

A Rare Case of Single Coronary Artery Arising From Right Sinus of Valsalva Presenting As Myocardial Infarction

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Abstract:

Background: Single coronary artery(SCA) is a rare anomaly with a reported incidence between 0.024 and 0.098%. It is classified into different types based on origin, branching pattern and course. Although majority of patients are asymptomatic ,some may present with ischemia , heart failure and sudden cardiac death. We present a rare case of single coronary artery arising from right sinus of Valsalva diving into three branches. LAD had malignant course and LCX,RCA having normal course presenting as Acute inferior wall ST elevation Myocardial infarction.

Case summary: A 45-year-old postmenopausal lady with history of Diabetes mellitus presented with acute inferior wall STEMI. Conventional coronary angiography revealed single coronary artery arising from right ostium with RCA proximal cut off and filling from LAD and LCX. CCTA(Coronary computed tomographic angiography) revealed single coronary artery arising from right ostium and dividing into three branches. LAD arising anteriorly from SCA had a malignant course and LCX arising posteriorly from SCA had normal course and calibre. SCA continued as RCA with proximal filling defect. In view of anomalous coronary artery with malignant course and proximal RCA cut off, CABG with RCA grafting was done.

Conclusion: Single coronary artery is a rare congenital anomaly which can have a benign or malignant clinical presentation, depending on the presence of luminal narrowing at the single coronary ostium or compression along its route. CCTA and MRI are the modalities of choice for diagnosis and risk stratification of single coronary artery. The presence of atherosclerotic disease in patients with coronary artery anomaly has a clinical implication, particularly when the decision is to be made between percutaneous coronary intervention (PCI) and coronary artery bypass graft (CABG).

Keywords: Single coronary artery (SCA), Coronary computed tomographic angiography (CCTA), Myocardial infarction, Anomalous coronary artery, Coronary artery bypass grafting(CABG).

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I. Introduction

Single coronary artery (SCA) is a rare congenital anomaly. SCA can be either an isolated anomaly or associated with other congenital abnormalities like coronary artery fistula, bicuspid aortic valve, or tetralogy of Fallot. Most cases are asymptomatic and incidental findings, but SCA can cause ischemia, congestive heart failure, and sudden cardiac death (SCD). We describe a rare case of single coronary artery arising from right sinus of Valsalva with malignant course ,with no associated congenital heart disease presenting with myocardial infarction.

II. Case Details

A 45-year-old postmenopausal diabetic woman presented to cardiology department with acute inferior wall Myocardial infarction of more than 24 hours window period. 2D Echocardiography was suggestive of regional wall motion abnormality of right coronary artery (RCA) territory with mild LV systolic dysfunction (ejection fraction of 42%). Patient was initially treated with Anticoagulants, antiplatelets, high dose statin, antidiabetic medication and angiotensin converting enzyme (ACE) inhibitor. Coronary angiography was done which showed anomalous coronary arteries with single coronary artery arising from right coronary sinus dividing into left anterior descending artery (LAD), non-dominant left circumflex artery (LCX) and right coronary artery (RCA). Proximal RCA cut off was noted with retrograde filling from LAD and LCX. CT coronary angiography (CCTA) was performed using 128 slice volumetric CT. A single main coronary artery arising from right coronary sinus diving into three branches noted . Anterior branch (LAD) was coursing between aorta and right ventricular tract and then continued in the direction of normal course of LAD. The posterior branch (LCX) was coursing between root of aorta and left atrium and then continued as normal course

of LCX. The walls and lumen of LAD and LCX were normal. RCA was dominant with proximal third of lumen not visualised, while mid and distal segments were patent with normal calibre. PDA and PLV were patent with near normal calibre. Calcium score is zero (AJ 130) and is in the zeroth percentile for patient's age group. Our case with malignant course of artery belongs to Lipton's classification class III and to category IV of Rigatelli's classification. In view of anomalous single coronary artery having malignant course with LAD coursing between great arteries and RCA proximal cut off ,CABG (Coronary artery bypass grafting) with graft to RCA was done and patient was discharged in stable condition.

