Anomalous connection of the coronary artery causing ST segment elevation myocardial infarction: A case report

Abstract

Background:

Anomalous connections of the Coronary arteries (ANOCOR) are a rare anatomical abnormality with an angiographic prevalence averaging 1%. The term connection is preferred over origin since the coronary arteries do not grow out of the aorta but grow into the aorta from the peri truncal ring of coronary arterial vasculature. In most cases, these anomalies are incidental findings with no clinical significance; however, it can cause angina, myocardial ischemia and sudden cardiac death (SCD), particularly if associated with a proximal course between the ascending aorta and the pulmonary artery called interarterial course.

Case presentation:

We report a rare case of a patient presenting with an inferior ST segment elevation myocardial infarction (STEMI) which was found to be caused by compression of the proximal right coronary artery (RCA) between the aorta and the pulmonary trunk. Surgery was indicated by the heart team for the treatment of this condition.

Conclusions:

This case presents a severe manifestation of proximal anomalous connection of the right coronary artery with a "malignant" course between the aorta and the pulmonary artery. This variant has been called malignant because of its association with sudden death especially in young asymptomatic athletes. Surgery is recommended as first line therapy in current guidelines for symptomatic ANOCOR with interarterial course.

Background

Proximal Anomalous connections of the Coronary arteries (ANOCOR) are a rare anatomical abnormality with an angiographic prevalence averaging 1% (1). The term connection is preferred over origin since the coronary arteries do not grow out of the aorta but grow into the aorta from the peri truncal ring of coronary arterial vasculature (2). In most cases, these anomalies are incidental findings with no clinical significance; however, it can cause angina, myocardial ischemia, and sudden cardiac death (SCD), particularly if associated with a proximal course between the ascending aorta and the pulmonary artery called interarterial course.

We report a rare case of a patient presenting with an inferior ST segment elevation myocardial infarction (STEMI) which was found to be caused by compression of the proximal right coronary artery (RCA) between the aorta and the pulmonary trunk.

Case presentation

A 29-year-old male presented to our emergency department with complaints of acute chest pain occurred while playing football. There were no risk factors or any significant past medical history. His electrocardiography (ECG) showed ST segment elevation in the inferior territory (Figure 1°), his troponin blood level was significantly high. The patient was referred immediately to the Cath lab where a coronary angiography showed normal left coronary network and an ectopic right coronary artery connected to the left coronary sinus with no visible narrowed segment (Figure 2). CT coronary angiography was then performed using a 64 slice CT scanner and showed the right coronary artery connected to the left coronary sinus, coursing between the aortic root and pulmonary artery with a narrowed proximal segment. (Figure 3 and 4). Cardiac magnetic resonance imaging was performed to eliminate concomitant viral myocarditis and showed subepicardial late gadolinium enhancement

suggestive of ischemic cause rather than myocarditis. Patient was informed of his congenital anomaly. Right coronary artery bypass using left mammary artery graft was indicated by the heart team, but the patient refused the intervention. The patient is until righting this words asymptomatic under betablockers with normal left ejection fraction; he was advised to avoid sports and significant efforts.

Discussion

How can An anomalous connection of the coronary artery causes STEMI?

This case presents a severe manifestation of proximal anomalous connection of the right coronary artery with a "malignant" course between the aorta and the pulmonary artery. Different possible courses of the ectopic right coronary artery are showed in (Figure 5). This variant has been called malignant because of its association with sudden death, especially in young asymptomatic athletes. The RCA connected to the left sinus of Valsalva as a separate vessel or as a branch of a single coronary artery has an incidence of 0.03-0.17% of patients undergoing angiography and associated with a pre-aortic course in nearly 90% cases (3). The frequency of an interarterial course is higher for the RCA than for the left coronary, due to a connection often close to the anterior commissure of the RCA (4). The incidence of sudden death with this anomaly is estimated at 25-40% and is associated with exercise in half of the reported cases (5). One major differential diagnostic in our case was viral myocarditis and this congenital anomaly is only an incidental finding , we had eliminate this diagnosis by performing an MRI study that showed an ischemic pattern of necrosis rather than myocarditis pattern. Various theories have been proposed for this association, including slit like ostium, acute angulation at the origin and compression of the vessel between the aorta and pulmonary artery (6). In an autopsy series of 242 congenital coronary anomalies, 49 ectopic LMCA connections were individualized from the RCS and 52 ectopic connections of the RCA from

the left sinus (3). In this series, 57% of left ANOCORs and 25% of right ANOCORs were associated with SCD, with in the majority of cases the presence of a intraarterial course. The usual profile encountered in this anomaly is a young subject (<35 years) without any cardiovascular history. Serious cardiac events (sudden death, syncope acute myocardial infarction) generally occur during (or just after) intense physical exertion and more particularly in sport practice which was the case for our patient.

