

Giant coronary fistula aneurysm presenting as a progressing left-sided asymptomatic mediastinal mass with systolic dominant Doppler flow: a case report

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Received 10 June 2021; first decision 10 September 2021; accepted 20 December 2021; online publish-ahead-of-print 9 January 2022

Background

Cases of giant coronary artery aneurysms (GCAAs) associated with coronary fistula are rarely reported, and they present with various symptoms, including coronary steal syndrome. We report an uncommon case of an asymptomatic giant coronary fistula aneurysm presenting as a progressing left-sided mediastinal mass that has been tracked for years.

Case summary

A 67-year-old healthy asymptomatic woman was referred to our hospital because of an abnormal shadow on her chest radiography revealing a left-sided mediastinal mass that had progressed in size over the past 4 years. Computed tomography revealed mass progression from 4 to 5 cm in diameter within 2 years. Coronary computed tomography and coronary angiography identified a GCAAs in a coronary fistula originating in the left anterior descending artery and draining into the main pulmonary artery. Transthoracic Doppler echocardiography revealed a unique systolic dominant flow. She underwent coronary artery aneurysmectomy and fistula ligation. The patient has been in good health without any events for 10 months since her discharge.

Discussion

A GCAAs in a coronary fistula can present as an asymptomatic left-sided mediastinal mass that has progressed in size for years in older adults. Echocardiography can provide clues of the steal phenomenon in coronary artery fistula. A close investigation of mediastinal abnormalities can facilitate the detection of coronary aneurysms.

Keywords

Giant coronary artery aneurysm • Coronary fistula • Mediastinal mass • Transthoracic Doppler echocardiography • Case report

ESC Curriculum

2.2 Echocardiography • 2.4 Cardiac computed tomography • 7.5 Cardiac surgery • 3.1 Coronary artery disease

Learning points

- A giant coronary fistula aneurysm can present as an asymptomatic left-sided mediastinal tumour that has progressed in size over time.
- Aneurysms in the coronary fistula can progress abruptly in older adults.
- Echocardiography can provide clues of the steal phenomenon in coronary artery fistula.

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Handling Editor: Luke Joseph Laffin

Peer-reviewers: Vincenzo Nuzzi; Golnaz Houshmand

Compliance Editor: Linh Ngo

Supplementary Material Editor: Fabienne Vervaat

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Introduction

Cases of giant coronary artery aneurysms associated with coronary fistula (GCAA-F) are rarely reported.¹ Patients with GCAA-F present with various symptoms and complications, including coronary steal syndrome, which may be difficult to diagnose. Very few cases of asymptomatic giant coronary aneurysm with fistula have been reported to date, and the progressive history has rarely been followed up.

Here, we report a case of asymptomatic GCAA-F presenting as a progressing left-sided mediastinal mass that was tracked for years.

Timeline

Date	Events
July 2013	Normal chest radiography
August 2018	Protrusion of left 3rd arch in chest radiography
October 2018	Plain chest computed tomography (CT): 4-cm mass
August 2020	Abnormal mass in chest radiogram
October 2020	Admission Transthoracic echocardiography and coronary CT angiography identified a giant coronary fistula aneurysm Coronary angiography confirmed the diagnosis
December 2020	Transfer to the corresponding surgery centre Coronary artery aneurysmectomy and coronary artery fistula ligation Discharge
October 2021	In good health without any events

Case presentation

A 67-year-old healthy woman was referred to the division of thoracic surgery at our hospital for an abnormal shadow detected during a regular annual medical check-up. The abnormal shadow was not apparent on the chest radiogram performed 7 years ago before this visit (*Figure 1*). A series of previous chest radiograms showed protrusion of the left 3rd arch that had progressed in size over the past 4 years. Computed tomography (CT) image taken 2 years prior to this visit for close examination of the protrusion showed a 4-cm mass (*Figure 2A and B*), although it was not noted by the physicians. She did not experience angina, either at rest or with exertion, or other symptoms. Her medical history was unremarkable from a cardiovascular perspective. Computed tomography revealed a mass ~5 cm in size in the mediastinum bordering the left side of the pulmonary artery, suspicious for lymphoma, neurogenic tumour, or germ cell tumour. Positron emission tomography-CT demonstrated negative uptake of 18F-FDG. The inside of the mass showed contrast enhancement equivalent to that of a blood vessel and thrombus formation, suggesting an aneurysm. The patient was referred to the Division of Cardiology.

