Coronary Artery Aneurysm with Associated Fistula:
A Rare Condition Detected by Computed Tomography
Coronary Angiography

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ABSTRACT
Coronary artery aneurysm with associated fistula is a rare condition. The complete anatomical delineation of this condition can be achieved with multi-detector computed tomography, which has advantages over echocardiogram and catheter angiography because of its ability to show the fistula separate from the surrounding cardiovascular structures, along with any aneurysm or obstruction along its course. Thus, multi-detector computed tomography may be preferred as the initial non-invasive imaging technique for characterisation of these rare congenital anomalies, and may be considered an alternative to conventional coronary angiography for follow-up studies because of its relative non-invasive nature.

Key Words: Coronary artery aneurysm; Coronary artery fistula; Coronary artery aneurysm with associated fistula

INTRODUCTION
A coronary artery fistula (CAF) is an abnormal communication between a coronary artery, bypassing the myocardial capillary bed, and either a cardiac chamber (coronary–cameral fistula) or a segment of the systemic / pulmonary circulation (coronary arteriovenous fistula). Rare cases such as CAF draining into the pericardium causing haematoma have also been reported.¹

CAF is an uncommon disorder with an incidence of 0.1% to 0.2% in patients undergoing coronary angiography.²³
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Most fistulae arise from the right coronary artery (60%) and terminate in the right side of the heart (90%). The termination sites of CAFs in decreasing order of frequency are the right ventricle, right atrium, coronary sinus, and pulmonary vasculature. Most CAFs are reported as congenital and have only rarely been reported as acquired. Acquired causes could be sequelae of previous open-heart surgery, post-endomyocardial biopsy and post-percutaneous transluminal coronary angioplasty, myocardial infarction, or penetrating/blunt trauma.

Coronary artery aneurysm with associated fistula (CAAAF) is even sparser, with only 50 cases reported in the English-language literature. Approximately 3% of CAFs are associated with coronary artery aneurysm. CAAAF occurs more frequently in women than in men (male-to-female ratio, 1:2.2). One-third of patients with CAAAF are asymptomatic, and two-thirds of patients may experience symptoms such as angina, dyspnoea on exertion, or palpitations. CAAAF may present with cardiac murmur or abnormalities on chest radiography.

This report is of an elderly man with CAAAF, which was likely to be a congenital abnormality because he did not have any previous cardiac interventions or procedures.

Figure 1. Computed tomography coronary angiograms with multiplanar reconstruction: (a, c, and d) axial oblique and (b) sagittal oblique images show a large aneurysm (A) arising near the ostium of the right coronary artery (arrowheads). There is also a coronary artery fistula (arrows) that subsequently drains via the dilated coronary sinus (CS) into the dilated right atrium (RA).
serious chest injury. The report describes the imaging techniques used to diagnose this patient and the role of multi-detector computed tomography (MDCT).

CASE REPORT
In May 2007, a 73-year-old man presented with a long history of atypical chest pain. He had previously undergone transthoracic echocardiogram in 1999 to 2000, which showed mild tricuspid regurgitation only. The results of other investigations done at the time of initial presentation — including physical examination, electrocardiogram, exercise treadmill test, thallium myocardial perfusion scan, and upper gastrointestinal endoscopy — were unremarkable.

The patient was subsequently referred to a cardiologist in 2008 because of worsening chest pain. At this referral, physical examination revealed pansystolic murmur all over the precordium, with palpable thrill. Electrocardiogram showed sinus rhythm with left axis deviation and partial right bundle branch block. Echocardiogram demonstrated dilated right and left atria, with aneurysmal dilatation of the right coronary artery and a suspected fistula to the right atrium. The left ventricular function appeared to be satisfactory, with no regional wall motion abnormality.

