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CONGENITAL HEART DISEASE

CLINICAL CASE

A Rare Case of Single Coronary Artery With Pulmonary Artery Fistula

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ABSTRACT

BACKGROUND Single coronary artery is a rare congenital anomaly. Its coexistence with coronary artery fistula is exceedingly uncommon.

CASE SUMMARY A 61-year-old woman with no cardiovascular risk factors underwent her first cardiological evaluation after incidental detection of atrial fibrillation. Imaging revealed a markedly dilated single coronary artery originating from the left coronary sinus, with a large fistula to the pulmonary artery trunk. Despite atrial enlargement and moderate mitral and tricuspid regurgitation, the patient was asymptomatic with preserved ventricular function. A conservative management strategy with close follow-up was adopted after multidisciplinary discussion.

DISCUSSION Management of rare coronary anomalies should be individualized. While surgery is recommended in symptomatic patients or those with evidence of ischemia, asymptomatic individuals may be managed conservatively; however, signs of adverse cardiac remodeling should prompt early reconsideration of intervention.

TAKE-HOME MESSAGE Conservative follow-up may be appropriate in asymptomatic patients with complex coronary anomalies, but progressive structural changes warrant timely reassessment of surgical options. (JACC Case Rep. 2025;30:104875) © 2025 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 61-year-old woman with no cardiovascular risk factors presented to our clinic for her first cardiological evaluation. She had no history of chest pain, dyspnea, or palpitations. However, some months earlier, paroxysmal atrial fibrillation was incidentally detected during an ergometric test performed before starting Pilates training. She underwent successful electrical cardioversion and was subsequently treated with bisoprolol and rivaroxaban. At presentation, she was asymptomatic and hemodynamically stable. Physical examination revealed rhythmic heart sounds with a systodiastolic murmur at the left sternal margin and a soft systolic murmur at the apex, suggestive of mitral regurgitation.

PAST MEDICAL HISTORY

The patient had been previously diagnosed with a congenital heart disease in early adulthood following the detection of a heart murmur, though documentation

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ABBREVIATIONS AND ACRONYMS

CAF = coronary artery fistula

CT = computed tomography

SCA = single coronary artery

regarding the exact anomaly was unavailable. At the age of 20 years, she had been advised to undergo surgical intervention, which she declined. She also had a history of left renal agenesis and recurrent right breast cancer, treated with quadrantectomy in 1997 and mastectomy in 2007. She had an uneventful pregnancy and maintained an active lifestyle.

DIFFERENTIAL DIAGNOSIS

Given her history of congenital heart disease and the presence of a heart murmur, possible diagnoses included anomalous coronary artery origin, valvular heart disease, intracardiac shunts, and vascular abnormalities such as coronary artery fistula (CAF).

INVESTIGATIONS

Electrocardiogram revealed sinus rhythm with a mean heart rate of 82 beats/min, occasional premature atrial and ventricular contractions, left atrial enlargement, and normal atrioventricular and intraventricular conduction (**Figure 1**).

Transthoracic echocardiography demonstrated enlargement of both atria, increased left ventricular end-diastolic volume with preserved ejection fraction, and moderate mitral regurgitation (Video 1). The aortic and tricuspid valves showed mild and moderate regurgitation, respectively, with an estimated pulmonary artery pressure of 45 mmHg. No pericardial effusion was detected. The left main coronary artery was found to be markedly dilated and gave rise to a tortuous circumflex artery. The right coronary artery was not visualized. A large fistula between the single coronary artery (SCA) and the pulmonary artery was evident. Cardiac computed tomography (CT) confirmed a dilated and serpiginous SCA (diameter 23 mm) originating from the left coronary sinus, supplying both left and right coronary territories, with a large fistulous connection to the anterolateral wall of the left pulmonary artery trunk (Video 2).

MANAGEMENT

According to the 2020 European Society of Cardiology[1] and the 2018 American Heart Association/American College of Cardiology[2] guidelines for the management of adult congenital heart disease, surgical intervention is recommended in symptomatic patients with coronary anomalies and should be considered in asymptomatic individuals with ventricular dysfunction or evidence of myocardial ischemia. Although the patient was asymptomatic at presentation, the presence of significantly enlarged atria, moderate mitral and tricuspid regurgitation, and arrhythmic events during exertion suggest a progressive trajectory toward biventricular dysfunction. Surgical correction might be advisable before irreversible chamber remodeling occurs. Iacona et al[3] demonstrated favorable outcomes with surgical correction of aneurysmal coronary artery fistulas, supporting a more proactive strategy in selected patients. Nevertheless, in our case, given the absence of ischemic symptoms and the patient's preference, a conservative approach with close monitoring was chosen, although this decision remains under continuous reevaluation.

