Case series: Peripartum cardiomyopathy- A nightmare

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ABSTRACT:
OBJECTIVE: To know the presentation, course, and outcome of the condition.
Peripartum means the final month of pregnancy through about five months after delivery. cardiomyopathy is a weakness of the heart muscle. So Peripartum cardiomyopathy (PPCM) is weakness of heart muscle in the last month of pregnancy and within five months after delivery in absence of any cardiac disease.
Risk factors for PPCM include advanced maternal age, multiparity, African race, twinning, gestational hypertension, and long-term tocolysis.
PPCM is the diagnosis of exclusion. It is a form of idiopathic dilated cardiomyopathy without any proven cause but it may be because of excessive production of abnormal prolactin, increased oxidative stress, coronary artery disease, viral infection in the heart, various inherited diseases, excess alcohol consumption, smoking and obesity.
PPCM usually presents in postnatal period with shortness of breath, swelling of limbs, palpitation, fatigue. Early diagnosis and initiation of treatment are essential to optimize pregnancy outcome. Treatment mainly concentrates on to improve heart function with Beta blockers, ACE inhibitors, ARB, Diuretics, Anti-arrhythmic, anticoagulants.

Key words: peripartum cardiomyopathy, prolactin,

INTRODUCTION

"Among all types of cardiomyopathy, peripartum cardiomyopathy has a relatively high recovery rate compared to other causes," Lili Barouch, M.D.
Peripartum means the final month of pregnancy through about five months after delivery. cardiomyopathy is a weakness of the heart muscle. So Peripartum cardiomyopathy (PPCM) is weakness of heart muscle in the last month of pregnancy and within five months after delivery in absence of any cardiac disease. It was first described in the 1800s, yet its aetiology is still unclear.
The incidence has been reported to vary by geographical location with rates ranging from 1:15,000 pregnancies in United States, to as frequent as 1:299 in a well-studied population in Haiti and 1:100 in a small region in Sub-Saharan Africa. Incidence not known in INDIA.

CASE SERIES

CASE 1

23year old Un-booked Primigravida, Breech with 39+4 days of pregnancy came with labour pain. Not a known case of hypertension, heart disease, asthma, COPD. Terminated by Emergency C-Section in view of Feto-pelvic disproportion. Postoperatively after 12 hours patient developed dyspnea, on examination Pulse rate was 128 beats per minute, SpO2 88%, respiratory rate 26 cycles per minute. Systolic murmur present. Lungs clear. Cardiologist called for 2D echo, which revealed dilated left ventricles with ejection fraction of 35%. Managed conservatively, recovered completely.

CASE 2

32year old Primi 36 weeks came with chest pain. Not a known case of hyper tension, diabetis, asthma, heart disease. On examination pulse rate 80bpm, BP 110/70mmHg no murmour lungs clear ECG shows ST depression 2D echo advised which shows early dialated cardiomyopathy and left bundle branch block. Patient shifted to high dependency unit. Managed conservatively, delivered by outlet forceps at 38 weeks.

CASE 3

Un-booked 21year old Primigravida with 39 weeks of pregnancy came with labour pains, delivered in casualty. Post-nataly on 2nd day developed breathlessness, cough, swelling of limbs. On examination SpO2 80%, pulse rate 140bpm, BP 80/60 mmHg, respiratory rate 30 cycles/min, chest X-ray taken, shows cardiomegaly with consolidation of lungs, 2D echo done shows dialated left ventricule with ejection fraction of 30%. Managed consevatevely, Recovered.

DISCUSSION

PPCM being a diagnosis of exclusion, a high index of suspicion is essential in detecting early signs of heart failure, thus aiding early recognition and intervention.
Risk factors for PPCM include advanced maternal age, multiparity, African race, twinning, gestational hypertension, and long-term tocolysis.
It's a form of idiopathic dilated cardiomyopathy without any proven cause but it may be because of excessive production of abnormal prolactin, increased oxidative stress, coronary artery disease, viral infection in the heart, various inherited diseases, excess alcohol consumption, smoking and obesity.
Increasing evidence has emerged that prolactin, a 23 kDa protein secreted by the pituitary gland, also plays an important role in the pathogenesis of PPCM. In addition to stimulating lactation from the late stage of pregnancy, prolactin also has cytokine-like effects on tumour growth, differentiation and apoptosis, and influences haemopoiesis, angiogenesis, and coagulation. In mice, the N-terminal fragment of prolactin (16 kDa-PRL) inhibits tumour growth and promotes fibrinolysis by binding to and inhibiting the effects of plasminogen activator inhibitor 1 (PAI-1).[3] PAI-1 is a member of the family of endogenous serine protease inhibitors

(serpins) that beyond inhibiting plasma fibrinolytic activity by blocking urokinase- and tissue-plasminogen activator (uPA and tPA, respectively), promotes tumour angiogenesis and growth.
oxidative stress stimulates the expression of cathepsin D by cardiomyocytes,[3] which cleaves prolactin into the 16 kDa-PRL fragments. This in turn inhibits angiogenesis and promotes endothelial apoptosis, resulting in heart failure.

