The right coronary artery originating from the distal circumflex artery

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Abstract
We report a case of anomalous origin of the right coronary artery in a 58-year-old male patient. The patient had a single coronary ostium originating from the left sinus valsalva. Right coronary artery was originating from the distal circumflex artery. There was no other cardiac anomaly in association with this rare case. Our patient had only severe atherosclerosis in the proximal left anterior descending artery with normal coronary flow in the circumflex artery. Generally, cases with single coronary ostium are considered to be benign. However, these patients experience the symptoms of coronary artery disease more critically because of dependence on one coronary artery.

Key words: coronary angiography, cardiovascular abnormalities, coronary vessel anomalies

Introduction
A single coronary artery (SCA) occurring in isolation, without being associated with other structural heart disease is very rarely reported in medical literature. Its occurrence in less than 0.02 to 0.06 % of the general population [1]. Classifications have been determined by different anatomical variations depend on either angiographically or necropsy findings [2]. We hereby present a case of a patient with one vessel disease and all coronary arteries originating from a single ostium in the left sinus of Valsalva with an anomalous course of the right coronary artery (RCA) originating as a branch of the distal circumflex artery (CX).
Case report

A 58-year-old man complaining of atypical chest discomfort with no family history of coronary artery disease (CAD) was admitted to İstinye University Liv Hospital. The associated risk factors included smoking and hypercholesterolemia. Cardiac auscultation and peripheral pulses were normal. The baseline electrocardiogram was normal. An echocardiogram revealed normal atroventricular morphology with no regional wall motion abnormality. Serial cardiac enzymes were normal. The treadmill test was positive for inducible myocardial ischemia. He underwent coronary angiography whereby RCA could not be selectively cannulated. A non-selective injection revealed no coronary artery originating from the right sinus valsalva. The selective injection of the left coronary sinus revealed only left main coronary artery (LMCA) (Figure 1). After a short course, LMCA was divided into two branches: left anterior descending artery (LAD) and CX. CX was the dominant vessel. CX was located in the normal anatomical region and there was no lesion in the CX. In the distal part of CX, a lateral branch was observed along the RCA track (Figure 2). This side branch was normal along the posterior sulcus atrioventricularis to the level of RCA. There was no other cardiac anomalies. A critical lesion was detected in proximal (ostial) LAD. Due to being not suitable for percutaneous coronary intervention (PCI), our patient was referred for surgery. During surgery, our patient was operated under cardiopulmonary bypass. His heart was arrested with intermittent antegrade cold blood cardioplegia. Distal anastomosis to LAD was performed by using left internal mammary artery insitu. The aortic cross-clamping time was 57 minutes and extracorporeal circulation was 70 minutes. Early postoperative course was uneventful. Our patient died in late postoperative course due to unknown reason of extracardiac manifestation.

Discussion

There are many factors in the embryological development of coronary vessels. These factors include chemotactic agents, adhesion molecules and multiple growth factors. Abnormalities of these signaling pathways may be responsible for coronary artery anomalies [3]. In our case, RCA ostium agenesis was present. There was a SCA originating from a single ostium in the left sinus of Valsalva.

The classification of SCA determined by Lipton et al. categorizes patterns according to the site of origin and anatomical distribution [1, 4-7]. Our patient was classified as L-1 subtype of SCA in accordance with the modified Lipton et al. classification and an abnormal RCA originating from the distal left circumflex artery. A single coronary ostium with RCA originating from the distal left circumflex artery is a very rare anomaly. In literature review, there are few patients similar to our case [2,3,5,8-31] (Table 1). There were five cases reported in Turkey.

Table 1Summary of characteristics of cases with right coronary artery (RCA) originating from distal circumflex artery (CX).

