Hybrid aortobrachial bypass for a giant subclavian and axillary artery aneurysm in a Marfan patient

Pedro J. Furtado Neves, MD, LeslieAnn S. Kao, MD, Andrea Simioni, MD, Emily A. Malgor, MD, Donald L. Jacobs, MD, *and* Rafael Demarchi Malgor, MD, *Aurora, CO*

Marfan syndrome was first described in 1896 and is now known to be an autosomal dominant disorder present in 1 of every 5000 live births. It is a mutation of the gene that encodes the fibrillin structural macromolecule (most often the *FBN1* gene).[1] This leads to a weakening of the media of the blood vessel wall, which manifests most frequently as aortic disease. However, a relationship between Marfan syndrome and peripheral aneurysms has also been shown, although axillary and subclavian artery aneurysms were rare, representing only 4.25% and 8.88% of all extrathoracic aneurysms according to a recent systematic review.[2] The average diameter ranged from 38.88 to 38.2 mm.[2] Because of the rarity of this clinical entity and the frequent previous operations in these patients, these aneurysms can be challenging to treat. We present the case of a giant 5.8-cm left subclavian and axillary artery aneurysm in a patient with Marfan syndrome with multiple prior aortic interventions that was successfully treated with a hybrid aortobrachial bypass. The patient provided written informed consent for the report of her case details and imaging studies.

We describe the case of a 57-year-old female patient with Marfan syndrome who presented with a complaint of severe left upper extremity burning pain, numbness, and frequent nocturnal wakening of several weeks' duration and a growing pulsating mass in the axilla. The patient had a history of ascending aortic replacement in 1995, a Yacoub valve-sparing procedure and coronary artery bypass grafting with a left internal mammary artery (LIMA) bypass to the left anterior descending artery in 2004, surgical aortic valve replacement with a mechanical valve in 2008, and open repair of abdominal aortic dissection in 2013. Computed tomography angiography showed a large, 5.8-cm left subclavian and axillary artery aneurysm and a patent LIMA graft (Fig 1). Because of the multiple prior thoracotomies, the patient was deemed at high risk of another redo sternotomy, and a hybrid approach was chosen in discussion with the cardiac surgery team and the patient. This would involve combining endovascular exclusion of the aneurysm, which has previously been described,[3] with open aneurysmal sac resection. Thus, the procedure involved cutdown of the distal brachial artery. Retrograde access was obtained into the ascending aortic arch, and stenting was performed, starting from the left subclavian artery. To maintain LIMA and left vertebral artery patency, an open-cell bare metal stent was deployed across the LIMA and left vertebral artery takeoff. Distally, auto-expandable covered stents were deployed into the axilla. Next, cutdown of the aneurysm in the axilla and of the distal subclavian artery to obtain proximal and distal control was performed, with endovascular balloon control. After both had been controlled with vessel loops, the aneurysm was dissected free of the surrounding nerves and brachial vein, with the vessels arising from the aneurysm double ligated and divided. The balloon was taken down, a Fogarty clamp with Hydragrip inserts was placed proximally to avoid damage to the stents. Distally, a regular vascular clamp was placed. The median and ulnar nerves were found to be intimately associated with the aneurysm, explaining the symptoms of paresthesia due to nerve compression (Fig 2). The aneurysm sac was opened, the aneurysm wall was resected, and a proximal anastomosis between the 9 × 50-mm auto-expandable Viabahn stent (W.L. Gore & Associates) and 7 × 4-mm tapered polytetrafluoroethylene (PTFE) graft was performed. Distally, the PTFE graft was cut to size and anastomosed to the distal brachial artery, measuring ~4 mm. The opened aneurysm sac with the stent graft and both proximal and distal PTFE graft anastomoses are shown in Fig 3. For the distal brachial artery and PTFE anastomosis, pledget-supported stitches were required because of arterial wall fragility. Finally, soft tissue rearrangement was performed as deemed most appropriate intraoperatively with subcutaneous tissue and muscle mobilization. This was necessary owing to the large amount of dead space resulting from resection of such a large aneurysm sac and thrombus. Layered closure was performed. The patient had an unremarkable postoperative course and was discharged home on postoperative day 15 with

From the Division of Vascular Surgery and Endovascular Therapy, University of Colorado, Anschutz Medical Center.

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Correspondence: Rafael Demarchi Malgor, MD, Division of Vascular Surgery, Department of Surgery, University of Colorado, Anschutz Medical Center, 12631 E 17th Ave, Rm 6111, Aurora, 80045 CO (e-mail: rafael.malgor@cuanschutz.edu).

