Endovascular Repair of Bilateral Internal Mammary Artery Aneurysms in a Patient with Marfan Syndrome

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Background: True internal mammary artery aneurysms are rare but rupture has been described.

Case Report: A 39-year-old male patient with Marfan syndrome was diagnosed with large asymptomatic bilateral internal mammary artery aneurysms (IMAAs) on contrast-enhanced CT scan, without other arterial lesions. Both aneurysms were coil embolized, a stent graft was deployed in the left subclavian artery to cover a wide neck that precluded complete and safe embolization of the left IMAA. Reintervention on the right side was performed 5 years later due to recanalization. Eight years after the initial procedure, the patient presented with a type A aortic dissection that was successfully repaired.

Conclusions: Although extremely rare, endovascular treatment should be considered to prevent rupture of internal mammary artery aneurysms.

Marfan syndrome (MFS) is one of the most common connective tissue genetic disorders affecting at least 1 in 20,000 people in the United States.[1] It has been established that FBN1 gene mutations are responsible for abnormal fibrillin found in most patients with MFS[2] and can cause a variety of cardinal manifestations, which involve the skeletal, ocular, cutaneous, and cardiovascular systems.[3]

Skeletal anomalies, including dural ectasia, can be found up to 90% of MFS patients and ocular manifestations in 50%.

Mitral valve prolapse, aneurysms, and dissection of the aorta are common in MFS patients and are known to have a high morbidity and are a frequent cause of death in this population. True aneurysms of carotid, subclavian, iliac, and visceral arteries are less frequently associated with MFS and only a few case reports are available. Aneurysms of smaller arteries are rare and exceptionally reported.[4]

We report a case of bilateral internal mammary artery (IMA) aneurysm in a patient with MFS that underwent successful endovascular treatment.

CASE REPORT

A 28-year-old male patient consulted with a significant family history of MFS with aortic complications. On a primary medical evaluation, he evidently had MFS stigmata, therefore chest, abdomen, and pelvis contrast-enhanced CT scan was obtained with no significant arterial findings.

Eleven years later, at the age of 39, he presented with an acute onset of lower back pain. CTA scan was ordered, which revealed tortuous bilateral internal mammary artery aneurysms with mural thrombus. Right internal mammary artery aneurysm was 28 mm large and located

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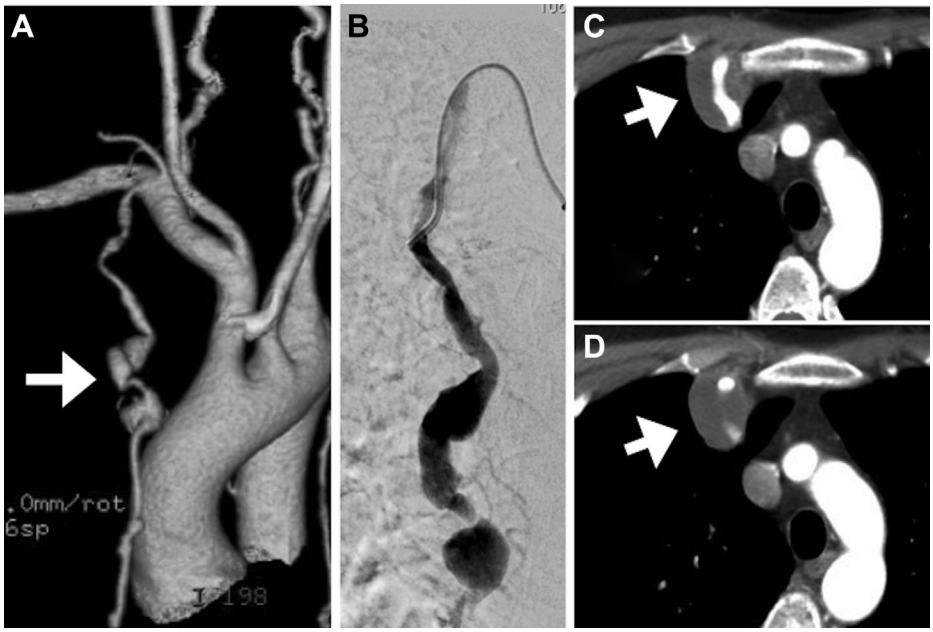

Fig. 1. Preoperative Computed Tomographic Angiogram. Volume rendering **(A)** shows right internal mammary artery aneurysm (*white arrow*) and its location. Selective angiogram during coil embolization **(B)**. Contrast phase **(C and D)** shows right IMA aneurysm with mural thrombus inside (*white arrows*).

in the mid third (Fig. 1) and left mammary artery aneurysms was located in the upper third with a diameter of 22 mm (Fig. 2). Lumbar scoliosis was the only finding related to his back pain.

