Case Report

Anomalous Origin of Left Main Coronary Artery from Right Coronary Artery in A Patient Presenting with Inferior Wall Myocardial Infarction: A Case Report

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Abstract:

Anomalous origin of the coronary arteries is a very rare phenomenon and is seen only in less than 1% of the general population. Single coronary artery (SCA) is a congenital anatomic abnormality identified by a single coronary ostium giving rise to one coronary artery. Our case presented at 40 years with intermittent chest discomfort, effort intolerance and a history of getting Streptokinase one month back due to AMI (Inferior). Diagnosis was confirmed with elective conventional coronary angiography and coronary CT angiography as an extremely rare variant of the left main coronary artery (LMCA) branching off from the right coronary artery (RCA) and then following a pre-pulmonic course. We did Off Pump CABG surgery with four grafts and discharged the patient uneventfully with guideline-directed medical therapy with a beta-blocker, statin, and dual antiplatelet agents and the patient is on follow up.

Key words: *Single coronary artery, Coronary artery anomaly, Pre-pulmonic, Anomalous left main coronary artery, CABG*

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Introduction:

The term 'coronary artery anomaly' (CAA) is used when the observed coronary pattern is seen in less than 1% of the general population.1 The overall incidence of CAA has been estimated between 0.9% and 5.6%.2–4 Based on the origin and course of the anomalous artery, CAA can either represent a benign incidental finding or can have severe cardiovascular sequelae. Coronary computed tomography angiography (CTA) is a reliable non-invasive tool for diagnosing CAA. The management varies based on the nature of symptoms. Here, we present a case of inferior wall ST-elevation myocardial infarction due to distal right coronary artery (RCA) occlusion with incidental findings of anomalous left main coronary artery (LMCA) branching off the RCA.

Case

A 40-year-old hypertensive and diabetic lady presented to our facility for elective CAG with a h/o AMI (Inferior) approximately one and half months back. Other relevant medical history included effort fatiguability and occasional

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left sided chest pain for two months with a positive family history. On presentation, the patient was haemodynamically stable and physical examination including cardiovascular auscultation was unremarkable. Electrocardiograms (EKG) displayed Q-wave and inverted T-wave in the leads II, III, and AVF (Figure 1A). The patient was admitted under Cardiology department and transferred to the cardiac catheterization lab where she underwent coronary angiogram. After an initial failed attempt to first cannulate the left coronary ostium to evaluate for concomitant left anterior descending (LAD) and circumflex disease with the suspected culprit being the RCA based on EKG, the right coronary ostium was engaged instead, which revealed a large caliber RCA descending through the coronary sulcus to the crux, giving rise to left anterior descending and obtuse marginal branches before giving rise to an early posterior descending artery (Early PDA) and continued to a postero-lateral vessel (PLV). Contrast injection into the RCA also showed the anomalous origin of the LMCA stemming from the proximal segment of the RCA. RCA was dominant, good size vessel having 98-99 stenosis at its proximal segment and 70-80 stenosis at its distal segment. The aberrant LMCA reached the left side of the heart anterior to the pulmonary artery and trifurcated into a small caliber LAD, ramus intermedius (RI), and left circumflex (LCX) artery that ends into a bifurcated obtuse marginal system (Figure 2). Patient was then referred back to Cardiac Surgery department for CABG operation. After proper pre operative evaluation among which Transthoracic echocardiogram (TTE) showed an ejection fraction (EF) between 45% and 50%, basal segment of inferior and inferolateral wall akinesis and no valvular abnormalities (Figure 1B). A coronary CTA confirmed the findings of coronary angiogram and a potential malignant course of anomalous LMCA was ruled out (Figure 3). Off-Pump Coronary Artery Bypass Surgery with four grafts (OPCAB×04 grafts) done with Left Internal Mammary Artery (LIMA) to LAD and Reverse Saphenous Vein Graft (RSVG) to OM, Early PDA & Poster-lateral Vessel (PLV) (Figure 4). Her immediate post-op and further hospital stay remained uneventful, and the patient was discharged home on guideline-directed medical therapy with a beta-blocker, statin, and dual antiplatelet agents. At 1-month follow-up visit, the patient remained asymptomatic and a repeat TTE showed no new findings.

