Heart Failure due to Peripartum Cardiomyopathy Presenting in the First Week of Puerperium - A Case Series from Nepal

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Abstract: Peripartum cardiomyopathy (PPCM) is a rare cause of heart failure associated with pregnancy without any other known cause. Most of the clinical presentation is similar to symptoms of advanced pregnancy making the diagnosis difficult. Reported are three patients who developed dyspnea, orthopnea, and dry cough during the first week of puerperium. On examination, bilateral lower limb edema and bilateral basal lung crepitation were present in all patients. Chest radiograph showed pulmonary edema in cases two and three, and pleural effusion in case one. All patients had reduced left ventricular ejection fraction and raised N-terminal pro-b-type natriuretic peptide (NT-proBNP) levels. Case two developed PPCM in the background of left pyelonephritis. Case three was complicated by acute kidney injury. All patients were managed with bromocriptine, diuretics, beta-blockers, ACE inhibitors, and fluid restriction. Hence, PPCM though rare should be considered as a differential in women presenting with features of heart failure in later months of pregnancy or within five months of delivery.

Keywords: heart failure, bromocriptine, peripartum dilated cardiomyopathy, pregnancy, peripartum cardiomyopathy.

Introduction-

Peripartum cardiomyopathy (PPCM) is a rare cause of heart failure affecting women in the later months of pregnancy or within five months of delivery. PPCM often presents when peak volume load, reached just before delivery is greatly reduced after delivery.[1,2]PPCM affects women from all ethnicities globally with wide variation in incidence from 1:20,000 live births in Japan to 1:100 in Zaria, Nigeria.[3,4] With a prognosis that can vary from the complete recovery of left ventricular function to maternal mortality as well as recurrence with subsequent pregnancies, the study of PPCM has been done sparsely in Nepal.[2,5] This case series attempts to increase awareness of disease, diagnosis, and treatment in Nepal.

CASE PRESENTATION

Case 1

A 33-years lady, G_2A_1 at 38+6 weeks of gestation, with no significant medical history underwent emergency LSCS for non-progression of labor secondary to arrest of descent. On the 4^{th} day of puerperium, she de-

veloped dyspnea on exertion, orthopnea, bilateral lower limb edema, and dry cough. On examination, she had increased blood pressure (160/110 mmHg) and bilateral lower limb pitting edema with bilateral basal crepitation on chest auscultation. Chest radiograph showed bilateral minimal pleural effusion. Transthoracic echocardiography (TTE) showed severe left ventricular systolic dysfunction, dilated left atrium (LA), moderate to severe mitral regurgitation (MR), severe tricuspid regurgitation (TR), moderate pulmonary artery hypertension (PAH) and reduced ejection fraction i.e< 25%. ECG showed T wave inversion in leads V1-V4 and aVL (figure 1). Serum N-terminal pro-b-type natriuretic peptide (NT-proBNP) was 10897 pg/ml (Normal <300). With the diagnosis of PPCM, she was managed with fluid restriction, diuretics, beta blockers, angiotensin receptor blockers (ARB), bromocriptine, and antibiotics. Repeat TTE done on the 12^{th} day of puerperium showed EF of 30%. However, she had 3 episodes of syncopal attack on the 19^{th} puerperal day which was probably due to orthostatic hypotension. She was discharged on the 22^{nd} puerperal day with oral ARB and bromocriptine and with close follow-up with cardiology and obstetrics and gynecology clinic.

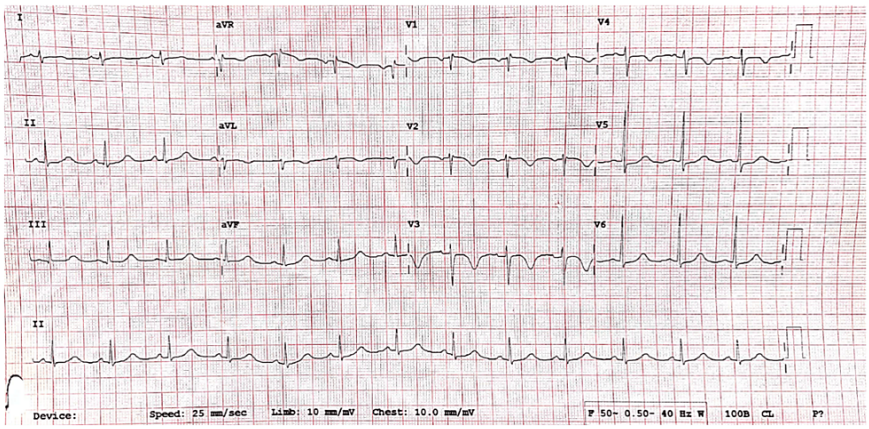

Figure 1: ECG of case 1 showing T wave inversion in leads V1-V4 and aVL.

