“An imprisoned heart” arrested by pillory of calcified ring surrounding both ventricles: An extremely rare case of constrictive pericarditis

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“An imprisoned heart” arrested by pillory of calcified ring surrounding both ventricles: An extremely rare case of constrictive pericarditis

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Keywords

localized constrictive pericarditis, heart failure, pericardial calcification, calcified ring

Abstract

We report the case of a 63-year-old man who was admitted to our hospital due to progressive dyspnea and leg edema. Echocardiography and cardiac computed tomography revealed localized severe pericardial calcification surrounding the basal segments of both ventricles, which caused pericardial constriction. Right heart catheter examination confirmed typical hemodynamic findings of constrictive pericarditis, and subsequently led to the definite diagnosis of constrictive pericarditis. We experienced a rare case of localized constrictive pericarditis caused by the thickened and calcified pericardial ring. Non-invasive and invasive multimodality evaluations should be encouraged for accurate diagnosis and better management of these cases.

Introduction

Constrictive pericarditis (CP) usually develops after an episode of acute pericarditis, cardiac surgery, radiotherapy, or collagen diseases[1]. In most CP cases, stiffness of the pericardium uniformly restricts the filling of the blood to the heart chambers. However, in a few cases, localized pericardial thickening can result in CP. Here, we report a rare case of a patient with localized CP whose circular pericardial calcification strangled the basal segments of both ventricles, which eventually led to typical CP hemodynamics.

Case report

A 63-year-old man who had been treated for refractory pleural effusion and liver cirrhosis of unknown origin was admitted to our hospital due to progressive dyspnea and systemic edema. Leg edema with stasis dermatitis and abdominal distention were observed. The jugular vein was distended with a prominent *y* descent (Movie clip 1), suggesting the presence of pericardial constriction. Furthermore, a widely split S_2 with loud P_2 and pericardial knock were observed upon auscultation. Transthoracic echocardiography revealed the localized cardiac constriction by the thickened and calcified pericardial ring; thus, the atrioventricular groove was peculiarly distorted（Figure 1, Movie clip 2-3）. Multi-detector row computed tomography clearly showed global pericardial thickening and a calcified pericardial ring surrounding the basal segments of both ventricles（Figure 2）. Eventually, the basal segment of the heart was tightly strangled, and the heart was shaped like a gourd

(Movie clip 4). Surprisingly, the left circumflex artery was also strangled by the calcified pericardial ring, which resulted in luminal narrowing. Cardiac catheter examination revealed the characteristic hemodynamics of CP which included a prominent *y* descent in the right atrial pressure, dip and plateau pattern in the right ventricular waveform (Figure 3), equalization of diastolic pressure of all cardiac chambers, and respiratory discordance between the right ventricular and left ventricular pressures. Based on these invasive and non-invasive multimodality assessments, the patient was diagnosed with localized CP resulting from the constriction by the band-shaped, thickened, and calcified pericardium.

The detailed history taking revealed that he had been suffering from arthralgia for about one year, moreover, he showed persistent fever after the admission. Based on the medical backgrounds and the results of immunologic examinations, he was eventually diagnosed with systemic lupus erythematosus. He was then treated with 1 mg/kg of prednisolone. After the decrease in the disease activity, he will undergo pericardiectomy.

Discussion

The normal pericardium can stretch to accommodate physiologic changes in cardiac volume. On the other hand, the pericardium becomes uniformly thickened, scarred and inelastic in CP. Hence, the thickened pericardium prevents transmission of the normal inspiratory decrease in intrathoracic pressure to the heart chambers. Thus, pulmonary venous pressure, but not left atrial pressure, decreases during inspiration, leading to a reduction in

left ventricular volume because of a decrease in the driving pressure from the pulmonary veins to the left heart. Moreover, the abnormal pericardium does not simultaneously expand to accommodate increased venous return to the right heart during inspiration. Due to the ventricular interaction, the right ventricular volume expands via shifting of the interventricular septum.

Only a few cases of localized pericardial constriction were previously reported[4]. Patients do not exhibit the typical hemodynamics of CP and sometimes mimic other cardiac conditions such as valvular stenosis[6] or cardiomyopathy[7] depending on the distribution of its adhesion and size. Therefore, it is actually difficult to diagnose localized CP in such cases. This case, however, demonstrated the specific findings consistent with typical CP by the cardiac catheter examination. It was speculated that although the thickened and calcified pericardial ring surrounded only the basal segments of both ventricles, the rest of the heart was also constricted by the thickened pericardium without calcification. A multimodality imaging approach is very helpful to diagnose various forms of pericardial constriction[5]. This case highlights the important role of a combination of assessments using both invasive and non-invasive modalities in such atypical cases of CP.

Conclusion

Herein we report a quite rare case of a patient with localized CP whose calcified pericardial ring strangled the basal segments of both ventricles. We need to recognize that

there is a unique phenotype of CP similar to this case. The possibility of this type of CP should be considered as a differential diagnosis for early diagnosis and better management, especially in cases with refractory right-sided heart failure.

Conflict of interest: The authors declare no conflict of interest.

