

A case of recurrent pericardial constriction presenting with severe pulmonary hypertension

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ABSTRACT

Chronic constrictive pericarditis (CP) is a relatively rare condition in which the pericardium becomes fibrotic and noncompliant, eventually resulting in heart failure due to impaired ventricular filling. The only curative treatment is pericardiectomy. Classically, CP does not usually cause severe pulmonary hypertension. When attempting to differentiate CP from restrictive cardiomyopathy, the presence of severely elevated pulmonary arterial pressure is used as a diagnostic criterion ruling against CP. We present a case of proven recurrent pericardial constriction following pericardiectomy presenting with severe pulmonary hypertension.

Key Words: constrictive pericarditis, heart failure, pericardiectomy, pulmonary hypertension

CASE REPORT

A 78-year-old man was admitted to the pulmonary hypertension service at Stanford Hospital in December, 2010 with elevated pulmonary arterial pressures and progressive right-sided heart failure. He reported progressive shortness of breath over a period of two months, culminating in dyspnea at rest. This was associated with a 20-pound weight gain, abdominal bloating, and leg edema.

He was first evaluated for dyspnea several years prior to admission, where he was diagnosed with bilateral pleural effusions and extensive pericardial calcification on chest computed tomography (CT). Echocardiography and cardiac catheterization were consistent with constrictive pericarditis. He subsequently underwent uneventful complete pericardiectomy in 2008. The pericardium was stripped from phrenic nerve to phrenic nerve and the diaphragmatic and posterior pericardium was removed as well.

The patient enjoyed symptomatic improvement for approximately one year but then developed recurrent dyspnea. An echocardiogram demonstrated right ventricular (RV) pressure overload and a right ventricular systolic pressure of 66 mmHg. A trial of sildenafil was

initiated but was discontinued after the patient developed a marked worsening of his dyspnea.

His medical history was remarkable for pericarditis diagnosed in 1978, which was presumed to be viral in origin. He had permanent atrial fibrillation/atrial flutter which did not respond to an attempted atrial flutter ablation in 2003. He had a history of chronic hypertension and gradually progressing stage 3-4 chronic renal impairment.

On admission, his jugular veins were distended to angle of his jaw when upright. Pitting edema was noted up to the mid thighs bilaterally. Crepitations were heard on auscultation of his lungs. N-terminal pro-brain natriuretic peptide was measured to be greater than 30,000 pg/mL. Chest x-ray showed blunting of the costophrenic sulci with coarse reticular linear basilar parenchymal opacities (Fig. 1).

Prior to admission, ventilation-perfusion scan was low probability for pulmonary embolism. Neither pulmonary function tests nor screening blood work revealed any other potential cause for his pulmonary hypertension.

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Echocardiogram on admission was technically difficult. Moderate RV enlargement and dysfunction were identified. Right ventricular systolic pressure was severely elevated at 82 mmHg. No findings of pericardial constriction were reported.

Cardiac catheterization was performed two days post admission after the patient had received intravenous diuresis. Pulmonary artery pressures were elevated at 70/13 mmHg with a mean of 35 mmHg. The pulmonary capillary wedge pressure was 13 mmHg. Cardiac output as calculated by the Fick equation was preserved at 5.3 L/min and pulmonary vascular resistance was calculated to be 4.3 WU. The pulmonary pressures did not respond to vasodilator challenge with inhaled nitric oxide at a dose of 20 ppm. The right atrial pressure tracing (Fig. 2A) showed abrupt x and y descents, and no fall in right atrial pressure with inspiration (positive Kussmaul's sign). Simultaneous pressure measurements of the RV and left ventricle (LV) revealed an early diastolic "dip and plateau" and equalization of end diastolic pressures (Fig. 2B). However, no evidence of discordant change in

RV and LV pressures was seen with respiration. Thus, the catheterization was supportive but not diagnostic for pericardial constriction. Volume challenge was not performed. Cardiac biopsies were performed at the time of the catheterization and showed only nonspecific changes.

