

A case report of peripartum cardiomyopathy with a review of literature

A case report of peripartum cardiomyopathy

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Abstract

Peripartum cardiomyopathy (PPCM) is a severe form of idiopathic dilated cardiomyopathy, which appears either in the last month of pregnancy or during the five months following delivery. This clinical entity leads to left ventricular dysfunction and eventually cardiac insufficiency. Left ventricle functions recover spontaneously in some patients with PPCM while left ventricular dysfunction persists to cause an indication for cardiac transplantation in some patients. This case report aims to increase the awareness of obstetricians about PPCM by describing an affected patient.

Keywords

Dilated Cardiomyopathy; Pregnancy; Peripartum Cardiomyopathy

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Introduction

Peripartum cardiomyopathy (PPCM) is a severe form of idiopathic dilated cardiomyopathy which appears either in the last month of pregnancy or during the five months following delivery. This clinical entity leads to left ventricular dysfunction and eventually cardiac insufficiency [1, 2]. It has been reported that the incidence of PPCM is 1: 299 live births in Haiti, 1:100 to 1:1000 live births in South Africa and 1:1149 to 1:4000 in the United States of America. Black race, advanced maternal age, multiparity, multiple pregnancies, assisted reproductive techniques, prolonged tocolysis, malnutrition, viral infections, hypertension, diabetes mellitus, obesity, and smoking are the risk factors for this disease [2, 3]. Left ventricle functions recover spontaneously in some patients with PPCM. On the other hand, left ventricular dysfunction persists to cause an indication for cardiac transplantation in 4% to 11% patients with PPCM and 3.3% to 30% of the affected cases result in death [1, 3]. Early diagnosis, meticulous treatment and the awareness of the obstetrician about this disease contribute positively to the prognosis of this disease.

This case report aimed to increase the awareness of obstetricians about PPCM by describing an affected patient.

Case Report

A 42-year-old, multiparous woman with a 37-week-old pregnancy was admitted to the study center due to difficulty in breathing. Physical examination revealed asthenia, pallor, dyspnea, and orthopnea. The vital findings of the patient were body temperature: 37°C, respiratory rate: 26/min, pulse: 100/min and systolic/diastolic blood pressure: 180/100 mmHg. There was nothing particular in pulmonary auscultation whereas only tachycardia was noticed in cardiac auscultation. Moreover, there was severe edema in both lower extremities.

Laboratory tests indicated hemoglobin: 8.3 g/dl, leukocyte count: 17.3×10^3 /ml, platelet count: 217.000/mm³, albumin: 3.15 g/dl, C-reactive protein (CRP): 12.1 mg/dl (range: <5 mg/dl) and troponin 79.6 pg/ml (range: <100 pg/ml). Thyroid, hepatic, and renal function tests were normal and serological tests were negative for viruses which might cause myocarditis. The electrocardiography of the patient showed sinus rhythm and R voltage decrease in anterior leads. Arterial blood gas analysis demonstrated metabolic alkalosis (pH=7.466, pCO₂=29.3 mmHg, pO₂=72.4 mmHg, HCO₃=22.3 mmol/L).

Obstetric ultrasonography revealed a single, 37-week-old fetus with a normal amniotic fluid index and placenta. Likewise, the non-stress test confirmed the fetal well being. Echocardiography reported dilated and hypokinetic left ventricle, left atrium diameter of 4 cm (range: 2.7-3.8 cm), left ventricle end-diastolic diameter of 5.5 cm (range: 3.9-5.3 cm), left ventricle end-systolic diameter of 4.2 cm (range: 3.3-5.5 cm) and ejection fraction of 40% (range: >55%). Moreover, mild to moderate mitral regurgitation, mild aortic regurgitation and mild tricuspid regurgitation were specified. The right ventricle dimensions and functions, as well as pulmonary artery systolic pressure, were assessed to be normal.

