Complex Coronary Cameral Fistulas Evaluated by Multi-Detector CT Angiography: A Report of Three Rare Cases and a Review of the Literature

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Congenital anomalies involving termination of a coronary artery and a fistulous communication with any cardiac chamber, pulmonary artery/vein, superior vena cava, or coronary sinus are defined as coronary cameral fistulas. Multi-detector computed tomography (MDCT) angiography with electrocardiographic gating precisely demonstrates complex and multiple coronary fistulas, their course and drainage routes, and relationships with surrounding structures, and MDCT is the primary modality for diagnosis, pre-treatment planning, and follow up of coronary cameral fistulas. We report three cases involving two elderly adults and a three-year-old child who were suspected to have coronary cameral fistulas and underwent MDCT to confirm the diagnosis and delineate the fistula, emphasising the unique role of MDCT in their diagnosis and management, along with a review of the literature.

Key words Coronary cameral fistulas · Coronary arterial fistulas · CT angiography · Anomalies, coronary vessel.

INTRODUCTION

Coronary cameral or coronary artery fistulas (CAFs) are anomalies involving termination of the coronary arteries and are, by definition, direct pre-capillary connections between a branch of the coronary artery with a cardiac chamber, pulmonary artery or vein, superior vena cava, or the coronary sinus [1,2]. Studies have shown that the incidence of coronary cameral fistulas is rare, about 0.0002% in the general population [3]. Multi-detector computed tomography (MDCT) angiography with electrocardiographic (ECG) gating precisely shows complex and sometimes multiple coronary fistulas, their intraluminal and extra-luminal morphology, drainage routes, and relationships with surrounding structures, helping guide treatment planning [1,2].

CASE REPORT

Case 1

A 58-year-old patient presented with dyspnoea on exertion for one year which had been aggravated over the previous three months. Echocardiography showed a dilated right coronary artery (RCA), and CAF was suspected. ECG-gated 64-slice MDCT coronary angiography showed a dilated and tortuous RCA arising from the right anterior coronary sinus (Fig. 1A and B) with a maximum diameter of about 9 mm. The RCA traversed the posterior interventricular groove with terminal posterolateral branches terminating into the left ventricle base adjacent to the upper posterior wall close to the bicuspid valve. A focal segment of constriction was noted in the intramural course, and the RCA had a small diverticular outpouching (4 mm) proximal to its junction with the left ventricle with a maximum calibre of 3 mm at the level of the diverticulum (Fig. 1C). In addition, multi-focal areas of atherosclerotic wall calcifications were noted in the RCA. The posterior descending artery, acute marginal branches of the RCA, and the left coronary artery (LCA) and its branches appeared normal. The diagnosis was RCA to left ventricular cameral fistula.

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Case 2

A 6-year-old girl presented with easy fatigability and chest pain. Echocardiography showed dilated coronary sinus and LCA, consistent with CAF. ECG-gated 64-slice MDCT showed, starting from its origin, a dilated left circumflex (LCX) artery which measured about 6 mm in calibre and directly communicated with the coronary sinus (Fig. 2). The coronary sinus was dilated, measuring 10 mm in maximum diameter, and enlarged up to the level of the confluence into the right atrium. The right atrium was mildly enlarged. The left anterior descending (LAD) artery and the remainder of its distal branches, the RCA and its branches, and the pulmonary artery appeared normal. Given the findings, a final diagnosis of LCX artery to coronary sinus coronary cameral fistula was made.

Case 3

A 65-year-old male patient presented with history of a recent anterior wall myocardial infarction (MI). Catheter coronary angiography showed 60% stenosis of the LAD artery and subtotal occlusion of the LCX artery. A fistula between the LCA

and the right ventricle was suspected. The patient underwent MDCT to evaluate the presence and course of the fistula, and it revealed calcified plaques in the LCA, LAD, and LCX arteries. The LAD was approximately 60% stenosed, and the LCX had subtotal occlusion. A small focal fistula was noted between the LAD and the right ventricular outflow tract just proximal to the pulmonary valve (Fig. 3).

DISCUSSION

Coronary cameral fistulas have a varied clinical presentation, with equal incidence in men and women, and they can be observed in any age group. Although most coronary cameral fistulas are congenital, they can be acquired from trauma, Takayasu's arteritis, post-radiotherapy, etc. Krause, who reported the first case of CAF in 1865, noted failure of normal regression of the intramyocardial trabecular sinusoidal system as the underlying embryological basis for the congenital form [3-5]. Clinical symptoms depend on the degree of left-to-right shunt, right or left heart overload, size, origin of the fistula, and drainage sites.

