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CASE REPORT
Heart failure

A case report of tuberculous constrictive pericarditis necessitating total pericardiectomy

Nooraldaem Yousif [1], Abdulla Alnuwakhtha[1], Abdulla Darwish[2], Zaid Arekat[1], and Seham Abdulrahman[1]*

[1]Mohammed Bin Khalifa Bin Sulman Al Khalifa Specialist Cardiac Centre (MKCC), Awali, Kingdom of Bahrain; [2]Department of Pathology, Royal Medical Services, Bahrain Defense Force Hospital, Riffa, Kingdom of Bahrain

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Background Constrictive pericarditis (CP) is one of the most serious sequelae of tuberculous pericarditis, which is characterized by heart constriction secondary to intense pericardial inflammation and thickening. Several invasive and non-invasive diagnostic modalities are crucial to address the challenges of confirming the diagnosis of CP and to expedite timely intervention.

Case summary This study reports the case of a Bahraini male with tuberculous lymphadenitis diagnosed with CP as a result of various evaluations. The patient underwent urgent total pericardiectomy and showed remarkable recovery with complete resolution of heart failure symptoms.

Discussion This case demonstrates the paramount importance of early diagnosis and treatment for patients with CP. In this unique case, the acoustic windows on echocardiography were suboptimal because of pericardial thickening. Further, computed tomography did not show significant calcification of the thickened pericardium. A novel approach of assessing haemodynamics through the right antecubital vein and right radial artery facilitated the accurate diagnosis of CP with confidence. Thereafter, successful pericardiectomy revealed a markedly thickened and stiff pericardium with many abscesses and dense adhesions encasing the heart, and pericardial biopsy showed large caseating granulomas. This case exemplifies the difficulty in diagnosing CP and the favourable outcomes achieved with well-timed surgical intervention.

Keywords Tuberculosis • Constrictive pericarditis • Right heart study • Swan-ganz catheter • Antecubital vein • Pericardiectomy • Case report

Learning points

- Constrictive pericarditis (CP) is an uncommon pathology that mimics various other diseases, which typically results in delayed or missed diagnosis of this peculiar condition.
- Care from a multidisciplinary team with multiple diagnostic modalities, both non-invasive and invasive, is imperative for distinguishing CP from other causes of heart failure.
- The mainstay and only definitive treatment for congestive heart failure secondary to CP is pericardiectomy.

* Corresponding author. Tel: 00973 17889934; Email: seham.mkcc@gmail.com
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Introduction

Bahrain is a country with a low incidence rate of mycobacterial tuberculosis (TB) infection (11 per 100 000); most TB cases (82%) were non-Bahraini, 68% were males, and 60% had pulmonary disease.[1] Pericarditis is a rare complication that occurs approximately in 12% of TB cases.[2] The most common pericardial manifestation is constrictive pericarditis (CP), which occurs in approximately 30% of patients.[2,3] Generally, CP is an uncommon disease that mimics other pathologies and is often unsuspected as an underlying mechanism of congestive heart failure, in which a fibrous thickening of the pericardium hinders the normal diastolic filling of the ventricles. This reduces venous return and cardiac output and eventually produces the so-called 'single diastolic chamber' that manifests clinical features of left and right heart failure.[4,5]

Here, we present a case of a young Bahraini male who has TB lymphadenitis and was diagnosed to have CP through various invasive and non-invasive investigations necessitating urgent total pericardiectomy with the dramatic improvement of his symptoms thereafter.

Case presentation

A 32-year-old Bahraini male presented with a 2-week history of exertional dyspnoea (New York Heart Association functional Class III) associated with orthopnoea, abdominal distension, and lower limb swelling in the emergency department. His past medical history was significant for TB lymphadenitis, which was diagnosed a few months ago via mediastinal lymph node biopsy. During that time, he was on appropriate anti-TB therapy as per the guidelines with isoniazid 300 mg o.d., rifampicin 600 mg o.d., ethambutol 1 g o.d., and pyrazinamide 1.5 g o.d. In addition, he was started on furosemide 40 mg b.i.d. and aldactone 25 mg o.d., 1 week prior to the current presentation.

