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Case Report: Peripartum Cardiomyopathy with Thromboembolic Episodes

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INTRODUCTION

Peripartum Cardiomyopathy is a type of dilated cardiomyopathy, of unknown origin, presenting as heart failure due to left ventricular systolic dysfunction in late 3rd trimester of pregnancy or within 5 months post delivery when no other cause of heart failure is detectable. Woman with Peripartum Cardiomyopathy commonly presents with complains of breathlessness, chest pain, palpitations, pedal odema near term or after delivery.

CASE REPORT

We report a rare case of Peripartum Cardiomyopathy with multiple thromboembolic episodes during postpartum period.

A 28 year old Russian lady was registered at Dr L. H Hiranandani Hospital in first trimester for Antenatal care and delivery. She was married to an Indian for 8years with obstetric history of Gravida 3 Para 1 Living 1 Abortion1 with 38 weeks of gestation. There was no evidence of Gestational Diabetes; Pregnancy induced hypertension or excessive weight gain in her antenatal period. There was no history of addiction to alcohol, cocaine or any other drug abuse. There no family history of hypertension, diabetes mellitus or any cardiac disease. She had a fullterm Cesarean section at Thailand due to intrapartum fetal bradycardia, 4yrs ago. In first delivery, there was no history of palpitations or breathlessness suggestive of cardiac disease during antenatal or postpartum period.

The patient was admitted for Elective Cesarean section at 39 weeks of gestation in view of previous cesarean section with poor Bishops score on 28/04/2014. Her pulse- 88/min and BP- 120/70 mm Hg, cardiovascular system (CVS) and respiratory system were normal, per abdomen- full term pregnancy, per vaginum- cervix posterior, os closed. Blood investigations advised were – Hemoglobin- 10.1gm%, TSH- 2.9mIU/ml, HbA1c – 5.2%, HIV, HbsAg, HCV- negative.

During cesarean section, uterine scar dehiscence was noted. There was no meconium stained amniotic fluid and the baby cried immediately after birth. She delivered a healthy female child, 3.29 kg. The uterine incision was closed in 2 layers with delayed absorbable suture No.1 Polyglactin. The patient's vital were stable during post-operative period and she was discharged after 3 days. After 1 day of discharge, the patient came to emergency room with complains of breathlessness and chest pain. O/E- pulse- 105/min, BP- 120/90 mm Hg, Respiratory system- basal crepts heard, CVS- S1S2 normal, no murmur, TROP- I- normal, TSH- normal. The ECG showed tachycardia, T wave inversion inV3-6. X-Ray chest showed right sided consolidation with pulmonary odema. 2 D Echo showed Dilated Cardiomyopathy with left ventricular ejection fraction (LVEF) - 20%. Patient was admitted in ICU and treated with intra venous (IV) beta-blockers, ACE inhibitors, Diuretics, Anticoagulants and Antibiotics. The patient was discharged after 5 days and advised Tablet Pradaxa (Dabigatran- direct Thrombin inhibitor), Tab Ramipril- (ACE inibitor), spironolactone + torasemide (diuretic) and Metoprolol (beta blocker).

The patient reported to emergency room after 12 hrs of discharge with complaints of sudden onset left sided weakness and numbness of upper and lower limbs, which had recovered in 10 mins.

MRI Brain showed no abnormality and Transient Ischemic attack was suspected. 2 D Echo done on Day 10 of delivery reported LVEF 30% with marked Global Hypokinesia and a left ventricular blood clot measuring 0.9 cm. The patient was started on Low Molecular Weight Heparin continuous infusion to prevent further thrombo-embolic episodes.

On day 11 of CS, the woman complained of acute right side pain in loin and iliac fossa. CT Scan of abdomen showed Infarction of Right Kidney cortex with hydroureter and uterine hematometra (13.9x8.2 cm). After withholding anticoagulant for 24 hrs the patient was posted for surgery- ultrasound guided evacuation of hematometra and D-J stenting of the right ureter. The patient was discharged after 8 days on oral anticoagulant and ACE inhibitors for 1 month. After 1 month, her 2 D Echo showed marked improvement in LVEF (40%), mild global hypokinesia and no left ventricular blood clot. The

patient is asymptomatic after 6 months of delivery and her LVEF had recovered to 55%.

We reported this rare case of peripartum cardiomyopathy presenting after 3 days of cesarean section with recurrent episodes of thromboembolism and cardiac recovery within 6 months of delivery.