The physical examination results were normal. No abnormalities were observed in the blood tests, e.g. troponin I, 3.3 pg/mL (normal <70 pg/mL). Transthoracic echocardiography revealed a large mass connecting to the left anterior descending artery (LAD). Blood flow into the mass cavity was detected, suggesting a coronary artery aneurysm. Coronary flow velocity by transthoracic Doppler echocardiography demonstrated a dominant systolic component at the left main coronary trunk proximal to the fistula and inside the mass, suggesting a steal phenomenon. (*Figure 3A*). The left ventricular wall motion was normal, and the ejection fraction was estimated to be 72%.

A 64-slice coronary CT angiography revealed a giant coronary aneurysm with coronary fistula (48 × 45 × 55 mm), with internal homogeneous contrast enhancement on arterial phase and thrombus formation, connected to the mid-portion of the LAD (*Figure 2C–F*). The aneurysm branched into a tortuous vessel connected to the main pulmonary artery. Another fistula from the proximal portion of the right coronary artery (RCA) drained into the left atrium and the main pulmonary artery.

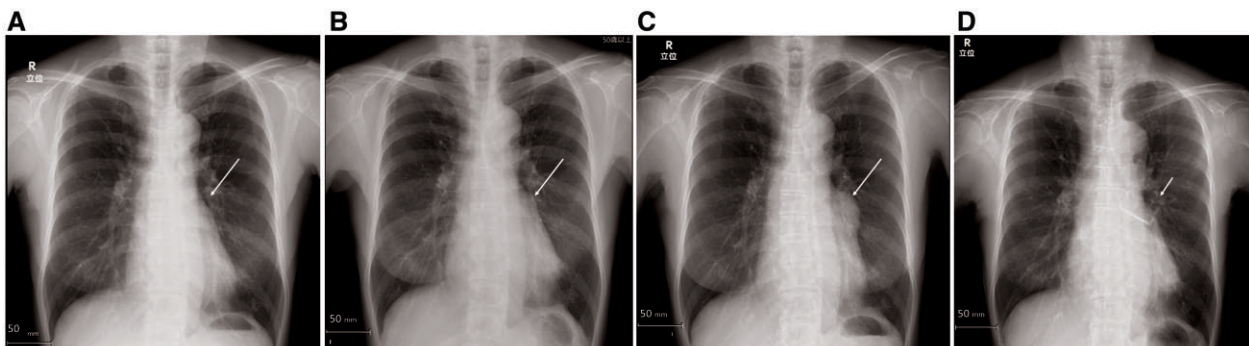


Figure 1 A series of chest radiograms performed (A) 7 years ago before this visit, (B) 4 years earlier, (C) on admission, and (D) after surgery. Arrows indicate the progression of the mass.

