

Case Report

Peripartum Cardiomyopathy: Case Report and Review of the Literature

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ABSTRACT

Peripartum cardiomyopathy (PPCM) is a rare but sometimes fatal form of heart disease. In this case report, we describe the development of PPCM in a young patient with a twin pregnancy. It is well-known from literature that twin pregnancy is considered a risk factor for

development of PPCM. However, the association of twin pregnancy and PPCM is unclear. Our patient improved remarkably despite the fact that PPCM has devastating consequences. We review the diagnostic criteria of PPCM and its management.

KEYWORDS: heart failure, peripartum cardiomyopathy, prognosis

INTRODUCTION

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' post partum^[1,2]. The incidence of PPCM ranges from 1:300 to 1:15,000 live births^[2]. Published mortality rates in PPCM generally range from 25 to 50% in the United States^[2].

Multiparity, twin births, advanced maternal age, pre-eclampsia, gestational hypertension, and black race are the known risk factors^[1-3]. The clinical presentation of patients with PPCM is similar to that of patients with congestive heart failure. Treatment includes digitalis, diuretic agents, and vasodilators. Anti-coagulation is also recommended in selected patients. The prognosis of PPCM is related to the recovery of ventricular function^[3]. Failure of heart size to return to normal is associated with excess morbidity and mortality^[3]. The risk of developing PPCM in subsequent pregnancies remains high, especially if left ventricular dysfunction is persistent^[3].

In this case report, we describe a female patient who developed PPCM after a twin delivery. The diagnostic criteria, therapeutic aspects, prognosis and future pregnancy of the PPCM are discussed.

CASE REPORT

A 23-year-old, non-Kuwaiti female, mother of two children, was admitted to the obstetrics department of Farwaniya Hospital in August 2000, for a twin delivery. Soon after admission she delivered the twins vaginally. On the following day,

she started to complain of progressive shortness of breath and chest discomfort. She gave no history of significant shortness of breath during her entire pregnancy. She had complained of fatigue but this was attributed to her twin pregnancy. She gave no history of any cardiac problems before and she denied any history of any similar problems in her past pregnancies. Her past medical history was otherwise unremarkable.

On examination, she was pale and mildly dyspnoeic. The blood pressure was 100/70 mm Hg, the pulse was 120 beats/minute, regular, and the respiratory rate was 34/minute. She was afebrile.

The chest examination revealed significant bilateral basal crackles. The cardiovascular examination showed a raised jugular venous pressure at about 8 cm above sternal angle. The cardiac auscultation showed normal first and second heart sounds, a third heart sound with a gallop rhythm. No significant cardiac murmurs were detected.

The abdominal examination was unremarkable. There was no pitting edema in lower extremities and no signs of deep vein thrombosis.

ECG showed sinus tachycardia with non-specific T wave inversion in the precordial leads. Chest X-ray showed pulmonary interstitial edema and increase in cardiac silhouette. Because of initial suspicion of pulmonary embolism, she underwent a V/Q scan, which was non-diagnostic. Doppler ultrasound of both lower limbs was also negative for deep vein thrombosis. The laboratory investigations were all normal.

Initial echocardiography showed dilated poorly contracting left ventricle. There was mild a mitral

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regurgitation, the ejection fraction was 32% and the left ventricular end-diastolic dimension was 60 mm (normal 35 to 56 mm).

A diagnosis of peripartum cardiomyopathy was made and the patient was treated for congestive heart failure with diuretics, digitalis and angiotensin converting enzyme inhibitor (ACE-I). She responded promptly to anti-failure therapy. Her improvement was rapid. She was discharged after two weeks of hospitalization. On follow-up visit, the improvement was sustained. Echocardiography was repeated on October 2000. It showed improvement in overall left ventricular function, the ejection fraction had increased to 40% and the left ventricular end-diastolic dimension was 55 mm.

She continued to do well and was seen again on January 2001. She denied any symptoms suggestive of recurrent congestive heart failure. She reported no adverse reactions to her medications. Repeated echocardiography showed an ejection fraction of 43%, and the left ventricular end-diastolic dimension was 55 mm.

The last time she was seen on follow-up was in March 2001 when her cardiac condition was stable. Clinically she had stable vital signs and no evidence of congestive heart failure signs. She was offered a beta-blocker but she refused. She was kept on the same regimen as she was on before with minor modification of dosage. She was advised strongly to avoid any future pregnancy and to continue regular medical follow-up.

