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PAMJ Clinical Medicine

Case report

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Received: 30 Jun 2020 - **Accepted:** 24 Jul 2020 - **Published:** 14 Aug 2020

Keywords: Unique coronary artery, single coronary ostium, anomalous coronary course, unstable angina, coronary angiography, CT scan angiography

Cite this article: Zakaria Qechchar et al. A rare case of a unique single ostium coronary artery from the right coronary sinus with anomalous left circumflex course. PAMJ Clinical Medicine. 2020;3(171). 10.11604/pamj-cm.2020.3.171.24704

Available online at: https://www.clinical-medicine.panafrican-med-journal.com//content/article/3/171/full

A rare case of a unique single ostium coronary artery from the right coronary sinus with anomalous left circumflex course

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Abstract

Among congenital coronary artery anomalies (CAA), the single coronary ostium is a very rare angiographic finding. Anomalous origin of the left anterior descending artery (LAD) arising from the right coronary artery (RCA) is an extremely uncommon variant of single coronary artery. In most cases, these anomalies are incidental finding with no clinical significance; however, it can cause angina, myocardial ischemia and sudden depending on the relation of the anomalous left coronary artery to the aorta and pulmonary artery. We report the case of a 66-year-old woman with a history of hypertension, diabetes type 2 and hypercholesterolemia who presented with unstable angina associated with New York Heart Association (NYHA) II dyspnea. She underwent coronary angiography that showed both right coronary artery (RCA) and left circumflex artery (LCx) originating from the same ostium in the right sinus of Valsalva (RSV) and an ectopic left anterior descending artery (LAD) arising from the proximal part of the RCA. Gated cardiac computed tomography (CT) confirmed these findings and showed no course between the aorta and pulmonary artery, therefore, surgery was not performed. Diagnosis and management of patients with this coronary anomaly are discussed.

Introduction

The incidence of coronary artery anomalies discovered in the largest reported coronary angiography series is about 1.3% [1]. Among these anomalies, the single coronary artery (SCA) in which one coronary artery emerges from a single coronary ostium in the aorta, is a very rare angiographic finding (0.02% to 0.06%) [2] with different subtypes depending on the course of the abnormal artery. A very rare variant of the SCA is the anomalous origin of the left anterior descending artery (LAD) arising from the right coronary artery (RCA). In most cases, this anomaly is incidental finding with no clinical significance; but it can also cause angina, myocardial ischemia and sudden cardiac death, particularly if associated with a proximal course between the aorta and pulmonary trunk, due to the anomalous artery compression between the two vessels during exercise [3]. We present a case of SCA with an ectopic left anterior descending artery (LAD) arising from the proximal part of the RCA, and an anomalous origin of the left circumflex artery (LCx) from the right sinus of Valsalva (RSV).

Patient and observation

We present the case of a 66-year-old Moroccan female with a history of hypertension, diabetes type 2 and hypercholesterolemia who presented with unstable angina. The patient reported complaints of typical effort angina few months prior to presentation, which progressed to become at rest, associated with NYHA II dyspnea. Vital signs were stable. Physical exam was unremarkable. Resting electrocardiogram was normal except for a type I atrioventricular block (AVB). Trans-thoracic echocardiography findings were unremarkable with normal left ventricular dimensions and function (LVEF = 60%) and no segmental wall-motion abnormalities, right ventricle parameters and heart valves were also normal. Both high sensitive troponins´ determinations were negative. Diagnostic coronary angiography was performed with multiple unsuccessful attempts for cannulation of the left coronary artery (LCA) by left Judkins. Selective approach (Figure 1) revealed a dominant RCA originating from the RSV. However, it showed a usual coronary artery anomaly: the left circumflex artery (LCx) also emerged from the RSV and an ectopic LAD arose from the proximal part of the RCA. No significant stenosis was associated. The gated cardiac computed tomography (CT) confirmed these findings and showed a pre-pulmonic, pre-pericardial course of the ectopic LAD and an anomalous retro aortic course of the LCx. (Figure 2, Figure 3). No « at risk » inter-aorto-pulmonary course was detected and no signs of artery compression or obstruction were found at rest. Therefore, prophylactic surgery was not considered. Medical management was sufficient to

control clinical signs during hospital course. However, we considered additional diagnostic stress imaging to evaluate the patient. No symptom nor cardiac event were detected in one year follow-up.

