CASE PRESENTATION

Constrictive Pericarditis - A Diagnostic Challenge

C A Nambiar DM(Card) FACC*
A K Chawla MD**

ABSTRACT

Constrictive pericarditis presents with protean manifestation and may masquerade as pleural effusion or as abdominal discomfort or ascites of unknown aetiology. Three cases who presented with atypical manifestations to Salmaniya Medical Centre are reported to emphasise the difficult diagnostic challenge the disease poses. The importance of eliciting subtle physical signs such as jugular venous pulsations, precordial movements and heart sounds and the common investigations to clinch the diagnosis are discussed. Timely diagnosis and prompt surgical resection of the pericardium result in gratifying results and the condition is potentially curable with good prognosis in the long term.

Constrictive pericarditis is a disease with protean manifestations and is known to masquerade as primary abdominal or pulmonary conditions[1-3]. Over the last eighteen months we came across three cases, each presenting with different symptomatology, thus posing a diagnostic challenge. These three cases form the material for this report.

THE CASES

CASE 1

ARH, a 28 year old Bahraini man presented with a history of dyspnoea, cough with mucopurulent expectoration, loss of appetite and weight of 5 months duration. He had physical findings and radiological evidence of right sided pleural effusion and was admitted under thoracic surgeon for investigations and management. The pleural fluid was exudative with lymphocyte dominance. The tuberculin test was positive and ESR was 15mm 1st hour. Anti Nuclear Factor and rheumatoid factor were negative, cold agglutinins for mycoplasma and antilegionella antibodies were negative, sputum was negative for AFB. Bronchoscopy showed no mass lesion, but there was collapse of right lower lobe due to pleural effusion. The patient was put on antitubercular drugs but the pleural effusion reaccumulated rapidly.

At this time cardiological consultation was sought. On clinical evaluation, his jugular venous pressure was grossly elevated to about 18 cms of water with prominent "Y" descent. The cardiac size was normal with an early

* Consultant Cardiologist

**Medical Doctor

Department of Medicine
Salmaniya Medical Centre
State of Bahrain

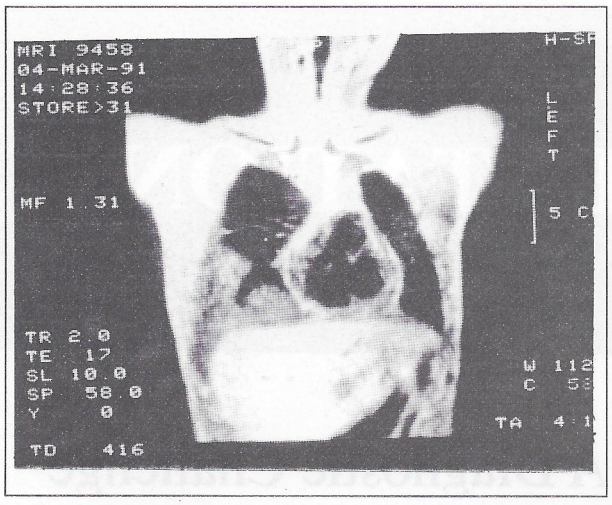

Figure 1: *MRI scan of chest of Case No. 1 showing thickened pericardium and right pleural effusion.*

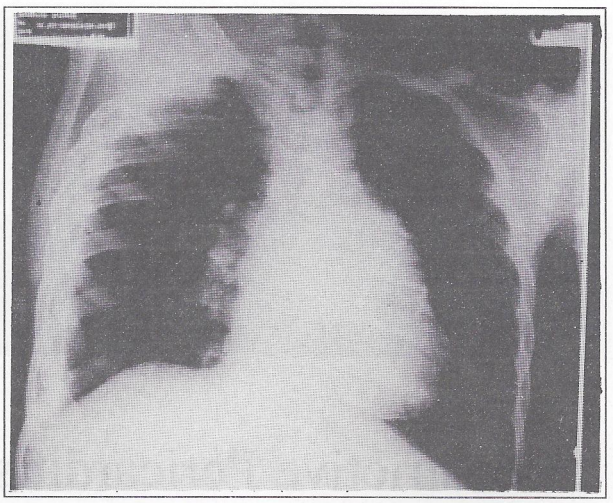

Figure 2: *Rt lateral decubitus X-ray chest of Case No.2 showing a rim of pericardial calcification along the apex of the heart and right pleural effusion.*

