Singapore Med J 2002 Vol 43(6) : 308-311

Constrictive Pericarditis – A Rare but Important Cause of Recurrent Cardiac Failure: A Case Report

V Y T Lim, R M L Kam, K K W Chen, F K Cheah, Z P Ding

ABSTRACT

Constrictive pericarditis (CP) is an uncommon cardiac disease which is often difficult to diagnose because of its vague and myriad clinical presentations. We report a case of a middle-aged lady who had non-specific symptoms and signs for six years before she was eventually diagnosed to have idiopathic constrictive pericarditis. An awareness and understanding of this condition is important, as it is a progressive condition and the likelihood of cure depends very much on its early identification and treatment.

Keywords: congestive cardiac failure, constrictive pericarditis, echocardiogram, magnetic resonance imaging, pericardiectomy

Singapore Med J 2002 Vol 43(6):308-311

Department of Cardiology
National Heart Centre
Mistri Wing
Third Hospital Avenue
Singapore 168752

V Y T Lim, MB ChB, MRCP (UK)
Registrar

R M L Kam, MMed (Int Med), MRCP (UK), FAMS
Senior Consultant

Z P Ding, MBBS, MMed (Int Med), FAMS
Senior Consultant

Department of Diagnostic Radiology
Singapore General Hospital
Outram Road
Singapore 169608

K K W Chen, MB ChB, MRCP (UK), FRCR
Registrar

F K Cheah, MB ChB, MRCP (UK), FRCR
Senior Consultant

Correspondence to:
Dr R M L Kam
Tel: (65) 6436 7542
Fax: (65) 6227 3562
Email: Ruth_KAM@nhc.com.sg

INTRODUCTION

There are few truly curable cardiac diseases. One of these is constrictive pericarditis (CP), a rare cardiac condition that usually manifests itself with non-specific symptoms and signs spread out over many months or even years. A better appreciation of this disease is important if one is to suspect its presence in a clinical scenario, which is an essential step that may lead to its diagnosis.

CASE PRESENTATION

This lady first presented six years ago at the age of forty, with breathlessness, easy fatiguability and atrial fibrillation (AF). The initial echocardiogram reported moderate mitral regurgitation and normal ventricular function. She had excellent effort tolerance, was treated with digoxin and warfarin, and declined further investigation.

She presented six years later with two episodes of congestive cardiac failure within a year. ECG showed AF with non-specific diffuse T wave inversions (Fig. 1). Chest X-ray did not show any sign of previous pulmonary tuberculosis, although the left cardiac border had an unusual double profile (Fig. 2). Echocardiogram was repeated and

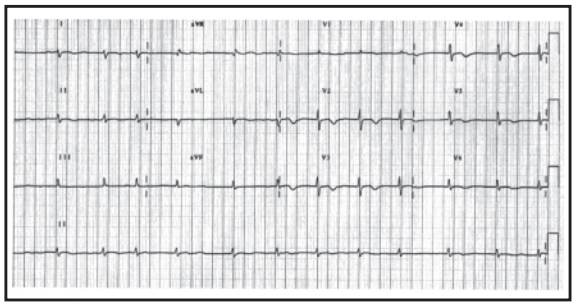

Fig. 1 ECG of patient showing non-specific features of atrial fibrillation, generalised low QRS voltages and diffuse small T wave inversions.

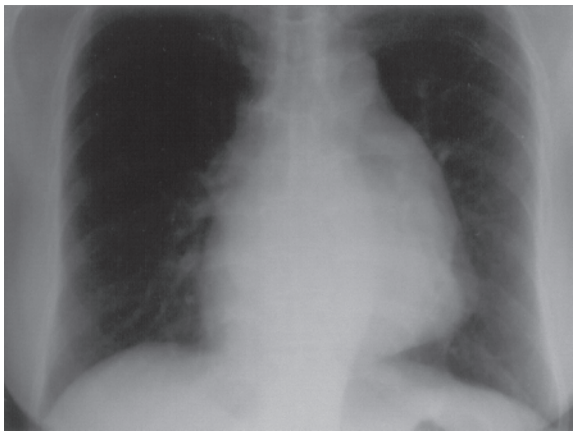

Fig. 2 Chest radiograph showing an unusual double profile of the left heart border.

this demonstrated normal left ventricular function. There was mild to moderate mitral regurgitation through a morphologically normal mitral valve and dilated mitral annulus. Biatrial dilatation was seen.

