

Middle aortic syndrome in a teenager

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Received: 26 March 2014, Revised: 21 April 2014, Accepted: 30 April 2014

SUMMARY: Eyileten Z, Taşar M, Yazıcıoğlu L, Kaya B, Fitöz S, Uysalel A. Middle aortic syndrome in a teenager. Turk J Pediatr 2014; 56: 658-660.

Middle aortic syndrome (MAS) is a rare pathology that involves diffuse/segmental narrowing of the distal thoracic or abdominal aorta. The most common clinical manifestation is severe hypertension, which requires multiple antihypertensive medications and/or surgical repair. We report the surgical repair of MAS in a 14-year-old male.

Key words: middle aortic syndrome, aortic coarctation.

Middle (or mid-) aortic syndrome (MAS) is considered a cause of curable hypertension. This uncommon condition is found in children and young adults. If left untreated, death results from cardiovascular complications such as cerebral hemorrhage or cardiac failure.

Case Report

A 14-year-old boy was referred to our department with MAS. His blood pressure was high (180/100 mmHg), despite being treated with three antihypertensive drugs. There was a pressure differential (70 mmHg) between the upper and lower limbs. Computed tomographic (CT) angiography showed a long segment (9 cm) with significant narrowing of the aorta between T₈ and T₁₂ with multiple aortico-aortic collaterals. The abdominal aorta lumen size was, at its smallest point, decreased by 95% (Fig.1). The visceral branches were normal. Surgery was planned, and a lumbar cerebrospinal fluid (CSF) drainage catheter was placed perioperatively.

In the operation, left thoracotomy was performed through the 6th intercostal space. The crux of the aortic hiatus was minimally dissected to expose the subdiaphragmatic portion of the abdominal aorta. Thoracic-abdominal aorto-aortic bypass was done. Because of the discrepancy of the pre- and poststenotic aortic segments, end-to-side anastomosis was performed using two different sizes of woven Dacron grafts (16 mm for distal, 18 mm for

proximal anastomosis) (Fig. 2). The gradient decreased to 10 mmHg when the anastomosis was completed. Postoperative follow-up was uneventful. The patient was discharged on the 10th post-operative day with administration of angiotensin-converting enzyme inhibitor. Four months later, the patient continued to be free of hypertension, and follow-up CT revealed a patent thoracoabdominal bypass (Fig. 3).

Discussion

Coarctation of the abdominal aorta, also known as mid-aortic syndrome, is a very rare condition^{1,2}. The etiology is unclear. Most patients are young, so the malformation may be congenital, which has been suggested to be the consequence of unequal fusion of the paired embryonic dorsal aortas. Acquired MAS is associated with Takayasu or temporal arteritis, neurofibromatosis, retroperitoneal fibrosis, fibromuscular dysplasia, mucopolysaccharidosis, Williams syndrome and CHARGE syndrome.

On the basis of Hallet's anatomic classification³, four types of MAS have been described. The coarctation sites are suprarenal in types I and III, and infrarenal in types II and IV. Types I and II are associated with renal artery stenosis, while types III and IV are not. The most common anatomic form is interrenal (19-52%), followed by suprarenal (11-40%), infrarenal (19-25%) and diffuse (12%)⁴. Our patient was compatible with type III.

Severe hypertension is the cardinal clinical

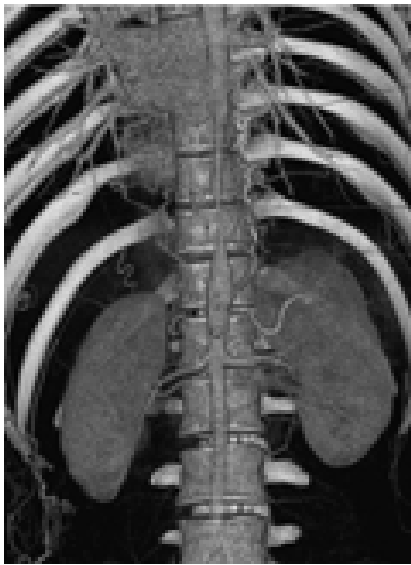


Fig. 1. Preoperative CT image of mid-aortic syndrome.

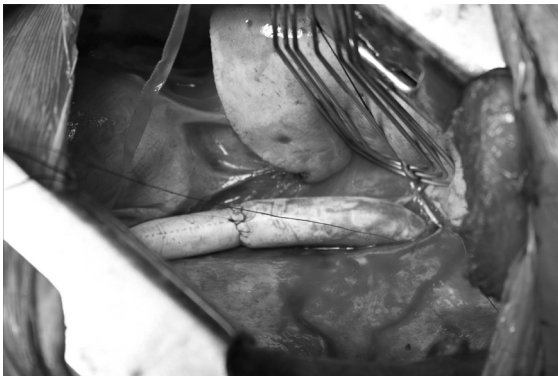


Fig. 2. Aorto-aortic bypass with two different sizes of Dacron graft.



Fig. 3. CT image of bypass graft between distal thoracic aorta and abdominal aorta at three months follow-up.

feature of mid-aortic syndrome and can be seen without signs of renal artery stenosis in the presence of sufficiently narrow suprarenal coarctation. The presence of collateral vessels may explain why our patient, with a severe stenosis above the celiac axis, did not have symptoms of visceral involvement.

Spiral CT angiography with 3D reconstruction gives additional information regarding the presence of any associated inflammatory changes: for instance, the presence of aortic wall thickening that angiography cannot evaluate. This method may reduce the need for invasive angiography and is currently the ultimate diagnostic tool in many specialized institutions.

Therapeutic approaches for MAS are medical, endovascular and/or surgical interventions^{5,6}. The treatment must be individualized on the basis of the length of the hypoplastic segment and associated renal artery involvement. Endovascular therapy has been shown to be successful in decreasing the gradient. However, little long-term success has been reported. Freedom from reintervention was only 58% at 1 year and 33% at 5 years. Freedom from reintervention after surgery was longer: 83% at 1 year and 72% at 10 years.⁷ Open surgery, in spite of its complexity, has been shown to be the primary treatment, as in our patient. Options for surgical reconstruction include resection of the stenotic segment and end-to-end anastomosis or interposition grafts, aorto-aortic bypass grafting, or patch aortoplasty; visceral reconstruction may also be required. We attempted to control the blood pressure medically, but due to intractable hypertension, we were obliged to proceed with surgery and so performed a thoracic-abdominal aorto-aortic bypass for the long-segment stenosis.

It is preferable to postpone surgery until the child has reached his full growth potential, except in the presence of severe intractable hypertension. In one study, it was reported that 42% of patients presented with hypertensive encephalopathy, and 45% died before 34 years of age⁸.

As in our patient, early normalization of hypertension is usually reported. Operative mortality rates have been low. Long-term patency is also good, and chronic anticoagulation is not required.

Conclusion

Timely referral for corrective surgery is essential, so any young patient with uncontrolled hypertension should be evaluated for abdominal aortic coarctation. Surgery remains the treatment of choice, offering immediate amelioration of the symptoms and durable clinical outcomes, controlling complications of hypertension and improving life expectancy.

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