



Peripartum cardiomyopathy: a case report

Cenk Gezer¹, Atalay Ekin¹, Mehmet Özeren¹, Cüneyt Eftal Taner¹,
Nazile Bilgin Doğan², Aşkın Doğan¹

¹Department of Perinatology, İzmir Tepecik Training and Research Hospital, İzmir, Turkey

²Cardiology Clinic, Menemen State Hospital, İzmir, Turkey

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.

Özet: Peripartum kardiyomiyopati: Olgu sunumu

Amaç: Peripartum kardiyomiyopati (PPKM) altta yatan sebebin tam olarak bilinmediği, gebeliğin potansiyel olarak hayatı tehlike arz eden komplikasyonlarından birisidir. Olgu sunumumuzda postpartum gelişen PPKM olgusunun yönetimini tartışmayı amaçladık.

Olgu: Biz bu olgu sunumunda, 37. gebelik haftasında doğum yapan, postpartum dönemde dispne, ortopne ve ayaklarda şişlik şikayetleri ortaya çıkan bir olguyu ele aldık. Yapılan tetkiklerde şiddetli sol ventrikül sistolik disfonksiyonu ortaya çıkması üzerine diğer tanılar ekarte edilerek hastaya PPKM tanısı konuldu.

Sonuç: Gebelikte de sıkça rastlanılan nefes darlığı ve ayaklarda şişlik şikayeti ile gelen bir hastanın ayırıcı tanısında PPKM mutlaka düşünülmelidir.

Anahtar sözcükler: Peripartum kardiyomiyopati, prognoz, eko-kardiyografi.

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.^[1] 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 transplan-

tation or death is observed between 12 and 18%.^[2] 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

Correspondence: Cenk Gezer, MD. İzmir Tepecik Eğitim ve Araştırma Hastanesi, Perinatoloji Kliniği, İzmir, Turkey.
e-mail: drcenkgezer@gmail.com

Received: January 29, 2014; **Accepted:** March 31, 2014

Available online at:
www.perinataljournal.com/20140222011
doi:10.2399/prn.14.0222011
QR (Quick Response) Code:



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 mm³, 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, PCO₂=37.6 mmHg, PO₂=35.8 mmHg, HCO₃⁻=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 (ECO) 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**).^[3] Pulmonary arterial systolic pressure was measured 40 mmHg (range: <36 mmHg).^[4] 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:^[5]

1. Cardiac failure developing at the last month of pregnancy or within the first 5 months after delivery,

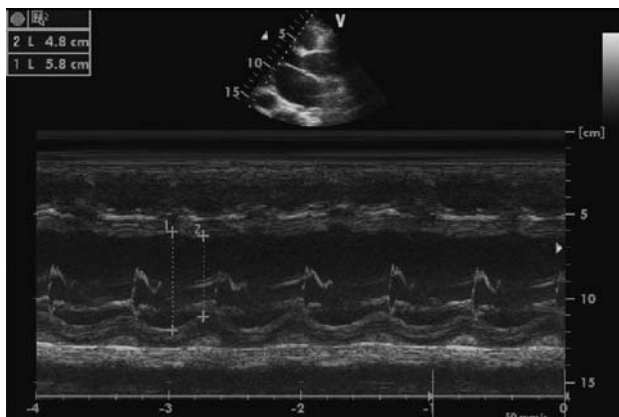


Fig. 1. Left ventricle end-systolic and end-diastolic diameter measurement in the transthoracic M-mode echocardiography of the case presented.

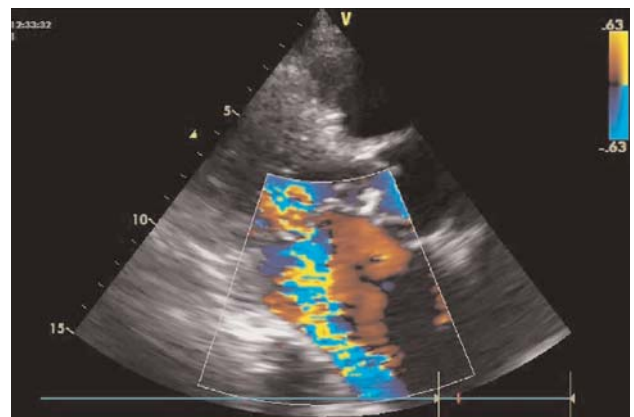


Fig. 2. Mitral insufficiency in the color Doppler in the transthoracic echocardiography of the case presented.

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.^[6] 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.^[6] 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.^[7] However, when cases are reviewed, 25-75% of them are seen in the young women who give birth for the first time.^[8,9]

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.^[5,6,10-12] Myocarditis findings were found in 50% of those who had endomyocardial biopsy in PPCM.^[11] 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.^[13] Unlike pregnant 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 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.^[1]

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 retar-

dation, 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.^[6] 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.^[14]

In the study of Biteker 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.^[15]

Peripartum cardiomyopathy may cause congestive cardiac failure, rise in increased atrioventricular arrhythmia, thromboembolism and sudden death.^[1] Thromboembolism may be seen especially in cases whose left ventricle EFs are severely depressed, and generally in 50% of patients.^[6] 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%.^[16] Duran et al. found mortality rate 30.3%, heart transplantation need 6.1% and persistent left ventricular dysfunction 39.4% in PPCM cases.^[17] Although there are mortality rates reported as 25-50% in previous publications, Felker et al. found 5-year survival rate 94%.^[2,18,19] 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.^[20] 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.^[21] 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%.^[22]

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.

