Aortic interruption in an adult female

Przerwanie aorty u dorosłej kobiety

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A 58-year-old woman complaining of exertional dyspnoea, fatigue and palpitations was admitted to the cardiology service as an outpatient. She had a history of hypertension for the last 12 years. Her blood pressure was measured as 148/78 mm Hg in the left arm and 145/80 mm Hg in the right arm; heart rate was irregular about 135 bpm; 2/6 grade systolic murmur was auscultated at the apex of the heart. Also inspiratory rales at the lower parts of both lungs were remarkable on chest auscultation. Peripheral pulses were palpable on both upper limbs, whereas pulses were diminished on lower limbs compared to upper limbs. Rhythm was atrial fibrillation on initial ECG recording. After clinical stabilisation via medical therapy with ACE inhibitor, beta-blocker and diuretics, spontaneous conversion to sinus rhythm was observed in the patient. Transthoracic echocardiography documented mitral regurgitation and left ventricular dysfunction (LVEF 35%). Descending aorta could not be evaluated because of poor echo window. On a subsequent transoesophageal echocardiography (TEE) examination, interruption of descending aorta distal to the origin of the left subclavian artery was revealed. On colour Doppler examination, there was no blood flow between the aortic arch and the descending aorta (Fig. 1). A thoracic computed tomography angiogram (CTA) also revealed interruption of aorta distal to the origin of left subclavian artery with severely enlarged internal mammarian arteries and intercostal arteries bilaterally. This indicated the compensatory development of collateral circulatory bypass vasculature perfusing the descending thoracic aorta (Fig. 2A, B). Coronary angiography as well as aortography was performed via the right radial artery. Coronary angiography showed normal coronary arteries. After contrast injection into the ascending aorta, interruption of thoracic aorta at the previously defined localisation was revealed (Fig. 3). The patient was referred for surgical intervention for the aortic interruption. Interrupted aortic arch (IAA) is defined as loss of luminal continuity between the ascending and descending aorta. It is a rare congenital anomaly that is detected in three per million live births. Based on the site of interruption, it is classified into three groups. Group A is where interruption occurs distal to the left subclavian artery (as in our case); Group B is where interruption occurs distal to the origin of left common carotid artery; and Group C is where interruption occurs proximal to the origin of left common carotid artery. The vast majority of patients today are diagnosed in the neonatal or prenatal period. Surgical repair of IAA is well compatible with long term survival. There are a few reported cases in the medical literature of successful survival until adulthood without surgical treatment, as in our case. In conclusion, it should be kept in mind that aortic interruption can be seen in adulthood and may present different clinical symptoms, as in our case.

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