

## Rupture of a Normal-Sized, Non-Dissected Distal Aortic Arch in a Marfan Patient

Yuji Maruyama, MD,<sup>1</sup> Masami Ochi, MD,<sup>1</sup> Ryuzo Bessho, MD,<sup>1</sup> Kenichi Yamada, MD,<sup>1</sup> Yosuke Ishii, MD,<sup>1</sup> Masahiro Fujii, MD,<sup>1</sup> Koichi Tamura, MD,<sup>2</sup> and Kazuo Shimizu, MD<sup>1</sup>

**We successfully repaired a rupture of a normal-sized, non-dissected distal aortic arch in a patient with Marfan syndrome. Six years previously she had undergone repair of the thoraco-abdominal aortic aneurysm with a 24-mm knitted Dacron graft for type B chronic aortic dissection. The rupture site was located at the back of the native distal aortic arch just 10 mm above the proximal anastomosis, and just below the left subclavian artery. This unexpected situation might be related to dilatation of the knitted Dacron graft up to 34 mm (142%), thus stretching out the fragile native aorta in this Marfan patient. (Ann Thorac Cardiovasc Surg 2006; 12: 438–40)**

**Key words:** Marfan syndrome, aortic arch, aortic surgery, rupture

### Introduction

The development of surgical therapies to repair aortic aneurysms would be predicted to prolong the life expectancy of patients with Marfan syndrome by preventing sudden death due to aortic rupture.<sup>1)</sup> A high incidence of subsequent operations on the remaining aorta has been observed among patients with Marfan syndrome. This is especially true in cases in which the site of the initial operation was more frequently the descending aorta than the ascending aorta including the arch.<sup>1,2)</sup> However, rupture of a normal-sized, non-dissected aorta following the first operation in a Marfan patient is very rare. We report on a case of a rupture of a normal-sized, non-dissected distal aortic arch after replacement of the descending aorta in a Marfan patient.

### Case

A 41-year-old woman with Marfan syndrome with a his-

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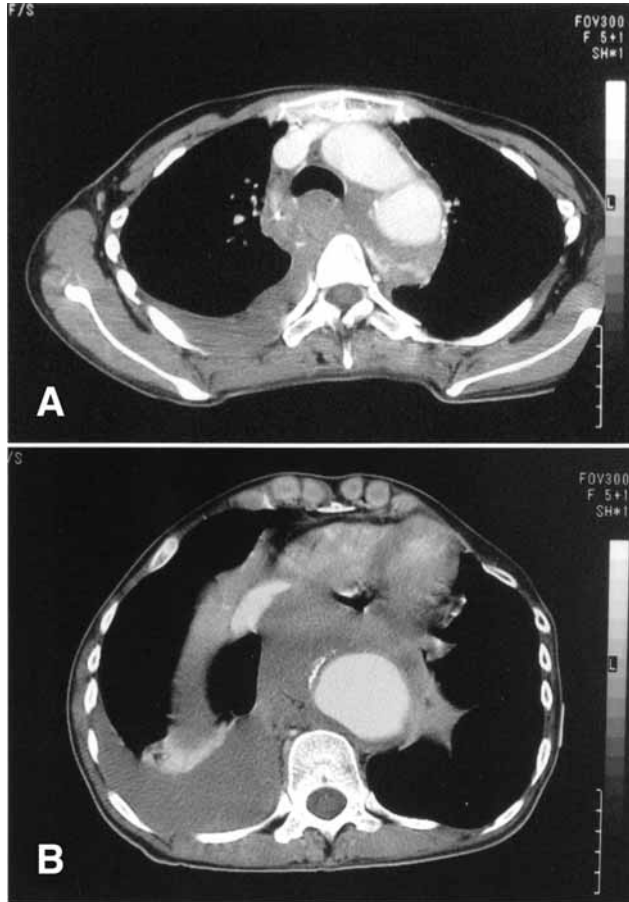
*From <sup>1</sup>Division of Cardiovascular Surgery, Department of Surgery, and <sup>2</sup>Department of Pathology, Nippon Medical School, Tokyo, Japan*