Course of management:

Day	Timing	Events
Day 1	17.08.2020	Patient presents with chest pain in a peripheral hospital. Diagnosed as having AMI (Inferior). Treated with Inj. Streptokinase and LMWH. Started on dual antiplatelet, statin therapy and released with advice for early CAG.
Day 43	30.09.2020	Elective CAG done at our center. Which revealed 98-99% stenosis at the proximal segment of LAD, 70-80% stenosis at the proximal segment of LCX, 98-99% stenosis at the proximal segment of RCA and 70 -80% stenosis at its distal segment right coronary artery (RCA) lesion and anomalous origin of left main coronary artery (LMCA) from RCA. Transthoracic echocardiography is performed which shows mild to moderately reduced ejection fraction.
Day 44	01.10.2020	Patient is scheduled for coronary computed tomography (CT) angiography and Evaluation for CABG. Coronary CT angiography (on 21.10.2020) demonstrates benign prepulmonic course of aberrant LMCA.
Day 74	01.11.2020	Off Pump CABG Surgery with 04 Grafts done.
Day 83	10.11.2020	Patient discharged on aspirin, clopidogrel, atorvastatin, metoprolol and furosemide with advice for follow up.

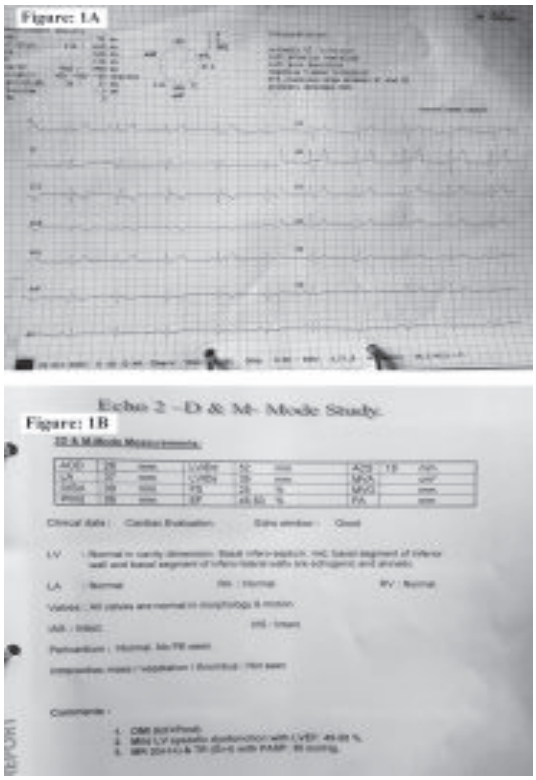

Fig.-1: *(A) Electrocardiograms (EKG) displayed Q-wave and inverted T-wave in the leads II, III, and AVF (B) Transthoracic echocardiogram (TTE) showed an ejection fraction (EF) between 45% and 50%, basal segment of inferior and inferolateral wall akinesis and no valvular abnormalities.*

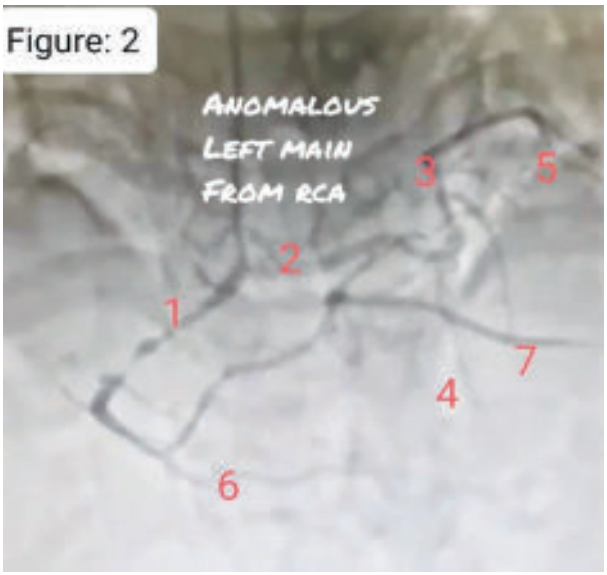

Fig.-2: *Contrast injection into the RCA showed the anomalous origin of the LMCA stemming from the proximal segment of the RCA. RCA was dominant, good size vessel having 98-99 stenosis at its proximal segment and 70-80 stenosis at its distal segment. The aberrant LMCA reached the left side of the heart anterior to the pulmonary artery and trifurcated into a small caliber LAD, ramus intermedius (RI), and left circumflex (LCX) artery that ends into a bifurcated obtuse marginal system. 1, right coronary artery; 2, left main coronary artery; 3, left circumflex artery; 4, left anterior descending artery; 5, obtuse marginals; 6, early posterior descending artery; 7, postero lateral vessel*