Discussion

Isolated congenital coronary artery anomalies (CAA_s) are very rare, with a prevalence ranging from 0.2-1.3% in the literature [1]. They can be classified into anomalies of origin, course and termination [4]. The majority of CAA are considered benign and are often asymptomatic, usually discovered as incidental findings at the time of catheterization. However, some CAA are considered « at risk » and are the second most frequent cause of sudden cardiac death (SCD) among young athletes, in particular CAAs with a course between the aorta and the pulmonary artery that induce ischemia. Among these, one of the rarest anomalies is the isolated single coronary artery (SCA), that was first described in 1903 and accounts for 0.02% to 0.06% [1,2,5] in angiographic series. SCA is defined by a single isolated coronary artery coming from a single coronary ostium in the aortic root, responsible for the perfusion of the entire heart myocardium [6]. This single coronary can originate either from the left or the right sinus of valsalva (RSV). SCA anomaly is rarely isolated and usually coexist with other congenital heart diseases such as transposition of the great vessels, bicuspid aortic valve, tetralogy of Fallot, truncus arteriosus, ventricular septal defect, patent ductus arteriosus, coronary arteriovenous fistula and patent foramen ovale [7]. In our patient, no other congenital cardiac defects was associated to the coronary anomaly. The various patterns of SCA are difficult to apprehend, some authors tried to make classifications to better understand this rare anomaly. For example, in 1950, Smith described 3 different types of SCA [6], completed by Ogden and Goodyer´s classification in 1976 [8]. Finally, in 1979, Lipton *et al*. proposed a more complete classification for isolated SCA that was more practical for angiographers [9]. The anomalous coronary artery is first called “R” (Right) or “L” (Left) depending on the sinus it is emerging from. It is then designated as group I, II, or III. Finally, the letters “A,” “B,” and “P” refer to “anterior,” “between,” and “posterior” patterns, depending on the relationship between the coronary artery and the aorta and pulmonary artery (Figure 4). In our patient´s case, the LCx arose from the RSV separately from the LAD that emerged from the proximal part of the RCA, which can be classified as an isolated single R-III subtype coronary artery anomaly accordingly to Lipton *et al*. classification. Another classification was Yamanaka and Hobbs´ in 1990 [1], who modified Lipton´s classification by adding “septal” and “combined” types, designated as “S” and “C,” in order to describe the anatomical variations with more precision.

The prognosis of SCA is not well defined and depends on the anatomical subtypes. Usually, SCA with left coronary artery (LCA) arising from the RSV, especially with an interarterial course, is classified among the potentially serious anomaly [1,10], responsible of several cases of SCD in young adults during exercise in the autopsy series [10]. Sometimes, the anomaly can affect myocardial perfusion and be responsible of ischemia, heart failure or SCD. SCA group I and III are usually completely benign with an extremely rare incidence of reported SCD unlike group II that is associated with the highest reported SCD [10]. However, cases of angina pectoris and myocardial infarction (MI) have been reported with group III [11], like it was the case of our patient who presented with unstable angina. The mechanism of ischemia in patients without atherosclerosis is not well known, some authors imputate it to the angulation at the origin from the RSV, others to coronary vasospasm or compression of great vessels, particularly in the context of an intense physical exercise. Venturini *et al*. [12] reported the case of a 74-year-old woman who presented with an acute MI with a SCA arising from RSV with a single ostium bifurcating into left main coronary artery (LMCA) and a dominant RCA. The angiography showed normal coronary arteries, with no significant stenosis. This case resembles

ours because of both patients´ advanced age and the combination of a benign course of SCA with acute MI without coronary atherosclerotic lesions. As far as the diagnosis is concerned, the gold standard for the diagnosis and classification of CAA remains coronary angiography. However, exact course determination and relationships can sometimes be difficult to assess. The use of cardiac CT scan can be helpful to visualize the course of the coronary artery in a three-dimensional image, can characterize plaques and assess intraluminal narrowing with high accuracy [13]. Furthermore, coronary CT scan has an excellent spatial resolution to describe the relationship of the anomalous arteries with the great vessels cardiac chambers [14]. Other imaging modalities include cardiac magnetic resonance imaging (MRI) that is a suitable alternative to CT despite its lower spatial resolution, and transesophageal echocardiography (TEE) [15]. All in all, coronary angiography and imaging modalities are complementary when the diagnosis and therapeutic approach are not clearly established by cardiac catheterization. In fact, the course of the anomalous coronary artery and its relation to the great vessels was unknown in more than two third of cases in a Spanish angiographic serie including 23 300 coronary angiographies, which was imputable to the rare use to imaging techniques (only 2 cases, by TEE and MRI) [16]. In our case, we diagnosed the SCA anomaly by cardiac angiography and used coronary CT angiography to determine its course and its relationship with the great vessels. The patient had a pre-pulmonic, pre-pericardial course and an anomalous retro aortic course of the LCx. Therapeutic modalities are individualized depending on the patient's age, ischemic symptoms and the associated coronary lesions. Patients of more than 50 years have a very low incidence of SCD, hence, the therapeutic approach is similar to normal population with ischemic heart disease. In younger patients with a potential malignant variant, the risk SCD is greater and surgery is the treatment of choice [17]. Our patient had advanced age and her anatomic variation was considered benign, therefore, surgical intervention was not offered and she was managed with conservative medical therapy and regular follow-up.

