Constrictive Pericarditis with a Calcific Mass Invading into the Right Ventricular Myocardium

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We present a rare and unique case of calcific constrictive pericarditis with a calcified pericardial mass invading the right ventricular myocardium. Perioperative two-dimensional and three-dimensional transesophageal echocardiography revealed the extent and structure of the pericardial mass and led to the repair of the right ventricular free wall as a surgical intervention. (Echocardiography 2013;30:E4-E6)

Key words: pericardiectomy, pericarditis, right ventricular function, right ventricular mass, transesophageal echocardiography, three-dimesional transesophageal echocardiography

A 68-year-old female was admitted to the hospital with increasing oxygen requirement and fluid overload. Medical history included chronic pericarditis, heart failure, paroxysmal atrial fibrillation, restrictive lung disease, and Langerhan's histiocytosis. Chest computed tomography (Brilliance CT-64-channel, Philips Medical Systems, Andover, MA, USA) demonstrated diffuse pericardial thickening with dense calcification predominantly along the atrioventricular grooves accompanied by significant left atrial enlargement. Cardiac magnetic resonance imaging (4.0 Tesla whole-body GE Signa scanner, GE Medical Systems, Milwaukee, WI, USA) revealed overall normal contraction of both left and right ventricles (RV) with normal ejection fraction, as well as diffuse thickening of both the visceral and parietal pericardial layers consistent with constrictive pericarditis. Right and left heart catheterization revealed marked elevation of the mean right atrial pressure of 35 mmHg and equalization of diastolic pressures. A diagnosis of constrictive pericarditis was made and pericardial stripping was scheduled.

Intraoperative two-dimensional (2D) transesophageal echocardiography (TEE) (IE-33, Philips Medical Systems) evaluation revealed bi-atrial enlargement, restrictive diastolic dysfunction, and normal systolic function. The pericardium was thickened and intensely echogenic with a calcified mass overlying the right ventricular free wall and intruding into the RV. (Fig. 1A; and

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movie S1, clip 1). Three-dimensional (3D) TEE was used to further evaluate the mass and revealed that it protruded into the RV as a donut shape structure with a thin apical cap (Fig. 1B; movie S1, clip 2). Cropping of the pericardial mass revealed what appeared to be an echolu-

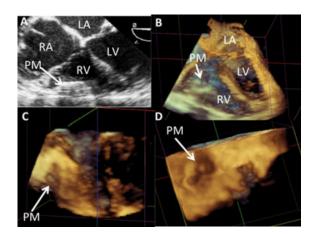


Figure 1. A pericardial mass was present in a patient with constrictive pericarditis. **A.** On a 2-dimensional (2D) transesophageal echocardiography four-chamber view, the pericardial mass is located on the free wall and is seen protruding into the right ventricle. **B.** On a 3D four-chamber view, the pericardial mass is protruding into the right ventricle as a donut shape structure with a thin apical cap. **C.** A three-dimensional (3D) image is focused on the pericardial mass of the right ventricular free wall. After cropping, a cavity can be seen inside the pericardial mass reaching the apical part of the mass. **D.** A 3D image of the pericardial mass from the surface of the right ventricular free wall shows a depression into the myocardium. RA = right atrium; LA = left atrium; RV = right ventricle; LV = left ventricle; PM = pericardial mass.

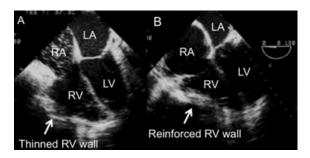


Figure 2. Two-dimensional transesophageal echocardiography four-chamber view of the right ventricular free wall after removal of the pericardial mass (**A**) and post reinforcement with purse-string sutures (**B**). **A.** A thinned free wall can be seen after removal of the pericardial mass. **B.** The right ventricular free wall can be seen after reinforcement with purse-string sutures. RA = right atrium; LA = left atrium; RV = right ventricle; LV = left vent.

cent cavity inside the mass (Fig. 1C; movie S2, clips 1 and 2). Further investigation using multiplanar reconstruction showed that the mass appeared to be a depression from the surface of the right ventricular free wall and also that the underlying right ventricular myocardium appeared to be thinned in this area from the direct protrusion of the mass into the myocardium. (Fig. 1D; movie S2, clip 3).

Surgical dissection of the pericardium above the right ventricular free wall revealed a thickened and calcified pericardial tissue with a cystlike structure. The cystic mass was found to be extended into the right ventricular myocardium. The mass was filled with a white paste-like material. After evacuation of this material, TEE revealed that the residual myocardium was severely thinned (Fig. 2A; movie S3, clip 1). Considering the TEE findings and the concern for aneurysm formation, the decision was made to reinforce the RV free wall. Three horizontal mattress sutures were placed circumferentially to purse-string the cavity. Postrepair TEE examination showed that the free wall thinning had been reinforced (Fig. 2B; movie S3, clip 2). The surgery was completed uneventfully, and the patient was taken to the surgical intensive care unit in a hemodynamically stable condition. Pathology of the pericardial biopsy and the paste-like material demonstrated chronic fibrosing pericarditis with calcifications and no growth from differential culture mediums. The patient had a complicated postoperative course with respiratory failure, significant arrhythmias, and suspected pulmonary embolus. She died on postoperative day 16.

