

Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery in an Elderly Patient Visualized by Three-Dimensional Multidetector Computed Tomograph Coronary Angiography

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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. We report a 65-year old female patient who presented with atypical chest discomfort. Coronary angiography and three-dimensional multidetector computed tomography coronary angiography demonstrated the ectatic right coronary artery (RCA) arising from the aorta, rich collaterals from the RCA to LCA and the ectatic tortuous LCA that originated from the pulmonary trunk. (Korean Circulation J 2005;35:84-87)

KEY WORDS : Anomalous origin of left coronary artery from artery ; Coronary vessel anomalies ; Collateral circulation.

Introduction

An anomalous origin of the left coronary artery (LCA) from the pulmonary artery, which is known as ALCAPA syndrome or Bland-White-Garland syndrome, is a rare congenital anomaly that accounts for approximately 0.5% of congenital malformations.^{1,2)} There is an almost 85% mortality during infancy as a result of myocardial ischemia. However, some patients can survive with this anomaly to adulthood if they have adequate collateral circulation.^{2,3)} We report a typical case of ALCAPA syndrome in a 65-year old woman, who was diagnosed by conventional coronary angiography including selective right coronary angiography and selective pulmonary angiography. This rare anomaly was visualized by multidetector computed tomography (MDCT).

Case

A 65-year old female presented with an intermittent aty-

pical substernal discomfort and mild exertional dyspnea for one year. She was generally healthy except for a one year history of diabetes mellitus and a several-year history of degenerative arthritis in both knee joints. She denied any specific acute febrile illness including Kawasaki disease during childhood. The physical examination, chest radiography, and electrocardiogram were normal. An exercise treadmill test (Bruce protocol) and stress perfusion myocardial imaging with 99m Tc-sestamibi at both rest and during physical stress were performed to disclose any inducible myocardial ischemia. However, there was no evidence of myocardial ischemia.

A transthoracic echocardiogram showed a dilated right coronary artery (RCA) arising from the right sinus of Valsalva (Fig. 1). Left ventricular function was preserved without regional wall motion abnormalities. The subsequent coronary angiography showed an absence of the LCA arising from the aorta. Selective right coronary angiography demonstrated an enormously dilated, tortuous and dominant RCA from the right aortic sinus, with profuse collateral channels feeding the left coronary system. The left coronary artery drained into the main pulmonary artery (Fig. 2). Selective catheterization of the pulmonary artery showed the origin of the anomalous dilated LCA from the pulmonary artery but there was no antegrade flow through the anomalous LCA.

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MDCT coronary angiography was performed by using a 16-channel multi-detector row CT scanner (Somatom Sensation 16; Siemens, Forchheim, Germany) with retrospective

ECG gating. The scanning parameters were 210-msec temporal resolution and 16×0.75 mm detector collimation, and the axial images were reconstructed with a 1 mm slice

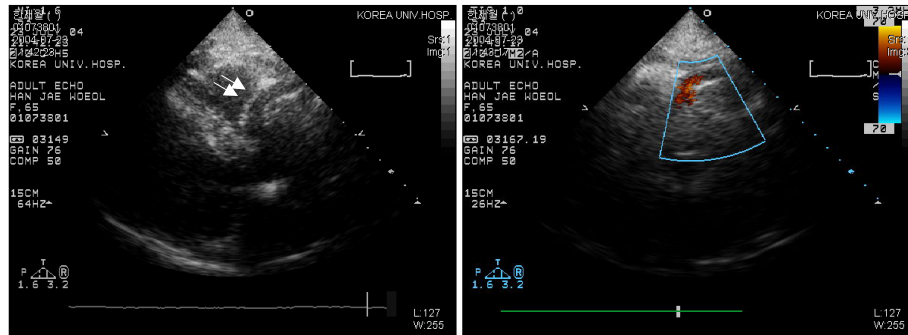


Fig. 1. Modified parasternal long axis view demonstrating a dilated right coronary artery (arrows) from the right coronary cusp of the aorta.

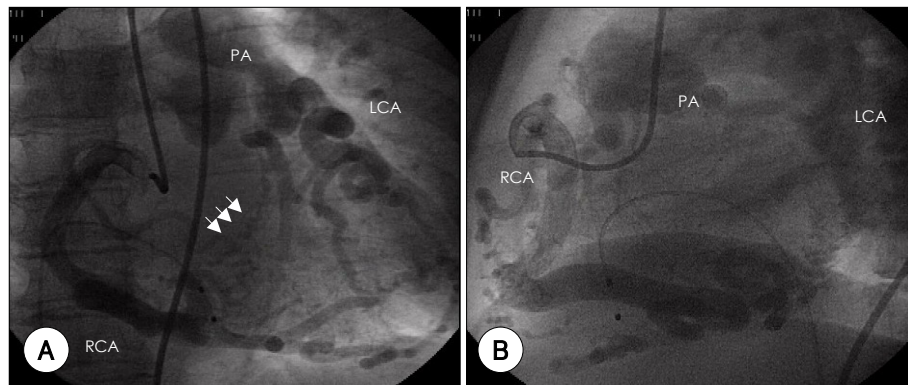


Fig. 2. Right coronary angiography (A: right anterior oblique view, B: left lateral view) showing well developed rich collaterals (arrows) from the ectatic right coronary artery (RCA) to the left coronary artery (LCA) originating from of the main pulmonary artery (PA).