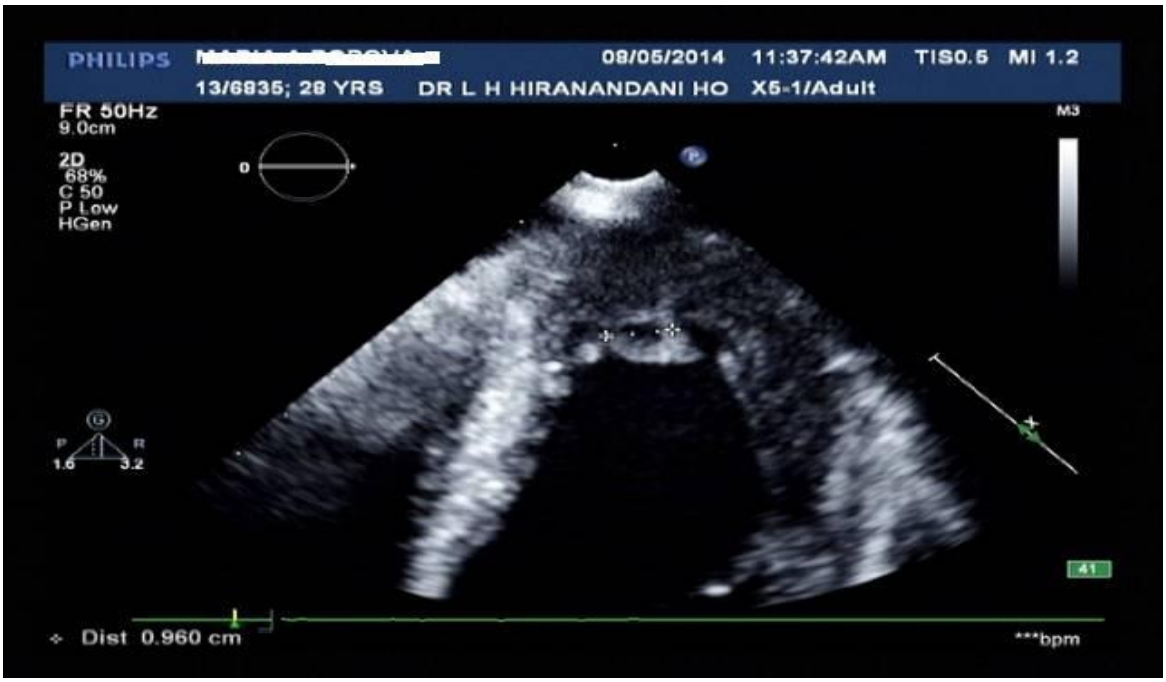


Fig 1: dilated left ventricle