

## CLINICAL VIGNETTE

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# Constrictive Pericarditis Presenting with Recurrent Pleural Effusions

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### *Introduction*

Constrictive pericarditis is a rare condition that requires a high index of clinical suspicion to make the diagnosis. The etiology of constrictive pericarditis varies widely depending on the population studied, with idiopathic or post-treatment (eg, post-operative or post-radiation) causes being more prevalent in developed countries, while infectious etiologies are more predominant in developing countries.<sup>1</sup> However, in the absence of risk factors, constrictive pericarditis should still be considered in the differential diagnosis if there is clinical suspicion. The disease can present with symptoms of heart failure, such as lower extremity edema, paroxysmal nocturnal dyspnea (PND), and dyspnea. However, despite the known association of pleural effusion with constrictive pericarditis, the diagnosis is often overlooked for more common etiologies including heart failure, pulmonary embolism, cirrhosis, pneumonia, or malignancy. A high index of suspicion is required because diagnostic studies may not always reveal all the features that are classically associated with constrictive pericarditis. We present a case of a young patient without history of cardiothoracic surgery, chest radiation, or obvious infection who presents with recurrent bilateral pleural effusions. Although diagnostic studies were equivocal for constriction, the diagnosis was eventually confirmed on final surgical pathology. We suggest that when evaluating patients with heart failure symptoms that are not well explained constrictive pericarditis should be considered in the differential. This is especially important with recurrent pleural effusions.

### *Case Presentation*

A 23-year-old male with non-ischemic cardiomyopathy and polysubstance abuse presented for evaluation for lower extremity edema and recurrent pleural effusions. He was diagnosed with heart failure, which was believed to be secondary to past methamphetamine use. His left ventricular ejection fraction (LVEF) on prior transthoracic echocardiogram was reportedly 20%, but was found to be 55% on repeat testing at our institution. He also reported recurrent pleural effusions requiring more than 20 prior therapeutic thoracenteses. He underwent VATS two years ago to assess etiology of recurrent pleural effusions, but the procedure was complicated by cardiac arrest, which he required placement of an automatic implantable cardioverter defibrillator (AICD). His home medications included carvedilol 12.5mg twice a day, furosemide 20mg daily, and methadone 5mg three times a day. On physical exam, he had decreased breath sounds over left lower lobe with

crackles at bilateral bases. There was no significant jugular venous distension or Kussmaul's sign and cardiac exam was otherwise unremarkable without lower extremity edema. Admission labs included hemoglobin 11.9, platelets 141, sodium 134, alkaline phosphatase 150. Troponin and BNP were not elevated.

Electrocardiogram (ECG) showed sinus rhythm, borderline right axis deviation, with nonspecific T-wave abnormalities in the lateral pericardial leads. Chest x-ray showed an implantable cardiac device overlying the left chest with epicardial pacing leads, congestion of the pulmonary vasculature and small bilateral pleural effusions with pleural fluid extending within the intralobar fissure on the right. Transthoracic echocardiogram showed a normal left ventricle (LV) size with ejection fraction of 60% and normal diastolic function. The right ventricle (RV) was grossly normal in size and function. Estimated right ventricular systolic pressure was 26mmHg. CT chest showed bilateral pleural effusions.

Patient underwent pigtail catheter placement in the right pleural space with drainage of 1700 milliliters of clear yellow fluid. Sampling of pleural fluid was consistent with transudative fluid per Light's criteria. Right heart catheterization (RHC) revealed the following:

Right atrial pressure 18mmHg  
Right Ventricle pressure 40mmHg/18mmHg with end diastolic pressure 22mmHg  
Pulmonary artery pressure 40mmHg/23mmHg  
Pulmonary capillary wedge pressure 20mmHg  
Left Ventricle pressure 90mmHg/20 mmHg with an end diastolic pressure of 14mmHg

A possible square root sign was noted in the right ventricle tracing indicating a rapid rise in filling pressures after diastole. Some respiratory variation was noted in the pressures. There was evidence of discordance between right and left sided pressures suggesting constrictive physiology, but the findings were not conclusive.

