Regression of Carotid Stenoses After Corticosteroid Therapy in Occlusive Thromboaortopathy (Takayasu's Disease)

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A 34-year-old, single Japanese woman with active Takayasu's disease presented with stenosed bilateral common carotid arteries, subtotal occlusion of the left subclavian artery, and complete occlusion of the ipsilateral proximal vertebral artery, as demonstrated by angiography. A right subclavian-left vertebral bypass using an autologous saphenous vein graft was successfully performed after 36 days of corticosteroid therapy. Subsequently, warfarin was administered in addition to prednisolone. After 32 months of treatment with a gradual reduction from 50 to 3.75 mg of prednisolone daily, angiography revealed a functioning bypass graft and regression of bilateral common carotid stenoses.

Report of a Case

This patient had an occasional mild stiffness in the left nuchal region and easy fatigability of the left upper extremity on exercise of the ipsilateral arm, lasting 5 years from April 1966, when she was 18 years old, to 1971. Subsequently, she had an asymptomatic period of about 10 years. In October 1981, at age 34, she began to notice bilateral tinnitus, general fatigue, and spontaneous pain in both the left nuchal and left anterior or neck region. The latter two symptoms occurred repeatedly, and she was admitted to the hospital. In January 1982, she had transient vertigo and often had to rest in bed. On April 3, she experienced a sudden onset of severe hoarseness and dysphagia, followed by vertigo and a throbbing headache. These symptoms did not completely subside even after 7 weeks. On May 23, she felt a numbness of the left upper lip, left buccal mucosa, and left side of the face.

She was admitted to Kyoto University Hospital on May 25, 1982. Examination revealed dysphagia, dysarthria, deviation of the tongue and uvula to the left, and hypesthesia of the face, upper lip, and buccal mucosa on the left. The pulses of the left upper extremity were absent, and the blood pressure was unmeasurable. There were palpable pulses on the bilateral common carotid arteries and systolic bruits in the bilateral supravacular fossae and the neck. Blood pressures in the right arm and right and left thighs were 128/70, 132/70, and 128/60 mm Hg, respectively. Right and left retinal arterial pressures in the supine position were 74/28 and 58/35 mm Hg, respectively. No other abnormal findings were detected at the bedside examination. She had slight fever and a high ESR (84 mm/hr, Westergren).

Results

Cerebral computed tomography scan including the posterior fossa, electrocardiogram, and chest x-ray were normal except for a decrease in the pulmonary vasculature in the right upper area. Subsegmental perfusion defects in the right upper lobe were revealed by pulmonary perfusion scintigraphy in a normal xenon-133 ventilation study. In retrograde arch aortograms, the left subclavian and ipsilateral vertebral arteries did not appear, and narrowing of the the bilateral common carotid arteries was evident (Figure 1 left). Retrograde right vertebralangiography and selective left subclavian arteriography revealed subtotal occlusion of the left proximal subclavian artery and complete occlusion of
the ipsilateral proximal vertebral artery (Figure 1 right).

On June 30, prednisolone, 50 mg daily, was prescribed because of the established diagnosis of active Takayasu's disease. The daily dose of prednisolone was gradually reduced to 20 mg on August 5, when the ESR was normal and complaints had subsided considerably. A right subclavian-left vertebral bypass using an autologous saphenous vein graft was successful.

The patient was discharged in August 1982 and put on warfarin in addition to prednisolone, 15 mg daily. She was examined every 2 weeks for 28.5 months and became asymptomatic about 1 year after discharge.

In February 1985, she was readmitted for angiography to evaluate effectiveness of the treatment. At that time, the inflammation was completely suppressed by alternate daily doses of prednisolone, 5 and 2.5 mg, respectively. Right and left brachial blood
pressures were 123/66 and 96/92 mm Hg, respectively. Right and left retinal arterial pressures were 98/42 and 101/46 mm Hg, respectively. Right heart catheterization was normal, and pulmonary arteriography revealed slight pulmonary arterial lesions, which mainly comprised subsegmental occlusion in the right upper lobe. In March, retrograde total aortography was carried out. Aortic arch aortograms revealed considerable regression of the bilateral carotid stenoses and a patent bypass graft (Figure 2). Abdominal aortograms were normal.

The patient was discharged in March 1985, but continued to be followed. In April 1986, steroid therapy and warfarin were discontinued. Right and left brachial blood pressures were 112/64 and 96/84 mm Hg, respectively. Pulses of the left upper extremity were faintly but clearly palpable. As of July 1986, she was asymptomatic with no restriction in normal physical activity. ESR was 7 mm/hr.

Discussion

In 76 autopsied patients with Takayasu’s disease, histology of the affected arterial wall showed that different stages of the inflammatory process were often present in different sites of the involved arterial segments in a single individual. However, which segment of the many involved segments will be steroid-sensitive or -resistant is clinically unknown. Corticosteroid therapy is usually first prescribed for patients in the clinically active stage of the disease. In the patient reported here, an obvious luminal improvement of stenosis of the aortic arch vessels, except for the left subclavian artery, was shown by arch aortography after 32 months on steroid treatment. There was no apparent improvement of the subtotal occlusion of the left subclavian artery, although in the left upper extremity unpalpable pulses and unmeasurable blood pressure did become faintly palpable and carefully measurable, respectively. There was a development of the left intercostal arteries. The involved left subclavian artery was probably already in a more steroid-resistant, advanced stage of the inflammatory process, or thrombi were probably superimposed on the luminal involvement before the steroid therapy, or both. Shelhamer et al reported that 8 of 16 patients with active Takayasu’s disease responded to corticosteroid therapy for 4 weeks, with improvement in their symptoms and stabilization of their angiographic abnormalities. Five of the remaining 8, who responded poorly to the therapy, had angiographic progression of the vascular lesions while on prednisone therapy. A period of > 4 weeks on steroids may be necessary for angiographic improvement of the stenosed arteries in patients with active Takayasu’s disease.

If the inflammation of the arterial wall in Takayasu’s disease is active, the possibility of suture failure or aneurysm formation as well as of occlusion of the graft is higher than in the case of arterial disease of other causes. In the present patient, a very high ESR was completely suppressed by steroid therapy before the reconstructive bypass surgery. No definite conclusion has been reached concerning medical vs. surgical treatment for the severely affected 4 cerebral arterial systems in patients with Takayasu’s disease.

In those patients with active Takayasu’s disease, the response to corticosteroid therapy was good. There was a suppression of the high ESR, a reduction in subjective symptoms, and a retardation of the progression of arterial disease. The results in our patient provide further evidence for the effectiveness of long-term corticosteroid therapy on the narrowed aortic arch vessels during the inflammatory active stage.

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References


Key Words: Takayasu’s disease • extracranial arteritis • corticosteroid therapy • vertebral arterial operation • aortic arch syndrome
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