Peripartum Cardiomyopathy and Respiratory Failure: Case Report

Peripartum Kardiyomiyopati ve Solunum Yetmezliği: Olgu sunumu

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Abstract

Peripartum cardiomyopathy (PPCM) is a rare, life-threatening heart disease of unknown etiology and occurs during the peripartum period in previously healthy women. When a female patient in peripartum has respiratory and cardiac failure, the physicians should be alert about this disease because the complaints are nonspecific. Herein, we present a previously healthy 29-year-old case of PPCM who showed nonspecific respiratory symptoms leading to a preliminary diagnosis of pulmonary thromboembolism (PTE). Echocardiography was an important tool for establishing the eventual diagnosis of PPCM with a consistent history and clinical background. The diagnosis, clinical course, and treatment of PPCM were reviewed in the light of pertinent literature. (Yoğun Bakım Derg 2013; 4: 18-20)

Key words: Peripartum cardiomyopathy, respiratory failure, cardiogenic pulmonary edema, pregnancy

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Özet

Peripartum kardiyomiyopati (PPKMP), daha öncesinde sağlıklı kadınlarda peripartum dönemde ortaya çıkan ve etyolojisi belli olmayan nadir görülen bir kalp hastalığıdır. Bu dönemde solunum ve kalp yetmezliği gelişen kadınlarda, yakınmaların nonspesifik olması nedeniyle uyanık olunmalıdır. Bu makalede, daha önce sağlıklı olan ve nonspesifik semptomları nedeniyle ilk olarak pulmoner emboli düşünülen 29 yaşında bir bayan olgu rapor edilmiştir. Anamnez ve klinik bulgular eşliğinde kesin PPKMP tanısına ulaşmada, ekokardiyografi önemli bir araç olmuştur. Hastalığın tanısı, klinik özellikleri ve tedavisi literatür ışığında tartışılmıştır. (Yoğun Bakım Derg 2013; 4: 18-20)

Anahtar sözcükler: Peripartum kardiyomiyopati, solunum yetmezliği, kardiyojenik pulmoner ödem, gebelik

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Introduction

Peripartum cardiomyopathy (PPCM) is a rare and life-threatening heart disease of unknown etiology. It can present within the interval from the beginning of the last trimester until the end of the first 6 months of postpartum period and causes circulatory and respiratory failure. The incidence of PPCM is 1 in 3000-4000 live births (1). Its etiology remains uncertain, but multifetal pregnancy, preeclampsia/eclampsia, advanced maternal age, viral infections, and black race are known as the risk factors for PPCM. The mortality rate is high; estimated maternal death due to PPCM is about 4% in the U.S. each year (2-5). PPCM can be misdiagnosed, especially as pulmonary thromboembolism (PTE). Echocardiography is a valuable tool in the differential diagnosis (6-8). Herein, we report a case of PPCM to emphasize critical clinical presentation of this rare disease, which when misdiagnosed, can be fatal.

Case Presentation

A 29-year-old woman presented to the emergency department with progressively increasing dyspnea within the previous 2 days. At the 36th week of her pregnancy, she was admitted to another hospital with pelvic inflammatory disease. During the treatment of pelvic inflammatory disease, she developed oligohydramniosis, and her pregnancy was terminated by cesarean section. Since her dyspnea, starting postoperatively, progressed to respiratory failure on the second postoperative day, she was referred to our center with a suspected diagnosis of PTE. Her baby had a low birth weight but was found to be healthy otherwise on examination after birth. The patient was a housewife and stated that she was capable of doing all active work at home and outside without feeling dyspnea or weakness until the first postoperative day. Her past medical history was unremarkable except for two spontaneous abortions.

Her body temperature was 36.8°C, heart rate: 135/min, blood pressure: 110/70mm Hg, and respiratory rate: 34/min. She had a body mass index within normal limits. She was dyspneic and tachypneic. On auscultation, bilateral inspiratory rales were heard at the bases of the lungs, and bilateral expiratory ronchi on the middle and lower parts of hemithoraxes. She had bilateral marked pretibial edema.

