Case Report

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An isolated single L-II type coronary artery anomaly: A rare coronary anomaly

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Summary **The incidence of congenital artery anomalies is 0.2-1.4%, and most are benign. Single coronary artery (SCA) anomalies are very rare. The right coronary artery (RCA) originating from the left coronary system is one such SCA anomaly, and the risk of sudden cardiac death (SCD) increases if it courses between the pulmonary artery and aorta and coexists with other congenital heart diseases. Additionally, coursing of the RCA between the great vessels increases the risk of atherosclerosis. We herein present the case of a 57 year-old man who was admitted to our cardiology outpatient clinic and diagnosed with an SCA anomaly in which the RCA arose from the left main coronary artery (LMCA) and coursed between the pulmonary artery and aorta. However a critical stenosis was not detected in imaging techniques, and myocardial perfusion scintigraphic evidence of ischaemia was found in a small area. Therefore, he was managed with conservative medical therapy.**

Keywords: Coronary vessel anomalies, coronary angiography, multidetector computed tomography

1. Introduction

Although most coronary artery anomalies are benign and are detected incidentally during diagnostic angiography, some anomalies result in catastrophic clinical outcomes such as sudden cardiac death (SCD) (*1*). Therefore, these anomalies are among the most complex and significant subjects in the field of cardiology. An anatomically correct definition of these anomalies is important to predict complications that may develop during myocardial revascularisation. We herein present a case involving a patient with a single coronary artery (SCA) anomaly in which the right coronary artery (RCA) arose from the left main coronary artery (LMCA) and coursed between the pulmonary artery and aorta.

2. Case report

A 57 year-old male was admitted to our outpatient clinic because of a 3-month history of exertional dyspnoea. He had no known history of coronary artery disease (CAD) or systemic disease. This patient's cardiovascular risk factors included a smoking habit (20 pack-years) and older age. Physical examination revealed a blood pressure of 120/80 mmHg and heart rate of 78 bpm. Increased bronchovascular branching was noted on telecardiography, and electrocardiography revealed a normal sinus rhythm. His echocardiographic findings were normal. A 1-mm ST depression was observed in the inferior leads during a treadmill exercise test; however, the patient did not have typical chest pain accompanying the electrocardiographic changes. He underwent coronary angiography (CAG) with a prediagnosis of CAD after he had been evaluated at a pulmonology clinic. The LMCA was selectively cannulated with a JL4-6F diagnostic catheter (Diagnostic catheter, Medtronic, New York, USA). No atherosclerotic lesions were observed on CAG; however, an SCA anomaly was seen in which the RCA originated from the LMCA (Figure 1). Additionally, the RCA coursed between the aorta and pulmonary artery, and a critical stenosis was not detected (Figure 2). Multislice computed tomography (CT) angiography was performed after the patient was discharged 1 week later on account of the fact that it can assist delineating the proximal course of the artery and

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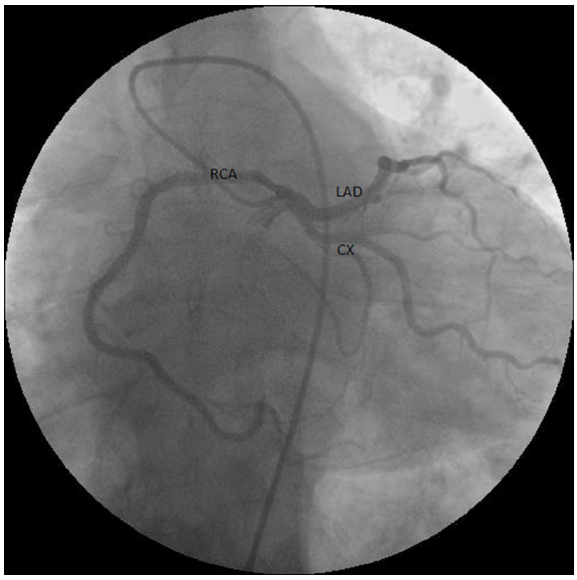

Figure 1. Coronary angiography revealed an SCA anomaly in which the RCA originated from the left main coronary artery (LMCA). SCA, single coronary artery; RCA, right coronary artery.

