

Interventional repair of a vascular aneurysm in a patient with Marfan syndrome

SAGE Open Medical Case Reports
Volume 6: 1–5
© The Author(s) 2018
Reprints and permissions:
sagepub.co.uk/journalsPermissions.nav
DOI: 10.1177/2050313X18788448
journals.sagepub.com/home/sco



Mohammed E Ghonem, Xun Yuan, Andreas Mitsis
and Christoph A Nienaber

Abstract

Marfan syndrome is a heritable connective tissue disorder affecting skeletal, ocular and cardiovascular systems. Cardiovascular manifestations comprise aneurysmal dilatation of aortic root, aortic dissection and rupture; peripheral arterial aneurysms have been reported in femoral, iliac and subclavian arteries with surgical reconstruction as the first-line therapeutic option. We report a Marfan patient with a symptomatic aneurysm of left subclavian artery in the intrathoracic retro-clavicular space; instead of open surgical resection, an endovascular solution was successfully applied by use of a flexible self-expanding stent-graft (W.L Gore® Viabahn® Endoprosthesis 9 × 100 mm) to exclude the aneurysm. This case exemplifies a modern option to manage vascular pathology even in patients with Marfan syndrome. Follow-up over 1 year proved very reassuring with complete remodelling and resolution of the aneurysm; long-term follow-up is certainly warranted, considering the potential of recurrence or initial reactive hyperplasia.

Keywords

Marfan syndrome, left subclavian aneurysm, stent-graft, endovascular management

Date received: 29 November 2017; accepted: 11 June 2018

Introduction

An isolated arterial aneurysm of the subclavian-axillary segment is unusual even in Marfan syndrome (MFS).¹ Depending on the location, extent and morphology, such aneurysms can rupture or cause thoracic outlet syndrome and will probably be seen more often with surveillance and increasing age.^{1–3} Once diagnosed, surgical resection is recommended in view of a high risk of both rupture or ischemic complications secondary to embolic events originating from arterial lesions proximal to carotid and vertebral territories, or in the case of retrograde embolization;^{1,3,4} only two cases are reported to be managed in an emergency setting by catheter-based procedures.^{5,6}

Case report

A 46-year-old lady had undergone multiple surgeries for vascular manifestations of a genetically confirmed MFS. In 2005, she had undergone a Crawford II repair for descending aortic dissection. In 2008, she received a valve-sparing aortic root replacement for ascending aortic dissection and had undergone bilateral mastectomy for breast cancer (BRCA2 mutation) in 2007.

In 2016, a routine computed tomography (CT) scan in an outside hospital revealed a newly developed circumscribed aneurysm (4.1 × 3.3 cm) of the left subclavian artery (LSA) in the retro-clavicular intrathoracic space (Figure 1). Clinically, a recent onset of pulsatile sensation in the left supra-clavicular fossa and increasing hoarseness were noted by the patient. Physical examination confirmed a palpable swelling in the retro-clavicular space. The patient was initially offered surgical resection, however, with limited mobility; after previous thoracotomies and restricted lung function from multiple thoracotomies, she declined open surgery and opted for an endovascular solution. The endovascular strategy eventually was to place a highly flexible self-expanding stent-graft (W.L Gore® Viabahn®

Cardiology and Aortic Centre, Royal Brompton & Harefield NHS Trust and Imperial College, London, UK

Corresponding Author:

Christoph A Nienaber, Cardiology and Aortic Centre, Royal Brompton & Harefield NHS Trust and Imperial College, Sydney Street, London SW3 6NP, UK.

