Case Report

Peripartum Cardiomyopathy

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Abstract

Peripartum cardiomyopathy (PPCM) is an idiopathic cardiomyopathy that presents with heart failure secondary to left ventricular systolic dysfunction. Onset is from the last trimester of pregnancy to 5 months postpartum. Diagnosis in the last trimester is complicated by the fact that the early symptoms of this disorder may mimic the symptoms of normal pregnancy. However, it is essential for the practitioner dealing with such population to have a high degree of clinical suspicion for early diagnosis and management. Echocardiography is used to diagnose this entity and monitor the therapy. We present a case report of a 40-year-old woman who presented two days post-partum with respiratory distress and early echocardiography helped in diagnosing PPCM. The aim of this report is to make health professionals aware of the possibilities of PPCM in a woman with dyspnoea in the postpartum period.

Keywords: *Peripartum cardiomyopathy, Left ventricular dysfunction, Echocardiography.*

Introduction

Peripartum cardiomyopathy (PPCM) is an uncommon type of heart failure of unknown cause occurring late in pregnancy or in the postpartum period. Although PPCM is associated with high morbidity and mortality it is often undetected or misdiagnosed because of its low incidence and nonspecific symptoms. Additionally, pregnancy-related diseases may have similar clinical presentations. Unfortunately, PPCM is a disease whose underlying etiology and natural history remain incompletely understood.

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Risk 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. This article 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 Report

A 40 years old woman came to the emergency department with sudden onset of dyspnoea two days after giving birth of her first child by normal vaginal delivery. She denied any chest pain or calf pain. Her medical history was uneventful, except for flulike symptoms approximately one month before childbirth that included coughing, nausea, vomiting, and diarrhea. In the emergency department, the patient appeared to be slightly anxious. She had a blood pressure of 140/80 mm Hg, a pulse rate of 110 beats per minute, a respiratory rate of 22 breaths per minute, and an oxygen saturation of 93% on room air. Physical examination showed S3 heart sound, bilateral basal crackles both lungs but no jugular venous distention, edema, or hepatosplenomegaly . She was slightly tachypnoeic but not in any acute respiratory stress. Laboratory tests revealed no proteinuria and a BNP level of 1250 pg/mL. ECG showed a sinus tachycardia, rate 110 beats per minute. Chest radiograph findings were noted to cardiomegaly with mild pulmonary congestion (Figure 1).

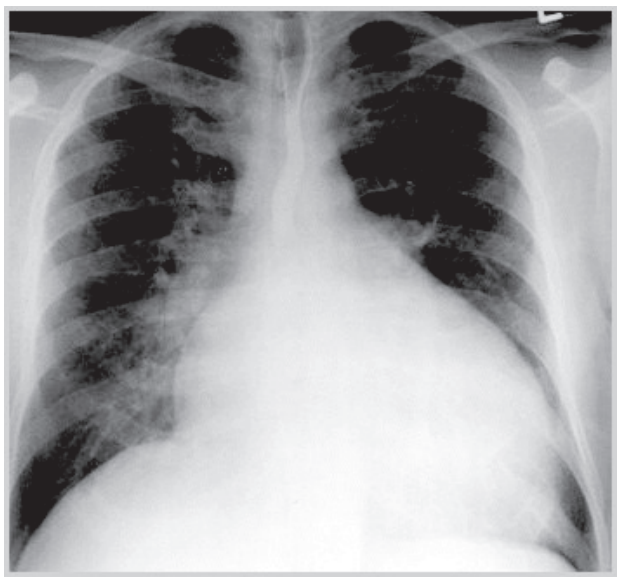

Figure-1: Chest X-ray P/A view

A CT scan of the chest was done which was negative for pulmonary emboli. An echocardiogram was done at that time which showed left atrial and LV dilation, global LV hypokinesia, and an estimated ejection fraction of 30% to 35% & Grade 1+ MR (Figure-2). 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 56-58%.

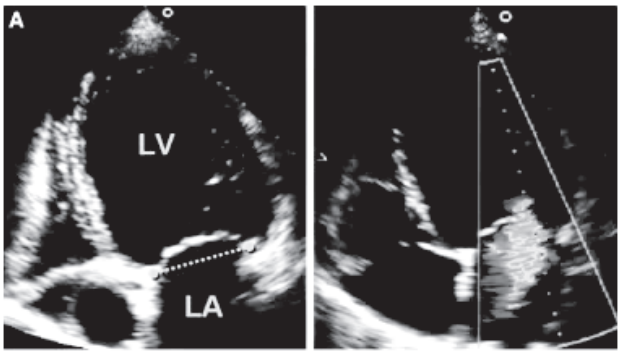

Figure-2: TTE, parasternal long-axis view: LV = left ventricle; LA = left atrial.

