Dissected Abdominal Aortic Aneurysm in a 24-Year-Old Female  
—Minimally Invasive Right Retroperitoneal Approach—

Shigeyoshi Gon, Takao Imazeki, Hiroshi Kiyama, Yoshihito Irie, Noriyuki Murai, 
Nobuaki Kaki, Souichi Shioguchi and Masahito Saito

A 24-year-old woman with an abdominal aortic aneurysm (AAA) caused by mucoid medial 
degeneration of the aortic wall in the absence of Marfan syndrome is reported. She required a 
Y-shaped graft replacement of the abdominal aorta through a minimal incision and recovered 

Keywords: abdominal aortic aneurysm (AAA), minimally invasive vascular surgery 
(MIVS), mucopolysaccharide

Case Report

The patient, a 24-year-old woman 154 cm in height and weighing 48 kg, was referred to our hospital with 
right waist and lower abdominal pain. At age 13, she was diagnosed with hypertension and an aplastic left 
kidney. Her blood pressure was 140/80 mmHg on 
hospitalization. A 40-mm diameter infrarenal dissect-
ed abdominal aortic aneurysm (AAA) was diagnosed 
at age 22. There was no family history of either 
Marfan syndrome or hereditary disease.

Abdominal X-ray examination showed nummular 
calcification in the right renal artery (Fig. 1). 
Abdominal CT and angiography showed the infrar-
enal AAA to be enlarged to 70 mm in diameter 
(Fig. 2).

A Y-shaped graft replacement was indicated, and 
a 7 cm transverse incision and retroperitoneal 
approach, i.e., a minimally invasive vascular surgery 
(MIVS), was performed (Fig. 3).

The location of the incision was determined by 
locating the aneurysm by ultrasonic echogram after the 
patient was anesthetized. The bowel was 
maintained within the peritoneal cavity and retracted 
to expose the aneurysm. The abdominal aorta and 
the bilateral common iliac arteries were controlled. 
After heparin (3,000 IU) was administered 
intravenously, the iliac arteries were occluded with 
balloon clamps. The aneurysmal sac was incised, 
lumbar vessels were ligated with sutures to control 
backbleeding. During proximal anastomosis, the 
surgical field was exposed by retracting the incision 
upward forward the head, while the distal retractors 
were released. For distal anastomosis, the distal site 
was exposed by retracting downwards. Dissection 
whole was exposed above bifurcation (Fig. 4).

Y-shaped graft replacement was accomplished by 
using a 16×8 mm Hemashield graft.

Operating time was 267 min, intraoperative bleed-
ing was 550 ml, and 600 ml of autologous transfusion 
was given.

Pathological examination of the aortic wall 
revealed a loss of elastic fibers, and deposition of 
mucopolysaccharide-like material (Fig. 5).

The patient didn't take an antihypertensive agent 
in a discharge, but she visit a hospital for treatment 
one in a half year.

Discussion

Employing the retroperitoneal approach to repair 
AAA offers many advantages resulting in improve-
ment of the bowel function and reduction in the length 
of postoperative hospital stay. In addition, the mini-
mal incision needed for abdominal aortic repair has 
cosmetic advantages and is less invasive.1-5) We 
performed the retroperitoneal approach because the 
patient was a young woman who wanted to return to 
work quickly.

The aorta is usually approached via the left 
retroperitoneal space because this approach provides 
easy access to the aorta below the renal artery. In 
this case, however, the abdominal aorta had shifted to 
the right, and there was no dilation of the left com-
mon iliac artery. Therefore we selected a right side 
transverse incision to approach the aneurysm.

The postoperative pain was mitigated and recovery 
was smooth. Ease of recovery was one reason for the 
decision to dissociate without cutting the abdominal 
external oblique and the rectus abdominis muscles.

The time from surgery to being able to walk to the 
lavatory was 19 h, oral intake was started within 23 h, 
and the postoperative hospital stay was 10 days.

AAA in young patients is extremely rare, and an 
abdominal aorta with localized dissection has not 
been previously reported. In this case, it is hard to
imagine that the aneurysm was caused by arteriosclerosis.

Most aneurysms in young patients are associated with congenital anomalies of the heart and great vessels or with Marfan syndrome. The patient in this case did not have ocular abnormalities or a highly arched palate or heart valve problems, and her extremities were not arachnodactylic nor was there cystic medial necrosis as seen in Marfan syndrome.

Pathological examination of the aortic wall did reveal a loss of elastic fibers, and deposition of mucopolysaccharide-like material. This is a likely cause of dissection. To the best of our knowledge, there are only 2 reports of aortic dissection caused by the deposition of mucopolysaccharide-like material in the absence of Marfan syndrome.6,7) Marcello postulated a genetically determined disease of connective tissue usually described as Marfan’s forme fruste.6)

Although familial dissecting aortic aneurysms in the absence of Marfan syndrome have been reported, in most cases, the aorta showed disruption of elastic fibers and cystic medial necrosis.8-11) Cystic medial necrosis has been shown in healthy persons and is related to age and hypertension.12,13) As Tobinaga reported, cystic medial necrosis is not specific to Marfan syndrome.

Our patient did not have Marfan syndrome, and dissection may have been caused by renovascular hypertension and deposition of the mucopolysaccharide-like material in the aortic wall. To determine the possibility of a hereditary connective tissue disease, it will be necessary to scrutinize the patient’s family history.
Fig. 5 Elastic van Gieson stain (×100) and Alcian blue stain (×400)

Pathological examination of the aortic wall revealed a loss of elastic fibers. Alcian blue stain revealed deposition of mucopolysaccaride-like material.

References


