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Case Report

Role of 256-slice MDCT in the evaluation of coronary artery fistula: A case series with review of literature

Lt Col Sunita Dashottar^{a,*}, Brig R.K. Singh (Retd)^b, Brig S.K. Malani^c,
Surg Cmde Arti Sarin^d, Lt Col H.S. Arora^e

^a Classified Specialist & Head of Dept (Radiodiagnosis), Command Hospital (Central Command), Lucknow, India

^b Associate Professor, Pacific Medical College, Udaipur, Rajasthan, India

^c Professor (Cardiology), Military Hospital (Cardiothoracic Centre), Pune 411040, India

^d Professor & Consultant (Radiodiagnosis), Command Hospital (Southern Command), Pune 411040, India

^e Assistant Professor (Cardiology), Military Hospital (Cardiothoracic Centre), Pune 411040, India

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CAF, two being coronary cameral fistula (CCF) in neonates and the other two cases of CAF in adults diagnosed on 256-slice CT scanner.

Case-1

A male neonate presented with features of cardiac failure at birth. On examination, besides tachycardia and tachypnoea, a loud continuous murmur was heard on auscultation. The patient was managed with injection digoxin (10 µg/kg/day). Echocardiography was done, which showed grossly dilated right coronary artery (RCA), which was found to be communicating with the right atrium (RA) and possibly with the right ventricle (RV) as well. Cardiac CT was done in 256-slice CT scanner, which showed grossly dilated RCA arising from right coronary sinus with normal course in the atrio-ventricular groove on the right side. The RCA was seen communicating first with the right atrium (RA) and then with the right ventricle (RV) (Fig. 1). The widest calibre of the RCA was 8.4 mm in the proximal portion and the size of fistulous opening at the RA was 5.4 mm and at the RV was 7.4 mm. The findings were consistent with CCF.

This neonate was initially managed with injection Digoxin for congestive cardiac failure. Due to the large size of the fistulous communication, surgical closure was advised. The patient underwent surgical closure of the CCF at a different centre and on 3 months follow-up was found doing well without any medication.

Introduction

Coronary artery fistula (CAF) is a rare congenital cardiac anomaly with an abnormal communication between the coronary artery and the cardiac chamber or great vessels.

CAF may be detected on routine echocardiography, or computerized tomography coronary angiography (CTCA) or even catheter coronary angiography (CCA) incidentally or in symptomatic patients. With the advent of ultrafast MDCT (multi-detector CT) scanners, it is now possible to confirm the diagnosis of CAF non-invasively, which otherwise would have required catheter angiography. Ultrafast MDCT Coronary Angiography is now considered as the preferred modality for diagnosing CAF. We present a rare series of four cases of

* Corresponding author. Tel.: +91 8806701108.

E-mail address: sunitadashottar70@gmail.com (S. Dashottar).

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Fig. 1 – (Case no-1) – Cardiac CT maximum intensity projection (MIP) axial images depict: (a) grossly dilated RCA arising from the right anterior coronary sinus and coursing in the atrio-ventricular (AV) groove anteriorly on right side (white block arrow). LCA is seen arising from the left coronary sinus (black simple arrow). (b) Fistulous communication between the RCA (white block arrow) and Right Atrium (RA) (simple black arrow) is demonstrated. (c) Mid portion of the RCA in the AV groove with reduced calibre distal to the fistulous opening (white arrow) is demonstrated. (d) Distal portion of RCA is dilated and is seen communicating (white block arrow) with the RV.

Case-2

A six-month-old male infant patient presented with repeated attacks of respiratory tract infection since birth in the year 2012. Echocardiography showed dilated left coronary artery (LCA) and left anterior descending artery (LAD) with a dilated sac-like structure seen at the distal end of LAD suggestive of distal LAD aneurysm. A small communication was suspected between the aneurysmal sac and the right ventricle suggestive of CCF. For further evaluation, CCA was done. CCA showed dilated LCA and LAD with aneurysm at the distal end of LAD. Faint contrast opacification was seen in the RV and the pulmonary trunk suggestive of fistulous communication between the aneurysmal sac and the RV (Fig. 2). The patient was managed conservatively and was observed clinically for 02 years.

Echocardiography was repeated at the age of two and half years in Dec 2014, which showed partial thrombus

formation in the aneurysmal sac of distal LAD. However, at this time, the fistulous communication between the distal LAD aneurysm and the RV could not be established.

CTCA was done for further evaluation and confirmation, which showed grossly dilated LCA and LAD with faintly opacified 34 mm × 31 mm × 28 mm size aneurysm at the distal end of LAD. However, no obvious CCF was demonstrated (Fig. 3). Contrast opacification of the LAD and the aneurysmal sac was less as compared to the LCA possibly due to partial thrombus formation and slow flow of contrast in the LAD and within the aneurysmal sac. These findings were suggestive of spontaneous closure of CCF with partial thrombus formation in the distal LAD aneurysm. CCA further confirmed the findings of cardiac CT.

Percutaneous transcatheter coiling/vascular occlusive device was advised for the patient so as to completely thrombose the aneurysmal sac and prevent any future complications.