S3 but had no murmurs. The liver was enlarged and pulsatile. Electrocardiogram showed low voltage complexes. Radiograph of the chest showed a rim of calcification along the left border of the heart. CT Scan of chest confirmed pericardial calcification. Echocardiography with doppler interrogation confirmed restriction to filling, with normal myocardial and valvular function. The diagnosis of constrictive pericarditis was confirmed and he was advised to undergo surgical treatment. The patient went to India, where MRI scan (Fig 1) and cardiac catheterisation were done before subjecting him to pericardiectomy. Histopathology revealed tuberculosis. He made uneventful recovery and is now completely asymptomatic. Anti-tuberculous treatment was given for a period of one year.

CASE 2

MS, an 18 year old Indian boy was seen by a private physician for swelling of his legs, of about 6 months duration and was referred for cardiac evaluation by him. He denied any history of dyspnoea or chest pain. About 3 months earlier he had consulted a general practitioner for abdominal discomfort. Hepatosplenomegaly and right pleural effusion were documented on abdominal ultrasound examination, but no follow up was done. On physical examination, he had pulsus paradoxus, grossly engorged jugulars with a venous pressure of 15 cms of water and prominent "X" and "Y" descents. There were peculiar rocking movements of the precordium with retraction of the apex. An early S3, consistent with pericardial knock was heard at apex. The liver was grossly enlarged and pulsatile, the spleen was also palpable and there was right pleural effusion. Electrocardiogram showed QRS axis of + 1200, R/S ratio of 1.5 in V1 and normal sinus rhythm. Radiograph of chest (Fig 2) showed a small right pleural effusion and extensive calcification along the left cardiac border around apex.

Echo doppler examination (Fig 3) showed pericardial calcification and restriction to ventricular filling. CT Scan of chest confirmed extensive pericardial calcification (Fig 4) and right pleural effusion. He was advised to undergo pericardectomy which was done in India. Histopathology showed tuberculosis. He was also put on antituberculous treatment and after one year's follow up he is completely asymptomatic.

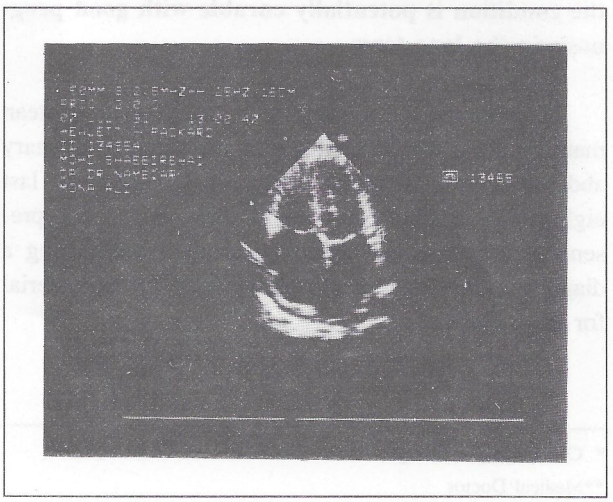

Figure 3: *2D Echocardiography of Case No. 2, shows thickened and Calcified pericardium.*

