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Case Report

Noninvasive Diagnostic Approach to a Rare Variant of Takotsubo Syndrome: From ESC Guidelines to Real World

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Abstract

Takotsubo syndrome is found in about 5%–6% of patients presenting with suspected ST-elevation myocardial infarction. Coronary angiography with left ventriculography is currently considered the modality of choice for the diagnosis. However, improvements of noninvasive diagnostic techniques have been allowing for definite assessment of ventricular function and anatomy. In this setting, the combined use of coronary computed tomography angiography and cardiac magnetic resonance may play a pivotal role for a complete noninvasive diagnosis and management of these patients. We present a case of a 52-year-old woman who presented to our department complaining chest pain and showing left ventricular systolic dysfunction, electrocardiography abnormalities, and mild elevation of cardiac-specific serum enzymes.

Keywords: Cardiac magnetic resonance, coronary computed tomography angiography, multimodality imaging, stress cardiomyopathy, Takotsubo syndrome

Introduction

Takotsubo syndrome (TTS) affects about 5%–6% of patients presenting with suspected ST-elevation myocardial infarction.[1] Coronary angiography with left ventriculography is currently considered the modality of choice for the diagnosis. However, improvements of noninvasive diagnostic techniques have been allowing for definite assessment of ventricular function and anatomy. In this setting, the combined use of coronary computed tomography angiography (CCTA) and cardiac magnetic resonance (CMR) may play a pivotal role for noninvasive diagnosis and management of patients with TTS, even in case of atypical presentation.

Case Report

A 52-year-old female smoker presented at the emergency department with acute chest pain began after vigorous exercise session. She had no family history of cardiovascular disease and metabolic disorders.

On physical examination, she had regular cardiac rhythm (HR: 80 bpm), clear lungs, and no heart murmurs at auscultation. Her blood pressure was of 115/75 mmHg, with oxygen saturation of 98% on room air.

Laboratory tests revealed hemoglobin of 14.8 gr % (normal range: 12–16), mild increase of cardiac-specific serum enzymes (troponin-I: 1.59 ng/mL; normal value: <0.05 ng/mL), myoglobin (83 ng/ml; normal range: 0–70), and creatine kinase-MB isoenzyme (13 U/L; normal value <10 U/L). Creatine phosphokinase concentration was normal (54 U/L; normal value <200 U/L).

Electrocardiography (ECG) showed that ST elevation in precordial leads from V1 to V5 with normal QTC interval. Transthoracic echocardiography was limited by the presence of a poor acoustic window, mainly due to the presence

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of breast implants. It revealed mild reduction of ejection fraction (EF: 45%) with akinesia of midventricular segments.

Because of her low clinical likelihood of coronary artery disease (CAD), the patient underwent ECG-gated CCTA, which showed no evidence of calcified plaques or coronary stenosis [Figure 1], with a reported CAD-RADS of 1.[2,3]

Subsequently, she underwent CMR on a 1.5 T scanner (Achieva, Philips, Best, The Netherlands). CMR protocol entailed T2-weighted short-time inversion recovery (STIR) sequences, balanced steady-state free precession (bSSFP) sequences, and phase-sensitive inversion recovery (PSIR) sequences performed 10–15 min after contrast injection for late gadolinium enhancement (LGE) imaging.[4]

On bSSFP sequences, CMR confirmed a moderate reduction of systolic function (EF: 40%) and circumferential akinesia of midventricular segments with sparing of apical portions [Figure 2a and b and Supplemental Video 1]. T2-weighted STIR sequences showed increased myocardial signal intensity (T2 ratio: 2.3; normal value: <2), suggestive of myocardial edema [Figure 2c]. LGE imaging did not reveal any myocardial scar [Figure 2d].

The provisional diagnosis of an atypical variant of TTS (midventricular type) was made. The patient underwent medical management with prescription of enalapril and bisoprolol. She remained asymptomatic with resolution of ECG changes and normalization of cardiac-specific serum enzymes and was discharged 2 days after.

Follow-up CMR at 3 months showed complete recovery of systolic function (EF: 65%) along with resolution of midventricular circumferential wall motion abnormalities [Figure 3a and b and Supplemental Video 2] and myocardial edema (T2 ratio: 1.8; normal value: <2) and without evidence of myocardial scarring [Figure 3c and d].

