Anomalous origin of the entire coronary system by three separate ostia within the right coronary sinus – a rarely observed coronary anomaly

Maha Al-Mohaissen MBBS¹, Brett Heilbron MD¹, Jonathon Leipsic MD², Andrew Ignaszewski MD¹

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The anomalous origin of the entire coronary system by three separate ostia within the right coronary sinus is a very rare anomaly with only 34 cases reported in the literature to date. A patient with this rare anomaly who developed coronary artery disease, requiring revascularization, is presented. His coronary computed tomography angiography and coronary angiographic findings are discussed. The present case demonstrates the complementary roles of coronary computed tomography angiography and conventional cardiac catheterization in managing a patient with anomalous coronary arteries and coronary artery disease.

Key Words: *Anomalous coronary arteries; Congenital heart disease; Coronary angiography; Coronary artery disease; Coronary computed tomography angiography*

CASE PRESENTATION

A 56-year-old man, who had been followed up in the heart function clinic at St. Paul's Hospital (Vancouver, British Columbia) for ischemic cardiomyopathy, presented complaining of left-sided chest heaviness that improved with nitroglycerin. In 1998, he had an inferior myocardial infarction that was treated medically. He had a coronary angiogram at that time, the report of which was not available; however, the test did show anomalous coronary arteries. A myocardial perfusion scan showed a fixed inferior wall defect; hence, he was continued on medical therapy. His ejection fraction was 30%. An implantable cardioverter defibrillator was placed for primary prevention of sudden cardiac death.

On examination, he was euvolemic and his B-type natriuretic peptide level was 26 ng/L (normal is lower than 40 ng/mL). A repeat myocardial perfusion scan revealed new inferolateral ischemia. The patient was referred for coronary angiography. This revealed an occluded right coronary artery (RCA), but selective cannulation of all the vessels was not achieved. He was referred for coronary computed tomography angiography (CCTA), which showed three coronary arteries arising from separate ostia in the right coronary sinus (RCS) (Figure 1). The occluded RCA and a septal branch were closely juxtaposed (Figure 1C). The left anterior descending artery (LAD), which ran anterior to the pulmonary artery (PA), had a moderate (50% to 75%) stenosis proximally. There was a moderate to severe (75% to 99%) stenosis in the left circumflex artery (LCX), which took a retroaortic course. The septal branch ran an interarterial course and was normal (Figure 2).

L'origine anormale de tout le système coronaire par trois orifices distincts dans le sinus coronaire droit : Une anomalie coronaire rare

L'origine anormale de tout le système coronaire par trois orifices distincts dans le sinus coronaire droit est une anomalie très rare puisque seulement 34 cas ont été déclarés dans les publications jusqu'à présent. Le cas d'un patient ayant cette rare anomalie et qui a présenté une coronaropathie nécessitant une revascularisation est décrit. Les résultats de son angiographie par tomodensitométrie cardiaque et de sa coronarographie sont exposés. Le présent cas démontre les rôles complémentaires de l'angiographie par tomodensitométrie cardiaque et du cathétérisme cardiaque classique dans la prise en charge d'un patient ayant des artères coronaires anormales et une coronaropathie.

Repeat coronary angiography, using the CCTA as a 'road map', confirmed the origin of the coronary arteries from the RCS. All vessels were imaged consecutively in one cine run (Figure 3). The LCX was a large vessel with 70% stenosis in a second obtuse marginal branch. The LAD had 50% stenosis in its mid segment and supplied collaterals to the RCA. The septal branch was normal (Figure 4). The patient was referred for surgical opinion.

DISCUSSION

Anomalous origin of the entire coronary system from three separate ostia within the RCS is a very rare anomaly; a recent review identified 34 cases reported in the medical literature. The course of the arteries once they originate from the RCS is variable. The RCA follows the usual pathway to the right atrioventricular sulcus in all cases. The LCX is typically posterior to the aortic root, but has been reported to run anterior to the PA or along the interventricular septum. The LAD may have an anterior (anterior to the PA or right ventricular outflow tract), septal or interarterial course (between the PA and aorta). Occasionally, a fourth 'accessory' vessel will originate from the left coronary sinus, in which case it may be mistaken for the LAD or LCX at the time of coronary angiography. Correct identification of these vessels is important; otherwise, the catheterization procedure may be terminated prematurely, before visualization of the coronary vessel is attempted (1).

