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Anomalous right coronary artery from the left sinus of Valsalva presenting a challenge for percutaneous coronary intervention

Dear Editor,

Treatment of atherosclerotic lesion(s) with percutaneous coronary intervention (PCI) in the setting of anomalous coronary artery origin from the opposite (improper) sinus (ACAOS) is always challenging. Çalışkan et al.[1] presented a case of right coronary artery (RCA) originating from the left aortic sinus, i.e. right ACAOS with a proximal obstructive lesion that was successfully treated with PCI. However, the authors did not describe the course of the aberrant RCA, obviously considering that such a defect is invariably associated with an interarterial course. Although this is true, we should have in mind that other courses such as retrocardiac, retroaortic, intraseptal, and prepulmonic courses are theoretically possible and may not always be benign. [2,3] The interarterial course, in particular, has the most potential for adverse sequelae, specifically exercise-related sudden cardiac death (SCD) in the young. [2-5] In such settings, identification of the anatomo-functional disturbances related to the specific circumstances of the crossing of the anomalous vessel towards its dependent territory is important for patient management.

During a 30° right anterior oblique ventriculography or aortography, a right ACAOS with an interarterial course will be depicted anterior to the aorta and may appear as a radiopaque "dot".[3] Based on the circle and loop approach for interpreting coronary angiograms, i.e. the atrioventricular groove is a circle and the interventricular septum forms a loop that intersects this circle both anteriorly and posteriorly. The straight rightward course of the anomalous RCA presented in an anteroposterior (frontal)-caudal view favors an interarterial trajectory being oriented directly towards the right atrioventricular groove. Carriers of a left or right ACAOS with the interarterial course aged less than 30 or 35 years comprise the group at the highest risk for SCD.[2,4] Other manifestations such as syncope, dyspnea, angina, and myocardial infarction are more prevalent among older individuals and may not be related to exercise but emotional stress, the onset of hypertension, and possibly aortic regurgitation or rapid weight gain.[2] Such defects result in clinically evident ischemia only occasionally; however, subclinical ischemic episodes may occur. The latter is supported by pathologic evidence for replacement-type fibrosis that could predispose to malignant arrhythmias and SCD.[4] Indeed, in more than 70%, SCD occurs without forewarning symptoms, being more frequently in carries of left ACAOS. Using intracoronary ultrasound studies, Angelini et al.^[2,5] showed that the interarterial course invariably entailed intussusception of the tangentially originating proximal ectopic vessel within the aortic media for a variable length. The intramural segment was stenotic (30-70% area stenosis at rest) due to hypoplasia and lateral compression. Moreover, stenosis worsened intermittently due to phasic systolic accentuation of compression, and it was further aggravated following pharmacologic simulation of exercise conditions. Such evidence favors the presence of a potentially decreased coronary flow reserve at baseline, which may further decrease to a critical level under conditions entailing increased stroke volume and/or aortic pressure, thus leading to ischemia. Spasm of the proximal intramural vessel, secondary to mechanical endothelial injury has also been proposed as a mechanism of ischemia.^[5,6] Although it has not been confirmed in a large series, there have been reported cases of right and left ACAOS where spasm, either spontaneous or induced was evident in the catheterization laboratory. It has been proposed that intracoronary ultrasound examination should be carried out, targeting the anatomo-functional behavior of the proximal intramural vessel under exercise-like conditions and spasmogenic stimuli in all the carriers of such defects presenting serious symptoms.^[5] Clinical manifestations are mainly determined by the size of the dependent myocardium, thus in right ACAOS.

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Such an investigation is justified in the presence of a large, dominant RCA; a small, non-dominant RCA will generally produce nonsignificant symptoms if does at all.[2,5] Currently, right ACAOS defects with the interarterial course and evidence for ischemia merit surgical correction, while this treatment may also be beneficial when there is evidence for coronary hypoplasia, lateral compression of the lumen, or restriction of flow, regardless of the absence of ischemia. On the other hand, a retrospective study involving Japanese patients with ACAOS (44/56 cases having a right ACAOS with interarterial course, 1 case having a right aortic sinus-derived left anterior descending artery with the interarterial course, none had an interarterial left main coronary artery) demonstrated that conservative therapy, i.e. limitation of physical exercise and/or drug therapy including a beta-blocker, may be a valid approach.[7] Of the 44 patients without significant atherosclerosis, none died or suffered a myocardial infarction as a direct consequence of the anomaly during follow-up (mean 5.6±4.2 years). However, four of the 22 patients with right ACAOS who underwent an exercise stress test developed ventricular tachycardia or hypotension.

In the case by Çalışkan et al.,[1] a 41-year-old patient with a history of anterior myocardial infarction, class IC unstable angina, i.e. postinfarction angina, and 2-vessel disease was successfully treated with PCI. The anomalous RCA in this case was large and dominant; hence, it could lead to a major ischemic event in the future or subclinical attacks of ischemia that could predispose the patient to lethal ventricular tachyarrhythmias. Furthermore, frustration and emotional stress may ensue in this patient due to limitations in physical exercise, potentially raising the possibility of anomaly-related ischemia. Although nuclear stress test frequently performed following the diagnosis of such defects in combination with echocardiography or computed tomography angiography may reveal exercise-related ischemia or scars and contribute to risk stratification, it is usually negative; hence, the potential for a reliable noninvasive functional assessment is limited. Given all these and the lack of prospective trials assessing the optimal treatment of such defects, one could advocate surgical revascularization in this patient.

In ACAOS with the interarterial course, investigation of the anatomo-functional features of the intussuscepted proximal ectopic segment with intravascular ultrasound may enable identification of those patients who need interventional treatment as well as correlation of the anatomic and functional data with clinical outcome in an attempt to define prognosis and optimal treatment.

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Author's reply

Dear Editor,

We appreciate with great pleasure the comprehensive critical review of the reader on our case report. In this case, we did not describe the course of the aberrant right coronary artery (RCA). Considering the fact that such an anomalous RCA had some part lying in the interarterial course, especially its proximal part, demonstration of the course would