

Usefulness of Multidetector-row Computed Tomography in Diagnosis of Anomalous Origin of Left Coronary Artery Arising From the Pulmonary Artery

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Anomalous origin of the left coronary artery arising from the pulmonary artery (ALCAPA) is a rare congenital heart defect that affects approximately 1 in 300,000 live births and accounts for 0.5% of all congenital heart disease. Without surgical intervention, most patients with this anomaly die in infancy. The diagnosis of ALCAPA syndrome is sometimes difficult, especially in children, because it often presents with atypical symptoms and signs. Echocardiography can yield some findings that are highly indicative of this syndrome. Cardiac catheterization usually establishes the diagnosis, but it is invasive and painful. Multidetector-row computed tomography (MDCT) is a valuable alternative tool to confirm the diagnosis of ALCAPA syndrome. In this report, we describe a 24-year-old woman with a dilated right coronary artery detected by transthoracic echocardiography, showing an interventricular abnormal flow over the diastolic phase of the cardiac cycle and the left main coronary artery not merging with the ascending aorta. The diagnosis of ALCAPA syndrome was confirmed by MDCT. [*J Chin Med Assoc* 2010;73(9):492–495]

Key Words: ALCAPA syndrome, echocardiography, multidetector-row computed tomography

Introduction

Anomalous origin of the left coronary artery arising from the pulmonary artery (ALCAPA) is a rare congenital heart defect that affects approximately 1 in 300,000 live births and accounts for 0.5% of all congenital heart disease. Without surgical intervention, most patients with this anomaly die in infancy. The diagnosis of ALCAPA is usually established by cardiac catheterization. With improvements in tomographic imaging and techniques, multidetector-row computed tomography (MDCT) now plays an increasingly important role in the diagnosis of many types of congenital heart disease, thus reducing the need for diagnostic cardiac catheterization.

In this report, we describe the case of a 24-year-old woman who presented with dyspnea on exertion. ALCAPA syndrome was diagnosed by MDCT, which

clearly showed the left coronary artery originating from the pulmonary trunk and well-developed collaterals between the right coronary artery and left anterior descending artery.

Case Report

A 24-year-old woman presented to our cardiovascular outpatient department because she had been experiencing dyspnea on exertion for 6 months. Physical examination revealed no specific findings with regard to her cardiovascular system. Electrocardiography showed ST segment depression in leads V4–6, and inverted T waves in lead I and AVL (Figure 1). Chest X-ray revealed cardiomegaly (cardiothoracic ratio, 52%) (Figure 2). Transthoracic echocardiography and 2-dimensional and color Doppler imaging demonstrated a dilated right



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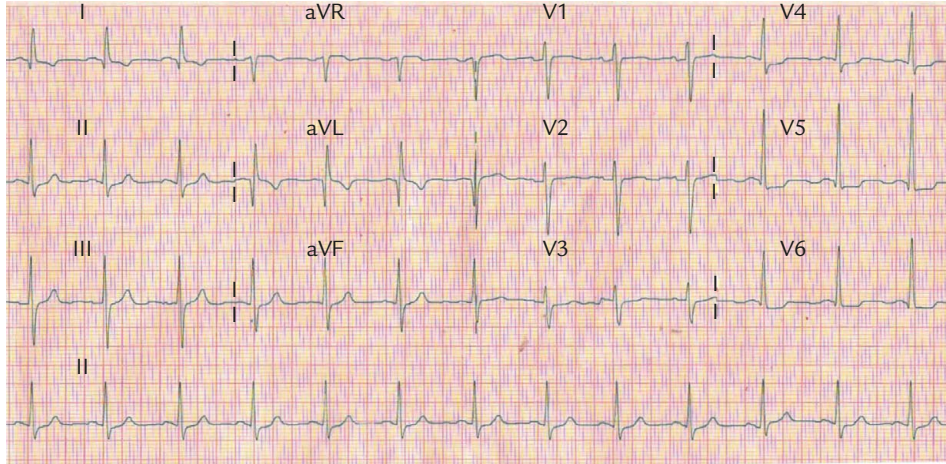


Figure 1. Complete 12-lead electrocardiogram shows ST segment depression over V4–6, which may represent lateral wall myocardial ischemia.



Figure 2. Chest X-ray shows cardiomegaly (cardiothoracic ratio, 52%).

coronary artery (1.0 cm in diameter), normal-sized left ventricle, moderate mitral regurgitation and mild hypokinesis of the anterolateral and lateral walls of the left ventricle. The Doppler color flow image showed right coronary artery flow in the diastolic phase, with the flow going from the inferior to the superior portion of the interventricular septum, then going into the pulmonary artery trunk (Figure 3). MDCT (Brilliance 40; Philips Medical Systems, Koninklijke Philips Electronics N.V., Best, The Netherlands) with contrast medium injection showed a dilated right coronary artery arising from the aorta, anomalous origin of the left coronary artery from the pulmonary trunk, and multiple well-developed collateral vessels between the right coronary artery and left anterior descending artery without atherosclerotic change (Figure 4). Coronary angiography was not performed. The patient was referred

to the cardiovascular surgeon, and she received reimplantation of the left coronary artery to the ascending aorta, angioplasty of the main pulmonary artery with autologous pericardium and mitral valve annuloplasty to recreate a 2-vessel coronary system. Her postoperative course was uneventful and she was discharged 1 week later.

