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Anomalous left coronary artery from the pulmonary artery (ALCAPA) – an adult who has remained asymptomatic (RCD code: I-1C.3)

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Abstract

A 21 year old woman with phenylketonuria, positive hepatitis B surface antigen and anomalous left coronary artery originating from the pulmonary artery (ALCAPA) was referred to our clinic to asses her current condition and to establish her further treatment. ALCAPA with concomitant mild mitral insufficiency were confirmed. She was asymptomatic and the studies provided evidence for not qualifying her to surgery. JRCD 2013; 1 (5): 25–27

Key words: anomalous left coronary artery from the pulmonary artery (ALCAPA), mitral insufficiency, phenylketonuria

A case report

A 21 year-old woman with phenylketonuria, positive hepatitis B surface antigen, ALCAPA with concomitant holosystolic murmur secondary to mitral insufficiency was admitted to our clinic from another center to assess her current condition and to establish her further treatment.

Her congenital anomaly was discovered by chance while being observed because of phenylketonuria. A holosystolic murmur at childhood of mild degree brought the doctors attention. Echocardiogram showed a mild mitral regurgitation with slight left ventricular hypertrophy. Hypertrophic cardiomyopathy was suspected but finally after coronary angiography the diagnosis of ALCAPA was made.

On admission she was asymptomatic. Her blood pressure was normal 110/70 mmHg with regular heart rate of 71 bpm. There was a holosystolic murmur a grade 3/6 heard along the left sternal border and the apex. Lungs were clear to auscultation. The abdomen was soft with normal bowel sounds. There was no hepatosplenomegaly and no peripheral edema. The electrocardiogram revealed sinus rhythm 71 bpm, with left axis deviation, a small progression of R wave in the precordial leads, small q in V2 lead and in aVL. The echocardiogram demonstrated a mild hypertrophy of left ventricle, with slightly hypokinetic anterior wall and ejection fraction of 60%. Mitral regurgitation was described as mild. The orifice of the left coronary artery was not identified at the aortic sinus. The pulmonary trunk and pulmonary arteries were not dilated (fig.1). Coronary angiography revealed the anomalous left coronary artery (LCA) originating from the pulmonary artery trunk and a single, large and tortuous right coronary artery (RCA) arising from the right sinus of Valsalva and giving off extensive collateral vessels coursing over the right ventricular wall, the interventricular septum, and the apex (fig.2). The diameter of RCA near the origin was 7.5 mm. There were no traces of coronary artery disease. MRI contrast study revealed subendocardial necrosis in the apex of left ventricle and in the antero-septal wall (fig.3,4). It did not exceed 50% of the wall thickness. The monitoring of the first-pass distribution of MR contrast medium in basal and vasodilated states showed the decreased myocardial perfusion in apical and antero-septal segments. The results of MRI and echocardiogram suggested that she might have had asymptomatic myocardial infarction in the past.

A patient was symptoms free which we confirmed in the exercise test (100 W, 3 min, no arrhythmia). The studies provided evidence for not qualifying her to surgery. The patient was discharged on medications and she was recommended a further follow up in our out-patients' department.

Conflict of interest: none declared.

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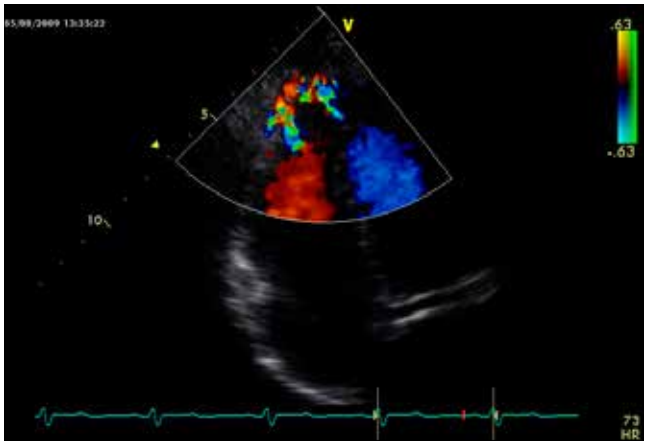

Figure 1. Echocardiography. Parasternal short-axis view. Colour Doppler flow demonstrating anomalous origin of the left coronary artery from the pulmonary artery. Retrograde drainage into the pulmonary artery under systemic pressure

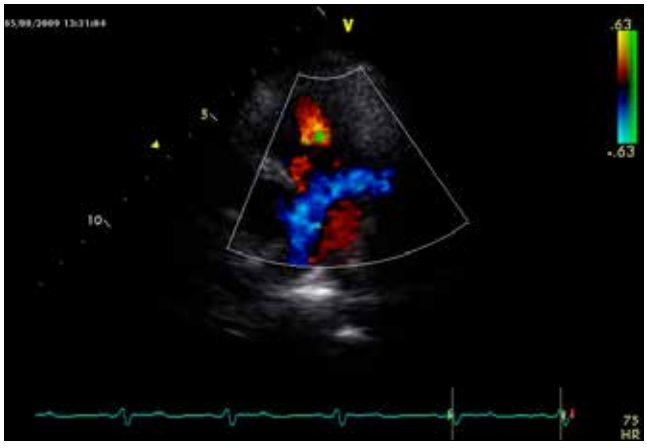

Figure 2. Echocardiography. Apical four-chamber view. Colour Doppler image documenting collaterals between the left and right coronary arteries in the apical region

