Coronary Anomalies

Anomalous Origin of Left Coronary Artery

with Intramural Aortic Course Causing Symptoms in a Teenaged Athlete

Daniel Garcia-Arribas, MD
Carmen Olmos, MD, PhD
Javier Higueras, MD, PhD
Pedro Marcos-Alberca, MD, PhD
Iñigo de la Pedraja, MD
Monica Garcia-Bouza, MD

Anomalous origin of the left coronary artery from the opposite sinus of Valsalva with an intramural aortic course is a rare congenital anomaly with a poor prognosis. We report the case of a 14-year-old soccer player who briefly lost consciousness while sprinting. He had exertional chest pain, syncope, ischemic changes on his electrocardiogram, and elevated cardiac troponin levels. Computed tomographic angiograms showed an anomalous origin of the left coronary artery from the right sinus of Valsalva and a course through the aortic wall toward the left coronary sinus. A surgically created neo-ostium in the left coronary sinus relieved the patient's ischemia, and he resumed playing soccer after cardiac rehabilitation. ***(Tex Heart Inst J 2020;47(2):165-7)***

Anomalous origin of the left coronary artery from the opposite sinus of Valsalva with an intramural aortic course (L-ACAOS-IM) toward the left coronary sinus (CS) is an exceptionally rare congenital coronary artery anomaly.[1,2] It is associated with severe adverse events such as ventricular arrhythmias, myocardial infarction, and sudden cardiac death.[3] We describe the diagnosis and treatment of L-ACAOS-IM in a teenaged athlete who presented with symptoms.

Key words: *Adolescent; coronary vessel anomalies/diagnosis/diagnostic imaging/epidemiology/surgery; death, sudden, cardiac/prevention & control; myocardial ischemia/etiology; sinus of Valsalva/abnormalities; treatment outcome; vascular malformations/physiopathology*

From: *Cardiovascular Institute (Drs. Garcia-Arribas, Garcia-Bouza, Higueras, Marcos-Alberca, and Olmos) and Department of Radiology (Dr. de la Pedraja), Instituto de Investigacion Sanitaria del Hospital Clinico San Carlos, 28040 Madrid, Spain*

Address for reprints: *Daniel Garcia-Arribas, MD, Instituto Cardiovascular, Hospital Clínico San Carlos, Prof. Martín Lagos s/n, 28040 Madrid, Spain*

E-mail: *danigara_1@hotmail.com*

Case Report

In September 2015, a 14-year-old boy who was sprinting while playing soccer had chest pain and palpitations that caused him to briefly lose consciousness and fall to the ground. He recovered rapidly and spontaneously. When an ambulance arrived, the boy was slightly dizzy and had chest pain. His blood pressure was 100/60 mmHg, and his heart rate was 84 beats/min. An electrocardiogram (ECG) was obtained at the field, after which he was brought to our emergency department.

The patient had no personal or family history of early ischemic heart disease or sudden cardiac death. He reported that he had not sought help after a similar episode 6 months earlier when he was playing in a game. Prerequisites for joining his soccer team had been a physical examination, an ECG, and a treadmill test; at that time, his results were normal.

The current ECG showed ischemic changes consisting of deep negative T waves in leads II, III, and aVF, and from leads V_4 through V_6 (Fig. 1). Serial cardiac troponin I levels were 0.6, 1.36, and 0.89 ng/dL. We admitted the boy to our coronary care unit.

The patient remained hemodynamically and clinically stable, and ischemic changes on the ECG gradually returned to normal over several days (Fig. 2). Echocardiograms suggested an anomalous origin of the left main coronary artery (LMCA); in the parasternal short-axis view, a vessel was seen between the aorta and the pulmonary artery, and abnormally directed blood flow was apparent in color-flow Doppler mode. In addition, the papillary muscles were hypertrophic and potentially noncompacted. Cardiac magnetic resonance images showed normal ventricular size and function, and ruled out pathologic hypertrophy and noncompaction.

Computed tomographic angiograms (CTA) revealed an anomalous origin of the LMCA from the right CS, with a proximal course inside the wall of the ascending aorta and continuing toward the left CS. The caliber of the artery was decreased in its intramural course; the area was reduced by a 68% stenosis, and the ratio between its vertical and transverse diameters was 2:1.

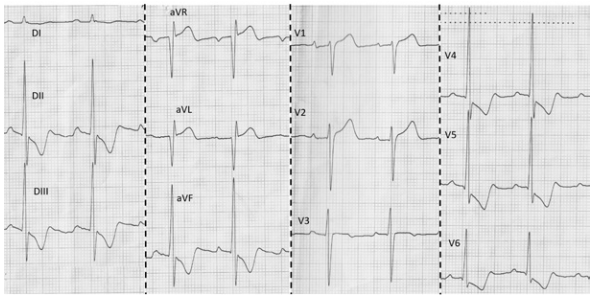

Fig. 1 *Initial electrocardiogram shows deep negative T waves in leads II, III, and aVF and from leads V_4 through V_6, along with 1-mm ST-segment elevation in lead aVR.*

