Editorial Commentary

Anomalous Origin of the Left Coronary Artery from the Opposite Sinus of Valsalva

Typical and Atypical Features

Paolo Angelini, MD, FACC

Having read the preceding case report by Lee and colleagues[1] with interest, I would like to make a few comments about the specific features of their case and to discuss the latest concepts regarding the clinical evaluation of the coronary artery anomaly (CAA) in question.

Specific Features of the Reported Case

Lee and co-authors reported the case of a 15-year-old boy who had an anomalous left coronary artery arising from the opposite sinus of Valsalva (L-ACAOS). The patient experienced 8 episodes of syncope (the last of which was accompanied by chest pain) before his condition was correctly diagnosed and effectively treated. This chain of events is not unusual, because sudden cardiac death (SCD) is consistently reported as the most common initial (and, frequently, the final) manifestation in patients who have a symptomatic CAA.[2] Only in about 20% of cases is a subsequently fatal CAA diagnosed due to prodromic symptoms, of which syncopal spells are the most predictive and common.[3,4] The initial mechanism of SCD is usually hypotension and bradycardia.[2] Ventricular fibrillation tends to occur only secondarily, after the patient has had persistent ischemia, hypotension, and cardiac standstill, especially if successful resuscitation has occurred (as in typical cases of reperfusion arrhythmias).[2]

The authors' claim that the distal left main coronary artery was narrowed beyond its intramural segment is surprising and not firmly supported by the evidence they present. In the preoperative coronary angiogram (Fig. 1A[1]), one can neither determine the exact severity of the stenosis nor differentiate between an intramural versus extramural location. In fact, the aortic wall cannot be seen at all with catheter angiography or even with computed tomographic angiography (CTA). Volume-rendered images are able to reveal this degree of anatomic detail only via dedicated tomographic imaging, which was not available in the case presented. In Figure 1A,[1] the end portion of the stenotic left main segment could still be intramural, as it usually is in cases of this type.[5]

Figure 1B[1] is an intraoperative photograph of the interior of the aortic root, showing the adjacent coronary ostia; again, the severity of the stenosis of the proximal left coronary artery cannot be established on the basis of this image. Figure 2A,[1] a CTA image, is more relevant: it shows a 3-dimensional reconstruction of the left main trunk, which seems to have a diffuse narrowing that extends from the ostium to a few millimeters proximal to the left main bifurcation. Unfortunately, the relationship of the stenotic segment to the aortic wall still cannot be determined with volume-rendered imaging, and the cross-section of the coronary luminal area cannot be evaluated.

External exploration of the aortic root, even after microdissection, cannot lead to reliable evaluation of an extramural stenosis of the left coronary main stem. When the coronary artery was transected in this case, the surgeon could have commented about the inner lumen, but this was not reported. Was the stenosis indeed extramural, and if so, how much of it? Was there thickening of the intima or a mural clot? These questions remain unanswered.

From: *Department of Cardiology, Texas Heart Institute at St. Luke's Episcopal Hospital, Houston, Texas 77030*

Address for reprints: *Paolo Angelini, MD, P.O. Box 20206, Houston, TX 77030*

E-mail: *PAngelini@leachmancardiology.com*

The intramural aortic course of the ectopic coronary artery is the basic culprit in ACAOS, because it implies hypoplasia, lateral compression, and further phasic narrowing during systole.[5,6] The location of the stenosis is especially relevant in choosing the optimal surgical technique (unroofing vs reimplantation), as the authors mentioned.[6]

The authors stated that they patched the extramural segment of the left main trunk, implying that they placed at this crucial location a vein graft that involved an extensive suture line. Postoperative imaging seems to show excellent early results (Fig. 2B[1]); however, such a patch and suture line may be vulnerable to late failure (accelerated atherosclerosis), especially in view of the patient's young age. In this regard, stress testing will probably be more important than CTA, but the latter approach could be quite revealing if it were performed with a precise (ultrafast) tomographic technique. If these methods yield uncertain results, either transthoracic echocardiography with Doppler interrogation or, better, intravascular ultrasonography (IVUS) may be able to provide clarification. Follow-up observation will be required for the rest of the patient's life.

