Tangled: Unraveling a Rare Combination of Multiple Coronary Artery Fistulas and Severe Aortic Stenosis in a 70-Year-Old Man

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INTRODUCTION

Coronary artery fistulas (CAFs) are rare congenital or acquired anomalies that connect one or more coronary arteries to a cardiac chamber or a great vessel without an intervening capillary bed.[1,2] Most are asymptomatic and are usually incidental findings on echocardiography or angiography for an unrelated cause.[3] Occasionally, complications occur, which include endocarditis, thrombosis, steal syndrome, and heart failure.[3–6] Here, we present a rare case of multiple CAFs draining to the main pulmonary artery in a patient with severe aortic stenosis (AS) presenting a unique management dilemma.

CASE PRESENTATION

Five weeks before admission, a 70-year-old man with good baseline functional capacity, independent on all activities of daily living, developed exertional dyspnea, orthopnea, and paroxysmal nocturnal dyspnea. His symptoms persisted for a week prompting consult at another institution where he was managed as a case of acute heart failure and given diuretics, which resolved his symptoms. His two-dimensional (2D) echocardiogram showed eccentric left ventricular hypertrophy with adequate segmental and global wall motion and contractility, a left ventricular ejection fraction of 62% by the Simpson method, normal right ventricular diameter with adequate wall motion and contractility, dilated left atrial dimensions with a left atrial volume index of 50.98 mL/m^2, moderate mitral regurgitation, normal right atrial diameter, thickened mitral valve leaflets without restriction of motion, thickened and calcified aortic valve, severe AS (aortic valve area of 0.37 cm^2, mean gradient of 78.77 mm Hg, and peak gradient of 124.88 mm Hg), severe aortic regurgitation, mild tricuspid and pulmonic regurgitation, and normal pulmonary artery pressure (13 mm Hg by tricuspid regurgitant jet).

Coronary angiography revealed a mild coronary artery disease involving the mid–right coronary artery, a type III left anterior descending (LAD) artery with a dilated proximal segment, and an incidental finding of multiple CAFs as seen in Figure 1. The largest CAF originates from the midsegment of the LAD and a smaller fistula originating from the left circumflex artery.

He was advised surgical intervention but opted to be discharged. He was prescribed with rosuvastatin 20 mg OD and furosemide 40 mg every other day as home medications. In the interim, he still had exertional dyspnea but could ambulate on flat surface with minimal assistance.

His medical history was significant for hypertension and dyslipidemia with no history of chest trauma or cardiac surgery, but he had undergone right knee and left humerus implants after a bike injury. Family history revealed hypertension and stroke. There was no history of premature coronary artery disease, diabetes, or cancer. He is a previous 30-pack-year smoker who quit 14 years prior and an occasional alcoholic beverage drinker.

Upon readmission, he was received conscious, coherent, and not in distress with the following vital signs: blood pressure, 130/60 mm Hg; heart rate, 77 beats/min; respiratory rate, 20 cycles/min; and temperature, 36.8°C. Neck veins were nondistended; jugular venous pressure was 6 cm. Chest findings showed no gross deformities; symmetric chest expansion; regular heart rhythm with a grade 4/6 systolic murmur best heard at the right upper parasternal border, which radiated to both carotid arteries; and a grade 3/6 low rumbling systolic murmur at the fifth to sixth intercostal space left anterior axillary line. There was no edema, and pulses were full and equal at both upper and lower extremities. He was then admitted for further management.

CASE TIMELINE

Shown in Figure 2 is the summary of the clinical and laboratory findings, diagnosis, and administered therapy of our patient from the onset of symptoms to initial presentation at our institution.

Diagnostics Assessment

A 12-L electrocardiogram showed sinus rhythm, left atrial abnormality, and complete right bundle-branch block (Figure 3).

A repeat resting 2D echocardiography was performed, which showed severe AS, as shown in Figure 4 (aortic valve area of 0.53 cm^2, peak velocity of 5.82 m/s, and mean gradient of 135.69 mm Hg); deterioration of left ventricular function shown as new segmental wall motion abnormalities with hypokinesia of the inferior, inferolateral, and anterolateral walls from base to apex; and a drop in left ventricular ejection from 62% to 52%. Pulmonary artery systolic pressure also increased from 13 to 28 mm Hg by tricuspid regurgitation jet.

A plain computed tomography (CT) scan of the chest and mediastinum was requested to determine if there were other structural abnormalities, which showed a calcified aortic valve, and a fistulous tract draining into the main pulmonary artery as seen in Figure 5.

