Multimodality Imaging Assessment of Anomalous Aortic Origin of the Left Main Coronary Artery Presenting With Syncope and Non-ST Elevation Myocardial Infarction

LT Jonathan J. Cho, MC USN†; ENS Samantha D. Fabrizio, MC USNR‡; LCDR Ariana K. Tabin, MC USN†; Gilbert E. Boswell, MD†; LCDR Gregory J. Condos, MC USN†

ABSTRACT  Anomalous aortic origin of a coronary artery (AAOCA) is a rare congenital abnormality associated with myocardial ischemia and sudden cardiac death. We present a case of a 20 year old previously healthy male presenting with exertional syncope and non-ST elevation myocardial infarction. Coronary computed tomography angiography showed an anomalous left main coronary artery arising from the right coronary cusp with a slit-like appearance, acute angle origin, intramural course, and a subsequent inter-arterial course between the main pulmonary artery and the proximal aorta. Cardiac magnetic resonance imaging demonstrated myocardial infarction in the distribution of the left main coronary artery. The patient underwent successful surgical correction with unroofing of the left main coronary artery. He has had no syncopal episodes or recurrence of chest pain and returned to full duty status in the United States Marine Corps. This case report demonstrates the evaluation and management of a patient with AAOCA.

HISTORY OF PRESENT ILLNESS
A 20 year old previously healthy active duty male presented after an episode of exertional syncope. He described two prior episodes of tightness between his shoulder blades and lightheadedness while performing aerobic exercise, but no prior syncope. Following recovery from syncope, he was asymptomatic while supine at rest, but complained of mild chest pressure with minimal exertion such as walking around the room. The patient was previously an avid athlete who had recently completed U.S. Marine Corps Recruit Training without exertional angina, pre-syncope, or syncope. There was no family history of congenital heart disease, sudden cardiac death (SCD), or atherosclerotic cardiovascular diseases, including in his identical twin brother. History was also notable for recent vaccination against SARS-CoV-2. He denied tobacco, alcohol, or recreational drug use.

DIFFERENTIAL DIAGNOSIS
The differential diagnosis in this patient included acute myocarditis, acute coronary syndrome, pulmonary embolism, aortic dissection, congenital or acquired coronary artery abnormality/anomaly, valvular heart disease, hypertrophic cardiomyopathy, and spontaneous coronary artery dissection.

DIAGNOSTIC EVALUATION
On presentation, his vital signs and physical exam were normal. A 12-lead electrocardiogram demonstrated sinus rhythm with non-specific T wave abnormalities in leads V2 and V3, a new finding compared to a prior EKG. He had a normal complete blood count, basic metabolic panel, C-reactive protein, pro-brain natriuretic peptide, and D-dimer. His initial troponin-T level was 1.97 ng/mL (ref 0.01–0.029 ng/mL) and peaked at 2.63 ng/mL 9 h after initial presentation. Respiratory viral evaluations were negative, including SARS-CoV-PCR nasal swab. Serum total cholesterol was 126 mg/dL, high density lipoprotein 34 mg/dL, low density lipoprotein 71 mg/dL, and triglycerides 104 mg/dL.

A transthoracic echocardiogram (TTE) was obtained, which demonstrated normal left ventricular (LV) systolic and diastolic function, ejection fraction (EF) of 59%, normal myocardial thickness, normal global longitudinal strain, normal right ventricular size and function, and no significant valvular disease.

To evaluate the etiology of myocardial injury, coronary computed tomography angiography (CCTA) was performed. The left main coronary artery (LMCA) was found to have an abnormal origin arising from a high location above the right coronary artery (RCA) (Figs. 1 and 2). The LMCA followed an inter-arterial course between the pulmonary artery and the proximal aorta (Figs. 1A and 2). The morphology of the LMCA was slit-like, with an acute angle origin (<45°), and followed an intramural course within the wall of the proximal aorta (Fig. 1B). Due to this anomalous aortic origin, his
left main coronary artery was severely stenotic with minimal luminal area within the intramural segment measured 5.5 mm² (Fig. 3A–D). The coronary system was right dominant, with a large posterolateral branch supplying the inferolateral wall.

To further investigate whether the cause of his symptoms and myocardial injury were attributed to the anomalous aortic origin of the left main coronary artery, he underwent cardiovascular magnetic resonance (CMR) imaging, which demonstrated mildly reduced LV EF (50%) and mild hypokinesis of the apical anterior left ventricle. Persistent subendocardial late gadolinium enhancement (LGE) involving up to 25% thickness of the LV wall was present in the mid to distal anterior and septal walls and basal lateral wall (Fig. 4A–D). Edema noted on T2 weighted imaging corresponded to these same territories. These abnormal areas of infarction correlated well with the area of myocardium subtended by the LMCA.

