Case Report

A rare case of isolated single coronary artery, Lipton’s type LIIB diagnosed by computed tomography coronary angiography✩,✩✩

Vikash Bhattarai, MBBS, MDa,*, Sandeep Mahat, MBBS, MDa, Asim Sitaula, MDb, Nirmal Prasad Neupane, MDh, Kritisha Rajlawot, MDb, Sujit K Jha, MDb, Saroj Chetry, MScMITb

aDepartment of Radiodiagnosis and Imaging, National Academy of Medical Sciences, Bir Hospital, Kathmandu, Nepal
bDepartment of Radiodiagnosis and Imaging, Shahid Gangalal National Heart Centre, Bansbari, Kathmandu, Nepal
cUC Davis Health, Sacramento, CA, USA

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Abstract

Single coronary artery (SCA) is a very rare coronary artery anomaly of origin and course with a reported prevalence of only 0.024%-0.066% among patients undergoing routine coronary angiography. The majority of the individuals remain asymptomatic and thus SCA is found only incidentally on conventional or computed tomography coronary angiography done for other reasons. A minority of the patients may have non-specific cardiac symptoms (such as ischemic pain, tachycardia, etc.) or even sudden death. SCA can occur in isolation or in association with other congenital cardiac defects like such as persistent truncus arteriosus, tetralogy of Fallot (TOF), pulmonary atresia, transposition of great vessels (TGA), ventricular septal defect (VSD), coronary arteriovenous fistula (AVF), patent foramen ovale (PFO) and bicuspid aortic valve. We present a case of 50 years male with incidental finding of SCA arising from the left coronary sinus which had an inter-arterial course before branching (SCA Type: LIIB based on the Lipton-Yamanaka classification) which was revealed on computed tomography coronary angiography (CTCA) performed after the patient complained of infrequent chest pain. Management of the diagnosed cases can be either conservative, stent placement or surgical correction based on the symptomatology and clinico-lab findings.

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* Corresponding author.
E-mail address: ivrisk@gmail.com (V. Bhattarai).
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Introduction

Any coronary morphology that occurs in more than 1% of the general population is considered a normal variant while the one that befalls in less than 1% is taken as an anomaly [1,2]. Coronary artery anomalies can be divided into anomalies of origin and course, anomalies of intrinsic coronary anatomy, and anomalies of termination [1]. Single coronary artery (SCA) supplying the entire heart is a very rare coronary artery anomaly of origin and course with reported prevalence of 0.024%-0.066% in the population who undergo routine coronary angiography [3,4]. SCA can be present in isolation or in association with other congenital cardiac anomalies [3]. There is a very useful classification system for SCA given by Lipton in 1979 based on the origin, course, and branching pattern of the artery, which was later modified by Yamanaka and Hobbs with the inclusion of an extra feature, that is, septal course [3]. Conventional coronary angiography is regarded as the gold standard to see coronary morphology however, non-invasive computed tomography coronary angiography (CTCA) has been increasingly used recently because of its superior capability of demonstrating the complex coronary anatomy and being a non-invasive procedure [3]. We report an incidental finding of a single coronary artery (SCA) arising from the left coronary sinus and having an inter-arterial course, in a middle-aged adult male undergoing CTCA after presenting with infrequent vague chest pain for one year.

Case presentation

A 50-year-old male, a known case of hypertension, was in regular follow-up with the cardiologist at our center, managed with regular anti-hypertensive medication for the last 10 years. The patients complained of a few brief episodes of non-radiating chest pain at rest over the period of 2 weeks. Non-specific T wave changes were noted in the electrocardiogram (ECG) after which a treadmill test (TMT) was done without significant new findings. All the relevant blood works were normal including the cardiac enzymes. Echocardiography showed no abnormality with the left ventricular ejection fraction > 60%. Then, the patient was asked to get a CTCA to see for the coronary arteries and rule out stenosis, if any present, secondary to atherosclerotic changes/plaques. A cardio-dedicated ECG gated 640 slice computed tomography (CT) scanner was used to acquire the coronary angiogram using the standard imaging protocols.

When CT images were analyzed, a single coronary artery (SCA) was discovered arising from the left coronary sinus which coursed in between the pulmonary artery and the aorta (inter-arterial course) while descending antero-inferiorly after which it bifurcated into 2 arteries, the left coronary artery (LCA) and the right coronary artery (RCA) (Fig. 2A and B). LCA was then divided into 2 arteries, the left anterior descending artery (LAD) and the (left) circumflex artery (LCX) (Fig. 2B and C). These features of a single coronary artery are consistent with SCA-LJIB type, according to the Lipton-Yamanaka classification system. The CT images did not reveal other congenital cardiac anomalies thus confirming the presence of an isolated single coronary artery in the patient.