A minimally invasive bilateral treatment was decided, a staged procedure was not considered due to the little morbidity related to bilateral IMA harvest for coronary bypass in nondiabetic patients. Percutaneous femoral access was obtained, innominate and right subclavian arteries were cannulated using a Vitek catheter (Cook Medical, Bloomington, IN). A triaxial system is composed of a 6 Fr. Ninety centimeter long sheath, a 5 Fr 45° angulated hydrophilic catheter, and a MicroFerret-18 microcatheter (Cook Medical, Bloomington, IN). This was advanced through the tortuous right IMA. Two 3 mm 0.018 embolization coils (Cook Medical, Bloomington, IN) were deployed distal and proximal to the aneurysmal area and several 8 mm coils were deployed inside the aneurysm, obtaining complete angiographic exclusion of the lesion.

The left subclavian artery was then accessed and the same triaxial system was used to coil embolize the distal IMA and the aneurysm. The neck of the IMA at the subclavian artery was wide and short, precluding safe and durable deployment of coils. After bilateral vertebral artery angiogram confirming patency of both vessels up to the basilar artery, a 10 × 60 mm FLUENCY stent graft (Bard Peripheral Vascular, Tempe, AZ) was deployed covering the ostium of the IMA and the left vertebral artery. A 10 Fr ProStar XL (Abbott Cardiovascular, Ill) was deployed at the access site (Fig. 3).

The postoperative period was uneventful. Mild and self-limited anterior chest pain was managed with nonsteroidal anti-inflammatories and attributed to aneurysm thrombosis.

At 6 months, the patient was asymptomatic, and a CT scan showed exclusion of both aneurysms and reduction of their diameters (Fig. 4).

At 5-year follow-up, the left IMA aneurysm was not visible. On the right side, the aneurysm had recanalized proximally with an increase in its diameter to 4.1 cm, the proximal IMA was 7 mm in diameter. The lesion was re-embolized with several 16 mm coils (Cook Medical, Bloomington, IN), and an AMPLATZER II plug (St Jude Medical, St Paul, MN) was deployed at the neck with complete exclusion on follow-up CTA scan.

At age 47, 46 mm dilatation of the ascending aorta was found on CT scan and both aneurysms remained excluded, the patient was referred for surgical repair. The patient postponed this procedure and 8 months later was admitted with a type A dissection undergoing successful emergent open repair.

Ten years after the original procedure, the patient remains asymptomatic, both aneurysms remain excluded, and his left subclavian stent remains patent on CT scan.

DISCUSSION

IMA aneurysms and pseudoaneurysms, although rare, have been described in patients with multiple connective tissue diseases,[4–7] vasculitis,[8,9] infection, direct trauma due to median sternotomy[10] or subclavian venous puncture,[11] being the later the most common causes. Atherosclerosis has been described

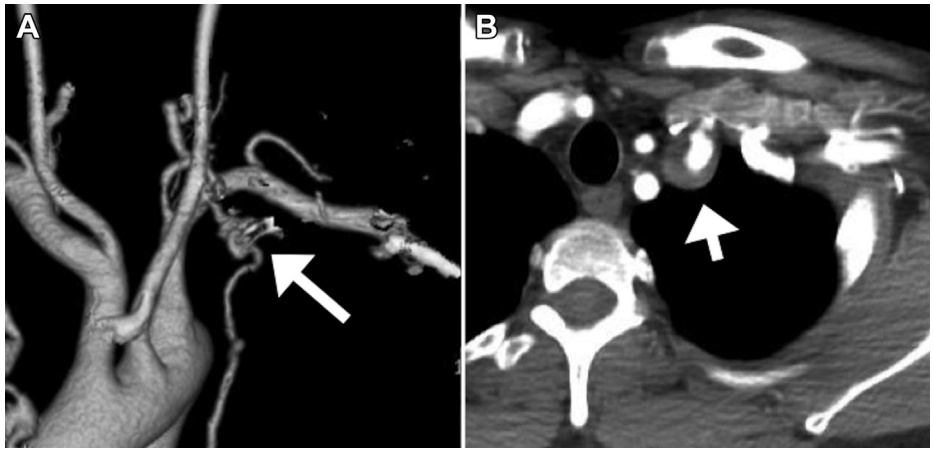

Fig. 2. Preoperative CT Angiogram. Volumen rendering **(A)** shows the location of a left internal mammary artery aneurysm (*arrow*). Contrast phase **(B)** shows the same aneurysm with mural thrombus inside (*arrow*).

