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Hypertrophic Obstructive Cardiomyopathy and Takotsubo Syndrome: Could They Coexist?

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Abstract

Introduction: Takotsubo syndrome (TKS) is generally caused by a stressful condition, and it usually has a good prognosis after the recovery of left ventricular function. About 70% of the cases of hypertrophic cardiomyopathy may develop obstruction in the left ventricular outflow tract (LVOT), which is responsible for heart failure.
Case Presentation: We present a unique case where TKS occurred in a middle-aged male patient with hypertrophic obstructive cardiomyopathy (HOCM) without a clearly identifiable initial stress trigger.
Conclusions: In the setting of acute left ventricular function depression in HOCM, a comprehensive differential diagnosis should be established. Treatment should be based on hemodynamic changes. After recovery, the prognosis is related to HOCM.

Keywords: Hypertrophic Cardiomyopathy, Takotsubo Cardiomyopathy, Heart Failure

1. Introduction

Takotsubo syndrome (TKS) can be described as a transient cardiac condition due to an intense stress trigger where heart failure may develop secondary to ventricular dysfunction. Middle-aged women with previous psychological trauma constitute the population primarily affected. TKS can mimic acute myocardial infarction in terms of the characteristics of the chest pain and the findings concerning electrocardiography (ECG) and troponin. It can also lead to heart failure because of depressed ventricular ejection fraction (EF). Once the acute situation is restored, the prognosis is benign (1).

On the other hand, the heart failure manifestations of hypertrophic obstructive cardiomyopathy (HOCM) are fundamentally due to the obstruction of the left ventricular outflow tract (LVOT), while LVEF is preserved (2). Its prognosis is not easy to establish because LV systolic function, LVOT obstruction, and other factors are the prognostic factors throughout the patient's life (2, 3).

We describe a male patient with HOCM who suffered a peculiar presentation of TKS.

2. Case Presentation

A 51-year-old man with dyspnea was diagnosed with HOCM in 2013. Because of the occurrence of syncope, hypotensive response to exercise (despite a maximum wall thickness of 19 mm), and LVOT obstruction gradient, he underwent a cardioverter defibrillator implantation. During the follow-up, several changes in medication were made in order to diminish the obstruction in the LVOT (about 85 mmHg at rest and 150 mmHg after exercise) and to improve the heart failure symptoms; there was, however, no positive response, LVEF was normal.

Clinical progress was finally achieved after the modification of the pacemaker parameters. Alpha-galactosidase activity in plasma was normal.

One month later, the patient suffered chest pain at rest, with irradiation to the left arm and jaw, followed by dizziness. No physical or psychological stress, or high temperature or any factor which may have played the role of a trigger was identified. The first examination showed blood pressure of 102/70 mmHg, normal oxygen saturation, and tachycardia. On auscultation, systolic mesocardial and mitral murmurs (grade III) were present, but there were no crackles in the lungs. The blood test revealed raised troponin T (4.47 nG/mL), renal and liver functions, hemogram, and coagulation, while inflammatory parameters were within normal limits. The first-day ECG was similar to the previous ones (Figure 1A), but on the following day, new deep negative T waves appeared from V3 to V6 and the QT segment was prolonged (Figure 1A-1B). No ventricular or supraventricular arrhythmias were found. The initial

echocardiogram discovered depressed LV systolic function (43% by the Simpson method, LV end-diastolic volume = 40 mL, and LV end-systolic volume = 23 mL) owing to severe anterior, septal, and apical hypokinesia (Figure 2A - 2C) (Appendix 1; videos 1 and 2).

