Congenitally Absent Right Coronary Artery with a Coronary Artery Fistula From the Left Anterior Descending Artery to the Main Pulmonary Artery: A Case Report

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Abstract

Introduction: Single coronary artery and coronary artery fistulas are unusual findings either alone or in combination. They may be incidental findings or can present with myocardial ischemia.

Case Presentation: A 46-year-old male had chest pain accompanied by heart failure symptoms. He sought consult in our institution where further workups revealed that he suffered an acute coronary syndrome. He underwent diagnostic coronary angiogram which showed significant coronary artery disease. There was also a single coronary artery with a coronary artery fistula. Patient was appraised regarding surgical revascularization and repair but did not consent. He was stabilized and improved on optimal anti ischemic therapy.

Discussion: Electrocardiogram revealed ST elevation myocardial infarction of the anterior wall. Two dimensional echocardiogram showed depressed systolic function with an ejection fraction of 48% and multisegmental wall motion abnormalities. Coronary angiogram revealed a 70-80% stenosis of the mid portion of the left anterior descending artery. There is a fistulous vessel draining to the main pulmonary artery. The left circumflex is super dominant with an 80-90% stenosis at its termination and supplying the right coronary circulation. The right coronary artery was absent.

Conclusion: Coronary artery fistula in combination with a congenitally absent coronary artery is extremely rare. The presence of these abnormalities alone or in combination can exacerbate ischemia, complicate pre existing coronary artery disease and may cause ischemic cardiomyopathy. This case emphasizes the importance of integration of clinical as well as imaging modalities to diagnose this uncommon abnormality and in guiding clinicians for definitive management.

Keywords: congenitally absent right coronary artery, single coronary artery, coronary artery fistula, case report

Introduction

Congenital coronary artery anomalies are often incidental findings found on angiography, imaging studies or necropsy. They affect one percent of the population, and the abnormalities range from anomalous origin, distribution, intercoronary communications, agenesis of the ostium and coronary artery fistulae.[1] These anomalies may be of no hemodynamic significance or may present with symptoms of myocardial ischemia, heart failure and arrhythmias. A single coronary artery and coronary artery fistulas are both extremely rare findings. There have only been a few reported cases of this combination. We present an unusual case of an absent right coronary artery with a coronary artery fistula to the main pulmonary artery.

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Case Presentation

A 46-year-old male, hypertensive and non diabetic, came in due to persistent chest pain on exertion accompanied by easy fatigability. Symptoms would be reportedly relieved by rest until few hours prior to admission when there was sudden onset of severe chest pain graded 9/10 and unrelieved by rest. Patient was then brought to our institution and was subsequently admitted. He is a known hypertensive, non diabetic. Family history is positive for diabetes and hypertension. He is a previous smoker and occasional alcoholic beverage drinker and denies history of illicit drug use. On physical examination, patient was conscious, coherent with a blood pressure of 140/80, heart rate of 76 beats per minute. He had clear breath sounds, no murmurs and no bipedal edema noted.

Electrocardiogram revealed an old anterior wall myocardial infarct. (Figure 1) Chest radiograph showed cardiomegaly with slight pulmonary congestion. Troponin I was negative at 0.008 ng/ml. Two-dimensional transthoracic echocardiogram showed an ejection fraction of 48% with

Figure 1. Electrocardiogram shows poor R wave progression in leads V1 to v5 indicative of an old anterior wall myocardial infarction

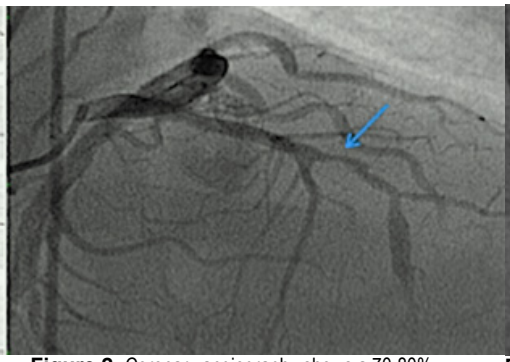

Figure 2. Coronary angiography shows a 70-80% mid left anterior descending artery stenosis (arrow) after the first diagonal branch

