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Case Report

Chronic constrictive pericarditis: an "Egg-Shell" calcification

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ABSTRACT

Chronic constrictive pericarditis is a condition marked by scarred and inelastic pericardium with excessive fibrous tissue and calcium. Here, we report a case of Egg-Shell like calcification in a 30-year-old young adult diagnosed with chronic constrictive pericarditis. The patient was successfully managed with very good prognosis post-pericardiectomy.

Keywords: Calcification, Computed tomography, Constrictive pericarditis, Fluoroscopy, Pericardiectomy

INTRODUCTION

Chronic constrictive pericarditis is an end-stage disease of pericardium, which has been considered as a quite unusual form of clinical heart failure. It involves chronic inflammatory responses that lead to fibrosis, calcification and adhesion of visceral and parietal pericardium.[1,2] The etiological parameters of constrictive pericarditis include idiopathic, viral infection, post-cardiac surgery, post-radiation therapy, post-infection (tuberculosis or purulent pericarditis) and other miscellaneous factors (malignancy, trauma, drug-induced, connective tissue disease such as rheumatological diseases and systemic lupus).[1,3,4]

Extensive pericardial calcification around the whole heart is very unusual in constrictive pericarditis and if recognised it indicates the chronicity and severity of the disease. Here, we report a very interesting case of 30-year-old male who was diagnosed with idiopathic chronic constrictive pericarditis with extensive "Egg-Shell" like calcification of entire pericardium. The patient underwent successful pericardiectomy and remained uneventful post-surgery.

CASE REPORT

The patient was a 30-year-old young male who presented with dyspnoea New York Heart Association (NYHA) functional class-III and peripheral oedema since last 5 months. An elevated jugular venous pressure (15 cm H_2O) with prominent 'x' and 'y' descents was observed on examination and auscultation revealed presence of pericardial knock. Hemodynamic examination showed elevated mean right atrial pressure (30 mmHg; normal: 2-6 mmHg), right ventricle end-diastolic pressure of (32 mmHg; normal: 2-8 mmHg) and left ventricle end-diastolic pressure (33 mmHg; normal: 10 mmHg) which showed nearly equal end-diastolic pressures of right atrium and right and left ventricles. The mean pulmonary capillary wedge pressure was also elevated (30 mmHg; normal: 4-12 mmHg). Both left and right ventricular pressure tracing suggested constrictive pericarditis through typical "dip-and-plateau" (square root sign) (Figure 1). All echocardiographic parameters suggested a classical picture of constrictive physiology with septal bounce and 11-mm thick calcified pericardium. Echocardiography also revealed 25 % variation in mitral inflow velocity with respiratory movements (Figure 2A) and diastolic flow reversal of hepatic vein during

expiration (Figure 2B). Fluoroscopic examination confirmed presence of dense circumferential calcification all around the heart (Figure 3). CT-scan of thorax also confirmed presence of Egg-Shell like calcification surrounding whole heart (Figure 4 and 5).

Figure 1: Ventricular pressure tracing shows typical "dip-and-plateau configuration" during diastole.

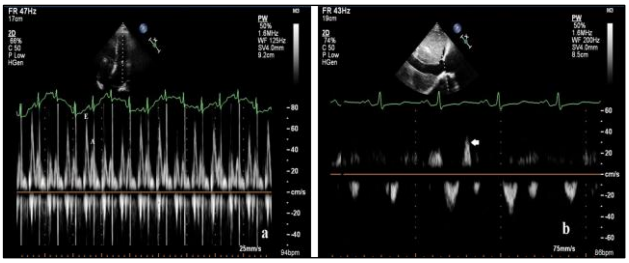

Figure 2: Echocardiographic examination (A) Echo image of variation in mitral in-flow with respiration and (B) Echo image of diastolic flow reversal of hepatic vein with expiration.

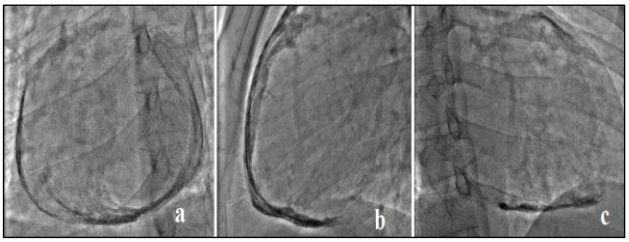

Figure 3: Fluoroscopy images in LAO 40 (a), LATERAL (b) RAO 40 (c) circumferential pericardial calcification around the heart.