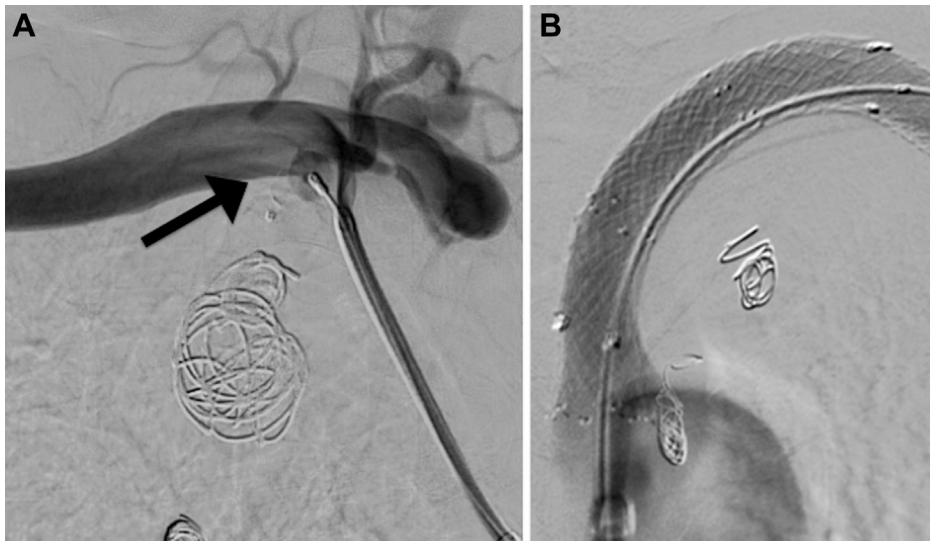

Fig. 3. (A). Intraoperative angiogram during reintervention of right IMA aneurysm, during deployment of the AMPLATZER® device. **(B)**. Final angiogram after deployment of FLUENCY® covered stent.

as a possible etiology, but at a different age and risk factor status.[12] The first report of an internal mammary artery aneurysm associated with MFS dates from 1999.[13]

Although it is common to associate MFS with aortic disease, aneurysms have been described in other locations. In this case, the diagnosis was timely obtained in a work-up CT scan. In regular clinical practice, an echocardiogram would be considered a reasonable alternative in an asymptomatic To rule out ascending aortic dilatation and aortic valve involvement in Marfan's patient, it would have missed the mammary artery dilatation.

Rupture of true IMA aneurysms has been well documented in cases of diseases of the arterial media[8] and in relation to neurofibromatosis.[14] There are reports of successful open surgical or endovascular treatment.

Coil embolization of the internal mammary artery was first reported in 1999 by Common et al.[13] and has been successfully used to treat pseudoaneurysm secondary to arterial injury during subclavian vein puncture for central venous access[11] or median sternotomy[10] and should probably be considered the treatment of choice if feasible, due to its minimally invasive nature.

Even though open repair through a limited thoracotomy or thoracoscopy allows excision or ligation of the aneurysm and provides tissue samples for histology,[9] embolization is far less invasive, can

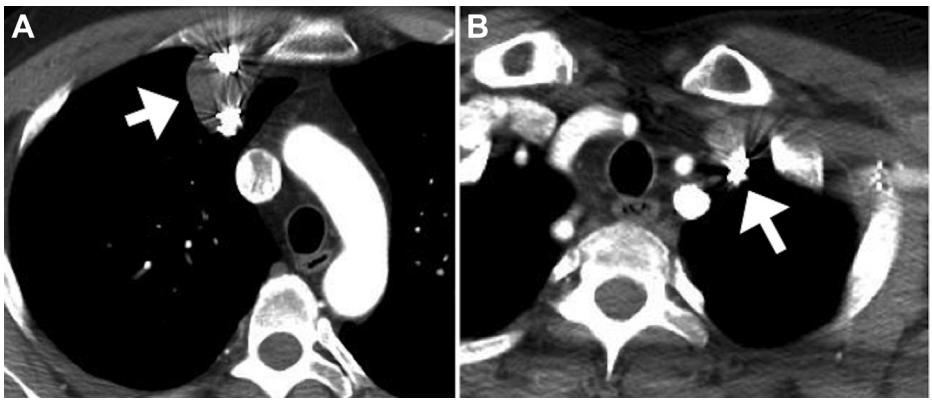

Fig. 4. Early follow-up CT scan after coil deployment and secondary thrombosis of **(A)** right IMA aneurysm (*arrow*) and **(B)** left IMA aneurysm (*arrow*).

be performed as an outpatient under local anesthetic, and results in prompt exclusion and thrombosis, without reconstruction of the vessel, therefore avoiding the decision between open surgical repair, involving anastomosis prone to pseudoaneurysm degeneration, or endografting and an unknown long-term result in a young patient. There are reports of IMA aneurysms in MFS, Loeys–Dietz syndrome (LDS), Ehlers–Danlos syndrome (EDS), and SMAD3 mutation patients successfully treated with coil embolization.[5–7,13,15]

Follow-up imaging is important since other arteries may be involved over time,[15] and failure of the procedure can be timely diagnosed and treated, as happened for this patient. This also allowed for diagnosis of the ascending aorta dilatation that caused type A dissection that could have been prevented in this patient if he had not postponed his treatment. Based on aortic experience, probably obtaining a CT scan close to the procedure to demonstrate successful exclusion of the aneurysms and then yearly would be a cost-efficient protocol.

Even though very rare, IMA aneurysms can be readily diagnosed using CT scan in a population at risk and can be efficiently treated with endovascular techniques with good long-term results.

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