Discussion

ALCAPA is the current term for what was previously known as Bland-White-Garland syndrome, named after the 3 physicians, Edward Bland, Paul Dudley White and Joseph Garland, who published the first comprehensive clinical presentation of the syndrome in 1933.¹ Most patients with this anomaly die in infancy.² Only 5–10% of patients survive an early myocardial infarction, with subsequent development of coronary collateral vessels from the right coronary artery to the left coronary artery, but most of them develop myocardial ischemia, mitral regurgitation, impaired left ventricular dysfunction and progressive heart failure, with sudden death occurring in some cases, depending on the development of collateral circulation.² Early diagnosis and treatment may prevent irreversible damage to the myocardium and subsequent complication.

ALCAPA syndrome develops before birth when the systemic and pulmonary arterial pressures are equal and there is anterograde flow in both the left and right coronary arteries. In the neonatal period, this gradually changes as the pulmonary blood pressure diminishes, the ductus arteriosus closes, and the flow in the left coronary artery reverses.³ The development

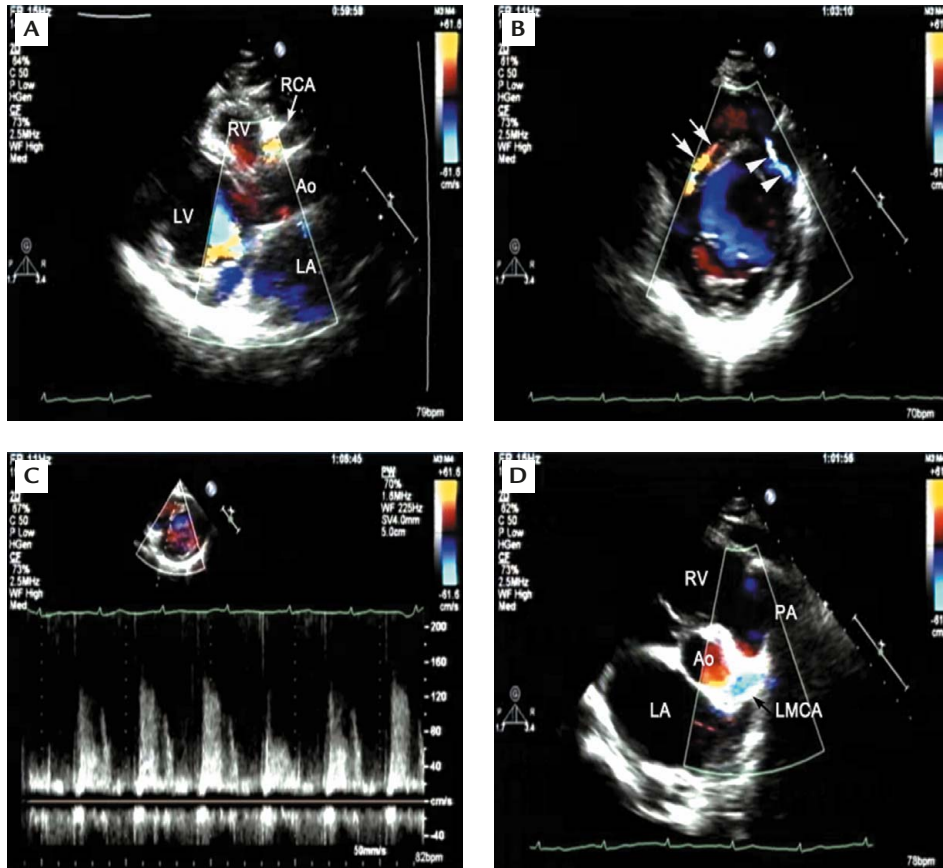


Figure 3. Transthoracic echocardiography. (A) Parasternal long-axis view, color Doppler image, shows a dilated right coronary artery (RCA, arrow) which is 1.0 cm in diameter arising from the right aortic sinus. (B) Parasternal short-axis view shows interventricular abnormal flow originating from the inferior septum (arrows) and continuing to the superior septum (white arrowheads). (C) Doppler flow image, pulse-wave mode, on the abnormal flow over the interventricular septum, shows dynamic blood flow over the diastolic phase of the cardiac cycle. (D) Parasternal short-axis view shows a dilated left main coronary artery (LMCA) with blood flow that does not merge with the ascending aorta (Ao, arrow). RV = right ventricle; LV = left ventricle; LA = left atrium; PA = pulmonary artery.

