Rare disease

Diagnostic dilemmas in cardiology

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Summary

A 29-year-old pregnant woman presented in accident and emergency with severe acute left ventricular failure requiring admission to the intensive care unit. A nasal swab was found to be positive for H1N1 making peripartum and viral cardiomyopathies, the most likely differential diagnoses. CT coronary angiography and subsequent invasive angiography revealed an anomalous coronary system thus making ischaemic cardiomyopathy also a possibility. Cardiac MRI played a vital role in identifying the underlying cause, which in this case was that of H1N1 influenza cardiomyopathy. Correct diagnosis in such patients is paramount as this impinges on patients' overall prognosis. In this case aggressive initial therapy including inotropic support and invasive ventilation followed by standard antifailure treatment with β -blockers and angiotensin-converting enzyme inhibitors led to restoration of the patient's left ventricular function and an associated marked improvement in symptoms.

BACKGROUND

This is one of the few published cases of H1N1 influenza cardiomyopathy. In our case, the possibilities of peripartum and ischaemic cardiomyopathy were also raised. Through an array of investigations H1N1 influenza was identified as the underlying cause of the patient's left ventricular systolic dysfunction. This case report very elegantly demonstrates the role of invasive and non-invasive imaging in the diagnosis of cardiac disease. It also emphasises that aggressive treatment including intensive care support is indicated in patients presenting with a new diagnosis of cardiomyopathy, as this can be potentially reversible and have a good prognosis depending on the underlying cause.

CASE PRESENTATION

A 29-year-old primigravida woman at 36 weeks into gestation presented in the accident and emergency department with a 5-day history of fever, general flu-like symptoms, and worsening shortness of breath. At the time of presentation her pregnancy was uncomplicated with normal fetal growth to date. There was no medical or family history and no history of recent travel, animal contact or alcohol abuse. On examination, she was pyrexial (38.2°C), tachycardic (110 beats/min), tachypnoeic (22 breaths/min) and with a blood pressure of 180/83 mm Hg. Auscultation revealed normal heart sounds with a soft systolic murmur throughout the praecordium and bibasal crepitations at the lung bases. Abdominal examination was consistent with a third-trimester pregnancy. Initial investigations showed a high white cell count (12.3×10^9 cells/l) with a predominant neutrophilia and a raised C-reactive protein (239 mg/l). Liver and renal function tests were normal. Troponin I and creatinine kinase were also within the normal range. Arterial blood gases on high-flow oxygen were consistent with Type I respiratory failure with a PO_2 of 11.8 kPa and a PCO_2 of 2.85 kPa.

INVESTIGATIONS

An electrocardiogram showed diffuse T-wave inversion and a chest radiograph left-sided consolidation with upper lobe diversion.

An echocardiogram was performed initially as it allowed us to assess left ventricular function, valvular structure and estimated pulmonary pressures. In this case it showed severely impaired left ventricular function (ejection fraction 0.23) with mild ventricular dilatation.

At an 8-week outpatient follow-up, she was stable and minimally symptomatic (New York Heart Association Functional Class II) with an ejection of 0.35 on repeat echocardiography. CT coronary angiography was arranged at this point for postpartum to exclude an ischaemic aetiology. This showed an absent left main stem, a small circumflex artery and a large dominant right coronary artery with a large right ventricular branch anastomosing to the proximal LAD, which itself appeared to be a small vessel. This was confirmed on subsequent coronary angiography (figure 1). A cardiac MRI (CMR) was arranged to differentiate the aetiologies of viral, peripartum and less likely ischaemic cardiomyopathies. CMR performed 6 months post discharge showed an improved ejection fraction at 0.56 with extensive subepicardial fibrosis but no evidence of subendocardial fibrosis, a hallmark of myocardial ischaemia (figure 2).

