

## Case Report

# Surgical Treatment of Multivessel Lesions in Takayasu's Arteritis: Report of a Case

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### Abstract

Takayasu's arteritis (TA) is a chronic vasculitis involving the aorta and its main branches, the pulmonary arteries, and the coronary tree. Here we report a case of TA complicated by severe stenosis of the left coronary ostium with multivessel brachiocephalic involvement. A combination of these abnormalities could complicate underlying illness in patients, posing an increased risk of surgical morbidity. Simultaneous surgical treatment of the ascending aorta to left carotid artery bypass and coronary artery bypass using the great saphenous vein were performed. We discuss the choice of simultaneous surgery and the options for surgical treatment of complicated lesions due to TA.

**Key words** Takayasu's arteritis · Coronary occlusion · Multivessel · Brachiocephalic surgery

### Introduction

Takayasu's arteritis (TA) is a rare chronic inflammatory disease of unknown cause that affects the large arteries. It most often affects the aorta and its primary branches. Its etiology is still not fully known. The majority of cases are seen in Asia and Africa. Coronary involvement has been reported in 6%–19% of cases. Most of the coronary lesions are thought to be secondary to the extension of aortitis,<sup>1,2</sup> and is usually limited to the ostia.<sup>2</sup> We herein report a case of TA complicated by severe stenosis of the left coronary ostium with multivessel brachiocephalic involvement. We discuss the choice of simultaneous bypass operations and the options for

surgical treatment of complicated lesions due to TA. After the surgery, the patient was followed up for 2 years, and she has remained completely asymptomatic with good exertional tolerance.

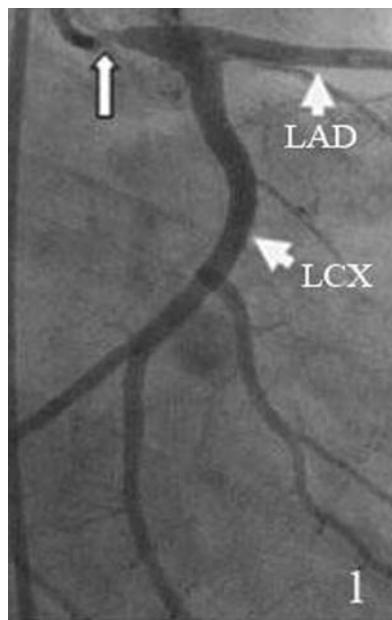
### Case Report

A 30-year-old local woman was admitted with retrosternal chest pain that had radiated to the left arm on exertion for 1 month. During the past 15 days, however, her pain had increased in intensity and duration. Three years prior to presenting with the chest pain symptoms the patient had suffered from visual disturbance and syncope, had been diagnosed with TA, and received steroid therapy. The patient also had experienced four transient ischemic attacks. She had no history of smoking, alcohol consumption, or use of illicit drugs. The patient also had no family history of coronary artery disease, cardiomyopathy, or arthritic disease. On admission, physical examination with the patient in the supine position revealed that the pulse over the left arm was absent. She had asymmetrical blood pressure of 100/60 mmHg in the left arm and 135/80 mmHg in the right arm. A systolic bruit was registered in the left cervical area, and heart sounds were normal on auscultation. The findings obtained from a central nervous system examination were normal. On the day of admission, the patient's C-reactive protein level (9.58 mg/dl) and erythrocyte sedimentation rate (32 mm/h) were high. The results of serology tests for syphilis, systemic lupus erythematosus, and rheumatoid arthritis were negative.

The patient's electrocardiogram showed significant ST-segment depression of the anterior chest leads. Two-dimensional echocardiography revealed a left ventricular ejection fraction of 0.60 and no regional wall motion abnormalities. Selected coronary angiography showed severe narrowing of the left main coronary ostium.

Reprint requests to: J. Zhang

Received: November 30, 2009 / Accepted: February 26, 2010

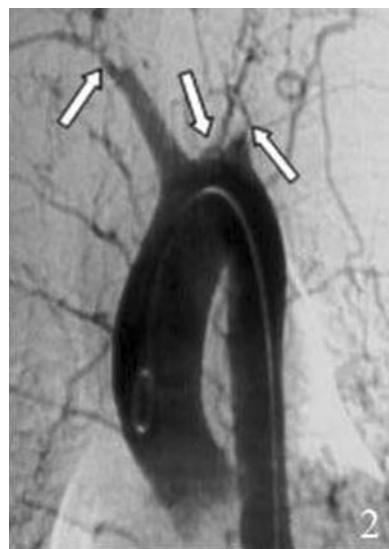


**Fig. 1.** Stenosis of the left main coronary artery ostium was evident (arrow with black outline). White arrows indicate the left anterior descending (*LAD*) and left circumflex (*LCX*) coronary artery, respectively

