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MINI-FOCUS ISSUE ON CARDIOMYOPATHIES AND GENETIC COUNSELING

INTERMEDIATE

CASE REPORT: CLINICAL CASE

An Exceptional Cause of Acute Right Heart Failure

Isolated Right Ventricular Takotsubo Syndrome

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ABSTRACT

We describe a patient with of acute right ventricular dysfunction secondary to right ventricular isolated Takotsubo syndrome (TTS). The importance of appropriate differential diagnosis for acute right ventricular dysfunction differential diagnosis of acute right ventricular dysfunction and the differences in diagnosis and management of right ventricular TTS and typical left ventricular TTS are highlighted. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2020;2:365–9) © 2020 The Authors. 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/).

HISTORY OF PRESENTATION

An 82-year-old female patient presented to the emergency department with nonspecific discomfort and in poor general condition. Physical examination demonstrated normal chest and cardiac auscultation. The patient showed signs of cardiogenic shock and respiratory failure: blood pressure 90/60 mm Hg, 110 beats/min, arterial oxygen saturation 80%, and lactate levels 7.3 mmol/l. An electrocardiogram (ECG) indicated sinus rhythm with no repolarization abnormalities. Right-sided ECG showed nondeep T waves in leads V_3R to V_6R (**Figure 1**). Transthoracic echocardiography revealed normal left ventricular (LV) systolic function but dilation and severe systolic dysfunction of the right ventricle (RV) with apical dyskinesia and basal hypercontractility, also known as the reverse McConnell sign (1).

LEARNING OBJECTIVES

- To review the differential diagnosis of acute RV dysfunction and highlight the importance of the clinical history.
- To understand the differences in clinical presentation and evolution of RV TTS and classic left ventricular TTS.
- Phosphodiesterase-3 inhibitors could be useful in cases of TTS complicated with cardiogenic shock.

PAST MEDICAL HISTORY

The patient had a chronic depressive affective disorder. Four days earlier, she had suffered a Colles fracture and coccyx fracture with persistent low back pain in the following days.

DIFFERENTIAL DIAGNOSIS

Taking into consideration the initial presentation with respiratory failure and RV dysfunction the initial

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ABBREVIATIONS AND ACRONYMS

AMI = acute myocardial infarction

ECG = electrocardiogram

LV = left ventricle

MRI = magnetic resonance imaging

RV = right ventricle

TTS = Takotsubo syndrome

differential diagnosis should include acute pulmonary embolism and acute myocardial infarction (AMI).

INVESTIGATIONS

Chest computed tomographic scan ruled out pulmonary embolism as the first suspected etiology. High sensitivity troponin T and creatine kinase levels were markedly elevated, with significant dynamic change (high sensitivity troponin T: 469 to 556 to 368 ng/dl; creatine kinase: 354 to 516 to 202 U/l), as was the N-terminal pro-B-type natriuretic peptide level (9,116 ng/l). Coronary angiography performed to rule out RV AMI showed normal coronary arteries (**Figure 2**). Right ventriculography confirmed mid-apical dyskinesia (Video 1).

