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Short Communication

Single Left Coronary Artery with Origin of the Right Coronary Artery from Distal Circumflex

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Summary: We report two very unusual cases of agenesis of the right coronary ostium with continuation of the left circumflex artery as the right coronary artery. The recognition of the anomaly in the first case lead to a better understanding of this finding in the second case, which translated into shorter procedure time, less contrast volume, and fewer catheter manipulations.

Key words: congenital coronary anomalies, single coronary artery

Introduction

Congenital coronary anomalies are found in 0.2–1.6% of the population undergoing diagnostic coronary angiography.[1–3] Many of these anomalies are incidental and benign findings while others may have serious implications. Anomalous origin of the left circumflex artery (LCx) from the right coronary sinus is the most common congenital coronary anomaly.[1,3,4] The absence of either coronary ostium (single coronary artery) is a rare finding, and the congenital absence of the ostium of the right coronary artery (RCA) with the origin of the RCA as a continuation of the distal LCx is extremely rare.[3,5,6]

We present two cases of this latter unusual single coronary artery anomaly. It is interesting that these cases presented within days of each other, and the encounter of such an anomaly in the first case facilitated early recognition of this finding in the second case with shorter procedure time, less contrast volume, and fewer catheter manipulations.

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Case Report

Case No. 1

A 44-year-old white female presented to our hospital with substernal chest pressure at rest. She had a strong family history for premature coronary artery disease but no other known risk factors. Her physical examination was unremarkable. The electrocardiogram (ECG) showed inverted T waves in leads V_1–V_3 with no previous ECG for comparison. Her troponin-I and creatine phosphokinase (CPK) isoenzymes were normal. A two-dimensional echocardiogram was unremarkable.

The patient exercised for 3 min on the Bruce protocol and complained of severe crescendo chest discomfort, following which the test was terminated. There were no additional ECG changes at submaximal heart rate. Cardiac catheterization showed a dominant left system with normal left main (LM), left anterior descending (LAD), and LCx arteries. The LCx was large and dominant, and its terminal part gave rise to the RCA (Fig.1). The right coronary ostium could not be cannnulated despite use of multiple catheters. A 45° left anterior oblique (LAO) aortogram confirmed the absence of the RCA ostium (Fig. 2). Left ventriculogram was normal. Total contrast used was 205 cc and fluoroscopy time was 10 min.

Case No. 2

A 30-year-old white man presented to our outpatient clinic with chest discomfort and dyspnea mostly during periods of physical exertion and emotional stress. He had a history of hyperlipidemia and heavy tobacco abuse. His cardiac and general physical examinations were normal.

The patient underwent technetium-99m sestamibi perfusion scan with good exercise effort and no ischemic ECG changes. The single-photon emission computed tomography (SPECT) images revealed evidence of mild posterolateral ischemia. Cardiac catheterization showed normal coronaries

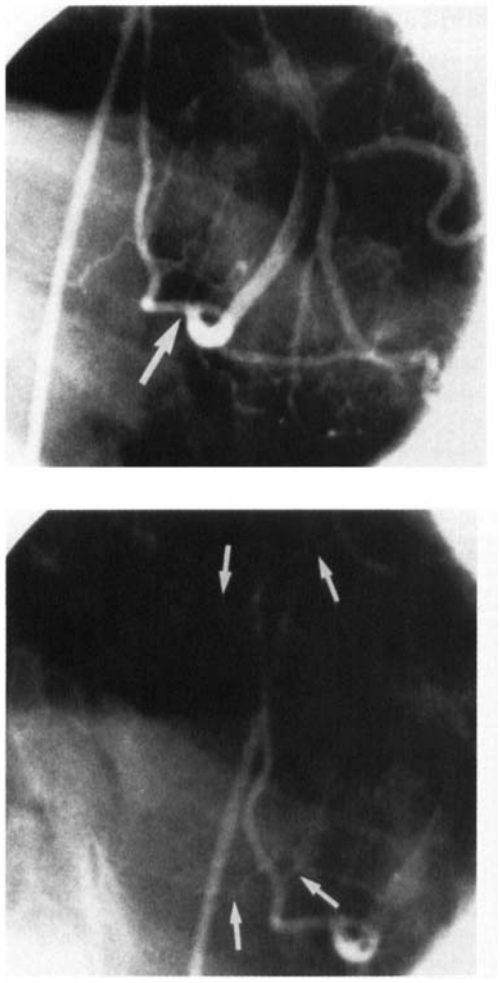

FIG. 1 (A) Right anterior oblique (RAO) caudal view showing the origin of the right coronary artery (RCA) from the terminal left circumflex (LCx) (arrow). The RCA courses up toward the right sinus of Valsalva. (B) Same view showing how the RCA continues in the right atrioventricular groove and up to the right coronary sinus. Notice the small right ventricular and terminal branches (arrows).