The patient was diagnosed with PPCM within the scope of these findings so that nasal oxygen (4 l/min), subcutaneous enoxaparin (40 mg/0.4 ml/day) and intravenous nitroglycerine (0.1 mg/

kg/day) treatment were administered. After the transfusion of two units of erythrocyte suspension, the patient underwent cesarean delivery due to the breech presentation on the second day of the medical treatment. A 2865 gram weighing baby girl with the first and fifth minute Apgar scores of 8 was delivered by cesarean section. Echocardiography examination in the third postoperative day displayed spontaneous recovery of left ventricle dimensions, systolic functions, and segmentary wall movements.

Discussion

The etiology and pathophysiology of PPCM have not been clarified totally. Although PPCM has been identified as a non-genetic and non-familial form of dilated cardiomyopathy, an association between PPCM and single nucleotide polymorphism near PTHLH gene has been defined. It has been also proposed that pregnancy-related immunosuppression might trigger myocardial inflammation by means of viral infections so that PPCM might occur. In addition, the production of autoantibodies against myocardial proteins, excessive activation of anti-angiogenesis signal transduction pathways and/or selenium deficiency may participate in the pathogenesis of PPCM. Another hypothesis is based on the generation of reactive oxygen radicals and marked an increase in oxidative stress during the last trimester of pregnancy and postpartum period. When this increase in oxidative stress cannot be balanced by anti-oxidant defense mechanisms, reactive oxygen species accelerate the release of cathepsin-d which subsequently degrades the prolactin molecules during pregnancy and puerperium. It has been suggested that these prolactin fragments may impair myocardial functions [1-3].

PPCM may affect pregnant women of every age. However, more than half of the patients with PPCM in literature are older than 30 years [4].

PPCM causes clinical symptoms that are specific to systolic dysfunction. According to the classification of New York Heart Association, the majority of PPCM patients have class III or IV (moderate to severe) cardiac insufficiency. Such a clinical entity may result in clinical symptoms and signs including dizziness, fatigue, chest pain, exertional dyspnea, orthopnea paroxysmal nocturnal dyspnea, persistent cough, palpitation, distended neck veins, bloating, leg edema, rales, rhonchi, third heart sound and hepatomegaly. It should be kept in mind that these symptoms and signs may mimic the physiological alterations induced by a nine-month-old pregnancy [1, 4]. In this case report, only dyspnea, orthopnea and tachycardia have been determined. Such a finding suggests that PPCM has been diagnosed at an early stage.

The diagnostic criteria for PPCM were as follows: (1) Cardiac insufficiency emerges in the last month of pregnancy or within five months following delivery, (2) No underlying cause could be defined for cardiac insufficiency, (3) No history of cardiac disease could be specified before and during pregnancy, (4) Echocardiography examination shows left ventricular dysfunction. Less than 10% of the PPCM patients are diagnosed in the last month of pregnancy [5, 6]. Complying with literature, the patient in this case report has been diagnosed with PPCM in the last month of pregnancy as the patient has echocardiography

findings related with PPCM in the absence of any underlying cause for cardiac insufficiency and previous history of cardiac diseases.

Most of the PPCM patients present with non-specific chest X-ray findings (i.e., cardiomegaly, pulmonary congestion and pleural effusion) and electrocardiography changes (i.e., sinus tachycardia, atrial fibrillation, atrial flutter and ventricular tachycardia). Electrocardiographic alterations such as ST and T-wave deformities, QT prolongation and QRS enlargement may indicate PPCM as well. Since the concentrations of N-terminal pro-brain natriuretic peptide (NT-proBNP) and troponin do not change during pregnancy and puerperium, any increase in these biochemical values points out PPCM [2, 5].

Echocardiography is the first diagnostic method used in patients who have a high index of suspicion for PPCM because it is an easily applicable and replicable procedure. The dimensions, structure, systolic and diastolic functions of the left ventricle can be evaluated accurately. The patients who have an ejection fraction < %45 and/or fractional shortening > %30 can be diagnosed with PPCM. Moreover, left ventricle dilatation, left ventricle systolic dysfunction, right ventricle dilatation, mitral regurgitation, tricuspid regurgitation and occasionally left ventricle thrombus can be identified [2, 6].