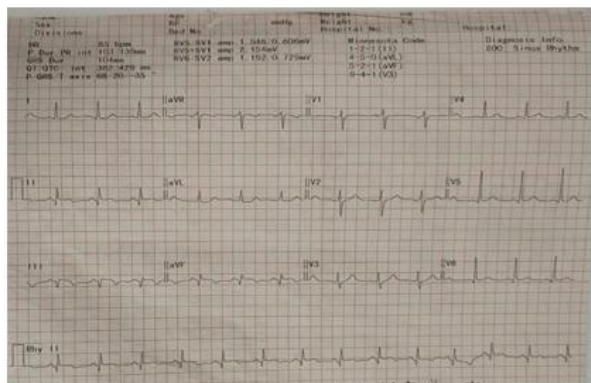


Fig.1 :ECG at presentation showing q with ST elevation in inferior leads

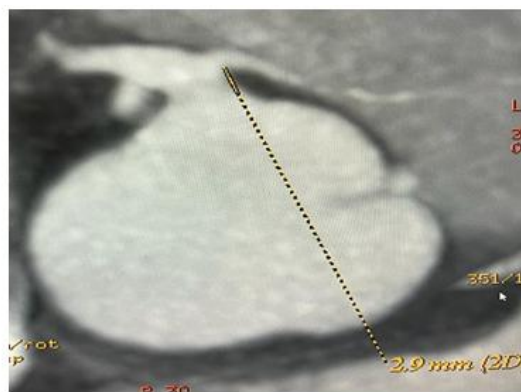


Fig.2 : CCTA image of single artery arising from right sinus of Valsalva and with normal ostial size

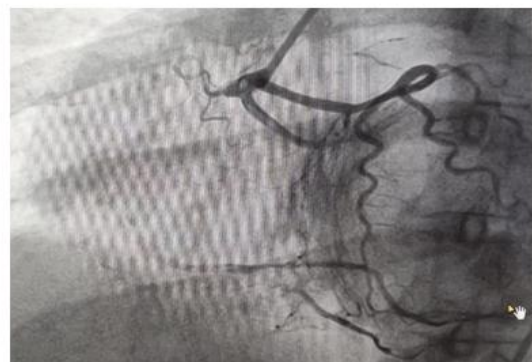
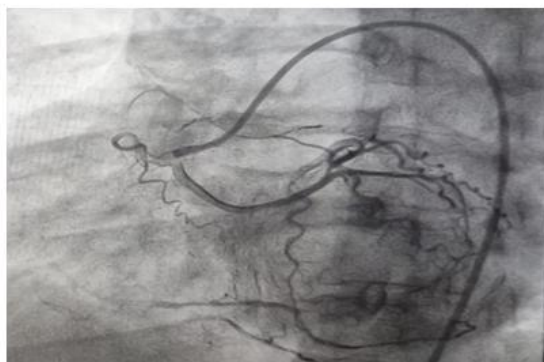


Fig.3 &4 : Coronary angiography showing single coronary artery arising from right sinus , normal LAD and LCX, Dominant RCA with proximal cut off ,filling from LAD and LCX.

Fig.5

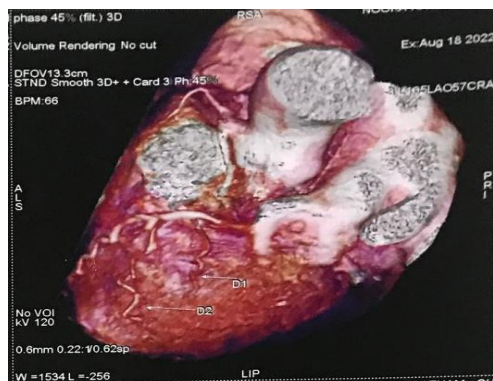
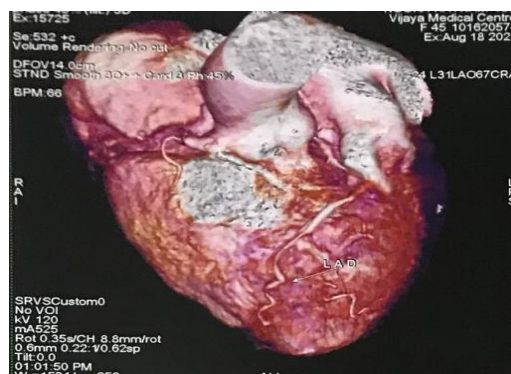


