Ascending to Descending Aorta Bypass for Middle Aortic Syndrome

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Though stenoses of the descending aorta and its branches are seen with congenital anomalies or systemic inflammation, occlusion of the descending aorta is extremely rare. A patient with an occluded hypoplastic descending thoracic aorta required re-operation because of graft failure between the descending thoracic aorta and the infrarenal abdominal aorta. The etiology of the aortic occlusion in this case is unknown, but inflammation, such as Takayasu disease, is speculated. (Circ J 2007; 71: 1162–1163)

Key Words: Bypass operation; Descending aorta; Inflammation; Occlusion

Middle aortic syndrome is rare vascular anomaly, which involves long segment narrowing of the descending thoracic and abdominal aorta. The most common clinical manifestation is severe uncontrolled hypertension. We present a successful re-operation by the ascending to descending aorta bypass for middle aortic syndrome patient.

Case Report

A 52-year-old woman complained of headache and easy fatigability of the lower extremities and had poorly controlled high blood pressure. Both femoral pulses were present but weak. Her blood pressure was 180/120 mmHg in the brachial artery and 108/76 mmHg in the thigh. Routine laboratory data were normal, with a blood urea nitrogen level of 12 mg/dl and a creatinine level of 0.6 mg/dl. C-reactive protein was negative. Twenty-seven years previously, she had undergone bypass grafting for Takayasu aortitis at another hospital. According to the operation record, an 8-mm-diameter prosthetic tube graft was bypassed from the descending thoracic aorta to the infrarenal abdominal aorta and it was passed through the aortic hiatus through a left thoracotomy and midline laparotomy.

In the present admission computed tomography scanning showed marked diffuse narrowing of the descending thoracic aorta with severe calcification (Fig1). The diameter of the aorta was 21 mm at the distal arch, 6 mm at the diaphragm level, 11 mm at the superior mesenteric artery level and 20 mm at the aortic bifurcation. The supradiaphragmatic descending aorta was completely occluded. The prosthetic graft was also severely atherosclerotic, but pseudoaneurysm formation was not detected on either the graft itself or at both anastomoses. The ascending aorta was normal and the both internal thoracic arteries were dilated. The right kidney was atrophic and non-functional. Preoperative angiography revealed total occlusion of the aorta from the mid-descending thorax to proximal to the celiac artery and multiple stenoses of the bypass graft. Blood flow to the celiac artery, the superior mesenteric artery, and the left renal artery was supplied retrogradely by the stenotic bypass graft. The right renal artery was occluded. No abnormality was detected in the ascending aorta, aortic arch, or supracervical branches. These findings indicated that her symptoms were related to the multiple stenoses of the graft, in addition to small size of the bypass graft itself. We decided to replace the graft.

The operation was performed through a midline sternotomy and pararectal retroperitoneal approach. The ascending aorta was side-clamped and a 16-mm-diameter prosthetic tube graft was anastomosed in an end-to-side fashion. The graft was passed through a small incision in the left lateral portion of the diaphragm. Adhesion of the former anastomosis, just distal to the left renal artery, was so dense that the distal end of the new graft was anastomosed at the aortic bifurcation, which appeared to be a normal part of the aortic wall. The patient’s postoperative course was eventful. Her hypertension was controlled without antihypertensive agents (brachial blood pressure of 110/68 mmHg and thigh blood pressure of 100/62 mmHg). At 1-year follow-up,
up, magnetic resonance angiography showed that the new graft was patent and there were no abnormalities of the anastomoses (Fig 2). She was doing well 3 years after operation, without headache or claudication.

Discussion

Long segment narrowing or hypoplasia of the descending aorta is a rare vascular anomaly. The aortic narrowing located in the descending thoracic aorta, abdominal aorta, or both is called “middle aortic syndrome” by Sen et al. Etiology of this vascular disease is not well known. Congenital, inflammation, developmental disorder, and infection are thought to be some of the causes. Panayiotopoulos et al said that middle aortic syndrome is the correct term because it describes the clinico-anatomical entity irrespective of its etiology and pathogenesis.

Though hypoplasia has been reported in all portions of the thoracic and abdominal aorta, total occlusion of the descending aorta is extremely rare. It was hard to explain the present case with a single etiology. The patient was diagnosed as having Takayasu disease at the time of her first bypass operation, but it was not a definitive diagnosis. Congenital coarctation at an atypical site and coarctation caused by Takayasu disease are indistinguishable on computed tomography. No other systemic change, such as neurofibromatosis, was detected preoperatively. At operation, we did not take specimens from the anastomosis sites, where inflammation was not supposed to occur. In Takayasu disease, the descending aorta is usually free of atheromatous changes. The fact that the present patient's descending thoracic aorta was severely calcified, with an almost normal ascending aorta, aortic arch, and supracervical branches, suggests inflammation rather than atherosclerotic change in the descending aorta. Though we do not have enough evidence to make a diagnosis of Takayasu disease, considering the patient's gender, age at first involvement, and the current findings, we speculate that this case was caused by inflammation such as Takayasu disease on a congenitally hypoplastic descending thoracic aorta. According to the classification of Takayasu disease, the present patient was type III: descending thoracic aorta, abdominal aorta, and/or renal arteries involvement. Inflammation thickens the arterial wall, leading to stenoses or occlusion, but the diameter of the artery does not change drastically. In the present case, the diameter of the descending thoracic aorta tapered to only 6 mm at the level of the diaphragm. It is thought that the occlusion of the aorta was induced because of both the hypoplasia of the descending thoracic aorta and Takayasu disease.

At re-operation in the presence of a failing but patent graft from the descending to abdominal aorta through a previous left thoracotomy and midline laparotomy, the descending aorta was not suitable as the inflow site of the bypass graft. The patient's descending aorta was small and calcified severely along the entire length. However, in the preoperative examinations the ascending aorta appeared normal without atherosclerotic change and it was easy to expose the distal abdominal aorta by the pararectal retroperitoneal approach because these portions were not involved in her type of Takayasu disease. Although axillofemoral bypass was another option, we chose ascending-descending aorta bypass for 2 reasons. First, long-term patency is expected and second, the afterload of the left ventricle is reduced more effectively. At operation, both the ascending aorta and aortic bifurcation were normal macroscopically. We left the failed graft untouched because there was no aneurysmal formation. In addition, it seemed difficult and hazardous to clamp and suture an atherosclerotic graft with heavy adhesion around the anastomoses. Regarding the timing of operation, she was in the “burned-out” phase of Takayasu disease according to the laboratory data, which was necessary and very important for avoiding complications.

References