CASE REPORT

Pericardiectomy as a diagnostic and therapeutic procedure

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SUMMARY

A 70-year-old man presented with recent onset, predominantly right-sided heart failure. Echocardiogram demonstrated features of hypertensive heart disease and was suggestive of, but non-diagnostic for, constrictive pericarditis (CP). CT demonstrated mild pericardial thickening. Right heart catheterisation showed elevation and equalisation of diastolic pressures in all cardiac chambers with early rapid filling, minimal ventricular interdependence, and no dissociation of intrathoracic and intracardiac pressures. While several features pointed towards CP, the minimal ventricular interdependence and no dissociation of intrathoracic and intracardiac pressures suggested other pathology. Diagnostic pericardiectomy was performed, after which the central venous pressure

decreased from 22 to 12 mm Hg. Pathology revealed pericardial fibrosis. The patient experienced sustained resolution of his heart failure. A potential explanation for lack of CP criteria was the presence of hypertensive heart disease. CP needs to be considered when approaching patients with heart failure as diagnostic evaluation can be multifaceted and treatment curative.

BACKGROUND

Advances in imaging techniques and combination of invasive haemodynamic criteria have resulted in greater diagnostic accuracy of constrictive pericarditis (CP). In some cases, however, despite careful integration of clinical, imaging and haemodynamic

Figure 1 (A) Pulsed-wave Doppler tracing of the mitral inflow with only minimal respiratory variation; (B) pulsed-wave Doppler tracing of hepatic veins; diastolic reversal ratio 0.90; (C) tissue Doppler imaging of septal mitral annulus; e' velocity 11 cm/s.

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Figure 2 Cardiac CT showed mild pericardial thickening, maximum 4 mm, without calcifications.

data, the diagnosis of constriction versus restriction remains uncertain. Since the consequences of missing a diagnosis of CP, which is amenable to a 'surgical cure', are profound, the definitive step may involve diagnostic pericardiectomy. If CP is present, pericardiectomy can result in complete resolution of heart failure symptoms. High index of suspicion for CP, a potentially reversible condition, is necessary when evaluating patients with predominantly right-sided heart failure symptoms.

CASE PRESENTATION

A 70-year-old man with history of long-standing hypertension, diabetes mellitus type 2, coronary artery disease and atrial fibrillation presented with fatigue, dyspnoea on exertion and extensive peripheral oedema of 6-month duration. Despite escalating doses of furosemide and daily metolazone, there was persistent extensive peripheral oedema. On examination, his blood

pressure was 115/87 mm Hg, and heart rate was 95 bpm. His jugular venous pressure was elevated above the angle of jaw when sitting upright, without appreciable decrease with inspiration. Heart rhythm was irregularly irregular, with variable first heart sounds. There were no additional heart sounds. No abdominal organomegaly and no ascites were present. Lungs were clear. There was a 3+ bilateral peripheral oedema up to the knees.

INVESTIGATIONS

Complete blood count and basic metabolic panel were within normal limits. Transaminases were normal. Alkaline phosphatase was mildly elevated at 155 U/L, as was the ESR (30 mm at 1 hour). ECG demonstrated atrial fibrillation with heart rate 102 bpm. Left ventricular (LV) hypertrophy was presented by the Cornell criterion, and there were non-specific ST and T wave abnormalities. Chest X-ray demonstrated normal cardiac silhouette; there were no pericardial calcifications, pulmonary congestion nor pleural effusions.

An echocardiogram showed normal LV ejection fraction and normal right ventricular (RV) systolic function. The interventricular septum measured 16 mm in end diastole. The inferior vena cava was distended without inspiratory collapse. Notably, the echocardiogram showed no septal shift or shudder. There were minimal respiratory variation in the mitral inflow velocity curves and mild expiratory diastolic flow reversals in the hepatic vein velocity curves (figure 1A, B). Tissue Doppler velocity of the medial and the lateral mitral annulus in early diastole (e') were 11 cm/s (figure 1C). Thus, the echocardiogram demonstrated only two of five criteria for CP described as Mayo Clinic criteria by Welch *et al*,¹ namely medial e' velocity ≥ 9 cm/s and expiratory diastolic flow reversals in the hepatic vein velocity curves. Medial e' velocity ≥ 9 cm/s is 83% sensitive and 81% specific, and hepatic vein reversal ratio ≥ 0.79 is 76% sensitive and 88% specific for CP. The criteria of ventricular septal shift and change in mitral E velocity $\geq 14.6\%$ and medial e' /lateral e' ≥ 0.91 were not met.

