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A Surgical Approach: A Heart in a Rigid Cage

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Abstract **Case Report**

Constrictive pericarditis is an uncommon disease with multiple aetiologies. The most common cause usually is idiopathic. In developed countries, constrictive pericarditis is usually a consequence of previous cardiac surgeries and tuberculosis remains as the common causes especially in developing countries. Medical treatment may temporarily relieves the symptoms but most patients undergoes pericardiectomy has better prognosis than medical treatment alone; in comparison on the outcome of total pericardiectomy and subtotal pericardiectomy done in these patients.

Keywords: Constrictive pericarditis, total pericardiectomy, subtotal pericardiectomy, tuberculosis.

INTRODUCTION

Pericardium is a fibroserous sac surrounding the heart and roots of the great vessels consisting of 2 components, the fibrous pericardium and serous pericardium. Fibrous pericardium is a though connective tissue outer layer that defines the boundaries of middle mediastinum. Serous pericardium is thin and consisting of 2 parts; the parietal and visceral layer (epicardium) [1].

Constrictive pericarditis is a form of diastolic heart failure that arises as the result of scarring and consequent loss of normal elasticity of pericardial sac. Pericardial constriction is typically chronic, but variants include subacute, transient and occult constriction.

Pathologically, constrictive pericarditis arises after a cycle of injury and inflammation, which might be initiated several days, months, or years before clinical presentation. Damage to pericardial mesothelial cells is associated with an acute reduction of tissue-type plasminogen activator and reduced fibrinolytic activity, with fibrinous inflammation and adhesion formation, which is a possible mechanism for pericardial fibrosis, although this mechanism is yet to be proven [2]. Grossly, the pericardium is considerably thicker than normal in approximately 80% of cases.

Risk factors of constrictive pericarditis include prior cardiac surgery (11 to 37%), radiation therapy (9 to 31%, mostly for Hodgkin's disease or breast cancer), associated with connective tissue disorder (3 to 7%), but most cases are still deemed to be idiopathic (42 to 49%) [3, 4]. Tuberculosis accounted for 49 percent of cases of constrictive pericarditis in a series reported in 1962 [5] but is now only a rare cause of constrictive pericarditis in developed countries. However, this disorder may be increasing among immigrants from underdeveloped nations and patients with HIV infection.

CASE REPORT (1)

A 27 years old male with no known medical illness, working as a trainer in a team building activities company; presented to emergency department with complains of worsening shortness of breath on exertion, reduced effort tolerance, orthopnea, paroxysmal nocturnal dyspnea and bilateral lower limbs swelling. Initial echocardiogram shows massive pericardial effusion; EF 40 to 45%, paradoxical movement, right atrium wall of 4cm, apical lateral wall of 4cm and posterior wall of 3cm. Patient was admitted for pericardial tapping; drained 1150cc of tea coloured fluid with some fibrin and sediments. Echocardiogram post pericardial tapping shows EF 55 to 60%, minimal pericardial effusion around 1 to 2cm with trivial mitral regurgitation and mild tricuspid regurgitation.

A CT thorax was performed post pericardial tapping; gross pericardial effusion, located right pleural effusion with differential diagnosis includes tuberculosis, malignancy with features of lung parenchymal are in favours of infection in which tuberculosis needs to be considered. Patient was treated as pulmonary tuberculosis with disseminated tuberculosis in view of CT thorax findings of possible

pulmonary tuberculosis and the nature of massive pericardial effusion with presence of fibrin materials. Anti tuberculosis medications was started. Pleuroscopy was done and noted minimal collection. In pleural cavity.

Subsequently, pericardial fluid sent for mycobacterium tuberculosis culture and sensitivity shows mycobacterium tuberculosis complex in the pericardial fluid samples. Patient was subsequently treated as tuberculosis pericardium and had completed anti tuberculosis medications for 9 months.

A cardiac MRI was done shows presence of focal pericardial thickening (about 4mm at its maximum thickness involving free right and left ventricles and also in apex), compressed / conical shaped right ventricle and diastolic septal bounce which suggestive of constrictive pericarditis, liver enlargement (around 19cm in craniocaudal length) and congestion, dilated right atrium (4.9cm in diameter in end diastole) which suggestive of right heart failure. With moderate tricuspid regurgitation.

Patient was preceded with total pericardiectomy. Intra operatively, post induction noted patient's blood pressure reduced and patient was placed on intra-aortic balloon pump (IABP) and was started on inotropes in view of persistent hypotension. An approach of anterior pericardiectomy was performed under off pump technique via median sternotomy route. Transesophageal echocardiogram post-surgery shows improved right ventricular contractility and patient was hemodynamically stable on minimal inotropic support. Post-surgery 4 hours noted patient persistently hypotensive and tachycardia despite on inotropes. Immediate transesophageal echocardiogram was done reveals the right ventricle is not contracting well. Patient diagnosed with right heart failure post pericardiectomy and was started on extracorporeal membrane oxygenation (ECMO).