The heart team was called to discuss options with the family. The patient consented to the intervention and underwent surgical aortic valve replacement (SAVR) with a bioprosthetic valve and ligation of the mid-LAD CAF.

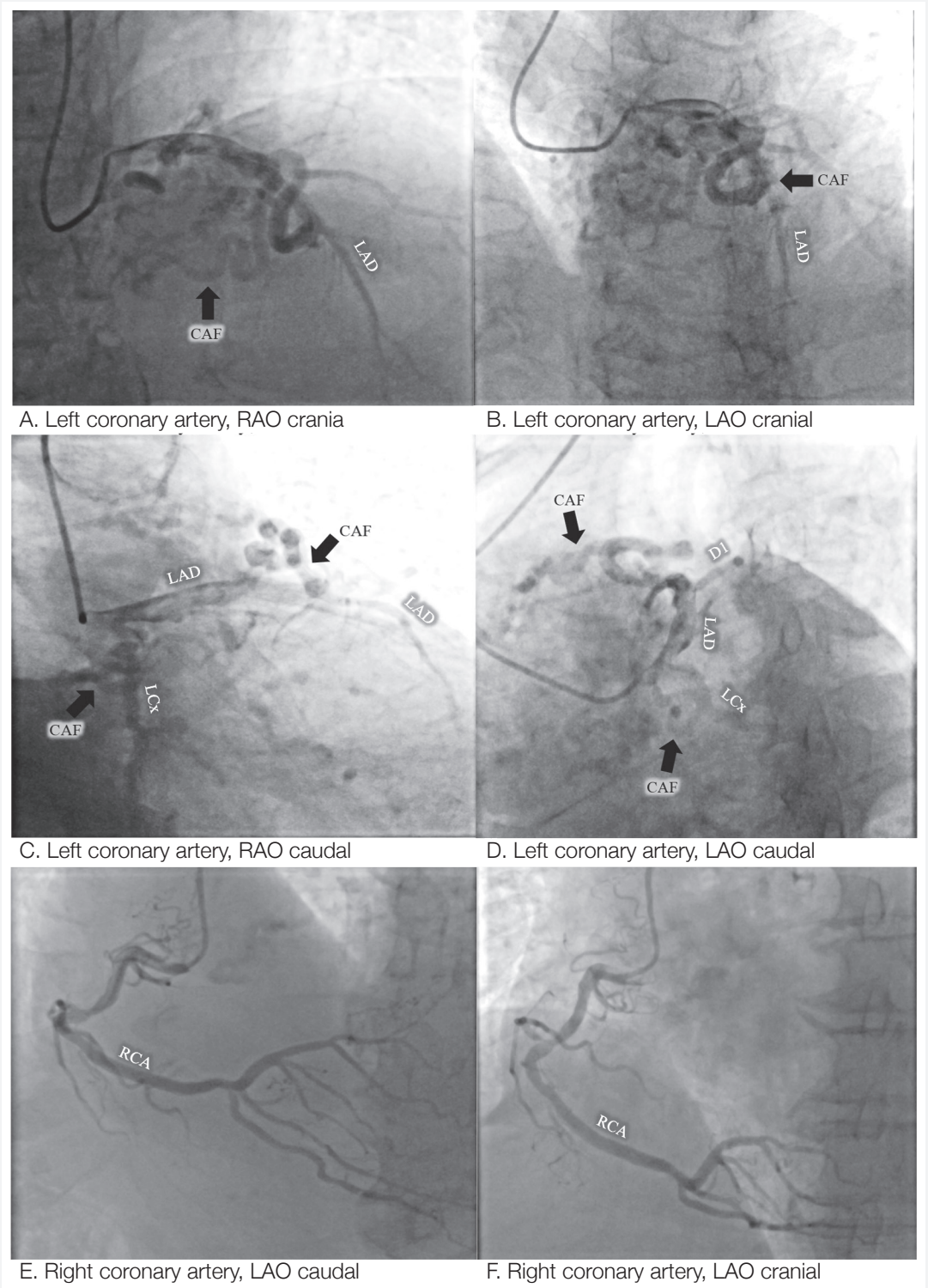
The postoperative course was uneventful, cardiac rehabilitation was started in-hospital, and he was eventually discharged well on rosuvastatin, carvedilol, warfarin, and lactulose. Three months after surgery, the patient had no heart failure symptoms already, and a repeat 2D echocardiogram showed improvement in left ventricular wall motion and contractility with associated increase in ejection fraction from 52% to 69.7%.

