

PERIPARTUM CARDIOMYOPATHY: A CASE REPORT

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About The Author: Afzal Azim is currently working as Associate Professor in the Department of Critical Care Medicine. He did his Postgraduation in Anesthesiology in 1999 from PGIMER Chandigarh. Then he did Post-doctoral certificate course (PDCC) in Neuroanaesthesiology and intensive care from SGPGIMS Lucknow. After finishing Senior residency he joined the Department of Critical Care Medicine at SGPGI, Lucknow in 2002 (first in any Government institute) It is a closed model 12 bedded intensive care unit that caters to both medical and surgical patients. In 2006 he had the opportunity to work as fellow at Westmead hospital, Sydney Australia. His special areas of interest are Antibiotic policies and Infection control in critically ill. He has worked on research project related to colonisation pattern in critically ill patients during their ICU stay.



Abstract:

Peripartum cardiomyopathy (PPCM) is an uncommon but life threatening disease that affects women in the last month of pregnancy or within the first five months after delivery. Very few Indian case reports are available. However, it is essential for the practitioner dealing with such population to have a high degree of clinical suspicion for early diagnosis and management. Echocardiography is used to diagnose this entity and monitor the therapy. Treatment options are available despite its somewhat poor overall prognosis. We present a case report of a 23-year-old woman who presented four days post-partum with respiratory distress and early echocardiography helped in diagnosing PPCM.

Key words: Peripartum cardiomyopathy, rarely reported, clinical suspicion, Echocardiography

Peripartum cardiomyopathy (PPCM) is a rare but life-threatening form of heart failure affecting women in the last month of pregnancy or the first five months post-partum without any previously recognizable heart disease^{1,2}. Its etiology is poorly understood. The present report

describes the case of a woman presenting with cardiac failure after emergency caesarean section for bleeding per vaginum and was diagnosed as a case of Peripartum cardiomyopathy.

CASE REPORT:

A 23 year old primi gravida with no comorbidities and uneventful antenatal period presented to our emergency department four days after undergoing emergency LSCS at 34 weeks of gestation with history of bleeding per vaginum in a private hospital. In the immediate postoperative period, she developed hemorrhagic shock with non-oliguric acute renal failure. She received multiple transfusions of blood and blood products to stabilize her hemodynamics and bleeding stopped. On the third postoperative day she developed respiratory distress for which she was referred to us for further management.

On examination, she was drowsy, mild pallor was present and she was in respiratory distress. Her blood pressure was 90/66 mm Hg, heart rate was 126 beats/minute, and the respiratory rate was 34/minute. She was afebrile. Chest auscultation revealed significant bilateral basal crepitations. The cardiovascular examination was normal. Abdominal examination was unremarkable. She was admitted in intensive care unit with a provisional diagnosis of post-partum hemorrhage with hemorrhagic shock with multiorgan failure. Her ICU admission APACHE-II was 19 and SOFA was 09.

Initial laboratory investigations were normal except for the serum creatinine level of 4.6 mg/dl and thrombocytopenia (90,000/cmm). ECG showed sinus tachycardia and Chest X-ray was suggestive of bilateral diffuse infiltrates with minimal bilateral pleural effusion. Invasive monitoring was instituted in the form of invasive blood pressure monitoring and central venous pressure monitoring. Arterial blood gas analysis was suggestive of severe metabolic acidosis ($\text{HCO}_3 = 11$) and hypoxemia (PaO_2 of 66 on fiO_2 of 40% through venturi mask). She was intubated and mechanical lung ventilation was started. Her opening central venous pressure was high (14 mmHg). She had high lactates (6.6mmol/L) suggestive of poor perfusion and low cardiac output. Vasopressor support was started in the form of Noradrenalin (0.5 to 1.0 micrograms/kg/ min) to maintain mean arterial pressure > 65 mmHg and echocardiography was planned. Echocardiography revealed global hypokinesia with ejection fraction of 30% and fractional shortening (FS) of 24%. Left ventricular end diastolic (LVED) dimension was 3.2 cm/m² with mild mitral regurgitation. There was no evidence of pulmonary

embolism. Her D-dimer was positive and Doppler ultrasound of both lower limbs was negative for deep vein thrombosis. Based on clinical presentation and echocardiography a diagnosis of Peripartum cardiomyopathy was made.

To augment the cardiac contractility dobutamine infusion was started. Other ICU supports included diuretics, analgesics and sedation, prophylaxis for stress ulcer, deep vein thrombosis prophylaxis and broad spectrum antibiotics. After a period of 48hr she showed improvement in acid base milieu, lactates normalized and noradrenalin was tapered off. Her renal parameters also improved gradually. She never required dialysis during ICU stay. After 5 days of dobutamine therapy she was switched over to oral digoxin therapy. Her respiratory parameters and chest radiology improved subsequently and she was weaned off from mechanical ventilation after 10 days of ICU stay. On day 16 of her ICU admission a repeat echocardiography showed persistent global hypokinesia, ejection fraction of 35%, FS 24.4%, LV end diastole dimension 3 cm/m². It was suggestive of minimal improvement as compared to echocardiography done at ICU admission. Cardiologist opinion was taken and she was discharged on digoxin and carvedilol after an ICU stay of 20 days.

