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Late Reoperation for Proximal Aortic Complication in a Marfan Patient Following Ascending Aortic Grafting

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ABBREVIATIONS

TTE = transthoracic echocardiogram

TEE = transesophageal echocardiogram

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ABSTRACT

We report a case of a female patient with Marfan syndrome who suffered a type A acute aortic dissection eleven years ago. At that time she underwent supracoronary ascending aortic grafting and resuspension of the incompetent aortic valve. Recently this patient presented to us with uncontrolled hypertension and a transthoracic echocardiographic study revealed severe dilatation of the sinuses of Valsalva. She required reoperation and a Bentall procedure was performed. Surgical treatment of the Marfan syndrome is discussed.

INTRODUCTION

Marfan syndrome is a variable, autosomal dominant connective tissue disorder, affecting mainly the cardiovascular system, eyes and skeleton.¹ It is a well-defined genetic defect (chromosome 15q) with both a structural and functional propensity for aortic dilatation and dissection. Acute type A aortic dissection is the most frequent cause of premature death in this syndrome. The immediate priority in surgery for these situations is survival. The gold standard of surgical treatment is the Bentall procedure. However, if a conservative surgical technique is employed (isolated replacement of the ascending aorta and resuspension of the incompetent aortic valve), the patient should be kept on a beta-blocker with continuous surveillance of the whole aorta by magnetic resonance imaging or computed tomographic scan. Not excluding the aortic sinuses results in aneurysmal dilatation and the need for reoperation.²

CASE REPORT

A 51-year-old Marfan female patient with a past history of surgery for type A aortic dissection presented to us with uncontrolled hypertension. The patient was known hypertensive and she had been on medical treatment which recently was insufficient to control the blood pressure. She was supposed to attend regular follow up, but admitted neglect of her appointments over the last three years.

Diagnosis of Marfan's syndrome was suspected at a young age, even if patient

matched only one major of the Ghent criteria (skeletal) and ocular involvement by that time. Her past medical history was significant for emergency cardiac surgery due to Type A aortic dissection eleven years ago in our hospital. Aortic dissection was extended from the aortic root to the iliac arteries. At that stage she was in a confusional state and there were bruits on both carotid arteries. There were no palpable pulses in the right upper and the left lower extremity. She was taken to the operating room in a catastrophic condition and underwent ascending aorta replacement with a tubular graft (Vascutec, 25 mm) and resuspension of the incompetent aortic valve.

Histopathological examination of the aortic wall revealed media with a profound decrease in the amount of elastin present and loss of the highly aligned and ordered lamellar arrangement. Extensive deposits of mucopolysaccharides were present throughout the media, and these changes were relatively uniform throughout the specimens. These findings were suggestive of Marfan syndrome. Patient responded well to surgery, had a very good recovery and discharged home with instructions for regular follow up.

On recent admission she was in sinus rhythm and her blood pressure was 170/95 mmHg. On auscultation a parasternal



FIGURE 1. Transesophageal echocardiogram. Midesophageal long axis view demonstrated significant aortic regurgitation.



FIGURE 3. Transesophageal echocardiogram. Mid-esophageal short axis view demonstrated dilatation of sinuses of Valsalva (diameter 67 mm); the right sinus of Valsalva is aneurysmal.



FIGURE 2. Transesophageal echocardiogram. Mid-esophageal long axis view demonstrated dilatation of Sinuses of Valsalva (diameter 67 mm). The right sinus of Valsalva is aneurysmal.



FIGURE 4. Transoesophageal echocardiogram. Short axis of the descending aorta demonstrated true and false lumens from the previous dissection.

LATE REOPERATION IN A MARFAN PATIENT

diastolic murmur was detected, heard best at the third left intercostal space radiating widely along the left sternal border. The electrocardiogram (ECG) revealed normal sinus rhythm and nonspecific ST segment changes. Chest x-ray showed cardiomegaly.

A transthoracic echocardiogram (TTE) demonstrated severe dilatation of the aortic root (diameter 67 mm) associated with severe aortic regurgitation. A right sinus of Valsalva aneurysm was present. The left ventricle (LV) appeared hypertrophic and dilated. LV function was mildly impaired. There was mild mitral regurgitation, due to the eccentric jet of the severe aortic insufficiency which was directed towards the anterior mitral valve leaflet, resulting in functional mitral valve prolapse.

