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CASE REPORT

INTERMEDIATE

CLINICAL CASE

Acute Myocarditis Mimicking Hypertrophic Cardiomyopathy in Marfan Syndrome and Morphologically Abnormal Mitral Valve

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ABSTRACT

A 40-year-old man with Marfan syndrome presented with chest pain and troponin elevation. Urgent echocardiography was suggestive of hypertrophic cardiomyopathy, but cardiovascular magnetic resonance identified features of acute myocarditis. Repeated imaging 4 months later showed resolution of septal thickness, confirming acute myocarditis. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2022;4:105–110) Crown Copyright © 2022 Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

A 40-year-old white man with a medical history of Marfan syndrome and mild aortic root dilatation presented with a 24-hour history of chest pain, intermittent palpitations, light headedness, and rigor. The results of admission observations were unremarkable. He quickly experienced signs of left-sided heart failure with increasing oxygen requirements, a cough, and pink frothy sputum.

LEARNING OBJECTIVES

- To underline the role of multimodality imaging in complex cases where there is diagnostic uncertainty.
- To revisit the central role of cardiovascular magnetic resonance in the diagnosis of myocarditis.
- To highlight the importance of serial imaging in confirming a correct diagnosis.

MEDICAL HISTORY

This patient was under surveillance for mild aortic dilatation secondary to Marfan syndrome. He had undergone an atrioventricular node ablation in 2016 for palpitations, and he took regular atenolol.

INVESTIGATIONS

Blood studies revealed a lymphocytosis of 12.21×10^9/L (reference range 4-11 $\times$ 10^9/L) and elevated C-reactive protein 218 mg/L (reference range <10 mg/L), with negative blood cultures. High-sensitivity troponin I was elevated at 1,405 ng/L (reference range <57 ng/L). The results of both D-dimer and

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ABBREVIATIONS AND ACRONYMS

CMR = cardiovascular magnetic resonance

HCM = hypertrophic cardiomyopathy

LV = left ventricle

LVEF = LV ejection fraction

LVH = left ventricular hypertrophy

LVOT = left ventricular outflow tract

MINOCA = myocardial infarction with non-obstructive coronary arteries

SAM = systolic anterior motion

COVID-19 polymerase chain reaction swabs were negative.

The admission electrocardiogram and chest x-ray are shown in **Figures 1 and 2**. A CT aortogram promptly excluded an acute aortic syndrome but demonstrated signs indeterminate for COVID-19 pneumonia (**Figure 3**). Echocardiography showed left ventricle hypertrophy (LVH) with chordal systolic anterior motion (SAM) resulting in left ventricle outflow tract (LVOT) obstruction and mitral regurgitation (Video 1). There was akinesia of the mid-anterior and septal walls and all apical segments, with overall severe LV impairment (LVEF 30%-35%) (Video 2). Coronary angiography revealed non-obstructive coronary arteries (Video 3). Subsequent echocardiography 6 days after admission confirmed asymmetric septal LVH (17 mm), significant LVOT obstruction (**Figure 4**), with resolution of the regional wall motion abnormalities and systolic dysfunction (LVEF 75%) (Video 4). The findings were reported to be in keeping with hypertrophic cardiomyopathy (HCM).

DIFFERENTIAL DIAGNOSIS

The initial differential diagnosis included causes of acute coronary syndrome. After invasive coronary angiography revealed unobstructed coronary arteries and aortic dissection was excluded, differentials were narrowed down to causes of myocardial infarction with non-obstructive coronary arteries (MINOCA) but additionally takotsubo cardiomyopathy, myocarditis, and HCM.

Given the atypical presentation, a cardiovascular magnetic resonance (CMR) scan was undertaken on day 7 and showed good LV systolic function (LVEF 58%) and asymmetric LVH (septum 17 mm vs lateral wall 7 mm) (Video 5), with chordal SAM resulting in a nonsignificant LVOT obstruction (Video 6). Native T1 and T2 mapping showed increased values (**Figure 5**). Late gadolinium enhancement images revealed focal linear midwall fibrosis in the basal to mid-septum (**Figures 5 and 6**), which was atypical for takotsubo cardiomyopathy and more in keeping with myocarditis.