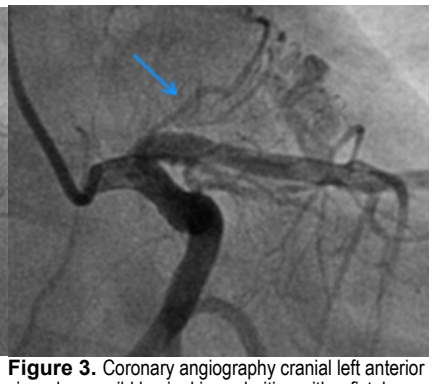

Figure 3. Coronary angiography cranial left anterior view shows mild luminal irregularities with a fistulous vessel arising near the ostium and appearing to drain to the main pulmonary artery

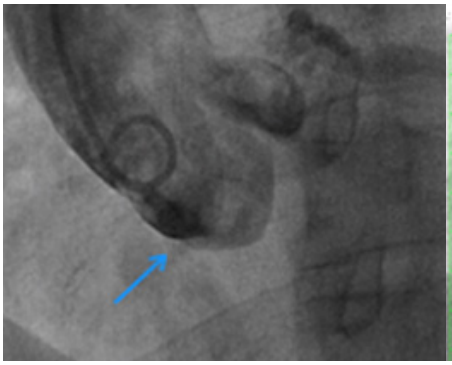

Figure 4. Aortogram contrast injection reveals an absent right coronary artery

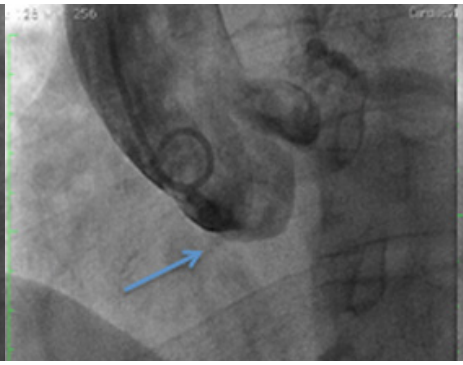

Figure 5. Coronary angiography left anterior view showing the right coronary cusp with absent right coronary artery (arrow) and contrast opacification of a coronary vessel underneath

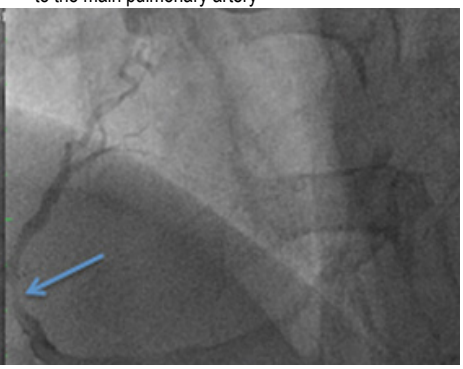

Figure 6. The right anterior caudal view showing a super dominant left circumflex artery which extends distally to the right atrioventricular groove to supply the absent right sided circulation. There is note of a 80-90% diffuse stenosis (arrow) near its termination

multisegmental wall motion abnormalities and Doppler evidence of grade one diastolic dysfunction. Patient was loaded with dual antiplatelet therapy (aspirin 325mg/tablet one dose followed by clopidogrel 75mg/tab four tabs), isosorbide drip was also started as well as atorvastatin 80mg/tab and metoprolol 50mg/tab. He was hooked to cardiac monitor and oxygen via nasal cannula at two liters per minute and was then sent in for urgent coronary angiogram. Cardiac catheterization showed the left main artery is a large normal sized vessel which bifurcated into the left anterior descending coronary artery (LAD) and the left circumflex coronary artery (LCX). There was a 70-80% stenosis of the LAD at its mid portion (Figure 2). The proximal portion of the left anterior descending artery has mild luminal irregularities with a fistulous vessel draining to the main pulmonary artery (Figure 3) The right coronary artery (RCA) has no ostium and is absent (Figure 4). The right coronary circulation receives retrograde filling from the left circumflex coronary artery (Figure 5) which was a large super dominant vessel and supplies the right coronary circulation. It has an 80-90% stenosis near its termination. (Figure 6)

Because of the presence of multivessel disease, complex anatomy and the presence of the coronary artery fistula, it was initially decided that the patient will best benefit from coronary artery bypass graft surgery with ligation of the fistula. Myocardial perfusion scan however revealed predominantly scarred myocardium in the apex, apical anterior and the adjacent septal, inferior and lateral segments. Surgery was deferred and he was then placed on anti ischemic medications with symptomatic improvement. Lifestyle advice such as exercise and proper diet were emphasized. He was advised to undergo cardiac computed tomography (coronary CT scan) to further delineate the course of the fistula and vessel anatomy but has not been able to facilitate undergoing the procedure. He is currently asymptomatic, compliant on optimal medical therapy and is clinically stable on follow up.