The clinical significance of this anomaly is mainly determined by the anatomical course of the arteries in relation to the great vessels. Patients with an interarterial course may develop coronary obstruction

¹Department of Cardiology; ²Department of Radiology, St. Paul's Hospital and University of British Columbia, Vancouver, British Columbia
Correspondence: Dr Maha Al-Mohaissen, St. Paul's Hospital, 1081 Burrard Street, Vancouver, British Columbia V6Z 1Y6. Telephone 604-682-2344, fax 604-806-8596, e-mail mahaabdulrahman@hotmail.com

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Figure 1) Coronary computed tomography angiography virtual angioscopy images. Three coronary arteries are seen arising from separate ostia in the right coronary sinus (RCS) (A to C). The right coronary artery (RCA) and a septal branch (S) are closely juxtaposed (C). The RCA is occluded in its proximal segment (D). LAD Left anterior descending artery; LCS Left coronary sinus; LCX Left circumflex artery; NCS Noncoronary sinus

secondary to aortic or PA dilation during exercise and, consequently, ischemia, which may be fatal. The anterior and posterior routes are believed to be benign.

Arteries with high take-off or with ostia that have acute angulations may also undergo sudden occlusion during exertion. Vasospasm due to endothelial damage is another potential mechanism of ischemia (2).

Anomalous coronary arteries, despite their rarity, are associated with a risk of developing coronary artery disease (CAD) that requires revascularization. The gold standard for the diagnosis of an anomalous coronary artery has been conventional cardiac catheterization. Occasionally, however, cannulation of the anomalous coronary ostia may be technically challenging and the precise course of the vessels may not be adequately defined. CCTA has emerged as a noninvasive diagnostic modality and as a complementary tool for coronary arteriography. CCTA can reliably identify the origin of the coronary arteries, characterize the proximal course of the vessels relative to the aorta and PA and, therefore, aid in clinical decision making. It is also useful for the detection of CAD (3).

CAD involving anomalous coronary arteries should be treated with the same approach applied for managing obstructive CAD in normally arising arteries (4). Although the majority of coronary anomalies are not associated with adverse clinical outcomes, origin of the left main artery or LAD from the RCS or origin of the RCA from the left coronary sinus, taking an interarterial course, is considered potentially fatal and is usually an indication for surgical repair. This finding is associated with poor prognosis, particularly in young athletes or military recruits. The retroaortic and prepulmonic courses are considered benign or nonmalignant. The retroaortic course is important to recognize, however, in patients with aortic valve disease because the artery may be compressed during aortic valve or aortic root surgery. The transeptal course is considered to be 'relatively benign' because there are reports of it being potentially fatal (5).

Figure 2) Coronary computed tomography angiography – three-dimensional volume-rendered image (A) and multiplanar reformats (B to D). The left anterior descending artery (LAD) is seen coursing superiorly and then anteriorly to the pulmonary artery (A), with moderate (50% to 75%) stenosis proximally (red arrow) (B). There is a moderate to severe (75% to 99%) stenosis (red arrow) in the left circumflex artery (LCX), which is taking a retroaortic course (C). The right coronary artery (RCA) is occluded proximally (red arrow) (D). S Septal branch

Figure 3) Repeat coronary angiography, using coronary computed tomography angiography as a 'road map', confirmed the origin of the three coronary arteries. All coronary arteries were imaged consecutively in one cine run (A to D). LAD Left anterior descending artery; LCX Left circumflex artery; RCA Right coronary artery; S Septal branch

Figure 4) Selective cannulation of the vessels and evaluation in multiple views revealed a 50% stenosis in the mid left anterior descending artery (LAD) (red arrow) (A) and a 70% stenosis in the second obtuse marginal branch (red arrow) (B). The right coronary artery (RCA) is seen occluded close to the ostium (red arrow), and a septal branch (S) is normal and attributing collaterals to the RCA (C). D1 First diagonal artery; PDA Posterior descending artery; LCX Left circumflex artery

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