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Stenting for infantile adult aortic coarctation with successful conception of zygomatic twins at 4 years' post-intervention

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ABSTRACT

INTRODUCTION: Aortic coarctation is a congenital defect which rarely presents in adulthood but results in significant morbidity and mortality. Endovascular techniques present novel therapeutic options for managing this anomaly with comparable results to traditional open surgical repair.
PRESENTATION OF CASE: We convey a case of postductal aortic coarctation in a symptomatic 31-year-old woman, undetected, despite 27 years history of congenital bicuspid valve with aortic incompetence. We staged a successful endovascular repair following presentation with hypertension, abdominal pain, and worsening lower limb claudication. This lady recovered well, eventually giving birth to twins after a successful caesarean section delivery. The anatomical, clinical and technical features of the case are appraised.
DISCUSSION: Aortic coarctation is classified anatomically as preductal (infantile) or postductal (adult) and the morphological spectrum of abnormality ranges from a discrete stenosis distal to the left subclavian to a hypoplastic transverse arch and aortic isthmus presenting in infancy. Its incidence is 0.2–0.6 per 100 live births. Traditional open surgical repair of aortic coarctation is associated with high morbidity and mortality. Advances in endovascular techniques with their associated shorter hospital time and lower costs, reduced short term morbidity and mortality mean that they are likely to become mainstays of treatment.
CONCLUSION: The presentation of aortic coarctation for the first time in an adult is extremely unusual. Error and delayed diagnosis can never be fully exempted in the practice of medicine even with the most advanced diagnostic tools. Follow-up is mandatory by trans-oesophageal echo in all endovascular patients.

1. Introduction

Aortic coarctation is a congenital anomaly that rarely presents in adults but results in significant morbidity due to secondary hypertension, intracranial haemorrhage, aortic rupture or dissection, and congestive cardiac failure. This condition represents 5–10% of all congenital cardiac lesions. The aetiology of congenital aortic coarctation is currently unknown; however secondary coarctation has been reported in cases of chest trauma, aortic arthrosclerosis and Takayasu's arteritis. The typical presentation of a patient with the defect is dependent on the severity of obstruction and associated cardiac lesions. In infancy patients can present with symptoms of congestive cardiac failure and severe acidosis. Adults typically present with early onset hypertension, headache, epistaxis and decreased lower limb perfusion.

2. Case report

Mrs. CM initially presented to the paediatric cardiology services at the 3 years of age. At this time her presenting symptoms were of fatigue with brief cyanotic episodes on crying and when distressed. Examination at that time was significant for a grade 3/6 diastolic murmur heard in the second, third and fourth left intercostal spaces and was loudest over the interscapular region posteriorly. On further examination she had normal carotid and femoral pulses without delay, together with a normal jugular venous pulse and in the absence of any stigmata of cyanotic congenital cardiac disease. Chest X-ray at that time was reported as normal. Her preliminary diagnosis was aortic valve incompetence without cardiac compromise. A follow-up chest X-ray at the 8 years of age demonstrated a dilated ascending aorta but normal pulmonary vasculature and lung fields. Fifteen years later, aged 27 years, CM underwent a 24 h blood pressure monitor due to underlying headaches which reported a mean blood pressure of 143/90 during the day and

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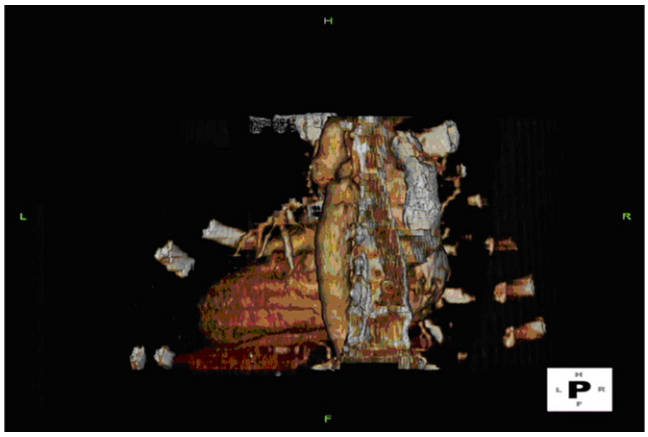

Fig. 1. Pre-operative CT angiogram displaying a postductal aortic coarctation with a true lumen measuring 2 mm approximately 1 cm distal to the origin of the left subclavian.

