Combined surgery for the treatment of bilateral subclavian artery aneurysm in Marfan syndrome

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Large bilateral aneurysm of the subclavian artery is an infrequent entity that can progress to thrombosis, embolization, or rupture if left untreated. Treatment consists of exclusion of the aneurysm by an endovascular procedure or open surgery. We present a case of large bilateral subclavian artery aneurysm in a patient with Marfan syndrome that was treated by a combination of endovascular and conventional surgery. This therapeutic approach provided good results for patency with lower morbidity and mortality. (J Vasc Surg 2007;45:180-2.)

Aneurysms of the subclavian artery account for <1% of all peripheral aneurysms.1 The most frequent cause is compression due to a narrow thoracic outlet. Other causes are arteriosclerosis and traumatic injury.2,3 Associated connective tissue disease is rare. Conventional therapy involves exclusion of the aneurysm. In patients with connective tissue diseases, however, this treatment may present complications owing to deterioration of the artery and prior procedures in the aortic region, which are common in these patients.4 We present a patient with a large bilateral aneurysm of the subclavian artery and Marfan syndrome who was treated by a combination of endovascular and conventional surgery.5

CASE REPORT

A 38-year-old woman had a history of chronic anemia due to malabsorption, bilateral internal saphenectomy for varices, and celiac disease under dietary treatment. In 2001, the patient presented with sudden chest pain radiating to the abdomen, which was diagnosed by computed tomographic (CT) angiography as a type B acute aortic dissection, extending from below the origin of the left subclavian artery to the right common iliac artery.

Visceral ischemia was detected, and an aortoaoortic prosthetic graft was implanted from below the subclavian artery to the supraaortic trunks, with reimplantation of the intercostal arteries, celiac trunk, and superior mesenteric artery. Complications included a subdural hematoma that resolved spontaneously and a left empyema that required drainage.

A follow-up CT scan at 1 month disclosed a fluid collection around the prosthesis. The patient was afebrile, had no signs of systemic infection, and blood cultures were negative. Empirical oral antibiotic treatment with ciprofloxacin 500 mg/12 hours for 4 weeks was prescribed.

In 2004, CT angiography evidenced two bilateral subclavian aneurysms, (maximum diameters: right, 5.5 cm; left, 3.5 cm) with no thrombi. The right aneurysm had no proximal neck and arose at the bifurcation of the brachiocephalic trunk. The left aneurysm presented a proximal neck. The patient was asymptomatic clinically. Painless supraclavicular masses with distal pulses could be palpated. There were no signs of ischemia or upper limb embolization, and no evidence of focal neurologic involvement (Fig 1). An ultrasound study and arteriography of the supraaortic trunks confirmed the CT angiography findings, demonstrated the origin of both vertebral arteries in the aneurysmal sacs, and disclosed a right axillary artery aneurysm (Fig 2).

Endovascular treatment was decided to exclude the left aneurysm. Eight days before the procedure, the left vertebral artery was revascularized to preserve vertebrobasilar circulation. The intervention consisted of vertebral artery reimplantation in the common carotid artery with a side-to-end anastomosis and proximal ligation of the vertebral artery.

After reimplantation of the vertebral artery, exclusion of the left subclavian artery was performed. By percutaneous access through the right common femoral artery and under arteriographic guidance through the left humeral artery, two overlapping Wallgraft endoprostheses (12 cm × 7 mm and 14 cm × 7 mm; Boston Scientific, Natick, Mass) were implanted in the left subclavian artery.

Perioperative arteriography showed exclusion of the aneurysm and no evidence of endoleaks. The patient presented a distal pulse, and there were no signs of ischemia or embolization. Chest plain radiographs showed no dislocation of the endoprosthesis. Follow-up CT at 1 month demonstrated exclusion of the aneurysm without endoleaks and patency of the left vertebral artery reimplanted in the primitive carotid artery.

Two months later, exclusion of the right subclavian artery aneurysm was undertaken. A right laterocervical incision was performed, and the common right carotid artery was dissected. The right vertebral artery was dissected so it could be reimplanted in the common carotid, but it was found to be of poor quality. Hence, it was ligated and a sample was taken for histologic study.

