ESC European Society of Cardiology

European Heart Journal - Case Reports (2022) **6**, 1–5
https://doi.org/10.1093/ehjcr/ytac231

CLASSICAL CASE REPORT
Heart failure

Heart in a shell—a cascade of classical findings: a case report

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Received 24 December 2021; first decision 18 January 2022; accepted 6 June 2022; online publish-ahead-of-print 10 June 2022

Background Idiopathic chronic constrictive pericarditis (CP) is an uncommon yet very important clinical entity as prompt diagnosis and early treatment are affiliated with improved outcomes. We describe an uncommon case of CP with a series of textbook findings and received successful treatment with surgical pericardiectomy (SP).

Case summary A 58-year-old male presented to the emergency department with exertional dyspnoea and anasarca. The past medical history was negative for infections like tuberculosis, viral, etc., connective tissue disease, prior cardiac surgery, human immunodeficiency virus, or any radiation therapy. The vital signs were within normal limits except mild hypoxia while the physical examination was consistent with congestive heart failure. Further investigations with laboratory testing, imaging with chest X-ray, computed tomography, echocardiogram, and invasive study with right heart catheterization were all consistent with idiopathic chronic CP. The patient underwent successful SP with significant improvement in clinical condition.

Discussion This case highlights the classical signs and symptoms along with important diagnostic features of CP. It is uncommon to see all the classical features of CP in one patient as described in the above case. Familiarity with these findings is crucial to make the diagnosis as early treatment is affiliated with improved outcomes.

Keywords Constrictive pericarditis • Heart failure • Cardiomyopathy • Pericardiectomy • Classical case report

ESC Curriculum 2.2 Echocardiography • 6.1 Symptoms and signs of heart failure • 6.5 Cardiomyopathy • 6.6 Pericardial disease • 2.1 Imaging modalities

Learning points

- Idiopathic constrictive pericarditis is a unique clinical condition associated with delayed treatment due to high latency between symptoms onset and diagnosis.
- The dissociation between intracardiac and intrathoracic pressures and increased interventricular dependence due to fixed pericardial space are the main pathophysiological mechanisms resulting in heart failure.
- Supportive treatment with diuretics and definitive therapy with surgical pericardiectomy are the mainstay of treatment.

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Handling Editor: Tom De Potter
Peer-reviewers: Rita Pavasini; Edoardo Conte
Compliance Editor: Brett Sydney Bernstein
Supplementary Material Editor: Mariame Chakir

Introduction

Constrictive pericarditis (CP) is of great clinical interest due to unique pathophysiology, hemodynamics, and significant recovery after treatment. Some reports of transient and subacute verities have been described in the literature, however, it is normally chronic and develops over months and years.[1] The exact prevalence is unknown with less than 0.5% of the patients with acute pericarditis ultimately develop CP.[2]

Tuberculosis remains the most common cause of CP worldwide up to 48% of the cases but the three most common aetiologies in North America and Europe are idiopathic, prior cardiac surgery, and radiation therapy.[3] Other aetiologies include post-viral infection, human immunodeficiency virus (HIV), and connective tissue diseases (CTD).[3]

We present a rare case of idiopathic CP with a cascade of classical findings who underwent successful surgical pericardiectomy (SP).

Timeline

Date	Event
1 year before admission	Progressive exertional dyspnoea
4 months before admission	Progressive anasarca
Day 1 admission	Presented with congestive heart failure symptoms Physical examination showed raised JVP, pulmonary rales, and anasarca The chest X-ray revealed fluid overload and calcified pericardium Started on supported treatment with diuresis
Day 3 admission	CT chest revealed severely thickened pericardium Echocardiogram revealed classic signs of constrictive pericarditis
Day 5 admission	Right heart catheterization revealed prominent '*x*' and '*y*' descent along with interventricular dependence
Day 14 admission	SP was performed
Day 19 admission	Discharge home

Case presentation

A 58-year-old Caucasian male presented to the emergency department with worsening exertional dyspnoea for 1 year and generalized body swelling for 4 months. The swelling started from lower extremities and slowly became anasarca. The review of systems was pertinent for fatigue, weakness, orthopnea, and paroxysmal nocturnal dyspnoea and negative for chest pain, cough, sputum, or syncope.