Discussion

Heart failure associated with pregnancy was first observed and recorded as early as the 19th century by Ritchie and Virchow. Adapted from work by Demakis et al.[1]. The exact incidence of PPCM is unknown but is estimated to be 1 in 3000-4000 deliveries in the United States[2]. Specific data is rare because mild forms are often unrecognized. PPCM constitutes less than 1% of all cardiovascular events related to pregnancy[3]. Risk factors classically identified in the literature include black race, increase age (>30 years), multiparity, twin pregnancy, pre eclampsia and gestational hypertension[1,4].

Despite many hypotheses, the cause and mechanism of pathogenesis of PPCM remain unknown. Early investigations proposed myocarditis as the cause for PPCM where anti-inflammatory treatment resulted in clinical improvement[5]. Further studies failed to establish this causal link between mycarditis and development of PPCM[6,7]. Additionally, Felker et al. confirmed that the absence or presence of inflammation on endomyocardial biopsy tissue did not predict outcome in patients with PPCM[8].

Persistent viral antigen has been also postulated as a trigger for the development of PPCM. Presence of viral genomic material, including enterovirus (coxsackie virus), parvovirus B19, adenovirus and hepatitis virus was isolated in biopsy material of patients with idiopathic dilated cardiomyopathy. Further, the authors demonstrated an association between clinical improvement of left ventricular systolic function and viral clearing[9]. Despite these promising findings, further studies are needed to clearly identify virus infection as the cause for development of PPCM. Many animal and clinical investigations support to the hypothesis that immune activation, dysregulation of cellular apoptosis pathways and upregulated humoral immunity contribute to the pathogenesis of PPCM. Whereas the importance of raised high sensitivity c-reactive protein in plasma of patients with new and evolving PPCM merits additional evaluation, concentrations of the inflammatory cytokine TNF-alpha, elevated immunoglobulins against cardiac myosin as well as markers of apoptosis (Fas/Apo-1) are well correlated with left ventricular function and mortality. Additionally, animal data suggest that the cardiac myocyte-specific STAT3 pathway is necessary for protection of the heart from postpartum stress[10].Taken all available data together, so far no cause has been clearly identified for the development of PPCM.

The clinical presentation of patients of PPCM is similar to that of other forms of left ventricular systolic failure. Clinical symptoms are dyspnea, cough, chest pain, and orthopnea. Additionally physical examination may reveal signs such as tachycardia, hypertension, ventricular gallop rhythm, mitral regurgitation murmur, hepatosplenomegaly, ascites, or jugular venous distension. Ancillary studies include electrocardiogram (ECG), chest radiograph, Doppler-echocardiography and possibly thoracic CT-scan. To recapitulate, proper medical history and an accurate physical examination combined with routine ancillary studies may ensure the diagnosis of PPCM. Clinicians should think of PPCM in any peripartum patient with unexplained disease.

Medical therapy of PPCM is similar to that for others forms of congestive heart failure. Thus digoxin, angiotensin-converting enzyme (ACE) inhibitors, diuretics, sodium restriction and after-load reducing agents are the mainstays of medical therapy[11-13]. If necessary, catecholamines like dopamine and dobutamine can be given. Arrhythmia treatment should be performed using standard guidelines and protocols. Low molecular weight heparin prophylaxis is routinely advised. Immunosuppressive therapy may be indicated when peripartum myocarditis and cardiomyopathy is present[5]. Intra-aortic balloon pumping or insertion of a left ventricular (LVAD) or biventricular assist device (BVAD) should be considered if the patient is not responding to medical management. As a last resort heart transplantation can be considered for non responders.

Peripartum cardiomyopathy is an uncommon complication of pregnancy with unknown cause and potentially life-threatening complications. Once the diagnosis is confirmed treatment of PPCM does not differ from that of other dilated cardiomyopathies. Echocardiography appears to be extremely valuable in diagnosing PPCM, formulating prognosis of recovery and following the course of the disease. Especially ejection fraction and left ventricular end diastolic dimensions at the time of diagnosis may be predictive of long-term cardiac dysfunction. Currently, there is no consensus regarding recommendations for future pregnancy after PPCM. Nevertheless the risk of developing PPCM, with future pregnancies, remains high. Particularly in the presence of persistent left ventricular dysfunction, a subsequent pregnancy should be discouraged and avoided[14].

Prognosis in the United States is variable with a maternal mortality of 25 to 50 percent[14]. However outcome differs widely between reports. Death is generally attributed to arrhythmia or thrombo-embolism. If congestive cardiomyopathy persists after 6 months it is likely irreversible and portends decreased survival.

References

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