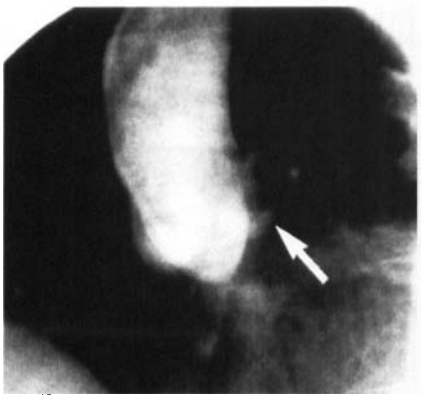

FIG. 2 Aortogram obtained in 45° left anterior oblique (LAO) projection. Note the absence of the right coronary ostium. The left main (LM) ostium is seen clearly (arrow).

with a left dominant system. The RCA ostium could not be engaged. Given our recent experience with the first case, we were quickly able to recognize the origin of the RCA from the terminal LCx with retrograde filling all the way up to the right sinus (Fig. 3). Aortogram confirmed the absence of the RCA ostium (Fig. 4). Left ventriculogram was normal. Right heart catheterization showed normal pressures and oxygen saturation values. Total contrast used was 150 cc and fluoroscopy time was 4.5 min.

Discussion

Anomalies in the origin and course of coronary arteries have been the subject of numerous reports. Some of these anomalies have been associated with sudden death and ischemic complications, particularly in cases of aberrant origin of the LM from the pulmonary artery and aberrant origin of the LM and RCA from the right and left sinuses, respectively.[7] In one postmortem study, 27.3% of patients with the LM artery arising from the right sinus had sudden unexplained death;[8] however, most patients with anomalous origin of the coronary arteries are asymptomatic.

The two cases described here represent a very rare and mostly benign form of isolated congenital coronary anoma-

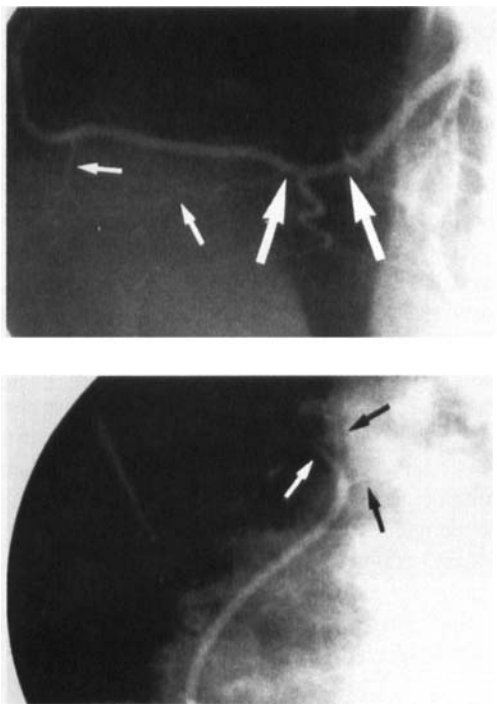

FIG. 3 (A) Left anterior oblique (LAO) caudal view showing the continuation of the left circumflex (LCx) as the right coronary artery (RCA) (large arrows). Note the right ventricular branches (small arrows). (B) Same view as in (A). The RCA continues beyond the crux and courses up in the same distribution as a normal RCA. It terminates at the right sinus after giving off several small branches (arrows).

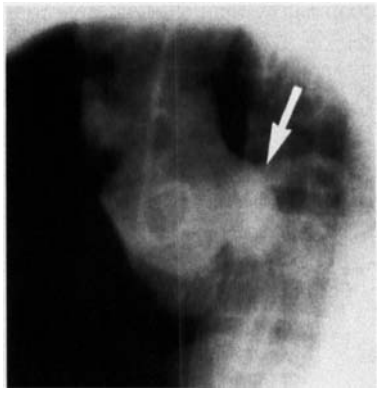

FIG. 4 A 45° left anterior oblique (LAO) aortogram showing the left main ostium (arrow) and agenesis of the right coronary artery (RCA) ostium.

lies. Such an anomaly is not expected to cause ischemia or any other complication. We believe that the chest discomfort in both of our patients was nonischemic. Our second patient had a mild posterolateral perfusion defect, which was most likely a false positive finding as he had no obstructive lesions or anomalies in that distribution.

Very few reports have addressed this particular anomaly in the past[7, 9–13] with an incidence of 0–0.035%.[3, 6, 7, 13, 14] Our cases fall under the L-1 pattern of the Lipton classification,[14] where there is a single left coronary artery with congenital absence of the RCA ostium. The LCx is dominant and provides the posterior descending branch after which it ascends along the atrioventricular groove in the distribution of the RCA and supplies the right ventricular branches. Such anomalies are usually considered to be benign, although one can wonder whether the retrograde perfusion of the RCA territory is equivalent to that in normal vessels.

Failure to visualize one of the coronary arteries should alert the cardiologist to the possibility of an aberrant vessel as coronary arteries rarely "vanish" and are almost always filled by collaterals. Being aware of anatomical variations is important for making the right diagnosis and therapeutic decisions. In one study, 38% of the patients with anomalous origin of a coronary artery had to undergo repeat catheterization before a definitive diagnosis could be made.[13]

The recognition of this anomaly in our first case lead to a "smoother" second case with shorter procedure time, less dye load, and fewer catheter manipulations. In addition, better angiographic views were obtained as the unusual course of the RCA was "expected."

Conclusion

Recognition of these rare anomalies is important for the right diagnosis and for avoiding prolonged procedures and the need for repeat catheterizations.

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