The Heart Surgery Forum #2008-1129
12 (1), 2009 [Epub February 2009]
doi: 10.1532/HSF98.20081129

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Ostial Patch Plasty of an Aberrant Right Coronary Artery in a Symptomatic Teenager

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ABSTRACT

An otherwise healthy 15-year-old girl presented with a history of transient myocardial ischemia. At the time, troponin levels were elevated to 20 µg/L. A myocardial perfusion scintigraphy evaluation revealed a minor scarring with an irreversible inferior defect at rest and at exercise. On admission, the patient was free from symptoms. Transthoracic echocardiography and computer tomography examinations revealed an aberrant origin of the right coronary artery (RCA) from the left sinus of Valsalva, just below the commissure. Repair was accomplished by incision into the RCA and patch plasty. An anomalous aortic origin of a coronary artery from the incorrect coronary sinus of Valsalva is a rare congenital cardiac defect that is associated with an increased risk of sudden death and cardiac morbidity; however, ischemia can occur even when the patient remains asymptomatic. Although this anomalous artery often shares a common orifice with the other coronary artery, it did not in the reported case. In conclusion, because of the high risk of sudden cardiac death, aggressive surgical management is indicated in symptomatic patients.

INTRODUCTION

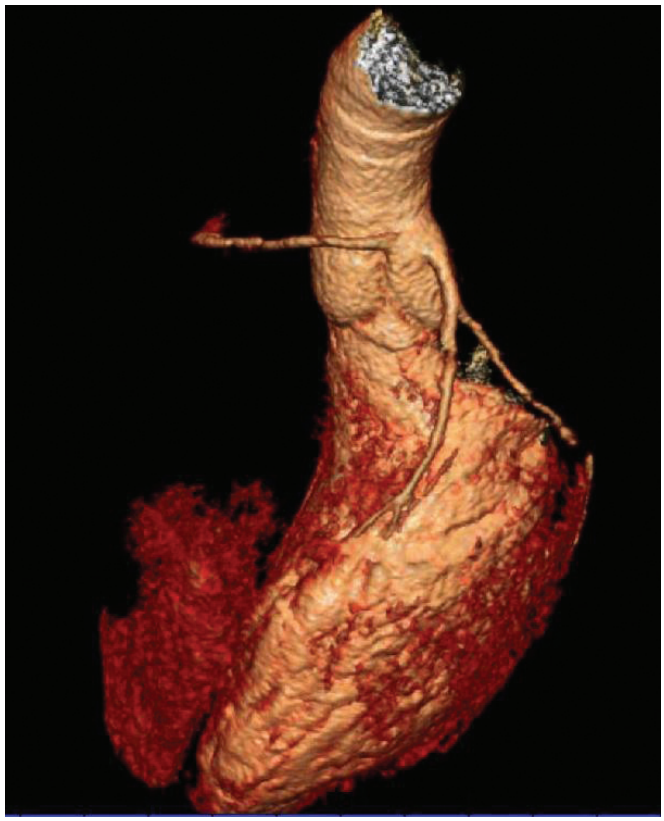
Anomalous aortic origin of a coronary artery from the incorrect sinus of Valsalva is a rare congenital cardiac defect. It is associated with an increased risk of sudden death and cardiac morbidity [Cheitlin 1974; Yamanaka 1990; Maron 1996]. The anomalous coronary artery may travel between the great vessels in either an extramural or an intramural course. The anomalous artery often shares a common orifice with the other coronary artery and it may be slit-like at its takeoff.

MATERIALS AND METHODS

A previously healthy 15-year-old girl had clinical signs of a myocardial infarction. Echocardiography and computer tomography examinations revealed an aberrant origin of the right coronary artery (RCA) from the left sinus of Valsalva (Figure).

RESULTS

After a median sternotomy, the heart was arrested with cold crystalloid cardioplegia, and an aortotomy was performed. Repair was accomplished by incision into the RCA and patch plasty with autologous pericardium [Jaggers 2008]. The RCA did not have an intramural course; however, at its ostium, which was slit-like just below the commissure, the RCA also had an acute takeoff from the lumen of the aorta (Video).



Computer tomography depicting an aberrant origin of the right coronary artery from the left sinus of Valsalva.

Received October 24, 2008; received in revised form December 16, 2008; accepted December 18, 2008.

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The patient's postoperative course was uneventful. An electrophysiological study excluded any arrhythmia. An echocardiographic evaluation demonstrated unrestrictive blood flow to the RCA.

DISCUSSION

Although anomalous origins of coronary arteries are rare in asymptomatic children, the prevalence is greater than found in other prospective studies [Davis 2001]. Davis and colleagues examined the proximal coronary artery anatomy of 2388 children and adolescents by echocardiography over a 3-year period. The patients had anatomically normal hearts, but further tests were performed in those with coronary anomalies. Four children (0.17%) were identified with anomalous origin of their coronary arteries, and angiograms, exercise perfusion studies, and/or stress tests were then performed. Ischemia can occur with an anomalous origin of either the left or right coronary artery.

The pathophysiology of compromised coronary blood flow has many possible mechanisms. The anomalous vessel may be compressed between the aorta and the pulmonary trunk, especially during exercise-induced distention of the sinus of Valsalva. Kinking or torsion of the aberrant vessel may also occur as it courses between the great vessels. An acute angle of origin of the anomalous artery with respect to the ostium may form a slit-like opening that could become further compressed or occluded during systole—apparently the case in our patient. Sudden cardiac death occurs most frequently when the anomalous vessel courses between the aorta and the pulmonary trunk. This high-risk anatomy was present in all 4 patients identified by Davis and colleagues.

Although other imaging tools, such as angiography and magnetic resonance angiography, can be used to identify anomalous vessels, echocardiography remains the imaging modality of choice. It provides an excellent, noninvasive tool to diagnose anomalous coronary arteries in young individuals, because its ability to visualize the origin and proximal segments of the coronary arteries is very high. In our described case, however, a computer tomography evaluation had already been performed at the referring institution. Therefore, after confirmation of the diagnosis by echocardiography, no other imaging study was needed.

Many surgical strategies have been suggested, including coronary reimplantation, coronary artery bypass grafting, or the patch plasty we used. Because of the high risk of sudden cardiac death, aggressive surgical management is indicated in symptomatic patients. Identification of coronary artery origins should be included as part of any echocardiographic examination. When premonitory symptoms such as exertional chest pain and syncope occur, an anomalous origin of coronary arteries should be suspected.

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