

CASE REPORT**RARE CASE OF BLAND WHITE-GARLAND SYNDROME WITH MYOCARDIAL INFARCTION IN PAEDIATRICS****Munnaza Saleem, Syed Sajid Hussain Shah, Tahir Mehmood**

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A 3.5 months old female infant presented with acute history of loose motions, fever and irritability for 5 days. The infant had history of respiratory distress since birth that exaggerated while feeding along with sweating and increased milk intake. Patient had poor weight gain. On investigations she was found to have anomalous origin of the left coronary artery from the left pulmonary artery (ALCAPA) with dilated cardiomyopathy and anterior wall myocardial infarction which is a very rare diagnosis in paediatrics and very rare condition diagnosed in our settings.

Keywords: Anomalous origin, coronary artery, myocardial infarction.

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INTRODUCTION

Anomalous origin of the left coronary artery from the left pulmonary artery (ALCAPA) was also previously known as Bland White-Garland syndrome. ALCAPA is a rare but serious congenital heart anomaly of coronary artery and it affects about 1 in 300,000 births.¹

In 1911 the anatomy of ALCAPA was first described by a Russian pathologist Abrikosoff while reporting autopsy of a 5 months old infant with left ventricular aneurysm. The first clinical description of the syndrome was published in 1933 by Edward Bland, Joseph Garland and Paul Dudley who were practicing in Massachusetts General Hospital.²

Patients with ALCAPA who survive childhood often have varying symptoms of myocardial ischemia, impaired left ventricular function, mitral regurgitation and progressive heart failure, depending on the development of collateral circulation. The majority of patients die in infancy.³

CASE REPORT

A three and half months old female infant was admitted through emergency department with history of fever, irritability and decrease oral intake with provisional diagnosis of acute gastroenteritis. On examination the infant was mildly distressed even after correction of fluid deficit. The infant was born via spontaneous vaginal delivery (SVD) to primigravida mother in a local private hospital, duly vaccinated until then. She had a history of respiratory difficulty during breast feeding since birth along with sweating. Though the infant was taking feed adequately yet not thriving well. Physical and systemic examination revealed ejection systolic murmur at left sternal edge grade II/VI with distress. Blood gases, electrolytes and renal function tests were normal. The X-ray chest (CXR) showed cardiomegaly (Figur-1). Echocardiography showed anomalous origin of left coronary artery from pulmonary artery with dilated cardiomyopathy and

anterior wall myocardial infarction (Figures 2-4). Patient was referred to paediatric cardiologist and management was done in collaboration of paediatric cardiologist and follow-up along with referral to paediatric cardiac surgeon. Though surgery was planned by the paediatric cardiac surgeon but unfortunately the patient expired after about a month.

DISCUSSION

ALCAPA syndrome is the commonest cause of myocardial infarction in paediatric patients.⁴ Bland, White and Garland was first to report the association of anomalous origin of left coronary artery from pulmonary artery with a hypertrophied left ventricle and attacks of dyspnoea, pallor and profuse sweating in a boy who died at the age of 3.5 months.² The estimated incidence of ALCAPA syndrome is 1/300,000 live births (0.24% to 0.46% of all congenital cardiac anomalies) as there may be significant underestimation of the true incidence due to undiagnosed deaths of asymptomatic patients.⁵ ALCAPA develops before birth when the systemic and pulmonary arterial pressures are equal and there is ante-grade flow in both left and right coronary arteries. In the neonatal period, ante-grade flow gradually changes as the pulmonary blood pressure diminishes, the ductus arteriosus closes and the flow in the left coronary artery reverses. Right and left coronary artery collateral circulation developments during closure of the duct and lowering of the pulmonary pressure determine the extent of myocardial ischemia. Patients with well established collaterals have been classified as the "adult type" and those with no collaterals as the "infantile type". These functional states actually represent different phases in collateral circulation and the changes engendered by each patient determine clinical course as reported by Edwards.⁶ In very rare cases patients may survive to the sixth or seventh decades of life.⁷

During first 3 months, onset of symptoms is usually observed as infant may present with feeding and crying induces dyspnea, profuse

sweating, pallor, fatigue and perception of pain. Physical examination is usually normal in between attacks. Though signs of heart failure and failure to thrive are common yet cases of totally asymptomatic adult patients have also been reported.⁸ The more extensive the coronary collaterals, the better preserved the myocardial function due to lack of ischemia, although the degree of coronary steal may then become significant.

Pulmonary hypertension develops gradually overtime due to combination of left-to-right shunting, left ventricular dysfunction, and mitral regurgitation. Mitral regurgitation is thought to result from the dysfunction of the ischemic papillary muscles and adjacent myocardium and it can be due to prolapsed or an associated lesion such as mitral cleft or short chordae.⁹



Figure-1: Chest X-Ray showing cardiomegaly



Figure-2: Echocardiography report image



Figure-3: Echocardiography report image



Figure-4: Echocardiography report image

Surgery is recommended in all patients with ALCAPA even in the absence of symptoms or a significant left-to-right shunt syndrome, given the risk of ventricular arrhythmias and sudden death.¹⁰

Although ALCAPA syndrome is a rare condition presenting in adulthood, awareness about it is important as early diagnosis and treatment may prevent irreversible damage to the myocardium and subsequent complications including myocardial infarction, heart failure, mitral regurgitation and sudden death.

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