Case Report

Radiological diagnosis of constrictive pericarditis - a rare disease in Bangladesh

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Abstract

A 35-years old man reported in radiology & imaging department of National Heart Foundation Hospital & Research Institute with palpitation, gradual dyspnoea on exertion and generalized body swelling. The patient had a history of tuberculosis & treated with anti-tubercular drugs for 06 months, one & half years back. His X-ray chest, ECG, Ultrasonogram, Echocardiogram, CT scan of chest & Cardiac CT angiogram were performed and the case was diagnosed as Constrictive pericarditis. A postero-anterior & lateral chest X-ray demonstrated severe, dense calcification of the pericardium. Constrictive pericarditis is a rare disease in Bangladesh. The objective of this report is to discuss the etiology, the patho-physiological features, clinical findings & diagnostic tools of this condition.

Keywords : Constrictive pericarditis, right heart failure & pericardium.

Introduction

Constrictive pericarditis (CP) is a reduction in the elasticity, or stiffening, of the pericardium, a sack-like covering that surrounds the heart, resulting in impaired filling of the heart with blood. Constrictive pericarditis, designated as concretio cordis dates back more than 300 years. The symptoms of Constrictive pericarditis may include exercise intolerance, liver failure, dyspnoea, and renal failure, which appear insidiously and may be misleading.[1,2] In many cases, constrictive pericarditis is late sequelae of an inflammatory condition of the pericardium. The inflammatory condition is usually an infection that involves the pericardium, but it may also occur after a heart attack or after heart surgery.[3]

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Almost half the cases of constrictive pericarditis in the developing world are idiopathic in origin. In regions where tuberculosis is common like Bangladesh, it is the cause in a large portion of cases.[4]

Patients may present with increasing weight gain, cardiac cirrhosis, and massive ascites.[5-7] But the diagnosis is rarely considered by the referring physician. Although rare but it has to be kept in mind as one of the differentials and investigations should be done accordingly.

Patients respond dramatically to a complete surgical pericardiectomy when it is performed early in the disease process; therefore, it is important to consider CP when making the diagnosis. Anatomic imaging findings, such as calcifications (see the images below) and thickening of the pericardium may be present, but the most reliable and most important findings are related to the filling pattern of the heart. Radiological investigations specially x-rays & CT angiogram are playing important roles in the diagnosis of constrictive pericarditis.[8]

Rare forms of constrictive pericarditis: In addition to classical chronic (rigid shell, calcific) & sub-acute (elastic) forms, new presentations such as effusive-constrictive, localized, transient, occult & constrictive pericarditis with normal pericardial thickness, have been described.[9]

Pathophysiology

In the classical form of constrictive pericarditis, the rigid, heavily fibrosed or even calcified pericardium causes

restriction of the myocardium, preventing adequate ventricular filling after an initial expansion.[2, 5-11] Due to decreased pericardial compliance & increased venous pressure, the early diastolic filling occurs very rapidly and stops when the intracardiac volume & pressure reach their maximum limits. As myocardium is not affected, relaxation of the left ventricle is usually normal. Elevated diastolic pressure in all four chambers is due to the rigid pericardium. Consequently, the atrial waves on the jugular veins show a prominent & deep diastolic Y descent (Friedreich sign - diastolic collapse of cervical veins due to adherent pericardium). The hallmarks of constrictive pericarditis are the hemodynamic changes observed during respiration. Increase in the right heart volume, causing higher pressure on the inter-ventricular septum. Subsequently, the septum moves to the left & the left-side volume decreases. Kussmaul's sign is positive (raised JVP on inspiration). Pulse is usually low volume with tachycardia. Pulsus paradoxus may be present - disappearance of peripheral pulse during inspiration. This interesting phenomenon is explained by impaired filling of the left ventricle with an enhanced decrease in the systolic pressure during inspiration caused either by fibrosed & calcified pericardium or by the accumulation of pericardial fluid. It has been found that constricted pericardium are not able to increase the flow velocity in either the vena cava or the pulmonary artery during inspiration, a finding that may be the mechanism for the appearance of pulsus paradoxus.[12] Pooling of blood in the pulmonary bed & filling competition of the ventricles in the presence of a fixed pericardial sac may serve as an additional cause for appearance of pulsus paradoxus. These dynamic changes throughout the respiratory cycle can be used to differentiate constrictive pericarditis from restrictive cardiomyopathy.

