Expect the Unexpected - Case Series of Peripartum Cardiomyopathy

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Abstract

Peripartum Cardiomyopathy (PPCM) is a serious and rare condition of unknown cause that affects childbearing woman. The symptoms of PPCM are likely to be confused with physiological changes associated with advanced pregnancy therefore high degree of suspicion should be present. In majority of the cases, the overall prognosis is good, although some patients may progress to irreversible heart failure. Early identification, diagnosis and prompt treatment at tertiary care hospitals reduces mortality and morbidity and improves the chances of complete recovery of heart function. Here we are presenting four clinical cases of peripartum cardiomyopathy, their course, treatment and outcome at Dr D.Y. Patil Medical College and Hospital, Pimpri, Pune, Maharashtra, who presented between January 2020 to November 2022. Ethical committee clearance was taken for this case study

INTRODUCTION

Peripartum Cardiomyopathy is a life-threatening primary, idiopathic form of heart disease. It occurs in the peripartum period associated with development of sudden onset cardiac failure in absence of any identifiable cause of heart failure in the last 6 weeks of pregnancy (34 weeks) or within 5 months of delivery. Its exact aetiology is unknown. Peripartum cardiomyopathy is diagnosed based on echocardiographic findings- Left ventricular end-diastolic dimension ≥2.7 cm/m2 or M-mode fractional shortening ≤30% or Left ventricular ejection fraction ≤0.451.

As it is a cause of significant mortality and morbidity among women of childbearing age throughout the world, we have reported four cases of PPCM from our setting of which one presented in the antepartum period and 3 presented in postpartum period. Their clinical presentations were variable leading to diagnostic challenges.

Case Presentation

Case One

A 30-year-old, primigravida (33 weeks of gestation) with severe preeclampsia was referred from primary health centre, she was planned elective LSCS after steroid cover. On day 2 of admission, patient had sudden onset dyspnoea and orthopnoea, emergency LSCS was immediately undertaken in view of severe Pre-Eclampsia with abruption and foetal distress. Intra-Operatively, Couvelaire uterus was noted, retroplacental clot of size 100 grams and thin meconium stained liqor was present. Baby was shifted to NICU in view of low birth weight and respiratory distress. 2 hours post LSCS, patient had dyspnoea of NYHA grade 3 in supine position, she was immediately shifted to intensive care unit for further management. 2D Echocardiography, chest x-ray, echocardiogram and blood investigations were sent. 2D-ECHO showed mild hypokinesia, ejection fraction 40% with severe MR. Chest Xray showed bilateral lung haziness and blunting of costophrenic angle. ECG was within normal limits. NT Pro BNP was elevated to 3749pg/ml and D-DIMER was 2498ng/ml. Patient was then diagnosed as a case of peripartum cardiomyopathy with severe MR with pleural effusion. The management was under combined team of intensivist and cardiologist. Patient was kept on non-invasive ventilator and supportive heart failure management comprising

of diuretics, beta blockers, ACE inhibitors. The signs of heart failure subsided within 2 weeks. The LVEF was 60% on repeat 2D-ECHO along with normal chest x-ray. Later, she was discharged with stable vitals with an advice to follow up.

Case Two

A 30-year-old woman presented to emergency department on post operative day 5 of LSCS done in view of preterm premature rupture of membranes, with dyspnoea NYHA grade 4 and palpitations for 3 hours. Her medical history was significant for hypothyroidism for which she was taking thyroxine. Her pregnancy had been otherwise uneventful. In emergency department, patient appeared to be anxious with blood pressure of 130/90 mmhg, heart rate of 128 beats per minute, respiratory rate was 18 breaths per minute and SPO2 was 96% on room air. She was then admitted in the cardiac care unit, her 2D ECHO showed LVEF of 20%, dilated RA, RV, LA, severe global LV hypokinesia and biventricular failure. Her ECG showed atrial tachycardia with 2:1 Block Rhythm. Laboratory blood tests were within normal limit. CT scan of chest was negative for pulmonary Emboli. Chest Radiograph findings was suggestive of right sided pleural effusion. Patient was given appropriate medications and treatment for heart failure. She gradually improved symptomatically and was discharged on day 10 with suggestion to follow up.

Case Three

A 26-year-old 4th gravida with previous 2 abortions and previous one LSCS presented in labor at 39 weeks of gestation with no significant obstetric, past, personal, or family history. Emergency LSCS was done in view of previous LSCS in labor. Antepartum, intrapartum, and immediate post-operative period was uneventful. On day 2 of post-partum period, she had dyspnoea (NYHA grade 3) and orthopnoea with bilateral lower limb oedema. On general physical examination, the woman was afebrile, orthopneic and mild pallor was present. Her respiratory rate was 24 breaths/min, SpO2 was 94% on room air, pulse was 130 beats per minute and blood pressure was 130/80 mmhg. On chest and cardiac auscultation, bilateral air entry was equal on both sides with no added sounds or crepts and S3 gallop heart sound was heard respectively. All laboratory blood investigations were within normal limits. ECG showed ST segment depression in lead II, III, V. Chest X-Ray showed borderline cardiomegaly. 2D Echo findings were consistent with severe left ventricular dysfunction with ejection fraction of 25%. Supportive management of heart failure including diuretic drugs, beta blockers, ivabradine, and inotropes like dobutamine and NTG infusion were given to patient, which alleviated her symptoms. 2D echo repeated on 5 days following admission showed increase in ejection fraction from 25% to 50%. Her follow up 2D Echo three months later showed normalisation of her ejection fraction to 60%.

