Vascular Specialist International
Vol. 33, No. 3, September 2017
pISSN 2288-7970 • eISSN 2288-7989

Case Report

Sequential Hybrid Repair of Aorta and Bilateral Common Iliac Arteries Secondary to Chronic Aortic Dissection with Extensive Aneurysmal Degeneration in a Marfan Patient

Carlos A. Hinojosa, Javier E. Anaya-Ayala, Hugo Laparra-Escareno, Rene Lizola, and Adriana Torres-Machorro

Department of Surgery, Section of Vascular Surgery and Endovascular Therapy, Instituto Nacional de Ciencias Médicas y Nutrición Salvador Zubirán, Mexico City, Mexico

Marfan syndrome is a connective tissue disorder associated with aortic dissection, aneurysmal degeneration and rupture. These cardiovascular complications represent the main cause of mortality, therefore repair is indicated. We present a 35-year-old woman who experienced acute onset of chest pain. Her imaging revealed a chronic DeBakey type I dissection with aortic root dilation and descending thoracic aneurysmal degeneration. She underwent a Bentall procedure and endovascular exclusion of the descending thoracic aortic aneurysm. She was closely followed and 2 years later a computed tomography angiography (CTA) revealed the aneurysmal degeneration of the thoracoabominal aorta and bilateral iliac arteries. The patient underwent a composite reconstruction using multivisceral branched and bifurcated Dacron grafts. At 5 years from her last surgery, a CTA revealed no new dissection or further aneurysmal degenerations. Aortic disease in Marfan patients is a complex clinical problem that may lead to secondary or tertiary aortic reconstructions; close follow-up is mandatory.

Key Words: Marfan syndrome, Chronic aortic dissection, Aneurysmal degeneration, Hybrid repair

Received May 10, 2017
Revised June 9, 2017
Accepted June 19, 2017

Corresponding author: Carlos A. Hinojosa
Department of Surgery, Section of Vascular Surgery and Endovascular Therapy, Instituto Nacional de Ciencias Médicas y Nutrición Salvador Zubirán, Vasco De Quiroga 15, Tlalpan, Sección XVI, Mexico City 14080, Mexico
Tel: 01152 55 5487 0900
Fax: 01152 55 5487 0900
E-mail: carlos.a.hinojosa@gmail.com
Conflict of interest: None.

Vasc Spec Int 2017;33(3):112-116 • https://doi.org/10.5758/vsi.2017.33.3.112

INTRODUCTION

Marfan Syndrome (MS) is a systemic disorder of connective tissue caused by mutations in the extracellular matrix protein fibrillin-1 [1]. Aortic dissection, aneurysmal degeneration and subsequent rupture remain the most life-threatening clinical manifestations of this syndrome, and without intervention death may occur in the early adulthood [2]. Acute ascending aortic dissection is an indication for surgical treatment, but despite operative success and improved survival after repair of the ascending aorta, these patients continue to experience aortic aneurysmal degeneration throughout their lives leading to the need for secondary or tertiary interventions of the aorta [3].

We herein present the case of a young woman with MS who suffered a type I DeBakey aortic dissection with subsequent aneurysmal degeneration of the ascending and thoracoabdominal aorta, and the common iliac arteries requiring complex open and endovascular reconstructions. We briefly discuss the clinical presentation, surgical planning, techniques and clinical outcome.

CASE

A 35-year-old woman with clinical features consistent with MS including tall stature (1.95 m), arachnodactyly and mild scoliosis presented to the emergency department with acute onset of chest pain in 2008. The year before she had experienced an episode of chest pain, but she did not seek medical attention at that time. Upon arrival a full work-up was initiated and a computed tomography angiography (CTA) revealed chronic DeBakey type I aortic dissection and aneurysmal degeneration of the aortic root measuring 7.74 cm and descending thoracic aorta (measurement 6.05 cm in maximum diameter) in axial view associated to a spiral dissection extending to both common iliac arteries (Fig. 1-3). Interestingly the patient had a bovine aortic arch with left carotid artery emerging from the innominate artery; the dissection followed the inner curvature of the aortic arch and it did not extend to the supra-aortic vessels. Aneurysmal degeneration of the aortic arch was not present (Fig. 3). An echocardiogram confirmed aortic valve insufficiency. She was taken to the operating room for Bentall and De Bono procedure, replacing the aortic root and valve with a mechanical prosthesis, the patient was discharged home on postoperative day nine on systemic anticoagulation. Two months later, she underwent endovascular treatment of descending thoracic aorta using a 200×40 mm Zenith (Cook Inc., Bloomington, IN, USA) thoracic stent-graft that successfully isolated the aneurysm from the circulation.

