Endovascular treatment of ruptured axillary and large internal mammary artery aneurysms in a patient with Marfan syndrome

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Marfan syndrome is an autosomally inherited disorder affecting the synthesis of connective tissues. Vascular manifestations of Marfan syndrome include aneurysmal dilatation of the aortic root, aortic dissection, and rupture. Peripheral aneurysms are mostly reported in the iliac, femoral, and subclavian arteries. We report a Marfan patient with a ruptured axillary artery aneurysm and a large left internal mammary artery aneurysm. The axillary aneurysm was successfully excluded using covered stent grafts, and the left internal mammary artery aneurysm was effectively coiled. Duplex ultrasound imaging at 4 months and computed tomography at 9 months demonstrated complete thrombosis and exclusion of both aneurysms with patent subclavian-axillary stent grafts. (J Vasc Surg 2011;53:478-82.)

CASE REPORT

A 68-year-old man underwent multiple interventions for vascular manifestations of MFS. In 1990 he underwent open infrarenal abdominal aortic aneurysm repair with a tube graft. The ascending aorta and the aortic valve were replaced in 1997. The thoracoabdominal aorta from the left subclavian to the renal arteries was subsequently replaced in 2000. Owing to cardiomyopathy, a heart transplant was performed in 2002.

Aneurysms of the left axillary artery and left internal mammary artery (LIMA), dissection of the right subclavian artery, and a 5.1-cm pseudoaneurysm from the ascending aorta had been noted on two previous computed tomography (CT) scans; however, these were not routinely monitored.

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Completion angiogram demonstrated no flow into the LIMA aneurysm sac, complete exclusion of the axillary aneurysm, and a patent vertebral artery (Fig 3, B). At the conclusion of the procedure, a brachial artery cutdown was performed to allow primary repair of the artery. Manual compression was held on the femoral artery puncture site.

The patient recovered well. The radial pulse was palpable postoperatively. The mass on his left chest was no longer pulsatile. His pneumonia resolved with antibiotics and pulmonary toilet. Because of chronic renal insufficiency, N-acetylcysteine, hydration, and minimal contrast (60 mL) were used. A rise in his serum creatinine concentration returned to baseline within a week.

An arterial duplex scan of his left subclavian and axillary arteries 4 months postoperatively showed no endoleak, triphasic flow, and completely thrombosed aneurysm sacs. A noncontrast CT of the chest 9 months later showed resolution of the left axillary aneurysm and shrinkage of the LIMA aneurysm from 6 to 2.5 cm in maximum diameter (Fig 4). The durability of endovascular stent grafts in the peripheral arteries of MFS patients is unknown; therefore, this patient will be monitored closely with duplex and CT imaging surveillance.

DISCUSSION

MFS is an inherited defect in connective tissue synthesis, most commonly caused by an autosomal dominant mutation of the fibrillin-1 gene (FBN1). The Marfan phenotype is most significant for skeletal abnormalities: tall, thin, short torso, and arachnodactyly. Other manifestations include high propensity for lens dislocation, arched palate, skin with striae, and fascia prone to incisional and inguinal hernias. At the cellular level, these patients have elastic f
calcification, increased deposition of matrix elements, and intimal hyperplasia. These changes may lead to fragmentation and loss of tensile strength of the vessel fibers, allowing the vessel walls to stretch and form aneurysms.\(^1\) The media is subject to cystic medial necrosis and vacuolization, which leads to both aneurysmal disease and dissection.\(^2,3\)

Cardiovascular complications account for most of the morbidity and mortality in these patients, many of whom have mitral valve regurgitation or prolapse, or both, aortic root dilatation, ascending aortic aneurysms (with high risk for dissection and rupture), and commonly, abdominal aortic aneurysms.\(^7\) The incidence of peripheral aneurysms in MFS patients is not well described in the literature.\(^3\)

