Surgical management of middle aortic syndrome in an adult

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ABSTRACT

Middle aortic syndrome (MAS), a coarctation of the lower thoracic and/or abdominal aorta, is typically diagnosed and treated in the pediatric population. We present a 48-year-old patient with a long-standing history of hypertension who was lost to follow-up owing to a lack of insurance coverage. After two myocardial infarcts owing to severe hypertension, a vascular workup including a computed tomography angiogram revealed a diagnosis of MAS. He underwent open vascular reconstruction with a thoracoabdominal Dacron bypass graft. He was discharged within 1 week with no hypertension or claudication. Adult patients diagnosed with MAS should undergo open or endovascular surgical repair with close follow-up. (J Vasc Surg Cases and Innovative Techniques 2020;6:38-40.)

Middle aortic syndrome (MAS) is a rare vascular pathology in which there is hypoplasia or coarctation of the distal thoracic and/or abdominal aorta. It can be congenital or acquired.[1] Most cases are congenital and are diagnosed in childhood, making MAS largely a pediatric disease. The lifespan of an individual with untreated MAS is less than 40 years, usually precipitated by the consequences of poorly managed renovascular hypertension such as myocardial infarct (MI), aortic rupture, or intracranial hemorrhage.[2]

We present an unusual case of an adult diagnosed with MAS after two MIs revealed distal thoracic aortic occlusion during an attempt to catheterize his cardiac vessels via the femoral artery. We discuss the operative management options for this type of patient population, and propose follow-up recommendations. Informed consent was obtained from the patient for publication.

CASE REPORT

A 48-year-old man was incidentally discovered to have hypertension during a routine medical visit at age 27. He was placed on two antihypertensive medications, but was lost to medical follow-up until age 42, when he suffered an MI. At age 47, he had another MI, prompting percutaneous coronary intervention with drug-eluting stent placement. At that time, an attempt to selectively cannulate the supraceliac aorta was unsuccessful owing to severe stenosis. A computed tomography angiogram revealed a thoracic aortic tapering and occlusion above the celiac artery, confirming MAS (Fig 1) with innumerable collateral vessels between the thoracic aorta and the superior mesenteric artery, celiac, and bilateral iliac arteries.

The patient was lost to follow-up owing to poor insurance coverage until 8 months later, when he presented to vascular surgery due to progressively worsening exercise-induced buttock and thigh claudication and worsening blood pressure requiring four antihypertensives. Ankle-brachial indices were 0.60 bilaterally. An open aortic bypass from thoracic to infrarenal aorta was recommended to avoid transecting the aorta near large spinal collaterals, thereby reducing the risk of spinal ischemia. An in situ bypass would not have been feasible given the degree of supraceliac aortic occlusion.

Preoperative approach and exposure of the thoracic and infrarenal aorta. A thoracic spinal drain was placed preoperatively and removed after 24 hours of normal neurologic examinations. Cardiopulmonary bypass was not indicated because the proximal aorta-graft anastomosis was planned to be in the distal thoracic aorta.

A posterolateral left thoracotomy was performed. Multiple large intercostal collateral arteries were suture ligated upon dissection of the seventh intercostal space. The aorta was dissected to the level of the inferior pulmonary vein. Preoperative imaging demonstrated a 5-cm segment proximal to the occlusion that was free of large intercostal branches. This aortic segment was identified and found to be soft and normal in caliber.

A second incision from the tip of 11th rib extended inferomedially toward the umbilicus. The dissection was carried down through the abdominal wall and spared the rectus (Fig 2). A retroperitoneal dissection identified the infrarenal aorta which appeared diminutive, but had pulsatile flow. The celiac artery, superior mesenteric artery, and internal mammary artery were palpable. The supraceliac aorta was palpated and had no flow. The spleen and left kidney were mobilized anteriorly.

Tunneling of the conduit and anastomosis. With the spleen and left kidney reflected medially, a small diaphragmatic incision was made lateral to the left crus, posterior to the diaphragmatic sulcus and adjacent to the vertebral body. Thoracic aortic clamps were placed distally then proximally in a bare area avoiding large intercostal collaterals and the inferior pulmonary

