

CASE REPORTS

A Case Report On Peripartum Cardiomyopathy

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Abstract:

A 32 year-old primigravida with gestational diabetes & subclinical hypothyroidism on replacement therapy and no previous cardiac problem, developed features of shock few hours after elective caesarian section at term, in the absence of any chest pain or palpitation. Following resuscitation she developed features of acute left ventricular failure. ECG showed nonspecific T changes, chest x-ray revealed enlarged cardiac shadow with pulmonary congestion, arterial blood gas analysis was normal with supplemental oxygen. Serial cardiac markers were normal & serum d-Dimer was negative. Echocardiogram revealed dilatation of all cardiac

chambers with global hypokinesia & severe left ventricular (LV) systolic dysfunction. She was diagnosed as a case of peripartum cardiomyopathy and treated conservatively with medications. Her condition improved dramatically & she became symptom-free by the 5th post-operative day (POD) and subsequently discharged on 9th POD. Follow-up echocardiogram after 6 weeks revealed regional wall motion abnormality, normal chamber dimensions and fair LV systolic function.

Key words: cardiomyopathy, Pericardium

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Introduction:

Peripartum cardiomyopathy (PPCM) is a rare but potentially fatal disease which presents with symptoms of heart failure primarily due to left ventricular (LV) systolic dysfunction presenting in the last part (mean 32-38 weeks) of pregnancy and up to 5-6 months post delivery¹⁻⁵. It is clinically very similar to other forms of non-ischemic dilated cardiomyopathy except for its unique relationship with pregnancy¹⁻² and the higher likelihood for full recovery in almost half of the cases³. However it can still result in chronic disability and ultimately death in relatively young women in their reproductive years. This emphasizes the need for a thorough understanding of PPCM, so that the early diagnosis & institution of effective multi-disciplinary management can influence patient's long term prognosis.

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Case Report

Mrs. X, a 32 year-old primigravida from old town, Dhaka, with history of amenorrhoea for 37⁺² weeks, got admitted in BIRDEM Hospital for elective lower segment caesarian section (LSCS). Her pregnancy period was uneventful. She had regular antenatal check-up & was properly immunized as per schedule. She was diagnosed of having gestational diabetes mellitus (GDM) & subclinical hypothyroidism in the early part of pregnancy on the basis of OGTT & thyroid function tests. There was no past history of any major illness. She was getting levothyroxine, iron, folic acid and calcium supplements. Her blood sugar was well controlled with dietary & nutritional measures alone. She was a homemaker with sedentary life style & came from an upper middle-class family. Both her parents were alive & suffering from diabetes. Prior to conception her menstrual history was normal.

At the time of admission, she was mildly anemic with normal vital parameters. There was no edema or cyanosis. Bed side urine test was normal. Abdomen was distended due to gravid uterus with fundal height corresponding 35 weeks of gestation; fetal movement was present. Fetal heart sounds were audible. Other systemic examinations were unremarkable. Investigations done prior to operation revealed

hemoglobin 8.9 gm% with normal total and differential count of WBC. Both fasting & post-prandial blood glucose levels were normal as well as total protein, serum albumin levels and renal function tests. Serum TSH was within normal range & USG revealed single live fetus with gestational age corresponding to 34 weeks.

She underwent LSCS two days following admission under spinal anesthesia. Bleeding during LSCS was average. Three hours later, in the post-operative ward, she suddenly developed shock with cold clammy extremities, feeble pulse & non-recordable BP. There was no associated chest pain or palpitation. ECG revealed sinus tachycardia with nonspecific T changes. Abdomen was soft with contracted uterus, dressing was dry. Per vaginal bleeding was average.