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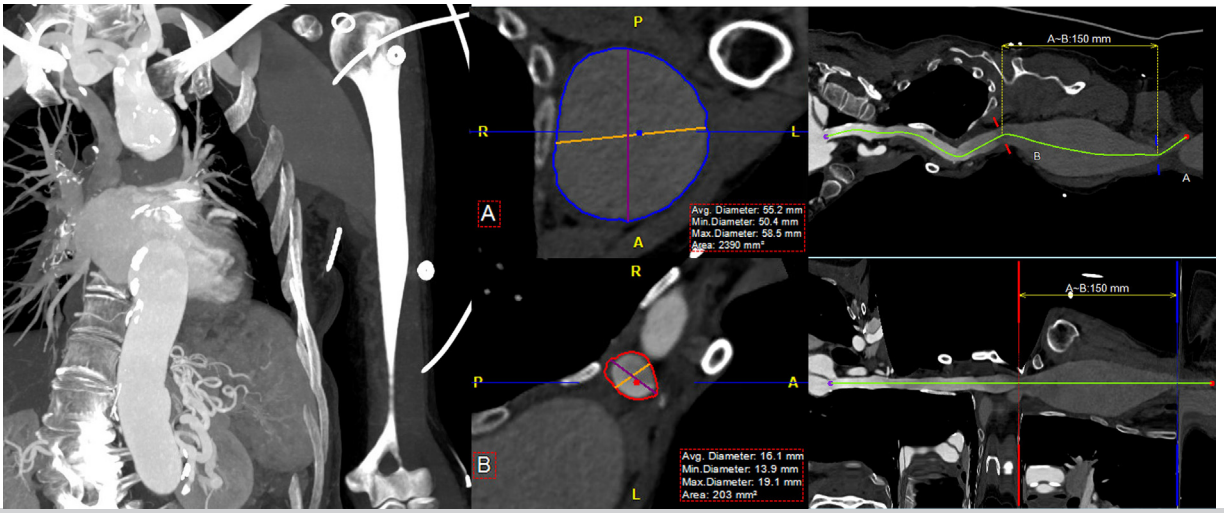

Fig 1. Computed tomography angiogram showing a large, 5.8-cm left subclavian and axillary artery aneurysm and patent left internal mammary artery (LIMA) graft.

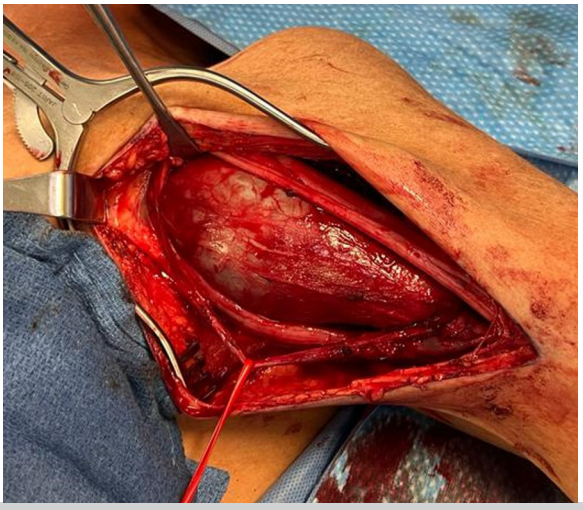

Fig 2. Intraoperative photograph showing median and ulnar nerves intimately associated with the aneurysm, explaining the symptoms of paresthesia due to nerve compression.

Fig 3. Intraoperative photograph showing opened aneurysm sac with the stent graft and both proximal and distal polytetrafluoroethylene (PTFE) graft anastomoses.

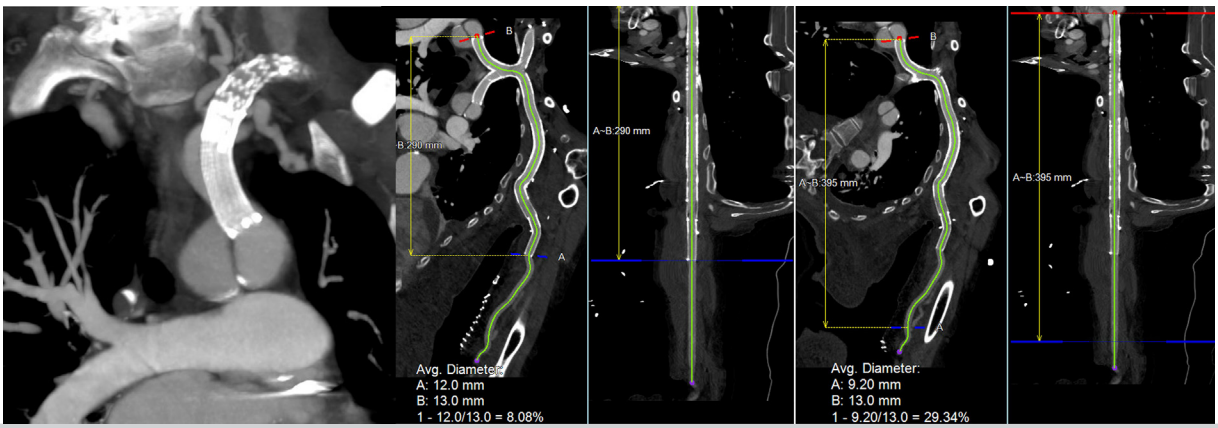

Fig 4. Follow-up computed tomography angiograms showing full extent of stenting over 29 cm and bypass over 39.5 cm, with a patent left internal mammary artery (LIMA) and left vertebral artery.

prescriptions for aspirin 81 mg daily, clopidogrel 75 mg daily, and warfarin with a goal international normalized ratio of 2.5 to 3.5 because of her mechanical aortic valve. At the 1-month follow-up, the patient's burning pain and numbness had resolved fully, and she had intact motor function and sensation of the hand and forearm. The incision healed fully without complications. Follow-up computed tomography angiography showed the full extent of stenting over 29 cm and bypass over 39.5 cm, with a patent LIMA and left vertebral artery (Fig 4). At the latest follow-up visit at 4 months after treatment, the patient was fully asymptomatic.

In conclusion, patients with Marfan syndrome often have complex disease and require lifelong surveillance. Because these patients often require multiple aortic and vascular procedures, those with multiple prior thoracotomies and a high risk of surgery could benefit from hybrid procedures to avoid additional redo sternotomy. These techniques can lead to lower rates of morbidity, a quicker return to activities, and lower mortality.

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