Conical Deformity of the Left Ventricle: A Case Report

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Abstract

Most published information on the diagnosis of constrictive pericarditis assumes that the left ventricle has normal or nearly normal systolic function. Noninvasive assessment of pericardial constriction remains crucial to the diagnosis, especially in high-risk subgroups of patients, such as patients after cardiac surgery.

We present a 63-year-old Caucasian man who was admitted to the hospital for chronic decompensated heart failure. He had a past history of 3 open-heart surgeries for rheumatic heart valve disease. He was diagnosed with mixed pericardial and myocardial disease. The patient had a left ventricular ejection fraction of 38%, a functional mechanical aortic prosthesis, and a mechanical mitral prosthesis with elevated gradients. Severe pulmonary hypertension was present. The major findings that strongly suggested pericardial constriction were the presence of a conical deformity of the left ventricle, preserved indices of myocardial relaxation, and pericardial thickening. A reversed pattern of the velocities of diastolic relaxation was seen (lateral higher than septal), highlighting the possibility of mixed disease.

This clinical case illustrates how noninvasive methods can be used to conclusively diagnose pericardial constriction in the presence of left ventricular systolic dysfunction.

Keywords: Constrictive pericarditis; Pulmonary hypertension; Pericardial constriction; Noninvasive methods; Left ventricular systolic dysfunction

Abbreviations: TTE: Transthoracic Echocardiography; LV: Left Ventricular; BNP: B-type Natriuretic Peptide, LVEF: Left Ventricular Ejection Fraction; RV: Right Ventricle; CT: Computed Tomography

Introduction

A history of previous cardiac surgery is currently associated with over 30% of patients with constrictive pericarditis [1]. The diagnosis of constriction remains challenging, and although clinical information is useful, confirmation is needed, and is usually obtained using multimodality cardiac imaging [2]. Transthoracic echocardiography (TTE) remains the most important diagnostic test, according to the algorithm proposed by Verhaert et al. [2]. However, in routine clinical practice, the shortcomings of echocardiography become apparent, as the following clinical case will illustrate. Tissue Doppler imaging velocities across the mitral ring and patterns of mitral inflow are less reliable for diagnosing constrictive pericarditis when mitral and aortic valve prostheses are present. In addition, atrial fibrillation and frequent premature beats impede the analysis of respirometry TTE tracings. Finally, most published information on the diagnosis of constrictive pericarditis assumes that the left ventricle has normal or nearly normal systolic function [3].

Case Presentation

A 63-year-old Caucasian man was admitted for decompensated chronic heart failure to the heart failure unit in December 2011. His past medical history was remarkable for mitral valve repair using a no. 34 Carpentier ring for rheumatic mitral valve disease 20 years previously. Mitral (no. 27 Medtronic-Hall) and aortic (no. 23 Medtronic-Hall) valve replacements were performed for symptomatic mixed mitral valve disease and severe aortic regurgitation 12 years previously. Two months before the current admission, the patient underwent an open-heart procedure to close a mitral prosthesis paravalvular leak. Before surgery he had a normal left ventricular (LV) ejection fraction

with mild-to-moderate LV enlargement. The surgical report of the last surgery before admission was remarkable for mild thickening of the pericardium (4 to 5 mm), especially over the left ventricle, with focal calcification in the pericardium at the apical region. Pericardial stripping was not performed. Regarding other comorbidities, the patient had mild chronic renal failure, chronic anemia, hyperuricemia, and dyslipidemia.

At the current admission, the patient presented with shortness of breath after minimal effort, marked fatigue, and presyncope. Physical examination revealed significant cachexia, edema of the lower extremities, ascites, enlarged liver (edge palpable 5 cm below the costal margin), and marked jugular venous distension (waveform visible at the angle of the mandible while sitting upright at 90 degrees) with marked Y descent and Kussmaul's sign. Cardiac auscultation revealed an irregular rhythm, 90/min, with good prosthetic mitral and aortic valve closure sounds. A systolic murmur was heard over the left sternal border. Pulmonary auscultation revealed absent murmur at both bases and rales bilaterally in 2/3 of the lung fields. His blood pressure was 130/95 mm Hg, and peripheral oxygen saturation was 90% while breathing room air. Intravenous diuretics and nitrates were started, and the patient had a good initial clinical response. Laboratory work revealed elevated B-type natriuretic peptide (BNP) (743.3 pg/mL), creatinine (1.4 mg/dL) and C-reactive protein (14.1 mg/dL). There were also laboratory findings suggesting liver congestion, and hyponatremia (131 mmol/L). Admission blood cultures were negative.

