

Right Coronary Artery Anomalous Origin from Left Anterior Descending Artery Detected by Computed Tomography Coronary Angiogram: A Case Report and Review of the Literature

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Abstract: Coronary anomalies are generally rare and recognized in approximately 1.3% of coronary angiograms. The right coronary artery originating from the left coronary system is an extremely rare variation of the single coronary artery anomaly in which the prognosis is usually benign provided that the anomalous vessel does not pass between the aorta and the pulmonary artery. Right coronary artery anomaly can be associated with other congenital cardiovascular anomalies such as bicuspid aortic valve, transposition of the great vessels and tetralogy of Fallot.

The clinical significance of coronary anomalies is usually determined by underlying anatomic features of the anomalous coronary origin and/or coronary atherosclerosis. Initial presentations include: chest pain, myocardial infarction, arrhythmias, sudden death, and quite rarely exertion syncope. Although coronary angiography is an important diagnostic method, new non-invasive methods such as coronary computed tomography angiography and cardiac magnetic resonance imaging have important roles to play in characterizing this coronary anomaly. It should be noted that the management strategy of these patients may vary based on clinical presentation and anatomical details.

We report a Case of exertion dizziness, diagnosed to have anomalous origin of right coronary artery from left anterior descending artery diagnosed by computed tomography coronary angiography. Medical treatment was effective in relieving the patient's symptoms and ischemia.

Keywords: Right coronary artery anomaly, diagnosis, CT Coronary angiography.

INTRODUCTION

Coronary anomalies are generally rare, carrying an incidence of 0.6 – 1.3% of angiographic series, and 0.3 % of autopsy series [1-28].

Anomalous right coronary artery (RCA) origin, an extremely rare condition, is known to cause myocardial ischemia, arrhythmia, syncope, and has recently become recognized as a cause of sudden cardiac death in the young [1-4].

The RCA originating from the left coronary system is an extremely rare variation of the single coronary artery anomaly in which the prognosis is usually benign provided that the anomalous vessel does not pass between the aorta and the pulmonary artery.

The incidence of anomalous RCA originating from the left coronary system ranges from 0.1% to 0.9%. The origin of an anomalous RCA may be from the left sinus of Valsalva, the posterior sinus of Valsalva, the ascending aorta, the pulmonary artery (PA), the left ventricle, the left main (LMCA), the left circumflex (LCX) or the left anterior descending (LAD) [1-28].

The origin of an anomalous RCA may also be viewed as an extension of the second diagonal branch or as a limb of the first septal perforator [29-31]. A recent report [32] identified that three different anomalous arteries originating from the mid LAD corresponded with the district of RCA. Cases of anomalous RCA from the LAD have only been rarely reported in the literature (30 cases in PubMed). In the majority of the cases published, anomalous RCA stems from the proximal or mid segment of the LAD and usually courses anterior to the PA to reach the right AV groove or in between the great vessels, and rarely crosses posterior to the aorta [1-36, 37-54].

We report a Case of exertion dizziness diagnosed to have anomalous origin of RCA from LAD by computed tomography (CT) coronary angiography. Medical treatment was effective in relieving the patient's symptoms and ischemia.

CASE PRESENTATION

A 35 year-old man was admitted to our hospital complaining of exertion dizziness and chest pain. His medical history consisted of hypertension and hyperlipidemia. He was a non-smoker and he had a one month history of exertion dizziness and chest pain. On admission, his ECG and cardiac enzyme levels were normal. Transthoracic echocardiography was within normal limits. Treadmill exercise electrocardiogram showed 1 mm ST segment depression in

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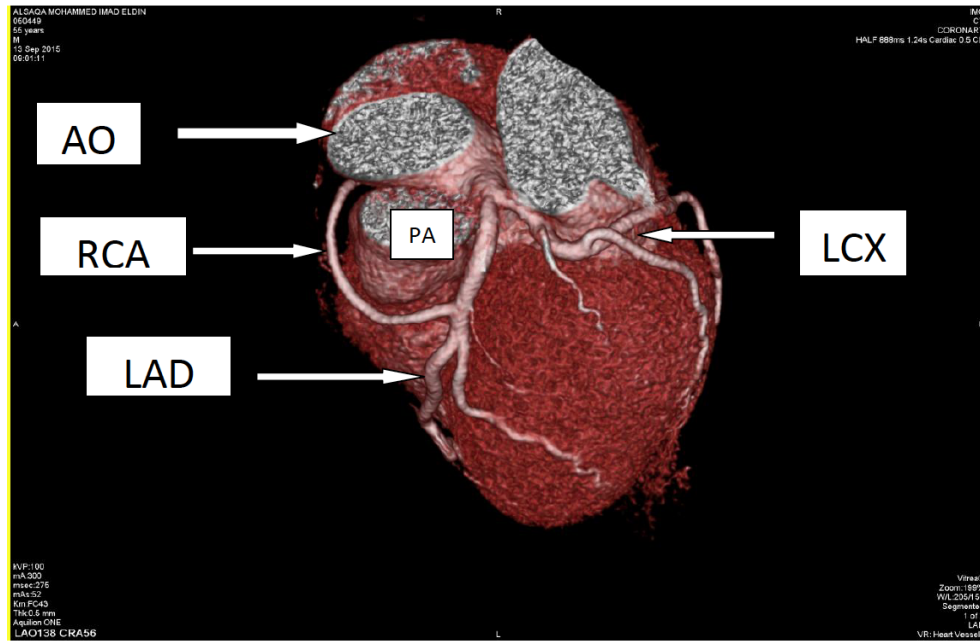