As the history of recurrent congestive cardiac failure was incongruent with her echocardiogram findings, further investigations were arranged. Coronary angiography showed normal coronary arteries. However, a small area of linear calcification was seen near the apex during fluoroscopy. Simultaneous left and right heart studies performed after infusion of 500 mls saline demonstrated elevation and equalisation of the diastolic pressures of all cardiac chambers to within 5 mmHg. The mean right atrial pressure (RAP) was 16 mmHg with a prominent Y descent, right ventricular end diastolic pressure (RVEDP) 16 mmHg, left ventricular end

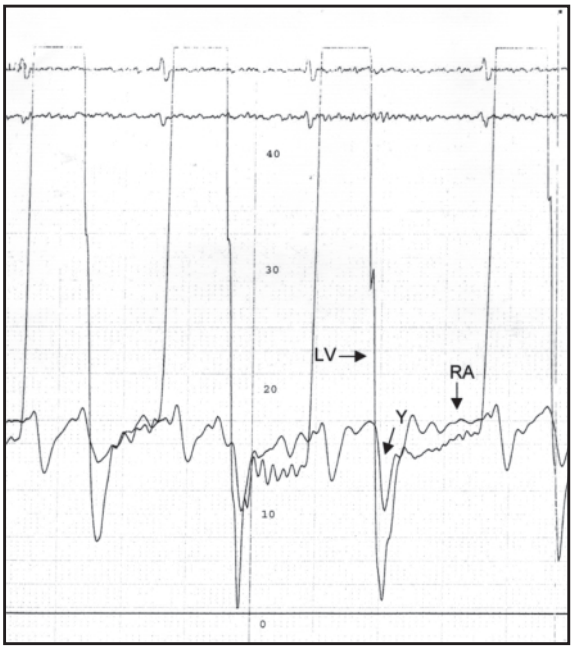

Fig. 3 Simultaneous left ventricular (LV) and right atrial (RA) pressure recordings obtained at cardiac catheterisation demonstrating equalisation of diastolic pressures and steep Y descent (Y) in RA tracing. (Pressures are in mmHg)

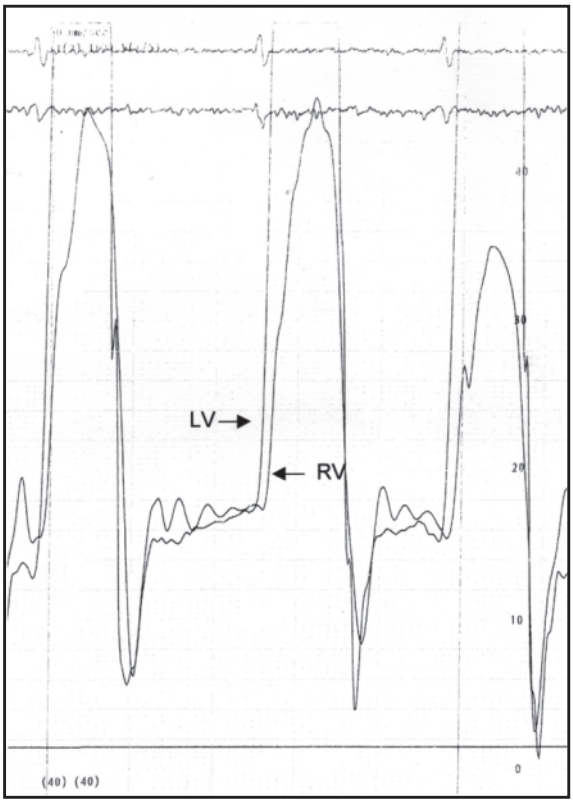

Fig. 4 Simultaneous left ventricular (LV) and right ventricular (RV) pressure recordings demonstrating equalisation of diastolic pressures and characteristic "dip and plateau" contour. (Pressures are in mmHg)

diastolic pressure (LVEDP) 15 mmHg and pulmonary-capillary wedge pressure (PCWP) 18 mmHg. The ventricular tracing showed a dip-and-plateau pattern ("square root sign") (Figs. 3 and 4).

