Reminder of important clinical lesson

The primary percutaneous coronary intervention for acute anterior myocardial infarction in a middle-aged male patient with bilateral coronary artery to pulmonary artery fistulas

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Summary

A 38-year-old man admitted to emergency department with 2 h of typical substernal chest pain, shortness of breath and nausea. The ECG revealed sinus rhythm with a 3 mm ST elevation in precordial leads V1–V6. The coronary angiography revealed acute total occlusion in left anterior descending artery (LAD) with normal circumflex and right coronary artery (RCA) along with bilateral fistulas arising from the proximal LAD and ostial RCA draining into the main pulmonary artery. Therefore, primary percutaneous coronary intervention and bare metal stent implantation was performed to culprit LAD lesion. The electrocardiographically gated 64-slice multidetector-row CT showed two large, tortuous abnormal vessels which arose from the both ostial part of the RCA and LAD draining into the main pulmonary artery. We report an unusual case of bilateral coronary artery to pulmonary artery fistulas leading to acute anterior myocardial infarction in a middle-aged male patient.

BACKGROUND

Congenital anomalies of the coronary arteries are rare and they have been discovered between 0.67% of the patients who undergo coronary arteriography and in 0.8% of patients at autopsy.^{1–2} Coronary artery fistulas (CAFs), which were first described by Krauss in 1865, are abnormal communications between epicardial coronary arteries and the cardiac chambers, the major vessels (the vena cavae, subpulmonary veins and pulmonary artery) or other structures.^{3–5} They are very rare congenital or acquired coronary artery anomalies with an estimated incidence of 0.002% in the general population. Also, they account for between 0.13% and 0.87% of all coronary anomalies in angiographic series^{6–7} and represent 0.27–0.40% of all congenital cardiac disease.⁸ Congenital CAFs draining into the pulmonary artery are very rare cardiac anomalies. Although many patients with CAF are asymptomatic, they may present with angina pectoris, atypical chest pain, palpitation, fatigue, bacterial endocarditis, congestive heart failure, pericardial effusion and sudden cardiac death. Although some reports of bilateral coronary to pulmonary arteries fistulas have been previously reported,^{4–10} there are no reports describing a case of coronary to pulmonary arteries fistulas with acute myocardial infarction requiring primary percutaneous coronary intervention. Bilateral fistulas which originate from the coronary system represent for 5% of all CAFs.¹¹

We report an unusual case of bilateral coronary artery to pulmonary artery fistulas leading to acute anterior myocardial infarction in a middle-aged male patient.

CASE PRESENTATION

A 38-year-old man admitted to emergency department with 2 h of typical substernal chest pain, shortness of

breath and nausea. His medical history was unremarkable without any cardiovascular risk factors except for smoking. Physical examination on admission revealed a blood pressure of 125/80 mm Hg, a regular heart rate of 95 beats/min. Cardiac auscultation revealed 2/6 grade systolic murmur radiating to the left axilla in lower left sternal region. The rest of the physical examination including respiratory sounds was unremarkable. The initial ECG revealed sinus rhythm with a 3 mm ST elevation in precordial leads V1–V6. The maximal troponin I elevation was 21 IU (normal range 0–0.06 IU) after the onset of clinical symptoms. The patient was transferred to the coronary care unit with diagnosis of acute anterior ST segment elevation myocardial infarction. Acetylsalicylic acid 300 mg, clopidogrel 600 mg and enoxaparin 60 mg were given intravenously as the initial medical therapy.

INVESTIGATIONS

The coronary angiography, which was done in 2 h of onset of symptoms, revealed acute total occlusion in left anterior descending artery (LAD) with normal circumflex (Cx) and right coronary artery (RCA). Also, it showed bilateral fistulas arising from the proximal LAD and ostial RCA draining into the main pulmonary artery, with an evidence of atherosclerotic coronary artery disease in left coronary artery tree (figure 1). Therefore primary percutaneous coronary intervention and bare metal stent implantation were performed to culprit LAD lesion. Thrombolysis in myocardial infarction-II (TIMI II) blood flow was obtained after procedure. Also, the right heart catheterisation was performed and there was seen significant step-up of oxygen saturation in the right heart and pulmonary arteries with remarkable pulmonary–systemic flow ratio ($Q_p/Q_s > 1.7$) and mild increased pulmonary

Figure 1 Selective left coronary angiography, right anterior caudal position: fistula between left anterior descending artery and pulmonary artery (arrow) (right). Selective right coronary angiography, left oblique position: tortuous fistula between proximal right coronary artery and pulmonary artery (arrows) (left).

artery pressure up to 40 mm Hg, respectively. Transthoracic echocardiography performed after the procedure showed mild tricuspid and mitral insufficiency, pulmonary artery pressure of 35–40 mm Hg, anterior and anteroapical moderate hypokinesia, left ventricular hypertrophy and marked decreased left ventricular systolic functions with an ejection fraction of 35%. There was a marked diastolic turbulent flow distal to the pulmonary artery as seen in the parasternal short-axis view (figure 2). The ECG-gated 64-slice multidetector-row CT showed two large, tortuous abnormal vessels which arose from the both ostial part of the RCA and LAD draining into the main pulmonary artery (figure 3). These abnormal vessels imaging were detected with coronary angiography as fistulas.