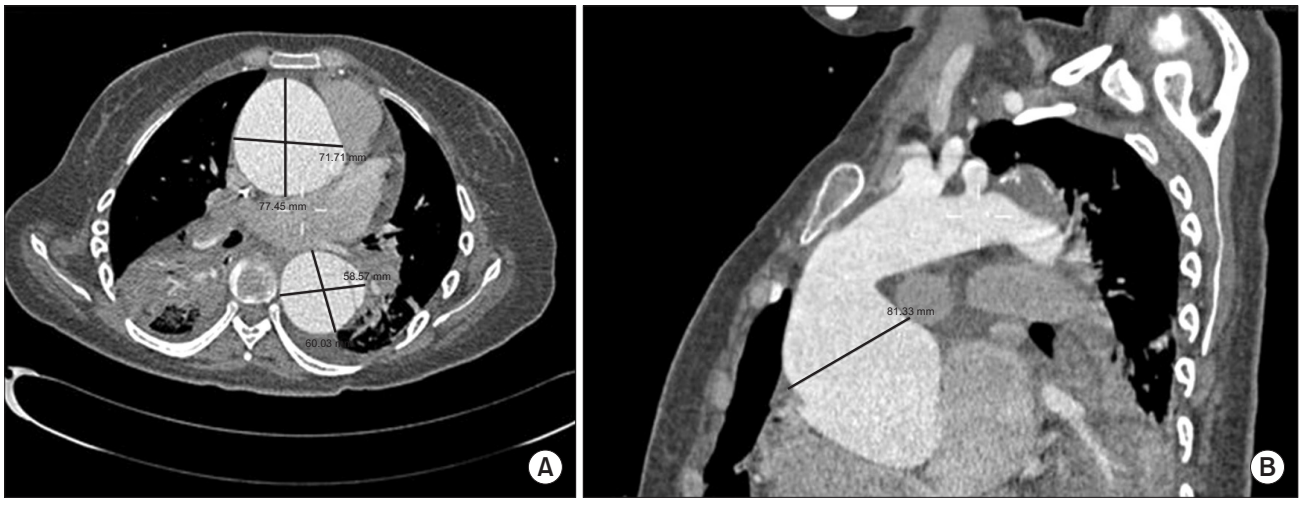

Fig. 1. Chest computed tomography angiography demonstrates the maximum diameters of the ascending aorta in (A) axial view (7.74 cm) and (B) lateral view (8.13 cm).

