Diagnosis and Endovascular Treatment of Coronary Artery Fistula: A Case of Symptomatic Left-to-Right Shunt

Tien En Edward Ong*, Justin Kwan, Sundeep Punamiya

Department of Diagnostic Radiology, Tan Tock Seng Hospital, Singapore

***Correspondence:** Tien En Edward Ong, Tan Tock Seng Hospital, 11 Jln Tan Tock Seng, Tan Tock Seng Hospital, Singapore 308433

edward.ong.tien.en@gmail.com

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Authors' Contributions
Tien En Edward Ong – Primary author, reporting radiologist
Justin Kwan – Supervisor, interventional radiologist (involved in patient treatment)
Sundeep Punamiya – Supervisor, interventional radiologist (involved in patient treatment)

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Consent
Yes

Human And Animal Rights
No experiments on human or animal subjects were carried out

ABSTRACT

Coronary artery fistulas (CAFs) are rare vascular malformations which constitute abnormal communications from coronary arteries to cardiac chambers, or to parts of the systemic or pulmonary circulation. While small CAFs are usually asymptomatic and often resolve spontaneously, medium or large CAFs can result in hemodynamically significant shunting with progressive dilation. We report a case of CAF originating from the right coronary artery and draining into the coronary sinus, which resulted in pulmonary hypertension and heart failure. The patient subsequently underwent percutaneous flow reduction with vascular plug and coils, leading to significant reduction of shunt flow and clinical improvement. CT and procedural fluoroscopic images are provided.

CASE REPORT

BACKGROUND

Coronary artery fistulas (CAFs) are rare vascular anomalies that can have significant clinical consequences when they result in substantial left-to-right shunting with eventual progression to pulmonary hypertension and heart failure. Traditionally managed with surgical ligation, advances in imaging and interventional techniques have paved the way for less invasive percutaneous approaches, particularly valuable in patients with high surgical risk. This case of an elderly patient with symptomatic CAF (draining into the coronary sinus) underscores the critical role of multimodality imaging in diagnosis and treatment planning. Moreover, it contributes to the growing body of literature by detailing a successful endovascular procedure, highlighting its feasibility and potential benefits in managing complex coronary anomalies.

CASE REPORT

A 73-year-old female presented with a 3-day history of progressive shortness of breath, orthopnea, and lower limb swelling. Her relevant cardiac history included atrial fibrillation and idiopathic pulmonary hypertension.

Imaging findings

A CT pulmonary angiogram was initially performed to rule out pulmonary embolism as a cause of her acute deterioration. No pulmonary embolism was detected, however there was an incidental finding of dilated right coronary artery (RCA) with likely fistulous connection to the right atrium (Figure 1). Ancillary features of pulmonary hypertension and right heart strain were also present, in the form of dilated pulmonary trunk

(4.3 cm diameter), right heart enlargement and inferior vena caval reflux of contrast.

Further evaluation with ECG-gated CT coronary angiography (CTCA) confirmed that the dilated and tortuous RCA (1.7 cm diameter) drained into an aneurysmal distal coronary sinus (3.5 x 2.9 cm) and thence into the right atrium (Figure 2).

Cardiac catheterisation

Right heart catheterization showed elevated right atrial pressure (8 mmHg) and mean pulmonary arterial pressure (25 mmHg), confirming the presence of pulmonary artery hypertension. Mixed venous sampling revealed a high venous oxygen saturation (SvO_2) of 86%, in keeping with a significant left-to-right shunt.

Management/Follow-up

Surgical and endovascular options were considered; due to the patient's multiple comorbidities and high surgical risk, a percutaneous transcatheter approach was chosen. Arterial and venous access was obtained (via the right common femoral artery, right common femoral vein and left subclavian vein), and retrograde contrast injection via the distal end of the fistula confirmed the presence of a high-flow shunt (Figure 3a). Both arterial and venous sides of the fistula were cannulated to provide better guidewire support. An 18 mm Amplatzer Vascular Plug (AVP II, Abbott) was deployed from the venous side in the mid-distal fistula (Figure 3b). Additionally, multiple detachable coils of various diameters (Concerto, Medtronic) were deployed from the arterial side to further retard flow (Figure 3c). Angiography demonstrated substantially reduced flow through the fistula, with preserved flow in the native RCA (Figure 3d). At this juncture, the coil mass was found to be nearing a normal branch of the RCA and the patient was clinically in volume overload; the procedure was concluded, with a view to further embolization if there was no clinical improvement.

