Coil embolization of bilateral internal mammary artery aneurysms is durable in a patient with Marfan syndrome

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ABSTRACT
Internal mammary artery (IMA) aneurysms are very rare, have a high risk of rupture, and can cause hemothorax. Here, we report the case of a 33-year-old man with metachronal and bilateral IMA aneurysms. He had Marfan syndrome diagnosed by genetic testing. We carried out endovascular repair with coil embolization. He has survived without additional treatment for 7 years. Endovascular repair of metachronal and bilateral IMA aneurysms is feasible even in a patient with Marfan syndrome. (J Vasc Surg Cases and Innovative Techniques 2018;4:216-9.)

Keywords: Internal mammary artery aneurysms; Marfan syndrome; Endovascular repair

Internal mammary artery (IMA) aneurysms are very rare, with only 40 cases reported in the last 40 years. Although IMA aneurysms are small, they often rupture and cause hemothorax, which can be life-threatening. Although the cause of IMA aneurysms is iatrogenic or traumatic in most cases, it can be inflammatory vasculitis or connective tissue disease, including Loeys-Dietz syndrome, Marfan syndrome (MFS), Ehlers-Danlos syndrome, neurofibromatosis type 1, and fibromuscular dysplasia. Here, we describe an extremely rare case of metachronal, bilateral IMA aneurysms due to MFS successfully treated by endovascular therapy. The patient consented to participation and publication of this case report.

CASE REPORT
We report the case of a 33-year-old man with a history of medical and surgical interventions for aortic anomalies. In August 2002, he presented with symptoms of acute type B aortic dissection and was treated by anti-impulse therapy using beta blockers, angiotensin II receptor blockers, and an angiotensin-converting enzyme inhibitor. In April 2006, he underwent an open repair of the proximal descending thoracic aorta for the treatment of an aneurysmal dilation of the false lumen of the descending aorta. In August 2006, he underwent a second-stage thoracoabdominal aortic repair with a reconstruction of intercostal arteries.

In May 2010, a follow-up computed tomography (CT) scan revealed a new aneurysmal dilation in the left IMA (LIMA; Fig 1, A-C). Hence, coil embolization of the aneurysm was performed. Furthermore, puncture of the left brachial artery and selective catheterization of the LIMA were performed using a 4F catheter followed by a microcatheter. Angiography revealed the aneurysm at the midportion of the LIMA (Fig 2, A). The LIMA was occluded with 37 detachable coils (Tornado; Cook Medical, Bloomington, Ind) that were distally and proximally placed to the aneurysm, and the aneurysm was packed with 20 detachable coils. Completion arteriography demonstrated the absence of filling in the coiled aneurysmal sac.

In October 2012, a CT scan revealed two aneurysmal dilations in the right IMA (RIMA; Fig 1, D). In May 2012, coil embolization of the aneurysms was performed using a 0.016-inch guidewire and a 0.018-inch microcatheter. Retrograde cannulation of the RIMA was performed through the right brachial artery. Angiography revealed both aneurysms at the midportion of the RIMA (Fig 2, B). Coil packing of the aneurysms was performed with detachable coils. A total of 35 detachable coils were placed at the proximal and distal necks of the aneurysms. Completion arteriography demonstrated the absence of filling in the coiled aneurysmal sac. The follow-up images revealed successful embolization of the RIMA and subsequent complete thrombosis of both aneurysms (Fig 3). Five years after the final intervention, three-dimensional CT revealed no recurrence of the aneurysms (Fig 1, E).

Diffuse and rapidly progressing vascular disease was diagnosed in 2017 in our patient who met the criteria for MFS, namely, a height of 181 cm, family history, dural ectasia, clubfoot, and hypertelorism. Genetic testing also revealed a novel frameshift mutation of the FBN1 gene (Fig 4).

DISCUSSION
Only a few studies have reported true IMA aneurysms in the medical literature. The noniatrogenic or nontraumatic etiology, including vasculitis, connective tissue diseases, neurofibromatosis type 1, fibromuscular dysplasia, atherosclerosis, and idiopathic causes, is very rare. MFS is an autosomal dominant inherited connective tissue
disorder due to genetic mutation in the FBN1 receptor. The likely causes of mortality are aortic aneurysms, dissections, and ruptures. Moreover, MFS is associated with multiple aneurysms and arterial tortuosity throughout the body. In this case report, we describe an MFS patient with metachronal and bilateral IMA aneurysms.

An appropriate selection of interventions is essential because arterial tortuosity may preclude an endovascular intervention. Proper diagnosis is necessary to avoid selecting unsuitable interventions in patients with connective tissue diseases that may phenotypically mimic more common connective tissue diseases. Before surgical
intervention, we considered coil embolization as an alternative option for the aneurysm. Coil embolization is rapidly becoming the treatment of choice for arteriovenous fistulas and small aneurysms, although only seven studies have reported short-term outcomes after the treatment of IMA aneurysms. Our patient has survived without recurrence of the aneurysms for 7 years after the operation. To the best of our knowledge, this is the first report of long-term results of catheter intervention for an IMA aneurysm.

Our experience with this patient has suggested that endovascular repair can yield excellent results even in MFS patients. Follow-up CT studies at 6-month intervals have demonstrated stable embolization and complete proximal and distal aneurysm sac thrombosis for up to 7 years. Furthermore, the patient tolerated additional major surgical interventions in the interim. We could demonstrate that the endovascular repair of focal peripheral aneurysms is an acceptable alternative to open surgical repair in properly selected MFS patients. For such high-risk patients, a closer and stricter follow-up (ie, every 3-6 months) with medication or adequate surgical treatments at appropriate times is mandatory. Moreover, frequent and long-term postoperative surveillance imaging is necessary in this population of challenging patients.

Serial CT suggested a rapid growth of the aneurysm because it was not detected in previous radiographs. Because the paper-thin wall of an aneurysm could have caused it to rupture, it was thought to require treatment. There is a possibility that this was a case of a sporadic class of aneurysm potentially resulting from mutations that affect connective tissue, thereby weakening the structural integrity of the arterial wall. It appears that a careful follow-up is necessary to detect a possible aneurysm in other arteries, and the possibility of an increased risk of arterial disease in this patient’s offspring cannot be excluded.

Noniatrogenic IMA aneurysms are so rare that there is limited information about their management and prognosis. Thus far, there have been no established intervention criteria. If an IMA aneurysm is diagnosed, the intervention must be considered urgent regardless of the size. Rupture of IMA aneurysms may be fatal because hemothorax with shock is the most common initial manifestation of IMA aneurysms.

CONCLUSIONS

We describe an extremely rare case of metachronous, bilateral IMA aneurysms due to MFS diagnosed by genetic testing. The aneurysms were successfully treated by endovascular repair with coil embolization. However, a careful follow-up by serial CT is necessary to detect possible aneurysms in the same or other arteries.
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