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CASE REPORT
Coronary heart disease

Percutaneous coil embolization and stent implantation for multiple coronary-to-pulmonary artery fistulas with giant coronary aneurysms: a case report

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Background	Multiple coronary-to-pulmonary artery fistulas (CPAFs) with giant coronary aneurysms (CAs) are extremely rare. The appropriate therapeutic indication and strategy for CPAFs have not been established.
Case summary	Herein, we report the case of an asymptomatic 74-year-old woman with multiple CPAFs associated with giant CAs that had gradually developed over a 4-year period. After heart team discussion, we were successfully treated by minimally invasive intervention using transcatheter coil embolization and coronary stent implantation to prevent ruptures.
Discussion	Coronary-to-pulmonary artery fistulas required evaluation of the appropriate timing of therapy initiation with reference to the presence of symptoms and fistula and aneurysm sizes, and determination of the optimal therapeutic approach with reference to the anatomy of the fistula with aneurysm and patient background characteristics.

Keywords	Coronary-to-pulmonary artery fistulas • Coronary aneurysms • Coil embolization • Coronary stent implantation • Case report
ESC Curriculum	3.1 Coronary artery disease • 2.1 Imaging modalities • 9.7 Adult congenital heart disease

Learning points

- Multiple coronary-to-pulmonary artery fistulas with giant coronary aneurysms are extremely rare complex coronary anomalies for which the ideal management and approach are not well established.
- The appropriate timing of therapy initiation is evaluated with reference to the presence of symptoms and fistula and aneurysm sizes.
- The optimal therapeutic approach is determined with reference to the anatomy of the fistula with aneurysm and patient background characteristics.

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Introduction

Coronary artery fistula is direct communication between a coronary artery and a great vessel or cardiac chamber and is noted in 0.1–0.2% of patients who undergo coronary angiography (CAG).[1] Among them, multiple coronary-to-pulmonary artery fistulas (CPAFs) with giant coronary aneurysms (CAs) are extremely rare. In cases with coronary artery fistula, surgical repair is associated with a high incidence of complications, including perioperative myocardial infarction.[2] Here, we reported a case of successful percutaneous coil embolization and stent implantation for multiple CPAFs with giant CA.

Timeline

Presentation	Referral to our hospital because of the gradual development of multinodular shadows within the anterior mediastinum
1 month later	Computed tomography angiography (CTA): multiple coronary-to-pulmonary artery fistulas (CPAFs) with giant coronary aneurysms (CAs). The CA diameter had grown from 23 to 30 mm over 4 years.
2 months	Coronary angiography (CAG): CPAFs originate from the proximal right coronary artery (RCA). CPAFs with giant CAs originating from the proximal and mid-left anterior descending artery (LAD). Intravascular ultrasound (IVUS) revealed the presence of an aneurysm-coronary septum at the orifice of the aneurysm.
3 months	Percutaneous coil embolization for CPAFs at RCA CAG: complete occlusion of CPAFs
4 months	Percutaneous coil embolization for CPAFs at the proximal LAD Percutaneous coil embolization and coronary stent implantation for CPAFs with giant CAs CAG: the mild residual flow into the aneurysm at mid-LAD
7 months (3 months after treatment)	CT: a partial thrombotic occlusion of the aneurysm at mid-LAD
16 months (12 months after treatment)	Clinically well and asymptomatic with no symptoms of coronary ischaemia CT: completely occlusion of the aneurysm at mid LAD

