Learning from errors

Unilateral presentation of postpartum cardiomyopathy misdiagnosed as pneumonia

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Summary

A 34-year-old woman presented to the emergency department with severe dyspnoea 10 days following a normal-course caesarean delivery. She had been experiencing shortness of breath throughout the third trimester of pregnancy accompanied by tachycardia (110 bpm); however, her evaluation did not include ECG or chest radiography to elucidate the cause. Following delivery, chest radiography was performed demonstrating predominantly unilateral findings interpreted as pneumonia. ECG revealed T-wave inversion in leads V₄–V₆, which was unaddressed. Overnight she deteriorated and a chest CT angiography was performed demonstrating heart enlargement and pulmonary oedema. An echocardiogram established a diminished ejection fraction (EF) of 15–20%, suggesting the diagnosis of peripartum cardiomyopathy. She was treated with angiotensin-converting enzyme inhibitors, spironolactone and furosemide, and was free of symptoms the following month with an EF of 40–45%. Though uncommon, heart failure is a potentially fatal cause of peripartum dyspnoea, often misdiagnosed, meriting further attention.

BACKGROUND

Peripartum cardiomyopathy (PPCM) is a condition of uncertain aetiology present in 1:1300–4000 deliveries.¹ Commonly defined as the de novo development of heart failure in the last month of pregnancy or within 5 months of delivery,² PPCM continues to be one of the most elusive diagnoses causing dyspnoea in pregnancy and the puerperium and should always be considered in this setting. Pulmonary oedema presenting unilaterally is especially prone to misdiagnosis as pneumonia causing substantial delay in proper treatment³ leading to a potentially high mortality.⁴ Therefore, we present a case of PPCM misdiagnosis, highlighting the proper differential diagnosis and management of peripartum dyspnoea, in an attempt to prevent such errors in the future.

CASE PRESENTATION

A 34-year-old woman arrived at the emergency department with severe shortness of breath at rest 10 days following delivery. Upon initial evaluation, her oral temperature was 36.7, blood pressure 163/102, pulse rate 146 and oxygen saturation 88% in room air.

The patient had been previously healthy outside a mild normocytic anaemia for which she underwent no evaluation. She was divorced with two children, of Iraqi-Jewish decent and smoked 'occasionally'. She had not been working and was living off welfare and child support. She had not been taking any medications, drinking alcohol or using illicit drugs, nor did she know of any allergies or illnesses in her family.

Her gynaecological and obstetric history included a prior vacuum-assisted delivery of a baby suffering from transposition of the great arteries 10 years prior to admission; she had undergone two terminations of pregnancy with dilatation and curettage for non-medical reasons and delivered

her second child by caesarean section 8 years prior to admission. Her pregnancies were unremarkable.

Her current pregnancy was uneventful until the third trimester when she started experiencing shortness of breath associated with a non-productive cough. On week 31 of her pregnancy she arrived at the emergency department for evaluation. She described her cough as non-productive, increasing in intensity over the past month and as sometimes accompanied by episodes of irregular uterine contractions. Upon examination, she was noted as being tachycardic with a pulse rate of 110 bpm. The remainder of her physical examination as well as her blood tests, including complete blood count and biochemical analysis, was all within normal. While chest radiography was suggested, she refused for fear of potential harm to the fetus. She received betamethasone for fetal lung maturation but refused tocolysis because she was 'tired of this pregnancy'. An ECG was not performed and she was discharged the following day with recommendation for an ambulatory evaluation of her cough.

Over the next month the patient remained at home, mostly in bed, somewhat relieving her shortness of breath. However, she did continue to experience irregular uterine contractions and episodes of decreased fetal movement for which she was examined in the emergency department on five occasions during weeks 36–37 of pregnancy. Her physical examinations, blood analyses and fetal heart monitoring were all within normal limits and she was treated with hydration; she was discharged with instructions for close fetal, blood pressure, urinary protein and glucose monitoring and an elective caesarean section was scheduled for week 38. On neither occasion was an ECG performed.

Two days prior to the scheduled delivery date, on week 38 of her pregnancy, the patient was admitted to the obstetrics department following rupture of the membranes. Due

Figure 1 Chest radiography at initial presentation demonstrating heart enlargement and pulmonary vascular congestion. A more apparent involvement of the right lung field has lead to the misdiagnosis of right-sided pneumonia.

Figure 2 ECG at presentation demonstrating T-wave inversion in leads V₄–V₆.

to high fetal weight estimation, a caesarean section was performed under spinal/epidural anaesthesia and a healthy 4175 g baby was delivered in a normal-course delivery. The patient was discharged 4 days later with no evidence of complications.