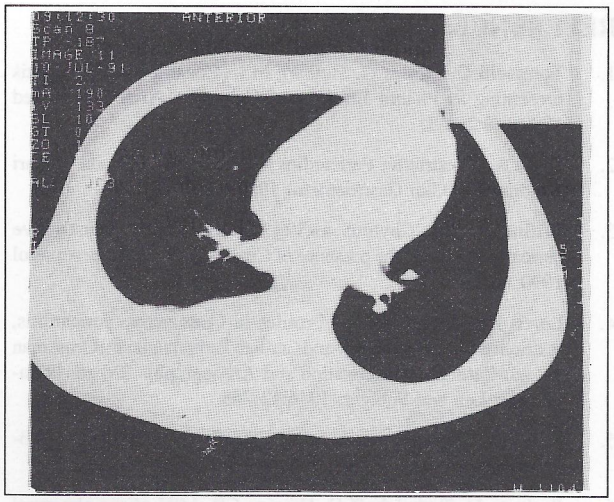

Figure 4: *CT Scan of chest of Case No. 2, showing Calcification of both Visceral & parietal pericardium along the apex.*

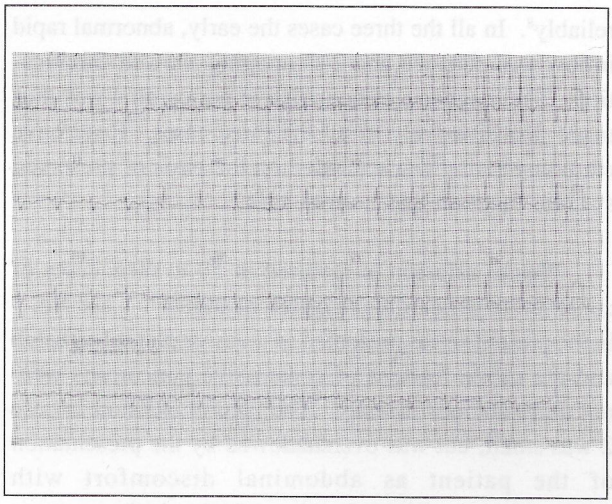

Figure 5: *ECG of case No.3, showing left axis deviation and widespread T inversions in leads II,III,avf and V1 to V6.*

CASE 3

AMJ, a 38 year old Pakistani patient, working in Public Security, was seen by a private physician in November 1990 for pain in the right hypochondrium and flatulence. He was found to have hepatomegaly and peripheral oedema. Pericardial disease was suspected and he was referred to the cardiac clinic. In the past he had pericardial effusion in October 1989, and was treated with anti-inflammatory drugs in another hospital. In January 1990, while he was in Pakistan, he was diagnosed to have a cavitary lesion of the right upper lobe and was treated with antituberculous drugs for about three months. Later he was found to have an abnormal ECG and was treated as Ischaemic Heart Disease with congestive Heart Failure.

Physical examination showed grossly engorged jugulars with prominent "Y" descent and Kussmaul's sign. Pulsus paradoxus was elicited and blood pressure was 110/80 mmHg.

The cardiac apex was felt feebly in the normal location with normal S1 and S2 and an early S3 at apex. Electrocardiogram (Fig 5) showed left anterior hemiblock (-45°) with rsŕ pattern in V1 and "T" wave inversion in leads V1 to V6. Radiograph of chest showed prominent pulmonary veins and calcification along the left cardiac border.

Echocardiogram showed restriction of diastolic filling of both the ventricles with normal systolic function, normal valves and thickened pericardium, consistent with constrictive pericarditis. A CT scan of the chest confirmed thickened pericardium with minimal calcification.

The patient has not so far agreed to undergo surgical treatment and is on decongestive medical treatment.

DISCUSSION

When a thick fibrotic and often calcific pericardium restricts the diastolic filling of the ventricles constrictive pericarditis is diagnosed. Cardiac catheterisation has been the gold standard so far for confirming the diagnosis when equilibration of the elevated diastolic pressure in all the four cardiac chambers is documented[4]. The jugular venous pulse reflects these events faithfully and careful observation often gives the clinician a helpful clue. Prominent "Y" descent and inspiratory engorgment (Kussmaul's sign) were present in all the three cases in the present series. In addition, the presence of pulsus paradoxus was also quite helpful.

