Coronary arteriovenous fistula with associated aneurysm

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DESCRIPTION
A 56-year-old man was seen in cardiology clinic with exertional chest pain, most marked when walking uphill. He was a heavy smoker but had no other cardiovascular risk factors. His 12-lead ECG was normal, and a CT coronary angiogram (CTCA) was organised with a suspected diagnosis of angina. Echocardiography showed normal cardiac chambers with preserved biventricular systolic function.

The CTCA showed mild coronary artery disease in the distal left main stem and left anterior descending artery (LAD) which was non-obstructive (figure 1). An aberrant vessel arose from the diagonal artery and connected to the main pulmonary artery with evidence of minimal shunting. This vessel was tortuous and contained a small well-defined aneurysmal focus of 3 mm diameter. A conservative strategy with medical treatment was initiated and his symptoms settled. Follow-up in cardiology clinic over the next year has shown no progression in his symptoms, and cardiac MRI has not been organised as there has been no clinical need.

Coronary artery fistulas are rare congenital anomalies (~0.9% of the adult population) which are often discovered incidentally during coronary angiography and increasingly so during CTCA given good anatomical detail obtained.1 In rare cases, they can also arise due to iatrogenic reasons.2 They may arise from any of the three major coronary arteries; however, the right coronary artery and the LAD are the most common with the circumflex coronary artery rarely involved.3 A fistula between the diagonal artery and the main pulmonary artery, as in this case, is a rare finding.

Coronary artery fistulae can present with symptoms of angina, congestive heart failure or arrhythmia. Ischaemia can occur due to the coronary steal phenomenon where blood is shunted away from coronary beds to the low-pressure pulmonary circulation during diastole, resulting in ischaemia in the affected territories.3 The main indications for closure of coronary fistulas are clinical symptoms, particularly heart failure and myocardial ischaemia, and in asymptomatic patients with high-flow shunts, to prevent the occurrence of undesirable complications.

Treatment options include surgical repair and catheter embolisation.4 In the past, surgical repair was the preferred treatment; however, surgery has been associated with a higher level of fistula recurrence, and for this reason a catheter closure procedure is now the preferred treatment of choice. This procedure is not without risk, hence the conservative approach adopted in our case.

Learning points
► CT coronary angiography has become the gold standard for defining coronary anatomy; however, its use has led to increased incidence of congenital coronary artery anomalies.
► Coronary artery fistulas are rare congenital anomalies which can cause angina, heart failure and undesirable complications.
► All physicians should be aware of this rare condition, as successful treatment can lead to improvement in symptoms and reduction in morbidity.

Figure 1 CT coronary angiogram. (A) CT coronary angiogram three-dimensional (3D) tree view demonstrating an aberrant vessel arising from the diagonal artery (D1) with a tortuous bend and passing superiority. The black arrow shows a small well-defined aneurysmal focus within the vessel. (B, D, E) CT coronary angiogram maximum intensity projections in axial (B), coronal (D) and sagittal (E) views, demonstrating an aberrant vessel arising from the diagonal artery (D1) with a tortuous bend and mid-vessel aneurysm (arrows); connection is seen to the unenhanced MPA with a tiny jet of contrast just visible. (C) The 3D heart reconstruction showing the aberrant vessel arising from D1 with an associated small vessel aneurysm (white arrow). AO, aorta; LV, left ventricle; MPA, main pulmonary artery.
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