

## Case Report

### Case Report: Trilogy of Anomalies

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#### Abstract

Congenital anomalies is interesting in the sense that many patients can manifest with symptoms in childhood while others may remain asymptomatic or with unusual symptoms and anomalies are diagnosed accidentally in adulthood. We are sharing this case of 66 year old female who presented to us symptoms suggestive of cardiac ischemia and three anomalies which might be the contributing factor in this patient. Anomalous right coronary artery originating from the left anterior descending artery, left anterior descending artery intra myocardial bridging and a fistulous communication of the septal perforators with the left ventricle.

**Methods and Materials:** Right radial area was sterilized with pyodine liquid and draped. The subcutaneous tissue was numbed with 2 ml of 2% lidocaine injection. The right radial artery vascular access was taken via 5 Fr sheath. Guide catheters JL 3.5 and JR 4.0 used to engage the left main and right coronary artery respectively. This case has been reviewed and approved by the IRB (Institution review board) of Bahria International Hospital, Lahore. The literature for discussion was reviewed from various articles from Pub Med and internet search engines before submission.

**Conclusions :** The anomalous origin of the right coronary artery from the left anterior descending artery is a unusual coronary anomaly. Congenital anomalies do occur very rarely but what is important is to stratify whether they can pose serious cardiac effects and appropriate measures to be taken beforehand.

**Received** | 03-07-2018: **Accepted** | 27-06-2019

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**Keywords** | left anterior descending artery (LAD), right coronary artery (RCA), intra myocardial bridging, fistulous communication

#### Introduction

Congenital coronary artery anomalies exist by birth but most remain asymptomatic to be incidentally discovered during angiography for another cause. The prevalence is about 1.3%.<sup>1,2</sup> Congenital anomalies account for 0.3 to 5.6% of the cases of coronary angiography and in 0.3% of autopsies<sup>3</sup>. Coronary Anomalies can be classified into benign and malignant; which carry a serious prognosis. The benign anomalies include Left Circumflex artery (LCx) from Right sinus or Right coronary artery

(RCA) itself and separate ostial origins of LCx and Left Anterior descending artery (LAD). The Malignant anomalies include Left main artery (LM) or LAD form Right sinus, RCA from LM, Anomalous left coronary artery from the pulmonary artery (ALCAPA), Anomalous right coronary artery from the pulmonary artery (ARCAPA) and Coronary Artery Fistula. The origin of an anomalous coronary artery is a relatively rare occurrence with an incidence of is about 1% in the general population<sup>1, 4</sup>. An anomalous RCA which originating from the LAD is seldom encountered and has been considered a single

coronary artery variant<sup>[4]</sup>.

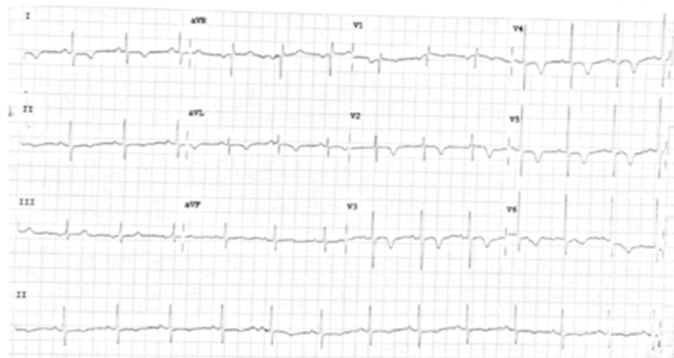
Malignant features of an anomalous coronary artery constitute four features; a slit like ostium, acute angle of takeoff, compression between the Aorta and the Pulmonary Artery and an intramural course.

We herein report a case of an Anomalous RCA arising from the LAD, in addition to this rare anomaly there were two coexistent other anomalies; LAD intra myocardial bridging and a fistulous communication of the septal perforators with the Left ventricle.

### Case Report

We account a case of a 66 year old female, with prior comorbidities including hypertension, dyslipidemia, HCV (Hepatitis C) positive chronic liver disease and long standing dyspepsia. She reported to us in the clinic with history of exertional chest heaviness and epigastric discomfort that she often confused with worsening of dyspepsia. Her symptoms had gotten worse over the past 2 months and was having functional class II-III symptoms. She had a strong family history of coronary artery disease; father and brother both suffered myocardial infarction in their 50s and ending up with coronary artery bypass grafting CABG (father) and a percutaneous coronary intervention to the left anterior descending artery (brother).

EKG performed showed T wave inversions in V2-V6 and also in lead I and aVL.



**Figure 1:** EKG showing T wave Inversions in V2-V6, I and aVL

Echocardiography performed revealed a preserved Left Ventricular systolic function with no regional wall motion abnormalities and no hemodynamically significant valvular pathology.

Keeping in mind her strong family history and the presence of worrying T wave changes, and detailed discussion with the patient which included options of all diagnostic modalities, decision to proceed with a coronary Angiography was made.