As previously mentioned, axillary and IMA aneurysms typically occur in a traumatic setting or in patients with connective tissue disorders. A limited number of case reports exist describing true axillary aneurysms, and the literature in MFS patients is especially sparse.\(^5\) In a comprehensive search, we identified no reported cases of simultaneous axillary and IMA aneurysms. The Table outlines the location, etiology, and treatment of reported axillary and IMA aneurysms in the current literature.\(^8-17\)

The decision to treat axillary and IMA aneurysms is based on size, presence of symptoms, and the risk for impending rupture.\(^4,5,18\) Axillary artery aneurysms can cause ischemia due to thromboembolism. Compression from large aneurysms, pseudoaneurysms, and hematomas can produce severe pain and brachial plexopathy. Spontaneous rupture can occur, as experienced by our patient.

Few published reports have described treating axillary pseudoaneurysms with percutaneous endovascular stent grafts; however, none were “true” aneurysms or patients with MFS.\(^4,18\) Much of the literature describes true axillary aneurysm resection with vein graft interposition.\(^19\) Endovascular techniques may be especially advantageous for MFS, because patients with various types of cystic medial necrosis have suboptimal outcomes with open repair. Self-expanding stents in the upper arm are likely a better option because they are less apt to deformation with movement and compression.\(^4\) The Viabahn stent graft is flexible, making it more adaptable to tortuous anatomy.\(^20,21\) Excluding
true axillary aneurysms with a covered stent seems prudent; however, some pseudoaneurysms, especially saccular ones, can be treated by placing coils in the aneurysm cavity, and an uncovered stent can be placed to allow maintenance of the side branches after repair.19

The literature primarily describes treatment options for IMA pseudoaneurysms after median sternotomy, but such options can be applied to true IMA aneurysms as well. Open repair through a limited thoracotomy allows excision or ligation of the aneurysm. Embolization results in prompt thrombosis and an earlier patient discharge.22 We identified a single report of a LIMA aneurysm in a Marfan patient that was successfully treated with coil embolization.13 If the patency of the LIMA needs to be preserved, coronary stents have been placed in the LIMA to exclude the aneurysm sac.22

CONCLUSIONS

The genetic defect in connective tissue synthesis in patients with MFS may cause aneurysmal degeneration not