Email: C.nienaber@rbht.nhs.uk



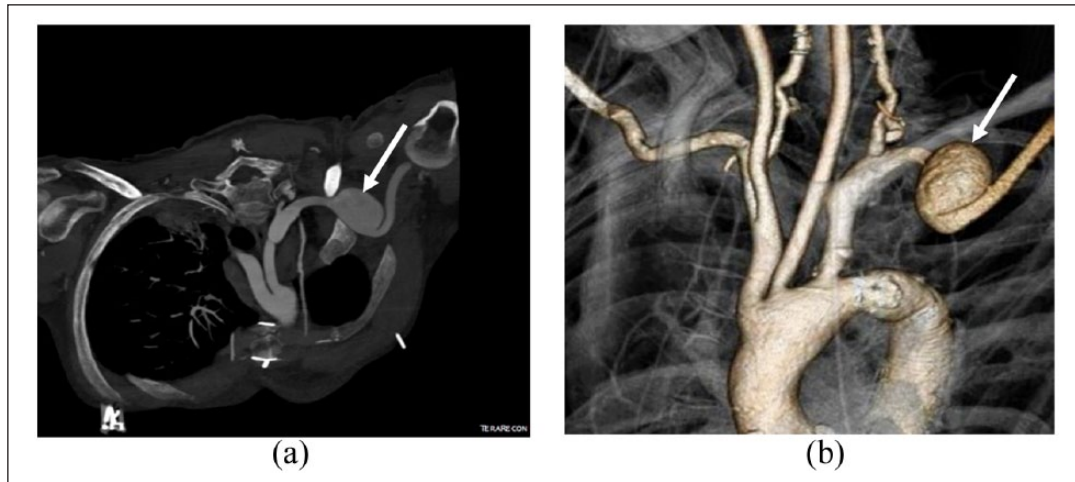


Figure 1. 2D (a) and 3D (b) computed tomography (CT) angiogram demonstrates a circumscribed aneurysm of left subclavian artery measuring 4.1×3.3 cm in the intrathoracic retro-clavicular space. (arrow).

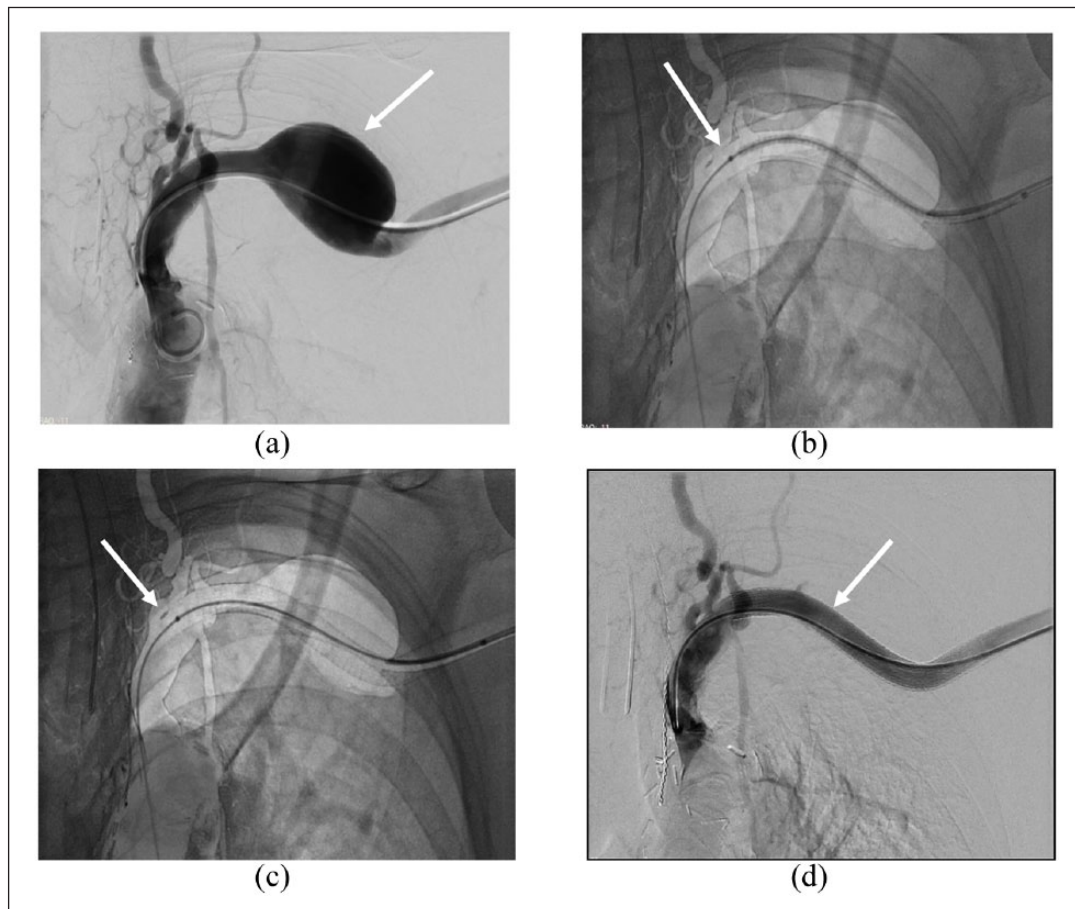


Figure 2. Intraoperative fluoroscopy images demonstrate: (a) 6F pigtail catheter navigated to the aortic arch across the aneurysm of left subclavian artery (LSA), arteriogram confirms a circumscribed aneurysm (arrow). (b) A 0.035 in guide wire was navigated across the aneurysm to the ascending aorta, and the highly flexible self-expanding covered stent-graft (Viabahn® 9×100 mm) was advanced along the wire, positioned correctly and ready for deployment (arrow). (c) LSA stent-graft has been successfully deployed from just beyond the proximal branches of LSA, to the junction with the axillary artery (arrow). (d) A completion arteriogram confirmed correct positioning of LSA stent-graft, the aneurysm sac has been successfully excluded (arrow) and the patency of axillary artery and left internal mammary artery.

