Rare disease

Spontaneous coronary artery dissection as a cause of sudden cardiac death in the peripartum period

Jatinder Singh Pabla,¹ Lawrence John,² William A McCrea¹¹ Wiltshire Cardiac Centre, The Great Western Hospital, Swindon, Wiltshire, UK² Department of Pathology, The Great Western Hospital, Swindon, Wiltshire, UK

Correspondence to Jatinder Singh Pabla, jatinderpabla@hotmail.com

Summary

Spontaneous coronary artery dissection is a rare cause of acute coronary syndromes and can be potentially fatal. The authors describe a case of multi-vessel coronary artery dissection occurring in a young postpartum mother. The diagnosis was established by coronary artery angiography. However, the patient died following a cardiac arrest. The authors discuss the methods of investigation available and the different treatment options.

BACKGROUND

Acute myocardial infarction is a common medical emergency. However, it is important to note that the majority of these cases in peripartum women are not due to coronary artery thrombosis but due to other causes such as vasospasm or coronary artery dissection.¹ Thrombolytic treatment in these cases can be harmful² and, therefore, greater awareness is needed of the differential diagnoses. Such cases should be managed with early or primary percutaneous coronary intervention.

CASE PRESENTATION

A Caucasian woman in her mid-30s presented with sudden onset transient syncope followed by retro-sternal crushing chest pain radiating to the neck lasting 1 hour before spontaneously resolving. There were no other symptoms. The patient was 8 weeks postpartum with an uncomplicated pregnancy. The medical history was otherwise unremarkable with no risk factors for cardiac disease. Clinical examination was unremarkable apart from a soft ejection systolic murmur in the aortic area. Blood pressure measurements were similar in both arms.

INVESTIGATIONS

The admission ECG showed transient ST elevation in the inferior leads and ST depression in the anterior leads before spontaneously resolving. An echocardiogram showed inferior left ventricular (LV) wall akinesis with overall good LV function. The aortic root appeared normal. A chest radiograph appeared normal. Troponin I was undetectable in the blood.

The patient declined antiplatelet drugs as she was breast feeding and was consented for emergency coronary angiography. This investigation revealed the left main stem (LMS) to be unobstructed, the left anterior descending artery (LAD) had a dissection in the proximal third, an intermediate coronary artery was occluded in the proximal course and the left circumflex (Cx) artery was co-dominant and

demonstrated a dissection in the mid third (figures 1 and 2). The right coronary artery (RCA) was co-dominant and appeared normal.

OUTCOME AND FOLLOW-UP

While changing to a RCA catheter the patient reported chest pain and ST segment elevation was noted in the anterior leads on ECG. Cardiac arrest occurred with pulseless electrical activity initially followed by refractory ventricular fibrillation. Continued angiography demonstrated an occluded left Cx artery and slow TIMI1 flow in the LAD. Despite prolonged advanced life support, and despite performing successful coronary angioplasty, the patient died. The subsequent postmortem findings confirmed the findings at coronary angiography: dissection of the LAD and Cx arteries and a normal RCA (figure 3).

At postmortem, the coronary arteries were cut at 2 mm intervals and then further sectioned at 4 µm for the microscopic slides. The sections of the artery on the slide look different to the appearance in life as after death the vessel collapses as there is no blood pressure to keep the vessel 'open'. Furthermore, in order for the vessel to be processed for histological examination, formalin is added that 'toughens up' the tissue allowing sectioning. This has the consequence of causing further distortion to the shape of the cross-section. This artefactual change does not produce dissection. The clefts in the histology slides produced by the coronary artery dissection are accompanied by inflammatory infiltrate (lymphocytes and neutrophils) and fibrin, which would not be present if the appearance was due to artefact.

It is unlikely that the initial dissection was produced by the coronary angiogram procedure since the left coronary system was engaged smoothly and without difficulty, the dissection sites were distal to the catheter, multiple vessels were involved and there was an indication of coronary dissection at the very initial partial opacification of the relevant segment of the blood vessel during the very first contrast injection.

Figure 1 Dissection flaps visible in the left anterior descending artery and circumflex arteries.

The initial ECG changes may possibly have been the result of RCA spasm or due to the co-dominant nature of the Cx artery.

