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Case Series

The myriad presentations of peripartum cardiomyopathy

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ABSTRACT

Peripartum cardiomyopathy (PPCM) is an idiopathic, non-ischemic systolic type of heart failure which can present anytime from the last month of pregnancy till the end of 5th month post-partum. The incidence of PPCM in the Indian population is 1:1340 with 60% of the cases occurring post-partum. PPCM has a mortality rate of 11.7% with unpredictable sequelae ranging from worsening heart failure, cardiogenic shock, development of arrythmias to complete recovery and recurrence in subsequent pregnancies. With an idiopathic aetiology with multiple theories, PPCM remains a diagnosis of exclusion, demanding a high index of suspicion and surveillance in pregnant women. The management involves a multidisciplinary approach involving the obstetrician, cardiologist and at times the anaesthesiologist and includes various drugs like beta- blockers, diuretics, digoxin, bromocriptine. In severe cases, maternal circulatory support may also be needed. We present three cases of PPCM diagnosed in the antepartum, intrapartum and immediate post-partum periods respectively. Out of 3 patients, one delivered vaginally and two underwent emergency caesarean sections. All of patients went home post-delivery with good outcomes and were doing well after 4 weeks of delivery.

Keywords: PPCM, Heart disease in pregnancy, Beta-blockers, Digoxin, Cardiogenic shock, Systolic dysfuction, Echocardiography

INTRODUCTION

The 2010 heart failure association of the European society of cardiology defines PPCM as "idiopathic cardiomyopathy presenting with heart failure secondary to left ventricular systolic dysfunction (left ventricular ejection fraction <45%) towards end of pregnancy or in months following delivery where no other cause of heart failure is found".[1] Thus, PPCM is essentially a diagnosis of exclusion.[2] Majority of cases occur post-partum (nearly 60-70%) with considerable ethnic and regional variability in incidence, severity of symptoms and even recovery rate.[3] A variety of risk factors have been identified by multiple studies like: African-American heritage, older maternal age, multifetal gestation and anaemia.[1,4]

Women with PPCM can present with non-specific symptoms like fatigue, ankle swelling, shortness of breath which may be confused with symptoms of pregnancy leading to delayed diagnosis. Specific symptoms of heart failure like orthopnoea, paroxysmal nocturnal dyspnoea, chest tightness or acute presentations of cardiogenic shock, arrythmias/thromboembolic complications are rare. Delay/misdiagnosis often leads to increased complications and poorer outcomes.[5] Echocardiography forms the mainstay of diagnosis which can not only accurately quantify the severity of systolic dysfunction but also give information about co-existent abnormalities/rule out other causes of heart failure like valvular heart disease, congenital heart disease, acquired/hereditary cardiomyopathies.[6,7] Additionally, electrocardiogram (ECG), chest X-ray, biomarkers like brain natriuretic peptide (BNP) and routine investigations like complete blood count (CBC) also have a role in establishing the diagnosis of PPCM.[7,8]

Here we present 3 cases diagnosed with PPCM with varied clinical presentations. All patients gave valid, informed consent for publication.

CASE SERIES

Case 1

A 32-year-old woman, 2nd gravida, with one living male child aged 8 years, delivered vaginally was referred to our institute at 36 weeks of gestation from her private physician in view of echocardiogram suggestive of markedly decreased systolic function (ejection fraction 35%) with global left ventricular hypokinesia. The patient had registered at 12 weeks of gestation, had regular follow ups and had had an uncomplicated antenatal course so far. She reported that she had been experiencing shortness of breath on and off since last 2 weeks which had prompted her primary physician to prescribe an echocardiogram in addition to chest radiogram (unremarkable), ECG (normal sinus rhythm) and CBC (within normal limits). She had no history of gestational hypertension or heart disease in her current or previous pregnancy. She did not have a family history of heart disease or similar complaints of breathlessness in her last pregnancy. Her physical examination on admission revealed normal blood pressure, sinus tachycardia (110 bpm), respiratory rate of 18/min and mild pallor. There was no oedema, adventitious breath sounds on auscultation or organomegaly. Her ECG confirmed sinus tachycardia with no other changes. Routine blood investigations including CBC, renal function test (RFT), liver function test (LFT) was within normal limits. Echocardiogram done at our institute was consistent with previous findings: global left ventricular hypokinesia with normal left ventricle size (left ventricle end systolic diameter 3.8cm and end diastolic diameter 4.4 cm) but markedly decreased systolic function (ejection fraction 35%) (Figure 1).

