A 9-year-old child presented with syncope during exercise. He received a diagnosis of congenital atresia of the left main coronary artery by angiography. He underwent successful coronary artery bypass grafting. On the third postoperative day, he experienced acute, precordial chest pain. An urgent computed tomographic scan showed an unrecognized anomalous origin of the right coronary artery (RCA) with a 1.5-cm intramural course. He was taken back to the operating room to undergo unroofing of the RCA. This case highlights the difficulty involved in making two rare diagnoses that can cause the same exact symptoms in a patient and the surgical challenges associated with it.


Congenital atresia of the left main coronary artery (LMCA) is a rare condition in which the left main trunk is atretic up to its bifurcation [1]. The left anterior descending (LAD) and the circumflex coronary arteries are typically confluent and do not originate from any vessel or cardiac chamber. They only receive retrograde flow through collaterals from the right coronary arterial system. Fewer than 35 cases have been reported in the literature, most of which have been congenital ostial atresia of the LMCA in children or secondary to advanced atherosclerotic disease in adults. This case report illustrates the diagnostic and surgical challenges associated with this rare entity in a child who was also found to have an anomalous aortic origin of the right coronary artery (AORCA).

A 9-year-old, presumably healthy boy presented to the emergency room with syncope. He had been at football practice, where he was witnessed to fall to the ground clutching his chest. He had been at football practice, where he was witnessed to fall to the ground clutching his chest. Although the initial electrocardiogram was normal, the troponin level was elevated. The following night, he experienced acute precordial chest pain while having a bowel movement. A 12-lead electrocardiogram was concerning for myocardial ischemia in the inferior and right precordial leads. The symptom abated in a few minutes without any intervention. A computed tomographic (CT) angiogram was emergently performed. The LMCA origin could not be visualized. The given clinical presentation and the suspicion of an unusual coronary pattern, coronary artery angiography was pursued. The LMCA could not be cannulated during cardiac catheterization. Selective RCA angiography (Fig 1) demonstrated a prominent conal branch off the RCA that supplied the LAD, with complete atresia of the LMCA. The circumflex coronary artery filled retrograde from the LAD. On the basis of the angiograms, this condition was thought to be congenital atresia of the LMCA, not a variant of a single coronary pattern. There was suspicion for myocardial bridging of the conal branch, which could potentially lead to left coronary insufficiency with exercise. The ostium of the RCA appeared widely patent. The origin of the RCA was slightly superior and more leftward than usual. The significance of the AORCA was downplayed by the diagnosis of the LMCA atresia and possible myocardial bridging of the conal branch of the RCA, which was the sole source of coronary blood flow to the left. The patient had normal systolic function with normal cardiac output. However, he had diastolic dysfunction and elevated biventricular, end-diastolic pressure of 14 mm Hg.

The decision was made to proceed with urgent coronary bypass grafting to the LAD with use of the left internal mammary artery (LIMA). The LIMA was obtained in standard fashion. A standard LIMA-to-LAD anastomosis was performed, and the patient was weaned from bypass without the need for support from inotropic agents. The conal branch from the RCA supplying the LAD was not ligated. The patient recovered well and was transferred out of the intensive care unit on the second postoperative day. The following night, he experienced acute precordial chest pain while having a bowel movement. A 12-lead electrocardiogram was concerning for myocardial ischemia in the inferior and right precordial leads. The symptom abated in a few minutes without any intervention. A computed tomographic (CT) angiogram was emergently performed. The LIMA graft to the LAD was widely patent. However, the RCA was noted to arise from the left coronary sinus immediately adjacent to the commissure with the right aortic cusp. The proximal RCA had an intramural course for an approximate length of 1.5 cm (Fig 2). The patient was urgently taken back to the operating room. The ostium of the RCA was tucked away in the left sinus, behind the commissure post between the left and right leaflets of the aortic valve. The commissure post needed to be taken down to enable full unroofing of the intramural RCA. This was carefully performed with use of a right angle in the coronary and unroofing the wall from inside the aorta. There was an ostial narrowing of at least 60% compared with the size of the RCA beyond the ostium. The RCA was unroofed well into the right sinus. The commissure post was then reimplemented. Transesophageal echocardiography showed good biventricular systolic function with no aortic valve insufficiency. The RCA could be easily visualized arising from the normal position.

The patient was discharged 3 days later. He remains asymptomatic while receiving low-dose aspirin. His echocardiogram continues to demonstrate normal biventricular systolic function and normal diastolic function based on tissue Doppler indices. He underwent a
Atresia of the LMCA is a rare condition. A literature review by Musiani and colleagues [1] collated 28 patients with LMCA atresia; 15 were children and 13 were adults. The pediatric patients in that series presented at varying ages. Their symptoms included dyspnea, syncope, myocardial infarction, ventricular tachycardia, and sudden death. Five children had associated cardiac anomalies, comprising supravalvar aortic stenosis (n = 2), stenosis of the RCA ostium (n = 1), ventricular septal defect (n = 1), and pulmonary valve stenosis (n = 1).

In adult patients, coronary artery bypass grafting (CABG) is definitely the procedure of choice, inasmuch as internal mammary artery bypasses have shown superior long-term patency and great physiologic adaptability to various flow patterns. However, in pediatric patients the long-term results of CABG can be questioned, especially if saphenous vein grafts are used [2]. Overall, in the literature there are reports of only 12 attempted revascularization operations in children with LMCA atresia [1, 3]. When only ostial atresia is present, coronary ostioplasty with the use of a homograft patch has been described [3]. There has been one elegant report of reconstructing the LMCA trunk with a baff of ascending aorta [4]. However, for long-segment LMCA atresia, CABG to the LAD is the only viable option in pediatric patients. We did not ligate the conal branch from the RCA supplying the LAD after CABG, thinking that this was safer in the context of the congenital atresia of the LMCA; this may be controversial because of competitive flow with a LIMA.

The patient in this report had an associated AORCA with a long intramural course. We could not identify any prior reports of this combination of coronary lesions. The significance of the AORCA seen in the initial angiogram was hard to gauge in the presence of the congenital LMCA atresia. This was compounded by the fact that the LAD was supplied by only one major collateral from the RCA, which had dynamic compression from a myocardial bridge. It was concerning that in this patient, coronary insufficiency could potentially develop with exercise. The intramural course of the RCA was not apparent on the initial angiograms, as is often the case. A CT angiogram with curved, multiplanar reformation is sometimes required to accurately demonstrate the intramural course of the RCA, as was the case in this patient. The development of symptoms after revascularization of the left coronary system led us to revisit the coronary anatomy, in particular the RCA, ultimately making the full diagnosis of bilateral coronary artery anomalies and providing the appropriate therapy.

This case highlights the difficulty involved in making two rare diagnoses of LMCA atresia and AORCA that can cause the same exact symptoms in a patient and the surgical challenges associated with it.

References

