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Case Report / Olgu Sunumu

The fate of a thoracoabdominal aortic bypass graft 10 years after surgery in a child with the middle aortic syndrome

Orta aort sendromlu bir çocukta torakoabdominal aortik baypas greftinin10 yıl sonraki akıbeti

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ABSTRACT

A 13-year-old boy who underwent thoracoabdominal aortic bypass when he was three years old for middle aortic syndrome was admitted with fatigue and need for an increased dose of antihypertensive mediations. The graft was patent, but there were stenoses at the juxta-proximal and juxta-distal anastomosis sites. A partial benefit was gained with endovascular stenting. Although postponement of surgery, until the child reaches full growth is preferred, surgery remains the inevitable treatment of choice in patients with middle aortic syndrome. In contrary, it is important to use the graft as large as possible during the initial operation to avoid patient-graft mismatch in the future.

Keywords: Aortic diseases; hypertension; thoracoabdominal aortic bypass; vascular patency.

ÖZ

Orta aort sendromu nedeniyle üç yaşında torakoabdominal baypas uygulanmış olan 13 yaşındaki bir erkek çocuk, çabuk yorulma ve antihipertansif tedavi dozundaki artış gerekliliği nedeniyle başvurdu. Greft açık olmakla birlikte, proksimal ve distal anastomoz yakınında darlıklar izlendi. Endovasküler stentleme ile kısmen fayda sağlandı. Çocuğun tam büyüme potansiyeline ulaşmasına kadar cerrahi tedavinin ertelenmesi tercih edilse de, orta aort sendromlu hastalarda cerrahi kaçınılmaz bir tedavi seçeneğidir. Buna karşın ilk ameliyat sırasında mümkün olan en geniş greftin kullanılması, gelecekteki hasta-greft uyumsuzluğundan kaçınmak için önem arz etmektedir.

Anahtar sözcükler: Aort hastalıkları; hipertansiyon; torakoabdominal aortik baypas; vasküler açıklık.

Middle aortic syndrome (MAS) is a rare and progressive congenital disease characterized by segmental narrowing of the abdominal aorta and/or distal descending aorta.[1-4] In this article, we discuss the long-term durability of a thoracoabdominal aortic bypass graft in a child with MAS in which endovascular stenting showed a partial benefit due to the outgrowth.

CASE REPORT

A three-year-old boy weighing 13 kg was referred with upper extremity hypertension (130/80 mmHg) in 2006. Contrast-enhanced computed tomography (CeCT) revealed luminal irregularities extending from the supra-diaphragmatic level to the supra-renal segment of the abdominal aorta involving visceral branches except renal arteries (Figure 1a). With the diagnosis of MAS, he previously underwent descending thoracic-to-infrarenal abdominal aortic bypass grafting using an 8 mm ringed polytetrafluoroethylene tube graft without visceral revascularization. Until one year before current admission, he was normotensive without anti-hypertensive medication.

In 2016, (13 years old, weight 35 kg), he was referred with fatigue and need for an increased dose of antihypertensive medications (enalapril 5 mg bid, amlodipin 5 mg bid) for the last six months. On physical examination, he had upper extremity hypertension (150/95 mmHg). Along with filiform femoral pulses, upper-to-lower extremity blood pressure difference was 60 mmHg. The ankle-brachial indices (ABI) were 0.5. He was asymptomatic for mesentery ischemia.

