Case Report

Post-Delivery Cardiomyopathy in a Patient Admitted to Critical Care Unit; A Rare Case Report

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Abstract

Background: Peripartum cardiomyopathy (PPCM) is an uncommon disease that affects women in the last month of pregnancy or within the first five months postpartum, occurring in about 1 in 3500 live births. The disease bears potentially devastating effects both on mother and the fetus if not treated early in its course.

Case Presentation: The case was a 34-year old woman with a triple pregnancy who presented to the ward immediately after cesarean section with signs of dyspnea, cyanosis and pulmonary edema. She was diagnosed with PPCM upon echocardiography. The patient improved remarkably despite the PPCM's devastating complications. This case report aims to describe a female patient who developed PPCM after a triple delivery.

Conclusion: Regarding the high risks of developing PPCM in subsequent pregnancies and avoiding multiparty, especially in older age, a reliable contraception in childbearing women would be helpful. The best prevention of PPCM is to avoid subsequent pregnancies.

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Background

Peripartum cardiomyopathy (PPCM) is a rare and life-threatening cardiomyopathy of unknown etiology that affects women in the last month of pregnancy or in the first five months postpartum. The true incidence of PPCM is unknown. The incidence of PPCM seems to range from 1:300 to 1:15,000 live births. The published mortality rates in PPCM, generally range from 25 to 50% in the United States (1-3). As the incidence of PPCM is 1: 3000 to 1: 4000 live births it seems to affect 1000–1300 women in the United States annually (4, 5). As the majority of PPCM cases occur during postdelivery period, it is important to know that PPCM is a diagnosis of exclusion by applying its diagnostic criteria (6).

Although the etiology of PPCM remains unclear, a number of potential risk factors for this disorder have been proposed. Among these factors are multiparty, advanced maternal age, multiple pregnancy, preeclampsia, gestational hypertension and women of African descent. Other risk factors include association with maternal cocaine abuse, selenium deficiency and long-term (>4 weeks) oral tocolytic therapy with beta adrenergic agonists such as terbutaline. However the disease may occur in women in the absence of these risk factors.

A number of possible causes have been proposed for PPCM, including myocarditis, abnormal immune response to pregnancy, maladaptive re-

sponse to the hemodynamic stresses of pregnancy and stress-activated cytokines. In addition, there have been a few reports on the genetic bases of familiar PPCM (7-18).

The diagnostic criteria of PPCM are based upon the clinical presentation and echocardiography (16). It is important to realize that the diagnosis of PPCM requires the exclusion of other causes of cardiomyopathy before its diagnosis is considered (19).

Treatment of PPCM is similar to that of other forms of congestive heart failure. A combination of digoxin, vasodilators, diuretics, beta blockers, sodium restriction and reduction of afterload forms the cornerstone of therapy. The goal of therapy in PPCM is reductions in both preload and afterload plus increased inotropy. Careful attention must be paid to fetal safety regarding excretion of the metabolites into breast milk. Furthermore, patients with PPCM are highly predisposed to thromboembolic phenomena. Thus, anticoagulants should be considered in these patients. Either heparin or coumadin can be used safely postpartum and neither of them is secreted into breast milk (20-32).

The prognosis for women with PPCM appears to depend on the normalization of left ventricular size and function within six months after delivery. About half of the patients with PPCM recover without any complication. Persistence of the disease after six months indicates irreversible cardiomyopathy and poor survival. The mortality estimates for patients with PPCM in the United States range from 25-50%. Most deaths occur within the first three months postpartum. Death is usually caused by progressive pump failure, arrhythmias or thromboembolic events.

Persistence of cardiac dysfunction 6-12 months after the initial diagnosis of PPCM usually indicates an irreversible problem and almost always represents an absolute contraindication to a subsequent pregnancy. Currently the recommendations for further pregnancies after PPCM vary and there are few reports that describe the outcomes of subsequent pregnancies in such patients (3, 5, 25, 33).

Case Presentation

The aim of this case report was to describe a female patient who developed PPCM after a triple delivery. A 34-year-old women, G1 (Triple) and gestational age of 32 weeks and 5 days, was admitted to the Gynecology and Obstetrics Department of Al-Zahra Medical Center in Isfahan, Iran during June 2008. She had a past history of measles at the age of two with right eye involvement and sequelae plus a history of UTI during pregnancy. Two days after admission, she underwent an emergent cesarean section because of frequent uterine contractions. During these two days she received betamethasone. She had no history of significant shortness of breath during her entire pregnancy. She had complained of fatigue but this was attributed to her triple pregnancy. She had no history of cardiac problems. Her past medical history was otherwise unremarkable except the aforementioned problems.

On physical examination, her blood pressure was 110/80 *mmHg*, the pulse was 80 beats/*min* and regular, and the respiratory rate was 14/*min*. She was afebrile.