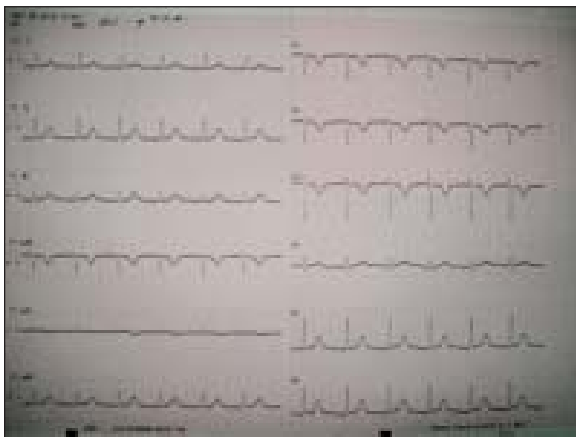


Fig-1:

She was resuscitated with oxygen, IV saline, noradrenaline, blood transfusion & shifted to ICU. In ICU, although blood pressure gradually became normal, she later on developed breathlessness along with tachypnea, diminished breath sounds in lower chest and bi-basal crepitations. She was diagnosed provisionally as a case of cardiogenic shock, acute left ventricular failure, status post (S/P) LSCS, GDM & subclinical hypothyroidism. A differential diagnosis of amniotic fluid embolism was kept in the mind.

Investigations in ICU revealed nonspecific T changes in follow-up ECG, normal arterial blood gas with oxygen saturation >90% with supplemental O₂ @ 4l/min via facemask. Serum D-dimer was negative. Serial serum troponins I levels and other cardiac markers were normal. Initial portable chest x-ray revealed pulmonary

congestion with probable enlarged cardiac shadow. Follow-up x-ray chest in upright posture revealed enlarged cardiac shadow with bilateral pulmonary congestion, more marked on the left.



Fig-2:



Fig-3:

Initial bed side 2-D, m-mode echocardiogram revealed global hypokinesia with dilatation of all 4 chambers & severe LV systolic dysfunction (EF-30%). Pulmonary trunk including the main branches was normal in dimension. Follow-up echocardiogram with color doppler study on the next day revealed global hypokinesia, dilated left ventricle, severe LV systolic dysfunction (EF-30%), grade ¼ PR, with trivial MR.

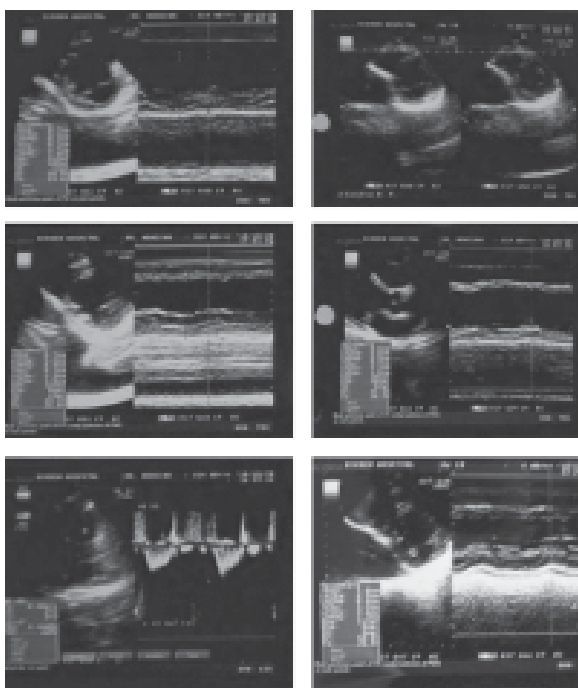


Fig.-4:

Her general conditions improved on conservative management with oxygen supplementation, frusemide (initially parenteral), digoxin, spironolactone, trimetazidine, fluid and salt restriction. The patient gradually improved and in the fifth post-operative day became symptom-free. She was finally diagnosed as a case of – Cardiogenic shock & acute left ventricular failure due to peripartum cardiomyopathy (PPCM), Status post LSCS, GDM & subclinical Hypothyroidism.

She was discharged on the 9th post-operative day with oral frusemide, spironolactone, losartan potassium, digoxin, calcium, iron, folic acid & vitamin supplements. She was properly counseled about contraception, time for future pregnancy & risk of recurrence of PPCM & was asked to come for follow-up after 6 weeks. Follow-up echocardiogram at that time revealed mild antero-septal wall hypokinesia with fair LV systolic function (EF-50%) and normal dimension of cardiac chambers.

