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Case Report
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ALCAPA Syndrome and Atrial Septal Defect In a 68-Year-Old Woman: An Extremely Rare Congenital Association

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Anomalous left coronary artery arising from the pulmonary artery (ALCAPA) has been generally reported as an isolated lesion that is also called Bland-White-Garland syndrome. Herein we report a case of ALCAPA syndrome with an atrial septal defect in a 68-year-old woman. She had been asymptomatic until the age of 68. Echocardiographic examination revealed atrial septal defect, and coronary angiography showed that the left main coronary artery originated from the pulmonary artery and intensive collateral connections between the right and left coronary artery. In this case, it would appear that ALCAPA is associated with atrial septal defect.

Key Words: ALCAPA • Atrial septal defect • Congenital cardiac defect • Coronary anomaly

INTRODUCTION

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA or Bland-White-Garland Syndrome) is a rare coronary anomaly found in approximately one in 300,000 live births.[1] Reversing blood flow of the left coronary artery after the birth, perfusion of the left myocardium depends on collateral connections with the right coronary artery.[1] Patients rarely survive to adulthood, and primarily depends on rapidly growing collateral circulation.[1] ALCAPA might be associated with other congenital heart defects such as coarctation of the aorta, tetralogy of Fallot or pulmonary atresia.[2] To the best of our knowledge, this is the second case report that shows the association between ALCAPA and atrial septal defect.

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CASE REPORT

A 68-year-old female patient presented with a 15-day history of shortness of breath and exertional angina, with no relevant medical history except hypertension. Her physical examination revealed a grade II/VI systolic murmur over the second intercostal space at the left sternal border, and the second heart sound was fixed splitting with increased intensity. Electrocardiogram showed normal sinus rhythm with non-specific ST and T wave changes. The patient's echocardiographic imaging revealed dilated right-sided heart chambers and an atrial septal defect, as well as normal left ventricular ejection fraction (60%), moderate tricuspid insufficiency and elevated systolic pulmonary artery pressure (80 mmHg). Transesophageal echocardiography confirmed the diagnosis of ostium secundum-type atrial septal defect (Figure 1). The patient underwent coronary angiography (CAG) and right heart catheterization that showed mean pulmonary artery pressure of 52 mmHg. CAG revealed rudimentary left coronary system and extremely tortuous, dilated right coronary artery (RCA). Furthermore, the distal part of the RCA extended collaterals to the left anterior descending artery, and some contrast drained into the main pulmonary artery (Figure 2A).

Multislice computed tomography with three dimensional reconstruction showed that the main coronary artery originated from the left pulmonary artery, confirming the diagnosis of ALCAPA (Figure 2B). The patient refused further intervention after complete study.

DISCUSSION

ALCAPA was first reported in 1866, but an exact description of this condition was defined by Bland, White and Garland in 1933.[3] ALCAPA is a life-threatening congenital coronary anomaly.[3] Most patients become symptomatic after birth when the pulmonary artery resistance has resolved and followed by the closure of ductus arteriosus; however, some cases could survive until adulthood, similar to our patient. The collateral circulation is important for supplying blood to the left coronary territory.[4] ALCAPA patients who survive until adulthood have various symptoms of myocardial ischemia, impaired left ventricular function, and mitral regurgitation and heart failure, depending on the collateral circulation between the two coronary systems.[4]

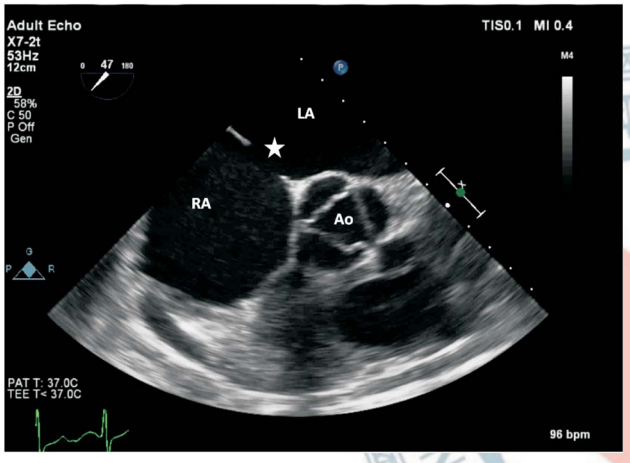

Figure 1. *Transesophageal echocardiography is showing atrial septal defect. Ao, Aorta 116 × 84 mm; LA, left atrium; RA, right atrium.*

ALCAPA patient may present with angina, exertional dyspnea, sudden cardiac death or may be diagnosed incidentally at upon coronary angiography.[4] Echocardiography may reveal that abnormal vessel originated from pulmonary artery or large right coronary artery.[1] Conventional coronary angiography is the main imaging modality, however, computed tomography or magnetic resonance imaging can also be used when concomitant congenital cardiac defect was suspected. CAG is not mandatory in those patients with atrial septal defect, especially in the young patients. In this case, the patient was older and had stable angina pectoris.

Laux et al. reported 12 cases with ALCAPA syndrome, where coarctation of the aorta was the most frequently associated heart defect in their series.[2] The main treatment modality for ALCAPA patients is open heart surgery to prevent sudden cardiac death; however, long term survival in patients with ALCAPA has been documented with previously reported case studies.[1,4,5] The oldest reported case with ALCAPA syndrome

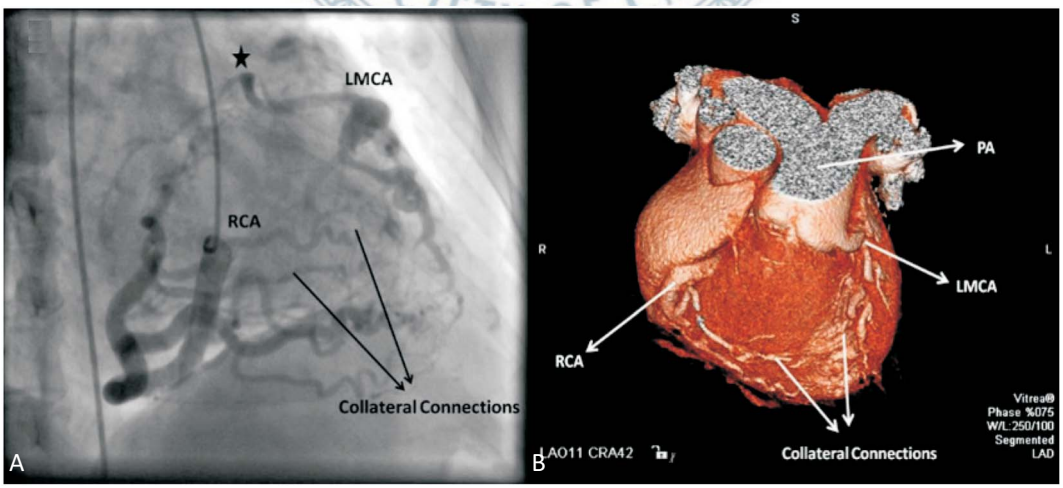

Figure 2. *(A) Right anterior oblique view of coronary angiogram is revealing dilated and tortuos right coronary artery and collateral connection between the coronary system. (B) Three dimensional reconstruction of multislice computed tomography is showing that left main coronary artery is originated from the pulmonary artery (star). LMCA, left main coronary artery; PA, pulmonary artery; RCA, right coronary artery.*

was an 88 year-old woman.[5] The present patient is an extremely unusual case in that she was asymptomatic until the age of 68, and had secundum-type atrial septal defect.

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