Proximal aortic perforation after endovascular repair of a type B dissection in a patient with Marfan syndrome

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Several successful cases of endovascular treatment of type B dissections in patients with Marfan syndrome have been reported. In our patient with Marfan syndrome, a type B dissection was successfully treated endovascularly. Three weeks after this procedure, a computed tomographic angiography (CTA) revealed a perforation of the aortic wall distal to the left subclavian artery by a bare strut of the stent graft. A second stent graft was placed to treat this complication. In patients with Marfan syndrome, complications might be prevented by using stent grafts specifically developed to treat dissections. However, specific complications, eg, perforation, must be taken into account and patients have to be followed attentively. (J Vasc Surg 2009;50:190-2.)

Marfan syndrome is an autosomal dominant connective tissue disorder that typically involves the cardiovascular, skeletal, and ocular systems. This genetic disorder is caused by mutations on the fibrillin gene, located on chromosome 15q. Vascular abnormalities, notably aortic dilation and dissection, are the most life-threatening manifestations of Marfan syndrome.[1] Recently, several technically successful and uneventful endovascular treatments of aortic dissections in patients with Marfan syndrome have been reported.[2-4] However, according to the instructions for use of stent grafts, patients with connective tissue disorders are not suitable for endovascular repair. We present a case wherein endovascular repair of a type B dissection in a patient with Marfan syndrome is complicated by a proximal strut perforation through the aortic wall.

CASE REPORT

A 55-year-old male, in 1992 diagnosed to have Marfan syndrome, presented with acute onset of severe back-pain, especially between his shoulders. A computed tomographic angiography scan (CTA) revealed a type B aortic dissection: the intimal flap originated just distal to the origin of the left subclavian artery and extended into the right iliac artery (Fig 1). A first entry site between the true and false lumen was seen 10 cm distal to the left subclavian artery (LSCA), a second 12 cm distal to the LSCA. The aorta proximal to the dissection had a diameter of 29 mm. The maximum diameter of the thoracic aorta was 40 mm (True lumen 22 mm), while the abdominal aorta had a maximum diameter of 37 mm (True lumen 17 mm). No stenoses were seen in the vertebral and carotid arteries.

Since the patient was hemodynamically stable, conservative treatment was started to decrease the systolic blood pressure below 130 mm Hg (analgesics, Acetaminophen and Adolan, and a combination of three anti-hypertensive agents, Triamterene, Perindopril, Metoprolol, were given. Metroprolol was exchanged for Labetalol hydrochloride after several days).

Six days after admission, the patient became hemodynamically unstable and the back-pain aggravated. On CTA, a rupture of the dissection in the descending thoracic aorta was seen. Therefore, the dissection entry points were excluded with two Valiant stent graft segments (Medtronic, Minneapolis, Minn); Proximal stent graft: diameter of 34 mm; length of 200 mm. Distal stent graft: diameter of 36 mm; length of 150 mm. The stent graft was inserted via the left femoral artery, and angiography was performed via the right brachial artery. Stent graft type and sizing was not considered ideal: we had preferred to use a less oversized non-bare stent graft. However, in the acute setting, such a stent graft was unavailable in our hospital. We deployed the stent graft just below the LSCA since the stent graft diameter was considered too big to overstent the LSCA. Besides, the most proximal detected entry was seen 10 cm below the LSCA. We decided to overstent the aorta with two stent graft segments to be sure that also possibly undetected entry sites were overstented. A spinal drainage device was placed direct postoperatively.

Postprocedural CTA showed closure of the intimal tear (Fig 2, *A*). Three weeks after the procedure, the back pain suddenly increased and a CTA was made, revealing a proximal pseudoaneurysm caused by a perforation of a proximal bare strut of the stent through the vessel wall (Fig 2, *B*).

The pseudoaneurysm was treated by endovascular placement of an extra stent, a Relay non-bare stent, 32 mm in diameter and 164 mm long (Bolton medical, Sunrise, Fla) (Insertion via the left femoral artery, angiography via the right brachial artery). The size of this stent graft was based on measurements proximal to the LSCA.

The periprocedural angiography confirmed the perforation (Fig 3). The Relay none-bare stent, which is especially designed for treatment of dissections, was placed between the left common carotid artery and the LSCA. A spinal drainage device was placed preoperatively. We did not revascularize the LSCA in this procedure primarily since we expected the risk on myelum ischemia to be

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Competition of interest: none.

