International Journal of Research in Medical Sciences
Baroopal A et al. Int J Res Med Sci. 2019 May;7(5):1968-1971
www.msjonline.org
pISSN 2320-6071 | eISSN 2320-6012
DOI: http://dx.doi.org/10.18203/2320-6012.ijrms20191711

Case Report

Anomalous left coronary artery from pulmonary artery in an adult

Anil Baroopal*, Rohit Mathur, Sanjeev Sanghvi, Swati Mahajan

Department of Cardiology, Dr. S. N. Medical College, Jodhpur, Rajasthan, India

Received: 27 February 2019
Revised: 02 April 2019
Accepted: 08 April 2019

***Correspondence:**
Dr. Anil Baroopal,
E-mail: anisabarupal@yahoo.com

ABSTRACT

Anomalous left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital cardiac anomaly that can cause myocardial infarction, heart failure and even death in paediatric patients. Only few untreated patients survive till adulthood. Here we present the case of a 28-year-old lady with exertional dyspnoea and chest pain who was diagnosed to have ALCAPA.

Keywords: ALCAPA, Congenital cardiac anomaly, Coronary angiography

INTRODUCTION

Anomalous origin of left coronary artery from the pulmonary artery (ALCAPA) is an uncommon congenital cardiac anomaly affecting 1 in 300,000 live births and survival to adulthood is furthermore rare accounting for only 10-15% of these cases.[1,2] In this report, authors present a rare case of ALCAPA in an adult who was successfully operated.

CASE REPORT

A 28-year-old lady presented in outpatient department of our institute with complaints of exertional dyspnea and chest pain radiating to left upper limb for fifteen days. She had past history of similar complaints since early childhood which used to get relieved after taking rest. She had no conventional risk factors for atherosclerotic coronary artery disease (CAD) such as family history of CAD, smoking, diabetes mellitus, hyperlipidemia and hypertension. She has two elder siblings and none of them has any congenital heart disease. In obstetric history, her first pregnancy was uneventful eight years back. She was told to have some congenital heart disease five years back during early second pregnancy. She was not investigated further as she did not consult a physician even after second delivery which was uneventful.

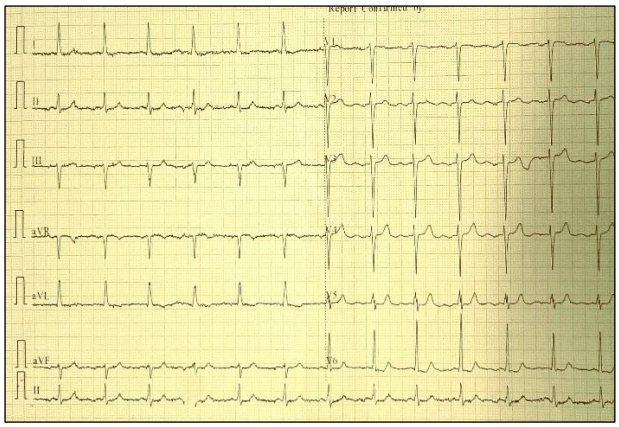

Figure 1(A): Electrocardiogram showing 0.5 mm down sloping ST segment depression in lead V6 and poor progression of R wave in leads V1 to V4.

On examination, her pulse was 80/min, regular and blood pressure was 100/70 mm of mercury. On auscultation a grade 3/6 continuous murmur was audible at left sternal border. There was no left ventricular (LV) S3 or flow murmur at apex. Laboratory investigations were within normal range along with negative troponin I. Her electrocardiogram showed 0.5 mm down sloping ST segment depression in lead V6 and poor progression of R wave in leads V1 to V4 (Figure 1(A)).

Treadmill test was not performed as the patient was symptomatic. Chest x-ray showed cardiomegaly with cardiothoracic (CT) ratio of 56%, LV type apex, normal pulmonary blood flow and lung parenchyma (Figure1(B)).

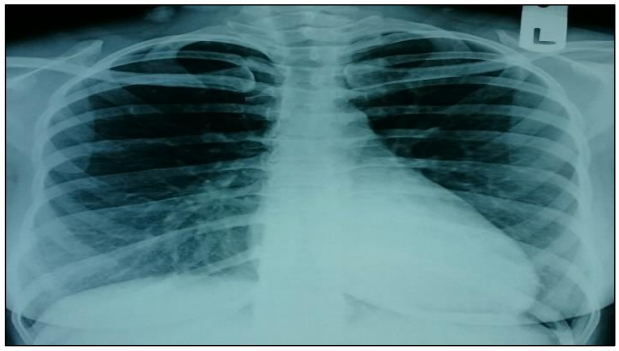

Figure 1(B): Chest x-ray showing cardiomegaly with CT ratio 56%, LV type apex, normal pulmonary blood flow and lung parenchyma.