Fig. 2 – (Case no-2) – Catheter coronary angiography (24/11/2012) when the baby was 06 months old. (a) Aortic root injection shows grossly dilated LCA and LAD (simple black arrow) with contrast opacification of the distal LAD aneurysm (white block arrow). RCA (small black arrow) is normal in calibre and course. (b) Selective catheter angiography of LCA shows dilated LCA and LAD with distal LAD aneurysm. Contrast opacification of the right ventricle (simple black arrow) and pulmonary trunk (block white arrow) is also seen suggestive of coronary cameral fistula.

Case-3

A 47-year old female patient presented with history of dyspnoea on exertion and recurrent episodes of chest pain for the past six months. Echocardiography demonstrated a 33 mm wide atrial septal defect (ASD) with left to right shunt and moderate tricuspid regurgitation (TR). Systemic to pulmonary blood flow ratio (Q_p/Q_s) was 3.2.

After putting the ASD closure device, routine CCA was done. Selective coronary angiography of LCA showed multiple small calibre tortuous arteries arising from LCA, proximal portion of the LAD and left circumflex artery (LCX) with contrast opacification of the main pulmonary trunk (MPA) (Fig. 4). Few contrast-filled round lesions were also seen in relation to the tortuous collaterals suggestive of small aneurysms of the collateral arteries. These findings were suggestive of CAF. After the ASD closure, the systemic to pulmonary blood flow ratio (Q_p/Q_s) was 1.34.

CTCA was performed to confirm and further delineate the coronary artery anatomy. Multiple tortuous collateral arteries were seen arising from the LCA, proximal portion of LAD and LCX with contrast opacification of the MPA in the arterial phase. The LCA, proximal portion of LAD and LCX, were dilated with abrupt tapering distally. Few small aneurysms of the collateral arteries were also noted. Multiple small collaterals arising from coronary arteries and draining into the MPA were consistent with findings of CAF (Fig. 5).

The patient was managed with conservative treatment as the Q_p/Q_s ratio before the ASD closure was 3.2 and after ASD closure was less than 1.5. The patient remained asymptomatic on medical treatment on regular 03 monthly follow-up.

Case-4

A 65-year-old male patient, a known case of diabetes mellitus and hypertension, presented with recurrent episodes of chest pain and breathlessness on exertion for the past 01 month. CTCA showed a prominent conus branch of the RCA communicating with the MPA. A few prominent arterial channels were also seen arising from the proximal portion of LCA and LAD draining into the MPA (Fig. 6). Based on the CTCA findings, diagnosis of bilateral CAF arising from RCA as well as from LCA and LAD communicating with the main pulmonary trunk was made.

CCA was done for further evaluation, which showed multiple small collaterals arising from LCA and the proximal portion of the LAD with contrast opacification of the MPA. In addition to this, a bunch of multiple small arteries was also seen arising from the large tortuous conus branch of the RCA with contrast opacification of the MPA. These findings were consistent with CAF (Fig. 7).

This was a rare case of bilateral CAF arising from RCA as well as LCA and LAD, which was communicating with the MPA. The patient was managed with conservative treatment and remained asymptomatic on regular 03 monthly follow-up.

Discussion

CAF is a rare congenital cardiac anomaly with an abnormal communication between the coronary artery and the cardiac chamber (CCF) or great systemic or pulmonary arteries or vena cava (coronary arterio-venous fistula) bypassing the

Fig. 3 – (Case no-2) – Cardiac CT of the same patient as in Fig. 2 in Dec 2014. (a) CTCA axial MIP image at the level of aortic root shows grossly dilated LCA and LAD with faint opacification of LAD (block white arrow). (b) Axial MIP image at the level of cardiac apex shows faint opacification of the distal LAD aneurysm (block white arrow). No obvious communication is seen between the aneurysmal sac and the adjacent cardiac chambers. (c) Coronal reformatted MIP images showing the distal LAD aneurysm (block white arrow). (d) Coronal reformatted MIP images showing the dilated LAD (simple white arrow) with distal LAD aneurysm (block white arrow).

myocardial capillary bed.¹ CAF is usually congenital, but acquired form has also been reported.

Lim et al. detected CAF in 56 patients (0.9%) in a retrospective study on 6341 patients who underwent CT coronary angiography. It was reported by the investigators that the incidence of the CAF detected by the CTCA was found to be more than what is generally detected by invasive catheter angiography (0.05–0.25%).²

CAF may present as an isolated finding in 50–80% of the cases, while in 20–45% of the cases, it may be associated with other congenital cardiac anomalies like atrial/ventricular septal defect, tetralogy of Fallot, aortic or pulmonary atresia and patent ductus arteriosus.³ Approximately 50% of the patients with CAF remain asymptomatic and a few disappear spontaneously during childhood.⁴

Majority of the patients of CAF are asymptomatic and are usually detected incidentally on echocardiography or CCA. The presence of symptoms depends upon the size of the CAF,

which can deprive the myocardium of the necessary amount of blood leading to myocardial ischaemia. 57% of symptomatic patients present with anginal chest pain, which occurs primarily due to coronary steal phenomena.⁵