Case presentation

Multinodular masses were detected within the anterior mediastinum of a previously healthy 74-year-old woman by plain chest computed tomography (CT) screening during a comprehensive medical check-up despite a normal chest X-ray. Despite having no symptoms, she was referred to our hospital because of the gradual development of multiple nodules over a 4-year period. She had no coronary risk factors and no history of Kawasaki's disease or vasculitis. Her vital signs were normal. Blood tests showed slightly elevated brain natriuretic peptide [38.9 pg/mL (≤18.4 pg/mL)] and D-dimer [1.8 μg/mL (≤1.0 μg/mL)] levels. Twelve-lead electrocardiography (ECG) showed a normal sinus rhythm and incomplete right bundle branch block. Transthoracic echocardiography showed a normal ventricular systolic function with no wall motion abnormalities or valvular heart disease. Contrast-enhanced cardiac CT showed multiple contrast-enhanced nodules adjacent to the ascending aorta (*Figure 1A*, *Video 1*). Three-dimensional reconstruction of the coronary arteries using CT angiography revealed multiple giant CA and abnormally enlarged and twisted arteries in the superior anterior part of the heart (*Figure 1B*). The largest aneurysm originating from the proximal left circumflex artery had become spontaneously occluded. Although the anterior mediastinal multinodular masses suggested mediastinal tumours, such as lymphoma, thymoma, thymic cyst, germ cell, and mediastinal thyroid mass, contrast-enhanced cardiac CT excluded these diseases. Iatrogenic causes and coronary injury were also excluded, as the patient had never underwent surgery or coronary intervention. Genetic testing excluded an underlying connective tissue disorder. A whole-body CT scan detected no aneurysms in any other part of the body. On admission, CAG revealed two CPAFs originating from the proximal right coronary artery (RCA) (*Figure 2A*). Although the CPAFs originating from the left anterior descending artery (LAD) were accompanied by giant CA, the orifices of the fistulas from the LAD could not be identified by CAG (*Figure 2B*, *Video 2*). Selective contrast injection using a guide-extension catheter (GuidelinerV3; Japan Lifeline, Tokyo, Japan) and intravascular ultrasound (IVUS) revealed a CPAF originating from the proximal LAD and a saccular aneurysm in the middle LAD. The orifice of the CA was small due to the presence of an aneurysm-coronary septum (*Video 3*, Supplementary material online, *Figure S1A*). Right heart catheterization (RHC) showed a significant increase in oxygen saturation between the right ventricle and pulmonary artery (71% and 78%, respectively), while the mean pulmonary artery pressure and pulmonary capillary wedge pressure were normal (19 and 9 mmHg, respectively). The ratio of pulmonary blood flow to systemic blood flow (Qp/Qs) was 1.3. There was no evidence of myocardial ischaemia with adenosine Tallium-201 myocardial perfusion scintigraphy, which might detect myocardial ischaemia caused by CPAF.[3]

While there were no subjective symptoms and complications related to CPAF and CA, the CA diameter had grown from 23 to 30 mm over 4 years, indicating a risk of aneurysmal rupture (*Figure 4A* and *B*). Therefore, we decided to treat the CPAFs with CA after the heart team held discussions with cardiac surgeons and paediatric cardiologists. Other than discussions in the heart team in our hospital, we discussed with adult congenital heart disease specialists and paediatric cardiologists in other hospitals before treatment, regarding the therapeutic indication and therapeutic strategy for this patient. Surgical correction was not feasible, since it will be difficult to identify the orifice of the fistula originating from the LAD, which requires a dissection of the cluster of CPAFs. Furthermore, the patient

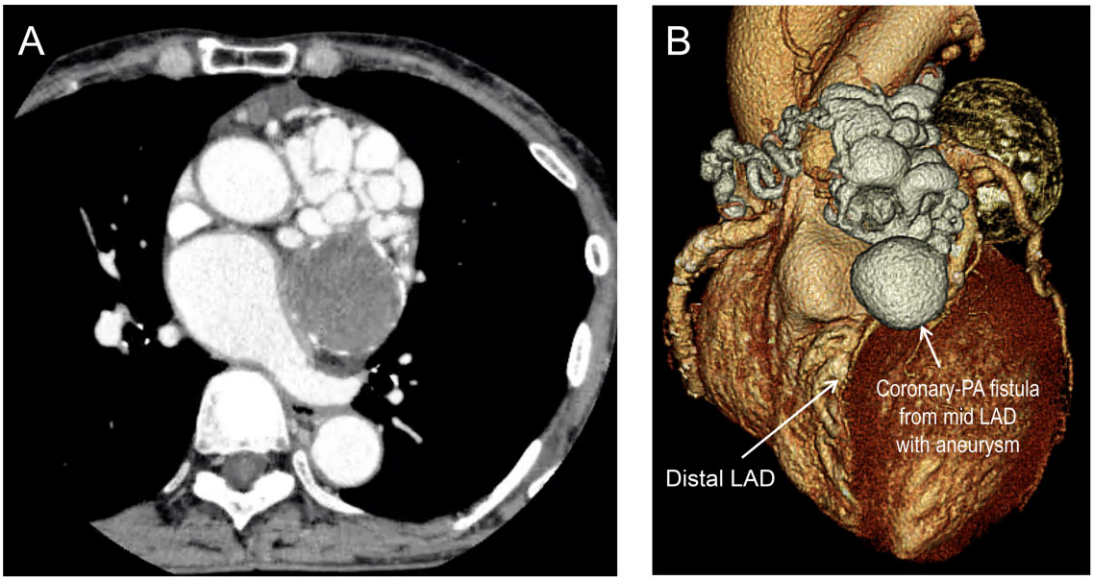
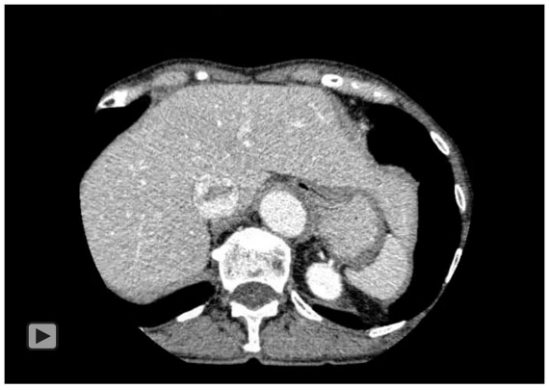


