Case Report

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Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery (ALCAPA) in an Infant

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Abstract

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is an extremely rare and fatal, but a treatable congenital anomaly of the coronary circulation. ALCAPA usually presents as myocardial ischemia, cardiomegaly and heart failure in infancy. About 90% of the infants die within the first year of life, if left untreated. Timely diagnosis by echocardiography and coronary computed tomography angiography with surgical correction is life-saving. Herein, we report a six-month-old infant with ALCAPA who presented with dilated cardiomyopathy, congestive cardiac failure and failure to thrive. The patient underwent surgical correction for the same but succumbed to the refractory congestive cardiac failure. All infants who present with cardiomegaly and congestive cardiac failure with electrocardiographic (ECG) changes suggestive of myocardial ischemia, should be evaluated for ALCAPA syndrome.

Keywords: ALCAPA; anomalous origin of the coronary arteries; Bland-White-Garland syndrome; congenital heart disease; dilated cardiomyopathy; echocardiography; heart failure.

Introduction

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital malformation of coronary vessels, in which the left coronary artery originates from the pulmonary artery instead of the left coronary sinus of the aorta.[1] The occurrence of myocardial ischemia is usually rare in the infantile age group.[2] Any infant presenting with symptoms of congestive cardiac failure (or angina equivalents), in the absence of a known structural heart disease, should be evaluated for causes of myocardial ischemia.[2] One of the important and rare causes of myocardial ischemia and congestive cardiac failure in infancy is the Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery (ALCAPA) syndrome.[2] Myocardial ischemia in ALCAPA is precipitated by impaired myocardial perfusion due to the "coronary steal" phenomenon.[2,3] ALCAPA can have a varied range of presentation, ranging from death in infancy to asymptomatic survival into adulthood.[1,2,3] If left untreated, majority of the affected infants die within the first year of life.[1,2,3] Herein, we report a case of ALCAPA with dilated cardiomyopathy, whose diagnosis was delayed elsewhere (before presentation to our institute), probably due to a lack of knowledge of the condition among medical practitioners.

Case Report

A six-months-old-male infant was brought to our institution with the complaints of irritability, increased precordial activity and failure to gain weight noticed by the parents since 3 months of age. There was history of excessive forehead sweating with suck-rest-suck cycle. There was absence of history of cyanosis or cyanotic spells.

The family history was unremarkable. On examination, the infant had tachycardia (166 beats per minute), tachypnea (56 breaths per minute) with subcostal retractions, blood pressure of 92/54 mmHg and oxygen saturation on pulse oximeter (SpO2) of 99% without any supplemental oxygen. His length was 59 cm (< -3 Z-score), weight was 4.5 kg (< -3 Z-score), head circumference was 42 cm (-1 Z-score) and mid-upper arm circumference was 11.5 cm. Cardiovascular system examination revealed laterally and downward displaced apical beat (in the sixth intercostal space, one centimetre lateral to the mid-clavicular line) with a gallop rhythm without any murmurs. Rest of the systemic examination did not reveal any abnormalities.

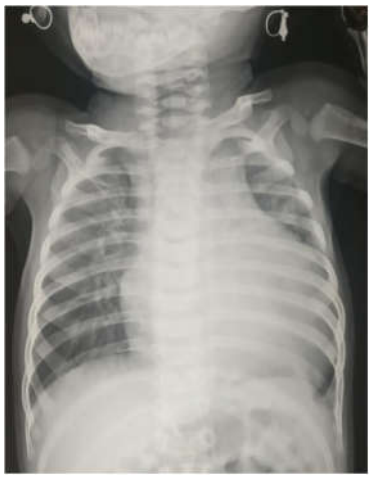

Fig. 1: Chest Radiograph Showing Cardiomegaly (Cardiothoracic Ratio of 0.65).

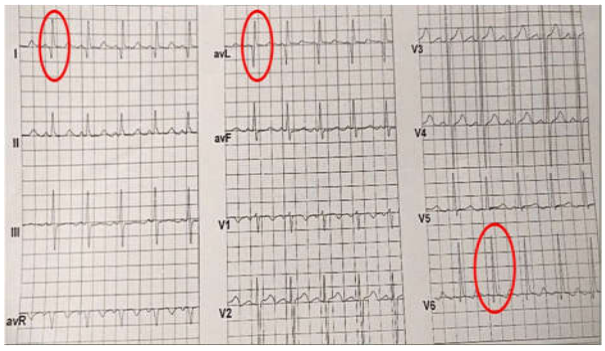

Fig. 2: Electrocardiogram Showing Prominent Q Waves (> 30 Milliseconds) in Leads I, aVL, and V6 Suggesting Myocardial Infarction of the Lateral Wall.

