Severe Hypertension in a Young Adult Resulting from Middle Aortic Syndrome

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Middle aortic syndrome is a rare condition which results in severe hypertension in children and young adults. This syndrome is characterized by severe segmental narrowing of the descending thoracic or abdominal aorta. The cause may be congenital or acquired. We report a 20-year-old male who presented with severe hypertension and hand tremor. Multi-detector computed tomography and aortography showed significant narrowing of the abdominal aorta and development of extensive collateral vessels. Indications for surgery in patients with middle aortic syndrome include refractory hypertension and severe ischemic symptoms. This case illustrates the importance of thorough and aggressive search for secondary hypertension and further imaging studies as indicated by clinical suspicion in young patients with severe hypertension.

Key Words: Middle aortic syndrome • Secondary hypertension

CASE REPORT

A 20-year-old male presented to outpatient clinic with two months of bilateral hand tremor and severe hypertension. Repeated measurements during office visits revealed blood pressure above 180/120 mmHg in both arms. The patient’s height was 172 cm, weight 68 kg, pulse rate 58 bpm and respiratory rate 18 breaths/minute. Physical examination revealed normal heart sounds without murmurs and no bruits in the neck or abdomen. Peripheral pulses were strong and symmetric. Neurologic and dermatologic examinations were unremarkable. The patient was a non-smoker with no significant medical or family history.

Initial laboratory findings, including complete blood count, electrolytes, thyroid profile, liver function test and renal function, were normal. The urinalysis showed no proteinuria. The levels of aldosterone and plasma renin activity were elevated to 254 pg/ml (normal range: 12-150 pg/ml) and 496 pg/ml (normal range: 1.8-59.4 pg/ml), respectively, while the levels of cortisol and adrenocorticotropin were within the normal limits. C-reactive protein (CRP) was less than 0.10 mg/dl, and erythrocyte sedimentation rate (ESR) was 2 mm/HR. A rheumatologic work-up including rheumatoid factor, anti-nuclear antibody, and anti-ds-DNA antibody did not suggest presence of autoimmune diseases. A presumptive diagnosis of renovascular disease was strongly suspected due to abnormal aldosterone and renin levels.

Further investigations with renal ultrasound and captopril renography were normal and suggested low probability of renal artery stenosis. The transthoracic echocardiography showed left ventricular hypertrophy. Multi-detector computed tomography (MDCT, Sensation 16, Siemens, AG, Munich, Germany) of the thoracic and abdominal aorta demonstrated a severe suprarenal abdominal aortic stenosis at the level between the celiac artery and the superior mesenteric artery orifices resulting in extensive collateral arteries (Figures 1 and 2).
prominent and dilated gastroduodenal artery provided collateral flow to the aorta distal to the narrowing (Figure 2). Moreover, there were enlarged internal mammary arteries communicating with the epigastric arteries, which in turn supplied blood flows to the bilateral common femoral arteries (Figure 3). The aortic arch and the subclavian arteries were normal.

Aortography confirmed the aortic stenosis in a suprarenal position with a pressure gradient of 68 mmHg across the stenotic aorta. Arteriography showed patent renal arteries bilaterally.

The patient’s blood pressure remained persistently above 160/100 mmHg despite aggressive antihypertensive therapy with three drugs: Doxazosin, amlodipine besylate, and nifedipine. Surgical intervention was suggested because of refractory hypertension. The patient refused to undergo surgical treatment and was lost to follow-up 3 months after the diagnosis.
DISCUSSION

Middle aortic syndrome (MAS) is a rare vascular condition characterized by segmental narrowing of the abdominal or distal thoracic aorta that was first described by Sen et al. in 1969.1 Whereas coarctation of the thoracic aorta at the level of ligamentum arteriosum is a well recognized cause of secondary hypertension, MAS is a much less common variety.2 It is usually found in children and young adults, with no sex predilection. Reports of patients over 40 years of age are rare, with only two cases noted over the age of 60 years.3

The severity and extent of narrowing of aorta and visceral vasculature account for the presence of symptoms.4 Abdominal bruit has a sensitivity of approximately 40 percent and is therefore absent in many patients, but has a specificity as high as 99 percent.5 Lower-limb ischemic symptoms such as intermittent claudication and leg weakness resulted from impaired blood flow distal to the stenosis can also be present. The absence of abdominal bruit and ischemic symptoms and the intact femoral pulses in our patient were likely due to the well developed collateral vessels. The patient’s neurologic examination was normal, and his hand tremor was probably non-specific.

Involvement of proximal renal arteries is reported in as many as 80% of cases of MAS;6 kidney atrophy, impaired renal functions and consequent hypertension may also be found. Although our patient presented with a short, localized suprarenal aortic stenosis without renal involvement, his severe hypertension was most likely secondary to decreased blood flow distal to the stenosis, leading to elevation of renin and activation of the renin-angiotensin system.

The exact etiology of MAS remains speculative and controversial. There are several hypotheses suggesting that it may be a congenital condition, or an acquired disease from insults in uterus or later in life. Maycock7 suggested that this congenital anomaly follows lack of unequal fusion of the two paired dorsal aorta with subsequent obliteration of one of those channels. Its known associations with neurofibromatosis, Williams and Alagille syndrome, intrauterine rubella infection, and tuberous sclerosis have also led to theory of a congenital process triggered by an intrauterine event.8,9 Support on the other hand for an acquired lesion comes largely from Asia and India,10 referring to cases with segmental aortitis and secondary atherosclerotic changes as variant of Takayasu’s arteritis.

Takayasu’s arteritis had been suspected initially in this patient since the disease can be confined to the abdominal aorta in the less common subtype.10 However, the patient had only met the criteria of onset before age of 40 and abnormal arteriogram, which made the possibility of Takayasu’s arteritis low.11 He had no history of inflammatory signs or symptoms suggesting arteritis, and inflammatory markers including CRP and ESR were within normal range. Dermatologic and neurologic examinations were unremarkable. Thus our patient may represent a congenital type of MAS.

Clinical clues suggesting possible renovascular hypertension warrant a more detailed and thorough evaluation, including additional imaging studies such as nuclear scan, magnetic resonance imaging (MRI) or MDCT. MDCT has been widely used in the evaluation of vascular disease.12 Angiography, however, is still the gold standard for confirming the abnormality and identifying coexistent vascular lesions.

The timing of surgical intervention is controversial and difficult to decide. Hypertension refractory to anti-hypertensive medications and severe ischemic symptoms are the major indications in proceeding to surgery.6,13 Without aggressive treatment, symptomatic patients usually die before the third or fourth decade of life from renal or cardiac failure or intracerebral hemorrhage.14 The current surgical management of MAS includes aorto-aortic bypass, vascular reconstruction by means of prosthetic or autologous venous graft, or percutaneous transluminal angioplasty. Patch aortoplasty may be considered in moderate or less lengthy aortic stenosis. Several reports had demonstrated satisfactory long-term results with the less invasive procedure of balloon angioplasty and stent placements10 in patients with middle aortic syndrome, although aneurysm formation and thrombosis of the stent are possible complications.15

REFERENCES

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