On examination, he was afebrile with a pulse rate of 98/min, respiratory rate of 18/min, and blood pressure of 98/60 mmHg. Physical examination revealed raised jugular venous pressure 4 cm above the sternal angle that increased with inspiration. The chest examination showed bibasal diminished air entry with minimal crackles. The cardiac examination revealed soft first and second heart sounds with no murmurs. The abdomen was distended with full flanks. There was bilateral lower extremity oedema to the knees.

Timeline

Three montds before admission	Diagnosed with tuberculosis (TB) lymphadenitis and kept on appropriate anti-TB therapy with isoniazid 300 mg o.d., rifampicin 600 mg o.d., ethambutol 1 g o.d., and pyrazinamide 1.5 g o.d.
Two weeks before admission	Exertional dyspnoea (New York Heart Association Class III) associated with orthopnoea, abdominal distension, and lower limb swelling
One week before admission	Started on furosemide 40 mg b.i.d. and aldactone 25 mg o.d. for unexplained right sided heart failure
Day 1	Admitted for heart failure work up and treatment. Electrocardiogram demonstrated low-voltage QRS complexes and sinus tachycardia. Echocardiogram showed preserved biventricular function, without any significant valvular disease. The pericardium was thick, and there was evidence of septal bounce. Tissue Doppler imaging showed annulus reversus. The inferior vena cava was dilated with no respiratory variations. Expiratory diastolic flow reversal was observed in the hepatic vein. The systolic pulmonary artery pressure was 40 mmHg.
Day 2	Computed tomography scan of the chest showed markedly thickened pericardium and mild bilateral pleural effusion. No evidence of significant pericardial calcification.
Day 3	He was able to lie flat and hence left–right heart catheterization was performed via right radial artery and right antecubital vein which confirmed constrictive pericarditis as underlying mechanism of congestive heart failure. • Mean right atrial (RA) pressure was elevated (30 mmHg) with prominent X and Y descents result in classic 'M' or 'W' pattern (Friedrich's sign). No significant variation in RA pressures during respiration, • Both right ventricular systolic pressure (RVSP) and right ventricular end-diastolic pressure (RVEDP) were elevated at 46 and 31 mmHg, respectively, with RVEDP-to-RVSP ratio of >1/3 (RVEDP/RVSP=0.67). • Simultaneous left ventricular–right ventricular (LV–RV) pressure tracings revealed equalization of LV and RV diastolic pressures with typical dip and plateau waveform (square root sign) and ventricular discordance (exaggerated ventricular interdependence) • Pulmonary artery systolic pressure was 30 mmHg • Coronary angiography showed normal epicardial coronary arteries.
Day 10	Uncomplicated total pericardiectomy
Day 20	Dramatic improvement of his symptoms and complete resolution of anasarca. He had significant 12 kg weight loss during his hospital stay and was discharged off all diuretics. Medications at discharge include the continuation phase of anti-TB therapy (isoniazid 300 mg o.d. and rifampicin 600 mg o.d.) along with pyridoxine 20 mg o.d. and vitamin C 1 g b.i.d. Additionally, paracetamol 1 g every 8 h and Tramadol 50 mg every 12 h (for 4 days) were prescribed to relief post-surgical pain.
One month after discharge	Follow-up via phone consultation (due to hospital COVID-19 related protocols), he reported doing very well, asymptomatic with no residual heart failure symptoms.
3 months after discharge	He remained asymptomatic with good functional status and normal exercise capacity. Follow-up echo showed normal LV systolic function, indeterminate diastolic dysfunction with no evidence of constrictive pattern in Doppler signals.

