Acute onset peripartum cardiomyopathy in a woman with severe pre-eclampsia: a diagnostic dilemma

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Summary: Peripartum cardiomyopathy (PPCM) is a form of dilated cardiomyopathy that can present as acute life-threatening pulmonary oedema in late pregnancy or early puerperium, its diagnosis is mainly by exclusion of other causes. Morbidity is high due to the reduced physiological reserve in pregnancy. PPCM and severe pre-eclampsia can co-exist and their clinical presentation may overlap, making the diagnosis more difficult and often delayed, with potentially devastating consequences. Here, we would like to share our experience of such a case and present to the readers how we dealt with the challenge. As obstetricians we often do not resort to transthoracic echocardiography, which in our case prompted the diagnosis timely. Lateral thinking and a heightened suspicion does help. Proper diagnosis is extremely important not only for the immediate appropriate management but also for advising long-term lifestyle modifications to minimize risk and counselling for future pregnancy.

Keywords: high-risk pregnancy, cardiac failure, pre-eclampsia, peripartum cardiomyopathy

INTRODUCTION

Acute pulmonary oedema during pregnancy can be due to cardiac problems including cardiomyopathy, pre-eclampsia-related heart failure, valvulopathy or myocardial ischaemia. Non-cardiogenic causes include iatrogenic fluid overload, tocolytics such as β -adrenoceptor antagonists, other medications such as magnesium, sepsis, anaphylaxis and amniotic fluid embolism. Morbidity is increased due to the physiological alterations associated with pregnancy such as tachycardia, hypervolaemia, increased cardiac output, decreased systemic vascular resistance and low colloid osmotic pressure.

Peripartum cardiomyopathy (PPCM) is one such cause of acute pulmonary oedema affecting previously healthy women in late pregnancy or early in the postpartum period. There is a strong association between PPCM and gestational hypertension.^{1,2} The clinical features of PPCM and pre-eclampsia may overlap, especially the predisposition towards pulmonary oedema, desaturation and dyspnoea, making the diagnosis extremely challenging and often delayed, with potentially devastating consequences. We would like to share our experience of diagnosing and managing a case of acute onset heart failure secondary to PPCM in a woman with severe pre-eclampsia, following induction of labour.

CASE REPORT

The mother was a young, primigravida and a chronic smoker with a body mass index of 32.6, who developed gestational hypertension in the third trimester which was well controlled

on Labetalol. At 38 weeks of gestation she presented with a blood pressure of 180/110 mmHg, 4 plus of proteinuria on urine dipstick, severe headache, vomiting and epigastric pain. Blood results were within normal limits apart from a raised urate (0.44 mmol/L). A diagnosis of severe pre-eclampsia was made and decision was made for induction of labour. Cardiotocograph for fetal wellbeing was normal. Fifteen to 20 minutes after administration of 3 mg vaginal prostaglandin E2 tablet, the patient became acutely unwell; confused, dyspnoeic and tachypnoeic. Peripheral oxygen saturation (SpO₂) dropped to 67–70% despite 15 L/minute supplemental oxygen via non re-breathing mask. No intravenous fluids had been administered till this time. Chest examination revealed minimal air entry but no audible crepitations or wheeze. Arterial blood gas analysis showed pH: 7.21, pO₂: 4.92, pCO₂: 5.82 and a BE of –10.0, i.e. metabolic acidosis.

She had a category 1 caesarean section under general anaesthesia. She was positioned with exaggerated left uterine tilt, and preoxygenation was done for three minutes but the SpO₂ remained at about 60%. Rapid sequence induction was done with thiopentone 375 mg and suxamethonium 150 mg, and endotracheal intubation was performed. Anaesthesia was maintained with sevoflurane. Adrenaline 0.5 mg was administered intramuscularly just prior to incision, because anaphylaxis to prostaglandin was a suspected cause for this deterioration. Blood pressure was maintained by metaraminol boluses. After the delivery of a healthy baby, it became much easier to ventilate the patient and SpO₂ increased to 92%. Magnesium sulphate infusion was started according to the pre-eclampsia protocol. Transthoracic echocardiography showed overall moderate to severe left ventricular systolic impairment with an ejection fraction <35% and fractional shortening of 17%. The left ventricular internal dimension at diastole was 4.9 cm, left ventricular internal dimension at systole was 4.1 cm and the

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Table 1 Arterial blood gas analysis and other vital parameters

