

CASE REPORT

Constrictive Pericarditis in the Light of Multimodal Cardiac Imaging. A Case Report

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ABSTRACT

Heart failure (HF) caused by constrictive pericarditis (CP) is very rare, but has a significant healing potential. In order to diagnose it, an initial high level of suspicion is imperative, given that HF presents in a setting lacking clinical signs capable of pinpointing a specific aetiology. However, current modern imaging techniques permit the accurate construction of a diagnosis for CP, clearing the way for surgical treatment. We are describing the case of a 60-year-old male who was hospitalised to our Cardiology Department due to a history of HF that gradually progressed to the congestion stage over the past six months. The diagnosis of CP was established after the examination of echocardiography and cardiac magnetic resonance imaging results. The patient underwent subtotal pericardectomy, and to this date, he has made a full recovery. The purpose of this case report is to highlight the importance of considering less common causes of HF, in addition to the common ones, in order to devise the most appropriate investigations and expedite surgical correction of this condition.

Keywords: constrictive pericarditis, heart failure, echocardiography, magnetic resonance imaging.

INTRODUCTION

Hear failure (HF) caused by constrictive pericarditis (CP) is very rare, but has a significant healing potential. In order to diagnose it, an initial high level of suspicion is imperative, given that HF presents in a setting lacking in clinical signs capable of pinpointing a specific aetiology. However, current imaging techniques allow nowadays for an accurate diagnosis of CP, paving the way for surgical treatment. □

CASE REPORT

History. A 60-years old male was referred to our Cardiology Department for a history of HF progressively developed to congestion stage within the last six months. The symptoms started with persistent exertional dyspnoea and fatigue progressing to dyspnoea at rest, orthopnoea and weight gain with general oedema. No febrile episode, nor cough or chest pain were described in his recent past previously to this clinical evolution. We have to mention also that he did not

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have a history of malignancy, irradiation, autoimmune or infectious diseases. At a certain moment he was considered to have dilated cardiomyopathy and based on this assumption he was administered a conventional treatment with digoxin, carvedilol, candesartan, spironolactone and furosemide without relieving symptoms. Two months before the current hospitalization he went through a mild infection with SARS-CoV-2.

Physical examination revealed orthopnea, jugular venous distension associated with Kussmaul sign bilateral pleural effusion syndrome, ascites in moderate quantity and important lower extremity edema. The patient's body mass index (BMI) was 34.5 kg/m². Vital signs were stable, with blood pressure 110/70 mm Hg, ventricular rate of 88 beats/minute and an oxygen saturation of 97% in ambient air.

The laboratory work-up has indicated normal values for common tests, including those for re-

nal and hepatic function, without anemia, a rise in inflammation tests or tumoral markers or abnormality of thyroid hormones. The serology for the infection with hepatitic viruses B and C or for HIV was negative, while D-Dimer levels were negative as well. NT-proBNP was the only abnormal parameter with a value equal to 1350 pg/mL.

Electrocardiogram (ECG) showed normal sinus rhythm, reduced voltage and nonspecific T-wave abnormalities.

Transthoracic echocardiography has shown the following features:

- Normal dimensions of the walls and ventricles, with a moderate enlargement of the left atrium and possible thickening of the pericardium in the postero-basal segment (Figure 1)
- Normal ejection fraction of the left ventricle, but with signs of decreased systolic function of the right ventricle: tricuspid annular plane systolic excursion (TAPSE) of 13 mm
- Dilated inferior cava vein diameter of 23 mm, without phasic variations with respiration
- No significant valvulopathies have been noticed
- Severe dyskinesia of the interventricular septum in early diastole (septal bounce)
- Mitral flow – restrictive pattern of diastolic filling with E/A ratio >2 and time (in expiration) of E wave deceleration < 160 msec; phasic variations of transmitral flow with respirations of 38% have been recorded (Figure 2).
- Aortic flow – phasic variations of transaortic flow with respirations of 34% have been recorded (Figure 2)



FIGURE 1. Parasternal long axis view indicating normal dimensions of the left cavities