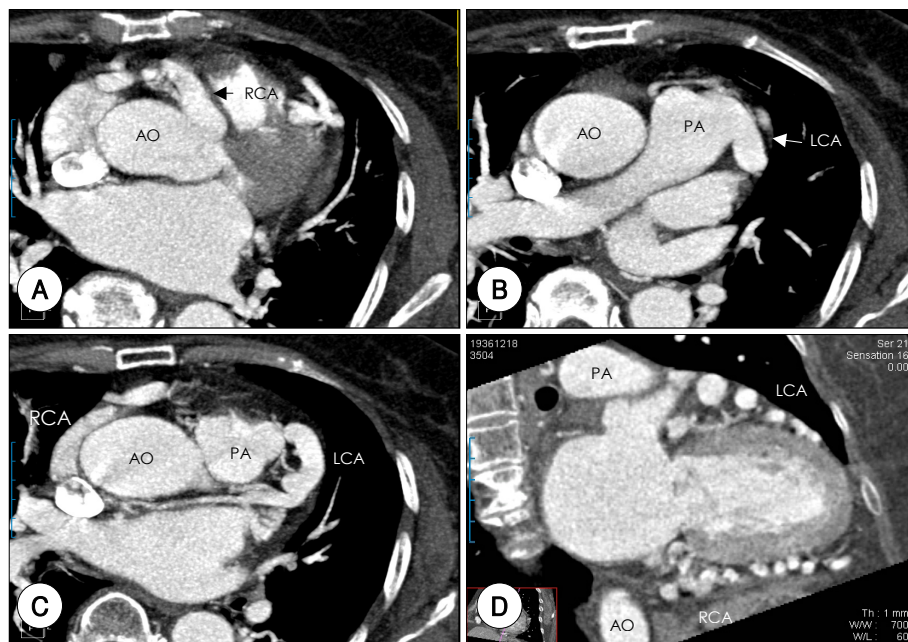


Fig. 3. Oblique axial view from the CT coronary angiography clearly showing the ectatic RCA with its origin at aorta (AO, A) and anomalous origin of LCA from PA (B). Markedly dilated both RCA and LCA (C) and tortuosity (D) were clearly demonstrated. RCA: right coronary artery, LCA: left coronary, PA: pulmonary artery, AO: aorta.

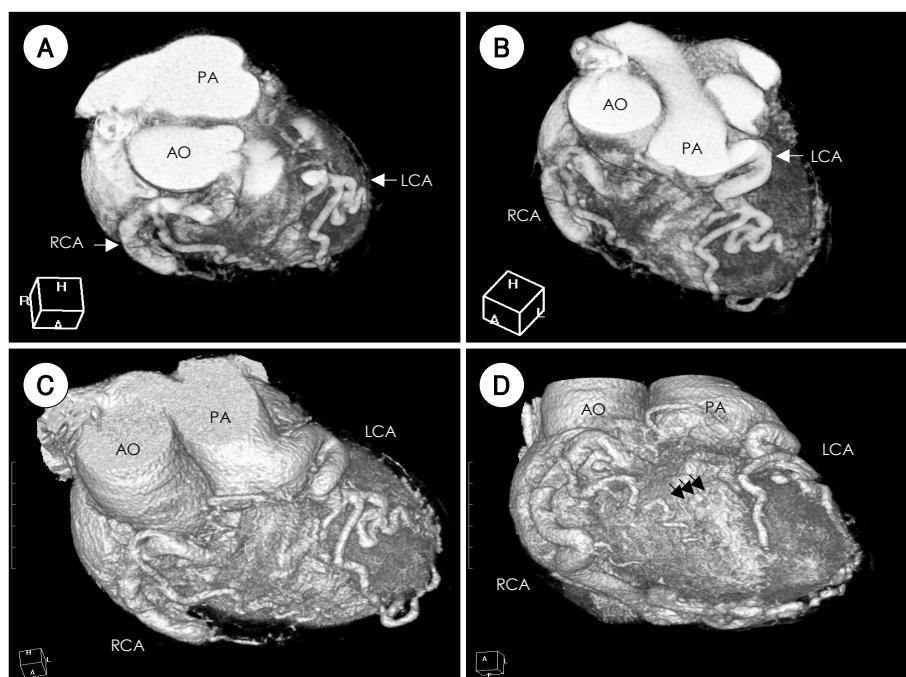


Fig. 4. Three-dimensional volumerendered view from the CT coronary angiography enabled the visualization of the dilated RCA from the aorta (A), anomalous origin of the LCA from PA (B), along with their anatomical relationship with surrounding structures (C). Rich collateral channels between RCA and LCA are shown in D (arrows). AO: aorta.

thickness and 0.7 mm increment. The reconstruction window, which was optimal for the anomalous coronary artery, was chosen at 30% of the R-R interval. A 110-mL dose of the non-ionic iodinated contrast material (Ultravist; Schering, Berlin, Germany) was infused intravenously at 4 mL/sec. A 30-mL bolus of normal saline was given after administering the contrast material to decrease the number of artifacts from the contrast material in the right heart.

