Peripartum Cardiomyopathy : A Rare Case Report And A Brief Review of Literature

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ABSTRACT : *Peripartum cardiomyopathy (PPCM) is a rare form of heart failure with a reported incidence of 1 per 3000 to 1 per 4000 live births and a fatality rate of 20%-50%. Onset is usually between the last month of pregnancy and up to 5 months up postpartum in previously healthy women. Peripartum cardiomyopathy is a relatively rare disease, which can have devasting consequences and should be promptly identified and correctly treated. Overall prognosis is good in majority of the cases, although some patients may progress to irreversible heart failure. We describe the case of a 22 year-old-female who presented with dyspnoea, fatiguability, peripheral edema and abdominal distension. Early diagnosis is important and effective treatment reduces mortality rates and increases the chance of complete recovery of ventricular systolic function.*

KEY WORDS : *Peripartum cardiomyopathy, heart failure, echocardiography.*

INTRODUCTION

Peripartum cardiomyopathy (PPCM) is an idipathic cardiomyopathy, characterized by the left heart failure, left ventricular systolic dysfunction towards the end of pregnancy or within 5 months of delivery, when no other cause of heart failure is found[1].There are no data about the incidence of PPCM in Lithuania. In the USA, there is one case in 2300 women giving birth and about 1300 new cases a year [2].

The incidence in South Africa is higher because of the Africo-American women. Peripartum cardiomyopathy (PPCM) is associated with one in every 3000 to 4000 live births, affecting thousands of women in the US each year[3].

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 [4,5].

CASE REPORT :

A 22 year-old-female, five days after giving birth presented to our intensive care unit with progressive dyspnoea, orthopnoea, and paroxysmal nocturnal dyspnoea associated with palpitation that had lasted for 2 days. She experienced chest pain, sweating and fatiguability and noticed peripheral edema & abdominal distension. Her past medical and family history was insignificant.

On examination, she was averagely built with weight of 56kg. She was conscious, oriented, pale and was in severe respiratory distress. She was febrile(100 F) with pulse rate of 130/min, blood pressure of 160/100 mm of Hg, respiratory rate of 36/min and oxygen saturation of 86% on room air. Her jugular venous pressure was elevated at 22cmH2O. The 1st heart sound was normal and second heart sound normal with left ventricular summation gallop. Chest examination revealed bilateral basal crepitations. Abdominal examination revealed pulsatile, tender hepatomegaly with a liver span of 13 cm and moderate ascites.

Laboratory evaluation revealed hemoglobin of 10g/dl, total leukocyte count of 8500/mm3 and platelet count 1,30,000/mm3. Kidney and liver function tests were normal. Glucose in blood chemistry is 100mg/dl. Urine examination was normal. C reactive protein 0.21mg/dl (normal value <0.5), BNP 689 pg/ml (normal –value <100pg/ml). Thyroid profile was normal. Serological tests for the usual viral agents responsible for myocarditis were negative. Chest X ray showed cardiomegaly with pulmonary congestion.

A computed tomography (CT) chest scan to evaluate for possible pulmonary emboli showed evidence of pleural effusion and cardiomegaly but no emboli. ECG demonstrated sinus tachycardia. The initial echocardiogram revealed a dilated left ventricle, grade 1 diastolic dysfunction of the LV & LA with severe LV systolic dysfunction with LVEF 25-30% and small pericardial effusion.

She was in frank failure and treated with I.V. Diuretics (furesemide), beta-blocker(carvedilol). She was anticoagulated initially with unfractionated with

heparin and maintained on heparin. Her fatigue and dyspnoea greately decreased with diuresis. She responded well to the treatment and was discharged on 15th day. Chest X ray on day 15 showed resolution of the pulmonary edema and pleural effusions.

She was recommended to continue her treatment along with a low salt diet and reduced physical activity. She was on regular follow-up and repeat 2Decho after 6 month revealed a normal LV, LA & RV and disappearance of the small pericardial effusion with LVEF was 30-35%. Here, we report a case of Peripartum cardiomyopathy which highlights the importance of recognition of symptoms and treatment of cardiac failure in the peripartum period.

DISCUSSION

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. PCM usually presents with classical symptoms and sign of systolic heart failure with ventricular enlargement and dysfunction seen on echocardiography. Often there is significant mitral and tricuspid regurgitation [6]. Risk factors include multiparty, black race, older maternal age, pre-eclampsia, and gestational hypertension[7]. Symptoms of PPCM, which include fatigue, edema, and dyspnea, are similar to those for the normal spectrum of peripartum states and pregnancy co-mordities such as pulmonary emboli and eclampsia. Therefore, diagnosis is often delayed and the disorder is under recognized, with devastating consequences: Mortality is as high as 20% to 50%[8].

