

Anomalous Origin of Left Coronary Artery from Pulmonary Artery (ALCAPA)

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ABSTRACT

Anomalous origin of the left coronary artery from the pulmonary artery also recognized as Bland White Garland syndrome is a very rare congenital condition. A two-months old baby boy presented with dyspnoea for two weeks and a pansystolic murmur on auscultation. The base line investigations showed cardiomegaly and bilateral basal haze on X-ray chest. ECG showed ST elevation in leads I and AVI and echocardiography showed situs solitus, levocardia, hypokinetic intra-ventricular septum, ejection fraction of 30%, mitral regurgitation of grade-I and an anomalous origin of the left coronary artery from pulmonary artery was diagnosed. Patient was in left heart failure. It was rectified surgically by creating a trans-pulmonary tunnel (Takeuchi repair). Postoperative course was uneventful and he was finally discharged in stable condition.

Key Words: ALCAPA. Bland white garland syndrome. Left ventricular failure. Echo cardiography. Anomalous left coronary artery. Pulmonary artery.

INTRODUCTION

Brooks in 1985 was the first to show that coronary arteries may originate from pulmonary artery.¹ ALCAPA (anomalous origin of left coronary artery from pulmonary artery) occurs in 1 out of 300,000 children and constitutes 0.24% of all congenital heart defects.² It may be an isolated anomaly or be associated with ventricular septal defect, tetralogy of Fallot and truncus arteriosus.³ The anomalous origin of the left coronary artery from the pulmonary artery, also known as Bland White Garland syndrome, requires surgical correction. Myocardial ischaemia and poor left ventricular function are the main consequences. Surgical correction can be done by Takeuchi repair, that demands an extensive intensive care unit stay with protracting inotropic support sometimes requiring left ventricular assisted device and extra-corporeal membrane oxygenation postoperatively. We describe this rare condition in an infant and its management.

CASE REPORT

A 2 months old baby boy was admitted at Armed Forces Institute of Cardiology / National Institute of Heart Diseases (AFIC/NIHD) with history of dyspnoea for the

last two weeks. Dyspnoea was progressive in nature. On examination, pulse was 130/minute. There was a pansystolic murmur on auscultation, otherwise the chest was clear. Chest X-ray showed enlarged cardiac shadow, mediastinal widening, prominent upper lobe vessels and bilateral basal haze (Figure 1). ECG showed ST elevation in leads I and AVI. Pre-operative echocardiography showed situs solitus, levocardia, hypokinetic intraventricular septum (IVS), ejection fraction of 30% and mitral regurgitation grade-I. There was no atrial septal defects/ventricular septal defects (ASD/VSD) and he was diagnosed as a case of anomalous left coronary artery arising from pulmonary artery with retrograde flow into pulmonary artery (Figure 2).

Patient was receiving treatment for left ventricular failure including captopril, digoxin and diuretics. He was planned for surgical correction of ALCAPA. It was done by creating transpulmonary tunnel (Takeuchi repair) procedure. Patient was shifted to the intensive care unit (ICU) after surgery. He was placed on inotropic support



Figure 1: Chest X-ray showing enlarged cardiac shadow.



Figure 2: Left parasternal short axis echo view on echo showing anomalous origin of LCA from PA (arrow).

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including infusion dopamine 7.5 microgram/kg/minute and infusion milrinone 0.5 microgram/kg/minute. He was kept on ventilator for 24 hours. Tablets Captopril and digoxin and injection frusemide were given intermittently after extubation on the second postoperative day. He was given prophylactic antibiotic injections cefoperazone and linezolid and continued for 5 days. Milrinone was halved on first postoperative day and stopped on the third postoperative day. Dopamine infusion was gradually tapered off till the seventh postoperative day. Patient had a smooth course in Intensive Care Unit and was shifted to ward on 7th postoperative day in stable condition. Patient had a stable 6 days stay in the ward and was discharged in a stable condition.

His parents were advised to continue medicines regularly and stay in this station for one week as per our routine protocols. They should report in emergency department in case of any problem, otherwise get checked-up in outdoor paediatric cardiac department after one week and fortnightly for 3 months. They were also counselled about the nature of disease and its potential complications.

DISCUSSION

ALCAPA, a rare congenital disorder was first shown by Brooks in 1885. Patients are usually asymptomatic before 01 month of age because of high pulmonary artery pressure.⁴ Increase in symptoms usually appear at the age of 1 – 2 months because of the left to right shunting from higher left coronary artery (LCA) system to the right coronary artery system. This most often results in death due to circulatory insufficiency from left ventricular dysfunction, mitral regurgitation, myocardial infarction or malignant dysrhythmias. Without treatment, most of children die within first year of life.⁵ Malignant dysrhythmias also exist in asymptomatic adult patients.⁶ The pathophysiology of ALCAPA includes changes in pulmonary artery pressure, intercoronary anastomosis and ischaemic injury to myocardium *in utero* and early neonatal life. Right ventricle performs the systemic workload, ejecting blood through the pulmonary artery, ductus arteriosus and into the aorta. Hence, the perfusion pressure in anomalous left coronary artery originating from pulmonary artery is normal. The left ventricle receives adequate blood supply and functions normally. Later in post-natal period, pulmonary arterial resistance decreases. This results in decreased pulmonary arterial pressures and therefore, decreased perfusion of the left coronary system. At this time, perfusion in the left coronary distribution depends on development of right to left coronary collateral blood flow through the intercoronary anastomosis. Once the anomalous left coronary artery pressure is less than the right coronary artery (RCA) pressure, blood flows retrogradely from right coronary artery (RCA) through

intercoronary anastomosis to the anomalous left coronary artery and drains into pulmonary artery. Because fully oxygenated arterial blood originating from right coronary artery (RCA) now drains into pulmonary artery, a left to right shunt is present, although a volume shunted blood may be insufficient to produce a change in oxygen saturation from right ventricle to pulmonary artery. The absence of antegrade flow in the anomalous left coronary artery leads to myocardial ischaemia. With progression, the left ventricle is generally hypertrophied and dilated, diffuse left ventricular fibrosis sets in and the patient may have evidence of old and recent myocardial infarction. Mitral insufficiency may also be seen. Pre-operatively, these patients are usually stabilized with diuretics and sometimes with inotropic support. Digoxin has debatable role because of ischaemia, may show high incidence of dysrhythmias. Diagnostic application of multidetector row computed tomographic coronary angiography to assess coronary abnormalities in paediatric patients can be done pre-operatively.⁷

Postoperatively, these patients usually require prolonged ICU stay due to poor left ventricle (LV). These patients require inotropic support, sometimes left ventricular assisted device (LVAD), extracorporeal membrane oxygenation (ECMO) or intra aortic balloon pump (IABP) in adult or relatively in elder kids.

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