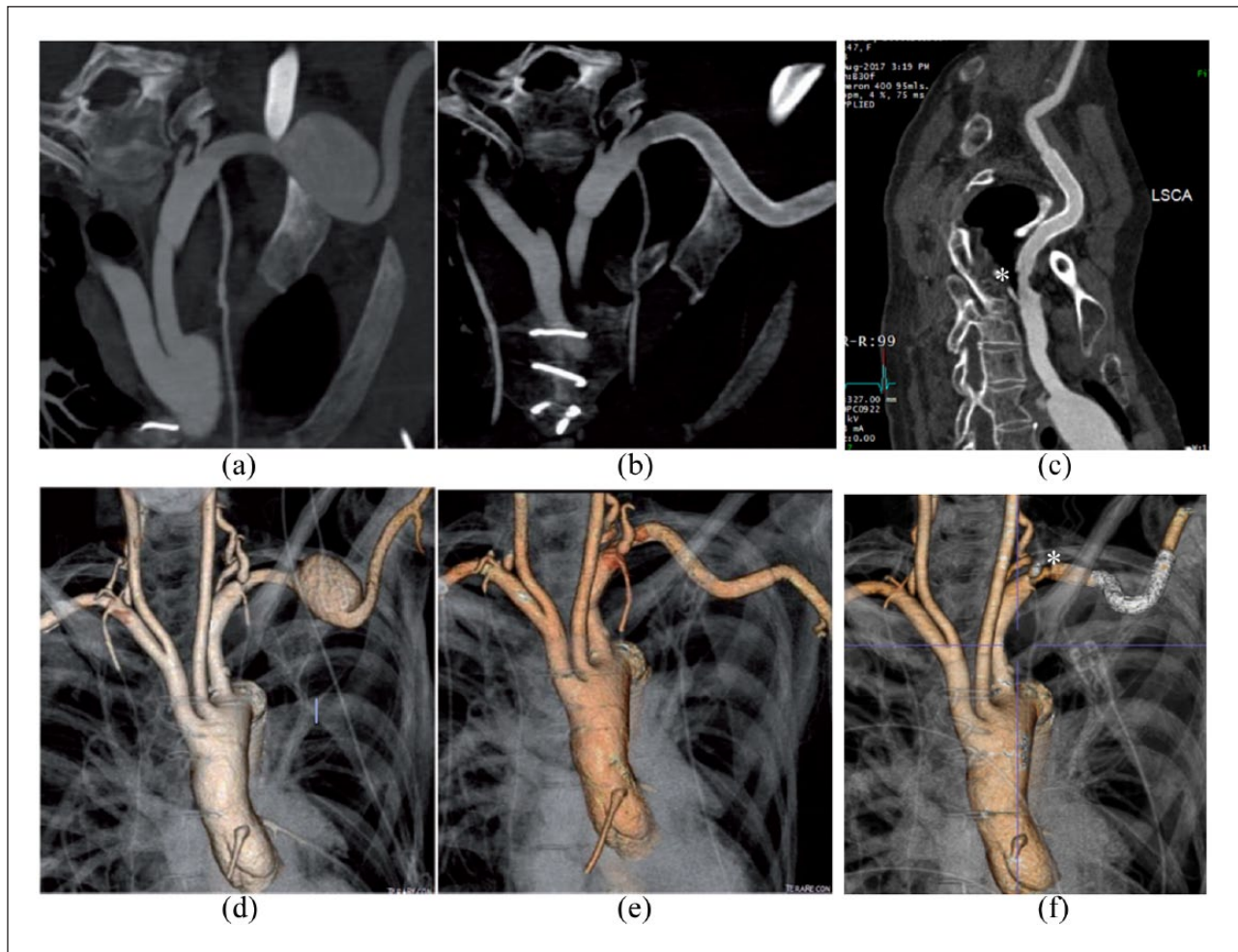


Figure 3. 2D and 3D volume rendered computed tomography (CT) angiogram demonstrates: (a) and (d) Pre-procedure, circumscribed LSA aneurysm measuring 4.1×3.3 cm. (b) and (e) Post-procedure; before hospital discharge, the LSA aneurysm was completely excluded after deployment of Viabahn® stent-graft (9×100 mm). (c) and (f) At 1-year follow-up, a CT scan showed the LSA stent-graft that remains widely patent with no evidence of the previous aneurysm. Immediately proximal to the stent and distal to the left vertebral origin, there is a moderate concentric narrowing with an estimated stenosis 30% (star).