Fig. 1. Coronary CT angiogram images in axial (A), oblique sagittal (B), and oblique coronal (C) planes showing dilated tortuous RCA (arrows in A & B) with a fistulous opening into the left ventricle base (arrow in C) close to the bicuspid valve (asterisk in C) with terminal intramural narrowing and outpouching. Volume rendered technique (VRT) image (D) showing the dilated RCA. RCA: right coronary artery.

Fig. 2. Coronary CT angiogram in oblique sagittal (A) and oblique coronal (B) planes, and curved multi-planar reformation (C) showing a dilated left circumflex branch (white arrows in A & B) directly terminating into the coronary sinus (black arrow in C), which is seen draining into the dilated right atrium (asterisk in C).

Fig. 3. Coronary CT angiogram in oblique axial (A) and curved planar reformation (B) showing obstructive stenosis of the left anterior descending artery with mural calcific specks (black arrow) and a fistulous opening into the right ventricular outflow tract (white arrow). The right atrium is marked with an asterisk.

Coronary arterial flow diversion away from normal myocardium can cause the coronary steal phenomenon with features of myocardial ischemia, namely angina, dyspnoea, and arrhythmia. The complications of large left-to-right shunts include congestive cardiac failure, rupture or thrombosis, endocarditis, pulmonary hypertension, or rare embolic phenomenon in the late adult period. Associated congenital cardiovascular anomalies have been reported in 5% to 30% of cases [2].

For decades, the diagnosis of coronary artery anomalies has been made with conventional angiography. When a CAF with complex anatomy is suspected or coronary arterial intervention is expected, coronary angiography may not be sufficient. In addition to being invasive, coronary angiography has difficulty in delineating the precise course of the anomalous vessel, as the three-dimensional geometry of the course of the fistula is fluo-

roscopically displayed in two dimensions [6]. Furthermore, coronary fistulas draining into the low-pressure chambers of the right heart are not well visualized on conventional angiography because of insufficient manual contrast volume injection and significant contrast dilution [2]. ECG-gated MDCT is more useful than a conventional coronary angiogram for evaluating complex CAFs; ECG-gated MDCT more precisely delineates ostial origin, the proximal paths of the anomalous coronary arteries, and the drainage sites of CAFs by displaying the complex vascular relationships in three-dimensional multi-planar reformations [7,8].

CAFs arise from the RCA in about 50%, the LCA in 42%, and from both coronary arteries in approximately 5% of patients. The drainage is predominantly (>90%) into the systemic venous circulation, i.e., right ventricle (41%), right atrium (26%),

pulmonary artery (17%), coronary sinus (7%), superior vena cava (1%), and less often into the heart, i.e., left atrium (5%) and left ventricle (3%) [1,2].

Coronary artery fistulas are divided into two groups

Systemic-to-pulmonary artery fistulas and systemic-to-systemic type of fistulas.

Coronary-to-pulmonary artery fistulas

Arise from the RCA (about 60%), LCA (40%), and from both the RCA and LCA (5% of affected patients) [9]. If the coronary-to-pulmonary artery fistulas shunts are small, patients may be asymptomatic, but patients with large shunts can be symptomatic from early childhood. Surgical correction is indicated for a large fistula characterized by high flow, multiple communications, tortuous course, multiple terminations, or significant aneurysm formation [5].

Systemic-to-systemic type

Coronary-to-ventricle fistulas

Coronary-to-left ventricle fistulas [10,11] are rare and account for about 2–3% of all CAFs [9]. Coronary artery-cardiac chamber fistulas are thought to be the result of an abnormality during embryonic development of the coronary circulation system. When the intramyocardial trabecular sinusoids, which normally narrow and persist only as Thebesian vessels in adults, are not obliterated, a fistulous communication persists between the coronary arteries and one of the cardiac chambers [10]. These sinusoid fistulas are the most common congenital coronary anomalies and result in significant hemodynamic change [11].

Coronary artery-to-coronary sinus fistula

There have been only a few reports of aneurysmal coronary arteries with a fistulous connection to the coronary sinus, as this is an extremely uncommon form, and the reported cases were mainly fistulas between the LCX and the coronary sinus [12–14].