125/73 at night. An echocardiogram was obtained which confirmed a bicuspid aortic valve with a mean gradient of 21 mmHg and mild to moderate aortic regurgitation. The decision by the cardiologist at that time was for yearly follow-up with consideration for an eventual aortic valve replacement. Over subsequent years CM remained largely asymptomatic however she reported occasional fatigue. Repeat echocardiogram at the age of 29 was unchanged from previously with a peak gradient across the bicuspid aortic valve of 15 mmHg and with normal systolic and diastolic function.

Two years later CM complained of bilateral leg pain which was worse on mobilising. She had a radial femoral delay and a grade 4 early diastolic heart murmur. She had a 10 mmHg difference in brachial pressure between the right and left arms. Consequently, a lower limb arterial duplex demonstrated an abdominal aorta measuring 1.4 cm (anteroposterior) × 1.5 cm (transverse) with turbulent flow throughout its course but with no atheromatous lesion visualised. The iliac, superficial and deep femoral and tibial vessels all appeared widely patent but with reduced flow. Exercise ankle brachial pressure indices however were reduced bilaterally to 0.61 on the right and 0.54 on the left. CT angiogram was performed and demonstrated coarctation of the aorta with 2 mm true lumen beyond the origin of the left subclavian artery and just proximal to the main left pulmonary artery (Fig. 1). The CTA confirmed normal patency of the abdominal aorta and peripheral vessels. A trans-oesophageal echo was obtained and revealed the coarctation with a gradient of 100 mmHg. Cardiac function was normal with an ejection fraction of 65%.

3. Technique

Following multidisciplinary discussion it was determined that endovascular intervention would be the most appropriate means of treatment. In January 2007 an 18 mm × 40 mm Wallstent-Uni Endoprosthesis (Boston Scientific, Natick, MA, U.S.A.) was inserted endovascularly via a right femoral cutdown. An arterial line in the right upper limb was placed in order to monitor the arterial pressure and Heparin was administered. The aortic lumen was gradually dilated using Cordis Powerflex Extreme balloons starting from 6 mm × 4 cm to 8 mm × 4 cm to 10 mm × 4 cm over 60 min (Cordis Corporation, Bridgewater, NJ, U.S.A.). Further sequential dilatation was carried out using Atlas balloons from 12 mm × 6 cm to 14 mm × 6 cm (Bard Peripheral Vascular, Inc., Tempe, AZ, U.S.A.) The pressure gradient between the upper and lower limb decreased accordingly and the 18 mm × 40 mm Wallstent was deployed (Fig. 2). Completion angiogram was satisfactory. The groin incision was closed in layers.

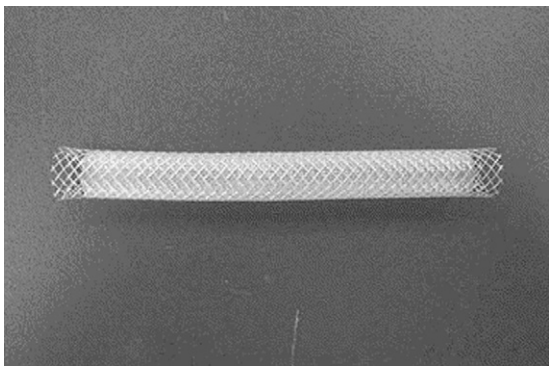

Fig. 2. 18 mm Wallstent (Boston Scientific, Natick, MA, U.S.A.) inserted endovascularly via femoral artery cut down and dilated in the coarcted segment from 4 mm to 16 mm over 70 min.

4. Post-operative

Post-operatively the patient was placed on 300 mg of aspirin and repeat ABPIs had improved to 0.92 on the right and 0.82 on the left. There were no post-operative complications. At 3, 6, and 9 months post-operatively, the patient was doing well clinically with improved control of her blood pressure (120/70 mmHg bilaterally) on Atenolol and complete resolution of her abdominal pain and lower limb claudication. A CT reconstruction post-operatively was imported to TeraRecon workstation showing a 16 mm channel of blood flow at the site of the coarctation (Fig. 3).

