

Peri-partum Cardiomyopathy Complicated with Pulmonary Embolism and Left Ventricular Thrombus: A Real Diagnostic and Therapeutic Challenge

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Abstract: *Peripartum (postpartum) cardiomyopathy (PPCM) is a rare dilated cardiomyopathy causing heart failure. It occurs in previously healthy women in the final month of pregnancy and up to 5 months after delivery. It is rare in prep partum and 90% of the cases occur in first two months of postpartum period. The signs, symptoms and treatment of PPCM are similar to that of heart failure. Early diagnosis and proper management are the corner stone for better outcome of these patients. We report a serious case of PPCM complicated by pulmonary embolism and left ventricle thrombosis that was successfully managed .The aim of this report is to make health professionals aware of this entity in a patient with dyspnea in the postpartum period.*

Keywords: Heart failure, postpartum cardiomyopathy, thromboembolism, left ventricle thrombus, cardiac MRI

1. Introduction

Peri-partum cardiomyopathy is a cardiac condition characterized by development of heart failure during the last trimester of pregnancy or during the first five months of post-partum period without any other identifiable cause of heart failure. Although its frequency is not high, clinicians have to keep in mind this condition because of high mortality rate in its severe cases. PPCM, even with a normal heart beat, may sometimes develop ventricular thrombus due to reduced EF and hypercoagulable status during the perinatal period (1). We report a serious case of PPCM complicated by pulmonary embolism and left ventricle thrombosis that was successfully managed.

2. Case Report

A 45-year-old female who had delivered her fifth child through normal vaginal delivery 4 months ago, she is complaining after child birth of gradual worsening dyspnea on effort and paroxysmal nocturne dyspnea. Actually, she was admitted to intensive care unit because of advanced heart failure with severe dyspnea. During pregnancy, she had no dyspnea. She had no previous history of heart disease. She did not have any other known risk factors for pulmonary embolism. She had blood pressure of 90/78 mmHg, heart rate of 100 bpm, respiratory rate of 25 breaths per minute and oxygen saturation was 88% on ambient air. Physical examination showed jugular venous distention, S3 heart sound and bibasilar rales and generalized edema.

An ECG showed sinus tachycardia, right axis deviation and poor R progression in chest leads. Chest radiographic appearance revealed increased cardiothoracic ratio and bilateral pulmonary congestion. Blood test showed the level of B-type natriuretic peptide increased to 1900 pg/ml, C reactive protein was 40 mg/l, and elevated serum troponin.

Transthoracic echocardiography on admission revealed dilated left ventricle and severe left ventricular dysfunction (EF=20%), tricuspid annulus planesystolic excursion (TAPSE) of 12 mm, distended right ventricular and free wall hypokinesia, second degree tricuspid and mitral valve regurgitation with a severe pulmonary hypertension (systolic pulmonary artery pressure =67 mmHg). There were two large thrombi in the LV apex (27 mm × 11 mm, 21 mm × 14 mm). Clinical judgment of pulmonary embolism was made and treatment with unfractionated heparin was initiated, followed by administration of furosemide, and supplementary oxygen. Rapidly, she presented hemodynamic instability based on persistent hypoxemia despite oxygenotherapy and collapse of blood pressure, thrombolysis with atenecteplase was proceeded associated with dobutamine. The outcome was favourable, marked by spectacular regression of symptoms.

A chest computed tomography angiography (CTA) was performed, which confirmed bilateral pulmonary embolism, Autoimmune panel and thyroid function tests were normal. Coronary artery was performed on the 5th day and revealed a normal coronary arteries. one month later a subsequent cardiac MRI (CMR) confirmed severe biventricular systolic dysfunction, with significant resolution of the thrombi previously detected on TTE, late gadolinium enhancement imaging showed subendocardial enhancement in apical and inferior wall with 25 to 50 % transmurality, Based on the facts, that the patient developed heart failure just after delivery without pre-existing cardiac disease, the normal coronary angiogram as well as the CMR findings, PPCM was assumed.

