Challenges in management of peripartum cardiomyopathy with diuretic resistance, hypoalbuminemia and hypothyroidism

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ABSTRACT:

Background: Peri-partum cardiomyopathy is an idiopathic cardiomyopathy that presents with heart failure secondary to left ventricle systolic dysfunction towards the end of pregnancy or in the months after delivery, in the absence of any other cause of heart failure. Approximately 75% of cases are diagnosed within the first month peri-partum, and 45% present in the first week.

Case presentation: In this report we present a 30 years old female patient, Para 1 living 1, who was admitted one month ago with complaints of abdominal distension, lower limb swelling and difficulty in breathing that started worsening two weeks prior. 12-lead ECG showed sinus tachycardia (100beats/min), low voltage and non-specific T inversion and flattening. Echocardiography showed dilated left ventricle (5.8cm) and left atrium (4.4cm), global hypokinesia with ejection fraction of 36% by biplane. Grade 1 diastolic dysfunction, no thrombus seen and Mild pericardial effusion.

Conclusion: Peripartum cardiomyopathy is a form of dilated cardiomyopathy associated with considerable morbidity and mortality, thus it should not be underestimated. It should be dealt with accordingly and when suspected, one must establish the diagnosis rapidly.

Key words: peri-partum cardiomyopathy, heart failure, difficulty in breathing

I. INTRODUCTION

Peri-partum cardiomyopathy is an idiopathic cardiomyopathy that presents with heart failure secondary to left ventricular systolic dysfunction toward the end of pregnancy or in the months after delivery, in the absence of any other cause of heart failure according to Heart Failure Association of the European Society of Cardiology Working Group on PPCM 2010.1 PPCM is a diagnosis of exclusion. Although the left ventricle may not be dilated, the ejection fraction is nearly always reduced below 45%.1

In contrast to other definitions, the Heart Failure Association’s definition specifically excludes women who develop cardiomyopathy early in their pregnancy and explicitly notes that not all cases present with left ventricular dilation. Pre eclampsia, hypertensive disorders, and multiple gestations are associated with occurrence of this cardiomyopathy.2

The severity of symptoms in patients with PPCM can be classified by the New York Heart Association system as follows:

- Class I - Disease with no symptoms
- Class II - Mild symptoms/effect on function or symptoms only with extreme exertion
- Class III - Symptoms with minimal exertion
- Class IV - Symptoms at rest

II. CASE PRESENTATION

We present a case of 30 years old female patient, Para 1 living 1, who was admitted with complaints of abdominal distension, lower limb swelling and difficulty in breathing that started worsening two weeks prior. She had similar symptoms that started four months after last child was born. The abdominal distension started gradually, progressively increased in severity and accompanied by lower limbs swelling. Difficulty in breathing was of gradual onset, progressively increasing in severity, initially on exertion, later even at rest necessitating use of up to 2 pillows while sleeping. History of paroxysmal nocturnal dyspnea, reduced urine frequency, occasional nocturnal dry cough, no chest pain and no history of fever. This was her index admission. During her course of illness, she was admitted several times due to heart failure, and treated with medications that included lasix. She attended outpatient follow-up monthly but later on irregularly and not adherent on her medications due to poor affordability. She is not having hypertension, diabetes or pregnancy induced hypertension. She is a form 2 leaver, not married, has a small shop and her child is one year eight months old. There was no history of pregnancy induced hypertension. Her father is hypertensive. On day of admission, she was conscious but orthopneic, tachypneic with respiratory rate 28breaths/min SPO2 98%, warm extremities and severe pretibial edema.
Sinus heart rate of 107 beats/ min regular, of low volume and normal character; blood pressure of 100/67 mmHg, JVP raised, with non-heaving shifted apex to 6th intercostal space anterior axillary line, S1 S3 gallop and pansystolic murmur grade 3 radiating to axilla suggesting mitral regurgitation and murmur of tricuspid regurgitation; moderate ascites and fine bilateral bibasal crepitations. The working diagnosis was acute decompensated heart failure secondary to peripartum dilated cardiomyopathy NYHA class IV with acute kidney injury (pre-renal).

**Plan of management**

She was put in cardiac bed, and started on intravenous frusemide 80 mg start dose then after review for adequacy of diuresis, continued at 60 mg twice daily, tablets enalapril 5 mg twice daily, tablets isosorbide mononitrate 10 mg twice daily. Daily weight measurement and Urethral catheterization to monitor urine output. **Blood investigations were as following:** WBC (K/UL): 6.6, Hb level (g/dl): 9.4, Platelets (K/UL): 229, Albumin (g/L): 28, Na (mmol/l): 132, K (mmol/l): 3.9, Cl (mmol/l): 93, Creatinine (umol/l): 127 (raised), BUN (mmol/l): 7.7 eGFR 55mls/min. Chest radiograph showed cardiomegaly with upper venous diversion suggesting left ventricular failure; 12-lead ECG showed sinus tachycardia (100 beats/min), low voltage and non specific T inversion and flattening.

Echocardiography showed dilated left ventricle (5.8 cm) and left atrium (4.4 cm) global hypokinesia with ejection fraction of 36% by bpline. Grade 1 diastolic diastolic dysfunction, no thrombus. Mild pericardial effusion.