Transthoracic echocardiogram was performed which revealed dilated left ventricle with hypokinesia in left anterior descending artery (LAD) and left circumflex artery (LCX) territory, LV systolic dysfunction with LV ejection fraction (EF) of 32% by Simpson method (Figure 1(C)), trivial mitral regurgitation and mild pulmonary regurgitation. Papillary muscle and mitral valve apparatus were normal.

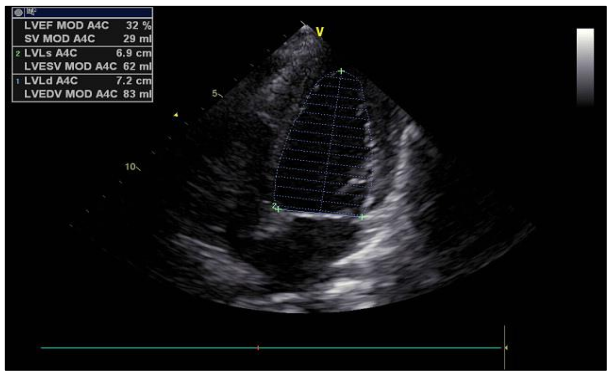

Figure 1(C): Two-dimensional transthoracic echocardiographic apical four-chamber view showing dilated left ventricle, left ventricular systolic dysfunction with LVEF 32% by Simpson method.

Origin of left main coronary artery (LMCA) was not seen from aorta. Dilated right coronary artery (RCA) arising from right sinus of Valsalva was seen. A continuous turbulent signal was observed in pulmonary artery which made us to suspect ALCAPA. Coronary angiography was performed which revealed a dominant, aneurysmally dilated and tortuous RCA arising from the right coronary sinus. LAD and LCX filled retrogradely via collaterals after RCA injection, with backflow from LMCA to main pulmonary artery (MPA) (Figure 1(D)).

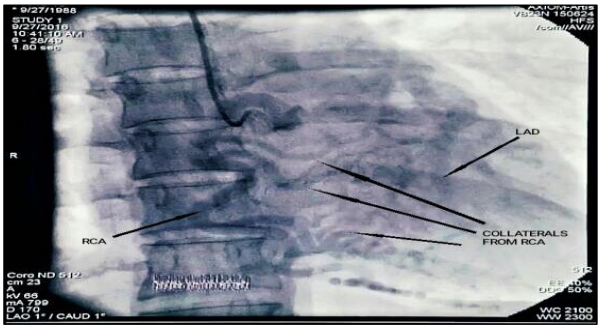

Figure 1(D): Coronary angiogram (LAO 10 /CAUD 10 view) showing aneurysmally dilated and tortuous RCA arising from the right coronary sinus with multiple inter coronary collaterals from RCA to LAD and LCX.

No evidence of pulmonary hypertension was detected clinically or by echocardiography, however right sided catheterization study was not done.

Multidetector computed tomography (MDCT) coronary angiogram with 3-D reconstruction and volume rendered images demonstrated LMCA arising from inferolateral aspect of the MPA with diffusely enlarged and tortuous RCA arising from the right coronary sinus with multiple inter coronary collateral formation from RCA to LAD and LCX (Figure 2(A) and (B)).

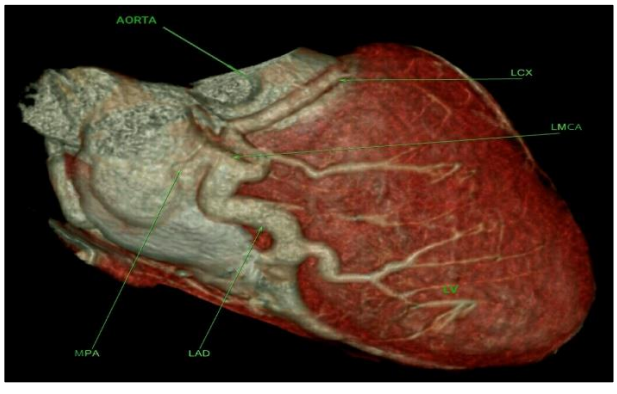

Figure 2(A): Multidetector computed tomography (MDCT) coronary angiogram with 3-D volume rendered image showing LMCA arising from inferolateral aspect of the MPA.

A diagnosis of ALCAPA was made and managed surgically by anastomosing left internal mammary artery to LAD, saphenous vein graft to obtuse marginal artery and LMCA was over sewn from MPA.