The admission TTE revealed a non-dilated conically deformed (tube shaped) left ventricle (Figure 1) with an estimated LV ejection fraction (LVEF) of 38% (confirmed by a nuclear multiple-gated acquisition scan). The right ventricle (RV) was dilated with moderately-to-severely depressed systolic function. Both a septal bounce and moderate biatrial enlargement were seen (videos S1 (http://youtu.be/pgI8Xyu81FE) and video S2 (http://youtu.be/9FBaRD9NNr0). The tilting-disk mitral prosthesis was well seated, with a small central regurgitation and no apparent leaks. The mitral gradients were 18 and 7 mm Hg. The tilting-disk aortic prosthesis was well seated, with a small central regurgitation and a small right-posterior leak. The aortic gradients were 23 and 12 mm Hg, with a dimensionless index of 0.41. The LV lateral E' was 10 cm/s, LV septal E' was 7 cm/s, and RV lateral E' was 8 cm/s (Figure 2.1-2.3). Global LV longitudinal strain was -10%. A dilated vena cava and reversed expiratory flow in the suprahepatic veins were identified. Respiratory variation of mitral inflow was not present, but a 15% variation in descending aortic blood flow was seen (Figure 2.4).

Right and left heart catheterization was also performed. There were no signs of coronary disease. The right atrial pressure waveform revealed a steep y descent (Figure 3.1). A dip-and-plateau was evident in the right ventricular diastolic tracing (Figure 3.2). Equalization of end-diastolic pressures was seen (RA 18 mm Hg, RV 16 mm Hg, pulmonary capillary wedge pressure 25 mm Hg [minus mean mitral gradient]) and pulmonary hypertension (95/27 mm Hg, mean 53 mm Hg, with an elevated transpulmonary gradient of 28 mm Hg). The cardiac index was calculated to be 1.84 L/min/m².

Thoracic computed tomography (CT) was performed on the following day and revealed mild circumferential thickening of the pericardium, sparing the RV, with focal thickening over the lateral and inferior LV walls. Also, areas of calcification were seen near the apex (Figure 4) contributing to the conical deformity of the left ventricle.

The patient improved with administration of intravenous diuretics, colchicine, and a course of inotropic therapy, but at the end of the first week of hospitalization, he was diagnosed with a hospital-acquired methicillin-resistant *Staphylococcus aureus* pneumonia and developed septic shock. He died three weeks later and was not autopsied at the family's request.

Discussion

Our patient probably had mixed pericardial and myocardial disease. The etiology of the myocardial disease was not completely

Figure 1: Conical deformity of the left ventricle. Mild LV wall thickening. Biatrial dilatation. Mechanical tilting disk mitral valve prosthesis. Cavitation.
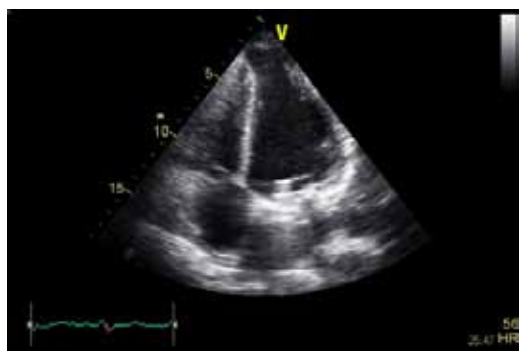

