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An unusual case of severe left ventricle outflow tract obstruction due to a coexistence of Takotsubo cardiomyopathy with septal hypertrophic cardiomyopathy

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Abstract

Hypertrophic cardiomyopathy (HCM) is a genetic disorder presenting with a pathological increase of left ventricle (LV) wall thicknesses. The most frequent morphological form is characterized by an abnormal LV basal septal hypertrophy. Takotsubo cardiomyopathy (TTC) is a transient left ventricular systolic dysfunction induced by high physical or emotional stress. Its occurrence with HCM is unusual. However, this presentation occurs more often with the classic asymmetrical septal hypertrophy compared with the apical variant. This case demonstrates that the coexistence of TTC with septal HCM in an elderly patient may lead to a severe hemodynamic instability picture.

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Introduction

Left ventricle (LV) basal septal hypertrophy is the most frequent morphological form of hypertrophic cardiomyopathy (HCM) [1]. Takotsubo is a transient cardiomyopathy related to high physical or emotional stress. It typically presents with substernal chest pain, electrocardiogram changes which include ST elevation, ST depression, T-wave inversion, pathologic Q-waves and QT prolongation, transient left ventricle apical ballooning associated with normal coronary angiogram [2,3]. This disease entity can clinically manifest in patients with HCM but typically occurs more often with the asymmetrical septal hypertrophy in comparison with the apical variant [1-3]. Our case demonstrates this unusual coexistence highlighting that its recognition is critical for the management and treatment, especially in the setting of cardiogenic shock, as inotropic agents are likely to aggravate and worsen the clinical condition.

Case Report

An 81-year-old female presented to the emergency department (ED) complaining of dyspnea and chest pain lasting for one day. She had hypertension and dyslipidemia associated with a familial history of sudden death. On physical exam, we found a severe hypotension (systolic blood pressure of 80 mmHg) associated with bilateral rales at chest auscultation. Cardiac auscultation revealed a harsh systolic murmur, best heard over the left sternal border. Heart rate was 60 bpm in sinus rhythm. Labs were significant for HS-I troponin of 6.035 ng/L (NV: ≤12) and NT-proBNP of 7.640 pg/mL (NV: ≤1800). A 12-leads electrocardiogram (ECG) at admission revealed a STEMI-like ST segment elevation from V2 to V6 (Figure 1A). For this reason, she was urgently taken to the cath-lab where she was found to have tortuous but normal coronary arteries. After coronary angiography, a trans-thoracic echocardiogram (TTE) revealed a pathological LV hypertrophy with a septal diastolic thickness of 19 mm, depressed LV ejection fraction (LVEF) due to a severe apical ballooning. At continous wave (CW)-doppler there

was a dynamic obstruction across the LV outflow tract (LVOT), with a late peak velocity of 4,9 m/s and an estimated peak gradient of 98 mmHg. The gradient was increased by a systolic anterior motion (SAM) of anterior mitral leaflet causing a moderate mitral regurgitation (MR). All these findings were consistent with obstructive septal HCM associated with Takotsubo cardiomyopathy (Figure 1 B,C; Figure 2; Video 1). After treatment with intravenous diuretics and metoprolol (5+5 mg i.v. bolus followed by oral dose of 100 mg daily), her clinical condition markedly improved. One week later, ECG (Figure 1D) demonstrated deeply inverted T waves on antero-lateral leads and QT prolongation (501 ms). Three weeks later, after a complete resolution of the LV apical dyskinesia, LVEF normalized (Figure 1E; Video 2). LVOT gradient decreased to 20 mmHg, with a dynamic increase to 70 mmHg during Valsalva maneuver (Figure 1F and Figure 3). Mitral SAM also disappeared when LV function normalized and peak gradient fell down.

Discussion

TTC, also known as stress cardiomyopathy, transient apical ballooning, or broken heart syndrome, was described by Armstrong *et al*., as the shape of the LV being similar to a Japanese octopus trap, with a round bottom and narrow neck [2].

In our patient cardiac catheterization was the initial intervention since she presented with a chest pain associated with a typical ST elevation at 12 leads electrocardiogram. Anterior STEMI had to be ruled out initially. After this, transthoracic echocardiogram demonstrated a LV apical ballooning associated with a severe basal septal hypertrophy and hyperkinesis resulting in an increased velocity flow through the LVOT, and inducing a mitral SAM through the Venturi effect [4]. Since the coronary angiogram resulted normal, TTC with coexisting HCM physio-pathology was the most likely diagnosis.

