Late presentation of middle aortic syndrome complicated with severe aortic regurgitation; the role of endovascular intervention as a bridging for Bentall surgery

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ABSTRACT

Middle aortic syndrome presents with segment narrowing of the descending thoracic and abdominal aorta. A common manifestation is uncontrolled hypertension, which can lead to severe aortic regurgitation in the long term. We have presented the case of a 31-year-old woman with worsening heart failure symptoms and longstanding uncontrolled hypertension. Echocardiography revealed severe aortic regurgitation. Aortic computed tomography showed severe stenosis of the aorta at the diaphragm level. Stent graft implantation was performed, followed by Bentall surgery 1 year later. Endovascular stent graft implantation of the descending aorta can be used safely as a bridging surgery for the Bentall procedure to reduce the patient's blood pressure and relieve heart failure symptoms. (J Vasc Surg Cases Innov Tech 2022;8:48-52.)

Keywords: Aortic regurgitation; Bentall procedure; Late presentation; Middle aortic syndrome; Stent-graft implantation

Middle aortic syndrome (MAS) is a rare vascular anomaly involving long segment narrowing of the descending thoracic and abdominal aorta.[1] It comprises 0.5% to 2.0% of aortic stenosis cases, which mostly affect children and young adults.[2,3] The location and length of the aortic narrowing in MAS vary greatly and, therefore, should always be considered for determining the intervention options.[2,4] The common clinical manifestations and complications include severe uncontrolled hypertension, congestive heart failure, and hypertensive encephalopathy.[3] Medical management of hypertension caused by MAS has been largely unsuccessful.[5,6] Thus, invasive intervention is often required to achieve adequate blood pressure (BP) control, relieve symptoms, and reverse or prevent end-organ damage.[7-9]

In the present case report, we have described the case of a patient with late presentation of MAS with uncontrolled hypertension and severe aortic regurgitation. The highlight of the present case was the use of endovascular intervention as a bridging therapy to Bentall surgery for aortic regurgitation to reduce morbidity. The patient provided written informed consent for the report of her case and imaging studies.

CASE REPORT

A 31-year-old woman had been referred to our center with complaints of worsening shortness of breath of 3 months' duration. She had symptoms of paroxysmal nocturnal dyspnea, orthopnea, and dyspnea with effort. She also had been experiencing headache and persistent hypertension for 14 years despite medical therapy. Her daily medications were amlodipine 1 × 10 mg, telmisartan 1 × 80 mg, spironolactone 1 × 25 mg, and bisoprolol 1 × 2.5 mg. The physical examination showed differences in the BP between the upper and lower extremities (215/83 mm Hg in the left arm, 240/69 mm Hg in the right arm, and 159/84 mm Hg in both legs). Bruit was also heard on the patient's back. Decreased palpable pulses in both lower extremities were noted. The initial laboratory findings showed normal renal function and inflammatory markers. Electrocardiography revealed left ventricular hypertrophy. The chest radiograph showed cardiomegaly and aortic elongation. Echocardiography revealed severe aortic regurgitation due to aortic root dilatation, aneurysm of the ascending aorta, left ventricular hypertrophy, good left and right ventricular systolic function with an ejection fraction of 81%, and tricuspid annular plane systolic excursion 3.2 cm. The findings from the other examinations were within normal limits.

Aortic computed tomography angiography revealed dilatation of the ascending aorta, severe stenosis of the aorta at the diaphragm level, a penetrating aortic ulcer, and an irregular aortic wall (shaggy aorta). The structure of the other aortic branches

