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CASE REPORT
Heart failure

Early-onset post-cardiotomy severe constrictive pericarditis: a case report

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Background Constrictive pericarditis (CP) can be one of the most challenging conditions to diagnose within cardiovascular medicine. Iatrogenic causes of CP are increasingly recognized in higher income countries. This case provides insight into the need for clinical suspicion when diagnosing this relatively under recognized clinical entity as well as the need for multimodality imaging combined with invasive haemodynamic assessment.

Case summary A 68-year-old man presented with decompensated heart failure 4 weeks after open-heart surgery. A diagnosis of early-onset post-cardiotomy CP was made using multimodality imaging and invasive haemodynamic assessment, which demonstrated the cardinal features of constrictive physiology. Surgical intervention with two pericardiectomy procedures was pursued given the aggressive and recalcitrant nature of his presentation. Our patient died shortly after his second surgery due to progressive multi-organ dysfunction.

Conclusion Constrictive pericarditis is a challenging but important clinical entity to diagnose. Differentiating CP from restrictive cardiomyopathy is important as there are key differences in management and prognosis. Our case supports the clinical utility of multimodality imaging combined with invasive haemodynamic assessment in patients with suspected CP.

Keywords Constrictive pericarditis • Pericardial disease • Iatrogenic pericarditis • Case report • Post-cardiotomy pericarditis

ESC Curriculum 2.2 Echocardiography • 2.3 Cardiac magnetic resonance • 6.6 Pericardial disease

Learning points

- Recognize the increasing prevalence of iatrogenic pericarditis.
- Understand the role of multimodality imaging and invasive haemodynamic assessment in the diagnosis of CP.

Introduction

Constrictive pericarditis (CP) can be one of the most challenging clinical entities to diagnose within cardiovascular medicine potentially, which can lead to under recognition of this potentially fatal disease. High income countries are increasingly recognizing the rising incidence of iatrogenic pericarditis.[1,2] Similarities in clinical presentation and diagnostic features of restrictive cardiomyopathy (RCM) and CP are often seen. Differentiating these distinct clinical entities is important as there are key differences in management options and prognosis. Multimodality imaging and invasive haemodynamic assessment are supported by consensus guidelines in the diagnostic pursuit of CP.[1] We present a case detailing the early-onset, post-cardiotomy CP demonstrating the cardinal diagnostic features of CP.

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Timeline

4 weeks prior to initial presentation	Coronary artery bypass grafting (CABG) and aortic valve replacement (AVR) for severe aortic stenosis and concomitant stable coronary artery disease
Initial presentation to hospital	Presented with features of acutely decompensated congestive cardiac failure
1 week after initial presentation	Had been investigated with transthoracic echocardiogram, cardiac magnetic resonance imaging, and simultaneous left and right heart catheterization to establish the unifying diagnosis of CP
2 weeks after initial presentation	Prolonged first pericardiectomy procedure complicated by tears to the right ventricular wall and pulmonary artery (densely adherent pericardium difficult to decorticate leading to decision for a second, staged pericardiectomy procedure)
2 weeks + 3 days after initial presentation	Had second pericardiectomy procedure
3 weeks after initial presentation	Acute right ventricular dysfunction necessitating veno-arterial extracorporeal membrane oxygenation (VA-ECMO) support
4 weeks after initial presentation	Progressive clinical deterioration with multi-organ dysfunction leading to transition to comfort cares and patient passing away

Case presentation

A 68-year-old man underwent CABG and bioprosthetic AVR without immediate peri-operative complications. One month post-operatively, he presented with dyspnoea on exertion, orthopnea, paroxysmal nocturnal dyspnoea, and extensive oedema. His physical examination was notable for pitting oedema extending from the feet up to the abdomen, a markedly elevated jugular venous pressure and a positive Kussmaul's sign. Auscultation of the chest demonstrated reduced air entry at the bases extending up to the mid zones. The heart sounds were dual with no appreciable murmurs.

His past medical history included multivessel coronary artery disease and severe aortic valve regurgitation. He was treated with CABG to the left anterior descending artery (LAD) and posterior descending artery (PDA) as well as bioprosthetic AVR insertion. He also had paroxysmal atrial fibrillation, hypertension, and hypercholesterolaemia. He was a lifelong non-smoker. There was no prior history of impairment in left or right ventricular function including diastolic parameters. There was no history of pericarditis, myocarditis, autoimmune, or rheumatological conditions.

