Peripartum Cardiomyopathy: Report of two cases and review of literature

Abstract:
Peripartum cardiomyopathy (PPCM) is a rare condition affecting women in late pregnancy or up to five months of the postpartum period. The signs, symptoms and treatment of PPCM are similar to that of heart failure. Early diagnosis and proper management are the cornerstones for a better outcome for these patients. We report two cases of peripartum cardiomyopathy which were treated in the surgical intensive care unit (SICU) of Hamad General Hospital, Qatar.

Key words: Peripartum cardiomyopathy, heart failure, thrombo-embolism.

Introduction:
Peripartum Cardiomyopathy (PPCM) is a rare dilated cardiomyopathy causing heart failure in women during late pregnancy or early postpartum. PPCM in 90 per cent of the cases occurs during the first two months of the postpartum period (1). Etiology of PPCM is yet to be known. PPCM manifestations and treatment are the same as those for heart failure with the added consideration of the effects that the medications would have on the fetus. PPCM causes significant morbidity and mortality in both mother and fetus; hence all clinicians, and particularly acute care physicians, should be aware of this condition.

Case One:
A thirty-nine-year old Caucasian (Gravida 4, Para 3) in her 38th week of gestation had no history of medical illness apart from being on regular treatment for diabetes mellitus. She presented to the Emergency Department with a history of sudden difficulty in breathing that had been worsening over the previous two hours. Shortly after she developed severe respiratory distress and cyanosis with pinkish frothy secretions. After immediate intubation and ventilation she developed severe bradycardia followed by cardiac arrest. She was resuscitated successfully onsite and then admitted to the Surgical Intensive Care Unit (SICU) for further management that included ventilatory support with the supplementation of proper analgesia and sedation under Bispectral Index (BIS) monitoring – the invasive monitoring included a pulmonary artery catheter and an arterial line. Her central venous pressure (CVP) was 20 mm Hg, and her pulmonary artery wedge pressure (PAWP) was 24 mm Hg, with a cardiac index of 2.2 liters/m2. Echocardiography showed global hypokinesia with a left ventricular ejection fraction of 18 percent. Peripartum cardiomyopathy was diagnosed according to the Demakis criteria and inotropes as well as diuretic infusion were instituted. On the fourth day the patient became stable and was weaned off the inotropes.

After weaning her off all sedation, analgesia and inotropic support she showed decerebrate posturing. Magnetic Resonance Imaging (MRI) of her brain showed diffuse hypoxic brain injury and an electroencephalography (EEG) demonstrated generalized slow activity. A follow-up echocardiogram showed an ejection fraction of 56 percent. Thereafter the patient was transferred to the rehabilitation ward for long-term care with a percutaneous tracheostomy.

Case Two:
A thirty-year-old female of African descent (Gravida 5, Para 3) in her 38th week of a twin pregnancy, with no history of any medical disease, underwent a cesarean section for non-progress of labor. She was explored postoperatively because of vaginal bleeding and hemostasis was achieved. Resuscitation included fluid, blood and blood products. She was then transferred to the SICU for further management. She was fully awake, breathing spontaneously, and hemodynamically stable. Invasive lines for monitoring were inserted and her CVP was 6-8 mm of Hg.

On the third day she developed tachycardia, had difficulty in breathing with a pink frothy secretion; her CVP had increased to 15-16 mm of Hg and a chest X-ray showed congested lungs.
She was intubated and ventilated with appropriate sedation and analgesia infusion guided by BIS monitoring. Echocardiography revealed global hypokinesia with a ventricular ejection fraction of 30 per cent. Inotropes and a diuretic infusion were instituted guided by Pulse Contour Continuous Cardiac Output (PiCCO) while fulfilling the Demakis criteria of PPCM. Digoxin was added to the treatment later. She improved clinically and when her chest and cardiac function became normal she was weaned off the ventilator and all inotropes and was discharged home a week later with no neurological deficits.