The presence of SAM is highly suggestive of LVOT obstruction, which is known to induce MR [3,4]. TTC has been associated with both inducing LVOT obstruction and masking an existing HCM-induced pressure gradient. In our patient, initial echocardiographic findings demonstrated apical ballooning with reduced LVEF, significant LVOT obstruction, mitral SAM and MR. Three weeks later, there was resolution of TTC, but the LVOT obstruction, with a dynamic gradient >50 mmHg, persisted. This finding allowed to exclude a "sigmoid LV septal hypertrophy" which is related to aging. Sigmoid morphology of the interventricular septum is an isolated hypertrophy of the basal antero-septum, which

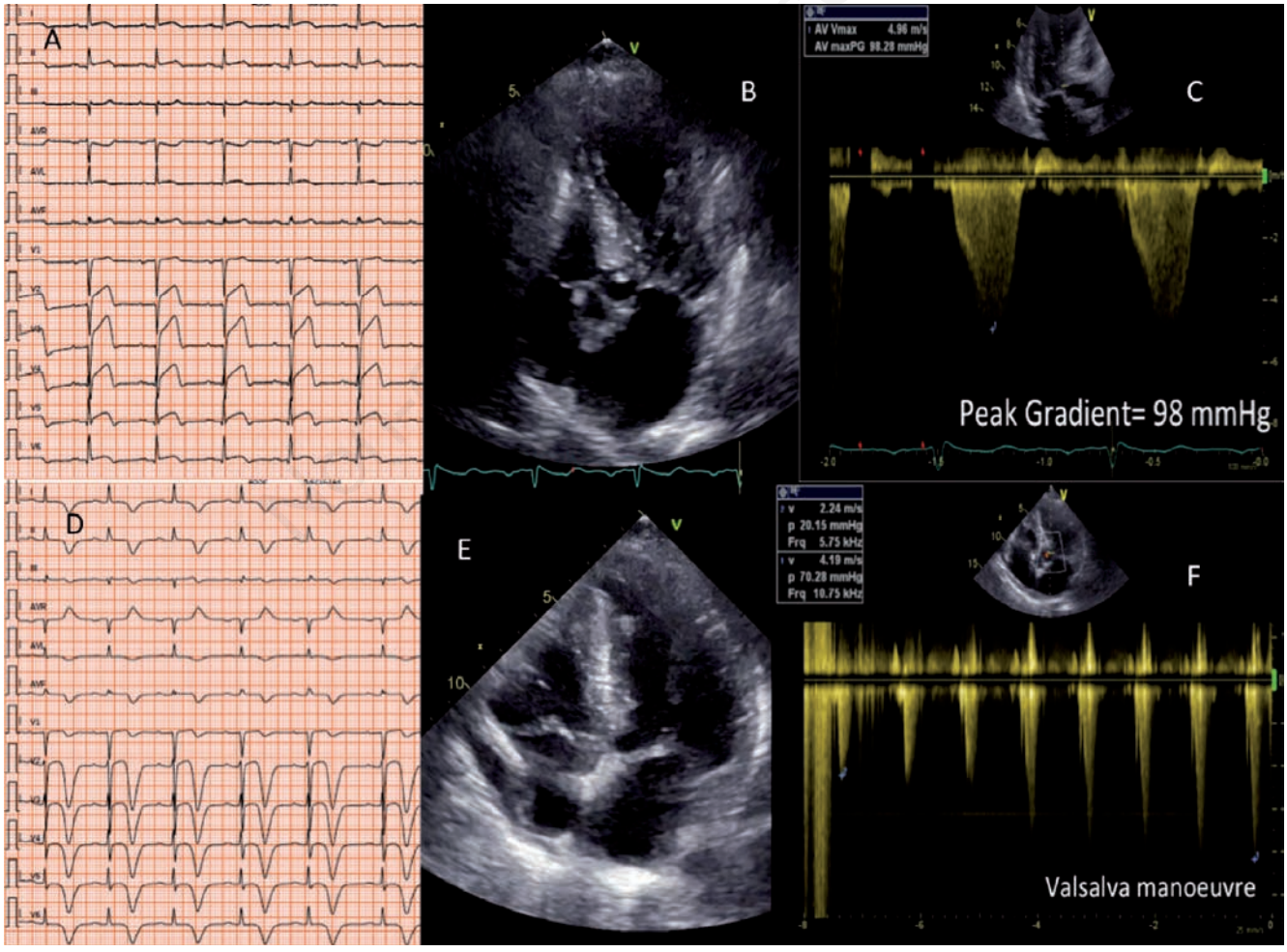

Figure 1. A) ECG at presentation. B) LV apical ballooning at TTE at presentation. C) LVOT peak gradient at presentation. D) ECG one week later. E) apical ballooning resolution at TTE one week later. F) LVOT peak gradient (at baseline and with Valsalva maneuver) one week later.

