CASE REPORT

Giant aneurysm in a left coronary artery fistula: diagnostic cardiovascular imaging and treatment considerations

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SUMMARY

Congenital coronary artery fistula complicated with giant coronary artery aneurysm is a very rare condition. In this case report, we present a 65-year-old woman, referred to us with a continuous heart murmur, occasional atypical chest pain and few episodes of fainting. A giant aneurysm and a coronary–pulmonary fistula were diagnosed using multiple cardiovascular imaging modalities to provide a sufficient anatomical picture. The patient was considered at high risk of sudden death from aneurysm rupture and received surgical treatment. Subsequent histopathological examination revealed a true aneurysm with severe wall calcifications, ulcerations and large areas with marked thinning of the wall. The postoperative course was uneventful.

BACKGROUND

Coronary artery fistula (CAF), congenital or acquired, is a very rare coronary anomaly with an estimated prevalence of 0.002% in the general population.¹ CAF is defined as an abnormal coronary artery termination into any cardiac chamber or a great vessel, bypassing the myocardial capillary network. CAFs are characterised by the number, origin, course, termination and presence of aneurysm or stenotic lesions.

We report a case with several congenital vascular malformations—a giant aneurysm in a coronary fistula fed by the left coronary artery (LAD) and a coronary–pulmonary fistula fed by the right coronary artery (RCA).

CASE PRESENTATION

A 65-year-old woman was referred to hospital with an accidental finding of an abnormal continuous heart murmur. A transthoracic echocardiogram (TTE) showed a huge abnormal cavity close to the pulmonary trunk (PT). Cardiovascular risk factors and symptoms included hypertension, hypercholesterolaemia, occasional atypical chest pain and few cases of unexplained fainting during physical activity. A grade 2 continuous heart murmur had been heard at the upper left parasternal region 25 years ago, but further examinations had been inconclusive. Recent electrocardiogram showed sinus rhythm with T-wave depression in V1 and V2.

INVESTIGATIONS

Repeated TTE revealed a continuous high-frequency flow (peak Doppler gradient 50 mm Hg) draining into the PT, consistent with a coronary–

pulmonary fistula. It also showed a giant aneurysm with a turbulent flow in close relation to the PT. Cardiac chamber sizes were all normal. A 320-slice multidetector CT (MDCT) coronary angiography (Toshiba Aquillion One) demonstrated a bundle of vessels arising from the proximal RCA emptying into the PT, and a fistula fed by the LAD supplying a 5.2×6.2 cm aneurysm. The aneurysm was found to the left side of the pulmonary artery, and contained a turbulent intraluminal flow and extensive wall calcification (figure 1). A coronary angiogram (CAG) confirmed a coronary–PT fistula arising from the RCA, forming a vascular conglomerate, terminating into the PT. Furthermore, it confirmed an aneurysm fed by the LAD only, with no visible connection to the coronary–pulmonary artery fistula (figure 2).

TREATMENT

Based on the MDCT and CAG findings, the patient was considered at high risk of sudden death from aneurysm rupture and was therefore referred for surgery.

OUTCOME AND FOLLOW-UP

Surgery confirmed MDCT and CAG findings and the aneurysm was found to have multiple wall calcifications, mural thrombi and several wall areas were considered to be dangerously thin. Intra-aneurysm sac pressure was measured to be 75% of the systemic pressure. Under cardiopulmonary bypass, the efferent vessels were ligated, and the aneurysm was resected without any complications.

A subsequent histopathological examination revealed a true aneurysm with severe wall calcifications, ulcerations and large areas with marked thinning of the wall. Microscopy showed severe degeneration of the muscle fibres, hyaline sclerosis, massive plaque formation in the vascular wall and intraluminal thrombi.

The postoperative course was uneventful and the patient discharged after 2 weeks.