References

- Nelson M, Moorhead A, Yost D, Whorton A. A 35-year-old pregnant woman presenting with sudden cardiac arrest secondary to peripartum cardiomyopathy. *Prehosp Emerg Care* 2012;16:299-302.
- O'Connell JB, Costazo-Nordin MR, Subramanian R, Robinson JA, Wallis DE, Scanlon PJ, et al. Peripartum cardiomyopathy: clinical, hemodynamic, histologic, and prognostic characteristics. *J Am Coll Cardiol* 1986;8:52-6.
- Lang RM, Bierig M, Devereux RB, Flachskampf FA, Foster E, Pellikka PA, et al.; Chamber Quantification Writing Group; American Society of Echocardiography's Guidelines and Standards Committee; European Association of Echocardiography. Recommendations for chamber quantification: a report from the American Society of Echocardiography's Guidelines and Standards Committee and the Chamber Quantification Writing Group, developed in conjunction with the European Association of Echocardiography, a branch of the European Society of Cardiology. *J Am Soc Echocardiogr* 2005;18:1440-63.
- Galiè N, Hoepfer MM, Humbert M, Torbicki A, Vachiery JL, Barbera JA, et al.; ESC Committee for Practice Guidelines (CPG). Guidelines for the diagnosis and treatment of pulmonary hypertension: the Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS), endorsed by the International Society of Heart and Lung Transplantation (ISHLT). *Eur Heart J* 2009;30:2493-537.
- Demakis JG, Rahimtoola SH. Peripartum cardiomyopathy. *Circulation* 1971;44:964-8.
- Pearson GD, Veille JC, Rahimtoola S, Hsia J, Oakley CM, Hosenpud JD, et al. Peripartum cardiomyopathy: National Heart, Lung, and Blood Institute and Office of Rare Diseases (National Institutes of Health) workshop recommendations and review. *JAMA* 2000;283:1183-8.
- Mielniczuk LM, Williams K, Davis DR, Tang AS, Lemery R, Green MS, et al. Frequency of peripartum cardiomyopathy. *Am J Cardiol* 2006;97:1765-8.
- Sliwa K, Forster O, Libhaber E, Fett JD, Sundstrom JB, Hilfiker-Kleiner D, et al. Peripartum cardiomyopathy: inflammatory markers as predictors of outcome in 100 prospectively studied patients. *Eur Heart J* 2006;27:441-6.
- Sliwa K, Skudicky D, Bergemann A, Cnady G, Puren A, Sareli P. Peripartum cardiomyopathy: analysis of clinical outcome, left ventricular function, plasma levels of cytokines and Fas/Apo-1. *J Am Coll Cardiol* 2000;35:701-5.
- Ansari AA, Fett JD, Carraway RD, Mayne AE, Onlamoon M, Sundstrom JB. Autoimmune mechanisms as the basis for human peripartum cardiomyopathy. *Clin Rev Allergy Immunol* 2002;23:289-312.
- Felker GM, Jaeger CJ, Klodas E, Thiemann DR, Hare JM, Hruban RH, et al. Myocarditis and long-term survival in peripartum cardiomyopathy. *Am Heart J* 2000;140:785-91.
- Midei MG, DeMent SH, Feldman AM, Hutchins GM, Baughman KL. Peripartum myocarditis and cardiomyopathy. *Circulation* 1990;81:922-8.
- Fett JD, Christie LG, Carraway RD, Murphy JG. Five-year prospective study of the incidence and prognosis of peripartum cardiomyopathy at a single institution. *Mayo Proc* 2005;80:1602-6.
- Biteker M, Duran NE, Ozkan M. The role of bromocriptine in peripartum cardiomyopathy. *Am J Obstet Gynecol* 2009; 201:e13.
- Biteker M, Duran NE, Kaya H, Gündüz S, Tanboğa Hİ, Gökdeniz T, et al. Effect of levosimendan and predictors of recovery in patients with peripartum cardiomyopathy, a randomized clinical trial. *Clin Res Cardiol* 2011;100:571-7.
- Heider AL, Kuller JA, Strauss RA, Wells SR. Focus on primary care: peripartum cardiomyopathy: a review of the literature. *Obstet Gynecol Surg* 1999;54:526-31.
- Duran N, Günes H, Duran I, Biteker M, Ozkan M. Predictors of prognosis in patients with peripartum cardiomyopathy. *Int J Gynaecol Obstet* 2008;101:137-40.
- Rickenbacher PR, Rizeq MN, Hunt SA, Billingham ME, Fowler MB. Longterm outcome after heart transplantation for peripartum cardiomyopathy. *Am Heart J* 1994;127:1318-23.
- Felker GM, Thompson RE, Hare JM, Hruban RH, Clemetson DE, Howard DL, et al. Underlying causes and long term survival in patients with initially unexplained cardiomyopathy. *N Engl J Med* 2000;342:1077-84.
- Rosner G, Wolchock Rosner SN, Heller I, Topilsky M. Congestive heart failure in pregnancy: a case of peripartum cardiomyopathy. *Neth J Med* 2004;62:290-2.
- Elkayam U, Tummala PP, Rao K, Akhter MW, Karaalp IS, Wani OR, et al. Maternal and fetal outcomes of subsequent pregnancies in women with peripartum cardiomyopathy. *N Engl J Med* 2001;344:1567-71.
- Elkayam U. Pregnancy and cardiovascular disease. In: Braunwald E, Zipes DP, Libby P, editors. *Heart Disease. A Textbook of Cardiovascular Medicine*. 6th ed. Philadelphia: WB Saunders Company; 2001; p: 2172-89.