

Bland-White-Garland Syndrome in a Middle Aged Patient-A Case Report & Review of Literature

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Abstract

Bland-White-Garland syndrome (anomalous origin of left coronary artery from the pulmonary artery) is a very rare congenital anomaly, reported in less than 0.5% of all the congenital heart disease which may result myocardial infarction, congestive heart failure and sometimes death during the early infantile period. We report a case with this anomaly, a 27 year old young lady, mother of one child presented with progressive dyspnea and cardiomegaly on X-ray. The diagnosis was made on noninvasive imaging procedure. This lady is now on optimal medications and follow up.

Key words: Bland-White-Garland (BWG) syndrome; Left coronary artery (LCA); Pulmonary artery (PA).

Introduction

Anomalous origin of left coronary artery (LCA) arising from the pulmonary artery (Bland- White-Garland syndrome also known as ALCAPA syndrome) is a rare congenital abnormality affecting 1 in 300,000 live births accounting for 0.5% of cases of congenital heart disease.¹ This congenital malformation leads to myocardial ischemia in the early infancy. If untreated, myocardial ischemia leads to severe LV dysfunction. Diagnosis before left ventricular (LV) dysfunction, though important, is quite difficult. The younger age and nonspecific symptoms of irritability and feeding problems make it difficult to diagnose even after myocardial ischemia is advanced to the stage of dilated poor LV, because the symptoms are usually considered to be attributed to acute myocarditis/dilated cardiomyopathy^{2,3} or heart failure due to large shunts. Once the timely diagnosis is made, pushing the patient for surgical option and more so the execution of coronary surgery at such a delicate age in our set up is much more than a gigantic task.

Case Report

A 27 years old normotensive, nondiabetic young mother of one child had history of puerperal cardiomyopathy and was admitted with the chief complaints of left sided chest heaviness associated with shortness of breath for three days. She also gave history of exertional dyspnea for 2 years. On physical examination, patient was conscious, oriented, pulse was 60 bpm, regular, respiratory rate was 18/min, BP-110/80 mm Hg and had a very soft pan systolic murmur (grade-1/6) at the apex. Blood routine investigation reports were normal. Chest X-ray showed cardiomegaly. ECG showed left bundle branch block (Fig-1). Echocardiography showed no regional wall motion

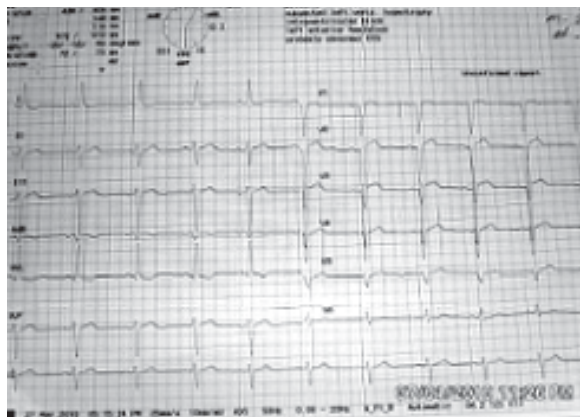


Fig-1: ECG- Left bundle branch block (LBBB)



Fig-2: Echocardiogram: short axis view, shows separate origins of circumflex artery (CX) from the aorta (AO) and LAD coronary artery from pulmonary artery (PA).

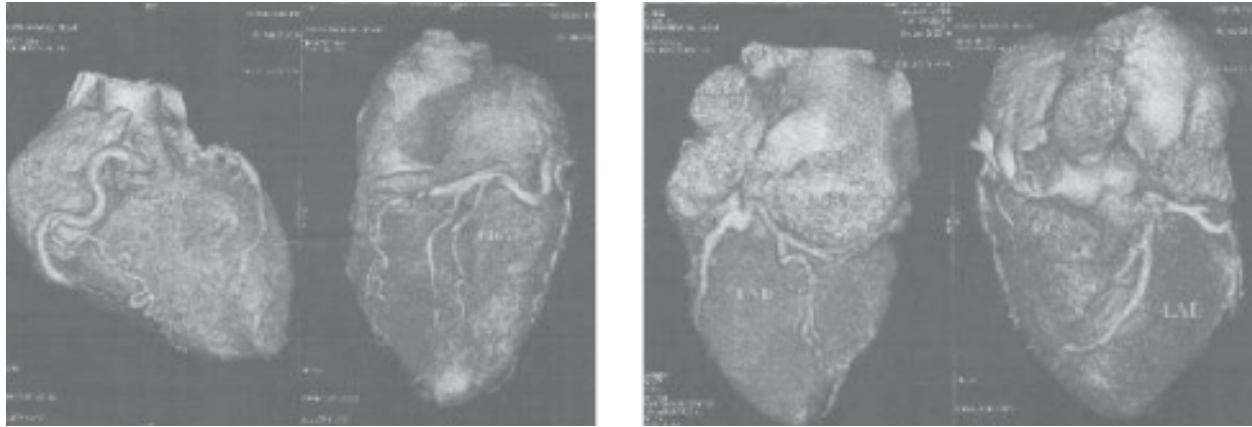


Fig.-3: (left & right)-CT coronary angiogram revealed Left Coronary origin from pulmonary artery.

