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CASE REPORT

Adult-type ALCAPA: missed at preoperative evaluation

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Abstract
The Bland-White-Garland syndrome, or Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery (ALCAPA) syndrome, is a rare congenital cardiac anomaly often associated with high mortality, if left untreated. We present a case of a 43-year-old female with undiagnosed ALCAPA who initially underwent mitral valve surgery for severe mitral regurgitation, only to require reoperation due to adult-type ALCAPA. Intraoperatively, the discovery of dilated right coronary artery and its branches and absence of the left coronary ostium prompted further investigation, leading to the diagnosis of adult-type ALCAPA. Surgical correction remains the definitive treatment, with various techniques available depending on patient age and anatomical considerations. In this case, a modified Cabrol graft was successfully employed due to anatomical constraints.

Keywords Coronary artery anomaly · Surgical correction of ALCAPA

Introduction

The Bland-White-Garland syndrome, also referred to as Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery (ALCAPA) syndrome, is a cardiac anomaly that is rarely observed [1]. The incidence rate in the USA is 1 in 300,000 live births. Failure of diagnosis and treatment results in a 90% mortality rate within the first year of life [1]. Some patients having adequate collateralized coronary circulation can infrequently reach adulthood without any symptoms [2].

We present a patient with missed diagnosis of ALCAPA operated for severe mitral regurgitation, and later operated for adult-type ALCAPA requiring reoperation.

Case report

A female patient, aged 43 years, with a medical history of severe mitral regurgitation, presented to the outpatient clinic complaining of shortness of breath. Echocardiography (Echo) revealed severe mitral regurgitation (vena contracta > 7 mm, regurgitation volume 65 mL, left atrial size 5.1 × 9.1 cm, systolic pulmonary arterial pressure (SPAP) 40 mmHg, and reduced left ventricular ejection fraction (LVEF 50%)), but ALCAPA was not reported. Preoperative coronary angiography was unnecessary owing to the patient's youthful age, premenopausal status, and lack of cardiac risk factors for coronary artery disease.

Standard mitral valve surgery was planned. After median sternotomy and cardiopulmonary bypass (CPB), multiple coronary aneurysms were detected. The right coronary artery (RCA) was severely dilated and tortuous with large collaterals (Fig. 1). After the cross-clamp was placed, antegrade Del-Nido cardioplegia was administered, and cardiac arrest was maintained for only 12 min. Aortotomy was performed, and it was observed that there was only a single coronary artery ostium and selective antegrade cardioplegia by switching to Custodiol was maintained through the single coronary ostium, and cardiac arrest was achieved. Size 33 CarboMedics® (CarboMedics, Austin, Tex.) mechanical valve was implanted successfully. However, premature cardiac electrical activity was noticed even

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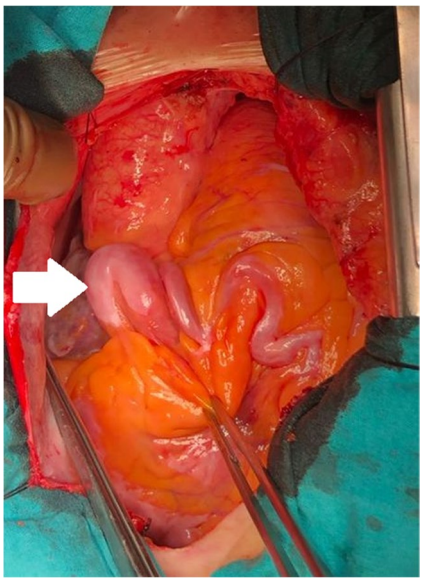

Fig. 1 White arrow: Dilated and tortuous right coronary artery

before the cross-clamp was removed suggesting shortening of period of cardioplegia. The patient was weaned from CPB. Postoperative period was uneventful. Computed tomography coronary angiography (CTA) was performed 10 days after the surgery to examine coronary anomalies. The CTA revealed adult-type ALCAPA; the left main coronary artery had originated from the pulmonary artery (PA) (Fig. 2). Echo revealed findings suggestive of ALCAPA (dilated right coronary artery (9 mm)), coronary collateral flow, anomalous origin of left main coronary artery (LMCA size 11 mm) from main pulmonary artery, and reverse flow in LMCA and circumflex artery (Video 1). Therefore, a reoperation decision was taken for corrective surgery.

She was operated on again 3 weeks after the first operation. Sternotomy was performed at the same incision line. After CPB was initiated, aorta and pulmonary artery were mobilized. Selective antegrade Custodiol cardioplegia was administered via the ostium of the left main coronary artery (LMCA) following pulmonary arteriotomy, and also via the aorta, resulting in cardiac arrest. LMCA was separated as a button. In infants, though the preferred technique remains LMCA button transfer and direct reimplantation to the aorta, but, due to long distance between pulmonary artery and aorta, mobilization of LMCA was not possible. Despite early reoperation, mobilization of the aorta and pulmonary artery was restricted. The creation of an aortopulmonary window proved to be challenging, rendering the Takeuchi operation unsuitable. Therefore, an end-to-end anastomosis was performed with a 10-mm dacron graft between LMCA and aorta. The defect on the PA was repaired with a pericardial patch. The patient was weaned from CPB.

After the second procedure, the patient was discharged in 10 days. At the postoperative third month, the patient was in good condition and there were no postoperative complications.

