Case Report



Surgical Reconstruction for Coarctation of the Abdominal Aorta

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Abstract

A 28-year-old man was referred for hypertension of 160/92 mmHg that was poorly controlled medically. However, he had no signs of lower limb ischemia; he had diminished femoral pulses and anklebrachial indices of 0.7 bilaterally on physical examination. Computed tomography revealed coarctation of the abdominal aorta, beginning just below the takeoff of both renal arteries and extending above the inferior mesenteric artery. A 14-mm woven dacron aorto-aortic bypass, sutured end-toside to the proximal descending thoracic aorta above and to the infrarenal abdominal aorta below, as well as right renal artery reconstruction were performed. The postoperative course was uneventful and he continues to have normal blood pressure and normal femoral pulses.

Key words: Middle aortic syndrome, abdominal aortic coarctation, hypertension

Introduction

Coarctation of the aorta is defined as significant luminal narrowing of the aorta that produces hemodynamically significant obstruction to the flow of blood [1]. Although 98% of aortic coarctation is located in the proximal descending thoracic aorta near the ligamentum arteriosum, 2% of coarctation affects the abdominal aorta, which is often called "middle aortic syndrome" or "mid-aortic dysplastic syndrome" [2,3]. Coarctation of the abdominal aorta represents a distinct clinical entity resulting from diffuse narrowing of the aortic lumen and occasional involvement of adjacent arterial trunks [4]. The etiology of this condition is poorly understood. The most common clinical manifestation of aortic hypoplasia is severe uncontrolled hypertension. Hypertension is often difficult to manage and requires treatment with anti-hypertensive medication and/or Department of Cardiovascular Surgery Saitama Medical Center Jichi Medical University Saitama-City, Japan

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Corresponding author: Manabu Shiraishi, MD Division of Cardiovascular Surgery Saitama Medical Center Jichi Medical University 1-847 Amanuma, Omiya-Ku Saitama-City 330-8503, Japan manabu@omiya.jichi.ac.jp surgical repair. We report a case with abdominal coarctation and hypertension that were operatively treated.

Case Report

A 28-year-old man was admitted to our hospital with complaints of hypertension. His medical history was significant for untreated hypertension that was first noted during childhood. During a primary care doctor follow-up, the hypertension was difficult to control with anti-hypertensive drugs. He was a nonsmoker and did not consume any alcohol. There was not another significant past medical history. Physical examination on admission revealed a normally developed man. His heart rate was 72 beats/min and regular with weak femoral, posterior tibial, and dorsalis pedis pulses. Brachial blood pressures were elevated, at 160/92 mm Hg, and lower thigh blood pressures were 124/82 mm Hg. The lung fields were clear; the heart was normal. No cervical or abdominal bruits were noted. There was no peripheral edema or cyanosis. Neurologic examination was found to be normal. Results of a complete blood count were normal, and the C-reactive protein level was normal. Serum blood urea nitrogen (BUN), creatinine levels and aldosterone were normal (8 mg/ dL, 0.69 mg/dL and 114 pg/mL). Peripheral venous rennin, angiotensin I and angiotensin II sampling were found to be elevated (11 ng/ml/hr, 420 pg/mL and 29 pg/mL). A 12-lead electrocardiogram revealed a normal sinus rhythm with left ventricular hypertrophy. Echocardiography showed left ventricular hypertrophy with good systolic function. Computed tomography (CT) showed a marked narrowing of the peri-renal abdominal aorta, distal to the inferior mesenteric artery (IMA) with no surrounding inflammatory changes. 3-dimensional contrast-enhanced CT of the abdominal aorta demonstrated that there was a long segmental narrowing of the abdominal aorta that was mostly marked in the upper portion (where it measures about 6 mm in diameter). A thin right renal artery arising from the narrow aorta was noted. The superior mesenteric artery (SMA) and the IMA were also meandering and enlarged, forming collaterals. There was some mural calcification, but no thrombosis or dissection were detected in the abdominal aorta and its branches (Figure 1). Angiographic examination demonstrated that the thoracic aorta and the origins of the great ves-



Figure 1. Computed tomography angiography showing coarctation of the abdominal aorta, beginning just below the takeoff of both renal arteries and extending above the inferior mesenteric artery. The white arrow is showing highly developed collateral between the superior and inferior mesenteric artery.