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Address reprint requests to Yuji Maruyama, MD: Division of Cardiovascular Surgery, Department of Surgery, Nippon Medical School, 1-1-5 Sendagi, Bunkyo-ku, Tokyo 113-8603, Japan.

tory of aortic disease was admitted with severe back pain. Six years previously, this patient underwent a prosthetic graft replacement, placing a 24-mm knitted Dacron graft, of the descending aorta from below the left subclavian artery to the bifurcated portion for type B chronic aortic dissection. At that time, a proximal anastomosis was performed using an open proximal procedure. Two pairs of intercostal arteries were connected side-to-side with the aortic prosthesis using a patch of the native aorta. The visceral arteries were each anastomosed end-to-end to a 10-mm knitted Dacron graft. Computed tomography revealed a massive hematoma around the aortic prosthesis, but the point of extravasation was unclear (Fig. 1). There was a normal-sized and non-dissected ascending aorta including the arch and no anastomotic aneurysm. The aortic prosthesis was dilated up to 42 mm (175%) in diameter at the level of the reconstruction of the intercostal arteries. An angiography demonstrated the point of extravasation was around the proximal anastomosis of the previous surgery and the aortic prosthesis was dilated up to 34 mm (142%) in diameter at the level of the proximal anastomosis (Fig. 2). Trans-thoracic echocardiography revealed no aortic insufficiency and a normal aortic root.

An emergency operation was conducted through a median sternotomy. Extracorporeal circulation was established by cannulating the femoral artery and vein. Total arch replacement with open distal anastomosis under pro-



**Fig. 1.** Computed tomography showed that (A) the aortic prosthesis was dilated up to 34 mm (142%) in diameter at the level of the proximal anastomosis and (B) 42 mm (175%) in diameter at the level of the reconstruction of the intercostal arteries and a massive hematoma around the aortic prosthesis.

found hypothermic circulatory arrest with antegrade selective cerebral perfusion was performed. The rupture site was located at the back of the native distal aortic arch just 10 mm above the previous proximal anastomosis, and just below the left subclavian artery. The native aorta was circumferentially torn, 12 mm in diameter, and was surrounded with massive fresh thrombi outside of the aortic wall. A distal anastomosis was completed using a 30-mm 4-branched woven Dacron graft connected end-to-end to the dilated aortic prosthesis of the previous surgery. A proximal anastomosis was connected to an ascending aorta reinforced with a felt strip. Therefore, a prosthetic graft replacement of the entire aorta except for the aortic root was completed. The post-operative course was uneventful. Pathologically, the resected aortic wall showed marked thinning and fragmentation of the medial elastic lamellae.

