High-Risk Variant of a Rare Coronary Anomaly

Bryan K. Austin, MD, MS; Milind S. Shah, MD, FACC

ABSTRACT

A 61-year-old man presented with typical chest pain and was treated via acute coronary syndrome protocol. Findings on his initial diagnostic evaluations prompted cardiac catheterization. His angiographic findings were suspicious for anomalous coronary artery origin. Advanced imaging confirmed an aberrant course of the left coronary artery, with the vessel arising from the right aortic sinus of Valsalva. It was identified that the patient possessed all known high-risk features associated with this anomaly, findings not previously documented in a living adult. The patient ultimately underwent surgical revascularization to mitigate his risk for sudden cardiac death.

INTRODUCTION

The rarest of coronary anomalies, the origin of the left main coronary artery from the right aortic sinus of Valsalva (RASV), also happens to be the most lethal. This variant, which accounts for only 1.3% of all anomalies, is notorious for precipitating sudden cardiac death, with approximately half of the instances discovered postmortem.[1,2] The course of the anomalous left main coronary artery (ALMCA) is what dictates the degree of risk, with interarterial passage between the aorta and pulmonary trunk accounting for most fatalities.[3] Other established high-risk features include acute angle take-off, intramural segment, and slit-like ostium.[4,5] Here we describe a male in his seventh decade of life who presented with typical symptoms, was found to have ALMCA arising from the RASV with evidence of all 4 high-risk features, and later underwent successful surgical revascularization. To our knowledge this is the first description of a living adult patient with this anomaly possessing all of the high-risk features.

CASE PRESENTATION

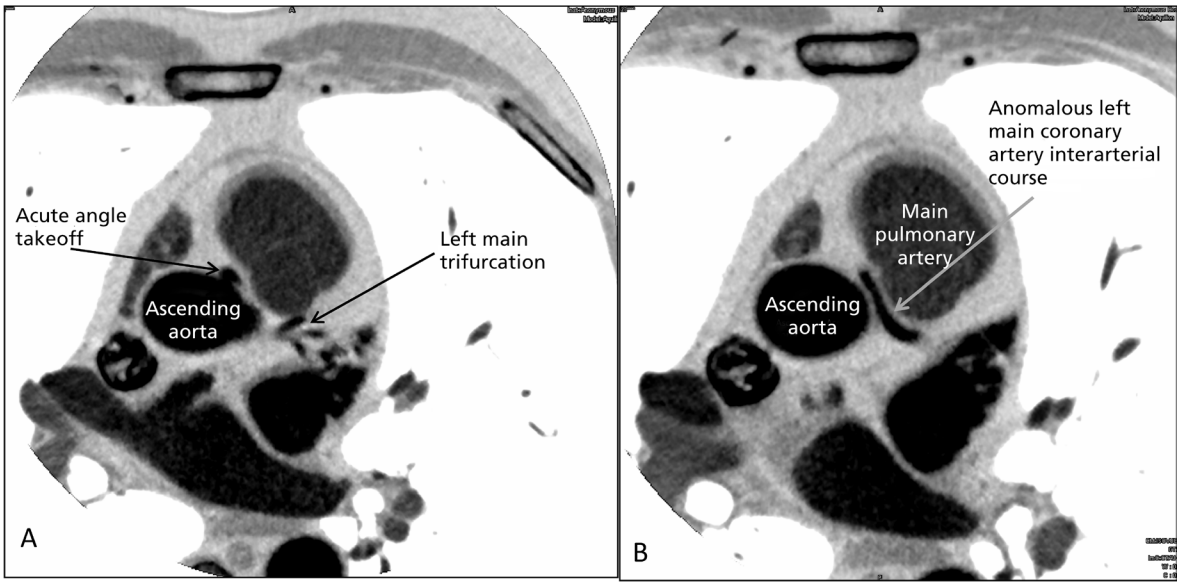
A 61-year-old, hypothyroid, dyslipidemic, nonsmoking, white man with no prior history of cardiovascular disease presented to the emergency department with 3 to 4 weeks of progressive exertional substernal chest pressure and associated dyspnea. Symptoms were most notable with lawn mowing and were relieved by rest. His initial electrocardiogram demonstrated right ventricular hypertrophy by voltage along with right axis deviation. Chest radiograph was inconspicuous. The chest pain was relieved with sublingual nitroglycerin. Heparin infusion was initiated, and the patient was admitted to the hospital with a diagnosis of unstable angina. Cardiac isoenzymes remained negative, and transthoracic echocardiogram was notable for mild right ventricular enlargement and mildly diminished right ventricular systolic function with normal estimated pulmonary artery systolic pressure. Left ventricular size and function were within reference ranges, and there were no regional wall motion abnormalities; no valvular disease was evident. Myocardial perfusion study was recommended by the consulting cardiologist, and a regadenoson nuclear scan was performed that revealed a small reversible defect in the inferolateral wall. Quantified ejection fraction was 55% to 60%. The patient subsequently underwent coronary angiography, and the dominant right coronary artery (RCA) was found to be free of any significant coronary artery disease. There was great difficulty in selectively engaging the left coronary artery, necessitating an aortogram, which disclosed an anomalous origin arising near the right coronary cusp. Despite using conventional coronary catheters, less than ideal opacification of the vessel was achieved. No high-grade obstruction was observed, but a coronary computed tomography angiogram was requested for further clarifica-

• • •

Author Affiliations: Department of Cardiovascular Medicine, Medical College of Wisconsin, Milwaukee (Austin); Department of Cardiology, Marshfield Clinic, Marshfield, Wis (Shah).

