Peripartum cardiomyopathy: a case report

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Abstract

Objective: Peripartum cardiomyopathy (PPCM) has an unknown etiology and it is one of the complications exposing a life-threatening risk for pregnancy potentially. In our case report, we aimed to discuss the management of PPCM case developing at postpartum period.

Case: In this case report, we analyzed a case which delivered at 37 weeks of gestation and had complaints of dyspnea, orthopnea and swelling feet. When severe left ventricle systolic dysfunction was observed in the examinations performed, other diagnoses were ruled out and the patient was diagnosed with PPCM.

Conclusion: Peripartum cardiomyopathy should be considered certainly in the differential diagnosis of a patient who refers with the complaints of shortness of breath and swelling feet which are common during pregnancy.

Keywords: Peripartum cardiomyopathy, prognosis, echocardiography.

Introduction

Peripartum cardiomyopathy (PPCM) has an unknown etiology and it is one of the complications exposing a life-threatening risk for pregnancy potentially. This rare type of dilated cardiomyopathy causes congestive cardiac failure at last months of pregnancy or within the first 5 postnatal months. Clinical course may vary between spontaneous recovery of ventricular functions and the need for cardiac transplantation when disease becomes refractory. Although mortality rate was found 50% in 1950s, it has been reported today that the need for cardiac transplantation or death is observed between 12 and 18%.[1] Early diagnosis, treatment and good knowledge of clinician on this disease are the basic factors improving prognosis.

Case Report

Nineteen-year-old patient who had her first pregnancy referred to our hospital upon spontaneous labor progress when her pregnancy was 37 weeks and 1 day, and she vaginally delivered 2450 g health male baby. Her postpartum first 24 hours were uneventful; however, her...
complaints began to occur. In the physical examination of the patient upon the complaints of shortness of breath and swelling feet, her general condition was at a medium level and she seemed pale. There were dyspnea and orthopnea. In the pulmonary auscultation, respiratory sounds were decreased at right lung basal and there were rales at left lung basal. In the cardiac auscultation, tachycardia, S3 gallop rhythm, and 3/6 apical systolic murmur spreading to axilla were found. In the bilateral lower extremity, +3 positive pitting edema was observed. The values found in the laboratory tests were as follow: Hemoglobin: 7.3 g/dL, hematocrit: 22.5%, platelet count: 502,000 mm$^3$, ALT: 152 U/L, AST: 84 U/L, LDH: 332 U/L, CK: 190 U/L, and albumin: 3.4 g/dL. The electrocardiography of the patient was in sinus rhythm; the pulse was 118/min, and T wave negativity was observed in V1-3 derivations. In the chest radiography, it was seen that cardiothoracic rate increased and there was increase in reticular density on bilateral lower zones. Metabolic acidosis (pH=7.05, PCO$_2$=37.6 mmHg, PO$_2$=35.8 mmHg, HCO$_3^-$=11.1 mmol/L) was found in the arterial blood gas.

After the first assessment of the patient, pulmonary embolism, cardiac failure associated with anemia and PPCM were considered as pre-diagnoses, and 4 L/min nasal oxygen, anticoagulant treatment and diuretic treatment were initiated. In the echocardiography (ECG) carried out after the first response, it was seen that the left cardiac structures were dilated, left atrium diameter was 4.8 (range: 2.7-3.8) cm, left ventricle end-diastolic diameter was 5.5 (range: 3.9-5.3) cm, and left ventricle end-systolic diameter was 4.8 (range: 3.3±0.5) cm (Fig.1).

In the left ventricle, global hypokinetic and ejection fraction (EF) was 30% (range: >55%), and moderate mitral insufficiency and mild tricuspid insufficiency were detected (Fig. 2). Pulmonary arterial systolic pressure was measured 40 mmHg (range: <36 mmHg). Pericardial fluid was observed which was surrounding left ventricle and not forming any constriction. By these findings, the patient was assessed as PPCM and transferred to the cardiology clinic. As a classical treatment of cardiac failure, salt restriction, digoxin, diuretic and vasodilator agents were administered. Blood transfusion was done for anemia. A dramatic response was received from the patient after the treatment. The patient was discharged when class 4 functional effort capacity according to the Cardiac Disease Classification of New York Heart Association (NYHA) and NYHA class 4 on postpartum 15th day were observed.

