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Radiology Corner Case #11

Coronary Artery Fistula

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History

A 38-year-old man with a single episode of vague sub-sternal chest pain was referred for evaluation of possible coronary artery disease. His medical history was significant for hypertension and type-II diabetes mellitus. The patient had no prior history of thoracic trauma or surgery. During an exercise stress test, the patient reported chest pain; however, no electrocardiogram (EKG) changes were documented. A cardiac MRI axial black blood (Figure A) and black blood oblique (Figure B), and coronary x-ray angiogram oblique projections (Figures C and D) were performed.

Imaging Findings

MR imaging was performed using cardiac-gated, "black blood" double inversion recovery fast spin echo pulse sequence (DIR FSE). On DIR FSE images (axial, Figure 1A; oblique, Figure 1B), the left circumflex artery was identified by its black central lumen and noted to arise normally from the left main coronary artery. However, it was also noted to be ectatic and dilated along its course especially in its mid to distal segments within the left atrio-ventricular groove. Most importantly, the left circumflex coronary artery was noted to drain directly into the coronary sinus, consistent with the diagnosis of a left circumflex coronary artery to coronary sinus fistula. The presence and course of the fistula was confirmed on conventional x-ray angiogram (Figures 1C and 1D).

Diagnosis

Coronary Artery Fistula

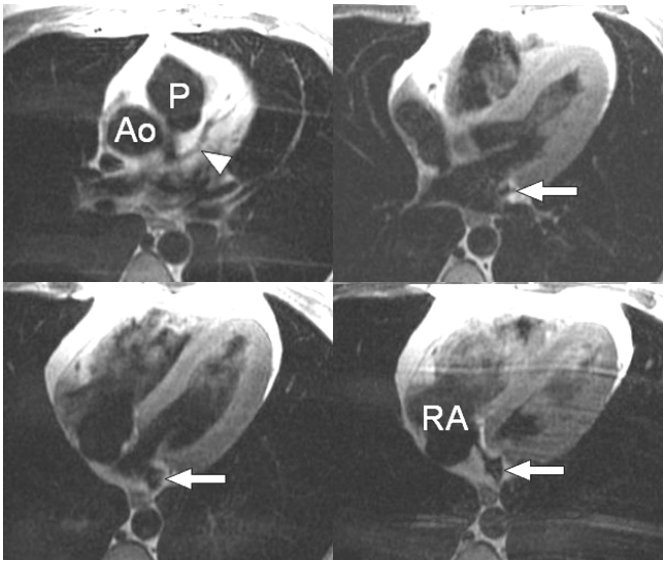

Figure 1A

Figures 1A. Axial black-blood double inversion recovery fast spin echo (DIR FSE; select images from top left through bottom right representing cranial to caudal) images through the thorax demonstrates normal origin of the left main coronary artery from the aorta and normal branching of the left anterior descending coronary artery (arrow head) from the left main coronary artery. On these images, the actual origin of the left circumflex coronary artery could not be confirmed; however, the majority of the left circumflex coronary artery (arrow) could be traced from its proximal to distal segments within the left atrio-ventricular groove. In particular, the left circumflex is noted to dilated distally (bottom images) and drain via the cavernous sinus into the right atrium (RA). Ao = ascending aorta; P = pulmonary artery

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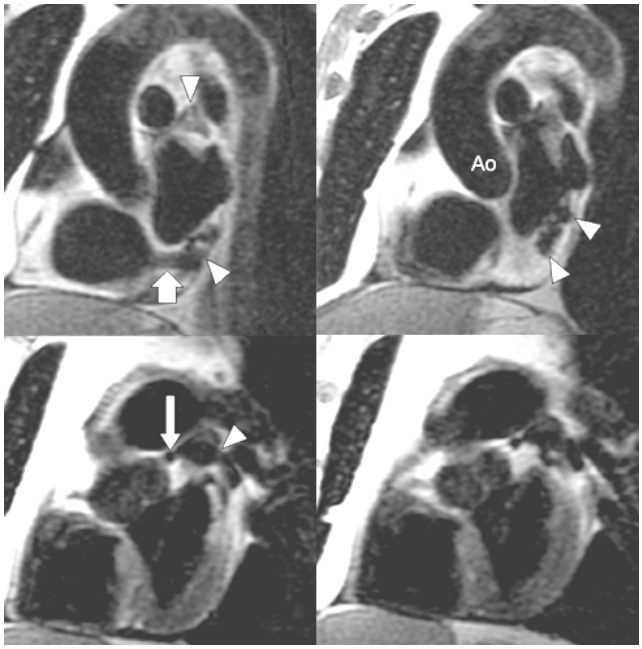

Figure 1B

Figures 1B. Double-oblique "short axis" black-blood DIR FSE images in a plane parallel to the left atrioventricular groove (top left to bottom right representing base to apex). The left circumflex coronary artery (arrowhead) is seen arising from the left main coronary artery (long thinner arrow, bottom left image) and draining into the coronary sinus (short wider arrow, top left image). The proximal and distal left circumflex coronary arteries were tortuous and dilated; but the mid left circumflex coronary artery was less well seen because of volume averaging with the left atrium and pulmonary venous confluence which were also dark. Ao = ascending aorta

