A rare case of long-term graft complication in a patient of middle aortic syndrome

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ABSTRACT

Middle aortic syndrome (MAS) is rare entity described as a progressive narrowing of the aorta between the aortic arch and terminal bifurcation of the aorta. Surgical treatment is with the use of grafts for bypassing the narrow aortic segment. Rarely, graft complications may occur over long periods of time due to the axial growth of the patient and loss of tensile strength of the graft material. This is a case of MAS, presenting 34 years after initial surgery due to aneurysmal dilation of the graft, treated with redo surgery and complete recovery of the patient.

Key words: *Dacron graft, Middle aortic syndrome, Redo surgery*

Middle aortic syndrome (MAS) is a rare condition, seen typically in the pediatric population as a stenotic aorto-arteriopathy, which was originally described by Sen *et al*. [1] as a progressive aortic narrowing occurring between the arch and terminal bifurcation of the aorta. It most commonly presents in children and young adults with refractory hypertension [1]. Management of these patients usually requires surgical intervention, although antihypertensive medications may be sufficient in patients with milder diseases and no other symptoms [2,3].

Despite minimally invasive endovascular treatment options, aortoaortic bypass by the use of autologous or prosthetic grafts of the stenosed segment is the most definitive treatment option [2,3]. Long-term complications may arise later in life when the graft material undergoes degeneration as the patient grows in a linear fashion, although the reported rates of such instances are very low [4].

We report a case of MAS in a 45-year-old female who was treated with extra-anatomic bypass grafting as an 11-year-old child and presented 34 years later with a pulsatile pseudoaneurysm of the graft due to degeneration of the graft.

CASE REPORT

A 45-year-old female presented with a pulsatile swelling in the abdomen, of 3 months duration. She did not report any other complaints; there was no history of fever, extremity pain or claudication, and peripheral edema. The patient had no history of smoking, tobacco use, or alcohol intake. The patient had been diagnosed to have MAS, for which she had been treated with extra-anatomic aortoaortic bypass surgery 34 years back, using a Dacron graft and had an uneventful post-operative recovery.

She had been asymptomatic and has also had two normal, full-term deliveries since then, without any complications. On clinical examination, the patient had stable vital signs. There was a pulsatile swelling in hypochondrium, non-tender, and on auscultation, a bruit could be heard over the swelling. Peripheral pulses were normal. She was not on any medication.

The patient was investigated with contrast-enhanced computed tomography (CT), which showed a dilatation of the graft material, measuring 7 cm×4 cm (Fig. 1, Panel A and Fig. 2, Panel A and B). The most important finding in addition to the aneurysm was that there was no antegrade flow in the descending aorta and the flow in the aorta post the occluded segment was dependent on the graft in a retrograde manner.

The patient was treated with redo aortoaortic bypass surgery, wherein the old graft was dissected and replaced using a new 18 mm silver-coated graft using deep hypothermic circulatory arrest, where the body temperature is lowered to 18°C.

The patient had an uneventful post-operative course. The post-operative CT shows a normal graft and the patient was discharged on the 10th post-operative day (Fig. 1 Panel B and Fig. 2 Panel C). At follow-up, the patient was doing well, with no complaints.

DISCUSSION

MAS is a rare cause of refractory hypertension in the pediatric population, and diagnosis of this entity requires a high degree of suspicion on the part of the treating physician in any patient who presents with the triad of the thoracoabdominal bruit, decreased femoral pulses, and absence of characteristic clinical or

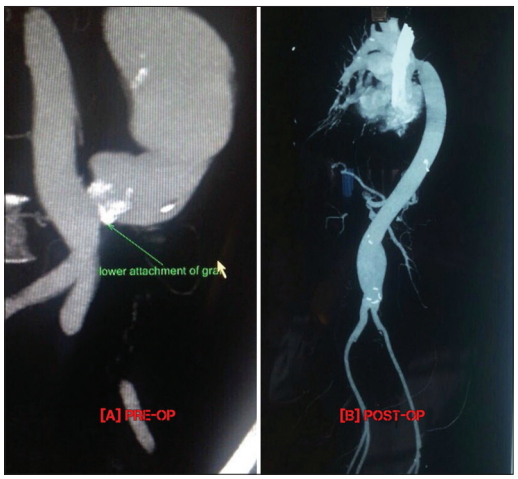

Figure 1: Contrast-enhanced computed tomography scan of the graft. Panel A: Pre-operative view; Panel B: Post-operative view after redo correction surgery

