A 52-year-old former recreational marathon runner with a history of permanent atrial fibrillation was referred to our institution because of fatigue and shortness of breath. His 12-lead ECG indicated atrial fibrillation with incomplete right bundle-branch block and inferolateral T-wave inversions (Figure 1). The chest x-ray showed an abnormal structure with a circular silhouette at the projection of the right atrium in the anterior-posterior view (Figure 2). Transsthoracic echocardiography revealed a vascular tubular structure adjacent to the atrioventricular groove (Figure 3A and 3B and Movies I and II in the online-only Data Supplement). Subsequently, we performed a coronary computed tomography angiography (CCTA) using a 256-slice multidetector-row CT (Philips Brilliance iCT, Best, The Netherlands) with a tube voltage of 100 kV and a tube current of 300 mA. Because of the atrial fibrillation (mean heart rate, 57 bpm; range, 45–110 bpm), an arrhythmia detection algorithm was used during the prospective ECG-triggered image acquisition. The CCTA depicted a normal left coronary system with no signs of atherosclerosis. The ostium of the right coronary artery (RCA) was dilated (10×8 mm), and the proximal segment of the vessel formed a giant aneurysm (Figure 4A and 4B). The location of the aneurysm was noted to be anterior to the right atrium, adjacent to the atrioventricular groove, and its size measured 62×60×86 mm (Figure 4C). The length of the whole RCA was 80 cm along its centerline. Subsequent invasive coronary angiography confirmed the CCTA findings (Movies III and IV in the online-only Data Supplement). Surgery was performed to repair the RCA and to stop the shunt. During the surgical procedure, the aneurysm was resected (Figure 5A–5C), the RCA was reimplanted into the aorta (Figure 5D), and the fistula was ligated at the orifice of the coronary sinus. Because of the permanent atrial fibrillation, the maze procedure and left auricle ligation were also performed, but restoration of the sinus rhythm was unsuccessful. The postoperative observation period was uneventful. At the 6-week regular follow-up examination, the patient was asymptomatic with normofrequent atrial fibrillation. Five months after the surgical procedure, the patient presented with cough, night sweats, and low-grade fever, which had persisted over the previous week. The chest x-ray was unremarkable (Figure 6A). The urgent chest CT examination showed lesions with central low attenuation and ring-like enhancement behind the left ventricle, which raised the possibility of an abscess (Figure 6B). Cardiac magnetic resonance imaging (1.5 T, Philips Achieva, Philips Medical Systems, Best, The Netherlands) demonstrated mildly elevated left and right ventricular volumes with preserved left and right systolic ventricular function (both left ventricular and right ventricular ejection fraction, 55%). The cine images showed no regional or segmental wall motion abnormalities. The tortuous distal two thirds of the RCA was completely filled with thrombus (Figure 7A and Movie V in the online-only Data Supplement). The luminal signal was inhomogeneous on both T1-weighted black-blood turbo spin echo (Figure 7B) and T2-weighted fat-suppression using spectral inversion recovery sequences (Figure 7C). On the diffusion-weighted imaging, the thrombosed lumen showed only a mildly increased variable signal instead of marked diffusion restriction pathognomonic to an abscess (Figure 7D). The phase-sensitive inversion-recovery early-enhancement series confirmed the lack of contrast uptake by the thrombus (Figure 7E). Interestingly, the coronary artery wall was thickened and demonstrated marked late enhancement (Figure 7F). On the basis of these findings, the symptoms were most probably caused by the inflammation of the large coronary artery filled with thrombotic material. The presence of thrombotic content and the inflammation of the coronary artery wall resemble the magnetic resonance imaging appearance of thrombophlebitis. However, in this case, a coronary artery was affected by the inflammatory process; therefore, thrombovasculitis might be a more appropriate term to describe this pathology. Considering the local nature of the inflammatory process, we initiated 500 mg/d acetylsalicylic acid therapy 3 times daily. The patient became symptom free in 1 week.

One year after the resection, the patient presented at our institution for a follow-up CCTA. The atrial fibrillation still persisted, but no other symptoms were present. The CCTA...
showed an RCA stump (11×11×12 mm) with contrast filling. Distal to the stump, the tortuous vessel was occluded (Figure 8A–8C). At the 3-year follow-up visit, the patient was asymptomatic with an excellent physical condition.

