

Aberrant Shape of the Heart, “Gourd Shaped”: A Rare Case of Localized Constrictive Pericarditis

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INTRODUCTION

Constrictive pericarditis (CP) is a chronic inflammatory process leading to scarring, fibrosis and calcification of the pericardium that leads to diastolic dysfunction.^{1,2} The leading cause of constrictive pericarditis are idiopathic and viral infection in developed countries, however tuberculosis remains the leading cause in developing countries.^{1,3-5} The importance in early identification and appropriate treatment is reflected in the potential curability of the condition.²

Here we present a case of 49 years old lady with the localized calcific constrictive pericarditis, with a rare site of calcification.

ABSTRACT

Calcific constrictive pericarditis results commonly from the tubercular lesions in developing countries. The localized calcifications associated with the calcific constrictive pericarditis on its own is rare and the calcification along the atrioventricular groove distorting the heart to a shape of gourd is extremely rare. We present a case to highlight the importance of the early diagnosis of the condition. We have approached the patient with echocardiography and computed tomography for the diagnosis and hemodynamic evaluation. Early treatment is equally important for this condition, as the calcific constrictive pericarditis is curable with pericardiectomy. However, there lacks a clear consensus for the pericardiectomy in patients with localized calcification and medical therapy can be initiated.

KEY WORDS

Constrictive pericarditis, Computed tomography, Echocardiography, Pericardiectomy

CASE REPORT

A 49 years old female presented at Shahid Gangalal National Heart Centre on 8th of July, 2024 with complaints of dyspnea and palpitation NYHA class II for 1 year. She also had bilateral pedal edema, pitting type for the past 1 year which was progressive. The pedal edema has progressed upto the knee for the past 2 months.

The examination revealed raised jugular venous pressure with prominent y descent. Pericardial knock could be heard during auscultation. Laboratory tests at the time of admission including complete blood count, renal function tests, liver function tests, thyroid function tests did not reveal any abnormality. The findings of electrocardiography and echocardiography, done during the time of admission, are noted.

Electrocardiography showed atrial fibrillation with controlled rate (Fig. 1).

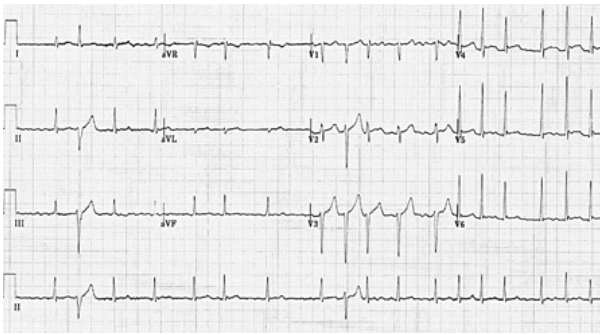


Figure 1. ECG of the patient with Atrial Fibrillation

Echocardiography: It was significant for diastolic septal bounce, annulus reversus, dilated left atrium, compression of both the ventricles, plethoric and non-collapsible inferior vena cava during inspiration (Fig. 2-7).

