Coarctation evolving into complete aortic isthmus obstruction

Proces ewolucji koarktacji w pełną niedrożność cieśni aorty

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A 52-year-old hypertensive man, an active paramedic until age of 45 was referred due to exercise intolerance, fatigue, dyspnea, and exertional chest pain. In his 30’s he was diagnosed with aortic coarctation but refused surgical or interventional correction, known with peak systolic gradient of 40 mm Hg (recorded 7 years earlier). First symptoms of heart failure appeared in his 40’s and on admission he was in the New York Heart Association (NYHA) class III with atrial fibrillation of unknown duration.

Admission physical examination revealed no murmurs characteristic for aortic coarctation; there was a 2/6 systolic murmur over cardiac apex. Brachial artery pressure was 130/80 mm Hg, popliteal artery pressure — 100/60 mm Hg. Echocardiography demonstrated severely dysfunctional left ventricle with ejection fraction 21% and mild functional mitral regurgitation (Fig. 1A) and significant left atrial enlargement. Ascending aorta was abnormal with tortuous course and irregular wall contour whereas no accelerated flow was detected in the isthmus region. Descending aortic flow showed diastolic runoff pattern. Cardiac catheterization via femoral access was impossible due to an inability to cross the aortic arch — aortography revealed aortic closure distally to the origin of the left subclavian artery (Fig. 1B). Descending aorta was supplied with the large collateral artery originating from the left subclavian artery. Angiography performed from the right radial artery revealed a huge conglomerate of abnormal vessels originated from the brachiocephalic trunk and directing the blood to descending aorta and lower part of thorax. Coronary arteries were normal. Right heart catheterization disclosed pulmonary hypertension (systolic pulmonary artery pressure 109 mm Hg) with moderately elevated (3.5 WU) but still reversible pulmonary vascular resistance.

Contact enhanced, 64-slice computed tomography confirmed the aortic arch interruption (IAA) distal to the origin of the left subclavian artery (Fig. 2) and abundant lower body collaterals from the extremely dilated brachiocephalic trunk, left subclavian artery and their branches, and vertebrospinal arteries. Aortic coarctation occurs in 0.2–0.6% of all live births and accounts for 5% to 8% of all cases of congenital heart disease [1] but IAA is a rare malformation that occurs in 3 per million live births [2, 3] an as our case demonstrates may develop as a consequence of untreated severe aortic coarctation [4].

Discussion

Therapeutic decisions in the presented case are difficult. Small difference between brachial and lower extremity pressures and absence of hypertension suggest effective low-resistance collaterals. Surgical repair of interrupted aorta (with graft implantation) could be complicated by hemorrhage due to extensive net of collaterals and altered vascular anatomy. Another surgical option to be considered because of severe left ventricular dysfunction with end-stage heart failure is orthotopic cardiac transplantation. Our patient had cardiac resynchronization therapy with defibrillator (CRT-D) device implanted but denies other treatment options despite progressive heart failure.

References


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