On investigations, his chest radiograph revealed cardiomegaly (cardiothoracic ratio of 0.65) (Fig. 1). The electrocardiogram (ECG) showed prominent Q waves (> 30 milliseconds) in the lateral leads (i.e. in lead I, aVL & V6) suggesting myocardial infarction of the lateral wall (Fig. 2). The transthoracic echocardiogram showed dilated left atrium and ventricle, severe left ventricular (LV) dysfunction with an ejection fraction of 10% and moderate mitral regurgitation with severe pulmonary hypertension. However, the origin of the left main coronary artery from the aorta could not be traced, whereas the right coronary artery was dilated with a normal origin, thus raising the suspicion of anomalous origin of the left coronary artery. A cardiac computed tomography (CT) with coronary angiography confirmed the anomalous origin of the left coronary artery from the pulmonary artery (i.e. ALCAPA) with a dilated and tortuous right coronary artery (Fig. 3).

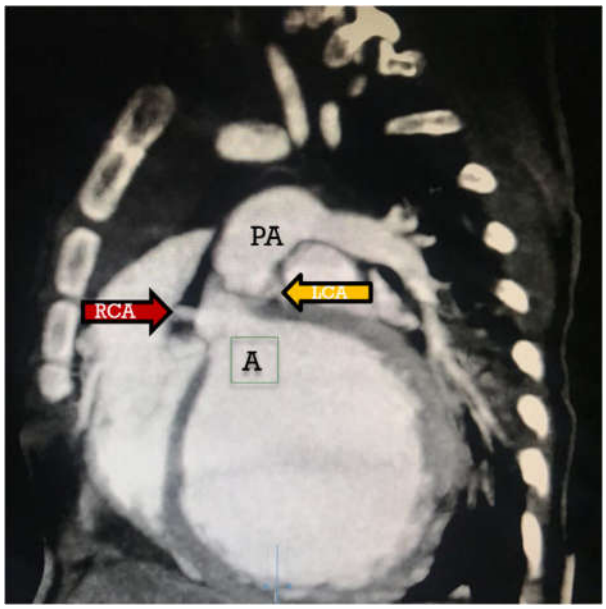

Fig. 3: Cardiac Computed Tomography with Coronary Angiogram Showing Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery (ALCAPA) and a Dilated Right Coronary Artery [PA.- Pulmonary Artery], A-Aorta, RCA- Right Coronary Artery (Wide Arrow), LCA-Left Coronary Artery (Narrow Arrow)].

The congestive cardiac failure was treated with oxygen by nasal prongs (2 L/min), inotropic agent (dobutamine, 10 microgram/kg/min continuous infusion) and diuretics (furosemide, 2 mg/kg/day). After stabilisation, the child underwent surgical correction (modified Takeuchi procedure). However, unfortunately the infant succumbed to refractory cardiac failure on the tenth post-operative day.

Discussion

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) syndrome, also known as the "Bland-White-Garland syndrome" is the most common major congenital malformation

of the coronary circulation, accounting for 0.25% - 0.5% of all congenital heart diseases.[1-4] It usually manifests as an isolated defect, but it may be associated with other congenital heart diseases as well.[2] Various theories have been proposed to explain the anomalous origin of coronary arteries from the pulmonary artery. The coronary arteries originate as two endothelial buds, and displacement of the origin of one or both of these buds can lead to either one or both of the coronary arteries to arise from the portion of the truncus arteriosus (which is destined to become the pulmonary artery).[6,7,8] It has also been hypothesized that faulty division of the truncus can incorporate the coronary arteries into the pulmonary trunk.[6,7,8] The "involution persistence theory" postulates the persistence of the pulmonary artery coronary anlagen, giving origin to the anomalous coronary with involution of the normal aortic coronary anlagen.[6,7,8]

