ALCAPA syndrome in a 56-year-old woman with dyspnoea on exertion

Zespół ALCAPA u 56-letniej kobiety z dusznością wysiłkową

Pawel Uruski¹, Dawid Lipski¹, Olga Trojnarska², Robert Juszkat³, Andrzej Tykarski¹

¹Department of Hypertension, Angiology and Internal Medicine, Poznan University of Medical Sciences, Poznan, Poland
²1st Department of Cardiology, Poznan University of Medical Sciences, Poznan, Poland
³Department of Clinical Radiology, Poznan University of Medical Sciences, Poznan, Poland

We present the case of a 56-year-old woman suffering from shortness of breath on exertion (NYHA class II) worsening for the last couple of years. Blood pressure on admission was 140/71 mm Hg and heart rate (HR) was 72 bpm. Further physical examination and routine laboratory tests were irrelevant. The electrocardiogram showed normal sinus rhythm (SR) with HR 62 bpm, intraventricular conduction delay, a low progression of R wave in V₁–V₄ leads and premature ventricular contractions. 24-hour Holter electrocardiogram monitoring was carried out. Normal SR with a mean HR of 67 bpm was recorded. There were 967 premature ventricular complexes (PVCs), including one run of three PVCs, two pairs of bigeminies and nine trigeminies. Moderate mitral regurgitation, akinesis of the apex, posterior and inferior walls and asynchrony of the septum contraction were diagnosed with echocardiography. The ejection fraction was 45%. Colour Doppler visualised the collaterals connecting the right coronary artery (RCA) and the left coronary artery (LCA). The last one appeared to conjoin with the pulmonary trunk. Coronarography confirmed the presence of wide RCA which connected with the branches of LCA. The origin of LCA could not be visualised in the ascending aorta. The coronary arteries anomaly was confirmed in angio-computed tomography (CT), which showed that left main coronary artery arises from the left-inferior part of the pulmonary trunk (Fig. 1). The left anterior descending artery and left circumflexus were patent, and sinuous with a diameter of 8 mm. RCA was dominant, very sinuous, and originating from the typical place. The junctions between distal branches of RCA and LCA were observed (Fig. 2). Magnetic resonance imaging of the heart revealed dilation of the left ventricle which was generally hypokinetic. Late gadolinium enhancement was not observed in the left ventricle muscle. A diagnosis of ALCAPA syndrome, anomalous origin of the LCA from the pulmonary artery, also called Bland-White-Garland syndrome, was established. This rare congenital anomaly appears in 1 in 300,000 live births and accounts for less than 0.5% of all congenital heart anomalies. In most cases, diagnosis and treatment is carried out in the first year of life. It is a very rare situation when ALCAPA is diagnosed in an adult because the natural history of this malformation mostly leads to death in childhood. There is a consensus that children with ALCAPA will benefit from a replantation of LCA to the ascending aorta. This procedure is technically impossible in most adults. Thus LCA bypass grafting with subsequent ligation of its anomalous origin should be performed. Another surgical method is the Takeuchi procedure where an intrapulmonary tunnel joining LCA and the left aortic sinus is created. Even asymptomatic patients should undergo surgery to prevent sudden cardiac death. Our patient was fully informed about the possible options but declined the surgical therapy. She was prescribed the following medications: carvedilol, perindopril, spironolactone, indapamide, rosuvastatin and acetylsalicylic acid, and returned home.

Address for correspondence:
Dawid Lipski, MD, Department of Hypertension, Angiology and Internal Medicine, Poznan University of Medical Sciences, ul. Długa 1/2, 61–848 Poznań, Poland, e-mail: dlipski@ump.edu.pl

Conflict of interest: none declared