| Author/year            | Age/sex | Presenting symptom | Angiography | Associated conditions | Treatment       |
|------------------------|---------|--------------------|-------------|-----------------------|-----------------|
| Tavernarakis et al.     | 57/M    | TCP                | LAD lesion  | None                  | NA              |
| Sheth et al.            | 60/M    | ATCP               | No lesion   | None                  | None            |
| Vrolix et al.           | 51/M    | TCP                | CX lesion   | None                  | CABG            |
| Shammas et al.          | 44/F    | Chest pain         | No lesion   | None                  | None            |
| Shammas et al.          | 30/M    | Dyspnea/Chest discomfort | No lesion | None                  | None            |
| Asha et al.             | 62/M    | TCP                | CX lesion   | None                  | CABG            |
| Turhan et al.           | 52/M    | ATCP               | No lesion   | None                  | None            |
| Yoshimoto et al.        | 63/M    | ATCP               | No lesion   | Atrial fibrillation   | Oral anticoagulation for AF |
| Chou et al.             | 42/M    | TCP                | No lesion   | None                  | Medical         |
| Nielsen et al.          | 55/F    | TCP                | No lesion   | None                  | None            |
| Kunimasa et al.         | 61/M    | MI                 | LAD lesion  | None                  | NA              |
| Celik et al.            | 57/M    | TCP                | No lesion   | None                  | Medical         |
| Canbay et al.           | 69/M    | Chest pain         | No lesion   | None                  | NA              |
| Araki et al.            | 76/M    | TCP                | CX lesion, LAD lesion | None                  | None            |
| Tanawuttiwat et al.     | 44/F    | ATCP               | No lesion   | None                  | Medical         |
| Datta et al.            | 69/F    | Chest pain         | No lesion   | None                  | Medical         |
| Cung et al.             | 77/F    | TCP                | LAD lesion  | None                  | PCI on LAD      |
| Choi et al.             | 68/F    | ATCP               | No lesion   | None                  | NA              |
| Ghaffari et al.         | 65/F    | Dyspnea            | No lesion   | Massive pulmonary embolism | Medical         |
| Voyce et al.            | 76/F    | AMI                | LAD and CX lesion | None                  | PCI on CX       |
| Sönmez et al.           | 63/F    | Subacute MI        | LAD lesion  | None                  | PCI on LAD      |
| Turfan et al.           | 58/M    | Exertional dyspnea and chest pain | Mid LAD lesion | Severe mitral regurgitation | Mitral valve surgery |
| Ma et al.               | 39/M    | RVMI               | Distal CX occlusion | None                  | PCI on CX       |
| Blaschke et al.         | 59/F    | TCP                | No lesion   | None                  | None            |
| Pourbehi et al.         | 47/M    | MI                 | CX & LAD lesion | None                  | PCI             |
| De Agustin et al.       | 40/M    | ATCP               | No lesion   | None                  | Conservative    |
| Pouraflari et al.       | 44/M    | MI                 | LAD lesion  | None                  | PCI             |
| Sing et al.             | 60/M    | ATCP               | No lesion   | None                  | Medical         |
| García-Blas et al.      | 87/M    | Senkop             | CX lesion   | Severe aortic stenosis | PCI on CX       |
| Present case            | 58/M    | ATCP               | LAD lesion  | None                  | CABG            |

Male: M, Female: F, ATCP: Atipical chest pain, TCP:Typical chest pain, MI: Myocardial infarction, AMI: Akut Myocardial infarction, RVMI: Right Ventricular Myocardial Infarction, PCI= Percutaneous coronary intervention, CABG= Coronary artery bypass grafting, NA: Not available.
Anomalous of coronary arteries are associated with ischemia and sudden death, which could be the result of compression by the aorta and pulmonary artery. However, ischemia is also reported when an anomalous coronary artery does not run between the great vessels as in our case. Although SCA is generally considered benign, it has been associated with myocardial infarction and heart failure [15,22]. In our case, our patient did not have any history of myocardial infarction even though he had severe stenosis in LAD (ostial). Myocardial ischemia was detected with treadmill test. Myocardium ischemia was found in 20.6 % of the cases [15,22,23,25,27,29]. Lesions were detected in both LAD and CX in three cases. Also, lesions were detected in LAD in six cases and CX in four cases. The location of the atherosclerotic lesion in this anomaly does not intervene clinically unless it corrupts the coronary flow. Percutaneous coronary intervention was performed in some cases with lesions. Only two patients with CX lesions underwent coronary artery bypass grafting (CABG) [3,10]. Due to being unsuitable for PCI, our patient was referred for CABG surgery.

The associated factors have been reported to be atrial fibrillation (AF), mitral valve insufficiency and massive pulmonary embolism in different cases. Ghaffari et al. reported massive pulmonary embolism in a SCA patient. They stated that this is the patient's hemodynamic instability. This coronary anomaly is considered as a factor contributing to right ventricular (RV) dysfunction and prolonged unstable state of the patient [21]. In another case, a SCA was detected in one patient with severe mitral valve insufficiency [24]. Yoshimoto et al. reported SCA in a patient with chronic AF [12]. As the L-I variant of SCA is extremely rare, it is difficult to predict whether patients of this type of coronary artery anomaly are at high risk or benign course. Even L-I type of SCA were reported benign in most cases, in our case the patient died on the first postoperative day of his postoperative course because of hemodynamic instability. This could be due to extracardiac manifestation such as pulmonary emboli, RV dysfunction as mentioned above.

Generally, cases with single coronary ostium are considered to be benign. However, these patients experience the symptoms of CAD more critically because of dependence on one coronary artery. Therefore, we think that the recognition of this coronary artery anomaly will contribute to interventional cardiologists and cardiac surgeons.

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