

Computed tomography coronary angiography diagnosis of single right coronary artery with congenital absence of left coronary artery system equivalents

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Abstract

The present case report is computed tomography (CT) coronary angiographic depiction of an exceedingly rare entity-single right coronary artery arising from the right sinus of Valsalva with the absence of equivalent left coronary artery system branches and associated mitral valve prolapse. Even though a statistical rarity, it is potentially fatal and can cause myocardial ischemia, sudden cardiac death, and warrants immediate clinical attention. Further, the report reveals the decisive role of CT coronary angiogram in the diagnosis of such rare entities, in contrast to catheter angiography, which may be inconclusive.

Key words: Absent left coronary with associated mitral valve prolapse; coronary CT angiogram; single right coronary

Introduction

Coronary artery anomalies have an incidence of 1.3% as per the largest series to date.^[1] Interestingly enough, single right coronary artery (RCA) anomalies are rarest, with estimated incidence of 0.0008%.^[1] Coronary anomalies may occur in isolation or association with congenital heart diseases such as bicuspid aortic valve, mitral valve prolapse (MVP), tetralogy of Fallot, and transposition of great arteries. Of these, the valvular anomalies are the most common, reportedly up to 29%.^[2,3] The clinical significance of coronary anomalies depends on their potential to cause myocardial ischemia and sudden death, which in turn is

dependent on their origin, course, associated congenital heart defects, and coexistent atherosclerotic lesions. Apart from a single coronary artery, malignant interarterial course of anomalous coronary artery and anomalous origin of the left coronary artery from pulmonary artery are the other potentially fatal coronary anomalies reported. Further, single coronary artery is also reported to have varied manifestations from being asymptomatic to causing sudden death.^[1,2,4]

Single anatomic RCA with superdominant branches and absent equivalents of the left coronary artery system is the rarest of the coronary artery anomalies with only three cases reported hitherto, two on necropsy,^[4] and third on dual

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source computed tomography (CT) coronary angiography.^[5] Significantly enough, none of these reported cases indicate associated MVP. The association with MVP makes the present entity (not documented in above-cited publications) at significant variance with single RCA anomalies, reported to date, and therefore notable.

Case Report

A 40-year-old male, a known case of MVP, presented with episodes of palpitations, underwent an electrocardiogram that was unremarkable with sinus rhythm. His serum lipid profile was within normal range. The two-dimensional transthoracic echocardiography revealed severe MVP with echogenic leaflets and mild diastolic dysfunction. Due to persistent symptoms in spite of medical management and reassurance, the patient was referred for surgical repair/mitral valve replacement. As part of the preoperative assessment, coronary angiography was undertaken to determine the course of coronary arteries. During catheter angiography, the left coronary artery ostium could not be cannulated. On aortogram, a blind left aortic sinus of Valsalva was noted [Figure 1A]. Upon cannulation and injection of the right coronary ostium, a single RCA was identified. No left coronary system arteries, i.e., left anterior descending (LAD) artery, left circumflex artery (LCX) were discernable [Figure 1B and C]. Multiple branches from superdominant posterior descending artery (PDA), posterolateral ventricular branch (PLVB) were observed supplying the left coronary artery territory. For further depiction of coronary anatomy, the patient was subjected to Single Source 64 Slice CT coronary angiography (Single Source GE LightSpeed VCT 64, GE Healthcare, UK), 70 ml of iohexol (300 mg I/ml) with 30 ml saline chase at rate of 4.5 ml/s using a dual-head power injector.

CT coronary angiography revealed that the left coronary ostium was absent, and a blind left aortic sinus of Valsalva was noted [Figure 2A and B (Video loop provided)]. In addition, no artery was noted to arise from noncoronary sinus or ascending aorta. A single coronary artery was observed to be arising from the right aortic sinus of Valsalva,

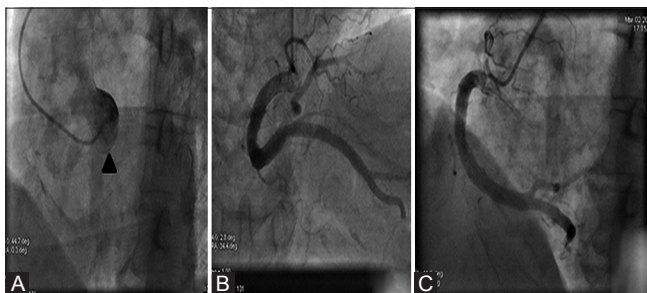


Figure 1 (A-C): (A) Aortic root angiogram depicting blind left coronary sinus of Valsalva (arrowhead). (B and C) Selective cannulation and injection of the right coronary ostium revealing the course and branches of the right coronary artery

at 12 o'clock position with a normal sinoatrial nodal branch emanating from it. Thereafter, it coursed through the right atrioventricular groove and divided into the right marginal artery, PDA, PLVB in keeping with the course of anatomic RCA. Further, a branch from proximal part of RCA was noted, supplying the proximal third of the interventricular septum. Multiple branches from PDA and PLVB supplied the left lateral ventricular wall, mid and distal third aspect of the interventricular septum, left atrium, i.e., LCX and LAD territories [Figure 2C-F]. The left main coronary artery (LMCA), LAD, and LCX were not discernable. Significantly enough, none of the superdominant branches of RCA followed the anatomic course of LCX and LAD to be considered as their equivalents. There was no evidence of

