Coronary arteriovenous malformation, as imaged with cardiac computed tomography angiography: A case series

Tigran Khachatryan¹, Sudha Karnwal¹, Yasmin S. Hamirani¹, Matthew J. Budoff¹*

¹. Los Angeles Biomedical Research Institute at Harbor - UCLA Medical Center, Torrance, CA, USA

*Correspondence: Matthew Budoff, MD, Los Angeles Biomedical Research Institute at Harbor-UCLA Medical Center, 1124 W. Carson Street, Torrance, California, 90502, USA (mbudoff@labiomed.org)

ABSTRACT

Introduction: Coronary Arteriovenous Malformations (AVM) can lead to various cardiovascular events. The severity of clinical manifestation depends on the degree of the malformation. The significance of major arterial shunt is that they can deprive the myocardium of the necessary amount of blood, leading to myocardial ischemia, and potentially tachycardia, left ventricular dilatation and heart failure secondary to increased stroke volume and cardiac output. Cardiac computed tomography (CCT) has proven to be a good non-invasive diagnostic tool in the detection of coronary Arteriovenous Malformations.

Case Presentation: We include 4 patients, who presented with various symptoms in relation to the coronary Arteriovenous Malformations. Diagnostic confirmation was made non-invasively using multi-row detector cardiac computed tomography (MDCT).

Conclusion: Coronary Arteriovenous Malformation is a rare congenital disease which can produce symptoms of varying severity depending on the size of the malformation. CCT angiography can help in non-invasively diagnosing the malformation along with revealing the anatomic details, which can be used for surgical planning, or for better medical management of the patients.

CASE SERIES

INTRODUCTION

Arteriovenous Malformation (AVM) is an abnormal connection between arteries and veins, and are termed arterio-arterial (AA) or arterio-venous (AV) malformations when the malformation is between two arteries or an artery and a vein, respectively. These connections can be congenital, occurring in a variety of sizes and in different locations (1,2,5). Coronary AVM also called coronary AV fistula, consists of abnormal communication between coronary arteries and one of the chambers of the heart (coronary-cameral fistula) or adjacent vessels (1,2,3,4). The presence and the extent of the effects and symptoms depend on the size of the coronary AVM, ranging from asymptomatic to myocardial necrosis, bacterial endocarditis and heart failure. In many cases, patients are asymptomatic (2,5,21). However, in case of large malformations, which lead to large shunts from the arteries to the veins, the myocardial tissue could be severely deprived of blood and the patient will present with severe symptoms and even death. We are presenting four cases of coronary AVMs that were detected with cardiac computed tomography (CCT) angiography.
CASES PRESENTATION

**Case 1:** A 69 year old white male with history of hypertension, diabetes mellitus and coronary artery disease (CAD), status post percutaneous transluminal coronary angioplasty (PTCA) and stenting of the left circumflex (LCX) and left anterior descending (LAD) arteries was admitted for further intervention of LCX to pulmonary artery fistula. The patient previously had closure of a fistulous connection from the LAD to the pulmonary artery using a combination of coiling and covered stent placement in the proximal LAD. Patient had no recent history of chest pain or shortness of breath. Previously he had smoked one pack of cigarettes a day for thirty five years which he reduced to six cigarettes a day. Upon examination patient had a regular S1 and S2 with grade 2/6 ejection systolic murmur at the aortic area. No carotid bruit was appreciated. The CCT angiography revealed a coil in the septal perforator and a persistent AV malformation with flow around the stent in LAD (Figure 1 & 2). The AVM connected the LAD and pulmonary artery and was partially occluded. Another malformation connected left main coronary artery (LMCA) to the pulmonary trunk (Figure 2). Smaller circumflex AVM was present proximally. There was a larger connection of the AVM to the pulmonary trunk from a separate origin in the aorta which might be a conus branch (Figure 1). It was later decided not to do any further intervention for the Arteriovenous Malformations on this patient and he was continued on medical management. At one year of follow up patient did not have any symptoms due to his coronary AVM.

