Bypass and embolization for a vertebral artery aneurysm in a patient with Marfan syndrome

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Extracranial vertebral artery aneurysms represent an uncommon presentation of collagen vascular disease. We performed staged proximal embolization of large left vertebral artery aneurysm after distal common carotid-to-vertebral bypass at C2 in a young adult patient with Marfan syndrome and a hypoplastic contralateral vertebral artery. Dilation of the autogenous saphenous vein graft occurred at 1 year with proximal graft stenosis requiring operative revision. Subsequent dilation of the basilar artery led to symptoms of pontine compression at 18 months that have resolved at 31 months of follow-up. (J Vasc Surg Cases 2015;1:77-80.)

Marfan syndrome comprises a connective tissue disorder inherited in an autosomal-dominant pattern that occurs in approximately one per 100,000 people. The pathogenesis involves a molecular defect in the microfibrillar protein fibrillin, the major component of elastin-associated microfibrils commonly found in the arterial wall. The most common manifestations of this syndrome are musculoskeletal, cardiovascular, and ocular, with the cause of death most often the result of an aortic aneurysm rupture. Vertebral artery aneurysms present sporadically, and extracranial vertebral artery aneurysms occur even more rarely, with most instances associated with dissection resulting from severe hypertension or trauma. We recently cared for a patient with Marfan syndrome with an extracranial vertebral artery aneurysm. Patient consent to publish this report was obtained.

CASE REPORT

The patient is a 21-year-old man with Marfan syndrome confirmed by genetic analysis who underwent ligation of a patent ductus arteriosus, closure of secundum atrial septal defect, and valve-sparing aortic root repair for aneurysmal dilation of his ductus arteriosus, closure of secundum atrial septal defect, and valve-sparing aortic root repair for aneurysmal dilation of his ascending aorta 6 years prior. In the interval, he developed chest, neck, and back pain. On examination he had a pulsatile mass in the left supraclavicular area, but no cleft palate, bifid uvula, or hypertelorism suggestive of Loeys-Dietz syndrome. A computed tomography arteriogram (CTA) obtained during that admission demonstrated a large left vertebral artery aneurysm with erosion and widening of the foramina of the transverse processes of the cervical vertebrae (Fig 1).

He was referred to our facility for further evaluation. The physical examination was significant for pectus carinatum and a well-healed median sternotomy scar from the prior surgical intervention. A transthoracic echocardiogram showed mild tricuspid regurgitation and trivial aortic valve insufficiency as the only findings. A two-stage approach was planned, with direct revascularization of the distal vertebral artery, followed by proximal endovascular embolization of the vertebral aneurysm.

Procedure. The distal left common carotid artery was exposed via a left-sided neck incision. The prevertebral fascia was mobilized medially to expose the inferior cervical spinal ligament. Further dissection was performed from C3 through C5, at which time a Kerrison punch was used to remove the anterior portion of the foramen transversarium ring. The vertebral artery was then mobilized from the foramen transversarium. After systemic administration of unfractionated heparin (5000 IU), clamps were placed on the common carotid artery just proximal to the bifurcation, and a 10-cm-long portion of the great saphenous vein was reversed and anastomosed end-to-side to the artery via a longitudinal arteriotomy. An ipsilateral carotid-carotid shunt was used during the procedure to maintain cerebral blood flow. An end-to-side anastomosis was then performed from the vein graft to the vertebral artery, which was then ligated proximal to the anastomosis. After flow was confirmed with a Doppler signal, the heparin anticoagulation was reversed with intravenous protamine (25 mg), and the wound was irrigated and then closed.

The patient was taken to the interventional radiology suite 4 days later for coiling of the left vertebral aneurysm to reduce the cyclic stress on the vertebral aneurysm wall from the proximal arterial pulsation. Via femoral artery access, a 5F catheter was used to perform a four-vessel arteriogram that confirmed patency of the carotid-vertebral bypass (Fig 2, A).

The catheter and femoral sheath were removed over a wire and replaced by a 6F shuttle sheath advanced into the left vertebral artery, which was efficiently accomplished despite the tortuosity of the vessel. A selective left vertebral arteriogram was done in multiple projections to define the aneurysm’s anatomy. The distal aspect of the bilobed aneurysm was coiled with a 14-mm × 30-cm detachable microcoil. The catheter was withdrawn into the main
portion of the aneurysm and packed with multiple smaller microcoils within the aneurysm and feeding the neck of the aneurysm. A repeat left subclavian arteriogram revealed satisfactory obliteration of the aneurysm (Fig 2, B).

**Follow-up.** The patient initially did quite well, with no neurologic sequelae. He returned 12 months after the intervention with complaints of neck pain and headaches. A CTA demonstrated stenosis of the proximal anastomosis and dilation of the vein graft.

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**Fig 1.** A, A three-dimensional reconstruction of the preoperative computed tomography arteriogram (CTA) demonstrates the left vertebral artery aneurysm. B, CTA demonstrates foraminal erosion and widening, indicated by the arrow.

