CASE STUDY

Peripartum Cardiomyopathy: Case Reports

Mary Wang, MD

Abstract

Peripartum cardiomyopathy (PPCM) is a dilated cardiomyopathy defined as systolic cardiac heart failure in the last month of pregnancy or within five months of delivery. PPCM, which affects thousands of women each year in the US, was first described in the 1800s, yet its etiology is still unclear. Its diagnosis is often delayed because its symptoms closely resemble those within the normal spectrum of pregnancy and the postpartum period. When PPCM is misdiagnosed or its diagnosis is delayed, the consequences for patients are deadly: The disorder carries a high mortality rate.

Introduction

Peripartum cardiomyopathy (PPCM) is associated with one in every 3000 to 4000 live births, affecting thousands of women in the US each year.[1] The definition of PPCM includes four criteria: 1) development of cardiac failure in the last month of pregnancy or within five months of delivery, 2) absence of an identifiable cause for the cardiac failure, 3) absence of recognizable heart disease before the last month of pregnancy, and 4) left ventricular (LV) dysfunction (ejection fraction of less than 45% or reduced shortening fraction).[2,3]

Risk factors include multiparity, black race, older maternal age, pre-eclampsia, and gestational hypertension.[1,4] Symptoms of PPCM, which include fatigue, edema, and dyspnea, are similar to those for the normal spectrum of peripartum states and pregnancy comordities such as pulmonary emboli and eclampsia.[5] Therefore, diagnosis is often delayed and the disorder is under recognized, with devastating consequences: Mortality is as high as 20% to 50%.[5] The following two case reports illustrate a typical presentation and an atypical one. This article also reviews the etiology, clinical symptoms, treatment, and prognosis for PPCM, which must be understood to provide patients with the most efficient and appropriate care.

Case 1

A white woman, age 29 years, presented to our urgent-care clinic five days after giving birth, reporting dyspnea and fatigue that had lasted two days. She said that because this was her first pregnancy, she thought her symptoms to be normal after delivery. However, when her husband insisted, she went to the clinic for evaluation. She was found to be dyspneic and hypoxic with saturation on room air in the low 80th percentile and was sent to the Emergency Department (ED) for further treatment.

Her medical history included obesity, but the patient was in relatively good health until approximately her last month of pregnancy, when she developed gestational hypertension (without other significant pre-eclampsia signs and symptoms), dependent peripheral edema, as well as some symptoms of an upper respiratory infection. She was given labetalol, 200 mg orally, twice daily, for blood-pressure management.

During examination in the ED, the patient was noted to be afebrile and had a blood pressure of 156/88 mm Hg, a pulse rate of 90 beats per minute, a respiratory rate of 20 breaths per minute, and an oxygen saturation of 95% while receiving oxygen through a 2-L nasal cannula. Her lungs were clear to auscultation and her heart rate was regular, with an S3 gallop. Her extremities were nonedematous, and she had no calf tenderness. Urinalysis results were negative for any proteins. Plasma levels of D-dimer and circulating levels of B-type natriuretic peptide (BNP) were 1981 pg/mL and 864 pg/mL, respectively. An electrocardiogram showed a normal sinus rhythm. Chest radiographs showed cardiomegaly with increased vascular congestion bilaterally. A computed tomography (CT) chest scan to evaluate for possible pulmonary emboli showed evidence of pleural effusion and cardiomegaly but no emboli.

The patient was subsequently admitted to the hospital for new-onset PPCM and was given furosemide intravenously for diuresis. A transthoracic echocardiogram done at admission showed an LV ejection fraction of 35% to 40%, with trace aortic and mitral regurgita-

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tion. Her fatigue and dyspnea greatly decreased with diuresis; she was discharged from the hospital three days later and instructed to take lisinopril and labetalol. Follow-up examination at six months showed a stable cardiomyopathy and well-controlled hypertension, and a repeat echocardiogram at the same point showed an improved ejection fraction of 55% to 60%.

Case 2

A white woman, age 25 years, presented at the ED with dyspnea six days after having given birth for the first time. She reported not having had any coughing, chest pain, or calf pain. Her medical history was significant for hypothyroidism, for which she was taking levothyroxine. Her pregnancy had been otherwise noneventful, except for flulike symptoms approximately one month before childbirth that included coughing, nausea, vomiting, and diarrhea.

In the ED, the patient appeared to be slightly anxious. She had a blood pressure of 159/87 mm Hg, a pulse rate of 58 beats per minute, a respiratory rate of 20 breaths per minute, and an oxygen saturation of 100% on room air. Physical examination showed no jugular venous distention, S3 heart sound, edema, or hepatosplenomegaly. She was slightly tachypneic but not in any acute respiratory stress. Laboratory tests revealed no proteinuria and a BNP level of 272 pg/mL. Echocardiography showed a normal sinus rhythm at 46 beats per minute. Chest radiograph findings were noted to be normal, with no cardiomegaly or pulmonary congestion. A CT scan of the chest was negative for pulmonary emboli.

The patient was discharged from the care of the ED physician with a diagnosis of dyspnea secondary to breast enlargement versus chest wall pain. She kept an appointment in our family medicine clinic four days later for a follow-up examination and still had the same symptoms. An echocardiogram ordered at that time showed left atrial and LV dilation, global LV hypokinesis, and an estimated ejection fraction of 25% to 30%. A cardiology consultation was done and the patient was given appropriate medications for heart failure, which alleviated her symptoms. Her follow-up echocardiogram three months later showed normalization of her ejection fraction to 60%.