She was advised to avoid any future pregnancy and to continue regular medical follow-up. Her echocardiography during follow-up after six months, showed normal left ventricular size and contractility with normal ejection fraction and FS of 38.6%.

DISCUSSION:

We report a case of PPCM which is very rarely reported in Indian literature. Our centre is a 700 bedded tertiary care centre and our ICU is a 12 bedded general purpose ICU which caters to sick patients requiring life support therapies. Informed consent was taken from the patient before writing the report.

PPCM is defined on the basis of four criteria which are: (a) development of cardiac failure in the last month of pregnancy or within five months of delivery; (b) absence of an identifiable cause for the cardiac failure; (c) absence of recognizable heart disease prior to the last month of pregnancy; (d) left ventricular systolic dysfunction demonstrated by classic echocardiographic finding such as left ventricular ejection fraction of less than 45 percent, fractional shortening of less than 30 percent on an M-mode echocardiographic scan, or both, and a left

ventricular end diastolic dimension of more than 2.7 cm per square meter of body-surface area^{3,4}.

The true incidence of PPCM is unknown, occurring in around one in 2,400 to one in 15,000 births in the western world^{1,5-9}, although its incidence is higher in African countries¹⁰. There is paucity of Indian literature regarding its incidence. A few case reports have been reported in India¹¹⁻¹⁵.

Although the etiology of PPCM remains unclear, a number of potential risk factors for this disorder have been proposed. Among these factors included are multiparity, advanced maternal age, multifetal pregnancy, pre-eclampsia, gestational hypertension and women of African descent³. Other risk factors include association with maternal cocaine abuse or selenium deficiency, long-term (more than four weeks) oral tocolytic therapy with beta adrenergic agonists such as terbutaline³. However, the disease can occur in women without these risk factors as was seen in our case.

A number of possible causes have been proposed for PPCM, including myocarditis, abnormal immune response to pregnancy, maladaptive response to the hemodynamic stresses of pregnancy, stress-activated cytokines³. In addition, there have been a few reports of familial PPCM¹⁶.

Diagnosis of PPCM requires exclusion of other causes of cardiomyopathy before it is considered. Our patient did not have any of the manifestations neither in the past nor during her ante-partum period. Though hemorrhagic shock was our primary diagnosis, a high CVP, low systemic blood pressures (90/66 mm Hg) and high lactates (6.6 mmol/L) were suggestive of cardiogenic shock in our patient. This prompted us for early echocardiography which was suggestive of cardiac failure. Septic shock induced myocardial dysfunction was ruled out because she was afebrile, her total leukocyte counts were normal (TLC-6,500) and she did not require fluid resuscitation or prolonged vasopressor support to maintain pressures. Noradrenalin was rapidly tapered off after initiation of dobutamine.

Treatment of PPCM is similar to that for other types of congestive heart failure. The combination of digoxin, diuretics and sodium restriction, anticoagulation, beta blockers and after load reduction forms the cornerstone of therapy¹⁷. All these interventions were done in our patient to which she showed a good clinical response. After initial dobutamine

infusion therapy she was switched over to oral digoxin therapy. Subsequently diuretics and carvedilol were added.

Women who fail maximal medical management may be candidates for cardiac transplantation¹⁸. Though this is far away in Indian setup, our patient fortunately responded to the usual line of management. The prognosis for women with PPCM appears to depend on the normalization of left ventricular size and function within six months after delivery.^{3,19} About half of the patients of PPCM recover without any complications¹⁹. Most hearts that are destined to recover normal function probably do so within six months from the time of diagnosis^{1,20}. Persistence of the disease even after six months indicates irreversible cardiomyopathy and portends worse survival. The mortality estimates for patients with PPCM in the United States range from 25 to 50 percent with most deaths occurring within the first three months post partum^{1,20}. Death is usually caused by progressive pump failure, arrhythmias or thromboembolic events^{2,21}. Although, our patient seemed to have full clinical improvement after six months with normalization of echocardiographic findings, concern remains because we could not have her further follow up. This concern evolves because studies like Lampert et al showed women with PPCM who have regained normal resting left ventricular size and performance have decreased contractile reserve as revealed by dobutamine challenge test²². Therefore, subsequent pregnancies, if they cannot be avoided, should be managed in a tertiary care centre with collaboration of a high-risk perinatal center^{1,3}

CONCLUSION:

Our report is one of the rare case report published in Indian literature. High degree of clinical suspicion supported by early echocardiography is highly important to diagnose this entity which can have a poor outcome despite optimal medical management.

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