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Anomalous Right Coronary Artery with LV Non-compaction having Malignant Course, A Rare but Lethal Combination

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ABSTRACT

Anomalous Right coronary Artery origin from contralateral sinus is a rare but lethal variant that may mimic like acute coronary syndrome and is a potential cause of sudden cardiac death. This needs early diagnosis with prompt treatment which may be conservative or interventional depending upon the clinical presentation, exact anatomy and route of the vessel. Here we present a unique case of combination of anomalous right coronary artery origin from opposite sinus causing ischemia, with LV non-compaction ending in non-hemorrhagic stroke.

Keywords: Anomalous coronary artery, inter-arterial course, computed angiography.

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Introduction:

Coronary artery anomalies are very rare disorders, accounting for only 1.3% of the patients, undergoing diagnostic investigations. In Yamanaka's report, RCA arising from the left coronary cusp was noted in 0.17% of coronary angiographies, while LCA from the right coronary cusp was seen in 0.047%.⁰¹

We report a case of aberrant origin of right coronary artery (RCA) having a malignant course causing ischemia, LV showing apical non-compaction as well and later on developed ischemic stroke. This course of the disease, to best of our knowledge, has never been reported in literature before.

Case Report:

A 57-years-man, diabetic & hypertensive, presented with chest pain of 4 days duration. The pain was of ischemic in nature. Electrocardiogram (ECG) showed ST-T changes in inferior leads. He was managed on the lines of Acute Coronary Syndrome (ACS) and coronary angiography (CA) was planned. The Left main (LM) was normal with mild irregularities in left anterior descending artery (LAD) and normal non-dominant Left circumflex (LCX). Surprisingly the injection in left system showed an aberrant vessel originating from the left cusp which was Anomalous Right coronary artery (ARCA). This was than selectively engaged with the help of XB guide and a BMW wire showing a mega dominant RCA with moderate ostial stenosis. (Figure 1)