DISCUSSION

Spontaneous coronary artery dissection (SCAD) is a rare cause of chest pain, acute coronary syndrome and death.² It is more common in younger patients with an average age of 46 years in men and 39 years in women.^{3 4} It is also approximately three times more common in women than men.² Sudden cardiac death is often the first manifestation and the majority of cases are diagnosed at autopsy. Mortality is high at 70%.²

In women, the risk of SCAD appears to be increased during the peripartum period with 25–30% of reports occurring in this setting.^{1 5} Myocardial infarction surrounding pregnancy has been associated with SCAD in 16–54% cases.¹ Most pregnancy related cases occur after delivery of the baby at 13 days postpartum on average,² although the latest reported case occurred at 3 months.^{2 6} Short-term mortality is as high as 38% although up to 82% of survivors were still alive at 3 years.^{1 6}

Women have a higher proportion of LAD artery involvement and LMS involvement whereas men are more likely to have RCA dissection. The Cx artery is infrequently

involved in men and women.⁷ Approximately 40% of SCADs are multifocal in nature.⁸

The aetiology is not fully understood. Most patients presenting with SCAD do not have risk factors for coronary artery disease. The haemodynamic stresses of pregnancy and delivery of the baby are thought to play a part.^{2 8 9} Other theories include structural changes in the arterial walls related to the hormones of pregnancy and eosinophilic infiltration resulting in separation of the tunicae intima and media.² Often dissection occurs without an initial tunica intimal damage suggesting the mechanism lies in degeneration of the tunicae media and adventitia.¹

Histologically, an inflammatory reaction in the adventitia has been described suggestive of periarteritis. The inflammatory infiltrate in this case's histology contained lymphocytes and neutrophils. However, this inflammatory response may be reactive rather than causative.⁹

SCAD should be suspected in all young patients without risk factors for coronary artery disease who present with chest pain, acute coronary syndromes or sudden death. This is especially true for women who present in the peripartum period.

Diagnosis is usually made by emergency coronary angiography,^{2 5} although trans-oesophageal ultrasound, intravascular ultrasound, optical coherence tomography

Figure 2 Dissection flap in the circumflex artery and an occluded intermediate artery.

Figure 3 Left anterior descending (LAD) artery dissection (top left); inflammatory cells and fibrin between the tunica media and adventitia of the LAD artery (top right); circumflex artery dissection (bottom left); normal right coronary artery (bottom right).

and CT coronary angiography have also been used where there is diagnostic difficulty or to guide management.^{1 2 5 6} Non-invasive methods such as CT angiography to diagnose and monitor may also decrease the risk of complications.^{2 6}

The high mortality is falling thanks to early diagnosis and aggressive management.⁵ The rarity of SCAD means that there is uncertainty and a lack of consensus in its management.^{1 2 5}

Medical management may be successful in half of women initially thought suitable for this approach. The remainder may still require intervention later.^{2 5} Medical management involves treatment with heparin, antiplatelets, nitrates and β -blockers.^{1 2 5} Antiplatelets and heparin are thought to help decrease thrombus formation in the false lumen; thus, allowing a more normal flow through the true lumen.^{5 8 10} The use of glycoprotein IIb/IIIa inhibitors is contraindicated.⁵ The use of thrombolysis remains controversial as some cases have reported benefit while others have shown no difference or even extension of the dissection.^{3 5 11–13} Immunosuppressive treatment with prednisone and cyclophosphamide, along with standard treatment, has also been used beneficially in one case lending potential support to the argument that SCAD is an inflammatory process.¹⁴

Where medical treatment is inappropriate (eg, due to haemodynamic compromise or the type of lesion) percutaneous coronary intervention, sometimes assisted by intravascular ultrasound, and coronary artery bypass grafting can be considered.⁵ A LV assist device implant followed by heart transplantation has also been successful.¹⁵

Peripartum SCAD is not an absolute contraindication to future pregnancies. Data on the risk of recurrence are limited and so it may be best to avoid further pregnancies.¹⁶ As such, pre-pregnancy counselling should be considered.

Learning points

- ▶ SCAD is a rare cause of acute coronary syndromes and sudden death.
- ▶ It should be considered in all peripartum women who present with chest pain.
- ▶ Thrombolytic treatment may be harmful.
- ▶ It should be diagnosed expediently by coronary or CT angiography.

Competing interests None.

Patient consent Not obtained.

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