Single Coronary Ostium Branching into the Right and Left Coronary Artery and Draining into the Pulmonary Artery: A Case Report

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Abstract

This is the case of a 38-year-old female diagnosed with chronic glomerulonephritis and on chronic hemodialysis who was admitted for worsening heart failure. Physical examination revealed a continuous murmur in the left parasternal border, which decreased with inspiration. A chest radiograph revealed an enlarged main pulmonary artery. Transthoracic echocardiogram revealed a dilated right coronary ostium and a continuous Doppler signal in the main pulmonary artery (MPA), indicative of a fistula draining into the MPA. A coronary angiogram and cardiac computed tomography angiogram revealed an absent left main coronary artery and a right coronary artery (RCA) giving rise to the left anterior descending artery (LAD) and left circumflex artery (LCx). An arteriovenous fistula from the LAD draining into the MPA was also noted. She was diagnosed with anomalous origin of the LAD and LCx from the RCA with a coronary arteriovenous fistula from the LAD, LCx and RCA to the MPA, and end-stage renal failure due to glomerulonephritis. Her heart failure and renal failure were managed with optimal medical therapy and continuous hemodialysis, respectively. Surgery was offered to ligate the arteriovenous fistula. This case represents a rare combination of coronary anomalies. Despite the rarity, such defects should be suspected among patients presenting with heart failure and a continuous murmur on examination.

INTRODUCTION

Congenital coronary anomalies are rare pathologies with paucity of data in adults. A coronary arteriovenous fistula is one of these congenital anomalies and is defined as a communication between a coronary artery and a segment of the pulmonary circulation. We present a case of an adult with multiple coronary anomalies and an arteriovenous fistula.

THE CASE

This is a case of a 38-year-old female who was admitted to our institution for a two-month history of progressive difficulty of breathing, orthopnea, bipedal edema and abdominal enlargement. She was diagnosed with chronic glomerulonephritis two years prior and had been on hemodialysis for 10 months. She had hypertension for a year and was maintained on enalapril. She had a 10-pack-year history of cigarette smoking with no family history of heart disease. She had given birth to five children without complications.

On physical examination, her blood pressure was 130/80 mmHg on all extremities and her heart rate was 90 beats per minute. She had a dynamic precordium with right ventricular heave, distinct heart sounds and a continuous murmur on the second intercostal space on the left parasternal border, with the diastolic component louder than the systolic and which decreased with inspiration. She had decreased breath sounds on both lower lung fields and rales on the right lower lung field. She had a globular, soft and nontender abdomen, which was positive for the fluid wave test. She also had grade III pitting, bipedal edema.

Electrocardiography revealed regular sinus rhythm, normal axis, left ventricular hypertrophy with nonspecific changes. Her chest radiograph revealed a dilated pulmonary artery and reticulonodular infiltrates in the right lower lung. A transthoracic echocardiogram revealed multi-chamber dilatation with hypokinesia of the mid-inferior, septal and inferoseptal walls. Ejection fraction was 41% by Simpsons method. The left atrium, right atrium and right ventricles were also dilated. The right ventricle had adequate contractility, with a tricuspid annular plane systolic excursion of 19 mm. All the cardiac valves were thickened but without restriction in opening and closing motion. The main pulmonary artery was dilated with a diameter of 50 mm. The right coronary artery (RCA) appeared dilated (Figure 1A) and exhibited increased flow (Figure 1B). A mosaic color flow was seen in the pulmonary artery in both systole (Figure 1A) and diastole (Figure 1B).

Coronary angiography and hemodynamic studies revealed an absent left main coronary artery, a dilated and tortuous RCA, left coronaries arising from the right coronary sinus, and a coronary artery fistula arising from the origin of the RCA and draining into the pulmonary artery. There was a significant step-up of oxygen saturation and pressure from the right ventricle (60% and 16 mmHg, respectively) to the main pulmonary artery (82% and 28 mmHg, respectively).

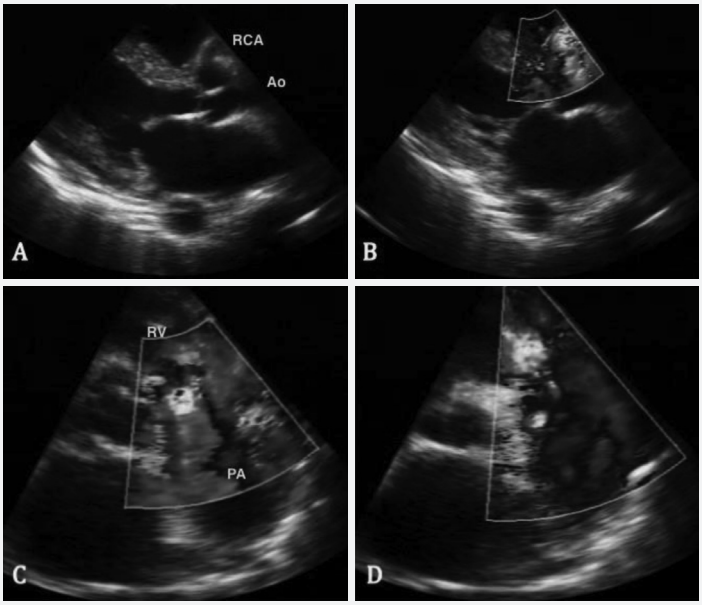

Figure 1. Transthoracic echocardiogram showing (A, B) a dilated right coronary artery and coronary ostium and a mosaic color flow in the main pulmonary artery during systole (C) and diastole (D).

