Case Report

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Anomalous origin of the left coronary artery from the pulmonary artery in a patient of advanced age: a case report and analysis

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Abstract

A 61-year-old Chinese man presented with a nearly 30-year history of an anomalous origin of the left coronary artery. He had been diagnosed with an anomalous origin of the left coronary artery in 1989. He then underwent regular echocardiographic examinations and it was found that his heart was gradually enlarging. After a >20-year asymptomatic period, he developed recurrent chest discomfort and palpitation. Coronary computed tomography angiography suggested that the left coronary artery anomaly originated from the pulmonary artery; additionally, the right coronary artery was tortuous and thickened. Coronary angiography showed that the right coronary artery was huge and buckling. The patient underwent corrective surgery of the anomalous origin of the left coronary artery from the pulmonary artery, aortic valve mechanical valve replacement, mitral valve plasty, and tricuspid valve plasty in Fuwai Hospital (National Center of Cardiovascular Disease of China), and the anatomic results of the surgery were good.

Keywords

Cardiovascular abnormalities, ALCAPA, Bland–White–Garland syndrome, echocardiography, coronary computed tomography angiography, corrective heart surgery

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Introduction

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA), also known as Bland–White–Garland syndrome, is a rare congenital coronary abnormality. Most patients with ALCAPA are diagnosed as infants, and few survive to adulthood without surgery. We herein describe a case involving a 61-year-old Chinese man who lived a healthy life until he was diagnosed with anomalous origin of the left coronary artery (LCA) at the age of 32 years. We followed him for 29 years before he developed symptoms, and treatment was successful. This case may raise our awareness of ALCAPA in patients of advanced age and further elucidate the pathogenesis of this disease.

Case description

A 61-year-old Chinese man presented with a 29-year history of an anomalous LCA and a 4-month history of chest discomfort and palpitation. He had been diagnosed with an anomalous origin of the LCA in 1989, at which time he had no discomfort. In 2007, he underwent digital subtraction angiography (DSA) at the First Affiliated Hospital of Zhejiang University School of Medicine, which showed no obvious stenosis in left main coronary artery. However, it revealed severe lesions and multiple stenoses in the LCA with a possible fistula from the LCA to pulmonary artery (PA). Additionally, the right coronary artery (RCA) had a large diameter and possible fistula to the PA. The proximal segment of the other branch was not stenosed, the middle segment was thin, the distal segment was not seen, and the collateral circulation was abundant. He sought treatment in several hospitals, but conservative treatment was performed; the patient declined surgery because he was asymptomatic and the operation was high-risk. During this period, the patient was regularly re-examined. We observed him for more than 10 years and recorded his examination findings (Table 1).

In September 2017, the patient developed chest discomfort, palpitation, and decreased activity tolerance. He visited several hospitals successively. Dynamic electrocardiogram at Zhejiang Provincial Hospital of TCM revealed sinus rhythm and sporadic atrial and ventricular premature beats. Echocardiography at Zhongshan Hospital revealed congenital heart disease characterized by the following: ectopic origin of the LCA from the PA and widening of the RCA, left ventricular hypertrophy with decreased left ventricular systolic activity, left atrial enlargement with severe mitral valve regurgitation, widening of the aortic sinus and ascending aorta with moderate to severe aortic regurgitation, and severe pulmonary hypertension with moderate pulmonary valve regurgitation. He was subsequently admitted to the Second Affiliated Hospital of Zhejiang University School of Medicine. He had no coronary risk factors

Table 1. Echocardiography reports.

