ALCAPA Presenting as Acute Coronary Syndrome in an Adult: An Interesting Case Report with Short Review of Literature

Mondal Prakas Chandra[1], Bera Debabrata[*1], Gupta Ashish[2] and Tiwari Sudama[1]

[1]*Department of Cardiology, Apollo Gleneagles Hospitals, Kolkata, India.*

[2]*Department of Cardiology, Burdwan Medical College, Burdwan, India.*

ABSTRACT

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital anomaly with an incidence of 1 in 300,000 live births and represents 0.25 to 0.5 per cent of all cases of congenital heart disease. Most of the cases (85%) are diagnosed within first month of life. Nearly 90 per cent of untreated patients die within one year with myocardial ischemia and infarction or congestive cardiac failure with mitral regurgitation. However, a few patients can survive into adulthood due to adequate collateral blood supply to the left coronary circulation through dominant right coronary artery. Regional wall motion abnormality is commonly seen in most of the patients with ALCAPA surviving till adult life. Adult ALCAPA can present as effort angina due to relative ischemia and coronary steal. Acute presentation though common in infancy, is relatively rare in patients who survive to adulthood due to adequate collaterals. Here we present a 44 year male patient presenting as acute coronary syndrome and being finally diagnosed as adult ALCAPA in the cath lab. The patient was successfully treated with surgery.

Key words: Acute coronary syndrome, Anomalous origin of the left coronary artery from the pulmonary artery, Coronary anomalies, Myocardial infarction, Echocardiography.

Key Messages: ALCAPA can present in adults and can have variable expression ranging from asymptomatic LV dysfunction, effort angina, and acute coronary syndrome to refractory arrhythmia or even sudden cardiac death. Though it is a congenital coronary anomaly it should still be kept in mind while dealing with an adult patient with such type of presentation. Echocardiography can sometimes miss the diagnosis, especially in adult patients, even if specially looked for.

INTRODUCTION

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital anomaly occurring in about 1 in 300000 births.[1] Anomalous origin of a coronary artery was first described in 1882 by Brooks [2] However, the first detailed clinical description was not published until 1933 who described[3] it in elaboration and the condition is now also known as Bland-White-Garland-syndrome. The typical clinical course is severe left sided heart failure and significant mitral valve insufficiency, presenting at the age of one to two months, when the pulmonary vascular resistance drops. Initial symptoms are feeding difficulties, irritability, diaphoresis, tachypnea and tachycardia. Chest pain due to myocardial ischemia may sometimes be mistaken with infantile colic. Around 85% of all cases of ALCAPA present within the first two months of life. However, symptoms may be misinterpreted in the young infants or even may be absent.

However, in some cases collateral blood supply from the right coronary artery may be sufficient and the patient could pass through childhood with relatively minor symptoms or no symptoms at all. In adult life symptoms may range from dyspnea and exercise intolerance to severe heart failure, mitral valve insufficiency, angina or arrhythmias generated from myocardial scar tissue.

***Corresponding address:**
Mr. Debabrata Bera
Department of Cardiology, MD, DM Associate Consultant,
Apollo Gleneagles Hospitals,
58, Canal Circular Road, Kolkata-700054, India.
Phone no : +91 8013894181 Fax no : +91 33 2320 5218
E-mail: debu2000pgi@gmail.com

DOI: 10.5530/jcdr.2015.1.8

The clinical expression of syndrome results from evolving morphological-functional alterations in pulmonary circulation that occur after birth. Soon after birth, resistance of the pulmonary circulation is high, permitting antegrade flow from the pulmonary artery (PA) to left coronary artery (LCA), which perfuses the left ventricle. Therefore, symptom is extremely rare in this age group. As pulmonary vascular resistance falls in following weeks flow from PA to LCA stops and left ventricular perfusion totally depends upon collaterals to LCA developed from right coronary artery (RCA). Death ensues if collaterals are poorly developed, while on the other hand if collaterals open up after an initial period of decompensation, improvement and survival into adulthood occurs - so called adult type of ALCAPA. In patients who survive into adulthood, pulmonary circulation acts as low resistance outlet and collaterals use LCA as only conduit into pulmonary circulation thus bypassing the left ventricular myocardium. This coronary steal may cause overt ischemia as well as left ventricular diastolic overload from left to right shunting when the shunt is large.[4] They also develop LV systolic dysfunction with mitral regurgitation (MR) usually in infancy.