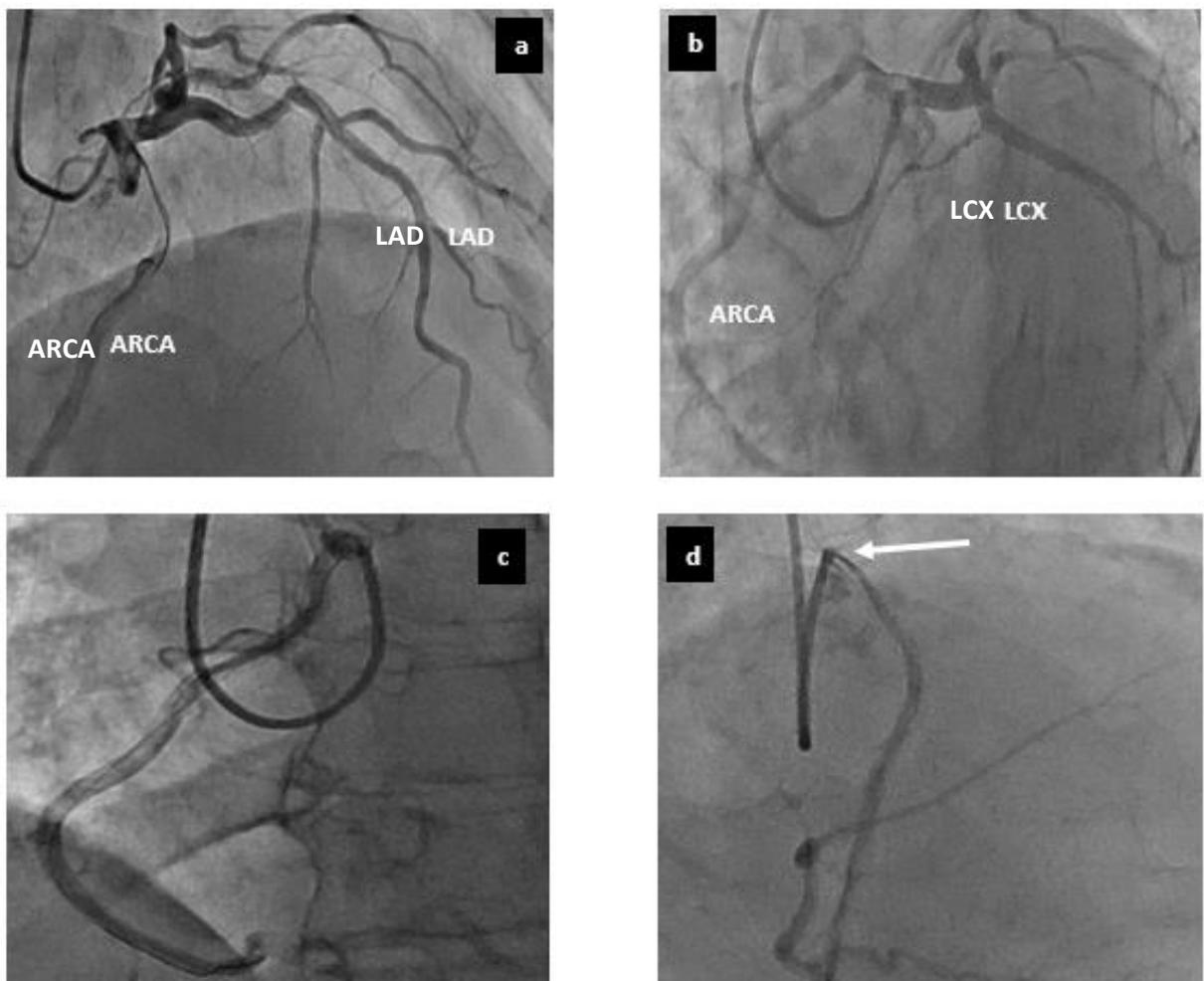


Figure 1: Coronary Angiography. 1a (RAO cranial view) and 1b (LAO caudal view), LM injection showing LAD and LCX with ARCA arising from Left coronary cusp (LCC), 1c (LAO view) showing selective engagement of Anomalous right coronary artery arising from LCC engaged with XB guide, 1d (RAO view) showing moderate ostial stenosis (Arrow).

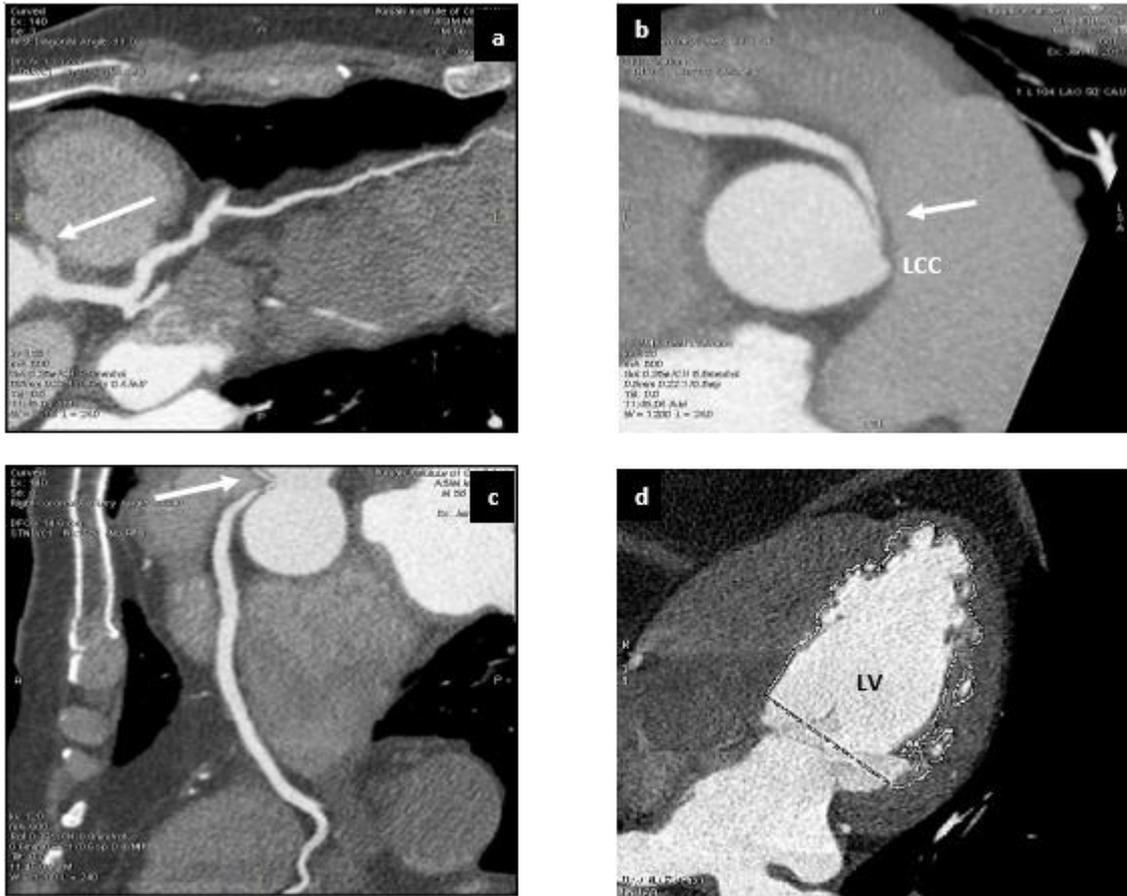


Figure 2: Multi-detector Computed tomography angiography. 2a, 2b and 2c showing aberrant origin of RCA from LCC (white arrows), 2d showing non compaction of LV.

