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CASE REPORT

Percutaneous treatment of native aortic coarctation in adults

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Abstract Aortic coarctation is a common congenital cardiac defect, which can be diagnosed over a wide range of ages and with varying degrees of severity. We present two cases of patients diagnosed with aortic coarctation in adulthood. Both patients were treated by an endovascular approach. These cases demonstrate the variety of indications in which percutaneous treatment is an excellent alternative for surgical treatment in adult native coarctation patients.

Keywords Native aortic coarctation in adults · Endovascular repair

Introduction

Aortic coarctation is a congenital cardiovascular malformation comprising a circumscript narrowing of the aortic lumen usually located distally of the left subclavian artery, at the site where the ductus arteriosus enters the aorta. Aortic coarctation accounts for 5–8% of all congenital heart defects and may be associated with other left-sided cardiac abnormalities, such as bicuspid aortic valve, hypoplastic aortic arch, and ventricular septal defect. When diagnosed in neonates, surgical reconstruction is the treatment of choice. In infants, surgery is preferred as well, but balloon angioplasty has also evolved rapidly over the past decade. Both treatment options carry the risk of restenosis and late aneurysm formation [1, 2].For recoarctation after initial surgical treatment, good long-term results have been reported [3, 4].

A small proportion of aortic coarctation is not diagnosed until adolescence or adult age. The presenting symptoms usually include hypertension, congestive heart failure, or intermittent claudication. There is an indication for treatment if the gradient is >20 mmHg in combination with hypertension, either at rest or during exercise [5]. When intervention is warranted, the risk of surgery is considerable because of the extensive collateral network surrounding the coarctation which results in a high risk of bleeding complications. As a consequence, balloon angioplasty with or without stenting has evolved as a treatment modality for adolescents with native aortic coarctation [6, 7].

Case A. Native aortic coarctation

The first case involves a male patient referred to our hospital, who had been diagnosed with severe aortic coarctation at the age of 24 in the Middle East. At the time of diagnosis, the gradient measured by catheterisation was 90 mmHg. The coarctation was left untreated despite the presence of hypertension. He presented to our hospital at the age of 33 with fatigue, shortness of breath on exercise, complaints of headache, and intermittent chest pain both at

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rest and during exercise. His blood pressure was 160/100 mmHg. On auscultation, normal heart sounds were heard, as well as an ejection murmur grade II/ VI at the right second intercostal space and a late-systolic murmur grade III/VI at the apex. Palpation of the femoral arteries revealed slightly decreased pulsations. The ECG showed a left anterior hemiblock. Echocardiography showed mild concentric left ventricular hypertrophy without dilatation. A tricuspid aortic valve was present with mild aortic regurgitation. The aortic root was dilated with a diameter of 41 mm. The diameter of the ascending aorta was 32 mm. Cardiac magnetic resonance imaging (MRI) confirmed a complete interruption of the descending aorta directly after the dilated left subclavian artery. Distal from the occlusion, the aorta was supported by many collaterals originating from the left mammary artery and the intercostal arteries (Fig. 1a and b). It was decided to treat this patient with a percutaneous balloon-expandable 28 mm covered stent. Retrograde passage proved impossible, but using the radial approach balloon passage was successful. After stent implantation with deployment to 18 mm, angiography revealed a well-expanded stent and there was no residual pressure gradient (Fig. 2a and b) (Tape a and b). Two months later, the patient was without complaints and follow-up angiography showed good stent position, no aortic aneurysms and there was no pressure gradient.

Case B. Native aortic coarctation

The second case concerns a 43-year-old man who had recently been diagnosed with severe aortic coarctation and a hypoplastic aortic arch, accompanied by a bicuspid aortic valve and moderate aortic valve regurgitation. In addition, he had a dilated left ventricle with impaired systolic function. He admitted severe alcohol and cannabis abuse over the past several years.

