Giant Left Circumflex Coronary Artery Aneurysm With Arteriovenous Fistula to the Coronary Sinus

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An 80-year-old woman with a history of breast cancer status post radiation therapy, paroxysmal atrial fibrillation, and congestive heart failure was referred to our institution for evaluation of a giant left circumflex (LCx) coronary artery aneurysm with fistulous communication to the coronary sinus. The patient initially presented with shortness of breath and non-ST elevation myocardial infarction associated with anterolateral T-wave inversion on ECG (Figure 1). Chest radiography showed a dense structure with a circular silhouette at the projection of the superior right mediastinum at the location of the right atrium in the posterior-anterior view and in the posterior mediastinum in the lateral view (Figure 2). She underwent cardiac catheterization, which showed no significant obstructive epicardial coronary artery disease. However, the angiogram revealed a large LCx coronary artery aneurysm with fistulous communication to the coronary sinus (Figure 3A and 3B; online-only Data Supplement Movies I and II). To better define the anatomic relationship of this aneurysm, a contrast-enhanced 64-slice multidetector computed tomography (MDCT) was performed. The location of the aneurysm was noted to be posterior to the left ventricle in juxtaposition with the left atrium, and its size measured 6.0 cm × 5.6 cm × 4.8 cm (Figure 4A through 4E; online-only Data Supplement Movie III). An intraoperative transesophageal echocardiogram was performed and confirmed the presence of the aneurysm and LCx fistula to the coronary sinus.
The patient underwent successful surgical resection of the LCx aneurysm with ligation of the LCx just proximal to the aneurysm (Figure 6A and 6B). The outflow of the aneurysm was ligated at the coronary sinus. Postoperative intraoperative transesophageal echocardiogram demonstrated residual ectasia of the LCx coronary artery proximal to the site of ligation and no evidence of coronary aneurysm or fistulous communication with the coronary sinus (online-only Data Supplement Movie VI). Surgical pathology revealed atheromatous changes in the vessel wall of the aneurysm with extensive fibrosis and calcification (Figure 6C). Her postoperative course was uneventful, and she has remained well at 4-month follow-up.

Coronary artery fistulous communication is rare, with a reported incidence of 0.1 to 0.2%. There have only been a few reported cases of aneurysmal circumflex coronary arteries with fistulous connection to the coronary sinus. Our case represents the largest reported LCx coronary aneurysm reported in association with fistulous connection between the circumflex coronary artery and the coronary sinus. Symptoms on presentation may include shortness of breath, fatigue, angina, and infective endocarditis. There remains no standard of management or clinical algorithm for this rare pathology, though diagnosis and treatment may stand to benefit from improved cardiac imaging modalities. MDCT may provide superior visualization of aneurysmal coronary fistulas and its anatomic relationship to the adjacent structures and could help guide surgical or endovascular treatment strategies.

Acknowledgments
We thank the Massachusetts General Hospital Echocardiography and Pathology laboratories for providing great clinical care.

Disclosures
Dr. Truong has received support from National Institutes of Health grant T32HL076136. The other authors report no conflicts.

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Figure 3. A, Conventional coronary angiography of the left coronary arterial system in the right anterior oblique caudal projection showed left main and circumflex coronary artery ectasia with a serpiginous course that subsequently becomes a large LCx aneurysm (asterisk). B, Left anterior oblique projection demonstrated the aneurysm (asterisk) as well as the inflow from the LCx coronary artery and the outflow fistulous communication and drainage into the coronary sinus.

Figure 4. A, Transaxial image from a contrast-enhanced MDCT showed the large eccentric aneurysm (asterisk) of the distal LCx coronary artery at the left atrioventricular groove. Its position is posterior to the left ventricle (LV) and posterolateral in the thorax. Heterogeneous contrast opacification is present within the aneurysm, which correlates well with the contrast twirling seen in the coronary angiogram. LA indicates left atrium; RA, right atrium; and RV, right ventricle. B, Maximum intensity projection image from the multidetector computed tomography demonstrated the course of the inflow of the dilated left main (black arrowhead) and LCx coronary artery (white arrowheads) into the aneurysm (asterisk) and its outflow drainage into the coronary sinus (arrows) and RA. The ectatic left main coronary originated normally from the left sinus of Valsalva of the ascending aorta (Ao). Minimal mural calcifications can be seen within the aneurysm near the ostia of the inflow and outflow. C through E, Volume-rendered 3-dimensional images of the MDCT showed the LCx aneurysm (asterisk), which arises from the dilated left main coronary artery and originates from the ascending aorta (C), its arteriovenous fistulous inflow and outflow course (D), and its drainage into the coronary sinus (E).
Figure 5. A, Intraoperative transesophageal echocardiography depicted the large LCx arteriovenous fistulous aneurysm (asterisk) with inflow from the dilated LCx coronary artery (arrowheads) and outflow into the coronary sinus (arrows). LA denotes left atrium. B, Intraoperative transesophageal echocardiogram with color Doppler flow demonstrated inflow from the dilated LCx coronary artery into the large LCx arteriovenous fistulous aneurysm (asterisk). The blue Doppler signal depicted blood flowing away from the transducer, whereas the orange Doppler signal represented blood flowing toward the transducer.

Figure 6. A and B, Intraoperative surgical images obtained after sternotomy showed LCx aneurysm (asterisk). The surgical forceps are pointing to the dilated fistulous connections (B). C, Histology of the aneurysmal segment showed atheromatous changes, fibrosis, and calcification of the wall.
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Circulation. 2008;118:2304
doi: 10.1161/CIRCULATIONAHA.108.781617

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/118/22/2304

Data Supplement (unedited) at:
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