Discussion
Coronary artery aneurysm is a rare condition detected in 0.3% to 5% of patients undergoing coronary angiography. Coronary artery aneurysm is defined as “coronary dilatation with a diameter of ≥1.5 times the normal coronary artery.” The proximal and the middle segments of the RCA are most frequently affected. Coronary aneurysms are usually asymptomatic. Symptoms such as chest pain, dyspnea, or palpitation can be attributable to the myocardial ischemia. It has previously been suggested that various pathological conditions such as Kawasaki disease can lead to the development of coronary aneurysms. Coronary artery fistulas can also increase the risk of aneurysm formation. Fistulas drain to the right atrium in 41% of the cases and to the coronary sinus in only 7%.

No specified protocols exist for the clinical care and treatment of coronary aneurysms. Cardiac magnetic resonance and CT not only may provide superior visualization of the aneurysms but also may support the assessment of potential complications during the follow-up period.

To the best of our knowledge, this case represents the largest reported RCA aneurysm with a fistulous connection to the coronary sinus evaluated by multiple imaging modalities. In addition, we believe that this is the only reported case in the literature that describes coronary artery thrombovasculitis as depicted by cardiac magnetic resonance.

Disclosures
None.

References

Figure 1. Twelve-lead surface ECG shows atrial fibrillation with incomplete right bundle-branch block and inferolateral T-wave inversion.

Figure 2. Chest x-ray demonstrates an abnormal structure with a circular silhouette (arrows) at the projection of the right atrium in the anterior-posterior view.
Figure 3. Transthoracic echocardiography image of the parasternal short-axis view at the level of the mitral valve (A) shows a complex tubular structure behind the ventricles, which corresponds to the dilated and tortuous right coronary artery (RCA; arrows). The apical 2-chamber view (B) also demonstrates several cross sections of the tortuous RCA (arrows) adjacent to the inferior wall of the left ventricle.

Figure 4. Maximum-intensity projection images in the coronal (A) and axial (B and C) views obtained from prospective ECG-triggered computed tomography angiography showing dilated ostium (arrows) of the giant aneurysm (asterisk) and its course toward the coronary sinus (arrowheads). Volume-rendered images (D, E, and F) depict the giant right coronary artery (RCA) aneurysm, which originates from the ascending aorta, and its location is anterior to the right atrium, adjacent to the atrioventricular groove. Distal to the aneurysm, the RCA remained enlarged and tortuous. LAD indicates left anterior descending artery.
Figure 5. The intraoperative surgical image obtained after sternotomy shows the giant right coronary artery (RCA) aneurysm (A, arrow). The surgical forceps are pointing to the aneurysm (B). Postoperative image shows the removed RCA aneurysm (C). After the resection, the RCA was reimplanted into the aorta (D).

Figure 6. Chest x-ray shows normal anatomy of the heart with postsurgical stitches (A). Acute chest computed tomography demonstrates ring-like enhancing lesions (arrows) with central low attenuation behind the left ventricle (B).

Figure 7. Diastolic phase of an axial cine balanced steady-state free-precession acquisition (A). Black-blood T1-weighted turbo spin echo image at the same location (B). Black-blood T2-weighted spectral inversion recovery image (C). In the diffusion-weighted axial image, the thrombosed lumen shows only mildly increased variable signal instead of marked diffusion restriction pathognomonic to an abscess (D). Axial early-enhancement image captured 3 to 4 minutes after administration of gadolinium-D03A-butrol using an intentionally long inversion time (E). An axial late-enhancement image was taken ≈12 minutes after contrast administration (F). The white arrows point to the thrombosed lumen of the tortuous ligated right coronary artery, adjacent to the coronary sinus. The black arrow in E points to the luminal thrombus showing lack of contrast uptake. In F, the central signal in the thrombus can be attributed to its long relaxation time (white arrow); the coronary wall demonstrates late enhancement related to the inflammatory process in the vessel wall (black arrow).
Figure 8. Maximum-intensity projection images in the axial, sagittal, and coronal views demonstrate the right coronary artery stump with contrast filling (white arrows) and distal to it the thrombosed lumen (arrowheads).
Multimodality Imaging of Giant Right Coronary Aneurysm and Postsurgical Coronary Artery Inflammation

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