**Discussion**

Cardiomyopathy developing in pregnant women having no underlying cardiac disease is characterized by the development of cardiac systolic dysfunction due to the decrease in left ventricle EF as in other cardiomyopathies. Clinical diagnosis criteria of the disease were defined by Demakis et al. in 1971 as follow:

1. Cardiac failure developing at the last month of pregnancy or within the first 5 months after delivery;
2. Non-existence of other reasons explaining cardiac failure,
3. Non-existence of any cardiac disease defined before the last month of pregnancy.

Later, presence of left ventricular dysfunction occurring with the EF decrease in the echocardiography was added to these criteria. Since our case had no cardiac disease history, her disease occurred at postpartum period and there was no other reason explaining cardiac disease and ECO findings were present, her diagnosis was established as PPCM. Although the actual incidence of the disease is not known, it is estimated that it is seen in about 1000-1300 women in the USA annually. It is thought to be more common in women on the upper limit of fertile period, those gave a lot of births, in multiple pregnancies, preeclampsia and in Afro-Americans. However, when cases are reviewed, 25-75% of them are seen in the young women who give birth for the first time.

Among the factors causing PPCM, there are cardiotoxic viruses, autoimmune diseases, toxins causing immune system dysfunction, abnormal serum relaxin levels, selenium deficiency, presence of proinflammatory cytokines, antibodies abnormally responding to cardiac tissues at high titers, and the underlying myocarditis. Myocarditis findings were found in 50% of those who had endomyocardial biopsy in PPCM. Even though the tocolytic agents, hypertension during pregnancy or preeclampsia causes cardiac failure to worsen, they do not cause PPCM, and these factors do not exist in the medical history of our case as in many cases. Unlike pregnant patients with underlying cardiac disease, cardiac failure begins at the end of pregnancy or after delivery in PPCM cases. The most common symptoms are chest pain, dyspnea, orthopnea and cough. Frequently observing these symptoms in pregnant patients due to physiological changes, and at postpartum period in patients who do operative delivery and even mistaking cardiac arrest for eclampsia crisis may cause difficulty during diagnosis.

Peripartum cardiomyopathy treatment is same as the conventional treatment of cardiac failure; oxygen supplementation, salt restriction, diuretics, digital and vasodilator agents are the basic instruments. ACE inhibitors frequently used in cardiac failure are contraindicated during antenatal period since they are associated with oligohydramnios, intrauterine growth retardation, neonatal renal failure and early neonatal death during pregnancy. As the data about beta-blocker use during pregnancy for PPCM is limited, such medication should be used after delivery. New agents are also tried in PPCM treatment. There are publications in the literature reporting that prolactin suppression and high dose of immunoglobulin accelerates healing process and provides rapid recovery in ventricular functions.

In the study of Bireker et al. carried out with levosimendan which is a new inotrope and vasodilator effective drug in PPCM cases, it was found that this drug did not change prognosis.

Peripartum cardiomyopathy may cause congestive cardiac failure, rise in increased atrioventricular arrhythmia, thromboembolism and sudden death. Thromboembolism may be seen especially in cases whose left ventricle EFs are severely depressed, and generally in 50% of patients. Therefore, it should be considered to add anticoagulant treatment to the treatment of standard cardiac failure treatment.

The option for the patients who are not responding to medical treatment is the cardiac transplantation in which 5-year survival rate is 60%. Duran et al. found mortality rate 30.3%, heart transplantation need 6.1% and persistent left ventricular dysfunction 39.4% in PPCM cases. Although there are mortality rates reported as 25-50% in previous publications, Felker et al. found 5-year survival rate 94%. Despite such optimistic statistics, there is also another patient group in which disease progress rapidly and irreversibly, and patient is lost within three months due to arrhythmia, thromboembolic complications and excessive pumping function insufficiency. It is cardiac function which actually determines the prognosis of patient. The progress of cardiac dysfunction more than 6 months seen in 50% of the cases shows that it is an irreversible disease. Although there is no consensus that the disease has no risk to repeat in subsequent pregnancies, Elkayam et al. stated that cardiac failure developed in subsequent pregnancies of 21% of the patients who regained their normal ventricular functions. Therefore, it cannot be guaranteed that subsequent pregnancies of patients whose cardiac function recovered will be uneventful. To decide and not to recommend another pregnancy is easy for patients who already have persisting cardiac disease since the mortality rate in subsequent pregnancies is 8-17%.
Conclusion
The diagnosis of peripartum cardiomyopathy is established by ruling out other possible diagnoses. PPCM is a gestational complication of which has an unknown etiology and lethal potential. Good knowledge of clinician on this disease increases the possibility of rapid diagnosis and proper treatment, and optimizes the prognosis.

Conflicts of Interest: No conflicts declared.

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