with apical blood clot

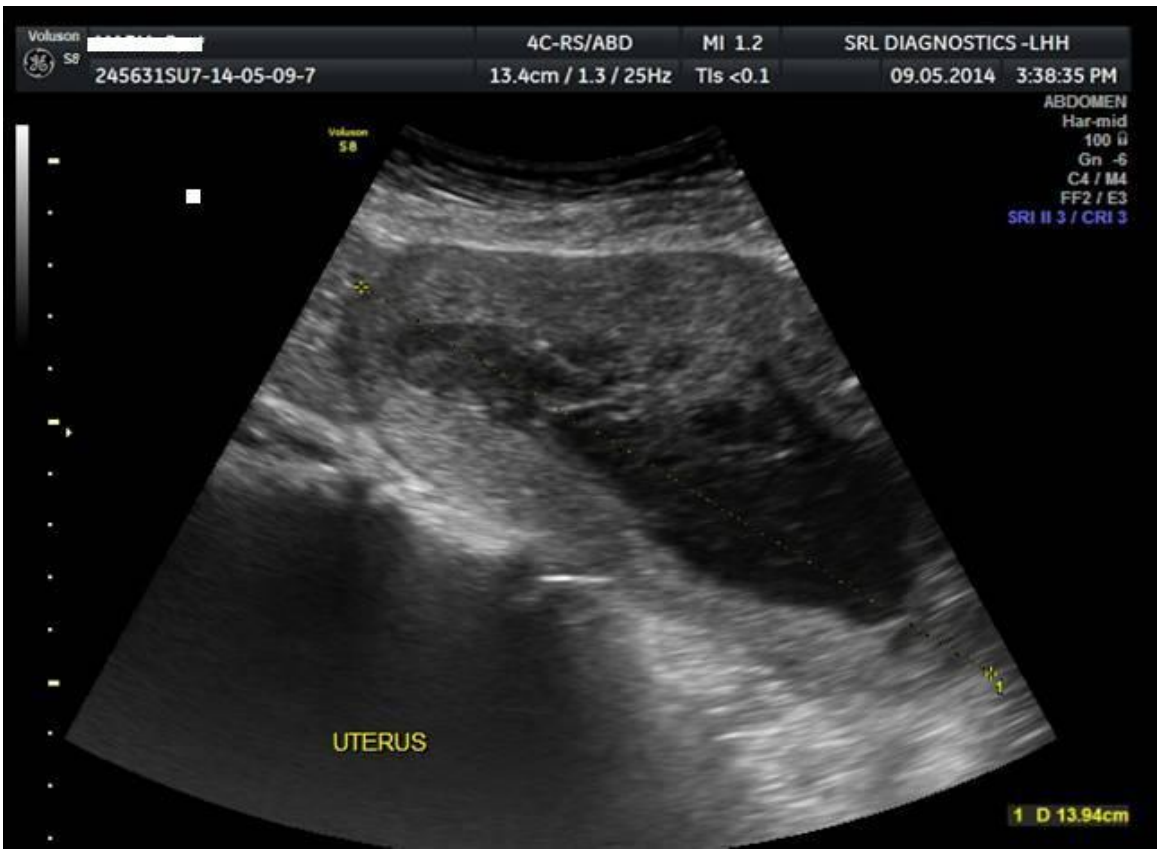