An echocardiogram was repeated on post pericardiectomy Day 4; right atrium was dilated with intertribal septum towards left atrium with moderate tricuspid regurgitation, right ventricle dilated relative to size of left ventricle with TAPSE of 7mm. Left atrium was dilated with moderate mitral regurgitation with LVEF of 54%. Patient was off from ECMO on post-surgery day 5 and was weaned down to CPAP ventilation. Patient was extubated on post-surgery day 9. Repeated echocardiogram on post-surgery day 12 reveals good left ventricular function with EF54%, right atrium seen dilated around 4.9cm with no pericardial effusion seen. Patient was discharged on post-surgery day 21.

During first follow up on post-surgery 2 months, noted improving in left ventricular function with EF 64%, good movement with no hypokinetia, no pericardial effusion and no pleural effusion seen.

CASE REPORT (2)

A 47 years old male, working as a bus driver, with underlying hypertension, dyslipidemia and history of pulmonary tuberculosis in 2009 and completed anti tuberculosis treatment for 6 months; presented to emergency department with fever, cough and constitutional symptoms. An echocardiogram was done reveals mild pericardial effusion of right ventricle wall of 0.7cm and post left ventricle of 0.6cm, EF 57%, mild tricuspid regurgitation and mild mitral regurgitation. A CECT thorax was done shows loculated pericarditis with proteinous complex fluid contains largest at lateral aspect 2.8cm with no evidence to suggest active pulmonary infection or inflammation.

Subsequently, patient came after 2 months presented with syncopal attack and failure symptoms. An echocardiogram was performed and shows satisfactory left ventricle function with EF 50 to 55%, left ventricle mildly paradoxical movement, left atrium dilated, mild mitral regurgitation, moderate tricuspid regurgitation and pericardial effusion at posterior, inferior and lateral wall. A CECT thorax and abdomen is repeated shows pericardial effusion (around 6.8 x 3.8 x 9.7 cm) with enchanting wall suggesting underlying inflammation causing effusive constructive pericarditis leading to hepatic passive congestion where tuberculous pericarditis has to be excluded.

A MRI cardiac was done reveals mild biventricular dysfunction (LVEF 46%, RVEF 42%) with features in keeping with constrictive pericarditis with complex pericardial collection. A right heart study was done shows constrictive pericarditis with evidence of square root sign in right ventricle tracing during diastole. The study also shows raised RVEDP (23mmHg) and LVEDP 27mmHg, equalisation of diastolic pressure in right ventricle and left ventricle, RVEDP is more than 1/3 right ventricle systolic pressure (0.7mmHg), discordance in left ventricle peak and right ventricle peak during inspiration and right atrium pressure raised during inspiration (Kaussmaul sign).

A subtotal pericardiectomy was done for the patient. Transesophageal echocardiogram pre operatively shows LVEF 43% with moderate left ventricle function, an irregular mass seen at the lateral border of left ventricle wall (about 3 x 6cm) causing left ventricle compression, thicker pericardium with hypoechogenicity, moderate right ventricle function with irregular mass seen at lateral border of right atrium causing compression, trivial mitral regurgitation, mild tricuspid regurgitation. Intraoperatively, noted thick pericardium over left ventricle, right ventricle and right atrium, solidified pericardial collection posterior to left ventricle and also right atrium to inferior vena cava

junction. The fluid is evacuated and pericardium is released over lateral wall to the left ventricle, right ventricle and right atrium wall.

Post operatively transesophageal echocardiogram shows preserved biventricular function, LVEF 50%, mass was removed from the lateral border fo left ventricle wall and the left ventricle is expanded, mass removed and compression is released with no pericardial effusion and pleural effusion seen, mild mitral regurgitation and tricuspid regurgitation. Patient was extubated on post-surgery day 2. Patient was discharged well on post-surgery day 10.

Upon follow up, patient was well with symptoms relieved. An echocardiogram was done reveals LVEF 51%, regional wall motion abnormality seen at mid septal mid anterior region, pericardial effusion seen at basal posterior (0.36 to 0.67cm) with no pleural effusion seen.

DISCUSSION

Chronic constrictive pericarditis is a chronic inflammatory process that involves both fibrous and serous layers of the pericardium, leading to pericardial thickening and compression of the ventricles, thus resulting to reduce cardiac output due to impairment in diastolic filling of ventricles. It was first described in human back in 17th century by the finding of Lower, who described both acute and chronic constrictive pericarditis in 1669. However only after more than 2 century later, both Rehn and Sauerbruck in Germany performed a successful partial pericardectomy in 1913 [6].