Figure 1 Contrast-enhanced cardiac computed tomography. (*A*) Contrast-enhanced cardiac computed tomography showed multiple contrast-enhanced nodules adjacent to the ascending aorta. (*B*) Three-dimensional reconstruction of coronary arteries using computed tomography angiography.



Video 1 Contrast-enhanced cardiac computed tomography image.

preferred minimally invasive treatment by a transcatheter approach. We, therefore, decided to perform transcatheter closure of these fistulas using detachable coils and a coronary stent.

Initial intervention was performed for the CPAFs of the RCA. A guidewire (SION blue; ASAHI INTEC, Aichi, Japan) and a steerable microcatheter (LEONIS Mova; Sumitomo Bakelite, Tokyo, Japan) were inserted into the fistula using a 7-Fr guiding catheter (JR-4, Heartrail II; Terumo Corp., Tokyo, Japan) from the right radial artery. The two fistulas of the RCA were embolized with five coils in total (Target XL360 soft; Stryker, Kalamazoo, Michigan) up to 6 mm in diameter and 20 cm in length. Post-embolization CAG confirmed the occlusion of all CPAFs from the RCA (*Figure 3A*). Embolization of the fistula at the proximal LAD was performed at a later date. The fistula was embolized with 6 coils up to 8 mm in diameter and 30 cm in length in a manner similar to the CPAFs of the RCA. The saccular aneurysm of the mid-LAD was subsequently treated. Based on the previous IVUS findings, we planned to treat the aneurysm with stent-assisted coil embolization.[4] Initially, a coil (3 mm in diameter and 90 mm in length) was deployed at the orifice of the aneurysm to disturb the blood flow into the aneurysm. A drug-eluting stent (DES) (CoCr-ZES 4.0/12 mm; Medtronic, Santa Rosa, CA, USA) was then delivered to cover the aneurysm orifice along with the deployed coil. Unfortunately, the coil migrated distally due to interference between the coil and stent. Since the first stent could not fully cover the coil, we decided to implant a second stent (PtCr-EES 3.0/16 mm; Boston Scientific, Marlborough, MA, USA). Although final angiography showed a mild residual flow into the aneurysm, IVUS findings showed the aneurysm orifice was mostly covered by the stents (Supplementary material online, *Video S1*, *Figure 3B*, Supplementary material online, *Figure 1B*). The periprocedural myocardial infarction (defined by elevation of cTn values > 5 × 99th percentile URL) was not documented with all procedures in this case.

Follow-up CAG showed that the blood inflow into the saccular aneurysm was minimal after 6 months (Supplementary material online, *Figure S2*). This patient presented with nasal bleeding 1 month after stent implantation. Therefore, she needed the de-escalation from DAPT with aspirin and clopidogrel to a single antiplatelet therapy (SAPT) with clopidogrel. Since then, her post-procedure course has been uneventful. A 3-month follow-up cardiac CT showed aneurysm shrinkage associated with partial thrombus formation (*Figure 4C*), and 1-year follow-up cardiac CT revealed that the aneurysm had become completely occluded, suggesting that aneurysm rupture had been successfully prevented by a transcatheter approach (*Figure 4D*).

