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【 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 mm^3, 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

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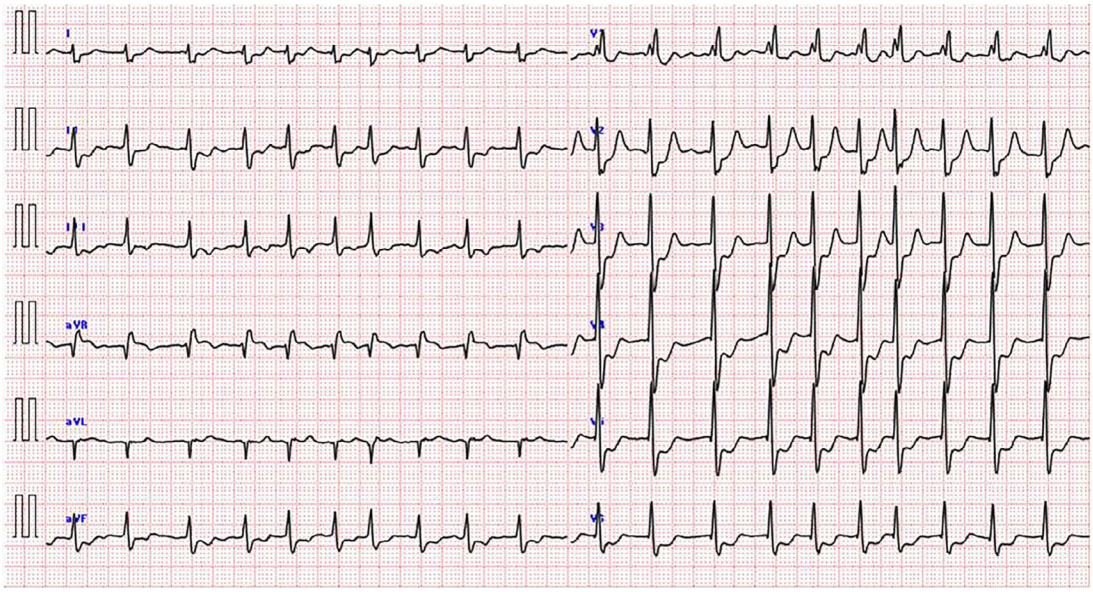

Figure 1. **An ECG on admission showing atrial fibrillation with a heart rate of 148/min and ST segment depression in leads I, II, aVf, and V1-6.**