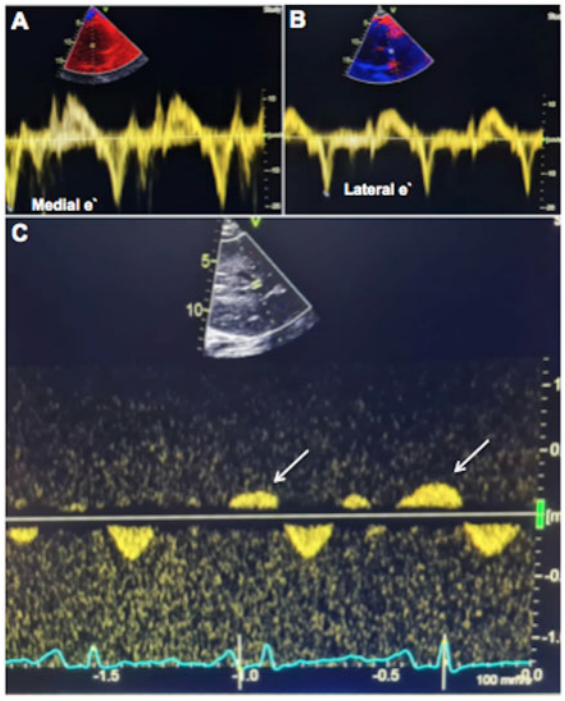

Figure 1 (*A and B*) Tissue Doppler imaging velocity of medial *E′* (0.19 m/s) is greater than that of lateral *E′* (0.14 m/s) indicating annulus reversus. (*C*) Hepatic vein expiratory diastolic flow reversal (white arrows).

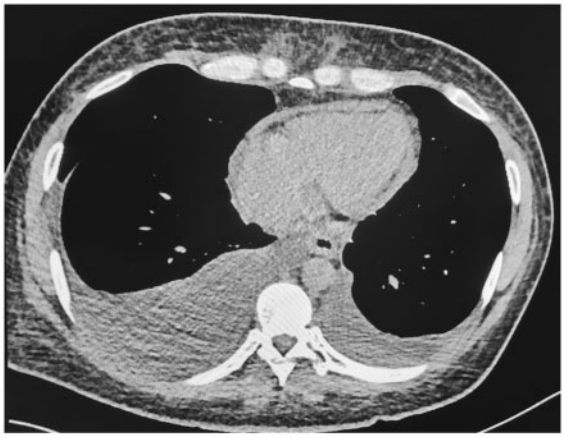

Figure 2 Computed tomography scan showing markedly thickened pericardium and mild bilateral pleural effusion.

Besides elevated erythrocyte sedimentation rate and C-reactive protein, laboratory investigations were normal. Electrocardiogram demonstrated low-voltage QRS complexes and sinus tachycardia. Echocardiogram (poor acoustic window) showed preserved biventricular function, without any significant valvular disease. The pericardium was thick, and there was evidence of septal bounce. There was a normal *E*-to-*A* ratio and a normal *E*-to-*E′* ratio. Tissue Doppler imaging velocity of medial *E′* (0.19 m/s) was greater than that of lateral *E′* (0.14 m/s) indicating annulus reversus (*Figure 1A,B*). There was no significant change in mitral or tricuspid inflow during respiration. The inferior vena cava was dilated with no respiratory variation, and the systolic pulmonary artery pressure was 40 mmHg. Finally, an expiratory diastolic flow reversal was observed in the hepatic vein (*Figure 1C*).

Computed tomography (CT) of the Chest showed markedly thickened pericardium and mild bilateral pleural effusion. No evidence of significant pericardial calcification or effusion was noted on CT (*Figure 2*).

Based on these findings, invasive haemodynamics was performed to confirm the diagnosis of CP and demonstrate the pathological impact of this disease. A right heart study was conducted via the right antecubital vein; simultaneous access of the right radial artery was gained for the left heart study and subsequent coronary angiography. Mean right atrial (RA) pressure was elevated (30 mmHg) with prominent X and Y descents (*Figure 3*). The combination of elevated mean pressure, inconspicuous positive waves, and prominent descents result in the classic 'M' or 'W' pattern (Friedrich's sign). Additionally, there was no significant variation in RA pressures during respiration, which indicates dissociation between respiratory and intracardiac pressures. Both right ventricular systolic pressure (RVSP) and right ventricular end-diastolic pressure (RVEDP) were elevated at 46 and 31 mmHg, respectively, with an RVEDP-to-RVSP ratio of >1/3 (RVEDP/RVSP = 0.67).

