CASE REPORT

Can Transthoracic Echocardiography Predict Anomalous Origin of the Left Circumflex Coronary Artery in Adults? A Case Report

*Abdul Wadud Chowdhury¹, Gobinda Kanti Paul², A K M Monwarul Islam¹, Kaniz Fatema Ananya¹

¹Department of Cardiology, National Institute of Cardiovascular Diseases and Hospital, Sher-E-Bangla Nagar, Dhaka, Bangladesh, ²Department of Cardiology, Mymensingh Medical College & Hospital, Mymensingh, Bangladesh

Abstract

Congenital coronary artery anomaly (CAA) is a rare entity. Recognition and precise detection of the CAA is essential for correct management of the condition. Various diagnostic techniques can be used to detect CAA and to assess the presence of high-risk features associated with CAA. Coronary computed tomography angiography (CCTA) is currently considered the gold standard test for diagnosing CAA and cardiac magnetic resonance (CMR) is considered as an alternative but both the techniques are expensive and not widely available in every corner of our country. Transthoracic echocardiography (TTE) is regarded as a key tool in the detection & diagnostic workup of CAAs in children, in whom the optimal acoustic windows usually allow the visualization of coronary ostia and there is no risk of radiation exposure. In adults, TTE plays a minor role in detecting CAA because here TTE can't identify coronary ostia clearly. Moreover, in adults it is difficult to visualize the course of coronary artery & it's relations with the great vessels by TTE. In spite of all limitations; in some adult cases, identification of two signs in TTE can predict anomalous origin of coronary artery (AOCA). These two signs are "RAC sign" (retro aortic anomalous coronary) and "Crossed aorta sign". Therefore, visualization of RAC sign & /or "Crossed aorta sign" in TTE can predict the presence of anomalous origin of left circumflex coronary artery (LCX). In this middle-aged woman while performing TTE, we found these two signs (RAC sign & Crossed aorta sign) and predicted her to have anomalous origin of LCX. Subsequently coronary angiogram (CAG) confirmed the diagnosis. So, careful echocardiographic observation may alert the interventional cardiologist to pay special attention during coronary catheterization to find out AOCA.

Key words: Anomalous origin of left circumflex artery (LCX), crossed aorta sign, retro aortic anomalous coronary (RAC)

Introduction

Actual prevalence of AOCA is unknown in general Bangladeshi population. CAA was found in 0.6% patients undergoing CAG at Bangladesh Medical University, Dhaka from 2004 to 2007. Among all CAA, 87.5% had AOCA¹. Antopol and Kugel first described anomalous origin of the LCX from the proximal right coronary artery (RCA) or right sinus of Valsalva (RSV) in 1933². It was found to be most common AOCA (58.3%) in an observational study conducted in Turkey from 2001 to 2011³. The relative frequency of this

anomaly demands a high level of anticipation during the performance of selective CAG which is possible to anticipate previously from TTE. This case will highlight the potential of echocardiography to identify abnormal origin of the LCX from the RCA.

Case Report

A 43 years old hypertensive, diabetic, postmenopausal woman presented with central, compressive chest pain for 16 hours at emergency department. On enquiry, she gave same type of chest pain and hospitalization 1 month back. She was on regular anti-ischemic treatment for the last 1 month. Her father died of stroke 10 years back.

Her blood pressure was 150/100 mm Hg, pulse was 94 beat/minute, respiratory rate was 16 breath/minute, jugular vein was not engorged, breath sound was

*Correspondence: Abdul Wadud Chowdhury, Department of Cardiology, National Institute of Cardiovascular Diseases and Hospital, Sher-E-Bangla Nagar, Dhaka-1207. Bangladesh.

Email: dr.wadud65@gmail.com ORCID ID: 0000-0003-4930-1448 vesicular. Cardiovascular system examination revealed no abnormality.

