A Rare Case of a Giant Coronary Sinus with Focal Aneurysm Secondary to Multiple Fistulous Connections Arising from a Dilated, Tortuous Left Circumflex Coronary Artery

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INTRODUCTION

Coronary arteriovenous fistula (CAF) is an abnormal connection directly linking one or more coronary arteries to either a cardiac chamber or major vessel without an interposed capillary bed.[1] CAFs are uncommon and are usually an incidental finding, with drainage into the coronary sinus being one of the least prevalent forms.[2] Two-dimensional and three-dimensional transthoracic echocardiography (TTE) often provides the initial diagnosis, along with identifying associated defects and hemodynamic changes, with additional multimodality imaging used to delineate the origin and termination of CAF. We describe a rare and incidental case of two fistulous connections between the distal left circumflex coronary artery (LCx) and a grossly dilated coronary sinus.

CASE PRESENTATION

A 55-year-old woman was referred to our institution with a history of paroxysmal supraventricular tachycardia. On presentation, the patient was in sinus rhythm, with no electrocardiographic evidence of preexcitation (heart rate 60 beats/min), and her blood pressure was 120/70 mm Hg. The patient denied chest pain and was usually fit, although in the 6 weeks before presentation, she had noticed more dyspnea than usual. On physical examination, no cardiac murmurs, right ventricular heave, or pedal edema were detected. Chest radiography revealed an increased cardiothoracic ratio. The patient had been commenced on metoprolol.

TTE showed a severely dilated left ventricle for the patient's body size (indexed left ventricular end-diastolic volume 101 mL/m^2), preserved systolic function (ejection fraction 57%), and normal global longitudinal strain (21%; normal range >20%). Right ventricular size was mildly increased, with normal systolic function (fractional area change 45%, RV S′ 15 cm/sec, tricuspid annular plane systolic excursion 29 mm). Both the left and right atria were dilated (85 and 49 mL/m^2, respectively) with a normal relative atrial index of 0.70 (normal range <1.0).[3] The coronary sinus appeared grossly dilated (51 × 55 mm), with a larger focal aneurysmal region (normal approximately 10 mm in diameter and 20–30 mm in length).[4,5] Color flow Doppler demonstrated continuous turbulent flow draining into the coronary sinus from a dilated LCx (Figure 1, Videos 1–5). The main pulmonary artery and its branches appeared dilated, and there was mild tricuspid regurgitation (estimated right ventricular systolic pressure 37 mm Hg, assuming right atrial pressure of 8 mm Hg; pulmonary vascular resistance 1.6 Wood units). The ratio of pulmonary to systemic flow could not be accurately calculated. There was no evidence of a sinus venosus atrial septal defect or persistent left superior vena cava. Transthoracic three-dimensional echocardiography demonstrated the dilated LCx artery draining into the coronary sinus aneurysm, posterior to the left ventricle (Figure 2, Video 6).

Cardiac magnetic resonance confirmed a large CAF, measuring 11 mm in diameter, arising from the LCx, tracking tortuously and inserting into a severely dilated coronary sinus (57 × 57 mm), with the possibility of two sites of connection (Figure 3, Videos 7 and 8). The coronary sinus appeared roofed and drained normally into the right atrium, with no evidence of persistent left superior vena cava. There was biventricular dilatation (indexed left ventricular end-diastolic volume 152 mL/m^2, indexed right ventricular end-diastolic volume 160 mL/m^2), with preserved biventricular systolic function. The ratio of pulmonary to systemic flow could not be calculated.

Computed tomographic coronary angiography revealed the true extent of the marked dilatation and tortuosity of the left main coronary artery and LCx. This imaging modality also confirmed two fistulous connections between the LCx and the coronary sinus. The proximal connection was larger, measuring 4 cm in diameter, connecting proximal to the coronary sinus ostium (opening 9 × 8 mm). Additionally, a fistulous connection by a tiny vessel was noted coursing from the distal LCx to the coronary sinus (~3.3 cm proximal to the ostium). There was corresponding dilatation of the coronary sinus with focal aneurysmal dilatation (57 × 56 × 69 mm maximum dimension; Figures 4 and 5, Video 9). Computed tomography was a useful imaging modality to aid in planning a possible intervention for this patient.