He presented with increasing fatigue but no exertional dyspnoea or orthopnoea. Physical examination revealed an important blood pressure difference between the right and left arm (125/77 versus 99/72 mmHg, respectively). On auscultation normal heart sounds with a grade II/IV crescendo/decrescendo murmur and a short diastolic murmur were heard. Palpation revealed a palpable liver 3 cm below the diaphragm, and weak peripheral pulsations. The electrocardiogram showed mild intra-ventricular conduction delay (QRS 120 ms), and left ventricular hypertrophy with secondary repolarisation abnormalities. Echocardiography showed a dilated, hypertrophied left ventricle with a moderately impaired function (Fig. 3a). Diffuse left ventricular wall motion disturbances with hypokinesis and akinesis of the interventricular septum were seen. The aortic valve was bicuspid with a maximal gradient of 26 mmHg and mild aortic regurgitation. The aortic root and ascending aortic diameters were 39 mm and 33 mm, respectively. Turbulent flow was seen in the descending aorta with a maximum velocity of 3.5 m/s and a typical saw-tooth shape (Fig. 3b). This patient was treated with balloon dilatation and implantation of a 22 mm covered stent. Before the procedure, mean systolic blood pressures were 70 mmHg proximal to the coarctation and 45 mmHg distal of the coarctation. Stent implantation was successful and resulted in pressure equilibration.

Discussion

Coarctation of the aorta can be diagnosed across a wide age spectrum and with a range of symptoms. An untreated

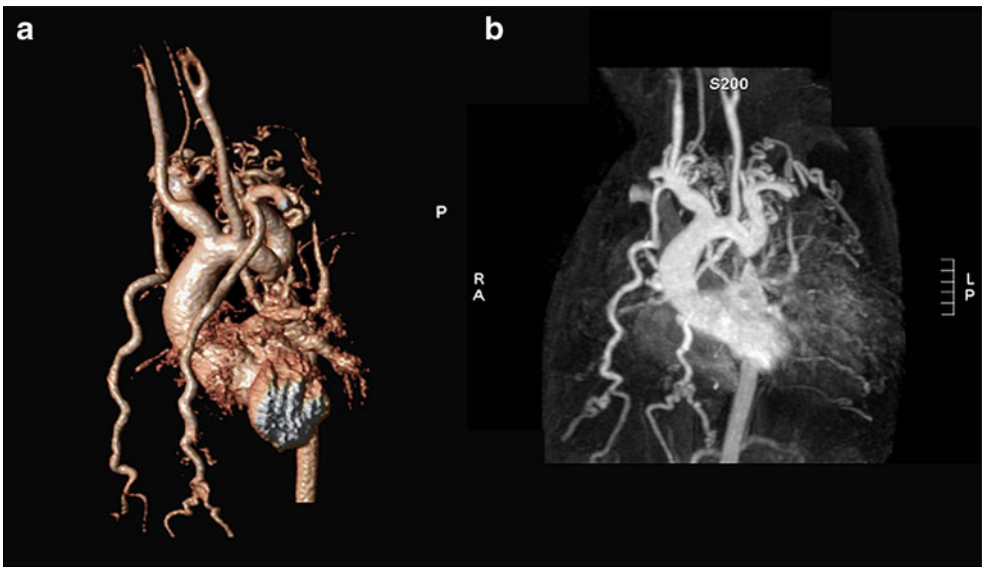

Fig. 1 Magnetic resonance imaging in native aortic coarctation. **a**. MRI reconstruction image demonstrating the many collateral arteries **b**. Late gadolinium MRI image demonstrating the severe coarctation accompanied by the collateral arteries

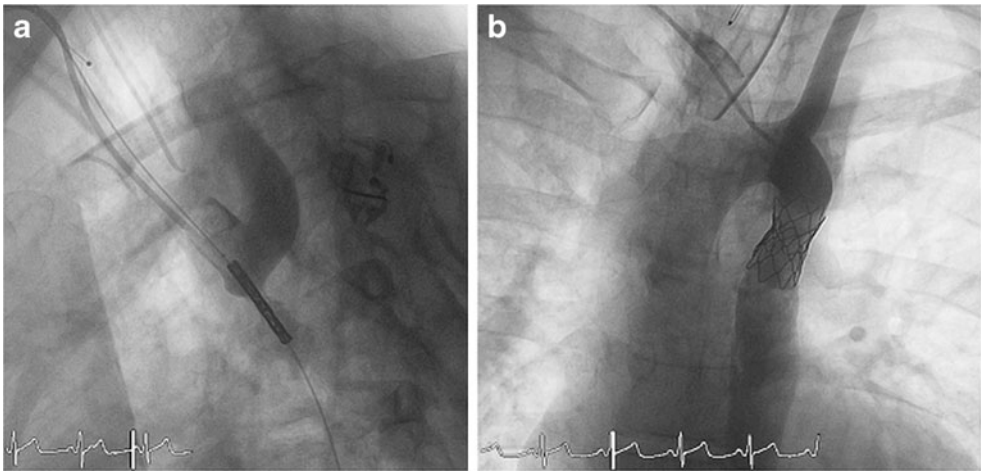

Fig. 2 Percutaneus stent-implantation for aortic coarctation **a**. Angiographic image before stent expansion in case A **b**. Angiographic image after stent expansion in case A