DISCUSSION

This patient had normal pregnancy and delivered twin boys and developed PPCM post-delivery. This is in keeping with the literature that the majority of PPCM occurs post-delivery. It is important to know that the diagnosis of PPCM was made after excluding other differential diagnosis and by applying the diagnostic criteria of PPCM.

PPCM is defined on the basis of four criteria. These diagnostic criteria are: (a) development of cardiac failure in the last month of pregnancy or within five months of delivery; (b) absence of an identifiable cause for the cardiac failure; (c) absence of recognizable heart disease prior to the last month of pregnancy; (d) left ventricular systolic dysfunction demonstrated by classic echocardiographic criteria, such as depressed shortening fraction or ejection fraction^[3].

The true incidence of PPCM is unknown. An accepted incidence is one per 3000 to one per 4000 live births, which would translate to between 1000 and 1300 women affected each year in the United States^[2,3].

Although the etiology of PPCM remains unclear, a number of potential risk factors for this disorder have been proposed. Among these factors included

are multiparity, advanced maternal age, multifetal pregnancy, pre-eclampsia, gestational hypertension and women of African descent^[3]. Other risk factors include association with maternal cocaine abuse or selenium deficiency, long-term (more than four weeks) oral tocolytic therapy with beta adrenergic agonists such as terbutaline^[3]. However, the disease can occur in women without these risk factors.

A number of possible causes have been proposed for PPCM, including myocarditis, abnormal immune response to pregnancy, maladaptive response to the hemodynamic stresses of pregnancy, stress-activated cytokines^[3]. In addition, there have been a few reports of familial PPCM^[4].

The diagnosis of PPCM has been based upon the established four diagnostic criteria that were used in the definition. It is important to realize that the diagnosis of PPCM requires excluding other causes of cardiomyopathy before the diagnosis of PPCM is considered. Had our patient had underlying heart disease, we would have expected some symptoms during pregnancy or around delivery period.

One important aspect of this case is that she improved remarkably, and was discharged within two weeks of presentation. The natural course of the disease is variable and not well understood. It is also not known who will improve and what are the criteria for good prognosis.

Treatment of PPCM is similar to that for other types of congestive heart failure. The combination of digoxin, diuretics and sodium restriction, beta blockers and after load reduction forms the cornerstone of therapy^[5]. Careful attention must be paid to fetal safety and to excretion of drug or drug metabolites during breast-feeding after delivery.

Patients with PPCM are highly predisposed to thromboembolic phenomena. Thus anticoagulants should be considered in these patients^[3,5]. Immunosuppressive therapy has no clear cut efficacy and, therefore, is not currently recommended^[5]. Intravenous immune globulin may play a role in women with PPCM^[6]. Women who fail maximal medical management may be candidates for cardiac transplantation^[7].

The prognosis for women with PPCM appears to depend on the normalization of left ventricular size and function within six months after delivery^[3,8]. About half of the patients of PPCM recover without any complications^[8]. Most hearts that are destined to recover normal function probably do so within six months from the time of diagnosis^[1,9]. Persistence of disease after six months indicates irreversible cardiomyopathy and portends worse survival. The mortality estimates for patients with PPCM in the United States range from 25 to 50 percent;

most deaths occur within the first three months post partum^[1,9]. Death is usually caused by progressive pump failure, arrhythmias or thromboembolic events^[2,10]. Although our patient seemed to have full clinical improvement, concern remains because she did not have normalization of left ventricular function on echocardiography.

The persistence of cardiac dysfunction six to 12 months after the initial diagnosis of PPCM usually indicates an irreversible problem and almost always represents an absolute contraindication to a subsequent pregnancy^[11]. Elkayam *et al* showed that in women who have had PPCM, subsequent pregnancies might be associated with deleterious fetal and maternal outcomes such as premature delivery, maternal cardiac dysfunction, symptomatic heart failure and even death^[12]. The data have been conflicting in patients with PPCM in whom left ventricular function recovers^[13], but these women appear to have some risk for recurrence. Women with PPCM who have regained normal resting left ventricular size and performance have decreased contractile reserve as revealed by dobutamine challenge test^[14]. Therefore, subsequent pregnancies, if they cannot be avoided, should be managed in collaboration with a high-risk perinatal center^[13].

CONCLUSION

Peripartum cardiomyopathy is a rare disease of unknown cause that strikes women in the child bearing years. Diagnosis of PPCM is challenging and requires vigilance. Once PPCM is identified, the primary goal of therapy is to alleviate symptoms of congestive heart failure.

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