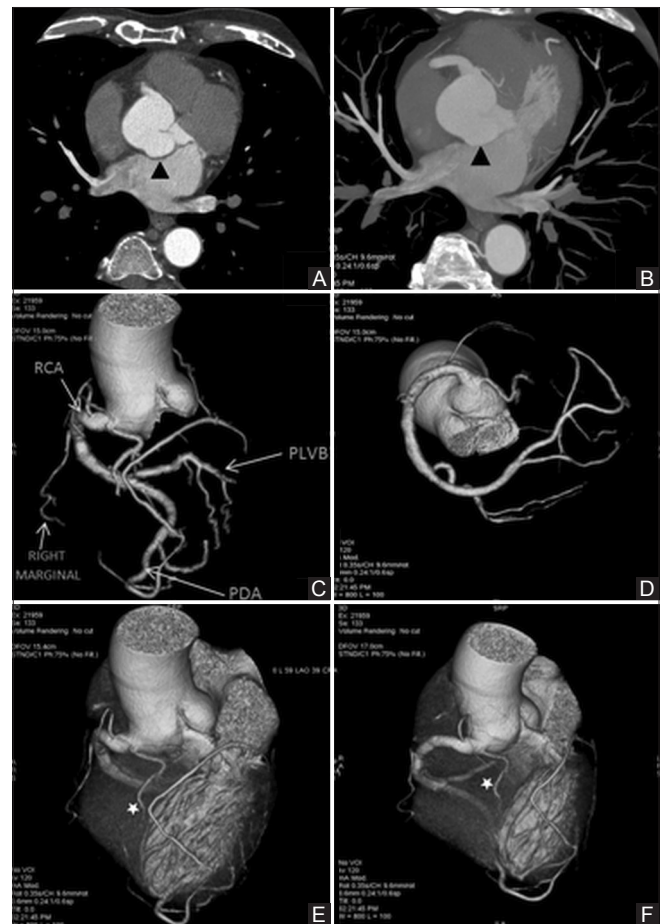


Figure 2 (A-F): (A) Axial source image and thick maximum intensity projection image (B) of computed tomography coronary angiogram illustrating single right coronary artery and a blind left sinus of Valsalva (arrowhead). (C-F) Volume rendered images showing the entire coronary tree with the right coronary artery and its branches (labeled). Branches from superdominant posterior descending artery, posterolateral ventricular branch supplying the left coronary artery territory, septal branch (*) arising from the right coronary artery. The left main coronary artery, left anterior descending, and left circumflex artery are conspicuous by their absence. MIP=Maximum intensity projection, VR=Volume rendered, RCA=Right coronary artery, PDA=Posterior descending artery, PLVB=Posterolateral ventricular branch, LMCA=Left main coronary artery, LAD=Left anterior descending, and LCX=Left circumflex artery

atherosclerosis in visualized coronary arteries or the aortic sinuses of Valsalva. A thickened mitral valve with annular calcifications, perhaps due to myxomatous degeneration was also noted. Hence, the entity was diagnosed as “single anatomic right coronary artery with no left coronary artery system equivalents and associated mitral valve prolapse.”

Discussion

Lipton's classification of single coronary artery anomalies is the most widely accepted and recognized system to date.^[6] The present coronary anomaly is at significant variance with Lipton's classification; therefore, precludes categorization into any of the types described by Lipton *et al.*, thus forming the basis for classification as a new subtype of R1 of Lipton's classification as proposed by Fu *et al.*^[5] The associated MVP makes already rare entity more noteworthy. An extensive literature search revealed yet another classification, by Shirani and Roberts,^[4] albeit least quoted and acknowledged, but indeed in reckoning the most comprehensive classification of single coronary anomalies and among others includes the presence or absence of equivalent arteries of one of the absent main coronary artery as criteria.^[4] They proposed a novel classification that included twenty possible types, encompassing all feasible anatomic combinations, and their manifestations. Following their classification, the coronary anomaly of this entity falls under class Type IIa,^[4] i.e., solitary coronary ostium in the right aortic sinus of Valsalva with absent aberrantly coursing left coronary system arteries. This category consists of single RCA with superdominant branches supplying the left coronary artery territory in place of the left main, LAD, and LCX equivalents.

The coronary anomaly reported here is rare among all the coronary artery anomalies due to absent left coronary artery equivalents. Only three prior such cases have been documented to date in literature two on necropsy^[4] and a third on dual source CT coronary angiogram by Fu *et al.*^[5] Notably enough, none of these reports had MVP as an association. Nonetheless, there exists report indicative of genetic propensity of MVP associated with the congenital coronary anomaly.^[2,3] The present coronary anomaly, with adjunct MVP as a coexisting feature, is significant variation from ones reported earlier and therefore noteworthy.

In practice, angiographic differentiation of congenital absence of coronary artery from severe atherosclerotic LMC ostial occlusion is often difficult. The presence of aberrantly coursing equivalent vessels of the left coronary artery system in addition to a blind sinus facilitates diagnosis of congenitally absent coronary artery over atherosclerotic LMC ostial occlusion on angiography.^[7] However, in view

of the absence of the left coronary artery equivalents in our case, the diagnostic dilemma of an occluded LMC ostium prevailed on angiography. The CT coronary angiographic depiction of blind left coronary sinus, as well as the arterial supply to entire heart from superdominant branches of RCA which have course different from the left coronary arterial branch equivalents, lead us to the diagnosis of “congenital single anatomic right coronary artery with absent left coronary artery system equivalents” rather than severe atherosclerotic occlusion of LMC artery.

Coronary CT angiography remains an unparalleled noninvasive imaging modality in congenital coronary anomalies and categorically demonstrates their origin, course, branches, and termination.^[8,9] The coronary anomaly with atypical characteristics reported upon represents a possible yet undescribed subtype of Lipton's Type R1^[5] or Shirani's Type IIa and the association with MVP has not documented earlier.

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Conflicts of interest

There are no conflicts of interest.

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