Congenital absence of left coronary artery: a case report and literature review

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March 31, 2022

Abstract

Single coronary artery (SCA) is defined as an isolated coronary artery that arises from a single coronary ostium and provides blood supply to the entire myocardium. At present, the absence of the right coronary artery is more common in SCA and the absence of the left coronary artery is rare. We report a case of a middle-aged man with congenital left coronary artery absence. The patient underwent coronary angiography after completing relevant examinations on admission, the results showed that the coronary artery blood supply was dominant in the right coronary artery; the right coronary artery was compensatively thickened and long, the part of the left coronary artery that functions as the anterior descending branch opens at the beginning of the right coronary artery and the blood vessels were small. The patient was given symptomatic supportive treatment during the hospitalization period and was discharged from the hospital. The patient's condition was stable during the follow-up. This case demonstrates that although congenital left coronary artery absence is a rare disease, correct diagnosis, appropriate treatment and patient education play a vital role in the treatment of these patients.

1. INTRODUCTION

The popularity of coronary angiography technology has increased the detection rate of coronary artery malformations, and at the same time, the understanding of the diagnosis and treatment of coronary artery malformations has become more and more in-depth[1].Coronary artery malformation is a rare congenital malformation. Different coronary artery malformations have different effects on the blood supply to the heart. Among them, SCA is defined as an isolated coronary artery that arises from a single coronary ostium and provides blood supply to the entire myocardium[2]. At present, the absence of the right coronary artery is more common in SCA and the absence of the left coronary artery is rare. This article reports the clinical data of a patient with congenital left coronary artery absence who was admitted to the Department of Cardiology, the Second Affiliated Hospital of Nanchang University and reviewed the literature.

2. CASE REPORT

A 44-year-old man presented with 1-year history of chest pain. The patient had chest tightness in the precordial area, severe chest pain, and tingling at night without obvious inducement 1 year ago. The duration was about 5 minutes per time, and it could relieve itself after a rest. No attention, diagnosis and treatment were given, and the chest pain reappeared 2 months ago, with the same symptoms as before, accompanied by a feeling of choking. The outpatient electrocardiogram (ECG) (Figure 1) showed sinus rhythm, poor r-wave progression in V1-V3 leads with ST-segment elevation must be combined with clinical exclusion of anterior septal myocardial infarction and J wave in inferior leads. He had undergone radical surgery for colon cancer in a local hospital in 2006. The patient's blood pressure was 108/71 mmHg and a heart rate was 88 beats/minute and his physical examination was unremarkable. Initial laboratory tests showed serum creatinine 74.00 μmol/L, estimated glomerular filtration rate 105.95, total cholesterol 4.40 mmol/L, triglycerides 2.72 mmol/L, and plasma homocysteine 11.24 μmol/L, cardiac enzymes, electrolytes and BNP were normal.

Transthoracic echocardiography (Figure 2) showed aorta ascendens 27mm, right atrium internal diameter 36mm, right ventricular anterior wall thickness 4mm, right ventricle diameter 19mm, interventricular septum thickness 10mm; left atrium internal diameter 34mm, left ventricular posterior wall thickness 9mm, left ventricularend diastolic dimension 44mm, left ventricularend systolic dimension 27mm, and the left ventricular ejection fraction is 68%. Eventually showed that the mitral valve, tricuspid valve and aortic valve insufficiency. Chest X-ray was unremarkable. Coronary angiography (Figures 3-4) was performed. Multi-position angiography showed that the coronary artery blood supply was dominant in the right coronary artery; the right coronary artery was compensatively thickened and long, the distal segment and branches extended to supply the left ventricle, as well as small plaques can be seen in the branches. Repeated attempts have failed to find the opening of the left coronary artery, the part of the left coronary artery that functions as the anterior descending branch opens at the beginning of the right coronary artery and the blood vessels were small. After coronary angiography, check the ECG (Figure 5) showed sinus rhythm and roughly normal ECG. The patient was given symptomatic supportive treatment(Aspirin、Atorvastatin、Pantoprazole) during the hospitalization period and was discharged from the hospital. The patient's condition was stable during the follow-up.

3. DISCUSSION

Coronary artery malformations are usually discovered accidentally during coronary angiography and autopsy, accounting for about 2% of the total population[3],SCA is a rare malformation in coronary artery malformations. Yamanaka O et al.[2]summarized 126595 cases of coronary artery disease. The data of patients with angiography showed that the incidence of SCA was 0.04%.Al Umairi, R et al.[4]included 4445 patients with coronary angiography and found that 12 cases were SCA, with an incidence of 0.27%. SCA can exist alone or coexist with other congenital abnormalities of the heart. Among the reported cases, 18%-40 % of patients are accompanied by other cardiac malformations (tetralogy of Fallot, ventricular septal defect, patent ductus arteriosus, patent foramen ovale, coronary arteriovenous fistula, etc.)[5].Congenital absence of the left coronary artery belongs to a special type of SCA, and its incidence is extremely low, only 0.024%, and there is no gender difference [6].