Subsequent computed tomography coronary angiography (CTCA) was performed on a 64-slice MDCT scanner (Aquilion 64; Toshiba Medical Systems Corp., Tochigi-ken, Japan) in Pok Oi Hospital, Hong Kong. Reconstructed images were then processed (Vitrea Enterprise Suite version 4.0; Vital Images Inc., Minnetonka, Minnesota, USA) using standard algorithms available in the CT cardiac package.

CTCA demonstrated a dilated and tortuous right coronary artery with a giant 5-cm aneurysm arising just beyond the ostium. Interval non-obstructive calcifications were also noted along the right coronary artery, which eventually drained via the coronary sinus into the right atrium (Figures 1 and 2). The origin, course, and calibre of the left coronary artery and its branches were normal. Three-dimensional volume-rendered (VR), multiplanar reconstruction (MPR), and curved MPR (cMPR) images were obtained (Figures 3 to 5).

The patient subsequently underwent left and right coronary artery catheterization, which confirmed the presence of a 3-cm proximal right coronary artery aneurysm that drained into the coronary sinus (Figure 6). Cardiothoracic surgery referral was arranged. However, the patient declined the option of definitive surgery despite the risk of rupture or thrombosis of the aneurysm explained to him. His condition remained stable for the subsequent 3 years, with only infrequent chest discomfort. In the 2011, he presented to Tuen Mun Hospital with chest discomfort and mild heart failure. He discharged himself against medical advice during the admission and was subsequently lost to follow-up.

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Figure 2. Volume-rendered images of the computed tomography coronary angiogram. (a) Coronal oblique and (b) axial oblique images show a right coronary artery aneurysm (A) and associated coronary artery fistula (arrows).
DISCUSSION

Conventional catheter coronary angiography is considered to be the gold standard for evaluation of coronary artery diseases and anomalies. However, owing to recent technological advancements in cardiac imaging, 64-slice MDCT permits whole-heart volumetric acquisition during a single (10-14 seconds) breath-hold with submillimeter isotropic resolution. In addition, the data set acquired can be reconstructed with different algorithms such as MPR, cMPR, and VR that provides the user with the best view for the evaluation of anatomical and pathological structures.
Traditional imaging techniques to diagnose a coronary artery fistula include echocardiography and catheter angiography. However, echocardiography has limitations, including operator dependency and limited acoustic window. The entire course of the fistula and its association with an aneurysm may not be readily visualised. Catheter angiography is an invasive procedure, and overlap between a tortuous fistula and adjacent cardiovascular structures may hamper complete evaluation of the lesion. Various studies have demonstrated the role of MDCT in the evaluation of an anomalous coronary artery, and case reports regarding its potential role in CAF are available.

MDCT is useful in the diagnosis of, and for delineating the complex anatomy of, a CAAAF. MDCT may be advantageous over catheter angiography because of its ability to show the fistula separately from the surrounding cardiovascular structures, along with any aneurysm or obstruction along its course. MDCT can provide accurate assessment of the size and location of a coronary artery aneurysm, and the amount of thrombus present inside the aneurysm. The high-quality two- and three-dimensional reformatted images in multiplanar and volumetric displays may be valuable in preoperative planning by showing spatial relations among the complex anatomy of the coronary artery fistula and aneurysm, great vessels, and heart.

Aneurysm repair, fistula closure, and coronary artery bypass grafts are definitive treatments for CAAAF. After diagnosis, the patient in this report was subsequently referred to the cardiothoracic surgeons for definitive treatment, but he declined the surgery.

CONCLUSION
A giant right CAAAF is a rare condition. The complete anatomical delineation of this patient’s condition was achieved with MDCT. MDCT has advantages over echocardiogram and catheter angiography because of its ability to show the fistula separate from the surrounding cardiovascular structures along with any aneurysm or obstruction along its course. Thus, MDCT may be preferred as the initial non-invasive imaging technique for characterisation of these rare congenital anomalies, and may be considered an alternative to conventional coronary angiography in follow-up because of its relatively non-invasive nature.

REFERENCES