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CASE REPORT
Coronary heart disease

A case report of unusual clinical features of a spontaneous coronary artery rupture: pathologic findings in the rupture site

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Background Spontaneous coronary artery rupture (SCAR) is an extremely rare but life-threatening state. The aetiology and the pathologic findings remain to be fully elucidated.

Case summary A 62-year-old woman, who had been on haemodialysis for 27 years, presented with chest discomfort worsening on deep inspiration that had been ongoing for the past 2 weeks. An echocardiogram and computed tomography showed diffuse pericardial fluid. ST elevation in the broad leads, especially in leads I, II, and aVF, and increased C-reactive peptide and Troponin I levels suggested pericarditis. The patient initially had a stable course with no medications. The chest symptoms disappeared and her vital signs were stable. On Day 13 after admission, however, she had a sudden cardiopulmonary arrest due to a cardiac tamponade. An emergency coronary angiography showed extravasation of the contrast into the epicardium from the branch of the circumflex artery. She was diagnosed with SCAR and underwent a successful coil embolization. However, she went into an irreversible coma due to the cerebral hypoxia. On Day 33, she died of pneumonia. An autopsy showed a rupture of the internal elastic layer of the coronary artery. However, no specific findings, such as aneurysm and dissection, were evident. The common atherosclerotic changes were observed.

Discussion The stable condition lasting for over 2 weeks was a rare clinical course for SCAR. Long-term hypertension and dialysis would have caused the rupture of the coronary artery with common atherosclerotic changes. We should consider SCAR as one of the differential diagnoses when we observe pericardial fluid.

Keywords Case report • Spontaneous coronary artery rupture • Pericardial effusion • Cardiac tamponade • Coronary atherosclerosis

Learning points

- In patients with pericardial effusion, spontaneous coronary artery rupture should be listed as a differential diagnosis.
- Spontaneous coronary artery rupture can present various clinical courses and its aetiology is different. Spontaneous coronary artery rupture can occur in the coronary artery even with the common atherosclerotic change.

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Introduction

Spontaneous coronary artery rupture (SCAR) is a life-threatening state. The incidence is extremely rare and most cases present with sudden cardiac arrest. Therefore, the aetiology of the SCAR remains to be fully elucidated.[1] We present a case who had an unusual clinical course of SCAR. The patient's condition was stable despite the bleeding, and the condition became critical suddenly. This is the first report that details pathophysiologic findings of the spontaneous rupture site of the coronary artery.

Timeline

Day -14	Chest discomfort that worsened on deep inspiration appeared.
Day 0 (admission day)	• Diffuse pericardial fluid was seen on echocardiography and computed tomography. • No evidence of cardiac tamponade and no contrast agent leakage in the pericardium. • ST elevation in broad leads and elevation of C-reactive peptide and high-sensitivity Troponin I.
Day +1∼+12	Pericardial effusion initially increased and then decreased; vital signs were stable.
Day +13	• Cardiopulmonary arrest occurred. • Coronary angiography showed a rupture of the branch of the left circumflex artery, and coil embolization was performed. • The patient went into an irreversible coma.
Day +33	• The patient died from pneumonia, and an autopsy was performed.

Case presentation

A 62-year-old woman was referred to our hospital with chest discomfort ongoing for 2 weeks. She had chronic renal failure secondary to malignant hypertension. She had been undergoing dialysis for 27 years. Her symptoms worsened when she took a deep breath, and she had experienced palpitations for a few hours before her arrival. At the time of her visit, her blood pressure was 85/54 mmHg, pulse rate was 150 ppm, and SpO_2 was 95% (room air). An electrocardiogram (ECG) demonstrated atrial fibrillation, which spontaneously converted to sinus rhythm within 1 h. Subsequently, the palpitations disappeared and the blood pressure increased to 100/62 mmHg. The sinus rhythm on the ECG showed a slight degree of ST elevation in the broad leads, especially in leads I, II, and aVF (*Figure 1*). A transthoracic echocardiogram (TTE) showed that the left ventricular wall motion, thickness, and size were within normal range. No valve dysfunction was observed. Diffuse mild pericardial fluid was seen. The maximum thickness of the fluid was 6 mm at the anterior right ventricle; however, no findings suggestive of heart chamber collapse were evident (*Figure 2A and B*; Supplementary material online, *Videos S1A* and *B*).