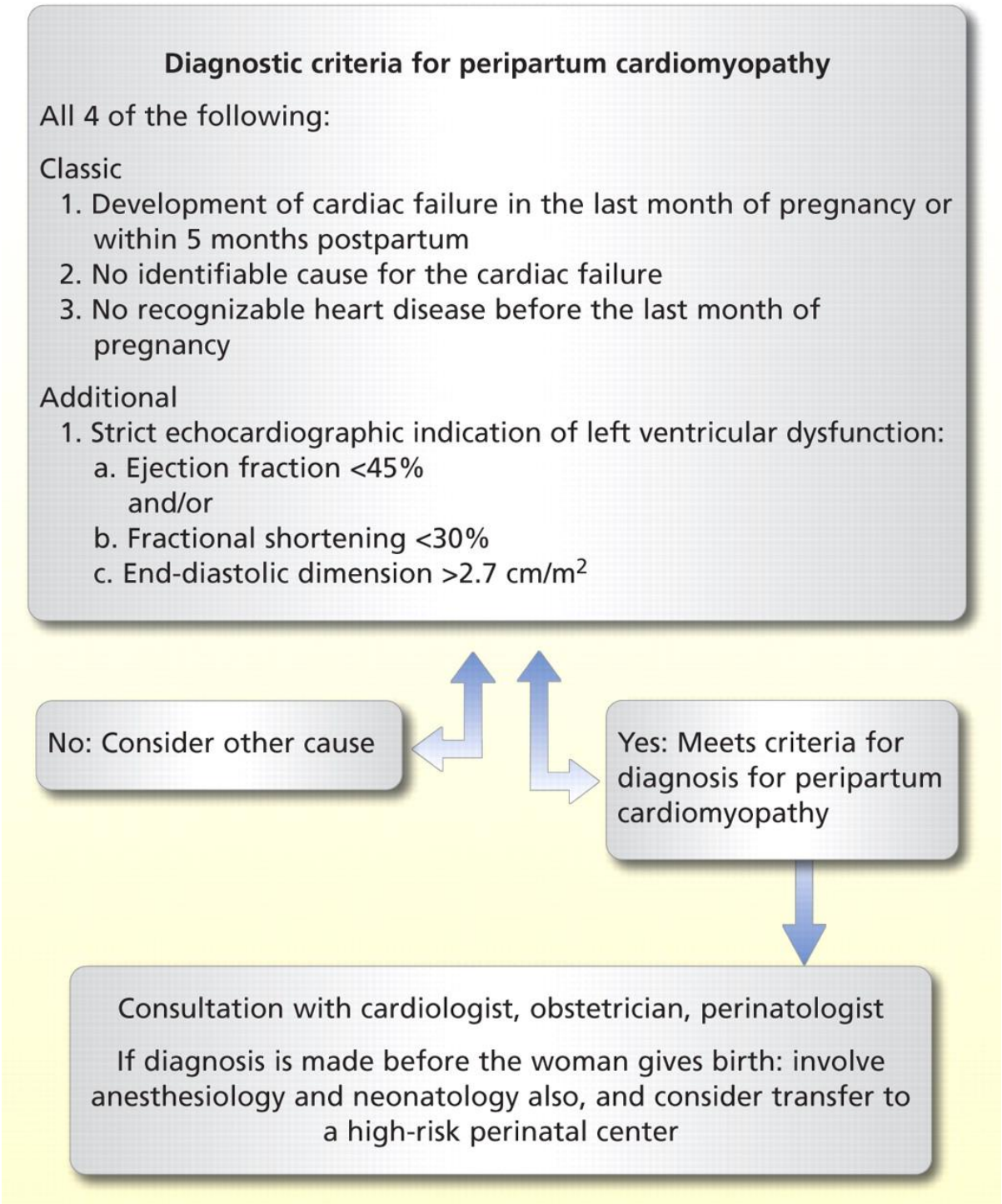
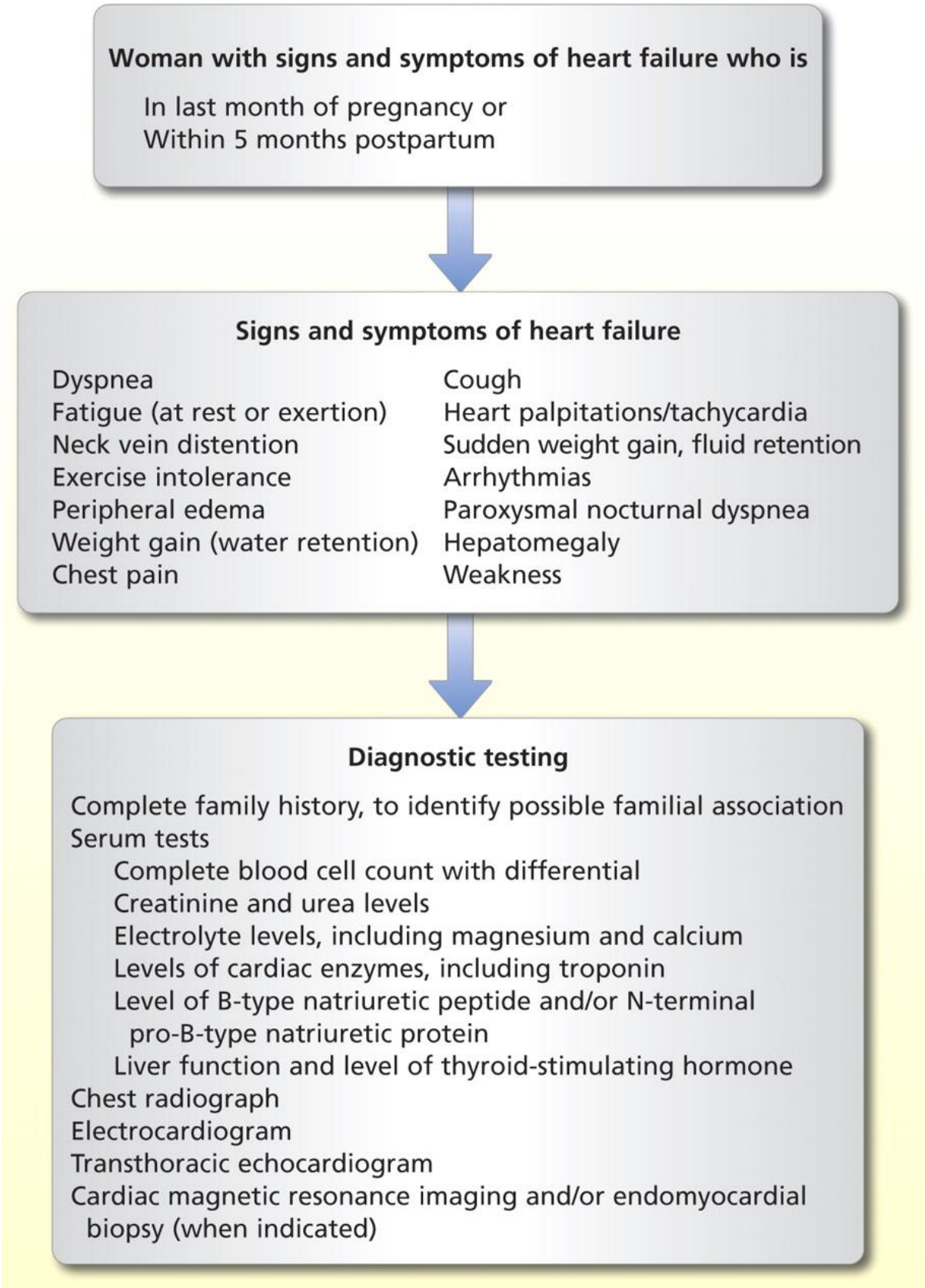