Nine days following delivery the patient presented to her local clinic with a marked deterioration in her shortness of breath. Upon examination, her oral temperature was 36.5 °C, blood pressure 134/98 and pulse rate 126. The remainder of her physical examination was unremarkable with no record of abnormal findings in heart and lung auscultation. Chest radiography was performed demonstrating predominantly right-sided findings (figure 1). The patient was diagnosed with right-sided pneumonia and was discharged with a recommendation for oral antibiotic treatment with cefuroxime and amoxicillin. An ECG was also performed demonstrating T-wave inversion in leads V₄–V₆ (figure 2), but the finding was unaddressed. Overnight her condition

continued to deteriorate and she was referred to the emergency department.

INVESTIGATIONS

Upon arriving at the emergency department, the patient had severe shortness of breath at rest 10 days following delivery. Her vital signs included an oral temperature of 36.7, blood pressure 163/102, pulse rate 146 and oxygen saturation 88% in room air. Treatment with supplemental oxygen by mask yielded an increase in oxygen saturation to 95%. Her physical examination revealed no jugular venous distension, hepatic enlargement or pedal oedema; heart sounds were fast and regular, with no evidence of murmurs or additional sounds. On lung auscultation bilateral crackles were present. Her laboratory analysis showed mild non-specific indicators of stress with a leucocyte count of 9.3×10^3 cells/mm³, haemoglobin value of 10.6 g/dl and a platelet count of 791×10^3 cells/mm³. Her ECG was similar

Figure 3 Chest CT performed following deterioration demonstrating heart enlargement, pulmonary oedema and bilateral pleural effusions mainly on the right.

to the one obtained a day earlier demonstrating T-wave inversion in leads V_4 – V_6 ; however, chest radiography showed a more bilateral presentation compared to the previous one demonstrating both heart enlargement and pulmonary oedema. A chest CT angiography (CTA) performed to exclude pulmonary artery embolisation confirmed the presence of cardiomegaly and pulmonary oedema with bilateral effusions (figure 3). An echocardiogram demonstrated a diminished ejection fraction (EF) of 15–20% confirming the diagnosis of PPCM.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of PPCM is wide and is highly dependent on the clinical setting. While physiological dyspnoea is an extremely common phenomenon during normal pregnancy, affecting up to 60% of healthy women during exercise and 20% of women at rest,⁵ pregnant women are also more likely to suffer dyspnoea related to respiratory tract infections, as well as exacerbations in asthma patients, requiring medical intervention in 20% of cases and affecting neonatal outcome.⁶ Although the patient presented herein was diagnosed with pneumonia, lack of typical signs upon physical examination, as well as prominent signs of inflammation, such as fever or leucocytosis, should motivate the physician to search for another potential cause.

Radiological evidence of pulmonary oedema, which has been initially mistaken in this case to be pulmonary infiltrates, has been described as present in 0.08% of deliveries in some series.⁷ Pulmonary oedema from a non-cardiogenic

cause may result from several reasons, including the low oncotic state of pregnancy, accompanied by hypoalbuminaemia,⁸ in response to certain triggers, such as systemic infections; acute respiratory distress syndrome should also be included in the differential diagnosis whether due to pre-eclampsia or infection, systemic or pulmonary.⁹ However, the lack of indicators of inflammation, as well as the lack of evidence of hypertension or proteinuria, precludes these diagnoses. Furthermore, the presence of ECG abnormalities highly suggests a cardiogenic cause of the oedema.

However, one major diagnostic pitfall is establishing whether the current symptoms are due to a new disease, acquired in pregnancy or an aggravation of a pre-existing one, such as valve disease, arrhythmia or cardiomyopathy. Identifying pre-existing heart disease should rely on a thorough history as well as demonstration of prominent structural defects on echocardiography. In the case of an established new-onset heart failure, two mainly encountered diagnoses that should be considered are PPCM and myocarditis. The diagnosis of PPCM relies on the classical criteria proposed by Demakis *et al*,² consisting of echocardiographic evidence of left ventricular dysfunction (ie, left ventricular EF<45%), with symptoms of heart failure becoming manifest in the last month of pregnancy or within 5 months of delivery, with no other established aetiology for the heart failure. While cardiac MRI has proven effective in both diagnosing PPCM and establishing a prognosis,¹⁰ it is not often found in routine use. Differentiating between PPCM and myocarditis is the subject of great debate; the current need for invasive tissue sampling to gain a definite

Figure 4 A typical ECG of a patient with peripartum cardiomyopathy showing inverted T-waves in leads V₃–V₆.²

diagnosis makes it highly unfavourable. Moreover, due both to the segmental nature of this disease¹¹ and the crucial timing of biopsy, tissue diagnosis is reserved for the subset of patients who do not improve within 1 week of the onset of symptoms.¹²

Late pregnancy and the puerperium are also times of increased risk for pulmonary embolism¹³ and, therefore, any case of acute shortness of breath or tachycardia in this context should alert the physician to pursue this diagnosis here excluded per CTA. Though a cause of severe mortality and morbidity, the natural course of amniotic fluid emboli¹⁴ does not fit well with the subacute nature of symptoms and is an unlikely cause; finally, in cases where tocolytics are used, one should also rule out tocolytic-induced pulmonary oedema demonstrated to be responsible for up to 25% of pulmonary oedema cases.⁷