A blood test showed creatine kinase and creatine kinase-MB levels in the normal range. The C-reactive peptide and high-sensitivity Troponin I levels were elevated (5.2 mg/dL and 86 ng/mL, respectively). Plain and contrast-enhanced chest computed tomography (CT) was performed (*Figure 3*). No findings indicated either aortic dissection or pulmonary embolism. The CT value of pericardial effusion was 35–74 HU, which indicated the bloody effusion. However, no contrast agent was observed in the pericardium. We diagnosed the patient with pericarditis and hospitalized her for treatment and observation for her pericardial effusion. Her inflammatory marker levels became normal, and the ST changes disappeared after several days with no medications. She had no chest pain and the creatinine kinase level was within normal range during the hospitalization. Her blood pressure during hospitalization was 120–140/80–90 mmHg. On Day 9 after hospitalization, the effusion increased (12 mm at the anterior right ventricle) and caused a mild diastolic collapse of right atrium; however, her haemodynamics remained stable. On Day 11, the pericardial effusion decreased significantly when compared with that on admission (5 mm at the anterior right ventricle).

On Day 13, after haemodialysis, she again experienced precordial discomfort and she had a sudden cardiopulmonary arrest (CPA) on the bed. The cardiac rhythm was pulseless electrical activity. Cardiopulmonary resuscitation was quickly initiated and a return of spontaneous circulation was achieved. A markedly increased pericardial effusion and chamber collapse which caused a cardiac tamponade were observed on the TTE. The systolic blood pressure was 60 mmHg. We performed pericardial drainage, and 300 mL of bloody fluid was drained. A blood test showed the troponin I level was 59.5 pg/mL. The emergency coronary angiography showed no significant abnormality of coronary artery forms. Mild stenosis was observed in the right coronary artery and the left circumflex artery (LCX). The middle part of the left anterior descending artery and the diagonal branch had severe stenosis with calcification. No coronary flow limitation was observed; however, the extravasation of contrast into the epicardium was seen from the branch of the LCX (Ellis classification: Type III) (*Figure 4A* and Supplementary material online, *Video S2A*). The patient was diagnosed with SCAR. The ruptured vessel was small, so we thought that the coil embolization would work for stopping the bleeding. The extravasation of contrast agent disappeared after inserting three coils (*Figure 4B* and Supplementary material online, *Video S2B*).

Despite the steadiness of the patient's haemodynamics after the coil embolization, she went into an irreversible coma. It took 21 min to return to spontaneous circulation from CPA. A head CT performed 6 days after the CPA showed an unclearness of the corticomedullary junction, which suggested cerebral hypoxia. Electroencephalography performed 5 days after the CPA indicated a continuous low voltage and semi-rhythmic delta activity, which suggested diffuse encephalopathy.

At 20 days after the CPA, she died from pneumonia. An autopsy revealed no specific findings in the coronary artery, such as aneurysm or dissection. Pathologically, intimal hypertrophy and medial thinning, which are common in atherosclerosis, were observed at the point of rupture. A rupture of the internal elastic layer was observed; however, no necrosis or inflammation was evident (*Figure 5*).

