CASE REPORT
IDIOPATHIC CALCIFIC CONSTRICTIVE PERICARDITIS: AN UNSETTLING REALITY

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ABSTRACT

Background: *Constrictive pericarditis is a disease characterized by the encasement of the heart by a rigid non-pliable pericardium due to dense fibrosis and adhesions. This causes impaired diastolic cardiac function leading to heart failure manifested as systemic without pulmonary congestion. Constrictive pericarditis should be suspected in all patients with findings suggestive of right heart failure or ascites. In around one third, no underlying cause is identified. Survival is dependent on the underlying aetiology and patients with idiopathic constrictive pericarditis tend to have best outcome after pericardiectomy.*

Methods: *We reviewed the case of a 35-year-old man who presented with a 6-year history of recurrent dyspnoea on exertion and a 3-year history of progressively increasing abdominal and bilateral leg swelling. There was no previous history suggestive of tuberculosis.*

Results: *Chest x-ray done showed a calcified pericardium. ECG revealed features of atrial fibrillation and echocardiography showed a moderately thickened pericardium with minimal effusion. It also showed evidence of constriction. The right side of the heart was dilated with evidence of pulmonary hypertension.*

Keywords: *Pericarditis, constrictive, calcific, idiopathic*

CASE REPORT

A 35-year-old man presented with a 6-year history of recurrent dyspnoea on exertion which progressed over time to occur at rest and a 3-year history of progressively increasing abdominal and bilateral leg swelling. He had no previous history suggestive of tuberculosis or irradiation. When examined, he was found to be acute-on-chronically ill-looking. He also had gynaecomastia, leuconychia, scanty axillary hair, generalized hyperpigmentation and bilateral pitting pedal oedema up to his knees. His pulse rate was 72 beats per minute, small volume and irregularly irregular. There was however no pulse deficit. Blood pressure was 80/50 mmHg and his jugular venous pressure was raised. Apex beat was located at the 4th left intercostal space, mid-clavicular line. Heart sounds were 1st and 2nd (both soft) with a pericardial knock. Breath sounds were vesicular and decreased over the right lung base. His abdomen was grossly distended with a girth of 66cm (44cm below the xiphisternum).His liver was ballotable 8cm below the right costal margin (firm and tender). He had ascites demonstrable by a fluid thrill.

Serum electrolytes done showed hyponatraemia (121 mmol/L). He also had hyperkalaemia (6.3 mmol/L) which was corrected. His packed cell volume was 34% and he had an erythrocyte sedimentation rate of 5 mm/hr. His total white blood cell count was 3.0 x 10^9/L with a normal differential. Human Immunodeficiency Virus screening was negative. Mantoux test was also negative (0 mm). His saturation of peripheral oxygen (SpO_2) was 93% in room air. His chest x-ray showed a calcified pericardium, a full pulmonary conus and features of pulmonary congestion (Figure 1). His ECG done showed atrial fibrillation, widespread T-wave inversion (inferior and anterior leads) and evidence of levorotation. Echocardiography done showed a moderately thickened pericardium with minimal effusion. It also showed evidence of constriction. The right side of the heart was dilated with evidence of pulmonary hypertension. His ejection fraction was however normal (70%). A diagnosis of calcific constrictive pericarditis was made and he was admitted for an abdominal paracentesis during

which 8 litres of fluid were removed. He was also placed on low-dose diuretics (frusemide and spironolactone), aspirin, hepatotonics. He felt better few days into admission and was subsequently referred for pericardiectomy.