Rarely ALCAPA has a late presentation in adulthood as few of these patients survive past childhood without surgical repair.[5] Among the reported adult cases, most of them, if not all, presented with evidence of irreversible impairment of cardiac function such as severe dilated cardiomyopathy, sudden cardiac death, acute myocardial infarction, malignant arrhythmias secondary to myocardial scar tissue, impaired LV contractile function, and development of significant mitral regurgitation.[6] There are few case reports of ALCAPA surviving into adulthood but without any LV dysfunction.[7]

CASE HISTORY

A 44 years old gentleman was admitted with acute onset chest pain for 3 hours. He was a smoker for 13 years but non-diabetic and non-hypertensive. On clinical examination he was mildly dyspnic. His blood pressure was 120/76 mm of Hg. His chest was having bilateral basal crepitation. Cardiovascular examination revealed S3 gallop. His chest discomfort had resolved by the time he reached emergency after 16 hours of onset of acute chest pain. He had history of effort angina (CCS II) for last 2 years. His ECG showed complete LBBB and prolonged PR interval (Figure 1). His cardiac biomarkers were significantly elevated (both CPK-MB and Troponin I) at admission. His initial echocardiography showed mildly compromised LV function (EF 45%) grade 1 diastolic dysfunction, RWMA present with akinetic anterior wall and anterior 2/3rd of septum, moderate MR. Thus a diagnosis of acute coronary syndrome (ACS) (NSTEMI) with ischemic MR was made. We did not have any previous ECG to suggest whether his LBBB is new-onset or pre-existing. He was stabilized medically with antiplatelets and enoxaparine and was posted for coronary angiography on 3rd day of admission. Surprisingly it showed only the RCA originating from aorta and the left system was filled retrogradely form the RCA with collaterals (Figure 2). The left system was draining into the pulmonary artery.

After being diagnosed as ALCAPA his echocardiography was repeated which still did not reveal any abnormal flow into pulmonary artery form the LMCA. But it showed only single coronary (RCA) [which was dilated] arising from the

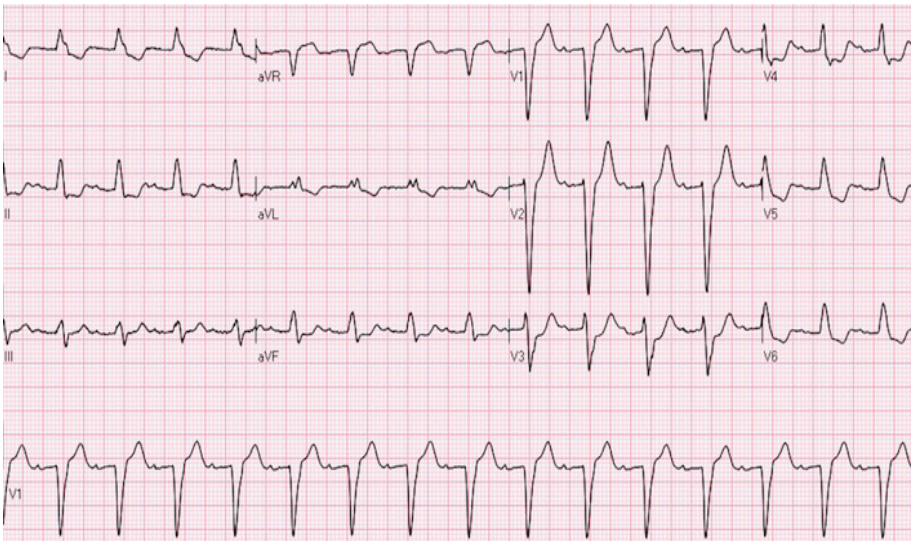

Figure 1: ECG showing complete LBBB with prolonged PR interval.