Discussion

Peripartum cardiomyopathy (PPCM) is an infrequent but critical disorder in which a destabilized heart is diagnosed within the last months of pregnancy or early puerperium and often complicating obstetrics as well as anesthetic management. Usually, it occurs early in

the postpartum period, with about 45% in the first week and 75% within the first month¹. It is largely a diagnosis of exclusion. Other causes of heart disease must be ruled out first before making a diagnosis of PPCM. It is then diagnosed in previously healthy women presenting with symptoms of heart failure and evidence of decreased LV systolic function from late pregnancy to early puerperium.

A relationship between pregnancy and dilated cardiomyopathy was first noted in 1870 when Virchow and Porak first reported autopsy evidence of myocardial degeneration in patients who died in the puerperium⁶. In 1937 Gouley et al⁷ described the clinical and pathologic features of seven pregnant patients who had severe and often fatal heart failure. The incidence of PPCM was one case per 1374 live births in an Indian study⁸. PPCM occurs in 1 in 3,000 to 1 in 4,000 pregnancies in the United States⁹. In South Africa, the reported incidence is higher (1: 1,000 live births)¹⁰. A much higher incidence of 1:300 live births has been reported from Haiti¹¹ and an extremely high rate of 1% has been described in Nigeria¹². Higher rates in developing countries may be due to variations in local cultural as well as puerperal practices, ecological factors, environmental influence, diagnostic criteria and reporting pattern used¹³.

Risk factors favoring development and recurrence of PPCM include the following: 1) advanced maternal age (>30 years), 2) multiparity, 3) Afro-American race, 4) twin pregnancy, 5) pre-eclampsia, 6) gestational hypertension, 7) chronic hypertension, 8) obesity, 9) prolonged use of tocolytics (â₂ stimulants e.g. terbutaline). PPCM is still regarded as a disease of unknown etiology. However there are several hypotheses like selenium deficiency, inflammatory pathology leading to myocarditis (viral infection or auto-immune response to released fetal antigen). Recent evidence in an animal (mice) model suggests a role for a 16 kDa prolactin derivative produced by proteolytic cleavage of prolactin secondary to unbalanced oxidative stress present during late pregnancy and early puerperium¹⁴⁻¹⁵. In certain cultures where the incidence of PPCM is high, certain cultural practices performed during the puerperium such as consuming lake salt or rock salt (known as ‘kanwa’ which has a particularly high sodium content) to promote the flow of breast milk and the heating of the body by sitting on a clay bed with a fire

beneath to keep warm (a belief felt to ward off infection) have both been suggested as contributory factors in its development as well¹⁶⁻¹⁷.

Presentation of PPCM is similar to that of patients presenting with Left ventricular failure due to other causes. The clinical presentation is most often dyspnea (90%), tachycardia (62%), and edema (60%)¹⁸. Some case studies also cite unusual presentations, including multiple thromboembolic events¹⁹ and acute hypoxia²⁰. The classical symptoms of heart failure can be masked - especially in obese women. Onset occurs one month prior to delivery and up to five months after delivery. However, the majority of women present postpartum. Possible complications include thromboembolism, arrhythmias, organ failure, obstetric & perinatal complications (premature delivery, small for date and low birth weight babies, intrauterine growth retardation and fetal deaths). PPCM is largely a diagnosis of exclusion. Other causes of heart disease such as congenital heart disease or acquired conditions that is, myocardial infarction causing LV dysfunction, pulmonary hypertension or valvular heart disease must be ruled out first before making a diagnosis of PPCM. The diagnosis of PPCM poses many challenges, as many women in the last month of normal pregnancy experience similar symptoms to that of early heart failure, such as shortness of breath on exertion, nocturnal dyspnea and cough, fatigue, palpitations and pedal edema, making differentiation difficult.

Original diagnostic criteria for PPCM were developed by Demakis et al in 1971⁵. They did not include echocardiographic findings because echocardiography was not readily available at that time. In 1999, echocardiographic criteria²¹ were incorporated in a new definition. Diagnostic criteria for PPCM include:

- Onset of heart failure in last month of pregnancy to 5-6 months post partum.
- Without any other demonstrable cause of heart failure.
- Absence of any heart disease before pregnancy.
- Echocardiography criteria to include
 - o An ejection fraction <45%, fractional shortening <30% or both, and
 - o End-diastolic dimension (LVIDd) >2.7 cm/m² body surface area.

