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Case Report Special Issue

Hybrid Endovascular Repair for Dysphagia Lusoria in a Patient with Marfan Syndrome

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Abstract

Objective: We present the case of a 51-year-old male with dysphagia lusoria and Marfan syndrome treated with a hybrid repair.

Methods: Review of chart and pre- and post-operative imaging was performed.

Results: The patient presented with a seven-year history of dysphagia with worsening symptoms including hoarseness. He had a prior aortic valve replacement and Bentall procedure, and no other significant comorbidities. Contrast CTA demonstrated an Aberrant Right Subclavian Artery (ARSA), with an area of focal dissection and aneurysmal degeneration measuring 26mm. The aorta measured 33mm proximally and distally, and the ARSA came off 15mm distal to the Left Subclavian Artery (LSA). An open right subclavian to carotid artery transposition was performed through a right neck incision. A 38mm×38mm×100mm proximal free flow Valiant™ device was used to cover the origin of the aberrant right subclavian artery. Completion arteriography and intravascular ultrasound were performed, demonstrating proximal and distal fixation with no evidence of endoleak or dissection.

The patient recovered well and was discharged on postoperative day three. At six weeks, he continued to recover uneventfully. CTA at three months demonstrated a stable repair. At one year, he has no complaints of chest or back pain and recently underwent a successful repair of an LSA aneurysm.

Conclusion: Aberrant right subclavian artery and Marfan syndrome are both rare conditions. When seen together, the treatment of this combined pathology can be challenging. A hybrid approach appears to be safe and effective for this clinical scenario.

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Introduction

Aberrant Right Subclavian Artery (ARSA), or arteria lusoria, is the most common congenital aortic arch anomaly, occurring in less than 1-2% of the general population [1-2]. Occasionally, the ARSA may compress the esophagus as it courses transmediastinal, causing dysphagia lusoria [3]. Successful endovascular and open repairs have been reported in the treatment of ARSA, however, the repair of ARSA in patients with Marfan Syndrome has been infrequently discussed in the literature [4-8]. We present a case of successful endovascular repair of an aneurysmal, aberrant right subclavian artery in a patient with Marfan Syndrome. We have the patient's expressed, written consent to publish case details and images related to his case for this purpose.

Case Report

A 51-year-old man with Marfan Syndrome presented with a seven-year history of dysphagia. For the past several years, his symptoms worsened to include hoarseness. His history includes aortic valve replacement and Bentall procedure six years prior for which he complied with daily warfarin therapy. His blood pressure and heart rate were well-controlled on a beta blocker and an ARB, and he had no other significant comorbidities. He was referred to our center from another hospital for discussion of endovascular repair to address his dysphagia lusoria.

The patient underwent CTA with contrast that demonstrated an aberrant right subclavian artery with an area of focal dissection and aneurysmal degeneration measuring 26mm (Figures 1,2). The aorta measured 33mm both proximally and distally, and the right subclavian artery came off the aorta 15mm distal to a widely patent left subclavian artery. There was moderate dilatation of the left subclavian artery, and it was unclear whether it would be possible to clamp this both proximally and distally. Therefore, the right carotid artery was identified and an open right subclavian to carotid artery transposition was performed through a standard neck incision. Then, a 38mm×38mm×100mm proximal free flow Valiant™ (Medtronic Inc, Santa Rosa, CA) device was used to cover the origin of the aberrant right subclavian artery. Completion arteriography and intravascular ultrasound were performed, demonstrating excellent proximal and distal fixation with no evidence of endoleak or dissection. There was antegrade flow through both carotids as well as the left subclavian artery.

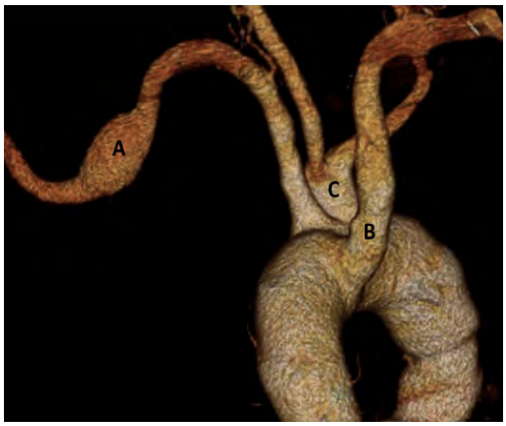

Figure 1: 3D CTA showing A) LSA with dilation preventing subclavian-subclavian bypass, B) Aberrant right subclavian artery, C) Shared origin of right and left carotid arteries.

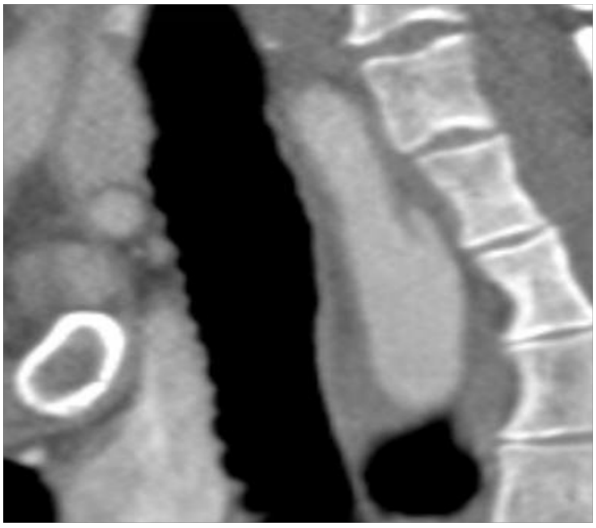

Figure 2: Focal dissection of the aberrant right subclavian artery seen between esophagus and spinal column.

