REVIEW ARTICLE

Peripartum cardiomyopathy (PPCM) – current review

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ABSTRACT

Peripartum cardiomyopathy, a rare life threatening type of dilated cardiomyopathy of unknown origin, occurs in previously healthy women causing a devastating form of cardiac failure affecting women mainly in their last months of pregnancy or early puerperium and often complicating their obstetrics as well as anesthetic management. Although the incidence is low—less than 0.1% of pregnancies - morbidity and mortality rates are high at 5% to 32%. The outcome of peripartum cardiomyopathy is also highly variable. For some women, the clinical and echocardiographic status improves and sometimes returns to normal, whereas for others, the disease progresses to severe cardiac failure and even sudden cardiac death. In acute care, treatment may involve the use of intravenous vasodilators, inotropic medications, an intra-aortic balloon pump, ventricular-assist devices, and/or extracorporeal membrane oxygenation. Survivors of peripartum cardiomyopathy often recover from left ventricular dysfunction; however, they may be at risk for recurrence of heart failure and death in subsequent pregnancies. Women with chronic left ventricular dysfunction should be managed according to guidelines of the American College of Cardiology Foundation and the American Heart Association.

Key words: Peripartum cardiomyopathy, PPCM-review
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Peripartum cardiomyopathy is a life threatening type of dilated cardiomyopathy. The cause of the same is of yet unknown origin. The condition occurs in the last months of pregnancy or early puerperium and complicates obstetric as well as anaesthetic management. Women with chronic left ventricular dysfunction should be managed according to guidelines of the American College of Cardiology Foundation and the American Heart Association. [1-5, 21].

Peripartum cardiomyopathy is defined based on four criteria by Demakis et al [6]:

Classic:

a. Development of cardiac failure in the last month of pregnancy or within 5 months of delivery.
b. Absence of identifiable cause for the cardiac failure.
c. Absence of recognizable heart disease prior to the last month of pregnancy

Additional:

Left ventricular systolic dysfunction demonstrated by classic echocardiographic criteria, such as decrease shortening fraction or ejection fraction.

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Etiology

The actual etiology of PPCM is unknown. Several hypotheses like myocarditis, viral infection, autoimmune factors, inflammatory cytokines, abnormal hemodynamic response to physiological changes in pregnancy, prolonged tocolysis and selenium deficiency have been postulated[5,7,8]. Reported incidences of myocarditis range between 29 to 100% [9, 10]. In a large study, Silwa et al [10] found higher concentrations of inflammatory cytokines like tumor necrosis factor α (TNFα), C-reactive protein (CRP), Interleukin-6 (IL-6) and Fas/Apo-1 (a marker of apoptosis) in PPCM patients, CRP levels correlated inversely with left ventricular ejection fraction (LVEF) in their study. High TNFα concentration may lead to advanced ventricular remodeling through specific cardiac receptors, resulting in ventricular dysfunction. Viral infection has also been implicated as a cause of myocarditis. In the 1970s, several reports in support of an autoimmune nature of PPCM joined reports of fetal chimerism (presence of fetal cells in maternal circulation during and after pregnancy). Such cells would be recognized as foreign antigens after normalization of maternal immunity following delivery and may trigger an immune response [8]. Abnormal hemodynamic response to physiological changes in pregnancy, PPCM may be an exacerbation of this normal phenomenon [8]. Abnormalities of relaxin, primarily an ovarian hormone produced during pregnancy, recently found in cardiac atria, shown to have positive inotropic and chronotropic properties and potentially involved in excessive relaxation of the cardiac skeleton [12].

Clinical Features

Symptoms of Dyspnea (90%), tachycardia (62%), and edema (60%) and fatigue [13]. These symptoms are often initially erroneously diagnosed as part of the normal puerperal process or pneumonia. Early diagnosis of PPCM may be difficult because many of the similarities of its presenting feature with that of advanced pregnancy. Diagnosis can be aided by Echocardiographic evidence of L V systolic dysfunction and significantly elevated serum brain natriuretic peptides (BNP). Elevations of troponin T (TnT) appear to have prognostic significance in this group. A TnT level 0.04 ng/mL at presentation predicts persistence of systolic dysfunction with a sensitivity of 55% and specificity of 91% [14].

