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Short Communication

Anomalous Coronary Arteries: A Familial Clustering

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Summary: We present two brothers with anomalous origin of left coronary artery from the right sinus of Valsalva. Screening of other members of the family revealed no coronary artery anomalies. The likelihood of finding such a rare anomaly in two members of a family by chance alone is extremely small; extensive review of the literature found only three other reports of such familial association.

Key words: familial, coronary anomalies, sinus of Valsalva, left main coronary artery, coronary angiography, interarterial, retroaortic

Introduction

Coronary artery anomalies are discovered in approximately 1% of patients undergoing coronary angiography;[1] the incidence at necropsy is 0.3%.[2] The majority are of no clinical significance. The most serious problem is the predisposition for sudden cardiac death when the coronary artery follows an interarterial course between the aorta and the pulmonary artery. Familial clustering of coronary artery anomalies is rare. We describe an unusual case of two brothers with anomalous origins of the left main coronary artery.

Case Reports

Case No. 1

A 32-year-old man presented to a local emergency department for evaluation of chest pain while driving to work. He described a squeezing discomfort with radiation into both arms, associated with palpitations, diaphoresis, and dizziness. Sublingual nitroglycerin relieved his chest pain, but no objective evidence for ischemia or myocardial infarction was discovered. Three similar episodes of chest pain had occurred prior to this visit. He had no significant past medical or family history. Risk factors included only tobacco abuse. He was discharged to home the same day with the diagnosis of noncardiac chest pain. He presented to his family physician 1 week later with recurrent chest pain. Electrocardiography, chest x-ray, and Holter monitor were normal. He underwent a nuclear perfusion study, exercising > 10 METS with no symptoms and normal perfusion images. A transthoracic echocardiogram revealed normal left ventricular structure and function. Because of recurrent symptoms, cardiac catheterization was performed. There was no atherosclerotic obstructive disease. The patient did have an aberrant origin of the left main coronary artery from the right coronary cusp; it followed a retroaortic course (Fig. 1), and a transesophageal echocardiogram confirmed the course (Fig. 2). The patient was reassured and no further therapy for the coronary artery anomaly was undertaken. At follow-up 4 years later, the patient states that he continues to experience rare episodes of chest pain, but this does not limit his lifestyle. He continues to perform all his activities, including softball and hunting, without difficulty and has not undergone further diagnostic testing.

Case No. 2

The 30-year-old brother of the patient in Case No. 1 presented to his primary care physician for evaluation of sharp, substernal chest pain. It occurred most frequently with exertion and occasionally occurred at rest. His past history was significant for hypertension and gastroesophageal reflux disease. He did not smoke or use illicit drugs and had no family history of premature coronary artery disease. Stress echocardiography revealed no evidence of ischemia, normal left ventricular function, and an exercise capacity of 15.2 METS with no symptoms. The patient continued to experience exertional chest pain. Because of ongoing symptoms, cardiac catheterization was performed. No angiographic evidence of atherosclerosis was observed, but the left main coronary artery had an aberrant origin from the right coronary cusp (Fig. 3). Visible compressions could be seen during systole (Fig. 4). Cardiac magnetic resonance imaging (MRI) confirmed the interarterial course

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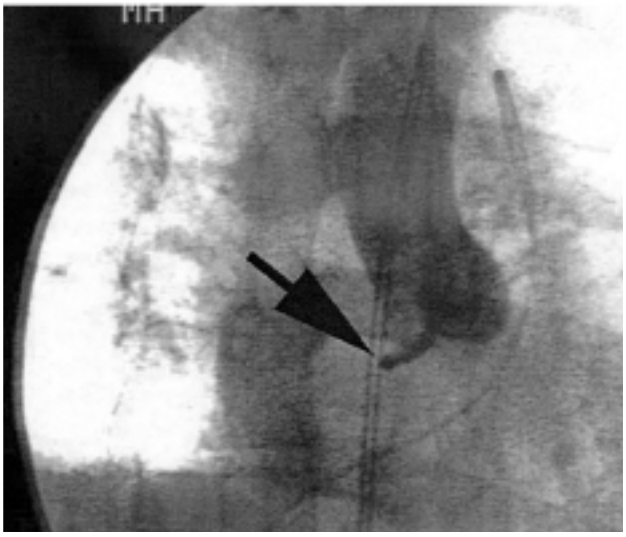


FIG. 1 Ascending aortogram (30° right anterior oblique [RAO] projection) demonstrates the retroaortic course of anomalous left coronary artery. Arrow indicates "Dot" sign seen to the left of the aortic root.

