We report a case of chronic aortic dissection in a patient with Marfan syndrome in which we performed thoracic endovascular repair after aortic root replacement, total arch replacement with open stent grafting and thoracoabdominal aortic repair. We consider that endovascular repair of the dissected descending aorta in a patient with Marfan syndrome can be effective when graft-to-graft bridging is performed as the “finishing procedure”.

Keywords: endovascular repair, aortic dissection, Marfan syndrome

Introduction

The treatment of thoracic aortic disease has changed with advances in endovascular therapy. Endovascular therapy has emerged as a viable alternative to open surgical repair with less perioperative mortality and morbidity in patients with Marfan syndrome. However, the feasibility of endovascular repair in patients with Marfan syndrome is controversial because of a high rate of endoleak and the need for re-intervention.1)

Case

The patient is a 51-year old woman with Marfan syndrome who initially presented 11 year prior with an acute Type A aortic dissection, for which she underwent total arch replacement and open stent grafting. Eight years later, she again presented with an acute type B aortic dissection. At that time the diameter of the thoracoabdominal aorta was 60 mm thus she underwent open thoracoabdominal aortic aneurysm repair (Coselli Thoracoabdominal Graft). Three years later, we performed aortic root replacement by a modified Bentall procedure (Trifecta aortic valve, Gelweave Valsalva) because of annuloaortic ectasia. After that, the diameter of the descending aorta gradually dilated, and we decided to perform stent graft repair.

Computed tomography showed dissection of the descending thoracic aorta with a maximum diameter of 46 mm, and the diameter of the true lumen was 10 mm (Figs. IA, and IB). There was thrombosis of the false lumen above the level of T9, and an Adamkiewicz artery was seen at the level of T10. There were small entry sites tears at the level of T10–12.

The descending thoracic aorta was stented with two stent grafts (Conformable GORE® TAG®) inserted through the right femoral artery. The proximal stent was 31 mm in diameter and 200 mm in length, and the distal stent was 28 mm in diameter and 200 mm in length. The proximal landing zone was an open-stent graft, and the distal landing zone was a Coselli thoracoabdominal graft.

Her postoperative course was uneventful, and enhanced CT showed complete thrombosis of the false lumen and no endoleak. Enhanced CT one year after surgery showed good expansion of the stent grafts and shrinkage of the false lumen (Figs. 2A, and 2B).

Discussion

Marfan syndrome is a heritable connective tissue disorder that results in aneurysm formation and dissection as its most severe complication.1,2) It is known that patients with Marfan syndrome continue to experience aortic degeneration throughout their lives, leading to remedial aortic interventions. As Marfan patients who have been treated with standard open operations advance in age and experience progression of their aortic pathology, the operative risk markedly increases. Open surgical treatment of proximal descending aortic dissection is associated with significant morbidity.9) The advent of endovascular repair provides an attractive alternative for patients who are high risk for open operations.
However, the feasibility of endovascular repair in patients with Marfan syndrome is widely debated because of the potential risk of aortic dilation induced by radial forces of the stent graft on the fragile aortic wall. Geisbusch et al. reported that an aortic dissection continued to dilate despite endograft exclusion of the aneurysm in patients with Marfan syndrome, and this resulted in endoleak.\textsuperscript{4}

Several reports of endovascular management of the descending aorta in patients with Marfan syndrome have been published, and small observational studies have shown that it is feasible and relatively safe.\textsuperscript{1–3,5–7}

Pacini et al.\textsuperscript{1} analyzed acute and chronic type B aortic dissection in 54 Marfan syndrome patients after stent grafting. They reported that the post-procedural endoleak rate was 22\%, and that the mortality during hospitalization was 2.5\%. During the follow-up period (average 2.5 years), mortality occurred in six (11.8\%) patients, and surgical conversion was required in seven (14\%) patients. Because of these high rates of mid-term mortality and surgical conversion, the authors concluded that stent grafting for type B dissection in Marfan patients, especially in chronic conditions, should be considered with great caution. However, they also reported that no endoleak occurred when the proximal landing zone was in a previously implanted graft. This is because the stent graft was deployed into a replacement aortic segment that provided a stable platform, which reduced the incidence of type 1 endoleaks.\textsuperscript{5}

We believe that it is not sufficient to just prevent dilation of the dissected aorta, because it is possible that progression of dissection or new dissection might be induced by the radial forces of the stent graft on the aortic wall when the distal landing zone is in the dissected aorta. Then a mismatch between the stent graft and aortic wall could occur, resulting in endoleak.

When both the proximal and distal landing zones are in locations where there are previously implanted grafts, including an open stent graft and elephant trunk, we believe it is unlikely that there will be new dissection, progression of the previous dissection or dilation of the aortic wall.
In this case, the patient had decreased strength because she underwent surgery three times. Therefore, stent graft repair was selected as a less invasive treatment than left thoracotomy. In addition, the stent graft was deployed into the previously implanted graft, which provided a stable platform for both the proximal and distal landing zones. Furthermore, enhanced CT showed shrinkage of the false lumen one year after surgery. Type II endoleak in the false lumen was not seen after surgery. However, the incidence rate of type II endoleak in the false lumen was unclear when the stent graft was deployed in the true lumen, and long-term follow up is needed.

This case had high risk of spinal cord injury because this procedure might occlude the blood flow from the intercostal arteries to the spinal cord. Spinal cord blood flow depends on the blood reserve from the collateral arteries and we think that maintenance of mean arterial pressure over 90 mmHg, correction of anemia and O₂ saturation and fluid overload are important to prevent spinal cord injury after surgery.

Conclusion

We consider that endovascular repair of the dissected descending aorta in a patient with Marfan syndrome can be effective when graft-to-graft bridging is performed as the “finishing procedure”. However, the long-term results of this procedure are not yet available, and strict follow-up is needed.

Disclosure Statement

None declared.

Author Contributions

Study conception: YY, NU
Writing: YY
Critical review and revision: all authors
Final approval of the article: all authors
Accountability for all aspects of the work: all authors

References