CLINICAL SUMMARY

A 4-year-old boy was noted to have a murmur by his pediatrician. He was referred to a cardiologist, who measured his blood pressure at 160/60. Echocardiogram revealed concentric left ventricular hypertrophy (LVH) with a “drag” in the descending thoracic aorta consistent with coarctation. Computed tomography (CT) angiogram of the abdomen revealed an extremely narrow upper abdominal aorta [Figure 1a and b], from which a diagnosis of midaortic syndrome (MAS) was made. There was no soft-tissue thickening surrounding the narrowed portion (as is often seen in Takayasu arteritis), nor was there involvement of branch vessels to specifically suggest other vasculopathy such as fibromuscular dysplasia as a cause. There was no clinical evidence of neurofibromatosis or Williams syndrome.

The patient was started on four blood pressure medications (beta-blocker – atenolol, vasodilator – minoxidil, angiotensin-converting enzyme inhibitor – enalapril, and calcium channel blocker – amlodipine), with blood pressure reduction to 120s/50s in the arms and 40s/20s in the legs. The patient was eventually unable to walk due to severe claudication and was referred to a tertiary hospital for a thoracoabdominal bypass graft. Polytetrafluoroethylene graft was successfully placed from the level of the diaphragm to just above the origin of the inferior mesenteric artery [Figure 2]. Postoperatively, his blood pressure was controlled with one antihypertensive medication (amlodipine). On follow-up echo 18 months later, LVH had resolved. He is now completely asymptomatic with no complaints of chest pain, dizziness, shortness of breath, syncope, or fatigue.

DIFFERENTIAL DIAGNOSIS

Takayasu arteritis, neurofibromatosis type 1, fibromuscular dysplasia.

DISCUSSION

MAS is characterized by narrowing of the descending aorta between the distal aortic arch and the aortic bifurcation, comprising 0.5%–2% of cases of aortic stenosis.[1] Disease involvement may be focal or long segment smooth narrowing and can extend into visceral arterial branches. Focal stenosis most commonly occurs in the suprarenal portion (60%–70% of cases), with focal renal (20%–25%) and infrarenal (10%–15%) involvement being less frequent.[2]
Patients with MAS present predominantly with refractory hypertension. If left untreated, symptoms can progress to intermittent claudication of the legs, congestive heart failure, and renal insufficiency. Symptoms include sequelae of hypertension, renal insufficiency, mesenteric ischemia, solid organ hypoperfusion, and lower limb ischemia. Chronic renal ischemia leading to renovascular disease can further exacerbate refractory hypertension. Untreated MAS has a 90% mortality rate by the sixth decade of life due to end-organ damage.

MAS can be associated with inflammatory arteritis including Takayasu’s, neurofibromatosis type 1, fibromuscular dysplasia, mucopolysaccharidosis, congenital Rubella syndrome, Noonan’s syndrome, and Williams syndrome.

MAS may be difficult to diagnose clinically, as it is one of the rarest causes of refractory hypertension. Renal function laboratory values may be abnormal, which could prompt further evaluation. Echocardiography and renal ultrasound can be useful initial imaging tests to evaluate for thoracic coarctation and renal parenchymal atrophy. While not definitive, both of these examinations may show secondary or direct evidence of aortic narrowing. Angiography was formerly the gold standard to diagnose MAS, but definitive diagnosis is now often achieved with either CT or magnetic resonance angiography.

The treatment of mid-aortic syndrome can be medical, endovascular, or surgical. A combination of medical management and invasive strategies can result in adequate control of hypertension and preservation of end-organ function.

Percutaneous interventions are effective in relieving obstruction in the acute setting, but there is a high incidence of refractory hypertension and recurrent stenosis requiring repeat intervention. Up to 50% of endovascular repairs require reintervention within 5 years of surgery. Serial dilation utilizing drug-eluting balloons may have promise as a percutaneous option. Percutaneous interventions can be associated with severe complications, including vascular tears, development of aneurysms, dissection, and even death.

Surgical interventions include aorto-aortic bypass, patch aortoplasty, and primary aortic repair. Improvement or resolution of hypertension after surgery in the mid to long term has been reported at 70%. Freedom from reintervention after surgery is reported to be 72% at 10 years. Complications occur in 9% of cases and include graft stenosis, bleeding, thrombosis, and iatrogenic tears. The mortality rate is 2.9%–4%.

While rare, MAS is a highly morbid and poorly understood disease process that occurs in both children and adults. The timing of disease progression is variable, but the risk of end-organ damage and death remains high no matter the age of presentation. It is important for imagers and clinicians to be aware of this disease and to have a high index of suspicion on screening. This can facilitate early diagnosis and treatment, as management is often a stepwise process that may require both noninvasive and surgical approaches.

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Conflicts of interest
There are no conflicts of interest.

REFERENCES


