Unusual case of single coronary artery: questions of methods and basic concepts

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Coronary artery anomalies continue to constitute a confusing subject in modern cardiology. While most anomalies are considered to have a benign prognosis, the literature and cardiologic culture frequently imply an intrinsic, systematic association of coronary anomalies with severe clinical presentations. We present a case of unusual single coronary artery, in order to elucidate the logical process that should be used to study similar cases.

A 56-year-old female presented with a 6-year history of atypical chest pain and an abnormal electrocardiogram. Heart catheterization revealed an abnormal coronary tree interpreted by some observers as a benign coronary anomaly, by others to indicate the need for coronary angioplasty.

A nuclear stress test was performed after 1 year of unrelenting symptoms and showed mildly abnormal findings, leading to a more definitive angiographic study that clarified the anatomy and the prognosis. The case is essentially and only an example of single coronary artery with origin of all branches from the right coronary sinus, but with an unusual triple origin of the branches serving the left anterior descending territory.

The notion that a case of single coronary artery may have significant prognostic and clinical repercussion is frequently repeated in the current inconclusive literature. A rational discussion should deal both with individual case objective evidence and theoretical general consideration.

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Introduction

Coronary artery anomalies continue to constitute a confusing subject in modern cardiology[1,2]. While most anomalies are considered to have a benign prognosis, the literature and cardiologic culture frequently imply severe clinical presentations, at least in some individuals, at least in some occasions of their life[3,4].

Case report

A 56-year-old female was referred to the Coronary Artery Anomalies Center of the Texas Heart Institute for a third specialistic opinion in the presence of a 6-year history of chest pain, in a hypertensive and hypercholesterolemic patient with family history of coronary artery disease. The chest pain syndrome was atypical for the following reasons: 1) the location of radiation was variable in time; 2) the precipitating factors were inconsistent: sometimes the occurrence was at rest, occasionally with exercise, usually during periods of intense emotional upset; 3) duration varied from minutes to days (in the absence of electrocardiographic changes or enzyme elevations); 4) response to sublingual nitroglycerin was equivocal. The resting electrocardiogram remained unchanged over the years and events, and found abnormal only because of poor progression of the R wave in the anterior leads, in the presence of a left anterior hemiblock pattern. A first heart catheterization was performed 6 years before the current admission and interpreted as showing a complex coronary anomaly described as "single right coronary artery" that "could not be treated surgically", "with diffuse disease of the left anterior descending branch". Medical treatment was initially recommended, but eventually questioned by a second opinion that introduced the possibility that an angioplasty of the left anterior descending artery could be indicated, even though technically difficult. A nuclear treadmill myocardial scintigraphy was interpreted as showing mild apical reversible ischemia. A second angiography was carried out at our institution before and after intracoronary injection of nitroglycerin. No fixed coronary obstructions were found. A single coronary ostium was located in the middle of a normal right coronary sinus of Valsalva. The short mixed trunk subdivided

promptly in a normally coursing, but quite dominant right coronary artery (which provided also a branch to the obtuse margin of the heart), and in two branches that crossed to the left side of the heart and reached both the anterolateral and anteroseptal areas. The two anterior branches (Figs. 1 and 2) differed in course and destination: one lied anteriorly, over the upmost area of the pulmonary infundibulum and then followed the upper interventricular sulcus, while providing perforating branches to the upper half of the septum. The second crossed the interventricular septum while providing a large septal branch, before ending into small diagonal and intermediate branches. The posterior descending branch of the right coronary artery wrapped around the apex and reached the distal anterior interventricular groove, providing anterior septal penetrating branches. A mitral valve prolapse was demonstrated on ventricular angiography. Reassurance and medical treatment with antihypertensive, vasodilating medication was recommended, jointly with statins. Clinical follow-up at 6 months revealed adequate resolution of the angina presentation.

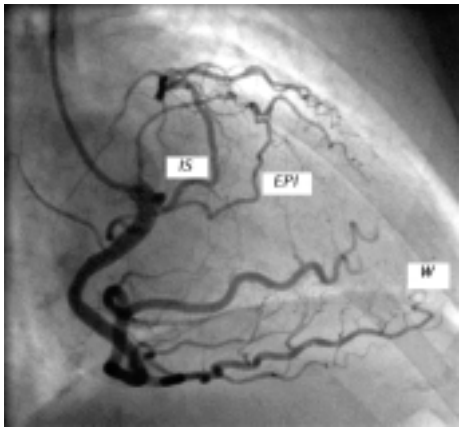

Figure 1. *Right anterior oblique, cranial projection. EPI = epicardial-course anterior subdivision (prepulmonic); IS = intraseptal branch; W = wrap-around branch of the posterior descending branch that provides the distal left anterior descending branch.*

