Mid Aortic Syndrome: a rare vascular disorder

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Abstract

Coarctation of abdominal aorta, Middle aortic syndrome (MAS) is a rare disease with only 200 reported cases. MAS may present clinically as uncontrolled hypertension , abdominal claudication or lower limb claudication. Surgical treatment is effective in controlling symptoms and improves life expectancy. We present another case of this rare entity, diagnosed and treated at Armed Forced Institute of Cardiology and Combined Military Hospital, Rawalpindi. This is the second reported case of MAS diagnosed and treated in Pakistan.

Keywords: Middle-aortic syndrome, Abdominal aortic coarctation, Hypertension, Lower limb claudication, Aorto-aortic bypass.

Introduction

Mid aortic syndrome or Middle aortic syndrome (MAS) is a rare entity affecting abdominal aorta in children and young adults. It is characterised by constriction of distal thoracic and/or abdominal aorta and its branches, therefore is also known as "Abdominal aortic coarctation".[1] MAS can present as hypertension or lower limb claudication or abdominal angina. MAS is characterized radiologically by severe narrowing of abdominal aorta and its branches. Most of these patients usually die due to progressive severe hypertension before the age of 35-40 if left untreated. Surgical treatment is challenging and requires careful assessment and planning for corrective surgery.

We present a case of MAS which was diagnosed and treated at Combined Military Hospital and Armed forces Institute of Cardiology Rawalpindi.

Case Report

A 20 years old male was referred to the Vascular Surgery department at Combined Military Hospital Rawalpindi with a history of uncontrolled hypertension. The patient presented with an episode of fit and unconsciousness. As a part of workup for hypertension, an echocardiogram revealed coarctation of abdominal aorta.

The patient was a well-built young man of relatively small stature. He presented with blood pressure of 300/130 mmHg with a combination therapy of captopril, amlodipine and a diuretic. Both his femoral pulses were weak and distal pulses in either of the lower limbs were not palpable. All the upper limb pulses were palpable. There was no cardiac murmur or neurological deficit on clinical examination. His laboratory workup including renal functions were within normal limit. Echocardiogram revealed ejection fraction of 47% with mild aortic regurgitation and grade 1 mitral and tricuspid regurgitation . A high resolution CT angiogram showed 70-80% segmental narrowing of abdominal aorta below the origin of the coelic trunk; 40-50% narrowing of superior mesenteric artery and 50-60% narrowing of left renal and 20-30% luminal narrowing of right renal artery. CT angiogram of cerebral circulation showed absent left internal carotid artery and posterior communicating arteries with no aneurysms.

Based on clinical picture and laboratory investigations, diagnosis of MAS was made and corrective surgery was planned. The patient had a thoracic epidural catheter placed perioperatively and operation was commenced under general anaesthesia. A thoracoabdominal incision into 6th intercostal space was made. Thoracic aorta was dissected and slung. The aortic hiatus was dissected and aortic opening enlarged. Abdominal aorta was exposed through left medial visceral rotation. Coeliac, superior mesenteric and left renal arteries were exposed. As expected, aorta was very narrow 1-2cm distal to the coeliac artery origin. Superior mesenteric, left renal arteries and infra renal aorta and common iliacs seemed to be hypoplastic (Figure-1).

Figure-1: Narrowed abdominal aorta (short broad arrow) and left renal artery (long arrow).

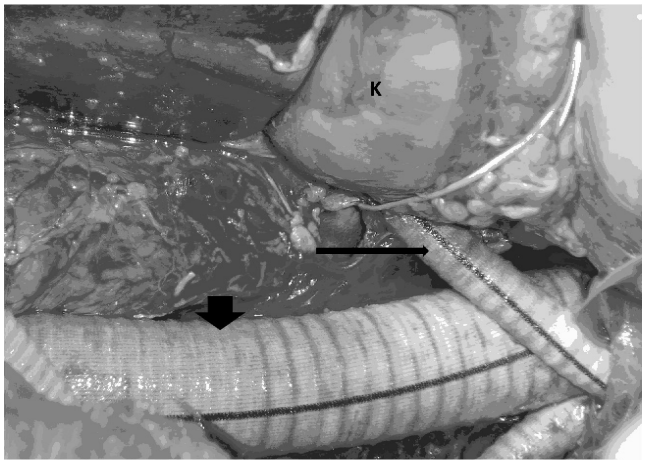

Figure-2: Jump graft (long arrow) to left renal artery from parent aorto-aortic bypass graft (short broad arrow), left kidney-K.

The Aortic vessel wall was soft and there was no evidence of perivascular fibrosis. Thoracic-abdominal aorto-aortic bypass was done using a 20mm Dacron graft. There was no pressure gradient between brachial artery and femoral artery when the anastomosis was completed, which was 100mmHg before the anastomosis. Aorto-left renal artery bypass was done using a 7mm Dacron jump graft from the parent graft (Figure-2). Flow in the left renal artery was confirmed using intraoperative Doppler Scan.

Post-operatively the patient required continuous Sodium nitroprusside infusion for a few days to keep the systolic BP less than 140mmHg. Post-operative oral antihypertensive drugs were given to control the BP. He was discharged on 12th post-operative day with a blood pressure of 120/70 mmHg with captopril and amlodipine. At six months after surgery, he is well and only requires captopril and amlodipine in twice daily doses to control blood pressure. There was no lower limb claudication or abdominal angina. Peripheral pulses were palpable and the Ankle brachial pressure index (ABPI) was 0.9, bilaterally.

