Peripartum Cardiomyopathy

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ABSTRACT

BACKGROUND

Peripartum cardiomyopathy is a rare, dilated cardiomyopathy with systolic dysfunction that occurs in the late pregnancy or more commonly in the early post-partum period. Risk factors include pre-eclampsia, advanced maternal age, multiple gestation. Although the complete pathogenesis is unclear, researches over the past few years suggest the importance of vasculo-hormonal pathways in women with underlying susceptibility and sarcomere gene mutation. Most of the patients will recover with adequate systolic function, although very few may go on to chronic cardiomyopathy. Other potential complications include thromboembolism and arrhythmias.

METHODS

This is a prospective study conducted at Kanyakumari Government Medical College between May 2019 and May 2021. 10 cases were observed and followed up. ANC cases with acute heart failure and no history of previous heart diseases were included in the study. Patients with known heart diseases were excluded from the study.

RESULTS

10 cases of ANC cases were diagnosed with peripartum cardiomyopathy with half of them having comorbidity and others were comorbid free. Diagnosis was confirmed with echocardiography and all the patients recovered with medical management.

CONCLUSIONS

Though peripartum cardiomyopathy is a rare occurring event, incidence of 10 cases in a period of 2 years highlights the need for us to be aware of this peripartum cardiac event and have it as a differential diagnosis in the situational setting at the back of the mind. Since this is a serious medical condition that affects pregnant women, prompt and timely medical intervention will save the patient from serious life-threatening complications

KEY WORDS

Peripartum Cardiomyopathy, Pregnancy, Heart Disease, Dyspnoea in Pregnancy.

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BACKGROUND

Peripartum cardiomyopathy is an idiopathic and often dilated cardiomyopathy that is marked by systolic dysfunction that presents in late pregnancy or in the early postpartum period. ESC - European society of cardiology defined peripartum cardiomyopathy as heart failure that occurs "towards the end of pregnancy or in the months following delivery, where no other cause of heart failure is found."[1]

Physical examination would reveal pulmonary rales, elevated JVP, S3 gallop. ECG showing sinus rhythm in most women with non-specific ST / T changes. Chest Xray shows pulmonary oedema and may show cardiomyopathy. NT pro BNP and BNP, troponin may be raised. Diagnosis requires echocardiography evidence of left ventricular dysfunction and often but not always left ventricular dilatation, with absence of any other cause. Echo may also show right ventricular dilatation and dysfunction, pulmonary hypertension, left atrial and biatrial enlargement, functional mitral and tricuspid regurgitation.

Although worldwide occurrence is common, most epidemiological data came from United states, South Africa, Nigeria and Haiti. In US its incidence risen up from one in 1181 live births in 2004 to one in 849 live births in 2011. In India it occurs in 1 in 3000 to 1 in 15000 pregnancies and the incidence may be increasing. The commonest risk factors include: advanced maternal age, pre-eclampsia, multiple gestation, hypertension, diabetes, obesity, twin gestation. Black patients are more susceptible than white patients.[2]

We intend to study the pattern of presentation, course of the disease and outcome of pregnancy in peripartum cardiomyopathy.

METHODS

This is a prospective study conducted at Kanyakumari Government Medical College between May 2019 and May 2021. Total number of samples were 10 and were taken as random sampling.

Inclusion Criteria

ANC cases with acute heart failure and no history of previous heat diseases were included in the study

Exclusion Criteria

Patients of known heat disease.

Pathogenesis and Biology

Although the complete pathogenesis of PPCM remains unclear, current hypothesis favours a "two hit" model of PPCM pathogenesis, whereby a vascular insult caused by anti-vascular or hormonal effects of late pregnancy and the early postpartum period induces cardiomyopathy in women with an underlying predisposition, which includes genetic predisposition causing alteration in sarcomeric proteins, hormonal effects which includes prolactin[2], placental angiogenic factors, autoimmune mechanism against adrenergic receptors and sarcomeric proteins with underlying risk factors. Thus, a heterogenous pathophysiology plays an important role in occurrence of peripartum cardiomyopathy.

When to Suspect Peripartum Cardiomyopathy

1. Heart failure within last month of pregnancy to 5 months postpartum
2. Absence of prior heart disease
3. No determinable cause
4. Echo findings of:
   - EF < 45 %
   - LV fraction shortening
   - End diastolic dimension > 2.7cm/m2

Clinical Presentation and Diagnosis

Women with PPCM experience symptoms of right heart failure or left heart failure which includes dyspnoea at exertion, orthopnoea, PND and oedema of lower extremities. Rare presentations include arrhythmia, shock, arterial thromboembolism. Physical examination reveals pulmonary rales, elevated JVP, S3 gallop. ECG showing sinus rhythm in most women with non-specific ST / T changes. Chest Xray shows pulmonary oedema and may show cardiomyopathy. NT pro BNP and BNP, troponin may be raised.