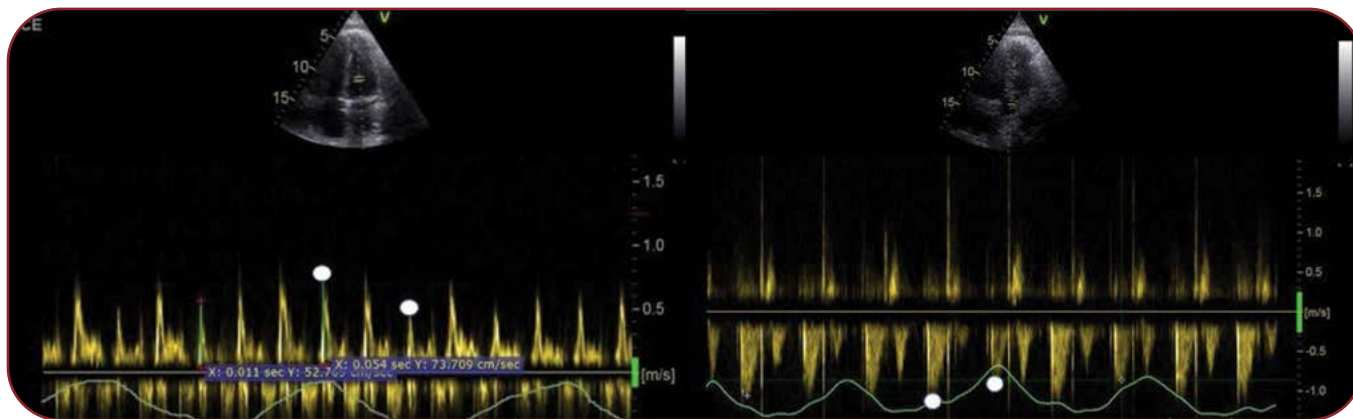


FIGURE 2. Panel A: restrictive pattern of transmitral flow with variations between expiration and inspiration of 38%; panel B: variations of transaortic flow between expiration and inspiration of 34%

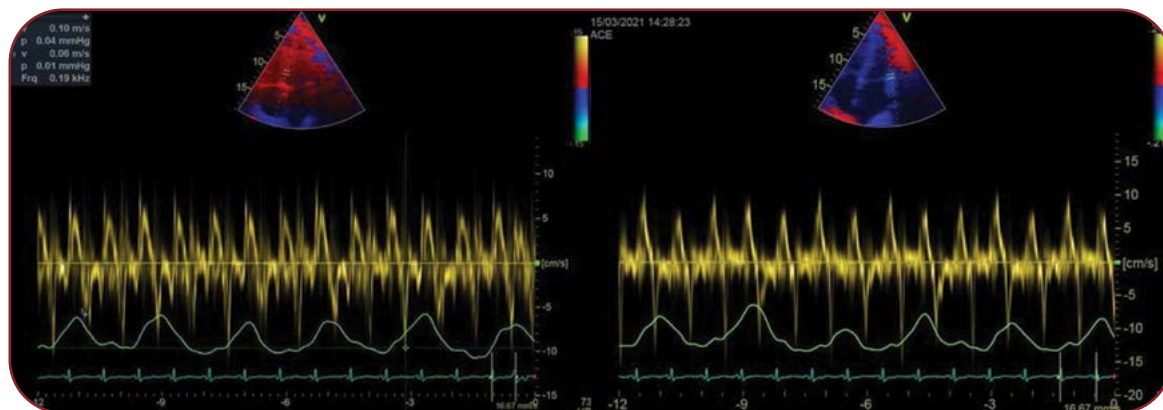


FIGURE 3. Panel A: in TDI application, mitral E' has normal values and varies with the respiratory cycles; panel B: in TDI application, lateral E' has normal values and varies with the respiratory cycles, without *annulus inversus* phenomenon

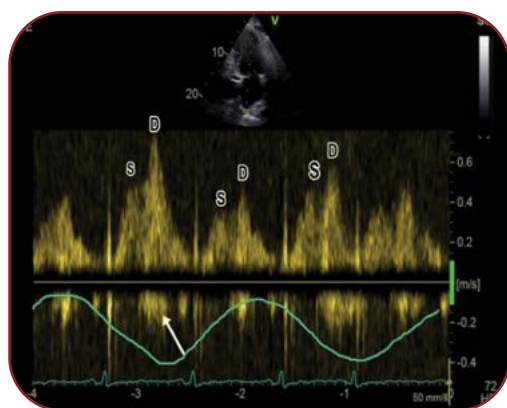


FIGURE 4. Pulmonary vein flow: decrease in S and D wave velocities during inspiration and increase of atrial reflux in expiration

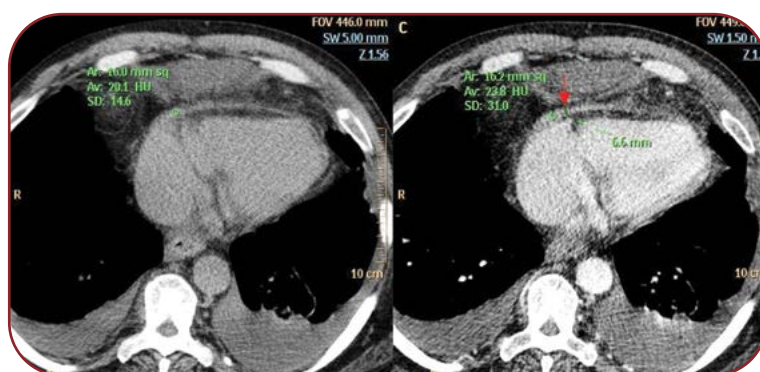


FIGURE 5. Thoracic computed tomography (CT) scan: thickening of pericardium with contrast enhancement; panel A: axial non-contrast thoracic CT; panel B: axial contrast-enhanced thoracic CT reveals the presence of pericardial thickening measuring up to 6.6 mm laterally to the right cavities (red arrow)

