A Case of Chronic Constrictive Pericarditis

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Abstract:

Constrictive pericarditis is a condition where normal cardiac filling is constricted by an inelastic pericardial sac. This case describes a 72 year old man who was admitted with peripheral edema and scrotal swelling. Physical examination confirmed significant peripheral edema and elevated jugular venous pressure. Chest Xray and CT showed calcification of the pericardium. Echocardiography findings were consistent with constrictive pericarditis with septal shudder. Cardiac catheterisation confirmed the diagnosis of chronic constrictive pericarditis.

Case history:

A 72 year old male was admitted with a long history of shortness of breath and peripheral edema that had recently worsened.

He presented 17 years earlier with chest pain and lethargy. There was a pericardial rub on auscultation and a diagnosis of pericarditis was made. Four years later he developed dyspnoea, pedal edema, rised JVP, and abdominal distension. He had an enlarged liver and alcohol use was felt to be a possible cause. Ultrasound confirmed hepatomegaly with fatty change, prominent IVC and some ascites. Liver biopsy showed fatty infiltration with no evidence of cirrhosis. He was started on diuretics for congestive cardiac failure (CCF). Eight years after this he presented with increasing shortness of breath, pedal edema, ascites and elevated jugular venous pressure to 12 cm above the sternal angle. Echocardiogram showed good LV function and normal valvular function at that time. One year ago he developed a right sided transudate pleural effusion.

On physical examination he was healthy-appearing and afebrile, with an irregularly irregular pulse of 88/min and blood pressure of 130/80 with no paradoxus. He had features of right sided heart failure with elevated jugular venous pressure, hepatomegaly, pedal edema and scrotal edema. On auscultation an early diastolic pericardial "knock" was audible with no murmurs.

Full blood count was normal and biochemistry showed mild renal impairment.

Electrocardiogram showed atrial fibrillation with controlled ventricular rate.

Chest X-ray on admission showed right pleural effusion and also showed a ring of calcification around the heart on lateral view (figure 1a- see arrows)

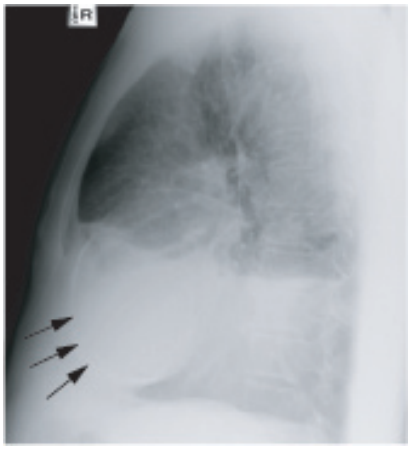


Figure 1a

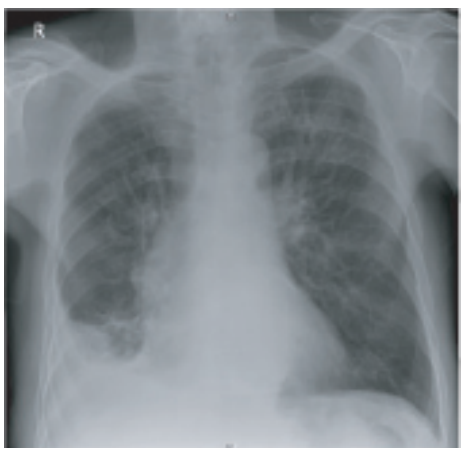
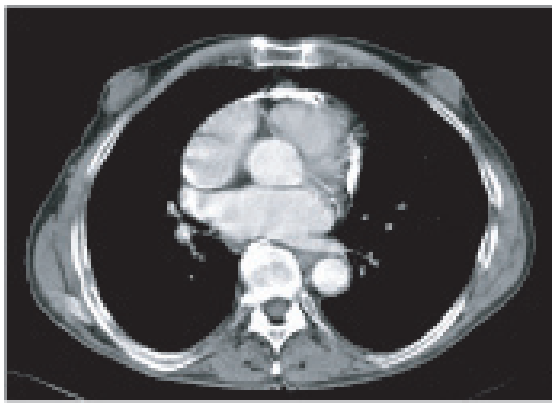
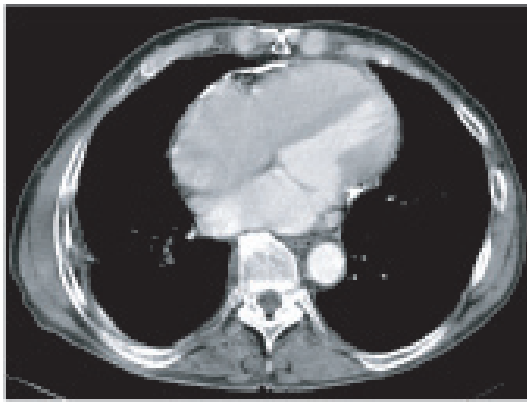