Diagnosis requires echocardiography evidence of left ventricular dysfunction and often but not always left ventricular dilatation, with absence of any other cause. Echo may also show right ventricular dilatation and dysfunction, pulmonary hypertension, left atrial and biatrial enlargement, functional mitral and tricuspid regurgitation, and intracardiac thrombus. Cardiac MRI can also be helpful when echo is technically not possible. Endomyocardial biopsy can show sarcomeric alteration which is not regularly indicated.

A total of ten patients were identified during the study period who fulfilled the inclusion criteria. Clinical data, including age distribution, gestational age at presentation, parity, presenting symptoms and identifiable risk factors was collected. Other causes of heart failure were ruled out by detailed clinical examination, ECG and cardiac enzymes for ischemic events, thyroid profile and ANA were also done.

2D ECHO with M mode and Doppler was used and data was analysed by Medialogic software. ECHO parameters measured were LV end diastolic dimension, LV fractional shortening and LV ejection fraction.[3] Echocardiography was repeated at one months and six months by the same cardiologist. A multidisciplinary team of physician, nephrologists' obstetricians and cardiologists were involved in the management. Management goals included preload optimization, afterload reduction and increasing the cardiac contractility. All patients received diuretics, digoxin, vasodilators and b-blockers. Selectively, ACE inhibitors in postpartum patients, inotropes and anti-coagulants were also used. Additionally fluid and salt restriction was instituted for an optimisation of blood pressure.[4] Mode of delivery was individualised as per obstetric requirement and pregnancy outcomes were systematically documented. Complications encountered such as pulmonary oedema, acute kidney injury, pleural effusion etc were appropriately managed.

Statistical Analysis

Data was collected and entered in an Excel sheet and the mean value and percentage were calculated for parameters under study.

RESULTS

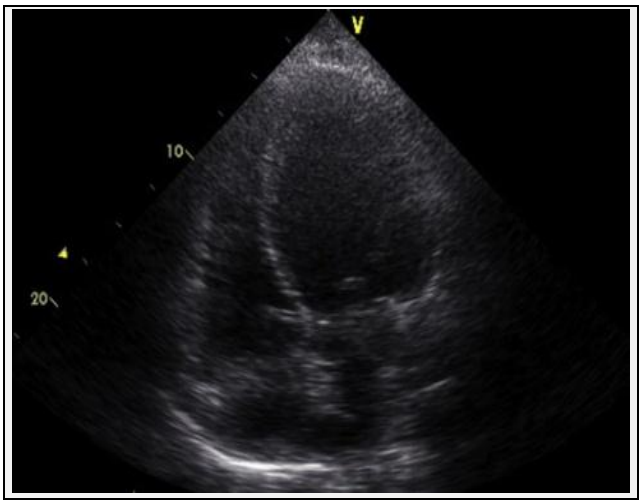
Our study revealed that out of 10 PPCM cases, 3 were primi gravid and 7 were higher gravida. Out of 10, 3 patients had gestational hypertension and 2 were having hypothyroidism[5]. The initial occurrence of symptoms of heart failure were predominantly during the postpartum period 6/10 (60 %). Cases presenting antenatally were at about 37 to 38 weeks gestation and postpartum patients were between first day postnatal up to four weeks of gestation. Symptoms presented were dyspnoea (100 %) at presentation, followed by cough (70 %), chest pain (25 %), shock (10 %). According to NYHA (New York Heart association) 1/10 (10 %) were in Class II, 3/10 (30 %) were in Class III and 3/16 (60 %) were in Class IV. All the cases were available for further follow up, all of them underwent serial 2D ECHO at one month and at six-month interval. PPCM mortality rates: zero mortality in our study is probably related to availability of advanced cardiac and critical care unit.

Prognosis based on 2D ECHO at diagnosis: Echocardiography parameters in our study like LV size, ejection fraction and fractional shortening at diagnosis differed significantly between those who needed longer in patient care and those who recovered earlier

