One Artery to Rule it All: A Case of 30-year-old Male with Single Coronary Artery

Andre Russell F. Banluta, MD | Jill Buensuceso, MD | Mylene Cornel, MD | Bernadette Semilla-Lim, MD | Renelene Macabeo, MD
Our Lady of Lourdes Hospital, Sta. Mesa, Manila

Abstract

Single coronary artery (SCA) is a rare congenital anomaly in which there is an isolated coronary artery that arises from a single coronary ostium and provides coronary blood supply to the entire myocardium. SCA is classified based on Lipton Classification which includes the origin, branching pattern and course. We aim to present a rare case of congenital anomaly that presented with atypical symptoms and to discuss the features and classification of a single coronary artery. This is a case of a 30-year-old Filipino, male, known type 1 diabetes mellitus came in with diarrhea and vomiting, which later had severe abdominal pain. A consideration of Mesenteric ischemia was entertained due to abdominal pain not compatible with physical exam findings. He then underwent CT angiogram of the abdomen which was unremarkable, so a consideration of atypical presentation of chest pain was considered since patient is diabetic hence he underwent CT coronary angiogram and was noted to have SCA as an incidental finding.

INTRODUCTION

Single coronary artery (SCA) is described as one coronary artery arising from the aortic trunk by a single coronary ostium and providing for perfusion of the entire myocardium.[1] Single coronary artery is a rare congenital anomaly of the coronary arteries, with an estimated incidence rate ranging between 0.024% to 0.066% of the general population of patients that underwent coronary angiogram. With currently unknown overall survival rate. Here, we report a rare case of congenital anomaly that presented with atypical symptoms and discussed the features and classification of an SCA.

CASE PRESENTATION

The patient is a 30-year-old Filipino male, diabetic, who initially presented with vomiting and diarrhea. He has no familial history of heart disease, and no known history of heart disease.

On initial assessment, patient was hypotensive, slightly tachycardiac, afebrile and not in cardiorespiratory distress. A fluid challenge was given, and patient became normotensive immediately. Laboratory exams and ECG were done which were all unremarkable.

Patient was treated as a case of acute gastroenteritis with severe dehydration. On the succeeding hospital days, patient was noted to have resolution of diarrhea and vomiting. However, he developed severe crampy epigastric pain which was not compatible with physical examination findings. A consideration of Mesenteric ischemia was entertained. He then underwent CT angiogram of the abdomen which was unremarkable, however a CT coronary angiogram was also done to rule out coronary artery disease as a possible atypical cause of the abdominal pain since patient is diabetic. The CT coronary angiogram showed a single coronary artery arising from the right aortic sinus measuring 5mm in diameter and free of disease [Figure 1 and 2]. Since the coronary arteries were free of disease, he was then treated clinically as mesenteric ischemia due to the atypical abdominal pain and managed conservatively by giving narcotic analgesics. He was subsequently discharged improved and sent home with insulin, antiplatelet and betablocker. Upon follow up, patient noted no recurrence of abdominal pain, and remains to have no chest pain or dyspnea. He was advised for further evaluation once he developed cardiac symptoms such as angina, exertional dyspnea or palpitations.

DISCUSSION

We have a 30-year-old male, known diabetic who came in due to vomiting and diarrhea. Whom later on during the course of the hospital stay developed severe abdominal pain. CT angiogram of the coronaries was done later in the course which revealed a rare vascular anomaly.