There was no pulmonary oedema or valvular abnormalities. The patient was started on oral Metoprolol 25 mg/ day and oral Furosemide 40 mg/day with fluid and salt restricted diet. She was admitted and monitored daily and did not report any fresh symptoms or worsening of breathlessness. She went into labour spontaneously at 38 weeks and delivered a healthy male child of 2.78 kg with good APGAR score. Intensive intrapartum monitoring of vitals and input-output was done. Outlet forceps was applied to cut short the 2nd stage of labour and active management of 3rd stage of labour was done followed by post-partum copper T insertion.

Immediately after delivery, 20 mg furosemide was given intravenously. The patient was breastfeeding and did not show any worsening signs of heart failure in the post-partum period. Enalapril 5 mg/day was added from day 2 post-partum after a cardiology review. She was discharged on day 7 and an echocardiogram done 2 weeks post-delivery showed an improved ejection fraction of 55% with no left ventricular wall motion abnormalities. The patient was followed up again after 4 weeks where she had no complaints. Metoprolol, furosemide and enalapril were stopped at 6 week follow up.

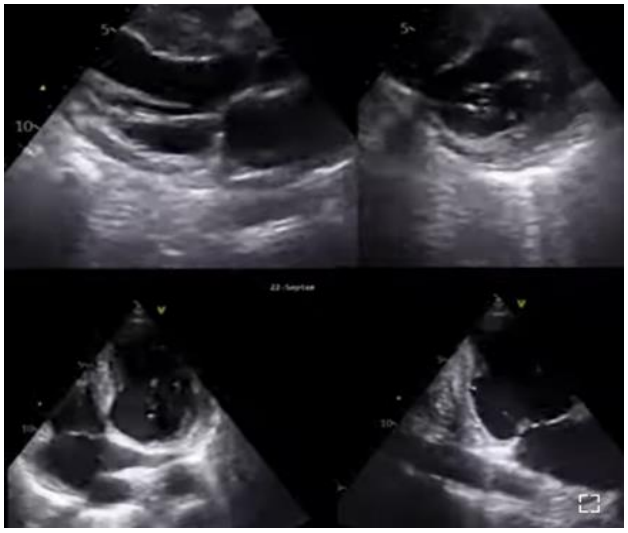

Figure 1: Antenatal echo of case 1 showing left ventricular ejection fraction 35%.

Case 2

A primigravida with monochorionic monoamniotic twin gestation at 34^{+2} weeks of gestation was referred to our institute for non-availability of neonatologist. She had no known comorbidities, had received regular antenatal check-ups and had no complaints at the time of admission. On admission, her CBC revealed microcytic hypochromic anaemia with a haemoglobin of 8.2 gm/dl with haematocrit of 29.9. All other parameters in the CBC were normal. Her RFT, LFT and blood sugars were normal. On physical examination she had normal blood pressure, pulse rate and respiratory rate. Signs of pallor were present. Chest auscultation revealed nothing abnormal. She was transfused with 1 unit of packed red blood cell and 2 doses of injection betamethasone 12 mg 24 hours apart were given to help with foetal lung maturity. She underwent caesarean section under spinal anaesthesia and delivered 2 female children weighing 1.8 and 2.0 kg with good APGARs. The procedure was uneventful with a blood loss of around 800 ml. 2 hours after surgery, the patient complained of breathlessness and chest tightness. Her saturation had dropped to 86% on room air with a respiratory rate of 30/min. Her blood pressure was 90/60 mmHg and pulse rate were 140/min. On auscultation there were coarse crepitations heard bilaterally. She was started on high flow oxygen by non-rebreather mask but her saturation remained 90%. She was shifted to the intensive care unit and electively intubated and put on volume AC mode of ventilation with 60% FiO_2. Portable chest radiogram showed pulmonary congestion, portable ECG was suggestive of sinus tachycardia and a bed side transthoracic echocardiogram revealed moderate systolic dysfunction with ejection fraction of 40% with mild mitral regurgitation and trivial tricuspid regurgitation (Figure 2). Her BNP level was 1500 pg/ml. Post-operative CBC, LFT and RFT were within normal limits. She was diagnosed as a case of PPCM. Intravenous furosemide 40 mg/day and bisoprolol were started for her along with fluid restriction.

