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CASE REPORT
Cardiac imaging

An unusual case of apical myocarditis: a case report

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Background Myocardial infarction with non-obstructed coronary arteries (MINOCA) syndrome accounts for ~6–8% of acute coronary syndrome presentations. Historically, MINOCA has been thought of as a benign condition, however, recent evidence suggests that some aetiologies of MINOCA such as cardiomyopathies are associated with significantly higher mortality than other causes such as myocarditis. Therefore, identifying the underlying cause of MINOCA is important in determining patient management and prognosis.

Case summary We describe the case of a 58-year-old lady with an acute admission with MINOCA syndrome. Cardiac magnetic resonance (CMR) examination on Day 9 demonstrated hypertrophy of the apical segments of the left ventricle (LV), with diffuse mid-wall hyper-enhancement on late gadolinium enhancement (LGE) images. T2-weighted imaging was suggestive of active inflammation in the hypertrophied segments. A repeat CMR scan was performed 3 months later showed normalization of LV wall thickness, LGE and T2 values in the apical segments.

Discussion This case report highlights the benefits of CMR with oedema-weighted imaging in the acute stages of MINOCA syndrome, as well as the importance of serial imaging in this patient cohort. While baseline imaging raised the possibility of apical hypertrophic cardiomyopathy, resolution of apical hypertrophy on follow-up CMR showed that the patient had acute myocarditis, specifically involving the apical segments.

Keywords MINOCA syndrome • Myocarditis • Takotsubo • Cardiac magnetic resonance • T2-weighted imaging • Case report

Learning points

- Myocarditis is a common cause of myocardial infarction with non-obstructed coronary arteries (MINOCA) syndrome.
- Acute myocardial oedema from myocarditis can result in transient thickening of the LV wall, which on standard echo and cardiac magnetic resonance (CMR) imaging can resemble LV hypertrophy.
- The resolution of LV wall thickness can be monitored through serial imaging.
- Differentiation between myocarditis and Takotsubo can be challenging without tissue biopsy, however, acute CMR with oedema-weighted imaging can help exclude chronic conditions such as apical hypertrophic cardiomyopathy.
- Differentiating myocarditis from other causes of MINOCA syndrome is important, as it often carries a favourable long-term prognosis and may not require long-term treatment.

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Introduction

Myocardial infarction with non-obstructed coronary arteries (MINOCA) is characterized by clinical evidence of myocardial infarction with normal or near-normal coronary arteries on angiography. The clinical syndrome encompasses a wide range of causes which include coronary (plaque rupture, spontaneous dissection, spasm, and thromboembolism) and non-coronary causes (myocarditis, Takotsubo, and other forms of cardiomyopathy).[1] Due to its multiparametric capabilities in assessing tissue characterization, cardiac magnetic resonance (CMR) imaging is considered an essential tool by the European Society of Cardiology in the diagnostic work-up of MINOCA patients.[2] We present a case of MINOCA syndrome, where diagnostic uncertainties were overcome by the use of early and serial CMR imaging.

Timeline

Timeline of clinical events

May 2019 Day 1	Presentation with chest pain, ST-elevation on ECG and raised serum troponin; coronary angiogram revealed unobstructed coronary arteries
Day 3	Echocardiogram showed globally mildly impaired left ventricle (LV) systolic function. The patient was diagnosed with 'MINOCA' syndrome, and discharged on ACE-inhibitor and beta-blockers.
Day 9	Cardiac magnetic resonance (CMR) scan demonstrated hypertrophy of apical LV segments, with diffuse mid-wall hyper-enhancement on late gadolinium enhancement images. T2-weighted imaging demonstrated high signal intensity in hypertrophied segments in keeping with acute inflammation. At this, the differential diagnosis included acute coronary syndrome with bystander apical hypertrophic cardiomyopathy and acute myocarditis. Due to diagnostic uncertainty, a repeat CMR scan was requested.
September 2019	Follow-up CMR scan at 4 months showed normal LV systolic function. The LV thickness of the apical segments returned to normal limits and there was complete resolution of late gadolinium enhancement. T2 values also returned to within normal range, indicating resolution of oedema. Apical hypertrophic cardiomyopathy was excluded and the diagnosis was changed to apical myocarditis; the patient was taken off ACE-inhibitors and beta-blockers and discharged from cardiology follow-up.

