CASE REPORT

A case of congenital coronary-pulmonary fistulas presenting as spontaneous mediastinal haemorrhage

Guramrinder Singh Thind,¹ Yashwant Agrawal,¹ Jagadeesh K Kalavakunta,² Robert Lapenna²

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¹Department of Internal Medicine, Western Michigan University School of Medicine, Kalamazoo, Michigan, USA
²Borgess Medical Center/ Michigan State University, Kalamazoo, Michigan, USA

Correspondence to

Dr Guramrinder Singh Thind, guramrinder.thind@med.wmich.edu

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SUMMARY

Coronary artery fistulas are rare anomalous vascular connections between coronary arteries and a cardiac chamber or a central vessel, without an intervening capillary bed. Coronary-pulmonary fistulas are a distinct subset of coronary artery fistulas. We present the case of a previously healthy 63-year-old-man who presented with chest pain and was found to have mediastinal haemorrhage. Upon further investigations, he was found to have multiple coronary-pulmonary fistulas with pseudoaneurysm formation in three of the fistulas. Two of these pseudoaneurysms showed inflammatory changes indicative of recent bleed. These were determined to be the source of the mediastinal bleeding and patient's initial presentation. The patient was managed medically after obtaining multiple expert opinions from various institutions.

BACKGROUND

Coronary artery fistulas (CAFs) are rare anomalous vascular connections between coronary arteries and a cardiac chamber or a central vessel, without an intervening capillary bed. Coronary-pulmonary fistulas (CPFs) are a distinct subset of CAFs. A large proportion of CPFs are asymptomatic and are detected incidentally. Alternatively, they may have various presentations and complications.

CASE PRESENTATION

A 63-year-old man with a prior medical history of hypercholesterolaemia presented to the emergency room with a complaint of left-sided chest pain that started 3–4 hours prior to arrival. Pain was sharp, left sided, 10/10 in intensity and radiated to the back. No aggravating or relieving factors for the pain were reported by the patient. The pain was associated with nausea and diaphoresis. His systolic blood pressure on presentation was 163 mm Hg. Due to concerns of aortic dissection, a CT scan of chest with contrast was done that revealed haemorrhage in the mediastinum associated with a pseudoaneurysm measuring 1.6 cm. The pseudoaneurysm was associated with a blood vessel that appeared to be originating from the descending aorta.

Cardiology service was consulted and they planned to do a coronary angiogram to better characterise the vascular anatomy and to localise the source of mediastinal haemorrhage. The angiogram showed a complex network of dilated anomalous vessels originating from the right aortic sinus very

close to the right coronary artery (RCA) (figure 1, see online supplementary video 1), as well as from the proximal left anterior descending artery (LAD) (see online supplementary video 2). These vessels showed further branching, but the exact location of their drainage was unclear. Other findings included mild coronary artery disease (CAD) and a normal left ventricular systolic function with no active site of bleeding. With resolution of symptoms and absence of continued bleeding, he was discharged in stable medical condition on metoprolol for blood pressure control and further outpatient workup planned.

INVESTIGATIONS

A coronary CT angiogram (CCTA) was done for further assessment of the vasculature. It confirmed that multiple small to medium calibre anomalous vessels originated from proximal RCA, proximal LAD, as well as some from the descending thoracic aorta, with all of them draining into the main pulmonary artery. The fistula between proximal RCA and pulmonary artery had the largest calibre, and its site of drainage was aneurysmal in nature measuring 1.8 cm×0.8 cm (figure 2). On the medial aspect of the aortic arch, two pseudoaneurysms were seen measuring 2.1 cm×1.9 cm and 1.0 cm×0.5 cm, respectively. They originated from the anomalous vessels arising from the aorta and showed inflammatory changes indicative of recent bleed. These were determined to be the source of the mediastinal bleeding and patient's initial presentation. Because of these connections between the systemic and pulmonary circulation creating a left-to-right shunt, there was a concern of development pulmonary hypertension. Right heart catheterisation was performed to assess right heart pressures, which were found to be normal.

DIFFERENTIAL DIAGNOSIS

Evaluation of the coronary vasculature with CCTA confirmed the presence of CPFs and detailed their anatomy. The presence of inflammatory changes in two of the associated pseudoaneurysms confirmed these as the cause of patient's mediastinal haemorrhage on initial presentation.

