Chest Pain due to Atherosclerotic Coronary Artery Disease in a Patient with Single Coronary Artery

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Abstract

Coronary artery anomalies are rare, and single coronary arteries are even rarer occurring 0.024%-0.066% in the general population. They range in presentation from being asymptomatic to severe chest pain and even sudden death. There are numerous variations of Coronary artery anomalies, some benign and others potentially lethal. Benign variations include separate origination of the left anterior descending and left circumflex arteries from the left sinus of Valsalva, an ectopic origin of right coronary artery or left main trunk from the posterior sinus of Valsalva, and intercoronary communication. Potentially serious anomalies, which constitute 19% of anomalies, include an ectopic coronary origin form the pulmonary artery, an ectopic origin of the left coronary artery from the right sinus of Valsalva, and a Single Coronary Artery. We present a case of a 65-year-old lady who presented with chest pain and was diagnosed, incidentally on cardiac catheterization as having a single coronary artery supplying the entire heart. The cardiac catheterization showed the patient did not have a right coronary artery. Rather, left circumflex branch of left main coronary artery continued as right coronary artery to supply the right side of the heart. The catheterization also showed a 90% stenosis to the proximal diagonal 1 branch of the Left anterior descending; a percutaneous coronary intervention was performed to the diagonal 1 lesion. The patient was chest pain free upon discharge and instructed to follow up in cardiology clinic.

Keywords

Coronary Artery Anomalies (CAA), Single Coronary Artery (SCA), Right Coronary Artery (RCA), Left Main Coronary Artery (LMCA), Left Anterior Descending Artery (LAD), Left Circumflex Artery (LCX), Percutaneous Coronary Intervention (PCI)

Introduction

Coronary artery anomalies are relatively rare in general and range in a variety of presentations. Single coronary artery anomalies are extremely rare with an incidence of only 0.024%-0.066% in the general population undergoing coronary angiography [1]. Although rare, these present clinically in a variety of different ways ranging from being asymptomatic to patients experiencing severe chest pain and sudden death depending on the course of anomalous anatomy of the vessels [2-4]. CAA come about from abnormalities during the third week of fetal development but usually do not manifest symptomatically until adulthood [5]. These anomalies have associated abnormalities such as transposition of the great vessels, coronary arteriovenous fistula, or a bicuspid aortic valve [5]. Most CAA are found incidentally on angiography. We present a case of a 65-year-old lady who presented with chest pain and was diagnosed, incidentally on cardiac catheterization as having a single coronary artery supplying the entire heart.

Case Presentation

A 65-year-old Polish female with a past medical history of hypertension and diabetes mellitus II presented to ED with the complaints of sharp, substernal chest pain 8/10 in severity, associated with diaphoresis, radiating to left arm, relieved by sublingual nitroglycerine. Her home medications included Metoprolol, Metformin, and Captopril. On physical examination, the patient’s vitals include BP 149/89, pulse rate 89, respiratory rate 18, and SaO₂ 99% on 2 L of O₂ by nasal canula. Her initial ECG showed ST-depression in lead II and III. The echocardiogram showed an ejection fraction of 55-60%. The patient was subsequently placed on nitroglycerin and heparin drip and transferred to the Coronary Care Unit for close monitoring until cardiac catheterization the following day. The cardiac catheterization showed the patient did not have a right coronary artery. Rather, the blood supply to the heart had all originated from the LMCA. It was found, incidentally, that the patient had a LMCA giving rise to the LAD and LCX arteries. The LCX artery then continued as the RCA to supply the right side of the heart. The LMCA showed 0% stenosis, the LAD showed 30% stenosis, the D1 artery showed 90% stenosis, and the LCX artery showed 30% stenosis. The post-catheterization diagnosis was a 90% proximal D1 lesion, and an absent RCA. The plan after catheterization was for the patient to do a Lexiscan stress test to assess for ischemia in the D1 territory, and if positive, a PCI to the D1 would be performed. The Lexiscan stress test showed nonspecific T wave changes at rest. Following Lexiscan infusion, ST segment depression appeared with 0.5-1 mm ST depression, especially in the inferior and lateral leads suggestive of myocardial ischemia. No significant arrhythmias were noted. Post Lexiscan gated images revealed no significant wall motion abnormality with an EF = 74%. Patient underwent successful PCI and stent to D1 branch of LAD. She was later discharged on Aspirin, Plavix, a β-blocker, and an ACEI and instructed to follow up in cardiology clinic.
Discussion