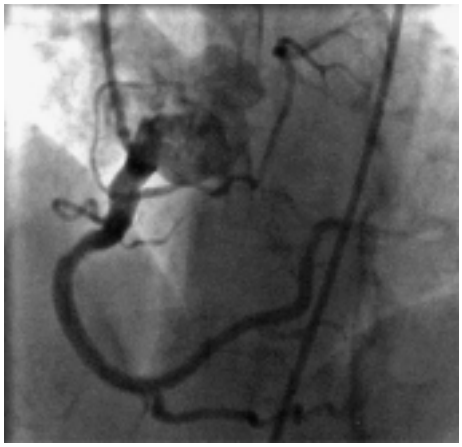

Figure 2. *Left anterior oblique, cranial projection.*

Discussion

Coronary anomalies are described in 1-2% of the general population[2], and frequently cause some confusion to the casual observer. The following notes are provided as a practical example/guide, in order to form a consistent method to interpret similar, unusual cases.

When describing (angiographically or anatomically) coronary anomalies, the observer is mainly concerned with the origin and course of each branch (with respect to the basic cardiac structures), and with the nature of each branch (which is provided by the dependent territory). Generally, sequential, multiplane coronary angiography consistently and adequately provides expert observers with a means for three-dimensional imaging, that enables to properly describe coronary anomalies.

The first issue in our patient is the exact anatomic and clinical diagnosis[5]. Coronary angiography performed at baseline, in the absence of vasodilating agents, suggested initially the presence of diffuse coronary disease in the territory of the reversible defect found at nuclear scintigraphy. The clear resolution of diffuse small branches narrowing after vasodilation suggested the presence of a hypertonic baseline state, more than Prinzmetal's angina, and surely not of fixed atherosclerotic changes.

The peculiar anatomic feature of our example of coronary anomaly was the total disruption of the left coronary pattern, with a circumflex territory supplied by the distal branch of the right coronary artery and a split, triple origin of the left anterior descending components. The circumflex anomaly had origin from the distal right coronary artery with a posterior course (at the posterior atrioventricular sulcus)[6]. The anterior-lateral-septal territory had a triple supply: a) an unusually prominent posterior descending branch of the right coronary artery that wrapped around the apex, to end at the mid-distal portion of the anterior interventricular groove; b) an infundibular branch off the right-sided mixed trunk prolonged to reach the proximal anterior interventricular sulcus; c) a septal branch off the right-sided mixed trunk provided both a large first septal branch and, after re-emerging to the epicardial position, also branches to the diagonal and to the ramus medianus.

The emerging concept, recently suggested by investigators of coronary anomalies, states that most anomalies do not cause ischemic clinical repercussion because of their abnormal origin or proximal course, but because of some specific variable features that require a quantitation[6]. Most notably, the class of anomalies that most frequently are related to myocardial ischemia and to sudden death in athletes is the so-called "anomalous origin of a coronary artery from the opposite sinus"[7]. A recent pathophysiological interpretation proposes that such anomalous arteries consistently exit from the aorta with a tangential angle, leading to an in-

tramural course of its proximal 1 to 3 cm segment (intussusception), before exiting to the subepicardial space. Such intramural segment seems to be hypoplastic (reduced circumference at a viable degree with respect to the more distal segment) and features a variable severity of lateral compression that increases in systole, as shown by intravascular ultrasound imaging[7]. Another likely pathophysiological mechanism is seen in myocardial bridges, which feature systolic compression of the intramyocardial segment, greatly variable in severity from patient to patient[6]. Not all the patients with an anomalous origin of a coronary artery from the opposite sinus or muscular bridges have clinical repercussions, but arguably only the ones who have either more extreme degree of functional obstruction and/or are involved in more extreme exertion (like athletes do) and/or have associated pathologic conditions, like spontaneous spasm and/or clot formation[6].

In our case, the only feature that we thought could potentially result in a clinical repercussion was the intramyocardial (septal) passage of the trunk leading to the diagonal and intermediate branches. The angiographic degree of systolic narrowing was trivial at baseline, but obvious after intracoronary injection of nitroglycerin, even though not severe. Intravascular ultrasound examination was not carried out because of the relatively small lumen of the vessel and the associated risk of inducing vasospasm while introducing the rigid ultrasound probe[8]. Noticeably, the scintigraphic area of reversible myocardial ischemia was related to the distal ventricular septum, that most likely was provided for by the distal portion of the posterior descending (right coronary artery) and not by this intramyocardial branch, anyway. We should probably conclude that the stress test must have yielded a false positive result in this case, possibly a breast artifact.

In summary, our patient seemed most likely to have been identified because of a casual finding of a coronary anomaly[9]. Reassurance and nonspecific medical treatment appeared to have resulted in symptomatic improvements at 6-month follow-up. At this stage of our studies on coronary anomalies, there is no quick substitute for the careful and extensive collection of long-term follow-up data in similar, well-described entities.

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