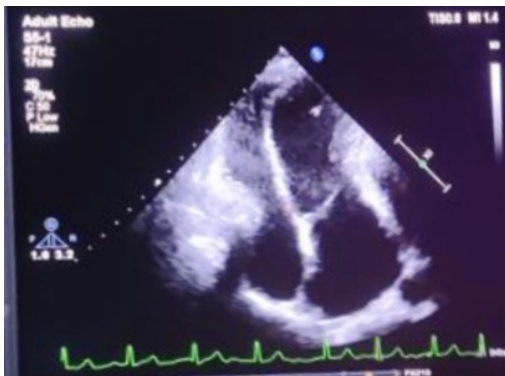


Figure 2. Echocardiography of the patient with gourd shaped heart heart

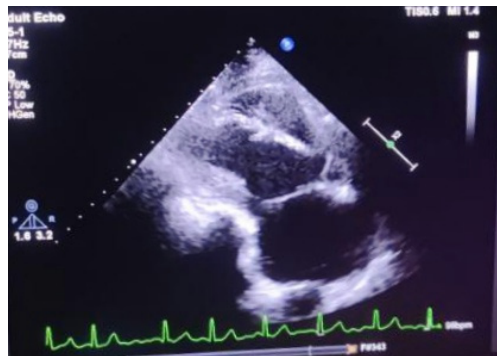


Figure 3. Echocardiography of patient with gourd shaped heart with localized constriction at left atrioventricular groove and dilate Left Atrium



Figure 4. Echocardiography of patient with gourd shaped heart showing inflow variation across tricuspid valve

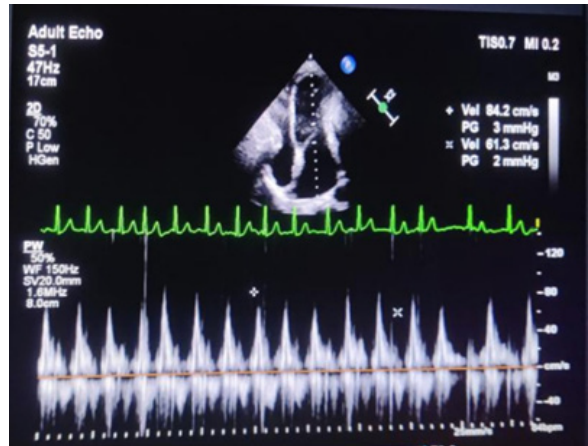


Figure 5. Echocardiography of the patient with gourd shaped heart with inflow variation across mitral valve

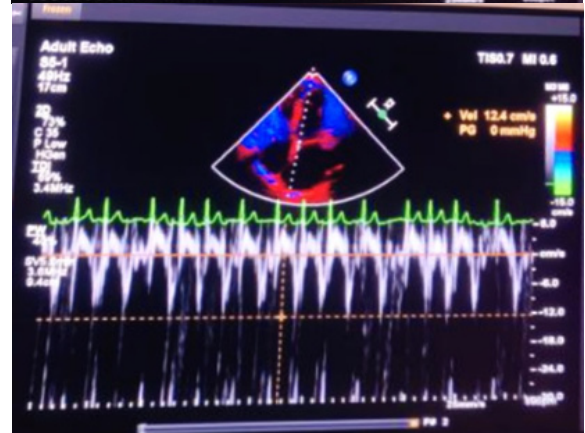
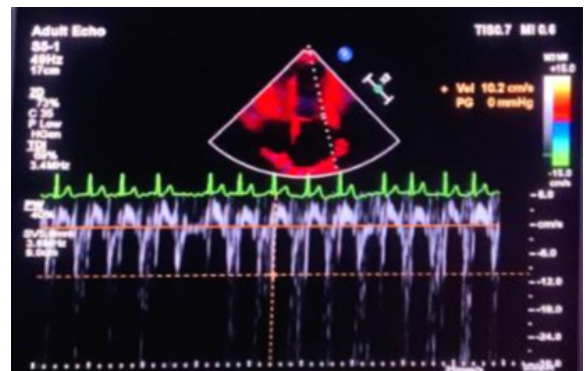


Figure 6. Echocardiography of the patient with gourd shaped heart showing annular reversus (medial annular velocity lower one is more than lateral annular velocity upper one)

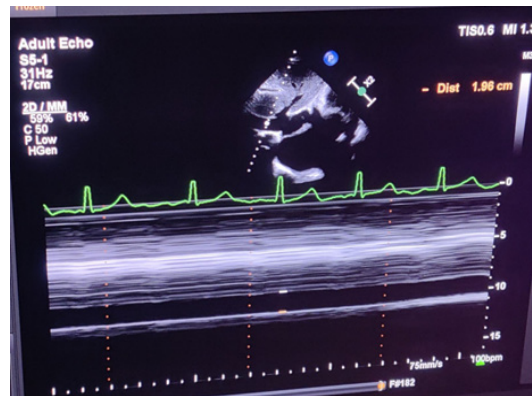


Figure 7. Echocardiography of patient with gourd shaped heart dilated and non-collapsible IVC



