CASE REPORT

Four Coronary Arteries Separately Originating from the Right Sinus of Valsalva

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Abstract:
An 82-year-old woman was admitted to our hospital with heart failure. Coronary angiography revealed one anatomically normal right coronary artery and three left coronary arteries (LCA-1, LSA-2, and LSA-3) separately originating from the right sinus of Valsalva, comprising multiple atherosclerotic lesions. LCA-1 became the obtuse marginal branch after branching off into the septal branches. LCA-2 was the main circumflex artery with an obstructive lesion. LCA-3 corresponded to the distal part of the anterior descending branch. The patient died 14 days after hospitalization. We describe the rarity of quadriostial origin, the unusual course, and the unusual branching of the coronary arteries.

Key words: quadriostial origin, coronary artery disease, coronary artery anomalies, dual left anterior descending arteries, twin circumflex arteries

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Introduction

Coronary artery anomalies, including an abnormal origin, course, and distribution of the epicardial arteries, are reported in 0.9-1.3% of patients undergoing coronary angiography (1-3).

We herein report an extremely rare case of a patient with four coronary arteries separately originating from the right sinus of Valsalva (RSV). We also describe the unusual course and branching of the coronary arteries as well as the rarity of quadriostial origin.

Case Report

An 82-year-old woman with a medical history of hypertension and pyelonephritis presented to our hospital with chest discomfort that had developed within 24 hours prior to hospitalization. The patient reported recurrent chest pain on exertion for approximately six months.

On the day of admission, her blood pressure was 102/84 mmHg. Electrocardiography (ECG) revealed atrial fibrillation with a heart rate of 148/min and ST segment depression in leads I, II, aVF, and V1-6 (Fig. 1). Plane chest radiography revealed heart enlargement and pulmonary congestion (Fig. 2). Echocardiography revealed extensive reduction of inferior and lateral wall motion with left ventricular ejection fraction of 40% and moderate mitral regurgitation. Blood examinations yielded a white blood cell count of 5,540 mm³, creatinine level of 1.12 mg/dL, aspartate transaminase level of 34 IU/L, alanine aminotransferase level of 15 IU/L, lactate dehydrogenase level of 214 IU/L, and creatine kinase level of 224 IU/L. Based on the clinical data, the patient was diagnosed with congestive heart failure accompanied by ischemic heart disease and atrial fibrillation.

Coronary angiography, performed on the day of admission, revealed four coronary arteries with multiple atherosclerotic lesions. The right coronary artery (RCA) was anatomically normal, whereas the three left coronary arteries (LCA-1, LSA-2, and LSA-3) originated from separate ostia in the RSV.

The RCA had significant stenotic lesions in the middle and distal segments (Fig. 3A). One of the three LCAs (LCA-1) arose from the immediate left side of the RCA...
Figure 1. An ECG on admission showing atrial fibrillation with a heart rate of 148/min and ST segment depression in leads I, II, aVf, and V1-6.

Figure 2. Plane chest radiography on admission showing heart enlargement and pulmonary congestion.

