



MDCT Depiction of Single Coronary Artery and Associated Cardiac Anomalies: Experience from A Tertiary Care Hospital

KEYWORDS

Single coronary artery (SCA); coronary sinus; multidetector CT; ventricular septal defect

* Dr. Abhishek Chauhan

MD Assistant Professor Dr Ram Manohar Lohia Institute of Medical Sciences Lucknow * Corresponding Author

Dr. Ragini Singh

MD DMRE FICRI Professor Dr Ram Manohar Lohia Institute of Medical Sciences Lucknow

Dr. Gaurav Raj

MD Associate Professor Dr Ram Manohar Lohia Institute of Medical Sciences Lucknow

Dr. Tushant Kumar

MD Assistant Professor Dr Ram Manohar Lohia Institute of Medical Sciences Lucknow

ABSTRACT Congenital coronary anomalies occur rarely, varying from 0.3-1.3% of the population. Although, most such anomalies are of no clinical significance and are found incidentally at coronary angiography, their impact on premature cardiac morbidity and mortality in young adults is not trivial. Knowledge of 3D spatial relations of the anomalies and understanding of clinical significance is important for the cardiac surgeons for accurate diagnosis and proper patient treatment. Increased use of multidetector CT helps in recognizing such anomalies on cross-sectional, multiplanar and volume rendered reconstructions. We present three cases, all of whom had a single coronary artery with associated congenital cardiac anomalies.

Introduction

Single coronary artery is an anomalous situation, in which one coronary artery arises with a single ostium from the aortic trunk. This is an extremely rare congenital anomaly seen in only 0.0024%-0.44% of the general population (1, 2). This condition is usually compatible with a normal life expectancy, however it can be devastating if artery has a malignant course or there is a proximal stenosis with inability to develop collaterals.

MDCT with ECG-synchronisation and new reconstruction algorithms is now accepted an accurate diagnostic technique for defining ostial origin and path of anomalous coronary branches (3).

Cases

Case I

S.K., 21 year old gentleman was admitted in cardiology department of our institute with complaints of dyspnoea on exertion (DOE) – grade II from childhood and DOE – grade III for last two months. He also complained of bluish discoloration of lips and nails from early infancy, with positive history of squatting, cyanotic spells and recurrent lower respiratory tract infections. Clinical examination revealed presence of cyanosis and clubbing. Further, cardiac evaluation was done in our department. Whole-heart coronary MDCT angiography (PHILIPS 64 slice MDCT) revealed levocardia, cardiac situs solitus with atrio-ventricular concordance. Atrial situs solitus was noted with large septum primum atrial septal defect (ASD) measuring 39.0mm. Anterior ventricular chamber was enlarged and hypertrophied (free wall-21.6mm) with morphological features of right ventricle. Aorta was seen arising from morphological right ventricle. Posterior chamber with morphological features of left ventricle was hypoplastic with no outflow tract s/o pulmonary atresia. A large membranous (28.6 mm) and small muscular (3.4mm) ventricular septal defect (VSD) was also noted. Right and left pulmonary arteries were hypoplastic and were filling from multiple collaterals. Patent ductus arteriosus was also seen communicating arch of aorta to left pulmonary artery. A single coronary ostium (Figure 1) was

noted in anatomical left coronary sinus with a common trunk, which was subsequently dividing into two branches, with branch on left continuing in left atrioventricular groove as LCX and giving origin to three marginal branches and posterolateral branch (Figure 1). Right branch after giving origin to branch in anterior interventricular groove (LAD) was continuing anteriorly to aorta in right atrioventricular groove as RCA. Prominent branches to anterolateral free wall of right ventricle were noted from RCA, with termination in posterior descending artery. No e/o any obstructive coronary disease of all three vessels was seen.

Patient was planned for right ventricular and left ventricular outflow tract repair.



