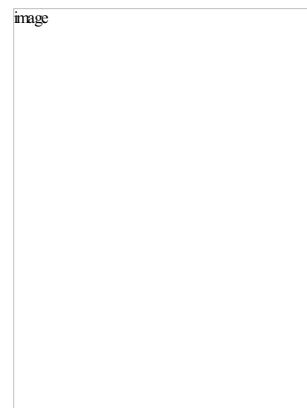


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Mid-cavity variant Takotsubo cardiomyopathy – MRI findings

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Introduction

Takotsubo cardiomyopathy (TCM) is a rare acute coronary syndrome characterized by transient left ventricular dysfunction. Typical findings include apical and mid-segment wall motion abnormalities in a non-vascular distribution¹. The relative compensatory hypercontractility of the basal segment may cause the heart to take on the appearance of a Japanese octopus trap for which the condition is named². Other patterns of involvement have been described, including mid-cavity variant TCM, where wall motion abnormalities are isolated to the mid-cavity with apical sparing^{3,4}. Cardiac magnetic resonance imaging (MRI) has been demonstrated to be an effective modality for the diagnosis and follow-up of TCM in a number of case reports³⁻⁵. Here we present an example of mid-cavity variant TCM with detailed cardiac MRI findings including the use of T2 myomaps, the first case report of its kind.

Case Report

A 56-year-old female presented to the emergency department with chest pain. Hashimoto's thyroid disease was the patient's only significant medical history. An ECG performed on presentation demonstrated no ST-segment elevation or conduction abnormalities however, evolving anterior biphasic T-waves were noted. Laboratory investigations revealed a raised high sensitivity (hs) Troponin T of 181 ng/L (normal <14 ng/L), a normal full blood count and a normal urea, creatinine, and electrolyte panel. There were no clinical features to suggest myocarditis with no history of fever or recent infection noted.

The patient was urgently transferred to the cardiac catheter lab and a coronary angiogram and left ventriculogram (LV gram) were performed. Findings revealed 50 – 60% stenosis of the left anterior descending artery (LAD) but otherwise angiographically normal coronary vessels. The LV gram demonstrated mid-anterior and mid-inferior wall hypokinesis consistent with mid-cavity variant Takotsubo cardiomyopathy (Figure 1). Echocardiography performed the next day demonstrated normal LV size with subtle mid-wall hypokinesis in the mid-inferior and mid-inferoseptal segments (Figure 2). Ejection fraction was 52% (normal $\geq 55\%$) and there was normal diastolic function and LV wall thickness seen. These clinical investigations and history were suggestive of TCM which was subsequently confirmed with cardiac MRI performed on a 3T scanner (Skyra, Siemens, Erlangen Germany).

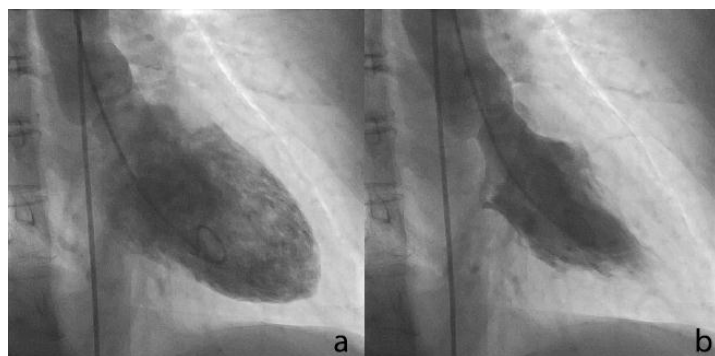


Figure 1: Left ventriculogram in diastolic (a) and systolic (b) phases demonstrating normal left ventricular size and overall preserved left ventricular function. The mid-anterior and mid-inferior walls are hypokinetic consistent with mid-variant TCM. LVEDP was 15 mmHg with no gradient across the aortic valve.