Investigations

Every patient should have an electrocardiography (ECG), chest X-ray (CXR) and doppler echocardiograph for diagnosis [7].

***ECG*:** ECG usually shows sinus tachycardia, though there may be features of atrial flutter/fibrillation, left atrial and ventricular

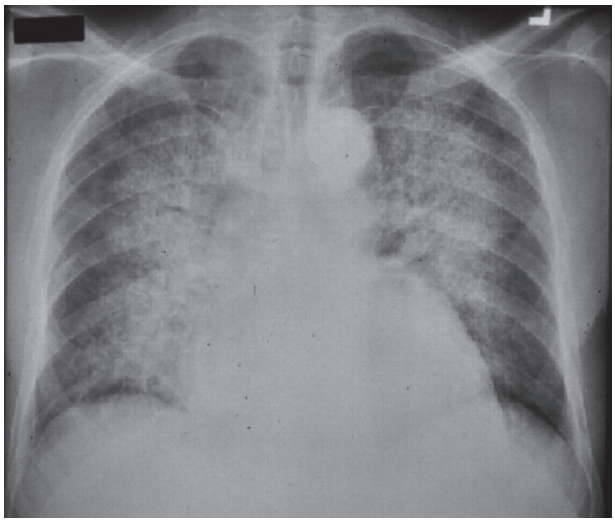

Fig 1. Cardiogenic pulmonary oedema with butterfly distribution due to LVF with perihilar opacity

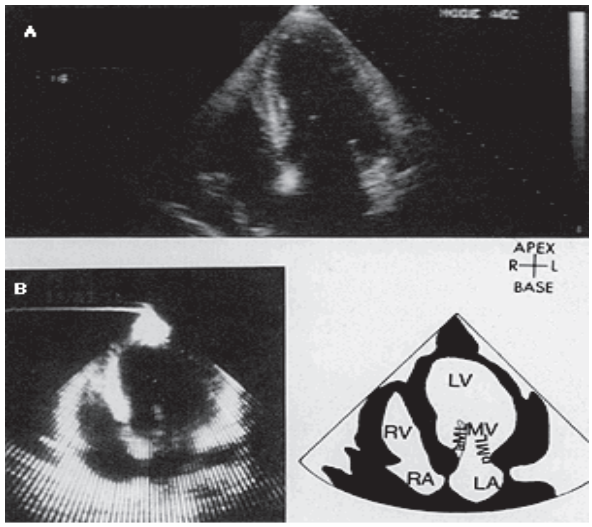

Fig 2. Heart is fairly spherical in Cardiomyopathy

hypertrophy (LVH), left axis deviation, nonspecific ST-T abnormalities, low voltage complex, arrhythmia, Q wave in anteroseptal leads and conduction abnormalities like prolonged PR, QRS intervals and bundle branch blocks [1-5, 15].

***Chest X-Ray*:** On CXR there may be evidence of cardiomegaly, Left Ventricular Hypertrophy, pulmonary edema, pulmonary venous congestion and bilateral pleural effusion (Fig. 1) [1, 2, 15].

***Doppler echocardiography*:** It is most essential diagnostic tool in assessing severity and prognosis. There is increase left ventricular end diastolic diameter (LVEDD), decrease left ventricular ejection fraction (LVEF), dilatation of all cardiac chamber with mitral tricuspid, aortic and pulmonary regurgitation with evidence of pulmonary hypertension (PAH). There may be thrombus in the cardiac chamber with pericardial effusion. (Fig2) [7, 8, 15].

Complications

All type of Cardiac arrhythmias atrial to ventricular tachycardia, atrial fibrillation and flutter, Premature Ventricular Contraction (PVC) to Wolf -Parkinson White Syndrome are reported in PPCM [2,16, 17]. Thromboembolic Phenomenon leading to mortality 30-50% reported with low ejection fraction less than 35 % reported in PPCM [1, 7, 15]. Low birth weight, premature delivery, IUGR, fetal death are common obstetrics complication after PPCM [3,15].