**Case 2:** A 34 year old Caucasian female presented with exertional chest pain and abnormal echocardiogram, the report of which was not available. The patient had a family history of coronary artery disease. She maintained an active but stressful lifestyle. The medical history did not reveal any other significant findings. Her electrocardiogram showed normal sinus rhythm with no ST, T changes. Laboratory examination including complete blood picture and comprehensive metabolic panel and three sets of cardiac enzymes were normal. A 64 detector cardiac computed tomography CCT angiography, high resolution volume study was performed to rule out anomalous coronaries. The CCT revealed a small AVM from the RCA to the pulmonary artery (Figure 3). There were no other significant findings. She was discharged with 3 months clinical follow up revealing no recurrence of her chest pain.

**Case 3:** A 56 year old Hispanic female with history of hypertension presents with a known coronary arterial fistula for evaluation with cardiac CT angiogram before closure. She described her life style as sedentary. The medical history was otherwise non-significant. A 64 detector cardiac computed tomography (CCT) angiography, high resolution volume study was performed. The CCT revealed a large (2.5mm) coronary fistula from the LAD to the main pulmonary artery. The fistula was evident due to the back flow of contrast (step up) in the pulmonary artery (Figure 4). The patient underwent surgical closure of her fistula afterwards.

**Case 4:** A 70 year old Hispanic female with history of hypertension, hyperlipidemia and diabetes mellitus was referred for possible coronary artery aneurysm. She had a one week history of difficulty sleeping, eating, and worsening shortness of breath on exertion. The patient had an unknown heart surgery 30 years and had rheumatic fever as a child. Both trans-thoracic and trans-esophageal echocardiogram (TEE) revealed a patent foramen ovale with right to left flow, which was confirmed with a bubble study (Figure 5 a). There was a severe amount of mitral regurgitation with posteriorly directed mitral regurgitent jet and a severe amount of tricuspid regurgitation (Figure not shown). Apart from the valvular regurgitation due to rheumatic valve involvement, there was suspicion of aneurysm left sided coronary artery (Figure 5 b). The patient was referred to our center to get an outpatient CCT angiography using Electron beam computed tomography (EBCT) performed. The EBCT revealed a massive AVM between RCA and coronary sinus (Figure 6). Furthermore, there was severe aneurysmal disease of RCA and LCX. The LAD and LM were normal. The patient was evaluated for mitral valve repair at that time. However, given the patient's unusual coronary anatomy, it was thought that a mitral valve repair would alter her hemodynamics in a negative way. Therefore, given that the patient was asymptomatic able to ambulate, and was without symptoms of heart failure, it was decided to continue medical treatment of her mitral regurgitation at that time and to follow up in the cardiology clinic as well as with her primary care physician.

**Cardiac Computed Tomography (CCT) image acquisition protocol at our center:**

CCT scan was performed with a 64- detector row Light speed VCT scanner (GE Healthcare, Milwaukee, WI). Individuals presenting with baseline heart rates >65 beats per minute (bpm) were administered oral beta-blocker therapy as the preferred method for slowing down the heart rate. IV administration was allowed in the protocol, using intravenous metoprolol at 5 mg increments to a total possible dose of 40 mg in order to achieve a resting heart rate <65 bpm. Following a scout radiograph of the chest (anteroposterior and lateral), a timing bolus (using 10-20 cc of contrast) was performed to detect time to optimal contrast opacification in the axial image at a level immediately superior to the ostium of the left main artery. Nitroglycerin 0.4 mg sublingual was administered immediately prior to contrast injection. During CCT acquisition, 80cc iodinated contrast was injected utilizing a triple-phase contrast protocol: 60cc iodixanol 'Visipaque', followed by 40cc of a 50:50 mixture of iodixanol and saline, followed by a 50cc saline flush. Retrospective ECG gated helical contrast-enhanced multi-detector row cardiac computed tomography angiography (MDCTA) was performed, with scan initiation 20 mm above the level of the left main artery to 20 mm below the inferior myocardial apex. The scan parameters were 64x0.625mm collimation, tube voltage 100 to 120 mV, effective mA 350- 780 mA. Radiation reduction algorithms using electrocardiography (ECG) modulation were employed, which reduces radiation exposure (mA) during systole and end-diastole. After scan completion, multiphasic reconstruction of MDCTA scans was performed, with
reconstructed images from 70 to 80% by 5% and 5 to 95% by 10% increment.