Discussion

Before going into the discussion on coronary anomalies it is worth knowing the gross normal anatomy of the coronary arteries. The aortic root consists of 3 sinuses of Valsalva, namely right, left, and non-coronary sinuses; right and left coronary arteries arising from the so-named sinuses and the non-coronary sinus being devoid of any arterial ostium [5]. LCA, in the majority, divides into the left anterior descending artery (LAD) and the left circumflex artery (LCX). The LAD descends towards the apex of the heart along the anterior interventricular sulcus giving septal and diagonal branches [5]. The RCA passes between the pulmonary artery and right auricle and descends along the right atroventricular groove which continues into the posterior aspect as the posterior descending artery (PDA) and gives off the posterolateral branch [5].

Any coronary morphology occurring in more than 1% of the general population should be taken as a normal variant while the one occurring in less than 1% is considered an anomaly [1,2]. Coronary artery variants are benign occurrences without much clinical importance while the rarer coronary artery anomalies can range from being benign and asymptomatic to malignant entities which might even cause sudden cardiac death (SCD) [1]. Many studies suggest that coronary artery anomalies can be fatal, especially in young individuals, causing sudden death during or after strenuous exercise which otherwise remains asymptomatic and thus undiagnosed in the majority [2].

Coronary artery anomalies are usually incidental findings but are of clinical significance as they are the second most common cause of sudden death of young sport-persons (contributing about 19%) after hypertrophic cardiomyopathy [2,5]. They can be broadly classified as the anomalies of origin and course, the anomalies of intrinsic coronary anatomy, and the anomalies of termination [1]. Separate origin of the left circumflex artery (LCX) and left anterior descending artery (LAD) is the most common coronary artery anomaly which is followed by the origin of LCX from the right coronary artery (RCA) and the third most common being the origin of RCA or left coronary artery (LCA) from the opposite sinuses [6].

SCA is a rare coronary artery anomaly with only one coronary artery arising from a single ostium at one of the coronary sinuses of the aortic root and perfusing the entire myocardium [3]. Most cases of SCA remain undiagnosed as they are asymptomatic, however individuals with some sub-types of SCA may suffer from chest pain, ventricular tachycardia, syncope, myocardial infarction, or even sudden cardiac death; for instance, SCAs with inter-arterial and sub-pulmonic trans-septal courses of the transverse artery (transverse artery meaning the SCA or the opposite coronary artery relative to the originating sinus of Valsalva, right or left) can be compressed during exercise causing the cardiac symptoms [7]. Various work of literature mention the frequent association of SCA with other congenital cardiac anomalies such as persistent truncus arteriosus, tetralogy of Fallot (TOF), pulmonary
Fig. 1 – Lipton-Yamanaka classification of SCA with the inclusion of variants of SCA LIIA and SCA R-III as described by Sampath et al. [6]. PA, Pulmonary artery; Ao, Aorta; L, Left coronary sinus; R, Right coronary sinus; N, Non-coronary sinus; RCA, Right coronary artery; LCA, Left coronary artery; LCX, (Left) Circumflex artery.  

(Box A) Shows normal origin and morphology of the right and left coronary arteries arising from the right and left coronary sinuses respectively. (Box B) Shows single coronary artery, right Type I (SCA RI), and left Type I (SCA LI) originating from the right and left coronary sinuses respectively. (Box C) Shows single coronary artery, right Type II (SCA RII) variants: RIIA (LCA after arising from RCA courses anterior to the pulmonary artery), RIIB (LCA after arising from RCA has inter-arterial course), RIIP (LCA after arising from RCA courses posterior to the aorta) and RIIS (LCA after arising from RCA courses via interventricular septum). (Box D) Shows single coronary artery, left Type II (SCA L-II) variants: LIIA (2 variants namely LIIA-V1 or LIIA-V2; RCA arising from the left main coronary artery or LAD respectively and coursing anterior to the pulmonary artery), LIIB (RCA after arising from LCA has an inter-arterial course), LIIP (RCA after arising from LCA courses posterior to the aorta) and LIIS (RCA after arising from RCA courses via interventricular septum). (Box E) Shows variants of SCA RIIC (ie, RCA giving off LAD and LCX with absent LCA and (C) indicating the combined courses of the branches of SCA) namely RIII-LAD-B, RCX-P; RIII-LAD-A, RCX-B; RIII-LAD-S, RCX-P; RIII-LAD-S, RCX-B, and RIII-LAD-A, RCX-P based on courses (A, B, P and S) of LAD, and LCX as described in Table 1.
Table 1 – Lipton-Yamanaka classification coding system for single coronary artery (SCA).

| Finding | Code: |
|---------|-------|
| Right coronary sinus | R |
| Left coronary sinus | L |
| Single dominant artery following the course of either RCA or LCA-LAD | I |
| RI: Single RCA continuing in left atrioventricular groove to become LCX and terminate as LAD |
| L1: LCA is dominant with RCA arising from LCX |
| Normally located one coronary artery gives another coronary artery by early branching (ie, from proximal aspect) | II |
| RI1: LCA branches from RCA originating at right coronary sinus |
| RI2: RCA branches from LCA originating at left coronary sinus |
| Absent LCA with RCA, LAD and LCX branching from SCA at right coronary sinus | III |
| Anterior to pulmonary artery | A |
| Inter-arterial course | B |
| Posterior to aorta | P |
| Trans-septal course through interventricular septum | S |
| Combination of diverse routes | C |

LAD, Left anterior descending artery; LCA, Left coronary artery; LCX, (Left) Circumflex artery; RCA, Right coronary artery; SCA, Single coronary artery.

