Three Echocardiographic Signs to Identify Anomalous Origin of the Circumflex Coronary Artery from the Right Sinus of Valsalva: A Case Report

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INTRODUCTION

The prevalence of congenital coronary artery anomalies, without clinical and hemodynamic repercussions, is estimated to range from 0.21% to 5.79% on the basis of coronary angiography (CA), computed tomography, and autopsy. The diagnosis is usually made easily on CA or computed tomography, but the presence of coronary anomalies may be suspected on transthoracic echocardiography (TTE), whenever feasible. Anomalous origin of the left circumflex coronary artery (LCx) from the right sinus of Valsalva (RSV) is the most frequent anomaly, with an estimated prevalence of about 0.39%. It is considered a benign anatomic variance, but cases of minor and major cardiovascular events have been described, such as angina, myocardial infarction, and less commonly sudden cardiac death. Specific echocardiographic signs associated with a retroaortic anomalous LCx course have been described: the "bleb sign" seen on transesophageal echocardiography (TEE), as a round structure in mitroaortic angle in the long-axis aortic view, and the "crossed aorta sign," a binary structure that seems to cross the aorta perpendicularly to its long axis in a five-chamber apical view, also described as the "tubular shape" or "retroaortic anomalous coronary" (RAC) sign on TTE.

CASE PRESENTATION

A 48-year-old male patient presented to our clinic with a 3-month history of angina. Physical examination was unremarkable. Electrocardiography showed normal sinus rhythm without any repolarization abnormalities.

The patient was known to have hypertension and dyslipidemia and was taking angiotensin receptor blockers and statins. No personal or family history of cardiovascular disease was reported.

Complete TTE revealed preserved global systolic function (left ventricular ejection fraction 55%, calculated using the biplane Simpson method) with regional hypokinesia of the basal to mid inferolateral wall and mild mitral regurgitation. From the four-chamber apical view, tilting the transducer to a shallower angle to a more anterior scan plane exploring the retroaortic region, we identified a binary structure above the mitral valve plane, in the atrioventricular groove, directed toward the right coronary sinus and overlapping the aortic root (Figure 1). This sign is known in the literature as the RAC sign, typically associated with anomalous retroaortic course of the left coronary artery. Searching for other typical signs of anomalous LCx coronary origin, angulating the scan plane to interrogate the aortic valve, we observed a small circle beneath the noncoronary cusp in the transthoracic parasternal long-axis view, similar to the bleb sign on TEE (Figures 2A-2C). Moreover, a tunnel-shaped structure extending behind the aortic root in a conventional short-axis view was also seen, which could represent the orthogonal view of the bleb sign (Figure 3). After visualizing the origins of the left and right coronary arteries above the aortic valve plane in the parasternal short-axis view (Figure 4), an anomalous origin of the LCx with a retroaortic course was hypothesized. Afterward, the patient underwent dipyridamole stress echocardiography (the accelerated high-dose infusion protocol for dipyridamole was 0.84 mg/kg over 6 min), and reversible regional akinesia of the basal to mid inferolateral wall appeared. As a consequence, the patient was admitted to the hospital for CA. Blood test results, in particular cardiac troponin T (11 ng/L; normal range, <14 ng/L) and serum creatinine (1.16 mg/dL) levels, were within the normal reference ranges. CA revealed a hemodynamically stable coronary tree without signs of significant stenosis or occlusion. Since the patient was asymptomatic and had no comorbidities that would contraindicate percutaneous coronary intervention, the decision was made to proceed with coronary angiography, which confirmed the presence of a retroaortic course of the LCx with a normal coronary tree. The patient was discharged on aspirin and statins, and follow-up appointments were scheduled.

Keywords: Echocardiography, Coronary vessel anomaly, Coronary circulation, Cardiovascular disease, Coronary angiography

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Figure 1 Modified transthoracic four-chamber apical view, obtained by tilting the transducer to a shallower angle to a more anterior scan plane, exploring the retroaortic region. A binary structure above the mitral valve plane into the atrioventricular groove directed toward the right coronary sinus, overlapping the aortic root, can be noted (RAC sign).
significant stenosis of the proximal LCx with origin from the RSV and no other significant stenoses (Figure 5). A drug-eluting stent was implanted in the anomalous coronary artery with good final angiographic flow (Figure 6); no complications were observed, and the patient was discharged 2 days after the procedure.

Three months later, an exercise stress test induced no signs or symptoms of myocardial ischemia. Subsequent clinical follow-up was unremarkable.

