Infrarenal Aortic Coarctation as a Cause for Hypertension

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Abstract

We present a case report of a 29-year-old male who was diagnosed with asymptomatic hypertension. Computed tomography angiography (CTA) and magnetic resonance angiography (MRA) showed a stenotic aorta, with extensive collateral flow called the middle aortic syndrome. The aetiology of middle aortic syndrome is poorly understood. Although treatment is preferably surgical, our case shows that medical therapy can be successful.

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Introduction

Segmental narrowing of the abdominal and its visceral branches is called middle aortic syndrome (MAS) and may be congenital or acquired. Segmental aortic stenosis may be located at the suprarenal, inter-renal or infrarenal aorta, with a high propensity for concomitant stenoses in both the renal (63%) and visceral (33%) arteries. Hypertension proximal to the aortic stenosis and relative hypotension distal to it are characteristic findings in MAS. Typical manifestations include headache, early fatigue on exertion and bilateral lower-limb claudication. Although the primary treatment in young patients is open surgery sometimes in stages, endovascular therapy may also be of use. 1 Here, we describe a case of a medically treated MAS.

Case description

A 29-year-old Caucasian male with no previous medical history was diagnosed with asymptomatic hypertension. On physical examination his blood pressure measured 231/122 mmHg in both arms, with a regular pulse of 69 beats per minute. Auscultation of the abdomen revealed a murmur in the upper abdomen. Peripheral pulsations were all palpable and there was no history of abdominal pain or claudication. Serum creatinine was 78 umol l−1 and modification of diet in renal disease-glomerular filtration rate (MDRD-GFR) was >90 ml/min/1.73 m2. The electrocardiogram was normal. Initial computed tomography angiography (CT-A) (Fig. 1) showed a stenotic aorta at the level of the renal arteries and was complemented with magnetic resonance angiography (MRA) (Fig. 2) as workup for a surgical correction. There was extensive collateral flow visible via the internal mammary arteries, the lumbar arteries and the left and right epigastric arteries to the iliac system. The patient was diagnosed with MAS based on these imaging findings. The patient was initially put on oral antihypertensive medication (enalapril 30 mg, nifedipine oros 30 mg, metoprolol 50 mg, doxazosine 8 mg and 12.5 mg hydrochlorothiazide). Six weeks later, his blood pressure was 145/75 mmHg lying down with a pulse of 80 beats per minute. The patient refused further surgical therapy. Two years after initial presentation, his renal function and blood pressure were stable, his last blood pressure measuring 135/78 mmHg.

Discussion

Since MAS is most frequently encountered in young adults, treatment is preferably surgical. Physical incapacitation and the severity of the hypertension are the main determinants to decide for surgical correction. Furthermore, since most patients are young, lifelong antihypertensive therapy is generally not advocated. Traditionally, the aortic stenosis is reconstructed with bypass grafting (aorto-aortic bypass) in the case of diffuse or patchy stenosis and patch aortoplasty in a small to moderate stenosis. When renal and visceral arteries are involved, multiple bypasses can be performed to improve bowel perfusion and reverse hypertension. 2 These interventions have a 30-day mortality rate of around 5% and offer hypertension improvement or normalisation in 94% of the cases. Endovascular therapy with balloon angioplasty and stenting is also an option. There is no long-term follow-up data,
so the efficacy and durability remain controversial. In our case, a surgical reconstruction was advocated but the patient refused. It shows that the medical treatment of MAS can be successful, at least for a follow-up of 2 years.

The aetiology of MAS is poorly understood. A congenital cause has been described due to incomplete fusion or over-fusion of the primitive embryonic dorsal aortas. Possible acquired causes include fibromuscular dysplasia or Takayasu’s arteritis. Patients usually present in the early decades of life with uncontrolled hypertension due to renal artery stenosis. Intestinal ischaemia and claudication are not a common feature due to extensive collateralisation. There are three main collateral pathways by which arterial blood flow can reach the lower extremities when the infrarenal aorta is occluded. The first is the visceral pathway through the inferior mesenteric artery and then via rectal branches to the iliac arterial system. A second route is the systemic pathway via intercostal and lumbar arteries, which supply branches of the iliolumbar and superior gluteal arteries, thereby filling the internal iliac arterial system.

A third, much less common route is via the internal mammary and epigastric arteries to the external iliac system, as in the presented case. This last route is called Winslow’s pathway. Knowledge of these potential collateral routes is essential to limit perioperative complications.

Conclusion

MAS is a poorly understood entity. Although primary treatment in young patients is preferably surgical, our observation in this patient suggests that medical therapy alone may be effective when extensive collaterals are present, and this can be considered in patients who are at high risk or refusing surgical intervention. When planning surgical intervention, knowledge of the collateral pathways is essential to limit complications.

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Conflict of Interest

None.

References