ANATOMIC ANOMALIES

Multimodality Imaging of a Giant Circumflex Check for updates Coronary Artery Aneurysm in a Septuagenarian

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INTRODUCTION

A giant coronary artery aneurysm is defined as an aneurysm measuring >8 mm.¹ Circumflex artery aneurysms are rare.² To date only 14 cases of giant circumflex artery aneurysm have been reported. These have been identified in infants as young as 6 months and in the elderly.^{3,4} We report a giant circumflex artery aneurysm with a fistula to the coronary sinus in an elderly woman, imaged on transesophageal echocardiography and multidetector cardiac computed tomography.

CASE PRESENTATION

A 74-year-old woman presented with dyspnea and chest pain of 1 month in duration. Her cardiac examination revealed a grade 2 continuous murmur and pansystolic mitral and tricuspid regurgitation murmurs. Her blood biochemistry was normal, including normal highsensitivity troponin levels. She had mildly elevated total cholesterol. Chest radiography revealed an enlarged cardiac silhouette. Findings on 12-lead electrocardiography were normal except for occasional atrial ectopic beats. Transthoracic echocardiography showed biatrial enlargement and mildly reduced left and right ventricular systolic function, with a left ventricular ejection fraction of 50% and tricuspid annular plane systolic excursion of 16 mm. The right ventricular enddiastolic diameter at the base was 30 mm. There was moderate mitral regurgitation secondary to myxomatous disease and moderate tricuspid regurgitation secondary to pulmonary hypertension. An aneurysm was suspected on the basis of a large pulsatile structure in the atrioventricular groove, but transthoracic echocardiography failed to provide further details regarding origin and course. Therefore, transesophageal echocardiography was performed. A large aneurysm involving the circumflex artery was noted to arise in the left atrioventricular groove; it had a beaded tortuous appearance, and continuous flow was noted in the structure on Doppler study (Figures 1A-1C). The aneurysm was pulsatile in nature, drained into the coronary sinus through a fistula, and compressed the lateral left atrial wall (Figures 2A and 2B, Videos 1 and 2).

The patient underwent multidetector cardiac computed tomography, which revealed a giant tortuous left circumflex coronary artery

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VIDEO HIGHLIGHTS

Video 1: Mid transesophageal echocardiographic view depicting flow through the giant circumflex artery aneurysm with compression of the left atrium.

Video 2: Three-dimensional transesophageal echocardiography showing compression of the left atrium by the circumflex artery aneurysm.

View the video content online at www.cvcasejournal.com.

(Figures 3A-3D and 4A-4C). The left anterior descending and right coronary arteries were normal. There was dilatation of the entire circumflex artery from the ostium to the distal artery. The distal artery was aneurysmal (56.8 \times 10.9 mm) as it drained directly into the coronary sinus (Figure 3D). The arteriovenous fistula resulted in a left-toright shunt without severe right ventricular dilatation, suggesting restriction of flow through the distal circumflex to the fistula, thus likely accounting for the aneurysmal circumflex dilatation. The beaded appearance was caused by the extreme tortuosity being viewed in cross-section (Figure 3A). The pulmonary hypertension noted was likely secondary to the mitral regurgitation and the leftto-right shunt to the arteriovenous fistula. The patient declined invasive coronary angiography or ischemia testing. She declined any invasive intervention and was initiated on medical therapy with a β -blocker, a loop diuretic, aspirin, and a statin.

DISCUSSION

This case highlights a coronary artery aneurysm as a rare cause of chest pain successfully defined using multimodality imaging. Giant coronary artery aneurysms can be congenital or secondary to atherosclerotic disease, inflammatory disorders, connective tissue disease, infections, and trauma.^{2,5-7} These aneurysms can be silent for long periods, with patients presenting as late as the eighth decade of life.⁸ They usually come to attention secondary to symptoms of heart failure, angina, myocardial infarction, thromboembolism, or hemopericardium.² Chest pain and dyspnea are usually a result of high flow state through the fistula, coronary steal phenomenon, or direct compression of the coronary arteries by the aneurysm.² The dilated, tortuous artery in this case likely resulted from high pressure in the artery draining into a restrictive distal fistula, with a resultant low-volume left-toright shunt.

Multimodality imaging provides excellent assessment of coronary artery aneurysms.^{9,10} Transthoracic echocardiography is useful for detection of the aneurysm and assessment of left and right ventricular function and hemodynamic evaluation, but



Figure 1 Transesophageal echocardiographic short-axis view showing beaded appearance of tortuous left circumflex coronary artery without color flow (**A**, *arrow*) and with color flow (**B**, *arrow*). Transesophageal echocardiographic view showing continuous flow through the circumflex artery aneurysm secondary to a fistula between the artery and the coronary sinus (**C**).



Figure 2 Mid-Transesophageal echocardiographic view depicting giant circumflex artery aneurysm (GCA) compressing the lateral wall of the left atrium (A). (B) Flow into coronary sinus through a fistulous connection (*left, arrow*) and a comparative image of the fistula without color (*right, arrow*).

transesophageal echocardiography allows a more detailed anatomic assessment of the aneurysm. Multidetector cardiac computed tomography provides useful information about the size, extent, and relationship of the aneurysm to adjacent structures. Important differential diagnoses on cardiac imaging include tumors, pseudoaneurysms of the coronary artery, pericardial cysts, and sinus of Valsalva aneurysms.^{11,12}

CONCLUSION

Management of patients with giant circumflex artery aneurysms has been varied and should be individualized depending on local expertise and patient preference.^{5,8} Options include watchful follow-up, anticoagulation, percutaneous stenting or coil place-

ment, surgical resection, and coronary artery bypass grafting.⁹ In general, surgical management is suggested when the likelihood of complications such as rupture, thromboembolism, and cardiac tamponade is high. Multimodality imaging studies are essential for diagnosing this rare case of a giant circumflex aneurysm with a fistula to the coronary sinus as a rare cause of chest pain in a septuagenarian.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi. org/10.1016/j.case.2020.02.004.



Figure 3 Multidetector cardiac computed tomographic image showing large circumflex artery aneurysm (**A**, **B**, *arrow*) draining into right atrium through the coronary sinus (**A**, *arrow*). (**C**) Tortuous left circumflex artery analogous to transesophageal echocardiography. (**D**) Large circumflex artery aneurysm measuring 5.68×10.92 mm in size draining into the coronary sinus.



Figure 4 Multidetector cardiac computed tomographic three-dimensional volume-rendered images showing origin of the tortuous circumflex (**A**, *vertical arrow*) from the left main coronary artery and its course in the left atrioventricular groove alongside the left anterior descending coronary artery in the anterior interventricular groove (**A**, *horizontal arrow*), its termination into a large aneurysm (**B**, *arrow*), and finally drainage into the coronary sinus posteriorly, adjacent to the great cardiac vein (**C**, *arrow*).

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