Atypical Coarctation of the Abdominal Aorta as a Cause of Acute Hypertensive Heart Failure

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SUMMARY

A case of atypical coarctation of the suprarenal aorta with symptoms of acute hypertensive heart failure is reported. A 62-year-old man was admitted because of dyspnea associated with severe hypertension. Chest x-ray demonstrated mild pulmonary congestion. Following antihypertensive management, symptoms of congestive heart failure improved. Abdominal computed tomography and digital subtraction angiography undertaken several days later demonstrated a narrowed segment of the suprarenal aorta. Control of hypertension was poor with currently available antihypertensives and a bilateral axillo-femoral bypass operation with artificial grafts was undertaken. Postoperative blood pressure was maintained at a normal level with a small dose of atenolol. The patient had an uneventful hospital course and remains well.

Additional Indexing Words:
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ORTIC coarctation is considered as a cause of curable hypertension.1-3) Coarctation of the abdominal aorta is a rare condition accounting for 2%4,5) of cases of aortic coarctation. Mortality and morbidity are determined by complications resulting from concurrent hypertension.6-8) If left untreated, death results from fatal cardiovascular complications such as cerebral hemorrhage or cardiac failure.1) Operative correction of aortic coarctation has improved survival by successful control of the refractory hypertension.1,8)

This report describes the case of a 62-year-old male with a markedly
stenosed suprarenal aorta associated with acute hypertensive heart failure.

CASE REPORT

A 62-year-old man was admitted on November 7, 1988, following the sudden onset of severe dyspnea. He had a history of hypertension for more than 10 years and had experienced leg numbness after prolonged walking. He also had claudication in both legs since the previous September, with blood pressure ranging from 170/100 mmHg to 180/110 mmHg in the arm. Blood pressure on admission was 242/112 mmHg in the right and 240/110 mmHg in the left arm. Femoral arterial pulses were not palpable on either side and blood pressure was unobtainable. Moist rales were noted over the chest and a high pitched systolic bruit was audible over the epigastric region. Chest x-ray demonstrated mild pulmonary congestion and cardiomegaly (Fig. 1). Abdominal x-ray showed calcification near the level of the first lumbar vertebra, which was recognizable on upper gastro-intestinal films taken 14 years previously. ECG showed high voltage (3.2 mV) in the left chest leads (V5) with depression of the ST segment, inverted T wave, and sinus tachycardia (110/min). Arterial blood gas analysis revealed low partial pressure of oxygen (45.3 mmHg), and a carbon dioxide partial pressure of 21.9 mEq/l resulting in a metabolic acidosis of pH 7.301. Urinalysis was
normal. Blood urea nitrogen and serum creatinine were slightly elevated (25.9 mg/dl and 1.7 mg/dl, respectively). Plasma renin activity was 11.0 ng/ml/h. On admission, the patient was treated for severe hypertensive heart failure with nasal oxygen, oral nifedipine 20 mg every 6 hours, and parenterally infused nitroglycerin at a dose of 50–100 μg/min. Two hours later, blood pressure dropped to 170/78 mmHg. Echocardiography showed hypertrophy of the left ventricular wall and interventricular septum with mild dilatation of the left ventricular dimension (Fig. 2). Computed tomography (CT) demonstrated severe stenosis of the aorta at the level of the first lumbar vertebra without diminished external diameter (Fig. 3). Digital subtraction angiography (DSA) via the left brachial artery demonstrated an atypical coarctation with severe stenosis of the suprarenal portion of the aorta and complete obstruction of the celiac and superior mesenteric arteries (Fig. 4). Extensive collateralization was seen between the branches of the inferior mesenteric artery and other visceral arteries. Subsequently, the patient was maintained on a combination of a diuretic (furosemide 40 mg), α₁-blocker (urapidil 60 mg), calcium antagonist (nifedipine 80 mg) and angiotensin converting enzyme inhibitor (enalapril 10 mg), but the hypertension remained uncontrolled. Bilateral axillo-femoral bypass operation was performed on March 8, 1989 with supported E-PTFE grafts (10 mm diameter) via the subcutaneous route. A histological study of the left femoral artery obtained

Fig. 2. Echocardiogram showing hypertrophy of the left ventricular wall and interventricular septum with mild dilatation of the left ventricular dimension.
Fig. 3. Computed tomography demonstrating severely stenosed aorta with calcification.

