Thomas A Kite, Benjamin A Marrow, Sarah Nduwayo and Gerry P McCann accepted manuscript.
Version of record available http://dx.doi.org/10.1136/bcr-2019-231731

BMJ Case Reports

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TITLE OF CASE

Non-ST elevation myocardial infarction, non-obstructive coronary arteries, and severe regional microvascular dysfunction in a patient with dilated cardiomyopathy

SUMMARY

Cardiac magnetic resonance (CMR) imaging with late gadolinium enhancement (LGE) is a key modality in providing localisation and characterisation of myocardial injury in patients diagnosed with myocardial infarction with non-obstructive coronary arteries (MINOCA). We present a case that demonstrates the unique ability of CMR to provide crucial information in instances of uncertainty. A 71-year-old patient with dilated cardiomyopathy (DCM) presented with symptoms suggestive of acute myocardial infarction. The diagnosis of MINOCA was confirmed following coronary angiography. CMR imaging with LGE confirmed presence of apical infarction. Quantitative myocardial perfusion mapping demonstrated severely reduced blood flow in the non-infarcted septal segments proximal to the distal infarcted territory. The precise aetiology of apical infarction remains uncertain and is likely attributed to coronary plaque rupture. However, concomitant severe regional microvascular dysfunction is also appreciated. This is a recognised, but not well-described, phenomenon in DCM and may contribute to repetitive ischaemic injury and disease progression.

BACKGROUND

This case highlights MINOCA and the role of CMR imaging with LGE in establishing aetiology. MINOCA is a heterogeneous condition in which the underlying cause is often difficult to ascertain. Fully quantitative myocardial perfusion mapping, a novel CMR technology, is likely to be widely available in the near future and provides additional strength to CMR as a diagnostic tool. The ability of CMR to identify unsuspected pathology, including regional microvascular dysfunction, is demonstrated in this case.

CASE PRESENTATION

A 71-year-old female presented to the emergency department with an acute episode of central chest and left arm pain. Medical history comprised “non-ischaemic” dilated cardiomyopathy (confirmed with non-obstructive coronary angiography performed in 2015), moderate left ventricular systolic dysfunction, and a right-sided total hip replacement. The patient was taking maximal tolerated doses of beta-blocker, ACE inhibitor and mineralocorticoid receptor antagonist therapy prior to admission. Clinical examination was unremarkable.

INVESTIGATIONS

12-lead electrocardiography showed sinus rhythm with normal axis, incomplete left bundle branch block, poor R-wave progression, and widespread ST segment depression with T-wave inversion (Figure 1). High-sensitivity Troponin I was elevated at 8445ng/L. Chest x-ray confirmed cardiomegaly with no evidence of acute cardiac decompensation. Dual antiplatelet (DAPT) and anti-thrombotic therapy was commenced as treatment for presumed non-ST elevation myocardial infarction.

Transthoracic echocardiography (TTE) demonstrated a dilated left ventricular cavity and regional wall motion abnormalities in mid to apical septal and inferior walls. Left ventricular ejection fraction (LVEF) was calculated at 22%. Coronary angiography revealed smooth, unobstructed epicardial coronary anatomy with mildly sluggish contrast flow throughout the coronary circulation (Videos 1-2). Pulmonary embolus was excluded with computed tomography pulmonary angiography. The patient was subsequently discharged on dual antiplatelet therapy (DAPT) with a plan for outpatient CMR imaging.

Outpatient CMR with adenosine stress perfusion to assess for microvascular dysfunction was performed on a 3 Tesla scanner 3 months after the patient’s index admission. The left ventricle was noted to be moderately dilated and LVEF calculated to be 25% (Video 3). Focal LGE involving the apical anterior and apical inferior segments with sparing of the apical septal and apical lateral segments was observed (Figure 2). Appearances are consistent with a small distal LAD infarction and the known diagnosis of DCM, as the degree of systolic dysfunction was disproportionate to the size of infarction. On visual analysis, first pass perfusion stress imaging did not indicate a perfusion defect (Video 4). However, inline quantitative myocardial perfusion mapping demonstrated severe reduction in stress blood flow (1.0-1.4ml/g/min) throughout the non-infarcted septal segments which was not apparent in the anterior, lateral or inferior walls (1.7-2.2ml/g/min) (Figure 3). Rest blood flow was in the low-normal range (0.5-0.65ml/g/min). This novel technique, described in recent literature[1], strikingly illustrates the intrigue of this case; severely reduced myocardial perfusion confined to the septal segments, proximal to the site of apical infarction.