5. Follow-up

Two years following her initial intervention CM represented with new onset claudication. A trans-oesophageal echo was performed and showed peak gradient of 38 mmHg. A CT angiogram demonstrated restenosis at the level of the coarctation with the wall stent squeezed and migrated distally by 15 mm. Further intervention was undertaken to fully dilate the coarcted segment. Under general anaesthesia, a right groin was cut down, and cannulation of both the femoral artery and the vein was carried out. Two Cook (Cook Medical Inc., Bloomington, IN, U.S.A.) 18Fr sheaths were introduced respectively. An atlas 22 mm balloon (Bard Peripheral Vascular, Inc., Tempe, AZ, U.S.A.) was placed in the inferior vena cava to impede the venous return and an expanded polytetrafluoroethylene (PTFE). A 4.5 cm long Cheatham-platinum (CP)-covered stent covered stent (NuMED, Inc., Hopkinton, NY, U.S.A.) was mounted on a BIB stent placement catheter which had an outer

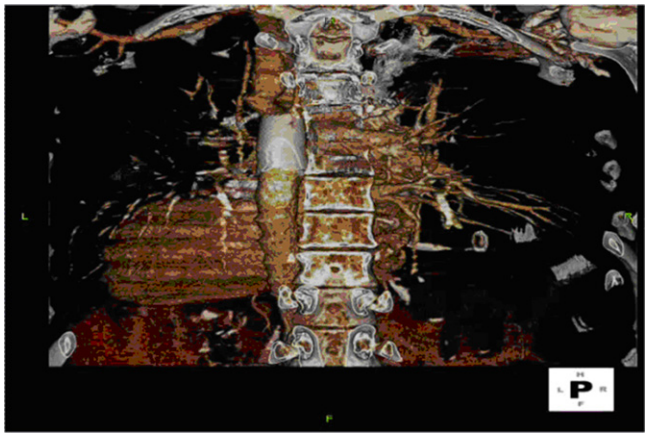

Fig. 3. Post-operative CT angiogram at 6-month post-intervention displaying a significant improvement in aortic calibre to 16 mm.

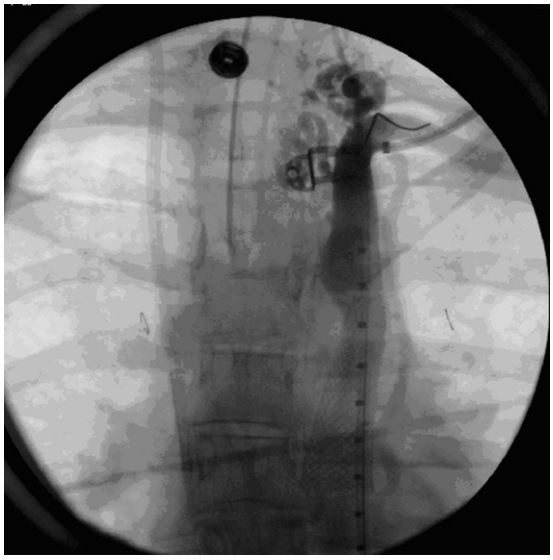

Fig. 4. Completion angiogram displaying good flow through the aorta post-insertion of CP stent.

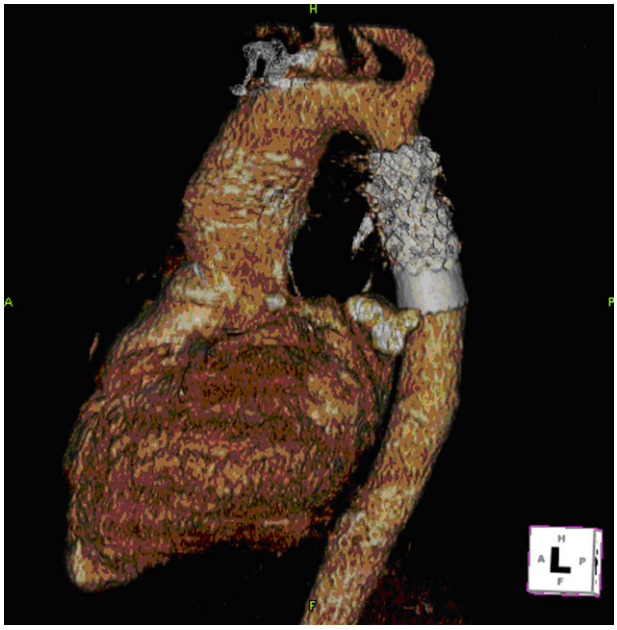

Fig. 5. CT angiogram at 4-year post-intervention showing normal flow though CP stent without graft migration.

