CLINICAL VIGNETTE

A Case of Constrictive Pericarditis

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Case Report

A 62-year-old male presented with persistent cough and dyspnea for the past year. He was a recreational cyclist and cycled about 45 miles/week with no difficulties prior to symptom onset. He also noted a rash, initially felt due to fish allergy and treated with steroids. Symptoms worsened on steroids and he developed weight gain of 15# and lower extremity edema.

He was evaluated by local pulmonary and cardiology specialists and was found to have large bilateral pleural effusions and a small pericardial effusion on chest CT. He was treated with furosemide and underwent thoracentesis with transient mild improvement. The pleural effusions and dyspnea recurred, prompting thoracic surgery referral and right sided pleurodesis with resolution of the large right pleural effusion. Pleural fluid showed lymphocytic predominance and pleural biopsy showed nonspecific chronic pleuritis.

His dyspnea and edema recurred and he was referred to UCLA Health for a second opinion. Rheumatology and infectious disease evaluations were unremarkable. Initial cardiology evaluation revealed a thin male in no distress. Vital Signs included BP 122/74, HR 74/min, with 98% oxygen saturation on room air. Exam was remarkable for elevated jugular venous pressures, and no pulsus paradoxus. Lungs were clear and he had a grade 2/6 holosystolic murmur at the left sternal border. The remainder of the exam was unremarkable except for mild bilateral lower extremity edema.

Echocardiogram showed normal systolic LV function with EF 55-60%, type II diastolic dysfunction, moderate tricuspid regurgitation, and evidence of mild pulmonary hypertension with pulmonary arterial pressure estimated at 38 mmHg. Because of his unexplained CHF, he underwent right and left cardiac catheterization and cardiac MRI to assess for possible constrictive pericarditis. Cardiac Catheterization show normal coronary arteries and normal systolic LV function. Hemodynamic assessment showed elevated right atrial pressures with mean of 17 mmHg, pulmonary arterial pressure 35/20, and wedge pressure with mean of 17 mmHg. Simultaneous left ventricular (LV) and right ventricular (RV) pressures showed equalization of end diastolic pressure tracings. During diastole, the "square root sign" was noted on RV and LV tracings (Figure 1). There was no discordance between RV pressures and LV pressures on inspiration. Cardiac MRI showed a thickened myocardium up to 7 mm in diameter without Gadolinium enhancement. The hemodynamic findings and MRI were consistent with constrictive pericarditis.

He was seen by cardiothoracic surgery and underwent successful pericardiectomy with uneventful recovery. He had complete resolution of symptoms, was weaned off diuretics and returned to his usual vigorous cycling.

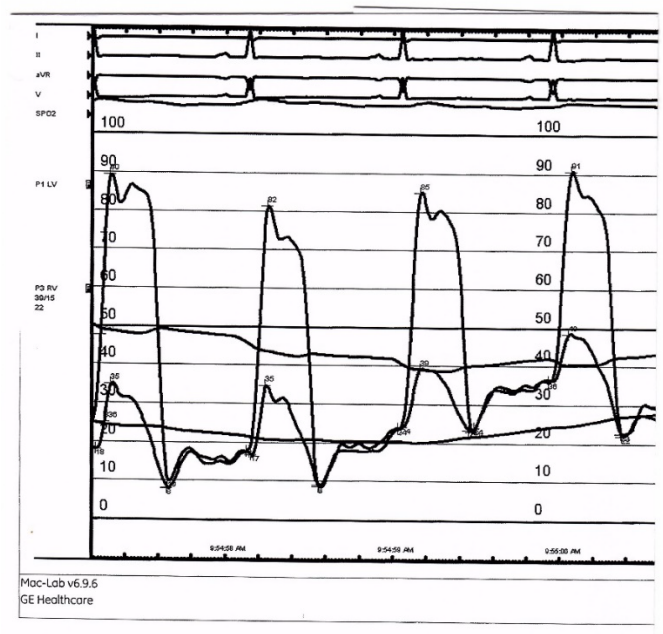

Figure 1

Discussion

Constrictive pericarditis is a disorder of diastolic filling of the heart. The normal pericardium is an elastic sac that houses the heart. The elastic pericardium expands with diastolic filling of the heart. With constrictive pericarditis, the pericardium loses it elasticity and constrains the heart filling during diastole. The limited filling causes decrease in stroke volume and cardiac output and results in congestive heart failure. Constrictive pericarditis may be difficult to diagnose. It should be considered in patients with preserved LV systolic function when standard CHF treatment does not improve or control symptoms.

The incidence of constrictive pericarditis varies by etiology. Post-procedure studies report incidence of 0.48% for viral/

idiopathic pericarditis while the non-idiopathic group (connective tissue disease, neoplastic, tuberculosis and purulent) had incidence of 8.3%.[1]

Etiologies of pericarditis in the developed world are predominently idiopathic, viral, post cardiac surgery and post radiation therapy. Less common causes include connective tissue disease, tuberculosis, non-viral infectious pericarditis, trauma, malignancy, uremic, sarcoidosis and drug induced pericarditis.[2,3] In non-developed countries TB is a common cause and should be considered in patients from endemic areas.