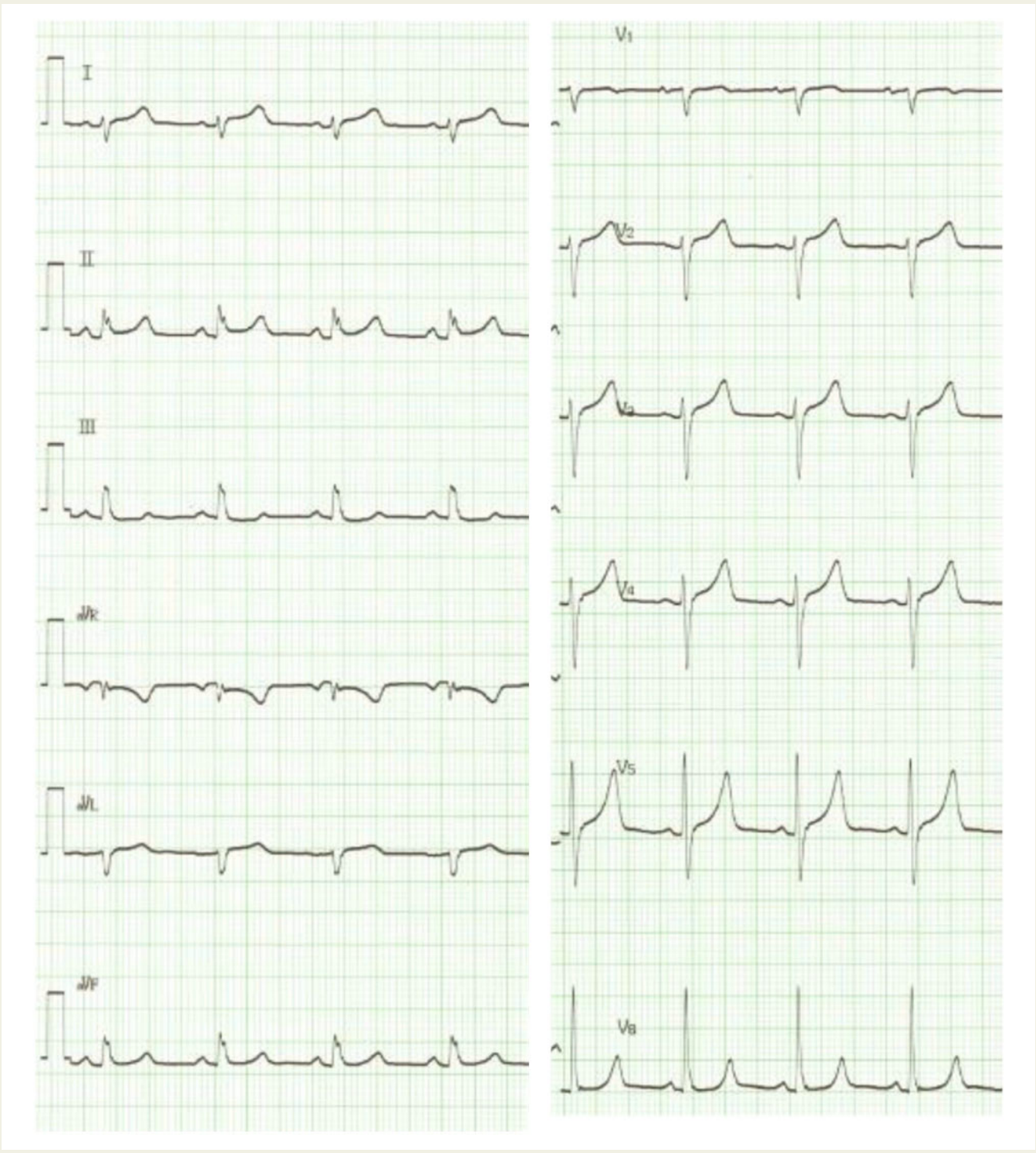

Figure 1 Twelve-lead electrocardiogram at admission.