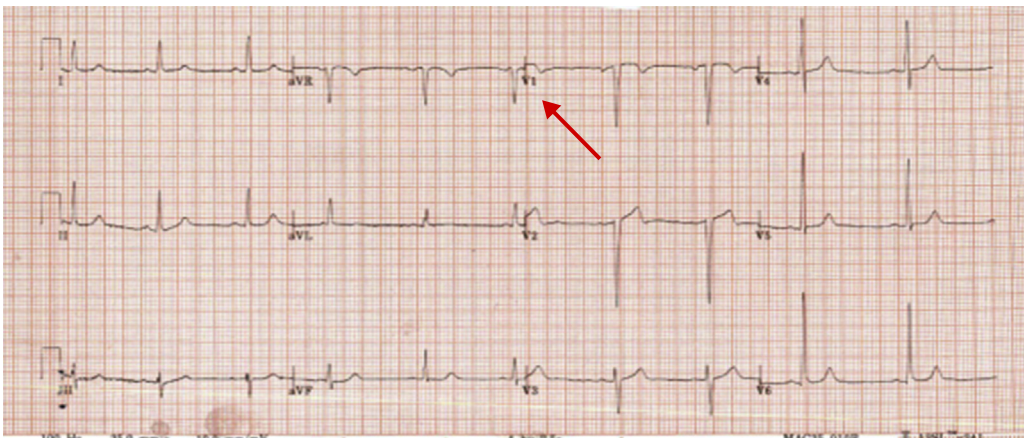
As part of aortic root surveillance, the patient underwent non-contrast CMR 26 months before the acute scan (scan 1) showing normal septal wall thickness of 10 to 11 mm (**Figure 7**) and thickening of the mitral valve tips with chordal SAM and flow acceleration in the LVOT (Video 7).

The CMR findings raised the suspicion of acute myocarditis. The asymmetric septal hypertrophy was assumed to be caused by acute edema rather than by HCM. Follow-up CMR was requested to confirm the final diagnosis.

MANAGEMENT

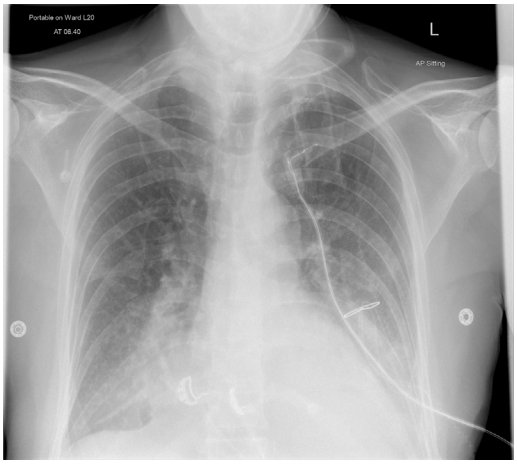
Initial management included dual antiplatelet therapy and fondaparinux for presumed acute coronary syndrome, which were quickly discontinued after normal coronary angiography results. Intravenous diuretic agents and continuous positive airway

FIGURE 1 12-Lead Admission Electrocardiogram



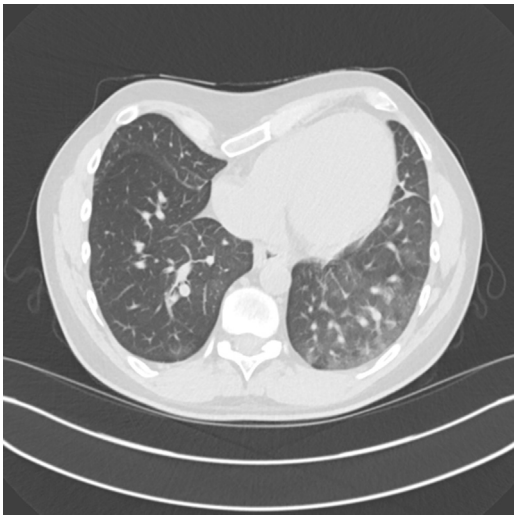
Admission electrocardiogram showing left ventricular hypertrophy and subtle ST-segment elevation and T-wave inversion in V_1.