Figure 1: An anomalous right coronary artery (RCA) arising from the proximal mid portion of the left anterior descending artery (LAD) passes anterior to the pulmonary artery (PA) before reaching the right atrioventricular groove; LCX — left circumflex artery; Ao — aorta.

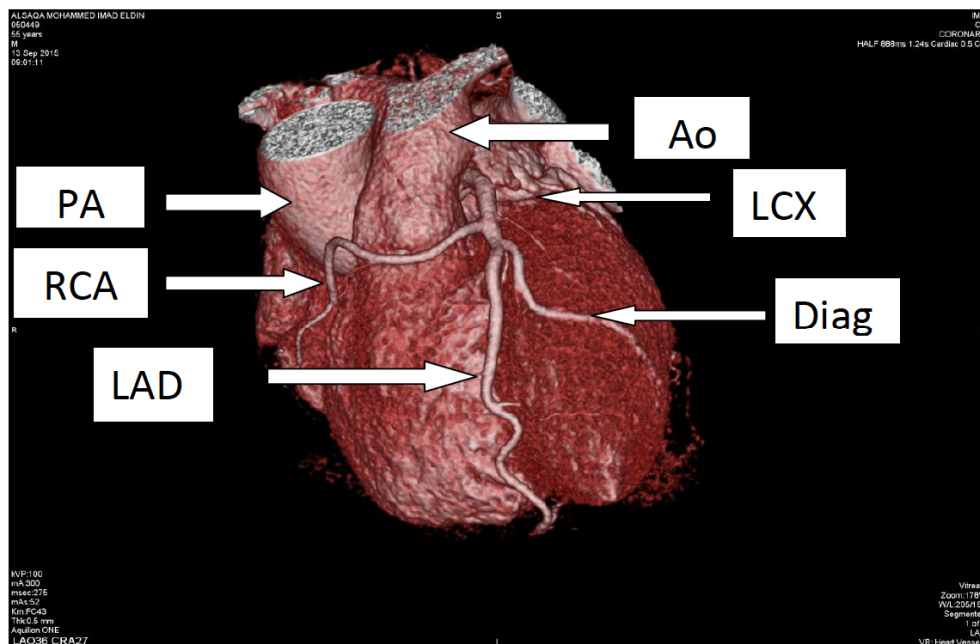


Figure 2: An anomalous right coronary artery (RCA) arising from the proximal mid portion of the left anterior descending artery (LAD) passes anterior to the pulmonary artery (PA) before reaching the right atrioventricular groove; Diag- diagonal artery; LCX — left circumflex artery; Ao — aorta.

inferior leads. Coronary computed tomography (CT) angiography displayed normal courses of the LMCA, dominant LCX, and LAD (Figure 1). An anomalous RCA as a separate small branch arose from the mid LAD close to the third diagonal branch, and then coursed anteriorly down the right atrioventricular (AV) groove (Figures 1,2). No vessel was seen originating

from the right aortic cusp or sinus. Neither aortography nor pulmonary artery (PA) CT angiography showed the presence of an origin of a supplementary RCA from another site, such as the PA and the aorta. This was concluded to be a benign anomaly from the data of a nuclear medical study that did not show any hypoperfused region in the myocardium. Therefore, the

patient was only given long-term medical therapy for hyperlipidemia and hypertension. At follow-up, it was observed that the patient had an asymptomatic clinical status.

DISCUSSION

Coronary artery anomalies (CAAs) occur during the third week of fetal development. The reported incidence of CAAs ranges from 0.6% to 5.6% of patients undergoing diagnostic coronary angiography, and in approximately 1% of routine autopsy examinations. The most common CAA is a separate origin of the LAD and LCX, with an incidence of 0.41%, followed by the LCX arising from the RCA, with an incidence of 0.37%. CAAs result from several anatomic aspects according to their origin, course and distribution.

