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CASE REPORT

Left Circumflex Coronary Artery–to–Coronary Sinus Fistula with Coronary Sinus Ostial Atresia and a Persistent Left Superior Vena Cava in an Adult Patient

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Understanding of coronary sinus (CS) anatomy and abnormalities is of critical importance due to their use in interventional procedures. Herein, the authors report a rare case of an asymptomatic 72-year-old man with a left circumflex coronary artery–to-CS fistula, together with CS ostial atresia and persistent left superior vena cava. These findings are described using both cardiac CT angiography and MRI with four-dimensional flow for anatomic and functional assessment.

Supplemental material is available for this article.

While of no hemodynamic significance (1), knowledge of anatomic variants of the cardiac venous system, and namely the coronary sinus (CS), is of great clinical importance given cardiac interventions that cannulate the CS for access, such as left ventricular pacing and arrhythmia mapping or ablation. In 1966, Mantini et al (2) established a classification schema for CS abnormalities that included the following: *(a)* enlargement of the CS (with and without left-to-right shunts), *(b)* absence of the CS, *(c)* atresia of the right atrial CS ostium, and *(d)* hypoplasia of the CS. While each of these cardiac anomalies is rare (1,2) and has been reported individually (1,3), a grouping of any number of these entities has not yet, to our knowledge, been described in the literature. Herein, we report a rare case of a left circumflex coronary artery (LCX)–to-CS fistula, together with CS ostial atresia (CSOA), CS enlargement, and a persistent left superior vena cava (PLSVC).

Case Report

A 72-year-old man with a past medical history of congenital LCX fistula to the CS complicated by atrial fibrillation visited our institution's outpatient cardiology clinic after being lost to follow-up for about 8 years. During previous visits, he had been evaluated for possible surgical treatment of his fistula, but the cardiologists decided to proceed with yearly imaging to monitor for interval changes. He presented with New York Heart Association functional capacity class I and denied any chest pain, shortness of breath, or palpitations for the past 10 years. He underwent 12-lead electrocardiography, which showed a normal sinus rhythm at 70 beats per minute with a first-degree atrioventricular and left posterior fascicular block. The transthoracic echocardiogram revealed a small but unobstructed left atrium compressed by an aneurysmal and partially thrombosed CS, measuring 9.0 × 10.4 cm, and duplicated superior vena cava (SVC). Atresia of the CS ostium was suspected given the large aneurysm. Thus, further characterization of the cardiac anatomy and function was planned with cardiac CT angiography (CTA) and cardiac four-dimensional (4D) flow MRI.

Cardiac-gated CTA of the chest (Movie 1 for axial, sagittal, and coronal cuts, and Movie 2 for three-dimensional [3D] reconstruction) showed an aneurysmal and tortuous LCX with drainage into a partially calcified and partially thrombosed aneurysm measuring 13.7 × 10.0 × 11.6 cm adjacent to the region of the CS (Fig 1A–1C). The aneurysm communicated superiorly with a PLSVC, which drained via a bridging vein into the right SVC, resulting in a left-to-right shunt (Fig 2). Atresia of the central portion of the CS was suspected.

This was also investigated with cardiac 4D flow MRI. CSOA was verified given that no flow between the CS and right atrium was identified (Fig 3, Movie 3). Furthermore, duplicated SVCs were verified with the aneurysmal structure draining retrograde (cephalad) into a left SVC that drained via a bridging vein into the right SVC (Fig 4, Movies 4 and 5). Because the patient had been asymptomatic for 8 years and the thrombus was partially calcified, the decision was made not to pursue surgical management and to continue with serial imaging.

Discussion

We report here a rare case of LCX-to-CS fistula with CSOA leading to a CS aneurysm with retrograde flow through a PLSVC. Both coronary artery–to-CS fistulas and CSOA with PLSVC are individually rare cardiac anomalies (4,5). To our knowledge, only one such case of CSOA together with coronary artery fistula to the CS

Summary

This case report presents a rare grouping of cardiac anomalies including a left circumflex coronary artery–to–coronary sinus (CS) fistula, together with a CS ostial atresia with a resultant aneurysm, and illustrates the advantage of complementary noninvasive modalities such as cardiac CT angiography and MRI to assist in diagnosing rare congenital cardiac defects.

