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Case Reports

Anomalous Origin of Left Coronary Artery from the Pulmonary Artery (ALCAPA) in an Infant - A Case Report

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Introduction

Anomalous origin of the left coronary artery (LCA) from the pulmonary artery (abnormal left coronary artery from pulmonary artery = ALCAPA) or Blade-White-Garland syndrome is a rare, but serious congenital cardiac malformation. ALCAPA represents one of the most common causes of myocardial ischaemia and infarction in children. If left untreated, the mortality rate is upto 90% within the first year of life. ALCAPA accounts for 0.25-0.50% of all congenital heart diseases.[1] Although the anomalous origin of the coronary arteries that arise from the pulmonary artery was first described in1886.[2] It was not until 1933 when Bland et al[3] described the first clinical features with an autopsy finding of anomalous left coronary artery arise from the pulmonary artery (ALCAPA). The anomaly has thus been called the Bland-White-Garland Syndrome.[3,4] A coronary artery anomaly may involve an abnormal number, origin and/or course, termination or structure of the coronary arteries.[5] The left main coronary artery normally arises from the left coronary sinus of valsalva. Instead of arising from the left aortic sinus, it may arises from the right sinus of valsalva or the proximal right coronary artery, from the non-coronary (posterior) aortic sinus, from the proximal part of the ascending aorta as well as from the pulmonary trunk or artery.[6] There are mainly two types of ALCAPA i.e. the infant type and the adult type, each of which has different manifestations and outcomes. Approximately 90% infant dies within the1st year of life, due to myocardial infarction and congestive heart failure (CHF). Restoration of a dual-coronary- artery system is the ideal surgical treatment for ALCAPA syndrome.[7] Here we report a case that presented in early infancy with respiratory distress and was diagnosed as ALCAPA on echocardiography.

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Case Report

Daniel, a 4 months old male infant, 1st issue born to a non-consanguineous parents from Sylhet was admitted to Dhaka Shishu Hospital with the complaints of cough and cold for 5 days & respiratory distress for 3 days. He had history of intermittent episodes of breathing difficulty associated with feeding difficulty & forehead sweating since 2 months of age. Mother was on regular antenatal checkup and had no history of antenatal fever with rash, hypertension or gestational diabetes mellitus. The baby was delivered at term by NVD at home. He had no history of cyanosis. With these complaints the baby was treated by several physicians initially with oral antibiotics & bronchodilator followed by injectable antibiotic, anti failure medication like Frusemide and Digoxin at local medical college hospital as a case of Pneumonia with Heart failure. Color doppler echocardiography was done by adult cardiologist & was diagnosed as a case of Dilated Cardiomyopathy (DCM). As the child was not improving satisfactorily he was referred to Dhaka Shishu Hospital for further investigation and proper management. On examination the baby was a febrile, acyanotic, SP02 was 96% in room air, mildly pale, dyspneic (RR-58/min), normotensive and having tachycardia, (HR-170 /min). All the peripheral pulses were palpable having no radio-femoral delay but pulse

volume was low, capillary refilling time was 2 seconds. Precordium was normal in shape, apex beat was at left 5th intercostal space just lateral to the mid clavicular line. There was no left parasternal heave, thrill or palpable P2. S1 & S2 were audible in all area but S1 was soft. There was a pansystolic murmur best heard over apical area, grade 3/6 and radiating towards the left axilla. Breath sound was vesicular without any added sound. There was mild hepatomegaly. Our provisional diagnosis was Dilated cardiomyopathy (DCM) with mitral regurgitation (MR) with Heart failure. Differentially we thought of viral myocarditis or ALCAPA. Chest X-ray showed huge cardiomegaly (Fig.-1). There was pathological Q wave in lead-I, aVL and T-wave inversion in lead I, aVL, V4 through V6 on electrocardiogram (Fig.-2).