causes a type of subaortic stenosis [5]. It has been linked to long-standing hypertension [1], which was noted in our patient, but usually it does not provoke a significant LVOT obstruction [1]. Previous publications [6] suggest that LVOT obstruction might be present in 25% of patients with TTC. In these cases, TTE reveals LV basal hyperkinesis and abnormal septal thickening associated with LVOT obstruction, mitral SAM and MR, similar to the findings associated with HOCM [6]. This finding may be a consequence of increased catecholamine levels [6]. In one retrospective study of 3,272 patients presenting with an acute coronary syndrome, 32 patients were ultimately diagnosed with TCM (1% of patients). Of these, 20% (n = 6) had LVOT obstruction identified or confirmed by TTE [7].

In cases like ours, it is still unclear if LVOT obstruction is a "result" of TTC or if an HCM-related LVOT obstruction may be the "cause" of stress cardiomyopathy [8]. Hemodynamic deterioration described in our case may have two combined mechanisms: 1) LVOT obstruction due to both TTC and HCM; 2) acute LV systolic dysfunction due to TTC. An additional assumption is that TTC worsens HCM related LVOT-gradient from a hemodynamic standpoint, and once the patient recovers from TTC, the gradient improves on echo-doppler.

Management of cardiogenic shock due to this double mechanism is a challenge. Fluids and cautious use of beta-blockers are recommended [9] but inotropic agents (such as adrenergic drugs) may be deleterious because they worsen the degree of obstruction. We treated our patient with intravenous diuretics because of pulmonary congestion, adding beta-blockers when congestion was solving. The final outcome was good.

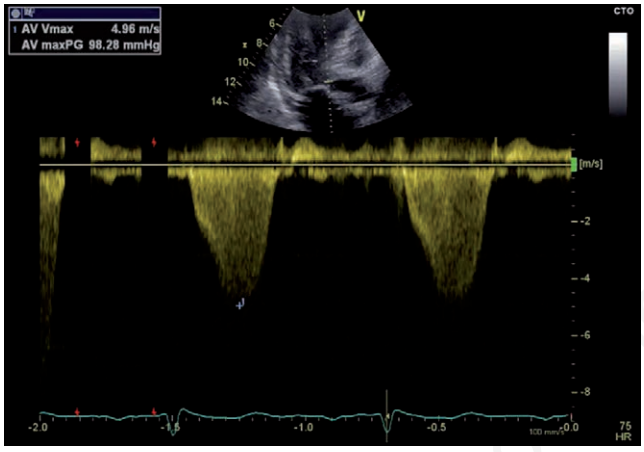

Figure 2. Continuous-wave doppler demonstrating a LVOT late peak gradient of 98 mmHg at time of TTC onset.

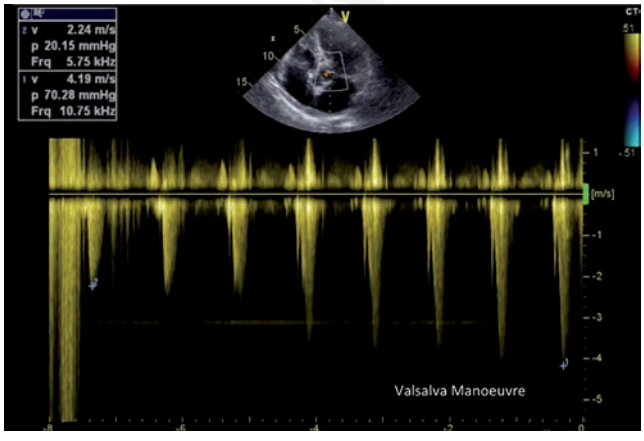

Figure 3. Continuous-wave doppler demonstrating a LVOT peak gradient decreasing to 20 mmHg, with a dynamic increase to 70 mmHg during Valsalva maneuver after LV apical ballooning resolution.

Conclusions

It is well known that TTC may be complicated by a reversible LVOT obstruction by itself but the combination with obstructive HCM can lead to low cardiac output and acute heart failure. This combination has been found to be not common and the correct treatment of this unusual type of cardiogenic shock is still unclear [9].

Careful initial evaluation and continuous monitoring must be warranted in such rare cases. Supportive care afterward with beta blockers, along with echocardiogram surveillance, are the mainstay of management. Cardiologists, intensivists, and clinicians alike need to recognize and comprehend the pathophysiology behind this unique clinical manifestation so that they may adjust their management and treatment accordingly.

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