Constrictive pericarditis presents with right heart failure symptoms. These include dyspnea and dyspnea on exertion, edema, abdominal discomfort from edema or hepatic congestion, and palpitation from associated arrhythmias.[4] Physical exam will show signs of CHF, including elevated Jugular venous pressure (JVP) with a prominent y descent on inspiration. Kussmaul sign (lack of JVP decline upon inspiration) may also be present with constrictive pericarditis. Lung exam may reveal pleural effusions. Cardiac exam may include a pericardial knock, a sound occurring in diastole prior to S3, which corresponds to a sudden termination of ventricular filling. One study reported presence of pericardial knock in 47% of patients with constrictive pericarditis.[5] Leg edema and generalized volume overload are common. Pulsus paradoxus (a drop of systolic blood pressure greater than 10 mmHg during inspiration) is reported in about 20% of patients with constrictive pericarditis.[5,6]

Initial evaluation of patient with CHF symptoms should include an EKG, labs, chest X-ray and an echocardiogram. EKG findings may be nonspecific with tachycardia, low voltage and atrial tachyarrhythmia such as atrial fibrillation. Chest x ray may show pericardial calcification, however this finding is more common with TB pericarditis. Echocardiography is a class I indication in patients with CHF and may assist in diagnosis.[7] Echocardiographic findings include thickening of the pericardium. This may be seen on transthoracic echocardiography, however less reliably than with transesophageal echocardiography which has higher resolution.[8] Moderate biatrial enlargement may be present. Respiration related dissociation between intracardiac and intrathoracic pressures are additional echocardiographic signs of constrictive pericarditis. On inspiration an abnormal septal bounce can be seen with shift of the intraventricular septum towards the left ventricle and shift back to the right ventricle with expiration.[9] Doppler assessment of the mitral inflow also shows flow variation >25% during respiration. On inspiration there is decrease in mitral inflow velocity while on expiration the mitral inflow velocity increases.[10,11] Similar respiratory flow velocity variation are noted on hepatic vein dopplers in constrictive pericarditis.[11] Another useful echo finding may be seen with tissue doppler assessment of the mitral annulus. Normal tissue doppler assessment has medial e' values lower than the lateral annular e' values. This reverses with constrictive pericarditis, especially with medial e' > 7cm/s.[12] When combined, these echocardiographic findings are highly specific, 91-97% for constrictive pericarditis with lower sensitivity of 64-87%.[13]

Constrictive pericarditis is usually associated with thickening of the pericardium, therefore imaging of the pericardium is important in the diagnosis. However, constrictive pericarditis can be seen without pericardial thickening with one case series reporting 18% of surgically confirmed patients with normal pericardial thickness.[14] CT remains useful in assessing pericardial thickness and presence of pericardial calcification. CT also provides high quality assessment of cardiac and vascular anatomy.[15]

Cardiac MRI also provides high resolution of the pericardium and cardiac anatomy. The sensitivity and specificity of MR imaging in the diagnosis of constrictive pericarditis is 88% and 100%.[16] MRI is also useful in distinguishing pericardium from pericardial fluid. Both gated Cine CT and MRI imaging can detect the abnormal septal motion seen with constrictive pericarditis.[14] Cardiac MR with late gadolinium enhancement can identify pericardial inflammation and guide treatment. Patients with pericardial inflammation may benefit from anti-inflammatory medications to lowered inflammation and occasionally lead to non-surgical resolution of constrictive pericarditis.[17,18]

Cardiac catheterization and invasive hemodynamic assessment may be required if non-invasive evaluation is indeterminate. Common hemodynamic findings include elevated right atrial pressures with a rapid y descent, which corresponds to rapid diastolic filling. Left ventricular and right ventricular pressures show equalization of diastolic filling pressures. The 'dip and plateau' or a 'square root sign' on diastolic tracing corresponds to rapid early filling with a cessation of filling caused by the external compression from the inelastic pericardium. Right ventricular pressure often shows an elevated end diastolic pressure, at least one third of the right ventricular peak systolic pressure. All of these findings may also be seen in restrictive cardiomyopathy and therefore are non-diagnostic by themselves.[19] Hemodynamic analysis during respiration relies on the dissociation between intracardiac and intrathoracic pressures seen with constrictive pericarditis. Simultaneous recordings of the right and left ventricle pressures show discordant pressures. With inspiration the right ventricular systolic pressure will increase while the left systolic pressure will fall. This respiratory interdependence is a unique finding in constrictive pericarditis and has a high sensitivity, specificity and positive predictive value.[20]

Treatment for transient or inflammatory constrictive pericarditis involves anti-inflammatory medications including non-steroidal anti-inflammatory drugs, colchicine and steroids. This approach has a reported 17% resolution without surgery.[18] Patients who responded to anti-inflammatory treatment had increased inflammatory markers and evidence of inflammation with late gadolinium enhancement on cardiac MR.[17] Treatment for chronic or non-inflammatory constrictive pericarditis remains surgical with pericardiectomy. Total pericardiectomy

is reported to have better long-term survival than partial pericardiectomy.[21] In-hospital mortality rates for pericardiectomy range from 7.1 – 18.6 %.[3,21,22] Surgical mortality is influenced by preexisting conditions, including older age, renal disease, liver dysfunction, decreased systolic LV function, pulmonary hypertension, need for cardiopulmonary bypass and atrial fibrillation. Patient with idiopathic constrictive pericarditis have the best long-term survival after surgery whereas radiation induced constrictive pericarditis patients have the worst survival rates.[22]

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