Endoprosthesis 9×100 mm) across the aneurysm with about 3 cm of landing zone on either end, leaving left vertebral and mammary artery unobstructed. For this percutaneous strategy, a standard 9F sheath was advanced into the left brachial artery. Heparin was given as soon as the access was established. Then, a '0.035' J-tip guide-wire was navigated across the aneurysm to reach the ascending aorta, followed by a Viabahn stent-graft (9×100 mm) advanced safely over this wire and precisely deployed to exclude the aneurysm (Figure 2(a)–(c)). A completion angiogram confirmed correct positioning and absence of endoleak (Figure 2(d)); the brachial artery access was closed by manual compression for 30 min and then a large radial TR-band® for 3 h. Post-procedure, the patient was stable and recovered well. Before hospital discharge, a CT angiogram demonstrated that the LSA stent had been precisely deployed, not compromising vital side branches and completely excluding the aneurysmal sac (Figure 3(b) and (e)). The patient went home on a combination medication of Aspirin 75 mg,

Bisoprolol 5 mg and Losartan 50 mg, and was followed for 1 year; at 1-year follow-up, a CT scan confirmed no endoleak, a completely thrombosed LSA aneurysm and eventually complete resolution of the aneurysmal sac (Figure 3(a)–(f)). Informed consent was obtained by the patient in writing to allow publishing the case and the associated images. As the durability of results after stent-graft peripheral arteries in MFS patients is still a matter of debate and possibly controversial, such patients are currently followed by CT Angiogram or Magnetic Resonance Angiography (MRA) according to surveillance protocol (in annual intervals).

Discussion

MFS is a heritable disorder of the connective tissue with large interfamilial variability, complete penetrance and autosomal dominant transmission, but with pleiotropic manifestation affecting cardiovascular, musculoskeletal and ophthalmologic

systems.⁷ MFS is caused mostly by a mutation in the fibrillin-1 (FBN1) gene.⁸ This gene produces fibrillin-1 protein, which is the major component of microfibrils in the extracellular matrix. These fibres are important and responsible for incorporating elastin into elastic fibres.⁹ The defect in collagen and elastin formation caused by FBN1 mutation may lead to fragmentation and loss of tensile strength of the vessel fibres, resulting in stretching of the vessel wall and formation of both aneurysm and dissection.¹⁰

The management of this challenging case embodies a new endovascular strategy for a vascular aneurysm in MFS when open surgical repair would incur substantial risk; percutaneous 'neo-branching' can provide a satisfactory solution with both excluding the aneurysmal sac and anchoring the graft snugly within target vessel.¹¹ The case highlights the possibility of a change from the conventional approach of resecting a subclavian artery aneurysm to a minimalistic intervention.^{12,13} Moreover, this novel endovascular solution was applied to a patient with MFS, a setting where endovascular strategies are far from being established.^{5,6} Conversely, particularly in cases with multiple previous thoracotomies and major surgery, a minimally invasive procedure may be the best option regardless of the Marfan background. Self-expanding flexible stents could be in fact the best option in such vascular pathologies of the upper arm, because they are less prone to deformation with movement of the shoulder joint;¹⁴ in contrast to previously used stiff balloon-expandable prototypes the heparin-coated Viabahn stent-graft is highly flexible allowing perfect apposition to the tortuous anatomy in the subclavian region.^{5,6,15} This makes it much more suitable than balloon-mounted stent-grafts in view of flexibility, wall apposition, freedom of movement and radial force especially considering delicate tissues. Finally, with follow-up over a full year and in view of the active lifestyle of the patient, no dislocation or graft failure was noted, but rather a suspicion of mild intimal hyperplasia at the proximal anchor zone of the Viabahn stent-graft, probably as a sign of excessive endothelialization, highlighting the need for further follow-up. MFS is considered a lifetime active disease that may chronically continue to degenerate with late signs of progression. The justification for a percutaneous stent-graft rather than an open surgical resection was the patient's preference (after multiple previous open operations) and the internal multidisciplinary decision to offer endovascular treatment even though utilization of a suitable Viabahn stent-graft in the LSA is currently not covered by instructions for use from the manufacturer. The successful use in this case with greater than 1-year follow-up, may in fact be an example to initiate a broader acceptance of the endovascular concept and certainly highlight the need for an international registry to collect information on peripheral interventions in patients with connective tissue disorders.