The patient's clinical signs and symptoms were suggestive of acute decompensated biventricular failure. The differential diagnosis included acute left ventricular dysfunction although there was no clear precipitant for this identified from the patient's clinical history. Other considerations included acute valvular pathology (although no murmur was identified on clinical exam), uncontrolled tachyarrhythmias (again there was no history of palpitations, pre-syncope, or syncope to support this), and pericardial/myocardial disease. Other diagnostic possibilities included superimposed pulmonary pathology such as pulmonary embolism or lower respiratory tract infection.

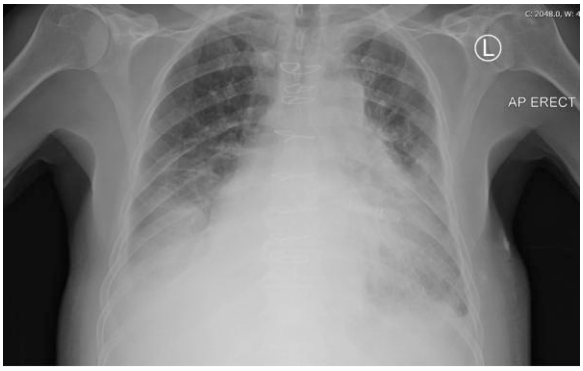

Figure 1 Chest roentgenograph demonstrating pleural effusions without pericardial calcification.

Chest *roentgenogram* demonstrated bilateral pleural effusions without evidence of pericardial calcification (*Figure 1*). A 12-lead electrocardiogram demonstrated sinus rhythm with no abnormalities (*Figure 2*). N-Terminal pro hormone B-type Natriuretic Peptide (NTproBNP) was elevated at 3034 ng/L (normal <126 ng/L). Transthoracic echocardiogram demonstrated normal sized cardiac chambers, normal left ventricular systolic function, and normal function of the aortic bioprosthesis. There was no regional wall motion abnormality noted. Note was made of a bright, thickened pericardium, and a small pericardial effusion. Respirophasic septal motion (septal bounce) consistent with ventricular interdependence was also noted. The inferior vena cava was noted to be distended without collapse on inspiration. Mitral inflow velocities increased by >25% during expiration while tricuspid inflow velocities increased by more than 40% on inspiration (*Figure 3*).

Given the atypically rapid presentation, diagnostic certainty prior to referral for potential repeat sternotomy and pericardial decortication was deemed necessary. Invasive haemodynamic assessment was pursued to confirm the diagnosis of CP. Concurrent invasive coronary angiography was performed as an alternative to coronary CT to limit delays in clinical assessment due to the limited availability of coronary CT at our centre. Coronary angiography demonstrated severe native, multivessel coronary artery disease. The left internal mammary artery graft to the LAD was patent but the saphenous vein graft to the PDA was atretic. Ventriculography confirmed normal left ventricular systolic function with no appreciable gradient across the aortic valve on pigtail catheter pullback. Right heart catheterization with simultaneous left ventricular pressure measurements was performed. The following cardinal features of pericardial constriction were demonstrated (*Figure 4*):

(1) Elevated mean right atrial pressure (17 mmHg) with rapid X and Y descents (the W or M sign) signifying rapid, early diastolic atrial emptying into an underfilled ventricle.
(2) The square root sign in the right and left ventricular pressure waveforms indicating rapid early ventricular filling with a sudden plateau as the ventricles reach capacity within a stiff pericardium.
(3) Equalization of left and right ventricular end-diastolic pressures reflective of a stiff, non-compliant pericardial sac.

