CASE REPORT

Congenital circumflex coronary artery fistulae; presentation and diagnosis

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SUMMARY

Small fistulae between coronary arteries and the pulmonary artery are common, but abnormal vascular communications between the coronary arteries and other cardiac structures are rare. They were first described by Krause in 1865. We report two cases of fistulous connexions between the circumflex coronary artery and the coronary sinus.

BACKGROUND

Congenital circumflex coronary artery to coronary sinus fistula is a rare condition and is one of the rare causes of heart failure. It was first described by Krause in 1865.¹ It can be an incidental finding during coronary angiography, but coronary CT or MR angiography are the diagnostic modality of choice to demonstrate the precise anatomy of the malformation. Both cases reported here were treated medically with no surgical intervention.

CASE PRESENTATION

Case 1

A 76-year-old man presented with 18 months history of gradually worsening shortness of breath on exertion. He has had no history of chest pain or syncope. On physical examination, the pulse rate was 95, the blood pressure was 168/79 and a loud pansystolic heart murmur was heard at the right second intercostal space along the sternal border with mild pitting oedema of both ankles.

Case 2

A 61-year-old man presented with symptomatic atrial fibrillation. Six months later, he developed leg swelling and shortness of breath on exertion and he was treated successfully with furosemide.

INVESTIGATIONS

Case 1

The ECG showed some non-specific lateral T wave changes; and the echocardiogram demonstrated a normal left ventricular systolic function, and a thickened and calcified aortic valve with restricted opening, the peak transaortic gradient was 40 mm Hg with a calculated valve area of 1.5 cm².

A coronary angiogram was performed with a view to aortic valve replacement, and it revealed large tortuous right and circumflex coronary arteries both approximately 3 cm across. Both vessels drained directly into the coronary sinus (see figures 1 and 2).

Case 2

The ECG confirmed atrial fibrillation, poor R wave progression with T wave inversion in leads I and the right augmented limb lead on ECG. An echocardiogram showed an abnormal flow in a structure lateral to the left atrium. A subsequent transoesophageal echocardiogram showed a grossly dilated circumflex coronary artery with multiple bends down its length and a grossly dilated coronary sinus. Cardiac MR confirmed a circumflex coronary artery to coronary sinus fistula (see figures 3 and 4).

DIFFERENTIAL DIAGNOSIS

Both patients presented with the heart failure syndrome. Common potential underlying diagnoses were coronary artery disease and aortic stenosis.

TREATMENT

In both cases the diagnosis was high output heart failure due to coronary artery to coronary sinus fistulae. Both were managed symptomatically, with diuretics causing resolution of symptoms. Although a surgical repair was considered, in both cases the risk was thought to be prohibitive as too much of the myocardium was supplied by side branches from the fistulous arteries. In both cases, the entry point from the fistulae into the coronary sinus was too wide to allow safe embolisation. The patients were managed with combination therapy with ACE inhibitor and β -blocker.

Figure 1 Ascending aortogram showing dilated and tortuous right coronary artery.

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Figure 2 Selective injection into circumflex coronary ostium showing dilated and tortuous circumflex coronary artery.

OUTCOME AND FOLLOW-UP

Case 1

He remains well 2 years later.

Case 2

He remains symptom-free on furosemide 40 mg once daily 6 months after diagnosis.

DISCUSSION

Although small fistulae between the left anterior descending coronary artery in particular and the pulmonary artery are

common incidental findings during coronary angiography, larger fistulous connexions with other cardiac structures are rare. The reported prevalence is 0.1–0.2% of all patients undergoing coronary angiography.² Fistulae are usually congenital, and thought to be due to incomplete closure of the sinusoids which normally connect the coronary arteries to the great vessels and chambers of the heart during embryological development³; occasionally acquired fistulae can develop, when they are commonly iatrogenic.^{3 4}

Coronary fistulae originate from the right coronary artery in approximately 50–58% of cases,⁵ the circumflex in 18.3% of cases and can involve both coronary arteries in 5% of cases. The most common site of drainage is the pulmonary artery in 29.8–43% and the right atrium in 20% of cases.^{4 6 7}

Patients with fistulae most commonly present with fatigue and dyspnoea. Patients may present with myocardial ischaemia due to coronary steal (3–7% of cases) or overt heart failure due to left-to-right shunt (19% of cases). Haemopericardium as a result of rupture of the fistula is rare. However, nearly half of patients with fistulae are asymptomatic at diagnosis.^{7 8}

The choice of investigation will be determined by patient presentation. Transoesophageal echocardiography and cardiac MR give similar detailed structural information of the heart, and coronary angiography allows more detailed study of the coronary tree which may be important in assessing suitability for surgery.

The management of these fistulae depends on their presentation and the magnitude of blood flow through the fistula. Surgical closure is possible in selected cases,⁷ but can be very high risk when the fistulous artery supplies a large amount of the ventricular myocardium.

Figure 3 Steady-state free precession MRI in the axial plane. The black arrow points to the proximal part of the dilated circumflex coronary artery. Ao=aorta; Desc Ao, descending aorta; LA, left atrium; RVOT, right ventricular outflow tract.

Figure 4 Steady-state free precession MRI in the short axis view at the level of the atrioventricular groove. The top black arrow points to the proximal part of the dilated circumflex coronary artery and the black arrow at the bottom of the image points to the coronary sinus just before it joins the right atrium. AOV, aortic valve; Desc Ao, descending aorta; LV, left ventricle; RVOT, right ventricular outflow tract.

Learning points

- ▶ Heart failure is not a final diagnosis.
- ▶ Patients presenting with heart failure should be investigated to find the underlying cause.
- ▶ Coronary artery fistulae can cause heart failure.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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