FIGURE 2 Anteroposterior Chest X-ray



Anteroposterior chest x-ray showing extensive consolidation within the left base, complete effacement of the left hemidiaphragm, and patchy opacification within the medial aspect of the right base.

FIGURE 3 Computed Tomography Aortography



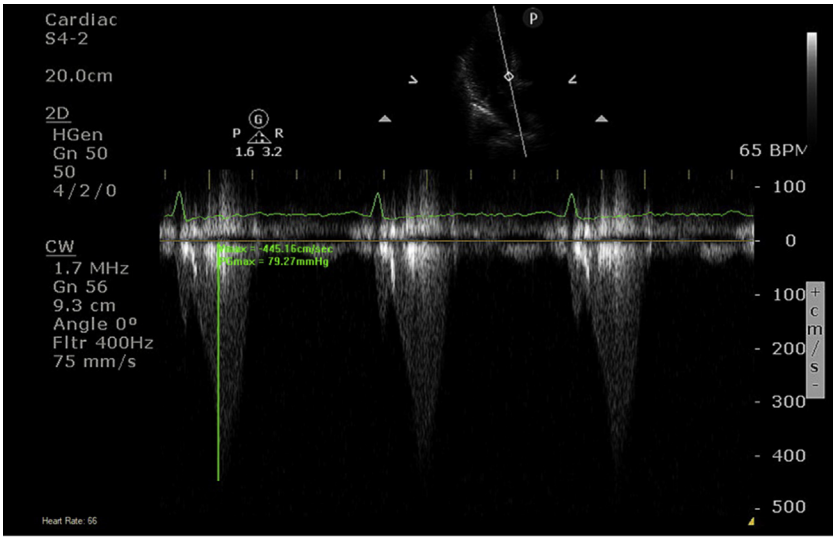
Transaxial computed tomography angiography slice showing nonspecific patchy peripheral ground-glass opacification in the left lower lobe, indeterminate for infection.

pressure were administered for pulmonary edema, and broad-spectrum intravenous antibiotic agents were administered for pneumonia. The patient made a quick recovery, with normalization of his LV function, and was discharged with a regimen of oral furosemide and bisoprolol.

FOLLOW-UP

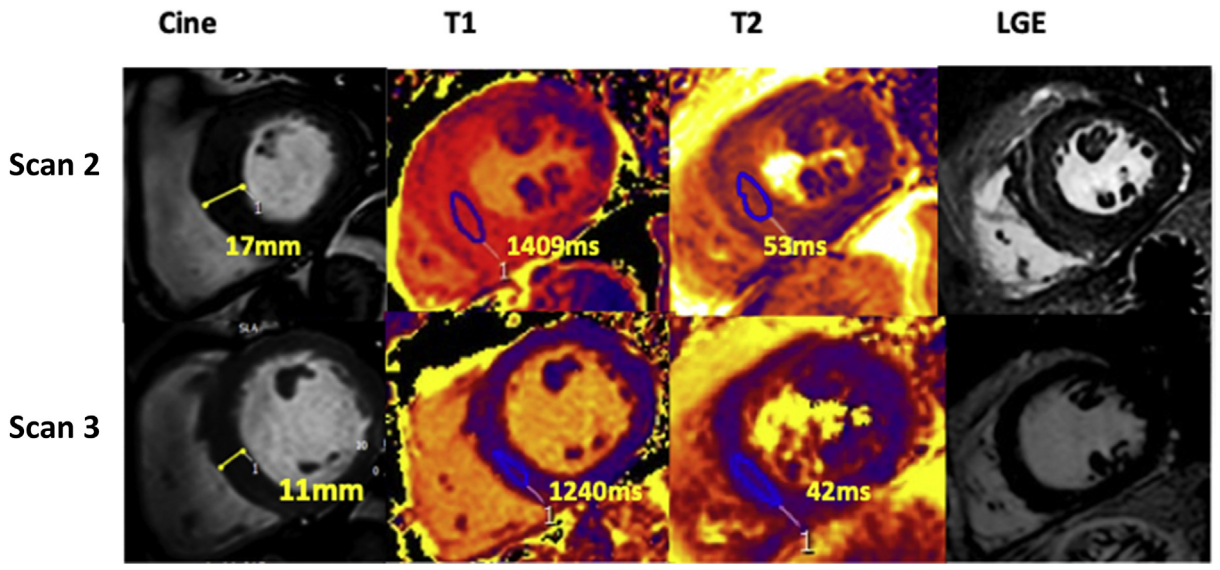
The patient was followed up in the cardiology clinic 6 weeks after discharge. Interval CMR imaging 4 months later showed significant reduction in the