Fig 2: uterine hematometra


Diagnostic criteria for peripartum cardiomyopathy
All 4 of the following:
Classic
1. Development of cardiac failure in the last month of pregnancy or within 5 months postpartum
2. No identifiable cause for the cardiac failure
3. No recognizable heart disease before the last month of pregnancy
Additional
1. Strict echocardiographic indication of left ventricular dysfunction:
a. Ejection fraction <45%
and/or
b. Fractional shortening <30%
c. End-diastolic dimension >2.7 cm/m2
No: Consider other cause
Yes: Meets criteria for diagnosis for peripartum cardiomyopathy
Consultation with cardiologist, obstetrician, perinatologist
If diagnosis is made before the woman gives birth: involve anesthesiology and neonatology also, and consider transfer to a high-risk perinatal center


Woman with signs and symptoms of heart failure who is
In last month of pregnancy or
Within 5 months postpartum
Signs and symptoms of heart failure
Dyspnea
Fatigue (at rest or exertion)
Neck vein distention
Exercise intolerance
Peripheral edema
Weight gain (water retention)
Chest pain
Cough
Heart palpitations/tachycardia
Sudden weight gain, fluid retention
Arrhythmias
Paroxysmal nocturnal dyspnea
Hepatomegaly
Weakness
Diagnostic testing
Complete family history, to identify possible familial association
Serum tests
Complete blood cell count with differential
Creatinine and urea levels
Electrolyte levels, including magnesium and calcium
Levels of cardiac enzymes, including troponin
Level of B-type natriuretic peptide and/or N-terminal pro-B-type natriuretic protein
Liver function and level of thyroid-stimulating hormone
Chest radiograph
Electrocardiogram
Transthoracic echocardiogram
Cardiac magnetic resonance imaging and/or endomyocardial biopsy (when indicated)

DISCUSSION

Peripartum Cardiomyopathy (PPCM) is a form of non-familial, non-genetic, dilated cardiomyopathy, with deterioration in cardiac function typically presenting between last month of pregnancy and up to 5 months post delivery. [1]

In PPCM, there is systolic dysfunction and reduced left ventricular ejection fraction (LVEF). The patient usually presents with congestive cardiac failure and there is higher risk of atrial and ventricular arrythmias, thromboembolic episodes, and sometimes even sudden cardiac death. The incidence of PPCM in overall pregnancies is very low (0.1%); but the maternal mortality and morbidity can be between 5% to 32%. The final prognosis of PPCM cannot be predicted only on the initial assessment of LVEF. [2]

Mielniczuk LM et al [3] reported the incidence of PPCM in Haiti was 1 in 299 livebirths and the incidence in Southern California was 1 in 2229 live births. Brar et al [4] reported that the incidence of PPCM in African American was found to be 2.9 times more than the white Americans and 7 times more likely Hispanic women. The common predisposing factors for peripartum cardiomyopathy are multiparity, elderly woman, multifetal gestations, pregnancy induced hypertension and gestational diabetes mellitus. Ntusi NB et al [5] proposed many theories for etiological causes of PPCM like Viral myocarditis, abnormal maternal response to hemodynamic changes in pregnancy, response to inflammatory cytokines, tumor necrosis factor, C-reactive protein, Fas/Apo- 1; a plasma marker for apotosis. There also have been co-relation of high level of prolactin hormone during pregnancy and chronic use of β-sympathomimetic medications as tocolytic agents. Ansari et al [6] in 2002, studied the role of abnormal autoimmune response to fetal micro- chimerism i.e. the presence of fetal cells in maternal circulation as the cause of PPCM.