Non-invasive imaging of the heart plays an important role in confirming the diagnosis[5-7]. Plain radiograph showed pericardial calcification in case 2. However CT scan was needed in case 1 and 3 to identify calcification of pericardium. MRI scanning is likely to be used with greater frequency in future for early detection of constriction as in case 1. Electrocardiogram shows low voltage and non-specific St-T changes. They may sometimes be misinterpreted as due to myocarditis or ischaemia, this delaying correct diagnosis. With the advent of Doppler echocardiography the diastolic function can be studied

reliably[8]. In all the three cases the early, abnormal rapid inflow was detected and 2-D imaging showed calcification. The diagnosis of constrictive pericarditis was thus made non-invasively in all the three cases. Restrictive cardiomyopathy was excluded on the basis of preserved systolic function and normal atria.

Pleural effusion is reported in 50 to 60% cases of constrictive pericarditis in different series[1,9]. Case No. 1 in the present series presented as massive pleural effusion and the subtle features of constrictive pericarditis were not quite obvious initially. The pleural effusion in case 2 was small, but was overshadowed by the presentation of the patient as abdominal discomfort with hepatosplenomegaly and ascites. Abdominal symptoms are known to be the only presenting symptoms in a significant proportion of cases. Case No. 3 also presented with indigestion. The elevated jugular venous pressure and the characteristic pulsations helped in the clinical diagnosis.

Successful treatment consists of specific treatment when aetiology is known and pericardiectomy[10]. All the three cases turned out to be tuberculous in aetiology. In developed countries tuberculosis accounts for only 15% of the cases at present[11,12]. However, in developing countries this is still the most common cause of constriction as exemplified by the present report. Surgical resection, however is essential in the vast majority of cases for complete cure to relieve the mechanical effects of compression.

REFERENCES

1. Tomaselli G, Gamsu G, Stulbarg MS. Constrictive Pericarditis presenting as Pleural Effusion of Unknown. Arch Intern Med 1989;149:201-3.
2. Arora A. Constrictive Pericarditis Masquerading as Budd - Chiari Syndrome. J Clin Gastroenterol 1991;13:178-81.
3. Williams AT. Chylous ascites should suggest constrictive pericarditis even in the presence of Cirrhosis. J Clin Gastroenterol 1990;12:581-4.
4. Lorell BH, Grossman W. Profiles in Constrictive pericarditis, restrictive Cardiomyopathy and Cardiac Tamponade. In: Grossman W, ed. Cardiac catheterisation and Angiography. 3rd ed. Philadelphia: Lea and Febiger, 1986:427-45.
5. Hoit BD. Imaging the pericardium. Cardiovasc Clin 1990;8:663-71.[65 ref]
6. Sechtem U, Tscholakoff D, Higgins CB. MRI of the abnormal pericardium. Am J Roentgenol 1986;147:245-52.
7. Brockington GM. Constrictive Pericarditis. Cardiovasc Clin 1990;8:645-61. [108 ref]
8. Mancuso L. Constrictive Pericarditis Versus restrictive cardiomyopathy: the role of Doppler echocardiography in differential diagnosis. Int J Cardiol 1991;31:319-27.
9. Weiss JM, Spodick DH. Association of left pleural effusion with pericardial disease. N Engl J Med 1983;308:696-7.
10. De Valeria PA. Current indications, risks and outcome after pericardiectomy. Ann Thorac Surg 1991;52:219-24.
11. Cameron J, Oestorle SN, Baldwin JC, Hancock EW. The etiologic spectrum of constrictive pericarditis. Am Heart J 1987;113: 354-60.
12. Roberts WC, Spray TL. Pericardial Heart Disease: A Study of its causes, consequences and morphologic features. In: Spodick DH, ed. Pericardial Diseases. Philadelphia: FA Davis Co., 1976:11-65.