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Case Report

Dilated cardiomyopathy in pregnancy: a case report

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ABSTRACT

The prevalence of dilated cardiomyopathy (DCM) is 1:2500 among the women of childbearing age. DCM often develops during second trimester or may be present prior pregnancy. We report a successful pregnancy outcome in a young pre-eclamptic woman with DCM who presented with cardiac failure. Multidisciplinary team involved. Anti-failure drugs, antihypertensives and thromboprophylaxis administered. Caesarean section was performed at 30 weeks because of spontaneous onset of labour to avoid undue stress on failing heart. Intra operative and post-operative recovery was uneventful. Follow up showed improvement in ejection fraction. Counselling prior pregnancy, stable cardiac status, regular monitoring, personalised care ensures safe maternal and foetal outcome.

Keywords: Dilated cardiomyopathy, Preeclampsia, Pregnancy

INTRODUCTION

Cardiovascular diseases are seen in approximately 1-4% of all pregnancies and it is one of the most common non-obstetric causes of maternal death. Estimated prevalence of dilated cardiomyopathy (DCM) is 1:2500, among which 50% is idiopathic.[1] Only very few case reports are available till date because pregnancy is not recommended in already diagnosed cases of DCM with low ejection fraction. Incidence of adverse cardiac events ranges from 39%-60% in moderate to severe left ventricular dysfunction. High index suspicion, timely diagnosis and prompt management with the involvement of a multi-disciplinary team are crucial in the management of DCM in pregnancy.

We report a successful pregnancy outcome in a young pre-eclamptic woman with DCM who presented with cardiac failure.

CASE REPORT

A 28-year-old primigravida was referred to our institute with acute onset breathlessness (New York Heart Association class 3) at 30 weeks of gestation. She had spontaneous conception. Her Nuchal Translucency (NT) scan and anomaly scan were normal. She had moderate anaemia (hemoglobin of 8 gm%) at 28 weeks, managed with 1000 mg parenteral iron. She was diagnosed with pre-eclampsia at 28 weeks and initiated on antihypertensives.

On examination, she was found to have heart rate of 118/minute, blood pressure 160/100 mmHg, SpO_2 94% in room air, Respiratory rate 26/min, jugular venous pulse was elevated. She had mild pallor, bilateral pitting pedal edema. Cardiovascular system examination unremarkable. Respiratory system showed bilateral basal crepitations. Echocardiography (ECHO) showed left ventricular (LV) dilatation, global hypokinesia of LV, severe systolic dysfunction with ejection fraction (EF) of 20% and moderate to severe pulmonary artery hypertension (PAH). Diagnosis of DCM was made.

She was started on parenteral antihypertensives and inotropes in cardiac intensive care unit. Her blood investigations revealed haemoglobin 9.9 g/dl, platelet 160000, coagulation profile, liver function within normal range. Serum electrolytes showed hyperkalaemia (6.1

mmol/l), elevated serum urea (72 mg/dl), creatinine (1 mg/dl) and TSH (8.38 miu/l). Nephrologist opinion sought for elevated renal parameters with hyperkalaemia. Electrolyte imbalance corrected, and was decided to rule out glomerular pathology in postpartum period. She was offered mechanical and pharmacological thromboprophylaxis. Bedside scan showed appropriately grown fetus with borderline oligohydramnios and normal doppler. Blood pressure was under control with antihypertensives.

Multi-disciplinary team (obstetrician, cardiologist, neonatologist and anaesthetist) decided for initial cardiac stabilization followed by delivery. Prophylactic steroids administered for fetal lung maturity. On day three of hospitalization, she went into spontaneous labor. Decision on emergency caesarean section was made to avoid stress on failing heart. It was performed under epidural anaesthesia with inotropic support after obtaining high risk consent. Intraoperative period was uneventful. She delivered a preterm boy baby with 1.3 kg weight and good APGAR. Baby admitted in neonatal intensive care unit (NICU) for preterm care.

Inotropes were tapered and stopped on postoperative day two. Thromboprophylaxis continued. A bedside ECHO performed showed EF of 29%. Anti-hypertensives were tapered. She was discharged on postoperative day seven in stable condition with angiotensin converting enzyme (ACE) inhibitors, beta blockers and thromboprophylaxis. Counselling regarding risk of recurrence and relatively high mortality rate in future pregnancies was done. She was followed-up at six weeks, initiated on contraception. ECHO done at six months showed improvement in EF to 53%.

DISCUSSION

Cardiovascular diseases affect 1-4 percent of pregnant women, and it is the most prevalent non-obstetric cause of maternal death. Cardiomyopathy is described as structural and or functional anomaly of the myocardium in the absence of coronary artery disease, hypertension, valvular disease, or congenital heart disease sufficient to cause the observed myocardial abnormalities.[2] Because pregnancy is not indicated in known heart disease with an EF of less than 30%, it is rarely described in pregnancy and only a few literatures exist.

