Extra-anatomic Aortic Bypass for the Management of Mid-Aortic Syndrome Caused by Takayasu arteritis

Jae Kwang Yun, M.D., Joon Bum Kim, M.D., Ph.D.

Lower limb ischemia caused by multiple stenosis of the thoracoabdominal aorta is one of the rare clinical manifestations of Takayasu arteritis. The optimal management of such mid-aortic syndrome related with Takayasu arteritis has not been established to date. Here we report a case of extra-anatomic aortic bypass through minimally invasive techniques to treat lower limb ischemia caused by Takayasu arteritis.

Key words: 1. Takayasu arteritis  
2. Aorta  
3. Bypass surgery

CASE REPORT

A 38-year-old woman was admitted due to claudication of the legs that had been sustained for 2 years. Eight months ago, she was diagnosed as having Takayasu arteritis during the work ups for chronic headache in primary clinic. Immunosuppressive medication with methotrexate was followed due to glucocorticoid resistance. Two months ago, embolic infarction of the kidney and the spleen resulted in anticoagulation therapy with warfarin. Her past medical history included hypertension requiring medication 10 years ago and a surgical history of appendectomy 9 years ago. On admission, physical examination revealed that the pulse over the right femoral area was absent. Asymmetrical systolic blood pressure of 142 mmHg in the arms and 88 in the right legs and 94 mmHg in the left leg was found. Thus, ankle-brachial index was 0.62 in right and 0.66 in left. The findings obtained from a central nervous system examination were unremarkable. The patient’s C-reactive protein level and erythrocyte sedimentation rate were within normal range. Three-dimensional computed tomography (CT) revealed multifocal severe luminal narrowing of descending thoracic and abdominal aorta (Fig. 1A, B). In addition, significant stenosis of the right carotid artery, brachiocephalic trunk, proximal celiac trunk ostium and total occlusion at left subclavian artery ostium with collateral development were observed. Whole body positron emission tomography-CT revealed no hypermetabolic lesions in the arterial system indicating that the vasculitis was not active. Cardiac evaluations including echocardiography and coronary angiography revealed normal cardiac function (left ventricular ejection fraction, 60%) without any stenotic lesions in coronary arteries. In order to relieve ischemic symptoms of the lower extremities, the patient underwent axillo-femoral bypass surgery in the right side using a 10 mm-ringed polytetrafluoroethylene graft (Fig. 1C). After the surgery, the difference in arterial blood pressure between the upper and lower extremities disappeared, and the symptom of claudication was relieved. Anticoagulation therapy was started on the first...
Postoperatively, vital signs were stable and the arterial blood pressure in the upper and lower extremities equalized. The patient was transferred from intensive care unit to general ward at postoperative day 1 and was discharged without complication at 8 days. During her 5 months of follow-up, the patient remained completely free of limb claudication with excellent exertional tolerance. Postoperative CT showed excellent graft configuration without architectural distortion (Fig. 3). Then the patient was given warfarin for 2 months with continued immunosuppressive medication.

**DISCUSSION**

In the early phase of Takayasu arteritis, systemic symptoms such as fatigue, weight loss, and low-grade fever usually present which indicate active inflammation in the body. As the disease progresses, evidence of vascular insufficiency becomes clinically apparent due to dilation, narrowing, or occlusion of the proximal or distal branches of the aorta. In advanced cases, occlusion of the vessels to the extremities may result in ischemic ulcerations or gangrene [1]. Collateral circulation usually develops in the areas involved by vasculitis because of the chronic nature of the disease [2].

Treatment of Takayasu arteritis should therefore be tailored in individual patients depending on the phase [1,3]. During the active phase of the disease, steroids have been used to improve systemic inflammatory symptoms within a few days.
Fig. 2. (A) An operative photograph showing an incision of upper hemi-sternotomy and mini-laparotomy. The ascending aorta-to-infrarenal abdominal aorta bypassing with the use of a Gelweave graft was performed. (B) The graft was anastomosed proximally to the ascending aorta and (C) distally to the infrarenal abdominal aorta.

Fig. 3. Follow-up computed tomography showing a widely patent bypass graft without any structural abnormality.

to weeks. Immunosuppressive drugs such as methotrexate or cyclophosphamide are usually added due to steroid resistance or relapse of the disease during steroid dosage reduction. In the chronic stage, revascularization of the affected organ may be needed either by surgery or endovascular treatment. Although endovascular treatment is less invasive than surgical repair, restenosis in the treated lesions still remains as a common complication [4]. Surgical revascularization has shown superior long-term artery patency than the endovascular treatment with the cost of increased risks of procedural complications [5]. Importantly, it has been reported that the revascularization therapy should be avoided during an active phase of Takayasu arteritis to avoid anastomotic dehiscence or restenosis. According to Fields et al. [6], performing surgery in patients with active-stage of Takayasu arteritis increases the risk of early graft revision and the procession of symptomatic disease in
other arterial beds. It is also important to maintain graft patency, as frequent vascular restenosis may lead to serious complications and a decrease in quality of life. Park et al. [7] reported restenosis occurred in 31.7% of Takayasu arteritis patients after revascularization intervention and a lower restenosis rate was observed when post-interventional immunosuppressive treatment was implemented.

In the present case, CT findings revealed multifocal luminal narrowing of descending thoracic and abdominal aorta constituting namely ‘mid-aortic syndrome’ which is not usual manifestation of Takayasu arteritis. Two years following the axillo-femoral bypass surgery, occlusion of bypass graft caused recurred claudication, and this let us to consider more durable solution to relieve ischemic symptoms. Considering the higher probability of graft occlusion in long and small bypass conduits, especially in arteries with projected poor distal runoff, we should have chosen a larger bypass conduit that are connected to larger arteries distally. An important consideration in the plan of aorta bypass was the site of proximal anastomosis. We undertake the proximal anastomosis on the ascending aorta for excellent anatomic exposure, easy bleeding control and reducing afterload. Anatomic repair surgery such as thoracoabdominal aorta replacement could have been another consideration, however, it was regarded too extensive a surgery carrying high risks of morbidity only to treat limb ischemia. Although the effect of anticoagulation therapy in Takayasu arteritis after surgical treatment was not reported in previous literature, we used short term anticoagulation therapy to maintain graft patency. Matsuno et al. [8] reported an extra-anatomic aortic bypass surgery which involved inserting a bypass graft between the ascending aorta and the infra-renal abdominal aorta to treat the mid-aortic syndrome. An important issue in this technique is the creation of a tunnel through which the graft passes through the diaphragm, lesser omentum and transverse mesocolon. Since the route allows the shortest course of the bypass graft, it can be advantageous to improve the graft patency while minimizing the risk of potential injuries of adjacent visceral organs. Basic concepts of the procedure performed in the present case go along with that of case of Matsuno et al. [8] with the only exception of ‘minimally invasive approach.’ Cosmetic issue was particularly important in the present case, since this patient was unmarried young woman who was strongly willing to minimize surgical incision for cosmetic purposes. With the use of current minimally invasive surgical techniques, such extra-anatomic bypass surgery of the aorta could be successfully performed with excellent surgical outcomes.

In conclusion, we report a case of extra-anatomic aortic bypass through minimally invasive techniques to treat lower limb ischemia caused by mid aortic syndrome related with Takayasu arteritis.

**CONFLICT OF INTEREST**

No potential conflict of interest relevant to this article was reported.

**REFERENCES**