ostium (Fig. 3B). The initial course of this artery in the left anterior oblique projection was leftward, slightly downward, across the aortic root, branching off into the septal branches, and it became the obtuse marginal branch or the ramus intermedius artery. Thus, the LCA-1 artery formed a caudal anterior loop and may have followed a trans-septal course. The LCA-2 originated from the left upper side of the RSV and corresponded to the main left circumflex artery (LCX) with an obstructive lesion in the middle segment (Fig. 3C). The distal segment of the LCA-2 was supplied through collateral flow from the LCA-1 artery (Fig. 3B right panel, blue arrowhead). The LCA-3 originated from the right upper side of the RSV and was divided into the distal part of the left anterior descending artery (LAD) and the diagonal branch, with severe stenosis observed at the bifurcation point (Fig. 3D). During selective coronary angiograms of each artery, when the contrast medium refluxed out of the ostium into the aorta, faint angiographic images of non-engaged coronary arteries were obtained (Fig. 4), demonstrating the spatial position relationships of the individual
have appeared on a CT scan. May have followed a trans-septal course, which might not have reached the left atrioventricular groove. The LCA-1 was reached through autopsies following sudden death or coronary angiography (CAG) for ischemic heart disease in middle-aged and elderly patients. In one of the six reported cases, the presence of two ostia was observed in the RSV (Table). Although this is a congenital anomaly, the diagnoses were reached through autopsies following sudden death or coronary angiography (CAG) for ischemic heart disease in middle-aged and elderly patients. In one of the six reported cases, the presence of two ostia was observed in the RSV (Table). Although this is a congenital anomaly, the diagnoses were reached through autopsies following sudden death or coronary angiography (CAG) for ischemic heart disease in middle-aged and elderly patients. In one of the six reported cases, the presence of two ostia was observed in the RSV (Table). Although this is a congenital anomaly, the diagnoses were reached through autopsies following sudden death or coronary angiography (CAG) for ischemic heart disease in middle-aged and elderly patients. In one of the six reported cases, the presence of two ostia was observed in the RSV (Table). Although this is a congenital anomaly, the diagnoses were reached through autopsies following sudden death or coronary angiography (CAG) for ischemic heart disease in middle-aged and elderly patients. In one of the six reported cases, the presence of two ostia was observed in the RSV. In all six cases, all four coronary arteries originated from the RSV (Fig. 5). These two LCAs had followed pre-pulmonary courses. Volume-rendered images revealed that the distribution of these two LCAs was consistent with that of the LCA-2 and the diagonal branch of the LCA-3 shown in the coronary angiogram. It was demonstrated that the LCA-2 had reached the left atrioventricular groove. The LCA-1 may have followed a trans-septal course, which might not have appeared on a CT scan.

Discussion

The spectrum of coronary anomalies is wide and includes anomalous coronary artery originating from the opposite sinus (ACAOS), single coronary artery, and coronary fistulae. The clinical significance of these coronary artery anomalies varies from benign to life-threatening.

The quadriostial origin of coronary arteries is an extremely rare anomaly (4-8). To our knowledge, only six such cases have been reported in the literature (Table). Although this is a congenital anomaly, the diagnoses were reached through autopsies following sudden death or coronary angiography (CAG) for ischemic heart disease in middle-aged and elderly patients. In one of the six reported cases, the presence of two ostia was observed in the RSV and the left sinus of Valsalva (one each). In the remaining five cases, all four coronary arteries originated from the RSV. In all six cases, as well as the present case, the main RCA arose from the normal site of the RSV and followed a normal course. Besides the main RCA, the LAD, and the
Figure 4. Faint images of the non-engaged coronary arteries during selective angiography. With the contrast medium refluxed out of the ostium into the aorta, some ostia and initial courses of other non-engaged coronary arteries are shown. Red, black, blue, and yellow arrowheads show the RCA, LCA-1, LSA-2, and LSA-3, respectively. (A) The coronary angiogram obtained in a left anterior oblique projection shows the selectively imaged LCA-1 and the secondarily imaged RCA. (B) The coronary angiogram obtained in a caudal projection shows the selectively imaged LCA-1 and the secondarily imaged LCA-2. (C-E) C, D, and E in Fig. 4 are still images taken from the same video obtained by one injection of contrast medium. The catheter is selectively engaged in LCA-2 in a left anterior oblique projection. The coronary angiograms show the secondarily imaged LCA-1, LSA-3, and RCA. (F) The coronary angiogram obtained in a cranial projection shows the selectively imaged LCA-3 and the secondarily imaged LCA-1 and RCA. LCA: left coronary artery, RCA: right coronary artery.