On control ETT, we observed an improvement of the left ventricular EF and a significant reduction of mitral and tricuspid valve regurgitation. The size of the thrombi was gradually reduced, until they resolved completely. The patient was hospitalized for 3 weeks and was discharged

with an EF of 30%. The treatment involved low doses of beta blocker, diuretic, anticoagulant .Then, an outpatient clinic follow-up was planned.

3. Discussion

Peripartum cardiomyopathy is a life-threatening disease characterized by left ventricular dysfunction during the pregnancy or early postpartum period in patients without a history of cardiovascular disease. The definition of PPCM includes the following criteria:

- 1) Development of heart failure in the last trimester of pregnancy or within 5 month after delivery
- 2) Absence of a pre-existing cardiac dysfunction
- 3) Absence of a determinable cause of the cardiomyopathy
- 4) Left ventricular dysfunction with an ejection fraction less than 45 % (2)

Our case met all the diagnostic criteria. Risk factors for PPCM include advanced age (>30 years), multiparity, obesity, preeclampsia, prolonged tocolysis, chronic hypertension and black race. Selenium, thiamine deficiency (3).

Etiology of PPCM is generally unrecognized, however, there are several presumable causes for PPCM, including viral infection, abnormal reactions in the immunology system, circulatory overload accompanied by pregnancy, and endocrine disorders. The role of a 16k Daprolactin derivative produced by proteolytic cleavage of prolactin secondary to unbalanced oxidative stress, which presents during late pregnancy and early post partum period has been noted (4). Medicines causing reduced secretion of prolactin from posterior pituitary gland or working as a D2 receptor antagonist such as bromocriptine have been reported to be effective in controlling PPCM patients (5).

Seventeen percent of PPCM cases are reported to have associated ventricle thrombus. This may be caused by the hypercoagulable status during the perinatal period or congestive blood flow with reduced cardiac function (6).

In accordance with some previously published studies and case reports, the present case suggest, that LGE in acute PPCM is associated with a bad prognosis, In the present case, interestingly, we observed a subendocardial LGE in apical and inferior wall of the left ventricle, which is a typical ischemic pattern of LGE, so that “myocardial infarction with angiographically normal coronary arteries (MINCA)” is a differential diagnosis in our case. However, MINCA is a broad description and its underlying pathophysiology is heterogeneous (spontaneously lysed thrombus, distal emboli, endothelial dysfunction) and often unclear. Anyway, due to the fact that our patient developed a dilated cardiomyopathy with a highly reduced left ventricular function in absence of a pre-existing cardiac dysfunction and any determinable cause of the cardiomyopathy in the last month of pregnancy, the formal diagnosis has to be PPCM. However, since we performed no myocardial biopsy, the true underlying pathophysiology remains unclear like in the majority of cases of PPCM.

Treatment for PPCM in its acute phase is a standardized strategy for heart failure, including ACE-I, diuretic drugs, beta blockers, catecholamine, and mechanical circulatory support devices (intra-aortic balloon pumping and percutaneous cardiopulmonary support). In its chronic phase, oral drugs, including angiotensin converting enzyme inhibitors, beta blockers, and diuretic drugs, are generally used. In patients with severe and refractory heart failure, heart transplant is required. In some patients with PPCM, immunosuppressive agents and anti-prolactin drugs are attempted. There is no established consensus or guideline on the cessation of medical treatment in the chronic phase (7).

Mortality of PPCM was reported to be significant lower than that in idiopathic cardiomyopathy (8). LVDd > 56 mm at the time of diagnosis, EF < 45% at two months after the disease occurrence, existence of LV thrombus at the time of diagnosis, and being African-American have been reported as factors of poor prognosis (9).

4. Conclusion

The diagnosis of peripartum cardiomyopathy is challenging since most women in last month of normal pregnancy or soon after delivery experience dyspnea, fatigue and pedal edema. The thromboembolic phenomenon is the most feared complication of PPCM. The reported mortality of PPCM is 15-50%. Early diagnosis, confirmation, and follow-up with appropriate modalities and prompt start of treatment are the main targets when confronted with such situations.