Following the procedure, the patient showed marked clinical improvement, with reduced features of volume overload and increased functional capacity. She was discharged 1 month post procedure and continued outpatient follow-up with Cardiology. 6 months of anticoagulation with Warfarin was commenced to ameliorate the risk of thrombus propagation in the event of delayed closure. At 1 year after discharge, the patient reported continued improvement in effort tolerance, lower limb edema and orthopnea. Unfortunately, further evaluation with repeat CT coronary angiography was declined.

DISCUSSION

Etiology & Demographics

Coronary artery fistulas (CAFs) are uncommon abnormal communications between a coronary artery and a cardiac chamber or another vascular structure. The majority of CAFs are congenital, and are thought to result from persistent embryological vessels (myocardial sinusoids) which perfuse the primitive tubular heart [1]. CAF can also be acquired following trauma, iatrogenic injury, or vasculitic insult [2]. In cases where a sufficiently large CAF drains into the right side of the heart, a left-to-right shunt can develop and enlarge, leading to volume overload. Over time, increased preload and pulmonary blood flow may result in pulmonary hypertension and right heart failure [3]. Additionally, a coronary "steal" phenomenon can develop, potentially leading to ischemia.

CAFs overall account for a small fraction of congenital cardiac anomalies, with an incidence of approximately 0.1–0.2% in the general population [4–8]. CAFs demonstrate no strong gender, race or age predilection [9].

Clinical & Imaging Findings

Patients with small CAFs are usually asymptomatic, while larger CAFs may initially present as an unexplained continuous murmur or electrocardiographic abnormality [10,11]. In cases where a hemodynamically significant shunt has developed, symptoms may include dyspnea, volume overload and myocardial infarction [12].

On imaging, CAFs can be classified based on size, origin, drainage site, complexity and accompanying cardiac anomaly [13]. The most common origin vessel is the right coronary artery (50%–55%), followed by the left anterior descending artery (35%–40%) [14,15]. The most common drainage site is a cardiac chamber (coronary-cameral fistula), with the right heart chambers being the most common drainage site [16–18].

CT angiography has emerged as the workhorse modality for evaluation of CAFs, offering rapid image acquisition and high spatial resolution with a non-invasive technique. Multiplanar reconstructions provide excellent anatomical detail, which is invaluable for treatment planning [19]. Conventional catheter coronary angiography remains the imaging modality of choice during endovascular intervention; however it carries procedural risks, and complex anatomy may be poorly appreciated on two-dimensional imaging [20].

Treatment & Prognosis

While small and asymptomatic CAFs may be monitored clinically and with echocardiography, intervention is indicated in patients with large fistulas or symptomatic fistulas of any size [21]. The primary treatment goal is to halt or significantly reduce flow through the CAF.

Surgical closure of the fistula may involve simple ligation, or may be accompanied by coronary artery bypass graft placement [22]. Surgical closure is favored in cases of fistulas with multiple communications, highly tortuous vessels, or for patients who require surgical management of concomitant cardiac abnormalities. Complications include postoperative myocardial infarction and incomplete closure of the fistula [23].

Percutaneous transcatheter closure has emerged as a valuable alternative to surgery, particularly in cases of high perioperative risk (as was the case in this patient with multiple comorbidities). CAFs with a single drainage site and absence of concomitant cardiac abnormalities are more amenable to transcatheter closure. A transarterial approach (cannulation of the fistula via its feeder artery) may be favored in fistulas originating from the proximal part of a coronary artery, while a transvenous approach (cannulation of the fistula via its drainage pathway) may be chosen in fistulas originating from the distal part of the coronary vessel. In this technically challenging case, a combined arterial and venous approach was used to cannulate the fistula from either end, providing enhanced guidewire support and allowing for precise closure device deployment. This dual-access strategy has been described as a technique for improving device control in particularly tortuous fistulas [24]. After access is secured, endovascular occlusion devices may be deployed to halt or slow flow. Vascular plugs are expanding braided nitinol mesh constructs which are generally available a wide variety of diameters, suitable for closing large fistulas. Embolization coils are particularly useful in occluding tortuous vessels due to their conformity to vessel shape; they can also be deployed via low-profile catheters. In cases of complex CAFs, covered stents may also be used to exclude side-branches. Procedural risks of endovascular CAF include vessel dissection and rupture, migration of the closure device and fistula recanalization [14,24,25]. Procedural success rates are estimated to range from 81-100% [26–28].