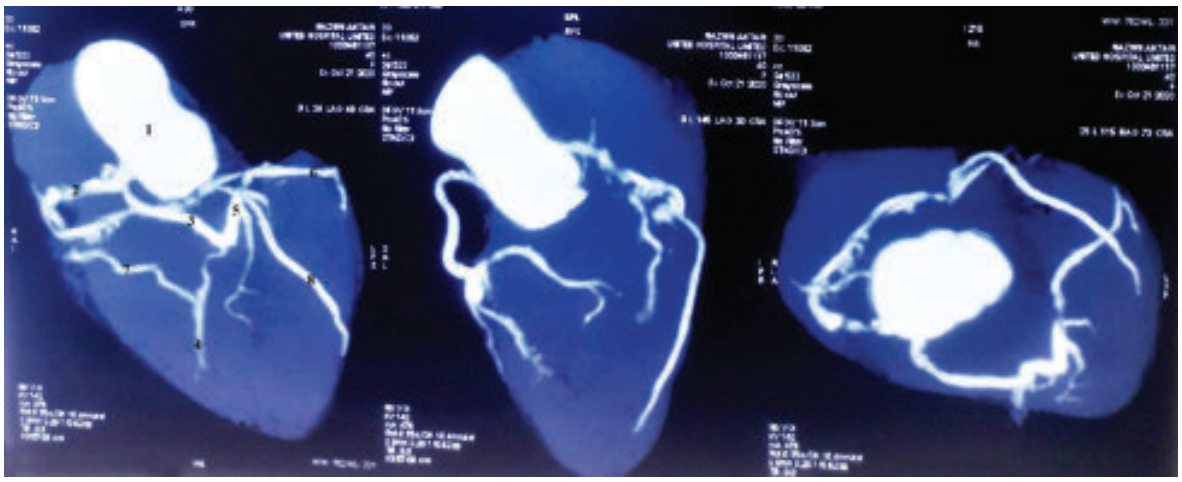

Fig.-3: *Computed tomography image of the heart (left) showing the right coronary artery giving rise to the left main coronary artery and the latter's course anterior to the main pulmonary trunk. 1, ascending aorta; 2, right coronary artery; 3, left main coronary artery; 4, left anterior descending artery; 5, left circumflex artery; 6, obtuse marginal branches; 7, early posterior descending artery; 8, postero-lateral vessel.*

Table-I

Lipton's classification for single coronary artery

Ostia location	Anatomic distribution subtypes
Right sinus of Valsalva (R)	
	RI—SCA follows the course of a normal RCA
	RII—SCA from the right sinus gives off an anomalous transverse branch that crosses the base to reach the contralateral side
	RIII—SCA from the right sinus, with LAD and LCx separate coronary trunks instead of single trunk
Left sinus of Valsalva (L)	
	LI—SCA follows the course of a normal LMCA
	LII—SCA from the left sinus gives off an anomalous transverse branch that crosses the base to reach al side

LAD, left anterior descending artery; LCx, left circumflex artery; LMCA, left main coronary artery; RCA, right coronary artery

Table-II

Subtypes of type II single coronary artery based on the course of the aberrant vessel

Anatomic distribution	SCA subtypes based on course of anomalous transverse branch
RII or LII Type A	—courses anterior to the pulmonary trunk
Type B	—courses between pulmonary artery and aorta
Type P	—courses posterior to the aorta
Type S	—septal type courses above the interventricular septum
Type C	—combined

Discussion:

Single coronary artery anomaly (SCA) is a congenital anatomic abnormality identified by a single coronary ostium giving rise to all arteries supplying the heart. Single CAA is uncommon and seen in only 0.024–0.06% of cases.5 In 1979, Lipton et al.6 provided the angiographic classification of SCA based on the site of ostial location, the anatomical course of the single vessel and relationship of the aberrant transverse branch with respect to the great arteries of the heart. This was further modified by Yamanaka et al.3 in 1990. Single CAA can either be right (R) or left (L) sided corresponding to the ostial location in the right or left sinus of Valsalva. Single CAA are classified into three groups from I to III with Group II consisting of various subtypes based on the course of the aberrant vessel (Tables 1 and 2). Villa et al.7 in 2016 classified CAA functionally as (i) anomalies with obligatory ischaemia, (ii) anomalies without ischaemia, and (iii) anomalies with episodic ischaemia that occasionally cause severe events but are otherwise compatible with normal life. In our case, the patient had congenital SCA; however, her acute presentation was unrelated to the aberrant vessel pathology. Based on the Lipton anatomic classification, the patient illustrated SCA type RIIA with the prepulmonic course of the aberrant vessel (LMCA). Potentially LMCA, RCA, and LAD can all branch from the SCA and take an aberrant course. Type II SCA involving the LMCA is fairly common and is seen in 5% of patients with tetralogy of