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MANAGEMENT

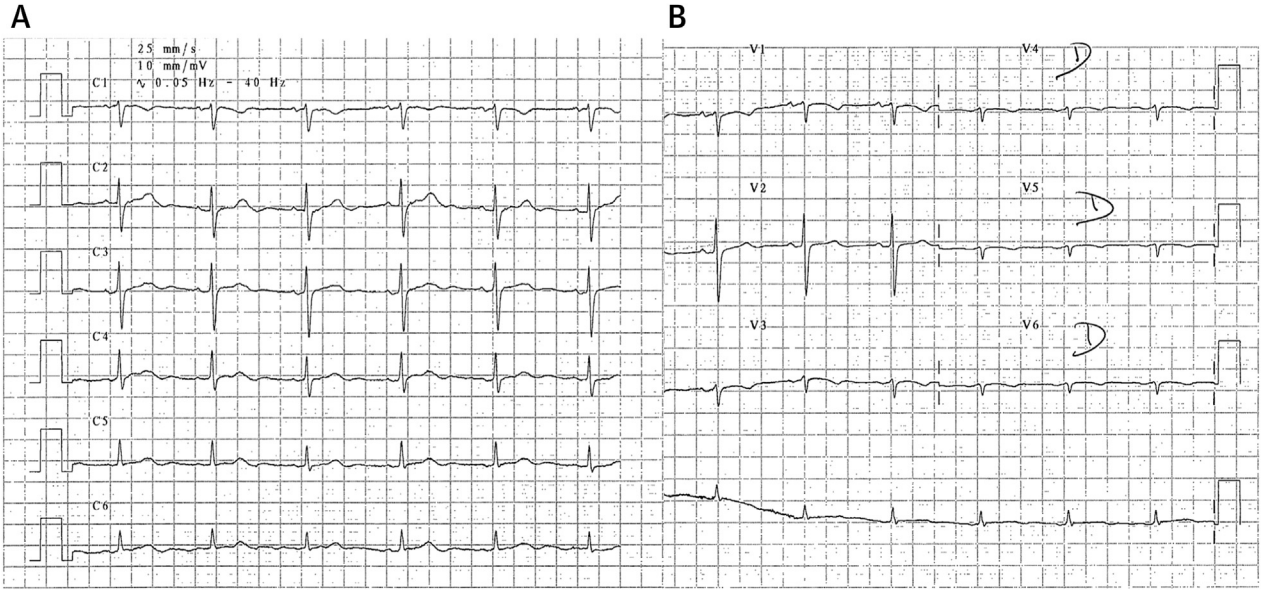
The patient was admitted to the coronary care unit. Oxygen therapy and administration of milrinone were initiated, with hemodynamic improvement. A pulmonary artery catheter was implanted and revealed pulmonary artery pressures of 34/14 mm Hg, central venous pressure 9 mm Hg, and cardiac index 2.5 l/min/m^2. In the following 24 h, the patient demonstrated progressive clinical improvement with resolution of respiratory failure and hemodynamic stability, allowing the withdrawal of inotropic support. Evolutionary ECG on day 2 showed the appearance of negative T waves in leads V_1 to V_4 with a slight QT-interval prolongation (**Figure 3**).

Cardiac magnetic resonance imaging (MRI) performed on day 2 showed a nonhypertrophic but dilated RV with dyskinetic movement of the apex and midanterior wall segments, conditioning mild RV systolic dysfunction (**Figure 2**, Video 2). Sequences to evaluate for the presence of myocardial edema could not be performed because the patient had low back pain due to prolonged recumbency.

On day 3, the patient was transferred to the cardiology ward. Control transthoracic echocardiography and cardiac MRI (days 7 and 10 after admission) showed normalization of volumes and regional/global RV systolic function without late gadolinium enhancement (**Figure 2**, Video 3).