Cardiac magnetic resonance imaging (MRI) was performed on a 1.5 T GE scanner, with no contrast given secondary to the patient's renal insufficiency. The main pulmonary artery, inferior vena cava and right atrium were found to be dilated. Residual thickened pericardium was identified in the basolateral and basal posterolateral segments (Fig. 3), with adhesive segments, most prominent in the basal anterior segment of the RV and the basal posterolateral segment of the LV. Septal flattening occurred on inspiration (Fig. 4A) and resolved completely on expiration (Fig. 4B), revealing heightened ventricular interdependence. These findings confirmed the diagnosis of recurrent pericardial constriction.

Given his significant comorbidity and the high-risk nature of repeat pericardiectomy, the patient was managed medically



Figure 1: PA chest x-ray demonstrating cardiomegaly, bilateral opacities, and pleural effusions.

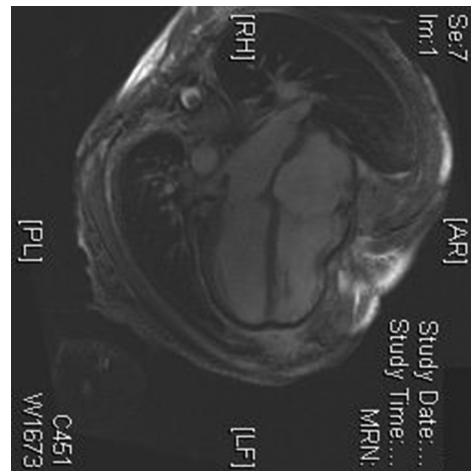


Figure 3: Cardiac MR Fiesta 4 chamber view demonstrating residual pericardial thickening and tethering in the basal posterolateral region of the left ventricle.



Figure 2: (A) Hemodynamic tracing from right atrium demonstrating abrupt x and y descents and no decline in pressure with inspiration. (B) Simultaneous hemodynamic tracings from left and right ventricle demonstrating early diastolic "dip and plateau" and equalization of end diastolic pressures.

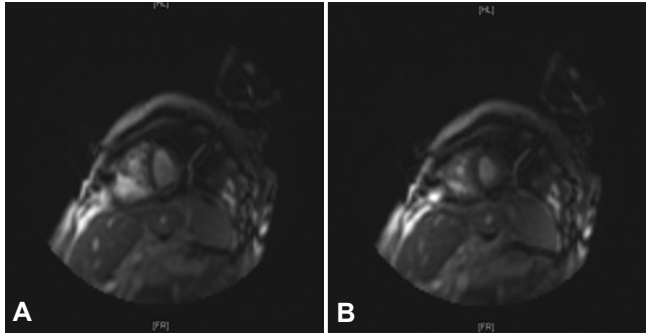


Figure 4: (A) Real-time cardiac MR. Short axis view at end inspiration showing flattening of the interventricular septum. (B) Real-time cardiac MR. Short axis view at end expiration showing normalization of the septal position.

with diuresis. He responded well to furosemide and was clinically euvolemic with improvement in his dyspnea on discharge.

DISCUSSION

Constrictive pericarditis must always be considered in the workup of occult heart failure with normal ejection fraction. While it may result from virtually any inflammatory process of the pericardium, the most common etiologies in North America include viral pericarditis, followed by cardiac surgery and mediastinal radiation injury.^[1] In constrictive pericarditis (CP), the pericardium becomes progressively fibrotic and noncompliant, impeding ventricular filling.