Year	1993	1997	2000	2004	2010	2012	2016
LVEF	60%	54%	61%	54%	56%	53%	54%
LVDd, mm	63	60	60	64	68	73	73
LA, mm	35	31	37	38	47	45	53

LVEF, left ventricular ejection fraction; LVDd, inner diameter of left ventricle at the end of diastole; LA, left atrial diameter

The echocardiography reports showed that the left heart, including both the left atrium and left ventricle, was growing larger and that the cardiac systolic function was deteriorating.

or family history of congenital heart disease. Physical examination findings were normal. Serial measurements of cardiac enzymes were negative. During his hospitalization, coronary computed tomography angiography (CTA) revealed the anomalous LCA originating from the PA; a tortuous and thickened RCA, with a few focal calcifications and mixed plaques in the proximal and middle segments of the RCA and slight localized narrowing of the lumen of the RCA; widening and tortuosity of the left main coronary artery, left anterior descending branch, diagonal branches, and left circumflex branch (LCX); significant enlargement of the left atrium and left ventricle; and widening of the PA and its branches (Figure 1).

Subsequent DSA showed an abnormal coronary artery, large RCA with no obvious stenosis and adequate provision of collateral circulation to the LCA, and origination of the LCA from the PA; pulmonary angiography did not show the LCA (Figure 2). The PA pressure was 77/31 mmHg, and the mean PA pressure was 46 mmHg.

The patient later sought surgical treatment at Fuwai Hospital (National Center of Cardiovascular Disease of China). Before surgery, myocardial perfusion imaging was performed to assess the myocardial viability. This examination showed that the left ventricular cavity was obviously enlarged and that the myocardial radioactivity was not uniform in each wall of the left ventricle, but there was no obvious radiation reduction or defect area. Because the patient had no surgical contraindications, corrective surgery of ALCAPA was performed.

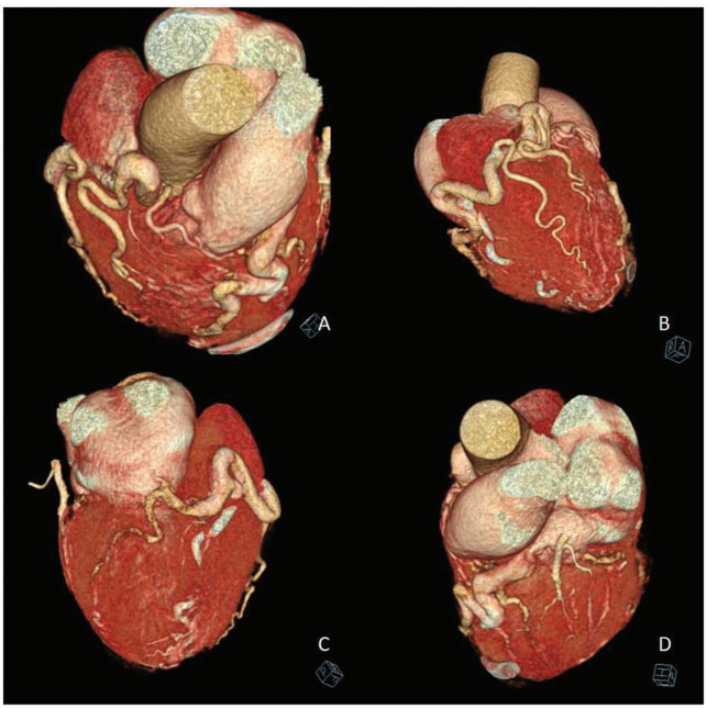

Figure 1. Coronary computed tomography angiography images. (a) The left coronary artery anomaly originates from the pulmonary artery, and the right coronary artery is tortuous and thickened. (b, c) The right coronary artery is tortuous and thickened. (d) The origin of the left coronary artery can be seen.