Patient underwent coronary Angiography via the radial approach. On engaging the Left Main artery (LM), we found that the Right Coronary Artery (RCA) was originating from the Proximal left anterior descending artery (LAD), this was the first anomaly discovered in this patient.

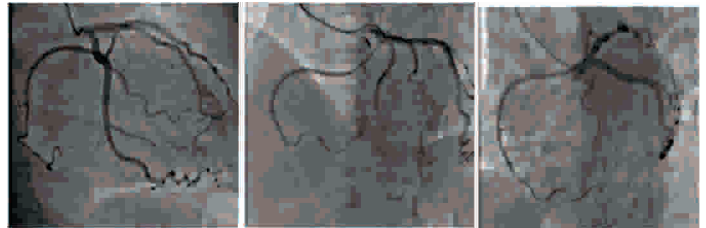


Fig. 2a

Fig. 2b

Fig. 2c

**Figure 2:** RAO (Right Anterior Oblique), Caudal (2a) LAO (Left Anterior Oblique) Cranial (2b) and LAO Caudal (2c) views Respectively Demonstrating RCA Originating from Proximal LAD.

On Further imaging it was also found that there was significant Myocardial Bridging of the LAD, the second anomaly. You can see in Figure 3(A) and 3(B) frame in diastole and systole showing compression of mid LAD in systole.

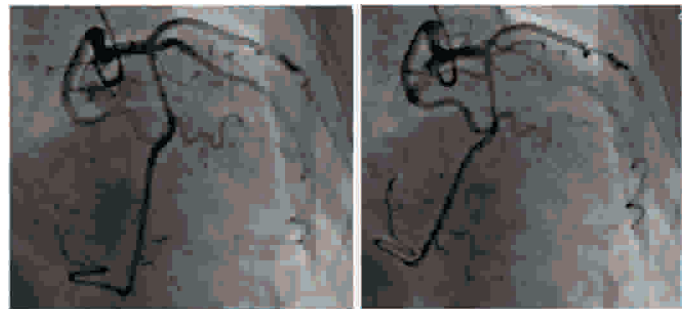


Fig. 3a

Fig. 3b

**Figure 3:** RAO Cranial view Showing LAD in Diastole (3a) and Systole (3b) Respectively with Significant Myocardial Bridging of the Mid LAD

It was also noted that the Left Ventricle (LV) was filling via a fistulous communication from the septal perforators, the third anomaly.



**Figure 4:** RAO Caudal view Demonstrating Septal Perforator Filling the Left Ventricle.

The patient was already on maximum tolerated dose of beta blockers and nitrates with BP of 100/ 65 mm Hg and heart rate of 78/min. It was, therefore, decided to add Ivabradine to existing therapy. The patient was advised an Exercise MIBI to assess the ischemic burden on the myocardium and a CT angiography to outline the course of the anomalous RCA. On telephonic contact she was feeling better and so far has decided not to pursue this further with more investigations.

## Discussion

The aberrant variant origin of the RCA from the LAD is rare, with only 40 occurrences being reported as solitary cases in published literature thus far.<sup>5,6</sup>

Previously a diverse array of differing origins of the RCA have been described; the left anterior sinus with varying courses, ascending aorta above the level of the sinus, below the aortic valve, descending thoracic aorta, the pulmonary arteries, left main coronary artery, or circumflex coronary artery.

Review of previous data on anomalous RCA arising from the LAD, describe an origin following the first septal perforator.<sup>7</sup> It carries a benign prognosis mostly, however, it may become clinical significant if it courses between the aorta and pulmonary artery, leading to ischemia of the myocardium or even sudden cardiac death.<sup>8</sup>

Usually, the anomalous vessel has been found arising from the proximal or mid segment of the LAD<sup>8,9</sup> Two courses of anomalous RCA have been well described as anterior to the pulmonary artery trunk or retro

aortic<sup>18,10</sup>. Majority of the published reports have described the anterior course of the aberrant RCA to the pulmonary artery trunk except for only two cases reporting a retro-aortic course.<sup>11,12</sup> The malignant course of RCA originating from the left main coronary artery, and coursing inter-arterially still remains to be reported.<sup>1,13</sup>

Different coronary anomalies can cause varying degrees of ischemia, most anomalies including split RCA and RCA originating from the left or Right cusp cause no ischemia. Intermittent ischemia can usually be encountered in patients with anomalous coronary arising from opposite cusp, coronary artery fistulas and in myocardial bridging. Obligatory ischemia is found in Anomalous left coronary artery from the pulmonary artery (ALCAPA), ostial atresia and severe stenosis.