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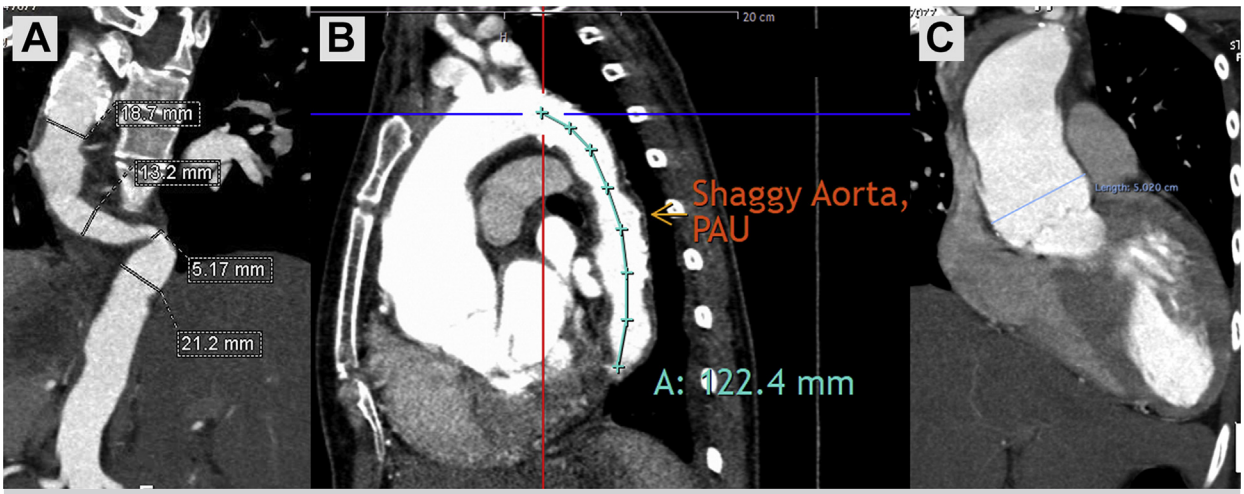

Fig 1. Computed tomography angiogram of the aorta. **A,** Severe stenosis at the level of the diaphragm. **B,** Sagittal view of computed tomography angiogram of the aorta showing the irregular aortic wall (shaggy aorta) and penetrating aortic ulcer (PAU) with a length ~122.4 mm (*A*). **C,** Aortic root diameter.

was within normal limits (Fig 1). Based on previous findings, MAS was diagnosed, concomitant with severe aortic regurgitation and New York Heart Association functional class III. Our team decided to proceed with stent-graft implantation of the descending aorta, followed by Bentall surgery later.

Aortography showed significant stenosis at the distal thoracic aorta, proximal to the celiac artery, with a mean pressure gradient of 35 mm Hg (Fig 2, *A*). A thoracic stent-graft, 26 × 150 mm (S&G Biotech, Inc, Yongin, South Korea) was deployed at the descending aorta, followed by postdilatation with a 14 × 60-mm Armada balloon (Abbott Vascular, Abbott Laboratories, Chicago, Ill) at the middle part of the stent. The final angiogram revealed <20% residual stenosis, and the mean gradient had subsided to 20 mm Hg (Fig 2, *B*). The patient was discharged in stable condition, and her BP had decreased reduced. At the 1-month follow-up visit, she had no symptoms, and her BP was controlled (143/42 mm Hg in the upper extremities and 126/61 mm Hg in the lower extremities) with antihypertensive medication consisting of candesartan 2 × 8 mg, amlodipine 1 × 5 mg, and spironolactone 1 × 12.5 mg. After 1 year of follow-up, her BP was 105/36 mm Hg, after which she underwent the Bentall procedure with good results. At the latest follow-up visit, her medications were aspirin 1 × 80 mg, clopidogrel 1 × 75 mg, bisoprolol 1 × 5 mg, candesartan 1 × 4 mg, and simvastatin 1 × 20 mg.

