Abstract

Single coronary artery anomalies are those where the entire myocardium is supplied by an artery arising from one ostium. It is a rare coronary anomaly and has been there in literature ever since 1867. Over the years, this has intrigued anatomists and physicians whether its presence has life-threatening consequences or is mere a benign entity. Most of the cases are generally silent except for the interarterial course variants which are associated with sudden deaths. We report the case of a patient presenting with recurrent inferior wall ST-elevation myocardial infarction who was found to have a single coronary artery on angiography. The patient underwent primary percutaneous coronary intervention and uneventful further course.

Keywords: Absent left main, congenital heart disease, single coronary ostium, acute coronary syndrome, Lipton's classification

Introduction

Single coronary artery is a rare anomaly occurring in about 5%-20% of major coronary artery anomalies.^[1] When occurring in morphologically normal heart, single coronary artery has normal distribution of branches in the left anterior descending and circumflex areas, but distribution may be ambiguous with congenital heart diseases.^[2] It is a benign entity in most patients, but it can be symptomatic too and may result in grave consequences including acute coronary syndrome (ACS).[3-5]

Case Report

A 35-year-old male presented with inferior wall ST-elevation myocardial infarction (STEMI) [Figure 1]. The patient was a known diabetic, hypertensive, and with dyslipidemia. He had a history of prior inferior wall infarction a year ago, for which he was treated by primary percutaneous intervention (PCI) to the right coronary artery (RCA). During prior ACS episode, the treating physician noted an additional vessel originating abnormally from the right coronary sinus (RCS) which was assumed to be left circumflex artery (LCX). Left injection was not be taken at that time as patient was hemodynamically unstable. He was discharged on secondary prevention drugs including dual antiplatelet therapy and was advised coronary computed tomography to delineate the course of anomalous artery which patient did not undergo. Noncompliant on treatment, he presented to us again with inferior STEMI. This time he was hemodynamically stable. Transthoracic echocardiogram revealed segmental wall-motion abnormalities in inferior, posterior, basal septal, and lateral walls of left ventricle. His cardiac biomarker (cardiac troponin I) levels were elevated (1.5 ng/ml, N < 0.4 ng/ml), but the rest of blood investigations including hemogram and renal function test were within normal limits. The patient was taken up for primary PCI through right radial access. Despite multiple attempts, left main coronary artery could not be engaged. Hence, a RCA injection was taken first. A short common vessel was noticed in the right sinus, from which two coronary arteries were emanating [Figure 2]. One of the vessels continued as RCA with a normal course. The second vessel originating from the common ostium was left coronary artery which was dividing into LAD and LCX [Figure 3]. Two discrete lesions were noted in RCA, and a stent was visible in proximal RCA which was patent. Proximal to stent, there was 70% stenosis

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and distal RCA also had 70% stenosis. LAD was also diseased in its proximal segment with 70%-80% stenosis. Because of multiple lesions in RCA and LAD and diabetic status of patient, he was given option between CABG and multivessel PCI. Fortunately, he opted for multivessel PCI. RCA was taken up for PCI, and lesion was wired with a floppy wire (Runthrough NS, Terumo Inc, Somerset, NJ, USA). The lesions were predilated using 2.5 mm \times 12 mm semi-compliant balloon catheter [Figure 4]. After that two nonoverlapping everolimus drug-eluting stents deployed in sequence, proximal one first followed by the distal one. The proximal stent was 3.5 mm \times 24 mm in size, while the distal stent was $3.0 \text{ mm} \times 18 \text{ mm}$ in size, respectively. Both stents were postdilated with upsized noncompliant balloons as IVUS run could not be performed. A staged PCI was planned for the noninfarct-related artery lesion in the proximal LAD.



Figure 1: A 12-lead electrocardiography at presentation depicting inferior wall MI (ST elevation leads II, II, and aVF; Left panel) with right ventricular (ST elevation in RV3-RV5 leads; right panel) and posterior wall involvement (ST elevation in V7-V9 leads; right panel)



Figure 3: Distribution of left coronary artery as seen in LAO projection with cranial angulation. LAD: Left anterior descending artery; LCX: Left circumflex artery; LMCA: Left main coronary artery; LAO: Left anterior oblique

The patient remained stable throughout the hospital course and was discharged on dual antiplatelet therapy (aspirin 150 mg and ticagrelor 90 mg bid) and other guideline-directed medical therapy for secondary prevention.