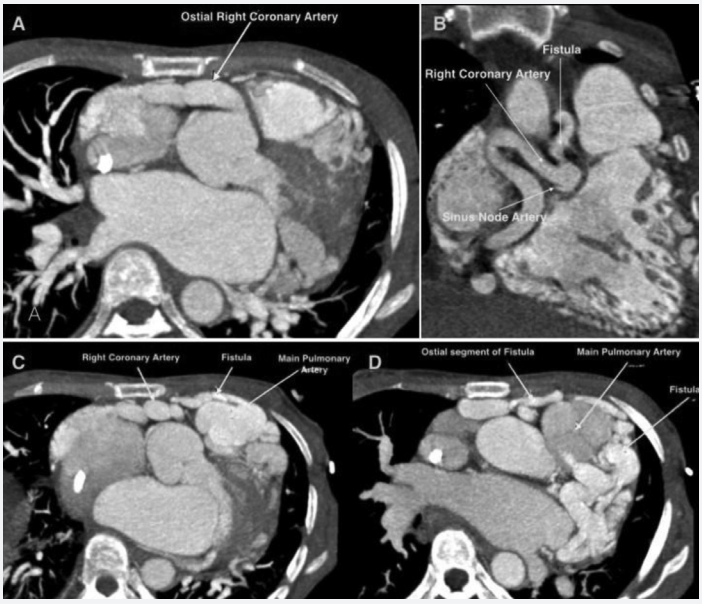

Figure 2. Computed tomography showing a dilated right coronary ostium (A) with three emerging branches (sinus node artery, right coronary artery and fistulous tract) (B), the fistulous tract from the right coronary ostium branching and draining into the pulmonary artery (C) and a confluence of fistulous arteries that have multiple connections to the pulmonary artery (D).

Computed tomography scan showed a missing left main coronary artery (MCA). The RCA was markedly enlarged (Figure 2A) and tortuous but free of disease along its entire length. A dilated sinus node artery emerged in its proximal segment and

a dilated acute marginal branch from its middle segment (Figure 2B). The sinus node artery further branched out posteriorly and drained into the posterior atrioventricular groove. The distal RCA branched out into a posterior descending artery and posterolateral artery in its distal segment, which were also free of disease. The posterolateral artery further extended into the posterior atrioventricular groove, meeting the sinus node artery to form the left circumflex artery (LCx). A fistula arose from the proximal portion of the RCA and coursed into the anterior aspect of the main pulmonary artery (MPA) just beneath the anterior rib and further into the apex of the left ventricle to form the left anterior descending artery (LAD). It also gave off small branches that drained into the MPA (Figure 2C, 2D). All of the branches of the RCA, including the anomalous branch, merged at the lateral aspect of the MPA. The pulmonary trunk was markedly dilated and branched out into the right and left pulmonary arteries (Figure 3).