Simultaneous LV–RV pressure tracings were recorded to document the equalization of LV and RV diastolic pressures (*Figure 4A*), and the tracings showed a difference of <5 mmHg (left ventricular end-diastolic pressure (LVEDP) - RVEDP = 2 mmHg) with typical dip and plateau waveform (square root sign). This waveform is characteristic of CP and represents early rapid LV diastolic filling that stops abruptly owing to a stiff pericardium that limits the amount of ventricular filling. Accentuated filling of the ventricles during early diastole is represented by deep left ventricular rapid flow wave similar to mitral annular E′ wave on tissue Doppler imaging. Left ventricular rapid flow wave measuring of >7 mmHg (as in our case) has a sensitivity of 93% in diagnosing CP (*Figure 4B*).

Furthermore, simultaneous right and left ventricular (RV and LV) pressure tracings showed ventricular discordance (i.e. during inspiration, peak systolic pressure in LV is reduced with a corresponding increase in RV pressure, and the opposite occurred during expiration), which confirmed that the filling of one ventricle is out of phase to the other, which is a distinctive feature of CP (*Figure 4C*). Moreover, in CP, the pulmonary artery systolic pressure (PASP) is generally limited to <50 mmHg, and in our patient, the PASP was 30 mmHg. Finally, coronary angiography showed normal epicardial coronary arteries. The main echocardiographic and invasive haemodynamic differences between CP and restrictive cardiomyopathy are depicted in *Table 1*.

Based on the history and significant haemodynamics findings and following a multidisciplinary discussion, he underwent a total pericardiectomy. Upon gross inspection, the pericardium appeared diffusely thickened and stiff with plenty of abscesses cavities (*Figure 5A,B*). Total pericardiectomy and debridement were performed successfully, and tissue specimens were sent for histological examination that confirmed the presence of necrotizing granulomas (*Figure 5C,D*), which coincided with the overall clinical picture of tuberculous CP.

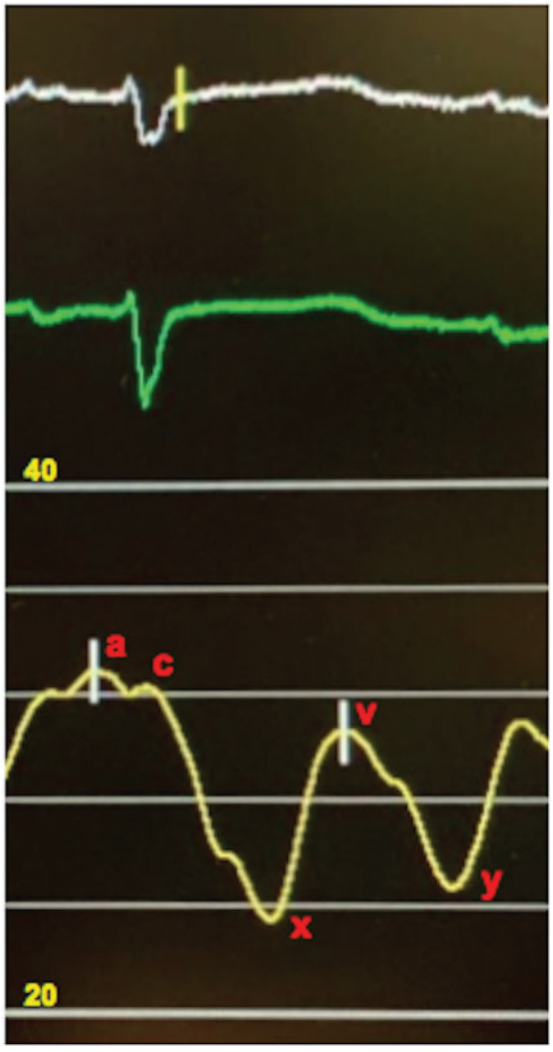

Figure 3 Right atrial pressure trace showing elevated mean right atrial pressure (30 mmHg) with prominent X and Y descents. The combination of elevated mean pressure, inconspicuous positive waves, and prominent descents result in the classic 'M' or 'W' pattern (Friedrich's sign).

