

Giant Unruptured Aortic Root Aneurysm After Repair of Ascending Aortic Dissection Due to Marfan's Syndrome

Marfan Sendromlu Hastada Assendan Aorta Diseksiyonu Operasyonundan Sonra Gelişen Rüp-türe Olmayan Aort Kökü Dev Anevrizması

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ABSTRACT The risk of aortic dissection and aortic aneurysm in patients with Marfan syndrome has been known for years. A 46-year-old man with Marfan's syndrome was admitted to our clinic with a complaint of chest pain and palpitation in the last year. Hemiarcus replacement, aortic valve re-suspension and graft interposition between innominate artery and prosthetic ascending aorta was performed 4 years ago due to type 1 aortic dissection. Echocardiography and computed tomography revealed dilatation of aortic root (68 mm in diameter), with advanced aortic valve insufficiency. Hence, we successfully performed aortic root replacement with Bentall procedure. The postsurgical course was uneventful. We conclude that aortic root aneurysm is a significant long-term complication after repair of the ascending aorta in patients with Marfan's syndrome, and these patients should be follow-up close and lifelong in terms of development of aneurysm at the level of the residual aorta.

Keywords: Aortic aneurysm; Marfan syndrome

ÖZET Marfan sendromlu hastalarda aorta anevrizması ve diseksiyonu gelişebilme riski yıllardır bilinmektedir. 46 yaşında Marfan sendromlu erkek hasta, geçen yıl göğüs ağrısı ve çarpıntı şikayeti ile kliniğimize başvurdu. Tip 1 aort diseksiyonuna bağlı olarak, 4 yıl önce hemiarcus replasmanı, aort kapak replasmanı ve innominate arter ile prostetik asendan aort arasında greft interpozisyonu yapılmış olan hastamızda, çeklen ekokardiyografi ve bilgisayarlı tomografide aort kökü dilatasyonu (68 mm çapında) ve aort kapak yetersizliği saptandı. Hastamıza, Bentall prosedürü ile başarılı bir şekilde aort kök replasmanı gerçekleştirdik. Postoperatif dönemde komplikasyon yaşanmadı. Aorta kökü anevrizmaları, assendan aorta diseksiyonu nedeniyle opere edilen Marfan sendromlu hastalarda rezidüel aortadan gelişir ve önemli uzun dönem komplikasyonlardır. Bu nedenle hastaların yaşam boyu yakın takibi sağlanmalıdır.

Anahtar Kelimeler: Aort anevrizması; Marfan sendromu

Marfan's Syndrome is a heritable disorder of connective tissue involving the skeletal, cardiovascular, and ocular system. It is characterized with qualitative and quantitative disorders in fibrillin syntese that is constituent glycoprotein of microfibrils, due to the defect of the fibrillin gene (FBN1) localised in chromosome 15.¹ It is an inflammatory disease of unknown etiology involving the aorta, and also causes aortic aneurysm and dissection.

Acute aortic dissection is a life-threatening disease. Priority treatment type is usually an emergency operation aiming at preserving life with minimal operative risk. A number of surgical options can be applied to the aorta and the root, in addition to supracoronary aortic replacement.^{2,3} Complete

resection of the intimal tear and prosthetic replacement of the ascending aorta with re-approximation of the proximal and distal edges are considered standard in acute aortic dissection surgery. Due to aortic disease associated with Marfan's syndrome, secondary aneurysm and dissections may develop in the native aorta after aortic surgery.

We presented a case in which we applied the Bentall procedure due to aortic root dilatation. The ascending aortic replacement had undergone for aortic dissection 4 years ago to the patient. We wanted to emphasize the importance of closely following patients with Marfan's syndrome who underwent aortic surgery.

CASE REPORT

A 46-year-old man who had Marfan's syndrome admitted with complains chest pain and palpitation that occurred in the last year. He had undergone hemiaricus replacement (28 mm prosthetic graft),

aortic valve resuspension and graft interposition (8 mm PTFE graft) between innominate artery and prosthetic ascending aorta due to type I aortic dissection 4 years earlier. Echocardiography revealed aortic root dilatation and advanced aortic valve insufficiency (Figure 1A, B). An aortic root of 6.8 cm in diameter was identified in the computed tomography in addition to descending aortic dissection (Figure 2). Furthermore, 3-D tomography supported the diagnosis of giant aortic aneurysms (68.1 mm) expanding into the left atrium and posterior including the valsalva sinuses, which indicated the necessity for an aortic root replacement. Continuing dissection in descending aorta and a graft anastomosis between innominate artery and prosthetic hemiaricus graft were also observed with this image (Figure 3). We recommended reoperation for the aneurysm. The patient received operation, and adhesions were carefully explored. The old hemiaricus graft was turned with elastic tape. After heparinization, the cardiopulmonary bypass

