A Case of Takayasu’s Arteritis with Total Occlusion of the Abdominal Aorta Treated Only with Oral Medication for More Than 40 Years

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Takayasu’s arteritis affects most commonly young women, often causing stenotic and occasionally dilated lesions of the medium-to-large-sized arteries with an acute inflammatory signs and symptoms. Here, we report a rare case of Takayasu’s arteritis with total occlusion of the abdominal aorta, which was successfully treated with medication alone and asymptomatic over 40 years. Magnetic resonance imaging revealed total occlusion of the abdominal aorta and stenosis of the right carotid artery. This is the first report of a very rare case of Takayasu’s arteritis, in which medical treatment only was successful against total occlusion of the abdominal aorta.

Key words: Takayasu’s arteritis, long-term, total occlusion of the abdominal aorta

Introduction

Takayasu’s arteritis causes lesions extended narrowing of the aorta with symptoms of acute inflammation, aortic coarctation affecting the cardiovascular system. It is therefore considered to be important to perform aggressive immunosuppressive therapy and early revascularization. Here, we report a rare case of Takayasu’s arteritis with total occlusion of the abdominal aorta, which was treated with medication alone and asymptomatic over 40 years.

Case Report

In 1971, a 30-year-old woman who had lost consciousness for 1 min and was found with blood pressure of 190/120 mmHg, and referred to our hospital after a second bout of feeling faint. The patient had begun to feel fatigue in her legs at the age of 15 and had a history of intermittent claudication when she came to our outpatient clinic. The patient’s right and left arm blood pressure were 182/102 mmHg and 180/108 mmHg, respectively. Renovascular hypertension was suspected based on the presence of abdominal bruits in this case, but the patient’s plasma renin activity was within normal limits (2.7 ng/mL). Renal function was normal (Cr 0.7 mg/dl) and no ischemic change was observed on electrocardiogram. The patient’s blood pressure was reduced to 144/88 mmHg with administration of antihypertensive medications, including trichlormethiazide, reserpine, and bethanidine sulfate. Aortic angiography performed in 1971 revealed aortic stenosis below the bifurcation of the renal arteries and severe stenotic lesions on the left renal artery with collateral arteries extending to the slightly atrophic left kidney (Fig. 1). However, the renal pelvises appeared normal on pyelography. The cardiothoracic ratio of the patient’s chest X-ray was 46%, and electrocardiography showed a high voltage (RV5 + SV1 = 4.3 mV). While erythrocyte sedimentation rate was elevated to 49 mm/h, C-reactive protein was negative. Based on these findings, a diagnosis of Takayasu’s arteritis was made. In 1975, bruits were detected over the bilateral carotid arteries as well as the left orbit. Furthermore, the right brachial artery showed attenuated pulse. The patient was treated with antihypertensive medication alone and has no difficulty when walking on flat surfaces, but a small amount of intermittent claudication while walking up stairs.

In 2009, magnetic resonance imaging (MRI) was performed and revealed right carotid artery stenosis and no visualization of the left subclavian artery (Fig. 2A). Further, total occlusion of the abdominal aorta (lesion length: 3 cm) and the left renal artery with atrophy of the left kidney were observed (Fig. 2B). Recently, in 2012, three-dimensional reconstitution of recent MRI findings demonstrated clear three-dimensional structure in which collateral circulation that goes down along the occluded aorta was observed (Fig. 3).

In 2015, the recent status of the patient was as follows. The blood pressure of the upper extremities was...
The systolic blood pressure of the lower extremities was 78 mmHg (right), 68 mmHg (left). Recent echocardiography showed no evidence of left ventricular hypertrophy (interventricular septal thickness 9 mm, left ventricular posterior wall thickness 9 mm), and systolic function was preserved (ejection fraction 68%). Erythrocyte sedimentation rate had been elevated and sustained (between 29 and 62 mm/h). Serum matrix metalloproteinase-3 concentration was within normal range (35.7 ng/ml). Up to now, plasma renin activity (less than 1.0 ng/mL/hr) and plasma aldosterone concentration have been within normal limits (51.3–110 pg/mL), which strongly supports the lack of severe renovascular hypertension.

Despite these severe arterial disorders, the patient has led a daily life for more than 40 years without any symptoms.

**Discussion and Conclusion**

Takayasu’s arteritis is relatively common in Asian countries, and can be subdivided into an early phase (i.e., systemic inflammatory, pre-pulseless) and a late phase (i.e., occlusive, pulseless). Diagnosis is difficult in the early phase because patients present with nonspecific signs and symptoms of systemic inflammatory illness, such as fever and myalgia, while many patients in the late phase (occlusive stage) present with symptoms of arterial manifestations rather than systemic inflammatory components. The variable nature of the presentation of this disease as well as the lack of symptoms in 30%–50% of cases reduces the likelihood of making an early diagnosis.
Classically, Takayasu’s arteritis affects most commonly young women, often causing stenotic and occasionally dilated lesions of the medium-to-large-sized arteries with an acute inflammatory signs and symptoms. However, it seems that middle-aged patients with chronic ongoing inflammation are being found more often lately. Surgical treatment is necessary without delay for the lesions such as aortic stenosis, aortic regurgitation, renal arterial stenosis, and aneurysms, leading to a lethal cardiovascular damage. However, there are some cases which follow a benign outcome without surgical treatment, in which ischemic lesions are improved by the development of collateral circulation. Although the patient described here came to our clinic complaining of faintness and severe hypertension and has been treated with antihypertensive medications alone, she has been free of ischemic episodes for more than 40 years. In addition, lack of severe renovascular hypertension would have contributed to the favorable clinical outcome of this patient. Whether or not Takayasu’s arteritis develops into fatal conditions such as arterial occlusion due to the progression of acute stenotic (dilative) lesion depends on the balance between the progression of the stenotic lesion and the development of compensatory collateral circulation. As in the present case, there are some cases that can be followed up with medication alone when the development of the collateral circulation sufficiently compensates for the progression of the stenotic lesion.

There have been no previous reports of patients with Takayasu’s arteritis with total occlusion of the abdominal aorta and right renal artery, who was treated with medication alone and was able to spend such an extended period almost asymptomatic like the present case. Collateral circulation that goes down along the occluded aorta was observed on MRA, which indicates a possibility of compensation being made by the development of this kind of collateral circulation. Besides this, it is also possible to
assume that several bypasses including collateral circulation such as internal thoracic arteries and iliac arteries developed outside of the imaging area. This is the first report of a very rare case of Takayasu’s arteritis, in which medical treatment only was successful against total occlusion of the abdominal aorta.

Disclosure Statement
All authors have no conflict of interest.

References