The characteristic patho-physiological mechanism of ALCAPA is the "coronary steal" phenomenon in which left to right shunt leads to abnormal left ventricular perfusion.[1-5] In the fetal and early neonatal life, the origin of left coronary artery (LCA) from the pulmonary artery is well tolerated because the pulmonary artery pressure equals the systemic pressure, which causes the antegrade blood flow in both anomalous LCA and normal right coronary artery (RCA).[2-4] But after birth, as pulmonary artery pressure decreases, blood flow in the LCA decreases initially and then eventually reverses, causing myocardial ischemia and infarction.[1,5] Initially, the viable myocardium is compromised, and hence the cardiac contractility is reduced.[5] Mitral regurgitation then occurs as a result of ischemic papillary muscle dysfunction. Eventually, the blood flow via the inter-coronary anastomoses results in a left to right shunt which is large enough to impose volume overload on the left ventricle.[2,5]

The clinical course of ALCAPA is a continuum, which ranges from death in infancy to asymptomatic adult survival.[2,3] About 90% of the affected patients die within the first year of life, if left untreated.[2,5] Affected infants appear normal at birth and usually do well for a short period before they become symptomatic, which is usually at age of 2 to 3 months (when pulmonary arterial resistance drops to adult level).[2,5] Symptoms may start with a paroxysmal attack of marked pallor, irritability, and sweating.[5] However, not all infants present in this way.[5] Many present with signs and symptoms of congestive heart failure including tachypnea, tachycardia, diaphoresis, poor feeding, and poor weight gain (as was seen in our case).[1,2,5] A few children may outgrow these symptoms and gradually become asymptomatic, although periodic dyspnea, angina pectoris, or sudden death may occur in their adulthood.[1,2,6,7] Chronic congestive cardiac failure is responsible for poor growth and development, as noted in our patient.[1,2,5] Two types of presentation are known- the "infantile" type and the "adult" type. In the "infantile" type, there is little or no collateral circulation between the RCA and LCA resulting in limited blood supply to the left ventricular myocardium, which leads to congestive cardiac failure and sudden cardiac death.[1,2] In the "adult" type, there is adequate collateral circulation from RCA to LCA, however, it is insufficient to supply the left ventricle and hence chronic left ventricular subendocardial ischemia ensues.[1,2,6,7] These patients may be asymptomatic or may present with mitral regurgitation, ischemic cardiomyopathy, or malignant dysrhythmias causing sudden cardiac death.[2,3]

For any infant presenting with heart failure, the ECG can provide an important clue for the diagnosis of ALCAPA.[9] Typical ECG findings include Q waves of > 30 milliseconds in lead I, aVL and V6.[8,9] Echocardiography with color Doppler identifies the origin of an anomalous left coronary artery from the pulmonary artery and also defines the flow patterns (retrograde flow pattern).[1,2,5] In our patient, the echocardiography could not conclusively diagnose the ALCAPA. ECG-gated multi-detector CT angiography and magnetic resonance (MR) imaging are sensitive non-invasive modalities used to diagnose ALCAPA.[3] Secondary findings of imaging like dilated and tortuous RCA and intercoronary collaterals, left ventricular hypertrophy with regional wall motion abnormalities, mitral insufficiency and delayed subendocardial enhancement (on MR imaging) are also seen in ALCAPA syndrome.[1,2,3,5]

Surgical correction is considered to be the standard of treatment, especially in infants, where early correction is associated with improvement in ventricular function.[7] Our patient received conservative management initially and later underwent surgical management. Various coronary artery system repair surgeries can be performed for ALCAPA. These include coronary button transfer, the Takeuchi procedure and placement of a coronary artery bypass graft.[2,9,10] Of these options, coronary button transfer is considered to be the most "anatomically correct" procedure and is the preferred method in infants.[2,10] The modified Takeuchi is preferred if the coronary anatomy is

unfavourable for button transfer, as was in our case.[2,10]

The most important purpose of clinical evaluation and diagnostic tests in children with suspected dilated cardiomyopathy (DCM) is to identify correctable conditions. ALCAPA syndrome is a rare, often fatal but a treatable cause of dilated cardiomyopathy. General physicians and pediatricians need to keep a high index of suspicion of ALCAPA in infants who present with symptoms of congestive cardiac failure, failure to thrive and dilated cardiomyopathy.

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