Clinical manifestations and diagnosis: The diagnosis of peripartum cardiomyopathy can be easily missed in a near term or post delivery women, as the symptoms like breathlessness, dyspnea, tachycardia, pedal odema, dizziness and fatigue are also common in pregnant women. These symptoms occur due to the physiological changes like increased blood volume, anemia, decreased peripheral vascular resistance and mild ventricular dilatation and increased cardiac output. The woman with PPCM mostly presents with chest pain, palpitations, dyspnea, arrythmias, jugular venous distention, cough, exercise intolerance, paroxysmal nocturnal dyspnea, hepatomegaly, thromboembolic episodes, and myocardial infarction.

Peripartum cardiomyopathy is a diagnosis of exclusion for heart failure. The most common causes of heart failure like cardiac infarction, pulmonary embolism, valvular heart disease, severe preeclampsia, sepsis and other cardiomyopathy should be investigated. Investigations for complete blood count, TSH, S creatinine, S. electrolytes and cardiac enzymes like Troponin are adivised. Troponin levels do help to rule out myocardial infarction, but it can be elevated in acute phase of PPCM.

Electrocardiograms (ECG) may show normal findings or may show evidence of left ventricular hypertrophy, ST-T wave abnormality, arrythmias and prolonged PR and QRS intervals. 2 D Echocardiography is the gold standard for diagnosis of cardiomyopathy, with findings of decreased ejection fraction, global dilatation, and thinned out cardiac walls. The American College of Cardiology Foundation and the American Heart Association[7] have given Endocardiographic criteria for PPCM like Ejection fraction < 45% and /or Fractional shortening < 30% and End- diastolic dimension > 2.7cm/m^2.

Cardiac magnetic resonance imaging and Endomyocardial Biopsy can be conducted to study the pathogenesis of the disease and the inflammatory processes. The treatment of PCCM is to treat the cardiac failure by reducing preload and afterload and improving cardiac inotropy. The standard protocol for heart failure includes the use of beta blockers (B-B), Diuretics, and (ACE-I) angiotensin converting enzyme inhibitors after delivery.

The aim is to reduce the blood pressure with use of diuretics and improve the cardiac function with Beta blokers and ACE-Inhibitors. In breastfeeding mothers, instead of ACE-I, Hydralazine with nitrates can be used. When the left ventricular ejection fraction is <35%, there is increased risk of left ventricular thrombus formation. Anticoagulation i.e Warfarin is advised to these women post delivery. Low molecular weight Heparin is advised in Antenatal period and continued after delivery till LVEF improves on 2 D Echo findings. Arrythmias should be promptly diagnosed and treated to reduce the risk of thrombus and improve cardiac function. Philips SD and Warnes CA[8] reported in refractive cases of PPCM with heart failure not responding to conventional treatment may need implantation of Left Ventricular Assist Device (LAVD) and sometimes Heart transplant is recommended as terminal treatment. When PPCM is diagnosed after fetal maturity i.e. after 37 weeks of gestation or with worsening heart functioning, termination of pregnancy may result in improving the maternal cardiac status. The delivery of woman with PPCM is to be referred to a High risk obstetric Unit which includes experienced Obstetrician, Cardiologist, Intensivist and Anesthesiologist. The patient with stable PPCM is recommended vaginal delivery to reduce the risk of associated to anesthesia and

surgery, to reduce blood loss, infections, post- op pain, and thromboembolic episodes.

An elective cesarean section has an advantage of rapid and safe delivery in the presence of experienced anesthesiologist and cardiologist.

In the United States, it has been observed at, atleast 50% of PPCM mother show complete cardiac recovery i.e LVEF> 55% within 1 year of delivery. It is advised to continue ACE inhibitors and beta – blockers till 1 year of post delivery. William et al [9] suggested that the subset of women with persistent left ventricular dysfunction should be counseled against future pregnancies. Also, the women with recovery of LVEF are at 19% higher risk of maternal death in subsequent pregnancy.

CONCLUSION

PPCM typically presents in healthy women in the 3rd trimester of gestation and up to 5 months post-delivery. The obstetrician should have high index of suspicion in pregnant women if she presents with breathlessness, palpitations, excessive weight gain due to fluid retention, and severe pre-eclampsia to diagnose peripartum cardiomyopathy.

Careful assessment of risk factors in pregnant women would help in early diagnosis and treatment of peripartum cardiomyopathy.

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