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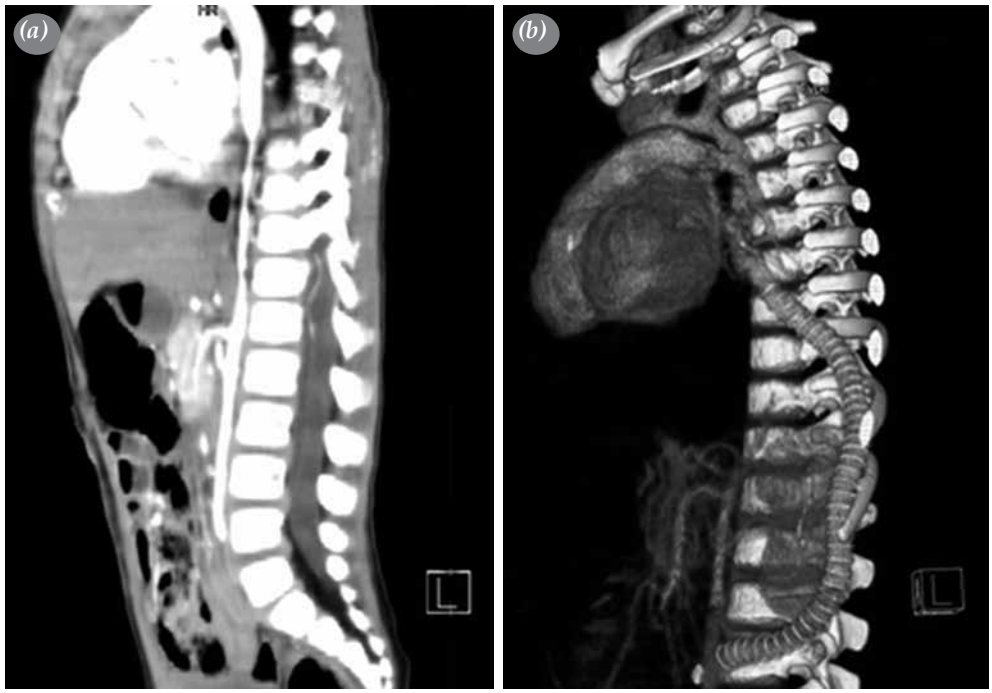

Figure 1. Computed tomography images. **(a)** Contrast-enhanced computed tomography of a three-year-old boy with congenital middle aortic syndrome. **(b)** Three-dimensional computed tomography scan of an 8 mm polytetrafluoroethylene graft 10 years after thoracoabdominal bypass grafting.

In laboratory, he had normal hepatic (aspartate aminotransferase: 38 U/L, alanine aminotransferase: 22 U/L) and renal (creatinine: 0.54 mg/dL) function. In three-dimensional reconstruction of the CeCT, the descending and the infa-renal abdominal aortic diameters were 15 mm and 12.5 mm, respectively. The graft was patent, but there were stenoses at the juxta-proximal and juxta-distal anastomosis sites (Figure 1b). During catheterization, stenoses were found to be 70% near the proximal and 60% near

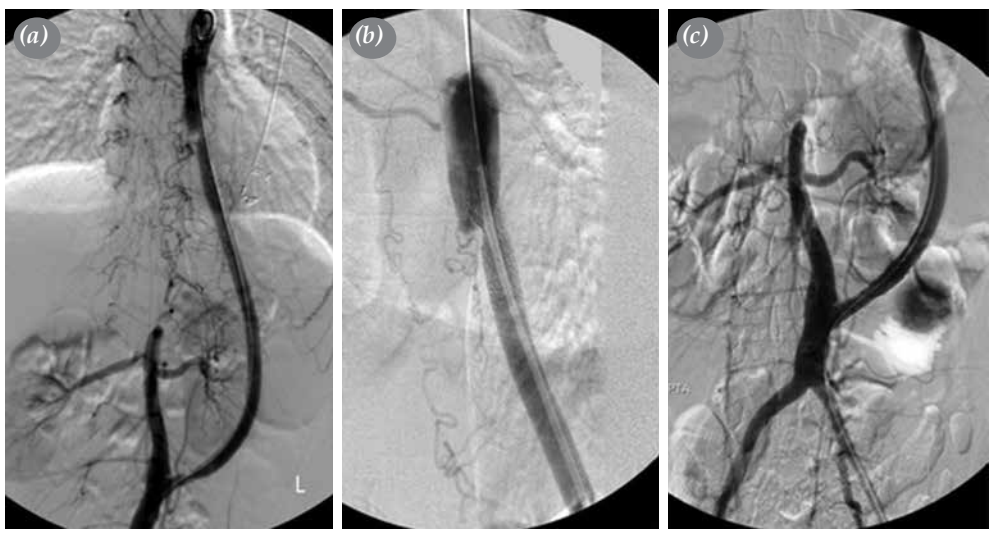

Figure 2. Conventional angiography images. **(a)** Patent tube graft with stenoses at proximal and distal site while he was 13 years old. **(b)** Stenting and subsequent balloon dilation of the proximal stenosis which was partially relieved due to proximal propagation of the disease. **(c)** Stenting of the distal anastomosis site stenosis.

the distal anastomosis sites. Additionally, along with a 30% stenosis of the right renal artery, there was intensive collateral vessels with total obstruction of the celiac and superior mesenteric arteries (Figure 2a).