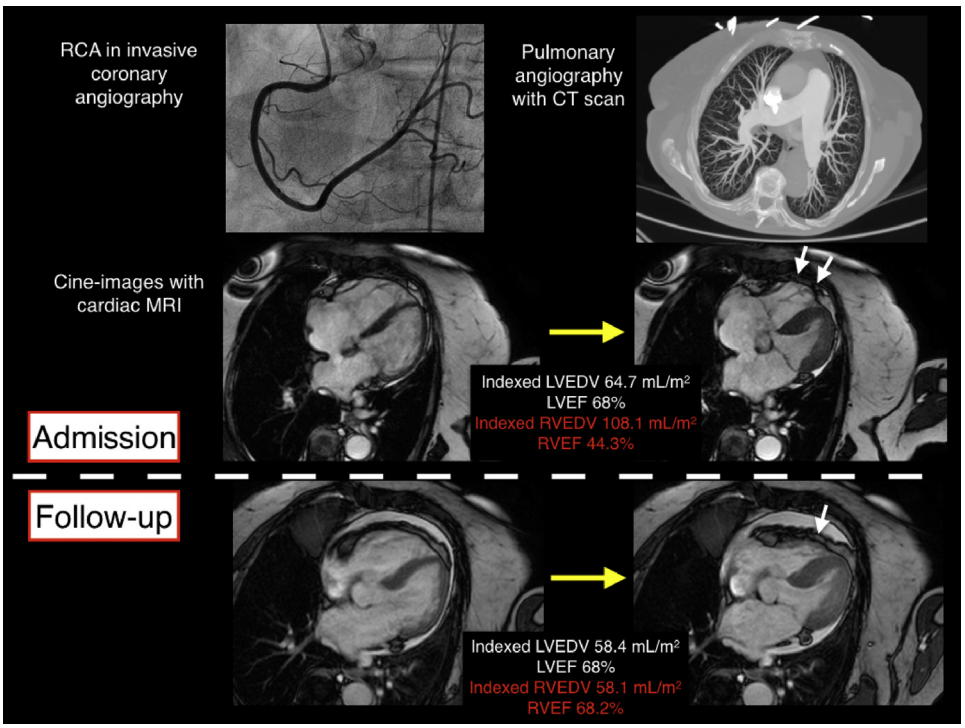
After pulmonary embolism, acute coronary syndrome, and other pathologies associated with transient acute RV dysfunction (e.g., acute myocarditis) were ruled out and considering the typical clinical scenario of a post-menopausal woman with a

FIGURE 1 Initial ECG



(A) ECG at admission showing precordial leads with no repolarization abnormalities. **(B)** Right-sided precordial leads **(right)** showing nondeep negative T waves in leads V_3R to V_6R. ECG = electrocardiogram.

FIGURE 2 **Right Coronary Angiography, Chest CT Scan, and Cardiac MRI**



(Top row) Angiography **(left)** showing normal right coronary artery (RCA) with no presence of lesions and chest computed tomographic (CT) scan **(right)** showing no evidence of pulmonary thromboembolism and normal diameter pulmonary trunk. **(Middle, bottom rows)** Four-chamber cine images by cardiac magnetic resonance imaging (MRI) in diastole **(left)** and systole **(right)** with dilated right ventricle (RV) and dyskinesia of the RV free wall at admission and recovery of the volumes and contractility of the RV at cardiac MRI performed 10 days after. LVEDV = left ventricular end-diastolic volume; LVEF = left ventricular ejection fraction; RVEDV = right ventricular end-diastolic volume; RVEF = right ventricular ejection fraction.

previous stressful situation and a compatible ECG evolution, the diagnosis of Takotsubo syndrome (TTS) with isolated RV involvement was finally established.

DISCUSSION

TTS, also known as stress cardiomyopathy, is an acute heart failure syndrome that usually affects post-menopausal women with a recent physical or psychological stressor. Unlike AMI, TTS is characterized by the absence of significant coronary lesions and the reversibility of cardiac dysfunction. However, because of the similar acute clinical presentations, it often triggers a differential diagnosis with AMI. TTS is increasingly recognized and reported, probably because of better knowledge of the disease and its particular characteristics, but the precise incidence is unknown (2,3). The apical and midregional systolic defect is the most frequent form of LV regional wall motion abnormality. It is classified as the typical anatomic variant of TTS, but other atypical forms have been described. Nevertheless, the isolated involvement of the RV is exceptional, and few cases have been reported to date (4,5), so this variant has not yet been included as a type of TTS in expert consensus (2,3). Unlike classic forms of TTS affecting the LV, some specific characteristics present in this case that we found in common with previously reported cases of isolated RV TTS are: 1) development of nondeep negative T waves in leads V_1 to V_4 on the evolutionary ECGs, in opposition to the deep and predominantly anterolateral negative T waves generally observed in the typical form of TTS; 2) dilation of the affected ventricle, which is not commonly observed in the LV forms of TTS, with characteristic systolic impairment resembling the reverse McConnell sign (i.e., apical dyskinesia and hyperkinesia of the basal segments); and 3) frequent severe clinical

