Case Report

Arteriovenous Connection Between the Aorta and the Coronary Sinus Through a Giant Fistulous Right Coronary Artery

Aristides Androulakis[1], Christina Chrysohoou[2], John Barbetseas[2], Stella Brili[2], Apostolos Kakavas[1], Dimitrios Maragiannis[2], Ioannis Kallikazaros[1], Christodoulos Stefanadis[2]

[1]Cardiac Department, [2]1st Cardiology Clinic, Athens University Medical School, Hippokration Hospital Athens, Greece

Key words:
Coronary arteriovenous fistula.

Coronary arteriovenous fistulas are rare. The right coronary artery (RCA) seems to be the most common site of origin, while the right ventricle, right atrium and the main pulmonary artery are the most common draining chambers. We report on a patient with fistulas in both coronary arteries: one arising from the circumflex artery and draining into the coronary sinus (CS), and a fistulous connection between the proximal aorta and the CS, represented by an enlarged and cirsoid RCA. Despite the fact that the non-invasive diagnosis of coronary fistulas has traditionally been difficult, in this case echocardiography led to the suspicion of the anomaly.

Manuscript received:
December 3, 2007;
Accepted:
December 10, 2007.

Address:
Aristides Androulakis
30 Dodekanisou St.
Vrilissia, 152 35 Athens, Greece
e-mail:
antaris@otenet.gr

Coronary arteriovenous fistulas are rare, with a reported incidence of 0.1-0.2% among all cardiac catheterisations and of 0.002% in the general population.[1,2] The right coronary artery (RCA) seems to be the most common site of origin, while the right ventricle (approximately 2/3 of cases), the right atrium and the main pulmonary artery are the most common draining chambers. We report on a patient with fistulas in both coronary arteries: one arising from the circumflex artery and draining into the coronary sinus (CS), and a fistulous connection between the proximal aorta and the CS, represented by an enlarged and cirsoid RCA.

Case presentation

A 60-year-old woman, with a history of smoking, arterial hypertension and diabetes mellitus under diet, was referred for evaluation of worsening angina and dyspnoea on exertion that had commenced a couple of years previously. She had been hospitalised for the past few days at another hospital because of severe angina triggered during an episode of atrial flutter. The electrocardiogram at that time showed more than 2 mm ST-segment depression in the anterior leads. Atrial flutter was self-terminated and she was treated with clopidogrel, b-blocker, nitrates, aspirin, and a statin thereafter. The electrocardiogram of the current hospitalisation revealed sinus rhythm with repolarisation changes in the precordial leads. The physical examination was unremarkable, with the exception of a mild systolic murmur at the cardiac apex; chest radiography was normal. No troponin elevations were detected during her hospitalisations. On echocardiographic examination, both the left and the right ventricle had normal dimensions and normal systolic function. Neither atrium was dilated. There was

trivial posterior mitral leaflet proptosis with mild mitral regurgitation. From the left parasternal short axis view the origin and the proximal part of a significantly dilated RCA were visualised (Figure 1), while from the left parasternal right ventricle inflow view, turbulent flow into the CS was demonstrated and increased blood flow to the right atrium was present on colour flow Doppler (Figure 2). No other congenital or acquired structural abnormality was detected.

On coronary arteriography, no atheromatous obstructive disease was present in any coronary artery. The circumflex artery, after giving rise to two considerable obtuse marginal branches, drained into the CS as a tortuous fistula (Figure 3). In addition, an enlarged, tortuous, dominant RCA was seen, representing a fistula presumably also draining into the CS (Figure 4). All the conventional RCA branches to the myocardium originated normally from the fistula. Subsequently, left and right cardiac catheterisation was undertaken at 85 beats/min. The pulmonary artery pressure was normal (26/10, mean 15 mmHg), as were the pulmonary capillary wedge and the left ventricular end-diastolic pressures (mean 12 mmHg). The oximetry run allowed for a calculated pulmonary-to-systemic flow ratio (Qp/Qs) of about 1.25.

In order to better visualise the fistulas,[3] we performed a cardiac magnetic resonance imaging (MRI) study that confirmed the drainage of the fistulous RCA into the CS. The diameter of this cirsoid vessel was calculated as 12 mm (Figure 5). MRI also verified that the Qp/Qs ratio was 1.3.

