An Uncommon Cardiac Etiology of Liver Cirrhosis, Recurrent Ascites, Atrial Fibrillation and Congestive Heart Failure

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Abstract

Objectives: To present a case of a long standing, recurrent ascites, liver cirrhosis under Gastro-enterology follow up, and AF rhythm who was recently diagnosed with congestive heart failure due to constrictive pericarditis with a typical hemodynamic pattern, and markedly improved after pericardiectomy.

Case Presentation and Intervention: A middle aged lady with a long history of recurrent ascites, liver cirrhosis and heart failure symptoms. Recently was reassessed, found to have atrial fibrillation rhythm with fast heart rate, markedly dilated atria, relatively small ventricles size and a restrictive filling pattern by echocardiography. Chest CT scan showed a thickened and heavily calcified pericardium. Cardiac catheterization showed a heavy calcification surrounding the heart, normal coronaries, and a typical hemodynamic pattern of constrictive pericarditis. Surgical pericardectomy was done and a sample of the resected pericardium was sent for histopathology which described a chronic inflammatory process with dense fibrosis and calcification. After surgery patient had remarkable improvement with no more ascites, less lower limbs edema, and reduced diuretics dose she needed.

Conclusion: Constrictive pericarditis is a reported cause of refractory left and right heart failure, recurrent ascites and cardiac liver cirrhosis, all of which can improve markedly after surgical pericardiectomy.

Keywords: Cardiac Etiology; Liver Cirrhosis; Recurrent Ascites; Atrial Fibrillation; Congestive Heart Failure

Introduction

The pericardium is a fibrous sac that surrounds the heart. It consists of 2 layers: the visceral and parietal pericardium. The visceral pericardium is composed of a single layer of cells adherent to the epicardium, while the parietal layer is a < 2 mm fibrous membrane. The 2 layers are separated by a potential space, normally contain < 40 mL of serous fluid [1,2].

The pericardium enhances mechanical interactions of cardiac chambers and limits acute cardiac dilatation. In response to long-standing stress, the pericardium dilates, shifting the pericardial pressure-volume relation substantially to the right [3]. This allows a slowly accumulating pericardial effusion to become quite large without compressing the cardiac chambers and for left ventricular remodeling to occur without pericardial constriction [4,5].

Constrictive Pericarditis is a rare underlying pathology of congestive heart failure which is mainly caused by a remarkable thickening, stiffness, and usually calcification of pericardium, with main pathophysiology is equalization of the end-diastolic pressures in all cardiac chambers [6-8]. The typical “Dip and Plateu” or “square root” pattern of ventricular hemodynamic tracing is one of the important clues for diagnosis, however, it can be reported less frequently in other pathologies.

Diagnostic challenges are usually present, these include clinical and hemodynamic resemblance of restrictive cardiomyopathy. Here I present a case with long standing history of liver cirrhosis and recurrent Ascites, that was recently diagnosed with constrictive pericarditis, having the typical hemodynamic pattern, and improved after pericardectomy.

**Case Presentation and Intervention**

A 39 year old lady, not known to have any chronic illnesses before, had a 3 years history of recurrent admission with ascites, lower limbs edema, dyspnea, and signs of liver cirrhosis, with which she was diagnosed by clinical features and imaging (Figure 1) and was under Gastroenterology follow up during that period, however, all investigations failed to prove a specific primary hepatic pathology. A previous echocardiography raised a possibility of restrictive cardiomyopathy but with no confirmation. When patient presented to our institution, she had a significant ascites, congestive heart failure status, lower limbs edema and distended neck veins, her ECG showed atrial fibrillation rhythm with relatively fast heart rate, while her echocardiography revealed markedly dilated atria, relatively small ventricles size (Figure 2) with a restrictive filling pattern, mildly impaired LV systolic function, and mild to moderate mitral and tricuspid regurgitation. A revision of her old chest X rays revealed some scattered calcification around the heart shadow which was not noticed before (Figure 3), a chest CT scan was then arranged and showed a thickened and heavily calcified pericardium (Figure 4), which raised the possibility of constrictive pericarditis as the cause of her condition. Patient denied any history of pulmonary TB symptoms, chest radiation, or previous surgery.