Fallot which can often complicate valve repair. As the vessel crosses the right ventricular outflow tract anteriorly, there are no significant haemodynamic consequences, however, isolated cases of angina have been reported.8 The aberrant vessel can also take an interarterial,

retroaortic, or septal (sub-pulmonic) route before reaching the left side of the heart. The five potential paths of the aberrant vessel before reaching the perfusion territory, correlate directly with the risk of SCA. Inter-arterial branch coursing between the pulmonary trunk and aorta has an aberrant intramural course within the aortic wall resulting in hypoplasia and lateral compression.9 Inter-arterial course can lead to sudden cardiac death (SCD). The mortality rates of left interarterial arteries (LAD or LMCA) are higher (57%) compared to right interarterial arteries such as RCA (25%). In the retroaortic subtype, the aberrant branch courses between the posterior aorta and the interatrial septum which can often complicate aortic valve surgery. This anomaly

usually arises from RCA and is not haemodynamically significant. In the subpulmonic type of SCA, the aberrant branch travels anteriorly and inferiorly through the interventricular septum before coursing through septal myocardium. The subpulmonic type has a lower position, is surrounded by septal myocardium and does not have slit like orifice differentiating it from interarterial course. When diagnosing suspected CAA, it is imperative that an exact anatomical course is established to assess the risk of SCD. The role of TTE in diagnosing CAA is limited. Inclusion of two new screening views by Thankavel et al.10 improved the diagnostic ability of TTE in anomalous coronary artery from opposite sinus from 0.02% to 0.22%. Currently, coronary CTA and magnetic resonance angiography are Class I indications for diagnosing congenital CAA11 Cardiac

CTA allows for improved, non-invasive visualization of CAA albeit at the expense of radiation and contrast exposure. Superiority of coronary CTA was depicted by Shi et al.12 in a report that showed conventional angiography was diagnostic in only 53% of CTA proven CAA cases. Magnetic resonance angiography is an alternative that can simultaneously assess structural abnormalities without requiring contrast or harmful radiation exposure however the use remains limited. Coronary angiography remains a useful test to diagnose and classify CAA and is the gold standard for identifying associated coronary artery disease. Traditional angiography offers limited visualization of the coronary ostia, proximal course, and surrounding structures. Ali et al.13 recommended looking for the presence of two signs that should raise suspicion for coronary anomaly (i) the unperfused myocardium sign where the vessel supplying that myocardial territory is not visualized. (ii) The 'aortic root' sign where the vessel appears to cross the aorta and the pulmonary artery at the level of aortic root. In cases with difficult LMCA cannulation, a cuspogram can be performed to visualize the partly opacified artery. When the cuspogram is unsuccessful, right, and non-coronary sinus should be engaged to look for the anomalous origin of LMCA. The 'dot and eye method' was tested by Ishikawa et al.14 to determine the true course of CAA radiologically. The role of non-invasive functional testing to assess the haemodynamic effect of SCA on myocardial perfusion remains controversial.

Both exercise treadmill and stress myocardial perfusion scan can yield false-positives and false-negative results. The use of fractional flow reserve (FFR) and intravascular ultrasound (IVUS) to guide therapy is increasingly gaining attention. Fractional flow reserve can help assess the haemodynamic flow of the aberrant vessel and IVUS can provide an insight on the ostial anatomy. Intravascular ultrasound can help identify slit like or stenosed orifice, acute angle takeoff, and intramural aortic segment (coursing through aortic wall) all of which can increase risk of SCD. Together FFR and IVUS can helps establish the need for intervention especially when the presenting symptoms are atypical.15

Treatment options include observation with conservative medical management, percutaneous intervention, or surgical repair. Proximal stenosis in cases with SCA can be devastating if adequate collaterals do not exist. The 2008 guidelines for adults with congenital heart disease recommend revascularization or surgical repair of interarterial course regardless of symptoms due to higher rates of myocardial infarction and surgical revascularization on follow-up11 Aberrant vessels with prepulmonic, retroaortic, or transseptal courses have excellent prognosis. Asymptomatic patients with high risk anatomic features and absence of ischaemia on stress testing need multidisciplinary approach.

Conclusion:

We report a rare case of an incidentally identified SCA with Lipton type RIIA pattern. Although the majority of the patients are asymptomatic further evaluation with coronary CTA is warranted to rule out potential malignant pathology. The risk of SCD is highest with the interarterial course of the aberrant vessel. Stress testing is often not reliable in assessing the functional status of the patient. The optimal approach involves risk stratification, multidisciplinary management, and surgical intervention in appropriate patients.

Conflict of interest: None

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