Figure 1. Lipton type L / A single coronary artery

Case II

N.P., 23 year old non-diabetic, non-hypertensive female was admitted with complaints of DOE II, easy fatigability and recurrent syncope for last eight months. Exaggerations of symptoms to grade III were complained of since last five months. History of one episode of symptomatic documented VT which was cardioverted with DC shock. There was no history of rest angina. Echocardiography and catheter angiography revealed dysplastic pulmonary valves with infundibular stenosis, for which she underwent right ventricular outflow tract (RVOT) repair. Five months post procedure; she was sent to our department for assessment of RVOT. MDCT cardiac angiography was done on PHILIPS 64 slice scanner. Reconstructed RVOT showed mild narrowing at outflow with normal sized pulmonary arteries. Sub-aortic VSD (5.4mm) along with small muscular VSDs were noted with enlarged and hypertrophied right ventricle. A

small atrial septal defect was noted measuring 4.8mm. Incidental finding of single coronary ostium was found with common trunk arising from the left sinus of Valsalva.

Single trunk after a short course was dividing in left and right branches (Figure 2).

Right branch was giving rise to a prominent branch just after its origin which was seen supplying antero-lateral free wall of right ventricle. The same artery was further extending in right atrio-ventricular groove as RCA and giving rise to AMA and terminally continuing as posterior descending artery. Left coronary trunk was giving origin to small ventricular branches and a prominent branch which was continuing in anterior interventricular groove as LAD with single diagonal branch. Left trunk after giving origin to LAD was continuing in left atrioventricular groove, giving OM1 and OM2 branches, and terminally as posterolateral branch (PLB) (Figure 2).

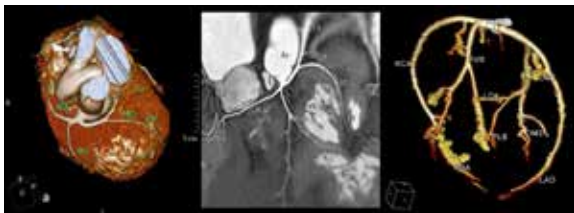


Figure 2. Lipton type L / A single coronary artery

Case III

R.Y., 6 year old male admitted to department of cardiology of our institute with complaints of fatigue for two years and bluish discoloration of lips and nails for one year. On examination positive findings of cyanosis and clubbing were seen. MDCT angiography done in our department showed left sided IVC draining into right atrium with situs ambiguus and right isomerism. All four pulmonary veins were draining into superior vena cava (SVC) suggesting total anomalous pulmonary venous connection (TAPVC) (Figure 9). Atrio-ventricular concordance was seen with normal anatomical location of ventricles. Double outlet right ventricle was seen with d-malposition of aorta. Septum primum ASD was seen measuring 19.1mm, with perimembranous VSD (17.1mm). Pulmonary infundibular narrowing with hypoplastic pulmonary arteries was also seen. Multiple aorto-pulmonary collaterals were noted. Incidental single coronary artery was noted from anatomical right posterior coronary sinus giving right branch in right atrio-ventricular groove as RCA and left circumflex trunk initially continuing between aorta and left atrium and distally giving origin to LAD (Figure 3). The above findings were further confirmed on catheter angiography.

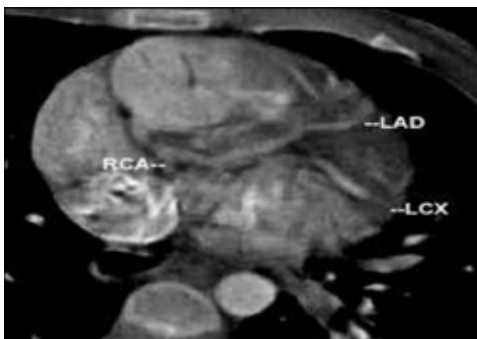


Figure 3. Lipton type R / P single coronary artery

Discussion

Single coronary artery is defined as the absence of a portion of one of the coronary arteries at the aortic root rather than the absence of a complete coronary artery. The more distal portion of the vessel is found in its expected location and communicates with the normally arising coronary artery by a connecting branch, creating a coronary pattern with a single source-thus the name single coronary artery (4). Single coronary artery is an extremely rare congenital anomaly with a quoted incidence of 0.0024% to 0.044% (2). It has been seen as an isolated occurrence and in association with congenital heart diseases.