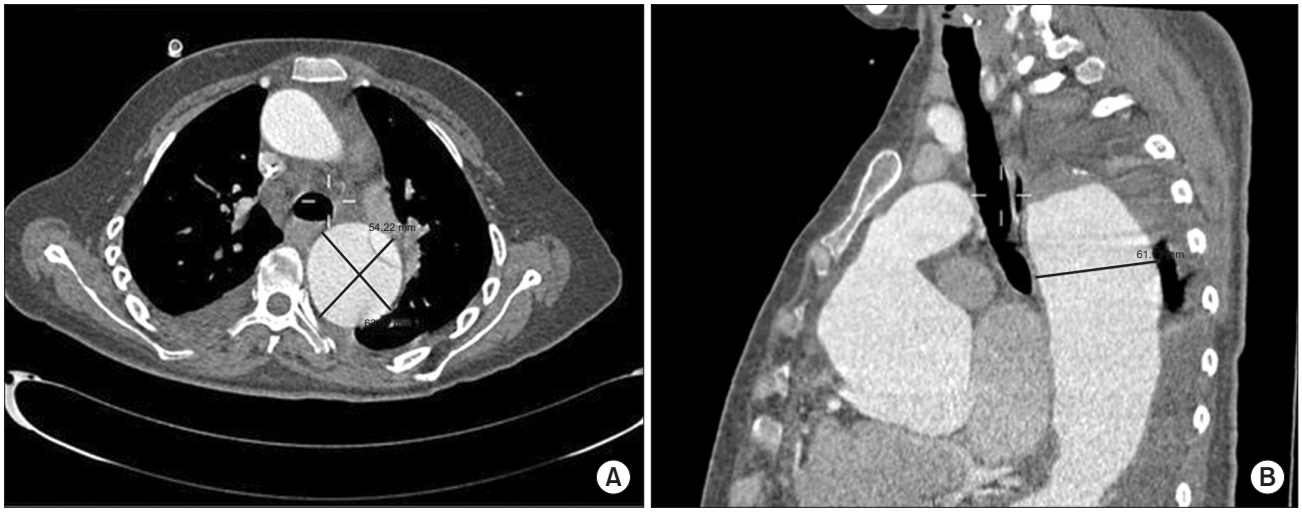

Fig. 2. Chest computed tomography angiography shows the maximum diameters of the descending thoracic aorta in (A) axial view (6.6 cm) and (B) lateral view (6.1 cm).

https://doi.org/10.5758/vsi.2017.33.3.112

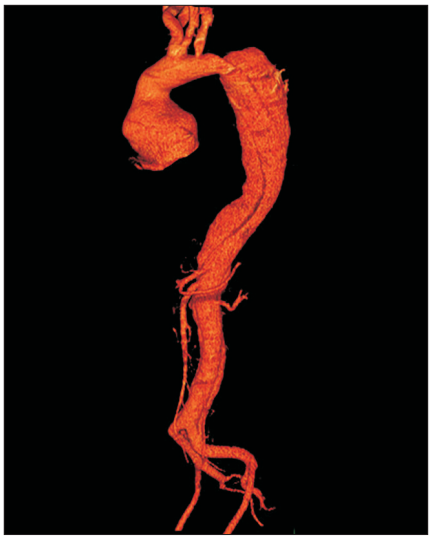

Fig. 3. Computed tomography angiography three dimensional reconstruction of the aorta demonstrates the type 1 DeBakey aortic dissection and aneurysmatic segments in the ascending and the descending thoracic aorta.

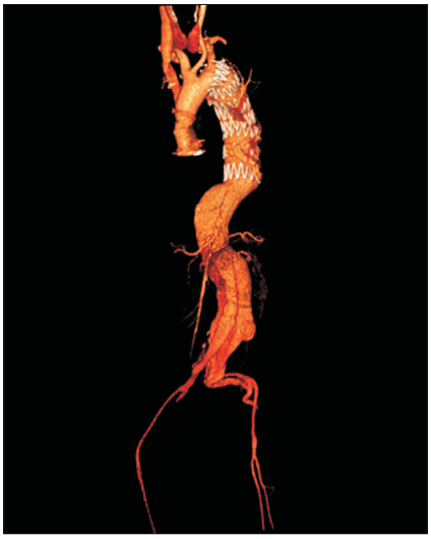

Fig. 4. Follow-up computed tomography angiography three dimensional reconstruction of the aorta from 2012, revealed the chronic dissection and extensive aneurysmal degeneration of the thoracoadominal aorta (measuring 6.4 cm) extending to both common iliac arteries (measuring 2.3 cm in the right and 2.9 in the left).

The distal thoracic aorta measured 3.3 cm and infrarenal portion 2.9, the decision was to maintain the patient under close surveillance. She was carefully followed in our institution by a multidisciplinary team, continuing her antihypertensive medical management with optimal blood pressure control. In 2012, a repeat CTA demonstrated aneurysm expansion of the thoracoadominal aorta measuring 6.4 cm and both common iliac arteries (measuring 2.3 cm in the right and 2.9 in the left) with imminent risk for rupture (Fig. 4). After discussions regarding the risks and benefits with the patient and her family, she was taken to the operating room for thoracoabdominal aortic and bilateral iliac arteries aneurysm repair.