The complete blood count was normal except for leukocytosis (12400/mm^3). The blood chemistry was normal except for mild hyponatremia (129 mEq/L). Her arterial blood gas analysis showed a pH of 7.52, a PaO_2 of 43 mmHg, a $PaCO_2$ of 29.2 mmHg, a HCO_3 of 21.6 while breathing room air. Her electrocardiogram showed supraventricular tachycardia.

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Bilateral infiltrations especially at the middle and lower zones, and cardiomegaly were notable on chest x-ray (Figure 1). Because of highly suspected PTE, spiral chest computed tomography (CT) was performed. A CT scan depicted bilateral reticulonodular and ground-glass infiltrations, especially in the lower lobes (Figure 2). There were no filling defects in pulmonary arteries consistent with PTE. Owing to the clinical and radiological findings, the lesions were more likely due to cardiac pulmonary edema. Echocardiography revealed mitral regurgitation secondary to dilatation of the left ventricle and an impaired function, with an ejection fraction of 35%. After consulting an obstetrician and a cardiologist, the case was diagnosed with PPCM. She was given treatment consisting of an angiotensin converting enzyme inhibitor, a calcium channel antagonist, furosemide and spironolactone for left ventricular failure during the hospitalization period. When the clinical state and echocardiographic findings improved, she was discharged from the hospital and referred to a cardiology clinic. Medical treatment was stopped at the end of the 5th month postoperatively because of complete recovery clinically, radiologically and echocardiographically. The patient was followed up to 9 months and no relapse occurred.

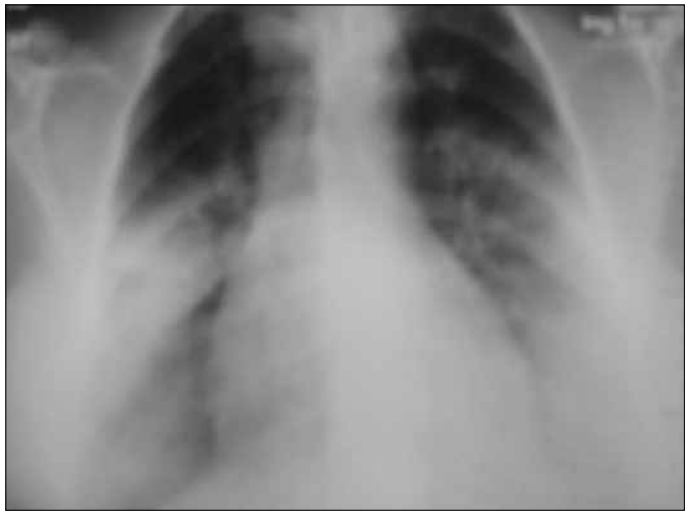

Figure 1. Chest x-ray depicting cardiomegaly and bilateral infiltrations predominantly in the middle and lower zones of the lungs

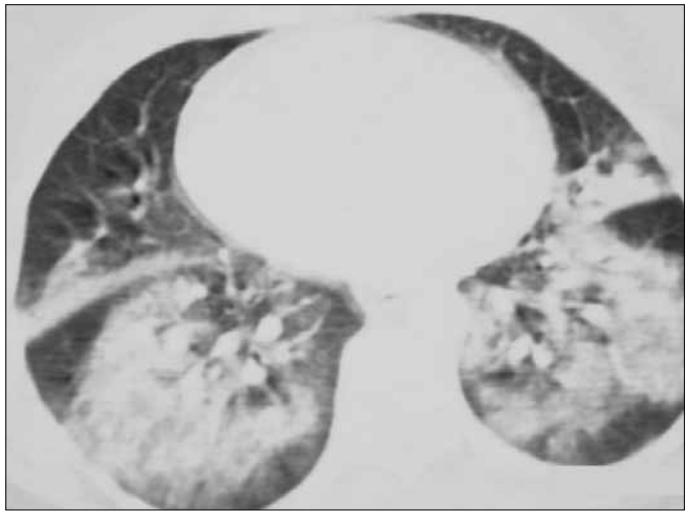

Figure 2. CT scan depicting bilateral patchy reticulonodular and ground-glass infiltrations in the lingula, middle lobe and lower lobes of the lungs

Discussion

Since PPCM can be easily misdiagnosed because of nonspecific symptoms, the emergency physicians should be alert about this disease. The true prevalance of PPCM is unknown. The incidence varies from 1 in 1300 to 1 in 15000 pregnancies. Because of geographical and diagnostic differences, the morbidity and mortality varies in a wide range of 25% to 90%. The causes of mortality are congestive heart failure, dysrhythmia and thromboembolic disease (5, 8-13).