Oblique axial view clearly demonstrated the RCA with its origin at the aorta and an anomalous LCA originating from the pulmonary trunk (Fig. 3). The three-dimensional reconstruction of the CT coronary angiography provided the anatomic features of the anomaly demonstrated in the coronary angiography (Fig. 4).

Discussion

Congenital coronary anomaly accounts for 0.2–1.3% of patients who undergo coronary angiography.⁴⁾ ALCAPA syndrome is one of the most frequent congenital coronary anomalies.⁵⁾ In Korea, Oh, et al reported a 45-year old man with ALCAPA syndrome who was treated successfully with a closure of the anomalous LCA orifice and saphenous vein grafting.⁶⁾ Sohn, et al reported a 32-year old woman with ALCAPA syndrome that was confirmed by coronary angio-

graphy who had reversible myocardial ischemia.⁷⁾ Our patient is the oldest typical case of adult ALCAPA syndrome reported in Korea. The patient had 'adult type' rich collaterals without ischemic symptoms and myocardial injury. Because of the absence of life-threatening symptoms or ischemic insults, the patient refused to have a definite surgical correction.

Most patients with ALCAP 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.^{2,3)} Regarding the collateral circulation, the 'infantile type' has no collaterals and no evidence of a left-to right flow whereas the 'adult type' has well established collaterals and a generous left to right flow.⁸⁾

ALCAPA syndrome is indicated by the patient's clinical history, electrocardiography, echocardiography, and treadmill test or stress myocardial perfusion imaging, but a definite diagnosis is made by conventional coronary angiography.⁹⁻¹¹⁾ Contrast-enhanced MDCT coronary angiography currently appears to fulfill the requirements of a noninvasive morphological assessment of the coronary arteries, as a result of its combination of unprecedented acquisition speed, spatial resolution, and robustness of use.¹²⁾ The three-dimensional volume rendered view derived from MDCT coronary angiography demonstrates the anomalous coronary arteries as well

as their relationship to the surrounding anatomy in this particular ALCAPA syndrome case.

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.¹³⁾

REFERENCES

- 1) Purut CM, Sabiston DC Jr. *Origin of the coronary artery from the pulmonary artery in older adults. J Thorac Cardiovasc Surg* 1991;102:566-70.
- 2) Saeed BT, Rosin MD, Murray RG. *Successful operation in an old survivor of anomalous origin of the left coronary artery from the pulmonary trunk (Bland-White-Garland Syndrome). Br Heart J* 1994;71:193-5.
- 3) Moodie DS, Fyfe D, Gill CC, et al. *Anomalous origin of the left coronary artery from the pulmonary artery (Bland-White-Garland syndrome) in adult patients: long term follow-up after surgery. Am Heart J* 1983;106:381-8.
- 4) Yamanaka O, Hobbs RE. *Coronary artery anomalies in 126,595 patients undergoing coronary angiography. Cathet Cardiovasc Diagn* 1990;21:28-40.
- 5) Keith JD. *The anomalous origin of the left coronary artery from the pulmonary artery. Br Heart J* 1959;21:149-61.
- 6) O SI, Lim HJ, Oh BH, et al. *A case of anomalous left coronary artery from pulmonary artery (Bland-White-Garland syndrome). Korean Circ J* 1993;18:468-73.
- 7) Shon TH, Moon KW, Yoo KD, et al. *Anomalous origin of left coronary artery from pulmonary artery: report of an adult case. Korean Circ J* 1999;29:528-31.
- 8) Selzman CH, Zimmerman MA, Capbell DN. *ALCAPA in an adult with preserved left ventricular function. J Card Surg* 2003;18:25-8.
- 9) Askenazi J, Nadas AS. *Anomalous left coronary artery originating from the pulmonary artery: report on 15 cases. Circulation* 1975;51:976-87.
- 10) Loya YS, Pinto RJ, Desai DM, Sundaram U, Bhagwat AR, Sharma S. *Selective coronary angiography via antegrade venous route in congenital cyanotic heart disease. Cathet Cardiovasc Diagn* 1993;28:179-82.
- 11) Elhendy A, Zoet-Nugteren S, Cornel JH, et al. *Functional assessment of ALCAPA syndrome by dobutamine stress thallium-201 SPECT and echocardiography. J Nucl Med* 1996;37:748-51.
- 12) Schoepf UJ, Becker CR, Ohnesorge BM, Yucel EK. *CT of coronary artery disease. Radiology* 2004;232:18-37.
- 13) Almeida de Oliveira S, Diamant J, Bezerra de Carvalho V, Arie S, Macruz R, Zerbini EJ. *Anomalous origin of the left coronary artery from the pulmonary artery: surgical repair of an unused form. J Cardiovasc Surg* 1977;18:599-605.