The images presented here are from a 44-year-old asymptomatic man. At age 36 years, he experienced non-sustained atrial fibrillation. A year later, a transthoracic echocardiogram disclosed an enlarged left ventricle (diastolic dimension, 7.4 cm; ejection fraction, 56%) and a very large coronary sinus with Doppler color flow evidence of diastolic and systolic turbulence consistent with entry of a coronary arteriovenous fistula. Current selective coronary angiography visualized dilated circumflex and right coronary arteries, both of which entered an aneurysmal coronary sinus. An MRI during breath-hold acquisition further delineated the enlarged coronary sinus into which the circumflex and right coronary arteries drained (Figure 1). A 3-dimensional (3D) image set was then obtained by sequential MRI acquisitions during repeated breath-holding with surface reconstructions (Figure 2). The diameter of the proximal right coronary artery was 10 mm, and the diameter of the left main coronary artery was 11 mm (Figure 2, left). The circumflex artery was 11 mm in its proximal diameter, then abruptly widened to 18 to 20 mm and became very tortuous (Figure 2, left). The left anterior descending artery, by contrast, was 5 to 6 mm in diameter. The enlarged coronary sinus measured $8.5 \times 4.5 \times 3.5$ cm (Figure 2, right) and compressed the inferior portion of the left atrium. These images assisted in planning surgical closure of the coronary arteriovenous fistulas, at which time the right atrial appendage with a portion of the enlarged right atrium was excised and a maze procedure was performed. Two months after operation, the left ventricular diastolic dimension was 6.1 cm and the ejection fraction was 45%. Six months after operation, the left ventricular diastolic dimension was 5.4 cm and the ejection fraction was 63%. An exercise radionuclide myocardial perfusion scan was normal.

Only 5% of coronary arteriovenous fistulas arise from both right and left coronary arteries, and only 7% drain into the coronary sinus.1 Our patient is uncommon if not rare on both counts.

Reference

Figure 1. Coronal MRI through posterior portion of heart during breath-hold acquisition by a standard 2D cardiac-trigger MR angiographic technique. Coronary sinus (CS) is strikingly enlarged and indents left atrium (LA). Entry points of dilated circumflex and right coronary arteries are shown as “fish-mouths” (arrows). Circumflex coronary artery enters upper portion of dilated coronary sinus (upper arrow), and right coronary artery enters lower portion (lower arrow). RA indicates right atrium.

Figure 2. Left, By use of sequential MRI acquisitions in transaxial planes during repeated breath-holding, a 3D MRI data set was obtained. This figure is an anterior view of 3D surface reconstructions using manual tracing of cardiac structures. Dilated, tortuous circumflex (Circ) and dilated right (RCA) coronary arteriovenous fistulas enter an aneurysmal coronary sinus (CS). LAD indicates left anterior descending coronary artery; Ao, ascending aorta. Right, Posterior view using same technique. LV indicates left ventricle.
Arteriovenous Fistulas of the Circumflex and Right Coronary Arteries With Drainage Into an Aneurysmal Coronary Sinus
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