DISCUSSION

Aortic stenosis with a mean gradient >40 mm Hg, peak velocity >4 m/s, and an aortic valve area <1.0 such as in our case is classified as severe AS. Heart failure is one of the three cardinal manifestations of severe AS, which include angina and syncope.[7] These symptoms develop typically after 70 years old for those with calcific AS and may present abruptly in some cases.[7–9] The mechanism may be left ventricular diastolic dysfunction with an excessive rise in end-diastolic pressure that is amplified by the limited ability to increase cardiac output leading to myocardial ischemia, ventricular dysfunction, and pulmonary congestion.[7,10] These pathologic mechanisms are further complicated by the presence of a CAF.

Coronary artery fistulas are rare congenital anomalies defined as abnormal connections between one or more coronary arteries to a cardiac chamber or a great vessel without an intervening capillary bed.[1,2] The prevalence varies from 0.05% to 0.25% in studies that used coronary angiography, but in studies that used CT angiography, the prevalence seems to be higher at 0.9% in the general population.[7–10] Most are asymptomatic and are usually incidental findings on echocardiography or angiography.[3] Complications due to CAFs are rare, which include endocarditis, thrombosis, steal syndrome, and heart failure.[3–6] Likewise, the combination of CAFs and severe AS is extremely rare and hence presents unique dilemmas in the approach and management.

The pathophysiologic mechanism of complications due to CAFs is due to decreased myocardial perfusion by myocardial stealing or reduction in myocardial blood flow distal to the site of the CAF connection.[11] This promotes left ventricular dysfunction and, as in the case of our patient, potentially amplifies the pathologic effects of severe AS.[11] The severity of stealing is dependent on the size of the communication and the resistance of the recipient chamber or vessel.[2,11] As shown in the video clip below, the caliber of the CAF is twice that of the coronary artery distal to it and drains into the pulmonary artery, which inherently has a lower resistance. This promotes preferential blood flow to the CAF and places the distal segments of the myocardium at



A. Left coronary artery, RAO crania
B. Left coronary artery, LAO cranial
C. Left coronary artery, RAO caudal
D. Left coronary artery, LAO caudal
E. Right coronary artery, LAO caudal
F. Right coronary artery, LAO cranial

Figure 1. Coronary angiogram views of a 70-year-old man presenting with dyspnea showing mild coronary artery disease of the midsegment of the right coronary artery and multiple coronary fistulas originating from the mid–left anterior descending artery and proximal left circumflex artery.
D1=first diagonal branch; LAD=left anterior descending; LAO=left anterior oblique; LCx=left circumflex; RAO=right anterior oblique; RCA=right coronary artery.

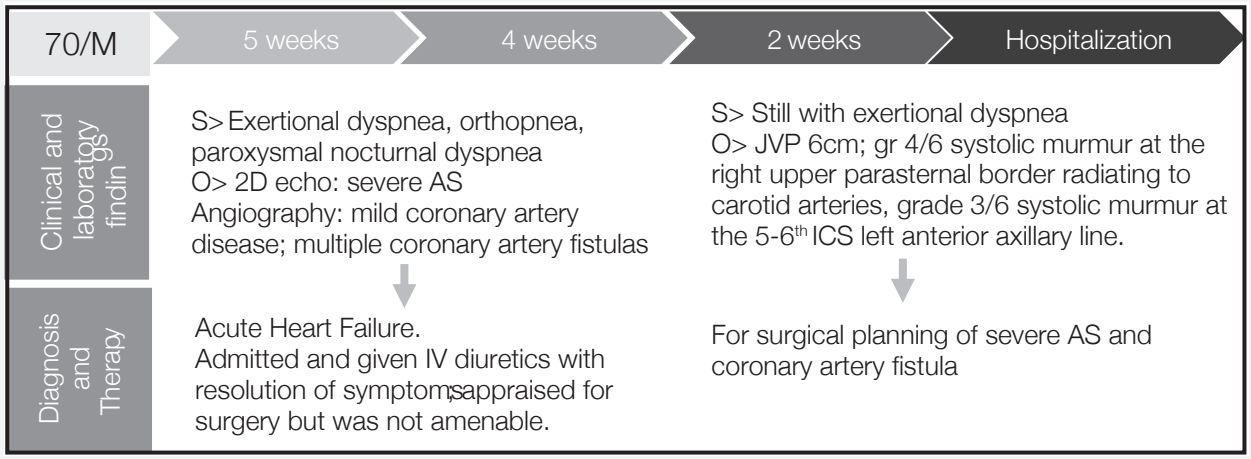


Figure 2. Case report timeline.
2D=two-dimensional; AS=aortic stenosis; ICS=intercostal space; IV=intravenous; JVP=jugular venous pressure.