Discussion

Clinical features and diagnosis

The clinical features of SCAR are usually acute, and the symptoms are similar to those of acute coronary syndrome or aortic dissection.[1,2] In contrast, mild symptoms, such as fatigue and shortness of breath, have been reported in some cases.[2,3] In patients with mild symptoms, the intrapericardial mass (haematoma) compressed the coronary artery, which slowed the bleeding from the ruptured vessels. In the present case, the patient had precordial discomfort lasting for 2 weeks, and this discomfort became worse when she took a deep breath. Although

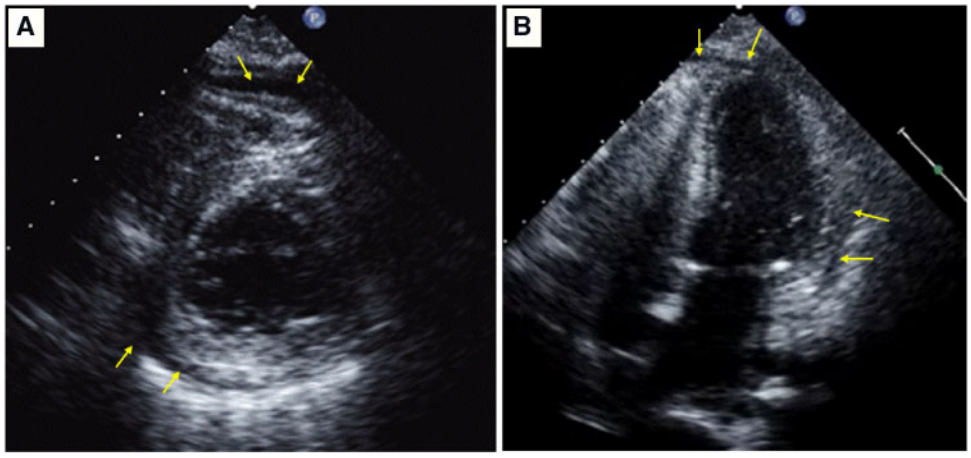

Figure 2 Echocardiography at admission showed diffuse pericardial fluid (yellow arrows). (*A*) Short-axis view and (*B*) four-chamber view.

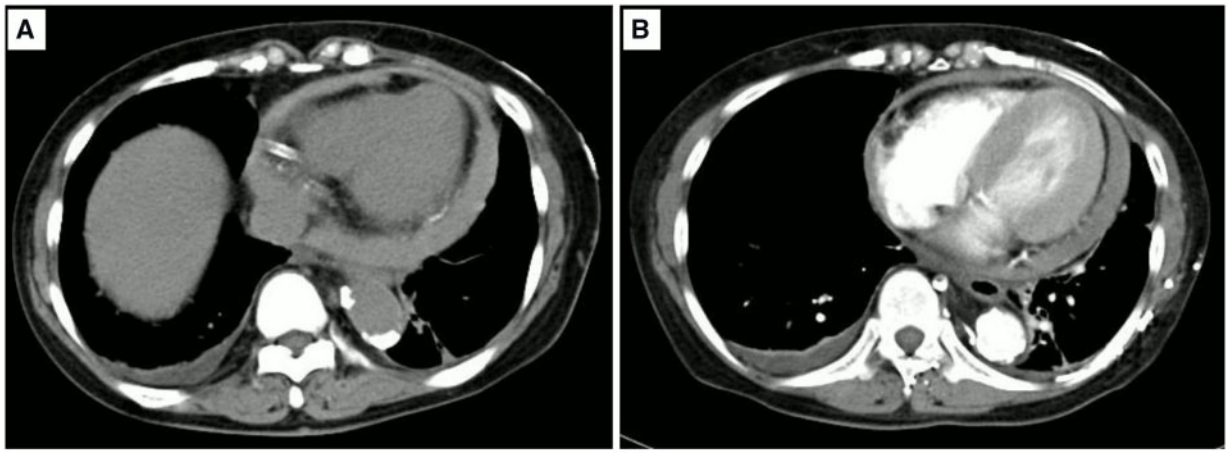

Figure 3 Chest computed tomography showed pericardial effusion. (*A*) Plain computed tomography and (*B*) contrast-enhanced.