DISCUSSION

MAS is a clinical condition resulting from segmental narrowing of the abdominal or distal descending thoracic aorta.[8] Most MAS cases will be diagnosed in children or adolescents, and most cases will be idiopathic; however, some have been described in association with genetic and acquired inflammatory disorders.[5] The cause of MAS in our patient was determined to be Takayasu arteritis in the stenotic or chronic phase because the patient's case had fulfilled three criteria based on the American College of Rheumatology guidelines: (1) age at onset of <40 years; (2) BP difference >10 mm Hg in systolic BP between the arms; and (3) irregular and narrowing of the aorta revealed by angiography. Aortic regurgitation can also develop in patients with Takayasu arteritis.[10]

The clinical manifestations of MAS depend on the anatomic location of the narrowing. Symptoms and complications can be associated with hypertensive end-organ damage and malperfusion syndrome.[1] In our patient, resistant hypertension, differences in the BP between the upper and lower limbs, and the presence of bruit at the back pointed to the diagnosis of MAS. This was confirmed by computed tomography angiography, with the stenotic part of the descending aorta at the level of diaphragm.

In the present patient, the aortic regurgitation was thought to have resulted from the longstanding uncontrolled hypertension due to MAS. The aortic valve anatomy was normal. Untreatable and persistent high BP in the later course could contribute to circumferential stress and elastin fragmentation, thus resulted in dilatation of the aorta.[11]

The management of MAS is complex and requires a multidisciplinary and individualized approach. The appropriate age for repair has not been established. However, only <20% of patients with untreated MAS will survive until the age of 40 years.[1,12] Although earlier repair is probably preferable because it can prevent or reduce target end-organ damage, repair in the earlier stage will be complicated owing to the growing size of the aorta.

Porras et al[1] found that stenosis of ≥60% will most likely require invasive management but that stenosis of <40%

Fig 2. Aortography. **A,** Aortogram showing significant stenosis. **B,** Completion aortogram after thoracic endovascular aortic repair deployment.

is unlikely to require invasive management. Other indications for repair include refractory hypertension, evidence of end-organ damage, and significant symptoms (eg, claudication or abdominal angina).[1] Because our patient had a history of persistent hypertension, left ventricular hypertrophy and, worsening congestive heart failure, invasive intervention was required to achieve adequate BP control, relieve her symptoms, and prevent further end-organ damage.

The choice of invasive strategy depends on the location and length of the stenosis and extent of the visceral vessel involvement.[1,13,14] Approximately 40% to 55% of patients with MAS will undergo open surgery such as aortoaortic bypass, patch aortoplasty, or primary aortic repair after aortic lengthening.[1,2] The complication and mortality rate after surgical procedures has been 9% and 4%, respectively. Complications have included graft stenosis, bleeding, thrombosis, and dissection.[2] Concomitant Bentall surgery with other aortic surgery can increase the operative risks, especially for patients with a New York Heart Association class of ≥3 and severe hypertension.[15]

Endovascular intervention of the middle aorta as an alternative surgery can successfully relieve the obstruction and improve the gradient across the lesion immediately after the procedure.[1] With recent advances in endovascular therapy, percutaneous transluminal angioplasty for stenotic aortic lesions has been reported to show excellent immediate and long-term results compared with open surgical procedures.[16,17]

Simple stent-graft implantation without fenestration can be considered for selected cases with discrete or web-like aortic stenosis located at some distance from the mesenteric or renal artery orifices.[8,18] Postdilation with a balloon will often be needed after stent deployment. However, recommendations for the best balloon size, type, and inflation pressure remain unknown. Morrissey et al[19] reported that the balloon size for postdilation should match the size of the adjacent normal luminal diameter, ~21 mm for our patient. However, the largest balloon size available at our center was 14 mm.

Early success of the intervention has been defined as a residual gradient of ≤20 mm Hg without the need for early reintervention.[8] In our patient, the residual gradient was 20 mm Hg, and she had remained free of early intervention for 1 year. Her BP and symptoms of heart failure had improved significantly, making her condition fit for another operation. Hypertension is an independent risk factor for late death after Bentall surgery.[15]

The issue for stenting in patients with MAS is the patency rate. Thoracic aorta lesions were more likely to obtain better improvement after aortic stenting compared with lesions involving the abdominal aorta.[16] Stents produce radial forces to the surrounding vessel wall, preventing elastic recoil, which then reduces the