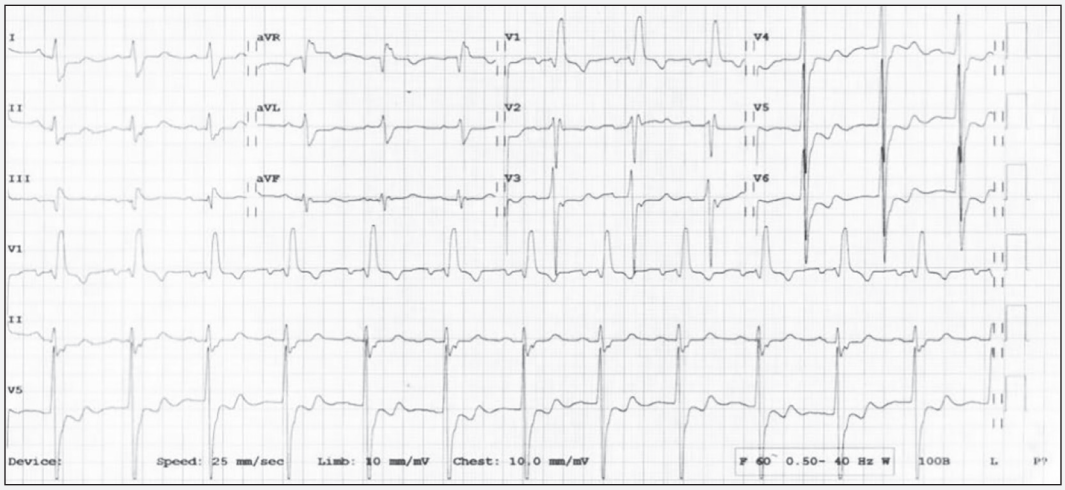


Figure 3. Twelve-lead electrocardiogram of the patient.

risk for ischemia. For our patient, the type III LAD supplies blood not only to the anterolateral walls via the diagonal branches but also to inferior walls of the left ventricle. Hence, a steal phenomenon involving the LAD would affect these walls as seen in our patient.

The imaging modality of choice to diagnose CAF is cardiac catheterization, which can define CAF patterns as it allows precise assessment of fistula anatomy including fine vessels and provides high temporal and hemodynamic information.[12,13] A cardiovascular CT CCT scan is another imaging modality that would be most beneficial to supplement coronary angiography. In the 2020 European Society of Cardiology guidelines for the management of adult congenital heart diseases, cardiovascular CT imaging is recommended to evaluate high-risk anatomy, including features such as an intramural course and orifice anomalies such as a slit-like orifice, acute angle take-off, or an orifice >1 cm above the sinotubular junction.[14] However, the performance of both tests may not be ideal as in our case. Our patient for planned cardiac surgery had several independent risk factors for postoperative acute renal failure such as male sex, age, and hypertension.[15–19] Furthermore, studies indicate that the incidence of postoperative acute renal failure is high among patients who recently underwent coronary angiography and may occur in as much as 39% among patients who underwent coronary angiography 7 to 30 days before cardiac surgery.[15,19] Thus, in our patient, who underwent angiography less than 3 weeks before cardiac surgery, CT angiography was foregone to avoid postoperative acute renal failure. Instead, a plain CT scan was performed to grossly assess the chest structures.

The choice to perform SAVR was straightforward in this case. Aortic valve replacement is indicated with a class I recommendation in symptomatic patients with severe, high-gradient AS such as in our case.[20] Surgical aortic valve replacement is also recommended in patients younger than 75 years who are at low risk for surgery (STS-PROM/EuroSCORE