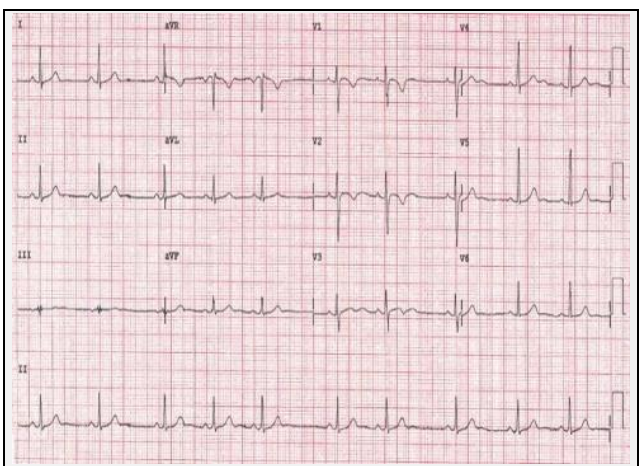
Special Presentation

Case 1: Peripartum cardiomyopathy with cardiorenal syndrome

28 years old primi at 38 weeks gestation, presented with exertional dyspnoea on flat surface (NYHA GR III) for 1 week. She is not a known case of SHTN, DM, pre-eclampsia, GHTN, GDM, CHD. O/E patient was dyspnoeic, tachypnoeic, B/L pedal oedema +, JVP +, BP; 126/78, PR; 104/min CVS: tachycardia +, PSM +, RS: B/L basal crepts +. Mild elevation of urea and creatinine were present. CBC. LFT, TFT, blood sugar was with in normal limits. Serum prolactin was in higher range. Chest Xray showed b/l costophrenic angle obliteration suggestive of pleural effusion with. ECG showed t inversion v1 to v3, echo was taken which showed left ventricular systolic dysfunction with ejection fraction about 35 %, increased left ventricular end diastolic pressure. Patient was managed with backrest, oxygen support, loop diuretics and digoxin. Termination of pregnancy was advised, nephrologist and cardiologist advices were obtained and carried out. 1 month later follow up echo was taken and systolic function was normal. Patient was tapered from drugs and was stopped.



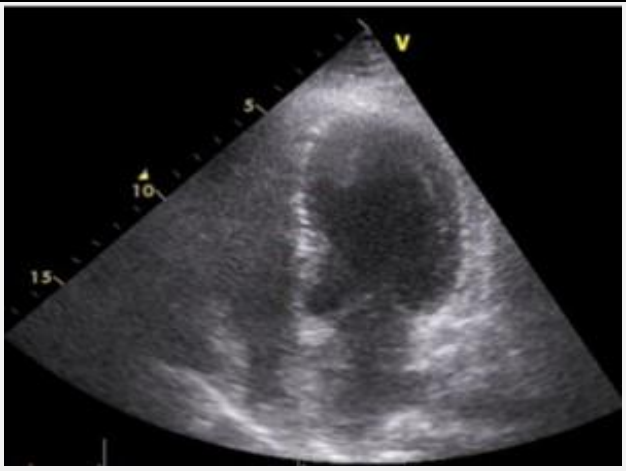
Echo Showing Dilated Left Ventricle



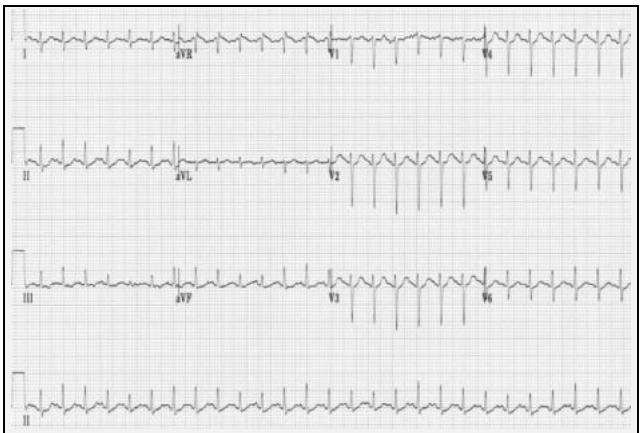
Sinus Tachycardia with Inversion in V1 to V3

Case 2: Peripartum cardiomyopathy with severe LVSD with LV apical ballooning takotsubo cardiomyopathy

24 years old para 3, post LSCS, on the day of surgery had c/o acute episode of dyspnoea 3 hours after lscs with decreased urine output. Patient was dyspnoeic, tachypnoeic, using accessory muscles of respiration, tachycardia +, BP was 130/90, JVP was raised, B/L pedal oedema +, LVS3 heard, B/L basal crepts +. CBC, RFT, LFT, prolactin, TFT, blood sugar was with in normal limits. Bed side Xray showed b/l batwing appearance with b/l costophrenic angle obliteration. ECG showed poor progression of R wave, tachycardia with secondary ST, T changes. Echo was taken which showed, LV apical ballooning, EF 19 %, mild MR, elevated LVEDP, LVEDD suggestive of peripartum cardiomyopathy. Patient was treated with backrest, PSV-NIV mode ventilation, loop diuretics, digoxin, beta blockers, spironolactone, and enoxaparin. Patient recovered gradually with medications and supportive measures. In-hospital repeat echo was taken which showed EF of 32 % after 1 week. Patient was discharged with medications. Repeat follow up echo was taken after 1 months which showed LV ejection fraction of about 46 %.