The previous history was negative for any co-morbidities or home medications. The social history was negative for nicotine or substance use disorders and positive for alcohol use disorder with eight beers a day for 30 years. The patient was living alone and working at a construction company, however, denied exposure to particles like asbestos etc.

Upon presentation, the vital signs revealed blood pressure of 123/76 mmHg, heart rate of 94 beats per minute, respiratory rate of 24 per minute, and oxygen saturation of 90% while the patient was breathing ambient air. The physical examination showed raised jugular venous pressure (JVP), normal S1 and S2, and generalized anasarca. The pulmonary examination revealed decreased breath sounds and bilateral crackles. The focused exam for CTD including lymphadenopathy, rash, ulcers, hair loss, joint swelling, etc. was negative.

In a patient with new-onset congestive heart failure (HF), the differential diagnosis of the acute coronary syndrome, cardiomyopathies including but not limited to alcoholic, constrictive vs. restrictive, thyrotoxicosis, hypertension, and valvular heart disease should be considered.

The laboratory data were within normal limits except for elevated brain natriuretic peptide levels of 274 pg/mL (reference range $<$100 pg/mL). The electrocardiogram was unremarkable while the chest radiography (CXR) revealed increased cardiac silhouette, pulmonary vascular congestion, and bilateral pleural effusions. The CXR also showed a bright layer surrounding the heart, suggestive of calcified pericardium (*Figure 1*). The computerized tomography confirmed the presence of calcified pericardium with thickness up to 23 mm (*Figure 2*).

The patient was started on oxygen supplementation and diuretic therapy with partial improvement of the symptoms. Further investigation with a transthoracic echocardiogram (TTE) revealed classical features of CP. The ejection fraction was normal along with bi-atrial enlargement (see Supplementary material online, *Figure S1*). The parasternal long-axis M-mode revealed rapid expansion and sudden diastolic flattening of the inferolateral wall due to thickened pericardium outside and septal bounce secondary to interventricular dependence (IVD) (*Figure 3A*). There was increased respiratory atrioventricular valve inflow variation due to intracardiac and intrathoracic pressure dissociations and IVD (*Figure 3B*). The inferior vena cava was dilated along with diastolic expiratory hepatic

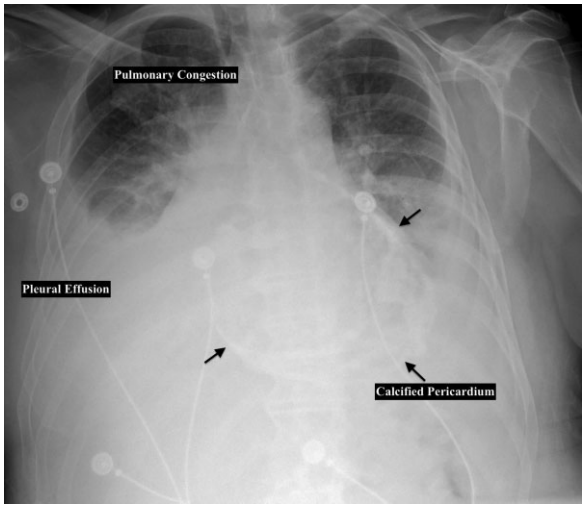

Figure 1 Chest radiography showing calcified pericardium, pulmonary congestion, and pleural effusions.

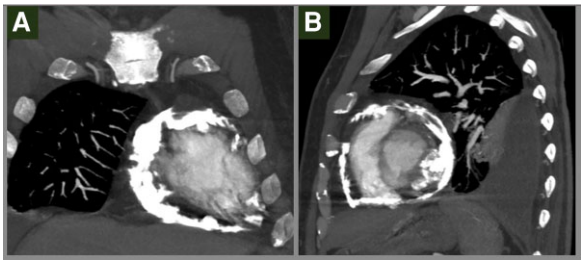

Figure 2 Computed tomography scan showing thickened pericardium (Arrows).