Additionally, the obstruction in the LVOT was reduced to 40 mmHg at rest. A diagnosis of non-ST-elevation acute myocardial infarction was made, and the patient was managed accordingly with dual antiplatelet and anticoagulation. A subsequent angiogram failed to identify any coronary obstruction. The ventriculogram demonstrated a typical LV TKS shape in systole (Figure 2G). Severe mitral valve regurgitation and significant LVOT obstruction (> 100 mm Hg) were also evidenced. In the following 48 hours, the patient developed not only severe heart failure due to severe LV systolic dysfunction (LVEF = 30%) and severe mitral regurgitation, but also signs of severe infection with a suspected origin in the urinary tract. Invasive mechanical ventilation, empiric antibiotic regimen, and hemodynamic support were required. No lung hemorrhage findings were identified with bronchoscopy. All microbiology, virus serology, and autoimmunity parameters were negative except for *E. coli*, found in urine as well as in blood. Transesophageal echocardiography was negative for endocarditis. Gradually, sepsis was controlled and remission commenced. Concurrently, LV contractility progressively returned to normal. The ECG changed toward normalization of the QT segment, although the negative T waves persisted. Once LVEF was restored (LVEF = 71%, end-diastolic LV volume = 41 mL, and end-systolic LV volume = 12 mL) (Figure 2D - 2F)(Appendix1; video 3 and 4), the LVOT obstruction reappeared. Titration with beta-blockers was initiated. In addition, the pacemaker auriculo-ventricular time was reprogrammed so that the ventricles were always under stimulation. No changes in LV wall thickness (interventricular septum = 19 mm and anteroapical septum = 15 mm) or in left atrial volume (18 cm^2) were registered.

At follow-up 15 days later, the patient was in good condition under the comprehensive treatment previously described: LVEF was preserved and LVOT obstruction was as low as only 7 mmHg at rest, with minimal mitral regurgitation.

3. Discussion

TKS is a disease whose diagnosis is made retrospectively once coronary artery disease and other entities such as reperfused myocardial infarction or myocarditis are excluded and wall motion abnormalities have recovered. It should be noted that TKS adopts a typical shape in systole during the acute phase: round-bottomed and narrow-necked. Only a few cases have been hitherto described in patients with a previously diagnosed HOCM, and women still constitute those primarily affected (4-9).

Our case occurred in a male patient with no recognizable initial stressor (4). Nevertheless, what it did have in common with the previously reported cases was its presentation insofar as it mimicked non-ST acute coronary syndrome. We excluded coronary artery obstruction, but such exclusion vis-a-vis other causes was rendered impossible because of the following reasons. Firstly, although cardiac magnetic resonance is exceedingly helpful in patients with chest pain, elevated troponin levels, and non-stenotic coronary arteries to determine the final cause of the entity (10, 11), our patient had a device not compatible with magnetic resonance imaging. Secondly, we could not perform nuclear imaging during the acute phase because of the patient's bad clinical situation. Furthermore, diagnostic profitability after almost 1 month, even after administrating anti-inflammatory and antibiotic treatment, was low. And thirdly, we were not able to plan cardiac biopsy because the LV was the region predominantly affected and the diagnostic yield itself was also poor. However, we made a diagnosis of TKS given the wall motion abnormality in the apical segments, the LV shape, and especially the recovery of LV contractility and function after some days. ECG changes were also in keeping with the diagnosis (12).