Conclusion

Left coronary artery emerging from the right sinus of Valsalva is a rare form of single coronary artery anomaly that can be responsible of ischemia sudden cardiac death. If the anomalous artery course is not between the great vessels, this anomaly is considered as relatively benign. Coronary CT scan and coronary angiography give the diagnostic confirmation. Despite this anomaly´s rarity, increased awareness may enable early diagnosis especially in active young individuals to prevent SCD. Multi-disciplinary management involving cardiologists, interventionists and surgeons, is required to provide individualized treatment strategies based on clinical presentation and coronary anatomy, and in some cases like ours conservative medical approach is preferred.

Competing interests

The authors declare no competing interests.

Authors' contributions

All the authors contributed equally in drafting the manuscript. All the authors read and agreed to the final manuscript.

Figures

Figure 1: left anterior oblique (LAO) angiogram showing single coronary artery emerging from the right coronary sinus

Figure 2: coronary CT angiography: single coronary artery with prepulmonic left anterior descending artery and retro aortic circumflex artery with anomalous course

Figure 3: 3D CT scan angiography reconstruction presenting single coronary artery with prepulmonic left anterior descending artery and retro aortic circumflex artery with anomalous course

Figure 4: classification of angiographic types of isolated single coronary artery patterns according to Lipton *et al*. Abbreviations: R = right, L = left, RCA = right coronary artery, LCA = left coronary artery, LAD = left anterior descending coronary artery, LCx = left circumflex coronary artery, Ao = aorta, PA = main pulmonary artery, A =anterior, B =between, and P = posterior to the great vessels

References

1. Yamanaka O, Hobbs RE. Coronary artery anomalies in 126,595 patient undergoing coronary arteriography. Cathet Cardiovasc Diagn. 1990 Sep;21(1): 28-40. **PubMed**| **Google Scholar**
2. Desmet W, Vanhaecke J, Vrolix M, Van de Werf F, Piessens J, Willems J *et al*. Isolated single coronary artery: a review of 50,000 consecutive coronary angiographies. Eur Heart J. 1992;13(12): 1637-40. **Google Scholar**
3. Bartoli CR, Wead WB, Giridharan GA, Prabhu SD, Koenig SC, Dowling RD. Mechanism of myocardial ischemia with an anomalous left coronary artery from the right sinus of Valsalva. J Thorac Cardiovasc Surg. 2012 Aug;144(2): 402-8. **PubMed**| **Google Scholar**
4. Wilkins CE, Betancourt B, Mathur VS. Coronary artery anomalies: a review of more than 10,000 patients from the Clayton cardiovascular laboratories. Tex Heart Inst J. 1988;15(3): 166-73. **PubMed**| **Google Scholar**
5. Flessas D, Mamarelis I, Maniatis V, Souretis G, Laschos N, Kotoulas C *et al*. An unusual pattern of three major components of the cardiovascular system: multimodality imaging and review of the literature. J Cardiothorac Surg. 2013 Apr 4;8: 61 **PubMed**| **Google Scholar**
6. Smith JC. Review of single coronary artery with report of 2 cases. Circulation. 1950 May;1(5): 1168-75. **PubMed**| **Google Scholar**
7. 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 Jan 1;5(1): 22-30. **PubMed**| **Google Scholar**
8. Ogden JA, Goodyer AV. Patterns of distribution of the single coronary artery. Yale J Biol Med. 1970 Aug;43(1): 11-21. **PubMed**| **Google Scholar**
9. Martin J Lipton, William H Barry, Ivo Obrez, James F Silverman, Lewis Wexler. Isolated single coronary artery: diagnosis, angiographic classification, and clinical significance. Radiology. 1979;130(1): 39-47. **PubMed**| **Google Scholar**
10. Cheitlin MD, De Castro CM, McAllister HA. Sudden death as a complication of anomalous left coronary origin from the anterior sinus of valsalva: a not so minor congenital anomaly. Circulation. 1974 Oct;50(4): 780-7. **PubMed**| **Google Scholar**
11. Murphy DA, Roy DL, Sohal M. Anomalous origin of left main coronary artery from anterior sinus of valsalva with myocardial infarction. J Thorac Cardiovasc Surg. 1978 Feb;75(2): 282-5. **PubMed**| **Google Scholar**
12. Venturini E, Magni L. Single coronary artery from the right sinus of Valsalva. Heart Int. 2011;6(1): e5. **PubMed**| **Google Scholar**