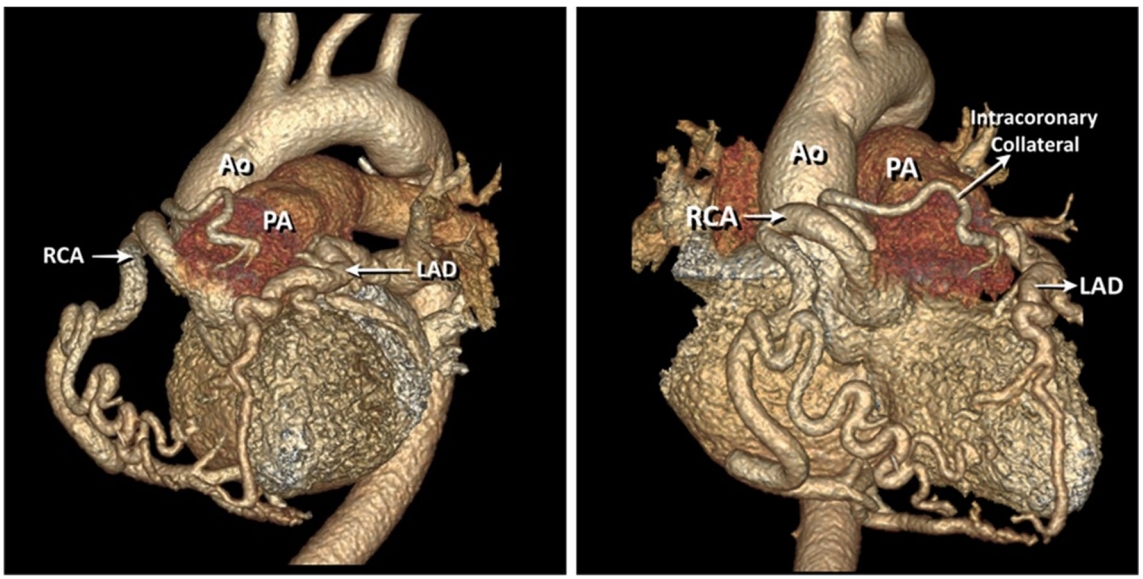

Fig. 2 Three-dimensional computed tomography coronary angiography (3-d CTA) images shows Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery (ALCAPA). Ao, Aort; PA, pulmonary artery; RCA, right coronary artery; LAD, left anterior descending coronary artery

Discussion

ALCAPA syndrome, which is very rare as a coronary artery anomaly, covers 0.2–0.5% of all congenital anomalies [3].

There are two types of ALCAPA: adult and infant type. Infant-type ALCAPA presents with symptoms of myocardial ischemia, heart failure, and even sudden death in early life. On the contrary, in adult-type ALCAPA syndrome, symptoms of myocardial ischemia may be suppressed due to adequate collateral circulation between right and left coronary arteries. Exacerbated pain, left ventricular dysfunction, ventricular arrhythmias, mitral insufficiency, and sudden death may develop with exercise due to subendocardial ischemia in the left ventricle [4]. Due to this ischemia, the cardiac damage could be sometimes irreversible even if the successful correction was performed. Despite our patient never experiencing angina before, dyspnea symptoms had begun for 1 year. Inherently, mitral insufficiency caused symptoms rather than ALCAPA.

Some hallmark signs can provide clues for a diagnosis for disguised ALCAPA. Dilated right coronary artery, ostial agenesis of coronary artery or coronary arteriovenous fistula, typical flow pattern called "to-and-fro," extensive collateral network, and mitral regurgitation developed as a consequence of ventricular dilatation following ventricular ischemia, papillary muscle ischemia, and dilated PA which suggest adult-type ALCAPA [5]. Although Echo was performed many times, ALCAPA was missed in our patient. There were several important points that could have led us to the diagnosis of ALCAPA syndrome during operation. First, the right coronary artery was aneurysmal and tortuous; second, dilated collateral vessels were present on the heart surface; third, the absence of the left coronary ostium; and finally, failure to achieve diastolic arrest despite adequate cardioplegia.

The definitive treatment of ALCAPA is surgical correction. Surgical treatment is required to avoid left ventricular dysfunction, arrhythmias, mitral insufficiency, or sudden death as soon as the diagnosis is made. Even if the patient is asymptomatic, conservative medical treatment is controversial due to the risk of sudden death and cardiac failure [6]. Various techniques are described; however, direct reimplantation is the most common. This technique can be performed more easily in infants. Contrarily in adulthood, it is not easy to perform because of the distance and stiffness of the dilated PA and aorta. Takeuchi procedure which involves the creation of an aortopulmonary window and an intrapulmonary tunnel connecting the aorta to the ostium of the abnormal left coronary artery, or LMCA occlusion followed by coronary artery bypass surgery, can be performed [7]. There is no consensus on the best surgical correction technique; however, reimplantation technique creating a dual coronary system is more likely to restore natural coronary anatomy [8]. Modified Cabrol technique provides satisfactory surgical correction and avoids kinking [9]. Since reimplantation was not suitable, because of the long distance of target, we performed modified Cabrol graft between the ostium of LMCA and the aorta.

Conclusion

ALCAPA is very rare in adulthood. Cardiologists and cardiovascular surgeons should keep in mind these rare coronary anomalies. In adult population, direct visualization of coronary artery origin is difficult and therefore indirect clues, as mentioned before, need to be looked for, by experienced echocardiographers and other modalities must be used, if required. A Cabrol bypass technique in adults may be preferable with ease to establish normal circulation without any complications; however, long-term follow-up is needed.

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