TEACHING POINT

Coronary artery fistulas (CAFs) are aberrant coronary arterial communications which drain into a cardiac chamber or systemic/pulmonary vessel; if left uncorrected, a haemodynamically significant left-right shunt may result in pulmonary hypertension and heart failure. Large or symptomatic CAFs can be treated with surgical or transcatheter closure, depending on fistula anatomy and patient comorbidities.

QUESTIONS

Question 1: Which imaging technique has become the noninvasive modality of choice for the evaluation of coronary artery fistulas?

A) Transthoracic echocardiogram
B) Transesophageal echocardiogram
C) Cardiac MRI
D) CT coronary angiography (applies)
E) Conventional catheter angiography

Explanation for question 1: CT angiography has emerged as the workhorse modality for evaluation of CAFs [offering rapid image acquisition and high spatial resolution with a non-invasive technique]

Question 2: Which of the following hemodynamic effects is frequently associated with large coronary artery fistulas?

A) Left ventricular outflow tract obstruction
B) Right-sided volume overload and pulmonary hypertension (applies)
C) Acute myocardial rupture
D) Complete heart block
E) Coronary artery dissection

Explanation for question 2: In cases where the CAF drains into the right side of the heart, [increased preload and pulmonary blood flow may result in pulmonary hypertension and right heart failure]

Question 3: Which factor most strongly influences whether surgical ligation or percutaneous closure is pursued for CAFs?

A) Patient gender
B) Presence of atrial fibrillation
C) Origin from the left circumflex artery
D) Complexity and number of fistulous tracts (applies)
E) Absence of any heart murmurs

Explanation for question 3: Surgical closure is [favored in cases of complex fistulas (with multiple communications) and highly tortuous vessels, or for patients who require surgical management of concomitant cardiac abnormalities].

Question 4: What is the proposed etiology for congenital CAFs?

A) Failure of embryonic myocardial sinusoids to regress, leading to a persistent vascular communication (applies)
B) Abnormal proliferation of coronary smooth muscle cells during development
C) Inflammatory damage to the coronary arteries during the neonatal period
D) Atherosclerotic changes in the coronary vessels occurring in utero
E) Abnormal deposition of connective tissue in the developing myocardium

Explanation for question 4: [The majority of CAFs are congenital and are thought to result from persistent embryological vessels (myocardial sinusoids) which perfuse the primitive tubular heart].

Question 5: What is the most common drainage site for CAFs?

A) Left cardiac chamber
B) Right cardiac chamber (applies)
C) Pulmonary artery
D) Coronary sinus
E) Pulmonary vein

Explanation for question 5: [The most common drainage site is a cardiac chamber (coronary-cameral fistula), with the right heart chambers being the most common drainage site (14-40% drain into the right ventricle and 19-26% into the right atrium)].

