Clinical and Hemodynamic Follow-up of a Patient After Operation for Dissection of an Ascending Aortic Aneurysm Secondary to Coarctation of the Aorta

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SUMMARY

We present clinical follow-up of a 20-year-old male with an aortic aneurysm secondary to aortic coarctation. The diagnosis of aortic aneurysm secondary to aortic coarctation was made in 1997. The patient did not agree to undergo any invasive or therapeutic procedures at that time. He presented to an emergency unit with severe chest pain after chest trauma obtained during judo exercises in 1998.

Two-dimensional echocardiography showed bicuspid aortic valves, an ascending aortic aneurysm 6 cm in diameter with an intimal flap and false lumen, aortic coarctation distal to the left subclavian artery, and aortic insufficiency secondary to annular dilatation. Type II aortic dissection was confirmed by transesophageal echocardiography, which showed the dissection was confined to the ascending aorta. The dissection extended to the beginning of the arcus aorta.

Following stabilization of the patient's clinical condition, balloon coarctation angioplasty was performed to reduce afterload and hypertension and to facilitate femoral artery cannulation for cardiopulmonary bypass. Surgical procedures included resection of the aortic valve and prosthetic valve implantation, resection of the ascending aorta, and interposition of a 22 mm Hamashied tubular vascular graft.

At a follow-up visit 6 years later, the patient reported being easily fatigued and having palpitations. He had been suffering from hemolytic anemia and mild renal function impairment. Cardiac catheterisation and angiography showed a 40 mmHg gradient due to kinking of the aortic graft and no gradient at the coarctation site. We postulated the kinking of the aortic vascular graft may be related to an inappropriate vascular graft length. We also thought that the severe hemolysis was attributable to the disturbance of blood flow by a jet of blood at the site of the kinking aortic vascular graft. A second operation was performed because the renal function of the patient had decreased progressively and hemolysis symptoms increased. After the second operation, hemolysis on peripheral blood smears had disappeared and renal function had shown progressive improvements. (Int Heart J 2005; 46: 1123-1131)

Key words: Aortic coarctation, Aortic aneurysm, Aortic dissection, Hemolysis

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Aortic dissection secondary to aortic coarctation is a well known complication, however, there are few reports of surgical correction of this condition. The optimal sequence and timing of repair, the best surgical exposure, the adequacy of blood perfusion, and the most appropriate arterial cannulation site are the most important issues in the repair of aortic dissection secondary to aortic coarctation. The early and late results of aortic graft surgery are other important issues. We present a case of acute ascending aorta dissection associated with aortic coarctation which we treated successfully with first balloon aortic dilatation and second aortic graft interpositioning. Hemolytic anemia and renal function impairment were seen due to hemodynamic aortic graft kinking at 6 years follow-up, and repair of the kinking site successfully corrected the haemolytic reaction and renal function impairment.

CASE REPORT

In 1998, a 20-year-old man came to our emergency unit with complaints of severe chest pain, shortness of breath, and palpitations after chest trauma obtained during judo exercises. A diagnosis of an aortic aneurysm secondary to aortic coarctation had been made in 1997 but he did not agree to undergo any invasive procedures at that time. The diameter of the aortic aneurysm was 6 cm and the trans-coarctation gradient was 82 mm Hg. The patient was taken to the intensive care unit. Blood pressure in the upper extremities was 150/110 mm Hg and his cardiac rate was 115/min. The patient was pale, perspiring, tachypneic, and irritated. A high-grade continuous murmur was heard through the precor-
dium. There was no significant pressure difference between the upper extremities. The femoral pulses were absent and collateral circulation was not visible on the anterior precordium.

There were signs of left ventricular hypertrophy on an ECG. Left ventricular enlargement was present in chest X-rays but rib notching was not. An echocardiographic examination was conducted from precordial and suprasternal windows. The origins of the carotid and subclavian arteries were patent. Shelf-like constric-

Figure 2. Biplane, longitudinal transesophageal echocardiogram demonstrating no dissection beyond and before coarctated aortic segment.

Figure 3. Surgical view of ascending aortic aneurysm, showing intramural haematomas.
tion just beyond the origin of the left subclavian artery was located from the suprasternal window. Parasternal long and short axis windows showed a 6 cm diameter ascending aortic aneurysm, an intimal flap, and a false lumen just below it (Figure 1). The aortic valve was bicuspid and colour Doppler showed high-grade aortic valve insufficiency and turbulence within the region of aortic coarctation. Continuous wave Doppler through the same area showed a 70 mmHg gradient. The gradient persisted well into diastole, indicating more severe obstruction. The intimal flap did not extend beyond the ascending aorta. Type II aortic dissection was confirmed with transesophageal echocardiography which showed no dissection just after or before the coarcted segment (Figure 2).