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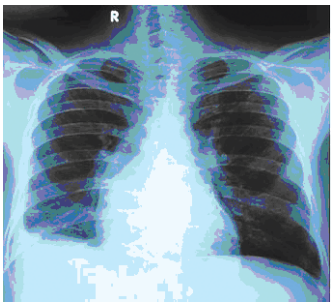
A 35-years old man reported in radiology & imaging department of National Heart Foundation Hospital & Research Institute with Palpitation, gradual dyspnoea on exertion and generalized swelling of the body. The patient had a history of tuberculosis & treated with anti-tubular drugs for 06 months, one & half years back.

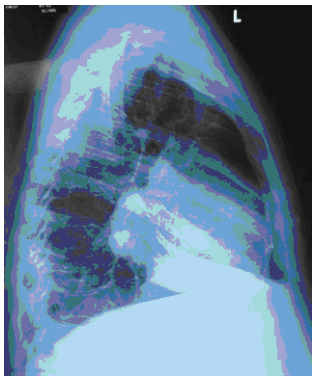
His X-ray chest, ECG, Ultrasonogram, Echocardiogram, CT scan of chest & Cardiac CT angiogram were performed. A postero-anterior & lateral chest X-ray demonstrated severe, dense calcification of the pericardium. ECG findings showed ischemia, atrial premature beats with evidence of atrial fibrillation. Ultrasonogram of abdomen showed hepatomegaly with evidence of hepatic venous congestion with ascites. Findings of echocardiogram were consistent with constrictive pericarditis. The pericardium was severely & diffusely calcified on CT scan (as seen also in 3D view).

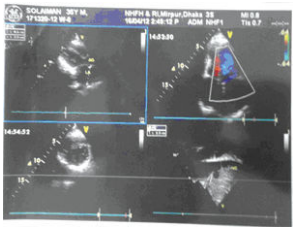
Investigations

(a) Chest X-ray :- Both the P/A & lateral views demonstrate severe, dense, calcification of the pericardium. Calcifications are mostly seen at the left heart border, anterior, inferior as well as diaphragmatic surfaces. Cardiac silhouette is not enlarged. There is also evidence of small pleural effusion on the right side.

(b) Electrocardiography : - This patient shows characteristic ECG changes for constrictive pericarditis. There is presence of tachycardia with low QRS voltage, ischemia, atrial extra systole with fibrillation.



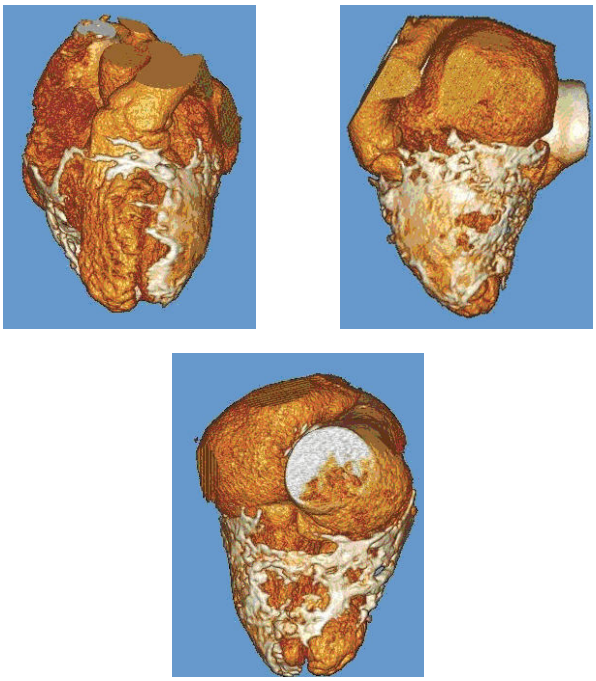




(c) Echocardiography : - In two dimensional Echocardiogram, parietal & visceral pericardium are adherent particularly at the left lateral & inferior wall of the heart with speckled calcification causing poor echo-window. There is an unusual motion of the

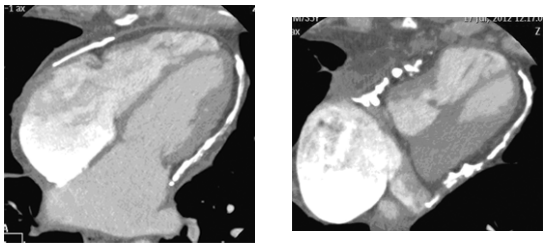
interventricular septum, designated as septal bounce, accompanied by an inspiratory septal shift to the left. The presence of a dilated, non-collapsing inferior vena cava is suggestive of constrictive pericarditis. On the other hand, a normal sized collapsing inferior vena cava at inspiration almost rules out heart constriction.