Table. Summary of MAS studies using endovascular intervention

Investigator	Study design	Baseline characteristics	Intervention	Outcomes
Kim et al[22]	Case report: 48-year-old man with MAS	Pressure gradient, 44 mm Hg; BP, 160/100 mm Hg; creatinine, 2.1 mg/dL	Stent-graft implantation with moderate dilation, followed by definitive dilation 3 months later	Pressure gradient, 28 mm Hg at first dilation; 18 mm Hg at second dilation; and 20 mm Hg at 20 months of follow-up; BP, 122/73 mm Hg; normal creatinine
Patel et al[23]	Retrospective cohort of 13 MAS patients	Patient 1, hypertension, abdominal pain; patient 2, chest pain, hypertension; patient 3, hypertensive, urgency	3 Patients had undergone endovascular intervention (covered stent)	Patient 1, hypertensive drug reduction (from 2 to 1 drug); patient 2, withdrawal of hypertensive drug; patient 3, reintervention, femoral artery repair, additional drug
Porras et al[1]	Retrospective cohort of 53 MAS patients	Hypertension	30 Hypertensive patients had undergone percutaneous intervention	1 patient was normotensive without medication; 13 patients were normotensive with medication; 5 remained hypertensive; 9 had undergone surgery; 2 had unknown outcomes
Glotzer et al[3]	Case report: 60-year-old woman with MAS and chronic hypertension	Pressure gradient >100 mm Hg; BP, 216/81 mm Hg	Covered stent	Patient lost to follow-up for 2 years; she presented with stenosis at distal portion of stent after 2 years; BP 220/85 mm Hg; pressure gradient, 60 mm Hg; stent was ballooned, and pressure gradient decreased to 20 mm Hg; 1 year later, the patient had excellent BP
Che et al[17]	Retrospective analytic study of 48 patients with MAS due to Takayasu arteritis	Mean NYHA functional class, 1.3; mean SBP, 179.06 ± 28.4 mm Hg; mean DBP, 95.36 ± 17.7 mm Hg	Covered stent	Reduction of mean NYHA functional class, 1.1 ($P = .06$); reduction of mean SBP, 149.56 ± 19.1 mm Hg ($P < .001$); reduction of mean DBP, 86.06 ± 14.3 mm Hg ($P = .006$)
Kim et al[24]	Case report: 42-year-old man with MAS and CHF	60 mm Hg; LVEF, 39%	Covered stent	Pressure gradient, 6 mm Hg; LVEF after 2 months, 44%; after 6 months, 57%; improved wall motion; BP, 120/90 mm Hg
Morrissey et al[19]	Case report: 48-year-old woman with MAS and Takayasu arteritis	SBP, 190-200 mm Hg	Bare metal stent	SBP, 140 mm Hg after 10 weeks

BP, blood pressure; *CHF,* congestive heart failure; *DBP,* diastolic blood pressure; *LVEF,* left ventricular ejection fraction; *MAS,* middle aortic syndrome; *NYHA,* New York Heart Association; *SBP,* systolic blood pressure.

obstruction. The choice of stent depends on the anatomy and size, operator preference, and stent availability.[4] Both bare metal stents (eg, Palmaz [Cardinal Health, Dublin, Ohio] and Cheatham Platinum [NuMED, Inc, Cross Roads, Tex]) and covered stent-grafts (eg, covered Cheatham Platinum [NuMED, Inc] and AndraStent [AndraMed GmbH, Reutlingen, Germany]) have been used for MAS or aortic coarctation with high success rates. Compared with bare metal stents, covered stent use has been associated with lower in-stent restenosis rates and lower aortic complications in patients with aortic coarctation.[20,21] Thus, the use of covered stents can yield good results in the management of MAS. Previous studies of MAS using endovascular intervention have been summarized in the Table.[1,3,17,19,22-24]

CONCLUSIONS

MAS will usually present with arterial hypertension and can lead to severe aortic regurgitation in the long term. Stent-graft implantation in the descending aorta can be used safely as a bridging therapy for Bentall surgery to reduce the patient's BP and relieve the symptoms associated with hypertension and heart failure.

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