Several classification systems for coronary artery abnormalities exist. Lipton et al (5) classified coronary variations based on origin and anatomical course relating to the ascending aorta and pulmonary trunk.

Ostial location

R à Right sinus of Valsalva

L à Left sinus of Valsalva

Anatomical distribution

I à Single coronary artery with normal right or left coursing (RC or LC)

II à After leaving the right or left sinus the single coronary artery crosses at the base of the heart as a large transverse trunk in order to supply the contralateral coronary artery

III à Single coronary artery arising from the right sinus, with the left anterior descending and circumflex arteries from separate coronary artery trunks instead of a single trunk immediately at the exit.

Course of the transfer branch

A à Anterior to the large vessels (anterior to the right ventricle)

B à Between the aorta and pulmonary artery

P à Posterior to the large vessels

S à Septal type (above the interventricular septum)

C à Combined type

In the first two cases, it was Lipton type L / A single coronary artery and in third case it was type R / P.

More recently, Angelini et al. (1) proposed a slightly different classification according to the anatomical course within the interventricular sulcus and atrioventricular groove, as well as the location of penetrating side branches.

A single, isolated origin of the coronary vessels may be associated with sudden cardiac death, particularly in cases with an interarterial course, although the risk has not been adequately quantified in specific studies. Its association with cardiac ischemia and congestive cardiac failure is likely due to the fact that a proximal stenosis of a single coronary artery may prove devastating if adequate collaterals do not exist [6]. The proposed management of this subgroup of patients is surgical because of the high incidence of sudden cardiac death, especially in the young who seldom have associated coronary artery disease or other abnormalities.

Coronary artery anomalies are also commonly associated with congenital heart diseases. Hence, principal purpose in these conditions is to forewarn the surgeon of important aberrant epi- or intramyocardial coronary arteries.

Until recently, delineation of the anatomical course relied on pulmonary artery catheter placement. However, the development of retrospectively ECG-gated MDCT and dual-tube cardiac CT technology as a suitable technique has allowed for noninvasive, accurate visualization of coronary artery anomalies, which may be difficult to delineate fluoroscopically at angiography in a two-dimensional plane.

To conclude, knowledge of single coronary artery anomalies is important. Most of these are benign, but a small number, principally those with a course between the great vessels are associated with myocardial ischemia and even sudden death. MDCT cardiac angiography highlights the value of recognizing such anomalies on cross-sectional, multiplanar and volume rendered reconstructions.

References

1. Angelini P, Velasco JA, Flamm S. Coronary anomalies: incidence, pathophysiology, and clinical relevance. *Circulation*. 2002;105:2449-2454
2. Desmet W, Vanhaecke J, Vrolix M, Van de Werf F, Piessens J, Willems J, de Geest H. Isolated single coronary artery: a review of 50,000 consecutive coronary angiographies. *Eur Heart J*. 1992;13:1637-1640
3. Shi H, Aschoff AJ, Brambs HJ, Hoffmann MH. Multislice CT imaging of anomalous coronary arteries. *Eur Radiol*. 2004;14:2172-2181
4. Smith JC. Review of single coronary artery with report of 2 cases. *Circulation*. 1950;1:1168-1175
5. Lipton MJ, Barry WH, Obrez I, Silverman JF, Wexler L. Isolated single coronary artery: Diagnosis, angiographic classification, and clinical significance. *Radiology*. 1979;130:39-47
6. Greenberg MA, Fish BG, Spindola-Franco H. Congenital anomalies of coronary artery: classification and significance. *Radiol Clin North Am*. 1989;27:1127-1146