A multiplane transesophageal echocardiogram (TEE) confirmed the above findings. True and false lumens in the descending aorta were visible as a consequence of the previous dissection. There was partial thrombosis of the false lumen in the aortic arch with some residual flow. A computer tomography angiography demonstrated severe aortic root dilatation and findings consistent with chronic dissection of the aortic arch and branches as well as of the descending aorta. Coronary angiography showed normal coronaries.

The patient was referred for urgent surgery. The diagnosis of dilated aortic root and right sinus of Valsalva aneurysm was confirmed intraoperatively. She underwent a Bentall procedure. The immediate postoperative course was uneventful, but the patient subsequently experienced ischemic injury of the liver and acute respiratory failure due to pulmonary aspiration which resulted in readmission in the intensive care unit and a long hospital stay. She was finally discharged home on the 55th post-operative day in a good condition.



FIGURE 5. Computer tomography angiography demonstrated aortic root sinus dilatation – right sinus of Valsalva aneurysm.

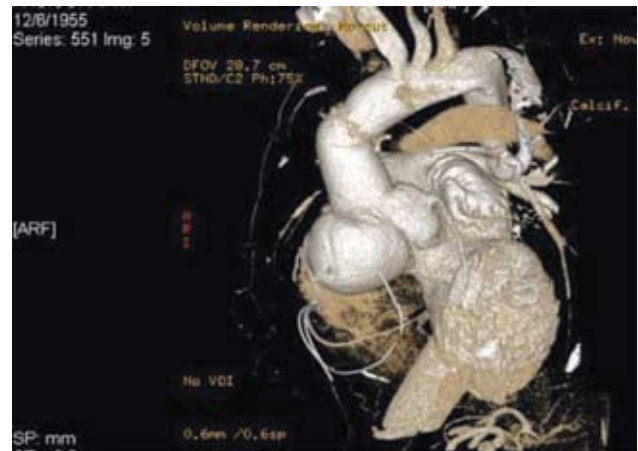


FIGURE 6. Computer tomography angiography demonstrated aortic sinus dilatation as well as findings consistent with chronic dissection of the aortic arch and branches along with the descending aorta.

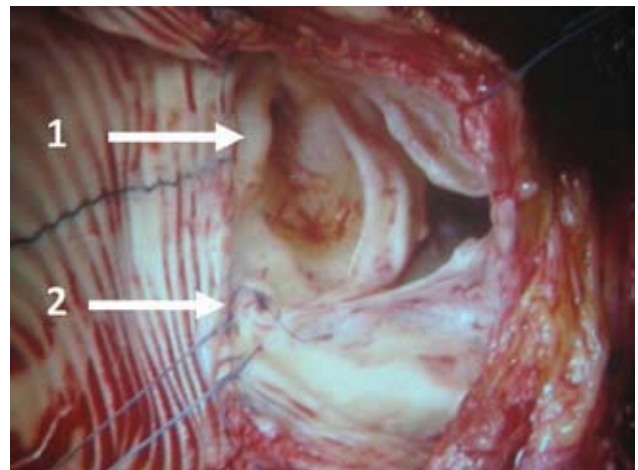


FIGURE 7. Intraoperative view of the dilated aortic root: arrows indicate the previously used dacron graft and left coronary ostium (1), and the pledget used to resuspend the commissure between left and non coronary cusps (2).

DISCUSSION

The Marfan syndrome is an autosomal dominant inherited connective tissue disorder with variable phenotypic expression of cardiovascular, ocular and musculoskeletal manifestations. The estimated prevalence of the syndrome is 1 in 10,000, and around 26% of cases have no family history, the condition resulting from a new mutation. The syndrome results from mutations in the fibrillin-1 (FBN1) gene on chromosome 15, which encodes for the glycoprotein fibrillin. Fibrillin represents the major structural component of connective tissue

microfibrils. Abnormalities involving microfibrils weaken the aortic wall. Severe elastic fiber degeneration occurs because mutant fibrillin is unable to bind calcium.¹⁻³

The characteristic features of the Marfan syndrome include progressive aortic dilatation associated with aortic valve incompetence, mitral valve prolapse and incompetence, lens dislocation and myopia, and a tall and thin body with long limbs, arachnodactyly, pectus deformities, and sometimes scoliosis. Marfan syndrome is currently diagnosed using criteria (*Ghent criteria*) based on an evaluation of the family history, molecular data, and 6 organ systems. The diagnosis cannot be based on molecular analysis alone because molecular diagnosis is not generally available, mutation detection is imperfect, and not all *FBN1* mutations are associated with Marfan syndrome. Life expectancy is primarily determined by the severity of cardiovascular involvement and has improved substantially in the past 30 years as a result of improved medical and surgical management. Without surgical intervention, many patients die in the third decade of their lives from complications of aortic root aneurysm, such as aortic rupture, aortic dissection, and aortic insufficiency.⁴