Case presentation

A 58-year-old lady was admitted to the coronary care unit of a tertiary cardiology centre with sudden onset of chest pain, associated with widespread ST-elevation on a 12-lead ECG (*Figure 1*). She denied any symptoms of recent viral illness and had no family history of ischaemic heart disease or cardiomyopathy. She also had no past medical history. Clinical examination was unremarkable; her heart sounds were normal, she was clinically euvolemic and her chest was clear to auscultate. Urgent invasive coronary angiography found her to have unobstructed coronary arteries. Serum high-sensitive troponin was significantly raised at 1998 ng/L (reference range <37 ng/L). Her inflammatory markers including white cell count and C-reactive protein were not elevated. Transthoracic echocardiogram showed marked global hypokinesia with overall mildly impaired left ventricular systolic function. Based on these findings an initial diagnosis of MINOCA syndrome was made and a CMR scan (1.5 Tesla) was performed for further clarification 9 days following her presentation. Cine images demonstrated non-dilated left ventricle (LV) with preserved overall systolic function (ejection fraction 67%). The apical segments had subtle hypocontractility but appeared hypertrophied (max thickness 14 mm). T2-weighted images (turbo spin-echo with dark bloods) were suggestive of active myocardial inflammation, most evidently in the apical segments, where the signal intensity (SI) ratio between myocardium and skeletal muscle was 2.8 (SI in the basal segments was 1.8). Late gadolinium enhancement (LGE) imaging demonstrated diffuse mid-wall hyper-enhancement in the apical segments (*Figure 2*), thereby fulfilling the updated Lake Louise criteria for myocardial inflammation.[3] Given this pattern and clinical context, differential diagnosis included acute coronary syndrome with bystander apical hypertrophic cardiomyopathy, acute myocarditis, and Takotsubo cardiomyopathy. She was discharged from hospital on an ACE-inhibitor (Ramipril 2.5 mg once a day) and a beta-blocker (Bisoprolol 2.5 mg once a day).

She returned for an interval CMR scan in 3 months (*Figure 3*). Cine imaging on this occasion again found non-dilated LV with preserved systolic function (ejection fraction 67%); however, the thickness of the LV apical segments had returned down to normal range (max 9 mm thickness). On T2-weighted images, the SI ratio in the apical segments had reduced to 1.7. Late gadolinium enhancement imaging demonstrated resolution of the apical diffuse mid-wall hyper-enhancement seen in the previous scan. It was therefore concluded that the thick appearance of the apex in the acute scan was caused by myocardial oedema from acute myocarditis rather than apical hypertrophic cardiomyopathy, which during the course of time had fully resolved.

Discussion

The prevalence of MINOCA syndrome is ~6–8% among patients presenting with acute coronary syndrome.[4] A meta-analysis of studies using CMR as a diagnostic tool in MINOCA has demonstrated myocarditis to be the leading cause and early CMR imaging was able to secure the diagnosis in up to 87% of the cases.[5] Historically, MINOCA has been thought of as a benign condition, however, differentiating myocarditis from other causes of MINOCA can carry significant prognostic relevance. Dastidar *et al.*[6] performed CMR on 388 consecutive MINOCA patients at a median of 37 days from presentation, and over a median follow-up of 3.5 years, 5.7% of patients died. Mortality rates were highest amongst patients with CMR evidence of