Unusual presentations include thrombo-embolism or hepatic failure secondary to heart failure. The development of heart failure and the usual time of diagnosis are during the post-partum period in more than 90% of the cases [9]. PCM can occur at any age with a higher incidence in women older than 30 years [10].

A possible relationship between pregnancy with dilated cardiomyopathy was recognized as early as the 1870s [11] and was classified as a distinct clinical entity in the 1930s. 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. Some evidence also suggests that it may be a pathologic autoimmune response to fetal cells that lodge in the maternal circulation and cardiac tissue [13,14]. There is also conflicting evidence whether nutritional deficiencies more specifically, selenium deficiency is a cause for PPCM [15].

Gestational hypertension, tocolytic therapy and twin pregnancy have been proposed as possible risk factors because they were commonly associated with PCM [10]. The association between PCM and twin pregnancy could support the theory of autoimmunity as a possible mechanism. This could depend on an excessive traffic of hematopoietic lineage cells from the fetus to the mother as manifest in twin pregnancy[16]. Multiparity could be another risk factor for the development of PCM [17] and again, this observation has not been confirmed from other authors. In fact more than 50% of the patients are at their first or second pregnancy [10]. Molecular markers of an inflammatory process are found in most of the patients. 90% of the patients show high levels of plasma C-reactive protein that correlated positively with LV end-diastolic and end-systolic dimensions and inversely with LV ejection fractions [6]. Clinical features of PPCM include symptoms of congestive heart failure and chest pain. Signs can include tachycardia, tachypnea, pulmonary rales, an enlarged Heart, and an S3 heart sound. Such signs and symptoms overlap with those of many other conditions, ranging from normal pregnancy to pulmonary emboli and upper respiratory infection.

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 [18]. Chest radiographs can show signs of pulmonary congestion, cardiac enlargement, and even pleural effusions in some cases. Echocardiograms usually show decreased contractility and LV enlargement without hypertrophy [19].

Overall prognosis of PCM is good in majority of the cases, although some patients may progress to irreversible heart failure. Progression of the condition requiring heart transplantation is described in 4% and death in 9% at a two years follow up . Other studies showed a much higher mortality rate such as 15% or 32% at 6 months[13]. Patients who eventually die tend to have worse NYHA functional class, LVEF and larger LV dimensions at diagnosis.There seems to be an initial high-risk period with 25-50% of the women dying within the first 3 months postpartum [20]. MRI could be a useful alternative to dobutamine

stress echocardiography in predicting outcome. Delayed gadolinium enhancement is more likely to be present in patients less responding to conventional therapy but could resolve over time in the recovering patients [21]. Management involves conventional therapy for heart failure with diuretics, ACE-inhibitors, beta-blockers and aldosterone antagonists. Angiotensin-receptor blockers should be added in case of ACE-inhibitors intolerance.

Anticoagulant therapy should be considered in view of the low left ventricular EF, which predisposes to thrombus formation, especially in the peripartum period when a hypercoagulable state exists. In patients not improving on conventional therapy or in patients with critical hemodynamic state with cardiogenic shock, hemodynamic support with pressors should be considered.

The duration of heart failure treatment is determined by the patient's heart performance at rest and with exertion. Patients with normal EF at rest and during dobutamine could taper off medical therapy in 6-12 months; patients with normal EF at rest and abnormal EF during dobutamine should be treated for longer period with ACE-inhibitors and beta-blockers [22]. Patients who continue to have a depressed ventricular function at rest have a poorer prognosis and should receive medical therapy indefinitely. In any case, it seems reasonable to continue at least for a year with ACE-inhibitors and beta-blockers even in case of complete recovery.

A subsequent pregnancy carries a high risk of relapse, significant decrease of left ventricular function and mortality. Mortality rate is described to be approximately 55% during subsequent pregnancy [23] even though it seems associated more with patients who entered the subsequent pregnancy with abnormal systolic function i.e. Without making a complete recovery [10]. Complete recovery from a relapse is very rare. There is no consensus regarding recommendations for future pregnancy after PCM but patients whose left ventricular size or function does not return to normal should be counseled strongly to avoid subsequent pregnancy[5].

CONCLUSION

PCM is a relatively rare disease, which can have devasting consequences and should be promptly identified and correctly treated. Early diagnosis is important and therefore women who develop symptoms of heart failure during pregnancy or shortly after should be investigated for this condition. Effective treatment reduces mortality rates and increases the chance of complete recovery of ventricular systolic function.

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