Clinical symptoms are often vague and develop insidiously. The most common clinical presentation is chronic heart failure.^[1] Patients typically complain of fatigue, dyspnea, weight gain, abdominal discomfort, increased abdominal girth, and edema.^[2,3] On examination, the jugular venous pressure is elevated and increases or does not fall upon inspiration (Kussmaul's sign). Both x and y descents are prominent. Cardiac auscultation may reveal a pericardial knock.^[3] Ascites and hepatomegaly occur early in the disease course, preceding the development of significant peripheral edema.^[2]

The diagnosis can be suggested on chest x-ray by the presence of pericardial calcifications, which are present in approximately 50% of cases.^[4] Pleural effusions and signs of pulmonary venous congestion are also common. Findings on two-dimensional echocardiography include abrupt motion of the interventricular septum during early diastole ("septal bounce") and signs of venous congestion. Doppler echocardiography reveals rapid early diastolic ventricular filling with an abrupt decrease in late diastolic filling.^[4] Heightened ventricular interdependence as shown by a $\geq 25\%$ increase in early diastolic mitral inflow velocity with expiration has a sensitivity and specificity of 85-90%

for CP, and is useful in the differentiation from restrictive cardiomyopathy.^[5]

Cardiac CT and MRI are also useful diagnostic modalities. Both modalities can detect pericardial thickening, which is supportive for CP. As in our case, cine images can show signs of abrupt cessation of diastolic filling and ventricular interdependence diagnostic of CP.^[4,6]

Cardiac catheterization has traditionally been employed to confirm constrictive physiology and to differentiate from restrictive cardiomyopathy. Both RV and LV pressures decrease rapidly in early diastole followed by an abrupt plateau ("dip and plateau" or "square root sign"). Classical features which favor CP over restrictive cardiomyopathy include equalization of left and right heart filling pressures to less than 5 mmHg, heightened ventricular interdependence with respiration, and only modestly elevated pulmonary artery systolic pressures (less than 50 mmHg).^[7] Diuresis may mask these findings, and rapid volume challenge with normal saline can improve the sensitivity of catheterization for the diagnosis of CP.

The treatment for symptomatic chronic CP is pericardiectomy. This is a technically challenging procedure and carries a mortality rate of 6-12%, with worse outcomes for more severely symptomatic patients.^[1,7] Thus, it is not recommended for mildly symptomatic or end stage patients. Complete normalization of cardiac hemodynamics is only seen in 60% of patients postoperatively.^[7] Recurrent constriction may be secondary to incomplete removal of the pericardium, immobilization, or myocardial fibrosis or atrophy. In our case, the MRI reveals that incomplete removal of the pericardium was at least partially responsible for the recurrence.

The performance of pulmonary artery systolic pressure exceeding 50 mmHg in distinguishing restrictive cardiomyopathy (RCM) from CP has not been ideal when used in isolation. In older series comparing surgically proven CP to RCM, significant overlap in pulmonary arterial systolic pressure (PASP) between the two entities was seen.^[8] In a more recent series, there was no statistically significant difference in pulmonary pressures at catheterization between CP and RCM, and $PASP < 55$ mmHg had a specificity of only 29% for the diagnosis of CP.^[9] The potential for the development of severe pulmonary hypertension due to any cardiac condition resulting in elevated LV filling pressures is well-documented.^[10] There is no pathophysiologic reason why CP could not lead to pulmonary hypertension of similar magnitude by the same mechanism.

There is a paucity of evidence regarding the optimal medical management of pulmonary hypertension secondary to CP. Treatment of these patients with pulmonary vasodilators

is hazardous given their potential for increasing LV-filling pressures further, thus worsening pulmonary edema. The deterioration our patient experienced after the initiation of sildenafil suggests this may have occurred.

Our case clearly documents significantly elevated pulmonary pressures on direct measurement in association with unequivocal constrictive physiology on cardiac MRI. The hemodynamic assessment did not show the same enhanced ventricular interdependence seen on MRI, yet our sensitivity may have been reduced by low filling pressures secondary to diuresis. The inconclusive nature of the echocardiogram was likely secondary to the technical limitations of the study. Though a mixed constrictive/restrictive state is technically possible, little evidence for restrictive cardiomyopathy was seen on cardiac imaging, hemodynamics, or biopsy. Moreover, an alternative cause for the patient's pulmonary hypertension could not be identified. This case illustrates the complex nature of chronic constrictive pericarditis and warns against excluding a diagnosis of CP solely on the basis of severely elevated pulmonary artery pressures in the absence of other findings.

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