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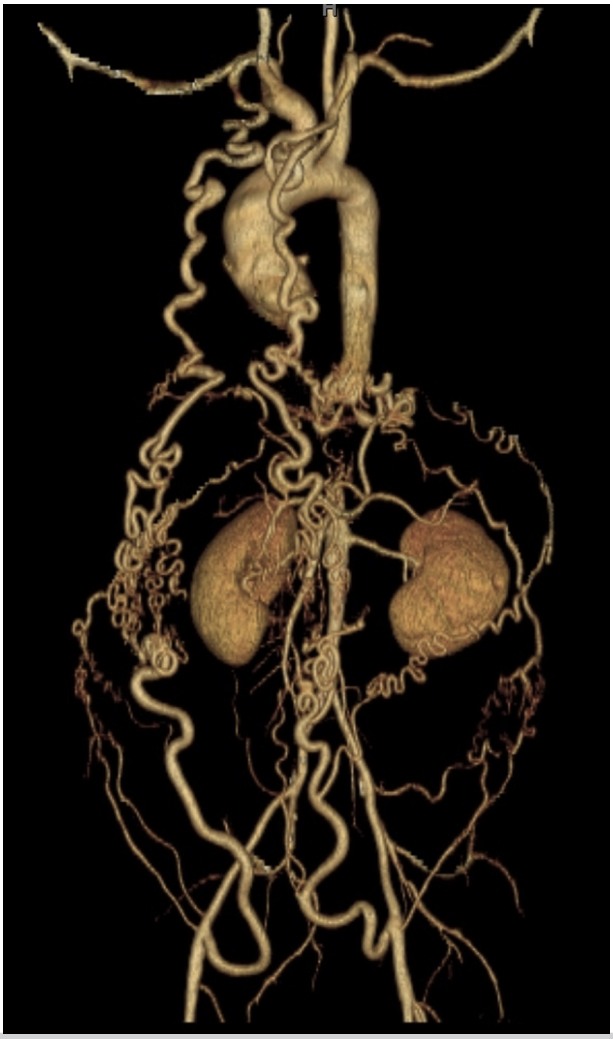

Fig 1. Preoperative computed tomography (CT) angiogram demonstrating thoracic aortic tapering, supraceliac occlusion, and innumerable large transhiatal collaterals.

vein. A 14-mm end-to-side Dacron graft was sewn to the thoracic aorta. The graft was tunneled through the diaphragmatic defect and laid end-to-side at the infrarenal aorta. At this level, the inferior mesenteric artery was controlled. A distal aortic clamp above the aortic bifurcation followed by proximal clamping several centimeters cephalad allowed for a Dacron to aorta end-to-side anastomosis. The thoracic anastomosis was covered with a bovine pericardial patch along the Dacron to decrease the risk of aortopulmonary fistula.

Postoperative course. The patient had no neurologic or vascular complications. His mean arterial blood pressure preoperatively was 110, on the day of discharge was 93, and at 1 month follow-up was 101. He was restarted on low-dose aspirin and clopidogrel to continue to protect the primary patency of his coronary stent. He was discharged on postoperative day 6 and seen in clinic 1 month later, off all antihypertensives and walking without claudication. A CT angiogram revealed an intact thoracoabdominal bypass graft and significantly diminished abdominal wall collateralization (Fig 3).

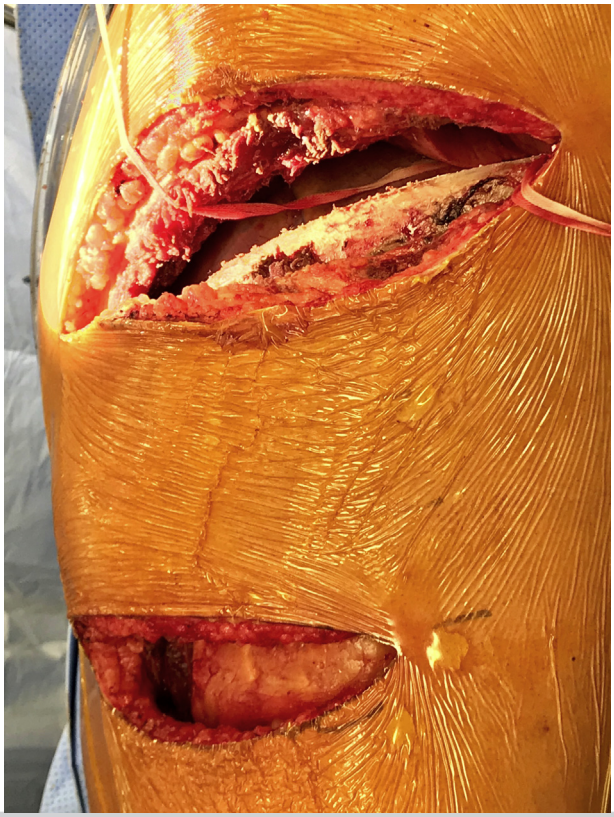

Fig 2. Intraoperative photograph of the thoracic and abdominal incisions facilitating aortic exposure and bypass.

DISCUSSION

MAS, also known as aortic coarctation or aortic hypoplasia, may be classified into either suprarenal (70%), infrarenal (7%), or intrarenal (23%) manifestations.[3] Renal and visceral artery involvement is seen in 80% and 22%, respectively,[3] which was not observed in this patient. The average age of diagnosis is 7 years old.[4] Largely associated with genetic syndromes, MAS likely arises from an early development error.

The predominant clinical manifestation of MAS is hypertension. Indications for surgery are medically refractory hypertension, the presence of symptoms (claudication, intestinal ischemia), and end-organ damage such as renal failure.[5] Optimal management of this patient would have included earlier diagnosis and close follow-up to encourage prompt surgical revascularization.