Magnetic resonance imaging (MRI) can be used to visualize intracardiac thrombi, cardiac structures, volumes, and functions more accurately. Besides, MRI may help to designate ischemic heart diseases or acute myocarditis. It has been recommended that MRI should be performed without contrast enhancement in pregnant women. However, MRI with gadolinium enhancement may ease the identification of fibrosis and infarcted areas [7]. The Endomyocardial biopsy could be performed in patients who are suspected to have myocarditis and patients who have shown no improvement after the administration of two-week-long medical treatment [2]. In this case report, both electrocardiography and echocardiography have been sufficiently utilized to assess the patient and diagnose her with PPCM.

The patients diagnosed with PPCM should be managed by a team of cardiologists, obstetricians, intensive care experts, pediatricians, and anesthesiologists. A standardized treatment of PPCM consists of oxygen support, fluid restriction, diuretics, angiotensin - converting enzyme inhibitors, angiotensin receptor antagonists, beta-blockers, aldosterone antagonists, and vasodilators. Although diuretics treat pulmonary congestion and peripheral edema by decreasing cardiac preload, these drugs may induce uterine hypoperfusion and maternal metabolic acidosis. That's why; HCO₃ monitorization and acetazolamide support is significant in pregnant women treated with diuretics. Digoxin can be safely prescribed for pregnant and puerperal women who have cardiac insufficiency symptoms and low ejection fraction. Angiotensin-converting enzyme inhibitors, angiotensin receptor antagonists and aldosterone antagonists decrease cardiac afterload efficiently but all of these drugs have severely toxic effects. It should be remembered that only angiotensin-converting enzyme inhibitors can be safely used in nursing mothers. A combination of hydralazine and isosorbide dinitrate can be administered to decrease cardiac afterload. Beta-blockers decrease the risk of ventricular arrhythmia and sudden cardiac death by attenuating tumor necrosis factor-alpha (TNF- α)

release and its myocardial expression. Metoprolol is the most commonly and safely used beta-blocker during pregnancy and postpartum period. On the other hand, beta-blocker treatment may impair uteroplacental circulation and, thus, end up with fetal bradycardia, hypoglycemia, and respiratory depression. The pregnant women receiving beta-blocker treatment should be monitored with respect to fetal well-being. Anti-coagulation with low molecular weight heparin should be started in PPCM patients who have an ejection fraction < %35 and high risk for left ventricle thrombus. Since warfarin has embryopathy effects, its administration should be avoided in the first trimester of pregnancy. As warfarin may cause fetal cerebral bleeding in the second and third trimesters, its administration should be performed cautiously [7, 8]. Intra-aortic balloon pump, ventricular support devices, and extracorporeal membrane oxygenation might be required in PPCM patients who do not respond to the medical treatment implemented for cardiac insufficiency [8].

Conclusion

This case report describes a PPCM patient who has been conservatively treated with oxygen, vasodilators, and anticoagulation followed by definitive recovery by cesarean section. There has been no need for support device in this case because of mild-to-moderate clinical course. Serious complications may appear within the first six months following the diagnosis of PPCM. These complications include severe cardiac insufficiency, cardiogenic shock, cardiopulmonary arrest, arrhythmias, thromboembolism, multiorgan failure and even death. Delayed diagnosis, QRS interval \geq 120 ms in electrocardiography, ejection fraction < 25% in echocardiography and NT-proBNP level > 300 pg/mL are the risk factors for PPCM-related complications [8]. Since the patient described by this case report does not have any risk factor, no complication related with PPCM has occurred during the postpartum period.

Scientific Responsibility Statement

The authors declare that they are responsible for the article's scientific content including study design, data collection, analysis and interpretation, writing, some of the main line, or all of the preparation and scientific review of the contents and approval of the final version of the article.

Animal and human rights statement

All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. No animal or human studies were carried out by the authors for this article.

Conflict of interest

None of the authors received any type of financial support that could be considered potential conflict of interest regarding the manuscript or its submission.

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