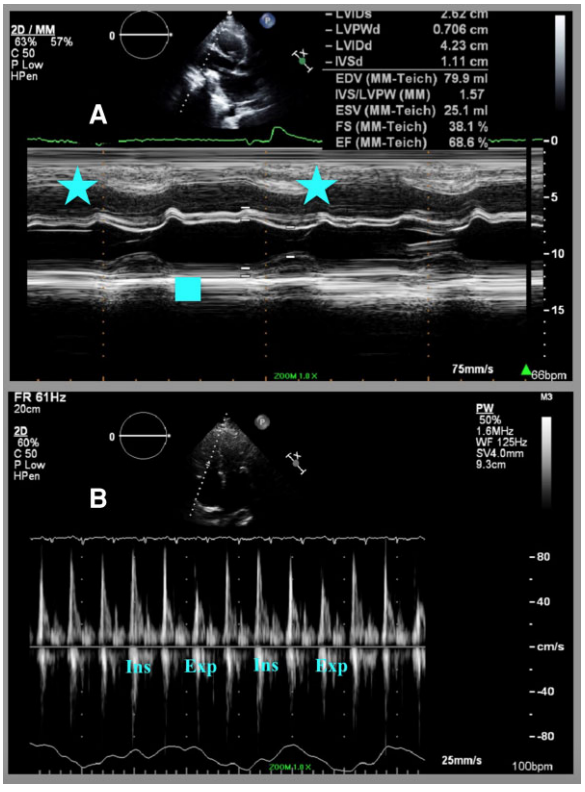

Figure 3 (*A*) Transthoracic echocardiogram M-mode showing diastolic inferolateral wall flattening and septal bounce. (*B*) Transthoracic echocardiogram Doppler showing atrioventricular valve respiratory variation.

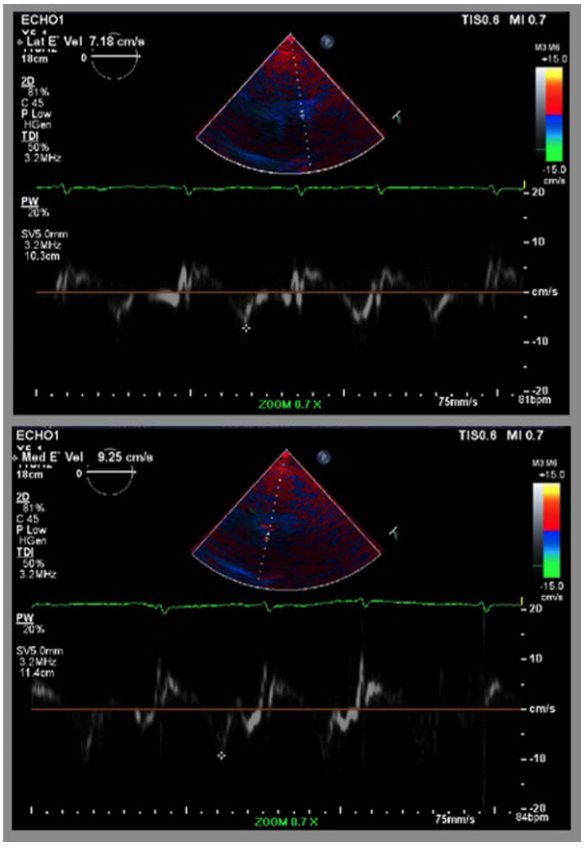

Figure 4 Transthoracic echocardiogram tissue Doppler showing medial and lateral annular velocities.

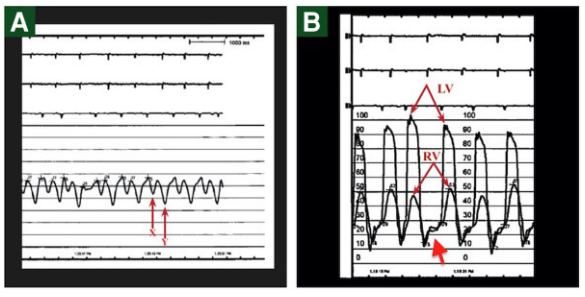

Figure 5 (*A* and *B*) Right atrial and simultaneous left and right ventricular pressure tracings.

Doppler flow reversal due to the right sided septal bounce (see Supplementary material online, *Figure S2*).

Upon tissue Doppler, the septal e' was more than 8 cm/s and more than the lateral e' consistent with annulus reversus (*Figure 4*). On invasive evaluation with right heart catheterization, the right atrial pressure tracing showed Friedrich's sign with a prominent '*y*' descent due to rapid diastolic emptying along with rapid '*x*' descent (*Figure 5A*). Simultaneous left and right ventricular pressure tracings revealed the 'Dip and plateau' or 'square root' sign (*Figure 5B*) characteristic for rapid early diastolic dip and equalization of diastolic pressures, along with IVD (*Figure 5B*). A thick shell of calcified pericardium with restricted heart motion was evident on fluoroscopy (see Supplementary material online, *Video S1*).