Case 2

A 31 years lady, G_2 P_1L_1 at 40 +6 weeks of gestation gave birth to a male baby via vaginal delivery with first-degree perineal tear following induction of labor for post-dated pregnancy. On the 4^{th}day of puerperium, she was admitted for puerperal pyrexia with left pyelonephritis, moderate anemia, and hypokalemia. She was treated with antibiotics, potassium supplements, and two units of packed red blood cell (PRBC) transfusion. However, on the 6^{th} day of puerperium, she developed shortness of breath, chest pain, dry cough, and orthopnoea. On examination, she had bilateral lower limb pitting edema with vitals within normal limits. In the chest radiograph, there were infiltrates in the bilateral lower zone and blunting of bilateral costophrenic angles (figure 2). Serum NT-proBNP was 1678pg/ml. TTE showed: dilated left atrium/left ventricle, mild tricuspid regurgitation, moderate pulmonary artery hypertension, left ventricle systolic dysfunction with an ejection fraction of 40%, and severe mitral regurgitation. She was then managed with the diagnosis of PPCM with left pyelonephritis. She was kept in a propped-up position, daily BP charting and renal function test was done and was managed with fluid restriction, diuretics, beta blocker, angiotensinogen converting enzyme inhibitors (ACEIs), bromocriptine, and antibiotics. She was symptomatically better and was discharged on the 14th day of puerperium with empagliflozin, torsemide, carvedilol, and cefixime with advice to follow up in cardiology and obstetrics and gynecology clinic.

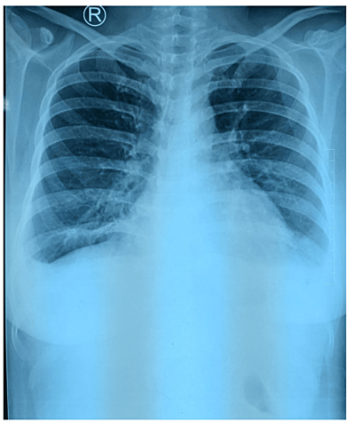

Figure 2: Chest X-ray of case 2 showing infiltrates in the bilateral lower zone and blunting of bilateral costophrenic angles

Case 3

A 28 years lady, $G_3P_1L_1A_1$at 37+1 weeks of gestation with impaired glucose tolerance, underwent Emergency LSCS for oligohydramnios (AFI 4.3cm). On the 3rd postoperative day, she developed sudden onset shortness of breath. On examination, she was tachypneic (40 breaths/minute) with low saturation of oxygen (SaO_2-80% in room air), raised blood pressure (140/100 mmHg), and had bilateral pedal edema. On chest auscultation, bilateral wheezes, and basal crepitations were heard, without any murmurs. ECG showed S1Q3T3 (right heart strain) pattern. Serum NT-proBNP was 7269 pg/ml. Renal function test (RFT)was deranged with urea 49 mg/dl, and creatinine 2.2 mg/dl with normal Na+/K+. TTE showed: dilated left atrium and left ventricle, global hypertrophy, left ventricular systolic dysfunction grade II with an ejection fraction of 30%, and mild mitral regurgitation. She was then managed under the diagnosis of PPCM with acute kidney injury: fluid and salt restriction, input/output charting, daily RFT, BP charting, propped-up position, diuretics, bromocriptine, and ACEIs. TTE repeated 9 days later showed mild hypokinesia of basolateral LV and LVEF of 40%. She was discharged on ACEIs and bromocriptine which was continued for 2 months. She was followed up for 6 months where she was symptomatically better and her RFTs were normal.

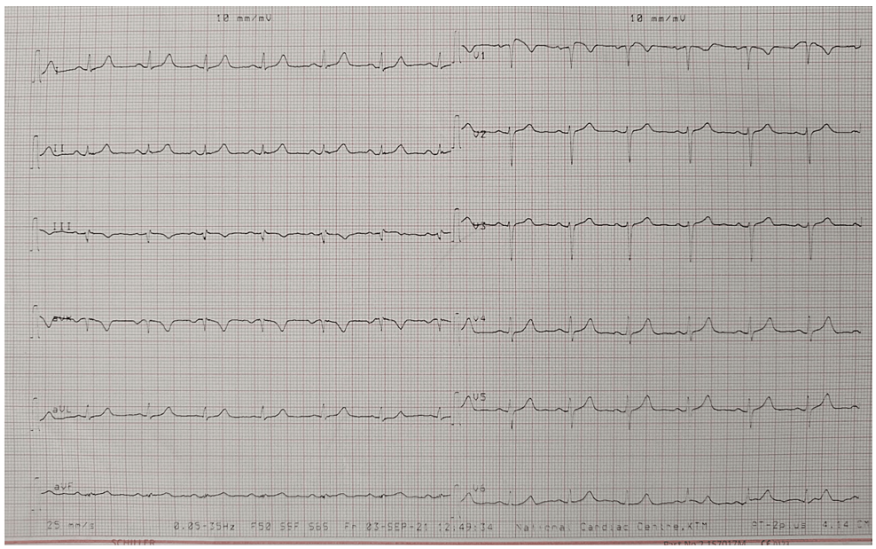

Figure 3: ECG of case 3 showing S1Q3T3 (right heart strain) pattern

Discussion:

Peripartum cardiomyopathy is a rare disease, often dilated cardiomyopathy of late pregnancy or early postpartum period without another known cause of heart failure.[5–7] PPCM has been defined as a heart failure that occurs in the last month of pregnancy or up to five months postpartum with left ventricular systolic dysfunction (left ventricular ejection fraction (LVEF) <45% or fractional shortening <30%, or both).[1]

Peripartum cardiomyopathy has been described in less than 0.1 percent of pregnancies with variable outcomes i.e. complete recovery or progression to severe cardiac failure and even sudden cardiac death.[8]

The etiology of PPCM is unclear, however, several risk factors have been identified so far. Among them, genetic predisposition, viral myocarditis, stress-activated cytokines, abnormal immune response to pregnancy, maladaptive response to hemodynamic stresses of pregnancy, excessive prolactin excretion, and prolonged tocolysis have been suggested as possible factors.[7,8] Although exact etiology could not be identified in our cases, the condition in case two might have been precipitated by pyelonephritis.

Since most of the clinical presentations are similar to symptoms of advanced pregnancy, diagnosis can be missed. Majority of patients present with typical features of heart failure such as dyspnea, orthopnea, cough and chest pain, hemoptysis, and paroxysmal nocturnal dyspnea.[5,9] Likewise, all of our three cases developed dyspnea, orthopnea, and dry cough during the first week of puerperium.

Physical examination usually reveals tachycardia, tachypnea, raised jugular venous pressure, displaced apical impulse, right ventricular heave, S3, and S4 gallop, murmurs of mitral and tricuspid regurgitation, rales, hepatomegaly and edema.[8,9] This was consistent with our cases who also had bilateral lower limb edema and bilateral basal lung crepitation.

PPCM can be investigated through several diagnostic modalities that include electrocardiography, chest radiography, echocardiography, and lab investigations such as BNP. ECG shows sinus rhythm, often with non-specific ST-segment or T-wave abnormalities. Chest radiography most often reveals indications of heart failure such as cardiomegaly, pulmonary congestion, and pleural effusions.[8] This finding was in keeping up with our cases as the Chest radiograph of cases two and three showed pulmonary edema, and that of case one showed pleural effusion. Patients with acute PPCM usually have elevated plasma concentrations of natriuretic peptides.[10] The diagnosis is confirmed by the echocardiographic findings of left ventricular systolic dysfunction.[1]

The differential diagnosis includes other causes of heart failure such as familial dilated cardiomyopathy, previous myocarditis, drug or toxin-induced cardiomyopathy, adult congenital heart disease, valvular disease, and pulmonary arterial hypertension among others.[6,7]

Acutely presented PPCM is similarly managed as acute heart failure of other etiologies.[10] Women in the peripartum period should be managed by a multidisciplinary approach including cardiologists, intensivists, obstetricians, neonatologists, anesthesiologists, and cardiac surgeons.[7,10]. Fluid and salt restriction is the mainstay of volume management, and loop diuretics may be added for symptomatic pulmonary or peripheral edema, taking care to avoid over-diuresis during pregnancy to prevent placental hypoperfusion.[8] Beta-blockers, hydralazine, and nitrates are also indicated for patients with PPCM. However, angiotensin-converting enzyme (ACE) inhibitors, angiotensin receptor blockers (ARBs), angiotensin receptor-neprilysin inhibitors (ARNI), ivabradine and mineralocorticoid receptor antagonists (MRAs) are contraindicated because of the possibility of teratogenicity.[7]

Important complications include severe heart failure, cardiogenic shock, arrhythmias, thromboembolic events, and death.[9]Moreover, there is an increased risk of PPCM in subsequent pregnancies with increased morbidity and mortality, particularly in women with persistent left ventricular systolic dysfunction after the first pregnancy.[11] In our experience, case three developed acute kidney injury during the disease.

Recovery typically occurs between 3 and 6 months postpartum, but there may be a delayed recovery as late as 48 months postpartum due to several factors such as delayed diagnosis, higher NYHA functional class, black ethnicity, LV thrombus, multiparity, and coexisting medical illnesses.[5]

To conclude PPCM despite being rare should be considered in women presenting with features of left ventricular failure in the later months of pregnancy or within five months of delivery. Acutely presenting PPCM that is managed as acute heart failure of other etiologies, can have devastating consequences if not diagnosed and treated early.

Author Contributions

S. Banmala, SA and LB contributed to the process of original draft preparation and introduction. PA, S. Basnet and BC led data collection, contributed in writing the case information and discussion. All the authors revised it critically for important intellectual content, contributed in review and editing.

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CONFLICT OF INTEREST

The authors report no conflicts of interest.

ETHICAL APPROVAL

This is a case report; therefore, it did not require ethical approval from ethics committee.

CONSENT

Written informed consent was obtained from the patient for publication of this case report and accompanying im-ages. A copy of the written consent is available for review by the editor- in- chief of this journal on request.

REGISTRATION OF RESEARCH STUDIES

Not applicable.

None

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