Analgesia and sedation were obtained by morphine injection. Sodium nitroprusside and β-blocking agents were used for blood pressure control.

Direct hemodynamic assessment of the gradient, associated lesions, and LV pressure and function were performed via catheterisation from the femoral artery. Aortography also showed dissection confined to the ascending aorta. Balloon dilatation of the coarctation was performed with 12 and 18 mm valvuloplasty balloons. The hemodynamic gradient between proximal and distal parts of the coarctation area was reduced from 68 mmHg to 10 mmHg. Emergency cardiopulmonary bypass systems were arranged during these procedures.

We saw no dissection of the aortic annulus or coronary ostia during the operation. The aortic valve was bicuspid. The ascending aorta aneurysm diameter was nearly 9 cm and there were intramural haematomas in some areas (Figure 3). We thought that the aneurysm may have progressed to an impending rupture. The intimal tear started 3-4 cm above the aortic annulus at the anterolateral region of the aorta. The aortic annulus and coronary ostia were intact. The aortic valve was

Figure 4. Surgical view showing graft interpositioned in aortic area.
resected and an aortic prosthetic valve was implanted. Because the aortic valve was very fragile and thin, resection of the ascending aorta was started from 4-5 cm above the aortic arch and ended at the arcus level. Teflon strips were introduced to the proximal and distal endings of the aorta (Teflon sandwich reinforcement method) and a 22 mm Hamashied tubular vascular graft was interpositioned (Figure 4). Cardiopulmonary bypass was terminated with no significant bleeding problem. The patient was discharged on the tenth postoperative day. No mechanical valve or graft problems were observed during cardiac catheterisation at one year follow-up. The patient did not comply with the follow-up program and only came to follow-up program in 2004 with complaints of easy fatigue and palpitations. He had also been suffering from anemia and mild renal function impairment. His hemoglobin was 8.7 g/dL. Mean corpuscular volume and haemoglobin concentrations were below the normal limit. His reticulocyte count was 20, creatinine 1.7 mg/dL, and lactate dehydrogenase 326 IU/L (normal, 100-230 IU/L). Serum iron and ferritin levels were below the normal limits. A peripheral blood smear displayed numerous fragmented erythrocytes, haptoglobin was less than 7 mg/dL, and hemosiderinuria was seen. Direct and indirect Coombs tests were

Figure 5. Aortic angiogram 7 years after the operation showing kinking of the aortic graft.
negative. Ultrasonography showed normal spleen size. His anemia was diagnosed as a combination of iron deficiency and intravascular haemolytic anemia. Follow-up cardiac catheterisation and aortography showed kinking of the upper part of the vascular graft (Figure 5). A 40-mmHg gradient was measured at that site. Mechanical aortic valve movements were normal. No gradient was measured at the corrected segment that was previously dilated with balloon angioplasty. After correction of the anemia with a erythrocyte suspension, the complaints of the patient were reduced and the patient was scheduled for a regular follow-up. We thought that kinking of the aortic vascular graft may have been related to inappropriate vascular graft length. We also concluded that the anemia may have been related to the destruction of erythrocytes at the kinking site of the vascular graft. On 13 January 2005, the patient's complaints and symptoms had increased and renal function impairment was apparent. His hemoglobin was 7.9 g/dL and creatinine 2.2 mg/dL. Peripheral blood smears showed severe fragmentation of erythrocytes. Lactate dehydrogenase increased to 385 IU/L. A decision to undertake a second operation was made and the inappropriate graft length was shortened during the procedure. After the second operation, fragmentation on the peripheral blood smear disappeared and the creatinine level decreased to 1.7 mg/dL, while haptoglobin levels remained decreased.

**DISCUSSION**

Hypertensive vascular complications, cerebrovascular emergencies, aortic valve destruction, and aortic aneurysms are usually seen in patients with unrepaired aortic coarctation. Dissection of an aortic aneurysm secondary to aortic coarctation was first described by Jordan in 1830.6,7) The primary cause of death in unrepaired cases is aortic rupture (23%) and the location of the rupture is usually the ascending aorta (75%).8)

The diagnosis and treatment of aortic coarctation are usually made in the early decades of life and complications are rarely seen. The dissection of an ascending aortic aneurysm has high mortality and unrepaired type I aortic dissection is associated with a nearly 88% mortality rate.9,10) This mortality rate may be higher in cases with aortic dissections secondary to aortic coarctation than simple aortic dissections.