As surgery is the only option for heavily calcified constrictive pericarditis, the patient underwent surgical pericardiectomy to remove densely calcified pericardium. Histo-pathological studies of resected tissues revealed presence of extensive fibrosis, hyalinisation and calcification without any evidence of granulomas which was consistent with constrictive pericarditis. Patient showed quick recovery without any adverse events and thus was discharged on 8th day post-surgery.

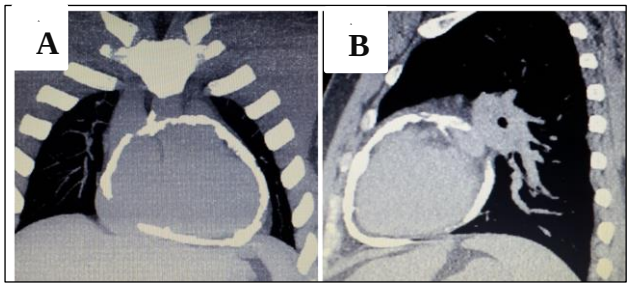

Figure 4: Non-contrast computed tomography (CT) reconstructed images in coronal (A) and sagittal (B) planes show thick, calcified pericardium around the heart.

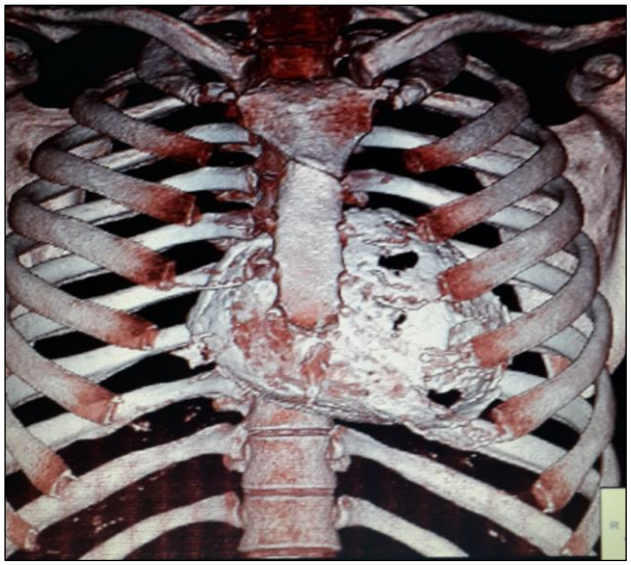

Figure 5: Non-contrast CT reconstructed volume rendered image shows pericardial "Egg-Shell" calcification around the heart.

DISCUSSION

Chronic constrictive pericarditis is characterised by marked restricted diastolic filling of heart due to chronic inflammation that results in accumulation of excess fibro-elastic tissues and calcium in pericardium. The diagnosis of constrictive pericarditis often overlooked as it mimics various other cardiac disorders but should be considered if the patient represent classic heart failure symptoms with preserved ventricular ejection fraction and elevated jugular venous pressure.[1,4,5] A typical diagnostic feature of constrictive pericarditis is an inconstant equalization of end-diastolic pressures of all four chambers of heart and pulmonary venous pressure. Echocardiography is used as a first-line imaging modality for the diagnosis of constrictive pericarditis. Other multi-modality imaging techniques such as chest X-ray, CT-scan and magnetic

resonance imaging act as secondary tool that help to confirm the presence of constrictive pericarditis with detailed anatomical specification that even detect minute pericardial calcification. Cardiac catheterization, in patients with suspected constriction, is also supportive in delineating hemodynamics, to differentiate constriction and restrictive cardiomyopathy and helpful to rule out coronary compression by calcified thickened pericardium.[1,3,5]

Surgical pericardiectomy is the principle treatment for the management of chronic constrictive pericarditis with very good prognosis and if left untreated it may progress to the worse level which even lead to death. The perioperative mortality of surgical pericardiectomy was 2-15% as observed in various previous studies and mostly dependent on preoperative NYHA class.[1,4,6]

Severely calcified constrictive pericarditis has higher risk of adverse events following surgical pericardiectomy as calcification point towards the chronicity of the disease. However, the patient in the present case successfully survived the surgical pericardiectomy without any perioperative events despite of having idiopathic chronic constrictive pericarditis with 11 mm thick calcified pericardium. Cases with this level of calcification are less commonly observed.

CONCLUSION

Timely diagnosis and appropriate management of chronic constrictive pericarditis is very crucial to expand life span as well as to improve quality of life of the patient. Appropriate use of various imaging modalities guide the investigators to discriminate among different analogous conditions and thus should be considered mandatory for diagnosis of such complex conditions. Similarly, in present case also the disease was diagnosed in time using various imaging techniques. Even though the patient had 11 mm thick “Egg-Shell” like calcified pericardium, he was wisely managed without any event post-pericardiectomy and was hemodynamically stable.

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