Unusual Rapid Evolution of Type B Aortic Dissection in a Marfan Patient Following Heart Transplantation: Successful Endovascular Treatment

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A patient with Marfan syndrome with previous Bentall operation for mitral and tricuspid valve repair, required orthotopic cardiac transplantation for end stage cardiomyopathy. Postoperatively he suffered type-B aortic dissection, despite normal aortic diameters. Following sudden increase of aortic diameters, two years later, he underwent successful stent graft implantation. In patients with Marfan syndrome, post transplantation morbidity is high, with a 40% incidence of thoracic aortic dissection. This case highlights the potential of endovascular approach for treating post-transplantation aortic dissection.

Keywords: Marfan syndrome; Heart transplantation; Type-B aortic dissection; Endovascular stent-graft.

Introduction

Marfan syndrome is an autosomal dominant disorder of connective tissue, characterised by highly variable clinical manifestations primarily in skeletal, ocular and cardiovascular organ systems.1 Usually the cardiovascular manifestations, such as progressive aortic dilation/dissection and mitral valve prolapse, represent the main cause of death. Patients who have undergone surgical correction for mitral valve prolapse and insufficiency may ultimately develop end-stage congestive heart failure2-3 for which heart transplantation is the only life-saving procedure. Aortic dissection is uncommon in cardiac allograft recipients3 but it is a potential complication in those with Marfan syndrome.

Case Report

In 2003 a 36-years-old male with Marfan syndrome was referred to our hospital for cardiac transplantation due to end stage cardiomyopathy. Ten years earlier, the patient has been underwent the Bentall operation (mitral and tricuspid valve repair for ascending aortic aneurysm, mitral and tricuspid valve regurgitation). An MRI study performed in 2003, before heart transplantation, did not show any morphologic or dimensional anomaly of the arch (30 mm) or descending thoracic aorta (29 mm). After heart transplantation the patient underwent a standardized immunosuppression therapy protocol with cyclosporin A, mychophenolato mofetil and prednisone, tapered as dictated by clinical status. Thirty days after transplantation the patient complained of a sudden, severe, infrascapular back pain. MR angiography showed Type-B aortic dissection (Fig. 1): the entry site was below the origin of the left subclavian artery, the intimal flap extended to the common iliac arteries and there was a slight increase of the descending aortic lumen (31 mm). Since the patient’s clinical condition was stable, with normal systemic pressure, he was discharged and followed with CT scan or MRI every 6 months. Initially the annual increase in aortic diameter was limited to 3–4 mm but 2 years later MRI revealed a sudden dimension increase, to 21 mm of the false lumen, with maximum diameter of true and false lumen.
of 55 mm in the proximal thoracic segment, 45 mm in the thoracic aorta below the diaphragm and 36 mm in the abdominal aortic segment. Because of the high risk of open surgical repair, the patient underwent endovascular stent-graft treatment. The procedure was performed under general anaesthesia with surgical exposure of common femoral artery. Three Valiant thoracic stent grafts segments (Medtronic, Santa Rosa, CA) were delivered under fluoroscopic and TEE guidance under hypotension, the most proximal stent with bare spring was placed below the left common carotid artery and the most distal with closed web 2 cm above the coeliac artery. The diameter of the stent-graft was oversized by 10% with respect to the aortic arch diameter. Post-procedural angiography and TEE showed closure of the primary and of two additional thoracic entry sites, with false lumen thrombosis down to the coeliac artery. The postoperative course was uneventful, with the exception of a pronounced post-implantation syndrome (fever, elevated C-reactive protein levels, and leukocytosis). The patient was discharged on the 14th postoperative day. CT scan performed during follow-up (one year) showed a complete occlusion of the thoracic false lumen down to the coeliac artery (Fig. 2) with initial external sac shrinkage of 3 mm. The false lumen remained open in the abdominal aortic segment but without any further increase in sac diameter.

**Discussion**

Management of patients with Marfan syndrome still constitutes a challenge and some are reluctant to place these patients on heart transplantation waiting lists. A variety of aortic complications can occur. The connection between the donor and the recipient aorta is a potential source of early and late complications as a result of infection, compliance mismatch and weakness of the aortic wall. In patients with Marfan syndrome, the risk of dissection is higher due to the aortic dilatation and the presence of aneurysms. Endovascular therapy offers a safe and effective alternative to open surgery, especially in high-risk patients. The use of endovascular stent grafts enables the treatment of complex and high-risk cases, improving the outcomes of patients with Marfan syndrome.
syndrome, there is a 40% incidence of post-operative thoracic aorta dissection, while dissection is extremely rare outside this setting, varying from 1–2%. The mechanism of post-operative aortic aneurysm and dissection formation is probably multifactorial. It has been suggested that, with improved cardiac output and abrupt change in systemic pressure, the aorta is exposed to increased stress by the transplanted heart. Another possible contributor is immunosuppression, because of collagen-weakening effect, secondary hypertension and accelerated atherosclerosis.

The expected increase of aortic diameter in uncomplicated type-B dissections also with Marfan syndrome is about 3 mm/year. Accordingly, an annual increase of 10 mm in aortic diameter has been reported in abdominal aortic aneurysm in heart transplant recipients. An adjunct problem in these patients is that beta-blockers therapy, useful to control the rate of pressure change with time (dp/dt) in aortic dissection, should be administered cautiously, since circulating catecholamines appear crucial for adequate cardiac function of the grafted heart during exercise.

The feasibility of endovascular repair in Marfan syndrome is widely debated, because of the potential risk induced by radial forces of the stent-graft upon the fragile aortic wall. However, conventional surgery for descending aorta dissection carries a high risk in these patients. In our case, open surgery could be aggravated by several adjunct problems, such as discontinuation of immunosuppressive therapy, increased risk of infection or alterations in the coagulation system.

In conclusion, endovascular repair is a recognised appropriate alternative to open surgery in high risk patients and may represent the only life-saving approach when multiple co-morbidities are present.

References

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