**Progress in ward**

She was attended and seen in the daily ward rounds, assessed for hemodynamic stability and medications were reviewed. She developed very low blood pressures with cold extremities accompanying severe heart failure and was put on infusion dobutamine starting at 5 ug/kg/min and nitroglycerin 15 ug/min. Blood investigations were repeated, serum albumin level and thyroid function test done. Abdominal ultrasound was also done. Albumin infusion was prescribed for hypoalbuminemia but was not available and not affordable. Ascitic tapping was also done due to tense ascites and respiratory embarrassment. Treatment charts were reviewed on daily basis. As per the medications prescribed, intravenous frusemide dose was increased upto 120 mg twice daily. Tablets thyroxine 100 ug daily was added for hypothyroidism and tablets metolazone 2.5 mg added for diuretic resistance. She was seen by the nutritionist on regular basis. High calorie high protein diet recommended. Soya, milk and eggs were recommended. The heart rate was mostly on higher side but later started lowering due to improvement in NYHA class and use of tablets carvedilol (Figure 1). The urine output initially ranged 200 – 700mls in 24 hours, later about 1200 to 2000mls in 24 hours. Weight reduced from 70 kg at admission to 56 kg at discharge (Figure 2). Repeated blood profile showed WBC (K/UL): 7.85, Hb level (g/dl): 10.16, Platelets (K/UL): 300, Albumin (g/L): 32, Creatinine (umol/l): 84.8, BUN (mmol/l): 5.4 eGFR 70mls/min. She was discharged to come to outpatient follow-up in two weeks, counselled on: medication adherence, fluid intake restrict to 1-1.5L/day, low salt, high protein foods, gradual ambulation/exercise, resumption of sexual activity, future pregnancy

**Figure 1:** Heart rate trend in hospital shows fluctuating rates.

**Figure 2:** There is a decrease in weight during course of stay in hospital as per chart below.
CASE DISCUSSION

Peripartum cardiomyopathy is a form of dilated cardiomyopathy associated with considerable morbidity and mortality. At least 7% of cases of peripartum cardiomyopathy may be part of spectrum of familial dilated cardiomyopathy. Peripartum cardiomyopathy occurs in women within last month of delivery and 5 months post-delivery. Recent data suggesting central role of unbalanced peripartum oxidative stress that triggers proteolytic cleavage of hormone prolactin into potent antiangiogenic, proapoptotic, and proinflammatory 16-kDa prolactin fragment and role of bromocriptine. Her echocardiography showed dilated left ventricle and left atrium with impaired ejection fraction; and global hypokinesia. Thinned-out walls also present as supported by Sliwa et al. In contrary to what was found in this patient, pre eclampsia, hypertensive disorders, and multiple gestations are associated with occurrence of this cardiomyopathy.

This patient presented with acute decompensated heart failure NYHA class IV with ascites and and renal dysfunction. Cardiorenal syndrome is a common problem in patients with advanced heart failure. Arterial underfilling with consequent neurohormonal activation, systemic and intrarenal vasoconstriction, and salt and water retention cause the main clinical features of cardiorenal syndrome which includes progressive decline in renal function, worsening renal function during treatment of heart failure and resistance to loop diuretics. Initially the patient was given intravenous dobutamine infusion in combination with nitroglycerin infusion due to shock. Dobutamine is a cardiac inotrope used in patients with heart failure when the mean arterial pressure is low. This patient was on dobutamine infusion for two weeks continuously. Spinarova et al reported that short term infusion of dobutamine has been recommended for low blood pressure. Though meta-analysis shows that dobutamine is not associated with improved mortality in patients with heart failure. Long-term use of either continuous or intermittent, intravenous parenteral positive inotropic agents, in the absence of specific indications or for reasons other than palliative care, is potentially harmful. Nitrovasodilator also improve short-term symptoms and are safe to administer but with no impact on mortality. The patient stayed in hospital more than a month with slow resolution of symptoms and signs, and poor weight loss. This may be partly be explained by unavailability of medications due to unaffordability by the patient relatives, diuretic resistance, coexisting hypoalbuminemia and hypothyroidism or inappropriate dosages as per recommended guidelines. Also the heart rate control was not adequate. The use of beta blockers in heart failure patients is recommended but, in this case, there was inappropriate introduction of carvedilol tablets when there was congestion and consequently not adequately uptitrated for better response as per recommendations. Upon further investigations, it was found that the serum albumin level was low (28g/L) and hypothyroidism was present. The hypoalbuminemia may interfere with the effectiveness of the frusemide due to poor delivery to site of action. According to Polat et al, up to 69.6% of patients with heart failure have hypoalbuminemia. Malnutrition and inflammation are factors causing hypoalbuminemia in patients with heart failure. This patient was started on high protein high calorie foods that included soya, eggs and milk that led to improvement in albumin levels from 28 to 32g/L. Albumin infusion was prescribed but very expensive to buy. Hypothyroidism causes a hypodynamic cardiovascular state, which is associated with reduced left ventricular systolic and diastolic function according to Fraczek et al. Thus she was started on tablets thyroxine with aim of correcting hypothyroidism, improving cardiac inotropy but the dose given was higher than the recommended. According to Curotto et al, patients with chronic heart failure and hypothyroidism significantly improve their physical performance when normal TSH levels were reached. Metolazone tablets were added together with increased dose of intravenous diuretics due to persistent edema as loop diuretic resistance was suspected. This will cause sequential nephron blockade. Other measures include fluid and salt restriction, increasing diuretic dose or giving diuretic continuous infusion. These measures led to improvement in NYHA class, increased albumin level, increased eGFRand weight reduction due to adequate diuresis.

References


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