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Adult ALCAPA Syndrome-Anomalous Origin of Left Coronary Artery from Pulmonary Artery: A Rare Coronary Artery Anomaly Diagnosed by 256-Slice Computed Tomography

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Abstract

We are presenting a rare case of anomalous origin of left coronary artery from the pulmonary artery (ALCAPA) in an adult patient. The patient underwent coronary CT angiography for exertional dyspnoea, suspected as coronary artery disease; however there were no atherosclerotic plaques. There was anomalous origin of left coronary artery from pulmonary artery. Conventional catheter angiography confirmed the origin of left coronary artery from pulmonary artery, which was retrogradely filling through right coronary artery.

Keywords

ALCAPA

Subject Areas: **Radiology & Medical Imaging**

1. Introduction

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA), otherwise known as Bland-White-Garland syndrome, is a rare congenital heart abnormality that occurs in approximately 1/300,000 live births or 0.5% of children with congenital heart diseases [1]. It most often presents as an isolated defect, but in 5% of cases it may be associated with other cardiac anomalies, including atrial septal defect, ventricular septal defect, and aortic coarctation.

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There are two types of ALCAPA syndromes: the infantile type and the adult type, each of which has different manifestations and outcomes; ALCAPA syndrome in adults may be an important cause of sudden cardiac death [2].

Conventionally, evaluation of coronary artery anomalies is performed by using catheter-based angiography. However, the precise course of the vessel may not be adequately defined with this technique. The development of multi-detector row computed tomography (MDCT) has permitted better definition of the coronary vessels with CT and its role in the identification of such coronary artery anomalies is well established [3].

2. Case Presentation

A 45-year-old woman came with history of exertional breathlessness.

Chest X-ray: revealed no significant abnormality (**Figure 1**).

Coronary CT angiography scan was performed using a retrospective ECG gated scan protocol on a 256-slice Brilliance i CT scanner (Philips Best, Netherlands Healthcare) with a gantry rotation time of 0.27 s, detector collimation of 2 × 128 × 0.625 mm, with 256 overlapping slices of 0.625 mm thickness and dynamic z-focal spot.

60 mL of non-ionic iodinated contrast agent was injected at the rate of 4.5 ml/sec through antecubital vein.

Coronary CT angiography showed normal origin of right coronary artery (RCA), which was dilated and tortuous in course, measuring approximately 13 mm in diameter. Left coronary artery (LCA) was seen arising from main pulmonary artery and was dividing into left anterior descending artery (LAD) and left circumflex artery (LCX), which were dilated & tortuous in course and there were multiple collaterals over anterolateral aspect of left ventricular wall.

LCA was retrogradely filling from RCA through multiple collaterals and was draining into main pulmonary artery, which was accurately detected on MDCT coronary angiography as positive contrast jet from LCA into pulmonary artery (curved arrow in **Figure 2(a)**). There were no atherosclerotic plaques in coronary arteries.

Above findings were confirmed on conventional coronary angiography. On aortogram only right coronary artery was opacified, with retrogradely filling of LAD & LCX from RCA through collaterals and which were seen draining into main pulmonary artery (**Figure 3**).

3. Discussion

Adult living with anomalous origin of left coronary artery from main pulmonary artery is extremely rare [2].

Embryologically, the anomaly arises from either abnormal septation of the aorta and the pulmonary artery or from persistence of aortic buds that form the coronary arteries [4].

ALCAPA syndrome results in the “coronary steal” phenomenon, in which a left-to-right shunt leads to abnormal left ventricular perfusion. Patients may be asymptomatic, or they may present with mitral insufficiency, ischemic cardiomyopathy or malignant dysrhythmias, which may lead to sudden death [2].

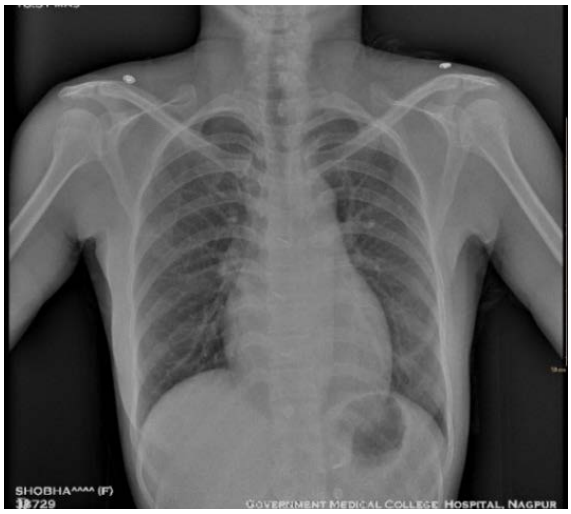

Figure 1. Chest X-ray PA view.