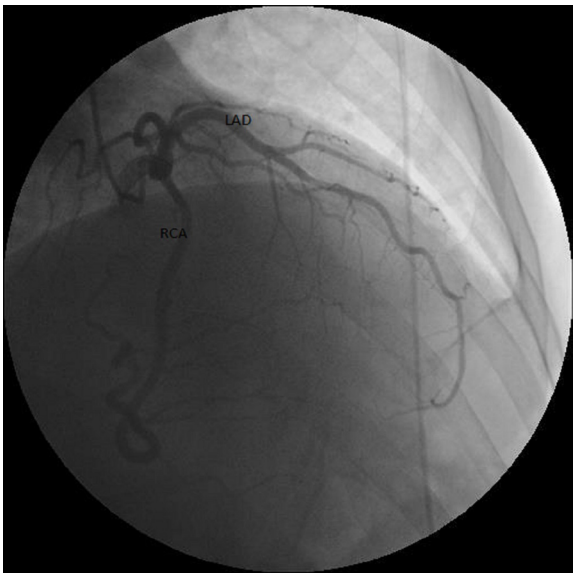

Figure 2. Coronary angiography revealed that the RCA coursed between the aorta and pulmonary artery.

provides excellent high-quality images. Thus, the origin and course of the RCA were able to be better evaluated with three-dimensional imaging (Figure 3). Resting and stress Tc-99m tetrofosmin single photon emission CT (SPECT) revealed 7% ischaemia in the RCA vascular territory. In addition, no other congenital cardiac defects accompanying the coronary anomaly were seen in our patient. Therefore, he was managed with conservative treatment comprising a betablocker and isosorbide mononitrate therapy.

3. Discussion

Congenital coronary artery anomalies are seen in 0.2-

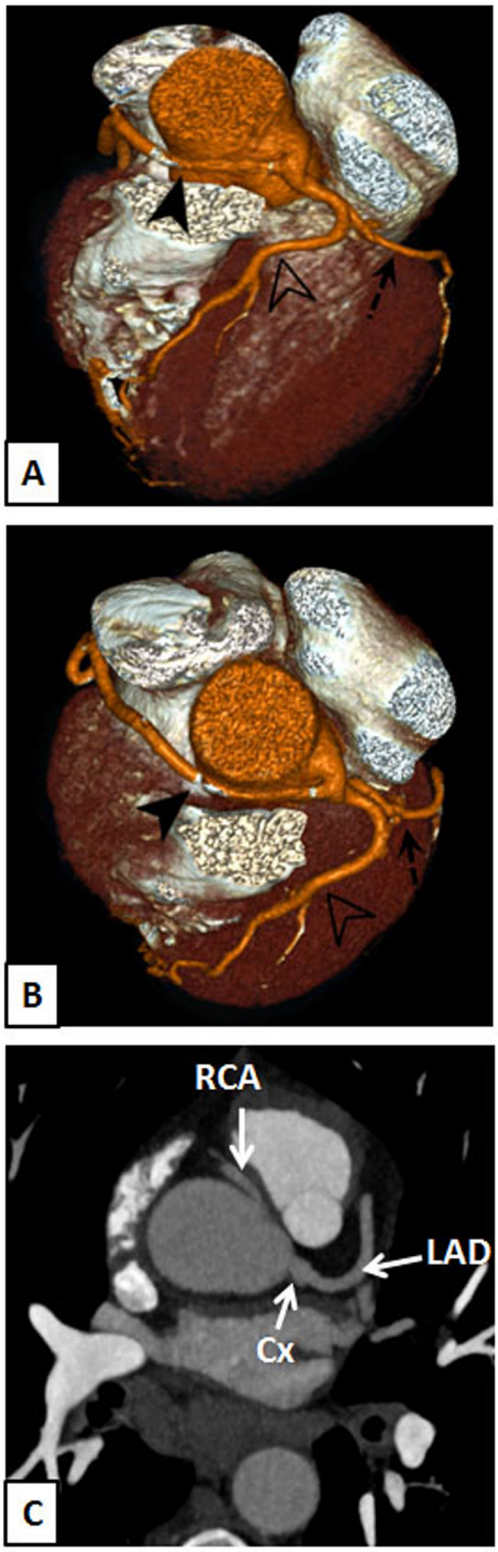

Figure 3. A three-dimensional volume-rendering image (A, B) and multiplanar reconstruction axial image (C) showing the SCA anomaly. RCA, black arrow head; LAD, white arrow head; CXA, dashed arrow.