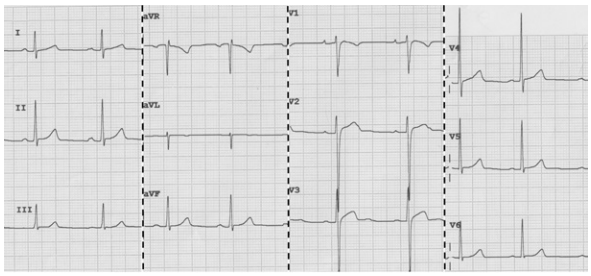

Fig. 2 *Electrocardiogram several days after admission shows gradual normalization of the inverted T waves in the precordial leads, and return to an isoelectric ST segment in lead aVR.*

After a nuclear treadmill test to rule out cardiac ischemia, the patient reported slight chest pain, which spontaneously resolved. No ECG changes or myocardial perfusion defects had been detected. His ischemia on the first ECG, symptoms, and troponin elevation prompted us to schedule him for cardiac surgery.

We performed a median sternotomy and established cardiopulmonary bypass (CPB) with aortic and cavoatrial cannulation. After cross-clamping the aorta and making an oblique aortotomy, we administered anterograde cold cardioplegic solution. On inspection, the aortic valve was normal. There were 2 separate coronary ostia in the right sinus of Valsalva. The ostium of the anomalous LMCA was high in the right CS, close to the right-to-left commissure, and it had an intramural course. To avoid disturbing the intercoronary commissure, we used a modified unroofing technique to create a neo-ostium in the left CS. We opened the lumen, inserted a blade through the anomalous ostium of the LMCA and advanced it until the left CS was reached, created an ostium where the LMCA left the aortic wall, and closed the lumen by using single 7-0 Prolene sutures. The neo-ostium had a diameter of 7.8 × 3.3 mm and an area of 20.2 mm^2; the 68% stenosis was reduced to 24% (Fig. 3). The uneventful surgery included a CPB time of 66 min and an aortic cross-clamp time of 54 min.

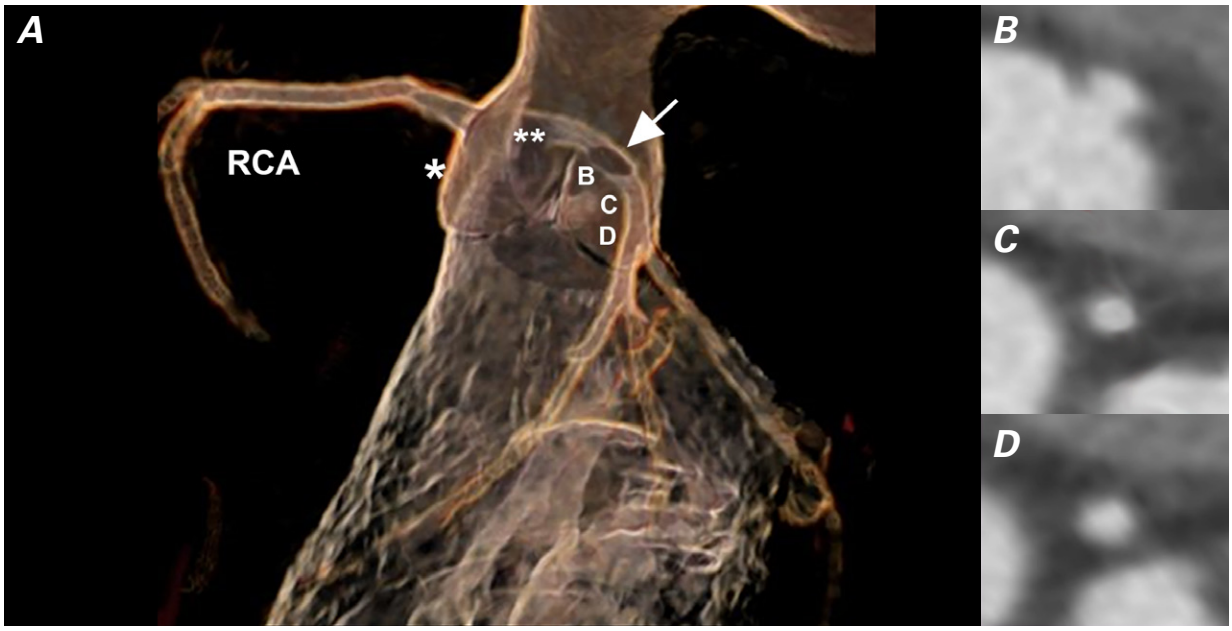

Fig. 3 *Low-dose 64-slice cardiac computed tomograms obtained postoperatively. **A**) Three-dimensional rendering of the coronary artery tree with myocardial perfusion imaging transparency shows where the left main coronary artery (LMCA, double asterisk) emerged from the right coronary cusp (asterisk), and where the neo-ostium connects the LMCA to the left coronary cusp (arrow). **B**), **C**), and **D**) are virtual intravascular reconstruction images (mild axial views) at different levels of the LMCA. **C**) The luminal area at the most stenotic point after the neo-ostium was 3.36 mm^2. The luminal area **B**) before the stenosis was 4.41 mm^2, and **D**) after it, 5.02 mm^2, which averaged to a 24% luminal stenosis.*

RCA = right coronary artery

The patient had no complications other than postcardiotomy syndrome. He was discharged from the hospital 20 days postoperatively, after a CTA showed good patency of the LMCA neo-ostium and no stenosis at the anastomotic site (Fig. 4). Two months postoperatively, the results of a treadmill test indicated no ischemia. The patient completed a cardiac rehabilitation program and was recertified to participate in sports. One year postoperatively, he was playing soccer without symptoms.