Figure 1C

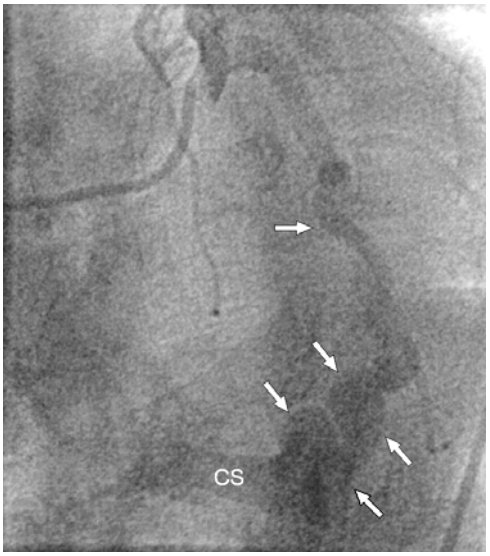

Figure 1D

Figures 1C-D. Select oblique conventional x-ray views (C, early phase; D, late phase) from a coronary catheterization study following injection of iodinated contrast into the left main coronary artery. On the early phase image (C), the large and tortuous left circumflex coronary artery (arrow) is seen coursing posteriorly overlying the posterior cardiac silhouette and the left anterior descending coronary artery (arrowhead) is noted to follow its normal course anteriorly. On the late phase image (D) the dilated distal left circumflex (arrows) is noted to communicate directly with the coronary sinus (CS), which drains into the right atrium (not shown on image).

Discussion

Coronary artery fistulas are uncommon; incidentally reported in 0.1% to 0.3% of patients undergoing coronary or cardiac angiography.[1-3] However, of coronary artery anomalies, coronary artery fistulas are one of the most common to be hemodynamically (and clinically) significant congenital coronary artery anomalies.[4, 5] Other congenital arterial lesions that can cause abnormalities of myocardial perfusion include anomalous coronary arteries (notably, those with inter-arterial course between the ascending aorta and pulmonary artery) and congenital coronary artery stenosis.

Most coronary artery fistulas are congenital but may also occasionally result from trauma such as gunshot, shrapnel, or stab injuries; or as a complication of a surgical procedure, such as coronary bypass surgery, mitral valve replacement, or myocardial biopsies after cardiac transplantation. [2, 3, 6, 7] Slightly more than half of all coronary artery fistulas involve the right coronary artery.[2-5, 8, 9] In a few cases (roughly 5% [4, 5, 9 - 11]), both left and right coronary arteries are involved. In roughly 85-90% of cases, coronary artery fistulas drain to the right side of the heart (i.e. coronary sinus, right atrium, right ventricle or pulmonary artery),[2-5, 8 – 11] thereby resulting in a left-to-right shunt. If the left-to-right shunt is large, blood may preferentially flow to the coronary fistula at the expense of the other coronary arteries, a phenomenon also known as "coronary steal." Coronary steal is particularly evident during times of stress and can result in myocardial ischemia to territories not supplied by the arterial fistula.[5, 8, 13]

Approximately half of individuals with a coronary artery fistula are asymptomatic, typically incidentally detected

during coronary angiography.[5, 8, 9] Though small fistulas can often go undetected in elderly patients, the likelihood and severity of symptoms generally increases with age if there is a left-to-right shunt. Symptoms when present can include angina, congestive heart failure, exertional dyspnea, bacterial endocarditis, and, even, acute myocardial infarction or spontaneous rupture with subsequent cardiac tamponade.[5, 8, 9, 12] The typical clinical sign for a coronary fistula is often non-specific, represented by a continuous heart murmur whose qualities vary based upon its location and size.[1, 2, 5, 8, 9, 12]

On chest radiographs, patients may have cardiomegaly or an abnormality of the cardiac contour;[5, 8, 10] however, these features are not distinctive and may be absent. The key to diagnosing a coronary fistula is proper identification of an anomalous communication between the donor coronary artery and the recipient cardiac chamber or structure. Since most fistulas result in a left-to-right shunt and increased blood flow, the coronary artery will often be dilated and tortuous as in our case. Doppler color flow mapping,[12, 14-16] transesophageal or transthoracic echocardiography,[16, 18] contrast-enhanced electron beam tomography,[18, 19] magnetic resonance imaging,[16, 20] and radionuclide cardioangiograms[13] have been proven to accurately locate the sites of origin and drainage of coronary artery fistulas. However, the presumptive gold standard is still coronary angiography.[4, 8, 15] With the increased performance of thoracic MR, especially cardiac MR, it is important to be aware of this lesion as a potential additional source of chest pain in younger adults, typical of the active duty military population.

Surgical ligation should be considered for patients with hemodynamically significant coronary artery fistulas.[1, 10] With age, worsening of a left-to-right shunt can result in the complications of coronary steal, congestive heart failure, angina, and endocarditis.[2, 4, 12]

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