CASE REPORT

Endovascular treatment of a rare cause of secondary hypertension in an elderly woman

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SUMMARY

A 70-year-old woman on four antihypertensive drugs including diuretics presented with accelerated hypertension and acute pulmonary oedema. She had a bounding brachial pulse with feeble femoral pulses. A 256 slice CT scan revealed the presence of severe diffuse thoracoabdominal atherosclerosis. Cardiac catheterisation revealed 125 mm Hg gradient across the atherosclerotic segment at the level of thoracic 10–11 vertebrae. A self-deploying stent was implanted in the thoracoabdominal segment reducing the gradient across the disease segment to 20 mm Hg with a significant improvement in the luminal diameter of the aorta. Her upper limb blood pressure normalised on two antihypertensive drugs 6 weeks later.

BACKGROUND

Middle aortic syndrome (MAS) is a rare condition characterised by diffuse hypoplasia or stenosis of the distal thoracic and abdominal aorta in association with stenoses within the renal or visceral arterial branches.^{1 2} It commonly presents in the second decade of life with symptoms of heart failure, chronic abdominal pain and lower limb claudication.^{2 3} Untreated patients typically succumb from congestive cardiac failure, hypertensive encephalopathy, or stroke.^{2–5} MRI and aortography show diffuse hypoplasia or stenosis of the middle portion of aorta between arch and terminal bifurcation. The treatment of this condition consists of either endovascular or surgical repair.^{2–4} There are few cases reported of this condition at an advanced age.⁴ We present one such case presenting at the age of 70 years who was treated using endovascular technique.

CASE PRESENTATION

A 70-year-old woman with a 7-year history of hypertension was on treatment with four drugs (optimal dose calcium-channel blocker, diuretic, β -blocker and α -blocker), with good compliance, when she presented with an episode of accelerated hypertension and acute pulmonary oedema. General examination revealed right arm blood pressure of 200/100 mm Hg. The brachial pulses were bounding and femoral pulses were feeble. A grade 2/6 systolic murmur was heard at the left upper sternal edge along with an abdominal bruit. Examination of the respiratory system revealed bilateral basal crepitations; examination of the other systems was unremarkable. The patient was managed with injection of nitroglycerine, morphine and frusemide. In addition, for a brief period, she

required non-invasive continuous positive airway pressure ventilation.

INVESTIGATIONS

Evaluation in hospital revealed normal haematological and biochemical parameters, the patient's blood urea and serum creatinine levels were 21 and 0.9 mg/dL, respectively. Ultrasonography of the abdomen revealed normal-sized kidneys (right kidney 9.8 cm, left kidney 10.2 cm) with an inconclusive renal Doppler. Work up for secondary hypertension including thyroid function test, urinary catecholamine estimation and serum electrolytes were negative. Chest radiograph showed evidence of bilateral pulmonary venous hypertension along with normal-sized heart. Echocardiography revealed concentric left ventricular hypertrophy with normal left ventricular systolic function. All valves were normal except for mild sclerosis of the aortic valve. A 256 slice CT scan performed on stabilisation showed diffuse thoracoabdominal atherosclerosis along with calcification.

DIFFERENTIAL DIAGNOSIS

Secondary cause of hypertension is suspected if there is a rapid onset of hypertension in young patients (<40 years), if there is sudden change in previously well controlled blood pressure and if hypertension becomes refractory to medical therapy. The aetiology of secondary hypertension could be vascular (renal artery stenosis, coarctation of aorta, preeclampsia), renal (chronic kidney disease, nephritic syndrome, glomerulonephritis, polycystic kidney disease), endocrine (pheochromocytoma, hyperaldosteronism, hypothyroidism, hyperthyroidism, hyperparathyroidism), toxic (alcohol abuse, medication such as contraceptive pills or non-steroidal anti-inflammatory drugs) and sleep apnoea syndrome.

TREATMENT

Cardiac catheterisation was undertaken and, under local anaesthesia (1% xylocaine), an 8 F sheath (Cordis Corporation, Miami Lakes, Florida, USA) was inserted into the right femoral artery using a modified Seldinger technique. Angiography was performed in the proximal descending thoracic aorta using a 6 F pig tail catheter to delineate the aortic coarctation segment. Diffuse narrowing of the thoracoabdominal aorta was noted along with a discrete calcific shelf-like lesion at the level of thoracic vertebrae 10–11 (D-10–11; figure 1). Pull back with a 6 F Judkins right coronary catheter revealed a gradient of 125 mm Hg across the coarctation

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Figure 1 Aortic angiogram showing diffuse narrowing of the thoracoabdominal aorta along with a discrete calcific lesion at the level of thoracic vertebrae 10–11.

segment (figure 2). A 12 F Cook sheath was exchanged in the right femoral artery. An 18×100 mm self-expanding Niti S vascular stent (Taewoong Medical Company Ltd, Seoul, South Korea) was deployed across the thoracoabdominal coarctation segment. Sequential dilation with 12 mm×40 mm and 14 mm×40 mm rival balloons (Bard Peripheral Vascular, Inc,

Figure 2 Aortic pressure tracing shows a gradient of 125 mm Hg across the prestenotic and poststenotic segment of the thoracic aorta.