Prognosis

Recovery of systolic function occurs in roughly half of affected women and usually occurs within 6 months of symptom onset [18]. A rapid recovery of EF is often seen in patients after initial diagnosis and dieresis [19]. EF 45% at 2 months after diagnosis predicts full functional recovery in 75% of women [20]. Of the approximately 50% of women without full recovery of systolic function, many benefit from improved EF or functional status, while others have persistent or progressive systolic dysfunction leading to transplant or death [8].

Management

Medical management of PPCM is similar to that of heart failure [1,5,7,11]. Fluid and salt restriction, digoxin, diuretics, vasodilators and anticoagulants are the mainstays of treatment. Strict bed rest of 6-12 months though advocated has chance for increasing risk of pulmonary embolism [7]. Anticoagulant therapy with heparin in antepartum and warfarin in post partum period is suggested in bed ridden patients with low LV EF [7,8,15].

Treatment

Treatment of heart failure with digoxin, diuretics for PPCM until the EF recovers. Medications include angiotensin converting enzyme inhibitors (ACE inhibitors) or angiotensin receptor blockers (ARBs), beta blockers, and diuretics. However, caution must be exercised as ACE inhibitors and ARBS are contraindicated in pregnancy. In addition, patients are advised regarding non-pharmacologic approaches such as salt restriction and the avoidance of offending medications (e.g., nonsteroidal anti-inflammatory drugs) [20].

An important therapeutic option to consider is anticoagulation. Pregnancy conveys a hypercoagulable state that extends approximately 3 months postpartum. During this time, patients with PPCM and depressed systolic function are at high risk for thrombus formation and thromboembolic events. Warfarin anticoagulation should be considered for prevention of morbidity or mortality from thromboembolism [5].

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 levels and increased EF in patients with PPCM.

Anaesthetic Management

The anesthesiologist plays a vital role in managing the patients in ICU, providing labor analgesia, optimizing medical condition of the mothers for cesarean section and administering anesthetics for urgent or elective cesarean section.

Anaesthesia for caesarian section

Anesthetic management for cesarean section in PPCM patients can be challenging to anesthesiologists. Both general anesthesia (GA) and regional anesthesia (RA) have been used.

Though Regional Anaesthesia (RA) has the advantages of sympathetic blockade-induced preload and afterload reduction. Graded Epidural Anaesthesia has been mostly used because of its better hemodynamic stability, while spinal anaesthesia is rarely used. Combined spinal epidural anaesthesia (CSE), with its lower failure rates, faster onset, good muscle relaxation, postoperative analgesia facilities and better maintenance of hemodynamic has also been successfully applied. Recommended precautions should be taken while using RA in anticoagulated patients. GA may be needed in emergency situations or when RA is contraindicated, particularly in anticoagulated patients. There is also increased risk of left ventricular failure (LVF) and pulmonary oedema [21].

Obstetric Management:

PPCM during the antepartum period demands intensive fetal and maternal monitoring. A multidisciplinary approach involving an obstetrician, cardiologist, anesthesiologist and perinatologist may be required to provide optimal care to such patients. Regional analgesia reduces the cardiac stress of labor pain, while application of outlet forceps or a vacuum device can minimize the cardiac stress of the second stage of labor. Following delivery, these patients need monitoring in an Intensive Care Unit (ICU) for early detection and management of uterine autotransfusion-induced pulmonary edema [21].

Summary

Although rare, when a woman is diagnosed with PPCM, only 50% will be expected to fully recover cardiac function. Obstetricians should suspect the diagnosis, particularly if the patient has risk factors, Evaluation should include an echocardiogram to assess the LV systolic function. Treatment includes ACE inhibitors or angiotensin receptor blockers, digoxin, beta blockers, and diuretics. Consideration should be given to anticoagulation. Early diagnosis and prompt treatment in patients with sign symptoms of heart failure with evidence of low EF in echocardiography can improve outcome.

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