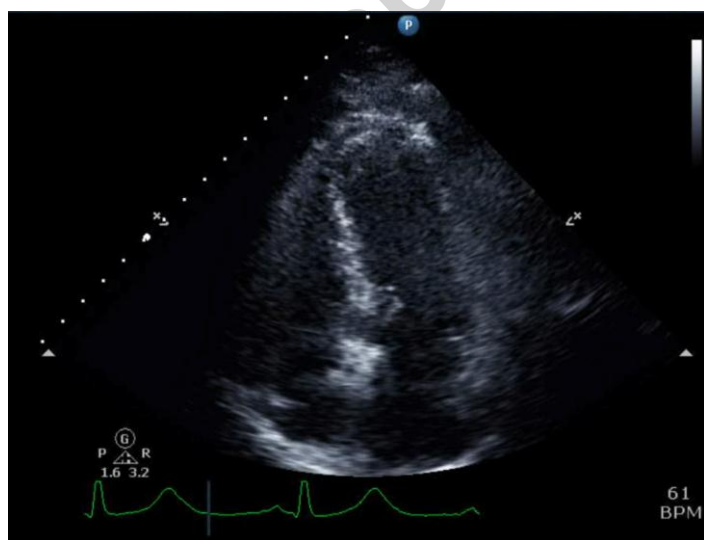


Figure 2: Still image of a four chamber view from the patient's echocardiogram demonstrating mild regional systolic dysfunction and hypokinesia of the mid-inferoseptal segment.

Cardiac MRI demonstrated the failure of contraction or thickening of the mid-ventricular septum, resulting in a focal regional wall motion abnormality that gave the appearance of “ballooning” of the mid-septum towards the right ventricle during contraction (Figure 3). This occurred in the absence of myocardial enhancement, excluding septal infarction as a possible cause for the regional wall motion abnormality. Black blood STIR imaging however, demonstrated high signal in keeping with focal myocardial oedema in this segment (Figure 4). Novel T1 and T2 mapping was performed using Modified Look-Locker Imaging (MOLLI) in addition to these traditional MRI sequences. T2 myomaps supported the presence of myocardial oedema with elevated values in the mid-septum of 53.1 msec (normal for 3T 40 msec) (Figure 5). Upon confirmation of the diagnosis of TCM, patient was commenced on heart failure medications and made a full and uneventful recovery on the ward.

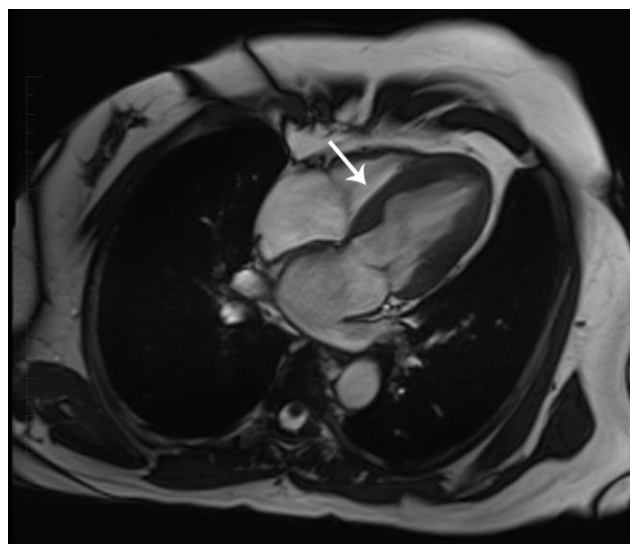


Figure 3: CINE MRI four chamber view demonstrates the appearance of focal ballooning of the mid-cavity interventricular septum (marked with an arrow). This appearance is caused by failure of the mid-septum to contract and thicken during systole.

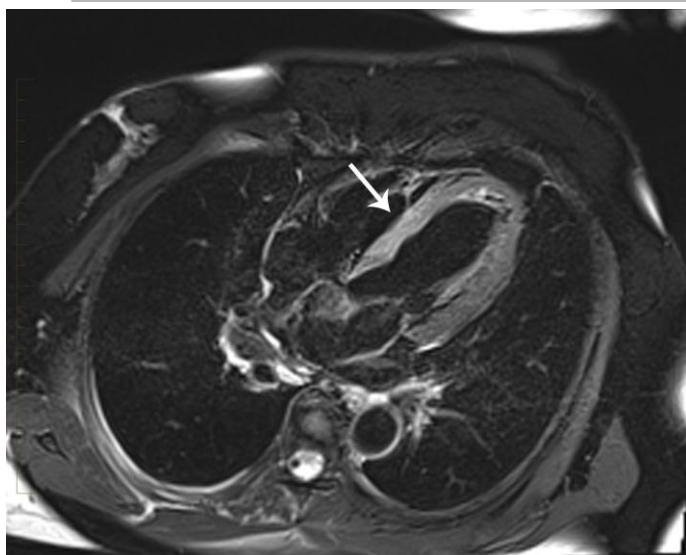


Figure 4: T2 black blood STIR demonstrates high signal in the mid-interventricular septum in keeping with focal myocardial oedema (arrow).

