Anomalous Left Coronary Artery from the Pulmonary Artery Syndrome: A Case Report in a 49-Year-Old Woman

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Anomalous left coronary artery from the pulmonary artery (ALCAPA) syndrome is a rare congenital coronary artery anomaly. Clinical presentation can range from asymptomatic conditions to sudden death with exertion. The majority of cases is diagnosed in childhood and dies within the first year, however medical literature has confirmed several cases in adults. We report a case of a 49-year-old woman hospitalized for vague chest pain. Echocardiography found collateral circulation on the anterior wall, and ALCAPA syndrome was diagnosed on multi-slice CT.

Key words ALCAPA · ALCAPA syndrome · Bland-white-garland syndrome.

INTRODUCTION

Anomalous left coronary artery from the pulmonary artery (ALCAPA) syndrome, first described in detail in 1933 by Bland et al., is a rare congenital abnormality of the coronary system in which the left coronary artery (LCA) originates from the pulmonary artery (PA) instead of the aorta [1]. The prevalence is 0.25–0.5% of congenital cardiac abnormalities [1]. Most of these syndrome cases appear in isolation, though 5% of cases are associated with other cardiac anomalies such as atrial or ventricular septal defect, tetralogy of Fallot or coarctation of the aorta [2].

Based on onset time, ALCAPA is divided into infantile and adult types, which have different manifestations and outcomes. Children account for the majority of cases, and 90% of them die within the first year, only 10–15% of patients are diagnosed in adulthood [3]. Because of the low prevalence in adults, this syndrome might be ignored, and adult patients might not receive appropriate treatment.

CASE REPORT

A 49-year-old female patient was admitted with current unspecific left chest pain. Her medical report was unremarkable, and she had a history of three vaginal deliveries. No family history of diagnosed cardiac disease was noted, and her vital signs were normal. Her echocardiogram and chest X-ray showed no progressive findings. Echocardiography revealed a dilated and tortuous vessel in the cardiac apex and middle interventricular zone, so coronary fistula was suspected (Fig. 1). Coronary CT angiography was performed using a GE Revolution 512-slice CT Scanner (GE Healthcare, Chicago, IL, USA), and the results were as follows: anomalous origin of the LCA from the PA was noted; the left main artery (LM), originating from the pulmonary trunk, was sandwiched between the aorta and the pulmonary trunk; the left anterior descending artery, left circumflex artery, and branches followed appropriate pathways; and tortuous dilated left and right coronary arteries (RCA) with multiple collaterals at the anterior cardiac zone were noted (Fig. 2). Further assessment by digital subtraction angiography (DSA) was advised; however, she declined the additional work-up due to personal reasons.

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DISCUSSION

Physiologically in ALCAPA syndrome, because minor circulation remains functional, the pressure in the PA suddenly decreases, and retrograde flow from the RCA through collaterals to the LCA reverses to the PA, which is known as a coronary steal phenomenon. Clinically, young infants can experience tachypnea and irritability. If multiple collateral circulation sources from the RCA or systemic artery to the LCA elevate the retro-

grade flow, left ventricular perfusion is enhanced, and patients could survive to adulthood. However, this compensation is sometimes inadequate, and chronic left ventricular ischemia ensues. Thus, patients may develop malignant ventricular dysrhythmia that can lead to sudden death [4].

Several imaging modalities can be utilized for diagnosis. Echocardiography reveals the dilated origin of the RCA from the aorta with a suggestive ratio of RCA to aorta >0.2 and coronary collateral flows in favor of ALCAPA in adults. However, LM that

Fig. 1. Echocardiography of the patient. Two-dimensional ultrasound with a parasternal long-axis view demonstrates a small anterior pericardial effusion and small dilated vessels at the surface of the right ventricular anterior wall (A). Doppler ultrasound shows small and tortuous diastolic flows in the interventricular septum in parasternal long-axis view (B), apical four-chamber view (C) and apical three-chamber view (D).

Fig. 2. Coronal multi-slice CT of the patient. The LCA originates from the PA on VRT (A) with a retrograde flow from the LCA into the PA (white band in PA) on MIP sagittal plan (B). The LCA origin is sandwiched (arrows) between the aorta and PA on posterior plan on VRT (C). Tortuous intercoronary collaterals at the pical zone (arrows) are shown on the anterior plan on VRT (D). PA: pulmonary artery, VRT: volume rendering technique, LCA: left coronary artery, MIP: maximum intensity projection.

originates from the aorta can be difficult to visualize on transthoracic echocardiography [5]. Although DSA is the gold standard in coronary diseases, this invasive procedure is not appropriate for all patients. Therefore, multi-slice CT (MSCT) becomes an effective preference because high spatial resolution is supported in the coronary artery. This generally includes direct visualization of the LCA that arises from the main PA with retrograde flow, dilated and tortuous RCA or intercoronary collateral vessels, left ventricular hypertrophy, dilation, abnormal wall motion, mitral insufficiency and prolapse, and dilated bronchial arteries [6].

Differential diagnosis should be considered in diseases that cause dilated coronary arteries. In cases of coronary artery fis-

tula, there is arteriovenous communication or a fistula to the cardiac chamber that is depicted by dilated and tortuous coronary artery and epicardial veins; however, only the affected artery is dilated. It is also possible that diffuse coronary artery dilatation could be caused by atherosclerosis. Additionally, coronary aneurysm is a notable feature in Kawasaki's disease or Takayasu arteritis [6].

ALCAPA treatment primarily involves surgical repair to restore a two-coronary-artery circulation system, including a coronary button transfer in newborns and a coronary artery bypass graft with an LCA origin ligation in adults. Associated complications could include tearing of the analogous LCA in a coronary button transfer or supravulvar pulmonary stenosis and

baffle obstruction or leakage in a bypass graft [6].

In this case study, a 49-year-old woman was admitted for vague chest pain. Echocardiography depicted collaterals in the cardiac apex, and because of her medical history and the rare incidence of ALCAPA in adults, this syndrome was not initially suspected. However, because coronary CTA showed obvious evidence of anomalous LCA from the PA, the clinical diagnosis was adjusted. Therefore, based on this study experience, we recommend that ALCAPA syndrome be considered when cardiac apical collaterals are visualized with imaging, and that MSCT should be indicated when a coronary anomaly is suspected on echocardiography.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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REFERENCES

1. Dilaveris P, Koutagiar I, Alexopoulos N, Tsiachris D, Gatzoulis K. Secondary prevention of sudden cardiac death in a 65 year untreated ALCAPA patient. *Int J Cardiol* 2014;176:e73-e74.
2. Cankurt T, Acar B, Aydın S, Maden O. ALCAPA syndrome and atrial septal defect in a 68-year-old woman: an extremely rare congenital association. *Acta Cardiol Sin* 2017;33:447-449.
3. Fierens C, Budts W, Denef B, Van De Werf F. A 72 year old woman with ALCAPA. *Heart* 2000;83:E2.
4. Mancini MC. Anomalous left coronary artery from the pulmonary artery. *Medscape.com* Web site. <https://emedicine.medscape.com/article/893290-overview#a6>. Accessed August 10, 2020.
5. Saedi S, Parsaee M, Farrashi M, Noohi F, Mohebbi B. The role of echocardiography in anomalous origin of coronary artery from pulmonary artery (ALCAPA): simple tool for a complex diagnosis. *Echocardiography* 2019; 36:177-181.
6. Peña E, Nguyen ET, Merchant N, Dennie C. ALCAPA syndrome: not just a pediatric disease. *Radiographics* 2009;29:553-565.