The echocardiogram was repeated specifically to look for features of constriction. This time subtle signs of diastolic checking of the posterior left ventricular wall and abnormal septal bounce were noted. Investigations looking for a possible cause of CP, for example, the Mantoux test, rheumatoid factor and antinuclear antibodies, were either negative or inconclusive. Cardiac MRI (Fig. 5) showed that the left ventricle was distorted close to its apex by an indrawing of the free wall and pericardium, associated with localised calcification. This feature was responsible for the double profile seen on the left cardiac border on chest X-ray. CT thorax clearly demonstrated the band of calcification (Fig. 6).

The diagnosis of constrictive pericarditis was confirmed on the evidence of pericardial calcification and thickening as seen on MRI, in combination with evidence of a constrictive physiology from cardiac catheterisation and echocardiogram. Subtotal pericardiectomy was performed in October 2000. During the surgery, it was confirmed that there was localised thickening and calcification of the pericardium over the apical and anterolateral wall of the left ventricle. The rest of the pericardium was not thickened. Multiple adhesions were present in the left pleural cavity. Histology of the excised pericardium only showed fibrosis, mild chronic inflammation and calcification.

The surgery was a complete success and the patient has remained well at nine months' follow-up. Currently she is only on digoxin and warfarin for the atrial fibrillation.

DISCUSSION

The underlying pathology of CP is variable thickening, fibrosis and frequently calcification of the pericardium, which results in a constrictive physiology where the normal diastolic filling of the heart is impaired[1]. The condition usually affects the whole pericardium, though rarely it may be localised, as in this case, where the appearance of the band of calcification is analogous to having a rubber band tied tightly to the left ventricle, causing a marked indentation of the myocardium and constriction. There is predominantly backward heart failure as a result of elevated filling pressures.

In the past, the commonest aetiology was tuberculosis, and this remains true in developing countries, but not for developed nations. In a recent study from United States, idiopathic CP is most common (34%) and the three most common identifiable causes were cardiac surgery (18%), pericarditis (16%) and mediastinal irradiation (13%). Other possible causes include connective tissue diseases, malignancy, trauma and infections[2].

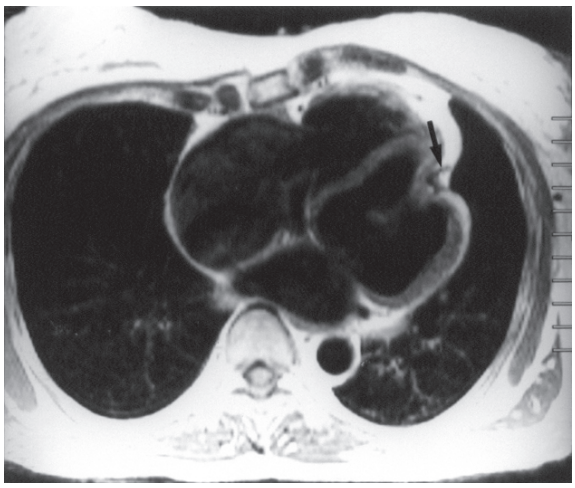

Fig. 5 Axial T1-weighted breath-hold MRI of thorax showing localised pericardial thickening (arrow) at the site where the left ventricle is inwardly indented. Enlargement of both atria, particularly of the right, are present.