FIGURE 4 Transthoracic Echocardiogram



Continuous-wave Doppler through the left ventricular outflow tract (maximum velocity 4.5 m/s, peak gradient 79 mm Hg).

FIGURE 5 Cardiac Magnetic Resonance Slices Showing Comparison Between Acute Scan (Scan 2) and Follow-Up (Scan 3)



Septal hypertrophy (17 mm) and edema in scan 2; septal T1 1,409 ms (1,190 ± 50 ms; 3-T Philips), T2 mGRaSE 53 ms (44-51 ms; 3-T Philips); resolution of septal hypertrophy (11 mm) and edema in scan 3; septal T1 1,240 ms (1,190 ± 50 ms; 3-T Philips), T2 mGRaSE 42 ms (44-51 ms). Late gadolinium enhancement shows septal midwall enhancement in scan 2 which has almost resolved by scan 3.

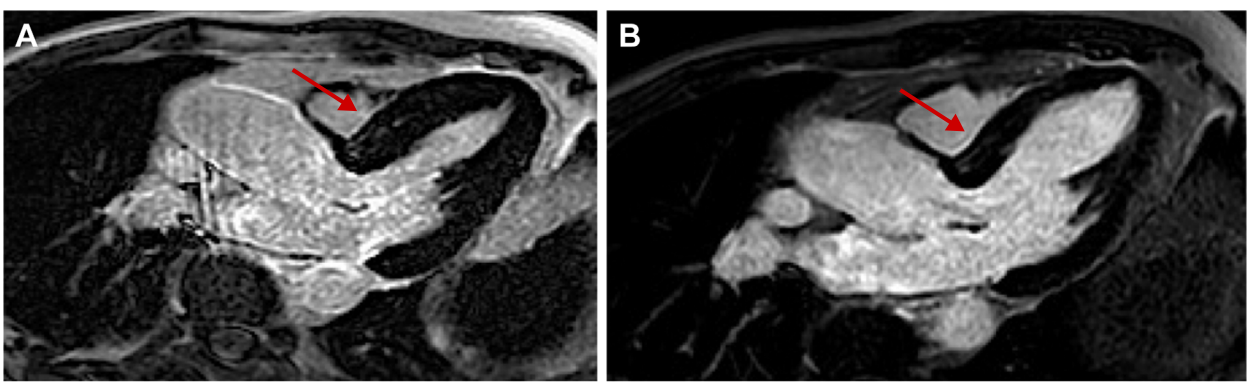
septal hypertrophy (Figure 7) consistent with resolution of the myocardial edema but persistent flow acceleration in the LVOT (Video 8). There was normalization of native T1 and T2 values, with near resolution of the midwall fibrosis (Figure 5).

DISCUSSION

Acute myocarditis is inflammation of the myocardium which commonly results from a viral illness however can be due to non-infectious etiologies. It has a heterogeneous clinical presentation, ranging from minor symptoms to fulminant heart failure, refractory arrhythmias, and cardiogenic shock.