Serial troponin-I was normal. So, we diagnosed her as a case of unstable angina (Intermediate risk).12 lead electrocardiography (ECG) showed left ventricular hypertrophy and non-specific ST-T changes in V2-V5. Her thrombolysis in myocardial infarction (TIMI) risk scoring was 3 points-Intermediate risk (13% risk of all-cause mortality, new or recurrent myocardial infarction or severe recurrent ischemia requiring urgent revascularization at 14 days).

TTE revealed interventricular septal thickness 15mm, posterior wall thickness 14mm, no regional wall motion abnormality, good left ventricular systolic function (Left ventricular ejection fraction 60-65%). Incidentally, we found a highly echogenic tubular structure, located slightly on the atrial side of atrioventricular groove in apical 2 chamber and apical 4 chamber view (RAC

sign) (Figure 1 & Figure 2). This was not thought to be an artifact because it was seen with a different appearance in other planes. Although it was tubular in appearance and suggestive of a vascular structure, it did not correlate with the typical location for any normal vessel. In parasternal short axis (PSAX) view at the level of great vessel, left main coronary artery and RCA were arising from their respective sinuses. The origin of the anomalous LCX was not seen in PSAX view. In apical 5 chamber view a binary structure was seen crossing the aorta perpendicular to its long axis-'crossed aorta sign'(Figure 3 & Figure 4).

According to American Heart Association (AHA) guideline of unstable angina with intermediate risk-we proceeded to diagnostic CAG. We selectively catheterize the left coronary sinus and found left main coronary artery (LM) and left anterior descending artery which was normal. But where was LCX? Yes, it could not be visualized from any conventional angiographic



Figure 1: Apical 2 chamber showing RAC sign.

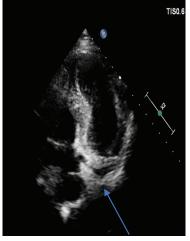


Figure 2: Apical 4 chamber showing RAC sign.

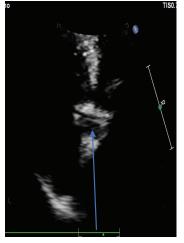


Figure 3: Apical 5 chamber showing crossed aorta sign.



Figure 4: Crossed aorta sign in apical 5 chamber view (TTE).



Figure 5: Origin of LCX from RCA



Figure 6: Origin of LCX from RCA.

views from left side. After locating the ostium of the RCA, origin of LCX was visualized from the proximal RCA (Figure: 5 & 6). Also, in a right anterior oblique projection, the anomalous LCX was seen in profile behind the aortic root as it coursed to the left atrioventricular groove. RCA was ectatic. Probably sluggish or turbulent flow within ectatic RCA produced symptoms of myocardial ischemia.

Discussion

The etiology of coronary anomalies is uncertain. There are no definite inheritance pattern and no sex predominance. AOCA has a wide spectrum of clinical presentation. Although anomalous retroaortic course of LCX is believed to be mostly benign, there have been reports of morbidity and mortality. The origin of the left coronary artery from the contralateral sinus, course of left main coronary artery between the aorta and right ventricular outflow tract, single coronary artery, origin of coronary artery from a pulmonary artery, coronary artery fistula and atresia of the coronary ostium has been clearly shown to be associated with adverse clinical outcome. Origin above sinotubular junction, acute angle take-off causing flow limitation during exercise are another high-risk feature of AOCA. AOCA arising from the aorta are a recognized cause of myocardial ischemia and sudden death. Death has been precipitated by exercise in most cases⁴. In many cases, conventional CAG may fail and multidetector computed tomography (MDCT) may be needed to delineate the origin and course of LCX. There are reports of chest pain, myocardial infarction and sudden cardiac death related to AOCA in the literature^{5,6}. These manifestations might be due to repeated compression of the anomalous artery by a dilated aortic root or to unusual angling as a result of the retroaortic course of the LCX, which can compress the coronary ostium and restrict blood flow. The variability of clinical presentation and prognosis of coronary anomalies depend on the proximal course of the anomalous LCX in relation to the great vessels which can be predicted by TTE where invasive catheterization is not available. Moreover, if any patient with RAC sign & crossed aorta sign in TTE have no guideline directed indication of CAG; we may predict the diagnosis of AOCA on the basis of noninvasive echocardiography only where MDCT is not available. Again, early detection and exact delineation of their proximal course are crucial in case of preoperative evaluation. Also, during CAG interventional cardiologist will invest less dye and less contrast material when LCX will not be visualized to take origin from left coronary sinus. Two angiographic signs have been proven reliable in recognizing the anomalous artery before its selective demonstration. These signs are a

