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

Hidekazu Maruyama 1*, Kumiko Habe2, Jo Kato1, and Makiko Nishikii1

1Department of Cardiology, National Hospital Organization Kasumigaura Medical Center, 2-7-14 Shimotakatsu, Tsuchiura 300-8585, Japan; and 2Department of Clinical Laboratory, National Hospital Organization Kasumigaura Medical Center, 2-7-14 Shimotakatsu, Tsuchiura 300-8585, Japan

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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 GCAA 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 GCAA 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.

* Corresponding author. Tel: +81 29 822 5050, Fax: +81 29 824 0494, Email: maruyama.hidekazu.mj@alumni.tsukuba.ac.jp

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Introduction

Cases of giant coronary artery aneurysms associated with coronary fistula (GCAA-F) are rarely reported. 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                                                             |
| December 2020| Transthoracic echocardiography and coronary CT angiography identified a giant coronary fistula aneurysm |
|            | Coronary angiography confirmed the diagnosis                            |
|            | Transfer to the corresponding surgery centre                           |
|            | Coronary artery aneurysmectomy and coronary artery fistula ligation     |
| 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 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.
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 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 × 45 × 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).
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 develop; the association with fistula was estimated to be 25% in giant coronary artery aneurysms (GCAAs). 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. 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 syndrome and angina 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 and coronary artery fistula. 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. In the present case, the growth was tracked for 4 years using chest radiography, and CT showed that the
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 Echocardiog; 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            | Chest radiography           | 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   | Cereus; 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 trans-thoracic 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.

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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.

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