

Anomalous Origin of the Left Main Coronary Artery with an Intraseptal Course Identified by Transesophageal Echocardiography and Role of various diagnostic modalities

M Kanjwal, R Bashir

Citation

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Abstract

A 41-year old male was seen in the cardiology clinic for risk assessment for a kidney and pancreatic transplant. Myoview stress test showed transient ischemic dilation and mild fixed anterior and inferior wall defects. Subsequent coronary angiography showed left main coronary artery arising from the right coronary cusp having an intra-septal course before bifurcating into anterior descending and left circumflex artery. The pre-operative transesophageal echocardiography confirmed the intra-septal course of the ectopic left main coronary artery. The intra-operative anatomy of this vessel also revealed a short pre-pulmonic course followed by an intraseptal course noted as the vessel dived into the septum.

INTRODUCTION

Congenital anomalies of the coronary arteries occur in 0.2-1.2% of the general population^{1, 2, 3}. Clinical significance of such findings has been speculated and a few variants implicated as a cause of sudden death, especially in young athletes⁴. The left main coronary artery arising from the right sinus of valsalva is found in 0.03%². Most of the reports of anomalous origin of coronary arteries have come from autopsy studies or using coronary angiography. The current diagnostic method of choice for detecting coronary artery anomalies is conventional x-ray coronary angiography. However, x-ray angiography provides only a two dimensional view of a vessel's complex three dimensional path, so the anatomic course of anomalous vessel with respect to the aorta and pulmonary artery may be difficult to discern. In addition, anomalous vessel may be overlooked or assumed to be occluded if not selectively engaged⁵.

Advances in non-invasive diagnostic modalities show a role of Transesophageal (TEE), Nuclear studies, MR Angiography and CT Angiography in recognizing such anomalies. We present a patient with an anomalous origin of the left coronary artery from the right sinus with an intraseptal course and the role of various modalities that could identify the same.

CASE HISTORY

A 41 year old male was referred to the out patient cardiology clinic for pre-operative risk assessment for a kidney and pancreatic transplant. Coronary risk factors included smoking, diabetes mellitus (with stage I diabetic retinopathy), hypertension, hypercholesterolemia, anticardiolipin antibody (with a history of stroke) and end stage renal disease. The patient was receiving hemodialysis thrice a week. Symptoms were significant for left shoulder pain and dysnea on exertion (NYHA 3). Physical examination was unremarkable except for the presence of a patent AV fistula on his left arm.

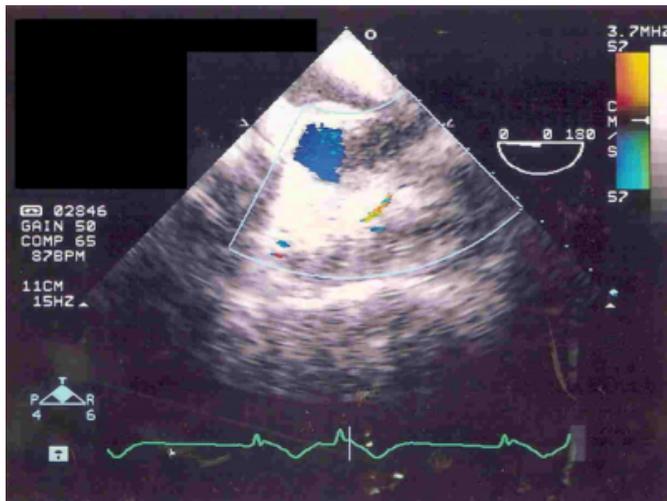
Electrocardiogram showed normal sinus rhythm with left ventricular hypertrophy and a strain pattern. Trans Thoracic echocardiography (TTE) revealed grossly normal left ventricular systolic function with mild diastolic dysfunction. There was no evidence of resting wall motion abnormality. A Persantine myoview stress test showed transient ischemic dilation along with a left ventricular function of 45%. There were also fixed anterior and inferior wall defects, with hypokinesis of the anterior, inferior, posterior and septal regions. Patient underwent a coronary angiography that showed an anomalous origin of the left main coronary artery from the right coronary cusp with, an initial short intra septal course.