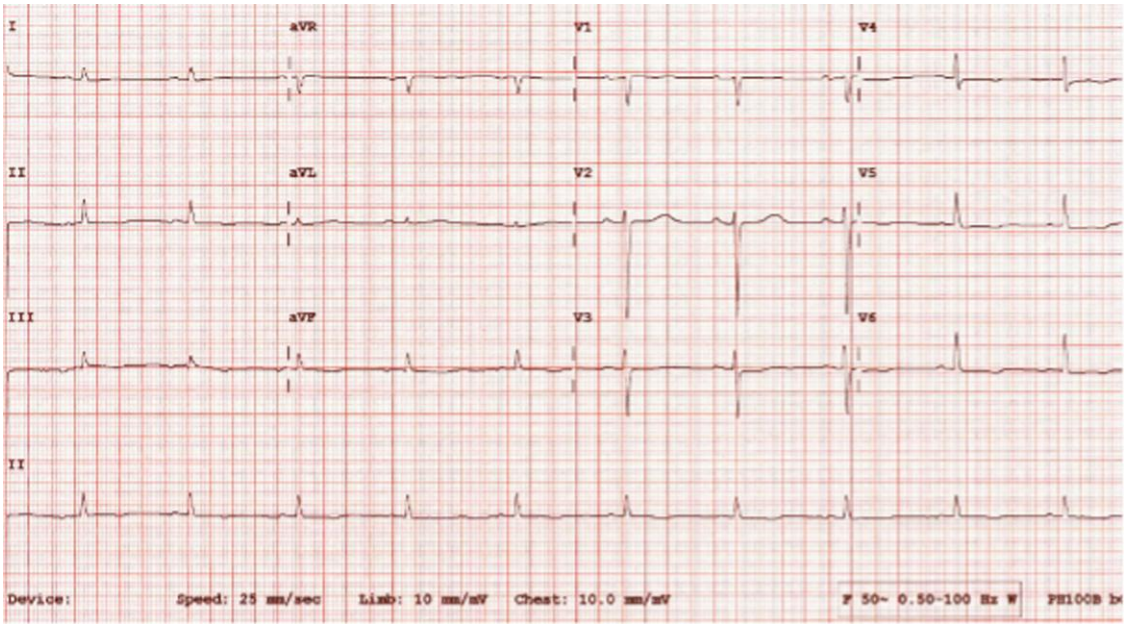

Figure 2 12-Lead electrocardiogram demonstrating sinus rhythm without acute abnormalities. Note that the low peripheral lead voltages, which is one of the non-specific signs of constrictive pericarditis on electrocardiogram.

