Peripartum cardiomyopathy: a forgotten diagnosis?

VO Olagundoye, Y Seow, MA Ashworth

INTRODUCTION

Peripartum cardiomyopathy (PPCM) is a rare form of left ventricular failure with potentially fatal consequences, occurring during the last month of pregnancy or within the first 5 months after delivery. Diagnosis is by excluding all other causes of left ventricular failure and demonstrating characteristic echocardiographic findings. Management is similar to that of left ventricular failure of other causes.

CASE REPORT 1

A 31-year-old primigravida with no previous medical or surgical history of note was referred at 38 weeks because of pre-eclampsia. She was asymptomatic. Her blood pressure (BP) was 160/100 mmHg and she had 2+ proteinuria. Her uterus was compatible with dates with the fetus in cephalic presentation. Her blood investigations showed haemoglobin 10.5 g/dl, white cell count 19.2×10^9/litre, platelets 169×10^9/litre, aspartamine transaminase 486 IU/litre and uric acid 382 μmol/litre. Urea and electrolytes were normal. A diagnosis of pre-eclampsia with elevated liver enzymes was made. She was started on methyldopa and later that day labour was induced but was delivered by emergency caesarean section for failure to progress. Her immediate postoperative period was satisfactory. On day 5, she complained of a cough and slight shortness of breath. Examination showed a temperature of 37.8°C with few basal crepitations. She was diagnosed as having a chest infection even though her blood and bacteriological tests were normal. Her symptoms improved with antibiotics and she was discharged on day 7. Over the next 2 days she became increasingly unwell with shortness of breath but was reassured by her GP. Her condition deteriorated the next day and she was readmitted urgently. On admission she had a BP of 150/100 mmHg, pulse 120/min with respiratory rate of 40/min. Her oxygen saturation was 90%. Cardiopulmonary examination revealed a raised jugular venous pressure, gallop rhythm, a soft systolic murmur and bilateral basal crepitations Urgent chest radiology showed acute pulmonary oedema and echocardiogram showed mitral regurgitation, global hypokinesia and an ejection fraction of less than 45%, consistent with peripartum cardiomyopathy. She was treated with frusemide, digoxin, ramipril, spironolactone and prophylactic subcutaneous heparin. She was discharged 2 weeks later to be followed up in the cardiology clinic with serial echocardiograms.

CASE REPORT 2

A 36-year-old woman presented in her third pregnancy with no previous history of cardiac disease. Her first pregnancy was delivered by emergency caesarean section 4 years previously for failure to progress; this was followed 1 year later by a spontaneous miscarriage at 6 weeks gestation. The current pregnancy was a spontaneous dichorionic twin pregnancy. Serial ultrasound scans from 24 weeks were normal except for a low-lying placenta as a result of which she was booked for elective caesarean section at 38 weeks. She presented to the delivery suite at 37 weeks and 5 days in early labour and was delivered by caesarean, which was uneventful. Two baby boys weighing 3340 g and 3135 g were delivered in good condition.

She complained of severe shortness of breath 18 hours after delivery. There was no chest or calf pain. On questioning she confirmed she had been short of breath with orthopnoea during the previous 4 days but she did not inform her doctors. Her blood pressure was 107/70 mmHg, pulse was 130/min, respiratory rate was 30/min with oxygen saturation of 92%. Cardiopulmonary examination showed raised jugular venous pressure, gallop rhythm and bilateral basal crepitations. Her haemoglobin was 12g/litre. Urea, electrolytes, liver function test, thyroid function test, ferritin and antinuclear antibody were normal.

Urgent chest X-ray confirmed cardiomegaly with pulmonary congestion and echocardiogram showed dilation of the left ventricle, moderate mitral regurgitation, global hypokinesia, severe impairment of systolic function with ejection fraction less than 45%, all consistent with peripartum cardiomyopathy. She was started on intravenous frusemide. She also received digoxin, lisinoprinol, spironolactone and prophylactic subcutaneous heparin. She was discharge on day 12 post-delivery to be followed up in the cardiology clinic with serial echocardiograms.

DISCUSSION

The aetiology of PPCM is unclear but predisposing factors include advanced maternal age, multiparity, multiple pregnancy, pre-eclampsia and tocolytic therapy. The exact incidence is unknown. In the USA, it is estimated to complicate 1:2400 to 1:15 000 pregnancies (Heider et al, 1999). There is increased risk among women of African descent. The disease has a mortality rate of up to 50%. Death is the result of progressive left ventricular failure and associated complications, usually arrhythmia and thromboembolism (Lampert and Lang, 1995).

The diagnostic criteria for PPCM are:

1. Development of cardiac failure in the last month of pregnancy or within 5 months of delivery
2. Absence of a determinable aetiology for the cardiac failure
3. Absence of demonstrable heart disease before the last month of pregnancy (Demakis et al, 1971).

More recently Hibbard et al (1999) added the fourth diagnostic criteria based on strict echocardiographic indication of left ventricular dysfunction. It includes ejection fraction less than 45% or M-mode fractional shortening less than 30%, or both, and end-diastolic dimension more than 2.7 cm/m^2.

The rarity of this condition means that most obstetricians and GPs will see very

Mr VO Olagundoye is Registrar in Obstetrics and Gynaecology, John Radcliffe Hospital, Oxford OX3 9DZ, **Dr Y Seow** is Senior House Officer in Obstetrics and Gynaecology and **Miss MA Ashworth** is Consultant Obstetrician and Gynaecologist, Stoke Mandeville Hospital, Aylesbury

Correspondence to: Mr VO Olagundoye

few cases during their career. This means that symptoms of early PPCM such as palpitations, malaise and shortness of breath can be confused with normal pregnancy symptoms, leading to delay in making the diagnosis. This problem was echoed by Veille and Zaccaro (1999). The cases described in this article also highlight the difficulty of early diagnosis. In the first case the early symptoms and signs of PPCM were mistaken for chest infection, while in the second case the patient mistook her symptoms for pregnancy symptoms and did not report them until she became very ill after her delivery. A high index of suspicion with prompt initiation of appropriate investigation is the goal. Treatment with immunosuppressive agents has been tried with varied success (Brown and Bertolet, 1998).

The advisability of further pregnancies even in those with complete resolution remains controversial. Lampert et al (1997) in a dopamine challenge test found reduced left ventricular contractile reserve among patients who have recovered completely. In a retrospective study by Elkayam et al (2001) involving 44 patients with subsequent pregnancies, the recurrence rate among 28 patients who had recovered completely was 21% with 0% mortality while the rates were 44% and 19% respectively among 16 patients with persistent left ventricular dysfunction. These two studies highlight the dangers of subsequent pregnancies in these women. Those wishing to have further pregnancies should therefore be appropriately counselled and must be closely monitored through the pregnancy. The two cases presented here were advised against further pregnancies. Oral contraceptives should be avoided in those with persistent left ventricular failure as these patients are at increased risk of thromboembolism.

CONCLUSION

The high mortality associated with PPCM and the difficulty of early diagnosis means that obstetricians and GPs should be vigilant at all times. It is very important that mild cases or initial symptoms are not confused with normal pregnancy symptoms. The application of Demakis's and Hibbard's diagnostic criteria should ensure standardization of reporting. HM

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