Figure 2. Angiography is showing abdominal coarctation at the renal artery level with severe stenosis of right renal artery ostia.

sels were normal; however, there was a segmental aortic coarctation beginning just below the level of the left renal artery, and including the right renal artery orifices (Figure 2).

The operation was performed with the patient in a supine position. An abdominal median incision was made for access to the abdominal aorta. The distal descending aorta and proximal abdominal aorta were found to have a normal diameter. No periaortic inflam-

266

matory changes were noted. Then, the distal abdominal aorta was exposed. The proximal portion of the coarcted segment was an obvious transmural atherosclerosis is with calcification. First, a partial occluding clamp was applied to the distal descending thoracic aorta, consistent with circulation of the lower part of the body with du femoro-femoral bypass. A 14 mm in diameter woven dacron vascular graft was anastomosed to the aorta in an end-to-side manner. The graft was tunneled through an opening in the diaphragm and brought up to the abdominal cavity retroperitoneally. A partially occluding vascular clamp was applied to the distal abdominal aorta just below the level of IMA, and end-to-side anasto-

mosis was performed. The right renal artery was divided distal to the stenotic area and was attached directly to the aortic graft. Pressures in the arteries were measured following the completion of the thoracoabdominal bypass. The pressure gradient between the upper extremities and the lower extremities was eliminated.

The postoperative course was uneventful, and he was discharged on the fifteenth day with his blood pressure regulated by an anti-hypertensive drug to 126/70 mm Hg. The lower thigh blood pressures were elevated at 132/73 mm Hg. There was no measurable rise of BUN or serum creatinine postoperatively. The control CT revealed a patent thoracoabdominal and right aortorenal bypass (Figure 3).



Figure 3. Postoperative computed tomography angiography is showing a patent prosthetic aorto-aortic bypass graft (gray arrow) and right renal artery. The collateral arteries underwent spontaneous regression (white arrow).

Discussion

The stenosis in coarctation of the aorta is characteristically located at the junction of the distal aortic arch and descending aorta. Atypical coarctation, located far from the aortic isthmus, affects about 0.5% to 2% of individuals with coarctation of the aorta [5].

Hypertension is the cardinal clinical feature of coarctation of the abdominal aorta and was present in 94% of the cases reviewed [6]. Symptomatic patients may present with a mental status change, headache, nosebleed, sleep disturbance and fatigue [6]. The hypertension is typically severe and, if left untreated, can result in life-threatening complications, such as a stroke and heart and renal failure [1]. Onat et al. reported that 42% of patients presented with hypertensive encephalopathy and 45% died before 34 years of age [7]. The hypertension is most likely the result of global renal hypoperfusion and activation of the rennin-angiotensin system [1].

Claudication is less common and is seen in cases of infrarenal hypoplasia. It may be associated with the early development of atherosclerosis in the aortoiliac distribution [1,8]. The visceral arteries are also involved in more than 20% of cases, but mesenteric ischemia is rare because of collateral formation [8].

Physical examination of patients with abdominal aortic hypoplasia has usually revealed normal upper extremity pulses, diminished pulses in the lower extremities, and occasionally an abdominal bruit. Blood pressure in the upper extremities is usually significantly greater than in the lower extremities [1].

Although it remains unclear, several hypotheses exist for the etiology of the abdominal aortic coarctation: congenital abdominal coarctation (2); virus infection [9]; non-specific aortitis [10]. Congenital abdominal aortic stenosis is thought to be the result of a primary underdeveloped aortic segment, and over-fusion of the two embryonic dorsal aortas or their failure to fuse during embryological development [2]. Certain viruses, notably rubella, may interfere with cell growth and may have direct cytopathic effects on developing tissue. Since this mechanism in the development of aortic hypoplasia has reported an association between hypoplasia and rubella syndrome, it is thought that viral inhibition of smooth muscle cell organization and proliferation may prevent normal aortic growth [1,9]. The main causes of inflammatory arteritis are Takayasu's arteritis, giant cell arteritis, and Behçet's disease. Neurofibromatosis, such as von Recklinghausen's disease and Williams syndrome [11], fibromuscular dysplasia [1,12], tuberculosis [1], and radiation therapy [1,13] at an early age have all been associated with hypoplasia of the abdominal aorta, but no causal relations have been identified [1].