In most patients the aetiology of chronic constrictive pericarditis is not known. Prior to its effective treatment, tuberculosis was the aetiology of chronic constrictive pericarditis in up to 17% of cases [7]. Currently, a prominent cause is mediastinal radiation for malignant disease. Trauma is other uncommon causes with hemoperitoneum usually present as the precursor of pericardial thickening and constriction. Both our patients initially presented with sign and symptoms of pulmonary tuberculosis that progress to pericardial tuberculosis. Despite completing anti tuberculosis, constrictive pericarditis occurs later as a complication form the pericardial TB. Classically, symptoms of chronic constrictive pericarditis are delayed for several years after the clinical or subclinical episode of acute pericarditis. The interval may, however, be as short as 3 to 4 weeks in those rare instances in which pericarditis develops after cardiac surgery , or 4 to 12 months after trauma or acute nonspecific pericarditis. Diagnosis of constrictive pericarditis is difficult because patients presented with vague and multiple symptoms. Prior cases series of constrictive pericarditis showed that 75% of patients presented with peripheral edema, 60 % with hepatomegaly and 40 % with ascites [8, 9]. The most important consistent finding in patients with constrictive pericarditis is elevation of jugular venous pressure (JVP) that reported to be present in more than 80% of cases; this is most important clinical sign in patients with constrictive pericarditis because it is not typically seen in patients with chronic liver disease.

Plain chest radiography may reveal pericardial calcification, however, this finding is not specific nor does it confirm constrictive physiology, only 25% of patients with constrictive pericarditis will have pericardial calcification on chest radiograph [10]. Transthoracic echocardiography (TTE) should be the 1st line investigation if constrictive pericarditis is suspected [11]. However there is no single pathognomonic echo finding to diagnose constrictive pericarditis. Echo findings that may present include right and left atrial dilatation, dilated non collapsing inferior vena cava and diastolic septal bounce. Multi detector CT (MDCT) should not be used as the first imaging modality for constrictive pericarditis except in certain condition like end stage calcific pericardial constriction, patients with prior cardiac surgery and radiation to the heart as it provides assessment of parenchymal lung disease and proximity of cardiovascular structures to the sternum. Magnetic resonance imaging (MRI) also can provide assessment and measurement of pericardial thickness and show characteristic right atrial dilatation and right ventricular compression. Cardiac catheterisation is another modality to diagnose constrictive pericardial where the hallmark findings is square root signs in which there is an early rapid fall in diastolic pressure in the right ventricle, followed by a rapid rise to an elevated diastolic plateau [11]. Both cardiac MRI and cardiac catheterisation were performed in our series to help in diagnosing constrictive pericarditis.

Pericardiectomy is the accepted standard of treatment in patients with chronic constrictive pericarditis[6]. The review by Seifert *et al*. states that 80 to 90% of hospital survivors achieved NYHA class I and II functional status after pericardial resection. Patients in class II to III are best recommended for pericardial resection. The mortality rate in Mayo Clinic series 132 was 14%, but Tuna and Danielson reported 1% in classes I and II, and 10% and 46% in class III and IV, respectively [12]. However, surgery should be considered cautiously in patients with either mild or very advanced disease and in those with radiation induced constriction, myocardial dysfunction or significant renal dysfunction [6]. A total pericardiectomy is defined as a wide excision of pericardium, with the phrenic nerves defining the posterior extent, the great vessels (including the intrapericardial portion and the superior vena cava - right atrium junction) defining the superior extent, and the diaphragmatic surface (including the inferior vena cava - right atrium junction) defining the inferior extent of pericardial resection. Both options are invasive with large incisions with significant postoperative pain and

relatively long post-operative recovery. They also confer a mortality rate up to 20% in some institutions [13]. During the procedure, it is important to avoid injury to the phrenic nerves and epicardial arteries; in particular, in fetching the pericardium from the area of left anterior descending (LAD) artery is particularly difficult, an island can be left in this area. Subtotal pericardiectomy is defined as any excision that is less than a total pericardiectomy. Both methods are approached through median sternotomy, either on pump or off pump procedure, where risk of perioperative bleeding is less with off pump method.

In the first case report, patient undergoes total pericardiectomy and post operatively noted patient has right ventricle failure requiring ECMO support. This may be due to long operative time and more manipulation of the heart during the surgery. In the second case report, patient undergoes subtotal pericardiectomy with the aim to relieve right and left ventricle compression, lead to a less stormy post-operative recovery. As seen, a comparison could be made where patient experience lesser post-surgery complications and shorter hospital stay by performing subtotal pericardiectomy for constrictive pericarditis. Both patients have been treated for tuberculosis previously and surgery was done within one year after diagnosis has been made.

CONCLUSION

In conclusion, chronic constrictive pericarditis remain difficult to diagnose and clinicians should have a high index of suspicion in patients presenting with failure symptoms. Tuberculosis remains a common culprit in this part of the world and patients can present years after completion of anti-tuberculosis. Pericardiectomy remain the mainstay of treatment in symptomatic patient in which subtotal pericardiectomy lead to less eventful perioperative period.

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