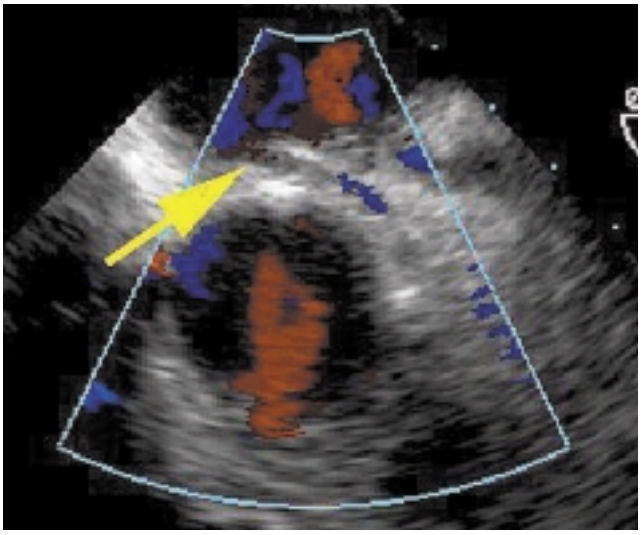


FIG. 2 Transesophageal echocardiogram demonstrating the retroaortic course of the anomalous left coronary artery (arrow).

between the aorta and the pulmonary artery (Fig. 5). Reimplantation of the left main coronary artery to the left coronary sinus of Valsalva was performed. A cardiac adenosine MRI was performed at 4-year follow-up, excluding ischemia and structural abnormalities. The patient continues to have chest pain, but different from his pain prior to surgery. Other family members were contacted and no others, including the index cases, were known to have anomalous coronary arteries.

Discussion

Coronary anomalies are rare. Their true incidence is unknown, since the majority are found during diagnostic coronary angiography or at necropsy. The anomalous origin of the left main coronary artery from the right aortic sinus of Valsalva can be subclassified into four types based on the arterial course: septal, anterior, retroaortic, and interarterial. Of these four courses, the interarterial course in which the left main coronary artery lies between the aorta and pulmonary trunk is associated with the most serious complications. During right anterior oblique ventriculography, aortography, or coronary arteriography the left main coronary artery will be "on end," anterior to the aorta, and appear as a radiopaque dot to the left of the aortic root.[3] Taylor *et al.* reported that in 60% of patients in whom the left main originates from the right coronary cusp, the interarterial course is found.[4] The most serious presentation of this anomaly is sudden cardiac death. In young patients involved in sports-related activities, coronary anomalies account for between 12 and 19% of reported sudden deaths.[5, 6] The proposed mechanism of sudden death is due to acute-angle take-off and slit-like orifice of the left main arising from the right coronary cusp. As the aorta expands with exercise, it compresses the left main coronary artery. This compression, in combination with the acute angle take-off and slit-like orifice, can lead to ischemia and sudden cardiac death. It may also cause angina, syncope, congestive heart failure, and myocardial infarction.[4]

Reports of coronary anomalies occurring in a familial clustering pattern are extremely rare. Horan *et al.* reported a father and daughter, both with a single coronary artery.[7] Bunce *et al.* reported sisters with anomalous coronary arteries identified by cardiac MRI. One sister had an anomalous right coronary artery originating from the left sinus of Valsalva and the other sister demonstrated an anomalous single coronary artery arising from the right sinus of Valsalva.[8] Rowe *et al.* reported an aberrant origin of the left circumflex artery from the right coronary cusp in a father and in both his son and daughter.[9] To our knowledge, there has been no report of familial clustering