Figure 1b

CT Thorax showed pericardial calcification (see figure 2a, 2b)



(Figure 2a)

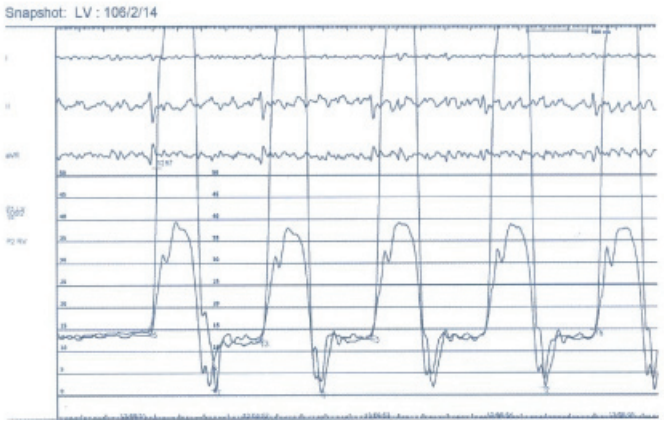


(Figure 2b)

Abdominal ultrasound showed hepatomegaly with fatty change. The hepatic veins and IVC were distended consistent with CCF.

Given these findings a diagnosis of constrictive pericarditis was suspected and an echocardiogram was requested. This showed good left and right ventricular function, a bright thickened pericardium, septal shudder, and markedly dilated veins with > 50 % collapse during inspiration. Abnormal mitral valve inflow velocities suggested pericardial constriction.

Cardiac catheterisation (see figure 3) with simultaneous LV/RV pressure tracing showed equalisation of diastolic pressures and the characteristic 'square root sign' or 'dip and plateau'- with an early diastolic dip (rapid filling of ventricles during early diastole) followed by a plateau (abrupt halt in filling due to non compliant pericardium in mid-late diastole) suggestive of constrictive pericarditis.



(Figure 3)

Discussion:

Pericardial constriction occurs when a scarred, thickened, and frequently calcified pericardium impairs cardiac filling, limiting the total cardiac volume.[1,2,3,4]

The pathophysiological hallmark of pericardial constriction is equalization of the end-diastolic pressures in all four cardiac chambers. Initial ventricular filling occurs rapidly in early diastole as blood moves from the atria to the ventricles without much change in the total cardiac volume. However, once the pericardial constraining volume is reached, diastolic filling stops abruptly. This results in the characteristic dip and plateau of ventricular diastolic pressures. The stiff pericardium also isolates the cardiac chambers from respiratory changes in intrathoracic pressures, resulting in Kussmaul's sign.

