Rare disease

An uncommon anatomy presenting with a common disease

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Summary

Coronary artery fistulas are rare anomalous communications, between coronary arteries and cardiac chambers and great vessels. They are often congenital, but usually present in adulthood. They can affect cardiac haemodynamic stability and are thought to predispose patients to heart failure, myocardial ischaemia, myocardial infarction, infective endocarditis, arrhythmias and rupture. Herein, a case is discussed where a patient with long-standing stable angina was found to have a coronary artery fistula to the main pulmonary artery and concomitant ischaemic heart disease with a chronically occluded left anterior descending artery, proximal to the fistula. It is thought that the fistula probably predisposed the patient's ischaemic heart disease. He underwent a successful coronary artery bypass grafting plus surgical ligation of the coronary artery fistula. This uncommon coronary artery anomaly, presenting with ischaemic heart disease, a common disease in adulthood, is discussed in the context of current recommendations.

BACKGROUND

Coronary artery fistulas (CAFs) are rare anomalies, which involve an abnormal communication between a coronary artery and any cardiac chamber or great vessel. They are often congenital but acquired forms can occur after cardiac surgery and catheter intervention. Congenital CAFs are thought to be the most common coronary anomaly affecting haemodynamic parameters.^{1–3} CAFs occur in about 2 in 1000 people in the general population and they are found in about 0.25% of those undergoing coronary angiography.^{4–5} Their clinical importance is usually in adulthood with an increased risk of heart failure, myocardial ischaemia, myocardial infarction, infective endocarditis, arrhythmias and rupture.^{6–7}

A large Dutch study by Salah in 2011 showed that most patients (80%) had unilateral fistulas, 18% presented with bilateral fistulas and 2% with multilateral fistulas. CAFs originated from the left coronary artery in 69% and from the right coronary artery (RCA) in 31% of the subjects. Termination into the pulmonary artery (PA) was reported in unilateral (44%), bilateral (73%) and multilateral (75%) fistulas. Although, 85% of patients were symptomatic, often presenting with dyspnoea (31%), chest pain (23%), angina pectoris (21%), palpitations (13%) and congestive heart failure (8%), only a small number of patients presented with a myocardial infarction (2%).⁸

Management of CAFs in symptomatic patients or patients with other cardiac abnormalities is well documented either with surgical ligation or catheter closure.^{9–12} However, consensus on management of asymptomatic patient remains elusive.

Herein we report a case of an adult male with a CAF arising from the left anterior descending artery (LAD) draining in the PA with concurrent coronary artery disease which may have been predisposed by his unusual coronary anatomy.

CASE PRESENTATION

A 61-year-old gentleman who was currently working as a fibreglass laminator presented with a 3 month history of stable angina. Although on further questioning it is likely that he may have had angina for several years, with left shoulder and arm numbness whenever he exerted himself dancing in the last 2–3 years. The patient was also known to have a background of hypertension, hypercholesterolaemia and a family history of atherosclerosis. He was a non-smoker and a social drinker.

INVESTIGATIONS

Exercise stress test under the Bruce protocol was positive for myocardial ischaemia with a reduced exercise capacity due to angina and dyspnoea with an upsloping inferolateral ST changes at 3 min. Preoperative transthoracic echocardiogram showed a normal left ventricular size and function, with mild inferolateral hypokinesis while his heart valves were normal. His preoperative ECG was normal. His preoperative blood tests were all within normal limits. Troponins tests were all negative.

Coronary angiography showed two vessel disease with a 100% occluded proximal LAD well collateralised from the RCA, 75% occluded left circumflex artery (LCx), minor RCA disease and CAF connecting the LAD and PA with its origin proximal to the LAD stenosis. The CAF took off from the proximal LAD, made a tortuous route, including a long hair-pin bend, before draining to the main PA.

TREATMENT

The patient subsequently underwent off-pump coronary artery bypass grafting via a median sternotomy with the left internal mammary artery grafted to the LAD and reversed saphenous vein graft to the lateral marginal branch of LCx. The CAF was found to be partly buried on the anterior wall of the main PA as it made a hair-pin bend.

Figure 1 Coronary Angiography. The distal end of the coronary artery fistula (black arrow) emptying into the pulmonary artery after a tortuous passage is highlighted. The occluded proximal LAD is shown only as a stump (gray arrow) where the coronary artery fistula originates. The 75% occlusion on the proximal LCx is also depicted (white arrow).