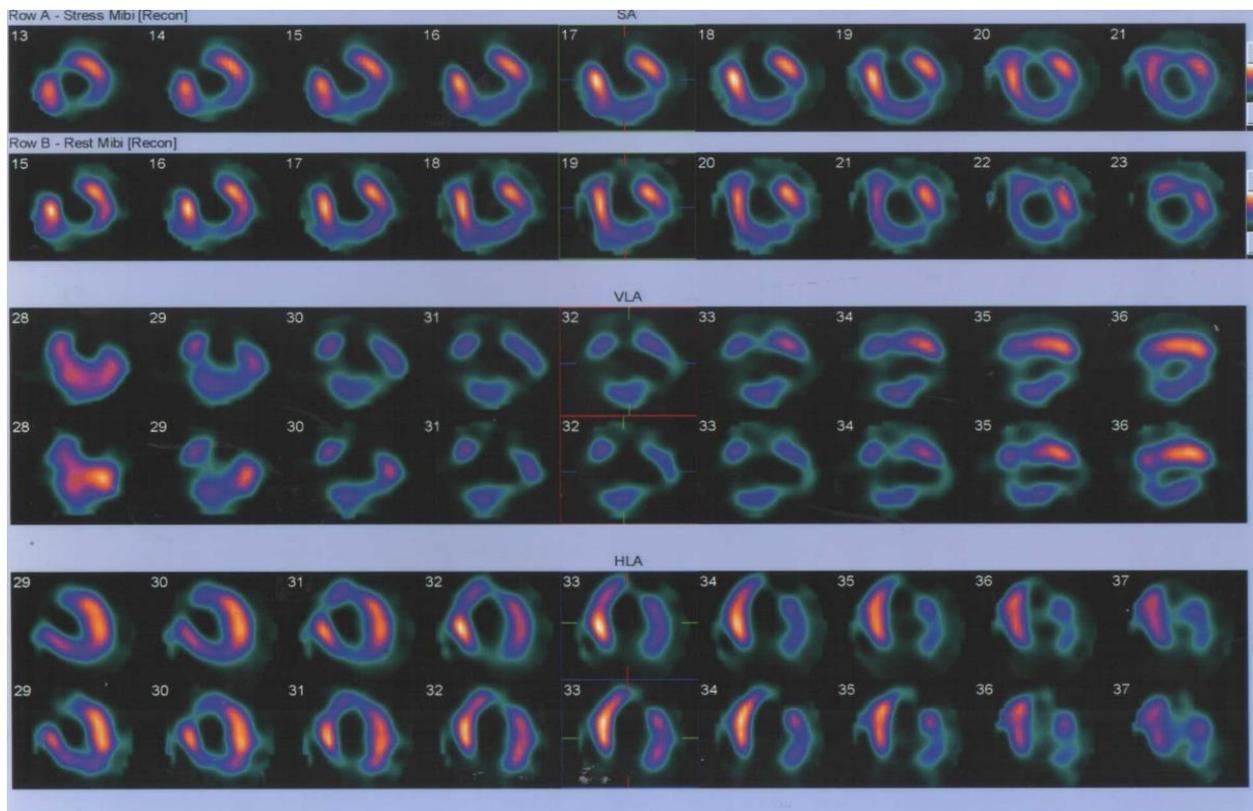


Figure 3: Stress test showing ischemic changes in inferior, lateral, apical and mid anterior wall of Left ventricle.

Multi-detector Computed tomography angiography (MDCT) was done, than, to rule out the inter-arterial course of the vessel, which confirmed it (Figure 2a, b and c). The Left ventricular (LV) function was depressed, about ejection fraction (EF) of 39% showing mid apical non-compaction (Figure 2d).

