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Case Report

Anomalous of the Left Coronary Artery Originating from the Pulmonary Artery in Children - A Mini Review and a Case Report

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Abstract

Anomalous of the Left Coronary Artery Originating from the Pulmonary Artery (ALCAPA) is a rare congenital heart disease. The onset time and severity of clinical symptoms of ALCAPA were determined by the rate of pulmonary artery pressure decline, the opening of collateral circulation, and the stenosis of the opening of the left coronary artery at the origin of ectopic location. If there is little or no collateral circulation between the coronary arteries, symptoms of myocardial ischemia and heart failure may occur early in the child. If abundant collateral circulation is formed between the left coronary artery of ectopic origin and the right coronary artery of normal position, the left coronary artery can maintain enough blood perfusion, and the symptoms will appear later. But "steal phenomenon" in the left coronary artery, can also cause hypoxia and ischemia of the myocardium, leading to significant cardiac enlargement and mitral regurgitation. Early diagnoisis and surgical treatment can significantly improve the prognosis of ALCAPA. We also conducted a retrospective case of a child with ALCAPA, to better understand the pathophysiology.

Keywords: Pulmonary Artery; Children; Congenital Heart Disease; ALCAPA

Case Report

A 2-years old boy, was hospitalized mainly due to "fever and cough for 4 days". There were no obvious symptoms of hyperhidrosis, difficulty in feeding and activity limitation, and no obvious abnormalities in growth and development. He had suffered from "bronchopneumonia" at 4 months and "upper respiratory tract infection" 5 times in the past 1 year. Family history was normal. Physical examination: body temperature 38.6°C, weight 13.3 kg, height 99 cm, breathing 38 times/min, pulse 158 times/min, blood pressure 93/60 mmHg. There was no rash, hyperemia of pharynx, no obvious swelling of bilateral tonsils, no purulent secretion. Moderate and fine moist rales could be heard in both lungs. There was no uplift in the precardiac area, the percussion cardiac boundary was slightly enlarged, the cardiac sound was strong, the rhythm was even, there was no hearing and splitting, and grade II systolic murmurs and short-term diastolic murmurs could be heard in the apex of the heart. The liver and spleen were not reached under the ribs. Auxiliary examinations before admission: routine blood hemoglobin 122 g/L, WBC 5.16 × 109/L, neutral 48.04%, lymph 41.43%, mononuclear 9.94%, eosinophilic 0.60%, basophilic 0.20%, platelet 229×109/L, CRP < 0.499 mg/L. Chest radiograph showed bronchopneumonia with enlarged heart shadow. An electrocardiogram examination after admission showed sinus tachycardia (heart rate 158 BPM), high left ventricular voltage (RV5 = 4 mv), Tv4-tv6 bidirectional, ST segment descending oblique or horizontal. Preliminary diagnosis: 1) bronchopneumonia; 2) congenital abnormal origin of coronary arteries; 3) dilated cardiomyopathy; 4) cardiac function level I.

Treatment: After admission, the patient was given symptomatic treatment such as anti-infection, cough and phlegm reduction, and nourishing myocardium, etc., and the body temperature fell to normal after 3 days of hospitalization, but the rales were continuously audible at the bottom of both lungs. The cough improved after 5 days of hospitalization, and the rales at the bottom of both lungs were reduced after auscultation. After 7 days in the hospital, The rales could not be heard, and the patient was discharged automatically. During the period of hospitalization, due to auscultation of precardiac murmur, chest radiograph enlargement of cardiac shadow, and electrocardiogram abnormality, ultrasonic echocardiography was recommended. The patient agreed the boy to have echocardiography examination after he recovered from pneumonia. After discharge, the patient was admitted to the department of cardiology. Echocardiography showed that the abnormal left coronary artery originated from the pulmonary artery, left ventricular enlargement, segmental motor abnormality of the ventricular wall,

and mitral regurgitation (moderate). After that, the patient was examined by enhanced CT and the CT showed anomalous origin of coronary artery, left coronary artery from the pulmonary artery root left side, left, left ventricular myocardial ischemic. The boy underwent “correction of coronary origin anomaly + mitral valvuloplasty "and an echocardiography one month after surgery showed improved mitral valve closure and normal left ventricular systolic function. So far no discomfort, no respiratory tract infection symptoms. At a 1-year follow-up, the boy recovered well and showed no signs of respiratory infection or cardiac insufficiency.

Discussion

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital cardiac anomaly, which is caused by malrotation of arterial trunk and pulmonary artery septa and dislocation of coronary artery germ. This congenital heart disease was first reported in 1885, the disease was first described clinically in 1933 by Bland, White, and Garland, hence called the bland-white-garland syndrome [1]. The incidence of is relatively low, about 0.25% ~ 0.50% of the total number of congenital heart disease [2] accounting for about 0.13% of congenital heart disease in Asian populations [3], 90% of ALCAPA infants died of congestive heart failure within 1 year old, about 10% ~ 15% of children can survive to adolescents and adults [4], surgery is the only treatment and long-term effect is good.