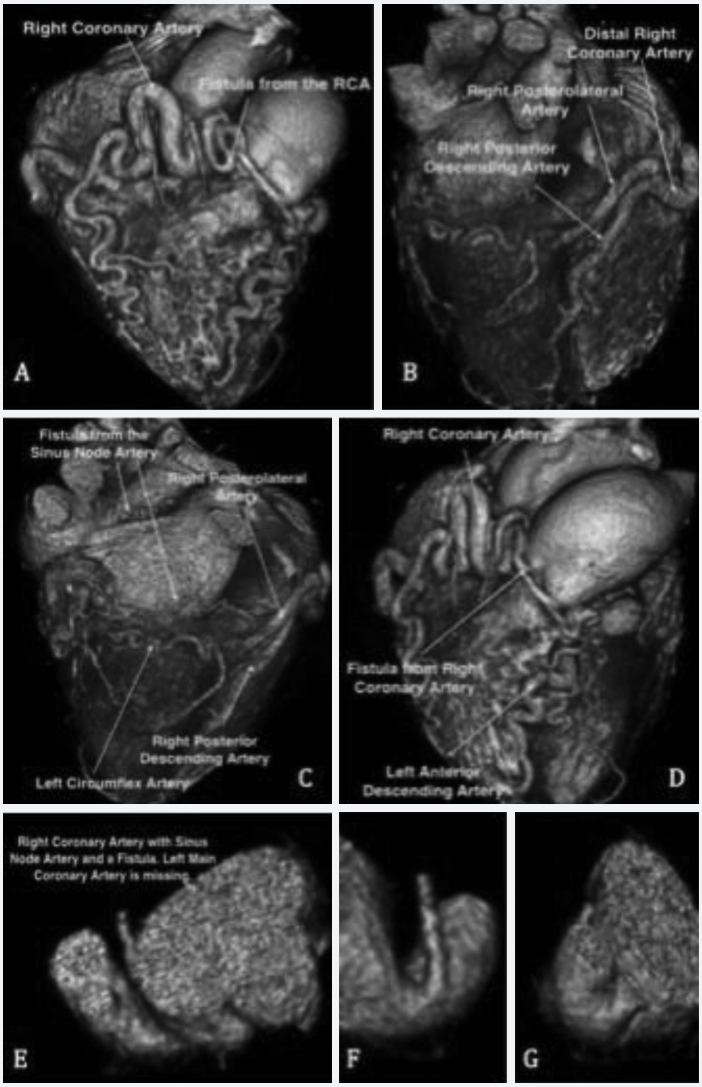

Figure 3. Three-dimensional reconstruction showing a dilated right coronary artery, a fistulous branch traveling anterior to the dilated pulmonary artery (A), a dilated right posterolateral and descending artery (B), fistulous branch from the sinus node artery joined by the right posterolateral artery becoming the left circumflex artery (C), fistulous tract from the right coronary artery becoming the left anterior descending artery (D), right coronary ostium giving rise to three branches (E), one of which is the sinus node artery (F), and the fistula that becomes the left anterior descending artery (G).

Her heart failure was treated with optimal medical therapy while her renal failure was treated with continuous hemodialysis. Surgery was offered to ligate the arteriovenous fistula. However, during her most recent admission, she had massive hematemesis, probably due to esophageal varices or uremic gastropathy, which led to her demise.

DISCUSSION

A single coronary artery arising from the right sinus of Valsalva is an extremely rare congenital anomaly, accounting for only 0.3% of all coronary anomalies and with an incidence of 0.024% to 0.066% in the general population.

The modified Lipton classification proposed by Yamanaka et al is frequently used to describe coronary artery anomalies and ascertain their incidence.[1,2] In the subgroup classification of patients with single coronary artery, lesions are described based on the ostium where they originate, the anatomical course of the arteries, and the relationship between the anomalous coronary artery and the aorta and pulmonary artery (Table 1). Our case has a RIII-C type of coronary artery anomaly, which has an incidence of 0.004%.

Coronary arteriovenous fistulas are usually congenital in origin but may also be acquired and due to coronary atherosclerosis, Takayasu arteritis, polymyositis, cardiac surgeries and procedures, acute myocardial infarction and cardiac trauma. Fistulas draining into the pulmonary artery are rare, which comprise 17% of all coronary arteriovenous fistulas.[3] Multiple fistulas draining into a single chamber is also rarer than fistulas with a single communication (33% vs 67%, respectively), according to Kose et al.[4] Given these rates, the estimated incidence of this case with multiple coronary anomalies is 0.0002% of the general population. The combination of these features has not been reported in literature.

Presentation of such patients with congenital coronary anomalies vary based on the course of the anomalous artery, the site draining the arteries and the severity of the shunt, if any. Arteries with intramural or inter-arterial course or with myocardial bridging are prone to myocardial ischemia, especially in the young.[5] Patients with coronary artery anomalies with fistulous connections to right-sided chambers and the pulmonary artery usually become aneurysmal and are associated with myocardial ischemia by creating a “steal” phenomenon from the coronary artery into the lower-resistance chamber or vessel. The chamber receiving the shunt may also undergo changes due to pressure and volume overload. Pulmonary hypertension and congestive heart failure are complications of large left-to-right shunts.[3]

Table 1. Modified Lipton Classification of Isolated Coronary Artery Anomalies

Origin	Course and Incidence	Relationship between the coronary artery and the aorta and pulmonary artery
Right ostium (R)	Anatomical course of either a right or left coronary artery (I), 0.0008%	Anterior (A) Between (B) Posterior (P) Septal (S) Combined (C)
	Anomaly arises from the proximal part of the normal right or left coronary artery and cross the base of the heart before assuming the normal position of the inherent coronary artery (II), 0.015%	
	Anomaly where the LAD and the LCx arise separately from the proximal part of the normal right coronary artery (III), 0.004%	
Left ostium (L)	Anatomical course of either a right or left coronary artery (I), 0.016%	
	Anomaly arises from the proximal part of the normal right or left coronary artery and cross the base of the heart before assuming the normal position of the inherent coronary artery (II), 0.009%	

It is imperative to accurately establish the particular type of coronary artery anomaly and the mechanism of its interference to normal blood flow for adequate medical and surgical planning.

In our case, the patient presented with multi-chamber dilatation and multi-segmental wall motion abnormality consistent with pressure and volume overload and myocardial ischemia from steal phenomenon. The patient's demise may have been caused by intra-abdominal bleeding from ruptured variceal vessels from her portal hypertension due to right heart failure, although uremic gastropathy is also a consideration.

CONCLUSION

We presented an extremely rare case of single coronary artery with its branches draining into the pulmonary artery. Cardiac imaging such as echocardiography, coronary angiography and computed tomography scanning are definitive in ascertaining the exact lesions in such cases. Despite its rarity, suspicion of such defects should be made among patients presenting with heart failure and a continuous murmur on examination.

DISCLOSURE

None

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