Tempe, Arizona, USA) at 6 atm was performed to optimise the stent apposition. The pull back pressure gradient across the coarctation segment was reduced from 125 to 20 mm Hg. Angiography following placement of the stent demonstrated a significant improvement in vessel calibre and no evidence of dissection or perforation. The postintervention period was uneventful.

OUTCOME AND FOLLOW-UP

Postprocedure the medication of the patient was revised to two antihypertensive drugs (nebivolol 2.5 mg twice daily and amlodipine 5 mg once daily). During follow-up her blood pressure normalised and a 256 slice CT scan revealed a uniformly dilated patent aortic stent.

DISCUSSION

MAS, a disease affecting mostly the young, constitutes 2% of all aortic obstruction and is characterised by diffuse hypoplasia or stenosis of the distal thoracic and abdominal aorta.^{1–5} Pathogenesis of the disorder is often complex and may be idiopathic or secondary to granulomatous disease (Takayasu's arteritis), atherosclerosis, non-specific inflammation, cystic medial necrosis, neurofibromatosis, tuberculosis or tuberous sclerosis.^{2–6} This condition has been reported rarely in individuals older than 60 years.⁴ In view of the advanced age of our patient and presence of hypertension, atherosclerotic disease of the distal thoracic aorta appears to be the probable aetiological factor in the present case.

Typical manifestations of MAS include headache, easy fatigability and bilateral lower limb claudication.^{1–4} Patients with MAS are often first detected due to refractory hypertension, as was noted in the present case. Other presentations include intermittent claudication, congestive heart failure, renal insufficiency, abdominal angina secondary to mesenteric vessel stenosis and symptoms of hypertensive end-organ damage.^{1–2–7}

The severity of hypertension is the primary indication for intervention.⁷ Open surgery is the primary treatment of MAS and entails aorto-aortic bypass of the diseased segment or, less often, patch aortoplasty along with bypass grafting of the stenosed renal and visceral arteries, if involved.^{2–3–7} Endovascular therapy may provide sound minimally invasive treatment in MAS caused by discrete aortic stenosis that does not encompass the mesenteric and renal arteries.⁸ Balloons and self-expandable stents are used for endovascular treatment.^{9–11} Proximity of the atherosclerotic disease segment to the visceral vessels made us select a long self-expandable stent in the present case. Self-expandable stents are easier to deploy, adapt better to the wall of the aorta and, in most patients, have similar efficacy in reducing coarctation gradient as balloon-expandable stents do. In addition, these stents do not cause vascular injury as they do not have sharp edges. Compared with surgical techniques, endovascular therapy provides comparable immediate haemodynamic results with reduced morbidity and hospital-stay length at the cost of higher risk of restenosis and recurrence of coarctation.^{8–11} Owing to good initial and intermediate results, stent implantation is now considered as preferred therapy in most adolescents and adults with coarctation of the aorta. Hypertension is improved or cured in more than 70% of patients after successful treatment.^{2–5} The aortic stenosis in the present case spared the renal and splanchnic vessels, producing good clinical response with optimal blood pressure control.

Endovascular treatment of MAS is emerging as a minimally invasive, fast, efficient and safe alternative to surgery.

Learning points

- ▶ Middle aortic syndrome (MAS), a condition characterised by diffuse hypoplasia or stenosis of the distal thoracic and abdominal aorta, is a rare cause of secondary hypertension in older individuals.
- ▶ It is suspected in patients with refractory hypertension presenting with flash pulmonary oedema, headache, claudication of lower limbs or abdominal angina.
- ▶ Diagnosis of MAS requires a high index of clinical suspicion and is confirmed by thoracoabdominal CT scan and invasive cardiac catheterisation.
- ▶ Endovascular therapy is emerging as a minimally invasive, fast and efficient alternative to surgical treatment for patients with MAS.

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Competing interests None.

Patient consent Obtained.

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