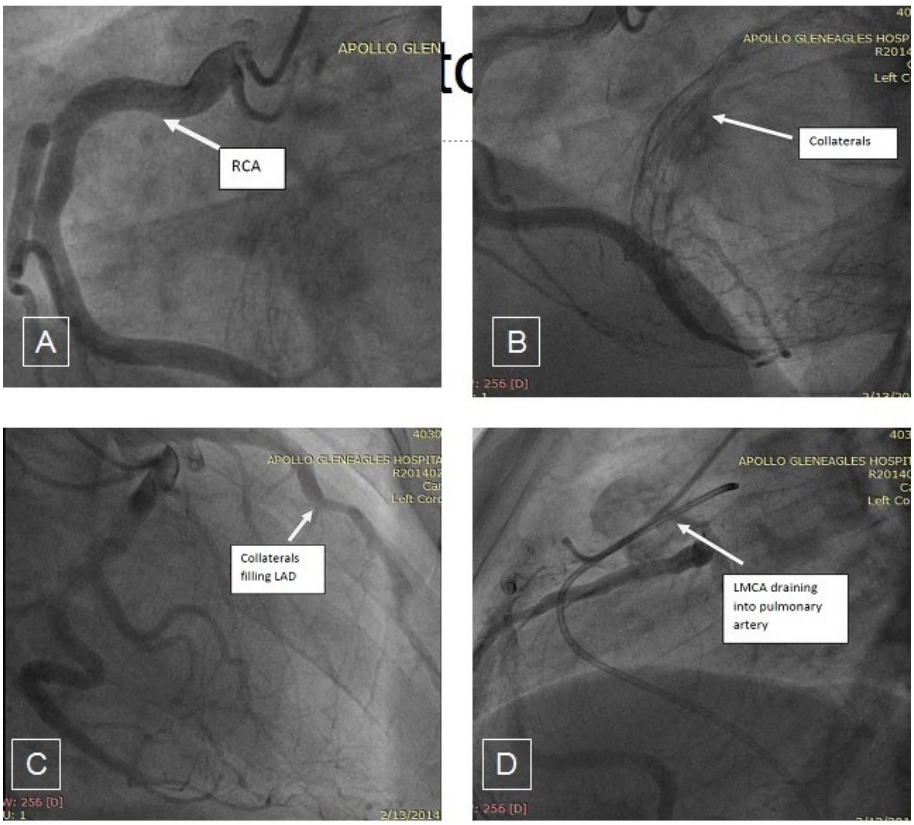

Figure 2: 2A: single coronary artery (dilated RCA) from aortic sinus. 2B and 2C: Collaterals from RCA filling left system retrogradely. 2D: LAD draining into pulmonary artery retrogradely.

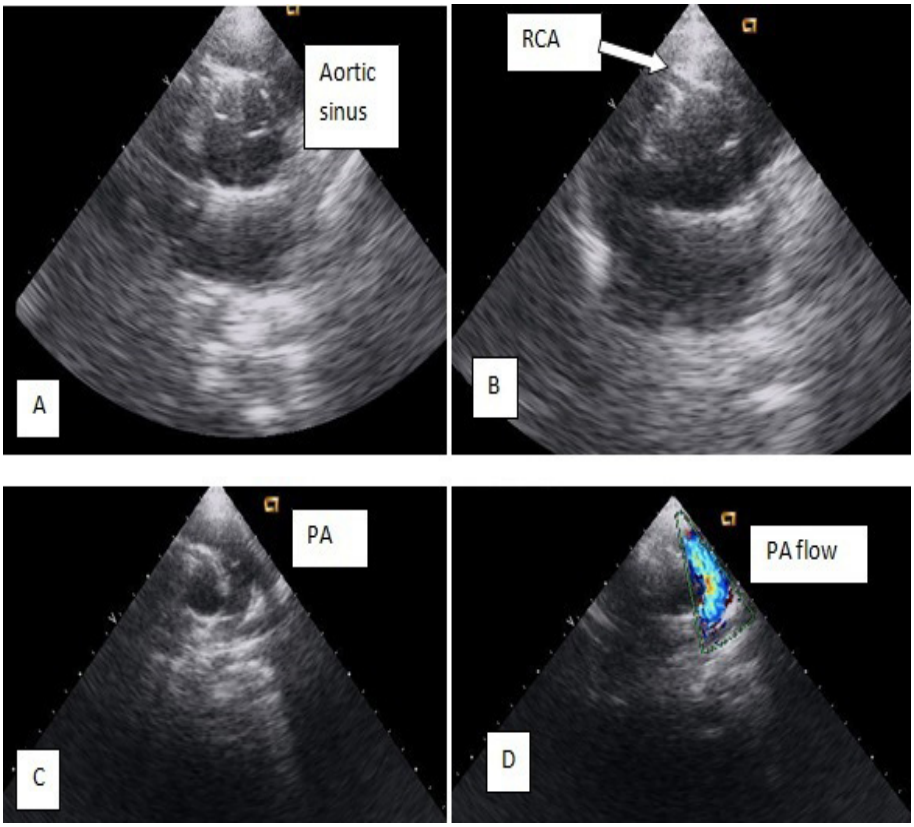

Figure 3: 3A and 3B: Aortic sinus showing origin of only RCA from aortic sinus. No LMCA origin could be found. 3C and 3D: No abnormal flow could be seen into pulmonary artery.

aorta. The origin of LMCA from the left aortic sinus could not be delineated (Figure 3).

The patient was treated with definitive surgery and is doing well on follow up.

DISCUSSION

We present a case of adult ALCAPA who mimicked an acute coronary syndrome (with myocardial infarction and LBBB). Literature review suggests that ALCAPA can present in adults mostly with LV systolic dysfunction. There are few case reports that they present with effort angina and chest discomfort but ultimately diagnosed as ALCALA on echocardiography or during angiography.[5-7] There is no case report describing any patient presenting as STEMI/new onset LBBB. A review article on STEMI-mimicker also does not enumerate ALCAPA as one of the mimicking culprit.[8] But our case shows that they can also present with symptoms of acute ischemia and may mimic acute myocardial infarction due to the pre-existing LBBB. The pathophysiology could be similar to coronary steal by the left system from the single functional coronary system (RCA). The supply demand mismatch can cause acute coronary insufficiency in childhood or can present late with gradually progressing LV systolic dysfunction. They can also present with acute coronary insufficiency despite profuse collaterals during the increased body demand. The least symptomatic ones can have preserved LV systolic function among whom a group can have symptoms of dyspnea despite normal LV systolic function. This is due to diastolic dysfunction and increased LVEDP.