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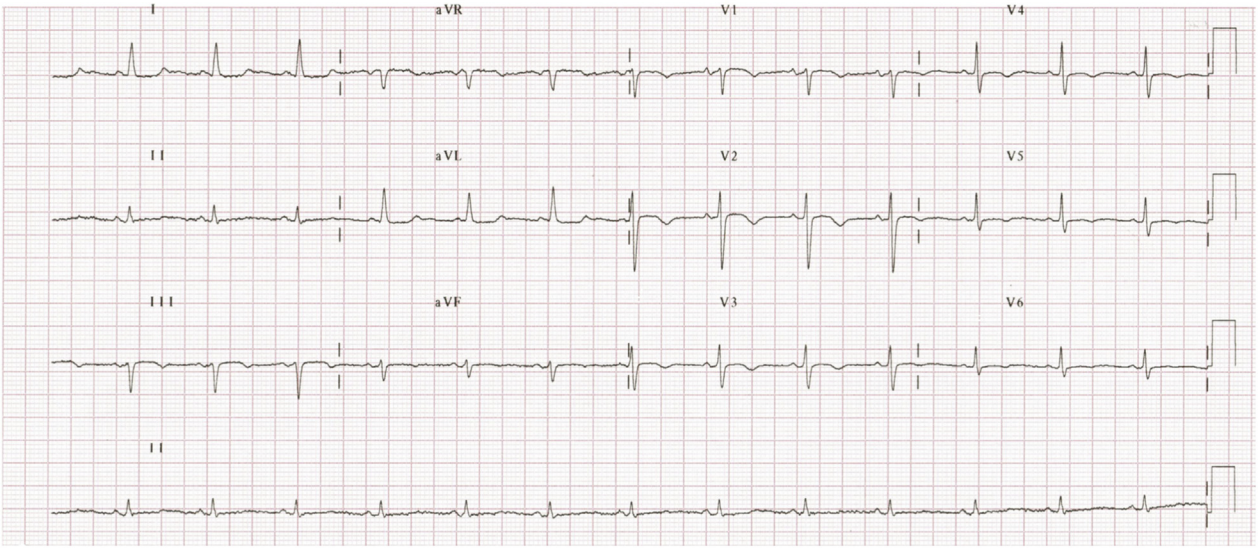
FIGURE 3 Evolutionary ECG



ECG performed on day 2 showing evolutionary negative T waves in leads V_1 to V_4 with slight QT-interval prolongation. ECG = electrocardiogram.

presentation, with a reasonably rapid clinical recovery in accordance with the rapid improvement of RV systolic dysfunction seen in the imaging tests. Interestingly, this recovery seems to occur faster than in patients with LV involvement.

In this case, some particular characteristics were noted. First, the initial ECG showed nonpathological negative T waves limited to lead V_1. The initial ECG can be normal in up to 14% of patients with classic LV TTS (6), and usually these patients develop deep negative T waves over the course of several days. This fact, in conjunction with the low magnitude of ECG changes reflecting RV pathology in the standard leads, could explain the normal ECG upon admission of this patient. Second is the choice of milrinone treatment and its successful response. We decided to administer a phosphodiesterase-3 inhibitor rather than dobutamine to avoid the use of catecholamines. Use of phosphodiesterase-3 inhibitors has been described for treatment of TTS complicated by cardiogenic shock, with successful results (7,8). Dobutamine use has been described as a trigger of TTS (9). Nonetheless, there is a lack of evidence on the benefit of using milrinone over dobutamine in clinical practice.

FOLLOW-UP

Within 2 years follow up with cardiology the patient is in New York Heart Association functional class I. Ambulatory echocardiography demonstrated normal biventricular function with no recurrence of TTS episodes.

CONCLUSIONS

Although very uncommon, RV TTS has to be considered in the differential diagnosis of acute right heart failure. This entity has several specific features that differentiate it from typical LV TTS. Previously reported cases describe a frequent severe clinical presentation, usually cardiogenic shock, with more rapid recovery than cases of LV TTS.

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KEY WORDS cardiac magnetic resonance, cardiogenic shock, right ventricle, stress cardiomyopathy, Takotsubo syndrome

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