Symptomatic Coronary Cameral Fistula

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Abstract

Coronary cameral fistula is a rare entity and is characterized by an abnormal communication between coronary artery and a cardiac chamber. It is usually congenital and asymptomatic in majority of patients. If symptomatic the patients usually present in childhood. We present a case of 45-year-old male who presented with anginal chest pain and dyspnea on exertion for last 1 year. His exercise treadmill test was positive for ischemic changes and ECG-gated contrast enhanced CT was done for further evaluation. CT showed a large right coronary artery to right atrium fistula. It also ruled out any coronary atherosclerosis as reason for chest pain and ischemic symptoms on exercise treadmill test. The fistula was successfully closed by surgery and there was resolution of chest pain and dyspnea.

Keywords: Chest pain, computed tomography, congenital, coronary fistula

INTRODUCTION

Coronary cameral fistulas (CCFs) are rare and are characterized by abnormal communication between coronary artery (CA) and cardiac chamber that usually results from aberrancy of normal embryological development.1 Majority of the CCFs are asymptomatic, and it is exceptionally rare to have a single fistulous communication presenting with chest pain in middle age. We here present a rare case of symptomatic CCF diagnosed by electrocardiogram (ECG)-gated multidetector computed tomography (MDCT) with resolution of the symptoms following successful repair.

CASE REPORT

A 45-year-old non-smoker, male, with no significant past medical history, presented to the outpatient clinic of our hospital with progressive dyspnea on exertion and anginal chest pain for one year. He reported having an echocardiogram in an outside hospital that showed a tortuous vascular channel along the right atrium concerning for CA aneurysm. The cardiac and respiratory exam was unremarkable. The cardiac apex was not displaced, and there was no murmur. ECG showed normal sinus rhythm with no ST/T wave changes. His exercise treadmill test was positive for ischemic changes with accompanying chest pain.

Electrocardiogram-gated contrast-enhanced (CE) MDCT was done for further evaluation of the anomaly. Prior to image acquisition, the patient received oral metoprolol for heart rate control and also received 0.8

https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4485206/?report=printable
mg of nitroglycerin immediately before scanning for coronary vasodilatation. CECT showed an enlarged and tortuous right CA [arrows in Figure 1a-d] in the right atrio-ventricular groove that drained into the right atrium [star in Figure 1b] consistent with a CCF. CT also showed the absence of any coronary atherosclerosis. Three-dimensional volume rendered (VR) images accurately depicts the relationship of the enlarged right CA to the aorta, right-sided chambers, and the CAs. Since the patient was symptomatic, surgical closure of the fistula was done. The postsurgical course was uneventful with resolution of chest pain and dyspnea.

**DISCUSSION**

Coronary cameral fistulas are usually a congenital communication between CA and cardiac chamber and are most commonly seen with right CA (approximately 55%), but it can be seen with left or both CAs.[2] In unselected patients undergoing diagnostic coronary catheterization, they are incidentally detected in approximately 0.1% cases.[1] Based on the type of communication with the cardiac chamber, they classified as arterio-luminal (direct communication with the cardiac chamber) or arterio-sinusoidal (communication via sinusoidal network rather than direct communication). They drain into the right-sided chamber or great vessel in approximately 90% cases.[2]

These fistulae are asymptomatic in the majority of cases and are detected incidentally. Symptomatic CCF is rare, symptoms are more common in patients with multiple CCF.[3] Although uncommon, even single CCF may be symptomatic. The presence or absence of symptoms may be related to the size of the fistula and the site of origin and termination of the fistula. These fistulae may cause angina by coronary steal phenomenon and diastolic overload. Classically, diagnostic coronary angiogram has been used for diagnosis and has been considered as a gold standard for diagnosis[4] but with advances in noninvasive cardiac imaging, these fistulae are being increasingly diagnosed by MDCT or echocardiogram.

Although there is no consensus on treatment of choice of symptomatic fistulae due to its rarity; surgical repair, catheter closure and medical management have been successfully tried. Arterio-luminal subtype, as in our case can be successfully closed by surgery, whereas arteri-sinusoidal type is less amenable to surgery and use of beta-blocker is described in the literature.[7]

**CONCLUSION**

To conclude, CCFs are rare and symptomatic CCFs are even rarer. We hereby present utility of ECG-gated MDCT for demonstration of origin, draining chamber and size of CCF. Given high negative predictive value (nearly 100%) of coronary CT in ruling out coronary atherosclerosis,[8] it also helps in ruling out coronary atherosclerosis as the cause of patient's symptoms. Hence, we propose that MDCT can be done as sole preoperative imaging in low-risk CA disease (CAD) patients with CCF. ECG-gated MDCT circumvents the need of invasive diagnostic coronary catheterization in low-risk CAD subset patients not only for diagnosing/preoperative planning, but also by ruling out CAD.

**Footnotes**

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**REFERENCES**


Figures and Tables

Figure 1
(a-d) Curved multiplanar reformatted contrast-enhanced computed tomography images (a and b) show a large and tortuous of right coronary artery (CA) fistula (arrows) draining into the right atrium in keeping with CA fistula. The left CA (block arrow in b) is arising normally from the left coronary cusp and is a normal caliber artery. Three-dimensional volume rendered (VR) computed tomography images (c and d) give an accurate depiction of the course of fistula and its relation to adjacent vascular structures.