DIFFERENTIAL DIAGNOSIS

- ▶ H1N1 influenza cardiomyopathy
- ▶ Peripartum cardiomyopathy
- ▶ Ischaemic cardiomyopathy

TREATMENT

- ▶ Intensive care support including invasive and non-invasive ventilation
- ▶ Emergency caesarean section
- ▶ Oseltamivir (Tamiflu)
- ▶ Diuretics

Figure 1 Coronary angiogram—right anterior oblique view (30°) illustrating a large dominant right coronary artery (RCA) with a large right ventricular branch (RVB) anastomosing to the proximal left anterior descending artery (LAD).

- β -Blockers
- ACE inhibitors

OUTCOME AND FOLLOW-UP

- The emergency caesarean section led to the delivery of a healthy baby.
- Intensive care unit supported led to stabilisation of patient and allowed initiation of antifailure treatment.
- At an 8-week outpatient follow-up she was stable and minimally symptomatic (New York Heart Association Functional Class II) with an ejection of 0.35 on repeat echocardiography.

Figure 2 Cardiac MRI—short-axis view following administration of Gadolinium-contrast agent. There is late enhancement of the inferior epicardial wall extending into the RV aspect of the septum (red arrows).

- At 16-weeks following initial presentation cardiac MRI showed restoration of left ventricular systolic function with an ejection fraction of 0.56.

DISCUSSION

Dilated cardiomyopathy is a common condition with a prevalence of 1:2500.¹ Correct diagnosis of this and its underlying aetiology requires careful history taking and a variety of invasive and non-invasive tests where appropriate. In our case, peripartum and viral-induced cardiomyopathies were the most likely diagnoses. The anomalous coronary anatomy raised the suspicion of an ischaemic component although cardiac biomarkers and the CMR findings did not provide any objective evidence to support this diagnosis. Viral myocarditis and peripartum cardiomyopathy can both cause myocardial fibrosis as the latter can have both inflammatory and non-inflammatory aetiologies.² The pattern of fibrosis seen on CMR was much more typical of a postviral rather than a peripartum aetiology.³ In addition, the temporal time frame and initial presentation were also consistent with an acute infective episode.

This case report very elegantly demonstrates the role of invasive and non-invasive imaging in the diagnosis of cardiac disease. The decision to proceed with invasive investigations that also expose patients to radiation always needs to be carefully considered especially in the younger patient. In a case such as ours where the probability of underlying coronary artery disease is low, CT coronary angiography is the best imaging modality to exclude this as it is not only cost-effective but also avoids the risks of an invasive coronary angiogram.⁴ The finding of an anomalous coronary anatomy however necessitated further evaluation with an invasive coronary angiogram. Further investigation with a cardiac MRI was also paramount as differentiating between ischaemic, viral and peri-partum aetiologies played an important role in further patient management as well as appropriate consultation regarding the feasibility of further pregnancies.

Our case is one of the few published cases of H1N1 influenza cardiomyopathy in pregnancy and the only one in our knowledge where an anomalous coronary anatomy was also present making the diagnosis of ischaemic cardiomyopathy thus a possibility. It emphasises the need for aggressive treatment, including intensive care support in such patients as the underlying left ventricular dysfunction, which can potentially be reversible and have a good prognosis depending on the underlying cause.

Learning points

- Aggressive treatment is important in patients presenting with a new diagnosis of acute left ventricular failure, as this can be reversible.
- Ischaemic cardiomyopathy is a rare cause of left ventricular failure in the younger population, but this needs to be excluded as an anomalous coronary system can lead to myocardial ischaemia. CT coronary angiography is a safe, non-invasive screening test in this population where the likelihood of underlying coronary atherosclerotic disease is low.

- Cardiac MRI is an invaluable imaging modality in patients with cardiomyopathy where the cause of this is unclear as it can differentiate between the different aetiologies something that has important implications in future patient management.
- This case report also touches upon the issues surrounding radiation exposure during pregnancy. Pulmonary embolism and cardiac failure are important differential diagnoses in peri-partum patients presenting with breathlessness. Echocardiography has an important role in the diagnosis of these as it accurately assesses left ventricular function, provides information regarding right ventricular strain and estimates pulmonary artery pressures, which can be raised in pulmonary embolism. It is a safe test in pregnancy that is also inexpensive and readily available.

Competing interests None.

Patient consent Obtained.

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