Corresponding Author: Bryan K. Austin, MD, MS, Cardiovascular Disease Fellow, Department of Cardiovascular Medicine, Medical College of Wisconsin, 8701 Watertown Plank Rd, Milwaukee, WI 53226; phone 414.805.0438; e-mail baustin@mcw.edu.

Figure. Cardiac-Gated Computed Tomography



(A) Origin and acute angle takeoff of anomalous left main coronary artery arising from the right aortic sinus of Valsalva. (B) Interarterial course of anomalous coronary artery between main pulmonary artery and aorta.

tion of the artery's course. The scan confirmed the aberrant left coronary artery, arising from the RASV, just inferior to the RCA. The vessel exhibited an acute angle takeoff (Figure [A]), then followed an interarterial course in the epicardial fat pad between the pulmonary artery and aorta (Figure [B]) before trifurcating into a left anterior descending (LAD), ramus intermedius (RI), and left circumflex (LCX) coronary artery. The LAD was free of significant atherosclerotic changes, but distal to the ostium of the first diagonal branch and first septal perforating branch there was a segment of intramural bridging. The nondominant LCX and RI were disease-free. Due to the patient's lifestyle-limiting chest discomfort and increased risk for sudden cardiac death, he was offered coronary artery bypass grafting (CABG). He successfully underwent double bypass, with the left internal mammary artery grafted to the mid-LAD and reversed saphenous vein graft to the first obtuse marginal branch of the LCX. The patient's course was notable only for postoperative atrial fibrillation, which resolved prior to discharge on postoperative day 6.

DISCUSSION

Patients with ALMCA arising from the RASV typically present with syncope, dyspnea, angina, acute myocardial infarction, or alternatively they are identified at autopsy. Symptoms usually occur with physical exertion or shortly thereafter. An interarterial course of the aberrant vessel enhances the risk profile, as does the presence of acute angle takeoff, intramural segment, and slit-like ostium. The postulated mechanism of ischemia is compression of the ALMCA by adjacent great vessels during times of cardiovascular exertion, resulting in myocardial ischemia.[3] Other established high-risk features also are thought to constrain coronary perfusion.[4,5] In 2006, Lorenz et al[6] performed an extensive literature review of the interarterial course of this anomaly. Upon examination of over 4 decades of data, they identified 104 cases of ALMCA arising from the RASV, but were unable to identify any surviving adult possessing all 4 of the high-risk features associated with sudden cardiac death.[6] A review of the literature published since 2006 resulted in similar findings, implying that this patient is unusual because of his advanced age at initial presentation, and unique due to his possession of all 4 high-risk features.

Of the 4 high-risk features described, slit-like ostium is the most difficult feature to document, even via the gold standard of coronary angiography. It has been reported that the presence of a slit-like orifice or membrane across the ostium can cause significant difficulty in engaging the ostium of the anomalous coronary, a finding exhibited by our patient. This patient's inferolateral wall perfusion defect at the time of presentation is difficult to explain given his coronary anatomy. His myocardial scan may have been a false positive. Four months following his CABG, he underwent

stress echocardiography as part of a preoperative evaluation prior to inguinal hernia repair, and this was negative.

Most patients with ALMCA arising from the RASV present with symptoms or sudden death early in life, with a mean age of reported cases in the literature of 33 years.[6] Additionally, to date, no living case of ALMCA arising from the RASV with all 4 high-risk features has been described. The patient presented here is unique in both age at presentation and in having all 4 high-risk features for cardiac ischemia and sudden death. Fortunately, the anomaly was correctable surgically, and the patient continues to do well.

Acknowledgement: The authors thank the Marshfield Clinic Research Foundation's Office of Scientific Writing and Publication for assistance in preparing this manuscript.

Funding/Support: None declared.

Financial Disclosures: None declared.

REFERENCES

1. Moustafa SE, Zehr K, Mookadam M, Lorenz EC, Mookadam F. Anomalous interarterial left coronary artery: an evidence based systematic overview. *Int J Cardiol.* 2008;126(1):13-20.

2. Safi AM, Rachko M, Tang A, Ketosugbo A, Kwan T, Afflu E. Anomalous origin of the left main coronary artery from the right sinus of Valsalva: disabling angina and syncope with noninterarterial courses case report of 2 patients. *Heart Dis.* 2001;3(1):24-27.

3. Selig MB, Jafari N. Anomalous origin of the left main coronary artery from the right coronary artery ostium – ineterarterial subtype: angiographic definition and surgical treatment. *Catheter Cardiovasc Diagn.* 1994;31(1):41-47.

4. Taylor AJ, Rogan KM, Virmani R. Sudden cardiac death associated with isolated congenital coronary artery anomalies. *J Am Coll Cardiol.* 1992;20(3):640-647.

5. Roberts WC, Shirani J. The 4 subtypes of anomalous origin of the left main coronary artery from the right aortic (or from the right coronary artery). *Am J Cardiol.* 1992;70(1):119-121.

6. Lorenz EC, Mookadam F, Mookadam M, Moustafa S, Zehr KJ. A systematic overview of anomalous coronary anatomy and an examination of the association with sudden cardiac death. *Rev Cardiovasc Med.* 2006;7(4):205-213.

advancing the art & science of medicine in the midwest

WMJ

WMJ (ISSN 1098-1861) is published through a collaboration between The Medical College of Wisconsin and The University of Wisconsin School of Medicine and Public Health. The mission of *WMJ* is to provide an opportunity to publish original research, case reports, review articles, and essays about current medical and public health issues.

Visit www.wmjonline.org to learn more.