Discussion:

Peripartum Cardiomyopathy (PPCM) was recognized as a separate clinical disease in the 19th century and in 1970 the criteria to diagnose PPCM were published (2). The etiology of PPCM is currently unknown although several possible causes have been hypothesized including increased inflammatory cytokines, viral infection, myocarditis, maternal immunologic response to fetal cells in maternal blood, and pregnancy-related hemodynamic changes. Although no causal links have been identified to date, the following factors have been associated with an increased risk: over 30 years of age, multiparity, African descent, multiple-gestation pregnancy, maternal cocaine abuse, history of pre-eclampsia, eclampsia, or postpartum hypertension and long term tocolytic therapy (4). The role of selenium deficiency is controversial (5). Other hypotheses have been proposed for the etiology of PPCM where a familial clustering of PPCM could be due to genetic factors (6). The abnormal immune response to the fetal cell is also blamed for PPCM (7) – Homan et al proposed that hemodynamic maladaptation to the stress of pregnancy causes PPCM (8).

The actual incidence of PPCM is unknown and reasons for geographic variations are not well understood. For example, in Haiti the incidence is 1 case per 299 live births; in South Africa 1 case per 1,000 live births; and in the United States 1 case per 2,000 live births (3). Unfortunately, we do not have an accurate incidence of PPCM at our hospital, possibly due to a lack of documentation or missed cases.

Patients with PPCM may present classic signs and symptoms of heart failure. Their histories are commonly notable for dyspnea on exertion, orthopnea, paroxysmal nocturnal dyspnea, and cough. Dyspnea and orthopnea are the common complaints of PPCM and the criteria for diagnosis are as follows:

- a. Echocardiographic evidence of heart failure (i.e. ejection fraction <45%),
- b. Onset between the last month of pregnancy and five months after pregnancy,
- c. No other identifiable causes of heart failure, and
- d. No pre-existing heart disease before the last month of pregnancy.

In the setting of acute pulmonary edema, urgent afterload and preload reduction is required. IV nitroglycerin or nitroprusside infusions can be uptitrated until BP is controlled. Diuretics, such as IV furosemide, help lower BP and cardiac filling pressures. Morphine is a potent vasoconstrictor and helps reduce preload and alleviate respiratory distress.

The aim of treatment in PPCM is to reduce preload, afterload and increase the contractility of the heart. Digoxin is safe to use during pregnancy and beta-blockers improve left ventricular function in patients of PPCM, but angiotensin-converting enzyme (ACE) inhibitors are the drugs of choice in the post partum period. Maintenance therapy for PPCM is the same as that for heart failure of another etiology. If the patient is pregnant ACE inhibitors should not be given due to the risk of oligohydramnios, fetal renal damage or fetal death. Hydralazine with nitrates can be substituted for ACE inhibitors.

The critically ill PPCM patients may need inotropes and other intravenous medications. Therapy should be guided by invasive monitoring, while taking into account the physiological changes of pregnancy (9). Ventricular arrhythmias should be treated aggressively with a class 3 anti-arrhythmic medication (10). Left ventricular (LV) stasis from severe heart failure and current or recent pregnancy places the patients with PPCM at a high risk for cardiac thrombosis and subsequent peripheral embolization. Therefore, patients with PPCM may exhibit lower-extremity arterial occlusion, cerebrovascular accident, mesenteric ischemia or infarct, and pulmonary embolism.

Clinicians must have a high index of suspicion for underlying PPCM when they encounter patients with these embolic diseases; anticoagulation is therefore a must in these patients. Occasionally PPCM patients may need mechanical cardiovascular support such as an intra-aortic balloon pump or ventricular assisting device – there have been cases reported where a cardiac transplant has been necessary (11). Unless the maternal hemodynamic is unable to support the fetus, or the support of the mother puts the fetus at risk, there would be no need to terminate the pregnancy in PPCM patients (12).

Despite its rarity PPCM causes considerable mortality and morbidity. The maternal mortality is 15-50 per cent (13), and up to 40 per cent of PPCM patients undergo a cesarean section (14). Many women with PPCM have a spontaneous recovery of LV function – this occurrence is distinct from outcomes with other causes of dilated cardiomyopathy. A retrospective study of 123 women in the United States with PPCM showed that mean LV ejection fractions increased from 29 per cent to 48 per cent over a period of one year. This same series showed an overall transplantation rate of 4 per cent and a mortality rate of 9 per cent.

Several studies have shown that patients with a history of
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PPCM are at risk for severe LV dysfunction should they become pregnant again. A recent study showed that of nine patients with a history of PPCM that became pregnant again, five (56%) died from severe heart failure (17). The prevention of subsequent PPCM is to avoid subsequent pregnancies (16) and patients must therefore be clearly advised of the risks of a subsequent pregnancy.

References