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Case Report

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How Not to Miss Pregnancy Related Cardiomyopathy?

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Case Report

36 year old G5P4 African American female at 37 weeks presented with 2 week history of fatigue, shortness of breath on walking a few steps, and cough. Examination was significant for mild tachycardia at 110 beats per minute, oxygen saturations of 90%, and crackles across both lung fields. Chest X-ray revealed bilateral pulmonary infiltrates. The patient was hospitalized with a presumptive diagnosis of community acquired pneumonia. She was started on broad spectrum antibiotics. Her condition deteriorated over the next 48 hours requiring emergent cesarean delivery due to category III fetal heart rate tracing under general anesthesia. Postoperative course was complicated by persistent hypoxemia and difficulty maintaining oxygen saturations. Further work up was initiated. Echocardiogram revealed 4 chamber dilatation and left ventricular ejection fraction (EF) of 20%. Diagnosis of peripartum cardiomyopathy was made.

This case highlights a combination of risk factors and warning signs for cardiomyopathy: advanced maternal age, African background, multiparity, persistent symptoms interfering with day to day life along with mild tachycardia and physical examination and radiographic evidence of pulmonary congestion. There is no evidence of a febrile illness that typically is seen in patients presenting with pneumonia. Presence of bilateral pulmonary infiltrates should alert the clinician to a likely diagnosis of cardiac source of pulmonary edema.

What is PPCMP?

Peripartum cardiomyopathy (PPCMP) also known as pregnancy associated cardiomyopathy (CMP) is an idiopathic dilated CMP presenting in the last month of pregnancy or within 5 months postpartum associated with depressed left ventricular systolic function. Additional criteria include absence of identifiable cause for heart failure, absence of recognizable heart disease prior to the last month of pregnancy, and left ventricular systolic dysfunction demonstrated by echocardiography [1-3]. It is a diagnosis of exclusion. It should be noted that about 20% of cases PPCMP present in the second or early part of third trimester. Clinical profile and outcomes in women presenting with early onset disease are similar to those who fulfill traditional criteria for PPCMP [4].

How often is it seen in Pregnancy and what are the Risk Factors?

Incidence varies across the world from as high as 1:300 in Haiti to 1:3,000-4,000 deliveries in the United States [5-7]. The precise etiology of PPCMP remains unknown. Several hypotheses have been proposed. Most recent data suggest that increased oxidative stress due to cleavage of prolactin by a protease cathepsin D transforming prolactin into an angiostatic and pro-apoptotic sub fragment leads to myocardial damage. Other theories indicate inflammation, infection, genetic predisposition and fetal chimerism may play a role. Risk factors for development of PPCMP include older age, non-Hispanic African American or Filipino heritage, grand multiparity, multiple gestations, hypertensive disorders of pregnancy, severe anemia, smoking, cocaine abuse and malnutrition [8].

Is PPCMP Dangerous?

PPCMP is among the leading causes of maternal mortality in the United States. Mortality rates vary from 1.4% to 30% in the highest risk populations. African American women are particularly vulnerable. They are at six times higher risk to die from this disease when compared to other ethnic groups [5,9,10]. Predictors of increased morbidity and mortality include severe LV dilation and depressed systolic function at the time of diagnosis, presence of LV thrombus, poor NYHA functional class, older age, multiparity, increased brain natriuretic peptide (BNP) levels, and evidence of myocardial damage, i.e. high troponin T values [11-14]. Prognosis varies from complete recovery to rapid deterioration. Approximately 50% of patients would recover, and this group generally would demonstrate significant improvements in left ventricular systolic function with normalization of their EF within 6 months of delivery.

How does PPCMP Present?

Patients usually present with symptoms of congestive heart failure (CHF) late in pregnancy or in the early postpartum period, i.e. dyspnea, cough, orthopnea, fatigue, palpitations, peripheral edema and chest pain. The most common presenting symptoms are shortness of breath and cough. Typical signs of heart failure (HF) including cardiomegaly, tachycardia and a third heart sound (S3) are noted in more than 85% of patients with PPCMP [15]. Anytime, a pregnant woman presents with shortness of breath, PPCMP should be strongly considered in the differential diagnosis.