TREATMENT

The patient was given initial treatment with oxygen by mask with a good response increasing her arterial blood oxygen saturation to 95%. Once heart failure was diagnosed, she was treated orally with angiotensin-converting enzyme inhibitors (captopril 6.25 mg three times daily), intravenous diuretic treatment (furosemide 40 mg twice daily) and 100 mg oral aspirin twice daily. During her admission to hospital she responded well to treatment allowing cessation of oxygen supplementation and tapering of her diuretic dose. She was discharged with instructions for a single daily dose of oral 20 mg furosemide, 5 mg ramipril, 25 mg spirinolactone and carvedilol 12.5 mg twice daily.

OUTCOME AND FOLLOW-UP

Upon examination the following month, the patient was asymptomatic with an EF of 40–45%. She currently continues to be completely free of symptoms 1 year following the event.

DISCUSSION

PPCM is a relatively well-documented disease with the earliest case description taking place as early as 1849.¹⁵ Becoming widely recognised as a discrete entity since the 1930s,¹⁶ PPCM emerged in a large published series in 1971.² Yet,

despite extensive data collection and new insight as for the nature of this disease, it is still continuously misdiagnosed in the clinical setting.

The most common form of PPCM misdiagnosis, described in several series, is pneumonia.^{3 17} In one series, 22% of patients with pulmonary oedema during the peripartum period had predominantly right-sided findings on radiography leading to an erroneous diagnosis of pneumonia, deferring initiation of proper treatment and delaying resolution from 2 days to 5 days.³ Although occurring in less than 1:1000 pregnancies, pneumonia is the most common non-obstetric infection responsible for severe maternal and neonatal outcomes.⁸ Properly differentiating pneumonia from other aetiologies should start with the same basic investigations as for non-pregnant patients, including searching for typical complaints such as productive cough, appropriate findings on lung examination and evidence of systemic inflammation (fever, leucocytosis); it is our view that in the case presented here, the lack of clinical support for this diagnosis should have prompted a search for a different aetiology.

Diagnosing pneumonia in the pregnant patient is especially challenging due to widespread patient refusal of chest radiography. Chest radiography results in a 0.002 Gy exposure—well below the 0.05 Gy threshold demonstrated to cause adverse results.¹⁸ Although in such cases of patient refusal radiography is usually contraindicated, application of lead abdominal protection and patient reassurance may both help in achieving greater cooperation leading to better diagnostic accuracy. Chest CTA is also useful in elucidating the cause of dyspnoea, excluding pulmonary embolisation as a cause, most importantly due to the pro-coagulatory effects of both pregnancy and surgery.¹⁹

Once a non-pulmonary cause for dyspnoea has been suspected, an ECG recording may prove highly useful. Though insensitive and unspecific for PPCM, an ECG is the screening tool of choice for heart disease during pregnancy due to its ease of use, non-invasiveness and lack of potential obstetric adverse effects. Nearly two-thirds of patients eventually diagnosed with PPCM have had a pathological ECG recording upon presentation: the most commonly encountered abnormality is T-wave inversion—the

majority of cases accompanied by evidence of left ventricular hypertrophy. Therefore, it is very probable that performing ECG earlier in this pregnancy might have resulted in a much swifter diagnosis; this is especially in light of the typical ECG pattern observed in this case (figure 2) as compared to the classic ECG of PPCM presented in the pioneering 1971 series publication (figure 4).

Timely diagnosis of PPCM is crucial for proper treatment.³ While patient refusal of chest radiography during pregnancy poses a challenge for lung assessment, we believe that it is mostly lack of awareness for this disease that led to the false diagnosis in this case. A higher degree of suspicion may alert the physician to perform an ECG recording earlier as well as to investigate any abnormalities promptly.

Learning points

- ▶ PPCM is a potentially fatal cause of dyspnoea in pregnancy and the puerperium often misdiagnosed as pneumonia.
- ▶ Awareness of this condition should be raised in order to prevent crucial misdiagnosis having deleterious effects on both mother and infant.
- ▶ Diagnostic measures should include thorough history taking and examination, performing an ECG early in the course of symptoms and obtaining a chest radiograph when possible.
- ▶ Any signs of heart disease on physical examination, evidence of cardiomegaly or pulmonary vascular congestion on chest radiography or ECG changes, most commonly T-wave inversion, should be addressed as soon as possible.

Competing interests None.

Patient consent Obtained.

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Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Amit BH, Marmor A, Hussein A. Unilateral presentation of postpartum cardiomyopathy misdiagnosed as pneumonia. *BMJ Case Reports* 2010; 10.1136/bcr.05.2010.3039, date of publication

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