The patient gradually improved with decreasing oxygen requirements and was extubated on post operative day 3. She started breastfeeding and was vitally stable till discharge. Echocardiogram done on day 10 post-delivery revealed improved ejection fraction of 60% with no left ventricular dysfunction. She was discharged on bisoprolol 1.25 mg/day and asked to follow up after 2 weeks in cardiology OPD. Eventually bisoprolol was stopped after 2 weeks and the patient went back to her normal life.

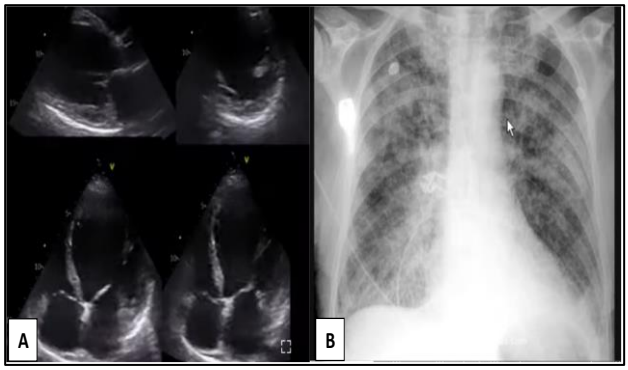

Figure 2 (A and B): 2D echo of case 2 done on postoperative day 1 showing left ventricular ejection fraction 40%, chest radiogram of case 2 showing pulmonary congestion.

Case 3

A 27-year-old primigravida at 35+2 weeks of gestation with intrauterine fetal death was admitted to our labour room. She had no history of gestational hypertension, heart disease. Her antenatal course had been uneventful. Her routine investigations including CBC, RFT, LFT and coagulation studies were all within normal. She underwent induction of labour and after 6 hours of induction developed acute breathlessness with saturation falling to 84%. Her respiratory rate had gone up to 30/min. Her pulse rate was 130/min and systolic blood pressure fell to 60 mmHg. She was electively intubated in the labour room by a team of anaesthesiologists and started on inotropes. Her arterial blood gas was suggestive of type 1 respiratory failure with metabolic acidosis. CBC was repeated which showed Hb 10.2 gm/dl, WBC of 8400/µl and platelet count of 1.58 lakh/µl. Her INR was 1.1, but d-Dimer was 780 mg/l and BNP was 2311 pg/ml. She was taken up for an emergency caesarean section to facilitate any resuscitative efforts required. Intraoperatively, uterus was completely atonic despite use of uterotonics. Hayman's compression sutures were taken. Following the procedure, patient was shifted to the ICU where a bedside echocardiogram showed left ventricular global hypokinesia with ejection fraction of 25% (Figure 3). ECG was suggestive of left axis deviation with normal sinus rhythm, chest radiogram was unremarkable. The patient was kept on ventilatory and circulatory support for 24 hours following which she was started on oral furosemide 40 mg/day, metoprolol 25 mg/day and cabergoline 0.25 mg/day for 2 days. Her vitals stabilised on day 4 post-operatively. Her echocardiogram done on Day 10 showed an ejection fraction of 45%. Her medications were changed to telmisartan 40 mg/day. She was discharged and asked to follow up. The patient was seen after 2 weeks where her ejection fraction was 60% and she had no complaints.