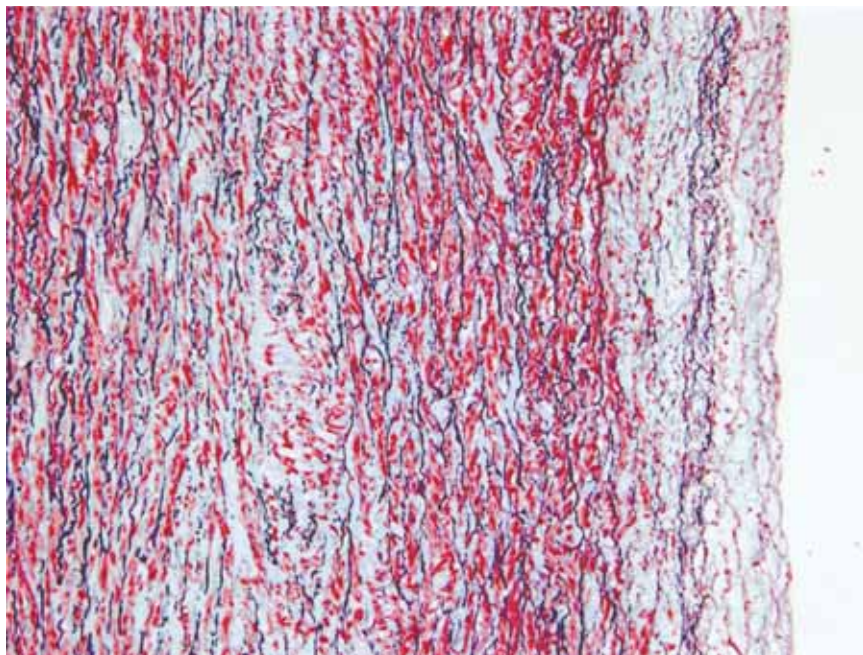


**Fig. 2.** Angiography showed the point of extravasation was around the proximal anastomosis of the previous surgery (arrow) and the aortic prosthesis was dilated up to 34 mm (142%) in diameter at the level of the proximal anastomosis.

Cystic medial necrosis was also observed. These were typical findings in the aorta of Marfan syndrome, suggesting fragility of the aortic wall. The rupture could not be confirmed in the histological specimen (Fig. 3).

## Discussion

The development of surgical therapies to repair aortic aneurysms would be expected to prolong the life expectancy of patients with Marfan syndrome by preventing sudden death due to aortic rupture.<sup>1)</sup> Although a high incidence of subsequent operations on the remaining aorta has been observed among the patients with Marfan syndrome, rupture of normal-sized, non-dissected distal aortic arch following the first operation in a Marfan patient is very rare.<sup>2)</sup> Carrel et al. reported on 2 patients who died from rupture of a normal-sized, non-dissected descending aorta following uncomplicated aortic root operation in 71 Marfan patients who received the initial surgery.<sup>3)</sup> Both had undergone minor surgery some days before rupture (one had an inguinal hernia repair, and the other partial excision and laser of a facial nevus flammeus). Arterial hypertension was not present in any of these 2 patients. Long-term survival and complications in Marfan patients are reported in the literature, including



**Fig. 3.** The resected aortic wall showed marked thinning and fragmentation of the medial elastic lamellae. Cystic medial necrosis was also observed. These were typical findings in the aorta of Marfan syndrome, suggesting fragility of the aortic wall. The tear of rupture could not be confirmed in the histological specimen. (elastica Masson-Goldner stain)

271 cases by Gott et al.,<sup>4)</sup> 192 cases by Finkbohner et al.<sup>1)</sup> and 78 cases by Tambour et al.<sup>5)</sup> Rupture of a normal-sized, non-dissected aorta following the first operation was not observed. An unexpected fatal rupture may occur in the downstream aorta following uncomplicated aortic surgery despite the presence of a normal-sized aorta. It is expected that patients with Marfan syndrome will require multiple interventions on the remaining aorta.

This unexpected situation in our case might be related to dilatation of the knitted Dacron graft up to 34 mm (142%), thus stretching out the fragile native aorta in this Marfan patient. Adachi et al. and Blumenberg et al. reported that the ratio of dilatation was 125–136% in a knitted Dacron graft higher than 105% in a woven Dacron graft, respectively.<sup>6,7)</sup> Sako et al. demonstrated that dilatation of a knitted Dacron graft for the descending thoracic aorta was related to a new dissection above the proximal anastomosis.<sup>8)</sup> In Marfan patients, a woven Dacron graft may be useful as an aortic prosthesis in view of preventing dilatation. In cases in which a knitted Dacron graft is selected as an aortic prosthesis, the use of a down-sized prosthesis may be recommended.

We successfully rescued a Marfan patient who had an aortic rupture of a normal-sized, non-dissected distal aortic arch after replacement of the descending aorta. This unexpected rupture might be related to dilatation of the knitted Dacron graft up to 34 mm (142%), thus stretching out the fragile native aorta in this Marfan patient.

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