- Tissue Doppler imaging (TDI) revealed extra-normal values of E' wave velocities (10 cm/sec) and its variations with respirations higher than 15%, with no *annulus inversus* phenomenon (mitral E' higher than lateral E') (Figure 3)
- Pulmonary veins flow – a decrease in S and D wave velocities in inspiration and an increase of atrial reflux in expiration have been noticed (Figure 4)
- Tricuspid flow – variations higher than 25% of inflow velocities in inspiration compared with expiration
- Hepatic vein flow was not recorded.

Before concluding a diagnosis, we ordered an urgent thoracic computed tomography (CT) scan in order to eliminate severe pulmonary thromboembolism, which was mostly suggested by the clinical signs of right heart failure and interventricular septum dyskinesia. This examination con-

firmed the presence of bilateral pleural effusions, indicated a dilation of the pulmonary artery and eliminated the presumption of pulmonary thromboembolism. Moreover, it revealed a slightly thickening of pericardium situated anterior to the right ventricle (6 mm vs >2 mm in normal situations) with contrast enhancement (Figure 5).

In this context, we returned to the information provided by echocardiography and checked the Mayo criteria for constrictive pericarditis. A high probability of diagnosis was raised by three out of four items: the septal bound, E/A > 1 and preserved/increased flow velocity of mitral E'. We were not able to provide informations about hepatic veins flow. To confirm this diagnosis, a cardiac magnetic resonance imaging (MRI) exam of the heart was performed, which detected the presence of pericardial thickening up to 5-6 mm

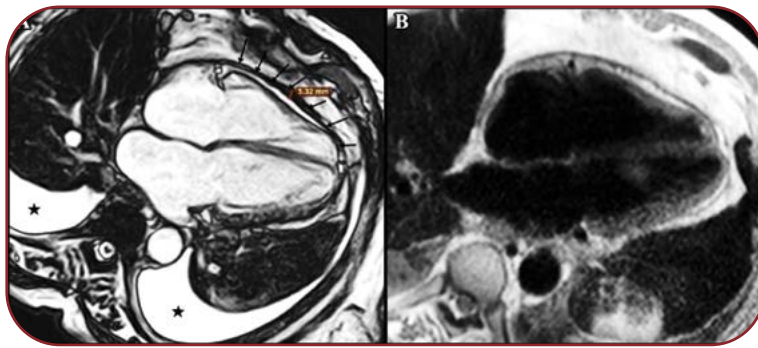


FIGURE 6. Panel A: cardiac magnetic resonance imaging (MRI), 4-chamber view cine 2D showing pericardial thickening up to 5.32 mm along the entire length of the right ventricular free wall (black arrows) and bilateral pleural effusions (black stars); panel B: cardiac MRI, 4-chamber view in T1-weighted showing the absence of myocardial injury

along with the free and posterior wall of right ventricle (Figure 6, panel A). T1-weighted image refuted myocardial injury (Figure 6). Panel B and T2-weighted image excluded myocardial edema. Mild tricuspid and mitral regurgitation without hemodynamic significance and dilation of inferior vena cava and main pulmonary artery were also described.

Based on the association of signs and symptoms of right heart failure and pericardial constriction with impaired diastolic filling, which was revealed by multimodality imaging, we established the definitive diagnosis of constrictive pericarditis. Afterwards, the heart team met and decided that surgical correction, with prior **angiographic assessment of coronary arteries** as part of the preoperative protocol, was the best treatment option. Subtotal pericardiectomy was performed and pericardial biopsy revealed fibrin deposits with structural organization of fibrous connective tissue, discrete, chronic inflammatory infiltrates and acute perivascular inflammation, focal reactive mesothelial hyperplasia, focal hemorrhages and deposits of hemosiderin pigment, without evidence of malignancy or tuberculosis. One day after surgery, the patient had an episode of symptomatic paroxysmal atrial fibrillation with dyspnea and palpitations, without hemodynamic instability. Successful electrical cardioversion was performed, without any further complications. He was discharged 10 days after surgery. Follow-up at one month post-discharge revealed weight loss with resolution of the generalized edema, improvement in diastolic function, decreased variation of the E wave in inspiration below 25%, absence of septal bounce

and decreased velocity of the septal e' wave on tissue Doppler examination. ■

DISCUSSIONS

Constrictive pericarditis is the result of a chronic inflammatory process which leads to fibrotic thickening of pericardium, loss of pericardial elasticity and restriction in the ventricular filling (1). The most common cause of CP is idiopathic or viral. Other possible etiologies include cardiothoracic surgery, radiotherapy, infectious or autoimmune diseases and malignancies (2). In the present case report, we have concluded on an idiopathic form of chronic CP. The epidemiologic data regarding CP are scarce. It is recognised as a rare pathology. According to some reports, CP is likely to represent less than 0.01% of hospital admissions (3).