bloody pericardial effusion was suggested by the CT value, there was no pericardial haematoma or mass. After hospitalization, the ST elevation normalized and her symptoms disappeared. We could not assume the possibility of SCAR because that condition is extremely rare, and the symptoms and clinical course in this patient were concordant with pericarditis. Retrospectively, we speculate that the bleeding and haemostasis of the coronary artery had been repeated, occurring both before and after her hospitalization. The ruptured vessel was small; therefore, the stable clinical course lasted over 2 weeks. In addition, the bleeding in the pericardium would lead to secondary pericarditis which showed the unusual symptom of the SCAR. On Day 13 after admission, the bleeding suddenly increased and resulted in a cardiac tamponade. The reason massive bleeding occurred at that time was unclear. However, the heparinization and blood pressure changes during the haemodialysis would have had a risk of the massive bleeding.

Treatment of spontaneous coronary artery rupture

We selected a coil embolization for the treatment of this SCAR. Previous reports have showed that emergency cardiothoracic surgery is usually performed.[2,4] The choice of the treatment strategy depends on the aetiology and the patient's condition. If the condition is unstable and the culprit vessel is small, then a coil embolization would be faster for controlling the bleeding when compared with cardiothoracic surgery. In this patient, although cardiopulmonary resuscitation was started immediately, it took 21 min to return to spontaneous circulation. The electroencephalography findings and head CT after the CPA suggested cerebral hypoxia, which would have caused her coma.

Pathophysiologic findings

Spontaneous coronary artery rupture is associated with known underlying diseased (e.g. aneurysm, Ehlers–Danlos syndrome, and Behcet's

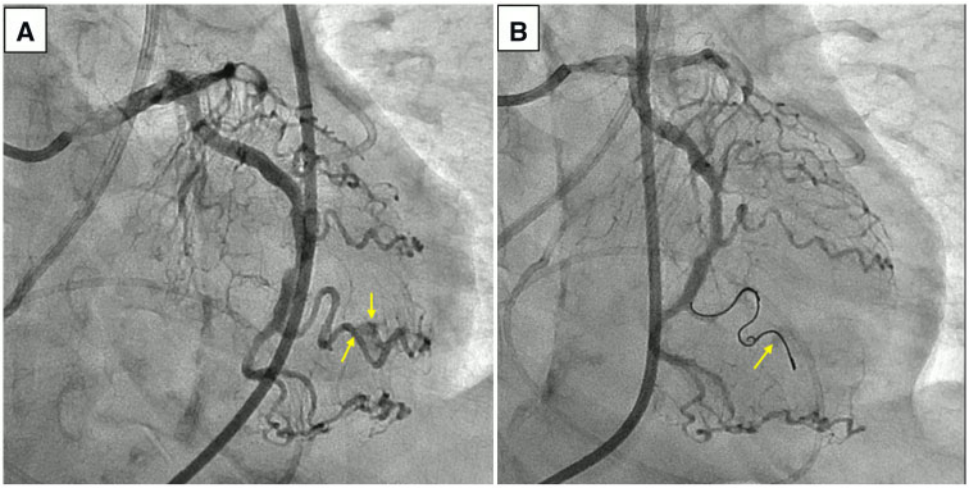

Figure 4 (*A*) Extravasation of contrast agent into the epicardium from the branch of the circumflex artery and (*B*) after coil embolization.