13. Korosoglou G, Mueller D, Lehrke S. Quantitative assessment of stenosis severity and atherosclerotic plaque composition using 256-slice computed tomography. European Radiology. 2010;20(8): 1841-1850. **PubMed**| **Google Scholar**
14. Aldana-Sepulveda N, Restrepo CS, Kimura-Hayama E. Single coronary artery: spectrum of imaging findings with multidetector CT. J Cardiovasc Comput Tomogr. 2013;7(6): 391-9. **PubMed**| **Google Scholar**
15. Zemanek D, Veselka J, Kautznerova D, Tesar D. The anomalous origin of the left coronary artery from the right aortic sinus: is the coronary angiography still a "gold standard"?. Int J Cardiovasc Imaging. 2006 Apr;22(2): 127-33. **PubMed**| **Google Scholar**
16. Barriales-Villa R, Morís C, Sanmartín JC, Fernández E, Pajín F, Ruiz Nodar JM. Registro de anomalías congénitas de las arterias coronarias con origen en el seno de Valsalva contralateral en 13 hospitales españoles (RACES). Rev Esp Cardiol. 2006 Jun;59(6): 620-3. **PubMed**| **Google Scholar**
17. Barriales-Villa R, Morís de la Tassa C. Anomalías congénitas de las arterias coronarias con origen en el seno de Valsalva contralateral: qué actitud se debe seguir. Rev Esp Cardiol. 2006 Apr;59(4): 360-70. **PubMed**| **Google Scholar**

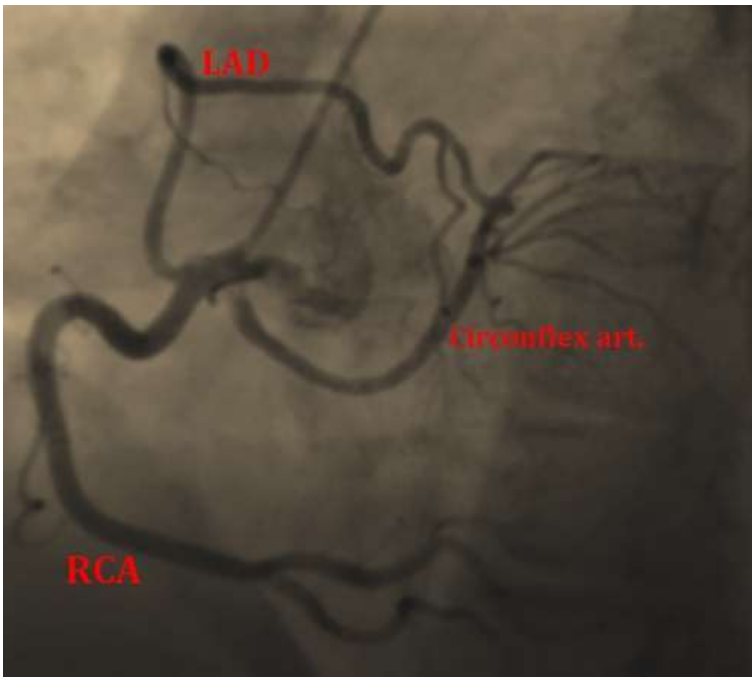


Figure 1: left anterior oblique (LAO) angiogram showing single coronary artery emerging from the right coronary sinus