**MDCTA Interpretation**

MDCTA images were interpreted by a reader with more than 10 years experience in reading cardiac computed tomography (MJB). All MDCTA images were evaluated on 3D image analysis workstation (GE Advantage Workstation, GE Healthcare, Milwaukee, WI). The post-processing image reconstruction algorithms used for interpretation included two-dimensional (2D) axial, or three-dimensional (3D) maximal intensity projection (MIP), multiplanar reformat (MPR), cross-sectional analysis and volume rendered technique (VRT).

**DISCUSSION**

**Coronary Arteriovenous Malformations:**

Coronary Arteriovenous Malformation (AVM) consists of abnormal communication between coronary artery and one of the cardiac chambers or vessels adjacent to the heart (1,2,3,4). We present our experience with diagnosing coronary Arteriovenous Malformation in our cardiac computed tomography laboratory.

Coronary Arteriovenous Malformation is a very rare anomaly present in 1 in 50,000 live births or 0.002% of the general population and is visualized in nearly 0.25% of patients undergoing catheterization (3,5,6,7,9,10,11,12,48). Coronary AVM may be congenital or acquired. It constitutes nearly half of all coronary artery anomalies and is the most common cause of hemodynamically significant coronary lesions (8,9,10,11,12,13,14). In 20-45% of the cases, Coronary AVMs are associated with some other congenital heart disease including atrial septal defect, tetralogy of Fallot, patent ductus arteriosus, ventricular septal defect or pulmonary atresia (8,10,11,13). It is present as an isolated finding in 55-80% of the cases (10,11,13,15). It was firstly described by Krause in 1865 (16,17). Most of the patients with coronary AVMs are older than 20 years. Approximately half of all patients with AVM remain asymptomatic and some coronary AVMs might disappear spontaneously during childhood (11,12,14,18,19). Symptoms and complications may develop with increasing age, and when surgery is performed in later life mortality and morbidity is increased (13,14).

Coronary AVM can arise from any of the coronary arteries. In the literature, the most common site reported is right coronary artery or its branches (55%), left anterior descending artery (35%) and circumflex coronary artery been rarely involved (8,11,12,13). Single origin is the most common form of coronary AVM, ranging from 74% to 90% (8,11,13,15) while multiple malformations have been reported in 10.7% to 16% of the cases. Over 90% of the fistulas drain into the venous structures of circulation which includes right ventricle (40%), right atrium (26%), pulmonary artery (17%), coronary sinus (7%), and superior vena cava (1%). Less frequently i.e. about 3-5 % of the AVMs drain into the left sided cardiac chambers (11,12,13). With increasing age, drainage into the main pulmonary artery is a relatively common occurrence (10).

Coronary AVMs cause shunting of blood and clinical presentation depends on the size of the malformation and thus degree of shunting (2). Coronary artery dilatation is reported but has not seen to be associated with the shunt size. In case of a low resistance malformation, the fistulous tract shunts significant amount of blood and causes a reversal of arterial flow in the segment distal to the AVM, resulting in a parasitic circulation, which causes decreased arterial pressures in the distal capillary beds and can cause tissue ischemia (2,7,20). About one half of the patients with AVM are asymptomatic (5,21). However, these malformations may result in severe complications and the patients may present with pulmonary hypertension because of an existing left to right shunt, congestive heart failure, subacute bacterial endocarditis, myocardial ischemia resulting from steal phenomena, rupture or thrombosis of the CAVM or associating arterial aneurysm (5,6,10,12,13,14,21,22). Symptoms and risk of these lethal complications increase with age (13,14,22).