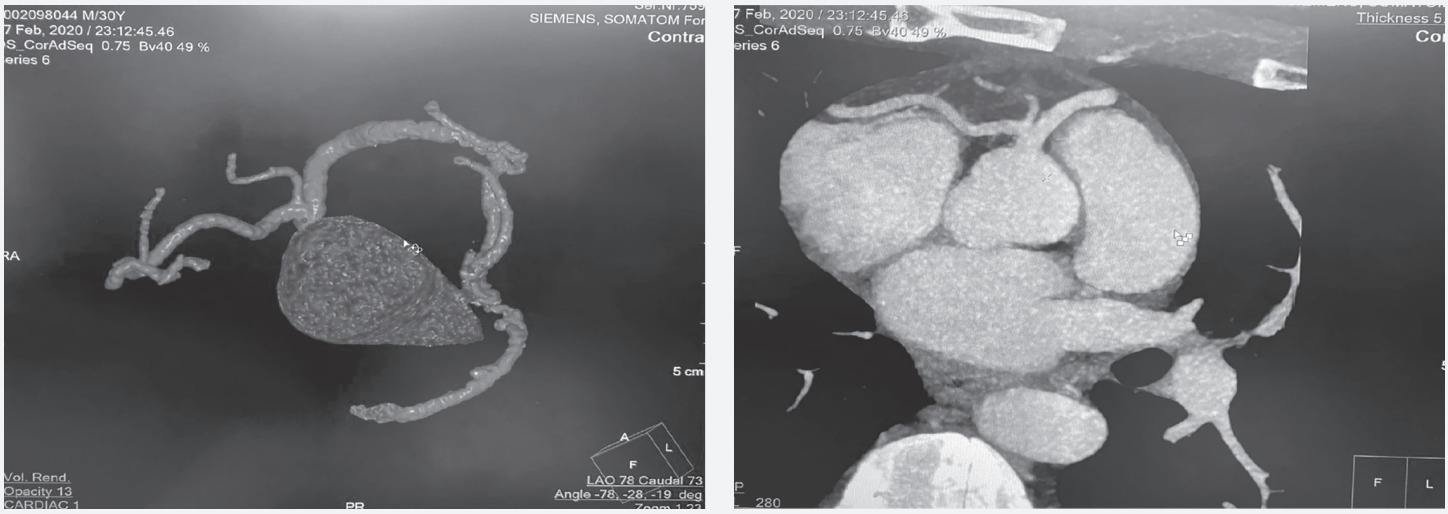

Figure 1. Shows the CT coronary angiogram (right) with the 3D reconstruction of the coronary arteries (left)