How do we manage patients with ANCOR?

The evaluation and management of patients with a "malignant" ANOCOR should be best discussed by a dedicated multidisciplinary team (cardiologists, radiologists, and surgeons with experience in the ANOCOR field). A standardized algorithm, regularly revised, should allow an optimal decision-making for each patient according to the initial presentation and diagnostic work-up. Vast majority of malignant ANOCORs may remain asymptomatic for a long time, ischemic symptoms, that may mimic a coronary artery disease, are possible. ANOCOR diagnosis is made by invasive coronary angiography or Coronary CT scan. Given the difficulty to demonstrate myocardial ischemia by functional non-invasive tests in ANOCOR, it seems acceptable to consider non-equivocal symptoms (angina, dyspnea, and syncope, especially on exertion) as ischemic symptoms, even if documented myocardial ischemia is lacking. Silent myocardial ischemia and severe ventricular arrhythmias must be considered in the same way (7).Surgery is recommended as first line therapy in current guidelines for symptomatic ANOCOR with interarterial course. In the expert consensus guidelines from the American Association for Thoracic Surgeons , the authors give a Class 1/Level B indication for any patient with a left ANOCOR at risk, with or without symptoms, or with a symptomatic right ANOCOR at risk (8).In the 2018 AHA/ACC guidelines for the management of adults with congenital heart disease, surgery is recommended (Class 1, Level B-NR) for ANOCOR at risk with ischemic symptoms or myocardial ischemia and is

reasonable (Class IIa/Level C-LD) for left ANOCOR at risk without ischemic symptoms or documented myocardial ischemia (9). Surgery or continued observation may be reasonable (Class IIb/Level B-NR) in asymptomatic right ANOCOR without inducible myocardial ischemia or anatomic severity criteria. The published literature of percutaneous coronary intervention (PCI) for ANOCOR with an interarterial course remains limited (10).

What is the place of PCI in these anomalies?

The place of PCI is only addressed in AATS guidelines for adults with high risk for surgery (Class IIb/Level C). Patients aged over 30 years with right ANOCOR associated with ischemic symptoms and/or documented myocardial ischemia represent the potential population eligible for percutaneous coronary intervention (PCI). PCI guidance by intravascular ultrasound (IVUS) is recommended for the evaluation of the ectopic segment (diameters, area, and length) and for the control after stenting.10 Direct stenting is recommended (11).

Conclusions

In Young patients presenting with myocardial infarction; anomalous coronary artery connection with an interarterial course should always be seeking for. Coronary CT scan plays an important role in the diagnosis and the therapeutic decision which must always be supported by a heart team.

List of abbreviations:

AHA/ACC: American heart association/ American college of cardiology

ANOCOR: Anomalous connections of the Coronary arteries

CT: Computed tomography

ECG: Electrocardiogram

IVUS: Intravascular ultrasound

LMCA: Left main coronary artery

PCI: Percutaneous coronary intervention

RCA: Right coronary artery

SCD: Sudden cardiac death

STEMI: ST segment elevation myocardial infarction

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Figures:

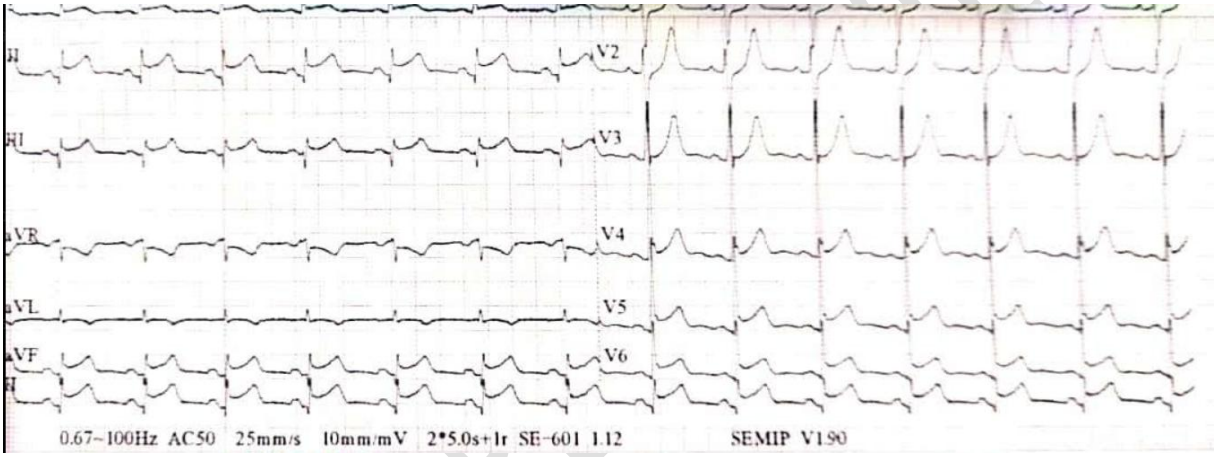

Figure 1: Electrocardiogram showing ST segment elevation in the inferior leads (DII,DIII,aVF)

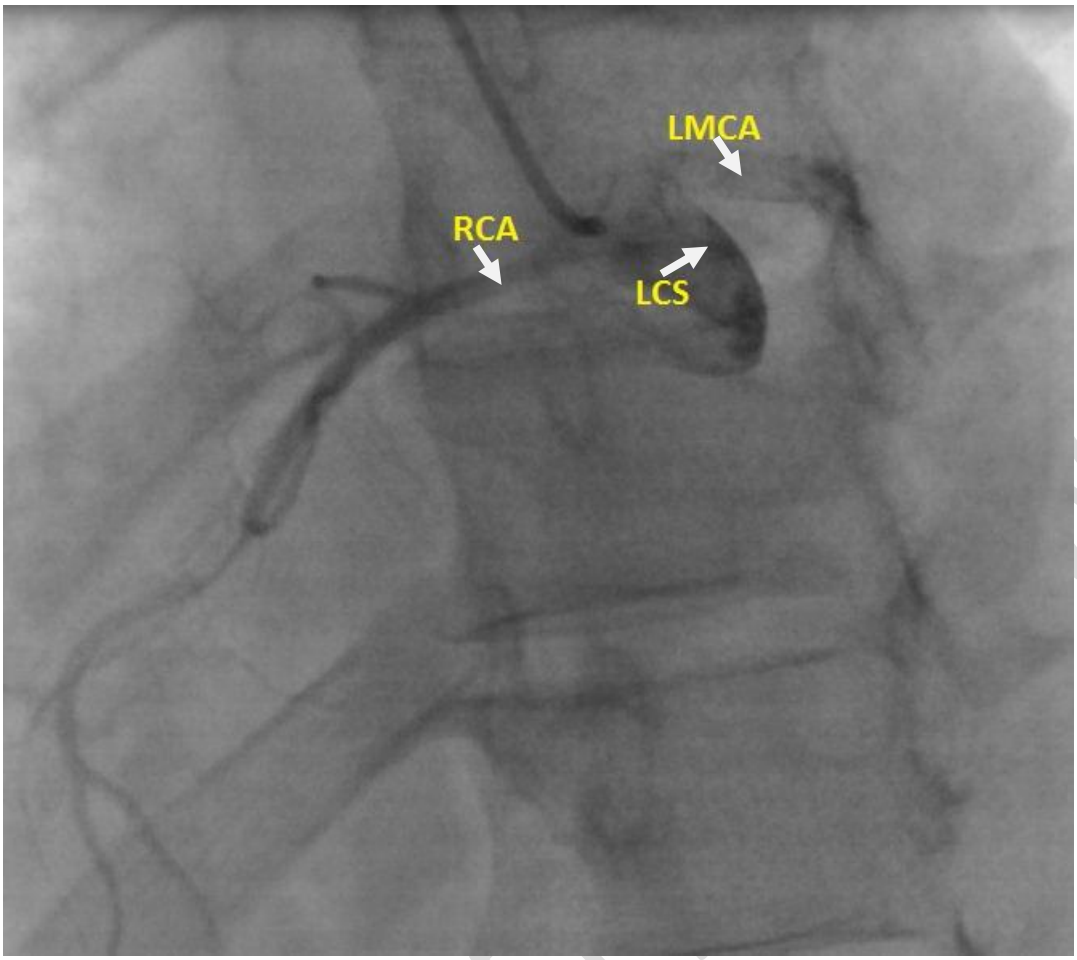

Figure 2: Coronary angiography showing: both right coronary artery (RCA) and left main coronary artery (LMCA) connected to the left coronary sinus (LCS).