The diagnosis of coronary AVM is challenging due to its low prevalence, yet it should be considered in many symptomatic or asymptomatic patients presenting with cardiac murmurs. Differential diagnosis includes patent ductus arteriosus, pulmonary arteriovenous fistula, ruptured sinus of Valsalva aneurysm, aortopulmonary window, prolapse of the right aortic cusp with a supracristal ventricular septal defect, internal mammary artery to pulmonary artery fistula, and systemic arteriovenous fistula (12,13,23). Invasive angiography remains the gold standard for diagnosis of coronary AVMs. However, the relation of coronary fistulas to other cardiac structures is at times unclear, and their origin and course can be limited due to overlap of adjacent structures (12,13,14,15,16,17,18,19,24,25,26,27,28,29,30,31,32). Most of the fistulas are small and found incidentally during coronary angiography. Non-invasive diagnostic modalities including transthoracic echocardiography combined with doppler and color flow imaging, transoesophageal echocardiography, magnetic resonance imaging and contrast enhanced multislice tomography can be used as an adjunct to coronary angiography (12,13,33,34,35).

**Cardiac Computed tomography and coronary Arteriovenous Malformations:**

The diagnostic performance of MDCT angiography has significantly improved with the latest 64-detector row scanners, with resultant high spatial and temporal resolution and high correlation with invasive coronary angiography (20,36). Cardiac computed tomography requires use of contrast and radiation exposure (2,4,5,13,37,38), however, currently CCT imaging protocols have been constructed to minimize the radiation exposure to the patient and has been shown to compare favorably with that of invasive coronary angiography (20,38). The images constructed are superior for anatomy but cannot assess the functional significance of the coronary AVM (e.g shunt volume) and CT angiography requires use of contrast material and the radiation exposure. Also, of note, the use of e.g. beta-blocker may be contraindicated in patient with tachycardia secondary to a large shunt volume in patients with coronary arteriovenous malformations.
Treatment of coronary Arteriovenous Malformations:

Although the natural history of the coronary AVM is variable and spontaneous closure is reported in some cases, it is recommended by most of the authors to treat symptomatic coronary Arteriovenous Malformations (7,11,18,19,37,38,39). The treatment of asymptomatic coronary AVM is still controversial with some authors recommending closure of coronary AVM in these patients to prevent fistula related complications that has been seen to increase with age (7,10,22,44). Currently there has been no consensus in surgical vs. medical treatment of patients with coronary AVMs. Surgical closure by epicardial and endocardial ligations are gold standard for treatment and remains safe and effective with good reported success (22,41,42). Successful surgical occlusion of these malformations without cardiopulmonary bypass has been reported (2,43). Ligation of the coronary AVM may be performed on the outside of the heart without CPB when there is a simple and easily accessible coronary AVM. The use of percutaneous Percutaneous closure technique needs several conditions: anatomy of the fistula should be favorable for this treatment (eg. Non-tortuous vessel, the fistula should be unique with distal narrowing to avoid embolism to the drainage site, and distal portion of the fistula should be accessible with the closure device (40,41,46). Transcatheter closure devices has been successful (6,41,44).

CONCLUSION

In conclusion, coronary AVM is a rare congenital disease and about half of the patients with the condition are asymptomatic. For patients who present with symptoms such as angina, dyspnea and arrhythmias, a coronary AVM should be suspected. Diagnosis of the condition can be easily and non-invasively made by cardiac CT angiography. Transthoracic echocardiography and magnetic resonance imaging are other diagnostic modalities which can be used for AVM evaluation and to calculate the shunt fraction. Shunt size, location, course and insertion site can be easily depicted, allowing for surgical or percutaneous procedural planning. Closure of the coronary AVM is recommended for symptomatic patients which can be done percutaneously if suitable anatomy and especially if patient needs percutaneous coronary intervention for a co-existing coronary pathology. Surgical closure can now be performed even without cardiopulmonary bypass in a beating heart.

