ventricular systolic dysfunction as evidenced by reduced tricuspid annular plane systolic excursion (TAPSE). 40% of the patients had mild to moderate mitral regurgitation, whereas 30% patients had mild to moderate tricuspid regurgitation.

We treat the patients with bed rest, water and salt restriction, loop diuretic, digitalis, selective Beta-blocker, vitamin B complex and an anticoagulant in relevant cases in antepartum period, and Angiotensin converting enzyme inhibitor (ACEI) or Angiotensin receptor blocker (ARB) was added in postpartum period. Comorbidities like anemia and hypertension were managed accordingly. 70% patients were clinically improved and in 45% the left ventricular functional status returned to normal with the above mentioned management. 30% developed persistent cardiomyopathy beyond six months of presentation. Maternal mortality was 1 (5%). Among all live births three had intra uterine growth retardation Regular clinical follow up with echocardiography were ensured for up to six months.

Discussion:
Epidemiology
The incidence and prevalence of PPCM have varied widely across different part of the world with highest incidence in north-south gradient, ranging from about one per 300 in Haiti, one per 1000 live births in South Africa and up to one per 3000 live births in the US and Western Europe. Genetic and environmental factors, lack of uniformity in diagnostic criteria, cultural and puerpertal practices may be responsible for these heterogeneity. The data regarding incidence and prevalence of PPCM in Bangladesh is lacking.

Pathogenesis and contributing factors
The pathophysiology of PPCM is not clearly elucidated. The identifiable contributing factors are black ethnicity, advanced maternal age, obesity, multifetal pregnancy, prolonged use of tocolytics and history of hypertensive disorders of pregnancy. Oxidative stress–prolactin axis hypotheses, angiogenic imbalance, viral myocarditis, abnormal response to hemodynamic stress of pregnancy, immune mediated injury, genetic predisposition, micronutrient deficiency are also some probable pathophysiologic mechanism.7-13 Oxidative stress–prolactin axis hypothesis suggests oxidative stress activates lysosomal enzyme, cathepsin D that in turn cleaves serum prolactin into its antiangiogenic and proapoptotic 16-kDa prolactin sub fragment. This subunit may be responsible for microvascular dysfunction and cardiac injury.

Diagnosis
Clinical presentation
High index of clinical suspicion is required as it’s a diagnosis of exclusion. Symptoms of PPCM (dyspnea, orthopnea, oedema, palpitations) may be found in normal pregnancy so late presentation with complications like ventricular arrhythmias, venous or arterial emboli are not infrequent.14 Approximately 75% of cases are diagnosed within the first month after delivery, and 45% occur in the first week.15 Physical examinations may reveal signs of heart failure, including tachycardia, hypotension, elevated jugular venous pulsation, peripheral edema, and pulmonary crackles. Signs of LV dilatation like displaced apical impulse, third heart sound may be present but not so frequent.

Electrocardiography
Sinus tachycardia with nonspecific changes are usual findings. Bundle branch block, T-wave changes (59%), P-wave abnormality (29%), QRS-axis deviation (25%) were found in a case series.17

Echocardiography
It is the single most important tool to diagnose PPCM, exclude the differentials, and find out the complications like embolism and pericardial effusion. It usually demonstrates global hypokinesia of LV with impairment of systolic function(LVEF<45%). LV dilatation is not mandatory.1 Diastolic dysfunction of LV may also predominate sometimes.18 Right ventricle is frequently affected though it’s not included in the diagnostic criteria. Functional regurgitation involving mitral and tricuspid valve may be found along with pulmonary hypertension. New echocardiographic modalities like speckle tracking is not validated in PPCM.1

Chest X ray
Evidence of pulmonary edema, cardiomegaly and pleural effusion are usually found.
Biomarkers
B type natriuretic peptides are increased in acute stage which reflects increased end diastolic pressure.

Novel biomarkers
Few potential novel biomarkers that need to be validated in near future are combination of cathepsin D, miR-146a, ratio VEGF/sFlt1 and serum asymmetric dimethylarginine (ADMA), a marker of endothelial dysfunction.

Cardiac magnetic resonance imaging (MRI)
CMRI can exclude other form of cardiomyopathy and detect LV volumes and function more precisely but cannot predict LV function recovery.

Endomyocardial biopsy
Endomyocardial biopsy is not currently recommended.

Differential Diagnoses
As it is a diagnosis of exclusion so more common causes of heart failure like rheumatic heart disease, myocarditis, dilated cardiomyopathy from other causes, coronary artery disease must be excluded prior to diagnosis.