1.4% of the normal population (*2*). Lipton *et al*. (*3*) reported that an SCA originating from the right or left coronary system constituted 3.3% of all congenital coronary anomalies in their study. The classification of SCA anomalies have been made by some authors.

For instance, in 1950, Smith described three different types of SCA (*4*). Recognizing the deficiency of such broad categories, Ogden and Goodyer in 1976 proposed a more complete classification (*5*). They classified the SCA into 14 basic distribution patterns. Yamanaka and Hobbs in 1990 modified the classification previously described by Lipton *et al*. (*6*). Based on the modified Lipton classification, each anomaly is coded with an R or L depending on the localisation of the sinus from which the coronary anomaly originates. Additionally, the anomalies are classified into three types according to the anatomical distribution on the ventricular surface: type I = the vessel follows the course of normal left or right coronary artery with a continuation into the missing artery's territory; type II = an anomalous artery arises from the proximal part of the other normal artery and courses the base of the heart before taking the native course; and type III = LAD and CX arteries originating from the proximal part of the RCA. The third component of the Lipton classification associated with the course of the transfer branch: the aberrant artery could take one of several different pathways to reach its vascular territory. These pathways are indicated as: type A (anterior to the right ventricular outflow tract); type B (between the aorta and pulmonary trunk); type P (posterior to the great vessels); type S (above the interventricular septum); type C (combined type). The present case can be classified as an isolated single L-II B subtype coronary artery anomaly.

Furthermore, RCA anomalies have many origins; they may originate from the left or posterior sinus of Valsalva, ascending aorta, pulmonary artery, LMCA, LAD, or CXA. The incidence of SCA anomalies involving the RCA originating from the left coronary system (L-I and L-II) was reported as 0.016-0.090% in the study by Lipton *et al*. (*3*). This incidence was reported as 0.036% among 16,573 patients included in the retrospective CAG screening study by Yuksel *et al*. (*7*). In the literature, most RCAs originated from the proximal or mid-LAD; RCA anomalies originating from the left coronary cusp or LMCA have been rarely reported.

SCA anomalies may coexist with other congenital heart diseases, mainly transposition of the great vessels followed by coronary arteriovenous fistula, bicuspid aortic valve, tetralogy of Fallot, truncus arteriosus, ventricular septal defect, patent ductus arteriosus, and patent foramen ovale (*8*). SCA anomalies may appear incidentally on CAG during screening and are known to increase the likelihood of the coexistence of congenital cardiac defects. No other congenital cardiac defects accompanying the coronary anomaly were seen in our patient.

Most patients are asymptomatic and have a benign clinical course. Nevertheless, an increased incidence of atherosclerosis is observed among patients with SCA anomalies although the relationship between atherosclerosis and SCA anomalies is not definitive. A few potential mechanisms have been proposed to explain atherosclerosis. Abnormal origin, long traveling distance, intramural course of the aberrant artery and particularly compression between the great vessels may precipitate endothelial injury and atherosclerosis. Atherosclerosis requiring medical therapy or percutaneous or surgical revascularisation has been seen in approximately half of the reported cases to date (*9*). This finding seems to support the theory that the risk of atherosclerosis is higher in patients with than without SCA anomalies. Symptoms including chest pain, dyspnoea, palpitation, syncope as in CAD, and myocardial infarction and SCD may be seen due to myocardial ischaemia. In patients without atherosclerosis, the development of ischaemia may be explained by the stenotic slit-like orifice, acute angle take-off, coronary vasospasm or compression of great vessels. If the RCA courses between the pulmonary artery and aorta and is under mechanical compression, coronary perfusion decreases. As a result, increased great vessel dilation leads to myocardial ischaemia, particularly during exercise, and arrhythmia and SCD may occur. Taylor *et al*. (*10*) investigated the association between SCD and congenital coronary artery anomalies. They reported that the incidence of exercise-related SCD was significantly higher in a nonatherosclerotic young population with an abnormal RCA originating from the left coronary system and coursing between the pulmonary artery and aorta. Our patient had an SCA anomaly with specific cardiovascular risk factors; however, his CAD was nonatherosclerotic and his RCA coursed between the great vessels. 7% ischaemia in the RCA vascular territory was seen on SPECT examination due to extrinsic compression. Therefore a surgical procedure was not offered to treat, and he was managed with conservative medical therapy.