profile view of the artery behind the aortic root during left ventriculography (the "aortic root sign") and recognition of absent arterial inflow to a significant area of the posterior lateral left ventricle during selective injections of the main left coronary artery (the "sign of non-perfused myocardium")7. From the apical fourchamber view, tilting the transducer to a shallower angle to a more anterior scan plane exploring the retro aortic region, a binary structure may be found above the mitral valve plane, in the atrioventricular groove, directed toward the right coronary sinus and overlapping the aortic root (RAC sign). A binary structure that crosses the aorta perpendicularly in apical five chamber view is called Crossed aorta sign. On the other hand, a rounded structure in the mitroaortic angle, visible best on transesophageal echocardiography (TEE) is termed as 'Bleb sign'. A small circle-like structure beneath the noncoronary cusp in the TTE at PLAX view may be seen similar to the bleb sign on TEE. A retrospective, investigator blinded, case-control study conducted at Mayo clinic cardiovascular department from 2005 to 2015 revealed that the RAC sign was seen in 63.3% patients with confirmed coronary anomalies and 6.1% with normal coronary anatomy. The sensitivity of the RAC sign was 63.3%, and the specificity was 93.9%⁸. Though TEE is an alternative and semi-invasive method capable of identifying AOCA; it has some limitations. TEE can't visualize the entire course of the coronary arteries and the level of yield vary with the experience of the sonographer, and invasiveness. Patient's discomfort might result in possible intolerance and post examination side effects (arrhythmias, damage to the esophageal mucosa, difficulty swallowing, chest or abdominal pain, vomiting). TTE is a valid alternative, being a fast, less expensive, noninvasive method requiring no ionizing radiation to suspect anomalous LCX origin from RCA. To minimize misdiagnoses, a suggestion would be to identify first the best-known RAC sign, described as a binary structure above the mitral valve plane directed toward the right coronary sinus in a modified four-chamber apical view. This view, obtained by tilting the transducer to obtain a more anterior scan that explores the retroaortic region. In TTE, the coronary sinus may be confused with a retroaortic course of LCX because both structures appear as a posterior vessel near the left atrium, but the key difference is that the coronary sinus is typically located more posteriorly, lower course than the atrioventricular sulcus, and thinner walled compared to the thicker walled LCX, especially when it takes a retroaortic path. So, to differentiate RAC sign from coronary sinus, careful analysis of different imaging plane is needed. The RAC sign must be distinguished from aortic valve calcifications, which instead have a consensual movement to that of the valve and also do not have internal anechogenicity. However, its simple identification, even in patients with poor acoustic windows, makes it the basis for the diagnosis.

Conclusion

RAC sign and crossed aorta sign found in TTE can predict the anomalous origin of LCX in adults. Methods like CCTA, MDCT are expensive; CAG is invasive. These are less available and expose patients to ionizing radiation and iodinated contrast agents. Failure to recognize and failure to properly demonstrate the coronary artery anomaly can be hazardous to patient management. Prediction of CAA can prevent catastrophe during coronary artery or prosthetic valve surgery. TTE may be a most accessible, available screening method to describe the RAC sign and crossed aorta sign. So, a careful echocardiographer having previous sensitization regarding RAC sign & crossed aorta sign may alert the interventional cardiologist to have preparation for difficulties which may arise during catheter engagement in CAG. However, mere incidental presence of RAC sign & crossed aorta sign in TTE should not allow arrangement of CAG or MDCT or CCTA in asymptomatic cases.

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