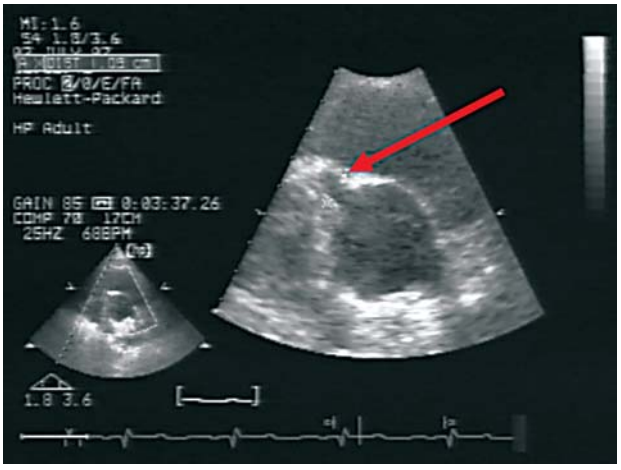

Figure 1. Left parasternal short axis view of the sinuses of Valsalva. The dilated origin of the right coronary artery (11 mm) is clearly visible at 11 o'clock (arrow).

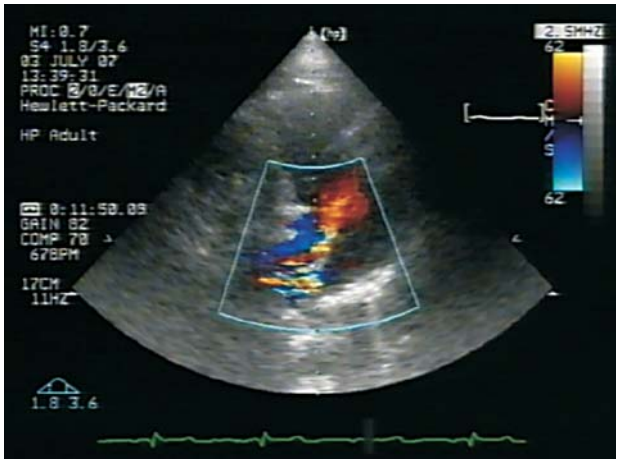

Figure 2. Left parasternal right ventricular inflow view, showing turbulent flow from the coronary sinus outflow into the right atrium.

Discussion

Most coronary arteriovenous fistulas are congenital in origin; few are acquired, due to trauma, or iatrogenic – after cardiac surgery or transcatheter interventions. Concomitant congenital anomalies occur in 40% of patients. Patients usually present symptoms after the third decade of life, such as exertional angina and/or dyspnoea, most often due to coronary flow 'steal' without obstructive atheromatous coronary artery disease, dyspnoea due to congestive heart failure in the case of significant right-to-left shunt, various arrhythmias, and syncope. Myocardial infarction, infective endocarditis, aneurysmal dilatation and rup-

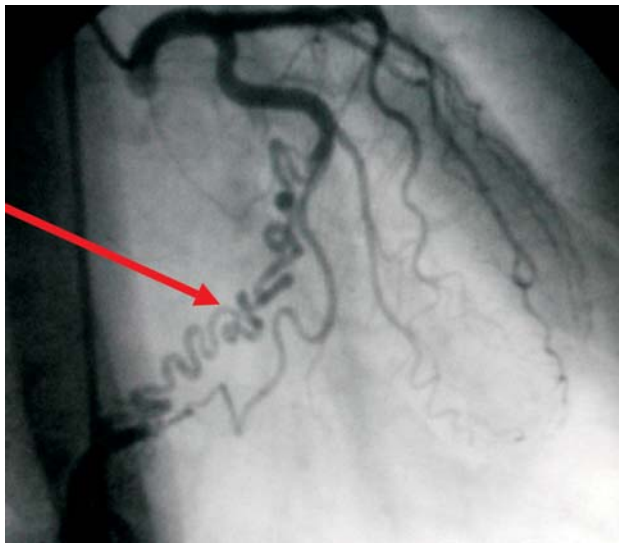

Figure 3. Right caudal oblique view of the left coronary artery. The distal left circumflex artery drains into the coronary sinus as a fistula.