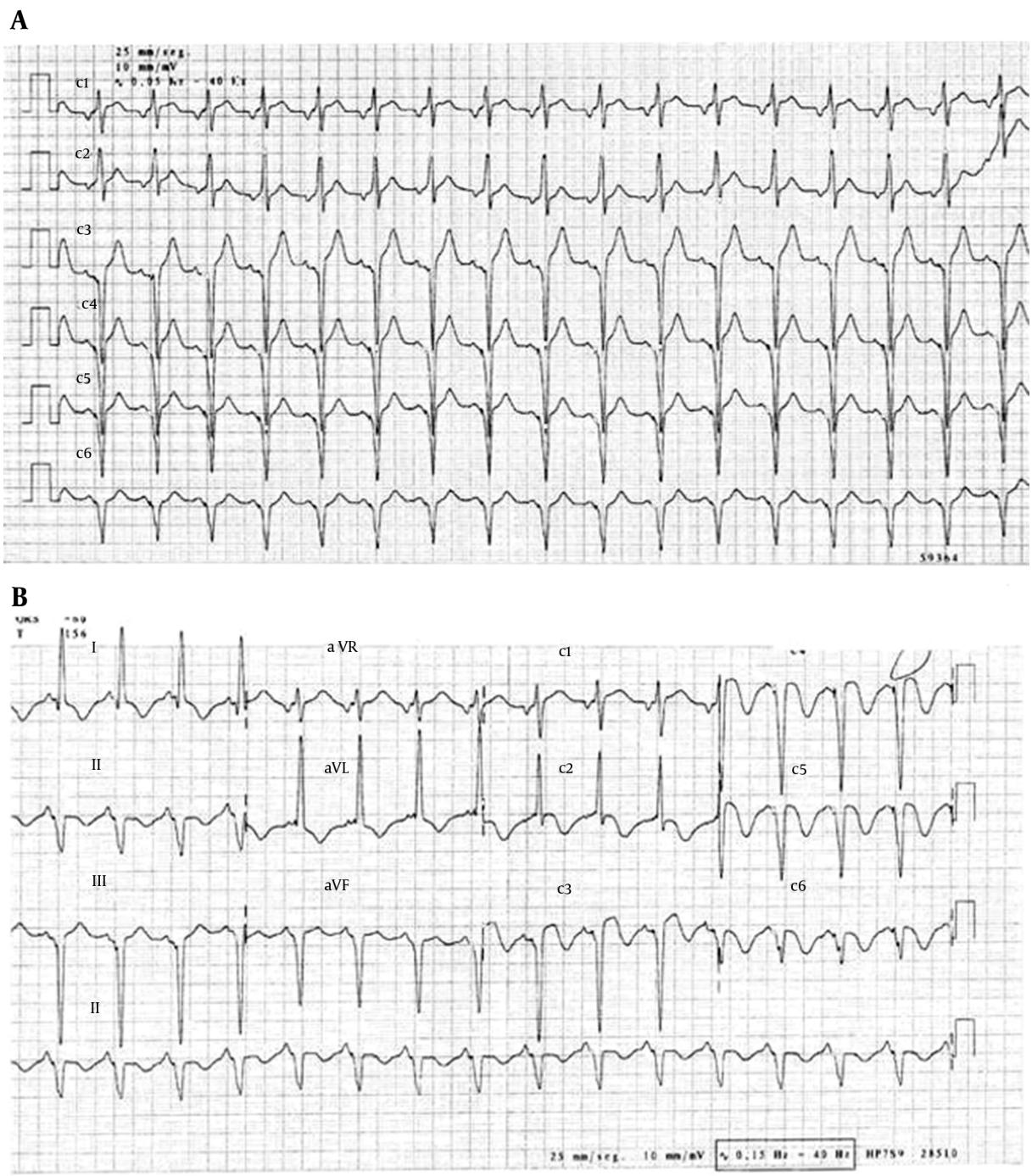
It is remarkable that the transient wall motion abnormality was settled on a pathological hypertrophy: although the maximum wall thickness was at the anteroseptal basal segment, it extended toward the lateral and inferolateral apical segments too. Contrary to other previously reported cases (13, 14) no changes in wall thicknesses during the acute or subacute phases were recognized.

The heart failure condition developed in the days following the patient's admission was due to LV systolic dysfunction. The pressure gradient across the LVOT diminished in the acute phase because of severe hypokinesia, which was not able to create an obstruction to the flow, resulting in a low LV stroke volume. As the contractility turned into normal, the LVOT pressure gradients reappeared.

The role of infective agents in the pathogenicity of TKS has been suggested. Whether the presymptomatic phase of urinary infection might have triggered TKS in our patient remains unclear.

The prognosis of TKS is assumed to be good after the restoration of systolic function, which only rarely relapses (1). The clinical management of the patient is now focused on HOCM symptoms and the prevention of complications.