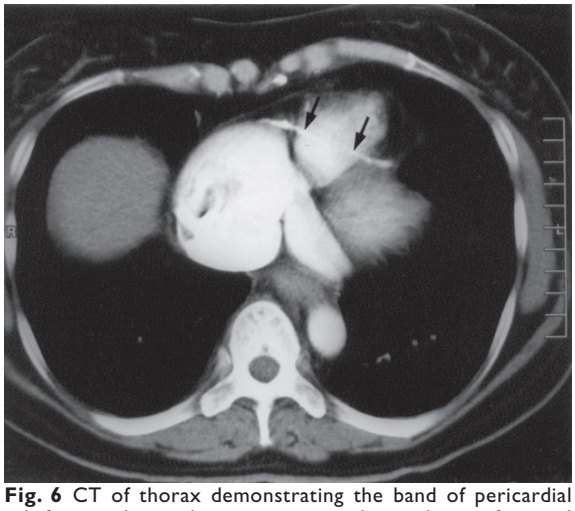

Fig. 6 CT of thorax demonstrating the band of pericardial calcification (arrows) running across the cardiac surface and causing inward indentation of the left ventricular free wall.

Presentations are often insidious and non-specific, such as reduced effort tolerance, orthopnoea or fatigue. In a recent European study, the average duration of symptoms before definitive diagnosis was 20 months[3]. The most important clinical finding is that of a raised jugular venous pressure with elevation on inspiration (Kussmaul's sign), though this in itself is not specific for CP, as it can occur in any condition with elevated right sided pressures. There may be a diastolic pericardial knock due to the sudden cessation of ventricular filling, which may be heard as a "third heart sound". Other clinical findings such as pulmonary congestion, hepatomegaly, ascites and peripheral oedema are a reflection of the elevated diastolic pressures causing heart failure.

The diagnosis of CP can be elusive and initial clinical suspicion often helps. Unfortunately, there is no single diagnostic test. Electrocardiograph is almost always abnormal and often shows non-specific low QRS voltages with generalised T wave inversion or flattening. Atrial fibrillation occurs in less than 50% of patients. Chest X-ray occasionally shows some pericardial calcification and though it may suggest CP, it is not diagnostic in itself. The most helpful investigations are the echocardiogram, computed tomography (CT) or magnetic resonance imaging (MRI), and cardiac catheterisation, and diagnosis of CP may require a combination of all these modalities.

On echocardiography, actual pericardial thickening is often not seen, but the inferior vena cava is often dilated. On M-mode echocardiography, diastolic "septal bounce" may be seen, as well as abrupt checking of the left ventricular posterior wall diastolic movement by the rigid pericardium (coinciding with the "pericardial knock")[4]. Another suggestive sign is early opening of the pulmonary valve in late diastole, due to an elevated RVEDP. There may also be respiration-related changes in valvular inflow velocities, such as an inspiratory decrease of more than 25% in the transmitral flow velocity. Reduction of preload by manoeuvres such as head-up tilt or diuresis may help to augment or unmask the typical respiratory variation[5].

Both CT and MRI of the heart can identify pericardial thickening much better than echocardiography, and are probably the most sensitive imaging techniques currently available for delineating the morphology and thickness of the pericardium[6]. The use of cine sequences during MRI also allow the depiction of morphological as well as kinetic dysfunction that result from the constriction. The ventricles may also be deformed as a result of the constriction. This is especially true in focal constriction where resultant movement abnormalities are well visualised. Severe constrictive physiology, however, could occur in the presence of a diseased but minimally thickened pericardium, and obvious pericardial thickening with calcification may not be associated with haemodynamically significant constriction.

Simultaneous recording of left and right heart pressures is helpful in documenting the presence of a constrictive physiology. The characteristic finding is elevation and equilibration of diastolic filling pressure in all cardiac chambers to within 5 mmHg. There is often a "dip and plateau" or "square root sign" for both right and left ventricle diastolic pressure curves, with a low early diastolic pressure and rapid rise to a high plateau, though this sign is not specific in itself. It is important to avoid prior diuresis before this test, as the characteristic findings may be obscured in the presence of hypovolaemia. Sometimes a rapid volume infusion to increase intravascular volume can help to unmask the characteristic findings[7].