5. Consent

A written informed consent was obtained from patient for the publication of this paper

6. Conflict of interest

The authors declare that they have no competing interest

7. List of Abbreviations

PPCM: Peripartum Cardiomyopathy

CMR: Cardiac Magnetic Resonance

LGE: Late Gadolinium Enhancement

LV: Left Ventricle

ACE-I: Angiotensin Converting Enzyme Inhibitors

LVDd : Left Ventricular End- Diastolic Diameter

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Figure Legend

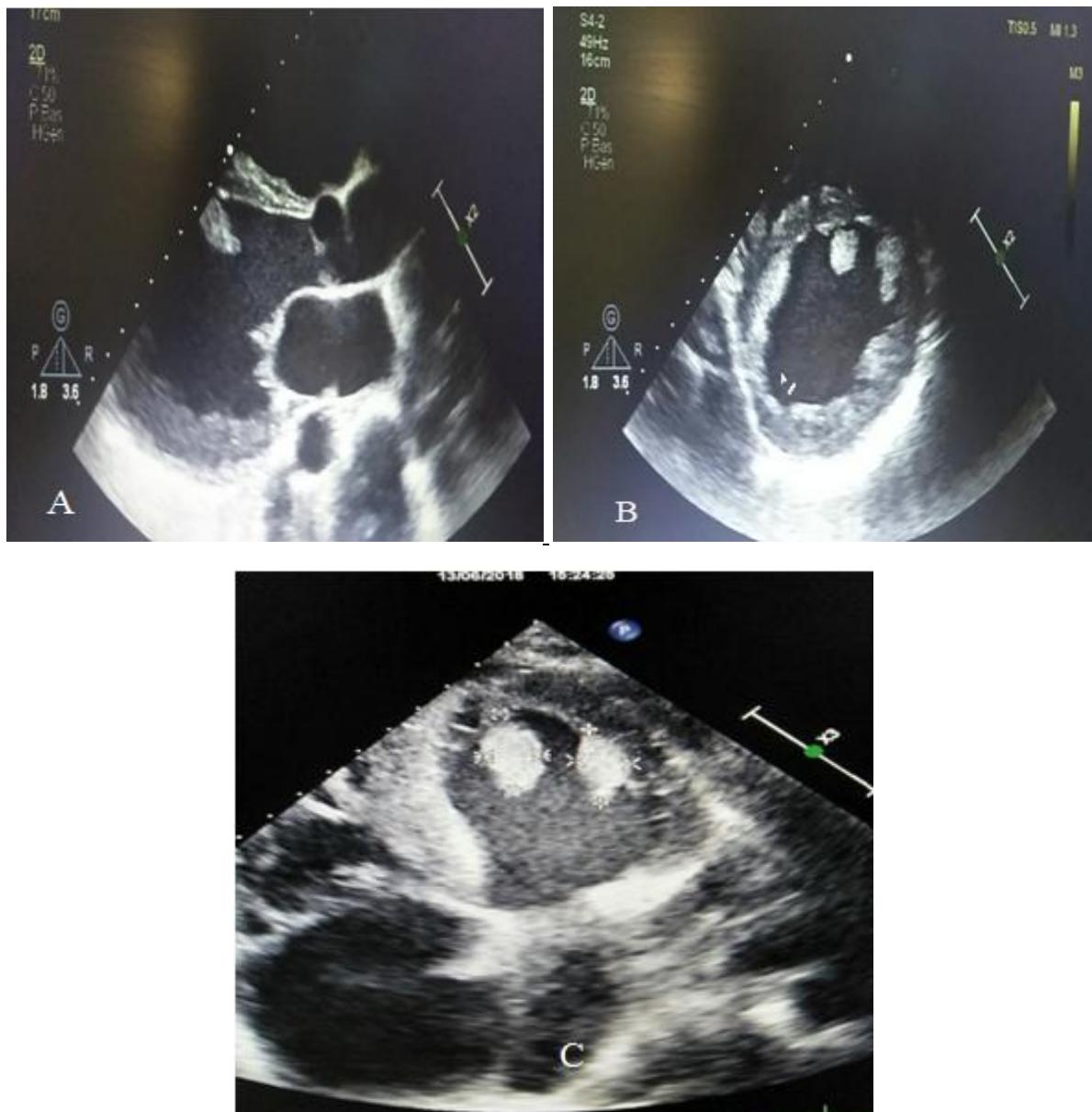


Figure 1: Parasternal long axis (A) and short axis (B) and four chambers views (C) of transthoracic echocardiogram: presence of 2 pedunculated thrombi attached at left ventricle apex .Thrombus dimensions seen in both views are measured as 27 x11 mm, 21x14 mm.