Peripartum cardiomyopathy causes cardiac failure between the beginning of the last period of pregnancy and the first period of puerperium in a patient with no history of previous cardiac disease (5, 14). The complaints and symptoms are nonspecific, thus making an accurate diagnosis of PPCM is not always easy. Because PTE is the leading and threatening cause of maternal death in developing countries, PPCM is not usually recognized early (2, 8). The current case had no history or previous symptoms of cardiac disease, manifested with nonspecific symptoms, and was initially suspected to have PTE as reported in the other cases of PPCM. Distinguishing between the PPCM and PTE is very important since managements of these two diseases are quite different (2). Dyspnea, orthopnea, paroxysmal nocturnal dyspnea and fatigue are the most common symptoms of PPCM. Increasing fatigue is the late symptom of PPCM occurring during late pregnancy and the puerperium (2, 7).

Interstitial and alveolar edema and cardiomegaly are frequent findings on the chest x-ray (2, 14). Electrocardiographic findings are usually nonspecific and demonstrate sinus tachycardia or atrial fibrillation and nonspecific ST and T wave changes (7, 14). Echocardiography is a useful diagnostic procedure for PPCM and can also be useful for revealing other diseases such as massive PTE. Another advantage of echocardiography is that it is a noninvasive procedure easily performed at the bedside. Main echocardiographic findings in PPCM are left atrial and ventricular dilatation and a decrease in myocardial systolic function, as manifested by a decrease in the ejection fraction of the left ventricle. Our patient also had an impaired ejection fraction, and additionally, mitral insufficiency secondary to the dilatation of the left ventricle (2, 3, 6, 8).

The main treatment of PPCM is similar to that in other forms of heart failure: restricting water and salt, adding digoxin, diuretics and agents decreasing afterload (3, 4). PPCM can cause thromboembolic complications because of stasis during left ventricular dysfunction so the treatment should also include an anticoagulant (4, 8, 14).

Hydralazine and a combination drug with nitroglycerine or amlodipine are the preferred drugs during pregnancy. In the postpartum period, angiotensin converting enzyme inhibitors or angiotensin II receptor blockers are effective drugs of choice in decreasing afterload but are contraindicated during pregnancy. These drugs have teratogenic effects associated with serious renal dysfunction and increased fetal loss. Angiotensin-converting enzyme inhibitors are also not recommended during breastfeeding because they can pass to the breast milk (3, 4). Diuretics and digoxin are reported to cause adverse effects on fetuses in animals, but there are no data in human. Digoxin is believed to be safe in pregnancy and the postpartum period even though it crosses the placental barrier and is secreted into breast milk (8). Our patient was given an angiotensin-converting enzyme inhibitor, furosemide, calcium channel blocker and spironolactone for left ventricular failure, and enoxaparin for PTE prophylaxis. Since our patient did not breastfeed the baby, angiotensin converting enzyme inhibitors were given freely (4, 8, 14).

Cardiac transplantation is recommended to patients who do not respond to intensive medical treatment mentioned above (7, 9). Usually,

recovery occurs within about 6 months after the diagnosis (10). Our patient was followed up for 9 months and had complete clinical, radiological and echocardiographic recovery 5 months postoperatively. The patients with persistent left ventricular dysfunctions have risk factors of complications and mortality. Survival and prognosis is directly correlated to recovery of left ventricular function. Echocardiography is an important tool also for the follow up of recovery as well as guidance of treatment (8, 15, 16).

Conclusion

Peripartum cardiomyopathy (PPCM) is a rare, life-threatening heart disease with unknown etiology and occurs during the peripartum period in previously healthy women. When a female patient in peripartum has respiratory and cardiac failure, the physicians should be alert to this disease because the complaints are nonspecific. Survival and prognosis is directly correlated to recovery of left ventricular function. Echocardiography is an important tool also for the follow up of recovery as well as guidance of treatment.

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