

Mixed constrictive pericarditis and restrictive cardiomyopathy in a 36-year-old female

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Abstract

Mixed lesion of Restrictive Cardiomyopathy and Constrictive Pericarditis is a rarely reported clinical entity which poses a diagnostic and therapeutic enigma to physicians. The management of both conditions differs markedly. Restrictive Cardiomyopathy is managed either conservatively or cardiac transplant may be offered. On the other hand, Constrictive Pericarditis can be surgically treated by pericardiectomy. We report a rare case of decompensated heart failure presenting with mixed features of both constrictive and restrictive cardiomyopathy.

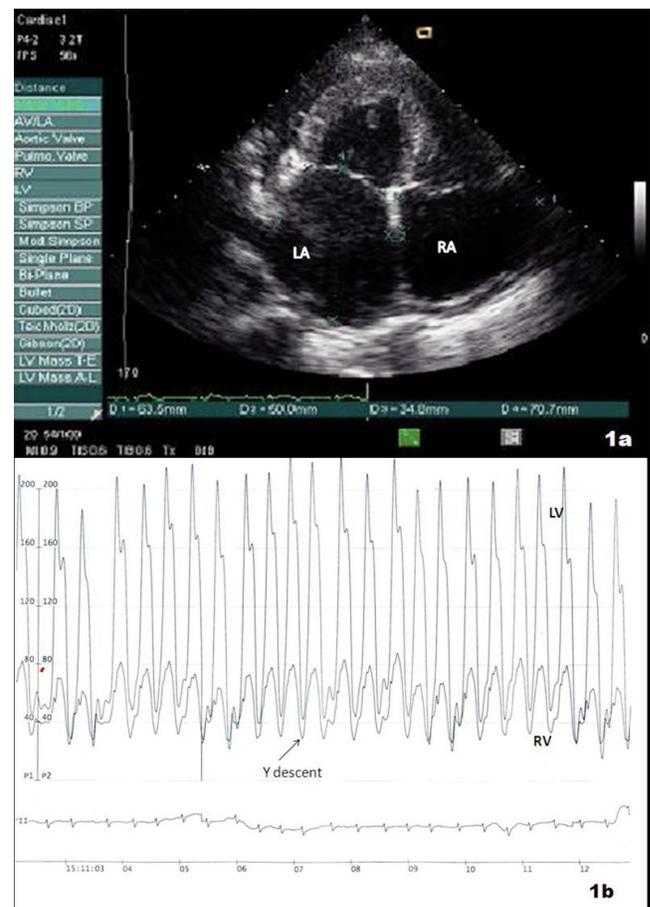
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Introduction

Constrictive Pericarditis (CP) is characterised by scarring and loss of elasticity of the pericardium, resulting in external impedance of normal diastolic cardiac filling.¹ Restrictive Cardiomyopathy (RC) is defined as a heart muscle disease that results in impaired ventricular filling, with normal or decreased diastolic volume of either or both ventricles.² Differentiating between Constrictive Pericarditis and Restrictive Cardiomyopathy is a difficult clinical challenge that requires multiple diagnostic modalities, using haemodynamics, imaging and biopsy studies. Appropriate selection of treatment is of critical value, as both conditions are managed differently. We report a rare case of a patient from adult age group who had mixed features of both Constrictive Pericarditis and Restrictive Cardiomyopathy.

Case Report

A 35-year-old female with no past significant medical history presented with signs of predominant right-sided heart failure. She had a two-month history of gradually increasing shortness of breath and orthopnoea. She had irregular pulse, pedal oedema and raised JVP. EKG revealed atrial fibrillation, while chest X-ray showed an increased cardiac shadow with increased bronchovascular markings in both lung fields with minimal right basal



LV: left ventricle
RV: right ventricle
RVSP: Right ventricle systolic pressure.