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0741-5214/$36.00

doi:10.1016/j.jvs.2009.01.045

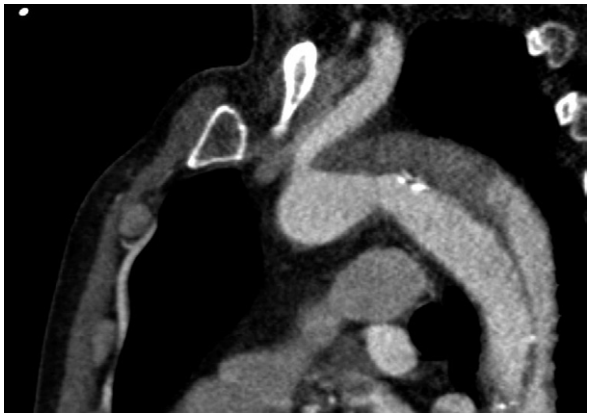

Fig 1. Computed tomographic angiography (CTA) of the thoracic aorta showing a type B dissection, sagittal view.

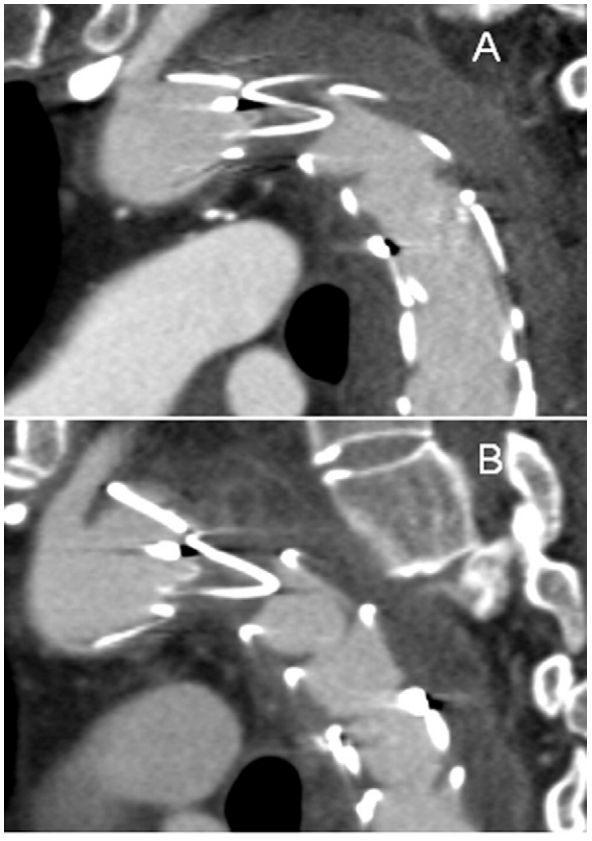

Fig 2. A, Computed tomographic angiography (CTA) performed after the first procedure, sagittal view. Successful closure of the primary intimal tear was seen. **B,** CTA performed 3 weeks after the first procedure, sagittal view. A proximal pseudoaneurysm, based on perforation of a proximal bare strut through the aortic wall, was seen.

very low in this patient with patent vertebral arteries and without severe atherosclerosis. A postprocedural CTA showed exclusion of the pseudoaneurysm (Fig 4).

After the reintervention, the back pain had disappeared. Four days after the second intervention, there were no more medical indications to keep the patient in the hospital. However, the patient was not dismissed out of the hospital until 20 days after the second intervention since no domiciliary care was available. Until now, 110 days after the second operation, no new complications have occurred and CTA reveals no abnormalities (Fig 4).

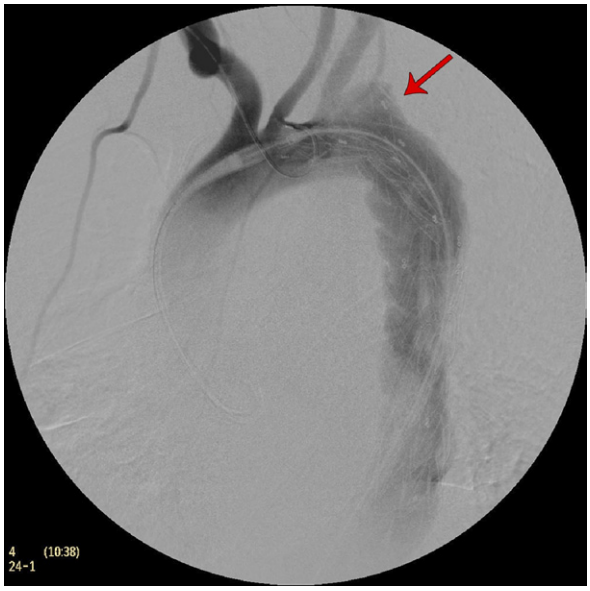

Fig 3. Angiography performed before deployment of the second stent graft. Note the obvious dislocation of the first stent graft with perforation of the aortic wall and contrast extravasation.