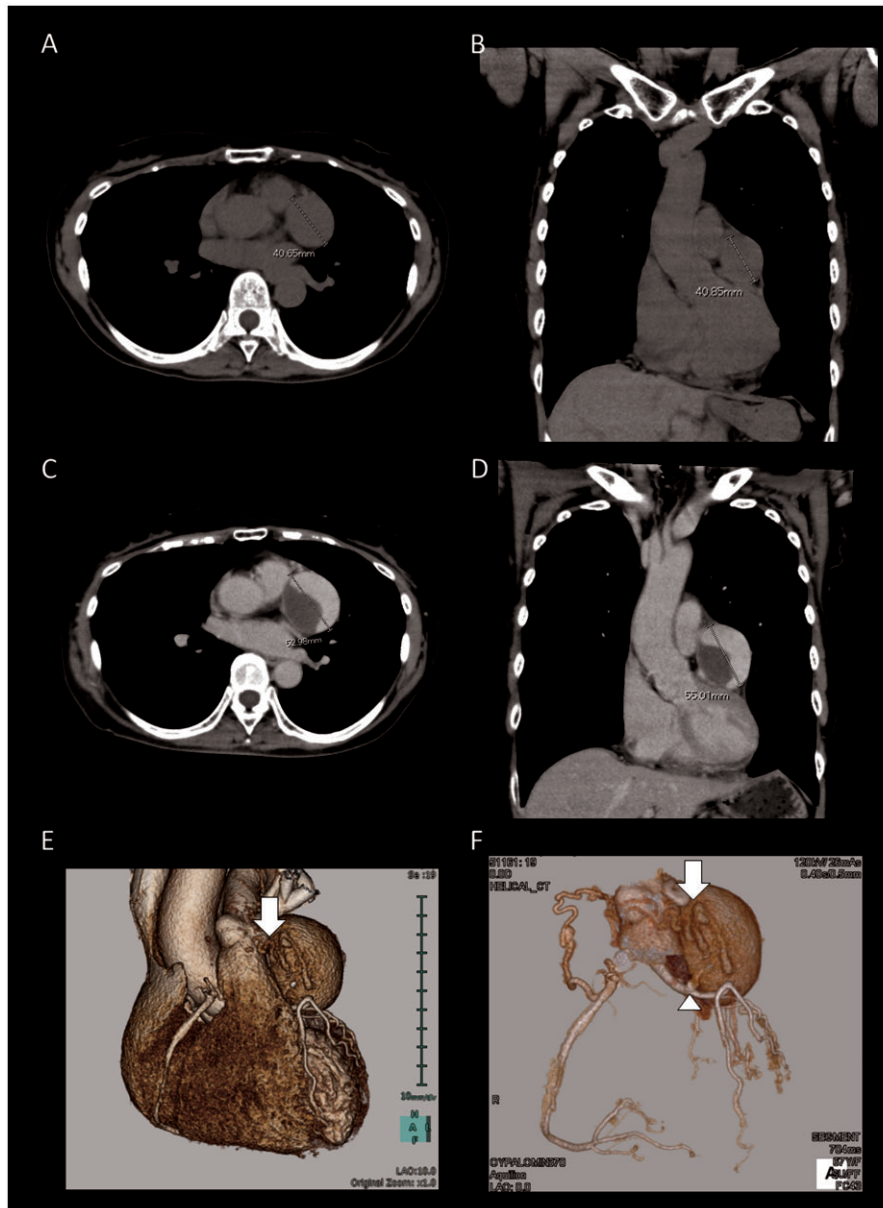


Figure 2 (A, B) Unenhanced computed tomography scan taken 2 years earlier showing a 4-cm mass. (C, D) Contrast-enhanced computed tomography angiography revealing a giant coronary aneurysm ($48 \times 45 \times 55$ mm) with internal contrast enhancement and thrombus formation. (E, F) Three-dimensional heart reconstruction of computed tomography images. Location of the aneurysm in the fistula originating at the mid-portion of left anterior descending artery (F arrowhead). The aneurysm branched into a tortuous vessel that connected to the main pulmonary artery (E, F, arrow). Another fistula from the proximal portion of the right coronary artery drained into the left atrium and the main pulmonary artery (F).

Adenosine triphosphate stress myocardial perfusion scintigraphy identified reversible perfusion defects in the anterior wall, representing myocardial ischaemia in the territory of the LAD (Figure 3B). Conventional coronary angiography confirmed a giant aneurysm originating from the LAD, to which blood flow was supplied during systole (Figure 4A). The aneurysm outflow was obscured (Figure 4B) due to the dilution of the contrast dye by the large aneurysmal sac. Other anomalous vessels started from the proximal portion of the

RCA (Figure 4C), as shown by CT. No occlusive atherosclerotic lesions were observed. Mean pulmonary artery wedge pressure and pulmonary artery pressure were measured to be 9 and 23/9/14 mmHg (systolic/diastolic/mean), respectively. The Qp/Qs ratio was calculated to be 1.2.

Because of the risk of rupture due to progressive enlargement of the aneurysm and the presence of ischaemia, the patient was immediately transferred to the cardiac surgery department