The hallmark of constrictive pericarditis on Doppler echocardiography is the respiratory flow variations through the heart valves, expressed as a decreased flow through the left side valves during inspiration & its increase with expiration, while on the right side valves the reverse phenomenon occur.



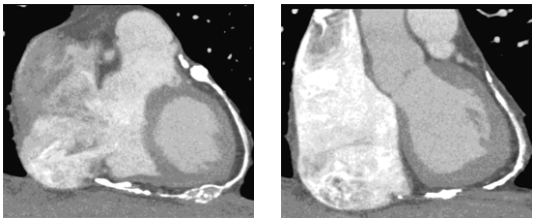
(d) CT angiogram : - This method allows direct visualization of the pericardium as well as heart. Pericardium is thickened (04 mm) with evidence of hyper-dense areas at the left lateral, anterior and inferior as well as diaphragmatic surfaces.

3D view shows calcified layer of the pericardium which looks like a cemented layer causing constriction of the heart.

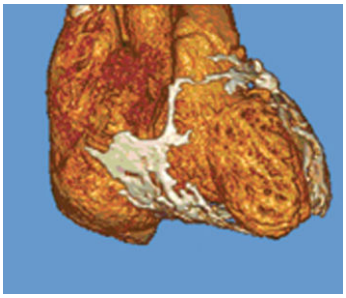


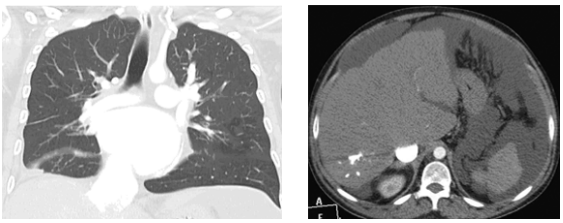
CT scan axial view showing the calcified pericardium

Four chambered heart is seen in axial views. There is evidence of increased concentration of the dye seen in the right side of the heart. Right atrium is enlarged in size. Both the ventricles are appearing tubular with evidence of pressure effects from outside (due to calcification) causing diastolic ventricular dysfunction secondary to constrictive pericarditis.



CT scan coronal views showing the calcified pericardium





Others

There is evidence of small pleural effusion on the right side. There is also huge ascites in the abdomen.

Discussion

The diagnosis of constrictive pericarditis is difficult because of its common clinical signs with many other possible diagnoses and the rarity of this condition. Constrictive pericarditis should be considered in patients with signs of right heart failure.[38]

Patients with constrictive pericarditis frequently present with symptoms of heart failure, such as dyspnoea, orthopnoea, and fatigability, and occasionally may present with liver enlargement and ascites. The causes of constrictive pericarditis have changed over time; at present, the most frequent causes are cardiac surgery and

radiation therapy.[21] Other causes include infection (viral or tuberculous), connective-tissue disease, uremia, neoplasm, or idiopathic condition.[22] Clinically, it is difficult to differentiate between constrictive pericarditis and restrictive cardiomyopathy. These two entities are characterized by similar clinical manifestations and similar findings at cardiac catheterization and echocardiography. In both conditions, ventricular filling is restricted, leading to an increase in diastolic pressure in all four cardiac chambers and to equalization of atrial and ventricular pressure. It is important, however, to distinguish between constrictive pericarditis and restrictive cardiomyopathy, because patients with constrictive pericarditis might benefit from pericardial stripping, whereas those with restrictive disease would not.

Traditionally, increased pericardial thickness has been considered a specific diagnostic feature of constrictive pericarditis, but these days there is also a subset of patients with hemodynamic signs of constrictive pericarditis and normal thickness of the pericardium.[39] Several criteria on invasive pressure tracings have been examined for the diagnosis of constrictive pericarditis. Among these, the change in the ventricular pressure curves during the respiratory cycle (reflecting ventricular interdependence) is unique to patients with constrictive pericarditis.[40]

Imaging is essential in arriving at the correct diagnosis.[41-43] Many imaging modalities offer potential choices for making the correct diagnosis. Plain radiography is helpful, but notoriously insensitive for demonstrating pericardial calcifications. Other plain radiographic evidence of constrictive pericarditis may be seen including cardiac enlargement, left atrial enlargement, and pulmonary vascular congestion.[44]

Echocardiographic findings indicate wall motion abnormalities and cardiac morphologic features consistent with constrictive pericarditis, and may demonstrate pericardial calcifications and thickening as well. Although transthoracic echocardiography is routinely performed for the evaluation of myocardial function in patients with symptoms of constrictive or restrictive physiologic change, it is not highly accurate in the depiction of pericardial thickening.[4] Transesophageal imaging allows better visualization of the pericardium, and respiration-correlated Doppler techniques are particularly useful in the diagnosis of constrictive pericarditis.[23] However, the transesophageal approach is limited by a narrow field of view and is relatively invasive.