Key Points

- Coronary sinus (CS) abnormalities are rare entities of critical importance due to their use in interventional procedures.
- Complementary noninvasive modalities such as CT angiography and four-dimensional flow MRI allow for anatomic and functional assessment of cardiac anomalies for conclusive diagnosis of CS abnormalities.

Keywords

Cardiac, Coronary Sinus, Aneurysms, Fistula, CT Angiography, MR Imaging

has been described, although this was without aneurysm or a PLSVC (6). To date, the case presented here is, to our knowledge, the first known multimodality description of a coronary artery–to-CS fistula, CSOA, and PLSVC co-occurring.

Coronary artery fistulas (CAFs) are rare anomalies, with a prevalence of about 0.9% in patients who undergo CTA (7); however, they have important clinical implications. The right coronary artery is the most common origin site for a CAF, occurring approximately 50% of the time, while the LCX, demonstrated in the present report, is the origin in only approximately 5% of cases (4). Conversely, the site of drainage for a CAF is typically either the right side of the heart (approximately 60%) or pulmonary arteries (approximately 27%) but is only rarely the CS (approximately 3%) (4). Although these are rare origin and drainage points for a CAF, LCX-to-CS fistulas have been described in the literature previously (8–11). Independent of origin and drainage points, CAF commonly become dilated and aneurysmal due to the creation of a left-to-right shunt, which may lead to thrombosis, embolism (8,9,11), or rupture (12). In cases where the CAF drains into the CS, the aneurysm usually occurs proximal to the CS (8,9,13). In the present case, not only do we demonstrate a rare LCX-to-CS fistula, but also a large, thrombosed aneurysm distal, as opposed to proximal, to the CS. The unexpected location of the aneurysm is likely a consequence of the jointly occurring CSOA and presence of a PLSVC.

Cases of CSOA are rare (6), usually noted as an incidental finding at autopsy (14). With atresia of the CS ostium, cardiac venous drainage cannot return to the right side of the heart; consequently, vessel anomalies must be present to allow for venous drainage. Half of the time, CSOA is associated with a PLSVC (6,15), which allows coronary venous blood to flow in a retrograde (cephalad) direction up the PLSVC into the left brachiocephalic vein and then into the right SVC and right atrium. The identification of a PLSVC in patients with CSOA is of clinical significance, as ligation could interrupt cardiac venous drainage, causing congestion and/or ischemia (15,16). Hence, it is of critical importance to appropriately diagnosis CSOA in the setting of PLSVC prior to manipulation.

Both cardiac CT and cardiac MRI are valuable imaging modalities for the evaluation of congenital cardiac defects, including CS abnormalities. While CT offers superior spatial resolution, full 3D manipulation with high anatomic fidelity, and is more widely accessible, MRI allows for physiologic assessment, such as shunt physiology and ventricular function. Thus, CT and MRI are complementary imaging modalities that can aid in both diagnosis and prognostication of rare congenital heart defects (3). As opposed to cardiac catheterization, the added capability of 4D flow MRI enables noninvasive, retrospective assessment of the magnitude and direction of blood flow (eg, retrograde flow through the PLSVC) or lack thereof (eg, CS to right atrium in CSOA) (17,18), as was performed in the present case.

In conclusion, we present an exceedingly rare grouping of cardiac anomalies that includes an LCX-to-CS fistula, a CSOA with a resultant CS aneurysm, and a PLSVC with retrograde flow. This case highlights the advantages of complementary noninvasive modalities such as CTA and 4D flow MRI in the