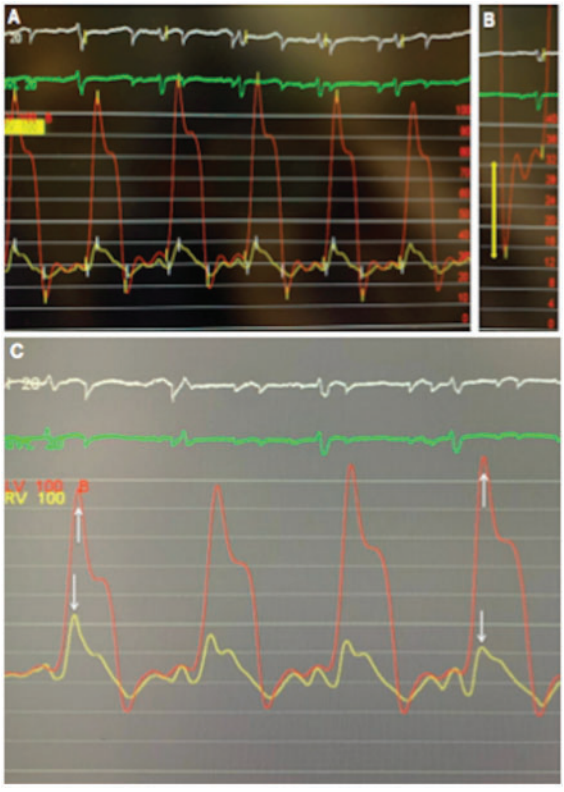

Figure 4 (*A*) Simultaneous left ventricular–right ventricular pressure tracings showing equalization of diastolic pressures. (*B*) Illustrating the typical dip and plateau waveform (square root sign) with deep left ventricular rapid flow wave measuring >7 mmHg (double headed arrow). (*C*) Simultaneous left ventricular–right ventricular pressure trace shows ventricular discordance (exaggerated ventricular interdependence).

The patient's postoperative stay was uneventful with a dramatic improvement of his symptoms and complete resolution of anasarca. He had a significant 12 kg weight loss during his hospital stay and was discharged 10 days later off all diuretics. Medications at discharge included the continuation phase of anti-TB therapy (isoniazid 300 mg o.d. and rifampicin 600 mg o.d.) along with pyridoxine 20 mg o.d. and vitamin C 1 g b.i.d. Additionally, Paracetamol tablet 1 g every 8 h and Tramadol capsule 50 mg every 12 h (for 4 days) were prescribed to relieve post-surgical pain.

In the 4-week of follow-up (via phone consultation due to hospital COVID-19 related protocols), he reported doing very well, asymptomatic with no residual heart failure symptoms.

Three months after discharge, he remained asymptomatic with good functional status and normal exercise capacity. Transthoracic echocardiogram was performed to rule out post pericardiectomy ventricular dilatation and low cardiac output syndrome, which showed normal LV systolic function, indeterminate diastolic dysfunction with no evidence of a constrictive pattern in Doppler signals.

Discussion

The mainstay and only treatment available for CP is pericardiectomy.[5] An observational study conducted over a 20-year period on 99 patients in Montreal showed 7.9% hospital mortality for isolated pericardiectomy. Surgical intervention within 6 months of the onset of symptoms was associated with reduced hospital mortality (odds ratio 0.11). Despite the high perioperative mortality rate, there was a significant improvement in functional status in the survivors.[6] The importance of diagnosing CP early has been well established; additionally, performing pericardiectomy early is associated with favourable outcomes both in the short and long terms.[7]

Our patient was not on steroids on admission, which is the most commonly used adjunctive therapy for tuberculous pericarditis.[5,8] To date, the largest trial of its use is the 'investigation of the management of pericarditis trial' that showed no significant difference in the

Table 1 The main echocardiographic and invasive haemodynamic differences between Constrictive pericarditis and restrictive cardiomyopathy

Parameters	Constrictive pericarditis	Restrictive cardiomyopathy
Echocardiographic features		
Right atrium pressure	Increased	Increased
Ventricular filling pressures	Increased (RV = LV)	Increased (LV>RV)
Diastolic filling	Impaired late filling	Impaired early filling
2D echo	Pericardial thickening with septal bounce	LV hypertrophy with biatrial enlargement
Respiratory variation in ventricular filling	Present	Absent
Tissue Doppler medial mitral annulus *E′* velocity	> 8 cm/s (annulus reversus)	<6 cm/s
Hepatic vein expiratory diastolic reversal/forward flow velocity ≥ 0.8	Present	Absent
Invasive haemodynamic parameters		
Prominent 'y' descent	Present (Friedrich's sign)	Variable
Equal right and left-sided filling pressures	Present	Usually left > right
Left ventricular rapid filling wave	>7 mmHg	≤7 mmHg
Square root sign	Present	Variable
Ventricular interdependence	Discordance	Concordance
Pulmonary artery systolic pressure > 50 mmHg	No	Common