Discussion

Coarctation of aorta is usually located in the thoracic aorta just distal to or sometime even proximal to ligament arteriosum. Middle aortic syndrome (MAS) is a rare entity characterized by localized narrowing of abdominal or distal thoracic aorta.[1,2] It constitutes about 0.5-2% of all the cases of aortic coarctation.[2]

Coarctation of abdominal aorta was first described by Quain in 1847,[3] however the first modern description of MAS was given by Sen in 1963[4] who named it as middle aortic syndrome.[3] Approximately 200 cases have been reported in the literature as of 2005.[1,5] Male to female ratio is 3:1.[1]

MAS may be congenital or acquired post-natally. Congenital coarctation has been thought to be due to incomplete fusion or overfusion of embryonic dorsal aortas during 4th week of gestation.[6-8] Another hypothesis implicates intra-uterine injury or infection, particularly rubella as the risk factor that precipitates aortic hypoplasia.[2] Acquired MAS is associated with neurofibromatosis, William's syndrome, Alagille syndrome, fibromuscular dysplasia, retroperitoneal fibrosis (Ormond disease), mucopolysaccharidosis, foetal alcohol syndrome and giant cell arteritides including temporal (cranial) and Takayasu arteritis.[1,9]

The most common anatomic form in congenital or idiopathic middle aortic syndrome is inter renal (19-52%), followed by supra-renal (11-40%), infra-renal (19-25%) and diffuse(12%).[1] Stenosis of the renal arteries is common (60-90%), with less common involvement of the coeliac and superior mesenteric arteries (20-40%), and infrequent involvement of the inferior mesenteric arteries.[1,10]

It usually presents as hypertension in young age group which is typically unresponsive to medical therapy. Rarely, it presents as lower limb claudication or abdominal angina due to proximal stenosis (hypertension) and distal stenosis causing hypotension.[1] The life expectancy of patients with untreated MAS is 30-40 years. The main reason of death is cardiovascular complications of progressive hypertension including cerebrovascular accidents, cardiomegaly, left heart failure and coronary artery disease.[1,11]

An initial effort to control hypertension with oral antihypertensives may be successful in a few cases with mild to moderate aortic and/or renal stenosis. The severity of hypertension is the primary indication for intervention and the factor determining procedural timing. Endovascular therapy may provide a sound minimally invasive treatment in MAS caused by discrete aortic stenoses that do not encompass the mesenteric and renal arteries.[1]

Open surgery is the primary treatment of tubular aortic narrowing associated with renovascular hypertension and visceral artery stenosis. For patients with active vasculitides surgery is not recommended in active phase of disease.[1,2,11] For children best results are achieved if definitive treatment can be delayed till they have achieved full growth.[1]

Aim of surgery is to control hypertension by improving tissue perfusion to renal, mesenteric vessels and lower limbs. Indications of surgery therefore depend upon severity of hypertension, lower limb claudication, involvement of kidneys and mesenteric ischaemia.[1,2] Surgical repair depends upon anatomy of the lesion and involvement of visceral arteries. Options include aorto-aortic bypass, patch aortoplasty, renal and mesenteric arterial reconstruction.[2] Results of surgery are good in most cases with improvement or normalization of blood pressure in 70% cases.[1]

In our case we performed a thoracic-abdominal aorto-aortic bypass for the long stenotic segment and only bypassed left renal artery, because it had greater function on differential renal scan and more stenosis. Both unilateral and bilateral renal artery revascularizations have been documented in literature.[12] Mesenteric artery bypass was not done as the patient was asymptomatic. If symptoms develop later, mesenteric artery bypass will be performed as a multi staged procedure.

Conclusion

MAS is a rare cause of uncontrolled hypertension with poor outcome if left untreated. Surgical correction is both feasible and effective in MAS. There is a need to promptly and correctly diagnose uncontrolled hypertension in young adults. Timely referral to a vascular surgeon for corrective surgery will reduce complications and mortality.

References

1. Delis KT, Gloviczki P. Middle aortic syndrome: from presentation to contemporary open surgical and endovascular treatment. Perspect Vasc Surg Endovasc Ther 2005; 17: 187-203.
2. Parvin S: Rare vascular disorders: a practical guide for the vascular specialist. Shrewsbury, UK: tfm Publishing limited; 2005.
3. Kumar S, Bury RW, Roberts DH. Unusual case of refradory hypertension: late presentation of the mid-aortic syndrome. Heart 2002; 87: E3-3.
4. Sen PK, Kinare SG, Engineer SD, Parulkar GB. The middle aortic syndrome. Br Heart J 1963; 225: 610-8.
5. Matsuno Y, Mori Y, Umeda Y, Imaizumi M, Takiya H. A successful case of ascending aorta--abdominal aorta bypass for middle aortic syndrome. Vasc Endovascular Surg 2009; 43: 96-9.
6. Cohen JR, Birnbaum E. Coarctation of the abdominal aorta. J Vasc Surg 1988; 8: 160-4.
7. Mickley V, Fleiter T. Coarctations of descending and abdominal aorta: long-term results of surgical therapy. J Vasc Surg 1998; 28: 206-14.
8. Vaccaro PS, Myers JC, Smead WL. Surgical correction of abdominal aortic coarctation and hypertension. J Vasc Surg 1986; 3: 643-8.
9. Gornik HL, Creaqer CM. Aortitis. Circulation 2008; 117: 3039-51.
10. Graham LK, Zelenock GB, Erlandson EE, Coran AG, Lindenaver SM, Standy JC. Abdominal aortic coarctation and segmental hypoplasia. Surgery 1979; 86: 519-29.
11. Connolly JE, Wilson SE, Lawrence PL, Fujitani. Middle aortic syndrome: distal thoracic and abdominal coarctation, a disorder with multiple etiologies. J Am Coll Surg 2002; 194: 774-81.
12. De Bakey ME, Garrett HE, Howell JF, Morris GC Jr. Coarctation of the abdominal aorta with renal arterial stenosis: surgical considerations. Ann Surg 1967; 165: 830-43.