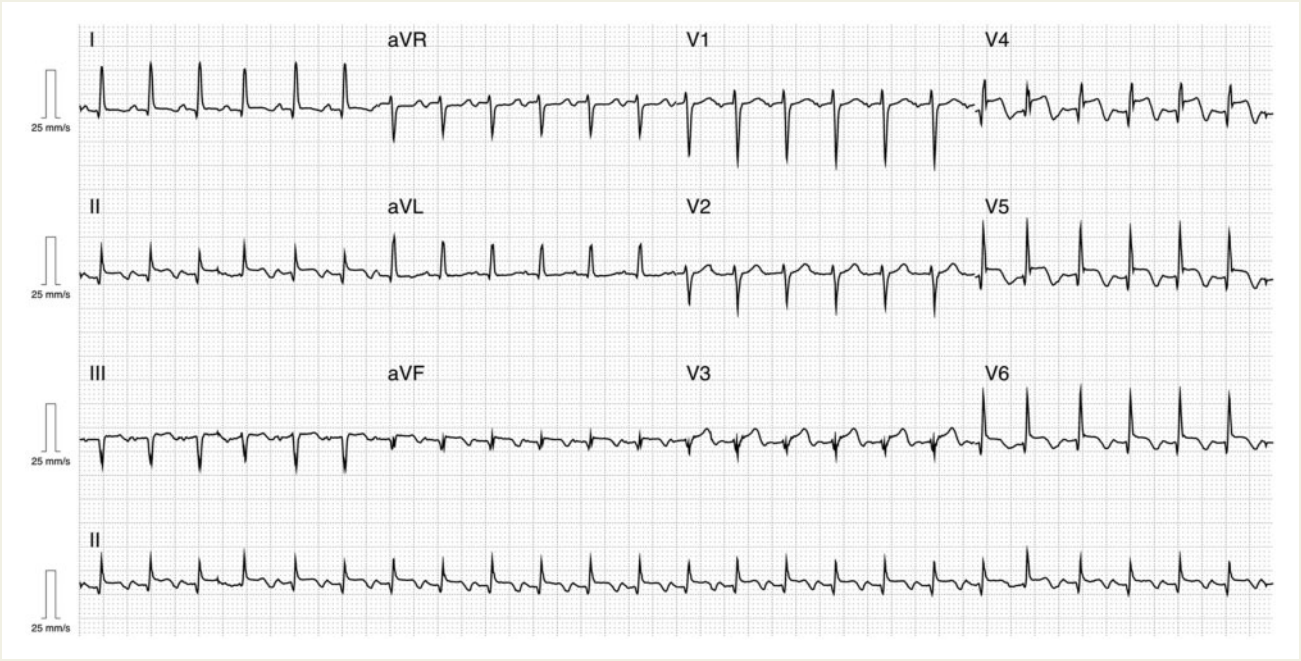

Figure 1 Twelve-lead ECG at presentation.

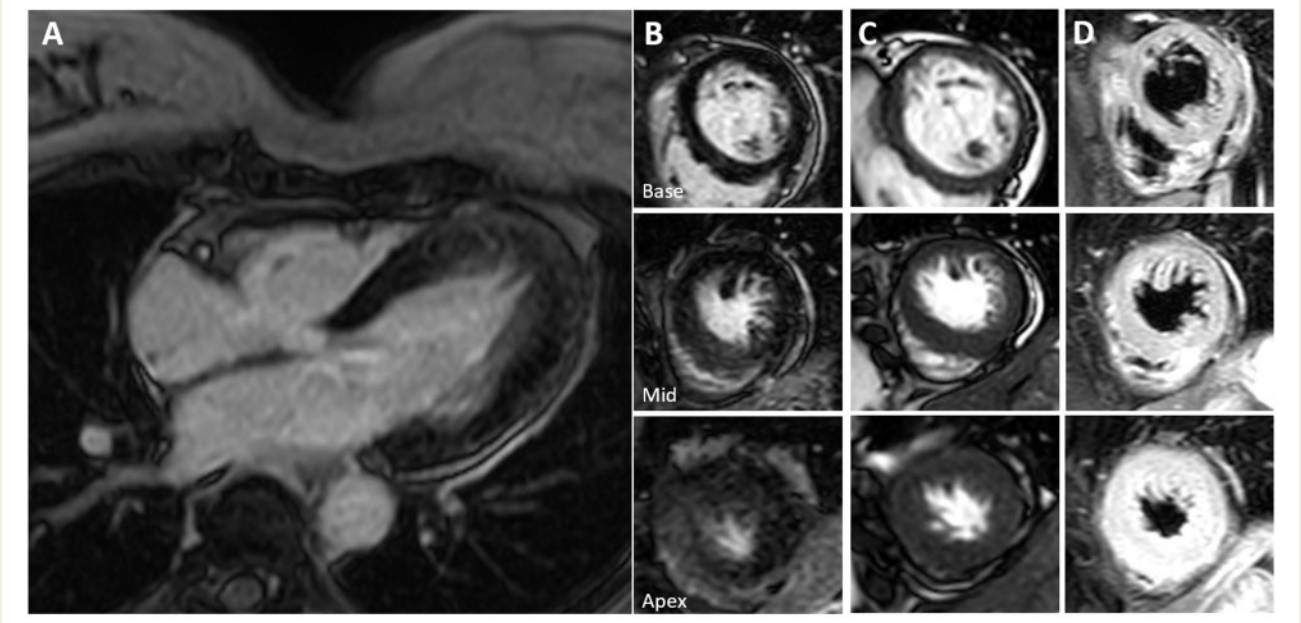

Figure 2 Acute MRI scan. Apical four-chamber view with late gadolinium enhancement (LGE) (*A*); basal, mid, apical short-axis slices by LGE (*B*), end-diastole cine (*C*), and T2-weighted images (*D*).

cardiomyopathy (15% mortality), followed by myocardial infarction (4% mortality), myocarditis and normal CMR (2% mortality in both). Following multivariate cox regression analysis including clinical and CMR parameters, ST-segment elevation on presenting ECG and cardiomyopathy on CMR imaging were the only two significant predictors of mortality, underpinning the added diagnostic and prognostic utility of performing CMR early after presentation in MINOCA patients.[7,8]

The phenomenon of transient LV wall thickening in the acute stages of myocarditis has been observed previously using

