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Case report

Anomalous left coronary artery from the pulmonary artery: A rare case diagnosed in an adult

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ABSTRACT

ALCAPA syndrome (anomalous origin of left coronary artery from the pulmonary artery) is an extremely rare congenital cardiac anomaly associated with high mortality rate at young age. If undiagnosed and uncorrected, the affected individuals rarely survive beyond infancy. This article reports on a 45-year-old asymptomatic man with the ALCAPA diagnosed thanks to a detailed cardiac examination, that followed the finding of the left bundle branch block and a decreased left ventricular systolic ejection fraction. Although coronary angiography and computed tomography are considered to be the key diagnostic methods for ALCAPA, in this case the diagnosis was established based on echocardiography. Previously, the patient has been under regular review by a cardiologist with the incorrect diagnosis of multiple ventricular septal defects. In fact, this diagnosis resulted from a misinterpretation of the ultrasound image of the intercoronary connections. The presence of a separate diagonal artery originating from aorta renders this case report even more interesting.

Introduction

Anomalous origin of left coronary artery from the pulmonary artery (ALCAPA or ALCA), or Bland–White–Garland syndrome, is a very rare congenital heart defect (CHD). It was first described in 1866, in detail in 1933 by the above mentioned physicians. In the work of Samánek et al., the incidence was 1.34 in 100,000 live births (more exactly: 11 cases per 816,569 live births in 10 years in Czech republic; all infants underwent ultrasound examination, the autopsy was performed in deceased ones) [1]. Out of all CHDs, ALCAPA represented only 0.22%. Nevertheless, these data do not include patients with ALCAPA diagnosed in adulthood. The anomaly is usually isolated, exceptionally may be associated with other CHD,

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such as persistent ductus arteriosus, ventricular septal defect, tetralogy of Fallot or coarctation of the aorta. ALCAPA is associated with high risk of death – it is the most frequent cause of ischemic heart disease (IHD) within the first year of life and the mortality rate up to 90% [2].

Coronary arteries differentiate from splanchnopleuric cells near the sinus venosus. These cells transform into mesenchymal and further into endothelial and smooth muscle cells, which, in the process of vasculogenesis, develop into coronary arteries. Their endothelial cells connect to the aortic endothelium thus enabling the circulation of blood. There are two theories explaining the anomalous origin of left coronary artery (LCA). According to Abrikossoff, the anomaly may result from an abnormal septation of truncus and conus arteriosus; according to the newer version by Hackensellner, all six semilunar valve regions have the propensity to develop the coronary arteries. The anomalies are explained on the basis of their defective involution or persistence [3].

During the prenatal period, ALCAPA does not become evident due to distinct anatomical and hemodynamic conditions. The pressure in fetal pulmonary circulation is higher than in the aorta and the blood flowing through the right cardiac chambers is mixed with high oxygen content. High pulmonary artery pressure enables sufficient prograde flow through the left coronary artery, and thanks to the high oxygen level, an adequate oxygenation of heart muscle. After birth, the oxygen inflow into the pulmonary alveoli triggers normal ventilation, leading to the tension change in smooth muscle cells in the pulmonary circulation and a dramatic decrease in the pulmonary vascular resistance. The pressure in aorta increases until it exceeds the pressure in the pulmonary artery. Afterwards, the occlusion of prenatal shunts (ductus arteriosus and foramen ovale) occurs. Following the pressure decline in the pulmonary artery, the blood flow in ALCAPA reverses and becomes retrograde. Coronary collaterals (intercoronary connections or arterio-arterial fistulae) supply the myocardium in a reduced manner, but, on the other hand, intensify the myocardial steal phenomenon, leading to the left-to-right shunt and results in left ventricular overload. These changes lead to ischemia and heart failure in the affected individuals – with the corresponding electrocardiogram findings (ECG), clinical and echocardiographic signs of heart failure, mitral regurgitation, left-sided heart overload, decreased systolic function of left ventricle.

Two types of ALCAPA syndrome are discussed in the literature. The first type is described as the ''infant type'' and presents with congestive heart failure or myocardial infarction early after birth [2]. Left untreated, 90% of affected children die within the first year of life. These individuals lack sufficient coronary collaterals. The second type presents later, but rarely as late as in the adulthood, and differs from the infant type in the anatomical pattern. In adults, rich collateralization of coronary arteries can be found with a hyperplastic right coronary artery. Affected individuals are at particular risk of sudden cardiac death, mostly at exercise.

According to Angelini et al., 19% of sudden cardiac deaths of young athletes were secondary to a coronary artery anomaly [4]. Basso et al. retrospectively analyzed 12 young athletes who died of sudden cardiac death, all of them had negative ECG and exercise stress tests previously. Ten of them reported previous cardiac symptoms – syncope or chest pain. In nine of them, anomalous origin of left coronary artery was found at autopsy, in three individuals the anomalous origin of right coronary artery [5].

Surgical repair is the treatment of choice, aiming at restoration of supply by one or two coronary arteries. In infancy, direct re-implantation of the coronary artery into the aorta is preferred, if unattainable due to anatomical conditions, then Takeuchi repair (transpulmonary baffle) may be performed. The procedure involves derivation of anomalous left coronary artery ostium through an intrapulmonary tunnel created within the pulmonary artery wall, leading to the aortopulmonary window in the aortic wall. The pulmonary artery is reconstructed with pericardial patch. In adults, it is possible to perform a bypass graft surgery with ALCAPA ligation. A less preferred method where a one-coronary artery system is created, is the isolated ALCAPA ligation [6]. In the work of Neumann et al., out of 30 patients with ALCAPA, 19 patients received coronary transfer (first group), 9 underwent Takeuchi repair (second group) and 2 were treated by closure of the ALCAPA (third group). All patients underwent surgery in infancy. One patient who received ALCAPA ligation died perioperatively. The rest of patients survived over 10 years, patients with the coronary transfer had lower rate of reoperations (5.9%), higher after Takeuchi repair (28.9%). 8 years after the Takeuchi repair, 79.9% of these patients had at least moderate pulmonary regurgitation, in contrast to 0% after coronary transfer [7]. In the work of Naimo et al., out of 42 patients, 9 had concomitant ALCAPA and mitral valve surgery. The early mortality was 2.4%. The follow-up period was 14 years, survival at 20 years of age 98%. 8 patients after concomitant mitral valve repair and ALCAPA repair underwent mitral valve reoperation. 90% of patients had preserved systolic function of LV (left ventricle), 10% moderately lower [8]. However, the published works encompass only small populations of pediatric patients with ALCAPA, in adults there are only case reports available.

The complications of isolated ALCAPA ligation include recanalization of the affected artery, severe mitral regurgitation and risk of sudden cardiac death due to persisting ischemia. Bypass surgery with LCA ligation may be complicated by a stenosis or occlusion of the graft with a high probability of reoperation. Coronary transfer is associated with the lowest complication rate, nevertheless, bleeding, leak or kinking of the left coronary artery may occur in adults. The complications related to Takeuchi repair involve supravalvular pulmonary stenosis, aortic valve stenosis, baffle obstruction or leak.

Case presentation

A 45-year-old man has been under regular review by a local cardiologist from age 32 due to the incidental finding of left bundle branch block (LBBB) and a systolic heart murmur. According to the patient's medical records, multiple muscular ventricular septum defects were suspected. Furthermore, he has had a mild mitral regurgitation with normal systolic LV function for a long time. The patient has been totally asymptomatic, with otherwise negative medical history,

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