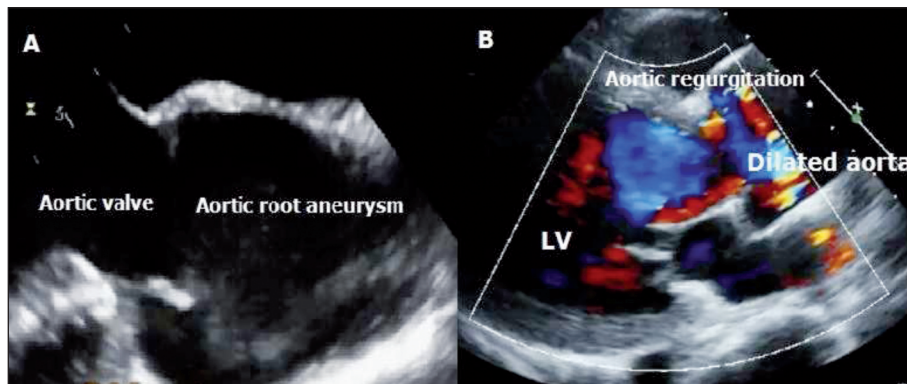


FIGURE 1: Echocardiography shows aortic valve insufficiency and aortic root enlargement.

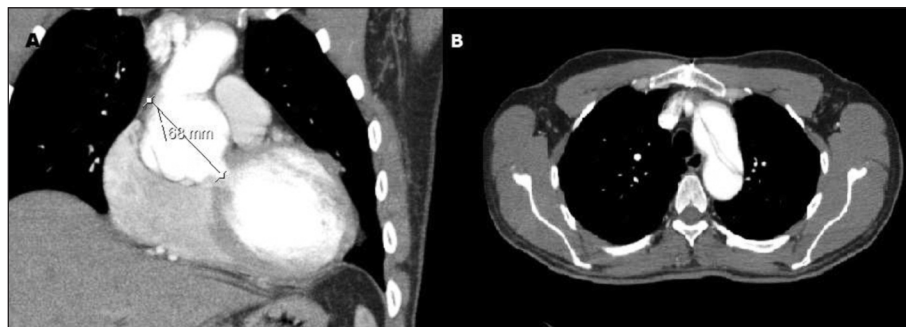


FIGURE 2: Computed tomography showed aortic root enlargement (68 mm in diameter) (A) and descending aortic dissection (B).

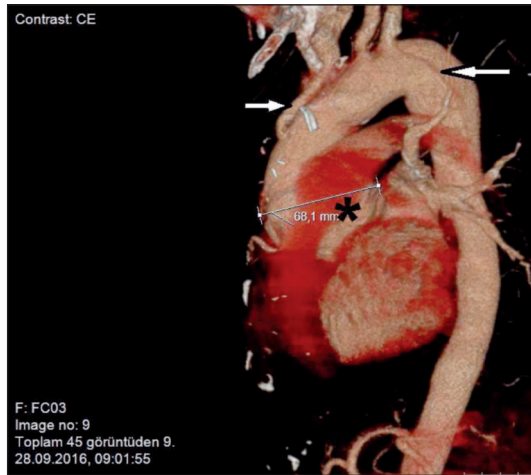


FIGURE 3: Continuing descending aortic dissection due to tip I dissection (big arrow) and the previous re-implantation between innominate artery and prosthetic graft bypass (small arrow) and aortic root aneurysm (asterisk).

was established by using the femoral artery and right atrium. A cross clamp was placed just below the previous innominate artery bypass. Prosthetic aortic graft was incised. The aortic wall of patient was remarkably thickened. The coronary sinuses of valsalva had dilated, and the native aorta and graft anastomosis line stretched. In addition, there were fenestrations on the intimal surface due to intimal damage and aneurysmatic dilatation (Figure 4). Aneurysmatic enlargement occurs through posterior with creating a pressure to left atrium. Aortic root was explored and coronary sinuses were released for root replacement. An aortic root replacement using the modified Bentall procedure was performed with conduit graft. Later, the conduit graft was sewed in old prosthetic graft. Anastomoses strengthened with fibrin glue. Weaning from the CPB was uneventful using a minimal dopamine infusion. Patient was stable without any problems in intensive care unit. The patient had an uneventful recovery and was discharged from the hospital without any complications on the 10th postoperative day. The patient has been followed for 6 months without any problems. A histopathological study of the resected aortic root wall showed a disruption of the elastic fibers in the media, and cystic necrosis with mucopolysaccharide deposits, and these findings were compatible with Marfan's syndrome.