Any young patient with uncontrolled hypertension should be evaluated for aortic coarctation, which may be suspected on the basis of the history and physical examination [1]. Accurate imaging is essential to allow planning of the vascular reconstruction. Angiography is the gold standard for the diagnosis of abdominal aortic coarctation [6]. Aortography not only shows the location and extent of the aortic disease but also may reveal associated abnormalities of the visceral and renal vessels [1]. Other imaging modalities, like CT, may be useful. The use of 3-Dimensional CT may reduce the need for invasive angiography [6]. CT may give additional information regarding the presence of any associated inflammatory changes: the presence of aortic wall thickening that angiography cannot evaluate [1]. A genetics evaluation may be needed in cases with stigmata of neurocutaneous conditions or Williams syndrome. Additional evaluation for abdominal aortic coarctation includes tests for baseline renal function and peripheral renin activity, renal ultrasound, echocardiography and a retinal exam [6].

The prognosis of aortic hypoplasia is generally poor in untreated cases. Untreated patients experience premature death secondary to the morbid sequelae of uncontrolled hypertension. Surgical correction of abdominal aortic coarctation remains the definitive treatment when technically feasible. However, with effective antihypertensive treatment, conservative therapy is often possible prior to surgery. Hypertension is typically severe and often difficult to manage, and it requires treatment with a combination of multiple classes of medications (angiotensin-converting enzyme inhibitors and angiotensin II receptor blockers) [6].

The primary indication for operative intervention has been uncontrolled severe hypertension. The exact timing of surgery should be individualized and based Four types of aortic coarctation have been described: type I, suprarenal coarctation and renal artery stenosis; type II, infrarenal coarctation and renal artery stenosis; type III, suprarenal coarctation and normal renal arteries; and type IV, infrarenal coarctation and normal renal arteries [8]. The operation must be individualized on the basis of the length of the hypoplastic segment and associated renal artery involvement. For types I to III, surgical intervention is aimed at improving renal blood flow and preserving renal function. Surgical intervention for type IV coarctation is indicated in cases of severe claudication and consists of aortic reconstruction [14]. Depending on the length and position of the pathological lesion, an appropriate operative procedure (aorto-aortic bypass, patch aortoplasty, extra-anatomic axillo-femoral bypass, percutaneous transluminal balloon angioplasty and stent placement) should be selected [6,15]. For renal artery stenosis, autotransplantation or aorto-renal bypass with prosthetic grafts and autogenous venous grafts are preferred [4,6]. The superior mesenteric and celiac arteries may be involved with the coarctation, but revascularization does not appear to be necessary because they are usually adequately filled from the inferior mesenteric artery by way of the meandering mesenteric artery [2,3]. However, there are some reports about the early and late complications after surgical reconstruction for coarctation of the abdominal aorta. Vaccaro et al. reported the occlusion of the renal artery bypass graft twice (the fifth postoperative day and two years later) after surgery [3]. Badmanaban et al. reported the dissection distal to the coarctation repair [16].

on the severity of symptoms, the response to medical

treatment and the consideration of surgical risks [6].

In the present case, the only presenting symptom was uncontrolled hypertension without any signs of occlusive disease. The patient had about a 15-cm segment of abdominal aortic coarctation extending from the renal artery to near IMA. Adequate distal perfusion via highly developed collaterals between the SMA and IMA preserved lower limb ischemia. We used moderate total body hypothermia as an adjunct to bypass grafting to minimize spinal complications and damage to abdominal organs, and performed combined correction of abdominal aortic coarctation and the right renal artery by thoracoabdominal bypass with a woven dacron graft and direct aortorenal bypass using the inherent renal artery.

Without early treatment, death from a cerebral hemorrhage, stroke and heart failure before the age of 40 years is very likely to occur [4]. Although the natural history of the disease is practically unknown, long-term follow-up (in excess of 20 years) demonstrates reconstructive success, symptom relief, and a normal quality of life [17].

Conclusion

In this report, a 28-year-old man with abdominal aortic coarctation with right renal artery stenosis is presented. Abdominal aortic coarctation is an important cause of renovascular hypertension in young patients and should be considered in the differential diagnosis of hypertension. A surgical bypass graft is the optimal management strategy for this condition to relieve the systemic hypertension and restore circulation to renal arteries.

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Conflict of interest statement

The authors have no conflicts of interest to declare. **References**

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