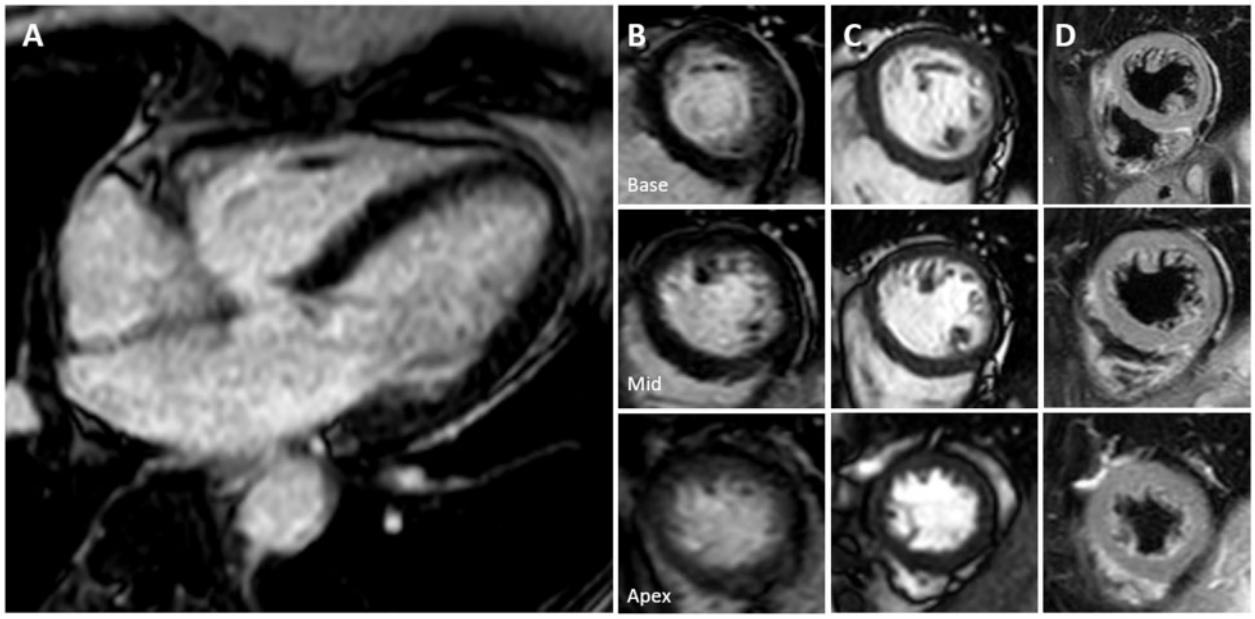

Figure 3 Follow-up scan. Apical four-chamber view with late gadolinium enhancement (LGE) (*A*); basal, mid, apical short-axis slices by LGE (*B*), end-diastole cine (*C*), and T2-weighted images (*D*).

echocardiography and CMR.[9–12] Zagrosek *et al.* performed CMR scans on 21 acute myocarditis patients and found them to have significantly higher LV mass in comparison to controls. By performing convalescent scans at 12 months, they observed a significant reduction these patients' LV mass, which paralleled the normalization of initially increased myocardial SI on T2-weighted images.[12] This acute change in ventricular wall thickness is thought to reflect oedema formation and cellular infiltration, as suggested by previous endocardial biopsy-based histological studies.[13] In the current case, the elevated T2 values on the acute CMR study suggested the thick appearance of the apical segments could be reflective of oedema rather than cellular hypertrophy. Most if not all case reports to date have shown a global concentric pattern of oedema and hypertrophy, whereas this case demonstrates an unusual focal involvement of the apical segments mimicking apical hypertrophic cardiomyopathy. Quantitative parametric mapping techniques such as native T1- and T2-mapping are highly sensitive to oedema and both have now been incorporated into the updated Lake Louise Criteria in 2018 for diagnosing myocarditis.[3]

Given the lack of viral symptoms and a normal CRP on admission, another differential diagnosis that also should be considered in this case is Takotsubo cardiomyopathy. Whilst this typically results in more marked regional wall motion abnormalities in non-coronary distributions, a large prospective study by Eitel *et al.*[14] found significant overlaps between the two conditions, with 67% of Takutsubo patients fulfilling the Lake Louise consensus criteria for the CMR diagnosis of acute myocarditis; and 9% having evidence of patchy LGE.

Conclusion

Myocarditis is a common cause of MINOCA syndrome and has a favourable prognosis compared with other causes such as cardiomyopathy and myocardial infarction. Early CMR imaging allows the detection of oedema which can help secure the diagnosis. Acute myocardial oedema can result in transient LV hypertrophy; most reported cases to date have shown global concentric hypertrophy at acute presentation, however, this case demonstrates a more focal pattern involving the apical segments which resolved over time. Differentiation between myocarditis and Takotsubo remains challenging without tissue biopsy, however, acute CMR with oedema-weighted imaging can help exclude chronic conditions such as apical hypertrophic cardiomyopathy.

Lead author biography

Dr Arka Das (MbChB, MRCP) is a cardiology trainee working at Leeds General Infirmary, Yorkshire, UK. He is currently undertaking a PhD with the University of Leeds, exploring the use of novel quantitative cardiac magnetic resonance techniques in cardiovascular disease.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

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Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none declared.

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