Case Four

A 32-year-old, third gravida, unregistered at 41 weeks of pregnancy, with gestational hypertension on labetalol 100mg twice a day in the last one month, presented to labor room in active labor and gave birth by spontaneous normal vaginal delivery. She had high BMI. There was no history of any heart disease, hypertension, or pre-eclampsia in her previous pregnancies. On day 2 post-delivery, patient had an episode of vomiting, following which she had dyspnoea, grunting and drowsiness, she then had cardiac arrest and collapsed suddenly. She was immediately intubated and CPR was initiated by code blue team of our hospital. Patient revived with return of spontaneous cardiac rhythm after continuous CPR and was shifted to critical care unit for further management. She was kept on mechanical ventilator and ionotropic support. She had frank pulmonary oedema with spo2 90%. Her heart rate was 128 beats per minute, blood pressure was 154/100 mmhg on Noradrenaline 5ml/hr and respiratory rate 22 breaths/ min. On auscultation, bilateral crepts were present with normal S1S2 heart sounds. Her ABG was suggestive of respiratory acidosis and metabolic acidosis with severe hypoxemia, chest x-ray showed right lower zone patch with bilateral infiltrates, ECG showed tachycardia, 2D-ECHO showed ejection fraction of 20% with global hypokinesia of left ventricle, CT-scan was suggestive of aspiration pneumonitis with bilateral pleural effusion and MRI brain was suggestive of changes of hypoxic ischemic injury. Her D-DIMER was 10000 ng/ml, Troponin-I was 5437.30 pg/ml, BNP Levels was 1981 pg/ml and COVID-19 RTPCR was negative. CT-Brain was negative for intracranial bleed. Patient was diagnosed as case of peripartum cardiomyopathy, with type 1 respiratory failure, with aspiration pneumonitis, with hypoxic ischemic encephalopathy, with status epilepticus. She was treated with inotropes, higher antibiotics, steroids, low molecular weight heparin, IV mannitol, IV valproate, midazolam infusion and other supportive heart failure treatment. Unfortunately, she succumbed on day 30 of post-natal period with further deterioration of cardiac function.

Discussion

The incidence and prevalence of PPCM varies widely all over the world. Incidence of PPCM in Indian population is 1 in 1340 live births2 however the exact cause is still unknown. According to previous studies, the risk factors are advanced maternal age, multiple gestation, smoking, obesity, history of pre-eclampsia, gestational hypertension, and eclampsia. Recent evidences show that oxidative stress-prolactin hypotheses, genetic and inflammatory mechanisms are also associated in peripartum cardiomyopathy. Through a genome wide study PPCM has been shown to be strongly associated with a gene on chromosome 123. PPCM is suspected to be an inflammatory response in pregnancy, given the increase in levels of tumor necrosis factor-alpha and interleukin-6 levels4,5. It can also be an autoimmune response associated with fetal cells lodging in the maternal circulation and cardiac tissue6. Evidences regarding nutritional deficiencies, more specifically, Selenium deficiency being a cause of PPCM, are under debate7. Abnormal effect of relaxin hormone secreted in pregnancy, causing excessive relaxation of cardiac muscle has also been associated as underlying cause of PPCM.

The age in the cases discussed ranged from 26 to 32 Years. Breathlessness was the most common complaint and dyspnoea with crepitations was the most frequent sign among them. Risk factors including gestational hypertension and severe pre-eclampsia showed strong association in all the cases. Pulmonary oedema was seen radiologically and NT Pro BNP marker for heart failure was significantly raised in all patients. As discussed, all four cases had different range of left ventricular dysfunction. 3 out of 4 patient showed clinical improvement after treatment with return to normal ventricular function, ensured with regular echo-cardiography. There was 1 maternal mortality among them. The patients were managed in intensive care unit under joint management of obstetricians, neonatologists, intensivist, physicians, and cardiologist. Treatment included conventional pharmacological heart failure therapies, also co-morbidities like anaemia and hypertension were managed accordingly. However, heart transplantation is often the last resort in severe cases, if medical treatments are not successful. Recently introduced PPCM targeted therapies have shown promising outcomes in small trials but require further evaluation like intravenous immunoglobulin, pentoxifylline and bromocriptine. Duration of treatment is that ACEI and beta blockers can be continued for one year depending on patient's clinical and echocardiographic improvement. Limited information and studies are available for appropriate time and mode of delivery in PPCM, the decision should be taken by combined team of obstetrician and cardiologist. In case of haemodynamic instability, immediate delivery should be done by caesarean section irrespective of the gestational age8. More than 50% of women have complete recovery of ventricular function within 6 months of delivery9. Despite treatment, mortality occurs in 10-25% of patients with peripartum cardiomyopathy. Family planning counselling and contraception should be advised always. In case pregnancy is desired, the patient should be advised to wait for at least five years after the ejection fraction has normalized.

Conclusion

These case reports highlight the diagnosis and management of a rare and unavoidable complication of peri partum cardiomyopathy which requires prompt identification. PPCM is a diagnosis of exclusion. Diagnosis requires a heightened awareness among multidisciplinary patient care teams. 2D- ECHO parameters are sensitive predictors in determining prognosis and recovery. As breathlessness is a common symptom in pregnancy and in the initial postpartum period, PPCM is often missed. Risk factors like advanced maternal age, pre-eclampsia and gestational hypertension appear to have strong association. Aggressive medical and obstetric management in tertiary care hospitals is essential for a good outcome. Thus, it is important that obstetricians should be familiar with PPCM and consider it when diagnosing dyspnoeic patients for immediate treatment to prevent potential maternal mortality and morbidity.

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