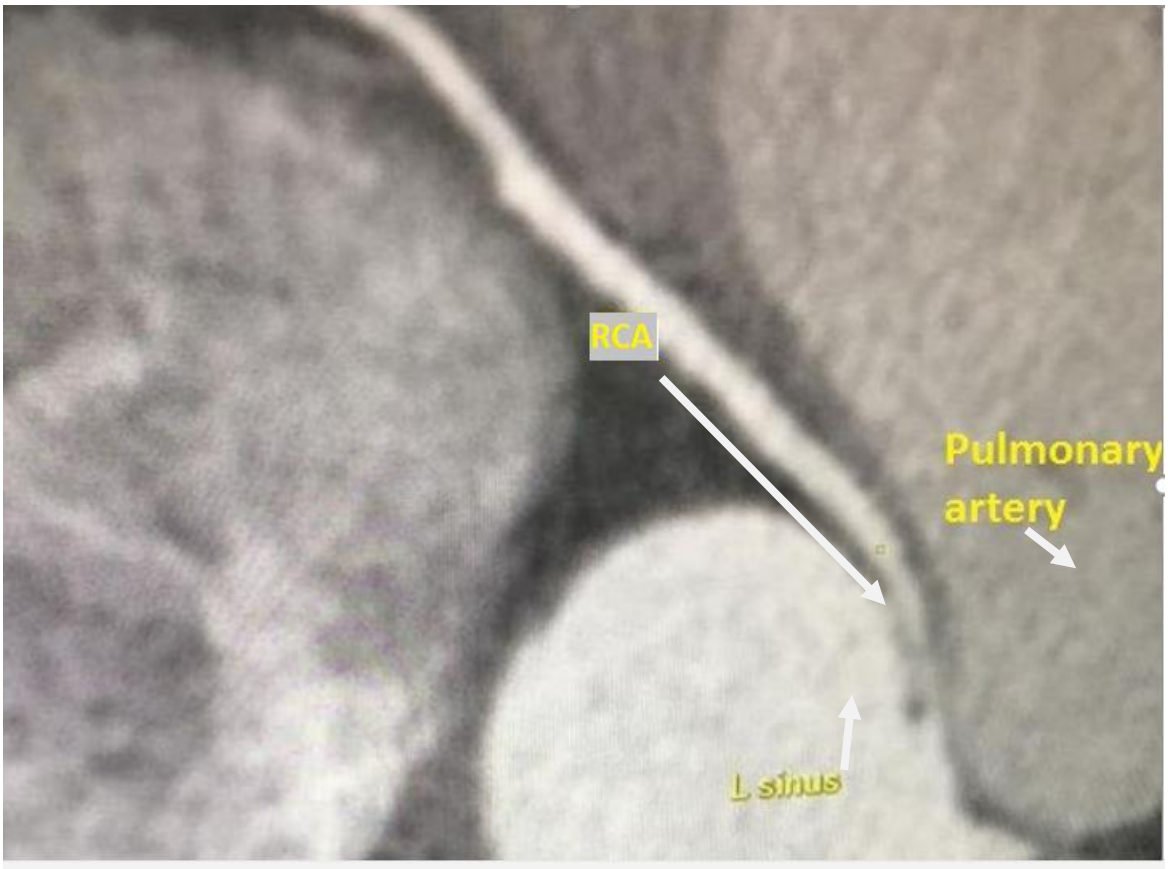

Figure 3: Cross section of coronary CT scan showing the right coronary artery (RCA) connected to the left coronary sinus (L.Sinus), coursing between the aortic root and pulmonary artery with a narrowed proximal segment