The incidence of anomalous RCA originating from the left coronary system ranges from 0.1% to 0.9%. The origin of an anomalous RCA may be from the left sinus of Valsalva, the posterior sinus of Valsalva, the ascending aorta, the PA, the left ventricle, the LMCA, the LCX or the LAD [1-28].

The origin of an anomalous RCA may also be viewed as an extension of the second diagonal branch or as a limb of the first septal perforator [29-31]. A recent report [32] identified that three different anomalous arteries originating from the mid LAD corresponded with the district of RCA. Cases of anomalous RCA from the LAD have only been rarely reported in the literature (30 cases in PubMed).

In the majority of the cases published, anomalous RCA stems from the proximal or mid segment of the LAD and usually courses anterior to the PA to reach the right AV groove or in between the great vessels, and rarely crosses posterior to the aorta [1-36, 37-54].

The distribution of an anomalous RCA typically corresponds to the region supplied by the RCA that derives from the normal right aortic sinus, except for the case of an anomalous RCA arising from the LAD with a coexisting proximal RCA. In such a case, while the area of proximal RCA is served by a separate branch originating in the normal right aortic trunk, an anomalous artery from the LAD serves the distal RCA region [35]. The RCA originating from the LAD is an extremely rare variation of isolated single coronary artery (SCA), which is a very rare congenital anomaly appearing in approximately 0.024–0.066% of the

general population undergoing coronary angiography [1-25, 29-33]. In these cases, one coronary artery stems from a single coronary ostium from the aortic trunk that nourishes the entire myocardium. It is well known that SCA is commonly associated with other congenital cardiovascular anomalies such as bicuspid aortic valve, transposition of the great vessels, coronary arteriovenous fistula, origin from the pulmonary artery, truncus arteriosus, ventricular septal defect, patent ductus arteriosus, patent foramen ovale and tetralogy of Fallot [1-38]. The most common co-existing cardiac anomaly with SCA is transposition of the great vessels. The great majority of cases of SCA with anomalous RCA from the LAD are less frequently associated with other congenital cardiovascular defects [1-38].

CLASSIFICATION

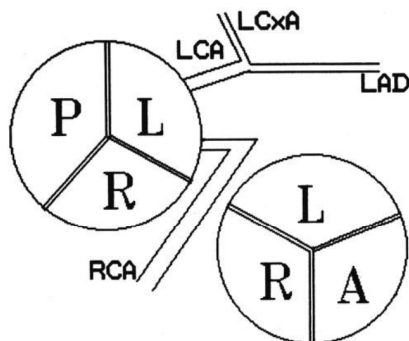
Angiographic identification of anomalous coronary arteries is crucial for the appropriate diagnosis and management of patients. Hence, attempts at classifications have been made by several authors [5,6,8-15,36]. Based on the modified Lipton classification [6], cases of anomalous RCA originating from the LAD correspond to an LII anomaly. However, anomalous RCA stemming from the mid LAD cannot be assigned an actual location according to these classifications. This is because group II anomalies have been only designed for “those arising from the proximal part of the normal right or left coronary artery”, and it has not been precisely defined for a coronary artery anomaly that derives from the mid-section of the LAD. For this reason, the real number of cases of RCA stemming from the mid LAD may have been misreported.

ANATOMICAL IMPORTANCE

The prognosis of anomalous RCA from the LAD is usually benign, and does not interfere with coronary perfusion. However, if an abnormal coronary artery crosses between the PA and the aorta, myocardial ischemia and sudden death may occur [1-16,27,34,41]. The clinical relevance of this coronary anomaly may be due to underlying coronary atherosclerosis. It has been proposed that abnormal origin and course of anomalous coronary arteries could make them more prone to atherosclerosis [40]. Coronary atherosclerosis or stenosis requiring medical, percutaneous or surgical revascularization has been observed in about 40% of reported cases [17,18,23,25-32,40-51]. In the minority of patients with CCAs (nearly 15%), myocardial

ischemia can develop even in the absence of atherosclerosis. Non-atherosclerotic causes of myocardial ischemia in coronary anomaly include: coronary vasospasm, acute angle take off, slit-like orifice, intramural course, and compression by the great vessels.

Several potential mechanisms have been proposed to explain myocardial ischemia and sudden death in patients with CAAs: i) spasm of the anomalous coronary artery, possibly as a result of endothelial injury or ischemia caused by its long distance of traveling; ii) the acute angle of takeoff of the anomalous vessel; iii) slit-like orifice; iv) intramural course of the anomalous vessel; and v) compression of the anomalous artery between the pulmonary and aortic trunks, particularly during or immediately after exercise, which leads to expansion of the aortic root and pulmonary trunk, creating external coronary artery compression and possibly increasing the pre-existing angulations of the coronary artery take off, with a reduction in the luminal diameter in the proximal portion of the anomalous coronary artery (Figure 3).



