

CASE REPORT

“UNUSUAL PRESENTATION OF PERIPARTUM CARDIOMYOPATHY (PPCM)- A CASE REPORT”

Saubhagya Kumar Jena¹, Soumya Samal², Suresh Kumar Behera³, Basanta Kumar Behera⁴.

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ABSTRACT: Peripartum cardiomyopathy (PPCM) is a rare but sometimes life-threatening form of dilated cardiomyopathy. The exact cause is unclear. It is associated with excess morbidity and mortality in women of childbearing age. Incidence of PPCM ranges from 1 in 2000 to 4000 pregnancies. The diagnostic criteria are onset of heart failure in the last month of pregnancy or in first 5 months postpartum, absence of determinable cause for cardiac failure, and absence of a demonstrable heart disease before the last month of pregnancy. There must be echocardiographic features of left ventricular dysfunction. The known risk factors include multiparity, twin births, advanced maternal age, preeclampsia, gestational hypertension, and black race. The usual clinical presentation of patients with peripartum cardiomyopathy is similar to that of patients with systolic congestive heart failure. Treatment is limited to use of drugs for symptomatic control, although recent studies indicate the usefulness of new drugs. About half the patients of peripartum cardiomyopathy recover without any complications. The prognosis is poor in patients with persistent cardiomyopathy. Persistence of disease after 6 months indicates irreversible cardiomyopathy and portends worse survival. Recurrence in subsequent pregnancies remains high. In this case report, we describe the unusual presentation of PPCM in a young woman with term pregnancy.

KEY WORDS: Anaesthesia, Heart failure, Peripartum cardiomyopathy, Pregnancy.

INTRODUCTION: Heart diseases of varying severity complicate 1% of pregnancies although the haemodynamic changes of normal pregnancy can mimic those of heart disease. ⁽¹⁾ These include rheumatic heart diseases (RHD), congenital heart diseases (CHD), cardiomyopathies, hypertensive heart diseases and coronary arterial disease (CAD) etc. ⁽¹⁾ Out of different types of cardiomyopathies like hypertrophic, dilated and restrictive, dilated cardiomyopathy is more common during pregnancy. ^(1, 2) Peripartum cardiomyopathy (PPCM) is a rare dilated cardiomyopathy causing heart failure in women in late pregnancy or early postpartum. ⁽³⁾ The incidence of PPCM has recently been estimated to be one in every 2000 to 4000 deliveries but it varies in different parts of world. ⁽⁴⁾ Its etiopathogenesis is still poorly understood, but recent evidence supports inflammation, viral infection and autoimmunity as the leading causative hypotheses.⁽³⁻⁵⁾ Multiparity, twin births, advanced maternal age, preeclampsia, gestational hypertension, and black race are some known risk factors. ⁽⁶⁾ Clinical presentation of PPCM is similar to that of systolic heart failure from any cause, and it can sometimes be complicated by a high incidence of thromboembolism. ⁽⁷⁾ The patients usually present with dyspnea (90%), tachycardia (62%), and edema (60%). ^(6, 7) Some case studies also cite unusual presentations, including multiple thromboembolic events and acute hypoxia. ⁽⁸⁾ We report an unusual form of presentation in a case of PPCM.

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The Case: A 25 year old female, gravida 2, para 1, 40 weeks pregnancy with previous caesarean section was referred from outside hospital during emergency hours in shock. She was admitted in our hospital at 18.12 hours. Her primary caesarean section was done for cephalo pelvic disproportion (CPD). She had an uneventful intra and postoperative period during that time. This time caesarean section was planned due to type III placenta praevia. After spinal anaesthesia, she went to cardiovascular collapse. The operation was stopped and she was referred to our hospital after resuscitation.

