Chronic Constrictive Pericarditis Presenting as Ascites in a Young Male

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Abstract

Constrictive pericarditis is an uncommon cause of right-sided heart failure. We present a case of idiopathic constrictive pericarditis presumably following a viral episode of acute pericarditis several months prior to the patient's presentation. This case highlights the necessity for a high clinical index of suspicion. Importantly, a series of imaging modalities were required to confirm the diagnosis and lead to a successful surgical intervention.

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Abstract: Constrictive pericarditis is an uncommon cause of right-sided heart failure. We present a case of idiopathic constrictive pericarditis presumably following a viral episode of acute pericarditis several months prior to the patient's presentation. This case highlights the necessity for a high clinical index of suspicion. Importantly, a series of imaging modalities were required to confirm the diagnosis and lead to a successful surgical intervention.

Clinical Vignette: A 31-year-old man presented to the emergency department with progressively worsening abdominal swelling and unintentional weight loss for five months. He denied any fever, chills, night sweats, chest pain, shortness of breath, palpitations, orthopnea, paroxysmal nocturnal dyspnea, or syncope. The patient had a history of generalized anxiety and major depression. He was not on any medications and denied drug or alcohol use. He was adopted and unable to provide any family history.

On presentation his blood pressure was 105/68 mmHg and heart rate was 106 beats per minute. On physical examination his abdomen was non-tender but distended with shifting dullness. His cardiac exam revealed normal S1/S2 heart sounds with no murmur, rub, or gallop. His lungs were clear to auscultation. There was trace pitting edema of his lower extremities and jugular venous distension (JVD) 3 centimeters above his clavicle. We were unable to appreciate prominent waveforms or x/y descents and an electrocardiogram showed a normal sinus rhythm.

Abdominal ultrasound was suggestive of cirrhosis with large volume ascites. A diagnostic and therapeutic paracentesis was performed with 11 liters removed. His peritoneal fluid studies yielded an albumin of 2.8g/dL and serum ascites albumin gradient (SAAG) of 1.5g/dL. While we initially suspected cirrhosis his high protein, high SAAG ascitic fluid shifted our differential to include a cardiac cause of his ascites as well.

A transthoracic echocardiogram showed a left ventricular ejection fraction (LVEF) of 50-55% and thickened pericardium (Figure 1). There was evidence of ventricular interdependence with resulting respiratory variation in a ortic flow velocity (Video 1). Medial mitral annular e' velocity was 27.4 cm/s and hepatic vein expiratory diastolic reversal ratio was 0.82(Figure 2A and 2B).

Cardiac MRI was subsequently completed which redemonstrated a thickened pericardium, measuring 6mm, without evidence of a pericardial effusion or calcification. A prominent diastolic interventricular septal bounce and LVEF of 53% was noted on Cardiac MRI as well (Video 2). He was diagnosed with chronic constrictive pericarditis and scheduled for pericardiectomy. However, the etiology for his pericardial disease was unclear. A purified protein derivative (PPD) skin test was non-reactive, and he had no previous surgeries or radiation.

In the operating room his thickened pericardium was directly visualized, and pericardial stripping was performed (Figure 3). Specimens were then sent for histopathologic examination (Figure 4) which ultimately revealed the cause of his constrictive pericarditis to be idiopathic (most likely post-viral).

The patient recovered rapidly after pericardial stripping and was discharged home four days later. At three-month follow-up he had complete resolution of his ascites.

Discussion: Constrictive pericarditis arises secondary to chronic inflammatory changes resulting in fibrous thickening of the pericardium. Scarring can progressively restrict ventricular filling past early diastole.¹ Clinical presentation can vary but patients tend to complain of chronic symptoms secondary to volume overload including peripheral edema, dyspnea on exertion, or worsening abdominal distension as seen in this case.

The first line imaging modality for patients with suspected constrictive pericarditis is transhoracic echocardiogram according to 2015 European Society of Cardiology Guidelines.² Key imaging findings include ventricular interdependence – a result of non-compliant pericardium preventing diastolic ventricular filling. Normally inspiration decreases intrathoracic pressure allowing inflow of blood into both ventricles. In constrictive pericarditis, inspiration results in decreased intrathoracic pressure without significant change in the intracardiac pressures. Intrathoracic-intracardiac dissociation leads to right ventricular (RV) expansion and shifts the interventricular septum to the left as the stiff pericardium limits left ventricular (LV) filling. In expiration the septum shifts towards the right lowering the RV's ability to fill. Additional echocardiographic findings suggestive of constrictive pericarditis include expiratory hepatic vein diastolic flow reversal ratio ([?] .79 cm/s) and elevated medial e' velocities ([?] 9cm/s). Each of these findings are independently associated with the diagnosis of constrictive pericarditis and were seen in our patient. The Mayo Clinic Criteria for echocardiographic evidence of constrictive pericarditis found the combination of these three variables had a diagnostic specificity of 97% for constrictive pericarditis.³

Accordingly, ventricular interdependence seen on echocardiogram can be demonstrated on cardiac catheterization. Simultaneous RV and LV pressure tracings with discordant respiratory changes are pathognomonic, and remain the most sensitive and specific finding, for constrictive pericarditis.⁴ As compared to a restrictive cardiomyopathy, or even a normal heart, concordant respiratory variation is seen instead.

Although direct visualization of pressure tracings is classically the gold standard when confirming constrictive pericarditis, it no longer remains a first-line method for diagnosis. Cardiac MRI has evolved into a more comprehensive imaging modality. Not only does it allow for visualization of the pericardium, but also the impact on the structure and function of the myocardium. Real-time imaging during free breathing can evaluate hemodynamics during cardiac filling which emulates what we elicit with cardiac catheterization. Over time Cardiac MRI has presented a viable non-invasive test to contrast catheterization as the gold standard for diagnosis. Another novel approach, Biderman et al. described the utility of a tagged myocardium study measuring visceral-parietal pericardial adherence to determine constriction offering another alternative method when trying to confirm an elusive diagnosis such as constrictive pericarditis.⁵

Constrictive pericarditis remains a difficult, but important diagnosis given surgical interventions available for treatment. The use of multiple imaging modalities allowed us to confirm the diagnosis of idiopathic chronic constrictive pericarditis followed by successful surgical intervention.

Figure Legend

Figure 1: Marked thickening of pericardium (arrow)

Figure 2A: Demonstrating medial E' velocity of 27.4cm/s, exceeding Mayo Clinic criteria of 9.0 cm/s for constrictive pericarditis

Figure 2B: The hepatic venous index of diastolic reversal velocity divided by systolic forward velocity equals 0.82 (NR = .78)

Figure 3: The pericardium was dense and fibrotic completely encapsulating the heart. At the beginning of the procedure, the central venous pressure (CVP) was 25mmHg. After the pericardium was incised to the level of the pericardial fat and removed the CVP was 15 mmHg.

Figure 4: Histopathologic examination revealed dense pericardial fibrosis with minimal microscopic calcification and epicardial neovascularization consistent with constrictive pericarditis.

Video 1: Transthoracic echocardiogram showing ventricular interdependence

Video 2: Cardiac MRI with evidence of septal bounce and pericardial thickening

Learning Objectives:

To illustrate the classic findings of chronic constrictive pericarditis in an otherwise healthy young male.

To emphasize that in patients with unexplained right-sided heart failure symptoms constrictive pericarditis should be considered as a strong diagnostic possibility.

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