Case Report

Peripartum Cardiomyopathy

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Abstract

Peripartum Cardiomyopathy (PPCM) is an idiopathic cardiomyopathy that presents with heart failure secondary to left ventricular systolic dysfunction toward the end of pregnancy or in the months after delivery. Incidence varies from 1:300 to 1:15000 pregnancies. Causes and pathogenesis are poorly understood. Clinical presentation includes signs and symptoms of heart failure. PPCM remains a major cause of maternal morbidity and mortality. Many cases of PPCM improve or resolve completely but others progress to heart failure; as early diagnosis and medical treatment may affect the patients long term prognosis. We are reporting the case of Peripartum Cardiomyopathy diagnosed in the puerperal period.

Keywords: Peripartum Cardiomyopathy, idiopathic cardiomyopathy, heart failure, pregnancy

Introduction

Peripartum Cardiomyopathy (PPCM)is a rare and potentially life threatening form of heart failure affecting women late in pregnancy or in the first months after delivery often complicating Obstetric and Anaesthetic management.[1] The Heart Failure Association of the European Society of Cardiology Working Group on PPCM has defined it as an idiopathic cardiomyopathy presenting with heart failure secondary to left ventricular (LV) systolic dysfunction towards the end of pregnancy or in the months following delivery, where no other cause of heart failure is found.[2] It is a diagnosis of exclusion. The LV may not be dilated but the ejection fraction is nearly always reduced below 45%. The echocardiographic criteria proposed for LV dysfunction are as follows:[3]

- Ejection fraction <45% or fractional shortening <30%
- End diastolic dimension >2.7cm/m^2

The etiology of PPCM is still unknown and many potential causes have been proposed but not conclusively proven. These include viral myocarditis, abnormal immune response to pregnancy, abnormal response to increased hemodynamic burden of pregnancy, hormonal abnormalities, malnutrition, inflammation and apoptosis. New experimental animal models indicate the role of a novel molecular pathogenetic mechanism involving a cardiomyoyte specific deletion of the transcription factor signal transducer and activator of transcription (STAT3) protein.[4] Experimental evidence has implicated abnormal prolactin metabolism as fundamental in the pathogenesis of PPCM and blockade of prolactin by bromocriptine has been shown to prevent PPCM.[4] It maybe caused by systemic angiogenic imbalance.[5,6,7]

The diagnosis of PPCM presents a challenge as its presentation may mimic other causes of systolic heart failure or even exaggerated signs and symptoms of late pregnancy and early puerperium. It is possible to misdiagnose PPCM for pulmonary embolism. An error in diagnosis is life threatening for the patient with a potential for fatal outcome in about 20%.

It is imperative to maintain a high index of suspicion with attention to timing of symptoms and perform thorough investigation to rule out other causes of heart failure.

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Case report

This is a case report of a 27 year old female who presented to the labour room at 38 weeks of gestation in her first pregnancy with swelling of both lower limbs since 15 days. At the time of admission, her blood pressure was 160/110mm Hg, with grade 3 edema and proteinuria. She was diagnosed as having Severe pre eclampsia.

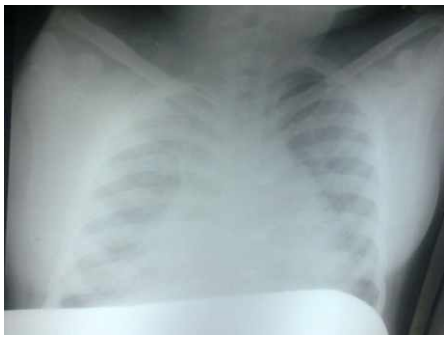

Figure 1: Chest X Ray showing grossly dilated cardia

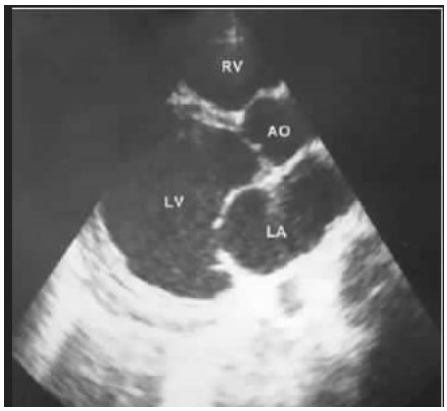

Figure 2: Transthoracic echocardiography showing dilated left atria and ventricle

Her complete blood count, liver function tests, renal function tests, uric acid, coagulation profile were all within normal limits. Antepartum fetal surveillance was non reassuring and she was taken up for caesarean section under spinal anaesthesia. Indication was fetal distress with severe pre eclampsia. A healthy male baby was delivered. Intra operative period was uneventful.