She was complaining of headache, shortness of breath and palpitation. There was no personal and family history of cardiovascular disease. She had an uneventful antenatal period. On examination, she was disoriented, dyspneic with sweating over face. Her peripheral pulses were not palpable, blood pressure was not recordable and respiratory rate was 32/minute. She was afebrile. Cardiovascular system examination revealed regular heart rate of 100 beats per minute, loud first and second heart sound with pansystolic murmur over apex. On auscultation of chest, there was vesicular breath sound with bilateral basal crepitations. On abdominal examination there was a sub-umbilical midline scar with term size relaxed uterus. The fetus was in longitudinal lie and cephalic presentation. Foetal head was 5th/5th palpable per abdomen. Foetal heart rate was 160 beats per minute and regular. On inspection of pelvic area there was no bleeding. She was on indwelling catheter with 300ml of concentrated urine.

Blood investigations revealed B +ve blood group, haemoglobin 13.3 gm%, PCV 39.3%. Antibody test for VDRL, HBsAg and HIV were non-reactive. Bleeding time 2.3 minute, clotting time 3.4 minute and total platelet count was 2.3 lakhs/cubic mm. PTT was 14.9 seconds (control 13.5 seconds), INR 1.1, aPTT 29.5 seconds (control 26 seconds). Serum sodium was 135 mEq/ L and potassium was 3.2 mEq/ L. There was no abnormality in urine examination.

ECG showed sinus rhythm, ST depression with T inversion in V₁ to V₆ leads and ST depression in I, V₅, V₆ leads. In echocardiography there was severe hypokinesia of basal and mid septum, left ventricular dysfunction, ejection fraction was 35%, and mild mitral regurgitation suggesting cardiomyopathy. Ultrasonography of gravid uterus showed a single live fetus with cephalic presentation and normal heart rate. There was type III placenta previa and liquor volume was reduced.

A diagnosis of peripartum cardiomyopathy was made after consulting cardiologist. Consultation of neonatologist and anesthesiologist was also taken. She was kept in cardiac position with 100% oxygen via venturi mask. Dopamine infusion was started at the rate of 5 µgm/kg/min. Right internal jugular venous cannulation and arterial catheterization was done. Central venous pressure was 5 cm of water and arterial pressure was 60/40 mm of Hg. Intravenous colloid was started and dopamine infusion was increased to 10 µgm/kg/min. The arterial pressure gradually increased to 80/50 mm of Hg. Then she was transferred to operation theatre for caesarean section.

General anaesthesia was induced using 200 mg intravenous thiopental and 100 mg of intravenous succinylcholine with cricoid pressure and tracheal intubation in presence of cardiologist. A female child of weight 3.2 kg was delivered by lower segment caesarean section (LSCS) at 19.47 hours. Apgar score at 1 and 5 minute was 3 and 7 respectively. There was type II posterior placenta previa and thick meconium stained scanty liquor. Fortunately there was no postpartum haemorrhage.

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She was kept in intensive care unit (ICU) during postoperative period and managed with broad spectrum antibiotics, intravenous fluids and analgesics. Her peripheral pulse was not palpable and blood pressure was not recordable. Dobutamine was started at 10 µgm/kg/min. She was given digoxin, furosemide and L-carnitine as prescribed by the cardiologist. Injection heparin was also given to prevent thrombosis. Gradually her peripheral pulses became palpable and blood pressure was recordable. On third postoperative day she developed carpopedal spasm. The total serum calcium and total serum protein was 8.6 mg/dl and 3.8 gm/dl respectively. It responded to treatment with intravenous calcium gluconate. Protein supplementation was also given. Suture removal was done on 7th day. Repeat echocardiography showed improving left ventricular function and ejection fraction. She was discharged on request on 10th day with the advice for regular follow up.

DISCUSSION: PPCM represents less than 1% of all cardiovascular diseases that occurs during pregnancy. Clinicians should consider this as a differential diagnosis in any peripartum patient with unexplained cardiovascular collapse. The National Heart, Lung and Blood Institute (NHLBI), with the National Institutes of Health (NIH), published diagnostic criteria for PPCM. The criteria include: (a) onset of heart failure signs and symptoms in the last month of pregnancy or within 5 months postpartum; (b) LV systolic dysfunction with ejection fraction (EF) measured $\leq 45\%$ or LV end diastolic dimension 2.7 cm/m^2 ; (c) no evidence of pre-existing heart disease prior to peripartum symptom onset; (d) no other identifiable causes of heart failure. ^(9,10)