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FIGURES

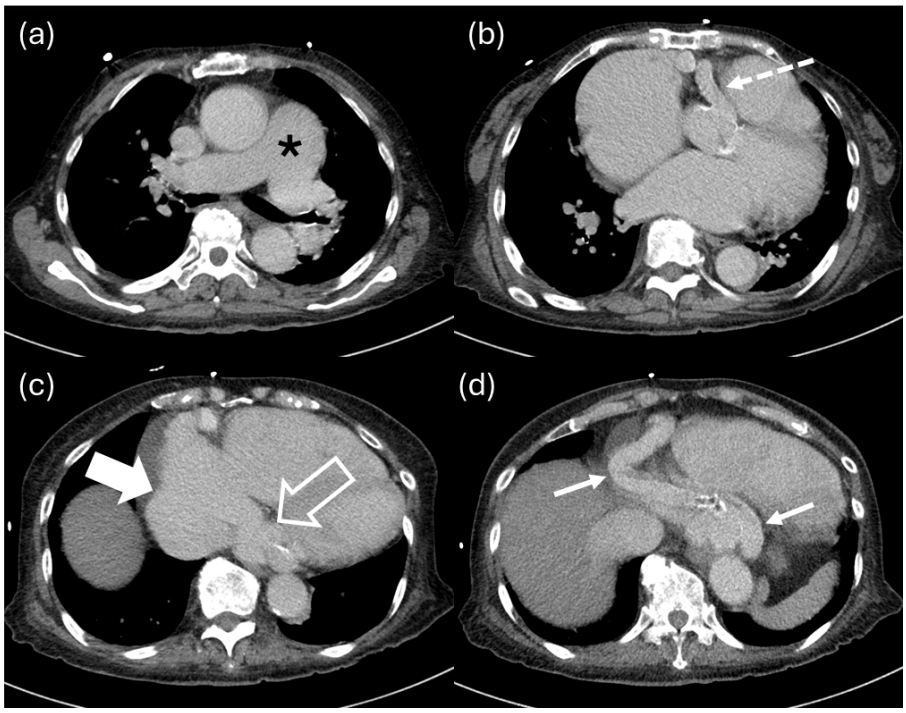

Figure 1: Selected slices from CT pulmonary angiogram – (a) Dilated pulmonary trunk measuring up to 4.3 cm in diameter (asterisk). (b) Origin of dilated RCA measuring up to 1.7 cm in diameter (dashed arrow). (c) Drainage into a dilated distal coronary sinus (hollow arrow) and enlarged right atrium (solid broad arrow). (d) Tortuous course of the RCA (arrows) and inferior vena caval reflux of contrast.
CT acquisition details: Siemens, 120kV, 154 mAs, 1 mm slice thickness, 60 ml IV Omnipaque 350

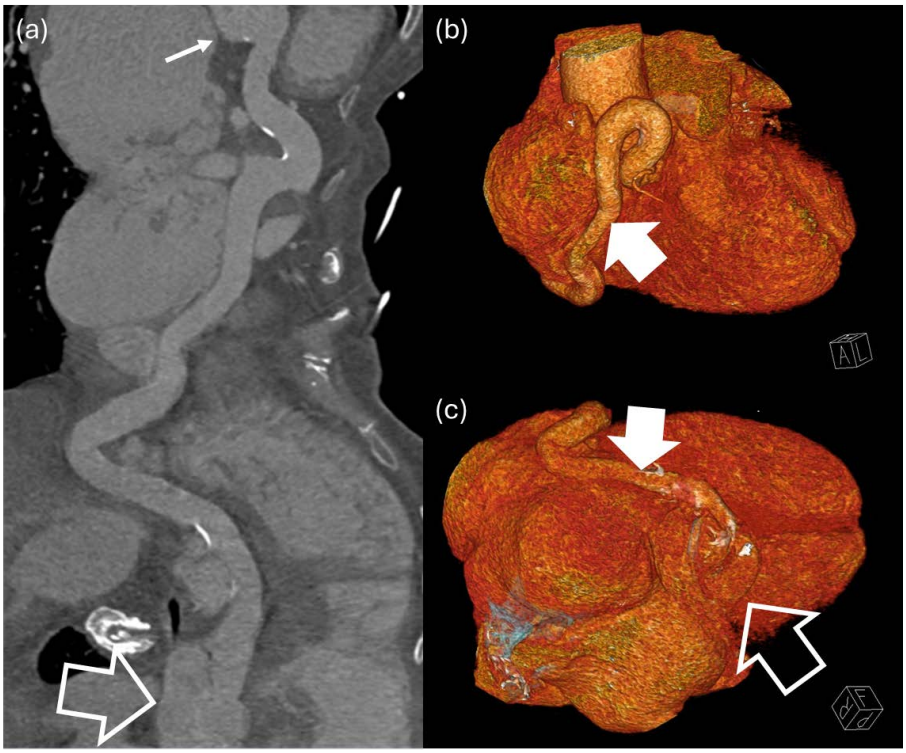

Figure 2: ECG-gated CT coronary angiogram – (a) Curved multiplanar reformat image. (b and c) Volume render. The tortuous course of the dilated RCA from the aortic root (white arrow) to its drainage into a dilated coronary sinus (hollow arrow) is demonstrated.
CT acquisition details: Siemens, 120kV, 360 mAs, ECG-gated, 60 ml IV Omnipaque 350, 10 mg IV Metoprolol