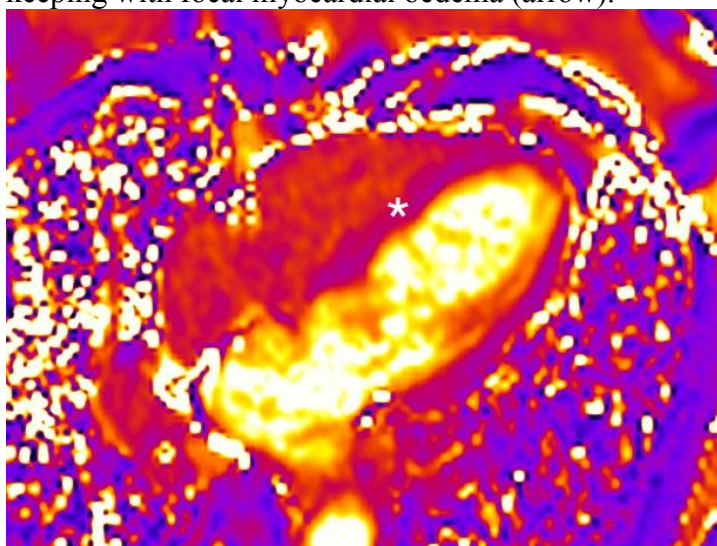


Figure 5: T2 myomaps supports the presence of oedema with T2 values raised to 53.1 (star) in the mid-septum (segment 4) compared to the unaffected left ventricular free wall which demonstrates normal values for 3T of 42. T2 myomaps allows for quantification of oedema.

Discussion

TCM typically presents in post-menopausal women and is often preceded by an acute physical or emotional stressor, leading to its colloquial name ‘Broken Heart Syndrome’⁶. Distinguishing between TCM and acute myocardial infarction can be challenging as both conditions frequently present with severe chest pain and dyspnea⁷. Likewise, both conditions may be associated with ST-segment elevation and T-wave changes on ECG, and a rise in cardiac Troponins¹. Echocardiography routinely plays a key role in diagnosis of TCM with typical findings demonstrating regional wall motion abnormalities out of the distribution of a single coronary artery¹. Unlike myocardial infarction, angiography demonstrates no flow limiting stenosis. Angiographic assessment of the left ventricle demonstrates transient apical left ventricular dilation and compensatory basal hyper-contractility⁸. Another condition which must be considered in patients with normal angiography and regional wall motion abnormalities is myocarditis⁹. Cardiac MRI allows differentiation between myocarditis and

TCM as delayed hyper-enhancement is often present in myocarditis and the pattern of myocardial involvement is different from that seen in TCM⁴. Treatment of TCM is supportive in nature and most patients make a full recovery within a few days or weeks. Recurrence is rare with an annual recurrence rate of approximately 1.5%¹⁰.

Although mid-cavity variant TCM has a different distribution of wall motion abnormalities, its clinical presentation and course is similar to classical TCM. One case series of 17 patients found no difference in clinical characteristics of patients suffering from variant TCM and classical TCM apart from higher ejection fraction on presentation in the variant group⁴. The pathophysiology of TCM is poorly understood however, it is speculated that variations in the distribution of the affected myocardium may be related to variations in autonomic innervation and/or the distribution of adrenergic receptors in the heart⁴.

Case reports of MRI findings in TCM are rare in the radiology literature as compared to the cardiology literature. This is despite the fact that cardiac MRI offers a non-invasive method that can differentiate between conditions such as myocarditis, myocardial infarction and TCM as well as identifying the extent of the disease⁵. Cardiac MRI also has clinical utility in identifying complications of TCM that may be missed on other imaging modalities such as dynamic obstruction of LV outflow and LV thrombus formation¹¹.

Conclusion

Cardiac MRI is an effective non-invasive modality for diagnosing mid-cavity variant Takotsubo cardiomyopathy. Characteristic findings of this variant of TCM include diffuse LV wall oedema isolated to the mid-septum, best appreciated on T2 STIR imaging and confirmed on T2 myomaps which allows for quantification of edema. The associated hypokinesis of the same segment is best appreciated on 4-chamber view CINE MRI. The absence of post-contrast enhancement in the affected segment confirms TCM as opposed to myocardial infarction or myocarditis.

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