Fig.6



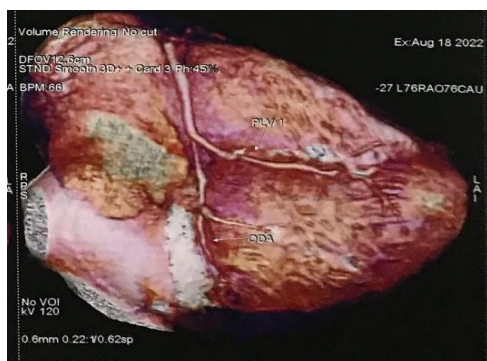


Fig.7

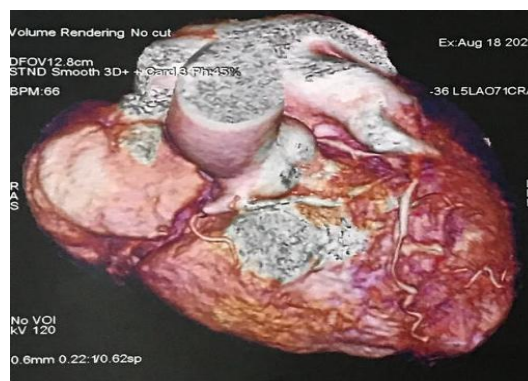


Fig.8

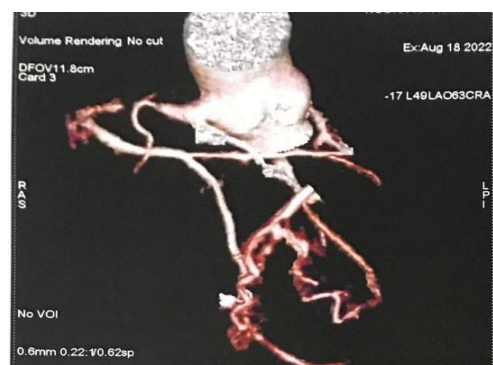


Fig.9

Fig.5-9 : CT Coronary angiography volume reconstructed images. Fig 5 showing single artery arising from Right sinus, LAD arising anteriorly and RCA with proximal filling defect. Fig 6:LAD course between aorta and pulmonary artery Fig 7&8 : Course of LAD arising anteriorly from SCA ,Fig 9 showing normal calibre PDA and PLV

III. Discussion

Single coronary artery is a congenital anomaly defined as a solitary coronary artery arising from a single coronary ostium in the aortic trunk and supplying the whole heart . There is no known gender predisposition.^{1,2} The incidence of single coronary artery(SCA)is rare, reportedbetween 0.024% and 0.098% of the population and are usually detected incidentally on coronary computed tomography angiography (CCTA).¹⁻⁵ SCA can either be isolated or coexist with other cardiac congenital anomalies including transposition of great vessels, coronary arteriovenous fistula , tetralogy of Fallot, truncus arteriosus, interventricular septal defect, patent ductus arteriosus, bicuspid aortic valve, or patent foramen ovale.⁶⁻⁸

In literatures, there are different classification systems of SCA based on conventional coronary angiography and necropsy findings. In 1979, Lipton et al. suggested a classification of single coronary artery in which two prior classification systems that were suggested by Smith(1950) and Ogden and Goodyer (1970) were incorporated.^{1,9}

As depicted in table 1and summarised in figure 10 , **Lipton’s classification of SCA**is summarised as :

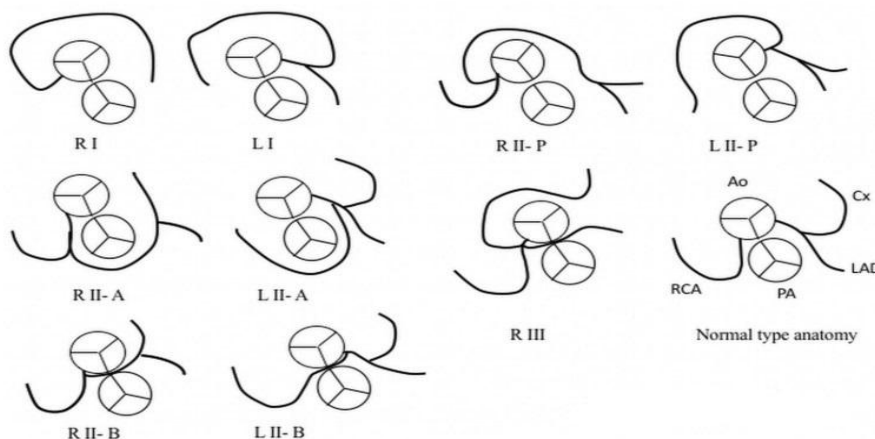
a)**Group 1**,the solitary vessel follows the course of either a normal right or left coronary artery.b)**Group 2**, single coronary artery arises from the right or left coronary sinus and from this a large trunk crosses the base of the heart to arrive at the normal contralateral coronary artery. This is further subdivided into six subgroups depending on the site of origin and route of the large transverse trunk.c)**Group 3** have a circumflex branch and anterior descending branch arising separately from a common trunk. The circumflex artery usually passes in a retro aortic route while the anterior descending branch courses between the aorta and pulmonary trunk to the intraventricular sulcus. SCA is divided into two main types: “R,” right type, and “L,” left type, to indicate the origin ofthe SCA from the right or the left coronary sinus, respectively. Further classification of these anomalies into malignant and benign forms depend on several factors, such as an interarterial course between the great vessels, proximal intramural course within the aorta, and ostial narrowing at the origin of the artery.¹

Rigatelli et al. proposed a new classification system based on the clinical significance of the single coronary anomaly and categorized SCA into four different classes. Class I is considered a benign type, class II is associated with fixed myocardial ischemia, class III is related to sudden cardiac death, and class IV is associated with superimposed coronary artery disease ,as depicted in table 2 .¹⁰In our patient, there was a single coronary artery arising from right coronary sinus, with LAD and LCX having separate origins ,LAD coursing between

great arteries and RCA with proximal cut off .This type of SCA is categorised as Lipton’s class R-III and Rigatelli’s class IV with malignant course.

