Case Report

Atypical Aortic Coarctation with Resistant Hypertension Treated with Axilloiliac Artery Bypass

Hiroko INAGAKI, Johji KATO, Atsushi YAMASHITA, Tomohiko UEMURA,
Kazuo KITAMURA, Yoshikazu YANO*, Kunihide NAKAMURA*,
Toshio ONITSUKA*, and Tanenao ETO

A 68-year-old woman was found to have atypical coarctation of the aorta, accompanied by systolic hypertension of the upper extremities despite administration of five types of antihypertensive drugs. Since the systolic hypertension was resistant to the conventional antihypertensive therapy, axilloiliac artery bypass grafting with a subcutaneous tunnel was performed to alleviate the pressure gradient. Systolic blood pressure was successfully reduced and hypertension was controlled after surgery.

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Key Words: atypical aortic coarctation, resistant hypertension, axilloiliac bypass grafting

Introduction

Hypertension secondary to coarctation of the aorta is often curable by surgical or non-surgical methods, but if untreated, it carries a poor prognosis due to the likelihood of congestive heart failure or cerebral hemorrhage (1-3). In Japan, atypical coarctation of the aorta, presumed to be mostly secondary to aortitis syndrome, is relatively common compared to that of congenital origin (4). The primary goal of the treatment for this disorder is to relieve stenosis, while the surgical or non-surgical procedure chosen may differ depending on factors such as patient age or the site of blood vessels involved (3, 5). We report here a case of atypical coarctation of the aorta with resistant hypertension, treated with axilloiliac bypass grafting.

Case Report

A 68-year-old woman was referred to our hospital as a result of hypertension that was resistant to antihypertensive

therapy with 40 mg furosemide, 10 mg enalapril, 10 mg amlodipine, 8 mg doxazosin, and 20 mg arotinolol. Hypertension had been detected thirty years prior to referral. Blood pressure had been controlled until, according to the patient's medical records, it began to rise and hypertension became more resistant two months prior to admission. Diet therapy had been recommended for diabetes mellitus at age 58, and cerebral infarction had occurred four years prior to admission.

On physical examination, brachial blood pressure was 212/60 mmHg without laterality, despite the administration of the antihypertensive medication listed above, while systolic blood pressure in the lower extremities was 110 mmHg by pulsation. Fundoscopic examination showed segmental narrowing of retinal arteries, without exudate, hemorrhage or papilledema. The chest was clear on auscultation, but bruit was audible at the upper umbilical region. Neurological examination revealed exaggerated deep tendon reflexes in the upper and lower extremities on the right side.

Table 1 summarizes laboratory data on admission. No remarkable abnormalities were observed in the urinalysis

From the First Department of Internal Medicine, Miyazaki Medical College, Miyazaki, Japan, and *Second Department of Surgery, Miyazaki Medical College, Miyazaki, Japan.

Address for Reprints: Johji Kato, M.D., First Department of Internal Medicine, Miyazaki Medical College, Kiyotake, Miyazaki 889-1692, Japan.

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Table 1. Laboratory Data on Admission

1. Urinalysis		
Protein		Negative
Glucose		Negative
Hemoglobin		Negative
2. Peripheral blood		
Red blood cell count		$397 \times 10^4/\mu\text{l}$
Hemoglobin		12.3 g/dl
Hematocrit		37.3%
White blood cell count		$6,300/\mu\text{l}$
Platelet		$11.7 \times 10^4/\mu\text{l}$
Erythrocyte sedimentation rate		11 mm/h
3. Blood chemistry		
Total protein		6.7 g/dl
Total cholesterol		179 mg/dl
Fasting plasma glucose		144 mg/dl
Urea nitrogen		18.7 mg/dl
Creatinine		1.1 mg/dl
Creatinine clearance		34 ml/min
Na		144 mEq/l
K		3.9 mEq/l
C-reactive protein		0.1 mg/dl
Hemoglobin A _{1c}		6.3%
4. Endocrinological test		
Plasma renin activity		1.4 ng/h/ml
Aldosterone		102 pg/ml
Adrenaline		<10 pg/ml
Noradrenaline		395 pg/ml