After induction of general anesthesia for cesarean section and intubation, oxygen saturation gradually dropped from 97% to 80% despite delivering 100% FIO2 under anesthesia. After extubation, she was agitated with some degrees of cyanosis in the post-anesthetic care unit (PACU). The chest examination revealed significant bilateral crackles. The cardiovascular examination showed a raised jugular venous pressure of about seven centimeters above the sternal angle. The cardiac auscultation showed normal first and second heart sounds and a third heart sound with a gallop rhythm. No significant cardiac murmurs were detected. The abdominal examination was unremarkable. There were no pitting edema in lower extremities and no signs of deep vein thrombosis. Chest X-ray showed pulmonary interstitial edema and an increase in cardiac silhouette with a cardiothoracic ratio (CTR) of about 68%. While still in post-anesthetic care unit (PACU), she felt better and pulmonary auscultation showed basilar crackles and mild cyanosis (pulse oximetry showed a SPO2 of about 80-83%) with moderate dyspnea and orthopnea after receiving furosemide, aminophylline and oxygen through facemask for two hours. Initial echocardiography showed a dilated, poorly contractile left ventricle with a hyperkinetic interventricular septum and an ejection fraction of 25%. The diagnosis of peri-

partum cardiomyopathy was made and the patient was treated for congestive heart failure with diuretics, digitalis and angiotensin converting enzyme inhibitors (ACE-I). She responded promptly to anti-heart failure therapy. Her improvement was rapid and cardiothoracic ratio (CTR) improved from a previous 68% ratio to about 50% on discharge from the hospital. A repeat echocardiography only after two days of treatment showed improvement in overall left ventricular function and the ejection fraction had increased to 40% with an improved left ventricular end diastolic function. She reported no adverse reactions to her medications and clinically was stable with no dyspnea on discharge. She was offered a beta blocker regimen (Carvedilol). She was strongly advised to avoid any further pregnancy and to continue regular medical follow-ups. She was discharged after nine days of hospitalization. Although the reported patient seemed to have full clinical improvement but concern still remains as her left ventricular function did not normalize (Mild Left Ventricular Dysfunction) on echocardiography.

Discussion

This patient had a normal term pregnancy and delivered three babies but developed PPCM in post-delivery period. PPCM is a rare and life-threatening cardiomyopathy of unknown etiology that affects women in the last month of pregnancy or in the first five month postpartum.

The exact etiology of PPCM is not known yet, but the following conditions have been suggested: familial, myocarditis, abnormal immune response and maladaption to pregnancy stress. Hyperdynamic circulation during pregnancy causes remodeling and transient hypertrophy of left ventricle. Exaggerated reduction in left ventricular systolic function alongside with the stress from gestational hypertension may contribute to heart failure in PPCM patients (15-18).

The clinical manifestation of patients with PPCM is similar to those of patients with congestive heart failure. PPCM presentations and treatments are the same as for heart failure but with regards to the unwanted effects of medications on the fetus.

Invasive hemodynamic monitoring will demonstrate elevated right and left heart filling pressures with low cardiac index. PPCM is uncommon but will cause significant morbidity and mortality in both mother and fetus. Therefore, all clinicians and particularly acute care physicians should be aware of this disease. It is important to note that PPCM can present with periodic thromboembolic phenomena (19).

In 1997, the National Heart Lung and Blood Institute (NHLBT) and the office for rare disease at National Institutes of Health (NIH) (16) convened a workshop that established the following diagnostic criteria:

1- The onset of cardiac failure with no identifiable cause in the last month of pregnancy or within five months after delivery.

2- The absence of any heart disease before the last month of pregnancy.

3- The absence of any identifiable cause for heart failure.

4- Presence of all the specific echocardiographic signs that follow (16).

Echocardiography evidence of left ventricular systolic dysfunction (LVEF < 45%, fractional shortening < 30%, or end-diastolic dimension > 2.7 cm/m^2).

Stricter echocardiographic criteria have been recommended including: a left ventricular ejection fraction of less than 45% t, fractional shortening of less than 30% on an M-mode echocardiography scan, or both, and a left ventricular end-diastolic dimension of more than 2.7 cm per square meter of body-surface area (10).

Medical management of PPCM is similar to other types of heart failure except from the effects of complications of treatment on the fetus or nursing infant. The main purpose of therapy in PPCM is to decrease preload and afterload plus increase in cardiac contractility.

Further management includes treatment with digitalis, diuretic agents, vasodilators (Amlodipine), β-blockers (Carvedilol), intravenous immune globulin, immunosuppressives, recombinant human relaxin and heart transplantation. Anticoagulants are also recommended in selected patients (20-31).

Thromboembolism is the most common complication of PPCM. A premature delivery rate of 25% has been described in patients with PPCM. PPCM cases have an increased frequency of cesarean section up to 40% (3, 25).

The prognosis of PPCM is related to the recovery of ventricular function. Failure of heart to return to a normal size is associated with excess morbidity and mortality.

The risk of developing PPCM in subsequent pregnancies remains high, especially if left ventricular dysfunction is persistent. The maternal mortality in PPCM has been reported to be 15-50%. Preterm delivery occurs in up to 25% of patients with PPCM. Intra-uterine fetal deaths have been reported in 40% of patients with PPCM undergoing cesarean section for obstetric indications. Patients who have recovered from PPCM run a high risk for the disease in subsequent pregnancies. Therefore, it is prudent that theses patients avoid further pregnancies (3,5,33).

Recommendations: Patients with PPCM are recommended to perform an echocardiogram every six months after the diagnosis and to avoid pregnancy until the ventricular ejection fraction has increased to greater than 50%, if they wish to have a further pregnancy at all.

Nurses and doctors should counsel patients on the potential health risks of future pregnancies and contraceptive use should be encouraged. Patients who decide to have additional pregnancies should be treated in high-risk prenatal centers (5, 33).

Conclusion

Peripartum cardiomyopathy is a rare disease of unknown etiology that strikes women in the childbearing years. The reported mortality of PPCM is 15-50%. The best prevention of PPCM is to avoid subsequent pregnancies.

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