Most of the patients with SCA are asymptomatic or present with nonspecific symptoms; however, patients with some variants of SCA can present with typical angina , myocardial infarction, syncope, ventricular tachycardia, and sudden cardiac death.^{1,7} SCA with an interarterial anomalous artery has been strongly linked with sudden death and myocardial ischemia especially among young competitive athletes .⁷ This is more common among patients with anomalous coronary artery arising from the right coronary sinus and coursing to the left between the aorta and the main pulmonary artery, as in our case.¹¹

SCA can be diagnosed by different diagnostic modalities like coronary computed tomography angiography, conventional coronary angiography and cardiac MRI. Conventional coronary angiography is the gold standard for assessment of the coronary artery; however, it is an invasive procedure and has a risk of procedure related complications.¹² Moreover, even with multiple projections and angiographic views, delineation of the anatomy of the coronary arteries in complex cases can be difficult.¹³ On the contrary, coronary computed tomography angiography (CCTA) is a non-invasive diagnostic tool with high temporal and spatial resolution that has emerged as a gold standard for detection and characterization of coronary artery anomalies.¹⁴⁻¹⁵ Management of patients with SCA includes observation, medical treatment and percutaneous or surgical intervention.¹⁶ In majority of asymptomatic patients with no atherosclerotic disease, usually, no invasive intervention is recommended.¹⁸ Invasive interventions including PCI with stent placement and surgical intervention are preserved for symptomatic patients and patients with a malignant course of the anomalous artery.¹⁹⁻²⁰ Surgical interventions include reimplantation of the anomalous artery to the aorta, osteoplasty, coronary artery bypass grafting (CABG) of the anomalous artery, and pulmonary artery translocation.¹⁶ In view of documented ischemia with anomalous SCA with proximal RCA cut off ,CABG was performed in our case with graft to RCA and discharged in stable condition.



Schematic diagram representing Lipton's classification of single coronary artery.

Fig.10

	Classification	Description
Ostial Location	R	Right Sinus of Valsalva
	L	Left Sinus of Valsalva
Anatomical Distribution	I	Solitary dominant vessel follows the course of either a normal Right or Left coronary artery
	II	One coronary artery arises from the proximal portion of the normally located other coronary artery
	III	LAD and Cx arise separately from a common trunk originating from the right sinus of Valsalva
Course of the transverse trunk	A	Anterior to the great vessels
	B	Between the aorta and pulmonary artery
	P	Posterior to the great vessels
	S	Septal type passes through the interventricular septum
	C	Combined type : a combination of diverse routes

Lipton's classification of single coronary artery.

Table 1: Lipton’s classification of single coronary artery

Table 2

Rigatelli's classification of coronary artery anomalies. Adapted from Rigatelli et al. ⁴	
Class	Coronary artery anomaly
I. Benign	Ectopic origin of LCx from right sinus Separate origin of LCx and LAD Ectopic origin of LCx from the RCA Dual LAD types I-IV Myocardial bridging (score <5)
II. Relevant Associated with myocardial ischemia	Coronary artery fistula Single coronary artery R-L, I-II-III, A-P Ectopic origin of LCA from PA Coronary artery atresia Hypoplastic coronary artery
III. Severe Related to SCD	Ectopic origin of LCA from the right sinus Ectopic origin of RCA from the left sinus Ectopic origin of RCA from PA Single coronary artery R-L, I-II-III B Myocardial bridging (score >5)
IV. Critical Associated with SCD/myocardial ischemia and with superimposed CAD	Class II and superimposed CAD Class III and superimposed CAD

Table 2: Rigatelli's classification of coronary artery anomalies

IV. Conclusion

We present a rare case of Single coronary artery (SCA) originating from ostium in right sinus of Valsalva with malignant course, documented ischemia and proximal cut off on coronary angiography. Given the vast variety of anatomical courses of SCA, the multidisciplinary approach involving interventional cardiologists and cardiothoracic surgeons should be considered to decide on the best treatment options. Awareness of these types is essential as it has an implication for planning patient management.

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