The patient was placed in left lateral decubitus, a drain was placed for spinal cord protection, and subsequently a left thoracoabdominal incision was made via the 7th intercostal space crossing the costal margin and brought downward through the abdominal midline line ending just above the pubic symphysis. The left hemidiaphragm was circumferentially incised along its parietal insertion and the peritoneal sac was detached from the abdominal wall, the viscera were moved medially and the thoracoabominal aorta was totally exposed. The thoracic aorta endograft was clamped just above the diaphragm and reconstructions of the aorta and vessels were performed using a 22 mm five-branched Dacron graft, with the proximal end anastomosed to the stentgraft using pledgets and the graft branches to both renal arteries, celiac trunk, superior mesenteric artery and finally to the artery of Adamkiewicz (Fig. 5A). Finally a bifurcated 20×10 mm graft was anastomosed to the distal tube graft and its limbs distally to the common iliac arteries bilaterally (Fig. 5B). Total circulation was reestablished and hemostasis was ensured, and subsequently the thorax and abdomen were closed by layers. The surgery was completed successfully without complication and her hospitalization course was uncomplicated. The patient was discharged in stable condition in postoperative day 12 with preserved kidney function and without neurologic deficits in her lower extremities. The patient continued to be imaged in annual basis to rule out recurrent aneurysms or graft stenosis. Fig. 6 demonstrate a CTA with three dimensional reconstruction of the aorta without new dissection or aneurysmal degenerations, and a schematic drawing of the complex hybrid repair with the prosthetic material utilized for the aorto-iliac reconstructions.

DISCUSSION

The expanding knowledge in this rare connective tissue disorder has demonstrated that up to 20% to 40% of the aortic dissections may progress to aneurysmal degeneration of the aorta, often resulting in the formation of extensive thoracoabdominal aortic aneurysms (TAAA) [4,5]. Over the last 30 years the medical and surgical management has improved substantially, allowing MS patients to survive an acute ascending aortic dissection; and this increase in the life expectancy has resulted in the emergence of descending aortic complications [6]. If untreated, MS patients with

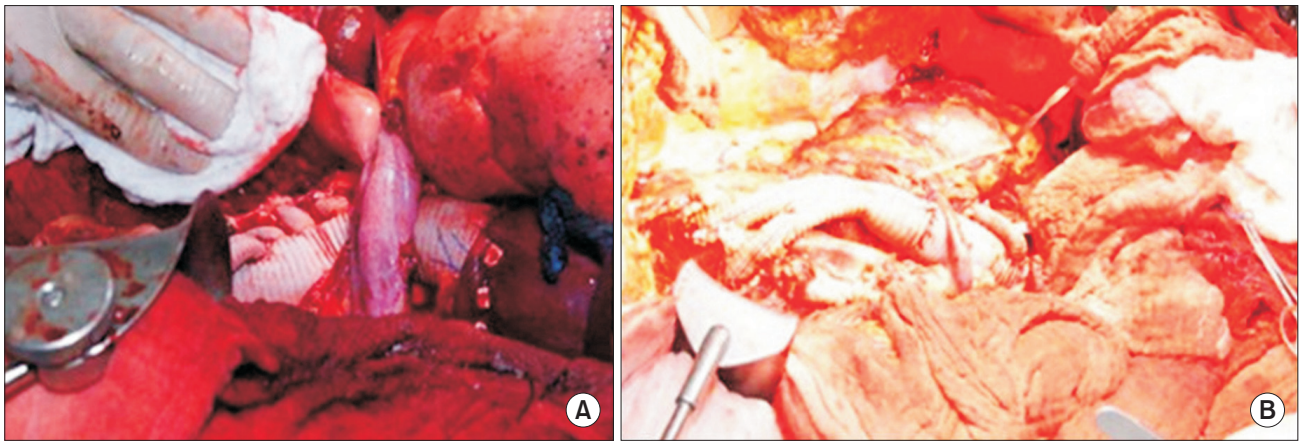

Fig. 5. (A) The thoracic aorta endograft was clamped just above the diaphragm and the aorta and vessels reconstruction was performed using a 22 mm five-branched Dacron graft. (B) Finally, a bifurcated 20×10 mm graft was anastomosed to the distal tube graft and its limbs distally to the common iliac arteries bilaterally.