and hematological studies, although serum creatinine level and creatinine clearance suggested impairment of renal function. Plasma renin activity (PRA) was in the normal range under antihypertensive medications of 10 mg amlodipine and 4 mg doxazosin, rising to 3.2 ng/h/ml 1 h after 25 mg captopril administration. No elevation was observed in the plasma aldosterone, adrenaline, or noradrenaline levels. Total blood volume was 63.2 ml/kg, a level within the normal limit. Chest X-ray showed an increased cardiothoracic ratio of 59%, and ST depressions and negative T waves were noted in the II, III, aVf, V5, and V6 leads in electrocardiogram. Echocardiogram revealed left ventricular hypertrophy with interventricular septum thickness at a diastole of 12.3 mm. As shown in Fig. 1, computed tomography (CT) revealed a markedly calcified descending aorta with a severe stenosis in the suprarenal portion (A) and the contracted right kidney (B). The stenotic lesion was further examined by angiography and by measurement of intraaortic pressure (Fig. 2). Two stenoses were found: one located at the suprarenal abdominal aorta with a pressure gradient of 70 mmHg at systole, the other at the infrarenal aorta with a gradient of 20 mmHg. In addition, a 50% stenosis was

Fig. 1. Computed tomography showed severe stenosis at the upper portion of the abdominal aorta with marked calcification (A) and contracted right kidney (B). The scale is in centimeters.

noted in the left renal artery, while a complete occlusion was demonstrated in the right.

Mainly because of the markedly calcified descending aorta, for which neither transluminal angioplasty nor aorto-aortic bypass grafting appeared an appropriate choice, axillo-external iliac artery bypass grafting was performed to relieve the pressure gradient. A Y-shaped knitted polyester prosthesis was subcutaneously grafted by end-to-side anastomoses to the right axillary artery and bilateral external iliac arteries. The operation was carried out safely and the patient recovered uneventfully. Blood pressure in the upper extremity declined to a level of 160/70 mmHg soon after the bypass grafting under antihypertensive therapy with 10 mg amlodipine, 4 mg doxazosin, and 1 mg indapamide. Hypertension has been well controlled with these drugs since then, without a significant difference in blood pressure between the upper and lower extremities. Postoperative angiography revealed that the left kidney was perfused not only by blood descending through the aorta but also by retrograde flow from the bypass graft, a finding that could be expected,

Fig. 2. Aortography and measurement of intraaortic pressure (mmHg). Two stenoses were seen in the abdominal aorta, as indicated by arrows. Suprarenal stenosis with pressure gradient of 70 mmHg at the systole was more severe than that of the infrarenal portion.

given the much less severe stenosis in the infrarenal aorta. Neither deteriorated renal function nor elevated PRA was observed postoperatively, and serum creatinine has remained unchanged during the year following surgery.

Discussion

Several mechanisms for hypertension in coarctation of the aorta have been proposed: these include an activated renin-angiotensin system, an imbalance of sodium and body fluid homeostasis, or an active role of the kidney(s) distal to the stenosis (6). In this case, PRA was within the normal range and rose slightly following captopril administration before surgery despite 50% stenosis of the left renal artery, suggesting a less active role for the renin-angiotensin system. In addition, total blood volume was in the normal range preoperatively and renal function remained unchanged after surgery. Alternative to the mechanisms described above, a lack of "Windkessel" properties may be considered the mechanism in this case (7). Indeed, the markedly elevated systolic blood pressure soon declined postoperatively, while diastolic blood pressure remained unchanged. An immediate reduction of

systolic blood pressure suggests a compliance increased by the bypass graft connected to the large arteries distal to the stenosis, which absorb the energy of pulsatile flow and pressure of the left ventricular ejection.

Inada *et al.* reported 13 cases of pulseless disease and/or atypical coarctation of the aorta, and based on the sites of blood vessels involved in the cases in their series, they concluded that atypical coarctation is most likely secondary to aortitis syndrome (4). Aortitis syndrome also seemed likely in our case; the clinical features were similar to those of their cases, though an etiology was unable to be identified given the remaining possibilities of involvement of atherosclerosis or congenital origin. Notably, axilloiliac artery bypass grafting with a subcutaneous tunnel was performed in this patient without any serious complication, successfully lowering systolic blood pressure by about 50 mmHg without deterioration of renal function or elevation of PRA. Based on the findings of the postoperative angiography, there may have been no apparent reduction in the renal blood flow after surgery. When compared with other extra-anatomical bypass graftings, axilloiliac artery bypass is less invasive with a lower surgical mortality rate, though the long-term patency of this procedure has been reported to be somewhat lower (8). Despite the fact that indications have not yet been well established, this bypass grafting procedure appears to be a suitable option for older high-risk patients with atypical coarctation of the aorta accompanied by resistant hypertension.

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