The CAF was surgically ligated with a 4.0 pledgeted suture in an over and under fashion and was occluded at the left lateral aspect of the main PA. The patient remained stable haemodynamically throughout the procedure and did not require any inotropic supports.

He was extubated in the intensive care unit as per protocol. His recovery was straightforward postoperatively, apart from some mild left hand weakness and paraesthesia. He progressed uneventfully on the ward and he was discharged on postoperative day 5.

OUTCOME AND FOLLOW-UP

The patient is doing very well 9 months after his operation. He is back to his normal exercise capacity with no recurrence of his dyspnoea or angina. His mild left hand weakness and paraesthesia is now resolved. (This is thought to be neuropraxia of lower region of the brachial plexus related to the sternotomy. Investigations of this were negative for a cerebrovascular accident).

DISCUSSION

CAFs were first described by Kraus in 1865 and was first surgically treated by Bjork and Crawford in 1947.^{13 14} Embryologically, they seem to represent persistent junctions of primordial epicardial vessels with intramyocardial sinusoidal circulation. They can be present with other congenital or acquired cardiac abnormalities such as valvular and septal pathologies and ischaemic heart disease.^{1 6} CAFs are uncommon malformations, they are thought to

cause haemodynamic compromise, myocardial perfusion abnormalities and accelerated atherosclerosis that may result in varying symptoms as common as dyspnoea to sudden death.

Many patients are asymptomatic in the first 2 decades of life. However, symptoms and complications may develop with increasing age and when surgical intervention is performed to correct the anomaly, there is increased morbidity and mortality. Complications from an untreated CAF include 'coronary steal' from the adjacent myocardium, thrombosis and embolism, cardiac failure, atrial fibrillation, rupture, endocarditis/endarteritis and arrhythmias.^{1 6 7}

The aim of treatment is to close the fistula and correct any other abnormality if need be. Surgical ligation has been the mainstay of CAFs closure with good long term results.^{9 15 16} Percutaneous catheter closure is now becoming a safe and effective alternative to surgical ligation. Different types of embolisation materials can be used such as detachable balloons, stainless steel coils or platinum microcoils.¹² The indications for treatment of CAFs include the presence of a large or increasing left-to-right shunt, left ventricular volume overload, myocardial ischaemia, left ventricular dysfunction, congestive cardiac failure and for prevention of endocarditis/endarteritis. Percutaneous catheter closure is usually indicated when the CAF is small, proximal, non-tortuous, has a single drain site, in older patient and with no other cardiac abnormality requiring surgical intervention. Surgical closure is usually indicated when a CAF is large, distal, tortuous, has multiple drains site and the patient has a concomitant cardiac disease. There are no current guidelines published as indications for closure of CAFs however, there is general consensus to treat if they have the aforementioned conditions especially when the patient is symptomatic. There remains to be controversy in treating patient who are asymptomatic and found to have a CAF. Interestingly, groups which are surgically orientated tend to advocate for ligation even for those who are yet to present with symptoms,^{11 15 17 18} while groups who intervene percutaneously, cardiologist and interventional radiologist, tend to slightly be more conservative.^{12 5}

The patient had what appears to be a small to medium sized congenital CAF originating from the proximal LAD, having a tortuous route including a hair-pin bend, before terminating at the main PA (figure 1). On his angiogram, his CAF, which was approximately 2 mm in diameter, did not appear to be shunting large amount of blood left to right, however, the patient had been complaining of angina for at least 2–3 years before presentation. This may indicate coronary steal syndrome predisposing him to atherosclerosis which likely lead to the complete occlusion of his proximal LAD just after the origin of his CAF.

The patient presented with a rare coronary artery anatomy which may have predisposed a common pathology, atherosclerosis. Had he presented earlier, when his symptoms initially started and if his CAF had been closed earlier, could his ischaemic heart disease been prevented or at least delayed? The answer remains unclear.

Learning points

- ▶ CAFs are rare coronary malformations but they are thought to predispose patients to significant sequelae and should not be missed.
- ▶ Treatment is advocated for patients with symptoms and for those asymptomatic patients who are at risk for future complications.
- ▶ Surgical ligation is safe and effective. Percutaneous closure is a good alternative for a select group of patients.

Competing interests None.

Patient consent Obtained.

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