abnormality with ejection fraction of 60%. There were moderate mitral regurgitation (MR), mitral annular calcification. Parasternal short axis view clearly demonstrated the left coronary artery arising from pulmonary trunk and dividing into left anterior descending and circumflex arteries (Fig-2). A diagnosis of ALCAPA was made by CT coronary angiogram (Fig-3). She was strongly recommended for conventional coronary angiogram. But the patient attendants refused to do conventional CAG. The patient was in optimal medical management and follow up.

Discussion :

Left coronary artery arising from pulmonary trunk is a rare but potentially lethal congenital heart disease with estimated incidence of 1/300,000 live births.^{1,3,4} Bland et al.²⁻⁴ in 1933 were first to correlate its clinical and autopsy findings on an infant. Thus the anomaly is also called Bland-White-Garland syndrome. In the fetus this condition is silent because blood supply to the LV myocardium is adequate due to high pulmonary artery pressures (PAP) and normal antegrade flow in the LCA.² After birth the pulmonary artery contains desaturated blood but as long as the high PAP ensure the antegrade flow from PA to LCA, the baby remains asymptomatic.²⁻⁵ With postnatal drop in the PAP the antegrade flow into LCA decreases and eventual flow reversal into the PA occurs resulting in a "coronary steal".² Initially the myocardial vessels dilate to increase the flow but soon the coronary vascular reserve is exhausted and myocardial ischemia and infarction occurs. Collateral vessels between the right and left coronary arteries develop in attempt to perfuse the LV myocardium but their efficiency is diminished due to "coronary steal".⁴ This usually occurs after the first month of life. Of all the children born with ALCAPA, about 87% present in infancy and about 65-85% die before first

birthday, usually after two months of age.² About 10-15% remains symptoms free and reach adult life, most probably due to large dominant right coronary and well-developed collaterals, but even they are at high risk of sudden death at a mean age of 35 years.³ The oldest reported case with the disease is a 72 years old woman.⁶ Typically the baby is well till 4-6 weeks of age and then gradually presents with profuse sweating, dyspnoea, pallor and irritability on feeding or crying (angina equillans),^{2,3,7} as was the presentation of our case. Physical examination may reveal signs of heart failure and/or mitral regurgitant murmur at the apex or even normal cardiac findings.^{2,3} Clinical diagnosis is extremely difficult even for the experienced physician because many of the more common conditions like acute myocarditis, dilated cardiomyopathy and even large left (L) to right (R) shunts & mixing lesion with unrestricted pulmonary flow may have similar presentation at this age. Standard investigation for the diagnosis is aortic root angiogram^{2,6} but solely for diagnostic purpose, that remains quite risky on an unstable patient.

Therefore many people are of the view that ALCAPA should be diagnosed by noninvasive tools.³ On ECG, broad and deep Q waves in lead I and aVL with absent Q in II, III and aVF is considered characteristic of ALCAPA.⁸ On 2-D echocardiography the abnormal origin of LCA can be visualized.³ Colour flow Doppler can detect an abnormal jet of LCA into PA. A dilated right coronary artery (if present) may also be helpful in the diagnosis.² Combining ECG and echocardiography ALCAPA can be diagnosed with reasonable certainty and with no need of angiography.⁹ In our case we demonstrated on the CT coronary angiogram revealed abnormal origin of LCA from pulmonary trunk. Treatment options include LCA reimplantation and Takeuchi's transpulmonary baffle; both

types establish dual coronary systems.³ Sometimes on a very sick unstable patient simple ligation of the LCA at origin is adequate to stop the “coronary steal” and stabilize the patient for later more optimal surgery.¹⁰ Surgery should be performed even if the LV function is poor because LV function improves and reaches normal within a year with restoration of dual coronary supply.^{11,12}

The present case illustrates that BWG syndrome can be asymptomatic over a long time even during pregnancy. Systemic collateral blood supply to the coronaries may cause for asymptomatic course and patient’s survival.

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