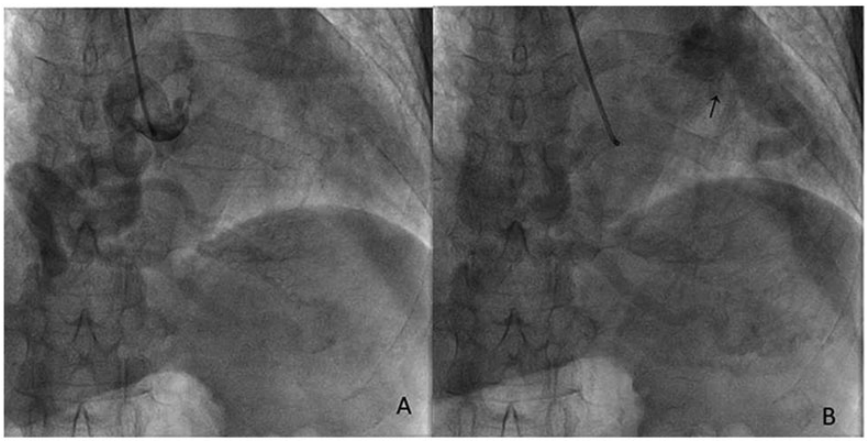

Figure 2. Digital subtraction angiography images. (a) Angiography shows a large and tortuous right coronary artery. (b) In the late phase of right coronary artery angiography, the left coronary artery was filled in reverse and the pulmonary artery was visible at the left coronary opening.

An autologous pericardial patch was used to implant the coronary artery into the aorta. During this procedure, mechanical aortic valve replacement, mitral valve plasty, and tricuspid valve plasty were also performed. The results of transesophageal echocardiography were normal after cardiac rebeating. Echocardiography after this operation showed that the inner diameter of the left ventricle at the end of diastole was 62 mm (versus 73 mm before the operation), aortic regurgitation was not seen (versus moderate to severe aortic regurgitation), and mitral valve regurgitation was mild to moderate (versus severe mitral valve regurgitation). After the surgery, warfarin, nitrates, and a diuretic were given as pharmacotherapy.

Ethics and consent

Written informed consent was obtained from the patient. This need for ethics approval was waived because the patient regularly presented for re-examinations, and we performed these re-examinations for clinical treatment rather than medical research. After he had successfully undergone surgery, we found this to be an interesting case and collected the examination findings to complete this case report.

Discussion

ALCAPA, also known as Bland–White–Garland syndrome, is a rare congenital coronary abnormality that appears in 1 in 300,000 live births and accounts for 0.25% to 0.50% of all congenital heart anomalies.[1] However, the actual incidence of this disease may be higher. It predominantly presents in the first year of life, and few patients survive to adulthood without surgery.[2] Adult patients with ALCAPA may present with angina, exertional dyspnea, or sudden cardiac death, or ALCAPA may be incidentally found on imageological examinations such as DSA.[3–5]

In the past, the diagnosis of ALCAPA was mainly dependent on DSA and autopsy.[4,6] However, as imaging technology has progressed, noninvasive techniques such as coronary CTA, magnetic resonance imaging, ultrasonography, and myocardial perfusion imaging have been used to assess coronary artery malformations and thus assist in the diagnosis of ALCAPA.[7,8]

For our patient, the first DSA examination did not result in a diagnosis ALCAPA, but coronary CTA revealed the abnormal origin of the left coronary artery from the PA and thus established the diagnosis of ALCAPA. Taking each characteristic finding of DSA and CTA into consideration, we consider that although DSA more clearly shows microcosmic structures of the vessels, CTA sometimes has advantages in terms of facilitating assessment of the macroscopic structures. Ultrasonography is a very convenient and practical examination. Two-dimensional ultrasonography can display abnormal connections between the LCA and PA, and color Doppler flow imaging can display the abundant collateral circulation between the LCA and RCA as well as the abnormal jet flow between the LCA and PA, thus playing an important role in identifying patients with ALCAPA. The diagnosis of the coronary artery malformation in this patient 29 years previously was based on echocardiography findings. Cardiac magnetic resonance imaging can be used to assess the coronary anatomy, ischemia, and heart function, but because it has not been widely developed in China, we adopted myocardial perfusion imaging to assess the ischemia as a supplement of CTA and ultrasonography.