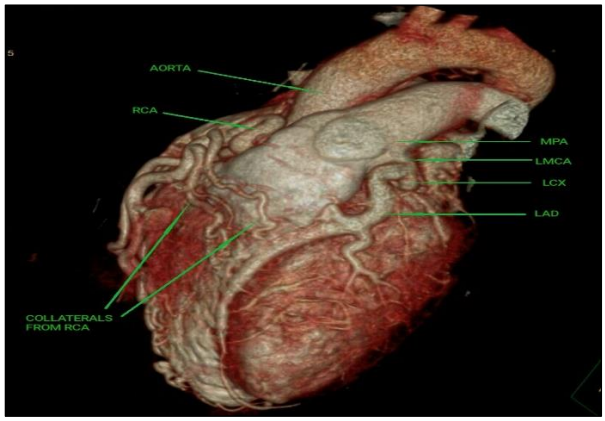

Figure 2(B): MDCT coronary angiogram with 3-D volume rendered image showing LMCA arising from inferolateral aspect of the MPA, dilated tortuous RCA arising from the right coronary sinus with multiple inter coronary collateral formation from RCA to LAD and LCX.

The patient was prescribed aspirin 150 mg daily after the operation. The patient improved symptomatically and was well after five months of surgery without any complications. Her transthoracic echocardiogram done five months after surgery showed improvement in LV systolic function with LV ejection fraction (EF) of 45% by Simpson method (Figure 2 (C)). Patient is now well after two years of follow up.

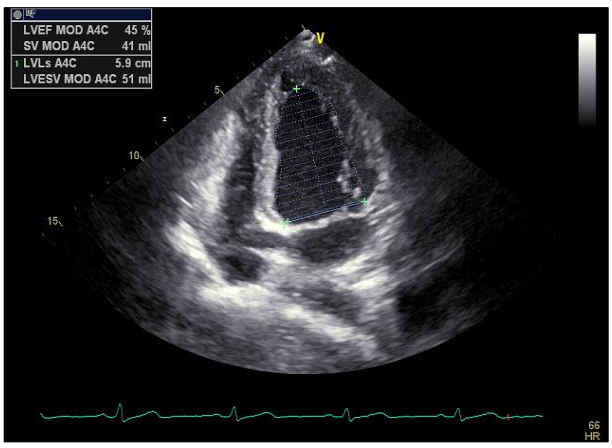

Figure 2(C): Two-dimensional transthoracic apical four-chamber view showing improved left ventricular systolic function with LVEF 45% by Simpson method after five months of surgery.

DISCUSSION

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare but serious congenital cardiac anomaly. In fetal and early neonatal life, ALCAPA is well tolerated because pulmonary arterial pressure equals systemic pressure but after birth, when pulmonary arterial pressure decreases in 7 to 10 days, flow in the LMCA decreases. This is called coronary steal phenomenon and leads to myocardial ischemia, infarction and congestive heart failure. Without treatment approximately 90% infants die within the first year of life.[3] In the adult type, survival to adulthood is dependent on the development of collateral circulation. Older ALCAPA patients clinically present as syncope, cardiac arrhythmias, exertional fatigue, nocturnal dyspnea and less commonly angina pectoris.[4] The average age of sudden death in untreated adult ALCAPA is 35 years and the defect is sometimes discovered during necropsy after sudden death.[5,6] Our patient reached adult age due to development of extensive collaterals from RCA to LAD and LCX as revealed by MDCT angiography and coronary angiography.

Historically, ALCAPA was diagnosed by conventional coronary angiography. However, MDCT coronary angiography and magnetic resonance imaging (MRI) are valuable noninvasive modalities that can be used to identify and define anomalous coronary arteries and their course with a very high accuracy.[7,8] MDCT coronary angiography was done to substantiate the diagnosis of ALCAPA in our patient. Surgical treatment immediately after diagnosis, with the aim of repairing the abnormal coronary circulation system is recommended. Direct re-implantation of the anomalous left coronary artery into the aorta by transferring it with a button of pulmonary artery is considered the standard treatment.[9] Takeuchi S et al, described an alternative approach in which an aortopulmonary window is created and an intrapulmonary tunnel is fashioned that directs blood from the aorta to the left coronary ostium.[10] There are several surgical alternatives for the establishment of a double coronary system, including subclavian-coronary artery anastomosis, ligation of the left coronary artery combined with coronary artery bypass grafting (CABG) with a saphenous vein graft or an internal thoracic artery as was done in our case.[11-13]

The mechanism of improvement in ventricular function following surgery in our patient is explained by the fact that, ALCAPA is an example of chronically hypo perfused myocardial fibers, supporting the hypothesis of a hibernating myocardium, in which myocyte adaptation to chronic ischemia and down regulation of contractility leads to systolic dysfunction but with preserved myocardial viability. The restoration of perfusion in hibernating myocardium after surgery leads to marked and rapid improvement in LV systolic function.[14,15] The extreme LV dilatation with a proportionately lesser increase in myocardial mass contributes to a very elevated LV wall stress and consequently to an increase in oxygen requirement. Thus, a severe imbalance of myocardial oxygen supply and demand appears to result in severe ischemia of entire LV myocardium. The

restoration of adequate oxygen supply to jeopardized myocardium is associated with improvement of LV systolic function, as occurred in our patient.[16]