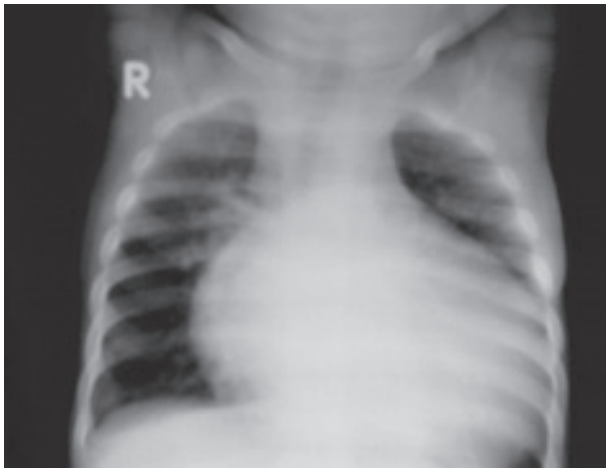

Fig-1:*Cardiomegaly on Chest X-ray.*

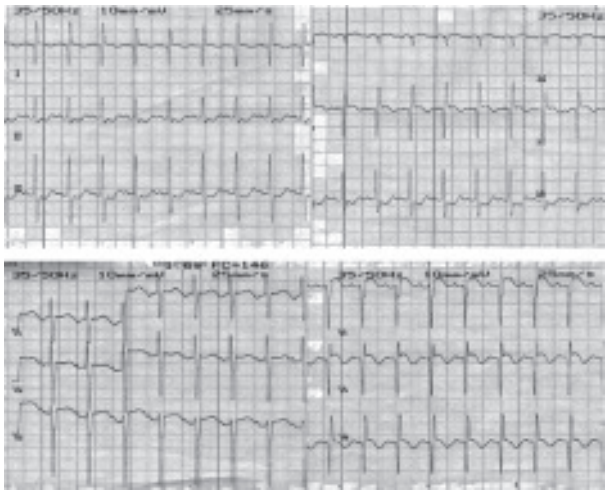

Fig-2: *ECG-Abnormal Q wave in Lead I and aVL & T inversion in Lead I, aV L, V4-V6.*

Color Dopplere chocardiogram revealed that the left coronary artery arises from the left posterior sinus of pulmonary artery and diastolic flow seen in pulmonary artery from left coronary artery (coronary steal), moderate MR, hugely dilated LV with global hypokinesiaand severe LV systolic dysfunction (LVEF-39.2%). The baby was treated with O2 inhalation, Inj. Frusemide, Tab. Spironolactone, Tab. Enalapril and Tab. Digoxin. After having clinical improvement the baby was discharged & reffered to an advanced cardiac center at Dhaka for immediate surgical correction.

Discussion

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital heart defect with an incidence of 1 in 300,00 live births and is the most common cause of myocardial infarction in children.[8] ALCAPA may result from abnormal separation of the conotruncus into the aorta and pulmonary artery or from persistence of the pulmonary buds together with involution of the aortic buds that eventually form the coronary arteries.[9] There are four types of ALCAPA defined by the path the left coronary artery takes after arising from the right coronary sinus. The path of the first type is anterior to right ventricular outflow tract before reaching the anterior sulcus, the usual area of bifurcation. The second type courses behind the right ventricular outflow. The third one courses dorsal to the ascending aorta. These three types in absence of atherosclerotic plaque obstruction are benign. The fourth type arises from the right sinus of Valsalva and passes obliquely between the aorta and pulmonary trunk. This latter type is the only one predisposing to sudden death.[10]

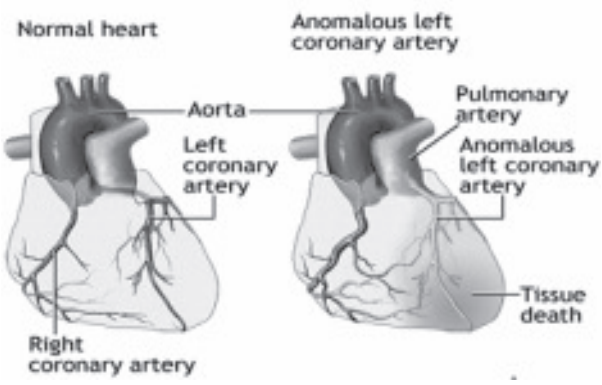

Fig.-3:*Normal coronaries & ALCAPA.*

ALCAPA usually manifests as an isolated defect, but in 5% of cases it may be associated with other cardiac anomalies such as Atrial septal defect, Ventricular septal defect, and Coarctation,[11] it also may be

associated with Tertralogy of Fallot (TOF), Transposition of great arteries (TGA) and Patent ductus arteriosus (PDA).[5]

In the neonatal period, the baby is asymptomatic as there is anterograde flow of desaturated blood from the pulmonary artery to the left coronary artery. As pulmonary arterial pressure drops, the combination of low flow and desaturated blood causes myocardial ischaemia, especially during exertion. Collateral vessels develop between the right and left coronary arteries. Further decreases in pulmonary arterial pressure result in reversal of flow, as the left coronary artery drains from the right coronary artery, through collaterals, into the pulmonary artery. This is known as myocardial steal or coronary steal.[12,13]

In the infants, the chief symptom was irritability elicited by only slight effort, such as feeding, with signs of poor peripheral perfusion. These features started within 2 months of birth, coinciding with substantial reduction in pulmonary vascular resistance that resulted in coronary steal from the anterolateral aspect of the LV. The ECG shows signs of ischemia accompanied the deteriorating ventricular function. This progression created strong suspicion of ALCAPA.[14]

Clinical finding sinlate presenters (adults) with ALCAPA were investigated by Yau et al[15] in a large review including 151 patients. They found a predominance of females 2:1, 66% presented with angina, dyspnoea, palpitations or fatigue. A further 17% presented with ventricular arrhythmia, syncope or sudden death and 62% of these had no antecedent symptoms.[15]

The diagnosis requires a high index of suspicion during history and physical examination.[9] Objective findings include Cardiomegaly on chest X-ray[14], this finding was perfectly present in our patient. In the study by