TEACHING POINT

Coronary Arteriovenous Malformation is a rare congenital disease which might be asymptomatic or present as symptoms of angina, dyspnea or cardiac arrhythmias. With the increased spatial and temporal resolution of cardiovascular computed tomography it is possible to identify the coronary Arteriovenous Malformation along with looking at the coronary artery stenosis and proper referral can be made for medical vs. surgical management.

REFERENCES


Cardiac Imaging: Coronary arteriovenous malformation, as imaged with cardiac computed tomography angiography: A case series

Khachatryan et al.

FIGURES

Figure 1: A 69 year old white male with history of hypertension, diabetes mellitus and coronary artery disease. A 64 detector-row cardiac computed tomography (CCT) high resolution volume study images shows the malformations, coil and stent. Green and blue arrows indicate the malformation into pulmonary artery from conus branch on volume rendered and axial maximum intensity projection (MIP) images, respectively. Yellow and red arrows indicate coil and stent in LAD, respectively.

Figure 2: A 69 year old white male with history of hypertension, diabetes mellitus and coronary artery disease. CCT angiogram images showing malformations (axial MIP images). The green and yellow arrows indicate malformations into pulmonary artery from the conus branch and LAD, respectively. The red arrow indicates malformation into pulmonary artery from LM.
Cardiac Imaging: Coronary arteriovenous malformation, as imaged with cardiac computed tomography angiography: A case series

Figure 3: A 34 year old Caucasian female presented with chest pain and abnormal echocardiogram. 3D CCT angiogram was obtained by a 64 detector-row CCT high resolution volume study. The blue arrow shows the malformation arising from RCA into the pulmonary artery.

Figure 4 (bottom): A 56 year old Hispanic female with history of hypertension presents with a known coronary AV fistula for evaluation with CCT angiogram before closure. The 64 detector-row CCT angiography high resolution volume study (left image) shows malformation arising from LAD (green arrow). The axial MIP image on the right shows contrast in the pulmonary artery (blue arrow).
Cardiac Imaging: Coronary arteriovenous malformation, as imaged with cardiac computed tomography angiography: A case series

Khachatryan et al.

Figure 5: Transesophageal echocardiogram. a) Mid esophageal view at the level of short axis of the aortic valve, showing the aneurysmal inter-atrial septum. Saline contrast study (bubble study) is showing right to left shunt with bubbles entering through foramen ovale from right atrium to the left atrium (shown by an arrow). B) Mid esophageal view at the level of short axis of the aortic valve, showing a larger than normal diameter coronary vessel arising from the left coronary cusp of the aortic valve (shown by an arrow).

Figure 6: A 70 year old Hispanic female with history of hypertension, hyperlipidemia and diabetes mellitus was referred for possible LCX aneurysm. CCT axial MIP images showing the malformation arising from RCA on Electron Beam Computed Tomography. The image on the left shows RCA (black arrow). The image in the middle shows coronary sinus and the malformation into the coronary sinus from the RCA (red and yellow arrows, respectively). The right image also shows coronary sinus and the malformation into the coronary sinus from the RCA (green and blue arrows, respectively).

ABBREVIATIONS

- AVM: Arteriovenous Malformation
- CAVM: Coronary Arteriovenous Malformation
- CCT: Cardiovascular Computed Tomography
- CT: Cardiac Computed tomography
- EBCT: Electron beam cardiac computed tomography
- LAD: Left anterior descending artery
- LCX: Left circumflex coronary artery
- LMCA: Left main coronary artery
- LV: Left ventricle
- MDCT: Multi-row detector cardiac computed tomography
- RCA: Right coronary artery
- TTE: Transthoracic echocardiogram

KEYWORDS

Coronary Arteriovenous Malformations, Cardiovascular Computed Tomography, Diagnosis, Treatment

Online access

This publication is online available at: www.radiologycases.com/index.php/radiologycases/article/view/313

Interactivity

This publication is available as an interactive article with scroll, window/level, magnify and more features. Available online at www.RadiologyCases.com

Published by EduRad

www.EduRad.org