Conclusion

MFS is a genetic connective tissue disorder causing degenerative aneurysms not only of the aorta but also in

peripheral arteries. Aneurysms of the subclavian artery are rare but once diagnosed, should be treated when expanding. Surgical treatment is recommended as a standard, but the endovascular solution can be an advantageous option for these patients, in view of the anatomic suitability and co-morbidities. As durability of the stent-graft in Marfan patients is controversial, structured follow-up with CT imaging and possibly intravascular ultrasound surveillance should be put in place.

Acknowledgements

The authors acknowledge Dr Michael Rubens for his invaluable input in imaging post-processing and 3D rendering.

Declaration of conflicting interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship and/or publication of this article.

Ethical approval

Our institution does not require ethical approval for reporting individual cases or case series.

Funding

The author(s) received no financial support for the research, authorship and/or publication of this article.

Informed consent

Written informed consent was obtained from the patient(s) for their anonymized information to be published in this article.

References

1. Salo JA, Ala Kulju L, Heikkinen S, et al. Diagnosis and treatment of subclavian artery aneurysms. *Eur J Vasc Surg* 1990; 4(3): 271–274.
2. Thomazinho F, Sardinha WE, Silvestre JMS, et al. Complicações arteriais da síndrome do desfiladeiro torácico: arterial complications of thoracic outlet syndrome. *J Vasc Bras* 2008; 7(2): 150–154.
3. Davidovi LB, Markov DM, Pejki SD, et al. Subclavian artery aneurysms. *Asian J Surg* 2003; 26(1): 7–11.
4. Malik MK, Kraev AI, Hsu EK, et al. Spontaneous axillary artery aneurysm: a case report and review of the literature. *Vascular* 2012; 20(1): 46–48.
5. Tassiopoulos AK, Nadalin BA, Labropoulos N, et al. Endovascular repair of a symptomatic subclavian artery aneurysm in a patient with Marfan syndrome: a case report. *Vasc Endovascular Surg* 2006; 40(5): 409–413.
6. Rose JF, Lucas LC, Bui TD, et al. Endovascular treatment of ruptured axillary and large internal mammary artery aneurysms in a patient with Marfan syndrome. *J Vasc Surg* 2011; 53(2): 478–482.
7. Pyeritz RE and McKusick VA. The Marfan syndrome: diagnosis and management. *N Engl J Med* 1979; 300(14): 772–777.
8. Faivre L, Collod-Beroud G, Child A, et al. Contribution of molecular analyses in diagnosing Marfan syndrome and type

- I fibrillinopathies: an international study of 1009 probands. *J Med Genet* 2008; 45(6): 384–390.
9. Godfrey M, Menashe V, Weleber RG, et al. Cosegregation of elastin-associated microfibrillar abnormalities with the Marfan phenotype in families. *Am J Hum Genet* 1990; 46(4): 652–660.
 10. Bunton TE, Biery NJ, Myers L, et al. Phenotypic alteration of vascular smooth muscle cells precedes elastolysis in a mouse model of Marfan syndrome. *Circ Res* 2001; 88(1): 37–43.
 11. Vandormael IL, Salmasi MY, Yeh JS, et al. Endovascular ‘neobranching’ to manage acute aortic syndrome. *Catheter Cardiovasc Interv* 2017; 190(2): 298–302.
 12. May J, White G, Waugh R, et al. Transluminal placement of a prosthetic graft-stent device for treatment of subclavian artery aneurysm. *J Vasc Surg* 1993; 18(6): 1056–1059.
 13. Schoder M, Cejna M, Holzenbein T, et al. Elective and emergent endovascular treatment of subclavian artery aneurysms and injuries. *J Endovasc Ther* 2003; 10(1): 58–65.
 14. Davey P and Wyatt M. Aneurysmal disease. In: Davies AH and Brophy CM (eds) *Vascular surgery*. London: Springer, 2006, pp. 191–219.
 15. Kasirajan K, Matteson M, Marek JM, et al. Covered stents for true subclavian aneurysms in patients with degenerative connective tissue disorders. *J Endovasc Ther* 2003; 10(3): 647–652.