The patient recovered well and was discharged on postoperative day three after restarting his warfarin. At six weeks, he continued to recover uneventfully. A CTA at three months demonstrated a stable repair with no evidence of endoleak (Figures 3,4). He continued without complaints of chest or back pain and at one year underwent resection of his left subclavian artery aneurysm with interposition graft with no complications.

Discussion

Arteria lusoria is characterized by the right subclavian artery originating directly from the aortic arch (usually distal to the left subclavian) rather than from the brachiocephalic trunk. Generally the condition is asymptomatic, but the proximity of the ARSA to the esophagus may result in dyspnea, chronic cough, or dysphagia lusoria if the ARSA compresses the esophagus [1,3,8].

Treatment is indicated to relieve dysphagia lusoria, as well as in cases where the ARSA is aneurismal [9]. Both open and endovascular approaches have been successfully performed and reported.[4,9] As TEVAR becomes more common, a variety of endovascular and hybrid techniques have been reported to treat ARSA [10-13]. A combined intervention with right subclavian artery transposition, distal or proximal artery ligation or endovascular occlusion, and TEVAR appears to be a promising endovascular solution for patients with ARSA [10]. Several hybrid procedures have been performed combining bilateral carotid-subclavian bypass with TEVAR, covering the origin of the ARSA [14-17]. Lacroix et al., also report successful TEVAR with a carotid-subclavian bypass and ARSA embolization to treat an aneurysmal ARSA [18]. Caution is necessary, however, when attempting endovascular occlusion of an aneurysmal ARSA, due to reports that proximal migration of a vascular plug deployed in an ARSA can cause recurrent dysphagia [19].

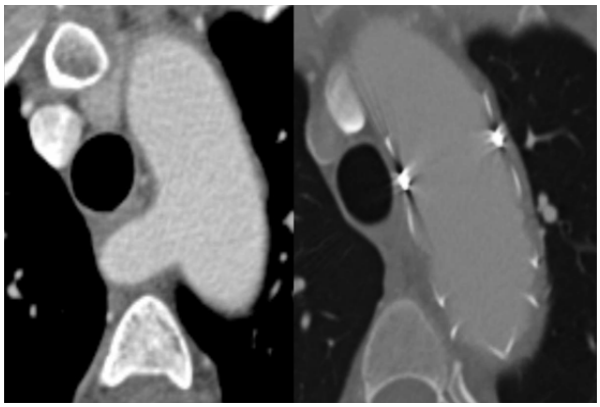

Figure 3: Comparison of pre-operative and post-TEVAR CTA showing successful repair.

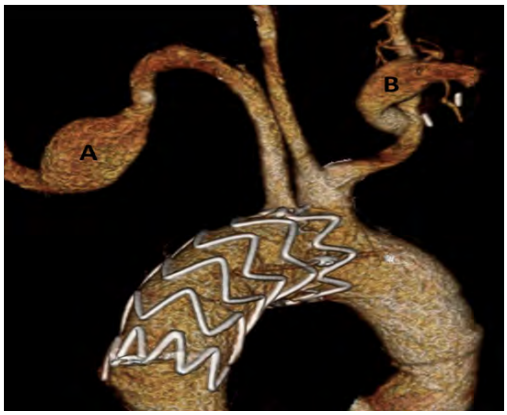

Figure 4: 3D CTA at 3 months post-TEVAR showing successful repair. A) LSA with dilatation, later repaired, B) Right subclavian to carotid artery transposition.

In particular, the use of an open right subclavian to right carotid artery transposition in conjunction with TEVAR in the presence of left subclavian artery dilatation is unique in this patient population. Mazzaccaro et al., echoed similar concern regarding a diseased left subclavian artery, and chose to use a periscope technique rather than a hybrid repair [13]. Multiple instances of successful right subclavian to right carotid bypass in conjunction with TEVAR for treatment of ARSA have been described [10,18]. We have also performed similar

procedures with success in other patients with dysphagia lusoria at our institution. However, the positive outcome in this patient show that hybrid endovascular repair of ARSA is a viable option even with the added complexities in Marfan syndrome.

Endovascular treatment of the thoracic aorta in patients with Marfan syndrome is the subject of some controversy [20]. In general, it has been suggested that while TEVAR may be a life-saving option in emergency situations, it has a high rate of reintervention and risk of complications in patients with Marfan syndrome, and thus should be used as a bail-out or bridge procedure to open surgical treatment [21-26].

In terms of Marfan syndrome patients with ARSA, few cases have been described. LeRoux et al., describe the management of 22 patients with ARSA, including one patient with Marfan syndrome [5]. However, it is not clear what technique was used in this patient. Kamiya et al., described 8 patients with aneurysmal ARSA, one of which had Marfan syndrome and a chronic type B aortic dissection, and was treated with a staged repair including a carotid-subclavian bypass, closure of the ARSA, and replacement of the descending aorta [6]. To our knowledge, few Marfan syndrome patients have undergone the type of hybrid endovascular repair of ARSA described in most of the literature regarding the endovascular treatment of ARSA. One case, described by Stanley et al., in 2012, was complicated by a Kommerell diverticulum and acute dissection of an aneurysmal ARSA [7]. Our patient, who had minimal dilation of his ARSA and dysphagia lusoria, now provides another important example of a successful endovascular repair of ARSA in a patient with Marfan syndrome.

Conclusion

In conclusion, we present the case of a 51-year-old male with combined dysphasia lusoria and Marfan syndrome effectively treated with a hybrid endovascular procedure, previously thought to the high risk in this patient population. We propose this to be a safe and effective treatment modality for patients with both dysphagia and aneurysmal dilatation in the setting of aberrant right subclavian artery and connective tissue pathology. Due to both the rarity and complexity of this combined condition, it would be hard to study this on a more prospective and randomized basis. Therefore, we believe this to be a new standard of care for this clinical scenario.

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