Cardiomyopathies are a group of illnesses that are either idiopathic (50%) or hereditary (35%), with PPCM being the most frequent cardiomyopathy seen during pregnancy. PPCM appears a month before or five months after delivery, whereas idiopathic DCM usually appears in the first trimester.[1] Hospitalization for cardiomyopathy in pregnancy was reported to be 0.46 per 1000 in a cross-sectional study conducted in the United States.[3] Viral infections, inflammatory disorders, arrhythmia and storage disorders are considered as risk factor for DCM and it accounts for 11% of maternal death.[4]

Most women present with features of cardiomyopathy in late trimester; however, she presented in early third trimester with features of heart failure. Adverse events like Atrial fibrillation (AF), ventricular tachycardia (VT), transient ischaemia or cardiac arrest secondary to cardiomyopathy add to maternal mortality. The signs of cardiac impairment should not be confused with those of pregnancy, because a delay in diagnosis can have a negative impact on the outcome.

EF of less than 30%, a higher degree of NYHA class (III or IV), first time diagnosing in pregnancy and a previous cardiac event are considered as poor prognostic factors in DCM.[1] In our case, the diagnosis was first made at 30 weeks of gestation and she falls under NYHA III. Pregnancy is contraindicated in WHO class IV cardiac diseases and termination is advised in case if women conceive.

The ROPAC study mentioned the risk of low-birth-weight babies in women using beta blockers and teratogenic effect of ACE inhibitors.[5] Beta blockers are recommended in case of arrhythmia, diuretics in case of cardiac failure and thromboprophylaxis to reduce thrombotic risk. The fact that most of the anti -failure medications including Renin Angiotensin-Aldosterone-System (RATS) are contraindicated in pregnancy will make the situation more troublesome. A multidisciplinary team with optimization of medication and timely intervention is crucial in this scenario. Proper counselling of the family and high-risk consent is mandatory.

Mazor and colleagues reported a case of idiopathic DCM first time diagnosed in second trimester of pregnancy with EF of more than 35%, and managed conservatively, similar to Chan F and colleagues.[6,7] Because the diagnosis was made in the early second trimester in both cases, the patients and family were offered termination of pregnancy but they refused. In the second case, an emergency caesarean section was performed under general anaesthesia at 31 weeks due to severe heart failure, and the patient suffered a thromboembolic stroke in the postoperative period despite being on therapeutic anticoagulation. Caesarean section (CS) is reserved only for obstetric indications and for certain cardiac circumstances in pregnant women with heart disease. We chose caesarean section for our patient in order to reduce the stress on failing heart. As per current literatures, regional anaesthesia is preferred for caesarean section complicated by heart disease as slow titrated epidural or even spinal with slow incremental dosage.[8] Maintenance of systemic vascular resistance (SVR) and cautious monitoring of vitals are crucial. Epidural anaesthesia was preferred by our anaesthesia team and the intra operative period was uneventful.

Few studies and case reports have been published on the co-existence of pre-eclampsia and PPCM. Behrens et al. discussed correlation between hypertensive disorders of pregnancy (HDP) and PPCM in a national cohort study.

The study showed increased incidence of PPCM with increase in severity of HDP.[9] HDP regardless of severity, have been linked to the development of cardiovascular disorders, particularly cardiomyopathies, beyond peripartum or later in life, even when blood pressure is normal in the post-partum period, implying a similar aetiology of these two conditions.[10] The evidence that pre-eclampsia during pregnancy causes remodelling of the left ventricle and diastolic dysfunction (altered circulatory structure and function) strengthens this link.[11]

Recent studies point towards the shared genetics (TTN gene mutation) between DCM, PPCM and pre-eclampsia., but more extensive studies are needed in this field.[12,13] A more recent study published in 2021 discussed a possible common aetiopathogenesis of pre-eclampsia and PPCM explained by elevated soluble FMS like tyrosine kinase (sFlt-1) in both.[14] Heart failure caused by pre-eclampsia and DCM must be distinguished for optimal patient management, and echocardiography is the investigation of choice in such cases.

Antenatal management of DCM is challenging because of foetotoxicity of cardiac medications, increased risk of thromboembolism with a failing heart, decision on timing of delivery, anaesthesia risk, preterm risk, risks of sudden intrauterine foetal demise and even maternal death. In our case it was further cumbersome because of the co-existence of severe pre-eclampsia. Involvement of a multidisciplinary team with aggressive medical and obstetric management, good anaesthetic care and preterm neonatal care made the outcome of this high-risk pregnancy successful.

CONCLUSION

A vigilant attitude is necessary for the timely diagnosis and management of DCM in pregnancy with severe pre-eclampsia. Shared decision making and optimization of drugs at the right time, involvement of a multidisciplinary team plays an important role in successful maternal and neonatal outcome.

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