Though the clinical presentation was unusual, our case fulfilled the clinical criteria for PPCM. Her rapid cardiovascular decline could have resulted, in part, from abrupt changes in ventricular loading after initiation of spinal anesthesia. There are reported incidences of sudden cardiovascular collapse after spinal anaesthesia for caesarean section and postoperative investigations confirming the diagnosis as PPCM. ⁽¹¹⁾ Alternative explanations for cardiovascular collapse such as anaphylactic shock, amniotic fluid embolism, and anesthesia-associated respiratory arrest were possible but not fully consistent with this patient's overall clinical condition during and after childbirth. ⁽¹²⁾

Obstetricians should be aware of PPCM, an uncommon but life-threatening condition. Once suspected the diagnosis should be confirmed with echocardiography after excluding other common causes of sudden cardiovascular collapse. ⁽¹⁰⁾ Treatment of PPCM is similar to that for other types of congestive heart failure. The combination of digoxin, diuretics and sodium restriction, beta blockers and after load reduction forms the cornerstone of therapy. ⁽¹³⁾ Anticoagulants should be considered in these patients as they are highly predisposed to thromboembolic phenomena. Patients with severe forms of heart failure may require more aggressive management in an ICU with monitoring of arterial blood pressure (ABP), central venous pressure (CVP), a pulmonary artery catheter (PAC) and echocardiography along with inotrope, vasodilator and ventilator therapy. Though some trials had reported benefits of pentoxifylline, intravenous immunoglobulin, and bromocriptine, no specific treatment has been identified to significantly alter the morbidity of PPCM. ⁽¹⁴⁾ Cardiac transplantation should be considered for cases who fail to maximal medical management.

Intensive fetal and maternal monitoring is required during the antepartum period. Induction of labour should be considered if the patient's condition deteriorates despite maximal medical management. A multidisciplinary approach involving an obstetrician, cardiologist, anesthesiologist and perinatologist may be required to provide optimal care to such patients. Regional analgesia reduces the cardiac stress of labor pain while application of outlet forceps or a vacuum device can

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minimize the cardiac stress of the second stage of labor. Caesarean section is best done for obstetric indications as well as in severe decompensated situations. Both general anaesthesia (GA) and regional anaesthesia (RA) can be used for caesarean section. Single-shot spinal anaesthesia is not preferred because of severe consequence like cardiac arrest and pulmonary edema. Epidural anaesthesia (EA) under invasive monitoring is a safe and effective method. Combined spinal-epidural (CSE) analgesia & continuous spinal analgesia (CSA) are also safer alternatives. Following delivery, these patients need monitoring in an ICU for early detection and management of associated complications. Counselling should be done about breast feeding and future pregnancy at the time of discharge. Because of the high risk of thromboembolism with oral contraceptive pills (OCP), barrier contraceptives are better options for family planning. Surgical sterilization should be considered if family is completed.

Although PPCM usually presents with features of systolic heart failure, rarely it can present with sudden cardiovascular collapse. It usually happens during labour or after anaesthesia. High degree of suspicion with intensive multidisciplinary approach can save the life of those patients.

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AUTHORS:

1. Saubhagya Kumar Jena
2. Soumya Samal
3. Suresh Kumar Behera
4. Basanta Kumar Behera

PARTICULARS OF CONTRIBUTORS:

1. Assistant Professor, Department of Obstetrics and Gynaecology, AIIMS, Bhubaneswar.
2. Assistant Professor, Department of Anaesthesiology, IMS & SUM Hospital, Bhubaneswar.
3. Assistant Professor, Department of Cardiology, IMS & SUM Hospital, Bhubaneswar.

4. Professor, Department of Community Medicine, KIMS, Bhubaneswar.

NAME ADDRESS EMAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Saubhagya Kumar Jena
AIIMS, Bhubaneswar.
At: Sijua, Po: Dumduma,
Bhubaneswar, Odisha
Pin-751019
E-mail: drsau@rediffmail.com

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