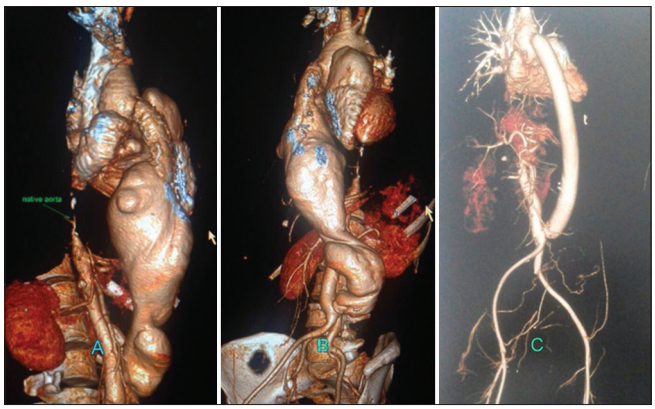

Figure 2: Volume-rendered computed tomography scan of the graft. Panel A and B: Pre-operative view, Panel C: Post-operative view after redo correction surgery

radiological features of other thoracic coarctations [5]. Surgical management provides the best outcome for the patient, with adequate control of hypertension and end-organ functioning.

This entity forms <2% of all aortic stenosis [6] and classically presents as refractory hypertension proximal to stenosis and hypotension distally, along with symptoms of nosebleeds, severe headaches, chest pains, lower limb claudication, and even cardiac failure [2]. Left untreated, this condition can progress to severe complications such as coronary artery disease, congestive heart failure, left ventricular hypertrophy, cerebrovascular accidents, hypertensive encephalopathy, and retinopathy, with most patients succumbing to the disease before 40 years of age [5]. On examination, absent femoral pulses and an abdominal bruit may be evident, depending on the degree of stenosis [2].

The etiology of this disease remains idiopathic for the majority of cases, although it is postulated to be due to an embryological defect, or associated with Mendelian disorders such as neurofibromatosis Type 1, Alagille syndrome, and Williams' syndrome [2]. The presentation of this disease may mimic other syndromes or present along with them, such as Takayasu arteritis, neurofibromatosis, fibromuscular dysplasia, retroperitoneal fibrosis, mucopolysaccharidosis, and congenital vascular abnormalities [2,3].

This disease process most frequently involves the abdominal aorta, along with visceral arteries such as renal (>80%), splanchnic (70–50%), superior mesenteric (60–30%), and celiac (60–20%), but the inferior mesenteric is almost never involved [2,3]. Based on the involvement of renal arteries, this syndrome can be classified into four major types: Type I - suprarenal coarctation and renal artery stenosis; Type II - infrarenal coarctation and renal artery stenosis; Type III - suprarenal coarctation and normal renal arteries; and Type IV - infrarenal coarctation and normal renal arteries [5]. Only the first three types require intervention for renal blood flow. Our patient had a stenosed segment extending from the descending aorta to the abdominal aorta with no collaterals.

The gold standard for diagnosis is intra-arterial digital subtraction angiography [7], which not only confirms the diagnosis but also delineates the sites of stenosis before surgical intervention. The stenosis may also be visualized by Color Doppler, magnetic resonance angiography, and computed tomographic angiography.

Treatment of this modality depends mainly on the severity of the disease, with mild cases being managed by medical treatment alone, but invasive procedures are required for more severe disease [2,3]. Various surgical procedures including aortoaortic bypass grafting, graft vascular replacement, patchy angioplasty, and renal autotransplantation have been described, of which prosthetic or autologous venous surgical arterial bypass grafting remains the treatment of choice [2]. Endovascular procedures such as percutaneous transluminal angioplasty with or without stenting have also been tried in some patients, but the reported rates of procedure failure are higher in this group [2].

Surgical correction in a pediatric patient carries the risk of graft stenosis or pseudoaneurysm formation as the patient grows in an axial fashion. The growth of the axial skeleton is comparatively less after the age of 10 years, and the growth of the aorta is mainly seen above and below the level of the graft, with the minimal lengthening of the bypassed segment [4]. Therefore, it is recommended to put the surgical treatment on hold till the patient has attained complete growth and manage the patient with antihypertensive medications [4]. To prevent these complications, measures such as the use of an end-to-side aortoaortic bypass, oversizing the length and diameter of the bypass, and use of crimped Dacron grafts may be undertaken [4,8]. The extra-anatomic axillofemoral bypass may be done if other procedures fail to provide relief as a last resort measure [9]. Some authors have recommended a follow-up with CT at the interval of every 5 years in patients with aortic surgery as prosthetic materials are subjected to wear and tear, similar to autologous grafts, which may then lead to graft degeneration [4,10]. In our extensive review of the published literature, few cases of non-anastomotic aneurysmal graft degeneration have been reported, and out of them, only one case was of a patient with MAS, who presented years later as an adult with an aortic graft-enteric fistula and went into pulseless activity during the redo operation [4,11-13].

CONCLUSION

MAS is a rare cause of unresponsive hypertension in children and treatment requires multimodality management, with surgical interventions forming the backbone of the treatment. Prosthetic grafting done in childhood may experience wear and tear as the patient grows and may show complications of graft material degeneration such as pseudoaneurysms. Periodic follow-up of the patient with a multispecialty team of experienced physicians is required to monitor a patient with these rare complications of this uncommon disease process.

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