Bilateral Subclavian Steal Associated With Severe Coarctation of the Thoracic Aorta and an Aberrant Right Subclavian Artery

A 10-year-old girl having bilateral subclavian steal associated with severe coarctation of the thoracic aorta and an aberrant right subclavian artery was found, on admission, to have no difference between upper and lower extremity blood pressure, but echocardiography revealed severe thoracic aorta coarctation and systolic blood pressure in the carotid arteries exceeding 200 mmHg estimated by Doppler ultrasonography. Magnetic resonance imaging and angiography demonstrated bilateral subclavian steal without esophageal compression. We reconstructed the aortic arch using the left subclavian artery and a reversed Blalock-Park procedure, then repaired the coarctation with a 14 mm woven double velor vascular graft. The girl was symptom-free following uncomplicated recovery from surgery. Doppler ultrasonography 2 weeks after surgery showed the pressure gradient across the aortic arch had decreased from 180 mmHg to 60 mmHg. This residual gradient at the anastomosis between the ascending aorta and left subclavian artery may improve as native vessels grow. (Jpn J Thorac Cardiovasc Surg 2003; 51: 110-112)

Key words: bilateral subclavian steal, aberrant right subclavian artery, coarctation of thoracic aorta

Subclavian steal is usually seen in adults with atherosclerotic lesions of the subclavian arteries. Subclavian steal, and bilateral steal in particular, is rare in childhood. Pediatric bilateral subclavian steal may occur with coarctation of the thoracic aorta and the origin of both subclavian arteries distal to coarctation. We report a case of bilateral subclavian steal associated with severe coarctation of the thoracic aorta and an aberrant right subclavian artery.

Case

A 10-year-old girl referred to the National Nagasaki Medical Center for evaluation of mild dyspnea and strong neck vessel pulsation on exertion was found, on examination, to have upper extremity systolic blood pressure of 100 mmHg but no difference in blood pressure between upper and lower extremities. She had regular rhythm with a normal first but accentuated second heart sound. A harsh, grade 3/6 systolic, ejection murmur was present, best heard at the left sternal border in the second intercostal space. Electrocardiography showed normal sinus rhythm with left ventricular hypertrophy. Chest radiography was unremarkable. Echocardiography revealed severe coarctation of the thoracic aorta with systolic blood pressure in the carotid arteries exceeding 200 mmHg estimated by Doppler evaluation. Magnetic resonance imaging (MRI) to delineate aortic arch anatomy showed a hypoplastic aortic arch with severe coarctation of the proximal descending aorta and the left subclavian artery and aberrant right subclavian artery originating from the stenotic aorta segment.

Angiography via a right femoral approach demonstrated bilateral subclavian steal associated with a hypoplastic aortic arch and severe coarctation, and a large caliber ascending aorta and proximal arch aorta with
normally originating carotid arteries. The gradient measured between the ascending and descending aorta was 180 mmHg (Fig. 1). Through a left thoracotomy, we reconstructed the aortic arch using the left subclavian artery and a reverse Blalock-Park procedure, then repaired the coarctation using a 14 mm woven double velor vascular graft (Fig. 2). Cardiopulmonary bypass was used to reduce the after-load against the left ventricle and cerebral tissue during aortic cross-clamping. The girl was symptom-free following an uncomplicated recovery from surgery. Doppler ultrasonography 2 weeks postoperatively showed the pressure gradient across the aortic arch had decreased from 180 mmHg to 60 mmHg. This residual gradient at the anastomosis between the ascending aorta and left subclavian artery may improve as native vessels grow (Fig. 3).