The incidence of CAF originating from LCA is 42.4%, from RCA is 12.1% and from both is 45.5%.⁵ The commonest draining site of CAF is pulmonary artery (76.8–84.8%),⁶ followed by cardiac chamber and bronchial artery (8.9%) and rarely in superior vena cava (1.8%).² Zhou et al. reported 24.2% of CAF cases to be associated with aneurysms of coronary artery.⁶

The imaging of the CAF includes non-invasive techniques, such as echocardiography, CTCA and cardiac magnetic resonance imaging (MRI). CT and MRI can demonstrate the origin, course, drainage site of CAF and coexisting abnormalities.⁶ MDCT is superior to catheter angiography in delineating the origin and proximal portion of the anomalous coronary artery.⁷

Fig. 4 – (Case no-3) – Selective LCA angiography: demonstrates multiple tortuous arterial channels arising from the LCA (black arrow), proximal portion of LAD and LCX with opacification of the MPA (white arrow). A few small aneurysms of the collaterals (small black arrow) are also seen.

The latest ultrafast MDCT scanners have significantly improved the diagnostic capabilities of CTCA due to its high gantry speed, and improved spatial and temporal resolution; it has been found to correlate well with conventional catheter angiography. CTCA can acquire volume acquisition of the entire heart and the coronary arterial tree within 4–5 s with higher temporal and spatial resolution than MRI.⁸ Protocols and techniques used in recent MDCT scanners have reduced

the amount of contrast and radiation exposure to the patient as compared to catheter angiography. The three-dimensional reconstructed images can give exquisite anatomical details of the CAF like shunt location, size, multiplicity, course, and insertion site for the surgical or percutaneous procedural planning. Volume rendered images provide an overview of the cardiac and vascular anatomy. As CTCA has become more popular, the incidentally detected CAF are increasing in

Fig. 5 – (Case no-3) – CTCA 3D volume rendered image: shows multiple tortuous arterial channels arising from the LCA, proximal portion of LAD and LCX (black arrow), draining into the MPA (white arrow). Proximal portion of LAD and LCX are dilated with abrupt tapering distal to the origin of the collaterals. A few small focal aneurysms of the collaterals are also seen (small black arrow).

Fig. 6 – (Case no-4) – Coronary CT angiography maximum intensity projection (MIP) image of the heart shows (a) few collateral arterial channels are seen arising from the LCA and LAD (block red arrow) and draining into the MPA. (b) Prominent conus branch (block yellow arrow) terminating into the pulmonary trunk.

numbers as compared to the CCA.^{9,10} CTCA has higher sensitivity (100%) in the visualization of the normal and abnormal coronary arteries.² The relative disadvantage of CTCA is radiation dose to the patient, which can be lowered by shielding the body parts and tube current modulation, especially with the newer generation of ultrafast CT scanners.

CTCA on ultrafast CT enables accurate delineation of anomalous communications, size and number of fistulae in CAF along with depiction of coexisting abnormalities. Therefore, it is considered as the preferred non-invasive imaging

modality for the diagnosis and further management of CAF.^{6,11} The radiation dose of prospective ECG-gated high-pitch spiral dual source CT angiography is below 1.0 mSv,¹¹ and with 320-row MDCT it is approximately 5.7 mSv.^{12,13} Leong et al. found that the median effective dose for CTCA on 128 slice dual source CT scanner was 2 mSv, while for diagnostic catheter angiography, it was 9.2 mSv ($p < 0.001$).¹⁴ They demonstrated that the radiation dose with the new-generation Ultrafast CT scanner has reduced from 5 mSv to 2 mSv, while there is no change in the radiation dose of CCA.

Fig. 7 – (Case no-4) – Selective catheter angiography. (a) Selective RCA angiography image showing dilated tortuous conus branch of RCA (white arrow) with multiple bunch of tortuous arterial channels (yellow arrow). (b) Selective LCA angiography image showing a few faintly opacified collaterals (yellow arrow) seen arising from the dilated proximal portion of the LAD and LCX with faint opacification of the MPA (white arrow).

Although coronary catheter angiography (CA) is considered to be the gold standard in the diagnosis of CAF, it remains a relatively invasive technique.¹¹ Other disadvantages of CCA are – as follows: technically difficult to cannulate all the vessels associated with CAF, and complex anomalous vessels and its relations with adjacent structures are not well delineated due to its 2D nature of Images.² CA cannot clearly demonstrate the CAF draining into the low-pressure chambers of the heart, due to the significant dilution of the contrast.¹⁵

Catheter angiography may not be as efficient and sufficient for the evaluation of complex CAF as it is not able to demonstrate multiple fistulae arising from different segments of coronary arteries. CTCA not only can demonstrate the complex anatomy non-invasively, but also it is easy to do the follow-up studies.¹⁰

Conclusions

Ultrafast CT scan can be considered as the non-invasive imaging modality of choice for the diagnosis of CAF. It provides excellent 3D images and can accurately delineate fine anatomical details, which are required for management of CAF.

Conflicts of interest

The authors have none to declare.

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