Cardiac catheterization (Video 10) was performed to determine the possibility of percutaneous closure of the fistulous connections. The coronary anatomy was right dominant and the left anterior descending coronary artery and LCx were dilated, with a coronary sinus

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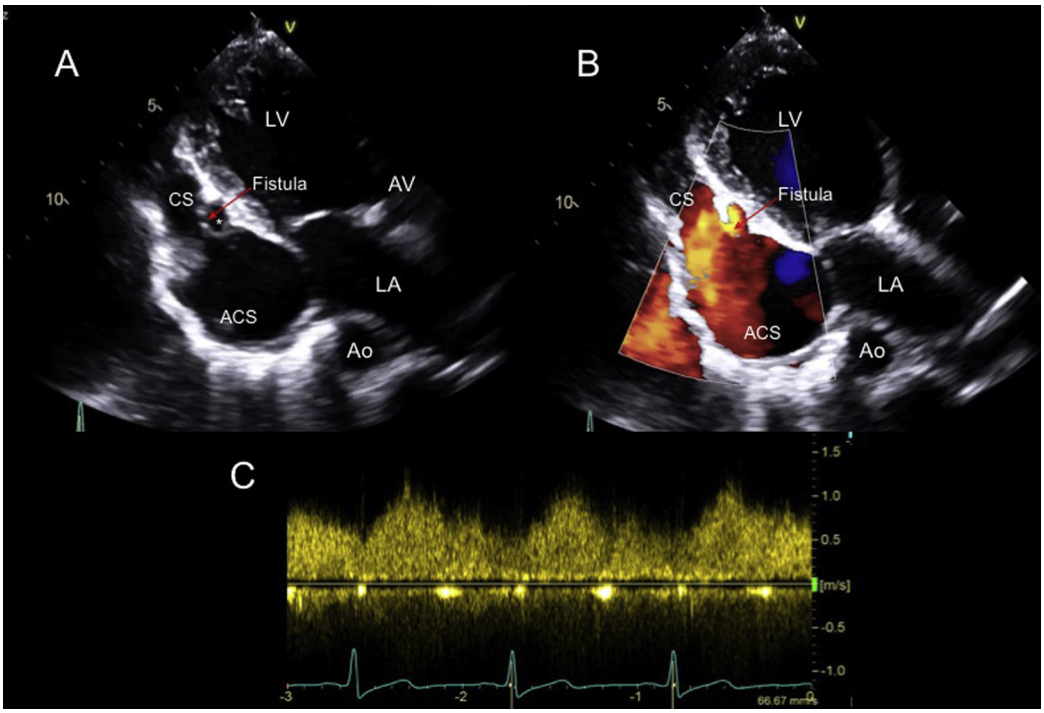

Figure 1 **(A)** Two-dimensional transthoracic apical long-axis view shows the dilated LCx (*asterisk*) draining into a dilated coronary sinus (CS, *arrow*). **(B)** Color Doppler echocardiography from the same view shows abundant and turbulent blood flow draining into the CS from the coronary artery fistula. **(C)** Continuous-wave Doppler demonstrates the continuous flow into the CS with a peak velocity of 1.1 m/sec. *ACS*, Aneurysmal coronary sinus; *Ao*, aorta; *AV*, aortic valve; *LA*, left atrium; *LV*, left ventricle.

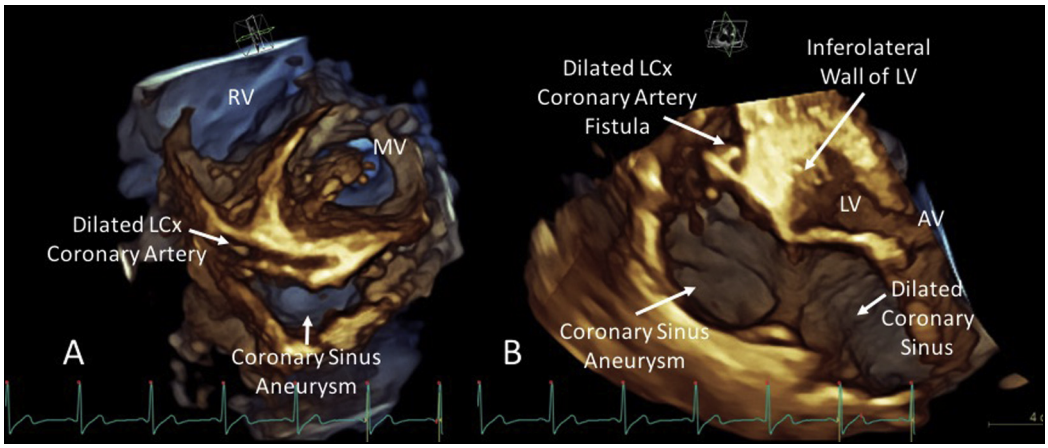

Figure 2 **(A)** Three-dimensional transthoracic echocardiographic image cut plane through the short-axis view of the left ventricle (LV). **(B)** Three-dimensional transthoracic echocardiographic image cut plane through the apical long-axis view. *AV*, Aortic valve; *LV*, left ventricle; *MV*, mitral valve; *RV*, right ventricle.

fistula confirmed. Although it was deemed technically possible to percutaneously close the fistulous connections, because of the risk for stasis and thrombosis, surgical resection of the large coronary sinus aneurysm was recommended.