In the fetal and early infant stages of patients with ALCAPA, the origin of the LCA from the PA can result in antegrade flow from the PA because of high PA pressure. However, as the pressure of the PA decreases after birth, RCA–LCA–PA shunting occurs. This is a true high-flow, low-pressure shunt that damages the blood supply of the LCA and LCX, affects the blood perfusion of the left ventricle, and leads to ischemia of the LCX and subendocardium. Because of this high-speed, low-obstruction blood flow, the local endothelium is stimulated by shear force and the epicardial artery expands. The lateral branch circulation between the RCA and LCA origin from the PA mainly develops at the level of the epicardium (anterior right ventricle, cardiac notch) and the compartment perforation branch.[9] The well-developed collateral circulation prevents local blood flow hypoperfusion and early acute symptoms. Our patient was asymptotic when he was diagnosed with the coronary artery malformation 29 years previously. Eleven years previously, RCA angiography showed that the RCA was thickened, abundant collateral circulation had developed, and severe lesions and multiple stenoses were present in the LCA. However, the rich blood supply of the collateral circulation from RCA compensated for the low left ventricular myocardial blood supply; thus, the cardiac function was not significantly impaired.

Follow-up of this patient for more than 20 years with recording of his echocardiographic parameters revealed that his left heart was large and that both his left ventricle and left atrium were gradually increasing. The progression was analogous to that of ischemic cardiomyopathy: insufficiency of the left heart blood supply led to myocardial dystrophy and shrinking, resulting in fibroplasia and cardiac dilatation. Because the insufficiency involved the papillary muscles, mitral valve, and subvalvular chordae tendineae, mitral incompetence and regurgitation occurred. At the same time, relative mitral incompetence occurred because of the expansion of the left heart, which aggravated the mitral regurgitation. The mechanism of aortic and tricuspid regurgitation is similar to that of mitral regurgitation. As the whole heart expanded, the aortic ring and tricuspid ring enlarged, resulting in aortic and tricuspid regurgitation. The larger the left heart, the more severe the regurgitation. ALCAPA is a potential, although uncommon, cause of mitral regurgitation and dilated cardiomyopathy.[10] In this case, although the left heart was significantly enlarged and severe

mitral regurgitation was present, the left ventricular systolic function was only slightly decreased, as shown in Table 1. As the patient's heart gradually enlarged, surgery was recommended at each subsequent visit, but he refused because he was asymptomatic and the surgery was very high-risk. During the asymptomatic period of more than 20 years, the patient's left heart function gradually lost compensation, and chest discomfort and palpitation appeared. These symptoms of coronary insufficiency may not only relate to coronary lesions such as atherosclerosis and stenosis but also to aortic regurgitation, which leads to a decrease in aortic flow and insufficiency of RCA perfusion. Because the symptoms affected his daily life, the patient began to consider surgery.

Surgery is the main treatment of ALCAPA. The technique to repair an anomalous origin of the coronary arteries can be achieved by direct reimplantation of the coronary artery into the aorta by transferring a button of the PA or by ligature of the origin of the artery along with coronary artery bypass grafting.[8] For our patient diagnosed with ALCAPA, the aortic ring was enlarged along with moderate to severe aortic regurgitation, the mitral ring was enlarged along with severe mitral valve regurgitation, and the tricuspid ring enlarged; thus, simple corrective surgery for ALCAPA would not have necessarily improved the structure and function of the heart. Thus, a combination of mechanical aortic valve replacement, mitral valve plasty, and tricuspid valve plasty was performed. The anatomic results of the surgery were good. The heart shrank and the regurgitation of the aortic valve and mitral valve decreased.

Conclusion

Imaging methods, especially echocardiography, are of great significance to the diagnosis and follow-up of ALCAPA. CTA may be better than DSA for assessment of the macroscopic structure. Because of the formation of collateral circulation, patients with ALCAPA can be asymptomatic in the early stages. However, as the disease progresses, cardiac structure changes, left heart expansion, and mitral regurgitation appear, gradually leading to symptoms of cardiac insufficiency. Surgery is the main treatment of ALCAPA.

Declaration of conflicting interest

The authors declare that there is no conflict of interest.

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