Figure 8. Echo assessment of left ventricle function

CT scan: It demonstrated ring shaped extensive calcification of the pericardium trapping along the atrioventricular groove altering the shape of the heart, resulting into a gourd shaped heart. There is associated thickening of pericardium with maximum thickness measuring approximately 6.0 mm (Fig. 8).



Figure 9. Calcification of the pericardium trapping along the atrioventricular groove

The patient was started on diuretics, oral torsemide 20 mg twice a day, and anticoagulant, oral rivaroxaban 20 mg once a day, in view of atrial fibrillation. The shortness of breath improved with the diuretics and edema also decreased. During her four days stay in hospital, the pedal edema decreased significantly. The dyspnea improved to NYHA I with improved in exercise tolerance, as evaluated on the fourth day of admission and before discharge from the hospital. The pericardiectomy was deferred due to patient's decision not to proceed with the procedure, NYHA class II symptoms, shape and location of the calcification, and relatively preserved diastolic function. The patient was advised for regular follow up and the need of pericardiectomy was explained to the patient should the symptoms worsen.

DISCUSSION

In constrictive pericarditis, noncompliant pericardium encases the heart. During inspiration, the increased

venous return cannot be accommodated by right ventricle. Increase in RV volume with expense of LV volume occurs via a shift of the ventricular septum to the left. This decreases the stroke volume of the left ventricle.^{1,2,6} Furthermore, the normal inspiratory decrease in intrathoracic pressure is not transmitted to intracardiac pressures which amplifies inspiratory decreases in pulmonary venous pressure and reduced left-sided inspiratory preload (diastolic failure).^{2,6}

Calcific constrictive pericarditis secondary to tuberculosis results in radiological pericardial calcification seen in 35-50% of patients.² These patients have the widespread calcification, but the localized calcification can be present over the diaphragmatic surface, over the right ventricle, apex and atrioventricular groove, the latter being one of the rarest site.^{2,7,8}

Echocardiography remains the standard diagnostic tool for the constrictive pericarditis with sensitivity of 87% and specificity upto 97%.⁹ In our case, the pathological hemodynamics such as diastolic septal bounce, plethoric and non-collapsible inferior vena cava during inspiration were seen.^{9,10} CT scanning can access the structure, but pathophysiological phenomena cannot be adequately assessed.¹⁰ The CT scan finding of our case demonstrated ring shaped extensive calcification of the pericardium trapping along the atrioventricular groove altering the shape of the heart, resulting into a gourd shaped heart. There is associated thickening of pericardium with maximum thickness measuring approximately 6.0 mm. The above finding contrasts with the common finding of the constrictive pericarditis. The patient was diagnosed with calcified annular constrictive pericarditis caused by constriction due to ring-shaped and localized calcified pericardium with multimodality imaging. Other modalities for the diagnosis and evaluation of the hemodynamics include cardiac MRI and cardiac catheterization.

The recommended treatment in the patients with significant symptoms is the pericardiectomy, which cures the disease as well as improve the symptoms. However, in our case, the patient was managed with oral diuretics and oral anticoagulants for atrial fibrillation. American College of Cardiology and European Society of Cardiology recommends surgery to 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.^{3,5} Furthermore, the surgical approach is not well defined in the cases of localized calcific pericarditis, which encourages further research in the future.⁸

Gourd shaped heart, a variant of chronic constrictive pericarditis, is a localized calcification of the pericardium. The condition is curable by pericardiectomy. Thus this case highlights the importance of the diagnostic measures to detect the disease and hemodynamics associated with the localized calcific constrictive pericarditis.

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