The case was discussed with cardiothoracic surgery and the plan was made to proceed with pericardiectomy. The patient underwent successful procedure with removal of the calcified

pericardium which allowed a significant distensibility of the heart (see Supplementary material online, *Videos S2–S3*). The patient had an uneventful recovery and significant improvement in the symptoms. The patient was discharged home on furosemide with close follow-up. The patient's symptoms were significantly improved at one month follow-up visit.

Discussion

Our patient was unique with no prior history that could point out a possible aetiology. Also, the work-up including C-reactive protein, sedimentation rate, HIV, tuberculosis, and CTD was negative. The pathophysiology of CP largely depends upon the dissociation between intracardiac and intrathoracic pressures and increased IVD due to the fixed pericardial space[4] resulting in septal bounce, respiratory atrioventricular flow variation, and expiratory diastolic hepatic flow reversal.

Heart failure is the most common presentation in these patients including exertional dyspnoea, raised JVP, Kussmaul sign, abdominal distension, and lower extremity oedema.[5] A careful physical examination can differentiate CP from restrictive pathophysiology as there are prominent *x* and *y* descent on jugular examination in CP due to enhanced annular motion and early ventricular filling while there is blunting of the *x* descent in restrictive cardiomyopathy due to atrial myopathy.[6]

The diagnostic modalities with Class I recommendation are echocardiography, cardiac computed tomography, and magnetic resonance imaging (MRI), and invasive evaluation with cardiac catheterization.[6,7] Transthoracic echocardiogram as an initial step has a combined sensitivity and specificity up to 87 and 91%, respectively, in the presence of ventricular septal shift with either hepatic vein expiratory diastolic flow reversal ratio $\geq$0.79 or medial e' $\geq$9 cm/s.[4] The radiological studies provide better assessment of the thickness and extension of calcified pericardium while invasive evaluation gives deeper look into pathophysiology.[6,7] Lately, MRI has become popular because of its additional benefit to detect inflammation. Moreover, variation in the pericardial late gadolinium enhancement can be used to follow the anti-inflammatory treatment response.[8]

The gold standard treatment is SP.[7] The most common indication is idiopathic CP with HF, resistant to medical therapy. The patients should be carefully screened for conditions like restrictive cardiomyopathy, active pericardial inflammation, chronic steroid use, liver failure, right ventricular dysfunction, and moderate to severe tricuspid regurgitation as presence of these are associated with adverse outcomes.[9,10] Our patient did not have any of these with MELD (Model for End-Stage Liver Disease) score of only seven indicating low-risk profile. SP can be either partial or complete. The historical approach is partial anterior phrenic to phrenic nerve resection through lateral thoracotomy, however, complete resection through median sternotomy has become more popular as it provides better outcomes.[9,10]

The medical therapy can be helpful in two scenarios; first, anti-inflammatory drugs can be given if there is active inflammation.[7,11] Second, diuretic therapy to relieve congestion, however, surgery should not be delayed to avoid poor prognosis and increased mortality.[12]

Conclusion

This case is unique as it has cascade of classical findings of CP. It is rare to see all the findings in every case which makes the diagnosis of CP very challenging. This case provides an overview of the possible aetiologies and pathophysiological mechanisms leading to diagnostic findings. Early diagnosis and prompt treatment are affiliated with improved outcomes.

Lead author biography

Hafiz Muhammad Waqas Khan, MD, is a cardiovascular disease fellow at Michigan State University/McLaren. He earned his Bachelor of Medicine and Surgery degree from Kind Edward Medical University, Pakistan. After completion of medical school, he pursued his career in USA and joined cardiovascular disease fellowship program after completing Internal Medicine residency. He is further pursuing his career in Interventional Cardiology and plan to subspecialize in Structural Heart Disease in the future.

Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports* online.

Acknowledgements

I would like to thank everyone who was involved in taking care of this patient.

Slide sets: A fully edited slide set detailing these cases and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for submission and publication of this case report including imaging and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: None declared.

Funding: None declared.

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