Fig. 4. Digital subtraction angiogram demonstrating severe stenosis of the suprarenal portion of the abdominal aorta with complete obstruction of the celiac and superior mesenteric arteries.

during the operation, showed thickening of the intima and tunica media with focal disruption of the lamina elastica and fibrosis of the tunica media without active inflammation and granuloma. After the operation, his ex-
tremities became warm with good pulsation in both feet, and the intermittent claudication was resolved. Blood pressure improved to 150/80 mmHg in the arms and 130/70 mmHg in the legs with atenolol (25 mg/day). Creatinine clearance improved from a preoperative value of 74.0 ml/min to 100.8 ml/min. Postoperative DSA disclosed good patency of the grafts, and demonstrated retrograde blood flow into the poststenotic aorta from the bilateral femoral arteries via the bypass grafts. The postoperative course was unremarkable and the patient remains active.

DISCUSSION

A case of atypical coarctation of the abdominal aorta with severe hypertensive heart failure and refractory hypertension was described. Since the first reported case by Quain in 1848, coarctation of the abdominal aorta has been a well-known but infrequent cause of hypertension. The disease is seen more frequently in younger people than in older. There has been only one report of a person over 60 years of age with the disease. The etiology is unclear. When a lesion is seen in a young patient and is near the aortic arch, the origin may be congenital. In patients with congenital disease, hypertension is frequently noted in childhood or in adolescence, and the external diameter of the stenotic aorta is diminished. Distal coarctation is mostly of acquired origin. Other reported etiologies include fibromuscular dysplasia, non-specific aortitis such as Takayasu’s disease, rubella, radiation aortitis, and some autoimmune diseases. In the present case, atherosclerosis does not seem to have been a primary cause because the aortic calcification was localized with no other sclerotic lesion on the DSA and a quite similar calcification had been recognized 14 years previously. Congenital origin does not seem likely because he was healthy during adolescence and the external diameter was normal on CT. In Japan, Takayasu’s disease is well known, and the arteriographic pattern of Takayasu’s disease and atypical coarctation are similar in appearance. The coarctation in the present case is similar to a variant of Takayasu’s disease as classified by Lande and Lupi-Herrera et al. Taken together, the coarctation of the aorta might have been caused by Takayasu’s disease, presumably active in adolescence. Regardless of the etiology, hypertension seen with abdominal coarctation is a life-threatening complication necessitating aggressive medical management and occasionally, surgical intervention.

It is considered that the hypertensive heart failure in this case accounted pathophysiologically for the episode of dyspnea noted on admission. The cardiac symptoms improved relatively quickly with antihypertensive therapy.
The exact mechanism of hypertension remains obscure. It is reported that in most cases with concomitant renal artery stenosis, elevated circulating renin is the origin of the hypertension.\textsuperscript{1,18} The present case had no renal artery involvement, but plasma renin activity was elevated on admission. This was probably related to a diminished renal blood flow due to severe suprarenal aortic stenosis. No response of high blood pressure to an angiotensin converting enzyme inhibitor (enalapril) is rare in the presence of elevated plasma renin activity. In the present case, essential hypertension, not related to the renin-angiotensin system, may have been present coincidentally and factors such as fluid retention or catecholamines might predominate in the maintenance of the hypertension. The evidence that blood pressure was not completely normalized after surgery supports the possibility that essential hypertension may have been the underlying cause. In such coarctation, the renin-angiotensin system, even if circulating renin is high, may not be the primary cause of hypertension.

Angiography is mandatory in order to obtain an accurate diagnosis, to demonstrate the location of the coarctation, and to plan surgical procedures. Hallett et al\textsuperscript{19} classified the disease into four types on the basis of the anatomical location: Type I=suprarenal coarctation with renal artery stenosis; Type II=infrarenal aortic coarctation with renal artery stenosis; Type III=suprarenal aortic coarctation with normal renal arteries; and Type IV=infrarenal aortic coarctation with normal renal arteries. The present case is compatible with Type III. This type can be successfully managed by a thoracoabdominal aortic bypass resulting in retrograde aortic flow to the renal arteries, and improvement of the diminished renal blood flow.\textsuperscript{19} In the present case, postoperative DSA demonstrated retrograde blood flow into the poststenotic portion of the aorta. It was thought that the postoperative improvement of renal function was due to the resulting increase in renal blood flow.

Prognosis depends upon the severity of hypertension. In a review of the literature on patients older than 40 with coarctation of the abdominal aorta, Bergqvist et al\textsuperscript{2} reported that of 26 patients, 2 died postoperatively, while for those surviving the operation, the long-term course was favorable. The present case should be given a careful long-term follow-up in order to ascertain the patency of grafts and the control of blood pressure.

**References**