Management
Acute phase
Treatment is focused on controlling volume status, counteracting maladaptive neurohormonal response and preventing complications like thromboembolism and arrhythmias. Concomitant comorbidities like anemia and hypertension should be managed appropriately. In volume overloaded patients, salt and fluid should be restricted. Judicious use of diuretics should be ensured during pregnancy as there is risk of placental hypoperfusion. If systolic blood pressure allows then intravenous vasodilators like hydralazine and nitroglycerine may be considered. Angiotensin converting enzyme inhibitors and angiotensin receptors blockers should be avoided during pregnancy due to fetotoxicity.

Enalapril, captopril and benazepril are preferred during breastfeeding. Beta blockers preferably metoprolol can be used throughout the entire period after stabilization of acute heart failure. In our center, digoxin is being used along with standard treatment with reasonably good outcome.

In case of low cardiac output syndrome, inotropes like dobutamine and levosimendan may be used. If the patient does not response to the medical management then mechanical circulatory support in the form of intra-aortic balloon counterpulsation (IABP) is instituted. In case of multiorgan dysfunction syndrome and non-responder to IABP is managed with Extracorporeal membrane oxygenation (ECMO) as a bridge to recovery, to LV assist device (LVAD) implantation or to heart transplantation.

Patient should be anticoagulated and continued for at least two months postpartum, if LVEF<35% or other indications for anticoagulations are present.

Targeted therapy
Bromocriptine, an ergot alkaloid and dopamine D2-receptor antagonist has emerged as a potential useful treatment for PPCM. Due to lack of consistent results in the trials and risk of thromboembolic complications its routine use is currently limited. Immunomodulation by pentoxifylline and intravenous immunoglobulin have failed to offer any benefit.

Long term management
As clinical course is variable and LVEF recovery may take up to six months in the majority of patients invasive therapies (cardiac resynchronization therapy and/or intracardiac defibrillator (ICD) implantation) may be wisely postponed for up to six months. However, in case of increased risk of sudden cardiac death, subcutaneous ICD or wearable external defibrillator may be considered immediately.

In advanced and refractory HF or in whom LVAD weaning fails, heart transplantation is the only therapeutic option though the prognosis is variable.

Treatment duration
There is no general agreement, for how long the treatment should be continued. ESC working group on PPCM has advocated to continue ACE inhibitors and beta blockers for at least one year but decision should be individualized based on clinical and echocardiographic recovery.

Obstetric management
Delivery
There are limited data available to support optimum timing and mode of delivery in PPCM and decision should be taken by a team consisting of cardiologists and obstetricians. Strategy and timing depend upon hemodynamic status of mother, fetal maturity and obstetrical factors. In case of hemodynamic instability, urgent delivery should be targeted preferably by caesarean section, irrespective of gestational age. Vaginal delivery is preferred in stable cases as it is associated with less blood loss, less thromboembolic and infectious complications.
Pain control
Epidural analgesia is preferred.1

Breast Feeding
Though breast feeding avoidance may be theoretically beneficial according to oxidative stress-prolactin axis hypothesis but it is not evidence based. Rather recovery rate is higher in lactating mother challenging this hypothesis.27

Subsequent pregnancies
Recurrence risk of PPCM depends upon extent of LV recovery. Patients with full recovery (LVEF>55%) have 17% chance of recurrent failure whereas who recover incompletely (LVEF<55%) have 46.2% risk of heart failure in subsequent pregnancies.28 So, family planning counselling is of paramount importance. Dobutamine stress echocardiography can be used to risk stratify women with subnormal LVEF.29

Predictors of recovery and prognosis
Improvement of LVEF>50% at six months is defined as recovery from PPCM.1 In our cases, 70% patients improved clinically, 30% developed persistent cardiomyopathy, and 5% died.

LVEF and LV dimensions are the best predictors of recovery.16 Lower level of plasma troponin and brain natriuretic peptide, diagnosis made after delivery, breast feeding, non-African ethnicity favorably affect the outcome.16,27

Conclusion:
Diagnosis of peripartum cardiomyopathy requires increased awareness among multidisciplinary patient care teams and a high degree of suspicion. Management of peripartum cardiomyopathy should aim first at improving heart-failure symptoms through conventional therapies, and then consideration of targeted therapies. Though the chance of recovery is high but chance of relapse is also not less in subsequent pregnancies.

References