DISCUSSION

LCx origin from the RSV with retroaortic course is generally considered a benign anatomic variation. Nevertheless, some cases of minor and major cardiovascular events have been described in the literature. Elements related to malignant outcomes are an acute angle of takeoff, a dilated aortic root, and an intramural course, resulting in compression of the proximal segment of the vessel. It has also been suggested that turbulent flow in anomalous coronary arteries might result in the development of atherosclerosis, although this theory requires further confirmation. Earlier and greater degree of atherosclerotic stenosis have been described in anomalous coronary arteries arising from the RSV with a retroaortic course.10

The diagnosis of this anomaly is generally made randomly on CA during the evaluation of patients with suspected coronary artery disease. Lately, coronary computed tomography, in particular multidetector computed tomographic angiography and the use of advanced dose reduction techniques, have been playing a major role in assessing the origins and courses of coronary arteries, with a higher detection rate than CA.11,12 However, these methods are expensive, are invasive, and expose patients to ionizing radiation and iodinated contrast agents. It has long been known that TEE is an alternative semi-invasive method capable of identifying anomalous origin of the coronary arteries and their relationship with the large vessels.13 There are some limitations associated with this method due to the inability to visualize the entire course of the coronary arteries, the use of many human and material resources, experience of the sonographer, and greater invasiveness, which might result in possible intolerance and postexamination side effects (arrhythmias, damage to the esophageal mucosa, difficulty swallowing, chest or abdominal pain, vomiting). It follows that TTE is a valid alternative, being a fast, less expensive, noninvasive method; furthermore, in experienced hands, it can provide additional information about the origin and course of the coronary arteries, with almost no side effects.

In 2017, Witt et al9 proposed the transthoracic echocardiographic RAC sign, related to a retroaortic coronary course anomaly.
Case reports have also proposed the bleb sign on TEE as a marker of LCx origin from the RSV. To the best of our knowledge, no other transthoracic echocardiographic signs have been described in patients with this anomaly.

In our case, we describe three simple signs on TTE in the same patient that can guide the noninvasive assessment of coronary anatomy. Although these three signs may allow the diagnosis of anomalous origin of the LCx from the RSV, some considerations are required. First of all, diagnosis of retroaortic course of the LCx originating from the RSV must be accompanied by individuation of the origin of both the right and left coronary arteries, to distinguish this anomaly from others characterized by a retroaortic course (such as origin of the left coronary artery from the RSV and origin of the right coronary artery from the left sinus of Valsalva).

Second, the presence of artifacts cannot be excluded; for example, in the parasternal short-axis view, the hyperechogenicity of the posterior aortic wall could be confused with the bleb sign in the orthogonal view. To minimize misdiagnoses, a suggestion would be to identify first the best known RAC sign (as in our case), described as a binary structure above the mitral valve plane directed toward the right coronary sinus in a modified four-chamber apical view. This view, obtained by tilting the transducer to obtain a more anterior scan that explores the retroaortic region, makes it possible to distinguish the RAC sign from the coronary sinus (the latter has larger dimensions, is located on a posterior plane, and has a lower course than the atrioventricular sulcus, where the LCx can originate from the RSV). The RAC sign must be distinguished from aortic valve calcifications, which instead have a consensual movement to that of the valve and also do not have internal anechogenicity; however, its simple identification, even in patients with poor acoustic windows, makes it the basis for the diagnosis. After identifying the more specific RAC sign, it is necessary to look for the other two signs in the parasternal short-axis view. If these two signs are isolated, they could be mistaken for artifacts; on the contrary, if they are identified together with the first, they can greatly increase the probability of diagnosis of anomalous origin of the LCx from the RSV. With regard to the structure identified on TTE by the bleb sign, more difficult to identify because of its small size, it could be useful to differentiate it from any mitroaortic calcifications through the anechogenicity inside which is a characteristic of all vessels; it could also be mistaken for an abscess of the mitral-aortic fibrosa, but it differs from the latter both for its smooth circular or ellipsoid appearance and the elongated retroaortic appearance in the orthogonal transthoracic echocardiographic view. It should be noted also that normally there are no other anatomic structures in that area of the heart.

Third, color Doppler is not useful in identifying the bleb sign in the orthogonal view, as the structure is without signal because the direction of the flow is perpendicular to the axis of the ultrasound beam; however, in some cases, color Doppler may allow identification of the origin of the LCx from the RSV and its initial course. These are some tips that sonographers and cardiologists can consider in normal clinical practice; however, the operator’s experience combined with the ultrasound setup (such as the reduction of gain and time gain compensation) certainly represent a more successful strategy.

In our case, the signs on TTE raised the suspicion of an aberrant coronary origin, while the role of stress TTE was to diagnose inducible inferolateral wall myocardial ischemia, supplied by the LCx artery.

**CONCLUSION**

This case underlines the possibility of TTE to identify abnormal origin of the LCx from the RSV and the retroaortic course using three echocardiographic signs. Additionally, we wish to emphasize how to recognize these signs and how to distinguish them from artifacts and other cardiac structures.

With this report we also wish to promote the use of TTE in the diagnosis of coronary anomalies, avoiding more invasive and harmful diagnostic tests.

Finally, we must not forget how a simple routine cardiac examination can prove time-saving and cost-effective in diagnostic workup.

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