DIFFERENTIAL DIAGNOSIS

Treatment

The patient was referred to the cardiovascular surgery department for fistula closure operation due to his persistent recurrent angina episodes under optimal anti-ischaemic, antithrombotic and antianginal treatment and increased pulmonary–systemic flow ratio ($Q_p/Q_s > 1.7$) with a mild increased pulmonary artery pressure; patient was referred to the cardiovascular surgery department for fistula closure operation. The patient refused any surgical

Figure 2 Parasternal short-axis view of the transthoracic echocardiography shows diastolic turbulent flow distal to the main pulmonary artery.

Figure 3 Multidetector-row CT (MDCT) imaging of the fistula between right coronary artery and pulmonary artery (black arrows) (left). MDCT imaging of the fistula between left anterior descending artery and pulmonary artery (black arrow) (right).

treatment and therefore was discharged from the hospital with an optimal anti-ischaemic, antithrombotic and antianginal medical treatment including oral acetylsalicylic acid 300 mg/day, clopidogrel 75 mg/day, metoprolol succinate 50 mg/day, perindopril 10 mg/day, atorvastatin 40 mg/day and isosorbide mononitrate 60 mg/day.

DISCUSSION

CAFs are very rare congenital or acquired (trauma, inflammation, myxoma and thrombus) coronary anomalies which can arise from any of the three major coronary arteries and drain into any of the cardiac chambers and great vessels. The incidence of these anomalies is 0.002% in the overall population. In a comprehensive review of cases with CAF, 50–58% of the fistulas were found to be originating from the RCA, 25% from the LAD, 18.3% Cx, 1.9% diagonal artery and 0.7% left main coronary artery.¹² Bilateral fistulas, arising from both left and right coronary arteries, represent for 5% of the total CAFs. The common sites of drainage are pulmonary artery (29.8–43%), right ventricle (14–40%), right atrium (19–20.2%), left ventricle (5.8–19%) and left atrium (5%). Approximately 56% of the bilateral fistulas and 17% of the unilateral fistulas have been found to drain into the pulmonary artery.¹³ In our case, the patient had bilateral coronary to pulmonary artery fistulas arising from LAD and RCA. The embryogenesis of CAF is uncertain. There are six coronary enlargements in the embryo: three in the developing aorta and three in the developing pulmonary artery. These enlargements normally involute, except for the two from the right and left aortic sinuses. A coronary to pulmonary artery fistula may result from the persistence of one or more of the pulmonary arterial enlargements, hence the term accessory coronary artery. The relatively high incidence of bilateral CAFs to the pulmonary artery is in accord with this theory.¹⁴ The clinical importance of CAF depends upon the amount of blood traversing the communication and the cardiac chamber or the great vessel into which the fistula drains. Even though many patients with CAF are asymptomatic, the most frequent clinical complaints are angina pectoris, palpitation, atypical chest pain, dyspnoea, orthopnoea, fatigue and lethargy. The pathophysiological mechanisms of the symptoms are volume overload and pulmonary congestion as a

consequence of left-to-right shunting, severe coronary steal that causes a diminished myocardial oxygen supply resulting acute myocardial ischaemia or infarction when combined with vulnerable thrombotic atherosclerotic plaques and the lack of collateral and capillary vessel formation.¹⁵ The main complications of CAFs are bacterial endocarditis, congestive heart failure, arrhythmia, pericardial effusion, atherosclerosis complicated with myocardial infarction or even sudden cardiac death.^{8–17}