**Figure 1**

![Figure 1](image1)

**Figure 2**

![Figure 2](image2)

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Cardiac catheterization (both left and right) was done to assess the hemodynamic tracing pattern as well as the coronary vessels prior to any surgical intervention. During cardiac catheterization, and before contrast injection, there was a clear evidence of heavy calcification surrounding the heart (Figure 5), with normal coronaries. The typical „dip and plateau“ or „square root“ pattern was shown in both ventricles hemodynamic tracing (Figure 6-8).
Figure 6

Figure 7

Figure 8

Patient was then referred to cardiac surgeon for surgical pericardectomy, which was done within 2 weeks after explaining the risks and benefits to the patient. During surgery around 80% of the thickened and heavily calcified pericardium was resected (Figure 9 and 10) and a sample was sent for histopathology which, in turn, described a chronic inflammatory process with dense fibrosis and calcification and no signs of granuloma (neither caseating or non-caseating) or any other specific pathology (Figure 11 and 12).

**Figure 9**

**Figure 10**

**Figure 11**
4 weeks later, patient was clinically reassessed, she had remarkable improvement, no more ascites, less lower limbs edema, and reduced diuretics dose. According to her, she started to have a better quality of life than she used to have over the past 3 years.

Discussion

Here we can raise some questions: Was it possible to diagnose this lady earlier, so her suffering could have been shortened?. Did she have another pathology beside constrictive pericarditis which could have attributed to her symptoms?. Any further investigations or work up were needed to role in/out any specific etiology for her constrictive pericarditis?

Constrictive pericarditis is an uncommon cause of congestive (mainly right) heart failure, however, it should be included in the differential diagnosis list. Restrictive cardiomyopathy is the main differential diagnosis due to similar echocardiographic, clinical, and some hemodynamic features. Differentiation between both is important since one of them is surgically manageable by removing the constricting pericardium, while the other is a non-curable disease. Pericardial constriction usually results from long-standing pericardial inflammatory process that leads to pericardial scarring, fibrosis, and calcification [8]. The most frequent causes are chronic idiopathic pericarditis, mediastinal radiation, post cardiac surgery, and tuberculous pericarditis [9-11]. Putting this pathology in mind, though rare, will make early diagnosis and proper management possible.

Differentiation between Constriction and restriction can be achieved through careful history taking (Previous surgery, chest radiation, recurrent viral pericarditis or Tuberculosis in constriction, vs conditions that may lead to amyloidosis, hemochromatosis, or other infiltrative disorders in restriction), in addition to the imaging methods that will show the heavy calcification of the pericardium. Simply, clinician needs to ask for proper chest X-ray and Chest CT scan with careful interpretation in patients who have refractory congestive (especially right side) heart failure, AF rhythm and restrictive cardiomyopathy features [12,13]. This can be enough to strongly suspect the diagnosis.

Traditionally confirmation used to be through invasive hemodynamic assessment of both ventricles showing the typical, yet not so specific, „dip and plateau“ or „square root“ appearance of the hemodynamic tracing, which was recorded in our case, in addition to equalization of all four cardiac chambers in end-diastole. Furthermore, cardiac cath will assess the coronary vessels prior to pericardectomy to exclude any significant coronary artery disease [14]. Later confirmation can be achieved by histopathologic assessment after surgery, like in our case.

Other comorbidities can associate the clinical scenario, however in our case, beside the negative investigations that did not prove any other hepatic or cardiovascular pathology that was not explained by her diagnosis, her marked clinical improvement after pericardectomy gives us a very good reason to think of constrictive pericarditis as the main single pathology. Further investigations to role out specific
pathology such as tuberculous pericarditis were reasonable in spite of absence of relevant history. However, histopathologic assessment of the resected pericardium failed to prove any specific pathology like granulomatous process, rather, a chronic inflammation, fibrosis, and dense calcification were noticed.

**Conclusion and Implication of Clinical Practice**

Constrictive pericarditis is a reported cause of refractory and recurrent ascites, liver cirrhosis, Congestive heart failure and atrial fibrillation rhythm. Patients can improve markedly after surgical pericardectomy. Putting this pathology in mind will make it possible to suspect and then confirm the diagnosis of this rare „curable“ disease earlier.

**Bibliography**


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