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There was a tubular 40% stenosis noted in the proximal part of this vessel and 70% stenosis in the mid left anterior descending artery. The left circumflex artery was non obstructive while the right coronary artery was dominant and was totally occluded distally with the posterior descending filling with left to right collaterals. As a percutaneous intervention was not feasible secondary to the abnormal anatomy of the coronaries, the patient underwent a coronary artery bypass graft. An intra operative transesophageal echo (TEE) was able to delineate the anomalous origin and the intra septal course of the left main coronary artery (fig 1). The intra-operative TEE also revealed a short pre-pulmonic course before it angled into a short intraseptal course. The left main divided into the LAD and the LCX intra septally. The patient had a left internal mammary artery graft to the left anterior descending and a saphenous vein graft to the right coronary artery. The patient did well after the procedure and recovered to go on for the transplant.

Figure 1

Figure 1: TEE showing intraseptal course of anomalous left main coronary artery.



DISCUSSION

The left coronary artery arising from right sinus of valsalva is a rare congenital anomaly. In a prospective study of 1950 angiograms at Texas Heart Institute, 0.15% showed a Left coronary artery arising from the right sinus. Other studies have also shown association of such a condition with sudden death.⁴ The mechanism responsible for such an adverse outcome is ischemia secondary to compromise of flow and factors like abnormalities of the coronary ostium,⁴ an aortic intra mural course and the initial course have been held responsible for the later. Yet, Yamanka and Hobbs⁶ found that most coronary anomalies are asymptomatic and are

discovered as incidental findings on coronary angiography. In fact many studies found that especially the initial pre-aortic anomalous path as a culprit for clinical ischemic manifestations and/ or sudden death^{4, 7}. Due to lack of a substantial database, it is unclear at this point if the mere presence of an anomalous coronary anatomy has a higher clinical risk.

After arising from the contra lateral sinus of valsalva, an anomalous coronary artery can take one of at least five pathways⁸: retrocardiac, retro aortic, preaortic (also called inter-arterial), intraseptal, and prepulmonic. Few authors consider the presence of only four variants⁹. Our patient had an initial pre-pulmonic course leading into an intra septal course. The presence of an inter-arterial course may lead to ischemia⁷, syncope and sudden cardiac death^{10,11} in the absence of coronary artery disease. Kothari et al¹² reported a patient with septal course of the left main coronary artery arising from right aortic cusp presenting with resistant ventricular tachycardia. The tachycardia resolved after CAGB to otherwise normal left anterior descending and circumflex arteries.

In the past, coronary angiography or autopsy were the only means by which these conditions could be diagnosed. However, improvement in ultrasound technology now allows delineation of coronary artery anatomy in many patients using TEE. Anomalous origin of left main from the right sinus and anomalous origin of right coronary artery from the left sinus of valsalva have been described using TEE^{13, 14, 15, 16}. Two dimensional TTE must be combined with color Doppler flow mapping to successfully identify anomalous origin of coronary artery¹⁴. TTE however fails to delineate AOCA with a course posterior to aorta and AOCA with intra mural course. TEE has been shown to be superior to TTE in diagnosis AOCA with retro-aortic course^{13, 15, 16}. Gaither NS et al¹⁴ in their report of five patients with anomalous origin of coronary artery, which were studied by monoplane TEE and selective coronary angiography. TTE was also performed in four of these five patients. Anomalous coronary ostia were visualized in four of five patients utilizing TEE, but in none of four patients by TTE, a proximal segment of an anomalous coronary vessel was identified in all five patients by TEE and only two of four patients by TTE. F Fernandes et al¹⁵, published series of nine coronary anomaly patients where transesophageal echocardiography was used to confirm the origin and initial course of the anomalous vessel found on angiography. They

found that TEE could demonstrate the course of anomalous left main coronary artery between the aorta and pulmonary artery better than angiography⁵. The image acquisition and analysis in this study was performed unblinded to the angiographic findings. Thus the ability of transesophageal echocardiography to identify coronary anomalies in a blinded analysis is unknown.