coarctation of the aorta presenting later in life has a negative impact on long-term survival, as compared with patients diagnosed and treated in early life [8]. Late complications such as aortic aneurysm formation, infective endocarditis, hypertension, premature (coronary) atherosclerosis and cerebrovascular accidents all account for the increased morbidity and mortality [9, 10]. The two patients described above were both diagnosed in adulthood. Traditionally, surgical repair has been an effective treatment option for adult patients with native coarctation, and it has been shown to improve the efficacy of postoperative antihypertensive treatment. Adult patients with native coarctation are usually at lower operative risk than re-coarctation patients because there are no concerns related to pleural or peri-aortic scar tissue associated with previous surgical repair. However, these patients do have many associated cardiovascular comorbidities including left ventricular hypertrophy and arrhythmias [8]. The first case presented with a severe aortic coarctation, with many collaterals due to the severe obstruction. In the second case the severe aortic coarctation was accompanied by a hypoplastic arch and an impaired left ventricular function possibly due to long-lasting alcohol abuse. In both cases, the risk of cardiac surgery was estimated to be too high. In case 1, the extensive collateral network surrounding the coarctation caused a substantial risk of peri-operative bleeding complications. In case 2 surgical risk was increased because of the impaired left ventricular function. In both cases endovascular repair with balloon angioplasty and stent placement was performed. Endovascular repair with or without stenting has been demonstrated to be an acceptable alternative to surgical repair with similar outcome in native coarctation. The use of covered stents has been promoted to avoid vascular complications [8]. The incidence of recoarctation after stent placement is lower than after balloon angioplasty alone, due to a lower degree of elastic recoil and avoidance of vessel overdilation and, as a consequence, reduced aortic injury. Concerns that arise in coarctation patients after endovascular repair are strut fractures, metal fatigue or aortic deterioration or aortic disruption at the coarctation site [11]. However, studies documenting the long-term outcomes of stent implantation confirmed the low rate of procedure-related adverse events and a long-term procedural success in both native and recurrent aortic coarctation or aneurysmal disease [12, 13]. Long-term outcome was comparable with other surgical and interventional modalities. This case report demonstrates the variety of indications in which percutaneous treatment

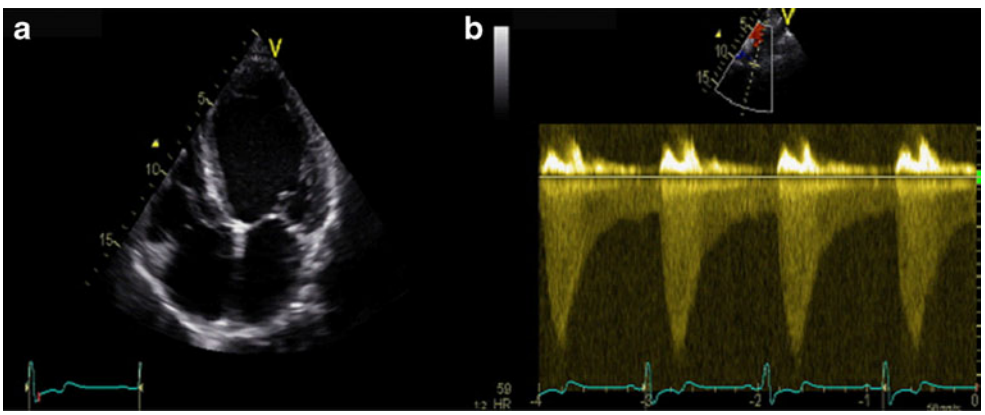

Fig. 3 Transthoracic echocardiography in case B **a**. Apical four-chamber view demonstrating the dilated left ventricle **b**. The typical sawtooth pattern with continuous flow on continuous wave Doppler interrogation in aortic coarctation

is preferable to surgical treatment in adult patients with native aortic coarctation. More research on the long-term assessment of adult native coarctation patients after percutaneous intervention will be important to determine the impact on survival in these patients.

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