Figure-1: a) Echo image showing massive biatrial enlargement. b) Simultaneous LV and RV pressure recordings showing RVSP of 80 mmHg and prominent Y descent.

pleural effusion. Echocardiography revealed dilated Left Ventricle (LV) with mild systolic dysfunction, EF-45% and preserved myocardial thickness and severe bi-atrial dilatation of left atrium (LA) 52mm and right atrium (RA) 54mm (Figure-1a), mild mitral regurgitation (MR), severe

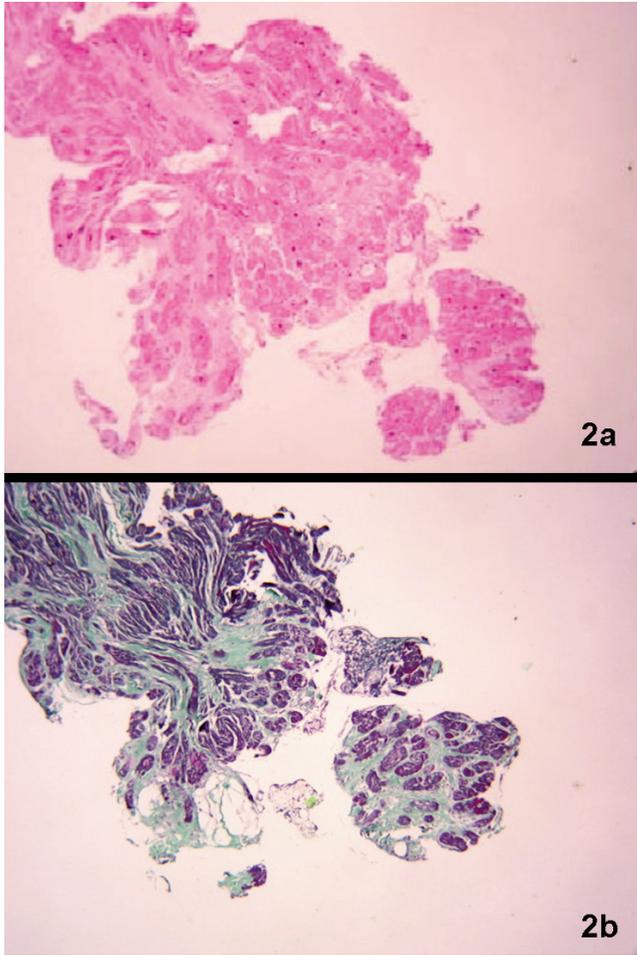


Figure-2: a) Myocardial biopsy showing focal myocardial necrosis, degenerative changes and moderate intramyocardial fibrosis. b) Special stains for collagen are positive.

tricuspid regurgitation (TR) with estimated pulmonary artery pressure of around 60mmHg. There were respiratory variations noted at mitral and tricuspid inflow with pulse wave Doppler suggestive of interventricular interdependence. Tissue Doppler Imaging (TDI) revealed a peak Ea velocity of 16cm/s. These features were suggestive of Constrictive Pericarditis, but, interestingly, pericardium was not thickened on 2D-echo. CT scan of the chest was performed to evaluate pericardial thickening, which was less than 2mm and did not meet the CP criteria. In view of the contradictory findings, we performed a right and left heart catheterization. It revealed LVEDP - RVEDP to be 10mmHg, while the right ventricular systolic pressure was 80mmHg. These findings favoured restrictive cardiomyopathy. However, interestingly again, her right ventricular pressure tracing showed a prominent Y descent and RVEDP/RVSP was 0.425 which indicated Constrictive Pericarditis (Figure-1b). Hence, she had mixed features of both the conditions. Keeping in view the therapeutic

implications of exact diagnosis, myocardial biopsy was performed, through a sub-xiphoid approach which revealed focal myocyte necrosis, degenerative changes and moderate intramyocardial fibrosis. Special stains were positive for collagen (Figure-2a & b). The pericardium showed sections of thickening and fibrosis. Based on the suspicion of RC, certain special investigations were done to rule out sarcoidosis, connective tissue disease, haemochromatosis and amyloidosis. These results were normal. After exhaustive investigations, it was concluded that the patient had mixed features of CP and RC. In view of extensive myocardial fibrosis with minimal pericardial thickening on the CT scan, and end-stage cardiomyopathy, we decided not to perform a pericardiectomy.

