ANOMALOUS ORIGIN OF LEFT CORONARY ARTERY FROM PULMONARY ARTERY - A CASE REPORT

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Abstract:
Anomalous origin of left coronary artery from pulmonary artery is a rare congenital anomaly. It usually manifests in infancy itself. Patient surviving into adulthood is rare in this anomaly. We report a case of Anomalous origin of left coronary artery from pulmonary artery in an 11 years old female child, previously diagnosed and treated as a case of dilated cardiomyopathy. Patient was advised surgical correction. A brief review of literature in Anomalous origin of left coronary artery from pulmonary artery is discussed.

Keyword:
ALCAPA, ALCAPA in Adolescence, Bland-White-Garland syndrome.

Introduction:
Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) syndrome, also known as Bland-White-Garland syndrome, is a rare congenital abnormality that affects 1 of every 300,000 live births (1) and accounts for 0.25%–0.5% of all congenital heart defects. It usually manifest 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 aortic coarctation. Infants experience myocardial infarction and congestive heart failure, and approximately 90% die within the 1st year of life. Rarely, ALCAPA syndrome manifests in adults; it may be an important cause of sudden cardiac death (2). We report 11 years old female, presented in infancy as a case of congestive cardiac failure due to dilated cardiomyopathy, now diagnosed as a case of ALCAPA.

Case report:
A 11 years old first born female child born out of non consanguineous marriage admitted with exertional breathlessness on less than ordinary activity. There was no history of chest pain, palpitation and syncope. There was no history of rheumatic fever, no history of similar ailment in other family members. At 1½ years of age, patient had respiratory distress due to bronchopneumonia for which patient was evaluated.
Echocardiographic examination at that time showed left atrium, left ventricle dilated with global hypokinesia of the left ventricle. Left ventricular Ejection fraction was 38% denoting moderate LV dysfunction. Patient was treated with anti failure measures. Patient had periodical follow-up with improvement in symptoms and ejection fraction over the years.

Patient has normal mental and physical development. Cardiovascular examination showed pulse rate of 92/min, blood pressure 90/70 mm Hg. JVP not elevated, heartsounds are normal in intensity and normal split of S2.2/6 soft early systolic murmur in left sternal border. Investigations showed normal blood counts and normal renal parameters. ECG showed no specific abnormality (figure 1).

X-ray chest showed normal size and shape of heart shadows with normal pulmonary vasculature (figure 2).

Echocardiographic evaluation showed normal size of the chambers with normal LV systolic function. Origin of Right coronary artery is dilated (figure 3).

Left coronary artery normal origin could not be found in left coronary sinus. A anomalous
channel found in the main pulmonary artery with continuous retrograde flow (figure 4,5).

There was no mitral regurgitation, papillary muscles appear to be minimally thickened. There was no regional wall motion abnormality. Coronary angiogram of right coronary artery showed dilated and tortuous right coronary artery which drains into dilated left coronary artery through multiple collaterals which in turn drains into main pulmonary artery (figure 7).

Turbulence noted in the inter ventricular septum with continuous flow (figure 6).
LV angiogram showed no ventricular septal defect and right coronary artery was dilated and tortuous and drains into main pulmonary artery via left coronary artery (figure 8).

Discussion:
Anomalous origin of the left coronary artery arising from the pulmonary artery (ALCAPA) is a rare but serious congenital anomaly. ALCAPA was first described in 1866. The first clinical description in conjunction with autopsy findings was described by Bland and colleagues in 1933, so the anomaly is also called Bland-White-Garland syndrome. The ALCAPA anomaly may result from abnormal septation 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. In this condition, in fetal life no abnormal hemodynamic changes occur as left ventricular myocardium gets adequate blood supply from anomalous left coronary artery from main pulmonary artery due to increased pulmonary vascular resistance (3). In the neonatal period (first stage) left ventricular myocardium gets adequate blood supply from pulmonary artery causing but in about 10% patients adequate collaterals develop (third stage). These patients survive to adulthood.

As the time passes in the children left coronary artery behaves as a conduit (stage four) between right coronary artery and pulmonary artery, so resulting in Coronary steal phenomenon. Left ventricle could not be perfused adequately with oxygenated blood which leads to myocardial infarction or sudden death. Papillary muscle dysfunction due to mitral regurgitation may happen, which produces volume overload of left ventricle. In some cases, presence of ventricular septal defect is beneficial, by increasing pulmonary artery pressure preventing ischemia. Infants after 6 weeks usually present with dyspnea, tachycardia and restlessness with incessant cry during feeding or nursing care or during defecation. Cardiomegaly with LV apex and hepatomegaly indicating congestive cardiac failure which are common signs in infants. In majority of infants, initially no murmur but subsequently develop a short systolic murmur or pan systolic murmur over the apex due to MR. Those who survive and attain adulthood may develop continuous murmur audible over left sternal border due to retrograde flow through inter coronary communications. This continuous murmur does not peak around second heart sound, moreover diastolic part is loud. ECG may be of very much diagnostic in ALCAPA. Typically, an anterolateral infarct pattern with abnormal deep (>3 mm) and wide (>30 msec) q waves is observed in leads I, aVL, V₅, and V₆, absent q waves in leads II, III, and aVF, and poor R wave progression across the precordial leads, with sudden shift to qR. Echocardiography with colour flow imaging is often diagnostic. In ALCAPA left coronary artery origin
left coronary artery origin is absent from aorta, but it can be traced from the pulmonary artery. Right coronary artery shows normal origin, often dilated. Demonstration of retrograde flow from left coronary artery towards pulmonary artery is diagnostic of ALCAPA. ECG-gated multidetector CT angiography and MR imaging are valuable noninvasive modalities that can be used to help diagnose ALCAPA. ECG-gated multidetector CT angiography plays an important role in assessment of ALCAPA syndrome, and it may be a valuable postoperative follow-up tool for adult patients. MR imaging is useful for assessment of left ventricular myocardial viability to determine if asymptomatic adult patients may benefit from surgical correction(4). Our case even though presented with features of congestive cardiac failure and dilated cardiomyopathy in early childhood patient was lucky enough to survive up to this age. Our case demonstrates need for high index of suspicion in these kind of presentations. Factors in infants that enable survival to adulthood are: abundant interarterial collateral vessels between the RCA and the LCA (Retrograde left ventricular perfusion from the RCA), RCA dominance (Smaller myocardial area supplied by the LCA leads to less extensive myocardial ischemia), Minimal coronary steal from the pulmonary artery (Ostial stenosis of the LCA or a restrictive opening into the pulmonary artery limits the left-to-right shunt), development of systemic blood supply to the LCA (Bronchial artery collateral vessels increase oxygenated blood flow and perfusion pressure to the ischemic myocardium)(5). Surgical correction is the gold standard in the therapy for ALCAPA. The most popular and fully anatomical correction of ALCAPA is direct reimplantation of ALCAPA into aorta by a button technique as described by Neches in 1974(6). Takeuchi described a pulmonary artery baffle when direct implantation is not possible for anatomical reasons(7). In adults, internal mammary artery may be used to revascularise the LAD(8). Establishment of two-coronary system is known to cause regression of intercoronary collateral network in course of time irrespective of the type of surgical technique used.

References