Parameters	Initial	After baby delivery	Before transfer to ITU
pH	7.219	7.077	7.174
PaO ₂ (kPa)	4.92	6.76	12.25
PaCO ₂ (kPa)	5.82	8.11	7.6
BE (mmol/L)	-10.0	-12.8	-8.4
Hb (g/dl)	11.5	12.4	13.5
K ⁺ (mmol/L)	-	5.09	6.3
Blood pressure (mmHg)	135/105	90/50	110/74
Heart rate (bpm)	101	95	94
Respiratory rate (bpm)	32	12–16 (ventilated)	12–16 (ventilated)
SpO ₂ (%)	67–70	92→100	100

posterior wall was 0.7 cm thick indicating left ventricular size at upper limits of normal when indexed to Body Surface Area (BSA) (3.1) with minimal myocardial thickening during systole. Left atrium and aortic valve was normal, mitral valve was very mildly thickened. Chest X-ray suggested bilateral florid pulmonary congestion. The electrocardiogram was within normal limits. Invasive monitoring (arterial and central venous access) were established demonstrating a mean arterial pressure of 85 mmHg and a mean central venous pressure (CVP) of 11 mmHg. The patient was transferred to the intensive care unit.

A presumptive diagnosis of PPCM was made. A glyceryl trinitrate (GTN) infusion and intravenous frusemide were commenced. A screen for infectious causes of myocarditis was negative. The clinical condition of the patient improved rapidly and she was extubated the next day. The GTN infusion was stopped and she was started on regular ramipril, bisoprolol and prophylactic enoxaparin. Serum tryptase levels collected during the caesarean section and six hours postoperatively were within normal limits, thereby excluding the diagnosis of anaphylaxis. Follow-up at four weeks postpartum showed full clinical recovery. An echocardiogram at six weeks postpartum showed return of normal ventricular function.

DISCUSSION

PPCM is a form of dilated cardiomyopathy defined by ^{2,3}

- Cardiac failure in the last month of pregnancy or within 5 months of delivery;
- Absence of preceding recognizable heart disease;
- Absence of identifiable causes of cardiac failure;
- Left ventricular systolic dysfunction demonstrated by echocardiography.

It is a rare life-threatening complication of pregnancy, and can present acutely. The aetiology remains largely unknown and is likely to be multifactorial. The diagnosis is made by excluding infectious, metabolic, toxic, ischaemic or valvular causes of myocardial dysfunction which could lead to similar acute heart failure. The situation becomes more complicated in case of co-existing problems such as pre-eclampsia. Some studies from the USA^{1,2} have reported significantly higher incidence of gestational hypertension or pre-eclampsia in patients with PPCM (22–68%), compared with the estimated overall PPCM incidence of 1 in 3000–4000 live births.⁴ This association with increased blood pressure brings in the postulation whether hypertension

could trigger the heart failure in PPCM. This is however not supported by the fact that structural changes within the myocardium secondary to hypertension causes impaired myocardial relaxation (lusitropy) resulting in diastolic failure. On the other hand, PPCM is characterized by systolic cardiac failure resulting from inadequate myocardial contractility.⁵

Prompt echocardiography is pivotal in establishing the aetiology of heart failure in the pregnant woman. Cardiac magnetic resonance imaging has an evolving role in complementing the diagnostic information provided by echocardiography. It can measure global and segmental myocardial contraction, and it can characterize the myocardium.⁶ In our patient, trans-thoracic echocardiogram showed essentially normal valvular function, left ventricular size within the upper limits of normal, minimal myocardial thickening of the left ventricle with moderate to severe impairment of overall systolic function and a small pericardial effusion. The presence of systolic dysfunction (rather than diastolic dysfunction) represented an intrinsic failure of the ventricular contractility seen in PPCM rather than left ventricular response to increased afterload (combination of increased systemic vascular resistance and contracted blood volume) characteristic of cardiac failure secondary to pre-eclampsia. A high index of suspicion should always be maintained.

The differentiation of PPCM from the cardiac failure associated with pre-eclampsia is of utmost importance not only to expedite early intervention and management but with regard to prognosis and counselling. Over 50% of women recover to completely normal cardiac function; however, regardless of recovery, PPCM has a recurrence rate of >30% in future pregnancies.⁴ On the other hand, the risk of recurrence for pre-eclampsia is around 15% if one previous pregnancy was affected and around 30% when two consecutive previous pregnancies were affected.⁷

DECLARATIONS

Competing interests: None.

Funding: Not applicable.

Ethical approval: Written patient consent obtained.

Guarantor: SB.

Contributorship: Both authors reviewed and edited the manuscript and approved the final version of the manuscript.

Acknowledgements: Not applicable.

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(Accepted 4 October 2012)