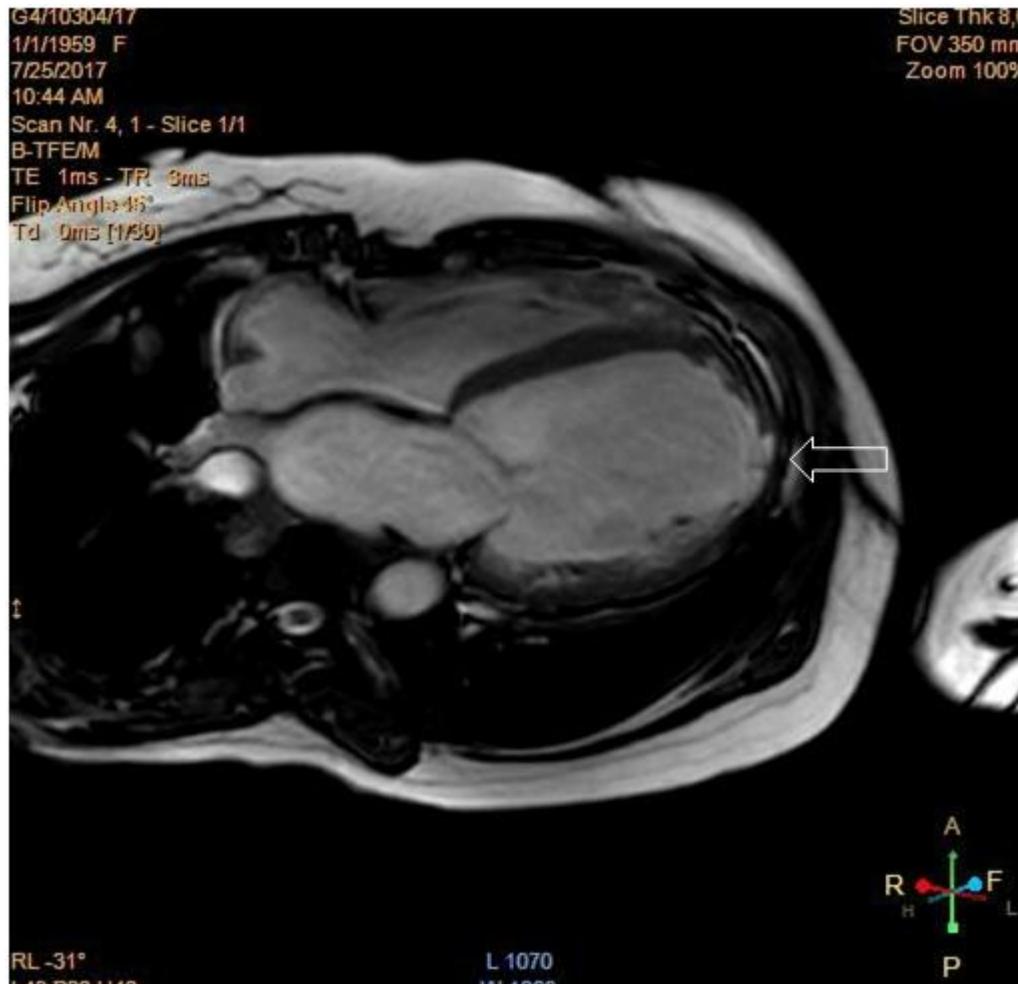


Figure 2: MRI of patient after one month of hospitalization shows dilatation of both ventricles with highly reduced left ventricle function. Subendocardial late gadolinium enhancement in apical wall.