

CASE REPORT

Unrecognised peripartum cardiomyopathy will have dire consequences

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ABSTRACT

We report an undiagnosed case of Peripartum cardiomyopathy (PPCM) in our tertiary care hospital, who presented for an elective cesarean section (CS) with cough and orthopnea in her late gestational period. She was treated for upper respiratory tract infection; whereas her heart failure leading to pulmonary edema causing cough and orthopnea, remained unidentified and thus uninvestigated. The disease was only diagnosed during the postoperative period when she suffered from cardiac arrest and had to be ventilated mechanically. She was effectively managed after correct diagnosis; weaned successfully off the ventilator and transferred to the Obstetrics/Gynecology ward in a stable condition.

Key words: Peripartum cardiomyopathy; Paroxysmal nocturnal dyspnea; Orthopnea; Cesarean section

Citation: Shafiq M, Khan RA, Khan A, Shah A, Hussain S. Unrecognised peripartum cardiomyopathy will have dire consequences. *Anaesth Pain & Intensive Care* 2013;17(2):195-197

INTRODUCTION

Peripartum cardiomyopathy (PPCM) is a disease which affects the parturient during late pregnancy or immediately after delivery.¹ The early signs and symptoms of PPCM are weight gain, pedal edema, dyspnea on exertion, orthopnea, paroxysmal nocturnal dyspnea and persistent cough, which are often seen in normal pregnancy due to physiologic changes. A low incidence of the disease, lack of awareness among health caretakers and lack of antenatal care due to poverty in the third world countries often lead to late/ missed diagnosis and treatment. A case report of this disease is presented which was missed being timely diagnosed and resulted in serious consequences for the patient, including need of mechanical ventilation. A strong suspicion of the condition is stressed in peripartum patients with moderate to severe dyspnea and persistent cough.

CASE REPORT

A 28 year old multigravida previously in good health, presented with cough to ER of our hospital in her late third trimester of pregnancy. She was sent home on antibiotics and cough suppressants. Few days later, she was admitted for emergency CS due to fetal distress. She

had two previous uneventful lower segment CS under GA. Her cough was unrelieved and so was her dyspnea. She was prepared for CS. In the Operating Room (OR) she had productive cough, dyspnea and required supplemental oxygen to correct low peripheral oxygen saturation. Spinal subarachnoid anesthesia was administered with standard dose of intrathecal Bupivacaine spinal. The patient had a systemic BP drop which responded partially to I/V fluids and inotropes. The operation went uneventful. A male baby with Apgar score of 10/10 was delivered. She was transferred to Surgical ICU for post-operative care. Her intractable cough and low oxygen saturation were her immediate issues. There were bilateral crepitations upon chest auscultations. Morphine Sulphate boluses in small doses and supplemental O₂ was administered. There was little improvement. ABGs showed metabolic acidosis, hypoxemia and upper normal PaCO₂ values. Chest x-ray had findings suggestive of pulmonary edema. Noninvasive ventilation with CPAP mask produced subjective relief but had no significant effect on the peripheral oxygen saturation. Three hours later the patient started showing signs of exhaustion. A decision was taken to invasively ventilate her. Unfortunately the patient suffered cardiac arrest during endotracheal intubation. It was managed according to the standard protocol. Stable cardiac

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rhythm was obtained in approximately 15 minutes and the patient was placed on the mechanical ventilator. 2-D echocardiography revealed dilated cardio-myopathy with EF of 33%. A cardiologist was called who established the diagnosis. The patient was weaned followed by tracheal extubation two days later when she fulfilled laid down criteria for cessation of ventilatory support/ extubation. She required cardiac medications, supplemental O₂, anti-biotics, nutrition, physiotherapy and psychological support. The patient was shifted to her ward after another three days. A repeat echocardiography revealed unremarkable improvement in cardiac functions. She was sent home on cardiac medications and advice to get reviewed both by gynecologist and cardiologist regularly. She was encouraged to breast feed her newborn baby.

