Case Report

Peripartum/ Postpartum Cardiomyopathy in a Young Woman with Breathlessness - A Case Report

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Abstract:
Peripartum cardiomyopathy (PPCM) is an unusual and uncommon form of dilated cardiomyopathy that is often fatal to young women. Though it can affect women of all races, data regarding its incidence is lacking in our country while in USA the disease occurs in 250-1350 women each year. It involves systolic dysfunction of the heart with increased risk of atrial and ventricular arrhythmias, thromboembolism and even sudden cardiac death. Here we report a young lady with postpartum cardiomyopathy, who presented to us with dyspnoea, palpitation and leg edema.

Keywords: Peripartum/ Postpartum cardiomyopathy (PPCM)

Introduction:
Peripartum cardiomyopathy is defined as deterioration in cardiac function presenting typically between the last month of pregnancy and up to five months postpartum[1]. As with other forms of dilated cardiomyopathy, PPCM decreases the left ventricular ejection fraction (EF) with associated congestive heart failure. Myocarditis of viral, immunologic or idiopathic etiology has been suggested. It is a diagnosis of exclusion, where patients have no prior history of heart disease and there are no other known possible causes of heart failure. Echocardiogram is used to both diagnose and monitor the effectiveness of treatment for PPCM. Primary therapy consists of bed rest, sodium and fluid restriction, vasodilators, digoxin, diuretics and sometimes cardiac transplantation, anticoagulation etc. Prognosis seems dependent on recovery of left ventricular function. The usual causes of death in patients with PPCM are progressive heart failure, arrhythmia or thromboembolism[2].

Case Report:
A 25 years old lady was admitted to the Medicine department of BIRDEM hospital with dyspnoea, palpitation, fatigue and pitting pedal edema that started 4 days after the birth of her first baby. She said that because this was her first pregnancy, she took her symptoms to be normal after delivery. However she was in relatively good health throughout pregnancy except excessive weight gain during last month of pregnancy. When she presented to us she had orthopnea, respiratory rate of 20 breaths per minute and an oxygen saturation of 95% while receiving oxygen through a 2-L nasal cannula. The patient was noted to be afebrile, mildly anaemic and had a blood pressure of 130/85 mm Hg, a regular pulse rate of 90 beats per minute and elevated jugular venous pressure. Cardiac auscultation revealed a systolic murmur at the left sternal edge with an S3 gallop. This pulmonic arterial flow murmur tends to become quieter during inspiration and her lungs revealed bibasal crepitation. There was no ascites, arrhythmias, embolic phenomenon and hepatomegaly.

The hemoglobin level was 11 g/dl and there was no thrombocytopenia. Troponin-I was normal and ECG showed sinus tachycardia. An electrocardiography revealed LV ejection fraction of 35%. Chest radiographs showed cardiomegaly with increased vascular congestion bilaterally. Urinalysis results were negative for any proteins. Serum creatinine was 0.8 mg/dl, serum electrolytes were within normal limit, liver enzymes were normal. EGFR was 100ml/min. Plasma levels of D-dimer was normal and circulating levels of BNP was high. Frequent cardiac consultation was taken. She was treated with digoxin, nitrates, carvedilol, furosemide, spironolactone, antiplatelet along with calcium and vitamins. She was also advised for fluid and salt restriction.

Her symptoms greatly decreased with medication. She was discharged from the hospital 7 days later and was instructed to take above medication. Follow-up examination at first month showed a stable cardiomyopathy and well-controlled BP, and a repeat echocardiogram at the same point showed an improved ejection fraction of 55% to 60%.

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Discussion:

Approximately 60% to 70% of women experience a sensation of dyspnea during the course of normal pregnancy. 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. 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. The cause of PPCM is still unknown. Most postulate that it is related to the cardiovascular stress of pregnancy; others have suggested myocarditis. Felker et al found that 26 of 51 women with PPCM had histologic evidence of myocarditis on endomyocardial biopsy[3]. Other researchers further postulated that PPCM may be an inflammatory response in pregnancy, citing an elevation of tumor necrosis factor-alpha and interleukin-6 levels[4]. Some evidence also suggests that it may be a pathologic autoimmune response to fetal cells that lodge in the maternal circulation and cardiac tissue. There is also conflicting evidence whether nutritional deficiencies—more specifically, selenium deficiency is a cause for PPCM[5]. Because dyspnea is a common finding in normal pregnancy and even in the initial postpartum state, PPCM is often missed[6].

Clinical features of PPCM include symptoms of congestive heart failure and chest pain. Signs can include tachycardia, tachypnoea, pulmonary rales, an enlarged heart, and an S_3 heart sound[7]. Such signs and symptoms overlap with those of many other conditions, ranging from normal pregnancy to pulmonary emboli and upper respiratory tract infection. 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[8]. 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[10]. These findings matched with ours as she was found to be dyspnoeic with bilateral pitting pedal edema that started 4 days after the birth of her first baby. Troponin-I was normal and ECG showed sinus tachycardia. ECHO revealed LV ejection fraction of 35%. Chest radiographs showed cardiomegaly with increased vascular congestion. Urinalysis was negative for proteins with normal creatinine, serum electrolytes, liver enzymes and D-dimer but BNP was high.

The treatment for PPCM is the same as for other forms of congestive heart failure - fluid and salt restriction, Beta-blocker, diuretic, and digoxin except for angiotensin-converting enzyme inhibitors and angiotensin-receptor blockers, which are contraindicated in pregnancy. Diuretics should be used cautiously during pregnancy to prevent dehydration and placental insufficiency. Patients with PPCM are also at high risk for thrombus formation, thus, anticoagulation should be considered especially for high-risk patients with severe LV dysfunction. In addition, physical activity should be encouraged according to patient's tolerance of symptoms. The best time to discontinue these medications is unknown, but their use should be continued for at least one year. We advised our patient accordingly.

Conclusion:

It is important that physicians be familiar with PPCM and therefore consider it when diagnosing dyspnoeic patients to expedite medical treatment for a potentially lethal condition. Patients with full recovery should be told that although a chance of recurrence exists, the mortality is low and the majority of such women have normal pregnancies. The patient may be willing to accept the risk of an adverse outcome, but the physician should make an objective recommendation, document it and not compromise his or her best medical judgement because of a patient's emotional desires.

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