In adults continuous murmur of collaterals are usually found but in presence of PAH the murmur can be purely systolic and can mimic MR or TR. On electrocardiography signs of infarction are absent in adults, which is consistent with our finding as electrocardiograph showed only LBBB with ST/T wave changes in leads I, avL and V6.[4]

Wesselhoft *et al*[1] classified the clinical spectrum of ALCAPA as follows:

- Infantile Syndrome: This is the most common form. Infants develops acute episode of respiratory insufficiency, cyanosis, irritability and profuse sweating. Most of them die within two years.
- Mitral Regurgitation: It is characterized by mitral regurgitation, congestive heart failure, cardiomegaly and atrial arrhythmias in children, adolescent and adults.
- Syndrome of Continuous Murmur: This occurs in asymptomatic patients with angina pectoris. A continuous murmur results from great volume of blood flowing through collateral branches between right and left coronary arteries.
- Sudden Death in Adolescents or Adults: Most of the patients are asymptomatic, but some may experience angina on exertion, cardiac arrhythmias and sudden death.

The adult form of ALCAPA may occur in approximately 10-15% of cases. It is characterized by exuberance in collateral coronary circulation, which allows survival until adulthood, with cases being reported up to the age of 72 years.

Surgical correction is the method of choice as soon as possible with medical management as only supportive and temporary. The two accepted corrective procedures for ALCAPA are ligation of the anomalous left coronary artery and reconstruction of a double coronary artery system. The two coronary artery system can be established in many ways: direct implantation of anomalous left coronary artery to aorta, aorta - left coronary artery saphenous vein graft, left coronary artery conduits using left common carotid artery or subclavian artery, left internal thoracic artery or by Takeuchi procedure (creation of an aorto-pulmonary window and an intra-pulmonary tunnel extending from the anomalous ostium to the window).[9]

Alixi-Meskishvili et al studied four adult patients with ALCAPA aged 27, 35, 54 and 60 years in whom clinical improvement was seen in all patients and concluded that patients with adult ALCAPA get benefit from surgical establishment of a two coronary system even at older age.[10]

CONFLICTS OF INTEREST

Authors declared no conflict of interest

REFERENCES

1. Wesselhoft H, Fawcet JS, Johnson AL. Anomalous origin of left coronary artery from pulmonary trunk: its clinical spectrum, its pathology and pathophysiology based on a review of 140 cases with seven further cases. Circulation 1968; 38(2): 403-25.
2. Brooks HS. Two Cases of an Abnormal Coronary Artery of the Heart Arising from the Pulmonary Artery: With some Remarks upon the Effect of this Anomaly in producing Cirsoid Dilatation of the Vessels. J Anat Physiol. 1885; 20: 26-9.
3. Bland EF, White PD, Garland J. Congenital anomalies of the coronary arteries: Report of an unusual case associated with cardiac hypertrophy. American Heart Journal 1933; 8: 787-801.
4. Parale GP, Pawar SS. Adult Type Anomalous Left Coronary Artery from Pulmonary Artery. J Assoc Physicians India 2006; 54: 397-9.
5. Singh TP, Di Carli MF, Sullivan NMM, Leonen MF, Morrow WR. Myocardial flow reserve in long-term survivors of repair of anomalous left coronary artery from pulmonary artery. Journal of the American College of Cardiology, vol. 31, no. 2, pp. 437–443, 1998.
6. Nielsen HB, Perko M, Aldershvile J, Saunamaki K. Cardiac arrest during exercise: anomalous left coronary artery from the pulmonary trunk. Scand Cardiovasc J. 1999; 33: 369-71.
7. Liu Y, Miller BW. ALCAPA presents in an adult with exercise intolerance but preserved cardiac function. Case Reports in Cardiology 2012.
8. Gu YL, Svilaas T, van der Horst ICC, Zijlstra F. Conditions mimicking acute ST- segment elevation myocardial infarction in patients referred for primary percutaneous coronary intervention. Neth Heart J. Oct 2008; 16(10): 325–31.
9. Arciniegas E, Farooki ZQ, Hakimi M, Green EW. Management of anomalous left coronary artery from the pulmonary artery. Circulation 1980; 62(2): I 180-I 189.
10. Alexi-Meskishivili V, Berger F, Weng Y. Anomalous origin of the left coronary artery from the pulmonary artery in adults. J Card Surg. 1995; 10(4): 309-315.