TREATMENT

Due to the extensive number and size of his CPFs with a history of mediastinal bleed, ligation of native vessels and CPFs, and coronary artery bypass

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Figure 1 Coronary angiogram showing CPF originating from the RCA. CPF, coronary-pulmonary fistula; RCA, right coronary artery.

grafting (CABG) were considered. However, expert medical as well as surgical opinion from at least four different institutions was obtained, and it was unanimously decided to treat the patient conservatively.

OUTCOME AND FOLLOW-UP

Patient has been followed regularly in the cardiology clinic for 8 years and has remained asymptomatic. He has had coronary angiograms done periodically with the last one done 1 year ago, which did not reveal progression of the pseudoaneurysms or the CPFs.

DISCUSSION

CAFs can be broadly classified as coronary cameral fistulas and coronary-vascular fistulas.¹ Coronary cameral fistulas communicate with one or more of the cardiac chambers. Coronary-vascular fistulas terminate into one of the major thoracic vessels (including pulmonary artery, coronary sinus, caval veins, pulmonary vein, bronchial or mediastinal vessels). Hence, CPFs are a subtype of coronary vascular fistulas. Although the majority of CPFs are congenital, rare cases of acquired CPFs have been reported after chest trauma,² or very rarely as a complication of percutaneous coronary intervention or CABG.^{3,4} Unilateral CAFs have been defined as fistulas originating from one major coronary artery or its main branches with one or more termination sites.

Figure 2 Coronary CT angiogram revealing the pseudoaneurysm aneurysm (A) and coronary-pulmonary fistula (B) coming originating from the right coronary artery (C).

Similarly, bilateral and multilateral CAFs originate from two and three coronary arteries, respectively.⁵ The incidence of CAF as a whole has been reported to be 0.13%–0.22% in adults undergoing coronary angiography.⁶ In a meta-analysis performed by Said including 304 adults, the percentage of fistulas terminating into the pulmonary artery was 44% for unilateral fistulas, and 73% and 75% for bilateral and multilateral fistulas, respectively.⁷

The embryological origin of these CPFs has not been fully elucidated. However, one popular hypothesis is based on Hackensellner's 'involution-persistence theory'.⁸ This theory suggests that there are six anlagen in the truncus arteriosus corresponding to the six cusps of aortic and pulmonary valves, of which two in the aortic sinuses persist in developing the coronary buds while the other four disappear. The normally involuted anlagen in the pulmonary sinus may persist and communicate with the aortic sinus, thereby resulting in anomalous vascular connections terminating in the pulmonary artery.

Clinical presentation of CPFs varies. They may be asymptomatic and may be detected incidentally. Two case series on CPFs have been published so far involving 32 and 14 patients, respectively.^{1,9} From combined analysis, 47.8% of the patients in these two studies were asymptomatic. Major presenting symptoms were chest pain (17.3%), angina (15.2%), palpitations (10.8%) and exertional dyspnoea (6.5%). Coronary steal is a theoretical concern in these patients as the fistula provides a low-resistance path for the blood and diverts the flow from the nutrient coronary vessel. The clinical significance of this phenomenon has been debated. It has been argued that increased cardiac stress causes increased coronary vasodilation, thereby minimising this effect. Nonetheless, in the present with moderate to severe degree of CAD, this effect can cause exertional chest pain and may warrant intervention. Fractional flow reserve (FFR) can be used during coronary angiography to objectively assess coronary steal in this scenario, as demonstrated by Härle *et al.*¹⁰ They temporarily occluded the CPF with balloon and measured the FFR in the nutrient coronary artery before and after the occlusion. These values were found to be 0.78 and 0.95, respectively, thereby demonstrating haemodynamically significant coronary steal in this particular case. Our patient only had minimal CAD and thus coronary steal was not a concern.

