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Case Report

A rare cause of dilated cardiomyopathy, the left coronary artery from the pulmonary artery (ALCAPA): Accurate diagnosis with dual-source CT

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ABSTRACT

An anomalous origin of the left coronary artery (LCA) from the pulmonary artery (ALCAPA syndrome) or Bland–White–Garland syndrome is a rare congenital cardiac anomaly. This syndrome is a rare cause of dilated cardiomyopathy. We report an 8-year-old male patient who presented with intermittent atypical substernal discomfort and mild exertional dyspnea. Dual Source CT and Coronary angiography demonstrated the dilated left atrium and left ventricle with consistent dilated cardiomyopathy and the right coronary artery (RCA) arising from the aorta, rich collaterals from the RCA to LCA and the LCA originated from the pulmonary trunk.

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1. 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.[2] It represents one of the most common causes of myocardial ischemia and infarction in children and if left untreated, results in a mortality rate of up to 90% within the first year of life.[3] ALCAPA is an important and treatable cause of dilated cardiomyopathy in infants. We report a typical case of ALCAPA syndrome in an 8-year-old male, who was diagnosed by Dual Source computed tomography.

2. Case report

An 8-year-old male presented with intermittent atypical substernal discomfort and mild exertional dyspnea. Upon auscultation, a grade 2 systolic murmur was heard over the right inferior sternal border. Chest radiography showed cardiomegaly and slightly increased pulmonary vascularity. The electrocardiogram was normal. Echocardiography showed dilatation of the left ventricle and left atrium and mitral regurgitation with an ejection fraction of 61%.

The patient was pretreated with atenolol (0.8 mg/kg) 2 h before the procedure. ECG-gated MDCT (multidetector computed tomography) (Somatom Definition, 128-slice Dual

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Source CT, Siemens Medical Systems, Erlangen, Germany) was carried out. Contrast material of 40 mm^3 (Ultravist 370 mg/mL, Schering, Germany) was given through the antecubital vein at a rate of 5 mm/s. Afterward, 25-cc of intravenous saline solution was given as a bolus at a rate of 5 cc/s. The ascending aorta was used as a reference point for location of the region of interest 1 cm distal to the tracheal bifurcation. Two seconds after the threshold level reached 140 Hounsfield units (HU), the scan was automatically started. Bolus tracking was employed for the control of contrast administration. The following protocol was used as a guidance for ECG-gated MDCT: detector collimation, 32 × 0.6 mm; gantry rotation time, 330 ms (temporal resolution, 83 ms); slice acquisition, 64 × 0.6 mm; pitch, 0.26 adapted to the heart rate; tube voltage, 100 kV; tube current, 320 mAs per rotation. The estimated effective radiation dose to the patient was 1.8 mSv.

Multiplanar view clearly demonstrated the RCA (right coronary artery) with its origin at the aorta and an anomalous LCA (left coronary artery) originating from the pulmonary trunk (Fig. 1a, b). The three dimensional reconstruction of the CT coronary angiography provided the anatomic features of the anomaly (Fig. 2a, b). The subsequent coronary angiography showed an absence of the LCA arising from the aorta. Selective right coronary angiography demonstrated the RCA originating from the right aortic sinus, with profuse collateral channels feeding the left coronary system (Fig. 3a). The left coronary artery drained into the main pulmonary artery. Selective catheterization of the pulmonary artery showed the origin of the anomalous LCA from the pulmonary artery but there was no antegrade flow through the anomalous LCA (Fig. 3b). An operation was planned; however, the patient's family did not want the surgery.