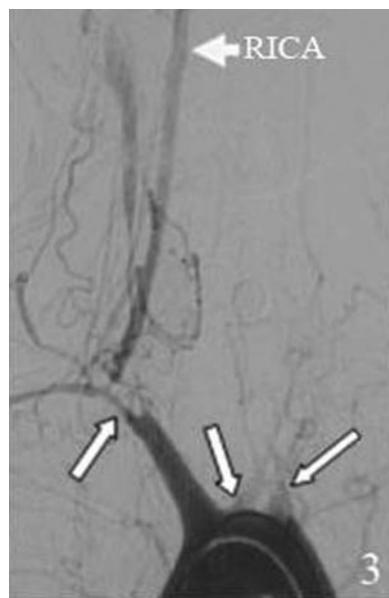
Aortic arch angiography showed total occlusion of the left carotid artery ostium and subclavian artery ostium. It also showed narrowing of the right carotid artery, right subclavian artery, and right vertebral artery ostium. The left vertebral artery and left carotid artery could not be observed in the delayed arterious phase image. Aortography also showed proximal narrowing of the left renal artery (Figs. 1–4). The diagnosis of TA was finally made in this case based on the modified American College of Rheumatology diagnostic criteria for TA.<sup>3</sup>

Methylprednisolone was given intravenously at 2 mg/kg per day to suppress the inflammatory arteritis the day after admission. Her C-reactive protein level and her erythrocyte sedimentation rate were normal after 7 days. Because of the patient's combination of coronary stenosis and multivessel brachiocephalic involvement, we decided that simultaneous surgical treatment using coronary artery bypass grafting (CABG) under cardiopulmonary bypass (CPB) and ascending aorta to left carotid artery bypass using the great saphenous vein was the best option.

The patient experienced an uneventful intraoperative and postoperative course. The operation was performed through a median sternotomy. Cardiopulmonary bypass was therefore instituted between the right common iliac artery and the right atrium. After CPB, a temporary vascular shunt was used to transfer blood to the left carotid artery. It was important to transfer enough blood



**Fig. 2.** Aortic arch angiography showed significant stenosis in the distal brachiocephalic trunk, right carotid artery, right subclavian artery, right vertebral artery origin, and total occlusions of the left carotid artery and subclavian artery origin (arrows with black outline)



**Fig. 3.** A delayed arterious phase angiogram showed the right internal carotid artery (*RICA*; white arrow) developing gradually, but the left carotid artery and left vertebral artery could not be observed in the image

to maintain cerebral blood flow perfusion. The patient underwent CABG with two vein grafts: one to the left anterior descending coronary artery (*LAD*) and the other to the left circumflex coronary artery (*LCx*) due to narrowing of the left main coronary ostium. During the operation, we found that the left coronary ostium



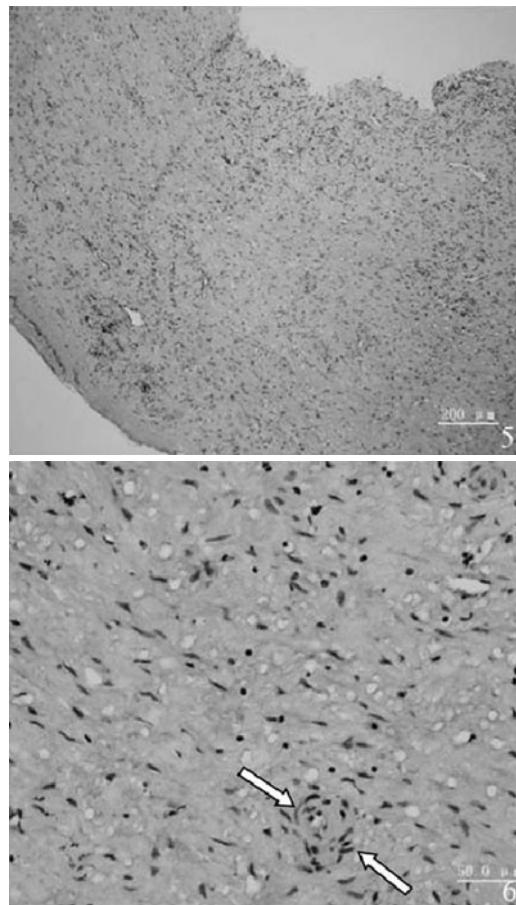
**Fig. 4.** Aortography showed proximal stenosis of the left renal artery (arrow with black outline)

was totally occluded, and the ascending aorta was thick and fibrotic. Nevertheless, a relatively preserved area was chosen for adequate construction of both proximal anastomoses. We also explored the left carotid artery and found that it was patent in the distal portion. After CABG, we performed an ascending aorta to left internal carotid artery bypass using the great saphenous vein. Transcranial Doppler ultrasonography was used to monitor perioperative cerebral blood flow perfusion, as there was a potentially high risk of brain damage. A photomicrograph of the ascending aorta of this patient shows marked intimal and adventitial thickening. Scattered lymphatic inflammation with neovascularization is seen in the media (Figs. 5 and 6). Steroid therapy was started on the second postoperative day on the basis of a clinical diagnosis of vascular disease.