Another concern with these patients is the development of pulmonary hypertension. Haemodynamically, CPFs cause left-to-right shunts and resemble the circulatory physiology of patent ductus arteriosus. Hence, CPFs may lead to pulmonary hypertension if the flow of the left-to-right shunt is significant enough. This may present as exertional dyspnoea and palpitations. Objective assessment of right heart pressure, as performed in our case, is warranted if pulmonary artery hypertension is suspected. Other complications of CPFs include aneurysm formation, which very rarely ruptures. Rare cases of haemopericardium and cardiac tamponade due to coronary fistula rupture have been reported in the literature.¹¹

CAF in general can be detected on routine imaging of chest or echocardiogram. Because of its popularity, coronary angiography remains the cornerstone of CAF detection. However, although not widely available, multidetector coronary CT angiogram can provide better anatomical characterisation including the course and termination of the fistulas, as exemplified with our case. Owing to its non-selective nature and superior three-dimensional imaging, CCTA can better demonstrate the multiple origins of bilateral or multilateral fistulas and fistulas arising from the thoracic aorta.⁷ Management decisions in adults with CAFs in general are mostly based on expert opinion and should be individualised. Asymptomatic cases can be managed conservatively.

As mentioned before, if coronary steal is suspected in patients with angina and some degree of CAD, objective assessment with FFR can be performed to guide therapy. Other candidates for intervention include patients with severe or worsening pulmonary hypertension, congestive heart failure, high risk of endocarditis or those undergoing an invasive procedure for another cardiac problem.¹

Learning points

- ▶ Coronary-pulmonary fistulas should be recognised as a potential cause of mediastinal haemorrhage.
- ▶ Coronary-pulmonary fistulas cause left-to-right shunting, which if significant may lead to pulmonary hypertension; patients should be evaluated for the same.
- ▶ Coronary CT angiogram can provide better anatomical details for evaluation of coronary-pulmonary fistulas.
- ▶ Although certain indications for surgical repair have been identified, the treatment plan should be individualised after interdisciplinary discussion.

Contributors All authors contributed to the manuscript as follows. GST and YA were both part of the team that admitted the patient. RL and JKK were part of the cardiology team that evaluated the patient. Hence, all authors were directly involved with patient care. GST and YA did the literature review and manuscript write-up. RL and JKK revised the manuscript critically for important intellectual content. All authors gave their final approval of the version to be published.

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REFERENCES

- 1 Said SA, Nijhuis RL, Akker JW, *et al*. Unilateral and multilateral congenital coronary-pulmonary fistulas in adults: clinical presentation, diagnostic modalities, and management with a brief review of the literature. *Clin Cardiol* 2014;37:n/a–45.
- 2 Bulur S, Satılmışoğlu MH, Özhan H, *et al*. Coronary-to-pulmonary artery fistula due to a penetrating trauma. *Türk Kardiyol Dern Ars* 2012;40:109.
- 3 Hagau AM, Dudea SM, Hagau R, *et al*. Large coronary pseudoaneurysm with pulmonary artery fistula, six months after left main trunk stenting with paclitaxel-eluting stent. *Med Ultrason* 2013;15:59–62.
- 4 Musleh G, Jalal A, Deiraniya AK. Post-coronary artery bypass grafting left internal mammary artery to pulmonary artery fistula: a 6 year follow-up following successful surgical division. *Eur J Cardiothorac Surg* 2001;20:1258–60.
- 5 Said SA, van der Werf T. Dutch survey of congenital coronary artery fistulas in adults: coronary artery-left ventricular multiple micro-fistulas multi-center observational survey in the Netherlands. *Int J Cardiol* 2006;110:33–9.
- 6 Latson LA. Coronary artery fistulas: how to manage them. *Catheter Cardiovasc Interv* 2007;70:111–8.
- 7 Said SA. Current characteristics of congenital coronary artery fistulas in adults: a decade of global experience. *World J Cardiol* 2011;3:267–77.
- 8 Hackensellner HA. [Accessory cardiac vessels arising from the pulmonary artery and their importance in understanding the etiology of the origin of one or both coronary arteries from the pulmonary artery]. *Frankf Z Pathol* 1955;66:463–70.
- 9 Lee CM, Song SY, Jeon SC, *et al*. Characteristics of coronary artery to pulmonary artery fistula on Coronary Computed Tomography Angiography. *J Comput Assist Tomogr* 2016;40:398–401.
- 10 Härle T, Kronberg K, Elsässer A. Coronary artery fistula with myocardial infarction due to steal syndrome. *Clin Res Cardiol* 2012;101:313–5.
- 11 Bauer HH, Allmendinger PD, Flaherty J, *et al*. Congenital coronary arteriovenous fistula: spontaneous rupture and cardiac tamponade. *Ann Thorac Surg* 1996;62:1521–3.

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