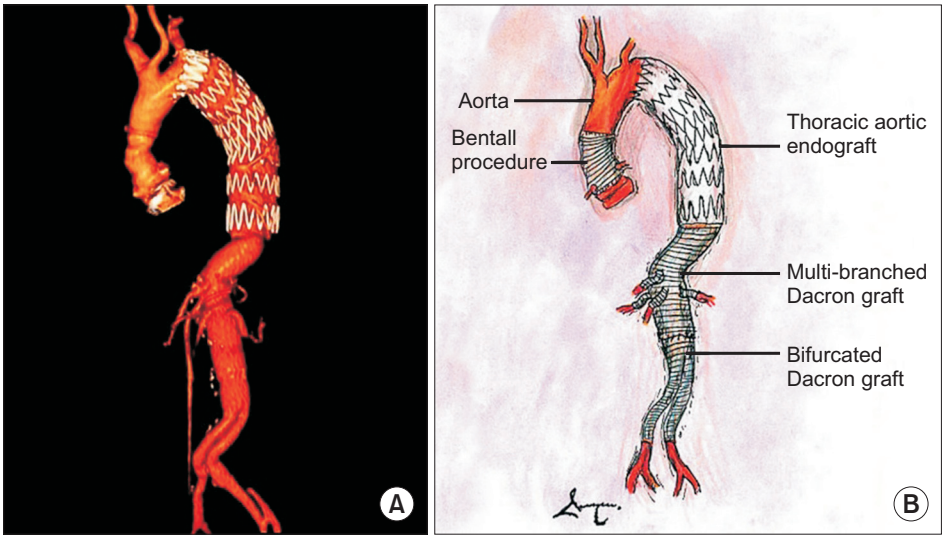

Fig. 6. (A) At 5 years from her last surgery, a computed tomography angiography three dimensional reconstruction revealed no new dissection in the remaining aorta or aneurysmal degenerations. (B) Schematic drawing of the sequential hybrid aortic and bilateral iliac aneurysm repair showing the components of the prosthetic material utilized for the complex reconstruction.

these extensive aortic aneurysms have only 10% to 20% 5-year survival rates, mostly secondary to rupture [5].

As these patients who have been previously treated with ascending aorta reconstructions, the option to perform a safe secondary or tertiary open intervention diminishes importantly, but the advent of endovascular technology has provided an attractive option for patients who are high risk for open operation. The select use of endovascular therapy in MS has been described in series [7]; however, the short and long-term benefits are unproven, and this practice remains controversial in patients with connective tissue disease as there is limited information regarding the impact of persistent radial forces of a stent-graft in an abnormal and weak aorta [8]. It is known that distal aortic failure after thoracic endovascular aortic repair (TEVAR) may occur as the distal aorta continues to grow; in a study by Geisbüsch et al. [7], MS patients who underwent TEVAR had 38% of reintervention rate and 50% developed de novo aneurysms during the follow-up. In another study by Norton et al. [1], these patients had a 33% reintervention rate and all distal thoracic aneurysm continue to grow at an average rate of 7 mm/year. Other authors as Waterman et al. [3], in their published series that included 16 patients that underwent TEVAR, there was a 12% of perioperative mortality and 44% required conversion to an open surgery due to diverse causes including retrograde dissections, type I endoleak, aortic rupture. Endovascular therapy should be seen with caution.

Despite the high rate of distal failures after TEVAR, some authors have proposed the use of stent-grafts as an important element to complete an aortic hybrid reconstruction, reducing the operative time significantly. Results have sug-

https://doi.org/10.5758/vsi.2017.33.3.112

gested that staged hybrid repair can be safely performed with acceptable results and at mid-term with favorable aortic remodeling and freedom from renal insufficiency, paraplegia or reinterventions [9]. We opted for this surgical approach for our patient as the result of a favorable remodeling of the stent-graft portion and this should be always in consideration; ultimately this decision reduced significantly the operative time of the complex surgical reconstruction.