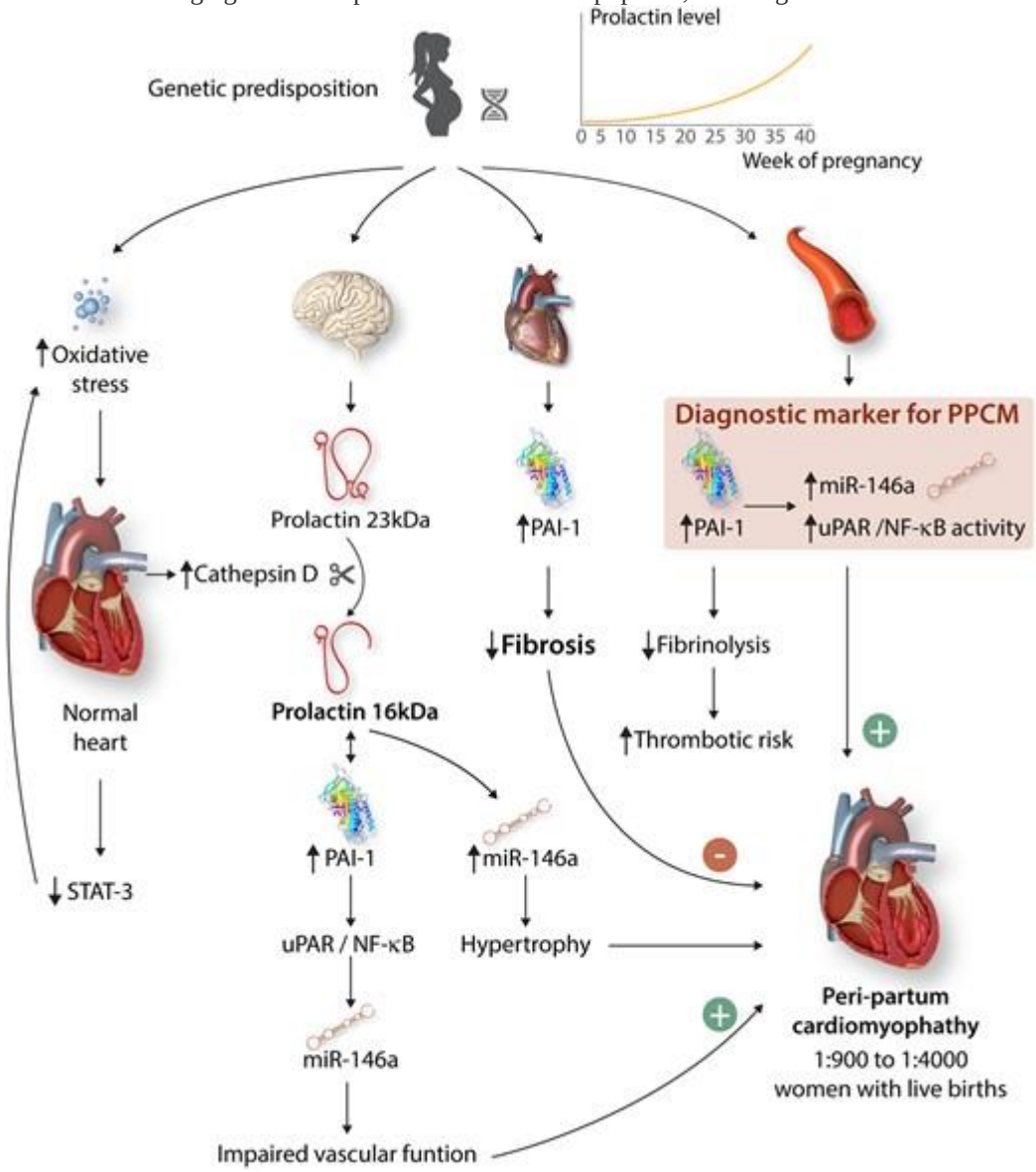

Figure:1 Pathogenesis of Peripartum cardiomyopathy

Amongst PPCM patients, there is an increased frequency of mutations in TTN, the gene that encodes titin, and TTN variants overlap considerably with mutations known to cause dilated cardiomyopathy (DCM).
In a mild case of peripartum cardiomyopathy, typical symptoms such as swelling in the feet and legs, and some shortness of breath can be similar to the symptoms of the third trimester of a normal pregnancy, so these symptoms may go undiagnosed. The patient may then go on to recover without further medical attention.
PPCM usually presents in postnatal period with shortness of breath, swelling of limbs, palpitation, fatigue. Early diagnosis and initiation of treatment are essential to optimize pregnancy outcome.
The New York Heart Association system classifies the severity of symptoms in patients with PPCM:

- Class I - Disease with no symptoms
- Class II - Mild symptoms/effect on function or symptoms only with extreme exertion
- Class III - Symptoms with minimal exertion
- Class IV - Symptoms at rest

PPCM being a diagnosis of exclusion, a high index of suspicion is essential in detecting early signs of heart failure, thus aiding early recognition and intervention. Diagnosis of PPCM rests on the echocardiographic identification of new left ventricular systolic dysfunction, depressed fractional shortening and ejection fraction during a limited period surrounding parturition.
Diagnostic criteria for PPCM (classical); a) Heart failure in the last month of pregnancy and up to five months postpartum. b) Absence of identifiable causes of heart failure. c) Absence of identifiable causes of heart failure.
Additional: Ejection fraction less than 45% and a left ventricular end diastolic dimension of more than 2.7 cm/m^2 of body surface area.

Medical therapy used was similar as in other forms of heart failure. Diuretics, vasodilators, digoxin, ACE inhibitors, inotropes formed the mainstay of treatment. Subsequently, after initial stabilisation, β-blockers were added. Recently dopamine agonist are also advocated.

Some patients recover only part of their heart function over a period of six months or longer. With others, the heart returns to full strength in as little as two weeks.

PREVENTION: To develop and maintain a strong heart, women should avoid cigarettes and alcohol, eat a well-balanced diet and get regular exercise. Universal screening of antenatal mother with echocardiography.

CONCLUSION

In our study two cases developed cardiomyopathy in postnatal period, one patient in antenatal period. Although PPCM is diagnosis of exclusion both antenatally and postnatally High index of suspicion is needed to detect early signs and symptoms of cardiomyopathy to prevent maternal mortality. Follow up and counselling regarding next pregnancy is very crucial.

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