Figure 5. Schematic representation of the four coronary arteries in this case.

main LCX, the patients had separately originating conus arteries, sinus node arteries, and/or ramus intermedius arteries. In the present case, the RCA and three LCAs originated from the RSV. ACAOS is rarely observed, with a reported rate of 0.15-3.9% (studies using catheter angiography) and 0.35-2.1% (studies using CT) (9, 10). Anomalous arteries
Figure 6. Non-electrocardiography-gated plane computed tomography (CT) scan performed two years prior to hospitalization. Plane CT images show the RCA (left upper panel, red arrow) and two of the three LCAs, which follow a pre-pulmonic course (right upper panel, yellow and blue arrows). Volume-rendered images show that the distributions of these two LCAs were consistent with those of LCA-2 and the diagonal branch of LCA-3 (right and left lower panel). Red, blue, and yellow colors represent the RCA, LCA-2, and LCA-3, respectively. LCA: left coronary artery, RCA: right coronary artery.

originating from the RSV take one of the following courses in relation to the aorta and pulmonary trunk: pre-pulmonic, retro-aortic, inter-arterial (between the aorta and pulmonary artery trunk), and trans-septal (10, 11). Opolski et al. (9) demonstrated that anomalous left main coronary arteries and the LADs originating from the RSV most commonly follow an intra-septal or anterior course. They also indicated that LCXs originating from the RSV most commonly follow a retro-aortic course. In the present case, the LCA-3 that perfused the distal part of the anterior descending branch followed an anterior (pre-pulmonic) course. The LCA-1 and the LCA-2 that perfused the circumflex region followed a trans-septal and a pre-pulmonic course, respectively. The initial courses of the three left coronary arteries in the present case were unusual compared with the previously reported cases.

ACAOS with an inter-arterial course, particularly with the intramural segment characterized by a coronary artery running within the aortic wall, is associated with a high risk of sudden cardiac death. In patients with such ACAOS, coronary artery surgery is often recommended (10-12).

Although the diagnosis of other benign ACAOSs is often incidental, certain cases may lead to ischemic heart disease. Click et al. (13) reported that an aberrant LCX originating from the RSV exhibited a greater degree of stenosis than an LCX originating from the left main coronary artery (13). However, these findings remain controversial (14).

In addition to the rarity of the quadriostial origin and unusual course of the coronary arteries in the present case, the presence of unusual branching was observed. The LCA-1 became the obtuse marginal branch or the ramus intermedius artery after branching off into the septal branch, the LCA-2 became the main LCX, and the LCA-3 became the LAD. These arteries formed “dual LAD” (15-17) and “twin LCX” (18-21).

Dual LAD is another rare anomaly, characterized by a short LAD and a long LAD (15-17). The short LAD terminates high in the anterior interventricular groove, while the long LAD follows a variable course outside of the anterior interventricular groove and returns to the interventricular
The authors state that they have no Conflict of Interest (COI).