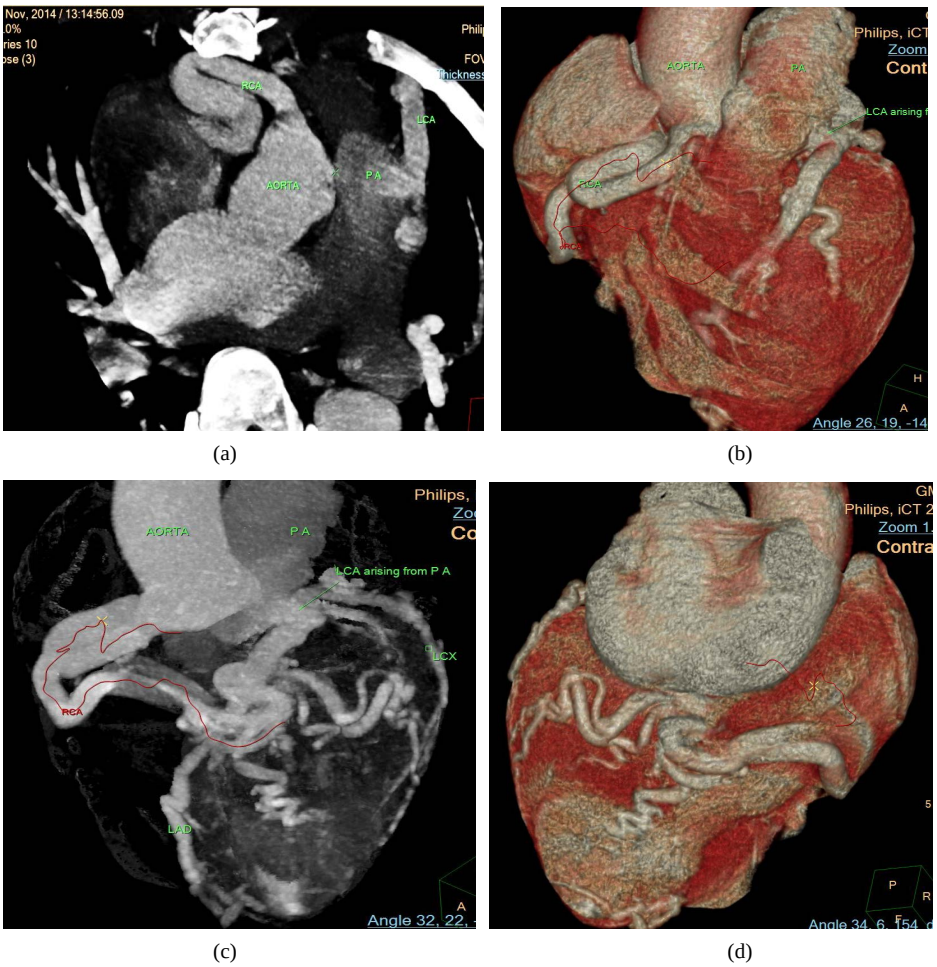

Figure 2. (a) MIP image at level of aorticsinus; (b) Three dimensional VRT image of heart & coronary arteries; (c) MIP image of coronary tree; (d) Three dimensional VRT image of heart & coronary arteries.

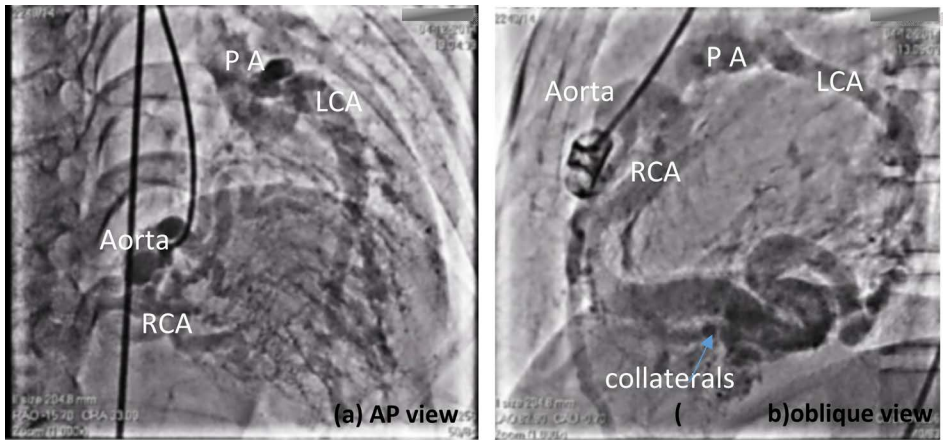

Figure 3. Conventional RCA angiogram reveals dilated & tortuous RCA, LCA is retrogradely filling through collaterals from RCA and draining into pulmonary artery (PA).

Factors that may lead to survival beyond infancy include the development of abundant intercoronary collateral arteries, an alteration in hemodynamics that encourages antegrade blood flow into the left coronary arterial

tree, and a reduction in the area of the left ventricular myocardium supplied by the LCA.

On CT and MRI, described imaging findings of ALCAPA, includes direct visualization of the LCA arising from the main pulmonary artery which is the diagnostic hallmark of ALCAPA syndrome, other findings include dilatation of the RCA and visualization of dilated intercoronary collateral arteries along the external surface of the heart or within the interventricular septum. Other differential diagnostic considerations for dilatation of the coronary arteries include Kawasaki's disease, coronary arterial fistulas, atherosclerosis, vasculitis (polyarteritis nodosa or Takayasu's arteritis), scleroderma, Ehlers-Danlos syndrome, hereditary haemorrhagictelangiectasia, trauma, and hyperlipidemia [1].

In the present case, direct visualisation of anomalous origin of LCA from the main pulmonary artery, along with dilatation and tortuosity of the RCA and external intercoronary collateral arteries, were well shown on CCTA and were diagnostic for ALCAPA syndrome.

256 slice MDCT is a noninvasive imaging technique which is fast and offers excellent spatial resolution, which is required to assess small vessels such as the coronary arteries. The short investigation time, simple preparation and minimal after care make MDCT coronary angiography advantageous over conventional coronary angiography.

As CCTA unambiguously delineates both origin and course of coronary vessels in patients with rare coronary anomalies like ALCAPA, CCTA should be considered as first line of investigation in suspected cases of coronary artery anomalies. The main disadvantages of MDCT angiography are its relatively high radiation dose and its inability to assess flow.

4. Conclusion

ALCAPA syndrome is a rare but potentially lethal coronary artery anomaly and one should be aware of this condition. The diagnosis can be accurately made with non-invasive 256 slice CCTA. Prompt treatment may prevent life threatening complications, such as myocardial infarction or sudden death.

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