A rare case of peripartum cardiomyopathy with shock and fetal demise in utero

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Abstract: Peripartum Cardiomyopathy (PPCM) is an uncommon disorder presenting as heart failure secondary to left ventricular systolic dysfunction. It presents in the last month of pregnancy or within 5 month of delivery with no underlying cardiac pathology. Its diagnosis is often delayed because its symptoms closely resemble those within the normal spectrum of pregnancy and the postpartum period. When PPCM is misdiagnosed or its diagnosis is delayed, the consequences for patients are deadly: The disorder carries a high mortality rate.

Keywords: Peripartum cardiomyopathy, High risk pregnancy.

INTRODUCTION
Peripartum cardiomyopathy (PPCM) is marked left ventricular systolic dysfunction that presents towards the end of pregnancy through the first five months post delivery, complicating obstetric as well as anaesthetic management. The strict time limit used in their diagnostic criteria was intended to exclude congenital and acquired causes of heart failure that usually manifest by second trimester. Overall, recent reports from various parts of the World show an incidence of 1 in 1,485 to 4,000 live births and the trend is increasing [1].

CASE REPORT
A 20 year old primigravida at 36 week 3 day period of gestation presented to Adesh Institute of Medical Sciences and Research, Bhucho emergency unit with history of acute onset of breathlessness 2 days ago. She was referred from a local hospital in shock with mechanical ventilation and inotropitic support. Referral report suggested acute renal failure and intrauterine foetal demise. There was history of one episode of generalized tonic clonic convulsions one day ago. Upto 36 weeks, the antenatal period had apparently been normal.

On examination, patient was intubated on assisted ventilation. Her blood pressure was 120/80 mm Hg and pulse rate was 140/min. Obstetric examination suggestive of term sized relaxed uterus with absent fetal heart sounds. On Per vaginum examination cervix was unfavourable.

On Investigating, blood counts were normal, liver function test and renal function tests were deranged. Coagulation profile was normal. ECG findings were suggestive of sinus tachycardia, left axis deviation, antero-septal infarct possibly acute inferolateral injury pattern. On ECHO, there was left ventricular systolic dysfunction with ejection fraction of 20%.

Fig-1: Chest X Ray showing dilated cardia
A multidisciplinary team consisting of obstetricians, physician and anaesthesiologists was swept into action. Removal of dead foetus from uterus was considered of paramount importance. In view of decompensated state of patient, emergency cesarean section under general anaesthesia was conducted. Intraoperative and postoperative fluid management was guided by central venous pressure monitoring. Post operatively she received intravenous diuretics, digoxin, intravenous antibiotics and low molecular weight heparin. She was on supportive therapy with ventilation and inotropes which were gradually weaned off.

ACE inhibitors (Ramipril) and cardioselective beta blockers (Carvedolol) were started in intensive care unit. Over a period of two weeks her cardiac function was found to improve gradually. There was no post operative complication and patient was discharged on ramipril and metoprolol with EF 55% at time of discharge. Ethical guidelines of institution were followed and permission obtained from institution.

**DISCUSSION**

Peripartum cardiomyopathy is a pregnancy associated idiopathic cardiomyopathy secondary to marked left ventricular systolic dysfunction. Diagnostic criteria for peripartum cardiomyopathy includes development of heart failure in the last month of pregnancy or within the 5 months following delivery, absence of determinable etiology of heart failure, absence of demonstrable heart disease prior to the diagnosis of heart failure, left ventricular ejection fraction <45%, or fractional shortening <30% or both.

PPCM is a significant cause of morbidity and mortality in pregnant patients. In a study occurring in series of 182 patient with PPCM, the rate of major adverse events was reported to be 25%, with 80% of these event occurring in the first six months after the diagnosis [2]. Mortality rates reported to date are around 15%, which is less than that associated with other forms of cardiomyopathy [3, 4]. In a series of 17 cases of death in PPCM patients, 18% of deaths occurred within one week of delivery and 87% within 6 months [5].

Overall, recent reports from various parts of the World show an incidence of 1 in 1,485 to 4,000 live births and the trend is increasing [1]. Only 5% of the cases of PPCM present in antepartum period, most are seen in postpartum period. Historically, advanced maternal age and multiple pregnancies are risk factors; contemporary trends show that there is increasing incidence in young primigravida as in this case. This entity presents a diagnostic challenge because many women in last month of normal pregnancy experience dyspnea, fatigue and pedal edema, symptoms identical to early congestive heart failure. There are no specific criteria for differentiating subtle symptoms of heart failure from normal late pregnancy. It is important that high index of suspicion be maintained to identify rare case of PPCM.

**CONCLUSION**

Prompt recognition and treatment of the disease can save the patient. Multidisciplinary team approach and intensive targeted therapy are necessary to achieve an optimal outcome.

**REFERENCES**


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