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Figure legends

Figure 1: (A) Parasternal long axis view showing severe pericardial carcification of the basal segment of the left ventricle (red arrows). (B) Apical 4-chamber view revealing the presence of a thickened and calcified pericardial ring surrounding the basal segments of both ventricles: thus, the atrioventricular groove is peculiarly distorted (red arrows).

LV=left ventricle; LA=left atrium; RV= right ventricle; RA= right atrium.

Figure 2: (A) Three-dimensional volume-rendering image showing the thickened and calcified pericardial ring (red arrows) that surrounds the basal segments of both ventricles. (B) Three-dimensional cardiac CT image from the right anterior oblique view demonstrating a calcified pericardial band (red arrows) strangling both ventricles.

LV=left ventricle; RV= right ventricle; PA= pulmonary artery; Ao= Aorta.

Figure 3: Right atrial (RA) and right ventricular pressure waveforms are shown. A prominent *y* descent of the RA waveform (red arrowheads) and the typical dip and plateau wave (red arrows) are observed.

ECG = electrocardiogram; RA = right atrial; RV= right ventricular.

Movie clips

Movie clip 1: Jugular venous distention with prominent *y* descent (white arrows) suggests the presence of pericardial constriction.

Movie clip 2: Parasternal long-axis view shows severe pericardial calcification (red arrows) of the basal segment of the left ventricle.

LV=left ventricle; LA=left atrium.

Movie clip 3: Apical 4-chamber view shows the thickened and calcified pericardial ring (red arrows) surrounding the basal segments of both ventricles, resulting in distortion of the cardiac chambers.

LV=left ventricle; LA=left atrium; RV= right ventricle; RA= right atrium.

Movie clip 4: Cardiac four-dimensional CT movie from the right anterior oblique view demonstrates the calcified pericardial band (red arrows) strangling the both ventricles.

LV=left ventricle.

Figure 1

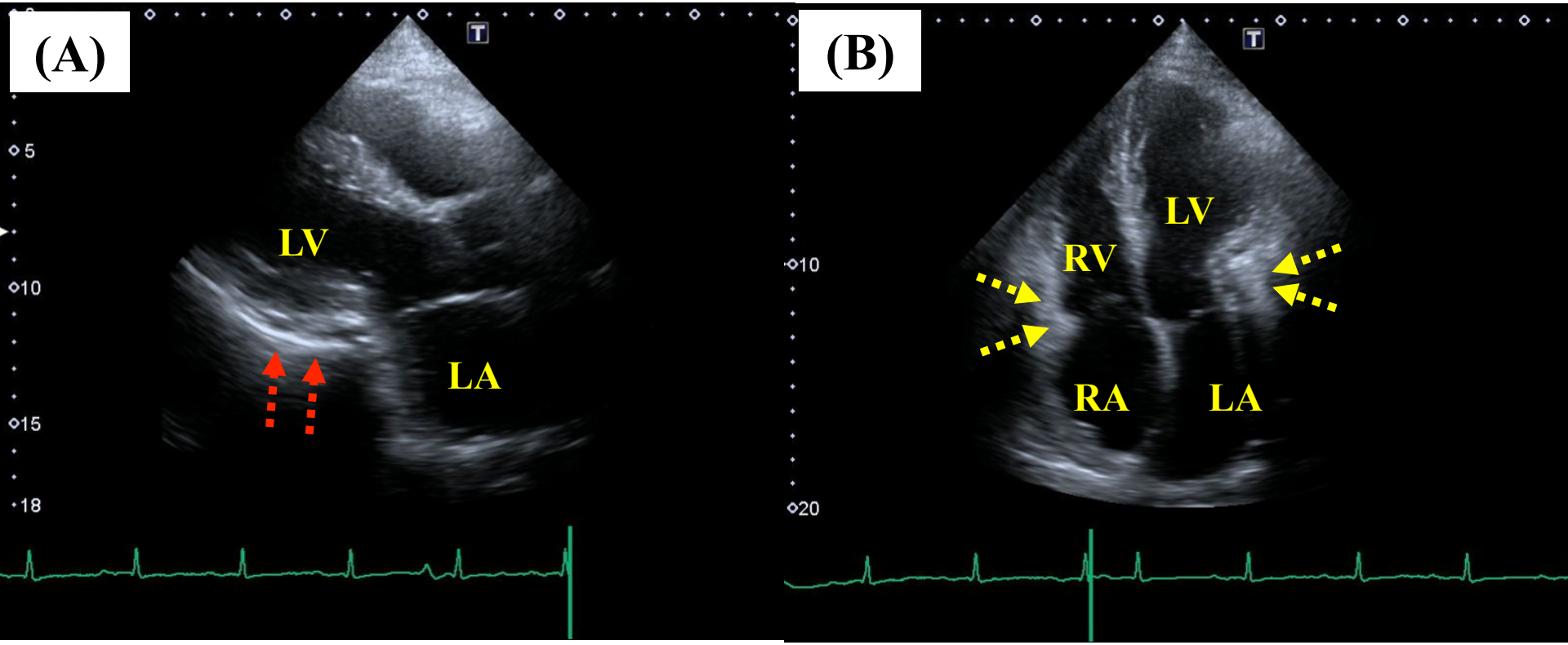
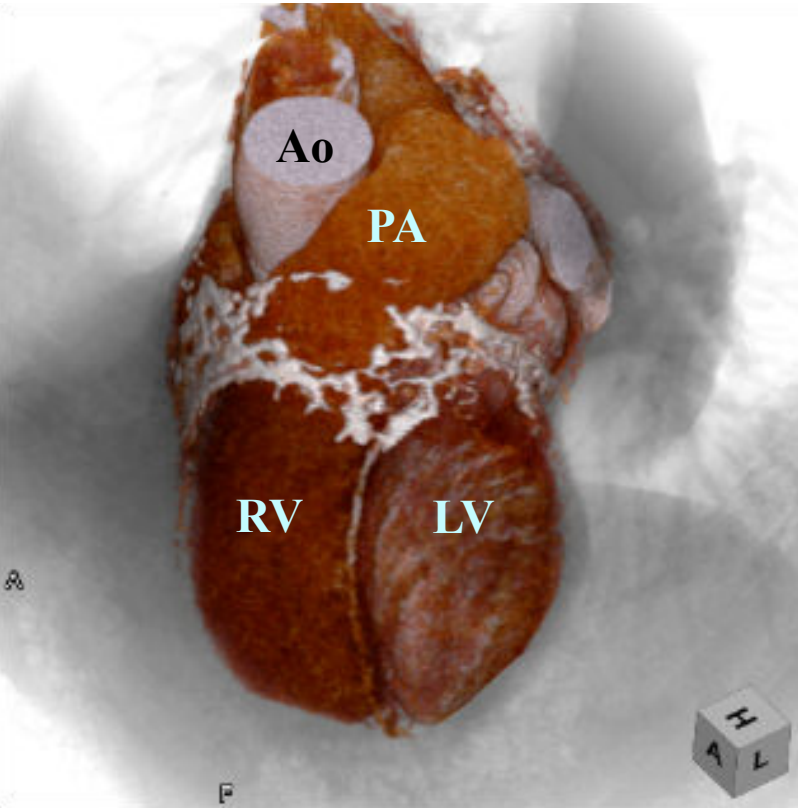