At her 2-year follow-up, the patient remained completely asymptomatic, with good exertional tolerance. Ultrasonic sound examination at a local hospital showed that the left internal carotid artery and great saphenous vein graft maintained their patency and that heart function was normal.

## Discussion

Takayasu's arteritis is a chronic vasculitis involving the aorta and its main branches, the pulmonary arteries, and the coronary tree. It may lead to stenosis with obstruction caused by fibrosis and thrombus formation, or



**Figs. 5 and 6.** A piece of the ascending aorta wall was observed by routine hematoxylin–eosin staining. Lymphocyte infiltration was remarkable in that the total ascending aorta wall and neovascularization is seen in the media (arrows with black outline)

aneurysm formation due to weakening of the vessel wall. The disease was first described in 1908 in a Japanese patient with retinal artery abnormalities, but was later characterized as a more generalized arterial disease.<sup>4</sup> Takayasu's arteritis has a female preponderance, with a reported female-to-male ratio of 8–10:1.<sup>5</sup> The disease is most common in the southeast Asian population, but its cause is unknown. The brachiocephalic vessels are the most frequent site involved in TA. Takayasu's arteritis with coronary artery involvement is rare, and there is little published information on the subject.

Coronary artery involvement in TA was first described by Frovig and Loken<sup>6</sup> in 1951, and CABG was first performed by Young et al.<sup>7</sup> in 1973. The incidence of coronary lesions complicating TA has been reported as being 10.5% in autopsy cases<sup>8</sup> and 9% in patients receiving a clinical diagnosis.<sup>9</sup>

Diagnosis of TA is based on the presence of symptoms and signs of ischemic, inflammatory large vessel disease, and supportive radiographic findings. According to the American College of Rheumatology diagnostic criteria for TA, we diagnosed this patient as having TA. The patient was a complicated patient because of the involvement of brachiocephalic multivessel lesions associated with left coronary artery stenosis. The incidence of cerebral complications is especially high among patients with associated bilateral carotid lesions. Based on the preoperative findings, we decided that simultaneous surgical treatment of CABG under CPB and ascending aorta to left carotid artery bypass using the saphenous vein was the best option. A temporary vascular shunt was used to transfer blood to the left carotid artery to maintain cerebral blood perfusion. Transcranial Doppler ultrasonography was used to monitor the perioperative cerebral blood flow. All these methods were used to decrease the risk of brain damage.

Patient management depends on the presenting phase of the disease. Pharmaceutical therapy has been successful in improving the symptoms in a majority of patients.<sup>10</sup> This includes high-dose corticosteroids and immunosuppressive therapy with cyclophosphamide, and low-dose methotrexate in patients in the inflammatory phase. Surgery is indicated for the relief of ischemic symptoms during the sclerotic phase. The surgical procedure always consists of a bypass operation to vessels normal on angiography, proximal and distal to the occlusive or stenotic lesion. Surgery should be avoided in the acute phase, because it is associated with a high rate of complications.

Patients positive for C-reactive protein (value >1.0 mg/dl) or whose erythrocyte sedimentation rate rose to >20 mm/h are considered to have active-stage inflammation, and should be placed on steroid therapy until these values return to normal, as previously reported.<sup>11</sup> Post-operative steroid therapy is strongly recommended for patients who are treated surgically with clinical and serological signs of an active phase of the disease.

A major issue in patients with TA undergoing CABG is restenosis of the grafts. The choice of conduit — either a great saphenous vein graft or an internal mammary artery (IMA) — is controversial. The long-term graft patency of the IMA versus the great saphenous vein in atherosclerotic patients favors the use of the IMA, provided that its origin and that of the proximal subclavian artery are normal. In our patient, the main drawback of the IMA was the compromise of the grafts if the disease were to recur, and it involves the origin of the subclavian artery or the brachiocephalic trunk. Conventional CABG with a saphenous vein graft can be used in cases of arch-vessel involvement if the IMA fails. Although internal thoracic artery graft patency is significantly

higher than that of vein grafts, it should not be used in patients with TA because of the high rate of subclavian artery occlusion.

The major pathognomonic histological feature of TA is the presence of giant cells at the interface between normal elastic tissue and disorganized damaged elastic tissue. The pathological process may result in narrowing of the vessel wall due to dense fibrosis with or without superimposed thrombosis, or progressive aneurysm formation due to weakening of the vessel wall. Marked thickening of the vessel wall is observed at surgery, and is also an early finding on ultrasonography.

Progression of the disease in the coronary arteries, aorta, and aortic branches is a possibility, and stenosis or occlusion of the grafts may occur. These patients should therefore be kept under close postoperative surveillance.

The present patient was event-free at 2 years' follow-up. The combination treatments (either preoperative, intraoperative, or postoperative) were successful. In conclusion, surgical treatment of symptomatic TA was highly effective and safe. Symptomatic improvement and excellent long-term graft patency should be expected after arterial reconstruction.

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