PERIPARTUM DILATED CARDIOMYPATHY

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Summary

Peripartum Cardiomyopathy is the development of heart failure in the last trimester pregnancy or with in six month after delivery without an identify cause in a previously healthy Female . It is a uncommons condition with better prognosis. We demonstrate eight cases of peripartum Cardiomypathy presented with sign symptoms of heart failure of different severity & was confirmed by Echocardiography.

Introduction

Peripartum dilated cardiomyopathy (PPCM) is a serious disease with poorly understood etiology with a mortality rate of about 9% [1].

Incidence in 1:3500 to 1:4000.The clinical picture of PPCM is that of dilated cardiomyopathy but differs from others by its rapid development in peripartum & better prognosis with 50% completely recover & most of the rest will improve. It is a diagnosis of exclusion i.e [2].

- Absence of identifiable cause of heart failure.
- Absence of recognizable heart disease before the last trimester pregnancy.
- Absence of identifiable cause of heart failure
- Left ventricular systolic function demonstrated by LVEF<40%,Fraction shortening

of <20% or both with or without left ventricular end diastolic dimension<2.7%/sq body surface area.

Predisposing factors seem to be multiparity childbirths,obesity, multiple pregnancy, ethnicity, smoking, diabetes, hypertension, pre-eclampsia, malnutrion,advanced age of mothers(>30) or teenage pregnancy and prolonged use of beta agonists. Genetic and familial association also reported.The aetiology of PPCM is uncertain. Infection, inflammation and autoimmune process may play a role [3].

PPCM is suspected to be the consequence of an unbalanced oxidative stress leading to proteolytic cleaveage of prolactin into a potent angiostatic and into pro-apoptotic Fragments leading to accelerated myocyte death, decrease pump function and clinical manifestation of PPCM [3].

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In third trimester or puerperial patients usually present with history and clinical sign symptoms acute heart failure of varing severities. Normal findings in late pregnancy overlap considerably with early stage of heart failure,so cardiac symptom may not be recognized until the extrems.Initial presentation may be cardiac arrest during L.S.C.S.3

ECG changes are multiple & non-specific sinus arrythmia, ST & Q wave abnormality (96%),ventricular hypertrophy, bundle branch block and atrial fibrillation.QRS time more than 120ms is predictor of mortality [2].

Echocardiography reveals dilated left ventricle or all four chambers of heart.

LVED diameter greater than 56mm, LVEF <40%, Fractional shortening <20%, left ventricular hypertrophy with left ventricular global hypokinessia [2].

Xray chest shows shadowing, cardiomegally, pulmonary oedema and pleural effusion.

Treatment

Aim of the treatment is to reduce preload and after load as well as to improve cardiac contractility by conventional therapy. Rest with propped up position, continuous haemodynamic and oxygen monitoring by face musk, CVP and arterial Canulation.If oxygen face musk fails to improve SPO_2>95% ventilation needed. Mild to moderate do not need fluid restriction.

Diuretic to reduce preload, Inj Lasix (40 to 80mg/day) followed by 40 mg tab accordingly.

Hydralazine i/v or in drip to prevent afterload in pregnant patient with heat failure.

ACE or ARB (25 to 50 mg/day) be used in puerperium not in antepartum. Among the ca channel blocker only amlodipine can be used in preeclampsia to control blood pressure.

Beta blocker carvidelol, metoprolol decrease heart rate, imptove left ventriculer diastolic function, protect against arrhythmia and safe in lactation. Digoxin if atrial arrythmia can be used both in pregnancy and puerperium & also increase myocardial force of control.

NTG 10 to 20 ug/min to 200ug/min afterload as well as dopamine dobutamine,milrinone can be used to give inotrophic support to failing heart if BP not maintain.

Levsimendin give inotrophic support in low output failure at a rate of.1 to.2ug/kg/min with or without loading dose of 3-12ug/kg over 10 minutes [4].