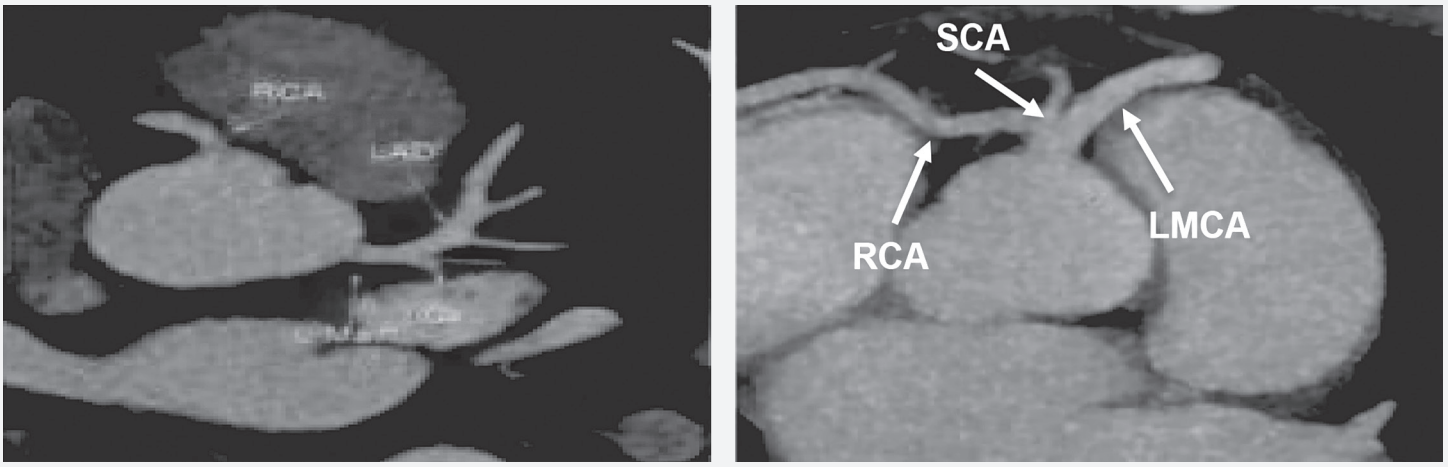

Figure 2. The picture on the left shows the normal configuration of the coronary arteries. The picture on the right is the CT coronary angiogram of the patient showing a single coronary artery (SCA) arising from the right aortic sinus and bifurcates into the right coronary artery (RCA) and left main coronary artery (LMCA)

Single coronary artery is a rare congenital anomaly of the coronary arteries, with an estimated incidence rate ranging between 0.024% to 0.066% of the general population of patients that underwent coronary angiogram. Currently there are no local statistics available for the Philippines. In the restrospective study of Al Umairi and Al-Khouri, the prevalence of SCA was 0.27% that is higher than the figures from previous reports. Among the 12 patients with single coronary artery, mean age was 56.4 years (age range: 34 to 71 years; male to female ratio 5:7.[2]

It is a vascular anomaly that only presents with one coronary artery arising from the aortic trunk supplying the entire heart.[1] Coronary congenital absence is thought to be caused by a defect of coronary development during the embryonic period resulting in a coronary artery anomaly.[3]

The detection of SCA is usually seen as an incidental finding in coronary angiogram. Most patients with SCA are usually asymptomatic at the time of diagnosis they can also present with a wide variety of symptoms, ranging from being completely asymptomatic, to anginal pain, syncope, ventricular tachycardia, myocardial infarction or sudden cardiac death.[4,5] In which, in our case, the diagnosis of SCA was an incidental finding. SCA is usually seen as an incidental finding during a conventional coronary angiogram, CT angiogram, and cardiac MRI.

We classify SCA according to Lipton's classification, wherein SCA is divided into two main types: "R," right type, and "L," left type, to indicate the origin of the SCA from the right or the left coronary sinus, respectively.[6] SCA is further divided into three subtypes based on the anatomical course of SCA. Type I in which there is a single coronary artery arising from either the right or the left coronary sinus is right coronary artery (RCA), and then it continues in the left atrioventricular groove as left circumflex artery (LCX) before it terminates as left anterior descending artery (LAD), or as Left main coronary artery (LMCA) that gives LCX and LAD; the RCA formed is a continuation of the LCX. In type II, a single coronary artery arises from either the right or the left coronary sinus, and from the proximal segment of this single coronary artery, an anomalous artery arises that crosses the base of the heart before it resumes its inherited course. In type III, the LCX and LAD have separate origins from the proximal RCA [5]. Based on the relationship of the SCA to the aorta and the main pulmonary artery, Lipton et al. categorized SCA into three categories: category A in which the anomalous artery passes anterior to the main pulmonary artery, category B in which the anomalous artery passes between the ascending aorta and the main pulmonary artery, and category P in which the anomalous artery passes posterior to the ascending aorta.[5] In this case, the classification is type IIB1. In one study, the most common class was RIII-C. Other SCA included RII-C, RII-A, and RII-S, two in each class. One patient had RI and one had LII-P. Two patients had coronary artery bypass graft. No major adverse cardiac events were reported over a mean follow-up of 25.3 months.[2]

Prognosis and future clinical course are currently unknown; there is no current guidelines on the management of patients with SCA. Coronary angiography remains the gold standard for diagnosis and classification. Echocardiography is useful mainly to delineate other structural abnormalities accompanying the SCA. Computed tomography angiography offers a less-invasive imaging modality despite requiring administration of contrast media. Cardiovascular magnetic resonance is a suitable alternative investigation for assessing coronary anatomy.[4] Treatment options include conservative medical management, percutaneous coronary intervention with stent placement, and surgical correction. Most asymptomatic patients do not require invasive therapy and should be managed with strict control of risk factors which was done in this case.

