



## Case report

## Malignant right coronary artery originating from left coronary sinus

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## ABSTRACT

Congenital coronary artery anomalies are rare and usually an incidental finding during selective coronary angiography. Most of these variations appear to be of no clinical significance but some do have potentially serious sequelae. Anomalous origin of the right coronary artery from the left sinus of valsalva with subsequent coursing between the aorta and pulmonary trunk called “malignant course” is a relatively uncommon congenital defect. In this report we present a patient with a malignant right coronary artery originating from left coronary sinus © 2015 The Society of Cardiovascular Academy. Production and hosting by Elsevier B.V. All rights reserved. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

## Introduction

Congenital coronary artery anomalies are rare and usually an incidental finding during selective coronary angiography. The incidence of coronary anomalies was reported as 1.3% in a large registry of patients undergoing coronary angiogram.<sup>1</sup> When there is only one coronary artery arising from the aortic trunk by a single coronary ostium the condition is called single coronary artery (SCA). Most of these variations appear to be of no clinical significance but some do have potentially serious sequelae.<sup>2</sup> Anomalous origin of the right coronary artery from the left sinus of valsalva with subsequent coursing between the aorta and pulmonary trunk called “malignant course” is a relatively uncommon congenital defect. The prevalence rate for the RCA branching from the left coronary sinus was reported as 0.43% in patients undergoing computed tomography coronary angiography.<sup>3</sup> Angiography based reports revealed this incidence between 0.03 and 0.17%.<sup>1</sup> It is well established that this anomaly can cause angina pectoris, myocardial infarction, or sudden cardiac death in the absence of atherosclerosis.<sup>4</sup> In this report, we present a patient with a malignant right coronary artery originating from left coronary sinus

was 120/85 mmHg and the pulse rate was 76 beats/min and a 2/6 degree apical pansystolic murmur was heard. The 12-lead electrocardiography (ECG) showed normal sinus rhythm. Transthoracic echocardiography showed a mildly dilated left atrium with mild-to-moderate mitral regurgitation and preserved ejection fraction. Treadmill exercise testing with the Bruce protocol was ambiguous. Ambulatory 24 hour ECG showed frequent ventricular extrasystoles (VES). The selective coronary angiography was performed. Right coronary artery was visualized as anomalously originating from left coronary sinus valsalva (Fig. 1A). The attempt to engage the right coronary catheter to the right coronary artery was unsuccessful and aortic root angiography showed no coronary ostium originating from the right sinus of valsalva. The coronary system was free of significant atherosclerotic disease. 320-slice ECG-gated multi-detector computed tomography (prospective gating) showed a right coronary artery originating from left coronary sinus with an interarterial course called “malignant course” (Fig. 1B–C–D). As the stress myocardial perfusion detected no ischemia, the patient was discharged with the medical treatment including beta blocker therapy for frequent VES and has been followed up at the outpatient clinic uneventfully for two years.

## Discussion

When a single coronary ostium provides blood flow for supplying the entire heart, the condition is called SCA. SCA is a rare congenital anomaly of the coronary circulation and is associated with other congenital cardiac malformations such as transposition of the great vessels, coronary arteriovenous fistula, or bicuspid aortic valve.<sup>1</sup> SCA has generally been regarded as a benign anomaly. The prevalence of anomalous origin and course of coronary arteries is about 0.7–1.96%.<sup>5,6</sup> The anomalous origin may have interarterial, retro-aortic, prepulmonic or septal course, the most common being inter-arterial. If RCA course is not