The most common and confusing differential diagnosis of PPCM is Idiopathic Dilated Cardiomyopathy (IDCM). Though PPCM is identical to IDCM in several ways, most researchers now accept PPCM as a distinct entity for the following reasons²²⁻²³:

- PPCM occurs at a younger age and is generally associated with better prognosis.
- The incidence of PPCM is higher than IDCM.
- PPCM occurs mostly postpartum (78 - 93%), whereas IDCM usually manifests by the second trimester.
- PPCM exclusively affects pregnant women and recurrent PPCM is seen to manifest again in the peripartum period.
- Varying types of hemodynamic patterns are seen in PPCM compared to IDCM.
- Unique sets of antigen and antibodies against myocardium are seen in PPCM compared to IDCM patients.
- The incidence of myocarditis is higher in PPCM than in IDCM.
- Heart size returns to normal after delivery in a greater percentage of PPCM patients compared to IDCM.
- Contrary to IDCM, PPCM may lead to rapid worsening of clinical condition.

All patients should have routine blood tests to exclude anemia, electrolyte disturbance, and kidney, liver and thyroid dysfunction, inflammatory markers, a septic screen and viral serology. Cardiac markers such as troponins and CK-MB are not helpful alone in reaching a diagnosis. Radiological signs of heart failure such as cardiomegaly, pulmonary congestion and pleural effusion may be found on chest x-ray. ECG may show sinus tachycardia or other arrhythmias such as atrial fibrillation, atrial flutter, ventricular tachycardia, intraventricular block pattern, nonspecific ST-T changes and LV hypertrophy pattern. Moreover, the ECG may demonstrate no significant changes²⁴. Echocardiography confirms ventricular failure with increased left ventricular end-diastolic dimensions and decreased ejection fraction (LVEF) and differentiates PPCM from other causes of heart failure, such as valve disease, etc. Invasive evaluation, such as cardiac

catheterization or endomyocardial biopsy, is often unnecessary for diagnosis or treatment. Coronary angiography is not routinely indicated, as coronary arteries are usually normal in PPCM. The role of endomyocardial biopsy in the diagnosis of PPCM is controversial. It is not routinely recommended, because of its limited availability, higher complication rate & low specificity. The pathology identified on endomyocardial biopsy is often nonspecific.

The treatment for peripartum cardiomyopathy is similar to that for other nonischemic dilated cardiomyopathies; however, consideration must also be given for the fetus. Non-pharmacological therapy includes low sodium diet (<4 gm/day), fluid restriction (<2 L/day) and modest daily exercise (i.e., walking).

The cornerstone of optimal oral pharmacologic therapy for cardiomyopathy begins with afterload reduction with use of angiotensin converting enzyme inhibitors (ACEI) or angiotensin II receptor blockers (ARB). Unfortunately, pregnancy is a contraindication to the use of ACEI and likely the ARB as well. In this circumstance, the combination of hydralazine and nitroglycerin or amlodipine can be safely used in pregnancy to provide needed afterload reduction. Preload reduction can be accomplished with diuretics and low-dose oral nitrates. In pregnancy diuretics must be used with caution as to avoid dehydration. Oral inotropic therapy is provided by digoxin. Furthermore, the deleterious effects of excessive sympathetic nervous system activation may be blocked and reversed with low-dose β -blockers. In the treatment of acutely ill or highly symptomatic patients, intravenous preload and afterload reducing agents (nitroprusside, nitroglycerin) or inotropic agents (dobutamine, dopamine, milrinone) should be considered. Intravenous nitroglycerin, dobutamine, dopamine, and milrinone can be used in pregnant patients if medically necessary. Invasive hemodynamic monitoring is often used to guide the acute phase of this therapy. As there is a high incidence in thromboembolism in this population, anticoagulation with either heparin or warfarin should be strongly considered. However warfarin should be avoided during pregnancy as it can cause birth defects.

