Constrictio Cordis: Can a Thin Pericardium Restrict the Heart?

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INTRODUCTION

Constrictio cordis (CC) is a rare condition usually caused by a thick, noncompliant pericardium, which can be inflamed, fibrotic, and/or calcified. Cardiac filling is impaired because of this, and full transmission of respiratory intrathoracic pressure changes to the cardiac cavities is prevented. Normal pericardial thickness is ≤2 mm. Most data show that a pericardial thickness >4 mm indicates pericardial constriction, and a thickness >6 mm has high specificity for CC. However, the classic thickened nature of the pericardium in CC is not always the case, and an apparent thin pericardium, in which the visceral layer of the pericardium is causing the constriction, can have the same hemodynamic impact and should therefore not exclude the diagnosis of CC. We report a case of a 55-year-old man with symptoms of right heart failure. Despite no signs of a thickened pericardium on multiple-modality imaging, surgical findings confirmed the diagnosis of CC despite a thin pericardium.

CASE PRESENTATION

A 55-year-old man with known non-insulin-dependent diabetes mellitus, hypertension, obesity, and atrial fibrillation was referred to our institution because of 18 months of symptoms consisting of dyspnea, peripheral edema, and ascites. The patient was initially hospitalized because of increased abdominal extent and weight loss but had no cardiac symptoms. Chest radiography and ultrasound showed pleural effusion and ascites (Figure 1). Pleural and abdominal drainage were performed, draining 2.5 and 9 L, respectively. Initially, malignancy was suspected, but multidetector computed tomography of the thorax and abdomen showed no signs of malignancy, and the pleural effusion lacked malignant cells. Multidetector computed tomography showed hydorenphrosis. Despite treatment with bilateral JJ catheters, production of ascites continued. As a result, the patient had ascites drained every eighth day. After examination, the cardiologist did not suspect any heart disease, and the results of echocardiography were interpreted as normal except for a minor pericardial effusion.

After thorough examination by both urologists and gastroenterologists at the local hospital, no diagnosis was established. Diuretic treatment was intensified with stable creatinine levels. Both ultrasound and multidetector computed tomography of the abdomen showed no signs of cirrhosis, and hepatitis B virus and hepatitis C virus screening were negative. The patient was then set up for regular pleural and ascites drainage.

The patient was once again referred to a cardiologist, and after a new echocardiographic examination, abnormal pericardium was suspected at the lateral-posterior segment of the left ventricle. A repeat computed tomographic scan of the thorax showed pericardial calcification, and the patient was then referred to a tertiary center (Rigshospitalet) with the diagnosis of possible CC. Here, the following examinations were performed.

Focused echocardiography showed a dilated inferior vena cava (2.87 cm) with retrograde diastolic liver vein flow, abnormal high Doppler tissue imaging velocity with a paradoxically high septal-to-lateral ratio and mitral filling pattern, as well as classical “septal bounce.” Furthermore, there was bilateral atrial dilatation (left atrium 49 mL/m²), small ventricular volumes (left ventricle 4.5 cm in the diastolic phase), and a low cardiac index (2.26 L/min/m²; Figure 2, Video 1).

Cardiac magnetic resonance imaging showed a nonthickened pericardium and no pericardial effusion, but tagging of the left ventricle suggested that the pericardium was fixated to the ventricle. The left ventricle was small and cone shaped, and cardiac output was below the normal range. Bilateral atrial enlargement and a large pleural effusion on the left side were present. Septal shift during respiration was not present, but the patient had difficulty cooperating because of dyspnea. The conclusion of cardiac magnetic resonance imaging was possible constrictive pericarditis due to adhesion of the pericardial layers (Figure 3, Video 2).

Right heart catheterization showed equalization of diastolic chamber pressure with a dip-plateau phase with increased pulmonary capillary wedge pressure of 24 mm Hg, pulmonary artery pressure of 45/23 mm Hg, right ventricular pressure of 43/21 mm Hg, right atrial mean pressure of 18 mm Hg, and high filling pressures of both ventricles. The filling of the left ventricle had an apparent plateau phase. The conclusion was suspicious for CC but with a need for comparison with other imaging modalities.

Because of high clinical and overall suspicion for CC, the patient was referred to surgery, where the pericardium was not thickened on inspection but adhesive to the ventricles sending septae into the myocardium, which could be peeled off as if peeling an onion.

A biopsy from the pericardium showed thickened tissue due to diffuse and cell-poor fibrosis, focal hemorrhage (possibly occurring intraoperatively), light perivascular infiltration of lymphocytes and plasma cells, and a few lymphocytic follicular accumulations in the transition to the attached fatty tissue. There was no abnormal...
Figure 1  Chest radiography. (A) Posterior-anterior projection. (B) Lateral projection. Images are taken the day before surgery. On the left side is a large pleural effusion (black arrow) and atelectasis. On the right side, minor atelectatic changes basal and a small pleural effusion (white arrow).

Figure 2  Echocardiographic images. (A) Dilated inferior vena cava of 2.9 cm. Less than 50% collapse during inspiration suggests increased pressure in the right atrium. (B) Flow curves through the inferior vena cava showing diastolic retrograde flow. (C) Apical four-chamber image demonstrating bialtrial enlargement, normal ventricles, and a high signal from the pericardium. (D) Pulse velocity flow through the mitral valve. High E peak and low deceleration time (110 msec). (E,F) Tissue Doppler of the medial and lateral mitral annulus demonstrating a high ratio of early diastolic septal versus lateral velocities (medial e'/septal e' = 1.2).
mesothelial proliferation or signs of malignancy. The picture corresponded well with the consequences of constrictive pericarditis.

In the postoperative phase, the patient was stable. He had a few episodes of atrial fibrillation that needed direct-current cardioversion but was discharged without further complications.

CONCLUSION

Traditionally, CC has been associated with a normal-sized heart and restricted diastolic filling secondary to a rigid calcified pericardial shell.4 The diagnosis of constrictive pericarditis remains a challenge because the physical findings and hemodynamics are similar to restrictive cardiomyopathy. Pericardial thickness may help distinguish between these. However, in some cases of CC, presence of normal thickness of the pericardium on imaging should not exclude the diagnosis of CC.4

The most specific echocardiographic findings suggesting CC are abnormal ventricular septal motion to the left in early diastole in inspiration (septal shift), abnormal high medial Doppler tissue imaging velocity, and diastolic reversal liver vein flow.5 This case explains how most patients must go through a multitude of examinations before final diagnosis of CC. Even though cardiac magnetic resonance imaging showed a nonthickened pericardium, a synthesis of all examinations—echocardiography, right-sided cardiac catheterization, and clinical presentation—were in strong favor of the diagnosis of CC, which was confirmed during surgery. Multi-modality imaging is essential for the diagnosis of CC because even a thin pericardium can restrict the heart.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at http://dx.doi.org/10.1016/j.case.2017.03.003.

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