Figure 2

(A)



(B)

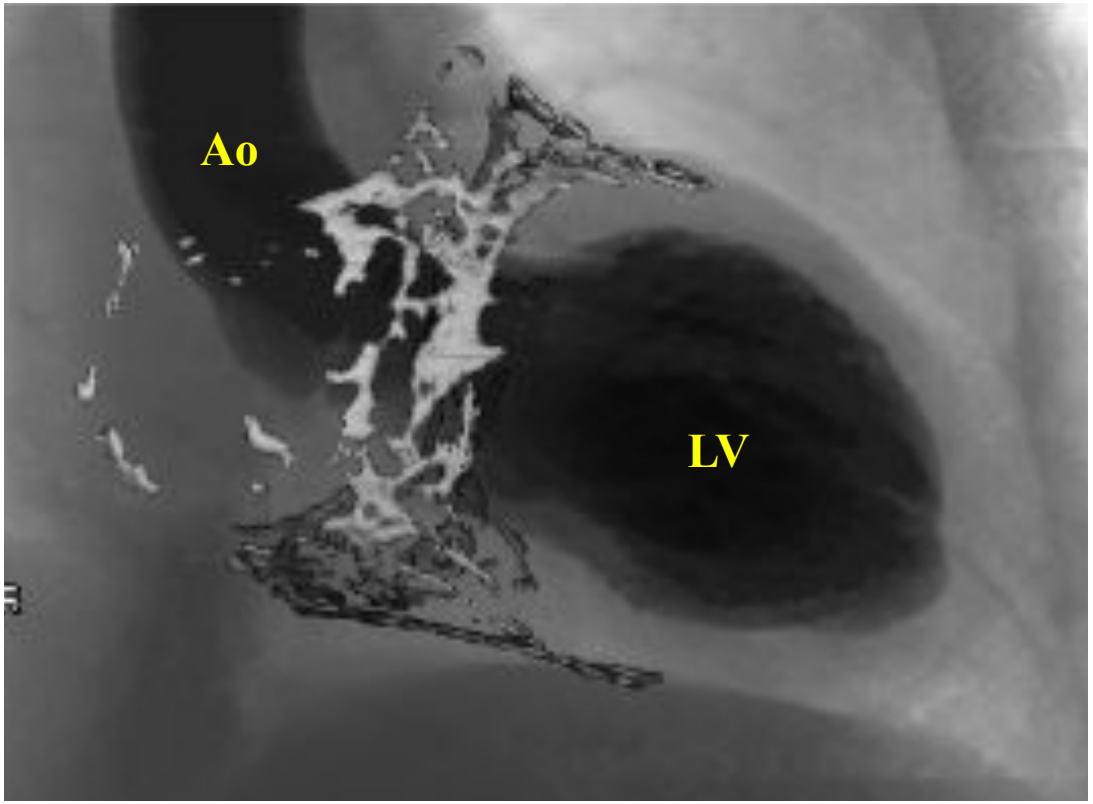


Figure 3