Cardiac transplantation offers a final yet very viable alternative for patients with peripartum cardiomyopathy who do not improve or who continue to deteriorate with

medical management. Due to limited availability of donor hearts, it may become necessary to support the patient with an intra aortic balloon pump (IABP) or ventricular assist device (VAD) as a bridge to transplant.

No specific treatment has been identified to significantly alter the morbidity of PPCM. Small trials have reported benefits of pentoxifylline, intravenous immunoglobulin, and bromocriptine. Pentoxifylline decreased TNF α levels and increased EF in patients with PPCM²⁵. Intravenous immunoglobulin (IVIG) improved EF compared to standard treatment in one small retrospective study of 6 cases compared to 11 controls²⁶. Case reports of recovery from PPCM with bromocriptine treatment have been described²⁷⁻²⁹.

There are several possible outcomes in PPCM. Some women remain stable for long periods, while others get worse slowly. Others get worse very quickly and may be candidates for a heart transplant. The death rate may be as high as 25-50%. The prognosis is poor in patients with persistent cardiomyopathy. Subsequent pregnancies are often associated with recurrence of left ventricular systolic dysfunction. The risk of recurrence in women who have completely recovered LV function after their previous pregnancy is lower than previously believed. Ideally, every woman planning a future pregnancy should have echocardiography performed, and, even if it is normal, they ought to have dobutamine stress echocardiography as well. Women with a full recovery of LV function on both echocardiography and dobutamine stress test can be advised that the risks of major complications are relatively low. Women with persistent LV dysfunction are advised not to pursue further pregnancies.

Conclusion

PPCM is an uncommon but potentially life threatening cardiac failure of undetermined etiology taking place in late pregnancy or early on in puerperium. Thromboembolism and cardiac arrhythmia are common complications. Diagnosis of PPCM should essentially include echocardiographic substantiation of left ventricular dysfunction. Treatment is generally the same as for heart failure with left ventricular systolic dysfunction with some possible exceptions because of the risks of certain drugs to the unborn child. In resistant cases, management with immunosuppressive drugs, immunoglobulin and pentoxifylline can be thought of.

For those who do not improve with conventional medical therapy, have persistent cardiomegaly, or have moderate to severe mitral regurgitation, referral to a cardiac transplant center should be considered. In the presence of persistent heart failure, further pregnancy is not recommended. If inevitable, subsequent pregnancy in patients with enhanced cardiac function should be managed in a multidisciplinary unit. Prognosis is linked to recovery of ventricular dysfunction.