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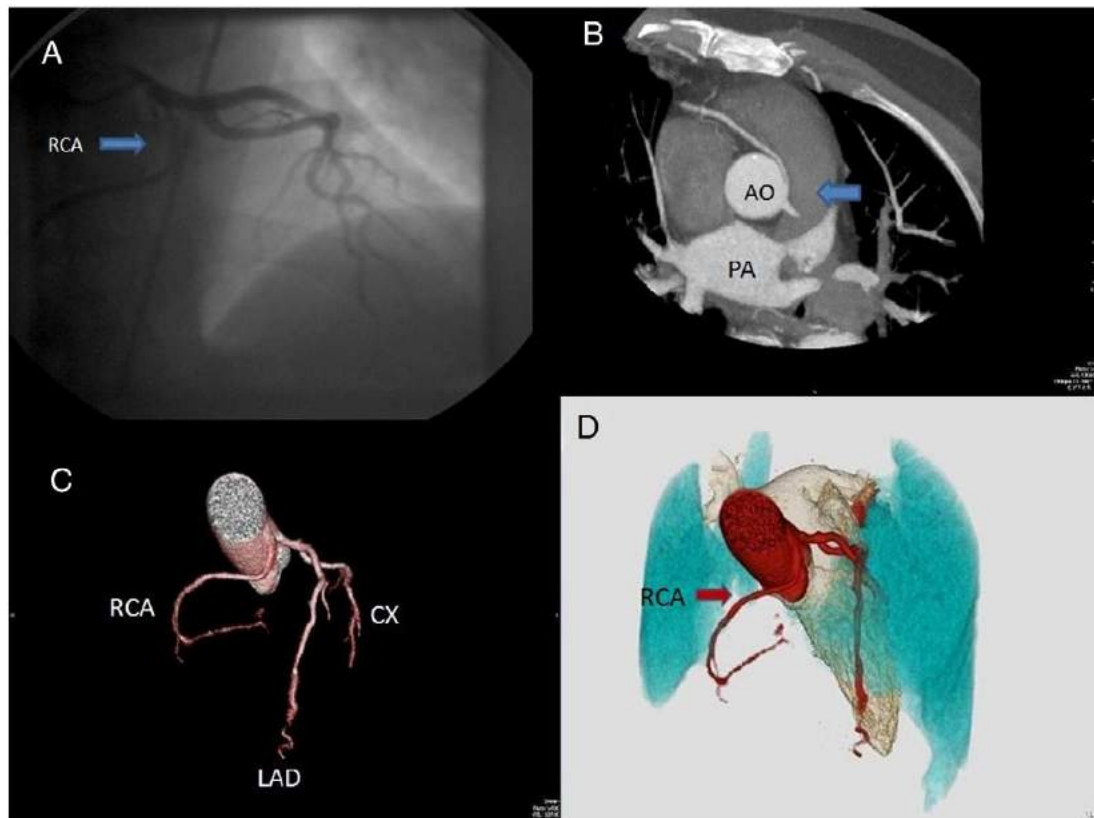
A 63-year-old man with hypertension and chronic obstructive pulmonary disease was admitted to our clinic for evaluation of atypical chest pain and palpitation. On admission, his blood pressure

☆ There is no conflict of interest.

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**Fig. 1.** A: Selective angiogram of the left coronary system in the right anterior oblique projection. The arrow shows right coronary artery (RCA) originating from left coronary sinus. B: Maximum intensity projection of top of heart by multidetector computed tomography. The arrow is showing both RCA and left coronary artery originating from left coronary sinus. RCA courses between pulmonary artery (PA) and aorta (AO). C: 3D-volume rendered image of multidetector computed tomography showing anomalous right coronary artery (RCA) (pulmonary artery is not demonstrated). D: 3D volume rendered image of multidetector computed tomography. The arrow is showing the RCA with an interarterial course. (pulmonary artery is not demonstrated).

between aorta and pulmonary artery, this anomaly is accepted as relatively benign form. A malignant or interarterial course of the right or left coronary artery arising from the opposite coronary sinus has been well described and may have serious sequelae. The interarterial course of a single coronary vessel is subject to compression, which may result in a higher incidence of angina, myocardial infarction, and sudden death. In a majority of previously reported cases, sudden death was triggered by exertion and most of these patients have a positive exercise stress test.<sup>7</sup> Several factors including acute angulation at the origin, compression of the vessel between the aorta and pulmonary artery, slit like ostium and intramural proximal intussusception of the ectopic artery at the aortic-root wall have been proposed for this association.<sup>8</sup> The traditional terminology (interarterial course) implies that the aberrant artery was liable to a scissors-like mechanism, created by the close proximity of the aorta and pulmonary artery. Compared with invasive angiography, multidetector computed tomography allows a more accurate depiction of the origin and course of the anomalous coronary artery. In combination with stress myocardial perfusion, multidetector computed tomography can provide an accurate diagnosis and a complete anatomic and functional assessment of this potentially lethal anomaly to guide patient management.<sup>9,10</sup> The prognosis in single coronary artery is unclear. The incidence of sudden death with this anomaly is estimated at 25–40% and is associated with exercise in most of the reported cases.<sup>11</sup> Revascularization is recommended for documented coronary ischemia in the setting of an anomalous coronary artery coursing between aorta and pulmonary arteries by American College of Cardiology and American Heart Association (ACC/AHA) guidelines for congenital heart diseases.<sup>12</sup> In the present case we decided a conservative strategy, as the patient lived uneventfully for 70 years and there was no documented ischemia with SPECT myocardial perfusion imaging.

## Conclusion

Most of these variations appear to be of no clinical significance but some do have potentially serious sequelae. Anomalous origin of the RCA from the left sinus of valsalva with subsequent coursing between the aorta and pulmonary trunk is called “malignant course”. It is well established that this anomaly can cause angina pectoris, myocardial infarction, or sudden cardiac death in the absence of atherosclerosis. Revascularization is recommended only if there is substantial atherosclerosis and documented ischemia.

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