Current Concepts Regarding L-ACAOS

In the recent medical literature, researchers have generally concluded that L-ACAOS, because of its intramural aortic course, is the most lethal kind of CAA in patients past infancy.[5,7] In these patients, the pathophysiologic mechanisms of ischemia were initially[8,9] speculated to be 1) external compression of the ectopic vessel related to the position of the anomalous trunk "between the aorta and pulmonary artery"; 2) a decrease in blood pressure related to the acute angulation of the artery's slit-like, tangential origin; 3) a flap- or valve-like mechanism; 4) superimposed spasm; or 5) an ostial ridge, possibly of an atherosclerotic nature. Even in studies involving semiquantitative analysis of individual case severity, such as those performed by Taylor and colleagues,[8] no single factor has seemed to be associated with the risk of SCD in L-ACAOS.

In addition, at our own center[5,6] and others, IVUS data have suggested that ACAOS (both left- and right-ACAOS) involves the following features: 1) The proximal segment is intramural, embedded in the aortic wall. This is a constant feature of any case that involves ectopic origination of a coronary artery from the opposite sinus of Valsalva, with a preaortic course. 2) Only the intramural segment is circumferentially narrowed by hypoplasia with respect to the distal portion of the left main coronary artery. 3) In addition, on cross-section, the intramural coronary trunk shows lateral compression that increases phasically during systole. 4) This lateral compression of the coronary cross-section increases during exercise, probably as a result of the increased cardiac output and especially stroke volume. Each of these features varies widely from case to case, but preliminary evidence suggests that the severity of stenosis correlates with clinical symptoms and the probability of SCD.

Conclusion

According to our current understanding, only the intramural course of the ectopic artery seems to be related to proximal coronary obstruction in L-ACAOS. An extramural stenosis of a congenital nature (vs an acquired, atherosclerotic nature) in L-ACAOS has never before been reported, and its possibility is hard to accept, because it implies the existence of another congenital mechanism of stenosis with respect to the intramural course itself. In the case presented, it is possible that the left main trunk was severely hypoplastic in the segment that had an intramural aortic course and that, extramurally, it gradually tapered toward the distal left main artery. The latter segment of the left main coronary artery was relatively ectatic, as suggested by the 4 CTA images that accompany this commentary (Fig. 1 below). The best means of precisely studying the severity, mechanism, and location of such stenosis is IVUS,

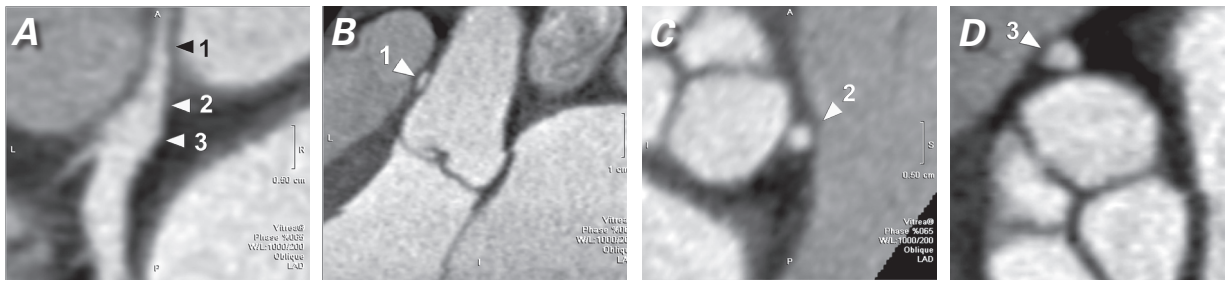

Fig. 1 *Computed tomographic angiograms of the left main trunk, which exits the aortic wall (not imaged with this technique).* ***A****) Sites 1, 2, and 3 are the locations from which the cross-sections of the left main trunk (****Figs. B****,* ***C****, and* ***D****, respectively) were obtained; the calculated cross-sectional areas are shown in parentheses.* ***B****) 1: intramural proximal left main trunk (5.7 mm²);* ***C****) 2: borderline segment (11.2 mm²); and* ***D****) 3: extramural distal left main trunk (16.7 mm²). As shown, the intima is always thin throughout the left main trunk, and the extramural proximal left main segment that appears smaller in angiography is round and progressively larger, in order to reach the relatively ectatic left main coronary artery at its bifurcation. The stenotic effect at that level is unsubstantiated (see text).*

which could have clarified the details of the current case. When performed by an expert operator, this technique is neither contraindicated nor difficult. Its use will enable physicians to establish firmer criteria for indicating interventional treatment and determining the optimal surgical technique.

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