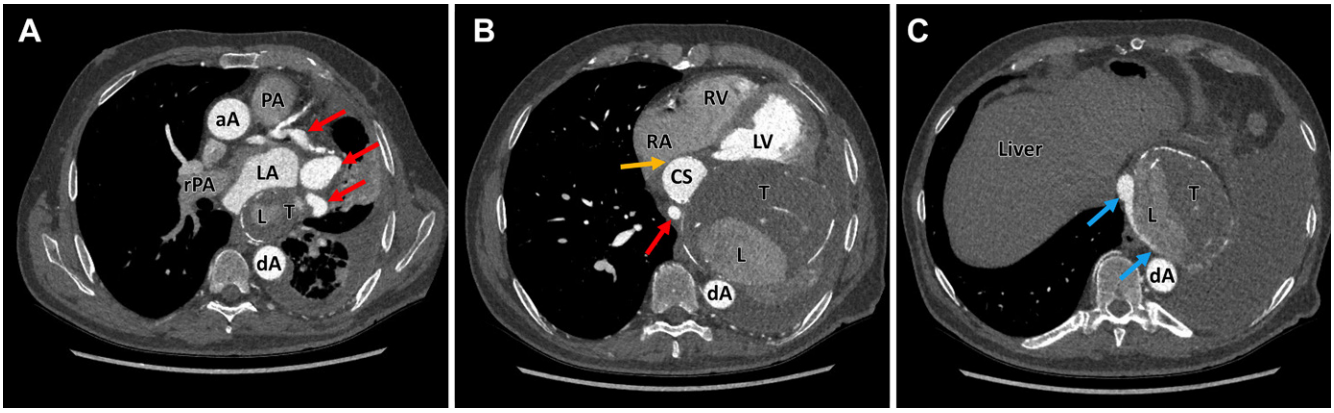

Figure 1: Axial cardiac CT angiographic images demonstrate the presence of a dilated left circumflex coronary artery with fistula to a dilated coronary sinus (CS) with ostial atresia and aneurysm. **(A)** Dilated and tortuous left circumflex coronary artery (red arrows). **(B)** Dilated, opacified CS adjacent to the right atrium (RA) without communication, demonstrating an ostial atresia (yellow arrow). A large, partially thrombosed aneurysm is visualized with a patent lumen. Red arrow indicates left circumflex coronary artery. **(C)** Drainage of the CS (blue arrows) into lumen of a large, partially calcified, partially thrombosed aneurysm. See Movie 1 for axial, sagittal, and coronal cuts, and see Movie 2 for three-dimensional reconstruction. aA = ascending aorta, dA = descending aorta, L = lumen of aneurysm, LA = left atrium, LV = left ventricle, PA = pulmonary artery, rPA = right PA, RV = right ventricle, T = thrombus of aneurysm.

anatomic and functional assessment of cardiac anomalies for conclusive diagnosis.

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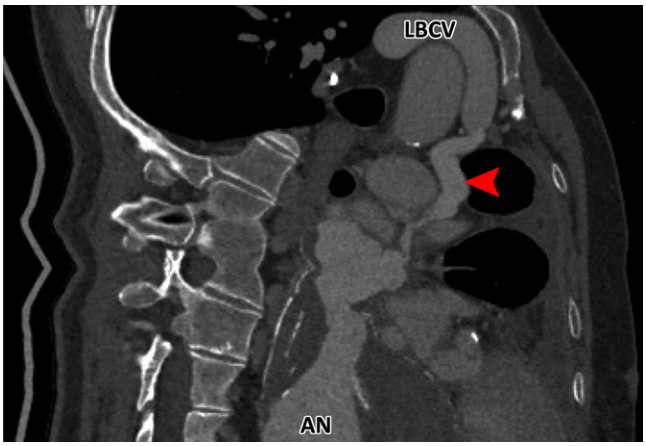

Figure 2: Curved multiplanar reconstruction of cardiac CT angiographic image demonstrates a partially thrombosed aneurysm (AN) draining superiorly through a persistent left superior vena cava (red arrowhead), connecting to a left brachiocephalic vein (LBCV).