According to the origin of SCA and the anatomical distribution of branches[7]: Type I, the distal segment of SCA continues as the contralateral coronary artery; Type II, after the origin of SCA, the larger branches are distributed through the root of the aorta to the contralateral coronary artery. According to this branch, it runs in the right ventricular conus or anterior pulmonary artery (IIA), between the aorta and pulmonary artery (IIB), or posterior to the aortic root (IIP); Type III, SCA originates from the right sinus, circumflex branch and the anterior descending branch travels through the back and front of the aorta, respectively. According to the above classification, this case belongs to the R-IIA type. The common features of the R-II type are: a single right coronary artery originates from the right coronary sinus, and a single right coronary artery has a large lumen and good filling, and can emit small branches during walking to supply blood to the left ventricle.

The etiology of SCA has not been fully determined. It is generally believed that the coronary arteries are abnormally or incompletely developed in the embryo, and the arterial occlusion occurs in the embryo. Many SCA case reports have been published in countries with high blood relationship rates[8]. Although blood relationship is related to a variety of heart diseases, the correlation with SCA has not yet been clear [9]. Some authors believe that certain mechanisms can explain the occurrence of SCA, such as the coronary steal phenomenon due to the abnormal vessel or microvascular damage, and slow controlled ischemia caused by long travel distance of abnormal coronary artery[10].

SCA usually does not have any clinical manifestations, and the clinical manifestations lack specificity. Some patients may show a simple angina pectoris. Arteaga et al[11] believe that angina pectoris does not occur in the absence of coronary artery stenosis. The ECG may have ST-T changes, sinus arrhythmia, atrioventricular block and other manifestations[3]. If SCA has risk factors such as age, stress, infection, trauma, etc., it can cause myocardial infarction, arrhythmia, heart failure, syncope, sudden death, without obstructive coronary artery disease[12]. In this case, due to chest pain, coronary angiography was performed to rule out coronary

syndrome, and the result was diagnosed as congenital absence of the left coronary artery.

In the clinic, the misdiagnosis rate of SCA is relatively high based on routine examinations or clinical symptoms[13],therefore, it is very important to accurately determine the specific conditions and subtypes of SCA patients at the diagnosis stage. At present, coronary angiography is considered to be the "gold standard" for diagnosing coronary artery disease.

Some literatures pointed out that with SCA examination as the reference standard, the negative predictive value of coronary CT angiography (CCTA) at the level of patients, blood vessels, and segments is about 90%, 97%, and 97%, respectively[4]. In recent years, the use of fractional flow reserve (FFR) and intravascular ultrasound (IVUS) to guide therapy is increasingly gaining attention. FFR can help evaluate the hemodynamics of blood vessels and IVUS can help identify slit like or stenosed orifice, acute angle takeoff, and intramural aortic segment[14].

For the treatment of SCA, there are no standards or guidelines based on evidence-based medicine. The treatment plan is mainly conservative treatment, such as the control of blood pressure, blood lipids and blood sugar, etc. And the use of β-blockers to control the rhythm can also be considered. If SCA patients have severe arrhythmias such as sick sinus syndrome, atrial fibrillation, and high-grade atrioventricular block, permanent pacemakers need to be implanted[15]. In this case, a conservative treatment plan was finally adopted, and the follow-up situation is stable.

4. CONCLUSION

In summary, congenital left coronary artery absence is a rare disease, and its clinical manifestations often lack specificity. CCTA or coronary angiography should be performed in time to rule out congenital left coronary artery absence and other congenital cardiovascular malformations. Correct diagnosis, appropriate treatment and patient education play a vital role in the treatment of these patients.

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FIGURE LEGENDS

Figure 1: Outpatient ECG showed sinus rhythm, poor r-wave progression in V1-V3 leads with ST-segment elevation must be combined with clinical exclusion of anterior septal myocardial infarction and J wave in inferior leads.

Figure 2: Transthoracic echocardiography showed the mitral valve, tricuspid valve and aortic valve insufficiency.

Figures 3-4: Coronary angiography showed the right coronary artery was compensatively thickened and long, the distal segment and branches extended to supply the left ventricle, as well as small plaques can be seen in the branches, the part of the left coronary artery that functions as the anterior descending branch opens at the beginning of the right coronary artery and the blood vessels were small.

Figure 5: Re-scrutinized ECG showed sinus rhythm and roughly normal ECG