The mechanism of limitation to coronary flow varying in severity from an ischemic pattern of angina with exertion to sudden cardiac Death, can be linked to the malignant features of anomalous artery origin i.e. including, slit-like orifice, acute take off angle and compression of the intramural segment by the aortic valve commissure. Lateral compression of coronary lumen in its intramural portion and compression of the coronary between pulmonary artery and aorta can be postulated as other mechanisms for induction of ischemia.<sup>14</sup> Post Mortem studies indicate that acute angle takeoff and narrow slit like orifice are most common findings in a sudden cardiac death patient.<sup>14</sup> Controversy remains over the mechanism causing the interarterial course compression between the aorta and pulmonary artery, an intravascular ultrasound study confirmed that this was caused most certainly due to the high aortic pressure as the pulmonary artery pressure was expectedly lower than the aorta.<sup>15</sup>

Coronaries and their major branches mostly course in sub-epicardial tissue of the myocardial surface. Occasionally, an epicardial segment of courses below overlying superficial myocardial fibers known as myocardial bridging. With a frequency estimated at 0.5-2.5 % in coronary angiography studies, usually involving the LAD artery.<sup>16</sup> Commonly, it is not clinically significant may lead to angina pectoris, coronary spasm, arrhythmias, myocardial infarction, and sudden death.<sup>16</sup>

In our case the exertional Ischemia of the patient was likely secondary to the LAD intra myocardial bridging rather than the RCA anomaly, the LV fistulous communication though present was not significant to warrant need of surgery. Patient was managed medically, by maximizing beta blocker dose to reduce the heart rate and decrease myocardial ischemia burden, nitrates for preload and calcium channel blockers for afterload reduction respectively. Ivabradine was also added as an adjunct to further reduce the heart rate.

## Conclusion

The anomalous origin of the RCA from the LAD remains a rare congenital coronary aberrancy in which the abnormally arising RCA originates from either the proximal or mid of the LAD. It commonly remains a benign coronary variation with certain features which can cause rare but malignant prognosis.

## References

- Bhatt A, Foster E, Kuehl K, Alpert J, Brabeck S, Crumb S et al. Congenital Heart Disease in the Older Adult. *Circulation*. 2015;131(21):1884-1931.
- Villa A, Sammut E, Nair A, Rajani R, Bonamini R, Chiribiri A. Coronary artery anomalies overview: The normal and the abnormal. *World J Radiol*. 2016;8(6):537.
- Motta P, Santoro J. *Congenital Heart Disease in Pediatric and Adult Patients*. Cham: Springer; 2017.
- Yurtdas M, Gülen O. Anomalous origin of the right coronary artery from the left anterior descending artery: Review of the literature. *Cardiology Journal*. 2012; 19(2):122-129.
- Tahir H, Ahmad S, Awan M, Omar B, Glass J, Cole J. Anomalous Origin of Left Anterior Descending Artery and Left Circumflex Artery from Right Coronary Sinus with Malignant Left Anterior Descending Artery Course: Role of Coronary CT Angiography Derived Fractional Flow Reserve in Decision Making. *Cureus*. 2018;
- Balghith M. Anomalous origin of the right coronary artery from the proximal left anterior descending artery and a single coronary artery anomaly: Three case reports. *J Saudi Heart Assoc*. 2013;25(1):43-46.
- Gungor M, Yıldırım E, Bugan B. Anomalous origin of the right coronary artery from the left anterior descending artery: A rare variant of single coronary artery. *International Journal of the Cardiovascular Academy*. 2016;2(3):137-139.
- Parviz Y, Rowe R, Rinze R. Emergency intervention in a case of an anomalous origin of the right coronary artery from the left anterior descending artery. *Am J Emerg Med*. 2017;35(6):926-928.
- Gholoobi A. Anomalous Origin of Right Coronary Artery from Distal Left Circumflex Artery: A Very Rare Variant of Single Coronary Artery Anomaly. *Int Cardio Res J*. 2016;10(3).
- Çanga Y, Güvenç T, Karataş M, Çalık A, Onuk T, Tanik V et al. Congenital coronary artery anomalies in adults: review of 111 cases from a single-centre experience. *Cardiol Young*. 2016;27(6):1041-1050.
- Suryanarayana P, Lee J, Abidov A, Lotun K. Anomalous right coronary artery: case series and review of literature. *Cardiovasc Revasc Med*. 2015;16(6):362-366.
- Meric M, Demircan S, Pinar M, Yuksel S. Single coronary artery: right coronary artery originated from middle of left anterior descending artery in a patient with severe mitral regurgitation. *Clin Cardiol*. 2010; 33:E48–50
- Yurtdaş M, Gülen O. Anomalous origin of the right coronary artery from the left anterior descending artery: review of the literature. *Cardiol J*. 2012; 19: 122–129
- Stout K, Daniels C, Aboulhosn J, Bozkurt B, Broberg C, Colman J et al. 2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease: Executive Summary. *Journal of the American College of Cardiology*. 2019;73(12):1494-1563.
- Visual Guide to Neonatal Cardiology [Internet]. Google Books. 2019 [cited 21 May 2019]. Available from: <https://books.google.com.pk/books?hl=en&lr=&id=1dtJDwAAQBAJ&oi=fnd&pg=PA233&ots=wlpvzq4Ka8&sig=WzWH5cR1MjEJl0re9EJLHM BID>
- Tarantini G, Migliore F, Cademartiri F, Fraccaro C, Iliceto S. Left Anterior Descending Artery Myocardial Bridging. *J Am Coll Cardiol*. 2016;68(25):2887-2899.