Initial presentations of sudden cardiac death, peripheral emboli (cerebral, mesenteric, pulmonary) and acute hepatic failure in the postpartum period have reported in rare instances. Diagnosis of PPCMP should also be entertained in a peripartum patient with unexplained disease [3].

What are the Common Symptoms of Pregnancy?

It is important to understand the cardiovascular physiology during pregnancy that includes increases in blood volume, cardiac output, heart rate and a decrease in diastolic blood pressure. These hemodynamic alterations lead to onset of new symptoms, changes in physical examination findings and restrict our ability to accurately interpret diagnostic tests. Most pregnant patients experience reduced exercise tolerance and easy fatigability. Light-headedness may occur due to mechanical compression of the inferior vena cava by the gravid uterus leading to poor venous return to the heart, especially in the

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third trimester. Other frequent complaints include hyper-ventilation or shortness of breath and orthopnea from mechanical pressure of enlarged uterus on the diaphragm [16,17]. Palpitations are common and probably are related to the hyper dynamic circulation of pregnancy rather than arrhythmias in most cases. The biggest challenge for an obstetrician is to differentiate complaints due to physiologic changes in pregnancy from those of potentially serious underlying cardiac disease.

Mild and gradual onset shortness of breath without additional symptoms is likely to be physiologic in nature. Whereas, additional symptoms of cough, fatigue, orthopnea, palpitations, chest pain, swelling or severe limitation of functional capacity may be suggestive of an underlying cardiac disease. The diagnosis is generally delayed due to overlap of symptoms with normal complaints of pregnancy. There is evidence to suggest that delays in diagnosis may lead to a significant increase in major adverse events including the need for cardiac transplantation and even death [11]. Cardiac etiology should remain a potential suspect when a pregnancy woman presents with shortness of breath or any other cardiac symptom Figure 1.

What Tests are Indicated?

Patients suspected of PPCMP should undergo electrocardiogram (EKG) and echocardiogram in addition to oxygen saturations and laboratory testing including complete blood count, serum electrolytes, thyroid and renal function.

BNP or N-terminal pro-BNP (NT-proBNP) is an extremely useful, relatively cheap, readily available test that may help triage patients with potential heart failure diagnosis [18]. Serial measurements may assist during follow up and evaluate response to heart failure therapy. BNP is a neuropeptide released by the ventricles in response to pressure or volume overload. Even though pregnancy is a state of volume overload, BNP levels are comparable to non-pregnant controls and remain normal (median <20 pg/mL) throughout uncomplicated pregnancy and postpartum period [19]. BNP levels offer tremendous value in establishing or excluding potential diagnosis of heart failure in an acute care setting [20].

Chest radiograph, cardiac catheterization, endomyocardial biopsy and viral serologies may be considered in select cases.

Is there any Treatment?

Patients with a diagnosis of PPCMP should be delivered after stabilization of the mother. Principles of therapy are similar to that in the non-pregnant state including bed rest, fluid and salt restriction and medical therapy. Medical therapy comprises of diuretics, vasodilators, angiotensin converting enzyme (ACE) inhibitors and beta blockers. ACE inhibitors are contraindicated during pregnancy, however, can safely be used in non lactating postpartum women. Digoxin may be considered in pregnant patients with PPCMP. Patients with EF of <35% are at risk or thromboembolism and therefore, anticoagulation should be considered. There is some evidence that treatment with