Fig. 2 – (A) Axial computed tomography angiographic image (in maximum intensity projection, MIP) at the level of sinuses of Valsalva demonstrates a single coronary artery (SCA) arising from the anterior aspect of the left coronary sinus; the right coronary artery (RCA) is arising from the SCA and not from the right coronary sinus; (B and C) show 3-dimensional reconstructed images confirming the same findings along with visualization of the inter-arterial course (ie, between the Ao, aorta and PA, pulmonary artery) of SCA during its antero-inferior descent before bifurcating into the RCA and the left coronary artery (LCA). These features are consistent with SCA-LIIB type according to Lipton-Yamanaka classification system. Ao, aorta; LAD, left anterior descending artery; LCA, left coronary artery; LCX, (left) circumflex artery; PA, pulmonary artery; RCA, right coronary artery; RCS, right coronary sinus; SCA, single coronary artery.

Lipton-Yamanaka classification categorizes SCA into 2 main types, “L,” that is, left type originating from the left coronary sinus, and “R,” that is, right type originating from the right coronary sinus (Table 1) [6–8]. Based on the course and distribution of SCA, there are 3 further sub-types: Type I, Type II and Type III as illustrated in Table 1 [6–9]. Lastly, based on the relation of the transverse artery with the aorta or the main pulmonary trunk, SCA is categorized as: “A,” if the course of the transverse artery is anterior to the pulmonary artery, “P,” if the transverse artery courses posterior to the ascending aorta, “B,” if the artery lies between the aorta and the pulmonary artery, that is, inter-arterial course, “S,” indicating the trans-septal course of the transverse artery, and “C,” for combined routes of the coronary arteries arising from the SCA (Table 1) [6–8]. Various types of SCA are also shown in illustrative diagrams (Fig. 1) based on the Lipton-Yamanaka classification with the inclusion of variants of some of the sub-types as mentioned in the literature by Sampath et al. [6].

Imaging modalities like conventional angiography, CTCA, and cardiac magnetic resonance imaging (MRI) can diagnose single coronary artery; conventional coronary angiography is considered the gold standard imaging modality for seeing coronary arteries [9]. Major drawbacks of the
conventional coronary angiogram are: the procedure is invasive with risk of related complications and it is inferior in delineating the anatomy of coronary arteries in complex cases [3,7]. CTCA is also emerging as the gold standard imaging modality with advantages like the procedure being noninvasive with high temporal/spatial resolution and the ability to produce 3-dimensional images for easier appreciation of coronary arteries in relation to adjacent great vessels [7]. MRI has an advantage over conventional coronary angiography and CTCA in regard to not having ionizing radiation exposure and is the sole imaging modality for assessing the viability of myocardium; however, demerits such as lesser availability of MRI machines, lower temporal/spatial resolution, and contraindication in individuals with cardiac pacemakers do not make MRI the imaging modality of choice [7]. Based on the symptomatology and clinico-lab findings, management of the diagnosed cases can be either conservative, stent placement, or surgical correction [9].

**Conclusion**

The single coronary artery is a very rare coronary artery anomaly which is usually an incidental finding in a patient undergoing routine coronary angiography for various cardiac symptoms. However, a minority of the patients can be the victims of sudden death before the diagnosis because of this anomaly. Though conventional coronary angiography is considered the gold standard modality, computed tomography coronary angiography (CTCA) is becoming an emerging gold standard modality used to study coronary morphology which can reveal coronary artery anomalies like our incidentally encountered case of a single coronary artery.

**Author contributions**

1. Dr Vikash Bhattarai – analyzed and interpreted the patient data and major contributor in writing the manuscript.
2. Dr Sandeep Mahat, Dr Asim Sitaula and Dr Kritisha Rajlawot – contributor in writing and the manuscript and literature review.
3. Dr. Nirmal Prasad Neupane and Dr Sujit K Jha – guided on how to write the manuscript and contributed in literature review and editing of the manuscript.
4. Saroj Chhetry – contributed in image selection for the manuscript.

All authors have read and approved the manuscript.

**Availability of data and materials**

All data generated or analyzed during this study are included in this published article.

**Ethics approval and consent to participate**

This case report did not require review by the Ethical committee for publication.

**Consent for publication**

Date of consent taken: August 1, 2022
Taken by: The author

**Patient consent**

Written informed consent was obtained from the patient for publication of this case report and any accompanying images assuring him about the confidentiality. A copy of the written consent is available for review by the Editor-in-Chief of this journal whenever required.

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