Anomalous aortic origin of the right coronary artery: Embryology over genetics

Ajay M. Patel, MD, Nirmal T. Sunkara, MD, Raylene M. Choy, BS, RDCTS, and Shaun P. Setty, MD
Orange and Long Beach, Calif

Anomalous aortic origin of the coronary artery (AAOCA) has an incidence of 0.1% to 0.6%,1-3 with debate over genetic and embryologic mechanisms as potential causes. We describe a unique case of anomalous aortic origin of the right coronary artery (RCA) found in 1 of a pair of identical twins.

CASE REPORT

A 24-year-old man was evaluated for resting angina at an emergency department visit. An initial 12-lead electrocardiogram showed T-wave inversion in leads II, III, and aVF. Troponin I level was 4.82 ng/mL and peaked after admission at 6.80 ng/mL (reference range, 0.00-0.09 ng/mL). A transthoracic echocardiogram demonstrated normal left ventricular systolic function without wall motion abnormalities. Given the patient’s young age and low pretest probability for coronary artery disease, a coronary computed tomography angiogram was ordered to evaluate the patient’s coronary arteries.

The coronary computed tomography angiogram revealed an anomalous take-off of the RCA from the left coronary sinus with an interarterial course between the aorta and the pulmonary artery (Figure 1).

Because of the presence of symptoms alone, the patient underwent a surgical repair. The anatomy consisted of an RCA origin near the right/left aortic commissure, which was slit-like and intramural for a distance of 8 mm with a steep angle. The specific findings of the coronary ostia and its course were made intraoperatively, as is commonly the case. The proximal RCA was unroofed (incising and excising the intramural aortic wall), dissected off the aortoventricular groove, and translocated as a large button to the right aortic sinus, allowing the coronary artery to have a gentle curve and not be under too much tension, which would alter the coronary ostial geometry. The patient’s recovery was uncomplicated, and he was discharged on postoperative day 3. He continues to do well 5 months later on follow-up. For surveillance, his identical twin had a transthoracic echocardiogram showing normal origins of both right and left coronary arteries.

DISCUSSION

The treatment of AAOCA is in evolution with different centers differing on surgical treatment options for asymptomatic individuals. There are many surgical approaches to treat patients with different AAOCA anatomic configurations.3 The presence of myocardial ischemia is a definite indication for surgery. The workup and evaluation of an asymptomatic individual are more controversial. At the present time, our preoperative imaging studies do not consistently provide the exact ostial anatomy that allows us to make a determination on high-risk AAOCA characteristics.3 There are no formal
guidelines for the screening of first-degree relatives with AAOCA.

The development of left and right asymmetry, the placement and patterning of internal organs, and the associated vasculature during embryologic development are fascinating and complex. Defects in situs determination can be associated with congenital heart disease, and multiple mechanisms have been proposed. The direction of ciliary motion in the embryonic node has been proposed as one potential mechanism. Shinohara and colleagues reported that 2 rotating cilia are sufficient to break left-right asymmetry in mouse models.

CONCLUSIONS

The presence of AAOCA in only 1 of the identical twins suggests an embryologic disturbance rather than a genetic mechanism. There are only a few published cases describing asymptomatic nonidentical siblings with AAOCA.1-3,5 There are reports of nonidentical siblings with AAOCA, as summarized by Laureti and colleagues.5 There are clearly additional factors that dictate this phenotypic anatomic presentation. To our knowledge, this is the first reported case of a pair of identical twins with and without AAOCA. The possibility of an embryologic insult that disturbs the coronary artery origins also may translate into embryologic insults that cause certain forms of congenital heart disease. This is only a hypothesis with no scientific proof, but it is worth investigating in the future. Although many genetic links are being established as the field of genomics grows, there are likely other, nongenetic reasons for certain anatomic configurations, such as the finding in this pair of identical twins.

References