Dilatation of the aortic root is a well-known and potentially life-threatening cardiovascular manifestation in patients with Marfan syndrome.³ Sinus dilatation and increased wall stiffness elevate pulse pressure on the structurally attenuated aortic wall. Impaired mechanotransduction (nitric oxide production) causes failure of flow-mediated vasodilation with increased wall stress and cardiac workload.² The mechanism causing aortic valve insufficiency is usually the enlargement of one or more components of the aortic root complex (e.g., aortic sinuses and/or annulus), hence the moving of the aortic valve cusps away from their optimal coaptation line. Aortic dissection, on the other hand, occurs in up to 20% of Marfan patients. In this condition, aortic valve incompetence may occur due to dilatation of the sinotubular junction with acute distraction of the valve leaflets, and/or unhinging and prolapsed of the leaflets secondary to sinus wall dissection.⁵

According to Cameron et al.⁶ the current indications for aortic root replacement in the Marfan syndrome are: 1) aortic sinus diameter ≥ 5 cm or ≥ 4.5 cm among patients with family history or rupture or dissection, 2) ascending aortic dissection, whether acute or chronic, 3) aneurysm growth >1 cm/year, and 4) worsening aortic regurgitation in a dilated root when a valve-sparing procedure is desired. The introduction of the Bentall procedure and its variants into clinical practice had a major impact on life expectancy of patients with Marfan syndrome. Today, implantation of a valved conduit for aortic root replacement is the gold standard for patients with the syndrome presenting with aneurysms of the aortic root. Although early and late postoperative outcomes of this procedure have been excellent, a significant proportion of patients with mechanical valve substitutes experience complications related to

long-term anticoagulation. This fact and the generally young age of patients with Marfan syndrome undergoing surgery of the aortic root render biologic or even valve-sparing operative concepts attractive. However, there is reluctance among surgeons with regard to preservation of the aortic valve in patients with Marfan syndrome because leaflets carrying a structural fibrillin-1 defect may not be stable enough for a long time, regardless of the type of valve reconstruction.⁷ Recently Patel et al reported that Bentall procedure and valve sparing root replacement have similar operative results in Marfan syndrome. The procedures are distinguished by higher rates of thromboembolism among Bentall patients and higher rates of reoperation among patients having the valve sparing technique.⁸⁻¹¹ However, Cameron and colleagues⁶ reviewing their series, which is the most complete in the literature, concluded that the valve sparing approach shows promise but has not yet proven as durable as the Bentall procedure. The valve sparing method has been used in dissection patients, but without extensive experience. The extra time and judgment required may prejudice survival.

Conservative operations (repair of the dissected aorta and resuspension of the aortic valve) in the Marfan patient result in aneurysmal dilatation and the need for early reoperation. Those who have undergone inappropriate root operation often develop aneurysmal dilatation within a few years of surgery. The patient should be kept on a beta-blocker with continuous surveillance of the whole aorta by magnetic resonance imaging or computed tomography scan. A close follow-up of all Marfan patients is necessary to detect asymptomatic changes requiring surgery.^{2,12,13}

Our patient suffered aortic dissection Type A, eleven years ago. Because of the catastrophic spontaneous prognosis of acute type A dissection of the aorta, surgeon's immediate priority was survival, performing a palliative procedure (aortic valve resuspension and replacement of ascending aorta with a tubular graft) aimed at preventing the vessel from rupturing into the pericardium. The patient came back with severe dilatation of the aortic root eleven years later and underwent a high risk re-operation (logistic EuroSCORE=23,96%). This operation would have been of lower risk if she had attended her regular follow up appointments and the dilatation of the aortic root had been detected earlier. In Marfan patients who suffered dissection, close attention is recommended to prevent any other cardiovascular complication, especially if a conservative operation for type A dissection has been performed.

We conclude that in patients with Marfan syndrome and acute ascending aortic dissection, replacement of the aortic root is usually required in addition to repair of the dissected aorta, in order to eliminate the re-operation rate which is of high mortality and morbidity. This will out of necessity increase the complexity and duration of the procedure when

LATE REOPERATION IN A MARFAN PATIENT

compared to simple tube graft replacement of the ascending aorta and resuspension of the aortic valve.

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