CT imaging of the chest showed mild pericardial thickening, maximum 4 mm, without calcifications (figure 2). Cardiac MRI showed mild pericardial delayed enhancement and non-specific delayed myocardial enhancement of the interventricular septum

Figure 3 Haemodynamic tracings with high-fidelity micromanometer catheters: (A) RA and LV; mean RA pressure 18 mm Hg and rapid y descent. (B) PAWP and LV; lack of intrathoracic-intracardiac pressure dissociation. Mean PAWP 24 mm Hg. (C) LV and RV; minimal enhancement of ventricular interaction during the respiratory cycle. The orange shade demarcates the LV area and the yellow shade the RV area. The ratio of the RV area to the LV area in inspiration versus expiration defines the systolic area index. LV, left ventricular; PAWP, pulmonary artery wedge pressure; RA, right atrial; RV, right ventricular.

at the site of RV insertion. While both studies provided further anatomical information, the presence of an abnormal pericardium does not necessarily reflect constrictive haemodynamics. Patients may have abnormal pericardial thickness in the absence of constriction, especially following cardiac surgery or chest radiation. However, the patient had no history of cardiac surgery or chest radiation. Pericardial enhancement after contrast administration on cardiac MRI is suggestive of ongoing inflammation. This feature may be characteristic of acute pericarditis; however, it is non-specific and should be interpreted within clinical context, and the patient had no symptoms of acute pericarditis. Cardiac MRI also provides insight into infiltrative cardiomyopathies such as amyloid heart disease. In this case, cardiac MRI showed non-specific delayed myocardial enhancement of the interventricular septum at the site of RV insertion.

Right and left heart catheterisation was performed to further investigate the primary cause of heart failure. Ventricular pacing was used to regularise the RR intervals. Mean right atrial pressure was severely elevated at 18 mm Hg with a prominent 'y' descent and lack of decrease with inspiration (positive Kussmaul's sign, [figure 3A](#)). The RV pressure showed a dip and plateau morphology with severe elevation in filling pressure (RV end-diastolic pressure 21 mm Hg). Pulmonary artery pressure was mildly elevated with a mean pulmonary artery pressure of 34 mm Hg. Pulmonary artery wedge pressure (PAWP) was severely increased at mean 24 mm Hg. LV pressure also showed a dip and plateau morphology with severely elevated filling pressure (LV end-diastolic pressure 21 mm Hg). On simultaneous LV and PAWP assessment, there was no evidence of intrathoracic-intracardiac pressure dissociation ([figure 3B](#)). There was only minimal enhancement of ventricular interdependence on simultaneous LV and RV pressure tracings ([figure 3C](#)). The lack of dynamic respiratory changes, indicating the absence of intrathoracic and intracardiac pressure dissociation, and only mild enhancement of ventricular interdependence were not classic findings for CP. However, the early rapid filling and elevation with equalisation of diastolic pressures in the cardiac chambers were unexplained. While this finding is also seen in restrictive cardiomyopathy, the thorough imaging evaluation did not reveal significant myocardial abnormality.

TREATMENT

Overall, the investigations were suggestive, but not diagnostic for CP. Thus, the patient was referred for 'diagnostic' pericardiectomy. On the intraoperative inspection, the pericardium was inflamed and thickened (up to 5 mm). Central venous pressure decreased from 22 to 12 mm Hg following total pericardiectomy. Pathology showed moderate to marked pericardial fibrosis and mild focal non-granulomatous plasmacytic infiltrates ([figure 4A–C](#)).

OUTCOME AND FOLLOW-UP

The patient reported that he felt 'almost 100%' improved at 4 months and continued to do well at 1-year follow-up.