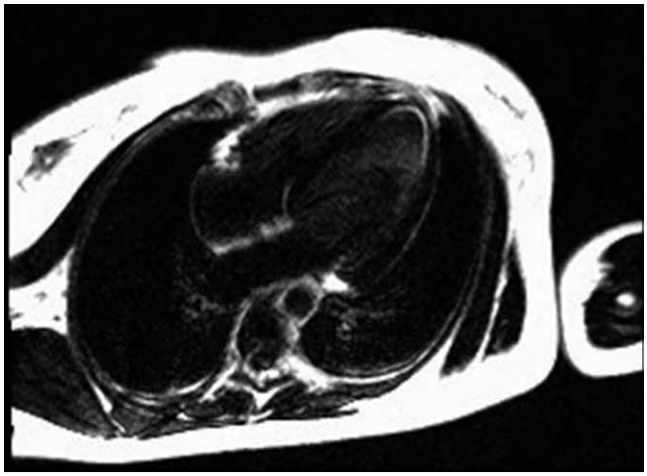

Figure 3. Cardiovascular magnetic resonance. Four-chamber delayed-enhacement image shows subendocardial necrosis of the apical region. It did not exceed 50% the wall thickness

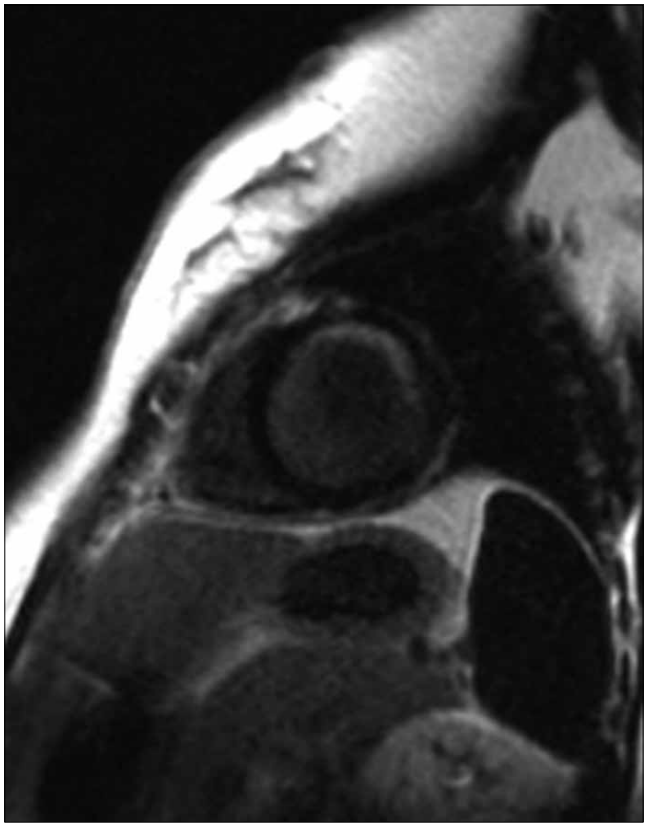

Figure 4. Cardiovascular magnetic resonance. Short axis delayed--enhacement image demonstrates late myocardial enhacement in the subendocardial antero-apical segment and indicates viability larger than 50% of myocardial thickness

Discussion

Anomalous coronary arteries are rare among congenital cardiac abnormalities. They are seen in approximately 0.3–1.0% of the population[1]. In 1885, Brooks et al. reported the first case of ALCAPA[2]. He hypothesized that blood was flowing into the pulmonary artery rather than away from it[3]. Anomalous origin of the left main coronary artery from the pulmonary artery also known as Bland-White-Garland syndrome is a rare congenital cardiac malformation[4] and accounts for about 0.25–0.5% of congenital heart defects[5]. ALCAPA is only one of several reported congenital coronary artery anomalies[6]. It is usually an isolated cardiac malformation but in rare cases, it was reported to coexist with ventricular septal defect, patent ductus arteriosus, tetralogy of Fallot and coarctation of the aorta[7]. Few patients survive childhood without surgical repair. The high number of sudden deaths was reported before 35 year-old[8]. Typical symptoms of ALCAPA range from chronic mitral insufficiency and angina, myocardial infarction, heart failure, malignant ventricular arrhythmias, sudden cardiac death, to ischemic cardiomyopathy[5]. We describe a case of 21 year-old patient who had a mild mitral insufficiency. Her congenital abnormality was discovered by chance while being observed by doctors dealing with phenylketonuria. It was suggested on echocardiography and confirmed

at cardiac catheterization where the left coronary artery (LCA) was shown to arise from the sinus of the left cusp of the pulmonary valve. The left descending artery gave many branches connecting it to the right coronary artery (RCA). The right coronary artery emerged normally from the right anterior aortic sinus and was dilated. Usually, collateral circulation between the right and left coronary systems ensues after birth and the left ventricular myocardium is chronically under perfused, since flow is preferentially directed into the pulmonary vessels because of the lower pulmonary vascular resistance (coronary steal phenomena). The presence or absence of these collaterals serves as the basis for the clinical classification of anomalous origin of the coronary arteries from the pulmonary artery. An infantile type can be differentiated, in which there are no or very few collaterals. An adult type is diagnosed when the collateral circulation is well developed. Patients with few collaterals have greater degree of myocardial ischemia and/or infarction. Moreover there is more severe mitral regurgitation and heart failure in this subgroup. MRI in ALCAPA shows evidence of chronic ischemia and severe interstitial fibrosis. Left untreated, the mortality rate in the first year of life is 90% secondary to myocardial ischemia or infarction and congestive heart failure[7,8]. In rare cases, the clinical presentation of myocardial ischemia may be delayed into early childhood, or even adulthood as in our patient, where collaterals from the RCA to the LCA can provide enough oxygenated blood to the myocardium.

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