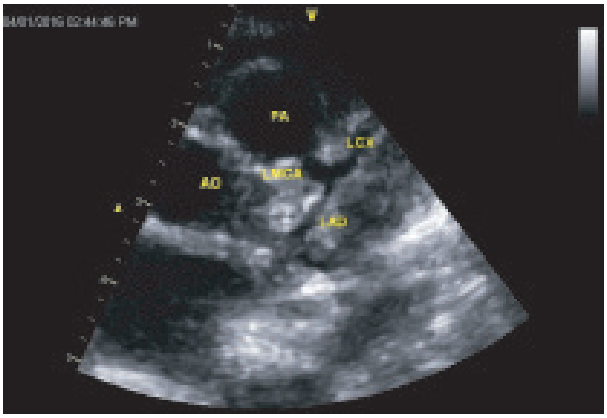

Fig-4: *Short axis view showing ALCAPA with coronary steal.*

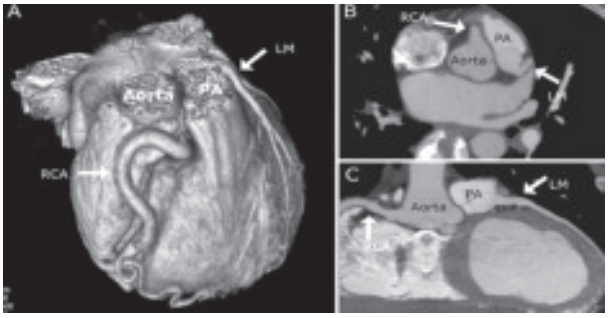

Fig-5: *CT angiogram of ALCAPA*

Yau et al[15], the ECG demonstrated Q-waves in 50%, left ventricular hypertrophy in 28% and left axis deviation in 15%. ECG was normal in 4%. In contrast, symptomatic infants with isolated ALCAPA all had ECG abnormalities with ST-depression, (pathological) Q-wave in V5-V6, and negative T-waves in V5.[16] 2-Dechocardiography may identify ALCAPA and color doppler shows an abnormal jet in the pulmonary artery and of retrograde blood stream in the system of LCA. An other finding is abnormal dilatation of the proximal RCA, an abnormal "brightness" (echogenicity) of left ventricular papillary muscles and sharply delimited sectors of the left ventricular endocardial. Mitral valve regurgitation, left ventricular dysfunction, and wall motion abnormalities may be present.[17]Color doppler echocardiography of our patient done in our Echo lab which revealed anomalous origin of left coronary artery from the left posterior sinus of the pulmonary artery with diastolic flow in the pulmonary artery from the left coronary artery (coronary steal), Hugely dilated LV and global hypokinesia, mild to moderate MR and severe LV dysfunction (LVEF - 39.2%). Cardiac catheterization and angiography, the classical gold standard for confirming the diagnosis of ALCAPA.[18]

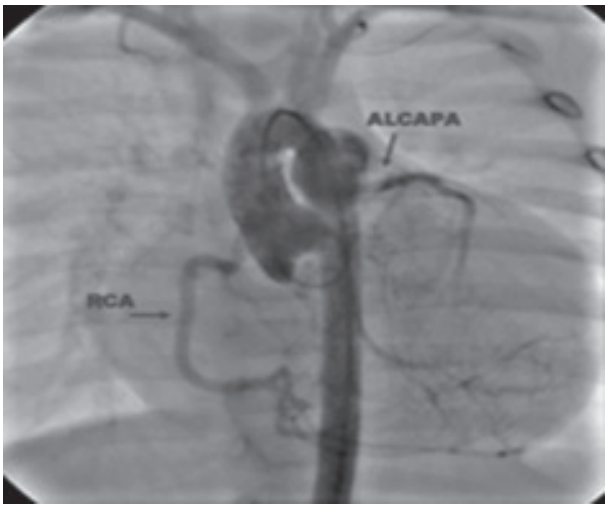

Fig-6: *Cardiac catheterization showing ALCAPA.*

Coronary angiography is indicated in all doubtful cases.

The current recommendation of management is to operate at any age when the diagnosis is made, due to the risk for ventricular arrhythmias and sudden death. Current surgical procedures are directed at establishing revascularization by creating a two-coronary artery system via either (i) Intra pulmonary Tunnel Operation i.e Takeuchi procedure (creation of an aorto pulmonary window and an intrapulmonary tunnel extending from the anomalous ostium to the window) (ii) Left coronary artery implantation, (iii) Subclavian artery to left coronary artery anastomosis, or (iv) Tashirorepair. By establishing a patent two-coronary artery system, most patients experience normalization of LV systolic function and improving long-term survival.[19,20] Based on clinical suspicion supported by X-ray, ECG, we did Echo cardiography which confirmed the diagnosis of ALCAPA.

Conclusion

ALCAPA is a rare but fatal congenital heart malformation. The diagnosis should be suspected in infants who have CHF that suggests DCM and who have ECG changes that suggest anterolateral myocardial ischemia. A combination of a high index of suspicion, typical ECG and echocardiographic findings in a young infant presenting with LV dysfunction could lead to an earlier diagnosis of ALCAPA. Re-implantation of the anomalous LCA in the aortic root is the treatment of choice. Early diagnosis and surgical intervention at optimum time generally results in an excellent prognosis.

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