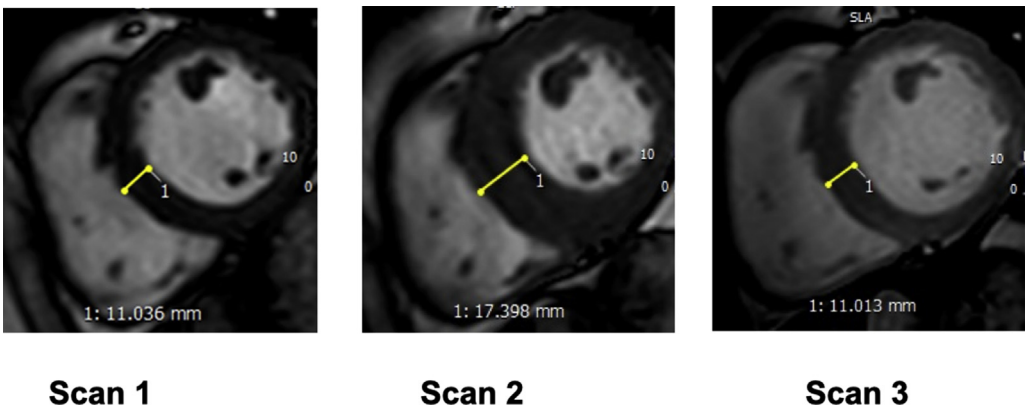
MINOCA is characterized by evidence of myocardial infarction with normal or near-normal coronary arteries on angiography. CMR plays a crucial role in the diagnostic workup for patients with MINOCA.[1] Myocarditis forms the final diagnosis in 25% of MINOCA CMR scans, as do various cardiomyopathies

FIGURE 6 Cardiac Magnetic Resonance 3-Chamber View Late Gadolinium Enhancement Images



Left ventricular outflow tract late gadolinium enhancement slices comparing scan 2 **(A)** showing septal midwall enhancement, which has almost resolved by scan 3 **(B)** 4 months later.

FIGURE 7 Cardiac Magnetic Resonance Short-Axis Cine Slices With Septal Measurements



Left ventricular septal wall thickness at scan 1 (11 mm), scan 2 (17 mm), and scan 3 (11 mm).

such as HCM, which also form 25%.[1] Although endomyocardial biopsy is widely considered to be the criterion standard for the diagnosis of myocarditis (Dallas criteria 1987),[2] CMR has overtaken as a noninvasive diagnostic alternative. The diagnosis of acute myocarditis has moved from being a definitive diagnosis based on histologic evidence to a working diagnosis supported by CMR findings.[3] The Lake Louise Criteria, revised in 2018, suggest a diagnosis of myocarditis in the presence of 1 positive T1-weighted criterion and 1 positive T2-weighted criterion.[4] In our case, native T1 and T2 mapping showed increased values in the septum as evidence of inflammation and myocardial edema, fulfilling the Lake Louise Criteria for a diagnosis of myocarditis.[5] Endomyocardial biopsy was not undertaken because repeated CMR imaging was sufficient to confirm the diagnosis of myocarditis, and the patient made a good and fast clinical recovery.[6]

The surveillance CMR undertaken 26 months prior to the acute admission scan, showed normal septal thickness, supporting the diagnosis of acute inflammation rather than HCM. An increase in wall thickness of 5 to 7 mm in 26 months was considered implausible secondary to HCM, and, as expected in myocarditis, septal hypertrophy resolved on follow-up. The finding of transient LVH in myocarditis has been rarely described in the literature previously,[7,8] and the incidence remains unknown.

CONCLUSIONS

This is a rare and interesting case requiring different imaging tests, over a period of months, to confirm the diagnosis. The unusual presentation of septal edema in myocarditis, compounded with a morphologically abnormal mitral valve, exacerbated the LVOT obstruction. Repeated CMR imaging showed complete resolution of the septal hypertrophy and edema with normalization of the T1 and T2 relaxation times, allowing a confident diagnosis of myocarditis and differentiation from HCM.

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KEY WORDS cardiac magnetic resonance, echocardiography, imaging, mitral valve

APPENDIX For supplemental videos, please see the online version of this paper.