One of the main differential diagnoses of CP is restrictive cardiomyopathy (RCM). The differentiation can be difficult, but Vaitkus and Kussmaul have identified three haemodynamic criteria which, if fulfilled, has excellent positive predictive value in diagnosing CP over RCM. A difference in RVEDP and LVEDP of ≤5 mmHg, a right ventricular systolic pressure (RVSP) ≤50 mmHg and a ratio of RVEDP to RVSP ≥1:3 all favour constriction[8]. These criteria were all met for this patient. A more sensitive and specific haemodynamic criterion is to demonstrate increased ventricular interdependence on cardiac catheterisation, as evidenced by respiratory discordance of left and right ventricular systolic pressures[9].

Chronic constrictive pericarditis is a progressive disease. Some patients may survive for many years controlled with diuretic therapy, but the majority will become increasingly symptomatic and disabled. Surgical pericardiectomy, which should be as complete as possible, is the definitive therapy which offers the possibility of a cure, with a low in-hospital mortality rate if performed in patients with good condition and without advanced disease. Predictors of poor outcome include radiation related constriction, New York Heart Association class III or IV at presentation, myocardial atrophy, inflammation or scarring[10,11].

CONCLUSION

Constrictive pericarditis is an uncommon condition that remains a diagnostic challenge. It should always be considered in the differential diagnosis of patients with congestive cardiac failure, especially if the ventricular function is good and there is no significant valvular disease. A correct diagnosis is rewarding, as pericardiectomy can be a curative procedure.

REFERENCES

1. Myers RBH, Spodick D. Constrictive pericarditis: clinical and pathophysiologic characteristics. Am Heart J 1999; 138:219-32.
2. Lieng LH, Oh JK, Schaff HV, Danielson GK, Mahoney DW, Seward JB, et al. Constrictive pericarditis in the modern era. Evolving clinical spectrum and impact on outcome after pericardiectomy. Circulation 1999; 100:1380-6.
3. Nataf P, Cacoub P, Dorent R, Jault F, Bors V, Pavie A, et al. Results of subtotal pericardiectomy for constrictive pericarditis. Eur J Cardiothorac Surg 1993; 7:252-6.
4. Oakley CM. Pericardial diseases. In: Julian DG, Camm AJ, Fox KM, et al, editors. Diseases of the heart, 2nd edition. London: Saunders, 1996:558-77.
5. Oh JK, Tajik AJ, Appleton CP, Hatle LK, Nishimura RA, Seward JB, et al. Preload reduction to unmask the characteristic doppler features of constrictive pericarditis. A new observation. Circulation 1997; 95:796-9.
6. Masui T, Finck S, Higgins CB. Constrictive pericarditis and restrictive cardiomyopathy: evaluation with magnetic resonance imaging. Radiology 1992; 182:369-73
7. Bush CA, Stang JM, Wooley CF, Kilman JW. Occult constrictive pericardial disease. Diagnosis by rapid volume expansion and correction by pericardiectomy. Circulation 1977; 56:924-30.
8. Vaitkus PT, Kussmaul WG. Constrictive pericarditis versus restrictive cardiomyopathy: a reappraisal and update of diagnostic criteria. Am Heart J 1991; 122:1431-41.
9. Hurrell DG, Nishimura RA, Higano ST, Appleton CP, Danielson GK, Holmes DR, et al. Value of dynamic respiratory changes in left and right ventricular pressures for the diagnosis of constrictive pericarditis. Circulation 1996; 93:2007-13.
10. DeValeria PA, Baumgartner WA, Casale AS, Greene PS, Cameron DE, Gardner TJ, et al. Current indications, risks and outcomes after pericardiectomy. Ann Thorac Surg 1990; 52:219-24.
11. Ni Y, von Segresser LK, Turina M. Futility of pericardiectomy for postirradiation constrictive pericarditis? Ann Thorac Surg 1990; 49:445-8.