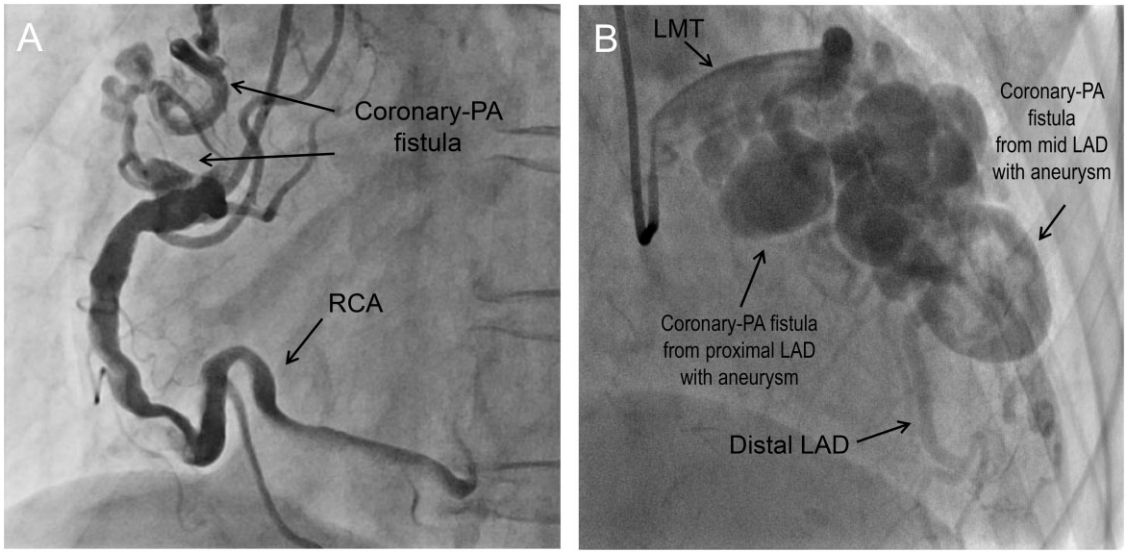
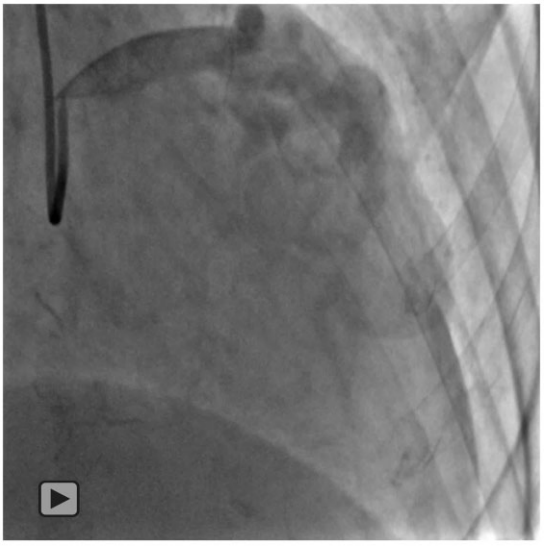
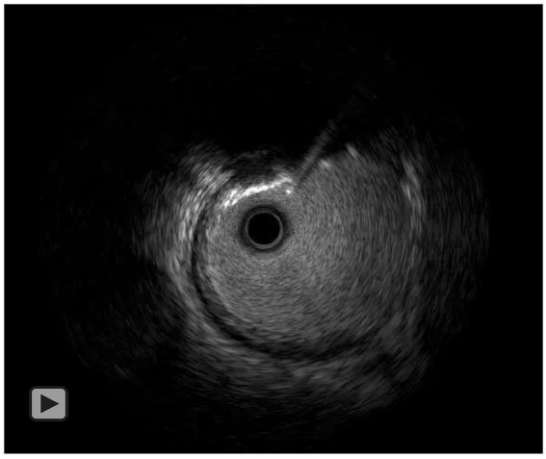


Figure 2 Coronary angiography. (*A*) Right coronary angiography showed two coronary-to-pulmonary artery fistulas originating from the proximal right coronary artery. (*B*) Left coronary angiography could not identify the orifices of the fistulas from the coronary artery.



Video 2 Left coronary angiography.



Video 3 Intravascular ultrasound findings of aneurysm anastomosis before treatment.