Discussion

The estimated prevalence of L-ACAOS-IM in the general population is 0.15%.[1,2] The condition can be asymptomatic; however, a course of the LMCA inside the aortic tunica media can cause chest pain, syncope, and ventricular arrhythmias, and the prognosis is poor.[3] Ischemia in L-ACAOS-IM results from lateral compression of the artery in its intramural course.[3-5] In autopsy studies, this anomaly was determined to be a frequent cause of sudden death in young people who had died of nontraumatic causes.[6] For this reason, when a young person has documented myocardial ischemia, surgical intervention should be considered.[3] Percutaneous coronary intervention may be considered in adult patients who have acute presentations or when surgical risk is unacceptable; however, the evidence supporting this approach is limited to a few case reports.[7,8]

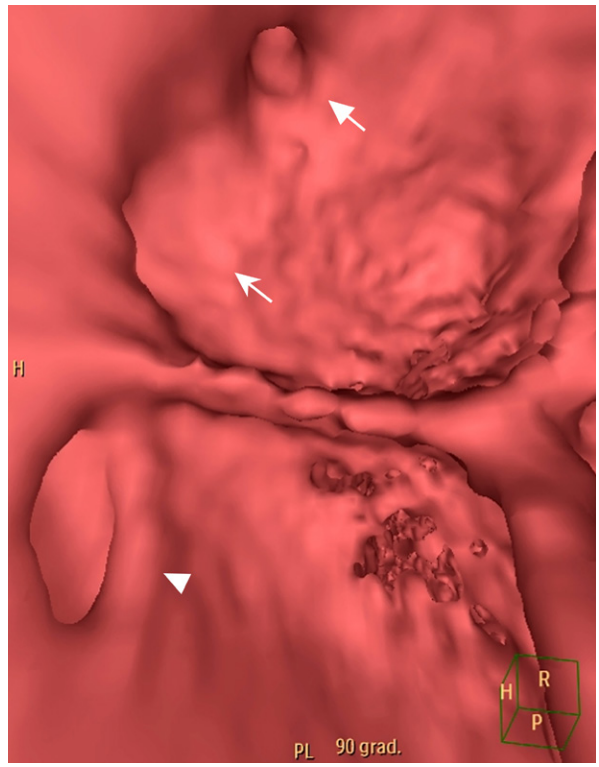

Fig. 4 *Postoperative cardiac computed tomogram (virtual reconstruction) shows the origin of the right coronary artery (arrow), the original left coronary ostium (notched arrow), and the neo-ostium (arrowhead).*

Acknowledgments

We appreciate the support from Dr. Luis C. Maroto in performing cardiac surgery, and from Dr. José Luis Rodrigo in acquiring and postprocessing cardiac images.

References

1. Davis JA, Cecchin F, Jones TK, Portman MA. Major coronary artery anomalies in a pediatric population: incidence and clinical importance. J Am Coll Cardiol 2001;37(2):593-7.
2. Angelini P, Villason S, Chan AV Jr, Diez JG. Normal and anomalous coronary arteries in humans. In: Angelini P, editor. Coronary artery anomalies: a comprehensive approach. Philadelphia: Lippincott Williams & Wilkins; 1999. p. 27-150.
3. Angelini P. Coronary artery anomalies: an entity in search of an identity. Circulation 2007;115(10):1296-305.
4. Angelini P, Uribe C, Monge J, Tobis JM, Elayda MA, Willerson JT. Origin of the right coronary artery from the opposite sinus of Valsalva in adults: characterization by intravascular ultrasonography at baseline and after stent angioplasty. Catheter Cardiovasc Interv 2015;86(2):199-208.
5. Angelini P, Uribe C. Anatomic spectrum of left coronary artery anomalies and associated mechanisms of coronary insufficiency. Catheter Cardiovasc Interv 2018;92(2):313-21.
6. Eckart RE, Scoville SL, Campbell CL, Shry EA, Stajduhar KC, Potter RN, et al. Sudden death in young adults: a 25-year review of autopsies in military recruits. Ann Intern Med 2004;141(11):829-34.
7. Lee JH, Park JS. Successful percutaneous coronary intervention in the setting of an aberrant left coronary artery arising from the right coronary cusp in a patient with acute coronary syndrome: a case report. BMC Cardiovasc Disord 2017;17(1):186.
8. Nef HM, Möllmann H, Möllmann S, Dill T, Hamm CW, Elsässer A. Primary stenting of the left main coronary artery with anomalous origin from the right sinus of Valsalva. Int J Cardiol 2007;114(1):137-8.