Coronary-to-bronchial artery fistula

The incidence of coronary-to-bronchial artery fistula (CBF) is reported to be 0.61% on cardiac MDCT [15]. CBF can present as a secondary result of occlusive disease of the pulmonary arteries or chronic pulmonary inflammation. Patients with CBF can remain clinically silent or can present with recurrent haemoptysis or chest pain due to the steal phenomenon. CBF usually originates from the LCX via a left atrial branch, and this branch crosses the pericardium in the retrocardiac space through the pericardial reflections.

Various imaging modalities have been used in the evaluation of CAFs. Conventional angiography provides high temporal and

spatial resolution, and hemodynamic assessment can be performed. Limited angles of projection of two-dimensional fluoroscopic images prevent delineation of complex configuration, multiplicity of coronary fistula, and drainage routes [16,17]. Echocardiography can depict the origin and draining site of fistula. Although microbubbles as contrast can increase depiction of the CAF, echocardiography has limited efficacy in the visualization of complex anatomy, distal parts of the CAF, occluded vessels, and the presence and course of collateral vessels. Magnetic resonance angiography (MRA) has the advantages of high temporal resolution, no radiation, and possible utility in assessing the proximal coronary arteries. However, MRA has lower spatial resolution and contrast-to-noise ratio compared to CT.

MDCT findings

Origin

The fistula is usually seen arising from a proximal segment of the coronary artery. The more proximal is the site of origin, the more likely it is that the affected artery will be dilated and atherosclerotic with features of myocardial steal. Fistulas arising from distal segments are small, but they are difficult to treat with coil embolization due to their tortuosity. Previous reports have shown that MDCT is superior to conventional angiography in complex CAF with dual origins.

Complexity

Rarely, complex communications and aneurysms can develop in congenital fistulas. On the other hand, acquired fistulas can show turbulent flow, atherosclerotic changes, and changes from trauma or inflammation.

Drainage sites

Drainage into a low pressure site can result in increased tortuosity and calibre of the fistula, causing clinical symptoms, aneurysm formation, and risk of rupture [18,19]. Confirming the presence and drainage site of the fistula, the “Contrast shunt sign” can be observed from contrast media being flushed into a non-opacified drainage chamber.

The clinical differential diagnosis includes other intra- and extra-cardiac shunts, such as patent ductus arteriosus, aorto-pulmonary window, and pulmonary arterio-venous fistula. The imaging differential diagnosis includes anomalous origin of the LCA with dilatation/aneurysm, atherosclerosis-related ectasias, sequelae of vasculitis (Kawasaki, Polyarteritis nodosa, Takayasu), scleroderma, and hereditary haemorrhagic telangiectasia. Identification of drainage sites with MDCT helps to limit the differential diagnosis [20].

Spontaneous closure of the fistula due to thrombosis is reported in up to 1–2% of cases. Careful periodic evaluations and close

follow up for months or even years may be needed. Usually, antiplatelet therapy and prophylactic precautions against bacterial endocarditis are recommended.

Mode of treatment

The options for permanent treatment include surgical ligation of the fistula and transcatheter embolization of the fistulous connection, and both modalities have similar early effectiveness, morbidity, and mortality. Indications for embolization are proximal location of the fistulous vessel, single drain site, extra-anatomic termination of the fistula away from the normal coronary arteries, older patient age, and absence of concomitant cardiac disorders requiring surgical intervention. Indications for surgery include a large CAF characterized by high fistula flow, multiple communications, very tortuous pathways, multiple terminations, significant aneurysmal formation, and need for simultaneous distal bypass [21-23]. When choosing between surgery or coil embolization for treatment, physicians can use MDCT as a "roadmap" to delineate the size, location, number, and tortuosity of the fistula site apart from associated changes both in the coronary system and cardiac lesions.

Long-term follow-up is essential due to the possibilities of postoperative recanalization, persistent dilatation of the coronary artery and ostium, thrombus formation, calcification, arrhythmias, and MI [12,23]. CAFs, a subgroup of coronary arterial anomalies, are rare cardiac defects. ECG-gated MDCT is a non-invasive and useful modality for diagnosis of CAFs, their course, and pre- and post-treatment follow up.

In conclusion, MDCT coronary arteriography is a non-invasive and useful imaging modality for diagnosis of CAFs. MDCT can also depict the origins, complex and tortuous pathways, and precise drainage site locations of CAFs. Thus, MDCT is of paramount importance in guiding the selection of proper treatment (surgical ligation vs. transcatheter embolization) and in revealing potential recanalization during post-treatment follow up.

Conflicts of Interest

The authors declare that they have no conflict of interest.

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