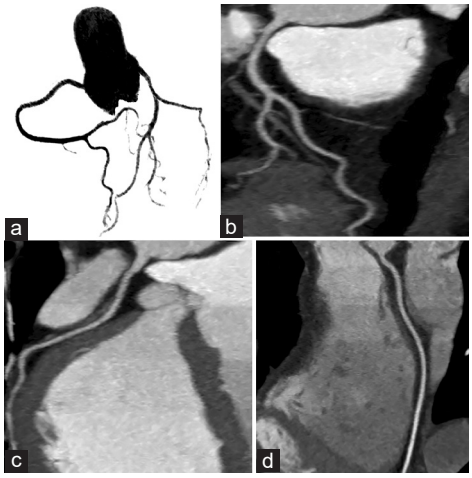

Figure 1: Coronary computed tomography angiography performed at patient's hospital admission, showing no evidence of coronary stenosis (coronary artery disease-RADS 1). Volume rendering with maximum intensity projection image of coronary vessels (a), curved maximum intensity projection reformatted images of left circumflex artery (b), left anterior descending (c), and right coronary artery (d)

Discussion

TTS syndrome is classically characterized by transient left ventricular systolic dysfunction, ECG abnormalities, and elevation of cardiac-specific serum enzymes that can be caused by a variety of physical or emotional triggers.[1]

Also known as "happy or broken heart syndrome," "stress cardiomyopathy," "apical ballooning syndrome," and "ampulla cardiomyopathy," its diagnosis is often challenging since symptoms are similar to acute myocardial infarction (i.e. acute chest pain, dyspnea, or syncope).[1]

Several pathophysiological mechanisms have been proposed, including plaque disruption, multivessel spasm, baroreflex abnormalities, and catecholamine surge, with proof of evidence confirming this latter to play a key role in the myocardial injury.[5]

Typically considered as a mimicker of acute coronary syndrome (ACS), it is characterized by no evidence of obstructive CAD at coronary angiography. Despite ventriculography is considered the current gold standard for diagnosis, noninvasive diagnostic approach has been suggested.[6]

CCTA has been recently proposed as a valid alternative to invasive angiography to rule out ACS in case of low-to-intermediate likelihood of CAD or when cardiac troponin levels with or without ECG are normal or inconclusive.[6]

Moreover, the current recommendations suggest to perform CMR in patients with nonobstructive coronary arteries on

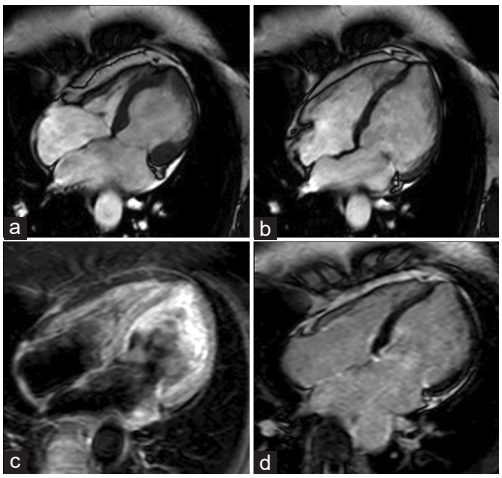

Figure 2: Cardiac magnetic resonance imaging performed at patient's hospital admission. Balanced steady-state free precession cine-images performed along the four-chamber view in end-systole (a) and end-diastole (b) show circumferential akinesia of midventricular segments. T2-weighted short-time inversion recovery (c) and T1-weighted phase-sensitive inversion recovery images (d) performed along the same plane, respectively, showing diffuse increase of myocardial signal and no evidence of late gadolinium enhancement

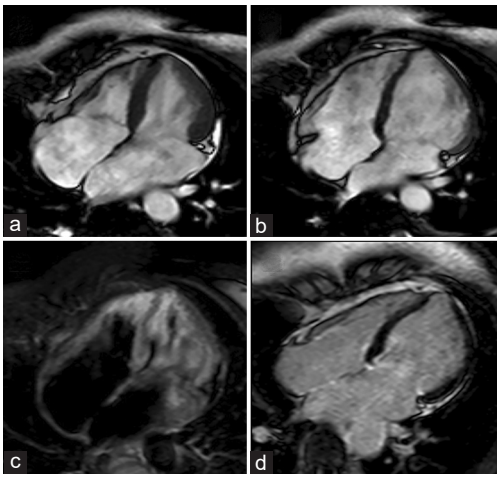

Figure 3: Cardiac magnetic resonance imaging performed at 3-month follow-up. Cine images performed along the four-chamber view in end-systole (a) and end-diastole (b) show complete recovery of midventricular akinesia. T2-weighted short-time inversion recovery (c) and T1-weighted phase-sensitive inversion recovery images (d) performed along the same plane, respectively, showing normalization of myocardial signal and no evidence of late gadolinium enhancement

angiography (i.e. no vessel stenosis >50% in a major epicardial vessel) and without obvious underlying causes.[6,7]

In our case, the combined CCTA and CMR imaging approach has allowed for a complete noninvasive diagnosis of TTS, confirmed at 3-month follow-up.

CCTA showed no evidence of moderate or severe coronary stenosis. On the other hand, CMR depicted noncoronary distribution of wall motion abnormalities. In addition, CMR allowed to show myocardial edema without evidence of scarring, thanks to its ability to provide noninvasive tissue characterization.[6]

In conclusion, a complete noninvasive diagnostic approach in patients with low-intermediate risk of CAD is feasible, even for the diagnosis of atypical and less frequent variants of TTS.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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