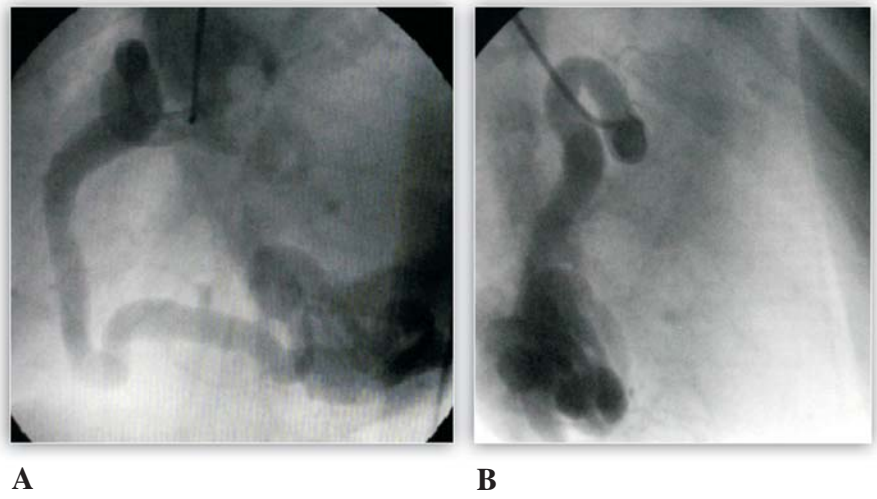

Figure 4. Left anterior oblique (A), and right anterior oblique (B) angiographic views of the fistulous right coronary artery.

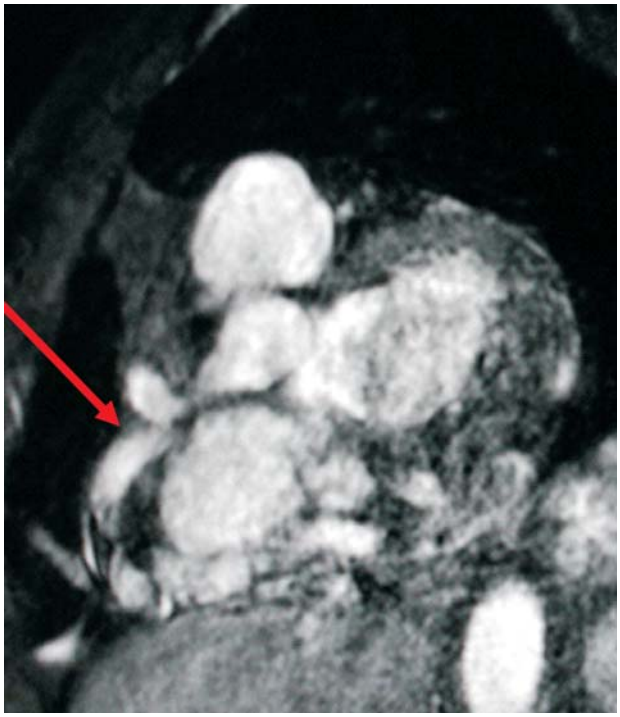

Figure 5. The large right coronary artery (arrow) as seen on magnetic resonance imaging.

ture may rarely develop. Diagnosis is almost always incidental during coronary arteriography. However, the most important issue about coronary fistulas has been the appropriateness of their therapy. It is generally believed that a symptomatic fistula has to be closed, either by surgical epicardial or endocardial ligation, or by an intravascular method with coil embolisation or occluder systems. All these interventions are considered to be rather safe, with a low perioperative morbidity and mortality.[3-5] Regarding asymptomatic fistulas, the decision might more accurately be driven by the severity of the left-to-right communication, or by any possible myocardial ischaemia due to the 'steal phenomenon' produced during an exercise test.

The interest of our case was the presence of two coronary fistulas in the same patient, arising from both the left and the right coronary arteries, while both drained into the CS. Also, despite the fact that the non-invasive diagnosis of coronary fistulas has traditionally been difficult, in this case echocardiography led to the suspicion of this anomaly (Figures 1, 2). Moreover, it is difficult to suggest invasive treatment in symptomatic patients like ours for the following reasons:

- the calculated Qp/Qs ratio was around 1.3 (by both MRI and heart catheterisation), while in addition the right cardiac chambers were not dilated and were functioning normally;
- her dyspnoea could be attributed to her smoking habit, and/or to left ventricular diastolic dysfunction due to her long-standing diabetic status and hypertension;
- intervention appeared difficult because the RCA (being a fistula itself) was dominant; occlusion systems or coils might compromise important posterolateral myocardial branches, while surgeons are not usually keen to intervene in the CS.

Cases similar, though not identical, to ours have already been described;[6,7] in both cases surgical consultation was suggested. We proposed that our patient should undergo an exercise thallium-201 myocardial perfusion scan, as well as a cardiopulmonary exercise test to measure maximum oxygen uptake and

the anaerobic threshold. However, she declined not only every intervention but also any further test.

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