Right coronary artery (RCA) originating from above left coronary cusp. Proximal portion curve in between the aorta (posterior) and pulmonary trunk (anterior).

Figure 3: An anomalous right coronary artery (RCA) arising from above the left coronary cusp (LCS) passes between the aorta (Ao) and pulmonary artery (PA) before reaching the right atrioventricular groove; LAD- left anterior descending artery; LCX — left circumflex artery; LCS — left main coronary artery.

The clinical significance and risk of a coronary anomaly usually depend on the place and course of the anomalous coronary artery origin, and the degree and localization of the culprit stenotic or atherosclerotic lesion [1-16,27,28,41, 43-52].

CLINICAL PRESENTATION

The clinical presentation of a patient with a coronary anomaly can include chest pain, dyspnea, palpitations,

syncope, ventricular fibrillation, myocardial infarction and sudden death especially following exertion. Yet coronary anomalies may also be asymptomatic. Taylor *et al.* [41], in their study of 52 patients with anomalous origin of the RCA, noted that 25% had died suddenly and in most cases asymptotically.

The delay in diagnosis may be fatal in patients with clinically silent coronary anomaly. Therefore, early identification of patients with a coronary anomaly is crucial as they can be saved by medical, percutaneous intervention and/or surgical therapy.

DIAGNOSTIC APPROACH

The diagnosis of these anomalies may not be exactly determined by defects observed by non-invasive tests such as electrocardiography, echocardiography, cardiovascular stress testing and nuclear medicine [1,2]. So far, the main diagnostic tool for the determination of coronary vessel anomalies has been selective coronary angiography. Although coronary angiography is an effective and important diagnostic method, it has some disadvantages owing to its invasive nature. Due to developments in technology, new non-invasive methods such as coronary computed tomography angiography (CTA) and cardiac magnetic resonance imaging (MRI) play an important clinical role in determining coronary anomalies [2]. Cardiac MRI and coronary CT angiography may be superior to conventional angiography, especially in patients with congenital defects and isolated coronary anomalies [2].

The most important feature of cardiac MRI is that it does not involve contrast agents and radiation. However, because of the low spatial resolution, this technique is currently less helpful in evaluating the distal coronary system [48,49]. In contrast, although it has some limitations associated with the administration of ionizing radiation and potentially allergenic or nephrotoxic agents, coronary CTA is a very good diagnostic tool in delineating coronary anomalies, because it has high spatial resolution and rapid acquisition. Additionally, with the use of electrocardiographic gating, coronary CTA provides excellent high-quality images of the coronary arteries, including their origin, course and termination.

Thus, this technique that allows for evaluating the comprehensive anatomy of coronary arteries could also be helpful in detecting future therapeutic methods [21,31,50-52]. In one reported case, a malignant right coronary anomaly coursing between the two great

arteries, which could not be seen by conventional coronary angiography, was demonstrated by multi-slice coronary CTA [53].

TREATMENT OPTIONS

Treatment for a coronary anomaly may be medical, percutaneous and/or surgical. Some difficulties may be encountered such as frequent catheter exchanges before finding the best-fitting catheter and guide wire, giving rise to increased fluoroscopic time and a greater amount of contrast agent used during coronary angiography and/or percutaneous coronary intervention (PCI) by reason of different anatomical structures in these patients. PCI for an anomalous RCA arising from the left coronary system has been reported by a variety of authors [22,28,37,40-42].

Cardiovascular surgery is the management of choice of a coronary anomaly of a patient who cannot be effectively treated by PCI. Proposed options for those who need surgery include ostioplasty, bypass grafting of the RCA, and translocation of the RCA to the aorta [29,43-47,53-57]. Gulati *et al.* [58] reported that a PA translocation procedure could be undertaken in patients with a single coronary artery arising from the opposite coronary ostium and passing between the great arteries in the absence of other factors associated with coronary insufficiency, such as fixed obstructive coronary lesions or slit-like ostium. However, the long-term benefits of such therapies have not yet been confirmed.

CONCLUSION

Anomalous RCA origin is extremely rare, has recently become recognized as a cause of angina, arrhythmia and sudden cardiac death in all age group. In single coronary subtype, RCA originates from the proximal or mid-portion of LAD, and crosses anterior to PA before reaching the right AV groove. Current classifications may have led to underestimation of the correct incidence. Approximately 15% of patients may have myocardial ischemia caused by the abnormal anatomy of the right coronary artery. In patients with a coronary vessel anomaly, an angiographic study such as conventional and coronary CT should be performed, and the management strategy of these patients may vary, based on clinical presentation and anatomical detail.

CONFLICT OF INTEREST

No Conflict of Interest exists.

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