Bromocriptine 2.5mg can be used for 6 month.
Anti coagulation with low molecular weight heparin if EF<35%,severely dilated ventricle, atrial fibrilation and presence of mural thrombus. Cardiac transplantation can be needed in 11% cases.
Silva et al showed treatment with i/v immunoglobulin and pentoxyphylline had significant improvement of PPCM NYHA class III & IV (52%vs 27%,p=0.03).Stem cell therapy is on trial [5].
Vaginal delivey is always preferable if patient is haemodynmically stable (EF>35%) and no obstetric indication for L.S.C.S. Close haemodynamic monitoring is needed. Epidural analgesia is preffered.Emergency.If L.S.C.S is to be done for obstetric indication regional anesthesia is the method of choice but contraindicated in a anticoagulated patient [6]. L.S.C.S is done irrespective of gestation in women with advanced failure and haemodynamic instability despite treatment.5General anesthesia is preffered in patient with poor cardiac reserve.
Recently the choice for combined spinal epidural may be opted for several reasons [7].
After complete recovery discontinuation of therapy should be done gradually with repeat ECHO evaluation of cardiac function Annual ECHO is recommended in patient with PPCM.

Case Reports

We report on eight cases diagnosed as PPCM in CCU Chittagong Metropolitan hospital Pvt Ltd in about last three years-

1. A 38 years Para 4+0 admitted on 5th postnatal day of her vaginal delivery at home with acute respiratory distress and collapse. She complains of inability to lie flat since 3rd postnatal day.
2. A 28 years 2nd gravid para 1+0 had emergency cesarean section (c/s) at 39weeks of pregnancy for P/H/O L.S.C.S with labour pain. She had no regular antenatal check up but gave H/O of respiratory distress on 2nd post operative day following 1st L.S.C.S, no document was available. Her postoperative period following 2nd L.S.C.S was uneventful and she was discharged on 5th post operative day. She was readmitted on 9th postoperative day with cardiogenic shock. Inspite all measure she died 3 hours after her admission.
3. A 25 years prime with regular antenatal check up admitted at term with sudden onset of dysponea and orthopnea . She had other sign symptom of heart failure. After consultation with cardiologist, ECG, ECHO & medical management planned for cesarean section. She needed resuscitation During L.S.C.S & ventilatory support for 48 hours. She was in intensive care for 10 days & recovered gradually.
4. A 30 years 2nd gravid para1+0 admitted with labour pain at 39 weeks of her pregnancy in active labour.She was haemodynamically stable . She gave history of PPCM on 3rd postnatal following her uneventful vaginal delivery. She was treated by cardiologist,recovered well & was under his supervision throughout 2nd pregnancy.ECHO revealed EF 42%. Her labour was progressing well & she delivered four hours after her admission under close supervision, second stage was cut short by ventouse. Her postnatal period was uneventful but on 5th postnatal day she felt mild orthopnea but ECG & ECHO findings revealed no deterioration of cardiac status. She was discharged on 10th postnatal day with treatment and advice for follow up.
5. A 35 years lady para 3+0 having L.S.C.S. for severe preeclampsia, admitted on 5th postoperative day with feature of acute heart failure. She was haemodynamically unstable and managed accordingly.Her ejection fraction was 33%.Breastfeeding was discoursed and tab bromocriptine 2.5mg twice daily was added.She improved with in 72 hours & was discharged 10days after admission with advice to continue treatment and to come for follow up. Repeat ECHO after 3 month showed 80% recovery of cardiac status.
6. A 30 years obese prime at 36weeks (twin) had L.S.C.S for severe preeclampsia. Her blood pressure controlled with in 24 hours of L.S.C.S. But on 5th postoperative day she developed respiratory distress. With other feature of heart failure and managed accordingly.

There were also two patients admitted in puerperium with signs of heart failure but haemodynamically stable. One had history of genital warts at 22weeks and other had viral fever at 30weeks of pregnancy. In all eight patients history,examination and ECHO lead ultimately to diagnosis of peripartom cardiomyopathy.

Discussion

PPCM is a pregnancy associated myocardial disease with high maternal mortality and morbidity. The etiology of PPCM is still unknown and many potential causes have been proposed but not proven.
These include viral myocarditis, abnormal immune response to pregnancy,abnormal response to increase haemodynamic burden of pregnancy, hormonal abnormality ,malnutrition, Chlamydia infection, inflammation and apoptosis.
In our report two patients had history of genital warts and viral fever in pregnancy. Pregnancy is a state of immunesuppression. There is records that haemodynamic change and immunosuppressive state of pregnancy may have exaggerated de-novo or

reactivated Latent virus like herpes simplex virus 6, parvovirus, Ebestein Bar Virus, cytomegalovirus, other cardiotrophic virus leading to viral Myocarditis & then cardiomyopathy.