DISCUSSION

With an incidence of 0.002% in the general population,[1,2,6-8] congenital CAFs arise from persistence of intratrabecular sinusoids between the coronary artery and coronary sinus.[2,9] CAFs vary widely in morphologic appearance and most commonly originate from the right coronary artery but may also arise from the left coronary artery or its branches.[2,10] The LCx is rarely involved.[11-13] Drainage into the right ventricle and pulmonary artery are generally seen,[2,3] with one of the least common draining sites being the coronary sinus (1.6%).[2,8,10] LCx–to–coronary sinus fistula causing marked dilatation to the coronary sinus as well as chamber enlargement from significant left-to-right shunting presented in this case is a rare occurrence. Differential diagnosis of dilated coronary sinus including right ventricular volume and pressure overload, persistent left superior vena cava,

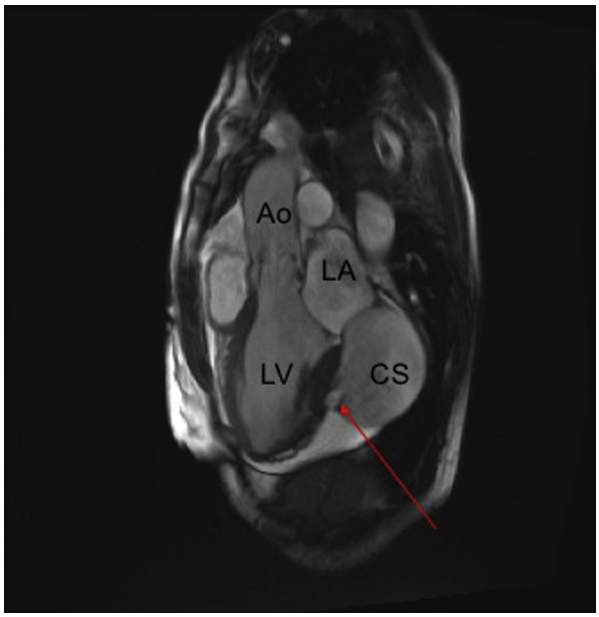

Figure 3 Cardiac magnetic resonance image obtained on a 1.5-T Siemens Aera using multiplanar cine steady-state free precession, flow imaging, navigator-gated noncontrast magnetic resonance coronary angiography shows a large coronary fistula (11 mm in diameter) arising from the left main coronary artery (*arrow*). *Ao*, Aorta; *CS*, coronary sinus; *LA*, left atrium; *LV*, left ventricle.

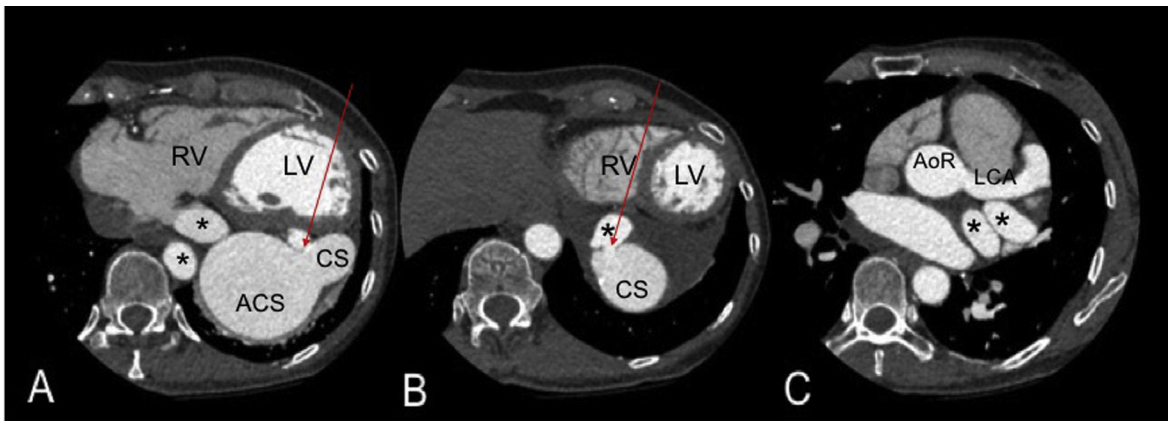

Figure 4 Prospective electrocardiographically triggered computed tomographic coronary angiography. **(A)** *Arrow* shows smaller proximal connection draining into the coronary sinus (CS), *asterisk* indicates the tortuous course of the LCx. **(B)** *Arrow* shows larger distal connection draining into the CS. **(C)** Dilated left coronary artery (LCA) arising from the aortic root (AoR). Tortuosity of the LCx is also demonstrated (*asterisk*). *ACS*, Aneurysmal coronary sinus; *LV*, left ventricle; *RV*, right ventricle.