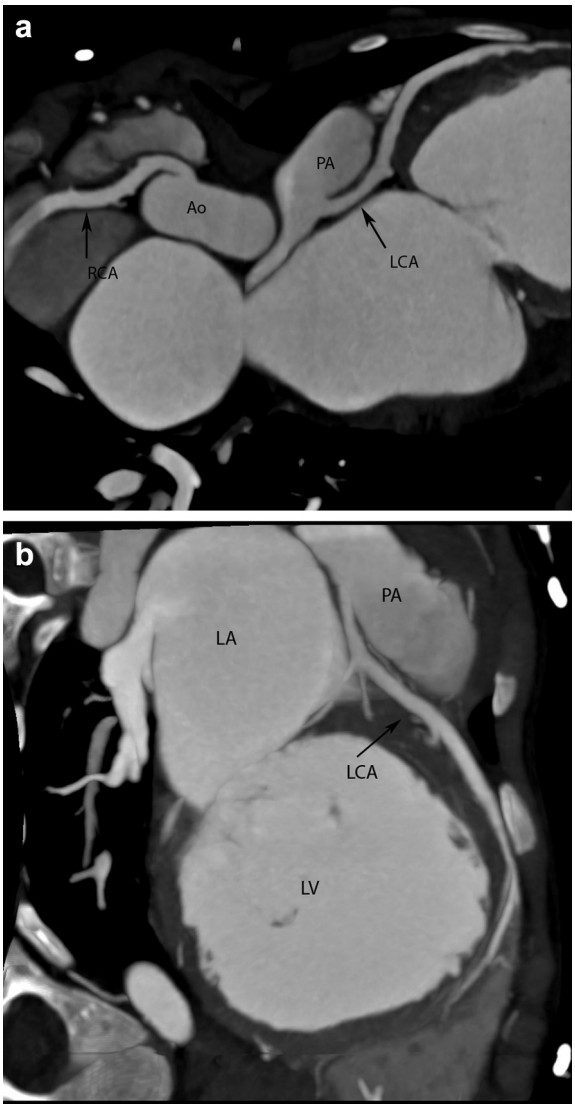

Fig. 1 – a. Axial and sagittal multidetector CT angiogram maximum intensity projection (MIP) image shows the origin of the left coronary artery from the main pulmonary artery. b. The dilated left atrium and left ventricle with consistent dilated cardiomyopathy. LCA; Left coronary artery, RCA; Right coronary artery, Ao; Aorta, PA; Pulmonary artery, LA; Left atrium, LV; Left ventricle.

3. Discussion

ALCAPA is a heart defect in which the left coronary artery is connected to the pulmonary artery instead of to the aorta. Collateral circulation between the right and left coronary systems ensues. Left coronary artery flow reverses and enters the pulmonic trunk due to the low pulmonary vascular resistance (coronary steal phenomena). As a result, the left ventricular myocardium remains underperfused. Consequently, the combination of left ventricular dysfunction and significant mitral valve insufficiency leads to congestive heart failure symptoms in the young infant.[4] Most of the patients with ALCAPA presented with signs and symptoms of congestive heart failure, dilated cardiomyopathy and a non-specific systolic cardiac murmur on physical examination. Most patients with ALCAPA syndrome die from severe congestive heart failure due to the chronic mitral regurgitation and global ischemic cardiomyopathy. However, a few patients can survive to adulthood if the collateral circulation is adequate.[5] It represents one of the most common causes of myocardial ischemia and infarction in children and if left untreated, results in a mortality rate of up to 90% within the first year of life.[3] Immediate surgical correction on diagnosis with the aim of restoring a two-coronary system circulation is the current standard in patients with ALCAPA.[6]

Surgery is indicated for the definite treatment of an anomalous origin of LCA from the pulmonary trunk. An effective treatment is either a reimplantation of the LCA in the aorta or an internal mammary artery-left anterior descending artery bypass.[7] Our patient's parents refused the operation.

Cardiac echocardiography is the first-line imaging modality used in diagnosis of ALCAPA. Although this modality is cheap and practical, it is user-dependent and not all cases may be correctly diagnosed.[8] The diagnosis should be confirmed by coronary angiography. Conventional coronary angiography is the modality of choice to diagnose coronary anomalies. However, it is an invasive method that requires hospitalization and at times causes some complications.

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