During the reconstruction of the visceral aortic segment, the standard TAAA involves the inclusion technique, characterized by direct reimplantation of visceral and intercostal vessels in the aortic graft in the form of a Carrel patch, however the use of this technique in MS patients leads to the development of visceral patch aneurysms with a reported 18% of incidence in compare to the 5.5% with patients with aneurysms associated to atherosclerosis [10]. The use of multi-branched grafts with direct end to end anastomosis to the visceral vessels leads to low morbidity and mortality with excellent vessel patency and preservation of kidney function. The formation of pseudoaneurysms in the anastomotic sites following the aortic reconstruction in patients with connective tissue disorders or vasculitis is a severe complication, and it has to be prevented by reinforcing sutures with felt pledgets as we have previously described [11,12]. In this particular case, the anastomosis with the thoracic stent graft allowed to minimize to surgical time to complete the aortic repair with the five branched tube graft safely and without complication. Additionally the aneurysmal involvement of the bilateral common iliac arteries added more complexity to our reconstruction, extending our surgical repair using a bifurcated 20×10 mm graft to each of the pelvic vessels, which is not commonly performed when treating a complex thoracoabominal aortic aneurysm.

In conclusion, we report the successful sequential hybrid aortic and bilateral iliac artery aneurysm repair in a young patient with MS with 5-year follow-up. Aortic disease in Marfan patients is a complex clinical problem that may lead to secondary or tertiary aortic reconstructions; close follow-up is mandatory.

REFERENCES

1) Nordon IM, Hinchliffe RJ, Holt PJ, Morgan R, Jahangiri M, Loftus IM, et al. Endovascular management of chronic aortic dissection in patients with Marfan syndrome. J Vasc Surg 2009;50:987-991.
2) Judge DP, Dietz HC. Marfan's syndrome. Lancet 2005;366:1965-1976.
3) Waterman AL, Feezor RJ, Lee WA, Hess PJ, Beaver TM, Martin TD, et al. Endovascular treatment of acute and chronic aortic pathology in patients with Marfan syndrome. J Vasc Surg 2012;55:1234-1240; disucssion 1241.
4) Crawford ES, Coselli JS. Thoracoabdominal aneurysm surgery. Semin Thorac Cardiovasc Surg 1991;3:300-322.
5) Safi HJ, Miller CC 3rd. Spinal cord protection in descending thoracic and thoracoabdominal aortic repair. Ann Thorac Surg 1999;67:1937-1939; discussion 1953-1958.
6) Mimoun L, Detaint D, Hamroun D, Arnoult F, Delorme G, Gautier M, et al. Dissection in Marfan syndrome: the importance of the descending aorta. Eur Heart J 2011;32:443-449.
7) Geisbüsch P, Kotelis D, von Tengg-Kobligk H, Hyhlik-Dürr A, Allenberg JR, Böckler D. Thoracic aortic endografting in patients with connective tissue diseases. J Endovasc Ther 2008; 15:144-149.
8) Svensson LG, Kouchoukos NT, Miller DC, Bavaria JE, Coselli JS, Curi MA, et al; Society of Thoracic Surgeons Endovascular Surgery Task Force. Expert consensus document on the treatment of descending thoracic aortic disease using endovascular stent-grafts. Ann Thorac Surg 2008;85(1 Suppl):S1-S41.
9) Jain A, Flohr TF, Johnston WF, Tracci MC, Cherry KJ, Upchurch GR Jr, et al. Staged hybrid repair of extensive thoracoabdominal aortic aneurysms secondary to chronic aortic dissection. J Vasc Surg 2016;63:62-69.
10) Dardik A, Perler BA, Roseborough GS, Williams GM. Aneurysmal expansion of the visceral patch after thoracoabdominal aortic replacement: an argument for limiting patch size? J Vasc Surg 2001;34:405-409; discussion 410.
11) Hinojosa CA, Anaya-Ayala JE, Torres-Machorro A, Lizola R, Laparra-Escareno H. Middle aortic syndrome in Takayasu's arteritis: report of two surgical cases. Ann Vasc Surg 2016;34: 270.e13-e17.
12) Hinojosa CA, Anaya-Ayala JE, Laparra-Escareno H, Torres-Machorro A, Lizola R. Complex aortic and bilateral renal artery aneurysm repair in a young patient with multiple arterial aneurysm syndrome. J Vasc Surg Cases Innov Tech 2016;2:84-87.