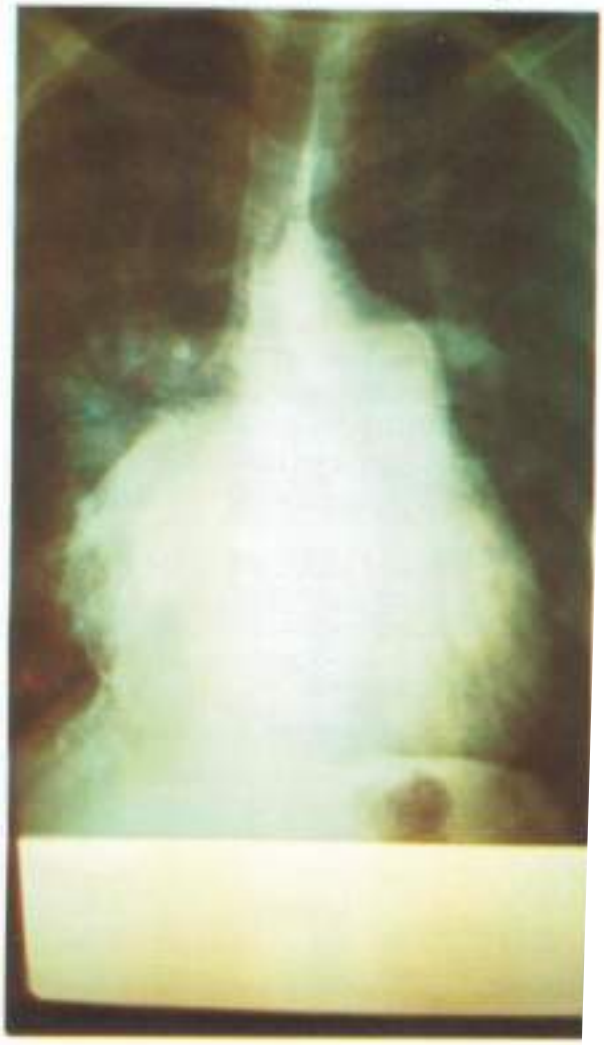

Figure 1: Plain chest x-ray showing cardiomegaly with calcification of the pericardium and fullness of the pulmonary conus.

DISCUSSION

Constrictive pericarditis is a process of chronic fibrous thickening of the pericardium, which is frequently accompanied with calcification and prevents the diastolic filling of the heart, reducing venous return and lowering output. The heart is usually encased by a rigid non-pliable pericardium due to dense fibrosis and adhesions. This causes impaired diastolic cardiac function leading to heart failure manifested as systemic without pulmonary congestion. The disease process usually results from chronic inflammation of the pericardium, which leads to pericardial scarring, thickening, fibrosis, and calcification.

Many aetiologies have been identified, such asinfection, previous cardiacsurgery and mediastinal radiotherapy. Other recognised but rare causes include connective tissue disorders, malignancy and local trauma. In around one third of cases, no underlying cause is identified (idiopathic), although adenovirus and echovirus infections have been implicated. A study published by Hancock in 1971 showed that most cases of constrictive pericarditis he studied were idiopathic. The next most common cause was radiation with tuberculosis being the least common cause (out of the three). This may have obviously been due to the geographical region in which the study was done. Bertog *et al.*, in 2004, reported the aetiology of constrictive pericarditis in the cohort they studied as idiopathic (or viral) in 46% (75 patients).

Tuberculosis is the commonest cause of pericarditis in our environment but this is not always the case. Tuberculosis is commonly presumed to be the cause of constrictive pericarditis but is infrequently proven. In only 3 of the 27 cases of constrictive pericarditis studied by Mabogunje *et al.* was there histological evidence of tuberculosis. Mabogunje *et al.* also observed that some of those in the non specific chronic inflammatory group may well havehad tuberculous or viral pericarditis initially, though others may have resulted from inadequately treated purulent pericarditis.

Clinical signs include oedema, ascites, raised jugular venous pressure, pleural effusion, and hepatomegaly. Around one third of patients with constrictive pericarditis have co-existent atrial fibrillation. Plain chest radiography may show pericardial calcification and unexplained pleural effusion. Echocardiography may confirm the presence of small ventricular dimensions with preserved systolic function (as observed in the index patient) and dilated atria. Abrupt termination of diastolic filling may cause a characteristic 'septal bounce'. All four cardiac chambers become encased so that diastolic pressure tend to equalise between them, which results in systemic venous engorgement and diminished ejection volume. Histopathological features include localised fibrosis and calcification within the pericardium.

Observational studies and case reports suggest that in most instances, constrictive pericarditis without surgical intervention causes a progression of symptoms and, frequently, early death. Furthermore, the overwhelming majority of operative survivors experience an improvement in

functional class or quality of life. Hence, with rare exceptions, pericardiectomy is the treatment of choice for symptomatic constrictive pericarditis. Total rather than partial pericardiectomy yields better outcome, and should be undertaken early, where possible.

It is however regrettable that many patients in our environment with chronic constrictive pericarditis present late when irreversible myocardial or hepatic insufficiency has developed. Death in these patients (post-pericardiectomy) is usually due to myocardial failure presumably the result of fibrosis or myocardial fibre atrophy. Death may also occur (post-pericardiectomy) due to liver failure as seen in the Mabogunje *et al.* Study. Survival is dependent on the underlying aetiology, and patients with idiopathic constrictive pericarditis tend to have best outcome after pericardiectomy.

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