Discussion

Restrictive Cardiomyopathy with overlapping Constrictive Pericarditis is a rarely accounted clinical condition and only isolated case reports have been cited.³ A number of studies using different techniques have been attempted to distinguish between the two conditions.¹ However, no technique is totally reliable and some patients may have mixed features of both conditions. In the case of our patient, multiple invasive and non-invasive diagnostic modalities were used, including two-dimensional echocardiography, tissue Doppler echocardiography, cardiac catheterisation, computed tomography (CT) chest scan and cardiac biopsy. There were elements of both restrictive and constrictive disease in these investigations which made treatment options difficult to choose from.

Simultaneous right and left heart pressure tracings revealed interesting haemodynamics. Vaikus described three haemodynamic criteria for differentiating restrictive from constrictive pathology.³ (i) equalization of right ventricular end-diastolic pressure (RVEDP) and left ventricular end-diastolic pressure (LVEDP), with constriction more likely with a difference of less than 5 mmHg, (ii) elevation of right ventricular systolic pressures (RVSP) to greater than 50 mmHg with restriction, and less than 50 mmHg with constriction; and (iii) RVEDP ratio to RVSP exceeding one-third in constriction and less than one-third with restriction. The overall predictive value of these criteria were 85%, 70% and 76%, respectively. Our patient had two features that suggested RC (LVEDP-RVEDP = 10mmHg and RVSP 80mmHg), and one feature that suggested CP (RVEDP/RVSP = 0.425) She also had a prominent Y descent on right heart pressure tracings that favoured CP.

There are several studies in adults which describe the use of tissue Doppler flow velocities for differentiating constrictive and restrictive pathology.⁴ Early diastolic mitral annular velocity (peak Ea) has been used to differentiate RC

from CP. It has been proposed that a cutoff value of Ea >8cm/sec favours constrictive, while <8cm/sec favours restriction. The peak Ea in our patient was more than 8.0cm/s. Hence, it favoured a CP diagnosis. Similarly, respiratory variations were noted at mitral and tricuspid inflow suggestive of interventricular interdependence also favouring CP. Interestingly, the pericardium was not thickened on 2D-echo which was further confirmed by CT chest. This made the exact diagnosis a dilemma. Therefore, we decided to perform cardiac biopsy which was suggestive of intramyocardial fibrosis with pericardial involvement. On the basis of an extensive intramyocardial fibrosis and positive collagen staining, RC was suspected. We carried out a series of specific investigations, which were negative for a probable RC cause. It was then contemplated that she could be suffering from idiopathic restrictive cardiomyopathy (IRCM) in which haemodynamic abnormalities occur in the absence of specific etiology. The disease is characterised by symptoms of progressive left and right-sided heart failure. The overall prognosis is poor, especially when the onset is in childhood, and patients often require cardiac transplantation.⁵ However, a remarkable paradox is that in IRCM, the pericardium is normal.² The patient's biopsy had extensive pericardial fibrosis. Thus, on basis of all the factors, it was concluded that she was suffering from mixed features of both RC and CP.

Mixed restrictive and constrictive pathology has been reported in a 17-year-old female, who had calcified pericardium and tissue Doppler imaging revealed evidence of intact active relaxation.³ Based on these findings, it was concluded that constriction rather than restriction was the dominant factor contributing to her pathology. In view of this, pericardiectomy was performed and the patient had recovered. In contrast, our patient had an advanced form of cardiomyopathy with severe myocardial fibrosis and pericardial involvement. It was decided not to operate upon the patient and to manage her conservatively.

Significant overlap can exist between pericardial and myocardial diseases.³ In the current era, exposure to radiation is attributed as the most important condition which

may involve both pericardium and myocardium.^{6,7} Our patient did not have any history of such exposure. Tuberculous constrictive pericarditis has been reported with myocardial and endocardial involvement in a case series.⁸ It has been suggested that tubercular insult to the heart may result in involvement of all three layers with varying severity and the hallmark of differentiation between constrictive and restrictive disease appears to be a diligent attempt at identification of a thickened pericardium. Our patients' clinical course and investigation did not suggest tuberculosis and her biopsy was also negative for any granuloma. Keeping in view all this, we postulate that our case could be a new variant or entity of pancarditis of unknown etiology leading to both constrictive and restrictive picture in this part of world.

Conclusion

To our knowledge, it is the first reported case of mixed features of both Constrictive Pericarditis and Restrictive Cardiomyopathy of unknown etiology presenting in this age group in the region.

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