References

1. Elkayam U, Akhter MW, Singh H. Pregnancy-associated cardiomyopathy: clinical characteristics and a comparison between early and late presentation. *Circulation* 2005;111:2050-55.
2. Cunningham FG, Pritchard JA, Hankins GD. Peripartum heart failure: idiopathic cardiomyopathy or compounding cardiovascular events? *Obstet Gynecol* 1986;67:157-68.
3. Demakis JG, Rahimtoola SH, Sutton GC. Natural course of peripartum cardiomyopathy. *Circulation*. 1971;44:1053-61.
4. Demakis JG, Rahimtoola SH. Peripartum cardiomyopathy. *Circulation* 1971;44:964-8.
5. Homans DC. Peripartum cardiomyopathy. *N Engl J Med* 1985;312:1432-37.
6. Sakakibara S, Sekiguchi M, Konno S. Idiopathic postpartum cardiomyopathy, report of a case with special references to its ultra-structural changes in the myocardium as studied by endomyocardial biopsy. *Heart* 1970;80:385-95.
7. Gouley B, McMillan T, Bellet S. Idiopathic myocardial degeneration associated with pregnancy and especially the puerperium. *Am J Med Sci* 1937;19:185-99.
8. Pandit V, Shetty S, Kumar A. Incidence and outcome of peripartum cardiomyopathy from a tertiary hospital in South India. *Trop Doct* 2009;39(3):168-69.
9. May HV. Peripartum cardiomyopathy: clinical and therapeutic characteristics. *J La State Med Soc* 1991;143(5):45-48.
10. Desai D, Moodley J, Naidoo D. Peripartum cardiomyopathy: experiences at King Edward VIII Hospital, Durban, South Africa and a review of the literature. *Trop Doct* 1995;25:118-23.
11. Fett JD, Christie LG, Carraway RD, et al. Five-year prospective study of the incidence and prognosis of peripartum cardiomyopathy at a single institution. *Mayo Clin Proc* 2005;80:1602-06.
12. Sanderson JE, Adesanya CO, Anjorin F. Postpartum cardiac failure- heart failure due to volume overload? *Am Heart J* 1979;97:613-21.
13. Veille JC, Zaccaro D. Peripartum cardiomyopathy: summary of an international survey on peripartum cardiomyopathy. *Am J Obstet Gynecol* 1999;181(2):315-19.
14. Reuwer AQ, Reuwer PJ, van der Post JA. Prolactin fragmentation by trophoblastic matrix metalloproteinase as a possible contributor to peripartum cardiomyopathy and pre-eclampsia. *Med Hypotheses* 2009 Sep 10. [Epub ahead of print].
15. Hilfiker-Kleiner D, Silva K, Drexler H. Peripartum cardiomyopathy: Recent insight into its pathophysiology. *Trends Cardiovasc Med* 2008;18:173-79.
16. Trevitt L. Attitudes and customs in child birth among Hausa women in Zaria City. *Savanna* 1973;2:223-26.
17. Davidson NM, Trevitt L, Parry EH. Peripartum cardiac failure an explanation for the observed geographic distribution in Nigeria. *Bull World Health Organ* 1974;51:203-08.
18. Chapa JB, Heiberger HB, Weinert L. Prognostic value of echocardiography in peripartum cardiomyopathy. *Obstet Gynecol* 2005;105(6):1303-08.
19. Carlson KM, Browning JE, Eggleston MK. eripartum cardiomyopathy presenting as lower extremity arterial thromboembolism. A case report. *J Rep Med Obstet Gynecol* 2000;45(4):351-53.
20. Cole WC, Mehta JB, Roy TM. "Peripartum cardiomyopathy: echocardiogram to predict prognosis," *Ten Med* 2001;94(4):135-38.
21. Hibbard JU, Lindheimer M, Lang RM. A modified definition for peripartum cardiomyopathy and prognosis based on echocardiography. *Obstet Gynecol* 1999;94:311-16.
22. Pearson GD, Veille JC, Rahimtoola S. Peripartum cardiomyopathy: National Heart, Lung and Blood Institute and Office of rare Diseases. (National Institute of Heart). Workshop recommendations and review. *JAMA* 2000;283:1183-88.
23. Avila WS, de Carvalho MEC, Tschaen CK. Pregnancy and peripartum cardiomyopathy. A comparative and prospective study. *Arq Bras Cardiol* 2002;79:484-93.
24. Witlin AG, Mable WC, Sibai BM. Peripartum cardiomyopathy: an ominous diagnosis. *Am J Obstet Gynaecol* 1997;176:182-88.
25. Sliwa K, Woodiwiss A, Candy G. Effects of pentoxifylline on cytokine profiles and left ventricular performance in patients with decompensated congestive heart failure secondary to idiopathic dilated cardiomyopathy. *Am J Cardiol* 2002;90(10):1118-22.
26. Bozkurt B, Villaneuva FS, Holubkov R. Intravenous immune globulin in the therapy of peripartum cardiomyopathy. *J Am Coll Cardiol* 1999;34(1):177-80.
27. De Jong JS, Rietveld K, Van Lochem LT. Rapid left ventricular recovery after carbergoline treatment in a patient with peripartum cardiomyopathy. *Eur J Heart Fail* 2009;11:220-22.
28. Habedank D, Kuhlenle Y, Elgeti T. Recovery from peripartum cardiomyopathy after treatment with bromocriptine. *Eur J Heart Fail* 2008;10:1149-51.
29. Hilfiker-Kleiner D, Meyer GP, Schieffer E. Recovery from postpartum cardiomyopathy in 2 patients by blocking prolactin release with bromocriptine. *J Am Coll Cardiol* 2007; 50:2354.