Case Reports

Repair of Multiple Subclavian and Axillary Artery Aneurysms

in a 58-Year-Old Man with Marfan Syndrome

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Dilation of the ascending aorta and aortic dissections are often seen in Marfan syndrome; however, true aneurysms of the subclavian and axillary arteries rarely seem to develop in patients who have this disease. We present the case of a 58-year-old man with Marfan syndrome who had undergone a Bentall procedure and thoracoabdominal aortic repair for an aortic dissection and who later developed multiple aneurysmal dilations of his right subclavian and axillary arteries. The aneurysms were successfully repaired by means of a surgical bypass technique in which a Dacron graft was placed between the carotid and brachial arteries. We also discuss our strategy for determining the optimal surgical approach in these patients. ***(Tex Heart Inst J 2016;43(5):428-9)***

In patients who have Marfan syndrome, aneurysmal enlargement and dissection of the ascending aorta are frequent cardiovascular sequelae; in contrast, peripheral artery aneurysms seem to develop only rarely. Aneurysmal dilation of the carotid, iliac, hepatic, and middle colic arteries sometimes occurs in patients who have Marfan syndrome, but aneurysms of the subclavian and axillary arteries are highly unusual.[1-3] We report a case in which a patient with Marfan syndrome developed multiple aneurysms of the subclavian and axillary arteries after aortic dissection, and we discuss our choice of surgical repair technique.

Key words: *Aneurysm/complications/surgery; diagnostic imaging/methods; Marfan syndrome/complications; peripheral arterial disease/complications; risk factors; treatment outcome*

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Case Report

In May 2015, a 58-year-old man with Marfan syndrome was admitted because of a pulsating mass on his neck. He had previously had a type I aortic dissection and had undergone an emergency Bentall procedure and elective thoracoabdominal aortic repair (19 and 11 years, respectively, before the current admission). The right axillary artery had been used for cannulation in the Bentall procedure.

On physical examination, a small, pulsating mass was observed in the patient's right lower neck; the radial pulses were palpable bilaterally. Systemic blood pressure measurements were equal in the right and left arms. Contrast-enhanced and 3-dimensional computed tomograms (CTs) revealed multiple dilations of the right subclavian and axillary arteries (Figs. 1 and 2); 3 aneurysms were identified. One aneurysm was in the subclavian artery, and the others were in the proximal and distal thirds of the axillary artery, respectively. Of note, the middle third of the axillary artery, the site of the previous cannulation, had no aneurysm. The brachial artery showed no stenosis or dilation.

We scheduled the patient for corrective surgery. A transverse incision was made above the right clavicle, and the branches of the brachiocephalic artery were identified. Another incision was made in the axilla, and the brachial artery, which was immediately distal to the axillary artery, was exposed. After heparin was administered, a 10-mm Dacron graft was anastomosed between the right common carotid artery and the proximal brachial artery. The subclavian artery was ligated proximally, and the axillary artery was ligated immediately distal to the aneurysm. After the procedure, distal pulses were present in the patient's right arm. His postoperative course was uneventful.

Dr. Coselli is principal investigator for clinical trials conducted by Medtronic, Inc.; W.L. Gore & Associates; and Cook Medical, Inc.; spoke at a Medtronic-sponsored meeting; serves on a W.L. Gore advisory board; and consults for and receives royalties from Vascutek Ltd., a Terumo company. Dr. Preventza consults for Medtronic and W.L. Gore and had travel paid by Cook Medical.