Figure 1. The ECG



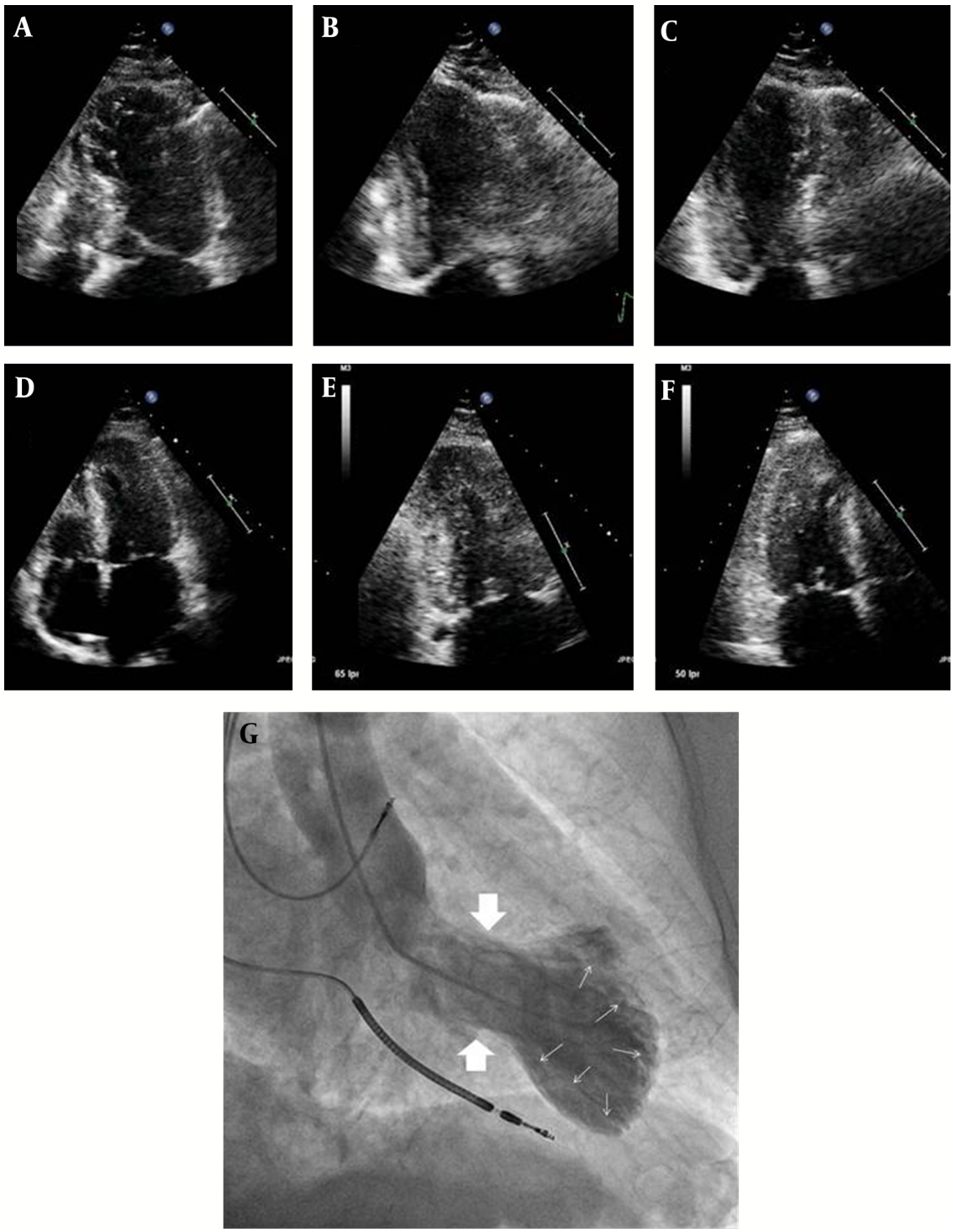
A, Presentation; and B, 48-hour admission ECGs demonstrate ST elevation in A and evolution to diffuse negative T wave in the precordial leads.

Supplements

id=56643

Supplementary material(s) is available at below link: http://cardiovascimaging.com/?page=download&file_-

Figure 2. Left Ventricular Images



A, B, and C, left ventricular end-systolic echocardiographic images during admission show an apical balloon appearance; D, E, and F, similar images after recovery; G, left ventricular angiogram demonstrates a typical Takotsubo appearance in end-systole.

Footnote

Authors' Contribution: Study concept and design, all authors; acquisition of data, all authors; analysis and interpretation of data, all authors; drafting of the manuscript, Pilar Egea-Serrano and Juan R Gimeno; critical revision of the manuscript for important intellectual content, Juan R Gimeno; statistical analysis, no relevant; administrative, technical, and material support, Pilar Egea-Serrano and Juan R Gimeno; study supervision, Pilar Egea-Serrano and Juan R Gimeno.

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