Discussion

Approximately 60% to 70% of women experience a sensation of dyspnea during the course of normal pregnancy.[6] Although historically PPCM risk factors occur in older women and in black women, contemporary trends show that there is an increasing incidence (24%–37%) in young primigravid and white patients.[7–9] The details of the two cases reported here support this trend; the women are both young primigravidas and are white. Because dyspnea is a common finding in normal pregnancy and even in the initial postpartum state, PPCM is often missed, especially if the patient population does not fit the typical epidemiology.

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Etiology

A possible relationship between pregnancy with dilated cardiomyopathy was recognized as early as the 1870s[10] and was classified as a distinct clinical entity in the 1930s.[11] Yet the cause of PPCM is still unknown. Most postulate that it is related to the cardiovascular stress of pregnancy (increased fluid load); others have suggested myocarditis. Felker et al[12] found that 26 of 51 women with PPCM had histologic evidence of myocarditis on endomyocardial biopsy. Other researchers further postulate that PPCM may be an inflammatory response in pregnancy, citing an elevation of tumor necrosis factor-alpha and interleukin-6 levels.[13,14] Some evidence also suggests that it may be a pathologic autoimmune response to fetal cells that lodge in the maternal circulation and cardiac tissue.[3] There is also conflicting evidence whether nutritional deficiencies—more specifically, selenium deficiency—is a cause for PPCM.[15,16]

Clinical Features and Diagnosis

Clinical features of PPCM include symptoms of congestive heart failure and chest pain. Signs can include tachycardia, tachypnea, pulmonary rales, an enlarged

Table 1. Signs and symptoms in peripartum cardiomyopathy vs normal pregnancy, pulmonary embolism, and upper respiratory infection

Pregnancy	PPCM	PE	URI
Fatigue	Fatigue	Fatigue	Fatigue
Tachycardia	Tachycardia	Tachycardia	
Dyspnea	Dyspnea	Dyspnea	
Edema	Edema	Edema	
	Chest pain	Chest pain	
	DOE	DOE	
	PND/orthopnea	PND/orthopnea	
	Rales	Rales	
	S3 heart sound	S3 heart sound	
	Cough		Cough
	Hepatosplenomegaly		

DOE = dyspnea on exertion; PE = pulmonary embolism; PND = paroxysmal nocturnal dyspnea; PPCM = peripartum cardiomyopathy; URI = upper respiratory infection.

heart, and an S3 heart sound.[4] Such signs and symptoms overlap with those of many other conditions, ranging from normal pregnancy to pulmonary emboli and upper respiratory infection (Table 1).

Diagnosis of PPCM includes the four criteria described at the start of this report. There are no specific laboratory abnormalities for PPCM, although BNP is often elevated. However, other exclusionary laboratory studies should also be considered, including cardiac enzymes assessment and a pre-eclampsia workup. Imaging studies include electrocardiography, chest radiography, and echocardiography. Electrocardiographic findings are often normal but can include sinus tachycardia, nonspecific ST- and T-wave abnormalities, and voltage abnormalities.[17] Chest radiographs can show signs of pulmonary congestion, cardiac enlargement, and even pleural effusions in some cases.[9] Echocardiograms usually show decreased contractility and LV enlargement without hypertrophy.[18]

Treatment and Prognosis

The treatment for PPCM is the same as for other forms of congestive heart failure (fluid and salt restriction, β-blocker, diuretic, and digoxin), except for angiotensin-converting enzyme inhibitors and angiotensin-receptor blockers, which are contraindicated in pregnancy.[1] Hydralazine can be used during pregnancy to reduce afterload.[4] Diuretics can be used cautiously during pregnancy to prevent dehydration and placental insufficiency. Patients with PPCM are also at high risk for thrombus formation;[19] thus, anticoagulation should be considered especially for high-risk patients with severe LV dysfunction. In addition, physical activity should be encouraged according to patients' tolerance of symptoms.

The best time to discontinue these medications is unknown, but their use should be continued for at least one year.[7] If medical treatments are not successful, heart transplantation is often the last resort. Fortunately, in recent years, the rate required transplantation has decreased to about 4% to 7%.[20] Transplantation success rates are good with favorable long-term survival rates.[21]

In about 50% of patients, the ejection fraction normalizes. Regardless of recovery, however, a second pregnancy is usually not recommended for these patients because PPCM recurs in more than 30% of subsequent pregnancies, which puts both mother and baby at great risk.[1]

Conclusion

The cases presented here demonstrate the variability of clinical presentation of PPCM. Case 1 illustrates a typical PPCM presentation, with gestational hypertension, S3 gallop, hypoxia, an elevated BNP level, cardiomegaly, and pulmonary congestion on chest radiographs. Case 2 illustrates an atypical PPCM presentation, with no abnormal findings on physical or imaging studies, yet with a lower ejection fraction shown by echocardiography. Thus, it is important that physicians be familiar with PPCM and therefore consider it when diagnosing dyspneic patients to expedite medical treatment for a potentially lethal condition. ❖

Disclosure Statement

The author(s) have no conflicts of interest to disclose.

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