Myocardial infarction is seen rarely in cases of coronary to pulmonary artery fistulas.¹⁸ In recent study, the prevalence of myocardial infarction was found to be 5%.¹⁹ Another retrospective study revealed myocardial infarction in 9 of 51 instances of CAF.²⁰ Myocardial infarction in patients with coronary to pulmonary artery fistulas has multiple potential aetiologies including a prothrombotic state, abnormal myocardial perfusion pressure, flow-mediated inflammatory vascular changes and coronary atherosclerosis.^{21–22} The decreased coronary blood flow distal to the fistula may promote myocardial ischaemia and myocardial infarction and it has been documented in patient with coronary to pulmonary artery fistulas with or without evidence of coronary atherosclerosis.^{22–23} Although, a coronary artery steal caused by the coronary to pulmonary artery fistulas is a rare cause of myocardial ischaemia by itself, it may trigger myocardial ischaemia and occurrence of coronary thrombosis or leading to myocardial infarction in the presence of significant coronary atherosclerosis.²⁴ As in our case, the relation between coronary to pulmonary artery fistulas and occurrence of coronary thrombosis leading to myocardial infarction has been demonstrated in some studies.^{25–26} This may be explained by coronary steal phenomenon which occurs due to flow shunting over the coronary fistula and by acute coronary thrombosis on an uncritical atherosclerotic plaque which appeared non-ulcerated and thrombus-free at coronary angiography. In our case, the patient was admitted to the hospital with an acute anterior myocardial infarction which may have taken place as a result of rupture of vulnerable atherosclerotic plaques concomitant with severe coronary steal effect of the bilateral CAFs drained into pulmonary artery. Pharmacological treatment is preserved for asymptomatic or small CAFs, patients without favourable coronary anatomy for surgery and percutaneous interventional procedures. While the main indications for surgery are symptomatic CAFs unresponsive to pharmacological treatment, aneurysmal degeneration, large left to right shunts, congestive heart failure, coronary rupture or side branch obstruction, small fistulas especially those that drain into the main pulmonary artery and the left ventricle with a pulmonary–systemic flow ratio exceeding 1.5:1 may be referred to percutaneous intervention.⁷ In our case, the surgical fistula closure operation was recommended to the patient for recurrent anginal episodes under optimal medical therapy after successful stent implantation procedure, and significant step-up of oxygen saturation measured in the right heart and pulmonary artery concomitant with an increased pulmonary artery pressure up to 50 mm Hg and the remarkable pulmonary–systemic flow ratio ($Q_p/Q_s > 1.7$) in the right heart catheterisation. Current treatment options include transcatheter embolisation, stent implantation or surgical ligation procedures. Although vessel tortuosity

and multiple drainage sites may lead CAFs unsuitable for transcatheter embolisation, the closure rate of this technique is nearly 85%.²⁷ The most widely used surgical techniques include an internal closure from within the cardiac chamber or pulmonary artery (46.1%), tangential arteriography (28.8%), ligation and coronary bypass grafting (3.8%), distal ligation alone (11.5%) or proximal and distal ligation/division (6.7%) with or without closure from within the aneurysmal coronary artery (2.9%).^{28–29} Surgical ligation is associated with a relatively low death rate compared to other surgical techniques (1.4%).²⁷ The definite treatment of CAF is unclear, especially in patients with small shunts. Liberthson reviewed 187 cases and found that operative mortality (1%) and complication rates (7%) were low in younger patients undergoing operation below the age of 20. However, it was detected that more serious postoperative mortality (7%), postoperative myocardial infarction (7%) and other complications (23%) occurred in adults. The authors recommended surgery in all cases of CAF in childhood independent of symptoms or size of the shunt.^{30–33} The main problem for surgery is the long-term results of these patients. In a series of 41 patients operated on for CAF, Cheung *et al* showed that 80.5% of the patients were asymptomatic in the follow-up (range 5.08 months–28.09 years). However, the authors revealed a recurrence rate of nearly 19% which was more important in the external ligation group as compared to the intracardiac closure group (22% vs 16%, respectively).²⁰ A new therapeutic option for the treatment of CAF is the percutaneous transcatheter embolic occlusion or transcatheter closure technique, which has been regarded as an alternative to surgery. A variety of materials have been used, including Gianturco coils, covered stainless-steel coils, detachable balloons, coaxial embolisation with platinum microcoils, double-umbrella device and the Gianturco Grifka vascular occlusion device.^{27–34–35} The most important difficulties of this technique are extreme vessel tortuosity, multiple drainage sites and coronary branches at the site of optimal device positioning.

Learning points

- In conclusion, multiple bilateral coronary artery to pulmonary artery fistulas are rare congenital or acquired anomalies.
- As in our case, the severe coronary steal through the fistula combined with a rupture of vulnerable thrombotic atherosclerotic plaques may result in clinically significant ischaemia or acute myocardial infarction.
- Coronary angiography and multidetector CT not only provide valuable information about coronary anatomy, but also help to determine treatment decision including percutaneous coronary intervention or surgery.
- While there are several treatment options available for CAF; the most safe and effective method to adopt requires further investigation in larger populations.
- Physicians should be aware of such coronary artery anomalies in order to diagnose and treat them properly.

Competing interests None.

Patient consent Obtained.

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