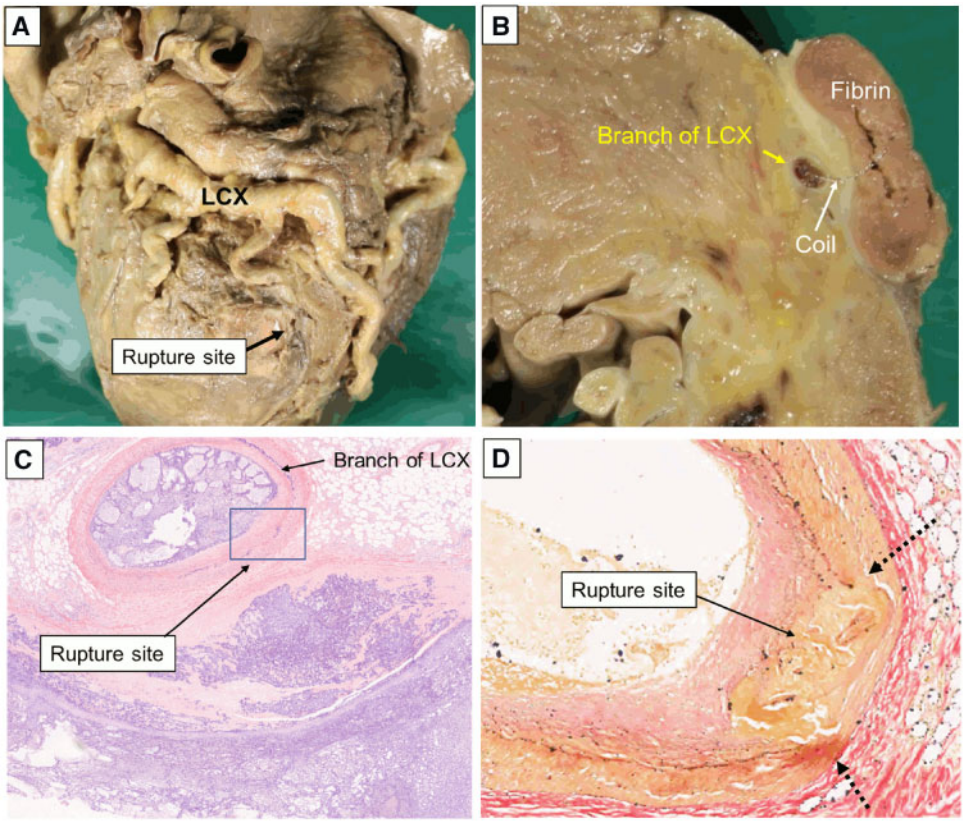

Figure 5 (*A* and *B*) Fibrin can be seen around the rupture site of the branch of the left circumflex artery. The coil for embolization is indicated (white arrow). (*C*) The short axis of the rupture site. (*D*) Elastica van Gieson staining revealed intimal hypertrophy, medial thinning, and rupture of the internal elastic layer (black dotted arrow).

disease),[1,4,5] but some reported SCAR cases have no underlying heart diseases.[2,6] In the present case, we first reported the details of the pathophysiologic findings of the rupture site, which showed no evident underlying coronary disease. Common atherosclerotic changes were observed in the culprit coronary artery, and a rupture of the internal elastic layer was seen. A possible mechanism for this SCAR could be the malignant hypertension and the dialysis. In addition to the atherosclerotic changes, the high diastolic pressure and haemodynamic changes due to the dialysis would weaken the vessel wall. The anticoagulant state during the dialysis would also promote bleeding.

Conclusion

We reported a case of unusual clinical features of a SCAR. The SCAR was successfully treated by a coil embolization; however, the patient subsequently died from pneumonia. The pathophysiologic findings of the rupture site revealed common atherosclerosis changes and a rupture of the internal elastic layer. Spontaneous coronary artery rupture is a rare disorder but it should be kept in mind when pericardial effusion is observed particularly in patients with poorly controlled hypertension.

Lead author biography

Dr Mitsuru Takami is an assistant professor of Division of Cardiovascular Medicine at Kobe University Hospital. In 2012, he was a winner of the Young Investigator Award of Japanese Heart Rhythm Society. From 2013 till 2015, he worked as a research fellow at the translational interventional electrophysiology laboratory, Mayo Clinic.

Supplementary material

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

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

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none declared.

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