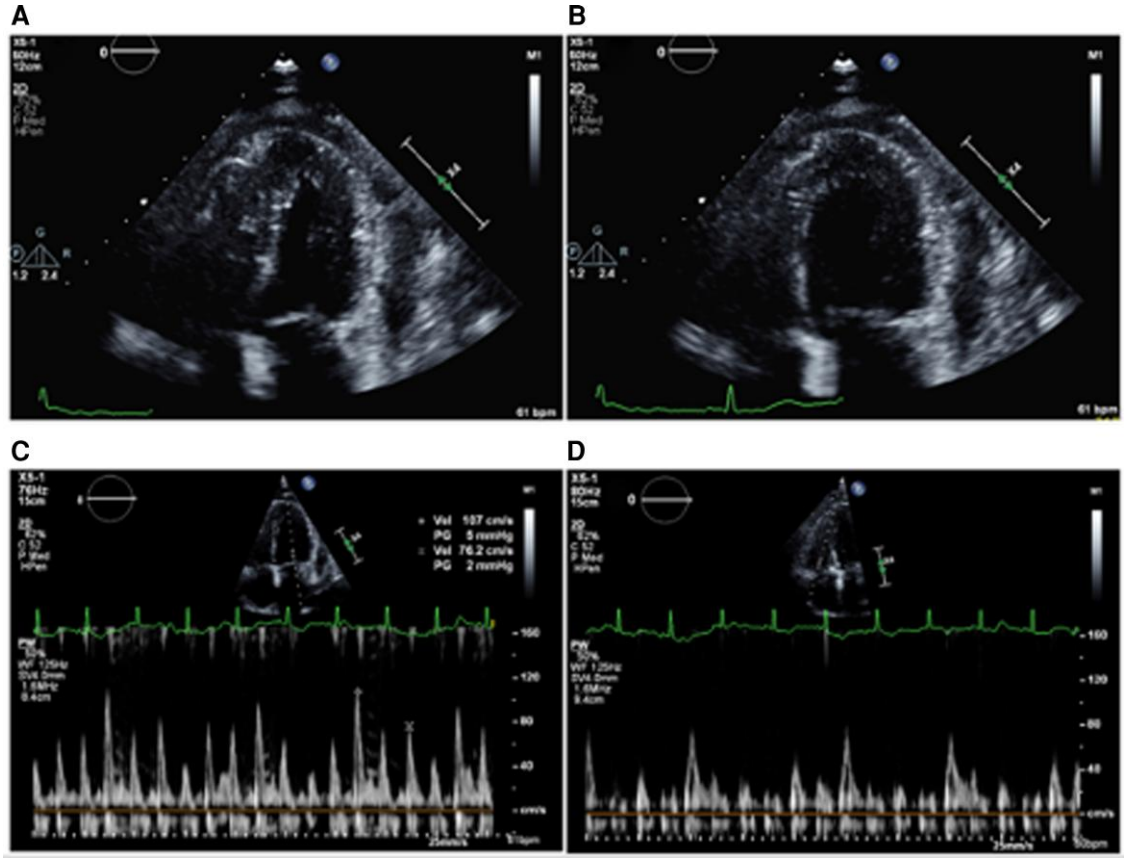

Figure 3 Transthoracic echocardiogram from the apical four-chamber view revealed respirophasic septal motion, seen at end-diastolic (*A*) inspiration and (*B*) expiration, and altered inflow velocities at the (*C*) mitral valve and (*D*) tricuspid valve.

(4) Discordance in the left and right ventricular systolic pressures during the respiratory cycle indicating ventricular interdependence.

Cardiac magnetic resonance imaging (CMR) demonstrated normal cardiac chamber size and respirophasic septal motion consistent with ventricular interdependence. Crescentic thickening (up to 24 mm) of the pericardium around the left ventricular wall was demonstrated but Phase Sensitive Inversion Recover (PSIR) confirmed that the true pericardium measured ~8 mm with up to 16 mm of pericardial fluid identified around the left ventricle (*Figure 5*). Pericardial signal