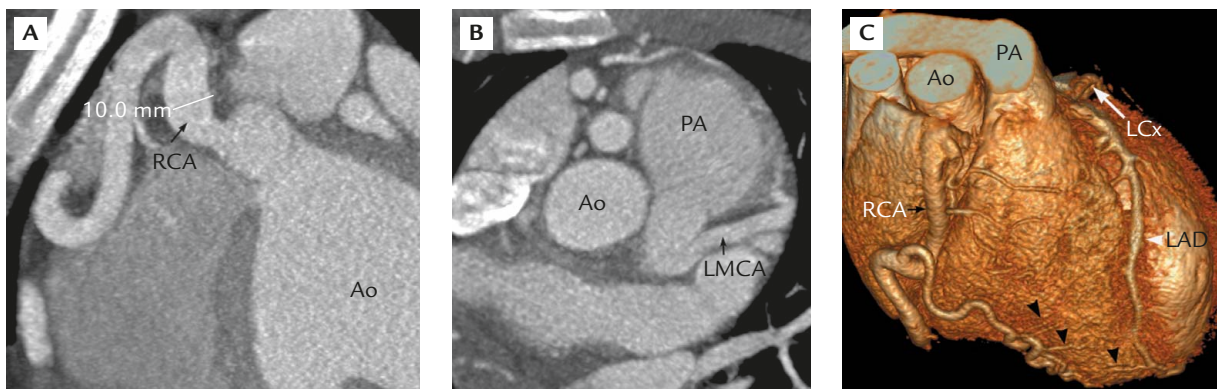


Figure 4. Multidetector-row computed tomography (CT). (A) Oblique coronary image of coronary CT angiography shows a dilated right coronary artery (RCA, arrow) which is 1.0 cm in diameter, arising from the right aorta (Ao) sinus. (B) Axial image of coronary CT angiography clearly shows a dilated left main coronary artery (LMCA, arrow) originating from the pulmonary artery (PA). (C) The 3-dimensional volume-rendered image reveals multiple well-developed collateral arteries (black arrowheads) between the RCA (black arrow) and the left anterior descending coronary artery (LAD, white arrowhead); the LMCA merges with the pulmonary trunk. LCx = left circumflex artery.

of collateral vessels between the right and left coronary arteries during the closure of the duct and lowering of pulmonary pressure will determine the extent of myocardial ischemia.⁴

Often, ALCAPA syndrome is suspected by clinicians when the findings of transthoracic echocardiography reveal a dilated right coronary artery arising from the aorta, diastolic blood flow from the inferior portion to the superior portion of the interventricular septum, visualization of diastolic blood flow from the left coronary artery into the pulmonary artery, and mitral regurgitation.⁵ However, these findings also demonstrate the possibility of Kawasaki's disease or arteriovenous fistula because they share similar echocardiographic findings such as a dilated right coronary artery, intramyocardial dilated coronary artery or shunting, which may confuse echocardiologists. The diagnosis of ALCAPA syndrome is usually established by cardiac catheterization with injection at the aortic root, which can reveal a dilated and tortuous right coronary artery with collateral filling of the left coronary artery system; variable degrees of shunting to the pulmonary artery may also be present.⁶

In the last decade, with the evolution in tomographic imaging and techniques, MDCT has been shown to be a promising noninvasive imaging modality for the demonstration of anatomy and evaluation of the coronary artery lumen with good image quality and high diagnostic accuracy.⁷⁻⁹ The potential of MDCT is enhanced by its inherent ability to acquire 3-dimensional volume data and reconstruction technique. For evaluation of the cardiac chamber, coronary artery and great vessel connection problems in complex congenital heart disease, MDCT is considered to be a very useful modality.^{10,11} MDCT serves as a rapid and convenient tool for the evaluation of ischemic heart disease, especially in the aspects of myocardium thickness, motion, viability, and perfusion.^{8,9,12} From the viewpoint of a cardiovascular surgeon, in the preoperative planning stage for a patient with complex congenital heart disease with or without coronary atherosclerotic disease, it is extremely important to fully understand where the defect is or what the abnormal vessel connection problem is. MDCT can serve as a one-stop shop to provide the cardiovascular surgeon with all this information. Moreover, the risk of vascular complications with coronary angiography is higher than the risk with CT angiography. Therefore, in adult patients with ALCAPA syndrome, MDCT is a valuable diagnostic tool and can provide the cardiovascular surgeon with information on the connection of the coronary

artery and pulmonary artery, assisting in preoperative planning.

We have described the case of a 24-year-old woman with a dilated right coronary artery who was diagnosed with ALCAPA syndrome by MDCT. She underwent an operation to create a 2-vessel coronary system.¹³ Her postoperative course was uneventful, and she was discharged 1 week later. MDCT is a useful alternative tool that can be used to establish the diagnosis of ALCAPA syndrome.

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