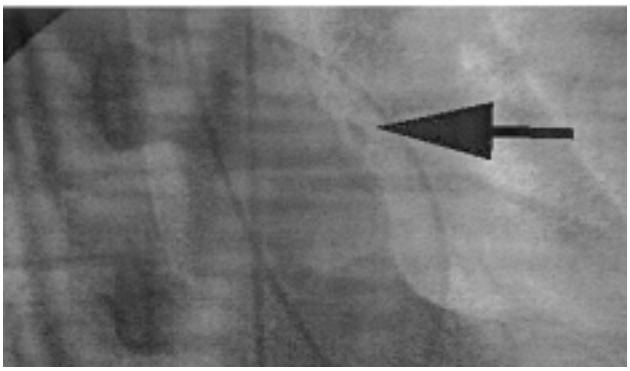
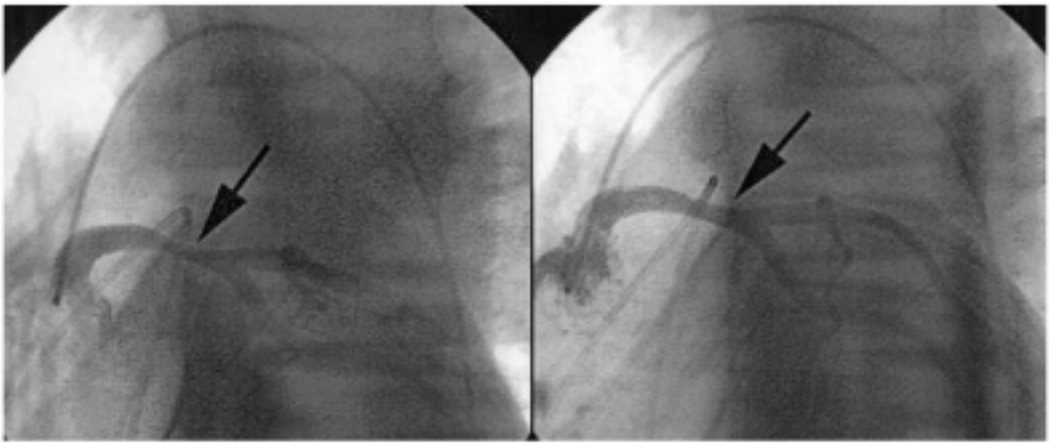


FIG. 3 A 30° right anterior oblique ventriculogram demonstrates the "Dot" sign to the right of the aortic root (arrow) indicative of the interarterial course of the left coronary artery.



(A) (B)

FIG. 4 Coronary angiogram (A) demonstrating systolic compression of the anomalous left main coronary artery (arrow). The comparative coronary angiogram (B) demonstrates resolution of the left main coronary artery (arrow).

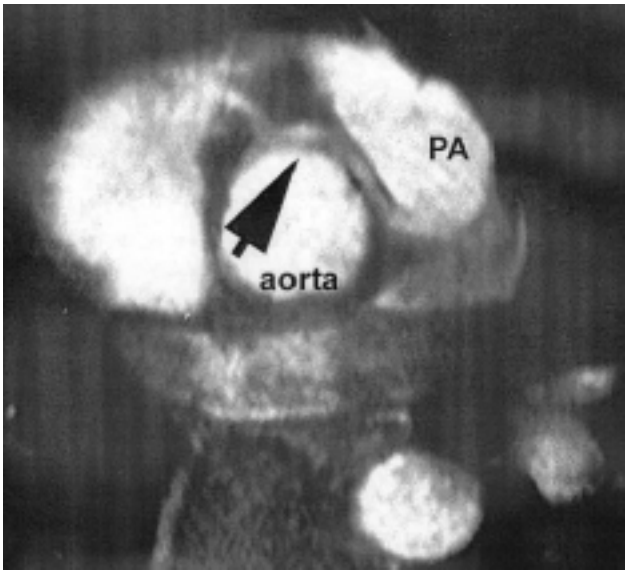


FIG. 5 Cardiac magnetic resonance imaging of the interarterial course of the left main coronary artery. Arrow indicates arterial course between aorta and pulmonary artery (PA).

involving anomalous origin of the left main coronary artery from the right sinus of Valsalva. There has been little research or reporting on the genetic patterns of this phenomenon. Leon and Bloor have studied coronary inheritance patterns in animal models, but no definitive data applicable to humans have been identified.[10] A combination of epidemiologic data collection and molecular genetic research may provide more insight in the future.

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