**DISCUSSION**

Anomalous aortic origin of a coronary artery (AAOCA) is the second leading cause of SCD in young athletes in the United States after hypertrophic cardiomyopathy. Presentation is variable and can include SCD, aborted SCD, and symptoms related to exertion such as chest pain, palpitations, pre-syncpe, or syncpe. Not infrequently, AAOCA is diagnosed incidentally in an asymptomatic patient. Chest pain is considered ischemic if accompanied by evidence of myocardial injury such as elevation in cardiac markers, ST segment abnormalities with exercise, ventricular arrhythmias, evidence of wall motion abnormality, or perfusion defects on nuclear scan or CMR in the distribution of the anomalous coronary artery. Treatment of AAOCA varies depending on the anatomic characteristics and risk stratification of the anomalous coronary artery and may include observation, activity modification, medical management, or surgical correction. Multiple options for surgical management have been developed, including coronary artery bypass grafting, coronary reimplantation, and unroofing of the intramural coronary segment.
Imaging Assessment of Anomalous Coronary Artery

FIGURE 3. Gated Cardiac CT. (A) and (B) Orthogonal curved planar reformatted images of left main and left anterior descending coronary artery. (C) and (D) Cross-sectional images of the proximal left main coronary artery indicating severe left main stenosis. Minimal luminal area within the intramural segment measured 5.5 mm². Note the oval configuration. (E) Volume rendered image of the left ventricle showing course of the LAD and large diagonal branches, distal to the small caliber left main coronary artery.

We present a case of a previously healthy 20 year old male with exertional syncope and angina that demonstrates use of multimodal imaging in the diagnosis and management of AAOCAs. TTE is recommended as the first line diagnostic modality due to its non-invasive, rapid, low cost, and widely available nature. TTE eliminated hypertrophic cardiomyopathy and valvular disease as likely etiologies in our case. However, low detection rate, lack of detailed characterization of AAOCAs features, and limited spatial resolution restrict the utility of TTE for diagnosis of AAOCAs. This case report further supports the recommendation to obtain further imaging if AAOCAs cannot be ruled out or confirmed with certainty.

We next performed CCTA that effectively identified the presence of AAOCAs. Nevertheless, diagnostic uncertainty remained because until recently, the patient had demonstrated very high fitness level despite his congenital abnormality. In light of his recent vaccination against SARS-CoV-2, myocarditis remained on the differential diagnosis and we felt CMR would best differentiate between these two competing causes. CMR clearly showed recent myocardial infarction with subendocardial LGE and edema in the LMCA coronary artery distribution, thus implicating the anomalous LMCA in his presentation. Definitive surgical management of his AAOCAs was warranted due to his young age, demonstrated infarction, and likelihood of repeat injury absent a significant reduction in physical activity for the rest of his life.

This case report is novel in that it is the first to describe the use of CMR to detect ischemia related to anomalous aortic origin of the LMCA. The frequency of inter-arterial anomalous left coronary artery is rare, with weighted prevalence of 0.03%, compared with that of inter-arterial anomalous right coronary artery, with weighted prevalence of 0.23%.

Of the case reports utilizing CMR to definitively characterize myocardial ischemia, all were attributed to anomalous aortic origin of right coronary artery. This case report, along with previous reports that showed myocardial ischemia on CMR caused by anomalous aortic origin of right coronary artery, supports the notion that CMR should be utilized to evaluate myocardial infarction in the setting of suspected AAOCAs. It is also worth noting that the entire pre-operative evaluation was non-invasive, with high quality CCTA negating the need for invasive angiography.

This patient’s rare anomalous aortic origin of the left main coronary artery demonstrated all of the most frequently identified high-risk features. This case supports the proposed fixed (slit-like ostium) and dynamic (acute angle origin and intramural segment) pathophysiology underlying the mechanism of
of variable myocardial ischemia by anomalous aortic origin of coronary artery arising from the opposite coronary sinus. Although this patient was previously an avid athlete without angina, pre-syncpe, or syncope associated with exercise, the recent high intensity exercise associated with military training appeared to have precipitated his MI. This supports evidence that myocardial ischemia associated with AAOCA may be intermittent and difficult to diagnose, and that additional factors, such as the type of physical activity (isotonic [cycling or running] vs. isometric [weight lifting]) and volume status may contribute. Fortunately, our patient had an excellent outcome and was able to resume his military career.

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REFERENCES