Discussion

We reported a case with multiple CPAFs with giant CA, to our knowledge, this is the first report of successful percutaneous coil embolization and stent implantation for multiple CPAFs with giant CA.

The appropriate therapeutic indication for CPAFs has not been established.[5] According to previous reports, the therapeutic indications depend on the size of the fistula, the presence of symptoms suggestive of myocardial ischaemia and heart failure, the size of aneurysms, anatomy of the fistula, patient's age, and the presence of associated cardiovascular abnormalities.[6] According to a previous case series in Japan, among 23 cases of coronary artery aneurysm rupture, 96% (22/23) of the patients had an aneurysm diameter of 3 cm or larger.[7] Hence, it is considered that therapeutic intervention may be indicated for aneurysms larger than 30 mm to prevent rupture. Moreover, some cases have reported that an aneurysm had progressively enlarged during follow-up period.[8,9] In our case, cardiac CT suggested that the saccular aneurysm had significantly enlarged to only 4 years (*Figure 4*), indicating the risk of rupture. Although our patient had neither clinical symptoms, including myocardial ischaemia, nor complications related to CPAF and CA, the size of the

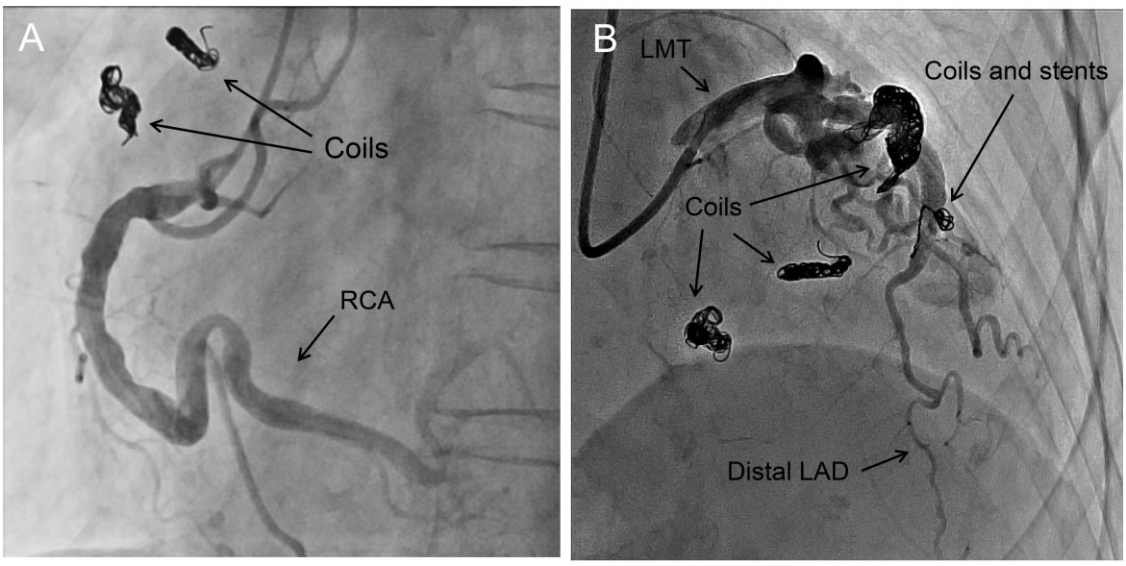


Figure 3 Coronary angiography. (*A*) Post-coil-embolization for coronary-to-pulmonary artery fistulas of the right coronary artery. (*B*) Post-coil-embolization for coronary-to-pulmonary artery fistulas of the proximal left anterior descending artery, and post coil-embolization and stent implantation for giant CA of the mid-left anterior descending artery.