High titre of auto-antibody against cardiac tissue protein, foetal Cell and Actin myocin released from uterine muscle during delivery are demonstrated in patient with PPCM. These autoantibody cross react with maternal myocardial and leads to PPCM. Among reported eight patients most of them had one or more predisposing factor in favour of PPCM.
Stress of severe preeclampsia and L.S.C.S in elderly can predispose to PPCM. A-E.Baruteau et al reported two sisters both with L.S.C.S for severe preeclampsia in first pregnancy at 34years to develop PPCM three years apart, also signifying familial PPCM, though none of our had familial predisposition.

Multiparity(>4) [8] though a risk factor for PPCM more recently ,John Abboud et al did not support a strong association between multiparity and developing of PPCM because almost 40% of the cases PPCM developed in first pregnancy and more than 50% case with the first 2 pregnancies. In our reports only one patient had 4 children and most of the patient were around 30 years or more [8].

Increased LV dimensions lower LVEF an usual ECHO findings in PPCM are always associated and correlated with elevated plasma level of proinflamatory TNF, Cytokines, oxidized LDL ,increased activity of prolactin cleaving enzyme Cathepsin D that increase 16KDA Prolactin. In addition peripheral blood film in patient PPCM demonstrate decrease oestrogen, progesterone,relaxin, regulatory T cell and cardioprotective STAT3 [9]. In one study performed in Niger,96% patients were positive for Chalamydia IgG & IgA antibody and has a prognostic value at the of diagnosis, high dilution associated with poor prognosis [10]. All the reported eight patients had typical ECG and ECHO changes in favour of PPCM.

But plasma markers of inflammation and hormonal levels were not detected in these patients.

One patient with LVEF 33% treated with tab bromocriptine 2.5mg twice daily for 14 days and adviced to continue for 6 month.She was also advised to discontinue breastfeed. Oxidized LDL and 16k DA prolactin are associated with accelerated myocyte death, Decrease pump fuction and clinical manifestation of PPCM. A study on 12 patients in South Africa all receiving standered heart failure treatment and 6 PPCM patient received additional bromocriptine for 6month. Cardiac function improved in all 6 cases with no death while 3 patient died within 4months of terminal heart faiure, 3 surviving patient developed recurrent left ventriculer dysfunction in no bromocriptine group. Ten similar study was done in Germany on 6 Patients [10].
Both concluded bromocriptine can prevent development of PPCM in women at high risk , improved pump function (EFfrom 15% to 51%) in NYHA Class III to IV and severly restricted pump function(EF 12% to 30%).10So clinically stable patient can continue breast feeding but acute severe PPCM patient with EF <30% preventing lactation can be considered.

In subsequent pregnancies there is a risk of recurrence and even mortality, risk is substantially higher in patient with persistent LV dysfunction before pregnancy. Patient sould be advised on the risk of subsequent pregnancy and never become pregnant again and on safest and most effective contraception [11]. Base line ECHO should be done before or in early pregnancy, at least 3 month after the discontinuation of ACE inhibitor or ARBs if patient is on heart failure treatment and isosorbide dinitrate-hydralazine combination is to be substituted.Patient should be followed with repeat ECHO during early second and third trimester,during the last gestation month,early after delivery and any time if new symptoms of hear failure develop and then annually [12] Early termination of unintentional pregnancy should be considered specially in persistent LV dysfunction [13].

Conclusion

The outcome of PPCM is better prognosis. 50% patient clinical improved and ECHO status complete recovery and return to normal, others deteriorate rapidly, develop persistent cardiac dysfunction (30%) while significant number do not respond to medical therapy, few patients needs cardiac transplantation or even die. So key point is to determine risk factor and prognostic variable, early diagnosis and interdisciplinary management, ascertain cardiovascular risk factor for subsequent pregnancy to have substantial recovery from PPCM. Evaluation of therapeutic intervention and estabilishment of acentra serum and tissue bank is recommended.

Disclosure

All the authors declare no competing interest.

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