The right femoral artery was accessed percutaneously for arteriographic monitoring. The common right carotid artery was punctured in a retrograde fashion and a 16-mm × 10-mm × 7-mm conical Excluder prosthesis (contralateral leg of abdominal bifurcated graft; W. L. Gore & Associates, Flagstaff, Ariz) was im-
planted from the brachiocephalic trunk to the common carotid, excluding the subclavian aneurysm proximally. We chose this graft because of the existing diameter difference between brachiocephalic trunk and right common carotid. Subsequently, a bypass with an 8-mm polytetrafluoroethylene graft (W. L. Gore & Associates) was performed from the right common carotid artery to the right axillary artery distal to the axillary aneurysm, with proximal ligation of the artery.

Thus, the two aneurysms were excluded and preserved the arm. Perioperative arteriography confirmed exclusion of both aneurysms without leaks and patency of the carotid-axillary bypass.

No dislocation of the grafts was observed on the chest radiographs. The patient presented a distal pulse, and there were no signs of ischemia or embolization. There were no femoral complications in these procedures. The patient received clopidogrel as antiplatelet therapy.

A genetic study of the right vertebral artery showed fibrillin 1 gene mutation. The diagnosis of Marfan syndrome was made with genetic and clinical criteria, including ascending aortic dissection, spinal scoliosis, and pes planus.

Follow-up CT and arteriography at 1 month showed exclusion of both subclavian aneurysms and the right axillary aneurysm, without endoleaks, and patency of the reimplanted left vertebral artery and right carotid axillary bypass (Fig 3). The angiography was performed for iliac aneurysm study. Additional follow-up with CT and duplex imaging after 6 and 12 months showed patency of the grafts and exclusion of both aneurysms without complications.

DISCUSSION

Subclavian artery aneurysms are uncommon. The singular features of this case include the bilateral involvement, large size, implication of both vertebral arteries and the right axillary artery, the etiology, and resolution by procedures combining endovascular treatment and conventional surgery.

The literature on subclavian artery aneurysm contains only small series or sporadic cases; hence, the natural history of this condition is uncertain. The most common causes are thoracic outlet syndrome, traumatic injury, and arteriosclerosis. Connective tissue disorders such as Marfan syndrome are among the less frequent etiologies, representing <10% of all subclavian aneurysms.

A subclavian aneurysm can be asymptomatic, presenting as a pulsatile supraclavicular mass, or become complicated and produce distal embolizations, thrombosis with ischemia of the limb, compression of adjacent structures, or rupture. In our case, the patient was asymptomatic, and the aneurysms were found on a follow-up examination for a prior surgery.
Classic treatment consists of resection of the aneurysm and insertion of a prosthetic graft or direct end-to-end anastomosis.\textsuperscript{9,10} The associated morbidity and mortality of this surgery is not negligible in patients with Marfan syndrome, although the rates are difficult to establish because the condition is uncommon and the series are limited. The introduction of endovascular surgery in this field has opened the possibility for less aggressive treatment, with lower morbidity and mortality, and good outcome with respect to patency.\textsuperscript{11,12} The first author to describe exclusion of a subclavian aneurysm by endovascular treatment was May et al.\textsuperscript{13} This therapy has been applied to aneurysms of various etiologies, with the use of several access routes.\textsuperscript{14-17}

Several problems had to be taken into account when the treatment strategy to use in this case was planned. The underlying disease that caused the prior aortic dissection and the two aneurysms was unknown, previous chest surgery made a new access difficult, CT showed a periprosthetic fluid collection, the origin of both vertebral arteries and the two aneurysms was unknown, previous chest surgery was not negligible in patients with Marfan syndrome.\textsuperscript{18} True aneurysm of axillary-subclavian artery with cystic medionecrosis: an unusual manifestation of Marfan syndrome. Ann Vasc Surg 2003;17:562-4.

One study in the related literature proposed treatment of a subclavian artery aneurysm by an innominate–carotid artery stent graft and a carotid-subclavian bypass and vertebral artery bypass.\textsuperscript{8} To our knowledge, combined treatment for a giant bilateral subclavian artery aneurysm in Marfan syndrome has not been reported.

CONCLUSION

Endovascular techniques combined with conventional revascularization offer a promising approach for use in patients with subclavian artery aneurysmal degeneration due to connective tissue disease because of their lower associated morbidity and mortality. However, because of the connective disorder, dilatations in the anchor zones of the graft may develop that lead to type I endoleak, which will require careful future surveillance.

REFERENCES