Etiology

Constrictive pericarditis is an uncommon condition with multiple etiologies. Worldwide,

tuberculosis remains the most common cause. However, in the western world, no definite etiology is established for a high percentage of cases of constrictive pericarditis, and many of these may be due to an inapparent viral infection. Postsurgical pericarditis is now an important cause of constrictive pericarditis. Other causes include chronic renal failure treated with hemodialysis, connective tissue diseases, neoplastic pericardial infiltration, incomplete drainage of purulent pericarditis, and fungal infections.[5]

Clinical features

Patients with pericardial constriction typically present with manifestations of elevated systemic venous pressures and low cardiac output.[6] Typically, there will be marked jugular venous distension, hepatic congestion with hepatomegaly, ascites and peripheral edema. The limited cardiac output typically presents as decreased exercise tolerance and may progress to cardiac cachexia with muscle wasting. In long-standing pericardial constriction, pleural effusions, ascites, and hepatic dysfunction may be prominent features.[2]

The jugular veins are distended with prominent X and Y descents. The normal inspiratory drop in jugular venous distention may be replaced by a rise in venous pressure (Kussmaul's sign). The classic auscultatory finding of pericardial constriction is a pericardial knock. This occurs as a high pitched sound early in diastole when there is the sudden cessation of rapid ventricular diastolic filling.[7]

Electrocardiogram may show low voltage QRS or non-specific T wave changes in 50% of patients and atrial fibrillation in 30% of patients.[8] Chest x ray shows pericardial calcification in up to 50% of patients, particularly on the lateral view.9 Pleural effusion is common and can be bilateral or unilateral.[5, 10]

Most patients with pericardial constriction have a thickened pericardium more than 2mm that can be imaged by CT or MRI.[2,4,11] It is important to recognize that pericardial constriction can be present without pericardial calcium. CT and MRI are also often used to delineate the thickened pericardium and to determine its extent, MRI being more sensitive.

On echocardiography, features of constriction are hard to interpret. They include pericardial thickening, paradoxical septal motion, marked variations in atrioventricular flow velocities with respiration, and absence of ventricular wall thickening.[8, 12] Doppler echocardiography is important in the evaluation of patients with suspected pericardial constriction. Doppler echocardiography frequently demonstrates restricted filling of both ventricles with a rapid deceleration of the early diastolic mitral inflow velocity (E wave) and small or absent A wave. In addition, there is substantial (>25%) respiratory variation of the mitral inflow velocity.[13]

Cardiac catheterisation is required to confirm the diagnosis. Tracings will show the characteristic early diastolic dip (rapid filling of ventricles during early diastole) followed by plateau (abrupt halt in filling due to non compliant pericardium in mid-late diastole) and respiratory variation between the left and right ventricular pressure tracings as a result of ventricular pressure interactions.[8]

Differential Diagnosis

Pericardial constriction should be considered in any patient with unexplained systemic venous congestion. Echocardiography is useful in differentiating pericardial constriction from right heart failure due to tricuspid valve disease and/or associated pulmonary hypertension.[2]

The most difficult differentiation is between pericardial constriction and restrictive cardiomyopathy (Appendix-1). Clinical manifestations of restrictive cardiomyopathy

most typically due to cardiac amyloid may be very similar to those due to pericardial constriction.[14, 15] Doppler echocardiography is the most useful method to distinguish constriction from restriction. Patients with pericardial constriction have marked respiratory variation (>25%) of mitral inflow, whereas this is not present in restrictive cardiomyopathies.[13]

The tissue Doppler measurement of mitral annular velocities is useful in distinguishing constriction from restriction. The early diastolic mitral annular velocity (Ea) is almost always reduced in patients with myocardial restriction, whereas it remains normal in patients with pericardial constriction.[16, 17, and 18] The optimal discrimination occurs with an Ea velocity of 8 cm/s. Similarly, rapid propagation of early diastolic flow to the apex is preserved in constriction and reduced in restriction. A slope >100 cm/s of the first aliasing contour in the colour M-mode best distinguishes the two.[16]

