Constrictive pericarditis – A challenging diagnosis

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ABSTRACT

Constrictive pericarditis is a challenging diagnosis. This case report describes a patient with constrictive pericarditis (CP) secondary to radiation therapy. As the patient presented with non-specific symptoms, various modalities (both invasive and non-invasive) were used in the diagnosis and differentiation of CP from restrictive cardiomyopathy (RCM). This case of surgically-proven CP demonstrates that a high clinical suspicion based on physical findings should be combined with multiple diagnostic modalities in the diagnosis of CP. It is crucial to differentiate CP from RCM due to the significant variance in treatment.

Key Words: Constrictive pericarditis, Echocardiography, Radiation, Restrictive cardiomyopathy, Diagnosis

1. INTRODUCTION

The pericardium is a fibrous sac surrounding the heart that consists of two layers: the visceral and parietal pericardium. With some variability, the thickness of the normal pericardium as measured on computed tomography (CT) and magnetic resonance imaging (MRI) is less than 2 mm.[1] Patients with constrictive pericarditis (CP) present with clinical features of congestive heart failure with right-sided features being more prominent than left-sided heart failure symptoms. More commonly, patients present with non-specific symptoms and different types of diagnostic modalities are needed to confirm the diagnosis. It is important to differentiate CP from restrictive cardiomyopathy (RCM) as both can present with similar symptoms but have dramatically different treatment modalities. The diagnosis and differentiation of these diseases can be quite challenging and we describe a case in which we were compelled to use multiple diagnostic tests to clinch the diagnosis.

2. CASE PRESENTATION

The patient is a 54-year-old man with a history of hypertension, coronary artery disease and inferior wall myocardial infarction revascularized with a drug-eluting stent in the right coronary artery nine months prior to presentation, and myasthenia gravis treated with thymectomy and radiation. The patient was hospitalized multiple times for shortness of breath and underwent numerous thoracenteses, but the etiology of recurrent pleural effusions had not been identified. Pertinent findings on physical exam included elevated jugular venous pulse, pericardial rub without evidence of murmur, ascites, and bilateral lower extremity edema. Chest X-ray showed bilateral pleural effusions. Kussmaul’s sign was negative.

Transthoracic echocardiography (TTE) demonstrated a septal bounce during inspiration, dilated and non-compressible inferior vena cava (IVC), and a thickened posterior pericardium (8-10 mm). Notably, TTE showed normal left ventricle (LV) inflow velocities and no annulus paradoxus. CT of the chest showed significant thickening and calcification of the pericardium and mediastinum secondary to radiation as well as a dilated superior vena cava and IVC (see Figure 1).

Cardiac MRI demonstrated calcification adjacent to the left
atrioventricular groove and the anterior aspect of the LV and right ventricle (RV), septal bounce, and exaggerated inspiratory septal flattening suggestive of constrictive physiology. LV ejection fraction was preserved at 55% and there was no pericardial effusion. Heart catheterization was performed showing discordance with an inspiratory rise in RV pressure and concomitant drop in LV pressure (see Figure 2). Unfortunately, most of the discordant waves were associated with premature ventricular contractions preventing definitive assessment of discordance in the catheterization lab. Upon arriving at the diagnosis of CP, the patient was evaluated by thoracic surgery and underwent anterior pericardiectomy with improvement in symptoms and resolution of pleural effusions.

Figure 1. CT scan showing thickened and calcified pericardium

Figure 2. Right heart catheterization showing discordance in the pressure waves with premature ventricular contractions

3. DISCUSSION

CP is the result of a number of disease processes causing pericardial inflammation. Radiation exposure accounts for nearly 31% of cases of CP.\(^2\) Pathologically, there is thickening and fibrosis of the visceral and parietal pericardium often with adhesions to adjacent myocardium. About 50% of these patients have some degree of pericardial calcification.\(^2\)
With CP, the stiffened pericardium prevents the ventricles from relaxing and forces ventricular filling to be dependent on each other (ventricular interdependence) leading to reduced filling. During inspiration, the interventricular septum shifts to the left because of decreased LV filling, during expiration, the increased filling of the LV forces the septum to return back to normal. This abnormal septal variation during respiration, which is most prominent in early diastole, is called a “septal bounce” and is a characteristic feature in CP that can be seen on TTE and cardiac MRI. Mitral valve (MV) and tricuspid valve (TV) peak flow velocities are also affected during respiration with CP. This respiratory variation is not seen in RCM thus helping in differentiating the two disease processes.

CT scan is another imaging modality used to differentiate between CP and RCM. CT provides a reliable assessment of pericardial calcification and thickness. The primary disadvantage of a CT scan is that it cannot be used to evaluate the ventricular function and hemodynamic parameters. For this reason, physicians often obtain both a CT scan and a TTE when evaluating for CP. Cardiac MRI is another imaging modality often utilized and the finding of interventricular dependence can differentiate CP from RCM.

It is not uncommon for the pericardium of patients with CP to appear normal on imaging studies and physical examination. This is when heart catheterization may be necessary to make the diagnosis. In this case, the TTE, CT scan, and heart catheterization showed a few features of CP but failed to provide a definitive diagnosis. Cardiac MRI sealed the diagnosis which was later proven during surgery.

Pericardiectomy is the only definitive treatment option for patients with chronic symptomatic CP; patients with only mild symptoms and those with end-stage disease are not suitable candidates. Although curative, there is documented evidence of significant mortality with this procedure and so the diagnosis needs to be confirmed before proceeding with treatment.

Radiation can cause both CP and RCM. The patient’s case is unique as it highlights the multiple different modalities that were utilized to confirm the diagnosis and thus guiding us in providing the appropriate form of treatment.

**CONFLICTS OF INTEREST DISCLOSURE**

The authors have declared no conflicts of interest.

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**REFERENCES**