Figure 2: Tissue Doppler imaging of the septum (2.1), lateral LV (2.2), lateral RV (2.3), and respiratory variations on the descending aorta (2.4). Note the preserved diastolic myocardial velocities, although with a higher value for the basal lateral LV than the septal LV.
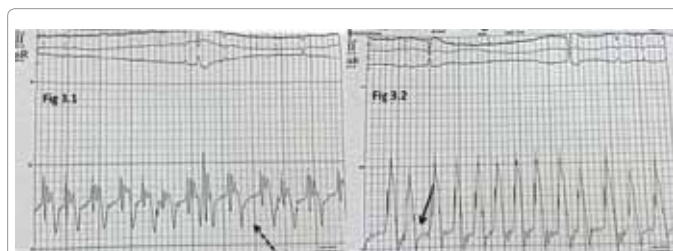
Figure 3: Cardiac catheterization. Figure 3.1: right atrial pressure waveform with a steep y descent (dashed arrow). Figure 3.2: right ventricular pressure waveform with a dip and plateau sign (solid arrow). Equalization of end-diastolic pressures.
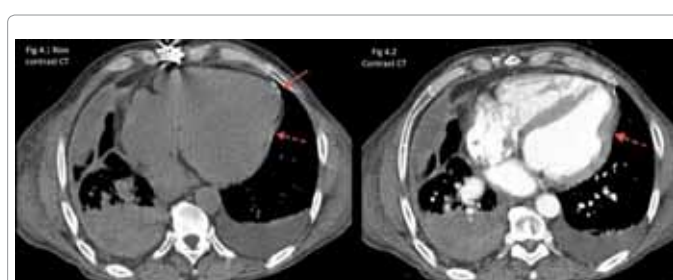
Figure 4: Computed tomography scans of the thorax (with and without contrast). Dashed arrow: localized thickening in the mid infero-lateral LV wall (6.4 mm). Solid arrow: pericardial calcification. There is also right-sided pleural thickening with bilateral pleural effusion (organized in the right pleural cavity).

understood, but could have been the result of end-stage constrictive pericarditis. Findings suggesting myocardial disease include the high BNP level at admission, decreased ejection fraction of both ventricles, and the reduced longitudinal global strain. Moreover, severe pulmonary hypertension (with a venous and arterial component) was present, which was probably also related to the long-standing mitral valve disease and a restrictive mitral valve prosthesis. Evidence for pericardial disease includes the morphological changes of the LV cavity, the septal bounce, CT findings and the results of the cardiac

catheterization. It is notable that pericardial thickening was seen to occur mainly around the left ventricle, sparing the right ventricle and calcium deposits were identified in the pericardium at the apex. The pericardial disease tethered the adjacent myocardium, and because of its asymmetrical distribution contributed to the LV morphological abnormalities seen in our patient. We therefore believe that the conical LV deformity could be explained by the myocardial involvement, the pericardial thickening around the LV, and the focal calcifications in the pericardium at the apex. Unfortunately, we could not obtain pathological confirmation. The reference to pericardial thickening with calcium deposits mentioned in the previous surgical report is consistent with the findings of the hemodynamic study with catheterization performed early during the course of this latest hospitalization, allowing us to assume he had constrictive pericarditis.

In our patient the diastolic velocities of the left ventricle were preserved (a less likely scenario if there were only myocardial disease), but contrary to what has been reported for other cases of typical pericardial constriction, the lateral E' was higher than the septal E´ (pseudo-normal pattern). We believe that the combination in our patient of mitral valve prosthesis, depressed LVEF, and severe pulmonary hypertension may have been responsible for the different pattern of myocardial relaxation despite the presence of pericardial constriction.

Conclusions

The diagnosis of constrictive pericarditis in the presence of depressed LV systolic function remains challenging. A detailed TTE is crucial for early diagnosis, which is based on LV morphology, septal bounce, and preserved and reversed Doppler velocities of myocardial relaxation. A conical LV deformity should alert the physician to the possibility of pericardial constriction.

Written informed consent was obtained from the patient relatives for publication of this manuscript and the accompanying images.

Authors' Contributions

RT performed the TTE examinations. RT, NM, and RM analyzed and interpreted the imaging data. FF analyzed and interpreted the clinical and hemodynamic data of the patient. All authors read and approved the final manuscript.

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