OUTCOME AND FOLLOW-UP

At the time of writing the patient has commenced Sacubitril/Valsartan in addition to established standard heart failure therapy, and reported no significant cardiovascular symptoms at a recent outpatient clinic appointment.

DISCUSSION

MINOCA is defined by clinical features of acute myocardial infarction alongside demonstration of non-obstructive coronary artery disease following coronary angiography (no coronary artery stenosis ≥50%)[2, 3]. Commonly considered a working diagnosis with varied possible aetiology (Table 1), MINOCA accounts for approximately 6% of all patients diagnosed with acute myocardial infarction, and carries a 5% risk of mortality at 12 months[4]. The European Society of Cardiology STEMI guidelines and MINOCA position paper, both published in 2017, outline the phenomenon of MINOCA and advise consideration of CMR imaging with LGE due to its unique ability of tissue characterisation and demonstration of oedema, fibrosis and scar[2, 3]. This case demonstrates the important additional information that CMR imaging with LGE and quantitative myocardial perfusion mapping can provide. It illustrates the diagnostic uncertainty often associated with MINOCA, and how novel imaging techniques can assist in establishing causal aetiology.

Concomitant pathology is the most likely explanation for the unusual CMR appearances in this patient. The LGE is typical for myocardial infarction in the distal LAD territory, and the small size consistent with the documented Troponin elevation. The mechanism of infarction is less certain, but most likely due to coronary plaque erosion and rupture. However, given the background of DCM and demonstration of unobstructed epicardial coronary arteries, coronary embolism is also a possible aetiology.

Interestingly, in addition to myocardial infarction we have demonstrated severe regional microvascular dysfunction, a recognised entity in DCM. Conventionally considered as a “non-ischaemic” cardiomyopathy, published data have suggested reduced myocardial blood flow in DCM[5]. In the absence of obstructive epicardial coronary artery disease, these findings may support a hypothesis of “microvascular ischaemia”, in which repetitive stress-induced myocardial stunning drives progressive left ventricular dysfunction and dilatation[6]. The combination of pathology demonstrated on CMR imaging in this case has, to the best of our knowledge, yet to be documented.

Table 1: Aetiology of MINOCA

Pathophysiological mechanisms of MINOCA	
Coronary causes	**Non-coronary causes**

Coronary plaque rupture Coronary artery spasm Coronary thrombus/embolism Coronary microvascular disorders • Microvascular angina • Microvascular spasm • Slow flow phenomenon	**Myocardial causes** Myocarditis Takotsubo cardiomyopathy Other cardiomyopathies (e.g. DCM, HCM) **Non-myocardial causes** Pulmonary embolism Renal disease Sepsis

LEARNING POINTS/TAKE HOME MESSAGES

- MINOCA is a syndrome characterised by clinical evidence of myocardial infarction, with non-obstructive coronary arteries confirmed on angiography
- MINOCA is a syndrome with wide-ranging aetiology, and often results in diagnostic uncertainty amongst physicians
- CMR imaging has an important role to play in establishing a diagnosis due to its excellent ability to localise and characterise myocardial injury
- Microvascular dysfunction, a recently recognised phenomenon in DCM, can be assessed by novel CMR myocardial perfusion techniques

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FIGURE/VIDEO CAPTIONS

Figure 1: Admission electrocardiogram

Figure 2: CMR late gadolinium enhancement imaging

Figure 3: CMR quantitative myocardial perfusion imaging

Video 1: Right coronary artery angiogram

Video 2: Left coronary artery angiogram

Video 3: 4 chamber cine CMR imaging

Video 4: Stress perfusion CMR imaging

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