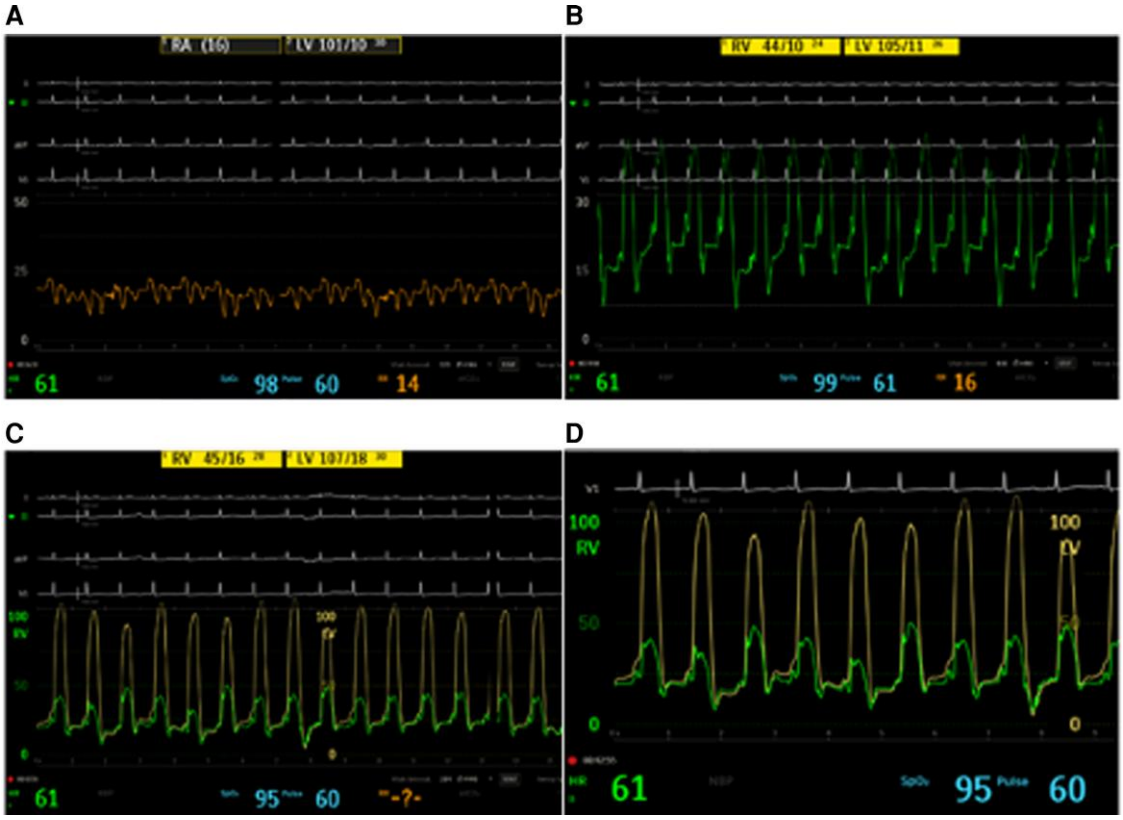

Figure 4 Right and left heart catheterization pressure measurements demonstrated (*A*) elevated right atrial pressure with rapid *x* and *y* descents, (*B*) square root sign, (*C*) equalization of left ventricular and right ventricular end-diastolic pressures, (*D*) discordance in ventricular pressures during respiration.

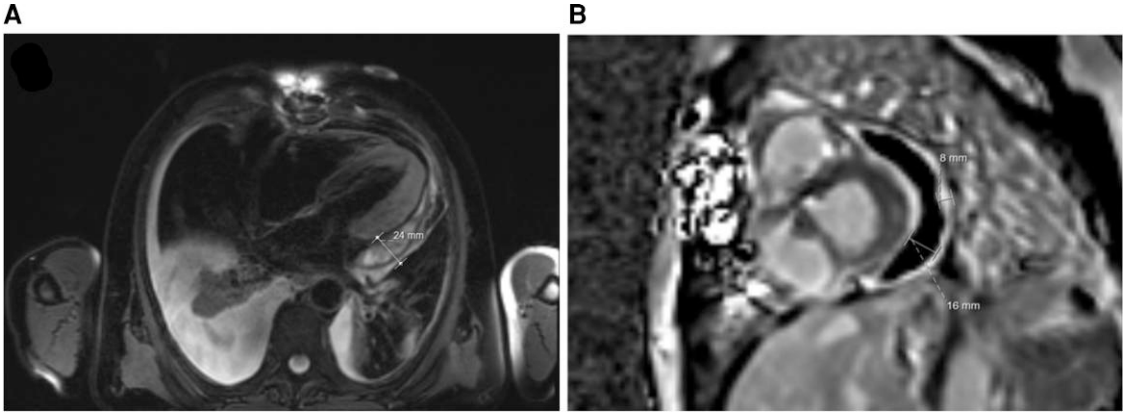

Figure 5 On T2 cardiac magnetic resonance imaging, thickened, fibrous crescent-shaped pericardial mass adjacent to the anterolateral, lateral, and inferolateral walls, measuring up to 24 mm (*A*). Phase Sensitive Inversion Recover sequences with gadolinium contrast demonstrated an 8 mm thick pericardium with up to 16 mm of pericardial fluid around the left ventricle (*B*). Evident interventricular dependence on free breathing module, indicative of contrictive physiology (Supplementary material online, *Video*).

enhancement was noted in short time inversion recovery sequence as well as late gadolinium enhancement on PSIR sequence indicating active inflammation. The left atrium:right atrium volume ratio was normal suggesting acute rather than chronic CP.

The clinical, echocardiographic, and haemodynamic findings were suggestive of post-cardiotomy CP; a diagnosis made more likely by the demonstration of a markedly thickened pericardium on CMR. The presence of the atretic vein graft was suggestive of adequate competitive flow within the native vessel. The absence of regional wall motion abnormalities on echocardiography combined with a normal troponin and the absence of ischaemic ECG changes indicated that the syndrome of heart failure was unlikely to be due to myocardial ischaemia. Despite medical therapy with intravenous frusemide and colchicine, there was no improvement in the patient's clinical condition. Our patient was referred for urgent surgical pericardiectomy after discussion at a multi-disciplinary heart team meeting. At surgery, extremely thickened pericardium, densely adherent to the heart over the right and left ventricles in particular was found. Decortication of the pericardium over the right side of the heart was technically challenging and caused tears to the pulmonary artery and right ventricle, which had to be meticulously sutured. The first operation lasted 8 h. A decision was made to stage pericardial decortication with completion pericardiectomy over the left ventricle (which had not been possible via sternotomy) via a left lateral thoracotomy approach. During the latter operation, extremely thickened pericardium was again found. The left lung was densely adherent to chest wall and pericardium, and injury to the lung occurred on entering chest (subsequently repaired). Unfortunately, the patient developed acute right ventricular dysfunction in the ensuing days, necessitating VA-ECMO support. Seven days after the second procedure, the patient developed multi-organ dysfunction with renal failure requiring continuous renal replacement therapy. His condition deteriorated further, and, after family discussion, active treatment was withdrawn. He passed away shortly afterwards.