On post operative day 2, she developed acute onset breathlessness, her pulse was 116/min, Blood pressure was 130/80mm Hg, respiratory rate was 28 cycles/min, she was afebrile, her oxygen saturation was 80% at room air. On auscultation, there were bilateral coarse crepitations. Her chest X ray showed cardiomegaly. (Figure 1) ECG showed left axis deviation. Transthoracic echocardiography (Figure 2) showed hypokinesia of left ventricle with decreased left ventricular systolic function, ejection fraction of 38% and left ventricular end diastolic dimension 52 mm and was diagnosed as a case of Peripartum Cardiomyopathy.

She was shifted to medicine ICU and given ventilator support in form of continuous positive airway pressure, she was managed with intravenous furosemide, bromocriptine and metoprolol.

Ventilator assistance was continued for 48 hours after which she was extubated.

She was discharged on post operative day 12 during which her pulse rate had improved to 80 beats per minute, respiratory rate was 18 cycles/min, blood pressure was 120/80mm Hg. Chest was clear and Cardiovascular system was normal. She was adviced to continue oral furosemide 20mg once daily and oral metoprolol 12.5 mg once daily for 2 weeks.

At 6 months follow up she was asymptomatic, her ejection fraction had improved to 67%.

Discussion

PPCM is a rare lethal disorder having an incidence of one per 1374 live births in an Indian study.[8] Higher rates in developing countries maybe due to variations in local cultural as well as puerperal practices, ecological factors, environmental influence, diagnostic criteria and reporting system used.[9]

Risk factors include multiparity, black race, older maternal age, pre eclampsia and gestational hypertension.[1] Symptoms of PPCM include fatigue, edema and dyspnoea which are similar to those for normal spectrum of peripartum states and pregnancy such as pulmonary emboli and eclampsia. Therefore, diagnosis is often delayed and the disorder in under recognised with devastating consequences. Mortality is as high as 20-50%.

Treatment of PPCM is the same as for other forms of congestive heart failure (fluid and salt restriction, beta blockers, diuretic and digoxin), except for angiotensin converting enzyme inhibitors and angiotensin receptor blockers which are contraindicated in pregnancy but maybe used in post partum. [10]

Pregnancy and puerperium are hypercoagulable states. Anticoagulation is advisable when LVEF falls below 35% or in case of atrial fibrillation or mural thrombi during pregnancy and atleast 2 months post partum.

Sudden death and ventricular arrhythmias are common in PPCM. Beta blockers maybe used to treat supraventricular tachyarrythmias.

Novel targeted therapy with bromocriptine is based upon experimental observation of prevention of PPCM in mice via prolactin blockade. A prospective study conducted by Sliwa K et al concluded that PPCM-bromocriptine patients displayed greater recovery of LVEF compared with PPCM- standard treatment patients at 6 months.[11]

Some experts including the 2010 European Society of Cardiology working group, suggest that breast feeding be avoided because of the potential effects of prolactin subfragments.[2] Overall, given the benefits of breast feeding, women who are clinically stable should not be discouraged from breast feeding if it is compatible with other medication.

In about 50% of the patients, ejection fraction normalize. Mortality rates have varied from 0-19%, while rates of cardiac transplantation have ranged from 6-11%.[12,13] The predictors of maternal mortality are poor NYHA status, LVEF <30%, higher maternal age, black race and multiparity.[14]

Predictors of persistent LV dysfunction at follow up are LVEF <30%, fractional shortening <20% and LV end diastolic dimension >6cm, elevated cardiac troponin T and black race.[14]

Regardless of the recovery, however, a second pregnancy is usually not recommended for these patients because PPCM recurs in more than 30% of subsequent pregnancies, which puts both the mother and baby at risk.[15]

Conclusion

Peripartum Cardiomyopathy is a form of systolic heart failure of relatively obscure etiology. It is important that Obstetricians should be familiar with PPCM and therefore consider it when diagnosing dyspneic patients to expedite medical treatment for a potentially lethal condition.

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