balloon measuring 22 mm × 4.5 cm and an inner balloon measuring 11 mm × 3.5 cm (NuMED, Inc., Hopkinton, NY, U.S.A.). The mounted stent was inserted into the coarcted segment and deployed just distal to the left subclavian artery. No post stent dilatation was required. Groin was closed and patient was discharged 24 h later, with 75 mg of Nuseal aspirin. The completion angiogram confirmed an excellent position of the second stent across the site of coarctation and repeat scans at 6 weeks, 6 months and 1 year demonstrated continued patency (Figs. 4 and 5). Furthermore, exercise ABIs had improved significantly to 1.05 on right and 1.02 on left. In 2010 CM became pregnant with twins and due to concerns regarding the risk of aortic dissection arising due to shear stress consequent to a normal vaginal delivery it was decided that elective caesarean section would be the safest method of delivery. This was successfully undertaken later that year enabling a full term gestation and delivery with no adverse consequences.

6. Discussion

Aortic coarctation is classified anatomically as preductal (infantile) or postductal (adult) and the morphological spectrum of abnormality ranges from a discrete stenosis distal to the left subclavian to a hypoplastic transverse arch and aortic isthmus presenting in infancy. Its incidence is 0.2–0.6 per 100 live births, accounting for 5–8% of all cases of congenital cardiac disease.[1,2] Previous autopsy studies suggest that the mortality rate in patients in whom coarctation of the aorta if not surgically corrected is 90% by the age of 50 years with a mean survival of 35 years.[3]

Traditional open surgical repair of aortic coarctation involves resection with end-to-end anastomosis, prosthetic patch aortoplasty, subclavian flap aortoplasty or resection with graft interposition. Significant complications associated with surgical repair, however, include aneurysm formation, paradoxical hypertension, paraplegia, injury to the phrenic or recurrent laryngeal nerves, pleural effusions and in the long-term recoarctation.

It is well recognised that the risk of paraplegia resulting from ischaemic spinal cord injury increases with age[4] and is postulated to be due to the lack of the abundant collateral circulation which is present in children. The incidence of paraplegia following open surgical repair of coarctation has been estimated as 0.5–1% and has been closely related to clamp time.[5] Additionally, in the largest study to date assessing long-term morbidity and mortality in patients undergoing surgical repair of aortic coarctation and spanning a 35-year period there was found to be a peak incidence of perioperative mortality in adult patients of approximately 4.5%.[6]

It is with this background that consideration has been given of late to an increasing role for endovascular interventions in the operative management of aortic coarctation. Balloon angioplasty has been used for the last 20 years, however controversy persists regarding its role in the management of native aortic coarctation.[7] Concerns continue regarding the incidence of aneurysm formation, calculated as between 5% and 20%. In a study, by Fawzy et al., the long-term outcome of balloon angioplasty for aortic coarctation in adolescents and adults was found to result in a significant reduction in peak aortic coarctation gradient from 66 ± 23 mmHg to 10.8 ± 7 mmHg in 93% of patients with successful outcomes in the remaining 7% following repeat angioplasty.[8] Additionally, blood pressure normalised without medication in 63% of the study group. Secondary aneurysm formation was seen in 7.5% of patients and there were no incidences of recoarctation on long-term follow-up. Another retrospective study by Phadke et al., examining balloon angioplasty in thirteen cases of adult aortic coarctation reported favourable results with significant reductions of preoperative gradient in all patients treated, together with significant improvements in upper limb blood pressure with only four patients requiring continuing antihypertensive treatment postoperatively.[9]