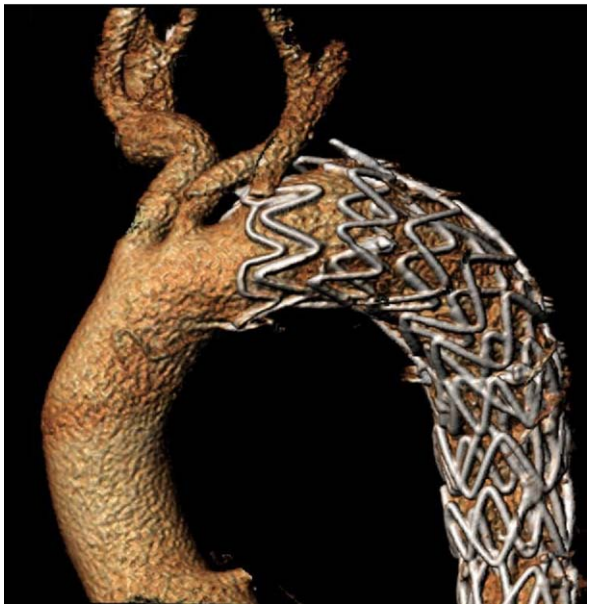

Fig 4. Computed tomographic angiography (CTA) of the thoracic aorta with three-dimensional volumetric reconstruction that shows successful exclusion of the pseudoaneurysm.

DISCUSSION

Complicated type B dissection in patients with Marfan syndrome requires surgical intervention.[5] However, conventional surgery for descending aortic dissection is unfavorable in patients with substantial comorbidity, and has a high mortality rate, ranging from 8 % to 57%.[6] An alternative to open repair is endovascular treatment, which is feasible and can be technically successful in patients with Marfan syndrome.[3] Although safe stent graft placements in patients with Marfan syndrome are published, evidence of long-term success is missing, due to the small number of published cases and limited follow-up.[4] In addition, publication bias has probably led to under-reporting of less successful cases. Next to this, it was stated in the 2008 expert consensus that endovascular repair in patients with Marfan syndrome is not recommended unless operative intervention is clearly indicated and the risk of conventional repair is deemed prohibitive.[7]

Although the placement of a stent graft can be technically successful, Marfan syndrome patients are intuitively more prone to several specific complications after endovascular repair. Aortic dilation, which frequently occurs in patients with Marfan syndrome,[1] may be manifest around fixation and sealing points of the endograft allowing for subsequent stent graft migration or periprosthetic leaks. Next to this, the mutation in the fibrillin gene in patients with Marfan syndrome results in weakened soft tissue, which thus makes the aortic wall more fragile. Therefore, the aortic wall may not withstand the pressure of the stent graft, with perforation as consequence. To the best of our knowledge, this is the first report on a perforation of the aortic wall by a stent graft in a patient with Marfan syndrome.

To prevent complications, we believe that endovascular treatment of aortic dissections requires specific types of stent grafts. Flexibility and radial expansion characteristics of stent grafts used for treatment of aortic dissections should be different than those used for treatment of aortic aneurysms. With treatment of aneurysms, a high localized radial force is essential at the relative short proximal and distal landing zones. With dissections, however, the stent graft most commonly has apposition over its entire length: less proximal and distal radial force is probably needed. Besides, the fragile aortic wall in patients with dissections, and especially in patients with Marfan syndrome, could be easily injured by bare struts and high radial force. This has led to our assumption that endografts with non-bare struts are preferable in patients with a dissection. Next to the use of stent grafts specifically designed to treat dissections, we advise to oversize stent grafts about 5% in patients with a dissection. More oversizing of stent grafts increases radial force, and the struts might perforate the fragile aortic wall.

Retrospectively, it is not unimaginable that the perforation of the bare strut through the aorta could have been prevented in this case. Overstenting of only the proximal entry site leads to a vulnerable stent graft landing zone: the aortic wall of a Marfan syndrome patient even weakened by a dissection. Therefore, we believe that overstenting of the entire initial dissection is possibly a better method than just overstenting the proximal entry site between the true and false lumen in patients with a connective tissue disease.

In the follow-up of Marfan patients after endovascular exclusion of a type B dissection, we recommend to perform CTAs regularly: within a few days postoperative, after 3 and 9 months, and thereafter yearly.

In conclusion, endovascular treatment of type B dissections in patients with Marfan syndrome should only be considered when operative intervention is clearly indicated, eg, in case of rupture. The risks on complications in patients with Marfan syndrome after endovascular treatment might be lowered by the use of (non-bare) stent grafts specifically developed to treat dissections. Moreover, Marfan syndrome patients who are treated endovascularly have to be monitored closely. Specific complications in patients with Marfan syndrome after endovascular repair of a type B dissection must be taken into account to permit, if necessary, early reintervention.

The authors thank Joffrey van Prehn, MD, and Gert J. de Borst, MD, PhD, for their helpful comments on a previous draft of this manuscript.

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