Discussion

By definition, single coronary artery is a condition, in which the entire myocardium is nourished by one artery arising by one ostium from an arterial trunk, regardless of its distribution. This rare coronary anomaly has been reported in literature as early as Bochdalek in 1867.^[6] It contributes about 5%–20% of major coronary artery anomalies.^[1] Sometimes, an atretic cord connects the part of the artery to a sinus of Valsalva that has no ostium. In 40% of cases, it is seen in association with other cardiac malformations such as tetralogy of Fallot, transposition of great vessels, truncus arteriosus, coronary-cameral fistulas, and bicuspid aortic valves. In morphologically normal



Figure 2: Angiographic views in LAO (left panel) and RAO (Right panel) projection depicting common origin of right and left coronary artery with single ostium with anterior course of LCA. LCA: Left coronary artery; RCA: Right coronary artery; CCA: Common coronary artery; LAO: Left anterior oblique; RAO: Right anterior oblique



Figure 4: Angiographic image of RCA showing lesions proximal to previous stent (white arrow) and in distal RCA (black arrow). The image is after predilatation with semi-compliant 2.5 × 12 balloon catheter. RCA: Right coronary artery



Figure 5: Classification of single coronary artery (Modified from Lipton *et al.*). CCA: Common coronary artery; *: Type III variant of single coronary artery usually arises only from right sinus and the distribution of traversing branches is combined type

heart, this anomaly has normal distribution of LAD and LCX; however, the distribution may be ambiguous if it occurs in association with congenital heart diseases.^[2]

The single artery can arise from either the left or RCS with varied distribution.^[6,7] On either side, it can have its normal course and then continue on to supply the other side of the heart or a main coronary artery can divide into right and left branches and course posteriorly or anteriorly to supply the other side of the heart. Branches also can course between the great vessels. The classification of single coronary artery with the distribution of branches was originally described in 1979 by Lipton *et al.* and a simplified version is described in Figure 5.^[6] Our case was a Type II single coronary artery variant from right sinus with an anterior course.

Although the coronary angiogram remains the gold standard for diagnosing congenital coronary anomalies, coronary computed tomography angiography (CCTA) has emerged as an essential tool to delineate the course of anomalous coronary vessels. Three-dimensional reconstructed images from CTA provide excellent spatial orientation for the origin and route traversed by the single coronary artery, especially for the inter aortopulmonary course. A recent CCTA-based study found that the prevalence of single coronary artery to be 0.27%. Moreover, all but one originated from right sinus.^[7] Type III variant was the most frequent among their cases.

Most single coronary artery cases are asymptomatic in the absence of coronary artery disease, but a small number of premature deaths have been reported with this anomaly.^[8] In 1956, E. Dubose Dent and Russell concluded that, in the absence of cardiovascular disease or other anomalies of the heart, a single coronary artery is not associated with decreased cardiac function.^[3] There may be symptoms of

myocardial ischemia such as angina and ischemic mitral regurgitation but variant with interarterial distribution of a major branch is at risk for sudden death.^[4,5,9,10] In the recent single-center experience discussed above, there were no adverse events at 2-year follow-up.^[7] Recurrence of STEMI, in this case, cannot be attributed solely to anomalous coronary anatomy only as noncompliance to the secondary prevention therapy after MI (as in our case) is also a pivotal contributor to adverse cardiovascular events.^[11]

Like various coronary anomalies, the presence of single coronary artery has surgical implication and demands expertise in congenital heart surgery operation, especially the arterial switch operation.^[12] However, because of rarity of the condition, we feel that every case deserves to be reported and added to the existing literature.

Conclusion

While a benign entity in most cases, single coronary artery can be symptomatic. We assume that pathology in our case was unrelated to coronary anomaly itself and rather of atherosclerotic origin. However, as cardiologists, we should be aware of this rare entity, as it carries a potential risk of sudden cardiac death, especially in association with other congenital heart disease and malignant course.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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