It has recently been reported that patients with pericardial constriction have only minimally elevated B-type natriuretic peptide (<200 pg/mL), whereas the B-type natriuretic peptide levels are typically markedly increased in patients with restrictive cardiomyopathy (>600 pg/mL).[19]

Traditionally, constriction and restriction were differentiated at cardiac catheterization by hemodynamic criteria. In constriction, there is a usually almost exact equalization of late diastolic pressure in both the right and left heart. With restriction, typically left ventricular end-diastolic pressure exceeds right ventricular pressure by at least a few mm Hg. Pulmonary hypertension is frequently seen with restriction but is not typically present with constriction. Thus, right ventricular diastolic pressure should be more than one third of the right ventricular systolic pressure in pericardial constriction.

It should be recognized that the aforementioned classic hemodynamic criteria have limited specificity (24% to 57%) in distinguishing pericardial constriction from cardiomyopathies.[20] By contrast, dynamic respiratory variations indicating increased ventricular interdependence are superior. In constriction during inspiration, right ventricular systolic pressures increase, while left ventricular systolic pressure decreases. The inverse occurs during expiration. This finding had >90% sensitivity and specificity in recognizing constrictive pericarditis versus restriction in a series of 36 patients from the Mayo Clinic.[20]

Treatment

The definitive treatment is pericardectomy with wide resection of both the visceral and parietal pericardium. This is recommended by the European Society of Cardiology.[1] Functional results are excellent in most patients who are suitable for the operation. However, even with selected cases surgical mortality is 6-12%.[1] This proportion may be reduced by excluding patients with extensive myocardial fibrosis or atrophy. The risks and benefits of surgery should be carefully considered, and a conservative approach may be more appropriate, particularly in elderly patients.[8, 21]

In some patients surgery does not immediately restore normal cardiac function, which may require some time after removal of the constricting pericardium to return to normal. The largest surgical series from the Mayo Clinic and the Cleveland Clinic indicate that patients with constriction due to idiopathic or viral pericarditis do best and patients with radiation-induced constriction fare poorly after surgery.[22, 23]

Conclusion

1. Constrictive pericarditis is a rare condition.

2. Symptoms are often non-specific, so that diagnosis can be delayed for 12-18 months after the onset of symptoms and it is not until the jugular venous pressure is carefully examined that the diagnosis is considered.[8]

3. It sometimes presents to gastroenterologists because of hepatomegaly, ascites, or abnormal liver function tests.[8]

4. The most important diagnostic tool is the clinical suspicion of constrictive pericarditis in a patient with signs and symptoms of right sided heart failure that are disproportionate to pulmonary or left sided heart disease.[24]

5. In patients presenting with congestive heart failure in the absence of cardiac dilatation or valvular disease, the diagnosis of constriction or restrictive cardiomyopathies should be suspected.[25]

6. Understanding the pathophysiology of this disease and using non-invasive and invasive techniques are helpful in diagnosis of chronic constrictive pericardtits.[24]

Appendix-1:

Differentiation of pericardial restriction from restrictive cardiomyopathy.

	Pericardial constriction	Restrictive Cardiomyopathy
Physical examination		
Pulmonary congestion	Usually present	Usually absent.
JVP	Prominent Y descent	
Early Diastolic sound	Pericardial knock	S3 low pitched
Pericardial thickness	Usually > 2 mm	< 2 mm
Echo/ Doppler		
LV Myocardium	Normal	"Sparkling"myocardium in amyloid
Atrial Size	+/- Atrial enlargement	Atrial enlargement
Mitral valve flow pattern	Restricted	Restricted
Respiratory variation in E wave	>25%	<20%
Mitral annular diastolic velocity	>8 cm/s	<8 cm/s
Biomarker		
B- type natriuretic peptide	<200 pg/ml	>600 pg/ml
Hemodynamics		
Y descent	Prominent	Variable
Reciprocal respiratory variation in RV/LV peak systolic pressure	Present	Absent

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