He was referred to cardiac surgeon for surgery who advised stress test. Stress test (Figure 3) showed dilated LV with segmental myocardial infarctions (MI) involving apex, apical ½ of inferior wall, apical septum, mid anterior wall and apical lateral wall. Partial thickness MI of basal inferior wall, infero-lateral part, basal and apical segments of anterior wall and reduced EF.

He was, than planned to undergo coronary artery bypass (CABG), as evidence of ischemia was there and the patient was still complaining of ischemic pain. His bilateral Carotid Doppler was normal. The patient, while awaiting for surgery, developed non-hemorrhagic stroke and right sided body weakness. His immediate CT scan of brain was unremarkable but after 48 hours, it showed a large infarct with hemorrhagic transformation within it. He needed ventilator support, was shifted back to ward after a week, followed by discharge in a stable condition.

DISCUSSION:

Anomalous coronary artery from the opposite sinus is a very rare condition reported to be 1.3% by Yamanaka's report ⁰¹ and 1.07% in P. Angelini's group study respectively. ⁰²

This anomalous origin mainly involves RCA, as a study shown by Garg that, 15 out of 4100 patients (0.37%) had abnormal origin of RCA from the left sinus of Valsalva, but only 1 patient (0.02%) had LCA from the right sinus. ⁰³ Although its prevalence is low, but the consequences can be dire ranging from myocardial ischemia to syncope and arrhythmias but the most dreadful condition that it leads to, is sudden cardiac death. ⁰⁴ In an autopsy report, the mortality percentage of Anomalous RCA arising from left sinus was

found to be 57% as compared to be 25% of the anomalous vessel arising from right sinus. ⁰⁵

The condition may have severe consequences, so its diagnosis should be made accurately. There are several methods to evaluate ARCA including Echocardiography (ECHO), angiography, Multi-detector computed tomography (MDCT) and MRI. Transthoracic ECHO is poor in diagnosing this anomaly but Transesophageal ECHO provides better more information. ⁰⁶ Non-invasive tools like MDCT ⁰⁷ and MRI ⁰⁸ are investigations of choice. When MDCT is combined with stress myocardial perfusion scan, the yield of diagnosing the anomaly gets very enhanced. ^{09 10}

The LV function, shown by both MDCT and stress test was depressed confirming obstruction at some part in the RCA causing infarction. There have been proposed many mechanisms of causation of ischemia in this type of vessel like, slit like RCA ostium in aortic wall, acute angulation of the vessel, unusual angle of take-off, tortuous proximal vessel leading to extended atherosclerotic process and lipid accumulation. ^{11, 12} In our case, the vessel was taking interarterial course, i.e. between aorta and pulmonary artery and the pulsations of these big vessels may render that critical part of RCA to episodic occlusion causing ischemia as well as the moderate ostial lesion.

The incidence of stroke after myocardial infarction (MI) is high. A study, by Witt and colleagues 2005 ¹³ showed a 44 fold rise within 30days after mi and even after 3 years it remained 2 to 3 times high. Another study showed a short term incidence of 3.0 to 5.2 percent of stroke ¹⁴ while the 5 year incidence was shown to be 8.1 percent. ¹⁵ Although the incidence of stroke is more in anterior wall MI but the major determinant for thrombus formation leading to embolic stroke is the low ejection fraction ¹⁵, severe apical-wall-motion abnormality ¹⁶ as its absence in both anterior or inferior infarction makes the patient a low risk for stroke. Our patient had, both, the low ejection fraction and apical wall motion abnormality, so

it's a strong suspicion that he formed a thrombus in left ventricle which embolised leading to stroke. Another possibility is that our patient was diagnosed having apical non-compaction which is a potential cause for systolic embolic events.¹⁷

The prevalence of non-compaction of LV in adults is only about 0.014%¹⁸ but more recent studies have found its increase prevalence to about 0.25 to 5 % in varying populations.¹⁹ The occurrence of aberrant RCA with LV non compaction has not yet been reported in literature to our knowledge so it's a unique case.

Revascularization, in the form Coronary Artery Bypass Grafting (CABG), is recommended (Class IA) for documented coronary ischemia in the setting of an anomalous right coronary artery coursing between aorta and pulmonary arteries by American College of Cardiology and American Heart Association (ACC/AHA) guidelines for congenital heart diseases.²⁰

CONCLUSION:

This is a rare case of association of anomalous RCA from LCC with apical non-compaction of LV. The condition may remain asymptomatic but it can leading to devastating conditions like myocardial ischemia, syncope and even sudden cardiac death, so proper and urgent evaluation followed by swift management is the need.

Conflicts of interests

All authors declare no conflict of interest.

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