Revascularization is recommended only if there is significant atherosclerosis and documented ischemia.[6] In cases of SCA poses certain technical challenges, so awareness of these variations is important for catheter-based treatment. Surgical options include osteoplasty, coronary artery bypass grafting of the anomalous artery, re-implantation of the anomalous artery to the aorta, and pulmonary artery translocation.[7]

A multidisciplinary team of cardiologists and cardiothoracic surgeons is recommended to manage the patient if there is atherosclerosis or defects in the single coronary artery to optimize coronary function and subsequent survival..

CONCLUSION

This is a case of a 30-year-old Filipino, male, known type 1 diabetes mellitus came who was initially treated as a case of severe Acute Gastroenteritis, but due to severe abdominal pain not compatible with physical exam findings, mesenteric ischemia was considered. CT angiogram of the abdomen was done which was unremarkable, hence a consideration of atypical presentation of chest pain was also considered. A CT coronary angiogram was done and noted to have SCA as an incidental finding. Patient was treated conservatively with IV narcotics with noted resolution of abdominal pain, and was sent home with an antiplatelet and betablocker. Upon follow up there was no reported incidence of chest pain, dyspnea, or any recurrence of abdominal pain. Single coronary artery is a very rare congenital anomaly that is discovered as an incidental finding or when investigating for a coronary artery disease. A CT coronary angiogram can provide us with a detailed picture of the anatomy and course of the coronary arteries. Since the patient was asymptomatic for cardiac disease, he does not require any invasive therapy and only managed with strict control of risk factors.

RECOMMENDATIONS

Currently, there are no clinical guidelines for treatment of patients with single coronary artery. Revascularization or surgical intervention is warranted in the presence of significant atherosclerosis and the presence of ischemia, which was not seen in this patient during the time of the CT coronary angiogram. Since patient is diabetic which predisposes the patient to atherosclerosis, careful monitoring and control of comorbidities is required.

The general population of patients with SCA is low, hence adequate prognostication is difficult. Studies on patients identified with SCA should be done to help us provide predictors and outcomes to the overall survival rate of future

patients diagnosed with single coronary artery disease. A prospective study is recommended for this purpose. Comparative studies with different imaging modalities is recommended to delineate the degree of accuracy of each imaging modality.

REFERENCES

1. Desmet W, Vanhaecke J, Vrolix M, et al. Isolated single coronary artery: a review of 50 000 consecutive coronary angiographies. *European Heart Journal*, vol. 13, no. 12, pp. 1637–1640, 1992. e
2. Al Umairi and Al-Khouri. Prevalence, Spectrum, and Outcomes of Single Coronary Artery Detected on Coronary Computed Tomography Angiography (CCTA). *Hindawi Radiology Research and Practice* Volume 2019, Article ID 2940148.
3. Forte E, Inglese M, Infante T, et al. Anomalous left main coronary artery detected by CT angiography. *Surg Radiol Anat* 2016;38:987–90.
4. Akcay A, Tuncer C, Batyraliev T, Gokce M, Eryonucu B, Koroglu S, Yilmaz R. Isolated single coronary artery. *Circ J.* 2008;72:1254–1258. doi: 10.1253/circj.72.1254
5. Yamanaka O, Hobbs RE. Coronary artery anomalies in 126,595 patients undergoing coronary arteriography. *Cathet Cardiovasc Diagn* 1990;21:28–40.
6. Yan GW, Bhetuwal A, Yang GQ, et al. Congenital absence of the right coronary artery: a case report and literature review. *Medicine* 2018;97:e0187.
7. Elbadawi A, Baig B, Elgendy IY, et al. Single Coronary Artery Anomaly: A Case Report and Review of Literature. Cardiol Ther. 2018 Jun;7(1):119-123. doi: 10.1007/s40119-018-0103-4. Epub 2018 Feb 6. PMID: 29411245; PMCID: PMC5986668.