A systematic review of childhood MAS showed that open surgical bypass was twice as frequent as endovascular intervention. Sixty-five percent of the surgical patients underwent an uneventful repair, and 8% experienced technical failure. Fifty-one percent of the endovascular patients underwent an uneventful repair, and 28% experienced technical failure.[6] The operative management for MAS includes aortoaortic bypass, patch aortoplasty, and visceral/renal artery reconstruction if needed.[2]

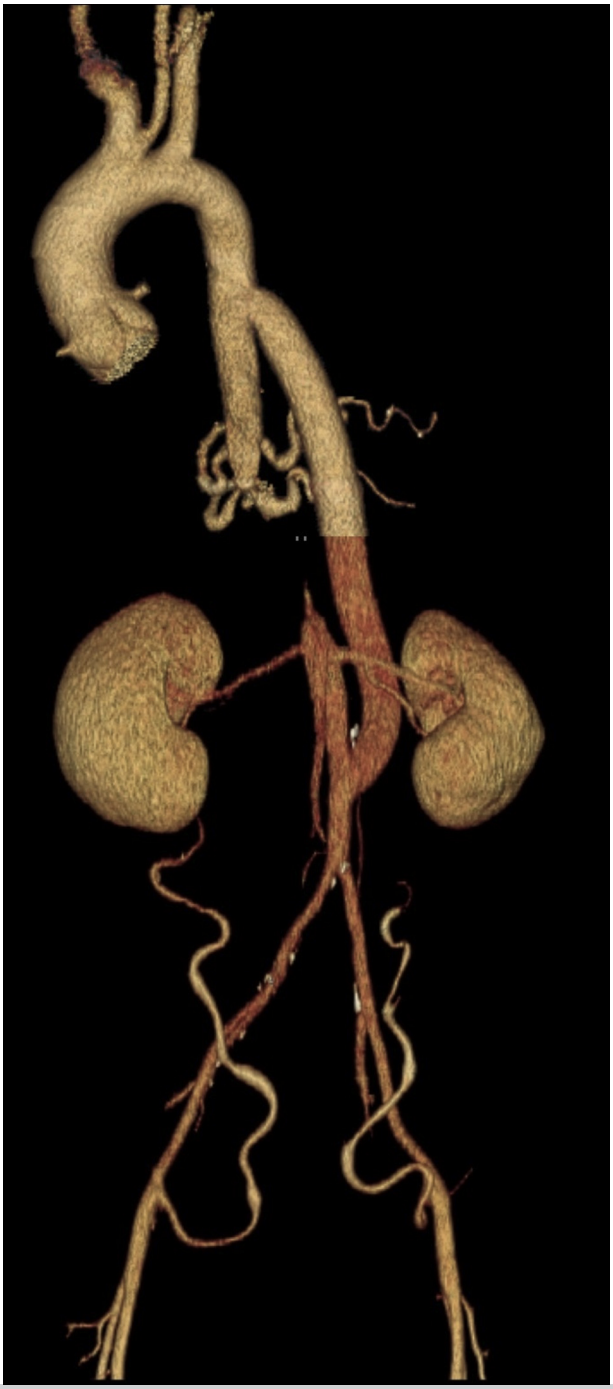

Fig 3. Postoperative computed tomography (CT) angiogram demonstrating thoracic aortic bypass to the infrarenal aorta and significantly diminished collateral networks.

MAS can include visceral artery stenosis. Visceral endarterectomy and endovascular options for revascularization may not be feasible owing to the congenital narrowing, which distinguishes this entity from atherosclerotic occlusive disease.[6] Similarly, endovascular approaches are less likely to be successful in MAS because of the diminutive vessels.[6] Extra-anatomic bypass (eg, axillary-femoral bypass graft, ascending aorta to femoral bypass graft) may be used in a hostile abdomen.[6] In young patients with an otherwise healthy abdomen, the lower durability and patency profile of extra-anatomic bypasses should be weighed against the need for long-lasting patency. This patient's known chronic aortic occlusion and young age directed the surgical decision making toward a technically feasible and highly durable aortoaortic bypass.

No established follow-up guidelines exist for patients who undergo surgical intervention for MAS. Previous authors have recommended lifelong follow-up, owing to the variable interval time between surgery and complications. Postoperative complications include anastomotic aneurysms, graft deterioration, heart failure, and cerebrovascular accidents.[7] This is illustrated by a case report of a patient who developed an aortoenteric fistula that presented as massive hematemesis 24 years after her repair.[5] We recommend a CT angiogram 1 month postoperatively to evaluate for pseudoaneurysmal degeneration at the anastomoses. If there are no anastomotic complications, repeat imaging at 3 to 5 years is reasonable, given that the original pathology is congenital occlusion and not aortic aneurysmal disease.

In conclusion, we present an unusual case of an adult patient with MAS who experienced two MIs and was diagnosed after difficulty cannulating the abdominal aorta. When clinicians encounter a young patient with medically refractory hypertension, evidence of end-organ ischemia, or bilateral peripheral claudication, their workup should include congenital vascular abnormalities and genetic syndromes such as MAS. These patients warrant lifelong surveillance.

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