Heart valve replacement with mechanical prostheses is known to induce mild but chronic intravascular hemolysis. Decompensated haemolytic anemia is, however, currently decreasing with the development of advanced generation cardiac valve prostheses. Repeat surgery for hemolysis six years after replacement of the ascending aorta for acute aortic dissection was described by Izumi S, et al in 2003.11)
In the literature, there were 11 cases of aortic dissection secondary to aortic coarctation in whom emergency surgery was performed.\(^1-4,12-15\) Four of the 11 cases died during surgery or the early postoperative period.

Surgical procedures are relatively easy when an aortic aneurysm secondary to coarctation is not dissected. Sampath, et al first repaired a coarctation in order to reduce afterload and then repaired an aortic aneurysm in their series of 3 cases.\(^5\) Their primary aim was to preclude dissection and rupture of the aorta. The correction of the coarctation solved the cannulation and perfusion problems seeing usually in aortic coarctation. As can be expected, if an aortic aneurysm secondary to coarctation is complicated with dissection, the surgery can be more complicated than a simple aneurysm and coarctation operation. The most important problems related to a dissected aortic aneurysm are arterial cannulation and perfusion problems because dissection impedes ascending and arcus aorta cannulation. If cannulation is performed via a femoral artery, this procedure can result in perfusion problems, especially in the upper extremities and brain. In the series of Westaby, et al, it was reported that 3 cases had died and 1 case had paraplegia.\(^12\) Lawson, et al, reported a case in whom they repaired the coarctation first and then the dissected aorta.\(^1\) Plunkett, et al, reported a case in whom they initially repaired the aneurysm using femoral arterial cannulation, but in the early postoperative period they had to repair the coarctation because of refractory heart failure\(^15\) The case described by Imamura, et al, was similar both pathologically and for the invasive and operative procedures employed. In their case, coarctation angioplasty initially and then Wheat's operation for dissection had been performed.\(^13\)

The early and late results of balloon coarctation angioplasty have been described as successful in different studies.\(^16-18\)

The patient's hemodynamic stability, secured with medical therapy, led us to consider diagnostic catheterisation and coarctation angioplasty. The successful coarctation angioplasty helped us achieve effective femoral cannulation and arterial perfusion. Aortic valve insufficiency and a bicuspid aortic valve necessitated aortic valve replacement therapy in our case. We preferred resection of the dissected aortic segment and interposition of an aortic graft because the dissected aortic wall was very fragile. The proximal and distal stumps were supported with teflon strips. This kind of support prevented excessive bleeding.

The one-year follow-up cardiac catheterisation showed slight kinking of the aortic graft and a 20-mmHg hemodynamic gradient was measured at that time. There was no hemodynamic gradient at the coarctation site.

The patient came to hospital with complaints of easy fatigability and palpitations in 2004. His hematological profile showed a combination of severe intravascular hemolysis and iron deficiency. The severe hemolysis was thought to be
attributable to disturbance of blood flow by a jet of blood at the site of the kinking aortic vascular graft. Izumi, et al described a case who had severe hemolytic anemia 6 years after replacement of the ascending aorta due to type A dissection. They ascribed the hemolysis to the compressed graft.

Hemodynamic measurements, cardiac catheterisation, and aortography in our case showed apparent kinking of the aorta at the upper end and a 40 mmHg hemodynamic gradient at that site. There was no hemodynamic gradient at the coarcted site dilated with balloon angioplasty previously. A second operation was undertaken because increased hemolysis and reduced perfusion to the kidneys progressively impaired the renal function of the patient. The inappropriate graft length was shortened in the second operation. The clinical symptoms improved, renal function increased, and intravascular hemolysis signs disappeared just after the second operation.

We concluded that coarctation angioplasty before intervention for type II aortic dissection secondary to aortic coarctation may be a logical way to solve arterial cannulation and perfusion problems because they are the most important problems in this kind of emergency operation. We also believe that kinking of the aortic vascular graft may be one of the complications seen in this kind of operation, causes severe intravascular hemolysis, and necessitates a second intervention.

**REFERENCES**

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