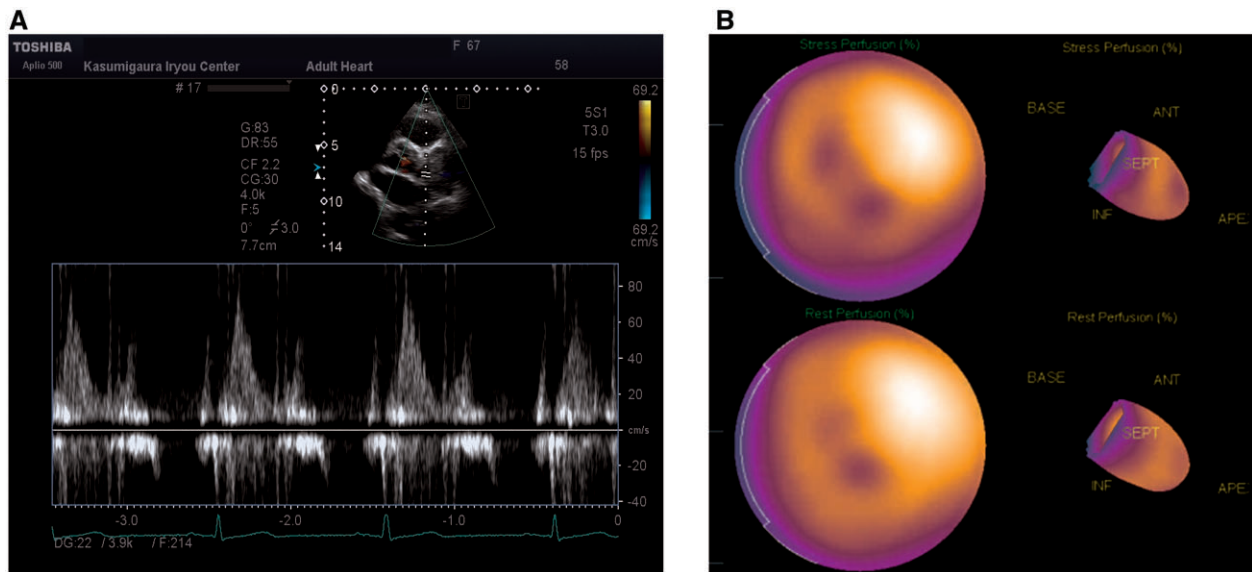


Figure 3 (A) Coronary flow velocity by Doppler echocardiography demonstrated a dominant systolic component at the left main coronary trunk. (B) Polar mapping of adenosine triphosphate stress myocardial perfusion scintigraphy identified reversible perfusion defects in the anterior wall.

without receiving additional medical treatment other than bisoprolol. The patient underwent elective surgery via median sternotomy and cardiopulmonary bypass. The fistula vessel between the proximal RCA and the main pulmonary artery was ligated. A 50-mm aneurysm was identified in the fistula vessel originating in the LAD and incised; a mural thrombus largely occupied it. Inflow path from the LAD was ligated. Two outflow vessels were identified and ligated; one connected to the main pulmonary artery, the other had not been detected during preoperative imaging tests. The aneurysm was resected, and subsequently, a left atrial appendage closure was performed due to paroxysmal atrial fibrillation detected during her hospitalization. The patient recovered uneventfully, except for a transient atrial fibrillation, and was discharged from the hospital. Edoxaban and bisoprolol were administered for 3 and 7 months, respectively. The patient has been in good health, and no cardiovascular events were reported for 10 months since her discharge.

Discussion

Here, we report an uncommon case of GCAA-F, and the course of this patient's disease provides two important clinical suggestions. First, a GCAA-F can present as an asymptomatic left-sided mediastinal mass that progresses in size for years in an elderly patient. Second, the steal phenomenon can be evaluated using transthoracic Doppler echocardiography.

Although no consensus definition exists, coronary artery aneurysm having diameters exceeding 20–50 mm is considered 'giant'. Coronary steal may cause aneurysms to develop²; the association with fistula was estimated to be 25% in giant coronary artery

aneurysms (GCAAs).^{1,2} Our patient had no history of known underlying disease for coronary artery aneurysm other than coronary artery fistula, which is considered congenital. The aneurysm progressed abruptly and asymptotically as the patient aged.

Most patients with coronary artery fistula are asymptomatic; however, symptoms develop depending on the pressure in its terminal chamber, the volume of the shunt, or the presence of coronary steal phenomenon via the fistulas.³ Symptoms may include angina, exertional dyspnoea, syncope and palpitations, associated with myocardial ischaemia, infarction, congestive heart failure, and cardiac arrhythmias. Giant coronary artery aneurysms are reported to compress adjacent structures and present with superior vena cava syndrome² and angina⁴ symptoms. As summarized in Table 1, only 18 asymptomatic GCAA-F patients with a maximum diameter >50 mm, including our patient, have been reported to date.