Although cardiac catheterisation is the gold standard for the identification of coronary anomalies, coronary CT angiography is a useful noninvasive method for evaluating the course of these abnormal coronary arteries, verifying the diagnostic accuracy, and determining the optimal treatment. The excellent spatial resolution of coronary CT angiography makes this technique very suitable to detect the relationship of the anomalous vessels with the aorta, pulmonary artery and cardiac structures (*11,12*). Besides, cardiac magnetic resonance imaging (MRI) is a convenient tecnique to determine coronary anomalies and it may be superior to conventional angiography, particularly in patients with congenital heart defects. However, due to low spatial resolution, this imaging technique is already less helpful in evaluating the distal coronary system (*13,14*). Therefore, we diagnosed the SCA anomaly by cardiac catheterisation and further preferred coronary CT angiography to delineate the course of the aberrant artery in relation to the great vessels.

4. Conclusion

An SCA anomaly involving the RCA originating from the left coronary system is an important risk factor for SCD if the RCA courses between the great vessels and a critical stenosis is detected. However, in our case a critical stenosis was not detected in imaging techniques. Therefore conservative medical therapy was preferred. While cardiac catheterisation is the gold standard for the identification of coronary anomalies, coronary CT angiography is a useful noninvasive imaging technique and plays an important role for the diagnosis of such anomalies.

References

1. Song SH, Suh SE, Jin SM, Moon JH, Cho YK, Lim SW. Myocardial ischemia caused by paroxysmal supraventricular tachycardia in a patient with anomalous origin of right coronary artery arising from left sinus of valsalva. Korean Circ J. 2013; 43:123-126.
2. Jacobs ML, Mavroudis C. Anomalies of the coronary arteries: Nomenclature and classification. Cardiol Young. 2010; 20 (supple 3):15-19.
3. Lipton MJ, Barry WH, Obrez I, Silverman JF, Wexler L. Isolated single coronary artery: Diagnosis, angiographic classification, and clinical significance. Radiology. 1979; 130:39-47.
4. Smith JC. Review of single coronary artery with report of 2 cases. Circulation. 1950; 1:1168-1175.
5. Ogden JA, Goodyer AV. Patterns of distribution of the single coronary artery. Yale J Biol Med. 1970; 43:11-21.
6. Yamanaka O, Hobbs RE. Coronary artery anomalies in 126,595 patients undergoing coronary arteriography. Cathet Cardiovasc Diagn. 1990; 21:28-40.
7. Yuksel S, Meric M, Soylu K, Gulel O, Zengin H, Demircan S, Yilmaz O, Sahin M. The primary anomalies of coronary artery origin and course: A coronary angiographic analysis of 16,573 patients. Exp Clin Cardiol. 2013; 18:121-123.
8. Poynter JA, Williams WG, McIntyre S, Brothers JA, Jacobs ML; Congenital Heart Surgeons Society AAOCA Working Group. Anomalous aortic origin of a coronary artery: A report from the Congenital Heart Surgeons Society Registry. World J Pediatr Congenit Heart Surg. 2014; 5:22-30.
9. Yurtdas 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.
10. Taylor AJ, Rogan KM, Virmani R. Sudden cardiac death associated with isolated congenital coronary artery anomalies. J Am Coll Cardiol. 1992; 20:640-647.
11. Aldana-Sepulveda N, Restrepo CS, Kimura-Hayama E. Single coronary artery: Spectrum of imaging findings with multidetector CT. J Cardiovasc Comput Tomogr. 2013; 7:391-399.
12. Yadav A, Buxi TB, Rawat K, Agarwal A, Mohanty A. Anomalous single coronary artery on low dose MDCT. J Radiol Case Rep. 2013; 7:6-15.
13. McConnell MV, Stuber M, Maning WJ. Clinical role of coronary magnetic resonance angiography in the diagnosis of anomalous coronary arteries. J Cardiovasc Magn Reson. 2000; 2:217-224.
14. American College of Cardiology Foundation Task Force on Expert Consensus Documents, Hundley WG, Bluemke DA, Finn JP, Flamm SD, Fogel MA, Friedrich MG, Ho VB, Jerosch-Herold M, Kramer CM, Manning WJ, Patel M, Pohost GM, Stillman AE, White RD, Woodard PK. ACCF/ACR/AHA/NASCI/SCMR 2010 expert consensus document on cardiovascular magnetic resonance: a report of the American College of Cardiology Foundation Task Force on Expert Consensus Documents. Circulation. 2010; 121:2462-2508.

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