DISCUSSION

Coronary–pulmonary fistula is a very rare condition often found incidentally during coronary angiography or TTE. The etiology of CAF is most frequently congenital and the clinical consequence mainly dependent on the magnitude of the left-to-right shunt. Approximately 50%, 42% and 5% of CAF arise from the RCA, the LAD and both vessels, respectively. The most common drainage

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Figure 1 Multidetector CT images. (A)–(D) A giant mass with calcifications is located to the left of the pulmonary trunk. The afferent vessels arising from the left anterior descending artery. Coronary–pulmonary fistula fed by the right coronary artery.

sites are low-pressure structures as: the right ventricle (41%), right atrium (26%), pulmonary artery (17%), coronary sinus (7%), left atrium (5%), left ventricle (3%) and the superior vena cava (1%), respectively.^{1 2} Most fistulas are single communications, but multiple fistulas and fistulas associated with fusiform or saccular aneurysms have been reported.³ The clinical presentation of CAF includes a continuous heart murmur, dyspnoea, fatigue, angina, arrhythmias, myocardial ischaemia, myocardial infarction or sudden death. Potential long-term complications caused by a large left-to-right shunt are right ventricular enlargement, pulmonary

hypertension, congestive heart failure, rupture or thrombosis of the fistula or associated arterial aneurysm.⁴

It is assumed that symptoms presented in this case are caused by haemodynamic changes following the progressive enlargement of fistula and aneurysm. These symptoms can be explained by the coronary steal phenomenon—a decreased coronary blood flow distal to coronary artery malformations due to diastolic runoff from the distal branches to low-pressure receiving cavities, resulting in myocardial ischaemia and possibly ventricular arrhythmias, explaining chest pain and syncope episodes

Figure 2 Digital subtraction coronary angiography. (A) The fistulous vessels arising from the proximal sites of the right coronary artery. (B) Huge aneurysm arising from the fistula fed by the left anterior descending artery, with no visible efferent vessels.

during physical activity. Coronary steal are more likely to occur when large fistulas shunt to the right side of the heart, which occur in over 90% of all cases.¹

Precise evaluation of the clinical presentation and the CAF morphology, including the anatomical origin and course, drainage site and the possible presence of an aneurysm, is necessary when considering treatment indication and options. The present case demonstrates a necessity of using several cardiovascular imaging modalities when investigating a complicated coronary anomaly including TTE, MDCT and invasive coronary angiography. The use of three-dimensional reconstruction and multiplanar reformation from an MDCT scan can provide details regarding the CAF's morphological features and spatial relationship with adjacent structures, which may help clarifying the diagnosis in incidentally found CAFs. In this case, an MDCT provided a partial anatomic description of the fistula's origin, proximal path, drainage site, and the size and shape of the aneurysm. The major limitation of MDCT is radiation exposure which, however, can be lowered using different reduction techniques.

Conventional coronary angiography is regarded as the gold standard for the diagnosis of multiple coronary abnormalities.⁵ In this case it provided a sufficient anatomical picture of the coronary–pulmonary fistula and the aneurysm fed by the LAD. However, the drainage site of a CAF, when being a low-pressure chamber, may not be well visualised, because of the significant contrast medium dilution in the drainage site.¹ Two-dimensional CAG projections and limited experience with complex coronary anomalies may result in many CAFs being overlooked or misinterpreted.⁶ Because of such limitations, transthoracic and transoesophageal echocardiography, magnetic resonance and MDCT have shown to be useful for a complete evaluation.

There is consensus about performing surgery in symptomatic patients with CAF. Treatment of adult asymptomatic patients with non-significant shunting to prevent fistula-related complications is still a matter of debate. In the present case, the CAF was combined with a significant aneurysmal formation with high intraluminal pressure and wall thinning, creating an absolute rationale for surgery to eliminate the risk of severe complications in terms of sudden death from rupture. Operative findings, including mural thrombi, high intra-aneurysm pressure and areas with very thin walls, confirmed our concerns regarding the aneurysm.

In conclusion, an MDCT is a valuable non-invasive technique for diagnosing, monitoring and follow-up of the patients with CAFs, but should always be supplemented with a conventional coronary angiography before treatment decision, especially in cases with complex vascular anatomy.

Learning points

- ▶ Patients with coronary artery fistula (CAF), complicated with an aneurysm, can be asymptomatic or present with an atypical clinical picture.
- ▶ Precise evaluation of the CAF morphology, and the possible presence of an aneurysm, is necessary when considering treatment indication and options.
- ▶ Multidetector CT and coronary angiogram can have an additive value in diagnosis of this cardiac anomaly.

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