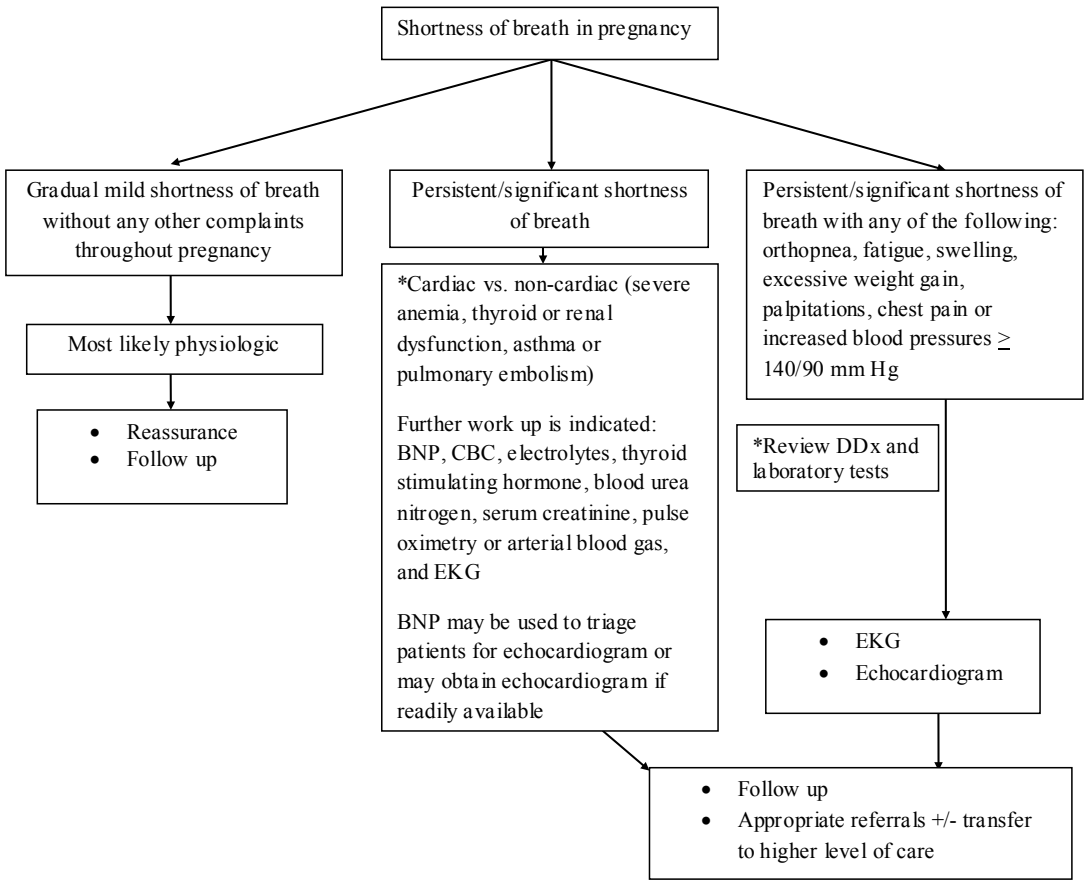

Figure 1: Shortness of breath in pregnancy.

prolactin inhibitors (prolactin) may improve survival in postpartum patients with PPCMP when used in addition to the standard heart failure therapy [3].

Can PPCMP recur?

Future outcomes with a previous history of PPCMP are related to the extent of recovery of left ventricular EF. Patients with recovered LV ejection fraction (EF) have a 20% risk of developing heart failure (HF); and those with persistent LV dysfunction have up to 30% risk of HF and 17% risk of maternal mortality in their subsequent pregnancy [21]. There is considerable controversy regarding the safety of subsequent pregnancy in patients with a history of PPCMP and normalization of LV function. It is recognized that LV systolic function may decline with the subsequent pregnancy even in patients who had normalization of their LV function. Careful pre-pregnancy counseling and discussion about the risks including the potential for life-threatening complications should be outlined with the patient prior to proceeding with a subsequent pregnancy.

How not to miss this deadly diagnosis?

- Maintain a high index of suspicion for PPCMP for any pregnant woman:
  - Who self reports symptoms of shortness of breath, fatigue, palpitations, swelling or chest pain
  - Has symptoms or complaints that are persistent and out of proportion to those anticipated due to routine pregnancy
  - With unexplained tachycardia or excessive weight gain
- Obtain B-type natriuretic peptide as a preliminary test for reassurance/triage if unsure
- EKG and echocardiogram
- Obtain appropriate consultations

Complacency in the diagnosis and management of PPCMP may have direct consequences for both the mother and the fetus. Therefore it is essential to maintain a high index of suspicion for underlying cardiac etiology for pregnant women with persistent cardiac symptoms. Heightened awareness will allow prompt diagnosis to establish management plan and outline the risks of adverse outcomes during pregnancy, delivery, and postpartum. In general, such women should be referred to a tertiary care center for a multidisciplinary management by a team of high risk obstetrician, cardiologist, anesthesiologist, and neonatologist.

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