DISCUSSION

The Heart Failure Association of the European Society of Cardiology Working Group on PPCM describes peripartum cardiomyopathy as “an idiopathic cardiomyopathy presenting with heart failure secondary to left ventricular systolic dysfunction towards the end of pregnancy or in the months following delivery, where no other cause of heart failure is found. It is a diagnosis of exclusion. The left ventricle may not be dilated but the ejection fraction is nearly always reduced below 45 %”.² Before establishing the diagnosis of PPCM some peripartum complications that result in depressed cardiac functions like pneumonia, pulmonary thrombo-embolism, amniotic fluid embolism, myocardial ischemia etc. need exclusion. The incidence of PPCM in United States is around 1:3,000 live births; but there are racial differences.³ The first case series of patients with PPCM was published in the year 1971 by Demakis *et al.*⁴ Pandit *et al* report it as 1 case per 1374 live births from Manipal, South India.⁵ Hasan *et al* state this as 1 in 837 deliveries in their analysis of 32 cases from October 2003 to September 2007 from Karachi, Pakistan.⁶ The etiology of PPCM remains unclear; however some risk factors have been identified. The strongest risk factor is African-American ethnicity.³ Other reported risk factors are age, pregnancy-induced hypertension or preeclampsia, multiparity, multiple gestations, obesity, chronic hypertension and the prolonged use of tocolytics. Some more causes which are being investigated include nutritional, infectious, and genetic, which hopefully will lead to more targeted treatment and better outcome.

The clinical presentation of patients with PPCM resembles systolic heart failure secondary to cardiomyopathy of other types. In many cases, the patient themselves and their attending doctors may believe that these symptoms are either due to gravidity approaching term; or due to the recent child birth causing physical and emotional stress. A breathless woman towards the end of pregnancy or in early

post natal period must be screened by Electrocardiography (ECG), plasma concentration of B-type natriuretic peptide (BNP) or N-terminal pro-BNP (NT-pro BNP) and cardiac imaging . A cardiology consultation is mandatory if there is any abnormality. Left Ventricular Ejection Fraction (LVEF) < 30 % and LV thrombus predict poor prognosis.⁷ Our patient had EF of 33% and there was no LV thrombus. This improved her recovery from cardiac arrest with no clinical neurological deficit.

The principles of managing acute HF due to PPCM are also similar to those applying to acute HF arising from any other cause. The patient is best managed in an intensive care unit with close hemodynamic and oxygenation monitoring. Supplemental oxygen should be administered in order to achieve an arterial oxygen saturation of $\geq 95\%$, using, where necessary, non-invasive ventilation.¹ Intravenous diuretics, nitrates and inotropic agents are usually employed. Angiotensin Converting Enzyme Inhibitors and Angiotensin Receptor Blockers are contraindicated in pregnant females due to the risk of fetal toxicity. For anticoagulation in very low LVEF pregnant patients, low-molecular-weight heparins are preferred as they do not cross the placenta. Left ventricular function normalizes in more than half of the patients, especially in those with LVEF >30% at the time of diagnosis,⁸ therefore intra-aortic balloon pump and Left Ventricular Assist Device need consideration. Cardiac transplantation is the last hope.

Patients with prenatal diagnosis of PPCM need to be treated by a multidisciplinary approach. The pregnancy is allowed to progress to term if there is no maternal or fetal deterioration. Effective labor analgesia decreases pain and anxiety. It reduces sympathetic activity which is detrimental to the maternal health. Similarly in case of vaginal delivery, assisted second stage is recommended to reduce maternal efforts and shorten labor. Multiple modes of anesthesia have been described in the literature for operative delivery of patients with PPCM. Generally Regional Anesthesia (combined spinal epidural, continuous epidural anesthesia and Infiltration anesthesia) has been used in patients undergoing non urgent Cesarean Section with relatively stable hemodynamics, while moderately symptomatic patients or those undergoing emergency surgery have received General Anesthesia (GA).¹ The hemodynamic goals of anesthesia are common to all approaches and these are to reduce cardiac preload and afterload, and to avoid any manoeuvre or drug causing cardiac depression in a patient who has already compromised cardiac contractility. Ergometrine should be avoided and oxytocin should be given as an infusion. The postpartum management requires conventional heart failure medications prescribed for a minimum of one year, with ongoing treatment guided by the extent

of recovery of cardiac function. Most drugs used for the management of HF are compatible with breast-feeding therefore clinically stable women with PPCM should not be discouraged from breast-feeding their infants. Women with persistent LV dysfunction are advised not to pursue further pregnancies. How to advise women with recovered LV function is more difficult. Some studies report good outcomes,⁹ while others report a high rate of recurrence.^{10,11}

CONCLUSION

We conclude that giving due attention to women during their last trimester and early post natal period with even subtle sign and symptoms of heart failure, like easy fatigue ability, unexplained cough, nocturnal dyspnea and insomnia will help in the diagnosis of PPCM. These patients will further be managed in line with the recommendations keeping in view their cardiac morbidity, thus preventing mishaps.

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