Pathophysiology

The non-compliant pericardium in CP is responsible for two pathophysiological phenomena: exaggerated ventricular interdependence and intrathoracic-intracardiac pressure dissociation (4). These pathogenic links result in dynamic respiratory changes of the ventricular filling patterns and trans-aortic flow (5, 6). During inspiration, the intrathoracic pressure decreases and augmented venous return from inferior vena cava increases the filling of the right ventricle. A normal pericardium allows the accommodation of the right ventricle to this influx and the variations of the transotricuspid flow are manifest but reduced. In CP, the expansion of the right ventricle is reduced and gives rise to a higher difference in tricuspid inflow velocities between inspiration and expiration. Moreover, the impaired lusitropy of the pericardium hinders the expansion of the right ventricle and therefore leads to the shifting of the septum towards the left side, thereby contributing to decreased blood inflow in the left ventricle and lower mitral velocities during inspiration. This effect is amplified by the reduction of left ventricular filling due to the drop in pulmonary veins (not encapsulated in the pericardium). On the contrary, during expiration, the intrathoracic pressure increases the left ventricular filling, causing a rightward movement of the interventricular septum and the underfilling of the right ventricle with backflow of blood into the hepatic veins (7-9).

Diagnosis

There are some important messages to be transmitted regarding the diagnosis of CP. First of all, a high level of suspicion from the clinical stage of diagnosis should be kept in mind, with the aim of starting an imagistic investigation as early as possible. Delays of more than 10 years have been reported in patients with chronic severe ascites (10).

For increased diagnostic accuracy, several diagnostic algorithms have been developed over time, especially based on multimodality cardiovascular imaging. Thus, transthoracic echocardiography is the first-line imaging test in patients with suspected CP (2). Detailed hemodynamic evaluation is the key advantage of echocardiography in the diagnosis of CP. The main hemodynamic criterion found in CP is the respiratory variation in mitral inflow velocity quantified by an increase of more than 25% in mitral E velocity during expiration compared with inspiration (5, 11). Another finding in CP is the thickening or calcification of the pericardium; however, it can be absent in 12-18% of patients (5). Septal bounce during early diastole, respirophasic ventricular septal shift and dilated inferior vena cava and hepatic veins are also included in echocardiographic findings of CP (5, 11). The systolic function of left ventricle is usually normal and biatrial enlargement is common. Tissue Doppler imaging and strain imaging can be helpful in differentiating from restrictive cardiomyopathy (4, 8, 11). During the validation of Mayo criteria for the echocardiographic diagnosis of CP it has been shown that interventricular septal shift in association with E' medial mitral annular velocity ≥ 9 cm/s had a diagnostic sensitivity and specificity of 87% and 91%, respectively (5).

The current guidelines of The European Society of Cardiology (ESC) recommend CT and/or MRI as second-level imaging procedures after echocardiography and chest X-ray (2). In most cases, non-invasive multimodality imaging is enough to confirm the diagnosis of constrictive

pericarditis. When constriction is not proven, the ESC Guidelines recommend the use of cardiac catheterization (2). In our case, the cardiovascular multimodality imaging alone was sufficient to prove the diagnosis of constrictive pericarditis.

The most important and difficult differential diagnosis of CP remains that of restrictive cardiomyopathy, but multimodal cardiovascular imaging helps the cardiologist to establish the correct diagnosis.

Treatment

Constrictive pericarditis is a potentially curable cause of diastolic heart failure through timely surgical treatment. Timely surgical pericardiectomy, before the onset of New York Heart Association (NYHA) Class III or IV symptoms, is associated with a significantly lower risk of 30-day surgical mortality (13). Total pericardiectomy is the main treatment in CP. Studies have shown that the rate of complications was higher in incomplete decortications compared with total pericardiectomy. However, the long-term outcome is dependent on not only the type of surgery but also the preoperative NYHA status and the etiology of pericardial disease. If there are adhesions and calcifications of the pericardial layers, it may be prudent to maintain a few islands of pericardium. Therefore, a subtotal pericardiectomy was performed with the improvement of both the NYHA functional class and echocardiographic parameters observed at one month follow-up. $\square$

CONCLUSIONS

The present case report emphasizes the importance of taking into account rare causes of HF, beyond the common ones, in order to plan the most appropriate investigations and to make surgical correction of this disease as promptly as possible. $\square$

Conflicts of interest: none declared.

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