There are numerous variations of coronary artery anomalies, some benign and others potentially lethal. Some benign variations include separate originations of the LAD and LCX arteries from the LSV, an absent LCX artery, a LCX artery originating from the RCA or RSV, an ectopic origin of RCA or left main trunk from the PSV, an ectopic coronary origin from the ascending aorta, and intercoronary communication [2]. Potentially serious anomalies, which constitute 19% of anomalies, include an ectopic coronary origin form the pulmonary artery, an ectopic origin of the LCA from the RSV, an ectopic origin of the RCA from the LSV, and a single coronary artery. Coronary artery fistulae may be benign or potentially serious [3]. Alarming symptoms of patients with serious anomalies include angina pectoris, MI, arrhythmias, syncope, sudden cardiac death (SCD) [6].

Fortunately the most common variants among these are the separate origination of LAD and LCX arteries from the LSV, occurring 0.41% of the time followed by the LCX artery originating from the RCA, with the RCA originating from the LSV or LMCA, both of which are benign variants [7,8]. As discussed in our case, we presented a patient with a SCA, which may or may not pose a serious threat based on the anomalous anatomy of the vasculature.

The classification system used to describe the variations of SCA anomalies was originally described by Lipton and further modified by Yamanaka & Hobbs [3]. The coronary artery system was classified as “R” when the vessel originated from the right sinus of Valsalva and “L” when it originated from the left sinus of Valsalva. The artery is further given a group I, II, or III classification. Group I indicates an anatomical course of either an RCA or LCA. Group II come from the proximal part of the normal left or right coronaries, and crosses the base of the heart before continuing in the normal position of the coronary artery. Group III anomalies are where the LAD and LCx arteries arise separately from a normal RCA. The final descriptor is either “A,” “B,” or “P” indicating the anatomical relationship between the anomalous artery and the aorta and pulmonary artery. The most important risk is when the anomalous artery originates from the opposite aortic trunk (i.e the RCA originating from the left aortic trunk). If the RCA were to originate from a left coronary artery, be it the LAD, LCX, or LMCA, the route in which it takes plays a critical role in the patient’s prognosis as noted earlier as “A,” “B,” & “P.” An anomalous artery that passes anteriorly or posteriorly poses less of a threat clinically than one that passes between the two main vessels. This is due to the compression of the anomalous artery by either the aorta or pulmonary artery, particularly during exercise leading to SCD or MI [4,9]. Roughly 15% of patients with a SCA might develop ischemia in those arteries due to this anatomic location relative to the pulmonary artery and the aorta, without having stenosis itself [10]. Therefore, in such cases surgery would be indicated.

Traditionally, pulmonary artery catheterization was used to determine the course of the anomalous artery but now multi-slice computed tomography (CT) is the preferred diagnostic tool as it has a high sensitivity and specificity and is non-invasive. People presenting with recurrent symptoms need imaging. One must also account for aortic valve abnormalities so an echocardiogram is encouraged. Treatment should be based on the course of the vessel and the development of atherosclerosis [5].

Our patient had a missing RCA with right heart perfusion originating at the LCA, which gave rise to the LAD and LCX. The LCX was found to be dominant and continued as the RCA branch that wrapped around the heart, classifying her as L1. Our patient has, without a doubt, a left-dominant heart. This anatomical configuration poses a potentially significant threat as a proximal lesion at the LCA can occlude all coronary circulation. Other mechanisms of SCD include a spasm of the anomalous artery secondary to endothelial damage or ischemia, an acute angle of take-off of the vessel, a slit-like orifice or an intramural course (9). Compression of the vessel between the aorta and pulmonary artery would not be of concern as this RCA is a continuation of the LCX and does not branch off the proximal LCA to trace between the two large vessels.

Figure 1: Cardiac Catheterization showing Left Main Coronary Artery (LMCA) dividing into Left Circumflex Artery (LCFx) and Left Anterior Descending Artery (LAD). LMCA continuing to supply Right side of Heart.

Figure 2: Congenital Absence of Right Coronary Artery (RCA)

Figure 3: Congenital Absence of RCA
As mentioned previously, the cardiac cath showed the patient missing a RCA with all blood supply to the right side of the heart originating from a dominant LCX artery. In this case, it is imperative that this vessel is maintained patent for adequate perfusion. There was no need to treat the anomalous vessel as it was patent (and found incidentally) but the D1 lesion was significant and was stented and since then the patient has done very well and remains chest pain free.

Disclosures

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References