CONCLUSION

In conclusion, although ALCAPA is a rare congenital anomaly, when an adult patient is admitted for dyspnea and chest pain, and has a continuous murmur, adult type ALCAPA should be suspected, because early surgical correction leads to marked improvement in morbidity and mortality.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Menahem SA, Venables AW. Anomalous left coronary artery from the pulmonary artery: a 15-year sample. Br Heart J. 1987;58(4):378-84.
2. Moodie DS, Fyfe D, Gill CC, Cook SA, Lytle BW, Taylor PC, et al. Anomalous origin of the left coronary artery from the pulmonary artery (Bland-White-Garland syndrome) in adult patients: long-term follow-up after surgery. Am Heart J. 1983;106(2):381-8.
3. Wesselhoeft H, Fawcett JS, Johnson AL. Anomalous origin of the left coronary artery from the pulmonary trunk: its clinical spectrum, pathology, and pathophysiology, based on a review of 140 cases with seven further cases. Circulat. 1968;38(2):403-25.
4. Wilson CL, Dlabal PW, Holeyfield RW, Akins CW, Knauf DG. Anomalous origin of left coronary artery from pulmonary artery. Case report and review of literature concerning teen-agers and adults. J Thorac Cardiovascul Surg. 1977;73(6):887-93.
5. Alexi-Meskishvili V, Berger F, Weng Y, Lange PE, Hetzer R. Anomalous origin of the left coronary artery from the pulmonary artery in adults. J Cardiac Surg. 1995;10(4):309-15.
6. Fernandes ED, Kadivar H, Hallman GL, Reul GJ, Ott DA, Cooley DA. Congenital malformations of the coronary arteries: the Texas Heart institute experience. Annals Thorac Surg. 1992;54(4):732-40.
7. Horisaki T, Yamashita T, Yokoyama H, Urasawa K, Kitabatake A. Three-dimensional reconstruction of computed tomographic images of anomalous origin of the left main coronary artery from the pulmonary trunk in an adult. Am J Cardio. 2003;92(7):898-9.
8. Khanna A, Torigian DA, Ferrari VA, Bross RJ, Rosen MA. Anomalous origin of the left coronary artery from the pulmonary artery in adulthood on CT and MRI. Am J Roentgenol. 2005;185:326-9.
9. Neches WH, Mathews RA, Park SC, Lenox CC, Zuberbuhler JR, Siewers RD, Bahnson HT. Anomalous origin of the left coronary artery from the pulmonary artery: a new method of surgical repair. Circulat. 1974;50(3):582-7.
10. Takeuchi S, Imamura H, Katsumoto K, Hayashi I, Katohgi T, Yozu R, Ohkura M, et al. New surgical method for repair of anomalous left coronary artery from pulmonary artery. J Thorac Cardiovascular Surg. 1979;78(1):7-11.
11. Meyer BW, Stefanik G, Stiles QR, Lindesmith GG, Jones JC. A method of definitive surgical treatment of anomalous origin of left coronary artery. A case report. J Thorac Cardiovasc Surg. 1968;56(1):104-7.
12. El-Said GM, Ruzyllo W, Williams RL, Mullins CE, Hallman GL, Cooley DA, et al. Early and late result of saphenous vein graft for anomalous origin of left coronary artery from pulmonary artery. Circulat 1973;48 (l): III2-6.
13. Kitamura S, Kawachi K, Nishii T, Taniguchi S, Inoue K, Mizuguchi K, et al. Internal thoracic artery grafting for congenital coronary malformations. Annals Thoracic Surg. 1992;53(3):513-6.
14. Cabrera AG, Chen DW, Pignatelli RH, Khan MS, Jeewa A, Mery CM, et al. Outcomes of anomalous left coronary artery from pulmonary artery repair: beyond normal function. Annals Thoracic Surg. 2015;99(4):1342-7.
15. Michielon G, Di Carlo D, Brancaccio G, Guccione P, Mazzera E, Toscano A, et al. Anomalous coronary artery origin from the pulmonary artery: correlation between surgical timing and left ventricular function recovery. Annals Thoracic Surg. 2003;76(2):581-8.
16. Rein AJ, Colan SD, Parness IA, Sanders SP. Regional and global left ventricular function in infants with anomalous origin of the left coronary artery from the pulmonary trunk: preoperative and postoperative assessment. Circulat. 1987;75(1):115-23.

Cite this article as: Baroopal A, Mathur R, Sanghvi S, Mahajan S. Anomalous left coronary artery from pulmonary artery in an adult. Int J Res Med Sci 2019;7:1968-71.