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

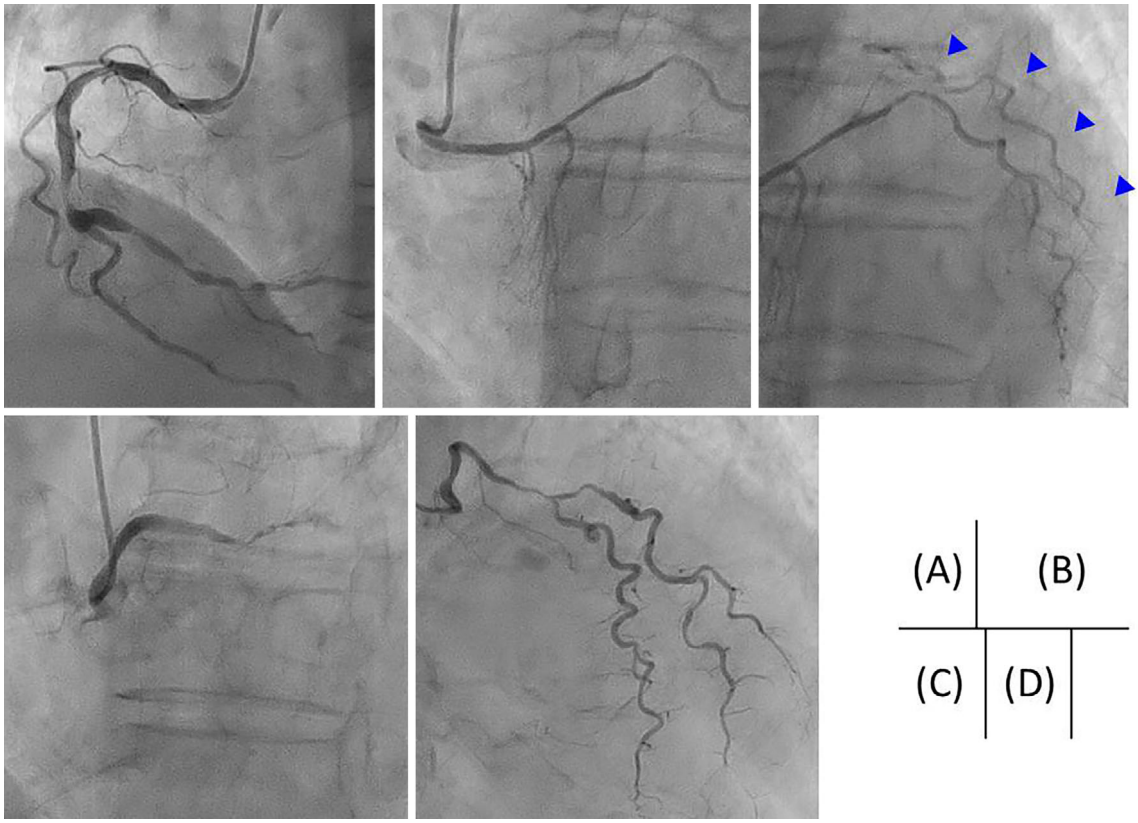

Figure 3. **Selective coronary angiograms of four coronary arteries separately originating from the RSV. (A) The RCA follows a normal course with severe stenoses in the middle and distal portions. (B) One left coronary artery arose from the immediate left side of the RCA ostium (LCA-1). The right panel shows the distal segment of this artery, supplying the distal portion of another LCA (LCA-2, blue arrowhead) through collateral flow. (C) The selective angiogram of the LCA-2, originating from the left upper side of the RSV, shows the obstructive lesion in the middle portion. (D) The third left coronary artery (LCA-3) arose from the right upper side of the RSV, supplying the distal portion of the left anterior descending artery and the diagonal branch showing severe stenosis at the bifurcation point. LCA: left coronary artery, RCA: right coronary artery, RSV: right sinus of Valsalva**

ostia. Fig. 5 is a schematic representation of the four coronary arteries.

Due to the deterioration of the patient's physical state, particularly her renal dysfunction, it was not possible to perform percutaneous coronary intervention or coronary computed tomography (CT) angiography. The patient ultimately died due to exacerbation of her heart failure 14 days after hospitalization.

Non-ECG-gated plane CT had been performed two years prior to the present hospitalization during an examination for the diagnosis of pyelonephritis. The examination revealed the RCA and two of the three LCAs originating from the RSV (Fig. 6). These two LCAs had followed prepulmonic 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