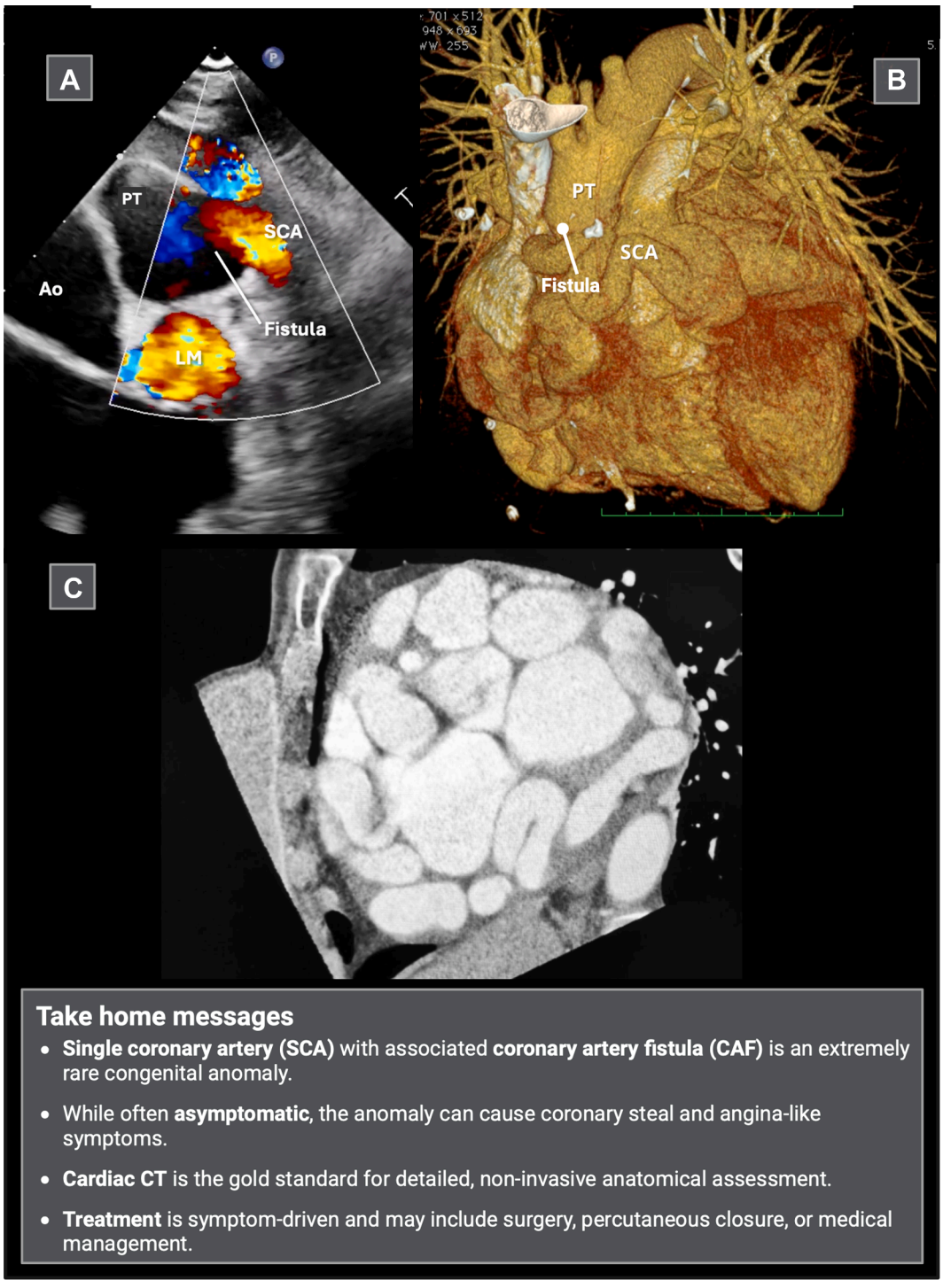
TAKE-HOME MESSAGES

- SCAs represent a rare coronary artery anomaly. They may be isolated or associated with other congenital anomalies. The coexistence of SCA and CAF is exceedingly uncommon, with only a few cases documented.
- Individuals with SCA or CAF are often asymptomatic and incidentally diagnosed. However, clinical manifestations can vary widely. The coexistence of SCA and CAF may lead to preferential blood diversion with coronary steal phenomena and angina-like symptoms.
- Cardiac CT is the gold standard for noninvasively characterizing the coronary anomaly.
- Surgical intervention is recommended in symptomatic patients with coronary anomalies. However, treatment should be tailored to each patient, with percutaneous transcatheter interventions or medical management being considered where appropriate. While conservative management may be considered in asymptomatic patients, progressive atrial dilation and valvular regurgitation should prompt reconsideration of early surgical intervention.