The differential diagnosis of a para-cardiac mass includes pericardial cysts, germinal cell neoplasms, thymic tumours, aneurysm of the cardiac wall, as well as coronary artery aneurysm. Patients with GCAA can also present with a mass, which can be misdiagnosed as a mediastinal tumour. The RCA is the most commonly affected in coronary artery aneurysm,⁵ and coronary artery fistula.⁶ However, most asymptomatic GCAA-F cases originate in the left coronary artery and present as left-sided mediastinal masses on chest radiogram (Table 1), making diagnosis challenging.

Since interventional treatment is performed in most cases and the progressive history is rarely seen, it is usually difficult to discern whether a GCAA was present at birth or was acquired later. Only one case of GCAA-F presenting with exertional dyspnoea and angina was reported to have been followed up by CT, in which the size had increased from 47.5 to 55 mm within 8 months.⁷ In the present case, the growth was tracked for 4 years using chest radiography, and CT showed that the

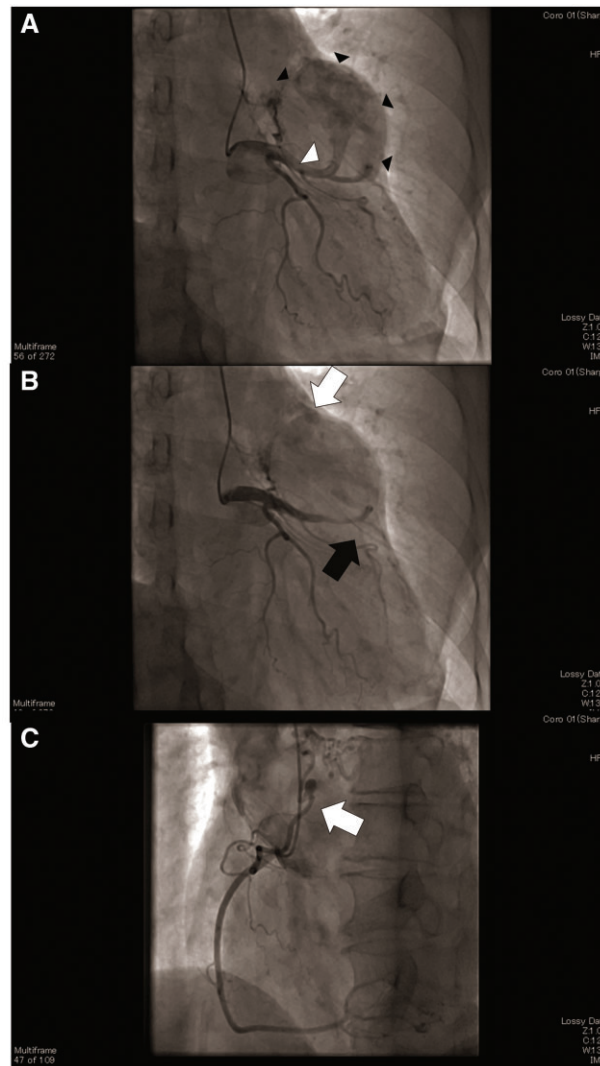


Figure 4 (A) Left coronary angiogram in the systolic phase demonstrated inflow of contrast (white arrowhead) from fistula originating from the left descending artery filling aneurysm (black arrowhead) with defect by thrombus. RAO 30° + CAUD 25°. (B) Left coronary angiogram in the diastolic phase following 'A' demonstrated distal left descending artery enhanced by contrast (black arrow). Aneurysm outflow (white arrow) towards the pulmonary artery was obscured. (C) Right coronary angiogram demonstrated coronary artery fistula draining into the main pulmonary artery (white arrow). LAO 45°.

size had increased from 40 to 55 mm within the last 2 years, suggesting that aneurysms can progress abruptly in older adults.

Transthoracic Doppler echocardiography can be used to evaluate the steal phenomenon. Okeie *et al.*⁸ reported typical findings of reversible myocardial ischaemia due to the coronary steal phenomenon in only two of seven patients with coronary artery pulmonary artery fistula. Hori *et al.*⁹ investigated coronary blood flow velocity in patients with coronary fistula aneurysm originating from the LAD to the pulmonary artery using the Doppler guidewire technique. The coronary flow pattern showed a prominent systolic component at the LAD site proximal to the fistula, suggesting that coronary steal affects coronary flow dynamics. Coronary flow velocity can be

clinically demonstrated using transthoracic Doppler echocardiography; the normal pattern is biphasic with a large diastolic predominance.¹⁰ The coronary flow pattern of our patient demonstrated a dominant systolic component at the left main coronary trunk proximal to the fistula and the aneurysm. She was asymptomatic, although myocardial ischaemia in the corresponding area was identified using myocardial perfusion scintigraphy. Echocardiography provides information regarding the morphological features of coronary artery aneurysms and fistulas, as well as further information regarding functional evaluation.

