Anomalous Origin and Interarterial Course of Right Coronary Artery Associated with Angina and Proven Ischemia

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Abstract

Keywords

- ▶ coronary anomaly
- ▶ ischemia
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- ▶ angina

Clinical significance of coronary arteries with anomalous origin and/or course is highly heterogeneous. Anomalies with the origin from the opposite sinus and interarterial course can be associated with angina, syncope, and sudden cardiac death. However, there are no clear guidelines for diagnosis and treatment of such cases. We present the case of a young lady who presented with typical angina, and later proved to have an anomalous right coronary artery (RCA) originating from the left sinus of Valsalva coursing between the aorta and pulmonary artery. This was associated with demonstrable stress ischemia with nuclear perfusion scan. The patient underwent surgery with a bypass graft to the anomalous RCA with complete relief of her angina.

In April 2012, a 25-year-old lady was seen in our clinic with the chief complaint of chest pain on exertion. The pain was central, crushing like, radiating to her neck, and was typically occurring on exertion. The patient noted that she experienced the pain especially in the mornings when she was rushing to catch her bus to work; the pain was worse when the weather was cold and if she had her breakfast. She was consistently experiencing the pain on exertion for the last 2 years. She did not have any risk factors for atherosclerotic coronary artery disease. Echocardiography findings were within normal limits. A treadmill exercise stress test was performed where she reached the target heart rate and complained of chest pain at peak exercise, but only non-diagnostic subtle upsloping ST depressions were observed. Because of low pretest probability of an obstructive coronary artery stenosis a conventional coronary angiography was not performed at that stage. With the possibility of a coronary artery anomaly the patient was referred for a computed tomography (CT) coronary angiography. The CT scan demonstrated anomalous origin of the right coronary artery (RCA) from the left sinus of Valsalva and the proximal course of the RCA between the aorta and pulmonary artery (►Fig. 1).

When questioned in depth, the patient did not report any history of syncope–presyncope or palpitations.

While the patient's symptoms were typical of angina, to aid in therapeutic decision making an objective criteria for ischemia was searched. A nuclear perfusion scan was performed that revealed a mild stress induced perfusion defect in the inferior segment of the left ventricle (►Fig. 2) (the interpreting physician was aware of patient's typical anginal symptoms and the presence of coronary anomaly, but not of the exact anatomy to avoid any bias). The decision was to refer patient for surgery. Robotic surgery through a minithoracotomy was performed and the right internal mammary artery was anastomosed to the distal RCA, the proximal RCA was tied off to prevent steal phenomenon. After an uneventful surgery, the patient recovered and her chest pain on exertion subsided completely. Two months after the operation, an exercise stress test was performed which confirmed complete relief of symptoms. A further nuclear perfusion study was not performed at that stage not to expose the patient to further radiation.

Anomalous coronary arteries are rare congenital abnormalities; however, they may be life threatening.

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Fig. 1 A 64-slice CT coronary angiograph. (A) Anomalous origin of the RCA from the left sinus and proximal course between the aorta and pulmonary artery. (B) Reconstructed image revealing common ostia of both the right and left coronary arteries, slit like orifice of RCA, proximal interarterial course, and intramural location in the aortic wall of the RCA. (C) The difference in the luminal diameter between the proximal interarterial segment and the distal normal segment of the RCA is clearly seen. Ao, aorta; PA, pulmonary artery; RCA, right coronary artery; LCx, left circumflex artery; LAD, left anterior descending artery.

Unfortunately, there are no clear proposed guidelines on their diagnosis and management. Clinical significance of coronary arteries with anomalous origin and/or course is highly heterogeneous. While some of the benign subtypes are totally clinically silent, others may be associated with sudden cardiac death (SCD).¹ One of the most severe forms of coronary anomalies is anomalous origin of the left coronary artery from the pulmonary artery. As the name implies, the left main artery arises from the pulmonary artery and hence supplies the myocardium with the deoxygenated blood from the pulmonary circulation. In addition, collaterals to the left coronary artery form and cause steal phenomenon draining to the low-pressure pulmonary artery. The anomaly manifests itself in the first years of life with congestive heart failure

and myocardial ischemia. While the mortality is high in uncorrected cases, rarely patients may survive to elderly ages; in this case, it is associated with ischemic cardiomyopathy, congestive heart failure, and SCD.²

Coronary anomalies with the origin from the opposite sinus and interarterial course are another group that has been linked to serious consequences.¹ In a series of 1950 consecutive angiograms, Angelini P et al defined the incidence of anomalous origin of RCA from the left sinus as 0.92% and origin of the left coronary artery from the right as 0.15%.¹ These anomalies have been linked to myocardial ischemia, angina, syncope, and SCD; however, the mechanisms causing these are poorly understood. Proposed mechanisms are intussusception of the proximal interarterial segment in

Fig. 2 Stress and rest bull's-eye representation (top row) and vertical long axis (bottom row) images of myocardial perfusion revealing stress induced and reversible perfusion defect in the inferior segments. INF, inferior.

the aortic wall making it prone to hypoplastic development and compression by the expanding aorta on increased cardiac output.^{1,3} Unfortunately, most of the patients who suffered an SCD had no prior symptoms, or if symptoms had been present they were mostly nonsignificant. Myocardial ischemia and angina are only episodic in these cases, and hence do not fit the typical exertion induced angina pattern.¹ Moreover, stress tests do not reveal any evidence of ischemia in most cases. In a series of 36 patients operated for anomalous origin from the opposite sinus and interatrial course, 81% of the patients were presenting with angina, shortness of breath, or syncope and only 43% of the patients had an abnormal stress test.⁴ Detection of the origin and proximal course of the coronary arteries is straightforward with the novel techniques, cardiac magnetic resonance, and CT coronary angiography and perhaps a lower threshold to refer suspected cases to these techniques may be appropriate. Luckily, our patient presented with typical exertion induced angina consistently present for a considerable duration of time that enabled correct and timely diagnosis. Robot-assisted minimally invasive surgery has been increasingly used in the correction of

anomalous coronaries from the opposite sinus.⁵ Apart from the obvious excellent cosmetic result, this technique appears to be associated with rapid healing and decreased rates of complication without having any compromise in the quality of the bypass grafting.⁵

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