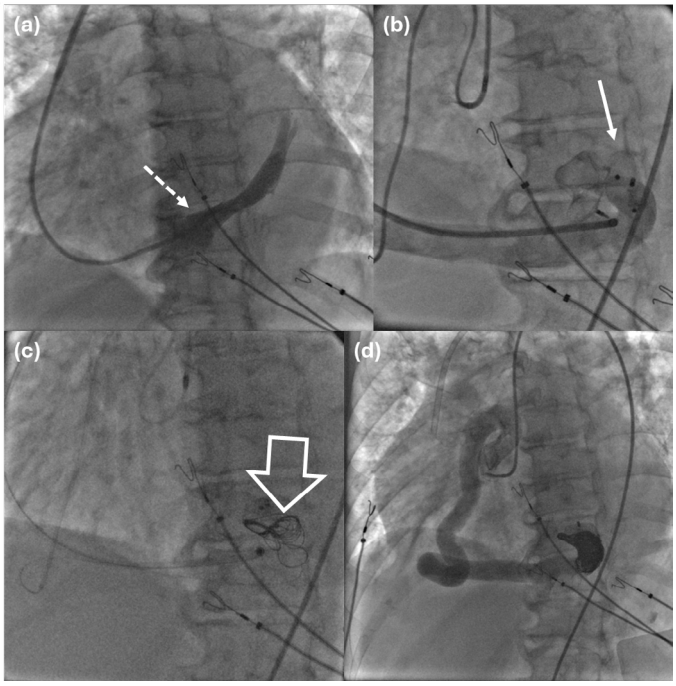

Figure 3: Fluoroscopic images acquired during endovascular flow retardation of CAF – (a) Retrograde injection of contrast via the CAF outflow demonstrating a high-flow shunt (dashed arrow). (b) Deployment of Amplatzer Vascular Plug in the distal fistula (arrow). (c) Deployment of detachable coils (hollow arrow) proximal to the vascular plug. (d) Post-treatment angiogram showing marked reduction of flow within the CAF.

Table 1: Summary table

Etiology	Congenital: Persistent primitive structures (e.g. myocardial sinusoids) Acquired: Trauma, iatrogenic injuries, or vasculitic changes
Incidence	0.1-0.2%
Gender ratio	No predilection
Age predilection	Poorly understood, diagnosed in all age groups
Risk factors	Unknown
Treatment	Surveillance if small and asymptomatic Surgical or endovascular closure if large or symptomatic
Prognosis	Variable, depending on size, anatomy and complications Risk of heart failure, pulmonary hypertension and ischemia if untreated shunt
Findings on imaging	Abnormal communication between coronary artery and cardiac chamber or systemic/pulmonary vessel

Table 2: Differential diagnoses

Differential diagnosis	**Clinical features**	**Imaging features**
Coronary artery fistula	Often asymptomatic if small May cause heart failure and pulmonary hypertension if left-right shunting occurs Continuous murmur	Abnormal communication between coronary artery and cardiac chamber or systemic/pulmonary vessel.
Coronary artery aneurysm	Associated with atherosclerosis and vasculitis e.g. Kawasaki disease Symptomatic complications (thrombosis, embolization) may present as myocardial infarction	Focal dilation of the coronary artery but no abnormal communication to a cardiac chamber or vessel; imaging shows an isolated aneurysmal segment.
Atrial septal defect	Often asymptomatic in childhood Development of symptoms in adulthood (right heart volume overload) Systolic ejection murmur with fixed splitting of S2	Deficient interatrial septum with left-right Doppler flow and associated right heart enlargement, but no abnormal coronary vessel
Ventricular septal defect	Usually small and asymptomatic In symptomatic, presents in infancy with congestive heart failure Holosystolic murmur	Deficient interventricular septum and left-right Doppler flow, but no abnormal coronary vessel
Pulmonary arteriovenous malformation	Present with hypoxemia or complications e.g. embolic stroke May have lung bruit	Pulmonary nidus with feeding pulmonary artery (rather than coronary artery) and draining pulmonary vein

KEYWORDS

Coronary artery fistula; Coronary arteriovenous malformation; CT coronary angiography; Heart Failure; Pulmonary hypertension