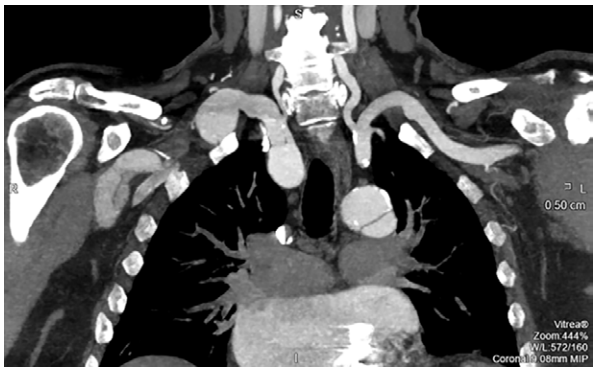

Fig. 1 *Contrast-enhanced computed tomogram of the chest shows a 4-cm aneurysm of the proximal right subclavian artery, a 4-cm aneurysm of the proximal right axillary artery, and a 2.5-cm aneurysm of the distal right axillary artery.*

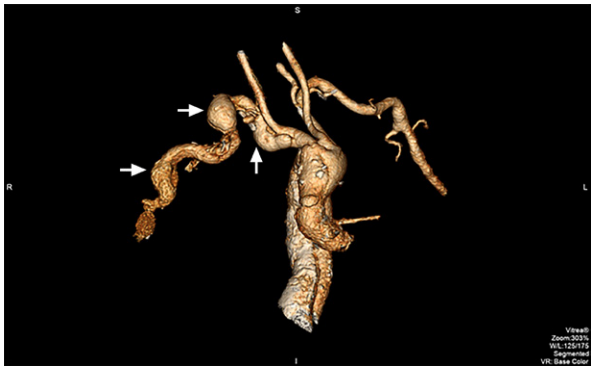

Fig. 2 *Three-dimensional computed tomogram shows consecutive aneurysmal enlargements (arrows) of the right subclavian and axillary arteries.*

Discussion

Aortic dissection and aneurysm formation in the ascending aorta are well-recognized sequelae of Marfan syndrome; conversely, peripheral artery aneurysms are less frequently reported, and subclavian and axillary artery aneurysms are extremely unusual. Peripheral artery aneurysms are thought to increase the morbidity and mortality rates of patients with Marfan syndrome because of potential rupture, thrombosis, and embolization.[4] Accordingly, we think that all symptomatic subclavian and axillary aneurysms in these patients should be treated. Although a strong correlation has not been found between aneurysm size and the occurrence of sequelae, repair has been recommended for all subclavian and axillary aneurysms greater than 2 cm in diameter.

Long-term monitoring of patients who present with Marfan syndrome is important. Some investigators have shown that the incidence of reoperation for aneurysmal disease or dissection is significantly increased in such patients.[5] Therefore, regular evaluation of the aorta and peripheral arteries with use of CT or magnetic resonance angiography (MRA) is recommended in these patients. We suggest that examinations be performed in the first 6 months after treatment and then repeated yearly for 2 to 3 years—with use of the same imaging method for each examination, to enable proper comparison until the rate of growth of the remaining aorta and its branches is established. If a slow growth rate is observed (<1 cm/yr), the imaging interval may be lengthened to every 2 years. Alternating between CT and MRA in the follow-up examinations might be advisable, to decrease the patient's total amount of radiation exposure during the lifelong monitoring period.

Aneurysms of the subclavian and axillary arteries typically can be repaired by either surgical or endovascular techniques; however, endovascular treatment of these vessels is not encouraged in Marfan patients, because the inherent weakness of arterial walls in connective-tissue disease results in poor stability of stents. Of multiple surgical methods to repair aneurysms of the subclavian and axillary arteries, resection and end-to-end anastomosis has been used most often. However, if the aneurysm involves a long vascular segment, or if multiple consecutive aneurysms are present, we think that it is better to perform bypass grafting with ligation of the aneurysmal section. A supraclavicular incision can be used for proximal control, and an infraclavicular or axillary incision for distal control. If more proximal control is required, median sternotomy is an option. The carotid artery, brachiocephalic artery, or proximal subclavian artery can be used for a proximal anastomosis, depending on the aneurysm's location.

Our patient had multiple aneurysms of the subclavian and axillary arteries, so we decided to perform bypass grafting. We used the right common carotid artery for proximal anastomosis because of the aneurysmal dilation of the proximal subclavian artery and the patient's previous median sternotomy (this last, associated with additional surgical risk). We think that close monitoring, including regular evaluation of the peripheral arteries, is mandatory in Marfan syndrome patients.

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