### References

1. Yamanaka O, Hobbs RE. Coronary artery anomalies in 126,595 patients undergoing coronary arteriography. Cathet Cardiovasc Diagn 21: 28-40, 1990.
2. Türkmen S, Cagliyan CE, Poyraz F, et al. Coronary arterial anomalies in a large group of patients undergoing coronary angiography in southeast Turkey. Folia Morphol (Warsz) 72: 123-127, 2013.
3. Yuksel S, Mercir M, Soylu K, et al. The primary anomalies of coronary artery origin and course: a coronary angiographic analysis of 16,573 patients. Exp Clin Cardiol 18: 121-123, 2013.
4. Virmani R, Chan PK, Rogan K, Riddick L. Anomalous origin of four coronary ostia from the right sinus of Valsalva. Am J Cardiol 63: 760-761, 1989.
5. Beach L, Burke A, Chute D, Virmani R. Anomalous origin of 4 coronary ostia from the right sinus of Valsalva in a patient with hypertrophic cardiomyopathy. Arch Pathol Lab Med 125: 1489-1490, 2001.
6. Kapoor A, Kumar S, Sinha N. Coronary angioplasty in a case of quadriostial origin of coronary arteries from right aortic sinus. Indian Heart J 56: 143-146, 2004.
7. Mohandas B, Singh B, Mehta JL, Sachdeva R. A man with four coronary ostia. Eur Heart J 28: 1357, 2007.
8. Jarwala P, Bhaita H, Kesava Rao RC, Padma Kumar EA. Quadriostial origin of 4 coronary arteries from the right coronary sinus of Valsalva: rare anomaly. JACC Cardiovasc Interv 11: 503-504, 2018.
9. Opolski MP, Pregowski J, Kruk M, et al. Prevalence and characteristics of coronary anomalies originating from the opposite sinus of Valsalva in 8,522 patients referred for coronary computed tomography angiography. Am J Cardiol 111: 1361-1367, 2013.
10. Agarwal PP, Dennie C, Pena E, et al. Anomalous coronary arteries that need intervention: review of pre- and postoperative imaging appearances. Radiographics 37: 740-757, 2017.
11. Eckart RE, Scoville SL, Campbell CL, et al. Sudden death in young adults: a 25-year review of autopsies in military recruits. Ann Intern Med 141: 829-834, 2004.
12. Krasuski RA, Magyar D, Hart S, et al. Long-term outcome and impact of surgery on adults with coronary arteries originating from the opposite coronary cusp. Circulation 123: 154-162, 2011.
13. Click RL, Holmes DR Jr, Vlietstra RE, Kosinski AS, Kronmal RA. Anomalous coronary arteries: location, degree of atherosclerosis and effect on survival-a report from the Coronary Artery Surgery Study. J Am Coll Cardiol 13: 531-537, 1989.
14. Mohsen GA, Mohsin KG, Fosberg M, Miller E, Taniuchi M, Klein AJ. Anomalous left circumflex artery from the right coronary cusp: a benign variant? J Invasive Cardiol 25: 284-287, 2013.

### Table. Review of the Previously Reported Cases with Quadriostial Origin of Coronary Arteries.

| Reference | Patient age/sex remarks | Ostium | RCA system | LAD system | LCX system |
|-----------|-------------------------|--------|------------|------------|------------|
| 4) 64/male | Sudden death Autopsy    | Four in the RSV | normal course → trans-septal | normal course → trans-septal | normal course → trans-septal |
| 62/male Traffic accident Autopsy | Four in the RSV | normal course | normal course | normal course | normal course |
| 5) 49/male | HCM Heart failure CAG   | Four in the RSV | normal course | normal course | normal course |
| 60/male Angina CAG | Four in the RSV | normal course | normal course | normal course | normal course |
| 7) 52/male Dyspnea CAG | Two each in the RSV and LSV | normal course | normal course | normal course | normal course |
| 8) 56/female | AMI Heart failure CAG   | Four in the RSV | normal course | normal course | normal course |

HCM: hypertrophic cardiomyopathy, CAG: coronary angiography. AMI: acute myocardial infarction. RSV: right sinus of Valsalva, LSV: left sinus of Valsalva, RCA: right coronary artery, LAD: left anterior descending artery, LCX: left circumflex artery, LCA: left coronary artery.

The black dot indicates that this is a coronary artery originating from the Valsalva sinus and not a branch.

groove distally. Twin LCX is another rarely reported anomaly. The main LCX usually branches off from the left main coronary artery and the aberrant LCX originating from the RSV. Aberrant LCXs most commonly follow a retro-aortic or inter-aortic course (18-21).

It has been reported that percutaneous coronary intervention for the treatment of anomalous coronary arteries, which was not applicable in the present case, may occasionally be accompanied by procedural challenges (21-23).

In conclusion, we described an extremely rare case of ischemic heart failure with four coronary arteries separately originating from the RSV. Apart from the incidence and the clinical significance of the anomalous form, the correct identification of the origin, course, and branching of the anomalous coronary arteries is clinically important.