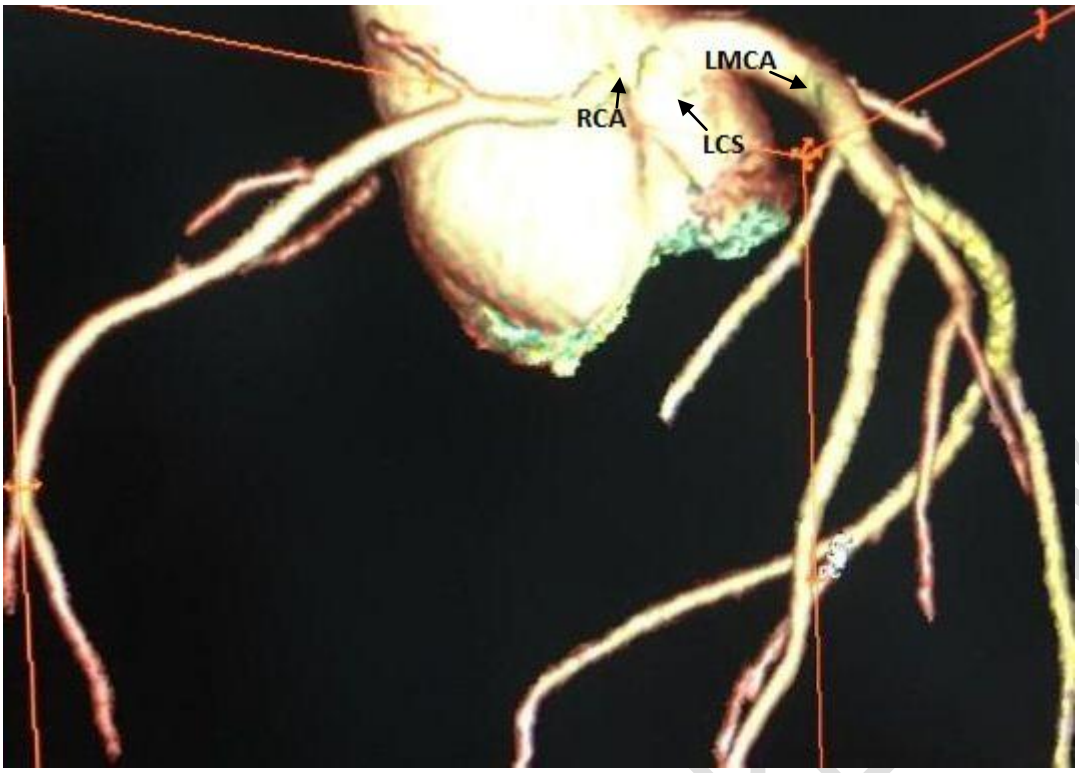

Figure 4: Cross section of coronary CT scan showing the right coronary artery (RCA) connected to the left coronary sinus (L.Sinus), coursing between the aortic root and pulmonary artery with a narrowed proximal segment

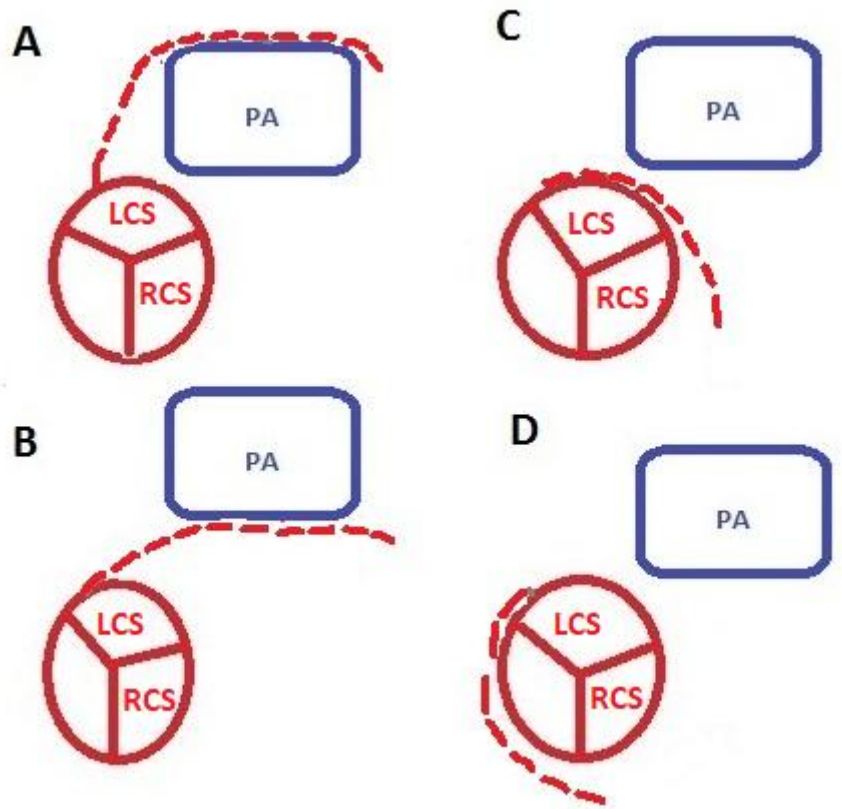

Figure 5: Different initial proximal courses in the case of a right anomalous coronary arterie connection: prepulmonary course (A),Retropulmnary course (B) interarterial course (our case) (C) retroaortic course (D)