Given inconclusive findings, a cardiac CT demonstrated mild prominence of the pericardium, with some areas of the pericardium with thickness of more than 4mm, highly suggestive of constriction.

The patient underwent surgical pericardectomy with findings notable for thickened pericardium over diaphragm and scar tissue around the pulmonary veins and inferior vena cava (IVC). Samples of pericardial tissue removed from procedure were sent to pathology, revealing pericardial and fibroadipose tissue with fibrosis with chronic inflammation. The patient's postoperative course was uncomplicated.

### Discussion

Constrictive pericarditis is a difficult clinical diagnosis to make. A high index of suspicion, multiple imaging modalities and hemodynamic data should be considered. Physical exam findings include elevated jugular venous pressure, in 93% of confirmed cases, pericardial knock, in 47% of cases, and pulses paradoxus, present in less than 20% of cases.<sup>2</sup> However, most of these findings are not specific. Kussmaul's sign, which is classically associated with constrictive pericarditis, can also be seen with restrictive physiology. Echocardiography can show increased pericardial thickness, abnormal septal motion, bi-atrial enlargement, dilated IVC and dilated hepatic vein with constrictive pericarditis. However, as this case demonstrates, some patients will have none of these findings.

The main hemodynamic features of constriction include equalization of the left and right sided pressures due to a limitation of the total volume that can be accommodated in the pericardial sac. This results in a rapid ventricular filling, "rapid y descent," followed by a sudden rise in pressure. Exaggerated ventricular longitudinal contraction results in an exaggerated x descent during ventricular systole. This results in elevated end diastolic pressures with the "square root" sign finding in hemodynamic data. Enhanced ventricular interdependence is also a result of the limited pressure that can be accommodated within the pericardium. As a result, the right ventricular (RV) pressure increase with inspiration results in a decrease in left

ventricular (LV) pressure. Our patient demonstrated some of these features, including elevated filling pressures with near equalization between the left and right side. His hemodynamic data demonstrated a possible square root sign and some discordance, though were not conclusive.

In a series of 143 patients with surgically confirmed constrictive pericarditis, 78 underwent cardiac catheterization. Of those, the mean right atrial pressure was 21mmHg and the square root sign was not seen in 23 percent of patients.<sup>3</sup> Furthermore, diastolic and equalization of pressures was absent in 19 percent of patients, while respiratory variation in the RV-LV pressure relationship was only seen in 44 percent of patients with confirmed pericarditis.<sup>3</sup>

While pleural effusions are common in constrictive pericarditis, constrictive pericarditis is frequently overlooked as a potential cause. Up to 55% of cases of constrictive pericarditis were associated with pleural effusions.<sup>4,5</sup> The pleural effusions known to be associated with constrictive pericarditis are usually left-sided at presentation.<sup>6</sup> As a result, the absence of unilaterality in pleural effusions can sometimes decrease the suspicion of constrictive pericarditis, skewing clinical decision making towards more common causes bilateral effusion such as heart failure, as in our patient, or chronic liver disease.

Given the variation in findings that may be evident in RHC with clinical suspicion of constrictive pericarditis, endomyocardial, or less commonly, pericardial biopsy may be useful to establish the diagnosis, especially when hemodynamic and imaging studies are not confirmatory.<sup>7</sup> However, the clinical assessment and correlation of all data is paramount to making the diagnosis. The fibrinous deposition with inflammatory cells, in conjunction with clinical signs of constrictive pericarditis found in this case is highly indicative of constrictive pericarditis as the unifying diagnosis.



Concurrent measurement of the LV and PA pressures demonstrating elevated filling pressures. Some evidence right and left heart pressure discordance is noted with the LV systolic pressure in fifth beat decreasing while the PA systolic pressure increases.

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