OUTCOME AND FOLLOW-UP

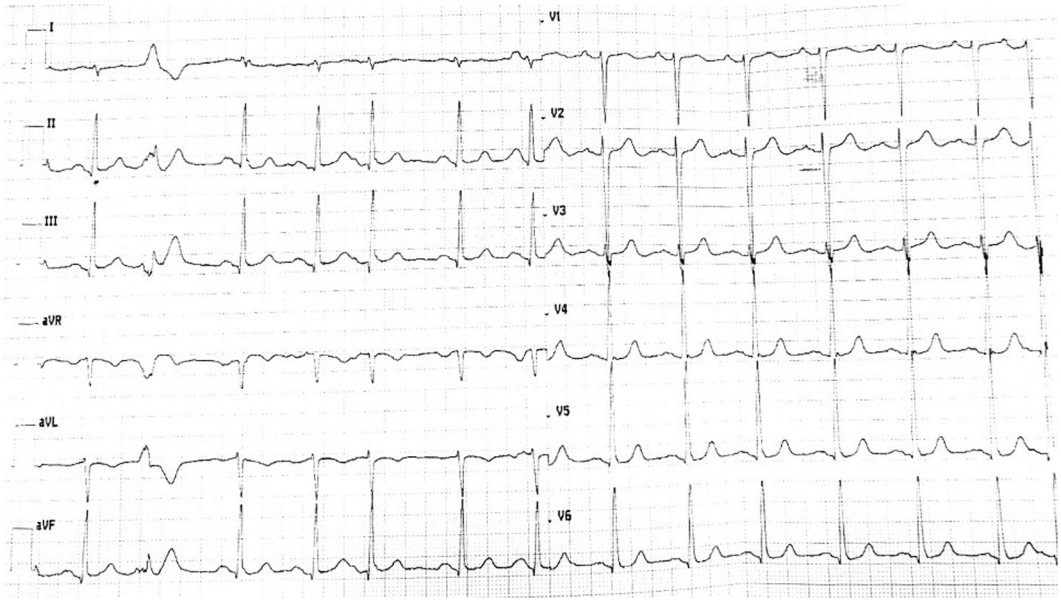
Large coronary artery fistulas can lead to complications such as angina, myocardial infarction, arrhythmias, heart failure, and endocarditis. Therefore, the patient was enrolled in long-term follow-up at our grown-up congenital heart disease clinic.

VISUAL SUMMARY **SCA and Pulmonary Artery Fistula**



(A) Echocardiographic visualization of the fistula between the single coronary artery (SCA) and the pulmonary artery. (B) Three-dimensional computed tomography (CT) reconstruction illustrating the fistulous connection between the SCA and the pulmonary trunk (PT). (C) Cross-sectional CT imaging of the heart, revealing the tortuous and serpiginous course of the anomalous SCA. Ao = aorta; LM = left main coronary artery.

FIGURE 1 Electrocardiogram at the Initial Presentation



Sinus rhythm at a mean heart rate of 82 beats/min with some premature atrial and ventricular contractions, signs of left atrial enlargement, and normal atrioventricular and intraventricular conduction.

Every 3 to 6 months, she will undergo a comprehensive cardiological evaluation including physical examination, electrocardiography, and echocardiography. Any new symptoms, particularly angina, will prompt reconsideration of surgical intervention. Given the presence of a shunt from the coronary to the pulmonary circulation, there is a risk of coronary steal and myocardial ischemia, necessitating close monitoring. Right heart catheterization remains a valuable tool for further assessment of shunt magnitude (Qp/Qs) and pulmonary hemodynamics, and may be considered at follow-up. Current guidelines also recommend nonpharmacological functional imaging in patients with coronary anomalies to confirm or exclude myocardial ischemia.

DISCUSSION

This case highlights an exceptionally rare congenital coronary anomaly, underscoring the need for individualized patient management. SCAs are rare, occurring in 0.024% to 0.066% of the population,[4] while CAF is identified in 0.1% to 0.2% of patients undergoing coronary angiography.[2] The coexistence of SCA and CAF is exceedingly uncommon, with only a few cases documented.[5-7] To our knowledge, this is the first reported case describing a large fistula between a SCA and the pulmonary artery. Although many cases of coronary anomalies remain asymptomatic, potential clinical manifestations include exertional angina, fatigue, palpitations, arrhythmias, heart failure, and sudden cardiac death. The presence of a coronary artery-to-pulmonary artery fistula may result in preferential blood flow diversion, leading to coronary steal phenomena and an increased risk of myocardial ischemia symptoms.

Cardiac CT with multiplanar reconstruction provides a detailed assessment of coronary anatomy, allowing for accurate diagnosis. Compared with traditional coronary angiography, it offers a superior, noninvasive approach for identifying small-caliber fistulas. While surgical repair remains the standard treatment for complex or symptomatic coronary fistulas, management must be individualized based on clinical presentation and anatomical features. In this case, despite anatomical complexity and evidence of chamber dilation and valvular involvement, the absence of ischemic symptoms and preserved ventricular function supported an initial conservative approach, with the understanding that ongoing

monitoring may necessitate reconsideration of surgical intervention.

CONCLUSIONS

Despite its rarity, this case provides several insights into the management of congenital coronary anomalies. The coexistence of SCA and CAF is exceptionally uncommon. While surgical correction is the preferred approach in symptomatic patients, asymptomatic individuals with preserved ventricular function can be managed conservatively with close follow-up. This case underscores the importance of a patient-centered approach, taking into account the anatomical complexity, symptomatology, and individual patient preferences.

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KEY WORDS computed tomography, congenital heart disease, coronary anomalies, single coronary artery

APPENDIX For supplemental videos, please see the online version of this paper.