<p>| Table. Description of reported axillary and internal mammary artery aneurysms, treatment modality, and outcomes |
|---------------------------------|---------------------------------|---------------------------------|---------------------------------|---------------------------------|</p>
<table>
<thead>
<tr>
<th><strong>Aneurysm location</strong></th>
<th><strong>Etiology</strong></th>
<th><strong>Presentation</strong></th>
<th><strong>Treatment</strong></th>
<th><strong>Outcomes</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>L axillary</td>
<td>No risk factors, exercise, mycotic</td>
<td>L arm pain, exercise, thromboembolism of pitching hand</td>
<td>Self expanding wall graft stent</td>
<td>Uneventful recovery</td>
</tr>
<tr>
<td>R axillary aneurysm</td>
<td>Major league pitcher</td>
<td>Numbness and coolness of pitching hand</td>
<td>Open resection with reverse saphenous vein graft</td>
<td>Uneventful recovery</td>
</tr>
<tr>
<td>R axillary aneurysm</td>
<td>Baseball pitcher</td>
<td>Numbness, cyanosis, and coolness of R hand with signs of small vessel embolization in digits 2-4</td>
<td>Open exploration, aneurysm ligation, ligation of anterior and posterior circumflex humeral arteries</td>
<td>Uneventful recovery</td>
</tr>
<tr>
<td>R axillary aneurysm</td>
<td>Baseball pitcher</td>
<td>Numbness, cyanosis of the right 2nd finger</td>
<td>Open exploration, aneurysm ligation and repair with reverse saphenous vein graft, ligation of anterior and posterior circumflex humeral arteries</td>
<td>Uneventful recovery</td>
</tr>
<tr>
<td>R axillary aneurysm</td>
<td>Alport’s syndrome, previous fistula, IV drug abuse, mycotic aneurysm</td>
<td>Embolization to fingers</td>
<td>Open resection with reverse saphenous vein graft, ligation of feeding vessels</td>
<td>Uneventful recovery</td>
</tr>
<tr>
<td>L axillary aneurysm</td>
<td>IV drug abuse, mycotic aneurysm</td>
<td>Finger tip ischemia</td>
<td>Open resection with reverse saphenous vein graft</td>
<td>Uneventful recovery</td>
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<tr>
<td>L axillary aneurysm</td>
<td>Long-term crutch use</td>
<td>Pain in L hand and absence of pulses</td>
<td>Open resection with end-to-end anastomosis</td>
<td>Uneventful recovery</td>
</tr>
<tr>
<td>L axillary aneurysm</td>
<td>Long-term crutch use</td>
<td>Coolness and pain of L hand</td>
<td>Open resection with reverse saphenous vein graft</td>
<td>Uneventful recovery</td>
</tr>
<tr>
<td>R axillary aneurysm</td>
<td>Long-term crutch use</td>
<td>Pain, numbness, and tingling of R hand</td>
<td>Open exploration with aneurysmorrhaphy</td>
<td>Uneventful recovery</td>
</tr>
<tr>
<td>R axillary aneurysm</td>
<td>Long-term crutch use</td>
<td>Pain in R hand, wrist with absence of sensation</td>
<td>Open resection with reverse saphenous vein graft</td>
<td>Uneventful recovery</td>
</tr>
<tr>
<td>L axillary aneurysm</td>
<td>Long-term crutch use</td>
<td>Pain, paralysis, coolness of L hand without pulses or sensation</td>
<td>Open resection with Dacron graft</td>
<td>Uneventful recovery</td>
</tr>
<tr>
<td>L IMA aneurysm</td>
<td>MFS, previous MVC</td>
<td>Incidental on routine CXR</td>
<td>Coil embolization</td>
<td>Uneventful recovery, died years later from type A dissection</td>
</tr>
<tr>
<td>Ruptured L IMA aneurysm</td>
<td>Ehlers-Danlos</td>
<td>Spontaneous hemothorax</td>
<td>Thoracotomy with ligation of the LIMA, drainage of hemothorax</td>
<td>Uneventful recovery</td>
</tr>
<tr>
<td>Ruptured L IMA aneurysm</td>
<td>No risk factors</td>
<td>Hemothorax</td>
<td>Angiographic embolization, thoracotomy for hemothorax evacuation</td>
<td>Uneventful recovery</td>
</tr>
<tr>
<td>BL IMA aneurysm</td>
<td>Polyarteritis nodosa</td>
<td>Hemothysis, incidental finding on imaging</td>
<td>Thoracotomy with bilateral aneurysmectomy</td>
<td>Uneventful recovery</td>
</tr>
<tr>
<td>I IMA aneurysm</td>
<td>Unknown</td>
<td>Incidental “coin lesion” on CXR</td>
<td>Exploratory thoracotomy and ligation of aneurysm</td>
<td>Uneventful recovery</td>
</tr>
<tr>
<td>R IMA aneurysm</td>
<td>Atherosclerosis</td>
<td>Anterior mediastinal mass</td>
<td>Open ligation and resection</td>
<td>Uneventful recovery</td>
</tr>
</tbody>
</table>

BL, Bilateral; CXR, chest x-ray; IMA, internal mammary artery; IV, intravenous; L, left; MFS, Marfan syndrome; MVC, motor vehicle collision; R, right.
only of the aorta but also of the peripheral arteries. Aneurysms of the axillary artery and IMA are rare but potentially morbidity. The presence of symptoms and an increase in the rate of growth are indications for treatment. Such aneurysms are amenable to endovascular therapy. The durability and patency of peripheral arterial stent grafts in patients with MFS is as yet unknown. Our short-term follow-up of this patient demonstrates an excellent result with symptom resolution significant sac shrinkage and complete exclusion of the aneurysm.

REFERENCES

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