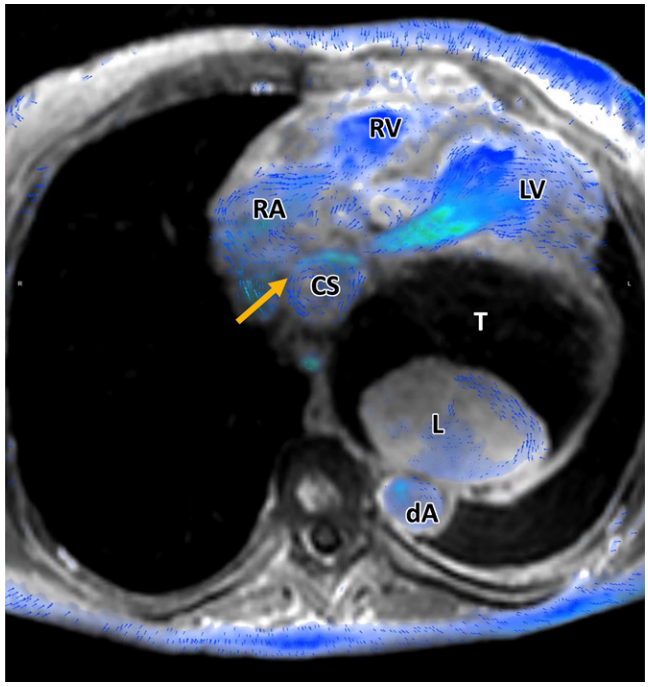

Figure 3: Image from axial cardiac four-dimensional flow MRI demonstrates no flow (ie, ostial atresia; yellow arrow) between the coronary sinus (CS) and the right atrium (RA). The flow through cardiac structures can also be appreciated with minimal flow within the lumen of the aneurysm (L), circular or turbulent flow within the CS, and linear high flow within the left ventricle (LV) toward the LV outflow tract. See Movie 3 for full loop. dA = descending aorta, RV = right ventricle, T = thrombus of aneurysm.

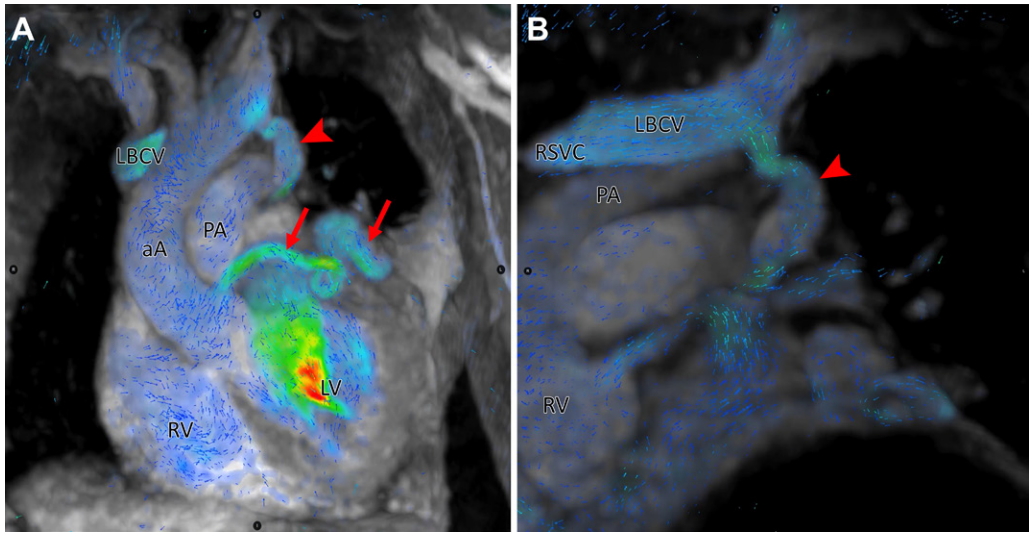

Figure 4: **(A, B)** Oblique reconstructions of cardiac four-dimensional flow MRI demonstrate retrograde (cephalad) blood flow through the persistent left superior vena cava (red arrowhead) into the left brachiocephalic vein (LBCV). Red arrows indicate left circumflex artery. See Movie 4 (for **A**) and Movie 5 (for **B**) for full loop. aA = ascending aorta, LV = left ventricle, PA = pulmonary artery, RSVC = right superior vena cava, RV = right ventricle.

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