The onset time and severity of clinical symptoms of ALCAPA are influenced by the factors including the rate of pulmonary artery pressure decline, the opening of collateral circulation, and the stenosis of the opening of the left coronary artery from the origin of ectopic location [5]. ALCAPA infants do not present no myocardial ischemia temporarily in 4~5 weeks after the birth due to high pulmonary circulation resistance and adequate coronary artery perfusion. From 1 month to a few months after birth, with the decrease of pulmonary vascular resistance, the pressure of the left coronary artery decreased and the blood perfusion decreased. If sufficient collateral circulations are not formed, hypoxia changes may occur in left ventricular cardiomyocytes, and clinical symptoms such as shortness of breath, hyperhidrosis, irritability and difficulty in feeding may occur. Clinical examination showed the manifestations of congestive heart failure, the apex of the heart can be heard and systolic blowing-like murmurs or continuous murmurs, X-ray may have heart shadow expansion. If abundant collateral circulation is formed between the left coronary artery of ectopic origin and the right coronary artery of normal position, the left coronary artery can maintain enough blood perfusion, and the symptoms will appear later. However, the "steal phenomenon" in the left coronary artery, that is, the blood carrying oxygen circulates from the collateral branch of the right coronary artery to the left coronary artery, and then it is injected into the pulmonary artery with low pressure to form the left-right shunt, which still affects the blood supply of the left ventricular cardiomyocytes and causes the symptoms of myocardial hypoxia. According to the pathophysiological characteristics of this disease, it can be divided into two types. Those with enough collateral circulation are called "the adult type", That is, there is no obvious clinical manifestation of cardiac insufficiency in infancy. In the "infantile" cases, there are no significant collateral angiogenesis between the left and right coronary arteries, and the corresponding symptoms of significantly reduced left ventricular myocardial perfusion appeared in the early infantile period. Typical clinical manifestations of ALCAPA infantile type include hyperhidrosis, shortness of breath, difficulty in feeding, recurrent heart failure, respiratory tract infection, and cardiac enlargement. Adult ALCAPA patients may be asymptomatic, or have symptoms associated with myocardial hypoxia, mitral regurgitation, cardiomyopathy, and malignant arrhythmia [1,6]. Some researchers divide ALCAPA into three types: in addition to the above adult type and infantile type, there are transitional types, which is characterized by both the above two types, with both the formation of collateral circulation and severe myocardial ischemia or myocardial infarction [7]. In our case, the patient did not have typical heart failure symptoms during growth. However, frequent respiratory infections and electrocardiogram and other examinations revealed myocardial ischemia, suggesting that collateral circulation of bilateral coronary arteries and “steal phenomenon” co-existed.

The clinical manifestations of ALCAPA are not specific, and its diagnosis depends on the imaging examination. 1) Electrocardiogram: under normal circumstances, the left coronary artery opens in the left coronary sinus, and the blood supply range of the left coronary artery is the anterior wall of the two ventricles, the anterior 2/3 of the ventricular septum, the right bundle branch and the left anterior branch, as well as the lateral wall, posterior wall and inferior wall of the left ventricle. Since the left coronary artery originates from the pulmonary artery, the oxygen content in the blood flowing into the left coronary artery is decreased, leading to local myocardial ischemia and hypoxia. Left ventricular subendocardial myocardial ischemic is often manifested as ST segment descending on the electrocardiogram. Subepicardial myocardial injury is often characterized by ST segment elevation and T - wave inversion. On the other hand, myocardial cells in the left ventricular wall distributed in the left coronary artery can produce infarction due to ischemic, and scattered scars can be found under the intima. Abnormal Q waves often appear in the leads of I, aVL, V 5 and V 6 of the electrocardiogram. Under normal circumstances, there is no Q wave in the lead of I, aVL of infant electrocardiogram. Once Q wave occurs, it is considered pathological. When I, aVL,V4-V6 leads show deep and wide Q waves, it is often suggested that the child

may have a left coronary artery originating from the pulmonary artery. Therefore, abnormal Q wave, ST segment depression and T wave inversion in the ecg lead I, avL and v4-6 lead have high sensitivity and specificity for the diagnosis of ALCAPA heart failure and cardiac enlargement in infants [8,9]. 2) Echocardiography is one of the main means of noninvasive examination of ALCAPA.If no left coronary artery is seen at the aortic root and abnormal blood flow appears in the pulmonary artery, ALCAPA can be diagnosed when the blood flow is connected to the left coronary artery trunk The widened right coronary artery and abundant collateral blood flow signals in the ventricular septum can be distinguished from cardiomyopathy and mitral regurgitation [10,11]. 3) Spiral CT and three-dimensional reconstruction of the heart, which can show the exact ectopic origin of left coronary artery from the path, as a reliable noninvasive diagnostic method, has been widely used in the diagnosis of congenital heart disease, through the three-dimensional imaging technology to provide information can partly replace cardiac coronary angiography, and to determine the left coronary artery and pulmonary artery connection relationship is very helpful. In the diagnosis of coronary artery disease in children, only the morphological anatomy of main coronary artery and branch can be shown. 4) coronary angiography: it can fully display the whole coronary artery system. The collateral circulation between coronary arteries and the origin of abnormal coronary arteries in the main pulmonary artery can provide important information for surgery, which cannot be replaced by other imaging examinations at present, but it is invasive and seldom used; 5) radionuclide myocardial imaging: mainly used for the evaluation of viable myocardium, providing important information for the selection of treatment options for children and the judgment of postoperative cardiac function recovery [12].

Because of the high mortality rate of ALCAPA in children and the risk of sudden death, early surgery is required upon diagnosis. The purpose of surgical treatment is to establish a dual coronary system, stop the " Steal phenomenon " of the left coronary artery of the pulmonary artery, and increase the blood supply of the left ventricular myocardium. The most common radical ALCAPA operations are intrapulmonary tunneling and left coronary replantation. Left coronary artery replantation is the most consistent with human physiological structure and has become the preferred surgical method of ALCAPA [13,14]. The short-term and long-term prognosis after replantation of the left coronary artery is good, and the factors influencing the surgical effect include the degree of left atrioventricular valve regurgitation, the decline of left cardiac function, the enlargement of left heart, and the age of surgery [15].

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