A Refined Coronary Computed Tomography Procedure Reveals an Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery, Including the Collateral Artery

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Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA), which has also been termed Bland-White-Garland syndrome, is a rare congenital anomaly of the coronary artery that accounts for 0.25–0.5% of all congenital cardiac malformations. Although computed tomographic coronary angiography allows for the evaluation of the connection and anatomy of ALCAPA, procedural details have not yet been reported. This case report describes how the appropriate dose and duration of contrast medium administration were determined, as well as the optimal direction of image acquisition to allow the most detailed anatomical visualization of the ALCAPA.

Key words ALCAPA · Bland-White-Garland syndrome · Computed tomography angiography.

INTRODUCTION

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital cardiac malformation in which the coronary artery arises from the pulmonary artery (PA) [1]. Although multi-detector computed tomographic coronary angiography (CTCA) is used to assess ALCAPA [2], detailed methods for the procedure have not yet been reported. The development of increasingly high-speed multi-slice X-ray computed tomography (CT) scanners has led to shorter imaging duration and the need to determine the best timing for imaging.

CASE REPORT

A 74-year-old female was admitted due to exertional dyspnea and underwent electrocardiography (ECG) and transthoracic echocardiography; the findings were suggestive of coronary artery disease with concomitant coronary fistula. Coronary angi-

ography (CAG) revealed a giant right coronary artery (RCA) channeling directly to the left coronary artery (LCA) without significant stenosis (Fig. 1, Movie 1 in the online-only Data Supplement) and coursing off the LCA into the PA without pulmonary hypertension (mean PA pressure: 15 mm Hg).

We performed an ECG-gated CTCA using a 256-slice multi-detector CT unit (Brilliance iCT, version 3.2.5, Philips, Best, the Netherlands) and made volume-rendering images on the portal workstation (IntelliSpace Portal, version 8, Best, the Netherlands). The baseline heart rate was 77 beats per minute (bpm), so intravenous Landiolol (Ono Pharmaceutical Co., Ltd., Osaka, Japan) was administered at a dosage of 0.125 mg/kg body weight into the antecubital vein; image acquisition was started with a heart rate of 63 bpm. In addition, 0.3 mg of sublingual nitroglycerin (Toa Eiyo Ltd., Tokyo, Japan) was given to the patient prior to imaging.

The standard protocol of contrast medium (iopamidol-370, Bayer Yakuhin Ltd., Osaka, Japan) administration for patients with a body weight of less than 50 kg calls for the injection of iodinated contrast at 20 mg/kg/second for 14 seconds followed by 30 mL of saline; the patient had a body weight of 44 kg.

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Fig. 1. CAG showing with anomalous origin of the LCA from the PA. (A) Schema of coronary flow based on CAG. (B-D) CAG (anterior-posterior cranial view) images showing an enlarged RCA and collateral circulation via the LCA into the PA from the RCA. RCA: right coronary artery, LAD: left anterior descending artery, LCX: left circumflex artery, CAG: coronary angiography, LCA: left coronary artery, PA: pulmonary artery.

However, the previously performed CAG imaging had already revealed abnormal dilation of the RCA. We therefore decided to use the protocol for patients weighing more than 50 kg (24 mg/kg/second) to enhance visualization of the entire coronary artery, including the RCA, collateral vessels, and LCA. The duration of the contrast injection was also prolonged because we anticipated a longer duration of blood flow through the diseased coronary artery.

The customary protocol calls for the initiation of data acquisition when the CT value of the region of interest (ROI) on the ascending aorta reaches 200 Hounsfield units and a direction of image acquisition from the side of the patient's head towards the cardiac apex to align with the direction of the coronary artery. However, since the LCA circulation in this case ran in the opposite direction (from the cardiac apex towards the head), CT images were obtained in the same direction as the flow.

It took about two seconds longer than usual to move the CT scan from the point set on the ROI to the cardiac apex and another two seconds more for the enhancement to reach the PA from the distal RCA via the collateral circulation. Based on the CAG, the duration of flow to the PA took 36 frames at a rate of 15 frames per second. Thus, we decided to combine the time required to move the CT and for the flow to reach the LCA and increased the contrast injection rate by four seconds. Detection

of the direction of coronary artery flow requires a higher density of contrast medium in the LCA than in the PA. Therefore, a prolonged contrast administration equalizes the density of the LCA and PA and prohibits detection of the direction of flow.

A total of 43 mL of non-ionic iodinated contrast was administered intravenously, followed by 30 mL of saline at a rate of 2.8 mL/s to obtain images with sufficient enhancement of both arteries that allowed acquisition of volume-rendering images and visualization of blood flow. These volume-rendering images identified the source of flow as originating from the RCA to the left anterior descending artery and to the main PA (Fig. 2), which was later confirmed by transesophageal echocardiography. It also revealed that the ostium of the LCA arose behind the main PA just above the pulmonary valve. Coronary flow was in the direction from the LCA to the PA.

DISCUSSION

In our protocol, we set the time duration of contrast medium administration to four seconds and the imaging direction from the side of the cardiac apex towards the patient's head. The four-second duration was based on performing a CTCA with chronic total occlusion (CTO) of the coronary artery, wherein the duration of contrast medium administration was two sec-

Fig. 2. (A) CECT showing a well-developed artery in the intraventricular septum. The vessel diameter at the apex was 6.6 mm. The arrowheads indicate a well-developed distal left anterior descending artery. (B) CECT showing a dilated LCA arising from the PA. The arrowheads indicate the LMT with a diameter of 12.7 mm. The lower CT value of the PA compared to the LMT suggested a retrograde direction of coronary blood flow into the PA from the LCA. (C and D) Three-dimensional computed tomographic coronary angiography showing developed collateral arteries. CECT: contrast-enhanced computed tomography, PA: pulmonary artery, LCA: left coronary artery, LMT: left main trunk.

onds longer than usual when imaging a typical coronary artery. The intuitive decision to prolong the contrast administration time for two more seconds compared to imaging a CTO was made in anticipation of the slower coronary flow through the anomaly. This imaging protocol was used by the authors in a previously encountered case of ALCAPA in whom CAG was not performed, and this method proved to be successful in detailing the coronary artery malformation. We compared this past injection protocol with our current patient using the timing information from the CAG and were able to validate the four-second duration of contrast administration and deem it as appropriate. It is therefore our impression that clear imaging of the coronary artery anatomy of an ALCAPA can be performed using this CTCA protocol.

This case presented CTCA images that clearly depicted ALCAPA in an elderly patient without pulmonary hypertension. Planning the dose and duration of administration of contrast medium and determining the appropriate direction for CTCA

acquisition are important to achieve superior visualization of the dilated and elongated arteries.

Supplementary Movie Legend

Movie 1. Left coronary artery and pulmonary artery were shown via collateral vessels in right coronary artery angiography.

Supplementary Materials

The online-only Data Supplement is available with this article at <https://doi.org/10.22468/cvia.2017.00164>.

Conflicts of Interest

The authors declare that they have no conflict of interest.

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