In the endovascular suite, the proximal stenosis was stented (9×30 mm balloon-expandable stent, Assurant-Cobalt, Medtronic, USA), but a 30% residual stenosis remained. The residual stenosis was unable to be relieved through balloon dilation (9 mm, Mustang, Boston Scientific, USA), which was thought to be due to the proximal propagation of the disease leading to a stiffness of the anastomosis site (Figure 2b). The distal stenosis was successfully relieved using a 10×40 mm, self-expandable stent (Natick, Boston Scientific, USA) (Figure 2c). In the post-interventional period, while the upper-to-lower extremity blood pressure difference decreased to 45 mmHg, the ankle-brachial indices increased to 0.7. The upper extremity hypertension was not adequately controlled; thus, the patient was listed for a redo thoracoabdominal bypass grafting with a higher diameter graft.

DISCUSSION

Although postponement of surgery to avoid redo surgery in the future and gaining time through endovascular interventions, until the child reaches full growth is preferred, surgery remains the absolute and, sometimes, the inevitable treatment of choice in patients with MAS with good long-term results.[1-4] In comparison to surgery, the long-term success of medical and/or endovascular treatment has of little value in which, freedom from reintervention for the endovascular and surgery groups for one year were 58% and 83%, respectively.[2,4] Additionally, while freedom from reintervention was reported to be 33% for the endovascular group at five years, it was 72% in the surgery group for 10 years.[2]

In the article of Hetzer et al.,[1] 14 children with a mean age of 6.7 years with the diagnosis of MAS were operated through extra-anatomic bypass grafting (descending-to-abdominal aortic bypass in two patients and ascending-to-abdominal aortic bypass in 12 patients). There was no reoperation or endovascular intervention for graft-related issues. In our patient, the proximal anastomosis site stenosis was unable to be relieved, even after balloon dilation mainly due to the proximal propagation of the disease, leading to a substantial stiffness at the anastomosis site. Stanley et al.[3] performed thoracoabdominal bypass grafting in 26 of 53 patients with MAS of which one patient required reoperation for proximal anastomosis site narrowing nine years after initial operation which resembled with the current case. In our opinion, when the progressive nature of the disease was considered, as seen in the current case, reconstruction of the proximal anastomosis at the ascending aorta that led the proximal anastomosis to be hold off the descending aorta[1] had a substantial benefit for the long-term patency of the extra-anatomic bypass graft.

For relief of upper extremity hypertension, thoracoabdominal bypass grafting was shown to be superior compared to other treatment modalities.[1-4] Our case was normotensive without antihypertensive medication through a period of nine years. Thus, the current case emphasized the long-term effectiveness of the thoracoabdominal bypass grafting on the control of hypertension, which led to a substantial period to pass in an asymptomatic and normotensive state.

The literature comprises successful endovascular interventions, both as an initial and a postsurgical procedure in patients with MAS.[2-4] In the current case, the distal aortic flow restored with an 8-mm graft was partially effective in lowering upper extremity hypertension which, in our opinion, was related to the patient-graft mismatch. The adequacy of the size of the graft with the patient's growth potential was emphasized.[1,3] Among 26 patients with thoracoabdominal bypass grafting, there was only one patient who received graft re-replacement seven years after surgery due to the outgrowth of the initial graft.[3] The diameter of the graft used 10 years ago in our case was 8 mm. Due to the concern about the graft to be too large which may have resulted in graft thrombosis, an 8-mm graft was used which was compatible with the recommendation of Stanley et al.[3] In the cohort of Hetzer et al.[1] with a mean age of 6.7±3.7 years, while the diameter of the graft used was 14 mm in 11, it was 12 mm in one patient, but the anthropometric data of the cohort were not reported. Additionally, none of the patients in their cohort required re-replacement due to outgrowth. The current case emphasized the importance of the necessity of an age-independent initial operation with the most appropriate sized-graft to avoid such a patient-graft mismatch.

In conclusion, thoracoabdominal bypass grafting is the most effective therapeutic modality in the treatment of middle aortic syndrome. In contrary, during the initial operation the most appropriate-sized graft should be chosen to avoid from the graft outgrowth in the future, since, in the event of graft stenosis, the benefit gained with an endovascular intervention would be limited.

Declaration of conflicting interests

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