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1. Yamanaka O, Hobbs RE. Coronary artery anomalies in 126,595 patients undergoing coronary arteriography. Cathet Cardiovasc Diagn 21: 28-40, 1990.
2. Türkmen S, Cagliyan CE, Poyraz F, et al. Coronary arterial anomalies in a large group of patients undergoing coronary angiography in southeast Turkey. Folia Morphol (Warsz) 72: 123-127, 2013.
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5. Beach L, Burke A, Chute D, Virmani R. Anomalous origin of 4 coronary ostia from the right sinus of Valsalva in a patient with hypertrophic cardiomyopathy. Arch Pathol Lab Med 125: 1489-1490, 2001.
6. Kapoor A, Kumar S, Sinha N. Coronary angioplasty in a case of quadriostial origin of coronary arteries from right aortic sinus. Indian Heart J 56: 143-146, 2004.
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8. Jarwala P, Bhaita H, Kesava Rao RC, Padma Kumar EA. Quadriostial origin of 4 coronary arteries from the right coronary sinus of Valsalva: rare anomaly. JACC Cardiovasc Interv 11: 503-504, 2018.
9. Opolski MP, Pregowski J, Kruk M, et al. Prevalence and characteristics of coronary anomalies originating from the opposite sinus of Valsalva in 8,522 patients referred for coronary computed tomography angiography. Am J Cardiol 111: 1361-1367, 2013.
10. Agarwal PP, Dennie C, Pena E, et al. Anomalous coronary arteries that need intervention: review of pre- and postoperative imaging appearances. Radiographics 37: 740-757, 2017.
11. Eckart RE, Scoville SL, Campbell CL, et al. Sudden death in young adults: a 25-year review of autopsies in military recruits. Ann Intern Med 141: 829-834, 2004.
12. Krasuski RA, Magyar D, Hart S, et al. Long-term outcome and impact of surgery on adults with coronary arteries originating from the opposite coronary cusp. Circulation 123: 154-162, 2011.
13. Click RL, Holmes DR Jr, Vlietstra RE, Kosinski AS, Kronmal RA. Anomalous coronary arteries: location, degree of atherosclerosis and effect on survival-a report from the Coronary Artery Surgery Study. J Am Coll Cardiol 13: 531-537, 1989.
14. Mohsen GA, Mohsin KG, Fosberg M, Miller E, Taniuchi M, Klein AJ. Anomalous left circumflex artery from the right coronary cusp: a benign variant? J Invasive Cardiol 25: 284-287, 2013.
15. Spindola-Franco H, Grose R, Solomon N. Dual left anterior descending coronary artery: angiographic description of important variants and surgical implications. Am Heart J 105: 445-455, 1983.

16. Tuncer C, Bayraliev T, Yilmaz R, Gokce M, Eryonucu B, Koroglu S. Origin and distribution anomalies of the left anterior descending artery in 70,850 adult patients: multicenter data collection. Catheter Cardiovasc Interv 68: 574-585, 2006.

17. Celik T, Bozlar U, Ozturk C, et al. A new anomaly of the left anterior descending artery: type X dual LAD. Indian Heart J 67 (Suppl 3): S14-S17, 2015.

18. Karabay KO, Uysal E, Buturta B, Vural M. A case of twin circumflex arteries associated with acute myocardial infarction. Arch Turk Soc Cardiol 38: 496-498, 2010.

19. Andreou AY, Theodorou S, Makrides C, Avraamides PC. Twin left circumflex arteries in a patient undergoing aortic valve replacement. Eur Rev Med Pharmacol Sci 18: 71-73, 2014.

20. Otlu YO, Bayramolu A, Hidayet Ş, Ermiş N. Transradial percutaneous coronary intervention in a patient with a rare coronary anomaly: twin circumflex arteries. Acta Cardiol Sin 31: 72-74, 2015.

21. Sinha SK, Mishra V, Abdali N, et al. Primary percutaneous coronary intervention angioplasty of occluded twin circumflex coronary artery in a patient of acute inferior wall myocardial infarction: a rare anomaly. Cardiol Res 8: 52-56, 2017.

22. Oral D, Berkalp B, Pamir G, Omürli K, Erol C. Significance of dual left anterior descending coronary artery in interventional cardiology. A case report. Angiology 47: 825-829, 1996.

23. Masuda N, Matsukage T, Ikari Y. Successful transradial intervention for two lesions with dual anomalous origins of coronary arteries. J Invasive Cardiol 23: E117-E120, 2011.