Despite the apparent successes of balloon angioplasty concerns remain regarding the risk of restenosis. Restenosis rates are particularly high in neonates. Rao et al. reported restenosis in up to 70% of neonates and approximately 40% in infants.[10] The benefits, however, of angioplasty in young children include symptomatic relief with the avoidance of thoracotomy. Recoarctation can also then be treated at a later date by further balloon angioplasty or surgery when necessary. In adults, although the restenosis rates are less in children, they still remain significantly higher than for open surgery.[11]

As such, endovascular stenting has been proposed as a technique that might improve upon balloon angioplasty, potentially reducing the incidence of aortic dissection and restenosis in the long term. The rationale for stent implantation is that over-dilatation of the coarctation segment is unnecessary thereby avoiding major transmural tears while simultaneously providing a scaffolding by which

smaller tears are contained preventing progression to dissection or aneurysm formation. Many of the studies currently available examining the use of endovascular stenting in aortic coarctation are of small sample size and there is a paucity of data from long-term follow-up. Excellent short to medium term results, however, were observed in a cohort of 33 patients examined by Handon et al.[12] This study consisted of cohorts undergoing angioplasty and stenting for both native coarctation and recoarctation following surgery and demonstrated successful outcomes in 32 of the patients examined with no further evidence of recoarctation, aneurysm formation, stent displacement or fracture.

Handon et al. also demonstrated some of the difficulties inherent to stent placement in coarcted segments of the thoracic aorta with three of their patients having inadequately positioned stents requiring implantation of a second optimally positioned stent similar to the case that we have illustrated. Self-expanding Nitinol stenting was selected for our patient to prevent elastic recoil and reduce the incidence of aneurysm formation by limiting the amount of dilatation required and thus reducing the degree of traumatic injury to the aortic wall. In many cases, there is near-complete resolution of the coarctation gradient as was the case with our patient initially. Recurrent stenosis in our case may be due to intimal hyperplasia and its outgrowth through the interstices' of the open cell design Wall stent and not enough stent radial force. We used the Cheatham platinum (CP) stent (NuMED Inc., Hopkinton, NY, U.S.A.) which is formed of an expanded PTFE membrane attached to the outer side of the platinum/iridium wire, in a zig pattern and designed for the treatment of congenital vascular lesions.[13] This has the theoretical advantage of the atraumatic edges of the CP stent. The use of a balloon mounted system avoided excessive flaring during implantation and the PTFE cover may help diminish the incidence of dissection, aneurysm formation and myointimal hyperplasia.[14,15]

The incidence of aneurysm formation may be as high as 13% with bare metal stents.[15] The first covered stent was introduced in 1999 and since then numerous different subtypes have emerged including self-expanding and balloon expanding stents. Currently, the Cheatam-Platinum (CP) covered stents (NuMED, Inc., Hopkinton, NY, U.S.A.) which were introduced in 2001, are considered the stent of choice for primary stenting of aortic coarctation in the adult. These stents can also be used to manage previous bare stent complications such as aneurysms, stent fractures and intra-stent thrombosis or in the acute setting of aortic dissection or aneurysm formation following balloon dilatation.

It is currently well documented in the literature that women have increased rates of pregnancy related risks post aortic coarctation repair. A study by Vriend et al., analysed 54 women who underwent 126 pregnancies.[16] They reported 22 miscarriages, 6 abortions and 2 neonatal deaths within their cohort. Furthermore they display that 21 pregnancies were complicated by maternal hypertensive disorders with 5 women suffering severe pre-eclampsia. Our patient underwent an uncomplicated pregnancy to full term; however, it was decided at a multidisciplinary team meeting that an elective caesarean section would be most appropriate to prevent the risk of aortic dissection arising due to shear stress consequent to a normal vaginal delivery.

7. Conclusion

The presentation of aortic coarctation for the first time in an adult is unusual. Error and delayed diagnosis can never be fully exempted in the practice of medicine even with the most advanced diagnostic tools and best expertise. Follow-up is mandatory by trans-oesophageal echo in all endovascular patients. CT scan is indicated in symptomatic patients.

Conflict of interest

None.

Funding

None.

Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contributions

PSW wrote the manuscript; DPM captured the images; WT reviewed the manuscript; NH contributed towards study design; SS reviewed the final draft of the manuscript.

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