Echo Showing Takotsubo Cardiomyopathy, Ballooning of LV



Sinus Tachycardia with Poor Progression of r Wave

DISCUSSION

These cases were evaluated and diagnosed at kgmch and managed with cardiologists' opinion. All of them recovered with good systolic function. While the underlying pathophysiology remains unclear, vasculo-hormonal influences and genetic susceptibility probably play an important role, clinical examination. Diagnosis of PPCM as previously mentioned, rests on the echocardiographic identification of new left ventricular systolic dysfunction, depressed fractional shortening and ejection fraction during a limited period surrounding parturition.[5] Management with early termination of pregnancy, back rest, oxygen support, fluid restriction, loop diuretics, digoxin, ACE- inhibitors, spironolactone will effectively stabilize the patient.[5] Novel drugs which include bromocriptine, and pentoxifylline.[5] Most of the women will recover and will have a normal systolic function. Complications includes arrhythmia, thromboembolism, which has to be managed appropriately with anti-arrhythmics and low molecular weight heparin. Most of the patients will recover well with prompt timely management, some patient may land up in chronic cardiomyopathy. If this patient plans for next pregnancy the probable complications depend on the adequate left ventricular function prior to conception, there are recommendations to do screening echo in these patients in their subsequent pregnancy at preconception, end of 1st trimester, end of 2nd trimester, 1 month before delivery, immediately after delivery, 1 month after delivery.[1] Awareness about the incidence will strike the diagnosis earlier and will help in earlier initiation of prompt treatment. Even with full recovery subsequent pregnancies carry a 30% risk of relapse.[5]

Predictors of Recovery

In addition to racial background and in part independently from it, EF at presentation best predicts rate of recovery.

In the IPAC study, only one third of the 27 women with LVEF < 30 % at presentation recovered EF to > 50 % at 1 year. None of the women who also had evidence of dilation (LV end-diastolic diameter > 6.0 cm) recovered compared with recovery in nearly 90 % of the 65 women who presented with LVEF > 30 %.[6] Events (death, LV assist device, or heart transplantation) occurred almost exclusively in women with LVEF < 30 % at presentation compared with those with LVEF ≥ 30 %.[6]

Interestingly, LVEF at presentation in the Soweto study did not predict outcome in this large cohort, 87 in contrast to US studies. Lower levels of plasma troponin or brain natriuretic peptide have also been associated with improved outcomes. However, like LVEF, their predictive accuracy is not sufficient to affect clinical decisions.

Patients in whom these conditions contribute to the development of PPCM, resolution of preeclampsia or gestational hypertension may, for example, accelerate resolution.

Recurrence with Recurrent Pregnancies

The question of the risks of carrying a second pregnancy often looms large in women who have had PPCM. A recent comprehensive review of the literature on this topic,[7] covering 191 recurrent pregnancies, showed that the risk of relapse in patients with persistent LV dysfunction before their recurrent pregnancy is much higher than in those who have normalize LV function: 48% of the former group (n=93) had significant deterioration of LV function and 16 % died, whereas 27 % of the latter group showed deterioration and no deaths were reported. The risk of worsening PPCM with recurrent pregnancy is thus substantial. The best predictor for relapse and deterioration of cardiac function is pre-pregnancy LVEF, but normalized LV function does not guarantee an uncomplicated subsequent pregnancy.

Many women, however, strongly desire subsequent pregnancies, the decision of whether to proceed is therefore difficult and highly individual. No firm recommendations on this subject can be made on the basis of available data. Nevertheless, waiting for normalization of LV function in the absence of medications is prudent, and women should be advised that they have a high risk of recurrence even if their LV function has recovered, which could lead to a severe and at times persistent decline in LV function or life-threatening complications, and that long-term outcomes are currently not known.[7] A team-based approach to decision making, including well-informed clinicians and patients, is highly advocated. If the decision to proceed with pregnancy is made, close monitoring of symptoms, LV function, and brain natriuretic peptide is highly recommended during and after pregnancy.[7]

Treatment after Recovery

Because of a lack of long-term follow-up data in women with PPCM, it is not clear if and when women with the condition can be considered to have fully recovered. This is an important issue, bearing on the decision of whether to discontinue long term medications in young and otherwise healthy women. One study of 15 patients with full LVEF recovery who stopped taking angiotensin-converting enzyme inhibitors or β-blockers reported no deterioration of LV function during a 2-year follow-up.[8]

A reasonable approach to the discontinuation of medications in women with complete recovery of LV function would include waiting until a few months after LV function has recovered, weaning of medications one at a time, and providing close clinical and echocardiographic monitoring during the discontinuation process followed by annual assessment of LV function.

CONCLUSIONS

Though peripartum cardiomyopathy is a rare occurring event, incidence of 10 cases in a period of 2 years highlights the need for us to be aware of this peripartum cardiac event and have it as a differential diagnosis in the situational setting at the back of the mind. Since this is a serious medical condition that affects pregnant women, prompt and timely medical intervention will save the patient from serious life-threatening complications.

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