A 39-year-old man with no significant past medical history and no conventional coronary risk factors presented with a history of gradually progressive exertional dyspnea and a systolic heart murmur. Chest x-ray showed an enlarged cardiac silhouette with pulmonary plethora. Initial assessment by transthoracic echocardiography demonstrated mild mitral regurgitation but also revealed an enlarged coronary sinus with abnormal Doppler flow patterns in the right atrium. A subsequent transesophageal echocardiogram demonstrated a large, serpiginous coronary artery fistula from the dominant left circumflex artery to the coronary sinus causing aneurysmal dilatation of the left circumflex artery extending back to the left main stem (maximum cross-sectional diameter, 17 mm). No other congenital heart defects were identified. A 64-slice multidetector computed tomography (GE Lightspeed VCT, Chalfont St Giles, Buckinghamshire, UK) coronary angiogram confirmed the transesophageal echocardiographic findings and demonstrated that the left anterior descending and nondominant right coronary arteries were both unobstructed with no significant atherosclerotic disease. A treadmill exercise ECG stress test did not provoke any chest pain or ischemic ECG changes to stage 4 of the standard Bruce protocol. Cardiac magnetic resonance imaging revealed that the main pulmonary artery was dilated at 3.7 cm maximum dimension (aorta measured 3.2 in the same plane) with an estimated flow through the fistula of $\sim 2.7 \text{ L/min}$ causing a high output state with subsequent dilatation of both right and left ventricles (right ventricular end-diastolic volume, 290 mL; left ventricular end-diastolic volume, 330 mL).
After consideration at multidisciplinary review, the patient was referred for surgical ligation of the fistula in December 2008. Macroscopic examination at surgery revealed a series of fistulous communications at the obtuse margin and a massively dilated (≈1 cm diameter) venous portion entering the coronary sinus. The fistulous communication was mobilized and ligated. Measurement of oxygen saturation in blood specimens taken from the coronary sinus and main pulmonary artery confirmed a marked drop in saturations in postligation specimens compared with those taken before ligation, confirming a substantial reduction in the shunt flow rate; therefore, it was elected not to proceed to formal exclusion and grafting of the aneurysmal left circumflex artery. The patient made an uncomplicated postoperative recovery and was discharged 5 days later.

The patient was readmitted in May 2009 complaining of chest pain with T-wave inversion in the inferior ECG leads and a serum troponin T concentration of 0.11 ng/L (normal range, <0.01 ng/L). Percutaneous catheter angiography demonstrated that the circumflex artery aneurysm contained a large, partially occlusive intraluminal thrombus (Figure 1). Anterograde flow was demonstrated down the obtuse marginal branch, but no anterograde flow was evident down the more distal left posterior descending artery. Subsequent repeat multidetector computed tomography angiography (Figure 2) indicated some retrograde filling of the left posterior descending artery. The thrombotic occlusion of the left posterior descending artery was managed conservatively by anticoagulation with warfarin, and the patient’s symptoms resolved without further complication.

To the best of our knowledge, this is the only reported case of a thrombotic occlusion of a giant left circumflex artery aneurysm leading to an acute coronary syndrome, and the large thrombus burden is illustrated dramatically in the angiographic images. The exact mechanism of thrombus formation in the circumflex aneurysm and thrombotic occlusion of the left posterior descending artery in this case is unclear, but it seems reasonable to speculate that the substantial reduction in coronary flow down the aneurysmal vessel was a significant contributory factor. Possible therapeutic options in the management of coronary arteriovenous fistulae include transcatheter embolization1 and surgical correction. Surgical closure has been recommended as the gold standard and remains safe and effective with good reported success,2,3 but this case illustrates a potential complication associated with simple distal ligation of a fistulous connection arising from a grossly aneurysmal coronary artery. Therefore, we would recommend that either dual antiplatelet therapy or full anticoagulation should be considered after surgery in such circumstances.

Disclosures

None.

References

Thrombotic Occlusion of Giant Circumflex Artery Aneurysm After Ligation of Arteriovenous Fistula

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