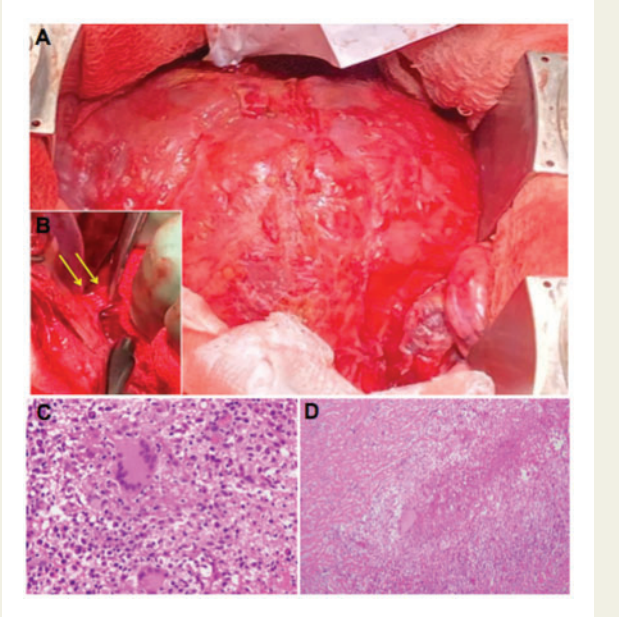

Figure 5 (*A and B*) During pericardiectomy, the pericardium appears diffusely thickened and stiff with plenty of abscesses cavities and dense adhesions (yellow arrows). (*B*) Haematoxylin and Eosin (H&E) stain shows granulomatous reaction with two separate multinucleated giant cells in the high power microscopic view. (*C*) The pericardial biopsy showing a large caseating granuloma with multinucleated giant cells.

composite of death, cardiac tamponade requiring pericardiocentesis, or CP between patients who received prednisolone and those who received placebo (23.8% and 24.5%, respectively; hazard ratio 0.95, 95% confidence interval 0.77–1.18; $P = 0.66$).[9]

In conclusion, our patient had a rapid decompensation. The diagnosis was made promptly owing to the high clinical suspicion, and total pericardiectomy was performed timely after the confirmation of the clinical diagnosis. There were no complications intraoperatively, and postoperatively, there was a rapid improvement in his symptoms. This further demonstrates the paramount importance of diagnosing and treating constriction early. This case is unique since the echocardiogram acoustic windows were suboptimal owing to pericardial thickening, and the CT did not show significant calcification of the thickened pericardium. Cardiac magnetic resonance imaging (MRI) plays an increasing role in the diagnostic work up of patients with suspected CP due to its capability to thoroughly depict both the morphologic aspects of the pericardium and the dynamic functional consequences of pericardial constriction. In our case, the patient refused a cardiac MRI scan due to claustrophobia.

A novel approach of assessing haemodynamics through the right radial artery and right antecubital vein facilitated the diagnosis of pericardial constriction pathology with confidence.

Lead author biography

Nooraldaem Yousif MD, FRCP, FACC, FESC is a Consultant interventional cardiologist at Mohammed Bin Khalifa Bin Sulman Al Khalifa Specialist Cardiac Centre (MKCC) in the Kingdom of Bahrain. He is board certified in internal medicine and cardiology. He completed adult cardiology fellowship at Zurich University Heart Centre, followed by interventional cardiology fellowship

at Andreas Gruentzig catheterization laboratories, Switzerland. His interests include: Complex, High-Risk and Indicated PCI (CHIP), Chronic Total Occlusion (CTO) PCI and clinical research in the field of Cardiology.

Supplementary material

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

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 images and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: None declared.

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