To our knowledge, this is the first report of asymptomatic GCAA-F presenting as a progressing left-sided mediastinal mass that was

Table 1 Reported cases with asymptomatic cases of GCAA-F with a maximum diameter >50 mm

Author	Year	Age	Sex	Max diameter (mm)	Originating site	Draining site	Examination detecting mass	Mediastinal mass	Intervention	Outcome	Journal
Okita	1992	61	F	50	LAD	PA	Chest radiography	Left	Surgery	Uneventful	Ann Thorac Surg; 54(4)
Abou Eid	1993	74	M	100	RCA	RA	Echocardiography	N/A	Surgery	Uneventful	Thorac Surg; 56(2)
Lee	1997	63	F	52	LAD	PA	Chest radiography	Left	NR	NR	Circulation; 95(8)
Mawatari	2000	56	F	70	RCA	RA	Chest radiography	Right	Surgery	Uneventful	Ann Thorac Surg; 70(4)
Niimura	2002	77	F	52	LAD	PA	Chest radiography	Left	Surgery	Doing well	N Engl J Med; 346(16)
Yamawaki	2006	71	M	88	LCx	LV	Chest radiography	Left	Surgery	Uncomplicated	Heart; 92(11)
Sawai	2008	35	F	80	RCA	LA	Echocardiography	N/A	Surgery	Uneventful	Anesth Analg; 106(4)
Branco	2008	66	F	60	Septal br	NR	Echocardiography	N/A	Medication	One episode of deterioration	Eur J Echocardiogr; 9(1)
Wei	2011	26	M	58	RCA	LV	CT	N/A	Surgery	No complaints	Intern Med; 50(3)
Ipek	2012	25	F	65	RCA	RA	CT	N/A	Surgery	NR	Tex Heart Inst J; 39(3)
Okamoto	2013	68	F	62	LAD	PA	Chest radiography	Left	Surgery	No symptoms	Intern Med; 52(2)
Li	2016	66	F	74	LAD	PA	CT	N/A	Surgery	No complications	Medicine; 95(46)
Rossi	2017	41	M	70	RCA	RA	Echocardiography	N/A	Clinical follow-up	NR	J Cardiovasc Med; 18(9)
Seo	2018	85	F	50	D1	PA	Chest radiography	Left	Percutaneous coil-embolization	No complications	Chin Med J; 131(24)
Suzuki	2018	79	M	71	LAD	PA	Chest radiography	Right	Surgery	Uneventful	Heart Surg Forum; 21(4)
Ren	2019	55	M	65	RCA	RA	Echocardiography	N/A	Surgery	Favourable	BMC Surg; 19(1)
Harada	2020	65	F	58	LMT	RA	Echocardiography	N/A	Surgery	In good health	Cureus; 12(3)
Maruyama	2021	67	F	55	LAD	PA	Chest radiography	Left	Surgery	No cardiovascular events	

CT, computed tomography; GCAA-F, giant coronary artery aneurysms associated with coronary fistula; LA, left atrium; LAD, left anterior descending artery; LCA, left coronary artery; LCX, left circumflex artery; LMT, left main coronary trunk; LV, left ventricle; N/A, not applicable; NR, not reported; PA, pulmonary artery; RA, right atrium; RCA, right coronary artery; Sep Br, septal branch.

tracked for years. Steal phenomenon was evaluated using transthoracic Doppler echocardiography. A close investigation of the mediastinal abnormality can facilitate the detection of coronary aneurysms.

Lead author biography



Hidekazu Maruyama was born in Wakayama in 1971. He graduated from the Faculty of Medicine at the University of Tsukuba in 1996 and from the Graduate School of Comprehensive Human Sciences at the University of Tsukuba in 2007. He is currently working as a cardiologist at the National Hospital Organization Kasumigaura Medical Center, Tsuchiura, Japan.

Supplementary material

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

Acknowledgements

The authors would like to express their appreciation to Department of Cardiovascular Surgery of University of Tsukuba, the radiologic technologists of the National Hospital Organization Kasumigaura Medical Center, and Masao Nishimura.

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.

Funding: None declared.

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