The diagnosis of constrictive pericarditis is greatly aided by the excellent depiction of the pericardium at CT and MR imaging. Normal pericardial thickness is less than 2 mm.[8,9] Pericardial thickness of 4 mm or more indicates abnormal thickening and, when it is accompanied by clinical findings of heart failure, is highly suggestive of constrictive pericarditis.

MR imaging has a reported accuracy of 93% for differentiation between constrictive pericarditis and restrictive cardiomyopathy on the basis of depiction of thickened pericardium (4 mm).[24] Pericardial thickening may be limited to the right side of the heart or to an even smaller area, such as the right atrioventricular groove[25]. But MRI has difficulty in demonstrating calcification. Other findings seen on MRI and present in this case include septal bounce, enlarged atria, and small cone-shaped ventricles.[45,46]

An additional advantage of CT is its high sensitivity in depicting pericardial calcification, which is also associated with constrictive pericarditis. CT scan exquisitely demonstrate pericardial calcifications, but gated imaging is necessary to prove constriction, and the temporal resolution of CT is inferior to other dynamic modalities for imaging the heart, including MRI and echocardiography.[46,47] It is important to remember, however, that neither pericardial thickening nor calcification is diagnostic of constrictive pericarditis unless the patient also has symptoms of physiologic constriction or restriction.

At both CT and MR imaging, the central cardiovascular structures may show a characteristic morphology in constrictive pericarditis. The right ventricle tends to have a reduced volume and a narrow tubular configuration. In some patients, a sigmoid-shaped ventricular septum or prominent leftward convexity in the septum can be observed.[25] In the setting of dense calcifications or in the presence of normal pericardial thickness, however, evaluation of the pericardium can be difficult.[42,49,50] Systemic venous dilatation (particularly in the inferior vena cava), hepatomegaly, and ascites also are frequently seen.

No single approach should be used to diagnose all cases of constrictive pericarditis. The diagnostic approach taken should be individualized for each patient. In some patients, the diagnosis may be made on the basis of the history, physical examination, and chest radiograph. In other patients, echocardiography, visualization of the pericardium, and cardiac catheterization all may be

required. The most important diagnostic tool is the clinical suspicion of constrictive pericarditis in a patient with signs and symptoms of right sided heart failure that are disproportionate to pulmonary or left sided heart disease. Understanding the pathophysiology of this disease and using non-invasive and invasive techniques are helpful in diagnosis, particularly in the patient who has myocardial and pericardial disease. The differential diagnosis is right atrial myxoma, tricuspid valve dysfunction & restrictive cardiomyopathy, nephrotic syndrome, obstruction of the superior vena cava, hepatic diseases & abdominal malignancies.

In the chronic stage of tuberculous constrictive pericarditis, pericardial decortication with wide resection of both the visceral and the parietal pericardium, remains the definitive treatment.[51] However, there are no clear-cut determinants for surgical intervention when managed at the early stage. Yang et al. once reported that decision largely depended on the clinical symptoms of cardiac temponade, progression of heart failure, and constriction that lead to jugular vein engorgement.[52] They highlighted the importance of pursuing early pericardiectomy, rather than pericardiocentesis and window placement, to achieve sustained relief of symptoms in patients with advanced stage disease.

Conclusion

It is very important that the sooner the diagnosis is established the better the outcome is. Constrictive pericarditis is caused by fibrosis and calcification of the pericardium, processes that inhibit diastolic filling of the heart. This condition has posed a diagnostic dilemma since it was first recognized clinically. Because surgical intervention can provide complete relief of symptoms in many patients, accurate diagnosis of this disorder is important.[3] By the use of diagnostic tools such as X-ray Chest, Echocardiography, CT scan for heart visualization are important & may save patient's anguish & complication. With the advent of CT Angiogram the diagnosis of the constrictive pericarditis have become very easy, especially 3D views.

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