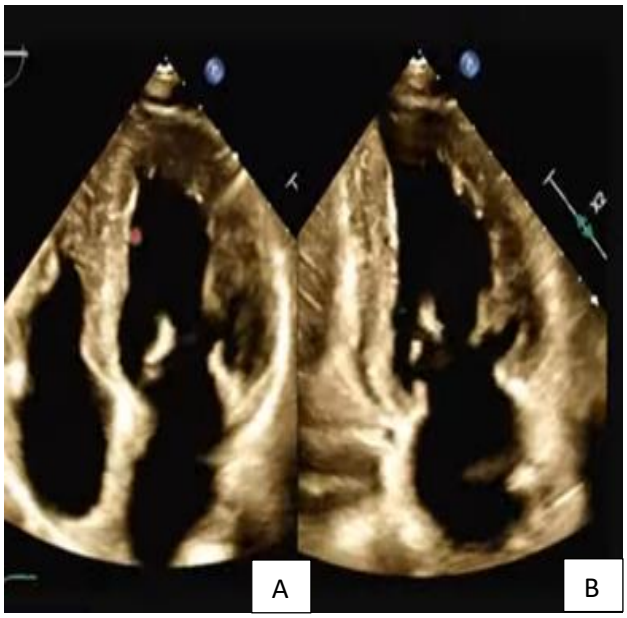

Figure 3 (A and B): Echo for case 3 done on post-operative day 1 suggestive of left ventricular ejection fraction 25%.

DISCUSSION

The pathophysiology of PPCM is multifactorial and poorly understood but there is strong evidence to indicate that it is largely a vascular disease triggered by the hormonal shifts of pregnancy.[9] Cytokines like Interleukin-6 (IL-6), Tumour necrosis factor-a (TNF α), C-reactive protein (CRP) and Interferon-gamma (IFNɣ) have been found to be elevated in PPCM and correlated with the severity of cardiac failure.[10] Activation of the 16-kDa prolactin fragment, decreased expression of the proangiogenic vascular endothelial growth factor (VEGF) and elevated soluble Fms-like tyrosine kinase 1 (sFlt1) levels have all been strongly implicated in various studies ono PPCM.[10] Multiple studies have also suggested a genetic basis for this disease with emphasis on mutations in the sarcomeric gene titin (TTN), and other genes like DSP, FLNC, and BAG3.[11] However, there is no role of genetic testing to diagnose or predict PPCM at present.

The treatment modalities are modified as per safety in pregnancy but largely comprise of beta blockers, digoxin, loop diuretics and injectable anti-coagulants. Post delivery, most medications are safe and the therapeutic arsenal can be extended to include drugs like angiotensin converting enzyme inhibitors (ACE I), aldosterone receptor blockers (ARB) and newer drugs like sacubutril-valsartan, ivabradine, levosimendan and warfarin. While the medications may be continued indefinitely in cases of

poor cardiac function, there is no clear consensus on the optimal duration of treatment in recovered cases.[12]

Labour and delivery need multidisciplinary planning and close monitoring especially for fluid balance. In most cases vaginal delivery is considered safe unless obstetrically contraindicated or in cases of hemodynamic instability.[13] In cases of worsening heart failure despite medical therapy, early delivery may be considered along with early initiation of maternal circulatory support (MCS). The role and timing of insertion of implantable cardioverter-defibrillator (ICD) devices to mitigate sudden cardiac death in PPCM is controversial.[14]

Patients who have survived PPCM need to be counselled on the risks of recurrence in future pregnancies. Relapse rates are higher in women with less than 50% left ventricular ejection fraction prior to subsequent conception and can be upto 20-25%.[15] Women with ejection fraction >50% have a lower rate of recurrence and lesser chance of severe deterioration. All patients with history of PPCM merit stringent follow up during their future pregnancies involving a cardiologist and an obstetrician. Clinical assessment, echocardiogram and/or BNP levels are recommended at the end of each trimester, one month prior to delivery, prior to discharge and 1 month post-delivery.[16] For women not keen on further childbearing, appropriate contraceptive counselling must be done. Intrauterine copper containing or progesterone containing devices, non-hormonal barrier method, injectable progesterone and even tubal ligation can be offered in patients of PPCM.

CONCLUSION

In spite of ongoing research, numerous uncertainties regarding the pathophysiology, risk factors, treatment and outcomes of PPCM patients continue to exist, indicating the need for continued investigations. We hope the above case series highlights not only the elusive diagnosis of PPCM, but also, how prompt intervention can be life-saving. Since the presentation of PPCM ranges from symptoms mimicking normal pregnancy to maternal collapse, a high index of suspicion is necessary among obstetricians to manage this potentially fatal condition.

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