**Fig 2.** Arch arteriogram after carotid-vertebral bypass. A, Left common carotid injection shows the vein bypass (black arrow), which is much smaller. B, Left subclavian injection shows microcoil embolization (white arrow) of the left vertebral artery aneurysm.
from a 3.3-mm internal diameter to 7.2 mm (Fig 3, A). He then underwent revision of the left carotid-vertebral artery bypass. Under general anesthesia, the left common carotid artery and vein graft were exposed surgically. An incision was made from the common carotid through the area of stenosis and extending 5 mm onto the vein graft, followed by a limited intimectomy and Dacron (DuPont, Wilmington, Del) patch closure. His neck pain and headaches improved but did not totally resolve.

He returned 6 months after this encounter with dysarthria and a right posterior headache, and the physical examination revealed normal strength bilaterally with deviation of his tongue to the right. Magnetic resonance imaging revealed significant redundancy and dilation of the distal intracranial vertebral artery leading to a mass effect on the right aspect of the medulla near the root zone of the hypoglossal nerve, thought to be responsible for his symptoms. Neurosurgical consultation felt that intervention with decompression of the posterior fossa would likely only temporize and that it would not be possible to adequately protect the brainstem from the enlarging vertebral artery.

The dysarthria resolved ≤3 months, and at 30 months of follow-up, he continues to complain only of intermittent moderate headache. CTA showed further dilation of the vein graft to 8.8 mm, with dilation of the common carotid to 8.5 mm (Fig 3, B). In addition, he developed aneurysmal dilation of his redundant right internal carotid artery to 10.6 mm, without evidence of mural thrombus (Fig 3, C).

DISCUSSION

Extracranial vertebral artery aneurysms are quite rare. Most of the reported vertebral artery aneurysms tend to be intracranial, arising near the junction between the posterior-inferior cerebellar artery and vertebral artery. The first documented repair for a vertebral artery aneurysm was in 1893, and the first reconstruction was performed in 1957. Current techniques for repair include primary ligation of the inflow and outflow, vertebral revascularization distal to the aneurysm before the aneurysm is excluded, aneurysmorrhaphy, and endovascular balloon and coil.

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**Fig 3.** Three-dimensional computed tomography arteriogram (CTA) reconstruction demonstrates (A) proximal anastomotic stenosis and early vein graft dilation at 12 months after the operation, (B) vein graft dilation, and (C) a right internal carotid aneurysm at 30 months. The saphenous vein graft diameter equals or exceeds that of common carotid. The black arrow indicates the saphenous vein graft, the white arrow indicates the common carotid artery, and the gray arrow indicates the right internal carotid aneurysm.
embolization. A retrospective review by Morasch et al of seven patients with extracranial vertebral artery aneurysms noted that all patients had a history of connective tissue disorder and that most aneurysms were located in the V2 segment. Eight of the nine aneurysms were managed operatively with ligation, bypass, and aneurysmorrhaphy, with one patient undergoing coil embolization after distal bypass, similar to our patient. At a follow-up as long as 5.5 years, all reconstructions, except one, were patent.

Treatment of a wide-necked intracranial vertebral artery aneurysm with combined endovascular stent implantation and endosaccular coils has been described. Shang et al reported their management of a 26-year-old man with a vertebral artery aneurysm near the origin from the right subclavian artery. He tolerated unilateral balloon occlusion for 30 minutes and successfully underwent endovascular intervention with stent graft placement into the subclavian artery and embolization of the right internal mammary artery and thyrocervical trunk. For our patient, simple ligation of embolization was not considered suitable due to his hypoplastic and incomplete contralateral vertebral artery.

Use of autogenous saphenous vein in our patient with Marfan syndrome was associated with dilation of the venous conduit. Dilation of autogenous vein grafts occasionally can occur and has been described in <15% of coronary bypass vein grafts. To our knowledge, such dilation has not been reported previously in association with Marfan syndrome. The fibrillin-1 misfolding defect of Marfan syndrome primarily affects synthesis and maintenance of extracellular tissues such as elastin, which does not comprise a major component of the vein wall. Whether the observed vein graft dilation occurred independently from the Marfan syndrome is uncertain, but we remain concerned that further dilation may occur. Certainly, any subsequent continued dilation of the more distal intracranial vertebral artery may lead to significant morbidity.

CONCLUSIONS

This patient is currently being monitored annually with CTA. The dilated but nonaneurysmal vein graft would have to increase in size quite substantially to consider any intervention, which would likely be endovascular in nature, such as use of a flow-diverting stent. Unfortunately, it seems more likely that he will require some sort of intervention for his enlarging right internal carotid artery aneurysm or his dilating distal vertebral or basilar artery before that. With the currently available technologies, no specific size criteria for intervention have been determined for this patient, and we hope that the occurrence of significant symptoms will not prompt an early intervention, because the options are limited.

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