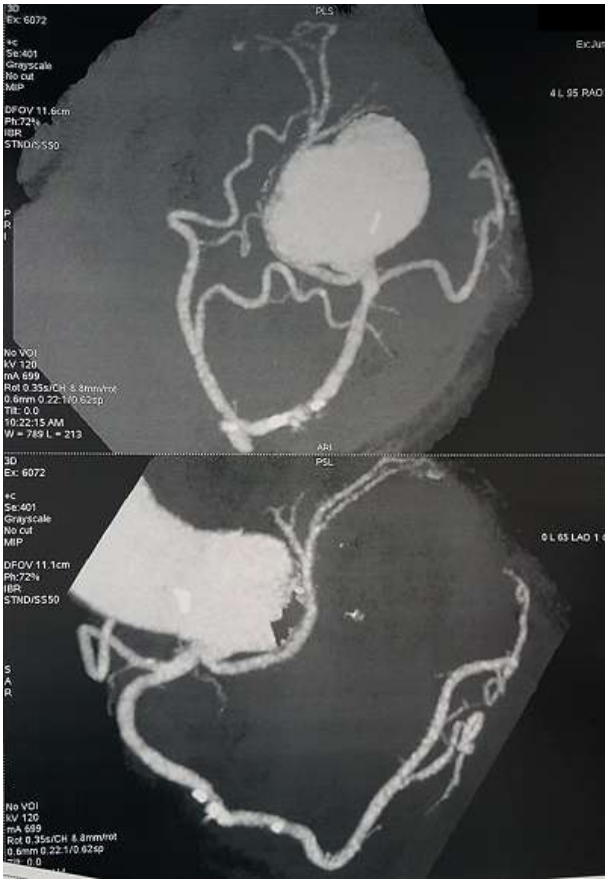


Figure 2: coronary CT angiography: single coronary artery with prepulmonic left anterior descending artery and retro aortic circumflex artery with anomalous course

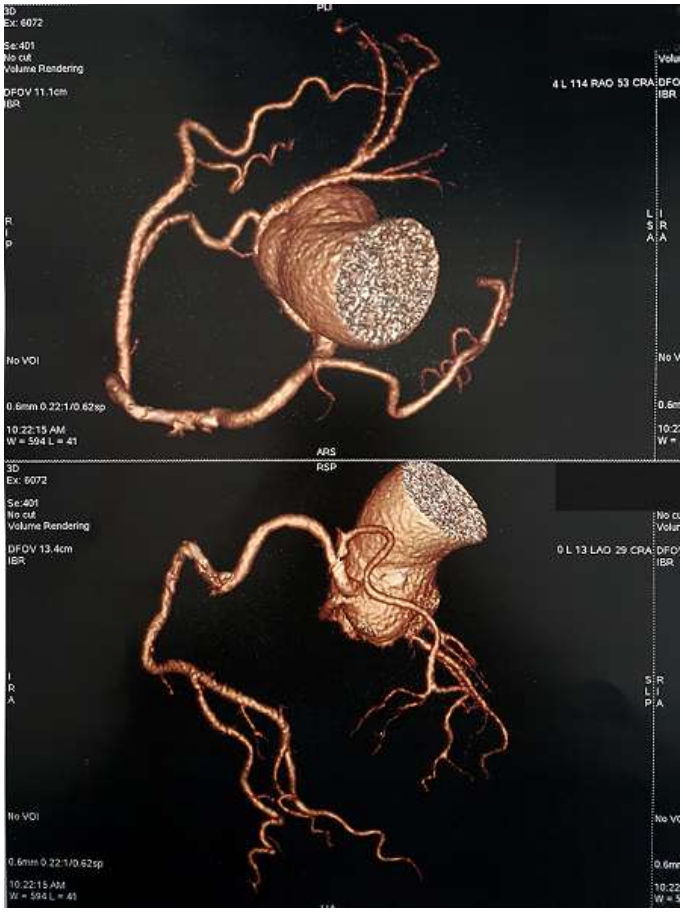
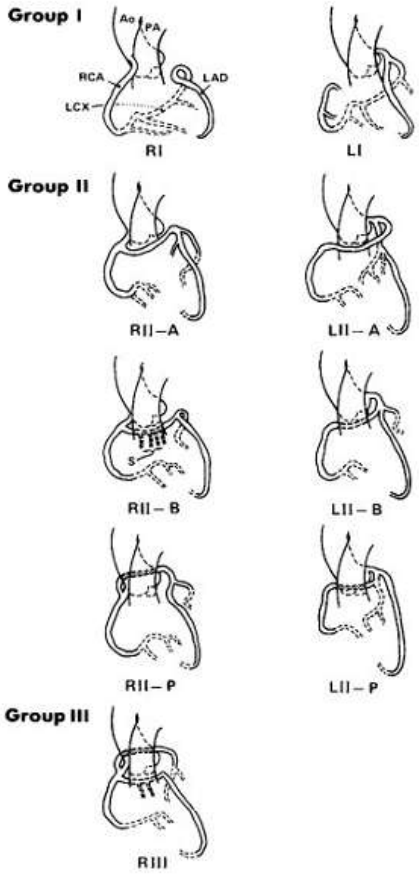


Figure 3: 3D CT scan angiography reconstruction presenting single coronary artery with prepulmonic left anterior descending artery and retro aortic circumflex artery with anomalous course



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