Discussion:

Constrictive pericarditis can occur after any pericardial disease process. Idiopathic and postcardiac surgery constrictive pericarditis being the 2

most frequently occurring etiologies in developed countries. Other causes include connective tissue disorder, malignancy, trauma, medications, asbestosis, sarcoidosis, and uremia. The rate of calcification of the pericardium is between 5% and 36%.¹

Diagnosis of constrictive pericarditis is usually made by clinical examination, echocardiography, or cardiac catheterization.² Once the diagnosis is confirmed, pericardectomy is the established treatment.

Constrictive pericarditis with mass formation is a rare occurrence, with few cases noted in the literature. Mass formation has been reported to be caused by hematoma, calcified hematoma, calcified tissue, tumor, and infection.^{3–10} Pusfilled mass formation by gram positive, gram negative, mycobacterial species, as well as sterile acellular debris containing cholesterol, calcium, and fatty acids, has been reported in various cases. 4-7,10 Very rarely, constrictive pericarditis with mass formation can invade the myocardium. A case of calcific constrictive pericarditis was reported with calcific invasion into the myocardium of the left ventricle, but it did not necessitate surgical reinforcement of the residual myocardium.9 Another case was reported in which a fibro-calcific mass lesion invaded the mitral annulus with infiltration into the base of the posterior mitral leaflet, causing mitral stenosis. Intraoperative management was not aided by echocardiography in this case.8

To our knowledge, this is the first case of calcific constrictive pericarditis involving the myocardium of the right ventricular free wall. The pericardial mass caused structural damage of the myocardium, which was diagnosed by intraoperative TEE and managed by surgical intervention. The initial 2D TEE revealed a pericardial mass of the right ventricular free wall, but the structure of the mass could not be evaluated. Further analysis of the mass using 3D TEE showed a donut shape protrusion in the RV with a thin apical part and exposed a cavity inside the mass extending into the myocardium of the RV (Fig. 1, movies S1 and S2). To our knowledge, this is the first time when a pericardial mass was visualized and characterized with 3D TEE.

This case illustrates the ability of 3D TEE to explore the structure of a pericardial mass. Although the paste-like material was not suspected from the TEE examination, the structure suggested the extension of the pericardial mass into the myocardium.

In this case, the etiology of the constrictive pericarditis was not conclusive because of lack of any characteristic cell types in the pericardial biopsy. The rare combination of Langerhans cell histiocytosis with Erdheim-Chester disease can be considered as a differential diagnosis. The combination of these 2 diseases can manifest in constrictive pericarditis and lung disease as it has been described in the literature but was not histologically proven in our case. ¹⁰

Conclusion:

This is the first intraoperative 2D and 3D TEE analysis of a pericardial mass that had compromised the structural integrity of the right ventricular myocardium. This pericardial mass structure with extension into the myocardium may represent a condition in which further surgical intervention may be indicated after pericardial stripping and removal of the lesion.

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Supporting Information

Additional Supporting Information may be found in the online version of this article:

Movie \$1. Constrictive pericarditis with pericardial mass. Clip 1: Two-dimensional TEE four-chamber view shows a pericardial mass protruded into the right ventricular free wall. Clip 2: On a 3D four-chamber TEE view, a donut shape pericardial mass is protruding into the right ventricle with a thin apical part. RA = right atrium; RV = right ventricle; RV = right ventricle; LA = left atrium; LV = left ventricle; PM = ericardial mass.

Movie S2. Constrictive pericarditis with pericardial mass. Clip 1: A 3D four-chamber view is focused on the pericardial mass of the right ventricular free wall. After cropping, a cavity can be seen inside the pericardial mass. Clip 2: A 3D four-chamber TEE view focused on the pericardial mass, which was rotated and cropped to show the extension of the cavity. The cavity extended into the apical part of the pericardial mass corresponding with the thin part of the protruded mass in the right ventricle. Clip 3: A 3D moving image of the pericardial mass from the surface of the right ventricular free wall shows a depression into the myocardium. RV = right ventricle; PM = pericardial mass.

Movie S3. Right ventricular free wall after removal of the pericardial mass. Clip 1: A 2D-TEE four-chamber view shows thinned right ventricular free wall after removal of the pericardial cyst. Clip 2: A 2D-TEE four-chamber view demonstrates the right ventricular free wall after reinforcement with purse-string sutures. RV = right ventricle; LV = left ventricle.