We could delineate AOCA in our patient using TEE. Color Doppler is particularly useful in diagnosing AOA with intramural course because the technique can give additional information of direction of flow of intramural segment that helps differentiate whether AOCA is from right or left sinus of valsalva. Yet, most of the TEE studies were done after initial identification of the anomalous coronaries and hence is difficult to interpret the utility of a TEE for primary screening of such a condition.

Electron beam computed tomography has been used to image coronary arteries noninvasively but experience with this modality remains very limited. Dieter Ropers et al,¹⁷ reported four cases variants of left anterior descending originating from right sinus of valsalva, where electron beam tomography was performed using C-150 XP EBT scanner. They could delineate Interarterial, Retroaortic, Prepulmonic and septal or subpulmonic course of left main artery illustrating the potential of contrast enhanced electron beam tomography to visualize the exact 3 dimensional course of anomalous coronary arteries.

MRI study using breath holding gradient echo technique has been used to delineate aberrant origin of the RCA from the extreme leftward aspect of the aorta, superior, posterior and lateral from its normal position. McConnell et al,¹⁸ performed breath hold transverse and breath hold oblique 2D coronary MR angiography in 16 patients with anomalous coronary arteries. They could identify 14 of 16 anomalous coronary arteries; one patient was unable to hold breath and image quality in another was inadequate. The mean imaging time was 50 minutes however some patients are unable to do repetitive breath holding and this inability can lead to section misintegration and respiratory motion artifacts. New, nonbreathhold method for respiratory motion compensation could accommodate for broader range of patients and would allow enhanced spatial and temporal resolution, and fewer registration errors between slices. Nicholas H, et al,¹⁹ studied 26 patients suspected of having coronary anomalies. Multiple 3D volume slabs were acquired at the level of Sinus of valsalva by using

diaphragmatic navigators for respiratory artifact suppression; they found free breathing MR angiography superior to conventional coronary angiography in delineating the course in anomalous artery between aortic root and RVOT.

It is important to correctly diagnose a vessel that passes between the aorta and the pulmonary artery or right ventricular outflow tract. This anomaly is associated with chest pain and sudden cardiac death. Although the actual pathophysiology remains unclear however a slit like orifice or an acute angled intramural segment that is compressed between high pressure aorta and the RVOT during exercise.

MR coronary angiography has certain potential clinical application. It can evaluate the three dimensional path of anomalous coronary vessel identified by xray angiography, especially when the vessel is having a course between aorta and RVOT. It can also differentiate an occluded vessel from an anomalous one in cases in which a vessel cannot be engaged by conventional angiography. And last but not the least it could potentially be used in young patients with unexplained syncope or chest discomfort in whom coronary anomaly is part of differential diagnosis.

An interesting thing that was seen in our patient was transient ischemic dilatation on persantine myoview stress. This might be a unique finding in patients with AOCA with non-obstructive coronary artery and should warrant further investigation. In our patient, coronary angiography also failed to delineate the intraseptal course, which was further delineated by using TEE.

We conclude that TEE is an adjunctive imaging modality in delineating the anomalous origin of left main arising from right coronary cusp with an intra septal course.

CORRESPONDENCE TO

Mohammad Khalil Kanjwal MD.
Clinical Instructor of Medicine.
Department of General Internal Medicine
University of Toledo Medical Center.
3000 Arlington ave Toledo OH 43614
Tele: 419-383-5388/4909
Fax 419-383-6252.

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Author Information

Mohammad Khalil Kanjwal, MD

Riyaz Bashir, MD