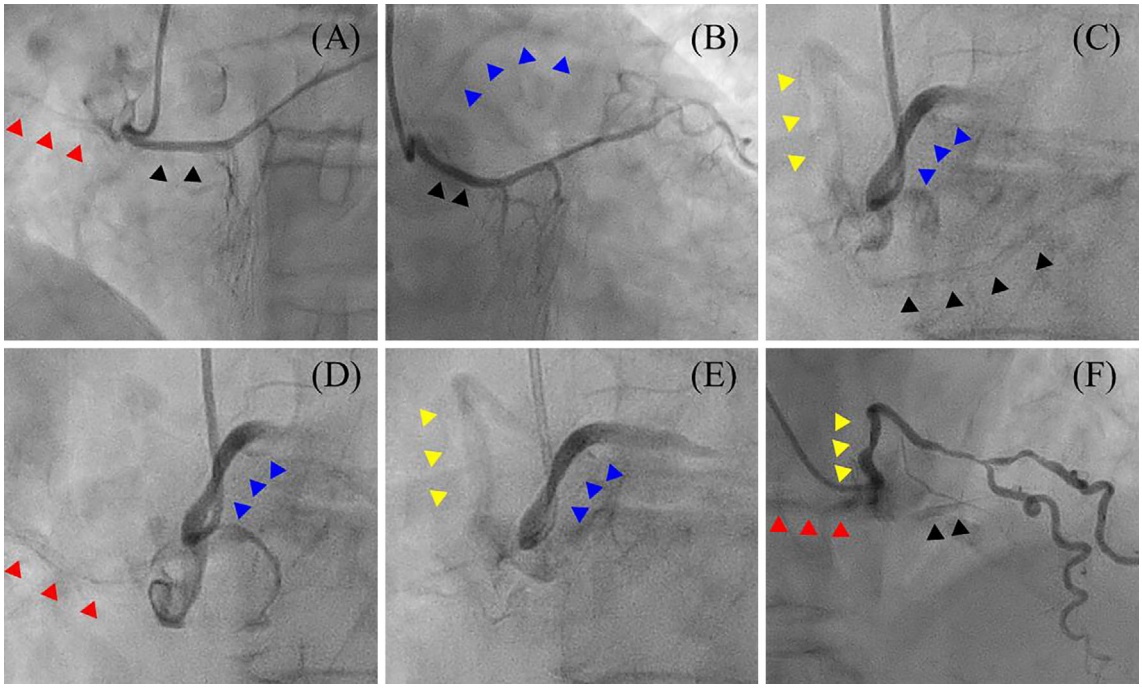

Figure 4. **Faint images of the non-engaged coronary arteries during selective angiography. With the contrast medium refluxed out of the ostium into the aorta, some ostia and initial courses of other non-engaged coronary arteries are shown. Red, black, blue, and yellow arrowheads show the RCA, LCA-1, 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**

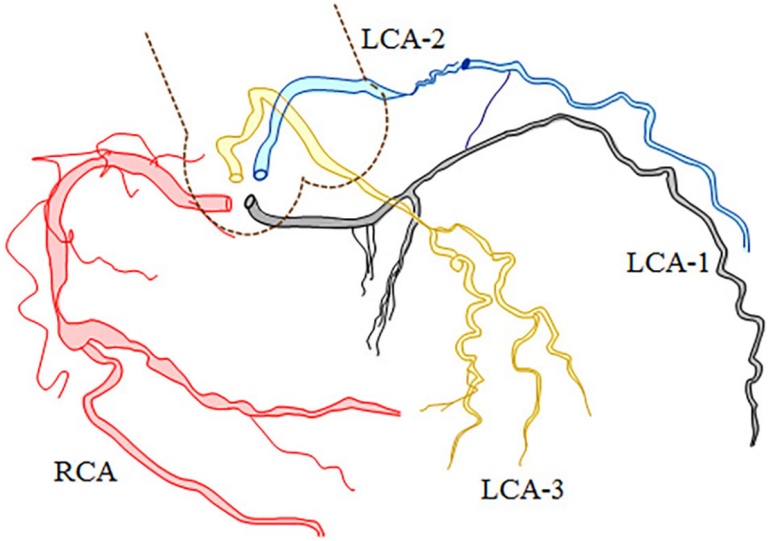

Figure 5. **Schematic representation of the four coronary arteries in this case.**