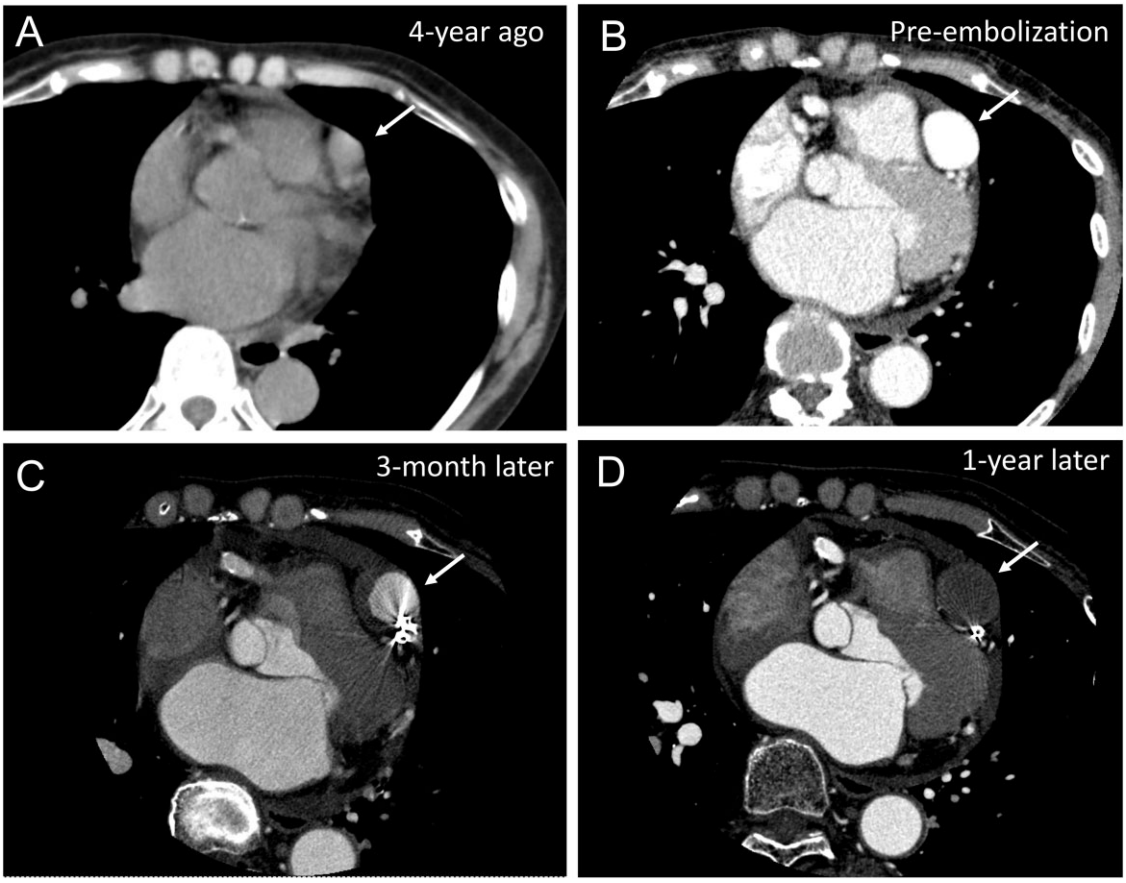


Figure 4 Serial cardiac computed tomography. Serial cardiac computed tomography findings (*A*) 4 years ago, (*B*) pre-embolization, (*C*) 3 months later, and (*D*) 1 year later. White arrows showed saccular aneurysm originating from the mid-left anterior descending artery.

aneurysms and their expansion supported the indication for interventional treatment in this case.

The management strategies for CPAFs with CA include surgical repair or catheter embolization.[5,6] In our case, the findings of IVUS revealed the anatomy of the aneurysmal orifice and aided us to determine a therapeutic strategy. After the heart team discussions, we decided to perform transcatheter closure for these CPAFs and CA using detachable coils and coronary stents. We applied DESs rather than a covered stent because a high restenosis rate and prolonged requirement of dual antiplatelet therapy (DAPT) are remaining concerns in the implantation of covered stents. Indeed, although we planned 3-month DAPT in this case, we de-escalated from DAPT with aspirin and clopidogrel to a SAPT with clopidogrel, due to frequent nasal bleeding 1 month after stent implantation.

In conclusion, multiple CPAFs with giant CA were successfully treated using percutaneous coil embolization and stent implantation. A transcatheter approach for multiple CPAFs is a promising therapeutic option with advantages over other approaches in safety and invasiveness.

Lead author biography

Yasuhiro Nakano was born on 2 October 1981. In 2006, he graduated from Saga University. From 2006 to 2010, he became a junior and senior resident in Saiseikai Fukuoka General Hospital. From 2010 to 2014, he conducted research on myocardial reperfusion injury at Kyushu University Graduate School of Medical Sciences. After working at Saga-ken medical center Koseikan and Kusatsu Heart Center, he is an interventional cardiology in Kyushu University Hospital since 2018.

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 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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