unroofed coronary sinus, postoperative obstruction, and anomalous drainage of pulmonary or hepatic veins must also be excluded.[12,13]

The majority of CAF connections are incidental findings during coronary angiography for evaluation of revascularization.[6,7,10] Patients generally remain asymptomatic, although exertional dyspnea, palpitations, fatigue, and chest pain have sometimes been reported.[6,10] Onset and severity of symptoms are dependent on patient age, location and size of connections, severity of left-to-right shunting, development of cardiac ischemia due to coronary steal phenomenon, and resistance of the recipient vessel or chamber.[2,7-9] Symptoms and complications from a significant shunt are used as indications for treatment with the fistula ligated or closed by direct suture or percutaneous closure.[9] Treatment aims to obliterate the fistula while preserving normal coronary blood flow either by surgical or percutaneous transcatheter closure with conservative treatment balanced with the risk of the procedure.[14]

Traditionally, coronary angiography is used to diagnose CAF. However, the relation of fistulas to other cardiovascular structures, origins, and courses may not always be apparent,[2] and therefore cardiac computed tomography is increasingly being used to better illustrate fistulas in three dimensions.[7] Xie *et al.*[2] assessed 63 patients using echocardiography before coronary arteriography and/or surgery and showed an accuracy rate of 95.2% by TTE. TTE has shown that the majority of CAFs are single fistulas (92%) and can include proximal dilatation of the involved coronary artery, tortuosity of the coronary vessel, and high-velocity flow signals in the ectatic coronary artery.[2,7] However, fistulas to the pulmonary artery may be more difficult to detect by two-dimensional echocardiography, because the

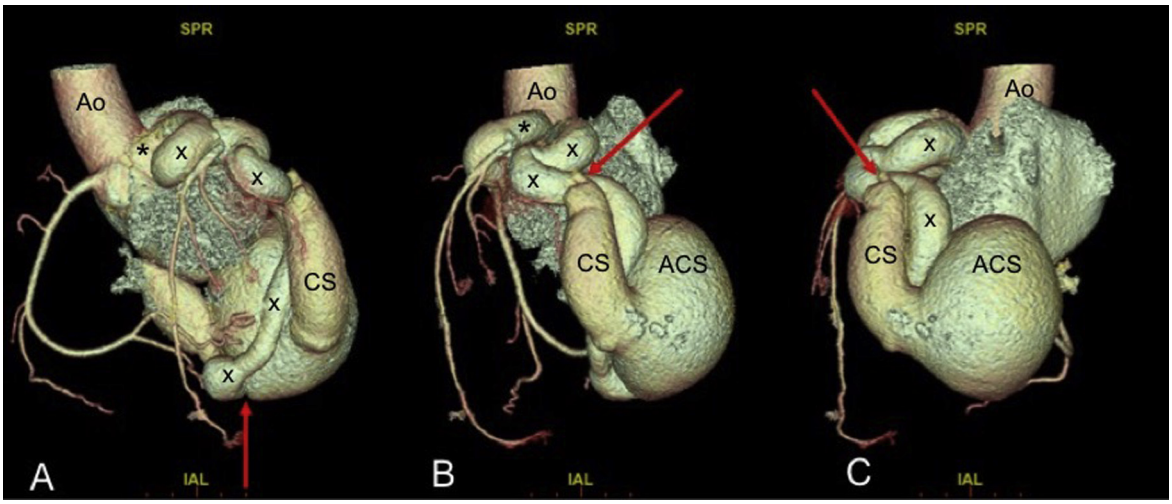

Figure 5 Three-dimensional cardiac computed tomography shows a dilated left coronary artery (*asterisk*) arising from the aortic root with a tortuous course of the LCx (x). **(A)** *Arrow* shows larger distal connection draining into the coronary sinus (CS). **(B,C)** *Arrow* shows smaller proximal connection draining into the CS. *ACS*, Aneurysmal coronary sinus; *Ao*, aorta.

coronary artery may be of normal size and the shunt small.[11] Overall, TTE is a reliable method of diagnosis of coronary artery fistula, including its origin, course, termination, associated defects, and hemodynamic changes.

CONCLUSIONS

This case illustrates a rare, incidental finding of an arteriovenous fistula, via two separate connections between the LCx and coronary sinus, that resulted in a grossly dilated coronary sinus with focal aneurysm. Additionally, this case exemplifies the role of comprehensive two- and three-dimensional TTE assessment in initially finding the underlying etiology well above and beyond the diagnosis of cardiac chamber enlargement and demonstrates the value of and complementary relationship of multimodality imaging in such cardiac anomalies.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found online at https://doi.org/10.1016/j.case.2017.12.004.

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