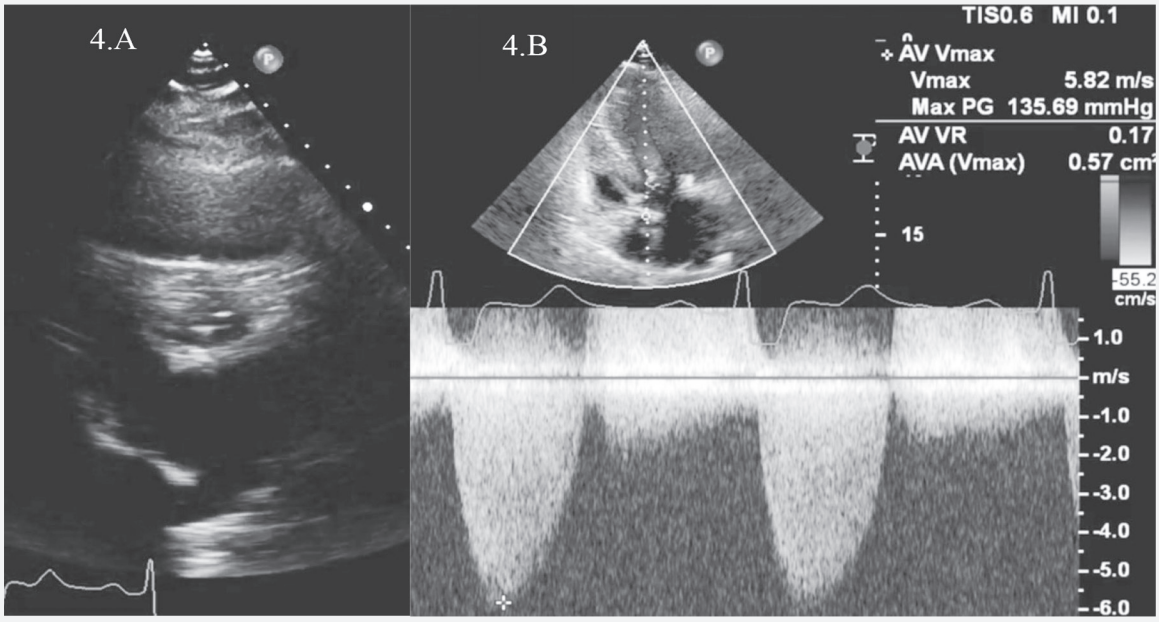


Figure 4. 4.A, 2DED of the patient's aortic valve, PSAX view, during systole showing restricted opening. 4.B, CW Doppler of AV showing peak velocity of 5.82 m/s, and peak gradient of 135.69 mm Hg.
AV=aortic valve; 2DED=2-dimensional echo/Doppler; CW Doppler=continuous wave Doppler; PSAX=parasternal short-axis.

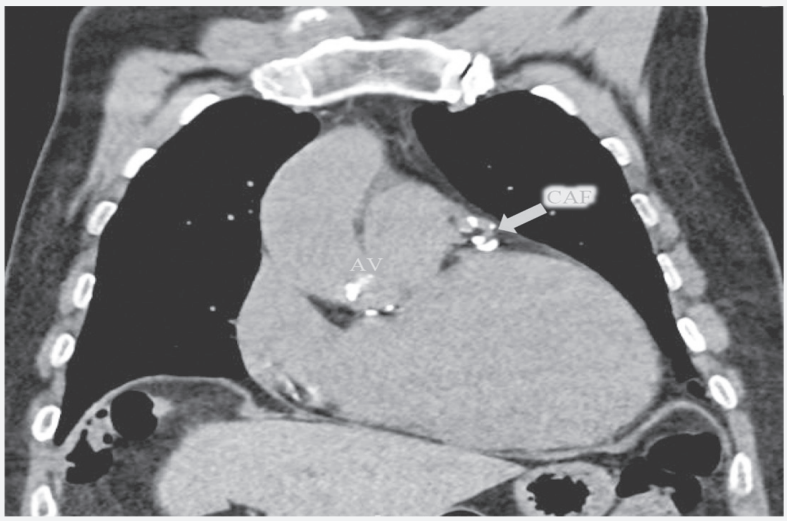


Figure 5. Plain chest computed tomography scans of the patient showing a calcified AV, and a fistulous tract (CAF) draining into the pulmonary artery.
AV=aortic valve; CAF=coronary artery fistula.

[Society of Thoracic Surgeons Predicted Risk of Mortality/ European System for Cardiac Operative Risk Evaluation] II <4%) or in patients who are operable and unsuitable for transaortic valve implantation.[20] In this case, because our patient had severely calcified aortic valves, which may occlude the coronary ostia, transaortic valve implantation was an unsuitable option. Hence, SAVR was opted.[20,21] On the other hand, the indication to ligate the patient's CAF needed further discussion due to lack of strong evidences regarding intervention. However, the 2020 European Society of Cardiology guidelines on the management of adult congenital heart disease mention that surgical or percutaneous closure may be indicated in the presence of symptoms, complications, and significant shunt such as in our patient.[14,20] Consequently, during the operation, coronary vessels were identified with the largest fistula originating from the mid-LAD draining to the main pulmonary artery, which was ligated.

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