Results from pericardial tissue, which was sampled during the patient's procedure demonstrated dense fibrous tissue with fibroblastic activity suggestive of reactive pericarditis. There was no evidence of suppurative inflammation, granulomas, or malignancy to suggest an alternative aetiology of CP.

Discussion

Our case highlights the high index of clinical suspicion required in diagnosing CP as well as the challenges involved in recognizing this potentially life-threatening condition. In this case, the recalcitrant-to-treatment and unusually severe nature of the patient's symptomatology led to consideration of CP with left and right heart catheterization to confirming constrictive physiology. Multimodal cardiac imaging supported the diagnosis of CP and excluded other pathological mechanisms such as RCM.

Constrictive pericarditis is a condition in which a non-compliant pericardium prevents ventricular filling.[3] Tuberculous pericarditis is the leading cause of CP in lower income countries.[1] Whilst almost half of all cases of CP in higher income countries are caused by viruses or are idiopathic in aetiology, iatrogenic causes such as chest radiotherapy and cardiotomy are increasingly recognized.[1,2] There is a paucity of data regarding post-cardiotomy CP with available data being limited to small retrospective case series and case reports. Consequently, the true incidence and risk factors for this potentially fatal clinical entity remain unclear.[4] In the largest available series we could find, patients experiencing post-cardiotomy pericarditis tended to be older, male patients with a history of myocardial infarction or Type 2 diabetes mellitus.[5] The presence of post-pericardiotomy syndrome has been implicated as a risk factor for the development of post-cardiotomy CP.[6]

Despite overlapping clinical and diagnostic features, CP and RCM have key differences in management and prognosis. Differentiating these clinical entities can be challenging. Both conditions impair the diastolic phase of the cardiac cycle. In CP, the pericardium is non-compliant with a fixed volume. Thus, during inspiration [when right ventricular filling is augmented and left ventricular (LV) filling is not], elevated right sided pressures cause translocation of the interventricular septum towards the LV. The converse occurs during expiration. This respirophasic phenomenon is termed ventricular interdependence and signifies compliant ventricles within a non-compliant pericardium: the hallmark feature of CP which is absent in RCM.[3]

Multimodality imaging is recommended in patients with suspected CP.[1,7–9] Chest X-ray or cardiac CT may demonstrate calcific/thickened pericardium.[1,3,7] Transthoracic echocardiogram provides valuable structural and haemodynamic information. Systolic function is typically preserved in both conditions provided the absence of alternative pathology. Strain imaging in CP is of limited benefit but a degree of latent systolic dysfunction may be observed in patients with RCM.[8] Respirophasic changes such as septal bounce, fluctuations in mitral and tricuspid inflow velocities, hepatic vein flow reversal, and variation in pulmonary veins peak diastolic flow are all consistent with CP.[3,7] Tissue Doppler changes such as annulus paradoxus and annulus reverses are also consistent with CP.[3] The role of CMR is still evolving. Free breathing modules used in real time can evaluate respirophasic changes within the cardiac chambers. In addition, CMR provides three-dimensional imaging to guide surgical intervention.[9]

Simultaneous left and right heart catheterization remains the gold standard for diagnosis of CP,[1,3] with the cardinal features being listed above.

Conclusions

Constrictive pericarditis is a rare but life-threatening condition. Iatrogenic causes are increasingly recognized. Diagnosis of CP and differentiation from RCM are challenging but necessary. Invasive haemodynamic assessment remains the gold standard for diagnosis.

Lead author biography

Dr Philopatir Mikhail is a Cardiology Advanced Trainee at Gosford Hospital in Australia with a special interest in coronary artery disease and coronary physiology. He hopes to pursue subspecialty training in Interventional and Structural Cardiology.

Supplementary material

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

Slide sets: A fully edited slide set detailing this case 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.

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