DISCUSSION

The definition, characteristics, clinical findings and treatment options of acute type I dissection are well described in the literature. Most patients have a clinic that carries the complications of the dissection when diagnosed. If surgery is not performed, intra-pericardial rupture is seen in more than 50% of patients with acute I dissection and death can occur within 48 hours.^{4,5} Because of this reason, our patient had undergone a dissection operation 4 years ago in urgent conditions.

Depending on intra-operative, early postoperative and late complications, patients undergoing surgical treatment may be exposed to a re-operation again or die.⁶⁻⁸ Clinical pathologies relation to aortic valve, native aortic tissue and continuing dissections may occur as aortic valve insufficiency, aortic dilatation and new dissection in the late postoperative period. In addition to this, aortic tissue pathologies and its clinic manifestations can often be observed in patients with connective tissue disease as Marfan's syndrome in the late period. An aortic root aneurysm is one of the problems encountered in late term for dissection operations as our case.

Marfan's syndrome is an autosomal dominant condition which may involve the cardiovascular, ocular, skeletal, and other systems. Mutations causing Marfan's syndrome are found in the FBN1 gene, encoding fibrillin-1, an extracellular matrix protein involved in microfibril formation. Ektopia

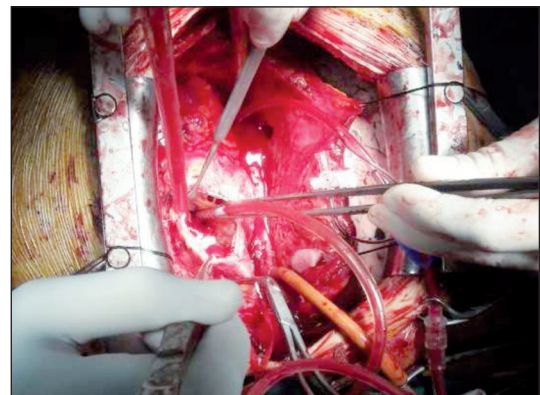


FIGURE 4: The picture shows fenestrations and aortic enlargement in the aortic intimal surface detected in re-operations.

lentis, ascending aortic dilatation, aortic dissection and dural ectasia as major manifestations has been identified for Marfan's syndrome in sinus valsalvas, sino-tubular junction and the proximal aorta.^{9,10} The presence of current connective tissue disease requires strict follow-up in patients treated surgically. As regards diagnostic tools, serial echocardiography, computed tomography, and magnetic resonance imaging are complementary for detecting progressive enlargement with high sensitivity and specificity. Whenever a tendency to root enlargement is diagnosed, one must shorten the interval for the next diagnostic imaging examination to 6 or even 3 months in patients with less aggressive surgical treatment, especially. As our patient with Marfan's syndrome, aneurysm formation may occur at the level of the residual aortic root because of prior replacement of the supracoronary aorta.

Surgeons may prefer supra-coronary aortic replacement and valve resuspension in order to minimize the peri-operative risk in patients with type I aortic dissection.² Other than this, as more aggressive technique, composite graft replacement or valve-sparing procedure may be preferable for aortic root. Although the more extensive surgery may be associated with higher perioperative risk, it eliminates the risk of future aneurismal dilatation and re-dissection of the aortic root or aortic valve incompetence.^{2,11,12} As in our case, only the supra-coronary graft interposition and valve resuspension were performed to reduce the risk of the first operation. But 4 years later, aneurysmatic expansion of the native aortic tissue occurred, and preoperative surgical risk increased significantly in the second operation.

If aortic dissection occurs in a patient with Marfan's syndrome, more aggressive surgical options should be preferred in relation to the aorta and the aortic valve. If a supra-coronary surgical intervention is performed to reduce the risk of surgery, a close and life-long follow-up is absolutely necessary for patients with Marfan's syndrome by echocardiography and computed tomography imaging. In a patient who had previously undergone supra-coronary aortic replacement, the optimal surgical treatment should include a complete resection of the fragile diseased aortic wall if the aortic root aneurysm develops. The modified Bentall procedure with the button technique seems to be the most preferable procedure.

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Conflict of Interest

No conflicts of interest between the authors and/or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: Ümit Arslan, Bilgehan Erkut; **Design:** Eyüp Serhat Çalık; **Control/Supervision:** Eyüp Serhat Çalık, Bilgehan Erkut; **Data Collection and/or Processing:** Ümit Arslan; **Analysis and/or Interpretation:** Ümit Arslan, Bilgehan Erkut; **Literature Review:** Ümit Arslan, Eyüp Serhat Çalık; **Writing the Article:** Ümit Arslan, Bilgehan Erkut; **Critical Review:** Bilgehan Erkut, Eyüp Serhat Çalık.

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