DISCUSSION

Classic signs of CP include volume overload, jugular venous distension with prominent 'x' and 'y' descents and Kussmaul's sign. Chest X-ray may show pericardial calcification, although chest CT is superior for assessment of pericardial calcification, pericardial thickening and deformation of the ventricles by the compression of the pericardium. On echocardiography, respiratory interventricular septal shift, respirophasic changes in mitral inflow velocities, expiratory diastolic flow reversal in the hepatic

Figure 4 Pericardial pathology specimens. (A) Gross pathology specimens. Parietal pericardial thickness is increased (normally <1 mm). (B) Low-power full-thickness view of pericardium (the outer surface at the top). (C) High-power views of chronic lymphoplasmacytic infiltrates.

veins and preserved early diastolic mitral annular velocity can confirm the diagnosis of CP with a high specificity such that many patients undergo pericardiectomy without need for haemodynamic catheterisation. Welch *et al* studied test

performance of five criteria for diagnosis of surgically confirmed CP at Mayo Clinic.¹ The sensitivities of the studied criteria are as follows: ventricular septal shift: 93%; change in mitral E velocity $\geq 14.6\%$: 84%; medial e' velocity ≥ 9 cm/s: 83%; medial e' /lateral e' ≥ 0.91 : 75%; and hepatic vein ratio in expiration ≥ 0.79 : 76%. The presented patient met only two of the five criteria, namely preserved medial e' velocity and reversal of hepatic vein ratio. However, the preserved medial e' pointed towards normal myocardial relaxation and was a clue for CP and not myocardial disease. In cases where echocardiography yields equivocal results, haemodynamic catheterisation is considered to be a gold standard and to provide a definitive diagnosis. Talreja *et al*² studied test characteristics of haemodynamic criteria considered classic for CP such as the inspiratory decrease in right atrial pressure < 5 mm Hg (sensitivity 71%, specificity 37%), difference between LV and RV end-diastolic pressures ≤ 5 mm Hg (sensitivity 46%, specificity 54%), ratio of RV end-diastolic to systolic pressure $> 1/3$ (sensitivity 93%, specificity 46%) and pulmonary artery systolic pressure < 55 mm Hg (sensitivity 90%, specificity 29%). The most valuable haemodynamic criterion to distinguish CP from restrictive cardiomyopathy was the systolic area index, defined as the ratio of the RV area (mm Hg $\times$ s) to the LV area (mm Hg $\times$ s) in inspiration versus expiration. The cut-off value of > 1.1 had 97% sensitivity, 100% specificity, 100% positive predictive accuracy and 95% negative predictive accuracy for CP. However, the calculation of systolic area index is not routinely calculated in our catheterisation laboratory.

However, in a minority of patients, the diagnosis of CP cannot be made with certainty despite echocardiography and haemodynamic catheterisation. This may be a case when there is mixed pathology of both CP-restrictive cardiomyopathy, for example, in patients with history of chest radiation. Given the potential of curable heart failure due to CP, diagnostic pericardiectomy may need to be considered in selected patients with clinical suspicion for CP.

The most common causes of CP in developed countries are iatrogenic (prior cardiac surgery and radiation) and idiopathic pericarditis. The aetiology of pericardial constriction in this case

remains uncertain. In this unique patient, typical haemodynamic findings of ventricular interaction were potentially masked by the septal hypertrophy due to previously undiagnosed hypertensive heart disease.³ However, the patient did have the classic findings of elevated diastolic pressures with early rapid filling. Additional important clues to the correct diagnosis were a preserved e' velocity of the septal mitral annulus on tissue Doppler imaging (11 cm/s) and pericardial thickening and ongoing inflammation. These findings raised a high index of suspicion for CP prompting surgical exploration.

Learning points

- Unexplained cause of heart failure, with findings suggestive of, though non-diagnostic for CP, may require pericardiectomy to make final diagnosis.
- Hypertensive cardiomyopathy may mask classic findings of CP.
- The diagnostic pericardiectomy may be curative.

Contributors EK wrote the manuscript. WE collected pathology pictures and pathology manuscript. JG involved in manuscript writing and mentorship. BG participated in mentorship.

Competing interests None declared.

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