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

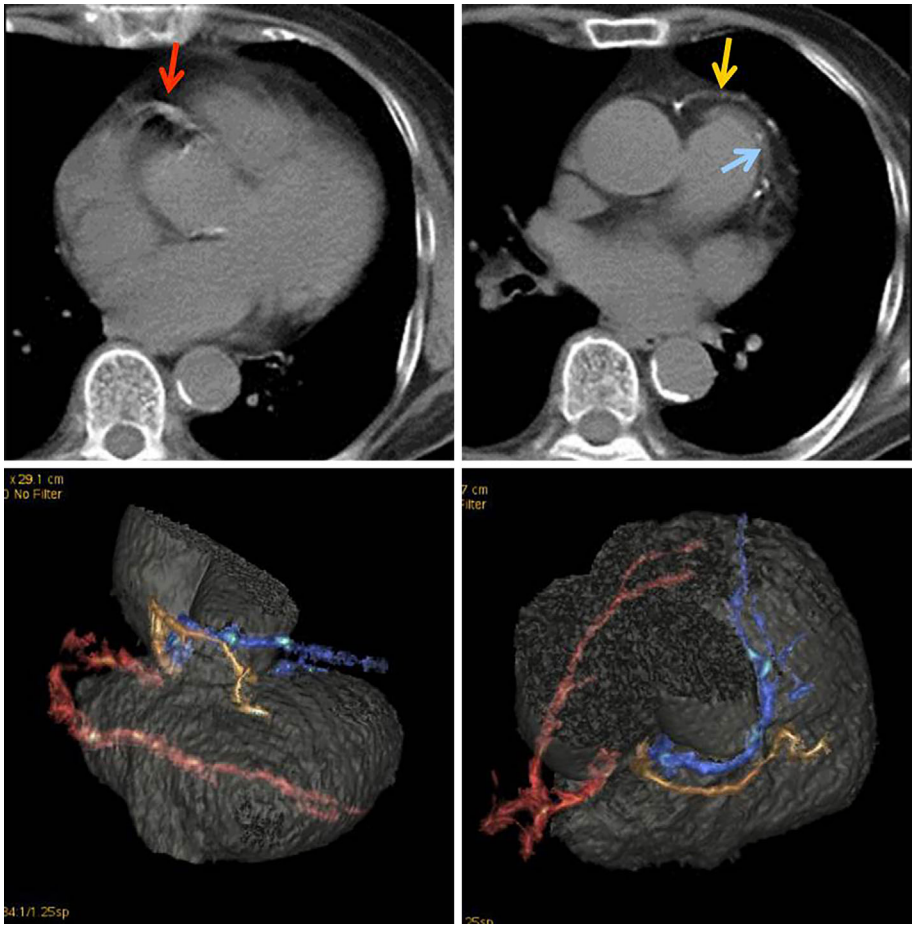

Figure 6. **Non-electrocardiography-gated plane computed tomography (CT) scan performed two years prior to hospitalization. Plane CT images show the RCA (left upper panel, red arrow) and two of the three LCAs, which follow a pre-pulmonic course (right upper panel, yellow and blue arrows). Volume-rendered 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

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	
			main RCA	Secondary RCA	LAD	Short LAD	Main LCX	Secondary LCX
4)	64/male Sudden death Autopsy	Four in the RSV	•normal course	•conus artery	•trans-septal course	-	•retroaortic course	-
	62/male Traffic accident Autopsy	Four in the RSV	•normal course →LCX	•conus artery •sinoatrial node branch	•trans-septal course	-	(from RCA)	-
5)	49/male HCM Sudden death Autopsy	Four in the RSV	•normal course	-	•trans-septal course	-	•retroaortic course	•ramus artery prepulmonic course
6)	60/male Angina CAG	Four in the RSV	•normal course	•conus artery	•trans-septal course	-	•retroaortic course	-
7)	52/male Dyspnea CAG	Two each in the RSV and LSV	•normal course (from RSV)	-	•normal course (from LSV)	-	•Inter-arterial (from RSV)	•ramus artery (from LSV)
8)	56/female AMI CAG	Four in the RSV	•normal course	-	•prepulmonic course	-	•retroaortic course	•ramus artery (trans-septal course?)
our report	82/female Heart failure CAG	Four in the RSV	•normal course	-	•LCA-3 prepulmonic course	•LCA-1 trans-septal course →secondary LCX	•LCA-2 prepulmonic course	(from LCA-1)

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.

The authors state that they have no Conflict of Interest (COI).

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