Discussion

A single coronary artery (SCA) is an unusual finding with an incidence of only 0.024% to 0.066% in the general population.[2] Very few cases describe the complete agenesis of the ostium of one coronary artery. It is suggested that the reason for the single artery to form is because of the underdevelopment of the proximal portion of the coronary artery which is expected to grow into the aorta during the final stages of the coronary artery formation during organogenesis in utero.[3] Lipton et al. classified the SCA in nine patterns according to the origin, anatomical course, and termination of the anomalous vessel.[4] Our patient has Type IA which is the most common type. Type IA is a single

coronary artery that divides into the left anterior descending coronary artery and the left circumflex coronary artery. The LCX then courses in the left AV groove to reach the crux, and then continues onward to occupy the anatomic position normally occupied by the RCA. Coronary artery fistulas are likewise rare findings occurring in about 0.2-0.4% of congenital cardiac defects.[5] They may arise from any of the major three coronary arteries, however, in most of the cases the coronary artery fistula arises from the right coronary artery, or the left anterior descending coronary artery; the circumflex coronary artery is rarely involved. Over 90% of the fistulas drain into venous structures such as right sided chambers, main pulmonary artery, coronary sinuses, and superior vena cava.[6] While many of these coronary anomalies may be asymptomatic, Shirani and colleagues[7] reported that 15% of the patients with SCA had myocardial ischemia as direct consequence of the anomaly. Various mechanisms, including anatomical features such as the compression of the anomalous vessel along its course between the aorta and the pulmonary artery or sporadic spasm of the anomalous coronary artery, have been postulated. Coronary artery fistula causes myocardial ischemia both by producing a coronary steal and by imposing an additional volume load on the left ventricle. Symptomatic adult patients may have symptoms of dyspnea, fatigue and angina, which may also be due to concomitant presence of underlying cardiac disease.

Coronary angiography remains the gold standard of diagnosis. In some cases however, it may sometimes fail to completely show the complex nature of these anomalies, as well as delineation of the exact course of the anomalous vessel. Other non-invasive methods such as contrast enhancing multislice computed tomography and magnetic resonance imaging (MRI) can be used with coronary angiography. They can also be crucial in planning for therapeutic intervention.[8]

It is possible that our patient had suffered a myocardial infarction prior to consult. The presence of significant two vessel coronary artery disease and the combination of his congenital anomalies were causing additional ischemic burden. Patients with a single coronary artery are particularly susceptible to the deleterious effects of atherosclerotic occlusive disease as the heart and the conduction system entirely depend on the SCA for oxygenated blood supply[9] while fistulas associated with atherosclerotic disease have been shown to cause myocardial infarction and heart failure[10] likely due to the mechanisms previously described. This combination likely had an additive contributory role to the presence and development ischemic cardiomyopathy in our patient as evidenced by extensive scarred myocardium on perfusion imaging. This made treatment particularly challenging, as we had to defer our initial surgical plan of coronary artery bypass graft with ligation of the fistula and instead optimize medical management for the time being. Our patient is compliant with treatment and is on close follow up with no fatal adverse events or re-hospitalizations.

In the review by Ishii et al. there have been approximately 35 cases of single coronary artery combined with coronary artery fistula in literature to which the authors were able to describe the varying combinations in 19 cases.[11] To the best of our knowledge the combination in our patient (absent right coronary artery with a coronary artery fistula from the left anterior descending artery to the main pulmonary artery) has never been previously reported.

This case emphasizes the importance of a meticulous and careful attention to detail integrating clinical history and physical examination as well as targeted adjunctive imaging procedures. As these are rare anomalies where findings can surprise and challenge the clinician, proper incorporation of diagnostic testing with sound clinical judgment as well as long tern follow up is required to determine the definitive management and prognosis of this uncommon abnormality.

Conclusion

Single coronary artery and coronary artery fistulas respectively are extremely rare findings as isolated entities. In combination they are even more unusual. Patients may be asymptomatic or may present with ischemic symptoms, which can place additional burden to existing coronary artery disease causing ischemic cardiomyopathy. Diagnosis is confirmed with a combination of clinical as well as invasive and non-invasive modalities that will serve to guide definitive medical and surgical management.

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